

Manual of Pediatric Anesthesia

Seventh Edition

Jerrold Lerman
Charles J. Coté
David J. Steward
Editors



Springer

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With an Index of Pediatric Syndromes

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Jerrold Lerman, MD, FRCPC, FANZCA

Clinical Professor of Anesthesiology
Women and Children's Hospital of Buffalo,
Department of Anesthesiology, State University of New York
Buffalo, New York, USA

Charles J. Coté, MD, FAAP

Professor of Anaesthesia, Harvard Medical School
MassGeneral Hospital for Children at the Massachusetts
General Hospital
Division of Pediatric Anesthesia, Department of Anesthesia,
Critical Care and Pain Medicine
Boston, MA, USA

David J. Steward, MB, FRCPC

Honorary Professor
Department of Anesthesiology, Pharmacology and Therapeutics
University of British Columbia
Vancouver, British Columbia



Springer

Jerrold Lerman
Buffalo, NY, USA

Charles J. Coté
Quincy, MA, USA

David J. Steward
Blaine, WA, USA

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In the tradition of the first six editions, this seventh edition of the *Manual of Pediatric Anesthesia* is designed as a concise but comprehensive pocketbook guide to pediatric anesthesia practice. We are honored and excited to join with the Springer Publishing family of anesthesia texts to produce this latest edition. Since its inception, this has been a book to be carried in the pocket or available on the desk for handy reference. With this edition, Springer will add an online version for even easier immediate pocketsize reference.

The Manual outlines the important considerations when anesthetizing infants and children, describes management problems, and presents a course of action for treating many of your pediatric patients. Each chapter also directs you to further reading. For the resident in training, it provides a compact but comprehensive source of current information concerning pediatric anesthesia practice.

The three authors have a combined total of more than a century of clinical experience in providing perioperative care for children of all ages having all types of surgical procedures. In recent years, our practices have extended outside the operating room to care for children having a wide variety of medical and minor surgical procedures as well as to provide pain management throughout the hospital.

In preparing this edition of the Manual, we have compiled an evidence-based approach based on the literature and fused this with our own experiences to synthesize optimal clinical strategies for each scenario. The information presented has been reviewed by all three authors and a consensus reached on controversial topics. In many instances, we recommend a course of action for a given clinical situation. When we do this, it is based on what has worked well for us. However, we recognize that others may have different ideas, approaches, and constraints on their practices that may require them to adapt to ensure a successful outcome.

It is now over 35 years since the first edition of this book appeared. Many changes in our practice have occurred during this time. Some occurred in response to the expanding scope of pediatric surgery; others resulted from the

progressive introduction of new anesthesia drugs and technologies, plus new visions of what constitutes optimal pediatric anesthesia care. It is satisfying that many of these changes were a result of the simultaneous proliferation of clinical investigations relating to anesthesia care for infants and children. We are now able to practice evidence-based anesthesia care much of the time.

Yet still, questions remain. The current debate regarding the safety of the use of general anesthetics in the very young child demonstrates that we must constantly be ready to meet new challenges and accept possible new concepts if we are to continue to deliver safe care to our youngest patients.

Children remain the most rewarding patients to manage. There is great satisfaction in the successful management of the tiny infant, the reticent child, or the many other children that you will meet in your practice. We hope that our handbook will help you to achieve this satisfaction.

Buffalo, NY
Quincy, MA
Blaine, WA

Jerrold Lerman
Charles J. Coté
David J. Steward

Abbreviations and Acronyms

A-aDN ₂	Alveolar-arterial nitrogen difference
A-aDO ₂	Alveolar-arterial oxygen tension gradient
ACE	Angiotensin-converting enzyme
ACT	Activated clotting time
ASD	Atrial septal defect
AV	Arteriovenous (AVM arteriovenous malformation)
BiPAP	Biphasic positive airway pressure
BP	Blood pressure
BSA	Body surface area
CBF	Cerebral blood flow
CF	Cystic fibrosis
CHD	Congenital heart disease
CHF	Congestive heart failure
CK	Creatine kinase
Cl	Clearance
CL	Lung compliance
CMRO ₂	Cerebral metabolic rate for oxygen
CNS	Central nervous system
CO	Carbon monoxide
CoHb	Carboxyhemoglobin
CPAP	Continuous positive airway pressure
CPB	Cardiopulmonary bypass
CPK	Creatine phosphokinase
CPP	Cerebral perfusion pressure
CSF	Cerebrospinal fluid
CT	Computed tomography
CV	Closing volume
CVP	Central venous pressure
DDAVP	1-deamino-8-D -arginine vasopressin
DIC	Disseminated intravascular coagulopathy
DN	Dibucaine number
2,3-DPG	2,3-diphosphoglycerate
EACA	Epsilon-aminocaproic acid
EBV	Estimated blood volume
ECF	Extracellular fluid

ECG	Electrocardiogram
ECHO	Echocardiography
ECMO	Extracorporeal membrane oxygenation
ED ₉₅	Effective dose in 95% of patients
EDTA	Ethylenediaminetetraacetic acid
EEG	Electroencephalography
EMG	Electromyography
ETT	Endotracheal tube
EUA	Examination under anesthesia
FESS	Functional endoscopic sinus surgery
FFP	Fresh frozen plasma
FIO ₂	Fraction of inspired oxygen
FN	Fluoride number
FRC	Functional residual capacity
GERD	Gastroesophageal reflux disease
GFR	Glomerular filtration rate
Hb	Hemoglobin
HbA	Adult hemoglobin
HbC	Hemoglobin C
3-HBDH	3-hydroxybutyrate dehydrogenase
HbF	Fetal hemoglobin
HbS	Sickle cell hemoglobin
Hct	Hematocrit
HFOV	High frequency oscillatory ventilation
HME	Heat and moisture exchanger
HpD	Hematoporphyrin derivative
IAP	Intra-abdominal pressure
ICF	Intracellular fluid
ICP	Intracranial pressure
ICU	Intensive care unit
ID	Internal diameter
IVJ	Internal jugular vein
IOP	Intraocular pressure
IPPB	Intermittent positive-pressure breathing
IPPV	Intermittent positive-pressure ventilation
IVAC	Intravenous accurate control [device]
IVH	Intraventricular hemorrhage
KTP	Potassium titanyl phosphate laser
L/S	Lecithin-sphingomyelin [ratio]
LDH	Lactate dehydrogenase
LES	Lower esophageal sphincter
LMA	Laryngeal mask airway

LV	Left ventricle
MABL	Maximal allowable blood loss
MAC	Minimum alveolar concentration
MEP	Motor evoked potential
MetHb	Methemoglobin
MH	Malignant hyperthermia or hyperpyrexia
MHS	Malignant hyperthermia-susceptible
MRI	Magnetic resonance imaging
N ₂ O	Nitrous oxide
NEC	Necrotizing enterocolitis
NGT	Nasogastric tube
NO	Nitric oxide
NSAID	Nonsteroidal antiinflammatory drug
NSF	Nephrogenic systemic fibrosis
OCR	Oculocardiac reflex
OELEM	Optimal external laryngeal manipulation
OLV	One lung ventilation
OR	Operating room
OSA	Obstructive sleep apnea
P ₅₀	PO ₂ with 50% hemoglobin saturation
PA	Pulmonary artery
PACU	Postanesthesia care unit
PaO ₂	Arterial oxygen pressure
PAR	Postanesthesia room
PC	Partition coefficient (λ)
PCA	Patient-controlled analgesia
PCEA	Patient controlled epidural analgesia
pCO ₂	Partial pressure of carbon dioxide (PaCO ₂ arterial carbon dioxide; PetCO ₂ end-tidal carbon dioxide)
PDA	Patent ductus arteriosus
PEEP	Positive end-expiratory pressure
PGE ₁	Prostaglandin E ₁
PIP	Peak inspiratory pressure
PNF	Protamine neutralization factor
pO ₂	Partial pressure of oxygen
PONV	Postoperative nausea and vomiting
PPIA	Parental presence at induction of anesthesia
PPM	Parts per million
PRBC	Packed red blood cells
PSV	Pressure support ventilation
PT	Prothrombin time
PTT	Partial thromboplastin time

PVC	Polyvinyl chloride
PVOD	Pulmonary vascular obstructive disease
PVR	Pulmonary vascular resistance
\dot{Q}	Perfusion
Qp:Qs	Ratio of pulmonary to systemic blood flow
RA	Right atrium
RAE	Ring, Adair, Elwyn
RAST	Radioallergosorbent testing
RBC	Red blood cell
RDS	Respiratory distress syndrome
REM	Rapid eye movement
RES	Reticuloendothelial system
ROP	Retinopathy of prematurity
RSI	Rapid sequence induction
RV	Right ventricle
SaO ₂	Arterial oxygen saturation
SBE	Subacute bacterial endocarditis
SCIWORA	Spinal cord injury without radiologic abnormality
SGOT	Serum glutamic oxaloacetic transaminase
SIADH	Syndrome of inappropriate antidiuretic hormone secretion
SNP	Sodium nitroprusside
SpO ₂	Saturation pulse oximetry
SSEP	Somatosensory evoked potentials
SVR	Systemic vascular resistance
TCI	Target controlled infusion
TEE	Transesophageal echocardiography
TEF	Tracheoesophageal fistula
TGA	Transposition of the great arteries
TIVA	Total intravenous anesthesia
TLC	Total lung capacity
URTI	Upper respiratory tract infection
V _A	Alveolar ventilation
V _D	Dead space volume
V _t	Ventilation
$\dot{V}O_2$	Rate of metabolism (or consumption) for oxygen
(\dot{V}/\dot{Q})	Ventilation-perfusion [matching]
V _t	Tidal volume
VACTERL association	The VATER association with added cardiac and limb defects
VAE	Venous air embolism

VATER association	Vertebral defects, anal atresia, tracheoesophageal fistula, esophageal atresia, radial and renal dysplasia
VATS	Video-assisted thorascopic surgery
VILI	Ventilator induced lung injury
VIP	Vasoactive intestinal polypeptide
VSD	Ventricular septal defect
VTV	Volume targeted ventilation
WBC	White blood cell

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Foundations of Pediatric Anesthesia

PSYCHOLOGICAL ASPECTS OF ANESTHESIA FOR CHILDREN

Hospitalization and/or medical procedures can have profound emotional consequences for infants and children. Some children demonstrate behavior disturbances that persist long after the event. The extent of the upset is determined by several factors, the most important of which is the child's age.

Infants younger than 6 months of age are not upset by separation from parents and readily accept a nurse as a substitute mother. From a psychological viewpoint, this is probably a good age for major surgery, although prolonged separation may impair parent-child bonding.

Older infants and young children (6 months to 5 years) are much more upset by a hospital stay, especially with separation from family and home; ambulatory surgery is much less upsetting. Separation of a preschool-age child from their parents at the time of surgery, even ambulatory surgery, is a stress that requires consideration. Explanations of procedures and the need for them are difficult at this age, and, not surprisingly, these children show the most severe behavior regression after hospitalization.

School-age children are usually less upset by separation and more concerned with the surgical procedure and its possible mutilating effect. They often have the wildest misconceptions of what their surgery involves. In contrast, adolescents fear the process of narcosis, the loss of control, of waking up during the surgery, and the possibility of not being able to face the process calmly. It is for these reasons that providing as much information as possible is essential, along with assuring them that they will not awaken during anesthesia and feel the pain of surgery and that they will awaken at the end.

The type and extent of the surgery is an important factor. Major surgery, craniofacial surgery, and amputation of a limb are especially distressing, and appropriate psychiatric support is essential. Surgery of the genitalia in particular may have important psychological implications in children over 18 months of age.

Factors other than age also influence the child's emotional response. For example, a prolonged hospitalization is much more disturbing than a brief admission, although the former has been mitigated in part by having parents "live in" with their children during hospital stays. Ambulatory surgery usually has a negligible emotional effect on most children, whereas repeated hospitalizations and surgeries may cause significant psychological disturbance; a previous bad experience may be long remembered.

Children vary in their responses to impending hospitalization or medical intervention. Some seek information and participate keenly in preparation programs; they have an active coping style. These children are likely to benefit from psychological preparation and can be expected to cooperate. Others maintain an air of disinterest; they have an avoidant coping style (the "silent child"). Children in the latter group may not benefit and indeed may be further sensitized by efforts at psychological preparation. They may benefit more from an effective anxiolytic premedication (see later discussion).

Psychological Preparation

Preoperative psychological preparation is very important and has been clearly demonstrated to benefit the children. In most cases, the parents prepare the children for surgery, although the extent to which the children can be prepared is determined by the child's age. The basic objective is to explain to the child in simple, understandable, and reassuring terms what will happen at the hospital. Older children and adolescents should be prepared well in advance, as soon as hospitalization is arranged. Younger children should not be prepared too far in advance—it is unnecessary and will be a continuing source of worry for them. Rather, they should be prepared a day or so beforehand.

Hospital tours, puppet shows, and/or audiovisual presentations should be available, as all have been shown to be beneficial. Videotapes are most useful and may be loaned to parents. In some centers, prehospital preparation programs for children have been televised via community television stations on a weekly basis. In this way, a whole population of children can be prepared for the possibility of hospitalization, rather than just those scheduled for surgery. Many older children (older than 5 years) benefit from preoperative information about the operation itself, how much it will hurt afterward and for how long, and what will be done to ease the pain.

Meeting with the Parents

Being unable to choose parents for your patients, you must make do with those who come with the child ... ; it would be abnormal if they showed no anxiety

Mellish (1969)

Parents are playing an increasingly active role in the perioperative care of their children; many expect to be present at induction of anesthesia and in the recovery room. However, some parents are more anxious than others, and this is readily perceived and may further upset the child. Good preparation of the parents reduces parental anxiety and indirectly helps the child.

There are many factors that influence the extent of parental anxiety when a child requires surgery. Even parents of children with only minor problems may initially be very anxious. Complete explanations and good communication with the medical and nursing teams usually do much to reduce their anxiety level. In particular, it is important to describe to parents how their child could respond during induction of anesthesia (eyes rolling up, movements of the arms and legs, and turning of the head) and to reassure them that these are normal and expected responses.

Obtaining Consent

The anesthesiologist is placed in a difficult situation when obtaining informed consent for general anesthesia; providing information on all the potential risks before a minor surgical procedure might well be expected to increase the level of anxiety of the parents. Parents benefit from an appropriate discussion of the risks of anesthesia in that this fulfills their own needs of responsibility and understanding. The parents should be permitted to dictate the extent of the information they wish to be given. Most parents of healthy children having minor procedures accept that there are risks and prefer to have the opportunity to discuss these risks. Such discussions should, of course, be outside the earshot of the young child.

In general, the anesthesiologist should rely on some well-established general principles in dealing with anxious parents. An approach that has been found most helpful in decreasing parental anxiety is one built on genuine warmth and friendliness, empathy, and understanding. Parents like to be listened to; discussions should allow ample time for questions and for the parents to express their concerns and ideas about the child and the proposed anesthesia process. Parents are reassured after a clear and thorough discussion of the plan for preoperative anxiolysis, how their child will be anesthetized, monitored (and the information

the monitors will provide), and provided with postoperative pain relief. A videotaped explanation may be helpful, but should be augmented by a personal interview. An overall discussion of risks, in particular those specific to their child, helps to place risk in perspective. Assurance that their child's anesthesia will be specifically designed with their child's safety and the surgeon's needs in mind also helps to relieve anxiety. Every parent will be pleased if you communicate the message, "We will design your child's anesthetic with his/her medical issues and the needs of the surgeon in mind. We will be with your child at all the times during the anesthetic!" (see Chap. 4 for further discussion re: consent).

Parental Presence at Induction

Although many parents express a desire to be present at induction of anesthesia for their child, a large body of evidence has demonstrated that while parental presence at induction of anesthesia (PPIA) may reduce parental anxiety, it has either no effect or less effect than other strategies (i.e., midazolam) in reducing the child's anxiety. Practitioners and institutions vary widely in embracing PPIA; some only permit PPIA under special circumstances, whereas others allow all parents to be present. The entire surgical team must endorse adopting PPIA. We believe that parents should be educated regarding their role and responsibilities at the induction through a presurgical education program (e.g., by video demonstration or attending a class). Such a practice will reduce disruptions and outbursts at induction, at a time when all attention should be focused on the child. Certainly, many parents of handicapped children or of children with life-threatening disease (e.g., cancer) can often be of great assistance to the anesthesiologist during the induction. Many parents are calm and supportive of their child and benefit from participating in the induction process. Having the child sit on their parent's lap while the parent hugs their child and holds their arms at the side allows the parents to feel that they are part of the process in a supportive way, while a mask is gently applied to the child's face.

The overanxious parent requires special consideration. Excessive anxiety is often of multifactorial origin and may not be entirely related to the child's present surgical condition. These parents may not reduce their anxiety levels from additional information about the forthcoming procedure. An anxious parent who insists on remaining with the child may do more harm than good and may increase the child's anxiety level. Such anxious parents should be counseled and excluded if possible. Adequate preoperative sedation of the child may help them to agree to this course. Certainly, there is no benefit for parental presence for a neonate or infant who is not fearful of strangers due to their young developmental level or for the child in need of a rapid sequence induction. It must be clear

that parental presence during induction is a privilege given at the discretion of the anesthesiologist in what you deem to be for the best interests of the child and the child's safety.

The Anesthesiologist and the Child

Anesthesia, and particularly the induction period, is recognized to have the potential to cause psychological trauma. Studies indicate that anesthesiologists vary in their ability to relate to children and minimize this upset. An *empathic approach* to the child before and during medical procedures is preferred (e.g., “*This may be a little uncomfortable and I know you are scared, but we are going to do all we can to help and it will soon be over. We don’t mind if you cry*”). The alternative *directive approach* (“*Hold still and be big and brave*”) is generally condemned.

Premedication with an oral anxiolytic is beneficial in decreasing anxiety during separation, increasing cooperation during induction, and decreasing post hospitalization behavior disturbance.

Caution should be exercised in caring for the silent child who has an avoidant coping style, especially the child who must undergo repeat procedures because such a child may not respond as well to routine preparation methods. Some may respond more favorably if they are allowed to continue with their avoidant coping pattern but are given a well-chosen preoperative medication.

Preparing Infants and Children for an Operation

1. Try to meet the young child with the parents so that the child can see them accept you.
2. Direct most of your attention at all times to the child, even if he or she is developmentally delayed. Try to maintain eye contact; it helps to sit alongside the child, on the floor if necessary.
3. Talk to the child in simple terms that the child can understand. Children who are old enough to understand and who have not had surgery previously should be informed that they will feel they were anesthetized for as long as it takes to “blink,” but the surgery will be finished.
4. Pay special attention to the silent child and recognize that he or she may be very upset. Consider the use of a suitable sedative premedication if not otherwise contraindicated.
5. Explain all the procedures to be undertaken in clear and simple terms while avoiding unnecessary and alarming details. Some children may ask about the operation: try to help them understand what is to be done, using

drawings if necessary. Older children in particular may be scared and grossly overestimate the extent of the procedure. They must be reassured, for instance, about the small size of the incision. They may benefit from pre-medication (see p. 52).

6. Do not use the phrase “put you to sleep”—this may worry some children if they recall a family pet that never came back! It may also cause them to worry that they might wake up from their “sleep” when the operation starts or while it is still in progress.
7. Do not present the child with unpleasant and difficult choices. For example, avoid questions such as, “*Do you want the needle or the mask?*” Tell the child what you intend to do and then try to meet any special requests (e.g., “*I do not want a needle, I want to go to sleep with the mask*” or “*I’d like to hold the mask myself*”). For inhalational inductions, let the child select a flavor to apply to their anesthesia mask.
8. Avoid uncovering the child more than necessary to complete the physical examination; many children get upset at being disrobed.
9. Many distraction strategies reduce children’s anxiety preoperatively as well as premedication including iPad tablets, hand-held computer games, television and cartoons, video glasses, music, and magic. Clowns are also effective but are much less practical for many facilities. Allow the child to continue using the distraction strategy until anesthesia is induced.
10. Allow the young child to bring a favorite toy or other security object to the operating room (OR). Label the toy with the child’s name; if it is a doll, suggest that perhaps the doll should also get a cast or a dressing applied during the operation. If the child is able, let him or her walk hand in hand to the OR rather than be carried or wheeled: children are quite independent and feel more at ease walking.
11. If appropriate, allow those parents who are judged to be calm and supportive to accompany their child during the induction. If this is not possible, both the child and the parents may be helped by premedicating the child (e.g., oral midazolam, see p. 52). The parents are much more satisfied if their child separates from them very well sedated. It is sometimes useful to start an intravenous infusion away from the OR with the parents present, especially for handicapped or developmentally delayed children. The intravenous route can then be used for both premedication (e.g., midazolam) and/or induction of anesthesia as soon as the child is in the OR. Some prefer to use local analgesia to insert the intravenous cannula; topical anesthetic cream is ideal if it can be applied well in advance (see p. 634).
12. Select the most appropriate induction technique for each child and proceed without delay. Do not allow the child to wait on the OR table longer than is absolutely necessary to apply the basic monitors: a pulse oximeter may be

- the only monitor that can be maintained during induction of anesthesia in a combative child.
13. Talk to the child throughout to explain or distract him or her as “magic laughing gas” (70 % nitrous oxide) is administered. Add 8 % sevoflurane to complete the induction.
 14. Ensure that all extraneous noises and conversations are excluded during this time. Only one person should be talking to the child. Quiet soothing music may help.
 15. An empathic approach should be used to prepare the child. Small children who are crying during venipuncture can often be calmed by telling them, *“We will put on a Band-Aid in a minute.”*
 16. Tell the child what to expect during the recovery, where recovery will take place, what discomfort they may experience, and for how long. Carefully explain such items as eye patches, nasogastric tubes, and catheters as necessary and that they will be inserted while the child is anesthetized. A urinary catheter may look like a giant worm to an unprepared child! Assure the child that any pain will be treated.
 17. Discuss in detail the plan for postoperative pain relief with both the family and the child.

Postoperative Care

The parents should be reunited with their child as soon as it is practical—before the child awakens, if possible. It is imperative that the parents are cautioned that if an emergency occurs in the recovery room, they will be returned to the parent waiting room until it is appropriate for them to return. Every effort should be made to provide good, but safe, analgesia. Regional nerve blocks, opioid infusions, patient-controlled analgesia, epidural opioids, and all ancillary techniques used in adults should be considered, discussed with the parents, and provided for infants and children when appropriate. A discussion of what the parents can expect postoperatively at home and the need for postoperative oral analgesics is also important.

In the intensive care unit (ICU), the child’s problems are similar to those for adults: pain, lack of sleep, and, later, boredom. In addition, children have their own special concerns, such as separation from the family. Special attention should be directed to pain relief, regular visitation by the parents, and provision of toys, games, and other distractions (e.g., television, computer games) as the child’s condition improves. Parents of children in the ICU benefit by being kept very well informed of their child’s condition and progress, and they must also be continuously updated on the treatment plans for their child.

Suggested Reading

- Banchs RJ, Lerman J. Preoperative anxiety management, emergence delirium, and postoperative behavior. *Anesthesiol Clin.* 2014;32:1–23.
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Anatomy and Physiology

CENTRAL NERVOUS SYSTEM

The central nervous system in the neonate differs from that in the older child: the myelination of nerve fibers is incomplete, and the cerebral cortex is less developed; its cellular elements continue to increase during the first years of life. Reflexes not seen in older children may be elicited. Neonatal spinal reflexes are more generalized and the threshold is lower. In the preterm, brainstem-evoked potentials are prolonged, and the normalization of these coincides with maturation of respiratory control mechanisms.

Anesthesia-induced developmental neurotoxicity has been demonstrated in some neonatal animal models, but the significance of this to humans remains uncertain and will be difficult to fully determine. Meanwhile, appropriate anesthesia and analgesia should be provided to all young children.

Sensitivity to Pain

Until quite recently, little was understood of the ability of infants and small children to appreciate pain. As a result, there was an unfortunate tendency to ignore the need for analgesia during painful procedures, even during and after surgical operations. It is now well established that neonates, including those born prematurely, may have increased sensitivity to pain and will react to it with tachycardia, hypertension, increased intracranial pressure, and a neuroendocrine response that exceeds that reported in adults. Infants demonstrate measurable behavioral responses to pain (e.g., crying, grimacing, restlessness); these responses have been used as a basis for pain scoring systems. Evidence suggests that infants who are subjected to painful procedures (e.g., circumcision) without adequate analgesia may experience an increased sensitivity to pain as older children. This has been attributed to the persistence of alterations in the infant's central processing of painful stimuli. Control of intraoperative and postoperative pain, by modifying stress responses, may possibly even improve survival in infants with critical illness. It is for these reasons that we provide optimal analgesia or anesthesia for all infants and children during and after *any* painful

procedure with the same care as we do for adults. As children grow from infancy into early childhood, their pain threshold remains reduced compared with that of older children or adults.

CRANIUM AND INTRACRANIAL PRESSURE

The skull is less rigid in infants than in adults. As a result, an increase in the volume of its contents—blood, cerebral spinal fluid (CSF), and brain tissue—can be accommodated to some extent by expansion of the fontanelles and separation of the suture lines. Palpation of the fontanelles can be used to assess intracranial pressure in infants. Intracranial pressure (ICP) increases with age ranging from 0–6 mmHg in infants to 13–15 mmHg in adolescents.

CEREBRAL BLOOD FLOW AND INTRAVENTRICULAR HEMORRHAGE

Autoregulation of cerebral blood flow (CBF) is impaired in sick neonates, rendering CBF pressure dependent. The threshold below which autoregulation of CBF is impaired is unknown, although some hold that this threshold (mean BP) is the gestational age in mmHg. Hypotension may lead to cerebral ischemia, and pressure fluctuations are transmitted to the capillary circulation. In the preterm infant, the cerebral vessels are very fragile, especially in the region of the germinal matrix overlying the caudate nucleus. Rupture of these vessels leads to intracerebral hemorrhage, which often extends into the ventricular system as an intraventricular hemorrhage (IVH).

Small preterm infants are very prone to IVH, which usually occurs during the first few days after birth and is a leading cause of mortality and morbidity; survivors have a high incidence of cerebral palsy, mental retardation, hydrocephalus, and psychiatric disorders. Potential predisposing factors to IVH include hypoxia, hypercarbia, hypernatremia, fluctuations in arterial or venous pressure, low hematocrit, overtransfusion, and rapid administration of hypertonic fluids (e.g., sodium bicarbonate, dextrose).

The anesthesiologist should avoid precipitating these factors in the small preterm infant: airway manipulations, including awake tracheal intubation, and suctioning have been shown to increase blood pressure and anterior fontanelle pressure to similar extents, similar to coughing as well. “Awake” intubation should be avoided whenever possible (even though a definite relationship between “awake” tracheal intubation and IVH has never been established). If the airway is known or appears to be difficult to secure or maintain by facemask, then an “awake” or preferably sedated tracheal intubation is a reasonable choice.

If an awake tracheal intubation is planned, topical analgesia to the mouth and palate (using weight-appropriate doses of local anesthetic [see p. 142]) may attenuate the infant's physiologic responses.

To further prevent blood pressure fluctuations during surgery or painful procedures, adequate anesthesia and analgesia should be provided. Rapid injections of undiluted hypertonic solutions, such as dextrose or sodium bicarbonate, should be avoided. Care should be taken to replace blood losses accurately. Severe anemia and/or coagulopathy should be corrected promptly. Periventricular leukomalacia is a major feature of persistent brain damage in small preterm infants and may follow IVH or may be a direct consequence of prematurity, hypoxia, ischemia, inflammation, and chronic persistent hypotension. Indomethacin therapy may reduce the incidence of severe IVH.

CEREBROSPINAL FLUID AND HYDROCEPHALUS

The CSF, which occupies the cerebral ventricles and the subarachnoid spaces surrounding the brain and spinal cord, is formed by choroid plexuses in the temporal horns of the lateral ventricles, the posterior portion of the third ventricle, and the roof of the fourth ventricle. Meningeal and ependymal vessels and blood vessels of the brain and spinal cord also contribute small volumes of CSF.

The choroid plexuses are cauliflower-like structures consisting of blood vessels covered by thin epithelium through which CSF continuously exudes. The rate of CSF formation is variable, increasing in the first two years to ~ 0.13 mL/min. Except for the active secretion of a few substances by the choroid plexus, CSF is similar in composition to interstitial fluid.

CSF flow is initiated by pulsations in the choroid plexus. From the lateral ventricles, CSF passes into the third ventricle via the foramen of Monro and along the aqueduct of Sylvius into the fourth ventricle, each ventricle contributing more fluid by secretion from its choroid plexus. CSF then flows through the two lateral foramina of Luschka and the midline foramen of Magendie into the cisterna magna and throughout the subarachnoid spaces. CSF is reabsorbed into the blood by hydrostatic filtration through the arachnoid villi, which project from the subarachnoid space into the venous sinuses. Hydrocephalus is an abnormal accumulation of CSF within the cranium that may be either obstructive or nonobstructive.

Obstructive hydrocephalus is caused by a blockage in the flow of CSF. It may be communicating (e.g., when the CSF pathway into the subarachnoid space is open, as after chronic arachnoiditis) or noncommunicating (e.g., when the fluid's pathway proximal to the subarachnoid space is obstructed, as in aqueductal stenosis or Arnold-Chiari malformation).

Nonobstructive hydrocephalus is caused by a reduction in the volume of brain substance, with secondary dilation of the ventricles; by overproduction of CSF (e.g., in choroid plexus papilloma); or by diminished reabsorption of CSF due to scarring.

EYES

Retinopathy of Prematurity

Retinopathy of prematurity (ROP), which is caused by retinal vessel proliferation and retinal detachment, is a leading cause of blindness. Improved survival of very-low-birth-weight infants increased the incidence of this condition. ROP is most common in neonates weighing less than 1500 g. The role of increased oxygen levels in the blood in the etiology of ROP has long been recognized. Limiting the inspired oxygen concentration reduces ROP but, if excessive, may increase both mortality and the incidence of necrotizing enterocolitis. The development of ROP is now considered to progress in stages. Preterm birth exposes the retina to oxygen levels in the blood that are greater than those during intrauterine development, which decreases local vascular growth factors, interrupting the orderly vascularization to the retinal periphery. In the second phase, the peripheral retina becomes metabolically active but with inadequate perfusion. Hypoxic ischemic damage ensues. Inspired oxygen levels in the blood accelerate this process and compound the situation by high levels of damaging “reactive oxygen species.”

The clinical factors associated with ROP include hyperoxia, hypoxia, hypercarbia, or hypocarbia, blood transfusion, recurrent apnea, sepsis, and other systemic illness. Occasionally, ROP occurs in infants who have never been given supplemental oxygen and even in infants with cyanotic congenital heart disease. A major multi-institutional study of tight control of oxygen administration failed to reduce the incidence of ROP. Nevertheless, the inspired oxygen concentration should be carefully controlled in all preterm infants to prevent unnecessary hyperoxia. Large fluctuations in inspired oxygen concentrations should be avoided. The safe level of PaO_2 is now considered to be 50–70 mmHg. Monitoring oxygen saturation at a preductal site (right hand or earlobe), maintaining the arterial oxygen saturation (SaO_2) at 90–95 %, and avoiding large fluctuations in oxygen levels are recommended. On occasion, it may still be necessary to err on the side of safety and administer greater inspired oxygen concentrations. Major surgical procedures do not appear to predispose infants to ROP.

RESPIRATORY SYSTEM

The respiratory system is of special interest to the anesthesiologist since this is the route of administration of inhaled anesthetic agents, and its functions may be significantly compromised before, during, and after anesthesia. Changes in the respiratory system occur continuously from infancy to about age 12 years as the child grows to maturity.

Anatomy

There are major anatomic differences in the airway between the neonate, the small infant, and the adult that are important for the safe conduct of anesthesia:

1. The head is relatively large and the neck is short, which may contribute to difficulty in laryngoscopy.
2. The tongue is relatively large and readily blocks the pharynx during and after anesthesia; therefore, an oropharyngeal airway or a jaw thrust is often needed. The large tongue may also hamper attempts to visualize the glottis at laryngoscopy.
3. The nasal passages are narrow and easily blocked by secretions or edema, which may cause serious problems. Neonates were previously described as “obligate nose breathers,” but whether this is always true has been questioned. It is certain that some neonates may not immediately convert to mouth breathing if the nasal passages are obstructed. Upper airway obstruction is more likely to occur when the neck is flexed. The anesthesiologist should extend the infant’s neck after extubation until the infant completely recovers from anesthesia. Insertion of a nasogastric tube (NGT) increases total airway resistance; if the nares are of unequal size, the NGT should be inserted into the smaller nostril where it will have the least adverse effect.
4. The larynx is situated more cephalad (C3–C4) and anteriorly, and its long axis is directed inferiorly and anteriorly. The high cervical level of the larynx in the infant means that elevating the head to the “sniffing position” during laryngoscopy will not assist in visualizing the glottis as it does in the adult. In the infant, if the head is elevated, the larynx also moves anteriorly (see p. 94).
5. The airway is narrowest at the level of the cricoid cartilage just below the vocal cords. This cartilage is the only solid circumferential component of the airway. The lining of the airway here is pseudostratified, ciliated epithelium that is loosely bound to the underlying tissue. Trauma to these tissues causes edema, and even a small amount of circumferential edema significantly

compromises the already small cross-sectional area of the infant's airway. The net effect is an increase in the resistance to airflow (which is turbulent [i.e., Reynold's number >2100]) resulting in stridor, where $R \propto 1/r^5$ and where R is the resistance and r is the radius of the airway. Insertion of an ETT similarly encroaches on this space, as the internal diameter of the ETT becomes the area for gas flow; hence, airway resistance increases. This is more significant in smaller children (<3 years). This is the reason that thinner-walled uncuffed tubes used to be advocated in pediatric cases. Today, the wall thickness of cuffed and uncuffed tubes is the same.

6. The epiglottis is relatively long and stiff. It is U shaped and projects posteriorly at an angle of approximately 45° above the glottis. Often, it must be lifted by the tip of a laryngoscope blade before the glottis can be seen. For this reason, a straight-blade laryngoscope is recommended for use in infants and children, although a properly used Macintosh blade may provide a similar view.
7. The trachea is short (approximately 5 cm in the neonate), so precise placement and firm fixation of the ETT are essential. The tracheal cartilages are soft and can easily be compressed by the fingers of an anesthesiologist holding a mask or collapsed (dynamic compression) by the infant's vigorous attempts to breathe against an obstructed airway.
8. The right main bronchus is larger than the left and is less acutely angled at its origin. For this reason, if the ETT is advanced too far, it almost invariably enters the right main bronchus. This most often occurs while taping the tube in place or with changes in position for surgery after tracheal intubation. Flexion of the head advances the tip of the tube. It is therefore essential to reassess the equality of bilateral breath sounds after repositioning for surgery (see Chaps. 4 and 8).
9. Because the ribs are almost horizontal, ventilation is mainly diaphragmatic. The abdominal viscera are bulky and can hinder diaphragmatic excursion, especially if the gastrointestinal tract is distended.

Table 2.1 lists the approximate airway dimensions in infants and children.

Physiology

Breathing movements begin in utero and are characteristically rapid, irregular, and episodic during late pregnancy. Normally, they are present 30 % of the time in the third trimester, vary with the sleep state of the fetus, and are subject to diurnal variation. Fetal breathing movements may play a role in lung development and provide exercise for the muscles of respiration. Monitoring of these movements may provide information on fetal health: hypoxemia leads to a

Table 2.1 Approximate airway dimensions in infants and children

Age (year)	Tracheal length (cm) (vocal cords to carina)	Trachea (AP diameter (mm))	Diameter (mm)	
			Right bronchus	Left bronchus
<0.5	5.9	5.0	4.5	3.5
0.5–1	7.2	5.5	4.8	3.7
1–2	7.5	6.3	5.1	3.9
2–4	8.0	7.5	6.4	4.9
4–6	8.6	8.0	6.7	5.3
6–8	9.5	9.2	7.9	6.1
8–10	10	9.75	8.4	6.5
10–12	11.5	10.5	9.2	6.8
12–14	13.5	11.5	9.8	7.5
14–16	14.5	13.2	11.5	8.8

AP anteroposterior

decrease in fetal breathing, and severe hypoxemia leads to gasping movements. The fetal lungs are filled with fluid, which is moved by this respiratory muscle activity. After 26–28 weeks of gestation, production of surface-active substances (surfactants) is established in the type II pneumocytes. Surfactant is secreted into the lung and can be detected in amniotic fluid samples, providing a diagnostic index of lung maturity and hence neonatal prognosis.

Passage of the fetus through the birth canal compresses the thorax, forcing much of the fluid from the lungs through the nose and mouth. On delivery, this compression is relieved and some air is sucked into the lungs. Peripheral (cold, touch, temperature, etc.) and biochemical (respiratory and metabolic acidosis) stimuli have been thought to initiate the onset of regular, continuous breathing. Other factors may be important, such as an increase in the PaO_2 or removal of central biochemical inhibitors. The first few spontaneous breaths are characterized by increased transpulmonary pressures (more than 50 cm H₂O). They establish the functional residual capacity (FRC) of the neonate's lungs. Remaining lung fluid is removed over the first few days of life by the pulmonary lymphatics and blood vessels. Infants who are delivered by cesarean section are not subjected to the same thoracic squeeze and may have more residual fluid in the lungs. This may cause them to have transient respiratory distress (transient tachypnea of the newborn).

The stability of the alveolar matrix in the neonate depends on the presence of adequate amounts of surfactant, which may be deficient in the preterm infant. Lack of surfactant leads to collapse of alveoli, maldistribution of ventilation, impaired gas exchange, decreased lung compliance, and increased work of breathing (i.e., RDS). Not surprisingly, pneumothorax occurs more commonly during the neonatal period than at any other age.

Control of Respiration in the Neonate

Control of respiration, which involves biochemical and reflex mechanisms, is well developed in the healthy full-term neonate but exhibits several differences from adults. Respiration in the infant in relation to body mass is greater for any given arterial carbon dioxide tension (PaCO_2), reflecting a greater metabolic rate. The ventilatory response to hypercapnia is less in the neonate than in older infants and less still in the preterm neonate. Any increase in the work of breathing is not well sustained. The slope of the carbon dioxide response curve is decreased in infants displaying episodes of apnea, and hypoxemia decreases the response of the neonate to hypercapnia.

The neonate is sensitive to changes in arterial oxygen tension (PaO_2). Administration of 100% oxygen decreases ventilation, indicating the existence of tonic chemoreceptor activity. The ventilatory response of the neonate to hypoxia is modified by many factors, including gestational and postnatal age, body temperature, and sleep state. Preterm and full-term infants younger than 1 week of age who are awake and normothermic usually demonstrate a biphasic response to hypoxemia—a brief period of hyperpnea followed by ventilatory depression. Hypothermic infants and very small preterm infants respond to hypoxemia with ventilatory depression without the initial hyperpnea. This depression of ventilation has been attributed to the central effects of hypoxia on the cortex and medulla. The peripheral chemoreceptors, although active in neonates, are unable to maintain a significant influence on this hypoxic response. Infants show a less sustained response to hypoxia during REM sleep. In neonates, hypoxia also depresses the ventilatory response to carbon dioxide. Hypoxia may induce periodic breathing in infants, and this may be abolished by oxygen administration. Full-term infants older than 2–3 weeks of age demonstrate hyperpnea in response to hypoxia, probably as a result of maturing of the chemoreceptor function.

Reflexes that arise from the lung and chest wall are probably more important in maintaining ventilation in the neonate, possibly compensating for inadequacies in other control mechanisms. They primarily determine the rate (f) and V_t but are also important for maintaining lung volume (i.e., terminating expiration). The Hering-Breuer inflation reflex, which is active in the neonate, is even more powerful in the preterm infant. This reflex disappears during rapid eye movement (REM) sleep and progressively fades during the early weeks of life. The paradoxical “head” reflex, a large inspiration triggered by small lung inflation, is active in the neonate. It may play a role in maintaining lung volumes in the neonate and may be present during anesthesia.

Periodic breathing (rapid ventilation alternating with periods of apnea lasting 5–10 s) occurs in many preterm and some full-term infants. It is associated with increased peripheral chemoreceptor activity. In the preterm infant, the PaCO_2 is greater than normal during these episodes of periodic breathing, but

the heart rate does not change significantly. In the full-term infant, hypocapnia may occur during periodic breathing, which seems to have no serious physiologic consequences and usually ceases by 44 to 46 weeks of postconceptional age. Periodic breathing generally only occurs about 3% of the time in full-term infants; a greater fraction of periodic breathing in a full-term infant is a warning sign of possible abnormal control of ventilation. Some preterm infants demonstrate far more serious and indeed life-threatening episodes of apnea. These commonly exceed 15 s and may be accompanied by bradycardia (possibly due to a chemoreceptor-mediated reflex) and hemoglobin oxygen desaturation. Brief apnea spells (<15 s) may also be accompanied by significant bradycardia (<80 beats/min). The pathogenesis of apnea in preterm infants is not fully understood. Apnea may reflect an immature central respiratory control system because it tends to resolve as the brain matures. However, a variety of pathophysiologic mechanisms are involved. Apneic episodes may result from a failure of central control mechanisms (central apnea); in such instances, there is no ventilatory effort. It may also result from airway obstruction (obstructive apnea), in which case ventilatory effort may be observed, but there is no gas exchange. Obstruction usually occurs in the infant's nasopharynx, pharynx, or hypopharynx. Mixed apnea (a combination of central and obstructive) commonly occurs, and one type may progress to another (i.e., obstructive apnea may progress to central apnea). Apnea may also result from failure of the ventilatory muscles. Many apneic episodes occur during REM sleep, when it is possible that fatigue of the ventilatory muscles is an important factor. Although neonatal apnea may be idiopathic, it may also be symptomatic of an underlying disease process, such as sepsis, intracranial bleeding, anemia, hypoglycemia, hypothermia, sensitivity to sedating medications, or patent ductus arteriosus.

Preterm infants must be carefully monitored to detect apneic episodes. Treatment is by tactile stimulation or, if this fails, by bag-mask resuscitation. The incidence of apneic episodes is decreased by therapy with aminophylline or caffeine (central stimulation) or by institution of continuous positive airway pressure (increased reflex activity of lung and chest wall reflexes and "splinting" of the airway). Preterm and former preterm infants up to 60-week postconceptional age, particularly those with anemia, are at risk for postoperative apnea even if apnea free at the time of anesthesia. These infants will benefit from appropriate postoperative monitoring in an ICU or similar close observation unit with apnea monitoring.

Muscles of Respiration

In the neonate, the muscles of respiration are subject to fatigue, a tendency that is determined by the types of muscle fiber present. In the diaphragm, 10% of the muscle fibers are type I (slow twitch, highly oxidative, fatigue resistant) in preterm

infants, which increases to 25% in full-term infants and reaches a maximum of 55% (the adult level) after 8 months postpartum. In the intercostals, 20, 46, and 65% of the fibers are type I for the same age groups, with the maximum reached by 2 months postpartum. Thus, the preterm infant is prone to ventilatory muscle fatigue, a predisposition that progressively disappears with maturity. Ventilation is also affected by changes that occur during changing sleep states. The preterm infant spends 50–60% of this time in REM sleep, during which time, intercostal muscle activity is inhibited, and paradoxical movement of the soft chest wall occurs. The reduced intercostal muscle activity is offset in part by an increase in diaphragmatic activity. Much of this activity is wasted when the ribs move paradoxically and may lead to diaphragmatic fatigue.

Respiratory Mechanics

The specific lung compliance (CL) increases slowly after birth as fluid is removed from the lung. Chest wall compliance in the infant (especially the preterm infant) is great, so that total compliance approximates CL. This highly compliant chest wall provides a relatively weak force to maintain the FRC and to oppose the action of the diaphragm. The FRC of the small infant is maintained by a rapid respiratory rate, the point of termination of expiration, the controlled expiration (“laryngeal braking”), and the tonic activity of the ventilatory muscles. This being so, it is not surprising that large decreases in FRC occur with apnea and during anesthesia when inhalation agents depress intercostal muscle function.

These large decreases in FRC are accompanied by airway closure and impaired oxygenation. Intercostal muscle inhibition during REM sleep or with inhaled anesthetic agents compounds the weakness of the chest wall and results in paradoxical movement. This paradoxical chest wall movement is markedly augmented by any airway obstruction. It may be inferred that infants generally require controlled ventilation during anesthesia and benefit from rapid respiratory rates or the use of positive end-expiratory pressure to maintain the lung volume and avoid airway closure and pharyngeal collapse during mask ventilation. As the child grows through infancy and childhood, the rib cage stiffens so that it becomes better able to oppose the action of the diaphragm and less reliant on intercostal muscle tone.

The transpulmonary pressures needed to optimally inflate the lungs are remarkably similar in healthy infants, children, and adults. During artificial ventilation, peak inspiratory pressures of 15–20 cm H₂O are normal.

The nasal air passages contribute up to 50% of the total airway resistance in infants and slightly less in African-American infants. Insertion of an NGT increases this resistance by as much as 50%. The nasal passages are usually of unequal size; if an NGT is inserted, it should be placed through the smaller

nostril, so as to have a lesser effect on total nasal airway resistance. The resistance of the neonate's peripheral airways is small but increases with age.

Lung Volumes

In the full-term infant, total lung capacity (TLC) is approximately 160 mL; the FRC is about half this volume. The tidal volume (V_t) is approximately 16 ml (6–7 mL/kg), and dead space volume (V_D) is about 5 mL (30 % of the V_t). Relative to body size, all of these volumes are similar to adult values. Note, however, that any dead space in anesthesia or ventilator circuits is much more significant in relation to the small volumes of the infant (e.g., a 5-mL apparatus dead space would increase the total effective V_D by 100 %).

In contrast to the static lung volumes, alveolar ventilation (V_A) is proportionally much greater in the neonate (~100–150 mL/kg/min) than in the adult (~60 mL/kg/min). This high V_A in the infant results in a V_A :FRC ratio of 5:1, compared with 1.5:1 in the adult. Consequently, the FRC is a much less effective “buffer” in the infant, so that changes in the concentration of inspired gases (including anesthetic gases) are more rapidly reflected in alveolar and arterial values.

The closing volume (CV) is relatively greater in infants and young children than in young adults; it may exceed the FRC to encroach on the V_t during normal respirations. Airway closure during normal respirations may explain the reduced normal values for PaO_2 in infants and neonates (Table 2.2). A decrease in FRC, which usually occurs during general anesthesia and persists into the postoperative period, further increases the significance of the large CV and increases the alveolar-arterial oxygen tension gradient ($A\text{-}a\text{DO}_2$). The younger the infant or child, the greater the decrease in FRC. The intraoperative decrease in FRC may be partially reversed by continuous positive airway pressure.

The total surface area of the air-tissue interface of the alveoli is small in the infant (2.8 m^2). When this area is related to the high metabolic rate for oxygen, it is apparent that the ratio between surface area and rate of oxygen consumption is smaller in the infant than the adult. As a result, the infant has a reduced reserve capability for gas exchange. This developmental fact assumes greater significance in the presence of congenital pulmonary hypoplasia or lung damage (e.g., from meconium aspiration). In such cases, the remaining healthy lung tissue may be inadequate to sustain life.

Table 2.2 Arterial oxygen tension in healthy infants and children

Age	Normal arterial oxygen (mmHg) in room air
0–1 week	70
1–10 months	85
4–8 years	90
12–16 years	96

Work of Breathing

The muscles of respiration generate the force necessary to overcome the resistance to airflow and the elastic recoil of the lungs and chest wall. These two factors dictate an optimal rate of ventilation and a V_t that delivers a given V_A while expending minimal muscular energy for each child. Because the time constant of the infant's lung is relatively small, efficient alveolar ventilation can be achieved at high respiratory rates. In the neonate, a respiratory rate of 37 breaths/min has been calculated to be most efficient, a rate that is close to the rate in healthy neonates. Full-term infants are similar to adults in that they require 1% of their metabolic energy to maintain ventilation; the oxygen cost of breathing is 0.5 mL / 0.5 L of ventilation. The preterm infant has a greater oxygen cost of breathing (0.9 mL/0.5 L), which is greatly increased if the lungs are diseased, as in respiratory distress syndrome (RDS) or bronchopulmonary dysplasia.

Ventilation-Perfusion Relationships in the Neonatal Lung

Ventilation (\dot{V}) and perfusion (\dot{Q}) are imperfectly matched in the neonatal lung. This may be in part a result of gas trapping in the lungs. \dot{V}/\dot{Q} mismatch is evident in the alveolar-arterial nitrogen difference (A-aDN₂), which is 25 mmHg immediately after birth and declines to about 10 mmHg within the first week. The normal PaO₂ in an infant breathing room air is about 50 mmHg just after birth and increases to 70 mmHg by 24 h of age. The large A-aDO₂ in infants is mainly caused by persisting anatomic shunts (see p. 26) and the relatively large CV.

Lung Surfactant

Surfactants in the alveolar lining layer stabilize the alveoli, preventing their collapse on expiration. Reducing the surface tension at the air-liquid interface in the alveoli also reduces the force required for their re-expansion. The principal surfactant in the lung is lecithin, which is produced by type II pneumocytes. The quantity of lecithin in the fetal lung increases progressively, beginning at 22 weeks of gestation and increasing sharply at 35–36 weeks as the lung matures. The lecithin production of the lung can be assessed by determining the lecithin/sphingomyelin (L/S) ratio in amniotic fluid, and this is used as a measure of lung maturity and predictor of RDS. The L/S ratio is usually less than 1 until 32 weeks' gestation, reaching 2 by 35 weeks, and 4–6 by full term.

Preterm infants with inadequate pulmonary lecithin production suffer from RDS. The biochemical pathways for surfactant production may also be depressed by hypoxia, hyperoxia, acidosis, or hypothermia; therefore, early correction of

these abnormalities in the sick neonate is vitally important. Inhaled anesthetic agents seem to have little effect on surfactant production. Maturation of biochemical processes in the lungs of the fetus in utero may be accelerated by the administration of corticosteroids to the mother. The use of exogenous surfactant therapy to treat RDS is now standard practice, reducing severity of the disease, air leaks, pneumothorax, and mortality. The optimal preparation, dose, and mode of delivery remain controversial.

Lung Growth and Development

The lungs continue to develop during the first two decades of life. The number of alveoli increases rapidly over the first 6 years, almost reaching adult levels, but growth continues into adolescence. In young children, the small size of the peripheral airways may predispose to obstructive lung diseases such as bronchiolitis.

Pulmonary Function Testing in Children

Children older than 6 years of age may cooperate sufficiently to enable standard tests of pulmonary function to be performed, and these may be an important part of the preoperative assessment. However, they should be interpreted within the context of their cooperation. Maximum expiratory and inspiratory flow-volume curves may be useful in determining the site and nature of airway obstruction; for example, they can differentiate between intrathoracic and extrathoracic obstruction. Such studies may be most useful in the preoperative assessment of children with a mediastinal mass. Spirometric studies may provide useful information as to the degree of reversible airway obstruction present in those with a disease such as asthma and may assist in preoperative planning. They also may indicate the extent of restrictive disease such as accompanies scoliosis, thereby predicting the likelihood of postoperative pulmonary insufficiency. There is renewed interest in forced oscillometry as a method to study lung functions in children as this does not require patient cooperation.

Respiratory System Changes with Anesthesia

The following is a summary of some of the major changes that occur in the respiratory system during and after anesthesia.

1. Inhaled anesthetic agents decrease tidal volume, and respiratory rate increases during spontaneous ventilation. This is thought to occur because of the

combined effects of anesthetic drugs on the central chemical control of respiration and on the muscles of respiration. Intercostal muscle activity is inhibited by inhaled anesthetic agents; consequently, diaphragmatic breathing predominates, and the chest wall may move paradoxically, even if the airway is only partially obstructed. Surgical stimulation tends to increase ventilation back toward normal levels. The effect of intravenous agents on ventilation in children is variable and not fully documented.

2. The FRC is reduced during inhaled anesthetic agents with or without neuromuscular blockade. This reduction is greatest in the youngest children and is caused by elevation of the diaphragm and loss of chest wall stability. As the FRC decreases, airway closure may occur during tidal ventilation, resulting in impaired oxygenation. It is for this reason that 5-cm PEEP is commonly advocated during anesthesia in infants and toddlers.
3. The ratio of physiologic dead space to tidal volume ($V_D:V_t$) remains constant in children breathing spontaneously but may increase in those whose ventilation is controlled. During controlled ventilation, major alterations in gas distribution within the lungs occur as a result of changes in the action of the diaphragm. This effect tends to markedly unbalance the \dot{V}/\dot{Q} matching within the lungs. Apparatus dead space may assume considerable significance given the small physiologic V_D and rapid rate of ventilation.
4. Compliance is little changed, and airway resistance is generally reduced by the bronchodilator action of inhaled anesthetic agents. Insertion of an ETT increases total flow resistance (see p. 14), especially in children less than 3 years of age.
5. The efficiency of gas exchange may be impaired by the effects of anesthetic drugs on the physiologic process that normally controls the regional distribution of inspired gases and blood flow throughout the lung (hypoxic pulmonary vasoconstriction).
6. Laryngospasm occurs more frequently in children compared with adults, especially those with an active or recent upper respiratory infection (URI), particularly during induction of anesthesia and after extubation (see p. 91). Laryngeal closure results from apposition of the vocal cords and supraglottic structures. The reason for the increased incidence of laryngospasm in children is unknown.

CARDIOVASCULAR SYSTEM

The Fetal Circulation

The fetal cardiovascular system perfuses the low-resistance placental circulation, directing 36–42 % of the combined ventricular output to this organ; only 5–10 % goes to the lungs. The high pulmonary vascular resistance limits flow to the fetal

lungs resulting in blood bypassing the lungs via the foramen ovale and the ductus arteriosus. Most of the blood returning from the placenta bypasses the liver via the ductus venosus. The pattern of flow from the inferior vena cava into the right atrium (RA) ensures that about one third of the oxygenated placental blood (partial pressure of oxygen (pO_2), 28–30 mmHg) is directed through the foramen ovale into the left atrium. This blood, which combines with the limited venous return from the lungs, is pumped by the left ventricle into the ascending aorta and thence to the coronary, cerebral, and forelimb circulations. Blood returning via the superior vena cava (pO_2 , 12–14 mmHg) passes through the RA into the right ventricle, from which most of the output flows through the ductus arteriosus into the descending aorta. Thus, blood supplied to the heart and upper body has a greater oxygen content (saturation, 65%; pO_2 , 26–28 mmHg) than that supplied to the abdominal organs, lower limbs, and placenta (saturation, 55–60%; pO_2 , 20–22 mmHg). In utero, the right ventricle pumps about 66% of the combined ventricular output, and the left ventricle pumps the remaining 34%.

Circulatory Changes at Birth

At birth, pulmonary ventilation is normally established quickly, and blood flow to the lungs is greatly increased while placental flow ceases. When the lungs expand and fill with gas, pulmonary vascular resistance (PVR) decreases markedly as a result of mechanical effects on the vessels and relaxation of pulmonary vasomotor tone when the pO_2 increases, and the partial pressure of CO_2 decreases in alveolar gas. PVR decreases by 80% from prenatal levels within a few minutes after normal initiation of respirations. As PVR decreases, blood flow to the lungs and then via the pulmonary veins into the left atrium increases, increasing left atrial pressure above that in the RA and closing the atrial septum over the foramen ovale.

Simultaneously, as flow to the placenta ceases because of clamping or umbilical artery constriction, a large, low-resistance vascular bed is excluded from the systemic circulation. This activity results in a large increase in systemic vascular resistance (SVR) and a decrease in inferior vena cava blood flow and RA pressure. The increase in SVR and the simultaneous decrease in PVR increase the aortic pressure above that in the pulmonary artery. Blood flow through the ductus arteriosus reverses (i.e., becomes left to right), and the ductus fills with oxygenated blood. This increased local pO_2 (to levels greater than 50–60 mmHg) causes the muscular wall of the ductus arteriosus to constrict secondary to a prostaglandin-mediated response. Shunts may persist through the ductus for some hours after birth, producing audible murmurs. Normally, however, flow through the ductus is insignificant by 15 h. Permanent closure of the ductus is usually complete within 5–7 days but may not be complete until 3 weeks.

The ductus venosus, which communicates between the umbilical veins, the portal vein, and the inferior vena cava, also remains patent for several days after birth. This channel provides a shunt past the hepatic circulation and therefore may delay the clearance of drugs metabolized in the liver (e.g., opioid analgesics).

The Transitional Circulation

During the early neonatal period, reversion to the fetal circulatory pattern is possible under some circumstances. If hypoxia occurs, the PVR increases, the foramen ovale opens, and the ductus arteriosus may also reopen; a significant proportion of blood then again bypasses the (now high-resistance) pulmonary circulation, causing a rapid decline in arterial oxygenation. Impaired tissue oxygenation then results in acidosis, which causes a further increase in PVR, establishing a vicious cycle of hypoxemia → acidosis → impaired pulmonary blood flow → hypoxemia. Reversion to a fetal pattern of circulation may complicate any condition that causes hypoxemia or acidemia (e.g., RDS or congenital diaphragmatic hernia).

The Neonatal Cardiovascular System

In healthy neonates, the wall thickness of the right ventricle exceeds that of the left. This preponderance is evident in the ECG, which shows an axis of up to +180° during the first week of life. After birth, the left ventricle enlarges disproportionately. By 3–6 months, the adult ratio of ventricular size is established (axis approximately +90°). During the immediate neonatal period, the heart rate is between 100 and 170 beats/min, and the rhythm is regular. As the child grows, the heart rate gradually decreases (Table 2.3). Sinus arrhythmia is common in children. All other irregular rhythms must be considered abnormal.

Systolic blood pressure is approximately 60 mmHg in the full-term neonate, and the diastolic pressure is 35 mmHg. These pressures vary considerably and may be 10–15 mmHg more if clamping of the umbilical cord is delayed or the cord is “stripped,” causing an increase in circulating blood volume. In either case, they decrease to normal values within 4 h. Preterm infants have reduced arterial pressures, as low as 45/25 mmHg in a 750-g infant (Table 2.4).

The myocardium of the neonate contains less contractile tissue and more supporting tissue than the adult heart. Consequently, the neonate’s ventricles are less compliant when relaxed and generate less tension during contraction. Because the reduced compliance of the relaxed ventricle tends to limit the size of the stroke volume, the cardiac output of the neonate is rate dependent. Bradycardia is invariably accompanied by reduced cardiac output. The less compliant ventricle of the neonate is also dependent on an adequate filling pressure,

Table 2.3 Normal heart rate

Age	Heart rate (beats/min)	
	Average	Range
Neonate	120	100–170
1–11 months	120	80–160
2 years	110	80–130
4 years	100	80–120
6 years	100	75–115
8 years	90	70–110
10 year	90	70–110
14 years		
Boys	80	60–100
Girls	85	65–105
16 years		
Boys	75	55–95
Girls	80	60–100

Table 2.4 Normal blood pressure^a

Age	Blood pressure (mmHg)		
	Systolic	Diastolic	Mean
<i>Neonate</i>			
Preterm (750 g)	44	24	33
Preterm (1000 g)	49	26	34.5
Full term	60	35	45
3–10 days	70–75	57	
6 months	95		
4 years	98		
6 years	110	60	
8 years	112	60	
12 years	115	65	
16 years	120	65	

^aReported normal blood pressure values for infants and children must be considered in light of their methods of determination. These values should serve as a guide only (*see* Monitoring During Anesthesia, p. 113)

so that hypovolemia is followed by a decrease in cardiac output. Thus, cardiac output is both rate dependent and volume dependent. Reduced compliance and contractility of the ventricles also predisposes the infant heart to failure with increased volume load. In the infant, failure of one ventricle rapidly compromises the function of the other and biventricular failure results.

The reduced contractility of the neonatal heart is also thought to be secondary to the immaturity of the myofibrils and to the less developed sarcoplasmic reticulum. It is postulated that the cyclic calcium flux within the neonatal

myocardium is more dependent on exchange across the cell membrane (sarcolemma) and less a function of the sarcoplasmic reticulum, thus a greater dependency upon ionized calcium levels. As the infant grows, the myocardial sarcoplasmic reticulum expands and progressively assumes a dominant role in intracellular calcium regulation, which is typical of the adult heart. The greater role of the sarcolemma in calcium regulation within the myocyte may explain the greater sensitivity of the neonate to myocardial depression because of inhalational anesthetics (calcium channel-blocking activity). It may also explain the severe cardiac depressant effects of calcium channel-blocking drugs or the rapid administration of citrated blood products such as fresh-frozen plasma or platelets in the neonate.

The autonomic innervation of the heart is incomplete in the neonate, and there is a relative lack of sympathetic elements. This may further compromise the ability of the less contractile neonatal myocardium to respond to stress. The differences in the neonate's myocardium are all particularly marked in the preterm infant.

In the neonate, shunts hamper the precise measurement of cardiac output, which averages 2–3 times that of the adult on a milliliter per kilogram body weight basis and is appropriate for the metabolic rate. The total systemic vascular resistance is reduced, reflecting the great proportion of vessel-rich tissue in the neonate (18%—twice that in the adult) and resulting in a reduced systemic arterial pressure despite the large cardiac output.

The Pulmonary Circulation

The changes in the pulmonary circulation that occur at birth continue with a slower progressive decrease in PVR over the first 3 months of life. This is associated with a parallel regression in the thickness of the medial muscle layer of the pulmonary arterioles. During the neonatal period, PVR is still high, and the muscular pulmonary vessels are highly reactive. Hypoxia, acidosis, and stress (e.g., from endotracheal suctioning) may all increase PVR. If the increase in the PVR is sustained by such stimuli, right-sided intracardiac pressures may exceed those on the left, and right-to-left shunting may ensue via the ductus arteriosus or foramen ovale. Right ventricular failure, rapidly progressing to biventricular failure, may occur.

In some circumstances, the normal regression of the muscular layer of the pulmonary vessels and the associated decrease in PVR may not occur. Continued hypoxemia, caused, for example, by continued high-altitude or excessive pulmonary blood flow as a result of left-to-right shunts (ventricular septal defect, patent ductus arteriosus, etc.) may lead to persistence of a high PVR into childhood

and beyond. Initially, this high PVR is reversible (e.g., with pulmonary vasodilators) and correction of the underlying defect. Later, this high PVR results in structural changes in the pulmonary vascular bed that are irreversible, causing pulmonary vascular obstructive disease.

Nitric oxide has been identified as an endothelium-derived relaxing factor that is normally produced continually in the lung to regulate pulmonary vascular tone. This has led to the use of nitric oxide inhalation to treat increased pulmonary vascular resistance.

Blood Volume

The blood volume varies considerably during the immediate postnatal period (the primary variable being the amount of blood drained from the placenta before the cord is clamped) and during the first year of life. Delay in clamping or stripping the cord at delivery may increase the blood volume by more than 20 %, resulting in transient respiratory distress. Conversely, fetal hypoxia during labor may vasoconstrict the cord, shift blood to the placental circulation, and cause hypovolemia in the already asphyxiated neonate.

The Response to Hypovolemia

The response to hypovolemia and restoration of the blood volume are of great importance to the anesthesiologist because surgery in the neonate may be accompanied by significant blood loss. Withdrawal of blood during exchange transfusion causes a progressive parallel decrease in systolic blood pressure and cardiac output. Reinfusion of an equal volume of blood restores these parameters to their original values. The changes in arterial blood pressure are proportional to the degree of hypovolemia. The capacity of the neonate to adapt the intravascular volume to the available blood volume is very limited, perhaps due to less efficient control of capacitance vessels. The baroreflexes of the infant, especially the preterm infant, are inactive during anesthesia, further compromising the response to hypovolemia.

In summary, the infant's systolic arterial blood pressure is closely related to the circulating blood volume. Blood pressure is an excellent guide to the adequacy of blood replacement during anesthesia, a fact that is amply confirmed by extensive clinical experience. The hypovolemic infant is unable to maintain an adequate cardiac output; hence, accurate early volume replacement is essential.

Table 2.5 shows approximate normal values for blood volume in infants and children. Values may be greater however, particularly in preterm infants.

The Response to Hypoxia

Because of the high rate of metabolism for oxygen ($\dot{V}O_2$), hypoxemia can develop rapidly in the neonate. The first observed response is usually bradycardia in contrast to the tachycardia observed in the adult. The anesthesiologist should treat any episode of unexplained bradycardia by immediately ventilating the lungs with 100% oxygen. During hypoxemia, pulmonary vasoconstriction occurs, and the pulmonary artery pressure increases more than in adults. The foramen ovale and the ductus arteriosus may reopen resulting in a large right-to-left shunt, further decreasing SaO_2 . Changes in cardiac output and SVR in infants also differ from those in older children and adults. During hypoxemia, the principal response in adults is systemic vasodilation, which, together with an increased cardiac output, helps to maintain oxygen transport to the tissues. The fetus and some neonates respond to hypoxemia with systemic vasoconstriction. During fetal life, this directs more blood to the placenta, but after birth, this response may reduce cardiac output, further limiting oxygen transport and forcing the heart to work harder. In the infant, the early and pronounced bradycardia in response to hypoxia may be caused by myocardial hypoxia and acidosis.

Neonates exposed to hypoxemia experience pulmonary and systemic vasoconstriction, bradycardia, and decreased cardiac output. Rapid intervention is necessary to prevent this state from proceeding to cardiac arrest.

Oxygen Transport

Blood volume in the neonatal period is approximately 80 mL/kg in the term infant and about 20% greater in the preterm infant (Table 2.5). The hematocrit (Hct) may be as great as 60% and the hemoglobin (Hb) 18–19 g/dL. The values for blood volume, Hct, and Hb vary from infant to infant, depending on the time of clamping of the umbilical cord. These values change little during the first week of life, after which the Hb level starts to decrease. This change occurs more rapidly in the preterm infant.

Most (70–90%) of the Hb present at birth in a full-term infant is fetal hemoglobin (HbF). The affinity of HbF for oxygen is greater than that of adult hemoglobin (HbA), primarily because of a lack of effect of 2,3-DPG on the HbF–O₂ interaction. HbF combines with more oxygen but releases it less readily in the

Table 2.5 Normal blood volume of children

Age	Blood volume (mL/kg)
Neonate	80–85
6 weeks to 2 years	75
2 years to puberty	70

tissues than does HbA. The pO_2 at 50% hemoglobin saturation (P_{50}) for HbF is approximately 20 mmHg, in contrast to 26–27 mmHg for HbA. Adequate oxygen transport to the tissues of the neonate therefore demands a greater Hb concentration. Less than 12 g/dL constitutes anemia, and greater levels are very desirable in hypoxic states. However, there are many risks associated with transfusion. Current thought is that correction of anemia by blood transfusion may be indicated to maintain the Hct greater than 40% in cases of severe cardiopulmonary disease, 30% in moderate cardiopulmonary disease or major surgery, and 25% in symptomatic anemia (apnea, tachycardia, lethargy, poor growth).

Transfusion with HbA-containing erythrocytes may improve oxygen transport to the tissues in the sick preterm infant. However, this treatment has also been reported to increase the risk of ROP. During the first weeks of life, the Hct and Hb levels decline steadily, in part because of a progressive increase in blood volume but also a result of suppression of erythropoiesis caused by improved tissue oxygenation (see Table 4.10). This physiologic anemia of infancy reaches a nadir at 2–3 months of age, with Hb levels of 9–11 g/dL. At this time, the HbF content of the blood has been largely replaced by HbA. Thus, oxygen delivery at the tissues is improved. Provided that nutrition is adequate, the Hb level now increases gradually over several weeks to 12–13 g/dL, which is maintained during early childhood.

The preterm infant demonstrates an earlier and greater decrease in Hb concentration, reaching 7–8 g/dL in infants weighing less than 1500 g at birth. This is the result of a short erythrocyte life span, rapid growth, and decreased erythropoietin production. The early “physiologic” anemia of the preterm infant is often followed by a continuing “late” anemia, which is secondary to nutritional deficiencies. In the infant in the neonatal intensive care unit, this anemia is accentuated by repeated blood sampling. Iron therapy is not effective in correcting this anemia and may cause other problems (e.g., hemolysis, infection). Anemia of the preterm infant may lead to tachycardia, tachypnea, poor feeding and growth, diminished activity, and apnea. In severe states, congestive heart failure may occur.

METABOLISM: FLUID AND ELECTROLYTE BALANCE

Glucose Homeostasis

The term neonate has stores of glycogen that are located mainly in the liver and myocardium. These are used during the first few hours of life until gluconeogenesis becomes established. Small-for-gestational-age and preterm infants may have inadequate glycogen stores and may fail to establish adequate gluconeogenesis. Hence, they are very dependent upon IV infusions to prevent hypoglycemia.

Table 2.6 Factors associated with hypoglycemia in neonates

Prematurity
Perinatal stress
Sepsis
Small for gestational age
Polycythemia
Hypoxia
Excess insulin
Infant of diabetic mother
Beckwith-Wiedemann syndrome

Hypoglycemia is common in the stressed neonate (Table 2.6). Blood glucose levels should be measured frequently in sick neonates, and if they decrease to less than 45 mg/dL or 2.2 mmol/L, they should be corrected by a slow continuous infusion of 10 % dextrose at 5–8 mg/kg/min (or 0.05–0.08 mL/kg/min). Symptoms of hypoglycemia (jitteriness, convulsions, apnea) should be treated immediately by slow injection of 10 % dextrose (1–2 mL/kg). Neurologic damage occurs in up to 50 % of infants with symptomatic hypoglycemia. Infants of diabetic mothers and those with Beckwith-Wiedemann syndrome must be treated with particular care because a bolus dose of intravenous glucose may precipitate hyperinsulinemia and serious rebound hypoglycemia. In these infants, a slow infusion of glucose as outlined above is recommended. Older infants and young children rarely become hypoglycemic even during an excessively long preoperative fasting period. Current pediatric practice minimizes preoperative fasting (see p. 77).

Hyperglycemia is a common iatrogenic problem of small infants receiving intravenous therapy, probably as a result of inadequate insulin release and continued hepatic glucose production. The effects of hyperglycemia can be serious. Osmotically induced cerebral fluid shifts may lead to cerebral hemorrhage, and glycosuria may cause diuresis resulting in water and electrolyte depletion. Hyperglycemia (glucose values >200 mg/dL) may also increase the extent of neurologic damage during a cerebral hypoxic-ischemic event. It is essential that glucose therapy be carefully controlled to avoid hyperglycemia. We recommend the use of a continuous infusion pump for maintenance glucose containing fluids with other intraoperative losses replaced with balanced salt solutions “piggy backed” to the maintenance fluids.

Calcium Homeostasis

Calcium is actively transported across the placenta to meet the needs of the fetus. This transport accelerates near term and may cause a decline in maternal calcium levels. After birth, the infant must depend on its own calcium reserves. However, parathyroid function is not fully established, and vitamin D stores may be inadequate. As a result, hypocalcemia must be anticipated—especially in the preterm infant—after birth trauma, neonatal asphyxia, any severe neonatal illness, or blood transfusion (especially fresh frozen plasma or platelets). Correction of metabolic acidosis in the neonate by administration of sodium bicarbonate may precipitate a significant decrease in ionized calcium levels. Continuing hypocalcemia may be a sign of parathyroid dysfunction and is typically present in DiGeorge syndrome (see p. 422, 543).

Symptoms of hypocalcemia include twitching, increased muscle tone, and seizures (hypocalcemia is not always easily distinguished from hypoglycemia). The Chvostek sign may be present, but confirmation depends on laboratory test results (total serum calcium, less than 7 mg/dL or 1.75 mmol/L; ionized calcium, less than 4 mg/dL or 1.0 mmol/L) or on the response to therapy. The infant prone to hypocalcemia is treated with continuous calcium chloride infusion at a rate of 5 mg/kg/h. Symptomatic hypocalcemia requires a slow infusion of either 10% calcium chloride (10–30 mg/kg) or 10% calcium gluconate (30–90 mg/kg), with continuous ECG monitoring. Note that calcium-containing solutions may cause severe skin damage, leading to sloughing if they leak into adjacent tissues. They should preferably be given through a central line.

Magnesium Homeostasis

Magnesium and calcium metabolism are closely related: an imbalance in one may affect the other. Magnesium levels affect parathyroid hormone secretion, and the renal excretion of calcium and that of magnesium are interrelated. Chronic hypomagnesemia is commonly accompanied by hypocalcemia secondary to the effect on parathyroid function.

Hypomagnesemia is more common in preterm infants, small-for-gestational-age infants, infants of diabetic mothers, and infants with intestinal disease. It may also complicate massive blood transfusion. Hypomagnesemia results in abnormal muscle activity, tremors, seizures, and cardiac arrhythmias and may alter sensitivity to muscle relaxant drugs.

Hypermagnesemia may complicate renal failure, or, in the neonate, it may be a consequence of the administration of magnesium sulfate to the mother. It may result in depression of the central nervous and respiratory systems, hyporeflexia, and hypotension.

Bilirubin Homeostasis

In the full-term neonate, unconjugated hyperbilirubinemia during the first week of life (physiologic jaundice) occurs secondary to an increased bilirubin load, limited hepatic cell uptake of bilirubin, and deficient hepatic conjugation to the water-soluble glucuronide. Serum bilirubin levels seldom exceed 7 mg/dL or 103 µmol/L. In preterm infants, greater levels (10–15 mg/dL or 170–255 µmol/L) are commonly reached. These persist for a longer period, owing to a greater bilirubin load and delayed maturation of the hepatic conjugation pathway. The preterm infant may sustain neurologic damage (kernicterus) at reduced serum bilirubin levels (6–9 mg/dL) than does the full-term infant (20 mg/dL). This predisposition is a result of the preterm infant's less effective blood-brain barrier and may be exacerbated by hypoxia, acidosis, infection, hypothermia, or a low level of serum albumin and hence decreased binding sites. The preterm infant must be carefully monitored for increased serum bilirubin levels, and specific treatment should be administered as required. Treatment includes phototherapy and possibly exchange transfusion. Some drugs (e.g., diazepam, sulfonamides, furosemide) displace protein-bound bilirubin (i.e., “acidic” binding sites for albumin) and therefore increase the danger of neurologic damage. There are no reports of anesthetic drugs (except benzodiazepines) producing adverse changes in bilirubin levels, but hypoxia, acidosis, hypothermia, and hypoalbuminemia may all increase the danger.

COMPOSITION AND REGULATION OF BODY FLUIDS

Body Water

The amount of total body water in neonates and infants is relatively greater than in adults. Its distribution also differs, the proportion of extracellular fluid (ECF) being greater in neonates and young children. In the preterm infant, the ECF exceeds the intracellular fluid (ICF), whereas in the older child and adult, the ECF is only half the volume of the ICF (Table 2.7). Normal levels of serum electrolytes in the neonate are listed in Table 2.8.

Table 2.7 Extracellular and intracellular fluid compartments

Fluid	Preterm neonate	Full-term neonate	Infant (7–8 months)	Adult
ECF	50	35–40	30	20
ICF	30	35–40	35	45

Data are % body weight

Table 2.8 Normal blood chemistry values

Parameter	Preterm neonate	Full-term neonate	2 years to adult
Serum chloride (mEq/L)	100–117	90–114	98–106
Serum potassium (mEq/L)	4.6–6.7	4.3–7.6	3.5–5.6
Serum sodium (mEq/L)	133–146	136–148	142
Blood glucose (mEq/L)	40–60	40–80	70–110
Total protein (g/dL)	3.9–4.7	4.6–7.7	5.5–7.8
PaCO ₂ (mmHg)	30–35	33–35	35–40

Neonatal Renal Function and Water Balance

In the neonate, renal function is limited by immaturity of tubular function and an increased renal vascular resistance, which results in reduced renal blood flow and glomerular filtration rate (GFR). GFR rapidly increases after birth as renal blood flow increases. The preterm infant has an even lower GFR, which increases less rapidly over the first weeks of life than it does in the full-term infant. The GFR of the neonate increases with fluid loading but only to a limited capacity. Consequently, the neonate cannot readily handle an excessive water load and may be unable to excrete excess electrolytes (especially sodium) or other substances that depend on glomerular filtration for their elimination. The GFR is further decreased by hypoxia, hypothermia, congestive cardiac failure, or mechanical ventilation; adult values are usually achieved by approximately 1 year of age.

The limited tubular function impairs the infant's ability to modify the glomerular filtrate for conservation or excretion. For this reason, sodium losses may be large, especially in the preterm infant, and must be balanced by intake. These losses are further increased if the GFR is increased by a high fluid intake, hence, the tendency of the neonate to hyponatremia. Glucose reabsorption is limited in the preterm infant, and glycosuria may occur. In the child with marked hyperglycemia, the resultant osmotic diuresis may lead to severe dehydration. The ability of the tubule to excrete acid is reduced in the preterm infant, thus impairing renal compensation during acidosis. The capacity to excrete H⁺⁺ increases with gestational age. The renal threshold for bicarbonate excretion is less in infants than adults, and this leads to reduced serum bicarbonate levels. The limitations of renal function summarized above necessitate careful fluid and electrolyte replacement therapy planned to match losses. Renal vascular resistance decreases and renal function matures rapidly over the first few weeks after birth in the full-term infant. Preterm infants show less rapid changes in renal function.

Fluid loss and hence the required fluid replacement are related to insensible fluid losses, urine output, and metabolic rate. Insensible fluid losses are

relatively great during infancy, major factors being the increased alveolar ventilation and the thin skin of low-birth-weight infants. Fluid losses are markedly increased by the use of radiant heat and/or phototherapy. Because of the infant's proportionally greater water turnover and the limited ability to concentrate urine and conserve water, dehydration develops rapidly when intake is restricted or losses occur.

Maintenance Requirements

Although maintenance requirements are directly related to the metabolic rate and caloric expenditure and are more accurately expressed in milliliters per square meter of surface area, it is most convenient to relate them to body weight. Fluid requirements for full-term neonates are reduced (40–60 mL/kg per 24 h) during the first few days of life as excess fluid present at birth is being excreted. By 1 week of age, the requirements are increased. Table 2.9 shows the volumes of fluid required during the period of great metabolic activity in infants weighing 4–20 kg.

Intraoperative fluid management in the neonate must include replacement of fluid deficits, third-space losses, blood loss, and maintenance fluid therapy. Fluid deficits and losses are generally replaced with a balanced salt solution such as lactated Ringer's solution, whereas maintenance therapy is replaced with an infusion of 5% glucose and one half to one fourth normal saline, containing 20 mEq of K⁺ per liter. If high-concentration glucose solutions (D-10 or D-20) are infusing preoperatively, these solutions should be continued either at the

Table 2.9 Daily maintenance requirements for fluid, electrolytes, and carbohydrates in relation to weight

Weight	H ₂ O (mL/kg)	Na ⁺ (mEq/kg)	K ⁺ (mEq/kg)	Carbohydrate (g/kg)
<i>Neonate^a</i>				
1000 g	≤200	3.0	2.0–2.5	≤10
1000–1499 g	≤180	2.5	2.0–2.5	
1500–2500 g	≤160	2.0	1.5–2.0	≤8
2500 g	≤150	1.5–2.0	2.0	≤5
4–10 kg	100–120	2.0–2.5	2.0–2.5	5–6
10–20 kg	80–100	1.6–2.0	1.6–2.0	4–5
20–40 kg	60–80	1.2–1.6	1.2–1.6	3–4
Adult	30–40	50 mEq total	50 mEq total	100–150 g total

^aAdjust according to postnatal age, exposure to phototherapy, reduced insensible losses with assisted ventilation, etc.

(From The Hospital for Sick Children: Residents' Handbook of Pediatrics, 6th ed. Toronto, Canada, 1979, with permission)

same or at a somewhat reduced rate throughout surgery. In critically-ill neonates, balanced salt solutions should be replaced with colloid solutions or plasma early, as hypoproteinemia is common.

PHYSIOLOGY OF TEMPERATURE HOMEOSTASIS

Because of their large surface area relative to body weight and their lack of heat-insulating subcutaneous fat, infants lose heat rapidly via four routes in order of importance: radiation (39 %)>convection (34 %)>evaporation (24 %)>conduction (3 %). Evaporative heat is lost into the respiratory tract and through the skin, the latter being related to increased skin permeability and is thus a particularly important factor in preterm infants. When heat loss occurs, heat production within the body must increase to maintain a normal core temperature. In adults and older children, this heat production is principally a function of involuntary muscular activity (shivering) accompanied by increased oxygen consumption, both of which can be prevented by the administration of a neuromuscular blocking drug. Infants rely primarily on non-shivering thermogenesis to generate heat. This mechanism, which also results in increased oxygen consumption, occurs mainly in the brown adipose tissue, which makes up 2–6 % of the full-term infant's body weight (less in the preterm infant) and is located around the scapulae, in the mediastinum, and surrounding the kidneys and adrenal glands. The cells of this “brown fat” have many mitochondria and fat vacuoles, and the tissue has a rich blood and autonomic nerve supply. Increased metabolic activity in brown fat is initiated by norepinephrine released at the sympathetic nerve endings. Hydrolysis of triglycerides to fatty acids and glycerol occurs with associated increased $\dot{V}O_2$ and heat production. Brown fat deposits decline during the first weeks of extrauterine life.

Exposure to a cool environment together with a decrease in central temperature normally triggers thermoregulatory vasoconstriction in unanesthetized infants and children. This vasoconstriction tends to limit further heat loss from the body surface. It is now recognized that the mechanisms for controlling body temperature are well developed in the full-term neonate. However, a decrease in core temperature results when compensatory increases in heat production cannot match heat losses. On exposure to a cool environment, increased metabolic activity is initiated in the brown fat so as to maintain the core temperature. This is accompanied by a progressive increase in oxygen consumption as the temperature gradient between the skin and the environment increases. Oxygen consumption is minimal when this gradient is less than 2 °C (i.e., neutral thermal environment). Exposure to a cool environment also leads to increased glucose use and acid metabolite formation.

Table 2.10 Neutral thermal environment temperatures (°C)

Age	Weight			
	1200 g	1200–1500 g	1500–2500 g	>2500 g
0–6 h	34–35.4	33.9–34.4	32.8–33.8	32.0–33.8
6–12 h	34–35.4	33.5–34.4	32.2–33.8	31.4–33.8
12–24 h	34–35.4	33.3–34.3	31.8–33.8	31.0–33.7
24–36 h	34–35	33.1–34.2	31.6–33.6	30.7–33.5
36–48 h	34–35	33.0–34.1	31.4–33.5	30.5–33.3
48–72 h	34–35	33.0–34.0	31.2–33.4	30.1–33.2
72–96 h	34–35	33.0–34.0	31.1–33.2	29.8–32.8
4–12 days	–	33–34 ^a	31–33.2	29.5–31.4
2–3 weeks	–	32.2–34 ^a	30.5–33.0	–
3–4 weeks	–	31.6–33.6 ^a	30.0–32.7	–

^a1500 g

The physiologic responses to cooling lead to increased oxygen and glucose use and result in acidosis, all of which may compromise the sick infant. The infant with chronic hypoxemia (e.g., cyanotic congenital heart disease) is unable to compensate if exposed to a cool ambient temperature and cools rapidly. To eliminate the need for compensatory responses, sick neonates should be maintained in a neutral thermal environment (i.e., in an ambient temperature that minimizes oxygen consumption [Table 2.10]).

During anesthesia, the normal thermoregulatory response of the infant to cold stress is lost, and oxygen consumption is unchanged in response to a cool environment. In addition, normal thermoregulatory skin vasoconstriction is inhibited. There is also a redistribution of body heat away from the central core to the periphery. Therefore, anesthetized infants and children have increased heat loss and a decrease in body temperature. Measures to minimize heat loss and avoid cold stress (warmed OR, warmed preparation and irrigation solutions, convective forced air warmers) are important during anesthesia (see p. 121).

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Clinical Pharmacology

ROUTES OF ADMINISTRATION

Intravenous. The intravenous route is the most reliable route to deliver drugs to target organs under all conditions. Check all drugs (i.e., *read the label*) and doses before administration. For less commonly used drugs, carefully follow the manufacturer's/pharmacy directions as to route of administration, speed of injection, and dilution. Rapid injection of some drugs (e.g., vancomycin) may cause severe physiologic effects (e.g., hypotension/cardiac arrest, "red man syndrome"). Administration of drugs using tuberculin syringes or when diluted (always double check) is particularly important in neonates and infants. Eject all air bubbles from the syringe before administration. Flush all intravenous tubing after every drug is administered to ensure that drugs do not remain in the tubing or the dead space of the injection port. Serious incidents have occurred when neuromuscular blocking drugs or anesthetics/opioids have been flushed into the child after a procedure has ended.

N.B. Drugs should not be injected into *hyperalimentation* fluids because infection or thrombosis of the line may result. However, some children may have a central line for repeated anesthesia/sedation procedures; it should be accessed using appropriate connectors under aseptic conditions. In other cases, it may be appropriate to "hep-lock" a peripheral IV for use in subsequent procedures.

Intramuscular. Drugs administered intramuscularly are rapidly absorbed, especially in small children, and preferably should be given in the lateral aspect of the thigh. (*Absorption is more rapid from the arm [deltoid] muscle than from the leg muscle; however, it carries a risk of nerve damage.*) Intramuscular injections are much less reliable in children in shock or who are hypovolemic, and there is a danger that repeated doses may have a cumulative effect when muscle tissue perfusion improves. Intramuscular injections are painful and generally avoided in conscious children as sterile abscesses may occur.

Intralingual. Injections into the tongue have been recommended for use in an emergency (e.g., succinylcholine) when parenteral access is limited. Systemic absorption is rapid via this route. *However, never inject in the midline base of the tongue as there is a risk of puncturing a blood vessel leading to a lingual hematoma. We have never found a need for this technique and only recommend it for an emergent situation.*

Intratracheal. Drugs sprayed into the trachea through a tracheal tube are rapidly absorbed, and this route is useful in an emergency if an intravenous route is not available (e.g., to administer atropine or epinephrine during CPR). However, this route is no longer recommended as a first choice for resuscitation; if it is used, larger drug doses are required. It is important to either dilute the drug to a larger volume or flush the drug into the tracheobronchial tree with 3–5 mL of saline to ensure delivery to a mucosal surface. Beware that local anesthetics sprayed into the trachea are rapidly absorbed; always check to ensure that the total dose given is safe.

Rectal. The use of suppositories (acetaminophen) or rectal administration of drugs (e.g., pentobarbital, methohexitol) is usually well accepted by children younger than 3 years of age. Absorption is less certain than with other routes, in part because of the partial first-pass effect of the liver when absorption occurs through the inferior/middle hemorrhoidal veins and the variable volume and pH of the rectal milieu. Unanesthetized children older than 3 years of age may be upset if medications are administered via this route. This route is infrequently used today.

Oral. Preoperative medication and postoperative analgesics may be given to selected children by this route. The oral route cannot be used if vomiting or other gastrointestinal dysfunction exists. Many drugs are very rapidly and predictably absorbed across the oral mucous membrane or via the oral transmucosal route in children.

Intranasal. Some drugs are well absorbed across the nasal mucous membrane and are rapidly effective by this route (e.g., sufentanil, midazolam). However, many children are upset at having drops instilled in their nose, and this route has not gained wide acceptance. There is also a concern that some drugs or their preservative (e.g., midazolam) might be neurotoxic if they penetrate the cribriform plate and enter the brain.

Intraosseous. This route is recommended for use during resuscitation in all age groups when an intravenous route is not available; drugs are rapidly absorbed and may be given in the usual doses. This route is advocated for pediatric resuscitation if initial attempts at venous access fail (see Chap. 4).

DISTRIBUTION OF ADMINISTERED DRUGS

In infants and young children, the relative sizes of the body fluid compartments differ from those in the adult. The extracellular fluid compartment is large; hence, drugs that are hydrophilic (e.g., succinylcholine) are required in larger doses.

Protein binding is less in neonates because of reduced total serum protein concentrations and reduced concentrations of specific proteins (e.g., α_1 -acid glycoprotein/albumen) thus more drugs are unbound and free to exert a clinical effect. For this reason, reduced doses of such drugs as barbiturates and local anesthetics are indicated.

Body composition also influences drug distribution; neonates have little fat or muscle tissue. Drugs normally distributed throughout these tissues will have greater plasma concentrations and prolonged duration of action.

METABOLISM AND ELIMINATION OF DRUGS

The half-lives of drugs metabolized in the liver in neonates and infants are generally greater than in the adult (e.g., opioid analgesics). Hepatic blood flow and hepatocellular enzymatic activity are the primary determinants of the rate of metabolism of many drugs by the liver. Hepatic blood flow is similar in the small infant and adult, although it may be reduced in the presence of increased intra-abdominal pressure and congestive cardiac failure. In the first few postnatal days, blood may shunt past the liver via a patent ductus venosus. The Phase 1 cytochrome enzymes mature during the first few years of life, some not reaching adult levels until adolescence. The Phase 2 conjugation pathways for drug metabolism are immature in preterm infants and are not fully active until several months of age. Hence, the elimination half-life of drugs such as morphine in this age group is increased. In addition, alternative pathways for drug metabolism may result in the accumulation of metabolites, some of which may be pharmacologically active (e.g., morphine-3- and morphine-6-glucuronide).

Older infants and young children demonstrate a rapid elimination of some drugs, reflecting the greater hepatic blood flow and enhanced metabolic activity in the child's liver.

Drug clearance (Cl) via the kidney (e.g., most antibiotics) depends on the glomerular filtration rate (GFR) or tubular secretion capacity, both of which are reduced during the first few weeks of postnatal life, (necessitating an increased interval between drug dosing). GFR increases with age, reaching adult values by ~2 years of age.

DRUGS USED IN ANESTHESIA

Inhalation Agents

The concentration of inhaled anesthetics in the alveoli increase more rapidly with decreasing age: infants > children > adults. This is the result of the increased alveolar ventilation to functional residual capacity ratio, the greater proportion

Table 3.1 Minimum alveolar concentration (MAC) (%) of inhalational agents

Age	Halothane	Isoflurane	Sevoflurane	Desflurane
Preterm neonate	Not available	1.3–1.4	Not available	Not available
Full-term neonate	0.87	1.6	3.3	9.1
Infant	1.2	1.8	3.2	9.4
Child	0.95	1.6	2.5 ^a	8.5 ^a

^a60% nitrous oxide reduces the MAC values ~25%

of vessel-rich tissues that rapidly equilibrate with blood concentrations, and the reduced blood-gas and tissue-gas partition coefficients (λ) of the inhaled anesthetics (except sevoflurane) in infants. Therefore, induction of anesthesia is more rapid in infants and small children. The rapid increase in alveolar, blood, and tissue concentrations of inhaled anesthetics may account in part for the precipitous decreases in blood pressure that occur when greater concentrations of these anesthetics are given to infants, particularly during controlled ventilation.

Excretion of inhaled anesthetic agents, and therefore recovery, is also more rapid in infants and small children than in adults, provided that ventilation is not depressed. The alveolar concentration of N₂O decreases to 10% within 2 min after discontinuation of 70% N₂O, a level not reached until 10 min in adults.

The minimum alveolar concentration (MAC) of inhaled anesthetics increases from preterm infants, peaks in infancy and decreases in children and adults (Table 3.1); the reasons for this are unknown. MAC values have been determined for a number of anesthesia interventions including tracheal intubation and extubation and for LMA insertion and removal (Table 3.2).

All inhaled anesthetics depress ventilation to a similar degree as evidenced by CO₂ response curves. A dose-related decrease in V_t and minute ventilation is accompanied by an increase in PaCO₂. Intercostal muscle activity is inhibited, particularly during halothane anesthesia. This contributes to the depressed ventilation and may lead to paradoxical chest wall movement, especially if any degree of airway obstruction occurs.

Inhaled anesthetics depress the myocardium in a dose-dependent manner, due to their calcium channel-blocking activity. The severity of the myocardial depression follows the order halothane > isoflurane \geq sevoflurane & desflurane. Halothane frequently and sevoflurane rarely cause bradycardia, which may be corrected by atropine administration. Inhaled anesthetics may prolong the QT interval (halothane > sevoflurane > isoflurane) in some children, especially young infants. Although in isolation this may be insignificant, if combined with an increased dispersion of repolarization (defined as the difference between the maximum and minimum QT interval) (>100 ms), it may predispose to torsades

Table 3.2 MAC for anesthesia interventions in children

	MAC (%)
Tracheal intubation	Halothane: 1.3 Enflurane: 2.93 Sevoflurane: 2.7
Tracheal extubation	Isoflurane: 1.4 Sevoflurane: 1.70, 2.3 Desflurane: 7.7
LMA insertion	Halothane: 1.5 Sevoflurane: 2.0
LMA extubation	Sevoflurane: 1.84 Desflurane: 3.39 (with caudal block) 3.56 (with fentanyl)
Tracheal intubation/skin incision ratio ^a	Halothane, enflurane, sevoflurane: 1.33
Awake	Sevoflurane: 0.3 Age: 2–5 years, 0.66 5–12 years, 0.43
BIS ₅₀	Sevoflurane 2.83

^aCalculated using the above MAC data

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de pointes. In children with long QT syndrome, medications that increase the Q-T interval may predispose to serious arrhythmias.

Inhaled anesthetics inhibit hypoxic pulmonary vasoconstriction and thereby disrupt the mechanism that normally redistributes perfusion away from under-ventilated alveoli. This increased shunt may cause a clinically significant decrease in SaO₂, especially in infants with lung disease (e.g., bronchopulmonary dysplasia).

Inhaled anesthetics increase cerebral blood flow (CBF) by decreasing cerebral vascular resistance in adults following the order from greatest to least: halothane > desflurane > isoflurane > sevoflurane. These effects are offset in part, by decreasing the cerebral metabolic rate for oxygen consumption. The net effect for each anesthetic is described below.

All of the inhaled anesthetics reduce the requirements for non-depolarizing neuromuscular blocking drugs (NMBDs) to produce a standard degree of block. This relaxant enhancing effect may be useful in reducing the total dose of NMBDs administered and facilitating antagonism of the neuromuscular blockade.

All inhaled anesthetics are capable of triggering malignant hyperthermia (MH) and should be avoided in children at risk for MH.

Nitrous Oxide (N_2O)

N_2O is commonly used to speed and facilitate induction and to provide analgesia/amnesia during maintenance. It may also be administered to sedate and to provide analgesia to facilitate IV catheter placement before an intravenous induction. N_2O is odorless and insoluble [blood-gas partition coefficient ($\lambda_{b/g}$) is 0.47] with a MAC of 104% in adults. In large concentrations, it enhances the rate of uptake of the inhaled anesthetics, accelerating induction (second-gas effect). The analgesic and amnestic effects of N_2O may complement the anesthetic regimen during maintenance. The effects of N_2O on ventilation appear to equal those of equipotent concentrations of halothane. N_2O mildly depresses cardiac output and systemic blood pressure in infants, but it has little effect on pulmonary artery pressure or pulmonary vascular resistance, even in those with pulmonary vascular disease. In infants and small children, the cardiovascular effects of N_2O combined with either halothane or isoflurane to 1.5 MAC are similar to those of equipotent (1.5 MAC) concentrations of either halothane or isoflurane in oxygen. N_2O rapidly diffuses into any gas-containing space within the body; this contraindicates its use in those with lung cysts, pneumothorax, lobar emphysema, necrotizing enterocolitis, bowel obstruction, and any other gas-filled cavity. N_2O (70%) doubles the size of a pneumothorax in 12 min and that of gas containing bowel in 120 min. The tenfold difference in doubling time is the result of the shrinking blood supply to the bowel, as the lumen of the bowel expands, and unchanged blood flow to the chest wall/pleura with expansion of the pneumothorax. N_2O also diffuses into the middle ear and may displace the graft during tympanoplasty. In some children with a normal ear and an intact eardrum, postoperative absorption of N_2O from the middle ear results in atelectasis of the drum and a later complaint of earache. N_2O does not appear to increase postoperative vomiting in children. However, when treating adolescents for procedures that are associated with a high incidence of PONV, it may be prudent to avoid nitrous oxide.

Sevoflurane

In the past decade, sevoflurane has become the agent of choice for inhalational induction in children. A fluorinated methyl isopropyl ether, it has a low solubility in blood ($\lambda_{b/g}$ is 0.68), a pleasant odor and is the least irritating to the airway of the potent inhaled anesthetics. As such, it is ideal for an inhalational induction. Induction can be most rapidly achieved by administering the maximum deliverable initial concentration (8%); it is not necessary to introduce sevoflurane in slow stepwise increases in inspired concentration except as a means of reducing its pungency for the child. Slowly increasing the inspired concentration of sevoflurane only prolongs the excitement period. To minimize any objection to the 8% inspired concentration of sevoflurane, the facemask may be

flavored and 70% N₂O in oxygen administered first until the child stops responding verbally. For older, cooperative children, the so-called single breath induction (i.e., a vital capacity breath) using an anesthesia circuit primed with 8% sevoflurane with (or without) 66% nitrous oxide in oxygen and fitted with a large reservoir bag will induce anesthesia rapidly and smoothly. The effect of age on the MAC of sevoflurane differs from that of the other anesthetics: it is 3.2% in neonates up to 6 months of age and 2.4% in children 6 months to 10 years. The large MAC values limit the effectiveness of the overpressure technique with an 8% vaporizer. The addition of 60% N₂O to sevoflurane in children only decreases the MAC by 25%. Attempts at intravenous cannulation should not be made until the child is adequately anesthetized and unresponsive. Satisfactory conditions for tracheal intubation without neuromuscular blocking agents can be achieved quite rapidly as anesthesia is deepened, especially if a single dose of propofol is administered before intubation or ventilation is controlled for a brief period.

Minute ventilation decreases about 50% during 8% sevoflurane as a result of a decrease in tidal volume and despite a compensatory increase in the respiratory rate. Surgical stimulation reverses in part the decreased minute ventilation. In practice, the addition of nitrous oxide to sevoflurane decreases the MAC of the latter and offsets in part the respiratory depression in the spontaneously ventilating child. Sevoflurane is a potent bronchodilator and effective for relaxing the airways in children with reactive airway disease.

Sevoflurane causes less myocardial depression than halothane, isoflurane, or desflurane and infrequently causes arrhythmias, even in the presence of epinephrine-containing solutions. Heart rate increases and blood pressure decreases minimally with induction of anesthesia. Occasionally, bradycardia occurs during induction of anesthesia in infants and particularly in children with trisomy 21. This may be prevented by pretreatment with an anticholinergic. Compared with halothane, sevoflurane causes less hypotension in children with congenital heart disease and less desaturation in those with cyanotic CHD.

Sevoflurane has similar effects on CBF and oxygen consumption as isoflurane. Sevoflurane increases CBF although cerebral autoregulation is maintained up to 1.5 MAC. Like isoflurane, prior hyperventilation restores autoregulation during sevoflurane anesthesia and attenuates the increase in CBF. Sevoflurane decreases the cerebral metabolic rate for oxygen the most of all inhalational anesthetics. The net effect is that sevoflurane provides the optimal ratio of CBF/oxygen consumption of these anesthetics. In sum, sevoflurane like isoflurane has a minimal effect of ICP, particularly if preceded by hyperventilation.

Sevoflurane decreases the dose requirement for non-depolarizing relaxants compared with TIVA, although the duration of action is prolonged compared with TIVA. These effects are similar to those of isoflurane.

Epileptiform EEG activity and myoclonic jerking movements have been rarely reported during sevoflurane anesthesia. The combination of large concentrations of sevoflurane and hyperventilation increase EEG epileptiform activity, although frank seizures are exceedingly rare. Epileptiform EEG activity during sevoflurane may cause paradoxical readings on “depth of anesthesia” monitors (e.g., the BIS monitor) as the depth of anesthesia appears to lessen as the inspired concentration exceeds 3%. Caution should be exercised when interpreting BIS values during sevoflurane anesthesia at large concentrations.

Emergence from sevoflurane anesthesia is smooth and rapid, although emergence delirium may occur in some infants and children, especially preschool-age children (see p. 213). This transient phenomenon (lasting <15 min in most) is characterized by restless and inconsolable behavior, inability to establish eye contact, lack of purposeful movement, and lack of interaction with their surroundings. Antinociceptive strategies including a regional block or systemic analgesics, combined with propofol, an alpha₂ agonist (e.g., dexmedetomidine) or an opioid during emergence may reduce the risk of emergence delirium.

Sevoflurane is metabolized (5%) *in vivo* by CYP2E1-releasing inorganic fluoride; maximum plasma concentrations occur within 2 h of terminating the anesthetic. Although nephrotoxicity after methoxyflurane was attributed to large-plasma inorganic fluoride concentrations, evidence has shown that the nephrotoxicity arises from the affinity of renal CYP2E1 for the ether anesthetics, which releases inorganic fluoride in the renal medulla impairing renal tubular function. The affinity of CYP2E1 for sevoflurane is one-fifth that of methoxyflurane; hence, nephrotoxicity does not occur with the former despite similar plasma concentrations of inorganic fluoride to those after methoxyflurane.

Sevoflurane is also degraded *in vitro*, in the presence of some carbon dioxide absorbents (Baralyme > soda lime >> Amsorb Plus). Amsorb is an example of a lithium-based absorbent, which does not degrade sevoflurane. The degradation yields five compounds, the most common being compound A, a vinyl compound that is nephrotoxic in rats. The production of compound A increases in parallel with the child's weight. In some countries, a minimum fresh gas flow (2 L/min) and concentration-time exposure have been recommended for sevoflurane in the presence of soda lime absorbents, although there is limited evidence that fresh gas flows < 2 L/min present any risk to humans. Absorbents that are free of potassium hydroxide and sodium hydroxide such as lithium-based absorbents do not degrade sevoflurane and are not regulated by any minimum fresh gas flows. To date, neither sevoflurane nor its degradation products have been shown to be toxic in humans.

The extent of the degradation of sevoflurane increases as the fresh gas flow decreases and the absorbent becomes desiccated. When sevoflurane is degraded in the presence of desiccated Baralyme, it causes an exothermic reaction that produces hydrogen gas at temperatures >200 °C. This resulted in several fires that led to the withdrawal of Baralyme from the market.

Desflurane

Desflurane is a fluorinated ether with a boiling point of 23 °C and the smallest $\lambda_{b/g}$, 0.42. In addition, it is the least soluble anesthetic. It is a very stable compound, with less than 0.02 % metabolized in vivo, although it is degraded to carbon monoxide in the presence of some desiccated absorbents (such as Sodalime). However, carbon monoxide is not produced in the presence of lithium-based absorbents. Desflurane is the least potent ether anesthetic agent with a MAC of 7–9.5 % in children. MAC increases with decreasing age, peaking in infants 6–12 months of age, and decreasing thereafter in neonates. Similar to sevoflurane, 60 % nitrous oxide decreases the MAC of desflurane in children only 25 %. In adults, the MAC of desflurane in homozygous redheads is 20 % greater than in non-redheads. The effects of 1 MAC desflurane on the cardiovascular system are similar to those of other ether inhalational anesthetics, although bradycardia is very rare. When desflurane is the sole anesthetic agent, sudden increases in the inspired concentration can lead to profound central sympathetic discharge, resulting in sudden and profound increases in blood pressure and heart rate. This may be prevented by prior administration of opioids.

Desflurane is very pungent and causes upper airway irritation; breath holding and laryngospasm are very common (~50 %), and for these reasons, desflurane is not recommended for inhalational inductions in children. It can, however, be safely used to maintain anesthesia after induction with other agents in children whose airways are intubated. There are limited data on airway responses to desflurane in children managed with either a mask or an LMA. Desflurane is not approved for maintenance in children whose airways are not instrumented; risks of adverse respiratory responses are increased when administered with an LMA in children <6 years. Emergence from anesthesia is very rapid as a result of its limited solubility. Airway reflex responses are infrequent after extubation during desflurane anesthesia; however, caution should be exercised when LMAs are removed during deep desflurane anesthesia. In this case, airway reflex responses are more common than when the LMAs are removed when the children are awake. To prevent the sudden onset of acute pain, analgesics should be administered before emergence. Emergence delirium has also been reported after desflurane, particularly if pain is present. The very rapid recovery may be useful in some children (e.g., small infants at risk for postoperative apnea, although this does not eliminate this risk).

Desflurane increases ICP more than isoflurane and sevoflurane, although this effect may be attenuated in part, if hyperventilation precedes its introduction. As a result, it is not the preferred inhalational anesthetic for children with increased ICP.

Desflurane decreases the dose of non-depolarizing relaxants compared with TIVA. Similarly, the time to recover neuromuscular function is delayed compared with TIVA. The effects may be similar to isoflurane.

Desflurane is less convenient to use than other agents because its low boiling point demands a specially designed, electrically heated vaporizer. A greater concentration is required to maintain anesthesia (because of its greater MAC), necessitating small fresh gas flows to limit the cost.

Isoflurane

Isoflurane is a polyhalogenated methyl ethyl ether anesthetic. It is a stable compound that is metabolized less than 0.2% *in vivo*, although it is degraded to carbon monoxide in the presence of some desiccated CO₂ absorbents. It is eliminated almost completely unchanged via the lungs; therefore, recovery should be very complete.

The $\lambda_{b/g}$ of 1.43 dictates that wash-in to the alveoli is slower than that of sevoflurane. In addition, isoflurane is not suited as an induction agent because its pungent odor irritates airway reflexes in children (coughing, laryngospasm, and breath holding). Nonetheless, isoflurane can be successfully administered after an intravenous induction, provided the concentration is increased slowly. Isoflurane depresses the respiratory system to a similar extent as sevoflurane. Recovery after isoflurane anesthesia is slower than after sevoflurane, although the frequency of emergence delirium appears to be similar. The incidence of laryngospasm during extubation and emergence is similar to that with halothane (see later discussion).

During isoflurane anesthesia, blood pressure decreases although heart rate does not change substantively. The decrease in blood pressure is due in part to myocardial depression, but also to peripheral vasodilation. Infusion of intravenous fluids usually restores the blood pressure. The vasodilating effect of isoflurane may be useful to control blood pressure, such as during induced hypotension. Isoflurane depresses the baroreflex in the neonate. This impairs the ability to compensate for changes in arterial blood pressure and for hypovolemia. Isoflurane does not sensitize the myocardium to the effects of catecholamines or theophylline.

As with the other ether inhalational anesthetics, isoflurane decreases cerebral vascular resistance and therefore increases CBF. At the same time, isoflurane decreases the CMRO₂, increasing cerebral volume up to ~1.5 MAC in adults. The effects in children are likely similar to those in adults. Nonetheless, with prior hyperventilation, isoflurane like sevoflurane, increases ICP the least of the current inhalational anesthetics.

Isoflurane potentiates non-depolarizing neuromuscular blocking drugs to a greater extent than sevoflurane or halothane, thus reducing the doses of relaxant drugs required. The neuromuscular effects of isoflurane are reversible when isoflurane is withdrawn, thereby facilitating reversal of neuromuscular blockade.

Halothane

Halothane was considered to be an ideal anesthetic for children, although its popularity in pediatric anesthesia has waned since the introduction of sevoflurane. Halothane is inexpensive and is still used in some parts of the world where more expensive anesthetics are less available. Pediatric anesthesiologists familiar with halothane consider that it may have some advantages over sevoflurane when a more prolonged emergence is needed to facilitate endoscopy with spontaneous ventilation (e.g., for laryngoscopy, bronchoscopy) or when managing a difficult airway. Its use will therefore be discussed.

With a $\lambda_{b/g}$ of 2.3 the wash-in of halothane is slow compared with sevoflurane. However, with a 5% maximum inspired concentration and a MAC in children of approximately 1%, the overpressure technique is more increased with halothane than with sevoflurane. The MAC in children who are cognitively challenged is 25% less than in children who are not challenged. Halothane provides a smooth inhalation induction with minimal irritation of the airways.

Halothane depresses respiration in a dose-dependent manner; minute ventilation decreases because of a decrease in tidal volume, despite an increase in respiratory rate. This increases the end-tidal carbon dioxide concentration but prevents an anesthetic overdose as long as respiration is spontaneous (see later discussion). Alveolar ventilation returns toward normal during surgical stimulation but is variable throughout anesthesia. Halothane inhibits intercostal muscle activity. Diaphragmatic ventilation predominates, and paradoxical movement of the chest wall may occur. Even very low blood levels of halothane severely depress the ventilatory response to hypoxia in young adult volunteers. It is likely that this effect also occurs in children of all ages. Laryngospasm may occur during light planes of halothane anesthesia, especially during extubation of the trachea. This effect can be avoided by extubating the trachea while the child is either still deeply anesthetized or completely awake. Lidocaine (1–2 mg/kg IV) given slowly before extubation may reduce coughing but will not prevent laryngospasm. Halothane is a potent bronchodilator and is very useful in children with asthma.

Halothane depresses myocardial contractility and heart rate, especially in neonates, leading to a decrease in cardiac output. Severe hypotension may ensue if large concentrations of halothane are administered, particularly with controlled ventilation. The combination of a rapid wash-in and decreased cardiac output speeds the increase in the concentration of halothane in the child's myocardium, leading to electromechanical dissociation/cardiac arrest.

Administration of atropine prevents bradycardia, which reverses in part, the myocardial depression in neonates. Atropine does not affect myocardial contractility in neonates. However, in infants (>3 months) and children, atropine not only increases heart rate but triggers a calcium-dependent, force-frequency response that increases myocardial contractility and restores cardiac output.

The infant's blood pressure is very sensitive to changes in cardiac output. However, vasoconstriction is less effective than in the adult, and halothane also depresses reflex baroreflexes. Accordingly, the blood pressure should be carefully monitored during halothane anesthesia and the inspired concentrations limited to 0.5–1% during controlled ventilation to prevent profound hypotension. In children with cardiac failure, the myocardial depressant effects of halothane are prominent, and severe hypotension may occur.

Arrhythmias and ventricular premature beats are common, especially during spontaneous ventilation in the presence of hypercarbia or increased endogenous catecholamines. If they occur, ventilation should be assisted or controlled and the depth of anesthesia deepened; if they persist, another inhalational anesthetic (e.g., isoflurane) should be substituted for halothane. Junctional rhythm and wandering pacemaker may also occur during halothane anesthesia.

Halothane increases CBF to the greatest extent of the inhalational anesthetics (see above) and therefore may increase ICP. At small concentrations, however, this effect is minimal, and if prior hyperventilation is instituted, the increase is limited even in those with intracranial space-occupying lesions.

Halothane is metabolized 15–20% in adults *in vivo*, but the extent of metabolism is less in children. Occasional cases of hepatic failure have been reported in adults, but many fewer have been reported in children, despite its wide and often repeated use in the latter age group. These rare cases of halothane hepatitis in children have been confirmed by the presence of halothane-related antibodies. Most episodes of halothane hepatitis in children run a less fulminant course than in adults. The reason for the reduced susceptibility of prepubertal children to halothane hepatitis is not known. Contraindications to halothane include a history of unexplained postoperative jaundice.

Summary

1. The MAC values of inhalational anesthetics increase with decreasing age from adults to peak in infancy. MAC decreases steadily as age decreases from infancy to neonates and preterm infants.
2. The smaller the child, the more rapid the wash-in of inhalational anesthetics.
3. Large concentrations of inhalational anesthetics can cause serious hypotension in infants and young children, particularly when ventilation is controlled.
Beware: overdose of inhaled agents is a leading cause of serious complications.
4. Sevoflurane is currently the most commonly used induction agent in infants and children; it is well tolerated by face mask, a respiratory bronchodilator and with limited cardiovascular depression.
5. All inhalational anesthetics may be used for maintenance, although recovery is more rapid with those of reduced solubility: desflurane < isoflurane ~ sevoflurane < halothane.

6. Emergence delirium occurs most commonly in preschool-age children after all ether anesthetics; analgesics, propofol, and α_2 agonists attenuate its frequency.

Miscellaneous Gases

Oxygen

Oxygen toxicity is rare, but two groups of children are vulnerable to large oxygen concentrations: those who are born premature and those who have received bleomycin and several other chemotherapeutic agents:

1. Preterm infants are vulnerable to the effects of reactive oxygen species produced by large concentrations of oxygen through at least two pathways. First, high inspired oxygen concentrations have been associated with ROP for many years. Although this association is far more complex than simply administering excess concentrations of oxygen, limiting the oxygen concentration in premature infants reduces the severity of this disorder. The controversy regarding the optimal oxygen concentration that limits the development of ROP without increasing mortality continues. Currently, the optimal concentration of oxygen recommended is that which maintains a SaO_2 of 90–95 %. Second, exposure to excess oxygen concentration in neonates at the time of delivery increases oxygen free radicals and oxidative stress, which may increase perinatal mortality. This has led to the practice of resuscitating neonates with room air provided the preductal $\text{SaO}_2 > 93\%$. If however, the infant remains hypoxic on room air, then supplementary oxygen should be added to achieve the target SaO_2 .
2. Children with cancer who are treated with bleomycin are at increased risk for pulmonary oxygen toxicity. Bleomycin increases the threshold for oxygen toxicity by promoting free radical formation in the presence of excess oxygen. Oxygen toxicity develops 4–10 weeks after bleomycin >400 mg. Risk factors for pulmonary toxicity include >400 mg cumulative dose, poor pulmonary reserves, radiotherapy, uremia, greater inspired oxygen concentrations, increased age (elderly), and multiple chemotherapeutic agents. Limiting the risk of toxicity is most effectively achieved by restricting the dose of bleomycin and the concentration of oxygen.

Nitric Oxide

Nitric oxide is a selective pulmonary vasodilator, which is not widely available in the OR. Its primary role is as a pulmonary vasodilator for infants with chronic pulmonary hypertension such as for persistent fetal circulation, congenital diaphragmatic hernia, and meconium aspiration. It has a very brief half-life, necessitating continuous administration. Nitric oxide is infrequently used by anesthesiologists; most commonly it is prepared and delivered by respiratory therapists. To maximize the delivery of nitric oxide to the lungs in patients with

tracheal tubes, the delivery tubing is “y”ed into the breathing circuit as close to the patient as possible.

Helium

Helium is a rare, noble gas with a molecular weight (MW) of 4. It has two roles in anesthesia: to reduce the density of gases during airway narrowing and as a nonoxidizable, nonflammable carrier gas. Gas flow in the upper airway and in the presence of airway narrowing is turbulent gas flow (Reynold's number >2100). Under these conditions, gas flow is inversely proportional to the square root of the density of the gas. Since density is directly proportional to the MW of an ideal gas, substituting 100% helium (MW 4) for 100% oxygen (MW 32) increases airway flow by the $\sqrt{8}$ or 2.65-fold. Changing from 100% oxygen to an 80:20 mixture of helium-oxygen (with a MW of 9.6) increases the gas flow 1.83-fold. If the oxygen requirements increase, the incremental increase in gas flow decreases by the square root of the MW ratio of the gases, reducing the therapeutic benefits of helium.

Intravenous Agents

If the child arrives in the OR with peripheral intravenous access, this route can be used to induce anesthesia. In general, injections should not be made into central or peripheral lines that are infusing intralipid or other hyperalimentation fluids to minimize the risk of sepsis. If there is no intravenous access, then a skillful, painless intravenous induction may cause fewer psychological sequelae than an inhalation induction. Needles and syringes should be kept out of the child's sight at all times, and the word *needle* specifically should be avoided. Music, pictures, video player, and bubble blowing are helpful distractions during venipuncture. The use of appropriately applied topical local anesthetic (see Appendix C, p. 634) facilitates painless venipuncture or intravenous cannula insertion. Alternatively, N₂O (inspired concentration, 50–70%) may be given by mask to sedate the child and provide analgesia for venous cannulation. “Poorly visible veins” may be improved by warming the site or applying nitroglycerine cream.

Midazolam

Midazolam is a water-soluble benzodiazepine that has been used for premedication (0.25–0.75 mg/kg PO or 0.05–0.1 mg/kg IV) and as an amnesic supplement to general anesthesia. It has an oral bioavailability in young children of 35% and an elimination half-life of about 3 h after single oral and IV dosing. Ventilation and cardiovascular homeostasis are generally maintained. Midazolam produces

satisfactory sedation in children in pediatric intensive care units; the loading dose is 0.2 mg/kg followed by an infusion at 0.4–2 µg/kg/min. It is not an effective agent for induction of anesthesia because of the large doses required and the variability in response. Midazolam has been most widely used as an oral pre-medication in children, although other routes including the nasal and rectal routes have been used (see Table 4.5). It should be noted that severe hypotension might occur in neonates when it is combined with fentanyl. Midazolam is the only benzodiazepine approved for use in neonates.

Propofol

Propofol (2,6-diisopropylphenol) is a short-acting general anesthetic that is associated with a very rapid onset and offset of anesthesia and pleasant recovery. Although recovery after a single dose of propofol is more rapid than after thiopental, recovery after propofol increases steadily after 1 h. Propofol has rapidly replaced thiopental as the induction agent of choice. Propofol is hydrophobic and therefore must be suspended in an emulsion of soybean oil. Asepsis is especially important to avoid bacterial contamination when handling this anesthetic, although all formulations in the USA contain an antibacterial agent (EDTA, sodium metabisulfite, or benzyl alcohol). Benzyl alcohol may trigger bronchospasm in asthmatic children and may be toxic in neonates. Anaphylactoid reactions have been reported after the use of the metabisulfite formulations. Unused propofol with preservative should be discarded after the vial has been opened or the syringe loaded for 12 h. The dose of propofol to induce anesthesia ranges from 1–5 mg/kg; larger doses may be required for younger infants and unpremedicated children, although the dose in neonates remains undetermined (Table 3.3). The respiratory and cardiovascular effects of a sleep dose (2.5–3.5 mg/kg) are similar to those of thiopental; a brief period of apnea may occur, and blood pressure may decrease transiently. An LMA may be inserted after a dose of 3.5 mg/kg. During induction of anesthesia with sevoflurane, up to 1.5–3 mg/kg propofol facilitates laryngoscopy and tracheal intubation without a prolonged apnea.

Table 3.3 Effect of age on the effective dose (ED_{50}) of common IV induction agents

Age	Thiopental	Propofol
Neonate	3–4	
Infant	6–7	3–5
Child	4–5	1–2.5

Doses are mg/kg

N.B. These are the ED_{50} values; larger doses are required to reliably induce anesthesia

Airway reflexes and oropharyngeal muscle tone are depressed by propofol; this is useful for cases involving airway instrumentation (e.g., LMA, intubation without muscle relaxant, difficult airway) and generally results in a good airway during emergence. The hypertensive response to instrumenting the airway is less after an induction sequence with propofol plus relaxant than after thiopental plus relaxant. In the presence of a difficult airway, small incremental doses of propofol may be used after an inhalational induction to facilitate tracheal intubation. The advantage of repeated doses of propofol is that spontaneous respiration returns quickly. Extraneous limb movements may occur during induction of anesthesia with propofol, especially if smaller doses are administered.

Pain is common at the site of IV injection, although it is less severe if propofol is injected into a free-flowing infusion or into a large antecubital vein. Pain is believed to be due to trace concentrations of propofol in the aqueous egg lecithin outer layer of the emulsion droplets that activate nociceptive nerve endings within the vein. Either administration of 70% N₂O by mask or application of a “mini Bier block” with 0.5–1.0 mg/kg lidocaine for 60 s before IV administration of propofol eliminates or substantively reduces the pain of injection.

Propofol may be infused continuously for maintenance of anesthesia in children to provide total intravenous anesthesia (TIVA). This approach may offer the advantage of rapid recovery with minimal sequelae and may also facilitate anesthesia in special locations where space is limited. The infusion rates for children vary depending on concurrent medications, but they tend to be greater than those in adults. In preterm and full-term neonates and infants less than 1 year of age, propofol clearance decreases dramatically; repeat doses or a large infusion rate may accumulate. When infusions are used to maintain anesthesia, the administration rate should be adjusted to match the predicted elimination of the drug (to maintain a constant blood level) and to prevent light anesthesia. The infusion may be adjusted manually or controlled by a computer, as in target-controlled infusion. In either case, the rate should be dictated by the pharmacokinetic parameters for the child’s age group. Although these parameters vary considerably more in children than in adults, clinically useful dosage schedules for children have been developed (Table 3.4). For brief or minor surgeries, intermittent intravenous boluses of propofol are both effective and efficient. Experience with propofol in pediatric hospitals suggests that it has widespread applications outside the OR—in MRI and other radiological studies, radiotherapy, and invasive procedures that include medical procedures, burn dressing changes, and endoscopies.

Propofol has a potent antiemetic effect and has been used in children with a history of persistent postoperative vomiting after anesthesia or when the risk of vomiting is substantial.

Propofol has been quite widely used in the neonate (e.g., before intubation in the NICU); however, there have been rare reports of severe hypotension in

Table 3.4 Infusion regimen for propofol anesthesia in children

Anesthesia may be induced by inhalation (in such cases, a loading dose may be unnecessary)
Loading sleep dose: 2.5–5 mg/kg
Initial infusion rate (first 20 min) ^a : 200–300 µg/kg/min or 12–18 mg/kg/h
Subsequent infusion rate (next 20 min): Reduce dose by 20%; to 160–240 µg/kg/min or 9.6–14 mg/kg/h
Final infusion rate: 50% of the initial rate; to 100–150 µg/kg/min or 6–9 mg/kg/h as tolerated
These rates must be adjusted or supplemental doses administered if signs of light anesthesia appear If an opioid is used, these infusion rates may be reduced by 25 %
^a Increasing rates (400–500 µg/kg/min) may be required to stop movement during radiological procedures in younger infants and children and in those who are cognitively impaired

neonates given propofol and one unpublished report of a subsequent cardiac arrest. Propofol should be used with caution in the neonate.

Propofol is no longer recommended for prolonged sedation in the pediatric intensive care unit. Propofol infusion syndrome (PRIS), which consists of acute refractory bradycardia progressing to asystole, associated with severe metabolic acidosis and other metabolic derangements, has been reported in children after prolonged propofol infusions (>5 mg/kg/h for 48 h), although incipient PRIS has been diagnosed after exposures of several hours and during anesthesia. Watch for unexpected arrhythmias and metabolic acidosis as signs that PRIS may be developing; the infusion rate should be decreased or discontinued. The mechanism underlying PRIS is unclear but may be due to impaired active transport of long chain triglycerides into myocardial mitochondria and to an interruption of the respiratory chain at complex II by propofol. Few effective treatments have been evaluated but only anecdotally because of the rarity of the disorder: hemodialysis, plasmapheresis, partial-exchange transfusion, and extracorporeal membrane oxygenation (ECMO). There is some concern with the use of propofol in children with mitochondrial disorders, particularly those associated with fatty acid metabolism. In the latter population, the smallest effective dose of propofol should be used and combined with an opioid as appropriate.

Ketamine Hydrochloride

Ketamine, a phencyclidine derivative, has been used to sedate and anesthetize infants and children for a wide variety of situations, although its appeal has been diminished with the introduction of propofol. Its oral bioavailability is 20%; 20–50% is protein bound. Recovery is achieved by redistribution of the drug from the brain to peripheral tissues. Ketamine is metabolized primarily by CYP3A4 to norketamine, which has about one-third the potency of the parent drug. Clearance is unaffected by age, when normalized to a 70-kg adult.

Ketamine 1–2 mg/kg IV induces general anesthesia within 1 min. For sedation, 3–10 mg/kg IM has a 2–5 min onset and about a 30–60 duration of action. Infants require larger doses than older children and adults. Oral ketamine (5–6 mg/kg) may be useful as a premedication or combined with midazolam (Table 4.5). These doses should be supplemented with atropine (0.02 mg/kg) or glycopyrrolate (0.01 mg/kg) to prevent excessive secretions. Low-dose ketamine has also been used for perioperative analgesia, particularly after tonsillectomy in children with OSA (see Chap. 10). Ketamine has generally been avoided in children with central nervous system diseases, although the evidence is conflicting. It produces profound analgesia, unconsciousness, cataleptic state, and amnesia. Ketamine increases CBF, ICP, and CMRO₂. The airway is usually well maintained, but secretions are increased (give atropine!) and airway obstruction or laryngospasm may occur. A mild degree of respiratory depression with brief periods of apnea may occur after induction. Because the protective laryngeal reflexes are depressed, gastric contents may be regurgitated and aspirated. Ketamine increases both the heart rate and the mean arterial pressure, although its direct effect on the isolated heart is a depressant one. In healthy subjects, cardiac output is increased, and peripheral vascular resistance is maintained. These indirect cardiovascular responses are mediated by adrenergic pathways.

Ketamine has limited gastrointestinal effects, although hypersalivation, nausea, and vomiting may occur. There have been no reports of hepatic or renal damage after its administration. Ketamine is infrequently associated with emergence phenomena, including hallucinations and nightmares. These emergence phenomena may be reduced by pretreatment with benzodiazepines and recovering the child in a quiet area. The effects of ketamine on ICP and CBF have led most to abandon this drug during neuroradiologic or similar procedures, although it remains in use for cardiac catheterization. Ketamine has no effect on visceral pain and therefore is unsatisfactory for abdominal surgery, unless regional analgesia is also provided.

Ketamine has been widely used for children with burns for dressing changes and for minor skin grafting procedures. For prolonged procedures, the advantage of an early return to normal nutrition is outweighed by the slow emergence (particularly if given in a prolonged infusion and in combination with a benzodiazepine). It is also useful for minor superficial procedures in infants. Ketamine may also be valuable for anesthesia in children with right-to-left intracardiac shunt, epidermolysis bullosa, or Stevens-Johnson syndrome; for induction of anesthesia in children in severe shock; for general anesthesia when facilities are limited as in some underdeveloped countries; and in large-scale disasters.

Etomidate

Etomidate is not widely used in children. The single induction dose recommended is 0.2–0.3 mg/kg IV. Pain on injection, random movements, myoclonus, and laryngospasm have been reported. Pretreatment with IV lidocaine may attenuate these effects. This drug should not be given by continuous infusion or repeatedly. Its main advantage is cardiovascular stability in hypovolemic patients or those with cardiovascular disease (e.g., cardiomyopathy). Etomidate has been used to sedate children for CT scans and emergency room procedures. However, suppression of adrenocortical function may persist for 24 h, even after a single dose. This suppression may have clinical implications for critically ill children.

Thiopental

Thiopental has been supplanted by propofol as the primary intravenous induction agent in infants and children. Induction dose depends on age (see Table 3.2). Onset of anesthesia is rapid and smooth, without pain but usually accompanied by a very brief period of apnea. Cardiovascular changes are minimal in the healthy child. Neonates are especially sensitive to barbiturates because of reduced protein binding. Barbiturates should not be used for children with porphyria and should be administered with caution in children who may be hypovolemic, those with limited cardiac reserve, and those with potential difficult airway. Thiopental reduces intraocular and cerebrospinal fluid pressure and hence may be especially useful for induction of anesthesia in children having ocular or neurosurgical procedures. Thiopental may cause arterial spasm if injected into an artery; injections should be made cautiously into the antecubital fossa as the vascular access may be an unrecognized artery that will spasm when thiopental with an alkaline pH (10.8) is injected. Extravasation into tissues should be avoided because of the alkaline pH but does not usually cause any serious problem in children. Thiopental precipitates if it is mixed/followed by an acidic drug (e.g., rocuronium), as thiopental requires an alkaline pH to remain soluble in water. Since the precipitate may occlude a small-gauge IV catheter, the IV tubing should be flushed between administration of thiopental and the acidic drug.

Methohexital

Methohexital is rarely used for induction of anesthesia, although some authors have reported that recovery after methohexital is faster than after thiopental. Methohexital often causes muscle twitching or hiccups—effects that can be minimized by avoiding large doses. Intravenous injection of 1% methohexital commonly causes pain along the injected vein; this can be attenuated by adding a small amount of plain lidocaine (e.g., 1 mg lidocaine/mL solution).

Methohexitol is most effective for sedation/induction of anesthesia in apprehensive children younger than 3 years of age by the rectal route, although it is rarely used today. Children may remain in their mother's arms until anesthesia is induced. Doses of 20–30 mg/kg of a 10% solution or 15 mg/kg of a 1% solution induce sedation/anesthesia in 6–8 min. Methohexitol should be administered from a syringe, using a well-lubricated No. 10 soft catheter, which should be inserted into the rectum only 3–4 cm. A diaper should be placed under the child, because soiling sometimes occurs. Rarely, ventilatory obstruction or depression may occur; therefore, children must be observed closely by an anesthesiologist with airway equipment readily available. Once the child is anesthetized, induction may be continued with an inhalational anesthetic; gently assisted ventilation may be needed at this stage.

Benzodiazepine Antagonists

Flumazenil

This antagonist for benzodiazepines has a rapid onset reaching its full effect within 5–10 min of IV administration. A dose of 2–20 µg/kg IV may be repeated as required to reverse the sedative/respiratory effects of the benzodiazepine and then repeated IM to prevent recrudescence. Children who receive flumazenil should be observed for a minimum of 2 h.

Alpha-2 Agonist Drugs

This class of drugs acts at presynaptic alpha-2 receptors in sympathetic nerves and postsynaptic sites in the central nervous system to inhibit the release of norepinephrine and other neurotransmitters, thus affecting the level of consciousness and perception of pain.

Clonidine

Clonidine is used most commonly in pediatric anesthesia as an adjunct to neuraxial blockade with local anesthetics. It is also used as an oral premedication to reduce post-sevoflurane delirium to reduce shivering, and to augment induced hypotensive regimens (see Table 4.5). In children, IV clonidine has a volume of distribution (Vd) of 1 L/kg, elimination half-life of 5.5 h, and clearance of 4.85 mL/kg/min. With a much smaller Vd and greater clearance than in adults, the elimination half-life is approximately one-half that in adults.

When combined with local anesthetics via the epidural route, clonidine 2 µg/kg extends the duration of caudal epidural analgesia by approximately 3 h without side effects. However, there are reports of apnea in preterm infants after the use of epidural clonidine.

For oral premedication, the sedation with 4 µg/kg clonidine is similar to that with midazolam, although the former requires 60–90 min to reach its full effect. Oral atropine should be added to prevent bradycardia. Mild hypotension may also follow its use. Clonidine reduces the cardiovascular responses to tracheal intubation and decreases the MAC for inhaled anesthetics. Analgesic and sedative effects may persist into the postoperative period, decrease agitation, and reduce analgesic requirements. Residual sedation from clonidine may be a safety concern for outpatient surgery.

Dexmedetomidine

Dexmedetomidine is eight times more specific for the α_2 -receptor than clonidine with similar physiologic effects. It has been used in the ICU, in the OR, and in the remote sites offering the advantage of sedation without respiratory depression. It is administered as a continuous IV infusion often after a loading dose. The elimination half-life after IV administration in children is approximately 2 h.

Various regimens have been proposed for effecting sedation with this drug. For MRI, 1 µg/kg loading dose over 10 min combined with 0.1 mg/kg midazolam and followed by an infusion at 0.7 µg/kg/h is effective. Recovery after this regimen in children is slightly delayed when compared with propofol. It has also been used for sedation to secure a difficult airway and to promote hypotension for large blood loss surgery and for cardiovascular stability during CV surgery.

As an oral premedication, it has a faster onset than clonidine. Recently, intranasal dexmedetomidine (1–2 µg/kg) has been used for premedication, although the permeable cribriform plate may facilitate its transfer into the central nervous system (see Table 4.5).

Neuroleptics

Droperidol

Droperidol is a powerful tranquilizer that potentiates sedatives and hypnotics. It has a potent antiemetic effect when administered in small doses. Droperidol was the subject of a “black box” warning by the US Food and Drug Administration after reports of prolonged QT interval and torsades de pointes surfaced in adults

given inappropriately large doses of the drug. Although droperidol may slightly increase the QT interval in children, it does not increase the dispersion of repolarization. Thus, droperidol itself does not pose a risk for torsades in healthy children.

Opioid Medications

Morphine, meperidine, and fentanyl have been administered extensively as part of balanced anesthesia in children. Alfentanil, sufentanil, and remifentanil have also been used; remifentanil may be particularly useful in a TIVA protocol. In addition to providing analgesia, fentanyl and sufentanil in adequate doses may block neuroendocrine and pulmonary vascular responses to intraoperative stress. Meperidine use in children is only indicated as a single dose to treat shivering in PACU; chronic administration is contraindicated due to accumulation of normeperidine, which may cause seizures, particularly in children with compromised renal function.

Morphine

Morphine provides excellent analgesia and sedation and remains a most satisfactory agent for postoperative systemic analgesia. Morphine is used in the perioperative period through the IV, PCA, and epidural and spinal routes. A standard initial dose of morphine in children during surgery is 0.05–0.1 mg/kg IV with additional doses as indicated. Dosing regimens for postoperative morphine IV and PCA are found in Tables 7.3 and 7.4 and for the epidural/caudal and spinal routes in Chap. 5.

Neonates have been considered more sensitive to the ventilatory depressant effects of morphine than to those of meperidine. Various factors have been postulated to account for this apparent sensitivity, including differences in permeability of the blood-brain barrier. Probably the most important factor is the relatively slower, less predictable clearance of morphine in the neonate, which accumulates in the blood during continuous infusions. The decrease in the blood concentration after discontinuation of a morphine infusion may also be delayed in small infants. Indeed, sometimes a transient increase in the blood concentration may be observed, possibly as a result of enterohepatic recirculation. Provided that careful monitoring and suitably low infusion rates are used, morphine can be safely administered by continuous infusion even in the neonate. The infant should be monitored for 24 h after discontinuation of a morphine infusion.

Codeine

Codeine is a naturally occurring opium alkaloid constituting approximately 0.5% of raw opium. It may be administered by the oral, rectal, or IM route but should not be administered by the IV route; severe hypotension may ensue, possibly as a result of histamine release. The oral or IM dose is 0.5–1.5 mg/kg.

Codeine is a prodrug, which must undergo O-demethylation via the CYP2D6 isozyme to release the active analgesic, morphine (5–15% of the administered dose of codeine). CYP2D6 is less active in the neonate, reaching adult level of activity by 10 years of age. This may explain the relative tolerance displayed by children to codeine. However, specific populations also show variations in the 2D6 activity resulting in little analgesia (poor metabolizers) in some patients and opioid toxicity possibly including respiratory arrest or death (ultrarapid metabolizers) in others. Several genetic polymorphisms of CYP2D6 explain these variable responses, e.g., up to 10% of the Caucasians and 30% of Hong Kong Chinese are CYP2D6 poor metabolizers, experiencing little or no analgesia from a standard dose of codeine. In contrast, 29% of Ethiopians and 1% of Swedes and Germans are CYP2D6 ultrarapid metabolizers (due to duplicate wild (or normal) genes or a rapid conversion polymorphism) that may yield large doses of and possible toxicity from unexpectedly large concentrations of morphine. Furthermore, repeated dosing of codeine to individuals with ultrarapid metabolizer polymorphisms and to others with opioid sensitivity (e.g., obstructive sleep apnea and/or obesity who desaturate at night (see Chap. 10)) may be potentially fatal; as a result, this drug is no longer recommended.

Oxycodone. This semisynthetic opioid derived from thebaine with potent antitussive and analgesic properties is prescribed for moderate to severe pain. The oral formulation is ten times more potent than codeine as an analgesic. The pediatric dose of oxycodone is 0.05–0.15 mg/kg orally 4–6 h. It is available in tablet and liquid formulations; caution should be exercised when it is combined with acetaminophen to avoid an overdose of acetaminophen in infants and young children. A sustained-release formulation, OxyContin, is available for oral use; the dose should be reduced in hepatic failure. However, OxyContin abuse has limited its use.

Bioavailability of the oral formulation is 40% and protein binding is 50%. The onset time is 20–30 min with a duration of action of 4–5 h. Terminal elimination half-life is 4 h in neonates and infants <2 months and 2 h in infants 2 months to 8 years. It is metabolized via 3A4 to noroxycodone (major metabolite) and 2D6 to oxymorphone (minor). The dose should be reduced in renal impairment.

Hydrocodone. This semisynthetic mu-receptor opioid analgesic is derived from codeine, with antitussive and analgesic properties for moderate to severe pain. It is 20–50% protein bound in blood, with a time of onset of approximately 20 min and duration of action of 4 h. Its potency exceeds that of codeine; however based on oral bioavailability after gastric absorption, 5 mg of hydrocodone is considered equianalgesic with 30 mg of codeine. Caution should be exercised if it is combined with acetaminophen to avoid an overdose of acetaminophen.

Hydrocodone undergoes hepatic metabolism via both 2D6 (to hydromorphone) and 3A4 (to norhydrocodone) pathway. Hydromorphone is a more potent analgesic than the parent compound, which could render ultrarapid metabolizers of 2D6 susceptible to exaggerated opioid effects (as in codeine). Inhibitors of 3A4 including clarithromycin and valproic acid may delay the elimination of hydrocodone. Furthermore, both hydromorphone and norhydrocodone depend on uridine diphosphate glucuronosyltransferase for their elimination, rendering those with reduced activity of this enzyme susceptible to prolonged effects of these metabolites.

Fentanyl

Fentanyl is a potent but short-acting synthetic opioid. Its metabolism in infants is age dependent: neonates, and especially preterm infants, metabolize fentanyl more slowly than older infants. Increased intra-abdominal pressure (e.g., repaired omphalocele, intestinal obstruction) further slows the clearance of fentanyl by reducing hepatic blood flow. As a sole analgesic agent during anesthesia in neonates, 15–50 µg/kg is required to prevent cardiovascular responses to surgery. Supplemental fentanyl may not be required for 60–90 min. If extubation is planned after surgery, an infusion of 2–4 µg/kg/h may be used to supplement N₂O during balanced anesthesia. However, larger doses should not be given to small infants unless they are ventilated or closely monitored postoperatively. The context-sensitive half-life of fentanyl after continuous infusion increases rapidly with the increased duration of the infusion. Rebound of fentanyl blood levels may occur and may cause depression of ventilation; therefore, if large doses have been given, the child must be carefully monitored. Infants older than 3 months of age may be less sensitive to fentanyl-induced ventilatory depression and have been demonstrated to metabolize the drug more rapidly. Bradycardia may occur after fentanyl unless it is preceded by an anticholinergic agent (e.g., atropine, pancuronium). Chest wall (muscle) rigidity may occur after IV fentanyl; it is infrequent in infants and children.

Infants who receive large doses of fentanyl over a prolonged period may develop tolerance and a significant number may also show signs of dependence.

This effect is common in infants who have been treated with a fentanyl infusion for a period of days. Such infants subsequently require escalating doses to prevent responses to stimulation. It may then be appropriate to substitute another anesthetic or analgesic drug. A methadone program may be required to wean the child off opioid dependency. Neonatal abstinence syndrome may occur when fentanyl is withdrawn after continued use for as little as 7 days; it is characterized by crying, hyperactivity, fever, tremors, abdominal distention, poor feeding and sleeping, and—in the extreme case—vomiting and convulsions. These sequelae can be avoided by tapering the opioid dose slowly, but may require intervention with methadone.

Remifentanil

Remifentanil, an ultrashort-acting synthetic opioid, represents a new class of opioids that must be administered as a continuous intravenous infusion. It is available as a lyophilized powder and requires reconstitution. The kinetics of remifentanil are unique: its elimination half-life, which is 3–10 min, is independent of both the dose and the duration of administration of the infusion as well as hepatic and renal dysfunctions. Its context-sensitive half-life is constant, approximately 4 min. Its action is terminated by hydrolysis of an ester bond by ubiquitous tissue esterase enzymes.

The intravenous loading dose is 0.5–2.0 µg/kg, and the maintenance dose is 0.05–2.0 µg/kg/min. The loading dose should be administered slowly to prevent chest wall rigidity. As an analgesic, the potency of remifentanil is 20–30-fold greater than that of alfentanil. The maintenance dose is reduced by half if a potent inhaled agent is administered. This is the only opioid with a shorter elimination half-life in neonates and infants less than 4 months of age than adults; this may offer a great advantage in instances where an intense but brief opioid effect is desired. The rapid offset of the analgesia once an infusion of remifentanil is discontinued requires that an alternative analgesic be administered before discontinuing remifentanil to prevent severe acute pain. This drug should only be administered by an infusion pump (best with a separate carrier by pump) and is best diluted to 5 µg/mL in neonates and infants, compared with the dilution of 50 µg/mL in adults. Remifentanil is a very useful adjunctive agent in a TIVA protocol. The side effects from remifentanil are similar to those of other opioids and include bradycardia, hypotension, apnea, chest wall rigidity, and vomiting. A vagolytic agent may be indicated when remifentanil is given to infants and children to prevent bradycardia. Acute tolerance after remifentanil has been reported, but is dose dependent; tolerance is less likely after smaller (0.3 µg/kg/min) rather than larger (>0.6 µg/kg/min) infusion rates.

Hydromorphone (Dilaudid)

Hydromorphone is a potent mu-receptor opioid that is more lipophilic and 5–7 times more potent than morphine. It is used to treat acute pain as an IV bolus or infusion (including PCA) as well as epidurally in children. The dose of hydromorphone is 0.03–0.08 mg/kg/dose q3–4 h orally and 0.01–0.02 mg/kg/dose IV q3–4 h. Continuous IV or epidural infusion rate is 1 µg/kg/h. Hydromorphone may also be used with PCA: bolus doses of 2–4 µg/kg with a lockout of 8–15 min are recommended with an optional basal infusion rate of 1–5 µg/kg/h.

It is available orally, with rapid duodenal absorption (within 30–60 min) resulting in 60% bioavailability as a result of first-pass metabolism. Hydromorphone is less than 20% protein bound, 95% is eliminated as the 3 glucuronide metabolite. Clearance is double that in adults, with a large volume of distribution that results in an elimination half-life, 1–3 h, similar to that in adults. Renal and hepatic failure delay elimination of both the parent compound and the 3-glucuronide metabolite. Although the metabolite is inactive, it may accumulate in renal/hepatic insufficiency producing neuroexcitatory side effects including allodynia and seizures.

Methadone

This racemic opioid is increasingly used in infants and children because of its prolonged duration of action, inactive metabolites, and lipophilicity. Parenteral methadone is fivefold more potent than morphine; thus, 20 µg/kg methadone should provide equivalent analgesia to 100 µg/kg morphine. It has a high bioavailability after oral administration. Protein binding in blood ranges from 60 to 90%. The clearance and volume of distribution are independent of age in childhood despite its dependency on CYP3A4 for 60% of its metabolism, with lesser contributions from CYP2D6 and CYP2B6. The metabolites are inactive analgesics. Elimination half-life for the racemate is 15–60 h. Side effects are similar to those of other opioids including apnea, nausea and vomiting, and sedation. Methadone has been reported to prolong QT interval, although there is no evidence that it increases the dispersion of repolarization. It has been associated with torsades de pointes and ventricular fibrillation, which have occurred in patients with chronic cardiac or liver disease or those abusing drugs.

Sufentanil

Sufentanil is ten times more potent than fentanyl and has a smaller elimination half-life. Clearance is slower in infants younger than 1 month of age. Sufentanil has been administered in large doses during cardiac surgery in infants, producing cardiovascular stability with minimal depression of ventricular function. Sufentanil in large doses may favorably influence the metabolic and neuroendocrine response to major cardiovascular surgery in infants.

Alfentanil

Alfentanil has a more rapid onset and a shorter duration of action than fentanyl and a greater incidence of vomiting than other opioids. It is less lipophilic than fentanyl and is extensively protein bound. Most of the drug is metabolized in the liver; <1% is excreted via the kidney unchanged. Clearance is slower and more variable in young infants, especially preterm infants. Otherwise, in older infants and children, the pharmacokinetics are similar to those in adults. The drug has minimal cardiovascular effects. Alfentanil, 35 µg/kg as a bolus followed by intermittent doses of 10 µg/kg every 10–15 min, has been suggested as suitable for children. A continuous infusion may be used but in this case remifentanil (see previous discussion) may be the preferred drug. Recovery after alfentanil is reported to be very rapid and complete. However, this is a very potent drug, and all children should be closely observed for signs of residual or recurring respiratory depression.

Opioid Antagonists

Provided opioids are carefully administered; it is rare that they need to be antagonized after general anesthesia. Remember that if you completely antagonize the opioid, your patient may experience severe pain and that resedation/airway compromise may recur.

Naloxone Hydrochloride

Naloxone hydrochloride (Narcan), an *N*-allyl derivative of oxymorphone HCl, antagonizes opioids; respiratory depression may be reversed with as little as 0.5 µg/kg IV, although larger doses (up to 100 µg/kg) may be required. Repeated small doses titrated to effect to reverse opioid-induced respiratory depression without affecting the analgesia is preferred unless apnea has occurred in which case a full dose is indicated. The same cumulative dose that reversed the respiratory depression may also be administered intramuscularly to prevent recrudescence. Naloxone is contraindicated in children who may be opioid dependent.

Neuromuscular Blocking Drugs

Neuromuscular blocking drugs are widely used to facilitate tracheal intubation and to provide muscle relaxation during controlled ventilation and surgery. The pharmacodynamic effects in infants often differ compared with adults. The neuromuscular junction in infancy has less reserve than that of the adult. Fade occurs at high rates of stimulation. This has led to the suggestion that infants show a myasthenic response and would be sensitive to non-depolarizing relaxants.

In fact, to produce a similar degree of block, infants and adults require similar doses of relaxants on a milligram-per-kilogram basis, but the variability in dose requirements and intervals between doses are greater. This may be attributed to the combined effects of a larger V_d plus a greater degree of block for a specific plasma concentration in the infant. It is prudent to monitor the degree of neuromuscular block to guide dosage.

Depolarizing Muscle Relaxant

Succinylcholine

Succinylcholine is the only depolarizing relaxant in clinical use. Its onset and offset of action are more rapid than those of other relaxants. Succinylcholine is metabolized by pseudocholinesterase of which there are five major (and several minor) alleles: typical (E^u), atypical (E^a), fluoride resistant (E^f), silent gene (E^s), and C₅ variant (or Cynthiana). E^a, E^f, and E^s have reduced pseudocholinesterase activity, with the homozygote genotypes expressing less activity than the heterozygote genotype thus prolonging the duration of action of succinylcholine, whereas the C₅ variant is an ultrarapid metabolizer, abbreviating the duration of paralysis.

Succinylcholine may be administered IV, IM, or intralingually. IV succinylcholine (2 mg/kg) has an onset of action of 20–30 s in children, reaching its maximum effect within 40 s. IM succinylcholine (4–5 mg/kg) has a slower onset of action compared with the IV route. Intralingual or submental succinylcholine has been used for emergencies when other routes are not accessible; we do not recommend this route because puncture of the vascular supply could cause a lingual hematoma. Infants and children require a relatively larger dose of succinylcholine than adults (2 vs. 1 mg/kg, respectively) due to their large extracellular fluid compartment.

Bradycardia occurs commonly after a single dose of IV succinylcholine in infants and children. This can be prevented by prior administration of intravenous atropine (0.02 mg/kg) or glycopyrrolate (0.01 mg/kg). IM succinylcholine (4–5 mg/kg) changes the heart rate and rhythm minimally, even in anesthetized children who have not received atropine.

Although myoglobinemia and myoglobinuria occur more commonly after succinylcholine in children than in adults, especially if halothane precedes succinylcholine, the incidence of strong fasciculations and muscle pain is less. Nonetheless, children who are ambulatory may benefit from pretreatment with a non-depolarizing relaxant (10% of an intubating dose) to reduce postoperative

muscle pain. Serious rhabdomyolysis, severe myoglobinuria, and hyperkalemic cardiac arrest may occur in children with myopathies (including Duchenne muscular dystrophy).

Masseter spasm has been reported in 1% of children who received IV succinylcholine after a halothane induction. However transient masseter spasm is a normal variant following succinylcholine and generally relaxes allowing the mandible to be opened. However a very small percentage of such cases result in masseter tetany ("jaws of steel") during which the mandible cannot be opened preventing insertion of the laryngoscope blade; generally these children can still be readily ventilated as they are otherwise paralyzed. This condition is strongly associated with MH and the halothane-caffeine contracture test is positive in ~50%, and the child should be monitored and investigated accordingly (see Chap. 6). In contrast, masseter spasm is extremely rare after an IV induction of anesthesia with thiopental (with atropine) followed by succinylcholine (1:3000 incidence). If masseter spasm occurs, it must be considered a significant warning sign of possible MH. The sequence of thiopental (or propofol), atropine, and succinylcholine do not commonly result in masseter spasm and remains the most effective means of securing the airway rapidly.

Succinylcholine does not decrease the gastric-esophageal barrier pressure. In part, because some muscles of the crura of the diaphragm are not skeletal in origin. Intraocular pressure increases transiently after succinylcholine administration (5–10 mmHg) because of greater tension in the smooth muscles lining the globe of the eye and possible dilation of the choroidal blood vessels. Although succinylcholine has been reported to transiently increase intraocular pressure, studies have indicated that large doses of thiopental attenuate this response. If intraocular pressure is to be measured (i.e., in glaucoma), or if forced duction testing is planned (i.e., in strabismus surgery), it may be best to avoid succinylcholine. If succinylcholine is considered for a child with an open globe injury, the ophthalmologist should be consulted as pinpoint punctures of the globe are less likely to lead to extrusion of intraocular contents after succinylcholine than are large (>4 mm) lacerations. Alternatively, high-dose rocuronium (1.2 mg/kg) provides similar intubating conditions in children 30 s after administration.

Serum potassium levels do not increase in children with cerebral palsy but do increase after succinylcholine administration in children with burns beyond the first 24 h, crush injury, muscle and neurologic diseases (upper and lower motor neuron lesions), chronically bedridden, and renal failure. These increases in K⁺ may result in ventricular tachycardia. Intravenous calcium chloride (or gluconate), and other measures to treat hyperkalemia must be initiated immediately to restore cardiac rhythm and output.

Non-depolarizing Neuromuscular Blocking Agents: Intermediate Duration

Rocuronium

Rocuronium is a steroid-based, non-depolarizing muscle relaxant that has a more rapid onset of action and greater stability in solution at room temperature than vecuronium. The rapid onset of action of rocuronium stems from its reduced potency, one-sixth that of vecuronium. The speed of onset of blockade is dose dependent. The ED₉₅ in infants is ~0.3 mg/kg and in children ~0.45 mg/kg IV. Inhalational anesthetics, notably sevoflurane, prolong the duration of action of neuromuscular blockade. The duration of action of an intubating dose (twice the ED₉₅), 0.6 mg/kg, in infants during nitrous oxide/opioid anesthesia is 50% greater than that in older children, 42 vs. 27 min. Recovery after similar weight-based dosing increases with decreasing age. A dose of 1.2 mg/kg is recommended for rapid sequence intubations with an expected prolonged duration of effect (~75 min). Rocuronium may also be administered as a continuous infusion (10 µg/kg/min and adjusted up or down by 2–3 µg/kg/min according to the train-of-four response). Rocuronium is minimally metabolized by the liver (possibly via CYP3A4), excreted primarily through the biliary system. Elimination is unaffected by renal failure, but may be prolonged by liver failure.

Rocuronium causes pain on injection, an effect that may be mitigated by prior treatment with lidocaine or remifentanil. It is devoid of cardiovascular and histamine effects. Low-dose rocuronium (0.3 mg/kg) provides satisfactory intubating conditions after 3 min and can usually be antagonized within 15–20 min. It should be noted that coadministration of thiopental and rocuronium precipitates thiopental and may block the IV cannula.

Vecuronium

Vecuronium (lyophilized powder) is an intermediate-acting non-depolarizing steroid neuromuscular blocking agent devoid of cardiovascular effects and histamine release that has effectively been replaced by rocuronium. Its duration of action in children is 35–45 min, but in neonates, its duration is much greater, 70 min or more. The ED₉₅ for vecuronium is the following: infants, 47 µg/kg; children 2–10 years old, 81 µg/kg; and adolescents, 55 µg/kg. The duration of action of vecuronium increases in the presence of some forms of liver disease and impaired renal function. Because vecuronium has no vagal blocking effect, bradycardia may occur if vagotonic drugs (e.g., fentanyl, halothane) are coadministered; atropine may be required. Vecuronium may also be administered as an infusion. Infants require a smaller infusion rate, ~1–2 µg/kg/min, than older children, ~2–3 µg/kg/min.

Atracurium

Atracurium, a benzylisoquinolinium, is a non-depolarizing neuromuscular blocking agent with an intermediate duration of action (~30 min). It is a mixture of ten stereoisomers that vary in potency and side effects. It is used infrequently now, having been replaced by *cis*-atracurium (see later discussion). Atracurium degrades spontaneously at physiologic pH to inactive compounds (Hofmann elimination) and hence has a predictable rate of elimination that is unaffected by the presence of severe hepatic or renal disease. Its brief duration of action and constant rate of metabolism made this drug ideal for administration by continuous infusion and for use in neonates with immature organ function. In children, an initial bolus of 0.3–0.4 mg/kg followed by an infusion of ~9 µg/kg/min (and the adjusted up or down by 1–2 µg/kg/min according to the train-of-four response) during TIVA and 6 µg/kg/min during an inhalational anesthetic provides satisfactory relaxation; note that infants <1 month of age will require ~25 % less. Slightly larger doses are required if opioids are substituted for inhaled agents. Atracurium usually has little effect on the cardiovascular system. It does release histamine, especially if large doses are given rapidly and should not be given to children with asthma as bronchospasm has occurred. A rash is common, but significant hypotension is uncommon. Rarely, precipitous and severe hypotension has occurred after atracurium, especially when it has been given in a large dose (more than 0.4 mg/kg) and preceded by thiopental.

Cis-Atracurium

Cis-atracurium, one of the ten stereoisomers of atracurium besylate, confers the most stable hemodynamics and least histamine release of the isomers having largely supplanted atracurium in clinical practice. It has a relatively slow onset of action. The dose for tracheal intubation is 0.15–0.2 mg/kg, which yields a duration of action of approximately 35 min. *Cis*-atracurium may also be administered by infusion at a dose of 1–2 µg/kg/min. Termination of its action is similar to atracurium, via Hofmann elimination and ester hydrolysis. The duration of action of *cis*-atracurium is unaffected by renal or hepatic failure; therefore, it is a drug of choice in such situations and in neonates with immature renal and hepatic function.

Prolonged Duration Relaxants

Pancuronium

Pancuronium is the only non-depolarizing relaxant with a prolonged duration of action. It is excreted principally via the kidney so its elimination half-life may be quite prolonged in those with impaired renal function.

An initial dose of 0.1 mg/kg permits intubation in about 2 min. Supplementary doses should be given carefully, using a nerve stimulator for guidance; each dose should be only 10–20 % of the initial paralyzing dose. Pancuronium has a vagolytic effect and causes an increase in heart rate and blood pressure, particularly when given as a rapid intravenous bolus. These effects are more pronounced in younger patients. In preterm infants, pancuronium causes a sustained tachycardia and hypertension and increased plasma epinephrine level. In practice, if pancuronium is combined with fentanyl, their effects on heart rate offset one another resulting in a relatively stable hemodynamic situation. Pancuronium causes little histamine release and is the preferred relaxant for prolonged surgery in children with asthma.

Antagonism of Neuromuscular Blockade

Non-depolarizing neuromuscular blocking agents should always be antagonized unless the child has completely recovered normal neuromuscular function as documented with a nerve stimulator. Antagonism may not be fully effective in children who are hypothermic (less than 35 °C); therefore, controlled ventilation is often continued until they are rewarmed. Rarely, antibiotics potentiate the neuromuscular blocking drugs in infants or children to the extent that they cannot be antagonized. This possibility must be considered, especially in those receiving aminoglycoside derivatives (e.g., neomycin, gentamicin, tobramycin).

The adequacy of antagonism may be difficult to judge, especially in infants. Hence, antagonism of neuromuscular blockade is recommended for all neonates and infants below 2 years of age. The train of four should demonstrate four equal contractions. Muscle tone can be examined and is often best judged in neonates and infants by flexion of the elbows and hips. The ability to generate a negative inspiratory pressure of 25 cm H₂O is also a useful index. When any doubt whatsoever exists about the adequacy of the antagonism, controlled ventilation should continue and recovery of neuromuscular function should be reevaluated periodically using a nerve stimulator.

Commonly used regimens to antagonize the non-depolarizing muscle relaxants in infants and children are as follows:

1. Neostigmine (50 µg/kg) mixed with atropine (20–25 µg/kg) is most effective and results in few and insignificant cardiac arrhythmias even in those with congenital heart disease. Glycopyrrolate (0.01 mg/kg) may not prevent neostigmine-induced bradycardia. In those in whom tachycardia is to be avoided, e.g., hypertrophic obstructive cardiomyopathy (HOCM) or aortic stenosis, it may be prudent to avoid neostigmine and edrophonium altogether, or if required, use an anticholinesterase in combination with glycopyrrolate.

2. Edrophonium (1 mg/kg) after atropine (20–25 µg/kg) may be used. Edrophonium has a more rapid onset of action than neostigmine, and its vagotonic action appears early. These findings have led to the practice of administering atropine before the edrophonium.
3. Sugammadex is a cyclodextrin that has been developed to trap rocuronium and eliminate it via the kidneys. It replaces anticholinesterases thus reducing the time to antagonize neuromuscular blockade (since sugammadex has a rapid onset of action) and avoiding side effects of anticholinesterases. Sugammadex (16 mg/kg IV) can reverse large doses of rocuronium within 1.5 min. Sugammadex has been approved to reverse rocuronium-induced neuromuscular blockade in adults in Europe and recently in the USA. Experience in pediatric patients is limited at present.

Nonsteroidal Anti-inflammatory Drugs

Acetaminophen

Acetaminophen is an analgesic and antipyretic drug without anti-inflammatory actions, but is commonly grouped with the NSAIDs. It is useful as an analgesic for mild pain and is also useful as an opioid-sparing adjunct for more severe pain. Oral dosing is 10–15 mg/kg q 4–6 h. For rectal administration, a loading dose of 35–45 mg/kg is recommended followed by 20 mg/kg PR q6h. The time to peak effect after oral administration is 10–20 min and after rectal administration is 60–180 min. The total daily dose should not exceed 100 mg/kg. *Beware: hepatic failure may occur with overdose and is a particular risk in the seriously ill debilitated child.*

There are two formulations of IV acetaminophen: paracetamol and proparacetamol. The latter is a water-soluble parenteral prodrug of acetaminophen (1 g = 0.5 g acetaminophen) that is available in Europe and many other countries outside of North America. Rapid hydrolysis by nonspecific esterases releases the active compound, paracetamol, within 10–30 min of IV administration. The pharmacokinetics are independent of age (beyond neonates), and the drug should be administered on a weight basis. In neonates, clearance is markedly diminished; with increasing age, clearance increases.

The recommended dose of IV paracetamol (Perfalgan) is 7.5 mg/kg for infants <10 kg, 15 mg/kg for children 11–50 kg, and 1 g for those >50 kg q6h. These doses provide adequate relief for mild to moderate pain in children. The maximum daily dose of IV paracetamol is 30 mg/kg in neonates, 60 mg/kg in infants and children (<50 kg), and 4 g in those >50 kg.

Several cases of IV paracetamol overdose that led to transient hepatic insufficiency in young children and a death in a young adult have been reported. These overdoses were attributed to calculation errors, a dosing mix-up between the prodrug and the native drug and a lack of knowledge of the maximum daily dose limit for IV paracetamol. Caution must be exercised when dosing IV acetaminophen to document the concentration of the acetaminophen formulation, the correct dose, and the maximum allowable daily dose. When an overdose is suspected, N-acetylcysteine (NAC) must be given as quickly as possible to limit the hepatotoxicity. NAC may be given by inhalation, orally, or IV. The oral route is not preferred because of the poor bioavailability; the IV route is effective although severe anaphylactic-like reactions have been reported in 3–6% of patients via this route. Most commonly, NAC is administered by inhalation.

Ibuprofen

Ibuprofen (4–10 mg/kg PO q6h max. 40 mg/kg/day) reduces postoperative morphine requirements and improves pain relief in children. Alternating doses of acetaminophen and ibuprofen may be a useful alternative to opioids after tonsillectomy in small children. An IV formulation is now available.

Diclofenac

Diclofenac is a nonsteroidal anti-inflammatory agent that is available in oral (25–75 mg tablets), intravenous, and rectal formulations (50 or 100 mg suppositories). Outside of North America, rectal formulations as small as 12.5 mg per suppository are available for use in children. Absorption and bioavailability after rectal administration is greater than after oral administration. Suggested pediatric dosing is 0.3 mg/kg IV and 1 mg/kg orally and rectally every 8 h. It provides similar analgesia to IV paracetamol. Rectal diclofenac is widely used in Europe for children after minor superficial surgery. Its effect on platelet activity is intermediate between placebo and ketorolac.

Ketorolac

Ketorolac is a nonsteroidal analgesic racemic mixture that is available for parenteral use either intramuscularly or intravenously. It is considered a moderately potent analgesic that is devoid of respiratory depression, vomiting, sedation, and urinary retention effects. In clinical trials, ketorolac has been shown to be effective and opioid sparing. The dose for children more than 6 months of age up to children <50 kg is 0.5 mg/kg IV (max. 15 mg/dose), repeated every 6 h (max. daily dose of 60 mg). The dose for children >50 kg is 0.5 mg/kg IV up to 30 mg/dose or 120 mg/24 h. The elimination half-life of ketorolac in children is similar to that in adults: approximately 4 h.

Ketorolac should be used only for brief periods (i.e., several days at a time) because of the risk of nephrotoxicity. Ketorolac should be administered with caution to children with impaired renal or hepatic function. Like most nonsteroidal anti-inflammatory drugs (NSAIDs), ketorolac reversibly inhibits platelet function and may cause increased bleeding, especially if it is administered before or early in the surgical procedure. Postoperatively, ketorolac does not appear to increase bleeding, although most clinicians avoid it in surgeries where bleeding may be a problem (adenotonsillectomy, cleft palate, etc.). There are limited data in animals that suggest ketorolac may impair bone healing, prompting some orthopedic surgeons to avoid this agent.

Tramadol

Tramadol is a weak mu-opioid receptor agonist that also inhibits the reuptake of noradrenaline and serotonin. The dose is 0.5–1 mg/kg with a maximum dose of 50 mg every 3–4 h. Its elimination half-life, 6–7 h, is greater than other oral analgesics. This is compounded by an active metabolite with a 10-h elimination half-life. Tramadol is metabolized by 2D6 to the O-methylated metabolite, which holds 200-fold greater affinity for the mu-opioid receptor than tramadol. Both the ontogeny and polymorphisms of 2D6 and renal maturation may impact the analgesic potency of Tramadol in young children. Tramadol causes less sedation and respiratory depression than opioids. It may lower the seizure threshold in children and is best avoided in those with seizures.

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Techniques and Procedures

ROUTINE PREPARATION FOR SURGERY

Preoperative Feeding Orders

Infants and children should not be subjected to prolonged preoperative fasting. Excessive fluid restriction rapidly leads to dehydration and hypovolemia because of their high metabolic rate; hypotension in infants during induction of anesthesia has been shown to be directly related to the duration of fasting. In addition, excessive fasting may precipitate hypoglycemia and/or metabolic acidosis, particularly in young infants. Healthy children may safely be given unlimited clear fluids up to 2 h before induction. The volume and acidity of the gastric contents are not increased when this regimen is used. Indeed, clear fluids 2 h before anesthesia may reduce gastric contents at induction, possibly by stimulating gastric emptying. Furthermore, this practice reduces hunger and thirst and makes for a happier child. Fasting guidelines need not be adjusted for obese children; the gastric fluid volume and pH after fasting are similar to those in nonobese children. Fasting guidelines are presented in Table 4.1.

Children who are scheduled for afternoon surgery may have a light breakfast (dry toast and clear fluids) early in the morning. These rules must, of course, be modified in special cases (e.g., for diabetics). Children who require emergency surgery, those with gastrointestinal disease, and any others at increased risk for vomiting during induction should receive intravenous fluids and a rapid sequence induction. When evaluating injured children, the interval between last food or fluid and the injury is the best predictor of the likely stomach contents. It is also suggested that ultrasound may be used to assess stomach contents. Children for whom any period of fluid deprivation might pose a risk (e.g., those with cyanotic heart disease) should have an intravenous infusion established at the commencement of any prolonged period of restricted oral intake.

Table 4.1 Preoperative fasting intervals for elective surgery

	Fasting interval (h)	Comments
Clear fluids	2	Water, ginger ale, apple juice; any particle-free fluids
Breast milk	4	
Formula/cow's milk	6	Also dry toast, black tea
Solids	8	

N.B.: Chewing gum should be expectorated; if swallowed the anesthetic should be delayed 8 h

Preoperative Assessment

A careful assessment of the child must be made during the preoperative visit. Although most children are healthy, some have diseases with significant anesthetic implications. A thorough history should be obtained both from the parents and from any medical records that are available. Review the systems and search for special problems that may complicate anesthesia (Table 4.2). When a significant history is obtained, it is important to establish the current status of the disease; this may require consultation with the pediatrician, the surgeon or radiologist, and other physicians. Pertinent historical questions for children include the delivery date, if preterm, any problems in the neonatal period (e.g., apnea risk), if they required a breathing tube (possible subglottic stenosis), family history of sickle cell disease (has the child been tested), problems with anesthesia (malignant hyperthermia, pseudocholinesterase deficiency), or family history of neuromuscular diseases (muscular dystrophy). Although most children take no medications, some may be taking medications that have implications for management during the perioperative period. If a child is taking routine medications, e.g., for seizure control, parents should be advised to administer them to the child with a sip of clear fluids the morning of surgery, document all medications, neutraceuticals, and drug allergies, and elicit details of any “allergic” reactions; many “allergies” are in fact sensitivities. The majority of rashes (e.g., associated with an oral antibiotic) are not immune-related allergic reactions. Common concurrent medications for children and their significance to anesthetic management are detailed in Table 4.3 (N.B. Some herbal preparations may have anesthetic implications [Table 4.4]). Throughout the preoperative interview, strive to gain the confidence of the child and the parents; always invite questions from both the parents and the child so as to help establish rapport and confidence.

Table 4.2 Review of the medical history—possible implications for anesthesia

Systems	History	Concerns for the anesthesiologist
Central nervous system	Seizures	Adequacy of seizure medication, recent control of convulsions. Phenytoin increases nondepolarizing relaxant and fentanyl requirements, produces gingival hyperplasia and bleeding, and may cause hepatic dysfunction. Ketamine and methohexitol are relatively contraindicated
Hydrocephalus		Possible raised intracranial pressure. Repeated anesthesias. Possible need for prophylactic antibiotics to prevent shunt infection
Head injury		Possible raised intracranial pressure. Current status
Cerebral tumor		Possible raised intracranial pressure, vomiting, change in electrolyte status. Chemotherapeutic agents and possible drug interactions
Cerebral palsy		Nutritional status, presence of chronic infections. Possible history of chronic aspiration and difficulties with positioning. Intelligence may be normal—careful psychological preparation needed
Down syndrome		Optimal cooperation at induction may be a problem. (Possibly get help from parent.) Airway (large tongue, subglottic stenosis). Heart disease. Evidence of joint hypermobility and indications of atlanto-axial instability
Neuromuscular disease		Difficulty positioning. Repeated surgery—careful psychological preparation needed
Meningomyelocele		Associated hydrocephalus. History suggesting latex allergy. Renal infections. Impaired renal function

Table continues on the following page.

Table 4.2 (continued)

Systems	History	Concerns for the anesthesiologist
Cardiovascular system	Heart murmur	Innocent murmur vs. significant lesion. Need for prophylactic antibiotics. Risk of paradoxic embolism
	Cyanosis	Hemoconcentration—avoid fluid restriction; risk of hyperviscosity. Coagulopathy secondary to hyperviscosity; check coagulogram. Reduced response to hypoxemia—caution with premedication
	Dyspnea, tachypnea	Evidence of congestive cardiac failure. History of digoxin and/or diuretic therapy. Digoxin level
	Sweating	Electrolyte levels
	Hypertension	Previous heart surgery Cardiac conduction defects. Pacemaker. History of arrhythmias Renal disease, coarctation of the aorta, endocrine disease
Respiratory system	Prematurity	Risk of perioperative apnea. If previously intubated, risk of subglottic stenosis
	Respiratory distress syndrome	Present postconceptual age, gestational age at birth. Anemia. History of apnea, residual chronic respiratory disease, impaired gas exchange. History of prolonged ventilation, residual subglottic stenosis. Possible bronchopulmonary dysplasia and need for diuretics and supplemental oxygen
	Recent upper respiratory tract infection	Evidence of acute infection. Pyrexia. Lower respiratory tract infection. Reactive airways prone to secondary infection. Increased risk of laryngospasm, bronchospasm, and oxygen desaturation
	Bronchiolitis	Reactive airways, evidence of bronchospasm
	Croup	Possible subglottic stenosis. Avoid intubation, use LMA if possible
	Asthma	Reactive airways. Current status. Theophylline therapy (blood level). β -Agonist drug therapy, history of corticosteroid therapy (prescription supplements). Develop a plan to ensure optimal status preoperatively. Avoid intubation if possible
	Cystic fibrosis	Present pulmonary function. Any acute infection? Can condition be improved? Can regional analgesia be used? Present drug therapy. Nutritional status. Emotional status. Inhaled hypertonic saline prior to induction and as soon as possible in PACU to mobilize secretions

Gastrointestinal system	Gastroesophageal reflux Vomiting	Evidence of aspiration pneumonia. Reactive airways and bronchospasm. Recent food intake, risk of regurgitation, and need for antacid and H ₂ -blockers. Evidence predicting difficult intubation danger Nutritional and hydration status. Electrolyte values. Urine output. Immediate full stomach
	Diarrhea Liver disease	Nutritional, fluid, and electrolyte status. Risk of hypoglycemia and hypovolemia Drug metabolism. Decreased requirements for nondepolarizing relaxants
Cenitourinary system	Renal failure	Anemia and coagulopathy, electrolyte abnormality especially hyperkalemia, volume status. Acid-base status. Hypertension, pericardial effusion, and incipient congestive cardiac failure. Date of last dialysis. History of infection? Impaired immunity? Psychological status
	Bladder surgery Bladder extrophy	Is history suggestive of latex allergy?
Endocrine system	Diabetes mellitus Thyroid disease Pituitary disease Adrenal disease	Current status and therapy. Plans for perioperative management. Need for planning with surgeon and endocrinologist Current status and medication? Euthyroid? Enlarged thyroid effect on the airway Intracranial pressure? Adrenal insufficiency? Thyroid function? Diabetes insipidus? Need for corticosteroid therapy? Volume and electrolyte status
Hematopoietic system	Anemia Bruising or bleeding Sickle cell disease	What is cause? Possible medical therapy. Urgency of surgery. Will anemia affect the course of anesthesia? Is transfusion indicated? Is coagulopathy present? Are further tests required? Preoperative therapy? Order blood products. Are the bruises in unusual locations and indicative of possible child abuse? Trait or disease? Are other abnormal hemoglobins present (Hb electrophoresis results)?
Muscular system	Muscular dystrophy	Is preoperative preparation such as transfusion to a hemoglobin of 10 g/dL required? Risk of hyperkalemia with succinylcholine. Avoid depolarizing relaxants. Use low or no doses of nondepolarizing muscle relaxants if possible. Avoid inhalation agents that might lead to rhabdomyolysis. Ventilatory reserve? Cardiac function? Will postoperative ICU admission be necessary?
Immune system and allergy	Latex allergy	Allergy to bananas, eggplant, passion fruit, and other fruits/vegetables Possibility of concurrent latex allergy (see p. 174 for further details)

Table 4.3 Concurrent medication—possible implications for anesthesia

Drug	Implications for anesthesia
Analgesic, anti-inflammatory Acetylsalicylic acid (ASA, aspirin)	Prolonged bleeding time due to platelet inactivation; check bleeding time if ASA given within 10 days
Nonsteroidal anti-inflammatory drugs (NSAIDs; e.g., ibuprofen, ketorolac)	Affect platelet aggregation and prolong bleeding time. Effect of antihypertensive agents may be decreased. Ketorolac decreases diuretic effect of furosemide
Antibiotics	Many of the “mycins” may potentiate neuromuscular blockade. Monitor neuromuscular block, check reversal carefully
Aminoglycosides	May potentiate succinylcholine and nondepolarizing relaxant drugs. Renal toxicity
Clindamycin	Cardiac depression when given rapidly. May potentiate nondepolarizing relaxant drugs
Erythromycin	May prolong the effect of alfentanil and midazolam. Decreases theophylline clearance rates. Potentiates anticoagulant effect of warfarin
Gentamicin	May prolong the effect of succinylcholine. Potentiates nondepolarizing relaxant drugs
Vancomycin	Potentiates nondepolarizing relaxant drugs. Rapid administration (<1 h) may cause “red man syndrome” with severe cardiovascular collapse
Anticancer agents	All may cause blood dyscrasias, coagulopathy, anorexia, nausea, stomatitis, and reduced resistance to infection
Doxorubicin (Adriamycin)	Cardiotoxic, may cause arrhythmias
Daunorubicin (Cerubidine)	Severe cardiac depression with halothane, especially likely when cumulative dose exceeds 250 mg/m ² (or 150 mg/m ² plus radiation)
Bleomycin	Pulmonary fibrosis—may be exacerbated by excess oxygen. Limit inspired oxygen concentration carefully
Busulfan	Inhibits plasma cholinesterase. May prolong the effect of succinylcholine
Cyclophosphamide	Inhibits plasma cholinesterases; prolonged effect of succinylcholine
Anticonvulsants Phenytoin	May cause blood dyscrasias, hypotension, bradycardia, arrhythmia
Mephenytoin	May increase requirements for nondepolarizing relaxants and may cause peripheral neuropathy
Valproic acid	May cause hypotonia. Hepatotoxic
Antihypertensive drugs	Severe hypotension may occur with potent anesthetics, especially if dehydrated

Table continues on the following page.

Table 4.3 (continued)

Drug	Implications for anesthesia
Captopril	Hyperkalemia with potassium-sparing diuretics (spironolactone). Indomethacin reduces antihypertensive effect
Clonidine	Must not be abruptly withdrawn—severe hypertension may result. Interaction with α -blockers—bradycardia, hypotension
Hydralazine (Apresoline)	May cause systemic lupus erythematosus (SLE)-type syndrome. Decreases tachycardia with atropine
Labetalol	Cimetidine may potentiate labetalol's action
Prazosin (Minipress)	May potentiate effects of ketamine. Diuretics potentiate antihypertensive effect
Antiviral agent Acyclovir	Nephrotoxic, bone marrow depression
α -Agonist agents (e.g., albuterol, Alupent)	May cause tachycardia, hypertension, arrhythmia. Albuterol has increased effect with tricyclic antidepressants or monoamine oxidase inhibitors. Blocked by α -blocking drugs
α -Blocking drugs	May cause bronchospasm, block effects of albuterol. Potentiate cardiac depression caused by halothane. May cause bradycardia with anticholinesterase drugs (e.g., neostigmine) and hypoglycemia
Calcium channel blockers: Verapamil, Nifedipine	Potentiate nondepolarizing relaxant drugs. Severe bradycardia or heart block with α -blocking drugs. Potentiate cardiac depression with potent inhalation agents May interact with α -blockers to cause severe cardiac depression
Corticosteroid preparations	Chronic therapy may lead to depression of the hypothalamic pituitary axis; severe collapse may occur perioperatively. Supplemental steroid therapy should be administered preoperatively
Digoxin	May potentiate bupivacaine toxicity Hypokalemia, if induced (e.g., by hyperventilation), predisposes to arrhythmias
Diuretics	All may result in electrolyte disturbances
Acetazolamide (Diamox)	Produces hyperchloremic metabolic acidosis
Furosemide	Hypokalemia, if present, may prolong action and delay reversal of relaxant drugs
Ophthalmic topical drugs Echothiophate (anticholinesterase)	Inhibits plasma cholinesterases Prolonged apnea with succinylcholine
Phenylephrine	May cause tachycardia and hypertension
Timolol (α -blocker)	May exacerbate asthma
Theophylline	Severe arrhythmias may occur with halothane; check blood level

Table 4.4 Some herbal preparations and their anesthesia implications

Herbal preparation	Reason for use	Anesthesia implication
Echinacea	URTI prophylaxis	Increased sedation with midazolam Decreased metabolism of phenobarbital, phenytoin, rifampin Increases liver damage with hepatotoxic agents Immunosuppression with long-term use
St John's wort	Anxiety, depression, sleep problems, etc.	Cardiovascular collapse, delayed awakening due to effect on GABA neurotransmitter Severe interactions with antidepressants Induces CYP3A4 system leading to increased dose requirements of many drugs (including midazolam)
Gingko	Improves memory and concentration	Increased surgical bleeding due to antiplatelet effects. Concurrent use of NSAIDs not recommended Decreases effect of anticonvulsants
Garlic	Cardiac health	Increased surgical bleeding Augments ASA, warfarin, and heparin
Ginseng	General health	Hypoglycemic effects in diabetics Antiplatelet effects and bleeding Hypertension, headaches, and vomiting
Aloe	Skin problems	Allergic skin reactions. GI constipation bleeding, renal damage with oral use

Obtaining Consent

Proper informed legal consent must be gained before any anesthesia procedure. Parents also will benefit from an appropriate discussion of the risks of anesthesia in that this fulfills their own needs of responsibility and understanding. Parents should be informed that their child will be constantly and closely attended throughout the procedure. Breathing, heart rate, blood pressure, the concentrations of inhaled anesthetics, and oxygen levels will be constantly monitored. Their child will be kept comfortable and warm. Common side effects that should always be mentioned include sore throat, postoperative pain, nausea, and vomiting, together with the steps that will be taken to minimize these effects. Any specific risks associated with a particular procedure should also be detailed. The plan for postoperative analgesia should be outlined in detail and the role of the parents in this process defined; for outpatients a written analgesic protocol

should be provided and explained in detail. Finally, parents should be warned that even though their child may be healthy and may have no family history of anesthetic problems, there is always a very remote chance that more serious complications could occur. In the unlikely event that this happens the child will be immediately cared for “*to the very best of our ability*” and that they will be kept fully informed of any developments.

In the past, once the risks of anesthesia were explained to the parents or guardian, consent was assumed as a verbal understanding. More recently many institutions have adopted a separate written consent for anesthesia. This should list the risks that a reasonable person would wish to hear about before proceeding with anesthesia, as well as very rare risks of anesthesia such as cardiac arrest.

Of late, many parents have become aware of concerns with the “safety” of anesthesia in infants and young children (<3 years of age) in terms of neurocognitive development. We advise parents that the decision to proceed with surgery in young children is a balance of risks of not operating versus the benefits of the surgery and anesthesia. Most surgery in young children is necessary, and delaying it increases the risks of complications. In terms of anesthesia, evidence from newborn animals (e.g., rats, primates) indicates that most current anesthetics (see Chap. 8) can damage brain cells and their circuitry resulting in memory and learning difficulties that continue into adulthood. However, current prospective evidence in humans has not associated poor outcomes with general anesthetics. We advise parents that there is currently insufficient evidence to conclude that anesthetics harm infants and children. Instead we emphasize that these issues are still theoretic regarding humans, that if such a problem were a major issue that likely this would have been noted many years ago, and that it is far more important for us to maintain blood pressure, oxygen levels in the body and to provide adequate analgesia than to be overly concerned with an as yet unproven problem to infants. We also reassure them that research is underway to address this question. Such discussions should, of course, be outside the earshot of the young child.

Prophylaxis Against Hemorrhage for Infants

Ensure that the neonate undergoing surgery has been given Vitamin K₁ to prevent Vitamin K deficiency bleeding (VKDB), which is secondary to low levels of factors II, VII, IX, and X. Aqueous Vitamin K₁ (1 mg IM or IV) corrects the deficiency within a few hours and therefore should be given as early as possible. Late onset VKDB may occur in breast-fed infants who were never given Vitamin K or not receiving routine vitamin supplements; check the history. If intraoperative problematic bleeding is thought due to such an omission, transfusion of FFP is indicated.

Basic Laboratory Tests

Preoperative laboratory tests are considered unnecessary for most children; however, in some regions, these tests are legislated requirements. Preoperative urinalysis has not been found useful in detecting significant diseases or in routine screening of children. However, because a history is unreliable, urine (or blood) for pregnancy testing has become mandatory in many centers for all menarchal females. Hemoglobin (Hb) determination is likewise of little value in otherwise healthy children. Mild degrees of anemia have not been shown to increase the risk of anesthesia and do not alter the anesthesia technique selected. Most authorities now recommend that these tests may be omitted in healthy children undergoing minor surgery. Infants <6 months of age, especially those who were born preterm, should have a preoperative Hb determination to exclude anemia, which is more common in such infants and may increase the risk of complications. Older children with systemic diseases, those with a history of anemia, and those who may lose significant amounts of blood intraoperatively should also have a preoperative Hb determination. A sickle cell test is now performed routinely in neonates immediately after birth in many regions. Older infants and children whose sickle status is unknown should be tested with a Sickledex, and if that is positive, a hemoglobin electrophoresis should be performed.

Premedication

Drugs may be given preoperatively to produce sedation and tranquility, facilitate separation from parents if necessary, and smooth the induction of anesthesia, although distraction techniques are also very effective (see Chap. 1).

Topical Local Anesthetics

If it is planned to perform venipuncture or insert an IV prior to induction of anesthesia a topical anesthetic should be applied to the chosen site well in advance. For topical local anesthetics, see Appendix C, p. 634.

Vagal Blocking Drugs

Vagal blocking drugs (atropine, hyoscine, glycopyrrolate) are no longer routinely given to children preoperatively; however, glycopyrrolate in particular may be useful to decrease secretions for specific procedures such as bronchoscopy, to smooth induction in the child with a suspected difficult airway or to reduce secretions in neurologically impaired children.

Brisk vagal responses may occur during anesthesia in infants and children. Atropine should immediately be available for injection (preferably IV) should it become necessary. Serious bradycardia may lead to significant hypotension or more dangerous arrhythmias. This can result from instrumentation of the airway, manipulation of the eye, traction of the peritoneum, or administration of cholinergic drugs (i.e., succinylcholine). If bradycardia occurs, it must be treated promptly; ventilate with 100% oxygen, withdraw the precipitating stimulus, and give intravenous atropine.

Atropine is the preferred anticholinergic in children. It is more effective in blocking the cardiac vagus nerve and causes less drying of secretions than hyoscine or glycopyrrolate. Respiratory tract secretions, in fact, are not a serious problem with current inhalational anesthetics.

Children require larger doses of atropine than adults to achieve the same increase in heart rate. If indicated, atropine (0.02 mg/kg; maximum, 0.6 mg) may be given IV at induction. This is the preferred route of administration since it ensures effective drug action and spares the child a painful intramuscular injection and a preoperative dry mouth. Although some ascribe to a minimum dose of atropine of 0.1 mg IV, there is no minimum dose. If successful venipuncture is in doubt, the same dose of atropine may be given orally 90 min or IM 30 min preoperatively to ensure a peak effect at the time of induction. In an emergency, the usual dose of atropine diluted in 2 mL of saline is rapidly effective by the intratracheal route.

Infants with established bradycardia have a longer onset time for the chronotropic effect of intravenous atropine because of their reduced cardiac output. Therefore, if bradycardia is attributed to a vagal response, atropine should be given as early as possible. *An alternative cause for intraoperative bradycardia in infants or children is hypoxia, so the first treatment for any unexpected bradycardia is ventilation with 100% oxygen (especially if the pulse oximeter readings have been questionable). Symptomatic bradycardia that does not immediately respond to atropine must be treated with chest compressions and epinephrine (10 mcg/kg).*

Contraindications to the use of atropine are few in the pediatric age group; infants and children tolerate tachycardia much better than bradycardia. Children with heart disease who might tolerate tachycardia poorly (e.g., aortic stenosis or cardiomyopathy) require special attention; if bradycardia develops, small incremental doses of atropine should be given until the desired heart rate is reached. Studies have failed to confirm a reputed increased sensitivity of children with Down syndrome to atropine and thus they should be given the usual doses if indicated.

Table 4.5 Premedications and route of delivery

	Midazolam (mg/kg)	Ketamine (mg/kg)	Clonidine (µg/kg)	Dexmedetomidine (µg/kg)
Intravenous	0.05–0.15	1–2		
Intramuscular	0.1–0.2	3–10		
Oral	0.25–0.75	5–6	4	2.6
Nasal	0.1–0.2	2–4	4	1
Rectal	0.75–1.0	6–10		

See Chap. 3 for further discussion of medications

Sedatives and Tranquilizers

There is a voluminous literature and many widely divergent opinions concerning the use of sedative premedication for children (Table 4.5). Sedatives, opioids, or hypnotics may not ensure calm cooperation at the time of induction, but they may be associated with postoperative respiratory depression, slowed emergence, delirium, and vomiting.

Midazolam. Oral midazolam is the most widely prescribed sedative premedication for children. Midazolam is a water-soluble benzodiazepine with a shorter duration of action than diazepam. It may be given orally, nasally, rectally, or intravenously. For healthy infants and children up to 6 years of age, the oral route is preferred; a dose of 0.25–0.75 mg/kg produces sedation and tranquility in greater than 95 % of children within 10–15 min, after which time its effects start to wane. Older children (>6 years) require smaller mg/kg doses (0.3–0.4 mg/kg, maximum dose of 20 mg) than children less than 6 years of age. In infants, midazolam may be applied sublingually with a medicine dropper; this technique ensures rapid oral transmucosal absorption.

Oral midazolam produces anxiolysis with an approximate 50 % incidence of antegrade amnesia, does not significantly affect the volume or acidity of gastric fluid, but does improve cooperation on separation from parents and during induction. Recovery is not delayed after surgeries lasting 1 h, but after a brief procedure, early recovery may be delayed. Some children given midazolam are more restless during emergence after a brief procedure possibly because of paradoxical excitation.

Midazolam premedication does not reduce the incidence of emergence delirium after sevoflurane administration. Although it may reduce adverse behavioral outcomes after hospitalization, midazolam premedication may increase the incidence of bad dreams postoperatively for several days.

Intranasal midazolam (0.2 mg/kg) is effective particularly in the uncooperative child who will not swallow an oral premedication, although giving it by this route is unpleasant and usually makes the child cry. Additionally intrathecal

midazolam produces neurotoxic changes to the blood–brain barrier and the spinal cord in rabbits. Therefore, we do not recommend this route. Rectal midazolam (0.3–1 mg/kg) may be useful for small infants who cannot take the drug orally, but the onset of sedation is less predictable. Children with established intravenous access may be given midazolam 0.05–0.1 mg/kg IV immediately before arrival in the OR for a rapid calming effect. It should be noted that the peak brain effect after IV administration occurs in 3 to 5 minutes (i.e., which is a delayed onset compared with IV diazepam).

Lorazepam. Lorazepam is very useful for adolescents greater than 12 years of age. In an oral dose of 1–2 mg, it produces anxiolysis with a significant degree of amnesia. There are insufficient data to recommend its use in children less than 12 years of age.

Ketamine. Ketamine may be given orally in doses of up to 6 mg/kg but must be accompanied by oral atropine if excessive secretions are to be avoided. Postoperative vomiting may increase after oral ketamine. The combination of oral midazolam (0.3–0.5 mg/kg) and oral ketamine (3–5 mg/kg) produces very effective sedation for the more disturbed child. If this combination is used, the child should be closely observed as the drugs become effective. The combination should not be used when heavy sedation might be dangerous (e.g., in the child with a difficult airway or OSA).

Clonidine. Clonidine 4 µg/kg has been used as a premedication, but given orally requires a 90 min onset time, a much greater onset time than oral midazolam (10–15 min). It reduces anesthesia requirements and smoothes emergence, reduces agitation after sevoflurane, and enhances postoperative analgesia. In addition, clonidine diminishes cardiovascular responses to tracheal intubation and facilitates planned induced hypotension. Clonidine may cause unwanted postoperative sedation, which may be a disadvantage in the outpatient. Occasionally atropine may be required to correct bradycardia after clonidine.

Dexmedetomidine. Intranasal dexmedetomidine (1.0 µg/kg) has been reported to be effective as a premedicant in children when given 60 min before separation from parents; it also reduces agitation after sevoflurane. Neither pain nor discomfort was reported when intranasal dexmedetomidine was used, but in general, children do not like having medications administered intranasally and often cry in response. Unfortunately, it is not as effective when given by the buccal route. Neurotoxicity studies of dexmedetomidine have not been forthcoming; because it is unclear whether dexmedetomidine causes nerve damage, this unapproved route of administration should be viewed with caution.

Opioids

Opioids are rarely used as premedications in children unless pain is present. Opioids have traditionally been given intramuscularly, which children find unpleasant. Dizziness, nausea, and vomiting are common after their use.

Special Considerations

1. Neurosurgical patients who may have increased ICP should not receive any sedative premedication.
2. Children with a suspected difficult airway or OSA should be sedated with great caution due to the possibility of respiratory depression or airway obstruction.
3. Atropine should not be given intramuscularly to children with a fever because it may exacerbate the fever by abolishing sweating. If indicated, it may be given intravenously at the time of induction.
4. Some children undergoing correction of strabismus are assessed by the ophthalmologist immediately before the operation and should not be premedicated with atropine. They may receive atropine (0.02 mg/kg intravenously) at induction to block the oculocardiac reflex.

MANAGEMENT OF THE AIRWAY

Mask Anesthesia

1. During mask anesthesia, always have age and size appropriate equipment for tracheal intubation immediately at hand:
 - (a) A selection of suitably sized tracheal tubes in place (Table 4.6). If cuffed tubes are preferred, use ~0.5 mm ID smaller than the corresponding uncuffed tubes
 - (b) Two properly functioning laryngoscope handles with several suitable blades
 - (c) Labeled syringes containing atropine, succinylcholine, and induction agent (propofol, thiopental (not available in North America), ketamine, or etomidate)
2. Select a mask that fits the contours of the child's face and minimizes the dead space if possible. Disposable cushion-type facemasks have all but replaced Rendell Baker masks for infants and small children
3. The relatively large tongue in infants and adenoid or tonsillar hypertrophy in older children may cause upper airway obstruction. If obstruction occurs,

Table 4.6 Approximate diameter and insertion depth of uncuffed pediatric tracheal tubes

Age of child (year)	Internal diameter (ID) (mm) ^a	Approximate depth of insertion (cm) to mid-trachea ^b from:	
		Teeth	Nares
Premature	2.5 (for infants <1500 g) to 3.0 (for infants >1500 g)	6–9	8–11
Neonate	3.5	9–10	12
1	4.0	11	13
2	4.5	12	14
4	5.0	14	16
6	5.5	15	17
8	6.0	16	18
10	6.5	17	19
12	7.0	18	20
14	7.5	19	21
16	8.0	20	22

^aFormula: (age (\geq 2 years) (year) \div 4) + 4.0 = size of tube ID (mm); for cuffed tubes (age (year) \div 4) + 3.5 = size ID (mm). The tube diameters listed are given only as a guide. Always prepare a selection of tubes and use the one with the best fit (see text)

^bSee also p. 96

sublux the temporomandibular joint by applying digital pressure to the apex of the ascending rami (behind the pinna) directing the force towards the frontal hairline. This maneuver translocates the mandible anteriorly and rotates the joint, thereby lifting the tongue and intraoral tissues off the posterior pharyngeal wall and opens the airway and the mouth. If necessary insert an oropharyngeal or nasopharyngeal airway of suitable size. Gentle positive end expiratory pressure will also stent open pharyngeal soft tissue.

4. Infants have soft laryngeal cartilages and tracheal rings. Therefore, the anesthesiologist should avoid applying pressure on the neck over the airway during mask anesthesia. Monitor breath sounds, PetCO₂, and the movement of the reservoir bag continuously.

Laryngospasm

Laryngospasm occurs most commonly during induction of anesthesia, but also during emergence and occasionally during maintenance of anesthesia if the child is stimulated while lightly anesthetized. It occurs more commonly in infants (compared with older children), in preterm infants, children with recent URIs, children with reactive airways disease, after airway surgery, and in those exposed to secondhand smoke. It is more commonly associated with desflurane and isoflurane as compared with sevoflurane or halothane. It is rare during

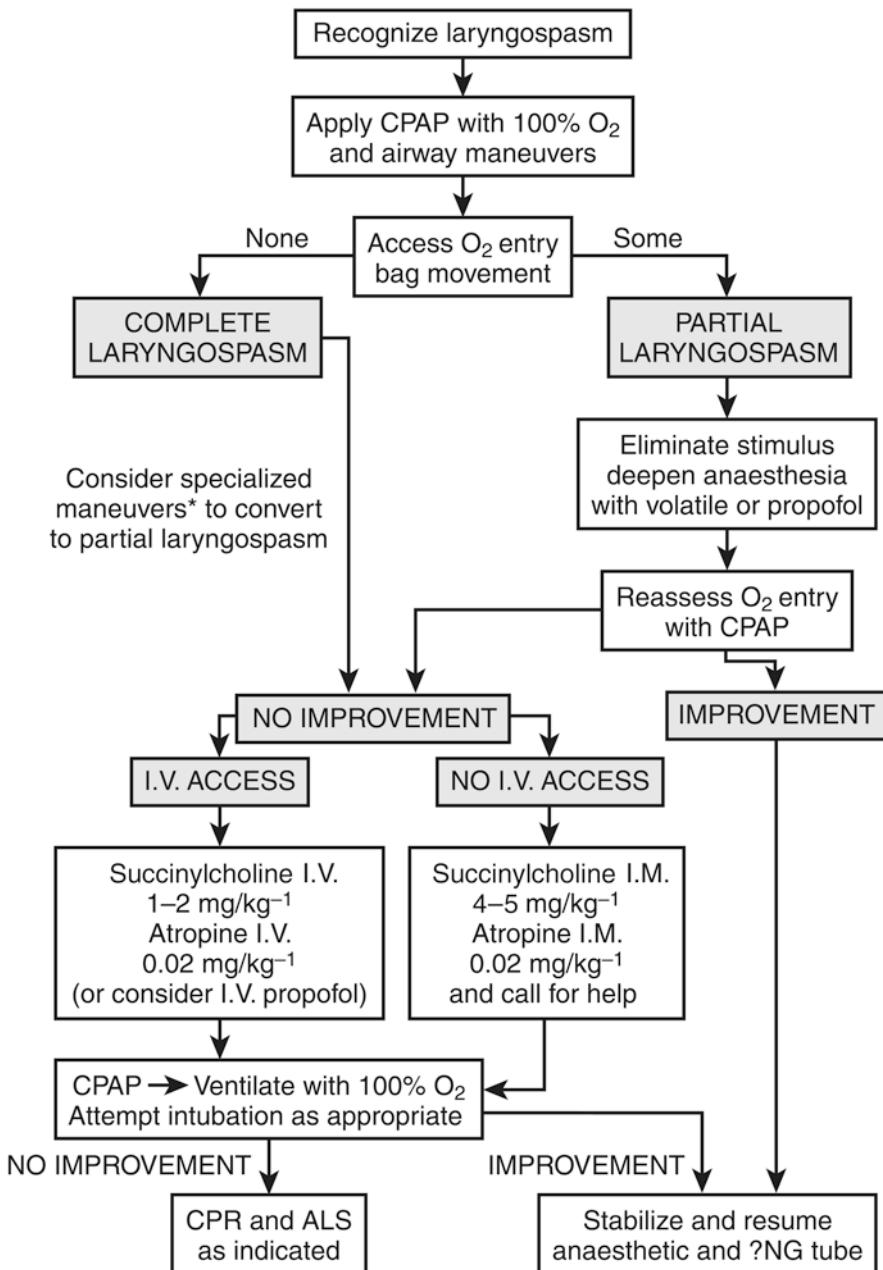
propofol-based anesthesia. The clinical hallmarks of laryngospasm include high-pitched sounds with inspiratory stridor that may progress to silence as the glottic aperture closes. Suprasternal and supraclavicular retractions occur with paradoxical chest wall motion and increased diaphragmatic excursions, and if the spasm persists, hemoglobin desaturation and bradycardia/asystole may ensue. As soon as laryngospasm is suspected, the precipitating agent (such as secretions, blood, or other foreign material) should be cleared. Initial treatment should include the application of a facemask to deliver 100 % oxygen with positive end-expiratory pressure (maximum pressure of 10–15 cm H₂O) (Fig. 4.1). If laryngospasm continues, the mandible should be displaced by transiently applying pressure to the superior end of the ascending ramus of the mandible (i.e., the condyles) using a single digit behind each pinna (see previous discussion). While subluxing the mandible, the thumbs should seal the facemask to the face to maintain positive pressure. Oral airways must be used with great caution in these circumstances as short oral airways may push the tongue into the glottic opening and long airways may push the epiglottis into the opening, in both instances obstructing rather than opening the airway. An additional benefit from pressing on the ascending ramus of the mandible is that the stimulation incurred increases respiratory effort. However, if these measures fail and the oxygen saturation and heart rate continue to decrease, and the child is in extremis (i.e., bradycardic and hypoxic), early treatment should include IV atropine (0.02 mg/kg) followed by IV propofol (1–2 mg/kg) and/or succinylcholine (1–2 mg/kg IV or 3–4 mg/kg IM) and tracheal intubation.

Postobstructive Pulmonary Edema

Postobstructive pulmonary edema is a complication that may occur after relief of acute (laryngospasm) or chronic upper airway obstruction (tonsillectomy). The mechanism appears to be the generation of extreme negative intrathoracic pressure against a closed glottis and its sudden release causing a dramatic increase in pulmonary blood flow resulting in low pressure, noncardiogenic pulmonary edema. This complication should be suspected when pink frothy sputum appears in the tracheal tube and oxygen desaturation continues unabated. Treatment is to continue positive pressure ventilation and administer IV furosemide (Lasix) as needed.

Postintubation Croup

Postintubation croup or subglottic edema is associated with traumatic intubation, tight-fitting endotracheal tubes, coughing on the tube, repositioning, prolonged intubation, surgery of the head and neck, and a history of croup.



*Specialized maneuvers

1. Pressure in laryngospasm notch
2. Pull mandible forward

Fig. 4.1 Laryngospasm algorithm (Modified from Hampson-Evans D, Morgan P, Farrar M: Pediatric laryngospasm. *Pediatr Anaesth* 18:303–307, 2008)

Treatment includes the inhalation of cool mist, steroids, and racemic epinephrine. Inhalation of racemic epinephrine (0.5 mL in 3 mL normal saline over 10 min) is usually effective although its effects are temporary and its use may be followed by rebound edema. Prolonged observation or overnight admission may be required.

Laryngoscopy

1. Ensure that the head is correctly positioned and supported.
 - (a) For infants and children younger than 6 years of age, the head should be on the level of the table and supported in a low head ring. At this age, the larynx is positioned high in the neck and there is no advantage to anterior displacement of the cervical spine because the relatively large head accomplishes this naturally. Extension of the neck at the atlanto-occipital joint aligns the oral, pharyngeal, and tracheal axis for optimal visualization. External pressure (the so-called optimal external laryngeal manipulation [OELM]) applied to the thyroid cartilage region of the anterior aspect of the neck (applied most effectively by the anesthesiologist using their second hand, in a posterior and cephalad direction) may further improve the view of the glottic aperture.
 - (b) For older children and adolescents, place a folded sheet or blanket under the head as a pillow to cause anterior displacement of the cervical spine and improve the alignment of the airway and the view of the glottis as in adults.
 - (c) Beware of conditions that may be associated with an unstable cervical spine (e.g., Down syndrome, Marfan syndrome). In such cases, note the range of spontaneous motion of the neck when the child is awake and inquire about the presence of neurologic symptoms preoperatively (wide-based gait, urinary, or fecal/urinary incontinence). There is no evidence that cervical spine films are helpful in ruling out unstable cervical vertebra in these children and X-rays of the cervical spine are not recommended preoperatively; however, we advise extreme care be taken to limit neck movement during tracheal intubation or surgical positioning.
2. Examine the teeth carefully; many young children have loose deciduous teeth. The teeth must be kept in view throughout laryngoscopy: retract the lip with your thumb and exert no pressure on the teeth during laryngoscopy. If loose teeth are present, it should be noted preoperatively and the parents should be informed that it may be safer to remove them once the child is anesthetized. If teeth are removed, they should be retained and given to the

parents after the anesthetic. A plastic tooth guard may be applied to the maxillary teeth and is particularly useful for older children and adolescents when a Macintosh (curved) blade is used.

3. In infants and children, the anesthesiologist's view of the glottis may be obscured by the epiglottis unless it is raised with the tip of the straight blade. (This is why we advocate the use of a straight blade in this age group.) In small infants, it is sometimes a problem to lift the epiglottis without it slipping off the laryngoscope blade. If this happens, advance the blade into the hypopharynx (with the entire tongue to the left of the blade) and slowly withdraw it until the glottis slips off the blade and the base of the epiglottis is firmly held. Alternatively, a straight blade may be used as would a curved blade and slowly advanced along the surface of the tongue. As the tip of the blade enters the vallecula, lift the base of the tongue to avoid trauma to other laryngeal structures.
4. Insufflation of oxygen into the pharynx during laryngoscopy (especially in infants or those with a difficult airway) improves oxygenation during attempts at intubation. Specially designed blades with an oxygen port are available (oxyscope, Anesthesia Medical Specialties, Santa Fe Springs, CA), or an oxygen catheter may be taped to the side of any blade to deliver approximately 2 L/min of oxygen.

Tracheal Intubation

1. The optimal size of the tube is the largest one that passes easily through the glottis and subglottic regions without incurring resistance. The presence of a leak around an uncuffed tube at 20 cm H₂O positive pressure depends on a number of factors (including head position and muscle relaxation). A leak should be tested and achieved between 20 and 30 cm H₂O peak inflation pressure. If the leak is too large, the next half size larger tube may be inserted. If the tube will not pass the cricoid ring or if it does and there is no leak at 30 cm H₂O, then a smaller diameter tube should be considered. In deciding that the tube size is appropriate, many clinicians depend more on the absence of any resistance when the tube passes through the cricoid region than on the presence of an audible leak. There is increasing use of cuffed tracheal tubes in children in the OR and PICU; these tubes should also have a leak when the cuff is deflated (see later discussion).
2. Clear, thin-walled polyvinyl chloride (PVC) tubes (Z79-approved) are preferred.
3. Cuffed tubes are preferred for major surgery, those with reduced lung compliance and those who may be at increased risk for regurgitation of gastric contents. Even in infants and young children, the use of an appropriately

sized cuffed tracheal tube does not increase postoperative morbidity. The use of a cuffed tube reduces the need to change tubes, reduces OR pollution, and may reduce the risk of aspiration. It also obviates the need to have a close fit in the subglottic region to ensure effective ventilation. For this reason it may be preferable, in children with a known tendency to subglottic stenosis (e.g., Down syndrome), to use a tube of smaller diameter together with a cuff to seal the airway. Small-sized cuffed tubes are now available with a wall thickness similar to that of uncuffed tubes. In infants and small children when a cuffed tube is used, the cuff may not require inflation to achieve an adequate seal for ventilation.

When using a cuffed tube, positioning the cuff below the cords places the tip of the tube much closer to the carina in small infants and may increase the risk of an endobronchial intubation. The MicroCuff¹ tube has a high compliance, elliptically shaped cuff that is more distally attached to the tube and may be advantageous: note that these tubes do not have Murphy eyes so right upper lobe ventilation may be compromised.

4. We recommend that tracheal tube connectors be firmly inserted. This is readily accomplished (particularly in small diameter tubes) by removing the connector, moistening with an alcohol swab, and reinsertion into the tube up to the shoulder.
5. The correct positioning of the tracheal tube should be immediately confirmed by observation of the chest excursion, the presence of humidity within the lumen of the tube, the capnograph trace, and chest auscultation. Ventilation should be auscultated in both axillae. The length of the trachea of the infant and young child is short, only 5 cm in the neonate. The tip must be accurately placed at mid-tracheal level to minimize the risk of an endobronchial intubation or accidental extubation. Note carefully the length of tube that is passed through the cords and check the tube length marking at the teeth. An easy method of determining the correct depth of insertion of the tracheal tube when it is even with the teeth or alveolar ridge is 10 cm in a neonate, 11 cm for a 1 year old, and 12 cm for a 2 year old (N.B. The second digit in this number is the child's age in years.); beyond 2 years, halve the child's age and add to 12 cm (see Table 4.6). Tubes must be firmly taped in place, preferably near the middle of the mouth, where they are less likely to kink. Alternatively, the tracheal tube may be taped in the corner of the mouth on the up side of the face so that secretions do not loosen the tape. When cuffed tubes are used, the cuff should be positioned just past the vocal cords; check for bilateral ventilation and secure the tube carefully. As previously mentioned, the margin of safety before the tip passes into the bronchus is shorter with cuffed tubes.

¹ Halyard Microcuff, www.halyardhealth.com

6. Avoid pressure by anesthetic hoses and other equipment on the child's face or head by the use of suitable padding. Facial nerve palsy has been reported after anesthesia because of pressure of anesthesia equipment.
7. The anesthesia circuit and tube must be carefully positioned and supported to prevent any traction on the tube that might cause it to kink or become dislodged. *Tracheal tubes used in children kink very easily, and this is a potential cause of disastrous accidents.*
8. Remember that extension of the neck withdraws the tip of the tube proximally in the trachea and flexion advances the tube; 1–3 cm of movement may occur between full flexion and full extension in infants. Position the tube carefully and consider the effects of changing the head position. Always reassess bilateral ventilation after repositioning the child.
9. Be aware that some neonates and infants have a congenitally short trachea, which increases the potential for endobronchial intubation (e.g., DiGeorge syndrome).

Nasotracheal Intubation

If the nasal route is chosen, the child should be prepared as outlined earlier. When performing nasotracheal intubation it is not unusual to cause some bleeding from the nose. As a result, several strategies have been developed to reduce this risk. Although warming the tip of the tube softens it, this has not been shown to be effective in reducing nose bleeding. Two effective strategies are application of a vasoconstrictor (e.g., oxymetazoline) to the nares and/or telescoping the tip of the tube into the flange end of a smooth red rubber catheter. With the latter, the catheter tip should be lubricated and passed along the floor of the (preferably left) nostril followed by the tube. (The left nostril aligns the tip of the tube [bevel on the left] directly with the glottic opening and aligns the bevel of the tube on the turbinate side.) The catheter is then advanced into the oropharynx where it is visualized in the hypopharynx and pulled off the tracheal tube with McGill forceps. The tube is then directed using McGill forceps into the trachea.

A Note on Awake Intubation in the Neonate. It was common practice to intubate the neonate awake; however, it is now usual to induce anesthesia first. Intubation is more likely to be successful and less desaturation to occur if the neonate is first anesthetized. Awake intubation might be considered in neonates who are hemodynamically unstable, have micrognathia or a difficult airway, or who are extremely premature. To perform an awake intubation, pretreat the neonate with oxygen and atropine, use a styletted tracheal tube, and have an assistant position the neonate's arms fully abducted against the head to stabilize

the neonate's head. With the laryngoscope on one hand and the tube on the second, remove the facemask and intubate the trachea rapidly. Awake intubation increases blood pressure and intracranial pressure to a similar extent as crying and coughing. There has been a concern raised regarding an increased risk of IVH in preterm infants whose tracheas were intubated awake; however, this risk has never been confirmed. If awake intubation is deemed to be necessary, a local anesthetic solution (diluted, with upper limits predetermined) may be applied to the oropharynx and palate to reduce the infant's distress and struggling. Small doses of fentanyl (0.5–1 µg/kg) and midazolam (0.05–0.1 mg/kg) may also facilitate the process. The use of a laryngeal mask airway (LMA) to establish the airway before anesthesia may also be considered (see later discussion).

Rapid Sequence Induction

Rapid sequence induction (RSI) is an intravenous induction technique used to rapidly and safely secure the airway with a tracheal tube when there is a risk of vomiting or regurgitation and aspiration. Careful preparation for an RSI is essential and requires the following:

1. Equipment: anesthesia machine, age appropriate laryngoscopes and tracheal tubes, functioning suction.
2. Predetermined drugs in weight-appropriate doses.
3. Skilled anesthesiologist and skilled assistant.

The equipment must include all standard monitors and the necessary airway instruments and supplies. The airway equipment should include two functioning laryngoscope blades and handles, two sets of suction tubing, cuffed tracheal tubes of age appropriate size (plus a half-size larger and smaller diameters), and a stylet. Prepared drugs include 100% oxygen to be given before the sequence begins, an intravenous induction agent (propofol 3–4 mg/kg, ketamine 1–2 mg/kg, or etomidate 0.2–0.3 mg/kg) in predetermined doses according to weight and physical status (N.B. Severity of dehydration, acidosis, or sepsis), atropine (0.02 mg/kg), and a muscle relaxant (succinylcholine 2 mg/kg or rocuronium 1.2 mg/kg). The child's head is optimally positioned during preoxygenation and as anesthesia is induced, cricoid pressure (see below) is applied by the assistant. (Before commencing, ensure that your assistant is instructed in the effective application of cricoid pressure.) Once paralyzed, the trachea is intubated without delay and the cuff inflated. In the modified RSI technique, the lungs may be ventilated manually with 100% oxygen while maintaining cricoid pressure (see later discussion).

Table 4.7 Contraindications to cricoid pressure

Contraindication	Potential complication
Active vomiting	Rupture of the esophagus
Airway issues	Fractured cricoid cartilage ⇒ may be worsened Sharp foreign body in the airway may perforate airway
Esophageal issues	Regurgitation from a Zenker diverticulum Sharp foreign body may perforate esophagus, larynx, aorta, or spinal canal
Vertebral/neurologic issues	Unstable C-spine may cause disruption of spinal cord Sharp foreign body may perforate spinal cord
Education	Lack of knowledge, expertise, and/or ability to properly apply cricoid pressure

Cricoid Pressure

Cricoid pressure (Sellick maneuver) should be applied to the cricoid ring with thumb and long finger as anesthesia is induced and should be maintained until tracheal position of the tube is confirmed. While we recommend the use of cricoid pressure, we have some reservation about its use in infants and children. Cricoid pressure has been popularized as an adjunct to the RSI to prevent passive regurgitation of gastric contents during induction of anesthesia. The use of this technique is widely advocated. However, evidence to support the effectiveness of cricoid pressure to either occlude the lumen of the esophagus or prevent passive regurgitation of gastric contents has not been forthcoming. MRI evidence indicates that the lumen of the esophagus is lateral to the cricoid ring in 50% of patients and is laterally displaced with cricoid pressure in 90%. Moreover, studies of the force required to occlude the lumen in adults report that inadequate force is applied in the majority of patients. Surveys indicated that cricoid pressure is used in only half of those children in whom it is indicated. In adults, 30–40 N (3–4 kg of force) must be applied to the esophagus; there are no comparable data in infants and children. However, as little as 5–10 N force distorts the cricoid ring in infants and children. Cricoid pressure may occlude the upper esophageal sphincter but it relaxes the lower esophageal sphincter. If mask ventilation is ever necessary in a child with a full stomach, it is probably prudent to continue to apply cricoid pressure to prevent gastric inflation and maintain small peak inflation pressures. Contraindications to cricoid pressure should be recognized (Table 4.7). Excessive cricoid pressure may increase the difficulty of laryngeal visualization; OELM combined with cricoid pressure may be needed.

The Laryngeal Mask Airway

The LMA Classic was originally designed as a substitute for mask anesthesia in spontaneously ventilating adults. Pediatric models were developed which were scaled down versions of the adult model, i.e., they were not specifically redesigned to suit the anatomical features of infants and children. LMAs are sized according to the child's weight (Table 4.8). If optimally placed, the LMA tip lies against the upper esophageal sphincter, the sides against the pyriform fossa, and the upper border against the base of the tongue. Thus it encircles the glottic opening. When the LMA is in position, studies show that the tip of the tube lies in the hypopharynx, although the relationship of the cuff to the epiglottis and laryngeal aperture may vary somewhat despite an apparently good airway. Successful initial insertion with a clear airway is more likely with larger-sized LMAs in older patients. Correctly positioned, the LMA causes less resistance to inspiration than the tracheal tube in spontaneously breathing children. The use of the LMA has been extended to some children whose lungs are ventilated; in children, controlled ventilation without distention of the stomach may be possible if peak airway pressures are maintained less than ~ 15 cm H₂O. The LMA does not protect the airway should vomiting or regurgitation occur.

The ProSeal LMA provides a better seal around the glottis (average leak at ~ 25 cm H₂O peak inflation pressure) and an esophageal conduit as a means for venting the stomach; this device is easier to insert successfully in small children.

Insertion of the LMA is performed after induction of adequate anesthesia by inhalation or by an intravenous injection of propofol (3–4 mg/kg for children). The cuff should be checked before it is inserted and it needs to be well lubricated. Several methods of insertion have been suggested:

- (a) After the cuff has been completely deflated and the mask well lubricated, it is inserted blindly along the curve of the palate into the pharynx, with the aperture positioned anteriorly, until resistance is felt. If the LMA becomes "hung up" on the posterior pharyngeal wall, a finger should be inserted into the mouth to lift the tip of the LMA and continue insertion. The LMA is then advanced using gentle pressure. When the cuff is inflated, the LMA should rise slightly out of the mouth. The adequacy of ventilation and the fit of the LMA are checked. Generally, a leak will become obvious at approximately 15 cm H₂O peak inflation pressure.
- (b) A second method is performed with the cuff partly inflated; the LMA is inserted with the mask facing posteriorly and then rotated 180° as it enters the pharynx. It may also be inserted with the cuff partially inflated but in the recommended position facing anteriorly.

Table 4.8 Guide to selection of LMA, endotracheal tube, and fiberoptic scope size

LMA size	Child's weight (kg)	Maximum cuff volume (mL)	Largest ETT (ID, mm) inside classic LMA	Largest ETT (ID, mm) inside Unique™ LMA ^a	Largest tracheal tube (ID, mm) ProSeal™ LMA ^b	Largest FOB inside ETT (mm)
1	<6.5	4	3.5	3.5	N/A	2.7
1.5	5–10	7	4.0	4.0	4.0, some manufacturers	3.5
2	10–20	10	4.5	4.5	3.5	4
2.5	20–30	14	5.0	5	4.0	5
3	30–50	20	6.0, cuffed	5.0, cuffed	4.5	5
4	50–70	30	6.0, cuffed	5.5, cuffed	5.0	
5	70–100	40	7.0, cuffed	6.0, cuffed	5.0, cuffed	
					6.0, cuffed	

^aNote that these sizes differ from the manufacturer's recommendations, but have been found by us to be the better alternatives to ensure easy passage of the tracheal tube through the LMA. In some cases, the tubes listed are smaller; in some cases a cuffed alternative is given as an option rather than an uncuffed tube

^bOlympus Surgical Inc., Orangeburg, NY

Fiberoptic bronchoscopes:

Olympus LF-P (outside diameter 2.2 mm): will pass through ETT 3-mm ID or larger. Will pass through 26 fr double lumen tube (no suction channel)

Olympus LF-2: will pass through 5-mm ID ETTs or larger (1.5 mm suction channel)

- (c) A third method is to insert the mask, partly inflated vertically into the mouth, and rotate it 90° as it is passed into place.
- (d) The ProSeal™ LMA may be “railroaded” into correct position over a suitable catheter advanced into the esophagus via the conduit in the mask. High success rates are claimed with this method.

In either case, a black guideline on the proximal visible tube should lie against the central incisors when the LMA is in place and correctly oriented. Coughing and laryngospasm may occur if the LMA is inserted at too light a level of anesthesia; such complications may be more common in infants and small children than in adults.

Some difficulty with insertion may be encountered in up to 25 % of children; this is more likely in the smaller child having a size 1 or size 1.5 LMA inserted. In a very small percentage of children, it may be impossible to place the LMA correctly.

When correctly positioned, the LMA provides a good airway with less resistance than a tracheal tube. It avoids instrumentation through the glottis and frees the anesthesiologist from holding a facemask. It is particularly useful during imaging procedures, radiotherapy, and for other short procedures where mask anesthesia with spontaneous ventilation might be used. Somewhat surprisingly, some have found the LMA to be useful during adenotonsillectomy, and it has been suggested that less aspiration of blood occurs with the LMA than with an uncuffed endotracheal tube (We, and most ENT surgeons, prefer tracheal intubation). A special application of the LMA is to use it as a guide to assist management of the child with a difficult airway, where it may be used as a prelude to fiberoptic endoscopy and intubation. The LMA can be inserted in awake infants (e.g., with Pierre Robin sequence) after inhalation of nebulized lidocaine and used to provide an airway to induce and deepen anesthesia before fiberoptic intubation. The LMA is useful when ventilation proves to be difficult in a child during multiple attempts at intubation. The LMA may also be advantageous in infants with tracheal stenosis, in whom passage of a tracheal tube would severely reduce an already compromised airway diameter. It must be remembered that the LMA does not guarantee the airway as does a tracheal tube, and it does not protect against aspiration.

The pressure in the cuff of the LMA should be measured; some models have a useful built in gauge for this purpose. Excessively high pressures (>60 cm/H₂O) may increase the leak and are a cause of postoperative sore throat. If the LMA is inserted with the cuff inflated, it may require some deflation to avoid excessive pressure when positioned.

At the end of the procedure, the LMA may be left in place until protective reflexes have returned, or it may be removed while the child is still deeply anesthetized. Removal while the child is anesthetized usually results in fewer airway

complications and less desaturation, but a facemask should be applied until the child is able to maintain a safe airway. If desflurane has been used, it may be safer to remove the LMA with the child awake. The incidence of postoperative sore throat is similar whether an LMA or a tracheal tube is used.

A number of other supraglottic devices are available but few have not been subject to the extensive evaluation of the LMA. Practitioners should decide which device is safest in their hands.

Difficult Tracheal Intubation

Preoperative Assessment

It is most important that the anesthesiologist carefully assess the airway before administering anesthesia to determine the likelihood of obstruction during induction and to judge the likely ease of tracheal intubation.

Beware of any child who does not look quite normal or who has any syndrome or association of defects. Always anticipate the possibility of an abnormal airway. When there is any doubt, assume the airway will be difficult and be prepared. Review the history and examine the child carefully. Always review any previous anesthesia records—but do not be lulled into a false sense of security by a previous easy airway and uneventful anesthetic. Not all anesthetic difficulties are detailed in the record or child's chart, but even if they were, the ease of intubation may change as the child grows. In some cases, intubation may become easier, as in the child with a cleft palate and Pierre Robin sequence. In others, it becomes more difficult, as in the child with Treacher Collins, Goldenhar, and Klippel–Feil syndromes.

The examination of the child may provide clues as to the likely ease of intubation:

1. Assess the extent of mouth opening.
2. Check the extent of neck flexion and extension.
3. Check the shape and size of the mandible and maxilla (profile view).
4. Examine the mouth, tongue, and palate.
5. External ear deformities are often associated with mandibular hypoplasia.
6. Assess the distance between the ramus of the mandible and the thyroid cartilage.

Limited mouth opening, restricted neck extension, a large tongue, or a short ramus of the mandible predicts difficulty with laryngoscopy and tracheal intubation. Inability to fully visualize the fauces and uvula suggests difficult intubation, although the Mallampati scoring system does not predict a difficult laryngoscopy. Successful laryngoscopy depends on the ability to displace the soft

tissues of the oropharynx into the mandibular space. Any deformity that limits this space (short or shallow mandible) or increases oropharyngeal tissue (large tongue) can be expected to compromise efforts to see the glottis.

Management Strategies

It is most important to be prepared for every option when facing a difficult airway. Confirm that all the age- and size-appropriate equipment you may require is readily available. It is essential to keep all the “difficult pediatric airway” supplies on a special cart that is located centrally and can be wheeled into any room where it is required. It is always advantageous to have expert assistance on hand and this second pair of hands is part of the ASA “known difficult airway” algorithm. If there are other members of the department with special skills, do not hesitate to enlist their aid, even if their initial role is simply to stand by, provide moral support, and be ready to intervene.

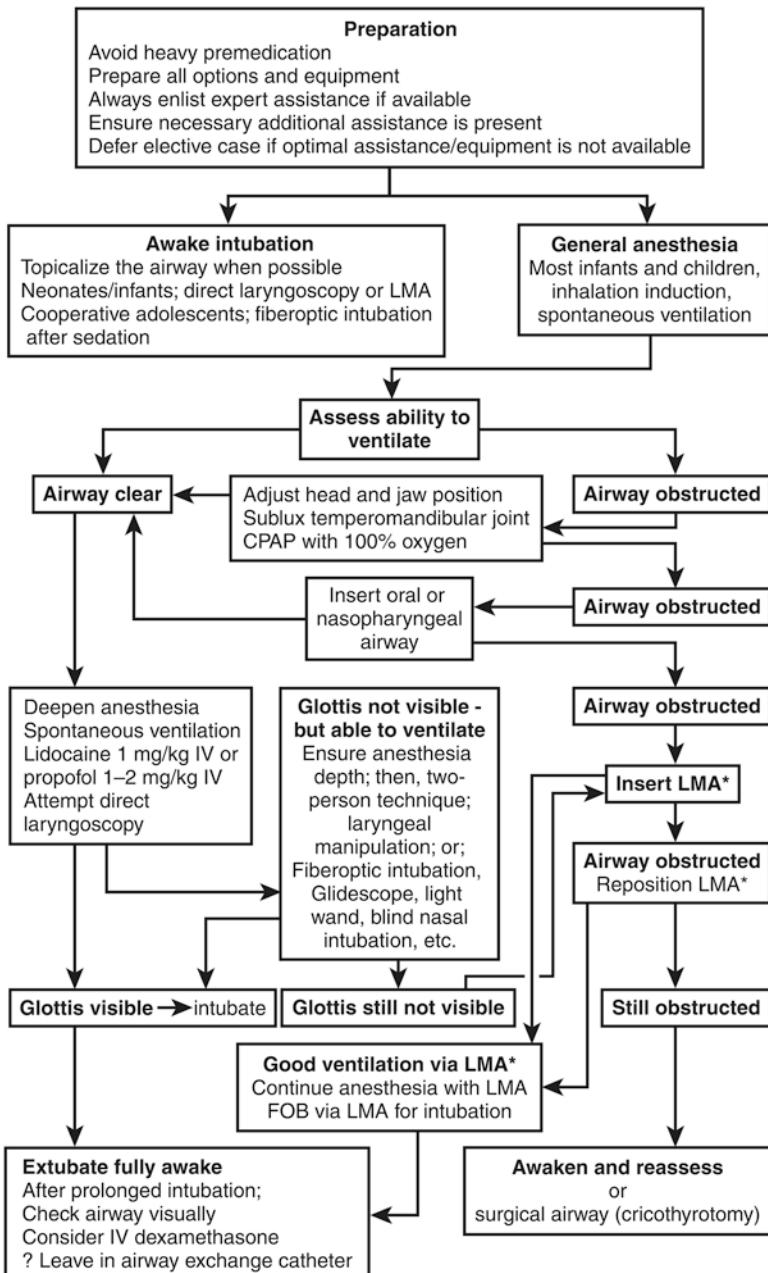
Preoperative administration of an anticholinergic drug may be advantageous to decrease secretions in the mouth and pharynx and minimize the possibility of laryngeal spasm. No heavy sedation should be administered; small doses of anxiolytics might be administered when necessary, using suitable caution and appropriate monitoring. Children who cannot be fasted and require emergency surgery may be prepared with the use of histamine₂-blocking drugs and intravenous metoclopramide.

A recommended sequence to follow is outlined in the Pediatric Difficult Airway Algorithm (Fig. 4.2).

The choice of anesthetized versus awake/sedated intubation is quite simple: children, unlike adults, almost always require general anesthesia. Children are very easily upset and will not cooperate during attempts at an awake/sedated intubation (even after local anesthesia is applied to the airway). Small infants may be severely stressed by attempts at awake intubation and their airways are more easily and rapidly intubated when they are anesthetized. An exception is advised for certain small infants (i.e., those with Pierre Robin sequence), in whom maintaining a patent airway and tracheal intubation may be very difficult under anesthesia and who may be more safely managed by topical anesthesia of the mouth and insertion of a well-lubricated LMA while the infant is awake. The LMA can then be used as a route to induce anesthesia and, if necessary, as a conduit for fiberoptic intubation (see Table 4.8).

Standard Management

The classic traditional approach to the management of the difficult pediatric airway is by inhalational induction, deep inhalational anesthesia, continued spontaneous ventilation, and direct laryngoscopy. This method is advantageous in that it does not require complex equipment and one can immediately determine the status of the airway and the degree of difficulty of direct laryngoscopy.



* Consider ProSeal™ LMA as it allows greater inflation pressures and the ability to decompress the stomach

Fig. 4.2 Difficult airway algorithm

These details, along with the type of laryngoscope blade used and other data, can then be clearly recorded in the anesthesia record. *Although this procedure is still recommended as a standard basic management plan, some children may be more effectively managed by early insertion of the LMA or other techniques (see Alternative Techniques below).*

Induction of anesthesia should be performed by inhalation; intravenous relaxant drugs are generally contraindicated. Sevoflurane is preferred for a smooth, rapid induction. If halothane is available, its greater solubility facilitates a more prolonged laryngoscopy than is possible with sevoflurane. As anesthesia is induced, the muscles of the tongue and pharynx relax. At this time, obstruction may occur, and immediate measures may be required to reestablish a clear airway.

1. Adjust the position of the head with increased jaw thrust to lift the tongue off the posterior pharyngeal wall and open the upper airway. A two-handed method is preferred, with one digit behind the most cephalad tip of the ascending ramus on each side of the mandible. The finger is wedged in the triangle formed at the base of the skull between the ascending ramus of the mandible anteriorly and the mastoid process posteriorly, immediately behind the tragus. Pull the mandible towards the frontal hairline. This maneuver subluxes the mandible anteriorly and rotates the temporomandibular joint, thereby opening the mouth. At the same time, two thumbs hold the mask on the face. With this maneuver, an oropharyngeal airway is often unnecessary.
2. If necessary, insert an oropharyngeal airway (see previous discussion), but be aware that if the child is too lightly anesthetized this may result in coughing and laryngospasm. Make sure that the airway is appropriately sized for the child; measure it against the outside of the face (the tip should extend just to the angle of the mandible). Initiate ~10 cm H₂O PEEP to distend pharyngeal soft tissues.

When an airway is established, anesthesia is deepened with 8 % sevoflurane in oxygen. Before the first attempts at laryngoscopy and intubation, 1.5 mg/kg of intravenous lidocaine and/or 1–3 mg/kg propofol are administered slowly to reduce the likelihood of breath holding or coughing during instrumentation. During laryngoscopy, oxygen may be insufflated into the pharynx via a catheter or by the use of a special laryngoscope blade. If an adequate view of the glottis is obtained, intubation can be performed; if not, other manipulations are required:

- (a) With the laryngoscope in place, apply posterior and cephalad pressure on the cricoid region of the neck to bring the larynx into view (OELM). In children with severe retrognathia, consider pushing the larynx to the right to visualize the glottis.

- (b) In some cases, a “two-person approach” to intubation is preferable. One person who holds the laryngoscope with one hand also applies pressure (as described previous discussion) to align the axes of the larynx and oropharynx. Once the glottis is visualized, the laryngoscopist tilts his head to the left to enable the second person to pass the tube through the cords.
- (c) A retromolar or paraglossal approach may be indicated. Insert a straight blade at the extreme right side of the mouth behind the bicuspid or last molar tooth while rotating the head to the left and retracting the corner of the mouth with a small surgical retractor. The epiglottis is visualized and the tracheal tube advanced off a stylet.
- (d) An alternative approach after induction of anesthesia and after confirming the ability to ventilate the child with bag and mask is to administer succinylcholine. Laryngoscopy can then be attempted during apnea with complete muscular relaxation. This may facilitate laryngoscopy and tracheal intubation, but at the same time, it limits the time available for each intubation attempt. Infants and small children desaturate more rapidly during apnea than do older children or adults. (N.B. Administration of a relaxant drug might result in a “can’t intubate/can’t ventilate” situation with an apneic child.)

If laryngoscopy proves impossible by direct vision, the mask should be reapplied, deep anesthesia continued, and other options considered (see Fig. 4.2). It is wise not to persist with prolonged attempts at direct laryngoscopy because these may become traumatic and result in bleeding, thus compromising the chances of success with other methods or by other individuals.

Alternative Techniques

The **GlideScope** (Verathon Inc., Bothell, WA, 98011). This device incorporates a small video camera to observe the glottis on a monitor screen. The “GlideScope AVL Preterm/Small Child” is recommended for preterm infants and small children whereas the “GVL” for larger children. The use of the GlideScope may allow intubation using a styletted tube. Be cautious not to injure the palate while passing this through the mouth. Since the introduction of the GlideScope, a host of other videolaryngoscopes have been introduced and studied in children including the Airtraq (Prodol Meditec, S.A., Vizcaya, Spain), Storz VL and C-MAC S Video Laryngoscope (Karl Storz GbmH, Tuttingen, Germany), and TruView EVO2 Infant (Truphatek International Ltd, Netanya, Israel). The reader should use the device with which he/she is most expert.

Laryngeal Mask Airway. The introduction of the LMA has made possible many new approaches to manage the difficult airway, provided the mouth and pharynx are of adequate size. If intubation under direct vision is impossible, the

LMA may be inserted without delay. Insertion of an LMA at any stage is usually successful in establishing a patent upper airway, but laryngeal spasm may occur if the child is inadequately anesthetized. Once in place, the LMA can be used as a route for ventilation and oxygenation, to continue anesthesia, and as a conduit for flexible bronchoscopes, endotracheal tubes, airway catheters, light wands, or other equipment needed to complete intubation (see Table 4.8).

Flexible Bronchoscope. In some children it is impossible to insert an LMA (i.e., very small or scarred mouth) and it may be necessary to perform flexible bronchoscopy via the mouth or nose. The smallest scopes do not have a suction channel (see Table 4.8). A suction channel on the scope is a very desirable feature because secretions frequently obscure the view and this channel may also be used to administer oxygen in children who are breathing spontaneously; external suction may then be required to clear secretions.

If the nasal route is chosen, anesthesia should be cautiously induced with sevoflurane while maintaining spontaneous ventilation as previously described. The flexible bronchoscope should be prepared in advance by sliding an appropriate size tracheal tube (without the connector) over the scope. When the child is adequately anesthetized, the flexible bronchoscope is passed through the left nostril, to the glottis and into the trachea. The tube should then be passed over the scope and into the trachea. If the tube does not pass the cords, the tube should be rotated 90° so that the tip of the tube is midline. Practicing flexible fiberoptic bronchoscopy in children with normal airways is essential if the anesthesiologist is to become adept at a similar intubation technique with a difficult airway. When a very small endoscope is not available, alternative methods for children have been suggested including visualizing the glottis with a larger scope to pass an airway exchange catheter into the trachea under direct vision, over which the tracheal tube can subsequently be threaded.

Light Wand Intubation. The use of a malleable lighted stylet, passed blindly into the trachea, makes it possible, with practice, to rapidly secure the airway. The stylet should be curved to suit the predicted shape of the child's airway, and a suitable tracheal tube is mounted on it. Always check that the lamp is screwed firmly in place. As the light passes into the trachea, the anterior neck can be seen to transilluminate. This is more easily seen with the room lights dimmed. The tracheal tube can then be advanced over the stylet into the trachea. The method requires practice but may be successful in many children with a difficult airway. It is limited by the size of tube that can be passed over the stylet; the Trachlight (Laerdal Medical Corp., Wappingers Falls, NY)² has two sizes: child accommodating tubes 4–6 mm ID and infant accommodating tubes 2.5–4 mm. This technique can be used with general anesthesia, appropriate sedation, and regional or

²Laerdal Inc., Long Beach, CA.

topical analgesia. The use of a light wand via the LMA has also been described. The Bonfils Optical Stylet (Karl Storz)³ provides a further advance since this device will accommodate a 2.5 mm ID tracheal tube with a video camera and monitor to facilitate visualization of the laryngeal inlet. The Shikani optical stylet (Clarus Medical, Minneapolis USA) best accommodates a 3.0 tracheal tube. These devices have, in general, been superseded by videolaryngoscopes.

Retrograde Intubation. This technique depends on threading a wire proximally through the vocal cords into the pharynx via a needle passed percutaneously into the trachea. This is a very dangerous technique with a reduced success rate particularly in infants and small children because of their compliant tracheas and the very small tracheal dimensions that together increase the risk of tracheal and extratracheal tissue injury. If cannulated successfully, the wire is then retrieved in the mouth and used to guide a tube into the trachea. A modification of this technique passes the retrieved wire retrograde up the suction port of a bronchoscope. The scope is then guided into the trachea by the wire and can be used to position the tube. This technique is rarely used in children except as a last resort to establish an airway.

Failed Intubation. If intubation options are failing, consider the following:

1. Should we awaken the child and reschedule?
2. Can this case be done safely with mask anesthesia?
3. Can this case be done safely with an LMA for airway support?
4. Do we need a surgical airway?

Extubation of the Trachea

1. Children are prone to laryngeal spasm during extubation, especially if extubated during a light plane of anesthesia. Therefore,
 - (a) Before extubation, ventilate with 100 % oxygen and ensure that all airway equipment for reintubation if necessary is available.
 - (b) Extubate the airway when the child is fully awake (or, if indicated, deeply anesthetized).
 - (c) Some children should not cough or strain with the tracheal tube in situ during emergence (e.g., those having neurosurgery or intraocular surgery). This may be achieved with a planned “deep” extubation, preceded by careful suctioning of the stomach and pharynx. Lidocaine, 1–2 mg/kg IV administered slowly before extubation, also decreases the risk of coughing and breath holding. After the tracheal tube is removed, a

³Storz Inc., Endoscopy-America Inc., Culver City, CA.

facemask should be applied, the airway maintained, and oxygen administered until the child is awake. Studies suggest that oxygen saturation (SaO_2) levels are better maintained if extubation is performed while the child is still anesthetized and oxygen is then given by mask until the child is fully awake.

- (d) When judging whether the child is "awake" enough for awake extubation, wait until the eyes and mouth open spontaneously, all limbs are moving, and the child resumes regular spontaneous ventilation after coughing.
 - (e) Do not disturb the child unnecessarily during the awaking stage, so as to minimize coughing and bucking on the tube before the child is fully awake. A "no touch" technique while waiting to extubate awake is very successful in many children.
 - (f) All monitors should be left in place until successful extubation is complete.
2. Severe laryngospasm upon extubation may be followed by pulmonary edema as the laryngospasm is relieved. If this occurs, it should be treated by continued positive pressure ventilation and a diuretic (e.g., Lasix).
 3. The following children should be fully awake before extubation:
 - (a) All those in whom tracheal intubation was difficult.
 - (b) All those having emergency surgery; these children may vomit gastric contents during emergence from anesthesia.
 - (c) All infants.
 4. Children who have had a mouth gag with tongue blade inserted by the surgeon (e.g., for cleft palate repair) are at risk for postoperative swelling of the tongue; always inspect the mouth before extubation.

Extubation of the Difficult Pediatric Airway

Extubation should be performed as a well-planned exercise, with the necessary equipment and personnel to reintubate the child readily available. In selected children a trial extubation, leaving an airway exchange catheter in situ, may be indicated.

All children with difficult airways should be extubated or have the LMA removed only after they have fully regained consciousness and when all danger of swelling in the region of the airway has passed. Corticosteroids (dexamethasone) have been used before extubation to decrease the likelihood of stridor, and all children should be given humidified oxygen after the tracheal tube is removed.

The golden rule: If there is any doubt about the airway, leave the trachea intubated.

Pediatric Anesthetic Circuits

The ideal anesthetic circuit for children should be lightweight; with low resistance and dead space; with low compliance; adaptable to spontaneous, assisted, or controlled ventilation; and readily humidified and scavenged. These conditions were most nearly met by the T-piece systems; however, modified circle systems are now almost exclusively used for children. The T-piece system is sometimes used during patient transport and by some anesthesiologists for small patients breathing spontaneously. A fresh gas flow rate of twice the minute ventilation is required to prevent rebreathing.

Circle Absorber Systems

The circle absorber system may be modified for use in children by incorporating a smaller-diameter breathing circuit. Though the resistance within a circle system is slightly greater than that of the T-piece, this is less significant with modern anesthesia workstations. The site of increased resistance is the small diameter tracheal tube. The integrity of the circle system and the presence and correct functioning of the valves and CO₂ absorbent (90% calcium hydroxide) must be carefully checked before each use. The use of very low flows (FGF < VE (expired minute ventilation)) delivered to a circle system has been associated with the accumulation of carbon monoxide (CO) within the circuit. The CO is thought to originate in accumulated expired gases or from degradation of inhalational agents (isoflurane, desflurane). In the United States, a minimum fresh gas flow of 2 Lpm is recommended for sevoflurane to prevent accumulation of compound A. Both CO and compound A may be avoided by using a lithium-based carbon dioxide absorbent.

Controlled Ventilation During Anesthesia

During anesthesia, ventilation may be controlled using manual or mechanical ventilation.

Manual Ventilation

This is used at times, especially during induction and when there is doubt about the adequacy of ventilation. It has been claimed that manual ventilation enables the anesthesiologist to monitor compliance continuously and to compensate rapidly for changes. Although this may be true for the experienced pediatric anesthesiologist with the Ayre's T-piece, there is some question about the ability of individual anesthesiologists to detect even complete airway obstruction just by the feel of the bag with circle circuits. However, if there is any doubt about the

adequacy of ventilation or in the event of sudden deterioration in the child's vital signs, it is wise to switch to manual ventilation. Then the adequacy of ventilation should be further confirmed by auscultation of the lungs, observation of chest movement, and the PetCO₂ waveform.

Rapid ventilation with small tidal volumes provides optimal results in the neonate because this pattern of ventilation tends to maintain the functional residual capacity and prevent airway closure. PetCO₂ levels should be monitored continuously because it is very easy to over-ventilate small infants. Hyperventilation (and consequent respiratory alkalosis) causes significant adverse physiological changes including reduced cerebral blood flow (apoptosis in animal models) and should be avoided.

Mechanical Ventilation

Current anesthesia workstations with compliance compensation incorporate circuitry and ventilators capable of delivering appropriate tidal volumes to even the smallest infants. However, it is also important to apply patterns of ventilation ("lung protective") that will avoid causing ventilator induced lung injury (VILI). This is essential in the critically ill neonate. Lung injury may be caused by excessive pressures (barotrauma) or tidal volumes (volutrauma). Volume targeted ventilation (VT) with at least 5 cm of positive end expiratory pressure (PEEP) is often preferred for small infants. V_t in the range of 4.5 mL/kg with PEEP is recommended for the sick preterm infant. The use of VT avoids the possibility of volutrauma to the lungs and the use of PEEP will maintain the FRC and optimize gas exchange. The use of a flow detector between the tracheal tube and the anesthesia circuit greatly facilitates this process. The adequacy of ventilation must be constantly monitored. Mild degrees of hypercapnia are preferred to hypocapnia in the neonate. The application of PEEP is essential to prevent a decrease in FRC below the closing volume, which inevitably accompanies anesthesia. In infants whose lungs are already being ventilated in the NICU, similar ventilator settings should be applied in the O.R. In some cases, a neonatal ventilator may be used in the O.R. or the decision may be to operate in the NICU, the latter especially for those infants who require high frequency oscillatory ventilation (HFOV). Older children may be ventilated applying the principles that are effective in adults; current recommendations suggest smaller tidal volumes with PEEP may be optimal in those with lung disease.

HFOV prevents volutrauma due to repetitive lung expansion and permits selection of a mean airway pressure that is optimal for gas exchange, yet minimizes lung damage. While it has often been used in children failing conventional ventilation methods, it has a probable role as primary therapy in those at risk of significant lung damage.

Pressure support ventilation (PSV) may be advantageous in spontaneously breathing patients, improving gas exchange and decreasing the work of breathing. PSV via an LMA has been found to be a useful technique for ambulatory pediatric surgery patients. PEEP is set to ~4 cm/H₂O and the flow trigger set at the lowest value that will avoid auto-triggering; usually about 0.4 L/min. The pressure should be set to achieve a V_t of approximately 6–8 mL/kg, greater pressures will be required in younger patients.

Humidification of Anesthetic Gases

Humidification of inspired gases during anesthesia has been recommended to prevent damage to the respiratory tract by dry gases and to minimize heat loss via the respiratory tract and thereby assist in maintaining normothermia. Dry gases inhibit ciliary activity and lead to the accumulation of inspissated secretions, which may, in the extreme, progress to obstruct the tracheal tube. Degenerative changes in cells exfoliated from the trachea after exposure to dry gas have been described, but an increased incidence of postoperative morbidity from pulmonary complications remains unproven. Gases should be humidified during very prolonged surgery and in the intensive care unit to reduce the risk of tube blockage from inspissated secretions.

Humidified anesthetic gases significantly reduce heat loss during surgery, particularly in neonates and infants. However, heated humidifiers are not as easily incorporated into circle circuits and accumulated water may plug capnogram tubing. An alternative means of humidification for older children is the use of a heat and moisture exchanger (HME) inserted at the connection of the tracheal tube to the circuit. The HME conserves approximately 50 % of the water normally lost via the respiratory tract and thus prevents a corresponding heat loss. The HME is most efficient with smaller tidal volumes and greater respiratory frequency, so it is quite useful in pediatric cases. These normally are very low resistance, but if blocked by secretions will significantly increase airway resistance; always monitor ventilation carefully when an HME is used. Use of low fresh gas flow in circle systems will also help to reduce water loss.

Monitoring During Anesthesia

Routine Monitoring Methods

Monitoring during anesthesia must always include the following:

1. Pulse oximeter: apply before induction and leave in place during transport and during the recovery room stay. The light source and sensor must be positioned to transilluminate a part of the body (earlobe, finger, toe, palm of

hand, or sole of foot, depending on the size of the child). Placement on the earlobe or buccal angle rather than the finger or foot may result in a slightly faster initial response time during acute desaturation. Placement at a preductal site (head or right hand) is desirable in infants with any potential for patency of the ductus arteriosus. A second postductal oximeter probe is useful to detect shunting. The sensor(s) should be protected to prevent outside light or pressure from interfering with the reading. Pulse oximetry has proved most effective in providing an early warning of developing hypoxemia. Failure of the pulse oximeter to detect and record a pulsatile flow may provide useful warning information about the child's circulatory status. However, if the pulse oximeter fails, check the child first (color of mucus membranes or nail beds, heart rate, breath sounds, blood pressure); then, if necessary, troubleshoot the equipment.

Pulse oximetry is relatively accurate throughout a wide variation in hematocrit. In children with cyanotic congenital heart disease the oximeter tends to overestimate saturation at lower readings (below SpO_2 70%). Similarly, the faster the rate of desaturation, the more the oximeter underestimates the true hemoglobin saturation.

Fetal hemoglobin (HbF), hemoglobin SS, and hyperbilirubinemia do not affect the pulse oximeter measurement.

Nail polish or disease of the nails may affect the performance of the monitor (blue or green nail polish); dark skin decreases the accuracy of oximeters at low saturations. Methemoglobin (MetHb) and carboxyhemoglobin (CoHb) affect the accuracy of readings: the former has a nonlinear effect, either underestimating or overestimating saturation, and the latter overestimates saturation.

An arterial saturation of 90–95%, which reflects a PaO_2 of 30–67 mmHg in infants <29 weeks is a safe range for the preterm infant. But because of the slope of the Hb/ O_2 dissociation curve, pulse oximetry is less precise in the assessment of hyperoxia than it is in hypoxia. If considered necessary, an arterial sample can be obtained to confirm which level of saturation is appropriate in terms of PaO_2 for each child. This level of saturation can then be maintained by varying the fraction of inspired oxygen (FIO_2).

The complications of pulse oximetry are few, but severe burns have occurred when an incorrect sensor from a different manufacturer has been substituted. Burns may also occur when pulse oximetry is incorrectly used in the magnetic resonance imaging suite (see Chap. 18).

2. Stethoscope, precordial or esophageal: there must be provision to monitor heart and breath sounds throughout anesthesia. Recently, there has been a trend away from the use of a stethoscope; however, should there be an equipment failure, this is an essential aid. If the monitors stop working, check the child first.

3. Blood pressure (BP) cuff of suitable width: the cuff should occupy two-thirds of the upper arm. If the cuff is too narrow, the BP readings are falsely high; if it is too wide, they are falsely low. A width of 4 cm is recommended for full-term neonates. A noninvasive blood pressure device (i.e., Dinamap) may be used, but ensure that it is set to provide readings at a maximum of 5 min intervals.
4. Electrocardiogram: It is standard to monitor the EKG; however, the EKG is of limited value in pediatric cases. Any arrhythmias that occur are usually benign, and bradycardia on the EKG is a very late sign that the child is in trouble.
5. Thermistor probe (axillary, esophageal, or rectal) (see Management of Body Temperature).
6. End-tidal carbon dioxide: This device noninvasively reflects the adequacy of ventilation and pulmonary perfusion. It also provides the most reliable indicator of successful tracheal intubation and should be used whenever tracheal intubation is performed. Two types of monitors are available: measuring carbon dioxide "in-line" at the connector and by sidestream sampling from the circuit. The latter method is more commonly used. However, it is not as easy to apply in infants and small children owing to the small size of the ventilatory volumes. When a partial rebreathing circuit is used (i.e., a T-piece plus ventilator), end-tidal sampling must be obtained from within the lumen of the tracheal tube for all small children (i.e., those weighing less than 12 kg) if useful numbers are to be obtained. When a circle circuit is used, proximal sampling at the tracheal elbow gives valid results even in small infants. The presence of a leak around the endotracheal tube may also affect end-tidal sampling, especially when positive end-expiratory pressure is applied; with a very large leak, the PetCO₂ waveform may disappear completely.

PetCO₂ measurements correlate poorly with the PaCO₂ in children who have congenital heart disease with a right-to-left shunt or mixing lesion; the lower the saturation, the greater the PaCO₂–PetCO₂ gradient. In those with left-to-right shunting, PetCO₂ readings are accurate.

A decrease in the PetCO₂ provides a very early indication of a reduction in pulmonary blood flow. This may be useful in the early diagnosis of cyanotic spells in the child with tetralogy of Fallot. A decrease may also be diagnostic of pulmonary embolism, air embolism, or a low cardiac output state. If CPR is required an PetCO₂ of 10–15 mmHg indicates adequate chest compressions and pulmonary blood flow; values less than this indicate inadequate compressions.

7. Peripheral nerve stimulator: should be used whenever nondepolarizing muscle relaxants are administered.
8. Arterial catheter: should be inserted for direct measurement of BP and to provide for intermittent blood gas analysis when required. The radial or

femoral artery is usually cannulated (see later discussion); rarely, the axillary artery may be used. We recommend checking for collateral flow from the ulnar artery when cannulating the radial artery. Do not use the brachial artery, which has poor collateral vessels. (See Precautions with Arterial Lines.) Cannulation of the superficial temporal artery has been described but this introduces the possibility of retrograde intracranial embolization. Children with Down syndrome may have a single (median) artery in the wrist, in which case cannulation should be avoided. (Always check the wrist vessels in Down syndrome children before attempting cannulation.)

9. Urine output: record this at regular intervals for all children undergoing major surgery and all who have hypovolemic shock or whose renal function may be impaired.
10. Central venous pressure (CVP): record from a catheter inserted centrally via the internal or external jugular vein (see later discussion). The external jugular is a less reliable route for CVP monitoring but is often useful for fluid replacement and drug infusions. The CVP should always be monitored in children in whom major blood loss and/or impaired cardiac performance is anticipated.

Cannulation Techniques

Radial Artery Cannulation

The left radial artery is often preferred for arterial puncture (in right-handed children).

1. Locate the artery by palpation; if this is difficult, use ultrasound, or in small infants, transilluminate the wrist with a bright cold light.
2. Use careful aseptic technique and prepare the skin with chlorhexidine.
3. Make a small skin incision over the artery with an 18-gauge needle. This prevents damage to the tip of the cannula during skin puncture.
4. Perform arterial puncture; as soon as blood issues into the hub of the needle, turn the needle so that the bevel faces down.
5. Advance the cannula gently into the artery (Fig. 4.3).
6. If it will not advance, withdraw until blood flows freely and carefully insert a fine guide wire,⁴ then advance the cannula over the wire.
7. If you fail to enter the artery at all, remove the needle and palpate the artery again, critically evaluating its alignment with the skin puncture. Then try again.

⁴0.018 (0.46 mm) dia.× 25 cm spring guide wire, AW-04018, Arrow International Inc., Reading, PA.

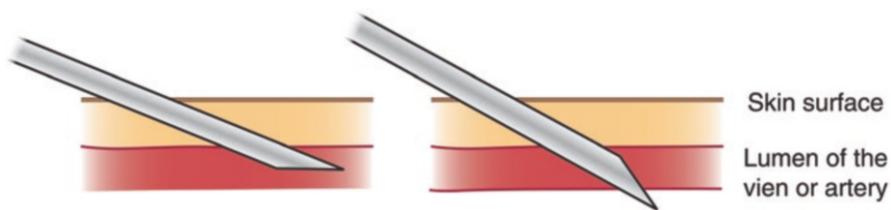


Fig. 4.3 Cannulation of small veins or arteries. *Hints:* Success may be improved if the bevel of the needle is turned down as in (a) so that the entire breadth of the end of the needle is intraluminal versus the situation in (b) where the tip may be through the distal wall and prevent advancing the catheter off the needle. Alternatively, when a flashback of blood is seen in the hub of the cannula a fine guide wire may be advanced through the needle to guide the cannula into the vein. Adapted from image in: (1) Filston HC, Johnson DG. Percutaneous venous cannulations in infants and children: a method for catheter insertion without cut-down. *Pediatric* 48: 896–901, 1971. (2) Steward DJ. Venous cannulation in small infants: A simple method to improve success. *Anesthesiology* 90: 930–31, 1999

8. Apply antibiotic spray or ointment to the skin puncture site and cover with a sterile dressing (e.g., Tegaderm, 3M Health Care Inc.).
9. Secure cannula carefully with adhesive tape. All connections should be Luer-Lok or similar to prevent accidental bleeding.
10. Place a red label indicating that this is an arterial line at the site of insertion and all connections to prevent confusion with a venous line.

Femoral Artery Cannulation

In some children the radial artery cannot be cannulated or is inappropriate (e.g., after surgery of the aortic arch). In such cases the femoral artery may be used.

1. Place a low pad under the child to elevate the pelvis.
2. Palpate the femoral artery *below* the inguinal ligament. If the artery is difficult to palpate, ultrasound may facilitate identification of the artery.
3. Use careful aseptic technique and prepare the skin with chlorhexidine.
4. Using the Seldinger technique, puncture the artery *below* the inguinal ligament using an appropriate-sized thin-walled needle. Punctures superior to the inguinal ligament introduce the risk of a retroperitoneal bleed.
5. Avoid needling the head of the femur; aseptic necrosis may result in infants and young children.
6. When the artery is entered, insert a guide wire and use it to introduce an appropriate size catheter. Secure the catheter carefully in place and cover it with antibiotic ointment and a clear plastic dressing.

Precautions with Arterial Lines. Regarding any arterial cannulation:

1. Insert the cannula with meticulous asepsis.
2. Secure all connections, using Luer-Lok fittings, to exclude the danger of accidental disconnection and hemorrhage. Plug sampling taps when not in use. Tape stopcocks in the “line-open” position if they will be hidden under the drapes and inaccessible. Clearly indicate all access ports with red tape or label as “arterial line” so as to avoid accidental drug administration through the arterial line.
3. For radial lines, immobilize the forearm and wrist on a padded splint to prevent accidental decannulation.
4. Use a continuous flush device, but beware of accidental fluid overload. Use 1 N or 0.5 N saline with heparin (1000 IU/500 mL). Do not use dextrose because of increased risk of infection of the line.
5. Beware of embolization.
 - (a) Do not reinfuse blood removed during sampling into the arterial line. Return the blood to an IV line.
 - (b) Do not use high pressure to attempt to clear a blocked cannula.
 - (c) Infuse only small volumes of flush fluid after sampling. In small infants, volumes of only 0.5–1.0 mL injected into the radial artery may flow retrograde into the cerebral vessels.
6. Remove the arterial line as soon as it has served its purpose. Complications (especially arterial thrombosis and sepsis) increase with the duration of cannulation of the vessel.

Internal Jugular Vein Cannulation

The use of an ultrasound probe improves the success rate and decreases the incidence of complications.

1. Position the child—head to slightly leftwards, 20° Trendelenburg, with a rolled towel under the shoulders to reduce concavity of the neck.
2. If available, use the ultrasound probe to locate and mark the position of the internal jugular vein (IJV) at the level of the cricoid cartilage. It is helpful to also note the relation of the IJV to the carotid artery at this level.
3. Prepare skin and drape, and gown and glove. (Gowning and gloving to insert CVP catheters decreases infection rates.)
4. Using the Seldinger technique, insert the thin-walled needle through the skin at 45° over the marked course of the IJV until the vein is punctured: a flow of venous blood is usually obtained as the needle is being slowly withdrawn.
5. Hold the needle very still, pass the guide wire; there should be no resistance to passage. The occurrence of premature ventricular beats indicates passage of the guide wire into the right ventricle; withdraw the wire several centimeters.

Remove the needle, make a skin nick with a scalpel blade, and complete the cannulation after advancing and removing the dilator. Note that gentle pressure on the liver will greatly increase the size of the IJV and facilitate cannulation.

If ultrasound is not available, cannulation may be performed using anatomic landmarks; after positioning the child as described in (1) previously:

1. Palpate the carotid pulse medial to the sternocleidomastoid and pick this muscle up to identify its bulk at the level of the thyroid cartilage prominence (the midpoint between the mastoid process and the sternal notch).
2. At the level of the bifurcation of the sternomastoid into the sternal and clavicular heads, aim the finder needle under the medial border of the clavicular head aiming for the ipsilateral nipple. Gradually fan medially until the vein is found. Thread the guide wire down the needle and withdraw the needle. Using a scalpel to create free access through the subcutaneous tissue, insert a dilator over the wire. After withdrawing the dilator, insert the catheter over the wire until it is properly positioned and then remove the wire. Aspirate each lumen of the catheter and flush with heparinized saline. Throughout this procedure, steer clear of the carotid pulsation.
3. Suture the catheter in situ, cover with antibiotic ointment and a sterile dressing.

N.B. In cyanotic children, it is advisable to attach the needle or cannula to a transducer and confirm that the pressure is venous before using the dilator.

To position the tip of the catheter at the junction of the superior vena cava and right atrium, the length should be equal to the distance from skin penetration to a point 2 cm below the upper border of the manubrium.

The tip of the guide wire, accurately placed to just protrude from the catheter, may be used as an internal EKG electrode to position the catheter. Connect the L (leg) lead to the wire with a sterile alligator clip and look at lead II; a biphasic P wave is seen if the catheter is correctly placed.

Always check the position of the tip of the catheter on the radiograph if the catheter is to be left in place postoperatively. Catheters that extend into the right atrium may perforate the heart.

If IJV puncture cannot be performed and the external jugular vein is visible, it may provide a useful alternative route for CVP monitoring.

External Jugular Vein Cannulation

1. Position the child with a 20° head-down tilt and a small roll of towels under the shoulders.
2. Locate the external jugular vein and prepare and drape the area.
3. Puncture the vein using a 22-gauge intravenous catheter.

4. Feed a J-wire through the catheter and advance it centrally, rotating as necessary. A J-wire with a 3 mm radius of curvature at the J is most likely to pass easily into the subclavian and towards the heart. The wire must be manipulated gently to avoid the possibility of puncturing the vein. It may be advantageous to abduct the child's arm to direct the wire towards the heart.
5. When the wire has advanced, a dilator may be gently inserted, taking care not to pass it more than 1–2 cm into the external jugular vein, otherwise it might tear the vein at the junction with the subclavian.
6. Advance the soft central venous catheter over the guide wire until it is sited at the junction of the superior vena cava and right atrium. This distance can be judged by measuring the distance from skin puncture to manubriosternal junction.
7. The position of long-indwelling CVP lines should always be checked by radiography. Complications (including perforation) may occur if the line is advanced too far.

Other Important Forms of Monitoring

Blood Glucose. Infants, especially preterm or small-for-gestational-age infants, are prone to hypoglycemia; their blood glucose levels should be checked frequently. This may be accomplished using an inexpensive handheld glucometer. The results obtained are accurate enough to detect important abnormalities. Hypoglycemia (less than 30 mg/dL in first 24 h of life and 45 mg/dL thereafter) should be corrected using a glucose infusion (6–8 mg/kg/min). Avoid excessive glucose administration because it may result in hyperglycemia and glycosuria leading to dehydration and electrolyte losses. Hyperglycemia may increase the extent of cerebral damage should a hypoxic/ischemic episode occur.

Fluid Administration. The intravenous administration of fluids must be very carefully monitored to avoid overload. Syringe pumps or controlled intravenous infusion pumps are recommended for all children, and especially for preterm infants and neonates. Sum up all fluids given, including those given with drugs. The use of a low-volume remote injection site⁵ or an injection cap at the infusion line site minimizes the need for large volumes of flushing fluid. Tuberculin syringes should be used to measure small doses accurately, avoiding the need to dilute drugs to give accurate doses. When medications are administered in small volumes, flush the IV line to be sure the medication reaches the bloodstream in a timely manner.

⁵ Mini-Vol micro bore extension sets, SIMS medical, Keene, NH.

Anesthesia Chart. The anesthesia chart is an important monitor and, if well kept, permits the anesthesiologist to detect important trends in the child's progress.

Management of Body Temperature

Monitoring

Continuous monitoring of body temperature with a thermistor probe is essential for all children undergoing general anesthesia. In larger children having minor surgery, the temperature is usually recorded from the axilla. This reflects the core temperature provided that the tip of the probe is close to the axillary artery and the child's arm is adducted. Adhesive skin temperature sensors (e.g., on the forehead) do not provide an accurate estimate of core temperature. In infants and children undergoing major surgery, core temperature is best monitored in the distal esophagus or rectum. Esophageal temperatures should be recorded at the lower third of the esophagus to avoid falsely low readings caused by gases flowing into the trachea. When using an esophageal stethoscope with a thermistor, adjust the position until the heart sounds are loudest; the thermistor is then optimally placed behind the left atrium.

Tympanic membrane probes have been used to monitor core temperature. The tympanic membrane temperature closely follows lower esophageal temperature, but care must be taken not to damage the ear. Whenever possible, we prefer to take the safe, easy, and reliable course and monitor the esophageal temperature during major surgery.

Conservation of Body Heat in Neonates

The objective of body heat conservation is to prevent cold stress and to avoid hypothermia. A reduced body temperature affects recovery from anesthetic and relaxant drugs, impairs coagulation, may depress ventilation, may result in arrhythmias, and increases postoperative oxygen consumption.

The sources of body heat loss are radiation (39 %) > convection (34 %) > evaporation (24 %) > conduction (3 %). Hence preheating the OR and wrapping the child reduces radiation heat loss; the use of a forced air warmer in a closed space decreases convection and conduction heat loss. The use of an HME reduces evaporative losses.

Preoperatively. Adjust the OR ambient temperature to 24 °C (75 °F) or greater. Prepare a forced air heater and mattress. Keep the infant in the heated transport incubator until you are ready to induce anesthesia.

Intraoperatively. Neonatal forced hot air blankets are under body blankets. Thus, they are placed on the table before the infant arrives and the blanket is turned on (temperature set point is 43 °C). If the room is warmed and the forced air blanket used, an infrared heating lamp is no longer needed for induction of anesthesia. However, the forced air blanket is turned off from just before surgical prep is applied to the skin until the infant is covered by the surgical drapes. If there is concern that this interval will result in heat loss, then an infrared heating lamp may be placed in position. Keep a cap on the infant's head whenever possible. Use warmed intravenous solutions when large volumes will be infused. A heated humidifier (at 36 °C) may be useful. Warmed (40 °C) skin preparation solution should be used, and any excess should be dried from the skin to prevent cooling by evaporation. The use of plastic drapes creates a "warm tent" around the infant.

Postoperatively. Use the forced air warmer during extubation and other procedures at the end of anesthesia. Alternately, the infrared heater may be used. Place the infant in a warmed incubator and return the infant promptly to the post-anesthesia care unit or the NICU.

Hyperthermia During Surgery

Hyperthermia sometimes develops during surgery if all of the described heat-conserving procedures are followed. If this occurs, the forced air heater should be set to ambient temperature until the infant's temperature begins to decrease and then discontinued. Other causes for hyperthermia during surgery include pyrexial reactions (e.g., from manipulation of an infected organ or a blood transfusion reaction) and very rarely, malignant hyperthermia (MH) (see p. 178), cocaine overdose and thyroid storm.

Intravenous Therapy

1. For all children weighing less than 20 kg, insert a buretrol or similar graduated reservoir between the intravenous bag and the administration set; this prevents accidental fluid overload and permits an accurate check of infused volumes.
2. Always use an infusion pump. This allows accurate control of the rate of infusion and easy monitoring of volumes administered. It also provides a warning if the infusion becomes obstructed.
3. Percutaneous insertion of a plastic cannula into a vein is considered optimal. If this is to be done with the child awake, apply a topical local anesthetic cream to the site (in advance, see p. 634). Use a 22-gauge cannula or larger if blood transfusion may be required. Observe strict asepsis when performing

cannulation; use an appropriate skin preparation solution and cover the puncture site with a sterile dressing. Label the intravenous line with the size of the cannula and the date and time of insertion.

4. For major abdominal surgery, intravenous access should be secured in the upper limbs.
5. Before surgery commences, ensure that the intravenous line is working well. Do not embark on any procedure with a doubtful intravenous line.
6. Do not be lulled into security if the child has a multi-lumen CVP since resistance to flow may be so great that infusion of blood is impossible. It is unwise to rapidly infuse blood into CVP lines in neonates; both the temperature and K⁺ content of stored blood may cause serious arrhythmias.

Venipuncture for Induction of Anesthesia

The ability to perform a venipuncture painlessly and to cannulate small veins successfully is essential for the pediatric anesthesiologist. Some tips that may help follow.

For venipuncture for drug administration:

1. Apply a topical local anesthetic cream in advance whenever possible (see p. 634). Alternatively, nitrous oxide may be administered (50–75% inspired concentration) to sedate the child during venous cannulation. Make sure that you have a skilled assistant who can distract the child while applying gentle restraint.
2. Do not use tourniquets in awake children; have your assistant grasp the arm to gently impede venous return and gently squeeze the hand or foot to shift blood into the veins. Do not attempt venipuncture unless the vein is obviously well filled; filling can be facilitated by having the assistant hold the child's hand below body level and apply gentle manual constriction to the limb.
3. Be aware that directly injecting medications into veins in the antecubital fossa, anatomical snuffbox, or scalp has a greater risk for accidental intra-arterial injection than at any other sites and the consequences, which include arterial spasm, may be catastrophic.
4. A vein on the dorsum of the hand or the foot is usually most suitable.
5. Use the smallest size needle and syringe possible, and keep the equipment from the child's view at all times.
6. Hold the needle and syringe firmly and avoid accidentally touching the skin with the needle until ready to puncture the vein.
7. When ready, puncture the skin and vein firmly with one rapid movement and then hold the butterfly needle firmly in place until the injection is completed.

Venous Cannulation

This may be performed after induction of anesthesia; otherwise, apply a topical local anesthetic cream in advance of the procedure (see page 634).

1. Select a suitable vein. The best sites usually are the dorsum of the hand, the medial aspect of the ankle (saphenous vein), the lateral aspect of the foot, a scalp vein (in infants), and the lateral aspect of the wrist (in older children). Consider which sites are appropriate, i.e., children with abdominal trauma or a tumor must have intravenous access in an upper extremity. Those children who will be using crutches after surgery should not have an intravenous line in the back of the hand.
2. Use careful aseptic technique and prepare the skin with appropriate preparation solution (chlorhexidine is now the preferred solution except in neonates).
3. Make sure the vein selected is well filled and make a small incision over it with an 18-gauge needle.
4. Note the direction of the bevel on the cannula needle. After the initial venous puncture is made and a flashback seen, turn the bevel face down before attempting to advance the cannula into the vein. This ensures that the point of the needle is unlikely to be in the distal wall of a small vein and that the cannula will advance unimpeded into the vein (see Fig. 4.2).
5. When the cannula is in place, cover it with a sterile dressing. Tape the cannula firmly in place and immobilize the limb on a splint.
6. When inserting a cannula into a very small vein it may help to pass a fine guide wire into the vein before attempting to advance the catheter:
 - (a) A 22-gauge Angiocath is inserted towards the vein at a shallow angle.
 - (b) As soon as there is a “flashback” of blood into the hub of the needle, the cannula is held absolutely still and the needle is very gently removed. Blood is usually seen flowing back into the cannula.
 - (c) A 0.018-in. (0.46-mm)⁶ diameter spring wire guide is then gently advanced through the cannula into the vein. In most instances this guide wire is easily inserted, even into very small veins, and can be seen tracking inside the vein for some distance up the limb. The cannula is now advanced over the guide wire with full confidence that it will end up lying freely within the lumen of the vein and will provide a very reliable intravenous route.

⁶Arrow RA-04018, Arrow International Inc., Reading, PA.

Intraosseous Infusions

When venipuncture is impossible and urgent fluid or drug therapy is indicated, the intraosseous route should be employed. Any drug or solution that can be given intravenously can also be given by this route. Continuous infusions can be given. In “shock” or “arrest” states, absorption from the intraosseous site may be more rapid than from a peripheral intravenous line.

The usual insertion sites are the distal femur (midline 1 cm above the patella) and the proximal tibia (medial on the tibial plateau 1 cm below the tuberosity); accidental injections into the epiphysis do not usually cause any harm. A bone marrow needle, a strong large-bore spinal needle, or a specifically designed intraosseous needle is firmly advanced through the bone until a “give” is noted and the needle stands rigidly. At this point bone marrow can be aspirated and fluid can be injected with very little resistance and with no swelling or extravasations. After initial fluid resuscitation by this route, it is often possible to start an intravenous infusion into a peripheral vein. The development of the EZ-IO which uses a battery-powered drill-like device limits the depth of insertion of a specifically designed needle and provides a low profile connection for the IV fluid.⁷

Preoperative Fluid Replacement

Most children will have been taking oral fluids up until 2 h before surgery and will be well hydrated. However, some infants may have serious fluid depletion secondary to their disease, which must be corrected before surgery. Preoperative dehydration in infants can be classified by the size of the deficit as mild, moderate, or severe:

Mild dehydration: 50 mL/kg (5% body weight loss) (normal mucous membranes, tears present, flat anterior fontanel, normal skin turgor, normal to slightly increased heart rate)

Moderate dehydration: 100 mL/kg (10% body weight loss) (dry mucous membranes, normal to reduced tears, possible sunken anterior fontanel, slightly increased skin turgor, tachycardia, oliguria)

Severe dehydration: 150 mL/kg (15% body weight loss) (parched mucous membranes, absent tears, sunken anterior fontanel, sunken eyeballs, increased skin turgor, tachycardia, hypotension, anuria)

⁷ Arrow® EZ-IO® Intraosseous Vascular Access System, Teleflex, Shavano Park, TX.

Replacement of water and electrolytes should proceed in three phases:

PHASE 1. *Treatment of overt or impending shock* (severe dehydration and hypovolemia): Start with balanced salt solution (Lactated Ringer's solution or normal saline—20 mL/kg) while ordering packed cells (10 mL/kg) if anemia is present or 5% albumin (20 mL/kg) if anemia is not present. If albumin is not available or a coagulopathy is present, consider plasma.

PHASE 2. *Replacement of extracellular water and sodium:* Half the estimated fluid deficit should be replaced over the initial 6–8 h as 0.3 N saline. If the deficit is severe, give an initial infusion of 1 N saline (20 mL/kg). The degree of success of this therapy can be gauged from the clinical signs (heart rate, arterial and venous pressures, and urine output). The following formula is useful in correcting sodium deficiency:

$$\text{Na}^+ \text{ deficit (mEq)} = [\text{normal Na}^+ (\text{mEq}) - \text{measured Na}^+ (\text{mEq})] \times 0.6 \times \text{weight (kg)},$$

where 0.6 = diffusion constant.

Severe metabolic acidosis should be treated simultaneously, using the formula

$$\text{Dose required (mEq of HCO}_3\text{)} = \text{base deficit} \times 0.3 (0.4 \text{ for infants}) \times \text{weight (kg)}$$

Give half the calculated requirement, then reassess the acid–base status.

PHASE 3. *Replacement of potassium:* Potassium (K^+) replacement should begin only when a good urinary output has been established, according to the following:

1. Replace a maximum of 3 mEq/kg of potassium per 24 h.
2. The rate of administration should not exceed 0.5 mEq/kg/h.
3. Complete correction of severe K^+ deficiency may take 4–5 days.

These rates only serve as estimates and must be adjusted for changes in metabolic activity, clinical conditions, and extrarenal losses (i.e., gastric suction).

N.B. A neonate's insensible water loss decreases by 30–35 % when nursed in a high-humidity atmosphere or ventilated with humidified gases. Insensible water loss is increased by crying, sweating, hyperventilation, and the use of a radiant heater or "bili" lights. Pyrexia increases water loss by 12 % per 1 °C.

Intraoperative Fluid Management

Calculation of the volume and type of fluid for infusion must take into account:

1. Dehydration present *before* preoperative fasting.
2. Fluid deficit incurred *during* preoperative fasting. (To determine the fluid deficit, use the 4:2:1 rule for the hourly requirements for neonates and infants <6 months (otherwise see later discussion for fluid management) and take the product of that requirement and the number of hours of fasting.

Replace half that deficit in the first hour and one-fourth the deficit in each of the subsequent 2 h)

3. Maintenance fluid requirement during surgery.
4. Estimated extracellular third space fluid loss related to surgical trauma.
5. Replacement of blood loss.
6. Increase in body temperature.

For brief surgical procedures (<15 min) in otherwise healthy children, intravenous fluids usually are not needed intraoperatively if the preoperative deficit was small, blood loss or tissue trauma was minimal, and oral intake is likely to be reestablished early (e.g., myringotomy and tubes). An intravenous line should, however, be set up and ready for use should an emergency develop.

For surgical procedures of greater duration and/or when reestablishment of oral intake may be delayed:

1. Intravenous access should be established.
2. A balanced salt solution (e.g., Lactated Ringer's) should be administered intraoperatively and postoperatively until oral intake is reestablished.
3. The goal of administering balanced salt solutions is to replace both fluid deficits and ongoing losses in order to restore euolemia while preventing hyponatremia from developing. To do this, antidiuretic hormone (ADH) must be suppressed. ADH is normally upregulated in the perioperative period as a result of many stimuli including pain, inflammation, and stress. Children are much more susceptible than adults to brain damage that may develop from hyponatremia. The most effective means to suppress ADH (and thus hyponatremia) is to establish euolemia. To that end, Holliday and Segar revised their perioperative fluid strategy by recommending 10–40 mL/kg of balanced salt solution infused over several hours (10–20 mL/kg in the first hour) for most simple, non-blood loss surgical procedures. For postoperative fluid management, see #7 below.
4. For neonates and infants <6 months, we recommend retaining the 4:2:1 rule for fluid management (see below).
5. For those who were fasted for a prolonged period and others at risk for hypoglycemia, a glucose containing solution may be indicated to prevent hypoglycemia. A 1–2.5% dextrose solution in Lactated Ringer's, delivered using an infusion pump at maintenance rates, may be piggybacked into the main IV line. Check the plasma glucose concentration periodically. The use of a 5% dextrose in Lactated Ringer's may cause hyperglycemia during prolonged infusions.
6. For extensive surgery, especially in neonates (especially preterm infants), it is advantageous to separate the administration of dextrose from other fluid therapy. An infusion of 5 or 10% dextrose can be established at a rate of 5–8 mg/kg/min (using an infusion pump) to prevent hypoglycemia in these infants. Blood glucose levels should be checked periodically.

7. For surgical procedures causing extensive tissue trauma and/or blood loss, give additional fluids to replace extracellular fluid lost in blood or sequestered into damaged tissue. This deficiency should be replaced with a balanced salt solution (e.g., Lactated Ringer's solution) in which the electrolyte concentrations are similar to those in extracellular fluid (Table 4.9). Superficial or extremity surgery (associated with minimal fluid loss) requires only 5 mL/kg/h fluid replacement, moderate procedures (e.g., spine surgery) require 10 mL/kg/h, and major procedures (e.g., laparotomy) require 15 mL/kg/h. Thoracotomy is associated with much less translocation of fluid, so smaller fluid volumes are required. When replacing blood loss with balanced salt solutions (such as Lactated Ringer's solution), the volume of salt solution required is 3 mL for every milliliter of blood lost. As a general rule, we recommend ordering coagulation studies when 75 mL/kg balanced salt solution has been infused. After infusion of 100 mL/kg balanced salt solution, consideration should be given to switching to colloid solutions (or blood products). Adequacy of fluid replacement is best judged by continuous monitoring of the cardiovascular indices and urine output. If urine output is less than 0.5–1 mL/kg/h, the fluid infusion rate should be increased.
8. Postoperatively, hyponatremia is a real danger. It is usually associated with the intraoperative and postoperative use of hypotonic solutions or occasionally inappropriate secretion of ADH. Holliday & Segar recommended halving their original fluid infusion rate (4:2:1 rule) to 2:1:0.5 postoperatively to prevent hyponatremia while avoiding salt overload when using a balanced salt solution: 2 mL/kg/h for the first 10 kg, 1 mL/kg/h for the second 10 kg, and 0.5 mL/kg/h for >20 kg for the first 12 h postsurgery and then reversion to the 4:2:1 rule (4 mL/kg for the first 10 kg, 2 mL/kg for the second 10 kg, and then 1 mL/kg for >20 kg) thereafter, until oral intake is resumed. Serum electrolytes should be monitored after major surgery to preclude electrolyte imbalances from developing.

Blood Replacement

Preoperative Assessment

Although minor surgical procedures are commonly performed in children who are mildly anemic with no problems, a normal Hb level (Table 4.10) is desirable in every case of *major* elective surgery. If the child is anemic preoperatively, elective surgery sometimes may be delayed until the anemia has been investigated and treated. In other cases, surgery is more urgent; anesthesia for these children must be administered with a technique that is compatible with their anemia (see Chap. 6). When surgery cannot be delayed despite a very low Hb (e.g., 5–7 g/dL)

Table 4.9 Composition of electrolyte solutions

Solution	Concentration (mEq/L)				Concentration of HCO_3^- (mEq/L)			Osmolarity
	Na^+	K^+	Mg^{2+}	Ca^{2+}	Cl^-	Acetate HCO_3^-	Gluconate	
Lactated Ringer's solution ^a	130	4	—	3	109	—	—	28
Normal saline (0.9%)	154	—	—	—	154	—	—	276
Plasmalyte 148	140	5	1.5	—	98	527	—	308
0.3 N saline in D ₅ W	51	—	—	—	51	—	—	294
0.2 N saline in D ₅ W	34	—	—	—	34	—	—	—
Normosol-M	40	13	3	—	40	16	—	—
Normosol-R	140	5	3	—	98	27	23	280

Magnesium sulfate (2 mL amp., 50% w/v (where w/v is weight/unit volume)): 4.0 mEq Mg^{2+} /mL
Sodium bicarbonate (50 mL amp., 7.5% w/v): 0.9 mEq HCO_3^- /mL
Calcium gluconate (10 mL amp., 10% w/v): 0.447 mEq Ca^{2+} /mL
Calcium chloride (10 mL amp., 10% w/v): 1.36 mEq Ca^{2+} /mL
D₅W, 5% dextrose in water

^aOr Hartmann's solution

Table 4.10 Normal hemoglobin concentrations

Age	Hb conc.^a (g/dL)
First day of life	20 (18–22)
Second week	17
3 months	10–11
2 years	11
3–5 years	12.5–13.0
5–10 years	13.0–13.5
10+ years	14.5

^aThe Hb concentration declines gradually to about 10–11 g/dL during the first few months of life of a full-term infant as fetal Hb is replaced. It then gradually increases and is maximal at about 14 years.

and blood loss is expected, packed red blood cells (PRBCs) should be infused preoperatively. Approximately 4 mL/kg of packed cells and 6 ml/kg of whole blood increase the Hb concentration 1 g. If no or minimal blood loss is expected, then have packed cells available for possible transfusion.

The hemoglobin content of stored whole blood is 12 g/dL, that of PRBCs is 24 g/dL, and that of buffy-coat-poor washed cells is 28 g/dL.

When significant blood losses (10 % of the estimated blood volume [EBV] or greater) are expected, the child's blood group should be determined and an appropriate number of units cross-matched. Insert a CVP line preoperatively in children who are hypovolemic and/or may require extensive blood replacement during surgery.

Perioperative Management

At commencement of the operation, record the estimated blood volume (EBV), the preoperative Hb level, and an estimate of the maximal allowable blood loss (MABL) on the anesthesia record:

$$\text{MABL} = \frac{(\text{Starting Hct} - \text{target Hct}) \times \text{EBV}}{\text{Starting Hct}}$$

Assessment of Blood Loss

Accurate estimates of blood loss must be maintained throughout the surgery.

1. Monitor cardiovascular system indices; in infants, the systolic BP is the most reliable indicator of blood volume.
2. Measure blood loss from the surgical site:

- (a) All sponges must be weighed before they dry out. This method is simple and accurate (assume 1 g = 1 mL blood and subtract the known dry weight).
 - (b) Measure blood from suction (in graduated flasks).
 - (c) Estimate blood on drapes.
3. Chart the running total continually.
 4. Be aware of the possibility that blood losses may accumulate in body cavities (e.g., peritoneum, pleura) or on the surgical drapes and on the floor.

Blood Transfusion

The decision whether to transfuse blood must be based on the preoperative Hb level, the measured surgical blood loss, and the child's cardiovascular responses. As a rough guide, in otherwise healthy children, blood replacement may be necessary after loss of ~15% of the EBV. The need for blood transfusion can be determined more accurately from serial hematocrit (Hct) measurements. Normally, the Hct should be maintained at or greater than 30–40% in infants and in those children with significant cardiac or respiratory disease, and at approximately 20–25% in otherwise healthy children.

Check each unit of blood against the child's identity bracelet and mix it well by repeated inversion of the bag. Blood should be warmed to 37 °C before administration; it should not be heated to more than 38 °C, otherwise red cells may be damaged. PRBCs are commonly diluted in saline before administration. Massive blood transfusions in smaller children may require that the PRBCs be diluted in fresh frozen plasma (FFP) to prevent a dilutional coagulopathy. Calcium is rarely necessary during massive transfusion in children but should be given if persistent hypotension follows apparently adequate volume replacement in infants. This is of particular concern after transfusing FFP because it has the greatest concentration of citrate per unit volume of any blood product. If the FFP transfusion is ≥1 mL/kg/min, then ionized hypocalcemia is likely and exogenous calcium should be administered during the transfusion (5 mg/kg calcium chloride or 15 mg/kg calcium gluconate, repeat as needed). In severely shocked children who require rapid massive transfusion, be prepared to also give sodium bicarbonate if indicated by serial acid–base determinations. Avoid direct transfusion of cold blood products into the right atrium in infants, otherwise life-threatening arrhythmias may ensue.

Massive Blood Transfusion

If it becomes apparent that massive blood transfusion will be required (i.e., more than one blood volume), institute a massive transfusion protocol (see Chap. 17) and monitor coagulation indices. Platelet counts, prothrombin time,

and partial thromboplastin time together with tests for fibrinolysis (determination of fibrin split products) should be repeated at least after every 50 % blood volume replacement. It is helpful to have a preoperative platelet count if massive transfusion is a possibility. A low initial count may necessitate early platelet transfusion. Platelet counts of less than $50,000/\text{mm}^3$ increase clinical bleeding and should be corrected. In practice, if platelets are monitored during a continuing massive replacement, platelets should be ordered as the count decreases below $100,000/\text{mm}^3$. Infusion of 5–10 mL/kg of platelet concentrate increases the platelet count by $50,000\text{--}100,000/\text{mm}^3$. Platelets must be stored at room temperature, not refrigerated, and they should be rocked periodically. Other deficiencies that become apparent should be dealt with by appropriate therapy (e.g., fresh frozen plasma—10–15 mL/kg increases factor levels by 15–20%). Anticipate that FFP will be required in a volume of at least 1/3 of each blood volume lost after the first blood volume is replaced; FFP should not be needed if the blood loss does not exceed one blood volume.

Cryoprecipitate may be required if bleeding persists despite all other measures in small infants (1–2 units/kg increase fibrinogen by 60–100 mg/dL). Remember that FFP, cryoprecipitate, and platelet solutions contain more citrate per unit volume than does whole blood. Therefore, calcium infusions may be required if hypotension occurs as these products are given rapidly.

Alternatives to Blood Transfusion

The risk of infection through transfusion has prompted the search for alternatives, many of which may be applicable in children:

1. *Autologous transfusion of blood donated preoperatively:* Suitable size donations may be collected at 4- to 5-day intervals, preoperatively. If blood donation is combined with measures to increase erythropoiesis (oral iron and Vitamin C 6–8 weeks in advance, erythropoietin 3–4 weeks in advance), significant volumes may be collected. These techniques are most applicable to older children and teenagers.
2. *Blood conservation:* Blood losses are minimized through the use of proper positioning, infiltration of vasoconstrictors, induced moderate hypotension, and meticulous surgical technique.
3. *Acute normovolemic hemodilution:* Blood is withdrawn after anesthesia induction but before surgery commences, and each mL removed is replaced with 3 mL of warmed Lactated Ringer's solution. Hemodilution to a Hct of 25 % is acceptable in otherwise healthy children. Blood is then reinfused as the surgery proceeds, saving the first collected unit of blood to be transfused last. The volume to be collected can be calculated from the following formula for estimating the MABL.
4. *Intraoperative autotransfusion of shed blood using a “cell saver”:* This technique has limited application, but it may be useful in orthopedic surgery. Shed red

cells may be collected by suction, washed, and reinfused. However, coagulation factors are discarded in the washing process, and extensive reinfusion of washed cells may lead to dilution of these factors and coagulopathy.

Special Considerations for the Preterm Infant

1. *Infection:* The immune system is immature, and the preterm infant is particularly prone to infection. Use careful aseptic technique for all invasive procedures.
2. *Intraventricular hemorrhage:* The preterm infant is prone to intraventricular hemorrhage. Avoid causing fluctuations in blood pressure, ensure adequate anesthesia, avoid overtransfusion, infuse hypertonic solutions slowly (e.g., dextrose, sodium bicarbonate), and treat anemia and coagulopathy.
3. *Apneic spells:* These are common in preterm infants, who must be monitored closely at all times and especially during and after anesthesia. Risk factors for perioperative apnea include:
 - (a) Preterm infants of less than 60 weeks postconceptional age are more likely to experience significant episodes of apnea than term infants; those less than 34 weeks gestational age at birth are at greater risk.
 - (b) Anemia (Hct <30%).
 - (c) A history of apnea episodes and use of an apnea monitor indicate greater risk.
 - (d) The presence of chronic lung disease increases the risk of apnea.
 - (e) The presence of any comorbidity.
 - (f) Parental cigarette smoking.

In very small infants, the risk of apnea may extend for as long as 72 h into the postoperative period. The following are general recommendations:

- (a) It is common practice to admit and monitor all preterm infants of less than 60 weeks postconceptional age after any general or regional anesthesia (see p. 16).
- (b) Older infants < 30 days of age are most commonly admitted and observed; older infants are admitted if there is any evidence of ventilatory disturbance in the perioperative period.

N.B. Apnea may be less common after surgery performed under spinal analgesia, but it may still occur, so the child must still be admitted and monitored. Caffeine therapy (10 mg/kg IV administered slowly after induction) may prevent or reduce the incidence of apnea, but monitoring is still advised.

4. *Temperature control:* The preterm infant is extremely vulnerable to heat loss—even more so than the full-term neonate. The surface area is even larger relative to body mass, there are no insulating subcutaneous tissues, and the brown fat stores are deficient. Be especially alert to prevent heat loss at all times because these infants cannot generate much heat.
5. *Oxygenation:* This must be very carefully controlled to minimize the risk of retinopathy of prematurity from hyperoxia and increased mortality from hypoxia in premature infants. Inspired concentrations must be kept to the minimum that will allow safe conduct of general anesthesia. Monitor with a pulse oximeter and maintain the saturation between 90 and 95 %.
 - (a) Ascertain the FIO₂ required preoperatively that ensures satisfactory oxygenation. During nonthoracic surgery with controlled ventilation, continue with this FIO₂ and check saturation.
 - (b) Whenever N₂O is contraindicated, use an air–O₂ mixture to achieve the desired FIO₂ and saturation.
 - (c) During intrathoracic surgery it is often essential to increase the FIO₂; monitor saturation and limit the O₂ concentration as far as possible while avoiding the possibility of inducing hypoxemia.
6. *Hypoglycemia and hyperglycemia:* Preterm infants are prone to hypoglycemia. Blood sugar levels should be checked frequently, and hypoglycemia (less than 40 mg/dL) should be corrected by infusions of glucose with an infusion pump. The preterm infant is also subject to hyperglycemia, which is usually iatrogenic but may also be caused by poor insulin response and continued glycolysis. Hyperglycemia leads to glycosuria, osmotic diuresis, and dehydration and should be avoided by frequent blood sugar determinations and controlled intravenous glucose administration.
7. *Fluid administration:* Avoid overload by very carefully metering the infusion of intravenous fluids. Determine the total intravenous fluids given, including those given with drugs. Use small (1 mL) syringes to accurately measure small volumes of drugs. Syringe pumps and controlled infusion lines are essential.
8. *Benzyl alcohol:* This is used as a preservative in multi-dose vials of some medications and has been linked with kernicterus, intraventricular hemorrhage, and mortality in preterm infants. Preparations containing large concentrations of this substance should be avoided.
9. *Coagulation:* Ensure that vitamin K has been administered. The preterm infant is also subject to coagulopathy associated with shock and sepsis. Thrombocytopenia is common. Perform coagulation studies on all seriously ill preterm infants. Platelet concentrates, FFP, or exchange transfusion maybe required.

ANESTHESIA FOR OUTPATIENT SURGERY

Advantages

1. The child's psychological upset is minimized.
2. There is less risk of hospital-acquired infection.
3. Cost of care is reduced; hospital beds are available for others.

Selection of Cases

1. The child must be healthy or have any chronic disease under good control.
2. The child's parents must be reliable and willing to follow instructions concerning preoperative and postoperative care.
3. The operation should be associated with minor physiologic upset, requiring no complex postoperative care or pain management.
4. Infants who were born preterm and are still less than 60 weeks postconceptual age should be admitted and monitored for 12 apnea free hours.⁸

Preoperative Preparation

Preoperative preparation is the same as for inpatient surgery. The parents should be given written instructions concerning preoperative fasting and methods to prepare their child for a visit to the hospital. They should also be given a health questionnaire to complete and bring with them to facilitate obtaining a medical history for the child (Fig. 4.4).

On the day of the operation, the child is brought to the outpatient department surgical unit. Hb determination is only required if the child appears anemic or suffers from chronic disease. A sickle cell preparation is obtained if indicated (i.e., in a child who has not been previously tested or whose status is unknown). The parent's or legal guardian's consent for operation must be properly obtained. A preoperative pregnancy test is mandatory in many hospitals for females who have reached menarche.

The anesthesiologist makes a preoperative assessment by taking the history, examining the child, and noting laboratory data. This may be done on the day of surgery or a few days before at an anesthesia assessment clinic.

⁸This remains the recommended practice in the United States. A recent meta-analysis suggested that completely healthy preterm infants who are more than 46 weeks PCA be monitored postoperatively for 6 h apnea-free before discharge. However, until prospective studies confirm the safety of the latter strategy, we recommend 12 h of apnea-free monitoring.

Anesthetic Techniques and Tricks

Premedication is not routinely given to outpatients so as to not prolong recovery. Most properly prepared children for outpatient surgery, especially when accompanied by their parents, are not very upset, and premedication is not necessary. In the event that the child is apprehensive, premedication may be administered, as described earlier (see p. 88).

Use simple general anesthesia techniques that are likely to result in rapid recovery. When possible, a light general anesthetic may be supplemented with the appropriate regional analgesia technique. If the block is established before surgery begins, it provides analgesia during and after surgery while reducing anesthetic requirements and the need for opioids. Regional blocks placed before surgery may also reduce the total pain experienced after surgery (preemptive analgesia) and reduce the incidence of postoperative nausea and vomiting [PONV]. Use local infiltration by the surgeon or peripheral nerve blocks to avoid opioids. Do not give unnecessary drugs that might increase postoperative morbidity (if needed use only low dose opioids and use double PONV prophylaxis, e.g., dexamethasone 0.1 mg/kg plus ondansetron 0.1 mg/kg up to 4 mg). Larger or repeat doses of any opioid increase the incidence of PONV or delays discharge. Inhaled agents have been widely used for pediatric outpatients, sevoflurane being the agent of choice for brief procedures. Simple inhalation anesthesia for brief procedures in children is followed by relatively rapid and complete recovery.

Total intravenous anesthesia (TIVA) using propofol, remifentanil, and a short-acting muscle relaxant (when required) is probably the optimal intravenous technique for outpatients; the intraoperative course is smooth, airway complications rare, and postoperative morbidity is minimal. A suitable regimen for TIVA for children undergoing relatively short procedures would be the following:

Propofol: 2.5–3.5 mg/kg for induction, then infusion of 250 µg/kg/min for 10 min; 200 µg/kg/min for next 10 min and 150 µg/kg/min for remainder of operation. In addition, remifentanil-loading dose 0.5–2 µg/kg; infusion at 0.1–2 µg/kg/min

Intravenous acetaminophen (10 mg/kg IV) and ketorolac (0.5 mg/kg IV, up to 15 mg for children <50 kg and up to 30 mg for those >50 kg) for mild to moderate pain (given after conferring with the surgeon and after hemostasis is achieved).

Tracheal intubation should be used whenever indicated; postintubation complications can be prevented by gentle laryngoscopy and by using a tracheal tube that passes easily through the glottis and subglottic space. A small leak should be present when the circuit is pressurized to 20–30 cm H₂O. In many cases the LMA is a preferable alternative for the healthy outpatient. For dental surgery, a nasotracheal tube is used (see previous discussion). At the end of the procedure,

Questionnaire for Outpatients

Please complete this form and bring it with you. This will assist in the preoperative evaluation of your child.

Has your child ever been hospitalized before?	yes/no
Why/when? _____	
Was your child premature?	yes/no
Has your child ever had an anesthetic before?	yes/no
Any problems? _____	
Has any family member had problems with anesthesia?	yes/no
Details: _____	
Is there any family history of muscle, nerve or bleeding disorders?	yes/no
Details: _____	
Does your child take any medicines or herbal remedies?	yes/no
Please list: _____	
Has your child ever been treated with prednisone or similar medicines?	yes/no
Does your child have allergies?	yes/no
If yes, please circle or list the relevant items:	
i. Latex or toy balloons, band-aids, bananas, eggplant, other fruit.	
ii. Antibiotics or other medicines.	
Type of allergic reaction: _____	
iii. Other foods or household items: _____	
Is your child exposed to cigarette smoke at home?	yes/no
Has your child had a head cold recently?	yes/no
Has your child ever had asthma?	yes/no
Ever admitted to hospital with asthma?	
Does your child have loose teeth, dental plate, or dental appliance?	yes/no
Does your child bruise or bleed easily?	yes/no
Does your child have muscular dystrophy or other muscle disease?	yes/no
Any other medical issues we should know about your child's health?	yes/no
Details: _____	
 For Adolescents	
Do you smoke or take any drugs?	yes/no
Details: _____	
Do you have any ear-rings or other body piercings?	yes/no
Details: _____	
Has menstruation commenced?	yes/no
Date of last menstrual period _____	
Child's name: _____	
Parent's name: _____	
Date: _____	

Fig. 4.4 Questionnaire for outpatients

always perform a laryngoscopy, suction the pharynx well, and ensure that all throat packs and other items have been removed.

For strabismus surgery, a propofol infusion with oxygen/air is associated with less nausea and vomiting than when inhalational agents are used. Current antiemetic management for strabismus surgery when using inhaled agents includes ondansetron (0.05–0.1 mg/kg, maximum 4 mg) and dexamethasone (0.1 mg/kg, maximum 8 mg) IV.

Some procedures in older children may be performed under regional analgesia; when possible, this is ideal for the outpatient. For example, for superficial surgery on the limbs, an intravenous block may provide excellent results.

Postoperative Care for the Outpatient

Many children require no analgesics immediately after surgery, especially if a regional block has been performed (e.g., for hernia; see pp. 149 and 382), but beware of the “analgesic window,” which may occur later at home as the block wears off. Analgesics should be ordered in anticipation of pain and should be administered by the clock rather than waiting for pain to occur. Plan for adequate continuing analgesia and thoroughly instruct the parents on how to dose their child; provide written instructions. Acetaminophen alternating with ibuprofen every 3 h around the clock may negate the need for opioids, depending on the severity of the pain. Codeine is no longer recommended for use in children, especially in those at risk for OSA and who are obese, because of the unrecognized risk that they are ultrarapid metabolizers of the CYP450 2D6 isozyme, rapidly converting codeine to morphine. (*See Chap. 2 re: caution regarding codeine in adenotonsillectomy surgery*). More potent analgesics are seldom required for outpatients, but hydrocodone (lortab elixir), oxycodone, and tramadol are used increasingly. Although few complications have been reported, these opioids are subject to the same metabolic pathways as codeine; thus they should be used with caution and the dosing should be carefully prescribed (see Chap. 3).

Many children take and retain oral fluids well before discharge, but it is unwise to “push” oral fluids before the child is ready to drink; doing so increases the incidence of postoperative vomiting. A useful method of fluid administration is to offer Popsicles ad lib, especially to children who have undergone tonsillectomy or adenoidectomy (see Chap. 10).

Every child should be discharged from the post-anesthesia care unit (PACU) by the anesthesiologist or his/her designee; in most cases standard discharge criteria may be used. Infants may be taken home when they have fully recovered. Children should be tested for street fitness and should be able to walk out; if

dizzy or nauseated, they must stay longer. If the anesthesiologist determines that a child is unfit for discharge, overnight admission is recommended.

Children must be accompanied home by an adult, who preferably should not also be the driver of the vehicle. Warn the parents that their child must not ride a bicycle or engage in dangerous activities for 24 h. A brochure containing basic information and a follow-up service should be provided. Parents should be carefully instructed regarding the treatment of postoperative pain. As more complex procedures are being performed in the day surgery unit, the need for effective postoperative analgesia is increasing. Parents should be instructed of the need to administer analgesic drugs before pain becomes severe. They can be instructed in the use of visual analog scores or other means to assess their child's pain and provided with suitable charts to record this. Parents must be encouraged to seek advice from the hospital if problems develop during the postoperative period. A follow-up phone call should be made to the parents on the evening of surgery.

Complications After Pediatric Outpatient Surgery

Complications are rare after pediatric outpatient surgery, with fewer than 1 % of children requiring overnight admission after a planned outpatient procedure. The most common reasons for admission are PONV or a surgical complication. Nausea and vomiting are a predictable consequence of some types of surgery (i.e., correction of strabismus, tonsillectomy). In such cases the choice of anesthesia techniques (i.e., propofol) or the preemptive administration of effective antiemetic drugs is recommended.

Complications that may occur at home include vomiting, cough, sleepiness, sore throat, and hoarseness. If the parents are well prepared, these can usually be treated effectively in the home.

Suggested Reading

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Regional Analgesia Techniques

LOCAL ANESTHETIC DRUGS

Local analgesic drugs are very widely used in children, particularly in the management of postoperative pain (Table 5.1).

Clinical Pharmacology

Local anesthetic drugs are amino esters (procaine, chloroprocaine, tetracaine) or amino amides (lidocaine, bupivacaine, ropivacaine) that interrupt nerve conduction by inhibiting the influx of sodium.

The pharmacokinetics in infants differs from those in older children and adults:

1. Absorption of the drugs is rapid, the cardiac output and regional tissue blood flows are greater, and the epidural space contains less fat tissue to buffer uptake. Drugs sprayed into the airway are very rapidly absorbed. The pK_a of local anesthetics is >7.4 ; hence, they exist in a quaternary, water-soluble form at physiologic pH.
2. The volume of distribution of the drug is larger. Plasma levels of local anesthetics injected into the epidural space in infants are less than in young children and adults; this greater volume of distribution also prolongs the elimination half-life.
3. The extent of protein binding in the neonate is less than in children because serum albumin and α_1 -acid glycoprotein levels are reduced in the neonate. Hence, the free fraction of local anesthetics is increased in neonates compared with children. Bilirubin binds to the acidic sites on albumin, but it does not interfere with the protein binding of bupivacaine or ropivacaine as these anesthetics only bind to the basic sites.
4. The rate of metabolism of local analgesic drugs is reduced in very young infants:
 - (a) Plasma cholinesterase activity is low in neonates, which may prolong the metabolism of the ester type of drugs.

Table 5.1 Maximum doses of local anesthetics^a

Local anesthetic	Dose (mg/kg)
Bupivacaine ^b	
Plain or with epinephrine	2.5
Ropivacaine ^b and levobupivacaine ^b	3.0
<i>Lidocaine^b</i>	
Without epinephrine	4.5
With epinephrine	7
<i>Mepivacaine^b or prilocaine^b</i>	
Without epinephrine	5
With epinephrine	7
Procaine	10
2-Chloroprocaine	10–20 (see p. 143)

^aMaximum dose may depend on site of injection

^bDoses for continuous infusions of amides should be reduced by 30 % in infants <6 months

Aarons CE, Fernandez MD, Willsey M, Peterson B, Key C, Fabregas J: Bier block regional anesthesia and casting for forearm fractures: safety in the pediatric emergency department setting. *J Pediatr Orthop* 34:45–49, 2014

- (b) The hepatic pathways (cytochrome P450) for conjugation of the amide local analgesics CYP450 3A4 and 1A2) are immature. The neonate has a reduced capacity to metabolize bupivacaine or ropivacaine; clearance at 1 month of age is only one-third of adult rates although, by 9 months, clearance reaches adult rates. This may lead to clinical problems in neonates during prolonged infusions of bupivacaine or ropivacaine (therefore limit infusions to 48 h at no more than 0.2 mg/kg/h). Older infants and children metabolize drugs more rapidly because of their relatively large liver size and maturation of the cytochrome pathways.
- 5. The metabolism of prilocaine results in methemoglobinemia. This may be more important in infants because of reduced levels of the enzyme methemoglobin reductase and may be significant if large areas of skin are covered with EMLA cream.

Local Anesthetic Drugs

Lidocaine

Lidocaine is commonly used for local infiltration. Total dose should not exceed 4.5 mg/kg of the plain solution or 7 mg/kg if epinephrine is added. Epinephrine prolongs the block and decreases the peak serum concentration by about 40 %.

Bupivacaine

Bupivacaine is a racemic mixture; the levo enantiomer is the clinically active form whereas the dextro is the more toxic form. Bupivacaine has been widely used for peripheral and epidural blocks. It has the disadvantage that overdosing or accidental intravascular injection may lead to severe myocardial depression that may be prolonged and difficult to reverse (use Intralipid see p. 144). Bupivacaine metabolism is reduced in small infants, thus requiring close attention to dose, particularly with infusions (see Clinical Pharmacology). The addition of epinephrine to bupivacaine is less effective in prolonging the block and decreasing peak serum concentrations than with lidocaine. However, epinephrine extends the duration of action of bupivacaine in infants and young children more than in older children.

Ropivacaine

Ropivacaine is also a levo enantiomer, which is less cardiotoxic than bupivacaine and produces an equal sensory block with a more rapid onset, less motor block, but similar duration of effect. Compared with bupivacaine, ropivacaine is less rapidly absorbed from the caudal epidural space, and peak plasma levels are less after an ilioinguinal nerve block. *Ropivacaine should not be used for penile or digital nerve blocks as vasoconstriction and ischemia could theoretically occur.* Epinephrine does not prolong the duration of epidural or other blocks with ropivacaine.

Chloroprocaine

This ester local anesthetic proffers two distinct advantages over other local anesthetics: a small risk of toxicity and an extremely short duration of action (plasma half-life is 20 s in adults and 40 s in neonates). With such a brief half-life, there is little risk of toxicity. It is metabolized by plasma cholinesterase. 2-Chloroprocaine contains EDTA as a preservative; back pain has not been reported in children. Some recommend maximum loading doses of 14–20 mg/kg for regional blocks although some practitioners exceed this dose in neonates and infants. For example, for caudal/epidural use, a loading dose of a 3 % solution of 2 mL/kg followed by a continuous infusion of 2 mL/kg/h in infants has been recommended for both upper and lower abdominal surgery. With its small risk of toxicity, caudal/epidural infusions of 3-chloroprocaine are particularly suited for neonates and infants.

Levobupivacaine

Levobupivacaine is the levo enantiomer of bupivacaine. It is less cardiotoxic, approximately 20 % more potent than bupivacaine, and may be more suitable for prolonged infusions.

Toxicity

Maximum doses of local anesthetics are recommended to prevent overdoses (see Table 5.1). N.B. Potential for toxicity is related to total dose and the uptake of drug from the injection site. The order of local anesthetic absorption with different blocks is easily remembered by the acronym “**ICE Blocks**”: I=intercostal >C=caudal, >E=epidural, >B=peripheral nerve blocks. Intravenous lidocaine in normal doses may produce toxic effects in children with CHD and right-to-left cardiac shunting because the normal first-pass absorption within the pulmonary circulation is bypassed. The dose should be reduced by ~50% in these children.

Compared with adults, neonates may exhibit signs of central nervous system toxicity (jitteriness and seizures) at smaller blood concentrations of local anesthetics. Local anesthetic blocks in children are most commonly performed during general anesthesia, which may mask signs of neurologic toxicity, but depending upon the anesthesia agent used might increase cardiac toxicity. If seizures occur, 100% oxygen should be administered, the airway secured, and medication to stop the seizure administered immediately: intravenous benzodiazepine, i.e., midazolam (0.05–0.2 mg/kg), thiopental (2–3 mg/kg), or propofol (1–2 mg/kg) in repeated doses as required.

N.B.[§] Acute intravascular (intravenous or intraosseous) injection of bupivacaine (and less likely ropivacaine or levobupivacaine) resulting in local anesthetic systemic toxicity (LAST) may cause persistent ventricular tachycardia resulting in a low cardiac output state. If asystole or pulseless electrical activity (PEA) occurs, then the standard PALS algorithm should be followed and a prolonged code can be anticipated. Consider cardiopulmonary bypass rescue. The most effective treatment in animals and anecdotal reports in humans suggest a role for intravenous Intralipid (20%) at 1.5 mL/kg loading dose followed by 1 mL/kg every 3–5 min (for 3 doses) and then an infusion of 0.25–0.5 mL/kg/min for myocardial dysfunction for at least 10 min. after restoring cardiovascular stability. The mechanism of rescue is believed to be elution of the local anesthetic from the myocardium by the lipid. N.B. Propofol should *not* be substituted for Intralipid if the latter is not available. The American Society of Regional Anesthesia also recommends avoiding vasopressin, calcium channel blockers, β blockers, or additional local anesthetic and reduce the dose of epinephrine to <1 μ g/kg for those with cardiac dysfunction but not asystole or PEA.

[§]<https://www.asra.com/content/documents/checklist-for-local-anesthetic-toxicity-treatment-1-18-12.pdf>

Adjvant Drugs

Epinephrine

Epinephrine is added to local anesthetics to extend their duration of action and limit absorption. It also acts as a marker for intravascular injection; peaked T waves and ST segment elevation are more reliable signs of intravascular injection than tachycardia. Epinephrine may interact with halothane and precipitate arrhythmias, but doses up to 10 µg/kg by infiltration are considered safe in children. Note that test dose EKG changes are unreliable in infants anesthetized with propofol and remifentanil and that an increase in blood pressure is a more sensitive marker of an intravascular injection.

Clonidine

Clonidine (1–2 µg/mL) may be added to local anesthetics for use in the caudal/epidural space. This will prolong the effect of the block approximately 3 h (based on a systematic review), but has inconsistent effects on the rate of elimination of the local analgesic from the epidural space.

Apnea has been reported after epidural clonidine administration in preterm infants although three toddlers who received a 100-fold overdose of clonidine experienced only prolonged postoperative sedation and no respiratory distress/apnea. Clonidine may also contribute to postoperative sedation (usually at doses >2 µg/kg), and this may be undesirable in outpatients.

REGIONAL ANALGESIA FOR PAIN CONTROL

Regional analgesia techniques alone are of limited value during pediatric surgical procedures; the overall nonacceptance and lack of cooperation in the awake young child results in the need for such large doses of sedatives that general anesthesia is preferable. However, in the management of postoperative pain, regional analgesic techniques have become an indispensable part of our pediatric anesthesia practice (see later discussion).

Regional analgesia may provide acceptable pain control intraoperatively for some selected older children. Rarely, regional blocks are also indicated for chronic pain therapy and/or diagnostic purposes.

1. Spinal and/or epidural anesthesia are useful for small infants, especially for the preterm infant (with or without lung disease) for herniorrhaphy repair, circumcision, or lower abdominal surgery. This is a means of reducing, but not eliminating the probability of postoperative apnea.

2. Epidural analgesia may be a suitable alternative to general anesthesia in some older children (e.g., those with cystic fibrosis), and it may then be continued into the postoperative period.
3. Some older children (5 years of age and older) can be “charmed” into cooperation and have their upper limb fractures reduced under a regional block.
4. Intravenous regional analgesia (Bier block) can be used for some older children having superficial surgery to lesions on the distal limbs or for some fracture reductions.
5. The possibility of using regional or local infiltration analgesia (see Table 5.1) should also be considered for any minor procedure in a high-risk patient, e.g., skeletal muscle biopsy in a child with cardiomyopathy, lymph node biopsy in a child with a mediastinal mass.

Basic rules for regional analgesia are as follows:

1. Calculate the allowable weight-based dose of the local analgesic agent for each child and do not exceed that dose.
2. Use as much of the allowable dose of agent as is necessary to ensure a good block.
3. Use careful aseptic technique; beware of intravascular injection. Test by aspirating frequently.
4. Plan ahead; allow a generous time period for the block to become well established before allowing the surgeon to approach the child.
5. Remember the special considerations for the use of local analgesic drugs in infants and young children (see previous discussion).
6. Always be prepared to deal with the complications of regional analgesia. Drugs and equipment to induce general anesthesia, secure the airway, and ventilate the lungs must be immediately available. Establish IV access before the block. A supply of Intraparalipid should be immediately available to treat an intravascular injection of local anesthetic (see Toxicity).
7. Be prepared; unsatisfactory regional analgesia may require administration of general anesthesia to permit completion of the surgical procedure.
8. If possible, apply topical anesthetic cream over the site of the proposed initial needle insertion point in advance (see p. 634).
9. Children are generally upset by paresthesias; techniques that do not rely on eliciting these effects are preferred (see below).
10. The use of ultrasound is very effective in accurately placing the local anesthetic solution, improving success rates, and reducing drug doses. This technique should be applied whenever possible.
11. Supplement your regional technique with age-appropriate sedation (e.g., oral midazolam), systemic analgesics (e.g., fentanyl intravenously), and/or distraction (e.g., video player or iPod and earphones).

12. Evidence supports performing a caudal or lumbar epidural block including passing an indwelling catheter in an anesthetized child. The risks of inserting the block under general anesthesia are considered less than the risks of inserting it in a distressed and mobile awake child.
13. When a choice exists, peripheral nerve blocks are generally safer than neuraxial blocks in infants and children.
14. Do not use epinephrine for blocks of areas with inadequate collateral blood supply (digits, penis).
15. Do not administer sedatives to former preterm infants who have a spinal or epidural block as this will markedly increase the potential for apnea.

AN OUTLINE OF PROCEDURES FOR REGIONAL BLOCKS IN INFANTS AND CHILDREN

For more detailed descriptions of the anatomic considerations and techniques of regional nerve blocks, the reader is referred to standard textbooks. (e.g., A Practice of Anesthesia for Infants and Children edited by CJ Coté, J Lerman, BJ Anderson, Elsevier 2013)

Spinal Anesthesia for Infants

In infants, awake spinal anesthesia is most commonly indicated for surgery at or below the umbilicus, but it has also been used for upper abdominal surgery in small infants with a history of respiratory disease. It avoids the necessity to intubate and ventilate the lungs and therefore reduces the risk of additional airway damage or ventilator dependence. Very little change in blood pressure after spinal block occurs in infants and children aged less than 6 years. Postoperative apnea of former preterm infants is less common after spinal anesthesia but may occur if supplemental sedation is given.

Special Considerations

1. The spinal cord may extend rostral as far as L3 in the infant <1 year (compared with L1–2 in the older child or adult); therefore perform the lumbar puncture at L4–5 or L5–S1.
2. The dural space extends to S3–4 in the neonate.
3. The volume of cerebrospinal fluid (CSF) in infants (4 mL/kg) is double the volume in adults (2 mL/kg).

Contraindications

1. Sepsis or infected lumbar puncture site
2. Coagulopathy
3. Lack of enthusiastic parental consent

Anesthetic Management**Preoperative**

1. The child should be fasted as for general anesthesia.
2. No premedication is necessary for small infants.

Perioperative

1. Observe all special precautions for infants, both term and preterm. Prepare the anesthesia machine, endotracheal tubes, and all ancillary equipment.
2. A sweetened soother is often useful to settle the infant. (Glucose may have analgesic properties of its own in neonates.) Sedatives such as ketamine or midazolam that supplement a spinal will cause a similar incidence of apnea as general anesthesia.
3. Establish a reliable intravenous infusion using local analgesia in the lower extremity.
4. Scrub, glove, and sterilize the skin.
5. Instruct your assistant to gently but firmly restrain the child in the chosen lateral or sitting position, but avoid neck flexion, which may compromise the airway.
6. Prepare and drape the child. Infiltrate the skin over the L4–5 interspace with 1% lidocaine.
7. Prepare a neonatal spinal needle (e.g., 22 or 25-gauge, 1 in. [26 mm]) and measure the dead space of this needle with a tuberculin syringe.
8. Prepare a 1 mL syringe containing hyperbaric spinal bupivacaine, 0.75% bupivacaine in 8.25% dextrose, at 0.1 mL/kg (or 0.75 mg/kg). Some use iso-baric bupivacaine 0.5% at 0.2 mL/kg (or 1 mg/kg).
9. Insert the needle at L4–5 with the bevel oriented laterally until CSF is obtained.
10. Slowly inject the local analgesic solution; rapid injection may result in a high or total spinal.
11. Carefully return the infant to the supine position and place the pulse oximeter and blood pressure cuff on the lower extremity. Motor function in the lower limbs usually ceases immediately. Do not allow the child's legs to be raised (e.g., to apply the cautery pad); log roll the child or an excessively high block may result.

12. Duration of anesthesia is usually about 1.5 h.
13. **N.B.** Total spinal anesthesia in infants is heralded by cessation of crying or apnea with little change in blood pressure and heart rate. Treat with controlled ventilation until recovery occurs.

Perioperative

1. Keep the child horizontal until motor function returns.
2. Monitor the former preterm infant for apnea; it is less common than after general anesthesia but may still occur.

Caudal Block

The caudal block is very useful in infants and children; it provides good postoperative analgesia after abdominal, lower limb, or perineal surgery. A caudal block may be an alternative to spinal anesthesia for lower abdominal surgery in infants. In young infants, the contents of the epidural space offer little resistance to the spread of local anesthetic solutions. Epidural blockade is accompanied by minimal changes in blood pressure or cardiac output in children <6 years. Continuous caudal catheters have been used intraoperatively for more prolonged surgery, and they may safely be threaded to a surprising distance cephalad (T6). Local infection has not been a problem when catheters have been left *in situ* for 3 days.

Caudal morphine provides analgesia for thoracic and abdominal procedures and reduces the need for systemic analgesic drugs. However, caudal opioids are associated with side effects that include nausea, vomiting, and rarely respiratory depression; hence, this regime is unsuitable for outpatients and for infants.

Preferred Technique

For postoperative analgesia, the block should be performed after general anesthesia has been induced but before the surgery commences. This allows for the block to become well established during surgery, offers the potential for pre-emptive analgesia, permits a reduction in general anesthetic agents, and allows a more rapid awakening.

The child is placed in the lateral decubitus position with the knees and hips well flexed. The landmarks are then identified (Fig. 5.1): the tip of the coccyx to fix the midline and the sacral cornua bounding the sacral hiatus. These lie at the apex of an inverted equilateral triangle, the base of which is a line drawn between the posterior superior iliac spines. The child is prepared and draped, and the operator wears sterile gloves and a mask. The skin over the sacral hiatus is nicked with an 18-gauge needle (to avoid tracking epidermal tissues into the caudal canal), after

Fig. 5.1 Caudal block:
landmarks

Posterior superior
iliac spine

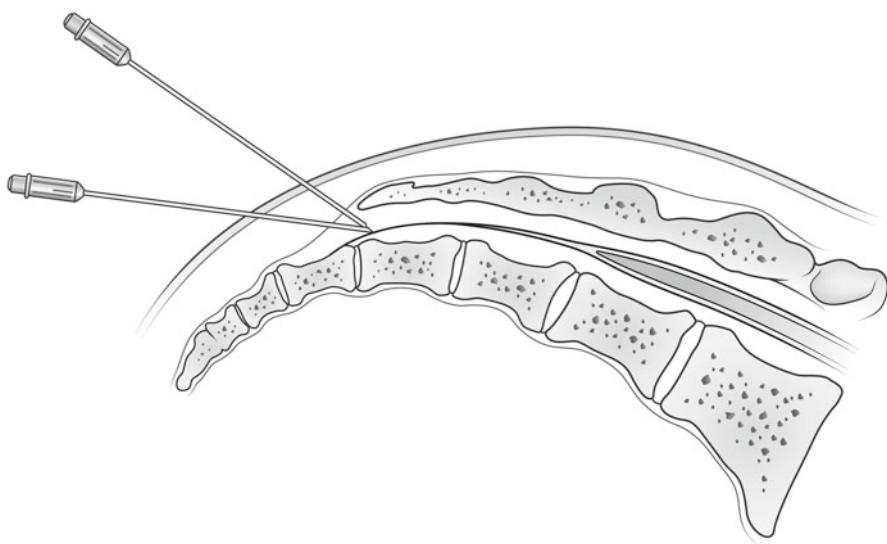
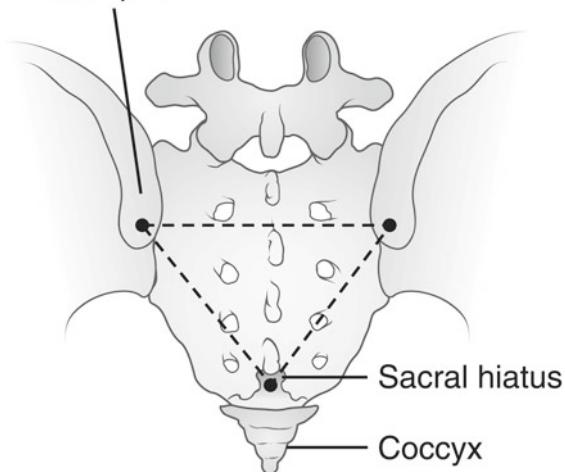


Fig. 5.2 Caudal block: direction of needle insertion

which an IV catheter (22 gauge for children <2 years, 20 gauge for those >2 years) is advanced cephalad at an angle of 45° to the skin with the bevel facing anteriorly. A distinctive sudden “give” is felt as the needle passes through the sacrococcygeal ligament. At this point, the angle of the needle is reduced and the catheter is advanced off the needle into the caudal canal (Fig. 5.2). The needle is then withdrawn, leaving the intravenous catheter in the caudal/epidural space. The catheter

should be observed for passive reflux of blood or CSF. If there is no evidence of blood or CSF, the local anesthetic is injected in incremental doses (there should be no resistance to injection; if there is resistance, then the catheter is either kinked or misplaced) while the electrocardiogram is observed. ST segment elevation and/or peaked T waves may be an early sign of intravascular injection during inhalational anesthesia; an increase in blood pressure is the most reliable change during TIVA. A finger should be placed over the sacrum to detect inadvertent subcutaneous injection. Advancing an intravenous catheter rather than a needle diminishes the risk of an intravascular or intraosseous injection. The use of ultrasound has been advocated by some to improve success rates.

Drugs, Concentrations, and Volumes

For single-shot caudal analgesia in outpatients, 0.125–0.175 % bupivacaine with epinephrine provides analgesia as effective as that of 0.25 % bupivacaine with less motor block. (A 0.175 % solution is prepared by combining 6 mL of preservative-free saline with 14 mL of 0.25 % bupivacaine with epinephrine (100 µg) [total volume 20 mL]. A 0.125 % solution is prepared with 10 mL of 0.25 % bupivacaine and 10 mL preservative-free saline with 100 µg epinephrine; this volume can be separated into two 10-mL syringes for ease of administration). Ropivacaine 0.2 % appears to provide similar analgesia and duration of action to bupivacaine and may be less cardiotoxic.

The following are volumes commonly injected via the caudal route:

1 mL/kg of 0.2 % ropivacaine is effective following lower abdominal surgery. 0.5 mL/kg provides good analgesia following genital surgery, e.g., hypospadias repair.

The addition of clonidine (1–2 µg/mL) to bupivacaine (where 1 mL/kg local anesthetic solution is instilled) has been reported in a systematic review to extend its duration of action approximately 3 h. Other drugs that have been used to prolong caudal analgesia include preservative-free midazolam and preservative-free ketamine; however, approved preparations are not currently available in the United States.

Caudal Morphine. Caudal morphine in a dose of 30 µg/kg, diluted in preservative-free saline to a volume of 0.5 mL/kg and administered as a single shot preoperatively, provides analgesia for up to 12 h and reduces the need for other analgesic drugs. Evidence suggests that maximum analgesia (and duration) is achieved with 30 µg/kg caudal morphine and that greater doses only increase the complication rate. *The child must be monitored continuously as respiratory depression may occur remote from the injection time.* In older children, intrathecal morphine administered by the surgeon (2–5 µg/kg) has provided excellent analgesia in those undergoing spine fusion.

Awake Single-Shot Caudal Analgesia for Lower Abdominal Surgery in Small Infants

As an alternative to awake spinal analgesia for lower abdominal surgery, bupivacaine 0.75 mL/kg of a 0.375% or 1 mL/kg of a 0.25% solution produces very effective caudal analgesia plus a motor block that lasts up to 90 min for surgery in awake infants. The dose of bupivacaine given when this technique is used may reach 3.125 mg/kg (to prevent leg movement), but has been considered acceptable because it is a single dose, the volume of distribution of the drug in the very small infant is large, and it has proven safe in practice. Alternatively, an equal volume of 0.2% ropivacaine might have a better safety profile while producing equal duration of anesthesia and motor blockade. A sugar-coated soother may help to calm the infant during the surgery; if sedation is administered to supplement the caudal block in a former preterm infant, the incidence of apnea may be as great as after general anesthesia.

Continuous Caudal Analgesia

In neonates, infants, and children up to 5 years of age, prolonged intraoperative and postoperative analgesia can be provided by means of a continuous caudal block. With the use of careful aseptic precautions, a 20-gauge epidural catheter may be threaded through an 18-gauge IV cannula and advanced to the desired level. In most infants, it is possible to pass the catheter to the thoracic level. If resistance is felt and is not relieved by rolling the catheter or slight flexion or extension of the child's spine, no attempt should be made to advance the catheter further. Threading the catheter to the lumbar and thoracic levels may not always be possible (particularly with catheters smaller than 20 gauge). However, the use of a nerve stimulator connected to a wire styletted catheter allows observation of muscle contraction as the catheter is advanced (<0.6 mA stimulation). Once the catheter is placed, tincture of benzoin is applied to the skin around the puncture site and a transparent occlusive dressing applied that should be regularly inspected. A second barrier drape is applied with benzoin to isolate the rectal area from the catheter insertion site. Some practitioners will obtain an epidurogram to assure correct positioning.

Although the risk of fecal contamination is very small, some tunnel the catheter subcutaneously to further reduce the infection risk. This is achieved by inserting the Tuohy needle into the skin several centimeters lateral and cephalad to the catheter insertion site and tunneling it subcutaneously such that the tip of the needle emerges at the catheter insertion site. Care must be taken to avoid severing the catheter with the tip of the Tuohy needle as it passes through the catheter site. The catheter is then passed retrograde through the tip of the Tuohy needle emerging from the upper end of the needle. The Tuohy is removed, both sites are covered and the catheter is secured.

Continuous caudal (and epidural) infusions of local anesthetics require ongoing vigilance. Trained floor nurses experienced in managing infants and children with caudal/epidural catheters are essential. If this is not available, the child should be monitored in an ICU or similar setting. The addition of opioids to a caudal/lumbar epidural infusion increases the risk of perioperative apnea as well as the risk of urinary retention, pruritus, and vomiting. Morphine may potentially spread more rostrally than fentanyl and may pose a slightly greater risk for apnea particularly in infants with multisystem diseases, respiratory compromise, or those who are neurologically impaired.

Neonates and infants <1 year metabolize bupivacaine and ropivacaine much more slowly than adults; hence, a much smaller infusion rate is needed to preclude toxicity (maximum infusion rate is 0.2 mg/kg/h) (see Table 7.5). Be very cautious if the infusion is continued more than 48 h in this age group.

Some centers use continuous infusions of lidocaine for continuous epidural analgesia. This technique has the advantage that blood levels of lidocaine can easily be measured thus avoiding toxic levels.

Lumbar and Thoracic Epidural Blocks

Lumbar epidural block has been widely used for postoperative pain relief in children and occasionally for surgery in older patients with special indications. The technique used is similar to that for adults; the “loss of resistance method” using saline is recommended to identify the epidural space. Air should not be used to test for loss of resistance because an intravascular air embolism may occur.

Thoracic epidural block may be performed in children using a technique similar to that used in adults. A midline approach is preferred, and the needle must be advanced at an angle determined by the configuration of the vertebral spine at the level selected. (Examination of the lateral chest radiograph is helpful before performing this block.) Thoracic epidural catheters are generally used in teenagers who may be sedated but not anesthetized during catheter placement so that paresthesia will be identified.

Special Considerations

In children who weigh more than 10 kg, the distance from skin to lumbar epidural space in millimeters is approximately numerically similar to the child’s weight in kilograms (i.e., the distance in a 20-kg child is about 20 mm).

A 19-gauge (5 cm) Tuohy needle and a 21-gauge catheter are usually used in children younger than 5 years of age. In older children, an 18-gauge Tuohy needle and a 20-gauge epidural catheter are used. Ultrasound guidance may improve success rate and reduce the risk of dural puncture and the number of needle passes.

Initial Volume to Be Injected

For a lumbar epidural block, bupivacaine 0.125% with 1:200,000 epinephrine or 0.2% ropivacaine is used in a dose of 1 mL/kg up to 20 mL, supplemented with a dose of 0.2 mL/kg to achieve the level of block required. For a thoracic epidural, a loading dose of 0.05 mL/kg × the number of dermatomes to be blocked appears to be effective.

Continuous Infusion Epidural Analgesia

Solutions of 0.05–0.125% bupivacaine or ropivacaine are widely used to which fentanyl (0–3 µg/mL), morphine (5–10 µg/mL), hydromorphone (3–7 µg/mL), or clonidine (0.5–1 µg/mL) may be added (see Table 7.5, p. 223). The maximum bupivacaine or ropivacaine infusion rate is 0.2 mg/kg/h in neonates and young infants (< 6 months of age) and 0.4 mg/kg/h for infants >6 months and children for up to 48 hours.

Beware of toxic effects of local analgesics when children are receiving continuous infusions, especially when the duration of the infusion exceeds 48 h. Recognize that children may not report the early symptoms of tinnitus, light-headedness, and visual effects. Children may find common side effects such as numbness (pins and needles) or weakness in their legs upsetting and disturbing but should be reassured. The concentration of local anesthetic may be reduced to attenuate the side effects. Some practitioners feel that intermittent “top-up” doses may be safer than continuous infusions in young children. Patient-controlled epidural analgesia (PCEA) has been effective in children as young as 5 years with both bupivacaine (0.1% with fentanyl 5 µg/mL) and ropivacaine (0.2%). The optimal dosing for PCEA in children has not been determined (see Suggested Reading). Unfortunately, urinary retention, pruritus, and vomiting may be more common after epidural opioids in children than in adults. Vomiting may be treated with round-the-clock ondansetron and pruritus treated with a background infusion of naloxone (0.25–1 µg/kg/h) or nalbuphine (0.025 mg/kg q6h). Urinary retention can be relieved with a urine catheter but may also respond to reduced opioid concentration and/or a naloxone infusion at the same rate as for treating pruritus. *Note: Be aware that respiratory depression is always possible and is generally heralded by a slowing of respirations; therefore, rescue orders for naloxone and narrow parameters for monitoring respiratory rate are essential.*

Intercostal Nerve Blocks

Intercostal nerve blocks may be performed to relieve pain after thoracotomy or some upper abdominal procedures.

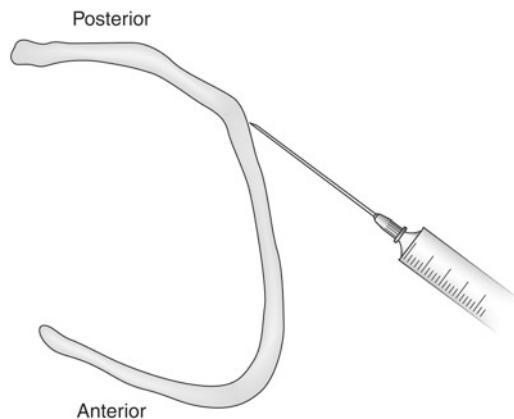
Special Considerations

1. Systemic absorption of local analgesics is the most rapid of any block from the very vascular intercostal space with a commensurate risk of toxic effects; take care not to exceed a total dose of 2 mg/kg of bupivacaine or ropivacaine. To limit this absorption, epinephrine (1:200,000) in the local anesthetic is recommended.
2. The risk of pneumothorax is substantial, especially in small children, in whom the distance from nerve to pleura is very small.
3. The intercostal nerves are sheathed in a dural layer posteriorly; injection near their origin can result in a total spinal block.

Preferred Technique

In infants and small children, the nerve in the intercostal space is approached by angling the needle posteromedially, almost parallel to the rib, rather than at right angles to it (Fig. 5.3). Palpate the lower margin of the rib, pull the skin upward, and advance until contacting the rib. The skin is released and the needle “walked” off the rib margin. The chosen volume (1–5 mL/rib 0.25% bupivacaine or 0.2% ropivacaine with epinephrine 1:200,000 not to exceed a total dose of 2 mg/kg) is then injected with only a 1–3 mm advancement and frequent aspiration for blood.

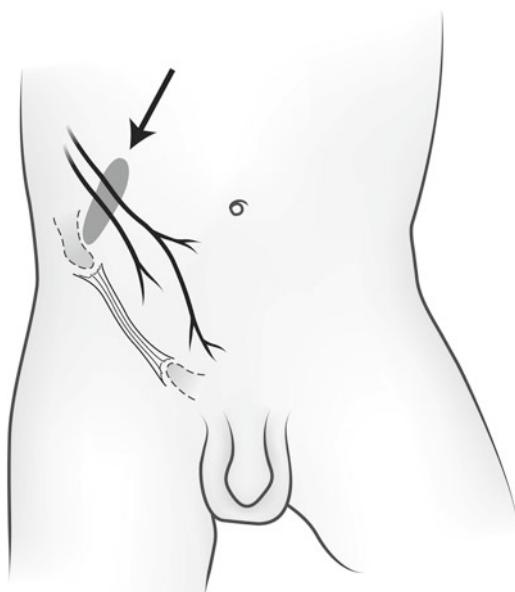
Fig. 5.3 Intercostal nerve block: direction of needle insertion in relation to rib



Ilioinguinal and Iliohypogastric Nerve Block

The ilioinguinal and iliohypogastric nerve block provides skin analgesia over the inguinal region providing postoperative analgesia after herniotomy. The block is preferably performed immediately after induction of general anesthesia and before the operation commences; alternatively, it can be placed by the surgeon following repair. The nerves run beneath the internal oblique muscle just medial

Fig. 5.4 Ilioinguinal-iliohypogastric nerve block:
area to be infiltrated with
local anesthetic solution



to the anterior superior iliac spine and may be blocked by a fan-shaped infiltration of the abdominal wall in this region (Fig. 5.4). Ultrasound guidance improves success, decreases the volume of drug required, and reduces the potential for intraabdominal puncture. Bupivacaine or ropivacaine 0.2–0.5% (up to 2 mg/kg) with epinephrine (1:200,000) may be used; more complete analgesia is obtained with the 0.5% solution, but occasionally this concentration produces a transient femoral nerve block with leg weakness.

Penile Block

The paired dorsal nerves pass inferior to the pubic bones on either side of the midline and supply the dorsal aspect of the penis and foreskin. A block of these nerves provides good pain relief after circumcision but does not provide adequate analgesia after hypospadias repair. Epinephrine-containing solutions should not be used because vasoconstriction might result in damaging ischemia to the penis. Complications have been very rare after properly performed penile blocks.

Volume to Be Injected

Up to 2 mg/kg of 0.5% bupivacaine *without epinephrine* is injected to a maximum volume of 1 mL in the small infant or 6 mL in the large child.

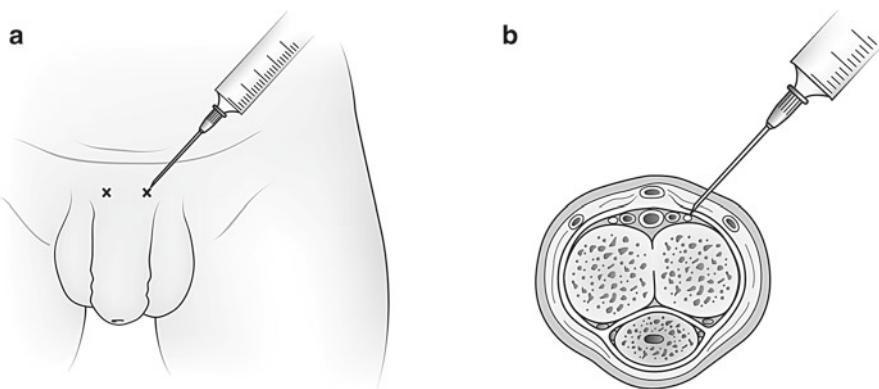
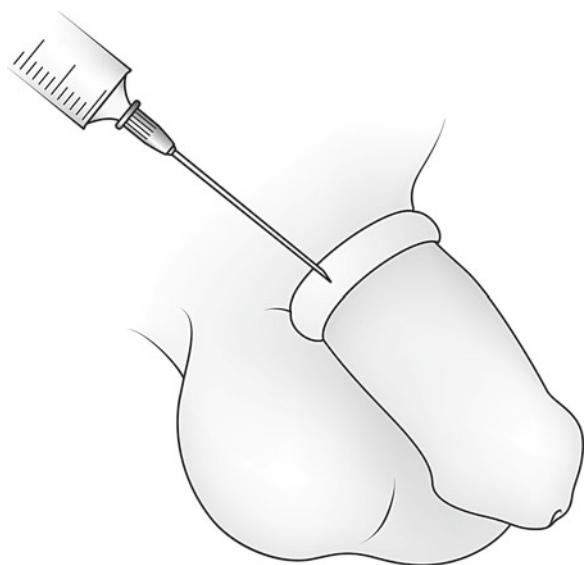


Fig. 5.5 Block of the dorsal nerves of the penis. (a) Injection sites. (b) Position of nerves at base of penis

Fig. 5.6 Ring block of the penis



Preferred Technique

With careful aseptic technique, bilateral injections are made beneath the pubis at the base of the penis at the 11 o'clock and 1 o'clock positions (Fig. 5.5). The needle should be felt to pass through Buck's fascia to deposit an equal volume adjacent to each nerve. In infants, it is possible to anesthetize both nerves by inserting the needle in the midline of the dorsal surface of the penis.

Alternatively, a ring block at the base of the penis may be performed (Fig. 5.6), at least one study suggests that epinephrine added to the local anesthetic is safe with a subcutaneous ring block at the base of the penis.

Head and Neck and Upper Limb Blocks

Supraorbital and Supratrochlear Nerve Blocks

These nerves are easy to block in children; the supraorbital nerve exits the supraorbital notch, which is readily palpated in children as one palpates the orbital rim from medial to lateral. The supratrochlear nerve exits the orbital rim several millimeters medial to the supraorbital notch.

Preferred Technique. For both blocks, a 27-gauge needle is positioned as described above and 1 mL bupivacaine 0.25% with epinephrine or 0.2% ropivacaine is injected. These blocks anesthetize the ipsilateral frontal region of the scalp for superficial surgery in this region (e.g., a frontal ventriculoperitoneal shunt).

Infraorbital Nerve Block

The infraorbital nerve block is also simple to perform and provides good analgesia following cleft lip repair or for children undergoing endoscopic sinus procedures. The infraorbital foramen can be palpated 1–1.5 cm below the infraorbital rim in line with the supraorbital notch and the pupil. With one technique a needle can be inserted at this point and 1–1.5 mL of local analgesic injected. However, in order to reduce the potential for a hematoma the preferred technique is subcutaneous through the mouth.

Preferred Technique. The upper lip is retracted and a long 25 or 27 g needle is advanced through the buccal mucosa and directed subcutaneously toward the infraorbital foramen. A finger is placed on the infraorbital foramen and the needle progress is palpated to reduce the risk of orbital puncture. After aspiration, 1 mL of 0.25% bupivacaine or 0.2% ropivacaine with epinephrine is injected.

Greater Occipital Nerve Block

A greater occipital nerve block may be performed by injecting 1–3 mL of 0.25% bupivacaine with epinephrine 1:200,000 or 0.2% ropivacaine as the nerves exit the skull next to the medial occipital artery at the level of the superior nuchal line. This will provide analgesia for superficial surgery involving the back of the head (e.g., a posterior ventricular peritoneal shunt).

Superficial Cervical Plexus Block

The nerves of the superficial cervical plexus may be readily blocked at the posterior aspect of the sternocleidomastoid muscle. A block of the greater auricular nerve and the lesser occipital nerve may be very useful in the child who has an incision behind the ear for mastoid surgery, tympanoplasty, or cochlear implant (Fig. 5.7).

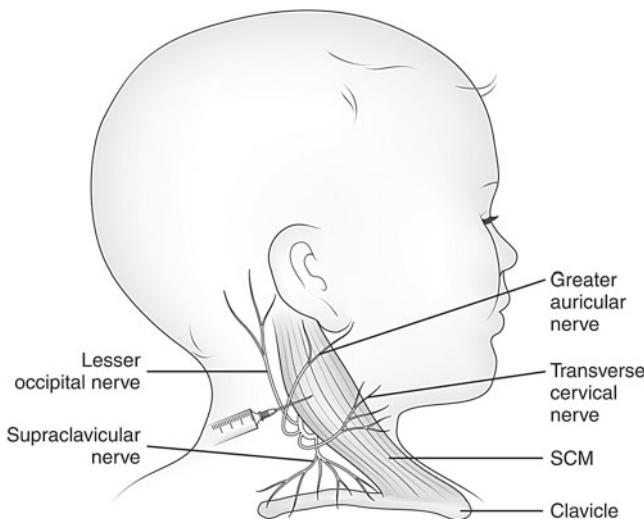


Fig. 5.7 Superficial cervical plexus block along the posterior aspect of the sternocleidomastoid muscle (SCM)

Brachial Plexus Blocks

There are several approaches to block the brachial plexus; the most commonly used are the axillary, supraclavicular, and infraclavicular routes.

Axillary Approach. The axillary approach is often recommended because of its simplicity and lack of serious complications (e.g., pneumothorax); it is easy to perform if the child can abduct the arm and is further simplified when possible by placing the hand behind the head. It is useful for forearm fractures, plastic surgery procedures, and insertion of shunts for dialysis, but the block does not include the area of the upper arm and sometimes not the area supplied by musculocutaneous nerves.

Preferred Technique. With careful asepsis, after skin analgesia, a short-beveled needle is advanced cephalad, at a 45° angle to skin, alongside and parallel to the axillary artery (Fig. 5.8). A slight “give” or “pop” should be felt as the neurovascular sheath is entered, and the arterial pulsations will be seen rocking the needle. With a nerve stimulator, a motor response should be detected in the ulnar, radial, or median nerve with 0.2 mA stimulation. The needle is supported gently, the plastic extension and syringe are connected to the needle, and after aspiration the drug is carefully injected with periodic aspiration. Pressure distally over the axillary artery may encourage proximal spread of the local analgesic solution and a more complete block.

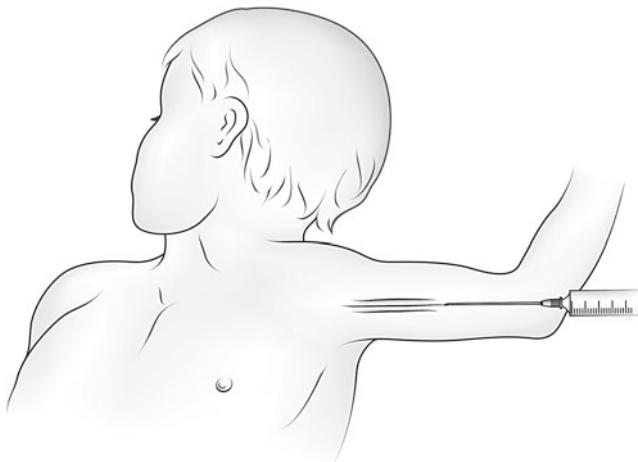


Fig. 5.8 Brachial plexus block, axillary route: direction of needle in relation to artery

Volume to Be Injected. Use 0.2–0.3 mL/kg of any local anesthetic (Table 5.1 for maximum doses); for older children, a maximum volume of 20 mL is usually satisfactory.

Lateral Infraclavicular Approach. This block may have advantages over the axillary block by providing more reliable analgesia in the musculocutaneous nerve distribution; however, pneumothorax is a potential complication. Ultrasound guidance is strongly encouraged to reduce this risk.

Preferred Technique. The block is performed inferior to the clavicle adjacent to the coracoid process of the scapula, which can readily be palpated in the child. The needle is inserted slightly caudal and slightly medial to the coracoid process and advanced vertically in a posterior direction. The use of ultrasound permits a high success rate, reduces the total volume of local anesthetic required, allows each of the three nerves to be individually blocked, and facilitates catheter insertion if a continuous brachial plexus block is indicated. A nerve stimulator may be used when ultrasound is not available in the anesthetized child to position the tip of the needle adjacent to the cords of the plexus. Wrist or forearm extensor movements (i.e., from posterior cord stimulation) when stimulated with 0.5 mA are predictive of success.

Volume to Be Injected. Local anesthetic 0.2–0.3 mL/kg may then be injected (Table 5.1 for maximum dose).

Supraclavicular Approach. This block can be performed in children for surgery involving the upper arm and forearm. With ultrasound guidance, there are few complications but if nerve stimulation is used, there is a risk of vertebral artery injection and pneumothorax.

Preferred Technique. The use of ultrasound is preferred. If stimulation is used, the plexus is located above the clavicle using a short bevel stimulating needle (1 mA). After piercing the skin, movement of any muscles of the upper extremity is accepted as correct placement and the local anesthetic is deposited.

Volume to Be Injected. Volumes of local anesthetic for this block are 0.15–0.2 mL/kg (Table 5.1 for maximum dose).

Lower Limb Blocks

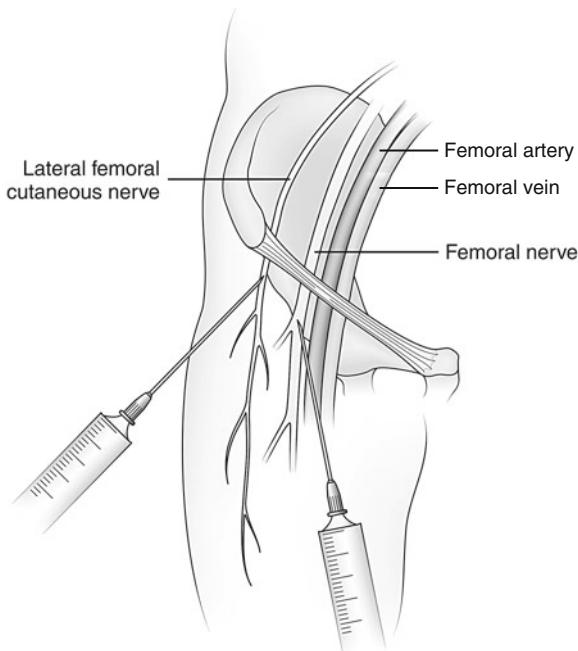
Femoral Nerve Block

The femoral nerve block is useful for fractures of the shaft of the femur and for muscle biopsy in patients with suspected myopathy (in which case it should be combined with a lateral femoral cutaneous nerve block).

For children with fracture of the femur, a continuous femoral nerve block may give very good continuing analgesia and also relieves the muscle spasms in the thigh.

Preferred Technique. The femoral artery is identified by palpation just below the inguinal ligament. After skin analgesia has been established, a 1-in., 22-gauge short bevel needle is advanced slowly just lateral to the artery at an angle of 45° to skin (Fig. 5.9). Slight resistance should be felt at two separate levels, the fascia

Fig. 5.9 Femoral nerve block and block of the lateral femoral cutaneous nerve of the thigh



lata and the fascia iliaca, after which the needle lies within the femoral sheath. The needle should then be gently supported, aspirated, and injected with periodic aspiration.

Volume to Be Injected. Use 0.2–0.3 mL/kg of local anesthetic with 1:200,000 epinephrine.

Continuous Femoral Nerve Block

With careful asepsis, after skin analgesia, a Tuohy needle with obturator is advanced, as outlined previously, through the two identified layers of resistance. The needle is positioned with the opening facing cephalad, and a standard epidural catheter is advanced proximally for 5–10 cm within the femoral sheath. When the Tuohy needle is correctly placed, the catheter should advance easily with little resistance. The Tuohy needle can then be withdrawn and the catheter left in situ, as when inserting an epidural catheter. Never attempt to withdraw the catheter through the needle because the tip may be sheared off and remain in the child. Ultrasound guidance will improve success rate.

Volume to Be Injected. Intermittent top-up doses of long acting local anesthetic or a continuous infusion may be used at a rate of 0.1 mL/kg/h.

Lateral Femoral Cutaneous Nerve Block

Block of the lateral femoral cutaneous nerve provides analgesia over the lateral aspect of the thigh.

Preferred Technique. The needle is inserted medial and inferior to the anterior superior iliac spine, just superior to the inguinal ligament, and advanced superiority and laterally until it impinges on the iliac bone (see Fig. 5.9). The local analgesic solution is injected as the needle is slowly withdrawn.

Volume to Be Injected. Use 2–5 mL of local anesthetic with 1:200,000 epinephrine injected in a fanlike manner.

Sciatic Nerve Block

The sciatic nerve in children may be blocked via an anterior, lateral, or posterior approach; the latter approach is considered to be the easiest and likely to be the most successful.

Preferred Technique. The child should be anesthetized and placed in a semi-prone position with the side of the nerve to be blocked elevated. The needle should be inserted at right angles to the skin at the midpoint of a line between the sacrococcygeal membrane and the greater trochanter of the femur, i.e., just lateral to the ischial tuberosity, until it is adjacent to the sciatic nerve). Confirm

by ultrasonography or by electrical stimulation (0.5 mA) via the needle. (In this case, a sheathed insulated needle¹ should be used).

If a catheter is needed for a continuous infusion, insert the needle just inferior to the gluteal fold and angle the needle slightly cephalad to approach the same site; the catheter will then be easier to secure and protect.

Volume to Be Injected. For a single dose, 0.2–0.3 mL/kg of local anesthetic with epinephrine 1:200,000. For a continuous block, use 0.1 mL/kg/h of local anesthetic.

Popliteal Fossa Block of the Sciatic Nerve

The sciatic nerve may also be blocked as it approaches the popliteal fossa. This block is useful to provide analgesia for foot surgery. Just proximal to the popliteal fossa, the nerve divides into the tibial nerve, which innervates the posterior calf and plantar area, and the peroneal nerve, which supplies sensation to the anterior leg. To be successful, the block must be performed above the division site; this is 3–7 cm above the popliteal crease in children under 8 years but slightly higher (within 3.5–8 cm) in older children. The approximate site to make the injection is 1 cm proximal to the popliteal crease for each 10 kg body weight; the needle should be angled at 45° to the skin surface pointing cephalad. The injection is made just lateral to the midline to avoid vascular structures. The block may be performed with the child in a lateral, supine, or prone position. Successful block can be achieved using a nerve stimulator (0.2–0.5 mA) to locate the main sciatic nerve or preferably by ultrasound. Continuous popliteal nerve blocks are becoming increasingly popular to replace caudal/epidural blocks for lower extremity surgery; a single dose of 5–10 mL of local anesthetic or a 20-gauge catheter is threaded 3–5 cm into the fossa and local anesthetic is infused at 0.1 mL/kg/h.

Ankle Block

Blocks of the branches of the sciatic nerve at the level of the ankle are very easy to perform and require only small doses of local anesthetics. An ankle block provides excellent analgesia following club foot repair. It is necessary to block the deep peroneal nerve alongside the anterior tibial artery, the saphenous nerve which lies alongside the saphenous vein, the posterior tibial nerve alongside the posterior tibial artery, and the sural nerve, which is just lateral to the Achilles tendon. Infiltration of the skin over the lateral aspect of the anterior tibia will block any branches of the superficial peroneal nerve supplying the dorsum of the foot.

¹ Stimuplex, Braun Medical, Bethlehem, PA.

Intravenous Regional Analgesia

The “Bier” (intravenous) block may be useful in older children having excision of lesions on either of the limbs (e.g., ganglion).

Preferred Technique

Insert a small intravenous cannula in the hand or foot. Use a reliable *double* pneumatic tourniquet. The success of the block varies with the degree of limb exsanguination, which can be achieved before injection of the local analgesic; use an elastic bandage, air splint, or lifting the extremity if possible. Inflate the proximal cuff (~200 mmHg) and inject the local analgesic solution. Do not exceed 5 mg/kg of 0.25–0.5% lidocaine (1 mL/kg). *Bupivacaine or ropivacaine should never be used for an intravenous block.* The addition of a very small dose of an intermediate acting muscle relaxant ($^{1/10}$ the usual intubating dose) will provide paralysis and perhaps improve surgical conditions by eliminating movements. When the block is established (~5 min), inflate the distal cuff and then deflate the proximal cuff. Do not release the remaining cuff until at least 30 min have elapsed, even if the operation is finished sooner.

This technique has been successfully used in children with forearm fractures requiring reduction and casting. The tourniquet was not released until the fiber-glass cast had been applied. Only a very small percentage of these patients experienced subsequent problems with a tight cast.

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Medical Conditions Influencing Anesthetic Management

UPPER RESPIRATORY TRACT INFECTION

Children often have or are recovering from a runny nose or other manifestations of an upper respiratory tract infection (URTI) when evaluated before general anesthesia. Indiscriminate cancellation is unwarranted and causes emotional and financial problems for children and their families; each case must be carefully evaluated. Some children seem to have a runny nose most of the time, possibly because of allergies or a chronic nasal infection; these children should be distinguished from those with an acute URTI.

Children with an uncomplicated URTI who are otherwise healthy have a greater incidence of intraoperative desaturation, bronchospasm, and laryngospasm. Although these complications can be disturbing, most are easily managed (see Laryngospasm, Chap. 4). Airway irritability after a URTI may last for 6–8 weeks, thus delaying the procedure 2 weeks will not reduce the incidence of these complications. Infants <1 year of age, infants who were born premature, children exposed to second-hand smoke, and children with reactive airway disease with a concomitant URTI may have more serious airway problems during and after anesthesia. Pulmonary complications in children having major (e.g., cardiac) surgery may be more common if there has been a recent (within 2 weeks) URTI. Anticholinergics do not affect the incidence of perioperative airway complications in children with URTI. Children with chronic rhinorrhea may receive 1–2 drops of either Neo-Synephrine (0.25 %) or oxymetazoline per nostril to dry up secretions during anesthesia.

Children with reactive airway disease (i.e., asthma) may benefit from their usual bronchodilators; bronchodilators and a vital capacity cough may resolve mild bronchospasm and airways plugged with mucus. Tracheal intubation is avoided if the airway can be managed with an LMA; this reduces the incidence of bronchospasm. However, evidence also suggests that the incidence of laryngospasm increases when LMAs are used in children with recent URTIs (where possible a face mask may be preferable).

**ALGORITHM FOR MANAGEMENT
OF UPPER RESPIRATORY TRACT INFECTION**

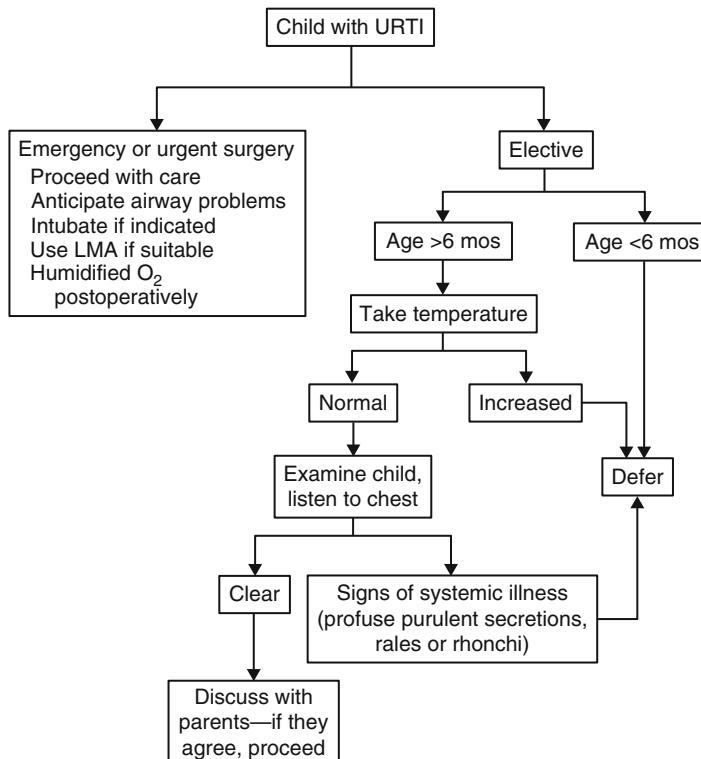


Fig. 6.1 Algorithm for management of upper respiratory tract infection (fever is $>38.5^{\circ}\text{C}$)

The following plan of action is suggested (Fig. 6.1):

1. Elective surgery
 - (a) The child should be carefully assessed. A history of the URTI and any other illnesses should be obtained. A careful physical examination should seek evidence of systemic illness, purulent nasal secretions, or lower respiratory tract disease.
 - (b) Children with a mild URTI but without pyrexia or any other evidence of disease are accepted for minor surgical procedures because evidence indicates little increased risk of complications. The decision to proceed should be discussed with the parents and the surgeon. Anesthesia via a face mask or laryngeal mask airway (LMA), avoidance of tracheal intubation when practical, and the use of sevoflurane may reduce airway complications.

- (c) Children who present for elective surgery with any one of the following should be rescheduled after 4 weeks:
 - Pyrexia greater than 38.5 °C
 - Change in behavior/diet/activity
 - Purulent secretions; or
 - Signs of lower respiratory tract involvement (e.g., wheezing) that do not clear with a forced cough.
 - (d) Children with a history of asthma and a URTI demand special attention; nonurgent surgery should be deferred for at least 4 weeks whenever possible.
2. Emergency or urgent surgery: Children who require emergency surgery cannot be deferred because of a URTI. The anesthetic prescription should account for the presence of a full stomach and the type of surgery but the presence of a URTI requires no special modifications. Sevoflurane is an acceptable anesthetic for children with mild URTI. Airway problems and laryngospasm must be anticipated, and all necessities for their treatment should be immediately at hand. When suitable, an LMA should be used as an alternative to tracheal intubation; it may result in fewer airway complications. Anticipate airway-related problems in PACU.

ASTHMA

Asthma affects 4–13 % of children in North America. The disease is characterized by variable cough, wheezing, and breathlessness and is episodic and seasonal in many. The symptoms result from bronchoconstriction, mucosal edema, and tenacious secretions in the small airways. Severe attacks may occur throughout childhood and may be life threatening. Acute exacerbations may be associated with URTIs, allergens, irritants, exercise, cold induced, or emotional stress. The treatment for mild asthma is inhaled β -adrenergics for bronchodilation and improved mucociliary clearance. For more severe asthma, inhaled corticosteroids may be added to control inflammation. Severe asthmatic attacks may require systemic corticosteroid therapy and ICU admission. The disease often improves as the child ages. Asthma severity is usually judged by the therapy required to control the symptoms; mild asthma is controlled by inhaled β -adrenergics with increasing severity suggested by the addition of inhaled corticosteroids. Rarely, oral steroids are prescribed but their use suggests a recent exacerbation.

When a child with asthma presents for elective surgery, it is most important to determine whether the child's current status is optimal. The following are important considerations:

1. If possible, surgery should be delayed for at least 1 month after the last acute attack; during this period, airway reactivity may be increased and residual mucosal edema and secretions may impair pulmonary function.

2. Elective surgery should be deferred if there is any evidence of an active URTI, which might precipitate and exacerbate symptoms.
3. The frequency of the asthma attacks, and the symptoms between attacks should be documented. Has admission to an intensive care unit or ventilator therapy ever been needed? If so, repeat severe attacks are likely.
4. What medications is the child taking, and are they controlling the child's symptoms? Have systemic corticosteroids been necessary? If so, in what dose? Has the child taken all the usual asthma medications today?
5. Physical examination is important to detect bronchospasm and exclude other current pulmonary pathology. If wheezing is present, the child should be encouraged to cough deeply. If the wheezing is unresolved, a trial of bronchodilator therapy should be administered. If the bronchospasm clears, then elective surgery that does not require intubation may proceed. If the signs do not resolve, then elective surgery should be deferred.
6. The results of recent pulmonary function tests and especially any response to bronchodilator therapy should be noted.
7. A chest radiograph should be ordered for any child with significant symptoms.

Anesthesia Management

Preoperative

1. Ensure that the child receives routine asthma medications up to the time of surgery. Oral medications can be taken with a sip of water; inhalation may be repeated, if necessary, just before transfer to the OR.
2. A stress dose of steroids (IV hydrocortisone [1–1.5 mg/kg]) should be administered at induction of anesthesia to those taking the equivalent of greater than 5 mg prednisone per day (adult dose), oral steroids within 3 months or high-dose inhaled steroids. A single dose of steroids is unlikely to do harm and will not precipitate a perioperative adrenal crisis.
3. Administer oral midazolam to calm the child if indicated. Atropine, if considered necessary, may be given intravenously at induction; it decreases secretions and causes some bronchodilation.

Perioperative

1. For intravenous induction, propofol is the preferred induction agent as thiopental may release histamine and possibly cause bronchoconstriction. Alternatively, ketamine may be used since it is a bronchodilator and may protect against bronchospasm; however, atropine should be given to limit secretions.
2. Inhalation induction with sevoflurane is preferred.

3. Avoid agents known to release histamine and cause bronchospasm (e.g., atracurium, morphine). Desflurane increases airway resistance in asthmatic children. Nitrous oxide, halothane, sevoflurane, fentanyl, rocuronium, and vecuronium are preferred drugs.
4. If tracheal intubation is necessary, it should be performed under deep anesthesia; otherwise, the procedure may trigger bronchospasm. Lidocaine (1.5 mg/kg IV) may decrease the potential for bronchospasm. If possible, avoid tracheal intubation for minor or brief procedures; an LMA is a useful alternative.
5. The anesthetic gases should be warmed and humidified—this is less essential if a circle circuit is used.
6. Intraoperative wheezing should be treated by deepening the level of anesthesia and by giving bronchodilator aerosols via the tracheal tube (e.g., albuterol). Be careful to exclude non-asthmatic causes for wheezing (e.g., a partially obstructed tracheal tube, endobronchial intubation, pneumothorax). Intraoperatively, albuterol may be delivered through an ETT by inserting its stem down into a 60 mL syringe (after the barrel is removed) and then reinserting the barrel. The syringe is screwed onto the capnography port of the elbow of the breathing circuit and the barrel compressed. Only 3–8 % of the aerosol delivered using this technique reaches the trachea the remainder layers out on the wall of the tracheal tube. To deliver most of the content of each puff to the tip of the tube, a long narrow gauge catheter may be inserted to the distal third of the tracheal tube. In this manner, the majority of the aerosol reaches the tracheal mucosa. Alternately a specially designed spacer with 15 and 22 mm connections can be placed at the ETT and puffs coordinated with inspirations.
7. At the end of surgery, antagonize the muscle relaxants as necessary. Neostigmine may increase bronchomotor tone, but this effect is counteracted by the atropine. Extubation is preferably performed with the child deeply anesthetized and breathing spontaneously so as to minimize the risk of precipitating bronchospasm. If awake extubation is planned, intravenous lidocaine or propofol may be administered beforehand to reduce laryngeal and bronchial reflexes.

Postoperative

1. Administer humidified oxygen.
2. When practical, regional analgesia is ideal for pain relief; for major surgery, continuous regional analgesia may be planned. Otherwise, use intravenous fentanyl or patient controlled analgesia (PCA) (see Chaps. 5 and 7).
3. Postoperative wheezing may require additional bronchodilator and appropriate adjustment of other medications.

CYSTIC FIBROSIS

Cystic fibrosis (CF) is an inherited disorder that results from a genetic defect on chromosome 9. Abnormal chloride and sodium transport result in increased electrolyte content and increased viscosity of secretions. This disorder affects many body systems and may present in the neonate as meconium ileus. During childhood, malabsorption due to pancreatic insufficiency predominates. In the second decade, increasing pulmonary problems occur because of abnormally viscous secretions and chronic infection. CF-induced diabetes is common in teenagers. Respiratory failure usually develops by the second or third decade of life secondary to retained secretions and chronic infection. Even if they appear fairly well, many of these children have severe pulmonary ventilation-perfusion (\dot{V}/\dot{Q}) inequality. CF patients are particularly susceptible to depression associated with long-term illness.

Surgery for children with CF is most commonly for nasal polypectomy (in many cases repeated), functional endoscopic sinus surgery (FESS), antral lavage, or bronchoscopy for removal of retained secretions and treatment of atelectasis. Children with advanced disease may present for lung transplantation; transplanted lungs do not appear to be affected by CF.

Special Anesthesia Problems

1. Copious, extremely viscous secretions are present in the respiratory tract.
Violent coughing and laryngospasm may occur.
2. Because of the \dot{V}/\dot{Q} disturbances:
 - (a) Hypoxia may develop rapidly.
 - (b) Inhalation induction is prolonged because of \dot{V}/\dot{Q} mismatch; this is exacerbated with less soluble anesthetics such as sevoflurane.
3. Lung compliance is reduced. In severe cases, high airway pressures may be required to adequately ventilate the lungs and prevent hypoxemia; a cuffed tracheal tube should be used.
4. Malnutrition and underweight for age is a result of malabsorption and chronic infection. Liver function may be reduced in older children. In such cases, drug dosages should be adjusted accordingly.
5. Many children with advanced CF suffer all the emotional upset of those with chronic disease; all of them require very careful and considerate handling and much reassurance.

Anesthesia Management

Preoperative

1. Assess the child's condition carefully; ensure that the pulmonary status is optimized before surgery by means of postural drainage, physiotherapy, and inhalation therapy. Commonly nebulized hypertonic saline to mobilize secretions and inhaled aminoglycosides to reduce infection are administered shortly before induction.
2. Pulmonary function is usually at its worst early in the morning. If possible, schedule the surgery at a time that allows for chest physiotherapy and clearing of secretions preoperatively.
3. Administer opioid premedication and midazolam judiciously as oxygenation may be compromised.
4. Ensure optimal hydration; fluids must not be withheld for prolonged periods. The child should be offered clear fluids until 2 h preoperatively.

Perioperative

N.B. Whenever possible, use local or regional anesthesia.

1. Place an IV for hydration and emergency drug administration.
2. Give 100 % O₂ by mask as tolerated in children with pulmonary involvement. Induce anesthesia with propofol, low-dose opioid, and a muscle relaxant. Be aware that inhalational induction in children with serious V/Q mismatch will be protracted.
3. Intubate with a cuffed tracheal tube.
4. Suction the trachea and remove secretions frequently using saline lavage.
5. Use humidified gases; children with severe CF may require 100 % O₂.
6. Minimize the use of long-acting medications and plan a technique that ensures rapid recovery.

Postoperative

1. Give IV fluids until the child is drinking.
2. Administer humidified oxygen and additional hypertonic saline nebulization. Whenever possible place the child in a semi-recumbent position that increases respiratory comfort.
3. Provide optimal analgesia. Use regional analgesia and NSAIDs for postoperative pain whenever possible.
4. Encourage early chest physiotherapy and ambulation.

LATEX ALLERGY

Latex allergy can cause a severe, life-threatening intraoperative immunoglobulin E-mediated anaphylactic reaction. Urticaria, bronchospasm, and/or circulatory collapse may occur. A history of repeated exposure to latex (e.g., frequent catheterization for neurogenic bladder, repeated surgery) and/or reactions to rubber balloons or dental dams may be elicited. Up to 40 % of children with spina bifida have latex allergy. The risk of developing allergy may be related to the number of surgeries (>5 surgeries) and the presence of atopy; hence, older children are more likely to have developed allergy. Cross-reactivity to bananas, avocado, kiwi fruit, and chestnuts may be found. Skin-prick testing or radioallergosorbent testing (RAST) to latex may confirm the allergy.

Latex allergy should be considered if signs of an anaphylactic reaction occur during surgery and cannot be related to drug administration. The initial signs usually develop 30–200 min after induction of anesthesia while the surgeon is handling large surface areas of mesentery or tissue; the signs include increased airway pressure, bronchospasm, decreased oxygen saturation, flushing and/or urticaria, hypotension, and tachycardia. One author has observed an urticaria the shape of a hand indicating the cutaneous contact with a latex glove.

The condition demands rapid, aggressive intervention including ventilation with oxygen, rapid infusion of warmed intravenous fluids to expand the blood volume, and epinephrine 1–5 µg/kg IV bolus followed by an infusion of 0.1–0.3 µg/kg/min. Other drugs that should be used include antihistamines (diphenhydramine, 1 mg/kg); histamine₂ (H₂) receptor-blocking drugs (cimetidine, 5–10 mg/kg); and corticosteroids (hydrocortisone 2 mg/kg). In some cases, a phenylephrine infusion may be required to maintain blood pressure.

Children at risk should be assessed for a history suggestive of latex allergy and skin tested if possible. Those who are positive may be candidates for desensitization therapy though the success of this has been variable. Some practitioners suggest pretreatment before surgery with corticosteroids (prednisone, 1 mg/kg) and H₁- and H₂-blocking drugs (diphenhydramine, 1 mg/kg; and cimetidine, 5–10 mg/kg); however, pretreatment does not prevent a reaction to latex. The most important method to avoid such reactions is to maintain a latex-free environment for those at risk for developing a latex allergy or with a known history of latex allergy. It is most important that all members of the surgical team use non-latex gloves and that all latex materials be excluded from the surgical and anesthesia equipment (Table 6.1).

In summary:

1. All parents and older children should be questioned about a history of lip swelling after touching toy balloons, tongue swelling after dental rubber dams in the mouth, and allergy to bananas, avocado, kiwi fruit, and chestnuts.

Table 6.1 Anesthesia equipment that may contain latex

Intravenous bag
Anesthesia circuit and bag
Airway or bite block
Plastic syringes
Vials
Intravenous bag
IV set (ports)
Catheters
Adhesive tape
Tourniquets
Face mask
Endotracheal tube
Anesthesia circuit and bag
Ventilator bellows
Blood pressure cuff
Gloves

2. Children in high-risk groups should be identified and offered skin tests.
3. All equipment should be clearly labeled as to its latex content.
4. All procedures on children who have a positive history or are at high risk for the development of latex allergy should be performed in a latex-free environment.

DOWN SYNDROME

Down syndrome (DS) (trisomy 21) is common (1.5 cases per 1000 live births). Developmental delay is invariably present but varies in severity; many DS children are quite cooperative and capable of relatively independent function in a protected environment.

Associated Conditions

1. Congenital heart disease (CHD) occurs in approximately 40–50 %, particularly atrioventricular canal, ventricular septal defect, patent ductus arteriosus, and tetralogy of Fallot. Pulmonary hypertension may accompany CHD and chronic airway obstruction.
2. Respiratory infections are common. This may be related to the genetic anomaly, an immune deficiency, and/or the social and institutional implications of the syndrome. Chronic sinus and ear infections are common.

3. Generalized hypotonia and joint laxity are present. Atlantoaxial joint instability occurs in 15% of afflicted children and may lead to cervical spinal cord injury. The neck is particularly unstable in the flexed position, but caution should be exercised to minimize excessive motion during intubation and positioning.
4. Congenital subglottic stenosis is common. Ensure that the tracheal tube is not too large and that there is an audible leak when positive pressure is applied.
5. Adenotonsillar hypertrophy is often present and obstructive sleep apnea (OSA) is common (see page 276).
6. Thyroid hypofunction is common as the child ages (15–20% of young children) and diabetes may develop.
7. Polycythemia is a frequent finding in neonates and may necessitate phlebotomy to relieve circulatory failure. Leukemia develops in 0.7% of children with DS.
8. Duodenal atresia is common in the neonate, as is umbilical hernia, renal malformations, undescended testis, and hypospadias.
9. Radial artery abnormalities (i.e., single median artery) are common and may make percutaneous cannulation difficult or impossible.

Special Anesthesia Problems

1. Airway: The large tongue and small nasopharynx predispose to respiratory obstruction, particularly during mask anesthesia and recovery stages. Have an oropharyngeal airway and LMA ready. Congenital subglottic stenosis predisposes to postoperative stridor.
2. Lungs: Is there any acute infection present that requires therapy before surgery? Is there a history suggestive of OSA?
3. Problems of associated cardiac disease: endocarditis prophylaxis may be required (see Chap. 14).
4. Atlantoaxial joint instability may predispose to injury during laryngoscopy and tracheal intubation. Inquire preoperatively for any signs or symptoms suggestive of C-spine compression (pain, gait disturbances, hand dysfunction, dizziness, bowel and bladder function). Gently examine the neck and perform a CNS examination. Routine neck X-rays are not warranted in the absence of signs or symptoms. Any excessive neck movement should be avoided (especially flexion). Position the head very carefully, and limit movement especially during ENT procedures.
5. Cognitively challenged children are more difficult to manage during induction of anesthesia; often the parents can be of great help.

N.B. Children with DS have been reported to be especially sensitive to the effects of atropine. This is not true; in practice, we have used the same dose for these as for other children without any problems.

OBESITY

The pandemic of childhood obesity impacts pediatric anesthesia in many ways. The diagnosis of obesity in children is more challenging than in adults because of the need to account for growth and development. Thus, accepted definitions for obesity in children are based on their growth curves (based on sex, weight, and height):

Overweight: BMI >85 percentile

Obesity: BMI >95 percentile

Super (morbid) obesity: BMI >99 percentile

Obesity negatively affects many organ systems although the complications from long-standing obesity are fewer than in adults. Respiratory problems include a restrictive respiratory pattern, an increased incidence of asthma and sleep apnea. Cardiovascular problems may include systemic hypertension, dyslipidemia, and left ventricular hypertrophy. Obesity leads to insulin resistance and type II diabetes mellitus. Other clinical implications include an increased incidence of gastroesophageal reflux, nonalcoholic steatohepatitis, and pseudotumor cerebri. Orthopedic and general surgeries are most common in this condition. Bariatric surgery is increasing in frequency with gastric sleeve surgery superseding earlier weight-loss surgeries.

All medical conditions should be investigated and optimized preoperatively. Obese children undergoing elective surgery are not at increased risk from pneumonia should aspiration occur; standard fasting intervals should be followed. However, obesity is an independent factor for difficult mask ventilation; airway obstruction necessitating an oropharyngeal airway may be necessary. Obese patients are also considered “difficult airways” because extensive fat deposits across their upper back effectively raise the larynx relative to the axis of the oropharynx. To optimize their airway, obese patients should be ramped at ~25–30° head-up and the head elevation doubled. In this position, the tragus is positioned at or above the sternal notch (lateral view), and the vocal cords should be in direct view during laryngoscopy. Basal atelectasis and oxygen desaturation occur commonly during brief apneas. The ramped position increases the functional residual capacity and the oxygen reserve, attenuating the severity of desaturation during tracheal intubation.

The pharmacokinetics of drugs depends on the drug’s hydro- and lipophilicity. To estimate the kinetics of drugs in obesity, it is instructive to consider the “fat mass” (FM) (as a compartment distinct from their “fat-free mass” (FFM)), as the compartment into which lipophilic drugs distribute, may be simply defined as:

$$FM = \text{total body weight (TBW)} - \text{the lean body weight (LBW)}$$

LBW is the ideal body weight (IBW) plus the added muscle mass to carry the FM or:

$$\text{LBW} = \text{IBW} + 0.3 (\text{TBW} - \text{IBW}) [\text{NB : Some use a factor of 0.4}]$$

IBW is the ideal weight for the sex, age, and height of the child. Several formulae may be used to estimate the IBW in children. One such example is:

For those <8 years: $\text{IBW} (\text{kg}) = 2 \times \text{Age} (\text{year}) + 9$

For those 8–12 years: $\text{IBW} (\text{kg}) = 3 \times \text{Age} (\text{year})$

The dosing of anesthetics in obese children based on these formulae still falls short of reliably predicting responses. Obese children have an increased cardiac output, increased extracellular fluid volume, increased LBW, and increased FM. The choice of which scalar (TBW, LBW, or IBW) best predicts the drug dose in obese children varies according with the drug. For example, the doses of propofol and thiopental (per kg) should use the LBW, non-depolarizing relaxants and morphine IBW, and succinylcholine boluses and propofol infusions TBW.

The uptake (and washout) of inhalational anesthetics in obesity vary with the anesthetic solubilities. In terms of the uptake, the greater FFM in obesity slows the washin of more soluble anesthetics. In terms of the washout, after intermediate duration surgery (<200 min) the washout is slowed because of the increased muscle mass and after prolonged surgery (>200 min), it is slowed because of the FM with soluble compared with less soluble anesthetics. Therefore, the use of less soluble anesthetics is preferred in obesity; the benefit of which is more apparent after surgery of greater duration.

Postoperative complications including respiratory issues (airway obstruction, desaturation), prolonged stay in PACU, and unplanned admissions are more common in obese children. Outcomes after cardiopulmonary resuscitation in obese children are worse.

MALIGNANT HYPERTHERMIA

Malignant hyperthermia (MH), a potentially fatal abnormal response to inhaled anesthetic agents and succinylcholine, is a pharmacogenetic disorder of skeletal muscles. All potent inhalational anesthetics including sevoflurane and desflurane, but not nitrous oxide, may trigger MH. It is characterized by a rapid increase in PetCO_2 , heart rate and respiratory rate, and the rapid onset of generalized muscle rigidity. Subsequently, body temperature increases. If untreated, profound biochemical changes will result.

MH is a rare condition, occurring in fewer than 1 in 100,000 cases in which general anesthetics are given, although the quoted incidence in children is 1 in 15,000. Children and young adults have most frequently been affected, and it has been reported in an infant as young as 2 months of age. Increased awareness of

MH and consistent operative monitoring of expired carbon dioxide has led to earlier diagnosis and prompt institution of treatment; the availability of a specific therapy (dantrolene) has reduced mortality from >70 to ~5 %.

The pathophysiology of MH is associated with altered calcium homeostasis in skeletal muscle. Malignant hyperthermia-susceptible (MHS) patients demonstrate an increased calcium ion concentration in skeletal muscle, and studies in MHS swine show a marked increase in the intracellular calcium concentration on exposure to triggering agents. This increased calcium level has been linked with a potential defect in the "calcium-induced calcium release" mechanism within the cell, now further linked with a defect in the calcium release channel of the sarcoplasmic reticulum. The mechanism of the disease process in the cell produces a sustained contracture of skeletal muscle, causing increased muscle metabolism with associated heat production. The further manifestations of the acute syndrome are secondary to the acceleration of metabolic processes within skeletal muscle, which is accompanied by large increases in oxygen consumption and CO₂ production. If the acute reaction persists, cellular energy substrates become depleted, with consequent failure of cellular functions, including those regulating the intracellular and extracellular chemical composition. Substances released by damaged cells (potassium, CK, myoglobin, etc.) trigger the continuing manifestations of the acute crisis, including coagulopathy and renal failure.

Detection of Susceptibility

There are still no simple, reliable screening tests to identify MHS individuals preoperatively. Creatine phosphokinase (CPK) levels may be increased, but this test is nonspecific. Many MHS children have local or generalized muscular disease; however, most with a muscle disease are not MHS. Although a positive family anesthetic history of MH is the most reliable clue, its absence does not guarantee an individual's non-susceptibility to triggering agents. Even uneventful previous anesthesia does not preclude an MH crisis during a subsequent anesthesia.

At present, MHS patients can be identified with reasonable certainty only by *in vitro* study of fresh living muscle tissue obtained at biopsy. Caffeine- and halothane-induced contracture of the biopsy specimen is usually diagnostic; this is presently the "gold standard." Up to the present time, a relatively large biopsy specimen has been required, so the test is not usually recommended for children younger than 10 years. Less invasive tests to determine MH susceptibility have been disappointing. MRI has been used to identify abnormalities of skeletal muscle energy substrates in MHS subjects, but a reliable, specific marker for the disease has not been identified. Genetic evaluation has shown some promise. Abnormalities of the ryanodine 1 gene on chromosome 19 have been

shown to be associated with MH susceptibility. If this test is positive, then no muscle biopsy is required; this genetic testing could be further used to define the proband's family tree of susceptibility. However if negative, then a biopsy is still needed. This test will be positive in ~30 % of susceptible patients but that means that it is negative in 70 %. Therefore, at the present time, there are no noninvasive methods to diagnose MH susceptibility with certainty.

Clinical Manifestations

Nonspecific, early signs include:

1. The hallmark of an evolving MH event is simultaneous and marked increases in expired carbon dioxide, tachypnea in non-paralyzed patients, and severe tachycardia. Other signs include sweating, desaturation, cyanosis, and overheating of the CO₂ absorbent. MH is most commonly triggered by the combination of inhalation agents and succinylcholine, each can trigger MH individually.
2. Hypertonus of skeletal muscles, particularly large muscle groups may be evident.
 - (a) It may occur immediately after the administration of succinylcholine. Failure of skeletal muscle to relax the so-called jaws of steel where one is unable to open the mouth for several minutes despite complete neuromuscular blockade. This is an indication to postpone surgery and reevaluate.
 - Tightness of large muscle groups such as upper arm, thigh, and calf muscles. This should arouse suspicion.
3. A rapid increase in body temperature (more than 1 °C) is a late sign.

The prognosis is much more favorable if the reaction is recognized early and dantrolene is administered before a significant increase in the body temperature occurs.

A venous blood gas obtained soon after the development of hypercarbia and tachycardia will generally demonstrate a pure respiratory acidosis (PvCO₂>80 mmHg) with a PvO₂<40 mmHg. If the reaction progresses prior to treatment, then severe metabolic acidosis (base deficit >25 mEq/L) and severe respiratory acidosis PvCO₂>80 mmHg will be present. *Venous desaturation is the hallmark of a hypermetabolic state and nearly pathognomonic of MH.* Early diagnosis and intervention with dantrolene will prevent the late manifestations of hyperkalemia, generalized muscle rigidity, ventricular arrhythmias, cyanosis, and increasing body temperature associated with a full-blown MH crisis.

Therapeutic Regimen¹

1. Discontinue all inhalational anesthetics and convert to TIVA; inform the surgeon of the diagnosis; insist that surgery be urgently terminated. **Send for help!**
2. Hyperventilate with 100% O₂ using a high flow. Either remove the patient from the anesthesia circuit and control respirations by an alternate means or change the circle absorber system and add Vapor-Clean agent filters to inspiratory and expiratory limbs of the circle circuit (Dynasthetics, Inc, Salt Lake City UT).
3. Immediately give the following:
 - (a) *Dantrolene*: The initial dose is 2.5 mg/kg, given as a rapid IV infusion. If necessary, repeat 2.5 mg/kg IV every 5 min (Max dose = 10 mg/kg but if in crisis and not responding to the dantrolene there would not appear to be harm in administering additional doses) until the end-tidal carbon dioxide decreases, the heart rate begins to slow and become regular, muscle tone decreases, and the child's fever abates. If tachycardia or tachypnea recur, then repeat IV dantrolene until clinical improvement is apparent. The sooner after the suspected diagnosis and initiation of dantrolene, the better the outcome. **N.B.:** The new injectable form of dantrolene (Ryanodex, Eagle Pharmaceuticals, Woodcliff Lake, NJ) is reconstituted with 5 mL of sterile water for a final concentration of 50 mg/mL and does not contain mannitol. Pediatric dosing may require use of a tuberculin syringe for infants.
 - (b) Dantrolene is a muscle relaxant so patients who require large doses may require respiratory support.
 - (c) *Sodium bicarbonate* (7.5%): 1–2 mEq/kg IV immediately; repeat in accordance with blood gas analyses to reverse the metabolic acidosis. Be aware that if alveolar ventilation is already maximized, bicarbonate may cause a transient respiratory acidosis.
 - (d) *Mannitol*: 0.5 g/kg; mannitol is no longer present in the dantrolene mixture but may be needed to maintain an adequate urine output (more than 1 mL/kg/h).
 - (e) **Do not use calcium channel blockers** as *calcium channel-blocking agents may interact with dantrolene to produce profound myocardial depression and their use has been associated with hyperkalemia and sudden death. They do not have a therapeutic role in MH.*
 - (f) Use standard drug therapy to treat dysrhythmias
 - (g) Use standard therapy to treat hyperkalemia

¹ Further advice can be obtained in North America from the MHAUS hotline telephone number: 209–634–4917.

4. If dantrolene is unavailable, commence active cooling. Symptomatic cooling should occur in cool/tepid water; iced temperature immersion may cause cutaneous vasoconstriction preventing heat loss. Place the child on a rubber sheet and use fans. Intragastric cooling and cold enemas may also be necessary. Infuse refrigerated saline solution intravenously at 10 mL/kg/h as necessary.
5. Continue monitoring the child closely:
 - (a) Monitor PetCO₂, oxygen saturation, and ECG.
 - (b) If the reaction is not aborted immediately after the first dose of dantrolene, an arterial line should be inserted to measure pressure and easily obtain blood samples.
 - (c) Insert a CVP to monitor volume status and venous oxygen extraction.
 - (d) Insert a urinary catheter and monitor urine output.
 - (e) Attach a multichannel thermometer (rectal, esophageal, skin, and muscle leads). Beware of overtreatment leading to hypothermia.
6. Obtain frequent arterial blood samples for the following studies:
 - (a) Blood gas and acid/base determinations (repeat every 10 min until stable)
 - (b) Serum electrolytes (Na, K, Cl, Ca, inorganic phosphate)
 - (c) CPK (obtain immediately at the onset of a reaction (normal value), peak concentration occurs at 12–18 h; a CPK > 10,000 IU is a presumptive sign of rhabdomyolysis and steps to prevent renal damage should be instituted (see below)).
 - (d) Coagulation studies
8. Correct any electrolyte imbalance on the basis of biochemical indices.
9. Maintain urine output at 1 mL/kg/h or greater; use mannitol or diuretics as necessary to prevent rhabdomyolysis-induced renal failure; alkalinize the urine.
10. In severe reactions, coagulopathy may develop and necessitate therapy on the basis of demonstrated factor deficiencies.
11. Avoid the administration of drugs that might complicate matters: calcium and adrenergic agents have previously been considered contraindicated. In fact, they may not cause further problems, but they are probably best avoided unless definitely indicated.
12. Monitor the child in the intensive care unit with the same aggressive monitoring as in the OR; MH may recur within the first 24–48 h. Sequelae of the episode may also demand aggressive therapy. Depending on the severity of the reaction, the airway may require ongoing intubation and hyperventilation. For example:

- (a) Recrudescence may occur in up to 25 % of those who have had MH reactions; IV dantrolene may be repeated as required to treat recrudescence. Various recommendations include dantrolene 1 mg/kg q4–6 h or an infusion of 0.25 mg/kg/h for the first 24 h after an event.
- (b) Cerebral edema may appear because of hypoxic insult.
- (c) Pulmonary edema may occur because of fluid overload or myocardial dysfunction.
- (d) Coagulopathy and renal failure may require continued care.

Anesthesia Regimen for Patients with Known Susceptibility to Malignant Hyperthermia

All personnel who may be concerned in the care of an MHS patient in the OR and PACU must be fully acquainted with a suitable protocol that describes the location of drugs and equipment and the procedures to be implemented if MH develops. Often the clinician must assume possible MH susceptibility on questionable evidence (i.e., a family history of “anesthetic difficulties” but no positive muscle biopsy in the family). The simple course then is to provide a trigger-free anesthetic.

Children at Risk

1. Survivors of an MH crisis
2. Children with a positive muscle biopsy
3. A first-degree relative of anyone known to be MHS (i.e., positive muscle biopsy or survivor of an MH crisis)
4. Those with muscle abnormalities and/or an increased serum CPK level, in whom MH may be suspected
5. Children with central core disease
6. Children with a “jaws of steel” response to succinylcholine (see further)

Preoperative Investigation

A preoperative investigation should be done for children with a positive or strongly suggestive family or personal history.

1. Review the family and personal anesthetic history carefully, noting especially muscle disease, and drug- or anesthesia-induced reactions. Deaths under anesthesia are particularly worrisome and require further exploration. There is no evidence for a relationship between mitochondrial disease or muscular

dystrophy and MH susceptibility although succinylcholine can induce rhabdomyolysis in children with the dystrophies.

2. An increased serum CPK may indicate an unrecognized muscle disorder but is not diagnostic.
3. If the history indicates a strong possibility of MH susceptibility and the child is older than 12 years of age, it may be advisable to arrange for a muscle biopsy at a suitably equipped center. Younger children and infants must be presumed to be at risk for MH and treated accordingly until they are old enough for testing (or until an improved, less invasive diagnostic test becomes available). A medic alert bracelet should be sought with any documented family history of MH

Preoperative Preparation

1. Oral midazolam may reduce preoperative anxiety.
2. Place topical local anesthetic cream on several possible IV sites.
3. Dantrolene pretreatment is not recommended for MHS children. However, dantrolene should be present in the OR prior to intravenous induction.
4. *If diagnostic muscle biopsy is to be performed, dantrolene must not be given (it affects the test results).*
5. Ensure that all necessary drugs and equipment have been prepared:
 - (a) Drugs for anesthesia—All intravenous induction agents, non-depolarizing relaxants, opioids, nitrous oxide, ketamine, atropine, and anticholinesterases are safe.
 - (b) Drugs for emergency use if MH develops, including dantrolene, refrigerated lactated Ringer's solution, normal saline, 7.5% sodium bicarbonate, warmed 20% mannitol and 50% glucose solutions, furosemide, soluble (Regular) insulin, and heparin.
 - (c) Equipment: The possibility of trace inhalation agent in the anesthesia machine and its components must be addressed. In general, changing the CO₂ absorbent, replacing the circuit with a clean disposable circuit, and flowing oxygen/air at 10 L/min for 20–40 min will cleanse most anesthesia machines but the success of this process is quite variable depending on the workstation. Inserting a charcoal filter (Dynasthetics, Inc, Salt Lake City UT) into the inspiratory and expiratory limb will reduce trace anesthetic concentrations to <5 ppm within several minutes and appear effective for approximately 1 h. Some recommend this process for ~10 min then placing new filters prior to induction. Appropriate temperature probes and perhaps a cooling blanket may also prove useful.
 - (d) If possible, remove all vaporizers from the machine or tape over the top so as to prevent accidental exposure; remove succinylcholine from the O.R.

Induction of Anesthesia

1. Induce and maintain anesthesia using only non-triggering agents; these include propofol, thiopental, nitrous oxide, opioids, benzodiazepines, non-depolarizing muscle relaxants, and local anesthetics.
2. For all major procedures, insert arterial and CVP lines and bladder catheter as indicated for the procedure (not simply because the patient is MH susceptible).
3. Closely observe the expired CO₂ level, heart rate, and respiratory rate [if breathing spontaneously]) for early signs of MH.

Postoperative (After Uneventful Anesthesia)

1. Transfer the patient to the PACU, with monitoring equipment and intravenous cannulas in place.
2. Ensure that *all* PACU staff are aware of the possibility of a delayed MH reaction and know what to do if one occurs.
3. Vital signs should be recorded at 5-min intervals initially.
4. Do not transfer the child to the ward or home until vital signs have been stable for 4 h and the results of any laboratory tests are satisfactory.
5. If the child is admitted vital signs are recorded hourly for 4 h and then every 4 h; assure that the nursing staff know that important increases in HR or respiratory rate should trigger a call for help.
6. Immediately administer dantrolene (2.5 mg/kg) if the child develops tachycardia and tachypnea unrelated to pain; once the reaction is controlled transfer to ICU for further monitoring and continued treatment.

Masseter Spasm

Masseter spasm is sometimes observed when succinylcholine is administered. It is now recognized that many children (~1%) respond with transient increased masseter tone, which generally lasts for a brief period of time (10–15 s); this is probably a normal response. However if the response is so severe that one is unable to open the mouth or the patient actively bites down ("jaws of steel") despite loss of response to a nerve stimulator then this may be associated with MH.

If masseter tetany occurs without other signs of MH, we recommend discontinuing MH-triggering agents (change to TIVA). We suggest taking blood for determination of serial CPKs and continued careful monitoring for signs of MH. Isolated masseter tetany after succinylcholine is very rare and must be considered a possible early sign of MH. Increased CPKs above 10,000 is suggestive of some degree of rhabdomyolysis and warrants admission and appropriate interventions to prevent renal injury (see above).

MUSCULAR DYSTROPHY

Pathophysiology

Duchenne's muscular dystrophy (DMD) is an X-linked recessive genetic defect in which a lack of dystrophin (<3% of normal) in the cytoskeleton (and increased intracellular calcium) of muscle renders this unstable and weak when stretched (e.g., by inhaled agents and succinylcholine) (see Appendix A). This disease primarily affects males. Skeletal muscle fragility becomes apparent between 2 and 6 years of age and continues to ~10 years of age. Thereafter, the significance of skeletal muscle instability wanes as myocardial involvement becomes predominant. "Pseudohypertrophy" of the calves is the result of fatty infiltration of muscles. Characteristic physical finding is Gower's sign. Acute skeletal muscle breakdown that may occur after succinylcholine and potent inhaled anesthetics, releases intracellular potassium, myoglobin, and CK, reaching >10,000 IU/L. Rhabdomyolysis may lead to sudden onset of cardiac arrest, myoglobinuria, and acute renal failure. Dystrophin also affects cardiac muscle; cardiac manifestations predominate (beyond age 10 years) with electrocardiographic (conduction defects) and echocardiographic changes (cardiomyopathy is present in 100% by 18 years). There is no treatment for DMD. See Appendix A for management.

Becker muscular dystrophy is a milder form of DMD that also affects primarily males but has a later onset, in the second decade. Dystrophin is reduced, not as severely as in DMD. Proximal muscle weakness is progressive but very slow. Cardiac involvement is rare in those <16 years of age.

Emery–Dreifuss muscular dystrophy is a disorder that affects the muscle proteins emerin or lamin A or C. Muscle weakness is usually apparent by 10 years of age and progresses slowly with age. However, cardiac conduction defects are predominant and syncope is common whereas cardiomyopathy is infrequent.

Management. Dystrophies are unrelated to MH; remove inhalational anesthetics to prevent rhabdomyolysis. For anesthetic management, see Duchenne's muscular dystrophy in Appendix A.

MITOCHONDRIAL MYOPATHY

Mitochondrial myopathies (MM) are a heterogeneous group of inborn errors of the respiratory chain (RC) in mitochondria that attenuate energy production. The RC, which comprises 5 enzymes (cytochrome oxidase), is under bigenomic control: (1) nuclear genomic control (most adult defects) yields 85% of the proteins and follows a Mendelian inheritance pattern; (2) mitochondrial genomic control (childhood defects) yields 15% of the proteins and follows maternal

lines of inheritance. Each cell contains thousands of mitochondria each requiring about 100 genes to generate energy. The variable presentation (phenotype) of MM depends on the ratio of mutated to normal genes in the mitochondria in each organ throughout the body. In infancy, the signs may be nonspecific, e.g., feeding difficulties, hypotonia, involving extraocular and skeletal muscles, and abnormal laboratory tests, e.g., lactic acidosis. Other organs including brain, heart, eyes, endocrine glands, nerves, and others may also be involved. Five common MM that begin in childhood affect multiple organ systems (see Table 6.2).

Management. All anesthetics suppress the RC complexes to varying degrees. Propofol infusion syndrome has NOT been associated with any perioperative complications in children with MM. Avoid prolonged fasting intervals. Preoperatively, it is essential to measure and stabilize the plasma glucose, lactic acid, and electrolyte concentrations. MELAS (see Table 6.2 for definition) is associated with lactic acidosis (>90% of children); lactated ringer's solution should be avoided in these children; a dextrose or saline solution is preferred. All anesthetics have been used in these children safely; specifically, adverse events have not been reported after dexmedetomidine, opioids, midazolam, and nitrous oxide. The response to neuromuscular blocking agents is unpredictable. The ideal anesthetic prescription for children with MM has not been determined; however, no anesthetic is contraindicated.

Table 6.2 Mitochondrial myopathies

Mitochondrial disorder ^a	Age of onset	Clinical manifestations
Leigh encephalopathy	Perinatal, early infancy	Nonspecific encephalopathy, hypotonia, dysphagia, central respiratory insufficiency
Pearson's Syndrome	Infancy, early childhood	Exocrine pancreatic dysfunction, DM, sideroblastic anemia, pancytopenia, nephropathy, hepatopathy, fatal
MERRF (myoclonic epilepsy with red-ragged fibers)	Childhood, young adult	Short stature, myopathy, neuropathy (peripheral and central incl. seizures), lactic acidosis (like KSS)
Kearns–Sayre Syndrome (KSS)	5–15 years	Short stature, myopathy, diabetes, retinitis pigmentosa and ophthalmoplegia, neuropathy, cardiac conduction defect, cardiomyopathy; sideroblastic anemia and Fanconi's disease
MELAS (mitochondrial encephalopathy, lactic acidosis and stroke-like episodes)	Late childhood, adulthood	Short stature, lactic acidosis, DM, myopathy, neuropathy, nephropathy, cardiac defects (conduction, cardiomyopathy), Fanconi's syndrome

^aAll are red-ragged fiber disorders on histologic staining except Leigh's disease

DM insulin-dependent diabetes mellitus

THE CHILD WITH CEREBRAL PALSY AND/OR SEIZURE DISORDER

Children with cerebral palsy (CP) frequently present for anesthesia; the increased survival of very-low-birth-weight infants is further increasing their numbers. CP is a collective term for a variety of clinical conditions resulting from a non-progressive neurologic insult early in life. The clinical state varies from mild local weakness with normal intelligence to severe spastic quadriplegia with gross cognitive and motor developmental delay. Common procedures in these patients include correction of scoliosis, rotation osteotomies, and baclofen pump insertion. Some with refractory seizures may require neurosurgery. Diagnostic procedures such as GI endoscopy and MRIs are common. It is important to obtain a careful history characterizing the severity, type, and frequency of seizures so as to know what to look for postoperatively, e.g., staring spells or tonic/clonic activity. Determine if the child is on a ketogenic diet for seizure control. Communicate with the child's pediatrician or neurologist to develop a plan for managing antiseizure medications postoperatively should the child be unable to take oral medications.

Special Anesthesia Problems

1. Communication with the child may be difficult or impossible, compromising the assessment of pain and making establishment of rapport with the patient difficult. In these circumstances, the parents or other caregivers should be co-opted and relied upon for assistance.
2. Many children have undergone repeated procedures and are extremely sensitized to the hospital environment. Careful planning is required to prevent further stresses.
3. The nutritional state may be suboptimal and may compromise anesthesia care and recovery from surgery. Preoperative hyperalimentation or placement of a feeding gastrostomy should be considered before extensive surgery.
4. Gastroesophageal reflux disease (GERD) is common and together with increased salivation may predispose to chronic pulmonary aspiration and recurrent pneumonia. Drugs to treat GERD should be administered on the morning of surgery.
5. Spasticity and contractures may make intravenous access and positioning difficult; carefully position and pad the child to prevent skin damage or nerve injury.
6. Baclofen (an agonist at GABA_A receptors in the dorsal horn) is commonly used to treat pain and contractures in CP. It is poorly absorbed via the GI tract and hence is given by an intrathecal pump. Overdose leads to drowsiness, respiratory depression, and delayed anesthesia recovery.

7. Antiseizure medication is frequently required. This must be continued perioperatively and may affect the actions of anesthesia drugs. Postoperatively, after extensive surgery, it is important to check anticonvulsant drug levels and restore these promptly. If the child remains NPO, fosphenytoin may be administered IV to prevent seizures. Be aware that older antiseizure medications such as carbamazepine, phenytoin, and valproic acid are associated with thrombocytopenia and bone marrow suppression. Some children will be on a ketogenic diet; in this situation, glucose and lactated ringers must be avoided and normal saline fluid therapy instituted. During extensive or prolonged procedures measure blood sugar periodically.
8. Latex allergy is common in children with CP; the history should be carefully evaluated for evidence of this and appropriate precautions taken (see above).

Anesthesia Management

1. Careful preoperative evaluation is required to assess comorbidities and to determine whether acute disease is present, especially the potential for pulmonary aspiration, atelectasis, or pneumonia.
2. The parents or other caregiver should be questioned about previous anesthesia experiences and their child's responses. Sedative premedication should be considered except for those who are severely obtunded or have a suspected difficult airway; a reduced dose may be appropriate in some cases. Anticholinergic premedication may be useful to decrease the sialorrhea and airway secretions.
3. If intravenous induction is planned, a suitable site should be selected and a topical local anesthetic (see p. 634) applied at the appropriate time. If the child is vasoconstricted, application of local warming may produce venodilation and improve success.
4. Intravenous propofol may be advantageous as it tends to decrease airway and bronchial reactivity. Otherwise, use sevoflurane to induce anesthesia.
5. The need for an RSI must be assessed for each child. Succinylcholine does not induce hyperkalemia in children with CP; the ED₅₀ for succinylcholine may be slightly less than in non-affected children. Children with excessive secretions should have the tracheal tube suctioned and sputum sent for culture for those with pulmonary disease.
6. The MAC of the inhalational anesthetics in children with CP is ~20 % less than in unaffected children.
7. Resistance to non-depolarizing relaxants as well as sensitivity have been observed in some children with CP and might be a result of interaction with anticonvulsant drug therapy or alterations in the neuromuscular acetylcholine receptors; a NM block monitor should be used. Assure appropriate recovery from neuromuscular blockade prior to extubation.

8. Children with CP are prone to hypothermia; monitor and maintain body temperature.
9. Endoscopic surgery is preferred because postoperative pain and complications are reduced.
10. Blood losses with extensive surgery (e.g., spinal instrumentation) may be greater than in normal children. This may be related to changes in vasomotor control and to platelet and clotting factor deficiencies. Appropriate supplies of blood and clotting factors should be available and used as indicated.

Postoperative Care

1. Regional analgesia or epidural/peripheral nerve catheters are ideal for postoperative pain control. The addition of clonidine to the infusion may reduce spasticity.
2. Parents or other caregiver should be present in the postoperative area to assist in the assessment of the child's discomfort.
3. After major spinal surgery in the child with CP the full resources of a pediatric intensive care unit are essential.
4. Antiseizure medications should be carefully continued. After major surgery blood levels of these drugs should be obtained to guide therapy. Fosphenytoin may be administered to those who remain NPO for any length of time.
5. Children with casts should be carefully monitored especially when epidural analgesia is employed. Compartment syndrome may follow osteotomies and is difficult to detect. Close cooperation with the orthopedic surgeon is essential.

ATYPICAL PLASMA CHOLINESTERASES

The genetically determined abnormal cholinesterases may result in prolonged apnea after the administration of muscle relaxants (e.g., succinylcholine). Five alleles of pseudocholinesterase found on chromosome 3 comprise the majority of phenotypes: normal, atypical, fluoride resistant, silent gene, and E Cynthiana (or Neitlich or C5 variant); further study added the H, J, and K variants. Atypical, fluoride resistant, and silent gene may significantly prolong recovery after succinylcholine, particularly in the homozygote state. Atypical and silent gene homozygous variants may prolong the duration of succinylcholine for several hours. The E Cynthiana variant rapidly degrades succinylcholine. In the homozygote atypical state (incidence 1:2500 children), recovery of muscle strength may be delayed for 5–6 h; in the heterozygous state (incidence 1:30), the delay may be unrecognized, 5–25 min.

Management of Prolonged Apnea

If muscle activity fails to recover after succinylcholine:

1. Continue to ventilate the lungs and provide sedation/anesthesia.
2. Confirm persistence of the neuromuscular block, using a nerve stimulator.
3. Allow the child to recover completely before ventilation and sedation/anesthesia are discontinued (this may take up to 3–6 h).

N.B. Prolonged apnea after succinylcholine due to an atypical cholinesterase is not serious provided the above steps are followed. Do not attempt to modify the neuromuscular block by giving reversal drugs; ventilate the lungs and be patient.

Blood samples for cholinesterase studies should be obtained:

1. Cholinesterase activity (normal range is 60–200 units but varies with the individual laboratory)
2. Dibucaine number (DN; normal range, 75–85)
3. Fluoride number (FN; normal range, 55–65)

Characteristically, children with the atypical enzyme have a low DN (15–25) and a low FN (20–25). Those heterozygous for the condition have intermediate values (DN, 50–70; FN, 40–50). The results of these tests may not be available for some days and are of no value during immediate management.

Other Considerations

When the diagnosis is confirmed, the child's family should have blood tests and be informed of their status. Those having homozygous atypical states should be advised to carry a warning card or wear a Medical Alert bracelet.

HEMATOLOGIC DISORDERS

Anemia

Children requiring surgery may be anemic. The hemoglobin (Hb) concentration, normally 18–20 g/dL at birth in full-term neonates, decreases to a nadir of about 10–11 g/dL by 3 months of age, and thereafter increases to 12–14 g/dL by 6 years (see Table 4.10). This is known as physiologic anemia of the newborn.

In the preterm infant, the Hb often decreases to lower concentrations because of a reduced red blood cell mass at birth, brief survival time of fetal red blood

cells, and poor erythropoietin response. Frequent blood sampling may exacerbate the anemia. In children, Hb levels below normal for age are most frequently caused by poor diet. Anemia discovered before elective surgery should be fully investigated and corrected before surgery. Most children with a chronic Hb level of 7–9 g/dL (i.e., renal failure) can be safely anesthetized provided no further blood loss takes place. If, however, the Hb is less than 7 g/dL, the physiologic consequences of anemia may significantly compromise the margin of safety during anesthesia, particularly if substantial surgical bleeding is expected.

The following considerations are relevant for children with anemia:

1. The major compensation of oxygen transport to tissues is by increased cardiac output; shift of the Hb-O₂ dissociation curve, caused by increased 2,3-diphosphoglycerate (2,3-DPG), contributes relatively little. At a Hb <8 g/dL, cardiac output increases to compensate for the decreased oxygen-carrying capacity.
2. Coronary sinus blood is normally very desaturated; therefore, in anemia, oxygen transport to the heart muscle is maintained only by increased coronary blood flow. At Hb levels <5 g/dL, myocardial oxygen needs are compromised resulting in subendocardial ischemia; high-output congestive cardiac failure may result.
3. Although unproven, significantly anemic children may be at increased risk for cardiac arrest during anesthesia due to a reduced margin of safety with desaturation events.
4. Children with cyanotic cardiac or respiratory disease compensate with a greater concentration of Hb than normal children; these children depend on an increased Hb but hyperviscosity may be an issue.
5. Anemic preterm infants are more prone to perioperative apnea but transfusion is not recommended.

The following plan of action is suggested for routine elective surgery:

1. If a significant anemia is discovered, delay elective surgery until the cause of anemia has been diagnosed and corrected. In children with iron-deficiency anemia, the Hb level increases significantly after 3–4 weeks of oral iron.
2. If surgery cannot be delayed, a decision must be made to proceed despite the anemic state or to transfuse packed cells to correct anemia. This decision depends on many factors, such as overall health of the child, expected surgical blood losses, and severity of anemia.
3. Use an anesthetic technique that is optimal for the anemic state:
 - (a) Avoid excessive preoperative sedation.
 - (b) Oxygenate the child before induction.
 - (c) Use increased concentrations of inspired oxygen during anesthesia.
 - (d) Always use a tracheal tube.

- (e) Use controlled ventilation and maintain normocapnia.
- (f) Be cautious with myocardial depressant drugs.
- (g) Carefully replace fluids to maintain the intravascular volume; hypovolemia must be avoided if the cardiac output is to be maintained.
- (h) Do not extubate until the child is fully awake.
- (i) Give additional oxygen continuously during transport to and in the postanesthesia care unit (PACU).
- (j) Keep the child normothermic.

Sickle Cell States

In sickle cell states, an abnormal Hb (sickle cell hemoglobin [HbS]) is present. When deoxygenated, sickle hemoglobin forms a gel, distorting the shape of erythrocytes, which then occlude vessels, causing infarction in lungs (acute chest syndrome), bones, spleen and brain. Additionally, erythrocyte life span is reduced with increased hemolysis and increased bilirubin level. Cholelithiasis is common. The course of the disease is one of many crises: sickling, hemolytic, or aplastic. Sickling crises result in ischemic pain, hemolytic crises result in further anemia, and aplastic crises may cause death. Ischemia due to sickling may involve many different organs. The disease is mainly confined to persons of African and Mediterranean descent.

In the USA, all neonates are screened at birth for sickle cell disease (SCD); however, those born outside the USA may not have been screened. SCD becomes evident during infancy (after 6 months of age) as HbS replaces fetal hemoglobin (HbF); the latter offers some protection against sickling. Hence, performing a Sickledex test in the first 6 months of life may not detect the presence of HbS. Infants with HbSS should receive pneumococcal vaccine and penicillin prophylaxis up to 6 years of age.

The severity of the disease depends on the percentage of HbS and the presence or absence of other abnormal forms of Hb (see Table 6.3):

1. Sickle cell trait (mild form, i.e., Hb AS-heterozygous): The incidence of sickle cell trait is approximately 8 % in the African American population. Sickling is unlikely to occur without severe hypoxemia and possibly during cardiopulmonary bypass.
2. SCD (severe form, i.e., Hb SS-homozygous): It occurs in 0.3–1.3 % of the Africa American population. This may cause serious complications in the perioperative period such as stroke, acute chest syndrome, and in severe cases a chronic lung disease.
3. The presence of another abnormal Hb may modify the disease. For example HbC, the second most common abnormal Hb, also promotes sickling. Children with HbSC have a normal Hb concentration but an increased risk

Table 6.3 Hemoglobin electrophoresis in older children

Syndrome	HbA (%)	HbS (%)	HbF (%)	HbC (%)
Sickle cell anemia	0	80–95	2–20	0
Sickle C disease	0	45–50	1–5	45–50
Sickle β-Thalassemia	0–30	65–90	2–15	0
Sickle cell trait	50–60	35–45	1–2	0
Normal	95–98	0	1–2	0

for sickling. Similarly, HbD promotes sickling when combined with HbS. On the other hand, HbF, if it is present (i.e., in thalassemia), may protect by reducing hemolysis and sickling. It is important to know the results of the Hb electrophoresis.

4. Neonates who have a large percent of HbF are not usually anemic or considered at risk for sickling. However, sickle cell crises have been reported in severely stressed neonates. Usually, the clinical signs appear by the time the child is a few months old.
5. Splenic function is impaired, serious infections may occur, and prophylactic antibiotics are indicated. Later, autosplenectomy may occur as a result of vaso-occlusive events.
6. Renal impairment (hyposthenuria) may occur in young children, leading to increased obligatory urine output and consequent increased risk of dehydration.
7. Acute chest syndrome, consisting of chest pain, respiratory distress, fever, and multilobe lung infiltrates, is common in children with SCD and may lead to severe hypoxemia. Therapy includes antibiotics, fluid therapy, transfusion, and pain control (PCA, NSAIDs, and possibly epidural analgesia). Often these children will present with “pneumonia” 1–3 days postsurgery.
8. In later life, pulmonary infarction leads to pulmonary fibrosis, pulmonary hypertension, and cor pulmonale. Stroke may develop in 10% of children.

Special Anesthesia Problems

1. A sickling crisis may be precipitated by general or local hypoxemia.
2. Sickling is more likely if the child is anemic, acidotic, hypotensive, dehydrated, septic, and/or hypothermic or if the child’s blood contains Hb C or D.
3. If the child has SCD, previous vascular occlusive crises may have permanently impaired cardiac, hepatic, and/or renal function.
4. Serum cholinesterase activity may be reduced.

Anesthesia Management

Perioperative management of SCD requires the assistance of a hematologist in deciding which measures are appropriate for each child.

Preoperative

1. All children of African American descent who require anesthesia must be screened for sickle cells:
 - (a) Neonatal screening using Hb electrophoresis is routine in the USA but children from other countries may not have been screened.
 - (b) Results of solubility tests (Sickledex, Sickleprep) are available in 5 min but do not differentiate SCD from trait for those who have not been screened.
 - (c) Screening tests are unreliable in infants less than 6 months of age because HbF masks the results.
 - (d) Hb electrophoresis provides the most accurate diagnosis.
2. In general, a severe form of the disease is less likely if anemia is absent, but a Hb electrophoresis is still essential to exclude any other abnormal Hb (e.g., Hb C and D) and to assess the severity of the condition (Table 6.3). Children with Hb SC disease may have a normal Hb concentration but are still at risk for sickling.
3. If the child has sickle cell trait (less than 50 % HbS):
 - (a) Avoid preoperative dehydration; encourage clear fluids until 2 h before surgery if appropriate, or start intravenous fluids during the fasting period.
 - (b) Avoid excessive preoperative sedation.
4. If the child has SCD (70–90 % HbS):
 - (a) Assess the child carefully, particularly for sequelae of previous sickling crises (i.e., CNS strokes occur in up to 30 %, cardiac, pulmonary, or renal infarction). Become familiar with previous pain management strategies and drug doses.
 - (b) Preoperative transfusions may be indicated. Exchange transfusion and simple transfusion to a Hb of 10 g/dL have been shown to be equally efficacious in reducing perioperative complications. Therefore, the most common approach is the following:

Transfuse packed cells to bring the hemoglobin up to 10 g/dL.
Hydrate the child preoperatively with 1–1/5 × maintenance balanced salt solution.

Be aware that with each transfusion bears the potential for the development of new antibodies in the recipient which could make it difficult to find compatible blood in the future.
 - (c) Those who require cardiopulmonary bypass with hypothermia should have packed cell infusions or exchange transfusion to reduce the HbS level to less than 5 %.
 - (d) In an acute emergency, exchange transfusion may be performed.

Perioperative

1. Use high concentrations of inspired oxygen (at least 50 %) to maintain 100% saturation and control ventilation.
2. Maintain normal acid-base status
3. Maintain normal body temperature.
4. Maintain fluid balance to avoid dehydration or excessive fluid administration.
5. Beware of regional ischemia:
 - (a) Position and pad the child carefully to avoid any regional vascular stasis.
 - (b) Do not use a tourniquet unless it is essential. If one is used, exsanguinate the limb well and use the tourniquet for a minimal time.
 - (c) Check the blood pressure cuff and other equipment frequently to see that no locally constricting effects are produced.

Postoperative

1. The child must be awake before extubation.
2. Continue supplemental oxygen and monitor SpO₂ continuously during transport to and in the postanesthesia care unit (PACU); continue supplemental oxygen for up to 24 h.
3. Hydration and warmth must be maintained.
4. Be alert for the possibility of pulmonary complications; they are common in children with SCD.
5. Provide effective pain management
6. Early mobilization
7. Incentive spirometry may be indicated

Hemophilia

Factor VIII Deficiency (Classic Hemophilia Type A)

Classic hemophilia is inherited as an X-linked recessive disorder (1:10,000 incidence) and characterized by episodes of bleeding, either spontaneous or after minimal injury. The presenting sign may be bleeding from the umbilical cord in neonates or after circumcision in infants. The diagnosis is confirmed by factor VIII assay. During childhood, many sites may be involved, hemarthrosis is common, and retroperitoneal bleeding may occur. Hemophilic children require special care during any operation, especially dental extractions.

Surgical Management. Children with hemophilia should undergo elective surgery only in hospitals with facilities to care for this condition. Team care by a

hematologist, a surgeon, and an anesthesiologist is essential. If an emergency operation is essential but the facilities of a hematology department are not available, give fresh frozen plasma (FFP) (20 mL/kg) preoperatively and consult with a hematologist.

Preoperative

1. If there is any doubt about the diagnosis, factor concentrations should be assessed. Preoperative investigation should also include screening for factor VIII inhibitors (found in 5–10 % of children), even if inhibitors were not detected previously.
2. One hour before surgery, an infusion of factor VIII concentrate (25–50 U/kg) is given, followed by an assay for plasma factor VIII activity (1 U/kg factor VIII increases plasma level by 2 %, $t_{1/2}$ is 12 h). Surgery can proceed if factor VIII activity is greater than 50 %; some procedures such as CNS or trauma require corrections to 80–100 %. In mild cases, desmopressin (1-deamino-8-D-arginine vasopressin, DDAVP) may be effective. Recombinant factor rVIIa eliminates infectious disease risk.

Perioperative

1. Exercise great care during airway instrumentation; avoid trauma that might provoke submucosal hemorrhage.
2. A continuous infusion of factor VIII (3–4 units/kg/h) may be advisable during major surgery (consult the child's hematologist).

Postoperative

1. Depending on the nature of the surgery, factor VIII levels should be maintained at 50 % for several days. This is achieved by giving factor VIII, as dictated by repeated assay, preferably by continuous infusion.
2. After dental extraction, ϵ -aminocaproic acid (Amicar) may help to inhibit fibrinolysis of formed blood clot.

N.B. When factor VIII inhibitors are present, therapy presents a major challenge. Various methods have been used, including very large doses of factor VIII combined with immune tolerance induction therapy, porcine factor VIII, factor eight inhibitor bypass therapy, and recombinant factor rVIIa.

Factor IX Deficiency (Christmas Disease, Hemophilia Type B)

This disease is similar to Hemophilia A. Children with factor IX deficiency are treated as for factor VIII deficiency except that factor IX levels are assayed and factor IX infusions are given. A recombinant factor rFIX is now available. Problems with inhibitors are common in this condition as well.

Von Willebrand Disease

Von Willebrand disease (VWD) is the most common congenital bleeding disorder; it may affect 1% of children. There are many forms of the disease, both congenital and acquired, and its incidence varies widely among different ethnic groups. It may occur secondary to another disease (e.g., chronic renal failure, congenital heart disease, Wilms tumor) and may resolve with treatment of that disease. The basic defect present in VWD is the lack of a plasma cofactor that is a carrier protein for factor VIII and is necessary for platelets to adhere to damaged endothelium. VWD is generally divided into types 1, 2, and 3 with many subtypes but in practical terms into mild, moderate, or severe depending upon bleeding history and factor values.

Type 1 is the most common (60–80%) with slightly reduced von Willebrand factor but often normal prothrombin (PT) and partial thromboplastin times (PTT). Most are asymptomatic until a dental extraction or surgical procedure.

Type 2 accounts for 20–30% of cases and there is a qualitative rather than a quantitative defect of von Willebrand factor (normal levels).

Type 3 is the most severe form (fewer than 1% of cases) in which von Willebrand factor is absent and factor VIII values are extremely low, leading to confusion with hemophilia A.

The clinical manifestations depend on the severity of the disease. Cutaneous and mucous membrane bleeding are common, but deep tissue bleeding may also occur. A history of easy bleeding or failure to clot after dental extraction should alert the anesthesiologist. In type 1 and type 2a disease, DDAVP is usually effective, but it may exacerbate type 2b and is ineffective in type 3 disease. The latter types must be treated with cryoprecipitate, fresh-frozen plasma, or recombinant rVIIa.

If the type of disease is unknown preoperatively, it may be appropriate to assess the effect on coagulation studies of an infusion of DDAVP. If the bleeding time is shortened, DDAVP may be useful perioperatively. If not, blood products will be necessary. Consultation and advice from a hematologist is advised.

DIABETES MELLITUS

Diabetes mellitus is the most common endocrine disorder of childhood. Children with symptomatic diabetes are all insulin dependent; type 2 diabetes can occur in obese children and adolescents; but it is asymptomatic and usually undiagnosed.

Young children typically have weight loss, polydipsia, and polyuria. However, the onset may be very abrupt, and the child may have ketoacidosis. The presentation may be accompanied by abdominal pain, leukocytosis, and mimic appendicitis. Vascular complications seldom occur in diabetic children, so the renal, cardiac, or peripheral vascular effects of the disease need not be anticipated.

The current approach to juvenile diabetes is to maintain blood glucose levels as close to normal as possible throughout the day by the use of twice- or thrice-daily insulin doses; some children may be using an insulin infusion pump. In some cases, control is difficult, and childhood diabetes is often unstable; the anesthesiologist should consult closely with the medical team in planning insulin management.

Anesthesia Management

Perioperatively, it is essential to achieve close control of the blood glucose level. Perioperative hyperglycemia (>200 mg/dL) is associated with increased wound infection rates in adults, similar data are not available in children.

Preoperative

1. Close monitoring of blood glucose should be instituted several days before elective major surgery to permit stabilization.
2. Minor elective surgery for children with well-controlled diabetes may be planned on an outpatient basis.
3. Severe hyperglycemia and ketosis should be treated with appropriate insulin adjustments before surgery is scheduled. HbA1c should be followed to assess metabolic stability. Defer emergency surgery, if possible, until the ketoacidosis is corrected.
4. Brief procedures (less than 1 h):
 - (a) Schedule as early as possible in the morning.
 - (b) Determine the preoperative blood glucose level.
 - (c) Give one half to two thirds the usual dose of insulin as intermediate-acting insulin only (NPH or Lente). No short-acting insulin should be given.

- (d) Start an intravenous infusion of 5% glucose-containing balanced salt solution and infuse at a maintenance rate (3–4 mL/kg/h) unless the blood glucose is less than 5 mmol/L (90 mg/dL), in which case the rate should be increased and the blood glucose rechecked.
 - (e) Determine the blood glucose concentration hourly during surgery and immediately after the procedure.
 - (f) Use an anesthesia technique that will minimize PONV and permit early return to normal oral intake.
5. Prolonged procedures (>1 h): The preferred technique is to give an infusion of insulin plus an infusion of dextrose and monitor blood glucose levels frequently.
- (a) Add 50 units of regular insulin to 500 mL of isotonic saline. (Each milliliter contains 0.1 unit of regular insulin.) Saturate the insulin-binding sites of the intravenous tubing by running 50–100 mL of the solution through the tubing.
 - (b) Use a Y-tube or piggyback connection to the maintenance intravenous line, and control the rate with an infusion pump.
 - (c) Infuse 1 mL/kg/h to deliver 0.1 units of insulin per kilogram per hour.
 - (d) Infuse a 5% glucose balanced salt solution at a rate to maintain blood glucose at 5–10 mmol/L (90–180 mg/dL).
 - (e) Assess intraoperative blood glucose levels hourly.
 - (f) Postoperatively, the insulin infusion may be continued as long as necessary, depending on the circumstances. The child's pediatrician should control the rate and amount of the infusion based on the blood glucose concentration.

Postoperative

1. Brief procedures:
 - (a) If the child is receiving subcutaneous insulin, give appropriate doses of short-acting insulin as required based on blood glucose levels. Continue intravenous hydration until the child is taking fluids orally.
 - (b) The child's usual insulin regimen can be resumed the next day if the child is able to tolerate fluids or food.
2. Prolonged procedures:
 - (a) Change to subcutaneous insulin when the child's condition allows (e.g., when the child is taking fluids orally and intravenous hydration has been discontinued).
 - (b) Pediatricians should follow-up and manage the child's diabetes.

Children Using an Insulin Pump

1. Ensure that the subcutaneous injection site is well protected and that the infusion needle does not become dislodged.
2. Plan management in consultation with the medical team. Generally, the basal rate will be maintained, omitting bolus doses as meals are missed. Boluses may be administered depending upon the results of blood glucose determinations.

Emergency Surgery

Diabetic ketoacidosis may result from the physiologic stresses of the surgical disease, but remember also that the symptoms of ketoacidosis may mimic those of an acute abdomen.

1. Attempt to stabilize the child before proceeding to the OR. Stabilization should be undertaken in cooperation with the medical team. Ketoacidosis must be corrected, intravascular volume restored, and electrolyte defects normalized.
2. Insert monitoring lines for central venous pressure (CVP) and arterial pressure to assist in correcting hypovolemia and hyperglycemia.
3. Infuse warmed fluids to correct hypovolemia; normal saline 10–20 mL/kg over 1 h may be required, as indicated by the CVP.
4. Intravenous regular insulin will be required. A bolus dose of 0.1 U/kg followed by an infusion of 0.1 U/kg/h may be used.
5. Monitor blood glucose frequently; the objective is to decrease the concentration by a maximum of 100 mg/dL each hour. More rapid declines may result in detrimental osmotic shifts.
6. When the blood glucose concentration is less than 300 mg/dL, continue rehydration with 0.45% saline and 5% dextrose solution.
7. Monitor serum electrolytes frequently; hypokalemia is to be expected. Potassium must be added to the infusion provided that renal function is satisfactory.
8. Metabolic acidosis corrects spontaneously as hypovolemia is corrected and insulin administered. Ketones are metabolized, and their production is halted. Sodium bicarbonate is unnecessary and may be deleterious except in very severe acidosis (pH less than 7.0).
9. Subclinical brain swelling occurs during the treatment of diabetic ketoacidosis and, rarely, dangerous cerebral edema may develop. It is recommended that children be closely monitored and that total fluid administration be limited to 4 L/m²/24 h.

MALIGNANT DISEASES

Anesthesiologists frequently care for children with malignant disease. Special problems may arise depending on the site and type of disease, but all of these children require special attention to their emotional status. Extreme care must be taken to ensure a minimum of discomfort and upset for both child and parents during painful diagnostic and therapeutic procedures (e.g., lumbar puncture, bone marrow aspiration).

Special Anesthesia Problems

The child with a malignancy may present special problems:

1. Abnormal anatomy, including the airway, may be present; be especially aware of the child with enlarged hilar lymph nodes.
2. Hematologic disease may result in anemia, coagulopathy, and immune deficiency. Coagulopathy may lead to intraoperative pulmonary hemorrhage and difficulty with ventilation. Urgent bronchoscopy is required to remove clots.
3. Increased susceptibility to infection means that care in asepsis is vital.
4. A history of long-term steroid therapy necessitates consideration of perioperative stress dose corticosteroids.
5. Nausea and vomiting may complicate radiotherapy and/or drug therapy and lead to dehydration and electrolyte disturbance. Check the biochemistry levels. Do not administer dexamethasone without consulting with the oncologist—tumor lysis syndrome may result.
6. Cardiomyopathy may follow total body irradiation, cyclophosphamide, doxorubicin, or daunomycin (see later discussion).
7. Hypercalcemia may accompany malignant bone tumors.
8. Nephropathy may lead to impaired renal function.
9. Increased intracranial pressure may occur with involvement of the CNS.
10. Peripheral neuropathy may occur.
11. Muscle weakness and hypotonia occur in advanced malignant disease.
12. Toxic effects of chemotherapy may be present (see later discussion).
13. Tumor lysis syndrome is a metabolic disorder that accompanies treatment of lymphomas and leukemia characterized by hyperkalemia, hypocalcemia, hyperuricemia, hyperphosphatemia, and acute renal failure.
14. Meticulous attention to aseptic techniques when accessing vascular access devices is essential.

Adverse Effects of Commonly Used Drugs

All antineoplastic drugs can cause the following effects:

1. Bone marrow suppression, with anemia, leukopenia, and thrombocytopenia
2. Anorexia, nausea, and vomiting
3. Stomatitis and alopecia
4. Decreased resistance to infection

Some of these agents produce additional adverse effects:

1. Hepatotoxicity (i.e., methotrexate, cyclophosphamide), check liver function indices.
2. Renal toxic effects (i.e., cisplatin, ifosfamide), which may be increased by the use of aminoglycoside antibiotics or diuretics (furosemide).

Furthermore, specific drugs have effects on cardiac or pulmonary function:

1. Cardiotoxic effects

ECG changes are nonspecific, but prolongation of the Q-T interval suggests toxicity. Echocardiography is the most useful index of cardiac function; recent studies should be reviewed before induction.

- (a) Both *daunomycin*, used in leukemia therapy, and *doxorubicin* (Adriamycin), used in therapy for solid tumors and leukemias, affect the heart, causing:

Nonspecific electrocardiographic (ECG) changes, with any dose. Disturbances of conduction, including supraventricular tachycardia, atrial and ventricular extrasystoles, and ventricular fibrillation. Drug-induced cardiomyopathy in 1–2 % of children, leading to congestive heart failure.

The cardiac effects of doxorubicin are dose related. A total cumulative dose of 250 mg/m², or 150 mg/m² if combined with mediastinal radiation, must alert the anesthesiologist and is an indication for a full cardiologic assessment particularly an echocardiogram to assess cardiac contractility. Children with a history of congestive heart failure are particularly likely to experience perioperative complications.

Myocardial depressant drugs (e.g., inhalation agents) should be used with caution or avoided and cardiac parameters should be closely monitored.

Beta blockers and calcium channel-blocking drugs may dangerously increase the cardiotoxic effects and should be avoided.

- (b) *Mitoxantrone* may cause cardiotoxicity, especially if given after previous anthracycline therapy and with cumulative doses greater than 120 mg/m².
 - (c) *Cyclophosphamide, cisplatin, 5-fluorouracil, amsacrine, mithramycin, mitomycin, vincristine, and actinomycin D* may be cardiotoxic or contribute to cardiotoxicity at large doses.
2. Pulmonary toxicity
- (a) *Bleomycin*, (testicular tumors and Hodgkin disease) causes pulmonary fibrosis in ~10% of patients and may result in death (1%). *The effects on the lung are accelerated by hyperoxia, and oxygen therapy should be carefully controlled at all times. Fluid therapy should be strictly limited since overload may further compromise lung function by causing pulmonary edema.*
 - (b) *Busulfan, carmustine, methotrexate, and mitomycin* may cause pulmonary fibrosis.
 - (c) *Cytosine arabinoside, vinblastine, and mitomycin* have been associated with noncardiac pulmonary edema.

3. Anticholinesterase inhibition

- (a) *Cyclophosphamide* and other alkylating agents—used for lymphomas, Hodgkin disease, and leukemias— inhibit serum cholinesterase; prolonged apnea with succinylcholine may occur.

THE CHILD WITH A TRANSPLANT

There are numerous children with transplanted organs; these children require special considerations:

- 1. These children and their families have survived extensive medical and surgical interventions; they require very considerate emotional care.
- 2. All of them receive a regimen of antirejection drugs, which may have important side effects.
 - (a) All of the antirejection drugs cause reduced resistance to infection. *Take extreme care with aseptic technique.* Insert only those intravenous and monitoring lines that are really needed, and removed them as soon as possible.
 - (b) Cyclosporine can cause hypertension, hyperkalemia, and nephrotoxicity; therefore, check renal function tests. In addition, it may interact with and potentiate barbiturates, fentanyl, and muscle relaxants.
 - (c) Azathioprine can cause bone marrow depression and hepatotoxicity; check liver function. It also has anticholinesterase effects and may prolong the action of succinylcholine and antagonize non-depolarizing relaxants.

- (d) Prolonged steroid therapy demands the usual considerations and appropriate supplementation.
 - (e) OKT3, usually used to treat rejection crises, can cause anaphylaxis and acute pulmonary edema, especially if fluid overload is present. It may also cause psychiatric disturbances.
3. Teenagers have a relatively high rate of noncompliance with their antirejection therapy. This may lead to problems with graft rejection.

Special Considerations for the Child with a Transplanted Heart

Children with a denervated transplanted heart have altered cardiac function.

- 1. Effects normally mediated via the autonomic nervous system are absent (i.e., vagal slowing, baroreceptor responses to blood pressure changes). Changes in heart rate as an index of light anesthesia or hypovolemia are unreliable.
- 2. Indirect drug effects that depend on autonomic pathways are absent (i.e., the chronotropic effects of atropine, pancuronium, or opioids).
- 3. Compensation for changes in blood volume and cardiac filling pressure is limited and delayed.
- 4. Small vessel coronary atherosclerosis is accelerated in transplanted hearts and may occur in children. In the denervated heart, ischemia may occur without pain and coronary angiography is required for diagnosis. Careful intraoperative ECG monitoring is essential. Manage anesthesia as one would for an adult with severe coronary artery disease.

The following are considerations for anesthesia in the child with a heart transplant:

- 1. The child should be carefully screened for signs of rejection, which if present may increase the risks of anesthesia and surgery. Signs include:
 - (a) Poor appetite, irritability, fluid retention
 - (b) Decreased cardiac function on echocardiogram
 - (c) Low-voltage ECG
- 2. Practice meticulous asepsis. Avoid unnecessary cannulation; use only essential invasive monitoring lines, and remove these as soon as is safe.
- 3. Maintain normovolemia; ensure adequate fluid replacement.
- 4. Avoid high doses of drugs that have a direct cardiac depressant effect (e.g., inhalation agents).
- 5. Maintain afterload; avoid agents and techniques that cause rapid changes in vascular tone.

6. If cardiotonic drugs are indicated, use direct-acting agents (e.g., isoproterenol, dopamine and epinephrine).

Most children are successfully managed using an opioid- and relaxant-based anesthetic. Despite theoretic considerations of interaction between muscle relaxants and immunosuppressive therapy, the usual doses are often required (but neuromuscular blockade must be monitored).

Special Considerations for the Patient with a Transplanted Lung

Heart and lung transplantation may be performed for children with cardiac disease complicated by pulmonary hypertension. Lung transplantation is performed in children with end-stage CF, primary pulmonary hypertension, or idiopathic pulmonary fibrosis. The transplanted lungs are prone to infection and also to obliterative bronchiolitis that progresses to respiratory failure.

Regular bronchial alveolar lavage with transbronchial biopsy is performed to monitor for infection or signs of rejection. General anesthesia usually is required. A technique using a propofol infusion with and without short-acting muscle relaxation with a tracheal tube or LMA has been satisfactory. The largest tracheal tube that can easily be passed should be used. Many of these children experience considerable dyspnea on emergence from anesthesia and are more comfortable if placed in a sitting position as they awaken. Secretions may also be troublesome.

Special Considerations for the Patient with a Transplanted Liver

After a successful liver transplantation, children can be expected to have normal metabolic functions and drug metabolism. Therefore, any suitable anesthesia regimen can be used, and no agents are contraindicated. However, these children are prone to infections, particularly by viral agents (cytomegalovirus, Epstein–Barr virus, hepatitis virus), and require very careful aseptic precautions.

Those children with abnormal liver function may have abnormal drug distribution, protein binding, metabolism, and clearance. In addition they may have a coagulopathy. The PT is considered one of the most useful tests of hepatic function because it becomes prolonged before most other tests are abnormal. In these children, anesthesia regimens should be carefully chosen; inhalational agents may be used cautiously, but the response to opioids can be unpredictable. *Cis*-atracurium and remifentanil offer advantage since neither require hepatic metabolism.

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Postoperative Care and Pain Management

THE POST-ANESTHESIA CARE UNIT

General Management

All children should be transported to the post-anesthesia care unit (PACU) in the lateral decubitus position with the head extended to provide an unobstructed airway; all children—other than absolutely healthy children having minor surgery—should receive oxygen or be monitored with SpO₂ during transport to PACU. Evidence suggests that decreases in SpO₂ during transport are most likely due to airway obstruction; be alert to this possibility. *However, administering oxygen while monitoring SpO₂ renders the oximeter an insensitive metric of hypopnea; respiration should be monitored by observing chest movements, mist in the facemask, or by feeling exhaled breathing.* The anesthesiologist walks at the head of the bed facing forward to continuously observe and monitor the child. If the airway is in doubt, do not leave the OR. If the airway becomes precarious while on route to the PACU, apply digital pressure to the condyle of the mandible behind the pinna to open the upper airway. Transport with a self-inflating bag or Mapleson D system at the ready is recommended. Children with a history of upper respiratory tract infections (URTI) are particularly at risk of airway problems.

In the PACU, the anesthesiologist:

1. Transfers the child to the care of the PACU nurses (see later discussion); summarizes the child's underlying medical and surgical conditions, the operative procedure, and any associated issues (e.g., bleeding, vascular compromise); describes the size tracheal tube used, distance of insertion, and ease of intubation; provides the timing and doses of antibiotics, analgesics, antiemetics, intravenous fluid therapy, and blood loss and replacement; and describes the anesthesia technique including any complications and their management. Specific orders should be given for any continuing anesthesia care (e.g., continuous epidural analgesia).

2. Completes the anesthesia chart by recording the initial vital signs obtained by nursing staff in the PACU.
3. Completes postoperative (PACU) orders, including pain and anti-emesis orders, intravenous fluids, and respiratory therapy.
4. Remains with the child until handover is complete and the PACU nurses accept care of the child.

In the PACU, every child receives oxygen via facemask or nasal prongs as tolerated. The anesthesiologist should not transfer the care of a child to the PACU nursing staff if there is any doubt about airway patency and the adequacy of ventilation. *A child who still requires an oropharyngeal airway or tracheal tube may still need an anesthesiologist.*

Small infants (younger than 3 months of age) may not rapidly convert to mouth-breathing if the nasal passages are blocked (i.e., after cleft lip or palate repair). If such obstruction occurs, insert an oropharyngeal or nasopharyngeal airway for patency until the child is fully awake.

The progress of recovery should be documented with a post-anesthesia scoring system along with regular recording of vital signs. All children should receive oxygen until they can maintain an adequate SpO₂ in room air. SpO₂ should be continuously monitored until the child is fully awake and ready for discharge.

As soon as the child awakens, has stable vital signs and is free from pain, the parents may be present at the bedside. This decreases the child's anxiety in the PACU, reduces crying, and reduces the need for sedation. It also clarifies whether the child is crying because of pain or the absence of a parent.

After ketamine anesthesia, recovery should take place in a quiet, dimly lit area with minimal tactile and auditory stimulation. If, despite these precautions, delirium and/or hallucinations develop, midazolam 0.05–0.1 mg/kg IV or diazepam (0.1–0.2 mg/kg IV) may be administered.

Complications in PACU

Laryngospasm

Laryngospasm may occur during emergence from anesthesia. During anesthesia, it may be treated as outlined elsewhere (Chap. 4) and possibly by deepening the level of anesthesia. During extubation and recovery, it is more likely to occur in children with blood or secretions in the pharynx or in those with a history of URTI (see Chap. 4). It should be managed by bag-mask ventilation with oxygen, maintaining positive airway pressure, and subluxation of the TMJ. Be prepared to administer either propofol or a short-acting muscle relaxant; reintubate the trachea if necessary and do not delay reintubation if desaturation progresses. Often a very small dose of succinylcholine (0.2 mg/kg) will relieve the laryngospasm.

Non-cardiogenic pulmonary edema may follow immediately upon relief of severe laryngospasm. If this occurs, treat with continued positive pressure ventilation, furosemide, fluid restriction, and supplemental oxygen as indicated.

Postoperative Stridor

Postoperative stridor caused by subglottic edema may occur especially after endoscopy, in children with a history of croup, those who were intubated as neonates, or after the unwise use of too large a tracheal tube diameter. Stridor is more common in children with Down syndrome and after surgery during which head movement occurred. Stridor usually appears within 30–60 min after extubation. The use of humidified oxygen and intravenous dexamethasone may reduce subglottic edema. If stridor persists, administer racemic epinephrine by spontaneous respirations or preferably intermittent positive-pressure breathing for 15 min; this is usually effective. If racemic epinephrine is used, then the child should be observed for an extended period of time for possible rebound edema. Very rarely it may be necessary to reintubate the airway in the PACU for persistent severe stridor. In such cases, a smaller-diameter tube that is accompanied by an audible leak should be used.

Emergence Delirium

Delirium occurs most commonly but not exclusively in children 2–6 years of age after sevoflurane anesthesia, with a reported incidence up to 80 %. Delirium has also been reported after desflurane or isoflurane. It is characterized by the presence of restless, thrashing, and inconsolable behaviors; disorientation; failure to establish eye contact; and a lack of purposeful movement and awareness of their surroundings. The delirium is usually transient, dissipating spontaneously within 10–20 min, without sequelae. The incidence may be attenuated by pre-treatment with propofol, opioids, dexmedetomidine, clonidine, or NSAIDs (ketorolac). Difficulty differentiating emergence delirium from postoperative pain has been addressed in part, with the introduction of a validated scale (Pediatric Anesthesia Emergence Delirium scale) to measure delirium in children. Treatment in the PACU may require small doses of propofol or fentanyl.

Shivering and Rigidity

Shivering and rigidity may occur during recovery from anesthesia, sometimes associated with hypothermia and may at other times, occur in normothermic children. This may increase the metabolic rate and oxygen requirement and is very undesirable after some procedures (e.g., orthopedic). It should be treated with low-dose intravenous meperidine (Demerol) (0.25 mg/kg IV) or dexmedetomidine (0.5 µg/kg IV slowly), which may eliminate the shivering.

Nausea and Vomiting

Postoperative nausea and vomiting (PONV) is troublesome and a leading cause of delayed discharge from the hospital or, more rarely, of unplanned admission of the day surgery patient. The incidence of PONV can be significantly reduced by some general measures:

1. Avoid the indiscriminate use of opioids; a single dose dramatically increases PONV. Use alternative analgesic drugs (e.g., nonsteroidal anti-inflammatory drugs [NSAIDs]) or regional analgesia whenever possible. However, pain itself may cause PONV—so ensure analgesia. Regional analgesia is ideal.
2. Administer large volumes (up to 30 mL/kg) of balanced salt solution intraoperatively.
3. Avoid oral fluids postoperatively; wait until the child asks for them or is thirsty.
4. Do not rush to mobilize the child, especially after eye surgery.

When nausea and vomiting can be anticipated (e.g., strabismus surgery, adenotonsillectomy), the incidence can be reduced by the choice of the anesthetic regimen (e.g., propofol), by avoiding nitrous oxide (in emetogenic surgery), by administering large volumes of intravenous fluids (20–30 mL/kg), and by prophylactic multimodal antiemetic therapy (dexamethasone and a serotonin-receptor antagonist, such as ondansetron). In children with unexpected nausea and vomiting or when the above therapies fail, rescue medication with an antiemetic drug is necessary (Table 7.1).

Ondansetron is the most effective antiemetic for PONV. Dexamethasone is not a rescue antiemetic. If it was not given intraoperatively, it may be given postoperatively but should be given over 5 min to reduce the risk of perineal irritation. Dimenhydrinate and metoclopramide are both moderately effective antiemetics and cause little sedation; they may be used for protracted vomiting in PACU. Droperidol in doses adequate to combat nausea and vomiting may cause sedation, with delayed recovery and discharge.

Table 7.1 Antiemetic drug doses for children

Dexamethasone	75–150 µg/kg (maximum 8 mg)
Ondansetron	50–100 µg/kg (maximum 4 mg)
Dimenhydrinate	0.5 mg/kg (maximum 25 mg)
Droperidol	10–15 µg/kg (maximum 1.25 mg)

Duration of Stay in PACU

Children remain in the PACU until they are fully awake and recovered from the effects of anesthesia. As a general rule, a minimum stay of 30 min or two sets of vital signs is required. Infants weighing less than 5 kg are usually kept in the PACU for a more prolonged period or transferred to a monitored bed. Be alert

for possible postoperative complications (e.g., stridor after surgery of or near the airway or after endoscopy; bleeding after a kidney or liver biopsy, tonsillectomy), and specify a longer stay in the PACU for such children. Children with lower respiratory tract signs and persistent desaturation may require deep-breathing and coughing exercises and chest physiotherapy.

Each child should be signed out of the PACU by an anesthesiologist except for the most simple of cases (e.g., myringotomy). If the discharge order from the PACU is delegated to the nurse, specific written clinical criteria should be documented. If an anesthetic complication occurs, the child must be re-evaluated by the anesthesia team before discharge from PACU. If the child receives IV opioids in the PACU, they should be monitored for an additional 30 min to ensure adequacy of ventilation before discharge.

MANAGEMENT OF PAIN

The ability of infants and children to feel pain was misunderstood in the past, and this led to its undertreatment. It is now recognized that the biochemical and nervous components of the pain perception pathways are completely formed during fetal life and that even the preterm infant can feel pain. Furthermore, the adverse effects of unmodified pain have been documented even in very young infants. Studies suggest that inadequate treatment of pain in infants may lead to increased sensitivity to pain later in life.

There are many reasons why pain in children was undertreated in the past and why even today it is inadequately treated:

1. Infants cannot tell us when they feel pain, and it is sometimes difficult to determine whether they are crying because they are in pain or for another reason.
2. The older child's response to pain differs from that of the adult; often these children are quiet and withdrawn, failing to announce their discomfort.
3. In the days when intramuscular injection of an opioid was the standard therapy for postoperative pain, children often feared the injection more than the pain and preferred to suffer in silence. This tended to perpetuate the myth that children do not feel pain as much as adults.
4. Physicians have been uncertain of the safety of the analgesic drugs given to infants. It was stated that infants are "exquisitely sensitive" to the respiratory depressant effects of morphine; this led to an ultraconservative approach in prescribing opioids.
5. Many physicians, and especially those junior staff to whom the responsibility for pain management was customarily delegated, were unsure of the correct dosage of analgesics for infants and children.
6. Nurses have tended to underestimate pain in children; many healthcare providers have overestimated the danger of the child's becoming addicted to opioids.

Currently, we know that all children can experience pain, we are better equipped to assess the severity of the pain (pain scoring systems), and we have better means to control pain (PCA, nerve blocks, continuous catheter techniques). Postoperative pain management should be planned during the preoperative evaluation and discussed with the child and parents; consent for additional procedures should be obtained where appropriate.

For outpatients, it is most important that the parents are well instructed in the management of pain when the child arrives home:

1. Analgesic drugs must be administered before pain becomes significant and repeated regularly by schedule rather than waiting for pain to be a problem. The only exception to this approach would be the child with OSA where opioids could pose a threat to their safety; techniques such as alternating acetaminophen/ibuprofen at fixed intervals are recommended.
2. The “analgesic gap” as regional analgesia wears off must be anticipated (most often manifest by the child becoming irritable or quiet) and suitable analgesics administered in advance.
3. Parents should be instructed to look for signs of pain, to use assessment tools (e.g., visual analog scales [VAS]), and to administer effective analgesics appropriately. A standard VAS may be sent home with the child and the parent instructed in its use. Written instructions should be provided to the parents and discussed with them.

Assessment of Pain

It is essential for optimal pain management to establish regular, objective pain level assessments recorded on the medical record. For infants, the level of pain is assessed by physiologic or behavioral indices. Indices of pain include tachycardia, tachypnea, increased blood pressure, sweating, facial expressions, posture, and crying. Of the behavioral indices, facial expression may be most reliable, but cry characteristics and body movement (especially flexion of the limbs) are also useful. The opinion of the parent and of the child’s nurse in interpretation of these behavioral signs is very useful. These indices are incorporated into a numeric scale that can be scored and recorded (Table 7.2).

Older children may be asked to report their pain level using one of a variety of VASs, such as the Wong-Baker FACES Pain Rating Scale (Fig. 7.1). They may also be asked to rate their pain on a color scale or to report it by coloring their pain on a body outline. Adolescents can be assessed with the use of standard adult self-report scales. Note, however, that at this age psychological and emotional factors may influence the response much more than in younger children.

When treating pain at any age, it is essential to monitor the response to therapy with an objective scoring system. Pain scores should be regularly recorded on the patient’s vital signs chart.

Table 7.2 A pain scale for preverbal and nonverbal infants (FLACC Scale)

Category	Score		
	0	1	2
Face	No particular expression or smile	Occasional grimace or frown, withdrawn, disinterested	Frequent to constant quivering chin, clenched jaw
Legs	Normal position or relaxed	Uneasy, restless, tense	Kicking or legs drawn up
Activity	Lying quietly, normal position, moves easily	Squirming, shifting back and forth, tense	Arched, rigid, or jerking
Cry	No cry (awake or asleep)	Moans or whimpers, occasional complaint	Crying steadily, screams or sobs, frequent complaints
Consolability	Content, relaxed	Reassured by occasional touching, hugging, or being talked to; distractable	Difficult to console or comfort

Reproduced with permission of Merkel SI et al: The FLACC: A behavioral scale for scoring postoperative pain in young children. Pediatr Nurs 23:392, 1997

Wong-Baker FACES® Pain Rating Scale



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Fig. 7.1 Wong-Baker FACES pain rating scale. (© Reproduced with permission of Dr. Donna L. Wong and Mosby Inc. Wong DL, et al. (eds): *Nursing Care of Infants and Children*, 6th ed. St. Louis, Mosby-Year Book, 1999)

POSTOPERATIVE PAIN

Postoperative pain has adverse physiologic and psychological effects. Optimal postoperative pain relief minimizes the metabolic rate for oxygen, reduces cardiorespiratory demands, promotes early ambulation, and speeds recovery. In addition, postoperative emotional disturbance is reduced if pain is well controlled.

Systemic Analgesic Drugs

After minor procedures, when no regional or local analgesia regimen is possible, the use of a systemic analgesic is indicated. Dosages in common use are listed in Table 7.3. Meperidine is no longer recommended for perioperative analgesia in children because of the potential for seizures associated with its metabolite normeperidine; its only indication is to treat shivering.

The appropriate drug should be chosen for the magnitude of the pain, and a satisfactory effect should be confirmed. It is preferable to administer the first analgesic dose before the child emerges from general anesthesia—for example, for tonsillectomy give 0.025–0.05 mg/kg IV morphine during surgery (if no OSA is present), and for minor superficial surgery give (Perfalgan) acetaminophen, 15 mg/kg IV or ~40 mg/kg PR, after induction of anesthesia. *Do not* cut suppositories of acetaminophen (or any other drug) as the acetaminophen may not be evenly distributed throughout the suppository. Remember that peak blood levels are achieved 60–180 min after rectal administration, so the suppository must be administered immediately after induction and this route is not appropriate for brief procedures; multiple suppositories with several strengths may be simultaneously administered to achieve the desired ~40 mg/kg PR dose. Avoid intramuscular injections in awake children; give analgesics by the intravenous, rectal, or oral route.

Mild Analgesics and NSAIDS

Acetaminophen. Acetaminophen is a mild analgesic and antipyretic drug, but it provides good analgesia and antipyresis after minor procedures, especially if given before the surgery. It is considered safe in neonates, but metabolism and elimination are delayed in neonates compared with adults, so repeat doses should be given at 6- rather than 4-h intervals. Excessive doses can cause hepatic failure; the total

Table 7.3 Common dosages for systemic analgesics

For minor procedures

Acetaminophen, 10–20 mg/kg PO, 30–40 mg/kg PR loading dose followed by 20 mg/kg q6h, maximum 90–100 mg/kg/24 h; 15 mg/kg IV for children (7.5 mg/kg for infants <10 kg)
 Ibuprofen, 5–10 mg/kg PO
 Ketorolac^a, 0.5 mg/kg IM or IV to a max. of 15 mg for <50 kg and 30 mg for >50 kg
 Tramadol^a, 1–2 mg/kg q6h

For major procedures

Morphine, 0.1–0.2 mg/kg IV q2–4 h
 Hydromorphone, 5–15 µg/kg IV q4–6 h
 Oxycodone^a, 0.1–0.2 mg/kg po q4–6 h
 Hydrocodone^a, 0.05–0.1 mg/kg po q4–6 h

^aThese opioids are subject to CYP2D6 polymorphisms that may result in drug overdose (in ultra-rapid metabolizers) or inadequate analgesia (in poor metabolizers)—see text for discussion

daily dose should not exceed 90–100 mg/kg (maximum daily dose for IV acetaminophen in children is <75 mg/kg). *Make sure that clear instructions are given to parents about dosage after the child is discharged.* Hepatic damage has been reported after excessive doses of acetaminophen or when given to debilitated children. After major surgery, acetaminophen combined with opioids reduces the dose of the latter, thereby reducing the risk of respiratory depression. Acetaminophen does not affect surgical bleeding. Oral acetaminophen dosing is 10–15 mg/kg q4–6 h; IV acetaminophen (Perfalgan) is given q6h to infants <10 kg as 7.5 mg/kg and to children 11–50 kg as 15 mg/kg IV. Children >50 kg may receive 1 g IV q6h. IV acetaminophen should be infused over 15–20 min; the dosing in infants should be carefully checked as a number of cases of several-fold overdoses have been reported.

Diclofenac

Diclofenac has been widely used in children and may be administered orally, intravenously or rectally; it is currently under development for parenteral use in children in North America. It is reported to be effective for pain control after minor surgical procedures. Suggested pediatric dosing is 0.3 mg/kg IV, 0.5 mg/kg rectal, and 1 mg/kg oral every 8 h. After tonsillectomy, it reduces the need for opioids and hence reduces PONV. There is greater bioavailability after rectal administration than with the enteric-coated formulation. It does not significantly affect bleeding or clot strength in children after tonsillectomy. The smallest PR dose currently available in the United States is 50 mg.

Ibuprofen

Ibuprofen, an NSAID, may be given by either the oral or rectal route, 4–10 mg/kg every 6 h. It reduces the child's opioid requirements postoperatively. However, ibuprofen can cause gastrointestinal upset (nausea, vomiting, diarrhea) and decrease platelet aggregation, which could increase bleeding.

Ketorolac

Ketorolac is another NSAID; its potent analgesic effects may rival those of morphine without the respiratory depressant effects of the latter. When given before surgery, ketorolac 1 mg/kg IV appears to provide postoperative analgesia comparable to 0.1 mg/kg of morphine; however, this dose is now considered excessive. The recommended IV dose is 0.5 mg/kg to a maximum of 15 mg in children <50 kg and 30 mg in children >50 kg. In common with other NSAIDs, ketorolac inhibits platelet aggregation and is not recommended when bleeding may be a problem. Impaired bone healing after ketorolac remains controversial. Other serious but uncommon potential side effects include gastrointestinal hemorrhage, interstitial nephritis, and acute renal failure. Once hemostasis has been

achieved, it is our practice to ask the surgeon if ketorolac can be administered given its potential negative effects on platelet function and bone healing.

Celecoxib

Recent evidence identified a reduction in the morphine requirements in children undergoing tonsillectomy, after 6 mg/kg oral Celecoxib was administered preoperatively followed by 3 mg/kg q12h. There was no effect on postoperative bleeding. Celecoxib may find a role in perioperative pain management in children.

Opioid Drugs

Morphine. Morphine remains a most useful drug in the management of postoperative pain. It produces effective analgesia together with sedation and a useful degree of euphoria. For children, it is preferably administered intravenously in a dose of 50–100 µg/kg every 7–10 min until comfortable or for ongoing pain, by continuous infusion/PCA (see later discussion).

Codeine. Codeine has been used to treat moderate pain. It may be given intramuscularly or orally, but never intravenously as severe hypotension may result. For most children, the usual dose of codeine is 1–1.5 mg/kg IM or PO (maximum, 60 mg). Codeine has been considered a safe drug for infants and children, but respiratory depression similar to that associated with morphine may occur, especially after repeated doses. There are some populations that carry polymorphisms of CYP2D6 isozyme that may either prevent or reduce the pain relief from codeine or convert codeine to morphine so rapidly that respiratory depression may suddenly occur (see Chap. 3 for details). Codeine is used much less frequently since reports of deaths after tonsillectomy in children with OSA have appeared and the FDA since issued a black box warning on its use after tonsillectomy (see Chap. 3). This drug is no longer recommended for routine use or postoperative analgesia in children.

Hydromorphone (Dilaudid). Hydromorphone is a long-acting opioid analgesic that is 5–7 times more potent than morphine in the IV form. Thus, the IV analgesic dose in children is 5–15 µg/kg. Its elimination half-life is 2.5 h. Hydromorphone may be given as an alternative postoperative analgesic to morphine.

Hydrocodone (0.05–0.1 mg/kg) is also a long-acting oral opioid commonly used for analgesia. This drug is metabolized by CYP2D6 to the active metabolite hydromorphone and subject to the same polymorphism issues as codeine that may result in both drug overdose in children who are rapid metabolizers and minimal analgesia in those who are slow metabolizers. Therefore, we recommend it with caution.

Oxycodone (Oxycontin). Oxycodone (the acetaminophen containing version of oxycontin) is most commonly prescribed as an oral analgesic, but is also approved for IV, IM, and sublingual routes in some countries. For chronic pain,

a sustained release oral preparation is also available. Single doses of oxycodone via all routes are 0.1–0.2 mg/kg every 6 h. It has been used in older children to transition from PCA to oral analgesics. Its side effect profile is similar to that of morphine. However, it too is subject to the problems associated with polymorphisms of 2D6, as the active form of oxycodone is its metabolite, oxymorphone.

Tramadol. Tramadol is a synthetic opioid with limited dosing recommendations in children (from studies outside of North America); a dose of 1–2 mg/kg IV provided good pain relief in children after tonsillectomy and may be useful in children with OSA. It has an elimination half-life of 6–7 h in adults. In North America, it is only available in an oral formulation and indicated only for adults. Tramadol is also subject to variable metabolism due to the CYP2D6 polymorphisms that could potentially lead to accumulation of active metabolites.

Continuous Opioid Infusions

A continuous infusion of morphine, using a dilute solution administered by a designated patient-controlled analgesia (PCA) pump provides for a constant level of analgesia with good sedation and is appropriate for many children after major surgery. The child must have close nursing supervision and be monitored by pulse oximetry. The dose administered should be frequently titrated against the observed and recorded pain level.

Recommended doses for continuous infusions of morphine.

1. Children >1 year: Loading dose, 0.1 mg/kg IV; infusion, 10–30 µg/kg/h
For some children, the loading dose may have to be repeated to establish an initial satisfactory level of analgesia.
2. Infants <1 year: Loading dose, 0.05 mg/kg IV; infusion, 5–15 µg/kg/h
Infants receiving a morphine infusion should be carefully monitored during the infusion and for 24 h after the infusion is discontinued to detect respiratory depression.

Reduced infusion rates may be adequate after cardiac surgery, especially in children who are receiving vasopressors, when the clearance rates for morphine are reduced.

Patient-Controlled Analgesia

Children older than 5 or 6 years of age are capable of using a PCA system to obtain excellent pain relief. Children may especially benefit from PCA; they do not have to ask for pain relief and can be “in control.” Most children are familiar with computer games and have no problem mastering the principles of PCA. It is important that a safe regimen be established and that both child and parents be reassured that the system has an appropriate lockout time and total dosage safeguards. The parents (and other adults) should be warned not to trigger the

Table 7.4 Dosages for patient-controlled analgesia with morphine

<i>For orthopedic surgery</i>	
Initial bolus doses	0.1–0.2 mg/kg IV until settled
PCA bolus dose	10 µg/kg
Lockout period	7–10 min
<i>Background infusion</i>	
For general surgery	0–20 µg/kg/h
For orthopedic surgery	0–25 µg/kg/h
For spinal surgery	0–40 µg/kg/h
Maximum hourly dose	0–100 µg/kg

PCA for the child. Recent evidence, however, suggests that parents or nurses can effectively and safely manage PCA for a child who is unable to do so for age, cognitive, or physical reasons. If such an approach is used, an educational program for the surrogate user must be delivered successfully before they are allowed to participate. *Always be aware that overdose is a potential complication when well-meaning parents are allowed to push the PCA button.*

All children being treated with opioids should have a loading dose. Whether a background infusion is used to supplement boluses is controversial, but in children a continuous infusion of a small dose of morphine complemented with PCA supplements may give the best results in terms of both pain control and sleep pattern.

The regimen used should be tailored to the type of surgery; after orthopedic surgery, children have greater morphine requirements than after general surgery, and after spinal surgery, the requirements are greater still. It is convenient to adjust the background infusion rate to suit the type of surgery (Table 7.4).

Side effects of PCA include the following:

1. *Nausea and vomiting:* This may be troublesome and may require a reduction in the opioid dosage and administration of promethazine (0.25–0.5 mg/kg), ondansetron (0.1 mg/kg up to 4 mg), or other antiemetics. Be aware that promethazine may cause sedation. Low-dose naloxone infusions (0.25 µg/kg/h) have attenuated opioid-associated side effects.
2. *Excessive sedation:* Monitor children carefully and have naloxone ready to treat excessive narcosis and respiratory depression. It may be prudent to keep naloxone and a bag-mask oxygen source at the bedside. Be alert to the possibility that someone who is unaware that the child is receiving a PCA may order a “stat dose” of another analgesic or sedative drug and thereby produce respiratory depression. Write specific orders for children with PCA pumps that they are to have no additional drugs without the knowledge of the PCA team.

Regional Analgesia for Postoperative Pain

The pain that occurs after many procedures can be effectively treated by regional analgesia, and this should be used whenever possible. Frequently, no additional drugs, or at the most, only mild analgesics (e.g., acetaminophen) will be required. Thus, the side effects of opioids are avoided and the child rapidly returns to full activity after minor surgery. These blocks are performed using surface landmarks (caudal epidural block), nerve stimulation, or preferably under ultrasound guidance (Table 7.5). The use of ultrasound increases success rates, decreases dose

Table 7.5 Suggested dosages for epidural/caudal blocks

<i>Bupivacaine</i>	
Loading dose (0.25 %)	0.5 mL/kg
Infusions ^a	
Children	0.4–0.5 mg/kg/h for (0.125 % at 0.3 mL/kg/h) ^c
Infants	0.25 mg/kg/h for (0.125 % at 0.3 mL/kg/h) ^c
Neonates ^b	0.2 mg/kg/h (0.1 % at 0.2 mL/kg/h)
<i>Ropivacaine</i>	
Loading dose (0.2 %)	0.5 mL/kg
Infusions ^a	
Children	0.4 mg/kg/h
Neonates-6 months ^b	0.2 mg/kg/h
PCEA (0.1 or 0.2 %)	
Bolus dose	0.1 mg/kg
Lockout interval	10 min
Background infusion	0.1 mg/kg/h
<i>Levobupivacaine</i>	
Loading dose (0.25 %)	0.5–1.0 mL/kg
Infusion ^a (0.0625–0.125 %)	0.3 mL/kg/h ^c
<i>2-Chloroprocaine</i>	
Loading dose (1–3 %):	10–20 ^d mg/kg
Infusion ^a (1–3 %)	0.3–1 ^d mL/kg/h
<i>Morphine</i>	
Single dose	0.030 ^e (range of 0.01–0.10) mg/kg preservative-free morphine in up to 15 mL preservative-free saline
<i>Fentanyl</i>	
Infusion	1–2 µg/mL @ 0.3 mL/kg/h without local anesthetic
PCEA	
Loading dose	1.4 µg/kg
Bolus dose	0.5 µg/kg
Lockout interval	15 min
Background infusion	0.5 µg/kg/h

^aStart infusion immediately after the loading dose

^bLimit infusions to 48 h

^cNo benefit adding fentanyl to this solution

^dLarger doses should be used with epinephrine and with caution in neonates (max. dose reported is 60 mg/kg)

^eMost effective and commonly used dose; larger doses generally increase side effects

requirements, and reduces the number of needle passes needed. Provision should be made, however, for transition to systemic analgesics after the block dissipates. Studies have shown that significant pain may occur at this time, especially in outpatients. The parent should be carefully instructed to administer an analgesic drug (e.g., acetaminophen or other analgesic) in anticipation of this need.

After major surgery, appropriate nerve blocks (e.g., intercostal nerve block) using local analgesic drugs may reduce the dose of opioids and facilitate earlier mobilization. Here, epinephrine should be considered.

The possibility that a regional block established before the surgical incision (preemptive analgesia) may modulate total postoperative pain by preventing biochemical changes (“windup”) within the central nervous system is appealing, but the results of well-designed studies have been disappointing. Regional blocks performed before the surgical incision do provide intraoperative analgesia, thus reducing the dose of general anesthetic drugs required. This reduction in anesthetic requirement indicates that the block is well established before emergence.

Studies of complications after pediatric regional analgesia procedures strongly suggest that peripheral nerve blocks are associated with fewer complications than neuraxial blocks. Hence, whenever there is a choice, a peripheral nerve block should be performed. Peripheral nerve and neuraxial blocks as well as their complications and treatments are detailed in Chap. 5.

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Neurosurgery and Invasive Neuroradiology

GENERAL PRINCIPLES

1. Perioperative management should be designed to minimize increases in intracranial pressure (ICP) and to ensure optimal operating conditions.
2. Light general anesthesia is adequate for most neurosurgical operations; interventions may be required to prevent or treat increased ICP. Short-acting and rapidly eliminated drugs are preferred to facilitate a prompt emergence from anesthesia to allow accurate and continuous postoperative neurocognitive assessment.
3. Prior infiltration of the scalp incision site with local anesthetic with epinephrine reduces blood loss, blunts responses to the initial incision, reduces the need for anesthetic drugs, and possibly minimizes postoperative pain.
4. Postoperative pain after intracranial surgery must be effectively treated, but respiratory depression must be avoided. For major procedures such as a craniofacial reconstruction, a morphine infusion may be titrated to achieve satisfactory analgesia. For minor procedures, oral opioids, ketorolac, or acetaminophen may suffice.
5. Postoperative systemic hypertension may occur after intracranial surgery, increasing the risk of bleeding and thus increased ICP. Aggressive treatment with antihypertensives may be indicated during emergence from anesthesia and thereafter.
6. Some children may benefit from a period of postoperative-controlled ventilation after major intracranial surgery. This is usually determined after consultation with the neurosurgeon.

Intracranial Physiology and Pathophysiology

1. Cerebrovascular autoregulation maintains constant blood flow to the brain during fluctuations in mean arterial blood pressure (MAP). Autoregulation is present in adults and children at MAPs between 50 and 150 mmHg and in the supine infant between 20 and 60 mmHg.

2. Cerebral blood flow (CBF) in infants and children (90–100 mL per 100 g/min) is greater than in adults (50–60 mL per 100 g/min). CBF varies directly with changes in PaCO₂ between 20 and 80 mmHg. CBF changes approximately 4% per mmHg change in PaCO₂.
3. Vasodilation of normal reactive cerebral vessels reduces blood flow in areas that have lost autoregulation (e.g., arteriovenous malformations (AVMs), vascular tumors, or areas of infection or trauma). This is known as intracerebral steal.
4. Vasoconstriction of normal reactive cerebral vessels has the opposite effect (i.e., inverse intracerebral steal). Hence, hyperventilation as a means for rapidly reducing cerebral blood volume is generally reserved for acute increases in ICP and is not recommended for prolonged periods of time.
5. In older children, the total volume of the intracranial contents is fixed. However, any of its three constituents—blood, CSF, and brain tissue—can increase or decrease if compensated by an equal and opposite change in the volumes of the others (revised Munro-Kelly hypothesis).
6. Infants have a less rigid skull than older children; an increase in the contents may be accommodated to some extent by stretching of the dura, expansion of the fontanel, and separation of the suture lines. ICP may be estimated by palpation of the fontanel.
7. The effect of a space-occupying lesion on ICP depends on its volume and rate of expansion and the rigidity of the skull. Initially, the lesion displaces CSF and/or venous blood from the skull, the sutures may separate in infants, and ICP increases slowly, if at all. As expansion continues, compensation is no longer possible, and small increases in volume cause progressively greater increases in ICP. With a rapidly expanding lesion (i.e., intracranial bleeding), ICP increases rapidly from the outset.

Effects of Specific Anesthetic Drugs on Intracranial Physiology

1. All inhalation agents increase CBF and may increase ICP unless preceded by mild hyperventilation (PaCO₂ ~30–35 mmHg):
 - (a) N₂O may cause a very small increase in CBF but has been used successfully in pediatric neurosurgery for many years. It may increase ICP if air is present within the cranium, and in these circumstances it is contraindicated.
 - (b) The increase in CBF follows the order: desflurane > halothane > isoflurane > sevoflurane.
 - (c) Cerebral autoregulation during changes in arterial BP is blunted as the concentrations of inhalational agents increase, but appears to be preserved at 1 MAC isoflurane and sevoflurane. This emphasizes the

- importance of using moderate concentrations of potent inhalation agents; during which the CBF responses to changes in PaCO₂ are retained. Moderate hyperventilation tends to attenuate or reverse the effects of agents that increase CBF (e.g., halothane, isoflurane, sevoflurane); this effect is more rapid during isoflurane anesthesia. Prior hyperventilation minimizes the increase in ICP with inhalational agents.
- (d) The cerebral metabolic rate for oxygen (CMRO₂) is reduced by halothane, isoflurane, and sevoflurane. Isoflurane and sevoflurane at greater concentrations may even provide some cerebral protection against hypoxia/ischemia.
2. Intravenous anesthetic agents (with the notable exception of ketamine) either have no effect on CBF or decrease it, but if hypercarbia is present, these effects are reversed:
- (a) Thiopental reduces ICP and therefore is an ideal induction agent in neurosurgery. However, it does not prevent increases in BP and ICP during laryngoscopy and intubation; these may be attenuated by prior administration of lidocaine (1–1.5 mg/kg IV) and/or an opioid (e.g., fentanyl 2–5 µg/kg).
 - (b) Propofol reduces CBF and CMRO₂, preserves autoregulation, and may offer some cerebral protection. Induction doses (3 mg/kg) may cause mild hypotension but also more effectively blunt the cardiovascular responses to laryngoscopy and intubation.
 - (c) Remifentanil, fentanyl, and sufentanil have little effect on CBF provided that constant ventilation is maintained. Autoregulation and the cerebrovascular response to PaCO₂ are also maintained. Alfentanil increases CSF pressure in children with cerebral tumors.
 - (d) Ketamine increases CBF and CMRO₂; CSF pressure is increased. This drug should not be used in neurosurgical patients with increased ICP.
 - (e) Midazolam and diazepam decrease CBF, CMRO₂, and ICP and may control seizures. Flumazenil, which antagonizes benzodiazepines, also antagonizes their effects on CBF and ICP. The latter should be used with caution.
3. Non-depolarizing muscle relaxants have no direct effect on CBF. (Vasodilation resulting from histamine release after atracurium is a possible exception.) The duration of action of vecuronium and rocuronium may be reduced in children taking chronic anti-seizure medications.
4. Succinylcholine may transiently and very slightly increase CBF and ICP in children with space-occupying lesions; this response may be attenuated by prior administration of a small dose of a non-depolarizing muscle relaxant.

Hyperkalemia has been reported after succinylcholine in children with cerebral trauma and other central nervous system diseases, including paraplegia, encephalitis, and subarachnoid hemorrhage.

5. Sodium nitroprusside (SNP), nitroglycerin, adenosine, and the calcium channel-blocking drugs impair cerebral autoregulation and may increase CBF and ICP.
6. Dexamethasone (0.15 mg/kg IV to a maximum of 8 mg) may decrease focal cerebral edema.
7. If an independent vasodilator effect is absent, drugs that decrease neuronal function decrease CBF (such as thiopental).
8. Drugs that increase neuronal function increase CBF (such as ketamine).
9. Somatosensory-evoked potentials (SSEPs) to monitor brain or spinal cord function are attenuated by inhalational anesthetics if these agents are given in more than minimal concentrations. Nitrous oxide, propofol, opioids, and muscle relaxants have little effect on SSEPs.
10. Motor-evoked potentials (MEPs) to monitor brain and spinal cord function are much more easily depressed by inhalational anesthetics than SSEPs. If MEP monitoring is planned, nitrous oxide and muscle relaxants should be withheld after intubation and inhalational agents limited to 0.5 MAC. Propofol, alpha-2 agonists, benzodiazepines, and opioids do not significantly compromise MEP monitoring.

Neuroapoptosis

NMDA receptor antagonists and GABA_A receptor agonists induce apoptosis (programmed cell death) in several regions of the brain in newborn animals. Most anesthetics (except α_2 agonists and opioids) induce apoptosis that is more severe if administered in combination and for prolonged periods. Additionally, these anesthetics cause structural and neurocognitive functional changes to the nervous system in animals after anesthesia. Curiously, other medications including acetaminophen, dexamethasone, and carbon dioxide have been shown to cause apoptosis. However, the positive predictive value for injury in humans is less than 10%. A number of salutary interventions after the injury attenuate the injury in newborn animals including exercising and socializing, which raise doubts about the translational nature of these data. The evidence that young children who received anesthesia may develop neurocognitive dysfunction is conflicting. Prospective studies to date have failed to demonstrate neurocognitive dysfunction in young children after general anesthesia. In the interim, parents should balance the known risks and benefits of proceeding with anesthesia and surgery.

ANESTHESIA MANAGEMENT

Premedication

Children with increased ICP should not receive doses of drugs that depress ventilation, prolong recovery, or hamper postoperative assessment. Therefore, with one exception (see below), do not give sedative premedication to those undergoing craniotomy. If IV access has not been established, topical local anesthetic or 50% nitrous oxide will facilitate pain-free insertion. Some children may benefit from a small dose of oral midazolam for anxiolysis before surgery, but they should be closely observed. Children with normal ICP who are undergoing elective or non-cranial surgery (e.g., laminectomy) may be premedicated as usual.

Exception. Children with a vascular aneurysm or AVM, especially if there is a history of hemorrhage, may benefit from being well sedated preoperatively in order to minimize changes in venous and arterial pressures with crying or stress at induction of anesthesia or tracheal intubation.

Induction of Anesthesia

Anesthesia should be induced aiming to minimize changes in ICP and fluctuations in arterial and venous pressures. Preoxygenation followed by an intravenous induction using thiopental or propofol followed by a muscle relaxant to facilitate tracheal intubation thereby ensuring optimal ventilation, is preferred. Lidocaine (1–1.5 mg/kg IV) and fentanyl (2–5 µg/kg) may be given 3 min before intubation to prevent increases in ICP associated with laryngoscopy and tracheal intubation.

Anesthesia for children with vascular anomalies should be induced as above but should then be deepened with an inhalation agent using controlled ventilation to prevent hypercapnia and to avoid hypertension and tachycardia during laryngoscopy, intubation, and placement of neurosurgical devices for securing the head.

Some children for emergency surgery will have a full stomach. They require a rapid sequence induction using succinylcholine or high-dose rocuronium (1.2 mg/kg) with all precautions to prevent regurgitation and aspiration.

For surgery in the prone position and for other procedures that entail changes in position, a nasotracheal tube is preferred. An orotracheal tube may kink in the prone patient or become dislodged if saliva loosens the adhesive tape; a nasotracheal tube is less likely to dislodge and is easier to secure at a specific depth in the trachea. In older children, a reinforced oral tube may be used. It should be secured using tape and benzoin. Administering glycopyrrolate may reduce

drooling. Bilateral ventilation should be checked after the tube is taped and the child is positioned; remember that flexion of the head (as in posterior fossa surgery) pushes the tip of the tracheal tube toward the carina whereas extension pulls the tracheal tube cephalad. (N.B. To determine whether an endobronchial intubation will occur after positioning prone, while the child is supine, flex the neck after the tube is taped. If wheezing or an endobronchial intubation is detected, untape the tube, withdraw it, and then retape it. Repeat the maneuver to ensure the tip of the tube is properly positioned.)

Sudden preoperative apnea may occur in neurosurgical patients awaiting surgery and may indicate an acute increase in ICP. If this occurs, hyperventilate the lungs with 100% oxygen and ask the surgeon to tap the CSF immediately.

Maintenance

If inhalational anesthetics are used, administer the smallest concentrations compatible with adequate anesthesia (to minimize increases in ICP). They may be accompanied by N₂O, muscle relaxants and mild-moderate hyperventilation. Otherwise, N₂O together with short-acting opioids (e.g., fentanyl, remifentanil, sufentanil), which ensure rapid postoperative recovery, may be preferred. Deep anesthesia is unnecessary. Propofol infusions may be a useful alternative in some children, particularly toward the end of a prolonged procedure when other drugs have been discontinued to facilitate a rapid emergence.

Ventilation

Limited controlled hyperventilation (PaCO₂ approximately 30–35 mmHg) may be used to decrease brain bulk and ICP during intracranial surgery.

Monitoring

The child should be monitored as follows:

1. Pulse oximeter, automated BP cuff, and ECG.
2. PetCO₂ monitor: this is useful both as a guide to the adequacy of ventilation and as a means of detecting air embolism.
3. Continuous recording of body temperature (esophageal or rectal).
4. Esophageal stethoscope.

5. For major neurosurgery, arterial and central venous access should be considered. Arterial access is useful to rapidly assess fluctuations in BP because of traction on neural tissues, blood loss, or air embolism and for laboratory testing. Central venous access will help assure a stable circulating blood volume, provide a means for administering vasoactive drugs (dilator or inotrope), and provide a possible route for aspirating embolized air. Central venous access is best sited in the subclavian (or femoral) vein to prevent misleading readings and occluding neck/intracranial veins. Rapid blood transfusions should not be given into CVP lines in small infants as cold, hyperkalemic blood may lead to cardiac arrest.
6. Measure urinary output during all major neurosurgeries, especially if mannitol or other diuretics will be given.
7. A precordial Doppler flowmeter should be used for operations with potential for air embolism. These include those performed with the child in the sitting or head-up position and all major cranial reconstructions (including cranioplasty for craniosynostosis). The Doppler probe should be placed over the right atrium (second right interspace adjacent to the sternum).
8. If neurophysiologic monitoring is planned (SSEPs, MEPs, or EEGs), ensure that the anesthetic prescription is consistent with producing optimal signals. Important considerations are:
 - (a) The depth of anesthesia should remain constant to compare sequential recordings.
 - (b) Ventilation (PaCO_2) and oxygenation should remain constant.
 - (c) Body temperature should remain constant.
 - (d) Opioids do not affect any neurophysiologic monitoring.
 - (e) Muscle relaxants do not affect SSEPs but are contraindicated in MEPs.
 - (f) Nitrous oxide has little effect on latency but depresses the amplitude of SSEPs; it is contraindicated for MEPs.
 - (g) Inhalational anesthetics generally increase latency and depress amplitude of SSEPs; they are limited to ≤ 0.5 MAC for MEPs.
 - (h) Propofol, ketamine, midazolam, and α_2 -agonists exert no significant effects.

In practice, a prescription using 0.5 MAC of an inhalational agent in an oxygen/air mixture, an opioid infusion (remifentanil, fentanyl, or sufentanil), midazolam, and a propofol infusion provides acceptable conditions for monitoring both SSEPs and MEPs. We prefer *cis*-atracurium for tracheal intubation when MEPs are planned (if succinylcholine is used and the child has pseudocholinesterase deficiency, MEPs may not be detectable). If remifentanil is used, administer a long-acting opioid to control postoperative pain before discontinuing the remifentanil.

INTRAVENOUS FLUID THERAPY AND CONTROL OF INTRACRANIAL PRESSURE

Strategies for Intravenous Fluid Therapy

A reliable intravenous cannula is essential for children undergoing neurosurgery; at least 22- or 20-gauge cannulas for infants and 18-gauge or larger cannulas for older children. Small infants undergoing major surgery should have at least two large and well-secured IV routes. (Exsanguination during neurosurgery can occur in small infants and happens rapidly!)

General Rules for Intravenous Therapy

1. Maintain the intravascular volume but avoid excessive fluid administration; third-space losses are very small in neurosurgical patients.
2. Avoid hypo-osmolar fluids because they increase brain edema; use normal saline. Syndrome of inappropriate antidiuretic hormone (SIADH) may follow neurosurgical procedures and result in hyponatremia; the use of hypotonic solutions increases this danger. Monitor plasma electrolytes postoperatively.
3. Avoid dextrose-containing solutions except for documented hypoglycemia. Dextrose administration may increase the risk of neurologic damage secondary to local ischemia, including that caused by surgical retraction of the brain. If hypoglycemia is a concern (i.e., in infants), plasma glucose concentrations should be measured periodically during surgery and a glucose-containing solution administered using an infusion pump to maintain normoglycemia.
4. Blood losses are difficult to measure intraoperatively; use cardiovascular indices (e.g., heart rate, BP, contour of the arterial wave form, and CVP) to guide the volume of fluids to administer. Colloid solutions and/or blood should be administered as indicated for extensive losses (see later discussion).

Control of ICP and Reduction of Brain Volume

The anesthetic prescription for neurosurgery should ensure that the surgeon has optimal intracranial operating conditions. This can be ensured as follows:

1. Prevent any episodes of hypoventilation or hypoxemia, coughing or straining, during induction of anesthesia.
2. Provide a clear, unobstructed airway at all times as increases in intrathoracic and airway pressures are directly reflected in ICP. The largest tracheal tube that will pass easily should be used. It should be positioned so that there is no possibility of kinking, compression or impinging on the carina. Reinforced tubes are preferred where possible.

3. Provide mild hyperventilation to a PaCO_2 of approximately 30–35 mmHg.
4. A slight head-up tilt is commonly used (15°). The veins in the neck should be totally unobstructed; avoid extreme neck rotation.
5. Administer furosemide 0.5 mg/kg IV followed by mannitol 20% (0.5–1 g/kg) infused over 20–30 min as the skull is being opened (or as requested by the neurosurgeon).

After administering a diuretic, the schedule of fluid therapy also depends on the urine output. When urine volume equals 10% of the estimated blood volume (EBV), further urine losses are replaced (volume for volume) with normal saline. Alternatively, fluid administration can be guided by the CVP; maintaining a constant CVP generally indicates a stable circulating blood volume. Subsequently, serum electrolyte concentrations should be determined to exclude abnormalities and guide fluid replacement.

Blood Replacement

Because blood loss during neurosurgery cannot be measured accurately, it must be gauged clinically from observation of the amount of bleeding and measurement of the child's cardiovascular indices and hematocrit. The systolic BP must be monitored as it is the most valuable guide to volume status; fluid replacement should maintain it at 60 mmHg in infants and 70–80 mmHg in older children. (N.B. The latter may lose up to 20% of EBV without a significant decrease in blood pressure). When surgery is complete but before the dura is closed, sufficient colloid or crystalloid should be given to return the arterial BP to the pre-“blood loss” level. The decision to transfuse blood is based on determination of the hematocrit together with clinical judgment of the blood losses occurring in relation to the allowable blood loss.

If a major blood transfusion has occurred, particularly in small infants, serum Ca^{++} should be measured. Hypotension unresponsive to volume replacement should be treated with parenteral calcium gluconate (30–45 mg/kg IV) or chloride (10–15 mg/kg IV). Assess coagulation indices and replace clotting factors as indicated.

Controlled Hypotensive Techniques

Controlled hypotensive anesthesia is rarely used in children, even in those with large AVMs or aneurysms, because these neurovascular defects are more often managed endovascularly rather than through a craniotomy. However, if controlled hypotension is planned, an arterial line is essential. A safe range of mean arterial pressure in the supine position is 50–65 mmHg in children up to

10 years of age and 70–75 mmHg in older children. The arterial transducer must be zeroed at the level of the head in order to accurately reflect cerebral perfusion pressure.

Drugs to Induce Hypotension

1. *Isoflurane or sevoflurane.* The inspired concentration can be increased until the desired BP is achieved. This results in very stable BP levels but is not readily reversed.
2. *Sodium nitroprusside* has been widely used to induce hypotension but may result in tachyphylaxis, often results in wide swings in BP and may cause toxic sequelae if large doses are used. Because SNP interferes with cerebral autoregulation and may increase ICP, it should not be infused until the skull is opened.
3. Remifentanil is an excellent choice for controlled hypotension since large doses provide moderate reductions in BP that are not associated with toxicity which are readily reversed by slowing or stopping the infusion. However, its onset of hypotension is slower than with SNP.

Venous Air Embolism

Venous air embolism (VAE) is a particular hazard when surgery is performed with the child in the sitting position, but it may also occur when the child is prone or supine if the head is elevated. It is relatively common during craniosynostosis surgery and has also occurred during laminectomy. Air may be drawn in rapidly if a venous sinus is entered, or it may trickle in through veins within the bone. VAE detected by Doppler ultrasonography has a similar incidence in children and in adults, but is more likely to produce cardiovascular instability in children.

VAE most often occurs during opening of the skull, but may also occur at the time of skin closure when the skin clips are removed. The signs, in order of decreasing sensitivity, include:

1. Changes in Doppler ultrasound over the precordium or appearance on trans-esophageal echocardiogram
2. Sudden decrease in PetCO₂ (or increase in end-tidal nitrogen level)
3. Hypotension
4. Change in heart sounds (windmill murmur or muffled/muted heart tones)

Early diagnosis and rapid therapy are required to prevent a serious outcome:

1. Inform the surgeon, who should compress and/or flood the wound with saline to prevent entrainment of additional air.
2. Lower the head to increase the venous pressure at the wound and augment venous return from the legs. If the child is prone but in a head-holding device,

the wound should be covered and consideration given to releasing the head and turning the child supine to perform chest compressions if needed.

3. Compress the jugular veins in the neck.
4. Ventilate with 100 % O₂, discontinue N₂O to prevent further expansion of air emboli within the bloodstream, and add 5–10 cm H₂O PEEP.
5. Attempt to aspirate air via the central venous catheter; this is successful in fewer than 60 % of cases.
6. Initiate cardiopulmonary resuscitation and other measures (e.g., inotropes) as required. *N.B.* measureable PetCO₂ > 10 mmHg indicates adequate chest compressions.

Postoperative Considerations

All children should be fully recovered from the effects of anesthetic drugs and awake at completion of the procedure. Extubation should be smooth, without coughing or bucking; this can be facilitated by giving lidocaine 1.5 mg/kg IV. If the child remains unresponsive or respirations are depressed, leave the tracheal tube in place and control ventilation until the cause is determined. After some major neurosurgery, it may be preferable to continue controlled ventilation into the postoperative period and extubate the trachea later, particularly if the surgery was in proximity to structures that control ventilation. Monitor BP closely during emergence; antihypertensives may be required.

An MRI is often performed immediately postoperatively to ensure intracranial bleeding has not occurred: this may follow directly from the OR before emergence and extubation or be scheduled later from the neurosurgical ICU. If the latter is planned, the child should remain intubated until the MRI is completed.

Postoperative nursing care includes neurologic monitoring. Assess fluid status as regulatory mechanisms (i.e., antidiuretic hormone levels) may be altered after craniotomy.

HYDROCEPHALUS

Hydrocephalus may be caused by a congenital defect (e.g., Arnold-Chiari malformation, aqueductal stenosis) or by acquired disease (e.g., hemorrhage, infection, tumor). In the neonate, hydrocephalus may be secondary to the Arnold-Chiari malformation. (In many cases, Chiari malformation is accompanied by meningomyelocele; this combined defect is present in 1–3 of every 1000 live births), although the latter defect has dramatically decreased with the use of prenatal folic acid supplements.

Surgical Procedures: Creation of Cerebrospinal Fluid Shunts

For noncommunicating hydrocephalus (see Chap. 2), the following procedures are performed:

1. Ventriculo-peritoneal (VP) shunt (lateral ventricle to peritoneum)—most common and preferred, because it allows the most room for growth
2. Ventriculo-atrial shunt (lateral ventricle to right atrium)—still used occasionally but may lead to long-term complications, particularly pulmonary thromboembolism and cor pulmonale because of pulmonary hypertension
3. Ventriculo-pleural shunt (lateral ventricle to pleural cavity)—rare
4. Fourth ventriculostomy

For communicating hydrocephalus (see Chap. 2), a lumboperitoneal shunt (lumbar subarachnoid space to peritoneum) is performed.

Endoscopic instruments are often used to position shunts in the lateral ventricles, for fourth ventriculostomy, and for making a connection between ventricles. Very occasionally, these endoscopic procedures may be accompanied by considerable bleeding. Alternatively, shunts may be positioned under ultrasound guidance.

Special Anesthesia Problems

1. Increased ICP may be present and is sometimes severe. Assess the child for vomiting and dehydration; check fluid and electrolyte status preoperatively. Occasionally, acute signs of increased ICP demand immediate surgery.
2. Children may have had repeated anesthesia for shunt revisions; check the previous records for difficulties with venous access, intubation, technical problems with successful shunt positioning, and latex allergy.
3. Blood loss is usually minimal, but very occasionally bleeding occurs from a large vessel. Always be prepared with large and secure intravenous access.
4. The child should be fully awake at the end of the procedure to permit rapid neurologic assessment.
5. Latex precautions should always be used in these children.

Anesthesia Management

Preoperative

1. Exercise special care if the ICP is increased. The child should be continuously monitored until surgery because the child's condition can deteriorate suddenly (e.g., becoming apneic and unresponsive), necessitating tracheal intubation, hyperventilation, and immediate ventricular tap or lumbar puncture (depending on whether the hydrocephalus is noncommunicating or communicating).
2. Assess fluid status. Administer IV fluids to maintain normovolemia.

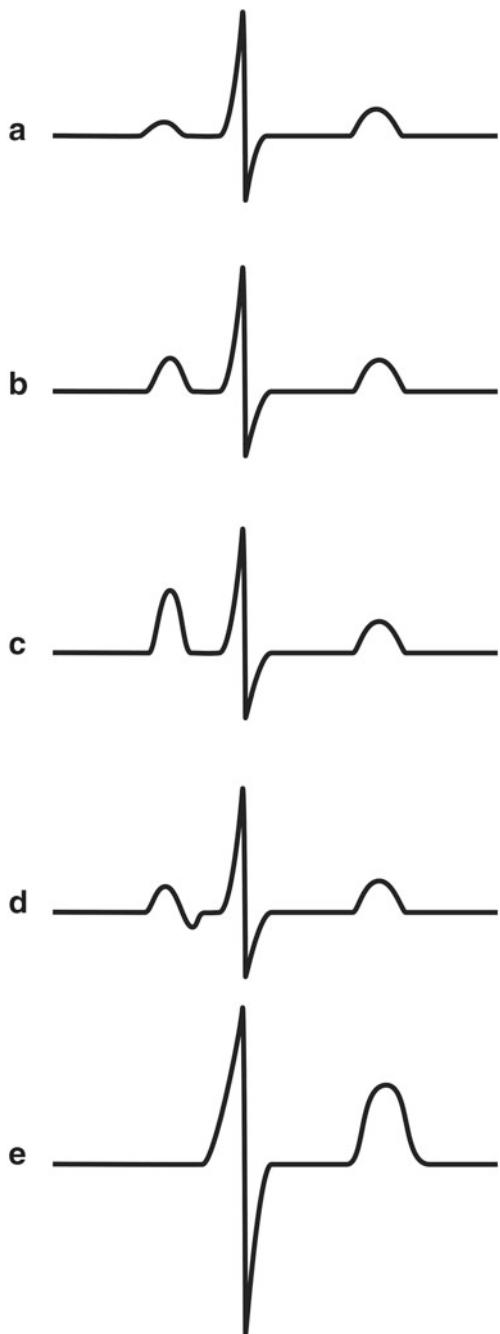
Perioperative

1. Induce anesthesia while avoiding hypoventilation, hypoxia, increased central venous pressure, and systemic hypertension:
 - (a) An intravenous induction with propofol or thiopental plus a muscle relaxant is preferred in order to secure the airway rapidly and control ventilation.
 - (b) Lidocaine (1.5 mg/kg IV) and fentanyl (2–5 µg/kg) may be given approximately 3 min before induction to attenuate the hypertensive response to laryngoscopy and intubation.
2. During surgery, maintain anesthesia with N₂O and low concentrations of isoflurane or sevoflurane. Non-depolarizing relaxant drug (*cis*-atracurium preferred for short cases) permits the use of minimal inhalational agent, with rapid recovery when antagonized. Controlled ventilation is preferred.
3. Note the following situations:
 - (a) Hypotension at the time of CSF tap. If the arterial BP was increased secondary to increased ICP and if inhalational agents have been given, the BP may decrease precipitously as the ICP returns to normal (i.e., at the time of CSF tap). Discontinue all anesthetic agents, ventilate with 100% O₂, and give a fluid bolus to restore the BP to a normal level. Pressors are rarely needed.
 - (b) Bradycardia and other arrhythmias may occur after or during placement of the intraventricular catheter, probably as a result of shifts in intracranial contents.
 - (c) VA shunts. Apply controlled positive pressure ventilation to prevent VAE while the vein is open for insertion of the cardiac end of the VA shunt. The ECG may be used as a guide for positioning the atrial end of the shunt. Fill the shunt tubing with hypertonic saline and attach it by a sterile extension wire to the left-arm ECG lead. Switch the ECG to lead III. Advance the tubing; as the tip approaches the right atrium the height of the P waves increase, and when it reaches its correct position in the atrium, they become small and biphasic (Fig. 8.1).
4. Blood loss is usually minimal; secure large-bore intravenous access.
5. Inject local anesthetic to block the supraorbital nerve for a frontal shunt and a block of the posterior auricular nerve for posterior shunts.
6. Discontinue anesthetic agents and extubate the airway when the child is completely awake and responsive before leaving the OR.

Postoperative

1. Routine post-craniotomy nursing care.
2. Analgesics as required. NSAIDs are often sufficient, but morphine may be required; if local analgesic drugs have been injected at the surgical sites, the need for systemic analgesics is usually minimal.

Fig. 8.1 Drawing of ECG tracings obtained as a VA shunt catheter is advanced toward the heart. (a) When the catheter tip is in the SVC, P waves become large as the tip approaches the right atrium (b, c), then smaller and biphasic as the atrium is entered (d). If the catheter is advanced too far into the ventricle, the QRS complexes become very large and the P waves disappear (e)



CRANIOSYNOSTOSIS

Premature fusion of a suture between bones of the cranial vault leads to this skull deformity (incidence 1:2000 births). Simple (single suture) in 75 % of cases or complex or syndrome (multiple sutures) in 25 %, most frequently only the sagittal suture is involved, leading to a cosmetic deformity. Fusion of more than one suture may lead to increased ICP and later to developmental delay and possibly optic atrophy. Early surgical repair (at <6 months of age) gives improved cosmetic results with less blood loss than repair at an older age.

Associated Conditions

1. Craniofacial abnormalities such as Crouzon disease and Apert syndrome, may be associated with OSA. See Appendix A for other syndromes and associated conditions.

Surgical Procedure

1. Open craniectomy—division of skull along suture lines
2. Endoscopic craniectomy
3. Spring-assisted strip cranioplasty—less blood loss, fewer hospital days

Special Anesthetic Problems

1. Possible increased ICP.
2. Sudden massive blood loss from damaged cerebral venous sinuses. Continued bleeding owing to the vascularity of the scalp and other membranes.
3. Difficult airway in children with craniofacial syndromes.
4. VAE is a very real potential intraoperative danger.
5. Take extra care to protect the eyes in those with proptosis.

Anesthetic Management

Preoperative

1. Packed red blood cells should be immediately available in the OR.
2. Use caution with premedication if the ICP is increased.

Perioperative

1. Intravenous induction with propofol (or thiopental) and a muscle relaxant is optimal to rapidly secure the airway and prevent hypoventilation in infants with increased ICP, but a standard inhalation induction may be used in

infants without an increased ICP. A nasotracheal tube is preferred for infants; otherwise, use an armored oral or RAE tube (if supine). Choanal atresia is occasionally associated with craniosynostosis.

2. Maintain anesthesia with N₂O/O₂, small concentrations of sevoflurane or isoflurane and a relaxant; control ventilation.
3. Establish a secure large-bore intravenous line and have blood checked in the OR. Monitor blood loss at scalp incision. An arterial line should be inserted if extensive surgery is planned (always discuss the surgical plan and the potential for rapid massive blood loss with the surgeon).
4. Mild hyperventilation and possibly diuresis to control ICP.
5. Use blood conserving measures. Antifibrinolytics may reduce blood loss: tranexamic acid (multiple regimens, e.g., 15 mg/kg IV loading then 10 mg/kg/h) or epsilon aminocaproic acid (75–100 mg/kg loading dose then 15 mg/kg/h IV infusion) until skin closed. Controlled hypotension is eschewed if increased ICP may be present.
6. A precordial Doppler probe to monitor for VAE with skull opening. Treat VAE as previously outlined.
7. Use low-dose opioids (e.g., 50 µg/kg morphine IV).
8. Extubate the airway when the child is wide awake.

Postoperative

1. Routine post-craniotomy nursing care.
2. Exercise caution when prescribing opioids. This procedure is not as painful as it seems especially if local anesthetic was used. Moreover, children with Crouzon disease may have OSA and be sensitive to the depressant effects of usual doses of opioids.
3. Ensure that an open cranium is appropriately protected and padded.

MYELODYSPLASIA: MENINGOMYELOCELE AND ENCEPHALOCELE

Meningomyelocele and encephalocele result from failure of the neural tube to fuse in the fetus. The incidence of meningomyelocele is approximately 1–4 per 1000 live births, with a large geographic variation. Encephalocele is much less common. Early operation should be performed because of the risk of infection and to prevent further damage to nerve tissue. Extensive skin dissection to mobilize flaps may be needed in some cases, and this may result in considerable blood loss.

Associated Conditions

Hydrocephalus, in many cases with Arnold-Chiari malformation and aqueductal stenosis, occurs in 80 % of infants with meningomyelocele or encephalocele.

Short trachea has been described in association with meningomyelocele. Inspiratory stridor may be present secondary to cranial nerve dysfunction causing vocal cord palsy.

Surgical Procedures

Excision of the sac and repair of the defect is usually performed as soon as possible after birth.

Special Anesthesia Problems

1. Potential difficulty in positioning the child for intubation; placing the defect in the middle of a “doughnut” will prevent pressure on the open defect but also necessitates additional padding beneath the shoulders and head. Beware that children with meningomyelocele may also have a short trachea. *Ensure that the tracheal tube is not endobronchial.*
2. Blood loss is difficult to measure and may be considerable.
3. Body temperature may be difficult to maintain during surgery. A forced air warmer is most effective for maintaining normothermia.
4. Postoperative stridor, ventilatory depression or apnea, or cranial nerve palsy is possible; ventilatory control may be abnormal in infants with meningomyelocele.

N.B. Succinylcholine may be used in these infants; it does not cause hyperkalemia in infants and children with myelomeningocele.

Anesthesia Management

Observe all special precautions for the neonate.

Preoperative

1. Cover the lesion with a sterile dressing.
2. Ensure that cross-matched blood is available in the OR.
3. Ensure that the OR has been warmed to at least 24 °C.

Perioperative

1. Use all modalities for thermal homeostasis (see Chap. 4).
2. Induce anesthesia by the inhalational or intravenous route. Secure the airway with a single dose of IV propofol (3 mg/kg) and/or rocuronium (0.3 mg/kg). This dose of relaxant will wear off by the time the nerve stimulator is needed.

Administer additional muscle relaxant only after the surgeon has confirmed that a nerve stimulator will not be required to identify nerve roots. In cases of encephalocele, laryngoscopy and intubation may be easier if the child is positioned in the left lateral decubitus, with an assistant applying forward pressure at the back of the head and backward pressure on the shoulders to prevent neck extension. If intubation cannot be performed in this position, place the infant supine, supported on a ring cushion to protect the spinal cord defect. Check the position of the tube.

3. Continue anesthesia with N₂O and isoflurane or sevoflurane, with controlled ventilation. An arterial line should be inserted if surgery is expected to be extensive.
4. For occipital defects, the infant is positioned prone on bolsters. Assure that all pressure points are adequately padded and the eyes are protected.
5. Blood loss cannot be measured accurately. Estimate the amount of bleeding and monitor the systolic BP and the hematocrit as guides to volume replacement.

Postoperative

1. Return the infant to a warm incubator, to be nursed prone.
2. Instruct the nursing staff to observe closely for signs of increased ICP, especially in cases of encephalocele. Monitor the adequacy of ventilation and for the development of stridor.
3. Do not give opioids until it is determined that there is sensory innervation of the surgical site.
4. Check hemoglobin and hematocrit on arrival in the postanesthesia care unit.

ARNOLD-CHIARI MALFORMATION

The Arnold-Chiari malformation consists of an elongated cerebellar vermis that herniates through the foramen magnum with associated compression of the brain stem. Infants with this disease may have difficulty swallowing, recurrent aspiration, stridor, and possible apneic episodes. The gag reflex may be depressed or absent.

Associated Conditions

Syringomyelia is often associated and leads to arm weakness and possible sensory deficit.

Surgical Procedure

The surgical procedure consists of decompression of the posterior fossa, enlargement of the foramen magnum with upper cervical laminectomy, opening of the dura, and lysis of adhesions. If syringomyelia is present, it is treated by drainage of the hydromyelia.

Special Anesthesia Problems

1. Control of ventilation is abnormal; stridor may require preoperative intubation and postoperative apnea may occur.
2. Recurrent aspiration frequently impairs pulmonary function, complicating the ventilatory status.
3. Stridor may not always improve immediately after surgery.
4. The child must be positioned prone and with the neck flexed for surgery; check the position of the nasotracheal tube before and after turning the child prone (see previous discussion).

Anesthetic Management

Anesthesia management is the same as for posterior fossa exploration. The child should be monitored postoperatively, preferably in an ICU or neurosurgical unit. Rarely, the tracheal tube must be left in place postoperatively if the child cannot protect the airway. If intubation is prolonged, tracheostomy may rarely be required.

CRANIOTOMY FOR TUMORS AND VASCULAR ANOMALIES

Intracerebral tumors are relatively common during childhood, with a peak incidence at the age of 5–8 years; about 60 % are in the posterior fossa. Benign vascular lesions may also require craniotomy.

Surgical Procedures

1. Exploratory biopsy and/or excision of lesion.

Special Anesthetic Problems

1. Increased ICP and/or hydrocephalus may be present and may result in nausea, vomiting, and electrolyte disturbance.
2. Anesthesia techniques must be designed to provide optimal intracranial conditions for surgery.

3. Blood loss may be rapid, massive, and difficult to measure accurately. Appropriate invasive monitoring and peripheral venous access will be required.
4. Small infants with AVMs may have associated high-output congestive heart failure. These infants have a low diastolic blood pressure and do not tolerate further reduction in blood pressure intraoperatively (cardiac arrest may occur). Anesthesia in older children with an aneurysm or AVM includes prevention of intraoperative hypertension; controlled hypotension may be required to facilitate the surgery.
5. Postoperatively, the child must be free of residual effects of anesthesia to permit accurate neurologic assessment and monitoring. Hypertension must be aggressively treated to prevent bleeding.
6. In a minority of cases, it may be necessary to perform intraoperative neurophysiologic studies; in others, it may be necessary to record cortical SSEPs. Refer to the neurophysiologic monitoring section for an anesthetic prescription.
7. In very few cases, the child must remain awake and cooperative during surgery (i.e., to map the speech area) after a brief period of deep sedation during the craniotomy.

Anesthesia Management

Preoperative

1. Review and understand the pathology and the surgical procedures that will be required. Check laboratory results.
2. Order packed red blood cells to the OR (at least 1000 mL for craniotomy and more for removal of vascular anomalies).
3. Do not give sedative premedications except to children with vascular lesions (see previous discussion). Establish rapport with older children, reassure them, and explain the planned procedures.

Perioperative

1. Induce anesthesia, preferably using intravenous propofol (2–3 mg/kg) (or thiopental 5–7 mg/kg), lidocaine 1.5 mg/kg, fentanyl (2–5 µg/kg), and a full dose of a muscle relaxant; this permits the airway to be secured rapidly and prevents hypoventilation, hypoxia, coughing, or straining, and adverse hemodynamic responses associated with tracheal intubation.
2. Intubate using the largest tracheal tube that passes the larynx easily. A nasotracheal tube may be preferred for small infants and for those in whom postoperative ventilation may be required.

3. Light general anesthesia is adequate for neurosurgery; little further opioid analgesia is required once the skull is open. Control ventilation with intermittent doses of intermediate-acting relaxants while monitoring neuromuscular block; antagonism should be readily accomplished. Alternatively, an infusion of muscle relaxant while monitoring the train-of-four response can maintain a stable degree of neuromuscular blockade (1–2 twitches) for many hours and simplifies management. For example, commence with 10 µg/kg/min rocuronium and increase or decrease the rate by 2–3 µg/kg/min until the desired steady state has been achieved.
4. For vascular tumors and vascular anomalies, an arterial line and CVP may be inserted (see previous discussion). A urinary catheter may also be used.
5. Encourage the surgeon to infiltrate the scalp with 0.125% bupivacaine with epinephrine 1:200,000 (maximum volume, 2.0 mL/kg) to minimize blood loss and pain.
6. The surgeon may request a diuretic to reduce cerebral edema.
7. Give dexamethasone 0.15 mg/kg IV (maximum dose, 8 mg) to minimize focal cerebral edema.
8. If extubation is planned at the end of surgery, extubated in the OR when the child is fully awake and responsive.

Special Considerations: Anterior and Middle Fossa Surgery

1. Use an armored orotracheal tube or a nasotracheal tube.
2. Position the child with a 15° head-up tilt.
3. Maintain anesthesia and monitor as previously described.
4. Arrhythmias or changes in BP may occur during dissections in the region of the pituitary gland and hypothalamus. If these occur, alert the surgeon to discontinue surgery until the situation resolves. Intravenous atropine may be required for bradycardia.

Special Considerations: Posterior Fossa Surgery (Prone Position)

1. Use a nasotracheal tube in small children. It can be precisely secured at the nostril, it is less likely to kink than an oral tube, and its taping is unlikely to be loosened by saliva. Assess the risk of an endobronchial intubation when the neck is flexed before turning prone.
2. The child should lie prone on a frame (e.g., Relton frame) or bolsters, with a 15° head-up tilt and with the thorax and abdomen hanging free.
3. Anesthetize as for anterior or middle fossa surgery (see previous discussion). Monitor vital signs particularly during manipulations in the region of the brain stem.

Special Considerations: Posterior Fossa Surgery (Sitting Position)

Many pediatric neurosurgeons use the prone or park-bench position, but unfortunately some still prefer the sitting position:

1. VAE is a prime concern; monitor with a precordial Doppler probe and capnograph and consider placing a CVP line at the junction of the superior vena cava and right atrium. The CVP line can be used to aspirate air in case of embolism (limited effectiveness) and also as a guide to fluid therapy.
2. Zero the arterial transducer at the level of the ear and the CVP transducer at the level of the heart. Cardiovascular stability is the next problem; avoid compression of vena cava and obstruction of venous return.

Postoperative

1. The child must be fully recovered from the effects of anesthesia before leaving the OR.
2. Routine post-craniotomy nursing monitoring and care.
3. Order small doses of opioids to avoid respiratory depression; a morphine infusion at an appropriately low rate is usually very effective. *N.B.* The child is often also given an anti-seizure medication.
4. Body temperature may increase; cool as necessary to maintain normothermia.
5. SIADH may occur with resulting oliguria and electrolyte disturbance.

CRANIOPHARYNGIOMA

This is the most common pituitary tumor in children, accounting for 5–10 % of childhood brain tumors; its incidence peaks in childhood between 5 and 14 years of age. It may become very large and compress adjacent structures (e.g., the optic chiasm), cause a considerable increase in ICP, and possibly cause significant endocrine disturbance. Even after aggressive removal of the tumor under microscopic guidance, recurrence is common (10 %).

Special Problems

1. Children with this disease often have growth retardation because of growth hormone deficiency, and they tend to be obese. They may also have behavior disturbances.
2. Adrenal insufficiency must be anticipated postoperatively; corticosteroid replacement therapy should be commenced before operation.
3. Diabetes insipidus may be present preoperatively and almost certainly will appear intraoperatively or postoperatively. Monitor CVP, urine output, and

serum electrolytes during the operation. Be prepared to replace excessive urine losses and to administer DDAVP if necessary.

4. The surgical approach via a frontal craniotomy requires optimal reduction in brain mass (i.e., perfect neurosurgical anesthesia with the liberal use of mannitol, furosemide, and mild hypocarbia) if good access to the tumor is to be obtained.
5. The procedure may be prolonged; the child must be very carefully positioned, padded and a forced air warmer in place.
6. After excision of a craniopharyngioma, children require multiple endocrine replacement therapy.

ANEURYSM OF THE VEIN OF GALEN

This uncommon disease, an AVM involving the great cerebral vein of Galen, is a considerable challenge to the pediatric neuroanesthesiologist. AVM may manifest in the neonate as evidenced by severe congestive cardiac failure and a cerebral bruit; the operative risk is greatest in this age group. Current management usually is by initial neurointerventional transcatheter coil occlusion of the feeding vessels (see below) possibly followed by surgical excision. When lesions of the vein of Galen manifest later in life, the course is more benign, and the lesion may be managed like other AVMs.

Special Anesthesia Problems

1. Neonates and very young infants are likely to have severe congestive cardiac failure preoperatively. This condition can sometimes be improved by embolization of some of the aberrant vessels. Staged closed embolization may represent the safest therapeutic approach to this disease.
2. The surgical mortality rate is high, usually due to uncontrollable bleeding or intraoperative cardiac arrest. Very close monitoring of the bleeding, adequate vascular access and the need to have blood products immediately available will help to manage massive bleeding.
3. Do not allow these patients to become hypotensive or cardiac arrest may rapidly occur (see below).

Anesthetic Management

1. It is important that hypovolemia or hypotension be avoided before the vessels are clipped. The AV shunt through the lesion places a great stress on the heart; failure is common, and myocardial perfusion is threatened by the low diastolic pressure. If the diastolic pressure further decreases, myocardial

perfusion will be inadequate, and cardiac arrest will occur. Hypotensive techniques are contraindicated; maintain BP until the aneurysm can be clipped. Aggressive cardiovascular monitoring is recommended for accurate monitoring of the intraoperative status and replacement of blood as needed.

2. When the aneurysm is clipped, the ventricular afterload suddenly increases and decompensation may occur. Administer vasodilators and inotropic agents if necessary.
3. A “cardiac anesthetic” (i.e., high-dose fentanyl analgesia or a remifentanil infusion) may be indicated for those in CHF.

ELECTROCORTICOGRAPHY AND OPERATIONS FOR EPILEPSY

Many older children (beyond 8 years of age) cooperate adequately during awake/sedated craniotomy for electrocorticography and operations for epilepsy (i.e., temporal lobectomy).

Special Anesthesia Problems

1. Drugs that modify the EEG significantly (e.g., barbiturates) must not be given. A propofol or dexmedetomidine infusion, supplemented with a short-acting opioid is preferred.
2. The child must be awake and cooperative (including being able to speak).
3. Anesthetic techniques must be designed to provide optimal intracranial conditions for surgery.
4. Blood losses are difficult to measure and may be considerable.
5. Postoperatively, the child should be awake to permit neurologic assessment and monitoring.

Anesthesia Management

Preoperative

1. At the preoperative visit, assess whether the child will be able to cooperate during surgery.
2. Explain to the child what will occur during the anesthetic and the reasons for it. Encourage enthusiastic cooperation in the procedure (i.e., explain that in this procedure children experience a dreamy state and feel no pain, but they themselves must help make the operation a success).

3. Do not premedicate.
4. Omit anticonvulsant drugs in the morning of the surgery.
5. Have blood available.

Perioperative

1. Ensure that equipment is at hand for emergency intubation and ventilation. Administer an oxygen facemask with a CO₂ sampling catheter in addition to standard monitors.
2. Insert a large-bore intravenous catheter and arterial line (use local anesthesia as needed).
3. The anesthetic technique must provide an initial period of sedation/anesthesia to prepare the child and perform craniotomy after which the child is allowed to awaken to respond to commands.
4. For the initial period of sedation/anesthesia, dexmedetomidine ± propofol may be used: dexmedetomidine may begin as a slow bolus of 1–2 µg/kg over 10 min followed by or simply begin with an infusion of 0.2–0.7 µg/kg/h. Alternately, a bolus of propofol 1–2 mg/kg IV followed by an infusion of 100–300 µg/kg/min may be used. A combination of propofol and dexmedetomidine has also been used. Titrate these to effect. Fentanyl 1–2 µg/kg IV may be titrated intermittently or a remifentanil infusion commenced until the skull is open. Low concentrations of N₂O may also be delivered via nasal cannulas while continuously monitoring EtCO₂ and SaO₂.
5. With the onset of sedation, surgeons shave the child's head and then insert a urinary catheter (ensure that lidocaine jelly is used).
6. A scalp block is performed on the side of the surgery. *Effective scalp analgesia is the key to success.*
7. Monitor the BP and arterial blood gases.
8. Although the skull is open, give mannitol 1–2 g/kg or furosemide 0.5–1.0 mg/kg and dexamethasone 0.2 mg/kg IV as needed.
9. If excessive respiratory depression occurs, titrate very small incremental doses of IV naloxone (0.05–0.25 µg/kg).
10. After the skull is open, the infusion rates of the sedative drugs should be reduced or discontinued until the child recovers, is able to respond to commands or speak, but remains still. Encourage deep respirations.
11. Management of children for awake craniotomy is not a procedure that one can master by reading a book. It requires considerable attention to detail that can only be gained by observing the procedure being performed.

Postoperative

1. If the child remains excessively drowsy, repeat small doses of naloxone.
2. Order routine post-craniotomy nursing care.

SPINAL CORD TUMORS AND TETHERED CORD

Spinal cord tumors are less common in children than in adults, but they can occur at any site in the spinal cord.

Tethered cord causes bladder and bowel symptoms and weakness of one or both lower limbs. This syndrome is confirmed by MRI and/or computed tomography, which demonstrate a low conus, a thickened filum, and a transverse orientation of nerve roots.

Surgical Procedure

Surgical division of the filum terminale is the treatment.

Special Anesthesia Considerations

1. General endotracheal anesthesia with N₂O, low concentrations of inhalational agent, and small doses of fentanyl with controlled ventilation is preferred. Standard monitoring may be used. Check for proper tracheal tube placement after turning prone.
2. Muscle relaxants should not be used if the surgeon plans to use nerve stimulation intraoperatively. Anorectal manometry or SSEPs from the pudendal nerve may also be used to monitor neurologic function intraoperatively.
3. The child must be carefully positioned on a frame or bolsters to avoid pressure on the abdomen. Such pressure diverts blood from the abdominal veins to the vertebral venous plexus and increases bleeding at the surgical site.

SELECTIVE POSTERIOR RHIZOTOMY FOR SPASTICITY

Some children with spasticity may benefit from rhizotomy of some of the fascicles of the posterior roots of L2 to S1 bilaterally. Intraoperative EMG monitoring is used to determine which fascicles demonstrate a normal response to stimulation (brief local contraction) and which give an abnormal response (a sustained or diffuse contracture). The latter are then divided. Many children benefit significantly, with a generalized reduction of spasticity, improved limb function, and even improved speech function. Sensation is not significantly affected.

Management of anesthesia should be as for tethered cord (see previous discussion) *and with all considerations for the child with CP* (see Chap. 6). Non-depolarizing muscle relaxants should not be administered because they may

compromise interpretation of the EMG findings. Succinylcholine may be given for intubation if required; it is safe to use in the child with CP. Alternatively, low doses of non-depolarizing muscle relaxants will wear off before the need to interpret the EMG.

Intrathecal morphine (10–30 µg/kg), placed by the surgeon, has been successfully used for postoperative analgesia in these children.

ANESTHESIA FOR PEDIATRIC INVASIVE NEURORADIOLOGY

The development of microcatheters and occlusive materials that can be delivered via microcatheters has altered the management of pediatric neurovascular lesions. Neuroradiologists are currently treating AVMs and vein of Galen and other intracranial aneurysms using endovascular techniques. Therapeutic materials include particulate and non-particulate embolic substances and microcoils.

Although morbidity after the use of these methods may be generally less than after open surgery, providing anesthesia to these children in the radiology suite introduces some problems for the anesthesiologist.

Special Anesthesia Problems

1. The radiology suite may be at a distance from the OR and anesthesia support services; ensure that all equipment and supplies that might be needed are available before commencing anesthesia. Make sure that communications to the OR are readily available so that additional equipment or help can be obtained if needed.
2. General endotracheal anesthesia with neuromuscular block (using a nerve stimulator) is always required for infants and young children; it is crucial that children do not move, especially during injections of embolizing materials. Brief periods of apnea may be requested.
3. Access to the children may be limited by the radiologic equipment; check bilateral ventilation after tracheal intubation and firmly secure the tracheal tube. Ensure that ventilator circuits and monitoring lines can be routed so that they are absolutely secure throughout the procedure.
4. Maintenance of normal body temperature may be difficult when the child is on the X-ray table. Use humidified, warmed gases and forced air warming mattresses. Keep the child covered as much as possible.
5. SSEPs may be monitored to guide the procedure; for this reason, inhalational anesthetics must be limited to small concentrations.

6. After the procedure, the child's neurologic status must be assessed and monitored. Therefore, a technique should be used that permits rapid and complete awakening.
7. Potential complications of the procedure include perforation of a vessel or aneurysm, accidental closure of normal vessels or draining veins (aberrant emboli), and adhesion of catheters. Some cases might need to proceed to craniotomy.

Anesthetic Management

1. No preoperative sedation is routinely administered, but it may be preferred for some children with intracranial aneurysm or AVM.
2. Induction and intubation should be planned as for craniotomy for AVM to minimize changes in ICP and adverse hemodynamic responses.
3. N₂O with sevoflurane (up to 1 %) may be used for maintenance and does not significantly affect SSEP monitoring. Opioids (e.g., fentanyl 1–2 µg/kg or remifentanil infusion IV) may minimize postoperative headache and pain. Midazolam (0.1 mg/kg IV) at induction of anesthesia may confer amnesia.
4. A non-depolarizing muscle relaxant should be used (rocuronium or vecuronium) and the degree of neuromuscular block monitored.
5. Special attention is required for positioning and padding to avoid pressure injuries. Means to maintain body temperature should be provided (see previous discussion).

Postoperative Care

1. It is desirable for the child to rest quietly but to not be obtunded, so that an accurate early postoperative neurologic assessment can be made.
2. The catheterization sites and the distal circulation should be regularly checked.
3. Suitable sedation and gentle restraint may be necessary to prevent movement that might result in bleeding or hematoma at the catheterization sites.

Suggested Reading

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Ophthalmology

GENERAL PRINCIPLES

1. General anesthesia is almost always required because children do not tolerate sedation and local analgesia for eye surgery.
2. Intraocular surgery and surgery of the nasolacrimal duct and eyelids require a bloodless field. Although induced hypotension is seldom indicated for these operations, all measures should be taken to ensure that the anesthetic does not increase bleeding. General anesthesia with a secure airway, proper positioning, and a quiet emergence without coughing or straining are important.
3. The oculocardiac reflex (OCR) is powerful in children but is effectively blocked by intravenous atropine (0.01–0.02 mg/kg) at induction of anesthesia. Do not rely on atropine given intramuscularly or on local anesthetic (retrobulbar) blocks to prevent this reflex. *Monitor the heart rate carefully during manipulation of the eyes and extraocular muscles.* In the rare event that atropine is contraindicated, remember that the OCR is more likely to be triggered by sudden traction than by gradually applied progressive traction on the extraocular muscles. The reflex usually fatigues rapidly, that is, a second pull does not elicit as powerful an effect as the first.
4. Some children may be taking medications that have significant side effects (Table 9.1).
5. Medications that are applied to the conjunctivas or injected into the eye during surgery may have important systemic effects or significant implications (Table 9.2).
6. The effects of anesthetic drugs and techniques on intraocular pressure (IOP) must be remembered:
 - (a) Atropine (by any route) causes only a very slight increase in IOP. Its use as a premedicant is not contraindicated in children with open angle glaucoma but is with narrow angle glaucoma.
 - (b) All potent inhalation anesthetic agents, intravenous agents (i.e., propofol, thiopental), and non-depolarizing relaxants decrease IOP; the effect may be dose related.

Table 9.1 Ophthalmic eye drops that may cause systemic side effects

Ophthalmic medication	Indication	Side effects
Echothiopate iodide (phospholine iodide), (long-acting plasma cholinesterase inhibitor)	Glaucoma, esotropia	Nausea, vomiting, abdominal pain, prolonged apnea after administration of succinylcholine
Timolol maleate topical (β -blocking agent)	Glaucoma	Bradycardia refractory to atropine, bronchospasm, exacerbation of the disease in asthmatics
Acetazolamide (Diamox) (carbonic anhydrase inhibitor)	Glaucoma	Metabolic acidosis, and depletion of sodium, potassium, and water. It may also rarely trigger anaphylaxis, Stevens-Johnson syndrome, and bone marrow depression
Dorzolamide (carbonic anhydrase inhibitor)	Glaucoma	Bradycardia

Table 9.2 Ophthalmic medications that may have significant effects during anesthesia

Medication	Potential adverse effects and implications
Epinephrine, phenylephrine	Hypertension and arrhythmias (especially dangerous during halothane anesthesia). Epinephrine eye drops are specifically contraindicated in children with tetralogy of Fallot because they may precipitate a cyanotic “tet” spell. Phenylephrine drops cause fewer problems, especially if the concentration is limited to 2.5%; but if instilled on a hyperemic conjunctiva, they may cause severe hypertension. Monitor the heart rate and blood pressure carefully after drug instillation. Ensure that you know what is being instilled: 10% phenylephrine drops should not be used in children; cardiac arrest may occur
Cyclopentolate (Cyclogyl)	Ataxia, disorientation, psychosis, and convulsions, especially if a 2% solution is used. A 0.5% solution should be used for infants, and a 1% solution for children
Tropicamide (Mydriacyl)	Behavior disturbance, psychotic reactions, and rarely, vasomotor collapse
Scopolamine eye drops	Excitation, disorientation, possible psychosis may be treated with physostigmine 0.01 mg/kg IV
Pilocarpine	Hypertension, tachycardia, bronchospasm, nausea, vomiting, and diarrhea
Intraocular injection of acetylcholine	Increased secretions, salivation, bronchospasm, and bradycardia. Treat with IV atropine (to produce miosis after lens extraction)
Sulfur hexafluoride gas or air injection to globe (to assist in retinal reattachment surgery)	Discontinue N ₂ O 20 min beforehand to prevent an increase in IOP followed by an even more dangerous decrease in IOP as the N ₂ O is withdrawn; this could damage the retinal reattachment

- (c) Intravenous succinylcholine may transiently increase the IOP, and this increase is not reliably prevented by pretreatment with a non-depolarizing relaxant drug. The increase in IOP occurs within 30 s after administration but abates quickly, returning to normal within 6 min. Succinylcholine is usually avoided in children undergoing intraocular surgery. The increase in IOP may be less in those in whom the IOP is already increased (i.e., glaucoma), but it seems prudent to omit succinylcholine in such children, especially if IOP is to be measured. The use of succinylcholine in children with penetrating eye trauma has been controversial. Although originally contraindicated, it has been shown to be safe in at least one very large series. Indeed, when given after thiopental in a rapid sequence induction, succinylcholine does not increase in IOP. However, with the intermediate duration, relatively rapid-acting non-depolarizing relaxants (i.e., rocuronium 1.2 mg/kg), there are alternatives to succinylcholine for RSI in children with penetrating eye injury.
- (d) Ketamine, originally thought to increase IOP, probably has little effect.
- (e) Diuretic drugs decrease IOP, and chronic diuretic therapy may reduce the increase in IOP after succinylcholine administration.
- (f) Pressure on the globe from a facemask increases IOP, avoid compressing the eyes; use a smaller size facemask. Laryngoscopy and tracheal intubation may increase IOP; this effect can be modified by the administration of lidocaine 1.5 mg/kg IV, preferably 3 min before laryngoscopy. The insertion of an LMA increases IOP to a lesser extent than tracheal intubation, and its removal may be associated with less coughing and straining. Therefore, the LMA may be useful for children undergoing elective eye surgery.
- (g) Coughing, bucking, crying, and straining all markedly increase IOP. Smooth extubation with reduced risk of coughing can be achieved by prior administration of lidocaine 1–2 mg IV, a small dose of propofol (1 mg/kg) immediately before removal of the tracheal tube, or by deep extubation (contraindicated in the presence of a full stomach).
- (h) Hypercapnia increases IOP, and hypocapnia decreases it.
7. Succinylcholine causes contracture of the extraocular smooth muscles and interferes with forced duction testing that is performed within 15 min after its administration.
8. The depth of anesthesia for ophthalmic surgery must be sufficient to ensure that the eyes are immobile and fixed centrally; during light anesthesia, the eyes often “roll up” cephalad. Ketamine is generally unsatisfactory for ophthalmic surgery because of the associated nystagmus.
9. Postoperative pain may be troublesome after eye operations, but nonsteroidal anti-inflammatory drugs (NSAIDs) such as preoperative oral or intraoperative intravenous acetaminophen (10–15 mg/kg) may be sufficient

after minor surgery. Immediately after intraocular surgery or trauma repair, a retrobulbar block placed during anesthesia may be very effective. Sub-tenon's block with ropivacaine also reduces pain after strabismus surgery. Topical analgesics (i.e., tetracaine ophthalmic drops) may significantly reduce postoperative discomfort.

10. Postoperative nausea and vomiting are common. It may be reduced by the use of propofol as the primary anesthetic, and it may be further reduced by the intraoperative administration of intravenous ondansetron (0.1 mg/kg), dexamethasone (0.0625–0.15 mg/kg), dimenhydrinate (0.5 mg/kg), or metoclopramide (0.15 mg/kg). Avoid dimenhydrinate, which produces sedation, if adjustable sutures are being used.
11. Be very cautious when using facemask anesthesia for surgery of the eyelids and similar operations (e.g., chalazion excision). Avoid high concentrations of oxygen, which might leak around the mask when the surgeon uses cautery; surgical drapes are flammable and serious facial burns have occurred. An air mixture is preferred; avoid N₂O, which supports combustion.

CORRECTION OF STRABISMUS

Correction of strabismus is the most common eye operation in children.

Associated Conditions

Malignant hyperthermia is very rare, but strabismus may be an associated condition.

Special Anesthesia Problems

1. OCR—Severe bradycardia and even cardiac arrest can occur as a result of traction on the extraocular muscles (see previous discussion).
2. “Oculogastric reflex”—vomiting after eye muscle surgery is very common and should be prevented as outlined previously. Vomiting may also be precipitated by “pushing” oral fluids postoperatively and by early ambulation. We recommend that children only ingest clear fluids when they express a desire to drink.
3. Postoperative pain after strabismus may be considerable in older children, requiring a prescription for an analgesic postoperatively. Consider requesting that the surgeon perform a sub-tenon's block using a recommended dose of ropivacaine.
4. If adjustable sutures are used, the child must be assessable postoperatively; excessive sedation should not be ordered. If a second anesthetic might be

required to adjust the sutures, an intravenous line or a heparin lock should be left in place to facilitate induction of a second anesthetic. Do not use droperidol as an antiemetic in such children because they may be too drowsy to cooperate; the combination of ondansetron and dexamethasone is preferred (see later discussion).

Anesthesia Management

Preoperative

1. Do not give heavy sedation.
 - (a) The surgeon may wish to examine the child immediately before the operation.
 - (b) Oral midazolam (0.25–0.75 mg/kg for children 1–6 years of age) is an effective premedication with rapid onset.
 - (c) Clonidine (4 µg/kg PO) is effective as a premedication although it must be given 60–90 min before surgery. This may cause bradycardia, hypotension, and postoperative sedation but it does provide postoperative analgesia.
2. Give atropine, preferably intravenously, at induction. If not administered, it must be readily available in a syringe in case the OCR is elicited.

Perioperative

1. Induction is accomplished by inhalation of sevoflurane in nitrous oxide or by intravenous propofol or thiopental followed by a relaxant.
2. Topicalization of the larynx with lidocaine is recommended after a sevoflurane induction and a dose of propofol (up to 3 mg/kg IV) or after an intravenous induction and a muscle relaxant. Maintain anesthesia with an inhaled agent. In both instances, an oral RAE tracheal tube is preferred. Alternatively, in suitable children, use a well-lubricated LMA.
3. Maintain anesthesia with N₂O/O₂/isoflurane or sevoflurane; allow spontaneous ventilation (provided the duration of surgery is not excessive).
4. From the start of surgery, monitor the EKG. If bradycardia occurs, ask the surgeon to discontinue traction on the extraocular muscle, additional intravenous atropine may be given; alternatively repeated gentle traction on the muscle may fatigue the OCR.
5. Give ondansetron (0.1 mg/kg) plus dexamethasone (0.0625–0.15 mg/kg) or dimenhydrinate (0.5 mg/kg) intravenously to reduce postoperative vomiting, or metoclopramide (0.15 mg/kg) immediately after the operation.
6. Provide for postoperative analgesia. Strabismus surgery causes limited pain in young children, manageable with acetaminophen or NSAIDs. In older children (>5 years of age), sub-tenon's block may be utilized. Occasionally, the pain after

this surgery is much greater, warranting the use of intravenous opioids. IV ketorolac decreases postoperative pain and is associated with less PONV than opioids. Dexmedetomidine and ketamine provide effective postoperative analgesia; the former also reduces emergence delirium and postoperative vomiting. Tetracaine eye drops instilled by the surgeon also provide analgesia.

Postoperative

1. To prevent subconjunctival hemorrhage, the trachea must be extubated or the LMA removed without causing coughing and straining by the child. Extubate while the child is still deeply anesthetized, and allow the child to awaken smoothly while supporting the airway and administering oxygen by mask. Intravenous lidocaine (1.5 mg/kg) or low-dose propofol (0.5–1 mg/kg) administered before extubation may reduce coughing during emergence.
2. Provide analgesics for pain as indicated above, avoid opioids where possible.
3. Intravenous rehydration during surgery should obviate the need for early oral ingestion in the post-anesthesia care unit. Delaying ingestion of oral fluids decreases the incidence of post-strabismus vomiting. Do not attempt to mobilize the child rapidly; there is a motion sickness component to PONV after strabismus surgery. Beware that vomiting may occur en route to home in outpatients. These children tend to sleep more than most in part because they have double vision in the immediate postoperative period. It is reasonable to inform parents that their child will be quite sleepy the rest of the day.
4. If nausea and/or vomiting occur, order additional antiemetic therapy and continue with intravenous fluids.

INTRAOOCULAR SURGERY AND EXAMINATION UNDER ANESTHESIA FOR GLAUCOMA OR TUMOR

Children most commonly require general anesthesia for cataract or glaucoma surgery, treatment of detached retina, or examination under anesthesia (EUA) for glaucoma or tumor.

Special Anesthesia Problems

1. The OCR (see previous discussion).
2. IOP—may be affected by anesthesia drugs and techniques (see previous discussion).
3. Coughing and straining—*may increase the IOP*. (Induction of and emergence from anesthesia should be as quiet and smooth as possible.)

Anesthesia Management

Preoperative

1. Give adequate sedation to prevent coughing and straining.
2. It is safe to give atropine to children with congenital open-angle but not narrow angle glaucoma.
3. Explain to older children that their eye will probably be covered with a patch after surgery.

Perioperative

1. Induce anesthesia as smoothly as possible, by inhalation of sevoflurane and N₂O or intravenously with propofol or thiopental.
2. Avoid succinylcholine.
3. For brief EUA procedures, use a facemask, but prevent pressure on the globe as it may increase IOP. Otherwise, either deepen anesthesia using a single dose of propofol (up to 3 mg/kg IV) or spray the larynx with lidocaine before intubating the trachea or inserting a well-lubricated LMA. For prolonged surgeries, a non-depolarizing muscle relaxant may be administered.
4. Maintain anesthesia with N₂O/O₂ and isoflurane, sevoflurane, or desflurane. Allow spontaneous ventilation for brief EUA procedures; otherwise, control ventilation to prevent hypercapnia. Alternatively, use a propofol infusion to maintain anesthesia because it may be advantageous in reducing postoperative vomiting.
5. Discontinue N₂O early if sulfur hexafluoride or air is to be injected into the eye.
6. A retrobulbar block with bupivacaine or ropivacaine may be useful to reduce postoperative pain.
7. At the end of surgery, suction the pharynx carefully and extubate the trachea, or remove the LMA, while the child is still deeply anesthetized. Lidocaine 1.5 mg/kg IV, administered before extubation decreases the risk of coughing or straining during emergence.
8. Reapply the facemask, support the airway, and give oxygen until the child awakens.

Postoperative

1. Order adequate sedation and analgesics.
2. Order an antiemetic as required.

PROBING OF THE NASOLACRIMAL DUCT AND CHALAZION EXCISION

These minor procedures are performed in the ambulatory setting and usually present few special challenges. The airway is usually managed with either a face-mask or LMA; tracheal intubation is rarely required. Nasolacrimal duct surgery is usually performed in infants (less than 12 months of age). Although it is usually brief, insertion of a silicon Crawford tube often requires additional time. Patency of the duct is confirmed by either injecting a dilute fluorescein solution into the ducts and detecting it on a pipe cleaner in the nose or by the touch of metal inserted into the duct to metal in the nose. Either fluorescein or blood from unblocking the duct may drain into the nasopharynx and trigger laryngospasm. To prevent such an occurrence, a roll may be inserted under the shoulders and the infant is positioned in Trendelenburg (head down). In this position, pharyngeal fluids drain away from the glottis. Beware of oxygen leaks under the mask if cautery (for chalazion surgery) is used (see previous discussion). If it appears that the procedure may be more difficult and prolonged, intubate the trachea or insert an LMA.

PENETRATING EYE TRAUMA

Penetrating eye trauma is a relatively common injury in children.

Special Anesthesia Problems

1. Any increase in IOP in the presence of an open eye injury may cause extrusion of anterior chamber structures and/or vitreous humor. Crying, coughing, and straining should be prevented as much as possible. Although one may be tempted to sedate the child preoperatively, in most cases a full stomach is present and sedation is relatively contraindicated. Whenever a foreign body is lodged in the eye, the ophthalmologist prescribes IV antibiotics immediately to prevent endophthalmitis, hence intravenous access will usually be established in the emergency room. If a rapid sequence induction is indicated, an intermediate-acting, non-depolarizing relaxant such as rocuronium is appropriate. Succinylcholine has traditionally been contraindicated in children with open eye injuries particularly with laceration injuries, although it has been used in a large series without serious sequelae (see discussion above). Hence, many would consider it justified if other indications exist for its use. Intravenous propofol or lidocaine minimizes the increase in IOP caused by laryngoscopy.
2. It may be difficult to position a mask if the eye is covered with a dressing.

Anesthesia Management

Preoperative

1. Give light sedation and analgesics as required provided a full stomach is not present; avoid upsetting the child.
2. If indicated, as early as possible before induction, give metoclopramide (Reglan) 0.1 mg/kg IV to expedite gastric emptying.
3. Give intravenous atropine at induction. (N.B. Atropine blocks the effect of metoclopramide and therefore should not be given earlier.)

Perioperative

1. Most children require a rapid sequence induction; give 100 % O₂ by mask if possible before inducing anesthesia.
2. Inject lidocaine 1 mg/kg IV, followed by propofol (or thiopental) and rocuronium (1.2 mg/kg).
3. Have an assistant apply cricoid pressure as needed (see Chap. 4).
4. Intubate the trachea before inflating the lungs. Aspirate the stomach.
5. Control ventilation and maintain anesthesia with N₂O/O₂ and isoflurane, sevoflurane, desflurane, or a propofol infusion.

Postoperative

1. Administer IV lidocaine before extubation to decrease coughing.
2. Extubate the trachea when the child is fully awake and in the lateral position.

THE PRETERM INFANT FOR LASER TREATMENT OF DETACHED RETINA

Improved management of very-low-birth-weight infants has led to an increase in the incidence of retinopathy of prematurity (ROP). The outcome of this disease is improved if laser treatment, possibly combined with intravitreal injection of bevacizumab (NOTE: this is an off-label use), or cryotherapy of the detached retina is performed when the disease is at threshold level. These interventions reduce ROP via reductions in vascular endothelial growth factor and insulin-like growth factor 1. Attempts to manage these patients using local analgesia and sedation are often unsuccessful and are accompanied by major complications. Therefore, general anesthesia is usually administered.

Special Anesthesia Problems

1. The infant is usually still very small and subject to all the problems of the preterm infant in the perioperative period. All precautions for the preterm must be observed (see Chaps. 2 and 4).

2. The infant may lose heat and become hypothermic during transport to the operating area. Preterm infants who are transported in an open infant bed invariably become hypothermic even if well covered. A proper heated transport incubator is required.
3. Access to the infant during treatment is limited. Airway, ventilation, and reliable comprehensive monitoring must be assured before the procedure commences. Thermal environment must be optimized (i.e., heated OR and forced air warmer).
4. Topical anesthetics and oral or rectal acetaminophen reduce pain. Postoperatively the infant's trachea should remain intubated and the lungs ventilated until judged stable by the neonatal team. A period of ventilation for 24–36 h is commonly required even if the infant had been weaned before the eye treatment. A period of ventilatory instability is common postoperatively.

ANESTHESIA FOR RETINOBLASTOMA

This is the most common ocular tumor in children, comprising up to 4% of tumors in childhood. Two forms of this tumor exist: unilateral comprising 75% of cases (with 90% nonhereditary) and bilateral comprising 25% (inherited as the RB1 gene on 13q14). Ten to 15% of children with retinoblastoma have metastatic disease. The objective of treatment is to preserve vision and remove the tumor. Treatment depends on the extent of the disease. Enucleation is reserved for massive tumors for which conservative treatment cannot preserve vision. Anesthesia for enucleation of an eye requires a completely still infant. General anesthesia with tracheal intubation is most appropriate. The anesthetic prescription should follow our standard management of any infant; avoid long-acting agents. The most serious, potentially fatal, complication is recession of the ophthalmic artery during dissection from the back of the orbit, resulting in massive blood loss until it is recovered.

Focal therapies (for tumors <6 mm in diameter) include argon laser therapy, cryotherapy, and thermotherapy; the therapy depends on the location of the tumor within the eye. Chemotherapy is reserved for metastatic disease. Radiotherapy and brachytherapy (see below) are also effective as retinoblastoma is radiosensitive, but may produce unrelated malignancies. Serial proton beam treatments may result in less potential for later malignancy.

Anesthesia for cryotherapy and laser therapy may be performed with an LMA or tracheal intubation. The goal of anesthesia is to prevent movement of the head while treatment is administered. There is no need for analgesics or antiemetics after these procedures in young infants.

ANESTHESIA FOR RADIOTHERAPY

Infants and children with retinoblastoma may require daily repeated radiotherapy for which they must remain absolutely still; hence, general anesthesia is required. This is usually a salvage procedure when all other treatments have failed; the risk with this approach is developing second (unrelated) malignancies. Children who will have serial radiation therapy or proton beam treatments generally undergo a series of procedures starting with insertion of a central access device, followed by MRI assessment, and then a CT planning session whereby a restraining facemask is constructed. During this series, it is preferable to administer anesthesia with propofol and spontaneous respirations with face-mask oxygen and to assure proper positioning that maintains a clear airway. All subsequent treatments will be undertaken in a similar manner with a natural airway with supplemental oxygen. Thus, it is essential to use a proper fitting mask created during the CT planning session. Generally such treatments will occur 5 days per week for 20–30 treatments depending on the severity of the tumor, and if it is bilateral.

The challenge is to administer short-acting anesthetics and return the child to normal activity and feeding as soon as possible. Nasal or facemask oxygen applied to the preformed mask and capnometry combined with pulse oximetry help to assure adequate ventilation and oxygenation during the procedures.

Some institutions use of the HeadFIX® Immobilization Device,¹ which applies suction to a mold of the palate, firmly supports the head and also generally maintains a very good airway without further instrumentation. Our experience is that a propofol infusion is safe and effective in these children.

ANESTHESIA FOR VISUAL EVOKED POTENTIALS OR ELECTRORETINOGRAPHY

Visual evoked potentials (VEPs) are exquisitely sensitive to the effects of sedative and anesthetic agents because of the complex cortical pathways involved. Hence, the anesthesia technique must be carefully planned if meaningful results are to be obtained.

1. Nitrous oxide significantly depresses the amplitude of VEPs.
2. Potent inhaled agents decrease amplitude and increase the latency of VEPs.
At 1.5 MAC, VEPs are unrecordable.

¹Medical Intelligence, Schwabmünchen, Germany.

3. Thiopental decreases the amplitude and increases the latency of VEPs. An induction dose of etomidate has less effect, only a slight increase in latency and no change in amplitude.
4. Propofol has a dose-related affect on amplitude and latency—hence, low doses may be acceptable.
5. Ketamine has a negligible effect on latency and a dose-related effect on amplitude of VEPs.
6. Fentanyl significantly decreases the amplitude.

In summary, it is difficult to provide anesthesia care for infants and small children for VEPs. A technique based on low-dose propofol or ketamine is recommended.

The results of electroretinography (ERG) are less affected by anesthesia and sedative agents. These are often performed in a completely dark room in the ophthalmology department, remote from the O.R. Special screens block the light from the anesthesia monitors, necessitating close observation of the monitors. We commonly use a propofol infusion, deliver oxygen via nasal prongs, and sample end-expired gases for CO₂ and spontaneous respiration.

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Otorhinolaryngology

GENERAL PRINCIPLES

Although much of it is simple and commonplace, ear, nose, and throat surgery has a disproportionately large potential for anesthetic and surgical complications. It demands meticulous attention to all aspects of the child's perioperative care:

1. Because many of these operations involve the airway, the anesthesiologist must be prepared to provide good surgical access while ventilating and oxygenating the child.
2. The advent of the surgical microscope has permitted development of delicate and precise surgery for the middle ear. Anesthesia for such procedures must provide quiet operating conditions with minimal bleeding, smooth emergence from anesthesia, and minimal disturbance postoperatively.
3. After surgery involving the airway, skilled nursing care in the postanesthesia care unit (PACU) is essential, so that signs of impending complications can be detected early and appropriate therapy instituted.
4. The use of the laser to treat lesions of the larynx creates additional potential problems for anesthesia management.
5. When topical vasoconstrictors are used, the anesthesiologist must be aware of the drugs and doses that will be used because significant systemic absorption may cause hypertension and bradycardia. A maximum initial dose of 20 µg/kg of phenylephrine has been recommended for children, but this is considerably less than has been commonly used. When topical vasoconstrictors are applied, the child should be monitored carefully; transient mild hypertension requires no therapy. Occasionally, severe hypertension may occur which may cause pulmonary edema; this should be treated with direct vasodilators (e.g., sodium nitroprusside) or α-adrenergic receptor antagonists (e.g., phentolamine). Do not use β-blockers or calcium channel blockers in the presence of hypertension; these may cause a disastrous decrease in cardiac output and cardiac arrest.

CHOANAL ATRESIA

Complete obstruction (90 % of cases) due to choanal atresia (membranous or bony occlusion of the posterior nares) causes respiratory distress immediately after birth. However, the distress is intermittent; infants are primarily nose breathers, but the obstruction is relieved whenever the infant opens the mouth and cries. The diagnosis is easily confirmed with a stethoscope by listening for air exchange over each nostril; the obstruction can generally be relieved with an oropharyngeal airway. The passage of an orogastric tube may open the oral airway in some infants and also facilitates feeding. Early endonasal puncture and stenting may be performed in the neonate, and KTP laser in the preterm infant. Incomplete choanal atresia may lead to chronic nasal problems; therefore, early repair is recommended.

Associated Conditions

The “CHARGE” association consists of Coloboma, congenital Heart disease, choanal Atresia, growth and growth Retardation, Genitourinary abnormalities with genital hypoplasia, and Ear anomalies.

Surgical Procedures

1. Endoscopic transnasal puncture may be preferred, especially in those with associated significant disease (e.g., the CHARGE association). In preterm infants, KTP laser is preferred.
2. Transpalatal repair if indicated is usually performed at age 1–2 days in the healthy, full-term infant. Stents are left postoperatively for varying periods.

Special Anesthesia Problems

The primary problem is maintenance of the airway until completion of surgery.

Anesthesia Management

Preoperative

1. Adequate ventilation requires continued use of an oropharyngeal airway.

Perioperative

1. Observe all special precautions for neonates (see Chap. 4).
2. Leave the oropharyngeal airway in place: give 100 % O₂ by mask.
3. Induce anesthesia by inhalation of sevoflurane, establish IV access, and intubate the trachea using a preformed oral RAE tube.

4. Maintain anesthesia with N₂O and sevoflurane or isoflurane with controlled ventilation; a remifentanil infusion may reduce the need for inhalation agent while providing excellent depth of anesthesia allowing the surgeon to place the necessary airway stents.
5. Suction the pharynx very carefully at the end of the operation, and ensure that the stents are clean and patent.
6. Extubate the trachea when the infant is fully awake.

Postoperative

1. Order humidified oxygen/air. The stents must be regularly suctioned with a fine catheter to keep them clear.
2. Constant observation is essential because aspiration during feeding commonly occurs after repair of choanal atresia.
3. Subsequent repairs may be necessary later in childhood for restenosis, but these operations present no other special anesthetic problems.
4. Order appropriate analgesia.

NASOPHARYNGEAL TUMORS

Teratomas, dermoid cysts, nasal encephaloceles, and other tumors require surgical excision. Juvenile nasal angiofibroma is a rare benign but very vascular tumor that may involve the nose. Biopsy of these tumors may result in extensive bleeding that is very difficult to control; therefore, diagnosis is usually made on the basis of imaging studies. An operation to remove the tumor may result in massive blood loss, and this problem should be anticipated. Postoperatively, nasal obstruction and bleeding may persist; the tracheal tube should be left in place until the child is fully awake.

SURGERY OF THE NOSE

The most common nasal surgical procedures are reduction of fractures, septoplasty, rhinoplasty, and excision of nasal polyps.

Special Anesthesia Problems

1. The nasal airway may be blocked. The surgeon may wish to pack the nose with gauze and a vasoconstrictor (e.g., 0.05% oxymetazoline or 1/2000 epinephrine) preoperatively.
2. Children with nasal polyps usually have cystic fibrosis.
3. Functional endoscopic sinus surgery (FESS) may precipitate special problems (see later discussion).

Anesthesia Management**Preoperative**

1. Assess the nasal airway.
2. If the child has cystic fibrosis, order appropriate preoperative care (see Chap. 6).

Perioperative

1. Induce anesthesia by inhalation or intravenously, followed by a muscle relaxant.
2. If the nose is blocked, insert an oropharyngeal airway before attempting mask ventilation.
3. Perform orotracheal intubation, with a cuffed RAE tube.
4. Insert a throat pack to prevent blood pooling in the pharynx and esophagus.
5. Position the child with a slight head-up tilt.
6. Consider the use of bilateral infraorbital and nasociliary nerve blocks for postoperative analgesia.
7. Extubate the trachea when the child is fully awake; premature extubation may lead to laryngospasm or airway obstruction.

Postoperative

1. Order analgesics as required.
2. Administer humidified oxygen/air by mask.
3. Postoperative airway obstruction may occur, resulting in postobstructive pulmonary edema requiring reintubation. Should this occur, apply standard therapy (IPPV with oxygen and a diuretic).

FUNCTIONAL ENDOSCOPIC SINUS SURGERY

FESS has become a standard surgical treatment for chronic sinus disease. Precise endoscopic resection of diseased tissue and relief of obstruction while preserving normal mucosa is the objective to restore normal sinus function.

Special Anesthesia Problems

1. Many of these children have chronic diseases (e.g., cystic fibrosis).
2. Successful endoscopic surgery requires extensive use of vasoconstrictors. Ensure that maximal permissible doses are not exceeded so as to prevent severe hypertension: 2–3-mg/kg cocaine or 10- μ g/kg epinephrine. If hypertension ensues, treat it by deepening anesthesia or using vasodilators; do not use β -blockers or calcium channel-blocking agents.

3. Bleeding may be considerable and may require that packing remain in place postoperatively. Because this is likely to cause complete nasal obstruction, have the child fully awake before extubation.
4. Consider the use of bilateral infraorbital and nasociliary nerve blocks for postoperative analgesia.
5. Rarely, the surgery may encroach on the orbit or intracranial space. In the latter case, intracranial bleeding may occur. There is also a danger of pneumocephalus if positive pressure is applied via a face mask.

TONSILLECTOMY AND ADENOIDECTOMY

Chronic inflammation and hypertrophy of lymphoid tissues in the pharynx may necessitate surgery to relieve obstruction or to remove the focus of infection. Repeated middle ear infections may be improved by adenoidectomy. Obstructive sleep apnea (OSA) is now the commonest indication for tonsillectomy and adenoidectomy (T&A) in North America. Rarely, acute tonsillitis leads to a peritonsillar abscess (quinsy tonsil).

T&A surgery is often performed in the ambulatory unit; this demands special considerations in the selection of suitable children and in their postoperative evaluation before discharge home. An efficient follow-up service must be provided to deal with unexpected complications. Some children may not be suitable for outpatient T&A.

The following are indications for admission after T&A:

1. Age <3 years
2. Those with abnormal coagulation studies or a history of increased bleeding tendency
3. Those with evidence of significant OSA (see later discussion)
4. Those with other systemic diseases that place them at additional perioperative risk (e.g., congenital heart disease, endocrine or neuromuscular disease, chromosomal abnormalities, obesity)
5. Those with craniofacial or airway abnormalities, including Down syndrome
6. Those with a history of peritonsillar abscess
7. Those who live at an excessive distance from the medical facility or whose home, social, or parental situation might preclude safe postoperative care.

T&A is still one of the most common procedures in children and should be very safe. However, T&A-related deaths still occur: the usual cause is excessive sedation of children with airway compromise, OSA, or postoperative bleeding. Postoperative nausea and vomiting are common after T&A but may be significantly reduced by withholding postoperative oral fluids until the child requests them, rehydration during anesthesia (20–25-mL/kg lactated Ringer's solution or

equivalent), and the administration of dexamethasone (0.0625–0.15 mg/kg up to 8 mg) and a serotonin-receptor antagonist (e.g., ondansetron 0.05–0.1 mg/kg up to 4 mg) IV intraoperatively.

Obstructive Sleep Apnea

Chronic obstruction due to lymphoid hyperplasia may result in OSA. Affected children may be obese (or asthenic), snore loudly, and show difficulty arousing in the morning or daytime somnolence, new onset of nocturnal enuresis and behavior problems (attention deficit disorder and limited attention span), cognitive impairment, night terrors, and nocturnal apnea and/or gasps.

If such a history is obtained preoperatively, ideally a sleep study (polysomnogram) is performed; if the results are significantly abnormal, admission after tonsillectomy is advised. Polysomnographic criteria for admission after tonsillectomy for the T&A with a history of OSA include:

1. A baseline value for partial pressure of carbon dioxide (PaCO_2) $\geq 50 \text{ mmHg}$
2. A baseline awake oxygen saturation value $\leq 92\%$
3. Episodes of oxygen desaturation $\leq 80\%$
4. Apnea/hypopnea index > 10

If polysomnography is unavailable, overnight oxygen saturation monitoring has been proposed as a valuable screening tool; resting saturations $< 90\%$ and episodes $< 80\%$ are strongly predictive of severe OSA. Although the positive predictive value of overnight SpO_2 monitoring is very high, a negative test is less helpful and should be interpreted cautiously. A promising, noninvasive test that offers to diagnose OSA in young children is the presence of ≥ 2 or 3 urinary proteins (increases in oromucoid-1, urocortin-3, and uromodulin and/or decreases in kallikrein) with 95–100% sensitivity and specificity.

Children with OSA have reduced responses to rebreathing CO_2 and may be exceedingly sensitive to opioids, requiring dramatically reduced doses of opioids. The latter is prominent in children whose nocturnal desaturation is $< 85\%$ (see below).

Children with mild OSA are reported to have few complications after T&A. Those with moderate/severe OSA may require more intensive monitoring and postoperative care.

The child with OSA should be closely monitored before and after surgery; supervised mild sedation (oral midazolam) is safe preoperatively but is often omitted. Children with OSA may be at risk for desaturation even after oral midazolam and should be observed closely. In all children with diagnosed OSA, spontaneous respirations are maintained after tracheal intubation to test their respiratory responses to small incremental doses of opioid (i.e., IV fentanyl 0.5 $\mu\text{g}/\text{kg}$ or morphine 0.025 mg/kg). Children with upregulated opioid

sensitivity (nocturnal desaturation <85%) may develop apnea after a single dose of opioid despite surgical stimulation. In such cases, no further opioids should be administered, and postoperative opioid doses should be markedly reduced or omitted. Either preoperative oral acetaminophen (10–15 mg/kg) or intraoperative IV acetaminophen (15 mg/kg) may also be effective. Post-tonsillectomy ketorolac has been criticized because of the risk of bleeding and reoperation, although those data are controversial. There may be roles for diclofenac, tramadol, dexmedetomidine, and celecoxib for post-tonsillectomy analgesia, although experience with these analgesics in children with OSA is limited. Ketamine 0.5-mg/kg IV during surgery reduces the need for other analgesics and may be particularly useful in children with OSA. Most importantly, codeine should not be prescribed for postoperative analgesia as deaths associated with children who have OSA (increased opioid sensitivity) and duplicated 2D6 isoforms (greater conversion of the prodrug codeine to morphine) have occurred, resulting in a black box warning from the FDA (see Chap. 3). It should be noted that other opioids (e.g., tramadol and oxycodone) may have altered pharmacokinetics related to duplicated cytochromes resulting in accumulation of tramadol and greater conversion of oxycodone to hydromorphone (see Chap. 3). Alternative analgesics as described above or alternating q6h ibuprofen and acetaminophen are recommended.

Postoperative sleep studies show that 90% or more of those with mild/moderate OSA improve by 6 months, but only 30% improve if obesity or severe OSA is present. Those who do not improve may benefit from intranasal steroids and nasal nocturnal oxygen therapy or should be investigated for residual soft tissue obstruction and evaluated for uvulopalatopharyngoplasty. In general, children do not tolerate CPAP or BiPAP devices.

Cardiorespiratory Syndrome

In very rare instances, severe chronic airway obstruction by adenoidal tissue may lead to pulmonary hypertension and right-sided heart failure (cardiorespiratory syndrome). This condition usually occurs in boys and is more common in African-American children. There is usually a history of symptoms lasting 1 year or longer. The child is usually febrile (due to associated adenoid or pulmonary infection) with tachycardia and tachypnea. Chest radiography may reveal cardiomegaly, and the electrocardiogram indicates right ventricular hypertrophy. Children with cardiorespiratory syndrome may be critically ill and require emergency tracheal intubation to relieve the obstruction. Heart failure is treated with digitalis and diuretics, and then T&A should be performed. They often need to remain intubated postoperatively, and all should be admitted to the ICU for further observation.

Special Anesthesia Problems for T&A Surgery

1. Sharing the airway with the surgeon.
2. Danger of postoperative bleeding (increased in obese children).
3. A history of bleeding tendency or recent salicylate therapy. If such a history is obtained, a bleeding time should be performed, and if prolonged (more than 10 min), the operation is deferred.
4. A history suggestive of OSA. Such children are at risk for perioperative apnea and demonstrated increased sensitivity to opioids (see previous discussion).

Anesthesia Management

1. For children with a history of chronic infectious tonsillitis, anesthesia may be induced intravenously or by inhalation and tracheal intubation.
2. Perform tracheal intubation with an oral RAE tube that is positioned under a slotted tongue blade of the mouth gag. Check airway patency after the gag is positioned. Check for bilateral ventilation because the tip of the RAE tube may become endobronchial after insertion of the mouth gag or if the neck is flexed. Some advocate a laryngeal mask airway (LMA) in place of a tracheal tube; however, in our opinion, the very low-associated morbidity and additional security of a tracheal tube argue against substituting an LMA.
3. Administer standard antiemetic therapy to all children: ondansetron (0.05–0.1 mg/kg up to 4 mg) and dexamethasone 0.0625–0.15-mg/kg IV (maximum 8 mg) and lactated Ringer's solution (20–25 mL/kg) to reduce nausea and vomiting and improve postoperative comfort.
4. When the indication for surgery is chronic infection, maintain anesthesia with N₂O and sevoflurane or isoflurane, assist ventilation (with or without a relaxant), and supplement with opioids. Plan for postoperative analgesia with morphine 0.05–0.1 mg/kg IV. When the indication is for OSA, titrate opioid dosing or use alternative analgesics as per the anesthetic technique described above.
5. Measure and chart blood losses carefully.
6. Carefully suction the pharynx; the presence of small amounts of blood in the pharynx may lead to laryngospasm. Extubate the trachea when the child is fully awake and airway reflexes are fully restored (especially with OSA). Do not pass suction catheters through the nose because doing so may make the adenoidal area bleed. Alternatively, the trachea may be extubated "deep" in older children. Transfer the child in the recovery position (lateral decubitus) to PACU.

Postoperative

1. Order morphine (0.05–0.1 mg/kg) IV in the PACU q5–10 min until comfortable in children without OSA. Children with OSA should be carefully monitored (pulse oximetry) in PACU; morphine doses based on the intraoperative responses (ranging from 0 to 50% of the usual dose for those who developed apnea after small doses to 100% of the usual dose of opioid for those whose breathing was

unaffected by the intraoperative morphine) should be titrated to ventilations. Caution: pulse oximetry is a poor measure of adequacy of ventilation if supplemental oxygen is being administered; use capnography in such cases.

2. An intravenous infusion should be maintained until the child is ready for discharge. It is particularly important to ensure that children undergoing T&A on an outpatient basis are well hydrated before discharge.
3. Do not push oral fluids; the child should have received the 20–25 mL/kg balanced salt solution infusion to restore euvoolemia. Order fluids by mouth as requested by the child (i.e., cola beverages, Popsicles) when the child is awake. If PONV occurs and further antiemetic therapy is required (e.g., ondansetron), be aware that this might prevent the child who is continuing to bleed from vomiting and thus conceal the hemorrhage.
4. Closely monitor children with OSA; airway obstruction may actually worsen in the first 24-h postsurgery. Such children may become apneic with opioids or sedatives, require constant nursing attention, and often require extended monitoring, e.g., ICU. Some children (especially the obese) may benefit from nasal CPAP therapy or nasal oxygen.
5. Be cautious when ordering opioids for the restless child, especially if there is any evidence of airway compromise. Restlessness may be a symptom of hypoxia secondary to obstruction, and opioids may produce apnea. Do not remove monitors once the child is settled.
6. The outpatient should be evaluated directly by the surgeon for bleeding and the anesthesiologist for adequacy of ventilation before discharge. In general, we recommend observing children for at least 4 h before discharge.
7. A written analgesic regimen should be provided to the parents and discussed with them. The use of acetaminophen in combination with ibuprofen has been found to be a satisfactory alternative to codeine-based regimes.
8. A telephone consultation service for follow-up of outpatients on the evening after surgery may improve patient/parent satisfaction.
9. Complaints of abdominal pain after T&A suggest the presence of swallowed blood from ongoing bleeding, especially after potent antiemetic therapy. Suspicion of tonsil/adenoidal bleeding should be raised.

Reoperation for Bleeding After Tonsillectomy

Special Anesthesia Problems

1. The stomach contains blood that may be regurgitated during induction.
2. Hypovolemia may be easily underestimated. There may be little visible bleeding, but much may have already been swallowed.
3. The child may have a bleeding disorder that has been not been identified; this should be evaluated.

Anesthesia Management

Preoperative

1. Ensure that sufficient fluids have been administered to restore euvoolemia, and circulatory homeostasis. Bleeding is rarely so brisk that complete restoration of blood volume cannot be achieved before operation.
2. Check that blood is available if needed. Transfuse to a minimum Hb of 7 or 8 Gm%.
3. Check coagulation indices.
4. In some instances, gentle restraint permits examination, insertion of packing, cautery, or ligation of bleeding vessels without the need for general anesthesia.
5. Rapid bleeding may prevent laryngoscopy; *be prepared for a surgical airway.*

Perioperative

1. Prepare all equipment for a rapid sequence induction (two suctions, two functioning laryngoscope blades, and two functioning handles).
2. Check again that the child has been adequately fluid resuscitated.
3. Give 100% O₂ by mask.
4. Rapidly inject propofol (or ketamine or etomidate if hypovolemia is a concern) with atropine added, followed immediately by succinylcholine (2 mg/kg). If there is a contraindication to succinylcholine, then give high-dose rocuronium (1.2 mg/kg), but anticipate a prolonged recovery from neuromuscular blockade after control of bleeding (~75 min). Do not be surprised if the tracheal intubation is difficult: edema, bleeding, and surgical tissue trauma may create a challenging view of the larynx.
5. Have an assistant immediately apply cricoid pressure if appropriate (see page 99).
6. Intubate the trachea as rapidly as possible with a styletted cuffed tracheal tube. Consider left lateral decubitus and Trendelenburg positions if bleeding obscures the larynx.
7. Maintain anesthesia as for T&A (see above), although surgery for tonsil rebleeding is much less painful than the original T&A, so a minimal opioid dose is usually needed, especially in children with OSA.
8. Extubate the trachea with the child in a lateral position when any residual neuromuscular blockade has been adequately antagonized and the child is fully awake.

Postoperative

1. Check the hemoglobin level to confirm adequacy of blood replacement.
2. Be alert to the possibility of further bleeding (increased in obese children).
3. Monitor oxygen saturation and vital signs closely.

4. Order suitable doses of analgesic as needed (not acetylsalicylic acid).
 - (a) Oversedation could result in complete obstruction of the airway.
 - (b) Restlessness may indicate hypoxia rather than a need for sedation.
 - (c) Do not remove monitors until the child is fully awake.

Peritonsillar Abscess (Quinsy)

Special Considerations

1. Trismus and swollen tissues in the pharynx may make tracheal intubation difficult.
2. There is danger that the abscess may burst and flood the pharynx with pus that leads to pulmonary aspiration.

Anesthesia Management

Preoperative

1. Children with tonsillar abscess should be closely observed for impending airway obstruction. Check the extent to which the mouth can be opened; significant trismus may be present.
2. Avoid premedications, particularly in children with evidence of airway obstruction.

Perioperative

1. Ensure that strong suction is available. Give atropine 0.02 mg/kg IV.
2. Induce anesthesia by inhalation of N₂O with sevoflurane. Maintain spontaneous ventilation. Position the head slightly down and turned to the affected side.
3. Do not give muscle relaxants (airway obstruction may occur).
4. When the child is *deeply* anesthetized, discontinue N₂O and continue with sevoflurane in 100% O₂; give 1–2 mg/kg lidocaine IV to reduce the risk of coughing or breath holding during laryngoscopy and tracheal intubation. Be careful not to rupture the abscess during airway instrumentation.
5. Maintenance is the same as for T&A for chronic infection (see previous discussion).
6. Suction carefully and extubate the fully awake child in a lateral position.

N.B. Sometimes the inflammatory swelling involves the supraglottic structures, and postextubation obstruction may occur. Dexamethasone (0.1 mg/kg up to 8 mg) IV should be considered. Close observation is essential.

OTOLOGIC CONDITIONS

Surgery for ear conditions ranges from (minor surgery) simple myringotomy and tubes (M & T) to (major) prolonged surgery for tympanomastoidectomy and cochlear implants. M & T surgery is brief (as brief as 5 min) but is not without risk because these infants and children often have or had recent URTIs. In contrast, tympanomastoidectomy and cochlear implantation require considerations for prolonged surgery, require tracheal intubation, and are associated with PONV.

Special Anesthesia Problems

1. The child may have had repeated procedures and may be very apprehensive.
2. The child's hearing may be impaired, making communication difficult.
3. During middle ear procedures, even a small amount of bleeding may interfere with surgery. Position the child carefully and avoid anesthetic causes of bleeding (e.g., hypoventilation, coughing, NSAIDs); induced hypotension is not usually required.
4. The surgeon may wish to use vasoconstrictor drugs (e.g., epinephrine). In such cases, the dose should not exceed the maximum (see previous discussion).
5. Otologic procedures can be lengthy; if this is the case, ventilation should be controlled, and careful attention should be paid to positioning, padding, and maintenance of body temperature.
6. In rare cases, the child's cooperation is required during surgery (see Chap. 8 Awake Craniotomy).
7. If the surgeon uses a nerve stimulator, relaxants are limited to induction of anesthesia.
8. Postoperative PONV secondary to labyrinthine disturbance is common. Prior therapy with antiemetics (e.g., ondansetron) may be useful.
9. The use of a great auricular nerve block before surgical incision and repeated at the end of surgery reduces vomiting compared with opioids.

Minor Otologic Procedures: Myringotomy and Tube (M & T) Placement

Minor otologic procedures are usually performed in the outpatient department. N₂O has been shown to pass into the middle ear cavity if air is present and may modify findings at operation, but in general, its use is not contraindicated. N₂O does not increase the incidence of postoperative vomiting.

Special Anesthesia Problems

1. Some children who require repeated minor otologic procedures have associated congenital deformities of the upper airway that predispose to their ear disease (e.g., cleft palate, Treacher Collins). Check previous anesthetic records carefully for potential airway problems.
2. Many of these children present for anesthesia with signs of an upper respiratory tract infection (URTI). In such instances, the decision to proceed must be based on the urgency of surgery (i.e., acute middle ear infection) compared with the severity of the URTI. If the child's temperature is normal, behavior and eating habits have been normal, and there are no mucopurulent secretions or chest wheezing/rales, then surgery should proceed (see Chap. 6).

Anesthesia Management

Preoperative

1. Sedation is often unnecessary, but oral midazolam is useful for the very upset child, though it may delay recovery after this brief procedure. Parental presence at induction may be a suitable alternative to premedication.

Perioperative

1. Induce anesthesia by inhalation of sevoflurane or intravenously with propofol or thiopental.
2. Maintain anesthesia with N₂O and sevoflurane by face mask.
3. Tracheal intubation is not required, but a laryngoscope and suitable endotracheal tubes should be immediately available in case of unexpected difficulties. An IV infusion is not usually required, but should be available.
4. Ensure that you can comfortably hold the child's head very still during the procedure—resting your elbow on the table will help with this.
5. With halothane, analgesics are usually not required; oral acetaminophen with local anesthetic ear drops (4% lidocaine) may be used. With sevoflurane, however, analgesia is required usually in the form of IM ketorolac (0.5–1 mg/kg) or IN fentanyl (2 µg/kg). Alternately, the auricular branch of the vagus nerve or the nerve of Arnold may provide effective analgesia when infiltrated with 0.2 mL of bupivacaine 0.25 %.

Postoperative

1. Acetaminophen may be required for ongoing pain.
2. Resume PO fluids when the child is awake.

Major Otologic Procedures: Cochlear Implant, Tympanomastoidectomy, and Tympanoplasty

Anesthesia Management

Preoperative

1. Order adequate sedation, especially for children who have had surgery previously.
2. Hearing aids may be worn until after induction of anesthesia after which time they should be removed until the child recovers.
3. If the child communicates by sign language, then either a parent or health-care professional who can sign should accompany the child to the OR. If the child can lip-read, do not cover your lips until the child is anesthetized.

Perioperative

1. Induce anesthesia by inhalation or intravenously with propofol (or thiopental), followed by a suitable relaxant. If nerve stimulation (usually for facial nerve) is required during surgery, only use a single low dose of intermediate-acting relaxant (rocuronium or *cis*-atracurium) for tracheal intubation. We caution against using succinylcholine because if the child has pseudocholinesterase deficiency, a nerve stimulator will be ineffective (1:3000 children have homozygote atypical pseudocholinesterase; see Chap. 3).
2. Spray the larynx with lidocaine, and then insert an orotracheal (regular or RAE) tube. Extended breathing circuit tubing is usually required as the anesthetic machine is located at the foot of the OR table. Ensure adequate ventilation for the increased breathing circuit compliance.
3. Maintain anesthesia with N₂O/O₂ and an inhalational anesthetic; anesthesia must be deep enough to prevent any possibility of bucking on the tube, which increases bleeding. Supplement with opioids as needed, but beware that this may increase PONV (consider remifentanil).
4. Position the child with a 15° head-up tilt to minimize bleeding.
5. A great auricular nerve block with 2–3 mL of 0.25% bupivacaine with epinephrine before surgical incision and then repeated at the end of surgery will greatly reduce the need for opioids and the incidence of vomiting.
6. If epinephrine is to be infiltrated, ensure that the dose does not exceed the maximum.
7. Prophylactic antiemetic therapy should be given. IV-balanced salt solution should include 20–25 mL/kg to decrease postoperative pain and emesis.
8. For tympanoplasty, discontinue N₂O from the inspired mixture before the graft is positioned. (N₂O bubbles might float the graft off the desired position.)
9. Smooth tracheal extubation, without coughing, is essential. Therefore, administer intravenous lidocaine 1–2 mg/kg or propofol 0.5–1 mg/kg 5 min

before the planned time of extubation, and remove the ETT while the child is still deeply anesthetized and breathing spontaneously. Maintain the airway and allow the child to awaken while administering oxygen by mask.

Postoperative

1. Order analgesics and antiemetics as required.

Awake Ear Surgery

For certain operations (e.g., ossicular reconstruction), the surgeon may wish to assess hearing during the surgical procedure. Most older children are very cooperative if such operations are performed under a combination of sedation and local analgesia.

Anesthesia Management

Preoperative

1. Explain in detail what will happen during the operation, and reassure the child that there will be no pain.
2. Administer PO midazolam in a sufficient dose to ensure adequate sedation preoperatively.

Perioperative

1. Establish an intravenous line using a local anesthetic.
2. Titrate propofol \pm dexmedetomidine infusions until an adequate degree of sedation is achieved (see Chap. 8, Awake Craniotomy, for dosing regimen). Titrate small doses of fentanyl (1–2 μ g/kg) or a remifentanil infusion until the child is comfortable.
3. Ensure that the child is positioned comfortably, and warn the child not to cough or move the head.
4. Talk with the child periodically to assess the effects of the drugs, but allow the child to sleep when cooperation is not required.
5. Consider a great auricular nerve block (see #5 under Major Otologic Procedures for details).
6. Monitor ventilation via nasal capnometry, administer supplemental oxygen, and, if necessary, remind the child to breathe deeply periodically.

Postoperative

1. Smaller than usual doses of analgesics are effective in most cases.
2. Antiemetic medications may be required.

ENDOSCOPY

Endoscopy is often indicated in infants and children for diagnosis (e.g., stridor) or for therapy (e.g., removal of a foreign body).

Procedures

1. Laryngoscopy
2. Bronchoscopy
3. Esophagoscopy

Special Anesthesia Problems

1. Existing airway problem or tracheotomy
2. Difficulty maintaining optimal ventilation during endoscopy, particularly in a child with a very small airway
3. Possibility of complete airway obstruction during some procedures (i.e., removal of foreign body)
4. Danger of airway fire if cautery or laser is used
5. Danger of postoperative reduction in the airway lumen by subglottic edema

N.B. Many conditions for which endoscopy is performed can progress to complete obstruction under anesthesia. Always have a selection of laryngoscopes and tracheal tubes prepared; from the start of anesthesia, ensure that the endoscopist is at hand in case surgical intervention with either a rigid bronchoscope or tracheotomy becomes urgently necessary.

General Anesthesia Management

1. *Spontaneous ventilation* is usually preferred during endoscopy in children. It may be safer than controlled ventilation if there is airway compromise, and it allows the endoscopist to examine the dynamic structures of the airway. Maintaining spontaneous respirations is particularly important when evaluating the child for airway compression, laryngeal, tracheal, or bronchomalacia. Abnormal airway compression or collapse may not be adequately detected during controlled ventilation.
2. *Controlled/assisted ventilation* is necessary for children who are in respiratory failure and for those who cannot maintain effective spontaneous ventilation when anesthetized.

Laryngoscopy

Anesthesia Management

Preoperative

1. Do not give heavy sedation to children with airway problems. Oral midazolam is useful for some older children having repeated endoscopy, but beware of sedating any child with a dubious airway. Children with laryngeal papillomata present a particular risk because of the possibility of ball-valve obstruction, airway fire, or embolism of tumor fragments distally. These cases require complete discussion with the surgeon so that the anesthetic prescription needed for that particular child is clear (i.e., spontaneous ventilation with insufflation, intermittent intubation, or paralysis with apneic oxygenation).

Perioperative

1. Apply monitors, including pulse oximeter, and induce anesthesia by inhalation of N₂O and O₂ with sevoflurane, and insert an appropriate size intravenous catheter.
2. Hydration with 20 mL/kg of lactated Ringer's solution will support the circulation during deep inhalation anesthesia.
3. When the child is deeply anesthetized, discontinue the N₂O, continue with sevoflurane in O₂, perform laryngoscopy, and spray the larynx and supraglottic structures with (preferably 10%) lidocaine (maximum dose, 7 mg/kg).
4. Replace the mask until the lidocaine becomes effective (2–3 min). The duration of the procedure may be brief in some cases but prolonged in others. Anesthesia may be continued with an infusion of propofol (\pm remifentanil).
5. Monitor ventilation visually, with a precordial stethoscope and by capnometry.
6. Additional topical lidocaine applied by the surgeon may be indicated.

N.B. Employing topical analgesia and propofol with spontaneous ventilation without a tracheal tube provides the most satisfactory surgical conditions. Tracheal tubes get into the surgeon's field of vision, and all other methods are cumbersome and complicated and therefore may fail. "Jet ventilation" methods can be dangerous in children, especially if the high-pressure jet migrates distal to an obstructing lesion; fatal pneumothorax and pneumomediastinum may occur.

Postoperative

1. Observe the child closely until awake.
2. Order humidified oxygen/air postoperatively.
3. Maintain NPO status until about 2 h after application of the lidocaine spray.

Special Considerations

Laryngomalacia. A common cause of inspiratory stridor in the neonate, laryngomalacia can be diagnosed during laryngoscopy, while the infant is awake or is awakening from anesthesia. The stridor usually disappears during deeper levels of anesthesia and with small amounts of positive end-expiratory pressure (PEEP); PEEP is especially useful in maintaining a patent airway during the initial phases of induction. In this condition, there is incomplete maturation of the cartilages of the larynx and a tendency for the epiglottis or one of the arytenoid cartilages to prolapse into the glottis during inspiration, causing marked inspiratory stridor. The condition is self-limited and disappears as the child ages; no special therapy is usually required. However, laryngoscopy is indicated to rule out other causes of stridor (e.g., cysts).

Congenital Cysts. Congenital cysts may occur in the region of the epiglottis and aryepiglottic folds. There may be inspiratory and expiratory stridor and a poor cry. The diagnosis is usually confirmed by endoscopic evaluation; therapy is by excision or marsupialization.

Congenital Webs. Complete tracheal webs are generally fatal at birth. However, most webs have a single central perforation (2–4 mm diameter) through which the neonate ventilates. Because of the limited diameter of the perforation and the oxygen requirements of the neonate, respiratory distress often occurs soon after birth. Laryngoscopy may reveal the presence of a web; otherwise, an appropriate sized tracheal tube may not pass below the vocal cords. When the diagnosis is confirmed by bronchoscopy, laser resection effectively restores the airway lumen.

Subglottic Hemangioma. Subglottic hemangioma may manifest with croup-like symptoms and a barking cough, usually commencing at a few months of age. The child frequently has other visible hemangiomas, especially on the face. The symptoms persist or recur, and diagnosis is confirmed with endoscopy. Commonly, therapy is by laser ablation; however, this is sometimes followed by subglottic stenosis. Open resection of the hemangioma is advocated by some.

Anesthesia Management

For laser resection:

1. Manage as for laryngeal papillomata (see later).
2. Humidified oxygen/air should be administered postoperatively after laser therapy.

For open excision:

1. An initial laryngoscopy and bronchoscopy will be performed to assess the lesion. This may be managed with inhalational anesthesia plus topical lidocaine (see later discussion).

2. Following the endoscopic examination, an oral tracheal tube is inserted and general anesthesia continued.
3. An anterior incision in the neck is made to display and open the larynx.
4. At this stage the oral tracheal tube is withdrawn and a second sterile tracheal tube passed by the surgical team via the lower end of the incision into the trachea.
5. The hemangioma on the posterior aspect of the larynx is now resected.
6. On completion of the operation, a nasotracheal tube is passed through the glottis and larynx into the trachea and left in situ.
7. The child is returned to the ICU for continued ventilation, sedation, and paralysis for the initial postoperative days. Immobility of the laryngeal structures is important during the early healing period.
8. The child is returned to the OR for examination before extubation several days later. Dexamethasone is administered before removing the tube. Close observation is required for a further period of 2–3 days.

Laryngeal Papillomas. These cauliflower-like papillomas, caused by the human papilloma virus (HPV), are the most frequent airway tumors in children. They can cause serious airway obstruction and, if untreated, lead to pulmonary hypertension and right-sided heart failure. Various nonsurgical therapies include cryoprobing, ultrasound, and immune sera, but the classic therapy is surgical resection and debulking by CO₂ laser. More recently, two lasers that only penetrate the superficial tissue layers, the KTP/532 and pulsed-dye lasers, and a non-laser approach, the microdebrider, have proven effective. The microdebrider offers the advantages of reduced cost and shorter surgical times although postoperative pain is similar to the CO₂ laser.

Children with this condition usually present at 2–4 years of age and return for repeated laryngoscopies and resection. Recurrences are almost certain until adolescence, at which time the lesions regress spontaneously. Increasing hoarseness and dyspnea are the usual indications for reoperation, and on each occasion, the extent of regrowth is impossible to determine before laryngoscopy. A cautious approach is indicated since extensive papillomas may completely obscure the glottis and create a ball-valve obstruction. Provide humidified gases postoperatively.

Special Anesthesia Problems

1. Acute airway obstruction may occur during induction of anesthesia; therefore, spontaneous ventilation is preferred.
2. The glottic opening may be difficult to visualize; therefore, barbiturates and relaxants are contraindicated.
3. Surgical therapy by laser demands an unobstructed view of the larynx and immobile vocal cords.

4. Instrumentation of the trachea below the glottis should be avoided because it may “seed” papillomas into the lower airways. Therefore, tracheal tubes should be avoided if possible, and tracheostomy is usually considered to be contraindicated.

Anesthesia Management for Laser Surgery

The safest and most widely used plan includes no premedication, careful inhalation induction followed by laryngoscopy, and lidocaine spray to the larynx. If obstruction develops during induction, a tracheal tube must be inserted to reestablish a patent airway. The tube can be removed when a deep plane of anesthesia is achieved and most of the papillomas have been resected. Usually, tracheal intubation is unnecessary, and laser resection can proceed once topical analgesia has been applied. Extremely rarely, critical airway obstruction may occur necessitating an urgent rigid bronchoscopy. In this rare circumstance, inhaled sevoflurane anesthesia may be supplemented or replaced with a propofol or propofol/remifentanil infusion.

Alternatively, a nonflammable tracheal tube¹ (or foil-wrapped tube) may be inserted. In this case ventilation can be controlled, but the surgeon must work around the tube. Another approach to laser surgery in the airway for lesions other than laryngeal papillomas is intermittent intubation by the surgeon with apneic oxygenation.

N.B. Jet ventilation can be very dangerous in cases of obstructing lesions of the airway. Laryngeal obstruction during jet ventilation may lead to pneumomediastinum and pneumothorax. If jet ventilation is used, extreme care must be taken to avoid barotrauma: the jet must not be advanced beyond the lumen of the laryngoscope, and peak airway pressure should be monitored and restricted to <15 mmHg.

Special Precautions for the Use of the Laser

1. Cover the child’s eyes with a wet towel.
2. All personnel in the OR should wear protective goggles for protection in case the laser beam is accidentally reflected in their direction. Post a warning sign on all doors that the laser is in use.
3. Special face masks should be worn to protect against inhalation of vaporized viral tissue.
4. Airway fires are a real danger during laser therapy. Use the minimal FiO₂ (preferable ≤30%) in either air or helium. Avoid N₂O; it supports combustion. If a tracheal tube should ignite, it must be immediately disconnected from the anesthetic circuit and removed from the airway. Injury results both from the burn and from the products of tube combustion.

¹ Phycon laser shielding tube, Fuji Systems Corp., Tokyo

Bronchoscopy

Bronchoscopy may be performed for various indications (e.g., removal of a foreign body, diagnosis of respiratory disease, removal of secretions, treatment of atelectasis, evaluation for tracheomalacia, bronchomalacia, or airway compression by vascular structure or mediastinal tumors).

Special Anesthesia Problems

1. Difficulty maintaining adequate ventilation during the procedure, when the airway must be shared with the endoscopist
2. Existing impaired ventilation

Anesthesia Management

Preoperative

1. Carefully assess the airway and the respiratory status.
2. Do not give heavy premedication if there is any doubt about the airway or ventilation.

Perioperative

Spontaneous ventilation is preferred for all children except those with respiratory insufficiency.

1. Induce anesthesia by inhalation of N₂O and sevoflurane in O₂, and establish adequate venous access.
2. Discontinue the N₂O and deepen anesthesia with sevoflurane in 100 % O₂.
3. When the child is adequately anesthetized, remove the mask and perform laryngoscopy. Spray the larynx and trachea with lidocaine (maximum 7 mg/kg).
4. Replace the mask; continue anesthesia with O₂ and sevoflurane until the lidocaine takes effect (2–3 min), at which time the bronchoscope can be inserted.
5. Supply O₂ and sevoflurane to the side arm and allow spontaneous ventilation, but remember that when a telescope is in use through a small bronchoscope (3.5 mm or smaller), the resistance to ventilation is high. At such times, ventilation should be assisted or controlled, and the telescope may have to be removed periodically. Sevoflurane anesthesia may be supplemented with a propofol or remifentanil/propofol infusion or bolus doses of propofol IV. Be aware that the dead space of the bronchoscope may not allow for accurate detection of expired carbon dioxide. It is important to assess the adequacy of ventilation by observing chest expansion.

6. Monitor oxygen saturation closely; set the sound at an adequate level so that the surgeon is immediately aware if the saturation begins to fall. During bronchoscopy it is useful to have a precordial stethoscope in place and to auscultate over other areas of the lungs.
7. Be alert to the possibility of pneumothorax, a rare complication of pediatric bronchoscopy.
8. When controlled ventilation is essential, as for children in respiratory failure, a Venturi device (e.g., Sanders injector) may be used, but remember that children whose severe chronic respiratory disease has reduced lung compliance may not ventilate well with this method. For such children, control ventilation using the anesthetic circuit connected to the side arm of the bronchoscope (Fig. 10.1).

Postoperative

1. Order nothing by mouth for 2 h (after lidocaine spray).
2. Order humidified O₂/air.
3. Watch for signs of stridor.
4. Check the postoperative X-ray for possible pneumothorax.

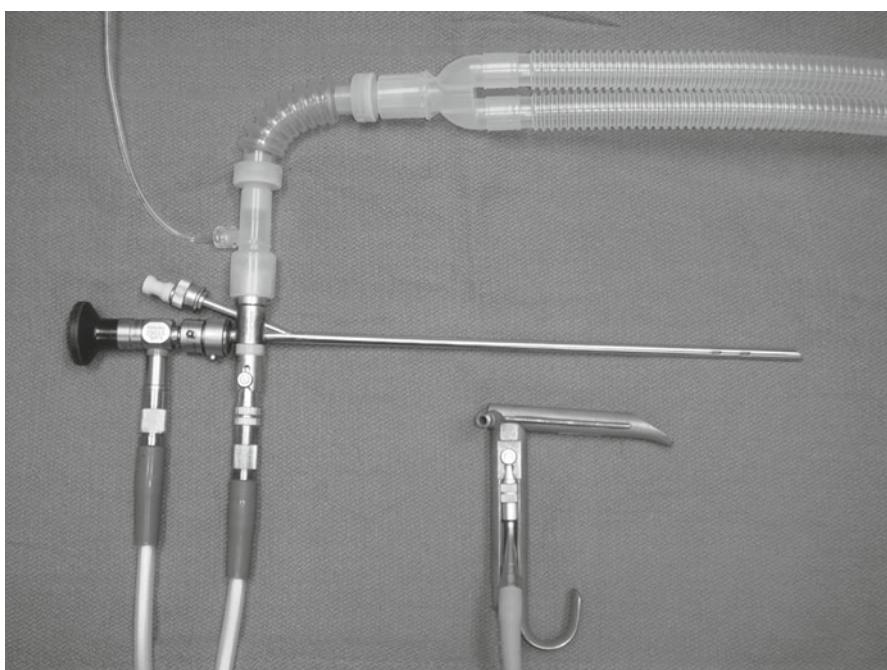


Fig. 10.1 Pediatric rigid bronchoscope with anesthesia circuit attached for controlled/assisted ventilation. Right is anterior commissure laryngoscope

Flexible Bronchoscopy

Small-diameter flexible fiberoptic bronchoscopes permit a diagnostic bronchoscopy to be performed with the child awake or with minimal sedation and topical anesthesia. In young children, however, general anesthesia is often preferred. In such cases, the bronchoscope is passed through an adapter at the elbow into an LMA or into the face mask and via the mouth or nose of the anesthetized child. The LMA is particularly useful as it assures adequate airway patency and a means for continuing to deliver anesthesia. Alternatively, a tracheal tube can be inserted as a conduit for the bronchoscope, although the stem of the LMA is much wider than the tracheal tube that will pass the larynx; thus, a larger bronchoscope can be used with the LMA. In all cases, topical anesthesia with lidocaine spray as described for rigid bronchoscopy is indicated.

Rigid Esophagoscopy

In children, rigid esophagoscopy is usually performed to dilate a stricture or for removal of a foreign body.

Special Anesthesia Problems

1. The child may have undergone esophagoscopy repeatedly and therefore may be very apprehensive. Children with TEF or EA repair are particularly vulnerable to having food, e.g., a piece of hot dog, stuck in the esophagus; be aware of possible associated subglottic stenosis.
2. In small infants, passage of an esophagoscope may compress the trachea and obstruct ventilation, even when a tracheal tube is in place.
3. Coughing, straining, or other movements can result in esophageal perforation during the procedure. Children must be anesthetized adequately to maintain complete immobility.
4. Lower esophageal strictures or achalasia may result in esophageal dilation more cephalad. Food and secretions accumulated in the dilated segment may be aspirated during anesthesia.
5. Rarely, a sharp or potentially damaging foreign body may lodge in the hypopharynx, possibly to be displaced into the airway if the child coughs or strains.
6. Special attention to securing the tracheal tube and manually holding the tube during movement of the esophagoscope will help to prevent unintended extubation of the trachea.

Anesthesia Management

Preoperative

1. Order adequate sedation, especially for children who have undergone esophagoscopy previously.
2. Check whether the radiographs show esophageal dilation and/or retained material.
3. Discuss with the surgeon any special considerations he/she may have, and alter the anesthetic prescription accordingly.

Perioperative

1. Give 100% O₂ by mask.
2. Induce anesthesia intravenously and intubate the trachea rapidly. (Use cricothyroid pressure when there is a risk of regurgitation and aspiration—except if contraindications are present; see Table 4.7)
3. In the case of a sharp foreign body in the hypopharynx, a gentle, smooth inhalation induction of deep anesthesia is indicated.
4. Deepen the anesthesia before permitting the endoscopist to proceed.
5. Monitor ventilation, oxygenation, and hemodynamics using standard monitors.

Postoperative

1. Observe the child until fully awake.
2. Be alert for signs of esophageal perforation, especially if difficulty was encountered. These signs include:
 - (a) Tachycardia
 - (b) Fever
 - (c) Signs of pneumothorax
 - (d) Radiographic evidence of pneumothorax or mediastinal air
3. Order nothing by mouth until 2 h after application of the lidocaine spray.

AIRWAY INFECTIONS

Epiglottitis

Supraglottic infections (epiglottitis) used to be most commonly caused by *Haemophilus influenzae* B (*HiB*) bacterial infection and less frequently by *Staphylococcus* and fungi in severely immunocompromised children. Since the introduction of a *HiB* vaccine, epiglottitis has become a rare disorder in children, but still occurs in adults. As the disease becomes rarer, there is an increased

danger that the diagnosis may be missed. However, epiglottitis can still occur, and there is recent evidence of resurgence in *HiB* epiglottitis because of non-immunized immigrant children, parents who refuse immunization of their children, and occasional ineffective/failed vaccinations.

Epiglottitis is most common in children 3–7 years old but also occurs in infants or adults. It is accompanied by severe systemic illness with pyrexia and leukocytosis. In addition to the epiglottitis, all the supraglottic structures are swollen and inflamed, contributing to the obstruction. Blood cultures are almost always positive for the infective agent, which is typically *HiB*, although other bacterial and fungal infections are possible, but rare. The common symptoms are sore throat, dysphagia, and drooling; severe airway obstruction may develop rapidly. Typically, the child appears toxic and anxious and sits in a tripod position with the chin extended and mouth open. In infants the presentation is less typical, occurring with sudden apnea during investigation of a high fever. Therefore, epiglottitis should be considered in the differential diagnosis of the infant with pyrexia and any respiratory difficulty.

Extra-epiglottic infection may occur: pneumonia, cervical adenitis, otitis media, septic arthritis, and meningitis are described in association with epiglottitis.

Anesthesia Management

Preoperative

1. Once the diagnosis is suspected, the child should be disturbed as little as possible. Avoid venipunctures or painful injections because the child may cry and become acutely obstructed. Do not try to visualize the pharynx because acute obstruction may result. Gently apply a face mask and give O₂.
2. The child must be attended constantly by a physician capable of establishing an emergency airway and equipped to do so.
3. Assemble the team and transfer the child rapidly to the OR; administer O₂, allowing the child to remain in the chosen posture.
4. Soft tissue radiographs of the neck may be misleading and are unnecessary in the typical case. If X-ray studies are required to make the diagnosis, the child must be accompanied to the radiology department by a physician who is capable of establishing an airway should the airway becomes obstructed during the examination. The child should not be made to lie down for the X-ray examination. The lateral neck film will reveal a dilated hypopharynx, thickened aryepiglottic folds, a thumb-like epiglottis obstructing the laryngeal inlet, and loss of the vallecula because of swelling of the tongue surface of the epiglottis.
5. The OR should be prepared for emergency bronchoscopy and possible tracheotomy (surgeon present, scrubbed, and ready to intervene if needed).

If Apnea Occurs (At Any Time)

1. Try to ventilate the lungs with O₂ by bag and mask. This is usually successful.
2. If unsuccessful, proceed to an immediate attempt at laryngoscopy and intubation. Also prepare for an emergency tracheotomy if intubation proves impossible.

Perioperative

1. Gently apply a precordial stethoscope, pulse oximeter, BP cuff, and EKG electrodes.
2. Do not place the child supine. Induce anesthesia with O₂ and sevoflurane (or halothane) by placing the mask gently over the child's face while the child remains seated, either on the OR table or on the anesthesiologist's or parent's knee. Having a parent present may reduce the child's upset and therefore minimize dynamic tracheal collapse with crying against an obstructed upper airway.
3. When anesthesia is induced, gently place the child in the supine position and escort the parents out of the OR. Assisted ventilation may be necessary at this time.
4. Apply other monitors and establish an intravenous infusion; administer intravenous atropine (20 µg/kg) and 20–30 mL/kg of lactated Ringer's solution. These children are febrile and usually dehydrated since swallowing is painful.
5. Anesthetic induction is likely to be very prolonged because of the shallow respirations and possible \dot{V}/\dot{Q} mismatch should the child also have pneumonia. If the airway is satisfactory, there is no urgency to rush the process. Our rule of thumb is that when you think the child is deep enough to perform laryngoscopy, wait 1 more full minute because it is quite easy to underestimate the depth of anesthesia.
6. Administer lidocaine 1–2 mg/kg IV to minimize the risk of coughing and laryngospasm; then perform laryngoscopy and orotracheal intubation with a styttled tracheal tube. In the rare event that the glottis is obscured by swelling and distortion of the supraglottic structures, compress the chest with one brisk compression. This usually expels a bubble through the larynx, providing a guide to the location of the glottis. Another useful trick is to insert the laryngoscope blade down the center of the tongue until the swollen epiglottis is observed. At this point, the tip of the blade is forced into the vallecula (which has been obliterated by the swollen tongue surface of the epiglottis). This will then apply pressure at the base of the tongue and force the epiglottis into a more favorable position for visualization of the laryngeal inlet.
7. Obtain a blood culture once the airway is secured.

8. When the child is anesthetized and well oxygenated, the tube may be changed from an oral to a nasotracheal tube (one size smaller than predicted for age) provided the intubation was not difficult, as follows:
 - (a) Move the oral tracheal tube to the left side of the mouth and allow an assistant to hold that tube while gently assisting respirations.
 - (b) Pass a well-lubricated nasal tracheal tube into the hypopharynx.
 - (c) Perform laryngoscopy and ascertain the location of the nasal tube.
 - (d) With small McGill forceps, position the tip of the nasal tube in the laryngeal inlet immediately next to the orotracheal tube.
 - (e) Remove the oral tube and insert the nasal tube (secure this very firmly—accidental extubation must be avoided).
9. Very rarely, pulmonary edema occurs immediately after intubation for epiglottitis. This has been attributed to a sudden release of the negative pressure caused by breathing against an obstructed upper airway, hypoxia, elevated catecholamines, and disturbed alveolar-capillary pressure gradient. Treatment is with controlled ventilation, PEEP, and diuretics.

Postoperative

1. Constant (24 h/day) nursing care in an intensive care unit is essential. Accidental extubation is a serious early complication and must be prevented by suitable restraints and adequate sedation.
2. Ensure adequate humidification of inspired gases and regular suctioning of the nasotracheal tube. Blockage of the tube may result from tracheal secretions.
3. Commence antibiotic therapy. Cefuroxime, a cephalosporin with a high margin of safety and good penetration of the cerebrospinal fluid, is considered the drug of choice for *Hib* infections.
4. Extubate the trachea after the pyrexia has resolved (usually within 12–36 h). Flexible laryngoscopy may be performed before extubation to examine the state of the supraglottic structures.
5. Observe the child after extubation for several hours. Very rarely does the trachea require reintubation for recurrent obstruction. It is reasonable to perform the extubation in the OR so that if the child fails extubation, reintubation is greatly facilitated.

Croup or Laryngotracheobronchitis

“Acute infectious croup” or laryngotracheobronchitis is most commonly caused by a virus and occurs most often in children 2–5 years of age. Do not overlook other causes including a foreign body. Inspiratory stridor is the principal symptom caused by swelling of the loose tracheal mucosa at the level of the cricoid cartilage. Symptoms are frequently worse at night.

Therapy varies according to the severity of the disease:

1. In mild to moderate cases, conservative measures (oral dexamethasone) is usually effective.
2. In more severe cases, epinephrine inhalations in addition to oral dexamethasone usually result in improvement.
3. Very rarely, nasotracheal intubation is required, generally to facilitate removal of inspissated secretions.

Epinephrine Inhalations

Inhalations of epinephrine are widely reported to be efficacious in cases that fail to respond to conservative measures.

1. Prepare the nebulizer solution of epinephrine: add 0.5 mL of 2.25 % racemic epinephrine to 3.0-mL distilled water.
2. Attach a suitable pediatric-size face mask, and hold this gently on the face, while the child is held comfortably by the mother or father.
3. These children are hypoxic. Add at least 40 % O₂ to the inspired gases; children usually then settle well and accept the mask quietly. Closely monitor oxygen saturation.
4. Monitor ventilation and heart rate via a precordial stethoscope. Some increase in heart rate may occur, but other arrhythmias are very rare.
5. Give the therapy for 20 min, by which time considerable improvement is usually apparent. (If not, the diagnosis of croup should be reconsidered.)
6. A single dose of IV dexamethasone (0.5–1 mg/kg) is also recommended.
7. After inhalations, observe the child carefully. Rebound stridor may occur. Rarely, the stridor increases rapidly, necessitating immediate establishment of an artificial airway.
8. Some children require more than one therapy. Total failure to respond with any improvement is an indication to review and question the diagnosis. The use of racemic epinephrine is contraindicated in infants with tetralogy of Fallot because a severe “tet” spell may be precipitated.

Nasotracheal Intubation

If conservative measures and epinephrine inhalations with IPPB fail to relieve symptoms, an artificial airway may be required. Nasotracheal intubation has been used successfully in many centers, with a small incidence of complications. The critical factor seems to be the diameter of the tracheal tube, which should be

sufficiently small as to provide a leak at approximately 20-cm H₂O peak inflation pressure. The tube is carefully secured and left in place until both oropharyngeal secretions and tissue swelling have diminished. Constant (24 h/day) expert respiratory care is essential; the presence of a small tube and thick secretions renders accidental blockage very likely.

On the rare occasion, a child does not respond as favorably to nasotracheal intubation and cannot be successfully extubated after the standard time. This occurs most commonly in infants, less than 1 year of age, in those with branchial arch deformities or a history of congenital subglottic stenosis and in those with a history of repeated croup. Tracheotomy may be necessary in these children.

Tracheotomy

Tracheotomy may become necessary in the therapy of upper airway obstruction or to facilitate respiratory care in other conditions.

Anesthesia Management

Preoperative

1. Give 100% O₂ by mask, and assist ventilation manually as necessary.
2. Do not give sedatives or opioids.

Perioperative

1. Continue 100% O₂ by mask, induce anesthesia with sevoflurane (or halothane) and O₂, and assist ventilation as required (the intravenous line must be checked for adequacy and replaced if necessary).
2. Deepen anesthesia and spray lidocaine on the larynx. The surgeon may perform a diagnostic bronchoscopy before the tracheostomy, and allow the surgeon to pass a bronchoscope. (Tracheotomy in children is usually performed after passage of a rigid bronchoscope. This makes it easy for the surgeon to identify the trachea and also enables the anesthesiologist to see immediately that the tracheotomy tube has in fact been passed into the lumen of the trachea.) If the surgeon performs the tracheotomy over a tracheal tube, reduce the inspired oxygen concentration, and discontinue nitrous oxide while maintaining a deep level of anesthesia with inhalational or TIVA to minimize the risk of an airway fire. After the surgeon opens the trachea, leave the tracheal tube within the larynx and slowly withdraw until the tip is above the tracheal opening. Only remove once the tracheostomy tube has been successfully placed. N.B. The cuff on a cuffed tracheal tube may be punctured when the surgeon makes his/her initial incision into the trachea.

3. In case of a “difficult airway” (i.e., Pierre Robin syndrome), anesthesia may be induced by mask and continued after insertion of a LMA, which may then be used as a conduit to intubate the trachea.
4. Before leaving the OR, confirm that the tip of the tracheostomy tube is above the carina with a fiberoptic scope while in the position the child will be cared for post operatively. If not, then another more suitable size should be inserted.

Postoperative

1. As soon as possible, obtain a chest radiograph. Check that the tracheostomy tube is positioned correctly and that pneumothorax (a rare complication of tracheotomy) is not present.
2. Be alert to the possibility of accidental extubation before the track into the trachea becomes established. If this happens, it may be very difficult to reinsert the tube. Many surgeons leave long black silk sutures through the edges of the trachea to facilitate emergency reinsertion of the tracheotomy tube.
3. Add an appropriate concentration of oxygen to the inspired gases (to overcome the continuing danger of hypoxemia).
4. Order close, constant observation of the child.
 - (a) Establishment of the airway does not result in immediate return to normal pulmonary function.
 - (b) Respiratory arrest may occur during the postoperative period.

Bacterial Tracheitis

As the incidence of epiglottitis has declined in the pediatric population, bacterial tracheitis has emerged as a more common cause of acute respiratory distress. The disease usually occurs with cough, hoarseness, stridor, and chest retraction. Drooling is rare, but the child is toxic and pyrexial with leukocytosis. Emergency bronchoscopy should be managed as for epiglottitis and reveals that the trachea is inflamed and edematous with purulent secretions and possible pseudomembranes. The responsible organism is usually *Staphylococcus* or *Streptococcus* pneumonia. Treatment is by tracheal intubation, frequent pulmonary toilet, and appropriate antibiotic therapy. Repeat bronchoscopy is indicated to monitor the progress of the disease and recovery.

SUBGLOTTIC STENOSIS

Subglottic stenosis is one of the most common causes of chronic airway obstruction in infants and children. The stenosis may be congenital or acquired—usually as a complication of prolonged endotracheal intubation. Severe subglottic stenosis requires a tracheotomy followed by surgery to reconstruct the subglottic space. The surgical procedure generally involves division of the cricoid cartilage and insertion of a cartilage graft to increase the diameter. A stent is then left in place to maintain the lumen. After extensive repair, it is often preferred to sedate, immobilize the neck, and ventilate the child for several days to optimize healing. Other approaches include a slide tracheoplasty and, more recently, application of 3D-designed external stents. The latter may offer greater success since there is no intraluminal suture line and therefore minimal irritation of intratracheal structures.

Subglottic stenosis, which may prevent successful extubation in neonates, may be treated by early anterior cricoid split without a preliminary tracheostomy.

Associated Conditions (Congenital Type)

1. Congenital heart disease
2. Down syndrome
3. Tracheoesophageal fistula

Anesthesia Management for Cartilage Graft to Cricoid Ring

Preoperative

1. If a tracheostomy is in place, all care and monitoring of the tracheostomy should be continued until the child arrives at the OR.

Perioperative

1. Anesthetize via the tracheotomy tube using sevoflurane in N₂O/O₂.
2. Remove the tracheotomy tube and insert an armored tube via the stoma; suture it firmly in place. (N.B. The lumen of the trachea will take a larger tube than is expected.)
3. Check ventilation to both lungs frequently.
4. Maintain anesthesia with N₂O and sevoflurane or isoflurane, and control ventilation.
5. Blood loss is usually minimal.
6. An endoscopic approach has been advocated by some surgeons.

Postoperative

1. Replace the tracheotomy tube.

2. Administer humidified O₂/air.
3. Intravenous fluids may be required for 1–2 days postoperatively until satisfactory oral intake.
4. A full diet can usually be resumed within a week.
5. The stent is removed, and laryngoscopy is performed under general anesthesia 3 months later.
6. The tracheotomy is left in place until the child is able to tolerate plugging of the lumen of the tube.

For Anterior Cricoid Split (Without Tracheostomy)

This procedure is performed for neonates who cannot be extubated as a result of narrowing at the cricoid.

1. The child is anesthetized via the existing tracheal tube.
2. When the child is anesthetized, the tube may be removed and a bronchoscope inserted to examine the remainder of the airway.
3. The trachea is reintubated and the cricoid cartilage divided anteriorly.
4. An age appropriate size nasotracheal tube is inserted into the trachea and left in place for 5–7 days.

Suggested Reading

American Society of Anesthesiologists Task Force on Perioperative Management of Patients with Obstructive Sleep Apnea. Practice guidelines for the perioperative management of patients with obstructive sleep apnea. An updated report by the American Society of Anesthesiologists task force on perioperative management of patients with obstructive sleep apnea. *Anesthesiology*. 2014;120:268–86.

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Dental Surgery

GENERAL PRINCIPLES

1. Children do not usually cooperate very well under moderate sedation (so-called conscious sedation) unless they are very motivated, older and specially prepared. Consequently, most require deep sedation/general anesthesia more frequently than adults for dental procedures.
2. Many children who present for general anesthesia for dentistry have had previous failed attempts at dental treatment under local anesthesia and sedation and consequently are very apprehensive.
3. Some have behavior disorders or developmental delay and require special consideration, especially those with autism.
4. Others have medical conditions that require special consideration (e.g., congenital heart disease).
5. Nasotracheal intubation is preferable for children having dental surgery. Nasal intubation per se is associated with bacteremia and is an indication for prophylactic antibiotics for endocarditis prophylaxis if heart disease is present (see later discussion). Children with extensive caries show positive blood cultures after dental procedures.
6. Special care must be taken to ensure that no foreign bodies remain in the airway at the end of the procedure (especially throat packs). Counting throat packs is essential. Direct laryngoscopy must be performed before extubation to ensure that the airway is clear.
7. Dental procedures may be prolonged when extensive disease is present. In such instances, recovery to a normal appetite is not as brisk as after short operations. Therefore, children should receive intraoperative intravenous fluids to restore euvoolemia. It is preferred to limit the duration of general anesthesia for the outpatient to a maximum of 4 h and to schedule surgery for such children to commence in the morning.
8. For procedures to be carried out under sedation plus local analgesia, monitoring should be applied as for general anesthesia.
9. Rarely, the use of air turbine dental drills has been a cause of intraoperative subcutaneous and mediastinal emphysema, leading to airway obstruction and possible pneumothorax. If facial swelling occurs, discontinue nitrous

oxide, check for pneumothorax, and be prepared to support ventilation. Very rarely, these complications may present in the postoperative period after tooth extractions.

MANAGEMENT FOR GENERAL ANESTHESIA

Preoperative

1. A careful preoperative history and physical examination should be performed as dentists are not authorized to perform “medical assessments” of patient conditions in most jurisdictions. Previously unrecognized significant disease is often discovered in children presenting for dental surgery.
2. Special investigations and treatments, as appropriate, should be ordered for children with other comorbidities.
3. Premedication should be tailored to the child’s needs. Upset children may benefit from a suitable dose of oral midazolam (0.75 mg/kg for children younger than 6 years of age, 0.3–0.5 mg/kg for older children) preoperatively. Every effort should be made to reassure and gain the confidence of the upset child.
4. Make sure that special drugs are administered at the right time (e.g., antibiotics for children with heart disease).
5. It may be helpful to have the parent accompany the child to the induction area or to insert an intravenous line with the parents present before admission to surgery.

Perioperative

1. Apply standard American Society of Anesthesiologists’ monitors.
2. Induce anesthesia by inhalation or with propofol intravenously.
3. If an inhalation induction is performed, establish appropriate IV access.
4. Nasotracheal intubation is associated with a bacteremia, and endocarditis prophylaxis is recommended for those at risk (see later discussion).
5. To reduce the risk of a nosebleed after nasotracheal intubation, a soft catheter may be used (see below) and/or a topical vasoconstrictor (oxymetazoline). Liberal lubrication and warming the tip of the nasotracheal tube have not been shown to reduce bleeding.
6. Administer propofol and/or a non-depolarizing muscle relaxant as indicated, and preoxygenate before performing a nasotracheal intubation. One published approach to minimize nosebleeding is to telescope the nasal tube into a soft red rubber catheter (usually latex) and drop the lubricated catheter tip along the floor of the nose until it reaches the nasopharynx. Using

the laryngoscope light to illuminate the oropharynx, extract the tip of the catheter via the mouth with Magill forceps, and dislodge the catheter from the tip of the tube. Thus, the soft catheter guides the tip of the tube atraumatically through the nasal passages. Laryngoscopy is then performed, and the tube is directed into the glottis with Magill forceps. Since this technique involves extra steps, oxygen desaturation is more likely if the child is already cyanotic, has intrinsic lung disease, and when this technique is practiced by less experienced anesthesiologists; therefore, preoxygenate well beforehand.

The left nostril is preferred for nasotracheal intubation since the bevel of the tube is on the left and the turbinates are less likely to be damaged. Furthermore, as the tube exits the nasopharynx, the leading edge (tip) of the tube (which is on the right side) is in the midline, directing the tube between the vocal cords.

7. The tip of the tracheal tube may fail to pass through the vocal cords despite the tube being the correct size. This occurs because the angle that the tube makes as it exits the nasopharynx is acute and the tip impacts the anterior commissure of the vocal cords. Any one of three remedies solves this problem: (A) rotate the tube so the bevel faces anteriorly and then advance, (B) pick up the tube with Magill forceps 2–3 cm proximal to the distal end and angle the distal end posteriorly so the tip passes, or (C) withdraw the tube slightly, lift the child's head to rest on the operator's abdomen, and flex the child's neck. This aligns the axes and the tip of the tube should pass easily. If none of these approaches is successful, consider an orotracheal tube.
8. Maintain anesthesia with N₂O and inhalation agent in O₂. For brief procedures, spontaneous ventilation will suffice. For more prolonged procedures, controlled ventilation may be more appropriate; if so, decrease the inhalational anesthetic concentration and monitor blood pressure carefully.
9. Administer maintenance fluids during the surgery; 20–30 mL/kg of a balanced salt solution.
10. Keep track of the total dose of local anesthetic administered by the dentist/oral surgeon to avoid toxicity.
11. At the end of the procedure, when all dental instrumentation has been removed, a gentle laryngoscopy should be performed to ensure that the airway is free of debris or foreign material before extubation. Beware of throat packs that may have been placed in the hypopharynx and forgotten.

Postoperative

1. Order analgesics as required. (Dental nerve blocks with local anesthetic reduce the requirement.) Acetaminophen and NSAIDs are usually sufficient after dental restorations. Primary teeth often have short roots and do not

cause pain when extracted. However, IV morphine or ketorolac may be required after multiple extractions. Ensure that the child is provided with analgesic drugs for use after discharge.

2. Antiemetics may be required, although dental surgery is not associated with a high incidence of PONV.
3. Continue intravenous fluids until the child is ready for discharge.

MANAGEMENT FOR DEEP SEDATION

1. Currently, propofol is probably the most useful drug for deep sedation, allowing good moment-to-moment control of sedation and ensuring a rapid recovery. Alternatives include ketamine and propofol in combination and dexmedetomidine infusions.
2. Preoperative care and intraoperative monitoring should be as for general anesthesia.
3. Preoperative medication with midazolam (see dosing above) and prior application of a topical anesthetic cream facilitate insertion of an intravenous catheter.
4. Drugs and equipment for intubation and ventilation with O₂ should be at hand.
5. Deep sedation should be commenced with small increments of (0.5–1-mg/kg propofol) as needed for induction so as to avoid apnea, followed by a continuous infusion, beginning with as much as 250 µg/kg/min (larger doses may be necessary for younger and cognitively impaired children), reducing the rate progressively to 75–100 µg/kg/min as the local anesthetic blocks become effective. The exact dose requirement for continued anesthesia varies from child to child.
6. Oxygen may be given and end-tidal carbon dioxide sampled by a septate nasal catheter. The airway is usually well maintained, but continuous close monitoring is essential.

THE CHILD WITH CONGENITAL HEART DISEASE

The child should be managed for anesthesia with all the necessary considerations for his or her cardiac disease (see “Noncardiac Surgery in Children with Congenital Heart Disease” in Chap. 14):

1. *Recommendations for prophylactic antibiotics in children with CHD having dental procedures.* The cardiac defects for which prophylactic antibiotics are now indicated are listed in Table 11.1. Antibiotic dosing based on the recommendations of the American Heart Association (2007) is listed (see Table 11.2).

Table 11.1 Indications for endocarditis prophylaxis

- Patients who should receive prophylactic antibiotics include those with:
- Prosthetic cardiac valve or prosthetic material used for cardiac valve repair
 - Previous infective endocarditis
 - Congenital heart disease (CHD)
 - Unrepaired cyanotic CHD, including palliative shunts and conduits
 - Completely repaired CHD with prosthetic material or device, whether placed by surgery or catheter intervention, during the first 6 months after the procedure
 - Repaired CHD with residual defects at the site or adjacent to the site of a prosthetic patch or prosthetic device (which inhibit endothelialization)
 - Cardiac transplantation and cardiac valvulopathy

Table 11.2 Antibiotic dosing for endocarditis prophylaxis

Regimen: single dose 30–60 min before procedure		
Considerations	Antibiotic	
Able to take oral medication	Amoxicillin	50 mg/kg
Unable to take oral medication	Ampicillin or Cefazolin or ceftriaxone	50 mg/kg IM or IV 50 mg/kg IM or IV
Allergy to penicillins or ampicillin—able to take oral medication	Cephalexin or Clindamycin or azithromycin or Clarithromycin	50 mg/kg 20 mg/kg 15 mg/kg
Allergy to penicillins or ampicillin <i>and</i> unable to take oral medication	Cefazolin or Ceftriaxone or Clindamycin	50 mg/kg IM or IV 50 mg/kg IM or IV 20 mg/kg IM or IV

- Dental procedures that require prophylactic antibiotics include all dental procedures that involve manipulation of gingival tissues or the periapical region of teeth or perforation of the oral mucosa, specifically, extractions, periodontal procedures including surgery, scaling and root planning, probing, dental implant placement, reimplantation of avulsed teeth, subgingival placements, intraligamentary local anesthetic injections, placement of orthodontic bands, endodontic instrumentation, surgery beyond the apex of the tooth, and prophylactic cleaning of teeth or implants (where bleeding may occur).
- Dental procedures that do *not* require prophylactic antibiotics include routine anesthetic injections through noninfected tissue, taking dental radiographs, placement of removable prosthodontic or orthodontic appliances, adjustment of orthodontic appliances, placement of orthodontic brackets, shedding of deciduous teeth, and bleeding from trauma to the lips or oral mucosa.

Suggested Reading

- Chicka MC, Dembo JB, Mathu-Muju KR, et al. Adverse events during pediatric dental anesthesia and sedation: a review of closed malpractice insurance claims. *Pediatr Dent.* 2012;34(3):231–8.
- Dorman ML, Wilson K, Stone K, Stassen LF. Is intravenous conscious sedation for surgical orthodontics in children a viable alternative to general anaesthesia? A case review. *Br Dent J.* 2007;202(11), E30.
- Oncag O, Aydemir S, Ersin N, et al. Bacteremia incidence in pediatric patients under dental general anesthesia. *Congenit Heart Dis.* 2006;1(5):224–8.
- Watt S, Pickhardt D, Lerman J, et al. Telescoping tracheal tubes into catheter minimizes epistaxis during nasotracheal intubation in children. *Anesthesiology.* 2007;106:238–42.

Plastic and Reconstructive Surgery

Many children require plastic surgery to correct congenital deformities. The head and neck are commonly affected, which may introduce special problems for the anesthesiologist. In addition, some children undergo plastic surgery for acquired lesions, such as burn scars and contractures or dog bites.

GENERAL PRINCIPLES

1. Many of these children have psychological upsets stemming from both the deformity and multiple surgical procedures. A careful, considerate approach by the anesthesiologist is essential. Review prior anesthetic records to adequately assess previous experience with premedication, airway management, venous access, and other associated medical or surgical issues.
2. A smooth induction of anesthesia and a quiet emergence limits the risk of damage to grafted areas and delicately sutured repairs.
3. Many children undergoing plastic surgery have potentially serious airway problems that require careful assessment and special management (See Chap. 4).
4. Congenital structural anomalies commonly affect more than one body system. The child with defects requiring plastic surgery may also have disease affecting other systems. If congenital heart disease is present, consider the need for prophylactic antibiotics (see Chap. 14); carefully review the most recent cardiac evaluations and the cardiac surgical history; seek consultation with the cardiologist when indicated.

CLEFT LIP AND PALATE

Cleft lip and cleft palate (CLP) are present in various combinations in as many as 1 of every 1000 live births. CLP defects may be isolated or occur as part of a syndrome or association of defects (syndromic CLP). Infants with these lesions

may be both malnourished and anemic as a result of feeding difficulties and may have a history of repeated respiratory infections. The treatment of CLP is optimally managed within a multidisciplinary team; the pediatric anesthesiologist is an essential member of this team.

The surgical management of CLP is evolving, and debates continue regarding the optimal age for stages in the repair and the use of such procedures as preoperative orthodontic maneuvers to mold the bony structure of the palate. An orthodontic appliance (e.g., Latham appliance) is sometimes applied to the neonatal palate with the object of progressively molding the bony alveolar ridge into alignment before soft tissue repairs.

1. Congenital heart disease—not specifically associated with isolated cleft palate, but may be present as part of a syndrome or association (see Appendix A).
2. Airway anomalies, for example, Pierre Robin sequence or Treacher Collins syndrome, may make intubation extremely difficult.
3. Syndromes associated with CLP may have other specific anesthesia implications (Table 12.1).

The surgical care of the patient with CLP now comprises many possible stages, not all of which will be required by all children.

Possible Surgical Procedures

1. Application of orthodontic splints or devices to the palate
2. Cleft lip repair—usually performed at 3–4 months of age, sometimes earlier
3. Cleft palate repair—usually performed at 12–18 months, but sometimes in younger infants
4. Alveolar bone graft
5. Pharyngoplasty—for velopharyngeal incompetence—usually performed at 5–15 years
6. Maxillary advancement (Le Fort procedure) required for some adolescent patients

Special Anesthesia Problems

1. Airway problems, including difficult tracheal intubation (sometimes extreme). Intubation with isolated micrognathia tends to be difficult in the young infant and becomes easier with age (e.g., Pierre Robin sequence). This may not apply if a syndrome is present (e.g., Treacher Collins syndrome).
2. Problems related to associated conditions (see previous discussion).
3. As plastic surgery involves soft tissue dissection, coagulopathy causes significant bleeding. Blood loss during cleft palate repair is usually insignificant but very occasionally may require transfusion.

Table 12.1 Some syndromes associated with cleft palate

Syndrome name implications	Features	Anesthesia
Arthrogryposis multiplex congenita	Limb contractures, CHD in 10% Stiffness of joints, GU defects	Difficult intubation due to limited mouth opening, antibiotics for CHD, position and pad carefully (see Page 531)
Beare-Stevenson syndrome	Craniosynostosis, hydrocephalus choanal atresia, midface hypoplasia proptosis, hypertelorism, cutis gyratum	Difficult ventilation (choanal atresia) difficult intubation, beware of tracheal stenosis, caution with neck. Cervical spine defects
Beckwith-Wiedemann syndrome	Exomphalos, macroglossia, visceromegaly, neonatal hypoglycemia	Danger of hypoglycemia, infuse glucose and monitor level, tongue reduction may be required at time of palate repair
CATCH 22	Cardiac defect, abnormal facies, thymic hypoplasia, hypocalcemia (DiGeorge syndrome)	Difficult airway. Antibiotics for CHD. Treat hypocalcemia use only irradiated PRBCs
Cornelia de Lange syndrome	Short stature, developmental delay (variable), CHD in 15%	Airway obstruction, difficult intubation, antibiotics for CHD
Down syndrome	Short stature, developmental delay (variable), macroglossia, unstable cervical spine, narrow subglottic space. CHD in 50%	Difficult intubation, caution with neck movement, caution with ETT size, antibiotics for CHD, prone to airway obstruction
EEC syndrome	Ectrodactyly, ectodermal dysplasia, hypohidrosis Chronic respiratory infections	Malnutrition, anemia, hypohidrosis, temperature control problems. Difficult intubation, protect eyes, position and pad carefully. Caution with atropine
Kabuki syndrome	Craniofacial and skeletal defects, hypotonia anomalies, CHD, visceral and urogenital defects, increased susceptibility to infections	Difficult airway, caution with relaxants, antibiotics for CHD, care with asepsis
King syndrome	Congenital myopathy, MH trait. Dysmorphic features (Noonan-like)	MH precautions
Miller syndrome	Mandibular defects, limb anomalies, renal defects	Difficult airway
Multiple pterygium syndrome	Webbing of skin, syngnathia, ankyloglossia, web neck	Difficult airway—more severe with age

Table continues on the following page.

Table 12.1 (continued)

Syndrome name implications	Features	Anesthesia
Nager syndrome	Malar hypoplasia, micrognathia, CHD, radial hypoplasia, absent thumbs, vertebral anomalies	Very difficult airway, limited mouth opening, cervical spine anomalies. Antibiotics for CHD
Oto-palatal-digital syndrome	Skull deformity, hearing loss, cervical spine defect, (Arnold-Chiari malformation), limb defects, possible thoracic hypoplasia	Possible brain-stem compression causing postoperative respiratory depression
Patau syndrome (trisomy 13)	Microcephaly, developmental delay, micrognathia CHD, usually fatal in infancy	Difficult airway, antibiotics for CHD
Pierre Robin sequence	Micrognathia, cleft palate	Difficult airway, postoperative airway obstruction
Seckel syndrome	Birdlike facies, dwarfism microcephaly, possible glottic narrowing; post-op apnea reported	Difficult airway, caution with ETT size. Monitor ventilation postoperatively
Smith-Lemli-Opitz syndrome	Growth failure, microcephaly, developmental delay, CHD, renal defects, hypotonia, GE reflux Thymic hypoplasia—prone to infection	Possible difficult airway, intraoperative muscle rigidity, temperature control problems, antibiotics for CHD
Spondyloepiphyseal dysplasia congenita	Dwarfism, C1–C2 instability	Caution with neck during intubation and positioning (use vacuum splint)
Stickler syndrome (Pierre Robin variant)	Midface hypoplasia, cleft palate, retromicrognathia, “moon face” one-third of “Pierre Robin” patients	Difficult airway (mask ventilation and intubation)
Walker-Warburg syndrome	Micrognathia, hypotonia, hydrocephalus, developmental delay, GU anomalies	Difficult intubation, postoperative hypoventilation
Trisomy 18	Cleft palate, lung hypoplasia, micrognathia, CHD	Difficult airway, ventilatory failure, antibiotics for CHD

CHD is congenital heart disease; GU is genito-urinary; ETT is tracheal tube; MH is malignant hyperthermia

Anesthesia Management

Preoperative

1. Conduct a thorough preoperative assessment.
 - (a) Special attention to the airway, lungs, and other systems that may be affected in congenital syndromes (see Appendix A).

- (b) Check for an acute upper respiratory tract infection; if such an infection is present, consider postponing surgery. It should be noted that children with cleft palate have frequent and often chronic URTI symptoms. If the problem is chronic then postponing surgery is not indicated.
 - (c) Check for anemia.
2. Check for any history of bleeding tendency. Check medication history (salicylates (e.g., aspirin), NSAIDs, gingko, garlic, or ginseng herbal medicines). If positive, determine the bleeding time; if it is prolonged, surgery should be deferred.
 3. Blood is rarely needed for cleft palate surgery, but check that the child has been typed for blood and serum saved (type and hold).

Perioperative

1. If there is any doubt about the ease of tracheal intubation, perform an inhalation induction and manage as for “difficult airway” (see Chap. 4). Consider starting an IV before anesthesia as tolerated.
2. For inhalation induction, administer nitrous oxide (N_2O) and oxygen (O_2) with sevoflurane and establish IV access. Then discontinue N_2O and deepen inhalational anesthesia until the child is anesthetized adequately for laryngoscopy. Give lidocaine (1.5 mg/kg IV) and propofol (1–2 mg/kg) before insertion of the laryngoscope to minimize the risk of coughing or laryngospasm.
3. If the cleft is large or bilateral, some recommend inserting a hard plastic “tooth guard” on the maxillary alveolar ridge to provide a smooth surface to stabilize the laryngoscope blade.
4. Use an oral RAE¹ preformed tracheal tube. Check carefully that bilateral ventilation of the lungs is present after the mouth gag is positioned. In infants, particularly with micrognathia, the preformed RAE tracheal tube may need to be pulled back to prevent an endobronchial intubation. Flexion of the neck during insertion of the mouth gag may advance the tip of the tube into a bronchus. Extension of the head tends to withdraw the tip of the tube, which could result in extubation if the tube is secured high in the trachea. It is essential to reassess breath sounds after positioning for surgery.
5. For cleft lip repair, tape the tube carefully to the mandible and ensure that the upper lip is free and not distorted by the surgical tape.
6. Reassess air entry and ventilation each time the gag is repositioned or the child is moved.

¹ RAE = Ring-Adair-Elwyn

7. Maintain anesthesia with N₂O, controlled ventilation, and sevoflurane or desflurane. The inhalational agent should be discontinued before the end of the operation so that the child awakens promptly. Otherwise, a TIVA regimen may be used.
8. Local anesthesia with epinephrine is often infiltrated by surgeons for analgesia and to reduce surgical bleeding. The use of an infraorbital nerve block (see Chap. 5) is an effective block for this type of surgery. IV acetaminophen should be considered (see Table 7.3 for infant dosing). To ensure an awake patient and a patent airway at extubation, we recommend administering opioids at the beginning of surgery (e.g., fentanyl (2–4 µg/kg) or morphine (50 µg/kg)).
9. Monitor blood loss and replace if indicated. The infiltration of a local anesthetic with 1:200,000 epinephrine reduces blood loss in cleft palate surgery and also provides some analgesia postoperatively.
10. Inspect the mouth and pharynx gently at the end of surgery; use a laryngoscope and remove all blood and clots. Extubate when the child is fully awake. After palate repair, acute swelling of the tongue causing obstruction has been reported as a complication of the use of the tongue blade on the mouth gag. Therefore examine the mouth carefully; if any signs of tongue swelling exist, the trachea should be left intubated.
11. A long suture is often inserted into the tongue to exert traction and facilitate immediate postoperative control of the airway in the PACU after cleft palate repair. This is usually removed when the child is fully awake but may be left in place overnight until a clear airway is ensured.

Postoperative

1. Cleft lip surgery is often performed as an ambulatory procedure because the surgery is superficial and postoperative problems are rare.
2. All children are admitted overnight after cleft palate surgery. Ensure constant observation for 24 h because airway problems or bleeding may occur. A small percentage of children may have to return to the OR for control of bleeding and some may require reintubation.
3. Carefully titrate postoperative opioids to avoid respiratory depression as upper airway obstruction is a particular risk in the postoperative period.

Application of Palatal Splints or Devices in the Neonate

This is sometimes performed to mold the palate and alveolar ridge and improve the aesthetic and dental results of subsequent surgery. All provisions for neonatal anesthesia should be observed. General endotracheal anesthesia is preferred with an oral RAE tube. After the procedure only a mild analgesic (e.g., acetaminophen or NSAIDs) is required.

Alveolar Bone Grafting

An alveolar bone graft may be required to close a gap in the bony alveolar ridge; this procedure may be performed on an ambulatory basis. The operation is preferably performed as the permanent dentition erupts so that teeth may be guided into the grafted area. The bone is often harvested at the iliac crest; pre-incisional infiltration of the donor site with bupivacaine with epinephrine may significantly reduce postoperative pain and bleeding. IV morphine is usually required intra- and postoperatively. Prophylactic administration of dexamethasone and ondansetron significantly decrease PONV and speeds return to normal activity.

Pharyngoplasty

Pharyngoplasty is performed to reduce velopharyngeal incompetence and improve speech. The procedure inevitably increases resistance to ventilation. Postoperative airway problems are common requiring close monitoring postoperatively.

Special Anesthesia Problems

1. Postoperative airway obstruction is a particular danger and may occur early in the PACU.
2. Chronic airway obstruction may persist after the operation and may lead to obstructive sleep apnea and possibly to later pulmonary hypertension.

Anesthesia Management

As for cleft palate (see previous discussion).

Postoperative

1. Observe closely in the PACU or PICU overnight for airway obstruction and/or bleeding.
2. A nasopharyngeal airway, placed by the surgeon intraoperatively and left in situ for 24 h, is effective in reducing serious postoperative respiratory complications.
3. Do not order large doses of opioids.
4. Monitor for signs of obstruction during sleep; a postoperative sleep study is recommended.

FRACTURED MANDIBLE

Surgical Procedures

1. Interdental wiring
2. Open reduction and wiring

Special Anesthesia Problems

1. The child may have a full stomach.
2. Intubation may be difficult because of tissue damage, trismus, and distortion of airway anatomy.
3. Foreign bodies may be present in the airway (i.e., teeth).
4. The mouth may be wired closed after the procedure; therefore postoperative vomiting is a potentially life-threatening event. Suction should be immediately available.
5. Wire cutters must be at the child's bedside at all times in case the trachea must be intubated.

Anesthesia Management

Preoperative

1. Assess the child for trismus that may limit mouth opening and preclude a thorough examination. Neither trismus nor a fractured mandible portends a difficult airway in general.
2. Determine the more patent nostril for intubation.
3. For children with a full stomach, delay surgery if possible and give metoclopramide 0.15 mg/kg IV.
4. Do not give heavy sedation.

Perioperative

1. For emergency surgery, use a RSI (see Chap. 4).
2. Examine the pharynx quickly but carefully during laryngoscopy to search for foreign debris. (Radio-opaque debris such as teeth may be visible on X-ray, consult radiographs before inducing anesthesia.)
3. For elective surgery, a nasotracheal tube may be inserted initially if optimal conditions for laryngoscopy are present. For emergency surgery, use an orotracheal tube initially to secure the airway and then change to a nasotracheal tube. (If you attempt a nasotracheal intubation initially, a nosebleed

may complicate the full stomach. Once the oral tube is in place, insert a nasogastric tube and aspirate the gastric contents. Then pass a nasal tube through one nostril, repeat the laryngoscopy, and exchange tracheal tubes.)

4. Ask the surgeon to pack the throat with sterile gauze.
5. Maintain anesthesia with N₂O and an inhalational anesthetic and/or a propofol infusion plus relaxant using controlled ventilation. Multimodal antiemetics should be administered. These approaches permit rapid recovery with minimal PONV.
6. Before final fixation of the jaw, remove the throat pack and inspect the pharynx with a laryngoscope; remove blood clots and other debris.
7. If the mandible is not wired to the maxilla, inspect the oropharynx for debris before discontinuing anesthesia in preparation for extubation. Extubate after assessing recovery from neuromuscular blockade, when spontaneous respirations are present and the child has recovered consciousness; this is especially important if the mandible is wired to the maxilla. With wire cutters present, withdraw the tube to the oropharynx and remove the tube completely from the nostril when you are certain the child can protect the airway and ventilation is sustainable.
8. Leave the nasogastric tube in place for use during the postoperative period.

Postoperative

1. Observe the child closely.
2. Ensure that wire cutters remain at the bedside at all times for children whose jaws are wired closed.

Removal of Interdental Wiring

General anesthesia is usually required for removal of the wiring and arch bars when the fracture is healed. The wires holding the jaws together can be removed before induction of anesthesia. However, jaw movement remains extremely restricted because of the prolonged immobilization, possibly rendering laryngoscopy and intubation difficult.

Ensure that the child has been fasted preoperatively. After removal of the securing wires, anesthesia may be induced with a variety of techniques such as inhalation agents, TIVA, or ketamine until a well-lubricated soft nasopharyngeal tube can be inserted. When the depth of anesthesia and ventilation are adequate, the surgeon may remove the arch bars; have equipment for emergency intubation and/or tracheotomy immediately at hand.

RECONSTRUCTIVE SURGERY FOR BURNS

Children who suffer a burn injury require acute and chronic (reconstructive) burn care and surgery. Anesthesia for acute burn care is addressed in Chap. 17. Anesthesia for repeat reconstructive plastic surgery for release of contractures and hand/foot reconstruction is addressed below.

Special Anesthesia Problems

1. Contractures that result from burns to the face and neck may make laryngoscopy and tracheal intubation and maintenance of the airway during anesthesia very difficult.
2. Extensive burns may make it difficult to obtain reliable venous access.
3. Severe emotional problems may result from the accident, from the subsequent disfigurement, and from the repeated surgical procedures. The last problem often gives rise to fear of anesthesia face mask and fear of venous access; ask the child or teenager which approach is preferred.
4. Premedication with oral midazolam is advisable, and some may benefit from the addition of oral ketamine and atropine if they have a history of inadequate response to oral midazolam premedication.
5. Blood losses for reconstructive procedures is generally not very extensive.
6. Temperature homeostasis is impaired, and special measures must be taken to avoid hypothermia.
7. Emergence from anesthesia should be quiet to avoid damage to recently grafted areas.

Anesthesia Management: General Endotracheal Anesthesia

General endotracheal anesthesia may be required if operating on the face or in the prone position. An LMA is usually appropriate for most other reconstructive procedures and for laser treatment of scars. Even after recovery from the acute burn, there are additional special considerations:

1. A 40 % larger dose of thiopental is required for children undergoing reconstructive surgery for several years after injury; comparable data for propofol are lacking. Some children may require IM ketamine for induction if no IV access is available.
2. Succinylcholine is contraindicated after the first 24 h after a burn and for 1.5–2 years after severe burns because it may cause cardiac arrest secondary to hyperkalemia.
3. The dose requirements for non-depolarizing muscle relaxants is increased compared with those in unburned children for several years after the injury and the magnitude of the increase is in proportion to the size of the burn.

Relaxants should be titrated to achieve the desired effect using a neuromuscular blockade monitor.

4. If the airway has been injured in the burn, subglottic stenosis may be present. Carefully select the size of the tracheal tube but prepare to use a smaller tube than expected. Some children may have a tracheostomy in place.

Preoperative

1. Conduct a standard preoperative assessment with particular emphasis on previous problems with the adequacy of premedication, a difficult airway or difficult venous access. Often the child is able to tell you where “the best vein” is located.
2. Take time to talk with the child. Encourage questions, answer them honestly, and reassure the child about the planned procedure. Having a parent present during induction is exceedingly important to this cohort of children.
3. Have difficult airway adjuncts (e.g., GlideScope, fiberoptic equipment, difficult airway cart) immediately available.
4. Administer adequate preoperative sedation either IV or by mouth.
5. Make sure that the OR is warmed to 25 °C.
6. Confer with the surgeon regarding any special issues such as a specific route of intubation (oral or nasal) or tracheal tube (standard or RAE), eye care, positioning, site of graft harvesting, limits on local anesthetics, and postoperative surgical concerns.

Perioperative

1. If airway problems are not present:
 - (a) Induce anesthesia by either IV or inhalational route.
 - (b) If an LMA is appropriate, then tracheal intubation is not required. If intubation is required then proceed as indicated following inhalation induction and establishment of IV access with propofol or propofol plus non-depolarizing relaxant.
 - (c) Measure blood losses and be prepared to replace them when excessive.
2. If airway problems are suspected (i.e., microstomia or neck contractures):
 - (a) If the chin cannot be extended or the mouth opened, direct visual intubation may be impossible. Select from the following alternatives:
 - Perform fiberoptic intubation, either sedated with topical analgesia or after an inhalation induction, depending on the child’s age and ability to cooperate.
 - Use a GlideScope or other advanced airway adjuncts to facilitate tracheal intubation while maintaining spontaneous respirations.
 - Release scar tissue under local analgesia and/or ketamine analgesia, induce anesthesia, and perform tracheal intubation under direct vision.

- (b) Once the airway has been secured, anesthesia can be maintained with either an inhalational anesthetic or TIVA-based technique.
- 3. These surgeries may be painful (particularly if a large skin graft is taken), and regional anesthesia or a field block is usually not possible. Opioids are essential adjuncts to these surgeries; the dose requirements are often increased because of residual tolerance from chronic use in the PICU.
- 4. All patients should receive multimodal PONV prophylaxis.

Postoperative

- 1. Emergence from anesthesia should be quiet; do not disturb the child. Order adequate analgesic drugs as required.
- 2. Emergence delirium and difficulty of pain control are common; often low-dose ketamine is highly effective in the PACU. Be aware that some children will have severe itching (opioids and old scars) requiring treatment with an antihistamine such as diphenhydramine.

MAJOR CRANIOFACIAL AND RECONSTRUCTIVE SURGERY

Extensive reconstruction is now widely performed to restore more normal facial features in children with severe facial deformities. The improvement in appearance frequently has a major beneficial effect on the child's future life; in some countries, these children cannot leave their home without restorative surgery. Much of this surgery is performed during infancy or early childhood with the objective of allowing the child to go to school looking as normal as possible.

In recent years, the technique of external maxillary distraction osteogenesis has been applied to children requiring maxillary osteotomies. The osteotomies are performed subperiosteally and external distraction appliances attached with which the healing bone is drawn slowly apart after a short period to allow callus formation. These procedures are usually brief in duration and associated with minimal blood loss. However, external appliances may limit the anesthesiologist's access to the airway.

General Principles

- 1. A team approach is essential for successful performance of this type of surgery.
- 2. Operations involving the jaws are usually delayed until dentition is complete (i.e., 13 years or older).

3. Operations not involving dentition (e.g., for craniofacial dysostosis) are usually performed at an earlier age.

Special Anesthesia Problems

1. Intubation of the airway may be difficult if the deformity is severe. In rare instances, the airway cannot be secured despite the use of all adjunctive intubation techniques; in these cases, tracheotomy is performed using local analgesia and sedation preoperatively.
2. Blood loss may be very extensive from the surgical and bone graft donor sites (e.g., pelvic girdle or ribs).
3. Surgery is of long duration, and special precautions must be taken to protect the child against complications of prolonged anesthesia (i.e., pressure sores, nerve compression, eye protection, and hypothermia).
4. Surgical manipulation involving the orbit and face may initiate the oculocardiac reflex.
5. Surgical manipulations may damage the tracheal tube intraoperatively (rare).
6. The children must awaken rapidly after surgery so that the surgeon can assess cranial nerve function.
7. Extensive postoperative swelling of adjacent tissues may dictate the need for prolonged intubation after the operation.

Anesthesia Management

Preoperative

1. A thorough preoperative assessment is required; particular attention to airway abnormalities and cardiopulmonary disease.
2. Consider the possibility of associated congenital defects or other features of a syndrome that may have implications for anesthesia (see Appendix A). Some children (i.e., those with Apert or Crouzon syndrome) may have OSA, consider reducing opioids if $\text{SaO}_2 < 85\%$ at night (see page 280).
3. Check all laboratory results, especially for the presence of a coagulopathy.
4. Have difficult airway adjuncts (e.g., GlideScope, fiberoptic equipment, difficult airway cart) immediately available.
5. Ensure that adequate supplies of blood and blood products will be available and that serum has been saved for further cross-matching if necessary.
6. Reassure the child and family and explain the planned procedures, discuss the reasons and need for venous and arterial lines, urinary catheter, nasogastric tube, and possible postoperative ventilation.
7. Administer preoperative sedation: midazolam for children or lorazepam (Ativan) for adolescents.

Perioperative

1. Induce anesthesia:
 - (a) If there are no airway problems, anesthesia may be induced by either IV or inhalational techniques; tracheal intubation should follow.
 - (b) If a difficult airway is anticipated, anesthesia may be induced using an inhalational technique and the airway instrumented using difficult airway adjuncts as needed (see Chap. 4).
 - (c) In some children with an extremely difficult airway, a tracheotomy may be performed with sedation under local analgesia or with general anesthesia with an LMA. This assures a secure airway for the procedure and postoperatively until swelling subsides.
2. If the trachea is intubated, the tracheal tube should be sutured or wired in position, with due consideration being given to the movements of the facial bones that may accompany the surgery. The tube should either be sutured or wired to a structure that will not be moved (e.g., teeth) or be so positioned in the trachea as to allow for the effects of movement at its point of fixation. An armored tube is useful as it cannot be compressed during surgery, but it does not prevent the tube from being incised.
3. Maintain anesthesia with N₂O (but if there is a risk of air embolism, substitute oxygen/air for oxygen/N₂O), an opioid (i.e., fentanyl (2–3 µg/kg/h), morphine (0.05–0.1 mg/kg/h), or remifentanil (0.05–0.15 µg/kg/min)) and an inhalational anesthetic. A non-depolarizing muscle relaxant may be used for prolonged surgery.
4. Control ventilation to an arterial carbon dioxide pressure (PaCO₂) of 30–35 mmHg.
5. Position the patient with 10°–15° head-up tilt.
6. Pad all pressure areas well, including occiput and areas compressed by the tracheal tube (i.e., nares, lip).
7. Place protective ointment in the eyes (the surgeon will usually perform tarsorrhaphy).
8. Monitor:
 - (a) Standard anesthetic monitors
 - (b) Arterial blood pressure and central venous pressure if indicated, by direct means; if head-up tilt is indicated, assure that the arterial transducer has been zeroed at the level of the child's auditory canal.
 - (c) Acid-base, blood gas, and hematocrit by serial determinations
 - (d) Temperature
 - (e) Coagulation indices (if a massive transfusion occurs)
 - (f) Urine output
 - (g) Precordial Doppler ultrasound/end tidal Pco₂—for air embolism during craniectomy

9. Be prepared for massive blood replacement (See Chaps. 4 and 17).
10. If reduction in brain mass is required, give furosemide (1 mg/kg) or mannitol (0.5 g/kg, slowly over several minutes to avoid hypotension).
11. When indicated, induce moderate hypotension using one of isoflurane, remifentanil, or sodium nitroprusside.
12. The child should be sufficiently awake in the OR at the end of surgery, so that the surgeon can check the child's vision and ascertain whether cranial nerve injury has occurred during surgery. The child can then be re-sedated if the tracheal tube is to be left in place postoperatively.

Postoperative

1. Order routine post-craniotomy nursing care when applicable.
2. Leave the tracheal tube in place until the child is fully awake *and* there is no further danger that postoperative tissue swelling might obstruct the airway. (Many children require intubation for 48–72 h postoperatively.) The timing of tracheal extubation must be based on the adequacy of spontaneous ventilation, the resolution of airway swelling, the presence of a leak around the tube, and the level of consciousness of the child. Dexmedetomidine sedation is very useful in these children. Some children will be most safely extubated if returned to the OR. Intravenous dexamethasone may be administered before extubation to prevent stridor.
3. Particular caution should be exercised if external distraction apparatus has been applied. In these cases and whenever in doubt, an airway exchange catheter may be inserted via the tracheal tube and left in place after extubation to aid in reintubation should this be required. The airway exchange catheter can then be removed when it is apparent that reintubation will not be required.
4. Observe caution when using opioid analgesics.
5. Check hemoglobin and hematocrit to ensure adequacy of blood replacement.

Children with Mandibular Distraction Osteogenesis

Neonates and infants with severe airway difficulties and associated difficulty feeding may be managed by one of several techniques: tongue-lip adhesion and a nasopharyngeal tube or tracheostomy. Although tongue-lip adhesion maintains a patent airway, it does not facilitate a better view, and in some cases, the fixation of the tongue may render the airway *more* difficult to secure by tracheal intubation. Recently, mandibular distraction osteogenesis (MDO) has been advocated in children with persistent respiratory distress, difficulty feeding, or a distance from the post-pharyngeal wall to the lingual root <3 mm. One clear indication for MDO is to ensure successful decannulation of the neonate with a tracheostomy for airway difficulties. Difficulty with tracheal intubation may be

anticipated based on the airway defect (Pierre Robin sequence or Treacher Collins syndrome). The infant's airway should remain intubated (particularly if the airway was difficult preoperatively) after application of the external fixator until the infant fully recovers or the mandible has been distracted. The mandible is usually distracted 0.5 mm three times a day beginning 2–3 days after surgery, a process that continues for 4–8 weeks. If the distraction device must be modified or adjusted with general anesthesia, mask anesthesia may present challenges, although once the mandible has been distracted, tracheal intubation is usually much easier.

TUMORS IN THE NECK: CYSTIC HYGROMA AND CONGENITAL CERVICAL TERATOMA

Cystic hygroma is a cystic lymphangioma that usually occurs in the neck and less commonly in the axilla. Intraoral extension of this benign tumor may cause airway obstruction. Three percent of cervical tumors extend into the mediastinum.

Teratomas may occur in the neonate, and though most occur in the sacral region, they may also involve the neck. Giant congenital cervical teratoma may cause life-threatening airway compromise.

Occasionally extremely large tumors in the neck are diagnosed by antenatal ultrasound and require intervention to establish an airway at the time of birth (see “EXIT procedure” below; see page 328).

Special Anesthesia Problems

1. Airway obstruction may be present.
2. Intubation may be difficult because the airway is distorted or difficult to visualize.
3. Complete removal of tumor may involve extensive dissection and be accompanied by major blood loss.

Anesthesia Management

Preoperative

1. Airway examination is a priority, radiological investigations should be reviewed; consider intrathoracic extension of the tumor.
2. Do not administer heavy sedation.
3. Ensure availability of blood and blood products for transfusion.

4. Have difficult airway adjuncts (e.g., GlideScope, fiberoptic equipment, difficult airway cart) immediately available.

Perioperative

1. Induce anesthesia cautiously by inhalation of N₂O and sevoflurane in oxygen. Maintain spontaneous ventilation.
2. When the child is anesthetized, establish a reliable large-bore intravenous route.
3. Before attempting intubation, discontinue N₂O and give O₂ and sevoflurane for several minutes. Coughing and breathholding and/or laryngospasm during attempts at intubation may be minimized by administering propofol (1–2 mg/kg) and lidocaine (1.5 mg/kg IV).
4. Intubate the trachea (consider an armored tube) and secure the tube firmly. For some children, if intraoral dissection is planned, a nasal tube may be preferable. In the case of a difficult intubation, insert an oral tube first and then change to a nasal tube if this is possible without risking losing the airway.
5. Maintain anesthesia with either inhaled anesthesia or TIVA and controlled ventilation.
6. For large tumors requiring extensive dissection, an arterial line and central venous access should be inserted; a urine catheter should also be placed.
7. Beware of vagal reflexes during dissection in the neck. Give a vagolytic (atropine or glycopyrrrolate) if these reflexes occur.
8. Replace fluid and blood losses guided by the blood pressure, CVP, and measured losses.
9. After the operation, prepare for a smooth emergence and extubation; prevent excessive coughing and bucking, which might cause bleeding at the surgical site.
10. If extensive surgery has been performed adjacent to the airway, extubation should be delayed until the extent of postoperative swelling is determined. If swelling is significant, the trachea should remain intubated until it resolves; rarely some children may require tracheotomy.

Postoperative

1. If extubated:
 - (a) Observation in the PICU or PACU overnight (because of the danger of bleeding into the surgical site or compression of the airway) is indicated.
 - (b) Avoid large doses of opioid analgesics.
2. If intubated:
 - (a) Confirm the position of the tracheal tube radiographically.
 - (b) Order appropriate humidified O₂ in air, sedation, and continuing care.

THE EXIT PROCEDURE: EX UTERO INTRAPARTUM TREATMENT

This procedure is designed to maintain the infant by means of the placental circulation until an airway can be established and pulmonary ventilation commenced (airway management on placental support). The essentials are as follows:

1. The position of the placenta is delineated to avoid damage at hysterotomy.
2. The mother is anesthetized using a technique that will cause uterine relaxation and prevent placental separation, high concentrations of inhalational anesthetics, and/or nitroglycerin. Maternal blood pressure is optimized by positioning and vasopressors as required. Massive rapid maternal blood loss is possible and a rapid infusion device should be set up and ready for use.
3. Hysterotomy is performed and the head of the fetus accessed. The airway is established by tracheal intubation, bronchoscopy, surgical tracheostomy, or initiation of ECMO.
4. Once airway and ventilation are satisfactory, the umbilical cord is clamped and the infant transferred to the NICU or another OR for further treatment as appropriate; some will require an urgent tracheostomy.

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General and Thoraco-Abdominal Surgery

GENERAL PRINCIPLES

1. Many children who require thoraco-abdominal procedures are neonates and preterm infants and that demands special considerations.
2. Understanding the pathophysiology of the lesion on normal neonatal physiology dictates the choice of optimal anesthesia management.
3. Immediate surgery is rarely required; usually, time is available for preoperative evaluation and resuscitation as indicated. Optimal timing for surgery is decided by consultation among the anesthesiologist, neonatologist, and surgeon.
4. For emergency abdominal surgery, the problem of the full stomach must be considered. Even if the child has not eaten for some time, gastric emptying may be delayed, e.g., by obstruction, ileus, or need for opioids. Children with severe gastroesophageal reflux are at increased risk.
 - (a) Remember the effects of drugs on the barrier pressure (i.e., lower esophageal pressure [LES] minus intragastric pressure). Barrier pressure is reduced by atropine, midazolam, ketamine, propofol, dexmedetomidine, inhalational anesthetics, and cricoid pressure. It is increased by metoclopramide, rocuronium, pancuronium, vecuronium, and little changed by succinylcholine.
 - (b) Consider administering drugs to reduce the volume and acidity of gastric contents: cimetidine, ranitidine, metoclopramide, and sodium citrate.

Possible plans of action for a full stomach:

- Neonates and small infants: aspirate stomach contents through a vented gastric tube in the supine, right, and left decubitus position (this may remove up to 95 % of fluid gastric contents), preoxygenate, and perform an RSI with cricoid pressure or awake sedated intubation (see Chap. 4).
- Older children: aspirate stomach contents (if appropriate) and perform an RSI with cricoid pressure (as indicated), which must be

commenced as soon drugs that may reduce LES are given. There is no relaxant that can replace succinylcholine for speed of onset, offset, and intensity of neuromuscular block; rocuronium (1.2 mg/kg) may be substituted if there is a contraindication to succinylcholine.

- Assure that the child is deeply anesthetized and relaxed before tracheal intubation. Struggling during attempts at intubation in an incompletely anesthetized and relaxed child is a common precursor to vomiting and aspiration.

Remember: During RSI, succinylcholine does not increase intragastric pressure in children <10 years of age; pretreatment with a low-dose non-depolarizing relaxant is not indicated (see Chap. 3).

5. During some thoraco-abdominal surgery procedures, blood loss may be considerable; be prepared for a major blood transfusion (see Chaps. 4 and 17).
6. For major abdominal surgery, always establish intravenous access in the upper limbs or neck. The inferior vena cava (IVC) may rarely be clamped, compressed, or lacerated rendering transfusion via lower limb veins ineffective.
7. N₂O diffuses into the lumen of gas-containing bowel, causing further distention and difficulties for the surgeon; do not use N₂O in children with intestinal obstruction.
8. The airways are small and, during lung surgery, bronchial secretions (often sanguinous) may accumulate; perform tracheobronchial toilet as indicated.
9. During thoracotomy, \dot{V}/\dot{Q} ratios in the lungs are disturbed; increase the inspired O₂ concentration to maintain an acceptable SaO₂.
10. In infants and small children, retracting the lungs may obstruct major airways, impairing ventilation, or compress the heart and great vessels, leading to a precipitous decrease in cardiac output and blood pressure. Constant monitoring of breath sounds via stethoscope, the shape of the capnogram, and observation of SaO₂ are essential. In the event of decreasing saturation, bradycardia, hypotension, or impaired ventilation:
 - (a) Ask the surgeon to remove all retractors immediately.
 - (b) Ventilate the lungs with 100% O₂.
 - (c) Use large manually delivered tidal volumes with relatively high peak inflation pressures to re-expand areas of atelectasis (or alveolar recruitment).
11. Even children who require minor surgery (e.g., herniotomy) may be preterm and/or have other conditions (e.g., anemia) that can complicate anesthesia and require special precautions (see Chap. 6).
12. Many general surgery procedures can now be performed with the use of video-assisted endoscopic techniques, which offer important advantages over open approaches.

SPECIAL CONSIDERATIONS FOR MINIMALLY INVASIVE/ ENDOSCOPIC SURGERY IN INFANTS AND CHILDREN

Advances in optical systems and instruments have allowed the extensive development of endoscopic techniques for infants and children. Advantages are smaller scars, less postoperative pain, earlier discharge, and presumably less psychological upset. Robotic surgery via endoscopic access is becoming more common for select surgical conditions.

Special Considerations

1. All endoscopic procedures may require urgent open operation should complications arise; always be prepared for emergency laparotomy or thoracotomy.
2. Bleeding may be difficult to rapidly control without open surgical exposure; ensure that reliable large-bore intravenous routes are established.
3. Access to the child may be limited by the equipment required for robotic surgery; ensure that you can adequately monitor the child and rapidly intervene should this be required.
 - (a) Laparoscopic procedures
 - Physiologic changes secondary to a CO₂ pneumoperitoneum.
 - The diaphragm is splinted with a consequent decrease in chest wall compliance; this effect is increased in the Trendelenburg position. Elevation of the diaphragm is followed by decreasing lung volumes, increasing the potential for airway closure during tidal breathing and consequent hypoxemia.
 - Increased intra-abdominal pressure (IAP) leads to decreased venous return, decreased cardiac output, and increased total systemic resistance. The magnitude of these effects is related to the IAP and is increased in the reverse Trendelenburg position. These hemodynamic effects are magnified in the volume-depleted child.
 - Carbon dioxide is readily absorbed into the circulation raising the PaCO₂ and resulting in the requirement for increased ventilation. Positioning, pneumo-peritoneum, and the requirement for increased ventilation may cause the peak inspiratory pressure (PIP) to markedly increase. Absorption of CO₂ may be greater in children than in adults owing to the physiologic properties of the peritoneum.
 - The extent of the above changes depends upon intra-abdominal pressure. Pressures up to 5 mmHg cause little effect; up to 12 mmHg is generally well tolerated by healthy children in the horizontal position. Positions other than horizontal or IAP greater than

12 mmHg may cause serious cardiorespiratory derangements that require interventions. Limiting the IAP and careful positioning may permit some children with cardiorespiratory disease to tolerate pneumoperitoneum but some may not be ideal candidates for this surgery (e.g., children with Fontan physiology and passive venous return to the lungs).

- Anesthesia management.

- Careful preoperative assessment of cardiorespiratory status, state of hydration, and volume replacement is necessary. Respiratory or cardiac disease and/or volume depletion could cause adverse responses to induced pneumoperitoneum. Volume status should be optimized (i.e., in infants with pyloric stenosis) and any cardiorespiratory limitations considered carefully preoperatively.
- Controlled ventilation with increased positive inspiratory pressure is required. Oxygenation and PetCO₂ should be closely monitored though the latter may be unreliable and give false low readings in infants. Increases in FIO₂ and minute ventilation (MV) are required. MV may need to be increased by 66% to maintain near homeostasis. Up to 5 cm PEEP may be required.
- Nitrous oxide should not be used:

When bowel distention is possible.

When air embolism might occur.

Intra-abdominal cautery is used. (N₂O supports combustion.)

- A gastric tube should be passed to empty the stomach and improve exposure. For lower abdominal surgery, either have the child empty the bladder preoperatively or pass a urinary catheter. For pyloromyotomy, a small amount of air may be injected into the stomach via the gastric tube to check the integrity of the duodenal mucosa.
- In small infants, the cephalad movement of the diaphragm and lungs may cause the tip of the endotracheal tube to enter a bronchus; frequently assess bilateral ventilation. Pneumothorax is rare complication and should be watched for. The addition of PEEP may decrease the adverse effects of high IAP and reduce the potential effects of pneumothorax.
- Heart rate should be carefully monitored during all intra-abdominal manipulations; a Bezold–Jarisch reflex may occur with initial insufflation and brisk vagal reflexes may also occur requiring immediate cessation of surgical manipulations and treatment with atropine.
- Carbon dioxide embolism is very rare, but could cause cardiovascular collapse. Monitor for sudden decreases in PetCO₂, unexplained decreases in blood pressure, or development of a windmill murmur.

(b) Visually assisted thoracic surgery (VATS)

- Physiologic considerations.

- Infants have a high metabolic rate for oxygen and a high V_a/FRC ratio. Any compromise in ventilation rapidly leads to hypoxemia.
- In older patients in the lateral decubitus position for intrathoracic procedures, ventilation of the uppermost lung exceeds that of the dependent lung, but perfusion favors the dependent lung. There is increased mismatch. In infants, because of their highly compliant ribs and small size, there is a smaller distribution of blood to the dependent lung and less \dot{V}/\dot{Q} mismatch.
- In order to visualize the intrathoracic structures, some degree of lung collapse must occur, resulting in further \dot{V}/\dot{Q} mismatch. This may be obtained by selective one-lung ventilation (OLV) or as a result of gas insufflation into one hemithorax.
- OLV tends to direct perfusion away from the unventilated lung because of hypoxic pulmonary vasoconstriction (attenuated in part by inhalational anesthetics) and mechanical effects within the collapsed lung thus normalizing \dot{V}/\dot{Q} relationships somewhat.
- Absorption of insufflated CO_2 from the pleural cavity requires increased pulmonary ventilation.
- PEEP is helpful in preserving ventilation of the dependent lung.

In summary, oxygenation and ventilation during VATS is unpredictable requires careful monitoring, and a higher FIO_2 will be necessary, especially in small infants.

- Anesthesia management for VATS.

- Carefully evaluate the cardiorespiratory system and volume status.
- Plan an approach for OLV if requested:

A tracheal tube (*no* Murphy eye) guided into the contralateral main bronchus is the usual option for small infants. An uncuffed tube 0.5 mm ID smaller or a cuffed tube 1 mm ID smaller than predicted for normal use should be used. The tube can usually be inserted blindly into the (R) main bronchus although right upper lobe atelectasis may occur in this situation. A fiberoptic bronchoscope or stylet is usually necessary to enter the (L) main bronchus. Using this technique, it is not possible to suction or inflate the operated side without withdrawing the tube.

A bronchial blocker may be placed on the operated side. A Fogarty catheter has been used in small infants although there is no central channel for suction or to allow the lung to collapse completely; in addition, it has a high pressure cuff, which might

cause damage to the bronchial mucosa. Low pressure cuffs are found on biliary catheters, which may also be used as a bronchial blocker. Bronchial blockers in small infants may be difficult to accurately position and may become displaced—possibly causing serious airway obstruction.

In children greater than 2 years of age, the Arndt 5 Fr pediatric bronchial blocker may be used, which has a high volume low pressure cuff. With the guide wire removed, this channel may also be used to suction or deflate the operated lung.

This blocker is inserted via a special adaptor into the endotracheal tube and guided into position using an FOB passed through the wire loop on the tip of the blocker. The use of the Arndt 5 Fr blocker passed through the trachea external to the tracheal tube has also been described. The blocker is passed into the trachea before intubation and positioned either by fluoroscopy or with the aid of the FOB. Another approach is to use an FOB to intubate the lung to be blocked and use the tube as a conduit to place a bronchial blocker; then remove this tube and replace it with a cuffed ETT with the tip above the carina.

The Univent tube may be used in children of 6 years or over and has the advantage of a channel to suction the operated side bronchus. It should be positioned with the aid of an FOB. The Univent tube is available in two pediatric sizes that may be suitable for older children:

3.5 mm Univent tube—external diameter 7.5–8 mm (=5.5 mm ID plain tube)

4.5 mm Univent tube—external diameter 8.5–9 mm (=6.5 mm ID plain tube)

Double lumen tracheal tubes (26 Fr) may be successfully placed in children of 6–8 years of age. Correct positioning should always be confirmed with the FOB.

- Anesthetic agents that interfere with normal hypoxic pulmonary vasoconstriction (e.g., halothane) should be avoided to ensure that pulmonary blood flow is optimally distributed away from the collapsed lung. Low concentrations of isoflurane or intravenous agents (e.g., propofol, fentanyl) are preferred.
- During OLV, the child must be closely monitored to detect any displacement of the device used. If obstruction to ventilation should occur, the cuff of a blocker should be immediately deflated.
- Following a period of OLV, the lung on the operated side should be carefully observed for reinflation. A postoperative chest X-ray should be obtained to exclude persistent atelectasis.

CONGENITAL DEFECTS THAT MAY NECESSITATE SURGERY DURING THE NEONATAL PERIOD

Congenital Lobar Emphysema

Abnormal distention of a lobe (usually the upper or middle lobe) compresses the remaining normal lung tissue and displaces the mediastinum; respiratory distress and cyanosis result. When severe, this condition manifests as an extreme emergency during the early neonatal period. Obstruction of the bronchus supplying the distended lobe may be extrinsic (e.g., abnormal blood vessels), intraluminal (e.g., bronchial papilloma), or it may be caused by a defect of the bronchial wall (bronchomalacia). More than one lobe may be involved, and sometimes the disease may be bilateral. The chest radiograph demonstrates a hyperlucent area with sparse lung markings (differentiating it from pneumothorax) and mediastinal shift. The lesion must also be differentiated from congenital diaphragmatic hernia (CDH) or cystic adenomatoid malformation, which can have a similar radiologic appearance. Less severe forms may pass unnoticed for months or even years, and conservative management may be appropriate in some cases.

Associated Condition

1. Congenital heart disease—an incidence of up to 37% in reported series.

Surgical Procedure

1. Lobectomy (if no intraluminal or extrinsic cause can be found and corrected). This may be performed using VATS in children other than those in extremis.

Special Anesthesia Problems

1. Severe respiratory failure may occur because of compression of normal lung tissue.
2. There is the possibility of a “ball-valve” effect, which further increases the size of the affected lobe during positive-pressure ventilation.
3. N₂O may cause further distention of the lobe and is contraindicated.
4. Considerations for the use of VATS.

Anesthesia Management

1. Observe the usual special considerations for neonates.

Preoperative

1. The child is cared for in the semi-upright position.

2. Give O₂ as needed. Avoid intermittent positive-pressure ventilation (IPPV) if possible (danger of “ball-valve” effect).
3. Insert a gastric tube and apply continuous suction to prevent gastric distension further compromising ventilation.
4. Check that blood is available.
5. Sudden serious deterioration of the child’s condition may demand immediate emergency thoracotomy to exteriorize the affected lobe and allow the normal lung tissue to ventilate.

Perioperative

1. Bronchoscopy to exclude intraluminal obstruction may be performed before thoracotomy or VATS.
2. Give atropine and preoxygenate.
3. If a bronchoscopy is planned, induce anesthesia with sevoflurane in O₂. Otherwise, induce anesthesia with either sevoflurane in O₂ or ketamine. Spray the vocal cords with 1% lidocaine while maintaining spontaneous respirations and intubate the trachea or pass the bronchoscope. After bronchoscopy, change to a tracheal tube.
4. Maintain anesthesia with either sevoflurane or ketamine with spontaneous respirations. If assisted ventilation is required this should be applied very gently.
5. VATS can usually be managed with a tracheal tube and gentle positive pressure ventilation during inflation of the thorax with CO₂. Rarely is there a need to isolate the lung. In such instances consider:
 - (a) Selective endobronchial intubation by advancing the tracheal tube into the bronchus of the contralateral lung. FOB should be used to verify the location of the tip of the tube while the neonate is supine. Intubation of the right bronchus may occlude the right upper lobe bronchus causing desaturation. Advancing the tube into the left bronchus may be accomplished by rotating the tube 180° before advancing it. Correct tube position should be verified after repositioning in the decubitus position.
 - (b) In older children, bronchial blockers may be inserted into the affected bronchus either endoscopically or radiographically. The blocker should be small, highly compliant, and its position verified. Blockers are inserted outside the tracheal tube lumen and positioned by passing an FOB through the tracheal tube and verifying the blocker location after inflation. These blockers may dislodge easily during surgery and suddenly obstruct the tracheal tube. Be prepared to deflate them should this occur.
6. Continue with spontaneous or very gently assisted ventilation until the thorax is open or accessed. In children in extremis, the affected lobe balloons out

of the chest on thoracotomy and ventilation must be controlled. If VATS is used, CO₂ will be insufflated into the affected hemithorax. Careful control of ventilation and monitoring of oxygenation during OLV is required. A high FIO₂ will be required. Permissive hypercapnia may be required during this period.

7. A non-depolarizing neuromuscular blocking drug may facilitate controlled ventilation and minimize the need for inhaled anesthetic vapors once the lobe has been controlled.
8. After the affected lobe has been excised, the remaining lung will expand to fill the thorax although a pneumothorax may remain.

Postoperative

1. Discontinue all anesthetic drugs, administer 100 % O₂ and antagonize muscle relaxants.
2. Suction and remove the tracheal tube when the infant is awake.
3. Place the infant in a heated incubator and supply O₂ as required to maintain SaO₂.
4. A chest drain (connected to underwater drainage and suction) is required.

Anesthesia Management: Older Children

In approximately 10 % of cases, a congenital emphysematous lobe is discovered at an older age. If surgery is planned, the children should be managed as outlined for younger children. A double lumen endotracheal tube may be used to facilitate surgery or VATS.

Congenital Diaphragmatic Hernia

The incidence of CDH is 1 in 4000 live births. There are three common types: anterior through the foramen of Morgagni, posterolateral via the foramen of Bochdalek, and at the esophageal hiatus. The most common type is through the foramen of Bochdalek, usually on the left side (80 %). Herniation of abdominal contents into the thorax causes respiratory distress, mediastinal displacement ("dextrocardia"), and a scaphoid abdomen. Breath sounds are absent over the affected side; bowel sounds may be heard over the thorax. The radiographic appearance is usually diagnostic.

In many children with CDH, both lungs may be severely hypoplastic. Currently, CDH is thought to be a primary failure of lung development associated with a failure of diaphragm development. The infant with CDH is usually in severe respiratory distress at or soon after birth, often the diagnosis has been made antenatally by fetal ultrasound.

Associated Conditions

1. Malrotation of the gut (40%)
2. Congenital heart disease (15%)
3. Renal abnormalities (less common)
4. Neurologic abnormalities
5. Cantrell's pentalogy (defined as CDH, omphalocele, sternal cleft, ectopia cordis, and intracardiac defect (VSD or diverticulum of left ventricle))

Surgical Procedure

1. Reduction of the hernia and repair of the diaphragmatic defect; usually, a trans-abdominal procedure—often performed via the laparoscope.

Special Anesthesia Problems

1. Optimal preoperative preparation of the child: The trend in recent years is not to rush to surgery. Relief of compression of the lungs by reduction of the herniated abdominal viscera usually does not solve the problem; indeed, there is evidence that respiratory mechanics and hemodynamics are worse postoperatively. It is now preferred to treat the respiratory insufficiency by muscle paralysis, controlled gentle ventilation, and therapy to reduce pulmonary vasoconstriction (including surfactant, hyperventilation, oxygenation, correction of metabolic acidosis, anesthesia and paralysis, and nitric oxide). If these measures fail, ECMO may be instituted. Surgical correction is performed later, as an elective procedure, when the infant is improving and can be weaned from respiratory support.

Anesthesia Management***Preoperative***

Preoperative management requires the facilities and trained staff of a specialist unit. The infant is nursed in a semi-upright, semilateral position, facing toward the involved side. A gastric tube is passed and maintained on low suction to prevent further distention of intrathoracic abdominal viscera. All but the exceptionally fit older infant require intubation and ventilation: bag-mask ventilation should be avoided because it may further distend the stomach and increase respiratory distress.

1. Muscle paralysis after intubation facilitates controlled ventilation and minimizes struggling, thereby decreasing the O₂ demand. It also reduces airway pressure, minimizes further lung damage, and diminishes the danger of pneumothorax. (Pneumothorax is a constant danger and must be watched for and immediately treated.)

2. Ventilation should be gentle—not to exceed positive inspiratory pressures of 20 cm H₂O. Some degree of hypercapnia is permitted rather than using high pressure that might cause further lung damage.
3. High-frequency oscillating ventilation (HFOV) may be applied while minimizing pressure swings, which might cause further lung damage. Surfactant therapy may be administered to preterm infants with CDH.
4. Pulmonary vascular resistance may be reduced by general measures such as fentanyl infusion and minimal handling of the child. Nitric oxide may be administered by inhalation and may further reduce pulmonary vascular resistance in some children although the results of NO in CDH children are unimpressive.
5. When all of these measures fail, ECMO is indicated and may permit survival of many infants until the pulmonary status improves.

Aggressive invasive monitoring using arterial pressure and ECHO-derived pulmonary artery pressures is required to ensure optimal treatment for the pulmonary status. The best predictors of the degree of pulmonary hypoplasia, and hence of survival, are the PaCO₂ and the respiratory index (the product of mean airway pressure and respiratory rate). Those children who are easy to ventilate and not grossly hypercarbic have a better prognosis. Those who are hypercarbic and hypoxic with a high mean airway pressure are less likely to survive. ECMO may increase survival of this latter group. If the child improves on ECMO, surgery is usually performed just before weaning.

Perioperative

CDH may be repaired in the NICU or in the OR. Children on ECMO and HFOV and those who have circulatory instability often undergo surgery in the NICU; most surgeons prefer to transport all other children to the OR for surgery.

1. Induce and maintain anesthesia with high-dose IV fentanyl (>12 µg/kg) as tolerated. Ventilate with low-dose inhalation agent as tolerated and O₂/air to maintain SaO₂. N₂O is contraindicated.
2. Monitor airway pressure. This should *not* exceed 20 cm H₂O (greater pressures may cause further lung damage or contralateral pneumothorax).
3. Do not try to expand the lungs after reduction of the hernia (lung damage may result).
4. Monitor blood gas and acid–base status frequently and correct as indicated.

For children having surgery while requiring ECMO:

1. A common approach is to administer additional doses of relaxant and opioids. However, infants on ECMO often develop tolerance to fentanyl and require very large doses to blunt cardiovascular responses to surgery. Instead, low concentrations of isoflurane may be titrated to blunt the responses by adding it to the oxygenator gas supply.

2. Ensure ECMO cannulas do not become kinked during positioning for surgery.
3. Even though the child may be heparinized, excessive bleeding usually is not a problem.

Postoperative

1. Return the child to the neonatal intensive care unit (NICU) for continued intensive respiratory care.
2. Some infants who have been salvaged by heroic intensive care measures may remain oxygen dependent for years.

Tracheoesophageal Fistula and Esophageal Atresia

Tracheoesophageal fistula (TEF) and interrelated esophageal atresia (EA) may occur in several combinations. The overall incidence is 1 in 3000 live births. Maternal polyhydramnios is present, and premature birth is common. EA may also be associated with duodenal atresia.

The most common form (approximately 90 % of cases) is EA with a fistula between the trachea and the distal segment of the esophagus (Fig. 13.1, Type 1). This condition might be detected when the neonate chokes at the first feeding, but ideally it should be diagnosed antenatally by ultrasound or at birth by the inability to pass a soft rubber catheter into the stomach. Plain radiography confirms the diagnosis, showing the catheter curled in the upper esophageal pouch and an air bubble in the stomach, indicating a fistula. Contrast medium should not be used because it may be aspirated and further damage the lungs.

EA without fistula is the second most common form of the disease (Fig. 13.1, Type 2); there may be a large gap between the upper and lower segments of the esophagus. In such children, it is not possible to pass a catheter into the stomach, and there is no gastric air bubble. Pulmonary aspiration from the upper pouch is an immediate danger. Constant suctioning of the upper pouch should be instituted pending surgical repair.

The third most common form is the H-type fistula without atresia (Fig. 13.1, Type 3); diagnosis may be more difficult and is often delayed. In such cases, there is usually a history of repeated respiratory infections. The fistula may be difficult to locate even when contrast studies and endoscopy are used. Once the fistula is identified, surgical ligation is usually performed via a neck dissection.

There are other, rarer anatomic variants of this disease; many of which include tracheal stenosis.

Associated Conditions

1. Prematurity (30–40 %)
2. Congenital heart disease (22 %)

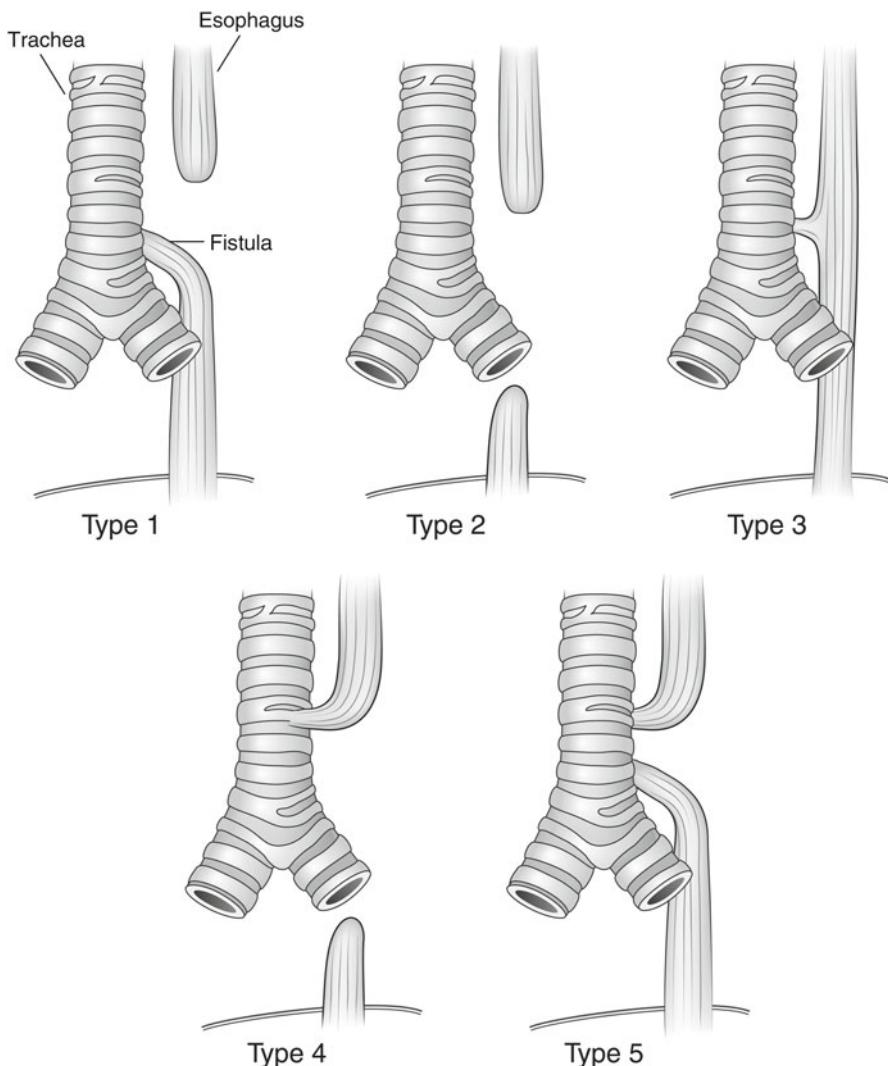


Fig. 13.1 Esophageal atresia and tracheoesophageal fistula (see text for details)

3. Additional gastrointestinal abnormalities (e.g., pyloric stenosis)
4. Renal and genitourinary abnormalities
5. The VATER and VACTERL associations: VATER association (Vertebral defects, Anal atresia, Tracheoesophageal fistula, *Esophageal atresia*, Radial and Renal dysplasia); VACTERL is VATER with Cardiac and Limb defects
6. Tracheomalacia and other abnormalities of the trachea (e.g., subglottic stenosis)

Surgical Procedures

The infant's general condition and the anatomy of the defect govern the choice of surgical management:

1. Primary complete repair (ligation of fistula and esophageal anastomosis) is preferred
2. Staged repair (gastrostomy followed by division of the fistula, followed later by repair of the esophagus) is less optimal but occasionally necessary

The current surgical trend is to perform an early primary repair with prior bronchoscopy to define the site of the fistula and exclude other tracheal defects.

Special Anesthesia Problems

1. Prematurity and other associated diseases (e.g., congenital heart disease) may complicate the case.
2. Pulmonary complications secondary to aspiration may be present.
3. There is a possibility of intubating the fistula.
4. Anesthetic gases may inflate the stomach via the fistula.
5. Surgical retraction during repair may obstruct ventilation.
6. Subglottic or tracheal stenosis may be present.
7. Children with repaired TEF are particularly prone to develop tracheal compression between the aorta and esophagus during feeding. The onset of symptoms of dyspnea and "dying spells" during feeding (i.e., parents report unresponsive infant, pale and apneic) is usually seen between 2 and 4 months of age. This condition is caused when an abnormally soft trachea becomes compressed against the aorta by a dilated esophagus. Aortopexy usually relieves symptoms, but in some children insertion of an external stent to reinforce the trachea may be necessary.

Anesthesia Management: Primary Repair

1. Observe the usual special considerations for neonates.

Preoperative

1. The infant is nursed in a semi-upright position.
2. The proximal esophageal pouch is suctioned continuously to prevent aspiration of secretions.
3. Institute intensive respiratory care to reduce pulmonary complications. (Even so, the lung condition seldom improves until after ligation of the fistula; therefore, surgery should not be delayed in the hope that pulmonary status will markedly improve.)
4. Examination of the infant to detect other associated lesions should be completed (i.e., echocardiography to rule out a congenital heart defect).

5. Establish a reliable intravenous route and ensure that blood is available for transfusion.
6. Give maintenance fluids but bear in mind that dehydration is not a major problem; neonatal fluid requirements are low during the first 24 h, and fluid and electrolyte depletion does not occur with esophageal obstruction.
7. In the preterm infant with respiratory distress syndrome and poor lung compliance, there is great danger of massive distention of the stomach (or massive leak from a gastrostomy if present) and consequent failure to ventilate. Rupture of the stomach and pneumo-peritoneum may occur. The risk of massive gastric distention in infants is reduced since the introduction of surfactant therapy. It has been suggested that a gas leak through the fistula may be controlled by:
 - (a) A balloon catheter passed via a gastrostomy retrograde into the lower esophagus.
 - (b) A balloon catheter inserted into the fistula under bronchoscopic guidance.
 - (c) Most commonly by early simple ligation of the fistula in infants with respiratory distress syndrome who might require high airway pressures. The operation is brief and can be followed by esophageal reconstruction after the child's respiratory function has improved.
8. Beware of the possibility of subglottic stenosis; have small tube sizes available.
9. TEF may be repaired using VATS in which case the considerations of one-lung surgery must be observed (see previous discussion).

Perioperative

1. Suction the upper pouch.
2. Apply the usual monitors and prepare to monitor ventilation using a precordial stethoscope in the left midaxillary line.
3. One of two approaches may be taken to secure the airway:
 - (a) Induce anesthesia with sevoflurane or halothane in O₂, maintaining spontaneous ventilation. Perform laryngoscopy and spray the larynx with lidocaine (1%, maximum dose, 5 mg/kg). Return to the mask and continue sevoflurane anesthesia for 2 or 3 min.
 - (b) Alternately, and if a bronchoscopy is *not* planned, sedate with fentanyl (0.5–1 µg/kg) and midazolam (25 µg/kg) and spray the larynx with lidocaine.

Then:

- (a) Intubate the trachea with the bevel of the tube facing posteriorly (to avoid intubating the fistula).

- (b) Alternately, if a rigid bronchoscopy is planned, once the surgeon passes the bronchoscope, attach the anesthesia circuit to the side arm of the bronchoscope and continue with spontaneous or gently assisted ventilation. When the bronchoscopy is completed, insert a tracheal tube with the bevel facing posteriorly, or
 - (c) If a fiberoptic bronchoscopy is planned, insert the tracheal tube with the bevel posterior and attach a bronchoscopic adapter to accommodate the fiberoptic bronchoscope through the tracheal tube. Gently assist ventilation while the endoscopy proceeds.
4. Immediately after intubation advance into the right bronchus and then slowly withdraw until breath sound are heard over the left lung and bilaterally; check ventilation throughout the lung fields. *If ventilation is unsatisfactory*, remove the tube, give O₂, and repeat the above measures. This will place the tube with its tip just above the carina but beyond the fistula in most cases. In some children, the fistula lies at the level of the carina. In this situation, the tube should be rotated so that the bevel faces anteriorly (to prevent inflating the fistula). More complicated methods to position the tube have been described but are unnecessary in our experience. Always check ventilation again after positioning of the child is complete. It is preferable to use a tube without a "Murphy eye," to minimize the possibility of leaks via the fistula.
 5. Once the tube has been properly placed, perform tracheobronchial suction to remove any accumulated secretions.
 6. Maintain anesthesia with air, O₂, and sevoflurane or isoflurane with spontaneous or gently assisted ventilation. If the inhalational anesthetic is not tolerated, small doses of fentanyl should be substituted (up to 10–12 µg/kg).
 7. *If spontaneous ventilation is inadequate:* assist ventilation cautiously, while observing and auscultating over the stomach for inflation. If gastric inflation occurs, allow the child to breathe spontaneously (with gentle manual assistance) until the fistula is ligated.
 8. Monitor ventilation carefully during surgical manipulation: large airways may be kinked by retraction, especially as the fistula is manipulated.
 9. Once the fistula is ligated, give a muscle relaxant and control the ventilation in the usual manner.
 10. Reports suggest that VATS may be useful in the repair of TEF, the view on the video-screen allows very precise repair of the esophagus. In this case, it may not be necessary to advance the endotracheal tube into the bronchus and isolate one lung. As the surgeon insufflates CO₂ under 5 cm H₂O pressure, the "up" lung is compressed and the lesion visualized, in effect, obtaining one-lung anesthesia by increasing intrathoracic pressure. During this period, ventilation may become difficult and a high FIO₂ required. Avoiding the necessity to intubate the bronchus may be advantageous as persistent upper lobe atelectasis may occur after endobronchial intubation.

Postoperative

1. The child with a clear chest who is awake and moving vigorously may be extubated in the OR. Some surgeons, however, may prefer to keep the trachea intubated and a gastroesophageal tube in place for several days to avoid reintubation and damage to the tracheal repair.
2. If there are pulmonary complications or any doubts about the adequacy of ventilation, continue controlled ventilation.
3. The pharynx is suctioned with a soft catheter that has a suitable maximum length of insertion clearly marked; it must not reach (and damage) the anastomotic site.
4. Prolonged intensive respiratory care may be required, swallowing is not normal postoperatively and aspiration may occur.
5. Prognosis after the repair depends on the maturity of the infant, whether other congenital anomalies are present, and whether pulmonary complications develop. In the absence of these conditions, the prognosis is excellent.
6. Postoperative analgesia may be provided by a caudal epidural catheter inserted intraoperatively and threaded to the thoracic level; careful management of local anesthetic doses is required (see Chap. 5).

Anesthesia Management-Staged Repair

If staged repair is planned, a preliminary gastrostomy is performed under local or general anesthesia. Management of the second stage (to repair the atresia) should follow the sequence outlined for primary repair, when the child's condition is optimal.

Late Complications

1. Diverticulum of the trachea at the site of the fistula is common. Be aware of this possibility and the danger of intubating the diverticulum during anesthesia in later life.
2. The tracheal cartilage structure is abnormal, and tracheomalacia may cause symptoms during infancy even after repair; episodes of stridor, dyspnea, and cyanosis ("dying spells") characteristically occur during feeding. This is caused by compression of the soft trachea between the dilated esophagus and the arch of the aorta. Severe symptoms require surgical treatment by aortopexy or tracheoplasty with an external splint. These children often have a deep barking cough much like children with croup.
3. Stricture may develop at the site of the esophageal anastomosis with episodes of esophageal obstruction with food (the classic "hotdog" in the esophagus); it may require repeated dilations and, later, possibly resection with replacement, using the colon or a gastric tube.

Congenital Laryngotracheoesophageal Cleft

This is a very rare anomaly in which there is a cleft in the posterior wall of the larynx that communicates with the esophagus. Four types are described, depending on the extension of this cleft distally. Type 1 is confined to the larynx, type 2 extends to the trachea, type 3 extends to the carina, and type 4 extends into the bronchi.

Mild type 1 forms may be missed initially and the child may later have a hoarse cry, cyanotic spells during feeding, and repeated episodes of aspiration pneumonia. Type 1 and type 2 clefts may be repaired endoscopically. These procedures are performed with the child breathing spontaneously and without a tracheal tube, thus ensuring good surgical access to the lesion.

Severe clefts manifest early in life with severe respiratory distress, which may be relieved by passage of a gastric tube. Pulmonary damage from aspiration may be severe. Early tracheostomy and gastrostomy may be lifesaving.

Anesthesia Management (For Type 1 and High Tracheal Lesions)

1. Careful preoperative assessment is performed to exclude other significant congenital lesions.
2. All the usual considerations for neonatal anesthesia are observed.
3. Inhalation induction of anesthesia using sevoflurane or halothane is performed.
4. A reliable IV route is established and administer atropine to limit secretions.
5. Topical anesthesia is thoroughly applied to the larynx and trachea (lidocaine 1%, 5 mg/kg maximum).
6. A suspension laryngoscope is positioned.
7. Anesthesia is maintained by insufflation of sevoflurane or halothane via a nasopharyngeal tube or alternatively by a propofol/remifentanil infusion.
8. An oxygen/air carrier gas should be used; if cauterity is planned, use the minimal FIO₂ to reduce the fire risk.
9. Postoperatively biphasic positive airway pressure (BIPAP) may be applied to ensure unobstructed respirations as required; a soft feeding tube is inserted into the stomach.

Note: This method of anesthesia (spontaneous ventilation, no tracheal tube, inhaled anesthetic, and meticulous topical anesthesia to the airway) is extremely useful for endoscopic surgery for the larynx and upper trachea. Scavenge gases around the head. We have used it successfully in many cases. However, obsessive attention is required—the child must be maintained at a constant level of anesthesia and not allowed to awaken or move. The topical anesthesia must be thoroughly applied and for long cases may need to be repeated. Halothane is a better anesthetic than sevoflurane for this purpose as awakening is slower if the concentration decreases for any reason—if you have it, use it. Otherwise, sevoflurane may be used—but maintain deep anesthesia. This is often difficult with

sevoflurane alone; supplementation with propofol either intermittently or by infusion is advised. Alternatively, a TIVA technique with a propofol/remifentanil infusion may be a better choice if halothane is unavailable.

Anesthesia Management (For Types 3 and 4 Clefts)

These defects require open surgery; definitive repair may involve a combined cervical and thoracic approach. Good intraoperative control of the airway is vital and demands close cooperation between the anesthesiologist and surgeon. A cuffed tube may be inserted into the esophagus to prevent ventilation of the stomach. In rare cases, cardiopulmonary bypass may be required for repair of lesions extending to the carina.

Congenital Hypertrophic Pyloric Stenosis

Congenital hypertrophic pyloric stenosis, a common surgical problem of infancy, occurs in up to 1 of every 300 live births. First-born male infants are more commonly affected. Hypertrophy of the muscle of the pyloric sphincter causes gastric outlet obstruction, leading to persistent vomiting. Dehydration, hypochloremia, and alkalosis develop. Today, the diagnosis is usually made promptly, resulting in normal electrolyte profiles in more than 50% of the infants. Diagnosis is made on the basis of the history, palpation of an olive-sized mass in the region of the pylorus, and confirmed by ultrasound.

Associated Condition

1. Jaundice (2% of children), which is caused by glucuronyl transferase deficiency: No special therapy is required, and the jaundice clears after pyloromyotomy.

Surgical Procedure

1. Pyloromyotomy—open or by laparoscopy (see Special Considerations)

Special Anesthesia Problems

1. Ensure that dehydration and electrolyte imbalance are fully corrected before surgery (pyloromyotomy is never a surgical emergency).
2. There is a danger of vomiting and aspiration.
3. Observe considerations for laparoscopic surgery (see page 331)

Anesthesia Management

1. Observe the usual anesthetic considerations for neonates.

Preoperative

1. Insert a gastric tube and apply continuous suction.
2. Rehydrate the child, correcting the electrolyte imbalance; this may require 24–48 h.
 - (a) Give 2:1 dextrose-saline solution and/or normal saline as indicated by serum electrolyte values; children with greater fluid and sodium deficits may require normal saline. Add potassium chloride (KCl supplements, 3 mEq/kg/day) when urine flow is established.
 - (b) Delay surgery until the infant appears clinically well hydrated and has normal electrolyte levels, acid–base balance, and good urine output.
3. Immediately before surgery, reassess the child to ensure that the fluid status and electrolytes are satisfactory:
 - (a) Check for clinical signs of adequate hydration (alertness, skin turgor, normal anterior fontanel, normal vital signs, moist tongue, urine output).
 - (b) Check biochemistry: chloride > 90 mmol/L; and bicarbonate < 28 mmol/L.

Perioperative

1. Give atropine intravenously.
2. Insert a vented orogastric suction catheter and suction the stomach in the supine, right and left lateral decubitus positions (even if the stomach has been on continuous gastric suction).
3. Give 100% O₂ by mask.
4. Use an RSI.
5. Maintain anesthesia with N₂O and a potent inhaled anesthetic or remifentanil.
6. Give a muscle relaxant (or propofol) and control ventilation. The choice of relaxant is dictated by the probable duration of surgery (i.e., the speed of the surgeon). Succinylcholine (2 mg/kg) and low-dose rocuronium (0.3 mg/kg) or cisatracurium (0.1 mg/kg) may also be used for brief procedures. No relaxant may be needed in some cases.
7. The surgeon may request that a small amount of air be insufflated into the stomach to check the integrity of the mucosa.
8. Acetaminophen (30–40 mg/kg PR or 7.5 mg/kg iv) may be administered at induction of anesthesia. Surgeons should infiltrate the wound with a long-acting local anesthetic to limit the perioperative pain. Administering opioids during surgery, especially during laparoscopic pyloromyotomy, will markedly delay emergence.
9. Extubate the airway when the infant is wide awake and in a lateral position.

Postoperative

1. If analgesia is insufficient, administer small doses of IV morphine (0.02–0.03 mg/kg).
2. Maintain intravenous infusion of fluids until oral intake is adequate (usually 24 h). Hypoglycemia may occur if glucose containing intravenous fluids is discontinued before oral intake is adequate.
3. Postoperative respiratory depression may occur as a result of the effect of preoperative alkalosis on the pH of the cerebrospinal fluid. Apnea has been reported in full-term infants after pyloromyotomy. The frequency of apnea and its causes are unclear, but infants should be carefully monitored (apnea monitor and/or pulse oximetry) for 24 h postoperatively.

Omphalocele and Gastroschisis

In omphalocele and gastroschisis, abdominal contents are herniated through the anterior abdominal wall. The incidence of these conditions varies regionally; 0.4–3 in 10,000 live births for gastroschisis and 1.5–3 in 10,000 for omphalocele.

With gastroschisis, the defect is lateral to the umbilicus (usually on the right side), the umbilical cord is situated normally, other congenital defects are rare, but the bowel is exposed to damaging effects of amniotic fluid in utero. Left-sided gastroschisis has a greater incidence of associated extra-intestinal defects. In omphalocele, the umbilical cord is continuous with the apex of the sac, which is covered by peritoneum and amnion, and associated congenital defects are common (75%). Preterm delivery is associated with a greater incidence of adverse outcomes.

The surgical objective is to close the abdominal defect without exposing the contained viscera to excessive pressure.

Associated Conditions (Omphalocele)

1. Prematurity: 30 %
2. Other gastrointestinal malformations (i.e., malrotation, diaphragmatic hernia): 25 %
3. Genitourinary anomalies: 25 %
4. Congenital heart disease: 10 %
5. Beckwith–Wiedemann syndrome (omphalocele, macroglossia, hyperviscosity, and severe hypoglycemia). These infants are usually large and have visceromegaly of the liver, kidney, and pancreas.

Surgical Procedures

The size of the abdomen in relation to the lesion determines the surgical procedure.

1. Primary closure is preferred because there is less risk of infection and other gastrointestinal complications. However, primary closure usually increases IAP, which, if excessive, may lead to impaired ventilation, reduced cardiac output, hypotension, and splanchnic ischemia, with impaired hepatic, renal, and bowel function. It has been recommended that the intragastric or bladder pressure be used as a guide to the safety of primary closure: pressures greater than 20 mmHg are poorly tolerated.
2. Skin closure only may be indicated in some infants.
3. In others, the procedure is staged; this approach involves initial closure using a preformed silo and the defect progressively reduced over time without anesthesia.
4. Very large omphaloceles may simply be painted with silver sulfadiazine and allowed to epithelialize over a period of weeks or months with delayed surgical closure.

Special Anesthesia Problems

1. Heat and fluid loss from exposed viscera (greater with gastroschisis).
2. Severe fluid and electrolyte disturbance and hypovolemic shock from transudation of fluid into the bowel and from exposure of viscera to air (hypoproteinemia may occur and is more likely with gastroschisis).
3. Increased IAP after closure may lead to compromised ventilation, decreased cardiac output, renal failure, and impaired hepatic function (including delayed clearance of drugs, notably fentanyl).
4. Possible hypoglycemia in infants with visceromegaly.

Anesthesia Management

1. Observe the usual special considerations for neonates.

Preoperative

1. The child is nursed in a semi-upright position with exposed viscera wrapped in sterile plastic film and covered by towels; this is done to prevent infection and minimize heat and fluid losses.
2. A gastric tube is usually inserted to decompress the bowel.
3. Monitor the blood glucose frequently; if the child is hypoglycemic (less than 40 mg/dl), infuse glucose continuously (6–8 mg/kg/min).

N.B. Children with Beckwith–Wiedemann syndrome have an increased insulin response; severe rebound hypoglycemia may occur if bolus doses of glucose are given.

4. Correct hypovolemia, electrolyte, and oncotic status. Initial fluid requirements are high (up to 140 mL/kg/24 h); normal saline plus colloid (albumin) is required to correct hypovolemia.
5. Order blood for possible transfusion.

Perioperative

1. Ensure that the OR is warmed to at least 24 °C and that heating lamps and forced-air warming devices are available.
2. Aspirate the gastric tube.
3. Give 100% O₂ by mask.
4. Perform a rapid-sequence induction with cricoid pressure. Note: children with Beckwith–Wiedemann syndrome may have a large trachea and require a larger or cuffed tracheal tube; their large tongue may interfere with normal laryngoscopy. In these children, a sedated awake intubation may be indicated. Have a GlideScope immediately available.
5. Maintain anesthesia with sevoflurane or halothane in O₂ and air.
 - (a) N₂O is contraindicated because it distends the bowel.
 - (b) Opioids (e.g., fentanyl) may not be metabolized as rapidly as expected because of the effect of increased IAP postoperatively.
6. Muscle relaxants may be required to facilitate ventilation.
7. Intraoperative pressure should be measured during abdominal closure to determine whether a primary closure will be tolerated. If the intraoperative pressure exceeds 20 mmHg, hemodynamic instability, and renal insufficiency may follow. As a simple adjunct, it is suggested that monitoring SaO₂ at post-ductal sites above and below the abdomen (i.e., left hand and foot) may predict increased IAP. If the reading from the left foot decreases significantly below the left hand, or if the quality of the waveform is markedly less in the foot compared with the hand, then IAP may be excessive.

Postoperative

1. Assess the adequacy of spontaneous respirations; if in doubt transfer to the NICU intubated for possible long-term airway management.
2. Continue nasogastric suction until bowel function recovers.
3. Continue IV fluids and glucose; fluid requirements remain high in the early postoperative phase. Some infants (i.e., gastroschisis) require intravenous alimentation because bowel function may be impaired for weeks and possibly months.

Biliary Atresia

Severe jaundice in the neonate most commonly results from neonatal hepatitis or biliary atresia. Atresia of the extrahepatic bile ducts may be a result of an intrauterine inflammatory process. Biliary obstruction leads to hepatocellular damage and ultimate cirrhosis. The incidence of biliary atresia is 0.5–1 in 10,000 live births. Other causes of persistent direct (mixed) hyperbilirubinemia must be ruled out and the diagnosis of bile duct atresia confirmed. This is usually achieved by percutaneous liver biopsy, scintigraphy, and endoscopic retrograde cholangiography.

Associated Anomalies

1. Polysplenia syndrome; intestinal malrotation, preduodenal portal vein, situs inversus, absent IVC, cardiac defects, and anomalous hepatic artery supply.

Surgical Procedure

1. Hepatic portoenterostomy (Kasai procedure). The extrahepatic bile ducts are resected, and the porta hepatis is dissected to a depth of 2–3 mm; then a Roux-en-Y jejunal anastomosis is performed. The major preoperative survival determinant seems to be the age at which the operation is performed. If the infant is older than 12 weeks of age, success is less likely. These older infants are often listed for liver transplantation.

Anesthesia Problems

1. Hepatic function is impaired, especially in the older child.
2. Hypoprothrombinemia develops and leads to impaired coagulation, especially in older infants.
3. Blood loss may be extensive, but this is unusual.
4. Intraoperative radiographs may be needed in some children.
5. The procedure may be performed using robotic surgery via laparoscope. (See previous Special Considerations)
6. Some children will have associated thrombocytopenia because of hypersplenism.
7. Re-operation may be required if bile drainage is inadequate.

Anesthesia Management

Preoperative

1. Observe the usual anesthetic considerations for neonates.
2. Check the coagulation profile and verify that the child has received vitamin K₁.

3. Check that adequate blood, fresh-frozen plasma, and platelets are available.

Perioperative

1. Administer 100 % O₂ by mask and place monitors.
2. Induce anesthesia intravenously or by inhalation of sevoflurane or halothane.
3. Administer a muscle relaxant and intubate; *Cis*-atracurium is preferred because of possible impaired hepatic function.
4. Maintain anesthesia with low concentrations of sevoflurane or isoflurane in an O₂/air mixture. Fentanyl or remifentanil may be used for analgesia.
5. Supplement relaxant drugs as necessary and control ventilation.
6. Insert large-bore intravenous lines into upper limbs or neck. Insert a gastric tube.
7. Be alert to the possibility of sudden hypotension because of IVC obstruction during surgical manipulation of the liver. Placing the infant in a slight head-down position may minimize falls in BP during manipulations of the liver.
8. Infuse glucose-containing solutions at maintenance rates and monitor blood glucose levels.
9. Monitor the temperature; use an overhead heater and a forced air heater (or both) in addition to all other measures to maintain normothermia.
10. Consider a continuous caudal or epidural catheter for postoperative analgesia provided the child is not coagulopathic.

Postoperative

1. Prolonged IV alimentation may be required.
2. Ascending cholangitis is common, and portal hypertension may develop.
3. Many children develop esophageal varices in later years with repeated bleeding episodes.
4. Postoperative ventilation for 24–48 h may be needed.
5. If the trachea is intubated postoperatively, titrate fentanyl or morphine to provide analgesia. In extubated children, a caudal catheter will provide satisfactory analgesia; recall that reduced total doses of long-acting local anesthetics are required in this age group.

Intestinal Obstruction in the Neonate

Intestinal obstruction in the neonate may result from various lesions (e.g., duodenal atresia, duplication, midgut volvulus, malrotation) or from accumulation of viscid meconium (“meconium ileus”). Laparoscopic rather than open procedures are common.

Associated Conditions

1. Prematurity.
2. Down syndrome and hence congenital heart disease (40–50%).
3. Cystic fibrosis (invariably accompanies meconium ileus).
4. Subglottic stenosis with duodenal atresia.

Special Anesthesia Problems

1. Hypovolemia; acid–base and electrolyte imbalance.
2. Gross abdominal distention in some cases.
3. Risk of regurgitation and aspiration.

Anesthesia Management

1. Observe special considerations for neonates.

Preoperative

1. Ensure adequate fluid volume replacement.
2. Check acid–base and electrolyte status and correct any imbalance as far as possible.
3. Ensure that the gastrointestinal tract has been decompressed as much as possible (via a gastric tube).
4. Ensure the availability of blood and blood products as indicated.

Perioperative

1. Give atropine intravenously.
2. Aspirate the stomach contents via a vented gastric tube in the supine, right and left lateral decubitus.
3. Give 100% O₂ by mask.
4. Intubate the trachea with the child awake or after an RSI. Beware of the possibility of subglottic stenosis.
5. Maintain anesthesia with isoflurane or sevoflurane in an air/O₂ mixture. Avoid N₂O because it may further distend the bowel.
6. Give small doses of relaxant drugs (cisatracurium is preferred) and control ventilation.
7. Despite apparently adequate preoperative fluid resuscitation, some infants become hypotensive upon opening the abdomen, especially those with small bowel atresia or midgut volvulus. They may need surprisingly large volumes of intravenous fluid (albumen and even blood or plasma) to restore the BP. Be prepared!
8. Some children will benefit with placement of a caudal catheter for postoperative pain management; recall that reduced total doses of long-acting local anesthetics are required in this age group.

Postoperative

1. Do not extubate the trachea until the infant is fully awake and vigorous.
Extubate the airway with the infant in the lateral position.
2. Prolonged intravenous or gastrostomy feeding may be required.
3. After meconium ileus, problems including prolonged bowel dysfunction, sepsis, and pneumonia must be anticipated.

Neonatal Necrotizing Enterocolitis

Neonatal necrotizing enterocolitis (NEC), a disease of low-birth-weight infants (usually less than 34 weeks gestation), is characterized by intestinal mucosal injury secondary to ischemia of the bowel. It often leads to perforation and peritonitis. Severe fluid and electrolyte disturbance, life-threatening hyperkalemia, endotoxic shock, renal failure, and coagulopathy, especially thrombocytopenia may develop. As the disease progresses, multisystem organ failure occurs. The cause of NEC is uncertain, but the disease usually affects infants with a history of birth asphyxia, respiratory distress syndrome, and shock. Other possible etiologic factors include early enteral feeding, liberal intravenous fluid administration, restrictive target oxygen therapy (SpO_2 85–89 % $>91\text{--}95\%$), infection, and umbilical artery catheterization. The administration of probiotics containing Lactobacillus or Bifidobacterium may reduce the incidence and severity of NEC.

The clinical picture is one of abdominal distention, bloody diarrhea, temperature instability, and lethargy; apnea may occur. Abdominal radiography may show intramural bowel or portal venous gas.

The medical management of NEC includes cessation of feeding and institution of continuous gastric suction, use of intravenous fluids and alimentation, IV antibiotics, and correction of anemia and/or coagulopathy. Steroids and inotropes may be used to treat shock. Bowel perforation is the usual indication for surgery.

Special Anesthesia Problems

1. Prematurity and respiratory distress.
2. Shock, hypovolemia, electrolyte disturbance, and coagulopathy.
3. Sepsis, acidosis, and congestive cardiac failure.
4. Interstitial gas in the bowel wall.
5. Interaction of antibiotics with relaxant drugs.

Anesthesia Management**Preoperative**

1. Restore blood and fluid volumes: blood, plasma, platelets, and crystalloid solutions may be required; third space losses are considerable. If volume replacement does not improve the BP, inotropes are indicated. Check Hct, blood gases, electrolytes, and the blood glucose level.
2. Check coagulation: thrombocytopenia must be corrected by platelet infusions.
3. Exchange transfusion may be required for severe sepsis. Correct the acid-base status.
4. Monitor carefully for apnea.
5. Prepare infusions of dopamine and epinephrine (on pumps and at appropriate concentrations for an infant) for possible use intraoperatively.
6. Prepare syringes of epinephrine for possible resuscitation or treatment of hypotension.
7. Prepare syringes of calcium gluconate or calcium chloride for anticipated hypocalcemia during rapid infusion of blood products.

Perioperative

1. Observe all special considerations for the preterm infant. Reliable intravenous routes and an arterial line are essential.
2. Use of fentanyl (10–12 µg/kg) and ventilation with appropriate concentrations of O₂ is the preferred method. (Avoid N₂O to prevent expanding intramural bowel gas.)
3. Administer cisatracurium as required.
4. Continue fluid resuscitation throughout surgery as dictated by clinical status and laboratory studies. Warm all intravenous fluids.
5. Anticipate that major fluid infusions may be required to maintain cardiovascular homeostasis when the abdomen is opened. Carefully monitor the BP and infuse adequate volumes of appropriate warmed crystalloid and colloid to correct hypotension. Up to one blood volume or more may be required to maintain vascular volume status due to massive third space losses.
6. Anticipate the possibility of massive rapid blood loss. Be prepared.
7. Anticipate the possible need for vasopressors (dopamine, epinephrine). Have them set up and in place ready for emergent use. Recall that very high rates of dopamine may be required (up to 20 µg/kg/min).
8. Anticipate possible severe hypocalcemia if rapid infusion of blood products (FFP or platelets) are needed.

Postoperative

1. Return the infant to the NICU for continued management of possible multi-system dysfunction (cardiac, pulmonary, renal, etc.).

OTHER MAJOR THORACOABDOMINAL LESIONS AND PROCEDURES

Neuroblastoma and Ganglioneuroma

Neuroblastoma is the most common solid malignant tumor of infancy. More than 50 % of such tumors appear in the retroperitoneal space, but they may occur anywhere along the sympathetic chain. Four stages of the disease are described: Stage 1, limited to the primary site; stage 2, extension from primary but not across the midline; stage 3, beyond the midline; and stage 4, beyond the midline with metastases. Metastases are present at diagnosis in more than 50 % of cases.

The diagnosis is sometimes based on fetal ultrasound. In neonates and young infants, the diagnosis may be made by palpation of an abdominal mass. The older child frequently has evidence of chronic disease, including fever, weight loss, and anemia. The tumor secretes catecholamines, which may cause hypertension, and a vasoactive intestinal peptide, which may cause watery diarrhea, dehydration, and electrolyte disturbance. The tumor may also cause symptoms as a result of its extension to involve other tissues; for example, the extradural space may be involved and neurologic signs may appear. The tumor may also wrap around or compress major vessels such as the vena cava or aorta. Most children have increased levels of catecholamines and metabolites in the urine (positive Vanillylmandelic acid). Computed tomography (CT) and magnetic resonance imaging (MRI) permit localization of the tumor and identification of metastases.

Treatment of stage 1 or 2 disease is by surgical excision. Stage 3 or 4 disease is treated by initial chemotherapy or radiotherapy, or both and may later be followed with surgical resection. Children with widespread disease may be treated by total body radiation, chemotherapy, and bone marrow transplantation.

Special Anesthesia Problems

1. Massive blood loss may occur during surgery, especially in those who had preoperative radiotherapy. Major vessels (e.g., IVC) may be invaded by tumor and may be a source of rapid blood loss and clot or tumor emboli.
2. Catecholamine levels are usually increased and hypertension may be present, but cardiac arrhythmias are unusual. Those very rare children with severe hypertension should be assumed to have a contracted blood volume and may benefit from preoperative preparation as outlined for pheochromocytoma.
3. Thoracic tumors may compress the lungs and produce respiratory failure. Cervical tumors may displace the trachea and compress the airway.
4. Children with extension to the extradural space (dumb-bell tumor) may need a combined abdominal and neurosurgical approach with laminectomy.
5. Hyperthermia (*not* MH) has been reported during surgery for neuroblastoma.

Anesthesia Management**Preoperative**

1. Carefully assess the systemic effects of the disease. Check the hemoglobin level and electrolytes. Check the results of CT or MRI for evidence of major blood vessel or neurologic involvement.
2. Assess the extent of hypertension and its control by preoperative therapy.
3. Administer appropriate sedation (e.g., midazolam 0.5 mg/kg PO or 0.05 mg/kg IV).
4. Important: Adequate supplies of blood for transfusion must be available, together with facilities to measure, warm, and replace large volumes of fluid. Be prepared. A rapid infusion device should be at hand.
5. Anticipate the need for vasopressors; set them up on a pump with a dedicated carrier available for immediate emergent use.
6. Anticipate possible severe hypocalcemia if rapid infusion of blood products (FFP or platelets) are needed.
7. Review the case with the surgeon so as to anticipate and plan appropriately.

Perioperative

1. Observe all special considerations for infants.
2. Place monitors and induce anesthesia.
3. Maintain anesthesia with inhalation agent, which can be titrated to control BP and avoid N₂O.
4. Rocuronium or vecuronium are preferred for relaxation; they provide cardiovascular stability and minimal histamine release.
5. Establish reliable large-bore intravenous routes (preferably rapid infusion catheters) in the upper limbs or neck.
6. Routine monitors should be supplemented with an arterial line and a urinary catheter. A double-lumen line central line provides routes for infusions and monitoring of central venous pressure (CVP).
7. Fluctuations in BP may occur during surgical manipulation of the tumor. These can usually be treated by adjusting the inspired concentration of inhalation agent. Rarely, it may be necessary to use small doses of phenotolamine (0.2 mg/kg) or an infusion of sodium nitroprusside.
8. Be prepared to provide rapid infusions of warmed blood. A decrease in BP is common as the tumor is removed. This usually responds to fluid infusions. Be aware that hypotension might also result from-surgical retraction compromising IVC flow.
9. If major blood transfusion becomes necessary, check coagulation indices and prepare to correct deficiencies.
10. Anticipate possible ionized hypocalcemia due to the rapid infusion of blood products (FFP or platelets).
11. Some children will benefit with placement of a caudal or epidural catheter for postoperative pain management.

Postoperative

1. In most children, the trachea can be extubated at the end of surgery; however, if the child is unstable, do not extubate.
2. Plan for optimal postoperative pain management; caudal or epidural analgesia is ideal for many children; recall that reduced total doses of long-acting local anesthetics are required in infants <6 months of age.
3. Continue to monitor volume status and urine output to ensure adequate fluid replacement.

N.B. Ganglioneuromas are benign tumors arising from sympathetic ganglia. They do not invade other tissues but gradually enlarge and may produce symptoms because of their size and pressure on adjacent structures. They do not generally secrete significant amounts of catecholamines or other active peptides; however, in atypical forms this may occur. In such a case, though preoperative catecholamine levels are normal, intraoperative hypertension may occur when the tumor is handled. Surgical excision is usually simpler than for neuroblastomas, but it may be complicated by the tumor's location and size. Many may be treated by endoscopic resection.

Lung Surgery

Lung surgery may be indicated for the following conditions:

1. Lung abscess
2. Bronchiectasis
3. Lung cysts
4. Bronchogenic cysts
5. Diagnostic biopsy (in children, usually to confirm or exclude infection by a virus or other pathogen)
6. Pulmonary arteriovenous malformation
7. Sequestered pulmonary lobe
8. Pulmonary neoplasm
9. Chronic pulmonary infection

Special Anesthesia Problems

1. For open thorax surgeries, major \dot{V}/\dot{Q} inequalities should be anticipated; increase the FIO_2 and monitor SaO_2 . Blood and secretions within the bronchial tree may compound this problem; if so, suction the tracheal tube frequently.
2. Major hemorrhage may occur suddenly if large vessels are accidentally cut or torn. Therefore, reliable large-bore infusion routes must be established and blood for transfusion must be immediately available in the OR. Have a rapid transfusion device nearby.

3. Postoperative pain limits coughing and deep breathing, especially in older children, predisposing to atelectasis. Plan for optimal analgesia (e.g., thoracic epidural catheter).
4. Pulmonary function may be seriously impaired, resulting in respiratory failure in some children (e.g., those admitted for lung biopsy). In such children, even minor fluid overload can precipitate serious deterioration in lung function postoperatively; be extremely cautious with fluid therapy.
5. Pus from purulent lesions (e.g., lung abscess, bronchiectasis) may become dispersed during surgery unless a lobe or lung can be isolated. Preoperative bronchoscopy to remove accumulated secretions is useful for some children.
6. Pulmonary arteriovenous malformation results in a very large right-to-left shunt, with consequent arterial desaturation that cannot be corrected by increasing the FIO₂.
7. Methods for VATS and OLV are described at the beginning of this chapter.

A further alternative in the case of unilateral purulent disease, thoracotomy may be performed with the child in the prone (Parry Brown) position with the head slightly dependent. In this position, purulent secretions tend to drain out via the tracheal tube and do not contaminate the dependent lung.

Anesthesia Management

Preoperative

1. Assess the child carefully.
 - (a) Evaluate pulmonary function as fully as possible (only limited data may be obtainable for children too young or otherwise unable to cooperate in the full range of tests).
 - (b) Check the results of blood gas and other available studies. Ensure that the child is in the best possible condition for the planned surgery.
2. Ensure that adequate supplies of blood products are ordered and adequate serum has been saved for additional cross-matching.
3. Ensure that appropriate respiratory care is ordered. For older children, explain the value of preoperative breathing exercises and the need for post-operative respiratory care.
4. Order appropriate preoperative sedation, taking care to prevent respiratory depression in children with impaired pulmonary function. Midazolam (0.5 mg/kg PO or 0.05 mg/kg IV) is often ideal.

Perioperative

1. Apply monitors.
2. Give 100% O₂ by mask.

3. Induce anesthesia by inhalation of either sevoflurane or halothane; if an IV is in place then propofol or thiopental followed by an intermediate or long-acting muscle relaxant to facilitate tracheal intubation.
4. Ventilate with 100 % O₂ until the child is fully relaxed, then perform a bronchoscopy if indicated. Otherwise, intubate the trachea as required for the surgery and the child's size. If lung isolation is required, use one of the techniques described (see page 333).
5. Establish a reliable wide-bore intravenous route.
6. Insert an arterial line (except for "minor" procedures in healthy children).
7. Maintain anesthesia with sevoflurane or isoflurane in oxygen/air with opioid, adding sufficient O₂ to maintain the saturation at 95–100 %. Nitrous oxide may be used if a gas-filled cavity is not present.
8. Control the ventilation (aim for a PaCO₂ of 35–40 mmHg).
9. After the child has been positioned for thoracotomy, recheck the ventilation of the lungs.
10. Suction the tracheal tube as needed.
11. If one-lung anesthesia is used, administer sufficient oxygen to maintain SaO₂ greater than 95 %.
12. Periodically inflate the lungs fully during thoracotomy and as the chest is being closed, give several large breaths with high peak pressures while observing reexpansion of the lung. The surgeon may also fill the chest with saline and ask for large breaths to check for possible air leaks.
13. When the chest is closed, ensure that chest drains are appropriately set.
14. At the end of surgery, before extubation, ensure that the child is awake and responding and has adequate spontaneous respirations. If in doubt, leave the tracheal tube in place and continue assisted ventilation until it is safe to withdraw this support.

Postoperative

1. Provide analgesia: A thoracic epidural block using bupivacaine or ropivacaine with fentanyl is ideal and may be left in place for up to 72 h postoperatively. Otherwise, intercostal nerve blocks plus an opioid infusion or patient-controlled analgesia may be used.
2. Ensure that chest drains are patent and are connected to an underwater seal and suction.
3. Order arterial blood gas determinations as necessary to assess the adequacy of ventilation.
4. Ensure that chest radiography is performed, and check the films for residual pneumothorax or atelectasis.
5. Order hemoglobin (Hb) and hematocrit (Hct) determinations to assess the adequacy of blood replacement.

Anterior Mediastinal Masses

Anterior mediastinal masses (AMM) (bordered by the retro-sternum, pericardium, xiphoid, and angle of Louis) may press on middle mediastinal structures (trachea/bronchi or heart (pulmonary artery, right atrium)) resulting in impaired or life-threatening loss of function. Four tumors that are common in the anterior mediastinum are:

1. Lymphoma, Hodgkin disease, etc.
2. Teratoma
3. Thyroid
4. Thymoma (less common)

Tumors also arise in the middle and posterior mediastinum (e.g., neuroblastoma, ganglioneuroma, and dermoid cysts) although they do not usually present substantive anesthetic challenges.

Children with AMM may need general anesthesia for cervical node biopsy, indwelling ports, radiologic investigations, or excision of the tumor. Children with significant airway compression may be best managed for tumor biopsy using local anesthesia and mild sedation.

Special Anesthesia Problems

1. Acute airway obstruction may occur during anesthesia, even in children with no history of dyspnea. Tracheal intubation may fail to relieve the obstruction, advancing the tube into a bronchus may be required to establish an airway and maintain ventilation. This is especially likely in children with massive enlargement of hilar nodes or a direct tumor effect causing tracheomalacia or extramural tracheobronchial compression. Even during simple, minor procedures, general anesthesia may be extremely dangerous.
2. The AMM mass may compress the heart (right atrium and pulmonary artery), compromise ventricular filling, and cause acute hypotension and sudden disappearance of the capnogram.
3. Major blood loss may occur during mediastinal surgery.
4. Myasthenic children require special consideration.

Anesthesia Management

Preoperative

1. Assess the child carefully, and anticipate potential airway and cardiovascular problems. A night cough and several pillow orthopnea are classic for upper airway compromise. The latter is a sign of potential disaster. Inquire about any postural dyspnea and which position gives the easiest breathing.

Look for signs of superior vena cava obstruction or upper body edema. In some cases, it is best to delay surgery until the size of the tumor can be reduced by a 12-h exposure to steroids or radiation. Discuss this with the surgeon and oncologist.

2. Examine radiologic studies and pulmonary function tests:
 - (a) Chest X-ray. Use the posteroanterior (PA) view to diagnose a compressed, deviated, or scabbard trachea (suggesting tracheomalacia).
 - (b) CT scan to define the extent of airway compromise; if the area of the airway is decreased by more than 50 %, extreme caution is recommended (i.e., consider preoperative steroids or operation under local anesthesia). Evidence of main stem bronchus compression is ominous. Examine the potential for heart and great vessel compression by the tumor mass.
 - (c) Echocardiogram to assess the heart for structural (compression of the right atrium or pulmonary artery) and pericardial (pericardial effusion or constrictive pericarditis) involvement.
 - (d) Flow-volume loop although most young children cannot cooperate well enough; if expiratory flow rates are decreased by 50 % or more, extreme caution is recommended (see previous discussion).
3. For mediastinal surgery, ensure that adequate blood is immediately available, that additional units can be secured, and that a rapid infusion device is nearby.
4. If there is any danger of airway obstruction, do not premedicate.
5. If airway problems are anticipated, prepare a full range of endotracheal tubes, stylet, and laryngoscope blades and have a skilled surgeon with a rigid bronchoscope present during induction. An armored tube on a rigid stylet may be required to secure the partially compressed airway.

Perioperative

N.B. Cervical lymph node biopsy in the child) with a very large AMM that prevents the child from lying flat requires special consideration. Preoperative steroids or radiation for 12 h should be considered. Lymph node biopsy may be accomplished with the child in the sitting position using local anesthetic. If that is not possible or if another surgery is planned, general anesthesia may be induced using inhalational anesthesia with the child in the left lateral decubitus position.

For general anesthesia:

1. Establish IV access using local anesthetic or nitrous oxide analgesia.
2. Check that the awake child can tolerate the position chosen for surgery. Anesthesia should be induced by inhalation while maintaining spontaneous respirations.

3. Breath sounds, the shape of the capnogram, and SaO_2 should be monitored continuously, especially after any changes in the child's position. If the airway obstructs or the capnogram loses its trace, before tracheal intubation:
 - (a) Intubate the trachea and turn the child to the left lateral decubitus position. If vital signs are not restored, turn the child prone. In this position, gravity pulls the tumor off the mediastinal structures. If anesthesia was induced supine and vital signs are lost, an alternative to turning the child is to have the surgeon apply towel clips to the xiphoid and suprasternal notch and lift the sternum to release the pressure) on the mediastinal structures.
 - (b) Advance the tube with a rigid stylet past the obstruction-endobronchial if necessary.
 - (c) If this fails, perform rigid bronchoscopy.
 - (d) In extreme cases, emergency thoracotomy may be required.
4. Maintain anesthesia with sevoflurane or isoflurane in 100 % O_2 ; spontaneous ventilation is preferable.
5. Monitor the cardiac rhythm during surgical dissection.
6. Ask the surgeon to place intercostal nerve blocks at appropriate locations
7. At the completion of surgery, check adequacy of respirations before extubation.
8. If respirations are inadequate, leave the trachea intubated and transfer to the PICU.

Postoperative

1. If applicable, ensure that the nurses in the PACU are aware that airway obstruction or cardiac compromise might occur in certain positions.
2. Order maintenance fluids and appropriate analgesia.
3. If blood replacement was required, check the Hct.
4. Obtain a chest radiograph to check for pneumothorax.
5. Admit to the PICU for more extensive observation and monitoring if there is any question.

Myasthenia Gravis

Myasthenia presents in three forms in infants and children:

1. *Transient neonatal myasthenia* in neonates of myasthenic mothers develop hypotonia and feeding difficulty within a few hours of birth. Improvement occurs within a few weeks although anticholinesterases are needed in the interim period.

2. *Juvenile myasthenia gravis* (autoimmune myasthenia) usually manifests in childhood or adolescence. The disease may be generalized or limited to the ocular muscles. Abnormal fatigability, limb weakness, and ptosis are the usual presenting clinical features. The diagnosis is confirmed by a decreased compound muscle action potential on repetitive nerve stimulation, or by improved muscle power after an intravenous anticholinesterase drug (edrophonium).
3. *Congenital myasthenic syndromes* due to various defects of neuromuscular transmission may cause symptoms during infancy or childhood and are difficult to distinguish from autoimmune myasthenia without complex testing.

Associated Condition

1. Hyperthyroidism maybe present.

Treatment

1. Anticholinesterase therapy produces symptomatic improvement, but secretions may become a problem; pyridostigmine is the drug of choice.
2. Plasmapheresis or intravenous immunoglobulin results in temporary improvement in many children and may reduce the need for surgery.
3. Prednisone and azathioprine have been effective in some children.
4. Thymectomy may increase the probability of remission, especially if it is performed early after the symptoms appear.

Surgical Procedure

Thymectomy is performed in some children with severe generalized myasthenia gravis who fail to respond to other treatment, even if there is no thymoma. The best remission occurs in young children with thymic hyperplasia. Complete thymectomy is considered the surgical management of choice and can be accomplished via minimally invasive surgery. This is preferred for the myasthenic child because it significantly decreases postoperative problems with incisional pain. Myasthenic children undergoing other procedures should be managed similarly but with the goal of regional blocks to provide postoperative analgesia.

Special Anesthesia Problems

1. Muscle weakness may lead to ventilatory failure.
2. Treatment with anticholinesterases increases respiratory tract secretions.
3. Potential sudden deterioration in muscle power may be caused by:
 - (a) A myasthenic crisis or
 - (b) A cholinergic crisis (sweating, salivation, bronchial secretions, miosis, muscle paralysis with respiratory failure) induced by excessive dosage with anticholinesterase

4. Postoperative pain may limit ventilation. Regional analgesia for postoperative pain is ideal (e.g., epidural analgesia).
5. Chest physiotherapy postoperatively rapidly fatigues the child if it is too vigorous.
6. Extreme sensitivity to non-depolarizing relaxants may occur. Inhalational anesthesia alone has proved very successful.

Anesthesia Management

Preoperative

1. The child should be admitted to the hospital for a period of rest, and anticholinesterase drugs should be reduced or withdrawn.
2. Ensure that blood is available.
3. Do not give heavy premedication—avoid opioids.
4. In children with severe symptoms preoperative plasmapheresis may be useful.
5. Consult with pediatric neurology.

Perioperative

1. Induce anesthesia by inhalation of sevoflurane or halothane or with IV thiopental (5–6 mg/kg IV) or propofol (4–5 mg/kg IV).
2. Deepen anesthesia with sevoflurane or halothane, or with TIVA (propofol and remifentanil)
3. Do not give any muscle relaxants; intubate the trachea when the child is adequately anesthetized and after applying topical analgesic to the larynx. Recall that these patients are very sensitive to the effects of non-depolarizing muscle relaxants but resistant to the relaxant effects of succinylcholine and its effects will be prolonged due to the anticholinesterase therapy. For VATS surgery, a double lumen tube or blocker is required (see previous discussion).
4. Control ventilation. Continue with TIVA or sevoflurane anesthesia.
5. Ensure reliable intravenous infusion routes for possible transfusions.
6. If a thoracotomy is planned, a thoracic epidural will provide postoperative analgesia and reduce inhalation agent requirements intraoperatively.
7. At the completion of surgery, allow the child to recover and resume spontaneous respirations with the tracheal tube in place. Check the vital capacity; if this is adequate (more than 20 mL/kg), proceed cautiously with extubation.

Postoperative

1. Close observation and respiratory care in the PICU is essential.
2. Reduce or discontinue anticholinesterase therapy (to lessen the likelihood of a cholinergic crisis).

3. Edrophonium (Tensilon test, initial dose 0.2 mg/kg or 1–2 mg IV at 1 min intervals up to 10 mg) testing may be performed periodically as a guide to anticholinesterase therapy (i.e., administer a small dose of edrophonium to confirm that this produces an improvement in muscle power). If there is no improvement, an impending cholinergic crisis is likely.
4. Use caution with opioids—regional nerve block (e.g., continuous epidural) is preferable.
5. Plan physiotherapy carefully to avoid overtiring the child. (Time the physiotherapy to take advantage of the increased muscle power after each edrophonium test.)
6. If fatigue and/or serious retention of secretions occur, intubate the trachea and control ventilation.

Splenectomy

See Idiopathic Thrombocytopenic Purpura (page 561).

Pheochromocytoma

Pheochromocytoma is rare in children (~5 % of all cases); when it appears, it is usually in the adrenal medulla and bilateral in 20 %. Most are benign; only 5–10 % are malignant. About 50 % are inherited as autosomal dominant (e.g., neurofibromatosis 1 and multiple endocrine neoplasia); the remainder are spontaneous. The common symptoms include headache, nausea, and vomiting, with sustained or, less commonly, episodic hypertension. Abdominal pain may occur. If the diagnosis is confused, this might prompt an unnecessary and dangerous exploratory operation in an unprepared child. Currently, the diagnosis is confirmed by measuring free plasma metanephrine and normetanephrine concentrations; rather than urinary vanillylmandelic acid. The secretion from the tumor is predominantly norepinephrine; sustained hypertension with vasoconstriction contracts the intravascular volume and increases hematocrit. Paragangioneuromas may mimic pheochromocytoma since they also secrete vasoactive peptides. Recent advances include a laparoscopic rather than open approach and to leave some viable adrenal cortex if the tumors are bilateral in order to preserve adrenal function; however, ~20 % will have tumor recurrence.

Associated Conditions

1. Neurofibromatosis 1
2. Thyroid tumor
3. Multiple endocrine adenomatoses (e.g., Sipple syndrome)

Special Anesthesia Problems

Note: Anesthesia is very dangerous in the unprepared child—extreme swings in BP may occur.

1. Management of the BP and volume status of the child can be difficult.
2. Major blood loss may occur from extensive surgery performed to locate and remove multiple tumors.
3. Avoid anesthetics that might increase the release of catecholamines (e.g., succinylcholine, pancuronium) or sensitize the heart to these substances (e.g., halothane). Pancuronium or droperidol may cause a hypertensive crisis. Drugs that release histamine (e.g., morphine, atracurium) should be avoided.
4. Good preoperative sedation and a smooth induction are essential to prevent release of catecholamines.

N.B. Dangerous cardiac arrhythmias are extremely rare in children.

Anesthesia Management

General anesthesia may be required for special investigations to locate the tumor, and for its extirpation, and must be conducted with the same considerations.

Preoperative

1. The child should be treated with α -blocking drugs (e.g., phenoxybenzamine HCl, 0.25–1.0 mg/kg/day) for several days, until:
 - (a) BP is consistently normal or there are signs or symptoms of mild postural hypotension.
 - (b) Hematocrit has decreased (indicating expansion of intravascular volume). It is not necessary to administer a β -blocker to children; β -blockade may be contraindicated when incomplete α -blockade is present (most of the time in children) as the latter may lead to cardiac failure. However, a brief infusion of a short-acting β blocker such as esmolol may rarely be appropriate in some older patients (500 μ g/kg over 2 min then an infusion of 50–250 μ g/min to treat tachycardia).
2. Ensure that adequate supplies of blood are available.
3. Check that drugs are at hand to treat any blood pressure disturbance or cardiac rhythm, including
 - (a) Phentolamine—to decrease BP (usually necessary)
 - (b) Isoproterenol—to increase heart rate
 - (c) Norepinephrine or phenylephrine—to increase BP
 - (d) Consider magnesium sulphate boluses (25–50 mg/kg over 20 min) to control hypertensive events
 - (e) Consider a remifentanil-based anesthetic to control heart rate and BP

4. Give adequate premedication to reduce anxiety (oral midazolam 1 mg/kg up to 20 mg or IV 0.05–0.1 mg/kg)

Perioperative

1. If the child still appears apprehensive after premedication, give supplemental midazolam (0.05 mg/kg IV) until the desired level of sedation is achieved.
2. Apply standard monitors.
3. Induce anesthesia with titrated doses of either propofol or thiopental.
4. Give rocuronium (0.6–1.0 mg/kg) or vecuronium (0.1 mg/kg) IV.
5. Ventilate with N₂O, O₂, and a clinically indicated dose of sevoflurane or isoflurane.
6. Give lidocaine 1.5 mg/kg IV and fentanyl (2–5 µg/kg) when the child is fully relaxed, intubate the trachea.
7. Maintain anesthesia with N₂O, O₂, and either isoflurane or sevoflurane with controlled ventilation.
8. Establish arterial and CVP access for monitoring. An epidural catheter may be useful to supplement intraoperative analgesia, postoperative pain control, and to improve hemodynamic stability. If so, be prepared to infuse intravenous fluids and/or initiate a phenylephrine or norepinephrine infusion as necessary as the block becomes established.
9. Infuse fentanyl 5 µg/kg when surgery commences and continue with an infusion of 2 µg/kg/h. Alternatively infuse remifentanil (0.05–0.15 µg/kg/min) piggybacked with a carrier infusion on a pump through a separate dedicated IV.
10. When the tumor has been excised:
 - (a) Transfuse fluids rapidly to maintain arterial pressure. Large volumes may be required.
 - (b) Maintain CVP at 9–11 cm H₂O, and check the Hct periodically.
 - (c) If hypertension persists, suspect additional tumors.
11. When the tumor or tumors have been removed:
 - (a) Discontinue inhalation agent
 - (b) Continue anesthesia with N₂O, O₂, fentanyl or remifentanil infusions, and muscle relaxant as indicated until the end of surgery.
 - (c) Recall that the duration of phenoxybenzamine will last far into the recovery period and that blood pressure support may be needed.

Postoperative

1. Check blood glucose levels frequently. (Hypoglycemia may occur as a result of the fall in catecholamine level and a secondary rebound hyperinsulinism.)
2. Order maintenance intravenous fluids that contain dextrose.
3. Anticipate an increase in Hct as the effect of phenoxybenzamine wears off.
4. Maintain epidural analgesia or order analgesics as required.

Wilms Tumor (Nephroblastoma)

Wilms tumors constitute 50% of the retroperitoneal masses in children but these tumors usually manifest as an abdominal mass. Stage 1 = one kidney only (~40%), Stage 2 = local spread outside of kidney but resectable (~23%), Stage 3 = spread outside of kidney but not resectable, e.g., to lymph nodes (~23%), Stage 4 = tumor spread to liver, brain, and bone (~10%), Stage 5 = tumors in both kidneys (~5%). Abdominal pain and fever are common symptoms. Hypertension may develop, possibly as a result of ischemia of renal tissue adjacent to the tumor, but the BP may remain elevated after removal of the entire affected kidney. Recent approaches include laparoscopic techniques, retroperitoneal approach (possible less pain), and renal tissue sparing techniques.

Associated Conditions

Hemihypertrophy, absence of the iris (aniridia), Beckwith-Wiedemann syndrome, Sotos syndrome, Denys-Drash syndrome, Fanconi anemia, and others.

Special Anesthesia Problems

1. Massive blood loss may occur (arterial and CVP lines should be placed) (see Chap. 17).
2. Surgical manipulations may kink the IVC and cause abrupt falls in cardiac output.
3. A thoraco-abdominal approach may be required for large tumors.
4. The size of the tumor and previous whole-body radiation may impair pulmonary function.
5. Hypertension may be present (60% of cases) secondary to renin secretion. It may be severe in some children and may require preoperative and intraoperative therapy. Angiotensin-converting enzyme (ACE)-inhibiting drugs (e.g., captopril) and beta blockers have been suggested as most appropriate preoperatively.
6. Rarely, tumor may invade the IVC; in such cases, an intraoperative pulmonary tumor or thrombus embolism may occur and cause sudden catastrophic hypotension.
7. A coagulopathy, acquired von Willebrand disease, may occur in association with Wilms tumor. This improves after resection of the tumor. Factor VIII concentrates may be required to reduce the bleeding time.
8. Anemia is commonly present.
9. Large intra-abdominal tumors are associated with delayed gastric emptying and potential regurgitation at induction of anesthesia.

Anesthesia Management

Preoperative

1. Check that an adequate supply of blood products is available.
2. Do not palpate the child's abdomen.
3. Consider the use of antacids and metoclopramide to reduce the risk of acid aspiration.
4. Anticipate a massive rapid blood loss. Be prepared.
5. Anticipate the need for vasopressors. Have them set up and in place ready for emergent use.
6. Anticipate possible severe hypocalcemia if rapid infusion of blood products (FFP or platelets) are needed.
7. Anticipate the possible need to treat episodic hypertension
8. Consider magnesium sulphate boluses (25–50 mg/kg over 20 min) to control hypertensive events
9. Consider a remifentanil-based anesthetic to control heart rate and BP
10. Consider an epidural for postoperative analgesia.

Perioperative

1. Apply monitors and induce anesthesia. An RSI is preferred.
2. Start intravenous infusions in an upper limb or neck vein using large cannulae.
3. Maintain anesthesia with isoflurane or sevoflurane, and a relaxant (rocuronium or vecuronium) or low-dose inhalation agent and remifentanil infusion through a separate dedicated IV piggybacked to a carrier infusion.
4. Insert an arterial line and a double lumen CVP to monitor volume status and provide a reliable route for vasopressors or vasodilators.
5. Beware of abrupt decreases in BP (due to surgical compression of the IVC). Notify the surgeon to desist immediately if this occurs.
6. If hypertension occurs (unusual), increase the inspired concentration of inhalation agent and consider treatment with magnesium, esmolol, or increased infusion of remifentanil.

N.B. Significant blood losses may occur during wound closure: continue transfusion to match losses. Be aware of possible hypotension after tumor excision when renin levels suddenly decrease. (Do not relax as soon as the tumor is out!)

Postoperative

1. Hypertension may continue and may require therapy (e.g., hydralazine).
2. Blood loss into the wound may continue, requiring ongoing transfusions.
3. Postoperative analgesia may be provided with an epidural catheter.

Acute Abdomen

In children, an “acute abdomen” most commonly represents acute appendicitis, intussusception, or perforated Meckel diverticulum.

Appendicitis

Appendicitis is the most common cause of acute abdomen in childhood. The concerns for the anesthesiologist are possible fluid and electrolyte disturbance (secondary to emesis) and the presence of sepsis and high fever. Adequate fluid resuscitation should be ensured before proceeding to general anesthesia.

Intussusception

Intussusception is a common cause of obstruction in the first 5 years of life. A segment of bowel passes into more distal bowel and may become ischemic and gangrenous. Enlarged Peyer patches, caused by viral infection, may precipitate this lesion by providing the lead point. The diagnosis is confirmed by contrast enema, which may also serve to reduce the intussusception. If this fails, a second attempt at hydrostatic reduction under general anesthesia may be successful; inhalation anesthetics may facilitate the process by relaxing abdominal muscles, decreasing smooth muscle activity, and reducing splanchnic blood flow. Pneumatic pressure of air or oxygen has also been used to reduce an intussusception. In this case, there is a risk of gas embolism, so N₂O should be omitted from the anesthesia technique. Laparotomy or a laparoscopic approach is indicated for peritonitis, failed reduction, and repeated episodes. It should be noted that occasionally significant and hidden blood loss may occur within the intussusception; some children will require transfusion.

Meckel Diverticulum

Meckel diverticulum is partial persistence of the omphalomesenteric duct, which is present in ~2% of the population; it may provide a site for bleeding, perforation, or intestinal obstruction. Severe bleeding may occur from ectopic gastric mucosa within the diverticulum and may result in hypovolemic shock.

Special Anesthesia Problems

1. Full stomach: even if the child has been NPO for several hours (and even if the child has vomited), do not assume that the stomach is empty.
2. Fluid and electrolyte disturbances may occur secondary to vomiting.
3. The child may have a high temperature; this increases the metabolic rate for oxygen and compounds the risk should any interruption of ventilation occur. It also increases the maintenance fluid requirements by 10–12% per 1C° increase in body temperature.

Anesthesia Management

Preoperative

1. Assess the child's general condition. Check the volume status, fluid intake, serum electrolytes, and urine output. Ensure that fluid replacement is sufficient to correct deficits and produce good urine output.
2. Ensure that blood has been cross-matched and sent to OR
3. If the child's temperature is increased, avoid atropine or hyoscine.
4. Prepare and check all equipment for a rapid-sequence induction (Chap. 4) and have suitable assistance available.

Perioperative

1. Check that a reliable intravenous route is available.
2. Apply standard monitors.
3. Preoxygenate as tolerated
4. Perform an RSI and intubate the trachea
5. Consider placement of an epidural catheter for postoperative pain relief
6. Maintain anesthesia with N₂O, inhalation agent, and a non-depolarizing muscle relaxant.
7. Children with high fever may benefit from intraoperative cooling, facilitated by the use of inhalational anesthetics and muscle relaxants.
8. At the end of surgery, stop all anesthetic agents and antagonize the relaxant.
9. When the child is fully awake, extubate the trachea with the child in the lateral decubitus position.

Postoperative

1. Order analgesics as required or continue local anesthetic infusion via epidural catheter for 48–72 h.
2. Monitor temperature postoperatively. Cool the child if fever persists

Testicular Torsion

Testicular torsion requires immediate surgery to save the testis. Therefore, in most cases, it is not possible to prepare the child. Always assume a full stomach is present.

Anesthesia Management

Preoperative

1. Prepare and check all equipment for an RSI (Chap. 4) and have suitable assistance available.

Perioperative and Postoperative

1. As for “Acute Abdomen” page 372.

Organ Transplantation: Care of the Donor

Transplantation of solid organs is now becoming common in children. Although transplantation (apart from kidneys) is limited to a few specialist centers, organ procurement may be performed in many hospitals. The anesthesiologist has a major role to play in the care of the donor to ensure that donated organs remain in optimal condition until harvesting. Donation from living-related donors is also increasing; thus, anesthesia care will be needed for two patients, often in adjacent operating rooms.

Determination of Brain Death

Determination of brain death has been based on the following: the presence of deep coma, lack of brain-stem function, unresponsiveness to stimuli, together with a host of criteria (including EEG) that vary from jurisdiction to jurisdiction. Before brain death is declared, all local criteria for “brain death” in an infant or child must be satisfied. A new advance to increase organ donations is donation after withdrawal of life support. In such situations, it is common for the brain dead donor to be brought to the OR where care is withdrawn and once either extreme hypotension or loss of heart beat occur, the organs are harvested. Another source of organs is from donors who have suffered prehospital admission cardiac arrest. A further strategy to improve organ donations is to institute extracorporeal support after circulatory determination of death. Further, even donation from brain dead neonates is being considered. All of these strategies have contributed to additional organs for pediatric transplantation. Each institution should develop specific guidelines based on national and local norms for the processes involved in the care of such donors.

Care of the Donor

When cerebral death occurs, a sequence of physiologic changes follow throughout the body that may compromise the survival of organs destined for transplantation. Hence, intensive measures to support these organs are indicated. The anesthesiologist will be involved in caring for the donor during organ retrieval. It is very useful to discuss management strategies with the surgical harvest teams as the goals of one (e.g., liver) may differ from that of another (e.g., heart). These strategies are particularly needed for donors after withdrawal of care and those that suffer out-of-hospital cardiac arrest.

After “brain death,” the following occurs:

1. Hemodynamic instability: Widespread vasodilation occurs, and the child tends to become pink and hypotensive. This hypotension may be compounded by hypovolemia secondary to the use of diuretics for previous attempts at cerebral resuscitation. Myocardial function may also be depressed. As the brain stem fails or in the presence of raised intracranial pressure, hypertension may occur. In the gravest situations, wide swings from hypotension to hypertension occur.
2. Central diabetes insipidus leads to polyuria, dehydration, hyperosmolarity, and hyponatremia.
3. Arrhythmias, atrial or ventricular, are frequent owing to intracranial pressure changes, electrolyte disturbances, and myocardial injury.
4. Hypothermia is present owing to loss of central thermoregulation.
5. Coagulopathy occurs secondary to disseminated intravascular coagulation as a result of released substrates from a necrotic brain.

To counter these changes, the following management regimen is suggested:

1. The circulating volume should be restored with a balanced salt solution (20–40 mL/kg); avoid over-hydration.
 - (a) Lactated Ringer’s solution or normal saline. Use 5% dextrose in water if the Na concentration is greater than 150 mEq/L but avoid hyponatremia and hyperglycemia.
 - (b) Albumin 5%. Volume expansion to maintain the CVP greater than 8 cm H₂O and an adequate urine output.
 - (c) Packed red blood cells for Hct less than 30%. Warm all fluids to prevent hypothermia.
2. If hypotension persists despite adequate volume expansion, check electrolyte levels (Ca⁺⁺) and then commence vasopressors with:
 - (a) Dopamine 5–15 µg/kg/min first.
 - (b) Epinephrine up to 0.1 µg/kg/min.

Vasopressors should, if possible, be discontinued or reduced in infusion rate before organs are harvested to minimize the chance of ischemic injury. The use of dopamine in cardiac donors should be limited to that required to stabilize hemodynamics; excessive doses may deplete energy stores and damage the myocardium; doses greater than 20 µg/kg/min are contraindicated.

3. Renal function should be maintained by fluid loading:
 - (a) If urine output decreases to less than 1 mL/kg/h, give furosemide 1 mg/kg.

- (b) For diabetes insipidus, give *either* 1-deamino-8-D-arginine vasopressin (DDAVP), 1–4 µg IV, or pitressin infusion, 0.5–2 mU/kg/h titrated to maintain the desired output.
 - (c) Suitable electrolyte solutions should be administered to correct hypernatremia and hypokalemia.
4. Hepatic function should be preserved by maintaining oxygenation and perfusion.
5. Other measures:
- (a) Measure esophageal and rectal temperature and maintain normothermia.
 - (b) Determine and correct acid–base and electrolyte status.
 - (c) Optimize ventilation (beware of pulmonary changes).
 - (d) Use careful aseptic technique. Prophylactic antibiotics may be requested. Blood cultures are taken immediately before organs are harvested.

Liver Transplantation

Major advances in the control of rejection, surgical and anesthesia techniques have made liver transplantation common for infants and children; transplantation of a portion of a living-related donor organ or split liver cadaver donor have improved organ availability. Common indications for pediatric liver transplantation include the following:

- 1. Biliary atresia
- 2. Metabolic disease (e.g., α_1 -antitrypsin deficiency)
- 3. Liver tumors—The results are significantly better in infants and children older than 1 year of age who weigh more than 10 kg.
- 4. Ingestion of liver toxins
- 5. Cirrhosis

Special Anesthesia Problems

- 1. Major blood losses may occur requiring massive blood transfusion.
- 2. Intraoperative circulatory instability may result from preexisting myocardial disease, plus mechanical factors (surgical manipulation), electrolyte disturbances (K^+ , Ca^{++}), acidosis, hepatic encephalopathy, and release of vasoactive and cardiotoxic factors on reperfusion.
- 3. Coagulation defects may preexist secondary to impaired preoperative hepatic function, secondary hypersplenism, and are compounded by massive blood losses.
- 4. Metabolic derangements may occur including hypothermia, hypoglycemia (rare), hyperglycemia (more common), hypernatremia secondary to bicarbonate therapy, ionized hypocalcemia secondary to citrate, and hyperkalemia on reperfusion.
- 5. Pulmonary function may be severely impaired secondary to liver disease (hepatopulmonary syndrome). Portopulmonary hypertension (PPHTN) may be

present in older children; it should be assessed by echocardiogram and if necessary by right heart catheterization. Mild PPHTN resolves after transplantation, severe PPHTN should be controlled preoperatively. Irreversible severe PPHTN is a contraindication to transplantation. Restrictive disease may be present secondary to ascites and pleural effusions.

6. Renal dysfunction may be present, as may hepatorenal syndrome, due to previous renal tubular damage.
7. Central nervous system dysfunction as a result of hepatic coma with increased ICP.

Anesthesia Management

Preoperative

1. Preoperative angiography to assess the vascular connections of the liver may require general anesthesia. Urgent admission at the time a donor organ becomes available is the norm.
2. Ideally, future candidates for transplantation should be electively evaluated in anticipation of pending transplant.
3. Children with PPHTN and those with possible cardiomyopathy will benefit from ECHO cardiogram assessment.
4. On date of surgery, examine the child carefully to exclude the presence of acute disease that might influence anesthesia. Assess coagulation status, BUN, creatinine, electrolytes (especially potassium which could be elevated or low), glucose, albumen, ionized calcium, magnesium, and correct as possible. Obtain a chest X-ray to assess possible pleural effusions.
5. Bowel preparation is performed.
6. Immunosuppressive therapy with high-dose steroids and other medications is initiated.
7. Anticipate increased blood losses in children younger than 2½ years of age, in those who have had previous abdominal surgery and in those with increased prothrombin times, acute liver disease, bleeding varices, or encephalopathy. Ensure that adequate quantities of blood products are immediately available. Be ready with a rapid infusion device. An increased INR is reliably predictive of likely massive blood loss.
8. Avoid intramuscular injections in children with coagulopathy; midazolam (IV or PO) premedication is preferred.
9. Many children are at risk for pulmonary aspiration; possible recent feeding, delayed gastric emptying, and abdominal distention may be present so plan for RSI.
10. Appropriate psychological support must be provided for the child and the family.
11. Anticipate the need for frequent (hourly) determination of glucose, electrolytes, acid-base, hematocrit and coagulation factors.
12. Prepare size appropriate infusions of dopamine and epinephrine.

13. Prepare infusions of fentanyl and muscle relaxant (cisatracurium).
14. Prepare resuscitation drugs (dilute epinephrine, calcium chloride, calcium gluconate, sodium bicarbonate).

Perioperative

First Stage:

The first stage includes mobilization of the diseased liver before its removal.

1. Apply basic monitors and induce anesthesia using etomidate and an RSI. If a small intravenous infusion must be started for induction, insert the catheter in a lower extremity.
2. Continue anesthesia using air/O₂/inhalation agent (sevoflurane) as tolerated, with remifentanil infusion supplementation. N₂O is contraindicated because it may cause bowel distention and exacerbate an intraoperative air embolization. Control ventilation to produce normocapnia with PEEP to prevent atelectasis.
3. Maintain neuromuscular block with cisatracurium. All drugs, including remifentanil, cisatracurium, and dopamine should be infused continuously via a central vein.
4. Insert at least three large-bore intravenous lines into the upper limbs and neck (20 gauge for infants, 14- or 16-gauge catheters or rapid infusion catheters for children). Place a large double/triple lumen CVP line; prime rapid blood transfusion and blood warming devices. Insert an arterial line, preferably in the radial artery (the abdominal aorta may have to be clamped); and a urinary catheter. Place esophageal and rectal temperature probes. Remember that a multi-lumen CVP is not adequate for rapid blood administration because of the luminal resistance and that large-bore IVs, rapid infusion catheters, or introducer sheaths are essential. The use of trans-esophageal echo to assess RV function is recommended if PPHTN is present.
5. The child should be carefully positioned and padded. A forced air warmer should cover the legs and head.
6. Monitor blood gases, electrolytes, glucose, ionized calcium, Hct, platelet count, prothrombin time, and partial thromboplastin time at frequent intervals, performing as many tests as possible on equipment in the OR suite. Other studies of coagulation (e.g., thromboelastogram) may be helpful to guide replacement therapy.
7. During mobilization of the liver, major bleeding may occur (especially if a Kasai procedure had been performed), depending on the extent of intra-abdominal adhesions from previous surgery. The intravascular volume should be replaced as necessary to maintain the CVP, BP, urine flow, and the Hct. Generally, surgeons prefer that the CVP be kept in a lower range so as to minimize venous pressure.
8. Hypotension may occur as a result of manipulation of the liver on its pedicle and compromise of IVC flow, but it may also be a result of low ionized

calcium levels. Mannitol (0.5 g/kg) may be administered to establish a brisk diuresis before clamping of hepatic and portal veins, and the hepatic artery.

Second (Anhepatic) Stage:

1. When the IVC is clamped, venous return from the lower body becomes dependent on collateral anastomotic channels unless a veno-venous bypass system is used. In this case, venous return is maintained but hazards of hypothermia, thromboembolism, and air embolism may be introduced. Veno-venous bypass may, however, reduce blood loss and improve intraoperative splanchnic and renal blood flow, with associated reduced morbidity. Veno-venous bypass is not usually used in infants weighing less than 10–15 kg because it is difficult to maintain flow in small cannulas and small infants seem to tolerate IVC clamping.
2. Hypoglycemia had been postulated as a problem of the anhepatic stage, but it is unusual because the dextrose content of infused blood products maintain normal to high blood levels. Monitor blood glucose levels frequently.
3. It is during this stage that patients are at greatest risk for ionized hypocalcemia, hypomagnesemia, and acidosis in association with transfusion of FFP because there is no liver to metabolize the citrate preservative.

Third Stage:

1. When the donor liver is reperfused, severe hypotension, arrhythmias, heart block, and cardiac arrest from hyperkalemia is possible. These arise from combined acute changes in acid–base and electrolyte levels and the effects of vasoactive and cardiotoxic factors released from reperfused, previously ischemic tissues.
2. To minimize these changes:
 - (a) Before reperfusion, the ionized calcium and bicarbonate (pH) levels should be increased with exogenous calcium and bicarbonate. Be prepared to treat hyperkalemia: calcium chloride, bicarbonate, beta agonist, possible glucose and insulin.
 - (b) Volume expansion with crystalloid or colloid solutions to maintain a CVP greater than 10 mmHg should be established. Further volumes should be immediately available for infusion.
 - (c) Rapid evaluation and correction of adverse electrolyte changes must be performed.
 - (d) Previously prepared vasopressors should be in line for instant infusion as required.
3. During the third stage, large blood losses may continue. Platelet transfusions are usually withheld until this stage to minimize the risk of vascular thrombosis in the transplanted liver.
4. The need to treat coagulopathy is based on both coagulation studies and observation of the surgical field. If oozing occurs, replacement therapy must

be instituted after discussion with the surgeon because after transplantation some children become hypercoagulable due to decreases in antithrombin III and protein C.

5. Hypertension is common late in the operation and is often unresponsive to additional opioids and antihypertensive medication. It has been attributed to multiple factors: volume overload, impaired renal function, cyclosporine, and steroid therapy. Postoperative treatment with salt restriction, diuretics, and ACE inhibitors may be indicated.
6. There may be difficulty in closing the abdomen because of the size of the implanted liver and distention of the bowel. Ventilation may be compromised, and in extreme cases the use of a Silastic pouch to close the abdomen temporarily (as in children with omphalocele) may be required.

Postoperative

1. Immediate extubation in the OR is preferred if the hemodynamics are stable and there are no other contraindications. A team discussion and decision as to the disposition of the child and airway management should occur. If surgery has been prolonged, or the child is cold or unstable, he or she is returned to the ICU intubated for continued support. Pulmonary problems are common and require aggressive therapy.
2. In some cases, return to the OR is necessary to re-explore for continued bleeding, impaired liver perfusion due to vascular thromboses, biliary obstruction, or hematoma washout.
3. Renal function may be impaired, and hypertension may be a continuing problem. Acid-base and electrolyte disturbances must be anticipated and treated.
4. There is a high risk of infection, and careful aseptic precautions are essential.
5. Neurologic complications, manifesting as seizures, are common.
6. Acute rejection—evidenced by headache, fever, malaise, nausea, and abdominal pain—may occur in 7–14 days. Liver enzyme levels may increase and synthetic functions diminish. Modification of the immunosuppressive drug regimen is required. The use of living-related donors may decrease the immunologic problems associated with liver transplantation.

COMMON MINOR SURGICAL PROCEDURES

N.B. Some children who require minor elective surgery have conditions that may complicate anesthesia and require special attention:

1. Anemia or an upper respiratory infection (see Chap. 6).
2. A history of prematurity and respiratory distress syndrome. These infants must not be considered absolutely normal even if they are now apparently healthy. See Chap. 6 for a discussion of these medical conditions.

Division of “Tongue-Tie”

If the frenulum is so short that the child has difficulty passing the tongue around the buccal sulcus, surgical division of the “tongue-tie” is probably advised. This is an outpatient procedure.

Special Anesthesia Problems

1. This is a very brief, minor procedure, but the surgeon must have good access to the oral cavity and a good airway must be ensured.
2. The surgeon may use one of the several techniques to free the tongue: clamp the frenulum and then cut with scissors or cauterize the frenulum. Sutures may also be used.

Anesthesia Management

Preoperative

1. Premedication: sedative premedication (oral midazolam) if required.

Perioperative

1. Induce anesthesia by inhaled sevoflurane or halothane or IV propofol.
2. If the surgeon does not cauterize the frenulum, maintain deep anesthesia with 8% sevoflurane in N₂O and O₂ and hold the elbow of the circuit (face mask removed) over the mouth while the surgeon clamps the frenulum. Alternately, cut a RAE tube (distal end in the oropharynx) and insert at the angle of the mouth to insufflate gasses. (Both of these methods require ingenious attempts at scavenging.) Caution is advised with electrocautery and the potential for an airway fire; use an air/O₂ blend with the lowest FIO₂. Otherwise, a tracheal tube may be inserted. IV anesthesia is probably an ideal technique (it avoids the need to scavenge), administer intermittent propofol.
3. Suction the pharynx to remove blood. Apply lidocaine gel to the sublingual wound.
4. The child should be fully awake before extubation and transfer to the PACU.

Postoperative

1. Further analgesics are not usually required.

Inguinal Herniotomy

Inguinal hernia is the most common elective general surgical procedure in children. Because these hernias readily become incarcerated during the first year of life, their repair should not be unduly delayed in this age group. Once incarceration

has occurred, conservative treatment is usually instituted. Virtually, all of these hernias can be reduced, and then, after 24–48 h, herniotomy can be performed.

If emergency surgery is to be performed as an outpatient procedure, select suitable anesthesia techniques.

The former preterm infant may benefit from spinal analgesia for herniotomy, especially if there is a history of residual pulmonary disease and as a means for reducing the potential for post-anesthesia apnea (see Chap. 4).

Anesthesia Management

Preoperative

1. Assess the child's general condition carefully.
2. Infants at risk, with a history of prematurity, should be admitted for postoperative apnea monitoring (see Chap. 4).

Perioperative

The choice of anesthesia technique for hernia repair depends on the surgeon. There are pediatric surgeons who can perform a unilateral inguinal herniotomy in 10 min—there are also those who can stretch this procedure out to last well over an hour. For the faster surgeon, general inhalational anesthesia delivered by face mask or LMA, or spinal or caudal anesthesia (for infants) is ideal; for more prolonged procedures, it is probably wise to use general anesthesia and place an LMA or intubate the trachea.

1. For general anesthesia, induce anesthesia by inhalation or with IV propofol or thiopental. Maintain anesthesia by mask with spontaneous ventilation and sevoflurane (or halothane) in N₂O/O₂.
2. For prolonged procedures, intubate the trachea or place an LMA and maintain anesthesia with N₂O and sevoflurane or halothane and assisted or controlled ventilation. Some surgeons will use a laparoscope through the hernia to assess the need to explore the opposite side; generally, very low intra-abdominal pressure is needed with insufflation.
3. Perform an ilioinguinal and iliohypogastric nerve block on the operative side (or sides) or a caudal block. Surgeons can perform these blocks during surgery while the nerves are exposed. Acetaminophen, 10–15 mg/kg PO/IV (7.5 mg/kg IV in infants) or 30–40 mg/kg PR is given before surgical incision, to prevent fever and augment postoperative analgesia. Opioids should generally be avoided because they increase the incidence of PONV.

Postoperative

1. Order additional analgesics as required. Patients who have intraoperative nerve blocks usually manage very well with acetaminophen by mouth.

Orchidopexy

Anesthesia management for orchidopexy is the same as for inguinal herniotomy. A caudal block should be performed after induction of anesthesia, before the surgery commences, to provide for postoperative analgesia. Ilioinguinal nerve blocks do not provide adequate analgesia.

Circumcision

Indications for circumcision vary in different communities and from time to time. It remains a common (often outpatient) procedure in pediatric surgery.

Special Anesthesia Problems

1. Management of postoperative pain

Anesthesia Management

Preoperative

1. Assess the child's general condition carefully.

Perioperative

1. Induction and maintenance of anesthesia as for herniotomy. Face mask or LMA anesthesia is usually adequate.
2. Provide for analgesia postoperatively. Perform either or both of the following:
 - (a) Dorsal penile nerve block using 0.25 % bupivacaine *without epinephrine*; maximal dose, 2 mg/kg.
 - (b) Perform a ring block around the base of the penis
 - (c) Apply lidocaine jelly to the wound.

Postoperative

1. If regional anesthesia is unsatisfactory (rare), administer opioid analgesia (e.g., morphine 0.05–0.10 mg/kg IV), which can be repeated in the PACU until the child is comfortable.

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Cardiovascular Surgery and Cardiologic Procedures

Heart surgery in children is performed almost exclusively for congenital heart disease (CHD). The incidence of CHD is approximately 6 per 1000 live births. The lesions listed in Table 14.1 account for more than 90% of all congenital heart defects. There are various classifications of CHD, but the one given in the table is most useful for the anesthesiologist.

THE CHILD WITH CONGENITAL HEART DISEASE

Diagnosis of Congenital Heart Disease

Today, many infants and children with CHD are diagnosed in utero, although some may remain undiagnosed throughout the neonatal period or even until later in life. In neonates, the diagnosis of CHD can be quite difficult:

1. Heart murmurs are common in the perinatal period and may not be indicative of significant heart disease.
2. Serious cardiac lesions may be present in the absence of a loud murmur.
3. The transitional circulation of the neonate may obscure the diagnosis: high pulmonary vascular resistance (PVR) limiting left-to-right shunts, and a patent ductus arteriosus (PDA) providing blood flow to the lower body (i.e., in children with coarctation or interrupted aortic arch).

Signs suggesting CHD in the neonate include cyanosis, tachypnea, prominent precordial pulsations, bounding peripheral pulses, hepatomegaly, poor feeding, cyanotic spells, and large heart on radiography. An echocardiogram (ECHO) is essential for the diagnosis and should be requested whenever CHD is suspected. Depending on the defect further workup may require additional imaging studies (cardiac catheterization, cardiac MRI, CT, or nuclear scan).

Table 14.1 Approximate incidence of congenital heart defects (CHD) in children

	Per 1000 live births	% of Total CHD cases
Ventricular septal defect	2.62	34
Atrial septal defect	1.64	13
Persistent ductus arteriosus	0.87	10
Pulmonary stenosis	0.5	8
Tetralogy of Fallot	0.34	5
Coarctation of aorta	0.34	5
Transposition great arteries	0.31	5
Aortic stenosis	0.22	4

Reference: Van der Linde D, Konings EEM, Slager MA et al. Birth Prevalence of Congenital Heart Disease Worldwide. J Am Coll Cardiol 58: 2241-7, 2011

N.B. The reported incidence has increased since diagnostic methods have improved and access to infant healthcare extended. There may be geographic and racial variations.

In older infants and children a murmur may be discovered during a routine medical examination or children may present with decreased exercise tolerance, recurrent respiratory infections, shortness of breath, chest pain, or syncope.

Congenital heart defects not only affect the growth and development of the cardiovascular system, but have significant impact on the whole body:

General Systemic Effects

- Poor weight gain and development delay.

Usually the child's height and weight are below average. Children with CHD, and especially those with cyanotic CHD, may also demonstrate some developmental delay. Children in cardiac failure, with increased work of breathing and cardiac volume or pressure load, are unable to meet the additional metabolic demands for age-appropriate growth and development. Any further metabolic stress (infection, fever, or hypothermia) can lead to decompensation.

Effects on the Respiratory System

CHD can have major effects on pulmonary function:

- Increased pulmonary blood flow:

Increased pulmonary blood flow results in small airway obstruction, decreased compliance, increased resistance, and ventilation-perfusion (\dot{V}/\dot{Q}) imbalance. Accumulation of interstitial lung water and pulmonary edema are additional factors leading to an increased work of breathing. Chronic

exposure to excessive pulmonary blood flow may lead to irreversible pulmonary hypertension (PAH) secondary to structural changes in the vessels; these include medial muscle hypertrophy and peripheral extension of the muscular layer into normally non-muscular arterioles. Early repair or temporary banding of the pulmonary artery can protect against these irreversible changes.

- Decreased pulmonary blood flow:

Children with decreased pulmonary blood flow have less efficient ventilation, requiring increased minute ventilation to eliminate carbon dioxide. The gradient between end-tidal and arterial carbon dioxide levels may be increased. The uptake of inhaled anesthetics into the blood is delayed; however, alveolar (and hence end-tidal) levels may increase rapidly. Cyanosis is associated with a reduced ventilatory response to hypoxemia.

Effects on the Heart

In addition to the special characteristics of the child's heart (see p. 24), CHD may cause other changes:

1. Obstructive lesions impose a pressure load on the affected ventricle. This ventricle then hypertrophies (becomes less compliant and less able to increase stroke volume). The thickened ventricle is subject to myocardial ischemia and consequent arrhythmias.
2. Large shunts or valvular incompetence are associated with a significant volume load on the ventricle. This ventricle initially responds with an increased stroke volume but later dilates and fails. The dilated ventricle requires a high wall tension to effect pressure change within the chamber (Laplace's law); it therefore is vulnerable to myocardial depressants and cannot cope with additional loads.
3. Myocardial ischemia may result from reduced aortic diastolic pressures and rapid heart rates in some children (e.g., those with a PDA).

Effects on the Blood

- Polycythemia and coagulopathy in cyanotic children.

Cyanosis induces compensatory changes in the blood: polycythemia and an increased blood volume. The increased hematocrit (Hct) may lead to poor microcirculation with thrombosis (subendocardial, cerebral) and abscess formation. Cyanotic CHD is also commonly accompanied by coagulopathy secondary to thrombocytopenia, impaired platelet function, and decreased clotting factors.

Effects on Hepatic and Renal Function

- Impaired organ perfusion and reduced drug clearance.

Hepatic and renal functions are impaired in cyanotic CHD and especially in those children with CHF. Splanchnic blood flow is reduced. Clearance of drugs via the liver or kidneys is delayed (i.e., morphine clearance is reduced in many children with CHD).

PRINCIPLES OF ANESTHETIC MANAGEMENT

Pathophysiologic Implications

1. Right-to-left shunts result in:

- Reduced PaO₂. This is often only minimally improved by increasing the FIO₂.
- Delayed uptake of inhaled anesthetics into the blood.
- Extreme danger of systemic emboli from venous air embolism. Make certain all IV solutions are bubble free.
- Short-arm-brain circulation time, with no pulmonary transit: potential for overdosing intravenous drugs.
- Less efficient ventilation and gas exchange; increased ventilation is necessary to maintain a normal PaCO₂, whether the child is awake or anesthetized.
- An increased arterial to PetCO₂ gradient; PetCO₂ levels underestimate arterial levels.
- Hemoconcentration
 - Polycythemia. (Hct > 55 % in cyanotic lesions) is associated with poor rheology, increased cardiac work, and greater risk for thrombosis, especially with additional dehydration or venous stasis, and cerebral abscess formation. However, acute hemodilution to Hct < 40 % may cause cardiovascular collapse: it is important to maintain the child's usual Hct.
 - Iron deficiency anemia may be present, but responds well to iron therapy.
- Coagulopathy may be present.

2. Left-to-right shunts result in:

- Pulmonary vascular overperfusion but good ventilatory efficiency and gas exchange initially.
- Later, PAH develops and progresses to irreversible increased PVR, which may limit the operability of associated cardiac lesions.

- (c) Eventual CHF.
- (d) Pulmonary hypertensive crisis: Some infants with large left-to-right shunts are at extreme risk of pulmonary hypertensive crises during and after surgery (e.g., truncus arteriosus, arteriovenous [AV] canal). A sudden increase in PVR can be triggered by hypoxia, hypercapnia, or a sudden catecholamine surge during stimulation and lead to RV failure, ischemia, and cardiac arrest. Management should include adequate anesthesia/analgesia intraoperatively, minimal handling postoperatively, controlled hyperventilation (PaCO_2 25–30 mmHg), fentanyl (2–5 $\mu\text{g}/\text{kg}/\text{h}$), or sodium nitroprusside (SNP) infusion (0.5–5 $\mu\text{g}/\text{kg}/\text{min}$), and/or inhaled NO. Prophylactic treatment before a stimulating procedure (such as suctioning the airway) postoperatively may include fentanyl (10 $\mu\text{g}/\text{kg}$) or lidocaine (0.5–1 mg/kg) IV bolus and 100 % oxygen.

3. *Obstructive lesions* may result in:

- (a) Fixed cardiac output, and therefore very limited ability to compensate for changes in metabolic demand or a decrease in SVR.
- (b) Myocardial hypertrophy, with possible inadequacy of myocardial perfusion, especially to the subendocardium. Reduced ventricular compliance results in dependence on a high cardiac filling pressure.
- (c) CHF.
- (d) Sudden serious arrhythmias.

4. *Complex “mixing” lesions:*

- (a) Venous and arterial blood are mixed at various levels before reaching the systemic circulation, often additional intra- or extracardiac flow obstructions are present.
- (b) Arterial oxygen saturation and adequate tissue perfusion depend to a large extent on a well-balanced ratio of pulmonary and systemic blood flow.
- (c) Pulmonary overcirculation will maintain oxygenation, but result in inadequate cardiac output and metabolic acidosis. Poor pulmonary perfusion will maintain systemic circulation, but lead to significant hypoxemia and cyanosis.

5. *Ductal-dependent defects:*

Some infants depend on the patency of the ductus arteriosus and the shunting of blood for adequate pulmonary or systemic blood flow (e.g., pulmonary atresia, TGA with intact septum, interrupted aortic arch, hypoplastic left heart syndrome (HLHS)). Prostaglandin E₁ (PGE₁) is used to keep the ductus open in such infants. An infusion of 0.05–0.1 $\mu\text{g}/\text{kg}/\text{min}$ should be continued until the appropriate surgical procedure is completed.

6. Congestive heart failure:

- (a) Depressed myocardial function with poor tolerance for increases in afterload or cardio-depressing effects of drugs. Prolonged circulation time.
- (b) Pulmonary edema and poor lung compliance. Rapid desaturation with apnea.
- (c) Hepatic and renal dysfunction, electrolyte disturbances.

General Principles**1. Minimize the demands on the cardiovascular system:**

- (a) Give adequate premedication to reduce anxiety, agitation, and O₂ requirements.
- (b) A rapid, smooth induction of anesthesia, with no crying or struggling, is most desirable.
- (c) Give adequate doses of analgesics and general anesthetics. High-dose opioid anesthesia combined with good postoperative analgesia may favorably influence the neuroendocrine and metabolic responses to surgery and improve survival. Prevent hypertension and tachycardia by ensuring adequate analgesia.
- (d) PAH must be controlled (See above).

2. Maintain myocardial function and cardiac output:

- (a) Avoid agents that cause excessive myocardial depression.
- (b) Adjust the fluid balance to maintain optimal cardiac filling pressures.
- (c) Assure adequate myocardial perfusion and oxygen supply:

- The duration of diastole and the diastolic pressure are important factors in maintaining perfusion of the myocardium, which is especially vulnerable if left-to-right shunting (causing low aortic root diastolic pressure) and ventricular hypertrophy are present. Avoid tachycardia, hypotension, and anemia, and also excessive hypertension, which may impair subendocardial perfusion.
- Maintain an adequate hematocrit for sufficient oxygen delivery. Cyanotic children are particularly dependent on a greater hematocrit (Hct > 40–45 %). Significant anemia may cause subendocardial ischemia in some defects.

3. Optimize ventilation for each child: Maintain normocapnia: avoid hyperventilation, unless indicated for the management of increased PVR. The resulting hypocarbia might:

- (a) Reduce cardiac output.
- (b) Cause vasoconstriction and increase SVR.

- (c) Decrease PVR and increase left-to-right shunts.
 - (d) Shift the hemoglobin/oxygen (Hb/O_2) dissociation curve to the left and limit O_2 transfer.
 - (e) Decrease myocardial blood flow.
 - (f) Decrease the serum potassium level, resulting in arrhythmias.
 - (g) Decrease cerebral blood flow.
4. *Prevent detrimental changes in cardiac shunts:*
- (a) Use anesthetic drugs that have minimal effects on SVR.
 - (b) Be aware of the possible effects of intermittent positive-pressure ventilation (IPPV) on shunts; avoid high intrathoracic pressures, but maintain the lung volume as necessary by the use of optimal positive end-expiratory pressure (PEEP). PVR is minimal at an optimal lung volume and increases at greater or lesser degrees of lung inflation.
 - (c) Drugs that produce a controllable degree of myocardial depression (e.g., inhalational agents) may be useful when hyperdynamic ventricular muscle causes obstruction to blood flow and increased shunting (e.g., TOF, IHSS).
 - (d) Children who depend on systemic-to-pulmonary shunts will desaturate if the systemic arterial pressure decreases.
 - (e) Anemia may increase left-to-right shunts. Conversely left-to-right shunting may be reduced by increasing Hct. Changes in blood viscosity have a greater effect on PVR than on SVR.
 - (f) Be prepared to use drugs or other methods to manipulate PVR or SVR.
5. *Give adequate doses of muscle relaxants to prevent movement or ventilatory efforts:* especially when the heart is open (danger of air emboli). The time to maximum effect of non-depolarizing agents in children with CHD is greater than in those without CHD; a more prolonged period of mask ventilation may be needed before intubation.
6. *Maintain body temperature* and prevent cold stress except when induced-hypothermia is indicated. Temperature control may be especially poor in neonates with cyanotic CHD; body temperature decreases rapidly if they are exposed to a cool environment. Vasoconstriction in the cold child impairs efforts at insertion of intravenous lines and may result in metabolic acidosis.
7. Heparin has a larger volume of distribution and a more rapid plasma clearance in infants than in adults. Therefore larger doses may be required initially, an activated clotting time (ACT) of 480 s or more is required before cardiopulmonary bypass (CPB), and the level of heparinization should be checked frequently (every 30 min). An ACT of 480 s is the commonly used threshold value to attain before and during CPB. However, some have questioned whether this is adequate to prevent thrombin formation in the hemodiluted, hypothermic infant, and suggest that a greater ACT is more appropriate (i.e., 600 s). Kaolin-activated ACT appears to produce more reproducible ACT results than Celite activated ACT. The determination of

blood heparin levels* may provide a better means to monitor heparinization and is being performed in some units.

8. During CPB, the myocardium may be protected by:

- (a) Cardioplegic solutions that are infused at a pressure of 100–150 mmHg into the coronary circulation after aortic clamping. Controversy still exists concerning the most advantageous type of solution, and this may differ in infants and adults. Most solutions contain increased concentrations of potassium with added dextrose and pH buffers. The addition of free radical scavenger agents and calcium ion channel blockers has been suggested. The ideal cardioplegic solution:
- Produces immediate arrest and prevents energy depletion.
 - Provides substrate for anaerobic metabolism.
 - Buffers metabolic acidosis in the tissue.
 - Minimizes tissue edema by its osmolar effects.
 - Stabilizes cell membranes.
 - Minimizes reperfusion injury.

Blood cardioplegia is preferred by many institutions. Repeated doses of cardioplegic solution are normally given at 15- to 20-min intervals.

- (b) Hypothermia. Remember that the heart has a great tendency to rewarm because of surgical manipulation and heat from operating room lights. Therefore, during prolonged surgery, cold cardioplegic solutions should be repeatedly applied, and a pericardial cooling bath should be used.
- (c) Pre-CPB systemic corticosteroids may help preserve myocardial tissue during periods of ischemic arrest, but this is controversial.
- (d) An optimal reperfusate solution may be used after a period of ischemic arrest. This may flush out metabolites and prevent reperfusion injury. This solution should be warmed and alkaline, contain a minimal concentration of ionized calcium and a slightly increased potassium concentration. In practice, a repeat dose of warmed cardioplegic solution is often given just before reperfusion.

Preoperative Assessment and Preparation

1. Review all the medical records (including previous anesthetic records and identify any difficulties) and obtain a history from the parents.
2. Identify additional defects or syndromes (e.g., cleft palate, Down syndrome, subglottic stenosis) that may complicate anesthesia and require special considerations.

*Hepcon HMS Plus system, Medtronic, Minneapolis MN or Hemochron® RxRx®, ITC, Edison, NJ.

3. Perform an independent physical examination, especially of the cardiovascular and respiratory systems, ears, nose, throat, teeth, and veins. Examine for signs of cardiac failure: tachypnea, sweating, and hepatomegaly in infants. Determine the respiratory status to exclude acute disease that might compromise the child perioperatively. If the child had a significant lower respiratory tract infection recently, elective surgery should be postponed for 2–3 weeks because of an increased susceptibility to pulmonary complications during this period.
4. Review the cardiology notes, most recent ECHO, cardiac catheterization, and angiographic data to fully understand the current cardiac pathophysiology. Note salient abnormalities and findings on the anesthesia chart.
5. Itemize all current medications and laboratory tests. With rare exceptions, ACE inhibitors, digitalis, and diuretics should be withheld on the day of surgery, whereas antiarrhythmic drugs and β -blockers should be continued (to avoid rebound effects). Many cardiac drugs have significant implications for the anesthetic management:
 - (a) Diuretics can cause hypovolemia and electrolyte disturbances (hypokalemia, hypocalcemia, and hypomagnesemia) and predispose the children to perioperative arrhythmias especially in association with hypothermia or digitalis.
 - (b) Digitalis is rarely used today. The therapeutic index is low, and toxicity is an ever-present hazard, especially in young children. If digitalis is prescribed, check a recent serum level (therapeutic range, 0.8–2 ng/mL).
 - (c) ACE inhibitors (particularly if taken on the day of surgery) can cause significant and often refractory hypotension after induction of general anesthesia
 - (d) β -Adrenergic blocking agents may impair cardiac contractility, but are often used as antiarrhythmics or to prevent “hypercyanotic spells” in those with TOF. They should not be stopped abruptly.
 - (e) Calcium channel-blocking agents are only occasionally used in children and adolescents, mainly for the treatment of PAH. In infants, they can cause severe, persistent myocardial depression. The combination of β -adrenergic blocking agents with calcium channel blockers is very dangerous and is best avoided.
 - (f) Anticoagulation with platelet inhibitors or warfarin can increase the risk of bleeding, but is often essential for thrombosis prophylaxis. Occasionally bridging with heparin is indicated.
6. If the child requires O₂ therapy and/or maintenance of the sitting position during transit to the operating room (OR), order these specifically.
7. Plan in advance for postoperative pain management and discuss this with the parents. In those whose tracheas may be extubated early, spinal or epidural

opioids may be useful. In most other cases, pain can be managed by an intravenous infusion of an opioid and/or PCA.

Premedication

1. Children with CHD require adequate preoperative sedation to reduce excitement, anxiety, and crying (and thus reduce O₂ consumption).
2. In recent years, an oral regimen has come to be preferred:
 - (a) Midazolam 0.5–1 mg/kg PO (maximum 20 mg) is very effective; allow 10–30 min for the peak effect to be achieved. Alternately, ketamine (5 mg/kg PO) or a combination of midazolam (0.25–1 mg/kg) and ketamine (3–5 mg/kg) may be effective. Children given this mixture should be constantly observed and SpO₂ monitored. (Suction and airway equipment should be available).
 - (b) Lorazepam 1–2 mg PO is effective for the adolescent patient. This pre-medication does not usually decrease SaO₂, even in cyanotic children. However, the child should be supervised as sedation occurs, and a pulse oximeter may be used as the child becomes settled.
3. Topical local anesthetic cream may be applied to a predetermined site for intravenous cannulation, covered with an occlusive dressing, to allow pain-free IV placement.
4. For cyanotic children with a high Hct, ensure that oral fluids are regularly offered up to 2 h before the operation to prevent dehydration. Alternatively, maintenance IV fluids should be administered during the preoperative fasting period.

Blood Supplies

During all cardiac surgeries, blood must be immediately available in the OR. In many centers, the surgical service is responsible for ordering blood. However, the day before surgery, the anesthesiologist should ensure that there will be an adequate supply of blood and blood products by the time surgery begins.

Special Blood Requirements

1. For cyanotic children with an Hb >16 g/dL, plasma should be available.
2. For all infants, check that the available blood is <3 days old and has been tested for cytomegalovirus. Washed cells should be ordered for small infants to prevent the danger of hyperkalemia. Radiated RBCs or leucocyte poor

blood may be indicated for some children (immune deficient, Di George Syndrome or transplant children).

3. For infants undergoing CPB, ensure that appropriate quantities of packed cells, FFP, and platelets (1 unit/5 kg) and cryoprecipitate have been ordered. Fresh whole blood is considered especially advantageous, and is reported to reduce bleeding after CPB, but may be difficult to obtain. Alternatively, "reconstituted blood," a mixture of packed cells and FFP is often used to prime the CPB circuit.
4. For all children likely to require prolonged CPB (greater than 1.5 h), ensure that cryoprecipitate, and platelets have been ordered.
5. Where "relatively minor" surgery is planned for older children and those with an initially high Hct, hemodilution with a clear fluid prime in the pump oxygenator may avert the need for blood transfusion. At the end of CPB, modified ultrafiltration may be used to remove the fluid prime and restore the Hct. Alternatively, the contents of the pump circuit may be washed and hemoconcentrated via a cell-saver for use in the postoperative period. However this does not conserve coagulation factors! (Blood should be ordered to be available on a standby basis.)

ANESTHESIA MANAGEMENT

Routine Anesthesia Management

Preoperative

1. Check all anesthesia and monitoring equipment before the child enters the OR
2. Have the following available in case of emergency:
 - (a) Sodium bicarbonate, 8.4 % solution 20 mL volume
 - (b) Atropine (0.4 mg/mL) diluted in 4 mL N saline (0.1 mg/mL solution).
 - (c) Calcium chloride, 10 % solution: 10 mL volume
 - (d) Epinephrine, 1:10,000 preparation: 10 mL volume
 - (e) Phenylephrine, 0.1 mg/mL: 10 mL volume

Solutions of inotropic drugs should be prepared, loaded on appropriate infusion pumps with appropriate initial settings and primed. These should be made in a concentration that will permit their infusion at a therapeutic rate without adding an excessive fluid load and set up in such a manner that a carrier infusion of balanced salt solution assures timely delivery. In practice, for small infants and children, it is useful to use a dilution that will deliver the required dose when infused at 1–2 mL/h (see Appendix C).

3. Check that preoperative medication has been given as ordered and is effective.
4. On arrival in the OR, gently apply basic monitors: pulse oximeters (one probe to a finger, thumb, or ear and one probe to a toe), precordial stethoscope, BP cuff, and ECG electrodes. Record HR and rhythm and BP. Do not prolong this process, especially if the child is apprehensive, but proceed carefully and rapidly.

Perioperative

1. Administer O₂ by mask. Often the child will be happier if the mask is held slightly away from the face. Use a high flow.
2. Induction of anesthesia.

Different induction methods may be used, and if they are well conducted, the SaO₂ will increase with all of them, even in cyanotic children; therefore, the anesthesiologist may choose whichever method seems most appropriate for the child.

In our practice, an intravenous induction using an opioid analgesic, a very small dose of propofol (or etomidate), and an intubating dose of a non-depolarizing relaxant are usually preferred particularly in children with right-to-left shunts, which slow inhalation inductions. This approach allows for good ventilation, rapid airway control, and very stable conditions. Use of a topical local anesthetic cream and suitable sedation facilitate the ease of venous cannulation. The advantage of establishing venous access outweighs the potential for upsetting the child. Ketamine IM has been commonly used, but an IM injection always upsets the child and leads to stress and crying. Furthermore, ketamine increases CMRO₂ and may cause hypertension!

For most children, fentanyl 2–5 µg/kg followed by propofol (3–5 mg/kg) given slowly produces a smooth induction with minimal cardiovascular effects. In small infants, precede the fentanyl by a small dose of atropine (0.01 mg/kg) or an appropriate dose of pancuronium to prevent bradycardia. For the very unstable child, use fentanyl (5–10 µg/kg) and incremental small doses of IV midazolam (up to 0.2 mg/kg) or etomidate (0.2–0.3 mg/kg) for induction. Be careful with rapid administration of large doses of fentanyl before muscle relaxation as it can cause significant chest wall rigidity.

3. Drugs given intravenously should be administered in small doses, *slowly*. (If a right-to-left shunt is present, they act very rapidly; but if the circulation time is slow, their effect may be less rapid.) Be patient and wait for the desired effect. Beware of overdose.
4. For tracheal intubation: give an initial dose of non-depolarizing relaxant and ventilate the lungs until relaxation is adequate; rocuronium 0.5–1 mg/kg or

vecuronium 0.1 mg/kg produce good or excellent intubating conditions within 3 min (see also item 9 in this list).

NB: Be aware of the possibility of subglottic stenosis in children with CHD, be prepared to use a smaller diameter tracheal tube.

5. Use a cuffed tracheal tube to ensure ability to ventilate well. The tube should pass easily through the glottis and subglottic space; otherwise use a smaller diameter tube. Carefully position the tube and check bilateral ventilation. The cuff does not usually need to be inflated in infants to prevent leaks at normal ventilator pressures.
6. Ventilate with a suitable mixture air/O₂. (It is rare to require >50% O₂. If a large right-to-left shunt is present, an increase in the FIO₂ has very little effect on the PaO₂.) It is probably advisable to avoid N₂O, which can increase the size of air bubbles and potentially exacerbate PAH, although the effect on PVR is small.
7. If myocardial function is good, for simple lesions, low concentrations of inhaled agents may be used. Otherwise, for all complex lesions, add opioids in adequate doses.
8. Control ventilation to produce desired carbon dioxide tension. Note that PetCO₂ is a satisfactory means to monitor PaCO₂ in acyanotic children but may underestimate the PaCO₂ in those with cyanotic CHD. Always compare the PetCO₂ with the PaCO₂; the PetCO₂ can then be used to follow trends.
9. The choice of muscle relaxant during maintenance of anesthesia should be influenced by the following: rocuronium and vecuronium have very little effect on cardiovascular parameters, have an intermediate duration of action, and, if properly dosed, can readily be reversed for early extubation. They are probably the agents of choice for many infants and children.
10. Insert a nasopharyngeal and rectal or bladder thermometer. An esophageal stethoscope cannot be inserted if a transesophageal echocardiography (TEE) probe will be used.
11. Insert adequate-bore intravenous routes, an arterial line, a double-lumen central venous line (see Chap. 4), and a urinary catheter. In older children who may be extubated early, it is preferred if possible to place the arterial line and the intravenous line in the same upper limb (usually the left). The other hand can then be used to operate a PCA pump.
12. Give maintenance fluids as outlined in Chap. 4. All fluid administration should be regulated by infusion pumps. Depending on NPO time and cardiac function, the cyanotic child with an increased hematocrit might benefit from a small fluid bolus of a balanced salt solution to improve the rheology and microcirculation. For CPB cases, slow cooling is preferred so no fluid warmer is needed pre-bypass.
13. For those whose trachea may be extubated early after surgery, consider the use of epidural or spinal opioids (see Chap. 5).

Open-Heart Surgery

1. Follow routine management (see previous discussion). Additionally, cerebral oximetry via near infrared spectroscopy (NIRS) and transcranial Doppler (TCD) can be used to monitor cerebral perfusion, but require specialized age and size-appropriate equipment; and for the TCD, training, and experience. For older children and adolescents, processed EEG monitoring can be helpful to minimize the risk of awareness, especially during the rewarming phase of CPB.
2. The intraoperative use of TEE has gained widespread acceptance: it helps to define the exact anatomy and pathophysiology and to assess the adequacy of the repair immediately after bypass. Residual shunts and air bubbles can be detected; valve function, ventricular filling, contractility, as well as flow characteristics in conduits and shunts may be evaluated. Special probes for children and even small infants are available. If a TEE probe is to be inserted, monitor ventilation, SaO_2 , the PetCO_2 waveform, and systemic BP very carefully. Passage of the probe is stimulating and may trigger autonomic reflexes in small infants, compromise ventilation, displace the tracheal tube, and/or compress the major vessels
3. Maintain anesthesia with:
 - (a) N_2O or Air/ O_2 in suitable proportions to ensure SaO_2 at an acceptable level. For lesions with significant pulmonary overcirculation, it may be necessary to reduce the FIO_2 ; deliver 21% oxygen (or close to it) and adjust the ventilator rate to achieve normoventilation.
 - (b) If tolerated, isoflurane 0.5–0.75% or sevoflurane 2% may be given and supplemented with generous doses of fentanyl.
 - (c) Children with a history of CHF who may benefit from afterload reduction will probably do well with minimal isoflurane (e.g., ventricular septal defect (VSD) with left-to-right shunt). Children with dynamic ventricular outflow obstruction who may benefit from a degree of controlled myocardial depression usually also do well with minimal isoflurane or even better low-dose halothane (0.5%), if available. (e.g., TOF with dynamic RV outflow obstruction, hypertrophic obstructive cardiomyopathy).
4. Give incremental doses of muscle relaxants as needed. Administer an additional generous bolus just before bypass to ensure complete immobility.
5. Give appropriate volumes of balanced salt solution based on the body weight to replace the calculated deficit during fasting (if any) and maintain urine output greater than 1 mL/kg/hr. Additional “fluid loading” before CPB has not been shown to be advantageous. Avoid dextrose-containing fluids; hyperglycemia may exacerbate a neurologic injury in case of cerebral

- hypoxia/ischemia. Monitor blood glucose concentrations in small infants to detect hypo or hyperglycemia.
6. Blood loss from sponges, suction, drapes, and blood specimens (i.e., blood withdrawn for analysis) must be measured, and the volume replaced. It is seldom necessary to transfuse blood before CPB unless major blood loss occurs during opening of the chest or dissection around the heart (i.e., during repeat operations). In repeat operations, warmed lactated Ringer's solution or blood (depending on the size of the child) should be hanging ready for infusion at the time of sternotomy; additional blood should also be immediately available in the OR.
 - (a) Aim to maintain the Hct near the preoperative level and the intravascular volume at a level to maintain CVP.
 - (b) If the Hct was markedly increased preoperatively, replace initial losses with albumin, but refrain from significant hemodilution before CPB. Excess hemodilution may compromise oxygen delivery especially in cyanotic CHD.
 - (c) During venous cannulation in small infants, a significant volume of blood may be sequestered into the cannulae. Ensure that this volume is replaced, usually by transfusion from the CPB circuit via the aortic cannula.
 7. Children with cyanotic CHD or those scheduled for repeat procedures may benefit from the administration of antifibrinolytic agents (e.g., tranexamic acid) to reduce bleeding.
 8. During dissection around the heart, monitor the ECG monitor and BP closely; arrhythmias are common, although most are innocuous. If hypotension or arrhythmia persists, notify the surgeon and stop surgery until the condition corrects itself. Continuing hypotension suggests hypovolemia, poor contractility or ischemia. Depending on the most likely cause it should be treated with fluids, inotropic support or surgical maneuvers to improve coronary perfusion.
 9. If N₂O has been used, discontinue it before cannulation to prevent expanding any potential air embolism.
 10. Obtain a baseline ACT. Before the heart is cannulated, give the initial dose of heparin 300–400 units/kg depending on the heparin manufacturer. Recheck the ACT after 2–3 min; the ACT should be at least 480 s before initiation of CPB. Small infants may require more heparin and demonstrate more variation in dose requirements than older children.
 11. Once CPB is established, the pump flow should be increased to establish a satisfactory perfusion. Indicators of adequate perfusion are cerebral oximetry (NIRS), urine output, and the absence of metabolic acidosis on repeated acid-base studies. In children with cyanotic CHD, perfusion

pressures may be low during early bypass because of the child's decreased vascular resistance (children with cyanotic CHD have larger vessels) and the use of a low-viscosity perfusate. Those with TOF may also have extensive collateral flow into the lungs. High flows may be required initially, but the systemic pressure will increase progressively, especially as cooling progresses. The use of vasoconstrictors is not usually necessary but should be considered if hypotension persists. When the perfusion pressure is low, it is vital that the superior vena cava (SVC) pressure should also be at or near zero. Under these circumstances, any increase in jugular venous pressure, often caused by a malpositioned obstructing SVC cannula may have a serious effect on cerebral blood flow. Monitor CVP and NIRS carefully to detect any compromise of SVC venous return and alert the surgeon immediately.

12. During partial bypass, ventilate the lungs with 100% O₂. Never use N₂O because of the possibility of expanding an air embolism.
13. During total bypass:
 - (a) Keep the lungs inflated at a low pressure.
 - (b) Add 0.5% isoflurane to the oxygenator to continue anesthesia and improve perfusion during bypass, or give additional doses of fentanyl. They are usually discontinued during rewarming. Fentanyl may bind to the plastic components of the CPB circuit, so blood levels decrease precipitously on bypass. Note that the tissue solubilities of inhalational agents are increased at low body temperatures and they may cause residual cardiac depression effects after rewarming and during weaning from CPB. Discontinue inhalational agents during rewarming and supplement with IV anesthetics.
14. Hypertension in the adequately anesthetized child may be treated by injection of phentolamine (0.2 mg/kg). During hypothermic CPB, children respond to the cold-induced stress with increased levels of vasoconstricting catecholamines. Phentolamine, by its α -adrenergic blocking action, improves perfusion and delays the development of metabolic acidosis.
15. During bypass (partial and total), repeat the ACT every 30 min and give additional doses of heparin as necessary to maintain the ACT above 480 s.
16. Take blood samples for acid-base, electrolyte, and Hct determinations every 30 min and just before CPB is discontinued. Monitor glucose levels in small infants.
17. Before discontinuing CPB:
 - (a) Inflate the lungs, suction the trachea/bronchi, and check ventilation by observing the movement and full re-expansion of both lungs.

- (b) Commence pacing if the heart rate is slow or if sinus rhythm is absent; infants and children need an atrial contraction to maintain a good cardiac output at this stage. Atrial pacing can be used for slow heart rates with normal conduction. AV sequential pacing is required if conduction is abnormal. If AV block is present, it is usually possible to sense the atrial contraction and use it to pace the ventricle (AV sequential pacing).
- (c) If the cardiac action is impaired, inotropic agents should be commenced well before weaning. For this purpose, dopamine, milrinone, dobutamine and/or low dose epinephrine may be used in various combinations.
- (d) If the myocardial contractility is very severely depressed, start an infusion of epinephrine 0.01–0.1 µg/kg/min.
- (e) Be aware of the impaired calcium regulation in neonates and infants, especially when administering blood products. Hypotension during transfusions should be immediately treated by injections of calcium chloride. A calcium infusion may be required for small infants.

18. For children with PAH, consider the following before weaning:

- (a) A pulmonary artery line to monitor pressure and assess treatment
- (b) Mild hyperventilation with 100 % oxygen.
- (c) Maintain adequate levels of anesthesia and analgesia.
- (d) Correct preexisting metabolic acidosis
- (e) In children with mild increase of pulmonary artery pressure, a bolus of milrinone (50 µg/kg) followed by an infusion of 0.5 µg/kg/min may be useful before weaning from CPB.
- (f) Add nitric oxide to the inspired gases if necessary to control high PVR.

19. As CPB is discontinued:

- (a) Administer calcium chloride to improve cardiac action if necessary. (Calcium should never be given until the heart has resumed a steady regular rhythm).
- (b) Request infusion of blood from the pump, and infuse cells until the heart appears adequately filled. Monitor CVP and/or left atrial pressure. Depending on the heart defect and the repair, the initial post-bypass ventricular compliance can be quite poor or rapidly changing; the pre-bypass levels may no longer apply. The Hct during bypass is usually reduced; age, type of repair, cardiac function and the presence of bleeding will determine the need for additional transfusion of packed cells. After complex repairs in neonates and small infants, a Hct of 35–40 % should be the goal. Older children, especially those having more minor procedures, may be weaned at a lower Hct, given a diuretic (furosemide), and have the pump contents re-infused over the ensuing period. In this way it may be possible to prevent the need for blood transfusion.

20. If the child remains hypotensive despite a good rate and rhythm:
 - (a) Adjust the dopamine infusion (5–10 $\mu\text{g}/\text{kg}/\text{min}$). Larger dopamine doses are often required in infants compared with older children and adults.
 - (b) Dobutamine 5–10 $\mu\text{g}/\text{kg}/\text{min}$ may be added. This drug also increases inotropy, but it may also increase heart rate and decrease SVR in children.
 - (c) Calcium infusion may improve performance in some children, especially small infants. It is required in children with DiGeorge syndrome.
 - (d) If all else fails, an infusion of epinephrine 0.1–0.5 $\mu\text{g}/\text{kg}/\text{min}$ may be indicated.
21. Modified ultrafiltration may be employed after CPB has been discontinued to remove excess intravascular fluid and increase the Hct. The bypass circuit is used to withdraw blood from the aortic cannula, pass it through an ultrafiltration unit, and return it to the right atrium. In addition, it may clear some inflammatory mediators released during CPB. It has been suggested that modified ultrafiltration may reduce bleeding and enhance postoperative cardiopulmonary function.
22. When the child's condition is stable, give protamine slowly (preferably diluted for small infants). Common practice is to give a dose of protamine at least equal to that of the heparin (mg per 100 U/kg), which was administered before bypass. After 50 % of the protamine is given, the surgeons stop using the pump suction to protect the CPB circuit and switch to the wall suction. If hypotension occurs after protamine, it usually can be reversed by calcium and/or a fluid bolus. After protamine has been reversed and cardiac function and filling pressures reassessed, the cannulae are removed.
23. Two to 3 min after the administration of protamine blood is sampled for an ACT, electrolytes, and blood gases. Give more protamine and repeat the ACT if indicated.
24. If bleeding persists, check coagulation indices; give platelets (1 Unit/5 kg), cryoprecipitate, and/or other coagulation factors (i.e., recombinant factor VII etc.) according to the coagulation results.

N.B. Anticipate continued bleeding because of platelet dysfunction and other factor deficiencies:

 - (a) After a long pump run
 - (b) In children with cyanotic CHD
 - (c) In small infants, in whom the pump-priming volume is very large in relation to blood volume
 - (d) After a complex re-do sternotomy

All bleeding must be well controlled before the chest is closed.

25. After some complex intracardiac repairs, sternal closure may be delayed, and a plastic membrane sewn in place to cover the heart in the interim. This is appropriate when myocardial edema is present, as leaving the chest open prevents the constricting effects of the closed sternum on cardiopulmonary function and may increase BP and urine output while decreasing CVP. It also allows for easy access to the heart in case of tamponade or the need for emergency cannulation for ECMO. The sternum is closed when myocardial function has improved and the child is able to tolerate this maneuver.
26. At the end of surgery, the decision must be made whether to extubate the trachea immediately or to continue with ventilatory support. The decision depends on the disease that was present and the intraoperative course.
27. The trend toward “fast-tracking” some children after cardiac surgery is now well-established; it may be very appropriate with minimally invasive surgical procedures. After simple operations (i.e., closure of ASD, resection of subaortic membrane), the trachea may be extubated in the OR and the child may be expected to have a very short stay in the ICU. In such cases:
 - (a) Employ an anesthesia technique that permits early brisk recovery; avoid large doses of opioids or long-acting relaxants.
 - (b) Plan for good postoperative analgesia; a single-shot caudal morphine injection is safe, effective, and easy (see Chap. 5).
 - (c) Ensure that excess pump-priming fluid is removed by means of modified ultrafiltration after CPB is discontinued.

It may be more appropriate for some children to be transferred to the ICU for extubation on arrival or soon thereafter, depending on local circumstances.

- Some infants with large left-to-right shunts are at extreme risk of pulmonary hypertensive crises during and after surgery (see Principles of Anesthetic Management, Chap. 14, page 393)
- (d) Milrinone infusion 0.5–0.75 µg/kg/min if hemodynamically tolerated. It is most important to maintain adequate coronary perfusion pressures for the “pressure loaded” right ventricle.
 - (e) Inhalation of nitric oxide (NO). NO has specific pulmonary vasodilating properties and is a very useful drug to control PVR. It is usually added to the inspired gas (as close to the tracheal tube as possible) in a concentration of 20–80 ppm, but does require special equipment and monitoring. When mixed with oxygen, nitrogen dioxide (NO_2) is formed, which is damaging to the lungs. Hence NO is added to the breathing circuit as close to the child as possible, FIO_2 should be limited to that needed to ensure an acceptable saturation level. Large doses can also lead to methemoglobinemia. A sudden withdrawal of NO can cause severe rebound PAH, therefore the child has to be weaned slowly from it.

28. Many children after CPB require postoperative ventilatory support.

(a) These include:

- Those with hypoxemia despite a high FIO₂
- Low cardiac output
- Pulmonary hypertension
- Diminished lung compliance
- Persistent arrhythmias
- Hypothermia (<34 °C)
- Continuing hemorrhage

(b) In such children:

- Plan for continuing IPPV and/or CPAP or pressure support ventilation. Controlled ventilation and CPAP are particularly beneficial during the immediate postoperative period; at this stage, the child predictably has a tendency toward reduced lung volumes and increased lung water (especially the infant). IPPV also permits excellent pain control by opioid infusion
- Do not antagonize the muscle relaxants.
- The choice of a nasal versus an oral tracheal tube depends on institutional preference and the age of the child; for infants we usually insert nasal tubes at induction of anesthesia, especially if the child may require postoperative ventilation. Infants tolerate nasal tubes extremely well; they are less likely to kink and cannot be occluded by the teeth. They are also easier to secure at a specific depth and less likely to be pulled out during removal of the TEE probe under the drapes. It has been suggested that nasal tubes might predispose to middle ear or sinus disease; this has not proved to be a major problem. We avoid changing tracheal tubes from oral to nasal at the end of the surgery to preclude bleeding from the nasopharynx or if the infant's condition is unstable. Older children are readily managed with oral endotracheal tubes.

29. During transport to the ICU (all children):

- (a) Attach a full bag of blood (or other appropriate fluid) to the IV line to ensure immediate replacement volume availability in case of sudden hemorrhage.
- (b) Cover the child with warm blankets.
- (c) Administer O₂ by mask or, if the trachea is still intubated, continue controlled ventilation with O₂. Make sure that the tank has an adequate supply of O₂ for the transfer to ICU.
- (d) A battery-powered monitor should provide:
 - Pulse oximetry
 - ECG

- Intravascular pressures
 - EtCO₂ monitoring is desirable.
- (e) Continue the infusions of inotropic drugs and/or vasodilators using battery-powered pumps. If NO is used, this must be continued during transport and in the ICU. Beware of interruptions or changes to the flow of any of these drugs during transport to the ICU. Raising or lowering an infusion pump may alter drug delivery rates. Kinking of infusion lines may interrupt flow and result in a bolus dose as the kink is relieved.

Deep Hypothermia with Circulatory Arrest

Deep hypothermia with circulatory arrest (DHCA) is used for some neonates and infants undergoing cardiac surgery. It is particularly advantageous when surgery involves the aortic root.

Hypothermia is achieved by means of bloodstream cooling on CPB. The debate concerning the safety of profoundly hypothermic circulatory arrest versus continued perfusion is ongoing; many infants have been successfully managed by DHCA and have shown little evidence of cerebral impairment as they grow to adulthood. However, with the extension of cardiac surgery to more complex repairs in smaller infants many centers now prefer to limit the duration of DHCA and possibly use antegrade cerebral perfusion (ACP) during aortic arch reconstruction procedures.

Anesthesia Management

Anesthesia management is as described previously, with the following modifications:

1. Dextrose-containing solutions are best avoided because hyperglycemia may increase the risk of cerebral damage during total circulatory arrest. However, the blood glucose concentration should be monitored to detect and treat hypoglycemia should it occur. Large doses of fentanyl (more than 50 µg/kg) are preferred and may limit the increase in blood glucose concentration that occurs as a metabolic response during hypothermic CPB.
2. Give methylprednisolone (Solu-Medrol) 15–30 mg/kg IV slowly, before cooling on CPB. Ensure that the child is given adequate doses of relaxant drugs. (Once circulatory arrest has occurred, no additional drugs can be given.)
3. Phenytoin (Dilantin) 5 mg/kg may be added to the CPB prime solution as a neuroprotective agent.
4. After CPB is begun, ensure that the difference between the esophageal temperature and the temperature of the pump's output does not exceed 10 °C. Set cooling mattresses to 10 °C. Turn the room temperature down. Pack the head in ice bags.

5. The optimal management of blood gas tensions and acid-base balance during profound hypothermia has been the subject of much debate.
 - (a) The “alpha stat” approach measures blood gases at 37 °C whatever the child’s body temperature (i.e., pH alkalotic when corrected for the actual body temperature) has been widely used in adults. Blood gas analysis shows a normal or low PaCO₂ during cooling.
 - (b) The “pH stat” approach corrects the blood gases for the child’s body temperature and requires that CO₂ be added to the oxygenator gases during cooling. This has the advantage of increasing CBF, allowing for more even cooling and better oxygen delivery, and thereby improved neurologic outcomes in infants. pH stat is now the recommended strategy for pediatric patients.
6. Administer phentolamine 0.2 mg/kg to improve tissue perfusion, ensure rapid even cooling, and minimize acidosis on rewarming.
7. When the esophageal temperature is 16 °C and the rectal temperature is <20 °C, CPB is discontinued, blood is drained to the oxygenator, and the venous cannulas are removed.
8. Record the duration of circulatory arrest. The duration of safe circulatory arrest at a given temperature is unknown, but it is generally preferred to limit it to 20–30 min at 15–18 °C core temperature.
9. Keep the lungs slightly inflated at 5 cm H₂O with an air/O₂ mixture; ventilator off.
10. When the repair is complete, the venous cannulas are replaced and the child is rewarmed until the esophageal temperature reaches 37 °C. The temperature of the blood from the pump should never exceed 39 °C, and the child’s temperature should not exceed 37 °C.
11. Do not correct the metabolic acidosis often seen during rewarming. It will spontaneously correct as the child’s metabolism resumes. Administration of sodium bicarbonate usually results in postoperative metabolic alkalosis.
12. Maintaining Hct levels at 30% during CPB cooling and rewarming may be preferable to a reduced HCT and may result in superior postoperative neurologic function.

PRINCIPLES OF POSTOPERATIVE CARE

Respiratory System

The status of the respiratory system after cardiac surgery in infants and children may be determined by the following factors:

1. Preexisting status
 - (a) Immaturity of respiratory system in young children (especially infants)
 - (b) Effects of the cardiac disease on the lungs

2. Effects of anesthesia, operation, and CPB on the respiratory system
 - (a) Decreased lung volume
 - (b) Increased lung water
3. Intraoperative events:
 - (a) atelectasis (esp. right upper lobe).
 - (b) phrenic nerve injury.
 - (c) pleural effusions

Many children benefit from a period of controlled ventilation and/or PEEP or CPAP. This assists in restoring the lung volume to normal and improves gas exchange. Levels of added O₂ and PEEP or CPAP can be reduced as the pulmonary status improves. Diuretic therapy may be indicated to reduce lung water. Special measures to control PVR may be required in children with PAH.

Cardiovascular System

After cardiac surgery, the cardiovascular status is determined by:

1. Preexisting status
 - (a) Immaturity of the heart and circulatory system in infants
 - (b) Effects of the cardiac disease on the cardiovascular system
2. Effects of anesthesia, surgery, and CPB, which are dictated by:
 - (a) Duration of anesthesia and surgery
 - (b) Duration of CPB
 - (c) Duration of induced cardiac arrest
 - (d) The success of myocardial protection measures.

After all but the most minor cardiac operations, cardiac function may deteriorate postoperatively. This deterioration progresses for the first few hours after surgery, probably associated with edema of the myocardium and other changes that decrease the compliance of the ventricles and reduce contractility. Treatment at this time must be directed towards:

1. Ensuring optimal filling pressures. Because the compliance of the ventricles is reduced in infancy and reduced in all children after cardiac surgery, increased filling pressures (i.e., 8–12 mmHg) may be required.
2. Producing an optimal cardiac rate and rhythm. This is most effectively achieved by the use of sequential pacing when necessary. Sinus rhythm (i.e., atrial contraction) significantly augments cardiac output.

3. Reducing afterload. The use of vasodilators in children with ventricular dysfunction increases cardiac output with little change in cardiac work or arterial BP. When vasodilators are used, the preload must be maintained by infusion of appropriate fluids; SNP infusion is commonly used to produce vasodilation (start at 0.5–2 $\mu\text{g}/\text{kg}/\text{min}$ and increase to 5 $\mu\text{g}/\text{kg}/\text{min}$). Alternatively, milrinone may be used to provide afterload reduction. In some units, phenoxybenzamine is administered to produce a long-lasting adrenergic blockade. Caution: some children do not tolerate LV afterload reduction; e.g., those with impaired RV function (TOF, etc.).
4. Inotropic agents: If a low cardiac output persists despite these measures, resorting to an inotropic agent becomes necessary:
 - (a) Dopamine is infused at 5–10 $\mu\text{g}/\text{kg}/\text{min}$ by infusion pump. In infants and children, dopamine has been shown effective in increasing cardiac output, *but*:
 - Larger doses are required than in adults.
 - The vasodilating effect is less than in adults. Hence the concurrent infusion of a vasodilator (SNP or milrinone) is usually warranted. The combined administration of dopamine and SNP may also be effective in reducing PVR in children with PAH, but it must often be given as soon as the child is weaned from bypass.
 - (b) Calcium is infused as necessary to maintain the serum Ca^{++} at a high-normal level (1–1.2 mmol/dL).
 - (c) If serious low output persists despite these measures, an epinephrine infusion 0.05–0.1 $\mu\text{g}/\text{kg}/\text{min}$ may rarely be needed.

Fluid and Electrolyte Therapy

1. Blood should be administered to maintain the hemoglobin level at near-normal levels (14–15 g/dL), especially when cardiac dysfunction is present.
2. Acid-base status should be monitored and acidosis corrected by sodium bicarbonate infusions.
3. Balanced salt solutions (i.e., lactated Ringer's solution) should be infused at rates sufficient to maintain urine output and CVP indices.
4. KCl 2 mEq/kg/day may be added, *provided* a urine output greater than 1 mL/kg/h.
5. Hypomagnesemia may occur perioperatively, especially with aggressive diuretic therapy. Magnesium sulfate 1 mEq/kg/day may be added to the intravenous fluid regimen if necessary.
6. If urine output decreases to less than 1 mL/kg/h in the absence of hypotension, fluid orders should be reviewed to ensure an adequate intake, and a “fluid challenge” may be administered. If there is no result, a diuretic may be ordered (furosemide, 1–2 mg/kg IV).

Postoperative Management in the Intensive Care Unit

1. Auscultate the chest to ensure that ventilation is adequate when switching ventilation to the ICU ventilator. Order a suitable FIO₂ concentration, and confirm ventilation and oxygenation by blood gas determination as soon as possible.
2. Ensure good analgesia; if regional analgesia has not been provided, order suitable opioids and sedative drugs:
 - (a) Morphine may be given intravenously as a continuous infusion (10–30 µg/kg/h for children, 5–15 µg/kg/h for infants) or less desirably, every 2 h.
 - (b) Midazolam infusion 60–120 µg/kg/h or 0.1 mg/kg IV every 2 h as needed.
3. Continue balanced salt solution for maintenance (with added KCL 2 mEq/kg/24 h provided that urine output is 1 mL/kg/h or more; otherwise withhold the KCL).
4. Check blood loss via the drainage tubes and instruct the nurses to replace this with volume and blood components as required.
5. A chest radiograph should be obtained. Examine it carefully for pneumothorax, hemothorax, and atelectasis, and to ensure that the tip of the tracheal tube is mid-tracheal. Check placement of all other indwelling lines; ensure that the tip of the CVP line is positioned at the level of the junction of the SVC and right atrium. (Cardiac perforation may complicate CVP line advanced to within the atrium.)
6. If bleeding persists order coagulation studies. Based on results, administer cryoprecipitate, platelets or other coagulation factors as indicated.

SPECIAL CONSIDERATIONS FOR SPECIFIC OPERATIONS

Ligation of Patent Ductus Arteriosus

A PDA typically presents as the premature baby who cannot be weaned off the ventilator, occasionally even with CHF and NEC; and the relatively asymptomatic older infant or child with a classical continuous murmur.

Older Infants and Children

In older infants and children, a persistent PDA is treated either by insertion of an occluding device in the catheterization lab (see page 440) or by ligation during a video assisted thoracotomy (VATS—see page 331). Exceptions may be made for the very large ductus in a small infant.

1. PDA as the sole lesion in older infants and children usually presents few problems. Ligation or device occlusion is necessary to prevent potential later complications (i.e., PAH and cardiac failure, SBE, or aneurysm).
2. If VATS is planned, prepare for the possibility that a thoracotomy may be necessary to manage bleeding. Always ensure that a reliable large-bore-IV route is available. Prophylactic antibiotics should be administered.
3. An arterial line is not essential. The BP must be monitored in the right arm; in the unlikely event of bleeding from the ductus, it may be necessary to clamp the left subclavian artery. Monitor SaO₂ in the right hand and a foot.
4. After VATS, infiltration of the small incision sites with a local anesthetic provides good analgesia. However for a thoracotomy, intercostal nerve blocks or an ultrasound guided paravertebral block is effective; these children may go to the ward after PACU. In infants and small children, a single dose of caudal morphine is another option, but the analgesia may have delayed onset. These children should be admitted to the PICU.
5. During the procedure, monitor carefully:
 - (a) For bradycardia during dissection near the vagus nerve.
 - (b) For vital signs after ligation; normally the continuous murmur ceases and a soft systolic murmur remains. The BP (especially diastolic) may increase slightly at this time, but large changes are unusual.

Major changes in BP might suggest that the wrong vessel has been ligated or occluded. If the aorta has accidentally been occluded, hypertension and loss of the oximeter signal from the foot will occur. If a PA has accidentally been ligated, the continuous murmur of the ductus will remain. If a bronchus has been ligated, airway pressures will increase and the murmur will persist unchanged.
6. Blood loss is usually minimal, seldom necessitating transfusion, but it may be sudden and massive if a major vessel is torn: check that blood is immediately available in the OR.

Postoperative Care

1. Routine post-thoracotomy care should be applied. Good analgesia facilitates deep breathing exercises.
2. Rarely, the thoracic duct may be injured during PDA ligation. The resulting chylothorax may require drainage, and continued losses of chyle may impose a severe nutritional challenge.
3. Damage to the left recurrent laryngeal nerve in the region of the ductus is also a rare complication.

Preterm Infants

Persistence of the ductus arteriosus may occur in preterm infants, especially those weighing less than 1500 g. In addition to prematurity, respiratory distress syndrome, excessive fluid therapy, neonatal asphyxia, hypoxia, and acidosis predispose to this condition. PDA results in a large left-to-right shunt, with pulmonary vascular engorgement and CHF. Clinical signs include tachypnea, hepatomegaly, and “bounding pulses.”

Diagnosis is confirmed by auscultation of the typical murmur, radiographic evidence of increased vascularity, and ECHO findings of a large left atrium:aorta ratio. PDA may prevent weaning from ventilatory support of infants with RDS.

Treatment for PDA

1. *Medical treatment.* Administration of indomethacin, a prostaglandin inhibitor, 0.1–0.4 mg/kg IV daily for several days may induce closure of the PDA. Indomethacin may also cause renal damage and suppress bone marrow. Therefore, it is contraindicated in children with renal failure or coagulopathy. Very small infants (those weighing less than 1000 g) do not respond as well with closure of the PDA after indomethacin as do larger, more mature infants.
2. *Surgical treatment (Clipping).* This is necessary if indomethacin therapy fails or is contraindicated.

Special Anesthesia Problems

1. Observe all special precautions for the preterm infant.
2. Be prepared for sudden blood loss; the ductus is very thin and tears easily.
3. It may be preferable to operate on very small preterm infants in the neonatal ICU. This prevents transport problems, allows the infant to remain on the NICU ventilator (incl. HFOV), but requires that the anesthesiologist adapt routines to safely administer adequate anesthesia and analgesia in the ICU setting. Check that the pulse oximeter and ECG function well during electrocautery.
4. Preoperative management:
 - (a) Assess the infant carefully. Anemia, if present, may predispose to CHF. In such circumstances, a transfusion of packed red blood cells may improve the cardiac status. The increased Hct improves myocardial oxygenation and may also reduce the extent of left-to-right shunting.
 - (b) For those infants transferred to the OR, a heated transport incubator with a ventilator is required to maintain body temperature.
 - (c) Premedication is not required.

5. Perioperative Management

- (a) Ensure that the OR is heated ($>80^{\circ}\text{F}$) and all warming devices are in position before transferring the infant to the OR table.
- (b) Attach monitors. Monitor SaO_2 in a preductal site (right hand) and in the foot; measure the BP in the right arm (see previous discussion).
- (c) Tracheal intubation:
 - If the trachea is already intubated, ensure that the tube is firmly fixed, patent, and optimally positioned; otherwise, re-intubate with a new tube.
 - If the trachea is not intubated, give atropine 0.02 mg/kg, fentanyl 10 $\mu\text{g}/\text{kg}$, and rocuronium 1 mg/kg or vecuronium 0.1 mg/kg. Ventilate with 100% O_2 (use caution with high FIO_2 if large left to right shunting) and then intubate the trachea.
- (d) Our preference is to induce anesthesia with fentanyl 10–25 $\mu\text{g}/\text{kg}$ and atropine (0.01–0.02 mg/kg) and maintain anesthesia with the fentanyl \pm isoflurane 0.5–1.0 %.
- (e) Rocuronium (0.3 mg/kg) may be used to facilitate ventilation and prevent movement during maintenance.
- (f) Establish a large intravenous line. Blood loss is usually minimal but can be catastrophic if a vessel is torn.
- (g) Give minimal intraoperative fluids. These infants are usually adequately hydrated preoperatively and do not have third-space losses.
- (h) Manual ventilation is often useful as the ductus is approached. Many of these infants have congested lungs and poor compliance. To ensure adequate surgical exposure and dissection, a large part of the lung will be compressed, which often necessitates increasing the FIO_2 . Watch for surgical retraction compromising cardiac output and oxygenation; if the infant becomes hypotensive or suddenly desaturates despite the above maneuvers, ask the surgeons to remove the retractors.
 - Intercostal nerve blocks by the surgeons are encouraged.

6. Postoperative

- (a) Continued ventilation is necessary for most infants, with increased attention to respiratory care in view of possible post-thoracotomy complications (e.g., atelectasis).
- (b) Improvement in respiratory status after ligation of PDA is dictated by the relative contributions of pulmonary vascular congestion and pulmonary disease (RDS or bronchopulmonary dysplasia) to the preoperative status.

Division of Vascular Rings and Suspension of Anomalous Innominate Artery

Abnormalities of the great vessels may encircle or compress the trachea, bronchi, and esophagus. Typical examples are a *double aortic arch*, a vascular ring that is completed by a PDA or ligamentum arteriosum, or an *abnormal course of the subclavian artery*. Severe compression by vascular rings can lead to stridor and/or difficulty with feeding during early infancy.

The infant with a vascular ring often assumes a characteristic opisthotonic position. A chest radiograph with barium swallow can be a helpful diagnostic tool. Anomalous vessels may compress the bronchi and lead to gas trapping in an individual lobe of the lung, with compression of the adjacent lung by the resultant emphysematous lobe. In addition, dynamic CT and cardiac MRI are frequently used to delineate the anatomy and plan the surgical repair (aortopexy, simple ligation, or complex reconstruction). Infants with vascular compression are prone to sudden cardiorespiratory arrest (see also Chap. 13, Trachea-esophageal fistula).

Special Anesthesia Problems

1. Respiratory failure may exist.
 - (a) Chronic or recurrent respiratory infection may cause impaired pulmonary function.
 - (b) Vascular compression may cause emphysema of one or more lobes, compressing other lung tissue.
2. Airway compression may be at the level of the carina or main bronchi; if so, a normally positioned tracheal tube will not relieve the obstruction.
3. Tracheal intubation may be required preoperatively to relieve serious symptoms. Air trapping in a lobe as a result of vascular compression can often be alleviated by the application of PEEP.
4. The use of an esophageal stethoscope in infants with vascular rings has been reported to cause acute airway obstruction.
5. VATS may be used for the procedure (see page 331). Be prepared for thoracotomy if problems arise.

Anesthesia Management

Preoperative

1. Monitor carefully and order intensive respiratory care to achieve optimal pulmonary status. The infant should be allowed to remain in a position that permits optimal ventilation.
2. Bronchoscopy may be required to evaluate the site of airway compression and is useful for endobronchial suction.

Perioperative

1. For intubation, use a method that ensures a good airway past the obstructing lesion. If the obstruction is mid-trachea, this is relatively simple. If the obstruction is low or at the carina, do one of the following:
 - (a) Pass a long tracheal tube past the obstruction into a main bronchus. Ensure a tube with a side hole (e.g. Murphy eye) is present to ventilate the other bronchus. Or:
 - (b) Ventilate the lungs via a rigid bronchoscope (which can be placed accurately under direct vision and adjusted intraoperatively if necessary).
2. Monitor the BP via an arterial line placed in the right radial artery. Operations on the great vessels can cause serious bleeding; establish a reliable, large-bore IV access.
3. For aortopexy, a bronchoscope may be used to ensure that the compression has been relieved. Compression of the trachea should always be assessed during spontaneous ventilation and coughing; if controlled ventilation is applied, the trachea is held open and always appears widely patent.
4. Use general anesthesia, maintain spontaneous ventilation, and spray the larynx with lidocaine before inserting the bronchoscope. During the remainder of the operation, ventilation can be assisted as necessary. Do not give relaxants.
5. Complex tracheobronchial reconstructions are occasionally performed on CPB.

Postoperative

1. Order constant care with added humidified oxygen for at least the first 24 h.
2. If residual obstruction persists, continue with tracheal intubation for 24 h, then reassess.
3. Partial obstruction may be improved by placing a small bolster, 1–2 in. thick, below the shoulders.
4. Racemic epinephrine and/or dexamethasone may be required for post-instrumentation croup.
5. After complex tracheobronchial reconstructions the children are usually immobilized for several days in a specific head-neck position to avoid tension on the sutures.

Resection of Aortic Coarctation

In the past, aortic coarctation (CoA) was classified according to its site in relation to the ductus arteriosus (i.e., preductal, juxtaductal, or postductal); all coarctations are now regarded as juxtaductal. However, it is useful to recognize the two distinct presentations of CoA; in infancy with CHF or in later life with hypertension. The infantile type is frequently accompanied by other anomalies

(e.g., VSD, PDA) and manifests as cardiac failure in an infant <6 months of age. In later life, CoA is usually diagnosed during investigation of hypertension in the upper limbs in preschoolers.

CoA can be treated medically by balloon angioplasty or surgically by resection. Controversy continues as to the optimal care, but there is some agreement that re-coarctation is best treated by angioplasty and that long segment coarctation is best treated surgically. General anesthesia may be required for balloon angioplasty to ensure immobility (see Interventional Cardiology, page 440).

Coarctation of the aorta is now recognized as a lifelong disorder; later in life many children will have continuing problems with hypertension and its complications. This is thought to be because of alterations in the renin angiotensin system or baroreceptors, and is more likely in those with a long history of hypertension preoperatively.

Preductal (Infantile Type)

Special Problems

1. Most infants have severe cardiac failure and are treated with inotropic support and diuretics. Assisted ventilation is often required.
2. Severe associated cardiovascular anomalies are common.
3. Blood flow to the lower portion of the body depends on the ductus arteriosus. Prostaglandin (0.05–0.15 µg/kg/min) is infused to maintain patency until surgical repair is performed.
4. Hypoplasia of the arch of the aorta may be present and, if significant, may require repair with CPB (see later discussion).

Anesthesia Management

Preoperative

1. Blood gases should be determined and abnormalities corrected. Maintain prostaglandin infusion.
2. Ensure that supportive drugs are immediately available (i.e., epinephrine, dopamine).
3. Ensure that an adequate volume of blood is available in the OR and checked before incision.

Perioperative

1. Passive cooling to 34 °C is commonly performed for neuroprotection.
2. While the aorta is clamped, do not allow the systolic pressure to exceed 100 mmHg. In the (rare) event that a drug is necessary to reduce the BP, administer small concentrations of isoflurane and titrate this against the BP.

A small dose of heparin (1 mg/kg) usually is given before clamping or balloon angioplasty. Check that the ACT has been prolonged to 250 s.

3. Be prepared to support the circulation when the aortic obstruction is removed; infusion of fluid and/or cardiotonic drugs may be required.
4. Prostaglandin may be discontinued once the ductus is ligated and the CoA relieved.

Postoperative

The course is sometimes stormy. Therefore:

1. Transfer the child to the ICU with a tracheal tube in place.
2. Controlled ventilation is usually necessary for at least 48–72 h.

Postductal (Adult Type)

Special Considerations for Surgical Repair

1. Clamping the aorta could compromise the blood supply to the spinal cord; hence the need to maintain an optimal mean BP in the distal aorta (~45 mmHg).
2. While the aorta is clamped, severe proximal hypertension may occur and require treatment.
3. Hypertension can be troublesome postoperatively; this may be controlled by the use of short acting β -blockers during the perioperative period, but it may also require the use of SNP.
4. Although blood must be available for transfusion, few children require it. (Bleeding from chest wall collateral vessels is much less profuse than in adults.)
5. In the very rare event that severe proximal hypertension cannot be controlled after aortic clamping or if the distal pressure is very low, a temporary shunt must be placed to bypass the site of anastomosis, or left heart bypass must be used.

Anesthesia Management

Preoperative

1. Monitor BP and SaO_2 in the right arm. Place a second oximeter probe on a foot.
2. Establish reliable intravenous infusions at sites other than in the left arm.
3. Epidural/caudal catheter may be placed to administer morphine and/or bupivacaine for postoperative pain management, although this may raise medicolegal concerns if paraplegia occurs.

Perioperative

1. Maintain anesthesia with N₂O and isoflurane; use rocuronium or vecuronium for relaxation.
2. Place an arterial line into the right radial artery, and a central venous line; the latter may be used to infuse drugs as necessary to control hypertension.
3. During the period of aortic clamping, control the BP if necessary, by increasing the inspired concentration of isoflurane. The BP should not be allowed to exceed 140 mmHg, but do not attempt to reduce the pressure if it remains below that level. Distal aortic pressure during clamp-off varies directly with the proximal pressure; distal pressure should be maintained at or above a mean of 45 mmHg to ensure perfusion of the spinal cord.
4. The surgeon first removes the distal clamp and then (slowly) the proximal clamp. Monitor the BP continuously. If hypotension develops, infuse fluids; if this is unsuccessful, ask the surgeon to partly reapply the proximal clamp briefly. The BP may remain slightly reduced for a while, but usually it is back to above-normal levels by the end of the operation. Anticipate the need to treat postoperative hypertension by preparing:
 - (a) SNP—1–10 µg/kg/min to control hypertension
 - (b) Esmolol—a loading dose 100–500 µg/kg over 1 min followed by an infusion of 50–100 µg/kg/min to control hypertension.
5. Blood loss is usually minimal and transfusion unnecessary.
6. In most cases, controlled ventilation is not required postoperatively and the trachea is extubated at the end of the operation.

Postoperative

1. The child should be monitored continuously in the ICU, special attention being paid to signs of blood loss. Measure the chest drainage and observe the clinical indices.
2. Hypertension usually persists for several days postoperatively; if severe, it may necessitate therapy with SNP and/or esmolol (see previous discussion). Prevention of hypertension is essential to prevent arteritis (see later discussion).
3. Very rarely, the postoperative course is complicated by post-coarctectomy syndrome; intestinal ileus caused by mesenteric arteritis secondary to increased pulsatile flow in the mesenteric artery. In extreme cases, bowel resection may be required.
4. Other serious postoperative complications include recurrent laryngeal nerve palsy, phrenic nerve palsy, chylothorax, and paraplegia because of spinal cord ischemia during repair (very rare).

5. Re-coarctation may occur in later years, particularly after repair in infancy. Repeat operation may be technically much more difficult and may involve major blood losses. Hence balloon angioplasty is now usually preferred.

Interrupted Aortic Arch

This lesion, which is frequently associated with VSD and DiGeorge syndrome, requires repair using CPB with profound hypothermic circulatory arrest. The patency of the ductus arteriosus must be maintained with a prostaglandin infusion until operation.

Palliative Surgery to Increase Pulmonary Blood Flow

The operations that may be used include the following:

1. Blalock-Taussig procedure (systemic artery anastomosed to the PA). A modified Blalock procedure using a synthetic graft between the aorta or innominate artery and the central portion of the PA is now the most commonly performed procedure.
2. The Potts operation (PA anastomosed to descending aorta) and the Waterston procedure (PA anastomosed to ascending aorta) are now rarely performed because they tend to become too large and may also cause unilateral pulmonary edema.

The operation is indicated in infants and children with TOF, tricuspid atresia, and other conditions in whom right-sided cardiac lesions result in decreased pulmonary blood flow. They are usually performed during infancy to increase pulmonary blood flow and stimulate growth of the PAs; they may then be followed by total correction of the defect at an older age. The performance of total repair of CHD in infancy has resulted in much less use of these shunting procedures.

Special Anesthesia Problems

1. Many of these children are severely hypoxemic and polycythemic.
2. During surgery, the PA is partly occluded so that the anastomosis can be completed; this causes a further temporary decrease in pulmonary blood flow.
3. Children with polycythemia may have a coagulation defect, although this is rarely a problem.
4. In small infants, the narrow lumen of the new shunt is prone to thrombosis. This may be prevented by using a small dose of heparin and appropriate fluid therapy.

Anesthesia Management

Preoperative

1. These are often urgent or semi-urgent surgeries to increase pulmonary blood flow after failure to stent the PDA or dilate a stenotic or atretic pulmonary valve in the cardiac catheterization laboratory.
2. Ensure that the respiratory system is optimal, with no active infection or recent history of URTI.
3. If the child is taking β -blocking agents, they should be continued to avoid rebound reactions.
4. Allow liberal fluids up to 2 h before operation. Otherwise, order intravenous maintenance fluids during any prolonged preoperative fasting period for children with an Hb greater than 16 g/dL.

Perioperative

1. Follow routine management, as on page 399.
2. Induce anesthesia intravenously (most of these children have a right-to-left shunt); a small dose of ketamine or propofol followed by fentanyl and rocuronium is preferred.
3. An arterial line should be placed either in the radial artery—*in the rare event that the subclavian artery will be used, place the line in the opposite side*—or in the femoral artery.
4. Maintain anesthesia with sevoflurane, isoflurane, or halothane in at least 50% O₂ in air.
 - (a) If halothane is available, it is useful for children with TOF because it decreases the RV contractility and outflow obstruction. Otherwise sevoflurane may be used.
 - (b) If the child with TOF desaturates, give esmolol (500 μ g/kg IV slowly) and/or small doses of phenylephrine (1–10 μ g/kg).
 - (c) If excessive hypotension occurs discontinue the volatile agent and administer fentanyl.
5. Immediately before the PA is clamped, switch to 100% O₂, inflate the lungs well, and give a further dose of relaxant.
6. Once the clamps are in place and if oxygenation is stable, allow the surgeon to proceed with the anastomosis. (Once this is commenced, the PA will be open and the clamps cannot come off until the anastomosis is completed.)
7. During creation of the anastomosis, if the systemic BP decreases profoundly or bradycardia occurs, administer cardiotonic drugs (i.e., epinephrine 1–5 μ g/kg, calcium chloride 10 mg/kg) until the anastomosis is completed and the clamps are removed.

8. A “modified” Blalock anastomosis is usually performed using a synthetic graft. In small infants, it is usual to give a small dose of heparin (100 U/kg) to prevent thrombosis of the shunt. A bolus of fluid after the clamps are released may enhance flow through the shunt.
9. Throughout the surgery, give 5% albumin as necessary to replace blood loss and decrease the Hct.

Postoperative

1. Auscultate the chest to document that a new murmur is present (this indicates that the shunt is functioning).
2. If the anastomosis is small and considered likely to be blocked by thrombosis, it is usual to order heparin in suitable dosage for several days postoperatively and start a small daily oral dose of an anti-platelet medication as soon as possible.

Palliative Surgery to Decrease Pulmonary Blood Flow

PA banding is performed to diminish blood flow to the lungs in infants who have a large left-to-right shunt, thereby improving systemic perfusion, decreasing pulmonary vascular congestion, and averting the development of fixed PAH. This is usually performed as an emergency procedure in the neonate who has significant other congenital abnormalities and/or is unable to tolerate a surgical repair on CPB. Many children present with CHF.

Anesthesia Management

Preoperative

1. Follow routine management, as on page 399.
2. Apply all measures possible to improve the infants’ general status before surgery (e.g., IPPV for several hours can be very beneficial) including treating any CHF.

Perioperative

1. Avoid myocardial depressants.
2. Monitor the infant closely as the band is applied. With an optimal band tightness:
 - (a) The systemic BP should increase, and the saturation may decrease slightly.
 - (b) The distal PA pressure should decrease to approximately 30–50 % of systemic pressure.
 - (c) The PetCO₂ value may decrease very slightly. (A large decrease indicates that the band is too tight.) This change in PetCO₂ is a most reliable guide to optimal PA banding.

Postoperative

1. Controlled ventilation may be required for several days.

SPECIFIC OPEN-HEART PROCEDURES

The considerations discussed here are a supplement to all the other important general principles outlined previously.

Atrial Septal Defect (Secundum Type)

The Secundum-type ASD is the most common form of an atrial septal defect and located in the fossa ovalis. Simple small ASDs are now commonly closed in the cardiac catheter laboratory using an “Umbrella” device (see page 440). Large ASD requires surgical closure, though this may be performed using minimally invasive techniques (i.e., small incisions). Other forms of (ASD) may be associated a cleft in the mitral valve (Primum ASD) or with partial anomalous pulmonary venous drainage (Sinus venosus type, most often located at the junction of the right atrium with the SVC), in which case a baffle is required to redirect this flow to the left atrium. This may complicate and prolong the simple ASD closure procedure slightly.

1. For simple secundum ASDs in healthy patients, plan to extubate the trachea at the end of the procedure. Many different “fast-track” techniques have been described, depending on the age and general condition of the patient. A single dose of caudal morphine (33 µg/kg of Duramorph) given after induction of anesthesia before surgery), can provide early postoperative analgesia and last up to 24 h but may require admission to PICU overnight.
2. Surgical closure of the ASD is performed either with cardioplegia or with induced ventricular fibrillation, so as to prevent the possibility that air may enter the LV and be pumped into the circulation. Ensure that the child is completely paralyzed during CPB to prevent the possibility that the child takes a breath while the atrium is open, which could draw air into the left side of the heart; give an additional dose of relaxant as the heart is being cannulated.
3. As the last suture is being tightened to close the defect, the surgeon may request sustained inflation of the lungs to promote flow of blood via the pulmonary veins into the left atrium to remove any residual air from the left side of the heart.
4. Bypass is short and post-CPB inotropic therapy is unlikely to be needed.

Total Anomalous Pulmonary Venous Drainage

The pulmonary veins drain into the right atrium or its venous connections. There are three common types:

1. *Supracardiac* (50% of cases): Pulmonary veins drain into the left SVC.
2. *Cardiac* (30% of cases): Pulmonary veins drain into the coronary sinus or right atrium.
3. *Infracardiac* (10% of cases): Pulmonary veins drain into the inferior vena cava via a common trunk below the diaphragm.

Total anomalous pulmonary venous drainage (TAPVD) manifests in early neonatal life with severe cyanosis and acidosis. Obstruction of the pulmonary veins may be present and may cause pulmonary edema and cardiac failure. Survival depends on a right-to-left shunt, usually via an ASD or patent foramen ovale (PFO), which may need enlarging by balloon dilation in the cath lab. The presence of obstruction to the pulmonary veins exacerbates the symptoms and if severe, may dictate emergency surgery.

Special Considerations

1. If the pulmonary veins are obstructed, pulmonary edema, PAH, and cardiac failure may be present.
2. Preoperative ventilation may be required to treat pulmonary edema and improve oxygenation.
3. The left atrium and LV may be small and the LV compliance reduced; aggressive inotropic therapy may be required after bypass.
4. The pulmonary vasculature may have a thick medial layer, and PVR may remain increased after repair.

Anesthesia Plan

1. Maintain controlled ventilation and PEEP; monitor acid-base status frequently.
2. High-dose opioid technique is preferred.
3. After repair, the LV may require generous inotropic support and afterload reduction to maintain cardiac output. High left atrial pressure should be avoided as this may lead to fluid overload.
4. Active measures to reduce PVR and prevent pulmonary hypertensive crisis must be instituted. The PA pressure should be monitored. Prepare to administer NO if it becomes necessary.
5. TEE may be contraindicated as it may further obstruct the venous drainage and induce a pulmonary hypertensive crisis.

Ventricular Septal Defect

VSD is the most common single defect (20 % of CHD cases). The position of the VSD is used to classify the disease and also may predict complications:

1. *Type I (supracrystal, 5 %)*: under the annulus of the aorta; may affect the adjacent cusp and cause aortic valve incompetence; also associated with narrow aortic isthmus
2. *Type II (infracrystal, most common, 80 %)*: in the membranous septum—often large with a big shunt
3. *Type III (AV canal type, 11 %)*: beneath the tricuspid valve
4. *Type IV (muscular 4 %)*: may be multiple (“Swiss cheese” defect)

The physiologic effects of the VSD depend on its size. If it is large (nonrestrictive), the left-to-right shunt is similarly large, resulting in early CHF and subsequent pulmonary vascular obstructive disease (PVOD). Small defects (restrictive) allow only a limited left-to-right shunting, which may be physiologically insignificant. Early operation is performed for large defects to prevent the onset of PVOD. Hybrid techniques (combined surgical and imaging) may be used to close small defects.

Special Considerations

1. Infants with severe CHF may benefit from intubation and ventilation preoperatively.
2. Infants with CHF cannot tolerate myocardial depressant drugs (e.g., halothane).
3. Postoperative conduction disturbances, which may be temporary (possibly due to edema around a suture), should be anticipated.
4. Pulmonary vascular crisis may occur postoperatively in small infants with previously large left-right shunts.

Anesthesia Plan

1. Avoid myocardial depressants; high-dose opioids are preferred. Titrate drugs slowly.
2. Bidirectional shunts are common; be careful to prevent air bubbles in IV lines.
3. Maintain PVR to prevent increasing left-to-right shunting; prevent hyperventilation. Minor reductions in SVR (perhaps minimal isoflurane) may be beneficial.
4. Be prepared to institute pacing if conduction is abnormal after repair.
5. In the case of large defects, post-bypass therapy to reduce PVR and to prevent pulmonary vascular crisis may be required.

Atrioventricular Canal

Atrioventricular (AV) canal is frequently associated with Down syndrome and may be complete (ASD, VSD, and cleft AV valve) or partial (ostium primum ASD plus cleft mitral valve). The most significant hemodynamic changes are a large left-to-right shunt, leading to PAH, and mitral incompetence. Early surgical repair in infancy is preferred.

Special Considerations

1. Down syndrome is often present (see Chap. 6).
2. PAH may be a problem postoperatively; prepare a transducer to measure PA pressure and institute therapy. Aggressive inotropic therapy may be required.
3. Disturbances of conduction are relatively common after repair and may persist; chronic pacemaker therapy may be required.
4. Mitral valve malfunction may occur in the early or late postoperative period, and reoperation to repair the valve may be required.

Tetralogy of Fallot

The clinical picture results from the large VSD in the presence of RV outflow obstruction, which together produce a large right-to-left shunt; the other features of TOF are overriding of the aorta and RV hypertrophy. RV obstruction may be infundibular (50%), valvular (10%), PA (10%), or combined (30%). Acute dynamic increases in infundibular obstruction may result in severe desaturation episodes (“tet” spells). Most children are now treated by complete repair in infancy, except those with small PAs, who are treated initially with a systemic-to-pulmonary shunt.

Special Considerations

1. Adequate preoperative sedation and sufficient anesthesia and analgesia to suppress any response to surgical stimulation are important to prevent “tet” spells. Avoid drugs that reduce SVR significantly (e.g., isoflurane); high-dose opioid technique is preferred. Titrate the drug carefully.
2. Halothane in low concentrations may be useful to depress the muscle of the RV infundibulum and prevent desaturation due to increase shunting.
3. Otherwise, intraoperative desaturation should be treated with oxygen, fluid infusion, and esmolol 0.5 mg/kg slowly and/or phenylephrine 1–10 µg/kg IV to increase SVR.
4. After CPB, a high filling pressure may be required owing to the thickened, poorly compliant right ventricle. Rarely, conduction defects require AV pacing. Junctional ectopic tachycardiac may be a problem in the postoperative period.

and is usually treated with gentle cooling, sedation, reduction of inotropic support and antiarrhythmic therapy (procainamide or amiodarone)

5. The use of LV afterload reduction may be poorly tolerated because the RV output is the limiting factor on overall cardiac output. In neonates, a small intra-atrial communication (PFO or ASD) is often left open or created to allow for RV decompression and maintenance of LV filling.

Transposition of the Great Arteries

The most common cause of cyanotic CHD in the neonate is transposition of the great arteries, whereby the aorta arises from the RV and the PA arises from the LV. The pulmonary and systemic circulations are thus separate and in parallel; survival depends on mixing via the PFO, ASD, VSD, or PDA. Without treatment, 90% of infants with transposition of the great arteries die within 12 months. Current surgical treatment is to perform an arterial switch procedure; this must be performed in the neonatal period for infants with an intact septum but may be performed later in those with a large VSD and subpulmonary obstruction.

Special Considerations

1. The neonate with an intact septum, dependent on mixing via the PFO and PDA, will become desperately hypoxic when the latter closes. Balloon atrial septostomy (BAS) has been used to improve mixing, or, if the neonate is having early surgery, PGE₁ may be used to maintain the PDA.
2. The effective pulmonary or systemic blood flow is limited to that volume of blood that shunts between the circulations.

Anesthesia Plan

1. A technique that maintains myocardial function and cardiac output should be used; a high-dose opioid technique is preferred.
2. Moderate hyperventilation with a high FIO₂ may reduce PVR and thereby increase pulmonary blood flow, mixing, and arterial saturation.
3. PGE₁ infusion must be continued, although the PDA is usually ligated once on bypass (to prevent run-off during surgery).
4. Post-CPB measures to optimize blood flow in the re-implanted coronary arteries are required. An infusion of nitroglycerin (1 µg/kg/min) should be commenced before weaning from CPB.
5. Maintain an optimal preload; must achieve an adequate balance between coronary perfusion and afterload reduction for the deconditioned LV that was previously exposed only to the lower PVR. (Monitoring the EKG for ST changes is an attractive concept but in practice, is not very reliable!)

6. Bleeding from multiple suture lines is to be expected. Order blood components to correct the coagulopathy that is common after CPB in neonates. Platelet suspensions, plasma, and cryoprecipitate may be required.

Aortic Stenosis

Critical aortic stenosis can cause severe CHF in the neonate. Older children with aortic stenosis may be asymptomatic, but they are at increased risk for angina, syncope, and sudden death. Aortic stenosis may be subvalvular; valvular, usually with a bicuspid valve (80% of cases); or supravalvular (often associated with Williams syndrome [see Appendix A]). Critical aortic stenosis in the neonate is now usually treated by balloon dilation. On occasion an infant with a small aortic annulus or with associated cardiac lesions may require operation. The principles outlined below apply equally to those children who require anesthesia in the cardiac catheterization lab.

Special Considerations

1. Infants with critical aortic stenosis are hypotensive, poorly perfused, and aci-dotic, with respiratory distress and hepatomegaly. The disease often becomes apparent as the ductus arteriosus closes. The thickened LV is prone to isch-emia and arrhythmias. Subendocardial fibroelastosis may also be present. These infants require aggressive resuscitation and early operation; even so, the mortality rate is high.
2. Older children with valvular aortic stenosis commonly have a bicuspid valve, and cardiac function is usually quite good despite a high gradient across the stenosed valve.

Anesthesia Plan: Infants

1. The infant will be receiving an infusion of prostaglandin to maintain the ductus arteriosus; this must be continued.
2. A high-dose opioid and relaxant technique is preferred.
3. Maintain body temperature carefully.
4. Serious arrhythmias, including ventricular fibrillation, may occur as the heart is manipulated, especially if any cooling has occurred.

Anesthesia Plan: Older Children

1. The aim is to maintain the heart rate constant, prevent tachycardia or brady-cardia, and prevent any major decrease in SVR and aortic root pressure. Opioid plus rocuronium or vecuronium is usually satisfactory.

2. Halothane may be useful to reduce dynamic LV outflow tract obstruction in those with subvalvular aortic stenosis.
3. Children with supravalvular aortic stenosis and Williams syndrome may have a difficult airway.
4. Postoperative hypertension is common after aortic valvotomy and a combination of esmolol and SNP infusion may be required.

Hypoplastic Left Heart Syndrome

In HLHS, the LV and ascending aorta are hypoplastic and the LV is nonfunctional. Immediate survival depends on the pumping action of the RV and systemic flow via the PDA with retrograde flow in the aortic arch. Blood mixes in the right atrium (via an ASD or PFO), and pulmonary-to-systemic flow ratio depends on the size of the intra-atrial communication, the PVR, and the SVR. Treatment of this condition involves conversion to a univentricular series type of circulation by means of a staged repair (very rarely heart transplantation may be an alternative).

1. *Stage 1:* Norwood procedure. Division of the PA, connection of the RV to a reconstructed aortic arch (neoaorta made from the PA and AO), atrial septectomy, and a modified Blalock-Taussig shunt (Inominate or subclavian artery to PA). A modification of the Stage 1 Norwood procedure is to place the shunt from RV to PA; this may improve myocardial perfusion (Sano Modification).
2. *Stage 2:* Bidirectional Glenn (SVC-RPA) anastomosis or hemi-Fontan procedure (and take down of the B-T shunt). This procedure is designed to direct some of the blood flow directly to the lungs and so reduce the load on the RV.
3. *Stage 3:* Completion of the Fontan procedure, which connects the IVC to the PA by means of an intratrial lateral tunnel or extracardiac Gortex conduit, and places the systemic and pulmonary circulations in series. There is no subpulmonary pumping chamber; the conduit/tunnel is fenestrated to decompress high right-sided pressures into the circulation.

Special Considerations

1. Preoperative management is aimed at maintaining the ratio of pulmonary to systemic blood flow ($Qp:Qs$) close to 1. The ease with which this can be achieved depends first on the child's anatomy (i.e., the size of the interatrial communication).
 - (a) If it is large and unrestrictive, pulmonary blood flow is excessive and systemic hypoperfusion with metabolic acidosis occurs.

- (b) Children with relative restriction (a high percentage) may achieve Qp:Qs near 1 when breathing room air. These infants must be managed to maintain the status quo (i.e., to prevent changes in ventilation or oxygenation.)
 - (c) Children with a very small or absent ASD have left atrial and PAH, decreased pulmonary blood flow and are profoundly hypoxic at birth. These infants require an immediate intervention in the cath lab (balloon atrial septostomy) and early surgery.
2. All infants require a continuous infusion of PGE₁ to maintain patency of the ductus arteriosus.
 3. Some infants may require inotropic agents to increase cardiac output and systemic perfusion; these must be used with caution to prevent adverse changes in SVR.

Anesthesia Plan-Norwood Stage 1

1. Take great care to maintain FIO₂ (an oxygen blender) and ventilation (self-inflating bag) unchanged during transport to and from the OR. It is very easy to hyperventilate the lungs accidentally and cause a disastrous decrease in PVR and thus in systemic blood flow.
2. Care during the pre-bypass stage is similar for those undergoing either the Norwood procedure or transplantation. Carefully maintain the level of ventilation and oxygenation to balance the PVR:SVR ratio. The arterial saturation should be $\pm 80\%$.
3. High-dose opioid anesthesia is preferred, but fentanyl (50–75 $\mu\text{g}/\text{kg}$) must be titrated slowly, balancing surgical stimulation, to prevent hypotension.
4. Post-CPB after a Norwood procedure and measures to maintain the PVR and limit pulmonary blood flow may still be required, depending on the size of the shunt. Expect the saturation to be 70–80%.
5. Rarely, if pulmonary blood flow is inadequate and the infant is severely hypoxic, a larger shunt may be needed.

Fontan Procedure

In addition to the HLHS, the Fontan procedure and its modifications are nowadays also used to treat many other forms of CHD in which there is a single functional ventricle: The SVC is directly connected to the PA (Glenn procedure), and the venous return from the IVC is redirected to the PA via an intra-atrial lateral tunnel or an extra-cardiac conduit. This creates a serial circulation in which the all the kinetic energy for blood flow through the pulmonary and systemic vascular beds is provided by the single ventricle.

Special Considerations

1. After the operation, the total cardiac output is directed to the systemic circulation. The pulmonary circulation is driven by the systemic venous return; hence a low PVR is vital. There is no shunting, so there is improved SaO_2 and there is also a reduced load on the ventricle.
2. For a successful operation, PVR less than 4 Wood units/ m^2 is desirable and PVR must not increase. However, in some children with mildly increased PVR, a Fontan operation is performed but a fenestration is left in the vena cava-to-PA baffle. This allows for some right-to-left shunting and relieves the right atrial pressure; cyanosis may occur, but ventricular filling and cardiac output are maintained.
3. Pleural and pericardial effusions are very common after the Fontan procedure and may require drainage for a prolonged period.
4. Plastic bronchitis leading to acute airway obstruction and requiring bronchoscopic removal of a membrane is described in post-Fontan children. The production of this membrane is considered to result from the high venous pressures.

Anesthesia Plan

1. Establish several reliable intravenous routes before the operation; postoperative edema may make later venous access difficult.
2. A reliable CVP line is essential for postoperative monitoring. The surgeons will place a direct atrial line to monitor the transpulmonary gradient (SVC pressure—atrial pressure)
3. Before terminating bypass, ensure that sinus rhythm is present or institute sequential pacing. Sinus rhythm is essential. Hyperventilate using a short inspiratory phase, minimal peak inspiratory pressure, and PEEP just adequate to maintain optimal lung volume.
4. After bypass, maintain the CVP at 14–16 mmHg and atrial pressures of 4–8 mmHg to maintain pulmonary blood flow. The gradient across the pulmonary bed should be less than 10 mmHg.
5. Early return to spontaneous ventilation and extubation augment pulmonary blood flow.
6. Fluid retention with peripheral edema, pleural and pericardial effusions, and ascites is common. Protein-losing enteropathy may occur in some children.

Tricuspid Atresia

Tricuspid atresia is a condition in which there is no communication between the right atrium and the RV, which is usually hypoplastic. Survival depends on the presence of an adequate atrial communication (PFO or ASD) and a systemic-to-pulmonary

shunt (VSD or PDA). Palliation in the neonate is required, and the ASD may have to be enlarged by balloon septostomy. For those children with diminished pulmonary blood flow, a systemic-to-PA shunt (e.g., modified Blalock operation) is performed. Those with increased pulmonary blood flow, CHF, and systemic hypoperfusion need a PA band. Later in life, when the pulmonary vasculature is more developed, palliative procedures towards the single ventricle circulation (bidirectional Glenn, Fontan) are possible.

Truncus Arteriosus

Truncus arteriosus occurs when the pulmonary artery fails to completely separate from the aorta resulting in a communication between the two vessels. Several types have been described:

1. *Type I*: PA arises from truncus and then divides
2. *Type II*: separate PAs arise from posterior truncus
3. *Type III*: separate PAs arise from sides of truncus

The different types are further classified according to the presence (type A) or absence (type B) of an additional VSD.

Special Considerations

1. DiGeorge syndrome is present in 25 % of cases (see Appendix A). In such cases monitor the Ca⁺⁺ level. Immune deficiency is present: use Irradiated red blood cells.
2. Increased pulmonary blood flow predisposes to pulmonary vascular hypertensive crises.
3. Low aortic diastolic pressure may result in inadequate coronary flow and myocardial ischemia.
4. The truncal valve is semilunar and commonly is incompetent.

Anesthesia Plan

1. Children may require preoperative ventilation and inotropic support.
2. SVR and PVR should be maintained to support aortic diastolic pressure and coronary flow. Therefore, prevent hyperventilation, excess O₂, or vasodilating drugs.
3. High-dose opioid technique is preferred.
4. After CPB and postoperatively, institute measures to protect against pulmonary vascular crisis.

HEART TRANSPLANTATION

Indications

1. Severe congenital malformations (e.g., HLHS) or failed previous surgical corrections of CHD not amenable to further surgery.
2. Cardiomyopathy—progressive with no chance of timely remission.
3. Myocardial tumors not amenable to resection (rare).

General Principles

1. The preoperative PVR is the most important determinant of the suitability of the infant or child for transplantation. Neonates have a high PVR, but a neonatal donor heart should be able to cope with this, and PVR may be expected to decrease over the first weeks of life. Otherwise, for older children, strategies must be used to reduce PVR or, failing this, heart–lung transplantation must be considered.
2. Other contraindications to transplantation include serious hepatic, renal, or central nervous system disease and chronic infections (i.e., hepatitis, cytomegalovirus, human immunodeficiency virus).
3. A stable family and social environment is most desirable to ensure the continued care that will be required after transplantation.

Care of the Donor

Care of the donor child during the organ harvesting is discussed in Chap. 13.

Anesthesia Management of the Recipient: Special Considerations

1. The basic management is similar to that for other open-heart procedures. However, the child may have been urgently admitted and may not have been fasted. Precautions for dealing with the full stomach may be necessary. If a rapid-sequence induction is planned, drugs and doses should be carefully worked out to prevent excessive cardiovascular effects. Ketamine or etomidate may be useful for the child with minimal reserve but exercise caution in children with chronic cardiomyopathy or depleted catecholamine stores.
2. Do not aspirate the stomach after induction if oral cyclosporin was recently given pre-transplant as a component of the anti-rejection therapy.
3. Most children have very poor cardiac function and a dilated heart; therefore take care to prevent inducing tachycardia, bradycardia, or any additional myocardial depression. A fentanyl-O₂ midazolam, muscle relaxant

anesthetic is usually preferred. Continue solutions of inotropic agents and/or PGE₁. Some children may need additional inotropic support as surgery commences.

4. Children who have had previous cardiac surgery should be given tranexamic acid to reduce postoperative bleeding.
5. Increased PVR should be managed to prevent any further pulmonary vasoconstriction.
6. Neonates with HLHS should be treated as for the Norwood procedure before CPB.
7. Infusions of dopamine and dobutamine, isoproterenol, and SNP should be primed for immediate use during weaning from bypass.
8. On weaning from CPB or as the clamps are removed and reperfusion starts, give methylprednisolone (Solu-Medrol) 15 mg/kg and other anti-rejection drugs as indicated. Sinus tachycardia is often present at this stage, and the action of two atrial pacemakers may be observed (one from the remains of the child's native atrium and one from the implanted atrium). In the absence of sinus rhythm, AV pacing should be commenced. A slow sinus rhythm usually responds well to an isoproterenol infusion. Infuse other inotropic solutions (e.g., dopamine) as necessary to maintain good cardiac action (see later discussion).
9. Measures to minimize PVR should be continued.
10. Remember the special properties of the newly implanted but denervated heart:
 - (a) Cardiac drugs exert *only their direct effects*; atropine will have no chronotropic effect; anticholinesterases will not affect heart rate. Epinephrine, isoproterenol, and norepinephrine will all increase the heart rate. Dopamine and dobutamine remain effective inotropic agents.
 - (b) Increased filling pressure will, through the Frank-Starling mechanism, result in increased stroke volume. A CVP of 10–12 mmHg is usually optimal. Hypovolemia is poorly tolerated.
 - (c) There is no change in heart rate with the respiratory cycle or with a Valsalva maneuver.
 - (d) Arrhythmias are not common in children, but the response to those antiarrhythmic drugs that have both direct and indirect effects on the heart will be altered:
 - Digoxin and procainamide normally exert a mixture of direct and indirect effects on the heart and will have a less predictable effect on the denervated heart.
 - The effects of lidocaine, phenytoin, β-adrenergic blocking drugs, and calcium channel-blocking drugs are direct and similar to those in the intact heart.

HEART-LUNG OR LUNG TRANSPLANTATION

Lung transplantation in children is usually performed with the aid of CPB; the considerations are similar to those for heart-lung transplantation.

Indications

1. The indications for heart-lung transplantation are
 - (a) Eisenmenger syndrome
 - (b) Other congenital defects with pulmonary vascular disease
 - (c) Complex CHD, inadequate pulmonary vessels not amenable to further repair
2. The indications for lung transplantation are
 - (a) Primary PAH
 - (b) Pulmonary fibrosis
 - (c) Cystic fibrosis

Anesthesia Management of the Donor

The general principles are outlined in Chap. 13. Selection of a donor for lung transplantation is more difficult:

1. Significant lung disease, infection, or damage from recent aspiration or pulmonary edema associated with resuscitative interventions and artificial ventilation must be excluded. It is suggested that a PaO₂ of 100 mmHg or more with an FIO₂ of 0.4, and peak inflating pressures of 30 cm H₂O or less with a tidal volume of 15 mL/kg and PEEP of 5 cm H₂O, indicate acceptability for transplantation.
2. The lungs must be an appropriate size to fit the thorax of the recipient; if they are too large, atelectasis will result. Perfect match or slightly smaller donor lungs are accepted; otherwise, lobar transplantation or tailoring of the donor lung may be required.
3. Before harvesting the lungs, the donor should receive 30 mg/kg of methylprednisolone and an infusion of PGE₁ (25 ng/kg/min) increased until the systemic arterial pressure decreases by 10–20 %. This produces maximal pulmonary vasodilation before infusion of the pulmoplegic solution.
4. Cardioplegia and pulmoplegia are induced, and the lungs are held inflated before tracheal clamping.
5. In selected cases, donor lung tissue may be obtained from living related donors (e.g., one lobe from each parent may be transplanted into a child).

Anesthesia Management of the Recipient

1. Older children with critical respiratory disease may be very apprehensive; consider the use of well-monitored preoperative sedation (e.g., midazolam or lorazepam intravenously with pulse oximetry in place and constant monitoring).
2. Before induction, verify administration of immunosuppressants and antibiotics as ordered.
3. Induction and maintenance of anesthesia should be planned as for CPB, bearing in mind the advanced respiratory disease.
4. Tracheal intubation: Use a cuffed tube and place the cuff just below the cords. For children with cystic fibrosis, suction the tube frequently during dissection of the native lungs.
5. Maintain meticulous aseptic technique for all procedures; remove all existing intravenous lines and replace them with new lines using strict asepsis. A pulmonary artery catheter should be inserted after induction.
6. Fluid therapy should be limited to basal rates.
7. Antifibrinolytic therapy (ϵ -aminocaproic acid or tranexamic acid) should be commenced after induction to decrease postoperative bleeding.
8. Plan for optimal postoperative pain management.
9. During bypass, be prepared to re-intubate with a new sterile tracheal tube, using appropriate aseptic technique. For children with pulmonary infections (e.g., those with cystic fibrosis), change the entire breathing circuit. In addition, once the native lungs are removed, the proximal trachea and bronchi should be lavaged with a solution of tobramycin. Once the new bronchi are anastomosed, fiberoptic bronchoscopy may be performed to assess the integrity of the anastomoses.
10. Dopamine, dobutamine, and PGE₁ infusions should be available. For those with pulmonary infections, prepare a phenylephrine infusion (see later discussion). Nitric oxide should be available.
11. Immediately on weaning from CPB or once reperfusion has started, administer methylprednisolone 15 mg/kg IV and furosemide 0.5–0.75 mg/kg. Give inotropic agents as required. Pulmonary function may be improved by albuterol inhalations and aggressive diuretic therapy.
12. Adjust the FIO₂ to maintain oxygen saturation at $\pm 93\%$ and to prevent hyperoxic damage to lungs. Hyperventilate slightly to maintain pulmonary flow using tidal volumes of 15 mL/kg. PEEP to 6–10 cm H₂O should be added to maintain optimal lung volume, minimize PVR, and prevent pulmonary edema.
13. Children with a history of severe lung infections (e.g., cystic fibrosis) may demonstrate signs of sepsis: low BP despite good cardiac action. In such cases, a norepinephrine infusion or low dose vasopressin may be required.

14. Persistent high PA pressures after transplantation should be treated by the addition of NO to the inhaled gases.
15. Extensive bleeding is to be expected, especially if the child has had a previous thoracotomy. Order appropriate supplies of replacement factors.
16. Postoperative problems may be related to damage to the phrenic, vagus, or recurrent laryngeal nerves.

CARDIOLOGIC PROCEDURES

Cardiac Catheterization

This is usually an elective procedure, and older children will benefit from preoperative teaching, a visit to the catheterization laboratory, and familiarization with the procedures to be performed. Cardiac catheterization may be performed under general anesthesia or with a combination of sedation plus local or regional analgesia. The important prerequisites for gathering reliable catheterization data are as follows:

1. Hemodynamic parameters during sedation/anesthesia should be as close to awake values as possible.
2. Maintain a constant inspired O₂ concentration throughout the procedure. Room air is preferred if tolerated by the child. Otherwise, a constant optimum inspired O₂ concentration should be selected.
3. Spontaneous ventilation is preferred when appropriate; controlled ventilation may change intracardiac shunts and modify intracardiac pressures.
4. The child should be maintained in an optimal physiologic state (e.g., normothermic and well hydrated).

Special Anesthesia Problems

1. The child may be seriously ill and in CHF.
2. The condition may further deteriorate during cardiac catheterization, especially if arrhythmias occur.
3. Contrast media used for angiograms may cause adverse effects.
4. Children with PAH are particularly prone to serious complications during cardiac catheterization and may require special consideration.
5. Cardiac perforation with tamponade is always a possibility.

Anesthesia Management

When the procedure is to be performed under sedation, the following technique has proved satisfactory:

1. Establish an intravenous route using local analgesia.

2. Apply monitors (ECG, pulse oximeter, BP cuff, temperature probe). If you have a Doppler probe positioned over an artery, it provides a very good means to monitor cardiac status moment by moment.
3. Maintain a thermoneutral environment, particularly in small infants (i.e., overhead warmer, forced air warming blanket).
4. Administer intravenous sedation. If hemodynamically tolerated, a propofol infusion with or without small doses of midazolam has been useful. When combined with local or regional analgesia, only very small doses of drugs are required to ensure sleep, and very stable cardiovascular parameters are maintained.
5. Small infants may be offered a “sucrose soother” and often settle with this alone.
6. Caudal analgesia may be useful for some children, especially if bilateral femoral catheterization is necessary or if large catheters are to be inserted (i.e., for balloon dilation).
7. Angiography requires that the child remain absolutely still; therefore augment the sedation if necessary. Contrast media are hyperosmolar (although nonionic agents are less so); they can cause aggregation of erythrocytes and, rarely, anaphylaxis. The total dose administered should be carefully recorded, especially in small infants and recommended limits observed.
8. The procedure may be carried out in a darkened room.
9. When it is considered advisable to ventilate the lungs during the procedure, the same technique (propofol infusion) may be used with the addition of a suitable muscle relaxant (e.g., rocuronium) and tracheal intubation. Normocapnia should be maintained.
10. In some children, the response of the pulmonary circulation to hyperventilation, hyperoxia, or the inhalation of NO may be studied.
11. Post-catheterization care: The child should be carefully monitored until the effects of sedation resolve and smooth awakening occurs. It is necessary that the child lie quietly to prevent bleeding and bruising at the catheter site; additional mild sedation may be necessary to achieve this state. The catheterization site should be examined for bleeding, and the pulses distal to arterial cannulation should be evaluated regularly. The child may be discharged from PACU when he/she is awake, with stable vital signs, and no evidence of vascular complications.

Interventional Cardiology

Sedation and/or general anesthesia are now quite frequently required for complex interventional techniques. These include:

1. Balloon dilation of pulmonary, mitral or aortic valves, or re-coarctation of the aorta
2. Occlusion of the ductus arteriosus

3. Closure of septal defects or other fistulas
4. Balloon dilations and stent placements in conduits, pulmonary arteries or veins
5. Percutaneous placement of pulmonary valves
6. Coiling of collateral vessels

Special Problems

1. The child may be critically ill.
2. Absolute immobility is essential during the critical stages.
3. An urgent call for open operation if complications occur is a real possibility. The heart may be perforated, vessels may be ruptured, or the occlusive device may become displaced. Complications are most likely during valve dilation procedures.
4. A steady hemodynamic state is required for measurements to be made.
5. Simultaneous TEE may be required to monitor results.

Anesthesia Management

1. Standard monitoring as per any open procedure. Two large-bore, reliable intravenous lines should be established. Inotropic medications and syringe pumps should be immediately available and defibrillator pads attached.
2. For some very simple procedures in older children, sedation with propofol with spontaneous ventilation and local analgesia may be suitable.
3. For more complex procedures (e.g., balloon dilation of a stenotic valve, "umbrella" closure of a septal defect), tracheal intubation with neuromuscular block and controlled ventilation is preferred. Anesthesia may be maintained with a propofol infusion if hemodynamically tolerated. Be prepared for major life threatening arrhythmias and hemodynamic instability during balloon dilations and stent placements.
4. Plans must be made for transfer to the OR should a complication occur. Supplies of blood for rapid transfusion should be immediately available.
5. During the procedure, the anesthesiologist must constantly monitor for signs of blood loss or cardiac tamponade.
 - (a) Cardiac perforation manifests with hypotension, and tachycardia with ectopic beats.
 - (b) Cardiac tamponade leads to hypotension, with reduced cardiac motion on fluoroscopy. Confirm by ECHO. Pericardiocentesis should be performed in either case. Continued bleeding from a cardiac perforation requires thoracotomy.
6. Children with PAH require special considerations:
 - (a) During catheterization the response of the pulmonary vasculature to increased levels of oxygen and/or inhalation of NO may be assessed.

- (b) Anesthesia management should be conducted to prevent precipitating increases in PVR.
- Excessive sedation leading to respiratory depression, increased PaCO₂, airway obstruction and hypoxemia must be prevented. Hypoventilation should be corrected early.
 - An adequate level of anesthesia should be ensured during potentially painful stimuli or instrumentation of the airway. Tracheal intubation should be avoided if possible in those with severe PAH.

ELECTROPHYSIOLOGIC STUDIES

Children may require anesthesia or sedation for electrophysiologic studies and radiofrequency catheter ablation of accessory conduction pathways in the treatment of dysrhythmias. These procedures are not painful, except during the moments of the actual radiofrequency ablation, but may be prolonged (consider inserting a urinary catheter), and absolute immobility is essential. Hence, general endotracheal anesthesia with controlled ventilation is recommended.

Isoproterenol may be infused during the procedure to elicit dysrhythmias to find accessory conduction pathways. If ablation of such pathways is the goal, defibrillation pads should be in place before the procedure and antiarrhythmic drugs should be immediately at hand.

An important consideration is the possible effect of anesthesia or sedative drugs and anesthesia techniques on cardiac rhythm generation and conduction. Some anesthetics (e.g., halothane) have much greater effects than others (e.g., propofol). Deep sedation or anesthesia with propofol or balanced anesthesia with nitrous oxide and low dose isoflurane do not interfere with EP studies or the ability to trigger SVT or VT. Therefore these agents are acceptable during electrophysiologic studies.

CARDIOVERSION

Cardioversion is usually an emergency procedure. The arrhythmia may be severe, markedly reducing cardiac output and producing shock.

Anesthesia Management

Preparation

1. Give 100% O₂ by mask until cardioversion can be performed.

2. Establish the fasting interval
3. Prepare and check all equipment. Standard monitors are required no matter the location of the cardioversion.
4. Establish reliable intravenous access.

During the Conversion

1. Continue 100 % O₂ by mask.
2. When ready to cardiovert, medicate with IV midazolam and induce anesthesia using a sleep dose of propofol (or ketamine if cardiac function is poor).
3. If the child has a full stomach:
 - (a) Continue 100 % O₂.
 - (b) Give atropine 0.02 mg/kg IV.
 - (c) Depending on hemodynamic status, give propofol 2.5–3.5 mg/kg or ketamine 1–2 mg/kg or etomidate 0.2–0.3 mg/kg (if a cardiomyopathy is present) and succinylcholine 1–2 mg/kg.
 - (d) Perform a RSI and secure the trachea.
4. As soon as anesthesia has been induced and good oxygenation achieved, countershock may be applied. Repeat propofol, ketamine, or etomidate doses as needed.

Post-procedure

1. The period of recovery is brief but the child should be closely monitored (including ECG) for several hours afterward.
2. If the trachea was intubated, remove the endotracheal tube after the child is fully conscious.

ANESTHESIA FOR CARDIAC MRI

MRI studies now provide a very important tool for mapping the anatomy of the heart and this is very useful in CHD. Studies can be satisfactorily performed with sedation (e.g., propofol). However, general anesthesia with intubation may be required if the child is unstable or if apneas are required for better imaging.

Special Anesthesia Problems

1. All considerations for sedation or anesthesia in the MRI unit must be observed (see page 510). For general tracheal anesthesia, an MRI compatible ventilator must be available.

2. The specific considerations of the child's cardiac disease must be addressed.
3. If tracheal intubation is required; children with potential difficult intubation should be intubated in the OR and then transferred to the MRI suite. All children should recover in a properly staffed and equipped PACU.

General Anesthetic Management

1. Monitors must all be applied and functioning before induction.
2. The technique chosen must depend on the physiologic status of the child; stable children may be induced by inhalation of sevoflurane or a titrated IV dose of propofol. Potentially unstable children may be managed using an infusion of opioid analgesics (e.g., remifentanil) and small doses of midazolam.
3. Cis-atracurium is preferred for neuromuscular blockade (brief duration of action).
4. The lungs may be ventilated with an FIO₂ that is most appropriate for the specific cardiac defect and will maintain adequate oxygenation during the frequent apnea periods.
5. Oxygen saturation and end-tidal CO₂ should be monitored.

ANESTHESIA FOR NON-CARDIAC SURGERY IN INFANTS AND CHILDREN WITH CONGENITAL HEART DISEASE

CHD often occurs in association with other congenital defects, some of which may require surgery in the neonate or infant. Older children with CHD frequently require anesthesia for non-cardiac procedures (e.g., dental surgery). Therefore, the anesthesiologist may be called on to provide care for children with CHD for other types of surgery. Some of these children have uncorrected cardiac lesions, others have undergone partial (palliation) procedures, and others have had complete repair of their defect. However, even after "complete repair" there may be important considerations, such as the need for prophylactic antibiotics (see page 308) the presence of a pacemaker for heart block, or other residual defects. After repair of complex lesions, the post-repair physiology (e.g., post-Fontan repair) may demand special perioperative considerations. Remember that the appearance of an intraoperative arrhythmia may be a very important observation indicating the potential for a more serious sudden cardiac event and should be reported to the child's cardiologist.

Identifying the Patient with Congenital Heart Disease

The first challenge may be to determine whether the child has CHD. Although some children with CHD may be diagnosed in utero, many still go unrecognized throughout the neonatal period. In neonates, the diagnosis of CHD can be quite difficult (see previous discussion). All neonates with defects that are commonly associated with CHD (i.e., tracheoesophageal fistula, diaphragmatic hernia, omphalocele) and those with the signs just listed should be screened with an ECHO before induction of anesthesia.

Older children may be found to have a previously undetected and undiagnosed murmur when examined before anesthesia. The problem is to determine whether significant heart disease is present and whether it is necessary to refer the child to a cardiologist. First, it is important to determine whether the child has normal exercise tolerance. It is very unlikely that a child with unlimited activity has a lesion that will cause problems during anesthesia. On physical examination, the characteristics of the murmur should be analyzed:

1. *Innocent murmurs* are soft, systolic, and not radiated; they may vary with position, may disappear on exercise, are not characteristic of any lesion, and are heard in healthy children.
2. *Non-innocent murmurs* include all diastolic murmurs, all pansystolic and late systolic murmurs, all loud murmurs, all continuous murmurs (except venous hum), and all transmitted murmurs.

If a soft and presumably innocent murmur is heard in a healthy child, surgery usually is not delayed. If a murmur suggestive of a definite, previously undiagnosed cardiac lesion is heard, delay elective surgery and refer to a cardiologist. Emergency surgery must proceed, and the child should be managed with due regard for the cardiac disease; prophylactic antibiotics should be administered when indicated (see page 309) and appropriate monitoring established.

Anesthesia Management

Preoperative

1. Many of the potential problems during non-cardiac surgery are similar to those associated with cardiac surgery, and the same considerations for each specific lesion apply. For major surgery, the child should be monitored as for cardiac surgery.
2. The anesthesiologist must clearly understand the pathophysiology of the child's heart disease and carefully assess the current physical status. See pages 389 for the physiologic changes associated with CHD.

3. The child's current medications must be reviewed and discussed with the parents. See previous discussion regarding which medications should be administered or withheld on the day of surgery.
4. Care must be taken to avoid excessive fluid restriction, especially in children with cyanotic CHD. The parents should be instructed to encourage clear fluids up until 2 h before the operation. If oral fluids cannot be taken, intravenous fluid therapy must be established.
5. Order appropriate preoperative sedation, but avoid producing respiratory depression. The child should be sedated but not depressed. Oral midazolam is ideal, but the child should be monitored with a pulse oximeter once sedation is achieved. Apply a topical analgesic cream to a likely intravenous site.
6. Ensure that equipment for cardiopulmonary resuscitation (including a defibrillator with paddles of suitable size) is available in the OR suite.
7. Check coagulation status in those children with cyanotic heart disease; coagulopathy is a common complication of polycythemia.
8. Anesthesia must be carefully planned to minimize the possibility of adversely affecting the cardiovascular status of the child.
 - (a) Use extreme care with potent inhalational agents or other cardiac depressants, especially in those with a history of or predisposition to CHF.
 - (b) Avoid causing major changes in PVR or SVR.
 - Carefully maintain ventilation and oxygenation.
 - Avoid vasodilating drugs (e.g., isoflurane >0.5 %)
 - Perform a rapid but atraumatic laryngoscopy and tracheal intubation.
 - Ensure adequate analgesia before surgery or anesthetic interventions.
 - Exercise extreme caution in children with PAH, anticipate their increased potential for complications and avoid precipitating a pulmonary hypertensive crisis (see page 393).
9. Administer antibiotic prophylaxis as indicated (Table 14.2). See page 308 for antibiotic prophylaxis regimens for children undergoing dental surgery. Routine prophylactic antibiotic therapy is no longer recommended for the prevention of infective endocarditis in children undergoing gastrointestinal or GU surgery. However, check with a cardiologist for their recommendations if in doubt.
10. Minimally invasive video assisted surgery is now used for many procedures and provides the same advantages of decreased postoperative pain and more rapid recovery for the cardiac patient. However, the special considerations for these procedures (see page 331) must be observed together with the requirements dictated by the cardiac lesion. Provided intra-abdominal pressure is maintained at below 10 mmHg most children will tolerate these procedures.

Table 14.2 Antibiotic routine for patients with cardiac disease**Dental procedures, oropharyngeal surgery, instrumentation of the respiratory tract including nasotracheal intubation**

Standard oral regimen for children includes those having prosthetic heart valves and other high-risk factors^a:

Amoxicillin 50 mg/kg PO 1 h preoperatively to a maximum of 2 g

Regimen for children allergic to amoxicillin/penicillin:

Clindamycin 20 mg/kg PO 1 h preoperatively

Alternative regimen for children unable to take oral medications:

Ampicillin 50 mg/kg IV or IM 30 min preoperatively

Regimen for children allergic to ampicillin/penicillin and unable to take oral medications:

Clindamycin 20 mg/kg IV 30 min preoperatively

Regimen for children with methicillin sodium-resistant staphylococcal infections:

Vancomycin 20 mg/kg (maximum, 1 g) IV given over 1 h, starting 1 h preoperatively

Gastrointestinal or genitourinary procedures or instrumentation

Standard regimen for children at high risk:

Ampicillin 50 mg/kg (up to 2 g) plus gentamicin 1.5 mg/kg (up to 120 mg); IV/IM, 30 min preoperatively and repeat ampicillin 25 mg/kg IV/IM 6 h later; or amoxicillin 25 mg/kg PO

Regimen for high-risk children allergic to ampicillin:

Vancomycin 20 mg/kg (maximum, 1 g) IV plus gentamicin 1.5 mg/kg

(maximum 80 mg) IV 1 h preoperatively

Alternative regimen for children at moderate risk:

Amoxicillin 50 mg/kg PO 1 h preoperatively or ampicillin 50 mg/kg IV/IM 30 min preoperatively

^aHigh-risk factors for subacute bacterial endocarditis include the presence of prosthetic valves or materials (e.g., Gore-Tex shunts), cyanotic lesions, and especially tetralogy of Fallot. (Wilson W, et al. see Suggested Reading)

11. The child who has had a Fontan procedure demands special consideration. Most important requirements are to maintain preload constant, prevent myocardial depression, and to ensure that PVR remains optimal. Spontaneous ventilation is ideal but if IPPV is required, ventilator settings to minimize intrathoracic pressure should be selected (brief inspiratory times, limited peak inspiratory pressure, and minimal PEEP). Fluid therapy must be carefully monitored and managed to maintain preload.

Perioperative

1. Attach all monitors before inducing anesthesia.
2. Establish a reliable intravenous route, but be aware of the risk of paradoxical emboli. Be careful to remove all bubbles from intravenous lines, especially in children with right-to-left shunts, but remember that others also may have bidirectional shunts.
3. Give 100% O₂ by mask unless the defect is an unrepairs single ventricle or large VSD.

4. Induce anesthesia with titrated doses of intravenous propofol or ketamine depending on the heart defect. Otherwise, inhalation induction with sevoflurane may be acceptable in some children, but beware of depressing the myocardium or triggering airway obstruction and hypoxemia-induced PAH.
5. Intubate the trachea for all but the most minor procedure (e.g., myringotomy). Give a suitable relaxant, but remember that if the circulation time is prolonged, there will be a longer delay before muscle paralysis is complete.
6. Children with less severe disease tolerate low concentrations of inhalational agents well; for minor procedures, maintain anesthesia with N₂O and sevoflurane or isoflurane with spontaneous or assisted ventilation. Children with more severe disease and those with any history of CHF cannot tolerate the use of myocardial depressant potent volatile agents. These children should be managed with a high-dose opioid, midazolam, and relaxant (pancuronium or rocuronium) technique, with controlled ventilation.
7. Maintain an adequate inspired concentration of oxygen, and monitor the oxygen saturation carefully. Pulse oximeters are less accurate at lower saturations; therefore err on the safe side.
8. For major non-cardiac surgery, insert arterial and other lines and monitor the child as for cardiac surgery. Consider the effects of previous surgery (e.g., systemic-to-pulmonary shunts) in choosing the BP cuff location.
9. PetCO₂ may not correlate well and tends to underestimate PaCO₂ in those children with right-to-left shunts but it does give valuable information as a trend indicator of cardiac output and pulmonary blood flow.
10. Replace blood and fluid deficits and losses accurately.

Postoperative

1. Continue to monitor the child (including ECG and oximeter) until the child has fully recovered from all effects of anesthesia. Transfer to the PICU or PACU as appropriate.
2. Give O₂ until recovery is complete. Those with cyanotic CHD have a reduced ventilatory response to hypoxemia.
3. Provide good pain control. Pain and restlessness increase oxygen demand.
4. Give maintenance fluids intravenously until oral intake is adequate, but avoid overhydrating.

Suggested Reading

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Orthopedic Surgery

A considerable proportion of children who undergo elective orthopedic surgery have multiple congenital anomalies and/or neuromuscular disease (see Appendix A). Underlying diseases, particularly those associated with muscle weakness, require special anesthesia care; even minor surgery may be fraught with major anesthesia complications.

GENERAL PRINCIPLES

1. Children with orthopedic deformities may require repeated surgery and spend much time in the hospital; sympathetic management is particularly important and the liberal use of preoperative sedation may be indicated.
2. Check the history carefully. Neuromuscular disease is particularly relevant. In general, muscle relaxants, particularly succinylcholine, should be avoided in children with myopathies (see Appendix A). Drug selection is influenced by the underlying disease and the type of neuro-monitoring that will be used intraoperatively.
3. Major surgery of the vertebral column deserves special consideration; the operations are extensive and may involve massive blood loss. Be prepared for a major transfusion. Preoperative blood donation, autotransfusion, and acute normovolemic hemodilution may be appropriate in some cases. Amicar and tranexamic acid decrease blood loss in spine surgery.
4. Malignant hyperthermia, though very rare, is more common in children with orthopedic diseases. Maintain vigilance for the early signs of a reaction (see Chap. 6).
5. When a tourniquet is used, blood loss is negligible. In other cases, surgery involving bone may result in significant blood loss (e.g., innominate osteotomy). Therefore, establish a reliable intravenous route and confirm that blood is available.
 - (a) When a tourniquet is inflated, there usually follows a progressive increase in the heart rate and blood pressure. The exact cause is unknown, but it has been attributed to sympathetic stimulation. If the anesthesiologist has been aggressively treating the tourniquet-induced

hypertension by giving greater concentrations of inhaled agents, the blood pressure may decrease precipitously on tourniquet release. Therefore, use caution with concentrations of potent inhalational agents and other drugs while the tourniquet is being used and reduce the concentration in anticipation of tourniquet release.

- (b) Hemodynamic and metabolic responses to tourniquet release in children are usually not clinically important. A transient decrease in arterial pH associated with an increase in base deficit and carbon dioxide tension (PaCO_2) does occur; this is most marked after prolonged tourniquet times or with the use of bilateral tourniquets. General recommendations include the following:
- Attempt to limit tourniquet times to less than 75 min.
 - Use controlled ventilation before and after tourniquet release to remove the respiratory component of the acidosis.
 - Do not release bilateral tourniquets simultaneously.
 - In children in whom metabolic or respiratory acidosis is not easily compensated for (i.e., those with renal disease or pulmonary disease), consider staged release of the tourniquets and close monitoring of SaO_2 and PetCO_2 .
 - Serious complications of tourniquet use in children are very rare; however, pulmonary embolism on release has been reported in an obese child. Consider subcutaneous heparin in such children and monitor cardiopulmonary function closely after release.
 - Ensure that the tourniquet is *removed* from the limb as soon as it is deflated. Residual pressure compromising perfusion of the limb has resulted in serious complications (including amputation).

6. Orthopedic surgery is associated with high levels of postoperative pain. Plan for optimal management, using regional analgesia when possible. The introduction of continuous peripheral nerve blocks using disposable pumps has opened up new avenues for postoperative pain control after limb or trunk surgery.
7. Compartment syndrome may complicate orthopedic injuries, especially supracondylar fractures of the humerus and forearm and tibial fractures. There is a concern that effective analgesia may mask the presenting sign of this syndrome (i.e., increased pain). This is especially of concern when the limb cannot easily be examined (i.e., encased in a cast). Compartment syndrome is sufficiently rare that withholding effective analgesia seems unreasonable; there is no evidence to confirm that regional analgesic regimens delay the diagnosis. Effective postoperative pain relief should therefore be provided for every child. Increased analgesic requirements, breakthrough pain, or pain remote from the surgical site should prompt direct measurement of compartment pressure.

8. Urinary retention may occur in children who have lower limb surgery and are treated with a regional block when an opioid is added to the block solution. This should be noted by postanesthesia care unit (PACU) staff and treated as necessary.
9. Children with cerebral palsy frequently present for orthopedic surgery. These children require special considerations (see Chap. 6).
10. Children with Duchenne muscular dystrophy may present for major scoliosis surgery and require careful assessment of their cardiorespiratory status and anticipation of increased bleeding (see later discussion and Appendix A).

MISCELLANEOUS ORTHOPEDIC PROCEDURES AND ANESTHESIA CONSIDERATIONS

Hip Arthrogram

Hip arthrograms are performed in infants to assess the head of the femur and other aspects of the hip joint. As part of the procedure, a small amount of air may be injected into the joint to ensure that the tip of the needle is in the joint space before injecting contrast material; serious air embolism and cardiac arrest have occurred.

Recommendation: If a hip arthrogram is planned, determine whether air will be injected. Monitor the child very carefully during injection, omit nitrous oxide, and monitor for evidence of an air embolism.

Club Feet

Beware of the high incidence of myopathies in children having club feet. Examine the child and the medical history carefully for any indications of a myopathy. If positive, revise the anesthesia technique appropriately.

In some cases, regional analgesia techniques combined with intravenous sedation may be optimal for clubfoot surgery. Caudal epidural analgesia may augment intraoperative management and provide for postoperative pain management.

LIMB FRACTURES

Closed Limb Fractures

Injuries to upper limbs are common; many of these children are older; therefore regional analgesia may be appropriate. If so:

1. Use adequate local analgesic drug to produce a profound block.

2. Perform the block well in advance of the scheduled surgery so that it has plenty of time to become well-established.
3. **Do not allow the surgeon to handle the limb until you know the block is working satisfactorily.**
4. If supplementary sedation is required, midazolam is usually satisfactory.

Fractures of the Forearm

1. Perform a block of the brachial plexus via the axillary route (see Chap. 5 for drugs and doses). Ultrasonography improves the success with brachial plexus blocks.
2. Intravenous blocks are usually not satisfactory for reduction of proximal fractures, but may be very efficacious for distal fractures. The cast may be satisfactorily applied to the exsanguinated limb.

Fractures of the Femur

1. A block of the femoral nerve with lidocaine or bupivacaine is easy to perform and relieves pain and muscle spasm if traction apparatus is being applied.
2. A catheter may be introduced to the femoral sheath and a continuous femoral nerve block maintained (see continuous femoral block Chap. 5).

General Anesthesia

Every child with a recent fracture must be considered to have a full stomach and a RSI should be performed. Vomiting frequently occurs during emergence from anesthesia; therefore, the child should be fully awake and in a lateral position before extubation.

Postoperative

1. Pain may be quite severe after routine orthopedic surgery and should be controlled by either systemic analgesic drugs or regional analgesia or a combination of these:
 - (a) Regimens combining acetaminophen and a nonsteroidal anti-inflammatory drug (NSAID) are more effective than either drug alone. Acetaminophen, ibuprofen, or diclofenac augmented by titrated doses of opioids as required is a suitable basic regimen. Pain should be assessed using standard objective measures (see Chap. 7).
 - (b) Patient-controlled analgesia (PCA) may be appropriate for many children over the age of 4 or 5 years (see Chap. 7).
 - (c) Regional analgesia provided by neuraxial or peripheral nerve block. When possible, the latter should be chosen as this is less likely to cause complications. In children, it is customary to perform blocks and insert

catheters under general anesthesia. Peripheral nerve blocks and catheter placement should be guided by ultrasonography when possible.

- (d) Peripheral nerve blocks may be initiated in the operating room and continued through the PACU to the ward or even to home using a disposable elastomeric infusion pump. The decision to continue this at home will depend upon the home circumstances and the attitude and abilities of the parents. A postoperative infusion of ropivacaine 0.2% at a rate of 0.1 mL/kg/h has been suggested for continuous peripheral nerve blocks.

KYPHOSCOLIOSIS

Kyphoscoliosis may be congenital (15%), idiopathic (65%), or secondary to neuromuscular disease (20%); 80% with idiopathic scoliosis are female. Pulmonary function may be impaired, and some children with associated diseases (myopathies, cerebral palsy) may be severely disabled, wheelchair bound, and physiologically debilitated.

Pulmonary Function

Changes in pulmonary function are related to the underlying cause, the speed of development of the scoliosis, and the severity of the curvature. The cardiorespiratory effects of scoliosis are summarized in Fig. 15.1.

The principal concern for young children with idiopathic scoliosis is the cosmetic effect of the spinal and pelvic or chest wall deformity, especially when the curvature increases during the years of rapid body growth. At this stage, respiratory symptoms are uncommon, but pulmonary function studies may reveal an abnormality. Although lung volumes can be normal, exercise tolerance may be reduced. In severe cases, the mechanical effects of scoliosis on respiratory function are apparent even at rest.

Pulmonary function is relatively normal in most children who present for correction of idiopathic scoliosis with a curvature of less than 65%. Respiratory disability is more likely to occur in association with congenital scoliosis or curvature of paralytic etiology.

The pulmonary abnormality is restrictive, with a reduced chest wall compliance. The vital capacity and the total lung capacity may be dramatically reduced, and the functional residual capacity somewhat less so. The residual volume tends to be maintained. The elastic resistance of the chest wall may be high, increasing the work of breathing. If left untreated, severe and prolonged lung compression impairs gas exchange, which becomes evident only in later stages of the disease.

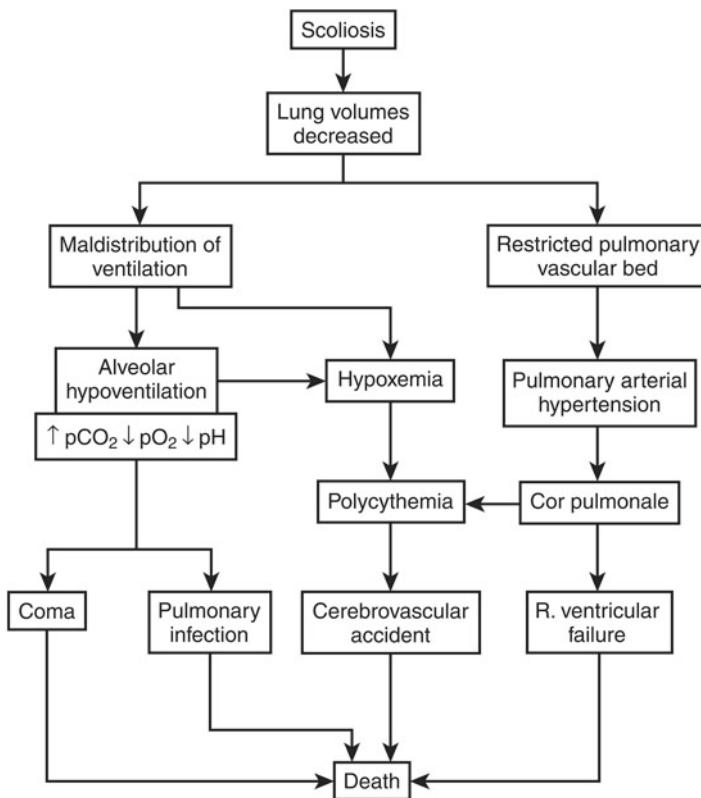


Fig. 15.1 Pathophysiology of the cardiorespiratory effects of kyphoscoliosis. Progressive alveolar hypoventilation, leading to hypoxia, may be accompanied by pulmonary hypertension and right ventricular failure (*Courtesy Henry Levison, MD, Former Director of Respiratory Physiology, The Hospital for Sick Children, Toronto*)

The High-Risk Scoliotic Child

Scoliosis surgery may be recommended for severely incapacitated children for the purpose of facilitating their ongoing care and arresting the progression of cardiorespiratory compromise. There is a very high incidence of perioperative complications in this cohort and many require prolonged postoperative intensive care; however, the ultimate outcome is judged by many of these children and their caregivers to be worthwhile. It is very important that these children be optimally prepared for their surgery:

1. The nutritional status should be assessed and optimized using total parenteral nutrition or via a gastrostomy as appropriate.
2. The pulmonary system should be evaluated for reversible disease that might be improved by physiotherapy or antibiotic therapy. Those with neuromus-

cular disease and a preoperative $\text{FEV}_1 < 40\%$ of predicted are likely to need postoperative ventilation.

3. The cardiac status should be evaluated by echocardiogram (i.e., myocardial contractility, RV function, etc.) and preoperative therapy initiated as indicated.
4. The pediatric intensive care unit (PICU) should be notified that the child will require admission and they should be familiar with his or her preoperative condition.

Surgical Procedures

1. Posterior spinal fusion may be performed using contoured metal rods to stabilize the spine postoperatively until bony fusion occurs. In some children with a flexible spine, the deformity is corrected solely by a posterior fusion. Adjustable rods may be placed in young children to permit adjustments during growth.
2. In some, an anterior thoraco-abdominal approach may be used to remove the intervertebral discs or a hemivertebra to correct a lateral curve. This may be an open procedure or an endoscopic procedure in which case the special considerations for endoscopic surgery apply (see Chap. 13).
3. In others, the two procedures are combined: an anterior release followed by a posterior fusion. In this case, surgery is often prolonged, and associated with significant blood loss.

Special Anesthesia Problems

1. Anesthesia management must take into account the following:
 - (a) The severity and cause of the curvature. The more severe the curve, the more pulmonary function is impaired, and greater the likelihood of postoperative pulmonary failure.
If the scoliosis is secondary to neuromuscular disease:
 - Pulmonary function impairment caused by the mechanical effects of the spinal curvature may be compounded by involvement of respiratory muscles in the disease process. Postoperative respiratory insufficiency is more likely.
 - Increased bleeding may be expected. This may be a result of altered vascular responses and platelet adhesion associated with myopathies (e.g., Duchenne muscular dystrophy).
 - Cardiomyopathy may occur in adolescents with Duchenne muscular dystrophy.
 - Selection of suitable drugs may be limited (i.e., avoid succinylcholine and inhalational anesthetics to prevent rhabdomyolysis if a myopathy is present). Titrate doses of non-depolarizing relaxants if required.

- (b) The degree of respiratory and cardiovascular impairment:
- Surgery may be expected to stabilize the cardiorespiratory effects of the disease but may not result in improvement.
 - A further impairment of pulmonary function must be anticipated in the early postoperative phase and may require respiratory support.
- (c) The type of corrective procedure proposed.
- Posterior fusion only.
 - Anterior and posterior fusion combined.
2. Preoperative assessment must include the following:
- (a) Detailed history and examination for an indication of disabilities, stamina, and the presence of any related medical conditions.
 - (b) Routine hematology, coagulation, biochemistry studies, and cross-match.
 - (c) Pulmonary function studies, including blood gas analysis. (These studies may not be possible in very young children due to lack of cooperation.)
 - (d) Echocardiogram to assess myocardial function for all children (particularly adolescents) with very severe curves or an associated myopathy.
3. Be alert for signs of significant respiratory impairment (i.e., tachypnea at rest, severely reduced vital capacity, abnormal blood gas values, inability to cough effectively). Postoperative hypoventilation, secretion retention, and atelectasis are likely in response to pain, analgesic drugs, and immobilization that will further compound existing problems.
4. Severe impairment of respiratory function is not a contraindication to surgery provided that resources are available for postoperative intensive respiratory care (including controlled ventilation, if necessary). Fixation of the spinal deformity is essential to prevent further deterioration of respiratory function (but usually does not result in significant early improvement).
5. Children with a vital capacity less than 40 % of normal or a Cobb angle of $>69^\circ$ may develop major postoperative respiratory complications and likely will require postoperative ventilation.

Corrective Surgery by the Posterior Approach

Special Anesthesia Problems

1. Pulmonary function may be severely impaired. The anesthesiologist should check that the present state is optimal and exclude any superimposed acute respiratory disease. Because the child will be in the prone position, extra care should be taken to secure the tracheal tube to prevent dislodgement. If preoperative curvature correction has been achieved with an exoskeletal

apparatus, intubation will likely be difficult; advanced airway adjuncts such as the GlideScope or a fiberoptic technique may be necessary to secure the airway.

2. Explain to the family and the child the need for invasive lines, postoperative ventilation (if likely), the need for ICU, and the potential for an intraoperative wake-up test.
3. Blood loss may be severe (in excess of 50 % of the estimated blood volume [EBV]). Most bleeding originates from the vertebral veins, which become engorged if there is any pressure on the anterior abdomen. Blood loss is also related to the extent of the surgery (length of spine to be fused) and to the surgeon's speed and expertise. Those with scoliosis secondary to a recognized neuromuscular disorder usually have a larger blood loss than those with idiopathic scoliosis. Alternatives to homologous transfusion should be considered:
 - (a) Preoperative autologous blood donation.
 - (b) Intraoperative acute normovolemic hemodilution may be used cautiously.

(These procedures may be augmented by oral ferrous sulfate and Vitamin C daily beginning 4 weeks before surgery and followed by twice-weekly intramuscular injections of erythropoietin commencing 2 weeks before surgery. The hematocrit (Hct) should not be allowed to exceed 55 % preoperatively).
 - (c) A cell saver may be used to salvage erythrocytes from suctioned blood. However, transfusion of large volumes of washed cells may lead to coagulopathy because of dilution of coagulation factors. Fresh-frozen plasma should be available and administered if blood loss exceeds one blood volume.
4. Spinal cord function is usually monitored during surgery: the use of somatosensory-evoked potentials (SSEPs) has been augmented by the addition of motor-evoked potentials (MEPs), stimulating the cord above the level of the surgery and recording the electromyogram from the limb.

Anesthetic techniques to facilitate the monitoring of evoked potentials limit inhalational agents to 0.5 MAC. Large doses of opioids may be given and benzodiazepines used to provide amnesia. Nitrous oxide decreases the amplitude of evoked potentials and is usually avoided. Propofol/opioid infusions are probably optimal for these procedures. During the testing of MEPs, better results are obtained with minimal neuromuscular blockade (i.e., two to four twitches on a train of four); the degree of neuromuscular block to be applied should be discussed with the neurophysiologist. Because of the difficulty in assessing depth of anesthesia during TIVA, a depth of anesthesia monitor is advised. For transcranial MEPs, insert a bite block to avoid tongue and lip lacerations.

Despite the use of multimodal neurophysiologic monitoring, a wake-up test may still be required (i.e., a surgical misadventure or failure of the monitoring system). Fortunately, the anesthesia technique that provides for neurophysiologic monitoring also allows for a rapid wake-up test should this be required.

Every child should be awake at the end of the operation, so that both sensory and motor function can be tested immediately. Any defects should be reported to the surgeon promptly.

5. Postoperative pain is considerable; intrathecal opioids administered intraoperatively have been effective for postoperative analgesia and may also facilitate intraoperative BP control and reduce blood loss. Otherwise, epidural analgesia or PCA should be planned and these options discussed with the child and family.
6. Visual loss is a rare (incidence <0.2 %) but serious complication after spine surgery.
 - (a) The most common ocular injury after surgery is a corneal abrasion. Far less common is visual loss because of posterior ischemic optic neuropathy (PION) (occurring three times more frequently than anterior AION) or central retinal artery occlusion (CRAO) may rarely occur after spine surgery—especially when performed in the prone position.
 - (b) Age: occurs rarely in pediatric patients (<18 years).
 - (c) Factors associated with visual loss because of ION include preoperative anemia, prolonged surgery, intraoperative hypotension, low hematocrit, and major blood loss. CRAO results from direct facial-orbital compression.
 - (d) A possible but unproven factor in the genesis of post-spinal visual loss is excessive crystalloid fluid administration intraoperatively. This is known to increase intraocular pressure (IOP) and periorbital edema.
 - (e) **N.B.** Visual loss recovers in 44 % of those with ION but in none of those with CRAO.

Precautions that should be taken to reduce the risk of intraoperative visual loss:

1. Assess the child carefully and encourage staged procedures if the proposed operation may be excessively long (>6 h) or associated with large blood losses (>45 % estimated blood volume).
2. Position carefully:
 - (a) Avoid pressure on the eyes—check frequently during surgery and document this in the anesthesia record.
 - (b) Position the head in a neutral forward position (no neck flexion, extension, or rotation) so that it is level with or above the heart.

3. Monitor the hematocrit frequently and avoid markedly reduced levels intraoperatively.
4. Monitor central venous pressure and administer balanced crystalloid and colloid solutions to maintain an adequate blood volume.
5. Induced hypotension is an accepted practice for spine surgery, but avoid excessively reduced levels from baseline (i.e., maintain pressure no less than 20–25 % below baseline mean arterial pressure or a minimum systolic pressure of 80–90 mmHg).

Check the child's vision immediately upon awakening and seek consultation if there are any defects.

Anesthesia Management

Preoperative

1. Premedication with oral midazolam is usually adequate if combined with reassurance and a full explanation of procedures to be performed. Lorazepam 1–2 mg PO 1 h preop is very effective for adolescents.
2. Do not give respiratory depressant drugs to children whose respiratory function is impaired.
3. Ensure that all equipment and drugs are prepared in case of emergency.
4. Check that an adequate supply of blood and other fluid replacements is at hand.
5. Review pertinent laboratory, echocardiographic and pulmonary function studies.

Perioperative

1. If halo-loop traction is in place, check that instruments to release the connecting rods are at hand.
2. Tracheal intubation:
 - (a) In uncomplicated cases, induce anesthesia with propofol or thiopental followed by a low dose of an intermediate-acting relaxant (e.g., rocuronium 0.3–0.4 mg/kg) and intubate. If evoked potential monitoring is planned, allow the child to recover from the neuromuscular blockade, then check the positioning and function of the stimulating electrodes over the posterior tibial nerve.
 - (b) If exoskeletal apparatus is present, direct laryngoscopy may not be possible.
 - Have appropriate difficult airway adjuncts at hand.
 - Select a suitable intubation technique (i.e., fiberoptic intubation awake or under general anesthesia).

3. Maintenance:

Generally, a balanced anesthesia with low-dose inhaled anesthetic and opioid (nitrous oxide is generally avoided because it depresses SSEPs) or propofol and opioid (i.e., total intravenous anesthesia [TIVA]) technique is used.

(a) Opioid infusions:

- Fentanyl-loading dose of 5 µg/kg; infusion at 3 µg/kg/h.
 - Morphine-loading dose of 100 µg/kg; infusion at 10–30 µg/kg/h.
 - Remifentanil infusion at 0.1 µg/kg/min and adjusted up or down to control blood pressure as needed.
- (b) Limited concentrations of isoflurane (0.5–1 %), sevoflurane (1–1.5 %), or desflurane (3–5 %) may control the BP as needed. This technique does not usually interfere with neurophysiologic monitoring. If MEPs are to be measured, neuromuscular block is generally permitted only for tracheal intubation.
- (c) A TIVA protocol. This is a useful technique when multimodal neurophysiologic monitoring is to be performed. A propofol infusion is administered; though propofol decreases the amplitude of transcranial MEPs, these can still be acceptably monitored. Propofol is commonly administered with remifentanil as follows: Propofol 100–150 µg/kg/min with remifentanil 0.1–0.3 µg/kg/min. The dose of propofol may be modified as surgery proceeds on the basis of BIS readings or other benchmarks of anesthetic depth. The remifentanil infusion may be adjusted according to hemodynamic parameters. It is also recommended that other sedative drugs may be administered during TIVA, including dexmedetomidine and/or midazolam, the latter to ensure amnesia.
- (d) Nitrous oxide is usually avoided as it interferes significantly with neurophysiologic monitoring.
- (e) Insert a nasogastric tube and a bite block to prevent bleeding from MEPs.

4. Position the child to avoid any external pressure on the anterior abdominal wall. Maintenance of the correct prone-suspended position is essential to ensure minimal blood loss. (If the child is mal-positioned so that pressure on the abdomen causes vertebral venous engorgement, heavy blood loss is inevitable.) Ensure that there is no pressure on the eyes and that the considerations for the prevention of postoperative visual loss are observed (see previous discussion).**5. Monitor the following throughout:**

- (a) Ventilation-esophageal stethoscope, airway pressure, PetCO₂.
- (b) Circulation—ECG, pulse oximeter, arterial line, and central venous pressure (CVP).
- (c) Temperature—rectal or esophageal probe.
- (d) Neuromuscular blockade—peripheral nerve stimulator.

- (e) Blood loss—gravimetric method and graduated suction bottles.
 - (f) Urine output—indwelling catheter.
 - (g) SSEPs and/or MEPs—before, during, and after correction.
 - (h) Hematology and biochemistry—acid-base and blood gas status, Hct and coagulation status as indicated by the duration and severity of the procedure.
 - Head/neck position; no pressure on the eyes; document periodic assessment in the anesthesia record.
6. Blood loss is minimized by:
- (a) Proper posture (see item 5, previously).
 - (b) Deep infiltration of the wound site (by the surgeon) with a large volume of dilute epinephrine/saline solution (up to 500 mL of 1:500,000 solution may be used).
 - (c) Controlled ventilation, maintaining PaCO₂ at 30–35 mmHg to avoid hypercarbia and vasodilation.
 - (d) Surgical technique (firm packing and meticulous subperiosteal plane dissection).
 - (e) Moderate hypotensive techniques may be used, but the MAP should be maintained no more than 20–25 % below baseline. In the prone child, the spinal cord is above the heart and, if hypotension occurs, spinal cord ischemia may ensue, especially while it is manipulated or stretched, leading to paraplegia. Do not combine hypotensive technique with hemodilution.
 - (f) Acute normovolemic hemodilution (i.e., an alternative blood conservation method) is carried out as follows:
 - A calculated volume of blood is withdrawn to reduce the Hct to 30 %. Use the preoperative Hct and the EBV (in milliliters) to calculate the volume to be withdrawn:
$$\text{Volume withdrawn} = (\text{Hct} - 30) \times \text{EBV/Hct}$$
 - When the child is anesthetized and lines have been inserted, a weighed volume of blood is withdrawn via the arterial line into citrate-phosphate-dextrose bags for storage.
 - During blood withdrawal, a volume of warmed lactated Ringer's solution equal to three times the blood volume withdrawn is infused or 1:1 replacement with 5 % albumen.
 - As surgery progresses, the blood that has been withdrawn is reinfused. This method results in loss of lower-Hct blood during surgery and conservation of the child's cells, clotting factors and platelets for reinfusion.
 - Monitor the oxygen tension (PaO₂), pH, and plasma lactate levels to ensure an adequate tissue supply of oxygen.

- (g) Both Amicar and tranexamic acid have been recommended to decrease blood losses during extensive or repeat spine surgery. The selected drug should be initiated after induction of anesthesia but before skin incision and continuously infused until the completion of surgery.
7. If the neurophysiologic recordings show any changes or cannot be obtained for technical reasons, or if the surgeon requires confirmation of spinal cord integrity after application of the distraction and compression apparatus or other manipulation, a wake-up test may be necessary:
- Discontinue inhaled anesthetics or TIVA infusions. Check the neuromuscular monitor for the extent of neuromuscular block (if present). Full antagonism is not advisable because it may lead to complications (see later discussion).
 - Decrease ventilation to return the PaCO₂ to normal levels. Flood the wound with saline to reduce the potential for air embolization. Ask the child to move the toes. (Voluntary dorsiflexion and plantarflexion of the feet confirms spinal cord integrity.) Re-anesthetize the child using midazolam (0.1 mg/kg) and propofol (3–5 mg/kg) IV. Beware of allowing the child to awaken too much; excessive movements may result in dangerous loss of position on the frame, and attempts to breathe spontaneously against the ventilator have been reported to result in air embolism.

Corrective Surgery by the Anterior Approach

For treatment of a curve in the lumbar region, the vertebral column is approached laterally on the convex side of the curvature. Thoracotomy is performed or using VATS; the diaphragm is divided at its peripheral attachments to provide access to the vertebrae.

N.B. This surgical trauma to the diaphragm and chest wall increases the risk of postoperative respiratory insufficiency.

Anesthesia management is the same as for the posterior approach with the following modifications:

- Selective endobronchial intubation of the contralateral lung may be advantageous, permitting easier access to upper thoracic curvatures. (Serial blood gas measurements dictate the feasibility of continuing this technique throughout the procedure, but it is usually well-tolerated.) The Arndt endobronchial blocker has proven useful for this procedure.
- If bilateral ventilation is selected, ventilation of the exposed lung will be impeded by surgical packing and retractors. Periodically, expand the lung fully (to avoid prolonged atelectasis).

Spinal Osteotomy

Spinal osteotomy with wedge excision consists of local resection of deformed vertebrae. This procedure may result in excessive blood loss owing to the proximity of the vertebral and epidural venous plexuses. It may be difficult to control the hemorrhage.

Monitor blood loss by:

1. Gravimetric method (weigh sponges and measure suction losses).
2. Continuous CVP measurement and direct BP readings via an arterial line.
3. Clinical observation (e.g., urine output).

If massive transfusion (>1 blood volume) is required:

1. Use packed cells in recently thawed FFP.
2. Warm all blood and intravenous fluids.
3. Monitor coagulation indices, especially platelets.
4. Order platelet concentrates (1 unit/5 kg body weight) if the platelet count is less than $100,000/\text{mm}^3$. Administer platelets if $<50,000/\text{mm}^3$.
5. Monitor the acid–base status and correct acidosis.
6. If citrate toxicity is suspected (hypotension despite volume replacement, usually during rapid infusion of FFP), administer calcium chloride (10 mg/kg) or calcium gluconate (30 mg/kg) intravenously (repeat as indicated).
7. Activate a massive transfusion protocol (see Chap. 17).

Postoperative Care After Scoliosis Surgery

1. *Idiopathic*: For the child with idiopathic scoliosis and good preoperative pulmonary function:
 - (a) The child should be awake and the trachea extubated before leaving the operating room. Give supplemental oxygen and monitor SaO_2 and respiration during transport to PACU.
 - Check for movement of legs and feet.
 - Check air entry throughout the lungs. (Pneumothorax is a possible complication of spinal surgery.)
 - (b) On arrival in the PACU:
 - Give 40% O_2 by mask.
 - Provide a detailed account of the intraoperative course to the PACU staff.
 - Supplement analgesia as necessary (e.g., morphine infusion, patient-controlled analgesia). There is evidence that morphine requirements may be increased after a remifentanil-based TIVA protocol.

- Obtain plain radiographs of the chest and vertebral column. Check the lung fields, looking especially for pneumothorax.
 - Obtain hemoglobin and Hct values; administer additional blood as indicated.
- (c) The child must remain supine for at least 12 h. Order physiotherapy; encourage breathing exercises.
- (d) Ensure that the child is nursed in a warm environment. (Body temperature usually decreases 1–2 °C during surgery because of large wound exposure, air-conditioning, and so on.)
2. *Neuromuscular*: For the high-risk child with neuromuscular disease and impaired cardiopulmonary function.
- (a) Transfer the child to the PICU intubated and ventilated if indicated. Anticipate that several days of IPPV may be required. X-ray chest on arrival; check endotracheal tube and CVP catheter tip position.
 - (b) Provide a detailed account of the intraoperative course to the PICU staff.
 - (c) Postoperative pulmonary insufficiency may result from:
 - Underlying neuromuscular disease and residual neuromuscular blockade.
 - Acute conditions (e.g., pneumothorax, hemothorax, pleural effusion).
 - Retention of secretions and atelectasis (due to pain, analgesics, and/or immobilization).
 - Perioperative aspiration of gastric contents.
 - Postoperative alteration in thoracic mechanics.
 - Fat embolism syndrome (rare, but may be fatal).
- (d) Other medical complications may follow scoliosis surgery:
 - Syndrome of inappropriate antidiuretic hormone secretion (SIADH).
 - Abdominal conditions; pancreatitis; cholelithiasis; superior mesenteric artery syndrome; ileus; pneumothorax; hemothorax; chylothorax.

Continuing Pulmonary Management

1. IPPV may be required for several days.
2. In some children, extubation may be possible with continuing assisted ventilation by means of noninvasive positive pressure ventilation.

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Urologic Investigation and Surgery

GENERAL PRINCIPLES

Anesthesia risk depends on the state of the child's renal function and any other disease process that may be present.

1. Most children who come for investigation or surgery of the lower urinary tract have good renal function.
2. Many of those who require renal biopsy have mild renal dysfunction (usually insufficient to influence anesthesia risk).
3. All children in renal failure are seriously ill and present multiple problems for the anesthesiologist.
4. Renal disease may be part of a syndrome and therefore requires consideration of all aspects of the condition (see Appendix A).
5. Surgery of the genitalia may have significant psychological effects on small children; effective postoperative pain relief may minimize these effects.
6. Many urologic procedures are now performed endoscopically and the provisions for laparoscopy should be applied (see Chap. 13).

CHILDREN WITH NORMAL RENAL FUNCTION

Anesthesia Management: Minor Procedures

Children require general anesthesia for minor procedures, such as cystoscopy, retrograde pyelography, circumcision, or hypospadias repair and healthy children are almost all managed as outpatients.

Preoperative

1. Oral midazolam is indicated for children who are anxious, especially those who require repeated surgery.

Perioperative

1. Induce anesthesia with sevoflurane or intravenous anesthetics.
2. Maintain anesthesia with N₂O, O₂, and either sevoflurane or isoflurane by mask, laryngeal mask airway (LMA), or tracheal tube.
3. Provide regional analgesia for postoperative pain control whenever possible (see Chap. 5). If the block is performed before the surgery commences, less anesthetic drugs will be required during surgery and awakening will be rapid and pain-free.
 - (a) For circumcision: perform a penile block (see Chap. 5) using bupivacaine *without* epinephrine. Alternatively, ropivacaine may be used for a ring block. EMLA cream may be applied to the incision to augment the block.
 - (b) For hypospadias repair, perform a caudal block (see Chap. 5).

Postoperative

1. Give supplementary analgesia as required; those children with a successful regional block will require few additional medications. Outpatients are usually discharged from the hospital 1 h after recovery. Anticipate the need for additional analgesia at home as the block wears off. For optimal pain relief, instruct parents to administer a suitable analgesic (e.g., acetaminophen) *before* pain occurs.

Anesthesia Management: Major Genitourinary Procedures

Surgical Procedures

Apply anesthesia management as for minor procedures (see previous discussion), plus the following:

1. Use general endotracheal anesthesia with muscle relaxants and controlled ventilation.
2. Be prepared for a major hemorrhage: Establish reliable large-bore intravenous access, measure blood losses carefully, and replace fluids as indicated.
3. Use regional analgesia to provide postoperative pain relief. For example, for reimplantation of ureters, a single-shot caudal provides analgesia for the first few hours or an indwelling epidural catheter provides analgesia for up to 3 days. Intercostal nerve block or preferably a lumbar epidural block provides excellent analgesia after renal surgery and pyeloplasty. Establish the block before surgery.
4. Children with dilated ureters may develop hypertension postoperatively. This may require treatment.
5. For special considerations for Wilms tumor, see Chap. 13.

CHILDREN WITH POOR RENAL FUNCTION OR RENAL FAILURE

These children may have many physiologic and also emotional disturbances.

Special Anesthesia Problems

1. Anemia (usually normochromic, normocytic) may be present, but with standard care under the supervision of a renal unit, this should not be severe.

Anemia in renal failure is caused by:

- (a) Decreased erythropoietin production. If erythropoietin is reduced, the hemoglobin concentration (Hb) will not exceed 7–9 g/dL. Recombinant human erythropoietin therapy may not be effective if severe uremia is present.
- (b) Decreased erythrocyte survival and increased hemolysis.
- (c) Increased bruising and bleeding from increased capillary fragility.
- (d) Iron and/or folic acid deficiency.
- (e) Bone marrow depression due to increased blood urea nitrogen (BUN).

The presence of anemia leads to a compensatory increased cardiac output and increased red blood cell (RBC) 2,3-diphosphoglycerate (2,3-DPG), although the latter is minimal. The P_{50} values are similar to those of children with normal renal function.

Treatment with erythropoietin, supplemental iron, and vitamin C will increase the Hgb to near 10 gm/dL, with possible further increase after a successful transplant. Blood transfusions may improve post-transplantation graft survival, but this remains very controversial.

2. Coagulopathy may be caused by:

- (a) Increased capillary fragility.
- (b) Functional platelet defect (decreased adhesiveness).
- (c) Thrombocytopenia due to bone marrow depression.
- (d) Altered components of the coagulation cascade.
- (e) Drugs (e.g., heparin, acetylsalicylic acid, newer anticoagulants, and altered excretion due to renal failure).

Coagulopathy, if present, is usually of minor significance and does not generally represent a contraindication to epidural catheter placement.

3. Acid–base imbalance:

- (a) Children produce more acid than adults; when urinary ammonia production decreases, a metabolic acidosis predominates and plasma bicarbonate decreases to 12–15 mEq/L. This is compensated to a variable degree by respiratory alkalosis.

- (b) In long-standing stable renal failure, H⁺ displaces Ca⁺⁺ from bone and K⁺ from intracellular fluid.
4. Fluid and electrolyte changes. Children undergoing dialysis (particularly hemodialysis) are likely to be hypovolemic. The timing of the last dialysis is critical as hypovolemia may cause precipitous hypotension in response to induction of anesthesia. Clarification regarding volume status may be obtained from the nephrology team (e.g., the number of liters of fluid that were removed and timing of the most recent dialysis).
- (a) Children with polycystic kidneys or severe pyelonephritis (tubular damage is disproportionately greater than glomerular injury) are “sodium losers”:
- Normotension or slight hypotension is present.
 - Edema is rarely present.
 - Hypokalemia is present in some.
 - Renal function is improved by increasing intake of sodium and water; sodium restriction may rapidly lead to severe hyponatremia.
- (b) Children with glomerulonephritis are often “sodium retainers”; salt retention, hypertension, and edema predispose them to cardiac failure.
- (c) Potassium shifts due to displacement of K⁺ from the cells by H⁺ leads to:
- Increased serum K⁺ concentrations.
 - Depressed excitability of muscles and nerves. This is particularly significant if the cardiac muscle is affected—further sudden increases in serum K⁺ (e.g., with succinylcholine [only if a peripheral neuropathy is present] or increased acidosis) may precipitate cardiac arrest.
- (d) Calcium shifts:
- If displacement of Ca⁺⁺ by H⁺ is prolonged, osteoporosis may develop.
- (e) Anion changes:
- Plasma bicarbonate (HCO³⁻) is decreased.
 - Plasma (SO₄⁻), (HPO₄⁻), and (Cl⁻) are increased.

5. Cardiovascular problems:

- (a) Hypertension may result from abnormalities of extracellular fluid regulation, fluid overload, and derangement of the renin-angiotensin-aldosterone system:
- In children with hypertension secondary to sodium and water retention, this can be controlled conservatively (i.e., by moderate salt restriction).
 - In some children, the BP can be titrated against sodium and water content during dialysis.

- In others, drug therapy with diuretics and vasodilators is necessary.
 - In a few, even large doses of antihypertensive agents fail to control the hypertension (which is probably caused by overproduction of renin). Retinopathy and/or encephalopathy may develop, and bilateral nephrectomy may become necessary.
 - Hypertensive crisis may occur in the perioperative period; an esmolol infusion is the treatment of choice.
- (b) Congestive heart failure and associated pericardial effusion may occur with advanced renal failure, as a result of hypertension, volume overload, anemia, electrolyte imbalance, and the effects of an arteriovenous (AV) fistula:
- Digitalis therapy is difficult to control.
 - Pericardial effusion and tamponade may occur.
- (c) Fatty degeneration of the myocardium may occur secondary to chronic renal failure.
6. Pulmonary congestion:
- (a) The alveolar-arterial partial oxygen pressure difference ($A-a\text{DO}_2$) may be large.
 - (b) Sodium and water retention, left ventricular failure, and hypoproteinemia contribute to the development of “uremic lung.”
 - (c) Pleural effusions may develop.
7. Gastrointestinal disturbances:
- (a) Anorexia, nausea, and vomiting (due to bacterial breakdown of urea to ammonia in the gastrointestinal tract) may aggravate the water, electrolyte, and acid-base imbalance. Gastric emptying may be delayed.
8. Multiple medications:
- (a) Many of these children are receiving long-term steroid therapy with resultant osteodystrophy, Cushingoid state, and glycosuria. Continue steroid therapy perioperatively.
 - (b) Antihypertensive polypharmacy: potential cardiovascular instability under anesthesia must be anticipated. (Do not discontinue beta-blockers before surgery; discontinue ACE inhibitors [e.g., lisinopril].)
 - (c) Diuretic therapy may lead to K^+ depletion and thus to increased susceptibility to cardiac arrhythmias (especially if digitalis is given).
 - (d) Antibiotics (e.g., gentamicin) may prolong the duration of action of non-depolarizing muscle relaxants.
 - (e) Drugs that are highly protein-bound, e.g., azathioprine, may increase the bioavailability of other protein-bound drugs by displacing them from proteins.

- (f) Renal failure may affect the clearance on non-renal cleared drugs by altering hepatic cytochromes and other as-yet unclear mechanisms.
- 9. Reduced immunity (increased risk of infection): it is vital to practice meticulous asepsis.
- 10. Poor quality of life for family and child with potentially major emotional disturbances:
 - (a) Resulting from chronic debilitating disease.
 - (b) Heightened by the uremic state and knowledge of a life-threatening condition.

In summary, these children may have:

- 1. Reduced O₂ carrying capacity, which depends on a stressed cardiovascular system.
- 2. Incipient or apparent cardiac failure:
 - (a) Left ventricular failure if hypertensive, hypervolemic, and anemic.
 - (b) Right ventricular failure (late).
- 3. Greater risk of cardiac arrest (e.g., due to increased K⁺ and acidosis).
- 4. Intolerance of inaccurate administration of blood, other fluids, and electrolytes.
- 5. Cardiovascular instability due to long-term administration of antihypertensive drugs.
- 6. Coagulopathy.
- 7. Low resistance to infection.
- 8. Very low tolerance to further discomfort, however minor (e.g., finger prick, movement from one bed to another).

N.B. Many children with impaired renal function undergo hemodialysis regularly and therefore have an AV shunt or fistula, usually in the arm. Special care must be taken to ensure that this shunt is well-protected throughout the perioperative period. The shunt may be monitored using a gently applied Doppler flowmeter. The child must not be allowed to lie on that limb at any time, and it should not be used for blood pressure determinations.

Preoperative Assessment and Preparation

Pay careful attention to the following physical and psychological aspects:

- 1. Children in a dialysis program are usually dialyzed 12–18 h before surgery. Check the post-dialysis fluid and electrolyte status and body weight.
- 2. Plan ahead so that the child's discomfort is not increased and any necessary disturbances are minimized.
- 3. Psychological preparation and premedication are of special benefit to these children.

4. Check results of laboratory tests:

- (a) Blood transfusions may be given if deemed desirable and may help pre-transplant. Packed RBCs are preferable.
- (b) Serum potassium: values less than 5 mEq/L are acceptable, even in an emergency; levels greater than 6 mEq/L are unacceptable.
 - If the serum K⁺ level is increased, surgery is usually delayed until hemodialysis has been performed.
 - In an emergency, the serum K⁺ concentration can be reduced by giving 5 mL/kg glucose as a 10% solution with 0.1 U/kg of regular insulin (maximum 10 U) followed by an infusion of insulin (0.1 U/kg/h) and D₁₀W with 0.45 normal saline at maintenance rates (onset of effect ~15 min). Acute treatment of hyperkalemia with or without arrhythmia includes IV calcium chloride, bicarbonate, hyperventilation, and inhaled albuterol or salbutamol.
- (c) Acid–base balance:
 - A pH greater than 7.32 is acceptable. If necessary, administer sodium bicarbonate (NaHCO³⁻) for correction of acidosis, even if sodium (Na⁺) levels are increased.
 - Correction must be cautious and gradual. Sudden correction may precipitate tetany or convulsions due to the resulting decrease in the serum calcium (Ca²⁺) level.

Anesthesia Management

1. Pay meticulous attention to details of asepsis.
2. For brief procedures (e.g., insertion of a peritoneal catheter) in a poor-risk child who is cooperative and emotionally stable, use sedation and local anesthesia (1–2% lidocaine without epinephrine; maximum, 3 mg/kg).
3. For all other cases and if in doubt, use general anesthesia.
4. Anesthesia drugs and renal failure:
 - (a) Children with renal failure vary in their response to opioids. Use these with caution. Morphine may exert prolonged effect owing to a failure to excrete/dialyze active metabolites (M6G); meperidine is no longer recommended for children due to the potential for toxic metabolites to cause seizures. In contrast, fentanyl, alfentanil, and sufentanil are relatively safe because their metabolites are inactive. Remifentanil may be the ideal intraoperative opioid for these children.
 - (b) Propofol and thiopental should be used cautiously and in reduced doses; less protein binding increases the free active fraction of drug. Ketamine and etomidate may cause less hypotension.

- (c) Inhalational anesthetics are eliminated via the lungs and hence are most useful but must be administered cautiously. Fluoride nephrotoxicity does not occur with the current ether anesthetics.
- (d) Succinylcholine may be used as a single dose, provided a peripheral neuropathy is not present. The serum potassium (K^+) concentration should be 5.5 mEq/L or less (serum K^+ increases 0.5–1 mEq/L after succinylcholine in children with renal failure; this produces no ECG changes because the hyperkalemia is chronic. If a peripheral neuropathy is present, however, succinylcholine may cause greater increases in serum K^+ and arrhythmias). Succinylcholine should only be used if a rapid sequence induction is deemed necessary and should be preceded by a small dose (~10% of the intubating dose) of a non-depolarizing relaxant.
- (e) Muscle relaxants, non-depolarizing: *cis*-atracurium and vecuronium are drugs of choice. Rocuronium has a slower onset of action, but a similar duration of action in children with and without renal failure. Pancuronium and gallamine are partially or completely excreted by the kidneys and should be avoided.
- (f) Local anesthesia drugs have not been extensively studied in children with renal failure; they may be used in normal doses for “single-shot” techniques. Ropivacaine appears safe for long-term infusions as its pharmacokinetics are unaffected by renal failure.

Preoperative

1. Dose requirements for drugs in children with renal failure are more variable than in those with normal renal function; titrate doses carefully.
2. Do not discontinue antihypertensive drugs (except ACE inhibitors).
3. Premedicate as required (e.g., PO or IV midazolam).
4. Check all medications that have been given and note their last dose before surgery.
5. Check the location of a shunt or fistula. Avoid any pressure to this area (*including blood pressure cuff*), and monitor function (Doppler flowmeter).
6. Ensure that all supportive drugs are available in the operating room.
7. Ensure that adequate supplies of blood and other fluids are available (including washed cells if and when indicated).

Perioperative

1. Give 100% O_2 by mask.
2. Apply monitors:
 - (a) Precordial stethoscope.
 - (b) ECG and pulse oximeter.
 - (c) Automated blood pressure—*do not use a limb with a shunt or fistula*.

3. Ensure that the limb with the shunt or fistula is easily accessible. Monitor function throughout the procedure.
4. Induce anesthesia with propofol (2–4 mg/kg), thiopental (2–3 mg/kg) or, if dehydrated or recently dialyzed, consider ketamine (1–2 mg/kg) or etomidate (0.3 mg/kg), followed by N₂O/O₂ and sevoflurane.
5. For tracheal intubation:
 - (a) Do not give succinylcholine if a peripheral neuropathy is present or there is an increased serum K⁺. Otherwise consider pretreatment with a low-dose non-depolarizing agent (~10% of the intubating dose). Always limit succinylcholine to a single dose.
 - (b) Alternatively, give cisatracurium, rocuronium, or vecuronium in the usual doses for tracheal intubation.
6. Maintain anesthesia with N₂O/O₂ and titrated doses of inhalation agent with a non-depolarizing muscle relaxant.
7. Controlled ventilation is recommended to maintain the arterial carbon dioxide pressure (PaCO₂) at the usual level for that particular child. Use moderate hyperventilation to compensate for metabolic acidosis and to encourage K⁺ movement back into the cells.
8. Administer fluids to ensure adequate blood volume for satisfactory BP, peripheral perfusion, and function of an AV fistula or shunt.
 - (a) Give balanced salt solutions, e.g., plasmalyte or lactated Ringer's solution to replace the preoperative deficit and for perioperative maintenance. Normal saline causes more acidosis and a greater increase in serum K⁺ than lactated Ringer's solution.
 - (b) For small blood losses, replace with balanced salt solution.
 - (c) For significant blood losses, replace with washed RBCs and salt-poor albumin.
 - Check Hb and hematocrit (Hct); keep Hct below 30 %.
 - Avoid over-transfusion.
9. At the end of surgery, antagonize muscle relaxants and extubate the trachea when full strength has returned and the child is awake.

Postoperative

1. Ensure good ventilation and oxygenation.
2. Carefully titrate opioid doses (repeated doses of morphine and its metabolites accumulate in renal failure; M6G cannot be removed by dialysis); monitor the effect and give supplements if necessary.
3. Ensure that the shunt or fistula is functioning; record this fact.
4. Check Hb, electrolytes, and blood gases.
5. Consult a nephrologist for continuing care.

RENAL TRANSPLANTATION

Transplantation offers the chance of a relatively normal life for the child with chronic renal failure. Organs for transplantation are in very limited supply; meticulous anesthetic management is essential to maximize the chance for graft survival.

Anesthesia Management

Preoperative

1. General management is the same as for children in renal failure.
2. Discuss with the nephrologist and urologist to ascertain the child's exact present status (state of hydration, electrolyte and acid-base status, renal function, cardiopulmonary state, coagulation issues).
3. If the child is not in optimal condition (e.g., volume overload), surgery should be postponed until after dialysis but with the objective of implant within 24 h.
4. Review the immunosuppressive therapy plan (i.e., drug, dose, timing). A typical plan includes administration of cyclosporin, methylprednisolone succinate (Solu-Medrol), and azathioprine; an antibiotic is also given.

Perioperative

1. General management is the same as for children in renal failure.
2. After induction of anesthesia:
 - (a) Insert a central venous line.
 - (b) Check its position with a pressure tracing and/or radiograph.
 - (c) Maintain the central venous pressure (CVP) at an acceptably high level to ensure diuresis (8–12 mmHg). Lactated Ringer's solution is the preferred initial maintenance fluid (*it causes less acidosis and hyperkalemia than Normal Saline*).
 - (d) The objective is to adequately replace any existing deficits and ongoing losses.
3. An arterial line is not essential in uncomplicated cases. Leaving the radial artery untouched may be advantageous in case the child needs another shunt in the future.
 - (a) If an external (Scribner) shunt is available, the arterial end may be used for monitoring and the venous end as an infusion route. (Use extremely careful aseptic technique.)

- (b) If an arterial line is indicated, insert the catheter in the radial artery (in the arm opposite to the fistula), do not use a cannula larger than a 22 gauge, and remove the catheter as soon as possible to minimize damage to the artery.
- 4. Provided no significant coagulopathy is present, an epidural catheter may be inserted to augment surgical anesthesia, improve hemodynamic stability, and ensure good postoperative analgesia.
- 5. Transfuse with packed RBCs to obtain an Hct of 35–40 % at the end of surgery. Children with chronic renal failure tend to lose third-space fluid extensively; greater Hct and colloid administration may limit this effect and improve graft perfusion.
- 6. During vascular anastomosis and before clamp release, give 1 mg/kg of furosemide and 1 g/kg of mannitol IV. Anticipate a decline in blood pressure as the clamps are removed, and prepare for rapid fluid infusions. Some surgeons will request an RBC transfusion just before releasing the clamp.
- 7. Systolic blood pressure (100–120 % of preoperative value) and a CVP > 14 cm H₂O are preferred before the clamps are removed to initiate and maintain renal perfusion and function. Lighten the level of anesthesia and use a dopamine infusion 5 µg/kg/min as necessary. A greater systolic pressure at this time increases the graft perfusion and facilitates good early function and graft survival. Optimal results may be expected if the graft functions within 10 min.
- 8. The solution used to preserve the kidney has a high potassium concentration; hyperkalemia and acidosis after release of the clamps have on rare occasions resulted in cardiac arrest. This may be a greater danger in small children.
- 9. Placement of an adult kidney in a small infant can be problematic. The kidney should be well-flushed by the surgeon before it is implanted. Fluids must be rapidly infused as the clamps are released to fill the vascular space of the graft. The CVP should be increased to 15–20 mmHg in anticipation of unclamping. The danger of hyperkalemia (due to preservative fluid) and acidosis (due to clamping of the aorta or iliac artery) is increased. Check the acid–base status frequently and give calcium chloride and sodium bicarbonate as necessary. Dopamine infusions may need to be increased.
- 10. Maintain the CVP at an increased level (i.e., up to 15–18 mmHg) to produce a good urine output. If the CVP is adequate but urine flow is still low:
 - (a) Give furosemide (1–4 mg/kg IV).
 - (b) If necessary, add mannitol (0.5–1 g/kg).
- 11. Anticipate the need to infuse large volumes of fluid (three to five times normal, i.e., 10–20 mL/kg/h) to compensate for third-space losses.

12. At the end of surgery, determine serum electrolyte concentrations:
 - (a) If urine output is adequate, serum K⁺ should be within the normal range.
 - (b) If the serum K⁺ is greater than 6 mEq/L and urine output is poor, continue to hyperventilate the lungs and plan to arrange for dialysis or therapy with glucose and insulin.
13. With such an aggressive approach to fluid management, pulmonary edema may threaten. In such cases, continue with controlled ventilation into the postoperative period.

Postoperative

1. General management is the same as for children in renal failure. Ensure good postoperative analgesia; epidural analgesia is ideal.
2. The pulmonary status of all children should be monitored by pulse oximetry, blood gases, and chest radiography as indicated. Small infants with large implanted kidneys may be at risk for pulmonary complications (especially atelectasis). This may be a combined result of the abdominal surgery, the mass of the implanted kidney, and aggressive fluid therapy. The need to “push fluids” to ensure diuresis may result in pulmonary edema. Very close monitoring in the PICU should be ensured.
3. Renal function in the transplanted kidney is as follows:
 - (a) Glomerular function is initially normal but wanes during the first 48 h as the kidney swells. Increased intravenous fluids are required to maintain diuresis at that time.
 - (b) Some degree of tubular damage is always present resulting in diuresis and loss of sodium; replacement of sodium and water is required. Other electrolytes must be infused as indicated by serum studies.
 - (c) Declining urine flow after 48 h despite fluid loading is indicative of mechanical problems (vascular) or rejection of the transplant.
4. Hypoglycemia has been described as a late complication after renal transplantation in small children. This may be secondary to the use of β-blocking drugs.

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Trauma, Including Acute Burns and Scalds

Children are commonly involved in accidents; trauma is the leading cause of death between 1 and 14 years of age. Even if the injury is relatively minor, some children require emergency anesthesia, the potential dangers of which may overshadow the injury.

Most children injured in accidents were previously healthy. Therefore considerations of past health are usually less important than in the adult. However, a complete medical history must be obtained as soon as possible. From the time of arrival in the emergency department, the anesthesiologist should be included in the treatment team. The anesthesiologist can contribute to immediate care while evaluating the child's condition for anesthesia and the need for further continuing care.

Basic trauma care involves three steps: primary survey, secondary survey, and definitive care. The primary survey involves initial resuscitation and stabilization: (a) airway, (b) breathing, (c) circulation, (d) disability (neurologic injury, modified Glasgow coma scale), and (e) exposure. Completely undress the child for full evaluation, but institute strategies to maintain thermoneutrality. The secondary survey focuses on additional, less life-threatening injuries (usually additional diagnostic testing). Definitive care involves transfer to ICU or OR.

MAJOR TRAUMA

Diagnosis and treatment must proceed rapidly and simultaneously. Vigorous resuscitative measures must be continued without interruption during anesthesia. The common major problems for the anesthesiologist include:

1. To secure and maintain a safe, reliable airway and to optimize ventilation and oxygenation
2. To restore adequate organ perfusion and oxygen delivery and crystalloid, colloid, and blood replacement as needed
3. To optimize cerebral perfusion pressure in children with a head injury
4. To maintain body temperature

N.B. Although injuries may appear limited to a single anatomic site or body system, consider the possibility of serious injuries elsewhere, e.g., the fractured femur may be obvious, but the as yet undiagnosed ruptured liver could be life-threatening.

Initial Urgent Procedures

1. Ensure a safe and protected airway and optimize ventilation and oxygenation.
2. The cardiovascular status must be determined:
 - (a) Correct hypovolemia.
 - (b) Maintain or restore cardiac output.
3. Send blood for type and cross-match; immediately advise the blood bank if a massive transfusion protocol must be activated (see Table 17.1).

Establishing the Airway

Airway obstruction is common in head and facial injuries and can have a disastrous effect on the outcome:

1. Oxygen should be given via face mask to all children with head injuries as soon as it is available, preferably at the trauma site.
2. For those with depressed consciousness, if simple positioning does not provide a clear airway or if the gag reflex is absent, intubate the trachea without delay.
3. Do not insert oropharyngeal airways in unconscious children. Resistance to ventilation is greater than with a tracheal tube, and they do not prevent aspiration of gastric contents.

Table 17.1 Massive transfusion protocol for possible immediate use

Protocol	Number of units to be issued			
	Red cell units	Plasma	Platelets	Cryoprecipitate
Adult	6	4	One 5-pack	One 5-pack
Pediatric				
0–10 kg	1	1	1	1
11–20 kg	2	2	2	2
21–30 kg	3	3	3	3
41–50 kg	4	4	4	4
>50 kg	5	5	One 5-pack	One 5-pack

Modified from: <http://surgery.med.umich.edu/pediatric/trauma/protocols/MassiveTransfusionProtocol4113.pdf>

4. Injury of the cervical spine must be suspected in all trauma; immobilize the neck using sandbags, a plaster shell, or a bean bag.* Avoid moving the neck. Note that a cervical spine injury may occur without any radiologic evidence (known as SCIWORA or Spinal Cord Injury WithOut Radiographic Abnormalities). Optimal airway management is the first priority, although all airway manipulations should assume the presence of a cervical spine injury (see later discussion). A supraglottic device (e.g., laryngeal mask airway) may be lifesaving for a difficult airway, but be aware that the cervical spine stabilization is still critical during placement.
5. Consider possible foreign bodies (i.e., teeth, bone fragments, food) in the mouth, pharynx, trachea, or bronchi, especially with facial injuries.
6. Assume that all trauma patients have a full stomach; perform a rapid-sequence intubation (RSI) to secure the airway. Succinylcholine does not increase intragastric pressure and the risk of regurgitation or raise intracranial pressure (ICP) in young children. The advantages of rapidly securing a clear airway and instituting controlled ventilation are the most important consideration. Beware of neck injury; stabilize the spine, but do not exert traction on the neck. (Remember: a cervical spine injury is a relative contraindication to cricoid pressure.) Assure that two functioning laryngoscope handles and size-appropriate blades are available as well as two suction sources, should one become blocked with food or vomit.

“Awake” laryngoscopy and intubation can markedly increase ICP, which may be detrimental in the head-injured child. In children with suspected increased ICP, deep sedation/general anesthesia is required for an RSI: give IV lidocaine 1–2 mg/kg, propofol 2–3 mg/kg (or thiopental 5 mg/kg), atropine 0.02 mg/kg, and succinylcholine 2 mg/kg for tracheal intubation. If succinylcholine is truly contraindicated, then substitute with 1.2 mg/kg rocuronium. If the child is hypovolemic, ketamine (use cautiously with increased ICP) and etomidate or midazolam (with IV lidocaine and fentanyl for the latter two) are acceptable alternatives to induce anesthesia. Airway adjuncts such as the GlideScope, ProSeal-LMA, and fiberoptic bronchoscope should be immediately available for airway rescue.

7. Vomiting and aspiration may occur. Immediately after intubation check PetCO₂ and auscultate air entry to all lung regions; suction the bronchi via the tracheal tube if necessary. Examine chest radiographs for tracheal tube position and evidence of lung pathology or aspirated foreign body.
8. Pass a gastric tube to decompress the stomach, especially in children with chest or abdominal injuries. Even children with relatively minor injuries often swallow a great deal of air causing gastric distention that may interfere

*Vac Pac surgical positioning system, Ventech Healthcare Inc., Toronto.

with ventilation and predispose to regurgitation and aspiration. Do not pass a tube through the nose in children who may have a basal skull fracture.

9. Be alert to the possibility of pneumo-/hemothorax.

Intravenous Therapy

Large-bore intravenous access must be established in the upper limbs or neck in children with injuries at or below the level of the thorax because flow through the inferior vena cava may be (or become) compromised. If the source of hemodynamic instability is unknown, establish IV access in both upper and lower extremities. *All intravenous fluids should be warmed since hypothermia rapidly develops in the injured child.*

Percutaneous cannulation of large veins, using rapid infusion catheters is preferable. If successful venipuncture is taking more than 90 s, consider the use of an intraosseous needle (e.g., Easy-IO). Internal jugular (IJ) or subclavian lines may be inserted if no other venous access is available; beware of further worsening the cardiorespiratory status if a pneumothorax or hemothorax occurs, and avoid IJ lines in head-injured children. A central venous catheter will be useful during the further management of the child, but it may be inserted after the initial acute fluid resuscitation.

Clues to blood volume status are as follows:

1. Cardiovascular indices:
 - (a) Tachycardia, i.e., a heart rate in excess of 140 beats/min in infants or 100 beats/min in older children, suggests hypovolemia.
 - (b) In infants, the systolic blood pressure (BP) varies directly with the intravascular volume and is a very good guide to volume status.
 - (c) In adolescents, as in adults, hypovolemia stimulates early constriction of venous capacitance vessels. Therefore, the systolic BP may remain near normal despite a loss of up to 20% of the blood volume. The central venous pressure (CVP) may also be unchanged initially. When vasoconstriction can no longer compensate and maintain the venous return to the heart, rapid decompensation may occur. At this point, the CVP decreases and becomes a more reliable guide to the adequacy of replacement of the blood volume. A 2–3 mmHg decrease in CVP may represent a loss of as much as 25% of the circulating blood volume in a healthy, supine child.
2. General appearance: Confusion, irrational behavior, pallor, mottling, sweating, and coolness of the skin, especially over the extremities, are signs of hypovolemia. Large differences between the skin and core body temperature indicates vasoconstriction and hypovolemia.
3. Urine output: Urine output should be assessed; urine flow of ≥ 1 mL/kg/h indicates adequate renal perfusion and volume replacement.

4. Acid-base assessment: Metabolic acidosis (lactic acidosis) may result from impaired organ perfusion and is a confirmatory sign of a low circulating blood volume. This acidosis may be corrected by adequate volume replacement. Sodium bicarbonate administration is not recommended except in the presence of severe acidosis that may compromise cardiac contractility (i.e., pH <7.2 despite normocapnia or hypocapnia).

Selection of Fluids

The initial fluid resuscitation should consist of balanced salt solutions rather than blood products. If at all possible, do not transfuse non-cross-matched universal donor blood. (A rapid cross-match can be performed in 20 min.) If blood loss is massive, and it has accumulated in the chest cavity, consider autotransfusion.

A balanced salt solution (e.g., lactated Ringer's solution) expands the circulating blood volume, but if used to excess (more than 100 mL/kg), it may lead to pulmonary insufficiency and/or dilutional coagulopathy. An initial bolus of 20 mL/kg is appropriate for the hypotensive child and may be repeated. If the child has a head injury, use normal saline (isotonic) to reduce the potential for cerebral edema. Dextrose-containing solutions should not be used (except, very rarely, to treat documented hypoglycemia); hyperglycemia may occur and increase the severity of neurologic damage (blood glucose >200 mg/dL) with head injury.

Expanding the blood volume with 5% albumin may be very effective in the immediate treatment of hypovolemia, but if large quantities are infused, "capillary leak" into the lungs may impair pulmonary function later.

Other plasma substitutes may be used (e.g., dextran 70, hetastarch). Note that dextrans may impair coagulation and interfere with cross-matching; do not exceed 7 mL/kg. Hetastarch does not impair coagulation, and may be used in volumes up to 25 mL/kg. (**N.B.** The starches have long residence times in the reticuloendothelial system in humans; limited data are available for their use in children.)

Indications for Blood Transfusion. It is seldom possible to measure the blood loss associated with a trauma. Large amounts may be lost from the intravascular volume but remain within hematomas (e.g., retroperitoneal) or body cavities. Volume replacement must therefore be judged on a case-by-case basis using the signs and symptoms listed above. We recommend that sufficient fluid be replaced to restore euvoolemia (with normal vital signs) and that blood be given in volumes sufficient to maintain the hematocrit (Hct) at ~21–25%.

The young child who is showing obvious signs of hypovolemia (pallor, sweating, hypotension) has lost at least 25% of the blood volume. In the emergency situation, the weight of the average child may be predicted from the child's age using the following formulae (Table 17.2) and assume the blood volume to be 70 mL/kg; 25% of this total is the initial volume to be rapidly replaced and then

Table 17.2 Weight prediction from child's age

Age (years)	Predicted weight (kg)
<1	$0.5 \times \text{age (months)} + 4^a$
1–7	$2 \times \text{age (year)} + 9$
8–12	$3 \times \text{age (year)}$

^aGraves L. et al. Resuscitation 2014;85:392–6

reassess the child's response. The need for additional fluid boluses or continuous infusion to maintain the BP, or deterioration after an apparently stable period, indicates persistent bleeding.

Children who have lost greater than one blood volume will require fresh-frozen plasma (FFP) to prevent dilutional coagulopathy. Packed cells resuspended in plasma (preferably recently thawed FFP) may be optimal.

Massive Blood Transfusion. In children, >30 mL/kg packed red blood cells transfusion with ongoing blood losses warrants activation of a massive transfusion protocol. Transfusions of this order of magnitude may be required in severely injured children, e.g., after thoracoabdominal injury. One practical massive transfusion protocol (developed at the University of Michigan) for children alerts the blood bank to send set packs of blood products based on the child's weight (Table 17.1).

Serious problems begin to develop after the rapid infusion of 200 % of the estimated blood volume (i.e., transfusion of ~140 mL/kg). These problems include:

1. Hypothermia and accompanying cardiac arrhythmias. Warm all blood and fluids to 40 °C.
2. Coagulation problems:

(a) Dilutional thrombocytopenia and impaired platelet function:

1. Check platelet count after each 50 % blood volume replacement.
2. If the platelet count is less than 50,000/mm³, administer platelet concentrates (0.1–0.3 units/kg). In general, ~40 % of the initial platelet count is lost with the first blood volume, ~20 % with the second blood, and ~10 % with the third, e.g., after three blood volumes only ~30 % of the initial platelet count remains.

(b) Deficiency of coagulation factors:

1. Measure prothrombin time (PT) and partial thromboplastin time (PTT).
2. If these are prolonged, give FFP (20 mL/kg). (Generally this occurs after loss of 1–1.5 blood volumes; for blood loss, >1.5 blood volumes give one-third of the further volume replacement as FFP.)

(c) Disseminated intravascular coagulation (DIC):

1. Likely if bleeding increases at all sites (i.e., old venipuncture sites).
2. Measure PT, PTT, clot lysis time, fibrinogen, and fibrin split product levels.
3. Prolonged PT and PTT, low fibrinogen level, and the presence of fibrin split products suggest DIC.
4. If DIC is suspected, enlist the aid of a hematologist if possible. Treatment must include removal of the cause (i.e., correction of hypovolemic shock and hypothermia), replacement of coagulation factors, and possibly heparinization.

3. Acidosis:

- (a) Check acid-base status frequently.
- (b) Correct severe metabolic acidosis with sodium bicarbonate (provided ventilation is not compromised).

4. Citrate toxicity (due to infusion of citrated blood or plasma):

- (a) May cause more problems in infants and small children than in adults.
- (b) Results in hypotension that persists despite adequate volume replacement. (**N.B.** Remember that FFP and platelet suspensions contain more citrate per unit volume than whole blood. Significant hypocalcemia may occur when infusions exceed 1 mL/kg/min FFP.)
- (c) May be diagnosed by measuring the ionized Ca^{++} concentration or by a prolonged rate-corrected QT interval on the ECG. If hypotension appears unresponsive to volume replacement, a therapeutic test of IV calcium chloride (10 mg/kg) is justified.
- (d) Correct by administering 10% calcium chloride 10–30 mg/kg slowly under ECG control. Frequent small boluses (5–10 mg/kg calcium chloride or 15–30 mg/kg calcium gluconate) during rapid FFP infusions prevent serious hypocalcemia.

5. Serum potassium disturbances: Monitor serum K^+ levels periodically. Serious hyperkalemia is likely in the presence of a low cardiac output, after transfusing old cold blood through a central line and the use of a rapid infusion device. The cardiac manifestations of hyperkalemia (peaked T waves, ventricular irritability, ventricular tachycardia) may be treated acutely by slow administration of intravenous calcium chloride 10–30 mg/kg, sodium bicarbonate 1–2 mmol/kg, hyperventilation, administration of an β -agonist, and, if more severe, with glucose, insulin, and Kayexalate. Contrary to expectations, hypokalemia may also occur after massive transfusions, although it is rare.

6. Posttraumatic pulmonary insufficiency. This is characterized by progressively decreasing pulmonary compliance, impaired gas exchange, and radiographic findings of diffuse infiltrates. The following factors may contribute:
- (a) Excess use of balanced salt solutions and/or albumin:
 1. Give diuretics (furosemide 1 mg/kg) if indicated.
 - (b) Microembolization of the pulmonary vessels by infused particulate matter (platelet or leukocyte clumps):
 1. Filter all blood products prior to administration.
 - (c) Damage to alveolar-capillary membrane:
 1. This results in low-pressure pulmonary edema.
 2. Large doses of steroids may help to prevent this.

Autotransfusion. Autotransfusion can be lifesaving for some trauma patients. Advantages are the ready availability of warm absolutely compatible blood. Do not use blood from the abdominal cavity if bowel injury is suspected.

Head Injury

Head injury is a major cause of pediatric mortality and morbidity, much of which might be reduced by early and efficient medical intervention. Children with head injury are less likely than adults to develop a mass lesion but are more likely to develop intracranial hypertension, secondary to diffuse hyperemia and edema. This secondary brain injury, which occurs after the primary trauma, must be minimized if recovery is to be optimized. Early aggressive treatment is essential to ensure cerebral oxygenation, control ICP and cerebral perfusion pressure (which is the mean arterial blood pressure—ICP), and minimize cerebral edema. The Glasgow coma scale (Table 17.3) is the most common means of initial assessment.

Glasgow Coma Scale and Pediatric Modifications (Table 17.3)

If consciousness is depressed (GCS <8), airway obstruction may seriously compromise the prognosis. The first priority must be to ensure an absolutely clear airway, excellent oxygenation, and optimal ventilation. If there is any doubt about the child's ability to maintain a patent airway and adequate ventilation:

1. Give oxygen and intubate the trachea without delay; use a cuffed oral tube. Suitable drugs should be given to obtund the cardiovascular responses to intubation (see previous discussion). Nasal tubes (and nasogastric tubes) are

Table 17.3 Glasgow coma scale and pediatric modifications

Standard	Score	Pediatric
Eye opening		
Spontaneous	4	Spontaneous
To speech	3	To speech
To pain	2	To pain
None	1	None
Verbal response		
Oriented	5	Coos, babbles (age appropriate)
Confused	4	Irritable, cries
Inappropriate words	3	Cries to pain
Incomprehensible sounds	2	Moans to pain
None	1	None
Motor response		
Follows commands	6	Spontaneous movement
Localizes to pain	5	Withdraws to touch
Withdraws to pain	4	Withdraws to pain
Abnormal flexion	3	Abnormal flexion
Extensor response	2	Extensor response
None	1	None

Utilize best score from each category for possible total score of 3–15

contraindicated in children with basilar skull fractures; perforation of the cribriform plate may occur. Beware of the possibility of SCIWORA; see later discussion.

2. Control ventilation to produce normocapnia pending further evaluation of the child's injuries. Hyperventilation should be avoided ($\text{PaCO}_2 < 30 \text{ mmHg}$) because it may compromise cerebral perfusion. PaCO_2 should preferably be checked by arterial blood gas analysis as end-tidal CO_2 may be misleading, particularly in children with associated chest trauma.
3. Continue anesthesia as required, preferably with intravenous agents (e.g., propofol, opioids, and muscle relaxants). Inhalational agents should be limited to $<1 \text{ MAC}$ to prevent possible increases in ICP. MAP should be maintained to preserve CPP. Avoid sufentanil, which increases ICP in head-injured patients.
4. Stabilize the hemodynamic state to ensure an optimal CPP. Glucose-free, isotonic fluids (normal saline) should be infused cautiously; excessive fluid therapy contributes to cerebral edema. If a vasopressor is required neosynephrine is the drug of choice.

The use of CT and MRI permits accurate anatomic mapping of traumatic lesions and removes the need for exploratory burr holes. CT or MRI is generally recommended for any child with GCS less than 13 or with vomiting. The characteristic appearance of diffuse cerebral damage on the scan can obviate the

need for craniotomy and permit early specific monitoring and therapeutic measures, such as the following:

1. ICP monitoring—most commonly includes the use of an extradural bolt connected to an external transducer. This is usually recommended for any child with a GCS<8. Once ICP monitoring is established, the CPP can be determined and maintained at an optimal level (see below).
2. Initiate treatment to control ICP and CPP:
 - (a) Optimal positioning, 35°–45° head up and face centrally forward.
(No internal or external jugular lines in children with head injuries.)
 - (b) Diuretics—mannitol and/or furosemide.
 - (c) Consider hypertonic saline infusions.
 - (d) Intermittent boluses of propofol may be effective in controlling ICP and seizures, but the use of infusions is controversial due to the potential of propofol infusion syndrome. Barbiturates (thiopental 2–4 mg/kg) (no longer available in the USA), which may be more effective in controlling ICP in children than in adults, may also be useful to control seizures.
 - (e) Controlled ventilation, preferably adjusted on the basis of measurements of ICP, and CPP. Excessive hyperventilation ($\text{PaCO}_2 < 30 \text{ mmHg}$) may be detrimental and is no longer recommended.
3. Maintain optimal CPP (>40 mmHg in 0–5-year-olds, >50 in 6–17-year-olds), hemoglobin values, and arterial oxygenation: Glucose-free isotonic or hypertonic fluids should be used for volume expansion. Dopamine may be required to treat hypotension (but see caution in later discussion). Hyperglycemia should be avoided because it may exacerbate secondary brain injury. Fluid therapy must be guided by constant monitoring of physiologic and biochemical variables. Endocrine functions (especially pituitary/hypothalamic) may be disturbed after head injury, which may lead to:
 - (a) Diabetes insipidus, with a large urine volume and hypernatremia.
 - (b) Inappropriate secretion of antidiuretic hormone (SIADH) with oliguria and hyponatremia.
 - (c) Hyperglycemia is common after head injury, secondary to catecholamine release; the value of treating this is controversial. Hypoglycemia may develop later and should be corrected.
4. Control of seizure activity, as evident clinically or by EEG monitoring.
5. Control body temperature:
 - (a) To prevent hyperthermia, which commonly follows brain injury
 - (b) Studies regarding the risk/benefits of moderate (32–33 °C) hypothermia in children with severe traumatic brain injury are conflicting

6. When all else fails to control ICP, a decompressive craniectomy may be required. If this is performed, be alert to the possibility of sudden severe hypotension as the dura is opened—especially in cases with a major midline shift.

Caution: Head injuries do not normally cause shock. When anesthetizing or caring for the child with a head injury, be alert for evidence of other injuries. Do not ascribe signs of hypovolemic shock (i.e., tachycardia, hypotension) to the head injury. If such signs are present, search for bleeding from wounds in the scalp and/or other sites (intra-abdominal, intrathoracic, or in the limbs). Be constantly aware that hemorrhage at another site may have been overlooked.

Cervical Spine Injury

SCIWORA is more common than previously thought in children. High cervical spine injury often occurs in young children during motor vehicle accidents. This contrasts with the lower cervical and upper thoracic spine injuries that occur in older patients. Severe high cervical spinal cord injury may result in apnea and cardiac arrest; this possibility should be considered in children with absent vital signs.

The anesthesiologist must ensure a rapid, safe tracheal intubation of the injured child while minimizing additional injury to the spinal cord. Early relief of an obstructed airway and controlled ventilation are essential for optimal recovery from severe head injury. Current opinion suggests that some children may experience immediate damage to the spinal cord at the time of their accident; the outcome from such established injuries will not be altered by the technique of tracheal intubation chosen.

Techniques of intubation other than by direct vision are more difficult (and often more prolonged) in children. Careful direct oral intubation can be performed without causing damage to the spinal cord. Hence, it is recommended that careful direct laryngoscopy and intubation be performed in injured children. Unnecessary head and neck movement should be avoided by using manual in-line stabilization (avoiding traction on the neck). Airway adjuncts such as the GlideScope may be particularly valuable to minimize neck movement during laryngoscopy.

Thoracoabdominal Injury

Blunt abdominal trauma is most commonly managed conservatively in children. Bleeding from the spleen or liver can be assessed by scanning techniques and usually resolves spontaneously; blood transfusion requirements are generally not increased during conservative management. Surgery is necessary for penetrating wounds and if continued bleeding causes hemodynamic instability; laparoscopy may be employed to assess intra-abdominal injuries.

Initial assessment must assess possible physiologic consequences of the injury and the additional effect of anesthesia on the child's condition. In those with intrathoracic injuries, the anesthesiologist's prime concerns are the pulmonary contusion/trauma, the hemo-/pneumothorax, the amount of blood lost, and the problem of securing a safe airway (possible tracheal disruption).

Special Anesthesia Problems

1. Major hemorrhage requiring massive blood transfusion (see Chap. 4 and Table 17.1).
2. The possibility of a full stomach (food or blood). Always use a cuffed tube in children with thoracoabdominal injuries.
3. Impaired cardiorespiratory function (in children with diaphragmatic or thoracic trauma); significant pulmonary contusion and serious arrhythmias may follow blunt thoracic trauma.

Immediate Management

1. Prepare for transfusion:
 - (a) Insert a wide-bore (a rapid infusion catheter is preferable) cannula into an upper limb or neck vein, by cutdown if necessary.
 - (b) Send blood for type and cross-match; immediately advise the blood bank if massive transfusion is likely.
 - (c) Insert a central venous cannula via a second upper limb or neck vein (use the femoral vein if a head injury is present) for measurement of CVP and to provide an alternative route for transfusion and vasopressors if necessary.
 - (d) Place a urine catheter to assess ongoing renal perfusion and function (blood in the urine may indicate renal fracture or disruption).
2. Assess the extent of hypovolemic shock.
3. Infuse appropriate solutions with the use of a blood warmer (boluses of 20 mL/kg balanced salt solution initially).
4. Never infuse large volumes of cold fluids (or blood) via a central vein.

Anesthesia Management

Preoperative

1. Before induction of anesthesia, restore euvoolemia. (In some cases, this may not be possible until the source of bleeding is controlled surgically.)
2. Assure adequate supplies of blood in the operating room (OR).
3. Premedication is usually unnecessary, although low-dose opioid may be used.

4. In children who are hypovolemic, titrate all IV drugs to effect.
5. Ensure availability of a rapid infusion device that will allow rapid administration of warmed blood and fluid.
6. Prepare appropriate vasoconstrictors and administration pumps (often through a separate IV with multiple stopcocks and a continuous carrier solution).
7. Prepare transducers for arterial and central venous access lines.
8. Consider a processed EEG monitor to estimate trends for depth of anesthesia although these are only useful in teenagers and often display paradoxical readings after ketamine and sevoflurane.

Perioperative

Induction:

1. Prepare and check all necessary equipment (two blades, two handles, two suctions) and drugs for RSI and tracheal intubation (see above).
2. Before induction of anesthesia, reassess the child's physiologic status.
3. Preoxygenate as tolerated.
4. Check patency of intravenous lines.
5. Connect monitoring equipment.
6. Consider the possibility of persistent hypovolemia:
 - (a) If hypovolemia has been corrected, induce anesthesia with propofol (up to 3 mg/kg) or thiopental (up to 4 mg/kg), atropine (20 µg/kg), and succinylcholine 1–2 mg/kg or rocuronium 1.2 mg/kg if there is a contraindication to succinylcholine. Inject these agents directly into a rapidly running IV cannula or flush drugs to avoid the delayed drug transit.
 - (b) For emergent surgery in children who are hypovolemic, induce anesthesia with ketamine (2 mg/kg) or etomidate (0.3 mg/kg).
7. Position the child supine and horizontally for intubation (to facilitate rapid insertion of a cuffed tube).
8. Do not inflate the lungs before laryngoscopy (ventilation may precipitate vomiting). Cricoid pressure may be considered, although it is contraindicated if cervical spine injury may be present (see management above). Release cricoid pressure after the tracheal tube is placed and the cuff inflated.
9. Do not give any drugs (except atropine and possibly low-dose fentanyl and midazolam) to moribund patients before intubating the trachea.

Maintenance

1. Give O₂, fentanyl, and a non-depolarizing neuromuscular relaxant (vecuronium, rocuronium, or pancuronium). N₂O may cause distention of air-containing spaces (e.g., bowel, pneumothorax) and should be avoided. Only low concentrations of inhalational agent should be used initially; midazolam should be given to minimize the risk of awareness.

2. Control ventilation to produce a near-normal PaCO₂. If the hypovolemia is still uncorrected, avoid positive end-expiratory pressure and adjust the inspiratory-expiratory ratio to give a low mean intrathoracic pressure. Use a passive or heated humidifier.
3. Monitor ventilation, heart rate and rhythm, temperature, BP, CVP, SaO₂, PetCO₂, and urine output. A processed EEG monitor may provide useful trend information in older children.
4. Insert an arterial cannula for direct BP monitoring and for repeated blood sampling for serial determination of the acid-base status, blood gases, Hct, and coagulation studies.
5. Insert a double-lumen CVP line. This provides for monitoring and drug infusions.
6. Maintain body temperature with a forced air warming mattress.
7. Be prepared to treat to possible sudden hypotension as the abdomen is opened in the child who has massive intra-abdominal bleeding.

Postoperative

1. In the absence of chest injury significant impairment of pulmonary function or need for damage control abdominal packing, remove the tracheal tube after antagonizing the muscle relaxant and with the child awake and responding and in a lateral position.
2. In the child with thoracic injuries or impaired pulmonary function, who remains unconscious—or whose condition is otherwise labile—continue ventilatory support and reevaluate later in the intensive care unit.
3. Plan postoperative pain relief.

Special Considerations

1. Overt or suspected hepatic injury: Drugs that are metabolized by the liver may have increased elimination half-lives. Cautious use of repeated doses of drugs that depend on the liver for metabolism (barbiturates, opioids) is required to prevent overdose. Use drugs that depend primarily on elimination via the lungs or kidneys (e.g., isoflurane, desflurane, *cis*-atracurium, remifentanil).
2. Overt or suspected renal injury is usually treated conservatively unless the child is hemodynamically unstable. Trauma patients with acute renal failure as a complication of prolonged hypovolemia and hypotension require special attention: Maintain an adequate mean arterial pressure, administer mannitol (0.25 gm/kg), and avoid drugs that are excreted predominantly via the kidneys (e.g., pancuronium).

3. Ruptured diaphragm: Rupture of the diaphragm is more common in children than adults and may be overlooked because respiratory distress is usually not severe. This condition requires thoracoabdominal repair:

- (a) Insert a gastric tube to decompress the upper gastrointestinal tract.
- (b) N₂O is contraindicated as in most cases of chest injury.

4. Injury to chest wall and lungs:

- (a) In young children, the ribs are relatively soft and less likely to fracture; however, the injury may separate the costochondral junctions, and this, in association with fractures of posterior ribs, may lead to "flail chest." If this results in hypoventilation, intubate the trachea and control the ventilation without delay.
- (b) Trauma to the chest wall is usually accompanied by contusion of the underlying lung, even when there are no rib fractures. This results in shunting of blood through damaged lung tissue and the need for O₂ therapy and possibly for positive pressure ventilation.
- (c) Pneumothorax, hemothorax, or both may be present. If these are suspected, insert a chest drain with a suitable valve or underwater seal before the child is anesthetized. These injuries should be suspected in the child with grunting respirations and may occur without rib fracture.

5. Overt or suspected tracheal or bronchial injury:

- (a) If there is evidence to suggest such injury, or if subcutaneous emphysema of the face, neck, or chest is present, bronchoscopy is required to define the extent of damage.
- (b) Induce anesthesia with sevoflurane in O₂ or IV propofol (very smoothly and deeply, avoiding coughing and straining). Maintain spontaneous ventilation and avoid positive pressure, which may increase any air leak. Avoid N₂O. In the case of a penetrating wound, cover the wound with a sterile dressing.
- (c) Give lidocaine (1.5 mg/kg IV), wait for 3 min, and then perform laryngoscopy and spray the larynx with lidocaine. The bronchoscope may then be inserted.
- (d) During bronchoscopy, give sevoflurane in 100% O₂ via the bronchoscope; alternately use a propofol infusion or a combination of the two techniques.
- (e) If thoracotomy is required and damage is limited to one bronchus, intubate the uninjured main bronchus using a fiberoptic scope to guide correct placement (see Chap. 4) and use 100% oxygen.
- (f) If a tracheal injury is present as a temporizing measure, pass the tracheal tube past the site of injury, almost to the carina. Check bilateral ventilation and allow spontaneous respirations. In some cases a tracheostomy may be required.

6. Injury to the heart and pericardium is rare in children but may occur in association with severe thoracic trauma. Cardiac contusion may result in changes in ventricular function that are detectable by echocardiography or scanning techniques; the ECG may show nonspecific ST changes but most often is unchanged. The clinical importance of such changes is not yet fully understood.
7. Injuries to the great vessels are less common than in adults, owing to the greater elasticity and mobility of the mediastinal structures; however, a widened mediastinum indicates the need for immediate exploration.
 - (a) If cardiac tamponade develops secondary to hemopericardium, induction of anesthesia may be very hazardous because the fixed low cardiac output cannot compensate for any drug-induced alterations in systemic vascular resistance.
 - (b) In hypotensive patients with hemopericardium, the surgeon should drain the pericardium (under local analgesia) before induction of general anesthesia. If tamponade is less severe, induction with ketamine may be possible. (**N.B.** Until the pericardium is open, keep the patient sitting or head-up and maintain spontaneous ventilation to augment venous return.)

Acute Burns and Scalds

Children are often the victims of burns and scalds. Extensive burns have widespread systemic effects: massive fluid shifts occur, plasma protein is lost, and all the major organ systems are affected. The physiologic response to acute burns may be divided into early (days 1–3) and late (after day 3) (Table 17.4):

1. Direct injury to the upper airway and lungs may cause obstruction secondary to edema, bronchospasm, or sloughed tissue early after a burn (see further). Later, pulmonary function may be affected by infection (pneumonia), acute respiratory distress syndrome (ARDS), or pulmonary vascular changes.
2. Hypovolemia and circulating myocardial depressant factor may lead to early cardiac dysfunction. After day 3, a hypermetabolic state occurs with a marked increase in cardiac output (two to three times baseline) to maintain oxygen delivery; this will persist until the child is fully grafted (see Table 17.4).
3. Circulating myoglobin and hemoglobin can directly damage the kidneys. Hypovolemia may cause acute tubular damage especially if compounded by hypoxia. After day 3, the marked increase in cardiac output increases GFR, which in turn dramatically reduces the half-life of many drugs excreted by the kidneys. In addition, hypertension occurs in approximately 50% of children with extensive burn injury. This is mediated by increased renin, endogenous catecholamines, atrial natriuretic factor, and aldosterone production. Some children require β blocker or calcium channel blockers to control the hypertension.

Table 17.4 Systemic effects of burn injury

System	Early	Late
Cardiovascular	↓ CO due to decreased circulating blood volume, myocardial depressant factor	↑ CO due to sepsis ↑ CO 2–3 times > baseline for months (hypermetabolism) Hypertension
Pulmonary	Upper airway obstruction due to edema Lower airway obstruction due to edema, bronchospasm, particulate matter ↓ FRC ↓ Pulmonary compliance ↓ Chest wall compliance	Bronchopneumonia Tracheal stenosis ↓ Chest wall compliance
Renal	↓ GFR (a) Secondary to ↓ circulating blood volume (b) Myoglobinuria (c) Hemoglobinuria Tubular dysfunction	↑ GFR 2° to ↑ CO Tubular dysfunction
Hepatic	↓ Function 2° to ↓ circulating blood volume, hypoxia, hepatotoxins	Hepatitis ↑ Function due to hypermetabolism, enzyme induction, ↑ CO ↓ Function due to sepsis, drug interactions
Hematopoietic	↓ Platelets ↑ Fibrin split products, consumptive coagulopathy, anemia	↑ Platelets ↑ Clotting factors
Neurologic	Encephalopathy Seizures ↑ ICP	Encephalopathy Seizures ICU psychosis
Skin	↑ Heat, fluid, electrolyte loss	Contractures, scar formation, difficult intubation
Metabolic	↓ Ionized calcium	↑ Oxygen consumption ↑ Carbon dioxide production & gluconeogenesis ↓ Ionized calcium
Pharmacokinetics	Altered volume of distribution Altered protein binding Altered pharmacokinetics Altered pharmacodynamics	Tolerance to opioids, sedatives Enzyme induction, altered receptors Drug interaction

Early refers to the first 24–72 h after injury, *Late* refers to days to weeks after injury; *CO* cardiac output, *FRC* functional residual capacity, *GFR* glomerular filtration rate, *ICP* intracranial pressure, *ICU* intensive care unit (With permission. Table 34-2 in Burn Injuries. Shank ES, Coté CJ, Martyn JA. In: A Practice of Anesthesia for Infants and Children. Coté CJ, Lerman J, Anderson BJ [eds], Philadelphia, 5th ed., 2013, Elsevier)

4. The liver may be damaged as a result of hypotension, hypoxemia, inhaled toxins, or sepsis. After day 3, marked increases in hepatic blood flow will increase excretion of medications metabolized by the liver.
5. Anemia of burns (decreased erythropoietin production), thrombocytopenia, and a consumptive coagulopathy may occur. Later, a notable thrombocytosis and increased fibrinogen may occur; beware of a sudden decrease in platelet count as it often heralds developing sepsis.
6. Gastric distention, intestinal ileus, and bleeding secondary to stress ulcers may develop; appropriate antacid therapy is indicated.

The anesthesiologist will be involved in the early treatment, consisting of:

1. Airway management and fluid resuscitation
2. Fasciotomy (electrical burns) and/or escharotomy (flame or scald burns) to improve adequacy of chest wall excursion or blood flow to extremities and digits
3. Debridement and grafting

Special Anesthesia Problems

1. Airway and pulmonary involvement may lead to airway obstruction, severe intrapulmonary shunting, and respiratory failure; edema of the tissues surrounding the upper airway may occur very rapidly and make intubation extremely difficult (intubate the trachea as soon as possible). Airway burns must be suspected in all children involved in enclosed-space fires (house/automobile), where inhaled temperatures may reach 1000 °F. Carbonaceous material in the mouth and nose or burned facial hair and/or eyebrows is a common warning sign. These children may have macroglossia, macrouvula, epiglottitis, and glottic edema (croup) simultaneously.

Early tracheal intubation with a cuffed tube is advised; any delay in intubation may render the airway impossible to secure due to swelling. Thermal injury is limited to the upper airway, but inhalation of products of plastic combustion, such as NO₂ or SO₂, is carried far down into the tracheobronchial tree and combines with water to form sulfuric and nitric acid and cause a distal airway burn. Loss of ciliated epithelium and other respiratory protective mechanisms predisposes to early infection and airway obstruction. Bronchospasm is common. Acid aldehydes may cause pulmonary edema and inhaled cyanide may poison the cytochrome oxidase system. CO poisoning is the most common cause of impaired oxygen delivery after burns and necessitates therapy with 100% oxygen to reduce the elimination half-life of CO (from 4 h to 30 min).

2. Maintenance of fluid balance and renal function: several formulas may be used to calculate fluid regimens for the burned patient; these are based on the burn area (excluding erythema). The Parkland formula prescribes 4 mL/kg

of lactated Ringer's solution for each 1% of burn area. One-half of this amount is given in the first 8 h and the other half over the next 16 h. Note that this formula will underestimate fluid requirements in children who weigh less than 10 kg who should receive their normal maintenance requirements as well. It should be further noted that the "rule of 9s" does not apply to toddlers and that burns of the head may account for 20% of the body surface area (it is easy to underestimate the severity of a burn). Many different resuscitation fluid regimens have been used and all are effective; see the reference (Fabia & Groner) for more discussion of this topic. In practice, fluid therapy should be adjusted as dictated by the child's clinical and biochemical status; urine output is the primary metric for hydration.

3. Acute gastric dilation: This commonly occurs and adds to the danger of regurgitation. The stomach should be decompressed, and special care should be taken during induction and emergence from anesthesia. For subsequent surgery, jejunal feeding tubes should not be removed.
4. Management of body temperature: Loss of normal skin severely impairs the patient's ability to conserve heat. Warm the operating room to maximal values (32 °C); do not unwrap all of the dressings at one time.
5. In children with extensive burns:
 - (a) Monitoring may be difficult.
 - (b) Sites for intravenous infusions may be limited; subclavian and femoral lines are common.
6. Circumferential thoracic burns: These may lead to severely compromised ventilation requiring multiple escharotomies to restore chest wall compliance.
7. Danger of infection: Extreme care must be taken to observe strict asepsis with invasive procedures.
8. Electrical burns: electrical current travels along neurovascular bundles and such an injury can produce a compartment syndrome. This type of injury will require immediate operation to perform multiple fasciotomies to preserve extremity function. Look for evidence of both entrance and exit wounds and assume everything in between may have been damaged.

Anesthesia Management

Preoperative

1. If the fire occurred in a closed space or involved burning hydrocarbons, suspect respiratory tract burns:
 - (a) Look for burning around the face (e.g., singed eyebrows).
 - (b) Assess the airway very carefully. (Children with airway burns may have considerable swelling of the pharyngeal and laryngeal tissues making intubation difficult.)

- (c) In the child with airway burns, early tracheal intubation with a cuffed tube should be performed before massive upper airway edema forms. This tube should be meticulously secured with cloth tracheostomy tape as adhesive tape cannot be used on burned tissue. Mark the appropriate distance of insertion with indelible ink so that misplacement does not occur with later dressing changes. Accidental extubation must be prevented as repeat intubation may be impossible.
 - (d) If the airway is already intubated, assure that the tracheal tube is properly placed and secured.
2. Check the adequacy of fluid and blood replacement:
- (a) The Hct should be maintained at 25–30%; a low serum albumin concentration is usually corrected to greater than 2 g/dL. Ensure adequate blood products are in the OR.
 - (b) The bladder should be catheterized, and urine output should be at least 1 mL/kg/h.
 - (c) FFP or platelets may be required to correct documented deficiency resulting in coagulopathy.

Beware of the use of glucose-containing solutions for fluid resuscitation. Hyperosmolar, hyperglycemic non-ketotic coma may occur in association with burns. If the child is receiving intravenous alimentation, then reduce the rate to two-thirds during anesthesia to avoid hyperglycemia.

- 3. Ensure that the OR temperature is at least >90 °F or 32 °C and that heating blankets and lamps are ready for use. Humidify anesthetic gases. Warm blood and intravenous fluids.
- 4. Check whether the child has been given opioids. Analgesics and hypnotics should be given to prevent pain during transportation to the OR.

Perioperative

General endotracheal anesthesia may be required, but remember:

1. In acute burns, use conservative dosing of propofol or thiopental if the child's volume status is uncertain; ketamine may be useful in such circumstances. Generally the child is induced in their hospital bed to prevent pain upon movement to the OR table.
2. Succinylcholine is contraindicated (after the first 24 h) because it may cause cardiac arrest secondary to massive potassium release from muscle.
3. Resistance to non-depolarizing muscle relaxants is increased in proportion to the burn area. Muscle relaxants must be titrated to achieve their desired effects. Monitor the neuromuscular blockade. Antagonism of very large doses of relaxants is not a problem.

4. Potent inhalational agents may cause hypotension in those with extensive burns in the acute phase, but may be useful in controlling blood pressure during the hypertensive phase. Generally a high-dose opioid technique with background inhalation agent is most useful. Alternately, TIVA with propofol infusion and an opioid or ketamine greatly simplify the anesthetic management of burned or scalded children.

For burn dressing changes:

- (a) Give atropine (0.02 mg/kg) and midazolam (50–100 mcg/kg) IV followed by ketamine (1–2 mg/kg) IV and supplemented doses as needed.
- (b) Alternatively, if IV access is not available, induce anesthesia with ketamine (4–6 mg/kg) IM combined with atropine (0.02 mg/kg) and midazolam (50–100 mcg/kg) IM. After an IV is placed, further incremental doses of ketamine (1–2 mg/kg) may be titrated IV.
- (c) A combination of propofol (1 mg/kg) and ketamine (1–2 mg/kg) IV, followed by a propofol infusion is another alternative.

For all children:

1. Carefully monitor the following:
 - (a) Cardiac rate via an esophageal stethoscope and the ECG plus pulse oximeter. The oximeter probe may be placed on the tongue or at the corner of the mouth if no other site is available.
 - (b) BP—place a sterile BP cuff at any available site; for severe burns, insert an arterial line.
 - (c) Temperature—esophageal and/or rectal.
 - (d) Blood loss—it is often difficult to estimate loss. Replacement must then be dictated by cardiovascular parameters and urine output. Blood loss is far greater with tangential excision of eschar until bleeding is observed from viable tissue, compared with full thickness eschar excision. Blood loss can be greatly reduced if the surgeon administers subcutaneous saline with dilute epinephrine (1:2,000,000 = 0.5 µg/mL) into both the donor and excised areas. Beware of fluid overload when large quantities of tumescent solution are injected; this must be considered in overall fluid management as late pulmonary edema may occur as this fluid is absorbed.
 - (e) CVP—consider utility versus danger of introducing infection. With large burns, a CVP may be the only venous access; beware that the small lumen produces very significant resistance to blood flow and may be inadequate if rapid blood loss occurs. (Avoid rapidly transfusing cold blood through a central line.) Short-term cannulation of a femoral vessel with a single lumen catheter may be indicated just for the procedure and may be removed afterward.

2. Replace blood losses carefully; large volumes may be required. Anticipate coagulation problems with massive transfusions.
3. Beware of hypocalcemia. Chronic low levels of ionized calcium have been described in burn patients. Plasma and platelet suspensions contain more citrate per unit volume than whole blood; give calcium chloride (5–10 mg/kg) or calcium gluconate (15–30 mg/kg) if unexpected hypotension occurs. If transfusion exceeds 1 mL/kg/min, anticipate hypocalcemia and administer calcium during these rapid infusions.

Postoperative

1. Order maintenance fluids:
 - (a) Clear fluids to maintain urine output at more than 1 mL/kg/h.
 - (b) Blood to maintain Hct at 25–30%.
 - (c) Albumen may be indicated to maintain total serum protein at greater than 3 g/dL.
 - (d) Electrolyte supplements as indicated by serial determinations.
2. Order analgesics by continuous infusion as required. Burn patients require constant nursing supervision so that a generous dosage of analgesics can be safely administered. Long-term infusions of ketamine and/or dexmedetomidine have been found to be very effective in reducing total opioid requirements in children with very extensive burns. Tolerance to opioids and sedatives rapidly develops. It is common for children with very extensive burn injury to require up to 1–3 mg/kg/h of morphine and 1–3 mg/kg/h of midazolam to maintain adequate analgesia, with additional bolus doses intraoperatively as needed.
3. Observe for evolving respiratory insufficiency. (This may occur during the first 24 h, even if the chest appears clear initially.) Rhonchi and a decreasing level of arterial oxygenation are the usual first signs of trouble. Inhaled β_2 -adrenergic agents may be useful as may nebulized acetyl cysteine and heparin. In severe airway burns, beware of the possibility of sudden airway obstruction because of sloughed mucosa. Bronchoscopy may be required.
4. Proton pump inhibitors and antacids should be administered to protect against stress ulcer. Burn patients may require larger than usual doses to reduce acid secretion.
5. Children who have been intubated for a period in the intensive care unit are at increased risk for development of postextubation stridor, especially if there was no detectable leak around the tube before extubation. Such children require careful monitoring for 24–48 h after extubation; treatment with racemic epinephrine, helium, and O₂ or even reintubation may be required.
6. Long-term administration of opioids and benzodiazepines may be associated with later withdrawal symptoms. Transition to oral methadone and gradual withdrawal of benzodiazepines reduce this risk.

Gunshot Wounds

Children frequently become victims of gunshot wounds. The free access to weapons in the USA and guns in the home contribute to this toll.

Anesthesiologists should be particularly aware of the tissue damage caused by modern high-velocity bullets. Tissue over an extensive area surrounding the path of the bullet is damaged or destroyed by the energy of the projectile. This is of particular importance when such wounds occur in the upper chest or neck region with the damage at the exit wound being far greater than the entrance wound. Tissue swelling can be expected to spread to involve a wide surrounding area, possibly jeopardizing the airway. The airway should be secured as early as possible, before distortion of the anatomy progresses further. If the child is relatively stable, ensure that adequate blood products are in the room and checked; otherwise, O-negative blood is necessary. Adequate venous access must be established in the upper extremities or central venous access before incision as tamponade of the intra-abdominal or intrathoracic wound will be relieved upon surgical incision and rapid hypotension may ensue. Be prepared for rapid transfusion and the need for vasopressors.

MINOR TRAUMA

Children with minor trauma must be provided with anesthesia that is safe, pleasant, and suitable for a young patient who may be ready to go home in an hour or so. Usually there is no extreme urgency in these cases, permitting a considered approach to the selection of both the anesthetic and the optimal time for surgery. However, fractures with vascular compression may require immediate intervention.

Keep in mind gastric emptying may be substantially delayed after even minor injury, resulting in a full stomach. The best guide to the status of the gastric contents after an injury is the time interval between last food and the injury; the greater the interval, the more likely the stomach has emptied. Once an injury occurs, pain and opioids may cause gastric paresis. Some children fasted more than 8 h after an injury still have a large volume of gastric contents. A safe period between oral intake, injury, and induction of anesthesia cannot be predicted. Hence, even if one can wait for a full normal fasting period, it is still advisable to use a regional technique or to induce general anesthesia using a rapid-sequence induction (with cricoid pressure) followed by extubating the trachea awake. The use of a laryngeal mask airway in children with minor injuries has been complicated by aspiration.

Although metoclopramide may help to speed gastric emptying, neither regional analgesia nor general anesthesia should be administered without fasting unless surgery is needed urgently. The need to convert an unsatisfactory regional anesthesia to general anesthesia arises more frequently in children than in adults.

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Anesthesia Outside the Operating Room

REMOTE ANESTHESIA

There is now a great demand for pediatric anesthesiologists to travel outside the operating room to provide general anesthesia or monitored sedation for a variety of medical investigations or procedures in infants and children of all ages. The concept that treatment in a children's hospital should be a pain- and stress-free experience is now well-accepted, and this has placed additional responsibilities on the anesthesia service. Anesthesia and deep sedation administered in remote locations may be associated with an increased incidence of serious complications; great caution and close attention to effective monitoring is advised for every case.

Equipment for the Remote Location

Each area should be fully equipped for the anesthesia care of the child and for any resuscitation that might be required. Each area should be provided with:

1. Primary and backup oxygen supply (checked O₂ tank), and means to provide intermittent positive-pressure ventilation (IPPV).
2. Facilities for gas scavenging if any inhalation anesthetics are used.
3. Functioning suction apparatus.
4. Good lighting.
5. Electrical outlets (operating room standards).
6. Means of immediate communication to the operating room personnel.
7. Equipment to transport the child while anesthetized.
8. Facilities and staff for the preparation and recovery of children according to published guidelines.¹

¹ Guidelines for monitoring and management of pediatric patients before, during, and after sedation for diagnostic and therapeutic procedures: update 2016. Coté CJ, Wilson S, American Academy of Pediatrics, American Academy of Pediatric Dentistry. Pediatrics 2016;138(1);e20161212.

9. Fully equipped, provisioned and regularly checked resuscitation cart and defibrillator (immediately available).

In addition, all drugs and equipment to manage the child during the procedure must be provided:

1. Monitoring for pulse oximetry, electrocardiography (ECG), PetCO₂, blood pressure, and body temperature.
2. Airway supplies; age and size appropriate facemasks, laryngoscope blades, tracheal tubes, oropharyngeal airways, laryngeal mask airways, suction catheters, and breathing circuits.
3. Appropriate regularly serviced anesthesia equipment, infusion pumps, etc.
4. All necessary drugs for anesthesia and sedation, and, syringes, a variety of IV catheters, intravenous fluid, and fluid administration sets.
5. Emergency resuscitation drugs.
6. Where indicated: equipment to maintain body temperature (e.g., forced air warmer for cardiac catheterization laboratory).

ADMINISTRATIVE PROCEDURE FOR REMOTE ANESTHESIA

For children anesthetized in remote locations, pre-procedure evaluation should be performed as for OR Patients. Children who require remote anesthesia may be urgent or semi-urgent cases; this should not exclude a full evaluation.

1. A full preanesthesia evaluation should be performed and reviewed just before sedation/anesthesia.
2. Informed consent for anesthesia should be obtained or confirmed to be on file (Note that a serial consent may be used for children undergoing serial procedures such as radiation treatments).
3. A fully detailed anesthesia record should be completed.
4. The child should recover in an appropriately staffed and equipped facility for children or be safely transported to the regular pediatric postanesthesia care unit (PACU).
5. A recovery record should be completed.
6. The child should be evaluated by the anesthesiologist when he/she has recovered and signed out of the unit (where appropriate this might be delegated using standard written criteria).

General Principles

1. The technique chosen should result in minimal (if any) post-anesthesia sequelae.
 - (a) Use short-acting drugs that will not delay recovery and are not associated with postoperative nausea and vomiting. (Therefore, when possible, avoid opioids, barbiturates, and ketamine). Propofol is ideal.
 - (b) Most children can be managed with an oxygen mask or nasal prongs, and a support to extend the neck. Occasionally, an oral or nasopharyngeal airway may be required. The laryngeal mask airway (LMA) may be a useful alternative. Finally, if all of the above maneuvers still result in an obstructed airway, tracheal intubation may be required. In general, tracheal intubation is avoided in remote locations to minimize post-extubation airway problems and to abbreviate the recovery time.
 - (c) When repeated anesthetics will be needed, a chronic intravenous line should be maintained: either a central line or a “Hep-Locked” peripheral line.
 - (d) Monitor every child as you would in the operating room.

DIAGNOSTIC AND THERAPEUTIC MEDICAL PROCEDURES

Computed Tomography (CT)

1. CT scans require absolute immobility of the child throughout. However, modern CT scan times are exceedingly brief, most scans are complete (even when contrast is required) within 10 min. Hence, many children can be managed without anesthesia and, for the others, the anesthetic plan must be tailored accordingly. Small infants can be bundled and restrained during the procedure without sedation. Older co-operative children may not require any form of sedation or anesthetic as they can be distracted and entertained for the short time necessary to complete a scan. It is sometimes necessary to sedate infants and children less than 3 years, and cognitively challenged children. Very rarely, a general anesthetic is required; usually for a child with significant comorbid disease or injury.
2. Intravenous sedation alone is suitable for many children:
 - (a) A propofol infusion is preferred.
 - (b) Intravenous pentobarbital is an alternative; an initial dose of 3 mg/kg of pentobarbital may be given. After 3 min, further doses of 1 mg/kg may be titrated up to a maximum of 7 mg/kg. (This is a regimen that may be used by specially trained nurses under supervision of the radiologist).

- (c) The child should be monitored with standard ASA monitors. All equipment required to establish an airway and ventilate the lungs should be immediately available.
- 3. If general anesthesia is required, use only plastic materials in the breathing circuit; metal components distort the image. Be aware that the metal spring in some LMA cuffs and tracheotomy tubes may cause an artifact, so the spring needs to be taped away from the field.
- 4. Contrast media may be injected intravenously to enhance the images obtained; very rarely, reactions may occur (see later discussion); be aware to limit the dose in children with renal dysfunction.

Magnetic Resonance Imaging

Magnetic resonance imaging (MRI) scans are commonly used to secure accurate anatomic diagnoses or valuable information on the physiologic changes associated with disease. The MRI is increasingly used to define vascular anatomy (Magnetic resonance angiography (MRA)), especially in children with congenital heart disease, and this requires special considerations (see later discussion). The basic component of the MRI system is a powerful superconducting magnet into which the child must be placed. This presents clinically significant problems:

- 1. The space is very confined which limits access to the child should an emergency occur; this limited space may frighten children with claustrophobia. Additionally, high and sudden noises (approximately 95 dB) and vibrations scare the child. Because of these considerations, deep sedation or general anesthesia is usually required for infants and children having MRI scans. Noise cancelling MRI-safe headphones² may help older children.
- 2. Ferrous metal objects are attracted to the magnet and become dangerous, life-threatening projectiles. Therefore, all equipment including oxygen tanks, IV poles, laryngoscope blades and handles, etc., which are taken into the unit must be MRI-compatible (non-ferromagnetic). Any “doubtful” items must be tested with a magnet before being brought into the scanner room.
- 3. It is essential that the child and accompanying parent are screened for implanted devices that might incorporate ferromagnetic material. These could be dangerously attracted to the magnet, their function could be affected by the magnetic field (e.g., cardiac pacemaker, transthoracic or transvenous pacing wires, vagal nerve stimulator), or they could seriously damage adjacent tissues (e.g., implanted prostheses, wires or vascular clips, cochlear implants, and metal foreign bodies). Cell phones, watches, credit

² www.optoacoustics.com

cards, parking receipts, some pens, etc. may be damaged or attracted to the magnet and must not enter the scanning room. Some children will have programmable VP shunts or vagal nerve stimulators (usually shut off prior to scan) that must be adjusted before or reprogrammed after the MRI. Any implanted device (e.g., spine fusion devices) must be checked for MRI compatibility and some devices may be compatible with 1.5 T units but incompatible with 3.0 T units. This situation may require a call to the manufacturer.

N.B. For children with non-removable ferromagnetic implanted material, an MRI is contraindicated.

4. The magnetic field may cause burns by inducing currents and heat in wire leads (ECG or pulse oximeter cables) or temperature probes. Prevent direct contact between the cables and the child's skin, avoid loops in the cables, and ensure that the cables are directed out of the center of the bore (not the sides) to minimize current induction. MRI-compatible ECG and pulse oximetry equipment must be used to reduce the risk of burns. Adolescents with tattoos may develop a burning sensation at the tattoo site if the dye contains ferromagnetic metals.
5. The magnetic field may affect the performance of anesthesia delivery systems; syringe pumps may be unreliable, and vaporizers may become inaccurate (bimetallic strip is non-ferromagnetic). Infusion pumps using extra long infusion lines threaded through a conduit in the wall of the scanner may be placed outside of the scanner room to avoid interference from the magnet. Note that the long tubing may have significant resistance that could affect drug delivery by some infusion pumps; check proper function beforehand.
6. MRI scanning is very motion-sensitive; if the child moves at any stage, a whole scan sequence may need to be repeated; breathing artifacts are common but generally tolerated by the radiologist.
7. The child may have a major life-threatening illness with the need for vasopressor support or invasive monitoring. This necessitates placing all pumps outside of the scanner and using long infusion lines which may interrupt the infusions and lead to cardiovascular instability. Arterial line transducers may also need to have extended lines placed to keep the transducer out of the bore of the scanner.
8. The child may have a difficult airway. In such a case, tracheal intubation should be performed outside of the scanner room so as to allow use of special airway adjuncts such as the GlideScope or a fiberoptic bronchoscope or alternately securing the airway in the operating room and transporting the child to the MRI unit anesthetized, on a properly equipped gurney.
9. Resuscitation equipment *cannot* be brought into the MRI scan room should it be required. If resuscitation is required, the child must be rapidly moved from the MRI scan room to an adjacent room in which full resuscitation equipment is available.

10. Higher power magnets (3 T) may reveal that equipment previously found to be safe and reliable in the environment of a 1.5 T unit may be unsafe and/or unreliable in a 3 T unit. The shielding installed and the distance a piece of equipment is sited from the magnet are also vital safety concerns. Even some MRI-compatible anesthesia machines will trigger an alarm should the machine be placed too close to the magnet. The anesthesiologist should be very familiar with the safety concerns of the local unit.
11. Gadopentetate dimeglumine (gadolinium) is commonly used as a contrast agent to enhance blood vessels in the MRI image. This may cause minor side effects (dizziness, nausea), but may also very rarely cause anaphylaxis. If administered to children with renal impairment, it is poorly cleared and may result in very severe complications; nephrogenic systemic fibrosis (NSF) with multiorgan involvement. Hence, the drug is contraindicated in children >2 years with renal impairment ($\text{GFR} < 30 \text{ mL/min}/1.73 \text{ m}^2$). (N.B. Radiology departments now have guidelines for gadolinium dose based on weight, creatinine, and BUN values, and these charts should be consulted). Also caution with use in those with sickle cell anemia.

Monitoring During MRI Scans

Monitoring during the scan is essential; a plastic precordial or esophageal stethoscope (without a thermistor probe) may be used, and MRI-compatible equipment is essential:

1. MRI-compatible pulse oximetry must be used continuously; assure that there are no loops in the cable leads to the child—otherwise currents may be induced and the child may be burned.
2. Continuous expired carbon dioxide monitoring is essential.
3. Blood pressure must be measured with most NIBP machines fitted with nylon connections.
4. The ECG may be monitored using non-ferromagnetic fasteners, cables, and electrodes. The electrocardiogram waveform may be distorted in the MRI environment; all other monitored parameters are less affected. The heart rate is most reliably monitored from the pulse oximeter.
5. Temperature may be monitored using probes that have radiofrequency filters or non-ferromagnetic skin temperature strips. Insulating the infant from the cool ambient magnet temperature is important; however, children usually increase their body temperature slightly during long MRI scans (likely due to absorption of RF energy).
6. MRI-compatible monitoring units are available with a remote slave monitor outside the MRI unit allowing the anesthesiologist to remain outside of the unit, but also able to view the patient via a window. Each multi-channel monitoring unit must be tested for compatibility with the local MRI magnet.

Management for Magnetic Resonance Imaging

Satisfactory immobilization of the child during the scanning process may be achieved by intravenous deep sedation/anesthesia or general anesthesia (with an LMA or tracheal tube).

Intravenous Deep Sedation/Anesthesia

An intravenous propofol infusion is the most reliable and efficient method, although some providers use combinations of dexmedetomidine, midazolam, and propofol.

1. The child should fast as for general anesthesia.
2. Monitors are placed and sedation is initiated with the child in the supine position on the MRI table beginning with a bolus dose of propofol 2–3 mg/kg followed by an infusion at an initial rate of 250 µg/kg/min. If IV access is unavailable, the child may undergo an inhalation induction before establishing IV access and then continue with a total intravenous anesthesia (TIVA) or a propofol infusion with nitrous oxide.
3. If the child moves, an additional bolus of propofol is given. Additionally, the infusion rate of propofol may be increased. Cognitively challenged children and younger infants require up to 400 µg/kg/min in the early period of anesthesia to achieve motionless conditions.
4. After 15–20 min, the infusion rate may be reduced to 200 µg/kg/min, and after a further 15–20 min (in older children), the rate may be further reduced to 150 µg/kg/min.
5. A shoulder roll is usually required to extend the neck to maintain a patent airway because the head holder slightly flexes the neck on the MRI table. Oxygen is delivered by facemask, nasal prongs, or an LMA during the scan. PetCO₂ must be monitored to assess adequacy of ventilation. If an LMA is not used, expired gases can be aspirated via an IV catheter that is inserted through the holes in the facemask with the tip positioned near the nares or mouth. Alternately, expired gases can be aspirated from nasal prongs that are designed to both administer oxygen and sample expired gases.
6. The airway is usually well-maintained during propofol sedation, however:
 - (a) If airway obstruction occurs when placed on the scan table, reposition the head and suction the airway for secretions.
 - (b) If obstruction persists, insert an oral or nasopharyngeal airway or a laryngeal mask airway.
 - (c) If obstruction persists or ventilation/oxygenation is poor, the trachea may require intubation.

7. At the end of the scan, the infusion is discontinued; recovery in an appropriately staffed and equipped pediatric recovery area is usually complete within 20–30 min.

General Anesthesia

1. General anesthesia with an LMA or tracheal intubation and controlled ventilation may be required for severely ill children, especially those with central nervous system disease, neonates and children with multiple injuries, and those who require abdominal MRI scans as these require periodic breath-holding during the scans. All anesthetics can be delivered during MRI scans; the choice should suite the child's needs. **N.B.** There is no pain and no stimulation during MRI scans, so avoid an excessive depth of anesthesia and opioids.

A useful technique for children without venous access is to induce with sevoflurane, nitrous oxide, and oxygen, establish venous access, administer 1 mg/kg of propofol to facilitate LMA placement while avoiding apnea, then reduce the inspired sevoflurane to 1.5–2.5% in 50% nitrous oxide while maintaining spontaneous respirations and 5 cm PEEP. At the end of the case, administer an additional 0.5–1 mg/kg propofol and remove the LMA; this reduces the potential for laryngospasm and for emergence delirium.

2. Children with a possible difficult airway should be managed as described above, i.e., the airway is secured outside of the scanner with all the usual difficult airway adjuncts or intubated in the operating room and transported to the scanner.

Special Considerations for Cardiac MRI Imaging

Cardiac imaging in the MRI unit successfully demonstrates the exact anatomy of congenital heart diseases and is a valuable diagnostic tool. To obtain consistent images, the scan must be successfully ECG “gated” to the cardiac cycle and obtained at a standard phase of ventilation, which can similarly be achieved by respiratory gating (“Free breathing”). Anesthesia or sedation for these children must recognize all the implications of their heart disease. Comprehensive monitoring should be provided appropriate to their disease. In many cases, tracheal intubation with paralysis and low-dose inhalation agent is required to facilitate scanning with breath holding; other patients can be managed using deep sedation with propofol. The technique chosen must balance the severity and type of cardiac anomaly and the needs of the cardiologist.

Other Radiologic Procedures

General anesthesia is often necessary for cerebral angiography in children because the procedure may be prolonged and uncomfortable and the child must remain immobile throughout.

Special Anesthesia Problems

1. Intracranial pressure (ICP) may be increased.
2. The radiologic examination may require special positioning and tilting of the child.
3. It may be difficult to maintain body temperature in infants.
4. Special attention must be paid to the total load of contrast material (mg/kg) particularly in infants with compromised renal function; low ionic contrast solutions are preferred. Reactions to contrast media may occur; though this is now quite rare with the use of nonionic agents.
5. It is desirable for children to recover rapidly so that their neurologic status can be checked.

Anesthesia Management

Preoperative

1. Assess the child's neurologic status and other underlying medical conditions carefully.
2. Check for a history of allergy, asthma, prior anesthetic experiences, or previous reactions to radiologic contrast media.
3. Avoid premedication if ICP is increased.

Perioperative

1. An intravenous induction is preferred. Give propofol or thiopental followed by a relaxant.
2. For children with increased ICP, give lidocaine 1.5 mg/kg IV and a short-acting opioid (i.e., fentanyl 2 µg/kg) to attenuate the hypertensive response to laryngoscopy and intubation.
3. Insert a tracheal tube. A nasal or armored tube should be used if the positioning or movement of the child might result in kinking of an oral tracheal tube.
4. Maintain anesthesia with sevoflurane or a propofol infusion (sevoflurane may provide more rapid awakening after a prolonged procedure).

Cerebral Arteriography

1. If a tumor or an arteriovenous malformation (AVM) is suspected, maintain anesthesia with N₂O, a low concentration of an inhalational agent, and a muscle relaxant. Ventilation should be controlled to produce a PetCO₂ of approximately 30 mmHg (confirm by sampling from the arterial catheter). This degree of hypocapnia may improve radiographic definition by constricting the normal vessels while abnormal vessels remain dilated.

N.B. If an AVM is suspected, preoperative management and induction of anesthesia should be as outlined in Chap. 8.
2. Chart the total volume of contrast medium and flush fluid carefully, especially in small infants. Beware of fluid overload, especially in small infants with AVMs.
3. A transient bradycardia with hypotension may occur at the time of injection into the carotid artery owing to baroreceptor activity; atropine prevents this response.

Postprocedure Management

1. The child must be awake or sedated but with a patent airway before transfer to PACU. After angiography, it is necessary to maintain pressure over the catheterization site to prevent hematoma formation; this is easier to achieve if the child is quiet and immobile.
2. Usually low-dose opioids combined with midazolam helps to reduce restlessness and movement that might dislodge the clot at the puncture site.
3. Check arterial puncture sites frequently; if a limb artery was used, check the circulation to that extremity.

Additional Possible Complications

In addition to the complications that may develop during administration of any anesthetic, some special problems may occur during neuroradiologic procedures.

1. Hypothermia due to difficulty in keeping warming blankets and heating lamps in place during frequent changes in the child's position and the cold temperature of the angiography suite.
2. Acute increase in ICP, leading brain stem herniation, is often associated with bradycardia. In this event:
 - (a) Hyperventilate the lungs.
 - (b) Request that a ventricular tap be performed as soon as possible.
 - (c) Give an osmotic diuretic (mannitol 0.5–1 g/kg or furosemide 0.5–1 mg/kg) and full doses of atropine to counter the bradycardia.

3. Allergic reaction to contrast media (hives, bronchospasm, hypotension). Nonionic contrast media containing iodine are most commonly used (i.e., iopamidol [Isovue]).
4. Stroke may occur if the embolizing glue or coil plugs a blood vessel for which it was not intended.

Reactions to Contrast Media

1. Although very rare in children, the anesthesiologist must be prepared for this possibility.
2. Minor allergic reactions (i.e., skin rashes) may be treated with diphenhydramine, 1 mg/kg IM or IV.
3. Major anaphylactic shock or bronchospasm (very rare) must be treated aggressively as follows:
 - (a) Ventilate with 100 % oxygen; cardiopulmonary resuscitation as required.
 - (b) Epinephrine 1–10 µg/kg IV (depending upon the severity of the reaction), followed by an epinephrine infusion 0.05–0.2 µg/kg/min.
 - (c) Hydrocortisone 10 mg/kg IV.
 - (d) Intravenous fluids as necessary to maintain the blood pressure.
 - (e) H1 blocker (diphenhydramine 1–2 mg/kg IV) and H2 blocker (ranitidine 1.5 mg/kg IV).
 - (f) Transfer to an ICU environment for further treatment and observation.
4. Contrast media may cause sickling in children with sickle cell disease.

RADIATION THERAPY

Infants and small children usually require general anesthesia or deep sedation to render them immobile for the period of radiotherapy. Treatments may have to be repeated daily or twice daily for many days. Therefore, a technique should be used that minimally interferes with the child's lifestyle and nutrition and causes the least emotional upset; intravenous sedation with propofol has been demonstrated to be very effective.

These children usually will have a central venous access device inserted to facilitate their care for the duration of their therapy. This can be used to administer the drugs, but:

1. Be meticulous with aseptic precautions to prevent catheter infection.
2. Carefully flush the catheter clear of propofol after the treatment. Residual propofol in the catheter might predispose to infection or to unexpected apnea when flushed later in PACU, on the ward, or at home.
3. Flush the system with an appropriate heparin solution so as to preserve device function and avoid clotting.

Alternatively, an intravenous catheter should be painlessly introduced (using a topical anesthetic cream) at the first treatment and maintained by the use of a splint and “Hep-lock” connection for use during subsequent sessions.

For radiotherapy to the head region, an immobilization frame that applies suction to the hard palate is commonly used. Surprisingly, this frame maintains the airway extremely well without the need for further interventions. Child-specific fiberglass masks crafted during pre-radiation therapy planning CT scan sessions that lock into position are also used for this purpose.

Expired CO₂ is monitored by inserting an IV catheter into the oxygen mask and locating the tip near the nose or mouth for optimal sampling. SaO₂ and NIBP monitoring are routine. All parameters can be monitored by closed-circuit television when the patient care team leaves the radiation suite during treatment. Recovery is very rapid after short procedures using propofol, and the child’s appetite soon returns. The child must be recovered in an appropriately staffed and equipped pediatric recovery facility.

INVASIVE MEDICAL PROCEDURES (ONCOLOGY)

Children who need lumbar puncture, bone marrow aspiration, or other painful procedures should be provided with optimal sedation and analgesia. This is particularly important for the child with a malignant disease who requires repeated sessions; an optimal management plan should be instituted at the outset of treatment. Apart from the use of well-selected sedative and analgesic drugs, there are some other important considerations:

1. The child and the parents must be well-prepared for the procedure and know exactly what to expect. Psychological preparation and behavioral training may be of value.
2. The procedure should be performed in comfortable, pleasant surroundings. The parents should be encouraged to accompany the child. The area should be provided with oxygen, suction, and emergency equipment to deal with potential complications; this equipment can be discreetly covered but should be immediately available.
3. A suitably equipped and staffed recovery area must be provided, and the parents should be allowed to remain with the child during awakening.

Suggested Routine

1. Fasting guidelines as for general anesthesia.
2. The child’s current health status should be fully reassessed and informed consent for the procedure should be obtained on file for serial treatments.

3. If the child does not have an established intravenous route (e.g., Hickman line or venous port), prepare to establish one by placing topical local anesthetic cream over a suitable vein as soon as the child arrives at the clinic. If the Hickman line is accessed be meticulous with asepsis.
4. If the child is anxious, premedicate with oral midazolam.
5. Sedation is provided with intravenous propofol, supplemented, if necessary, by small doses of fentanyl. Frequently, if local analgesics are properly administered, propofol alone is adequate. It is preferable to avoid opioids as they increase the risk of postprocedure nausea and vomiting. Small doses of ketamine (0.25–1 mg/kg) may be added to the propofol prescription to provide additional analgesia. This will not usually increase vomiting but may delay recovery slightly.
6. During the procedure, standard ASA monitoring should be utilized.
7. Age- and size-appropriate equipment to establish an airway and ventilate the lungs should be immediately available.
8. Monitor ventilation carefully; when propofol is being used, the child may be positioned in the lateral decubitus position, or even prone on bolsters; usually a clear airway is maintained without the need for tracheal intubation. Minor degrees of airway compromise usually can be corrected by repositioning the head or by insertion of an oropharyngeal airway, which is well-tolerated during deep propofol sedation/anesthesia.

Gastrointestinal Endoscopy

Infants and children may require anesthesia during diagnostic upper or lower endoscopy and for therapeutic procedures such as injection of esophageal varices and percutaneous endoscopic gastrostomy.

Gastroduodenoscopy

There are two alternative methods for upper endoscopy and therapeutic procedures; an unprotected airway or a tracheal tube. Since these procedures are almost always performed in the lateral decubitus position, which facilitates the drainage of oropharyngeal fluids from the mouth, with a gastroscope in the stomach, many provide anesthesia without instrumenting their airways. In such cases, anesthesia is induced by inhalation and an IV placed. Maintenance of anesthesia requires either intermittent propofol (1–1.5 mg/kg IV boluses) or a continuous infusion of propofol with or without remifentanil. An infusion is usually unnecessary as these procedures are very brief. Additionally, there is little need to use an opioid in non-painful procedures. Care should be taken to prevent nerve compression in the lateral decubitus position. If the decision is to use a tracheal tube, then the larynx may be sprayed with lidocaine to reduce coughing and

bucking before intubation. Maintenance may include propofol with or without remifentanil infusions, titrating the doses to the child's requirements or sevoflurane. With both approaches, the endoscopist should be reminded to empty all air from the esophagus and stomach after the procedure.

In infants <10 kg, severely obese children, or those with abnormal airway anatomy, the scope may cause upper airway obstruction. In such cases, the trachea is intubated to maintain a patent upper airway. Alternatively, after the larynx is topicalized, the endoscopist can pass the scope first, followed by an LMA, one size smaller than that estimated by weight. A third approach is to place the LMA and then have the endoscopist pass the scope after all air is removed from the cuff. After passage of the scope, the cuff is reinflated. Similarly, if injection of varices is planned in any age child, the trachea should be secured with a tracheal tube and a large-bore intravenous line should be in place. The stomach should be aspirated at the end of the procedure in all cases before the endoscope is removed, and the child should be awake before tracheal extubation. If a child is to undergo insertion of a gastrointestinal camera (PillCam), then intubation may be the safest method of airway management since this device is quite large and could accidentally be deployed obstructing the airway.

Colonoscopy

Colonoscopy may follow the EGD or be a separate procedure. Either anesthesia may be continued as per the EGD (e.g., IV propofol) or 70% N₂O administered by facemask (or LMA) with supplemental doses of propofol as needed. If it is a separate procedure, general anesthesia or deep sedation may be continued using propofol; tracheal intubation is usually unnecessary. In young children, anesthesia may be maintained using either a propofol infusion with oxygen delivered by nasal prongs or facemask or sevoflurane through an LMA or breathing tube after general anesthesia is induced. The child is positioned either in the left lateral decubitus position or supine and care must be directed at preventing nerve compression. The extent of abdominal distention caused by insufflation of air into the bowel should be monitored. The endoscopist should be encouraged to remove all air possible before terminating the procedure.

Percutaneous Endoscopic Gastrostomy (PEG)

Insertion of a PEG requires tracheal intubation with the addition that the skin area at the proposed gastrostomy site should be infiltrated with a long-acting local anesthetic.

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Anesthesia Implications of Syndromes and Unusual Disorders¹

This Appendix contains brief descriptions of common and rare syndromes; their associated anesthetic considerations for safe pediatric anesthesia practice are outlined. Many of these shared features make precise identification difficult; the reader should consider all the information provided for informing anesthetic considerations. *Whenever possible, the referenced literature should be consulted before anesthesia* is undertaken. A brief list of several excellent resources that are up-to-date follows this introduction. In some rare syndromes, the only references are in foreign language journals; we have listed these when an English abstract was appended. Very occasionally, the reference refers to the disease in adults; we still include it when no published pediatric literature was available and where we thought that the adult information provided might be useful. Space limits a complete literature review for each condition, but we have attempted to list the most important and recent publications or case reports.

There are now more than 10,000 medical syndromes recorded, so it is inevitable that this list is incomplete. Although the number of syndromes has increased and existing syndromes have become better understood in part because of genetic studies, anesthesiologists may still encounter unreported difficulties and complications. When in doubt as to the identity and implications of a particular syndrome, the anesthesiologist should make preparations that take into account all possible associated disorders.

Recurring challenges that are common in many of these syndromes influence the choice of an anesthetic technique, specifically the difficult airway and congenital heart defects. The reader is encouraged to consult the chapters that provide approaches to management of the difficult airway and specific congenital heart defects, which may be adapted to the particular syndrome presented and in accordance with the practice and experience of the anesthesiologist. Those conditions that include impaired renal function require special care when administering radiologic contrast media. Classic descriptions of the clinical

¹ Originally adapted from Jones EP, Pelton DA: Can Anaesth Soc J 23:207, 1976 and extensively augmented and revised.

problems of a syndrome may allow the reader to decide when newer drugs and techniques can be used safely in individual cases.

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Name	Description	Anesthesia implications
Achondroplasia	Most common form of dwarfism. Defective fibroblastic growth factor 3 (FGFR3) at chromosome 4. Defective bone formation with decreased rate of endochondral ossification leads to shorter tubular bones. Foramen magnum or spinal stenosis may occur. Sleep apnea may be related to brain stem compression. May need suboccipital craniectomy, laminectomy, or cerebrospinal fluid (CSF) shunts	Intubation may be difficult but usually is not. Tracheal tube size and depth of insertion are best judged by weight (not age)—most require a smaller tube than indicated by their age. Caution with neck movements—avoid excessive extension. IV access is difficult due to excess lax skin. High incidence of complications when operated on in the sitting position Krishnan BS, Eipe N, Korula G. Anaesthetic management of a patient with achondroplasia. <i>Paediatr Anaesth</i> . 2003;13(6):547–9.
Acrocephalopolysyndactyly	See Carpenter syndrome and Saethre–Chotzen syndrome	
Acrocephalosyndactyly	See Apert syndrome	
Adrenogenital syndrome	Inability to synthesize hydrocortisone; virilization of females. All need perioperative steroid supplementation, even if not salt losing	Preoperatively check electrolytes and ensure that supplementary corticosteroids are administered even if anesthesia is unaccompanied by surgery (i.e., for MRI or other investigations)
	Abel M, von Petrykowski W. Perioperative substitution therapy in congenital adrenogenital syndrome with salt loss. [German–English abstract]. <i>Anaesthetist</i> . 1984;33(8):374–6.	
Adrenoleukodystrophy	See leukodystrophy	
Aicardi syndrome	Absent corpus callosum, chorioretinopathy, and infantile spasms. Marked myotonia and drowsiness. Repeated aspiration pneumonia	Caution with muscle relaxants and residual weakness. No other special recommendations
	Mayhew J. Anesthesia in a child with Aicardi syndrome. <i>Paediatr Anaesth</i> . 2007;17(12):1223.	
	Terakawa Y, Miwa T, Mizuno Y. Anesthetic management of a child with Aicardi syndrome undergoing laparoscopic Nissen's fundoplication: A case report. <i>J Anesth</i> . 2011;25:123–6.	

Table continues on the following page.

Name	Description	Anesthesia implications
Alagille syndrome	Disorder of the bile ducts with cholestasis. May have cardiac (97%), musculoskeletal (inc. vertebral), ocular, facial, and neurologic abnormalities. Variable presentation of an autosomal dominant inherited condition. Severe cases necessitate liver transplantation	<p>Assess cardiac status (echocardiogram) preoperatively. Bilirubin, coagulation profile, and vitamin K level should be checked preoperatively.</p> <p>Hepatosplenomegaly encourages regurgitation; a rapid sequence induction may be necessary to prevent aspiration. Caution with drugs handled by the liver.</p> <p>Avoid drugs that decrease hepatic blood flow (HBF); isoflurane has least effect on HBF. Maintain intravascular volume to preserve HBF. Epidural anesthesia may be preferred over opioids, but check clotting state and vertebral anatomy (check X-rays).</p> <p>Caution with transport and positioning; osteoporosis may be present (vitamin D deficiency)</p>
Albers-Schönberg disease (marble bone disease; osteopetrosis)	Disorder of osteoclasts and bone overgrowth. Infantile malignant form presents at less than 1 year of age with failure to thrive and seizures because of hypocalcemia. Lethargy, macrocephaly, frontal bossing, and obligate mouth breathing (overgrowth nasopharyngeal bone) are common. Hydrocephalus and pneumocephalus may occur. Brittle bones, pathologic fractures. Anemia from marrow sclerosis; hepatosplenomegaly	<p>Ensure that anti-seizure medications will be given the morning of surgery. Check hemoglobin and Ca^{++} level preoperatively. Care in moving, positioning, and use of restraints. Beware difficult airway; have difficult airway cart at hand. Nasal airway unreliable; may obstruct when anesthetized; prepare airways and laryngeal mask airway (LMA). N.B. Limited mobility of joints. Avoid nitrous oxide (pneumocephalus)</p> <p>Ozer AB, Erhan OL, Demirel I, et al. Administration of general anaesthesia to a paediatric patient with osteopetrosis. BMJ Case Reports. 2005;15:256-7 doi:10.1136/bcr-2012-006901</p>
Albright-Butler syndrome	Renal tubular acidosis, hypokalemia, osteomalacia, rickets, renal calculi. Treated with alkali and K^+ supplementation	<p>Check and correct electrolytes to normal values. Renal impairment; caution with renally excreted drugs and fluid therapy</p> <p>Unwin RI, Capasso G. The renal tubular acidoses. J Royal Soc Med. 2001;94(5):221-5</p>

Abricht hereditary osteodystrophy (pseudohypoparathyroidism)	Ectopic bone formation, developmental delay. Hypocalcemia: possible ECG conduction defects, neuromuscular problems, convulsions. May present for cataract surgery	Preoperatively check ECG and electrolytes. Avoid hyperventilation and respiratory alkalosis (exacerbates hypocalcemia). Monitor ECG for an increased QT interval or conduction defects. Extreme caution with muscle relaxants, possible residual weakness
Aldrich syndrome	See Wiskott-Aldrich syndrome	
Alexander disease	See leukodystrophy	
Alpha-mannosidosis	Developmental delay, skeletal and muscle abnormalities, psychiatric symptoms, and impaired pulmonary function and infections. Short stiff unstable cervical spine	Assess airway; use in-line stabilization during laryngoscopy; prepare for difficulty—but usually routine. Antiemetic Rx recommended
Hallas P, Borgwardt LG, Roed J, et al. Anesthesia for patients with alpha-mannosidosis: a case series of 10 patients. <i>Pediatr Anesth.</i> 2011;21:1269–70		
Apert syndrome	Nephritis and nerve deafness; renal pathology variable. Renal failure in second to third decade. May present for renal transplantation	Use caution with drugs excreted by kidneys. Check ECG as AV conduction defects may occur
Ferrari E, Nascimento P Jr, Vianna PT. Complete atrioventricular block during renal transplantation in a patient with Apert's syndrome: case report. <i>Sao Paulo Med J.</i> 2001;119(5):184–6		
Alström syndrome	Obesity, blindness by 7 years, hearing loss, diabetes after puberty, hepatic dysfunction, and glomerulosclerosis. Decreased liver renal function. Dilated cardiomyopathy	Check liver function. Diabetes and obesity require special consideration. Caution with drugs excreted by kidneys. Echo cardiac assessment required. Great caution with inhalation agents; avoid tachycardia
Avazou M, Tanaka T, Sato S, et al. Hepatic dysfunction in two sibs with Alstrom syndrome: case report and review of the literature. <i>Am J Med Gen. 1997;69(1):13–6</i>		
Corbetti F, Razzolini R, Bettini V, et al. Alstrom syndrome: cardiac magnetic resonance findings. <i>Int J Cardiol.</i> 2013;167(4):1257–63		

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Name	Description	Anesthesia implications
Amaurotic familial idiocy	See gangliosidosis GM2 (Tay-Sachs disease)	Sensitive to thiopental, propofol, and respiratory depressants (due to reduced muscle mass). Avoid muscle relaxants where possible: unpredictable response to non-depolarizing relaxants
Amyotonia congenita (infantile muscular atrophy)	Degeneration of motor neurons. Progressive muscular weakness and respiratory failure. Prone to aspiration pneumonia	Check baseline ventilatory status (spirometry). Do not use succinylcholine: possible K ⁺ release and cardiac arrest. Use minimal doses of thiopental, propofol, and relaxants (cisatracurium preferred). Rocuronium and sugammadex may also be used. Monitor neuromuscular blockade. Avoid respiratory depressants. Consider regional analgesia
Amyotrophic lateral sclerosis	Degeneration of motor neurons. Progressive muscular weakness and respiratory failure. Prone to aspiration pneumonia	Check baseline ventilatory status (spirometry). Do not use succinylcholine: possible K ⁺ release and cardiac arrest. Use minimal doses of thiopental, propofol, and relaxants (cisatracurium preferred). Rocuronium and sugammadex may also be used. Monitor neuromuscular blockade. Avoid respiratory depressants. Consider regional analgesia
Analbuminemia	Extremely low level of serum albumin (4–100 mg/dL)	Very sensitive to drugs that bind to albumin (e.g., propofol, thiopental, vecuronium)
Analphalipoproteinemia	See Tangier disease	Koot BG, Houwen R, Pot DJ, et al. Congenital analbuminemia biochemical and clinical implications. A case report and literature review. Eur J Pediatr. 2004;163:664–70
Andersen disease (glycogen storage disease type IV)	Deficiency of glucosyltransferase (brancher enzyme). Early severe hepatic cirrhosis; liver failure; splenomegaly; hemorrhagic tendency	Check coagulation factors; treat excessive bleeding with fresh frozen plasma. Risk of hypoglycemia under anesthesia; measure glucose values and infuse dextrose perioperatively
Andersen syndrome	Periodic paralysis, long QT interval, dysmorphic features; severe midfacial hypoplasia → relative mandibular prognathism; abnormal structure and angle of mandible (triangular facies), kyphoscoliosis	Possible airway problems; mask ventilation and intubation may be difficult. Have difficult airway cart at hand. Assess respiratory status. Observe precautions for long Q-T syndrome. Avoid succinylcholine
Young DA. Anesthesia for the child with Andersen's syndrome. Paediatr Anaesth. 2005;15(11):1019–20		

Angelman syndrome	Developmental delay, craniofacial anomalies, drooling, ataxia, seizures, paroxysmal laughter, muscle atrophy. Genetic defect in maternal 15q chromosome in 75% of cases affecting GABA _{A_{gq}} subunit receptors may alter response to anesthetic drugs. Vagal hypertonia	Usually uncooperative. Give anti-seizure medications on the morning of surgery. Caution with IV hypnotics (GABA _{A_{gq}}) and muscle relaxants (myopathy). Propofol may be useful. Normal response to inhaled agents and opioids. Possible difficult airway. Prophylactic anticholinergic to limit vagal overactivity
Bevinetto CM, Kaye AD. Perioperative considerations in the patient with Angelman syndrome. <i>J Clin Anesth.</i> 2014;26:75-9.		
Angioedema (hereditary angioneurotic edema)	Episodic brawny edema of extremities, face, trunk, airway abdominal viscera, lasts 4 h to 1 week. Mutation on chromosome 11 responsible. Onset in childhood differentiates this from idiopathic form. Etiology: (1) deficiency of C1 esterase inhibitor, reduced to 20% of normal levels or (2) normal levels of dysfunctional type of C1 esterase inhibitor. Accumulation of vasoactive substances → increased vascular permeability → edema. Usually painless; may have prodromal focal tingling or “tightness.” Often induced by trauma or vibration. May have bouts of abdominal pain, diarrhea; hemoconcentration leading to hypotension, shock, pharyngeal edema (usually develops slowly). Most deaths from laryngeal edema; mortality rate up to 33%. Treatment with antifibrinolytic and hormonal agents	Check complement assay, Hct, fluid status, treatment history, previous drug reactions. Note voice change or dysphasia. Prophylaxis (e.g., for dental manipulation). C1 esterase inhibitor (C1INH) replacement therapy should be used. Otherwise fresh frozen plasma for 1–3 days, preoperatively. Continue EACA IV perioperatively and postoperatively. Danazol (androgen) is useful. Acute attack: epinephrine, steroids, antihistamine (in case diagnosis is a true anaphylaxis), fresh frozen plasma, or purified C1 inhibitor. If pharyngeal edema develops: tracheal intubation (leave in place for 24–72 h); if this is not possible, perform tracheotomy. Regional anesthesia when possible. Otherwise, extreme care when instrumenting airway

Wall RT, Frank M, Hahn M. A review of 25 patients with hereditary angioedema requiring surgery. *Anesthesiology.* 1989;71(2):309–11.
 Riedl M. Hereditary angioedema therapies in the United States: Movement toward an international treatment consensus. *Clin Ther.* 2012;34:623–30.
 Williams AH, Craig TJ. Perioperative management for patients with hereditary angioedema. *Allergy Rhinol. (Providence.)* 2015;6(1):50–5.

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Name	Description	Anesthesia implications
Angio-osteohypertrophy	See Klippel-Trénaunay-Weber syndrome	
Anhidrotic ectodermal dysplasia	See Christ-Siemens-Touraine syndrome	
Antley-Bixler syndrome	Recessive condition with bony and cartilaginous abnormalities: craniostenosis, midface hypoplasia, choanal atresia, and joint contractures. May have cardiac, gastrointestinal, and renal abnormalities. Respiratory obstruction may require early intervention (including tracheostomy). Need major cranial surgery in neonates to relieve craniostenosis	Potential respiratory problems and difficult intubation. Have the difficult airway cart available. Care with positioning. Extremity deformities may preclude easy vascular access
	Boswell D, Mayhew J. Anesthesia for an infant with Antley-Bixler syndrome. <i>Paediatr Anaesth</i> . 2007;17(5):97-8	
Apert syndrome (acrocephalosyndactyly)	Developmental delay. Hypoplastic maxilla and exophthalmos. Syndactyly. Craniostenosis, possibly with increased ICP; fused cervical vertebrae. Trachea may be narrow with fused rings ("bamboo trachea") CHD may be present	Mask anesthesia may be difficult; have supraglottic airway devices ready. Orotracheal intubation is almost always easy. Nasotracheal intubation may be difficult because of narrowed nasal passages. Ensure that a leak is present around the tracheal tube. ICP may be increased. High incidence of respiratory complications—caution if history of recent URI
	Barnett S, Moloney C, Bingham R. Perioperative complications in children with Apert syndrome: a review of 509 anesthetics. <i>Paediatr Anaesth</i> . 2011;21(1):72-7	
Arachnodactyly	See Marfan syndrome	
Arima syndrome	Malformation of the brain stem with congenital amaurosis and psychomotor retardation. Renal dysfunction or failure because of polycystic kidneys. May also have hepatic failure	Preoperatively check serum electrolytes if chronic renal failure present. Caution with renally excreted drugs. Hyperkalemia during surgery can produce ECG changes necessitating treatment
	Koizuka S, Nishikawa K-I, Nemoto H, et al. Intraoperative QRS-interval changes caused by hyperkalaemia in an infant with Arima syndrome. <i>Paediatr Anaesth</i> . 1998;8:425-8	

Arthrogryposis multiplex	Multiple congenital contractures, stiffness of joints; CHD in about 10% of cases. Intraoperative tachycardia, hypermetabolism, and hyperthermia may occur but without classic biochemical or genetic markers for MH	Minimal thiopental/propofol required; muscles replaced by fat. Sensitivity to non-depolarizing muscle relaxants. Difficult intubation and airway problem because of limitation of temporomandibular movement; have difficult airway cart available. Tachycardia and increase in body temperature often observed for unclear reasons (not prone to MH). Monitor body temperature and be prepared for cooling measures. Regional analgesia may be used, consider ultrasound guidance
Asplenia syndrome	Absent spleen; malposition of abdominal organs. Very complex cardiovascular anomalies (i.e., single ventricle); cyanosis and heart failure in many cases. Increased susceptibility to overwhelming infection	Preoperatively assess cardiac status (echocardiogram), SBE prophylaxis if indicated; use sterile technique, reverse isolation. Do not use cardio depressants; ketamine, midazolam, and fentanyl recommended (Uchida K, Ando T, Okuda C et al. Anesthetic management of an infant with a single ventricle (asplenia syndrome) for noncardiac surgery. [Japanese] English abstract. Masui. 1992;41(11):1793-7)
Ataxia telangiectasia	Cerebellar ataxia, skin and conjunctival telangiectasia; decreased serum IgA or IgE. Defective immunity → recurrent pulmonary and sinus infections; bronchiectasis. Severe anemia may be present. RES malignancy in about 10% of cases	Check Hb and Hct levels and pulmonary function as needed. Treat anemia. Use antibiotic prophylaxis if indicated. Use sterile technique (reverse isolation). Supplemental O ₂ may be required in post-op period
<i>Lockman JL, Iskander AJ, Bembra M, et al. Anesthetic and perioperative risk in the patient with Ataxia-Telangiectasia. Pediatr Anesth. 2012;22:256-62.</i>		<i>Table continues on the following page.</i>

Name	Description	Anesthesia implications
Bardet-Biedl syndrome	Developmental delay, pigmentary retinopathy, polydactyly, obesity, hypogenitalism, diabetes, and hypertension. (Spastic paraplegia, typical in Laurence-Moon syndrome, is absent.) May have renal abnormalities and congenital heart defects	Preoperatively assess cardiac status (echocardiogram), renal (BUN/creatinine), endocrine, and fluid status. SBE prophylaxis if indicated. Use contrast material with caution. May have bifid epiglottis
Bartter's syndrome	Bauaman ML, Hogan GR. Laurence-Moon-Biedl syndrome. Report of two unrelated children less than 3 years of age. <i>Am J Dis Child.</i> 1973;126:119-26 Chittoor dan S, Crowe S. Day care general anaesthesia for a child with Bardet-Biedl syndrome. <i>Case Reports in Medicine.</i> 2010;pii: 239239, doi: 10.1155/2010/239239	Hypokalemic, hypochloremic metabolic alklosis. Normotensive but hypovolemic. Chloride reabsorption defect with urinary potassium loss. Juxtaglomerular cell hyperplasia, hyperaldosteronism, prostaglandin overproduction and activation of the renin-angiotensin-aldosterone system
Beare-Stevenson syndrome	Kannan S, Delph Y, Moseley HS. Anaesthetic management of a child with Bartter's syndrome. <i>Can J Anaesth.</i> 1995;42:808-12	Craniosynostosis with cloverleaf skull, hydrocephalus, proptosis, choanal atresia, cleft palate, cutis gyratum, and abnormal genitalia. Associated cervical spine and foramen magnum abnormalities Airway maintenance and intubation may be very difficult; have difficult airway cart available. IV access may be complicated by skin changes. Caution with neck movement; use in-line stabilization if indicated. Protect the eyes with ointment and tape to close or use eye shields. Monitor ventilation postoperatively

Upmeyer S, Bothwell M, Tobias JD. Perioperative care of a patient with Beare-Stevenson syndrome. *Paediatr Anaesth.* 2005;15(12):1131-6

Becker syndrome	See Duchenne muscular dystrophy	Rare disease caused by genetic defect with variable inheritance patterns. Birth weight greater than 4000 g, macroglossia, and exophthalmos. Omphalocele, visceromegaly, hyperviscosity syndrome, umbilical hernias, congenital heart disease, and hypoglycemia are common (see neonatal hypoglycemia). Cleft palate may be associated, and if this is repaired, tongue reduction may be indicated to prevent severe airway obstruction	Preoperatively assess cardiac status (echocardiogram) in neonate to rule out CHD. Airway problems and difficult intubation because of large tongue; have difficult airway cart available. Trachea may be large for age—use a cuffed tracheal tube. Monitor blood glucose frequently and treat hypoglycemia by slow infusion of dextrose (bolus dose may cause rebound hypoglycemia). Nasopharyngeal airway useful for postoperative airway obstruction. May require phlebotomy to reduce high hematocrit
Beckwith-Wiedemann syndrome (Beckwith-Wiedemann syndrome, infantile gigantism)	Suan C, Ojeda R, Garcia-Perla JL, et al. Anaesthesia and the Beckwith-Wiedemann syndrome. <i>Paediatr Anaesth</i> . 1996;6:231–3 Kimura Y, Kamada Y, Kimura S, et al. Anesthetic management of two cases of Beckwith-Wiedemann syndrome. <i>J Anesth</i> . 2008;22(1):93–5 Eaton J, Atiles R, Tuchman JB. GlideScope for management of the difficult airway in a child with Beckwith-Wiedemann syndrome. <i>Paediatr Anaesth</i> . 2009;19(7):696–8 Batra M, Valecha UK. Anesthetic management of tongue reduction in a case of Beckwith-Wiedemann syndrome. <i>Anes Clin Pharm</i> . 2014;30:562–4	Gross ulceration of mouth (usually first sign; may extend to esophagus) and genital area; uveitis, iritis, conjunctivitis, skin lesions, nonerosive arthritis. May have vasculitis, myocardial, and CNS involvement; risk of sepsis at sites of skin punctures, etc.	Preoperatively assess cardiac status (echocardiogram and ECG) to rule out cardiac involvement. Use sterile technique. May have history of steroid therapy; nutritional status may be very poor. Tracheal intubation may be very difficult because of scarring in pharynx; have difficult airway cart available
Behcet syndrome	Turner ME. Anaesthetic difficulties associated with Behcet's syndrome. <i>Br J Anaesth</i> . 1972;44:100–2 Sahihoglu Z, Dikmen Y, Demirogluk S, et al. Oral aphthous ulcers after difficult intubation in a patient with Behcet's disease. <i>Anaesthesia</i> . 2002;57(6):620–1		
Binder syndrome	Maxillonasal dysplasia; if severe, may be corrected surgically	Advancement of maxilla and wiring of maxilla and mandible may cause airway problems perioperatively and postoperatively	
	Chummun S, McLean NR, Nugent M, et al. Binder syndrome. <i>J Craniofac Surg</i> . 2012;23(4):986–90		<i>Table continues on the following page.</i>

Name	Description	Anesthesia implications
Blackfan-Diamond syndrome	Congenital idiopathic RBC aplasia. Liver and spleen enlarged; hypersplenism, thrombocytopenia. Craniofacial and cardiac defects may be present. Treatment with steroids and repeated transfusions; hemochromatosis may develop. Bone marrow transplant may be successful. Increased incidence of malignancy (leukemia)	Preoperatively assess cardiac status (echocardiogram). Coagulation studies preoperatively; treat anemia and have platelets available. Give additional steroids. Considerations of hemochromatosis
Bland-White-Garland syndrome	Coronary artery malformation with left coronary arising from the pulmonary trunk. Myocardial ischemia leading to acute heart failure. Lethal if not corrected early	Preoperatively assess cardiac status (echocardiogram). Anesthesia as for coronary artery disease. Left ventricular dysfunction may require aggressive therapy to wean from CPB
Bowen syndrome	See cerebrohepatorenal syndrome	Kleinschmidt S, Grueness V, Molter G. The Bland-White-Garland syndrome: clinical picture and anaesthesiological management. <i>Pediatr Anaesth</i> . 1996;6:65-8 Minkovich LL, Brister SJ, Slinger PD. Transthoracic echocardiography in adult-type Bland-White-Garland syndrome. <i>Anesth Analg</i> . 2007;104(6):1348-9
Brachmann-de Lange syndrome	Developmental delay with craniofacial, cardiac and GI malformations, hirsutism, and strabismus.	Preoperatively assess cardiac status (echocardiogram), check pulmonary status, anticipate difficult airway and intubation; have difficult airway cart available.
	Gastroesophageal reflux and aspiration leads to frequent pulmonary infections	SBE prophylaxis if indicated
	Fernandez-Garcia R, Perez Mencia T, Gutierrez-Jodra A, et al. Anesthetic management with laryngeal mask in a child with Brachmann-de Lange syndrome. <i>Paediatr Anaesth</i> . 2006;16(6):698-700	
Branchio-Oto-Renal (BOR) syndrome (Melnick-Fraser syndrome)	Branchial cysts or fistulae, hearing loss, pre-auricular pits, external ear malformations, renal abnormalities	Monitor heart rate. Episodic bradycardia requiring atropine or epinephrine may occur during sevoflurane administration
Taylor MH, Wilton NC. Bradycardia with sevoflurane in siblings with Branchio-oto-renal syndrome. <i>Paediatr Anaesth</i> . 2007;17(1):80-3		

Brugada syndrome	Rare in Occidentals but more common in Southeast Asia. Results from Na^+ channel defect in the myocardium. ST segment elevation in precordial leads and incomplete RBB with anatomically normal heart. Prone to ventricular tachycardia and fibrillation	Avoid parasympathetic stimulation (give anticholinergic, caution with reversal agents) and caution with drugs that affect Na^+ channels (local analgesics). Thiopental, propofol, and inhaled agents probably OK. Maintain normothermia. Apply defibrillator pads intra-op. Monitor ECG postoperatively as indicated
Camurati-Engelmann disease (progressive diaphyseal dysplasia)	Cortical thickening of long bones and skull. Skeletal pain, muscle weakness, and cranial nerve compression. Pathological fractures	Possible difficult airway; have difficult airway cart available. Care with moving and positioning. Caution with relaxants
Canavan disease	See leukodystrophy	Passariello M, Almenrader N. Anesthesia for a child with Camurati-Engelmann disease. <i>Pediatr Anesth</i> . 2013;23: 464–5
Cantrell pentalogy	Defect in the recti muscles of the abdominal wall above the umbilicus, agenesis of sternum and diaphragm, pericardial defect and cardiac malformations: cardiac septal and valvular defects present. Prone to develop severe respiratory distress and hypoxemia	Preoperatively assess cardiac status (echocardiogram), monitor for arrhythmias. Caudal epidural anesthesia during general anesthesia for noncardiac surgery has been employed successfully
		Saito T, Suzuki A, Takahata O, et al. Anesthetic management of a patient with Cantrell's pentalogy diagnosed prenatally. <i>Can Anesth Soc J</i> . 2004;51(9):946–7
		<i>Table continues on the following page.</i>

Name	Description	Anesthesia implications
Capillary angioma with thrombocytopenic purpura syndrome	See Kasabach-Merritt syndrome	
Carcinoid tumors	Carcinoid tumors (secrete vasoactive peptides) are more common in adults but may occur in children, often in the appendix but also in other sites (i.e., testis, bronchus, GI tract). Usually the diagnosis is made at histology. Carcinoid syndrome (flushing, hypotension, etc.) is very rare in children but may occur especially with malignant carcinoid tumors. Cardiac fibrosis (carcinoid heart disease) may be present	Preoperatively assess cardiac status (echocardiogram) for valvular disease. Slow stress-free induction and maintenance of anesthesia. Avoid drugs that stimulate the sympathetic system (i.e., ketamine) or release histamine (morphine, atracurium, meperidine). Octreotide is the drug of choice for perioperative control of carcinoid manifestations
	Mancuso K, Kaye AD, Boudreaux JP, et al. Carcinoid syndrome and perioperative anesthetic considerations. <i>J Clin Anesth.</i> 2011;23:329–41 Castillo JG, Silvay G, Solis J. Current concepts in diagnosis and perioperative management of carcinoid heart disease. <i>Semin Cardiothorac Vasc Anesth.</i> 2013;17(3):212–23	
Cardioauditory syndrome	See Jervell-Lange-Nielsen syndrome	
Carpenter syndrome (acrocephalopolysyndactyly)	Obesity, developmental delay, oxycephaly, peculiar facies, syndactyly, deformed extremities, cardiac defects, hypogenitalism	Preoperatively assess cardiac status (echocardiogram), SBE prophylaxis if indicated. Hypoplastic mandible may make intubation difficult; have difficult airway cart available. Monitor airway Post-op
	Batra YK, Rajeev S, Nishtala S, Grover G. Anesthetic implications of Carpenter syndrome (Acrocephalopolysyndactyly type II). <i>J Pediatr Anesth.</i> 2008;18:1235–7 Kadakia S, Helmam S N, Healy NJ, et al. Carpenter syndrome: a review for the craniofacial surgeon. <i>J Craniofac Surg.</i> 2014;25(5):1653–7	
Central core disease	Muscular dystrophy; hypotonnia without muscle wasting. Increased risk of MH	Preoperatively, assess respiratory status. Sensitive to thiopental, propofol, and respiratory depressants: avoid muscle relaxants (postoperative ventilation may be required if relaxants used). Avoid all MH triggers and prepare workstation appropriately (see Chap. 6)
	Brislin RP, Theroux MC. Core myopathies and malignant hyperthermia susceptibility: a review. <i>Pediatr Anesth.</i> 2013;23:834–41 Klingler W, Rueffert H, Lehmann-Horn F, et al. Core myopathies and risk of malignant hyperthermia. <i>Anesth Analg.</i> 2009;109(4):1167–73	

Cerebrohepatorenal syndrome (Bowen syndrome, Zellweger syndrome)	Neonatal jaundice, hepatomegaly, polycystic kidneys, muscular hypotonia, coagulopathy, dysmorphic facies, and developmental delay. CHD may be present. Hypotonia and gastroesophageal reflux predisposes to recurrent pneumonia	Preoperatively assess cardiac status (echocardiogram). Assess pulmonary status; have difficult airway cart available. Treat hypoprothrombinemia, fresh-frozen plasma may decrease surgical bleeding. Drug metabolism is impaired; titrate muscle relaxants and drugs excreted by kidneys. Ensure complete reversal of muscle relaxants
Catch 22 syndrome	See DiGeorge syndrome	Platis CM, Kachko L, Peled E, et al. Anesthesia for the child with Zellweger syndrome: a case report. <i>Pediatr Anaesth</i> . 2006;16(3):361-2.
Charcot-Marie-Tooth syndrome (peroneal muscular atrophy)	Hereditary polyneuropathy. Muscle weakness in legs and arms. Cardiac involvement: arrhythmias, conduction defects, cardiomopathy. MH has been described in two patients with CMT syndrome; however, a clear relationship is not established	Preoperatively assess cardiac status (echocardiogram). Responses to non-depolarizing muscle relaxants are usually normal. Beware of arrhythmias. MH triggering agents have been used in many patients without problems
CHARGE association	An association of Coloboma, congenital heart disease, choanal atresia, renal abnormalities, genital hypoplasia, and ear defects	Antognini JF. Anesthesia for Charcot-Marie-Tooth disease: a review of 86 cases. <i>Can J Anaesth</i> . 1992;39(4):398-400
Che'diak-Higashi syndrome	Disorder of neutrophil function, histiocyte infiltration of multiple organs. Partial albinism, immunodeficiency, panhypoproteinemia, hepatosplenomegaly, recurrent bacterial infections. Neurologic disorders and developmental delay. Steroid therapy and cytotoxic drugs may be given to induce remission	Stack CG, Wyse RK. Incidence and management of airway problems in the CHARGE association. <i>Anaesthesia</i> . 1991;46:582-5 Hara Y, Hirota K, Fukuda K. Successful airway management with use of a laryngeal mask airway in a patient with CHARGE syndrome. <i>J Anesth</i> . 2009;23(4):630-2
Ulcsoy H, Erciyas N, Ovali E, et al. Anesthesia in Che'diak-Higashi syndrome—case report. <i>Mid East J Anaesth</i>. 1995;13(1):101-5	Use sterile technique (reverse isolation). Use disposable equipment. Repeated pulmonary infections may have impaired pulmonary function. Aggressive therapy to prevent postoperative complications is required. Give supplemental steroids. Thrombocytopenia may require platelet transfusions	<i>Table continues on the following page.</i>

Name	Description	Anesthesia implications
Cherubism	Fibrous dysplasia of mandible and maxilla with intraoral masses may cause respiratory distress	Tracheal intubation may be extremely difficult; if there is acute respiratory distress, tracheotomy may be required. Have the difficult airway cart at hand. Profuse bleeding may occur during surgery of the disease mass
Monclús E, Garcés A, Artes D, et al. Oral to nasal tube exchange under fibroscopic view: a new technique for nasal intubation in a predicted difficult airway. <i>Paediatr Anaesth.</i> 2008;18(7):663–6		
Chozen syndrome	See Saethre-Chotzen syndrome	Hypoplastic mandible may make tracheal intubation difficult; have the difficult airway cart at hand. Monitor body temperature and be prepared to institute cooling. Humidify inspired gases. Use chest physiotherapy preoperatively and postoperatively
Christ-Siemens-Touraine syndrome (anhidrotic ectodermal dysplasia)	The absence of sweating and tearing. Heat intolerance due to inability to control temperature by sweating. Poor mucus formation → persistent respiratory infections	Hypoplastic mandible may make tracheal intubation difficult; have the difficult airway cart at hand. Monitor body temperature and be prepared to institute cooling. Humidify inspired gases. Use chest physiotherapy preoperatively and postoperatively
Hotta M, Koitabashi T, Umemura N, et al. Anesthetic management of a patient with hypohidrotic ectodermal dysplasia. [Japanese] English abstract. <i>Masui.</i> 2000;49(4):414–6		
Kuriakose R, Balakrishnan M. Anesthetic problems in Christ Siemens-Touraine syndrome—a case report. <i>Mid East J Anaesth.</i> 2005;18:647–50		
Chronic granulomatous disease	Inherited disorder of leukocyte function: recurrent infections with nonpathogenic organisms (Bacteria or fungi) and disordered inflammation. Poor pulmonary function. Multiple organ system involvement. Hepatomegaly in 95 % of cases, advanced liver disease leads to portal hypertension. Thrombocytopenia may be present. GI lesions predispose to regurgitation and aspiration. Bone marrow transplant may be effective therapy	Preoperatively assess respiratory status. Check coagulation. Use sterile technique (reverse isolation). Consider rapid sequence induction. Caution with drugs metabolized in the liver
Wall RF, Buzzanell CA, Epstein TA, et al. Anesthetic considerations in patients with chronic granulomatous disease. <i>J Clin Anesth.</i> 1990;2(5):306–11		
Qasim W, Gennery R. Gene therapy for primary immunodeficiencies: current status and future prospects. <i>Drugs.</i> 2014;74(9): 963–9		
Mahdaviani SA, Mohajerani SA, Rezaei N, et al. Pulmonary manifestations of chronic granulomatous disease. <i>Expert Rev Clin Immunol.</i> 2013;9(2):153–60		

CINCA syndrome	Chronic infantile neurologic cutaneous articular syndrome. Genetic autoinflammatory syndrome characterized by repeated attacks of fever and skin and joint inflammation starting in infancy. Progressive developmental delay and muscle wasting may occur. Facial dysmorphia may be associated with a CINCA syndrome. Paediatr Anaesth. 2008;18(6):575–7	Concern that stress of anesthesia and surgery may exacerbate inflammatory state. TIVA with propofol and remifentanil recommended on this basis. Caution with relaxant drugs. Potential difficult tracheal intubation; have the difficult airway cart available
Cockayne syndrome	Dysmorphic dwarfism, developmental delay, and premature senescence; patients present in early childhood. Prominent maxillae, large teeth, and sunken eyes. Ataxia, peripheral neuropathy, and flexion contractures. Associated hypertension, arteriosclerosis, and renal disease. Survival beyond second decade is unusual	Difficult tracheal intubation; have the difficult airway cart available. Associated subglottic stenosis may require a smaller diameter tube (weight vs. age appropriate). May be difficult to position. Considerations of associated cardiovascular and renal disease. A preoperative ECG for evidence of myocardial ischemia/infarction may be indicated
Collagen diseases	Systemic connective tissue diseases with variable systemic involvement. Osteoporosis, fatty infiltration of muscle, anemia, pulmonary infiltration with fibrosis. Renal involvement common. Frequently receiving steroid therapy (dermatomyositis; polyarteritis nodosa; rheumatoid arthritis; systemic lupus erythematosus)	Raghavendran S, Brown KA, Buu N. Perioperative management of patients with Cockayne syndrome—recognition of accelerated aging with growth arrest. Paediatr Anaesth. 2008;18(4):360–1 Rapin I. Disorders of nucleotide excision repair. Handb Clin Neurol. 2013;113:1637–50 Smith BL. Anaesthesia for patients with juvenile chronic arthritis (Still's disease) Anaesthesia. 1998;53(3):314 LaMont LE, Doyle SM. Orthopedic aspects of collagen disorders. Curr Opin Pediatr. 2014;26(1):79–84

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Name	Description	Anesthesia implications
Congenital heart block	Comprises less than 1% of congenital heart disease, may be associated with other CHD lesions. Defect of conduction between atrioventricular node and bundle of His or within bundle of His. Supraventricular arrhythmias may occur, and up to 20% progress to congestive heart failure and Stokes-Adams attacks. Heart rates less than 55 are poorly tolerated by infants and the response to chronotropic drugs is usually minimal	Preoperative consultation with the cardiology team is strongly recommended. Because of the possibility of intraoperative arrhythmia or increased atrioventricular block, preoperative insertion of a temporary transvenous pacemaker is usually recommended; this may be achieved via the umbilical vein in the neonate. Alternatively, transcutaneous pacing via pads may be considered. Transesophageal pacing is often not effective in infants. Sevoflurane may be useful for anesthesia as it tends to increase heart rate
Congenital insensitivity to pain and anhidrosis (CIPA)	Rare autosomal recessive disorder due to deficient nerve growth factor. Insensitivity to pain and temperature, lack of sweating, and possible developmental delay. Hyperpyrexia may occur. Delayed gastric emptying has been reported	Kussman BD, Madril DR, Thiagarajan RR et al. Anesthetic management of the neonate with congenital complete heart block: a 16-year review. <i>Paediatr Anaesth</i> . 2005;15(12):1059–66 Brändes JF, Stuth EA. Use of BIS monitor in a child with congenital insensitivity to pain with anhidrosis. <i>Paediatr Anaesth</i> . 2006;16(4):466–70 Zlotnik A, Gruenbaum SE, Rozet J, et al. Risk of aspiration during anesthesia in patients with congenital insensitivity to pain with anhidrosis: case reports and review of the literature. <i>J Anesth</i> . 2010;24(5):778–82

Conradi syndrome (chondrodyplasia epiphysealis punctata; chondrodyplasia calcificans congenita; koala bear syndrome)	Chondrodyplasty with contractures, saddle nose, microcephaly and micrognathia, developmental delay, dwarfing, congenital cataracts. CHD and renal anomalies in some other cases	Preoperatively assess cardiac status (echocardiogram), if indicated. Possible difficult airway; have difficult airway cart available
Tasker WG, Mastri AR, Gold AP. Chondrodyplasia calcificans congenita (dysplasia epiphysealis punctata): recognition of the clinical picture. <i>Am J Dis Child.</i> 1970;119:122-7.		
Porter FD, Herman GE. Malformation syndromes caused by disorders of cholesterol synthesis. <i>J Lipid Res.</i> 2011;52(1):6-34.		
Cori disease	See von Gierke disease	
Cornelia de Lange syndrome	Short stature, microcephaly, developmental delay, hirsute. Short or dysmorphic extremities, hypoplastic nipples, rib and sternal defect. Low hairline, thin lips, and downturned ("cod") mouth. Cry is low-pitched growl. CHD in 30%. Pulmonary aspiration is common, susceptible to infections (immune system defect). There is one case report of respiratory depression following midazolam	Preoperatively assess cardiac status (echocardiogram). Care with asepsis. Intubation may be difficult, and airway obstruction develops easily; have the difficult airway cart available. Caution with (avoid) midazolam
Tsusaki B, Mayhew JF. Anaesthetic implications of Cornelia de Lange syndrome. <i>Paediatr Anaesth.</i> 1998;8(2):181.		
Stevic M, Milojevic I, Bokun Z, et al. Unpredictable drug reaction in a child with Cornelia de Lange syndrome. <i>Int J Clin Pharm.</i> 2015;37:1-3.		
Boyle MI, Jespersgaard C, Brondum-Nielsen K, et al. Cornelia de Lange syndrome. <i>Clin Genet.</i> 2015;88(1):1-12.		
Costello syndrome	Developmental delay and delayed growth, coarse facies, redundant skin (neck, palms, soles), and papillomata (oral, nasal, anal). Cardiac involvement is common. CHD in 30%, hypertrophic cardiomyopathy in 20 %. Endocrine problems include hypopituitarism, hypothyroid, and hypoadrenal states. Hypoglycemia may occur. Potential airway problems include short neck, choanal atresia, macroglossia, and laryngeal papillomata	Preoperatively assess cardiac status (echocardiogram). Check endocrine status. Caution with the airway; have the difficult airway cart at hand. Monitor postoperative ventilation. Monitor blood glucose during long procedures
Katcher K, Bothwell M, Tobias JD. Anaesthetic implications of Costello syndrome. <i>Paediatr Anaesth.</i> 2003;13(3):257-62.		

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Name	Description	Anesthesia implications
Gretinism (congenital hypothyroidism)	Goiter; hypothyroidism secondary to defective synthesis of thyroid hormone. Large tongue. Respiratory center very sensitive to depression; CO ₂ retention common. Hypoglycemia, hyponatremia, hypotension, low cardiac output. Early treatment with levothyroxine is essential to prevent developmental delay	Correct hypothyroidism and anemia preoperatively if possible. Intravenous triiodothyronine may be useful. Airway problems due to large tongue; have difficult airway cart at hand. Monitor body temperature; use forced hot air warming blankets. Do not use myocardial depressants. Transfuse carefully; overtransfusion is poorly tolerated because of decreased myocardial contractility
Mason KP, Koka BY, Eldredge EA, et al. Perioperative considerations in a hypothyroid infant with hepatic haemangioma. <i>Paediatr Anaesth.</i> 2001;11(2):228–32 Grosse SD, Van Viet G. Prevention of intellectual disability through screening for congenital hypothyroidism: how much and at what level? <i>Arch Dis Child.</i> 2011;96(4):374–9 Krude H, Kuhnen P, Biebermann H. Treatment of congenital thyroid dysfunction: Achievements and challenges. <i>Best Pract Res Clin Endocrinol Metab.</i> 2015;29(3):399–413		
Cri du chat syndrome	Chromosome 5p abnormality causing developmental delay, abnormal catlike cry, microcephaly, round face, hypertelorism. In some, ears abnormal, micrognathia, epiglottis, and larynx small. CHD may be present	Preoperatively assess cardiac status (echocardiogram) if indicated. Airway problems: stridor, laryngomalacia. Tracheal intubation may be difficult; have the difficult airway cart at hand. A small size tube may be required. Risk of postextubation croup
Brislin RP, Stayer SA, Schwartz RE. Anaesthetic considerations for the patient with cri du chat syndrome. <i>Paediatr Anaesth.</i> 1995;5(2):139–41 dos Santos KM, de Rezende DC, Borges ZD. Anesthetic management of a patient with Cri Du Chat syndrome. <i>Case report. Rev Bras Anestesiol.</i> 2010;60(6):630–1		
Crouzon syndrome	Craniostenosis, hypertelorism, parrot beak nose, hypoplastic maxilla, and exophthalmos because of chromosomal bony defect causing premature closure of cranial sutures and intracranial hypertension	Eye protection important. Mask ventilation difficult, requires jaw thrust ± oral airway; have the difficult airway cart at hand. Tracheal intubation usually easy. Postoperative airway obstruction is common; elective tracheostomy may be indicated. Beware of additional airway problems related to external fixation devices postsurgery for maxillary distraction which may limit access to the mouth and lead to trismus
Payne JF, Cranston AJ. Postoperative airway problems in a child with Crouzon's syndrome. <i>Paediatr Anaesth.</i> 1995;5:331 Roche I, Frawley G, Heggie A. Difficult tracheal intubation induced by maxillary distraction devices in craniosynostosis syndromes. <i>Pediatr Anaesth.</i> 2002;12(3):227–34		

Cutis laxa	Elastic fiber degeneration; pendulous skin, frequent hernias. Recurrent pulmonary infections, emphysema and cor pulmonale, arterial fragility	Preoperatively assess pulmonary status. Use sterile technique. Difficulty maintaining IV line due to poor tissues. Excess soft tissues around larynx may cause upper airway obstruction; have the difficult airway cart at hand
Pandey R, Garg R, Manikandan R, et al. Perianesthetic management of generalized congenital cutis laxa syndrome associated with pulmonary stenosis undergoing inguinal hernia repair. <i>Pediatr Anesth.</i> 2008;18:907-9		
Dandy-Walker syndrome	See hydrocephalus (page 237)	
Deletion 9p syndrome	Partial deletion of short arm of chromosome 9 is associated with developmental delay, trigonocephaly, dysmorphic facies, small mouth, cleft palate, choanal stenosis, cardiac and renal disease. Gastroesophageal reflux and aspiration leading to repeated pulmonary infections	Difficult airway and intubation; have the difficult airway cart available. A smaller diameter tracheal tube may be required. Use short-acting drugs for rapid recovery of airway reflexes
Cakmakkaya OS, Balkan M, Altintas F, et al. Anesthetic management in a child with deletion 9p syndrome. <i>Pediatr Anaesth.</i> 2007;17(1):88-9		
Dermatomyositis	See collagen disease	
DiGeorge syndrome (Catch 22 syndrome, deletion 22q syndrome, Velocardiofacial syndrome, third and fourth brachial arch/pharyngeal pouch syndrome)	Aortic arch and cardiac abnormalities. Thymus and parathyroids absent, hypoparathyroidism, low serum Ca resulting in tetany and stridor. Often associated with chromosome 22 defect. Immune deficiency: susceptibility to fungal and viral infections; recurrent chest infections. Treated by thymic transplants	Preoperatively assess cardiac status (echocardiogram) and consult cardiology. Use sterile technique (reverse isolation). Donor blood must be previously irradiated (30 Gy) to prevent graft-versus-host reaction. Check calcium levels Ca^{++} ; an infusion may be required. Caution with intubation; may have laryngomalacia and short trachea
Singh VP, Agarwal RC, Sanyal S, et al. Anesthesia for DiGeorge's syndrome. <i>J Cardiothorac Vasc Anesth.</i> 1997;11:811		
Huang RX, Shapiro NL. Structural airway anomalies in patients with DiGeorge syndrome: a current review. <i>Am J Otolaryngol.</i> 2000;21:326-30		
Yeoh IY, Scavonetto F, Hamlin RJ, et al. Perioperative management of patients with DiGeorge syndrome undergoing cardiac surgery. <i>J Cardiothorac Vasc Anesth.</i> 2014;28(4):983-9		
<i>Table continues on the following page.</i>		

Name	Description	Anesthesia implications
Donohue syndrome	See leprechaunism	
Down syndrome (see Chap. 6)		
Duchenne muscular dystrophy	Progressive pseudohypertrophy of muscles with cardiomyopathy in most cases. Predominantly occurs in males; a milder form, Becker syndrome, also occurs in females. Genetic cause: X-linked recessive mutation in dystrophin gene at chromosome 21. May be subclinical until 2–6 years and many die before 20 years of age	Preoperatively assess cardiac status (echocardiogram) particularly in adolescents. Succinylcholine contraindicated (may cause hyperkalemic cardiac arrest). DMD may be undiagnosed in infancy, leading to recommendation to avoid elective use of succinylcholine in boys less than 6 years of age. Inhalational agents may cause rhabdomyolysis; TIVA is preferred. Respiratory depression occurs easily; titrate drug dosage to limit cardiorespiratory depression. Give non-depolarizing muscle relaxants judiciously and monitor block. Use local analgesia whenever possible. IPPV support may be needed postoperatively

- Yemen TA, McClain C. Muscular dystrophy, anesthesia and the safety of inhalational agents revisited: again. [Editorial] *Pediatr Anaesth*. 2006;16(2):105–8.
- Segura LG, Lorenz JD, Weingarten TN, et al. Anesthesia and Duchenne or Becker muscular dystrophy: review of 117 anesthetic exposures. *Pediatr Anesth*. 2013;23:855–64.
- Kako H, Corridore M, Kean J. Dexmedetomidine and ketamine sedation for muscle biopsies in patients with Duchenne muscular dystrophy. *Paediatr Anaesth*. 2014;24(8):851–6.
- Brunklaus A, Parish E, Muntoni F, et al. The value of cardiac MRI versus echocardiography in the pre-operative assessment of patients with Duchenne muscular dystrophy. *Eur J Paediatr Neurol*. 2015;19(4):395–401.

Dutch-Kentucky syndrome	See Trismus-pseudocamptodactyly	
EEC syndrome (ectrodactyly, ectodermal dysplasia, and cleft lip and palate)	Congenital anomaly complex. Lobster claw deformity, dysplasia of all ectodermal elements (including central nervous system), with disordered temperature control (hypohidrosis plus central defect). Decreased tearing, conjunctivitis, blepharitis. Cleft lip and palate, respiratory tract infections, genitourinary anomalies, malnutrition, and anemia. Developmental delay in 8 %	Assess nutrition and anemia. Preoperative chest physiotherapy advised; avoid anticholinergics (i.e., atropine [effect on sweating]). Extreme care with skin required; position and pad carefully. Protect eyes. Tracheal intubation may be difficult with cleft palate; have the difficult airway cart available. Be prepared to maintain normothermia using heating/cooling blankets, etc.
Mizushima A, Satoyoshi M. Anaesthetic problems in a child with ectrodactyly, ectodermal dysplasia and cleft lip palate: the EEC syndrome. <i>Anaesthesia</i> . 1992;47:137-40		
Edwards syndrome (trisomy 18 [E])	Developmental delay and dysmorphic changes, micrognathia in 80 %, hypotonia. CHD in 95 %, renal malformations in 50-80 %. Most die in infancy	Preoperatively assess cardiac status (echocardiogram). Airway and tracheal intubation may be difficult; have the difficult airway cart available. Use caution with drugs excreted by kidney
Courreges P, Nieuviarts R, Lecoutre D. Anaesthetic management for Edward's syndrome. <i>Paediatr Anaesth</i> . 2003;13(3):267-9		
Ehlers-Danlos syndrome (cutis hyperelastica)	Collagen abnormality: hyperelasticity and fragile tissues; dissecting aneurysm of aorta, fragility of other blood vessels; ECG conduction abnormalities. Bleeding diathesis; hernias. May have heart, lung, and gastrointestinal malformations	Preoperative ECG. Difficult to maintain IV line and prevent complications of IV infusions. Poor tissues and clotting defect may lead to increased surgical bleeding. Spontaneous pneumothorax may occur. Monitor for ECG conduction abnormalities. Caution with neck movement. Neuraxial anaesthesia relatively contraindicated (risk of bleeding)
Lane D. Anaesthetic implications of vascular type Ehlers-Danlos syndrome. <i>Anaesth Intens Care</i> . 2006;34(4):501-5		<i>Table continues on the following page.</i>

Name	Description	Anesthesia implications
Eisenmenger syndrome	Association of high pulmonary vascular resistance (pulmonary hypertension) and an intracardiac or extracardiac R-L shunt. Dyspnea, fatigue, cyanosis, finger clubbing, and cardiac failure. Often associated with Down syndrome	<p>Assess cardiac status (echocardiogram) and consult cardiology. Assess severity of R-L cardiac shunt; shunt may increase with hypoxia, hypercarbia, or acidosis.</p> <p>Inhalation induction with potent anesthetic agents has been utilized with great caution. Alternatively, a slow intravenous induction may be performed as rapid effect from IV agents and may occur. Avoid drugs or airway events that may increase PVR (ie., hypercarbia, hypoxemia, acidosis) or decrease SVR (ie., high-dose thiopental or propofol, intravenous vasodilators) significantly. Caution with controlled ventilation to maintain lung volume but minimize intrathoracic pressure. Care with fluid therapy; hypovolemia is not well tolerated, and overtransfusion may lead to R ventricular failure. Polycythemia increases viscosity but caution required if hemodiluting as it decreases oxygen carrying capacity. Despite all the potential problems, many children tolerate a well-conducted anesthetic.</p> <p>Epidural anesthesia has been used successfully</p> <p>Lyons B, Motherway C, Casey W, et al. The anaesthetic management of the child with Eisenmenger's syndrome. <i>Can J Anaesth.</i> 1995;42:904-9</p> <p>Bennett JM, Ehrenfeld JM, Markham L, et al. Anesthetic management and outcomes for patients with pulmonary hypertension and Eisenmenger syndrome: a review of institutional experience. <i>J Clin Anesth.</i> 2014;26(4):286-93</p>
Elfin facies syndrome	See Williams syndrome	
Ellis-van Creveld syndrome (chondroectodermal/mesoectodermal dysplasia)	Ectodermal defects causing skeletal dwarfism, cardiac anomalies (50%), chest wall defects, and poor lung function. Short limbs, polydactyly, and hypoplastic nails. May have abnormal maxillae, cleft palate, cleft lip, peg teeth, hepatosplenomegaly. Patients often die in infancy	<p>Preoperatively assess cardiac status (echocardiogram). Tracheal intubation can be routine, but airway problems and peg teeth may make intubation difficult; have the difficult airway cart available</p> <p>Abelès AI, Tobias JD. Anesthetic implications of Ellis-van Creveld syndrome. <i>J Clin Anesth.</i> 2008;20:e18-21</p>

Bosinophilic granuloma	See histiocytosis X	
Epidermolysis bullosa (Herlitz syndrome)	<p>Skin cleavage at dermal-epidermal junction, resulting in erosions and blisters from minor trauma to skin or mucous membrane. The disease occurs in several forms: Simplex: dominant, maps to chromosome 17. Relatively mild with rapid healing and little scarring. Lethal: recessive, maps to chromosome 12. Junctional epidermolysis bullosa. Severe, presents at birth, leads to extensive scarring and death (often from sepsis) usually before 2 years of age.</p> <p>Dystrophic: Recessive, maps to chromosome 12. Very rare but severe; lesions heal slowly with extensive scarring. Structures may form and involve the pharynx, larynx and esophagus. Digital fusion occurs ("mitten hand").</p> <p>Nutritional deprivation leads to growth retardation and anemia. Infections are common</p>	<p>Antibiotic prophylaxis perioperatively to prevent secondary infections. Check history of steroid therapy.</p> <p>Use sterile technique (reverse isolation). Airway difficulty: oral lesions, adhesion of tongue, intraoral scarring; avoid tracheal intubation and/or instrumentation of the airway if possible as bullae may develop; otherwise, lubricate tube and laryngoscope generously. Prevent trauma to skin or mucous membranes, especially from friction or shearing movements. Use very generous lubricated padding. Use insufflation or a well-padded and lubricated mask for inhalation anesthesia or use propofol or ketamine. Care with a tourniquet and use optical ointment. Avoid adhesive tapes (patients/parents often know which tapes can be tolerated); an oximeter probe may be held in place with a lubricated gauze bandage. ECG pads should be coated with surgical lubricant and placed under the child. Regional analgesia may be appropriate for limb surgery</p>

Herod J, Denyer I, Goldman A, et al. Epidermolysis bullosa in children: pathophysiology, anaesthesia and pain management. *Paediatr Anaesth*. 2002;12(5):388-97

Saraf S V, Mandawade NJ, Gore SK, et al. Epidermolysis bullosa: Careful monitoring and no touch principle for anaesthesia management. *J Anaesthesiol Clin Pharmacol*. 2013;29:390-3

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Name	Description	Anesthesia implications
Erythema multiforme	See Stevens-Johnson syndrome	
Eulenburg periodic paralysis	See paramyotonia congenita	
Escobar syndrome (multiple pterygium syndrome)	Autosomal recessive progressive disease; multiple joint contractures, facial and genital anomalies, severe kyphoscoliosis. Normal intellect	Difficult airway and intubation; have the difficult airway cart available. Difficulty of intubation increases with age. IV access may be limited. Caution with padding and positioning. Epidural analgesia may be appropriate despite deformity Kuzma PJ, Calkins MD, Kline MD, et al. The anesthetic management of patients with multiple pterygium syndrome. <i>Anesth Analg</i> . 1995;83:430–2 Kachko L, Platis CM, Konen O, et al. Lumbar epidural anesthesia for the child with Escobar syndrome. <i>Paediatr Anaesth</i> . 2006;16(6):700–2 Sertoz N, Gunay H, Karaman S. Anesthetic approach to a patient with multiple pterygium (Escobar) syndrome. <i>Pediatr Anesth</i> . 2012;22:490–2 Matthew S, Chaudhuri S, Arun Kumar H, et al. Airway management in Escobar syndrome: A formidable challenge. <i>Indian J Anaesth</i> . 2013;57(6):603–5
Fabry disease (angiokeratomas corporis diffusum)	X-linked lipid storage disorder. Lipid deposition in blood vessels causes periodic very severe pain and fever crises. Corneal opacities. Dark telangiectasia, particularly around genitals and buttocks; hypertension, myocardial ischemia, renal failure. Hypertension and myocardial ischemia	Preoperatively assess cardiac status (echocardiogram) for myocardial function; ECG for myocardial ischemia; BUN and creatinine for renal function; caution with drugs excreted by the kidneys if renal dysfunction is present Woolley J, Pichel AC. Peri-operative considerations for Anderson-Fabry disease. <i>Anesthesia</i> . 2008;63(1):101–2
Familial dysautonomia	See Riley-Day syndrome	
Familial osteodysplasia	See Andersen syndrome	
Familial periodic paralysis	Periodic muscle weakness secondary to serum K ⁺ disturbance (hypokalemia or hyperkalemia). Muscle weakness in the hypokalemic variety is caused by massive uptake of K ⁺ into muscles and thus decreased serum K ⁺	Monitor serum K ⁺ , glucose, and the ECG; maintain normokalemia and normoglycemia. Avoid muscle relaxants; hyperkalemia from Sch. maintain body temperature. Avoid excessive glucose solutions. TIVA with propofol and remifentanil has been successful in adult patients. Regional analgesia may be useful Kim JB. Channelopathies. <i>Korean J Pediatr</i> . 2014;57:1–18 Bandschapp O, Iaizzo PA. Pathophysiology and anesthetic considerations for patients with myotonia congenita or periodic paroxysms. <i>Pediatr Anesth</i> . 2013;23:824–33

Fanconi syndrome (anemia with renal tubular acidosis)	Usually secondary to cystinosis. Proximal tubular defect: impaired renal function; acidosis, K ⁺ loss, dehydration. Older children may have thyroid and pancreatic dysfunction secondary to cysteine deposition. May present for renal transplant in second decade	Treat electrolyte and acid-base abnormalities: Caution with drugs excreted by kidneys. Cisatracurium is the preferred muscle relaxant. Be aware of possibility of other metabolic or endocrine defects
Farber disease (lipogranulomatosis)	Sphingomyelin deposition: widespread visceral lipogranulomas, especially in CNS. General systemic involvement leading to cardiac, renal failure	Preoperatively assess cardiac (ECHO) and renal status. Deposits in oral cavity, pharynx, and larynx; possible difficult intubation. Have the difficult airway cart available
Favism (glucose-6-phosphate dehydrogenase (G6PD) deficiency)	Diathesis for spontaneous/induced (drugs, fava beans, infection) hemolytic anemia	Asada A, Tatekawa S, Terai T, et al. The anesthetic implications of a patient with Farber's lipogranulomatosis. <i>Anesthesiology</i> . 1994;80:206–9 Do not give drugs that cause hemolysis (i.e., acetylsalicylic acid, phenacetin, sulfonamides, quinidine, methylene blue). Midazolam, sevoflurane, nitrous oxide, and rocuronium are all acceptable. Anemia; transfuse if necessary
		Wada R, Hino H, Ando Y. Case of laparoscopic cholecystectomy in a patient with glucose-6-dihydrogenase deficiency. [English abstract.] <i>Masui</i> . 2008;57(2):200–2 Elbassi AR, Rowshan HH. Perioperative management of the glucose-6-phosphate dehydrogenase deficient patient: a review of literature. <i>Anesth. Prog.</i> 2009;56(3):86–91

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Name	Description	Anesthesia implications
Fetal alcohol syndrome	Abnormalities of the infant due to maternal heavy alcohol consumption: growth retardation, intellectual impairment, craniofacial abnormalities (microcephaly, microphthalmia, hypoplastic upper lip, flat maxilla), cardiac defects (especially ventricular septal defect), renal abnormalities, and inguinal hernia	Preoperatively assess cardiac status (echocardiogram). May have difficulty with intubation; have the difficult airway cart available. Evaluate renal function
	Clаррен SK, Smith DW. The fetal alcohol syndrome. N Engl J Med. 1978;298:1063-7 Finucaine BT. Difficult intubation associated with the fetal alcohol syndrome. Can Anaesth Soc J. 1980;27:574-5 Burd L, Deal E, Rios R, et al. Congenital heart defects and fetal alcohol spectrum disorders. Congenit Heart Dis. 2007;2(4):250-5	
Fibrodysplasia ossificans progressiva	See myositis ossificans	Airway may contain papillomas resulting in difficulties with ventilation. Potential difficult airway; have the difficult airway cart available
Focal dermal hypoplasia (Goltz syndrome)	Multifarious features, including multiple papillomas of mucous membranes, skin, ontogenetic cysts, and giant cell tumors of bone	Airway may contain papillomas resulting in difficulties with ventilation. Potential difficult airway; have the difficult airway cart available
	Holtzman RS. Airway involvement and anesthesia management in Goltz syndrome. J Clin Anesth. 1991;3:422-5 Gosavi KS, Mundada SD. Anesthetic management in Gorlin-Goltz syndrome. Indian J Anaesth. 2012;56(4):394-6	

Forbes disease (glycogen storage disease type III)	See Von Gierke Disease	
Freeman-Sheldon syndrome (whistling face syndrome)	Progressive congenital myopathy and dysplasia with autosomal or X-linked recessive inheritance. Increased tone and fibrosis of facial muscles. Hypertelorism, microstomia, and micrognathia. Leads to flexion contracture of limbs. Strabismus and inguinal hernia common. Later, kyphoscoliosis causes restrictive lung disease. There is a rare association with malignant hyperthermia	Very difficult intubation primarily due to microstomia and micrognathia; tight facial muscles will not relax with neuromuscular blockade, and masseter muscle rigidity may follow halothane or succinylcholine. Venous access difficult due to limb flexion contractures. Pulmonary function may be impaired (late). Insertion of an LMA (if microstomia is not severe) facilitates fiberoptic bronchoscopy and intubation. Regional analgesia maybe useful for surgery and/or postoperative pain. An MH trigger-free anesthetic technique may be indicated in these children
Friedreich ataxia	Progressive degeneration of the cerebellum, lateral and posterior column of spinal cord; scoliosis; myocardial degeneration and fibrosis, leading to failure and serious arrhythmias. Glucose intolerance; 10 % are diabetic	Preoperatively assess metabolic state and cardiac status (ECHO). Care with cardiac depressant drugs; monitor ECG. TIVA with propofol and remifentanil has been recommended. Bis responses appear normal. Response to relaxants uncertain; avoid if possible. Otherwise cisatracurium with block monitor suggested

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Name	Description	Anesthesia implications
Gangliosidoses	Invariably fatal. Supportive measures only treatment	Administer anti-seizure medications on the morning of surgery. Preoperatively assess cardiac status (echocardiogram). Progressive neurologic loss leads to respiratory complications; assess cardiopulmonary status
GM1, type 1	Acute onset in infancy: rapid neurologic decline, severe bone abnormalities; pulmonary infiltration common.	
GM1, type 2	Death by 2 years of age	
GM2 (Tay-Sachs disease; Sandhoff disease)	Onset in infancy: progressive psychomotor deterioration, blindness, seizures. Predominantly in Ashkenazi Jewish heritage. Death by 5 years	
	Onset in early childhood: few somatic changes. Death from cardiopulmonary causes by 10 year of age	
	Rare juvenile variants: same features; longer survival	
	Maegawa GH, Stockley T, Tropak M, et al. The natural history of juvenile or subacute GM2 gangliosidosis: 21 new cases and literature review of 134 previously reported. Pediatrics. 2006;118(5):e1550–62	
	Venuopalap P, Joshi SN. Cardiac involvement in infantile Sandhoff disease. J Paediatr Child Health. 2002;38(1):98–100	
GAPO syndrome (growth retardation, alopecia, pseudo-anodontia, and optic atrophy)	Growth retardation, alopecia, optic atrophy, large tongue, and unerupted teeth, associated with dilated cardiomopathy, pulmonary hypertension, glaucoma	Preoperatively assess cardiac status (echocardiogram) and consult cardiology. Facial anomalies, large tongue, and short neck may lead to difficult airway; have the difficult airway cart available. Nasal passages too small for FOB intubation: oral approach preferred. Airway improved in sitting position
Sinha R, Trikha A, Laha A. Anesthetic management of a patient with GAPO syndrome for glaucoma surgery. Pediatr Anesth. 2011;21:910–12		
Nanda A, Al-Ateeqi WA, Al-Khwari MA, et al. GAPO syndrome: a report of two siblings and a review of literature. Pediatr Dermatol. 2010;27(2):156–61		

Gardner syndrome	Familial polyposis of colon; bone tumors, sebaceous cysts, fibromas. Associated with bowel adenocarcinoma and intussusception	No specific anesthesia problems described but anemia is possible due to bleeding polyps
Gaucher disease	Cerebroside accumulation in CNS, liver, spleen, etc. due to deficient lysosomal enzyme glucocerebrosidase. Serum acid phosphatase increased. May have cardiac involvement. Pulmonary disease from aspiration (pseudobulbar palsy) and hepatosplenomegaly. Hypersplenism may cause platelet deficiency. If obvious neurologic signs: usually fatal in infancy (neuropathic type 2 and 3). If nonneuropathic (type 1), course is more chronic with bone pain, fractures, etc. Some may respond to enzyme replacement therapy	Preoperatively assess cardiac status (echocardiogram). Assess pulmonary status; beware of aspiration. Tracheal intubation usually routine but may be difficult if trismus, neck, or airway infiltration; have the difficult airway cart available. Surgical bleeding may be a major problem; treat coagulation disorders and correct anemia
Glanzmann disease (thrombasthenia)	Abnormal platelet function, leading to mild thrombocytopenic purpura; abnormality of high-energy phosphate mechanisms. Considerable bleeding risk with any surgical procedure	No specific therapy for bleeding; platelet transfusions disappointing. Therapy with recombinant activated factor VII plus antithrombolytic agents may be helpful. May have history of steroid therapy
	Gunaydin B, Ozkose Z, Pezek S. Recombinant activated factor VII and epsilon aminocaproic acid treatment of a patient with Glanzmann's thrombasthenia for nasal polypectomy. <i>J Anesth.</i> 2007;21(1):106-7	<i>Table continues on the following page.</i>

Name	Description	Anesthesia implications
Glucose-6-phosphate dehydrogenase (G6PD) deficiency	See favism	
Glycogen storage disease		
Type I	See von Gierke disease	
Type II	See Pompe disease	
Type III (Cori disease; Forbes disease)	See von Gierke disease	
Type IV	See von Gierke disease	
Type V	See Andersen disease	
Type VI (Hers disease)	See McArdle disease	
Type VII	See muscle phosphofructokinase deficiency	
Type VIII	See hepatic phosphorylase kinase deficiency	
Goldenhar syndrome (oculoauriculovertebral syndrome; hemifacial microsomia)	Unilateral mandibular hypoplasia; CHD in 35%. Embryonic malformation due to chromosome 22 trisomy. Vertebral abnormalities may limit neck extension	Preoperatively assess cardiac status (echocardiogram) if indicated. Airway problems; may be extremely difficult to hold a mask in place and maintain an airway once anesthesia induced. Tracheal intubation may be very difficult (bilateral) or very easy (unilateral or left-sided lesion). If right TMJ and mandible are involved or bilateral disease, increased difficulty for intubation. Have an LMA (ProSeal if available) ready plus all difficult airway supplies (difficult airway cart). A GlideScope and fiberoptic laryngoscope may be very useful. Ready for a surgical airway. TIVA with propofol and remifentanil may facilitate rapid emergence. Extubate the trachea awake

Nargozian C, Ririe DG, Bennun RD, et al. Hemifacial microstomia: anatomical prediction of difficult intubation. <i>Pediatr Anesth.</i> 1999;9:393–8	
Altintas F, Cakmakkaya OS. General anesthesia for a child with Goldenhar syndrome. <i>Pediatr Anesth.</i> 2005;15(6):529–30	
Milne AD, Dower AM, and Hackmann T. Airway management using the pediatric GlideScope in a child with Goldenhar syndrome and atypical plasma cholinesterase. <i>Pediatr Anaesth.</i> 2007;17(5):484–7	
Ozlu O, Simsek S, Alacakir H, et al. Goldenhar syndrome and intubation with the fiberoptic bronchoscope. <i>Paediatr Anaesth.</i> 2008;18(8):793–4	
Aydogan MS, Begec Z, Erdogan MA, et al. Airway management using the ProSeal laryngeal mask airway in a child with Goldenhar syndrome. <i>Eur Rev Med Pharmacol Sci.</i> 2012;16(4):559–61	
Goltz syndrome	See focal dermal hypoplasia and Gorlin-Goltz syndrome
Gonadal dysgenesis	See Turner syndrome
Gorham syndrome (disappearing bone disease)	Massive osteolysis and lymphangiomatosis. Pathologic fractures and bony deformities with neurologic and respiratory complications. Severe kyphoscoliosis may be present. Problems relate to bony involvement: cervical spine subluxation, thoracic deformity leading to respiratory failure. Pleural effusions or chylothorax may be present. Normal intellect
Szabo C, Habre W. Gorham syndrome: anaesthetic management. <i>Anaesthesia.</i> 2000;55(2):157–9	Tracheal intubation may be difficult; have the difficult airway cart available. Cervical spine precautions indicated. Caution with protein-bound drugs if chylothorax has caused hypoproteinemia. Avoid succinylcholine to prevent fasciculations that could break bones. Caution with transport and positioning. Postoperative ventilation may be required
Gorlin-Chaudhry-Moss syndrome	Craniofacial dysostosis, patent ductus arteriosus, hypertelorism, hypoplasia of labia majora, dental and eye anomalies. Normal intelligence
Ortalli G, Tiberio I, Mammmana G. Gorlin-Goltz syndrome. Observation of a case. [Italian] <i>English abstract.</i> <i>Minerva Anesthesiol.</i> 1991;57(4):161–3	Asymmetry of the head may cause a difficult airway; have the difficult airway cart available
Gorlin-Goltz syndrome (basal cell nevus syndrome)	Multiple nevoid basal cell carcinomas, hypertelorism, mandibular prognathism, multiple jaw cysts and fibrosarcomas, kyphoscoliosis, incomplete segmentation of cervical and thoracic vertebrae; congenital hydrocephalus, developmental delay, etc.
Debu A, Sleth JC, Girard C, et al. The use of subcutaneous infusion tumescent anesthesia in photodynamic therapy pain control. <i>Pediatr Anesth.</i> 2012;22:600–1	<i>Table continues on the following page.</i>

Name	Description	Anesthesia implications
Grönblad-Strandberg syndrome (pseudoxanthoma elasticum)	Degeneration of elastic tissue in the skin, eye, and cardiovascular system; rupture of arteries, especially in gastrointestinal tract; hypertension; arterial calcification; occlusion of cerebral and coronary arteries	Preoperatively assess cardiac status (echocardiogram and ECG). Manage as for coronary artery disease. Difficult to maintain IV cannula in situ. Prevent tachycardia and hypertension (rupture of aneurysms). Avoid an arterial line (vessel damage). Avoid NG tube (bleeding)
Guillain-Barré syndrome (acute [idiopathic] polyneuritis)	Acute polyneuropathy; progressive peripheral neuritis; usually involving cranial nerves; bulbar palsy with hypoventilation and hypotension. May follow an infection or surgery. Early treatment by plasma exchange and immunotherapy is highly desirable to limit the disease. Some require tracheotomy and ventilatory support	Krechel SL, Ramirez-Inawat RC, Fabian LW. Anesthetic considerations in pseudoxanthoma elasticum. <i>Anesth Analg</i> . 1981;60(5):344-7 Do not use succinylcholine for at least 3 months after onset of polyneuritis and until lower motor neuron deficit resolves (risk of ↑ K ⁺ release). May have serious hemodynamic instability. Disease may first present in the postoperative period with weakness, loss of tendon reflexes, and other signs of polyneuropathy
Hallervorden-Spatz disease	Autosomal recessive disorder of basal ganglia: leading to dementia, dystonia, and chorea. Torticollis, scoliosis, and trismus develop. Episodes of airway obstruction and desaturation may occur during posturing. Stereotactic thalamotomy may improve the neurologic state considerably	Jones GD, Wilmsurst JM, Sykes K, et al. Guillain-Barre syndrome: delayed diagnosis following anaesthesia. <i>Pediatr Anaesth</i> . 1999;9(6):539-542 Hughes RA, Swan AV, van Doorn PA. Intravenous immunoglobulin for Guillain-Barre syndrome. <i>Cochrane Database Syst Rev</i> . 2014;9:CD002063 Preoperatively assess pulmonary status. Inhalation induction of anesthesia leads to relaxation of abnormal posturing and trismus and facilitates intubation. Avoid succinylcholine (↑ K ⁺ release may intensify rigidity) or rapid sequence induction (in case of difficult intubation); have difficult airway cart available. Responses to other anesthetics are normal

Keegan MT, Flick RP, Matsumoto JY, et al. Anesthetic management for two-stage computer-assisted, stereotactic thalamotomy in a child with Hallervorden-Spatz Disease. *J Neurosurg Anesthesiol*. 2000;12(2):107-11
Sinha R, Riyani G, Bhattacharjee S. Anaesthetic management of a child with panthothenate kinase-associated neurodegeneration. *Indian J Anaesth*. 2015;59(1):43-6

Hand-Schüller-Christian syndrome	See Histiocytosis X	
Harlequin syndrome	Skin color changes with demarcation line bisecting the body. Hemifacial sweating and flushing due to unilateral sympathectomy	No contraindications to routine anesthesia. Hemifacial flushing may develop during neck surgery due to interference with sympathetic ganglia
Hecht-Beals syndrome	Transient postoperative harlequin syndrome combined with Horner's syndrome in a pediatric patient after neck mass excision.	Developmental delay, arachnodactyl, kyphoscoliosis, and multiple congenital joint contractures
Hemangioma with thrombocytopenia		Negata O, Tateoka A, Shiro R, et al. Anaesthetic management of two paediatric patients with Hecht-Beals syndrome. <i>Paediatr Anaesth</i> . 1999;9(5):444-7 Kumar A, Chandran R, Khanna P, et al. Successful difficult airway management in a child with Hecht-Beals syndrome. <i>Indian J Anesth</i> . 2012;56(6):591-2
Hemolytic uremic syndrome	Usually occurs in 1- to 2-year-olds; prodromal (usually gastrointestinal) infection followed by sudden onset of renal failure, hemolytic anemia, and thrombocytopenia. All systems may be involved: cardiovascular system—severe hypertension, myocarditis, and congestive cardiac failure; respiratory-pulmonary insufficiency. Central nervous system: depression progressing to drowsiness, seizures, and coma. Hepatosplenomegaly with hepatic dysfunction, seizures, and coma. Coagulopathy: thrombocytopenia, decreased platelet function, prolonged prothrombin time, and bleeding time. Treatment is by blood transfusion, renal dialysis, and symptomatic therapy for other disorders	Administered anti-seizure medications are given on the morning of surgery. Preoperatively assess cardiac status (echocardiogram and ECG) for cardiac function or evidence of myocarditis. Comprehensive respiratory assessment required. Correct electrolyte, acid-base, and coagulation abnormalities. May have full stomach (gastrointestinal dysfunction) requiring RSI. Short-acting anesthetics that do not require normal hepatic or renal function recommended. Intensive continuous monitoring of biochemistry needed intraoperatively and postoperatively
		Johnson GD, Rosales JK. The haemolytic uraemic syndrome and anaesthesia. <i>Can J Anaesth</i> . 1987;34:196-9 Grisaru S. Management of hemolytic-uremic syndrome in children. <i>Int J Nephrol Renovasc Dis</i> . 2014;7:231-9

Table continues on the following page.

Name	Description	Anesthesia implications
Hepatic phosphorylase kinase deficiency (glycogen storage disease type VIII)	Hepatomegaly; increased liver glycogen concentration. Minor growth retardation and delayed motor development. Mild to moderate hypoglycemia may occur. Many children are asymptomatic and lead normal lives on a special diet. Occasionally disease status is more severe with hypoglycemia and acidosis. Very rarely a form of this disease may cause severe neonatal hypoglycemia	Preoperatively assess metabolic status and history. No specific anesthesia complications reported. Monitor glucose levels and administer glucose-containing solution; check acid-base if indicated perioperatively
Echaniiz-Laguna A, Akman HO, Mohr M, et al. Muscle phosphorylase b kinase deficiency revisited. Neuromuscul Disord. 2010;20(2):125-7		
Herlitz syndrome	See epidermolysis bullosa	
Hermansky-Pudlak syndrome	Albinism: bleeding diathesis due to platelet Abnormality and life threatening pulmonary fibrosis	Monitor coagulation. Careful preoperative assessment of pulmonary status. May require platelet transfusion during surgery
Haddadin AS, Ayoub CM, Sevarino FB, et al. Evaluation of hemostasis by the Clot Signature Analyzer: a potentially valuable device for the anesthesiologist. J Clin Monit Comp. 1999;15(2):125-9 Bin Saeedan M, Faheem Mohammed S, Mohammed TI. Hermansky-pudlak syndrome: high-resolution computed tomography findings and literature review. Curr Probl Diagn Radiol. 2015;44(4):383-5		
Hers disease	See von Gierke disease	
Histiocytosis X (eosinophilic granuloma; Hand-Schüller- Christian disease, Letterer-Siwe disease)	Osteolytic lesions in bones and lesions in viscera (larynx, lungs, liver, and spleen). Clinical course similar to acute leukemia. Hypersplenism, pancytopenia, anemia, purpura, hemorrhage; hepatic involvement. Pulmonary-diffuse hilar infiltration: respiratory failure, cor pulmonale. Gingival inflammation and necrosis, with loss of teeth. Diabetes insipidus if sella turcica involved. May be on chemotherapy and/or radiotherapy	Correct anemia and coagulation defects. Assess cardiorespiratory status. Check electrolytes and fluid balance. May have history of steroid therapy. Laryngeal fibrosis; intubation may be difficult; have the difficult airway cart at hand. Beware of loose teeth
Broscheit J, Eichelbrodner O, Greim C, et al. Anesthetic management of a patient with histiocytosis X and pulmonary complications during Cesarean section. Euro J Anaesth. 2004;21(11):919-21 Howard JE, Dwivedi RC, Masterson L, et al. Langerhans cell sarcoma: a systematic review. Cancer Treat Rev. 2015;41 (4):320-31		

Holt-Oram syndrome (heart-hand syndrome)	Upper limb abnormalities; CHD in 80% (usually ASD) but arrhythmias may occur with normal cardiac anatomy; possibility of sudden death from arrhythmia, pulmonary embolus, coronary occlusion	Preoperatively assess cardiac status (echocardiogram and ECG). Upper limb venous system may be abnormal. Difficult venous cannulation. Potential difficult intubation, check airway; have the difficult airway cart available. May be difficult to position BP cuff
Homocystinuria	Thromboembolic phenomena due to intimal thickening; ectopia lentis, osteoporosis, kyphoscoliosis. Hypoglycemia may occur. Angiography may precipitate thrombosis, especially cerebral	Give fluids to maintain urine output. Consult with hematologist for treatment options to reduce risk of thromboembolism. Dextran 40 may reduce blood viscosity and platelet adhesiveness and increase peripheral perfusion. Pneumatic stockings may be indicated to prevent venous stasis. Infuse dextrose and monitor glucose levels. Avoid nitrous oxide (impairs conversion of homocysteine to methionine and increases level)
Hunter syndrome (mucopolysaccharidosis type II)	Similar, but less severe than Hurler syndrome (see page 561). See also mucopolysaccharidoses	Lowe S, Johnson DA, Tobias JD. Anesthetic implications of the child with homocystinuria. <i>J Clinical Anesth.</i> 1994;6(2):142-4 Koblin DD. Homocystinuria and administration of nitrous oxide. <i>J Clinical Anesth.</i> 1995;7(2):176 Asghar A, Ali FM. Anaesthetic management of a young patient with homocystinuria. <i>J Coll Physicians Surg Pak.</i> 2012;22(11):720-2 Hunter syndrome (mucopolysaccharidosis type II)
		Management as for Hurler syndrome. Difficult intubation due to large tongue. Attempts to secure airway using the LMA have not always been successful; have difficult airway cart at hand. Delayed recovery from anesthesia and postobstructive pulmonary edema has been reported
		Busoni P, Fognani G. Failure of the laryngeal mask to secure the airway in a patient with Hunter's syndrome (mucopolysaccharidosis type II). <i>Paediatr Anaesth.</i> 1999;9:153-5 Kreidstein A, Boorin MR, Crespi P, et al. Delayed awakening from general anaesthesia in a patient with Hunter syndrome. <i>Can J Anaesth.</i> 1994;41:423-6

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Name	Description	Anesthesia implications
Hurler syndrome (mucopolysaccharidoses type I H; formerly classed as type I)	Developmental delay, gargoyle facies, deafness, stiff joints, dwarfing, pectus excavatum, kyphoscoliosis. Abnormal tracheobronchial cartilages; severe coronary artery disease at early age, valvar and myocardial involvement. Hepatosplenomegaly. Most die from respiratory and cardiac failure before 10 years of age; sudden death common after 7 years of age. See mucopolysaccharidoses	Preoperatively assess cardiac status (echocardiogram and ECG). Antibiotic prophylaxis and chest physiotherapy preoperatively. If severe coronary artery disease or myocardial dysfunction, avoid tachycardia, do not give atropine. Upper airway obstruction due to profuse lymphoid tissue infiltration. Caution with neck movement; hypoplasia of the odontoid, atlantoaxial subluxation may occur. Difficult intubation, especially in older children, due to micrognathia, short neck, and limited movement of temporomandibular joint. Have advanced airway adjuncts such as GlideScope and fiberoptic scope immediately at hand. Nasal or oral airways may not relieve obstruction. An LMA may be successfully placed but does not always relieve obstruction. Propofol, sevoflurane, and cisatracurium are agents of choice. Epidural analgesia may fail (due to lymphoid deposition)
Hurler-Scheie compound syndrome (type I HS)	See Scheie syndrome	Belani KG, Kravit W, Carpenter BJ, et al. Children with mucopolysaccharidosis: perioperative care, morbidity, mortality, and new findings. <i>J Ped Surg.</i> 1993;28(3):403–8 Gurumurthy T, Shailaja S, Kishan S, et al. Management of an anticipated difficult airway in Hurler's syndrome. <i>J Anaesthet Clin Pharmacol.</i> 2014;30(4):558–61
Hutchinson-Gillford syndrome	See Progeria	
Hyalinosis, cutaneous-mucosal	See Urbach-Wiethe disease	

Hyperekplexia	See stiff baby syndrome
Hyperpyrexia/ hyperthermia, malignant I-cell disease (mucolipidoses)	Developmental delay, Hurler-type bone changes, severe joint limitation, chronic pulmonary disease; cardiac involvement, valvar insufficiency common. Atlantoaxial subluxation. Death in early childhood common but some survive 1 or 2 decades
	Preoperatively assess cardiac status (echocardiogram and ECG) and pulmonary status. Tracheal intubation and airway maintenance difficult due to limited jaw movement, stiffness of neck and rib cage. Caution with neck movement; use in-line stabilization if indicated. Have advanced airway adjuncts such as GlideScope and fiberoptic scope immediately available. No specific anesthesia recommendations.
	May be difficult to wean from ventilatory support M. Mahfouz AK, George G, Al-Bahlani SS, et al. Difficult intubation management in a child with I-cell disease. <i>Saud J Anaesth</i> . 2010;4(2):105-7
Idiopathic thrombocytopenic purpura	Autoimmune disease in which an antiplatelet factor is present, resulting in destruction of platelets in the spleen with thrombocytopenia and the potential for bleeding. May be acute or chronic; severe gastrointestinal or intracranial bleeding are rare in children; most recover in a few weeks. Chronic ITP is more likely in children over 10 year of age. Treatment with high-dose steroids and γ -globulin is effective in raising the platelet count (i.e., for a surgical procedure). Splenectomy is very rarely recommended in children
	May have history of steroid therapy. Platelet counts may be very low, but platelet transfusions are ineffective. Do not give NSAIDs. Avoid intramuscular injections (if splenectomy is performed, do not give platelets until the spleen is out). Preoperative pneumococcal vaccine may be indicated. Prophylactic antibiotics after splenectomy may be indicated.
	British Committee for Standards in Haematology General Haematology Task Force. Guidelines for the investigation and management of idiopathic thrombocytopenic purpura in adults, children and in pregnancy. <i>Br J Haematol</i> . 2003;120(4):574-96
	<i>Table continues on the following page.</i>

Name	Description	Anesthesia implications
Ivemark syndrome	See asplenia syndrome	
Jervell and Lange-Nielsen syndrome (Romano-Ward syndrome, congenital long QT syndrome)	Congenital deafness and cardiac conduction defects: arrhythmias and syncopal attacks (may be misdiagnosed as epilepsy). ECG shows large T waves, prolonged QT interval. Sudden death may occur. Serious arrhythmias (ventricular fibrillation) under anesthesia. Acquired long QT syndrome may be a result of drug therapy	Preoperatively assess cardiac status (ECG); consult with the child's cardiologist. General anesthesia may precipitate arrhythmias; pretreat with β-blockers to decrease risk. Avoid atropine, halothane, and desflurane. Propofol may improve rhythm disturbances. TIVA with propofol and remifentanil (or other opioid) may be the optimal technique. Left stellate ganglion block has been recommended in adults to decrease the QT interval. Ventricular fibrillation may respond to lidocaine and defibrillation. Watch for hypoglycemia as a complication of β-blockade
		Curry TB, Gaver R, White RD. Acquired long QT syndrome and elective anesthesia in children. <i>Paediatr Anaesth.</i> 2006;16(4):471–8.
		Saussine M, Massad I, Raczkia F, et al. Torsade de pointes during sevoflurane anaesthesia in a child with congenital long QT syndrome. <i>Paediatr Anaesth.</i> 2006;16(1):63–5.
		Kim HS, Kim JT, Kim CS, et al. Effects of sevoflurane on QT parameters in children with congenital sensorineural hearing loss. <i>Anaesthesia.</i> 2009;64(1):3–8.
		Aypar E, Karagoz AH, Ozer S, et al. The effects of sevoflurane and desflurane anaesthesia on QTc interval and cardiac rhythm in children. <i>Paediatr Anaesth.</i> 2007;17(6):563–7.
Juvenile hyaline fibromatosis	Autosomal recessive disease; multiple subcutaneous nodules, flexion contractures of large and small joints, radiolucent bone destruction (especially femur and humerus), hypertrophic gingiva. Systemic manifestations may involve pleura, lung, renal, and digestive system. Entrapment of nerves and vessels may occur. Intelligence normal	Check preoperatively for evidence of other organ involvements. Difficult intubation due to gingival hyperplasia and limited motion at neck and temporomandibular joints; have the difficult airway cart at hand. Careful positioning and padding required
		Norman B, Soni N, Madden N. Anaesthesia and juvenile hyaline fibromatosis. <i>Br J Anaesth.</i> 1996;76(1):163–6.
		Seefelder C, Ko JH, Padwa B. Fibrooptic intubation for massive gingival hyperplasia in juvenile hyaline fibromatosis. <i>Paediatr Anaesth.</i> 2000;10(6):682–4.

Jeune syndrome (asphyxiating thoracic dystrophy)	Severe thoracic malformation leading to neonatal asphyxia. A high rate of cervical spine stenosis and instability is present. Associated pulmonary hypoplasia. Milder forms may present in older children. Cystic renal changes, progressing to renal failure	Extreme care with intubation; in-line stabilization may be indicated. Have advanced airway adjuncts such as GlideScope and fiberoptic scope immediately available. Avoid high-pressure ventilation (hypoplastic lungs). Surgery to enlarge thorax may necessitate prolonged periods of assisted ventilation. Care with drugs excreted by kidneys
Joubert syndrome (Mohr syndrome variant, familial cerebellar vermis agenesis)	Rare autosomal recessive disorder. Cerebellar vermis dysplasia or agenesis and brain stem cysts. Hypotonia, ataxia, jerky eye movements, and tongue protrusion. Developmental delay. Abnormal respiration: alternating tachypnea and apneic spells. May be lethal in early childhood	Life-threatening respiratory problems perioperatively. Very sensitive to anesthetic agents and opioids. Inhalational induction, controlled ventilation, and local or regional analgesia advised. Apnea monitoring postoperatively; caffeine may be useful
Kabuki syndrome	Developmental delay and craniofacial anomalies; 50% have CHD and 25% have renal disease. Muscular hypotonia may be present but muscle biopsies are normal. Scoliosis develops in many	Preoperatively assess cardiac status (echocardiogram). May have difficult airway but no other specific anesthesia problems reported; have the difficult airway cart at hand
	Johnson G, Mayhew JE. Anesthesia for a child with Kabuki syndrome. <i>Paediatr Anaesth</i> . 2007;17(9):900–1 Dentici ML, Di Pede A, Lepri FR, et al. Kabuki syndrome: clinical and molecular diagnosis in the first year of life. <i>Arch Dis Child</i> . 2015;100(2):158–64	<i>Table continues on the following page.</i>

Name	Description	Anesthesia implications
Kartagener syndrome	Dextrocardia, situs inversus. Immotile abnormal cilia, deficient mucociliary clearance; sinusitis, bronchiectasis. Often undergo endoscopic sinus surgery. Defective immunity	Order respiratory physiotherapy preoperatively. Use careful aseptic technique (reverse isolation). Assess respiratory status. Affected lung lobes may need to be isolated for lobectomy
Kasabach-Merritt syndrome	Hemangioma that suddenly increases in size; thrombocytopenia, hypofibrinogenemia → purpura, bleeding, anemia, increased fibrinolytic activity. Treated by radiotherapy (surgery may precipitate disseminated intravascular coagulation). Treated with embolization, steroids, chemotherapy, and everolimus and sirolimus.	Sahajananda H, Sanjay OP, Thomas J, et al. General anaesthesia for lobectomy in an 8-year-old child with Kartagener's syndrome—anaesthetic considerations for ENT surgery. <i>Otolaryngol Pol.</i> 2012;66(4):291–4 Burdzik PK, Wawrzyniak K, Kazmierczak W, et al. Kartagener's syndrome—anaesthetic considerations for ENT surgery. <i>Paediatr Anaesth.</i> 2003;13(8):714–7 Recovery follows destruction of tumor

Kawahara M, Takeshita T, Akita S. Anesthetic management of a patient with Kasabach-Merritt syndrome. *Anesth Prog.* 1987;34(1):17–19
Kumar S, Taneja B, Saxena KN, et al. Anaesthetic management of a neonate with Kasabach-Merritt syndrome. *Indian J Anaesth.* 2013;57(3): 292–4

Kawasaki disease (mucocutaneous lymph node syndrome)	Acute febrile exanthematous disease secondary to vasculitis with cardiac involvement (pancarditis, valvular dysfunction, arrhythmias, and coronary artery vasculitis with aneurysms). Seen in infants and young children, endemic in Japan. Signs include fever, conjunctivitis, oral erythema, strawberry tongue, and red hands and feet. Cardiac involvement in 20% of cases: ranges from asymptomatic ECG changes to severe congestive failure and massive myocardial infarction. Salicylates are used in treatment and may reduce coronary lesions. Biliary tract or bowel symptoms may require laparotomy. Hepatic involvement in 10% of patients. CNS vasculitis with symptoms ranging from headache to seizures and death reported	Administer anti-seizure medications on the morning of surgery. Preoperatively assess cardiac status (echocardiogram and ECG); consult with cardiologist. Avoid myocardial depressants and anesthetic as for a patient with coronary artery disease. Monitor for cardiac ischemic changes (V_5 and lead II). Be prepared with vasoactive and antiarrhythmic drugs. Regional peripheral nerve blocks may improve ischemic limb circulation. Sevoflurane anesthesia has been used satisfactorily. Salicylates may increase surgical bleeding
Kenny-Caffey syndrome	Normal intellect. Dwarfism, macrocephaly, thoracic skeletal abnormalities, anemia, hypocalcemia, recurrent aspiration and can have mandibular hypoplasia	Intubation may be difficult; have the difficult airway cart at hand. The use of LMA is an option for securing the airway. Monitor ionized calcium levels perioperatively

Morrison JE, Anderson M, Chan KC et al. A 15-year review of children with Kawasaki's syndrome having general anesthesia or deep sedation. *Paediatr Anaesth.* 2005;15(12):1033-8
 Duzova A, Bakalkoglu A. Central nervous system involvement in pediatric rheumatic diseases: current concepts in treatment. *Curr Pharm Des.* 2008;14(13):1295-301

Janke EL, Fletcher JE, Lewis IH. Anaesthetic management of the Kenny-Caffey syndrome using the laryngeal mask. *Paediatr Anaesth.* 1996;6(3):235-8

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Name	Description	Anesthesia implications
Ketouria, branched-chain	See maple syrup urine disease	
Klinefelter syndrome (gonosomal aneuploidy with tubular dysgenesis)	Confined to males. Sex chromosome defect (47 XY, XXXY). May have ambiguous genitalia at birth. Tall, reduced intelligence, behavior problems, hypogonadism, vertebral collapse due to osteoporosis. May have diabetes mellitus. Testosterone replacement therapy is initiated at puberty	No anesthesia problem reported, except as related to diabetes. Position very carefully to prevent spinal cord damage (osteoporosis)
Watendorff DJ, Menke M. Klinefelter syndrome. Am Fam Phys. 2005;72(11):259-62		
Klippel-Feil syndrome	Congenital fusion of two or more cervical vertebrae, causing neck rigidity. Occipital encephalomyelocele may be associated in the neonate. Arnold-Chiari malformation and/or scoliosis may be associated. Rib defects, cardiac and renal disease occasionally related	Preoperatively assess cardiac (echocardiogram and renal status). Intubation may be very difficult because of immobile neck: should be carried out "awake/sedated" if possible; otherwise inhalation induction without muscle relaxant. Have advanced airway devices immediately at hand (GlideScope, fiberoptic laryngoscope, LMS, etc) Do not extubate until fully awake
Stallmer ML, Vanaharam V, Mashhour GA. Congenital cervical spine fusion and airway management: a case series of Klippel-Feil syndrome. J Clin Anesth. 2008;20:447-51.		
Klippel-Trenaunay-Weber syndrome (angio-osteohypertrophy)	Hemangiomas with hypertrophy of adjacent bone; thrombocytopenia. AV fistulas and anemia lead to high cardiac output, with possible cardiac failure; thrombocytopenia in association with visceral hemangiomas. Macrocephaly, scoliosis, and pectus excavatum. Severe bleeding may occur from hemangiomas	Assess cardiac status (echocardiogram); correct coagulopathy. Be prepared for major transfusion. Assess airway, intubation usually not a problem; have the difficult airway cart available
Barbara DW, Wilson JL. Anesthesia for surgery related to Klippel-Trenaunay syndrome: a review of 136 anesthetics. Anesth Analg. 2011;113:98-102		
Krabbe disease (globoid cell leukodystrophy)	See leukodystrophy	

Lamellar ichthyosis	Disorder of keratinization with widespread severe scaling; severe dental disease likely	Limited mouth opening, difficult airway; have the difficult airway cart at hand. An LMA will be most helpful. Prone to thermal instability; use forced air heater. Difficulty with IV access and fixation
Larsen syndrome	Multiple congenital dislocations: knees, elbows, hips. Characteristic facies, hydrocephalus, cleft palate, flat face, upturned nose. Connective tissue defect of cartilage of ribs, epiglottis, arytenoids, and tracheomalacia. Cervical spine abnormal and unstable, kyphoscoliosis, chronic respiratory problems, and CHD	Preoperatively assess cardiac (echocardiogram) and respiratory status. Intubation may be difficult and subglottic stenosis may be present. Have advanced airway devices immediately available (Glidescope, fiberoptic laryngoscope, LMS, etc.). Caution with neck; cervical spine instability; use in-line stabilization during laryngoscopy. Possible increased ICP
Laurence-Moon-Biedl syndrome	Developmental delay, pigmentary retinopathy, hypogenitalism, and spastic paraplegia. (Polydactyly and obesity, typical in Bardet-Biedl syndrome, are absent.) May have renal abnormalities and CHD	Preoperatively assess cardiac (echocardiogram) and renal status
	Banman ML, Hogan GR. Laurence-Moon-Biedl syndrome. Report of two unrelated children less than 3 years of age. <i>Am J Dis Child.</i> 1973;126:119-26.	<i>Table continues on the following page.</i>

Name	Description	Anesthesia implications
Leigh disease (subacute necrotizing encephalomyopathy)	A genetic neurologic and metabolic disease considered to be a mitochondrial myopathy may occur in infancy or childhood. Infants develop hypotonia, somnolence, optic atrophy, deafness, and pyramidal tract signs. Altered respiratory patterns may occur and may lead to sudden infant death syndrome. Impaired intracellular metabolism secondary to mitochondrial involvement. Older children have acute neurologic deterioration and respiratory failure. General anesthesia may be followed by respiratory failure and death	Preoperatively, assess pulmonary status and treat acute infections. Ensure adequate hydration, give dextrose infusion, and monitor glucose levels. Use normal saline and avoid lactated Ringer's solution. Treat acidosis. Monitor ventilation in the perioperative period. Propofol and remifentanil may be useful for many procedures
LEOPARD syndrome	A cardio-cutaneous syndrome; multiple large freckles; hypertelorism, eyelid ptosis, deafness. CHD, progressive hypertrophic cardiomyopathy (pulmonary stenosis in 95%); ECG anomalies include aberrant conduction; serious arrhythmias may occur. Growth retardation common; pectus carinatum, kyphosis, etc., in some. Genitourinary anomalies (hypospadias, cryptorchidism, ovarian hypoplasia, etc.)	Preoperatively assess cardiac (echocardiogram and ECG) and respiratory status. Avoid tachycardia and myocardial depressant drugs. Be prepared to support cardiac function and treat arrhythmias; have vasoactive drugs and antiarrhythmic drugs on hand. Intubation may be difficult; have the difficult airway cart on hand

Terkawi AS, Wahab TM, Al-Shuaib KM, et al. Anesthetic considerations in Leigh disease: Case report and literature review. *Saudi J Anesth.* 2012;6:181-5
 Gozal D, Goldin E, Shafran-Tikva S, et al. Leigh syndrome: anesthetic management in complicated endoscopic procedures. *Pediatr Anesth.* 2006;16(1):38-42

Yehoh TY, Wittner ED, Weingarten TN, et al. Anesthesia and LEOPARD syndrome: a review of forty-nine anesthetic exposures. *J Cardiothorac Vasc Anesth.* 2014;28:1243-50

Torres J, Russo P, Tobias JD. Anaesthetic implications of LEOPARD syndrome. *Pediatr Anesth.* 2004;14(4):352-6

Leprechaunism (Donohue syndrome)	A severe insulin-resistance disease due to a mutant insulin receptor gene with associated endocrine disorders such as hypothyroidism. Elfin face with mandibular hypoplasia. Failure to thrive and severe developmental delay. Hypoglycemia due to hyperinsulinism from hyperplastic islets of Langerhans; renal tubular defects → impaired renal function with nephrocalcinosis due to hypercalcemia. Lung may be dysmorphic. Most die before 1 year of age [Epib ahead of print]	Assess for multiple organ disease. Administer glucose-containing solutions and monitor blood glucose. Intubation may be difficult; have the difficult airway cart available. Use drugs excreted by kidneys with caution
Lesch-Nyhan syndrome	Disorder of purine metabolism, occurs in males. Developmental delay and growth retardation, malnutrition, choreoathetosis secondary to reduced basal ganglia volume. Very aggressive with compulsive self-destructive behavior. Hyperuricemia leads to renal calculi, RBC damage, hypertension, gouty arthritis, and coronary artery disease. Associated tracheal diverticulum has been reported. Renal failure by age of 20 years	Use drugs excreted by the kidney with caution. Beware of regurgitation and give metoclopramide. Benzodiazepines for behavior management. Midazolam, propofol, thiopental, isoflurane, and cisatracurium (Hoffman degradation independent of renal dysfunction) are recommended. Caution with catecholamines

Kallo A, Lakatos I, Szijarto L. Leprechaunism (Donohue's syndrome). *J Pediatr.* 1965;66:372-9
 Simpkin A, Cochran E, Cameron F, et al. Insulin receptor and the kidney: nephrocalcinosis in patients with recessive INSR mutations. *Nephron Physiol.* 2014;10:24

Larson LO, Wilkins RG. Anesthesia and the Lesch-Nyhan syndrome. *Anesthesiology.* 1985;63(2):197-9
 Salhotra R, Sharma C, Tyagi A, et al. An unanticipated difficult airway in Lesch-Nyhan syndrome. *J Anaesthesiol Clin Pharmacol.* 2012;28(2):239-41

Table continues on the following page.

Name	Description	Anesthesia implications
Letterer-Siwe disease	See histiocytosis X	
Leukodystrophy (Alexander disease, Canavan disease, Krabbe disease, Pelizaeus-Merzbacher disease, adrenoleukodystrophy, metachromatic leukodystrophy)	Inherited lysosomal storage disorder of myelin formation due to a deficiency of arylsulfatase A, progressive degenerative disease with spasticity, gait disturbance, poor motor development, seizures, extrapyramidal movements, and choreoathetosis. Disordered swallowing and gastroesophageal reflux lead to aspiration pneumonia. Malnutrition and anemia. Acid-base abnormalities may occur	Administer anti-seizure medications on the morning of surgery. Preoperatively assess pulmonary status and anticonvulsant medications. If oral secretions are copious, use an antisialagogue. Danger of pulmonary aspiration; suction the stomach and give antiemetics. Position and pad carefully. Avoid succinylcholine (theoretical risk of hyperkalemia). Seizure medications may result in altered responses to induction agents and muscle relaxants. Give glucose-containing solutions and avoid lactated Ringer's solution. Maintain body temperature. Extubate awake, monitor closely postoperatively. (N.B. Adrenal dysfunction in adrenoleukodystrophy; give steroids. Lumbar epidural analgesia may be appropriate for postoperative pain.) Propofol infusions are acceptable for MRI
Lipodystrophy with diabetes (Seip syndrome, Berardinelli-Seip syndrome)	Generalized loss of all body fat, fibrotic liver leading to failure, portal hypertension; splenomegaly, nephropathy, diabetes. May have renal failure. Hypersplenism may lead to anemia and thrombocytopenia. May develop dilated cardiomyopathy	Preoperatively assess cardiac (echocardiogram) renal, and coagulation status. Considerations for diabetes; monitor blood glucose. Caution with drugs metabolized by liver and those excreted by the kidneys

Hernandez-Palazon J. Anaesthetic management in children with metachromatic leukodystrophy. *Pediatr Anaesth.* 2003;13(8):733-4

Mattioli C, Gemma M, Baldoli C, et al. Sedation for children with metachromatic leukodystrophy undergoing MRI. *Pediatr Anesth.* 2007;17:64-9

Lorioli L, Cicalese MB, Silvani P, et al. Abnormalities of acid-base balance and predisposition to metabolic acidosis in Metachromatic Leukodystrophy patients. *Mol Genet Metab.* 2015;115(1):48-52

Bennett T, Allford M. Delayed emergence from anaesthesia in a child with congenital generalized lipodystrophy (Berardinelli-Seip syndrome). *Pediatr Anesth.* 2012;22:299-300

Khalife WI, Mourad MC, Khalil J. Dilated cardiomyopathy and myocardial infarction secondary to congenital generalized lipodystrophy. *Tex Heart Inst.* 2008;35(2):196-9

Lipogranulomatosis	See Farber disease	
Loeys-Dietz syndrome (similar to Marfan's)	This is an inherited defect in transforming growth factor-beta receptors resulting in dilated aortic root, hypertension, tortuous vessels, joint laxity, easy bruising. Cervical spine instability and focal kyphosis is common	Assess for cervical spine subluxation preoperatively. Difficulty with intubation and unstable cervical spine; have difficult airway cart at hand and use in-line stabilization. Avoid tachycardia and hypertension. Concurrent antihypertensives may complicate anesthesia unless withheld. Drugs to manage hyper/hypotension should be at hand. Consider regional blocks when appropriate
	Bunting AC, Bould MD. Hemodynamic instability during anesthesia in an adolescent with Loeys-Dietz syndrome: a case report. <i>Pediatr Anesth</i> . 2014;24:1302-4.	Fuhrhop SK, McElroy MJ, Dietz HC, III et al. High prevalence of cervical deformity and instability requires surveillance in Loeys-Dietz syndrome. <i>J Bone Joint Surg Am</i> . 2015;97(5):411-9.
Long QT syndrome	See Jervell and Lange-Nielsen syndrome (Romano-Ward syndrome)	
Lowe syndrome (oculocerebrorenal syndrome)	Affects males. Cataract, glaucoma, developmental delay; hypotonia, renal tubular acidosis, proteinuria, sodium and potassium wasting, osteoporosis, and rickets. Glomerulosclerosis leads to renal failure	Check electrolyte and acid-base balance and correct acidosis and hypokalemia and low serum Ca^{++} (treated with vitamin D and Ca^{++}). Use reduced doses of non-depolarizing muscle relaxants; use a blockade monitor. Caution with opioids. Avoid hyperventilation or excess glucose infusion (decreases serum K^+). Caution with drugs excreted by kidneys
	Saricaoglu F, Demirtas E, Aypar U. Preoperative and perioperative management of a patient with Lowe syndrome diagnosed to have Farconi's syndrome. <i>Pediatr Anaesth</i> . 2004;14(6):530-2.	<i>Table continues on the following page.</i>

Name	Description	Anesthesia implications
Lupus erythematosus disseminatus	See collagen diseases	
Maffucci syndrome	Enchondromatosis and hemangiomas with malignant change. Pathologic fractures, gastrointestinal bleeding from hemangiomas, orthostatic hypotension	Transport and position carefully, protect hemangioma. May show orthostatic hypotension and be sensitive to vasodilator drugs. Careful positioning to avoid fractures. Caution with intubation (airway hemangioma)
	Chan SK, Ng SK, Cho AM, et al. Anaesthetic implications of Maffucci's syndrome. <i>Anaesth Intens Care</i> . 1998;26(5):586-9.	
Malignant hyperpyrexia/ hyperthermia	See page Chap. 6	
Mandibulofacial dysostosis	See Treacher Collins syndrome	
Mannosidosis type I (severe), type II (milder)	Primary metabolic deficiency of α -mannosidases A and B → lysosomal accumulation of mannose-rich substrates. Abnormal neutrophil immunologic function. Hepatosplenomegaly, severe recurrent infections, and early death. Hearing loss, developmental delay, Hurler-like skeletal changes, gargoyle-like facies, clumsy motor function, weak connective tissues	Be alert for hepatic dysfunction and for hypoventilation perioperatively and postoperatively. Check airway, but intubation is usually without difficulty, have difficult airway cart available
	Hallas P, Borgwardt LG, Roed J, et al. Anesthesia for patients with alpha-mannosidosis—a case series of 10 patients. <i>Pediatr Anesth</i> . 2011;21:1269-70	
Maple syrup urine disease (MSUD; branched-chain ketonuria)	Inability to metabolize leucine, isoleucine, and valine and accumulation of branched-chain amino acids and keto acids lead to severe neurologic damage and respiratory disturbances. Episodes of hypoglycemia. Treated by diet from birth. Acute, life-threatening episodes with stress due to infection or surgery may lead to ketoacidosis and may require peritoneal dialysis or exchange transfusion	Consult the child's endocrinologist. Check acid-base balance, plasma amino acids preoperatively. Check serum glucose before, during, and after anesthesia. Start glucose infusion (at least 10–15 mg/kg/min) preoperatively and continue until oral intake is reestablished. Prevent overhydration. TIVA has been recommended
	Kahraman S, Erkan M, Alkukus O, et al. Anaesthetic management in maple syrup urine disease. <i>Anaesthesia</i> . 1996;51(6):575-8	

Marfan syndrome (arachnodactyly)	Tall, thin predominantly male patients with long fingers, long face, and high arched palate. Mutant gene at chromosome 15 for fibrillin causes connective tissue disorder leading to joint instability and dislocation (including cervical spine), dislocation of lens, kyphoscoliosis, hernia, pectus excavatum, lung cysts. High incidence (~4%) of spontaneous pneumothorax. Aortic root dilation may lead to aortic incompetence or aneurysm; pulmonary artery or mitral valve may be diseased	Preoperatively assess cardiac status (echocardiogram). Intubation may be difficult and the cervical spine unstable at the atlantoaxial joint. Laryngoscopy should be gentle to prevent cervical spine or temporomandibular joint damage. The use of advance airway devices and in-line stabilization is recommended. Tracheomalacia leading to difficult ventilation has been described. Position carefully to prevent dislocations. Avoid myocardial depressants, but do not allow the patient to become hypertensive (danger of aortic dissection). Beware of pneumothorax with controlled ventilation
Kane MG, Pyeritz RE. Medical management of Marfan syndrome. <i>Circ.</i> 2008;117(21):2802–13 Oh AY, Kim YH, Kim BK, et al. Unexpected tracheomalacia in Marfan syndrome during general anesthesia for correction of scoliosis. <i>Anesth Analg</i> 2002;95(2):331–2 Hebert ID, Auld BC, Sasaki T, et al. Infant repair of massive aortic aneurysm with prosthetic valved conduit. <i>Ann Thorac Surg</i> 2013;96(3):1070–2		
Maroteaux-Lamy syndrome (mucopolysaccharidosis type VI)	Normal intellect. Kyphoscoliosis with poor lung reserve; chronic respiratory infections; hypersplenism, anaemia, thrombocytopenia. Myocardial involvement with heart valve thickening, cardiomyopathy, fibroelastosis and conduction defects; heart failure by 20 years of age	See mucopolysaccharidoses. Assess cardiac (echocardiogram and ECG) and pulmonary status (chest X-ray). Assess hemoglobin and platelet counts. Care with cardiac depressant drugs. Spinal cord compression may occur. May have challenging intubation; have difficult airway cart available. May require ventilation postoperatively
Suh SH, Okutani R, Nakasui M, et al. Anesthesia in a patient with mucopolysaccharidosis type VI (Maroteaux-Lamy syndrome). <i>J Anesth</i> . 2010;24(6):945–8 Walker RW, Colovic V, Robinson DN, et al. Postobstructive pulmonary oedema during anaesthesia in children with mucopolysaccharidoses. <i>Pediatr Anesth</i> . 2003;13(5):441–7 Golda A, Jurecka A, Opoka-Winiarska V, et al. A Mucopolysaccharidosis type VI: a cardiologist's guide to diagnosis and treatment. <i>Int J Cardiol</i> . 2013;167(1):1–10		<i>Table continues on the following page.</i>

Name	Description	Anesthesia implications
Marshall-Smith syndrome	Skeletal dysplasia and dysmorphic facial features including micrognathia. Hypotonia and failure to thrive. Possible atlantoaxial instability. Respiratory tract anomalies lead to complications. Prone to nontraumatic fractures	Flexion/extension lateral neck films to rule out atlantoaxial instability. Airway problems and difficult intubation; have the difficult airway cart available. May require in-line stabilization during laryngoscopy. Association with laryngomalacia and tracheomalacia described as a cause of failure to ventilate. May require oropharyngeal or nasopharyngeal airway during induction and recovery. Cautious use of muscle relaxants if hypotonia present. Caution with positioning due to fragile bones
Mastocytosis syndrome (urticaria pigmentosa)	Abnormal aggregates of histamine- and heparin-containing mast cells; skin lesion is a brownish-red maculopapular rash mainly on trunk. Mast cell degranulation with systemic histamine and heparin release may occur with trauma, temperature changes, alcohol, and drugs (including salicylates, morphine, papaverine, polymyxin, vancomycin and atropine). Often with a history of gastroesophageal reflux. Anesthesia for minor surgical procedures have led to generalized anaphylaxis and death, but most are uneventful	Dernedde G, Pendeville P, Veyckemans F, et al. Anaesthetic management of a child with Marshall-Smith syndrome. <i>Paediatr Anaesth.</i> 1998;8(5):429–32 Artilla H, Laitio T, Aantaa R, et al. Difficult airway in a patient with Marshall-Smith syndrome. <i>Paediatr Anaesth.</i> 1998;8(5):429–32 Avoid stimuli and drugs known to cause mast cell degranulation. Prophylactic treatment with antihistamines (i.e., diphenhydramine, ranitidine, and montelukast (Singulair)) and steroids has been recommended. Inhalation anesthetics may be safely used. Propofol, rocuronium, succinylcholine, fentanyl, remifentanil, and/or meperidine (Demerol) may be used safely. Avoid amphotericin B, anticholinergics, dextran, dextromethorphan, vancomycin, ketorolac, atracurium, succinylcholine, benzocaine, chloroprocaine, tetracaine, procaine; caution with NSAIDs. Bleeding secondary to heparin release may require protamine therapy

Carter MC, Uzzaman A, Scott LM, et al. Pediatric mastocytosis: routine anesthetic management for a complex disease. *Anesth Analg.* 2008;107(2):422–7
Klein NJ, Missedine S. Anesthetic considerations in pediatric mastocytosis: a review. *J Anesth.* 2013;27:588–98

McArdle myopathy (glycogen storage disease type V)	Muscle phosphorylase deficiency; serum lactate not increased by exercise. Initially, increased fatigability; progresses to muscle cramps and weakness (all skeletal muscles affected), myoglobinuria may lead to renal failure. Myocardium may be involved; ECG abnormalities have been reported. Patients may test positive for MH with the in vitro contracture test but no reports of clinical MH have been reported	Preoperatively assess cardiac status (echocardiogram and ECG). Do not use turniquets; maintain infusion of dextrose during surgery; do not use succinylcholine. IV fluids to maintain urine output. Prevent shivering. Care with cardiac depressant drugs; monitor ECG
Meckel syndrome (dysencephalia splanchnocystica)	Occipital encephalocele, microcephaly, micrognathia, and cleft epiglottis, CHD, bilateral cystic renal dysplasia, hepatic ductal proliferation and fibrosis, and polydactyly. Most die in infancy	Preoperatively assess cardiac status (echocardiogram). Airway and intubation may be difficult; have the difficult airway cart at hand. Care with drugs excreted by liver or kidneys
Medium chain acyl-CoA dehydrogenase deficiency (MCAD)	Salonen R, Paavola P. Meckel syndrome. J Med Genet. 1998;35(6):497-501 A disorder of fatty acid metabolism secondary to deficiency of mitochondrial enzyme. Hypoglycemia, seizures, or coma may result. Tachyarrhythmias have been reported treated with carnitine	Administer anti-seizure medications on the morning of surgery. Assess cardiac rhythm with ECG preoperatively. Avoid prolonged fasting, give IV dextrose infusion, and check blood glucose perioperatively. Avoid propofol due to its high fat content. Avoid lactated Ringer's solution and use normal saline

Justiz AC, Mayhew JF. Anesthesia in a child with medium-chain acyl-CoA dehydrogenase deficiency. Paediatr Anaesth. 2006;16(12):1293-4

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Name	Description	Anesthesia implications
Median cleft face syndrome	Various degrees of cleft face; lipomas, and dermoids over frontal bone. Other intracerebral deformities are often present. Choanal atresia may be present	Assess for associated defects. Cleft nose, lip and palate may cause intubation difficulties; have difficult airway cart at hand. Caution with nasal instrumentation; nasal encephaloceles may be present
Bonneburg T, Lenz W, Eusterbrock T. Median cleft face syndrome in association with hydrocephalus, agenesis of the corpus callosum, holoprosencephaly and choanal atresia. <i>Euro J Ped.</i> 1987;146(3):301–2		
Menkes syndrome (kinky hair disease)	X-linked disorder of copper metabolism. Onset in first months of life; retarded growth and developmental delay, seizures, progressive cerebral degeneration. Gastroesophageal reflux commonly leads to aspiration pneumonia. Death from seizures or pneumonia in a few years	Administer anti-seizure medications on the morning of surgery. Risk of acid aspiration; suction the stomach. Prone to hypothermia. Avoid succinylcholine (neurologic disease). Anticonvulsants may interact with non-depolarizing muscle relaxants. Postoperative ventilation may be required
Passariello M, Altmendorfer N, Pietropaoli P. Anesthesia for a child with Menkes disease. <i>Pediatr Anesth.</i> 2008;18:1225–6		
Methylmalonyl-coenzyme A mutase deficiency	Autosomal recessive defect of protein metabolism. Protein metabolism leads to high plasma methylmalonic acid levels, producing lethargy, vomiting, dehydration, acidosis, ketonemia, and hyperammonemia. Treated by limiting protein intake, plus supplemental bicarbonate and cobalamin. Anesthesia and surgery may increase protein metabolism and lead to acidemia	Avoid excessive fasting or accumulation of blood in gastrointestinal tract. Maintain intravascular volume. Monitor blood gases, electrolytes, and ammonia level. Avoid nitrous oxide. (May exacerbate metabolic defect)
Sharar SR, Haberkern CM, Jack R, et al. Anesthetic management of a child with methylmalonyl-coenzyme A mutase deficiency. <i>Anesth Analg.</i> 1991;73(4):499–501		
Moebius syndrome (congenital oculofacial paralysis)	Congenital paralyses of sixth and seventh cranial nerves results in inability to smile. Limb deformities, micrognathia. Feeding difficulties and aspiration may cause chronic pulmonary problems. Not associated with MH although one case suggested this	Preoperatively assess respiratory status. Intubation may be difficult (but not usually); have the difficult airway cart available. May be sensitive to opioids, central apnea may occur. Monitor ventilation postoperatively
Gondipalli P, Tobias JD. Anesthetic implications of Moebius Syndrome. <i>J Clin Anesth.</i> 2006;18:55–9		
Fernandes CR, Pinto Filho WA, Cezar LC, et al. Fatal recrudescence of malignant hyperthermia in an infant with Moebius syndrome. <i>Braz J Anesthesiol.</i> 2013;63(3):296–300		

Morquio syndrome (mucopolysaccharidosis type IV)	Normal intellect. Severe dwarfing; aortic incompetence; kyphoscoliosis with poor lung function (cardiorespiratory symptoms and pulmonary hypertension by second decade). Unstable atlantoaxial joint leading to spinal cord compression; deafness. Inguinal hernia common. See mucopolysaccharidoses	Preoperatively assess cardiac (echocardiogram and ECG) and pulmonary status (X-ray). Care with cardiac depressant drugs. Assess atlantoaxial stability preoperatively and use in-line stabilization during laryngoscopy if indicated. Have advanced airway devices immediately available (GlideScope, fiberoptic laryngoscope, LMA, etc.). Care with positioning and avoid excessive neck manipulation. Regional analgesia may be appropriate for some children
Morgan KA, Rehman MA, Schwartz RE. Morquio's syndrome and its anaesthetic considerations. <i>Pediatr Anaesth</i> . 2002;12(7):641–4.		
Theroux MC, Nerkir T, Ditrano C, et al. Anesthetic care and perioperative complications of children with Morquio syndrome. <i>Pediatr Anesth</i> . 2012;22(9):901–7	Same as type IV: preparation and anesthetic	
Mucopolysaccharidosis type VII (β -glucuronidase deficiency)	Severe developmental delay. Skeletal anomalies similar to type IV	
Moschcowitz disease (thrombotic thrombocytopenic purpura)	Hemolytic anemia and thrombocytopenia, arteriolar and capillary disease, neurologic damage, renal disease. Treatment: plasmapheresis and steroids; splenectomy for resistant cases. Assess for history of steroid therapy	Check platelet count and hemoglobin. Assess BUN/creatinine. Possible steroid supplement. Avoid IM injections. Smooth induction and intubation (prevent hypertension as might cause CNS bleed). Nasotracheal intubation contraindicated (due to risk of bleeding). Care with drugs excreted by kidneys. Platelet transfusions should be avoided (may exacerbate disease); transfuse with PRBCs and FFP
Pivalizza EG. Anesthetic management of a patient with thrombotic thrombocytopenic purpura. <i>Anesth Analg</i> . 1994;79(6):1203–5		

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Name	Description	Anesthesia implications
Moyamoya disease	Severe carotid artery stenosis with a fine network of vessels around the basal ganglia ("puff of smoke"). Initially manifests as transient ischemic attacks leading to ischemia and later paroxysmal hemiplegia. Treatment is with low-dose aspirin or calcium channel blockers and by surgical revascularization using a scalp artery to the pial surface of the brain Baykan N, Ozgen S, Ustalar ZS, et al. Moyamoya disease and anaesthesia. Paediatr Anaesth. 2005;15(12):1111-5	Hypocapnia leads to severe cerebral ischemia: prevent hyperventilation, maintain normocapnia. Isoflurane may be useful as a cerebral vasodilator, propofol may provide cerebral protection. Prevent hypothermia. Maintain hydration (1.5× maintenance) and maintain cerebral perfusion pressure. Scalp blocks may be useful during neurosurgery
Mucopolysaccharidoses	Type I H, I HS, II, VII Type III Type I S, IV, VI Affects bones and intellect Affects intellect only Affects bones only See previous classifications I H: Hurler syndrome I S: Scheie syndrome; formerly classified as type V HS: Hurler-Scheie compound (see Scheie syndrome) II: Hunter syndrome III: Sanfilippo syndrome IV: Morquio syndrome V: Formerly Scheie syndrome VI: Maroteaux-Lamy syndrome VII: Morquio syndrome Walker RW, Darowski M, Morris P, et al. Anaesthesia and mucopolysaccharidoses. A review of airway problems in children. Anaesthesia. 1994;49(12):1078-84 Frawley G, Fuenzalida D, Donath S, et al. A retrospective audit of anaesthetic techniques and complications in children with mucopolysaccharidoses. Pediatr Anesth. 2012;22:737-44	All may be difficult to intubate. LMA may not relieve obstruction. Spinal cord compression may occur because of thickening of dura and odontoid hypoplasia; preoperative MRI of spinal cord suggested. Assess cardiac status (echocardiogram and ECG) to assess severity of cardiac dysfunction. All are subject to postobstructive pulmonary edema. (Early stem cell therapy may reduce the difficulty of airway management later)

Multiple endocrine adenomas	See Wermer syndrome See Sipple syndrome	Caution with all muscle relaxants. Succinylcholine results in very high CK levels or hyperkalemia and should be avoided
Muscle, eye, brain disease (MEB)	Muscular dystrophy, eye disease (glaucoma, strabismus, nystagmus), and developmental delay. Severe muscle weakness, secretion retention, bedridden	Karhunen U. Serum creatine kinase levels after succinylcholine in children with "muscle, eye and brain disease." <i>Can J Anaesth.</i> 1988;35:90-2 Kose EA, Bakar B, Ates G, et al. Anesthesia for a child with Walker-Warburg syndrome. <i>Brazil J Anesth.</i> 2014;64:128-30
Muscle phosphofructokinase deficiency (glycogen storage disease type VII) (Tarui disease)	Neither glycogen nor glucose can be used as metabolic fuels. May have myoglobinuria, rhabdomyolysis, muscle cramps, exercise intolerance, hemolytic anemia and hyperuricemia. Reduced RBC life span (13–16 days)	Monitor glucose levels. Infuse dextrose. No specific anesthesia complications have been reported
Myasthenia congenita	Similar to myasthenia gravis in older children. See page 364	Do not use respiratory depressants or muscle relaxants. Ventilatory support may be required postoperatively. Possibility of cholinergic crisis with anticholinesterase therapy
White MC, Stoddart PA. Anesthesia for thymectomy in children with myasthenia gravis. Paediatr Anaesth. 2004;14(8):625-35		<i>Table continues on the following page.</i>

Name	Description	Anesthesia implications
Myofibrillar myopathy	Skeletal (including respiratory), cardiac, and smooth muscle involved in progressive disease. Contractures and rigidity in affected skeletal muscles. Risk of respiratory failure, cardiomyopathy, conduction defects, and sudden death	Preoperatively assess cardiac status (echocardiogram). Airway may be difficult due to contractures; have difficult airway cart at hand. Avoid succinylcholine and caution with other relaxants. Difficulty with positioning. Risks of MH and rhabdomyolysis are unknown
Myositis ossificans (fibrodysplasia ossificans progressiva)	Bony infiltration of tendons, fascia, aponeuroses, and muscle. Thoracic involvement greatly reduces thoracic compliance: progressive respiratory failure. Risk of any further minor trauma causing progression of disease	Check respiratory function and history of steroid therapy. Airway and intubation problems if neck rigid and mouth fixed; have difficult airway cart and advanced airway devices available. Forced manipulation of the jaw may cause local progression of disease; very gentle fiberoptic intubation indicated. Avoid IM injections. Careful padding and prevent all trauma to joints and tissues. Specific recommendations at www.IFOPA.org
Myotonia congenita (Thomsen disease)	Decreased ability to relax muscles after contraction; diffuse hypertrophy of muscle (similar to myotonia dystrophica but more benign and nonprogressive). Paradoxical response to non-depolarizing muscle relaxants possible (generalized muscle spasms); avoid succinylcholine (possible hyperkalemia, masseter spasm)	Use short-intermediate-acting non-depolarizing muscle relaxants with caution; avoid succinylcholine; avoid inhalation anesthetics; TIVA technique recommended; use opioids with caution. Postoperative respiratory complications common due to poor cough. Regional nerve blocks for analgesia recommended

Latham GJ, Lopez G. Anesthetic considerations in myofibrillar myopathy. *Pediatr Anesth.* 2013;23:231–8

Tunolo M, Moscatelli A, Silvestri G. Anaesthetic management of a child with fibrodysplasia ossificans progressiva. *Br J Anaesth.* 2006;97(5):701–3
Kilmartin E, Grunwald Z, Kaplan FS, et al. General anesthesia for dental procedures in patients with fibrodysplasia ossificans progressiva: a review of 42 cases in 30 patients. *Anesth Analg.* 2014;118:298–301

Bandschapp O, Iaizzo PA. Pathophysiologic and anesthetic considerations for patients with myotonia congenital or periodic paralyses. *Pediatr Anesth.* 2013;23:824–33

Myotonia dystrophica (myotonic dystrophy, Steinert disease)	Weakness and myotonia; eyelid ptosis, cataracts, frontal baldness; cardiac conduction defects and arrhythmias, possible cardiomyopathy. Reduced pulmonary function, very sensitive to respiratory depressants. Esophageal motility disorder; dysphagia and tendency to GERD and aspiration. Endocrine abnormalities (hypothyroidism, diabetes) may be present in older patients. May present in the neonate with weakness and hypotonia	Preoperatively assess cardiac status (echocardiogram and ECG). Assess respiratory function. Do not use succinylcholine (which causes myotonia in 50%). Cautious use of inhalational agents as may cause myocardial depression. Monitor ECG continuously. Non-depolarizing relaxant drugs may produce poor relaxation and may interact with patient's medication (ie., phenytoin). Caution with antagonizing muscle relaxants; neostigmine may induce myotonia; halothane may cause shivering and myotonia postoperatively. Sugammadex is useful after rocuronium. Extremely sensitive to respiratory depressants—use regional analgesia when possible. Anticipate postoperative pulmonary complications; ventilatory support may be necessary. Caution with opioids
Nager syndrome	Micrognathia, fishlike face, cleft palate (similar to Treacher Collins), limb deformities. Tetralogy of Fallot may be associated. Cervical spine anomalies	Preoperatively assess cardiac function (echocardiogram); SBE prophylaxis if indicated. Mouth opening can be very limited and intubation very difficult. Have difficult airway cart in OR and use advanced airway adjuncts such as LMA and fiberoptic technique. Upper airway obstruction may necessitate tracheostomy in the neonate. Postoperative ventilatory obstruction may occur; Monitor postoperatively

White RJ, Bass SP. Myotonic dystrophy and paediatric anaesthesia. *Pediatr Anaesth*. 2003;13(2):94–102
Veyckemans F, Scholles JL. Myotonic dystrophies type 1 and 2: anesthetic care. *Pediatr Anesth*. 2013;23:794–803

Groper K, Johnson JO, Braddock SR, et al. Anaesthetic implications of Nager syndrome. *Paediatr Anaesth*. 2002;12(4):365–8

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Name	Description	Anesthesia implications
Nail-patella syndrome (arthro-osteonychodyplasia)	Dysplasia of nails and absent or hypoplastic patellas. Fragile teeth. May have “lilac horns” abnormality of elbows, nephropathy; increased mucopolysaccharide excretion.	Caution with intubation (fragile teeth). Prepare for vasomotor instability. Care with drugs excreted by kidneys. Position and pad carefully (abnormal muscle insertions)
Nemaline rod myopathy	Hennessey TA, Backman SB, Meterissian SH, et al. Nail-Patella syndrome: a case report and anesthetic implications. Can J Anaesth. 2007;54(10):835-9 Congenital myopathy, may be related to central core disease. Commonly present as neonates with hypotonia, weak cry, and poor feeding. Dysmorphic features, micrognathia, slender face, high arched palate. Motor development delayed, muscle weakness of trunk and limbs plus respiratory and pharyngeal; leads to respiratory failure, aspiration pneumonia. Congenital heart disease may be associated	Preoperatively assess cardiac (echocardiogram) and pulmonary status. Preoperative physiotherapy and antibiotics for infection if indicated. Intubation may be difficult; have the difficult airway cart at hand. Brisk vagal responses noted; atropine or glycopyrrrolate may be indicated. Possible central sensitivity to depressant drugs. Avoid succinylcholine (abnormal response) and inhalation agents. Postoperative ventilation may be required. Link to central core disease suggests possibility of MH but not yet reported in nemaline myopathy. Regional analgesia may be appropriate. TIVA technique may be the best alternative

- Cunliffe M, Burrows FA. Anaesthetic implications of nemaline rod myopathy. Can Anaesth Soc J. 1985;32(5):543-7
 Shenkman Z, Sheffer O, Erez J, et al. Spinal anaesthesia for gastrostomy in an infant with nemaline myopathy. Anesth Analg. 2000;91(4):858-9
 Klingler W, Rueffert H, Lehmann-Horn F, et al. Core myopathies and risk of malignant hyperthermia. Anesth Analg. 2009;109(4):1167-73

Neonatal hypoglycemia, symptomatic	Symptomatic hypoglycemia in infants: (1) small for gestational age, (2) diabetic mothers, and (3) premature infants. If untreated: convulsions, lethargy; and developmental delay; no ketosis. Rarely, insulinoma or pancreatic hypertrophy requiring subtotal pancreatectomy. See also Beckwith syndrome	Start IV glucose infusion (5–10 mg/kg/min on a pump; no bolus) preoperatively and monitor blood glucose until condition stable postoperatively. (Boluses would precipitate rebound hyperglycemia.) The child may be receiving steroids, diazoxide, and glucagon. (N.B. Normal full-term neonates may occasionally be found to have asymptomatic “hypoglycemia” <40 mg/dL)
Nevroid basal cell carcinoma syndrome	Rozance PJ. Update on neonatal hypoglycemia. <i>Curr Opin Endocrinol Diabetes Obes.</i> 2014;21:45–50 Tin W. Defining neonatal hypoglycemia: a continuing debate. <i>Semin Fetal Neonatal Med.</i> 2014;19:27–32	See Gorlin-Goltz syndrome
Niemann-Pick disease (see also Wolman disease) types A, C, D, and type B (onset in infancy)	Hepatosplenomegaly and accumulation of sphingomyelin and other lipids throughout the body. Bone marrow, liver, and spleen involvement leads to anemia and thrombocytopenia. Diffuse foam cell infiltration of lungs leads to pulmonary insufficiency, pneumonia. May have coronary or valvular heart disease. Developmental delay. Epilepsy, ataxia. Death usually by the 3rd year (type A) to 15th year (type C). Normal intellect. Pulmonary disease (foam cells in alveoli)	Preoperatively check cardiac (echocardiogram), pulmonary (chest X-ray), and coagulation studies. Anticipate difficulty with ventilation (pulmonary restrictive disease and ascites) and possible requirement for postoperatively ventilatory support. Caution with drugs metabolized in the liver. Possibility of seizures with sevoflurane. Propofol is usually OK. Monitor and maintain body temperature

Bujok LS, Bujok G, Knapik P, Niemann-Pick disease: a rare problem in anaesthesiological practice. *Paediatr Anaesth.* 2002;12(9):806–808
Miao N, Lu X, O’Grady NP. Niemann-pick disease type C: implications for sedation and anesthesia for diagnostic procedures. *J Child Neurol.* 2012;27:1541–6

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Name	Description	Anesthesia implications
Noack syndrome (variant of Pfeiffer syndrome)	See Pfeiffer syndrome	
Noonan syndrome	Short stature, web neck, hypertelorism, mild developmental delay. Similar to Turner syndrome. Cardiac anomalies: usually pulmonary stenosis, hypertrophic cardiomopathy. Micrognathia, hydronephrosis, platelet dysfunction	Preoperative assess cardiac (echocardiogram and ECG), renal (BUN creatinine) and coagulation status. Possible difficult intubation; have the difficult airway cart at hand. Care with drugs excreted by kidney's
Oculoauriculovertebral syndrome	Aggarwal V, Malik V, Kapoor PM, Kiran U. Noonan syndrome: an anesthesiologist's perspective. Ann Cardiac Anaesth. 2011;14:214–7 Colquitt JL, Noonan JA. Cardiac findings in Noonan syndrome on long-term follow-up. Congenit Heart Dis. 2014;9(2):144–50	
Oculocerebrorenal syndrome	See Goldenhar syndrome	
Oculodento-osseous dysplasia (ODOD)	Microphthalmia and microcornea, small nose with anteverted nostrils, cleft palate, dental enamel dysplasia, plus a generalized defect of bony modeling; mandibular dysplasia, thick ribs, abnormal long bones	Airway difficulties due to nasal, oral, and mandibular defects. Brittle teeth. Difficult intubation; have difficult airway cart available
Oculofacial paraparesis, congenital	Coleheavy F, Colbert S, Dunphy J. Oculodento-osseous dysplasia: a review of anaesthetic problems. Paediatr Anaesth. 1994;4:179–82	
Ollier syndrome	See Moebius syndrome	
	See also Maffucci syndrome (enchondromatosis with cavernous hemangioma). Multiple chondromas within bones, usually unilaterals; pathologic fractures. Asymmetric dwarfism. 25 % risk for skeletal, visceral or brain malignancy	Position carefully. Hemangioma considerations as for Maffucci syndrome

Opitz-Frias syndrome (G syndrome, hypopadias, dysphagia syndrome)	X-linked or autosomal dominant, affects males more than females. Craniofacial and genital abnormalities (bifid scrotum). Dysphagia and recurrent aspiration, achalasia, hiatal hernia. Hypertelorism, micrognathia, and a high arched palate. Laryngeal malformations (including laryngotracheal cleft and subglottic stenosis) and pulmonary hypoplasia	Difficult airway, small larynx (prepare small tracheal tubes); have difficult airway cart immediately available. Danger of regurgitation; empty stomach before induction
Oral-facial-digital syndrome (Mohr syndrome)	Cleft lip and palate, lobed tongue, hypoplastic mandible and maxilla, digital anomalies; hydrocephalus, polycystic kidneys. Possible tracheomalacia and laryngomalacia. Corpus callosum anomaly (may result in delayed recovery from anesthesia)	Assess respiratory status. Airway problems and difficult intubation; have difficult airway cart available. Preoperatively renal function (BUN/creatinine); caution with drugs excreted by the kidneys
Osler-Rendu-Weber syndrome (hemorrhagic telangiectasia)	Multiple capillary and venous dilation, most commonly affects the skin and nasal mucosa, but any organ may be affected. High incidence of pulmonary and hepatic AV fistula; massive hemoptysis possible	Anemia; internal hemorrhage may occur perioperatively. Blood loss difficult to control. Difficult to maintain IV due to fragile vessels. Check pulmonary status. Positive pressure ventilation may decrease oxygenation in patients with pulmonary AV malformation
		Sharma D, Pandia MP, Bithal PK. Anaesthetic management of Osler-Weber-Rendu syndrome with coexisting congenital methaemoglobinemia. <i>Acta Anaesthet Scand.</i> 2005;49(9):1391-4 Weingarten TN, Hanson JW, Anusonwu KO, et al. Management of patients with hereditary hemorrhagic telangiectasia undergoing general anesthesia: a cohort from a single academic center's experience. <i>J Anesth.</i> 2013;27:705-11

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Name	Description	Anesthesia implications
Osteogenesis imperfecta (fragilitas ossium)	1. Congenita—usually stillbirth or rapidly fatal 2. Tarda—pathologic fractures, blue sclera, deafness Osteoporosis → kyphoscoliosis → lung pathology Fragility of vessels results in subcutaneous hemorrhage. Dentine deficiency results in carious, fragile teeth	Use extreme care in positioning (to prevent breaking bones) and intubating. Teeth are easily broken. Difficulty in maintaining IV due to fragile vessels. Intraoperative hyperthermia (not MH) has been described in patients receiving inhaled anesthetics; temperature remains unchanged or may decrease during TIVA
	Karabiyik L, Capan Z. Osteogenesis imperfecta: different anaesthetic approaches to two paediatric cases. <i>Paediatr Anaesth.</i> 2004;14(6):524–5 Ogawa S, Okutani R, Suehiro K. Anesthetic management using total intravenous anesthesia with remifentanil in a child with osteogenesis imperfecta. <i>J Anesth.</i> 2009;23(1):123–5	
Osteopetrosis	See Albers-Schönberg disease	Check serum K ⁺ level. Unpredictable response to non-depolarizing muscle relaxants; avoid succinylcholine. (See also myotonic dystrophy)
Paramyotonia congenita (Eulenburg periodic paralysis)	Myotonia on exposure to cold; paroxysmal weakness; serum K ⁺ may be high or low	Ay B, Gercerk A, Dogan VI, et al. Pyloromyotomy in a patient with paramyotonia congenita. <i>Anesth Analg.</i> 2004;98(1):68–9
Patau syndrome (trisomy 13 syndrome)	Developmental delay, microcephaly, microglossia, cleft lip or palate. May have cardiac anomalies (usually ventricular septal defect and/or dextrocardia). Patients die in infancy. Possible spinal malformations	Assess cardiac status (echocardiogram and ECG); SBE prophylaxis if indicated. Possible difficult intubation; have difficult airway cart at hand. Avoid neuraxial regional analgesia if spine is abnormal.
	Pollard RC, Beasley JM. Anaesthesia for patients with trisomy 13 (Patau's syndrome). <i>Paediatr Anaesth.</i> 1996;6:151–3 Cohen IR. Caudal block complication in a patient with trisomy 13. <i>Pediatr Anesth.</i> 2006;16:213–5	

Pelizaeus-Merzbacher disease	See leukodystrophy		
Pendred syndrome	Autosomal recessive inheritance of sensorineural deafness and goiter; incomplete block of thyroxine production. May be euthyroid or hypothyroid. Increased risk for thyroid cancer. Candidates for cochlear implants	Preoperatively ensure that patient is euthyroid, otherwise as for cretinism	
	Fraser GR, Morgans ME, Trotter WR. The syndrome of sporadic goitre and congenital deafness. Queensland J Med. 1960;29:279 Choi BY, Muskett J, King KA, et al. Hereditary hearing loss with thyroid abnormalities. Adv Otorhinolaryngol. 2011;70:43-9 Richards ML. Familial syndromes associated with thyroid cancer in the era of personalized medicine. Thyroid. 2010;20(7):707-13		
Periodic paralysis	See familial periodic paralysis and paramyotonia congenita		
Pfeiffer syndrome	Cloverleaf skull (bicornal craniostenosis), proptosis, hypertelorism, small mandible, ankylosis of elbows, may have hydrocephalus. May have associated obstructive sleep apnea. Normal intelligence. High mortality rate	Anticipate difficult airway. Have advanced airway devices (GlideScope, fiberoptic scope, and difficult airway cart) available. If OSA, reduce opioids and closely monitor respirations and oxygen saturation postoperatively. Eye care for proptosis	
	Caruselli M, Giretti R, Pallotto R, et al. Intubation using a "bonfils fiberscope" in a patient with Pfeiffer syndrome. J Bronchology Inter Pulmonol. 2011;18(4):374-5 Gupta A, Ahmed M, Prabhakar C, et al. Unique airway finding in a case of Pfeiffer syndrome and its management. J Anaesthesiol Clin Pharmacol. 2011;27(3):414-5		
PHACE syndrome	Posterior fossa brain malformations, hemangiomas, arterial anomalies, coarctation of the aorta, eye defects (PHACE). May be associated with bilateral agenesis of the carotid arteries, cerebral perfusion via the vertebral arteries. Airway hemangioma may be present. Renal artery stenosis may cause hypertension	Preoperatively assess cardiac status (echocardiogram). Risk of cerebral ischemia; CVA. Review vascular anatomy. Monitor cerebral oxygenation and function. Hypertension may need therapy	
	Javault A, Metton O, Raisky O, et al. Anesthesia management in a child with PHACE syndrome and agenesis of bilateral internal carotid arteries. Paediatr Anaesth. 2007;17(10):989-93 Imada T, Okurani R, Oda Y. Anesthesia for aortic reconstruction in a child with PHACE syndrome. J Anesth. 2014;28:919-23		<i>Table continues on the following page.</i>

Name	Description	Anesthesia implications
Phenylketonuria	Phenylalanine hydroxylase deficiency. At risk for B12 deficiency. Vomiting, CNS irritability, developmental delay, hypertension, convulsions. Phenylalanine-deficient diet must be maintained; may present with megaloblastic anemia if poorly controlled	Induction and maintenance by inhalation technique. Control ventilation. Give dextrose infusion and monitor glucose levels perioperatively (tendency to hypoglycemia). Ensure adequate phenylalanine and hemoglobin concentration (no megaloblastic anemia) preoperatively. Nitrous oxide is contraindicated as it irreversibly oxidizes the cobalt in vitamin B12 and has lead to subacute spinal cord degeneration. Sensitive to opioids and other CNS depressants; monitor body temperature. If patient has epilepsy, continue anti-seizure medications

Dal D, Celiker V. Anesthetic management of a strabismus patient with phenylketonuria. *Pediatr Anaesth.* 2004;14(8):701-2
 Baum VC. When nitrous oxide is no laughing matter: nitrous oxide and pediatric anesthesia. *Pediatr Anaesth.* 2007;17(9):824-30
 Flippo TS, Holder WD, Jr. Neurologic degeneration associated with nitrous oxide anesthesia in patients with vitamin B12 deficiency. *Arch Surg.* 1993;128(12):1391-5

Pierre Robin syndrome	Cleft palate, micrognathia, glossoptosis due to first branchial arch embryologic defect. CHD may be present. Neonates: upper airway obstruction may occur and can lead to cor pulmonale. Maintain airway by nursing prone on a frame; may require tongue suture, intubation, or tracheostomy	Preoperatively assess cardiac status (echocardiogram). Micrognathia and airway improve with growth. Intubation may be extremely difficult. Difficult airway adjuncts must be immediately available, and a fiberoptic or GlideScope intubation should be anticipated; topical airway analgesia with nebulized lidocaine will facilitate awake insertion of an LMA, used to induce anesthesia and as a conduit for intubation in the neonate or infant. The child should be fully awake before extubation
Asai T, Nagata A, Shingu K. Awake tracheal intubation through the laryngeal mask in neonates with upper airway obstruction. Paediatr Anaesth. 2008;18(1):77–80		
Frawley G, Espenell A, Howe P, et al. Anesthetic implications of infants with mandibular hypoplasia treated with mandibular distraction osteogenesis. Paediatr Anaesth. 2013;23(4):342–8		
Cladis F, Kumar A, Grunwaldt L, et al. Pierre Robin Sequence: a perioperative review. Anesth Analg. 2014;119(2):400–12		
Plotz syndrome	Vocal cord paralysis, psychomotor retardation, and sixth nerve palsy. Stridor at rest, respiratory distress, and cyanotic or choking spells	Anticipate airway obstruction and potential for aspiration perioperatively
Poland syndrome	Absent or hypoplastic pectoral muscles with chest deformity. Ipsilateral syndactyly or microdactyly. May have CHD, renal and gastrointestinal anomalies. Extreme form: Moebius syndrome has facial paralysis. Lung herniation on crying; paradoxical movement of chest wall on inspiration	Preoperatively assess cardiac (echocardiogram) and renal function (BUN/creatinine). Controlled ventilation is recommended due to the chest deformity
Sethuraman R, Kannan S, Bala I, et al. Anaesthesia in Poland syndrome. Can J Anaesth. 1998;45(3):277–9		
Soccorso G, Parkkh DH, Worollo S. Customized silicone implant for the correction of acquired and congenital chest wall deformities: A valuable option with pectus excavatum. J Pediatr Surg. 2015;50(7):1232–5		

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Name	Description	Anesthesia implications
Polyarteritis nodosa	See collagen disease	
Polycystic kidneys	Associated cysts in the liver, pancreas, spleen, lungs, bladder, thyroid (one-third); cerebral aneurysm in 15%	Preoperatively, assess renal function (BUN/creatinine). Lung cysts may lead to pneumothorax; avoid high-peak inflation pressures. Prevent hypertension (possible associated cerebral aneurysm)
Polyneuritis, acute	See Guillain-Barré syndrome	
Pompe disease (glycogen storage disease type II)	Deposits of glycogen in muscles, severe hypotonicity; large tongue; hypertrophic cardiomyopathy and conduction defects. Death from cardiorespiratory failure before 2 years of age. Replacement therapy with recombinant human α -glucosidase enzyme (rhGAA) has proven effective in extending the life span	Preoperatively assess cardiac function (echocardiogram and ECG); SBE prophylaxis if indicated. Extreme care required with cardiac depressants; avoid tachycardia and maintain preload. Monitor ECG for rhythm and ST segment changes; serious arrhythmias may occur. Use muscle relaxants with caution; etomidate and ketamine are recommended agents. Large tongue may cause airway problem; have the difficult airway cart at hand. TIVA is recommended. Regional analgesia may be useful in some cases

Ing RJ, Cook DR, Bengur RA, et al. Anaesthetic management of infants with glycogen storage disease type II: a physiological approach. *Paediatr Anaesth.* 2004;14(6):514-9
 Walker RW, Briggs G, Bruce J. Regional anaesthetic techniques are an alternative to general anaesthesia for infants with Pompe's disease. *Pediatr Anesth.* 2007;17:697-702

Porphyrias	Paralysis, psychiatric disorder, autonomic imbalance—hypertension, tachycardia; abdominal pain precipitated by drugs, stress, infection, etc. High incidence of diabetes	Avoid prolonged fasting and dehydration. Avoid barbiturates (including thiopental) and certain other IV agents (i.e., etomidate, ketamine, hydantoin, derivatives, sulfonamides, antipyretics, or hypoglycemic agents). See Jensen et al. for drug concerns in porphyria. The following have been used safely: atropine, glycopyrrolate, propofol (brief exposure), succinylcholine, N ₂ O, sevoflurane, vecuronium, atracurium, cisatracurium, fentanyl, morphine, barbiturates, epinephrine, neostigmine, chloral hydrate, diphenhydramine, chlorpromazine, and bupivacaine	Jensen NF, Fideller DS, Striepe V. Anesthetic considerations in porphyrias. <i>Anesth Analg</i> . 1995;80:591-9 Sheppard L, Dorman T. Anesthesia in a child with homozygous porphobilinogen deaminase deficiency: a severe form of acute intermittent porphyria. <i>Paediatr Anaesth</i> . 2005;15(5):426-8
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Name	Description	Anesthesia implications
Prader-Labhart-Willi syndrome	Sporadic mutation. Cytogenetic deletion at chromosome inherited from father (same genetic defect in Angelman syndrome is inherited from mother). Hypothalamic type “Pickwickian syndrome.” Neonate: hypotonia, poor feeding, reflexes absent. Second phase: hyperactive, uncontrollable polyphagia, thermoregulation disturbed, developmental delay. Extreme obesity leading to cardiorespiratory failure. May have a small mouth or mandible	Danger of hypoglycemia so monitor blood glucose and infuse IV glucose solution in perioperative period. Obesity makes venous cannulation difficult. The small mandible may make intubation difficult; have the difficult airway cart at hand. Low-grade pyrexia may occur during scoliosis, strabismus, or hernia surgery. No risk of MH. Hypothermia may also occur. Sleep apnea common: assisted or controlled ventilation may be necessary during and after operation, or apnea monitoring postoperatively. Use only low-dose opioid or avoid opioids with regional blocks and postoperative NSAIDS. Beware of postoperative airway obstruction; nasal CPAP may improve airway. Regional analgesia may be appropriate in some patients for intraoperative management and/or postoperative pain

Dearlove OR, Dobson A, Super M. Anaesthesia and Prader-Willi syndrome. Paediatr Anaesth. 1998;8(3):267-71
 Lam H, Landsman IS. Are children with Prader Willi syndrome at higher risk for anaesthetic complications? Paediatr Anaesth. 2014;24(4):457-9
 Mantadakis E, Spanaki AM, Geromarkaki E, et al. Near demise of a child with Prader-Willi syndrome during elective orchidopexy. Paediatr Anaesth. 2006;16(7):790-3

Progeria (Hutchinson-Gillford syndrome)	Premature aging starts at 6 months to 3 years; cardiac disease-ischemia, hypertension, cardiomegaly. Diabetes may be present. Death from coronary artery disease may occur before 10 year of age. Thin skin and fragile blood vessels	Assess cardiac status preoperatively particularly evidence of coronary artery disease and myocardial ischemia. Intubation may be difficult because of small mouth and receding mandible; have difficult airway cart available. Anesthesia as for adults with coronary artery disease and myocardial ischemia; avoid tachycardia and hypertension. Administer glucose-containing solutions and check glucose values perioperatively
Capell BC, Collins FS, Nabel EG. Mechanisms of cardiovascular disease in accelerated aging syndromes. <i>Circ Res.</i> 2007;101(1):13–26 Liesmann CD. Anaesthesia in a child with Hutchinson-Gillford progeria. <i>Paediatr Anaesth.</i> 2001;11(5):611–4 Hansda U, Agarwal J, Patra C, et al. Extradural hematoma surgery in a child with Hutchinson-Gillford progeria syndrome: Perioperative concerns. <i>J Ped Neurosci.</i> 2013;8:165–7		
Proteus syndrome	A highly variable disease with progressive overgrowth of connective tissues, bone, skin lesions (nevi), and abnormal distribution of fat. Cystic lung lesions. Scoliosis is common. The neck may be elongated and twisted because of vertebral deformities. Vascular malformations may be present, and pulmonary emboli without venous thrombosis have been reported	Check preoperative chest X-ray for cystic lesion. If present, avoid N ₂ O and high-peak inflation pressures; possible pneumothorax. Caution with airway, intubation may be difficult; have difficult airway cart available. Postoperative airway obstruction may occur, monitor ventilation

Cekmen N, Kordan AZ, Tuner B, et al. Anesthesia for proteus syndrome. *Paediatr Anaesth.* 2004;14(8):689–92

Nakane M, Sato M, Hattori H, et al. Perioperative respiratory complications caused by cystic lung malformation in Proteus syndrome. *J Anesth.* 2006;20:26–9

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Name	Description	Anesthesia implications
Prune belly syndrome	Agenesis of abdominal musculature with renal anomalies. Poor cough; risk of postoperative atelectasis, respiratory infections, and respiratory failure	Preoperatively assess renal function (BUN/creatinine). Treat as for a full stomach: intubate and control ventilation. Intubation may be difficult in some; have difficult airway cart available. Use muscle relaxants and drugs excreted by kidneys with caution. Thoracic epidural useful for postoperative analgesia and may prevent respiratory compromise
Pseudohypoparathyroidism	See Albright osteodystrophy	Baris S, Karakaya D, Ustun E, et al. Complicated airway management in a child with prune-belly syndrome. Paediatr Anaesth. 2001;11:501-4
Pseudoxanthoma elasticum	See Grönblad-Strandberg syndrome	Henderson AM, Vallis CJ, Sumner E. Anaesthesia in the prune-belly syndrome. A review of 36 cases. Anaesthesia. 1987;42(1):54-60
Pyle disease (metaphyseal dysplasia)	Craniofacial abnormalities; enlarged mandible; cranial nerve paryses	Assess airway; possible difficult intubation; have difficult airway cart at hand

Rett syndrome	Disabling neurologic disorder affecting only females. Underweight, developmental delay, autism, seizures, scoliosis, abnormal pain sensation, vaso motor instability, cardiac arrhythmias (long QT syndrome), marked irregular respiration: hyperventilation alternating with apneic spells	Administer anti-seizure medications on the morning of surgery. Preoperative ECG. Assess pulmonary function. Severe risk of respiratory complications. Often present for spinal surgery for scoliosis. SSEPs can be monitored but MEPs may be contraindicated if history of seizures. Postoperatively, apnea monitoring or ventilatory support needed. Insensitive or hypersensitive to pain. May be sensitive to respiratory depressant drugs. Benzodiazepines to control seizures. Cautions as for long QT syndrome; prevent tachycardia
Reye syndrome	Severe metabolic encephalopathy and fatty degeneration of viscera (especially liver): hyperaminoacidemia; increased prothrombin time, blood ammonia, serum transaminases. Suspected cofactor is ingestion of ASA (aspirin) during prodromal illness. Most reliable diagnosis is by liver biopsy. If untreated, increased ICP is usually fatal. There have been very few cases in the last 15 years	Anesthetize for investigation of and decompression of increased ICP. Patient may be receiving steroids and controlled hypothermia. Avoid drugs metabolized by the liver. Control ventilation and continue hypothermia and all supportive measures
	Schröt, K. Aspirin and Reye syndrome: a review of the evidence. <i>Paediatr Drugs.</i> 2007;9(3):195-204 Pugliese A, Beltramo T, Torre D. Reye's and Reye's-like syndromes. <i>Cell Biochem Funct.</i> 2008;26(7):741-6	<i>Table continues on the following page.</i>

Name	Description	Anesthesia implications
Rheumatoid arthritis	See collagen diseases	
Rieger syndrome	Hypodontia and malformations of anterior chamber of eye. May have other developmental abnormalities including maxillary hypoplasia	Possible difficult airway; have difficult airway cart available
Riley-Day syndrome (familial dysautonomia)	Recessive disorder of autonomic ganglia and sensory neurons found in Ashkenazi Jews. Deficiency of dopamine- β -hydroxylase; autonomic dysfunction and decreased sensation, paroxysmal hypertension, and orthostatic hypotension. Emotional lability, absent lacrimation, abnormal sweating, poor sucking and swallowing. Recurrent aspiration pneumonia and chronic lung disease. Dysautonomic crisis (vomiting, profuse sweating, heart rate and hemodynamic instability) can occur in response to stress	Asai T, Matsumoto H, Shingu K. Difficult airway management in a baby with Axenfeld-Rieger syndrome. <i>Paediatr Anaesth</i> . 1998;8(5):444. Avoid prolonged fasting. Premedication with midazolam and H ₂ -receptor antagonist. Parental presence may help. Atropine can be given. Require IV hydration; replace fluid losses to maintain volume status (monitor CVP if extensive blood loss is anticipated). Sensitive to anesthetic agents; titrate inhalational agents to effect; can use barbiturates, propofol, etomidate, opioids, and relaxants. Respiratory center unresponsive to CO ₂ ; use opioids with caution, may require postoperative ventilation. Risk of aspiration, postoperatively and at induction. Diazepam often controls an autonomic crisis, ranitidine for gastric acidity, and clonidine may be useful to manage postoperative hypertension. Epidural and spinal anesthesia was thought to be contraindicated but has been used successfully in a few cases with increased cardiovascular stability and superior analgesia reported. Caution with eyes, lubricate and cover

Ngai J, Kreymin I, Kim JT, et al. Anesthesia management of familial dysautonomia. *Paediatr Anaesth*. 2006;16(6):611–20
 Ahmed N, Watve MM, Ahmed M. Spinal anesthesia in Kiley-Day syndrome (familial dysautonomia). *Paediatr Anesth*. 2008;18:1136–7

Robin Pierre syndrome	See Pierre Robin syndrome	
Robinow syndrome (fetal face syndrome)	Limb-shortening, facial (midface hypoplasia), and spinal deformities, CHD, renal disease, and hypoplastic genitalia. May be associated with Crigler-Najjar liver disease. (Both diseases a result of consanguinity)	Preoperatively assess cardiac (echocardiogram), renal (BUN/creatinine), and hepatic (if Crigler-Najjar suspected) status. If Crigler-Najjar is present, evaluate coagulation status. Caution with airway recommended but usually not difficult; have difficult airway cart at hand
Romano-Ward syndrome	See Jervell and Lange-Nielsen syndrome	
Rubinstein-Taybi syndrome	Broad thumb and great toes, developmental delay, microcephaly. May have CHD (usually pulmonary stenosis), frequent chest infections, repeated aspiration leading to pneumonia and chronic lung disease. Estimated frequency: 1 of every 500 institutionalized development delayed persons	Preoperatively assess cardiac (echocardiogram) and pulmonary (chest X-ray) status. Anticipate difficult intubation; have difficult airway cart at hand. Caution with respiratory depressants. Secretions may be a problem. Beware of postoperative ventilatory depression or apnea
	Altintas F, Caknakkaya S. Anesthetic management of a child with Rubinstein-Taybi syndrome. <i>Paediatr Anaesth.</i> 2004;14(7):610-11 Agarwal S, Ahmad YH, Talpesh M, et al. Anesthetic management of children with Rubinstein-Taybi syndrome—case reports. <i>Mid East J Anaesth.</i> 2011;21:309-12	
Russell-Silver syndrome	See Silver-Russell dwarfism	
Sandhoff disease	See gangliosidosis GM2	
Saethre-Chotzen syndrome	Acrocephalosyndactyly III, premature closure of cranial sutures with syndactyly of the hands and feet. Hypoplasic maxilla, hypertelorism, malformed ears may be present. Intelligence usually normal. Mutation in TWIST1 gene	Anticipate difficult airway; have difficult airway cart at hand. No other special considerations
	Easely D, Mayhew JF. Anesthesia in a child with Saethre-Chotzen syndrome. <i>Paediatr Anaesth.</i> 2008;18(1):81 Sharma A, Patel N, Arora S, Ramachandran R. Child with Saethre-Chotzen syndrome: anesthetic management and literature review. <i>Acta Anesth Belg.</i> 2014;65:179-82	

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Name	Description	Anesthesia implications
Sanfilippo syndrome (mucopolysaccharidosis type III)	CNS malfunction in childhood progresses to developmental delay and dementia. Emotional disturbance and agitation. No hepatosplenomegaly, cardiac problems, or major bone problems	See mucopolysaccharidoses. No other specific anesthesia problems described
Andrade F, Aldaniz-Echevaria I, Llarena M, et al. Sanfilippo syndrome: overall review. <i>Pediatr Int.</i> 2015;57(3):331–8 Cingi EC, Beebe DS, Whitley CB, et al. Anesthetic care and outcomes in children with Sanfilippo Syndrome Type A. <i>Pediatr Anesth.</i> 2016;26(5):531–8	Congenital hypoparathyroidism, hypocalcemia, hyperphosphatemia, seizures, dwarfism, developmental delay, and dysmorphic features. Recurrent pulmonary infections. Confined to children of Arab descent	Administer anti-seizure medications on the morning of surgery. Preoperatively assess respiratory status (chest X-ray), electrolytes and ionized calcium. Airway and intubation may be difficult; have difficult airway cart at hand. Use short-acting drugs. Monitor postoperative respiratory status
Sanjad-Sakati syndrome (SSS)	Normal or almost normal intellect. Corneal clouding, hernias; joint stiffness, especially of hands and feet; aortic insufficiency. Sleep apnea may occur	Platis CM, Wasersprung D, Kachko L, et al. Anesthesia management for the child with Sanjad-Sakati syndrome. <i>Paediatr Anaesth.</i> 2006;16(11):1189–92 Preoperatively assess cardiac status (echocardiogram). See mucopolysaccharidoses. Monitor for apnea. Caution with opioids, reduce dose. Position with care
Scheie syndrome (mucopolysaccharidosis type IS, formerly classified as type V)	Dwarfism, microstomia, micrognathia, cleft palate, myotonia limiting joint movement, bowing of long bones, thermoregulatory disorder. Usually normal intellect (some degree of developmental delay in 25%)	Difficult intubation; have difficult airway cart available. Fiberoptic intubation has been recommended. Succinylcholine contraindicated; possible abnormal response to nondepolarizing relaxants, avoid inhalation agents. (See also myotonia and paramyotonia)
Schwartz-Jampel syndrome	Perks WH, Cooper RA, Bradbury S, et al. Sleep apnea in Scheie's syndrome. <i>Thorax.</i> 1980;35:85–91	Ray S, Rubin AP. Anaesthesia in a child with Schwartz-Jampel syndrome. <i>Anaesthesia.</i> 1994;49(7):600–2

Scleroderma	Diffuse cutaneous stiffening. May have hemifacial atrophy. Plastic surgery required for contracture and constrictions. May have cardiac fibrosis or cor pulmonale (rare in children—but common cause of death). Esophageal dilation leads to GERD. Children have less multiple organ involvement than adults—more arthritis and myositis. Raynaud phenomena may rarely be present. Therapy may include steroids, methotrexate, Ca^{++} channel blockers	Preoperatively assess cardiac status (echocardiogram). Check history of steroid therapy and other drug history. Scarring of face and mouth, possible difficult airway and intubation; have difficult airway cart at hand. Chest restriction, poor compliance. Diffuse pulmonary fibrosis—hypoxia. Veins may be invisible, impalpable, difficult to enter. Prevent hypothermia
Sebaceous nevi syndrome, linear	Zulian F. Systemic sclerosis and localized scleroderma in childhood. <i>Rheum Dis Clin N Am.</i> 2008;34(1):239–55 Roberts JG, Sabar R, Gianoli JA, et al. Progressive systemic sclerosis: clinical manifestations and anesthetic considerations. <i>J Clin Anesth.</i> 2002;14(6):474–7	Linear nevi from the forehead to the nose; hydrocephalus, developmental delay; may have coarctation and/or hypoplasia of aorta
Seckel syndrome	Autosomal recessive disorder, developmental delay, dwarfism, microcephaly (“birdlike” facies), prominent maxilla, micrognathia, pointed nose	Preoperatively assess cardiac status (echocardiogram). May have increased ICP

Gurkan Y, Hosten T, Dayioglu H, et al. Anesthesia for Seckel syndrome. *Pediatr Anesth.* 2006;16:359–60
Deniralp G, Mayhew J. Anesthesia in a child with Seckel syndrome. *Pediatr Anesth.* 2007;17:1121
Ural Y, Dogan AI, Ozkose Z, et al. Anesthetic management of a patient with Seckel syndrome and implanted pacemaker. *Paediatr Anaesth.* 2008;18(7):676–7

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Name	Description	Anesthesia implications
Seip syndrome	See lipotrophy with diabetes	
Shy-Drager syndrome	Orthostatic hypotension; diffuse degeneration of central and autonomic nervous systems; inability of pulse and blood pressure possibly because of defective baroreceptor response; decreased sweating; hypersensitivity to catecholamines and angiotensin	Caution with potent inhalation anesthetics; accurate fluid replacement important; treat hypotension with IV fluids and phenylephrine; vasopressin may be the best agent to treat refractory hypotension. Use muscle relaxants with caution
	Hutchinson RC, Stugden JC. Anaesthesia for Shy-Drager syndrome. <i>Anaesthesia</i> . 1984;39(12):1229-31 Vallejo R, DeSouza G, Lee J. Shy-Drager syndrome and severe unexplained intraoperative hypotension responsive to vasopressin. <i>Anesth Analg</i> . 2002;95:50-2	
Silver-Russell dwarfism	Short stature, skeletal asymmetry, micognathia. Low birth weight. Café au lait spots, endocrine abnormalities, hypogonadism	Possible difficult mask ventilation and intubation; have difficult airway cart at hand. Monitor blood glucose level. Prone to hypothermia. Caution with relaxants; monitor block
Dinner M, Goldin EZ, Ward R, et al. Russell-Silver syndrome: anesthetic implications. <i>Anesth Analg</i> . 1994;78(6):1197-9 Passier RH, Verwijs E, Driessens J. Anaesthesia for orphan disease: management of an infant with Silver-Russell syndrome. <i>Eur J Anaesth</i> . 2014;31:336-8		
Sipple syndrome (multiple endocrine adenomatosis type 2 (MEN 2))	Three forms: MEN 2A, familial medullary thyroid carcinoma, and MEN 2B. Pheochromocytoma more common in MEN2A and MEN2B (bilateral in 75% of cases), medullary thyroid carcinoma, parathyroid adenoma (common in MEN 2A), multiple endocrine neoplasia. MEN 2B also presents with mucocutaneous neuromas and muscular hypotonia	See pheochromocytoma, page 367. Problem of multiple endocrine disorders. For MEN 2B, evaluate for the presence of muscular hypotonia before considering muscle relaxants
Carney JA. Familial multiple endocrine neoplasia: the first 100 years. <i>Am J Surg Pathol</i> . 2005;29(2):254-74		

Sleep apnea syndromes	<p>Disorders of breathing during sleep, including the following</p> <ol style="list-style-type: none"> Central sleep apnea due to CNS immaturity (sudden infant death syndrome), trauma, infections, or neoplasms, and primary central alveolar hypoventilation (Ondine curse). Apnea occurs without evidence of respiratory muscle activity Obstructive sleep apnea due to obesity, adenotonsillar hypertrophy, Pierre Robin syndrome, or any other condition causing chronic airway obstruction. Apnea occurs because of obstruction and is accompanied by increased respiratory muscle activity. Response to CO₂ is blunted. Ventilatory depression with opioids is markedly increased and opioid requirements for analgesia markedly decreased Mixed forms. Medical history may include daytime somnolence, loud snoring, restless sleep, insomnia, fatigue. Children may be hyperactive and aggressive. 	<p>Review sleep study if done with attention to severity of nocturnal desaturation (if nocturnal SaO₂ nadir <85% = opioid sensitivity may be increased). Assess airway. Small risk of desaturation with preoperative sedation. Beware of acute airway obstruction during induction of anesthesia. Intubate airway; allow spontaneous respiration to assess opioid sensitivity. May require only one-half to one-third of usual dose of opioids (morphine), normalized to ideal body weight. Use opioid sparing approach such as around the clock acetaminophen and ibuprofen. Awaken patient completely before transfer to postanesthesia care unit (PACU). Monitor closely for apnea postoperatively. (See Chap. 10 for sleep apnea strategy during anesthesia)</p>
Smith-Lemli-Opitz syndrome	Inborn error of cholesterol synthesis. Microcephaly, cleft palate, developmental delay, genital and skeletal anomalies (including micrognathia), thymic hypoplasia, hypotonia; may have increased susceptibility to infection	<p>Use sterile technique. Anticipate airway and intubation problems; have the difficult airway cart at hand. Use muscle relaxants with caution. Muscle rigidity with inhalational anesthetics has been described, but this syndrome is not associated with MH and has not been associated with rhabdomyolysis.</p> <p>Consider TIVA</p>

Bandla P, Brooks LJ, Trimarchi T, et al. Obstructive sleep apnea syndrome in children. *Anesth Clinics N Am.* 2005;23(3):535-49
 Lerman J. A disquisition on sleep-disordered breathing in children. *Pediatr Anesth.* 2009;19 Suppl 1:100-8
 Coté CJ, Posner KL, Domino KB. Death or neurologic injury after tonsillectomy in children with a focus on obstructive sleep apnea: houston, we have a problem! *Anesth Analg.* 2014;118(6):1276-83

Quezado ZM, Veithmeyer J, Schwartz L, et al. Anesthesia and airway management of pediatric patients with Smith-Lemli-Opitz syndrome. *Anesthesiology.* 2002;97(4):1015-9

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Name	Description	Anesthesia implications
Sotos syndrome (cerebral gigantism)	Macrocephaly, dilated cerebral ventricles but normal ICP. Developmental delay. Hypotonia. Accelerated growth during childhood. Prone to hernias. Cardiac and GU abnormalities in a few patients. Reduced immune response	Assess cardiac status (echocardiogram and ECG); SBE prophylaxis if indicated. Care with asepsis. Intubation reported to be easy. Care with padding and positioning head. Hyperthermia during anesthesia is reported (not MH); monitor temperature and institute cooling as indicated. Regional analgesia may be indicated in some cases
Spinal muscle atrophy	Degeneration of anterior spinal neurons leads to generalized progressive muscle weakness. Bulbar dysfunction may be present. Intellect and sensation normal	Aldhami EJ, Cancio-Babu CV. Anaesthesia in a child with Sotos syndrome. <i>Paediatr Anaesth</i> . 2003;13(9):835-40 Chierichini A, Messina A, Vergari A, et al. Regional anaesthesia in a child with Sotos syndrome. <i>Int J Immunopharm Regional</i> . 2011;24(1 Suppl 2):21-3 Intubation may be difficult; have difficult airway cart at hand. Risk of GI reflux. Avoid succinylcholine. Caution with relaxants—may have prolonged effect. Monitor blood glucose. Evaluate post-op ventilation and provide support. Provide good analgesia; caution with opioids. Regional analgesia may be difficult but is ideal

Islander G. Anesthesia and spinal muscle atrophy. *Pediatr Anesth*. 2013;23: 804-16

Stevens-Johnson syndrome (erythema multiforme)	Urticular lesions; erosions of the mouth, eyes, genitalia. Possible hypersensitivity to exogenous agents (drugs, infections, etc.). If pleural bleeds are present, pneumothorax may occur. Dehydration and malnutrition are common. May have myocarditis, pericarditis. Medical care is similar to that of children with a burn injury; some may be on high-dose steroid therapy	Preoperatively assess cardiac status (echocardiogram and ECG), fluid status, and pulmonary function. Check for recent steroid therapy. Use sterile technique (reverse isolation). Oral lesions: avoid intubation and insertion of esophageal stethoscope, gentle pharyngeal suctioning. Use soft face mask with Vaseline gauze on skin. If intubation is required, secure tracheal tube with tracheostomy tape that is well padded; do not use adhesive tape. Monitoring is difficult (because of skin lesions) but essential; cover ECG pads with surgical lubricant and place beneath the patient. Warm the operating room to approximately 35–37 °C; danger of severe hypothermia. Monitor closely; serious arrhythmias and ventricular fibrillation may occur. IV infusion essential but avoid cutdowns if possible (possibility of infection). Ketamine is probably the best anesthetic agent
Stickler syndrome	Autosomal dominant disorder with midface hypoplasia, micrognathia, cleft palate, and “moon-face” appearance. Progressive myopia, retinal degeneration, and hearing loss. Spinal abnormalities and mitral valve prolapse are also common	Preoperatively assess cardiac status (echocardiogram). Anesthesia problems similar to Pierre Robin patients. Airway maintenance and intubation may be very difficult; have the difficult airway cart at hand

Smith GB, Shribman AJ. Anaesthesia and severe skin disease. *Anaesthesia*. 1984;39(5):443–55

Madan R, Chawla R, Dhar P, et al. Anesthesia in Stevens Johnson syndrome. *Indian Pediatr*. 1989;26(10):1038–40

Stickler syndrome

Autosomal dominant disorder with midface hypoplasia, micrognathia, cleft palate, and “moon-face” appearance. Progressive myopia, retinal degeneration, and hearing loss. Spinal abnormalities and mitral valve prolapse are also common

Kucukyavuz Z, Ozkaynak O, Tuzuner AM, et al. Difficulties in anaesthetic management of patients with micrognathia: report of a patient with Stickler syndrome. *Oral Surg Oral Med Oral Pathol Radiol Endod*. 2006;102(6):e33–6

Fujii M, Tachibana K, Takeuchi M, et al. Perioperative management of 19 infants undergoing glossopexy (tongue-lip adhesion) procedure: a retrospective study. *Pediatr Anaesth*. 2015;25(8):829–33

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Name	Description	Anesthesia implications
Stiff baby syndrome (hyperkplexia, “startle disease”)	Rare, genetic syndrome. Severe muscle rigidity appears at birth and persists for several years. Exaggerated startle response is present. Life-threatening spasms may be terminated by flexing the head and legs toward the trunk. Choking, vomiting, and difficulty swallowing may occur. EMG shows continuous muscle activity	Use caution with muscle relaxants; monitor effects carefully. (Sevoflurane may cause fade on TOF monitor). May be resistant to succinylcholine but have normal response to non-depolarizing muscle relaxants. Effect of neostigmine is normal. Opioids increase rigidity. Propofol may be appropriate. Monitor for perioperative apnea
Still disease (juvenile rheumatoïd arthritis)	See collagen diseases	Garg R, Ramachandran R, Sharma P. Anaesthetic implications of hyperkplexia—“startle disease”. <i>Anaesth Intens Care</i> . 2008;36(2):254–6 Murphy C, Shorten G. Train of four fade in a child with stiff baby syndrome. <i>Paediatr Anaesth</i> . 2000;10(5):567–9
Sturge-Weber syndrome	Cavernous angioma over trigeminal nerve distribution, usually unilateral. Developmental mesodermal capillary defect. Glaucoma. Intracranial calcification, convulsions, developmental delay. Possible laryngeal and tracheal involvement	Often have port wine stains treated. Care with instrumentation of larynx in case of undiagnosed angioma. Care to prevent hypertension or raised intraocular pressure during intubation or extubation. Often treated with repeat laser therapy; LMA well suited for this procedure. No other specific anesthetic problems
Supravalvar aortic stenosis with idiopathic infantile hypercalcemia	See Williams syndrome	Batra RK, Gulaya V, Madan R, et al. Anaesthesia and the Sturge-Weber syndrome. <i>Can J Anaesth</i> . 1994;41(2):133–6 Ceyhan A, Cakan T, Basar H, et al. Anaesthesia for Sturge-Weber syndrome. <i>Eur J Anaesthesiol</i> . 1999;16(5):339–41

Tangier disease (alpha-1lipoproteinemia)	Low-plasma high-density lipoproteins; accumulation of cholesterol esters in large orange tonsils, spleen, and lymph nodes. Anemia and thrombocytopenia. Peripheral neuropathy and abnormal EMG; premature coronary disease (lipid deposits found in coronary arteries of a 6-year-old). (No reports of ischemic heart disease in children)	Preoperatively assess cardiac/coronary status (echocardiogram and ECG), Hb, and platelet counts. Use caution with cardiac depressants and muscle relaxants. Monitor for cardiac ischemia and avoid tachycardia and hypertension as per adult with ischemic heart disease
Tay-Sachs disease	See gangliosidosis GM2	
Thomsen disease	See myotonia congenita	
Telangiectasia, hemorrhagic	See Osler-Rendu-Weber syndrome	
Thalassemia major (Cooley anemia)	Hereditary disease that may affect any race but is most common in Mediterranean and Southeast Asians. Slow rate of Hb synthesis and high percentage of HbF are present. Low Hb levels require repeated transfusion leading to hemosiderosis and cardiac dysfunction. MRI may be used to assess severity of iron overload. May require chelation therapy from birth. Partial or total splenectomy may reduce the need for transfusions	Preoperatively assess cardiac status (echocardiogram and ECG). Hemosiderosis may affect heart and hepatic function. Anemia may be severe. Facial deformity: overgrowth of maxilla may cause difficult intubation; have difficult airway cart at hand. Anesthesia considerations for the anemic patient (see page 191). Special attention to aseptic techniques for children who have had a splenectomy. Heterozygous form (thalassemia minor) poses no special anesthesia problems

Firth PG. Anesthesia and hemoglobinopathies. *Anesthesiology Clinics*. 2009;27:321-36

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Name	Description	Anesthesia implications
Thromboasthenia	See Glanzmann disease	Preoperatively assess cardiac function if CHD present (echocardiogram). Platelet transfusion for surgery or bleeding; consult with hematologist. Avoid elective surgery in the first year (35–40% mortality from intracranial hemorrhage)
Thrombocytopenia with absent radius (TAR syndrome)	Thought to be due to a 1.q21.1 chromosomal deletion. Episodic thrombocytopenia precipitated by stress, infection, surgery, etc. Platelets increase to normal by adulthood. CHD in 30% of cases (especially tetralogy of Fallot and ASD); also associated with tracheoesophageal and renal anomalies	Goldfarb CA, Wall L, Manske PR. Radial longitudinal deficiency: the incidence of associated medical and musculoskeletal conditions. <i>J Hand Surg Am.</i> 2006;31(7):1176–82
Thrombocytopenia with eczema and repeated infections	See Wiskott-Aldrich syndrome	Morrison JE Jr, Lockhart CH. Tourette syndrome: anesthetic implications. <i>Anesth Analg.</i> 1986;55(2):200–2
Thrombotic thrombocytopenic purpura	See Moschcowitz disease	Yoshikawa F, Takagi T, Fukayama H, et al. Intravenous sedation and general anesthesia for a patient with Gilles de la Tourette's syndrome undergoing dental treatment <i>Acta Anesthesiol Scand.</i> 2002;46:1279–80
Tourette syndrome	Complex neuropsychiatric disorder with onset in childhood. Attention-deficit disorder progresses to spasmodic repetitive movements that may become powerful muscle jerks. Patient also may exhibit coprolalia (profane speech) and echolalia (repetitions). High incidence of migraine headaches. Treated with haloperidol, clonidine, or pimozide	Establish rapport with patient and family. Continue medications. Sedate preoperatively. No specific anesthesia regimen is indicated, except that pimozide may cause prolonged QT interval syndrome (see page 571)

Treacher-Collins syndrome (mandibulofacial dysostosis)	Micrognathia and slanting jawbone, aplastic zygoma, microstomia, choanal atresia, coloboma of eyelids, microtia. Patients often have cleft palate and cardiac anomalies. Airway becomes progressively more difficult to manage with age. Most have normal intelligence, but often are hearing impaired	Preoperatively assess cardiac status (echocardiogram). Mask ventilation may not be possible. Having a surgeon capable of establishing a surgical airway is reasonable. Difficult airway adjuncts must be immediately available and a fiberoptic or GlideScope intubation should be anticipated; topical airway analgesia with nebulized lidocaine will facilitate awake insertion of an LMA and is used to induce anesthesia and as a conduit for intubation in the neonate or infant. The child should be fully awake before extubation. Some children require tracheotomy
Nargozian CN.	The airway in patients with craniofacial abnormalities. <i>Pediatr Anesth.</i> 2004;14:53-9	
Hosking J, Zoanetti D, Carlyle A.	Anesthesia for Treacher Collins syndrome: a review of airway management in 240 pediatric cases. <i>Pediatr Anesth.</i> 2012;22:752-8	
Fawley G, Espenell A, Howe P, et al.	Anesthetic implications of infants with mandibular hypoplasia treated with mandibular distraction osteogenesis. <i>Pediatr Anesth.</i> 2013;23(4):342-8	
Trismus- pseudocampiodactyly (Dutch-Kentucky syndrome)	Autosomal dominant condition. Decreased mouth opening due to enlarged coronoid process of the mandible and/or abnormal ligaments plus flexion deformity of the fingers when wrist is extended. Short stature and foot deformities may occur. May be present for surgery to mandible	Extremely difficult intubation; have the difficult airway cart at hand. Difficult airway adjuncts must be immediately available, and a fiberoptic or GlideScope intubation should be anticipated
Vaghadia H, Blackstock D.	Anaesthetic implications for the trismus pseudocampiodactyly (Dutch-Kentucky or Hecht Beals) syndrome. <i>Can J Anesth.</i> 1988;35:80-85	
Toydemir RM, Chen H, Proud VK, et al.	Trismus-pseudocampiodactyly syndrome is caused by recurrent mutation of MYH8. <i>Am J Med Genet A.</i> 2006;140(22):2387-93	

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Name	Description	Anesthesia implications
Trisomies		
Trisomy 13	See Patau syndrome	
Trisomy 18 [E]	See Edwards syndrome	
Trisomy 21	See Down syndrome	
Tuberous sclerosis (TSC)	Neurocutaneous condition with hamartoma growth in body. Multisystem disease: sebaceous adenoma of skin, epilepsy, developmental delay, intracranial calcification, seizures. May have tumors in the brain, heart, lungs, and kidneys; pyelonephritis and renal failure may occur. Cardiac rhabdomyomas are benign but may occur in up to 90% of neonates. These tumors tend to remain the same in size (as the heart grows less important) or resolve spontaneously	Administer anti-seizure medications in the morning of surgery. Preoperatively assess cardiac (echocardiogram) and renal (BUN /creatinine) status; cardiac tumors tend to resolve spontaneously but may cause right outflow tract obstruction, particularly in infants. Care with drugs excreted by the kidney, especially radiocontrast material during repeat MRI assessments that generally occur q6 months. Possible cardiac arrhythmia and rupture of lung cysts.
		Anesthetic management depends on preoperative examination and limitations of organ functions found. Maintain normothermia and normocarbia to prevent seizures. Check anticonvulsant levels postoperatively
		Diaz JH. Perioperative management of children with congenital phakomatoses. Pediatr Anesth. 2000;10:121-8 Mettin RR, Merkenschlager A, Bernhard MK, et al. Wide spectrum of clinical manifestations in children with tuberous sclerosis complex—follow-up of 20 children. Brain Dev. 2014;36(4):306-14

Turner syndrome (gonadal dysgenesis)	XO females. Short stature, infantile genitalia, webbed neck; possible micrognathia. CHD coarctation, dissecting aneurysm of aorta (teenagers) or PS. Hypothyroidism in some cases. Renal anomalies in more than 50% of cases	Preoperatively assess cardiac (echocardiogram) and renal (BUN/creatinine) status. Intubation may be difficult; have the difficult airway cart at hand. Maintain normal blood pressures. Ensure thyroid replacement therapy is up to date. Care with renal excreted drugs
Umbilical hernia in infancy	Be alert to possibility of Beckwith syndrome	Avoid anticholinergics (excessive drying). Gentle laryngoscopy and intubation to prevent mucosal trauma. Vocal cords may be thickened. Airway and intubation may be difficult; have the difficult airway cart at hand. Administer anti-seizure medications in the morning of surgery
Urbach-Wiethe disease (cutaneous mucosal hyalinosis)	Hoarseness or aphonia (eosinophilic hyaline-like deposits in larynx and pharynx) and skin eruption. Tongue may be thickened, dry and friable. Mucous membranes thickened, dry and friable. Intracranial calcification and epilepsy may develop. Highly associated with consanguineous parents	Avoid anticholinergics (excessive drying). Gentle laryngoscopy and intubation to prevent mucosal trauma. Vocal cords may be thickened. Airway and intubation may be difficult; have the difficult airway cart at hand. Administer anti-seizure medications in the morning of surgery
VATER association (VACTERL association)	A nonrandom association of defects: <i>V</i> vertebral anomalies (congenital scoliosis), <i>A</i> anal atresia, <i>T</i> tracheoesophageal fistula, <i>E</i> esophageal atresia, <i>R</i> renal anomalies (<i>C</i> cardiac disease and <i>L</i> limb defects also in VACTERL)	Examine neonates with any of these features for other congenital lesions. Preoperatively assess cardiac (echocardiogram) and renal (BUN/creatinine) status. Anesthesia management dictated by considerations of individual lesions

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Jain D, Supriya S, Gurpreet K. Association of difficult airway to VACTERL anomaly: Anesthetic challenge. *Anaesth Pain & Intens Care*. 2013;17(2):192–4
Solomon BD, VACTERL/VATER Association. *Orphanet J Rare Dis*. 2011;16(6):56

Name	Description	Anesthesia implications
Velocardiofacial syndrome	A phenotypic variant of the 22q deletion syndrome, separate from DiGeorge syndrome. Speech difficulties due to velopharyngeal anomalies, learning disability (mild), CHD (especially VSD), and characteristic facies: large nose with broad nasal bridge, vertically long face, narrow palpebral fissures, and retruded mandible. Associated OSA. Long-term risk for psychiatric disorder especially schizophrenia.	Preoperatively assess cardiac status (echocardiogram). Airway and intubation may be difficult; have the difficult airway cart at hand. May present for pharyngoplasty. Obstructive sleep apnea may occur after pharyngoplasty; use one-third to one-half the normal weight normalized dose of opioid or opioid-sparing strategy with acetaminophen and ibuprofen. Postoperative monitoring for apnea is recommended Gotheff D, Frisch A, Michaelovsky E, et al. Velo-Cardio-Facial Syndrome. J Ment Health Res Intellect Disabil. 2009;2(2):149-67 Kirschner RE and Baylis AL. Surgical considerations in 22Q11.2 deletion syndrome. Clin Plast Surg. 2014;41(2):271-82
Very long-chain acylcoenzyme A dehydrogenase deficiency	Disorder of fatty acid metabolism leads to potential for hypoglycemia, liver failure, cardiomyopathy, and rhabdomyolysis	Preoperatively assess cardiac (echocardiogram) and liver status. Minimal fasting and administer glucose infusions perioperatively. Propofol is contraindicated (due to emulsion vehicle). Avoid succinylcholine. Avoid myocardial depressant medications. Low-dose volatile agents, opioids, regional analgesia, and NSAIDS are considered safe Redshaw C, Stewart C. Anesthetic agents in patients with very long-chain acyl-coenzyme A dehydrogenase deficiency: a literature review. Pediatr Anesth. 2014;24:1115-19

von Gierke disease type I (glycogen storage disease)	Developmental delay, marked hepatomegaly, renal hyperplasia, stomatitis, lactic acidosis, leucopenia, and bleeding diathesis (usually nose bleeds secondary to impaired platelet aggregation). Fasting causes hypoglycemia and convulsions. Severe biochemical disturbances; hypoglycemia is unresponsive to epinephrine and glucagon. May also have Fanconi syndrome (see Fanconi syndrome). Associated hepatic tumors and pancreatitis	Preoperatively evaluate renal (BUN/creatinine) and hepatic status. Continuous IV glucose infusion preoperatively and perioperatively; avoid lactated Ringer's solution. Monitor blood sugar and acid-base balance. Caution with propofol. Report of postoperative pancreatitis in a patient with this disease. Type III (Cori disease; Forbes disease) similar to but milder than type I. Type VI (Hers disease) similar to but milder than type I
von Hippel-Lindau syndrome	Retinal angiomas and cerebellar hemangioblastomas; pheochromocytoma in some; may have pulmonary, pancreatic, hepatic, adrenal, renal cysts. Paroxysmal hypertension due to cerebellar tumor or pheochromocytoma	Preoperatively assess renal (BUN/creatinine) and hepatic status. Investigate for pheochromocytoma (urinary vanillylmandelic acid). Hypertensive crises may occur. Preoperative management for pheochromocytoma for several days prior to surgery. Laparoscopic approach to tumor excision is now most common

Shenkman Z, Golub Y, Meretyk S, et al. Anaesthetic management of a patient with glycogen storage disease type 1b. *Can J Anaesth.* 1998;43:467-70
Bustamante SE, Appachi E. Acute pancreatitis after anaesthesia with propofol in a child with glycogen storage disease type 1A. *Paediatr Anaesth.* 2006;16:680-3

Gurunathan U, Korula G. Unsuspected pheochromocytoma: von Hippel-Lindau disease. *J Neurosurg Anesthesiol.* 2004;16:26-8
Widimsky J Jr. Recent advances in the diagnosis and treatment of pheochromocytoma. *Kidney Blood Press Res.* 2006;29(5):321-6

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Name	Description	Anesthesia implications
von Recklinghausen disease (neurofibromatosis)	Café au lait spots (>5); tumors in all parts of the CNS and peripheral tumors associated with nerve trunks. Tumors may occur in the larynx or trachea and right ventricular outflow tract; kyphoscoliosis in 50%; neck may be unstable. May have fibroblasts "honeycomb (cystic) lung" predisposing to pulmonary problems. Renal artery dysplasia (hypertension) common. Pheochromocytoma in 1% (all these patients should be investigated—urinary vanillylmandelic acid). Often require a q6-month MRI surveillance	Preoperatively assess pulmonary (chest X-ray), renal (BUN/creatinine), and cardiac function (echocardiogram) as indicated. Intubation could be complicated by tumor; have difficult airway cart at hand. Caution with neck; may need in-line stabilization. Titrate neuromuscular blocking drugs; effects of non-depolarizing muscle relaxants may be prolonged. If kidneys are involved, care with drugs excreted by kidneys including contrast material
von Willebrand disease (pseudohemophilia)	Prolonged bleeding time (decreased von Willebrand factor and associated decreased factor VIII activity leading to defective platelet adhesiveness) and capillary abnormality. History of bruising and bleeding (menorrhagia, epistaxis, etc.). Several types of the disease exist and determine the response to therapy. IV desmopressin acetate (DDAVP) is effective therapy for type 1 and some type 2 patients, but may exacerbate others. (Consult hematologist.) Types 2 and 3 require factor VIII concentrates. Bleeding may be controlled by transfusions of fresh-frozen plasma and/or cryoprecipitate. Antifibrinolytics may help	Preoperatively consult with surgeon and hematologist to develop optimal management strategy. Monitor factor VIII and bleeding time and maintain factor VIII at >50% activity

Allen GC, Armfield DR, Bontempo FA, et al. Adenotonsillectomy in children with von Willebrand disease. Arch Otolaryng Head & Neck Surg. 1999;125:547-51
 Maquoi I, Bonhomme V, Bonf D, et al. Perioperative management of a child with von Willebrand disease undergoing surgical repair of craniosynostosis: looking at unusual targets. Anesth Analg. 2009;109(3):720-4
 Stone ME, Mazzetti M, Derham J, et al. Current management of von Willebrand disease and von Willebrand syndrome. Curr Opin Anaesthesiol. 2014;27(3):353-8

Weaver syndrome	Skeletal overgrowth leading to craniofacial and digital abnormalities. Relative micrognathia, short neck, and anterior larynx lead to difficult intubation	Caution with airway and intubation; have difficult airway cart at hand. Difficulties may be less in older children
Crawford MW, Rohan D. The upper airway in Weaver syndrome. <i>Paediatr Anaesth</i> . 2005;15:893–6		
Weber-Christian disease (chronic nonsuppurative panniculitis)	Necrosis of fat in any situation, including the following: Retoperitoneal tissue—may cause acute or chronic adrenal insufficiency. Pericardium: leads to restrictive pericarditis. Meninges: causes convulsions	Preoperatively assess cardiac (echocardiogram) and renal status. Prevent trauma to fat by heat, cold, or pressure. Maintain blood volume; use cardiac depressant drugs and drugs excreted by the kidneys with caution
Spivak JL, Lindo S, Coleman M. Weber-Christian disease complicated by consumption coagulopathy and microangiopathic hemolytic anemia. <i>Johns Hopkins Med J</i> . 1970;126:344–9		
Werdnig-Hoffmann disease (infantile muscular atrophy, SMA type I)	Onset in infancy; severe scoliosis with onset in first year of life. >95% require gastrostomy feeding by 24 months of age. Fundoplication is another common procedure to prevent aspiration. Chronic respiratory problems. Most patients die before puberty	Preoperatively assess severity of pulmonary system and extent of weakness. Succinylcholine is contraindicated (due to hyperkalemia). Use muscle relaxants or respiratory depressant drugs with caution. Ventilatory support may be required, and weaning may be difficult
Mercuri E, Bertini E, Iannaccone ST. Childhood spinal muscular atrophy: controversies and challenges. <i>Lancet Neurol</i> . 2012;11(5):443–52 Bach JR. Medical considerations of long-term survival of Werdnig-Hoffmann disease. <i>Am J Phys Med Rehabil</i> . 2007;86(5):349–55		

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Name	Description	Anesthesia implications	
Werner syndrome (multiple endocrine adenomatosis type 1 (MEN1))	Hyperparathyroidism, anterior tumors of pituitary, insulinomas of the pancreas (hypoglycemia), hyperparathyroidism, and gastric ulcers are common. Carcinoid tumors of bronchial tree may occur. Renal failure due to kidney stones is possible. These patients require frequent MRI evaluations of many organ systems usually on a yearly basis	Preoperatively assess for renal and pituitary dysfunction; presence of carcinoid tumor. Preoperative consult with endocrinologist for specific management issues. Administer glucose at maintenance rates and monitor perioperative glucose values. See also carcinoid tumors (page 536)	
Werner syndrome	Premature aging; diabetes in 50 %, developmental delay in 50 %, early cataracts, osteomyelitis-like bone lesions, cardiac infarction, and failure	Harrison, B. Endocrine surgical aspects of multiple endocrine neoplasia syndromes in children. <i>Horm Res.</i> 2007;68(Suppl 5):105–6 Peranteau WH, Palladino AA, Bhatti TR, et al. The surgical management of insulinomas in children. <i>J Pediatr Surg.</i> 2013;48(12):2517–24 Romero AMA, Morris LF, Rich TA, et al. Preoperative multiple endocrine neoplasia type 1 diagnosis improves the surgical outcomes of pediatric patients with primary hyperparathyroidism. <i>J Pediatric Surgery.</i> 2014;49(4):546–50	Preoperatively assess cardiac status (echocardiogram and ECG) for evidence of myocardial ischemia and dysfunction. If diabetes is present, manage accordingly. Anesthesia conducted as for patients with coronary artery disease; avoid tachycardia and hypertension

Capell BC, Collins FS, Nabel EG. Mechanisms of cardiovascular disease in accelerated aging syndromes. *Circ Res.* 2007;101:13–26
 Dominguez-Gerpe L, Araujo-Vilar D. Prematurely aged children: molecular alterations leading to Hutchinson-Gilford progeria and Werner syndromes. *Curr Aging Sci.* 2008;1(3):202–12

West syndrome	Infantile spasms, hypersarrhythmia on EEG, and psychomotor impairment. Seizures, neurologic deficits, lissencephaly; and severe developmental delay. Long-term treatment with anticonvulsants required. Some patients successfully treated with ketogenic diet	Administer anti-seizure medications in the morning of surgery. If on ketogenic diet, then avoid glucose-containing solutions and lactated Ringer's; administer normal saline. One report describes very low BIS values noted in an "awake" patient with paradoxical changes during anesthesia induction. BIS monitor may be unreliable to monitor anesthesia depth. Parents may be helpful in determining when their child is "awake"
	Valkenburg AJ, de Leeuw TG, Machotta A, et al. Extremely low preanesthetic BIS values in two children with West syndrome and lissencephaly. <i>Pediatr Anaesth.</i> 2008;18:446-8 Kossoff EH, Wang HS. Dietary therapies for epilepsy. <i>Biomed J.</i> 2013;36(1):2-8 McTague A, Cross JH. Treatment of epileptic encephalopathies. <i>CNS Drugs.</i> 2013;27(3):175-84	<i>Table continues on the following page.</i>

Name	Description	Anesthesia implications
Williams syndrome (elfin facies syndrome)	Cardiac anomalies: usually supravalvular aortic stenosis, peripheral pulmonary artery obstruction, and developmental delay (IQ 40–80), elfin facies, some with microgathia, “party personality.” Fixed cardiac output and myocardial ischemia leading to dyspnea and angina. Very high anesthetic risk and difficult to resuscitate. Sudden death may occur. Hypercalcemia, require low calcium diet, steroids, and cardiac corrective surgery. Hypothyroidism and renal abnormalities may be present	<p>Preoperatively assess cardiac (echocardiogram, ECG, and coronary artery anatomy/obstruction), renal function (BUN/creatinine), and calcium. Check steroid history. Monitor calcium levels perioperatively. Mask ventilation and intubation may be difficult; have difficult airway cart at hand. High incidence of cardiac arrest during induction of anesthesia. A careful smooth very slow inhalation induction with modest doses of inhalation agent may be used, but an intravenous induction with an opioid-based anesthetic is recommended and preferred. Avoid cardiac depressants and drugs causing tachycardia. Generous volume loading is essential to avoid hypotension. Transesophageal echocardiography may aid anesthetic management. This cohort has a high representation in the perioperative cardiac arrest registry! Be extremely cautious! Also check website: www.williams-syndrome.org</p>

- Medley J, Russo P, Tobias JD. Perioperative care of the patient with Williams syndrome. *Pediatr Anaesth*. 2005;15:243–7.
- Gupta P, Tobias JD, Goyal S, et al. Sudden cardiac death under anesthesia in pediatric patient with Williams syndrome: a case report and review of literature. *Ann Cardiac Anesth*. 2010;13:44–8.
- Olsen M, Fahy CJ, Costi DA, et al. Anaesthesia-related haemodynamic complications in Williams syndrome patients: a review of one institution's experience. *Anaesth Intens Care*. 2014;42:619–24.

Wilson disease (hepatolenticular degeneration)	Decreased ceruloplasmin; copper deposits, especially in the liver and CNS motor nuclei. Renal tubular acidosis; hepatic failure due to fibrosis	Preoperatively assess renal (BUN/creatinine) and hepatic status. Use muscle relaxants that do not depend upon hepatic or renal function (cisatracurium) to avoid unpredictable responses. Care with drugs excreted by kidneys
Green DW, Ashley EM. The choice of inhalation anaesthetic for major abdominal surgery in children with liver disease. <i>Paediatr Anaesth.</i> 2002;12:665–73 Kaler SG. Inborn errors of copper metabolism. <i>Handb Clin Neurol.</i> 2013;113:1745–54		
Wilson-Mikity syndrome	Prematurity (<1500 g birth weight); severe chronic lung disease leading to generalized fibrosis with cystic interstitial emphysema, repeated chest infection, aspiration, right ventricular failure. Steroids may be given to try to prevent pulmonary fibrosis. Pathogenesis unknown; possibly due to O ₂ toxicity or barotrauma	Preoperatively assess cardiac (echocardiogram) and pulmonary (chest X-ray) status. May have a history of steroid therapy requiring supplementation. Monitor postoperatively for apnea
	Stasic AF. Perioperative implications of common respiratory problems. <i>Sem Pediatr Surg.</i> 2004;13:174–180 Hoepker A, Seear M, Petrochellou A, et al. Wilson-Mikity syndrome: updated diagnostic criteria based on nine cases and a review of the literature. <i>Pediatr Pulmonol.</i> 2008;43(10):1004–12	
Wiskott-Aldrich syndrome	This is an x-linked recessive disease. Decreased production of microplatelets; hypersusceptibility to severe herpes simplex infections (disordered immune mechanism), eczema, asthma, and lymphoid malignancy. Most die before 10 years of age, many from generalized herpes or opportunistic infection. Stem cell transplant has been attempted with mixed results	Antibiotic prophylaxis may be indicated preoperatively. Transfusions of blood and platelets may be required. All blood products must be irradiated to prevent graft-versus-host reaction. Use sterile technique (reverse isolation)
	Notarangelo LD, Miao CH, Ochs HD. Wiskott-Aldrich syndrome. <i>Curr Opin Hematol.</i> 2008;15:30–6 Worth AJ, Booth C, Veys P. Stem cell transplantation for primary immune deficiency. <i>Curr Opin Hematol.</i> 2013;20(6):501–8	<i>Table continues on the following page.</i>

Name	Description	Anesthesia implications
Wolff-Parkinson-White (WPW) syndrome	Anomalous conduction path between atria and ventricles. ECG: Short P-R interval, prolonged QRS with phasic variation in 40%. Prone to paroxysmal supraventricular tachycardia (SVT). May have other cardiac defects. Infants, especially preterm, are very prone to SVT. May be associated with CHD. Prone to arrhythmias. Paroxysmal SVT on induction of anesthesia has been reported; treat with adenosine, beta blockers or calcium channel blockers (e.g., verapamil)	Preoperatively review cardiac status with cardiologist; review echocardiogram and ECG if associated with CHD. Avoid atropine or pancuronium. During electrophysiologic studies, propofol may be anesthetic of choice having little effect on abnormal conduction pathways
Wolman disease (familial xanthomatosis)	Failure to thrive because of xanthomatous visceral changes: adrenal calcification. Resembles Niemann-Pick disease, with hepatosplenomegaly, hypersplenism, and foam cell infiltration (of all tissues, including myocardium.) Death usually by 6 months of age. Trials with enzyme replacement are ongoing and show some promise	Treatment is entirely supportive, but if anesthesia is required, then appropriate assessments of cardiac, pulmonary, hepatic, and hematologic systems are advised
Worster-Drought syndrome	A form of cerebral palsy with childhood onset epilepsy	Postoperative apnea is possible; monitor appropriately
Zellweger syndrome	Chhabra A, Baidya DK. Postoperative cyanotic breath-holding spells in a child with Worster-Drought syndrome. <i>J Anesth.</i> 2010;24(6):982-3 See cerebrohepatorenal syndrome	

Cardiopulmonary Resuscitation, Including Neonatal Resuscitation

Cardiopulmonary resuscitation (CPR) is concerned with the restoration of pulmonary and cardiovascular function and the prevention of neurologic damage. Initially, it consists of artificial ventilation and artificial circulation by whatever means are immediately available. This is termed *basic life support*. Its object is to prevent clinical death from progressing to biologic death before other remedial measures (i.e., *advanced life support*) can be instituted.

As in adults, heroic resuscitative efforts may not be indicated in children with lethal terminal disease. This is a decision that should be made in advance and clearly documented and communicated.

The overall success rate for pediatric CPR is worse than for adults, especially if success is defined as long-term survival without neurologic deficit. A possible reason is that the majority of cardiac arrests in children result from hypoxemia. In such children, it must be assumed that by the time the heart has suffered hypoxia enough to stop it, the brain has also suffered hypoxia enough to severely damage it. This being so, every effort must be directed at detecting and treating any respiratory compromise before it leads to serious hypoxemia.

PREVENTION OF CARDIAC ARREST

Awareness of precipitating factors is essential in preventing cardiac arrest in children.

Common causes include:

1. Failure of ventilation:
 - (a) Due to central depression, airway obstruction, or primary pulmonary disorders
 - (b) Secondary to regurgitation and pulmonary aspiration
 - (c) Secondary to neurologic and neuromuscular disorders (i.e., residual neuromuscular block)
2. Hypovolemia.

3. Toxicity (drugs, poisons, toxins).
4. Primary cardiac disorders. (These account for only a small percentage of cardiac arrests on the general wards of a pediatric hospital.)

Prevention requires:

1. Recognition of potential causes
2. Constant surveillance
3. Early recognition of respiratory failure

Special Hazards for Children

Anesthesiologists should constantly be aware of factors that may be insignificant in the adult but may rapidly be life-threatening in infants and children;

1. The upper airway may become obstructed by:
 - (a) Laryngospasm (common); due to small amounts of mucus or blood or inadequate or unwisely planned anesthesia (see Chap. 4)
 - (b) Hypertrophied adenoidal tissue and/or enlarged tonsils, which may completely block the airway
 - (c) The relatively large tongue, associated with:
 - Muscle flaccidity in the anesthetized patient
 - Inadvertent displacement or compression of submental soft tissue and tongue by the anesthesiologist's fingers
 - Inadequate neck extension
 - Inadequate elevation of the mandible
 - Premature removal of an artificial airway
 - (d) Regurgitated stomach contents—a common occurrence because of the frequency of feedings
Remember: Infants have large tongues and may be primarily nose breathers. If the nasal airway is inadequate, an oral airway should be inserted without delay.
2. Ventilation may be compromised if the stomach becomes inflated, usually a result of:
 - (a) Excessive inflation pressures
 - (b) Partial airway obstruction
 - (c) Crying and swallowing air

After protecting the airway with a tracheal tube, pass a No. 10 or 12 suction catheter into the stomach and aspirate the contents to reduce the possibility of aspiration.
3. Blood volumes are small—significant hypovolemia may develop rapidly.

Routine Precautions

Preoperative

1. Be prepared to give atropine to all young children who are scheduled for laryngoscopy/tracheal intubation or to receive cholinergic drugs (i.e., halothane, succinylcholine) or have surgery that may elicit vagal reflexes that may result in bradycardia and asystole. Vagal reflexes are brisk and may lead to cardiac arrest. The greater the delay before giving atropine, the greater the time until it takes effect. (**N.B.** atropine does not correct hypoxemia!)
2. Always give 100 % O₂ before intubating the trachea in children. (Desaturation occurs much more rapidly in children, particularly infants, than in adolescents). Intubate the trachea as quickly and smoothly as possible.
3. Select the tracheal tube size carefully, secure it firmly, and check its position; confirm EtCO₂ and listen to both sides of the chest. Position and support the tracheal tube so that it cannot kink. (These procedures are critical in children.)

Perioperative

1. Carefully maintain a patent airway and adequate ventilation.
2. Monitor the following constantly:
 - (a) Heart and lung functions by stethoscope, ECG, and NIBP
 - (b) SaO₂ and PetCO₂ (do not disable alarms)
 - (c) Body temperature
 - (d) Blood loss
3. Meticulously measure all gases, vapors, and drugs; always read the drug label.
4. Measure fluid losses accurately and replace as indicated. (Even a small loss is significant in a small child.) Ensure that there is a reliable, generous sized IV route before allowing surgery to start.
5. Remember that rapid infusion of blood products may cause hyperkalemia or hypocalcemia (especially when given into a central vein in small infants).
6. Prevent unintentional pressure on the chest and abdominal wall from dressings, hands, and surgical assistants leaning on the drapes.
7. If problems become apparent, advise other members of the team (especially the surgeon) immediately.

Postoperative

Note: Cardiac arrest in the postanesthesia care unit (PACU) is as common as in the operating room:

1. For all infants and all seriously ill children: do not extubate the trachea unless and until the child is reacting vigorously.

2. Whenever possible, all children should be transported to the PACU in the lateral position, with the upper leg flexed at the hip and knee and the neck moderately extended (the “tonsil” or “recovery” position). Administer oxygen and monitor respiration. SaO₂ is an unreliable monitor of ventilation in the presence of oxygen.
3. In the PACU:
 - (a) Immediately monitor SaO₂, NIBP, and ECG.
 - (b) Provide a full report to the PACU nursing staff regarding underlying medical problems, surgical problems, medication doses and time of administration, and anticipated possible PACU problems.
 - (c) Ensure that the child remains safely positioned with a clear airway:
 - Order humidified O₂ by mask until the child is responding well.
 - Document stable vital signs at handoff.
 - (d) Do not leave until you are assured vital signs are stable and have handed over care of your child to a nurse.
 - (e) Before discharge from the PACU, ensure that the danger of drug-induced respiratory depression has passed and that the child is fully conscious.
 - (f) Some neonates and infants need to be stimulated frequently to maintain ventilation.
 - (g) All former preterm infants of less than 60 weeks conceptual age and those with a history of chronic respiratory disease should be monitored on an apnea alarm for at least 24 h or 12 h apnea-free (see Chap. 2).
 - (h) Children with obstructive sleep apnea (OSA) require extended observation and monitoring and may be sensitive to the respiratory depressant effects of opioids (see Chap. 10).

Treatment of Arrhythmias

Arrhythmias that cause hemodynamic compromise or those that might progress to cardiac arrest must be promptly treated. The advice of a pediatric cardiologist should be obtained whenever this is possible.

Supraventricular Tachycardia (SVT)

- (a) SVT (heart rates >220 bpm in infants or greater than 180 bpm in children suggest SVT) may be difficult to differentiate from sinus tachycardia, but the history and the heart rate usually clarify the diagnosis.
- (b) Early consultation with a pediatric cardiologist is recommended for hemodynamically stable VT. Vagal maneuvers (Valsalva maneuver or ice applied to face) are recommended as initial therapy. Adenosine may be administered and if this is unsuccessful amiodarone may be indicated

(see Appendix A for dosing). If circulatory instability is present, immediate synchronized electrical cardioversion (0.5–1 J/kg) is recommended.

Ventricular Tachycardia (VT) and Ventricular Fibrillation (VF)

- (a) For VT with hypotension but with a palpable pulse, immediate synchronized cardioversion is recommended. Children with less hemodynamic compromise should be assessed for the cause of the VT and may be sedated before cardioversion.
- (b) For pulseless VT and VF, chest compressions followed by very prompt defibrillation is recommended. If this is unsuccessful or VT recurs, amiodarone 5 mg/kg may be considered.

Non-shockable Rhythm (Asystole and Pulseless Electrical Activity [PEA])

Asystole and PEA are the most common ECG findings in cardiac arrest in infants and children. PEA is a cluster of slow, wide QRS complexes in the absence of palpable pulses. CPR should be continued; defibrillation is not indicated. Underlying causes should be sought.

CARDIOPULMONARY RESUSCITATION

The guidelines for cardiopulmonary resuscitation and neonatal resuscitation are based on peer reviewed scientific publications that are critically analyzed to derive a consensus. This appendix incorporates the recommendations from their most recent International Liaison Committee on Resuscitation (ILCOR) deliberations (2010).

Basic Life Support

N.B. For basic life support in hospitals, use a bag and mask as soon as possible to prevent risk of infection to hospital personnel. Make sure that such equipment is immediately available and functioning in all patient-care areas. In rare instances, it may be necessary to resort to mouth-to-mouth resuscitation.

Do not leave the child. Call for help and equipment (including a defibrillator).

Begin with the ABCs:

1. Airway: check patency and apply jaw thrust.
2. Breathing (four ventilations) preferably with bag and mask.
3. Cardiac activity: check with stethoscope—palpate brachial artery in an infant and femoral or carotid artery in a child.

When called to resuscitate a child, assess the situation immediately according to the following priorities:

1. Check ventilation:

(a) *If there are respiratory efforts:*

- Position the child in the lateral position (recovery position) to provide a clear airway and decrease the risk of aspiration should vomiting occur.
- Give O₂ by mask as soon as it becomes available.

(b) *If respiratory efforts are present, but evidence of airway obstruction is present* (breath sounds absent, intercostal retraction, flaring of lateral chest margins, cyanosis):

- Pull the tongue or mandible forward (by pressing behind the mandibular condyles) and remove any foreign matter from the pharynx, keeping the mouth slightly open.
- Extend the neck if necessary (caution if neck injury—but remember that ventilation is the first priority).
- Give O₂ by mask.
- Check for improved chest movement and breath sounds.

2. If there is no respiratory effort or ventilation appears inadequate:

(a) Begin positive pressure ventilation at once.

(b) Ventilate directly, mouth to mouth if necessary, until resuscitation equipment is placed in your hand. (The small infant face necessitates application of your mouth to the infant's mouth and nose.) An infant's tidal volume is small (8–10 mL/kg); therefore, only puffs are necessary.

As soon as possible, begin ventilation using a bag and mask with oxygen. Assure adequate expansion of the chest with each breath.

3. If cardiac activity is undetectable by auscultation or by femoral, carotid, or brachial artery palpation (or if the heart rate is <60 bpm despite ventilation and oxygenation and there are signs of poor perfusion [pallor, cyanosis]):

(a) Start external cardiac compression at once ("push hard, push fast"):

- The site of compression in an infant is one finger-breadth below the intermammary line; in a child, it is over the lower sternum, one finger-breadth above the xiphisternum. In an infant, two fingers may be used. In a child, the heel of the hand should be used.
- Depth of compression: one-third to one-half of the anteroposterior diameter of the chest; between compressions, release completely to allow the chest wall to fully recoil.
- Rate of compression: 100/min for infants, children, and adolescents.

- Rate of ventilation (basic life support—unintubated airway):
 - Single-person technique: 2 ventilations for 30 compressions.
 - Two-person technique: 2 ventilations to 15 compressions.
 - For intubated airways, a rate of 100 cardiac compressions per minute and 8–10 ventilations with 100% O₂ per minute should be initiated immediately. Prevent hyperventilation.
 - Do not interrupt the compressions for ventilation.
 - Apply cardiac compressions to infants by encircling the chest with your hands (Fig. B.1). This method results in a larger cardiac output than anterior sternal compression alone.

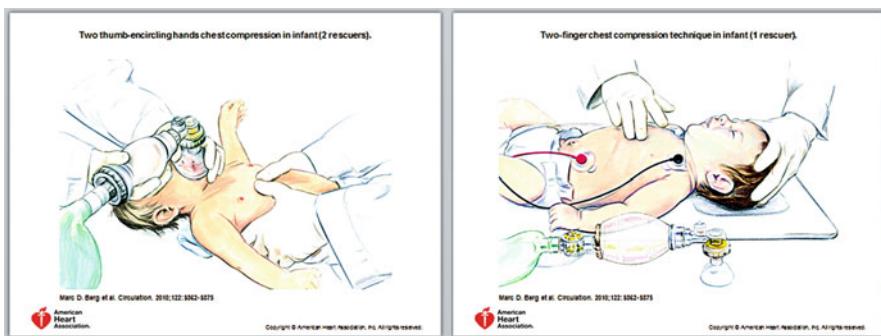


Fig. B.1 Two-handed method of external cardiac compression. Note how both hands encircle the chest and how both thumbs are used for cardiac compression. (From Todres D, Rogers MC: Method of external cardiac massage in the newborn infant. J Pediatr 86:781, 1975, with permission)

Defibrillation

Early defibrillation is recommended for children who have ventricular fibrillation or pulseless ventricular tachycardia. Defibrillation is more successful after effective chest compression in adults. These are more likely when there has been a sudden unexpected arrest:

1. For infants and children weighing less than 20 kg, use pediatric defibrillator plates (diameter of 4.5 cm for infants and 8 cm for children).
2. Set the machine to deliver shocks appropriate to the child's size (to maximize the chance of success and minimize the danger of electrically induced myocardial damage); 2 J (W s)/kg should be the initial setting.
3. Give one shock and then immediately resume CPR for 2 minutes and assess response. If unsuccessful, the dose should be doubled to 4J.
4. Many AEDs have the potential to detect pediatric “shockable” rhythms and can be adjusted to deliver appropriate energy to each shock. Ensure that AEDs installed in a pediatric care environment conform to these requirements.

5. Children who are digitalized should be treated with the reduced power settings initially; then the power setting is gradually increased. Normal doses of countershock may cause irreversible cardiac arrest in the presence of bound digitalis in the heart muscle.

ADVANCED LIFE SUPPORT

The foregoing provides only basic interim resuscitation. Most children also require:

1. Ventilation with O₂ as soon as it is available. Ventilation and oxygenation are the first line of therapy for the acidosis that accompanies cardiac arrest.
2. Establish an airway by rapidly intubating the trachea; a cuffed tube may be used when appropriate to protect the airway. (The laryngeal mask airway may be useful in some instances.) Exhaled CO₂ detection is recommended for early confirmation of successful intubation and to ensure adequate pulmonary circulation with chest compressions. Do not delay ventilation during lengthy attempts at intubation: ventilation is essential, intubation is optional.
3. Definitive ECG diagnosis of cardiac activity and defibrillation if indicated (see previous discussion).
4. Establish an intravenous or intraosseous route for drug administration. Intratracheal drug administration is no longer recommended.
5. Supportive drugs, primarily epinephrine (see later discussion).
6. Further pharmacologic and medical treatment, including fluid replacement, as indicated.
7. Consideration of the possibility of lung injury by aspirated acidic gastric contents.
8. Early assessment of neurologic function—plan early and continue treatment to minimize and prevent further hypoxic-ischemic brain damage.

DRUG THERAPY

Although subsequent drug therapy is necessarily individualized, a standard initial protocol is advantageous:

1. Epinephrine: To be maximally effective, it must be given intravenously (preferably into a central vein) or, if this is not possible, by the intraosseous route:
 - (a) Initial and subsequent doses: 10 µg/kg (0.1 mL/kg of a 1:10,000 solution). Continue CPR for 2 minutes and assess response.
 - (b) High-dose (100 µg/kg) epinephrine is no longer recommended (except possibly in the treatment of β-blocker overdose).

2. Vasopressin has been successful in some cases of prolonged cardiac arrest but has not resulted in increased survival to a neurologically intact hospital discharge.
3. Dopamine infusion may be required for continued hypotension and poor tissue perfusion; 5–20 µg/kg/min may be titrated to achieve the desired effect.
4. Sodium bicarbonate administration is no longer recommended as a routine; but it might be considered in prolonged cardiac arrest for documented continuing severe metabolic acidosis despite adequate ventilation, oxygenation, and chest compressions. It may also be useful in the treatment of hyperkalemia, hypermagnesemia, or tricyclic antidepressant overdose.

N.B. Administration of excessive doses of sodium bicarbonate produces hyperosmolarity, hypernatremia, hypokalemia, decreased ionized calcium, impaired cardiac action, and possibly severe alkalosis after recovery.
5. Calcium is no longer recommended except as the definitive treatment for hyperkalemic-induced arrhythmias or arrest and citrate-induced hypocalcemia.
6. Glucose: Documented hypoglycemia should be treated by glucose infusions. Otherwise, avoid any glucose administration because hyperglycemia (glucose >200 mg/dL) may compromise neurologic outcome after a hypoxic event.

Route of Administration of Drugs

Inject epinephrine into a central line, if one is available; otherwise, use a peripheral intravenous or intraosseous line. The tracheal route is only used as a last resort.

Intracardiac injections should not be made. Damage to the heart and coronary arteries and/or a pneumothorax may result.

Fluid Replacement

1. Insert a large-bore intravenous cannula as soon as possible:
 - (a) To provide a route for drug therapy.
 - (b) For rapid replacement of fluid. (In cardiac arrest, hypoxic capillaries leak rapidly, diminishing the circulating blood volume.)
2. Replace losses initially with crystalloids and later with colloids (plasma or blood) as indicated.

N.B.:

 - (a) Even a child previously in congestive heart failure needs infusions totaling at least 10% of the expected blood volume (EBV; equal to approximately 1% of body weight).

- (b) With recovery, the extravasated fluid returns slowly to the vascular compartment, giving time for assessment of fluid volume and a decision as to whether diuretic therapy is necessary.
3. Avoid the use of dextrose-containing solutions. They may cause hyperglycemia, which may compromise cerebral survival. If hypoglycemia is suspected, it should be confirmed by blood glucose determination and treated accordingly.

Post-resuscitation Care

Emphasis is placed on this as being critical to a favorable outcome:

1. Hyperventilation may be harmful and should be avoided: maintain normocarbia.
2. Induced hypothermia (32–34 °C) for 12–24 h may be considered for children who remain comatose after CPR.
3. Prevention of hyperthermia is essential.
4. Hemodynamic support using vasoactive drugs (e.g., dopamine) should be used when necessary to improve the circulatory status.
5. Target blood glucose levels to achieve normal levels.
6. Restrict fluid replacement: avoid large infusions of crystalloid solutions once cardiovascular stability is ensured.
7. Treat seizure activity with phenobarbital and/or phenytoin (Dilantin).
8. Obtain an early neurologic consultation.
9. Maintain cerebral perfusion pressure.

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NEONATAL RESUSCITATION

The anesthesiologist is frequently called upon to assist at or manage the care of the neonate immediately after birth.

Neonatal resuscitation must be based on a detailed knowledge of the normal physiologic changes that occur during transition to extrauterine life (see Chap. 2) plus a recognition of the pathologic processes in the mother or the fetus that may affect the infant at this time.

Most infants require little help. Those born at term, with clear amniotic fluid, who are crying or breathing and have good tone should be dried and kept warm. No further interventions are necessary.

Others, however, require rapid intervention if serious sequelae are to be prevented. Preexisting maternal or fetal disease and/or events during labor may affect the neonate's status after delivery. Frequently, infants at risk may be recognized before birth, and preparations can then be made for their immediate resuscitation on delivery. However, some infants who have demonstrated no antenatal signs of distress may need urgent intervention after birth.

Immediate Assessment of the Neonate

A rapid assessment must be made to determine the extent of stepwise treatment that is required. This determination is made on the basis of respirations, heart rate, and color. The steps in resuscitation are:

1. Clearing the airway, positioning, stimulating
2. Ventilation
3. Chest compressions
4. Medications or volume expansion

Procedures for Neonatal Resuscitation

These are the latest recommendations (ILCOR 2005) with some background information:

1. Assess respirations.
If the infant is breathing but cyanotic, give oxygen; if cyanosis persists, then ventilate.
If the infant has apnea or HR less than 100, then ventilate.
2. Assess heart rate.
If heart rate is less than 60 bpm, continue to ventilate and initiate chest compressions.
Reassess heart rate; if heart rate remains less than 60 bpm, despite effective ventilation and compressions, then:

Administer epinephrine (10–30 µg/kg IV).

Augment blood volume (10 mL/kg isotonic crystalloid).

Some Notes

If bag-mask ventilation is unsuccessful in achieving good ventilation, intubate the trachea immediately. (In some instances, a laryngeal mask airway may be useful as an alternative.)

Initial Resuscitation: Oxygen vs. Room Air

Whether to use supplementary oxygen or air to ventilate the lungs in the neonate is controversial. The use of oxygen has been associated with potentially adverse pulmonary and cerebrovascular effects and may result in tissue damage from oxygen-free radicals. Oxygen use during neonatal resuscitation has been linked to the development of childhood cancer. The results of animal studies are conflicting. However, human studies suggest that the results of resuscitation with room air are equal to or better than those when oxygen is used. Hence current recommendations are:

1. If respiratory efforts are absent or inadequate, priority should be given to lung inflation/ventilation using room air.
2. If the heart rate remains low after ventilation is established, priority should be given to support cardiac output with chest compressions.
3. Supplementary oxygen should be considered for infants with continuing central cyanosis. Monitor SpO₂ to prevent hypoxia and hyperoxia.
4. Excessive oxygen may cause damage and should be avoided, especially in the preterm.
5. Avoid overventilation and hypocapnia.

Ventilation Strategies

Initial breaths of the neonate establish the functional residual capacity (FRC). The peak pressures and inflation time required to initiate ventilation and establish the FRC in the apneic neonate have not been determined. Pressures varying from 30 to 60 cm H₂O and ventilation rates of 30–60 per minute have been used successfully in reported series. Preterm lungs may be damaged by overinflation. Continuous positive airway pressure (CPAP) may help stabilize and improve lung function in sick neonates and may be useful in preterm infants, decreasing the need for intubation.

Current recommendations are:

1. Establishing effective ventilation is the primary objective.
2. If bradycardia is present, an increase in heart rate is the primary index of adequate ventilation.

3. Chest wall movement should be assessed if heart rate does not increase.
4. If airway pressure is being monitored, an initial inflation pressure of 20 cm H₂O may be effective but some full-term infants may require 40 cm H₂O.
5. For preterm infants, prevent excessive distention of the lungs as evidenced by chest wall movement. Initial inflation pressures of 20–25 cm H₂O are usually adequate.

Self-inflating bags, flow-inflating bags, or T-piece systems can be used. The LMA is not recommended as a primary airway device but may be useful if intubation is unsuccessful or “not feasible.” No recommendations are made regarding the use of CPAP in resuscitation.

Chest Compressions

Chest compressions are indicated for a heart rate less than 60 bpm despite adequate ventilation with supplementary oxygen for 30 s.

1. Compressions should be delivered at a rate of 100 per minute over the lower third of the sternum, preferably using the chest encircling/thumbs compressing technique.
2. Ventilation to compression ratio should be 1:3, coordinated to prevent compression during an inspiratory phase of ventilation.
3. Check the heart rate every 30 s (stethoscope).
4. Compressions should continue until the spontaneous heart rate equals 60 bpm.

Medications

These are rarely indicated in neonatal resuscitation, and there is a lack of data regarding the value of drugs in improving outcomes. High doses of epinephrine may reduce survival rates and increase neurologic damage. Naloxone, if given to an infant of opioid addicted mother, may cause seizures:

1. Epinephrine may be indicated if the heart rate remains less than 60 bpm despite adequate ventilation and cardiac compressions. The IV route is recommended (0.01–0.03 mg/kg); high doses are not recommended. The intratracheal route is not recommended.
2. Naloxone is not recommended for initial neonatal resuscitation. Depressed ventilation should be treated with bag and mask.
3. Sodium bicarbonate is not recommended.

Volume Expansion

Volume expansion is indicated for infants when blood loss is suspected, if the infant appears pale with a weak pulse and has not responded fully to other measures:

4. Isotonic crystalloid solution, 10 mL/kg, is recommended and may need to be repeated.
5. Caution: In preterm infants, large volumes rapidly infused have been associated with intraventricular hemorrhage.

Meconium

Intrapartum suctioning has not been demonstrated to reduce the incidence of meconium aspiration syndrome and is no longer recommended. Tracheal suctioning should be performed on meconium-stained depressed infants before stimulation. Meconium stained vigorous infants do not require suctioning.

Post-resuscitation Care

1. Temperature control; hyperthermia is bad and increases the risk of mortality and morbidity. Selective head cooling may decrease the incidence of cerebral morbidity following encephalopathy.
2. Check blood glucose level and treat hypoglycemia.

The Preterm Infant

Some special considerations are necessary for the very small infant:

1. Special care must be taken to prevent heat losses; immediately dry and place the infant on a warm mattress under a heating lamp. Use humidified oxygen.
2. Infants weighing greater than 1000 g should be given O₂, suctioned, and stimulated.
3. Infants weighing less than 1000 g are very likely to require early intubation and ventilation. Be prepared to intervene rapidly unless the infant obviously is in satisfactory condition.
4. Any preterm infant displaying respiratory difficulty should be intubated to provide for optimal ventilation and oxygenation.

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Drug Doses

PREOPERATIVE PERIOD

N.B. Avoid giving drugs intramuscularly (IM) if possible. IM injections are painful and children do not like them. If IM drugs are necessary and more than one has to be given, combine them in the same syringe whenever possible.

Drugs for Premedication

Anticholinergics

Atropine: IV 0.02 mg/kg at induction (maximum dose, 0.6 mg); IM 0.02 mg/kg 30–60 min preoperatively (maximum, 0.6 mg). PO same dose, 60–90 min preoperatively.

Glycopyrrolate: 0.01 mg/kg IV or IM.

Sedatives

Midazolam (Versed): 0.5–0.75 mg/kg PO, 0.2 mg/kg intranasally, 1.0 mg/kg PR, 0.1 mg/kg IM, or 0.05–0.1 mg/kg IV (in a monitored area).

Clonidine: 4 µg/kg oral, or 1–2 µg/kg intranasal.

Dexmedetomidine: 2.5 µg/kg oral or 1 µg/kg intranasal. (**N.B.** No data on neurotoxicity for intranasal injection.)

Lorazepam (Ativan): for adolescents, 1–2 mg PO.

Midazolam/ketamine mixture: 0.3–0.5 mg/kg midazolam combined with 2–6 mg/kg ketamine plus 0.02 mg/kg atropine PO (This combination may result in considerable sedation—use in a monitored setting).

Antacids: H₂-Histamine blocking agents

Cimetidine: 10 mg/kg PO, or 30 mg/kg PR, or 5 mg/kg IV.

Ranitidine: 2–5 mg/kg PO, or 1.5 mg/kg IV or IM.

Sodium citrate: 0.4 mL/kg PO.

Drugs to Speed Gastric Emptying

Metoclopramide: 0.15 mg/kg IV. (Note: Atropine blocks the effect of metoclopramide and should be withheld until induction of anesthesia.)

Topical Local Anesthetics

Eutectic mixture of local anesthetics (EMLA): *prilocaine* (2.5%) and *lidocaine* (2.5%). Apply to skin 60–90 min before procedure. Cover with occlusive dressing. Caution: metabolism of prilocaine may result in methemoglobinemia in neonates—limit application to 1 g EMLA cream over 10 cm² skin.

Amethocaine gel (Ametop): *Tetracaine* (4%). Apply to skin 45 min before procedure. Cover with occlusive dressing. Caution: not recommended for infants <1 month.

Ela-Max (4% Lidocaine): Apply to skin 30 min before procedure. Cover with occlusive dressing.

S-Caine Patch: Eutectic mixture of 70 mg *lidocaine* and 70 mg *tetracaine* in each patch. Apply 20 min before procedure. Cover with occlusive dressing.

INTRAOPERATIVE PERIOD

Induction Agents

Thiopental sodium (Pentothal): neonates (younger than 1 month), up to 3–4 mg/kg; infants (1 month–1 year), up to 7–8 mg/kg; children, up to 5–6 mg/kg.

Etomidate: 0.25–0.3 mg/kg.

Methohexitol: up to 2 mg/kg IV or 15–25 mg/kg of a 1% or 20–30 mg/kg of a 10% solution PR.

Propofol (Diprivan): infants 1.5–2 mg/kg, children 2.5–3.5 mg/kg.

Ketamine: 2 mg/kg IV or 4–8 mg/kg IM (plus atropine 0.02 mg/kg IM/IV).

Drugs for Intubation

Succinylcholine: infants, 2 mg/kg IV; older children, 1 mg/kg IV or 4–5 mg/kg IM.

Rocuronium: 0.3–1.2 mg/kg IV.

(N.B. Large doses of rocuronium in infants have a rapid onset but may result in prolonged blockade).

Vecuronium: 0.1 mg/kg IV.

(N.B.: Do not inject rocuronium or vecuronium immediately after thiopental; thiopental precipitates and may occlude IV).

Cis-atracurium: 0.1–0.2 mg/kg IV.

Pancuronium: 0.1 mg/kg IV.

Topical lidocaine for laryngeal spray: maximum dose 4 mg/kg.

Maintenance

Fentanyl: bolus doses 1–2 µg/kg IV prn.

IV infusion for major surgery: loading dose 5 µg/kg, infuse at 2–4 µg/kg/h.

Hydromorphone: 0.01–0.02 mg/kg IV.

Morphine: 10–100 µg/kg IV or intravenous infusion (for children older than 5 years of age); loading dose 100 µg/kg over 5 min, infusion at 40–60 µg/kg/h.

Remifentanil: Loading dose—0.5 to 2 µg/kg IV.

IV Infusion—0.05–0.3 µg/kg/min.

Acetaminophen: 30–40 mg/kg single-dose PR followed by 20 mg/kg q6h or 10–15 mg/kg PO q4–6 h (maximum daily dose 90–100 mg/kg); 7.5 mg/kg q6h IV for neonates and infants <10 kg or 10 mg/kg q6h IV for children >2 years (maximum daily dose 75 mg/kg)

Neuromuscular Blocking Drugs

1. Usual route of administration IV; IM succinylcholine only in emergency situation.
2. Give initial and repeat doses preferably as indicated by nerve stimulator, especially in infants (whose response to these drugs is extremely variable).
3. Remember that potent inhalational anesthetics (especially sevoflurane and isoflurane) reduce the dose requirement of non-depolarizing drugs.
4. Infusion rates are given as a guide only and should be modified as indicated by neuromuscular blockade monitoring.

Cis-atracurium: initial dose 0.1 mg/kg IV, repeat dose 0.03 mg/kg.

Infusion: loading dose 0.1 mg/kg, infusion at 2–3 µg/kg/min.

Pancuronium: initial dose 0.06–0.1 mg/kg IV; repeat doses should not exceed one sixth of the initial dose.

Rocuronium: initial dose 0.3–1.2 mg/kg IV/IM, incremental doses 0.15 mg/kg.

Infusion: rate: 10–12 µg/kg/min.

Vecuronium: loading dose 0.1 mg/kg, incremental doses 0.02 mg/kg.

Infusion rate 0.1 mg/kg/h.

Antagonism of Neuromuscular Blockade

Atropine 0.02 mg/kg or *glycopyrrolate* 0.01 mg/kg mixed with *neostigmine* 0.05 mg/kg-administer IV slowly; use a nerve stimulator to monitor effect;

OR

Atropine 0.02 mg/kg, followed by *edrophonium* 1 mg/kg IV.

POSTOPERATIVE PERIOD

Analgesics

Acetaminophen (*Tylenol*): 10–20 mg/kg q4–6 h PO or 30–40 mg/kg dose PR followed by 20 mg/kg q6h (maximum daily dose 90–100 mg/kg); 10 mg/kg q6h IV (children >2 years) (maximum daily dose <75 mg/kg)

Acetaminophen with codeine elixir: each 5 mL contains 120 mg acetaminophen + 12 mg codeine

[**N.B. The FDA issued a box warning regarding the use of codeine in children with obstructive sleep apnea after tonsillectomy because of deaths in ultra-rapid metabolizers. Codeine is no longer recommended for use in any child**]

Volume of elixir to administer should be equivalent to 1 mg/kg codeine PO q4 h

Codeine (useful for minor surgery): 1–1.5 mg/kg IM. (Note: Codeine must not be given intravenously).

[**N.B. Codeine is no longer recommended for use in children**]

Diclofenac: 1 mg/kg PO or PR.

Hydromorphone (*Dilaudid*): Bolus dose: 0.01–0.02 mg/kg IV q3 to 4 h.

Continuous infusion rates: 3–5 µg/kg/h.

Caudal analgesia: 10 µg/kg.

Ibuprofen: 10 mg/kg PO.

Ketorolac: 0.5–1.0 mg/kg IV (maximum 15 mg for children <50 kg and 30 mg for >50 kg).

Morphine: children, 0.05–0.1 mg/kg; infants, 0.05 mg/kg IM or IV.

Morphine infusion: children, 10–30 µg/kg/h. To prepare a solution mix: [0.5 × the child's weight (kg)] mg morphine in 50 mL saline. The solution then contains 10 µg/kg/mL. Infuse at 1–3 mL/h for postoperative analgesia (equivalent to 10–30 µg/kg/h).

For infants, give 5–15 µg/kg/h (i.e., 0.5–1.5 mL/h).

Caudal or epidural morphine: 30 µg/kg single dose.

Spinal morphine (preservative free): 10 µg/kg single shot.

Nalbuphine: 0.1–0.2 mg/kg IV (Max 5 mg/kg)

IV Infusion 0.1–0.2 mg/kg/h.

Oxycodone: 0.1–0.2 mg/kg PO q4h.

Tramadol: 1.0–2.0 mg/kg q6h PO. (>4 years)

Opioid Antagonist

Naloxone (Narcan): 0.5–2 µg/kg IV or IM. This drug should be titrated slowly until undesired opioid effects are reversed. Rapid administration of an excessive dose results in loss of analgesia, pain, and extreme restlessness. The same dose of naloxone (µg IV) that resulted in desired effect should then be given IM to prevent recrudescence.

Prophylaxis Against PONV

First-line medications

Dexamethasone: 0.0625–0.15 mg/kg IV (maximum 8 mg).

Ondansetron: 0.05–0.15 mg/kg IV.

Metoclopramide: 0.15 mg/kg IV.

Second-line medications

Dimenhydrinate (Gravol, Dramamine): 1 mg/kg IV or 2 mg/kg PR.

Ancillary Drugs

Single IV antibiotic doses are listed below (maximum daily dose is shown in parentheses). The smaller dose should be given to neonates less than 1 week of age (limited neonatal liver and renal function). Antibiotics should be infused over several minutes only (i.e., *never* give antibiotics as IV boluses!) to minimize the possibility of adverse reactions. Some must be given over a greater period (e.g., vancomycin). Regimens for antibiotic prophylaxis against subacute bacterial endocarditis are listed on page 447.

*Ampicillin*²: 25–100 mg/kg (300 mg/kg).

Cefazolin: 20–40 mg/kg (100 mg/kg).

Cefoxitin: 20–40 mg/kg (160 mg/kg).

Cefuroxime: 20–50 mg/kg (240 mg/kg).

Clindamycin: 5–10 mg/kg (30 mg/kg).

Cloxacillin: 12–25 mg/kg (100 mg/kg).

Erythromycin: 2.5–5 mg/kg (20 mg/kg).

Gentamicin: 2.0 mg/kg (7.5 mg/kg).

Benzyl penicillin (see Footnote 2): 30,000–50,000 IU/kg (250,000 IU/kg).

Vancomycin (see Footnote 2): 10 mg/kg (60 mg/kg) (must be given over a period of at least 1 h).

²Use caution in children with renal failure.

Adrenocorticosteroids

Dexamethasone (Decadron): 0.2–0.5 mg/kg IV (maximum, 10 mg).

Methylprednisolone (Solu-Medrol): 5–25 mg/kg IV slowly over 10 min.

Hydrocortisone sodium succinate (Solu-Cortef): 1–5 mg/kg IV over 8–10 min.

Cardiovascular Drugs (titrate infusions to effect) (see below for infusion setup)

Adenosine: 100 µg/kg as a rapid IV bolus. Repeat up to maximum dose 0.3 mg/kg or 12 mg.

Amiodarone: loading dose 5 mg/kg (over 30–60 min) IV.

Amrinone: loading dose 0.75 mg/kg IV; infusion 3–5 mg/kg/min in neonates and 5–10 mg/kg/min in children.

Calcium chloride: 5–15 mg/kg IV.

Calcium gluconate: 10–30 mg/kg IV.

Dopamine: 5–20 µg/kg/min infusion.

Dobutamine: 5–20 µg/kg/min infusion.

Epinephrine: 0.1–1 µg/kg/min infusion.

Esmolol: 100–500 µg/kg IV, 50–100 µg/kg/min infusion.

Hydralazine: 0.1–0.2 mg/kg IM or IV.

Isoproterenol (Isuprel): 0.025–0.1 µg/kg/min infusion.

Lidocaine: 1–2 mg/kg IV.

Milrinone: 50–100 µg/kg loading dose over 15–30 min (reduce loading dose if hypotension occurs) followed by 0.5–1.0 µg/kg/min infusion.

Norepinephrine (Levophed): 0.1–1 µg/kg/min infusion.

Nitroglycerin: 1–10 µg/kg/min infusion.

Phenoxybenzamine: loading dose 0.25 mg/kg × 4 over 2–4 h; maintenance, 0.25 mg/kg q6h.

Phentolamine: 0.2 mg/kg IV.

Phenylephrine: 0.1–1 µg/kg/min infusion.

Procainamide: 5–15 mg/kg IV.

Propranolol: 0.01–0.1 mg/kg IV over 10 min.

Prostaglandin E₁: 0.05–0.1 µg/kg/min (starting dose). (Maintenance infusion rate, e.g., after PDA open, may be between 0.005 and 0.4 µg/kg/min.)

Sodium nitroprusside: 0.5–10 µg/kg/min infusion.

Verapamil: 0.1–0.3 mg/kg IV. (Do not give to infants younger than 1 year of age).

Diuretics

Ethacrynic acid: 0.5–1 mg/kg IV.

Furosemide (Lasix): 1 mg/kg IV.

Mannitol: 0.5–1.0 g/kg IV (administer over several minutes to prevent transient hypotension).

Anticonvulsants

Diphenylhydantoin (Dilantin): loading dose 15–20 mg/kg IV slowly; maintenance, 2.5–5 mg/kg bid IV or PO.

Phenobarbital: loading dose 10 mg/kg IV; maintenance, 1.5–2.5 mg/kg bid IV.

Bronchodilators

Salbutamol (Albuterol): loading dose 5–6 µg/kg IV; infusion 0.1–1.0 µg/kg/min; inhaled aerosol 100 µg dose q6 h (delivery of aerosol through pediatric tracheal tubes is only 3–10%; to deliver a more effective dose, activate the canister (during inspiration); (1) into a spacer, (2) through a catheter inserted part way down the tube, or (3) after inserting it into the barrel of a 60 mL syringe that is connected to the CO₂ port and reinserting the plunger into the barrel).

Aminophylline: loading dose 5 mg/kg over 30 min IV; infusion 1 mg/kg/h (if no recent doses). Monitor blood levels (therapeutic range, 10–12 µg/mL).

Local Anesthetics

Recommended safe maximum doses

Lidocaine plain: 5 mg/kg

Lidocaine with epinephrine: 7 mg/kg

N.B. The maximum recommended dose of epinephrine to be infiltrated during halothane anesthesia is 10 µg/kg.

Bupivacaine: 2.5 mg/kg

Ropivacaine: 2.5 mg/kg

DRUGS INFUSIONS FOR INFANTS AND CHILDREN

These formulas are designed to permit medications to be infused with limited fluid volumes. (**N.B.** Weight = the child's weight in kilograms).

Dopamine or dobutamine:

Weight × 6 mg of drug in 100 mL; then 1 mL/h = 1 µg/kg/min

In the case of neonates and infants <10 kg use:

(Weight × 30) mg of drug in 100 mL; then 1 mL/h = 5 µg/kg/min

Epinephrine:

(Weight × 0.6) mg of drug in 100 mL; then 1 mL/h = 0.1 µg/kg/min

Sodium nitroprusside or nitroglycerin:

(Weight × 6) mg of drug in 100 mL; then 1 mL/h = 1 µg/kg/min

Isoproterenol:

(Weight \times 0.15) mg of drug in 100 mL; then 1 mL/h = 0.025 $\mu\text{g}/\text{kg}/\text{min}$

Prostaglandin:

(Weight \times 60) μg of drug in 20 mL; then 1 mL/h = 0.05 $\mu\text{g}/\text{kg}/\text{min}$

DRUGS TO REDUCE BLEEDING

Desmopressin: May improve platelet function and reduce bleeding in some platelet diseases. Dose: 0.3 $\mu\text{g}/\text{kg}$ by slow infusion over 20 min after weaning from cardiopulmonary bypass. Monitor cardiovascular parameters carefully during infusion.

ϵ -Aminocaproic acid (Amicar): Used to treat fibrinolytic states. May reduce post-operative bleeding, especially in cyanotic children. Should be administered before sternotomy. Loading dose: 40–100 mg/kg (maximum 5 g) diluted and infused slowly over 20–60 min followed by continuous infusion during surgery at 10–30 mg/kg/h.

Tranexamic acid: a synthetic drug that forms a reversible complex with both plasminogen and plasmin by combining at lysine binding sites inhibits fibrinolysis and reduces bleeding. Loading dose 100 mg/kg IV followed by infusion during surgery at 10 mg/kg/h.

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