Chapter 10: Preoperative Evaluation

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INTRODUCTION

FOCUS POINTS

1. Past medical history with emphasis on prior anesthetic experiences and familial disorders (ie, bleeding, malignant hyperthermia, hemoglobinopathies) should be explored during the preoperative visit.
2. Fasting guidelines have been modified to allow for the age of the child and for clear liquids up to 2 hours prior to general anesthesia.
3. In general, children with chronic disorders should take their medications on the day of surgery (exceptions are diuretics, antihypertensives).
4. Asthma is the leading cause of chronic illness in children; specific questions such as history of emergency department visits, recent oral or intravenous steroid use, and hospitalizations should be inquired during the preanesthetic period.
5. Sleep-disordered breathing (SDB) affects about 10% of the population with about 1% to 4% progressing to obstructive sleep apnea syndrome (OSAS).
6. Children with cold symptoms have an increased risk for perioperative complications such as atelectasis, bronchospasm, laryngospasm, and postoperative pneumonia.
7. Former premature infants are at risk for postanesthetic apnea especially if the hematocrit is less than 30%.
8. Children with history of congenital heart disease should have the most updated cardiology note and procedures (ECG, Echo, cardiac catheterization, CXR) documented in the chart prior to any anesthetic administration.

There is an increasing desire among patients and families to be involved in the perioperative decision-making process.[1] Informed consent in pediatric surgical and interventional procedures requiring general anesthesia involves a shared decision-making process between the multidisciplinary physicians, patient, and family. Shared decision making has the potential to increase satisfaction with care, reduce decisional conflict and regret, improve understanding of and participation in care, and thereby improve health-related quality of life. It is important that the anesthesiologist should be able to accurately estimate and describe the risks of the proposed anesthetic management to the family. Pediatric risk assessment tools may be used to communicate these patient-specific risks.[2]

Anesthetic risk can be decreased by maximizing the information known about the patient's health prior to induction of anesthesia. The overall incidence of cardiac arrest in children under 18 years of age has been reported to be 2.9–4.95/10,000 and of these 18.28/10,000 or 0.18% were below 1 year of age.[3] Since there is no substitute for the long-term relationship that pediatrician, family, and patient have, it is the responsibility of the perioperative anesthesia care team to assess the patient's current health status as it compares with the usual state of health. Both acute and chronic diseases should be evaluated and optimized prior to anesthesia. The choice of anesthetic agent and mode of delivery is a multifactorial decision; however, state of health, both current and prior, is the major determinant. Appropriate laboratory examination should be performed prior to the time of surgery to provide adequate opportunity to adjust or optimize a patient's current health status. If consultation by another specialist is warranted, this can be planned and accomplished prior to anesthesia and surgery. Not only will this planning optimize anesthetic risk factors, but will also prevent cancellation as a consequence of inadequate documentation of health status or data that cannot be retrieved.

PSYCHOLOGICAL PREPARATION

The understanding of and response to illness is affected by a child's maturity. The medical practitioner should anticipate the child's needs and concerns and be able to interpret the child's nonverbal expressions and actions when the child's communication skills are not highly developed. Diseases carry with them different psychosocial aspects for children as compared to adults. For many healthy children who undergo elective surgery, the emotional disruption may surpass the medical issues. Children respond to the prospect of surgery in a varied and age-dependent manner, and the anesthesiologist must consider this during the preoperative interview.

The toddler's greatest fear is the loss of control for actions and choices. The preschooler fears injury, loss of control, the unknown, and abandonment. The preschooler interprets words literally and is unable to differentiate between what is heard and what is implied. The words adults use with children are as important as the messages they try to convey. Because preschoolers are unable to distinguish between reality and fantasy and exist in a world of magical thinking, they cannot recognize the difference between safe sleep during anesthesia and the kind of "sleep" from which their animal did not awaken. The school-aged child fears loss of control, injury, inability to meet the expectations of adults, and death. Between the ages of 6 and 12, children begin to think more logically; yet they may nod with understanding and listen intently, when in fact they do not fully grasp the explanation. These children may fail to ask questions or admit a lack of knowledge because they feel that they should know the information. Adolescents fear loss of control, an altered body image, and segregation from peers. They are usually convinced that the anesthesiologist will not be able to put them to sleep and that, if the anesthesiologist does succeed, they will never wake up.[4]

PAST MEDICAL HISTORY

The child's prior anesthetic experience should be explored during the preoperative visit, since reactions to previous anesthetics may guide the choice of techniques to utilize or avoid. Was general anesthesia previously induced with a mask? Was the parent present for induction? Was premedication used? Was the induction stormy? Were there any sequelae after the hospital experience, such as nightmares, regression to earlier behavior, or new fears of odors? Family history should be explored for anesthesia-related events. Malignant hyperthermia (MH) is always a concern in the pediatric age group and high fevers or unusual perioperative events in the operating room or in family members should be investigated.[5] Although most pediatric anesthesiologists refrain from routinely using succinylcholine, questions about prolonged paralysis or mechanical ventilation after general anesthesia in family members should be asked. If there is a possible history of pseudocholinesterase deficiency, laboratory examination should be performed to determine if that child is at risk. Family members should be queried for a history of unexpected death, sudden infant death syndrome (SIDS), genetic defects, or familial conditions such as muscular dystrophy, cardiomyopathy, cystic fibrosis, sickle cell disease, bleeding tendencies, or human immunodeficiency virus (HIV) infection.

A complete review of systems should be included with emphasis placed on medical comorbidity, which might influence either the choice or outcome of the anesthetic. The presence of cough, asthma, or a recent upper respiratory infection might predispose the child to laryngospasm, bronchospasm, atelectasis, or pneumonia. The new onset of a heart murmur, cyanosis, hypertension, exercise intolerance, or a history of rheumatic fever might suggest an evolving problem which could become exacerbated during the administration of an anesthetic or with a surgical procedure. Parents should be questioned for the presence of vomiting, diarrhea, malabsorption, black stools, gastroesophageal reflux, or jaundice to reveal electrolyte imbalance, dehydration, hypoglycemia, anemia, or the need for a rapid sequence induction. The presence of seizures, head trauma, or swallowing problems may indicate a metabolic derangement, increased intracranial pressure, or sensitivity to muscle relaxants and the anesthetic plan should be altered accordingly. The presence of a urinary tract abnormality should be sought in an attempt to evaluate the state of hydration and integrity of renal function. Abnormal development, alterations in serum glucose levels, or a history of chronic steroid use may indicate an endocrinopathy, diabetes mellitus, hypothyroidism, or adrenal insufficiency. Finally, a history of anemia, bruising, or excess bleeding may suggest a transfusion requirement or coagulopathy, which should be investigated prior to the time of surgery.

PREGNANCY TESTING

The decision to require pregnancy testing prior to administration of anesthesia is institution dependent. Most hospitals do, however, have guidelines for mandatory pregnancy testing for all post-menarchal females prior to anesthesia.[6]

The rate of undiagnosed pregnancy in adolescent females is approximately 1%. Fetal exposure to some anesthetic agents may increase the risk of

spontaneous miscarriage, teratogenic effects, and apoptosis in the rapidly developing brain. For these reasons it has been suggested that elective surgery should be postponed in pregnant female patients.[7,8]

FASTING GUIDELINES

Liberalization of oral intake results in a less anxious child, calmer parents, better maintenance of hemodynamic parameters, and less risk of intraoperative hypoglycemia. Fasting guidelines for children before general anesthesia have been modified to recommend that restricting children to fasting after midnight is no longer common practice and children should be encouraged to drink clear fluids up to 2 hours prior to the time of general anesthesia.[9]

In general, most institutions allow the ingestion of clear fluids until 2 to 3 hours before the time of surgery. These include water, electrolyte solutions (Pedialyte), glucose water, apple juice, white grape juice, and frozen pops without fruit pulp. Clear fluids are defined as any fluid through which a newspaper can be read. No evidence exists that volume has an impact on gastric emptying time or residual volume; therefore, the quantity of clear fluids is not limited.

Formula and breast milk are not clear fluids. Breast milk is considered to be intermediate between clear fluids and formula usually restricted for 4 hours prior to general anesthesia. Clear policies regarding formula, breast milk, and solids should be established for each institution.

MEDICATIONS, ALLERGIES, ADJUNCT THERAPY, AND BEYOND

The parent should be questioned for current use of antibiotics, antihistamines, or other medicines. Many children have never been on medications, some only have been exposed to antibiotics for a simple illness yet others may have received many medications for complex disease processes. It is essential to obtain a full medication history including nonprescription medications administered for minor illness since many over-the-counter (OTC) cold remedies contain aspirin, nonsteroidal anti-inflammatory drugs (NSAIDs), or other compounds which may interfere with coagulation and platelet function. The use of alternative therapies as well as herbal remedies should be documented since these may complicate the anesthetic management. The American Society of Anesthesiologists does not have a formal position on phytopharmaceuticals or other forms of alternative therapy; however, taking “all natural” agents during the perioperative period may put a patient at risk for untoward events. Weight loss aids may augment sympathetic function, and agents designed to enhance muscle growth (eg, creatine) may alter hepatic and renal functions. It should be common practice during the preoperative interview to document intake of any herbal therapies and determine if an alteration in anesthetic technique is warranted. Similarly, the practice of body piercing is becoming increasingly more common. Metal objects in the skin during surgery and anesthesia increase the risk of burn injury if there is an intraoperative electrocautery malfunction. Additionally, metal objects may become caught on equipment in the OR resulting in tearing of skin and subcutaneous tissue. Large metal objects pierced through the midline of the tongue may interfere with effective laryngoscopy and make securing the airway unnecessarily challenging. These objects may also tear nondisposable laryngeal mask airways. Patients, especially adolescents, should be counseled to remove all metal objects and disclose any body piercing that can’t be seen during the preoperative interview.

Queries regarding known drug allergies should be made in children just like in adults. Inquiry regarding primary and secondhand smoke exposure should be made since there is evidence to suggest that these result in an increase in perioperative airway complications.[10,11] Inquiry regarding illicit drug use should also be included in the adolescent population.

Several groups of pediatric patients are at increased risk for latex allergy including children with spina bifida. Adverse reactions to bananas, latex balloons, other latex-containing toys, or the rubber dam used by a dentist should alert the practitioner to the possibility of latex allergy.[12] Allergy consultation and preoperative RAST testing and skin prick testing should be considered in children with a high index of suspicion. Latex sensitization in general pediatric surgical patients is becoming more common; therefore, there is a need for increased screening of patients.[13]

Most regular medications (exceptions maybe drugs such as diuretics and antihypertensives) should be taken on the morning of surgery with a sip of water including oral suspensions. For children who cannot ingest oral medication without food, a spoonful of grape or apple jelly may be substituted as an acceptable alternative.

SPECIAL CONSIDERATIONS

Anesthesia-Induced Neurotoxicity

There have been recent allegations that commonly used anesthetic drugs are deleterious to the developing brain. Most of the epidemiological studies are retrospective and include significant confounders of underlying pathology and surgery.[14,15]

Animal studies have demonstrated that commonly used anesthetic, sedative, and analgesic agents are associated with neuroapoptosis and neurobehavioral deficits; however, the mechanisms underlying the neurotoxic effects have not been elucidated. It is likely that the group of patients potentially susceptible to the effects of prolonged exposure to anesthetics and sedatives are premature infants requiring neonatal intensive care and complex pediatric patients undergoing long, complicated procedures.[16]

Obstructive Sleep Apnea and Sleep-Disordered Breathing

Obstruction of the oropharyngeal airway by hypertrophied tonsils leading to apnea during sleep is an important clinical entity. Despite only mild-to-moderate tonsillar enlargement on physical examination, these patients have upper airway obstruction while awake and apnea during sleep.[17] In children with long-standing hypoxemia and hypercarbia, increased airway resistance can lead to cor pulmonale. Patients may have electrocardiographic evidence of right ventricular hypertrophy, and radiographic evidence consistent with cardiomegaly.[18] These patients often have dysfunction in the medulla or hypothalamic areas of the central nervous system causing persistently elevated CO_2, despite relief of airway obstruction as well as a hyperreactive pulmonary vascular bed. The increased pulmonary vascular resistance and myocardial depression in response to hypoxia, hypercarbia, and acidosis are far greater than what is expected for that degree of physiologic alteration in the normal population. A thorough investigation of this is essential in children who are at risk.

Sleep-disordered breathing (SDB) is a spectrum of disorders ranging from primary snoring to obstructive sleep apnea syndrome (OSAS). SDB affects 10% of the population but only 1% to 4% will progress to OSAS.[19] Proper screening for and diagnosis of obstructive sleep apnea prior to surgery is important in reducing the associated risks. The STOP-BANG questionnaire which has been developed as a tool to screen adult patients for obstructive sleep apnea (*S*noring, *T*iredness, *O*bservation of apnea during sleep, high blood *P*ressure, *B*MI >35 kg/m^2, *A*ge >50, *N*eck size, *G*ender) is not applicable to children.[20] The STBUR questionnaire has been proposed as an alternative for pediatric patients. It screens for *S*noring, *T*rouble *B*reathing and whether a child is *U*n-*R*efreshed after sleep and has potential to be a reliable predictor of children at risk for perioperative respiratory events.[21,22] Repetitive arousal from sleep to restore airway patency is a common feature as are episodic sleep-associated oxygen desaturations, hypercarbia, and cardiac dysfunction as a result of airway obstruction. Individuals who experience obstruction during sleep may have snoring loud enough to be heard through closed doors or observed pauses in breathing during sleep and the presence of these findings should be documented. Obesity changes craniofacial anthropometric characteristics; therefore, a body mass index of a greater than or equal of 95% for age or greater is a predisposing physical characteristic that increases the risk of developing OSAS.[23] Children with craniofacial abnormalities including a small maxilla and mandible, a large tongue for a given mandibular size, and a thick neck have a similar increased risk.

A history of sleep-disordered breathing should be sought. The physical examination should begin with observation of the patient. The presence of audible respirations, mouth breathing, nasal quality of the speech, and chest retractions should be noted. Mouth breathing may be the result of chronic nasopharyngeal obstruction. An elongated face, a retrognathic mandible, and a high-arched palate may be present. The oropharynx should be inspected for evaluation of tonsillar size to determine the ease of mask ventilation and tracheal intubation. The presence of wheezing or rales on auscultation of the chest may be a lower respiratory component of upper airway infection. The presence of inspiratory stridor or prolonged expiration may indicate partial airway obstruction from hypertrophied tonsils or adenoids.

Chest radiographs and electrocardiograms (ECGs) are not required unless specific abnormalities are elicited during the history, such as recent pneumonia, bronchitis, upper respiratory infection (URI), or history consistent with cor pulmonale, which is seen in children with obstructive sleep apnea syndrome. In those children with a history of cardiac abnormalities, an echocardiogram may be indicated.

The Child with a Cold

Children usually experience three to nine episodes of respiratory infection each year. It is, therefore, likely that especially during the winter months a child has an acute upper respiratory infection, is just recovering from one, or is about to have another. The risks vary from minor complications to death under anesthesia as a result of pathophysiology associated with a respiratory infection.[24,25] Children with a respiratory tract infection have a 2-

to 7-fold increase in perioperative respiratory complications and an 11-fold increase if endotracheal intubation is performed.[26] Complications include atelectasis, oxygen desaturation, bronchospasm, croup, laryngospasm, and postoperative pneumonia.[27] Although definitive criteria for cancelling surgery have not been established, the decision is often subjective. Decisions to cancel or postpone surgery should be made in conjunction with the surgeon and be based on the type of procedure, the urgency of the procedure, and the child's overall medical condition.[28]

Criteria which are suggestive of cancellation include the necessity of endotracheal intubation, parental observation that the child is acutely ill on the day of surgery, the presence of nasal congestion, cough and active sputum production, and whether the child is exposed to passive smoke. If the airway can be adequately maintained with a face mask or laryngeal mask airway, the possible morbidity associated with the respiratory infection may be minimized.

Cough is a sign of lower respiratory involvement and should be evaluated for origin (upper airway or bronchial) and quality (wet or dry). Most children will have clear breath sounds when auscultated during quiet respirations. It is during coughing and crying that rales and rhonchi will best be detected. Bronchial hyperreactivity may exist for up to 7 weeks after the resolution of URI symptoms; although it is often impossible to delay surgery for this length of time, most anesthesiologists would agree that surgery may be scheduled after the acute symptoms have resolved and no sooner than 3 weeks after the initial evaluation.

Asthma

Asthma is a leading cause of chronic illness in the pediatric population and consists of bronchoconstriction, hypersecretion of mucus, mucosal edema, and desquamation of inflammatory cells. The hyperreactive airways are very sensitive to stimuli and endotracheal intubation is one of the most potent stimuli for this.

A thorough history should be taken and must include the age of onset of symptoms, the severity of symptoms, the frequency of wheezing, prior steroid therapy, the frequency of emergency room visits, and the number of hospital admissions for pulmonary problems including necessity of mechanical ventilation. The current medical therapy should be noted as well as any additional therapy which is required during acute exacerbations of the disease. It is important to note if a child is on maximal medical therapy and if wheezing persists despite this. It may not be possible for some children to be free of wheezing prior to the administration of general anesthesia.

All medications, both inhaled and oral, should be administered up to and including the morning of surgery. If children are not on maintenance therapy and only require treatment during acute exacerbations, this therapy should be considered for the 24 hours prior to anesthesia even if the child has no respiratory symptoms. This provides an added protection against intraoperative pulmonary complications.

Cystic Fibrosis

Cystic fibrosis is an inherited multisystem disorder and the major cause of severe chronic lung disease in children. It is characterized by chronic obstruction and inflammation of the airway as well as exocrine gland dysfunction. Children with cystic fibrosis may also have nasal polyps, pansinusitis, rectal prolapse, pancreatitis, cholelithiasis, and insulin-dependent diabetes mellitus. An attempt should be made to optimize pulmonary function prior to general anesthesia. This may be accomplished through the use of bronchodilator therapy, antibiotics, and vigorous pulmonary toilet. Recent pulmonary function tests (PFTs) should be reviewed so that the baseline pulmonary status may be documented. Children as young as 5 years of age can cooperate and complete PFT and these should be checked within 6 months of the planned procedure.

Cardiac Disease

Most children with significant cardiac disease are followed regularly by a cardiologist and should have an interval evaluation by the cardiologist in the preoperative period to detect and document any change. Children who have corrected congenital heart disease should have a description of the repair and current anatomy documented and made available to the anesthesia team. If a defect still exists, management recommendations should be requested from the cardiologist. All current cardiac catheterization data should be reviewed. Children with cardiac disease can be divided into two categories: those who have structural congenital heart disease (corrected and uncorrected) and those who have a heart murmur without structural abnormalities (previously diagnosed or new) and this should be clarified. Heart murmurs in children should be identified as innocent or pathologic and this is easily accomplished by a screening echocardiogram. If a murmur is pathologic, the degree of physiologic and hemodynamic compromise should be determined and the need for antibiotic prophylaxis should be assessed during the preoperative visit.

The Former Premature Infant

Premature infants frequently have complex medical histories and require surgery for a variety of reasons. Many former premature infants have developed bronchopulmonary dysplasia (BPD), which is the result of prematurity itself, mechanical ventilation, and respiratory distress syndrome early in the neonatal period. These infants may have interstitial fibrosis, increased airway resistance, decreased pulmonary compliance, fluid retention, and hyperinflation of the lungs. They may require supplemental oxygen, steroids, and diuretics and should have preoperative radiologic documentation of their pulmonary status. Spinal anesthesia is recommended for appropriate surgical procedures.

Apnea associated with bradycardia is another entity frequently observed in former premature infants. Apnea is usually central in origin and is the result of brainstem immaturity which predisposes these infants to more significant apnea during the postoperative period. This risk is less than 1% in a child born at 35 weeks' gestation if surgery is delayed until after 54 weeks postconceptual age.[29] If surgery cannot wait, then postanesthetic apnea monitoring is required for 24 hours. Postanesthetic apnea has been reported in full-term infants less than 4 weeks of age, so similar monitoring is required. A hematocrit less than 30 is associated with an increased risk of postanesthetic apnea in the former preterm infant; therefore, a preoperative hematocrit is warranted in all patients.[30] The presence of retinopathy of prematurity should be sought and documented as well as a history of intraventricular hemorrhage requiring ventriculoperitoneal shunt placement.

CENTRAL NERVOUS SYSTEM DISORDERS

Myelomeningocele

The incidence of myelomeningocele is 1 per 1000 live births and although 75% of lesions occur in the lumbosacral region, affected children may present with a defect anywhere along the neuraxis.[31]

Dysfunction of the skeletal system, skin, genitourinary tract, and peripheral and central nervous systems may also be present so these organ systems should be fully evaluated during the preoperative visit. There is high incidence of sensitivity to latex-containing products, so an attempt to limit exposure to latex should be made. This caution should be noted during the preoperative visit and "Latex Sensitivity" should be posted clearly in the child's chart.

Seizure Disorders

Seizures are a frequently encountered component of many childhood illnesses and afflictions and occur in 4 to 6 children per 1000. They are a symptom of an underlying central nervous system disorder that must be fully investigated and understood. The history should provide a detailed description of the seizure, including the type, frequency, and severity of symptoms as well as the characteristics of the postictal state so that it may easily be recognized by the OR team should it occur during the perioperative period. All anticonvulsant therapy should be recorded and serum drug levels should be checked. All anticonvulsants should be taken up to and including the morning of surgery. If the child has seizures despite adequate therapy, this should be noted.

Cervical Spine Instability

Children who have had significant trauma as well as children who have a variety of congenital abnormalities are at risk for cervical spine instability. Altered mucopolysaccharide metabolism may predispose children to deformities of the odontoid process resulting in cervical spine instability. Atlantoaxial instability and superior migration of the odontoid process may occur in children with rheumatoid arthritis and skeletal dysplasia. Children with Trisomy 21 have laxity of the transverse ligament and abnormal development of the odontoid process which results in cervical spine instability in 15% of cases. Symptoms include clinical manifestations of cord compression which usually are not manifested until after 5 years of age. Although there are no uniform guidelines regarding preoperative testing in these children, it has been suggested that children who are symptomatic have flexion-extension radiographs of the cervical spine and a neurological consultation. If cervical abnormalities are noted, intubation of the trachea should be undertaken in a neutral head position or with somatosensory evoked potential (SSEP) monitoring of the upper extremities.[32]

HEMATOLOGIC DISORDERS

Sickle Cell Disease

Sickle cell disease is a genetically transmitted autosomal recessive disorder that occurs in 8% of the African American population in the heterozygous form and 0.16% in the homozygous form.[33] Heterozygous sickle cell trait does not affect anesthetic management or perioperative outcome, whereas homozygous sickle cell disease increases the risk of perioperative acute chest syndrome, stroke, myocardial infarction, and sickle cell crises. Preoperative preparation should include a measurement of hemoglobin or hematocrit to ascertain deviation from baseline (usually in the 7–8 g/dL range) with the need for presurgical procedure blood transfusion. Partial exchange transfusion should be performed in order to decrease the level of Hgb S to less than 30% to 40% or transfusing to 10 grams of hemoglobin in severe cases where anemia is severe or there is a history of prior stroke or acute chest syndrome. Patients should be admitted to the hospital 12 to 24 hours in advance of their scheduled procedure and receive vigorous intravenous hydration to optimize intravascular flow.

Hemophilia

Hemophilia is the most common and serious of the inherited coagulation disorders. It occurs in approximately 1 in 10,000 males since the defective gene is carried on the X chromosome. Since factor VIII does not cross the placenta, bleeding during the neonatal period suggests the diagnosis. Ninety percent of children have had a significant bleeding episode by the end of the first year of life. The hallmark of the disease is hemarthrosis so children frequently present to the operating room for surgical treatment of this as well as other emergent and routine surgical procedures.

Factor VIII levels should be measured during the preoperative period and orders for replacement therapy should be written when indicated. A consultation with a pediatric hematologist is advisable to determine if therapy with desmopressin (DDAVP) is indicated in addition to guiding factor replacement. The partial thromboplastin time (PTT) will be prolonged but platelet count and prothrombin time will be normal so routine measurement is not necessary. An order for topical analgesic cream or use of pressurized injectable lidocaine (j-tip) should be provided so that preoperative intravenous cannulation will be less painful if preoperative Factor VIII or DDAVP administration is required. These children exhibit an increased frequency of hematoma formation; therefore, intramuscular premedication should be avoided. Due to the high probability of prior blood component therapy, the HIV and hepatitis status of all patients with hemophilia should be documented so that proper precautions may be taken by caregivers.

von Willebrand Disease (vWD)

This bleeding disorder occurs equally in both sexes and is inherited as an autosomal dominant trait. Clinical manifestations include nosebleeds, bleeding from gums, prolonged bleeding from bruises and lacerations, and increased bleeding during surgery. In contrast to hemophilia, hemarthrosis is rare in vWD. The defect is in the platelet binding protein but the platelet count is normal. The prothrombin time (PT) is normal and the partial thromboplastin time (PTT) may be slightly prolonged. Consultation with a pediatric hematologist is suggested to obtain specialized tests to identify the type of vWD to guide therapy during the perioperative period. Therapy consists of DDAVP or replacement therapy with von Willebrand factor from either fresh frozen plasma or cryoprecipitate.

ENDOCRINE DISORDERS

Diabetes Mellitus

The prevalence of diabetes mellitus in school aged children is 1.9 per 1000. The goal of perioperative glucose management is to maintain serum glucose as close to the patient's usual level despite the effects of stress on serum hormonal levels and fasting. It is essential that the physician (pediatrician or endocrinologist) who manages the child's glucose control on a regular basis should be consulted for input on the acceptable range of serum glucose values in each specific patient and how those are best achieved. There should also be clear guidelines for glucose and insulin management if serum glucose measurement either increases or decreases from the acceptable range. Diabetic children should be scheduled as the first morning case to minimize the period of fasting. The serum glycosylated hemoglobin (HbA1C) should be measured during the preoperative period to determine the efficacy of long-term glucose control. Age-appropriate fasting guidelines should be observed and serum glucose should be measured on the morning of surgery.

Diabetes Insipidus

Children with diabetes insipidus may be effectively managed in a variety of ways during the perioperative period and consultation with an endocrinologist at the specific institution is suggested. One effective method of maintaining electrolyte balance without the compounded difficulty of over- or underhydration involves a two-tiered process. Surgery in children with diabetes insipidus should be scheduled as the first case of the day. All patients should have serum electrolytes and osmolality documented preoperatively to establish a baseline. For children who are scheduled for ambulatory or minor surgery, DDAVP should be administered at the usual time and in the usual dose on the day prior to and morning of surgery.

For children who are scheduled for major surgery in which a significant blood loss or fluid shift is anticipated, the DDAVP administration should be modified. A complete history of the dose and timing of DDAVP should be documented as well as the time that urinary breakthrough occurs. If a child receives DDAVP twice a day, the evening dose should be administered prior to surgery but the morning dose should be omitted. If a patient receives a single daily dose it should be omitted on the day of surgery if it is usually taken in the morning but administered as half the usual dose in the evening prior to surgery if it is usually administered at night. In children who have had their DDAVP omitted preoperatively, intraoperative fluid management may be aided by the administration of intravenous vasopressin. Postoperative observation in the ICU should be arranged during the preoperative visit.

Neuromuscular Disorders

Diseases of the motor unit are not uncommon in children and since the combination of certain muscular diseases and specific anesthetic agents may be lethal, identification of these children during the preoperative period is essential. The specific association of malignant hyperthermia and hyperkalemia and certain muscular dystrophies remains unclear; however, the opportunity for the OR team to use a “clean” non-triggering technique should be explored as well. Children with Duchenne muscular dystrophy, central core disease, and other myopathies are in this category.

All children who have either a presumed or confirmed diagnosis of myopathy should have a preoperative electrocardiogram and chest radiograph to identify a rhythm disturbance or dilated cardiac chambers which might be suggestive of an associated cardiomyopathy. In children with a heart murmur or those less than a year of age, a consultation with a pediatric cardiologist should be sought. Poor neuromuscular function may result in compromised respiratory function. Pulmonary function testing may be helpful in predicting which children will have difficulty with extubation at the conclusion of surgery. If postoperative ventilation is considered, appropriate discussion with the family as well as an arrangement for appropriate postoperative disposition should be made. Children who have a diagnosis of mitochondrial myopathy may have metabolic derangements if NPO status is maintained for a prolonged period of time. These children should be specifically instructed to drink glucose containing clear fluids up to 2 hours prior to the time of surgery.

Oncologic Disease

Children with current or prior malignancy should have their chemotherapy documented. The anthracycline class of drugs may cause myocardial dysfunction and others such as mitomycin and bleomycin may cause pulmonary dysfunction. Children who have received chemotherapy regimens which include anthracycline agents require echocardiographic evaluation if the cumulative dose is greater than 150 mg/m^2. Any child with a history of congestive heart failure, who has not had a post-chemotherapy echocardiogram or who has not had an echocardiogram within 2 years prior to the time of anesthesia irrespective of length of time since completion of therapy, requires a preoperative echocardiogram.

Specific chemotherapeutic agents may cause toxicity and abnormal function in other organ systems including renal, hepatic, and the central nervous system. Specific laboratory testing and imaging should be completed preoperatively when compromised function is suspected.

The Child of a Jehovah’s Witness Parent

The belief of Jehovah’s Witnesses that requires them to refuse transfused blood is based on scriptural passages that define the “life force” as residing in the blood. Many Jehovah’s Witness patients would rather die than receive blood or blood products. Although adult Jehovah’s Witness patients may choose to refuse lifesaving blood transfusions, pediatric patients as minor children do not have that same right. It is, therefore, the responsibility of the anesthesiologist to define a plan with the parents in the event that blood is required.[34]

The anesthesiologist should explore the particular family beliefs of each patient since some Jehovah’s Witness patients will allow the use of blood conservation in an attempt to minimize intraoperative blood loss and transfusion requirement. Perioperative volume expanders (ie, albumin), hemodilution, and blood salvage are acceptable to some individuals depending on their interpretation of biblical passages, while others will not allow their administration.[35]

Most medical personnel are in agreement that in an emergency situation it is unacceptable for a parent to make a conscious decision that could result in the loss of the minor child's life and in such cases appropriate medical therapy, including transfusion or blood and/or blood products are administered against the wishes of the family. In most circumstances, the courts have intervened to allow blood transfusions over the religious objections of the parents. No child of Jehovah's Witness parents should die for lack of transfused blood, as courts have uniformly intervened to allow hospitals to give blood transfusions to minors irrespective of the religious objections of Jehovah's Witness parents. Thus, if a minor requires blood and the parents object, the anesthesiologist may contact the Office of General Counsel to obtain a court order authorizing the blood transfusion. In the event of an emergency, the blood can be administered while the Office of General Counsel is simultaneously being contacted. This process should be disclosed during the preoperative discussion. Additional preoperative preparation includes optimization of hemoglobin with oral iron therapy 2 to 3 weeks prior to surgery.

Autism

Autism is a heterogenous neurodevelopmental disorder that impairs social communication capabilities as well as the way an individual perceives his or her surroundings. Children present early in life with deficits in their information processing skills and their ability to cope with stress. The initial interview provides valuable information regarding patient's behavior, likes and dislikes, favorite activities, triggers for negative behaviors, modeling, and coping techniques. It is widely accepted that collecting information from parents regarding their child's behavioral patterns and needs, and previous anesthetic experiences lead to a more successful anesthetic experience. A connection can be established with some autistic children; however, low-functioning patients present a significant challenge to the anesthesiologist. Establishing good rapport with the family is helpful since gaining parental trust, acknowledging their anxiety and previous experiences, willingness to listen to their concerns, and formulating management plan together can all contribute to a smooth anesthetic course. Collecting pertinent information about the behavioral profile of the patient including baseline behaviors, triggers of emotional outbursts, and signs of escalating anxiety during the preoperative interview is beneficial to the anesthesia team in planning the perioperative anesthetic management. The family should be encouraged to bring favorite toys, electronic devices, and comforting items on the day of the procedure. In addition careful detailed discussion with the surgical team regarding the plan and workflow is important to the design of the perioperative plan.[36]

Immunization

Frequently, children present for general anesthesia shortly after an immunization has been administered. Post-immunization effects include fever, pain at the injection site, malaise, and irritability. These clinical symptoms must not be confused with perioperative complications. The effect of anesthesia on the immune response during elective surgery is minor and usually persists for 48 hours and there is no contraindication to the immunization of healthy children scheduled for elective surgery. However, a delay prior to anesthesia of 2 days for inactivated vaccines such as diphtheria-pertussis-tetanus (DPT) or 14 to 21 days for live attenuated vaccines such as measles-mumps-rubella (MMR) immunization may be prudent.[37] The time interval between immunizations and procedures may be important in preventing misinterpretation of vaccine-driven adverse events as postoperative/postprocedure complications. This includes, but is not limited to, routine scheduled vaccines as well as seasonal and flu vaccine. When a vaccine is administered within 7 days of a scheduled procedure or in the case of unplanned procedures, both the anesthesiologist and surgeon should be informed. Cases should not be automatically cancelled but left to the discretion of the anesthesiologist and/or surgeon based on individual patient considerations.

Concussion

Sport- or recreation-related concussion may occur in the setting of trauma requiring surgical intervention under general anesthesia especially in adolescents. The effect of surgery and general anesthesia on brain recovery, either favorable or unfavorable, in this population is unknown. There is no evidenced-based, clinical pathway describing the optimal timing of surgery and anesthesia for semi-elective surgical procedures following a concussion. The decision to proceed with surgery in patients with known concussion is based on individual clinician's judgment. Elective surgery is often delayed in patients with known concussion until the patient is cleared to "return to play" or "free of symptoms." Rest and avoidance of second or recurrent collisions are essential to the patient's recovery; however, there is no clear clinical definition of "rest" or how much time is needed for full recovery.[38]

Physical Examination

The physical examination of the pediatric patient must begin with simple observation from a distance since the infant or child may become frightened when approached directly. A great deal may be learned about physical findings that relate to the anesthetic without touching the child. The color of the skin including the presence of pallor, cyanosis, rash, jaundice, unusual markings, or prior surgical scars may reveal the presence of organ system dysfunction. Since one congenital abnormality is often associated with others, abnormal facies should be explored as an indication of a syndrome.

The respiratory system should be evaluated by noting the rate and quality of respirations, the presence of noisy breathing, coughing, purulent nasal discharge, stridor, and wheezing. Signs of an acute upper respiratory infection should be evaluated. The ease of mouth opening should be determined as well as the presence of loose teeth.

If a heart murmur is detected on the cardiovascular examination there are specific concerns that must be addressed. An innocent murmur may be due to turbulent blood flow during a growth spurt, whereas a pathologic murmur is usually due to a structural abnormality; this distinction must be made. Lesions in which bacterial endocarditis prophylaxis or protection from paradoxical air embolus are required must be documented.

The patient's neurologic evaluation should include the level of consciousness, presence of an intact gag reflex, and adequate cervical spine movement. General muscle tone and the presence of signs of an increase in intracranial pressure should also be noted.

Diagnostic Testing

It is important to remember that phlebotomy is often traumatic for children and an event that they do not easily forget. For this reason, it is best to limit the number of invasive tests performed. The diagnostic studies should be selected based on the general medical health of the patient and the procedure being performed. In general, measurement of hematocrit in a healthy child undergoing elective surgery is unnecessary. If significant blood loss is anticipated or if the child is less than 6 months of age or was born prematurely a hematocrit should be measured. Neither the routine measurement of the coagulation profile nor a history of "easy bruising" is reliable in predicting surgical bleeding. The presence of prior hematoma, bleeding from circumcision, or large bruises should prompt an investigation; however, a negative history for bruising in an otherwise healthy child would require no further testing. Routine preoperative urinalysis is not indicated in children and serum chemistry analysis should only be performed when an abnormality is suspected. Children who are treated with anticonvulsants should have these medication levels checked and an electrocardiogram or chest radiograph should only be ordered if the general medical condition warrants. Routine pregnancy testing is controversial and the policy of the specific medical facility should be followed.

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