

# THE **HARRIET LANE** HANDBOOK

THE JOHNS HOPKINS HOSPITAL

KEITH **KLEINMAN**  
LAUREN **MCDANIEL**  
MATTHEW **MOLLOY**



TWENTY-SECOND  
EDITION

## PEDIATRIC PARAMETERS AND EQUIPMENT

	Premie	Newborn	6 mo	1 yr	2-3 yr	4-6 yr	7-10 yr	11-15 yr	>16 yr
WT (kg)	2.5-3.5	3.5-4	6-8	10	13-16	20-25	25-35	40-50	>50
BAG VALVE MASK	Infant	Infant	Small child	Small child	Child	Child	Child/small adult	Adult	Adult
NASAL AIRWAY (Fr)	12	12	14-16	14-16	14-18	14-18	16-20	18-22	22-36
ORAL AIRWAY	Infant 50 mm	Small 60 mm	Small 60 mm	Small 60 mm	Small 70 mm	Small 70-80 mm	Med 80-90 mm	Med 90 mm	Med 90 mm
BLADE	MIL 0	MIL 0	MIL 1	MIL 1, MAC 2	MIL 1, MAC 2	MIL 2, MAC 3	MIL 2, MAC 3	MIL 2, MAC 3	MIL 2, MAC 3
ETT	2.5-3.0	3.0-3.5	3.5-4.0	4.0-4.5	4.5-5.0	5.0-5.5	5.5-6.0	6.0-6.5	7.0-8.0
LMA	1	1	1.5	2	2	2.5	2.5-3	3	4
GLIDESCOPE	1	1 or 2	2	2	3	3	3	3 or 4	3 or 4
IV CATH (ga)	22-24	22-24	20-24	20-24	18-22	18-22	18-22	18-20	16-20
CVL (Fr)	3	3-4	4	4-5	4-5	5	5	7	7
NGT/OGT (Fr)	5	5-8	8	10	10-12	12-14	12-14	14-18	14-18
CHEST TUBE (Fr)	10-12	10-12	12-18	16-20	16-24	20-28	20-32	28-38	28-42
FOLEY (Fr)	6	8	8	8	8	8	8	10	12

## ESTIMATED BLOOD PRESSURE BY AGE

Measurement	50th %	5th %
Systolic BP	90 + (age × 2)	60 (neonate); 70 (1 mo-1 yr) 70 + (age × 2) (for 2-10 yr) <90 (>10 yr)
MAP	55 + (age × 1.5)	40 + (age × 1.5)

## NORMAL VITAL SIGNS BY AGE

Age	Heart Rate (beats/min)	Blood Pressure (mmHg)	Respiratory Rate (breaths/min)
Premie	120-170	55-75/35-45 (gestational age approximates normal MAP)	40-70
0-3 mo	110-160	65-85/45-55	30-60
3-6 mo	100-150	70-90/50-65	30-45
6-12 mo	90-130	80-100/55-65	25-40
1-3 yr	80-125	90-105/55-70	20-30
3-6 yr	70-115	95-110/60-75	20-25
6-12 yr	60-100	100-120/60-75	14-22
>12 yr	60-100	100-120/70-80	12-18

## ENDOTRACHEAL TUBE FORMULAS

Uncuffed ETT size: age (years)/4 + 4; Cuffed ETT size: age (years)/4 + 3

ETT depth (from lip to mid-trachea): ETT internal diameter (size) × 3

## GLASGOW COMA SCALE

Activity	Score	Child/Adult	Score	Infant
Eye opening	4	Spontaneous	4	Spontaneous
	3	To speech	3	To speech/sound
	2	To pain	2	To painful stimuli
	1	None	1	None
Verbal	5	Oriented	5	Coos/babbles
	4	Confused	4	Irritable cry
	3	Inappropriate	3	Cries to pain
	2	Incomprehensible	2	Moans to pain
	1	None	1	None
Motor	6	Obey commands	6	Normal spontaneous movement
	5	Localizes to pain	5	Withdraws to touch
	4	Withdraws to pain	4	Withdraws to pain
	3	Abnormal flexion	3	Abnormal flexion (decorticate)
	2	Abnormal extension	2	Abnormal extension (decerebrate)
	1	None	1	None (flaccid)

Adapted from Hunt EA, Nelson-McMillan K, McNamara L. The Johns Hopkins Children's Center Kids Card, 2016.

## RESUSCITATION MEDICATIONS

<b>Adenosine</b>	<b>0.1 mg/kg IV/IO RAPID BOLUS (over 1-2 sec), Flush with 10 mL normal saline</b>
Supraventricular tachycardia	May repeat at 0.2 mg/kg IV/IO, then 0.3 mg/kg IV/IO after 2 min Max first dose 6 mg, max subsequent dose 12 mg Administer using a 3-way stopcock attached to a 10 ml NS flush
<b>Amiodarone</b>	<b>5 mg/kg IV/IO</b>
Ventricular tachycardia	No Pulse: Push Undiluted Pulse: Dilute and give over 20-60 minutes Max first dose 300 mg, max subsequent dose 150 mg Only give max of 3 IV push doses Monitor for hypotension Strongly consider pretreating with IV calcium in patients with a pulse to prevent hypotension
<b>Atropine</b>	<b>0.02 mg/kg IV/IO/IM, 0.04–0.06 mg/kg ETT</b>
Bradycardia (increased vagal tone)	Max single dose 0.5 mg Repeat in 5 minutes if needed (up to twice) to max total dose 1 mg
Primary AV block	
<b>Calcium chloride (10%)</b>	<b>20 mg/kg IV/IO</b>
Hypocalcemia	Max dose 1 gram
<b>Calcium Gluconate (10%)</b>	<b>60 mg/kg IV/IO</b>
	Max dose 3 grams
<b>Dextrose</b>	<b>Weight-Based Dosing: 0.5–1 gram/kg</b> <b>Volume-Based Dosing (“Rule of 50”):</b> <5 kg: 10% dextrose 5-10 mL/kg IV/IO 5-44 kg: 25% dextrose 2-4 mL/kg IV/IO ≥45 kg: 50% dextrose 1-2 mL/kg IV/IO Max single dose 50 grams = 100 mL
<b>Epinephrine</b>	<b>0.01 mg/kg IV/IO every 3–5 min (max single dose 1 mg)</b>
Pulseless arrest	<b>0.1 mg/kg ETT every 3–5 min (max single dose 2.5 mg)</b>
Bradycardia (symptomatic)	<b>Anaphylaxis:</b> <b>0.01 mg/kg IM (1 mg/mL) in thigh every 5-15 min PRN; max single dose 0.5 mg</b>
Anaphylaxis	<b>Standardized/Autoinjector:</b> <7.5 kg: no autoinjector, see above 7.5 to <15 kg: 0.1 mg IM 15 to <30 kg: 0.15 mg IM ≥30 kg: 0.3 mg IM
<b>Hydrocortisone</b>	<b>2 mg/kg IV/IM/IO</b>
Adrenal Crisis/Insufficiency	Max dose 100 mg
<b>Insulin (Regular or Aspart)</b>	<b>0.1 units/kg IV/IO with 0.5 gram/kg of dextrose</b>
	Max dose 10 units
<b>Hyperkalemia</b>	
<b>Lidocaine</b>	<b>1 mg/kg IV/IO (ETT dose is 2-3x IV dose)</b>
Antiarrhythmic	Max single dose 100 mg May repeat in 10-15 min x2
<b>Magnesium sulfate</b>	<b>50 mg/kg IV/IO</b>
Torsades de pointes	No Pulse: Push
Hypomagnesemia	Pulse: Give over 20-60 minutes Max single dose 2 grams Monitor for hypotension/bradycardia
<b>Naloxone</b>	<b>Respiratory Depression: 0.001–0.005 mg/kg/dose IV/IO/IM/Subcut (max 0.1 mg first dose, may titrate to effect)</b>
Opioid overdose	<b>Full Reversal/Arrest Dose: 0.1 mg/kg IV/IO/IM/Subcut (max dose 2 mg)</b>
Coma	ETT dose 2–3 times IV dose, IN dose 2–4 mg. May give every 2 min PRN
<b>Sodium Bicarbonate (8.4% = 1 mEq/mL)</b>	<b>1 mEq/kg IV/IO</b>
Administer only with clear indication:	Dilute 8.4% sodium bicarbonate 1 : 1 with sterile water for patients <10 kg to a final concentration of 4.2% = 0.5 mEq/mL
Metabolic acidosis	Hyperkalemia: Max single dose 50 mEq
Hyperkalemia	
Tricyclic antidepressant overdose	

ETT Meds (NAVEL: naloxone, atropine, vasopressin, epinephrine, lidocaine)—dilute meds to 5 mL with NS, follow with positive-pressure ventilation.

Adapted from Hunt EA, Nelson-McMillan K, McNamara L. *The Johns Hopkins Children's Center Kids Kard, 2018 and the American Heart Association, PALS Pocket Card, 2015*.

$$\text{IV INFUSIONS* } 6 \times \frac{\text{Desired dose (mCg/kg/min)}}{\text{Desired rate (mL/hr)}} \times \text{Wt (kg)} = \frac{\text{mg drug}}{100 \text{ mL fluid}}$$

Medication	Dose (mCg/kg/min)	Dilution in 100 mL in a Compatible IV Fluid	IV Infusion Rate
Alprostadil (prostaglandin E <sub>1</sub> )	0.05–0.1	0.3 mg/kg	1 mL/hr = 0.05 mCg/ kg/min
Amiodarone	5–15	6 mg/kg	1 mL/hr = 1 mCg/kg/min
DOPamine	5–20	6 mg/kg	1 mL/hr = 1 mCg/kg/min
DOBUTamine	2–20	6 mg/kg	1 mL/hr = 1 mCg/kg/min
EPINEPHrine	0.1–1	0.6 mg/kg	1 mL/hr = 0.1 mCg/ kg/min
Lidocaine, post resuscitation	20–50	6 mg/kg	1 mL/hr = 1 mCg/kg/min
Phenylephrine	0.1–2, up to 5 in severe circumstances	0.3 mg/kg	1 mL/hr = 0.05 mCg/ kg/min
Terbutaline	0.1–4 (up to 10 has been used)	0.6 mg/kg	1 mL/hr = 0.1 mCg/ kg/min
Vasopressin (pressor)	0.17–8 millionunits/kg/min	6 units/kg	1 mL/hr = 1 millionunit/ kg/min

\*Standardized concentrations are recommended when available. For additional information, see Larsen GY, Park HB et al. Standard drug concentrations and smart-pump technology reduce continuous-medication-infusion errors in pediatric patients. Pediatrics. 2005;116(1):e21–e25.

Special thanks to Lisa Hutchins, Clinical Pharmacy Specialist, for her expert guidance with IV infusion and resuscitation medication guidelines.

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TWENTY-SECOND  
EDITION

A MANUAL FOR PEDIATRIC HOUSE OFFICERS

# THE **HARRIET LANE HANDBOOK**

THE HARRIET LANE HOUSE STAFF AT  
THE CHARLOTTE R. BLOOMBERG CHILDREN'S CENTER OF  
THE JOHNS HOPKINS HOSPITAL

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### **To our families**

Michael and Debbie Kleinman, you have always been there for guidance and support and have allowed me to follow my dreams. Mary Buckley Kleinman, thank you for being such a loving and devoted wife; you push me to be better every day. Dr. Kimberly Erica Kleinman, you are such a wonderful sister whom I have always looked up to. Camper Whitney Kleinman, you are beautiful in every way. Ina Zun, you were the perfect grandmother and the reason that I am a doctor; I miss you every day.

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From an early age, you instilled in me a love of books, a passion for medicine, and an unwavering belief that with hard work and a sense of humor, anything is possible. Michael McDaniel, thank you for being the best brother I could ever ask for and for always believing in and supporting me.

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### **To our patients and their families**

We will never forget the lessons you have taught us and the trust you place in us.

### **To our residents**

We are inspired by your brilliance, boldness, and dedication to caring for children.

### **To the wonderful pediatricians and educators who trained us**

Especially Nicole Shilkofski, Janet Serwint, George Dover, Tina Cheng

**In loving memory of Dr. Michael Burke**

# Preface

*“Why this child? Why this disease? Why now?”*

—Barton Childs, MD

*The Harriet Lane Handbook* was first developed in 1953 after Harrison Spencer (chief resident in 1950–1951) suggested that residents should write a pocket-sized “pearl book.” As recounted by Henry Seidel, the first editor of *The Harriet Lane Handbook*, “Six of us began without funds and without [the] supervision of our elders, meeting sporadically around a table in the library of the Harriet Lane Home.” The product of their efforts was a concise yet comprehensive handbook that became an indispensable tool for the residents of the Harriet Lane Home. Ultimately, Robert Cooke (department chief, 1956–1974) realized the potential of the Handbook, and, with his backing, the fifth edition was published for widespread distribution by Year Book. Since that time, the handbook has been regularly updated and rigorously revised to reflect the most up-to-date information and available clinical guidelines. It has grown from a humble Hopkins resident “pearl book” to become a nationally and internationally respected clinical resource. Now translated into many languages, the handbook is still intended as an easy-to-use manual to help pediatricians provide current and comprehensive pediatric care.

Today, *The Harriet Lane Handbook* continues to be updated and revised *by* house officers *for* house officers. Recognizing the limit to what can be included in a pocket guide, additional information has been placed online and for use via mobile applications. The online-only content includes references, expanded text, and additional tables, figures, and images.

In addition to including the most up-to-date guidelines, practice parameters, and references, we will highlight some of the most important improvements in the twenty-second edition of *The Harriet Lane Handbook*:

The Emergency Management and Trauma, Burn, and Common Critical Care Emergencies chapters have been reorganized. The **Emergency and Critical Care Management** chapter now focuses on the medical management of common critical care emergencies, while the management of trauma, including burns, has been consolidated into the **Traumatic Injuries** chapter.

The Development, Behavior, and Mental Health chapter has been separated into two chapters with expanded content: **Behavior, Development, and Developmental Disability** and **Psychiatry**, reflecting the growing need for pediatricians to understand mental and behavioral health.

The **Genetics** chapter has been reorganized to present categories of metabolic disease in easily referenced tables and to provide an organization to different patterns and etiologies of dysmorphology.

The **Hematology** chapter has been restructured with much of the text re-organized and expanded into tables and figures, including a new algorithmic approach to anemia. Content on the management of transfusion reactions has been added.

The **Immunoprophylaxis** chapter includes a new section on vaccine hesitancy.

The **Nutrition and Growth** chapter now includes expanded content on the management of overweight and obese children, definitions of various degrees of malnutrition, information on refeeding syndrome, and a table with instructions on the preparation of fortified formula. Enteral formulas have been reorganized based on clinical indications.

The **Radiology** chapter has been reorganized with all-new images and more focused content.

The **Rheumatology** chapter has been refocused for the general pediatrician and includes a section on the primary care management of rheumatologic diseases.

*The Harriet Lane Handbook*, designed for pediatric house staff, was made possible by the extraordinary efforts of this year's Johns Hopkins Harriet Lane Pediatric Residency Program senior resident class. It has been an honor to watch these fine doctors mature and refine their skills since internship. They have balanced their busy work schedules and personal lives while authoring the chapters that follow. We are grateful to each of them, along with their faculty advisors, who selflessly dedicated their time to improve the quality and content of this publication. The high quality of this handbook is representative of our residents, who are the heart and soul of our department.

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The Formulary is complete, concise, and up-to-date thanks to the tireless efforts of Carlton K.K. Lee, PharmD, MPH. With each edition, he carefully updates, revises, and improves the section. His herculean efforts make the Formulary one of the most useful and cited pediatric drug reference texts available.

We are grateful and humbled to have the opportunity to build on the great work of the preceding editors: Drs. Henry Seidel, Harrison Spencer, William Friedman, Robert Haslam, Jerry Winkelstein, Herbert Swick, Dennis Headings, Kenneth Schuberth, Basil Zitelli, Jeffery Biller, Andrew Yeager, Cynthia Cole, Peter Rowe, Mary Greene, Kevin Johnson, Michael Barone, George Siberry, Robert Iannone, Veronica Gunn, Christian Nechyba, Jason Robertson, Nicole Shilkofski, Jason Custer, Rachel Rau, Megan Tschudy, Kristin Arcara, Jamie Flerlage, Branden Engorn, Helen Hughes, and Lauren Kahl. Many of these previous editors continue to make important contributions to the education of the Harriet Lane house staff, none more than Dr. Nicole Shilkofski, our current residency program director. We are constantly impressed by her enthusiasm for education and her advocacy for the residents. As recent editors, Drs. Helen Hughes and Lauren Kahl also have been instrumental in helping us to navigate this process. We hope to live up to the legacy of these many outstanding clinicians, educators, and mentors.

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# Contents

## PART I Pediatric Acute Care

- 1 Emergency and Critical Care Management, 3  
*Kelsey Stayer, MD and Lisa Hutchins, PharmD*
- 2 Traumatic Injuries, 33  
*Nymisha Chilukuri, MD*
- 3 Toxicology, 52  
*Maria D. Latham, MD*
- 4 Procedures, 61  
*Andrew Percy, MD*

## PART II Diagnostic and Therapeutic Information

- 5 Adolescent Medicine, 101  
*Christine Krueger, MD and Harita Shah, MD*
- 6 Analgesia and Procedural Sedation, 126  
*Courtney Altshuler, MD and Kelsey Gladen, MD*
- 7 Cardiology, 145  
*Aoibhinn Nyhan, MD*
- 8 Dermatology, 189  
*Jennifer Reed DiBiagio, MD and M. Cooper Lloyd, MD, MPH*
- 9 Development, Behavior, and Developmental Disability, 211  
*Brittany Badesch, MD*
- 10 Endocrinology, 228  
*Samar Atteih, MD and Jessica Ratner, MD*
- 11 Fluids and Electrolytes, 261  
*Lauren Burgunder, MD*
- 12 Gastroenterology, 283  
*Matthew Buendia, MD and Natalie Thoni, MD*
- 13 Genetics: Metabolism and Dysmorphology, 300  
*Jasmine Knoll, MD, RaeLynn Forsyth, MD and Sarah Pryor, MD, MPH*
- 14 Hematology, 328  
*Jessica Calihan, MD*
- 15 Immunology and Allergy, 368  
*Carlos A. Salgado, MD*
- 16 Immunoprophylaxis, 382  
*Xiao P. Peng, MD, PhD*
- 17 Microbiology and Infectious Disease, 408  
*Kevin Klembczyk, MD and Samuel McAleese, MD*

- 18**    Neonatology, 447  
*Niana Carter, MD and Bethany Sharpless Chalk, PharmD*
- 19**    Nephrology, 472  
*Paul M. Gallo, MD, PhD*
- 20**    Neurology, 502  
*Ania K. Dabrowski, MD and Lindsay Schleifer, MD*
- 21**    Nutrition and Growth, 523  
*Jaime La Charite, MD, MP*
- 22**    Oncology, 546  
*P. Galen DiDomizio, MD and Chana Richter, MD*
- 23**    Palliative Care, 566  
*Joshua Natbony, MD*
- 24**    Psychiatry, 574  
*Christopher Morrow, MD*
- 25**    Pulmonology and Sleep Medicine, 586  
*Stephanie Tung, MD, MSc*
- 26**    Radiology, 606  
*Brittany Hunter, MD*
- 27**    Rheumatology, 627  
*Shani Jones, MD*

### **PART III Reference**

- 28**    Blood Chemistry and Body Fluids, 641  
*Lauren McDaniel, MD*
- 29**    Biostatistics and Evidence-Based Medicine, 653  
*Matthew Molloy, MD, MPH*

### **PART IV Formulary**

- 30**    Drug Dosages, 665  
*Carlton K.K. Lee, PharmD, MPH*
- 31**    Drugs in Renal Failure, 1075  
*Elizabeth A.S. Goswami, PharmD and Namrata Trivedi, PharmD*

# Chapter 1

## Emergency and Critical Care Management

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This chapter is presented in accordance with the universal acronym **C-A-B** (circulation, airway, breathing) to emphasize the reduction of “no blood flow” time.<sup>1-3</sup> However, given the high prevalence of asphyxial cardiac arrest in the pediatric population, ventilation remains fundamental to the resuscitation of the critically ill child.<sup>4</sup> This chapter serves to function as a guide to caring for “sick” children, spanning the principles of resuscitation and stabilization, as well as management of the most common pediatric medical emergencies.

### I. APPROACH TO THE UNRESPONSIVE CHILD

#### A. Circulation<sup>1-3,5-10</sup>

##### 1. Assessment

- a. **Pulse:** Spend no more than **10 seconds** assessing pulse. Assess brachial pulse in infants, carotid or femoral pulse in children.
- b. **Perfusion:** Check for pallor, mottling, or cyanosis. Capillary refill time  $>2$  seconds is delayed and  $<1$  to 2 seconds or “flash” may indicate warm shock.
- c. **Rate:** Bradycardia **<60 beats/min** with **poor perfusion** requires immediate cardiopulmonary resuscitation (CPR). Tachycardia **>220 beats/min** suggests pathologic tachyarrhythmia.
- d. **Rhythm:** Attach patient to defibrillator or continuous electrocardiography. In arrest, check rhythm every 2 minutes with minimal interruptions in chest compressions (e.g., during compressor change).
- e. **Blood pressure (BP):** Hypotension in a pediatric patient is a **late** manifestation of circulatory compromise.
- f. **Urine output:** Normal output is 1.5 to 2 mL/kg/h in infants and young children and 1 mL/kg/h in older children.

##### 2. Management: Initiate CPR immediately if patient is pulseless or bradycardic ( $<60$ beats/min) with poor perfusion.

- a. **Chest compressions:** See **Box 1.1** for an outline of the five components of **high-quality CPR**.
- b. **Monitoring:** Continuous capnography and invasive hemodynamic monitoring may guide effectiveness of chest compressions.
  - (1) Target **end-tidal CO<sub>2</sub>** (EtCO<sub>2</sub>) **>20 mmHg**. If consistently less than, improve compressions and assess for excessive ventilation.
  - (2) Abrupt and sustained rise in EtCO<sub>2</sub> is often observed just prior to clinical return of spontaneous circulation (ROSC).

**BOX 1.1****FIVE COMPONENTS OF HIGH-QUALITY CARDIOPULMONARY RESUSCITATION**

- “Push fast”: Target rate of **100–120 compressions/min**
- “Push hard”: Target depth of **at least  $\frac{1}{2}$  anteroposterior diameter of chest**
  - Place step stool at side of bed to assist compressor
  - Slide backboard under patient or place on hard surface
  - Use the compression technique that achieves the best results
    - Consider two-handed, one-handed, two-finger, or two-thumb-encircling hands techniques
  - Aim for 1 fingerbreadth below intermammary line in infants, 2 fingerbreadths in prepubertal children, and the lower half of the sternum in adolescents
- Allow full chest recoil between compressions
- **Minimize interruptions** in chest compressions
  - Rotate compressor every 2 min or sooner if fatigued
  - Check cardiac rhythm at time of compressor change
- Avoid excessive ventilation
  - If no advanced airway (endotracheal tube, laryngeal mask airway, tracheostomy) secured, perform **30:2 compression-ventilation ratio** (with single rescuer or for any adolescent/adult) or **15:2 ratio** (in an infant/child only if 2 rescuers present)
  - If advanced airway secured, give **one breath every 6–8 sec** with continuous compressions
  - Ventilation volume should produce no more than minimal, visible chest rise

(3) If a patient has an indwelling arterial catheter, assess waveform for feedback to evaluate chest compressions. Target **diastolic BP >25 mmHg** in infants and **>30 mmHg** in children.

- c. **Defibrillation:** Shockable arrest rhythms include **ventricular fibrillation** and **pulseless ventricular tachycardia**. Nonshockable arrest rhythms include asystole, pulseless electrical activity, and bradycardia with poor perfusion.
- (1) Use age- and size-appropriate pads as recommended per manufacturer.
  - (2) Initial shock dose is **2 J/kg**, second dose is **4 J/kg**, subsequent doses are **≥4 J/kg (maximum 10 J/kg or adult maximum dose)**.
- d. **Cardioversion:** A synchronized electrical shock delivered for hemodynamically **unstable** patients with **tachyarrhythmias** (i.e., supraventricular tachycardia, atrial flutter, ventricular tachycardia) and **palpable pulses**.
- (1) Initial dose is **0.5 to 1 J/kg**. Increase to **2 J/kg** if ineffective, repeating doses if necessary. Reevaluate diagnosis if rhythm does not convert to sinus.
  - (2) Consultation with a pediatric cardiologist is recommended for elective cardioversion for stable patients with tachyarrhythmias.
- e. **Resuscitation**
- (1) **Access:** Place intraosseous access immediately if in arrest or if intravenous (IV) access difficult.

- (a) If previously established, central access is preferred for drug administration.
- (b) Endotracheal tube (ETT) drug administration is acceptable if required. Lidocaine, epinephrine, atropine, and naloxone (LEAN) and vasopressin can be administered via endotracheal route.
- (2) **Pharmacotherapy:** See [Table 1.1](#) detailing medications for pediatric resuscitation. If actual body weight is unavailable, use length-based habitus-modified (e.g., Mercy method, PAWPER tape) estimation methods, parental estimates, or length-based (e.g., Broselow tape) estimation methods, in order of accuracy.
- (3) **Fluids:** Administer isotonic crystalloid for treatment of shock even if BP is normal.
- (a) Administer up to 60 mL/kg of isotonic crystalloid in 20 mL/kg increments in non-neonates during the first 20 minutes until perfusion improves. Frequently reassess for hepatomegaly, pulmonary crackles, and respiratory distress.
- (b) Special consideration for **cardiogenic shock:**
- (i) Administer an initial fluid bolus of 5 to 10 mL/kg over 10 to 20 minutes if cardiac insufficiency suspected or unknown (consider in neonate).
  - (ii) Be prepared to support oxygenation and ventilation in case of pulmonary edema.
- (c) Special consideration for **septic shock:** Specific goals of therapy include  $\text{ScvO}_2$  (central venous saturation)  $\geq 70\%$ , adequate BP, normalized heart rate (HR), and appropriate end-organ perfusion.
- f. **Extracorporeal-CPR (E-CPR):** Rapid deployment of venoarterial (VA) or venovenous (VV) extracorporeal membrane oxygenation (ECMO) to artificially provide oxygenation, ventilation, and circulation as a means of CPR for in-hospital arrest refractory to conventional interventions. Contraindications are limited but may include extremes of prematurity or low birth weight, lethal chromosomal abnormalities, uncontrollable hemorrhage, or irreversible brain damage. Should not be offered if likely to be futile.

1

## B. Airway and Breathing<sup>1,7,11-17</sup>

1. Assessment
  - a. **Airway patency:** Perform head tilt and chin lift or jaw thrust to open airway. Avoid overextension in infants.
  - b. **Spontaneous respirations:** Assess spontaneous patient effort.
    - (1) If breathing regularly, place patient in **recovery position** (turn onto side).
    - (2) If the patient has a palpable pulse but inadequate breathing, **provide a 1-second breath every 3 to 5 seconds.**
  - c. **Adequacy of respiration:** Evaluate for symmetric chest rise. Auscultate for equal breath sounds with good aeration.

**TABLE 1.1****PEDIATRIC RESUSCITATION MEDICATIONS<sup>5,7,11,17</sup>**

Medication	Indication	Dosing	Mechanism	Side Effects
Adenosine	SVT secondary to AV node reentry or accessory pathways	Initial: 0.1 mg/kg IV (max 6 mg) Sec: 0.2 mg/kg IV (max 12 mg) Third: 0.3 mg/kg IV (max 12 mg) Wait 2 min between doses Administer with three-way stopcock rapid push/flush technique	Purine nucleoside blocks AV node conduction	Brief period of asystole (10–15 sec)
Amiodarone	Shock-refractory VF or pVF, stable SVT, unstable VT	5 mg/kg (max 300 mg) IV/IO No pulse: Push undiluted dose Pulse: Dilute and run over 20–60 min Repeat dosing: 5 mg/kg (up to max 150 mg) up to 15 mg/kg total Infusion: 5–15 mCg/kg/min (max 20 mg/kg/day or 2200 mg/day)	Potassium-channel blockade suppresses AV node, prolongs QT and QRS	Risk of polymorphic VT, hypotension, decreased cardiac contractility
Atropine	Bradycardia from increased vagal tone, cholinergic drug toxicity, second- and third-degree AV block	0.02 mg/kg IV/IO/IM (min 0.1 mg/dose, max 0.5 mg/dose; larger doses may be needed for organophosphate poisoning) or 0.04–0.06 mg/kg ET Repeat dosing: may repeat once after 5 min	Cholinergic blockade accelerates atrial pacemakers, enhances AV conduction	Tachycardia, risk of myocardial ischemia, paradoxical bradycardia with lower than minimal dosing
Calcium chloride	Hypocalcemia, hyperkalemia, hypermagnesemia, calcium channel blocker overdose	20 mg/kg (max 1 g) IV/IO Administer over 10–20 sec in arrest Consider calcium gluconate in nonarrest if access is peripheral only	Binds myocardial troponin to increase cardiac contractility	Risk of myocardial necrosis
Dextrose	Documented hypoglycemia	0.5–1 g/kg IV/IO Newborn: 5–10 mL/kg D <sub>10</sub> W Infants, children: 2–4 mL/kg D <sub>25</sub> W Adolescents: 1–2 mL/kg D <sub>50</sub> W	Restores energy metabolite	Risk of poor neurologic outcomes in setting of hyperglycemia

Epinephrine	Asystole, PEA, VT, pVT, diastolic hypotension, bradycardia	Bolus: 0.01 mg/kg IV/IO (0.1 mg/mL; max 1 mg) or 0.1 mg/kg ET (1 mg/mL; max 2.5 mg) Repeat dosing: Bolus every 3–5 min as needed Infusion: 0.1–1 mCg/kg/min	$\alpha$ -Agonism increases heart rate and cardiac contractility	Tachycardia, ectopy, tachyarrhythmias, hypertension
Lidocaine	Shock-refractory VF or pVT (second-line after amiodarone)	Bolus: 1 mg/kg (max 100 mg) IV/IO, 2–3 mg/kg ET Repeat dosing: 1 mg/kg (max 100 mg) every 10–15 min up to 3–5 mg/kg in first hr Infusion: 20–50 mCg/kg/min	Sodium-channel blockade shortens the duration of the action potential	Myocardial depression, altered mental status, seizures, muscle twitching
Magnesium sulfate	Torsades de pointes, hypomagnesemia	50 mg/kg (max 2 g) IV/IO No pulse: Push dose Pulse: Run over 20–60 min	Calcium antagonist depresses abnormal secondary depolarizations and AV node conduction	Hypotension, bradycardia
Naloxone	Opioid overdose	Full reversal: 0.1 mg/kg/dose (max 2 mg/dose) IV/IO/IM, 0.2 mg/kg to 1 mg/kg/dose ET, or 2–4 mg IN Repeat dosing: every 2–3 min as needed	Opioid antagonist reverses opioid-induced respiratory depression, sedation, analgesia and hypotension	Rapid withdrawal, agitation, pain, pulmonary edema
Procainamide	Stable SVT, atrial flutter, atrial fibrillation, VT	Load: 15 mg/kg IV/IO, run over 30–60 min Infusion: 20–80 mCg/kg/min (Max 2 g/24 hr)	Sodium-channel blockade prolongs effective refractory period, depresses conduction velocity	Proarrhythmic, polymorphic VT, hypotension
Sodium bicarbonate	Routine use in arrest is <b>not</b> recommended; hyperkalemia, arrhythmias in tricyclic overdose	1 mEq/kg IV/IO Hyperkalemia: Max single dose 50 mEq	Buffers acidosis by binding hydrogen ions to improve myocardial function, reduce SVR and inhibit defibrillation	May impair tissue oxygen delivery, hypokalemia, hypocalcemia, hypernatremia, impaired cardiac function

AV, Atrioventricular; D<sub>10</sub>W, dextrose 10% in water; ET, endotracheal; HR, heart rate; ICP, intracranial pressure; IM, intramuscular; IO, intraosseous; IV, intravenous; IN, intranasal; mCg, microgram; PEA, pulseless electrical activity; pVF, pulseless ventricular fibrillation; pVT, pulseless ventricular tachycardia; SVR, systemic vascular resistance; SVT, supraventricular tachycardia; VF, ventricular fibrillation; VT, ventricular tachycardia.

- d. **Distress:** Recognize tachypnea, grunting, flaring, retractions, stridor, or wheeze. Infants may exhibit head bobbing.
2. Securing airway
    - a. **Bag-mask ventilation** (BVM): May be used indefinitely if ventilating effectively.
      - (1) Avoid pushing mask down, which can obstruct airway. Bring face into mask.
      - (2) Consider **oropharyngeal** airway in the **unconscious** patient with obstruction. Correct size will extend from corner of mouth to mandibular angle.
      - (3) Consider **nasopharyngeal** airway in the **conscious** (gag reflex intact) or unconscious patient with obstruction. Correct size will extend from tip of nose to tragus of ear.
      - (4) Cricoid pressure (Sellick maneuver) may be used to minimize gastric inflation and aspiration. Avoid excess pressure leading to tracheal obstruction.
    - b. **Laryngeal mask airway** (LMA): Supraglottic airway placed blindly. Useful to emergently secure access to a difficult airway.
      - (1) Use manufacturer-specific weight-based mask size estimation systems or the combined width of the patient's index, middle, and ring fingers to estimate mask size.
      - (2) Continuous chest compressions can be performed once LMA is placed.
    - c. **Endotracheal intubation:** Rapid sequence intubation is indicated in patients presenting with (presumed) full stomach. Immediately sequential sedation and neuromuscular blockade help to avert the need for positive pressure ventilation, minimizing aspiration risk.
      - (1) **Preparation:** Always have a secondary plan to manage the airway if intubation is unsuccessful.
        - (a) **Preoxygenation:** Deliver 100% oxygen via a nonrebreather mask for at least 3 minutes. Children have higher oxygen consumption than adults and can rapidly become hypoxicemic.
        - (b) **Equipment:** Collect monitoring, suctioning, and oxygen delivery equipment.
          - (i) If available, quantitative **EtCO<sub>2</sub>** is recommended as primary method to confirm ventilation.
          - (ii) Place suction catheter at head of bed. Set suction device from **-80 mmHg to -120 mmHg**.
          - (iii) Consider nasogastric tube for stomach decompression. See Chapter 4 for placement.
        - (c) **Airway supplies:** Both cuffed and uncuffed ETTs are acceptable. Cuffed tubes may decrease risk of aspiration.
          - (i) If available, use a length-based estimator (e.g., Broselow tape) of ETT size and laryngoscope blade size.
          - (ii) To estimate age-based ETT size (internal diameter) for patients 2 to 10 years:

Cuffed ETT (mm) = (age in years / 4) + 3.5

Uncuffed ETT (mm) = (age in years / 4) + 4.0

- (iii) To approximate depth of insertion:

Depth (mm) = ETT size (mm) × 3

- (iv) Choose laryngoscope blade type and size based on patient age and airway.

- (v) Straight (i.e., Miller) blades are typically reserved for children <2 years age or difficult airways.

[1] Miller #00-1 for premature to 2 months age

[2] Miller #1 for 3 months to 3 years age

[3] Miller #2 for >3 years age

- (vi) Curved (i.e., Mac) laryngoscope blades are often more effective for children >2 years age.

[1] Mac #2 for >2 years age

[2] Mac #3 for >8 years age

- (d) **Pharmacology:** See Table 1.2 for rapid sequence intubation medications.

- (e) **Positioning:** Place patient in “sniffing” position with neck slightly extended to align the airway.

- (i) Infants and toddlers may require towel roll beneath **shoulders** due to large occiput.

- (ii) Children and adolescents may require towel roll beneath **neck**.

- (2) **Procedure:** Advanced airways should be placed by experienced healthcare providers with appropriate training.

- (a) Confirm placement by detecting EtCO<sub>2</sub>, observing chest wall movement, auscultating for symmetric breath sounds, and monitoring oxygen saturation. Evaluate placement via chest radiograph.

- (3) **Failure:** Acute respiratory failure in an intubated patient may signify **D**isplacement of the ETT, **O**bstruction, **P**neumothorax, or **E**quipment failure (**DOPE**).

- d. **Surgical airway:** Consider needle or surgical cricothyrotomy if BVM, endotracheal intubation, and LMA fail. If available, consult emergently with difficult airway specialists (pediatric anesthesiologist, intensivist, and/or otolaryngologist).

### 3. Oxygenation and Ventilation

- a. Oxygen delivery systems:

- (1) Low-flow systems (e.g., nasal cannula, simple face mask) **do not meet** the inspiratory flow demand of the patient. Delivery of set fraction of inspired oxygen (FiO<sub>2</sub>) is difficult due to room air mixing.
- (2) High-flow systems (e.g., nonrebreather, oxygen hood) **do meet** the inspiratory flow demand of the patient. Measurable FiO<sub>2</sub> is delivered.

**TABLE 1.2****RAPID SEQUENCE INTUBATION MEDICATIONS<sup>11,15-17,20</sup>**

Medication	Benefit	Indication	Dosing	Side Effects
<b>1. Adjuncts</b>				
Atropine	Prevent bradycardia associated with laryngoscope insertion, decrease oral secretions	Bradycardia in any patient, infants <1 year, children 1–5 years receiving succinylcholine, children >5 years receiving a second dose of succinylcholine	0.02 mg/kg IV/IO/IM (max 0.5 mg)	Tachycardia, pupil dilation
Glycopyrrolate	Decrease oral secretions, may cause less tachycardia than atropine, preserves pupillary exam in trauma	Hypersalivation	0.004–0.01 mg/kg IV/IM/IO (max 0.1 mg)	Tachycardia
Lidocaine	Blunts rise in ICP associated with laryngoscopy	Elevated ICP, shock, arrhythmia, and status asthmaticus	1 mg/kg IV/IO (max 100 mg)	Myocardial depression, altered mental status, seizures, muscle twitching
<b>2. Induction Agents</b>				
Etomidate (sedative)	Minimal cardiovascular side effects, minimally decreases ICP	Multitrauma patient at risk for increased ICP and hypotension Caution in patients with adrenal suppression; avoid in septic shock	0.3 mg/kg IV/IO	Suppresses adrenal corticosteroid synthesis, vomiting, myoclonus, lowers seizure threshold
Fentanyl (analgesic, sedative)	Minimal cardiovascular effect	Shock	1–5 mCg/kg slow IV/IM push (max 100 mCg)	Chest wall rigidity, bradycardia, respiratory depression
Ketamine (sedative, analgesic)	Catecholamine release causes bronchodilation, abates bradycardia associated with laryngoscope insertion, increases HR and SVR, produces a “dissociative amnesia”	Status asthmaticus, shock and hypotensive patients Caution in patients at risk for elevated ICP or glaucoma history	1–2 mg/kg IV/IO (max 150 mg) 4–6 mg/kg IM	Vomiting, laryngospasm, hypersalivation, emergence reactions (hallucinations)

Midazolam (sedative, amnestic, anxiolytic)	Minimal cardiovascular effect	Mild shock	0.05–0.3 mg/kg IV/IM/IO (max 10 mg)	Dose-dependent respiratory depression, hypotension
Propofol (sedative)	Ultra-short acting	Role in RSI unclear Avoid in shock or patients who require maintenance of CPP	1 mg/kg IV initial bolus, then 0.5 mg/kg boluses every 3 min as needed	Hypotension, myocardial depression, metabolic acidosis; may cause paradoxical hypertension in children
<b>3. Neuromuscular blockade</b>				
Succinylcholine (depolarizing)	Shortest acting neuromuscular blockade agent, reversible with acetylcholinesterase inhibitor	Role limited due to adverse events Contraindicated in neuromuscular disease, myopathies, spinal cord injury, crush injury, burns, renal insufficiency	IV: ≤2 years: 2 mg/kg >2 years: 1 mg/kg (30–60 sec onset, 4–6 min duration) IM: 3–4 mg/kg (3–4 min onset, 10–30 min duration) Max dose: 150 mg/dose IV/IM	Hyperkalemia, trigger of malignant hyperthermia, masseter spasm, bradycardia, muscle fasciculations, increased intracranial, intraocular, and intragastric pressure
Rocuronium (nondepolarizing)	Minimal cardiovascular effect, reversible with sugammadex	Caution in patients with difficult airway	1.2 mg/kg IV/IM/IO (30–60 sec onset, 30–40 min duration) Max dose: 100 mg	Prolonged duration in hepatic failure
Vecuronium (nondepolarizing)	Minimal cardiovascular effect, reversible with sugammadex	Caution in patients with difficult airway	0.15–0.2 mg/kg IV/IO (1–3 min onset, 30–40 min duration) Max dose: 10 mg	Prolonged duration in hepatic failure

CPP, Cerebral perfusion pressure; HR, heart rate; ICP, intracranial pressure; IM, intramuscular; IV, intravenous; IO, intraosseous; mcg, microgram; SVR, systemic vascular resistance; RSI, rapid sequence intubation.

(3) High-flow nasal cannula (**HFNC**):

- (a) High-flow, noninvasive respiratory support provides a heated and humidified air-oxygen mixture that may improve gas exchange by providing airway-distending pressure.
- (b) Optimal and maximal flow rates are unknown. Consensus supports a maximum flow rate of up to **2 L/kg/min** or 12 L/min for infants and toddlers, 30 L/min for children, and up to 50 L/min for adolescents and adults.

b. Noninvasive positive pressure ventilation (**NIPPV**):

- (1) **CPAP:** Delivery of a continuous, distending positive airway pressure independent of patient inspiratory effort.
- (2) **BiPAP:** Pressure-limited ventilatory mode in which the clinician sets an inspiratory positive airway pressure (IPAP) and expiratory positive airway pressure (EPAP).
  - (a) EPAP is started at 4 to 5 cmH<sub>2</sub>O and increased to a maximum of 8 to 12 cmH<sub>2</sub>O.
  - (b) Set 4 to 6 cmH<sub>2</sub>O of pressure support, or the difference between IPAP and EPAP.
  - (c) Consider setting a “backup rate,” or respiratory rate just shy of the spontaneous respiratory rate to be delivered in case of apnea.

## c. Mechanical Ventilation:

(1) **Parameters:**

- (a) Rate: Number of mechanical breaths delivered per minute.
- (b) FiO<sub>2</sub>: Fraction of oxygen in inspired gas.
- (c) PIP: Peak inspiratory pressure attained during respiratory cycle.
- (d) Positive end-expiratory pressure (PEEP): Distending pressure that increases functional residual capacity (FRC), or volume of gas at the end of exhalation.
- (e) Mean airway pressure (P<sub>aw</sub>): Average airway pressure over entire respiratory cycle, which correlates to mean alveolar volume.
- (f) Tidal volume (V<sub>T</sub>): Volume of gas delivered during inspiration.
- (g) Time: May indicate a function of time spent in inspiration (T<sub>i</sub>), in high pressure (T<sub>high</sub>), or in low pressure (T<sub>low</sub>).

(2) **Modes of Ventilation:**

- (a) **Controlled Ventilation:** Ventilation is completely mechanical with no spontaneous ventilation efforts expected from the patient.
  - (i) Pressure-controlled ventilation (**PCV**): A preset respiratory rate and T<sub>i</sub> delivers a pressure-limited breath (the set pressure is maintained during inspiration). V<sub>T</sub> is determined by the preset pressure as well as lung compliance and resistance.
  - (ii) Volume-controlled ventilation (**VCV**): A preset respiratory rate and T<sub>i</sub> delivers a preset V<sub>T</sub>.

- (b) Intermittent mandatory ventilation (**IMV**): Allows the patient to breathe spontaneously between a preset number of (mandatory) mechanical breaths.
- (i) Synchronized IMV (**SIMV**): If patient initiates spontaneous breath, mandatory breath is synchronized with patient effort rather than spaced evenly over each minute.
  - (ii) If spontaneous breathing rate is less than mandatory rate, some mandatory breaths will be delivered in the absence of patient effort.
  - (iii) Delivered breaths may be volume regulated or pressure limited.
- (c) Airway-pressure-release ventilation (**APRV**): Most of the respiratory cycle is spent at a high distending pressure (a functionally high CPAP phase) with intermittent, short release to a low pressure for a brief ventilation phase. Spontaneous breathing can be superimposed at any point in the cycle.
- (d) **Support ventilation:** Mechanical breaths support patient-initiated breaths, but no mandatory breaths are provided.
- (i) Pressure support (**PS**): Delivers a preset amount of pressure to assist spontaneous respiratory effort.
  - (ii) Often used in combination with other modes of ventilation to support spontaneous breaths greater than preset respiratory rates.
- (e) High-frequency oscillatory ventilation (**HFOV**): Gas flow pressurizes the system to the preset  $P_{aw}$  while a piston moves backwards and forwards to force air and withdraw a small  $V_T$  (that approximates anatomic dead space) into the lungs at rates exceeding normal respiratory rates.
- (3) **Management:** The three subdivisions of mechanical ventilatory support are the acute (lung recruitment), maintenance (lung recovery), and weaning phases.
- (a) **Acute:** See Table 1.3 for ventilation parameter initial settings and titration effects.
  - (b) **Maintenance:** To avoid volutrauma, barotrauma, or oxygen toxicity, maintain  $V_T$  at 4–6 mL/kg,  $PIP < 35 \text{ cmH}_2\text{O}$ , and  $\text{FiO}_2 \leq 60\%$ .
  - (c) **Weaning:**
    - (i) Assess daily for clinical signs of readiness, such as spontaneous breathing efforts.
    - (ii) Standard indices indicating readiness include:  $\text{FiO}_2 < 50\%$ , PEEP of 5  $\text{cmH}_2\text{O}$ ,  $PIP < 20 \text{ cmH}_2\text{O}$ , normalized rate for age, and absence of hypercapnia or acidosis.
    - (iii) The general approach combines gradual weaning of parameters and reliance on pressure-support modes.
  - (d) **Extubation:**
    - (i) Provide humidified inspired oxygen after extubation.

**TABLE 1.3****MECHANICAL VENTILATION PARAMETER SETTINGS AND EFFECTS<sup>11,14,17</sup>**

Parameter	Initial Setting	Effect of ↑ on PaCO <sub>2</sub>	Effect of ↑ on PaO <sub>2</sub>
PIP	≤28 cmH <sub>2</sub> O or ≤29–32 cmH <sub>2</sub> O for reduced chest wall compliance	↔	↑
PEEP	3–5 cmH <sub>2</sub> O	↑	↑↑
V <sub>T</sub>	5–8 mL/kg or 3–6 mL/kg for poorly compliant lungs	↔	↑
Rate	Normal rate for age	↔	Minimal ↑
I:E ratio	(33%) 1:2 (67%)	No change	↑
FiO <sub>2</sub>	<50% and/or to maintain PaO <sub>2</sub> between 80 and 100 mmHg and SpO <sub>2</sub> ≥95%	No change	↑↑
<b>High-Frequency Ventilation Parameters</b>			
Amplitude (ΔP)	Set to produce a visible wiggling motion to the level of the lower abdomen	↓	No change
Frequency (Hz)	Range from 3–20 Hz (180–1200 breaths per min)	↑↑	↓
P <sub>aw</sub>	5 cmH <sub>2</sub> O > than P <sub>aw</sub> of previous conventional ventilation	Minimal ↓	↑

FiO<sub>2</sub>, Fraction of inspired oxygen; I:E, inspiratory to expiratory; Hz, hertz; P<sub>aw</sub>, mean airway pressure; PaCO<sub>2</sub>, partial pressure of carbon dioxide (arterial); PaO<sub>2</sub>, partial pressure of oxygen (arterial); PEEP, positive end-expiratory pressure; PIP, peak inspiratory pressure; V<sub>T</sub>, tidal volume.

- (ii) In case of uncuffed tube or the absence of an air leak at delivered pressure <30 cmH<sub>2</sub>O, consider 24 hours of dexamethasone (airway edema dosing) to prevent postextubation stridor.

**II. MANAGEMENT OF SHOCK<sup>3,5,7,11</sup>****A. Definition: Physiologic state characterized by inadequate oxygen and nutrient delivery to meet tissue demands**

1. **Compensated:** Perfusion to vital organs is maintained by compensatory mechanisms.
  - a. Tachycardia is often the first and most sensitive vital sign change.
  - b. Blood flow is redirected from nonvital organs and tissues to vital organs by a selective increase in systemic vascular resistance (SVR), resulting in reduced peripheral perfusion and decreased urine volume.
  - c. Cardiac contractility increases to maintain cardiac output.
  - d. Increased venous smooth muscle tone improves preload and stroke volume.
2. **Decompensated:** Perfusion to vital organs is compromised. Denoted by **hypotension**, poor perfusion, oliguria/anuria, and altered mental status.

**B. Etiology: Categorized into four basic types:**

1. **Hypovolemic:** inadequate fluid intake, increased fluid losses (hemorrhage, gastroenteritis, burns). Assess for tachycardia, narrow pulse pressure, delayed capillary refill, cool extremities.
2. **Cardiogenic:** congenital heart disease, myocarditis, cardiomyopathy, arrhythmia. Assess for increased respiratory effort from pulmonary edema, hepatomegaly, jugular venous distension, and cyanosis.
3. **Distributive:** sepsis, anaphylaxis, neurogenic (e.g., high cervical spine injury)
  - a. Assess for tachycardia, fever, and petechial, purpuric, or urticarial rash.
  - b. Warm septic shock is characterized by bounding peripheral pulses, flash capillary refill, and wide pulse pressure.
  - c. Cold septic shock is characterized by decreased peripheral pulses, delayed capillary refill, and narrow pulse pressure.
  - d. Neurogenic shock is characterized by hypotension with a wide pulse pressure, normal HR or bradycardia, and hypothermia.
4. **Obstructive:** tension pneumothorax, cardiac tamponade, pulmonary embolism, ductal-dependent congenital cardiac abnormalities
  - a. Early clinical presentation is indistinguishable from hypovolemic shock. Progression of shock leads to signs and symptoms similar to cardiogenic shock.
  - b. Cardiac tamponade is characterized by muffled heart sounds and pulsus paradoxus.
  - c. Ductal-dependent lesions may be characterized by higher preductal versus postductal BP or arterial oxygen saturation.

**C. Management**

1. Administer 100% supplemental oxygen initially regardless of oxygen saturation to optimize oxygen delivery. Once perfusion restored, titrate as able to avoid adverse effects from hyperoxia.
2. See **Table 1.4** for type- and etiology-specific pathophysiology and management of shock.
3. See **Table 1.5** for vasoactive agents to support cardiac output. Vasoactive agents affect SVR (vasodilators and vasoconstrictors), cardiac contractility (inotropes), or HR (chronotropes). Some agents increase blood flow via contractility and vasodilation (inodilators) or increase perfusion pressure via contractility and vasoconstriction (inoconstrictors).

**III. MANAGEMENT OF COMMON EMERGENCIES****A. Anaphylaxis<sup>18</sup>**

1. **Definition:** Rapid-onset (minutes to hours) usually immunoglobulin E (IgE)-mediated systemic allergic reaction involving multiple organ systems, including two or more of the following:
  - a. **Cutaneous/mucosal** (80% to 90%): flushing, urticaria, pruritis, angioedema

**TABLE 1.4****PATHOPHYSIOLOGY AND MANAGEMENT OF SHOCK<sup>3,5</sup>**

Type	HR	Preload	Contractility	SVR	Management
Hypovolemic	↑	↓↓	Normal or ↑	↑	Rapid administration of isotonic crystalloids Replace blood loss with 10 mL/kg PRBCs boluses Consider colloids if response is poor to crystalloids and loss of protein-containing fluids is suspected
Distributive	↑ or ↓	Normal or ↓	Normal or ↓	±	Administer isotonic crystalloids to expand intravascular volume Support with vasoressors if fluid-refractory
Septic	↑	↓↓	Normal or ↓	↓	<b>Within 1st hour:</b> Administer isotonic crystalloid boluses, broad-spectrum antibiotics, and consider stress-dose hydrocortisone <b>Warm:</b> Support with norepinephrine or high-dose dopamine <b>Cold:</b> Support with epinephrine or dopamine
Neurogenic	Normal or ↓	↓↓	±	↓↓	Position patient flat or head-down Administer a trial of isotonic crystalloid therapy If fluid-refractory, support with norepinephrine or epinephrine Maintain normothermia
Cardiogenic	±	↑	↓↓	↑	Consider cautious administration (10–20 min) of isotonic crystalloid (5–10 mL/kg); stop if fluid overload develops Support with inodilator milrinone Decrease metabolic demand with oxygen therapy, ventilatory support and antipyretics
Obstructive	↑	±	Normal	↑	Correct underlying cause Start prostaglandin E <sub>1</sub> if ductal-dependent lesion suspected Consider initial fluid challenge with isotonic crystalloid (10–20 mL/kg)

HR, Heart rate; PRBCs, packed red blood cells; SVR, systemic vascular resistance

**TABLE 1.5****MEDICATIONS TO SUPPORT CARDIAC OUTPUT<sup>5,7,11</sup>**

Medication	Dose	Mechanism	Comments
Dobutamine	2–20 mCg/kg/min	Selective $\beta_1$ agonist	Inotrope May predispose to arrhythmia Indicated for normotensive, poorly perfused shock
Dopamine	5–20 mCg/kg/min	Direct dopamine receptor agonist, indirect $\beta$ and $\alpha$ agonist (stimulates norepinephrine release), direct $\alpha$ agonist at high dose (>15 mCg/kg/min)	Low to moderate dose: inotrope, chronotrope, splanchnic vasodilator High dose: vasopressor Indicated for shock with poor contractility and/or low SVR and cold septic shock if epinephrine unavailable
Epinephrine	0.1–1 mCg/kg/min	$\beta_1$ and $\beta_2$ agonist at low dose (<0.3 mCg/kg/min), $\alpha_1$ agonist at high dose (>0.3 mCg/kg/min)	Low dose: inotrope, chronotrope, vasodilator High dose: vasopressor Indicated for hypotensive shock with marked circulatory instability and cold septic shock
Milrinone	Loading: 50 mCg/kg over 15 min, then 0.25–0.75 mCg/kg/min	Type III phosphodiesterase-inhibitor	Inodilator Improves cardiac output with little effect on heart rate Indicated for normotensive shock with myocardial dysfunction and cold septic shock refractory to epinephrine
Norepinephrine	0.05–2.5 mCg/kg/min	$\alpha_1$ and $\beta_1$ agonist	Vasoconstrictor, mild inotrope Indicated for shock with low SVR (warm septic, anaphylactic, spinal) and cold shock refractory to epinephrine if diastolic BP low
Phenylephrine	Loading: 5–20 mCg/kg/dose (max 500 mCg), then 0.1–0.5 mCg/kg/min	Pure $\alpha_1$ agonist	Vasoconstrictor
Vasopressin (ADH)	0.17–8 mUnits/kg/min	Vasopressin receptor agonist	Vasoconstrictor Consider for cardiac arrest, refractory hypotension in septic shock and GI hemorrhage

ADH, Antidiuretic hormone; BP, blood pressure; cGMP, cyclic guanosine monophosphate; GI, gastrointestinal; mCg, microgram; NO, nitric oxide; SVR, systemic vascular resistance.

- b. **Respiratory** (70%): laryngeal edema, bronchospasm, dyspnea, wheezing, stridor, hypoxemia
  - c. **Gastrointestinal** (45%): vomiting, diarrhea, nausea, crampy abdominal pain
  - d. **Circulatory** (45%): tachycardia, hypotension, syncope
2. Management:
- a. **Stop exposure** to precipitating antigen.
  - b. While performing A-B-Cs, immediately give intramuscular (**IM**) **epinephrine** into midanteriorlateral thigh.
    - (1) For child, administer **0.01 mg/kg of 1 mg/mL solution** up to a max dose of 1 mg/dose. For adult-sized patients, first administer **0.2 to 0.5 mg of 1 mg/mL solution**, increasing as necessary up to max single dose of 1 mg.
    - (2) Autoinjector dosing: 7.5 to <15 kg use **0.1 mg**, 15 to <30 kg use **0.15 mg**, ≥30 kg use **0.3 mg**.
    - (3) Repeat dosing every 5 to 15 minutes as needed.
  - c. Provide oxygen and ventilatory assistance. Consider early endotracheal intubation.
  - d. Obtain IV access. For management of shock, resuscitate with 20 mL/kg isotonic crystalloid fluid boluses and vasoactive agents as needed.
  - e. Place patient in Trendelenburg position (head 30 degrees below feet).
  - f. Consider adjuvant pharmacologic agents:
    - (1) **Histamine receptor antagonist:** Diphenhydramine (H1-antagonism) and ranitidine/famotidine (H2-antagonism)
    - (2) **Corticosteroid:** Methylprednisolone or dexamethasone
    - (3) **Inhaled β<sub>2</sub> agonist:** Albuterol
  - g. Symptoms may recur (“biphasic anaphylaxis”) up to 72 hours after initial recovery.
    - (1) Observe for a minimum of 4 to 10 hours for late-phase symptoms.
    - (2) Discharge with an epinephrine autoinjector and an anaphylaxis action plan.

## B. Upper Airway Obstruction

1. Epiglottitis<sup>19-20</sup>
- a. **Definition:** Life-threatening, rapidly progressive inflammation (usually infectious) of the supraglottic region.
    - (1) Most often affects children between 1 and 7 years, but may occur at any age.
    - (2) May be caused by infection, thermal injury, caustic ingestion, or foreign body.
    - (3) Most common infectious organisms include *Haemophilus influenzae* (unvaccinated), *Streptococcus pneumoniae*, group A streptococci, and *Staphylococcus aureus*.
    - (4) Patients often present febrile, toxic-appearing, and tripodding in respiratory distress. Drooling, dysphagia and inspiratory stridor are common. Barky cough is absent.

- b. **Management:** Avoid *any agitation* of the child prior to securing airway to prevent impending complete obstruction.
- (1) Allow child to assume a position of comfort. Unobtrusively provide blow-by oxygen. Monitor with pulse oximetry.
  - (2) To secure airway, emergently consult difficult airway personnel (pediatric anesthesiologist, intensivist, and/or otolaryngologist).
    - (a) If unstable (unresponsive, cyanotic, bradycardic), emergently intubate.
    - (b) If stable with high suspicion, escort patient to OR for laryngoscopy and intubation under general anesthesia. Equipment for tracheotomy should be readily available.
    - (c) If stable with moderate or low suspicion, obtain lateral neck radiograph to assess for “thumb sign” of an inflamed epiglottis.
  - (3) Initiate broad-spectrum antibiotic therapy (e.g., vancomycin and Ceftriaxone).

## 2. Croup<sup>21-22</sup>

- a. **Definition:** Common infectious inflammation of the subglottic area.
- (1) Most common in infants aged 6 to 36 months.
  - (2) 75% of infections are caused by parainfluenza virus.
  - (3) Patients present with fever, barking cough, inspiratory stridor, and increased work of breathing, often worse at night.
- b. **Management:**
- (1) Administer oxygen to children with hypoxemia or severe respiratory distress. Consider humidified air, although current consensus suggests it is ineffective for mild to moderate disease.
  - (2) If **no stridor at rest**, give dexamethasone. Consider nebulized budesonide in patients vomiting or who lack IV access.
  - (3) If **stridor at rest**, give dexamethasone and nebulized racemic epinephrine. Observe for 2 to 4 hours given short duration of action of nebulized epinephrine.
  - (4) Indications for hospitalization include >1 racemic epinephrine nebulization required, atypical age (<6 months), severe respiratory distress, or dehydration.
  - (5) Consider heliox (helium and oxygen mixture) to improve turbulent airflow in moderate to severe croup, although benefit is controversial.

## 3. Foreign body aspiration<sup>1,20,23</sup>

- a. **Definition:** Acute airway obstruction from aspiration of an organic (e.g., nuts, seeds, grapes, hot dogs) or inorganic (e.g., coins, pins, beads, balloons, small toy parts) foreign body.
- (1) Male children younger than 3 years of age are most susceptible.
  - (2) Patients (<40%) present with classic triad of paroxysmal cough, wheezing, and decreased air entry. Other manifestations include cyanosis, fever, stridor, and persistent pneumonia or notably may be asymptomatic.

- (3) The most common location is the right main bronchus (45% to 57%), then left main bronchus (18% to 40%), and trachea (10% to 17%).
- b. **Management:** Care is taken to avoid converting a partial airway obstruction into complete obstruction.
- (1) If **not** breathing (no cough or sound):
    - (a) Infant: Deliver repeated cycles of 5 back blows followed by 5 chest compressions until object is expelled or victim becomes unresponsive.
    - (b) Child: Perform subdiaphragmatic abdominal thrusts (Heimlich maneuver) until object is expelled or victim becomes unresponsive.
    - (c) Patients should be taken to the OR for emergent removal under direct laryngoscopy and bronchoscopy.
  - (2) If **breathing** (forcefully coughing, phonating):
    - (a) Obtain posteroranterior chest (including neck) radiograph to screen for radiopaque body or mediastinal shift. Consider inspiratory and expiratory films (or bilateral lateral decubitus in young patients) to assess for air trapping. A normal chest radiograph does not rule out foreign body.
    - (b) If clinical concern is high, consider urgent bronchoscopy or laryngoscopy.
  - (3) If patient becomes **unresponsive**: initiate CPR immediately.
    - (a) After 30 chest compressions, open airway and remove foreign body if visible. Do **not** perform blind sweep.
    - (b) Attempt to give two breaths and continue with cycles of chest compressions and ventilations until object expelled.

### C. Status Asthmaticus<sup>24-28</sup>

1. **Definition:** Inflammatory airflow obstruction secondary to triad of airway edema, bronchoconstriction, and hyperresponsiveness.
2. **Examination:** Assess breathlessness, speech, alertness, respiratory rate, accessory muscle use, wheezing, HR, pulsus paradoxus, peak expiratory flow, SpO<sub>2</sub>, and pCO<sub>2</sub>.
3. **Management:**
  - a. Provide oxygen to achieve SpO<sub>2</sub> ≥90%. If hypoxemia not readily corrected with supplemental oxygen, consider pneumothorax, pneumonia, methemoglobinemia, or other process.
  - b. See **Table 1.6** for pharmacologic agents used in acute asthma exacerbations.
  - c. Ventilation interventions:
    - (1) Normalizing pCO<sub>2</sub> can be a sign of impending respiratory failure.
    - (2) NIPPV (e.g., BiPAP) may be used in patients with impending respiratory failure to avoid intubation but requires a cooperative patient with spontaneous respirations.

**TABLE 1.6****STATUS ASTHMATICUS MEDICATIONS<sup>24-28</sup>**

Medication	Dose	Comments
<b>Short-acting <math>\beta_2</math> agonist</b>		
Albuterol	<b>Mild to Moderate:</b> Administer up to 3 doses in the first hour MDI: 4–8 puffs (90 mCg/puff) q20 min–4 hr Nebulizer: 0.15 mg/kg (min 2.5 mg, max 5 mg) q20 min–4 hr <b>Severe:</b> Continuous nebulization: 0.5 mg/kg/hr (max 30 mg/hr)	Inhaler (with spacer) is preferred delivery method given equal or greater efficacy, fewer side effects, and shorter length of stay
<b>Anticholinergics</b>		
Ipratropium bromide	Administer q20 min for 3 doses with albuterol MDI: 4–8 puffs (17 mCg/puff) Nebulizer: 0.25–0.5 mg	No additional benefit shown in inpatient setting
<b>Systemic corticosteroids</b>		
Dexamethasone	<b>Mild to Moderate:</b> 0.6 mg/kg/day PO/IV/IM for 1–2 days (max 16 mg/day)	Equally as efficacious as prednisone or prednisolone with fewer side effects, better compliance and palatability
Prednisone, Prednisolone Methylprednisolone	<b>Mild to Severe:</b> 2 mg/kg/day PO for 5–7 days (max 60 mg/day) <b>Severe:</b> Loading: 2 mg/kg IV (max 60 mg) Maintenance: 2 mg/kg/day IV divided q6–12hr (max <12 years 60 mg/day, ≥12 years 80 mg/day)	Taper if course ≥7 days or bounce back from recent exacerbation No known advantage in severe exacerbations for higher dosing or IV administration over oral therapy, provided normal GI transit and absorption
<b>Injected <math>\beta_2</math> agonist</b>		
Epinephrine	0.01 mg/kg of 1 mg/mL IM (max 1 mg) q15–20 min for up to 3 doses	Consider for severe exacerbation with minimal air entry Consider quickly accessed autoinjector
Terbutaline	SC: 0.01 mg/kg (max 0.25 mg/dose) q20 min for up to 3 doses, then as needed q2–6 hr IV load: 2–10 mCg/kg IV IV continuous: 0.1–0.4 mCg/kg/min (doses as high as 10 mCg/kg/min have been used)	Consider for severe exacerbation with minimal air entry IV administration may decrease the need for mechanical ventilation
<b>Adjunct therapies</b>		
Magnesium sulfate	25–75 mg/kg/dose IV (max 2 g), infuse over 20 min	Smooth muscle relaxant May cause hypotension; consider simultaneous fluid bolus Reduces hospitalization rates in severe exacerbations

1

**TABLE 1.6—CONT'D**

Medication	Dose	Comments
Ketamine	1–2 mg/kg IV bolus followed by 1 mg/kg/h infusion, titrated to affect	Used as a sympathomimetic adjuvant in effort to avoid endotracheal intubation Preferred induction-sedative agent for endotracheal intubation in asthma
Aminophylline	6 mg/kg IV bolus over 20 min followed by 0.5–1.2 mg/kg/h infusion (age-dependent, see formulary)	Use limited to severe exacerbations refractory to traditional interventions May improve lung function and oxygen saturation but is associated with greater length of stay and time to symptom improvement
Heliox	Optimal helium-oxygen ratio unknown, most commonly 70:30 or 80:20 mixture	Low density gas that promotes laminar airflow and improves $\beta_2$ agonist delivery to distal airways Useful in severe or very severe exacerbations
Inhaled anesthetics (e.g., halothane, isoflurane, sevoflurane)	Consultation with pediatric anesthetist recommended	Rescue therapy for intubated patients with life-threatening exacerbation Associated with prolonged length of stay and increased cost Isoflurane may cause hypotension Sevoflurane may cause renal tubular injury, hepatotoxicity, neuropathy

*GI*, Gastrointestinal; *IM*, intramuscular; *IV*, intravenous; *mcg*, microgram; *MDI*, metered-dose inhaler; *SC*, subcutaneous.

- (3) Intubation should be approached cautiously given the risk of worsening air-trapping and difficulty in managing the transition from extremely negative to positive pressure ventilation.
  - (a) Indications include severe airway obstruction, markedly increased work of breathing, refractory hypoxemia, and impending respiratory arrest.
  - (b) Ventilation strategies include slower rates with prolonged expiratory phase, minimal end-expiratory pressures, and short inspiratory times to minimize hyperinflation and air trapping.
- (4) Consider inhaled anesthetics or ECMO as rescue therapies.

#### **D. Pulmonary Hypertensive Crisis<sup>11,29</sup>**

1. Definition:
  - a. Pulmonary hypertension (PH) is defined as resting elevated mean pulmonary artery pressure (PAP)  $\geq 25 \text{ mmHg}$  in children  $> 3$  months of age.

- b. A **pulmonary hypertensive crisis** is a sudden increase in PAP and pulmonary vascular resistance (PVR) that causes acute right-sided heart failure.
- (1) May be triggered by pain, anxiety, tracheal suctioning, hypoxia, acidosis, or respiratory illness. Most commonly described after cardiac surgery or in the setting of rapid withdrawal of PH-specific therapies.
  - (2) Patients present with systemic hypotension, oxygen desaturation (if atrial or ventricular communication present), and decreased EtCO<sub>2</sub> on capnography (reduced pulmonary blood flow).
  - (3) Assess for increased intensity of systolic murmur (worsening tricuspid regurgitation) and increased hepatomegaly.
2. **Management:** Timely consultation with providers with expertise in managing PH is recommended.
- a. Implement efforts to keep patient calm. Consider opiates, sedatives, and neuromuscular blockade to reduce stress response, especially postoperatively. Avoid agents that decrease SVR.
  - b. Administer supplemental oxygen to treat hypoxemia or as an adjunct to pulmonary vasodilators.
  - c. Avoid acute hypercarbia and acidosis, which abruptly increase PVR. Consider brief hyperventilation or sodium bicarbonate infusions.
  - d. Diuretics treat congestive symptoms. Avoid excessive reduction in intravascular volume leading to decreased cardiac output.
  - e. NIPPV may improve oxygenation, treat hypoventilation, and reduce work of breathing. Weigh benefits against increasing patient anxiety and delaying mechanical ventilation.
  - f. PH-specific pharmacologic therapies aim to induce pulmonary vasodilation, support the right ventricle, and maintain cardiac output.
- (1) **Inhaled pulmonary vasodilators:** Nitric oxide
    - (a) Indicated to reduce need for ECMO in patients with an oxygen index >25.
    - (b) Rapid withdrawal of low doses may cause rebound PH. Gradually decrease dose when weaning.
    - (c) Monitor for methemoglobinemia.
  - (2) **Phosphodiesterase type-5 inhibitors:** Sildenafil, tadalafil
    - (a) Often used to prevent rebound PH associated with cessation of nitric oxide.
    - (b) Monitor for acute hypotension or hypoxemia secondary to increased alveolar-arterial gradient.
  - (3) **Synthetic prostacyclin analogs:** Epoprostenol (Flolan), treprostinil, iloprost
  - (4) **Endothelin receptor antagonist:** Bosentan
- g. Consider ECMO or emergent atrial septostomy in case of failed medical management.

### E. Hypertensive Crisis<sup>11,30</sup>

1. Definition:
  - a. For normal BP values based on age and height, see Chapter 7.
  - b. **Hypertensive emergency:** Acutely elevated BP (usually significantly >99th percentile for age and gender) with evidence of end-organ damage.
    - (1) Most commonly secondary to renal disease, catecholamine-producing tumors, endocrine syndromes, toxidromes, medication withdrawal, or elevated intracranial pressure (ICP).
    - (2) Presents with encephalopathy (e.g., headaches, vomiting, seizures, altered mental status), vision disturbance, congestive heart failure (e.g., dyspnea, peripheral edema, gallop rhythm), and acute kidney injury.
  - c. **Hypertensive urgency:** Acutely elevated BP (usually >5 mmHg greater than the 99th percentile for age and gender) without evidence of end-organ damage.
    - (1) Most commonly primary hypertension in children >7 years age, followed by renal disease.
    - (2) Present with minor complaints (e.g., headaches, nausea).
2. Management:
  - a. Rule out increased ICP before instituting antihypertensive treatment given critical need to maintain cerebral perfusion.
  - b. Goal is to reduce BP by **≤25%** in the **first 8 hours**, then gradual normalization over the next **24 to 48 hours**.
  - c. See [Table 1.7](#) for hypertensive emergency and urgency medications.

### F. Hypercyanotic Crisis (“Tet spell”)<sup>20,31</sup>

1. **Definition:** Cyanotic emergency secondary to an acute worsening of a preexisting right ventricular outflow tract obstruction (e.g., in a patient with tetralogy of Fallot) that prevents pulmonary blood flow and induces a right-to-left intracardiac shunt.
  - a. Peak incidence occurs between 2 and 4 months of age.
  - b. Usually occurs in the morning after crying, feeding, or defecation.
  - c. Patients present with extreme cyanosis, hyperpnea, tachypnea, and agitation.
2. **Management:** Follow stepwise approach, escalating if spell is not broken.
  - a. Make every effort to calm the child. Allow parent to comfort. Consider oral sucrose analgesia (e.g., Sweet-Ease).
  - b. Bring knees to chest in infants or encourage squatting in older children to increase SVR and decrease shunting.
  - c. Administer 100% oxygen **if patient tolerates**, although effect is limited given absence of effective pulmonary blood flow.
  - d. For stepwise pharmacologic abortive management, see [Table 1.8](#).
  - e. Consider isotonic crystalloid resuscitation (5 to 10 mL/kg boluses) to ensure adequate preload if patient is dehydrated.

**TABLE 1.7****HYPERTENSIVE CRISIS MEDICATIONS<sup>11,30</sup>**

Drug	Dose	Pharmacokinetics	Mechanism	Side Effects
<b>PARENTERAL THERAPY</b>				
Esmolol	Bolus: 100–500 mCg/kg Infusion: 100–500 mCg/kg/min (max 1000 mCg/kg/min)	Onset: Immediate Duration: 10–30 min	$\beta_1$ blocker	Bradycardia, bronchospasm (at high doses)
Hydralazine	0.1–0.2 mg/kg/dose IV/IM (max 2 mg/kg/dose or 20 mg) q4–6 hr PRN	Onset: 5–30 min Duration: 2–6 hr	Direct arteriole vasodilator	Reflex tachycardia, flushing, lupus-like syndrome
Labetalol	Bolus: 0.2–1 mg/kg (max 40 mg) Infusion: 0.4–1 mg/kg/hr (max 3 mg/kg/hr)	Onset: 2–5 min Duration: 2–6 hr	$\beta_1$ , $\beta_2$ , and $\alpha_1$ blocker	Hyperkalemia, bronchospasm; caution in liver failure due to prolonged duration of action
Nicardipine	Start at 0.5–1 mCg/kg/min (max 5 mCg/kg/min or 15 mg/hr)	Onset: 1–2 min Duration: 2–4 hr	Calcium channel blocker	Reflex tachycardia
Nitroprusside	0.3–4 mCg/kg/min (max 10 mCg/kg/min)	Onset: 30 sec to 2 min Duration: 1–10 min	Arterial and venous vasodilation via NO	Cyanide toxicity
<b>ENTERAL THERAPY</b>				
Captopril	0.3–0.5 mg/kg (max 6 mg/kg/day or 450 mg/24h)	Onset: 15–30 min Duration: 2–6 hr	ACE inhibitor; lowers blood pressure without causing tachycardia	Hyperkalemia, neutropenia, angioedema, cough; contraindicated in bilateral renal artery stenosis or solitary kidney
Clonidine	2–10 mCg/kg/dose q6–8 hr (max 25 mCg/kg/24 hr up to 0.9 mg/24 hr)	Onset: 30–60 min Duration: 6–10 hr	Peripheral vasodilator	Bradycardia, rebound hypertension
Nifedipine	0.1–0.25 mg/kg/dose q4–6 hr PO/SL (max 10 mg/dose, 1–2 mg/kg/24 hr)	Onset: 15–30 min Duration: 4–6 hr	Calcium channel blocker	Precipitous hypotension, reflex tachycardia

ACE, Angiotensin-converting enzyme; IM, intramuscular; IV, intravenous; mCg, microgram; NO, nitric oxide; PO, oral; PRN, as needed; SL, sublingual

**TABLE 1.8****HYPERCYANOTIC CRISIS ABORTIVE MEDICATIONS<sup>20,31</sup>**

Medication	Dose	Comment
Ketamine	1–2 mg/kg IM or IV, administer IV dose over 60 sec	Sedating, increases SVR
Morphine	0.05–0.2 mg/kg IM, SC or IV; do not wait for IV access	Calms agitation, suppresses hyperpnea Monitor for respiratory depression
Phenylephrine	5–20 mCg/kg IV bolus	$\alpha$ Agonist, increases SVR
Propranolol	0.15–0.25 mg/kg, via slow IV push Max initial dose 1 mg	$\beta$ Blockade decreases heart rate, promoting ventricular filling Monitor for hypotension

IM, Intramuscular; IV, intravenous; SC, subcutaneous; SVR, systemic vascular resistance.

- f. Treat acidosis with sodium bicarbonate.
- g. For refractory spells, consider general anesthesia and emergent surgery for palliation with a systemic to pulmonary shunt or full repair.

**G. Altered Level of Consciousness<sup>20,32</sup>**

1. **Definition:** A spectrum of impaired consciousness spanning confusion, disorientation, agitation, stupor, lethargy, and coma.
  - a. Fluctuations in level of consciousness are common and progression may occur rapidly.
  - b. **Coma:** Refers to an unarousable state.
  - c. **Lethargy:** Refers to a depressed consciousness resembling sleep from which a patient can be aroused but immediately returns to depressed state.
  - d. **Stupor:** Refers to a state of depressed responses to external stimuli but not totally asleep.
  - e. Standard descriptors of level of responsiveness include:
    - (1) The **Glasgow Coma Scale** (and modified scale for infants): See **Table 1.9** to score level of responsiveness.
    - (2) **AVPU** mnemonic: Graded as **A** if alert, **V** if responsive to verbal stimulation, **P** if responsive to painful stimulation, or **U** if unresponsive.
  - f. Broad differential considerations include **Drugs**, **Infection**, **Metabolic**, and **Structural causes (DIMS)**.
  - g. See **Table 1.10** for common etiologies and targeted work-up recommendations.
2. **Management:** Stabilize initially. Further management is aimed at correcting underlying etiology.
  - a. Airway, Breathing, Circulation:
    - (1) Administer supplemental oxygen to patients presenting with seizure or with signs of shock, regardless of pulse oximetry reading.
    - (2) Intubation is indicated in patients unable to protect their airway.
    - (3) Consider delaying administration of atropine unless necessary secondary to the loss of pupillary light reflex.

**TABLE 1.9****COMA SCALES<sup>20</sup>**

Grading	Glasgow Coma Scale	Modified Coma Scale for Infants
<b>EYE OPENING</b>		
4	Spontaneous	Spontaneous
3	To speech	To speech
2	To pain	To pain
1	None	None
<b>VERBAL</b>		
5	Oriented	Coos or babbles
4	Confused	Irritable
3	Inappropriate words	Cries to pain
2	Nonspecific sounds	Moans to pain
1	None	None
<b>MOTOR</b>		
6	Follows commands	Normal, spontaneous movements
5	Localizes pain	Withdraws to touch
4	Withdraws to pain	Withdraws to pain
3	Abnormal flexion	Abnormal flexion
2	Abnormal extension	Abnormal extension
1	None	None

Data from Shaw KN, Bachur RG. *Fleisher & Ludwig's Textbook of Pediatric Emergency Medicine*. 7th ed. Philadelphia: Lippincott Williams & Wilkins; 2016.

- (4) Avoid hypercarbia, maintaining  $\text{PaCO}_2$  in normal range. Prophylactic hyperventilation is not recommended.
- b. **Dextrose:** Correct hypoglycemia immediately with a 5 to 10 mL/kg bolus of 10% dextrose or 2 to 4 mL/kg of 25% dextrose. After bolus, start a continuous infusion of dextrose-containing fluids to avoid recurrent hypoglycemia.
- c. **Imaging:** Request emergency head computed tomography (CT) if patient stable for transport. Consult with neurosurgical team if indicated.
- d. **Hyponatremia:** Often asymptomatic unless sodium decreases rapidly or becomes severe (i.e.,  $<125 \text{ mmol/L}$ ).
  - (1) Treat **symptomatic** hyponatremia immediately with a 3 to 5 mL/kg bolus of 3% hypertonic saline over 15 to 30 minutes until seizure activity ceases or serum sodium level is  $>125 \text{ mmol/L}$ .
  - (2) See Chapter 11 for subsequent, slow correction of asymptomatic hyponatremia.
- e. **Infection:** If presentation concerning for severe sepsis, treat empirically with broad-spectrum antibiotics (e.g., ceftriaxone and vancomycin) within the first hour. Include antiviral therapy (e.g., acyclovir) if viral encephalitis is suspected. Lumbar puncture should be performed only if there is no clinical suspicion of increased ICP and the patient is stable.

**TABLE 1.10****ETIOLOGIES AND TARGETED EVALUATION OF ALTERED LEVEL OF CONSCIOUSNESS**

Category	Etiologies	Work-up
Drugs	Opiates (e.g., oxycodone, fentanyl, heroin) Sympathomimetics (e.g., cocaine, MDMA) Anticholinergics (e.g., diphenhydramine, TCAs) Cholinergics (e.g., organophosphates) Serotonin syndrome (e.g., SSRIs, dextromethorphan)	Urine toxicology screen Acetaminophen level ASA level Ethanol level ECG Blood gas Serum chemistry
Infection	Systemic sepsis Meningitis Encephalitis Abscess	Blood culture Complete blood count Urine analysis and culture CSF analysis and culture (if indicated)
Metabolic	Hypoglycemia Electrolyte abnormalities (e.g., hypernatremia/hyponatremia) Uremic encephalopathy Hyperammonemic encephalopathy Diabetic ketoacidosis Inborn error of metabolism Hepatic failure Renal failure	Blood gas Lactate Glucose Electrolytes Liver enzymes Renal function Ammonia Serum amino acids Urine organic acids Acylcarnitine profile Coagulation studies Serum/urine osmolarity
Structural	Space-occupying lesions (e.g., tumor, blood, abscess, cyst, cerebral edema secondary to trauma) Obstructions to cerebral blood flow (e.g., thrombus, vasculitis)	Head CT or MRI
Other	Anoxia Hypothermia/hyperthermia Seizure/postictal state Psychiatric/psychogenic	EEG

ASA, Acetylsalicylic acid (aspirin); CSF, cerebrospinal fluid; CT, computed tomography; ECG, electrocardiogram; EEG, electroencephalogram; MDMA, 3,4-Methylenedioxymethamphetamine (ecstasy); MRI, magnetic resonance imaging; SSRI, selective serotonin reuptake inhibitor; TCAs, tricyclic antidepressants.

Data from Krmpotic K. A clinical approach to altered level of consciousness in the pediatric patient. *Austin Pediatr*. 2016;3(5):1046.

- f. **Ingestion:** General management includes decreasing absorption, altering metabolism, and enhancing elimination.
  - (1) Contact the regional poison control center for specific treatment recommendations.
  - (2) See Chapter 3 for toxicology management.
- g. **Naloxone:** Administer opioid antagonist (full reversal: 0.1 mg/kg/dose IV/IM/subcutaneous [SC], max 2 mg/dose) if opioid ingestion

is suspected. Repeat dosing every 2 to 3 minutes. Short duration of action may necessitate multiple doses.

- h. **Thiamine:** Consider administration prior to hypertonic glucose for patients with eating disorders, chronic disease, or alcoholism to prevent Wernicke encephalopathy.
- i. If patient is an infant or toddler, consider evaluation for inborn error of metabolism, hepatic failure, renal failure, or nonaccidental trauma.

1

#### H. Status Epilepticus<sup>33-34</sup>

1. **Definition:** Prolonged seizure (clinical or electrographic) or recurrent seizure activity without return to baseline lasting **5 minutes** or more.
  - a. Common acute etiologies: febrile seizures, metabolic disturbances, sepsis, head trauma, stroke/hemorrhage, drug toxicity, inadequate antiepileptic therapy, hypoxia, hypertensive encephalopathy, autoimmune encephalitis
  - b. Common chronic etiologies: preexisting epilepsy, tumor, stroke, inborn error of metabolism, ethanol abuse
2. **Management:** Timely administration of anticonvulsant therapy is associated with a greater likelihood of seizure termination and better neurologic outcomes. See [Table 1.11](#) for timed evaluation and treatment outline.

**TABLE 1.11**  
**STATUS EPILEPTICUS TREATMENT GUIDELINE<sup>33-34</sup>**

#### IMMEDIATE APPROACH (0–5 min)

##### Management:

Protect airway, intubate if needed

Assess vitals

Bedside fingerstick blood glucose

Establish peripheral IV access: administer emergent AED, fluid resuscitation, nutrient resuscitation  
(thiamine, dextrose)

Labs: laboratory blood glucose, CBC, BMP, calcium, magnesium, antiseizure medication drug levels

Medication	Dose	Comment
Diazepam (Valium)	0.15–0.5 mg/kg IV (max 10 mg/dose) 2–5 years: 0.5 mg/kg PR (max 20 mg/dose) 6–11 years: 0.3 mg/kg PR (max 20 mg/dose) ≥12 years: 0.2 mg/kg PR (max 20 mg/dose) May repeat dose once in 5 min	Monitor for hypotension, respiratory depression
Lorazepam (Ativan)	0.1 mg/kg IV (max 4 mg/dose) May repeat dose once in 5–10 min	Monitor for hypotension, respiratory depression
Midazolam (Versed)	0.2 mg/kg IM/IN 0.5 mg/kg buccal Max: 10 mg all forms Single dose recommended	Monitor for hypotension, respiratory depression

*Continued*

**TABLE 1.11—CONT'D****URGENT APPROACH (5–15 min)****Management:**

Secondary AED control therapy

Initiate vasopressor support if indicated

Neurological examination

CT if indicated

Labs: Liver function tests, coagulation studies, toxicology screen, inborn error of metabolism

screening

Neurologic consultation

<b>Medication</b>	<b>Dose</b>	<b>Comment</b>
Fosphenytoin	20 mg PE/kg IV/IM (max 1500 mg PE/24 hr) May give additional 5 mg PE/kg repeat dose	Monitor for arrhythmia, hypotension
Levetiracetam (Keppra)	20–60 mg/kg IV (max 4500 mg/dose)	Minimal drug interactions Not hepatically metabolized
Phenytoin	20 mg/kg IV (max 1500 mg/24 hr) May give additional 5–10 mg/kg repeat dose	Monitor for arrhythmia, hypotension, purple glove syndrome
Phenobarbital	15–20 mg/kg IV (max 1000 mg) May give additional 5–10 mg/kg repeat dose	Monitor for hypotension, respiratory depression
Valproic Acid	20–40 mg/kg IV May give additional 20 mg/kg repeat dose (max 3000 mg/dose)	Use with caution in TBI Monitor for hyperammonemia, pancreatitis, hepatotoxicity, thrombocytopenia

**REFRACTORY APPROACH (15–60 min)****Management:**

Refractory AED control therapy

Continuous EEG monitoring if indicated

MRI if indicated

Lumbar puncture if indicated

Consider broad-spectrum antibiotics and antivirals if indicated

Intracranial pressure monitoring if indicated

Urinary catheter

<b>Medication</b>	<b>Dose</b>	<b>Comment</b>
Midazolam (continuous infusion)	Load: 0.2 mg/kg Infusion: 0.05–2 mg/kg/hr Breakthrough: 0.1–0.2 mg/kg bolus	Tachyphylaxis with prolonged use Monitor for respiratory depression, hypotension
Pentobarbital	Load: 5–15 mg/kg Infusion: 0.5–5 mg/kg/hr Breakthrough: 5 mg/kg bolus	Monitor for hypotension, respiratory depression, cardiac depression, paralytic ileus
Propofol	Load: 1–2 mg/kg Infusion: 20–65 mCg/kg/min Breakthrough: 1 mg/kg bolus	Monitor for hypotension, respiratory depression, cardiac failure, rhabdomyolysis, metabolic acidosis, renal failure, hypertriglyceridemia, pancreatitis (propofol related infusion syndrome)

AED, Antiepileptic drug; BMP, basic metabolic panel; CBC, complete blood count; CT, computed tomography; EEG, electroencephalogram; IM, intramuscular; IN, intranasal; IV, intravenous; mCg, microgram; PE, phenytoin equivalent; PR, per rectum; TBI, traumatic brain injury

## I. Increased Intracranial Pressure<sup>35-37</sup>

1. **Definition:** An increase in the volume of an intracranial component (brain, blood, or cerebrospinal fluid) within the fixed volume of the skull that exceeds the limits of compensation, generally accepted as a sustained increase  $\geq 20 \text{ mmHg}$ .
  - a. Intricately related to cerebral perfusion via the following equation:
$$\text{Cerebral perfusion pressure (CPP)} = \text{Mean arterial pressure (MAP)} - \text{ICP}$$
  - b. Most commonly caused by brain trauma, tumors, or intracranial infections.
  - c. Patients present with headache, diplopia, nausea, vomiting, or decreased level of consciousness.
  - d. Assess for signs of trauma, ataxia, pupillary asymmetry, papilledema, cranial nerve dysfunction, bulging fontanelle, or abnormal posturing.
    - (1) Foramen magnum herniation: hypertension, bradycardia, irregular respirations (Cushing triad)
    - (2) Transtentorial herniation: ipsilateral papillary dilation, contralateral hemiparesis
  - e. Evaluation may include infectious studies, electrolytes, toxicology screen, and stat CT head. Lumbar puncture is contraindicated due to herniation risk if cause is obstructive.
2. **Management:** Adequate CPP ( $>40 \text{ mmHg}$ ) is critical to overcome the resistance of increased ICP.
  - a. Stabilize initially as per resuscitation guidelines.
    - (1) Maintain normal oxygenation and ventilation to treat increased metabolic demand and avoid hypercarbia-related cerebral vasodilation.
    - (2) Consider hyperventilation (EtCO<sub>2</sub> target between 25 and 30) for patients with **active** evidence of herniation. Prophylactic hyperventilation is otherwise not recommended.
    - (3) Support MAP with adequate isotonic fluid resuscitation and vasoactive agents.
  - b. Consultation with neurosurgical team is recommended and required immediately if evidence of herniation is present.
  - c. Administer **mannitol** (0.25 to 1 g/kg) and/or **hypertonic saline** (5 to 10 mL/kg of 3% hypertonic saline) in case of acute neurologic deterioration or cerebral herniation.
    - (1) Continuous infusions of 3% hypertonic saline (0.5 to 1.5 mL/kg/h) may be titrated as necessary to maintain ICP less than 20 mmHg.
    - (2) Rapid osmotic diuresis from mannitol may cause hypovolemia and hypotension, especially in polytrauma patients.
  - d. Request **noncontrast head CT** to evaluate for emergent surgical pathology.
  - e. Treat acute seizure activity given the associated increased cerebral metabolic rate and subsequent increased cerebral blood flow. Consider prophylactic antiseizure therapy (e.g., phenytoin, levetiracetam), if transport or delayed definitive care is anticipated.

- f. Sedation and analgesia prevent increases in ICP related to pain and agitation, although benefit is balanced with risk of hypotension and alteration of neurologic exam.
- g. Avoid secondary brain injury by maintaining neuroprotective parameters: Maintain head midline and elevated at 30 degrees, normoglycemia, normonatremia, normothermia, and correct acidosis.
- h. If elevated ICP is refractory to medical management, consider draining an existing ventriculoperitoneal shunt or acute neurosurgical intervention (external ventricular drain or decompressive craniectomy).
- i. For elevated ICP refractory to medical and surgical management, consider barbiturate coma.

#### IV. CRITICAL CARE REFERENCE DATA

1. Minute ventilation ( $V_E$ ):

$$V_E = \text{Respiratory rate} \times \text{Tidal volume } (V_T)$$

2. Alveolar gas equation:

$$P_{A\text{O}_2} = [\text{FiO}_2 (\text{P}_{\text{atm}} - \text{PH}_2\text{O})] - (\text{P}_a\text{CO}_2/\text{R})$$

- a.  $P_{A\text{O}_2}$  = Alveolar partial pressure of oxygen
- b.  $\text{FiO}_2$  = Inspired fraction of oxygen (0.21 at room air)
- c.  $\text{P}_{\text{atm}}$  = Atmospheric pressure (760 mmHg at sea level; adjust for high altitude)
- d.  $\text{PH}_2\text{O}$  = Water vapor pressure (47 mmHg)
- e.  $\text{P}_a\text{CO}_2$  = Arteriolar partial pressure of carbon dioxide (measured via arterial blood gas)
- f. R = Respiratory quotient (0.8;  $\text{CO}_2$  produced/ $\text{O}_2$  consumed)

3. Alveolar-arterial oxygen gradient (A-a gradient):

$$\text{A-a gradient} = P_{A\text{O}_2} - P_a\text{O}_2$$

- a.  $P_{A\text{O}_2}$  = Alveolar partial pressure of oxygen (estimated from alveolar gas equation)
- b.  $P_a\text{O}_2$  = Arteriolar partial pressure of oxygen (measured via arterial blood gas)
- c. Normal gradient is 20 to 65 mmHg on 100% oxygen or 5 to 20 mmHg on room air
- d. The A-a gradient is increased in hypoventilation, diffusion limitations, pulmonary blood-flow shunts and ventilation/blood flow (V/Q) mismatch.

4. Oxygenation index (OI):

$$OI = P_{aw} \times \text{FiO}_2 \times 100 / P_a\text{O}_2$$

- a.  $P_{aw}$  (mmHg) = Mean airway pressure
- b.  $OI > 40$  in hypoxic respiratory failure is historically considered an indication for extracorporeal life support.

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# Chapter 2

## Traumatic Injuries

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 See additional content on Expert Consult

### I. COMPONENTS OF THE TRAUMA ASSESSMENT

#### A. Primary Survey

1. The primary survey includes assessment of ABCDE (airway, breathing, circulation, disability, exposure/exsanguination). This includes intravenous (IV) access, preferably two large-bore catheters.
2. **NOTE:** The Advanced Trauma Life Support algorithm developed by the American College of Surgeons continues to support the ABC sequence in the primary survey. For nontraumatic cardiorespiratory arrest, the circulation, airway, and breathing (CAB) sequence is currently in use by the American Heart Association as part of the Pediatric Advanced Life Support algorithm (see [Chapter 1](#)).

#### B. Secondary Survey ([Fig. 2.1](#))

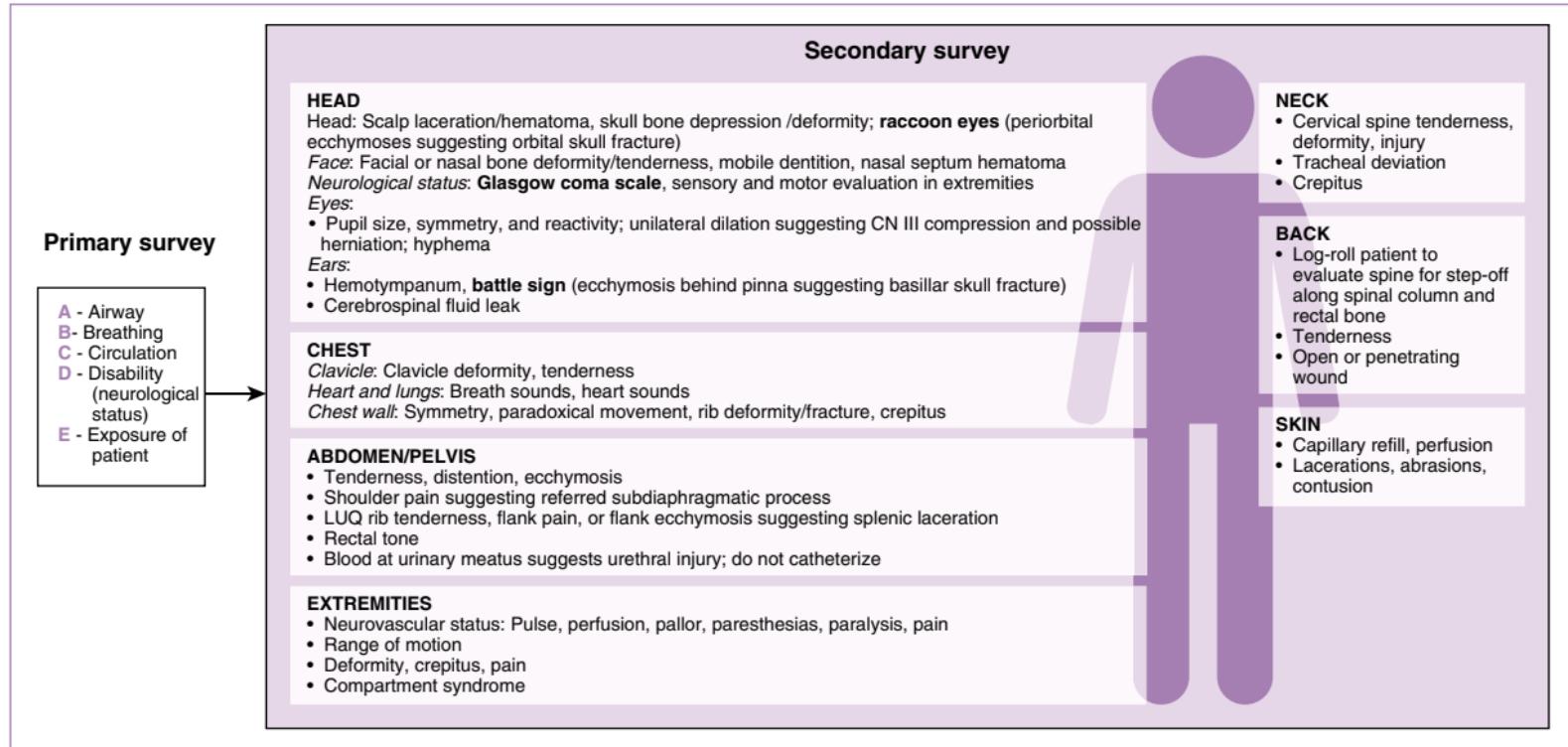
### II. HEAD AND NECK TRAUMA

#### A. Head Imaging

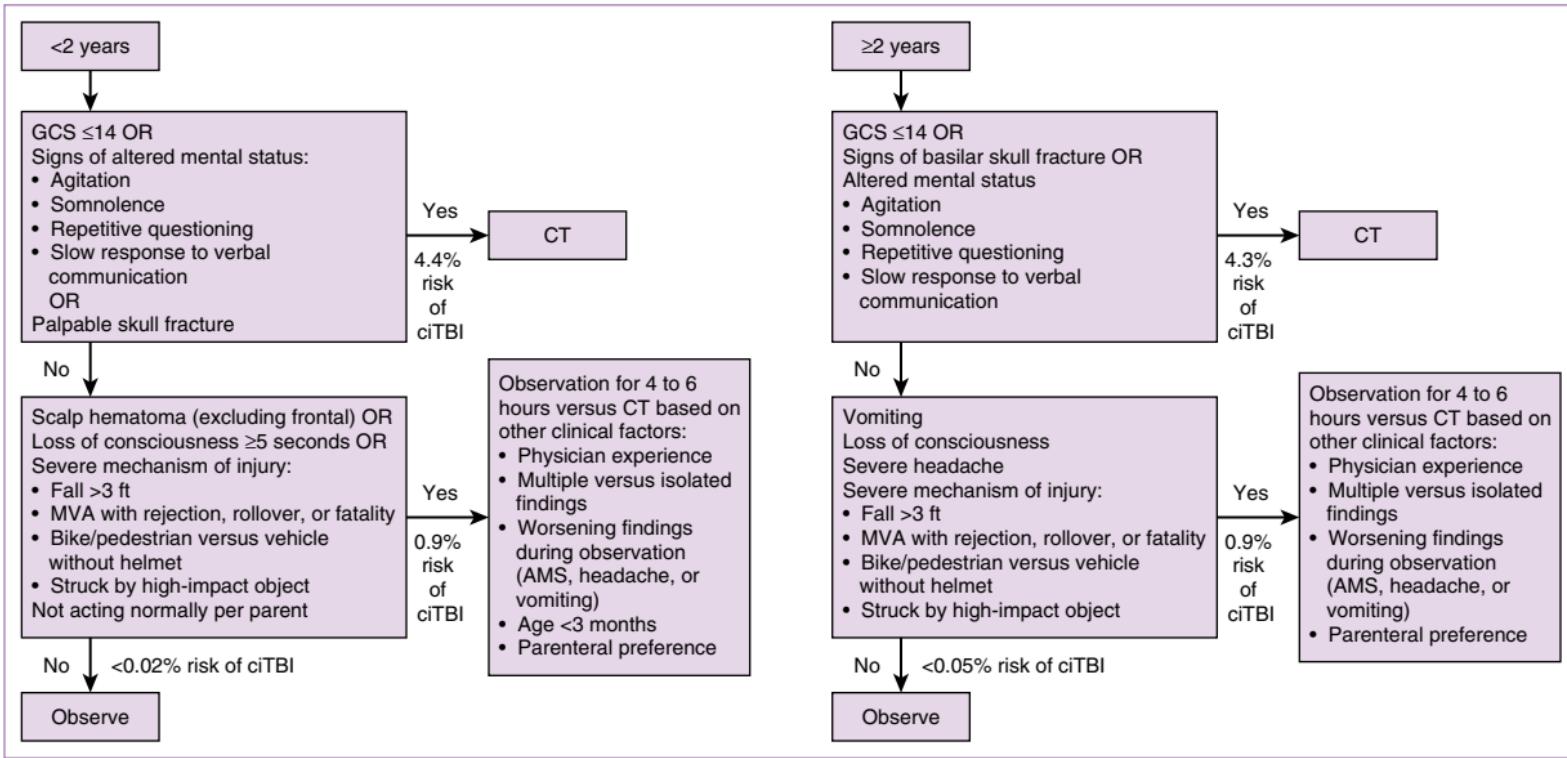
1. The PECARN algorithm ([Fig. 2.2](#)) is often used to assess risk for clinically important traumatic brain injury.<sup>1</sup>
2. If signs of traumatic brain injury on computed tomography (CT), consider consultation by pediatric neurosurgery/trauma surgeon.

#### B. Cervical Spine and Neck Imaging

1. There are currently no unified protocols or clinical guidelines for pediatric cervical spine clearance after blunt trauma.
2. Based on PECARN C-Spine criteria,<sup>2</sup> consider obtaining imaging if any of the following are present in a patient  $\leq 16$  years old:
  - a. Altered mental status
  - b. Focal neurologic deficits
  - c. Complaint of neck pain
  - d. Torticollis
  - e. Substantial injury to the torso
  - f. Predisposing condition
  - g. High-risk motor vehicle crash
  - h. Diving accident
3. Note, many institutions alternatively use NEXUS criteria for clinical c-spine clearance. This is validated in children  $\geq 8$  years old<sup>3</sup> and includes #1, 2, and 3 of PECARN c-spine plus presence of intoxication or painful, distracting injury.<sup>4,5</sup>

**FIGURE 2.1**

Trauma primary and secondary survey.



**FIGURE 2.2**

Recommended algorithm for obtaining head computed tomography in children after head trauma by age. (From Kuppermann N, Holmes JF, Dayan PS, et al. Identification of children at very low risk of clinically-important brain injuries after head trauma: a prospective cohort study. The Lancet. 2009;374(9696):1160–1170.)

4. Recent guidelines by the Pediatric Cervical Spine Clearance Working Group Algorithm<sup>6</sup> additionally highlight the following factors:
  - a. In patients  $\leq 3$  years old, consider plain radiographs, if c-spine cannot be cleared clinically.
  - b. Clinical clearance can be done regardless of mechanism of injury if a child is  $\geq 3$  years and is asymptomatic with normal mental status and normal physical examination.
  - c. Clinical clearance CANNOT be performed if the child is observed to have or reports persistent neck pain, or if there is abnormal head posture or difficulty in neck movement.

### C. Specific Imaging Studies

1. C-spine x-ray (XR) with minimum of two views (lateral, anteroposterior, and/or odontoid views) (90% sensitivity in identifying bony cervical spine injury).<sup>7</sup>
2. Consider further cross-section imaging for further evaluation of vertebral cervical fracture.<sup>8</sup>
3. Consider magnetic resonance imaging (MRI) scan for further evaluation of ligamentous and cord cervical spine injury.<sup>9,10</sup>
4. Spinal cord injury without radiographic abnormality (SCIWORA): Neurologic symptoms persist with no radiographic abnormality. Of note, recent research found that MRI revealed abnormal features only in those patients with complete neurologic deficits and may lack sensitivity with abnormal features associated with partial or temporary neurologic deficits.<sup>11,12</sup>
5. If signs of spinal column or vascular injury on imaging, consider consultation by trauma, spine, and/or neck surgeon.

## III. CONCUSSION

### A. Concussion Evaluation

1. The Acute Concussion Evaluation (ACE) can be used in multiple settings (see [Section XI. Resources](#)), including the clinic and emergency department (ED).<sup>13–15</sup>
2. Patients should be referred to a concussion specialist if symptoms persist greater than 10 to 14 days, if they worsen, or if a patient has a history of multiple concussions.

### B. Return-to-school and Return-to-play Guidelines ([Table 2.1](#))

1. Overarching goal is to allow healing from first injury in an attempt to prevent “second impact syndrome”: Diffuse cerebral swelling in the setting of a second concussion that occurred while still symptomatic from an earlier concussion. This is a rare but potentially fatal complication of concussions.
2. Consider providing the ACE Care Plan for parent and child guidance (see [Section XI. Resources](#)).
3. Brain rest: Although evidence-based guidelines for brain rest following concussion are limited, current research suggests that extreme rest (i.e., bed rest) can hinder recovery from concussions.<sup>16</sup> Other studies

**TABLE 2.1****RETURN-TO-PLAY AND RETURN-TO-SCHOOL<sup>15</sup>**

<b>Return-to-Play Guidelines</b>	<b>Return-to-School Guidelines</b>
<b>BRIEF GUIDELINES</b>	<b>BRIEF GUIDELINES<sup>41</sup></b>
Each step should last a minimum of 24 hr. Move to the next level of activity only if no symptoms are experienced. If symptoms return, patients should stop activities and notify a health professional. After evaluation, once the patient has not had symptoms for minimum 24 hr, patients should resume play at the previous tolerated step of the return to play guidelines.	If symptoms affect concentration or if unable to tolerate stimulation for more than 30 min without symptoms, consider remaining at home with light mental activities (watching TV, light reading, and interaction with the family), so long as they do not provoke symptoms. Minimize computer use, texting, and video games. If able to tolerate stimulation for minimum of 30–45 min without symptoms, consider returning to learning with modifications. Providers should provide school notes.
Step 1: No physical activity Step 2: Low levels of physical activity Examples: Walking, light jogging, light stationary biking, light weightlifting (lower weight with higher repetitions, no bench, no squat) Step 3: Moderate levels of physical activity with body/head movement Examples: Moderate jogging, brief running, moderate-intensity stationary biking, moderate-intensity weightlifting (reduced time and/or reduced weight from typical routine) Step 4: Heavy noncontact physical activity Examples: Sprinting/running, high-intensity stationary biking, regular weightlifting routine, noncontact sport-specific drills (in three planes of movement) Step 5: Full contact in controlled practice Step 6: Full contact in game play	<b>SUGGESTED SCHOOL MODIFICATIONS:</b> Shortened school days Frequent breaks during classes Extra time to complete coursework/assignments and tests Decreased homework load No significant classroom or standardized testing at this time Consider 504 Plan and/or Individualized Education Plan (IEP)

have found that some degree of cognitive rest can be beneficial and that patients presenting with signs of injury following concussion (e.g., loss of consciousness, posttraumatic amnesia) are more likely to benefit from rest following concussion than those patients presenting with symptoms alone (somatic, cognitive, affective, and sleep-related symptoms).<sup>17</sup>

- For further guidelines, please discuss with concussion specialist.

**IV. THORACIC AND ABDOMINAL TRAUMA EVALUATION<sup>18</sup>****A. Physical Exam**

“Seat belt sign” is a significant predictive factor for surgical abdominal injury after blunt trauma (sensitivity 70.6%, specificity 82.4%).<sup>19</sup>

**B. Laboratory Studies to Consider**

Type and cross-match, complete blood cell count (CBC; low hemoglobin indicates possible hemorrhage; however, this is a late sign), electrolytes,

liver function tests (high AST/ALT indicate liver injury), lipase (high level indicates pancreatic injury), and urinalysis (hematuria indicates possible renal/bladder injury).

### C. Imaging Studies to Consider

1. Chest radiograph
  - a. Look for rib fracture, pneumothorax and/or hemothorax, pulmonary contusion, pneumomediastinum.
  - b. Consider chest CT with IV contrast, if recommended by radiologist and/or trauma surgeon.
2. Pelvis radiograph
  - a. Look for pelvis fracture.
  - b. Consider pelvis CT if recommended by radiologist and/or trauma surgeon.
3. Abdominal/pelvis CT with IV contrast
  - a. This is the “gold standard” for intra-abdominal injury diagnosis; however, radiographs should be obtained first if there is concern for additional injuries that would compromise clinical stability.
  - b. For blunt abdominal trauma, routine oral contrast is not indicated, whereas IV contrast can help to identify visceral, vascular, or bowel injury.<sup>20</sup>
  - c. For penetrating abdominal trauma, triple contrast (oral, rectal, IV) CT to identify peritoneal penetration or intra-abdominal organ injury in stable stab wound victims.<sup>21</sup>
  - d. Look for duodenal hematoma, hemoperitoneum, bladder injury, solid organ hemorrhage (e.g., spleen and/or liver).
  - e. If gross hematuria or urinalysis with greater than 50 RBC/hpf, consider genitourinary tract trauma and consider CT abdomen and pelvis with and without IV contrast (CT urography) and CT cystogram, in consultation with radiology/urologist/trauma surgeon.
4. Extended focused assessment with sonography for trauma (eFAST)
  - a. Can help to identify intra-abdominal free fluid and parenchymal injury (sensitivity 50%, specificity 85%).<sup>22</sup>
  - b. eFAST with bilateral anterior lung views is highly sensitive for pneumothorax.
  - c. Consider performing if qualified personnel available.
5. If any workup is positive for thoracic or abdominal trauma, immediate consultation with nearest pediatric trauma center/surgeon is indicated.

## V. ORTHOPEDIC/LONG BONE TRAUMA

### A. Physical Exam

1. Look for swelling, ecchymosis, or deformity. Look for breaks in the skin (abrasions, lacerations) overlying the apex of the fracture suggestive of open fracture.
2. Bleeding
  - a. Consider arterial bleed if absent pulses and cool extremity with bleeding.
  - b. Consider venous bleed if persistent pulse with bleeding.

3. Compartment syndrome: Tense, swollen area at site of injury, pain, paresthesia, paresis, pallor, pulselessness (if unable to palpate pulse, consider using vascular ultrasound with Doppler).
4. If signs/symptoms of compartment syndrome or open fracture, consultation with a pediatric orthopedic surgeon is recommended.

### B. Imaging

1. Children's bones are less densely calcified, have thickened periosteum, and have a growth plate, all of which increase their vulnerability to fractures.
2. Obtain radiographs if bony point tenderness or deformity, decreased sensation, decreased range of motion, or overlying skin discoloration.
3. Radiographs with anterior-posterior and lateral views ± oblique and including areas above and below the suspected area of injury are recommended.

### C. Fractures Unique to Children

1. Physeal or Salter-Harris fractures<sup>18</sup>: Fractures involving growth plates (see [Chapter 26](#)).
2. Plastic fractures: Pliability of bones in response to compressive and transverse forces.
  - a. Torus or buckle fracture: Compression injury with buckled cortex
  - b. Greenstick fracture: Fracture on one side of the diaphysis with cortex intact on other side of diaphysis
  - c. Bowing or bending fractures
3. Avulsion fractures: Tendon or ligament dislodging a bone fragment. These are more common among adolescents participating in sports.

### D. Fractures Requiring Urgent Orthopedic Surgeon Consultation

1. Open fractures
2. Unacceptably displaced fractures
3. Fractures with associated neurovascular compromise (consider emergent reduction to improve neurovascular status if orthopedic surgery is not available on-site)
4. Significant growth plate or joint injuries
5. Complete or displaced fractures of the long bones of the extremities
6. Pelvic fractures (other than minor avulsions)
7. Spinal fractures
8. Dislocations of major joints other than the shoulder

### E. Fractures That Are Appropriate to Manage Acutely With Outpatient Referral to Orthopedics ([Table 2.2](#))

## VI. DENTAL TRAUMA

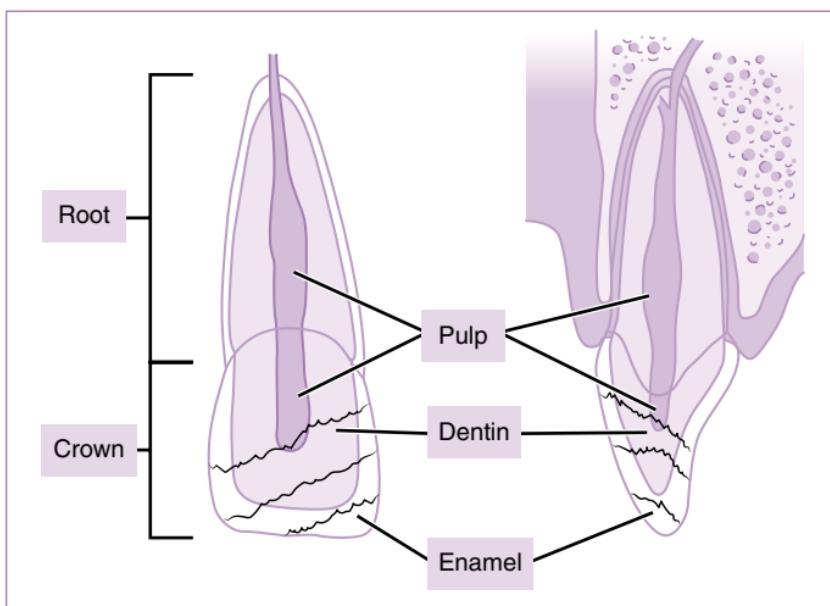
### A. Components of a Tooth ([Fig. 2.3](#))

### B. Differences Between Primary and Permanent Teeth ([Fig. 2.4](#))

1. Primary teeth appear 6 months to 3 years of age, are relatively smaller, whiter, front teeth have a smooth biting surface.

**TABLE 2.2****COMMON PEDIATRIC ORTHOPEDIC INJURIES AND MANAGEMENT**

Injury	ED Management	Follow-up
Clavicle fracture without tenting or displacement (if present, orthopedic surgery consultation required)	Sling	Primary care provider in 2 weeks
Acromioclavicular joint separation	Sling	Orthopedics in 1 week
Proximal humerus fracture WITHOUT deformity, displacement, neurovascular injury	Sling	Orthopedics within 1 week
Distal radius or ulna fracture WITHOUT deformity, displacement, neurovascular injury	Volar splint	Orthopedics within 1 week
Salter-Harris Type 1—Distal radius	Volar splint	Orthopedics within 1 week
Salter-Harris Type 1—Distal fibula	Posterior splint, crutches	Orthopedics within 1 week

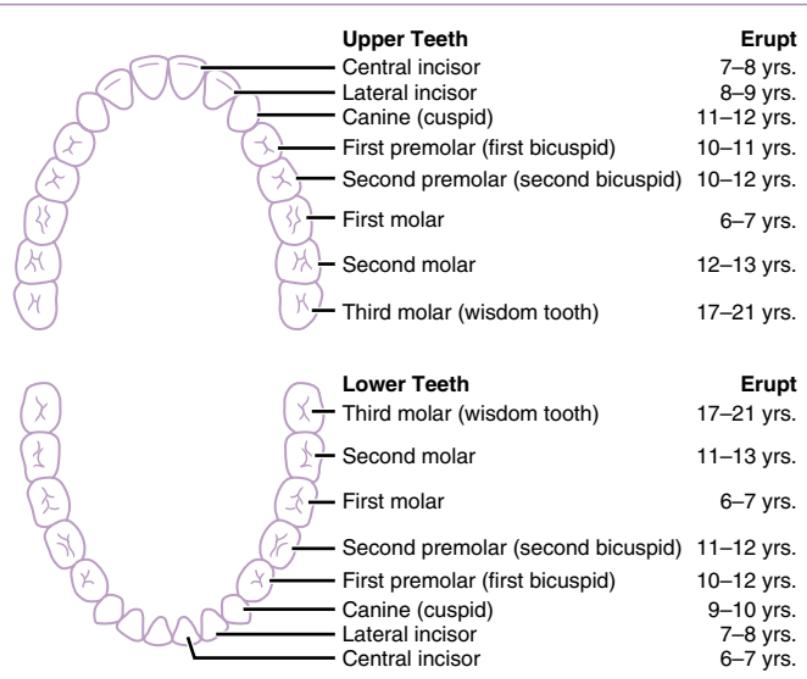
**FIGURE 2.3**

Normal anatomy of a tooth. (Modified from Textbook of Pediatric Emergency Medicine.<sup>18</sup>)

2. Permanent teeth appear 6 years to 21 years of age, relatively larger, front teeth have a ridged biting surface.

### C. Dental Injuries

1. Avulsion
  - a. An avulsion injury involves complete displacement of the tooth from the alveolar socket.<sup>23</sup>
  - b. If a primary tooth, outpatient dental follow-up is appropriate.
  - c. If a permanent tooth, this is a dental emergency!

**FIGURE 2.4**

Development from primary to permanent teeth by location. (Modified from American Dental Association. [www.mouthhealthy.org](http://www.mouthhealthy.org))

- d. **Most important: Immediate reimplantation should occur within 60 minutes to maximize tooth viability. Without a tooth present, the periodontal ligament can degenerate.**
- e. Method:
  - (1) Pick up the avulsed tooth by the crown and avoid touching the root to prevent injury to the periodontal ligament.
  - (2) Wash the tooth briefly with saline or Hanks Balanced Salt Solution (HBSS).
  - (3) Administer local lidocaine into the gum if time permitting.
  - (4) Insert the root into the alveolar socket with concave part facing the tongue.
  - (5) Ask the patient to bite on gauze to hold it in position.
  - (6) **Refer to a dentist emergently for splinting.**
- f. Reimplantation should always be attempted. If reimplantation is not possible, place the tooth in a container in osmolality balanced media (e.g., HBSS, cold milk) and **refer to a dentist emergently for reimplantation and splinting.**
- 2. Luxation
  - a. Luxation injuries result from physical displacement of tooth within the alveolar socket, tearing of the periodontal ligament with possible injury to the alveolar bone.<sup>24</sup>

**TABLE 2.3****TOOTH FRACTURE TYPES AND FOLLOW-UP RECOMMENDATIONS<sup>24,25</sup>**

Fracture Type	Follow-up recommendations
Enamel Fracture	Dental evaluation outpatient for possible binding of tooth fragment, if available
Enamel-Dentin Fracture	Dental evaluation 48–72 hr to place a dressing of calcium hydroxide to prevent injury to the pulp
Enamel-Dentin-Pulp Fracture	Immediate dental evaluation within 48 hr
Alveolar Ridge Fracture	Emergent dental evaluation

- b. Primary tooth: If tooth is loose, there is an increased risk of aspiration, and the tooth may be extracted with firm pressure with gauze. If tooth is not loose, may need repositioning and splinting. In both situations, refer to a dentist for evaluation within 48 hours.
  - c. Permanent tooth: Immediate dental evaluation required if significant tooth mobility; otherwise, outpatient evaluation within 48 hours is appropriate.
3. Subluxation
- a. Subluxation is characterized by tooth injury with minor mobility without displacement.
  - b. Regardless of whether permanent or primary tooth, outpatient dental follow-up, ideally within 48 hours, is needed to rule out root fracture.
4. Tooth fracture:
- a. Classify the fracture per involvement of enamel, dentin, and pulp.<sup>25</sup>
  - b. For management guidelines: [Table 2.3](#).
- D. Anticipatory Guidance Following Dental Trauma**
1. Avoid contact sports
  2. Analgesics as needed for pain control (e.g., acetaminophen, ibuprofen, cold compresses)
  3. Soft diet
  4. Use a soft toothbrush, if able to brush teeth
  5. Regular follow-up with a dentist

**VII. OPHTHALMOLOGIC TRAUMA<sup>26</sup>****A. Chemical Injury to the Eye<sup>18</sup>**

1. Determine if substance is an acid or alkali. Alkali solutions tend to be more damaging because they penetrate more deeply.
2. Obtain a baseline pH by touching Litmus paper to the conjunctiva.
3. Retract eye lids as much as possible and irrigate immediately with normal saline or lactated Ringer solution. This can be performed at the eyewash station or with a standard bag of fluid with tubing placed at the medial canthus. Allow the liquid to pass over the open eye to the lateral canthus.

4. Continue irrigation for a minimum of 30 minutes with minimum 1 to 2 L of solution or until pH becomes neutral (7.0 to 7.4). Additional fluid may be required.
5. Monitor conjunctival pH with Litmus paper 10 to 20 minutes after irrigation.<sup>27</sup>
6. Consider ophthalmologic consultation and discuss with Poison Control.

#### B. Ruptured Globe

1. A ruptured globe is caused by laceration or puncture of the cornea and/or sclera following trauma caused by projectile, sharp, or blunt trauma.
2. Key physical exam findings include: Teardrop shaped pupil pointing towards perforation, hyphema (hemorrhage in the anterior chamber) and/or subconjunctival hemorrhage, severe pain, decreased visual acuity, edema.
3. Stop the exam and place a rigid eye shield.
4. Elevate the head of the bed.
5. Keep patient as calm as possible and control symptoms (e.g., antiemetics and pain control) to avoid increased globe pressure and further extrusion of vitreous/aqueous humor.
6. Immediately consult ophthalmology and administer antibiotics.

#### C. Corneal Abrasion

1. Key physical exam findings include red eye with tearing, intense pain, resistance to eye opening, photophobia, foreign body sensation.
2. Consider application of topical anesthetic before examination. If foreign body sensation is present on your exam, evert eyelids to look for retained foreign body.
3. Apply fluorescein staining and examine with Wood lamp. Focal uptake indicates abrasion.
4. Consider ophthalmic ointment or artificial tears for lubrication and pain relief.
5. Consider ophthalmologic consultation in the ED if concern for larger corneal abrasions involving visual axis, corneal laceration, ulceration, embedded foreign body, or prolonged healing (i.e., symptoms not improving after several days).

#### D. Superglue to the Eye<sup>28</sup>

1. Trim eyelashes as needed with blunt-tip scissors.
2. Apply copious amounts of ointment, such as bacitracin ophthalmic ointment or baby shampoo, and gently massage eyelashes to break down the glue. Advise that the patient continue this as often as possible. Dissolution of glue may take several days.
3. Consider consultation with ophthalmology if several days of ointment is unsuccessful.

#### E. Eyelid Laceration

1. Consider consultation with ophthalmology if: Full-thickness lacerations (exposed adipose tissue), laceration through the lid margin or tarsal plate,

lacerations involving lacrimal canaliculi (medial third of the upper/lower lids), or ptosis (unequal lifting of lids with upward gaze would suggest this).

2. Some superficial lacerations that occur in the direction of a natural skin fold may not require repair.

#### F. Orbital Floor Fractures

1. This injury is usually caused by blunt trauma and is often referred to as a “blow out fracture,” because the weakest area of the orbital bones is the orbital floor/maxillary roof.
2. Key physical exam findings include: Eyelid swelling, ecchymosis, enophthalmos of affected eye, ptosis, diplopia, anesthesia of the cheek (involvement of infraorbital nerve), decreased extraocular eye movements (especially decreased superior range of the globe due to inferior rectus entrapment).
3. Evaluate for other eye injuries (e.g., retinal trauma, ruptured globe).
4. Consider consultation with ophthalmology and plastics/otorhinolaryngology surgeon.

#### G. Other Instances Requiring Ophthalmologic Consultation

1. Traumatic iritis is associated with blunt trauma with painful red eye, pupillary constriction, and photophobia, often with delayed presentation of symptoms (24 to 72 hours) after trauma.
2. Sudden loss of vision could suggest retrobulbar hemorrhage or retinal detachment.

### VIII. ANIMAL BITES

#### A. Wounds at the Highest Risk of Infection

1. Bites over hand, foot, genitalia, or joint surface
2. Bites from a cat or human
3. Wounds in an asplenic or immunocompromised patient
4. Wounds with delayed presentation to care >12 hours

#### B. Decision to Suture

1. Avoid closing wounds at high risk of infection (see earlier) unless for cosmetic reasons, large wounds or wounds with edges far apart where loose approximation can be helpful.
2. Wounds on head and neck can be safely sutured after copious irrigation and wound débridement if within 6 to 8 hours of injury and there are no signs of infection. Avoid skin glue due to high risk of infection.
3. In large wounds, subcutaneous dead space should be closed with a minimal number of absorbable sutures, with delayed closure in 3 to 5 days, if there is no evidence of infection.
4. Wounds that involve tendons, joints, deep fascia, or major vasculature should be evaluated by a surgeon.

#### C. Antibiotic Prophylaxis<sup>29</sup>

1. Table 2.4.

**TABLE 2.4****ANTIBIOTIC MANAGEMENT OF ANIMAL AND HUMAN BITES**

Type of Bite	Organisms	Treatment
Animal bite	<i>Staphylococcus aureus</i> , <i>Streptococci</i> , Oral Anaerobes, <i>Pasteurella</i> , <i>Capnocytophaga canimorsus</i>	Amoxicillin/clavulanate for 5 days TMP/SMX and clindamycin, if allergy to penicillin
Human bite	<i>Streptococcus viridans</i> , <i>S. aureus</i> , Oral Anaerobes, <i>Eikenella corrodens</i>	Amoxicillin/clavulanate for 5 days Clindamycin AND ciprofloxacin, if allergy to penicillin

- Consider IV antibiotics if patient is critically ill or unable to tolerate PO intake.

**D. Tetanus Postexposure Prophylaxis: See Chapter 16**

**E. Rabies Postexposure Prophylaxis: See Chapter 16**

## IX. BURNS

**A. Burns That Should Prompt Consideration of Elective Intubation**

- Signs of inhalational injury (e.g., singed nasal hairs, soot at the nares, oropharyngeal erythema)
- Early onset stridor
- Severe burns of face and/or mouth
- Progressive respiratory insufficiency

**B. Estimation of the Surface Area of Burns**

- See Fig. 2.5.
- Only include partial- and full-thickness burns and exclude superficial burns in the calculation of body surface area.

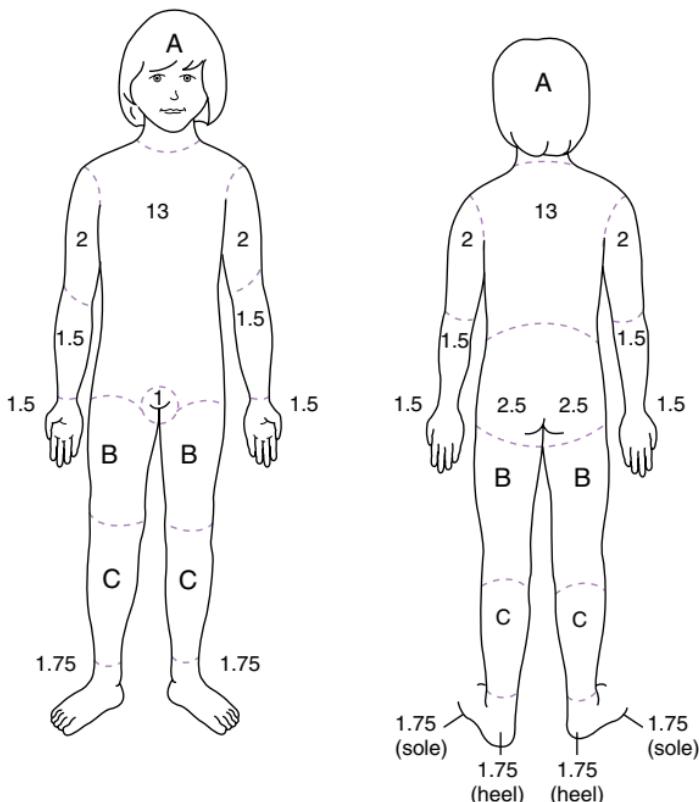
**C. Estimation of the Depth of Burns (Table 2.5)**

**D. Fluid Resuscitation in Patients With Burns (Fig. 2.6)**

- Consider central venous access for burns greater than 25% BSA.
- Withhold potassium from IV fluids generally for the first 48 hours because of a large release of potassium from damaged tissues.
- Foley catheter placement is recommended to monitor urine output during fluid resuscitation phase.

**E. Indications for Transfer to a Burn Center<sup>30</sup>**

- ≥10% partial-thickness and/or full-thickness burns
- ≥5% full-thickness burns
- If burn débridement is warranted (e.g., any partial-thickness burn >2 cm in diameter)
- Respiratory involvement and/or major trauma
- Electrical, chemical, or inhalation injury
- Burns of critical areas, such as face, hands, feet, perineum, or joints
- Circumferential burns

**FIGURE 2.5**

Burn assessment chart. All numbers are percentages. (Modified from Barkin RM, Rosen P. Emergency Pediatrics: A Guide to Ambulatory Care. 6th ed. St. Louis: Mosby; 2003.)

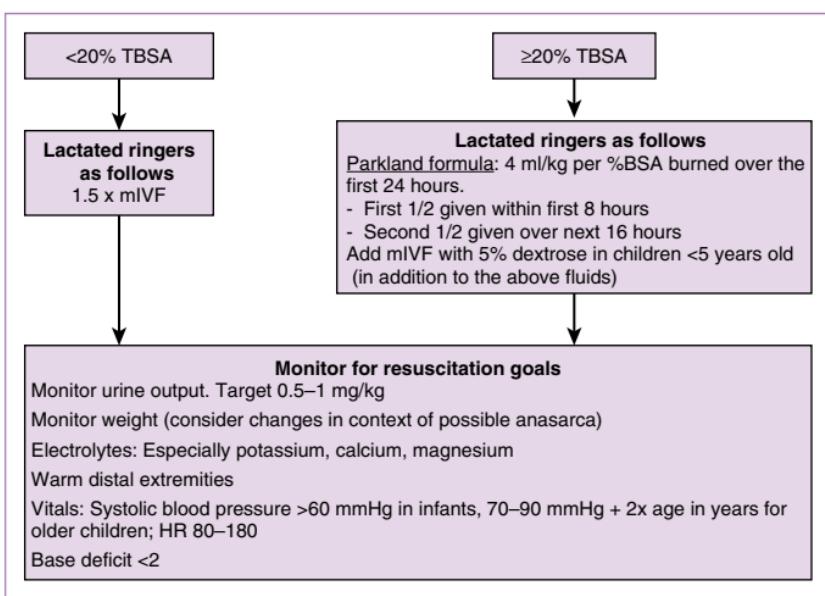
8. Patient with underlying chronic illness
9. Suspicion of abuse or unsafe home environment

#### **F. Management of Burns Not Referred to Burn Center**

1. For a partial-thickness burn not requiring débridement:
  - a. Clean with warm saline or mild soap and water.
  - b. Apply topical antibacterial agent such as bacitracin (requires daily dressing changes) or silver-impregnated dressings (dressing can be

**TABLE 2.5****BURN CLASSIFICATION**

Wound Depth	Layer Involved	Clinical Findings
Superficial	Epidermis	Dry, painful, erythematous (like a sunburn)
Partial Thickness	Dermis	Moist, painful, erythematous Blistering present, blanches Disruption of nails, hair, sebaceous glands
Full Thickness	Subcutaneous, fascia, muscle, bone	Pale, charred, waxy, leathery, insensate No bleeding or blanching

**FIGURE 2.6**

Formulaic fluid resuscitation for pediatric burns.<sup>18</sup>

- left in place until follow-up) and cover with nonadherent dressing.
- Follow-up inspections of wound should occur at 24 and 72 hours.
  - Follow-up within one week at a pediatric burn center is highly recommended.
  - Oral antibiotics are not indicated.

#### G. Other Special Considerations With Burns

- Circumferential burns can increase risk of compartment syndrome.
- Tetanus prophylaxis is warranted with burns. Refer to Chapter 16 for details.

#### H. Other Types of Burns

- Household electrical burn<sup>31</sup>: In general, household outlets are 120 to 240 V and rarely cause serious injuries or cardiac arrhythmias.
- High-voltage burns (>1000 V), including lightning burns:
  - Patients are at increased risk of ventricular arrhythmias or asystole. Consider cardiac monitoring for 48 hours.<sup>31</sup>
  - Patients are also at increased risk of compression spine fractures or

spinal cord injury due to tetany, as well as compartment syndrome, rhabdomyolysis, and hyperkalemia due to muscle swelling.

## X. NONACCIDENTAL TRAUMA

### A. Physical Abuse

#### 1. Red flags in history

- a. Delay in presentation
- b. Inconsistent/incomplete/vague/changing explanations for significant injury
- c. History is inconsistent with age, pattern, or severity of the injury
- d. History is inconsistent with child's physical or developmental capabilities
- e. Different witnesses provide different explanation

#### 2. Concerning physical exam findings<sup>32</sup>

- a. Bruises: In protected areas (chest, abdomen, back, buttocks), multiple, in various stages of healing, those that do not fit history or developmental stage of child, in unusual places (e.g., postauricular, neck, inner aspect of arms), those consistent with slap of hand or pinch.
- b. Burns: Multiple, well-demarcated, stocking/glove distribution, symmetrically burned palms/soles, buttocks and/or lower legs, mirror image burns of extremities, spared inguinal or other flexural creases, appearance of a cigarette burn.
- c. Other: Frenulum tears, loop marks from cord or cable, bites.
- d. See [Figs. 2.7–2.10](#) (color plates) and [Figs. EC 2.A-D](#) for examples.

#### 3. Imaging guidelines

##### a. Skeletal survey<sup>33–35</sup>

- (1) In children less than 2 years of age, use skeletal survey to evaluate for bony injury. This includes frontal and lateral views of the skull, lateral views of the cervical spine and thoracolumbosacral spine, and single frontal views of the long bones, hands, feet, chest, and abdomen.



**Figure EC 2.A**

Bite mark outlining the dental arch. (Modified from Zitelli BJ, McIntire SC, Nowalk, AJ. Atlas of Pediatric Physical Diagnosis. 7th ed. Philadelphia: Elsevier; 2018.)<sup>39</sup>



**Figure EC 2.B**

Cigarette burn appearing as a circular punched out lesion. (Modified from Zitelli BJ, McIntire SC, Nowalk, AJ. Atlas of Pediatric Physical Diagnosis. 7th ed. Philadelphia: Elsevier; 2018.)<sup>39</sup>



**Figure EC 2.C**

Loop marks from a cord or cable. (Modified from Zitelli BJ, McIntire SC, Nowalk, AJ. Atlas of Pediatric Physical Diagnosis. 7th ed. Philadelphia: Elsevier; 2018.)<sup>39</sup>



**Figure EC 2.D**

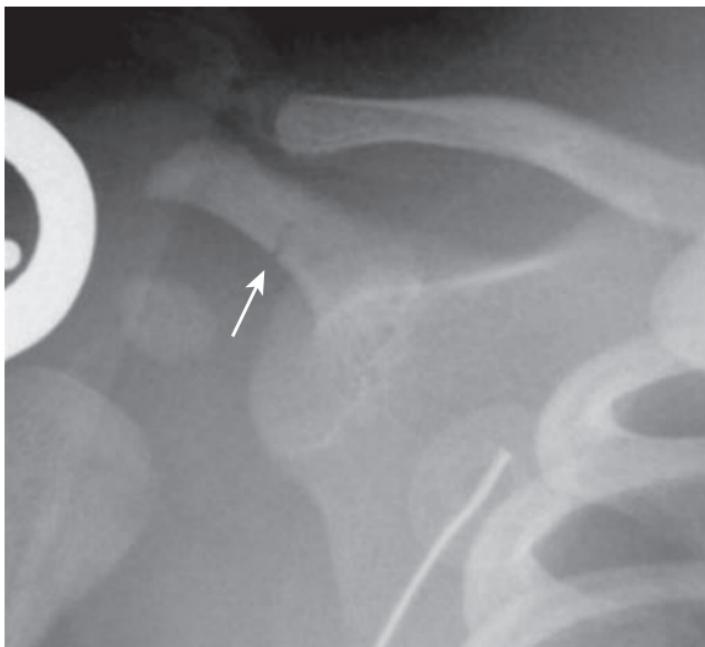
Multiple parallel lines equally distributed due to a slap from a hand. (Modified from Zitelli BJ, McIntire SC, Nowalk AJ. *Atlas of Pediatric Physical Diagnosis*. 7th ed. Philadelphia: Elsevier; 2018.)<sup>39</sup>

- (2) In children greater than 5 years of age, targeted imaging to the area(s) of suspected injury is usually appropriate. The utility of screening with skeletal survey diminishes after 5 years of age.
  - (3) In children 2 to 5 years of age, decisions about type of imaging are open to clinical judgement.
  - (4) Do not use “babygrams” (i.e., whole-body x-rays in one image) because of the high rate of false-negatives.
  - (5) Follow-up skeletal survey approximately 2 weeks after the initial examination should be performed when abnormal or equivocal findings are found on initial study and when abuse is suspected on clinical grounds to identify fractures missed on initial survey.
  - (6) Fractures with an association with child abuse include rib fractures, metaphyseal bucket and corner fractures, spine and scapula fractures, and complex skull fractures ([Fig. 2.11](#) and [Figs. EC 2.E-G](#) for examples).
- b. Head CT without contrast if:
- (1) Less than 6 months of age with suspected abuse
  - (2) Neurologic changes
  - (3) Facial injuries concerning for abuse
- c. Additional imaging/consultation
- (1) Ophthalmologic evaluation for retinal hemorrhages.
  - (2) MRI may identify lesions not detected by CT (e.g., posterior fossa injury and diffuse axonal injury).
4. **What to do if physical abuse is suspected**
- a. All healthcare providers are required by law to report suspected child maltreatment to the local police and/or child welfare agency.
  - b. In addition, consider consultation with local child injury/abuse specialist.
  - c. Medical stabilization is the primary goal; prevention of further injuries is the long-term goal.
  - d. The professional who makes such reports is immune from any civil or criminal liability.
  - e. Carefully and legibly document the following:
    - (1) Reported and suspected history and mechanisms of injury.
    - (2) Any history given by the victim in his or her own words (use quotation marks).
    - (3) Information provided by other providers or services.
    - (4) Physical examination findings, including drawings of injuries and details of dimensions, color, shape, and texture. Consider early use of police crime laboratory photography to document injuries. If taking photos, start with full patient, then part of patient, then zoomed into wound, and then take a separate photo of wrist identification band.



**Figure EC 2.E**

Healing right clavicular fracture and nine fractures of the right ribs and four fractures of the left ribs. (Modified from Coley BD. Caffey's Pediatric Diagnostic Imaging. 13th ed. Philadelphia: Elsevier; 2019.)<sup>40</sup>

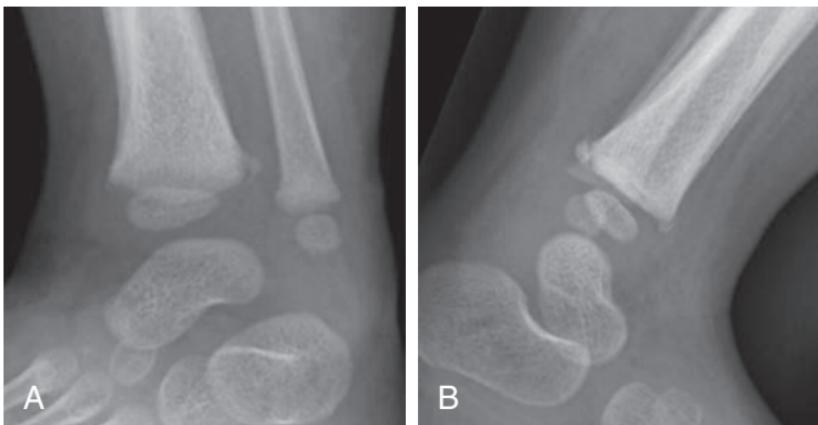


**Figure EC 2.F**

Right acromial fracture (arrow). (*Modified from Coley BD. Caffey's Pediatric Diagnostic Imaging. 13th ed. Philadelphia: Elsevier; 2019.*)<sup>40</sup>

**Figure EC 2.G**

Bilateral parietal fractures of the skull. (*Modified from Coley BD. Caffey's Pediatric Diagnostic Imaging. 13th ed. Philadelphia: Elsevier; 2019.*)<sup>40</sup>

**FIGURE 2.11**

"Bucket-Handle Fracture" (A) and a "Corner Fracture" of the distal tibial metaphysis (B). (Modified from Coley BD. Caffey's Pediatric Diagnostic Imaging. 13th ed. Philadelphia: Elsevier; 2019.)<sup>40</sup>

## B. Sexual Abuse

### 1. Physical Exam Findings

- Normal genital examination does not rule out abuse; most examinations are normal in cases of abuse.<sup>36</sup>
- Table 2.6 for physical exam findings highly suggestive of sexual abuse.<sup>37</sup>

### 2. What to do if sexual abuse is suspected<sup>38</sup>

- If suspected sexual abuse occurred within 72 hours to a child younger than 12 years or within 120 hours to a child older than 12 years, defer interview and examination and urgently involve a multidisciplinary team with a sexual assault nurse examiner with expertise in the evaluation of sexual abuse.
- Nonacute examinations falling outside of the above time windows should be deferred to a child advocacy center.
- Genital examination should be performed by a trained forensic specialist.
- Evaluate the need for sexually transmitted infection (STI) testing.

### 3. STI testing

- Tests include: Serum human immunodeficiency virus (HIV), serum syphilis, gonorrhea (culture or NAAT from pharynx and anus in boys and girls, vagina in girls and urethra in boys), chlamydia (culture or NAAT from anus in boys and girls, vagina in girls).
- In adolescents, recommended for all patients.
- In prepubertal children, consider testing if:
  - Experienced penetration of the vagina or anus
  - Abuse by a stranger

**TABLE 2.6****PHYSICAL EXAM FINDINGS SUGGESTIVE OF SEXUAL ABUSE<sup>37</sup>****ACUTE TRAUMA TO GENITAL/ANAL TISSUES**

Acute laceration(s) or bruising of labia, penis, scrotum, or perineum, posterior fourchette or vestibule not involving the hymen

Bruising, petechiae, or abrasion on the hymen

Acute laceration of the hymen of any depth, partial or complete

Vaginal laceration

Perianal laceration with exposure of tissues below the dermis

**RESIDUAL (HEALING) INJURIES TO GENITAL/ANAL TISSUES**

Perianal scar

Scar of the posterior fourchette or fossa

Healed hymenal transection/complete hymen cleft—a defect in the hymen below the 3–9 o'clock location that extends to or through the base of the hymen with no hymenal tissue discernible at that location

Signs of female genital mutilation or cutting, such as loss of part or all of the prepuce (clitoral head), labia minora or majora, or vertical linear scar adjacent to the clitoris

**FINDINGS DIAGNOSTIC OF SEXUAL ABUSE**

Pregnancy

Semen identified in forensic specimens taken directly from the child's body

- (3) Abuse by a perpetrator known to be infected with an STI or at high risk of being infected (e.g., IV drug use, men who have sex with men, people with multiple sexual encounters)
- (4) Child with sibling or other relative in the household with STI
- (5) Child living in an area with high rate of STI in the community
- (6) Signs/symptoms of an STI
- (7) Already been diagnosed with one STI

**XI. RESOURCES**

**A. Acute Concussion Evaluation Forms for Emergency Department and Physician/Clinician Office:** <https://www.cdc.gov/headsup/providers/tools.html>

**B. Acute Concussion Evaluation Care Plans for Work and School:** <https://www.cdc.gov/headsup/providers/discharge-materials.html>

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A complete list of references can be found online at [www.expertconsult.com](http://www.expertconsult.com).

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**FIGURE 2.7**

Frenulum tear due to direct blow to the face. (Modified from Zitelli BJ, McIntire SC, Nowalk, AJ. *Atlas of Pediatric Physical Diagnosis*. 7th ed. Philadelphia: Elsevier; 2018.)<sup>39</sup>



**FIGURE 2.8**

Postauricular bruising. (Modified from Zitelli BJ, McIntire SC, Nowalk, AJ. *Atlas of Pediatric Physical Diagnosis*. 7th ed. Philadelphia: Elsevier; 2018.)<sup>39</sup>



**FIGURE 2.9**

Petechial lesions due to choking. (Modified from Zitelli BJ, McIntire SC, Nowalk, AJ. *Atlas of Pediatric Physical Diagnosis*. 7th ed. Philadelphia: Elsevier; 2018.)<sup>39</sup>



**FIGURE 2.10**

Pinch marks signified by two small bruises separated by clear space. (Modified from Zitelli BJ, McIntire SC, Nowalk, AJ. *Atlas of Pediatric Physical Diagnosis*. 7th ed. Philadelphia: Elsevier; 2018.)<sup>39</sup>

# Chapter 3

## Toxicology

Maria D. Latham, MD

 See additional content on Expert Consult

**Whenever ingestion is suspected, contact local poison control at 1-800-222-1222.**

Each year the American Association of Poison Control Centers records more than 1.2 million childhood poisoning exposures. Of these exposures, 76% occur in children younger than the age of 6 years. Exposures in young children are often unintentional, whereas adolescents are more likely to have intentional ingestions.<sup>1</sup>

### I. INITIAL EVALUATION

#### A. History

##### 1. Exposure history

Obtain history from witnesses and/or close contacts. Route, timing, and number of exposures (acute, chronic, or repeated ingestion), prior treatments or decontamination efforts.<sup>2,3</sup>

##### 2. Substance identification and quantity ingested

Attempt to identify exact name of substance(s) ingested, including: product name, active ingredients, possible contaminants, expiration date, concentration, and dose. Attempt to estimate the missing volume of liquid or the number of missing pills from a container. Poison control can assist with pill identification.

##### 3. Environmental information

Accessible items in the house or garage; open containers; spilled tablets; household members taking medications, visitors to the house, herbs, or other complementary medicines.<sup>2</sup>

#### B. Workup and Laboratory Investigation

##### 1. Electrocardiogram (ECG):

Several medications will cause ECG changes, including QRS prolongation.

##### 2. Blood Tests

- Individual drug levels such as acetaminophen, aspirin, and ethanol are helpful general screenings in an acute, unknown ingestion.
- Acetaminophen levels are especially important to test in suicidal ingestions. Acetaminophen is detected in 1/500 of all suicidal ingestions even when it is not reported as an ingested agent.<sup>3</sup>
- Venous blood gas, blood glucose, and serum electrolytes.

##### 3. Urine Toxicology Screens

- Basic screens include amphetamines, cocaine, opiates, phencyclidine (PCP), and tetrahydrocannabinol (THC).

- b. Positive results are presumptive only; must be confirmed by gas chromatography/mass spectrometry.<sup>4</sup>

### C. Clinical Diagnostic Aids (Table EC 3.A)

## II. TOXIDROMES

See Table 3.1.

3

## III. INGESTIONS AND ANTIDOTES

See Table 3.2.

### A. Decontamination

#### 1. Activated charcoal<sup>5</sup>

- a. Most effective when used within first hour after ingestion but can be given after first hour, especially for sustained-release preparations. Should be given PO to an awake and alert patient. Nasogastric (NG) tube should be used only if a patient is intubated.
- b. Substances not absorbed by charcoal: Iron, alcohols, lithium
- c. Contraindications: Unprotected airway, caustic ingestion, disrupted gastrointestinal tract, concern for aspiration

#### 2. Whole bowel irrigation

- a. Indicated for evacuation of substances not bound to activated charcoal such as iron, lead-containing foreign bodies, fatal sustained release products, drug packing.
- b. Use a polyethylene glycol electrolyte solution preparation to irrigate the bowel. Recommended rates: 9 months to 6 years (500 mL/hr), 6 to 12 years (1000 mL/hr), more than 12 years (1500 to 2000 mL/hr).

### B. Enhanced Removal

- 1. Hemodialysis or exchange transfusions may be indicated to remove a drug/toxin.
- 2. Ingestions that may require enhanced removal therapies: Salicylate, lithium, methanol, ethylene glycol, metformin-associated lactic acidosis, valproate, theophylline

### C. Other Considerations

- 1. Many ingestions managed primarily with supportive care of any associated toxic effects, such as hypotension or hyperpyrexia.
- 2. Seizures: First line agents are benzodiazepines. Barbiturates or propofol should be considered as second line agents. Phenytoin has no role in the treatment of toxin-induced seizures.<sup>6</sup>
- 3. Patients with severe poisoning and refractory cardiorespiratory failure after ingestion are potential extracorporeal membrane oxygenation (ECMO) candidates because the toxic effects are transient.

**TABLE EC 3.A****CLINICAL DIAGNOSTIC AIDS**

Clinical Sign	Intoxicant
<b>VITAL SIGNS</b>	
Hypothermia	Alcohol, antidepressants, barbiturates, carbamazepine, carbon monoxide, clonidine, ethanol, hypoglycemics, opioids, phenothiazines, sedative-hypnotics
Hyperpyrexia	Amphetamines, anticholinergics, antihistamines, atropinics, $\beta$ -blockers, cocaine, iron, isoniazid, monoamine oxidase inhibitors (MAOIs), phencyclidine, phenothiazines, quinine, salicylates, sympathomimetics, selective serotonin reuptake inhibitors (SSRIs), theophylline, thyroxine, tricyclic antidepressants (TCAs)
Bradypnea	Acetone, alcohol, barbiturates, botulinum toxin, clonidine, ethanol, ibuprofen, opioids, nicotine, sedative-hypnotics
Tachypnea	Amphetamines, barbiturates, carbon monoxide, cyanide, ethylene glycol, isopropanol, methanol, salicylates <i>Direct pulmonary insult:</i> Hydrocarbons, organophosphates, salicylates
Bradycardia	$\alpha$ -Agonists, alcohols, $\beta$ -blockers, calcium channel blockers, central $\alpha_2$ -agonist, clonidine, cyanide, digoxin, opioids, organophosphates, plants (lily of the valley, foxglove, oleander), sedative-hypnotics
Tachycardia	Alcohol, amphetamines, anticholinergics, antihistamines, atropine, cocaine, cyclic antidepressants, cyanide, iron, phencyclidine, salicylates, sympathomimetics, theophylline, TCAs, thyroxine
Hypotension	$\alpha$ -Agonists, angiotensin-converting enzyme (ACE) inhibitors, barbiturates, carbon monoxide, cyanide, iron, methemoglobinemia, opioids, phenothiazine, sedative-hypnotics, TCAs <i>Profound hypotension:</i> $\beta$ -blockers, calcium channel blockers, clonidine, cyclic antidepressants, digoxin, imidazolines, nitrates, quinidine, propoxyphene, theophylline
Hypertension	Amphetamines, anticholinergics, antihistamines, atropinics, clonidine, cocaine, cyclic antidepressants (early after ingestion), diet pills, ephedrine, MAOIs, nicotine, over-the-counter cold remedies, phencyclidine, phenylpropanolamine, pressors, sympathomimetics, TCAs <i>Delayed hypertension:</i> Thyroxine
Hypoxia	Oxidizing agents
<b>NEUROMUSCULAR</b>	
Nervous system instability	<i>Insidious onset:</i> Acetaminophen, benzocaine, opioids <i>Abrupt onset:</i> Lidocaine, monocyclic or tricyclic antidepressants, phenothiazines, theophylline <i>Delayed onset:</i> Atropine, diphenoxylate <i>Transient instability:</i> Hydrocarbons
Depression and excitation	Clonidine, imidazolines, phencyclidine
Ataxia	Alcohol, anticonvulsants, barbiturates, carbon monoxide, heavy metals, hydrocarbons, solvents, sedative-hypnotics

*Continued*

**TABLE EC 3.A—CONT'D****CLINICAL DIAGNOSTIC AIDS**

Clinical Sign	Intoxicant
Chvostek/Trousseau signs	Ethylene glycol, hydrofluoric acid–induced hypocalcemia, phosphate-induced hypocalcemia from Fleet enema
Coma	Alcohol, anesthetics, anticholinergics (antihistamines, antidepressants, phenothiazines, atropinics, over-the-counter sleep preparations), anticonvulsants, baclofen, barbiturates, benzodiazepines, bromide, carbon monoxide, chloral hydrate, clonidine, cyanide, cyclic antidepressants, γ-hydroxybutyrate (GHB), hydrocarbons, hypoglycemics, inhalants, insulin, lithium, opioids, organophosphate insecticides, phenothiazines, salicylates, sedative-hypnotics, tetrahydrozoline, theophylline
Delirium, psychosis	Alcohol, anticholinergics (including cold remedies), cocaine, heavy metals, heroin, lysergic acid diethylamide (LSD), marijuana, mescaline, methaqualone, peyote, phencyclidine, phenothiazines, steroids, sympathomimetics
Miosis	Barbiturates, clonidine, ethanol, opioids, organophosphates, phencyclidine, phenothiazines, muscarinic mushrooms
Mydriasis	Amphetamines, antidepressants, antihistamines, atropinics, barbiturates (if comatose), botulism, cocaine, glutethimide, LSD, marijuana, methanol, phencyclidine
Nystagmus	Barbiturates, carbamazepine, diphenylhydantoin, ethanol, glutethimide, MAOIs, phencyclidine (both vertical and horizontal), sedative-hypnotics
Paralysis	Botulism, heavy metals, paralytic shellfish poisoning, plants (poison hemlock)
Seizures	Ammonium fluoride, amphetamines, anticholinergics, antidepressants, antihistamines, atropine, β-blockers, boric acid, bupropion, caffeine, camphor, carbamates, carbamazepine, carbon monoxide, chlorinated insecticides, cocaine, cyclic antidepressants, diethyltoluamide, ergotamine, ethanol, GHB, <i>Gyromitra</i> mushrooms, hydrocarbons, hypoglycemics, ibuprofen, imidazolines, isoniazid, lead, lidocaine, lindane, lithium, LSD, meperidine, nicotine, opioids, organophosphate insecticides, phencyclidine, phenothiazines, phenylpropanolamine, phenytoin physostigmine, plants (water hemlock), propoxyphene, salicylates, strychnine, theophylline

**CARDIOVASCULAR**

Hypoperfusion      Calcium channel blockers, iron

Wide QRS complex      TCAs

**ELECTROLYTES**

Anion gap metabolic acidosis      Acetaminophen, carbon monoxide, chronic toluene, cyanide, ethylene glycol, ibuprofen, iron, isoniazid, lactate, methanol, metformin, paraldehyde, phenformin, salicylates

Electrolyte disturbances      Diuretics, salicylates, theophylline

Hypoglycemia      Alcohol, β-blockers, hypoglycemics, insulin, salicylates

**TABLE EC 3.A—CONT'D****CLINICAL DIAGNOSTIC AIDS**

Clinical Sign	Intoxicant
Serum osmol gap	Acetone, ethanol, ethylene glycol, isopropyl alcohol, methanol, propylene glycol
<b>SKIN</b>	
Cyanosis unresponsive to oxygen	Aniline dyes, benzocaine, nitrites, nitrobenzene, phenazopyridine, phenacetin
Flushing	Alcohol, antihistamines, atropinics, boric acid, carbon monoxide, cyanide, disulfiram
Jaundice	Acetaminophen, carbon tetrachloride, heavy metals (iron, phosphorus, arsenic), naphthalene, phenothiazines, plants (mushrooms, fava beans)
<b>ODORS</b>	
Acetone	Acetone, isopropyl alcohol, phenol, salicylates
Alcohol	Ethanol
Bitter almond	Cyanide
Garlic	Heavy metal (arsenic, phosphorus, thallium), organophosphates
Hydrocarbons	Hydrocarbons (gasoline, turpentine, etc.)
Oil of wintergreen	Salicylates
Pear	Chloral hydrate
Violets	Turpentine
<b>RADIOLOGY</b>	
Small opacities on radiograph	Halogenated toxins, heavy metals, iron, lithium, densely packaged products

**TABLE 3.1****TOXIDROMES**

Drug Class	Temp	HR	RR	BP	Pupils	Skin	Mental Status	Other Signs	Causative Agents
Anticholinergic “Mad as a hatter, red as a beet, blind as a bat, hot as a hare, dry as a bone” <sup>a</sup>	↑	↑	↑/nl	↑/nl	Dilated	Dry, flushed	Delirium, psychosis, paranoia	Urinary retention, decreased bowel sounds, thirst, garbled speech	Antihistamines, atropine, antipsychotics, phenothiazines, scopolamine, TCAs
Cholinergic “SLUDGE, Killer B’s” <sup>a</sup>	nl	↓	↑ (bronchospasm)	↓/nl	Constricted	Sweaty	Depressed, confused	Salivation, lacrimation, urination, defecation, emesis. Liquid nicotine can cause fasciculations and paralysis.	Organophosphates, pesticides, nerve agents, tobacco, liquid nicotine
Opioids	↓/nl	↓/nl	↓ (hypoventilation)	↓/nl	Constricted	No change	Sedated		Morphine, fentanyl, oxycodone, methadone
Sympathomimetics	↑/nl	↑	↑/nl	↑	Dilated	Sweaty	Agitated	At risk for seizures, coronary vasospasm	Amphetamines, cocaine
Sedative/Hypnotics “Coma with normal vitals”	nl	nl	↓/nl	↓/nl	Normal	No change	Depressed		Benzodiazepines, barbiturates, ethanol
Serotonergic	↑	↑	↑	↑/nl	Dilated	Flushed	Confusion	Shivering, muscle rigidity, at risk for seizures, hyperreflexia and clonus of lower extremities	SSRIs, SNRIs, MAOIs, trazadone, dextromethorphan, LSD, TCAs, MDMA (ecstasy)

<sup>a</sup>The “mad as a hatter” mnemonic references delirium, flushed skin, mydriasis, hyperpyrexia, and dry skin/urinary retention seen in the anticholinergic toxidrome. The “SLUDGE” mnemonic references salivation, lacrimation, urination, diaphoresis, gastrointestinal distress (including diarrhea), and emesis seen in the cholinergic toxidrome. The “Killer B’s” mnemonic references bronchospasm, bronchorrhea, and bradycardia seen in the cholinergic toxidrome.

↑ refers to increased or elevated vital sign, ↓ refers to decreased or depressed vital sign, nl refers to vital sign within normal limits.

BP, Blood pressure; HR, heart rate; LSD, lysergic acid diethylamide; MAOIs, monoamine oxidase inhibitors; RR, respiratory rate; SNRIs, serotonin and norepinephrine reuptake inhibitors; SSRIs, selective serotonin reuptake inhibitors; TCAs, tricyclic antidepressants; Temp, temperature.

**TABLE 3.2****COMMONLY INGESTED AGENTS**

Ingested Agent	Signs and Symptoms	Antidote <sup>a</sup>
Acetaminophen	See Section IV	
Amphetamines	See sympathomimetics toxidrome in Table 3.1	Supportive care <b>Benzodiazepines</b> for agitation
Anticholinergics	See anticholinergic toxidrome in Table 3.1	<b>Physostigmine</b>
Anticholinesterase (insecticides, donepezil, mushrooms)	See cholinergic toxidrome in Table 3.1	<b>Atropine</b>
Antihistamines	See anticholinergic toxidrome in Table 3.1; paradoxical CNS stimulation, dizziness, seizures, prolonged QT	Supportive care
Benzodiazepines	See sedative/hypnotic toxidrome in Table 3.1	<b>Flumazenil</b>
β-Blockers	Bradycardia, hypotension, AV conduction block, bronchospasm, hypoglycemia	<b>Glucagon</b> See <b>insulin/dextrose</b> treatment in calcium channel blockers
Calcium channel blockers	Bradycardia, hypotension, AV conduction block, pulmonary edema, hyperglycemia	<b>Calcium chloride (10%)</b> <b>Calcium gluconate (10%)</b> <b>Glucagon</b> <b>High-Dose Insulin/Dextrose<sup>b</sup>:</b> 1 unit/kg bolus → infuse at 1–10 unit/kg/hr; give with D25W at 0.5 g/kg/hr. Monitor BG frequently.
Clonidine	Symptoms resemble an opioid toxidrome. CNS depression, coma, lethargy, hypothermia, miosis, bradycardia, profound hypotension, respiratory depression	<b>Naloxone</b>
Cocaine	See sympathomimetics toxidrome in Table 3.1	Supportive care
Detergent pods	Vomiting, sedation, aspiration, respiratory distress	Supportive care
Ecstasy	Hallucinations, teeth grinding, hyperthermia, hyponatremia, seizures	Supportive care
Ethanol	See sedative/hypnotic toxidrome in Table 3.1	Supportive care
Ethylene glycol/ methanol	Hypoglycemia in young children Similar to ethanol; additionally, blurry or double vision (methanol), renal failure/hypocalcemia (ethylene glycol), osmol gap with severe anion gap metabolic acidosis	<b>Fomepizole</b> Ethanol (only to be used as second line agent when fomepizole unavailable; risk of inappropriate dosing, CNS depression, aspiration, and hypoglycemia). Consider dialysis.

**TABLE 3.2—CONT'D****COMMONLY INGESTED AGENTS**

Ingested Agent	Signs and Symptoms	Antidote <sup>a</sup>
Iron	Vomiting, diarrhea, hypotension, lethargy, anion gap metabolic acidosis, cardiogenic shock, renal failure	Deferoxamine
Lead	See Section V	
Nicotine	Vomiting and cholinergic toxidrome in <b>Table 3.1</b>	Supportive care
NSAIDs	Nausea, vomiting, epigastric pain, headache, gastrointestinal hemorrhage, renal failure	Supportive care
Opioids	See opioid toxidrome in <b>Table 3.1</b>	<b>Naloxone</b>
Organophosphates	See cholinergic toxidrome in <b>Table 3.1</b>	<b>Atropine</b> <b>Pralidoxime</b>
Salicylates	Gastrointestinal upset, tinnitus, tachypnea, hyperpyrexia, dizziness, lethargy, dysarthria, seizure, coma, cerebral edema	<b>Sodium bicarbonate</b> Consider dialysis
Serotonergic Agents	See serotonergic toxidrome in <b>Table 3.1</b>	<b>Benzodiazepines (first line)</b> Cyproheptadine
Sulfonylureas	Hypoglycemia, dizziness, agitation, confusion, tachycardia, diaphoresis	<b>Food (if able)</b> <b>Dextrose:</b> 0.5–1 g/kg (2–4 mL/kg of D25W) <i>After euglycemia achieved:</i> <b>Octreotide:</b> 1–1.25 mCg/kg SQ Q6–12 hr (max dose 50 mCg) if rebound hypoglycemia
Synthetic cannabinoids	Agitation, altered sensorium, tachycardia, hypertension, vomiting, mydriasis, hypokalemia	Supportive care
TCAs	Tachycardia, seizures, delirium, widened QRS possibly leading to ventricular arrhythmias, hypotension	<i>For wide QRS complex:</i> <b>Sodium bicarbonate:</b> 1–2 mEq/kg IV push, followed by D5W + 140 mEq/L NaHCO <sub>3</sub> and 20 meq/L KCl at 1.5× maintenance fluid rate with goal serum pH 7.45–7.55
Warfarin	Bleeding	<b>Phytonadione/Vitamin K<sub>1</sub></b>

<sup>a</sup>See Formulary for dosing recommendations.BG, Blood glucose; CNS, central nervous system; KCl, potassium chloride; NaHCO<sub>3</sub>, sodium bicarbonate; NSAIDs, nonsteroidal antiinflammatory drugs; TCA, tricyclic antidepressant.Data from Gummie DD, Mowry JB, Spyker DA, et al. 2017 Annual Report of the American Association of Poison Control Centers' National Poison Data System (NPDS): 35th Annual Report. *Clin Toxicol.* 2017;56(12):1213–1415.

## IV. ACETAMINOPHEN OVERDOSE<sup>7</sup>

NAPQI metabolite is hepatotoxic.

### A. Four Phases of Intoxication

- Phase 1 (first 24 hours):** Nonspecific symptoms such as nausea, malaise, vomiting.
- Phase 2 (24 to 72 hours):** Above symptoms resolve; right upper quadrant pain, hepatomegaly, and increasing transaminases develop.
- Phase 3 (72 to 96 hours):** Return of nonspecific symptoms as well as evidence of liver failure (increased prothrombin time, lactate, phosphate), renal failure, and encephalopathy.
- Phase 4 (4 days to 2 weeks):** Recovery or death.

### B. Treatment Criteria

- Serum acetaminophen** concentration above the possible toxicity line on the Rumack-Matthew nomogram after single acute ingestion (Fig. 3.1).
- History of ingesting more than 200 mg/kg or 10 g** (whichever is less) and serum concentration not available or time of ingestion not known.
- If time of ingestion is unknown or multiple/chronic ingestion,** check acetaminophen level and AST. Treat if either is elevated.

### C. Antidote: *N*-Acetylcysteine (See Formulary)

- PO:** 140 mg/kg loading dose followed by 70 mg/kg Q4 hours for 17 doses (18 total doses including loading dose).
- Intravenous (IV):** 150 mg/kg *N*-acetylcysteine IV loading dose over 60 minutes, followed by 50 mg/kg over 4 hours, followed by 100 mg/kg over 16 hours for a total infusion time of 21 hours. Some patients may require more than 21 hours of *N*-acetylcysteine administration.
- Liver failure:** Continue the 100 mg/kg over 16 hours infusion until resolution of encephalopathy, AST less than 1000 units/L, and INR less than 2.

## V. LEAD POISONING<sup>8</sup>

### A. Definition:

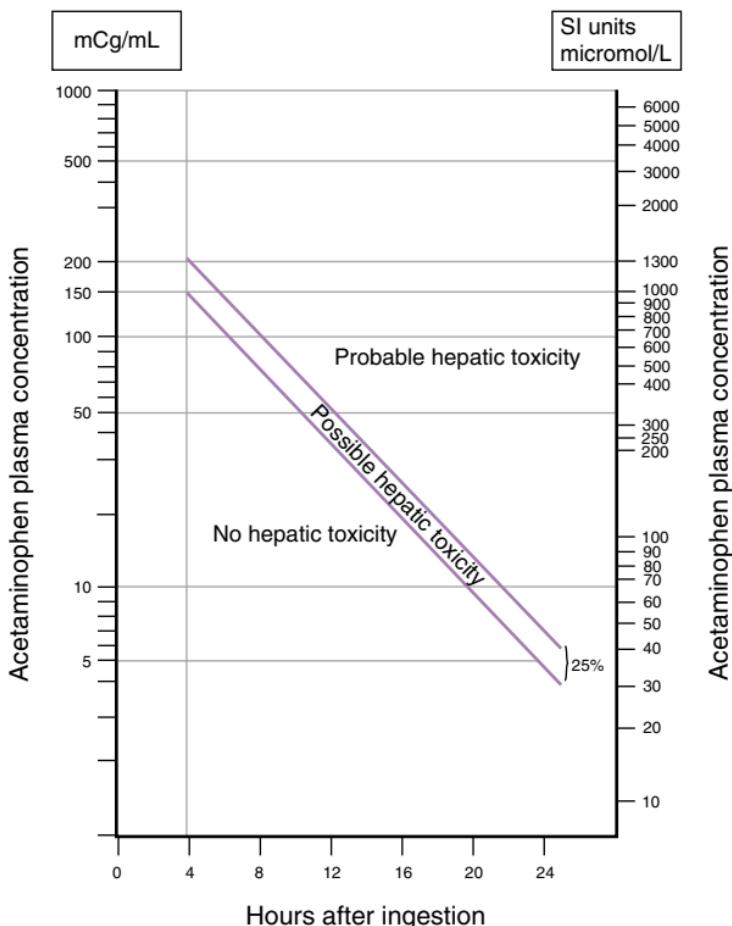
Centers for Disease Control and Prevention (CDC) defines a reference level of 5 mCg/dL to identify children with elevated blood lead levels (BLLs).

### B. Sources of Exposure:

Paint, dust, soil, drinking water, cosmetics, cookware, toys, and caregivers with occupations and/or hobbies using lead-containing materials or substances.

### C. Overview of Symptoms by Blood Lead Level:

- BLL ≥40 mCg/dL:** Irritability, vomiting, abdominal pain, constipation, anorexia
- BLL ≥70 mCg/dL:** Lethargy, seizure, and coma. **Note:** Children may be asymptomatic with lead levels greater than 100 mCg/dL.

**FIGURE 3.1**

Rumack-Matthew nomogram. Semilogarithmic plot of plasma acetaminophen levels versus time. This nomogram is valid for use after single acute ingestions of acetaminophen. The need for treatment cannot be extrapolated based on a level before 4 hr. (Data from Pediatrics 55:871, 1975 and Micromedex.)

#### D. Management

1. See Tables 3.3 and 3.4 for general management and repeat testing guidelines.

2. Chelation therapy<sup>2</sup>

- a. Asymptomatic children with BLL 45 to 69 mCg/dL

**Succimer:** 1050 mg/m<sup>2</sup>/day PO divided Q8 hours × 5 days, then 700 mg/m<sup>2</sup>/day divided Q12 hours × 14 days. See Formulary for more details.

**TABLE 3.3****MANAGEMENT OF LEAD POISONING<sup>8</sup>**

Blood Lead Levels (BLL)	Recommended Guidelines See <i>Table 3.4</i> for Repeat Testing Guidelines.
5–9 mCg/dL	<ol style="list-style-type: none"> <li>Obtain detailed environmental exposure history to assess for possible sources.</li> <li>Provide education about reducing environmental lead exposure and reducing dietary lead absorption<sup>a</sup></li> </ol>
10–19 mCg/dL	<ol style="list-style-type: none"> <li>As above for BLL 5–9 mCg/dL</li> <li>Consider iron studies.</li> <li>Environmental investigation may be available based on local resources.</li> </ol>
20–44 mCg/dL	<ol style="list-style-type: none"> <li>As above for BLL 5–9 mCg/dL</li> <li>Environmental investigation</li> <li>Iron level, complete blood cell count (CBC), abdominal radiography with bowel decontamination if indicated</li> <li>Complete exam including neurodevelopmental assessment</li> </ol>
45–69 mCg/dL	<ol style="list-style-type: none"> <li>As above for BLL 20–44 mCg/dL</li> <li>Administer oral chelation therapy, consider hospitalization</li> </ol>
≥70 mCg/dL	<ol style="list-style-type: none"> <li>Hospitalize and commence chelation therapy</li> <li>Contact local poison control</li> </ol>

<sup>a</sup>Iron, calcium, and vitamin C help to minimize absorption of lead.

**TABLE 3.4****REPEAT BLOOD LEAD TESTING GUIDELINES<sup>8</sup>**

If Screening BLL is: (mCg/dL)	Time Frame of Confirmation of Screening BLL	Follow-Up Testing (After Confirmatory Testing)	Later Follow-Up Testing After BLL Declining
≥5–9	1–3 months	3 months	6–9 months
10–19	1 week–1 month <sup>a</sup>	1–3 months	3–6 months
20–24	1 week–1 month <sup>a</sup>	1–3 months	1–3 months
25–44	1 week–1 month <sup>a</sup>	2 weeks–1 month	1 month
45–59	48 hr	<i>Repeat testing as soon as possible after chelation therapy</i>	
60–69	24 hr		
≥70	Urgently		

<sup>a</sup>Per provider discretion.

BLL, Blood lead level.

b. Asymptomatic children with BLL ≥70 mCg/dL

(1) **Succimer:** Per above dosing.

(2) **Edetate (EDTA) calcium disodium:** 1000 mg/m<sup>2</sup> (max dose 2 to 3 g) as 24-hour IV infusion × 5 days. Begin two hours after first dose of succimer. Monitor renal function closely.

**Warning:** Do not mistake edetate disodium for edetate calcium disodium. Edetate calcium disodium is the correct medicine used for the treatment of lead poisoning.

- c. Symptomatic children (e.g., lead encephalopathy, seizure)
  - (1) **Dimercaprol (BAL):** 450 mg/m<sup>2</sup>/day IM divided Q4 hours × 3 to 5 days (number of days based on clinical course). Do not give to patients with peanut allergy. Do not use concomitantly with iron, as BAL-iron complex is a potent emetic. Use with caution in patients with G6PD deficiency, as it may cause hemolysis.
  - (2) **Edetate (EDTA) calcium disodium:** 1500 mg/m<sup>2</sup> (maximum dose 2 to 3 g) as 24-hour IV continuous infusion × 5 days. Begin four hours after first dose of BAL.

## VI. WEB RESOURCES

- American Association of Poison Control Centers: <http://www.aapcc.org/>
- American Academy of Clinical Toxicology: <http://www.clintox.org/index.cfm>
- Centers for Disease Control and Prevention, Section on Environmental Health: <http://www.cdc.gov/nceh>

## REFERENCES

A complete list of references can be found online at [www.expertconsult.com](http://www.expertconsult.com).

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# Chapter 4

## Procedures

Andrew Percy, MD

 See additional content on Expert Consult

### I. GENERAL GUIDELINES

#### A. Consent

Before performing any procedure, it is crucial to obtain informed consent from the parent or guardian by explaining the procedure, the indications, any risks involved, and any alternatives. Obtaining consent should not impede life-saving, emergency procedures.

#### B. Risks

1. All invasive procedures involve pain, risk for infection and bleeding, and potential injury to neighboring structures.
2. Sedation and analgesia should be planned in advance, and the risks of such explained to the parent and/or patient as appropriate. (See Chapter 6 and the AAP Guidelines for Monitoring and Management of Pediatric Patients Before, During, and After Sedation for Diagnostic and Therapeutic Procedures.<sup>1</sup>)
3. Universal precautions and proper sterile technique should be followed for all patient contact that exposes the healthcare provider to bodily fluids.

#### C. Documentation

**It is important that the physician performing the procedure document the informed consent process.** Include the date, time, additional personnel present (if applicable), brief summary of the consent conversation, the diagnosis, recommended procedure, specific risks and benefits, and alternative treatments. It is equally important to document if a patient refuses a procedure and that the risks associated with refusal were discussed.

#### D. Attending to the Needs of a Fearful Child

Children represent a vulnerable population in that they often lack the capacity to understand why a potentially uncomfortable procedure is being performed. All efforts should be made to provide information about the procedure to the child at an age-appropriate level. Utilize Child Life Specialists as able. When possible, allow the child to touch unfamiliar objects in the examination room to desensitize them and enhance trust. Address the child's fears. Toddlers often fear separation from the parent. Older children often fear pain. Adolescents often worry about embarrassment sustained by expressing anxiety or fear. Encourage active parent participation and presence. Allow all children a degree of basic autonomy such as selecting the postprocedure bandage color.

## II. ULTRASOUND FOR PROCEDURES

### A. Introduction to Ultrasound

Ultrasound has become an increasingly important bedside diagnostic and procedural aid, and it can improve visualization of subcutaneous structures during procedures.

### B. Ultrasound Basics

#### 1. Probe Selection

- Linear transducers use higher frequencies to produce higher resolution images and are primarily used for procedures in pediatrics. A wide area of contact at the skin surface facilitates needle placement in procedures.
- Curvilinear transducers use low to midrange frequencies and permit deep structure visualization. Though they provide a wide area of skin contact to facilitate procedures near concave and convex surfaces, larger curvilinear probes are difficult to use in small children.
- There are a variety of other probes (phased-array, microconvex) that generate sector shaped images but are predominantly used for diagnostic purposes.

#### 2. Image Optimization

- Ensure adequate contact by using enough ultrasound gel and applying comfortable pressure on the skin.
- Gain: Measure of image brightness which is used for optimizing images and reducing artifact.
- Frequency: Increase to improve image resolution of shallow structures. Decrease to improve imaging of deep structures.
- Depth: Adjust to visualize structure of interest and at least a centimeter of tissue below that structure.

## III. NEUROLOGIC PROCEDURES: LUMBAR PUNCTURE<sup>2,3</sup>

### A. Indications:

Examination of spinal fluid for suspected infection, inflammatory disorder, or malignancy, instillation of intrathecal chemotherapy, or measurement of opening pressure.

### B. Complications:

Local pain, infection, bleeding, spinal fluid leak, hematoma, spinal headache, and acquired epidermal spinal cord tumor (caused by implantation of epidermal material into the spinal canal if no stylet is used on skin entry).

### C. Cautions and Contraindications:

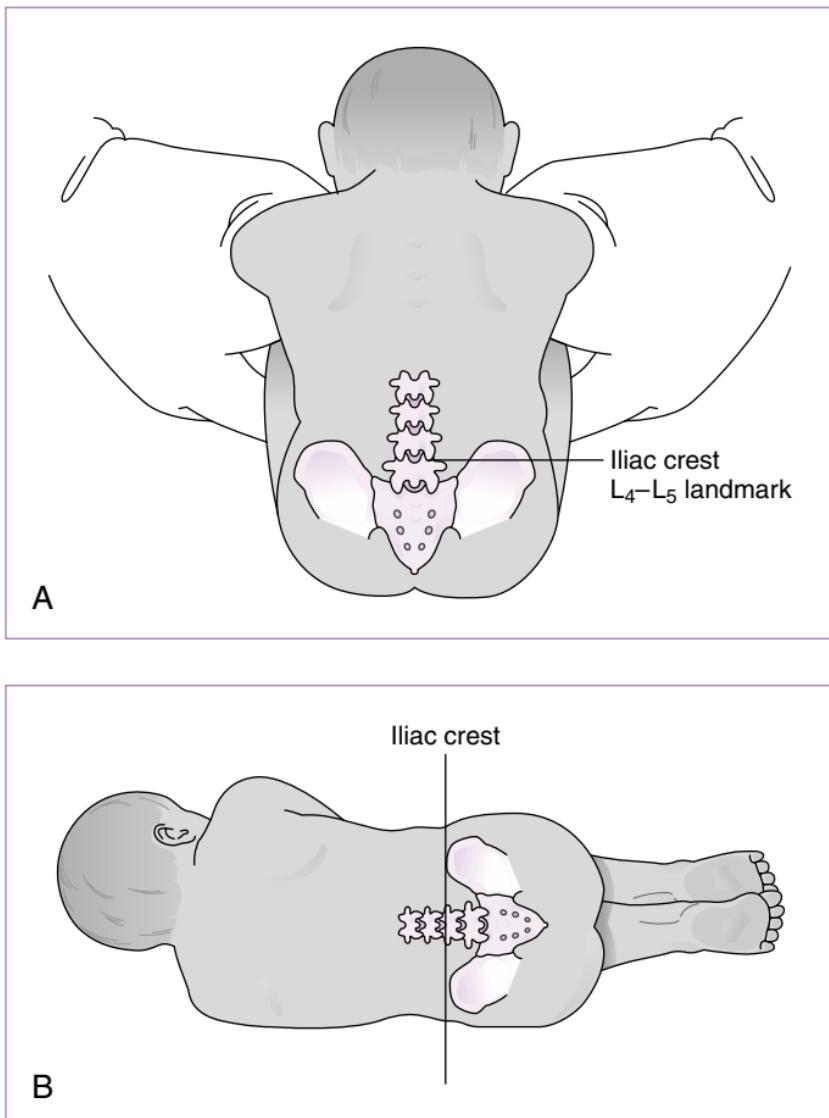
- Increased intracranial pressure (ICP): Before lumbar puncture (LP), perform a funduscopic examination. Presence of papilledema, retinal hemorrhage, or clinical suspicion of increased ICP should prompt further evaluation and may be a contraindication to the procedure. A sudden drop in spinal canal fluid pressure by rapid release of cerebrospinal fluid (CSF) may cause fatal herniation. Computed tomography (CT)

may be indicated before LP if there is suspected intracranial bleeding, focal mass lesion, or increased ICP. A normal CT scan does not rule out increased ICP but usually excludes conditions that may put the patient at risk for herniation. Decision to obtain CT should not delay appropriate antibiotic therapy, if indicated.

2. Bleeding diathesis: Platelet count greater than 50,000/mm<sup>3</sup> is desirable before LP, and correction of any clotting factor deficiencies can minimize the risk for bleeding and subsequent cord or nerve root compression.
3. Overlying skin infection may result in inoculation of CSF with organisms.
4. LP should be deferred in unstable patients, and appropriate therapy should be initiated, including antibiotics, if indicated.

#### D. Procedure:

1. Apply local anesthetic cream if sufficient time is available.
2. Position child (Fig. 4.1) in either the sitting position or lateral recumbent position, with hips, knees, and neck flexed. Keep shoulders and hips aligned to avoid rotating the spine. *Do not* compromise a small infant's cardiorespiratory status with positioning.
3. Locate the desired intervertebral space (either L3 to L4 or L4 to L5) by drawing an imaginary line between the top of the iliac crests. Alternatively, ultrasound can be used to mark the intervertebral space (see [Section XI](#), Online Content).
4. Prepare the skin in a sterile fashion. Drape conservatively to make monitoring the infant possible. Use a 20- to 22-G spinal needle with stylet (1.5, 2.5, or 3.5 inch depending on the size of the child). A smaller-gauge needle will decrease the incidence of spinal headache and CSF leak.
5. Overlying skin and interspinous tissue can be anesthetized with 1% lidocaine using a 25G needle.
6. Puncture the skin in the midline just caudad to the palpated spinous process, angling slightly cephalad toward the umbilicus. Advance several millimeters at a time, and withdraw stylet frequently to check for CSF flow. **Needle may be advanced without the stylet once it is completely through the skin.** In small infants, one may *not* feel a change in resistance or "pop" as the dura is penetrated.
7. If resistance is met initially and the needle cannot be advanced, withdraw needle to just under the skin surface and redirect the angle of the needle slightly.
8. Send CSF for appropriate studies. In general, send the first tube for culture and Gram stain, the second tube for measurement of glucose and protein levels, and the last tube for cell count and differential. Additional tubes can be collected for viral cultures, polymerase chain reaction (PCR), or CSF metabolic studies, if indicated. If subarachnoid hemorrhage or traumatic tap is suspected, send the first and fourth tubes for cell count, and ask the laboratory to examine the CSF for xanthochromia.



**FIGURE 4.1**

Lumbar puncture site. (A) Infant placed in sitting position. (B) Infant placed in lateral (recumbent) position. (From Dieckmann R, Fiser D, Selbst S. Pediatric Emergency and Critical Care Procedures. St. Louis: Mosby; 1997.)

9. Accurate measurement of CSF pressure can be made only with the patient lying quietly on his or her side in an unflexed position. It is not a reliable measurement in the sitting position. Once the free flow of spinal fluid is obtained, attach the manometer and measure CSF pressure. Opening pressure is recorded when the CSF level is steady.

E. A video on **lumbar punctures** is available on the *New England Journal of Medicine's* website.

#### IV. OTOLARYNGOLOGIC PROCEDURES

##### A. Cerumen Impaction Removal<sup>4,5</sup>

1. **Indications:** Symptomatic (decreased hearing, pain) and/or assessment of the ear. Clinicians should *not* routinely disimpact asymptomatic patients whose ears can be adequately assessed.
2. **Complications:** Allergic reaction to cerumenolytics, trauma, earache, dizziness, nystagmus, retention of water, tympanic membrane perforation.
3. **Procedures:**

###### a. Cerumenolytics

- (1) There is no high-quality evidence suggesting that one cerumenolytic is more effective than another. Water and saline are equally as effective as cerumenolytics. There is no difference in efficacy between oil-based and water-based treatments.
- (2) Avoid hydrogen peroxide; may exacerbate cerumen impaction.
- (3) Apply 5 to 10 eardrops twice daily for no longer than 4 days. Keep the head tilted for several minutes for cerumenolytic retention.

###### b. Irrigation

- (1) Direct visualization is not necessary.
- (2) Irrigation of the ear canal with a large syringe containing luke-warm water is equally effective as a commercial mechanical jet irrigator.<sup>5</sup>
- (3) Place a small bucket (e.g., emesis bin) under the patient's ear to collect water.
- (4) Straighten the ear as much as possible by lifting the auricle up and posteriorly. Gently apply a continuous stream upwards in the canal.
- (5) **Note** that irrigation is contraindicated in patients with tympanostomy tubes or perforated tympanic membranes, and for removing vegetables/legumes (increases swelling) and button batteries (enhances current flow).

###### c. Manual Removal/Instrumentation

- (1) Most useful for cerumen removal in the outer one-third of the ear.
- (2) Direct visualization is essential, and may render manual removal impossible in an uncooperative patient.
- (3) Tools include curettes (plastic or metal), spoons, alligator forceps. Do not attempt to break through the cerumen. Advance the loop of the curette behind the cerumen and retrieve.

###### d. A video on **cerumen removal** is available on the *New England Journal of Medicine's* website.

##### B. Foreign Body Removal from Ear<sup>6</sup>

1. **Indications:** Retained foreign body in the external auditory canal.

2. **Contraindications:** Urgent referral to an otolaryngologist **prior** to attempted removal is indicated if object is a button battery or penetrating the ear canal (e.g., pencil, cotton-tipped swab).
3. **Complications:** External auditory canal trauma (most common), perforation of the tympanic membrane, retained foreign object.
4. **Procedure:**
  - a. Insects should be killed with mineral oil, ethanol, or lidocaine prior to attempted removal.
  - b. Irrigation is useful for hard objects resistant to grasping that are nonocclusive.
  - c. Instrumentation is most successful for irregularly shaped objects that are graspable.
  - d. Refer to otolaryngology if removal is unsuccessful.
5. A video on removal of foreign bodies from the ear and nose is available on the *New England Journal of Medicine's* website.

### C. Foreign Body Removal from Nose<sup>6,7</sup>

1. **Indications:** Retained foreign body in the nasal cavity. Button batteries and magnets attached to the nasal septum require urgent removal.
2. **Contraindications:** Most nasal foreign bodies do not require subspecialty referral. Consider otolaryngology referral for posterior objects, button batteries, and unsuccessful initial attempts.
3. **Complications:** Epistaxis, perforation of cribriform plate, retained foreign object.
4. **Procedure:** Lidocaine or any vasoconstrictor (e.g., crushed ice) may be used to minimize bleeding and edema.
  - a. **Self-Removal:** The easiest and least invasive method. Typically, only effective for patients older than 3 years. Instruct the patient to occlude the unobstructed nostril and blow his/her nose.
  - b. **Parent Kiss:** Provides up to a 60% successful removal rate.
    - (1) Instruct the caregiver to place his/her lips around the patient's lips (similar to a "mouth-to-mouth" resuscitation breath) and occlude the uninvolved nostril with one finger.
    - (2) Quickly and forcefully exhale one puff into the child's mouth. This maneuver often expels the foreign body.
  - c. **High-Flow Oxygen:** Best for foreign bodies that *completely* occlude the anterior nasal cavity. Place suction tubing into the unobstructed nostril while the child's mouth is closed. Deliver 10 to 15 L/min of oxygen flow through the tubing.
  - d. **Instrumentation:** Best for foreign bodies that are *nonocclusive*.
    - (1) Equipment: alligator forceps, right-angle hook, Foley catheter (5 to 8 Fr), irrigating devices
    - (2) Use alligator forceps to extract compressible objects that have rough surfaces.
    - (3) Use a right-angle hook for smooth objects that cannot be easily grasped.

- (4) Use a Foley catheter for small round objects (e.g., marble). Lubricate the catheter, advance the uninflated catheter past the object, inflate the catheter balloon, and withdraw the catheter and the object.

#### D. Management of Epistaxis<sup>6,8</sup>

1. **Indications:** Simple nosebleed. Most cases of epistaxis in children have a benign etiology. Referral to an otolaryngologist is only indicated for uncontrollable bleeding, posterior epistaxis, hemodynamic instability, or anatomic abnormalities (e.g., tumors, polyps). See [Chapter 14](#) for management of epistaxis in patients with hemophilia, von Willebrand disease, immune thrombocytopenia, or other bleeding disorders.
2. **Complications:** Persistent bleeding, swallowing blood, toxic shock syndrome (from packing material), septal hematomas/abscesses from traumatic packing.
3. **Procedure:** The child should sit upright and bent forward at the waist to minimize swallowing blood.
  - a. **Direct compression:** Instruct the child or parent to compress the nasal alae together for a minimum of 5 to 10 minutes. Most simple bleeds will clot after 5 to 10 minutes.
  - b. **Topical vasoconstriction:** Use oxymetazoline-soaked cotton pledgets or gauze. Phenylephrine is associated with morbidity when used topically and should be avoided in patients younger than 6 years of age. However, if bleeding is refractory to other interventions, the minimum dose of phenylephrine needed to cease bleeding should be used. Use a squirt bottle or apply the vasoconstrictor on a piece of cotton, applying direct pressure on the nose for 5 to 10 minutes.
  - c. **Nasal packing**
    - (1) Apply topical anesthetic (4% lidocaine or tetracaine) on a cotton pledge and insert into the nasal cavity.
    - (2) Rub antibiotic ointment into a quarter-inch × 72-inch gauze ribbon. Using a nasal speculum or forceps, pack the nasal cavity by grasping the gauze ribbon approximately 6 inches from its end and placing the packing as far back as possible. Ensure that the free end protrudes from the nose and secure with tape.
    - (3) Maintain packing for 72 hours and prescribe antistaphylococcal antibiotic for 7 to 10 days to minimize risk of toxic shock syndrome. If bleeding persists after 72 hours, packing should be replaced and the child referred to an otolaryngologist.

### V. CARDIOVASCULAR PROCEDURES

#### A. Vagal Maneuvers for Supraventricular Tachycardia (SVT)<sup>9,10,11</sup>

1. **Indications:** Supraventricular tachycardia, 2:1 atrioventricular (AV) block, evaluation of cardiac murmurs.

2. **Contraindications:** Carotid sinus massage is to be avoided in patients with prior stroke within the past 3 months or any history of ventricular arrhythmia.
3. **Complications:** Typically transient (resolve within seconds to minutes) and include prolonged sinus pause, hypertension (increased intrathoracic pressure), hypotension (decreased venous return/decrease in intrathoracic pressure on exhalation).
4. **Procedure:**
  - a. **Cold stimulus to the face:** Briefly place an icepack or washcloth soaked in ice water on the forehead or bridge of the nose. The ice should not be applied for longer than 30 seconds to avoid frostbite.
  - b. **Valsalva maneuver:** Place the patient in supine position and instruct to exhale forcefully against a closed glottis. The strain should be maintained for 10 to 15 seconds before resuming normal breathing.
  - c. **Modified Valsalva maneuver:** Greater success rate at restoring sinus rhythm than standard Valsalva. Place the patient in a semi-recumbent position (45-degree angle), and apply standard Valsalva strain. Immediately reposition supine with 15 seconds of passive leg raise at a 45-degree angle.
  - d. **Carotid sinus massage:** Place the patient in a supine position with neck extension. Apply steady pressure for 5 to 10 seconds to **one** carotid sinus (inferior to the angle of the mandible where pulsation is detected). If unsuccessful, wait 1 to 2 minutes and repeat on the contralateral side.

### B. Heelstick and Fingerstick<sup>12</sup>

1. **Indications:** Blood sampling in infants, obtaining point of care whole blood samples such as serum glucose
2. **Complications:** Infection, bleeding, osteomyelitis.
3. **Procedure:**
  - a. Warm heel or finger.
  - b. Clean with alcohol.
  - c. Using a lancet puncture heel on the lateral aspect, avoiding the posterior area, or finger on the distal palmar lateral pad.
  - d. Wipe away the first drop of blood, and then collect the sample using a capillary tube or container.
  - e. Alternate between squeezing blood from the leg toward the heel (or from the hand toward the finger) and then releasing the pressure for several seconds.

### C. Peripheral Intravenous Access

1. **Indications:** Blood sampling and access to peripheral venous circulation to deliver fluid, medications, or blood products.
2. **Complications:** Thrombosis, infection.
3. **Procedure:**
  - a. Apply tourniquet around the extremity proximal to chosen site.
  - b. Prepare site with alcohol or chlorhexidine.

- c. Insert IV catheter, bevel up, at an angle almost parallel to the skin, advancing until a *flash* of blood is seen in the catheter hub. Advance the plastic catheter only, remove the needle, and secure the catheter.
- d. After removing tourniquet, attach a syringe and apply gentle negative pressure to withdraw blood for serum sampling. Then, attach T connector filled with saline to the catheter, flush with normal saline (NS) to ensure patency of the IV line.

**4. Ultrasound-Guided Procedure:**

- a. With linear ultrasound probe, identify a vein that does not appear to be prohibitively tortuous or stenotic. Perform this by sliding the probe along the course of the vessel and identifying its direction and branching. The saphenous veins in the calves, veins in the forearms, antecubital areas, inside of the upper arms, and external jugular veins are areas where ultrasound guidance can help. An ideal vessel appears less than 1 cm below the skin surface. Deeper vessels are prone to through-and-through perforation of the vessel. Infiltration around deeper vessels is also a risk, as a shorter length of catheter resides in the vessel after insertion.
- b. Prepare the site, and in the case of limb vessels, place a tourniquet proximal to the insertion site.
- c. Under ultrasound visualization, insert the needle into the skin at a shallow (usually <30 degrees) angle to the skin at the midline of the probe near where it contacts the skin. With the probe visualizing the vessel transversely, slowly advance the needle and follow the tip of the needle by sliding the probe away from you. Advance the ultrasound probe until the needle punctures the vessel wall.
- d. Proceed with cannulation of the vessel and secure the intravenous catheter per standard procedure.

**5. Infiltration and Extravasation**<sup>13</sup>: Common injury secondary to fluid infusion into subcutaneous tissues around the venipuncture site. Typically occurs due to puncture of the vein or if the catheter slips out of the vein. The difference between infiltration and extravasation is the type of fluid that has leaked (nonvesicant vs. vesicant). Infiltrations are generally benign, although they can still inflict damage via exertion of mechanical forces on surrounding structures. Extravasation due to a vesicant can cause blistering and burns, leading to necrosis of the tissue. To determine if infiltration/extravasation has occurred, firmly occlude the vein 1 to 2 inches proximal to the insertion site. Continued infusion without resistance indicates infiltration. Immediately stop the infusion. Refer to institutional policy for guidelines regarding application of medication (e.g., hyaluronidase, phentolamine, nitroglycerin ointment). Elevate the affected limb to reduce swelling; apply a warm compress for 10 to 15 minutes; encourage movement of the affected arm. Reevaluate the site every 8 hours.

**6. A video on peripheral IV placement is available on the *New England Journal of Medicine's* website.**

7. A video on **ultrasound-guided peripheral IV placement** is available on the *New England Journal of Medicine's* website.

**D. External Jugular Puncture and Catheterization (see Section XI, Online Content)**

**E. Radial Artery Puncture and Catheterization<sup>2,3</sup>**

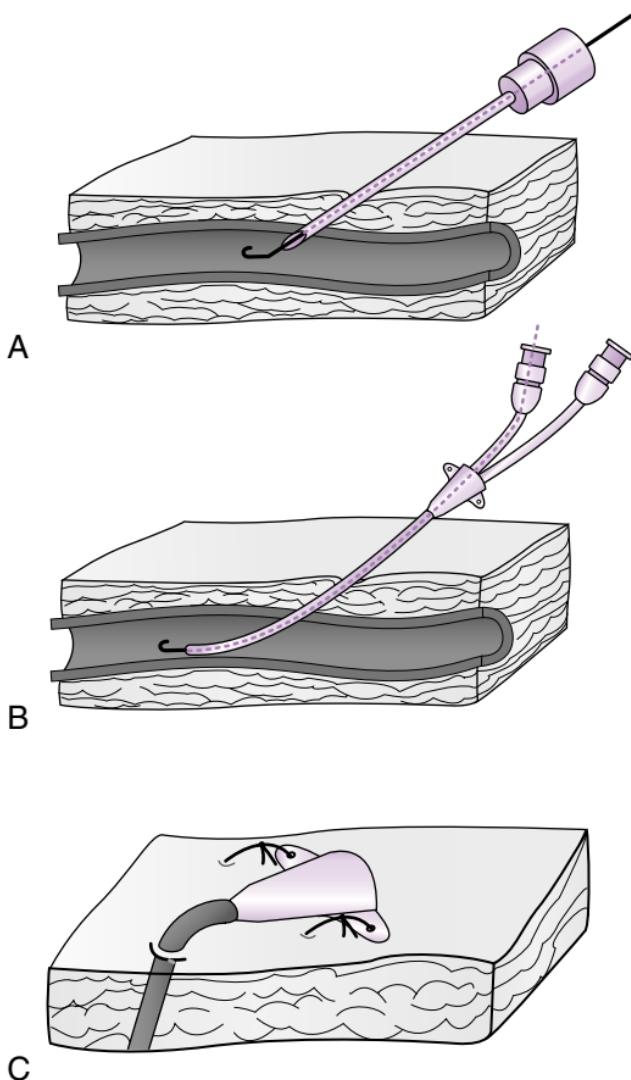
1. **Indications:** Arterial blood sampling or frequent blood gas and continuous blood pressure monitoring in an intensive care setting.
2. **Complications:** Infection, bleeding, occlusion of artery by hematoma or thrombosis, ischemia if ulnar circulation is inadequate.

**3. Procedure:**

- a. Before the procedure, test adequacy of ulnar blood flow with the Allen test: Clench the hand while simultaneously compressing ulnar and radial arteries. The hand will blanch. Release pressure from the ulnar artery, and observe the flushing response. Procedure is safe to perform if the entire hand flushes.
- b. Locate the radial pulse. It is optional to infiltrate the area over the point of maximal impulse with lidocaine. Avoid infusion into the vessel by aspirating before infusing. Prepare the site in a sterile fashion.
- c. Puncture: Insert a butterfly needle attached to a syringe at a 30- to 60-degree angle over the point of maximal impulse. Blood should flow freely into the syringe in a pulsatile fashion. Suction may be required for plastic tubes. Once the sample is obtained, apply firm, constant pressure for 5 minutes and then place a pressure dressing on the puncture site.
- d. Catheter placement: Secure the patient's hand to an arm board. Leave the fingers exposed to observe any color changes. Prepare the wrist with sterile technique and infiltrate over the point of maximal impulse with 1% lidocaine. Insert an IV catheter with its needle at a 30-degree angle to the horizontal until a flash of blood is seen in the catheter hub. Advance the plastic catheter and remove the needle. Alternatively, pass the needle and catheter through the artery to transfix it, and then withdraw the needle. Very slowly, withdraw the catheter until free flow of blood is noted, then advance the catheter and secure in place using sutures or tape. Seldinger technique (Fig. 4.2) using a guidewire can also be used. Apply a sterile dressing and infuse heparinized isotonic fluid (per institutional protocol) at a minimum of 1 mL/hr. A pressure transducer may be attached to monitor blood pressure.
- e. Suggested size of arterial catheters based on weight:
  - (1) Infant (<10 kg): 24 G or 2.5 Fr, 2.5 cm
  - (2) Child (10 to 40 kg): 22 G or 2.4 Fr, 2.5 cm
  - (3) Adolescent (>40 kg): 20 G

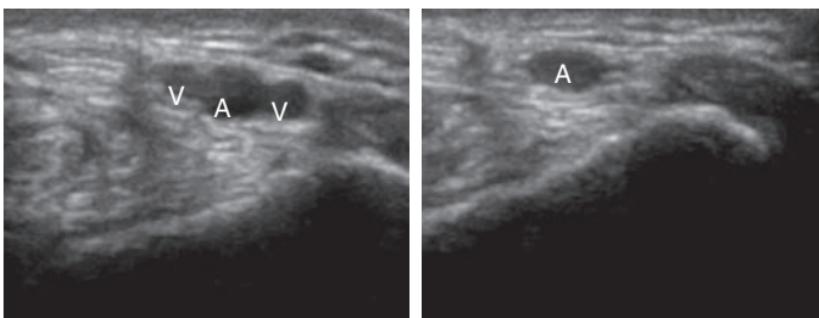
**4. Ultrasound-Guided Procedure**

- a. Use the linear probe. After the sterile field has been prepped, apply gel to the probe and place within a sterile cover. Place the ultrasound probe transverse to the artery on the radial, posterior tibial, or

**FIGURE 4.2**

Seldinger technique. (A) Guidewire is placed through introducer needle into lumen of vein. (B) Catheter is advanced into vein lumen along guidewire. (C) Hub of catheter is secured to skin with suture. (Modified from Fuhrman B, Zimmerman J. Pediatric Critical Care. 4th ed. Philadelphia: Mosby; 2011.)

dorsalis pedis pulse. Identify the artery, which will appear pulsatile with some compression. Once the artery has been identified, center the probe over the vessel (Fig. 4.3). Insert the needle into the skin at a 45-degree angle at the midline of the probe near where it contacts the skin. With the probe visualizing the vessel transversely, slowly

**FIGURE 4.3**

Ultrasound transverse view of radial artery. In the left image, the radial artery is seen in cross section with veins on either side. On the right image, pressure has been applied and the veins are collapsed while the artery remains patent. A, Artery; V, vein. (From Weiner MM, Geldard P, Mitnacht AJC. Ultrasound guided vascular access: a comprehensive review. J Cardiothorac Vasc Anesth. 2013;27(2):345–360.)

advance the needle and follow the tip of the needle by sliding the probe away. Advance the ultrasound probe until the needle punctures the vessel wall. Proceed with the rest of the procedure after vessel puncture, as described previously.

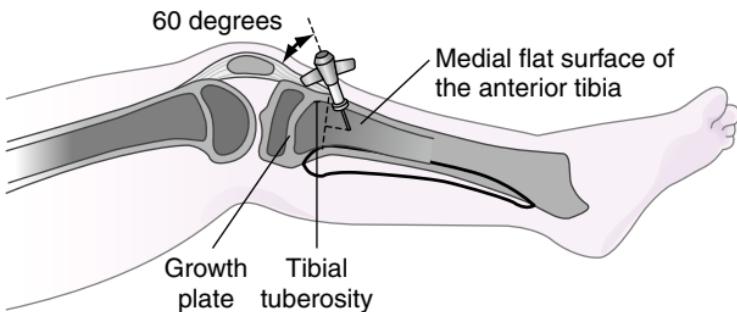
5. **Videos on arterial puncture and radial artery catheterization are available on the New England Journal of Medicine's website.**
6. **A video on ultrasound-guided radial artery catheterization is available on the New England Journal of Medicine's website.**

#### **F. Posterior Tibial and Dorsalis Pedis Artery Puncture**

1. **Indications:** Arterial blood sampling when radial artery puncture is unsuccessful or inaccessible.
2. **Complications:** Infection, bleeding, ischemia if circulation is inadequate.
3. **Procedure:**
  - a. Posterior tibial artery: Puncture the artery posterior to medial malleolus while holding the foot in dorsiflexion.
  - b. Dorsalis pedis artery: Puncture the artery at dorsal midfoot between first and second toes while holding the foot in plantar flexion.

#### **G. Intraosseous (IO) Access<sup>2,3</sup> (Fig. 4.4)**

1. **Indications:** Obtain emergency access in children during life-threatening situations. This is very useful during cardiopulmonary arrest, shock, burns, and life-threatening status epilepticus. Any crystalloid, blood product, or drug that may be infused into a peripheral vein may also be infused into the IO space. The IO needle should be removed once adequate vascular access has been established. Insertion of IO needle into fractured bones should be avoided.
2. **Complications:**
  - a. Complications are rare, particularly with the correct technique. Frequency of complications increases with prolonged infusions.

**FIGURE 4.4**

Intraosseous needle placement using standard anterior tibial approach. Insertion point is in the midline on medial flat surface of anterior tibia, 1 to 3 cm (2 fingerbreadths) below tibial tuberosity. (From Dieckmann R, Fiser D, Selbst S. Pediatric Emergency and Critical Care Procedures. St. Louis: Mosby; 1997.)

- b. Complications include extravasation of fluid from incomplete or through and through cortex penetration, infection, bleeding, osteomyelitis, compartment syndrome, fat embolism, fracture, epiphyseal injury.

**3. Sites of entry (in order of preference):**

- a. Anteromedial surface of the proximal tibia, 2 cm below and 1 to 2 cm medial to the tibial tuberosity on the flat part of the bone.
- b. Distal femur 3 cm above the lateral condyle in the midline.
- c. Medial surface of the distal tibia 1 to 2 cm above the medial malleolus (may be a more effective site in older children).
- d. Proximal humerus, 2 cm below the acromion process into the greater tubercle with the arm held in adduction and internal rotation.
- e. Anterosuperior iliac spine at a 90-degree angle to the long axis of the body.

**4. Procedure:**

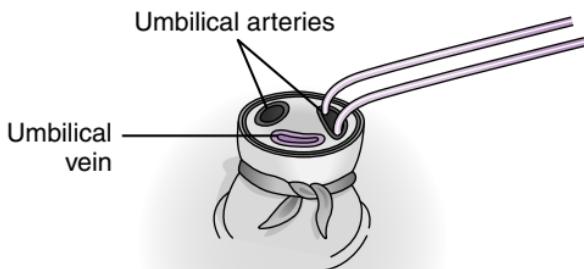
- a. Prepare the selected site in a sterile fashion.
- b. If the child is conscious, anesthetize the puncture site down to the periosteum with 1% lidocaine (optional in emergency situations).
- c. Choose between a manual IO or drill-powered IO insertion device:
  - (1) For manual IO needle: Insert a 15- to 18-gauge IO needle perpendicular to the skin at an angle away from the epiphyseal plate, and advance to the periosteum. With a boring rotary motion, penetrate through the cortex until there is a decrease in resistance, indicating that you have reached the marrow. The needle should stand firmly without support. Secure the needle carefully.
  - (2) For drill-powered IO needle: Enter skin with the needle perpendicular to the skin, as with the manual needle, and press the needle until you meet the periosteum. Then apply easy pressure

while gently depressing the drill trigger until you feel a decrease in resistance. Remove the drill while holding the needle steady to ensure stability prior to securing the needle. Use an EZ-IO AD for patients greater than 40 kg, and use EZ-IO PD for patients greater than 6 kg and less than 40 kg.

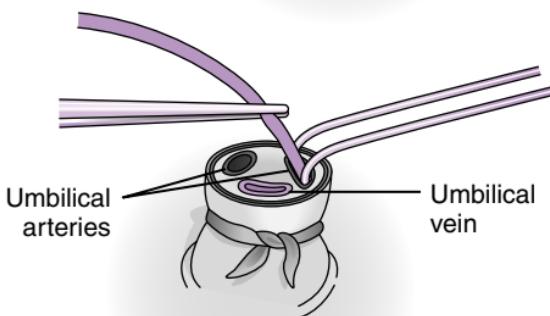
- d. Remove the stylet and attempt to aspirate marrow. (Note that it is not necessary to aspirate marrow.) Flush with crystalloid solution. Observe for fluid extravasation. Marrow can be sent to determine glucose levels, chemistries, blood types and cross-matches, hemoglobin levels, blood gas analyses, and cultures.
  - e. Attach standard IV tubing. Increased pressure (through pressure bag or push) may be necessary for infusion. There is a high risk for obstruction if continuous high-pressure fluids are not flushed through the IO needle.
5. A video on IO catheter placement is available on the *New England Journal of Medicine's* website.

## H. Umbilical Artery and Umbilical Vein Catheterization<sup>2</sup>

1. **Indications:** Vascular access (via umbilical vein [UV]), blood pressure monitoring (via umbilical artery [UA]), or blood gas monitoring (via UA) in critically ill neonates.
2. **Complications:** Infection, bleeding, hemorrhage, perforation of vessel, thrombosis with distal embolization, ischemia or infarction of lower extremities, bowel, or kidney, arrhythmia if catheter is in the heart, air embolus.
3. **Contraindications:** Omphalitis, peritonitis, possible/confirmed necrotizing enterocolitis, intestinal hypoperfusion.
4. **Line placement:**
  - a. Umbilical arterial catheter (UAC) line: Low line vs. high line.
    - (1) Low line: Tip of catheter should lie just above the aortic bifurcation between L3 and L5. This avoids renal and mesenteric arteries near L1, possibly decreasing the incidence of thrombosis or ischemia.
    - (2) High line: Tip of catheter should be above the diaphragm between T6 and T9. A high line may be recommended in infants weighing less than 750 g, in whom a low line could easily slip out.
  - b. Umbilical venous catheter (UVC) lines should be placed in the inferior vena cava above the level of the ductus venosus and the hepatic veins and below the level of the right atrium.
  - c. Catheter length: Determine the length of catheter required using either a standardized graph based on shoulder-umbilical length or the following birth weight (BW) regression formula:
    - (1) UAC Low Line (cm) = BW (kg) × 7
    - (2) UAC High Line (cm) = (3 × BW (kg)) + 9
    - (3) UVC Length (cm) = 0.5 × UAC high line (cm) + 1.
5. **Procedure for UAC line (Fig. 4.5):**



A



B

**FIGURE 4.5**

Placement of umbilical arterial catheter. (A) Dilating lumen of umbilical artery. (B) Insertion of umbilical artery catheter. (From Dieckmann R, Fiser D, Selbst S. Pediatric Emergency and Critical Care Procedures. St. Louis: Mosby; 1997.)

- a. Determine the length of the catheter to be inserted for either high (T6 to T9) or low (L3 to L5) position.
- b. Restrain infant. Maintain the infant's temperature during the procedure. Prepare and drape the umbilical cord and adjacent skin using sterile technique.
- c. Flush the catheter with sterile saline solution before insertion. Ensure that there are no air bubbles in the catheter or attached syringe.
- d. Place sterile umbilical tape around the base of the cord. Cut through the cord horizontally about 1.5 to 2 cm from the skin; tighten the umbilical tape to prevent bleeding.
- e. Identify the one large, thin-walled UV and two smaller, thick-walled arteries. Use one tip of open, curved forceps to gently probe and dilate one artery. Then use both points of closed forceps, and dilate artery by allowing forceps to open gently.

- f. Grasp the catheter 1 cm from its tip with toothless forceps and insert the catheter into the lumen of the artery. Aim the tip toward the feet and gently advance the catheter to the desired distance. *Do not force.* If resistance is encountered, try loosening umbilical tape, applying steady and gentle pressure, or manipulating the angle of the umbilical cord to the skin. Often the catheter cannot be advanced because of the creation of a “false luminal tract.” There should be good blood return when the catheter enters the iliac artery.
  - g. Confirm catheter tip position with x-ray or ultrasound. Secure catheter with a suture through the cord, a marker tape, and a tape bridge. The catheter may be pulled back but *not* advanced once the sterile field is broken.
  - h. Observe for complications: Blanching or cyanosis of lower extremities, perforation, thrombosis, embolism, or infection. If any complications occur, the catheter should be removed.
  - i. Use isotonic fluids containing heparin per institutional policy. Never use hypoosmolar fluids in the UA.
6. **Procedure for UVC line (see Fig. 4.5):**
- a. Determine the desired length and follow steps “a” through “d” for UA catheter placement.
  - b. Isolate the thin-walled UV, clear thrombi with forceps, and insert catheter, aiming the tip toward the right shoulder. Gently advance the catheter to the desired distance. *Do not force.* If resistance is encountered, try loosening the umbilical tape, applying steady and gentle pressure, or manipulating the angle of the umbilical cord to the skin. Resistance is commonly met at the abdominal wall and again at the portal system. *Do not infuse anything into the liver.*
  - c. Confirm catheter tip position with x-ray or ultrasound. Secure catheter with a suture through the cord, a marker tape, and a tape bridge. The catheter may be pulled back but *not* advanced once the sterile field is broken.
7. A video on **UVC/UAC line placement** is available on the *New England Journal of Medicine’s website.*

## VI. PULMONARY PROCEDURES

### A. Use of Metered-Dose Inhalers and Spacer<sup>6</sup>

1. **Indications:** Delivery of medication to distal airways in the lungs.
2. **Complications:** Failure of medication delivery. **Note** that there are risks associated with the medication rather than the delivery method.
3. **Procedure:**
  - a. Shake the inhaler, remove the cap, and attach it to the spacer device.
  - b. Instruct the child to exhale completely.
  - c. Place the mouthpiece of the spacer into the patient’s mouth, and instruct the child to make a complete seal with the lips. Alternatively, a spacer with a mask can be placed over the child’s mouth if they are unable to make a seal with their lips.

- d. Spray 1 puff from the inhaler into the spacer and instruct the patient to breathe slowly and deeply, holding the breath for 10 seconds.
- e. Wait 1 minute and repeat as indicated.

### B. Needle Cricothyrotomy<sup>6,14</sup>

1. **Indications:** When an emergency airway is required and the clinician is unable to use bag-mask ventilation or secure an orotracheal or nasotracheal airway. Common indications include facial fractures, blood or vomitus in the airway, airway obstruction (e.g., foreign body, tumor, edema from trauma).
2. **Contraindications:** No absolute contraindications. Relative contraindications include unable to locate landmarks, laryngotracheal damage, coagulopathy, bleeding dyscrasias.
3. **Complications:** Bleeding, hypoxia, pneumothorax, esophageal laceration, vocal cord injury, posterior tracheal wall perforation, infection.
4. **Procedure:**
  - a. Immobilize the larynx with the nondominant hand and identify the cricothyroid membrane. This is located by palpating the laryngeal prominence at midline of the thyroid cartilage and then moving distally 1 to 2 cm to a small depression. This depression overlies the cricothyroid membrane.
  - b. Insert a 12 to 14-gauge *angiocatheter* caudally at a 30- to 45-degree angle through the cricothyroid membrane while aspirating the needle as it is inserted.
  - c. Attach the needle to an oxygen source that can deliver roughly 30 psi. Alternatively, a bag-valve device can be connected using a 7.0 endotracheal tube adapter and a 3 mL syringe with plunger removed.
  - d. Intermittent ventilation can be achieved by cutting a small hole in the oxygen tubing, and covering the hole in the tubing. Allow for expiration by uncovering the hole for 4 to 10 seconds.

### C. Needle Thoracostomy<sup>2,15</sup>

1. **Indications:** Evacuation of a pneumothorax, hemothorax, chylothorax, large pleural effusion, or empyema for diagnostic or therapeutic purposes.
2. **Complications:** Infection, bleeding, pneumothorax, hemothorax, pulmonary contusion or laceration, puncture of diaphragm, spleen, or liver, or bronchopleural fistula.
3. **Procedure:**
  - a. Prepare and drape the skin as clean as possible, with goal of sterility.
  - b. Insert a large-bore angiocatheter (14- to 22-gauge based on patient size and likely depth of the chest wall) into the anterior second intercostal space in the midclavicular line. Insert needle over superior aspect of rib margin to avoid neurovascular structures. If the angiocatheter permits, a 3- to 10-mL syringe with 1 to 2 mL of saline can be connected to it. Aspirating the syringe while inserting the IV will pull air bubbles through the saline if an air collection exists. A rush of bubbles signifies successful access.

- c. When pleural space is entered, withdraw needle and attach catheter to a three-way stopcock and syringe, and aspirate air. The stopcock is used to stop air flow through the catheter when sufficient evacuation has been performed.
  - d. Subsequent insertion of a chest tube is often necessary for ongoing release of air. It is advised not to completely evacuate chest prior to placement of chest tube to avoid pleural injury.
4. A video on **needle decompression of spontaneous pneumothorax** is available on the *New England Journal of Medicine's* website.

## VII. GASTROINTESTINAL PROCEDURES

### A. Nasogastric Tube Placement<sup>6,16</sup>

- 1. **Indications:** Enteral nutrition, administration of medications, treatment of ileus or obstruction, gastric decompression.
- 2. **Contraindications:** Esophageal stricture, esophageal varices, severe mid-face trauma (cribriform plate disruption), bleeding diatheses, alkaline ingestion.
- 3. **Complications:** Malposition, coiling of tube, esophageal perforation, pneumothorax.
- 4. **Procedure:**
  - a. Approximate the length of 6- to 10-Fr tube insertion by positioning the tube from the nares or mouth to the ear, then to the mid-xiphoid-umbilicus. Mark this length on the tube with marker.
  - b. The patient should be sitting as upright as possible. The head should be tilted toward the chest.
  - c. Lubricate the tube and insert the tube through the nose. Advance the tube to the length mark, asking the patient to swallow while the tube is inserted. It may be helpful to provide a cup of water with a straw.
  - d. Confirm placement of the tube with a radiograph of the lower chest/upper abdomen. Ensure that the tube is located distal to the carina, crosses the diaphragm, and rests in a central position in the gastric region. The tube should not cross the midline. Additional confirmation can be obtained by testing the pH of aspirated contents. A of pH 1 to 4 confirms proper positioning. Alternatively, insert a small amount of air (20 to 30 mL) through the tube while listening to the gastric area with a stethoscope.
  - e. Secure the tube.

### B. Gastrostomy Tube Replacement<sup>6,17</sup>

- 1. **Indications:** Dislodged, blocked, or replacement of gastrostomy tube (G-tube) or gastrostomy button.
- 2. **Complications:** Perforation, bleeding, pneumoperitoneum, creation of "false track" particularly if tube is newly placed. **Note** that misplacement and associated complications are rare for children with a mature G-tube track undergoing tube replacement in a pediatric emergency room.

**3. Procedure:**

- a. Deflate balloon completely with a syringe and pull the tube out steadily.
- b. Insert new tube in the stoma and inflate balloon fully with water. Gently tug on the tube to assess whether the balloon is inflated. Secure the tube.
- c. Confirm intragastric placement by aspirating gastric contents.
- d. If replacement tube is not immediately available, a Foley catheter of similar size may be placed using the method above to maintain tract patency.
- e. If the G-tube track is too constricted for placement of G-tube, consider upsizing with Foley catheter serial dilation.

**4. A video on gastrostomy tube exchange is available on the *New England Journal of Medicine's* website.**

### VIII. GENITOURINARY PROCEDURES

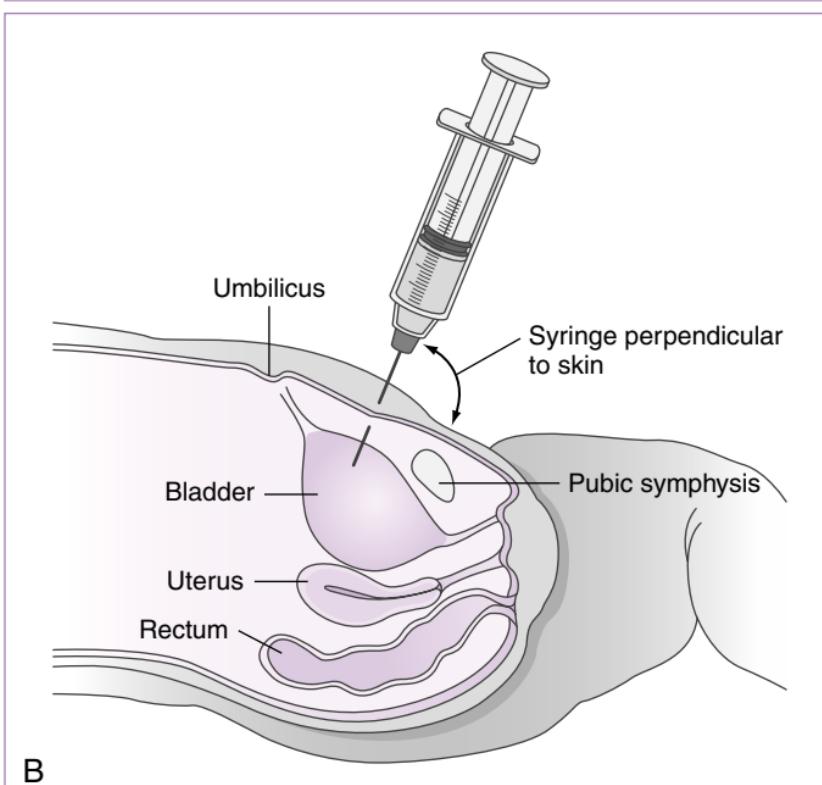
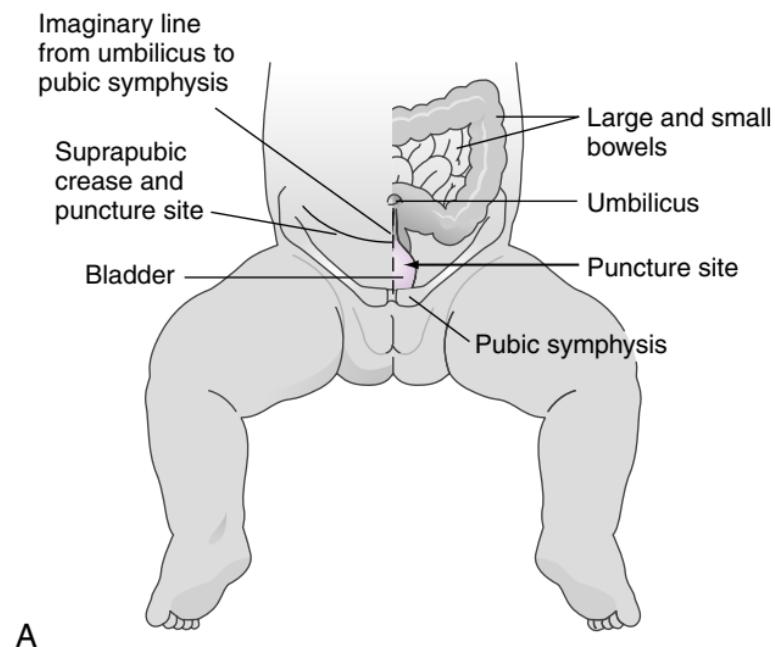
**A. Urinary Bladder Catheterization<sup>3,6,18</sup>**

1. **Indications:** To obtain urine for urinalysis and sterile culture, to accurately monitor hydration status, and bladder decompression.
2. **Complications:** Hematuria, infection, trauma to urethra or bladder, intravesical knot of catheter (rarely occurs).
3. **Contraindications:** Pelvic fractures, known trauma to the urethra, or blood at the meatus.
4. **Catheter Selection:** 5 Fr for children younger than 6 months; 8 Fr for children between 6 months and adolescence; 10 Fr for adolescents.
5. **Procedure:**
  - a. For collection of urinalysis and/or urine culture, the infant/child should not have voided within 1 hour of procedure.
  - b. Prepare the urethral opening using sterile technique.
  - c. In males, apply gentle traction to the penis to straighten the urethra. In uncircumcised male infants, expose the meatus with gentle retraction of the foreskin. The foreskin has to be retracted only far enough to visualize the meatus.
  - d. In girls, the urethral orifice may be difficult to visualize, but it is usually immediately superoanterior to the vaginal orifice.
  - e. Gently insert a lubricated catheter into the urethra. Slowly advance catheter until resistance is met at the external sphincter. Continued pressure will overcome this resistance, and the catheter will enter the bladder. Only a few centimeters of advancement are required to reach the bladder in girls. In boys, insert a few centimeters longer than the shaft of the penis.
  - f. Carefully remove the catheter once specimen is obtained, and cleanse skin of iodine.
  - g. If indwelling Foley catheter is inserted, inflate balloon with sterile water or saline as indicated on bulb, then connect catheter to drainage tubing attached to urine drainage bag. Secure catheter tubing to inner thigh.

6. Videos on catheterization of the male urethra and catheterization of the female urethra are available on the *New England Journal of Medicine's website*.

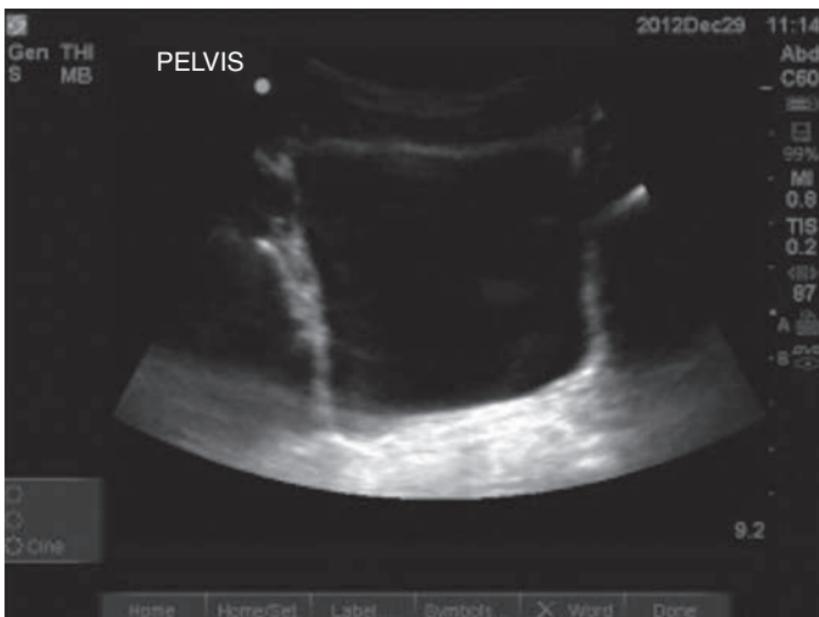
### B. Suprapubic Bladder Aspiration<sup>2</sup>

1. **Indications:** To obtain urine in a sterile manner for urinalysis and culture in children younger than 2 years (avoid in children with genitourinary tract anomalies, coagulopathy, or intestinal obstruction). This bypasses distal urethra, thereby minimizing risk for contamination.
2. **Complications:** Infection (cellulitis), hematuria (usually microscopic), intestinal perforation.
3. **Procedure (Fig. 4.6):**
  - a. Anterior rectal pressure in girls or gentle penile pressure in boys may be used to prevent urination during the procedure. Child should not have voided within 1 hour of procedure.
  - b. Restrain child in the supine, frog leg position. Prepare suprapubic area in a sterile fashion.
  - c. The site for puncture is 1 to 2 cm above the symphysis pubis in the midline. Use a syringe with a 22-gauge, 1-inch needle, and puncture at a 10- to 20-degree angle to the perpendicular, aiming slightly caudad.
  - d. Ultrasound guidance:
    - (1) Ultrasound can be used to visualize the urinary bladder for this procedure as follows: Use the curvilinear or linear probe. Apply the probe in transverse position in the midline of the lower abdomen, positioning it to locate the bladder. The bladder is a midline structure with a dark center and bright margins. The shape of the bladder is usually rounded; however, it can appear spherical, pyramidal, or even cuboidal (Fig. 4.7).
    - (2) The bladder may be empty as well with no dark cavity. If no clear structure, give fluids and reassess in 30 minutes. This technique can also be used in the evaluation of anuric patients, to differentiate between decreased urine production and urinary retention. This is also useful in the case of patients with a urinary catheter as the catheter is usually visible. If it is visualized and the bladder also has urine around it, the catheter is likely malfunctioning.
    - (3) Aspiration can be performed after marking the site with ultrasound, proceeding with preparing and draping the patient and proceeding to puncture.
  - e. **Gently exert suction as the needle is advanced until urine enters syringe.** The needle should not be advanced more than 3 cm. Aspirate urine with gentle suction.
  - f. Remove needle, cleanse skin of iodine, and apply a sterile bandage.
4. A video of suprapubic bladder aspiration is available on the *New England Journal of Medicine's website*.



**FIGURE 4.6**

Landmarks for suprapubic bladder aspiration. (Modified from Dieckmann R, Fiser D, Selbst S. Pediatric Emergency and Critical Care Procedures. St. Louis: Mosby; 1997.)

**FIGURE 4.7**

Ultrasound of bladder. In this transverse midline view of the pelvis the bladder appears black (anechoic) and cuboid in the midline. This is the typical appearance of a full bladder on ultrasound, although the shape may vary. (From Leeson K, Leeson B. *Pediatric ultrasound: applications in the emergency department*. Emerg Med Clin North Am. 2013;31(3):809–829.)

## IX. MUSCULOSKELETAL PROCEDURES

### A. Basic Splinting<sup>2</sup>

1. **Indications:** To provide short-term stabilization of limb injuries while accommodating swelling associated with acute injuries.
2. **Complications:** Pressure sores, dermatitis, neurovascular impairment.
3. **Procedure:**
  - a. Determine style of splint needed (see [Section IX.B](#)).
  - b. Measure and cut fiberglass or plaster to appropriate length. If using plaster, upper-extremity splints require 8 to 10 layers and lower-extremity splints require 12 to 14 layers.
  - c. Pad extremity with copious cotton roll padding, taking care to overlap each turn by 50%. In prepackaged fiberglass splints, additional padding is not generally required. Bony prominences may require additional padding. Place cotton between digits if they are in a splint.
  - d. Immerse plaster slabs into room temperature water until bubbling stops. Smooth out wet plaster slab, avoiding any wrinkles. Fiberglass splints will harden when exposed to air; however, application of a small amount of room temperature water can accelerate this process.
  - e. Position splint over extremity and mold to desired contour. Wrap with an elastic bandage to hold molded splint onto extremity in position of

function. Continue to hold desired form of splint upon extremity until fully hardened.

- f. **NOTE:** Plaster becomes warm while drying. Using warm water will decrease drying time. This may result in inadequate time to mold splint. Turn edge of the splint back on itself to produce a smooth surface. Take care to cover the sharp edges of fiberglass.
- g. Use crutches or slings as indicated.
- h. The need for orthopedic referral should be individually assessed.
- i. Emergent orthopedic referral may be required, including when there is concern for neurovascular compromise or compartment syndrome of the affected extremity.

#### 4. **Postsplint Care:**

- a. Standard rest, ice, and elevation of affected extremity should be performed.
- b. Avoid weight bearing on splinted extremity.
- c. Do not get splint wet. Splints can be wrapped in water-resistant items such as a plastic bag or a specially designed splint bag to allow for showering. Use a hair dryer in instances where the splint has accidentally gotten wet.
- d. Do not stick items such as a pen or clothes hanger to scratch inside the splint.
- e. If areas in or distal to the splint develop numbness, tingling, increased pain, turn blue or pale, or become swollen, patient should loosen the elastic bandage of the splint. Instruct to seek immediate medical care if this does not quickly (<30 minutes) resolve these symptoms.

#### 5. A video on basic splinting techniques is available on the *New England Journal of Medicine's* website.

### B. Selected Splints and Indications (Fig. 4.8)

#### 1. Long Arm Posterior Splint

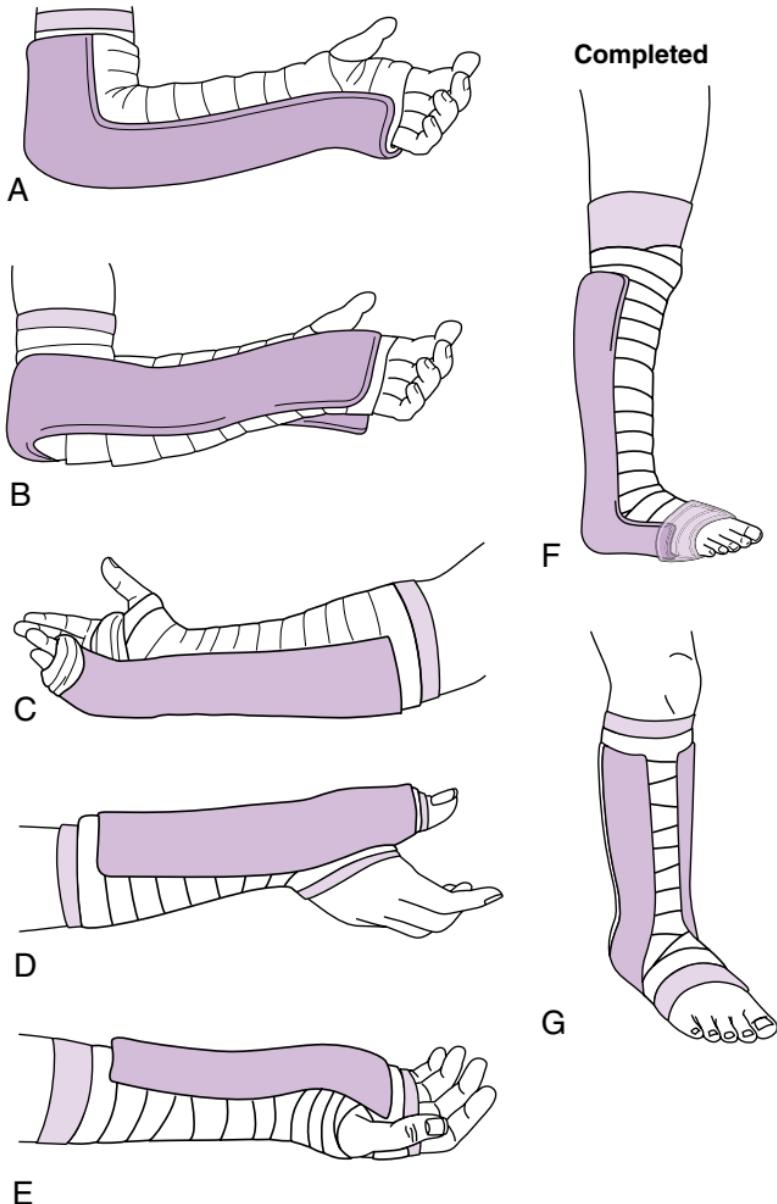
- a. Indications: Immobilization of elbow and forearm injuries.
- b. Procedure: Elbow flexed at 90 degrees, forearm in neutral position, slight dorsiflexion of the wrist. Splint extends from palmar crease of the hand to mid upper arm along the ulnar side of the forearm and the posterior aspect of the humerus. Width should be semicircumferential.

#### 2. Sugar Tong Forearm Splint

- a. Indications: For distal radius and wrist fractures; to immobilize the elbow and minimize pronation and supination.
- b. Procedure: Elbow flexed at 90 degrees, forearm in neutral position, and slight dorsiflexion of the wrist. Splint extends from palmar crease along volar aspect of forearm, around elbow, and dorsally to the metacarpals. Fingers and thumb remain free. Width should support arm on both sides but not overlap.

#### 3. Ulnar Gutter Splint

- a. Indications: Nonrotated fourth or fifth (boxer) metacarpal metaphyseal fracture with less than 20 degrees of angulation, uncomplicated fourth and fifth phalangeal fracture.

**FIGURE 4.8**

Selected splint types. Light purple layer is stockinette, white layer is cotton roll, dark purple layer is the splint. (A) Long arm posterior splint. (B) Sugar tong forearm splint. (C) Ulnar gutter splint. (D) Thumb spica splint. (E) Volar splint. (F) Posterior ankle splint. (G) Ankle stirrup splint.

- b. Assess for malrotation, displacement (especially Salter I type fracture), angulation, and joint stability before splinting.
- c. Procedure: Elbow in neutral position, wrist in slight dorsiflexion, metacarpophalangeal (MP) joint at 60 to 90 degrees, interphalangeal (IP) joint at 20 degrees. Apply splint in U shape from the tip of the fifth digit to 3 cm distal to the volar crease of the elbow. Splint should be wide enough to enclose the fourth and fifth digits.

#### 4. **Thumb Spica Splint**

- a. Indications: Nonrotated, nonangulated, nonarticular fractures of the thumb metacarpal or phalanx, ulnar collateral ligament injury (gamekeeper's or skier's thumb), scaphoid fracture or suspected scaphoid fracture (pain in anatomic snuff box).
- b. Procedure: Wrist in slight dorsiflexion, thumb in some flexion and abduction, IP joint in slight flexion. Apply splint in U shape along radial side of forearm extending from tip of thumb to mid-forearm. Mold the splint along the long axis of the thumb so that thumb position is maintained. This will result in a spiral configuration along the forearm with maintained apposition of the index finger and thumb.

#### 5. **Volar Splint**

- a. Indications: Wrist immobilization for wrist sprains, strains, or certain fractures.
- b. Procedure: Wrist in slight dorsiflexion. Apply splint on palmar surface from the MP joint to 2 to 3 cm distal to the volar crease of the elbow. It is useful to curve the splint to allow the MP joint to rest at an 80- to 90-degree angle.

#### 6. **Posterior Ankle Splint**

- a. Indications: Immobilization of ankle sprains and fractures of the foot, ankle, and distal fibula.
- b. Procedure: Place patient in prone position with ipsilateral knee flexed at 90 degrees and affected ankle held in flexion at 90 degrees. Splint should extend from base of toes to upper portion of the calf. Width should match that of the foot. An ankle stirrup (sugar tong) splint can be added to increase stability for ankle fractures.

#### 7. **Ankle Stirrup Splint**

- a. Indications: Immobilization of the ankle.
- b. Procedure: Ankle held in flexion at 90 degrees. Splint extends in U-shaped fashion from fibular head underneath the ankle to just below the knee. Width should be one half of the narrowest circumference of the lower leg and not overlapping. May be used alone or in combination with (placed after) posterior ankle splint.

#### C. Radial Head Subluxation (Nursemaid's Elbow) Reduction<sup>19</sup>

- 1. **Presentation:** Commonly occurs in children aged 1 to 4 years with a history of inability to use an arm after it was pulled. Child presents with affected arm held at the side in pronation, with elbow slightly flexed.

2. **Caution:** Rule out a fracture clinically before doing procedure. Consider radiograph if mechanism of injury or history is atypical or if exam is concerning for fracture (swelling, bruising, tenderness, etc.).
  3. **Procedure:**
    - a. Two most common techniques include hyperpronation (HP) and traditional supination-flexion (SF) maneuvers. Recent meta-analyses of randomized trials evaluating the two techniques favor HP for both efficacy and pain tolerance.
    - b. Support the elbow with one hand, and place your thumb laterally over the radial head at the elbow applying pressure medially. With your other hand, grasp the child's hand in a handshake position or at the wrist. The child's thumb should point downward.
    - c. HP method: Forcefully pronate the wrist. You may feel a click as reduction occurs.
    - d. SF method: Quickly and deliberately supinate and externally rotate the forearm, and simultaneously flex the elbow.
    - e. Most children will begin to use the arm within 15 minutes, some immediately after reduction. If reduction occurs after a prolonged period of subluxation, it may take the child longer to recover use of the arm. In this case, the arm should be immobilized with a posterior splint.
    - f. If procedure is unsuccessful, consider obtaining a radiograph. Maneuver may be repeated if needed.
  4. A video on reduction of nursemaid's elbow is available on the *New England Journal of Medicine's* website.
- D. Finger/Toe Dislocation Reduction<sup>2</sup>**
1. **Indications:** IP and MP/metatarsophalangeal dislocations.
  2. **Complications:** Fracture of phalanges, entrapment of neurovascular structures.
  3. **Cautions:** Volar dislocations and dorsal dislocations with interposition of the volar plate or entrapment of the metacarpal/metatarsal head often cannot be performed using closed reduction.
  4. **Procedure:**
    - a. Evaluate for neurovascular compromise in the affected digit. Perform radiographs to evaluate for possible fracture.
    - b. Consider procedural sedation or a digital block prior to procedure.
    - c. Grasp the digit proximal to fracture to allow for stabilization.
    - d. Grasp the tip of the distal digit and apply longitudinal traction, with the joint typically slipping into place.
    - e. Alternatively, grasp the distal phalanx and mildly hyperextend to accentuate the deformity while applying longitudinal traction.
    - f. After reduction, again evaluate neurovascular status and obtain radiographs to ensure proper position and to further evaluate for fracture.
    - g. Immobilize the joint using a padded splint using full extension for distal IP joints and 20 to 30 degrees of flexion for proximal IP joints.

## E. Knee Arthrocentesis<sup>2</sup>

1. **Indications:** Evaluation of fluid for the diagnosis of disease, including infectious, inflammatory, and crystalline disease, and removal of fluid for relief of pain and/or functional limitation.
2. **Contraindications:** Bleeding diathesis, local fracture, overlying skin infection.
3. **Complications:** Pain, bleeding, infection.
4. **Procedure:**
  - a. Place child supine on exam table with knee in slight flexion, with use of a padded roll underneath the knee for support, if unable to slightly flex.
  - b. The lateral or medial approach can be made, with the lateral approach preferred to avoid the vastus medialis muscle.
  - c. The puncture point should be at the posterior margin of the patella in both cases.
  - d. Prepare the overlying skin in a sterile fashion, and once cleaned, numb the area using 1% lidocaine with a small-gauge needle. Then, using an 18-gauge needle attached to a syringe, puncture the skin at a 10- to 20-degree downward angle, and advance under continuous syringe suction until fluid is withdrawn, indicating entry into the joint space.
  - e. In large effusions, several syringes may be needed for complete fluid removal if so desired, and the needle may have to be redirected to access pockets of fluid.
  - f. Upon completion, withdraw the needle and cover the wound with a sterile gauze dressing.
  - g. Synovial fluid can then be sent for studies as indicated.
5. A video on knee arthrocentesis is available on the *New England Journal of Medicine's* website.

## F. Hematoma Blocks<sup>20</sup>

1. **Indications:** Analgesia for closed fracture of the extremity that requires manipulation or closed reduction. It is an alternative when procedural sedation is not possible or is impractical.
2. **Contraindications:** Allergic reactions to local anesthetic agents, open fracture, cellulitis overlying fracture site, presence of a neurovascular deficit or vascular deficit.
3. **Complications:** Rare, but include compartment syndrome, local anesthetic toxicity (circumoral and tongue numbness, dizziness, tinnitus, and visual disturbances), and osteomyelitis.
4. **Procedure:**
  - a. Perform using aseptic technique.
  - b. Draw up the local anesthetic solution into a syringe with a 22- or 23-gauge, 2-inch-long needle. Bupivacaine, for postprocedure analgesia, is desired and can be used alone or mixed with lidocaine in a 50:50 ratio. (See Chapter 6 for dosage maximum.)
  - c. Place a wheal of 1% lidocaine subcutaneously over the fracture site. Allow 1 to 2 minutes for the anesthetic to take effect.

- d. Slowly insert and advance the needle attached to the local anesthetic solution through the skin wheal and aimed at the fracture site. C-arm fluoroscopy can aid fracture/hematoma localization. Slowly advance the needle.
  - (1) Aspirate with the syringe to **ensure there is no free flow of blood**, which indicates that the needle is within a blood vessel. If there is free blood flow, do not inject the local anesthetic solution.
  - (2) A flash, without flow, of blood indicates entry of the tip of the needle into the hematoma.
  - (3) Redirect needle if you strike bone or if no flash of blood is returned.
- e. Once flash, without flow, is obtained, slowly inject the local anesthetic solution into the hematoma.
- f. Reposition the needle to different areas within the hematoma and inject small amounts of the local anesthetic into each area. This technique distributes the local anesthetic solution to increase the efficacy of the hematoma block and minimizes the risk of intravascular injection of the entire dose of local anesthetic.
- g. Withdraw the needle, apply a bandage to the skin puncture site, and await analgesia.

## X. SKIN/DERMATOLOGIC PROCEDURES

### A. Immunization and Medication Administration<sup>3</sup>

**NOTE:** Please see [Chapters 16](#) and [30](#) for relevant vaccines and medications and their appropriate administration routes.

#### 1. Subcutaneous Injections

- a. **Indications:** Immunizations and medications.
- b. **Complications:** Bleeding, infection, allergic reaction, lipohypertrophy or lipoatrophy after repeated injections.
- c. **Procedure:**
  - (1) Locate injection site: Upper outer arm or outer aspect of upper thigh.
  - (2) Cleanse skin with alcohol.
  - (3) Insert 0.5-inch, 25- or 27-gauge needle into subcutaneous layer at a 45-degree angle to the skin. Aspirate for blood, if none present, inject medication/immunization.

#### 2. Intramuscular (IM) Injections

- a. **Indications:** Immunizations and other medications.
- b. **Complications:** Bleeding, infection, allergic reaction, nerve injury.
- c. **Cautions**
  - (1) Avoid IM injections in a child with a bleeding disorder or thrombocytopenia.
  - (2) Maximum volume to be injected is 0.5 mL in a small infant, 1 mL in an older infant, 2 mL in a school-aged child, and 3 mL in an adolescent.

**d. Procedure**

- (1) Locate injection site: Anterolateral upper thigh in smaller child or outer aspect of upper arm (deltoid) in older one. The dorsal gluteal region is less commonly used because of risk for nerve or vascular injury. To find the ventral gluteal site, form a triangle by placing your index finger on the anterior iliac spine and your middle finger on the most superior aspect of the iliac crest. The injection should occur in the middle of the triangle formed by the two fingers and the iliac crest.
- (2) Cleanse skin with alcohol.
- (3) Pinch muscle with free hand, and insert 1-inch, 23- or 25-gauge needle until hub is flush with skin surface. For deltoid and ventral gluteal muscles, needle should be perpendicular to skin. For anterolateral thigh, needle should be at a 45-degree angle to the long axis of the thigh. Aspirate for blood; if none present, inject medication.

**B. Basic Laceration Repair<sup>2</sup>**

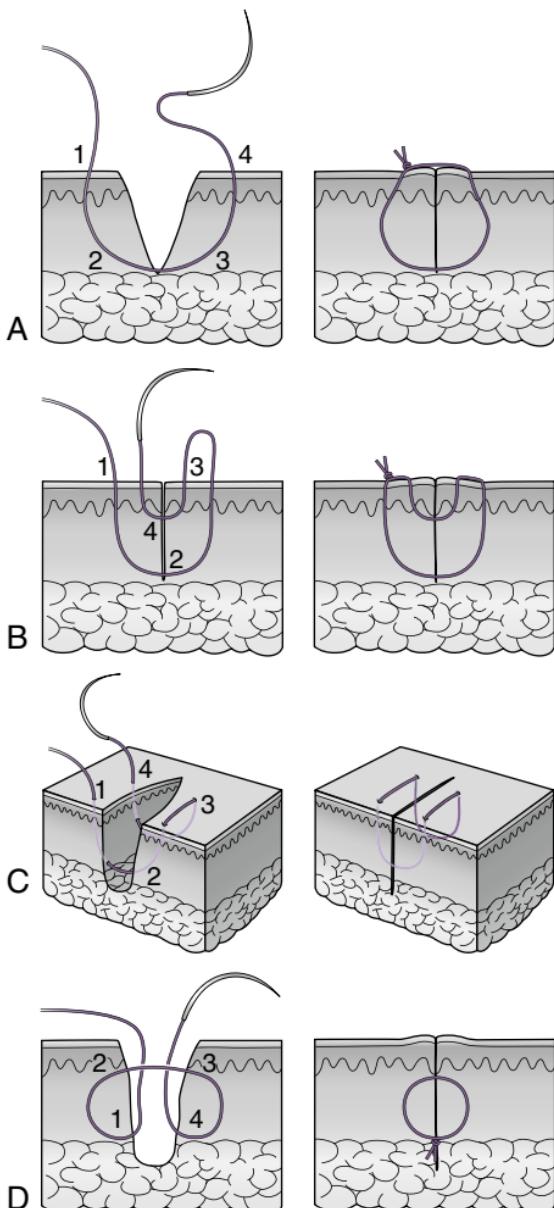
1. **Wound Irrigation<sup>21,22</sup>:** Numerous studies, including a large Cochrane review, conclude that there is no difference in the infection rates of wounds irrigated with either tap water or sterile NS. The volume of irrigation depends on the location and size of the wound; 100 mL/1 cm of laceration is a good approximation for relatively uncontaminated wounds.

**2. Suturing:****a. Basic Suturing Technique (Fig. 4.9):**

- (1) Simple interrupted: Basic closure of most uncomplicated wounds.
- (2) Horizontal mattress: Provides eversion of wound edges.
- (3) Vertical mattress: For added strength in areas of thick skin or areas of skin movement; provides eversion of wound edges.
- (4) Running intradermal: For cosmetic closures.
- (5) Deep dermal: For bringing together deeper portions of wounds with dissolving sutures to allow improved approximation and closure of superficial surfaces.

**b. Procedure:**

- (1) See **Tables 4.1–4.3** for sutures material, size, and time for removal.<sup>23</sup>
- (2) **NOTE:** Lacerations of the face, lips, hands, genitalia, mouth, or periorbital area may require consultation with a specialist. Ideally, lacerations at increased risk for infection (areas with poor blood supply, contaminated, or crush injury) should be sutured within 6 hours of injury. Clean wounds in cosmetically important areas may be closed up to 24 hours after injury in the absence of significant contamination or devitalization. In general, bite wounds should not be sutured except in areas

**FIGURE 4.9**

A–D, Suture techniques. (A) Simple interrupted. (B) Vertical mattress. (C) Horizontal mattress. (D) Deep dermal. (*Modified from Srivastava D, Taylor RS. Suturing technique and other closure materials. In: Robinson JK, Hanke CW, Siegel DM, et al., eds. Surgery of the Skin. 3rd ed. Elsevier: Philadelphia, PA; 2015:193–213.*)

of high cosmetic importance (face) or if significant gaping is present. These wounds can be closed loosely to aid in healing by secondary intention. The longer sutures are left in place, the greater potential for scarring and infection. Sutures in

**TABLE 4.1****GUIDELINES FOR SUTURE MATERIAL, SIZE, AND REMOVAL<sup>23</sup>**

Body Region	Nonabsorbable	Absorbable	Duration (Days)
Scalp	5–0 or 4–0	4–0	5–7
Face	6–0	5–0	3–5
Eyelid	7–0 or 6–0	—	3–5
Eyebrow	6–0 or 5–0	5–0	3–5
Trunk	5–0 or 4–0	3–0	5–7
Extremities	5–0 or 4–0	4–0	7–10
Joint surface	4–0	—	10–14
Hand	5–0	5–0	7
Foot sole	4–0 or 3–0	4–0	7–10

**TABLE 4.2****CHARACTERISTICS OF COMMON ABSORBABLE SUTURES<sup>23</sup>**

Material	Type	Tensile Strength	Absorption Time (Weeks)	Uses
Vicryl	Braided	75% at 14 days	8–10	Subcutaneous closure, vessel ligature
		50% at 21 days		
		5% at 30 days		
Vicryl rapide	Braided	50% at 5 days 0 at 14 days	6	Mucosa, dermis
Surgical gut plain	Twisted	Poor at 7–10 days	6–8	Subcutaneous closure
Surgical gut chromic	Twisted	Poor at 21–23 days	8–10	Subcutaneous closure
Monocryl	Monofilament	60%–70% at 7 days 30%–40% at 14 days	13–17	Subcuticular

**TABLE 4.3****CHARACTERISTICS OF COMMON NONABSORBABLE SUTURES<sup>23</sup>**

Material	Type	Tensile Strength
<b>NYLON (USED FOR SKIN CLOSURE)</b>		
Ethilon	Monofilament	20% per year
Dermalon	Monofilament	20% per year
Surgilon	Braided	Good
Nurolon	Braided	Good
<b>POLYPROPYLENE (USED FOR SCALP, EYEBROWS)</b>		
Prolene	Monofilament	Permanent
Surgilene	Monofilament	Permanent

cosmetically sensitive areas should be removed as soon as possible. Sutures in high tension areas (e.g., extensor surfaces) should stay in longer.

- (3) Prepare child for procedure with appropriate sedation, analgesia, and restraint. Utilize Child Life or age-appropriate distraction.

- (4) Anesthetize the wound with topical anesthetic or with lidocaine mixed with bicarbonate (with or without epinephrine) by injecting the anesthetic into the subcutaneous tissues (see [Chapter 6](#)).
- (5) Forcefully irrigate the wound as per above. This is the most important step in preventing infection.
- (6) Prepare and drape the patient for a sterile procedure.
- (7) Débride the wound when indicated. Probe for foreign bodies as indicated. Consider obtaining a radiograph if a radiopaque foreign body is suspected.
- (8) Select suture type for percutaneous closure (see [Tables 4.1–4.3](#)).
- (9) Match layers of injured tissues. Carefully match the depth of the bite taken on each side of the wound when suturing. Take equal bites from both wound edges. Apply slight thumb pressure on the wound edge as the needle is entering the opposite side. Pull the sutures to approximate wound edges but not too tightly, to avoid tissue necrosis. In delicate areas, sutures should be approximately 2 mm apart and 2 mm from the wound edge. Larger bites are acceptable where cosmesis is less important.<sup>2</sup>
- (10) When suturing is complete, apply topical antibiotic and sterile dressing. If laceration is in proximity of a joint, splinting of the affected area to limit mobility often speeds healing and prevents wound dehiscence.
- (11) Check wounds at 48 to 72 hours in cases where wounds are of questionable viability, if wound was packed, or for patients prescribed prophylactic antibiotics. Change dressing at checkup.
- (12) For hand lacerations, close skin only; do not use subcutaneous stitches. Elevate and immobilize the hand. Consider consulting a hand or plastics specialist.
- (13) Consider the need for tetanus prophylaxis (see [Chapter 16](#)).

c. A video on [basic laceration repair](#) is available on the *New England Journal of Medicine's* website

### 3. Skin Staples

#### a. Indications:

- (1) Best for scalp, trunk, extremity lacerations.
- (2) More rapid application than sutures, but can be more painful to remove.
- (3) Lower rates of wound infection.

#### b. Contraindications:

- (1) Not for areas that require meticulous cosmesis.
- (2) Avoid in patients who require magnetic resonance imaging (MRI) or CT.

#### c. Procedure:

- (1) Apply topical anesthetic, as above. Injection of lidocaine is not routinely used when using staples.
- (2) Clean and irrigate wound, as with suturing.

- (3) Appose wound edges, press stapler firmly against skin at center of apposed edges, and staple.
- (4) Apply antibiotic ointment and sterile bandage.
- (5) Left in place for the same length of time as sutures (see [Table 4.1](#)).
- (6) To remove, use dedicated staple remover.

#### 4. **Tissue Adhesives**<sup>24</sup>

##### a. **Indications:**

- (1) For use with superficial lacerations with clean edges.
- (2) Excellent cosmetic results, ease of application, and reduced patient anxiety.
- (3) Lower rates of wound infection.

4

##### b. **Contraindications:**

- (1) Not for use in areas under large amounts of tension (e.g., hands, joints).
- (2) Use caution with areas near the eye or over areas with hair such as the eyebrow.

##### c. **Procedure:**

- (1) Use pressure to achieve hemostasis and clean the wound as explained previously.
- (2) Hold together wound edges.
- (3) Apply adhesive dropwise along the wound surface, avoiding applying adhesive to the inside of the wound. Hold in place for 20 to 30 seconds.
- (4) If the wound is misaligned, remove the adhesive with forceps and reapply. Petroleum jelly or similar substance can aid in removal of skin adhesive.
- (5) Adhesive will slough off after 7 to 10 days.
- (6) Antibiotic ointments or other creams/lotions should not be applied to the adhesive, as this can cause premature loosening of the glue and subsequent wound dehiscence.

#### C. **Incision and Drainage (I&D) of Abscesses**<sup>2</sup>

##### 1. **Indications:** Diagnostic and therapeutic drainage of soft tissue abscess.

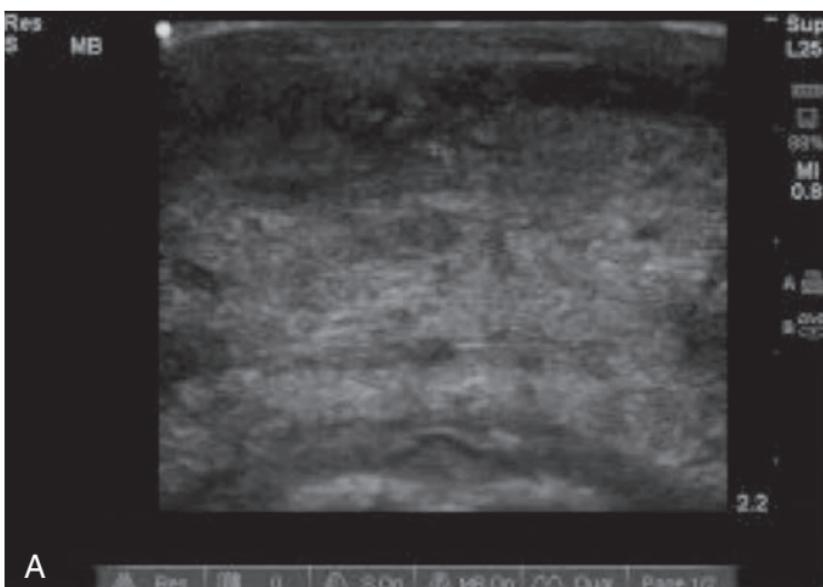
##### 2. **Complications:** Inadequate abscess drainage, local tissue injury, pain, scar formation, and, in rare cases, fistula formation. Consider specialized surgical evaluation for abscesses in cosmetically or anatomically sensitive areas such as the face, breast, or the anogenital region.

##### 3. **Ultrasound Identification:** Ultrasound imaging can be used to differentiate cellulitis from abscess.

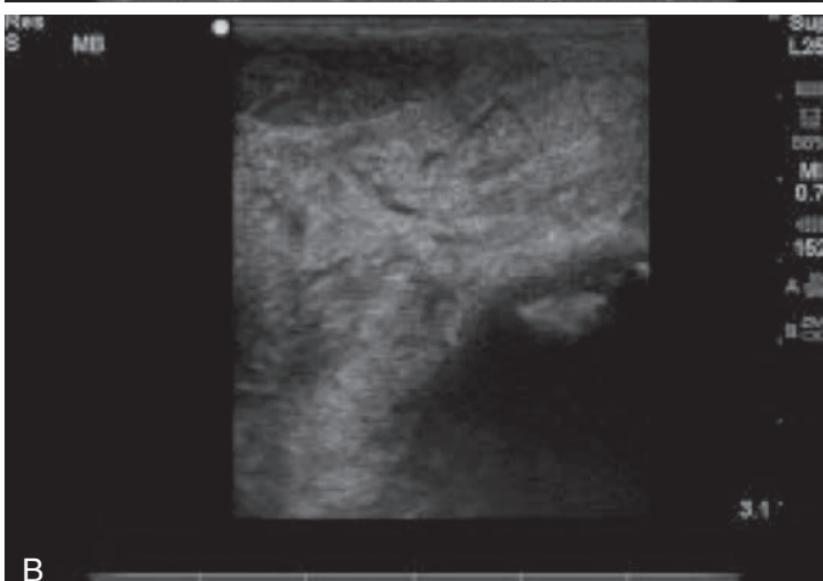
- a. Use a linear probe, place the probe over the area of interest, and scan it systematically such that the entire area of interest is examined.
- b. Cellulitis characteristics on ultrasound:
  - (1) Increased edema; tissue may appear slightly darker, and will have distorted, indistinct margins.
  - (2) Areas may have a “cobblestone” appearance caused by edema ([Fig. 4.10](#)).

c. Abscess characteristics on ultrasound:

- (1) Dark fluid collection distinct from surrounding tissue (see Fig. 4.10).
- (2) Often round or oval in shape.
- (3) Doppler can help to distinguish between lymph nodes and fluid collections.



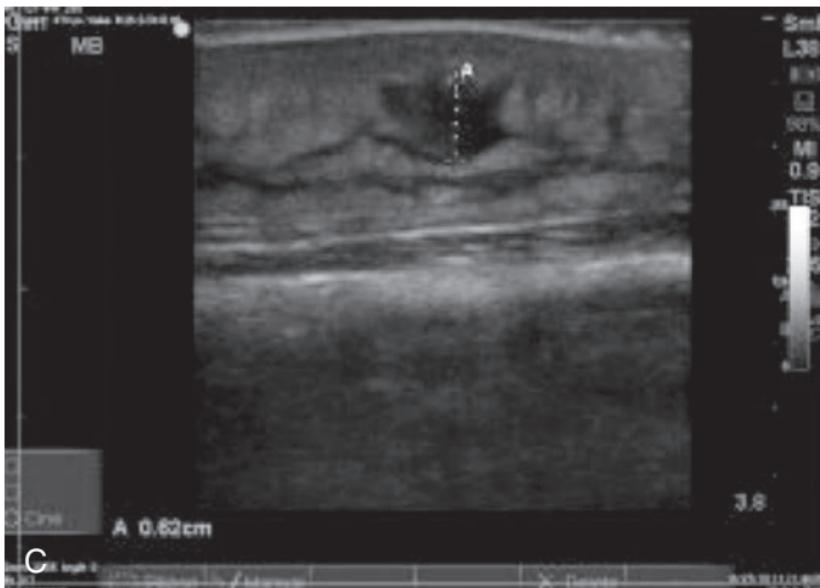
A



B

**FIGURE 4.10**

Ultrasound characteristics of soft tissue cellulitis and abscess. (A) Cellulitis characterized by bright (hyperechoic) tissue due to edema and inflammation in the tissue. (B) This image demonstrates the classic "cobblestone" appearance which is a later ultrasound finding in cellulitis.



**FIGURE 4.10, cont'd**

(C) A black (anechoic) rounded structure is noted in the soft tissue, which is characteristic of a soft tissue abscess. Some abscesses may appear dark gray depending on the characteristics of the fluid within the abscess. (*From Leeson K, Leeson B. Pediatric ultrasound: applications in the emergency department. Emerg Med Clin North Am. 2013;31(3):809–829.*)

#### 4. Procedure:

- Consider procedural sedation based upon the child's expected tolerance of the procedure and the location/size/complexity of the abscess.
- Apply topical anesthetic cream to the abscess to numb superficial epidermis (see [Chapter 6](#)).
- Prepare the overlying skin in a sterile fashion, and once cleaned, numb the area using 1% lidocaine and a small gauge needle, performing first a circumferential field block of the abscess area followed by direct injection to the planned incision site.
- Incise the skin over the abscess down to the superficial fascia using a scalpel blade, cutting parallel to the natural crease of the skin, if present.
- Using a hemostat, bluntly widen and undermine the incision to break up any septations or loculated fluid collections. Vigorously irrigate the wound using sterile saline to improve removal of purulent material.
- If desired, introduce a sterile packing strip into the wound using a hemostat, making sure to fill in an outside-to-inside pattern without overfilling.

- g. Leave a 2- to 3-cm tail outside the wound to facilitate removal and cover the wound with an absorbent dressing. Packing material should be removed in 1 to 2 days with a minimum of daily dressing changes until healed.
  - h. Consider starting antibiotics that cover staphylococcus and streptococcal species per local guidelines and resistance patterns.
5. A video on **I & D of Abscesses** is available on the *New England Journal of Medicine's* website.

#### D. Soft Tissue Aspiration<sup>25</sup>

1. **Indications:** Cellulitis that is unresponsive to initial standard therapy, recurrent cellulitis or abscesses, immunocompromised patients in whom organism recovery is necessary and may affect antimicrobial therapy.
2. **Complications:** Pain, infection, bleeding.
3. **Procedure:**
  - a. Select site to aspirate at the point of maximal inflammation (more likely to increase recovery of causative agent than leading edge of erythema or center).
  - b. Cleanse area in a sterile fashion.
  - c. Local anesthesia with 1% lidocaine is optional.
  - d. Fill tuberculin syringe with 0.1 to 0.2 mL of *nonbacteriostatic* sterile saline, and attach to needle.
  - e. Using 18- or 20-gauge needle (22-gauge for facial cellulitis), advance to appropriate depth, inject saline, aspirate fluid, and apply negative pressure while withdrawing needle.
  - f. Send fluid from aspiration for Gram stain and cultures. If no fluid is obtained, needle can be streaked on agar plate. Consider acid-fast bacillus (AFB) and fungal stains in immunocompromised patients.

#### E. Tuberculin Skin Test Placement<sup>26</sup>

1. **Indications:** Concern for exposure to tuberculosis.
2. **Contraindications:** History of severe reactions to prior placements (e.g., necrosis, anaphylactic shock, ulcerations). **Note** that there is no contraindication for any other individuals including infants, children, pregnant women, or persons who have been vaccinated with bacille Calmette-Guérin (BCG). Note that although a tuberculin skin test (TST) may be placed on the same day as a receiving a live vaccine, a TST must otherwise be placed 4 to 6 weeks after administration of a live vaccine (if not placed that same day).
3. **Complications:** Soreness, necrosis.
4. **Procedure:** Inject 0.1 mL of tuberculin purified protein derivative (PPD) with a tuberculin syringe (bevel up) into the forearm at a 5- to 15-degree angle. The bevel should be visible just below the skin surface. The injection should produce a pale elevation of the skin 6 to 10 mm in diameter.

5. **Follow-Up:** A TST should be read between 48 and 72 hours after administration. The reaction is measured across the forearm (perpendicular to the long axis) in millimeters of induration (palpable, raised, hardened area or swelling). Do not measure erythema.

6. **Interpretation:** see Chapter 17.

#### F. Tick Removal<sup>27</sup>

1. **Indications:** Visualization of tick. Urgent removal is essential, as the risk of Lyme disease transmission significantly increases after 24 hours of attachment with the risk highest at 48 to 72 hours.

2. **Complications:** Retention of tick fragments (particularly mouthparts), infection, granuloma formation.

3. **Procedure:** Only the use of blunt, medium-tipped, angled forceps or protected fingers have been shown to result in effective removal of ticks.

a. Use blunt forceps to grasp the tick at the skin surface. Lift up firmly, applying steady pressure and without a twisting motion. Take care to not squeeze the body of the tick, because its fluid may leak infectious material.

b. Apply antiseptic solution to the attachment site and provide patients with signs and symptoms of both local and systemic illness.

#### REFERENCES

A complete list of references can be found online at [www.expertconsult.com](http://www.expertconsult.com).

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## XI. ONLINE CONTENT

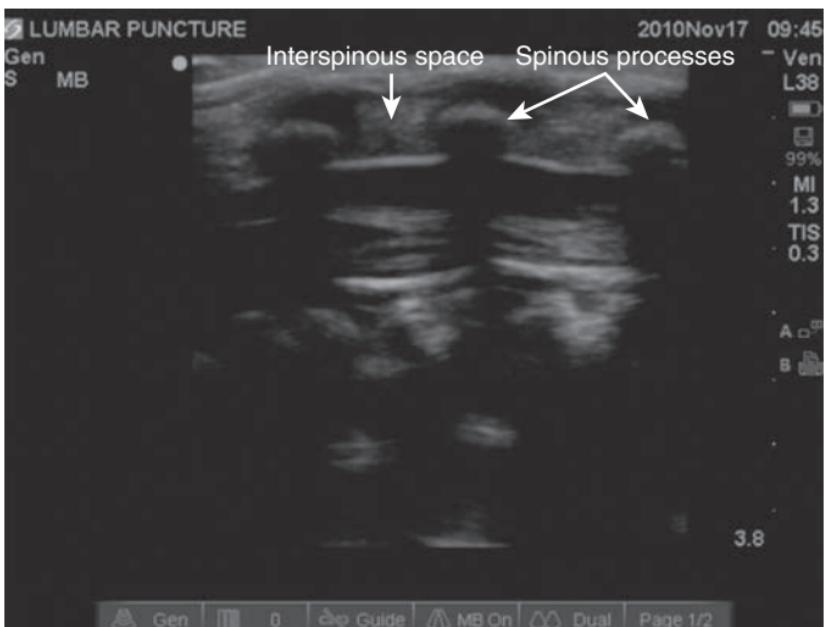
### A. Ultrasound-Guided Lumbar Puncture

1. Use the linear probe. Before preparing the patient, obtain a transverse view of the spine perpendicular to its axis. In the transverse view, identify the anatomic midline by locating the spinous process. The periosteum of the spinous process will appear as a hyperechoic, rounded structure with dark, posterior shadowing. Center the spinous process in the middle of the probe and mark a line in a cephalad-caudad direction on the patient's back to identify the midline (Fig. EC 4.A).
2. Rotate the probe 90 degrees to obtain a longitudinal view (probe parallel to the spine). Identify the vertebral bodies and an intervertebral space above or below L4. Mark a line on either side of the skin correlating with the space (Fig. EC 4.B).
3. The intersection of the marks identifies the area to be punctured. The crosshairs formed by the marks should leave the actual insertion site clean (Fig. EC 4.C).
4. The procedure should progress with no further movement of the patient. Preparation and draping should proceed from this point toward completion of the procedure.



**FIGURE EC 4.A**

Transverse ultrasound view of the lumbar spine. The spinous process is labeled in this ultrasound image of the lumbar spine, marking the anatomic midline for a lumbar puncture. A marking line should be drawn in the cephalad-caudad direction on the skin over the spinous processes. (From Marin J. Novel applications in pediatric emergency ultrasound. Clin Pediatr Emerg Med. 2011;12[1]:53–64.)

**FIGURE EC 4.B**

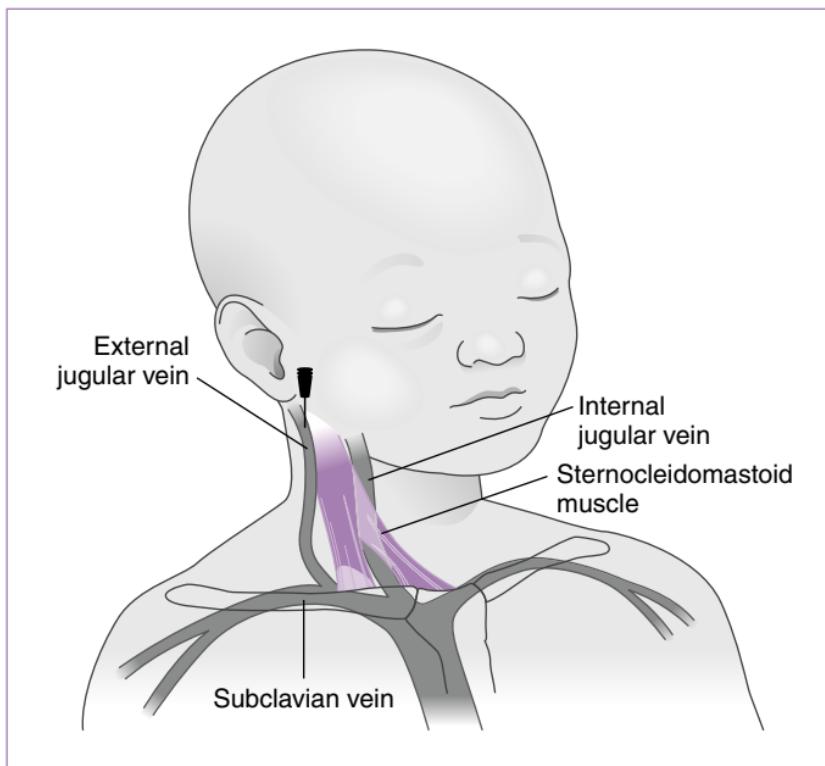
Longitudinal ultrasound view of the spine. The spinous processes are visualized as hyperechoic (bright) lines with posterior shadowing. In between the rounded spinous process is the interspinous space, which should be marked with a line for the procedure. (From Marin J. Novel applications in pediatric emergency ultrasound. Clin Pediatr Emerg Med. 2011;12[1]:53–64.)

**FIGURE EC 4.C**

Lumbar area marked for lumbar puncture. The lines from ultrasound markings should make a cross as seen in this image. Ideally there will be an area free of marking in the center where the actual puncture site will be. (From Strony R. Ultrasound-assisted lumbar puncture in obese patients. Crit Care Clin. 2010;26[4]:661–664.)

**B. External Jugular Puncture and Catheterization<sup>2</sup>**

1. **Indications:** Blood sampling in patients with inadequate peripheral vascular access or during resuscitation.
2. **Complications:** Infection, bleeding, pneumothorax.
3. **Procedure:** (Fig. EC 4.D)
  - a. Restrain patient securely with head turned 45 degrees to the contralateral side of cannulation. Position with towel roll under shoulders or with head over side of bed to extend neck and accentuate the posterior margin of the sternocleidomastoid muscle on the side of venipuncture. Place patient in the 15- to 20-degree Trendelenburg position.
  - b. Prepare area in a sterile fashion.
  - c. The external jugular vein will distend if its most proximal segment is occluded or if the child cries. The vein runs from the angle of the mandible to the posterior border of the lower third of the sternocleidomastoid muscle.

**FIGURE EC 4.D**

External jugular catheterization. (From Dieckmann R, Fiser D, Selbst S. Illustrated Textbook of Pediatric Emergency and Critical Care Procedures. St. Louis, MO: Mosby; 1997.)

- d. With continuous negative suction on the syringe, insert the needle at approximately a 30-degree angle to the skin. Continue as with any peripheral venipuncture.
- e. Apply a sterile dressing, and put pressure on the puncture site for 5 minutes.
- f. Enter the vein at the point where it crosses the sternocleidomastoid muscle.
- g. Proceed with peripheral catheter placement as described previously.

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# Chapter 5

## Adolescent Medicine

*Christine Krueger, MD and Harita Shah, MD*

 See additional content on Expert Consult

### I. ADOLESCENT HEALTH MAINTENANCE

#### A. Confidentiality

Begin integrating one-on-one time with the provider and patient into adolescent visits as early as age 11 to provide teens with regular opportunities to discuss concerns and sensitive topics in an open manner.<sup>1</sup> Adolescents are concerned about the confidentiality of their interactions with healthcare providers.<sup>2,3</sup> Providers should be aware of barriers to confidentiality related to consent laws and billing/explanation of benefits by insurance companies.<sup>4</sup>

1. **Consent laws:** All states and the District of Columbia allow minors to consent to sexually transmitted infection (STI) services (diagnosis and treatment), although some states have a minimum age to consent. Laws surrounding consent to HIV testing and treatment, contraception, abortion, and other healthcare services vary by state. Current information on consent laws by state can be found at the Guttmacher Institute's website (<https://www.guttmacher.org/state-policy/explore/overview-minors-consent-law>).<sup>5</sup>
2. **Breach of confidentiality:** Confidentiality must be breached if the adolescent is at risk of harming themselves or others (e.g., suicidal or homicidal ideation). Cases of child abuse or neglect must be reported to child protective services. The definition of statutory rape and reporting laws vary by state, with minimum age to consent to sexual activity ranging from 16 to 18 years old. Current information on reporting laws by state can be found at the Rape, Abuse & Incest National Network's website (<https://rainn.org/public-policy-action>).<sup>6</sup>

#### B. History Elements Unique to the Adolescent Patient

1. **Psychosocial development**<sup>7</sup>: Progression through adolescence is characterized by cognitive, psychosocial, and emotional developments, which help adolescents to establish their identity and autonomy. See **Table EC 5.A** for detailed psychosocial development by age.
2. **HEADSS assessment**<sup>8-10</sup>: A screening tool for psychosocial factors, which impact adolescent mental, physical, and sexual health (**Box 5.1**).
3. **Screening, brief intervention, and referral to treatment (SBIRT)** for substance use<sup>11</sup>:
  - a. **Screening:** If adolescent has used any alcohol, marijuana, or other drugs in the past 12 months, administer CRAFFT questionnaire (**Box 5.2**). If not, administer only the "Car" question (Have you ever ridden in a car with a driver who had used alcohol or drugs?).

**BOX 5.1****HE<sup>2</sup>ADS<sup>3</sup> (MODIFIED HEADSS) ASSESSMENT<sup>8</sup>**

- (H)**OME: Household composition, family dynamics and relationships, living and sleeping arrangements, recent changes, any periods of homelessness, running away from home
- (E)**DUCATION/Employment, **(E)**ATING: School performance, attendance, suspensions; attitude toward school; favorite, most difficult, best subjects; special educational needs; goals for the future; after-school job or other work history; body image and dieting
- (A)**CТИVITIES: Friendships with same or opposite sex, ages of friends, best friend, dating, recreational activities, physical activity, sports participation, hobbies, and interests
- (D)**RUGS: Personal use of tobacco, alcohol, illicit drugs, anabolic steroids; peer substance use; family substance use and attitudes; if personal use, determine frequency, quantity, binge, injury with use; consider use of **CRAFFT** questionnaire (Box 5.2)
- (S)**EXUALITY: Sexual orientation, gender identity, and relationship(s) should be explored with open-ended questions. If the adolescent is sexually active, discuss age of first sexual act, number of lifetime and current partners, ages of partners, knowledge of contraception and sexually transmitted infection/human immunodeficiency virus (STI/HIV) prevention, reproductive life plan, prior testing for STI/HIV, prior pregnancies and/or abortions, and history of nonconsensual intimate physical contact or sex. See Box EC 5.A for the “Five Ps” of the sexual history
- (S)**UICIDE/DEPRESSION: Feelings about self, both positive and negative; history of depression or other mental health problems; sleep problems (difficulty getting to sleep, early waking); changes in appetite or weight; anhedonia; irritability; anxiety; current or prior suicidal thoughts or attempts; other self-harming or injurious behavior; screen for depression using the Patient Health Questionnaire (PHQ-2)
- (S)**AFETY: Feeling unsafe at home, at school, or in the community; bullying; guns in the home; weapon carrying, what kinds of weapons; fighting; arrests; gang membership; seatbelt use

**BOX 5.2****CRAFFT QUESTIONNAIRE<sup>10</sup>**

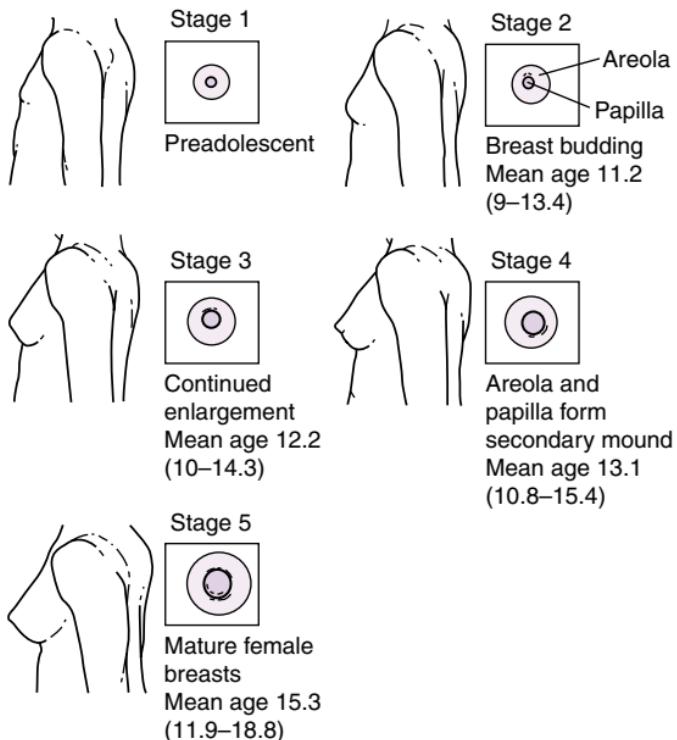
- C**—Have you ever ridden in a **CAR** driven by someone (or yourself) who was “high” or had been using alcohol or drugs?
- R**—Do you ever use alcohol or drugs to **RELAX**, feel better about yourself, or fit in?
- A**—Do you ever use alcohol/drugs while you are **ALONE**?
- F**—Do your family or **FRIENDS** ever tell you that you should cut down on your drinking or drug use?
- F**—Do you ever **FORGET** things you did while using alcohol or drugs?
- T**—Have you gotten into **TROUBLE** while you were using alcohol or drugs?
- NOTE:** Answering yes to two or more questions is a positive screen

- b. **Brief Intervention:** Stratify risk based on responses to screening questions.
- (1) Low risk (abstinent): Reinforce decisions with praise and provide anticipatory guidance regarding riding in a car with a driver under the influence.
  - (2) Yes to "Car" question: Counsel and encourage safety plan.
  - (3) Moderate risk (CRAFFT negative): Advise cessation of substance use, educate regarding health risks of continued use, and praise personal attributes.
  - (4) High risk (CRAFFT  $\geq 2$ ): Conduct in-depth assessment using motivational enhancement techniques, conduct brief negotiated interview, or refer as appropriate.
- c. **Referral to Treatment:** Further evaluation by a specialist in mental health/addiction can guide referral to an appropriate level of care.
4. **Social media**<sup>12</sup>: Explore how social media is used and for what quantity of time.
- a. Benefits: Communication and engagement.
  - b. Risks of excessive/inappropriate use: Impaired sleep, attention, and learning; obesity; depression; viewing of unsuitable content; decrease in caregiver-child interactions; compromised privacy; meeting high-risk sexual partners or sexual predators, sexting, and cyberbullying.
    - (1) Guidance for teens and families: <https://www.brightfutures.org/development/adolescence/social-media.html>
    - (2) AAP Family Media Use Plan: [www.healthychildren.org/MediaUsePlan](http://www.healthychildren.org/MediaUsePlan)
5. **Menstrual history:** Age of menarche, last menstrual period (LMP), frequency/regularity and duration of menstrual cycle, reproductive life plan, condom use, and contraceptive use.
- C. Physical Examination Elements Unique to the Adolescent**<sup>13,14</sup>
1. **Dentition and gums:** Caries, enamel defects from tobacco use, and enamel erosion from induced vomiting.
  2. **Skin:** Acne (see Chapter 8 for treatment guidelines), atypical nevi, acanthosis nigricans, rashes, evidence of cutting, piercings, and tattoos.
  3. **Thyroid:** Size, nodules.
  4. **Breasts:** Sexual maturity rating for females, masses (most commonly fibrocystic changes and fibroadenomas in females, or gynecomastia in males), breast asymmetry (common occurrence in adolescence; more pronounced between Tanner pubertal stages 2 and 4).
    - a. Normal female breast development: See Fig. 5.1.
    - b. Physiologic gynecomastia in males:
      - (1) Epidemiology: Generally, occurs in middle to late stages of puberty (usually peaks in Tanner pubertal stage 3); occurs in 50% of boys (50% unilateral, 50% bilateral).
      - (2) Etiology: Breast growth stimulated by estradiol.
      - (3) Clinical course: Regression usually occurs over a 2-year period.

**TABLE EC 5.A****PSYCHOSOCIAL DEVELOPMENT OF ADOLESCENTS**

<b>Task</b>	<b>Early Adolescence (10–13 years)</b>	<b>Middle Adolescence (14–16 years)</b>	<b>Late Adolescence (&gt;17 years)</b>
Independence	Less interest in parental activities Wide mood swings	Peak of parental conflicts	Reacceptance of parental advice and values
Body image	Preoccupation with self and pubertal changes Uncertainty about appearance	General acceptance of body Concern over making body more attractive	Acceptance of pubertal changes
Peers	Intense relationships with same-sex friend(s)	Peak of peer involvement Conformity with peer values Increased sexual activity and experimentation	Peer group less important More time spent in sharing intimate relationships
Identity	Increased cognition Increased fantasy world Increasing sexual attractions Idealistic vocational goals Increased need for privacy Lack of impulse control	Increased scope of feelings Increased intellectual ability Feeling of omnipotence Risk-taking behavior Emerging sexual identity	Practical, realistic vocational goals Refinement of moral, religious, and sexual values Ability to compromise and to set limits Sexual identity becomes more secure

From Joffe A. Introduction to adolescent medicine. In: McMillan JA, DeAngelis CD, Feigan RD, et al., eds. *Oski's Pediatrics Principles and Practice*. 4th ed. Philadelphia: Lippincott Williams, & Wilkins; 2006.

**FIGURE 5.1**

Tanner stages of breast development in females. (Modified from Johnson TR, Moore WM. *Children Are Different: Developmental Physiology*. 2nd ed. Columbus, OH: Ross Laboratories; 1978. Mean age and range [2 standard deviations around mean] from Joffe A. Introduction to adolescent medicine. In: McMillan JA, DeAngelis CD, Feigin RD, et al., eds. *Osaki's Pediatrics: Principles and Practice*. 4th ed. Philadelphia: Lippincott Williams & Wilkins; 2006:549–550.)

- (4) Physical examination: Firm glandular tissue in a concentric mass beneath the areola/nipple is consistent with physiologic gynecomastia. A testicular examination should also be performed.
- (5) Differential diagnosis: Nonphysiologic gynecomastia. Common causes include medication or substance use, primary or secondary hypogonadism, cirrhosis, hyperthyroidism, tumors, and pseudogynecomastia (excess adipose tissue on exam).
- (6) Red flags: Symptom duration over 2 years, nipple discharge, skin changes, breast masses, and coincident testicular abnormalities.
- (7) Treatment: Often no treatment is necessary. Severe or non-regressing cases may warrant referral to pediatric surgeon, endocrinologist, or oncologist, depending on suspected etiology.

**TABLE 5.1****TANNER STAGES OF GENITAL DEVELOPMENT (MALE)**

Tanner Stage	Comment ( $\pm 2$ Standard Deviations Around Mean Age)
1	Pre-pubertal
2	Enlargement of scrotum and testes <sup>a</sup> ; skin of scrotum reddens and changes in texture; little or no enlargement of penis; mean age 11.4 years (9.5–13.8 years)
3	Enlargement of penis, first mainly in length; further growth of testes and scrotum; mean age 12.9 years (10.8–14.9 years)
4	Increased size of penis with growth in breadth and development of glans; further enlargement of testes and scrotum and increased darkening of scrotal skin; mean age 13.8 years (11.7–15.8 years)
5	Genitalia adult in size and shape; mean age 14.9 years (13–17.3 years)

<sup>a</sup>Testicular volume of greater than 4 mL or a long axis of greater than 2.5 cm is evidence that pubertal testicular growth has begun.

Data from Joffe A. Introduction to adolescent medicine. In: McMillan JA, DeAngelis CD, Feigin RD, et al, eds. *Osaki's Pediatrics: Principles and Practice*. 4th ed. Philadelphia: Lippincott Williams & Wilkins; 2006:546–557.

**TABLE 5.2****TANNER STAGES OF PUBIC HAIR**

Tanner Stage	Appearance
1	No hair
2	Sparse, downy hair at base of symphysis pubis
3	Sparse, coarse hair across symphysis pubis
4	Adult hair quality, fills in pubic triangle, no spread to thighs
5	Adult quality and distribution including spread to medial thighs

Data from Alario AJ, Birnkrant JD. Sexual maturation and tanner staging. *Practical Guide to the Care of the Pediatric Patient*. 2nd ed. St. Louis: Mosby; 2007:798–800.

5. **Genitalia:** For both male and female genital examinations, a chaperone should be present, an explanation should occur before the examination, and findings should be discussed.

a. **Male**<sup>15</sup>:

- (1) Normal male genital development: [Table 5.1](#).
- (2) Normal pubic hair development: [Table 5.2](#).
- (3) Assess for signs of STIs (rashes, warts, ulcers, erosions, discharge), inguinal hernias, masses, hydroceles, and varicoceles. If there are symptoms of proctitis with history of receptive anal intercourse (e.g., rectal pain, rectal bleeding, or tenesmus), a digital rectal examination should be performed.

b. **Female**<sup>16,17</sup>:

- (1) Normal pubic hair development: see [Table 5.2](#).
- (2) External examination: Assess for signs of STIs (rashes, warts, ulcers, erosions, discharge), discharge suggestive of candidiasis or bacterial vaginosis, and evidence of trauma.

- (3) Pelvic examination: Speculum exams are not routinely recommended for healthy asymptomatic women under 21 years of age. Indications for bimanual exam or speculum exam include: vaginal discharge with lower abdominal or pelvic pain (assess cervix for mucopurulent discharge, friability, large ectropion, foreign body, or cervical motion tenderness), menstrual disorders (amenorrhea, abnormal vaginal bleeding, or dysmenorrhea refractory to medical therapy), and Pap smear (see [Section I.D.6](#)).
- (4) For suspected or reported sexual abuse or rape, refer to a specialized center if not appropriately trained and equipped to document evidence of trauma and collect forensic specimens.

Note: See [Chapter 10](#) for information about precocious and delayed puberty.

#### D. Screening Laboratory Tests and Procedures

A lack of evidence has led to variability in guidelines for topics such as screening for dyslipidemia, iron-deficiency anemia, diabetes, and tuberculosis. The recommendations that follow are largely based on the AAP, CDC, U.S. Preventive Services Task Force (USPSTF), and the American College of Obstetricians and Gynecologists (ACOG).<sup>1,18–20</sup>

1. **Immunizations:** See [Chapter 16](#).
2. **Cholesterol screening:** All children should undergo cholesterol screening once between ages 9 and 11 years and once between ages 17 and 21 years.
3. **Diabetes screening:** Consider screening for type 2 diabetes with hemoglobin A1c, fasting plasma glucose, or oral glucose tolerance test in children who have a BMI greater than 85% for age and sex with other risk factors such as family history.<sup>21</sup>
4. Consider selective screening for tuberculosis, anemia, and vision and hearing abnormalities if patient screens positive on risk screening questions.
5. **STI screening:** See [Section II](#).
6. **Papanicolaou (Pap) smear cervical cancer screening:** Cytologic evaluation should be used. Human papilloma virus (HPV) testing is only recommended for routine screening above age 30. Recommended screening intervals and follow-up depend on age, medical history, and result of previous Pap smear, as presented in [Table 5.3](#).

## II. SEXUAL HEALTH

### A. Sexually Transmitted Infection Screening Guidelines by Sexual Behavior<sup>18,22,23</sup>

1. **All adolescents over age 13:** The CDC recommends universal screening for HIV (via HIV 1/2 antigen/antibody test) at least once using an opt-out approach, or more frequently based on risk factors.

**TABLE 5.3**  
**GUIDELINES FOR PAPANICOLAOU (PAP) SMEAR SCREENING AND FOLLOW-UP**

Immune Status	Recommended Timing of Pap Screening	Recommended Follow-Up Based on Pap Result
Immunocompetent	Age 21	<p>Normal</p> <p>Atypical squamous cells of undetermined significance (ASC-US)</p> <p>Low-grade squamous intraepithelial lesion (LSIL)</p> <p>High-grade squamous intraepithelial lesion (HSIL)</p> <p>Atypical squamous cells, cannot rule out HSIL (ASC-H)</p>
Immunosuppressed	Age 21	<p>Normal</p> <p>Abnormal</p> <p>Normal</p> <p>Abnormal</p>
HIV+	Within 1 year of HIV diagnosis, or if perinatally acquired, within 1 year of onset of sexual activity	<p>Repeat Pap smear every 3 years</p> <p>Repeat Pap smear in 1 year</p> <p>Repeat Pap smear in 1 year</p> <p>Gynecology referral for colposcopy</p> <p>Gynecology referral for colposcopy</p> <p>Repeat Pap smear every year</p> <p>Gynecology referral</p> <p>Repeat Pap smear every year. If three consecutive Pap smears are normal, space Pap interval to every 3 years.</p> <p>Gynecology referral</p>

Data from American College of Obstetricians and Gynecologists. Committee on Practice Bulletins—Gynecology. Practice Bulletin No. 168: Cervical Cancer Screening and Prevention. *Obstet Gynecol*. 2016;128(4):e111–e130.

**2. Heterosexual with one lifetime partner:**

- a. **Male:** Screen once with HIV 1/2 antigen/antibody test; gonorrhea and chlamydia urine nucleic acid amplification test (NAAT) in high prevalence clinical settings (e.g., adolescent clinics, correctional facilities, STI clinics). Repeat as indicated by sexual risk.
- b. **Female:** Screen once with HIV 1/2 antigen/antibody test; self- or provider-collected vaginal NAAT for gonorrhea and chlamydia (routine screening recommended in sexually active women under age 25). Repeat as indicated by sexual risk. Vaginal swab is the preferred method to screen for gonorrhea and chlamydia; self-collected specimens may have higher patient acceptability. Vaginal swabs are as sensitive and specific as cervical swabs, and both are more accurate than urine samples.<sup>24</sup>

**3. Heterosexual with risk factors** (new partner, multiple partners, partner with STI, intravenous drug use):

- a. **Male:** Annual HIV 1/2 antigen/antibody test, rapid plasma reagin (RPR), and gonorrhea and chlamydia urine NAAT.
- b. **Female:** Annual HIV 1/2 antigen/antibody test, RPR, and self- or provider-collected vaginal NAAT for gonorrhea, chlamydia, and trichomonas.
- c. In adolescents with a history of an STI, repeat testing is recommended 3 months after treatment given high risk of reinfection.

**4. Men who have sex with men (MSM):** Annual HIV 1/2 antigen/antibody test, RPR, and gonorrhea and chlamydia NAAT (test sites of sexual contact: pharynx, urethra, and/or rectum) are recommended. For MSM with multiple or anonymous partners, consider 3 to 6 month interval STI testing.**5. Women who have sex with women (WSW):** Equivalent STI screening as heterosexual women, guided by sexual practices (e.g., gonorrhea and chlamydia NAAT should be done at sites of sexual contact) and risk factors.**6. Pregnant women:** At the first visit, pregnant patients should be screened with HIV 1/2 antigen/antibody test, RPR, vaginal gonorrhea and chlamydia NAAT, and hepatitis B surface antigen test.**7. Transgender:** Given the diversity of transgender persons regarding patterns of sexual behavior, hormone use, and surgery, clinicians should assess STI risk based on the patient's sexual behaviors and current anatomy (the latter of which should guide method of NAAT testing, if indicated).**8. Concern for recent exposure to STI:**

- a. Fourth generation HIV 1/2 antigen/antibody tests detect acute infection within 10 to 14 days. If there is concern for acute or early HIV exposure, consider an HIV RNA nucleic acid test.<sup>25</sup>
- b. Screen with RPR, gonorrhea and chlamydia NAAT (method of testing as described above by sexual behavior), and vaginal trichomonas NAAT for women.
- c. Consider HSV PCR testing in individuals presenting for STI evaluation with genital lesion(s).

9. **Persons living with HIV:** Screen at least annually with RPR, gonorrhea and chlamydia NAAT (method of testing as described above by sexual behavior), and vaginal trichomonas NAAT for women. Screen more frequently if indicated by sexual risk behaviors.

## B. Sexually Transmitted Infection Evaluation and Management

### (Table 5.4)

1. **HIV:** See Chapter 17 for information on diagnosis and treatment of HIV, pre-exposure prophylaxis (PrEP), and postexposure prophylaxis (PEP). PrEP should be initiated in the primary care setting, when possible.
2. **Syphilis**
  - a. Etiology: *Treponema pallidum*
  - b. Early syphilis (within 1 year of initial infection)
    - (1) Primary syphilis (chancre): Firm, usually painless sore(s) or ulcer(s) develop at the site of initial infection (genital, rectal, or oral). Chancres typically develop within 3 weeks of infection and heal 3 to 6 weeks after development in the absence of treatment.
    - (2) Secondary syphilis: Within weeks to months after a chancre appears, patients may develop body rash involving palms and soles, mucocutaneous lesions, lymphadenopathy, constitutional symptoms, and/or early neurosyphilis (e.g., meningitis or ocular syphilis).
    - (3) Early latent syphilis: Asymptomatic stage.
  - c. Late syphilis (over 1 year after initial infection)
    - (1) Late latent syphilis: Asymptomatic stage.
    - (2) Tertiary syphilis: Organ involvement may progress to cardiovascular syphilis (e.g., aortitis), late neurosyphilis (e.g., tabes dorsalis or paresis), or gummatous syphilis.
  - d. Diagnosis: Testing algorithm varies by laboratory and typically includes a nontreponemal test (RPR or Venereal Disease Research Laboratory [VDRL] test) and a treponemal test (e.g., fluorescent treponemal antibody absorbed [FTA-ABS] test, enzyme immunoassay) for confirmation.
  - e. Treatment: See Table 5.4. Clinical and serologic evaluation should be performed 6 and 12 months after treatment to ensure a fourfold reduction in nontreponemal titers or seroreversion. Monitor for Jarisch-Herxheimer reaction (fever, headache, and myalgias) within 24 hours of treatment.
  - f. Partner treatment: Partners with sexual contact within 90 days of a patient's diagnosis should be treated empirically. Partners with sexual contact over 90 days prior to diagnosis should be evaluated for treatment based on CDC 2015 STD Treatment Guidelines: <http://www.cdc.gov/std/tg2015/>.
  - g. See Chapter 17 for information on neonatal syphilis and interpretation of maternal and neonatal syphilis testing.

**TABLE 5.4**  
**SEXUALLY TRANSMITTED AND GENITOURINARY INFECTIONS: GUIDELINES FOR MANAGEMENT<sup>a</sup>**

Infection	Clinical Diagnosis	Empiric Therapy <sup>a</sup>	Comments
Chlamydia Infections	Uncomplicated infection of the cervix, urethra, rectum, or pharynx	Azithromycin 1 g PO once <i>OR</i> doxycycline 100 mg PO BID for 7 days <i>Alt:</i> <i>Erythromycin OR fluoroquinolone</i>	Consider empirical treatment for gonorrhoea secondary to common coinfection Test of cure 3 weeks posttreatment in all pregnant patients
	<i>Chlamydia</i> infection in pregnancy	Azithromycin 1 g PO once <i>Alt:</i> <i>Amoxicillin OR erythromycin</i>	
	Lymphogranuloma venereum (LGV)	Doxycycline 100 mg PO BID for 21 days <i>Alt:</i> <i>Erythromycin</i>	
		Ceftriaxone 250 mg IM once <i>PLUS</i> azithromycin 1 g PO once <i>Alt for ceftriaxone:</i> <i>Cefixime OR gemifloxacin OR gentamicin</i>	Dual treatment is recommended for gonorrhoea secondary to organism resistance For MSM, replace doxycycline with a fluoroquinolone for 10 days
Gonorrhea Infections	Uncomplicated infection of the cervix, urethra, rectum, or pharynx	Ceftriaxone 250 mg IM once <i>PLUS</i> azithromycin 1.00 mg PO BID for 10 days <i>Alt for ceftriaxone:</i> <i>Cefixime OR gentamicin</i>	Can switch to oral therapy 24–48 h after clinical improvement. Total course: 7 days
	Epididymitis	Ceftriaxone 1 g IV/IM daily <i>PLUS</i> azithromycin 1 g PO once <i>Alt for ceftriaxone:</i> <i>Cefotaxime</i>	
	Disseminated gonococcal infections		
Pelvic Inflammatory Disease (PID)	PID warranting outpatient treatment	Ceftriaxone 250 mg IM once <i>PLUS</i> doxycycline 100 mg PO BID for 14 days ± metronidazole 500 mg PO BID for 14 days Regimen A for 14 days: (Cefotetan 2 g IV q12h <i>OR</i> cefotixin 2 g IV q6h) <i>PLUS</i> doxycycline 100 mg IV or PO q12h Regimen B for 14 days: Clindamycin 900 mg IV q8h <i>PLUS</i> gentamicin 2 mg/kg loading dose, then 1.5 mg/kg IV q8h maintenance (or 3–5 mg/kg IV single daily dosing)	Switch to oral therapy 24 h after clinical improvement to complete 14 days of treatment with doxycycline 100 mg PO BID or clindamycin 450 mg PO QD, respectively

Syphilis	Primary, secondary, or early latent syphilis (<1 year duration)	Benzathine penicillin G 50,000 U/kg (max 2.4 million units) IM <i>Alt: Doxycycline OR tetracycline</i>	Data is limited for penicillin alternatives. Pregnant women should be treated with penicillin G regimen appropriate for stage of syphilis
	Late latent syphilis (>1 year duration); tertiary syphilis	Benzathine penicillin G 50,000 U/kg (max 2.4 million units) IM weekly for 3 weeks <i>Alt: Doxycycline OR tetracycline</i>	
	Herpes (Genital, Nonneonatal)	Acyclovir or valacyclovir	See Formulary for treatment for initial infection and recurrence

*Alt.*, Alternative; *IM*, intramuscular; *IV*, intravenous; *MSM*, men who have sex with men; *PQ*, per os.

<sup>a</sup>For dosing for children aged ≤8 years or weighing less than 15 kg, or for dosing of alternative regimens, please refer to the CDC Treatment Guidelines, 2015: <http://www.cdc.gov/std/tg2015/>. Partner notification and treatment is recommended for most sexually transmitted infections. Patients treated for a sexually transmitted infection should refrain from all sexual activity for 7 days posttreatment.

**3. Chlamydia and gonorrhea<sup>18</sup>**

- a. Etiology: *Chlamydia trachomatis* and *Neisseria gonorrhoeae*
- b. Clinical manifestations: Urethritis, cervicitis, pharyngitis, proctitis, epididymitis, prostatitis. Other manifestations include:
  - (1) Lymphogranuloma venereum (LGV): Lymphoproliferative reaction caused by *C. trachomatis* serovars L1 to L3 that most frequently presents as proctitis and lymphadenopathy in patients who are MSM or HIV positive.
  - (2) Disseminated gonococcal infection: Bacteremic spread of *N. gonorrhoeae* results in septic arthritis or arthritis-dermatitis syndrome (polyarthralgia, tenosynovitis, and dermatitis). In addition to urogenital, rectal, and pharyngeal NAAT, workup should include blood, synovial, or CSF cultures, as applicable.
- c. Diagnosis: Site-specific NAAT, including urogenital (urine NAAT in males, urine or vaginal NAAT in females [see Section II.A.2.b]), pharyngeal, and rectal.
- d. Treatment: See **Table 5.4**.
- e. Partner treatment: Partners should be treated. For partners for whom providers are concerned about access to prompt clinical evaluation and treatment, expedited partner therapy may be an option depending on local and state laws.

**4. Pelvic inflammatory disease (PID):** Acute infection of the female upper genital tract.<sup>18</sup>

- a. Etiology: Often polymicrobial in nature, however *N. gonorrhoeae* and *C. trachomatis* are the most commonly identified pathogens, followed by *Mycoplasma genitalium*.
- b. Differential diagnosis: Endometriosis, tubo-ovarian abscess (TOA), ovarian cyst, ectopic pregnancy, acute surgical abdomen, inflammatory bowel disease (IBD), pyelonephritis, dysmenorrhea, septic/threatened abortion
- c. Workup: Pelvic and bimanual examination, gonorrhea/chlamydia and HIV testing, human chorionic gonadotropin (hCG), wet preparation, erythrocyte sedimentation rate (ESR), C-reactive protein (CRP), and urinalysis/urine culture if clinically indicated. Consider a complete blood cell count (CBC) with differential and pelvic ultrasound if the patient is ill-appearing, has an adnexal mass on bimanual examination, or is not improving after antibiotics.
- d. Minimum diagnostic criteria: Uterine, adnexal, or cervical motion tenderness without other identifiable causes. One or more of the following additional criteria enhances specificity: fever ( $>38.3^{\circ}\text{C}$ ), mucopurulent vaginal or cervical discharge, leukocytes on saline microscopy, increased ESR or CRP, laboratory documentation of chlamydial or gonorrhea infection.
- e. Treatment: See **Table 5.4**.
- f. Admission criteria: Cannot exclude acute surgical abdomen, presence of TOA, pregnancy, immunodeficiency, severe illness, inability

to tolerate or follow outpatient oral regimen, failure to respond to appropriate outpatient therapy, or follow-up cannot be ensured.

### 5. Trichomoniasis<sup>18</sup>

- a. Etiology: *Trichomonas vaginalis*
- b. Diagnosis and treatment: See [Table 5.5](#).
- c. Follow-up: Women treated for trichomoniasis should be retested 3 months after treatment due to high rates of reinfection.
- d. Partner treatment: Partners should be treated to prevent reinfection.

### 6. Mycoplasma genitalium<sup>18</sup>

- a. Etiology: *Mycoplasma genitalium*
- b. Clinical manifestations: Persistent urethritis, cervicitis, or PID despite appropriate treatment.
- c. Diagnosis: No FDA-approved diagnostic test; NAAT is available in some large medical centers and commercial laboratories.
- d. Treatment: Moxifloxacin has been used successfully; refer to latest CDC treatment guidelines.

### 7. Vaginal infections, genital ulcers, and warts

- a. Diagnostic features of vaginal infections (see [Table 5.5](#)) can assist in differentiating normal vaginal discharge from bacterial vaginosis, trichomoniasis, and yeast vaginitis.
- b. Diagnostic features of genital lesions, as well as management of warts and ulcers, are presented in [Table 5.6](#).

## C. Gender and Sexual Behavior

### 1. Terminology and definitions:

- a. **Sexual orientation**<sup>26,27</sup>: Sexual orientation relates to sexual attraction, identity, and behavior. It is not related to gender identity. It should be defined by the individual patient.
- b. **Gender identity**: An individual's self-awareness as male or female.
- c. **Gender expression**: The way an individual expresses their gender (e.g., clothing and speech); may differ from gender identity.
- d. **Sex assigned at birth**: Often based on phenotype (external genitals, gonads, and internal sex organs) and karyotype (XX, XY, XO, XXY, etc.); assigned at birth.
- e. **Transgender**: An individual whose gender identity differs from the sex assigned at birth.
- f. **Cisgender**: An individual whose gender identity is the same as the sex assigned at birth.
- g. **Gender nonbinary**: Gender expression by an individual that does not match masculine and feminine gender norms.
- h. **Gender dysphoria**<sup>28</sup>: Discomfort or distress caused by discrepancy between a person's gender identity and sex assigned at birth. DSM-V criteria recommends a diagnosis occur after 6 months of continuous incongruence. For prepubescent children, the desire to be of the other gender must be present and verbalized.

### 2. Special considerations in adolescents:

- a. Adolescents may engage in a variety of sexual activities (penile-vaginal, anal, or oral intercourse) that do not reflect their sexual orientation (e.g.,

**TABLE 5.5**  
**DIAGNOSTIC FEATURES AND MANAGEMENT OF VAGINAL INFECTIONS**

	No Infection/Physiologic Leukorrhea	Vulvovaginal Candidiasis	Trichomoniasis	Bacterial Vaginosis <sup>a</sup>
Etiology	—	<i>Candida albicans</i> and other yeasts	<i>Trichomonas vaginalis</i>	<i>Gardnerella vaginalis</i> , anaerobic bacteria, mycoplasma
Typical symptoms	None	Vulvar itching, irritation, ↑ discharge Scant to moderate	Malodorous frothy discharge, vulvar itching Profuse	Malodorous, ↑ discharge Moderate
Discharge Amount	Variable, usually scant	White	Yellow-green	Usually white or gray
Discharge Color	Clear or white	Clumped; adherent plaques	Homogenous	Homogenous, low viscosity
Discharge Consistency	Heterogeneous	Yes	Yes	No
Vulvar/vaginal inflammation	No	Usually <4.5	Usually >5.0	Usually >4.5
pH of vaginal fluid <sup>b</sup>	Usually <4.5	None	May be present	Present, positive "whiff-amine" test
Amine ("fishy") odor with 10% potassium hydroxide (KOH)	None	Leukocytes, epithelial cells, yeast, mycelia, or pseudomycelia in 40%–80% of cases	Leukocytes; matile trichomonads seen in 50%–70% of symptomatic patients, less often if asymptomatic	Clue cells, few leukocytes; <i>Lactobacillus</i> outnumbered by profuse mixed flora (nearly always including <i>G. vaginalis</i> plus anaerobes)
Microscopy <sup>c</sup>	Normal epithelial cells; <i>Lactobacillus</i> predominates		Metronidazole 2 g PO once OR Fluconazole 150 mg PO once OR intravaginal azole cream	Metronidazole 500 mg PO BID for 7 days OR metronidazole gel 0.75% 5 g intravaginally daily for 5 days OR clindamycin cream 2% 5 g intravaginally daily for 7 days
Usual treatment	None			

**NOTE:** Refer to Formulary for dosing information.

<sup>a</sup>Despite more sensitive and specific laboratory tests, cost and practicality make the Amsel criteria the best in-office method to diagnose Bacterial Vaginosis. To diagnose BV, at least 3 criteria must be present: (1) Homogenous, thin, gray/white discharge; (2) vaginal pH >4.5; (3) Positive whiff-amine test; (4) Clue cells on wet mount.

<sup>b</sup>pH determination is not useful if blood is present.

<sup>c</sup>To detect fungal elements, vaginal fluid is mixed with 10% KOH before microscopic examination; to examine for other features, fluid is mixed with saline.

From Workowski KA, Bolan GA. Centers for Disease Control and Prevention. Sexually transmitted diseases treatment guidelines. 2015. *MMWR Recomm Rep*. 2015;64(RR-3):1–137.

**TABLE 5.6** DIAGNOSTIC FEATURES AND MANAGEMENT OF GENITAL ULCERS AND WARTS

Infection	Clinical Presentation	Presumptive Diagnosis	Definitive Diagnosis	Treatment/Management of Sex Partners
Genital herpes	Grouped vesicles, painful shallow ulcers to mild clinical manifestation (redness, pain, excoriations); HSV-2 more common cause of genital lesions	Tzank preparation with multinucleated giant cells	HSV PCR	No known cure. Prompt initiation of therapy shortens duration of first episode. For severe recurrent disease, initiate therapy at start of prodrome or within 1 day. Transmission can occur during asymptomatic periods. See Formulary for dosing of acyclovir, famciclovir, or valacyclovir.
Chancroid	Etiology: <i>Haemophilus ducreyi</i> Painful genital ulcer; tender, suppurative inguinal adenopathy	No evidence of <i>Treponema pallidum</i> (syphilis) on dark-field microscopy or serologic testing; negative HSV	Use of special media (not widely available in United States); sensitivity <80%	Single dose: Azithromycin 1 g orally <i>Or</i> ceftriaxone 250 mg IM. Partners should be examined and treated, regardless of whether symptoms are present, or if they have had sex within 10 days preceding onset of patient's symptoms. Syphilis is a common co-pathogen with chancroid.
Primary syphilis	Indurated, well-defined, usually single painless ulcer or chancre; nontender inguinal adenopathy	Nontreponemal serologic test: VDRL, RPR, or STS	Treponemal serologic test: FTA-ABS, dark-field microscopy or direct fluorescent antibody tests of lesion exudates or tissue	Parenteral penicillin G (see Table 5.4 for preparation[s], dosage, and length of treatment). Treat presumptively for persons exposed within 3 months preceding the diagnosis of primary syphilis in a sex partner or who were exposed >90 days preceding the diagnosis and in whom serologic tests may not be immediately available or follow-up is uncertain.

*Continued*

**TABLE 5.6—CONT'D**

<b>DIAGNOSTIC FEATURES AND MANAGEMENT OF GENITAL ULCERS AND WARTS</b>				
Infection	Clinical Presentation	Presumptive Diagnosis	Definitive Diagnosis	Treatment/Management of Sex Partners
HPV infection (genital warts)	Soft, fleshy, papillary or sessile, painless growth(s) around anus, vulvovaginal area, penis, urethra, or perineum; no inguinal adenopathy	Typical clinical presentation	Papanicolaou smear revealing typical cytologic changes	<p>Treatment does not eradicate infection. Goal: Removal of exophytic warts. Exclude cervical dysplasia before treatment.</p> <ol style="list-style-type: none"> <li><b>Patient-administered therapies include:</b> Podofilox gel or imiquimod cream</li> <li><b>Clinician-applied therapies include:</b> Bichloroacetic or trichloroacetic acid, surgical removal, or cryotherapy with liquid nitrogen or cryoprobe.</li> </ol> <p>Podofilox, imiquimod, and podophyllin are contraindicated in pregnancy. Period of communicability unknown.</p>

**NOTE:** Chancroid, lymphogranuloma venereum (LGV), and granuloma inguinale should be considered in the differential diagnosis of genital ulcers if the clinical presentation is atypical and tests for herpes and syphilis are negative.

*EIA*, Enzyme immunoassay; *FTA-ABS*, fluorescent treponemal antibody absorbed; *HPV*, human papillomavirus; *HSV*, herpes simplex virus; *IM*, intramuscular; *RPR*, rapid plasma reagent; *STS*, serologic test for syphilis; *TP-PA*, T. pallidum passive particle agglutination assay; *VDRL*, Venereal Disease Research Laboratory.

Modified from Worobski KA, Bolan GA. Centers for Disease Control and Prevention. Sexually transmitted diseases treatment & guidelines. 2015. *MMWR Recomm Rep*. 2015;64(RR3):1–137.

heterosexual, homosexual, bisexual). Conversely, adolescents may self-identify with a particular sexual orientation but not be sexually active.

- b. Sexual identity emerges during adolescence. It is important to provide a safe environment for adolescents to discuss questions about their sexual identity and behavior, and ask questions about sexual activities regardless of sexual orientation.

### 3. Medical interventions for gender-dysphoric or transgender patients<sup>29-31</sup>:

- a. Prepubertal children: Parental support and education to create a safe environment for the child. Familial support of social transition for transgender children has been associated with better mental health outcomes.
- b. Pubertal suppression (reversible): GnRH analogue (e.g., Lupron and Supprelin) can be used to suppress endogenous hormones after onset of puberty.
- c. Gender-affirming hormone therapy (partially irreversible): Estradiol or testosterone therapy with gradual dose escalation initiated after a multidisciplinary team of medical and mental health providers has confirmed the persistence of gender dysphoria/gender incongruence and sufficient mental capacity to give informed consent (generally by the age of 16 years). Treatment can be considered as early as age 13.5 to 14 years.
- d. Gender-affirming surgery (including gonadectomy, hysterectomy, mastectomy, and genital surgery): Not recommended until age of majority.

## D. Contraception<sup>32,33</sup>

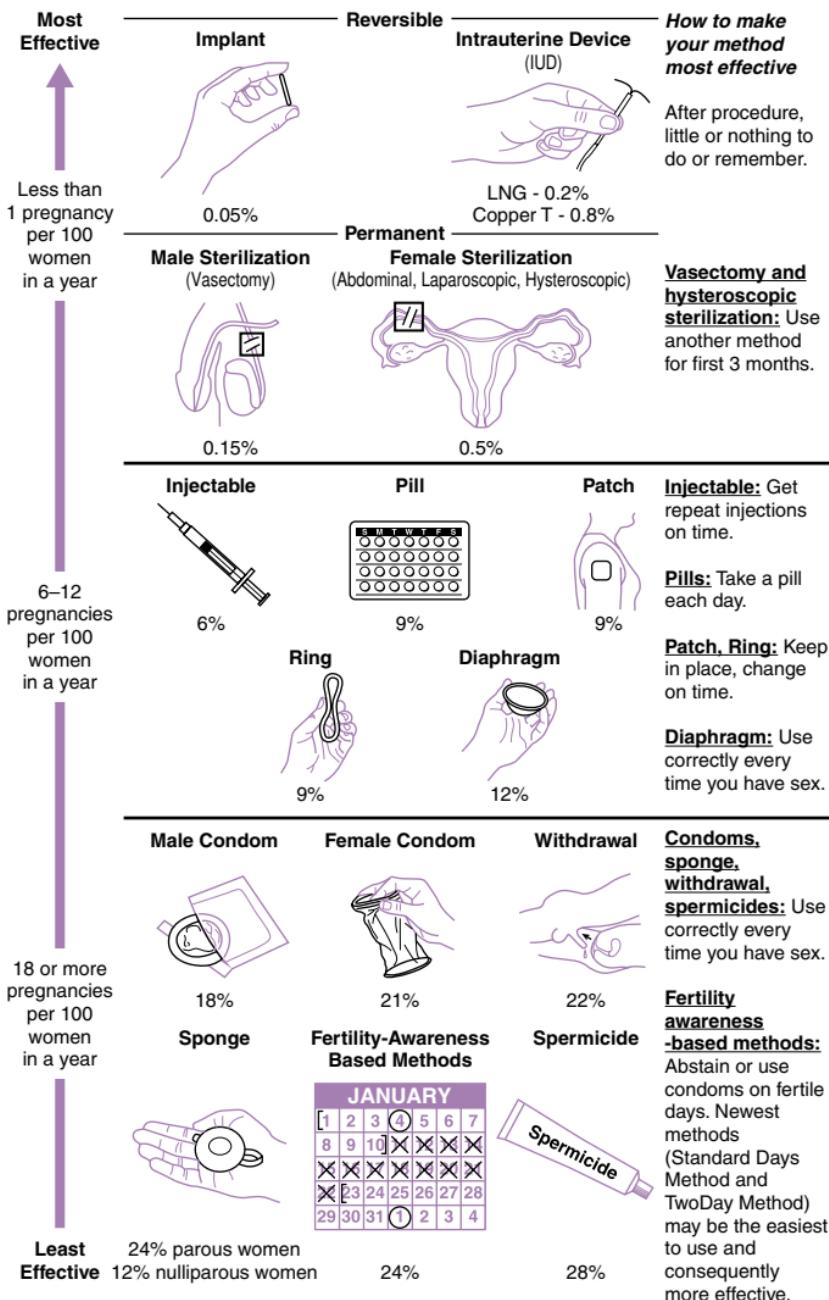
### 1. Special considerations in adolescents:

- a. Over 40% of adolescents in the United States have had sexual intercourse and over 75% of adolescent pregnancies are unplanned.<sup>34,35</sup>
- b. Barriers may include confidentiality concerns (e.g., fear of parental disclosure), fear of pelvic examination, and fear of medication side effects.
- c. **Long-acting reversible contraception (LARC)** methods have well-established safety and efficacy and are first-line contraceptive methods according to the ACOG and the AAP. Adherence and continuation rates of LARC methods in adolescents are superior to short-acting contraceptives. To avoid a delay in initiation, quick start method should be considered for most adolescents.<sup>36</sup>
- d. Providers should pay special attention to informed consent, confidentiality, parental involvement, insurance coverage, and cost. If an adolescent does not have or does not want to use their insurance coverage, refer to a clinic with Title X or other public funding (<http://www.hhs.gov/opa/>).
- e. Counseling should include discussion of need for barrier method to prevent STIs.

### 2. Method comparison (Fig. 5.2)<sup>37</sup>:

### 3. Contraception selection and initiation:

- a. **Selecting a contraceptive method:** Refer to the CDC Medical Eligibility Criteria (<http://www.cdc.gov/reproductivehealth/unintendedpregnancy/usmec.htm>) for relative and absolute contraindications for each



**CONDOMS SHOULD ALWAYS BE USED TO REDUCE THE RISK OF SEXUALLY TRANSMITTED INFECTIONS**

#### Other Methods of Contraception

**Lactational Amenorrhea Method:** LAM is a highly effective, temporary method of contraception.

**Emergency Contraception:** Emergency contraceptive pills or a copper IUD after unprotected intercourse substantially reduces risk of pregnancy.

**FIGURE 5.2—CONT'D**

Comparing effectiveness of family planning methods. The percentages indicate the failure rate of each contraceptive method, or the number of women who experienced an unintended pregnancy within the first year of typical use. (From Centers for Disease Control and Prevention. U.S. Selected Practice Recommendations for Contraceptive Use, 2016. *MMWR*. 2016;65(4):1–66. Available at: <https://www.cdc.gov/mmwr/volumes/65/rr/rr6504a1.htm>.)

hormonal contraceptive method and the CDC's Selected Practice Recommendations (<http://www.cdc.gov/reproductivehealth/unintendedPregnancy/USSPR.htm>) for minimum requirements to start each method.

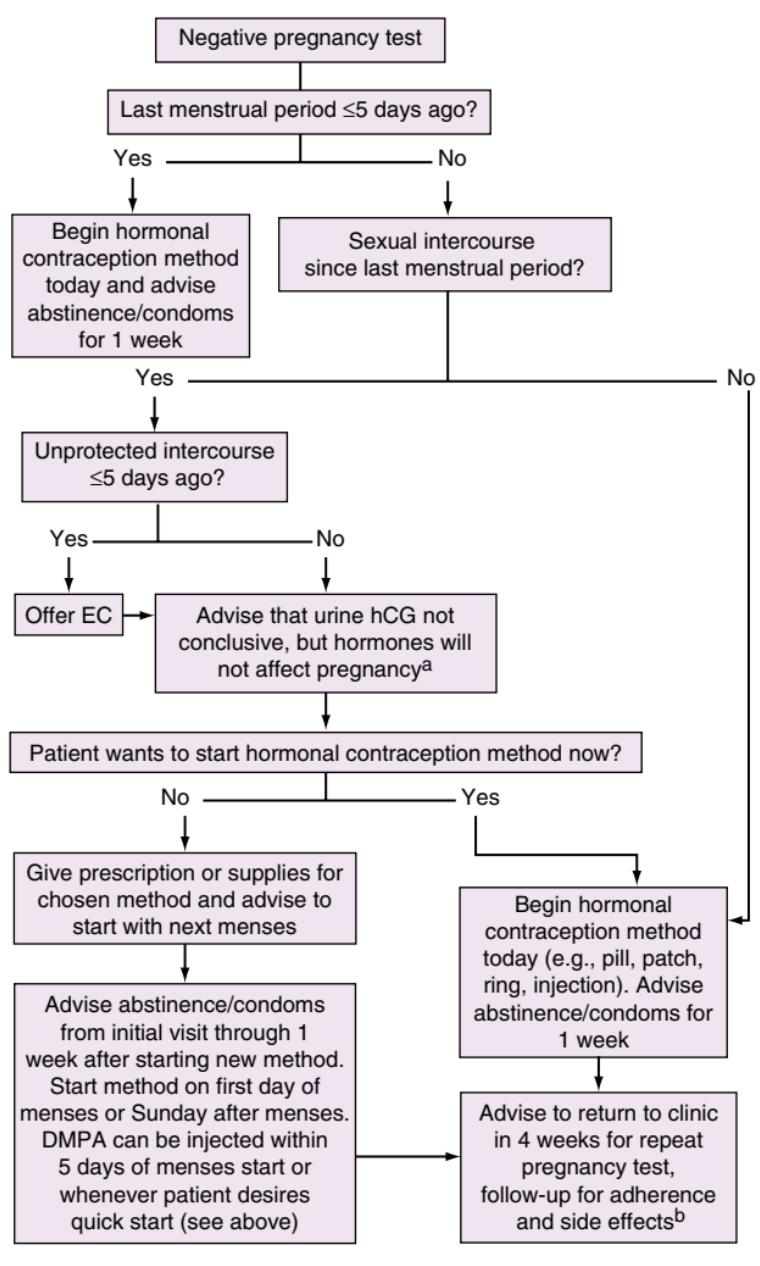
- (1) To start a hormonal method, the basic medical history should include assessment of clotting risk, blood pressure, pregnancy status, and any other pertinent medical comorbidities.
  - (2) Combined hormonal contraception is associated with a small increase in risk for thrombosis including deep vein thrombosis, myocardial infarction, and stroke.<sup>38</sup> The risk is higher in women who smoke more than 15 cigarettes a day, women over 35 years old, and women with other risk factors for cardiovascular disease.
  - (3) To support adherence and continuation, use a patient-centered approach, review method effectiveness, and provide anticipatory guidance regarding side effects of each method when assisting an adolescent in selecting a new contraceptive method.
- b. **Quick start** (Fig. 5.3): Starting a method of contraception on the day of the visit (not waiting until a new menstrual cycle begins) should be considered for most adolescents. Can be used for all methods, including LARC, if there is reasonable certainty that the patient is not pregnant (Box 5.3). A urine pregnancy test should be performed when using this approach.<sup>39</sup>

#### 4. Specific contraceptive methods:

Note: Contraceptive methods are described in order of effectiveness.

- a. **Intrauterine device (IUD)**<sup>36</sup>: LARC inserted into the uterus. Safe to use among adolescents, may be inserted without difficulty in most adolescents and nulliparous women; expulsion is uncommon. Among the most effective forms of birth control. Does not increase risk of infertility; baseline fertility returns rapidly after removal. Increased risk of pelvic infection with placement, but the absolute risk of infection is low and exists only within the first 3 weeks after placement. Screening is recommended for gonorrhea and chlamydia at the time of insertion based on the CDC guidelines as age (<25 years old) is a risk factor for STIs. Insertion should not be delayed for test results; treatment can occur without IUD removal.

- (1) Copper (Paragard): Hormone-free, FDA approved for 10 years of use although data supports potential efficacy for an additional 1 to 2 years.<sup>36,40</sup>

**FIGURE 5.3**

Algorithm for quick start initiation of contraception. *EC*, emergency contraception; *hCG*, human chorionic gonadotropin.

<sup>a</sup>Pregnancy tests may take 2 to 3 weeks after sex to be accurate.

<sup>b</sup>Consider pregnancy test at second depot medroxyprogesterone acetate (*DMPA* [*Depo-Provera*]) injection if quick start regimen was used and patient failed 4-week follow-up visit. (Modified from Zieman M, Hatcher RA, Cwiak C, et al. *A Pocket Guide to Managing Contraception*. Tiger, GA: Bridging the Gap Foundation; 2010:142.)

**BOX 5.3****HOW TO BE REASONABLY CERTAIN THAT A WOMAN IS NOT PREGNANT**

If the patient has no symptoms or signs of pregnancy and meets any of the following criteria:

1. Is  $\leq 7$  days after the start of normal menses
2. Has not had sexual intercourse since the start of last normal menses
3. Has been correctly and consistently using a reliable method of contraception
4. Is  $\leq 7$  days after spontaneous or induced abortion
5. Is within 4 weeks postpartum
6. Is fully or nearly fully breastfeeding, amenorrheic, and  $<6$  months postpartum

Adapted from Curtis KM, Jatlaoui TC, Tepper NK, et al. U.S. selected practice recommendations for contraceptive use, 2016. *MMWR Recomm Rep*. 2016;65(4):1–66.

- (2) Progestin-containing (Levonorgestrel): There are four types with differing amounts of progestin. Mirena is FDA approved for 5 years; data supports an additional 2 years.<sup>36,41</sup> Kyleena is FDA approved for 5 years. Liletta is FDA approved for 4 years; data supports an additional year.<sup>36,42</sup> Skyla is FDA approved for 3 years.
- (3) Changes in bleeding patterns are common in first months of use. Copper IUD may cause heavier menses. Many women using the levonorgestrel IUD will have a decrease in bleeding over time. First-line treatment for bleeding and spotting is NSAIDs.<sup>43</sup> Bleeding concerns that are not associated with device insertion should be evaluated for other etiologies.
- b. **Subdermal implant:** Progestin-only LARC, 4-cm rod inserted into the upper arm. Newer model (Nexplanon) is radio-opaque. FDA approved for 3 years; studies show efficacy for up to 5 years.<sup>41</sup> Minimal or no effect on bone density or body weight; causes a change in bleeding patterns. Return to fertility is rapid after removal. May be less effective for women who are overweight or obese. Among the most effective forms of birth control.
- (1) Removal requires a small incision and takes an average of 1 minute.
  - (2) Persistent irregular bleeding is the most common complaint resulting in implant removal, but continuation rates among adolescents remain high.<sup>44</sup> As opposed to levonorgestrel IUD, bleeding changes persist throughout duration of use. The bleeding pattern in the first 3 months of use is predictive of future bleeding. Important to provide preinsertion anticipatory guidance. Consider postinsertion management of bleeding with NSAIDs, combined OCPs, or doxycycline.<sup>37,45</sup>
- c. **Depot medroxyprogesterone acetate (DMPA [Depo-Provera]) injection:** Progestin-only injection into arm every 13 to 15 weeks. Typical failure

rate: 6%. Delayed return to fertility (9 to 18 months). Menstrual irregularity is common, but often resolves after several cycles. May cause weight gain, but not a uniform finding in studies.

- (1) Patient should be encouraged to receive adequate calcium and vitamin D due to association with decrease in bone mineral density.
- (2) FDA black box warning: Should not be used for longer than 2 consecutive years unless other forms of birth control are inadequate due to bone mineral density concerns. Bone density returns after discontinuation. The risk of loss of bone mineral density should be weighed against the need for effective contraception in the context of each adolescent.<sup>39</sup>

d. **Combined hormonal oral contraceptive pills (OCPs):** Combination of estrogen and progestin taken daily. "Low-dose" (35 mCg or less of ethinyl estradiol) pills are the recommended dosing for adolescents. Back-up method needed for at least 7 days after initiation. Typical failure rates are approximately 9% and may be higher in teens. Known to improve dysmenorrhea and are first-line therapy for endometriosis. Newer formulations exist, known as extended-cycle regimens, which reduce the number of menstrual cycles per year.

- (1) The first pill should be taken either on the day of the visit (quick start) or between the first and seventh day after the start of the menstrual period (most commonly Sunday).
- (2) Some pill packs have 28 pills, others have 21 pills. When the 28-day pack is empty, immediately start taking pills from a new pack. When the 21-day pack is empty, wait 1 week (7 days), then begin taking pills from a new pack.
- (3) If a pill is missed, it should be taken as soon as remembered, even if it means taking two pills in 1 day. If two or more pills missed, two pills should be taken daily until back on schedule and a backup method should be used for 7 days.

e. **Progestin-only pills:** Can be used for those with contraindications to estrogen-containing formulations. Require daily use and are more sensitive to timing (should be taken at same time each day); have no pill-free interval. Considered less effective than combined hormonal methods. Irregular bleeding is a common adverse effect.

f. **Transdermal (patch) contraceptive:** Contains estrogen and progestin, measures 1.75 x 1.75 in. Place on abdomen, upper torso, upper outer arm, or buttocks. Use one patch for 3 weeks, then remove for 1 week for withdrawal bleed. Greater exposure to estrogen than with other methods; may have more estrogen-related side effects. There may be a greater risk for thromboembolism compared to OCPs, though the data is not clear.<sup>46</sup> May be less effective in women who weigh more than 90 kg.

g. **Vaginal ring:** Flexible latex-free ring that contains estrogen and progestin. Placed in vagina for 3 weeks, removed for 1 week for

withdrawal bleeding. May be used continuously (avoiding week of menses) by replacing with a new ring every 4 weeks (or the same day every month) to help reduce pelvic pain and dysmenorrhea. May be removed for up to 3 hours (not recommended in adolescents).

Requires user comfort with insertion and removal. Screen for comfort with this method by asking if the adolescent is comfortable using tampons. Typical use failure rate similar to other combined hormonal methods (9%).

- h. **Barrier methods:** Require placement prior to sexual intercourse. Include cervical sponge, cervical cap, cervical shield, diaphragm (these methods are used in conjunction with spermicide), as well as female and male condoms. Male condom is most commonly used birth control among adolescents with a failure rate of 18% with typical use.<sup>37</sup> Use latex condoms only with water-based lubricants; oil-based lubricants are not recommended. While barrier methods are less effective than other methods of contraception, their use should still be emphasized for prevention of STIs.

- i. **Fertility awareness-based methods of pregnancy prevention:** Involves following a woman's menstrual cycle to help prevent pregnancy.

5. **Emergency contraception (EC)**<sup>47</sup>: Used to prevent pregnancy following unprotected sex (including sexual assault) or a recent possible failure of another method of contraception.

- a. Mechanism: Hormonal methods work by delaying ovulation. Copper IUD inhibits fertilization by affecting sperm viability and function. All methods are only effective before implantation takes place and will not disrupt an implanted pregnancy.

- b. Efficacy<sup>48</sup>: Copper IUD is the most effective method, but requires a clinic visit for insertion. Ulipristal is the next most effective, but requires a prescription. Levonorgestrel is the next most effective and is available over the counter. The Yuzpe regimen is the least effective and has the most side effects.

- c. Timing: Hormonal methods are most effective when given as soon as possible. Efficacy declines linearly with time but there is efficacy up to 120 hours after intercourse. Ulipristal and copper IUD maintain high efficacy when taken up to 120 hours after intercourse.

- d. Pregnancy should be excluded based on history, physical exam, or pregnancy testing before prescribing ulipristal or placing an IUD, as they may adversely affect an established pregnancy.

- e. Methods:

- (1) Progestin only: Levonorgestrel. Take 1.5 mg orally once (may be packaged as 1.5 mg tablet or two 0.75 mg tablets).

- (2) Antiprogestins: Ulipristal ("Ella"). Take 30 mg orally once.

Mifepristone is an alternative agent used in some countries as EC, but is not available in the U.S. for this purpose.

- (3) Ethynodiol diacetate plus levonorgestrel (Yuzpe regimen): Patients take multiple OCPs from packets designed for 28-day use. Take

equivalent of 100 mCg of ethinyl estradiol plus 500 mCg of levonorgestrel. Twelve hours later, take the same dose. For more precise instructions for a particular combination pill, refer to <https://ec.princeton.edu>.

- (4) Copper IUD may be inserted within 120 hours of coitus.

f. Guidelines:

- (1) Counseling about EC should be a routine part of anticipatory guidance for all female and male adolescents. Advance prescriptions should be considered for all adolescents.
- (2) Antiemetics can be used to prevent nausea and should be used prophylactically in the Yuzpe regimen.
- (3) May be combined with other ongoing methods of birth control.
- (4) OCPs may be started immediately after progestin-only or combined hormonal EC dosing has been completed. DMPA may be given the same day.
- (5) Patient should abstain from sexual intercourse or use barrier contraception for 7 days (14 if using ulipristal) or until next menses, whichever comes first.
- (6) Scheduled follow-up is not required after use of EC. However, women whose menses are delayed by a week or more, or have any signs of pregnancy (e.g., irregular menses, abdominal cramping), should be evaluated clinically or have a pregnancy test.

6. **Follow-up recommendations for contraception:** Two or three visits per year to monitor compliance, blood pressure, side effects, and satisfaction with chosen birth control option.

E. **Pregnancy**<sup>49,50</sup>

If pregnancy is suspected in an adolescent patient, take a sexual history and explore how the patient feels about a possible pregnancy in order to guide the rest of the visit.

1. **Diagnosis:**

- a. Perform urine hCG testing to diagnose the pregnancy. False-positives and false-negatives are possible; repeat urine testing or serum hCG testing may be indicated.
- b. If pregnancy is diagnosed, estimate the gestational age using the LMP. Confirm with a brief exam of uterine size. When in doubt, arrange an ultrasound and obstetric consultation promptly, as gestational age will affect counseling options.
- c. Share the diagnosis with the patient privately. Encourage them to involve a parent or legal guardian and facilitate the discussion, if necessary. Be familiar with local confidentiality laws, which vary by state.
- d. Review the patient's medications to ensure they are safe for pregnancy. Start patient on prenatal vitamin if not taking.

2. **Prenatal testing:** All pregnant adolescents should be tested for HIV, syphilis, hepatitis B, chlamydia, and gonorrhea at the first prenatal visit. If an infection is suspected or if there may be a delay in obstetric care, the pediatrician should perform the testing.

3. **Options counseling:** Counsel the adolescent on the importance of making a timely decision. The options depend on gestational age, but may include continuing the pregnancy and raising the infant, continuing the pregnancy and making an adoption plan, or terminating the pregnancy. If a pediatrician has personal limitations in offering a discussion of all three options, he/she should make a prompt referral to a colleague or consultant. Medical and surgical abortion may be available depending on the gestational age of the pregnancy and coexisting medical conditions. Medical abortion is generally available under 9 weeks of gestation; surgical abortion is generally available under 20 to 24 weeks of gestation.
4. **Complications:** First trimester complications include ectopic pregnancy and spontaneous abortion; immediate obstetric referral may be indicated for abdominal pain and/or vaginal bleeding in the pregnant patient.

### III. TRANSITIONING ADOLESCENTS INTO ADULT CARE<sup>51</sup>

All adolescents, particularly those with special healthcare needs or chronic conditions, benefit from careful attention to the process of transitioning to adult care. Transition planning should routinely occur during well-visits and should start at age 12. Resources for how to approach and organize the transition process including guidance on transition readiness and planning are available at <http://www.gotransition.org/>. See Chapter 9 for discussion of transition to adult care for youth with developmental disorders and disabilities.

### IV. WEB RESOURCES

#### A. Websites for Clinicians

- Centers for Disease Control and Prevention (CDC) on contraception: <http://www.cdc.gov/reproductivehealth/unintendedpregnancy/contraception.htm>
- CDC on sexually transmitted infections (STI): <https://www.cdc.gov/std/life-stages-populations/adolescents-youngadults.htm>
- Society for Adolescent Health and Medicine: <http://www.adolescenthealth.org>

#### B. Websites for Patients

- Drug abuse: <http://www.teens.drugabuse.gov>
- Sexual health: <http://www.plannedparenthood.org/>, <http://www.bedsider.org>
- CDC resources for Lesbian, Gay, Bisexual, & Transgender (LGBT) youth: <https://www.cdc.gov/lgbthealth/youth-resources.htm>

### REFERENCES

A complete list of references can be found online at [www.expertconsult.com](http://www.expertconsult.com).

**BOX EC 5.A****OBTAINING THE SEXUAL HISTORY: THE FIVE PS**

1. Partners
  - “Do you have sex with men, women, or both?”
  - “In the past 2 months, how many partners have you had sex with?”
  - “In the past 12 months, how many partners have you had sex with?”
  - “Is it possible that any of your sex partners in the past 12 months had sex with someone else while they were still in a sexual relationship with you?”
2. Prevention of Pregnancy
  - “What are you doing to prevent pregnancy?”
3. Protection from STIs
  - “What do you do to protect yourself from STIs including HIV?”
4. Practices
  - “To understand your risk for STIs, I need to understand the kind of sex you have had recently.”
  - “Have you had vaginal sex, meaning ‘penis in vagina sex?’” If yes, “Do you use condoms: never, sometimes, or always?”
  - “Have you had anal sex, meaning “penis in rectum/anus sex?”” If yes, “Do you use condoms: never, sometimes, or always?”
  - “Have you had oral sex, meaning ‘mouth on penis/vagina sex?’”
  - “Have you had vaginal or anal sex using fingers or sex toys?”

For condom answers:

- If “never”: “Why don’t you use condoms?”
- If “sometimes”: “In what situations, or with whom, do you not use condoms?”

5. Past History of STIs

- “Have you ever had an STI?”
- “Have any of your partners had an STI?”

Additional questions to identify HIV and viral hepatitis risk include:

- “Have you or any of your partners ever injected drugs?”
- “Have any of your partners exchanged money or drugs for sex?”
- “Is there anything else about your sexual practices I need to know about?”

HIV, Human immunodeficiency virus; STI, sexually transmitted infection.

Modified from the Centers for Disease Control and Prevention (CDC). *Sexually Transmitted Diseases Treatment Guidelines 2015, Clinical Prevention Guidance*. Available at <https://www.cdc.gov/std/tg2015/clinical.htm>.

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# Chapter 6

## Analgesia and Procedural Sedation

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 See additional content on Expert Consult

### I. PAIN ASSESSMENT

#### A. Infant<sup>1</sup>

##### 1. Physiologic response

- Characterized by oxygen desaturation, crying, diaphoresis, flushing or pallor, and increases in blood pressure, heart rate, and respiratory rate.
- Seen primarily in acute pain; subsides with continuing/chronic pain.

##### 2. Behavioral response

- Observe characteristics and duration of cry, facial expressions, visual tracking, body movements, and response to stimuli.
- Neonatal Infant Pain Scale (NIPS): Behavioral assessment tool for the preterm neonate and full-term neonate up to 6 weeks after birth.
- FLACC scale ([Table 6.1](#)): Measures and evaluates pain interventions by quantifying pain behaviors, including **F**acial expression, **L**eg movement, **A**ctivity, **C**ry, and **C**onsolability, with scores ranging from 0 to 10.<sup>2</sup> The Revised FLACC scale is reliable in children with cognitive impairment.<sup>3</sup>

#### B. Preschooler

In addition to physiologic and behavioral responses, the **FACES** pain scale revised (FPS-R) can be used to assess pain intensity in children as young as 3 years of age by having the patient point to the image on the scale that best characterizes their pain ([Fig. 6.1](#)).

#### C. School-Age and Adolescent

Evaluate physiologic and behavioral responses; ask about description, location, and character of pain. Starting at the age of 7 years, children can use the standard subjective pain rating scale, in which 0 is no pain and 10 is the worst pain ever experienced.

### II. ANALGESICS<sup>1</sup>

#### A. Safety

##### 1. Combined Analgesics

- Danger of acetaminophen toxicity when using combined opioid-acetaminophen products (oxycodone or hydrocodone with acetaminophen).

**TABLE 6.1****FLACC PAIN ASSESSMENT TOOL****FACE**

- 0—No particular expression or smile  
 1—Occasional grimace or frown, withdrawn, disinterested  
 2—Frequent to constant frown, quivering chin, clenched jaw

**LEGS**

- 0—Normal position or relaxed  
 1—Uneasy, restless, tense  
 2—Kicking or legs drawn up

**ACTIVITY**

- 0—Lying quietly, normal position, moves easily  
 1—Squirming, shifting back and forth, tense  
 2—Arched, rigid, or jerking

**CRY**

- 0—No cry (awake or asleep)  
 1—Moans or whimpers, occasional complaint  
 2—Crying steadily, screams or sobs, frequent complaints

**CONSOLABILITY**

- 0—Content, relaxed  
 1—Reassured by occasional touching, hugging, or being talked to; distractible  
 2—Difficult to console or comfort

Modified from Manworren R, Hynan L. Clinical validation of FLACC: preverbal patient pain scale. *Pediatr Nurs.* 2003;29:140–146.

**Wong-Baker FACES® pain rating scale****FIGURE 6.1**

Wong-Baker FACES Pain Rating Scale (From [wongbakerfaces.org](http://wongbakerfaces.org): Wong-Baker FACES Foundation [2016]. Wong-Baker FACES® Pain Rating Scale. Retrieved with permission from <http://www.WongBakerFACES.org>. Originally published in Whaley & Wong's *Nursing Care of Infants and Children*. © Elsevier Inc.)

- b. **Preferable to prescribe opioids and acetaminophen separately.**
2. Codeine
  - a. **Not recommended for use in children.**
  - b. Five percent of the population show ultra-rapid metabolism of codeine to morphine (the active metabolite), which can lead to dangerously high levels. This is especially unsafe after tonsillectomy and adenoidectomy (T&A) performed for sleep apnea.<sup>4</sup>

- c. Little to no analgesic effect in newborns and approximately 10% of the U.S. population.<sup>5</sup>
- 3. Meperidine
  - a. Not recommended for use in children due to risk of neurotoxicity (agitation, tremors, myoclonus, and seizures), especially when renal dysfunction is present.<sup>6</sup>
  - b. Contraindicated in patients receiving monoamine oxidase (MAO) inhibitors.
- 4. Tramadol
  - a. Opioid pain reliever, with additional effects on nonopioid receptors.
  - b. May be over-metabolized to an active opiate metabolite, resulting in potentially fatal respiratory depression.
  - c. In 2017, the FDA issued its strongest warning against use in children; therefore, administration is considered off-label.<sup>7</sup>

### B. Nonopiod Analgesics

Weak analgesics with antipyretic activity are commonly used to manage mild to moderate pain of nonvisceral origin. Can be administered alone or in combination with opioids.

- 1. **Acetaminophen [by mouth (PO)/per rectum (PR)/intravenous (IV)]:** Weak analgesic with no antiinflammatory activity, platelet inhibition, or gastrointestinal (GI) irritation. **Hepatotoxicity can occur with high doses.**
- 2. **Aspirin (PO/PR):** Associated with platelet inhibition and GI irritation. **Avoid for analgesia in pediatrics due to risk of Reye syndrome.**
- 3. **Nonsteroidal antiinflammatory drugs (NSAIDs):** Ibuprofen (PO/IV), ketorolac [IV/intramuscular (IM)/PO/intranasal (IN)], naproxen (PO), diclofenac (PO/IV), and celecoxib (PO).
  - a. Use with extreme caution in children less than 6 months of age due to concern for adverse GI effects and risk of renal failure.
  - b. Especially useful for sickle cell disease, bony, rheumatic, and inflammatory pain.
  - c. Concurrent histamine-2-receptor blocker or proton pump inhibitor is recommended with prolonged use given GI side effects.
  - d. Other adverse effects include interference with platelet aggregation, hepatitis, bronchoconstriction, hypersensitivity reactions, and azotemia. Avoid in patients with severe renal disease, dehydration, or heart failure.

**NOTE:** Ketorolac is a potent analgesic. Limit duration of therapy to less than 5 days to limit renal toxicity.

### C. Opioids (Table 6.2)

- 1. Produce analgesia by binding to  $\mu$  receptors in the brain and spinal cord.
- 2. **Side effects:** Pruritus, nausea, vomiting, constipation, urine retention, and (rarely) respiratory depression and hypotension. Prescribe a bowel regimen when prescribing opioids.

**TABLE 6.2****COMMONLY USED OPIOIDS**

Drug	Morphine Equivalence Ratio	Onset (min)	Duration (hr)	Side Effects	Comments
Fentanyl	80–100	IV: 1–2	0.5–1	Pruritus Bradycardia <b>Chest wall rigidity with doses &gt;5 mCg/kg (but can occur at all doses); treat with naloxone or neuromuscular blockade.</b>	Risk of cardiovascular instability is lower than other opioids, making it relatively safer in hypovolemia, congenital heart disease, or head trauma Respiratory depressant effect much longer (4 hr) than analgesic effect Most commonly used opioid for short, painful procedures, but transdermal route is more effective in chronic pain situations <sup>a</sup>
Hydromorphone	4–7	IV/SQ: 5–10 PO: 30–60	3–4		Less sedation, nausea, and pruritus than morphine
Methadone	0.25–1 <sup>b</sup>	IV: 5–10 PO: 30–60	4–24		Initial dose may produce analgesia for 3–4 hr; duration of action is increased with repeated dosing Useful for neuropathic pain and opioid weaning due to unique mechanism of NMDA blockade
Morphine	1	IV: 5–10 IM/SQ: 10–30 PO: 30–60	IV: 3–4 IM/SQ/PO: 4–5	Seizures in neonates. Can cause significant histamine release.	Available in sustained-release form for chronic pain
Oxycodone	1.5	30–60	3–4		Available in sustained-release form for chronic pain

<sup>a</sup>Removing a transdermal fentanyl patch does not stop opioid uptake from the skin; fentanyl will continue to be absorbed for 12–24 hours after patch removal (fentanyl 25-mCg patch administers 25 mCg/hr of fentanyl).

<sup>b</sup>Morphine-to-methadone conversion in the tolerant/dependent patient is variable. Consider starting at the lowest conversion ratio: 0.25.

IM, Intramuscular; IV, intravenous; mCg, microgram; PO, by mouth; SQ, subcutaneous.

Data from Yaster M, Cote C, Krane E, et al. *Pediatric Pain Management and Sedation Handbook*. St. Louis: Mosby; 1997:29–50.

**3. Patients with renal failure.**

- a. **Morphine:** Avoid use secondary to decreased excretion of the active metabolite that can result in respiratory depression.
- b. **Preferred choices:** Fentanyl, remifentanil, methadone, hydromorphone, oxycodone.
4. Long-acting opioids (methadone, extended-release tablets, and patches) are not recommended for acute pain.
5. Although opioids are essential for the treatment of moderate to severe pain, a thoughtful approach is recommended with the quantity that is dispensed, as studies have shown that over 50% of opioid doses dispensed are not consumed.<sup>8</sup> There is need for further research to develop evidence-based opioid prescribing guidelines for treating acute pain in children.

**D. Local Anesthetics<sup>9-12</sup>**

Administered topically or subcutaneously to surround peripheral nerves (peripheral block) or centrally (epidural/spinal block). Temporarily block nerve conduction at the sodium channel.

**1. For all local anesthetics, 1% solution = 10 mg/mL<sup>b</sup>**

**2. Topical local anesthetics (Table 6.3)<sup>12</sup>**

**3. Injectable local anesthetics (Table 6.4):**

- a. Subcutaneous infiltration of the skin at the site: Used for painful procedures such as wound closure or lumbar puncture.
- b. Use of a 27- to 30-gauge needle, alkalization, warming the solution to 37°C to 42°C, and a slow injection can reduce stinging from injection. Alkalize by adding 1 mL (1 mEq<sup>c</sup>) sodium bicarbonate to 9 mL lidocaine (or 29 mL bupivacaine).
- c. To enhance efficacy and duration, add epinephrine (5 to 10 mCg<sup>d</sup> of epinephrine to 1 mL of local anesthetic) to decrease vascular uptake. **Never use local anesthetics with epinephrine in areas supplied by end arteries (e.g., pinna, fingers, toes, nasal tip, penis).**
- d. **Maximum volume (mL) = (maximum mg/kg dose × weight in kg)/(% solution × 10).** See Table 6.4 for maximum doses.
- e. Toxicity: Central nervous system (CNS) and cardiac toxicity are of greatest concern. CNS symptoms are seen before cardiovascular collapse. Always calculate the maximum volume of the local anesthetic and always draw up less than that. Bupivacaine is associated with more severe cardiac toxicity than lidocaine.
  - (1) Progression of symptoms: Perioral numbness → dizziness → auditory disturbances → muscular twitching → unconsciousness → seizures → coma → respiratory arrest → cardiovascular collapse.

<sup>a</sup>mL, milliliter.

<sup>b</sup>mEq, milliequivalent.

<sup>c</sup>mCg, microgram.

**TABLE 6.3****COMMONLY USED TOPICAL LOCAL ANESTHETICS**

Components		Indications	Peak Effect	Duration <sup>a</sup>	Cautions
EMLA	Lidocaine 2.5% Prilocaine 2.5%	<b>Intact skin only</b> Venipuncture, circumcision, LP, abscess drainage, BMA	60 min	90 min	Methemoglobinemia: Not for use in patients predisposed to methemoglobinemia (G6PD deficiency) Infants <3 months of age: Use sparingly (up to 1 g is safe)
LMX	Lidocaine 4%	Same as EMLA	30 min	60 min	Same as EMLA
LET	Lidocaine 4% Epinephrine 0.1% Tetracaine 0.5% Can be mixed with cellulose to create a gel	<b>Safe for nonintact skin/ lacerations</b> Can be used to attain hemostasis with simple lacerations	30 min	45 min	Vasoconstriction: Contraindicated in areas supplied by end arteries (e.g., pinna, nose, penis, digits) Avoid contact with mucous membranes Not for use in contaminated wounds
Viscous lidocaine	Lidocaine 2% (May be mixed with Aluminum/Magnesium Hydroxide/Simethicone (Maalox) and diphenhydramine in a 1:1:1 ratio for palatability when administered orally)	<b>Safe for nonintact skin</b> Mucous membranes (e.g., urethral catheter placement, mucositis)	10 min	30 min	Overuse can lead to life-threatening toxicity Not to be used for teething

<sup>a</sup>Approximate.

BMA, Bone marrow aspiration; EMLA, eutectic mixture of local anesthetics; G6PD, glucose-6-phosphate dehydrogenase; LP, lumbar puncture; min, minutes.

Data from Krauss B, Green SM. Sedation and analgesia for procedures in children. *N Engl J Med*. 2000;342:938–945; and Zempsky W, Cravero J. Relief of pain and anxiety in pediatric patients in emergency medical systems. *Pediatrics*. 2004;114:1348–1356.

**TABLE 6.4****COMMONLY USED INJECTABLE LOCAL ANESTHETICS<sup>1,10</sup>**

Agent	Concentration (%) <sup>a</sup>	Max Dose (mg/kg)	Onset (min)	Duration (hr)
Lidocaine	0.5–2	5	3	0.5–2
Lidocaine with epinephrine	0.5–2	7	3	1–3
Bupivacaine	0.25–0.75	2.5	15	2–4
Bupivacaine with epinephrine	0.25–0.75	3	15	4–8
Bupivacaine with Lidocaine mixture	Variable	b	3–15	0.5–4

<sup>a</sup>(1% solution = 10 mg/mL).

<sup>b</sup>[(mg/kg used of bupivacaine)/2.5 mg/kg × 100] + [(mg/kg used of lidocaine)/5 mg/kg × 100]. Toxicity occurs when the sum is >100%.

Data from St Germain BA. The management of pain in the emergency department. *Pediatr Clin North Am.* 2000;47:651–679; Yaster M, Cote C, Krane E, et al. *Pediatric Pain Management and Sedation Handbook*. St. Louis: Mosby; 1997:51–72.

<b>Lipid emulsion 20%</b> (Precise volume and flow rate are not crucial)	
Greater than 70 kg patient	Less than 70 kg patient
<b>Bolus 100 mL lipid emulsion 20%</b> rapidly over 2–3 minutes <ul style="list-style-type: none"> <li>Lipid emulsion infusion 200–250 mL over 15–20 minutes</li> </ul>	<b>Bolus 1.5 mL/kg lipid emulsion 20%</b> rapidly over 2–3 minutes <ul style="list-style-type: none"> <li>Lipid emulsion infusion ~0.25 mL/kg/min (ideal body weight)</li> </ul>
<b>If patient remains unstable:</b> <ul style="list-style-type: none"> <li>Re-bolus once or twice at the same dose and double infusion rate; be aware of dosing limit (12mL/kg)</li> <li>Total volume of lipid emulsion can approach 1 L in a prolonged resuscitation (e.g., &gt;30 minutes)</li> </ul>	

**FIGURE 6.2**

Lipid emulsion 20%.

- (2) Summary of American Society of Regional Anesthesia and Pain Medicine (ASRA) Checklist for Treatment of Local Anesthetic Systemic Toxicity (LAST) ([https://www.asra.com/content/documents/asra\\_last\\_checklist\\_2018.pdf](https://www.asra.com/content/documents/asra_last_checklist_2018.pdf))
  - (a) Stop injecting local anesthetic.
  - (b) Call for help and obtain 20% lipid emulsion (see Fig. 6.2 for dosing).
  - (c) Manage airway: ventilate with 100% FiO<sub>2</sub> (fraction of inspired oxygen), insert advanced airway if needed.
  - (d) Control seizures with benzodiazepines; avoid large doses of propofol due to the potential to exacerbate hypotension.

- (e) Treat hypotension and bradycardia—start cardiopulmonary resuscitation (CPR) if pulseless. Avoid hyperventilation.
- (3) If concerned for systemic toxicity, contact an anesthesiologist and call poison control (1-800-222-1222).

6

## E. Nonpharmacologic Measures of Pain Relief<sup>13,14</sup>

### 1. Sucrose for neonates (Sweet-Ease):

- a. Indications: Painful minor procedures (heel lance, venipuncture, and intramuscular injection) in neonates and infants. Has not been shown to be effective for relief of circumcision pain. Strongest evidence for infants aged 0 to 1 month,<sup>13</sup> but additional evidence suggests efficacy up to 12 months.<sup>14</sup>
- b. Procedure: Administer up to 2 mL of 24% sucrose into the infant's mouth by syringe or from a nipple/pacifier ~2 minutes before the procedure. Effective doses in very low-birth-weight infants may be as low as 0.05 to 0.1 mL.
- c. An additional dose may be administered within a relatively short period of time for multiple procedures, but it should not be administered more than twice in 1 hour.
- d. Use along with other age-appropriate nonpharmacologic measures listed below.
- e. Avoid if patient is unable to appropriately feed by mouth or cannot safely handle oral secretions.

### 2. Other: Parental presence/holding, distraction with toys, child life specialists, guided meditation/coping skills, virtual reality simulations.

## III. PATIENT-CONTROLLED ANALGESIA (PCA)

### A. Definition

PCA enables a patient to receive a limited number of small doses (boluses) of an analgesic with or without a continuous (basal) infusion on an as-needed basis. In children younger than 6 years old or with physical/mental disability, a family member, caregiver, or nurse may administer supplemental (bolus) doses.

### B. Indications

- 1. Moderate to severe pain of acute or chronic nature. Commonly used in sickle cell disease, postsurgery, posttrauma, burns, and cancer.
- 2. Useful for preemptive pain management (e.g., dressing changes).

### C. Routes of Administration

IV or epidural

### D. Agents (Table 6.5)

### E. Adjuvants

- 1. Low-dose naloxone (Narcan) infusion reduces incidence of pruritus and nausea associated with narcotic administration.

**TABLE 6.5****ORDERS FOR PATIENT-CONTROLLED ANALGESIA**

Drug	Basal Rate (mCg/kg/hr)	Bolus Dose (mCg/kg)	Lockout Period (min)	Boluses (hr)	Max Dose (mCg/kg/hr)
Morphine	10–30	10–30	6–10	4–6	100–150
Hydromorphone	3–5	3–5	6–10	4–6	15–20
Fentanyl	0.5–1	0.5–1.0	6–10	2–3	2–4

mCg, Microgram.

Data from Yaster M, Cote C, Krane E, et al. *Pediatric Pain Management and Sedation Handbook*. St. Louis: Mosby; 1997:100.

- Low-dose ketamine infusion has a narcotic sparing effect. It is especially helpful in chemotherapy-induced mucositis, visceral pain, and neuropathic pain. Its mechanism of action is by N-methyl-D-aspartate (NMDA) blockade. May be used with or as an alternative to methadone.

#### **F. Side Effects of Opioid Patient-Controlled Analgesia**

Pruritus, nausea, constipation, urine retention, excessive drowsiness, and respiratory depression.

### **IV. OPIOID TAPERING**

#### **A. Indications**

Because of the development of dependence and potential for withdrawal, a tapering schedule is required if the patient has received frequent opioid analgesics for >5 days.

#### **B. Withdrawal**

- See Box 18.1 for symptoms of opioid withdrawal.**
- Onset of signs and symptoms:** 6 to 12 hours after the last dose of morphine and 36 to 48 hours after the last dose of methadone.
- Duration:** 7 to 14 days, with a peak intensity reached within 2 to 4 days.

#### **C. Recommendations for Tapering**

- Conversion:** All drugs should be converted to a single equi-analgesic member of that group (see Table 6.2).
- PCA wean:** Drug dosing should be changed from continuous/intermittent IV infusion to PO basal/bolus therapy. If the patient is on PCA, once the first PO dose is administered, the PCA basal infusion should be stopped 30 to 60 minutes later. PCA bolus doses should be continued but reduced by 25% to 50%. If no further bolus doses are administered in the next 6 hours, the PCA should be discontinued. If the patient continues to experience pain, consider increasing scheduled PO dose, administering a rescue one-time PO bolus dose, or adding an adjuvant analgesic (e.g., NSAID).
- Slow dose decrease:** During an intermittent IV/PO wean, the total daily dose should be decreased by 10% to 20% of the original dose every 1 to 2 days (e.g., to taper a morphine dose of 40 mg/day, decrease the daily dose by 4 to 8 mg every 1 to 2 days).

4. **Oral regimen:** If not done previously, IV dosing should be converted to equivalent PO administration 1 to 2 days before discharge, and titration should be continued as outlined previously.
5. **Adjunctive therapy:**
  - a. Clonidine in combination with an opioid decreases the length of time needed for opioid weaning in neonatal abstinence syndrome, with few short-term side effects. Long-term safety has yet to be thoroughly investigated, but follow-up after 1 year on motor, cognitive, and language scores showed no difference in those treated with clonidine.<sup>15,16</sup>
  - b. PO and transdermal clonidine have a potential role for sedation, analgesia, and iatrogenic drug withdrawal in critically ill children, but current reports are retrospective or small clinical trials with significant heterogeneity in dosing, so further research is necessary. Transdermal dosing should not be used in children aged <1 year due to altered skin absorption.<sup>17</sup>
  - c. Studies have shown efficacy of  $\alpha_2$ -adrenergic agonists in treating opioid withdrawal and reducing doses of methadone, but the duration of treatment was longer with  $\alpha_2$ -adrenergic agonists, and there were fewer adverse effects with methadone.<sup>18</sup>
  - d. Dexmedetomidine is an  $\alpha_2$ -adrenoreceptor agonist, which produces sedation and mild analgesia, with minimal to no respiratory depression. Administered as a continuous infusion, it has been shown to reduce opioid requirements and facilitate opioid weaning.

#### D. Examples

See Box EC 6.A for example of opioid wean.

## V. PROCEDURAL SEDATION<sup>1,9–12,19–21</sup>

#### A. Definitions

1. **Mild sedation (anxiolysis):** Intent is anxiolysis with maintenance of consciousness.
2. **Moderate sedation:** Formerly known as *conscious sedation*. A controlled state of depressed consciousness during which airway reflexes and patency are maintained. Patient responds appropriately to age-appropriate commands (e.g., “Open your eyes”) and light touch. Practically obtained any time a combination of a sedative-hypnotic and an analgesic are used.
3. **Deep sedation:** A controlled state of depressed consciousness during which *airway reflexes and patency may not be maintained*, and the child is unable to respond to physical or verbal stimuli. In practice, deep sedation is required for most painful procedures in children. Practically obtained with propofol.
4. **Dissociative sedation:** Unique state of sedation achieved with ketamine characterized by a deep level of depressed consciousness and

**TABLE 6.6****FASTING RECOMMENDATIONS FOR ANESTHESIA**

Food Type	Minimum Fasting Period (hr)
Clear liquids	2
Breast milk	4
Nonhuman milk, formula	6
Solids	8

Data from Practice guidelines for preoperative fasting and the use of pharmacologic agents to reduce the risk of pulmonary aspiration: application to healthy patients undergoing elective procedures. A report by the American Society of Anesthesiologists Task Force on Preoperative Fasting and Use of Pharmacological Agents to Reduce the Risk of Pulmonary Aspiration (Online). <http://anesthesiology.pubs.asahq.org/article.aspx?articleid=1933410>.

analgesia. Airway reflexes and patency are generally maintained; however, excessive oral secretions may become problematic, occasionally resulting in micro-aspiration or laryngospasm.

### B. Preparation

1. The patient should be **NPO** for solids and liquids (Table 6.6).<sup>19</sup> Per American Society of Anesthesiologists (ASA) and American Academy of Pediatrics (AAP) guidelines, children receiving moderate sedation for elective procedures should follow the same fasting guidelines as those for general anesthesia.<sup>20,22</sup> For urgent/emergent sedation when children are not NPO, the risks of sedation and possible aspiration must be balanced against the benefits of performing the procedure promptly. Recent studies suggest that NPO status for liquids and solids may not be statistically associated with aspiration, although studies are limited as aspiration is a relatively uncommon complication.<sup>23</sup>

2. **Focused patient history:**

- a. Allergies, medications, and any history of a previous reaction to anesthesia or sedation.
- b. Assess for the possibility of an adverse airway event occurring with sedation (hypoxemia, hypercarbia, inability to mask ventilate, etc.). This can occur from: (1) mechanical airway obstruction (micrognathia, tonsillar and/or adenoid hypertrophy, large tongue, history of snoring, presence of noisy breathing, diagnosis of obstructive sleep apnea, obesity, presence of a craniofacial syndrome), (2) lung disease (history of prematurity, chronic lung disease or bronchopulmonary dysplasia, asthma), or (3) presence of a recent upper respiratory infection (URI). A history of a URI increases the risk of laryngospasm and/or bronchospasm; therefore, one must weigh the risks/benefits of providing sedation after a recent URI versus need for immediate interventional procedure. For elective procedures requiring sedation, it is best to wait 2 to 4 weeks after resolution of illness to reduce the risk of an adverse event.<sup>24</sup>
- c. Assess aspiration risk (neuromuscular disease, esophageal disease, altered mental status, obesity, pregnancy).

**BOX EC 6.A****EXAMPLE OF OPIOID TAPERING**

Patient on morphine patient-controlled analgesia (PCA) to be converted to oral (PO) morphine with home weaning.

For example: morphine PCA basal rate = 2 mg/hr, average bolus rate = 0.5 mg/hr

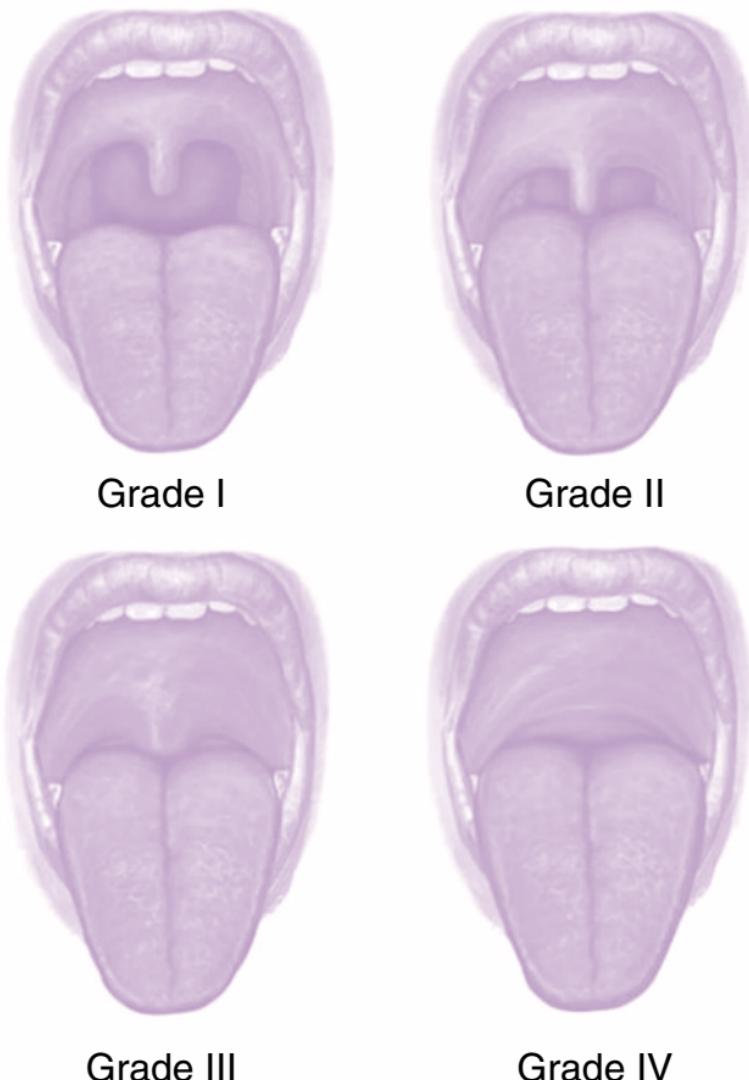
Step 1: Calculate daily dose: basal + bolus =  $(2 \text{ mg/hr} \times 24 \text{ hr}) + (0.5 \text{ mg/hr} \times 24 \text{ hr}) = 60 \text{ mg}$  intravenous (IV) morphine

Step 2: Convert according to drug potency: morphine IV/morphine oral = approx. 3:1 potency;  $3 \times 60 \text{ mg} = 180 \text{ mg}$  PO morphine

Step 3: Prescribe 90 mg BID or 60 mg TID; wean 10%–20% of original dose (30 mg) every 1–2 days

*BID*, Twice daily; *IV*, intravenous; *mg*, milligram; *PCA*, patient controlled analgesia; *PO*, by mouth; *TID*, three times daily.

Data from Yaster M, Cote C, Krane E, et al. *Pediatric Pain Management and Sedation Handbook*. St. Louis: Mosby; 1997:29–50.



**FIGURE 6.3**

Mallampati classification system.

- d. Presence of kidney/liver disease (may prolong sedative effect) and cardiac disease (potential for hemodynamic instability with sedative administration).
3. **Physical examination:** Specific attention to mouth opening and neck extension. Use the Mallampati classification system to assess the airway for likelihood of difficult direct laryngoscopy and intubation ([Fig. 6.3](#)).
4. **Determine ASA Physical Status Classification (Table 6.7):** Class I and II patients are generally good candidates for mild, moderate, or deep sedation outside of the operating room.<sup>20</sup>

**TABLE 6.7****ASA PHYSICAL STATUS CLASSIFICATION**

Class I	A normally healthy patient
Class II	A patient with mild systemic disease (e.g., controlled reactive airway disease)
Class III	A patient with severe systemic disease (e.g., a child who is actively wheezing)
Class IV	A patient with severe systemic disease that is a constant threat to life (e.g., a child with status asthmaticus)
Class V	A moribund patient who is not expected to survive without the operation (e.g., a patient with severe cardiomyopathy requiring heart transplantation)
Class VI	A declared brain-dead patient whose organs are being removed for donor purposes

**5. Always have an emergency plan ready:**

- a. Make sure qualified backup personnel and equipment are close by.
- b. Complications most often occur 5 to 10 minutes after administration of IV medication and immediately after a procedure is completed (when the stimuli associated with the procedure are removed).<sup>11</sup>

**6. Personnel:** Two providers are required. One provider should perform the procedure, and a separate provider should monitor the patient during sedation and recovery.

**7. Ensure IV access** prior to induction by flushing with saline.

Subcutaneous infiltration of a sedative can cause unpredictable or prolonged sedation.

**8. Have airway/intubation equipment immediately available** (see [Chapter 1](#)).

**9. Emergency medications:** Always have emergency medications for rapid sequence intubation and CPR immediately available.

**10. Reversal agents** should be readily available (naloxone, flumazenil).

**C. Monitoring**

**1. Vital signs:** Baseline vital signs should be obtained. Heart rate, oxygen saturation, and respiratory rate should be continuously monitored, and blood pressure monitored intermittently (every 3 to 5 minutes) until a pre-sedation level of consciousness is achieved.

**NOTE: Unrecognized apnea is often followed by desaturation within 1 to 2 minutes when not receiving supplemental oxygenation.**

**Administration of supplemental oxygen can further delay recognition of apnea because the onset of desaturation may occur more than 2 minutes after apnea.**

**2. Airway:** Airway patency and adequacy of ventilation should be frequently assessed through capnography (e.g., continuous end-tidal

carbon dioxide [CO<sub>2</sub>]), auscultation, and direct visualization. This can help ensure immediate recognition of apnea, and appropriate measures may be taken before desaturation occurs.

#### D. Pharmacologic Agents

1. **Goal of procedural sedation:** The administration of medications to provide appropriate levels of analgesia, sedation, and anxiolysis so that the procedure can occur without the need to secure the airway.
2. **CNS, cardiovascular, and respiratory depression** may always occur; occurs more commonly when combining sedative drugs and/or opioids, or with rapid drug administration. It is always best to titrate medications to the desired level of sedation.
3. **Common sedative/hypnotic agents (Box 6.1).** Also see **Table 6.2** and **Table 6.8** for more information on opioids and barbiturates/benzodiazepines.
4. Reversal agents:
  - a. Naloxone: Opioid antagonist. See **Box 6.2** for naloxone administration protocol.
  - b. Flumazenil: Benzodiazepine antagonist.

6

#### E. Discharge Criteria<sup>20</sup>

1. The patient can maintain a patent airway without requiring supplemental oxygen. There should also be no compromise in cardiovascular function.
2. The patient should be easily arousable with intact protective airway reflexes (swallow, cough, and gag).
3. The patient should have the ability to talk and sit up unaided (if age appropriate). Alternatively, for very young or intellectually disabled children, the goal is to return to their pre-sedation level of responsiveness.
4. Ensure ability to maintain adequate hydration (i.e., the patient can tolerate enteral fluids).

#### F. Examples of Sedation Protocols (Table 6.9 and Table EC 6.A)

#### BOX 6.1

#### PROPERTIES OF COMMON SEDATIVE-HYPNOTIC AGENTS

##### Sedating Antihistamines (Diphenhydramine, Hydroxyzine)

- Mild sedative-hypnotics with antiemetic and antipruritic properties; used for sedation and treatment of opioid side effects
- No anxiolytic or analgesic effects

##### Barbiturates

- Contraindicated in patients with porphyria
- Suitable only for nonpainful procedures
- Not reversible with flumazenil
- Narrower margin of safety than benzodiazepines
- No anxiolytic or analgesic effects

**BOX 6.1—cont'd****Benzodiazepines**

- Reversible with flumazenil
- Anxiolytic effects; no analgesic effects

**Opioids**

- Reversible with naloxone
- Analgesic effects; no anxiolytic effects

**Ketamine<sup>1,10–13</sup>**

- Causes potent dissociative amnesia and analgesia
- Nystagmus indicates likely therapeutic effect
- Vocalizations/movement may occur even with adequate sedation
- Onset: IV, 0.5–2 min; IM, 5–10 min; PO/PR, 20–45 min
- Duration: IV, 20–60 min; IM, 30–90 min; PO/PR, 60–120+ min
- **CNS effects:** Emergence delirium with auditory, visual, and tactile hallucinations
- **Cardiovascular effects:** Inhibits catecholamine reuptake, thereby causing increased HR, BP, SVR, and PVR. Rarely causes hemodynamic instability; however, in catecholamine-deplete patients (e.g., shock) it can cause direct myocardial depression and hypotension.
- **Respiratory effects:** Bronchodilation (useful in asthmatics), increased secretions (can result in laryngospasm), maintenance of ventilatory response to hypoxia, relative maintenance of airway reflexes
- **Other effects:** Increased muscle tone, myoclonic jerks, nausea, emesis
- **Contraindications:** Hypertension and preexisting psychotic disorders. Controversy exists on its safety in patients with elevated ICP or IOP. Evidence suggesting ketamine elevates intracranial pressure or causes harm in these patients is limited.

**Propofol**

- For deep sedation or general anesthesia
- Administered as single or multiple IV boluses +/- infusion
- Rapid onset and brief recovery (5–15 min) with bolus administration
- Can have antiemetic and euphoric effects
- Caution: Respiratory depression, apnea, hypotension
- Anxiolytic; no analgesic effects

**Dexmedetomidine**

- Give IV load over 10 min, followed by infusion.
- Dexmedetomidine can also be given intranasally. It will take 30–60 min to attain natural sleep, and patients will briefly awaken with stimulation.
- Rapid onset and brief recovery (5–15 min)
- Does not cause respiratory depression or apnea. Can cause hypotension and bradycardia, especially when IV load given too quickly.
- Anxiolytic and analgesic effects
- Increased cost compared with other medications

**Nitrous Oxide**

- Inhaled gas delivered as a mixture with oxygen
- Amnestic, anxiolytic, and analgesic effects

**BOX 6.1—cont'd**

- Extremely rapid onset and recovery
- Due to risk for delivery of hypoxic gas mixture, avoid concentrations higher than 70% (30% oxygen)
- Must be given in combination with other sedative drugs for more painful procedures

*BP*, Blood pressure; *CNS*, central nervous system; *HR*, heart rate; *ICP*, intracranial pressure; *IM*, intramuscular; *IOP*, intraocular pressure; *IV*, intravenous; *PO*, oral; *PR*, rectal; *PVR*, pulmonary vascular resistance; *SVR*, systemic vascular resistance.

Data from Yaster M, Cote C, Krane E, et al. *Pediatric Pain Management and Sedation Handbook*. St. Louis: Mosby; 1997:376–382; St Germain BA. The management of pain in the emergency department. *Pediatr Clin North Am*. 2000;47:651–679; and Cote CJ, Lerman J, Todres ID, et al. *A Practice of Anesthesia for Infants and Children*. Philadelphia: WB Saunders; 2001.

**TABLE EC 6.A****SUGGESTED ANALGESIA AND SEDATION PROTOCOLS**

Pain Threshold	Procedure	Suggested Choices
Nonpainful	CT/MRI/EEG/ECHO	Midazolam <sup>a</sup>
Mild	Phlebotomy/IV	EMLA
	LP	EMLA ( $\pm$ midazolam), lidocaine
	Pelvic exam	Midazolam
	Minor laceration, well vascularized	LET
	Minor laceration, not well vascularized	Lidocaine
Moderate	BM aspiration	EMLA ( $\pm$ midazolam)
	Arthrocentesis	Lidocaine (local) for cooperative child or ketamine for uncooperative child
	Fracture reduction	Ketamine
	Major laceration	Ketamine or fentanyl + midazolam
	Burn debridement	Ketamine or fentanyl + midazolam
Severe	Long procedures (>30 min)	Consider general anesthesia
	Fracture reduction	Ketamine
	Long procedures (>30 min)	Consider general anesthesia

<sup>a</sup>Consult with neurologist prior to administering a benzodiazepine for sedation during EEG.

*BM*, Bone marrow; *CT*, computed tomography; *ECHO*, echocardiogram; *EEG*, electroencephalogram; *EMLA*, eutectic mixture of local anesthetics; *LP*, lumbar puncture; *LET*, lidocaine, epinephrine, tetracaine; *MRI*, magnetic resonance imaging.

Modified from Yaster M, Cote C, Krane E, et al. *Pediatric Pain Management and Sedation Handbook*. St. Louis: Mosby; 1997:551–552.

**TABLE 6.8****COMMONLY USED BENZODIAZEPINES<sup>a</sup> AND BARBITURATES<sup>1,4</sup>**

Drug Class	Duration of Action	Drug	Route	Onset (min)	Duration (hr)	Comments
Benzodiazepines	Short	Midazolam (Versed)	IV	1–3	1–2	Has rapid and predictable onset of action, short recovery time
			IM/IN	5–10		Causes amnesia
			PO/PR	10–30		Results in mild depression of hypoxic ventilatory drive
	Intermediate	Diazepam (Valium)	IV (painful)	1–3	0.25–1	Poor choice for procedural sedation
			PR	7–15	2–3	Excellent for muscle relaxation or prolonged sedation
			PO	30–60	2–3	Painful on IV injection
	Long	Lorazepam (Ativan)	IV	1–5	3–4	Poor choice for procedural sedation
			IM	10–20	3–6	Ideal for prolonged anxiolysis, seizure treatment
			PO	30–60	3–6	
Barbiturates	Short	Methohexital	PR <sup>b</sup>	5–10	1–1.5	PR form used as sedative for nonpainful procedure
	Intermediate	Pentobarbital	IV	1–10	1–4	Predictable sedation and immobility for nonpainful procedures
			IM	5–15	2–4	Minimal respiratory depression when used alone
			PO/PR	15–60	2–4	Associated with slow wake up and agitation

<sup>a</sup>Use IV solution for PO, PR, and IN administration. Rectal diazepam gel (Diastat) is also available.

<sup>b</sup>IV administration produces general anesthesia; only PR should be used for sedation.

IM, Intramuscular; IN, intranasal; IV, intravenous; min, minute; PO, by mouth; PR, per rectum.

Data from Yaster M, Cote C, Krane E, et al. *Pediatric Pain Management and Sedation Handbook*. St. Louis: Mosby; 1997:345–374; St Germain BA. The management of pain in the emergency department. *Pediatr Clin North Am*. 2000;47:651–679; and Cote CJ, Lerman J, Todres ID, et al. *A Practice of Anesthesia for Infants and Children*. Philadelphia: WB Saunders; 2001.

**BOX 6.2****NALOXONE (NARCAN) ADMINISTRATION<sup>a</sup>****Indications: Patients Requiring Naloxone (Narcan) Usually Meet All the Following Criteria**

- Shallow respirations or respiratory rate <8 breaths/min<sup>b</sup>
- Pinpoint pupils
- Unresponsive to physical stimulation

**Procedure**

1. **Stop opioid administration** (as well as other sedative drugs), assess **ABCs** (Airway, Breathing, Circulation), and **call for help**.
2. **Dilute naloxone:**
  - a. If child >40 kg: Mix 0.4 mg (1 ampule) of naloxone with 9 mL of normal saline (final concentration 0.04 mg/mL = 40 mCg/mL)
  - b. If child <40 kg: Mix 0.4 mg (1 ampule) of naloxone with 9 mL of normal saline to make a concentration of 40 mCg/mL (as above). **Then, repeat dilution** by mixing 1 mL of the 40 mCg/mL solution with 9 mL of normal saline for final concentration of 4 mCg/mL.
3. **Administer and observe response:** Administer dilute naloxone *slowly* (1–2 mCg/kg/dose IV over 2 minutes). Observe patient response.
4. **Titrate to effect:** Within 1–2 minutes, patient should open eyes and respond. If not, continue until a total dose of 10 mCg/kg is given. If no response is obtained, evaluate for other cause of sedation/respiratory depression.
5. **Discontinue naloxone administration:** Discontinue naloxone as soon as patient responds (e.g., takes deep breaths when directed).
6. **Caution:** Another dose of naloxone may be required within 30 min of first dose (duration of action of naloxone is shorter than that of most opioids).
7. **Monitor patient:** Assign a staff member to monitor sedation/respiratory status and remind patient to take deep breaths as necessary.
8. **Alternative analgesia:** Provide nonopioids for pain relief. Resume opioid administration at half the original dose when the patient is easily aroused, and respiratory rate is >9 breaths/min.

6

<sup>a</sup>Naloxone administration for patients being treated for pain. Higher doses may be necessary for patients found in the community or those with signs of cardiopulmonary failure. Please see formulary for additional dosing.

<sup>b</sup>Respiratory rates that require naloxone vary according to infant's/child's usual rate.

IV, Intravenous; kg, kilogram; mCg, microgram; mg, milligram; mL, milliliter.

Modified from McCaffery M, Pasero C. *Pain: Clinical Manual*. St. Louis: Mosby; 1999:269–270.

**TABLE 6.9****EXAMPLES OF SEDATION PROTOCOLS**

Protocol/Doses	Comments
Ketamine × 1–3 doses	Lowest rates of adverse events when ketamine used alone <sup>a</sup>
Ketamine + midazolam + atropine (“ketazolam”)	Atropine = anticholinergic Midazolam = counters emergence delirium
Ketamine × 1–3 doses	Can be given IM or IV. If giving IM, combine all 3 agents in 1 syringe (using the smallest volume possible, preferably <3 mL total).
Midazolam × 1 dose	
Atropine × 1 dose	
Midazolam + fentanyl	High likelihood of respiratory depression
Midazolam × 3 doses PRN	Give fentanyl no more frequently than every 3 min
Fentanyl × 3 doses PRN	Risk of rigid chest—give no faster than 1 mCg/kg/min

<sup>a</sup>Green, SM, Roback MG, Krauss B, et al. Predictors of emesis and recovery agitation with emergency department ketamine sedation: an individual-patient data meta-analysis of 8,282 children. *Ann Emerg Med.* 2009;54:171–180.

IM, Intramuscular; IV, intravenous; mCg, microgram; PRN, as needed.

Modified from Yaster M, Cote C, Krane E, et al. *Pediatric Pain Management and Sedation Handbook*. St. Louis: Mosby; 1997.

**VI. WEB RESOURCES**

- International Association for the Study of Pain: <http://childpain.org/>
- American Pain Society: <http://www.ampainsoc.org/>
- American Society of Anesthesiologists: <http://www.asahq.org/>

**REFERENCES**

A complete list of references can be found online at [www.expertconsult.com](http://www.expertconsult.com).

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# Chapter 7

## Cardiology

*Aoibhinn Nyhan, MD*

 See additional content on Expert Consult

### I. PHYSICAL EXAMINATION

#### A. Heart Rate

Refer to the inside front cover for normal heart rate (HR) by age.

#### B. Blood Pressure

##### 1. Blood pressure (BP):

See Tables 7.1 and 7.2 for normal BP values (systolic blood pressure [SBP], diastolic blood pressure [DBP]) by age.<sup>1,2</sup>

##### 2. Mean arterial pressure (MAP)

- MAP = diastolic pressure + (pulse pressure/3) OR 1/3 systolic pressure + 2/3 diastolic pressure.
- Preterm infants and newborns: Normal MAP = gestational age in weeks + 5.

##### 3. Abnormalities in BP

- Four-limb BP measurements can be used to assess for coarctation of the aorta.
- Pulsus paradoxus: Exaggeration of the normal drop in SBP with inspiration. Determine SBP at the end of exhalation and during inhalation; difference >10 mmHg consider pericardial effusion, tamponade, pericarditis, severe asthma, or restrictive cardiomyopathies.

##### 4. Hypertension (HTN)

- See Chapter 1 for management of acute HTN.
- See Chapter 19 for screening, work-up, and management of chronic HTN.

#### C. Heart Sounds

- S<sub>1</sub>:** Associated with closure of mitral and tricuspid valves; heard best at the apex or left lower sternal border (LLSB).
- S<sub>2</sub>:** Associated with closure of pulmonary and aortic valves; heard best at the left upper sternal border (LUSB) and has normal physiologic splitting that increases with inspiration.
- S<sub>3</sub>:** Heard best at the apex or LLSB.
- S<sub>4</sub>:** Heard at the apex.

#### D. Systolic and Diastolic Sounds

See Box 7.1 for abnormal heart sounds.<sup>3</sup>

#### E. Murmurs<sup>4</sup>

Clinical characteristics are summarized in Table 7.3.<sup>3</sup>

**TABLE 7.1****BLOOD PRESSURE LEVELS FOR THE 50TH, 90TH, 95TH, AND 99TH PERCENTILES OF BLOOD PRESSURE FOR GIRLS AGED 1–17 YEARS BY PERCENTILES OF HEIGHT**

Age (years)	BP Percentile	SBP (mmHg) Height Percentile or Measured Height							DBP (mmHg) Height Percentile or Measured Height						
		5%	10%	25%	50%	75%	90%	95%	5%	10%	25%	50%	75%	90%	95%
1	Height (in)	29.7	30.2	30.9	31.8	32.7	33.4	33.9	29.7	30.2	30.9	31.8	32.7	33.4	33.9
	Height (cm)	75.4	76.6	78.6	80.8	83	84.9	86.1	75.4	76.6	78.6	80.8	83	84.9	86.1
	50th	84	85	86	86	87	88	88	41	42	42	43	44	45	46
	90th	98	99	99	100	101	102	102	54	55	56	56	57	58	58
	95th	101	102	102	103	104	105	105	59	59	60	60	61	62	62
	95th + 12 mmHg	113	114	114	115	116	117	117	71	71	72	72	73	74	74
2	Height (in)	33.4	34	34.9	35.9	36.9	37.8	38.4	33.4	34	34.9	35.9	36.9	37.8	38.4
	Height (cm)	84.9	86.3	88.6	91.1	93.7	96	97.4	84.9	86.3	88.6	91.1	93.7	96	97.4
	50th	87	87	88	89	90	91	91	45	46	47	48	49	50	51
	90th	101	101	102	103	104	105	106	58	58	59	60	61	62	62
	95th	104	105	106	106	107	108	109	62	63	63	64	65	66	66
	95th + 12 mmHg	116	117	118	118	119	120	121	74	75	75	76	77	78	78
3	Height (in)	35.8	36.4	37.3	38.4	39.6	40.6	41.2	35.8	36.4	37.3	38.4	39.6	40.6	41.2
	Height (cm)	91	92.4	94.9	97.6	100.5	103.1	104.6	91	92.4	94.9	97.6	100.5	103.1	104.6
	50th	88	89	89	90	91	92	93	48	48	49	50	51	53	53
	90th	102	103	104	104	105	106	107	60	61	61	62	63	64	65
	95th	106	106	107	108	109	110	110	64	65	65	66	67	68	69
	95th + 12 mmHg	118	118	119	120	121	122	122	76	77	77	78	79	80	81

*Continued*

TABLE 7.1—CONT'D

Age (years)	BP Percentile	SBP (mmHg) Height Percentile or Measured Height							DBP (mmHg) Height Percentile or Measured Height						
		5%	10%	25%	50%	75%	90%	95%	5%	10%	25%	50%	75%	90%	95%
4	Height (in)	38.3	38.9	39.9	41.1	42.4	43.5	44.2	38.3	38.9	39.9	41.1	42.4	43.5	44.2
	Height (cm)	97.2	98.8	101.4	104.5	107.6	110.5	112.2	97.2	98.8	101.4	104.5	107.6	110.5	112.2
	50th	89	90	91	92	93	94	94	50	51	51	53	54	55	55
	90th	103	104	105	106	107	108	108	62	63	64	65	66	67	67
	95th	107	108	109	109	110	111	112	66	67	68	69	70	70	71
	95th + 12 mmHg	119	120	121	121	122	123	124	78	79	80	81	82	82	83
5	Height (in)	40.8	41.5	42.6	43.9	45.2	46.5	47.3	40.8	41.5	42.6	43.9	45.2	46.5	47.3
	Height (cm)	103.6	105.3	108.2	111.5	114.9	118.1	120	103.6	105.3	108.2	111.5	114.9	118.1	120
	50th	90	91	92	93	94	95	96	52	52	53	55	56	57	57
	90th	104	105	106	107	108	109	110	64	65	66	67	68	69	70
	95th	108	109	109	110	111	112	113	68	69	70	71	72	73	73
	95th + 12 mmHg	120	121	121	122	123	124	125	80	81	82	83	84	85	85
6	Height (in)	43.3	44	45.2	46.6	48.1	49.4	50.3	43.4	44	45.2	46.6	48.1	49.4	50.3
	Height (cm)	110	111.8	114.9	118.4	122.1	125.6	127.7	110	111.8	114.9	118.4	122.1	125.6	127.7
	50th	92	92	93	94	96	97	97	54	54	55	56	57	58	59
	90th	105	106	107	108	109	110	111	67	67	68	69	70	71	71
	95th	109	109	110	111	112	113	114	70	71	72	72	73	74	74
	95th + 12 mmHg	121	121	122	123	124	125	126	82	83	84	84	85	86	86
7	Height (in)	45.6	46.4	47.7	49.2	50.7	52.1	53	45.6	46.4	47.7	49.2	50.7	52.1	53
	Height (cm)	115.9	117.8	121.1	124.9	128.8	132.5	134.7	115.9	117.8	121.1	124.9	128.8	132.5	134.7
	50th	92	93	94	95	97	98	99	55	55	56	57	58	59	60
	90th	106	106	107	109	110	111	112	68	68	69	70	71	72	72
	95th	109	110	111	112	113	114	115	72	72	73	73	74	74	75
	95th + 12 mmHg	121	122	123	124	125	126	127	84	84	85	85	86	86	87

Continued

TABLE 7.1—CONT'D

Age (years)	BP Percentile	SBP (mmHg) Height Percentile or Measured Height							DBP (mmHg) Height Percentile or Measured Height						
		5%	10%	25%	50%	75%	90%	95%	5%	10%	25%	50%	75%	90%	95%
8	Height (in)	47.6	48.4	49.8	51.4	53	54.5	55.5	47.6	48.4	49.8	51.4	53	54.5	55.5
	Height (cm)	121	123	126.5	130.6	134.7	138.5	140.9	121	123	126.5	130.6	134.7	138.5	140.9
	50th	93	94	95	97	98	99	100	56	56	57	59	60	61	61
	90th	107	107	108	110	111	112	113	69	70	71	72	72	73	73
	95th	110	111	112	113	115	116	117	72	73	74	74	75	75	75
	95th + 12 mmHg	122	123	124	125	127	128	129	84	85	86	87	87	87	87
9	Height (in)	49.3	50.2	51.7	53.4	55.1	56.7	57.7	49.3	50.2	51.7	53.4	55.1	56.7	57.7
	Height (cm)	125.3	127.6	131.3	135.6	140.1	144.1	146.6	125.3	127.6	131.3	135.6	140.1	144.1	146.6
	50th	95	95	97	98	99	100	101	57	58	59	60	60	61	61
	90th	108	108	109	111	112	113	114	71	71	72	73	73	73	73
	95th	112	112	113	114	116	117	118	74	74	75	75	75	75	75
	95th + 12 mmHg	124	124	125	126	128	129	130	86	86	87	87	87	87	87
10	Height (in)	51.1	52	53.7	55.5	57.4	59.1	60.2	51.1	52	53.7	55.5	57.4	59.1	60.2
	Height (cm)	129.7	132.2	136.3	141	145.8	150.2	152.8	129.7	132.2	136.3	141	145.8	150.2	152.8
	50th	96	97	98	99	101	102	103	58	59	59	60	61	61	62
	90th	109	110	111	112	113	115	116	72	73	73	73	73	73	73
	95th	113	114	114	116	117	119	120	75	75	76	76	76	76	76
	95th + 12 mmHg	125	126	126	128	129	131	132	87	87	88	88	88	88	88
11	Height (in)	53.4	54.5	56.2	58.2	60.2	61.9	63	53.4	54.5	56.2	58.2	60.2	61.9	63
	Height (cm)	135.6	138.3	142.8	147.8	152.8	157.3	160	135.6	138.3	142.8	147.8	152.8	157.3	160
	50th	98	99	101	102	104	105	106	60	60	60	61	62	63	64
	90th	111	112	113	114	116	118	120	74	74	74	74	74	75	75
	95th	115	116	117	118	120	123	124	76	77	77	77	77	77	77
	95th + 12 mmHg	127	128	129	130	132	135	136	88	89	89	89	89	89	89

Continued

TABLE 7.1—CONT'D

Age (years)	BP Percentile	SBP (mmHg) Height Percentile or Measured Height							DBP (mmHg) Height Percentile or Measured Height						
		5%	10%	25%	50%	75%	90%	95%	5%	10%	25%	50%	75%	90%	95%
12	Height (in)	56.2	57.3	59	60.9	62.8	64.5	65.5	56.2	57.3	59	60.9	62.8	64.5	65.5
	Height (cm)	142.8	145.5	149.9	154.8	159.6	163.8	166.4	142.8	145.5	149.9	154.8	159.6	163.8	166.4
	50th	102	102	104	105	107	108	108	61	61	61	62	64	65	65
	90th	114	115	116	118	120	122	122	75	75	75	75	76	76	76
	95th	118	119	120	122	124	125	126	78	78	78	78	79	79	79
	95th + 12 mmHg	130	131	132	134	136	137	138	90	90	90	90	91	91	91
13	Height (in)	58.3	59.3	60.9	62.7	64.5	66.1	67	58.3	59.3	60.9	62.7	64.5	66.1	67
	Height (cm)	148.1	150.6	154.7	159.2	163.7	167.8	170.2	148.1	150.6	154.7	159.2	163.7	167.8	170.2
	50th	104	105	106	107	108	108	109	62	62	63	64	65	65	66
	90th	116	117	119	121	122	123	123	75	75	75	76	76	76	76
	95th	121	122	123	124	126	126	127	79	79	79	79	80	80	81
	95th + 12 mmHg	133	134	135	136	138	138	139	91	91	91	91	92	92	93
14	Height (in)	59.3	60.2	61.8	63.5	65.2	66.8	67.7	59.3	60.2	61.8	63.5	65.2	66.8	67.7
	Height (cm)	150.6	153	156.9	161.3	165.7	169.7	172.1	150.6	153	156.9	161.3	165.7	169.7	172.1
	50th	105	106	107	108	109	109	109	63	63	64	65	66	66	66
	90th	118	118	120	122	123	123	123	76	76	76	76	77	77	77
	95th	123	123	124	125	126	127	127	80	80	80	80	81	81	82
	95th + 12 mmHg	135	135	136	137	138	139	139	92	92	92	92	93	93	94
15	Height (in)	59.7	60.6	62.2	63.9	65.6	67.2	68.1	59.7	60.6	62.2	63.9	65.6	67.2	68.1
	Height (cm)	151.7	154	157.9	162.3	166.7	170.6	173	151.7	154	157.9	162.3	166.7	170.6	173
	50th	105	106	107	108	109	109	109	64	64	64	65	66	67	67
	90th	118	119	121	122	123	123	124	76	76	76	77	77	78	78
	95th	124	124	125	126	127	127	128	80	80	80	81	82	82	82
	95th + 12 mmHg	136	136	137	138	139	139	140	92	92	92	93	94	94	94

Continued

TABLE 7.1—CONT'D

Age (years)	BP Percentile	SBP (mmHg) Height Percentile or Measured Height							DBP (mmHg) Height Percentile or Measured Height						
		5%	10%	25%	50%	75%	90%	95%	5%	10%	25%	50%	75%	90%	95%
16	Height (in)	59.9	60.8	62.4	64.1	65.8	67.3	68.3	59.9	60.8	62.4	64.1	65.8	67.3	68.3
	Height (cm)	152.1	154.5	158.4	162.8	167.1	171.1	173.4	152.1	154.5	158.4	162.8	167.1	171.1	173.4
	50th	106	107	108	109	109	110	110	64	64	65	66	66	67	67
	90th	119	120	122	123	124	124	124	76	76	76	77	78	78	78
	95th	124	125	125	127	127	128	128	80	80	80	81	82	82	82
	95th + 12 mmHg	136	137	137	139	139	140	140	92	92	92	93	94	94	94
17	Height (in)	60.0	60.9	62.5	64.2	65.9	67.4	68.4	60.0	60.9	62.5	64.2	65.9	67.4	68.4
	Height (cm)	152.4	154.7	158.7	163.0	167.4	171.3	173.7	152.4	154.7	158.7	163.0	167.4	171.3	173.7
	50th	107	108	109	110	110	110	111	64	64	65	66	66	66	67
	90th	120	121	123	124	124	125	125	76	76	77	77	78	78	78
	95th	125	125	126	127	128	128	128	80	80	80	81	82	82	82
	95th + 12 mmHg	137	137	138	139	140	140	140	92	92	92	93	94	94	94

BP, Blood pressure; DBP, diastolic blood pressure; SBP, systolic blood pressure.

From Clinical Practice Guideline for Screening and Management of High Blood Pressure in Children and Adolescents. *Pediatrics*. 2017;e20171904; <https://doi.org/10.1542/peds.2017-1904>.

*Continued***TABLE 7.2****BLOOD PRESSURE LEVELS FOR THE 50TH, 90TH, 95TH, AND 99TH PERCENTILES OF BLOOD PRESSURE FOR BOYS AGED 1–17 YEARS BY PERCENTILES OF HEIGHT**

Age (years)	BP Percentile	SBP (mmHg) Height Percentile or Measured Height							DBP (mmHg) Height Percentile or Measured Height						
		5%	10%	25%	50%	75%	90%	95%	5%	10%	25%	50%	75%	90%	95%
1	Height (in)	30.4	30.8	31.6	32.4	33.3	34.1	34.6	30.4	30.8	31.6	32.4	33.3	34.1	34.6
	Height (cm)	77.2	78.3	80.2	82.4	84.6	86.7	87.9	77.2	78.3	80.2	82.4	84.6	86.7	87.9
	50th	85	85	86	86	87	88	88	40	40	40	41	41	42	42
	90th	98	99	99	100	100	101	101	52	52	53	53	54	54	54
	95th	102	102	103	103	104	105	105	54	54	55	55	56	57	57
	95th + 12 mmHg	114	114	115	115	116	117	117	66	66	67	67	68	69	69
2	Height (in)	33.9	34.4	35.3	36.3	37.3	38.2	38.8	33.9	34.4	35.3	36.3	37.3	38.2	38.8
	Height (cm)	86.1	87.4	89.6	92.1	94.7	97.1	98.5	86.1	87.4	89.6	92.1	94.7	97.1	98.5
	50th	87	87	88	89	89	90	91	43	43	44	44	45	46	46
	90th	100	100	101	102	103	103	104	55	55	56	56	57	58	58
	95th	104	105	105	106	107	107	108	57	58	58	59	60	61	61
	95th + 12 mmHg	116	117	117	118	119	119	120	69	70	70	71	72	73	73
3	Height (in)	36.4	37	37.9	39	40.1	41.1	41.7	36.4	37	37.9	39	40.1	41.1	41.7
	Height (cm)	92.5	93.9	96.3	99	101.8	104.3	105.8	92.5	93.9	96.3	99	101.8	104.3	105.8
	50th	88	89	89	90	91	92	92	45	46	46	47	48	49	49
	90th	101	102	102	103	104	105	105	58	58	59	59	60	61	61
	95th	106	106	107	107	108	109	109	60	61	61	62	63	64	64
	95th + 12 mmHg	118	118	119	119	120	121	121	72	73	73	74	75	76	76

TABLE 7.2—CONT'D

Age (years)	BP Percentile	SBP (mmHg) Height Percentile or Measured Height							DBP (mmHg) Height Percentile or Measured Height						
		5%	10%	25%	50%	75%	90%	95%	5%	10%	25%	50%	75%	90%	95%
4	Height (in)	38.8	39.4	40.5	41.7	42.9	43.9	44.5	38.8	39.4	40.5	41.7	42.9	43.9	44.5
	Height (cm)	98.5	100.2	102.9	105.9	108.9	111.5	113.2	98.5	100.2	102.9	105.9	108.9	111.5	113.2
	50th	90	90	91	92	93	94	94	48	49	49	50	51	52	52
	90th	102	103	104	105	105	106	107	60	61	62	62	63	64	64
	95th	107	107	108	108	109	110	110	63	64	65	66	67	67	68
	95th + 12 mmHg	119	119	120	120	121	122	122	75	76	77	78	79	79	80
5	Height (in)	41.1	41.8	43.0	44.3	45.5	46.7	47.4	41.1	41.8	43.0	44.3	45.5	46.7	47.4
	Height (cm)	104.4	106.2	109.1	112.4	115.7	118.6	120.3	104.4	106.2	109.1	112.4	115.7	118.6	120.3
	50th	91	92	93	94	95	96	96	51	51	52	53	54	55	55
	90th	103	104	105	106	107	108	108	63	64	65	65	66	67	67
	95th	107	108	109	109	110	111	112	66	67	68	69	70	70	71
	95th + 12 mmHg	119	120	121	121	122	123	124	78	79	80	81	82	82	83
6	Height (in)	43.4	44.2	45.4	46.8	48.2	49.4	50.2	43.4	44.2	45.4	46.8	48.2	49.4	50.2
	Height (cm)	110.3	112.2	115.3	118.9	122.4	125.6	127.5	110.3	112.2	115.3	118.9	122.4	125.6	127.5
	50th	93	93	94	95	96	97	98	54	54	55	56	57	57	58
	90th	105	105	106	107	109	110	110	66	66	67	68	68	69	69
	95th	108	109	110	111	112	113	114	69	70	70	71	72	72	73
	95th + 12 mmHg	120	121	122	123	124	125	126	81	82	83	84	84	84	85
7	Height (in)	45.7	46.5	47.8	49.3	50.8	52.1	52.9	45.7	46.5	47.8	49.3	50.8	52.1	52.9
	Height (cm)	116.1	118	121.4	125.1	128.9	132.4	134.5	116.1	118	121.4	125.1	128.9	132.4	134.5
	50th	94	94	95	97	98	98	99	56	56	57	58	58	59	59
	90th	106	107	108	109	110	111	111	68	68	69	70	70	71	71
	95th	110	110	111	112	114	115	116	71	71	72	73	73	74	74
	95th + 12 mmHg	122	122	123	124	126	127	128	83	83	84	85	85	86	86

Continued

TABLE 7.2—CONT'D

Age (years)	BP Percentile	SBP (mmHg) Height Percentile or Measured Height							DBP (mmHg) Height Percentile or Measured Height						
		5%	10%	25%	50%	75%	90%	95%	5%	10%	25%	50%	75%	90%	95%
8	Height (in)	47.8	48.6	50	51.6	53.2	54.6	55.5	47.8	48.6	50	51.6	53.2	54.6	55.5
	Height (cm)	121.4	123.5	127	131	135.1	138.8	141	121.4	123.5	127	131	135.1	138.8	141
	50th	95	96	97	98	99	99	100	57	57	58	59	59	60	60
	90th	107	108	109	110	111	112	112	69	70	70	71	72	72	73
	95th	111	112	112	114	115	116	117	72	73	73	74	75	75	75
	95th + 12 mmHg	123	124	124	126	127	128	129	84	85	85	86	87	87	87
9	Height (in)	49.6	50.5	52	53.7	55.4	56.9	57.9	49.6	50.5	52	53.7	55.4	56.9	57.9
	Height (cm)	126	128.3	132.1	136.3	140.7	144.7	147.1	126	128.3	132.1	136.3	140.7	144.7	147.1
	50th	96	97	98	99	100	101	101	57	58	59	60	61	62	62
	90th	107	108	109	110	112	113	114	70	71	72	73	74	74	74
	95th	112	112	113	115	116	118	119	74	74	75	76	76	77	77
	95th + 12 mmHg	124	124	125	127	128	130	131	86	86	87	88	88	89	89
10	Height (in)	51.3	52.2	53.8	55.6	57.4	59.1	60.1	51.3	52.2	53.8	55.6	57.4	59.1	60.1
	Height (cm)	130.2	132.7	136.7	141.3	145.9	150.1	152.7	130.2	132.7	136.7	141.3	145.9	150.1	152.7
	50th	97	98	99	100	101	102	103	59	60	61	62	63	63	64
	90th	108	109	111	112	113	115	116	72	73	74	74	75	75	76
	95th	112	113	114	116	118	120	121	76	76	77	77	78	78	78
	95th + 12 mmHg	124	125	126	128	130	132	133	88	88	89	89	90	90	90
11	Height (in)	53	54	55.7	57.6	59.6	61.3	62.4	53	54	55.7	57.6	59.6	61.3	62.4
	Height (cm)	134.7	137.3	141.5	146.4	151.3	155.8	158.6	134.7	137.3	141.5	146.4	151.3	155.8	158.6
	50th	99	99	101	102	103	104	106	61	61	62	63	63	63	63
	90th	110	111	112	114	116	117	118	74	74	75	75	75	76	76
	95th	114	114	116	118	120	123	124	77	78	78	78	78	78	78
	95th + 12 mmHg	126	126	128	130	132	135	136	89	90	90	90	90	90	90

Continued

**TABLE 7.2—CONT'D**

Age (years)	BP Percentile	SBP (mmHg) Height Percentile or Measured Height							DBP (mmHg) Height Percentile or Measured Height						
		5%	10%	25%	50%	75%	90%	95%	5%	10%	25%	50%	75%	90%	95%
12	Height (in)	55.2	56.3	58.1	60.1	62.2	64	65.2	55.2	56.3	58.1	60.1	62.2	64	65.2
	Height (cm)	140.3	143	147.5	152.7	157.9	162.6	165.5	140.3	143	147.5	152.7	157.9	162.6	165.5
	50th	101	101	102	104	106	108	109	61	62	62	62	62	63	63
	90th	113	114	115	117	119	121	122	75	75	75	75	75	76	76
	95th	116	117	118	121	124	126	128	78	78	78	78	78	79	79
	95th + 12 mmHg	128	129	130	133	136	138	140	90	90	90	90	90	91	91
13	Height (in)	57.9	59.1	61	63.1	65.2	67.1	68.3	57.9	59.1	61	63.1	65.2	67.1	68.3
	Height (cm)	147	150	154.9	160.3	165.7	170.5	173.4	147	150	154.9	160.3	165.7	170.5	173.4
	50th	103	104	105	108	110	111	112	61	60	61	62	63	64	65
	90th	115	116	118	121	124	126	126	74	74	74	75	76	77	77
	95th	119	120	122	125	128	130	131	78	78	78	78	80	81	81
	95th + 12 mmHg	131	132	134	137	140	142	143	90	90	90	90	92	93	93
14	Height (in)	60.6	61.8	63.8	65.9	68.0	69.8	70.9	60.6	61.8	63.8	65.9	68.0	69.8	70.9
	Height (cm)	153.8	156.9	162	167.5	172.7	177.4	180.1	153.8	156.9	162	167.5	172.7	177.4	180.1
	50th	105	106	109	111	112	113	113	60	60	62	64	65	66	67
	90th	119	120	123	126	127	128	129	74	74	75	77	78	79	80
	95th	123	125	127	130	132	133	134	77	78	79	81	82	83	84
	95th + 12 mmHg	135	137	139	142	144	145	146	89	90	91	93	94	95	96

*Continued*

**TABLE 7.2—CONT'D**

Age (years)	BP Percentile	SBP (mmHg) Height Percentile or Measured Height							DBP (mmHg) Height Percentile or Measured Height						
		5%	10%	25%	50%	75%	90%	95%	5%	10%	25%	50%	75%	90%	95%
15	Height (in)	62.6	63.8	65.7	67.8	69.8	71.5	72.5	62.6	63.8	65.7	67.8	69.8	71.5	72.5
	Height (cm)	159	162	166.9	172.2	177.2	181.6	184.2	159	162	166.9	172.2	177.2	181.6	184.2
	50th	108	110	112	113	114	114	114	61	62	64	65	66	67	68
	90th	123	124	126	128	129	130	130	75	76	78	79	80	81	81
	95th	127	129	131	132	134	135	135	78	79	81	83	84	85	85
	95th + 12 mmHg	139	141	143	144	146	147	147	90	91	93	95	96	97	97
16	Height (in)	63.8	64.9	66.8	68.8	70.7	72.4	73.4	63.8	64.9	66.8	70.7	70.7	72.4	73.4
	Height (cm)	162.1	165	169.6	174.6	179.5	183.8	186.4	162.1	165	169.6	174.6	179.5	183.8	186.4
	50th	111	112	114	115	115	116	116	63	64	66	67	68	69	69
	90th	126	127	128	129	131	131	132	77	78	79	80	81	82	82
	95th	130	131	133	134	135	136	137	80	81	83	84	85	86	86
	95th + 12 mmHg	142	143	145	146	147	148	149	92	93	95	96	97	98	98
17	Height (in)	64.5	65.5	67.3	69.2	71.1	72.8	73.8	64.5	65.5	67.3	69.2	71.1	72.8	73.8
	Height (cm)	163.8	166.5	170.9	175.8	180.7	184.9	187.5	163.8	166.5	170.9	175.8	180.7	184.9	187.5
	50th	114	115	116	117	117	118	118	65	66	67	68	69	70	70
	90th	128	129	130	131	132	133	134	78	79	80	81	82	82	83
	95th	132	133	134	135	137	138	138	81	82	84	85	86	86	87
	95th + 12 mmHg	144	145	146	147	149	150	150	93	94	96	97	98	98	99

BP, Blood pressure; DBP, diastolic blood pressure; SBP, systolic blood pressure.

From Clinical Practice Guideline for Screening and Management of High Blood Pressure in Children and Adolescents. *Pediatrics*. 2017;e20171904; <https://doi.org/10.1542/peds.2017-1904>.

**BOX 7.1****SUMMARY OF ABNORMAL HEART SOUNDS**

- **Widely split S<sub>1</sub>:** Ebstein anomaly, RBBB
- **Widely split and fixed S<sub>2</sub>:** Right ventricular volume overload (e.g., ASD, PAPVR), pressure overload (e.g., PS), electrical delay in RV contraction (e.g., RBBB), early aortic closure (e.g., MR), occasionally heard in normal child
- **Narrowly split S<sub>2</sub>:** Pulmonary hypertension, AS, delay in LV contraction (e.g., LBBB), occasionally heard in normal child
- **Single S<sub>2</sub>:** Pulmonary hypertension, one semilunar valve (e.g., pulmonary atresia, aortic atresia, truncus arteriosus), P2 not audible (e.g., TGA, TOF, severe PS), severe AS, occasionally heard in normal child
- **Paradoxically split S<sub>2</sub>:** Severe AS, LBBB, Wolff-Parkinson-White syndrome (type B)
- **Abnormal intensity of P2:** Increased P2 (e.g., pulmonary hypertension), decreased P2 (e.g., severe PS, TOF, TS)
- **S<sub>3</sub>:** Occasionally heard in healthy children or adults or may indicate dilated ventricles (e.g., large VSD, CHF)
- **S<sub>4</sub>:** Always pathologic, indicative of decreased ventricular compliance
- **Ejection click:** Heard with stenosis of the semilunar valves, dilated great arteries in the setting of pulmonary or systemic HTN, idiopathic dilation of the PA, TOF, persistent truncus arteriosus
- **Midsystolic click:** Heard at the apex in mitral valve prolapse
- **Diastolic opening snap:** Rare in children; associated with TS/MS

AS, Aortic stenosis; ASD, atrial septal defect; CHF, congestive heart failure; LBBB, left bundle-branch block; LV, left ventricle; MR, mitral regurgitation; MS, mitral stenosis; PA, pulmonary artery; PAPVR, partial anomalous pulmonary venous return; PS, pulmonic stenosis; RBBB, right bundle-branch block; RV, right ventricular; TGA, transposition of the great arteries; TOF, tetralogy of fallot; TS, tricuspid stenosis; VSD, ventricular septal defect.

Modified from Park MK. *Pediatric Cardiology for Practitioners*. 5th ed. St Louis: Elsevier; 2008:25.

**1. Grading of heart murmurs:** Intensified by states of higher cardiac output (e.g., anemia, anxiety, fever, exercise).<sup>3</sup>

- Grade I: Barely audible
- Grade II: Murmur softer than heart sounds, but audible
- Grade III: Murmur moderately loud, equally loud as heart sounds, not accompanied by a thrill
- Grade IV: Murmur louder than heart sounds, associated with a thrill
- Grade V: Audible with a stethoscope barely on the chest
- Grade VI: Audible with a stethoscope off the chest

**2. Benign heart murmurs<sup>4</sup>:**

- Caused by a disturbance of the laminar flow of blood; frequently produced as the diameter of the blood's pathway decreases and velocity increases.
- Present in >80% of children sometime during childhood, most commonly beginning at age 3 to 4 years.
- Accentuated in high-output states, especially with fever and anemia.
- Normal electrocardiogram (ECG) and radiographic findings.

**NOTE:** ECG and chest radiograph are not routinely used, nor are they cost-effective screening tools for distinguishing benign from pathologic murmurs.

**TABLE 7.3****COMMON INNOCENT HEART MURMURS**

Type (Timing)	Description of Murmur	Age Group
Classic vibratory murmur (Still's murmur; systolic)	Maximal at LMSB or between LLSB and apex Grade 2–3/6 in intensity Low-frequency vibratory, twanging string, groaning, squeaking, or musical	3–6 years; occasionally in infancy
Pulmonary ejection murmur (systolic)	Maximal at LUSB Early to midsystolic Grade 1–3/6 in intensity Blowing in quality	8–14 years
Pulmonary flow murmur of newborn (systolic)	Maximal at LUSB Transmits well to left and right chest, axilla, and back Grade 1–2/6 in intensity	Premature and full-term newborns Usually disappears by 3–6 months
Venous hum (continuous)	Maximal at right (or left) supraclavicular and infraclavicular areas Grade 1–3/6 in intensity Inaudible in supine position Intensity changes with rotation of head and disappears with compression of jugular vein	3–6 years
Carotid bruit (systolic)	Right supraclavicular area over carotids Grade 2–3/6 in intensity Occasional thrill over carotid	Any age

LMSB, Left lower sternal border; LMSB, left middle sternal border; LUSB, left upper sternal border

From Park MK. *Pediatric Cardiology for Practitioners*. 5th ed. St Louis: Elsevier; 2008:36.

- A murmur is more likely to be pathologic when one or more of the following are present:** Symptoms (e.g., chest pain, dyspnea with exertion, syncope with exertion); cyanosis; a systolic murmur that is loud (grade  $\geq 3/6$ ), harsh, pansystolic, or long in duration; diastolic murmur; abnormal heart sounds; presence of a click; abnormally strong or weak pulses.<sup>3,4</sup>
- Systolic and diastolic heart murmurs (Box 7.2).**

**II. ELECTROCARDIOGRAPHY****A. Basic Electrocardiography Principles**

- Lead placement (Fig. 7.1)**
- ECG complexes**
  - P wave: Represents atrial depolarization.
  - QRS complex: Represents ventricular depolarization.
  - T wave: Represents ventricular repolarization.
  - U wave: May follow the T wave and represents late phases of ventricular repolarization.
- Systematic approach for evaluating ECGs (Table 7.4 shows normal ECG parameters):<sup>3,5</sup>**
  - Rate**

**BOX 7.2****SYSTOLIC AND DIASTOLIC HEART MURMURS****RUSB**

Aortic valve stenosis (supravalvular, subvalvular)

*Aortic regurgitation***LUSB**

Pulmonary valve stenosis

Atrial septal defect

Pulmonary ejection murmur, innocent

Pulmonary flow murmur of newborn

Pulmonary artery stenosis

Aortic stenosis

Coarctation of the aorta

Patent ductus arteriosus

Partial anomalous pulmonary venous return (PAPVR)

Total anomalous pulmonary venous return (TAPVR)

*Pulmonary regurgitation***LLSB**

Ventricular septal defect, including atrioventricular septal defect

Vibratory innocent murmur (Still's murmur)

HOCM (IHSS)

Tricuspid regurgitation

Tetralogy of Fallot

*Tricuspid stenosis***Apex**

Mitral regurgitation

Vibratory innocent murmur (Still's murmur)

Mitral valve prolapse

Aortic stenosis

HOCM (IHSS)

*Mitral stenosis*Murmurs listed by the location at which they are best heard. *Diaстolic murmurs are in italics.*

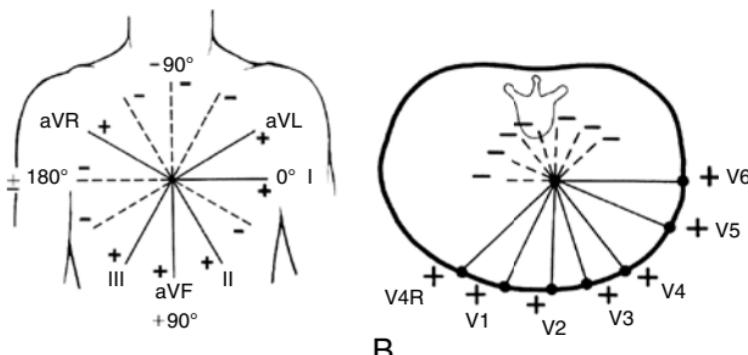
HOCM, Hypertrophic obstructive cardiomyopathy; IHSS, idiopathic hypertrophic subaortic stenosis; LLSB, left lower sternal border; LUSB, left upper sternal border; RUSB, right upper sternal border. From Park MK. *Pediatric Cardiology for Practitioners*. 5th ed. St Louis: Elsevier; 2008:30.

- (1) Standardization: Paper speed is 25 mm/sec. **One small square = 1 mm = 0.04 second. One large square = 5 mm = 0.2 second.**  
**Amplitude standard: 10 mm = 1 mV.**

- (2) Calculation: HR (beats/min) = 60 divided by the average R-R interval in seconds, or 1500 divided by the R-R interval in millimeters.

b. **Rhythm**

- (1) Sinus rhythm: Every QRS complex is preceded by a P wave, normal PR interval (although PR interval may be prolonged, as in first-degree atrioventricular [AV] block), and normal P-wave axis (upright P in leads I and aVF).

**FIGURE 7.1**

(A) Hexaxial reference system, (B) Horizontal reference system. (Modified from Park MK, Guntheroth WG. *How to Read Pediatric ECGs*. 4th ed. Philadelphia: Elsevier; 2006:3.)

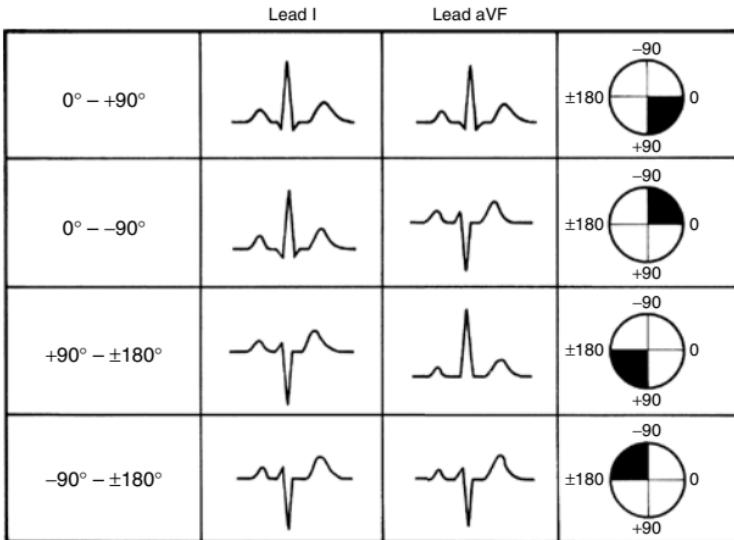
- (2) There is normal respiratory variation of the R-R interval without morphologic changes of the P wave or QRS complex.
- c. **Axis:** The direction of the QRS in leads I and aVF should be observed, the quadrant determined, and comparison made with age-matched normal values (Fig. 7.2 and Table 7.4).
- d. **Intervals** (PR, QRS, QTc)
  - (1) See Table 7.4 for normal PR and QRS intervals.
  - (2) The QTc is calculated using the Bazett formula: **QTc = QT (sec) measured / √R-R** (the average of three measurements taken from the same lead, usually lead II).
  - (3) The QT interval is measured from the beginning of the QRS complex to the end of the T wave. Divide this value by the square root of the preceding R-R interval to obtain the QTc.
  - (4) **Normal values for QTc are:**
    - (a) 0.44 second is the 97th percentile for infants 3 to 4 days old.<sup>6</sup>
    - (b) ≤0.45 second in all males aged >1 week and in prepubescent females.
    - (c) ≤0.46 second for postpubescent females.
- e. **P-wave size and shape:** A normal P wave should be <0.10 second in children and <0.08 second in infants, with an amplitude of <0.3 mV (3 mm in height, with normal standardization).
- f. **R-wave progression:** In general, there is a normal increase in R-wave size and a decrease in S-wave size from leads V<sub>1</sub> to V<sub>6</sub> (with dominant S waves in the right precordial leads and dominant R waves in the left precordial leads), representing dominance of left ventricular forces. However, newborns and infants have a normal dominance of the right ventricle.
- g. **Q waves:** Normal Q waves are usually <0.04 second in duration and <25% of the total QRS amplitude. Q waves are <5 mm deep in the left precordial leads and aVF, and ≤8 mm deep in lead III for children age <3 years.

TABLE 7.4

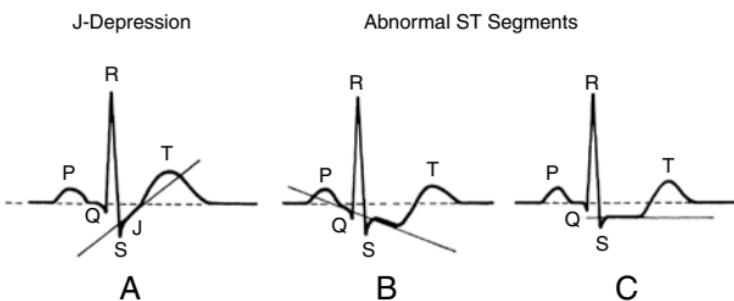
## NORMAL PEDIATRIC ELECTROCARDIOGRAM PARAMETERS

Age	Heart Rate (bpm)	QRS Axis <sup>a</sup>	PR Interval (sec) <sup>a</sup>	QRS Duration (sec) <sup>b</sup>	Lead V <sub>1</sub>			Lead V <sub>6</sub>		
					R-Wave Amplitude (mm) <sup>b</sup>	S-Wave Amplitude (mm) <sup>b</sup>	R/S Ratio	R-Wave Amplitude (mm) <sup>b</sup>	S-Wave Amplitude (mm) <sup>b</sup>	R/S Ratio
0–7 days	95–160 (125)	+30 to +180 (+110)	0.08–0.12 (0.10)	0.05 (0.07)	13.3 (25.5)	7.7 (18.8)	2.5	4.8 (11.8)	3.2 (9.6)	2.2
1–3 weeks	105–180 (145)	+30 to +180 (+110)	0.08–0.12 (0.10)	0.05 (0.07)	10.6 (20.8)	4.2 (10.8)	2.9	7.6 (16.4)	3.4 (9.8)	3.3
1–6 months	110–180 (145)	+10 to +125 (+70)	0.08–0.13 (0.11)	0.05 (0.07)	9.7 (19)	5.4 (15)	2.3	12.4 (22)	2.8 (8.3)	5.6
6–12 months	110–170 (135)	+10 to +125 (+60)	0.10–0.14 (0.12)	0.05 (0.07)	9.4 (20.3)	6.4 (18.1)	1.6	12.6 (22.7)	2.1 (7.2)	7.6
1–3 years	90–150 (120)	+10 to +125 (+60)	0.10–0.14 (0.12)	0.06 (0.07)	8.5 (18)	9 (21)	1.2	14 (23.3)	1.7 (6)	10
4–5 years	65–135 (110)	0 to +110 (+60)	0.11–0.15 (0.13)	0.07 (0.08)	7.6 (16)	11 (22.5)	0.8	15.6 (25)	1.4 (4.7)	11.2
6–8 years	60–130 (100)	-15 to +110 (+60)	0.12–0.16 (0.14)	0.07 (0.08)	6 (13)	12 (24.5)	0.6	16.3 (26)	1.1 (3.9)	13
9–11 years	60–110 (85)	-15 to +110 (+60)	0.12–0.17 (0.14)	0.07 (0.09)	5.4 (12.1)	11.9 (25.4)	0.5	16.3 (25.4)	1.0 (3.9)	14.3
12–16 years	60–110 (85)	-15 to +110 (+60)	0.12–0.17 (0.15)	0.07 (0.10)	4.1 (9.9)	10.8 (21.2)	0.5	14.3 (23)	0.8 (3.7)	14.7
>16 years	60–100 (80)	-15 to +110 (+60)	0.12–0.20 (0.15)	0.08 (0.10)	3 (9)	10 (20)	0.3	10 (20)	0.8 (3.7)	12

<sup>a</sup>Normal range and (mean).<sup>b</sup>Mean and (98th percentile).Data from Park MK. *Pediatric Cardiology for Practitioners*. 5th ed. St Louis: Elsevier; 2008; and Davignon A, et al. Normal ECG standards for infants and children. *Pediatr Cardiol*. 1979;1:123–131.

**FIGURE 7.2**

Location of quadrants of the mean QRS axis from leads I and aVF. (From Park MK, Guntheroth WG. *How to Read Pediatric ECGs*. 4th ed. Philadelphia: Elsevier; 2006:17.)

**FIGURE 7.3**

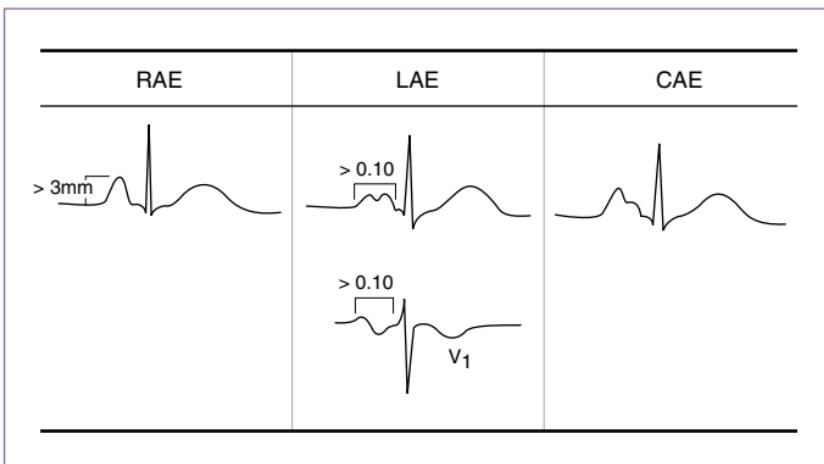
Non-pathologic (non-ischemic) and pathologic (ischemic) ST and T changes. (A) Characteristic non-ischemic ST-segment alteration called J-depression (note that ST slope is upward), B-C. Ischemic or pathologic ST-segment alterations, (B) Downward slope of ST segment, (C) Horizontal segment is sustained. (From Park MK, Guntheroth WG. *How to Read Pediatric ECGs*. 4th ed. Philadelphia: Elsevier; 2006:107.)

- h. **ST-segment** (Fig. 7.3): ST-segment elevation or depression of >1 mm in the limb leads and >2 mm in the precordial leads is consistent with myocardial ischemia or injury. **NOTE:** J-depression is an upsloping of the ST segment and a normal variant.

**TABLE 7.5****NORMAL T-WAVE AXIS**

Age	V <sub>1</sub> , V <sub>2</sub>	AVF	I, V <sub>5</sub> , V <sub>6</sub>
Birth–1 day	±	+	±
1–4 days	±	+	+
4 days to adolescent	–	+	+
Adolescent to adult	+	+	+

+, T wave positive; –, T wave negative; ±, T wave normally either positive or negative.

**FIGURE 7.4**

Criteria for Atrial Enlargement. CAE, Combined atrial enlargement; LAE, left atrial enlargement; RAE, right atrial enlargement. (From Park MK. *Pediatric Cardiology for Practitioners*. 5th ed. St Louis: Elsevier; 2008:53.)

**i. T wave:**

- (1) Inverted T waves in V<sub>1</sub> and V<sub>2</sub> can be normal in children up to adolescence (Table 7.5).
- (2) Tall, peaked T waves may be seen in hyperkalemia.
- (3) Flat or low T waves may be seen in hypokalemia, hypothyroidism, normal newborns, and myocardial/pericardial ischemia and inflammation.

**j. Hypertrophy/enlargement**

- (1) Atrial enlargement (Fig. 7.4).
- (2) Ventricular hypertrophy: Diagnosed by QRS axis, voltage, and R/S ratio (Box 7.3; see also Table 7.6).

**B. ECG Abnormalities**

1. **Nonventricular arrhythmias** (Table 7.6; Fig. 7.5)<sup>7</sup>
2. **Ventricular arrhythmias** (Table 7.7; Fig. 7.6)
3. **Nonventricular conduction disturbances** (Table 7.8; Fig. 7.7)<sup>8</sup>
4. **Ventricular conduction disturbances** (Table 7.9)

**BOX 7.3****VENTRICULAR HYPERTROPHY CRITERIA****Right Ventricular Hypertrophy (RVH) Criteria**

Must Have at Least One of the Following:

Upright T wave in lead V<sub>1</sub> after 3 days of age to adolescence

Presence of Q wave in V<sub>1</sub> (QR or QRS pattern)

Increased right and anterior QRS voltage (with normal QRS duration):

R in lead V<sub>1</sub>, >98th percentile for age

S in lead V<sub>6</sub>, >98th percentile for age

Right ventricular strain (associated with inverted T wave in V<sub>1</sub> with tall R wave)

**Left Ventricular Hypertrophy (LVH) Criteria**

Left ventricular strain (associated with inverted T wave in leads V<sub>6</sub>, I, and/or aVF)

**Supplemental Criteria**

Left axis deviation (LAD) for patient's age

Volume overload (associated with Q wave >5 mm and tall T waves in V<sub>5</sub> or V<sub>6</sub>)

Increased QRS voltage in left leads (with normal QRS duration):

R in lead V<sub>6</sub> (and I, aVL, V<sub>5</sub>), >98th percentile for age

S in lead V<sub>1</sub>, >98th percentile for age

### C. ECG Findings Secondary to Electrolyte Disturbances, Medications, and Systemic Illnesses (Table 7.10)<sup>7,9</sup>

#### D. Long QT

1. Diagnosis:
  - a. In general, QTc is similar in males and females from birth until late adolescence (0.37 to 0.44 second).
  - b. In adults, prolonged QTc is generally >0.45 second.
  - c. In ~10% of cases, patients may have a normal QTc. Patients may also have a family history of long QT associated with unexplained syncope, seizure, or cardiac arrest, without prolongation of QTc on ECG.
  - d. Treadmill exercise testing may prolong the QTc and will sometimes induce arrhythmias.
2. **Complications:** Associated with ventricular arrhythmias (torsades de pointes), syncope, and sudden death.
3. **Management:**
  - a. Congenital long QT: β-blockers and/or defibrillators; rarely requires cardiac sympathetic denervation or cardiac pacemakers.
  - b. Acquired long QT: Treatment of arrhythmias, discontinuation of precipitating drugs, and correction of metabolic abnormalities.

#### E. Hyperkalemia:

ECG changes dependent on the serum potassium (K<sup>+</sup>) level; however, the ECG may be normal with serum K<sup>+</sup> levels between 2.5 and 6 mEq/L.

1. **Serum K<sup>+</sup> <2.5 mEq/L:** Depressed ST segment, biphasic T wave.

2. **Serum K<sup>+</sup> >6 mEq/L:** Tall T wave.

**TABLE 7.6****NONVENTRICULAR ARRHYTHMIAS**

Name/Description	Cause	Treatment
<b>SINUS</b>		
<b>TACHYCARDIA</b>		
Normal sinus rhythm with HR >95th percentile for age (usually infants: <220 beats/min and children: <180 beats/min)	Hypovolemia, shock, anemia, sepsis, fever, anxiety, CHF, PE, myocardial disease, drugs (e.g., $\beta$ -agonists, albuterol, caffeine, atropine)	Address underlying cause
<b>BRADYCARDIA</b>		
Normal sinus rhythm with HR <5th percentile for age	Normal (especially in athletic individuals), increased ICP, hypoxia, hyperkalemia, hypercalcemia, vagal stimulation, hypothyroidism, hypothermia, drugs (e.g., opioids, digoxin, $\beta$ -blockers), long QT	Address underlying cause; if symptomatic, refer to inside back cover for bradycardia algorithm
<b>SUPRAVENTRICULAR<sup>a</sup></b>		
<b>PREMATURE ATRIAL CONTRACTION (PAC)</b>		
Narrow QRS complex; ectopic focus in atria with abnormal P-wave morphology	Digitalis toxicity, medications (e.g., caffeine, theophylline, sympathomimetics), normal variant	Treat digitalis toxicity; otherwise no treatment needed
<b>ATRIAL FLUTTER</b>		
Atrial rate 250–350 beats/min; characteristic saw-tooth or flutter pattern with variable ventricular response rate and normal QRS complex	Dilated atria, previous intra-atrial surgery, valvular or ischemic heart disease, idiopathic in newborns	Synchronized cardioversion or overdrive pacing; treat underlying cause
<b>ATRIAL FIBRILLATION</b>		
Irregular; atrial rate 350–600 beats/min, yielding characteristic fibrillary pattern (no discrete P waves) and irregular ventricular response rate of about 110–150 beats/min with normal QRS complex	Wolff-Parkinson-White syndrome and those listed previously for atrial flutter (except idiopathic), alcohol exposure, familial	Synchronized cardioversion; then may need anticoagulation based on stroke risk

*Continued*

**TABLE 7.6—CONT'D**

Name/Description	Cause	Treatment
<b>SVT</b>		
Sudden run of three or more consecutive premature supraventricular beats at >220 beats/min (infant) or >180 beats/min (child), with narrow QRS complex and absent/abnormal P wave; either sustained (>30 sec) or non-sustained	Most commonly idiopathic but may be seen in congenital heart disease (e.g., Ebstein anomaly, transposition)	Vagal maneuvers, adenosine; if unstable, need immediate synchronized cardioversion (0.5 J/kg up to 1 J/kg); consult cardiologist; refer to the back of the book for tachycardia with poor perfusion and tachycardia with adequate perfusion algorithms
I. <i>AV Reentrant</i> : Presence of accessory bypass pathway, in conjunction with AV node, establishes cyclic pattern of reentry independent of SA node; most common cause of non-sinus tachycardia in children (see Wolff-Parkinson-White syndrome, <i>Table 7.9</i> )		
II. <i>Junctional</i> : Automatic focus; simultaneous depolarization of atria and ventricles yields invisible P wave or retrograde P wave	Cardiac surgery, idiopathic	Adjust for clinical situation; consult cardiology
III. <i>Ectopic atrial tachycardia</i> : Rapid firing of ectopic focus in atrium	Idiopathic	AV nodal blockade, ablation
<b>NODAL ESCAPE/JUNCTIONAL RHYTHM</b>		
Abnormal rhythm driven by AV node impulse, giving normal QRS complex and invisible P wave (buried in preceding QRS or T wave) or retrograde P wave (negative in lead II, positive in aVR); seen in sinus bradycardia	Common after surgery of atria	Often requires no treatment; if rate is slow enough, may require pacemaker

<sup>a</sup>Abnormal rhythm resulting from ectopic focus in atria or AV node, or from accessory conduction pathways. Characterized by different P-wave shape and abnormal P-wave axis. QRS morphology usually normal. See Fig. 7.5.<sup>6</sup>

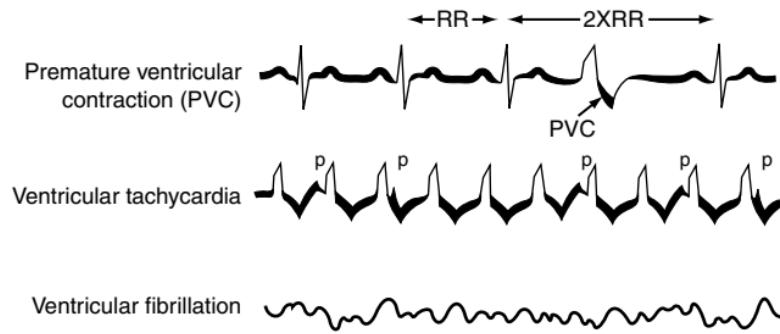
AV, Atrioventricular; CHF, congestive heart failure; HR, heart rate; ICP, intracranial pressure; PE, pulmonary embolism; SA, sinoatrial; SVT, supraventricular tachycardia.

**FIGURE 7.5**

Supraventricular Arrhythmias. *p<sup>1</sup>*, Premature atrial contraction. (From Park MK, Guntheroth WG. *How to Read Pediatric ECGs*. 4th ed. Philadelphia: Elsevier; 2006:129.)

**TABLE 7.7**  
**VENTRICULAR ARRHYTHMIAS**

Name/Description	Cause	Treatment
<b>PREMATURE VENTRICULAR CONTRACTION (PVC)</b>		
Ectopic ventricular focus causing early depolarization. Abnormally wide QRS complex appears prematurely, usually with full compensatory pause. May be unifocal or multifocal	Myocarditis, myocardial injury, cardiomyopathy, long QT, congenital and acquired heart disease, drugs (catecholamines, theophylline, caffeine, anesthetics), MVP, anxiety, hypokalemia, hypoxia, hypomagnesemia;	None; more worrisome if associated with underlying heart disease or syncope, if worse with activity, or if they are multifocal (especially couplets); address underlying cause; rule out structural heart disease
<b>Bigeminy:</b> Alternating normal and abnormal QRS complexes.		
<b>Trigeminy:</b> Two normal QRS complexes followed by an abnormal one		
<b>Couplet:</b> Two consecutive PVCs		
<b>VENTRICULAR TACHYCARDIA</b>		
Series of three or more PVCs at rapid rate (120–250 beats/min), with wide QRS complex and dissociated, retrograde, or no P wave	See causes of PVCs (70% have underlying cause)	Refer to front of book for tachycardia with poor perfusion and tachycardia with adequate perfusion algorithms
<b>VENTRICULAR FIBRILLATION</b>		
Depolarization of ventricles in uncoordinated asynchronous pattern, yielding abnormal QRS complexes of varying size and morphology with irregular, rapid rate; rare in children.	Myocarditis, MI, postoperative state, digitalis or quinidine toxicity, catecholamines, severe hypoxia, electrolyte disturbances, long QT	Requires immediate defibrillation; refer to front of book for asystole and pulseless arrest algorithm

**FIGURE 7.6**

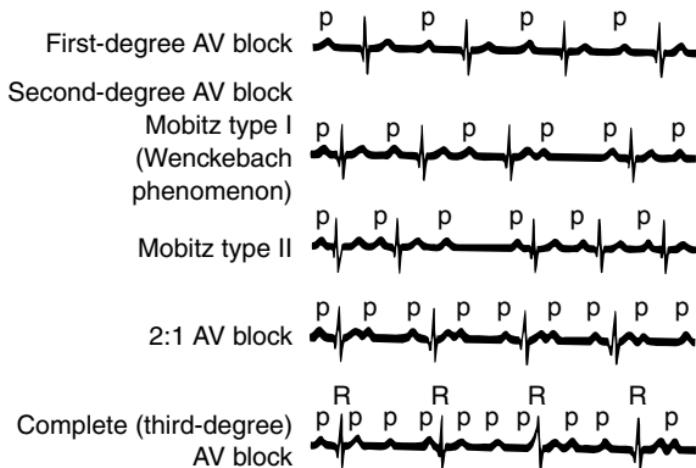
Ventricular Arrhythmias. *p*, P wave; PVC, premature ventricular contraction; RR, R-R interval. (From Park MK, Guntheroth WG. *How to Read Pediatric ECGs*. 4th ed. Philadelphia: Elsevier; 2006:138.)

**TABLE 7.8****NONVENTRICULAR CONDUCTION DISTURBANCES**

Name/Description <sup>a</sup>	Cause	Treatment
<b>FIRST-DEGREE HEART BLOCK</b>		
Abnormal but asymptomatic delay in conduction through AV node, yielding prolongation of PR interval	Acute rheumatic fever, tick-borne (e.g., Lyme) disease, connective tissue disease, congenital heart disease, cardiomyopathy, digitalis toxicity, postoperative state, normal children	No specific treatment except to address the underlying cause
<b>SECOND-DEGREE HEART BLOCK: MOBITZ TYPE I (WENCKEBACH)</b>		
Progressive lengthening of PR interval until a QRS complex is not conducted; common finding in asymptomatic teenagers	Myocarditis, cardiomyopathy, congenital heart disease, postoperative state, MI, toxicity (digitalis, $\beta$ -blocker), normal children, Lyme disease, lupus	Address underlying cause, or none needed
<b>SECOND-DEGREE HEART BLOCK: MOBITZ TYPE II</b>		
Loss of conduction to ventricle without lengthening of the PR interval; may progress to complete heart block	Same as for Mobitz type I	Address underlying cause; may need pacemaker
<b>THIRD-DEGREE (COMPLETE) HEART BLOCK</b>		
Complete dissociation of atrial and ventricular conduction, with atrial rate faster than ventricular rate; P wave and PP interval regular; RR interval regular and much slower	Congenital due to maternal lupus or other connective tissue disease	If bradycardic and symptomatic, consider pacing; refer to back of the book for bradycardia algorithm

<sup>a</sup>High-degree AV block: Conduction of atrial impulse at regular intervals, yielding 2:1 block (two atrial impulses for each ventricular response), 3:1 block, etc.

AV, Atrioventricular; MI, myocardial infarction.

**FIGURE 7.7**

Conduction Blocks. *p*, P wave; *R*, QRS complex. (From Park MK, Guntheroth WG. *How to Read Pediatric ECGs*. 4th ed. Philadelphia: Elsevier; 2006:141.)

**TABLE 7.9**  
**VENTRICULAR CONDUCTION DISTURBANCES**

Name/Description	Criteria	Causes/Treatment
<b>RIGHT BUNDLE-BRANCH BLOCK (RBBB)</b>		
Delayed right bundle conduction prolongs RV depolarization time, leading to wide QRS	<ol style="list-style-type: none"> <li>1. Prolonged or wide QRS with terminal slurred R' (m-shaped RSR' or RR') in V<sub>1</sub>, V<sub>2</sub>, aVR</li> <li>2. Wide and slurred S wave in leads I and V<sub>6</sub></li> </ol>	ASD, surgery with right ventriculotomy, occasionally seen in normal children
<b>LEFT BUNDLE-BRANCH BLOCK (LBBB)</b>		
Delayed left bundle conduction prolongs septal and LV depolarization time, leading to wide QRS with loss of usual septal signal; there is still a predominance of left ventricle forces; rare in children.	<ol style="list-style-type: none"> <li>1. Wide negative QRS complex in lead V<sub>1</sub> with loss of septal R wave</li> <li>2. Wide R or RR' complex in lead V<sub>6</sub> with loss of septal Q wave</li> </ol>	Hypertension, ischemic or valvular heart disease, cardiomyopathy
<b>WOLFF-PARKINSON-WHITE (WPW)</b>		
Atrial impulse transmitted via anomalous conduction pathway to ventricles, bypassing AV node and normal ventricular conduction system; leads to early and prolonged depolarization of ventricles; bypass pathway is a predisposing condition for SVT	<ol style="list-style-type: none"> <li>1. Shortened PR interval</li> <li>2. Delta wave</li> <li>3. Wide QRS</li> </ol>	Acute management of SVT if necessary, as previously described; consider ablation of accessory pathway if recurrent SVT; all patients need cardiology referral

ASD, Atrial septal defect; *LV*, left ventricle; *RV*, right ventricle; *SVT*, supraventricular tachycardia.

**TABLE 7.10****SYSTEMIC EFFECTS ON ELECTROCARDIOGRAM**

	Short QT	Long QT-U	Prolonged QRS	ST-T Changes	Sinus Tachycardia	Sinus Bradycardia	AV Block	Ventricular Tachycardia	Miscellaneous
<b>CHEMISTRY</b>									
Hyperkalemia			X	X			X	X	Low-voltage P waves; peaked T waves
Hypokalemia		X		X					
Hypercalcemia	X					X	X	X	
Hypocalcemia		X			X				
Hypermagnesemia								X	
Hypomagnesemia		X							
<b>DRUGS</b>									
Digitalis	X			X		T	X	T	
Phenothiazines		T							T
Phenytoin	X								
Propranolol	X					X	X		
Tricyclic antidepressants		T	T	T	T		T	T	
Verapamil						X		X	
<b>MISCELLANEOUS</b>									
CNS injury		X		X	X	X	X		

**TABLE 7.10—CONT'D**

	Short QT	Long QT-U	Prolonged QRS	ST-T Changes	Sinus Tachycardia	Sinus Bradycardia	AV Block	Ventricular Tachycardia	Miscellaneous
Friedreich ataxia				X	X				Atrial flutter
Duchenne muscular dystrophy					X	X			Atrial flutter
Myotonic dystrophy			X	X	X		X		
Collagen vascular disease				X			X	X	
Hypothyroidism						X			Low voltage
Hyperthyroidism		X		X	X		X		
Lyme disease			X				X		
Holt-Oram, maternal lupus							X		

CNS, Central nervous system; T, present only with drug toxicity; X, present.

Data from Garson A Jr. *The Electrocardiogram in Infants and Children: A Systematic Approach*. Philadelphia: Lea & Febiger; 1983:172; and Walsh EP. Cardiac arrhythmias. In: Fyler DC, Nadas A, eds. *Pediatric Cardiology*. Philadelphia: Hanley & Belfus; 1992:141–143.

**TABLE 7.11****MAJOR SYNDROMES ASSOCIATED WITH CARDIAC DEFECTS**

Syndrome	Dominant Cardiac Defect
CHARGE	TOF, truncus arteriosus, aortic arch abnormalities
DiGeorge	Aortic arch anomalies, TOF, truncus arteriosus, VSD, PDA
Trisomy 21	Atrioventricular septal defect, VSD
Marfan	Aortic root dilation, mitral valve prolapse
Loeys-Dietz	Aortic root dilation with higher risk of rupture at smaller dimensions
Noonan	Supravalvular pulmonic stenosis, LVH
Turner	COA, bicuspid aortic valve, aortic root dilation as a teenager
Williams	Supravalvular aortic stenosis, pulmonary artery stenosis
FAS	Occasional: VSD, PDA, ASD, TOF
IDM	TGA, VSD, COA, cardiomyopathy
VATER/VACTERL	VSD
VCFS	Truncus arteriosus, TOF, pulmonary atresia with VSD, TGA, interrupted aortic arch

ASD, Atrial septal defect; CHARGE, a syndrome of associated defects including Coloboma of the eye, Heart anomaly, choanal Atresia, Retardation, and Genital and Ear anomalies; COA, coarctation of aorta; FAS, fetal alcohol syndrome; IDM, infant of diabetic mother; LVH, left ventricular hypertrophy; PDA, patent ductus arteriosus; TGA, transposition of the great arteries; TOF, tetralogy of Fallot; VATER/VACTERL, association of Vertebral anomalies, Anal atresia, Cardiac anomalies, Tracheoesophageal fistula, Renal/radial anomalies, Limb defects; VCFS, velocardiofacial syndrome; VSD, ventricular septal defect.

From Park MK. *Pediatric Cardiology for Practitioners*. 5th ed. St Louis: Elsevier; 2008:10–12.

3. **Serum K<sup>+</sup> >7.5 mEq/L:** Long PR interval, wide QRS, tall T wave.
4. **Serum K<sup>+</sup> >9 mEq/L:** Absent P wave, sinusoidal.

### III. CONGENITAL HEART DISEASE

#### A. Pulse Oximetry Screening for Critical Congenital Heart Disease

1. **To be done as late as possible, but before discharge from nursery, preferably >24 hours of life, due to decreased false-positive rate.**  
Recommended to use the right hand and 1 foot, either in parallel or direct sequence.
2. **The screening result would be considered positive if:**
  - a. Any oxygen saturation measure <90%.
  - b. Oxygen saturation <95% in both extremities on three measures, each separated by 1 hour.
  - c. There is a >3% absolute difference in oxygen saturation between the right hand and foot on three measures, each separated by 1 hour.

#### B. Common Syndromes Associated with Cardiac Lesions (Table 7.11)

#### C. Acyanotic Lesions (Table 7.12)

#### D. Cyanotic Lesions (Table 7.13)

A hyperoxia test is used to evaluate the etiology of cyanosis in neonates. A baseline arterial blood gas (ABG) with saturation at  $\text{FiO}_2 = 0.21$  is obtained. Then the infant is placed in an oxygen hood at  $\text{FiO}_2 = 1$  for a minimum of

**TABLE 7.12****ACYANOTIC CONGENITAL HEART DISEASE**

Lesion Type	Examination Findings	ECG Findings	Chest Radiograph Findings
Ventricular septal defect (VSD)	2–5/6 holosystolic murmur, loudest at the LLSB, $\pm$ systolic thrill $\pm$ apical diastolic rumble with large shunt With large VSD and pulmonary hypertension, S <sub>2</sub> may be narrow	Small VSD: Normal Medium VSD: LVH $\pm$ LAE Large VSD: BVH $\pm$ LAE, pure RVH	May show cardiomegaly and increased PVMs, depending on amount of left-to-right shunting
Atrial septal defect (ASD)	Wide, fixed split S <sub>2</sub> with grade 2–3/6 SEM at the LUSB May have mid-diastolic rumble at LLSB	Small ASD: Normal Large ASD: RAD and mild RVH or RBBB with RSR' in V <sub>1</sub>	May show cardiomegaly with increased PVMs if hemodynamically significant ASD
Patent ductus arteriosus (PDA)	40%–60% in VLBW infants 1–4/6 continuous “machinery” murmur loudest at LUSB Wide pulse pressure	Small–moderate PDA: Normal or LVH Large PDA: BVH	May have cardiomegaly and increased PVMs, depending on size of shunt
Atrioventricular septal defects	Most occur in Down syndrome Hyperactive precordium with systolic thrill at LLSB and loud S <sub>2</sub> $\pm$ grade 3–4/6 holosystolic regurgitant murmur along LLSB $\pm$ systolic murmur of MR at apex $\pm$ mid-diastolic rumble at LLSB or at apex $\pm$ gallop rhythm	Superior QRS axis RVH and LVH may be present	Cardiomegaly with increased PVMs
Pulmonary stenosis (PS)	Ejection click at LUSB with valvular PS; click intensity varies with respiration, decreasing with inspiration and increasing with expiration S <sub>2</sub> may split widely with P <sub>2</sub> diminished in intensity SEM (2–5/6) $\pm$ thrill at LUSB with radiation to back and sides	Mild PS: Normal Moderate PS: RAD and RVH Severe PS: RAE and RVH with strain	Normal heart size with normal to decreased PVMs

*Continued*

**TABLE 7.12—CONT'D**

Lesion Type	Examination Findings	ECG Findings	Chest Radiograph Findings
Aortic stenosis (AS)	Systolic thrill at RUSB, suprasternal notch, or over carotids Ejection click that does not vary with respiration if valvular AS Harsh SEM (2–4/6) at second RICS or third LICS, with radiation to neck and apex ± early diastolic decrescendo murmur due to AR Narrow pulse pressure, if severe stenosis	Mild AS: Normal Moderate–severe AS: LVH ± strain	Usually normal
Coarctation of aorta may present as: 1. Infant in CHF 2. Child with HTN 3. Child with murmur	Male/female ratio of 2:1 2–3/6 SEM at LUSB, radiating to left interscapular area Bicuspid valve is often associated, so may have systolic ejection click at apex and RUSB BP in lower extremities will be lower than in upper extremities Pulse oximetry discrepancy of >5% between upper and lower extremities is also suggestive of coarctation	<i>In infancy:</i> RVH or RBBB <i>In older children:</i> LVH	Marked cardiomegaly and pulmonary venous congestion Rib notching from collateral circulation usually not seen in children younger than 5 years because collaterals not yet established

AR, Aortic regurgitation; ASD, atrial septal defect; BP, blood pressure; BVH, biventricular hypertrophy; CDG, congenital disorders of glycosylation; CHD, congenital heart disease; CHF, congestive heart failure; HTN, hypertension; LAE, left atrial enlargement; LICS, left intercostal space; LLSB, left lower sternal border; LUSB, left upper sternal border; LVH, left ventricular hypertrophy; MR, mitral regurgitation; PVM, pulmonary vascular markings; RAD, right axis deviation; RAE, right atrial enlargement; RBBB, right bundle-branch block; RICS, right intercostal space; RUSB, right upper sternal border; RVH, right ventricular hypertrophy; SEM, systolic ejection murmur; VLBW, very low birth weight (i.e., <1500 g); VSD, ventricular septal defect.

**TABLE 7.13****CYANOTIC CONGENITAL HEART DISEASE**

Lesion	Examination Findings	ECG Findings	Chest Radiograph Findings
Tetralogy of Fallot: 1. Large VSD 2. RVOT obstruction 3. RVH 4. Overriding aorta  Degree of RVOT obstruction will determine whether there is clinical cyanosis; if PS is mild, there will be a left-to-right shunt, and child will be acyanotic; increased obstruction leads to increased right-to-left shunting across VSD, and child will be cyanotic	Loud SEM at LMSB and LUSB and a loud, single $S_2$ ± thrill at LMSB and LLSB  <i>Tet spells:</i> Occur in young infants; as RVOT obstruction increases or systemic resistance decreases, right-to-left shunting across VSD occurs; may present with tachypnea, increasing cyanosis, and decreasing murmur	RAD and RVH  RAD and RVH (due to RV acting as systemic ventricle); after 3 days of age, upright T wave in $V_1$ may be only abnormality	Boot-shaped heart with normal heart size ± decreased PVMs
Transposition of great arteries	Nonspecific; extreme cyanosis; loud, single $S_2$ ; no murmur unless there is associated VSD or PS	RAD and RVH (due to RV acting as systemic ventricle); after 3 days of age, upright T wave in $V_1$ may be only abnormality	Classic finding: “egg on a string” with cardiomegaly; possible increased PVMs.
Tricuspid atresia (absent tricuspid valve and hypoplastic RV and PA; must have ASD, PDA, or VSD to survive)	Single $S_2$ + grade 2–3/6 systolic regurgitation murmur at LLSB if VSD is present. Occasional PDA murmur.	Superior QRS axis; RAE or CAE and LVH.	Normal or slightly enlarged heart size; may have boot-shaped heart.

*Continued*

**TABLE 7.13—CONT'D**

Lesion	Examination Findings	ECG Findings	Chest Radiograph Findings
Total anomalous pulmonary venous return: Instead of draining into LA, pulmonary veins drain into the following locations (must have ASD or PFO for survival): <i>Supracardiac (most common): SVC</i> <i>Cardiac:</i> Coronary sinus or RA <i>Subdiaphragmatic:</i> IVC, portal vein, ductus venosus, or hepatic vein <i>Mixed type</i>	Hyperactive RV impulse, quadruple rhythm, S <sub>2</sub> fixed and widely split, 2–3/6 SEM at LUSB, and mid-diastolic rumble at LLSB	RAD, RVH (RSR' in V <sub>1</sub> ); may see RAE	Cardiomegaly and increased PVMs; classic finding is “snowman in a snowstorm,” but this is rarely seen until after age 4 months
<b>OTHER</b>			
Cyanotic CHDs that each occur at a frequency of <1% include pulmonary atresia, Ebstein anomaly, truncus arteriosus, single ventricle, and double outlet right ventricle			

ASD, Atrial septal defect; CAE, common atrial enlargement; ECG, electrocardiogram; IVC, inferior vena cava; LA, left atrium; LLSB, left lower sternal border; LMSB, left midsternal border; LUSB, left upper sternal border; LVH, left ventricular hypertrophy; PA, pulmonary artery; PDA, patent ductus arteriosus; PFO, patent foramen ovale; PVM, pulmonary vascular markings; PS, pulmonary stenosis; RA, right atrium; RAD, right-axis deviation; RAE, right atrial enlargement; RV, right ventricle; RVH, right ventricular hypertrophy; RVOT, right ventricular outflow tract; SEM, systolic ejection murmur; SVC, superior vena cava; VSD, ventricular septal defect.

10 minutes, and the ABG is repeated. In cardiac disease, there will not be a significant change in  $\text{Pao}_2$  following the oxygen challenge test. A  $\text{Pao}_2$  of  $>200$  after exposure to  $\text{Fio}_2$  of 1.0 is considered normal, and  $>150$  indicates pulmonary rather than cardiac disease. **Note:** Pulse oximetry is not useful for following changes in oxygenation once saturation has reached 100% (approximately a  $\text{Pao}_2$  of  $>90$  mmHg).<sup>12-17</sup> See Table EC 7.A for interpretation of oxygen challenge test (hyperoxia test).

#### IV. ACQUIRED HEART DISEASE

##### A. Myocardial Infarction (MI) in Children (Box 7.4; Fig. 7.8)

##### B. Endocarditis

1. **Common causative organisms:** Approximately 70% of endocarditis is caused by streptococcal species (*Streptococcus viridans*, enterococci), 20% by staphylococcal species (*Staphylococcus aureus*, *Staphylococcus epidermidis*), and 10% by other organisms (*Haemophilus influenzae*, gram-negative bacteria, fungi).
2. **Presentation:** Heart murmur, recurrent fever, splenomegaly, petechiae, fatigue, Osler nodes (tender nodules at the fingertips), Janeway lesions (painless hemorrhagic areas on the palms or soles), splinter hemorrhages, Roth spots (retinal hemorrhages).
3. **Diagnosis**—Duke's Criteria:
  - a. Pathologic criteria:
    - (1) Direct evidence of endocarditis based upon histologic findings.
    - (2) Gram stain positive or cultures of specimens.
  - b. Clinical criteria: 1 major criterion and 1 minor OR 3 minor criteria:
    - (1) Major: Persistently positive blood cultures (2 sets 1 hour apart), positive echocardiogram for vegetations, new regurgitant murmur, single positive blood culture for *Coxiella burnetii*.
    - (2) Minor: Fever, predisposing valvular condition (prosthetic heart valve, valve lesion OR intravenous drug user [IVDU]), vascular phenomenon (e.g., emboli), immunologic phenomenon (e.g., Roth's spots, Osler's nodes), positive blood cultures that do not meet major criteria.
4. Management: Daily blood cultures while febrile; support heart failure symptoms with diuretics, digoxin, etc.

##### C. Bacterial Endocarditis Prophylaxis

See Box 7.5 for cardiac conditions that meet criteria for prophylaxis.<sup>18</sup>

1. **All dental procedures** that involve treatment of gingival tissue, the periapical region of the teeth, or oral mucosal perforation.
2. **Invasive procedures** that involve incision or biopsy of respiratory mucosa, such as tonsillectomy and adenoidectomy.
3. **Not recommended** for genitourinary or gastrointestinal tract procedures; solely for bacterial endocarditis prevention.
4. **Treatment:** Amoxicillin is preferred PO; ampicillin if unable to take PO; cephalexin if allergic to penicillins.<sup>28</sup>

**TABLE EC 7.A****INTERPRETATION OF OXYGEN CHALLENGE TEST (HYPEROXIA TEST)**

Condition	$\text{FiO}_2 = 0.21$ $\text{PaO}_2$ (% Saturation)	$\text{FiO}_2 = 1.00$ $\text{PaO}_2$ (% Saturation)	$\text{PaCO}_2$
Normal	70 (95)	>200 (100)	35
Pulmonary disease	50 (85)	>150 (100)	50
Neurologic disease	50 (85)	>150 (100)	50
Methemoglobinemia	70 (85)	>200 (85)	35
Cardiac disease			
• Separate circulation <sup>a</sup>	<40 (<75)	<50 (<85)	35
• Restricted PBF <sup>b</sup>	<40 (<75)	<50 (<85)	35
• Complete mixing without restricted PBF <sup>c</sup>	50 (85)	<150 (<100)	35
Persistent pulmonary hypertension	<i>Predictal</i>	<i>Postductal</i>	
PFO (no R to L shunt)	70 (95)	<40 (<75)	Variable 35–50
PFO (with R to L shunt)	<40 (<75)	<40 (<75)	Variable 35–50

<sup>a</sup>D-Transposition of the great arteries (D-TGA) with intact ventricular septum.

<sup>b</sup>Tricuspid atresia with pulmonary stenosis or atresia, pulmonary atresia or critical pulmonary stenosis with intact ventricular septum, or tetralogy of Fallot.

<sup>c</sup>Truncus arteriosus, total anomalous pulmonary venous return, single ventricle, hypoplastic left heart syndrome, D-TGA with ventricular septal defect, tricuspid atresia without pulmonary stenosis or atresia.

$\text{FiO}_2$ , Fraction of inspired oxygen; PBF, pulmonary blood flow; PFO, patent foramen ovale.

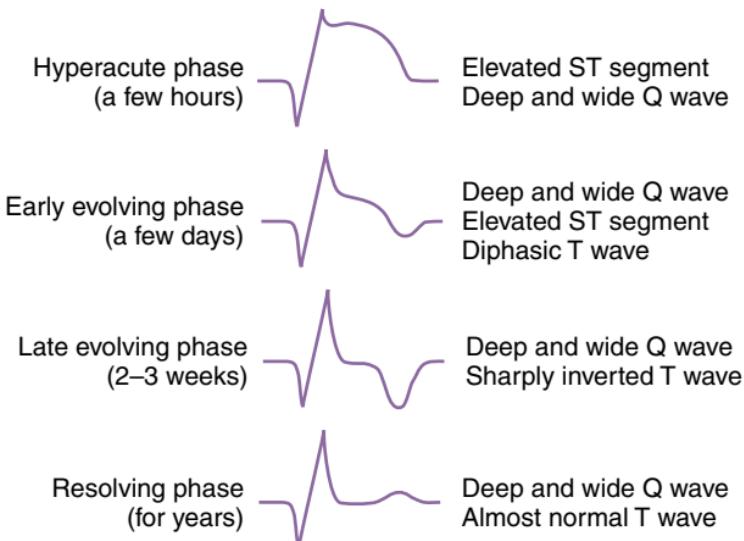
From Lees MH. Cyanosis of the newborn infant: recognition and clinical evaluation. *J Pediatr*. 1970;77:484; Kitterman JA. Cyanosis in the newborn infant. *Pediatr Rev*. 1982;4:13; and Jones RW, Baumer JH, Joseph MC, et al. Arterial oxygen tension and response to oxygen breathing in differential diagnosis of heart disease in infancy. *Arch Dis Child*. 1976;51:667–673.

**BOX 7.4****MYOCARDIAL INFARCTION IN CHILDREN<sup>25,26</sup>**

Etiologies	Diagnosis
Anomalous origin of coronary artery	<b>ECG findings</b> <sup>10,11</sup> : See Fig. 7.12
Kawasaki disease	<b>Biomarkers</b>
Congenital heart disease	Troponin I, CK-MB nonspecific for ischemic injury in children
Dilated cardiomyopathy	
Severe hypertension	
SLE	
Myocarditis	
Drug ingestion (cocaine, adrenergic drugs)	

7

CK-MB, Creatine kinase-MB; ECG, electrocardiogram; MI, myocardial Infarction; SLE, systemic lupus erythematosus.

**FIGURE 7.8**

Sequential Changes During Myocardial Infarction. (From Park MK, Guntheroth WG. *How to Read Pediatric ECGs*. 4th ed. Philadelphia: Elsevier; 2006:115.)

#### D. Myocardial Disease

1. **Dilated cardiomyopathy:** End result of myocardial damage leading to atrial and ventricular dilation with decreased systolic contractile function of the ventricles.
  - a. Treatment: Management of congestive heart failure (CHF) (digoxin, diuretics, vasodilation, angiotensin-converting enzyme [ACE] inhibitors).
  - b. Anticoagulants should be considered to decrease the risk of thrombus formation. Cardiac transplant may eventually be required.

**BOX 7.5****CARDIAC CONDITIONS FOR WHICH ANTIBIOTIC PROPHYLAXIS IS RECOMMENDED**

- Prosthetic cardiac valve
- Previous bacterial endocarditis
- Congenital heart disease (CHD)—Limited to the following conditions:
  - Unrepaired cyanotic defect, including palliative shunts and conduits
  - Completely repaired CHD with prosthetic material/device (placed by surgery or catheterization), during first 6 months after procedure
  - Repaired CHD with residual defects at or adjacent to the site of prosthetic patch or device (which inhibits endothelialization)
  - Cardiac transplantation patients who develop cardiac valvulopathy

Data from Wilson W, Taubert KA, Gewitz M, et al. Prevention of infective endocarditis: Guidelines from the American Heart Association: A guideline from the American Heart Association Rheumatic Fever, Endocarditis, and Kawasaki Disease Committee, Council on Cardiovascular Disease in the Young, and the Council on Clinical Cardiology, Council on Cardiovascular Surgery and Anesthesia, and the Quality of Care and Outcomes Research Interdisciplinary Working Group. *Circulation*. 2007;116(15):1736–1754.

2. **Hypertrophic obstructive cardiomyopathy (HOCM):** Abnormality of myocardial cells leading to significant ventricular hypertrophy (usually left ventricle) with small to normal ventricular dimensions. Increased contractile function, impaired filling secondary to stiff ventricles. Most common type is asymmetrical septal hypertrophy with (HOCM) or without left ventricular outflow obstruction. There is a 4% to 6% incidence of sudden death in children and adolescents.
  - a. Treatment: Moderate restriction of physical activity, negative inotropes ( $\beta$ -blocker, calcium channel blocker) to help improve filling, and maintenance of adequate hydration. If at increased risk for sudden death, may consider implantable defibrillator. If symptomatic with subaortic obstruction, may benefit from myectomy.
  - b. Additional management: HOCM is a preload dependent lesion and, therefore, patient may benefit from higher rates of fluid administration. Avoid inotropes, tachycardia, and afterload reduction.
3. **Restrictive cardiomyopathy:** Myocardial or endocardial disease (usually infiltrative or fibrotic) resulting in stiff ventricular walls with restriction of diastolic filling but normal contractile function. Results in atrial enlargement. Associated with a high mortality rate. Very rare in children. Treatment is supportive with diuretics, anticoagulants, calcium channel blockers, a pacemaker for heart block, and cardiac transplantation, if severe.
4. **Myocarditis:** Inflammation of myocardial tissue
  - a. Etiology:
    - (1) Infectious: viral (coxsackie virus, echovirus, adenovirus), bacterial, rickettsial, fungal, parasitic
    - (2) Other: immune-mediated disease (Kawasaki disease, acute rheumatic fever), collagen vascular disease, toxin-induced

- b. Presentation: Symptoms can be nonspecific, including fatigue, shortness of breath, emesis. Exam includes signs of CHF, soft systolic murmur, arrhythmia.
- c. Testing:
  - (1) Imaging: ECG: Low QRS voltages throughout (<5 mm), ST-segment and T-wave changes (e.g., decreased T-wave amplitude), prolongation of QT interval, arrhythmias (especially premature contractions, first- or second-degree AV block); echo shows enlarged chambers and impaired LV function
  - (2) Labs: CK, troponin
- d. Treatment: Bed rest, diuretics, inotropes (dopamine, dobutamine, milrinone), digoxin, gamma globulin, ACE inhibitors, possibly steroids.
- e. May require ventricular assist device and/or heart transplantation ( $\approx 20\%$  to 25% of cases).

## E. Pericardial Disease

- 1. **Pericarditis:** Inflammation of visceral and parietal layers of pericardium. It is often self-limited.
  - a. Presentation: Chest pain (often pleuritic in nature), fever, tachycardia, distant heart sounds, friction rub.
  - b. EKG: Diffuse ST-segment elevation in almost all leads (representing inflammation of adjacent myocardium); PR-segment depression.
  - c. Treatment: Address underlying condition and provide symptomatic treatment with rest, analgesia, and anti-inflammatory drugs.
- 2. **Pericardial effusion:** Accumulation of excess fluid in pericardial sac.
  - a. Etiology: Acute pericarditis, serous effusion from increased hydrostatic pressure (CHF), decreased plasma oncotic pressure, increased capillary permeability.
  - b. Presentation: Can be asymptomatic, chest or abdominal pain, muffled heart sounds, dullness to percussion, vital sign instability from cardiac compression (e.g., hypotension).
  - c. EKG: Decreased QRS voltage, electrical alternans.
  - d. Treatment: Address underlying condition. Observe if asymptomatic; use pericardiocentesis if there is sudden increase in volume or hemodynamic compromise. Nonsteroidal antiinflammatory drugs (NSAIDs) or steroids may be of benefit, depending on etiology.
- 3. **Cardiac tamponade:** Accumulation of pericardial fluid under high pressure causing compression of cardiac chambers, limiting filling, and decreasing stroke volume and cardiac output.
  - a. Etiology: Same as pericardial effusion.
  - b. Presentation: Dyspnea, fatigue, signs of CHF (jugular venous distension, hepatomegaly, edema, tachypnea/rales), pulsus paradoxus.
  - c. EKG: Same as pericardial effusion.
  - d. Echocardiogram: RV collapse in early diastole, RA/LA collapse in end diastole and early systole.

- e. Treatment is pericardiocentesis with temporary catheter left in place if necessary; pericardial window or stripping, if it is a recurrent condition.

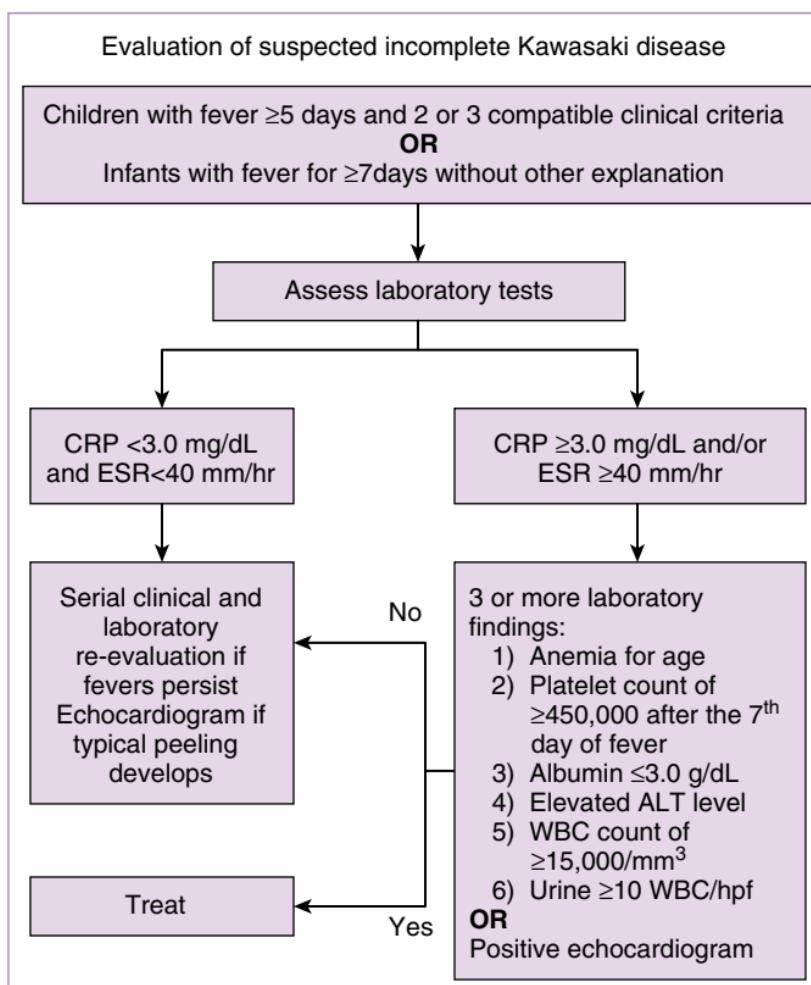
## F. Kawasaki Disease<sup>19</sup>

Acute febrile vasculitis of unknown etiology, which is common in children aged <8 years and is the leading cause of acquired childhood heart disease in developed countries.

1. **Etiology:** Unknown; thought to be immune regulated in response to infectious agents or environmental toxins.
2. **Diagnosis:**
  - a. Typical Kawasaki disease: Based on clinical criteria. These include high fever lasting 5 days or more, plus at least four of the following five criteria:
    - (1) Bilateral, painless, bulbar conjunctival injection without exudate
    - (2) Erythematous mouth and pharynx, strawberry tongue, or red cracked lips
    - (3) Polymorphous exanthem (may be morbilliform, maculopapular, or scarlatiniform)
    - (4) Swelling of hands and feet with erythema of palms and soles
    - (5) Cervical lymphadenopathy (>1.5 cm in diameter), usually single and unilateral
  - b. Atypical/incomplete Kawasaki disease: A suspicion of Kawasaki disease but with fewer of the criteria required for diagnosis. Even without all criteria, there is a risk for coronary artery abnormalities.
    - (1) More often seen in infants. Echocardiography should be considered in any infant <6 months with fever >7 days duration, laboratory evidence of systemic inflammation (CRP >3 and/or ESR >40), and no other explanation for the febrile illness.
    - (2) See Fig. 7.9 for evaluation of incomplete Kawasaki disease.
    - (3) Supplemental laboratory criteria: Albumin  $\leq 3.0$  g/dL, anemia for age, elevation of alanine aminotransferase, platelets after 7 days  $\geq 450,000/\text{mm}^3$ , white blood cell count  $\geq 15,000/\text{mm}^3$ , and urine white blood cells/hpf  $\geq 10$  (non-catheterized specimen).
  3. **Other clinical findings:** Often associated with extreme irritability, abdominal pain, diarrhea, vomiting. Also seen are arthritis and arthralgia, hepatic enlargement, jaundice, acute acalculous distention of the gallbladder, carditis.
  4. **Laboratory findings:** Leukocytosis with left shift, neutrophils with vacuoles or toxic granules, elevated C-reactive protein (CRP) or erythrocyte sedimentation rate (ESR) (seen acutely), thrombocytosis, normocytic and normochromic anemia, sterile pyuria (33%), increased transaminases (40%), hyperbilirubinemia (10%).
  5. **Subacute phase (11 to 25 days after onset of illness):** Resolution of fever, rash, and lymphadenopathy. Often, desquamation of the fingertips or toes and thrombocytosis occur. Cardiovascular

complications: If untreated, 20% to 25% develop coronary artery aneurysms and dilation in subacute phase (peak prevalence occurs about 2 to 4 weeks after onset of disease; rarely appears after 6 weeks) and are at risk for coronary thrombosis acutely and coronary stenosis chronically. Carditis; aortic, mitral, and tricuspid regurgitation; pericardial effusion; CHF; MI; left ventricular dysfunction; and ECG changes may also occur.

6. **Convalescent phase:** ESR, CRP, and platelet count return to normal. Those with coronary artery abnormalities are at increased risk for MI, arrhythmias, and sudden death.
7. **Management** (see also [Table EC 7.B](#))<sup>19</sup>



**FIGURE 7.9**

Evaluation of Incomplete Kawasaki Disease. (From Diagnosis, Treatment, and Long-Term Management of Kawasaki Disease: A Scientific Statement for Health Professionals from the American Heart Association. *Circulation* 2017; Mar 29.)

- a. Intravenous immunoglobulin (IVIG)
  - (1) Shown to reduce incidence of coronary artery dilation to <3% and decrease duration of fever, if given in the first 10 days of illness. Current recommended regimen is a single dose of IVIG, 2 g/kg over 10 to 12 hours.<sup>19</sup>
  - (2) Can be given to children after 10th day of fever if ESR or CRP elevated with persistent fever.
  - (3) Approximately 10% of patients treated with IVIG fail to respond (persistent or recurrent fever  $\geq 36$  hours after IVIG completion). Retreat with second dose.<sup>19</sup>
- b. Aspirin is recommended for both its anti-inflammatory and antiplatelet effects. In the United States, high-dose aspirin (80 to 100 mg/kg/day divided in four doses) is recommended 48 to 72 hours after defervescence. This is given with IVIG. Low-dose aspirin (3 to 5 mg/kg/day as a single daily dose) is continued for 6 to 8 weeks or until platelet count and ESR are normal (if there are no coronary artery abnormalities). Aspirin may be continued indefinitely, if coronary artery abnormalities persist.<sup>19</sup>
- c. Dipyridamole is sometimes used as an alternative to aspirin, particularly if symptoms of influenza or varicella arise while on aspirin (due to concern for Reye syndrome).
- d. Follow-up: Serial echocardiography is recommended to assess coronary arteries and left ventricular function (at time of diagnosis, at 2 weeks, at 6 to 8 weeks, and at 12 months [optional]). More frequent intervals and long-term follow-up are recommended if abnormalities are seen on echocardiography. Cardiac catheterization may be necessary.
- e. Follow up with Cardiology depending on presence of coronary aneurysms and Z score of aneurysm (see [Table EC 7.B](#)).

## G. Rheumatic Heart Disease

1. **Etiology:** Believed to be an immunologically-mediated, delayed sequela of group A streptococcal pharyngitis.
2. **Clinical findings:** History of streptococcal pharyngitis 1 to 5 weeks before onset of symptoms. Often with pallor, malaise, easy fatigability.
3. **Diagnosis:** Jones criteria ([Box 7.6](#)).
4. **Management:** Penicillin, bed rest, salicylates, supportive management of CHF (if present) with diuretics, digoxin, morphine.

## V. IMAGING

- A. **Chest Radiograph** ([Fig. 7.10](#))
- B. **Echocardiography** ([Table EC 7.C](#))

## VI. PROCEDURES

- A. **Cardiac Surgery** ([Fig. 7.11](#), [Table 7.14](#))

- B. **Cardiac Catheterization**<sup>13,14</sup>

1. Performed in pediatric patients for diagnostic and interventional purposes, including pressure measurements, angiography, embolization of

**TABLE EC 7.B****GUIDELINES FOR TREATMENT AND FOLLOW-UP OF CHILDREN WITH KAWASAKI DISEASE**

Risk Level	Pharmacologic Therapy	Physical Activity	Follow-Up and Diagnostic Testing	Invasive Testing
I. No coronary artery changes at any stage of illness	None beyond initial 6–8 weeks	No restrictions beyond initial 6–8 weeks	Counsel on cardiovascular risk factors every 5 years	None recommended
II. Transient coronary artery ectasia that resolves by 8 weeks after disease onset	None beyond initial 6–8 weeks	No restrictions beyond initial 6–8 weeks	Counsel on cardiovascular risk factors every 3–5 years	None recommended
III. Small-to-medium solitary coronary artery aneurysm	3–5 mg/kg/day aspirin, at least until aneurysm resolves	For patients in first decade of life, no restriction beyond initial 6–8 weeks; during the second decade of life, physical activity guided by stress testing every 2 years; avoid competitive contact and high-impact sports while on antiplatelet therapy	Annual follow-up with echocardiogram and electrocardiogram	Angiography, if stress testing or echocardiography suggests stenosis
IV. One or more large, >6 mm, aneurysms and coronary arteries with multiple small-to-medium aneurysms, without obstruction	Long-term aspirin (3–5 mg/kg/day) and warfarin or LMWH for patients with giant aneurysms	Annual stress testing guides physical activity; avoid competitive contact and high-impact sports while on anticoagulant therapy	Echocardiogram and electrocardiogram at 6-month intervals, annual stress testing, atherosclerosis risk factor counseling at each visit	Cardiac catheterization 6–12 months after acute illness with additional testing if ischemia noted or testing inconclusive
V. Coronary artery obstruction	Long-term aspirin (3–5 mg/kg/day); warfarin or LMWH if giant aneurysm persists; consider use of $\beta$ -blockers to reduce myocardial work	Contact sports, isometrics, and weight training should be avoided; other physical activity recommendations guided by outcome of stress testing or myocardial perfusion scan	Echocardiogram and electrocardiogram at 6-month intervals, annual Holter and stress testing	Cardiac catheterization 6–12 months after acute illness to aid in selecting therapeutic options, additional testing if ischemia noted

*LMWH*, Low molecular weight heparin.

**BOX 7.6****GUIDELINES FOR DIAGNOSIS OF INITIAL ATTACK OF RHEUMATIC FEVER (JONES CRITERIA)**

Major Manifestations	Minor Manifestations
Carditis	Clinical findings: Arthralgia
Polyarthritis	Fever
Chorea	Laboratory findings: Elevated acute phase reactants (erythrocyte sedimentation rate, C-reactive protein)
Erythema marginatum	Prolonged PR interval
Subcutaneous nodules	

**Plus Supporting Evidence of Antecedent Group A Streptococcal Infection**

- Positive throat culture or rapid streptococcal antigen test
- Elevated or rising streptococcal antibody titer

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**NOTE:** If supported by evidence of preceding group A streptococcal infection, the presence of two major manifestations or of one major and two minor manifestations indicates a high probability of acute rheumatic fever.

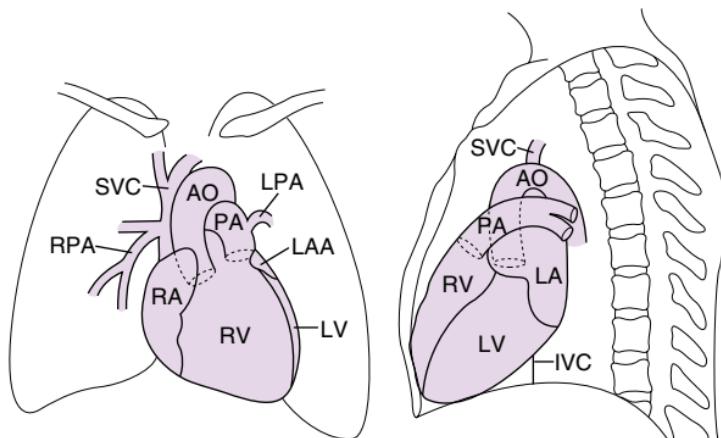
abnormal vessels, dilation of atretic valves and vessels, device closure of cardiac defects, and electrophysiology procedures.

- Relatively common complications to be aware of: Arrhythmias (SVT, AV block, bradycardia, etc.), vascular complications (thrombosis, decreased/absent pulses), intervention-related (balloon rupture, etc.), bleeding.
- Other less common complications: Myocardial/vessel staining, cardiac perforation, cardiac tamponade, air embolus, infection, allergic reaction, cardiac arrest, and death.

**VII. COMMON CARDIAC COMPLAINTS****A. Non-Traumatic Chest Pain<sup>20</sup>****1. Etiologies**

- Life-threatening causes
  - Cardiac: Congenital heart disease (CHD) with left ventricular outflow tract obstruction, coronary artery abnormality, pericarditis, myocarditis, dilated cardiomyopathy, aortic root dissection; cardiac etiologies are rare in children (prevalence <6%).<sup>25</sup>
  - Non-cardiac: Pneumothorax, pulmonary embolism, pulmonary HTN, acute chest syndrome.
- Common, non-cardiac causes (94% to 99% patients):
 

Musculoskeletal (costochondritis), respiratory (asthma, pneumonia,

**FIGURE 7.10**

Radiological Contours of the Heart. *AO*, Aorta; *IVC*, inferior vena cava; *LA*, left atrium; *LAA*, left atrial appendage; *LPA*, left pulmonary artery; *LV*, left ventricle; *PA*, pulmonary artery; *RA*, right atrium; *RPA*, right pulmonary artery; *RV*, right ventricle; *SVC*, superior vena cava.

pleuritic), gastrointestinal (gastroesophageal reflux disease [GERD]), psychiatric (panic attack, hyperventilation syndrome).

2. **When to consider referral to cardiologist:** Symptoms that suggest cardiac etiology (palpitations, syncope with exertion, and decreased exercise tolerance), ECG changes, new murmur.

## B. Syncope<sup>21</sup>

### 1. Etiologies

#### a. Cardiac etiologies:

- (1) Electrical disturbances: Long QT syndrome, Brugada syndrome, congenital short QT syndrome, catecholaminergic polymorphic ventricular tachycardia.
- (2) Structural heart disease: Hypertrophic cardiomyopathy, coronary artery anomalies, valvular aortic stenosis, dilated cardiomyopathy, acute myocarditis, pulmonary HTN.

#### b. Non-cardiac etiologies<sup>21</sup>:

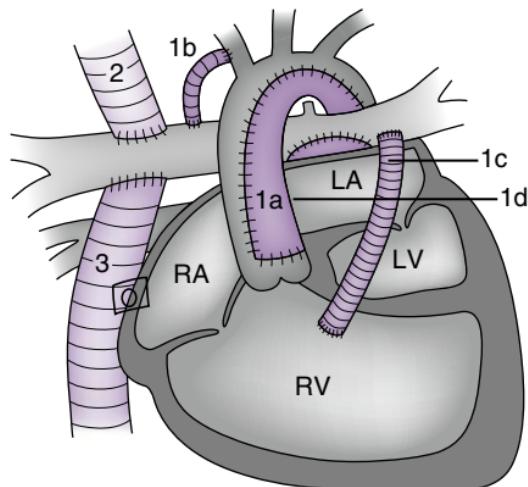
- (1) Common: Vasovagal syncope (50% pediatric syncope), breath holding spells, orthostatic hypotension.
- (2) Life-threatening: Heat illness/stroke, anaphylaxis, toxic ingestion, hypoglycemia.

### 2. When to consider referral to cardiologist:

- a. History: Congenital/acquired heart disease, syncope with exertion, associated chest pain or palpitations.

**TABLE EC 7.C****ECHOCARDIOGRAMS**

	<b>Transthoracic Echocardiogram (TTE)</b>	<b>Transesophageal Echocardiogram (TEE)</b>
Approach	Transducer placed on chest externally	Transducer on end of modified endoscope to view heart from esophagus
Pros	Does not require general anesthesia Simpler to perform than TEE	Better views in obese patients Good for intraoperative use Better visualization of small lesions/ vegetations
Cons	Limited views in certain patients (uncooperative, obese, suspected endocarditis)	Requires general anesthesia More difficult to perform

**FIGURE 7.11**

Schematic diagram of cardiac shunts, including the modified Blalock-Taussig (*BT*), Sano modification, bidirectional Glenn, and Fontan shunts.

7

- b. Family history: Early sudden cardiac death, arrhythmia, cardiomyopathy.
- c. Evaluation: Abnormal cardiac exam or abnormal ECG.

### VIII. EXERCISE RECOMMENDATIONS FOR PATIENTS WITH CONGENITAL HEART DISEASE

See [Table EC 7.D](#) for exercise recommendations for patients with CHD.<sup>22</sup>

### IX. LIPID MONITORING RECOMMENDATIONS

#### A. Screening of Children and Adolescents<sup>23</sup>

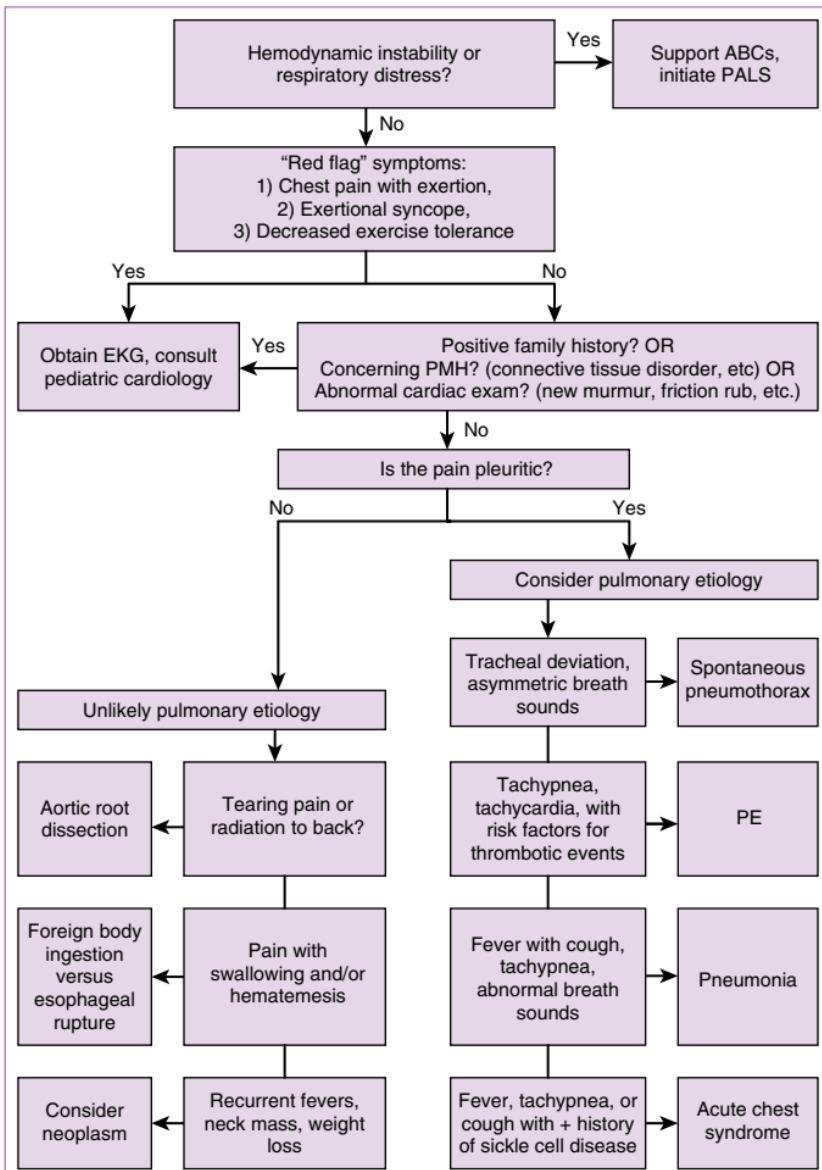
1. **Universal screening:** Children 9 to 11 years old (prior to onset of puberty) and at 17 to 21 years.
2. **Targeted screening:** 2 to 8 years old and 12 to 16 years old with risk factors:
  - a. Moderate or high-risk medical condition: History of prematurity, very low birth weight, CHD (repaired or unrepaired), recurrent urinary tract infections, renal or urologic malformations, family history of congenital renal disease, solid organ transplant, malignancy or bone marrow transplant, treatment with drugs known to raise BP, other systemic illness associated with HTN (e.g., neurofibromatosis, tuberous sclerosis), evidence of elevated intracranial pressure.
  - b. Other cardiovascular risk factors, including diabetes, HTN, body mass index  $\geq 95^{\text{th}}$  percentile, cigarette use.

**TABLE 7.14**  
**CARDIAC SURGERIES<sup>29</sup>**

Intervention	Indication	Procedure
Atrial septostomy	Common: TGA, HLHS with restrictive atrial septum Less common: tricuspid/mitral/aortic/pulmonary atresia, TAPVR	Percutaneous procedure with balloon-tipped catheter, intra-arterial opening created to allow mixing of blood between systemic and pulmonary systems
Palliative systemic-to-pulmonary artery shunts (e.g., Blalock-Taussig shunt) Norwood procedure, stage 1 (neonatal period)	Lesions with impaired pulmonary perfusion (TOF, HLHS, tricuspid atresia, pulmonary atresia) HLHS	Shunt created to increase pulmonary blood flow MPA anastomosis to aorta with arch reconstruction Modified BTS or Sano performed to provide pulmonary blood flow ASD created for decompression of left atrium Expected oxygen saturation 75%–85%
Bidirectional Glenn shunt or hemi-Fontan (3–6 months)	HLHS Intermediate step between Norwood 1 and Fontan	Bidirectional Glenn shunt or hemi-Fontan to reduce volume overload of single right ventricle Expected oxygen saturation 80%–85%
Fontan procedure	Functionally single ventricle (tricuspid atresia, HLHS)	Anastomosis of right atria and/or IVC to pulmonary arteries, separates systemic and pulmonary circulations Expected oxygen saturation >92%
Modified Fontan	Single ventricle	Completely separates systemic and pulmonary circulations Expected oxygen saturations >92%
Arterial switch	TGA	Connects aorta to LV and PA to RV, reconnects coronary arteries to aorta Normal oxygen saturations
Ross procedure ("switch procedure")	Aortic stenosis	Pulmonary valve used to replace diseased aortic valve, pulmonary valve replaced by homograft, avoids long-term anticoagulation Normal oxygen saturations

ASD, Atrial septal defect; BTS, Blalock-Taussig shunt; HLHS, hypoplastic left heart syndrome; LV, left ventricle; MPA, main pulmonary artery; PA, pulmonary artery; PVR, pulmonary arteriovenous return; TGA, transposition of great arteries; TOF, tetralogy of Fallot.

- c. Family history of early cardiovascular disease (CVD) or severe hypercholesterolemia:
- (1) Parent or grandparent who at <55 years old (males) or <65 years old (females) suffered an MI or sudden death, underwent a coronary artery procedure, or who had evidence of coronary atherosclerosis, peripheral vascular disease, or cerebrovascular disease.
  - (2) Parent with total cholesterol  $\geq 240$  mg/dL or known dyslipidemia.

**FIGURE 7.12**

Algorithm for the evaluation of non-traumatic chest pain. ABCs, airway, breathing, and circulation; EKG, electrocardiogram; PALS, pediatric advanced life support; PE, pulmonary embolism; PMH, past medical history.

**B. Goals for Lipid Levels in Childhood<sup>23</sup>**

1. **Total cholesterol**
  - a. Acceptable (<170 mg/dL): Repeat measurement in 3 to 5 years.
  - b. Borderline (170 to 199 mg/dL): Repeat cholesterol and average with previous measurement. If <170 mg/dL, repeat in 3 to 5 years. If ≥ 170 mg/dL, obtain lipoprotein analysis.
  - c. High (≥200 mg/dL): Obtain lipoprotein analysis.
2. **Low-density lipoprotein (LDL) cholesterol**
  - a. Acceptable (<110 mg/dL)
  - b. Borderline (110 to 129 mg/dL)
  - c. High (≥130 mg/dL)

**X. CARDIOVASCULAR SCREENING****A. Sports<sup>24</sup>**

There is no established or mandated pre-participation sports screening. There is a recommended history and physical examination screening from the AHA.<sup>24</sup> Routine ECGs are not required unless there is suspicion of underlying cardiac disease **Box EC 7.A**.

**B. Attention-Deficit/Hyperactivity Disorder (ADHD)<sup>27</sup>**

1. **Obtain a good patient and family history as well as physical examination.**
2. **There is no increased risk of sudden cardiac death in children without cardiac disease taking ADHD medications.** There is no consensus on universal ECG screening. ECGs should be obtained in those who screen with positive answers on history, in cases of polypharmacy, in those with tachycardia while on medications, and in those with a history of significant cardiac disease. If a patient has significant heart disease or concern for cardiac disease, have patient evaluated by a pediatric cardiologist.

**XI. WEB RESOURCES**

- <http://www.pted.org>
- <https://murmurquiz.org>

**REFERENCES**

A complete list of references can be found online at [www.expertconsult.com](http://www.expertconsult.com).

**TABLE EC 7.D****EXERCISE RECOMMENDATIONS FOR CONGENITAL HEART DISEASE AND SPORTS ALLOWED FOR SOME SPECIFIC CARDIAC LESIONS<sup>18</sup>**

Diagnosis	Sports Allowed		
Sports Classification	Low Dynamic (A)	Moderate Dynamic (B)	High Dynamic (C)
Small ASD or VSD		No restriction	
Mild aortic stenosis		No restriction	
MVP (without other risk factors)		No restriction	
Moderate aortic stenosis		IA, IB, IIA	
Mild LV dysfunction		IA, IB, IC	
Moderate LV dysfunction		IA only	
Long QT syndrome		IA only	
Hypertrophic cardiomyopathy		None (or IA only)	
Severe aortic stenosis		None	
I. Low static	Billiards Bowling Golf Riflery	Baseball/Softball Table tennis Volleyball Fencing	Racket sports Cross-country skiing Field hockey <sup>a</sup> Race walking Running (long distance) Soccer <sup>a</sup>
II. Moderate static	Archery Auto racing <sup>a,b</sup> Diving <sup>a,b</sup> Equestrian <sup>a,b</sup> Motorcycling <sup>a,b</sup>	Fencing Field events (jumping) Figure skating <sup>a</sup> Football (American) <sup>a</sup> Surfing Rugby <sup>a</sup> Running (sprint) Synchronized swimming <sup>b</sup>	Basketball <sup>a</sup> Ice hockey <sup>a</sup> Cross-country skiing (skating technique) Swimming Lacrosse <sup>a</sup> Running (middle distance) Team handball
III. High static	Bobsledding Field events Gymnastics <sup>a,b</sup> Rock climbing Sailing Windsurfing <sup>a,b</sup> Waterskiing <sup>a,b</sup> Weight-lifting <sup>a,b</sup>	Bodybuilding <sup>a,b</sup> Downhill skiing <sup>a,b</sup> Skateboarding <sup>a,b</sup>	Boxing/Wrestling <sup>a</sup> Martial arts <sup>a</sup> Rowing Speed skating Cycling <sup>a,b</sup>

<sup>a</sup>Danger of bodily collision.<sup>b</sup>Increased risk if syncope occurs.

ASD, Atrial septal defect; LV, left ventricular; MVP, mitral valve prolapse; VSD, ventricular septal defect.

Data from Maron BJ, Zipes DP. 36th Bethesda Conference: Eligibility recommendations for competitive athletes with cardiovascular abnormalities. *J Am Coll Cardiol*. 2005;45(8):1318–1321; and Committee on Sports Medicine and Fitness, American Academy of Pediatrics. Medical conditions affecting sports participation. *Pediatrics*. 2001;107(5):1205–1209.

**BOX EC 7.A****THE 12-ELEMENT AMERICAN HEART ASSOCIATION RECOMMENDATIONS FOR PARTICIPATION: CARDIOVASCULAR SCREENING OF COMPETITIVE ATHLETES****Medical History<sup>a</sup>****Personal History**

1. Exertional chest pain/discomfort
2. Unexplained syncope/near syncope<sup>b</sup>
3. Excessive exertional and unexplained dyspnea/fatigue, associated with exercise
4. Prior recognition of a heart murmur
5. Elevated systemic blood pressure

**Family History**

1. Premature death (sudden and unexpected, or otherwise) before age 50 years due to heart disease, in  $\geq 1$  relative
2. Disability from heart disease in a close relative  $< 50$  years of age
3. Specific knowledge of certain cardiac conditions in family members:  
Hypertrophic or dilated cardiomyopathy, long-QT syndrome or other ion channelopathies, Marfan syndrome, or clinically important arrhythmias

**Physical Examination**

1. Heart murmur<sup>c</sup>
2. Femoral pulses to exclude aortic coarctation
3. Physical stigmata of Marfan syndrome
4. Brachial artery blood pressure (sitting position)<sup>d</sup>

<sup>a</sup>Parental verification is recommended for high school and middle school athletes.

<sup>b</sup>Judged not to be neurocardiogenic (vasovagal); of particular concern when related to exertion.

<sup>c</sup>Auscultation should be performed in both supine and standing positions (or with Valsalva maneuver), specifically to identify murmurs of dynamic left ventricular outflow tract obstruction.

<sup>d</sup>Preferably taken in both arms.

From Maron BJ, Thompson PD, Ackerman MJ, et al. Recommendations and considerations related to preparticipation screening for cardiovascular abnormalities in competitive athletes: 2007 update: a scientific statement from the American Heart Association Council on Nutrition, Physical Activity, and Metabolism: endorsed by the American College of Cardiology Foundation. *Circulation*. 2007;115:1643–1655.

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# Chapter 8

## Dermatology

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 See additional content on Expert Consult

### I. EVALUATION AND CLINICAL DESCRIPTIONS OF SKIN FINDINGS

#### A. Primary Skin Lesions

1. **Macule:** Small, flat, well-circumscribed discolored lesion (<1 cm)
2. **Patch:** Large macule (>1 cm)
3. **Papule:** Small, elevated, firm, well-circumscribed superficial lesion (<1 cm)
4. **Plaque:** Large papule (>1 cm)
5. **Pustule:** Small, well-circumscribed elevation of skin containing purulent material (<1 cm)
6. **Vesicle:** Small, well-circumscribed elevation of skin containing serous fluid (<0.5 cm)
7. **Bulla:** Large vesicle (>0.5 cm)
8. **Wheal:** Transient, raised, well-circumscribed lesion with erythematous periphery and central pallor
9. **Nodule:** Soft or firm lesion in dermis or subcutaneous fat (>1 cm)
10. **Tumor/mass:** Solid, firm lesion (typically >2 cm)

#### B. Secondary Skin Lesions

1. **Scale:** Small, thin plates shedding from the surface of the skin
2. **Crust:** Solidified exudative material from erosions or ruptured vesicles/pustules
3. **Erosion:** Loss of the most superficial layers of the epidermis from friction, pressure, or inflammation
4. **Ulcer:** Full thickness loss of the epidermis and dermis, with clearly defined edges
5. **Fissure:** Linear or wedge-shaped epidermal tear associated with inflammation and pain
6. **Excoriation:** Superficial linear abrasions secondary to scratching
7. **Lichenification:** Thickening of the epidermis with accentuated skin lines, secondary to chronic inflammation and/or scratching
8. **Scar:** Formation of new connective tissue after full thickness injury to skin, leaving permanent change in skin

#### C. Shapes and Arrangements

1. **Linear:** Distributed along a line
2. **Dermatomal:** Following a dermatome
3. **Filiform:** Thread-like

4. **Serpiginous:** Wavy, coiled, serpentine pattern
5. **Annular:** Ring-like configuration
6. **Nummular/discoid:** Disk-like lesion
7. **Targetoid:** Resembling a bull's eye target with central erythema surrounded by pale edema with a peripheral border of erythema
8. **Clustered:** Lesions in a group
9. **Herpetiform:** Clustered vesicular lesions on erythematous bases
10. **Reticulated:** Net or lacey distribution
11. **Geographic:** Resembling outlines on a map such as a continent
12. **Morbilliform:** Eruption of erythematous to dusky coalescing macules with interspersed healthy skin

## II. VASCULAR ANOMALIES<sup>1</sup>

### A. Vascular Tumors

1. Infantile hemangiomas (Fig. 8.1, Color Plates).<sup>2,3</sup>
  - a. **Pathogenesis:** Benign vascular tumor with rapid proliferation followed by spontaneous involution. Most present before 4 weeks of age. Undergo rapid growth between 1 and 2 months of age, with 80% of size reached by 3 months. Most begin to regress between 6 and 12 months of age, with the majority of tumor regression occurring by 4 years of age. 50% to 70% resolve completely.
  - b. **Clinical presentation:** Newborns may demonstrate pale macules with threadlike telangiectasias that later develop into hemangiomas. May be superficial, deep, or mixed. After involution, can have residual skin changes including scarring and atrophy.
  - c. **Indicators that should prompt consideration for early treatment:**
    - (1) Potential for life-threatening complications: Airway hemangiomas, liver hemangiomas (associated with high-output heart failure and severe hypothyroidism), and profuse bleeding from an ulcerated hemangioma.
    - (2) Risk of functional impairment: Interference with the development of vision (if near eye) and interference with feeding (if near mouth).
    - (3) Ulceration: Most common complication (5% to 21%). Can be extremely painful and usually scars; risk greatest in large hemangiomas and those located in skin creases, particularly the diaper area.
    - (4) Associated structural anomalies: PHACES syndrome (**P**osterior cranial fossa malformations, **H**uge hemangiomas, **A**rterial lesions, **C**ardiovascular anomalies (aortic anomalies), **E**ye anomalies, **S**ternal cleft anomalies/supraumbilical raphe<sup>4</sup>) and LUMBAR syndrome (**L**ower body hemangioma, **U**rogenital anomalies, **M**ulberry hemangioma, **B**ony deformities, **A**norectal malformations, **A**rterial anomalies, **R**enal anomalies).
    - (5) Potential for disfigurement: Risk of permanent scarring or distortion of anatomic landmarks.

**TABLE 8.1****INDICATIONS TO OBTAIN IMAGING OF INFANTILE HEMANGIOMAS**

Indication	Imaging Modality
1. Diagnosis of infantile hemangiomas (IH) is uncertain (e.g., atypical appearance or behavior)	Ultrasound with Doppler
2. Five or more cutaneous IH	Abdominal ultrasound with Doppler (screen for hepatic IH)
3. Associated structural abnormalities (e.g., PHACE syndrome or LUMBAR syndrome) are suspected	<ul style="list-style-type: none"> <li>1. If PHACE syndrome is suspected, MRI/MRA head/neck with and without contrast; echocardiography</li> <li>2. If LUMBAR syndrome is suspected, spinal ultrasound and abdominal ultrasound with Doppler are initial screen, with MRI likely to follow</li> <li>3. May wish to consult with hemangioma specialist on exact imaging to be ordered</li> </ul>

From Krowchuk D, Frieden IJ, Mancini AJ, et al. Clinical practice guideline for the management of infantile hemangiomas. *Pediatrics*. 2019;143(1):1–28.



- d. **Diagnosis:** Usually diagnosed clinically. Atypical clinical findings, growth pattern, and equivocal imaging should prompt tissue biopsy to exclude other neoplasms or unusual vascular malformations. See **Table 8.1** for indications to order imaging.
- e. **Treatment:**
- (1) Most are uncomplicated and can be observed with watchful waiting. Photo documentation is used to follow the growth and regression process.
  - (2) If an infantile hemangioma is identified as high risk, the child should be evaluated by a hemangioma specialist promptly, as there is a narrow window of opportunity in which to intervene and prevent poor outcomes.
  - (3)  $\beta$ -adrenergic blockers such as propranolol are considered first-line therapy for complicated infantile hemangiomas and should be initiated under supervision of a pediatric dermatologist or experienced practitioner.<sup>5</sup> While patients should be clinically screened for cardiac disease, EKG and/or echocardiogram are not required unless there is clinical concern. Contraindications include: Reactive airways, sinus bradycardia, decompensated heart failure, greater than 1st degree heart block, hypotension, hypoglycemia, hypersensitivity to propranolol. Off label use of selective beta-blockers may be considered in certain patients. Duration should be at least 6 months and up to 12 months of age.<sup>6</sup>
  - (4) Corticosteroids are considered second line. Similar efficacy to propranolol in a prospective, randomized, investigator-blinded trial, but propranolol is better tolerated and with fewer severe side effects.<sup>7</sup>
  - (5) Topical timolol is effective in superficial, uncomplicated hemangiomas (recommend 0.5% gel forming solution).

2. Pyogenic granuloma (Lobular Capillary Hemangioma) ([Fig. 8.2](#), Color Plates)
  - a. **Clinical presentation:** Benign vascular tumor, appears as small (usually 3 to 10 mm but occasionally much larger) bright red papule that grows over several weeks to months into sessile or pedunculated papule with a “collarette,” scale, or crust. Can bleed profusely with minor trauma and can ulcerate. Rarely spontaneously regresses. Seen in all ages; average age of diagnosis 6 months to 10 years. Located on head and neck, sometimes in oral mucosa but can be at any skin site and often misdiagnosed as hemangiomas.
  - b. **Treatment:** Usually required, given frequent bleeding and ulceration. Options include shave excision or curettage with cautery of base, surgical excision, carbon dioxide laser excision, or pulsed dye laser therapy. For most cases, shave and cautery are quick, safe, low risk, and can be performed quickly with local anesthesia.

### B. Vascular Malformations

Include capillary (port-wine stains and salmon patch/stork bite/angel kiss), lymphatic, venous, and arteriovenous malformations.

Note: For a comparison of vascular malformations to vascular tumors, please see [Table EC 8.A.8](#)

## III. INFECTIONS

### A. Viral

1. Warts
  - a. **Pathogenesis:** Human papillomaviruses (HPVs) of the epithelium or mucus membrane.
  - b. **Clinical presentation:**
    - (1) Common warts: Skin-colored, rough, minimally scaly papules and nodules found most commonly on the hands, although can occur anywhere. Can be solitary or multiple, range from a few millimeters to several centimeters, may form large plaques or a confluent linear pattern secondary to autoinoculation. Sometimes persistent in immunocompromised patients.
    - (2) Flat warts: Flesh to brown/yellow-colored, smooth, flat-topped papules commonly found over the hands, arms, and face. Usually <2 mm in diameter and often present in clusters.
    - (3) Plantar warts: Occur on soles of feet as inward growing, hyperkeratotic plaques and papules. Trauma on weight-bearing surfaces results in small black dots (petechiae from thrombosed vessels on the surface of the wart). Can be painful.
  - c. **Diagnosis:** Clinical diagnosis.
  - d. **Treatment**<sup>9</sup>:
    - (1) Spontaneous resolution occurs in greater than 75% of warts in otherwise healthy individuals within 3 years. No specific treatment clearly better than placebo, except possibly topical salicylic acid.

**TABLE EC 8.A****DIFFERENTIATING VASCULAR TUMORS AND VASCULAR MALFORMATIONS**

Vascular tumor (infantile hemangioma, pyogenic granuloma, kaposiform hemangioendothelioma, tufted angioma, other tumors)	Vascular malformation (venous, arterial, AVM, capillary, lymphatic)
<ul style="list-style-type: none"><li>• Usually not present at birth</li><li>• Dynamic</li><li>• Regressing</li><li>• Proliferative</li></ul>	<ul style="list-style-type: none"><li>• Present at birth</li><li>• Static</li><li>• Persistent</li><li>• Non-proliferative</li></ul>

AVM, Arteriovenous malformation.

Adapted from Cohen BA, Rozell-Shannon L. Early diagnosis and intervention of vascular anomalies (infantile hemangiomas and malformations). *Pediatric Care Online*. <http://pediatriccare.solutions.aap.org>. Accessed September 2018.





- (2) Keratolytics (topical salicylates): Particularly effective in combination with adhesive tape occlusion. Response may take 4 to 6 months.
- (3) Destructive techniques, candida antigen, cantharidin, or “beetle juice” are not clearly more effective than placebo. Additionally, destructive techniques can be painful and cause scarring. These options are not recommended in children.
2. Molluscum contagiosum (**Fig. 8.3**, Color Plates)
- Pathogenesis:** Large DNA poxvirus. Spread by skin-to-skin contact.
  - Clinical presentation:** Dome-shaped, often umbilicated, translucent to white papules that range from 1 mm to 1 cm. Occur anywhere except palms and soles, most commonly on the trunk and intertriginous areas. Can occur in the genital area and lower abdomen when obtained as a sexually transmitted infection. May be pruritic and can be surrounded by erythema, resembling eczema.
  - Diagnosis:** Clinical diagnosis.
  - Treatment:** Most spontaneously resolve within 6 to 18 months and do not require intervention other than monitoring for secondary bacterial infection. Surrounding eczematous changes may indicate an immunologic reaction and serve as a harbinger of regression. Treatment may cause scarring and may not be more effective than placebo. Recurrences are common.
3. Herpes simplex virus
- Pathogenesis:** Either HSV-1 or HSV-2 may be implicated, regardless of lesion location. During the initial outbreak, oral lesions last 2 to 3 weeks whereas genital lesions may last 2 to 6 weeks. Recurrent episodes are usually much shorter.
  - Clinical presentation (**Fig. 8.6**, Color Plate):** Symptoms include prodrome of tingling, itching, or burning followed by painful vesicles on erythematous base that may last 7 to 10 days, break open, and crust prior to healing, flu-like symptoms, dehydration (gingivostomatitis), dysuria (genital), ophthalmologic symptoms (keratitis). May be triggered by stress, illness, sun exposure, and menstruation. The first outbreak is typically the worst.
  - Diagnosis:** Diagnosed clinically and, in many centers, with viral DNA PCR (more sensitive than culture). To culture a lesion, clean with alcohol, unroof lesion with sterile needle or wooden side of cotton swab, collect vesicular fluid on sterile swab, and send in viral transport medium.
  - Treatment:** Acyclovir or valacyclovir for 7 to 14 days (see Formulary for dosing). For children with herpetic gingivostomatitis, antiviral therapy should be initiated within 72 to 96 hours of onset if they are unable to drink or have significant pain. Valacyclovir is generally preferred as it is more bioavailable than acyclovir and, as a result, is dosed less frequently.

4. Erythema infectiosum (“fifth disease”)
  - a. **Pathogenesis:** Parvovirus B19.
  - b. **Clinical presentation:** Pediatric presentation of nonspecific febrile illness with headache, coryza, and gastrointestinal complaints. Two to five days after onset of symptoms, the classic malar rash with “slapped cheek” appearance erupts, followed by a reticular rash to the trunk several days later. Associated signs and symptoms include arthralgias (more common in adults) and a transient aplastic crisis, which may be more of a problem in patients with hemoglobinopathies and pregnant women.
  - c. **Diagnosis:** Clinical diagnosis, serum IgM, or serum DNA PCR.
  - d. **Treatment:** Supportive care and avoiding contact with pregnant women.
5. Pityriasis rosea
  - a. **Pathogenesis:** Viral etiology (possibly HHV-6, HHV-7) has been hypothesized but no definitive cause has been described.
  - b. **Clinical presentation:** Typically asymptomatic or may have mild pruritus. Classic presentation with a round to oval, sharply demarcated, scaly, salmon-colored herald patch with central clearing on trunk followed by a “Christmas tree” distribution of oval crops of lesions similar to herald patch. Pediatric patients may have an atypical distribution involving the scalp, face, distal extremities, and sparing of the trunk. Lesions typically resolve in 4 to 6 weeks.
  - c. **Diagnosis:** Clinical diagnosis.
  - d. **Treatment:** Typically self-resolving.
6. Roseola infantum ([Fig. 8.5](#), Color Plates)
  - a. **Pathogenesis:** Human Herpesvirus 6 (HHV-6).
  - b. **Clinical presentation:** Typically diagnosed in children <2 years old with peak 7 to 13 months. Febrile phase of 3 to 5 days of high fever (often >40°C), viremia, and irritability. As febrile phase resolves, patients develop a morbilliform rash on neck and trunk that spreads centripetally to face and extremities for 1 to 2 days.
  - c. **Diagnosis:** Clinical diagnosis.
  - d. **Treatment:** Self-resolving.
7. Hand, foot, and mouth disease
  - a. **Pathogenesis:** Most commonly Coxsackievirus A serotypes.
  - b. **Clinical presentation:** Oral lesions on the tongue, buccal mucosa, and palate that initially are 1 to 5 mm erythematous macules and evolve to vesicles and ulcers with a thin erythematous halo. Erythematous, non-pruritic 1 to 10 mm macules, papules, and/or vesicles on the palms and soles. Typically resolve in 3 to 4 days. Usually non-tender, unless caused by Coxsackie A6 (associated with high fevers, widespread lesions, longer duration [12 days], palmar and plantar desquamation, and nail dystrophy).
  - c. **Diagnosis:** Clinical diagnosis.
  - d. **Treatment:** Supportive care.



## 8. Reactive erythema (Fig. 8.4; Figs. 8.5–8.10, Color Plates)

- Pathogenesis:** Represent cutaneous reaction patterns triggered by endogenous and environmental factors (e.g., viral infections, drug reactions).
- Clinical presentation:** Group of disorders characterized by erythematous patches, plaques, and nodules that vary in size, shape, and distribution.

## B. Parasitic

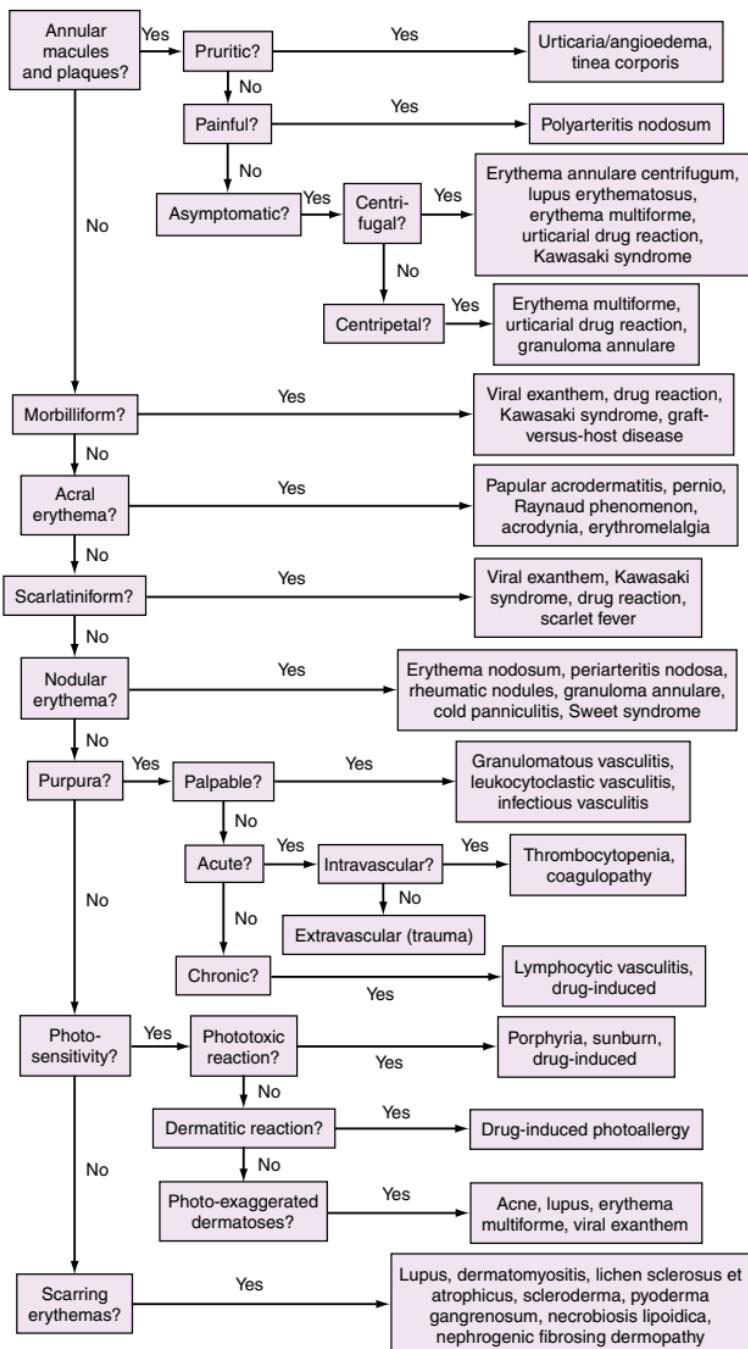
### 1. Scabies (Fig. 8.11, Color Plates)

- Pathogenesis:** Caused by the mite *Sarcoptes scabiei*. Spread by skin-to-skin contact and through fomites. Can live for 2 days away from a human host. Female mites burrow and lay eggs under the skin.
- Clinical presentation:** Initial lesion is a small, erythematous papule that is easy to overlook. Can have burrows (elongated, edematous papulovesicles, often with a pustule at the advancing border) which are pathognomonic. Most commonly located in interdigital webs, wrist folds, elbows, axilla, genitals, buttocks, and belt line. In temperate climates, the face and scalp are usually spared. In young infants, the palms and soles are also commonly involved. Burrows are most dramatic in patients who are unable to scratch (e.g., infants). Disseminated eczematous eruption results in generalized severe pruritus, especially at night. Can become nodular, particularly in intertriginous areas, or be susceptible to superinfection due to frequent excoriations. Immunosuppressed patients may develop diffuse scaly crusted eruption and lack pruritus.
- Treatment<sup>10</sup>:**
  - Permethrin cream: 5% cream applied to skin from neck down in normal hosts including under fingernails and toenails. Rinse off after 8 to 14 hours. Can repeat in 7 to 10 days.
  - Ivermectin (off-label use): Single dose; can repeat in 2 weeks. Efficacy comparable to permethrin cream. May be the best choice for immunodeficient patients where total body application may be difficult.
  - Environment: Mites cannot live away from human skin for more than 2 to 3 days. Launder clothing and sheets. Bag and seal stuffed animals and pillows for 2 to 3 days. Consider treatment of close contacts.

## C. Fungal (Figs. 8.12–8.16, Color Plates)

### 1. Tinea capitis (see Fig. 8.12, Color Plates)

- Pathogenesis:** Mostly caused by fungi of the genus *Trichophyton* in North America (95%), less commonly *Microsporum* (5% or less), and spread through contact and fomites.
- Epidemiology:** Usually occurs in young children, with higher incidence in African American children, but any age and ethnicity can be affected.

**FIGURE 8.4**

Reactive erythema. (Modified from Cohen BA. *Atlas of Pediatric Dermatology*. 4th ed. China: Elsevier Limited; 2013:206.)



- c. **Clinical presentation:**
    - (1) Black dot: Most common. Slowly growing, erythematous, scaling patches. These areas develop alopecia and black dots are visible on scalp where hair has broken.
    - (2) Gray patch ("seborrheic dermatitis type"): Erythematous, scaling, well-demarcated patches that grow centrifugally. Hair breaks off a few millimeters above the scalp and takes on a gray/frosted appearance.
    - (3) Kerion (see Fig. 8.13, Color Plates): Complication of tinea capitis or tinea corporis. Type IV hypersensitivity to fungus. Raised, boggy/spongy lesions, often tender and covered with purulent exudate. Most commonly occurs months after primary infection.
    - (4) All can be associated with posterior cervical lymphadenopathy.
  - d. **Diagnosis:** Can be made clinically, but since oral antifungal therapy is indicated, tinea capitis should be confirmed by direct microscopic exam of a potassium hydroxide (KOH) preparation of the proximal ends of hairs, gently and painlessly scraped from the affected area. Cultures may be obtained by using a sterile toothbrush or cotton swab. The scale can be scraped directly into a sterile plastic cup and/or the cotton swab tips can be broken off and placed into the sterile plastic cups.
  - e. **Treatment**<sup>11</sup>: Always requires systemic therapy. First-line therapy includes oral griseofulvin for 10 to 12 weeks (which should be taken with fatty foods for improved absorption) and terbinafine for 6 weeks (see Formulary for dosing). Most experts consider terbinafine superior to griseofulvin for *T. tonsurans* because of its shorter duration of therapy and superior effectiveness. The FDA recommends baseline and follow-up hepatic function testing in children taking terbinafine, though most clinicians forego laboratory testing in healthy children without history of liver disease if treatment is 6 weeks or less. Though not FDA-approved for tinea capitis, fluconazole at 6 mg/kg/day (max 400 mg/day) for 6 weeks is recommended by the AAP Red Book as an alternative treatment of tinea capitis in children younger than 2 years old.<sup>12</sup> All family members, particularly other children, should be examined carefully for subtle infection. Selenium sulfide 2.5% shampoo may shorten the period of shedding of fungal organisms and reduce risk of infection of unaffected family members.
2. Tinea corporis and pedis<sup>11</sup> (see Figs. 8.14 and 8.15, Color Plates)
- a. **Pathogenesis:** Spread through direct contact and fomites, especially in sports with close contact.
  - b. **Clinical presentation:** Pruritic, erythematous, annular patch or plaque with central clearing and a scaly raised border. Typically affects glabrous skin (smooth and bare).
  - c. **Diagnosis:** Usually diagnosed clinically, but a KOH preparation or fungal culture can be used to help guide diagnosis.
  - d. **Treatment:** Topical antifungals (terbinafine, azole) through 1 to 2 weeks past lesion resolution. Widespread eruption may require oral antifungals.

3. Tinea versicolor (see Fig. 8.16, Color Plates)
  - a. **Pathogenesis:** Caused by *Malassezia*. Exacerbated by hot/humid weather, hyperhidrosis, topical skin oil use. Most people are colonized with *Malassezia* but only a small number are prone to develop clinical lesions. Not associated with poor hygiene. Not contagious.
  - b. **Clinical presentation:** Well demarcated, minimally scaly, hypopigmented macules or patches. Hypopigmented areas tend to be more prominent in the summer because affected areas do not tan. Lesions often have a fine scale that may be noted following gentle rubbing and can be mildly pruritic but are usually asymptomatic.
  - c. **Diagnosis:** KOH microscopy reveals pseudohyphae and yeast cells that appear like “spaghetti and meatballs.”
  - d. **Treatment:** Topical antifungal shampoos and/or creams (miconazole, oxiconazole, ketoconazole) or selenium sulfide are effective. Given the risk of hepatotoxicity, oral azole antifungals are reserved for resistant or widespread disease. Oral terbinafine is not effective. Pigmentation changes may take months to resolve despite successful treatment.

#### D. Bacterial

1. Impetigo
  - a. **Pathogenesis:** Contagious bacterial infection of the skin, most commonly caused by *Staphylococcus aureus* (99% MSSA), with a minority of cases caused by Group A *Streptococcus*.
  - b. **Clinical presentation:**
    - (1) Nonbullous impetigo: Papules that evolve into erythematous pustules or vesicles that break and form thick, honey-colored crusts and plaques. Commonly overlying any break to skin barrier. Primarily face and extremities.
    - (2) Bullous impetigo: Painless vesicles that evolve into flaccid bullae and crusted patches with undermined border. Seen more in infants and young children. Caused by *Staphylococcus aureus* exfoliative toxin A.
  - c. **Diagnosis:** Clinical diagnosis.
  - d. **Treatment:** When impetigo is contained to a small area, topical mupirocin may be used for 5 days. When the infection is widespread, an oral antibiotic such as cephalexin should be used for 7 days. Consider broader coverage if MRSA is suspected, although MSSA accounts for most infections.
2. Staph scalded skin syndrome
  - a. **Pathogenesis:** *Staphylococcus aureus* infections of the skin with hematogenous dissemination of exfoliative toxin A or B to the epidermis.
  - b. **Clinical presentation:** Typical presentation is Ritter disease (generalized exfoliation) in a 3- to 7-day-old infant who initially is febrile and irritable with conjunctivitis and perioral erythema. In addition to newborns, this presentation is seen in young children who do not have antibodies to the toxin and often do not clear the toxin-antibody complex quickly due to decreased renal excretion. One to two days after the prodromal onset, patient develops diffuse erythema, fragile,

flaccid bullae and erosions that are Nikolsky positive in areas of mechanical stress such as intertriginous areas. Lesions are not scarring as they are intraepidermal. Older children tend to have a localized bullous impetigo with tender scarlatiniform eruption. Infants and toddlers usually have a combination of the presentations seen in neonates and older children along with white to brown thick flaking desquamation of the entire body, especially the face and neck.

- c. **Diagnosis:** Typically clinically. However, cultures should be obtained from any potential source site of infection or colonization such as the medial canthi or nares.
  - d. **Treatment:** Nearly all cases are MSSA, with an increasing number being clindamycin resistant. First-line treatment may include oral penicillinase-resistant beta-lactams such as first or second-generation cephalosporins. Vancomycin should be considered in patients who fail to respond to treatment and/or in areas with a high prevalence of MRSA. Management should also include supportive care with topical emollients and close monitoring of fluid and electrolyte status.
3. Scarlet fever ([Fig. 8.17](#), Color Plates)
- a. **Pathogenesis:** Exotoxin-mediated response to a *Streptococcus pyogenes* infection, typically pharyngitis.
  - b. **Clinical presentation:** Sandpaper-like, coarse, erythematous, blanching rash that originates in the groin and axilla then spreads to the trunk then extremities but spares the palms and soles. May have Pastia lines. Associated with pharyngitis, circumoral pallor, and a strawberry tongue.
  - c. **Diagnosis:** Clinical diagnosis. May benefit from rapid strep test and throat culture.
  - d. **Treatment:** No additional treatment aside from treating the patient's Strep pharyngitis.
4. Cellulitis: See [Chapter 17](#).



#### IV. HAIR LOSS (FIGS. 8.18–8.20, COLOR PLATES)

##### A. Telogen Effluvium (see [Fig. 8.18](#), Color Plates)

- 1. **Pathogenesis:** Most common cause of diffuse hair loss. Mature hair follicles switch prematurely to the telogen (resting) state, with shedding within 3 months.
- 2. **Clinical presentation:** Diffuse hair thinning 3 months after a stressful event (major illnesses or surgery, pregnancy, severe weight loss).
- 3. **Treatment:** Self-limited. Regrowth usually occurs over several months.

##### B. Alopecia Areata (see [Fig. 8.19](#), Color Plates)

- 1. **Clinical presentation:** Chronic inflammatory (probably autoimmune) disease that starts with well-circumscribed small bald patches and normal-appearing underlying skin. New lesions may demonstrate subtle erythema and be pruritic. Bald patches may enlarge to involve large areas of the scalp or other hair-bearing areas. Many experience good

hair regrowth within 1 to 2 years, although most will relapse. A minority progress to total loss of all scalp (alopecia totalis) and/or body hair (alopecia universalis).

2. **Diagnosis:** Usually clinical diagnosis.
3. **Treatment<sup>13</sup>:** First-line therapy is topical steroids. Referral to dermatology is warranted for consideration of other treatments. No evidence-based data that any therapy is better than placebo. Older children, adolescents, and young adults with longstanding localized areas of hair loss have the best prognosis.

#### C. Traction Alopecia (see Fig. 8.20, Color Plates)

1. **Pathogenesis:** Hairstyles that apply tension for long periods of time.
2. **Clinical presentation:** Noninflammatory linear areas of hair loss at margins of hairline, part line, or scattered regions, depending on hairstyling procedures used.
3. **Treatment:** Avoidance of styling products or styles that result in traction. If traction remains for long periods, condition may progress to permanent scarring hair loss.

#### D. Trichotillomania and Hair Pulling

1. **Pathogenesis:** Alopecia due to compulsive urge to pull out one's own hair, resulting in irregular areas of incomplete hair loss. Mainly on the scalp; can involve eyebrows and eyelashes. Onset is usually after age 10 and should be distinguished from hair twirling/pulling in younger children that resolves without treatment in most cases.
2. **Clinical presentation:** Characterized by hair of differing lengths; area of hair loss can be unusual in shape.
3. **Treatment:** Behavioral modification and consider psychiatric evaluation (can be associated with anxiety, depression, and obsessive-compulsive disorder).

## V. ACNE VULGARIS

#### A. Pathogenetic Factors

Follicular hyperkeratinization, increased sebum production, *Cutibacterium acnes* (formerly *Propionibacterium acnes*) proliferation, and inflammation.

#### B. Risk Factors

Androgens, family history, and stress. No strong evidence that dietary habits affect acne.

#### C. Clinical Presentation

1. **Noninflammatory lesions**
  - a. Closed comedone (whitehead): Accumulation of sebum and keratinous material, resulting in white/skin-colored papules without surrounding erythema.
  - b. Open comedone (blackhead): Dilated follicles filled with keratinocytes, oils, and melanin.



2. **Inflammatory lesions:** Papules, pustules, nodules, and cysts with evidence of surrounding inflammation. Typically appear later in the course of acne. Nodulocystic presentations are more likely to lead to hyperpigmentation and/or permanent scarring.

#### D. Treatment<sup>14–16</sup> (Table 8.2)

- Skin care:** Gentle nonabrasive cleaning. Avoid picking or popping lesions. Vigorous scrubbing and abrasive cleaners can worsen acne.
- Topical first-line therapies:** Recommended for mild to moderate acne.
  - Retinoids** (Table EC 8.B)
    - Normalize follicular keratinization and decrease inflammation.
    - A pea-sized amount should be applied to cover the entire face.
    - Risks: Cause irritation and dryness of skin. Retinoids should be used at night due to inactivation by sunlight. This class should not be used during pregnancy.
    - Three topical retinoids (tretinoin, adapalene, and tazarotene) are available by prescription in the United States. Adapalene 0.1% gel has been approved for over-the-counter (OTC) use with significant efficacy.<sup>17</sup>

TABLE 8.2

PEDIATRIC TREATMENT RECOMMENDATION FOR MILD, MODERATE, AND SEVERE ACNE

Acne Classification	Initial Treatment	Inadequate Response
Mild	Benzoyl peroxide (BPO) or topical retinoid <b>OR</b> <i>Topical combination therapy:</i> BPO + Antibiotic or Retinoid + BPO or Retinoid + Antibiotic + BPO	Add BPO or retinoid if not already prescribed OR change topical retinoid concentration, type, and/or formulation OR change topical combination therapy
Moderate	<i>Topical combination therapy:</i> Retinoid + BPO or Retinoid + BPO + Antibiotic <b>OR</b> Oral Antibiotic + Topical Retinoid + BPO or Topical Retinoid + Topical Antibiotic + BPO	Change topical retinoid concentration, type, and/or formulation and/or change topical combination therapy OR add or change oral antibiotic. Consider oral isotretinoin (dermatology referral). Females: consider hormonal therapy.
Severe	<i>Combination therapy:</i> Oral Antibiotic + Topical Retinoid + BPO ± Topical Antibiotic	Consider changing oral antibiotic AND consider oral isotretinoin. Females: consider hormonal therapy. Strongly consider referral to dermatology.

Topical fixed-combination prescriptions available.

Data from Eichenfield LF, Krakowski AC, Piggott C, et al. Evidence-based recommendations for the diagnosis and treatment of pediatric acne. *Pediatrics*. 2013;131:S163–S186.

**TABLE EC 8.B****FORMULATIONS AND CONCENTRATIONS OF TOPICAL RETINOIDS**

Retinoid	Vehicle <sup>a</sup>	Strength (%)
TRETINOIN Pregnancy Category C	Cream	0.025, 0.05, 0.1
	Gel	0.01, 0.025
	Gel (micronized)	0.05
	Microsphere gel	0.04, 0.1
	Polymerized cream	0.025
	Polymerized gel	0.025
ADAPALENE Pregnancy Category C	Cream	0.1
	Gel	0.1, 0.3
	Solution	0.1
	Lotion	0.1
TAZAROTENE Pregnancy Category X	Gel	0.05, 0.1
	Cream	0.05, 0.1

<sup>a</sup>Numerous generic retinoids are available. Branded products are available under the following trade names: Atralin, Avita, and Retin-A Micro for tretinoin; Differin for adapalene; and Tazorac for tazarotene.

Data from Eichenfield LF, Krakowski AC, Piggott C, et al. Evidence-based recommendations for the diagnosis and treatment of pediatric acne. *Pediatrics*. 2013;131:S163–S186, Table 4.



- b. Benzoyl peroxide (BPO)
  - (1) Oxidizing agent with antibacterial and mild anticomедolytic properties.
  - (2) Washes may be most convenient formulation, as they can be used in the shower.
  - (3) Risks: Can bleach hair, clothing, towels, and sheets.
- c. Salicylic acid: Topical comedolytic agent that may be found in OTC face washes and serves as an alternative to a topical retinoid.

**3. Topical antimicrobials:**

- a. Azelaic acid: Antimicrobial, comedolytic, and anti-inflammatory. Recommended by the American Academy of Dermatology (AAD) for the treatment of postinflammatory dyspigmentation (see [Figures EC 8.P](#) and [EC 8.S](#)). Available in a 15% gel and a 20% cream (more efficacious).<sup>18</sup>
- b. Erythromycin and clindamycin: Avoid topical antibiotics as monotherapy. Topical BPO should be concurrently used to optimize efficacy and avoid bacterial resistance.

**4. Oral antibiotics (Table EC 8.C):** Recommended for moderate to severe inflammatory acne that is resistant to topical treatment. These medications should be used with BPO or topical retinoid. Do not use as monotherapy. Limit to 3 months to minimize bacterial resistance.

- a. ≥8 years old: Doxycycline or minocycline
- b. <8 years old, pregnancy, or tetracycline allergy: Azithromycin, erythromycin, or trimethoprim/sulfamethoxazole.
- c. Erythromycin should be used with care due to increased risk of resistance. The AAD recommends reserving trimethoprim/sulfamethoxazole for patients who have failed other treatments or are unable to tolerate tetracyclines and macrolides.

**5. Hormonal therapy:** Reduces sebum production and androgen levels.

Good option for pubertal females who have sudden onset of moderate to severe hormonal acne (often on lower face, jawline) and have not responded to conventional first-line therapies. Should not be used as monotherapy. Combination oral contraceptives (Ortho Tri-Cyclen, Estrostep, and Yaz) or spironolactone (antiandrogen).

**6. Oral isotretinoin:** Reserved for patients with severe nodular, cystic, or scarring acne who do not respond to traditional therapy or who cannot be weaned from oral antibiotics. Should be managed by a dermatologist. Most patients have complete resolution of their acne after 16 to 20 weeks of use.

a. Side effects:

- (1) Teratogenicity: Patients and physicians are mandated by the FDA to comply with the iPledge program to eliminate fetal exposure to isotretinoin. Female patients with child-bearing potential must use two forms of birth control with routine pregnancy testing.
- (2) Hepatotoxicity, hyperlipidemia, and bone marrow suppression, a complete blood cell count, fasting lipid profile, and liver function tests should be obtained before initiation of therapy and repeated at 4 and 8 weeks.

**TABLE EC 8.C****FIRST-LINE ORAL ANTIBIOTICS USED FOR TREATMENT OF MODERATE TO SEVERE ACNE VULGARIS**

Antibiotic	Potential Adverse Effects	Comments
DOXYCYCLINE	Pill esophagitis; photosensitivity; staining of forming tooth enamel (<8 years of age); vaginal candidiasis	Take with large glass of water and maintain upward position ~1 hr; optimize photoprotection; avoid in children without permanent teeth
MINOCYCLINE (IMMEDIATE RELEASE)	Cutaneous and/or mucosal hyperpigmentation; DHS (systemic, within first 1–2 months); LLS; SJS; vestibular toxicity (within first few days); staining of forming tooth enamel (<8 years of age); vaginal candidiasis	Can be taken with meals; warn patient about dizziness/vertigo; avoid in children without permanent teeth; monitor for pigmentary changes on skin
MINOCYCLINE (EXTENDED RELEASE)	Same as above although above side effects reported predominantly with immediate release formulations; lower incidence of acute vestibular side effects with weight-based dosing	Less accumulation of drug over time due to pharmacokinetic properties of extended release formulation, may correlate with decreased hyperpigmentation

DHS, Drug hypersensitivity syndrome; LLS, lupus-like syndrome; SJS, Stephens-Johnson syndrome.

Modified from Eichenfield LF, Krakowski AC, Piggott C, et al. Evidence-based recommendations for the diagnosis and treatment of pediatric acne. *Pediatrics*. 2013;131:S163–S186, Table 5.



## VI. COMMON NEONATAL DERMATOLOGIC CONDITIONS (FIG. 8.21; FIGS. 8.22–8.30, COLOR PLATES)

### A. Erythema Toxicum Neonatorum (see Fig. 8.22, Color Plates)

1. **Clinical presentation:** Most common rash of full-term infants; incidence declines with lower birth weight and prematurity. Appears as small erythematous macules and papules that evolve into pustules on erythematous bases. Rash most often occurs by 24 to 48 hours of life but can be present at birth or emerge as late as 2 to 3 weeks.
2. **Course:** Self-limited, resolves within 5 to 7 days; recurrences possible.

### B. Transient Neonatal Pustular Melanosis (see Figs. 8.23–8.24, Color Plates)

1. **Clinical presentation:** More commonly affects full-term infants with darker pigmentation. At birth, appears as small pustules on non-erythematous bases that rupture and leave erythematous/hyperpigmented macules with a collarette of scale.
2. **Course:** Self-limited macules fade over weeks to months.

### C. Miliaria (Heat Rash) (see Fig. 8.25, Color Plates)

1. **Clinical presentation:** Common newborn rash associated with warmer climates, incubator use, or occlusion with clothes/dressings. Appears as small erythematous papules or pustules usually on face, scalp, or intertriginous areas.
2. **Course:** Rash resolves when infant is placed in cooler environment or tight clothing/dressings are removed.

### D. Milia (see Fig. 8.26, Color Plates)

1. **Clinical presentation:** Common newborn lesions. Appears as 1- to 3-mm white/yellow papules, frequently found on nose and face; due to retention of keratin and sebaceous materials in pilosebaceous follicles.
2. **Course:** Self-limited, resolves within first few weeks to few months of life.

### E. Neonatal Acne (see Fig. 8.27, Color Plates)

1. **Clinical presentation:** Seen in 20% of infants. Appears as inflammatory papules or pustules without comedones, usually on face and scalp. Secondary to effect of maternal and endogenous androgens on infant's sebaceous glands.
2. **Course:** Peaks around 1 month, resolves within a few months, usually without intervention. Does not increase risk of acne as an adolescent.

### F. Seborrheic Dermatitis (Cradle Cap) (see Figs. 8.28–8.29, Color Plates)

1. **Clinical presentation:** Erythematous plaques with greasy yellow scales. Located in areas rich with sebaceous glands, such as scalp, cheeks, ears, eyebrows, intertriginous areas, diaper area. Unknown etiology. Can be seen in newborns, more commonly in infants aged 1 to 4 months.
2. **Course:** Self-limited and resolves within a few weeks to months.



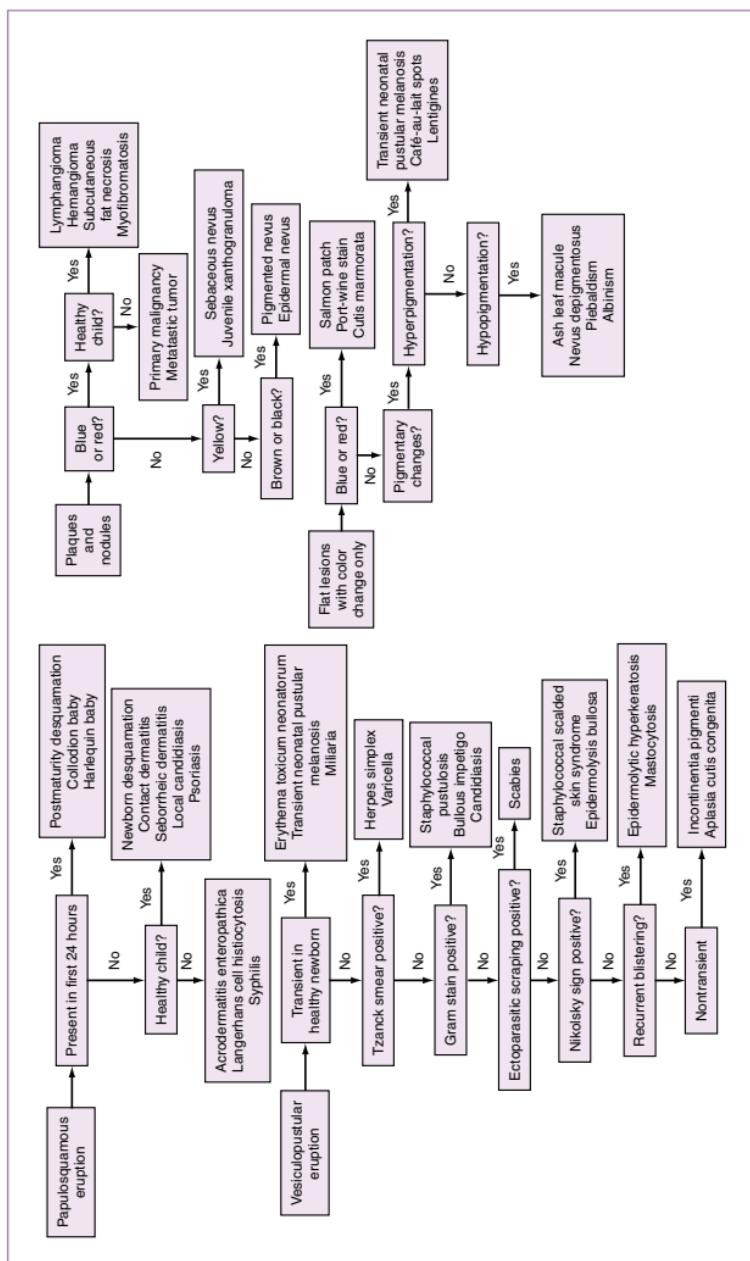


FIGURE 8.21

Evaluation of neonatal rashes. (Modified from Cohen BA. *Atlas of Pediatric Dermatology*. 4th ed. China: Elsevier Limited; 2013:62.)

3. **Treatment:** Can remove scales on scalp with an emollient (e.g., mineral or olive oil, or petroleum jelly) and a soft brush/fine comb. In more severe cases, antifungal shampoos or low-potency topical steroid can shorten the course, although no shampoos are FDA-approved for children less than 2 years of age.

#### G. Congenital Dermal Melanocytosis (formerly known as Mongolian Spots)

1. **Clinical presentation:** Most common pigmented lesion of newborns, usually seen in babies with darker skin tone. Appear as blue/gray macules without definite disappearance of dermal melanocytes. Can be mistaken for child abuse thus accurate documentation at newborn and well-child visits is important.
2. **Course:** Spots typically fade within first few years of life, with majority resolved or much improved by age 10 years.

#### H. Diaper Dermatitis<sup>19</sup> (see Fig. 8.30, Color Plates)

1. **Clinical presentation:** Irritant contact dermatitis characterized by erythematous eruption on buttocks and genital areas with exclusion of other potential causes. Rarely associated with diaper candidiasis, characterized by a red, raised papular rash with small pustules at the periphery. Tends to involve the skin creases.<sup>19</sup>
2. **Treatment:** Frequent diaper changes, air exposure, adequate drying, gentle cleaning, and judicious use of topical barrier preparations. If persistent, can use low-potency topical steroid until cleared. For candidiasis, treatment with topical nystatin, miconazole, or clotrimazole is sufficient. Combination steroid/antifungal creams should be avoided due to steroid-related side effects and association with persistent fungal infections.<sup>20</sup>



## VII. AUTOIMMUNE AND ALLERGIC DERMATOLOGIC CONDITIONS (FIGS. 8.31–8.38, COLOR PLATES)

#### A. Contact Dermatitis

1. **Irritant dermatitis:** Exposure to physical, chemical, or mechanical irritants to the skin. Common irritants include frequent hand washing, hot water, lip-licking, thumb-sucking, and exposure to chemicals, paints, or certain foods like citrus fruits.
2. **Allergic dermatitis** (see Fig. 8.31, Color Plates):
  - a. **Pathogenesis:** Immune reaction to an environmental trigger that comes into contact with the skin. After initial sensitization period of 7 to 10 days in susceptible individuals, an allergic response occurs with subsequent exposures.
  - b. **Common allergens:** *Toxicodendron* spp. (poison ivy, oak, sumac), nickel, cobalt, gold, dyes, fragrances, formaldehyde, and latex.
  - c. **Clinical presentation:** Pruritic erythematous dermatitis that can progress to chronic scaling, lichenification, and pigment changes. Poison ivy (see Fig. 8.32, Color Plates): Exposure to urushiol causes streaks of erythematous papules, pustules, and vesicles. Highly pruritic, can become edematous, especially if rash is on face or genitals. In extreme cases, anaphylaxis can occur.

- d. **Diagnosis:** Careful history taking and recognition of unusual shapes and locations suggesting an “outside job” allow for clinical diagnosis. Patch testing may also be helpful when trigger cannot be identified.
- e. **Treatment:**
  - (1) Remove causative agent. Moisturize with ointment like Vaseline or Aquaphor twice per day. Use antihistamine and/or oatmeal baths as needed for itching, sedation, and sleeping, though they do not directly impact the rash.
  - (2) Mild/moderate: Topical steroids twice a day for 1 week, then daily for 1 to 2 weeks.
  - (3) Widespread/severe: Systemic steroids for 2 to 3 weeks, with taper. There is no role for short courses of steroids because eruption will flare when drug is stopped.
  - (4) For poison ivy contact, remove clothing and wash skin with mild soap and water as soon as possible.

### B. Atopic Dermatitis (Eczema) (See Figs. 8.33–8.37, Color Plates)

- 1. **Pathogenesis:** Due to inadequate skin barrier function from combination of genetic and environmental factors, resulting in transepidermal water loss. Can be associated with elevated serum IgE.
- 2. **Epidemiology<sup>21</sup>:** Affects up to 20% of children in the United States, the vast majority with onset before age 5 years. Other comorbidities may follow including asthma, allergic rhinitis, and food allergies. Eczema resolves or improves in over 75% of patients by adulthood.
- 3. **Clinical presentation:** Dry, pruritic skin with acute changes, including erythema, vesicles, crusting, and chronic changes, including scaling, postinflammatory hypo- or hyperpigmentation (see Figures EC 8.P and EC 8.S), and lichenification.
  - a. Infantile form: Erythematous, scaly lesions on the cheeks, scalp, and extensor surfaces. Covered areas (especially the diaper area) are usually spared.
  - b. Childhood form: Lichenified plaques in flexural areas.
  - c. Adolescence: More localized and lichenified skin changes. Predominantly on skin flexures, hands, and feet.
- 4. **Treatment<sup>21</sup>:** See Chapter 15
  - a. Lifestyle: Avoiding triggers (products with alcohol, fragrances, astringents, sweat, allergens, and excessive bathing). Avoid scratching (eczema is the “itch that rashes”).
  - b. Bathing: Should be less than 5 minutes in lukewarm water with a gentle bar soap and no washcloth or scrubbing. Skin should be patted dry (not rubbed) and followed by rapid application of an emollient (“soak and smear”).
  - c. Consider diluted bleach baths once or twice a week (mix 1/4 cup of bleach in full tub of lukewarm water and soak for 10 minutes, then rinse off with fresh water).
  - d. Skin hydration: Frequent use of bland emollients with minimal water content (Vaseline or Aquaphor). Avoid lotions, as they have high water and low oil content, which worsens dry skin.



- e. Oral antihistamines: There is little evidence that antihistamines improve skin lesions in atopic dermatitis. Non-sedating antihistamines can be used for environmental allergies and hives. Sedating antihistamines may be of transient benefit for sedation at bedtime.
- f. Treatment for inflammation:
  - (1) **Mild disease:** **Topical steroids**<sup>22</sup> (**Table 8.3**): Low and medium potency steroid ointments once or twice daily for 7 days during a flare. Severe flares may require a high-potency steroid for a longer duration of therapy, followed by a taper to a low-potency steroid. Use of topical steroids in areas where skin is thin (groin, axilla, face, under breasts) should generally be avoided. Short durations of low-potency steroids may be used as needed in these areas. Ointments can be applied over steroid.
  - (2) **Moderate disease:** **Crisaborole** is a topical PDE4 inhibitor approved for mild to moderate eczema with preliminary studies of the 2% ointment showing improvement in the majority of clinical signs and symptoms, particularly pruritus.<sup>23</sup> Topical calcineurin inhibitors (**tacrolimus ointment, pimecrolimus cream**) are second-line therapies which should only be used in consultation with a dermatologist due to FDA “black box” warnings on these medications for theoretical increased risk of cancer, although there are no data to confirm and long-term safety studies are pending.<sup>24,25</sup>
  - (3) **Severe disease:** **Phototherapy** with narrowband UVB light is a treatment option for older children and adolescents. Low-dose **methotrexate** is a consideration before cyclosporine. For many dermatologists, low-dose oral methotrexate is the first oral option for severe disease unresponsive to aggressive topical therapy. Oral **cyclosporine** is only used in severe cases of older children and adolescents who have failed other treatments due to concern for renal compromise. **Dupilumab** is an IL-4 receptor alpha antagonist prescribed for refractory cases, currently with FDA approval only for treatment in adults.

## 5. Complications<sup>26</sup>:

- a. Bacterial superinfection: Usually *S. aureus*, sometimes Group A *Streptococcus*. Depending on extent of infection, treat with topical mupirocin or systemic antibiotics.
- b. Eczema herpeticum superinfection with herpes simplex virus can cause severe systemic infection. Presents as vesiculopustular lesions with central punched-out erosions that do not respond to oral antibiotics. Must be treated systemically with acyclovir or valacyclovir. Should be evaluated by ophthalmologist if there is concern for eye involvement.

## C. Papular Urticaria (See Fig. 8.38, Color Plates)

- 1. **Pathogenesis:** Type IV hypersensitivity reaction to fleas, mosquitos, or bedbugs; also known as insect bite-induced hypersensitivity (IBIH).

**TABLE 8.3****RELATIVE POTENCIES OF TOPICAL CORTICOSTEROIDS**

Class	Drug	Vehicle(s)	Strength (%)
I. VERY HIGH POTENCY	Augmented betamethasone dipropionate	Ointment	0.05
	Clobetasol propionate	Cream, foam, ointment	0.05
	Diflorasone diacetate	Ointment	0.05
	Halobetasol propionate	Cream, ointment	0.05
II. HIGH POTENCY	Amcinonide	Cream, lotion, ointment	0.1
	Augmented betamethasone dipropionate	Cream	0.05
	Betamethasone propionate	Cream, foam, ointment, solution	0.05
	Desoximetasone	Cream, ointment, gel	0.25 (C,O), 0.05 (G)
	Diflorasone diacetate	Cream	0.05
	Fluocinonide	Cream, gel, ointment, solution	0.05
	Halcinonide	Cream, ointment	0.1
III-IV. MEDIUM POTENCY	Mometasone furoate	Ointment	0.1
	Triamcinolone acetonide	Cream, ointment	0.5
	Betamethasone valerate	Cream, foam, lotion, ointment	0.1
	Clocortolone pivalate	Cream	0.1
	Desoximetasone	Cream	0.05
	Fluocinolone acetonide, Fluradrenolide	Cream, ointment	0.025 (C), 0.05 (O)
	Fluticasone propionate	Cream, Ointment	0.05 (C), 0.005 (O)
V. LOWER-MEDIUM POTENCY	Triamcinolone acetonide, Mometasone furoate	Cream	0.1
	Hydrocortisone butyrate	Cream, ointment, solution	0.1
	Hydrocortisone probutate, Prednicarbate	Cream	0.1
VI. LOW POTENCY	Hydrocortisone valerate	Cream, ointment	0.2
	Alclometasone dipropionate	Cream, ointment	0.05
	Desonide	Cream, gel, foam, ointment	0.05
VII. LOWEST POTENCY	Fluocinonide acetonide	Cream, solution	0.01
	Dexamethasone	Cream	0.1
	Hydrocortisone	Cream, lotion	0.25
		Cream, ointment	0.5
		Cream, solution	1
	Hydrocortisone acetate	Cream, ointment	0.5-1

C, Cream; G, gel; O, ointment.

Modified from Eichenfeld LF, Tom WL, Berger TG, et al. Guidelines of care for the management of atopic dermatitis.

Section 2. Management and treatment of atopic dermatitis with topical therapies. *J Am Acad Dermatol*. 2014;71(1):116-132, Table 5.



2. **Clinical presentation/epidemiology:** Summarized by the SCRATCH principles<sup>27</sup>:
  - a. **Symmetric eruption:** Exposed areas and scalp commonly affected. Spares diaper region, palms, and soles.
  - b. **Cluster:** Appear as “meal clusters” or “breakfast, lunch, and dinner” which are linear or triangular groupings of lesions. Associated with bedbugs and fleas.
  - c. **Rover not required:** A remote animal exposure or lack of pet at home does not rule out IBIH.
  - d. **Age:** Tends to peak by age 2. Not seen in newborn period. Most tend to develop tolerance by age 10.
  - e. **Target lesions:** Especially in darkly pigmented patients. **Time:** Emphasize chronic nature of eruption and need for patience and watchful waiting.
  - f. **Confused pediatrician/parent:** Diagnosis often met with disbelief by parent and/or referring pediatrician.
  - g. **Household:** Because of the nature of the hypersensitivity, usually only affects one family member in the household.
3. Management (3 Ps):
  - a. **Prevention:** Wear protective clothing, use insect repellent when outside (AAP guidelines recommend up to 30% DEET or 12% picaridin containing repellents), launder bedding and mattress pads for bedbugs, and maximize flea control for pets.
  - b. **Pruritis control:** Topical steroids or antihistamines may be of some benefit.
  - c. **Patience:** Can be frustrating because of its persistent, recurrent nature. Ensure patients that their symptoms will resolve and they will eventually develop tolerance.

#### D. Stevens-Johnson Syndrome and Toxic Epidermal Necrolysis

1. **Pathogenesis:** Severe mucocutaneous reaction with partial to full epidermal necrosis due to keratinocyte necrosis. Stevens-Johnson syndrome (SJS) has less than 10% involvement of body surface area (BSA), whereas toxic epidermal necrolysis (TEN) has greater than 30% BSA involvement. SJS/TEN defines the gap of 10% to 30% involvement. Overall mortality for pediatric patients is less than 8%. Commonly caused by medications initiated in previous 8 weeks including sulfonamide antibiotics, lamotrigine, carbamazepine, phenobarbital, and several oncologic drugs. May also be caused by *Mycoplasma pneumoniae* infections. Nearly one third of cases have no identified trigger.
2. **Clinical presentation:** Fever and flu-like prodrome for 1 to 3 days prior to mucocutaneous lesions. Ophthalmologic and oropharyngeal symptoms are often first sites of mucosal involvement. Urogenital mucosal involvement seen in two-thirds of patients may lead to urinary retention and have significant long-term anatomic changes in female patients. Epidermal lesions are described as exquisitely tender (with pain out of

proportion), ill-defined, coalescing macules and patches of erythema with central purple-to-black areas. Lesions typically start on face and trunk then spread in a symmetric distribution sparing the scalp, palms, and soles. Bullae form with disease progression. Then, the epidermis sloughs with positive Nikolsky and Asboe-Hansen (lateral expansion of bullae with pressure) signs. Acute phase may last 8 to 12 days with reepithelialization requiring up to four weeks.

3. **Diagnosis:** Although usually not necessary, clinical diagnosis may be confirmed with a skin biopsy. Additional work up includes CBC, CMP, ESR, CRP, bacterial and fungal cultures, *M. pneumoniae* PCR, and CXR.
4. **Treatment:** Remove offending agent, supportive care, and close monitoring of all organ systems in the inpatient/ICU setting. There is controversy regarding IVIG and single dose of TNF-alpha inhibitor early in course. Systemic steroids probably should not be used.
5. **Complications:** At risk for serious complications including secondary bacterial infections (*Staphylococcus aureus* and *Pseudomonas aeruginosa*), septic shock, pneumonia, acute respiratory distress syndrome (ARDS), and epithelial necrosis of the GI tract. Most common complication in children is corneal scarring and dry eye.

E. Autoimmune Bullous Diseases: See **Section X, Online Content.**

**VIII. NAIL DISORDERS<sup>28</sup>: SEE SECTION X, ONLINE CONTENT**

**IX. DISORDERS OF PIGMENTATION: SEE SECTION X, ONLINE CONTENT**

## REFERENCES

A complete list of references can be found online at [www.expertconsult.com](http://www.expertconsult.com).

## X. ONLINE CONTENT

### A. Autoimmune and Allergic Lesions

#### 1. Autoimmune bullous diseases

- a. Very rare in children but should be considered if bullous lesions do not respond to standard therapy. Suspicion for any of the following should warrant referral to a dermatologist for diagnosis and management.

- b. **Pemphigus vulgaris** (Figure EC 8.A):

- (1) Pathogenesis: IgG autoantibodies to epidermal adhesion molecules, which interrupt integrity of epidermis and/or mucosa and result in extensive blister formation.
- (2) Clinical presentation: Flaccid bullae that start in the mouth and spread to face, scalp, trunk, extremities, and other mucosal membranes. Positive Nikolsky sign. Ruptured blisters are painful and prone to secondary infection. Can lead to impaired oral intake if there is significant oral mucosal involvement.
- (3) Treatment: Systemic glucocorticoids, rituximab, and/or intravenous immunoglobulin.

- c. **Pemphigus foliaceus:**

- (1) Pathogenesis: IgG autoantibodies bind to the same antigen as in bullous impetigo and staphylococcal scalded skin syndrome; thus lesions are superficial and rupture easily. Can be triggered by certain drugs, including thiol compounds and penicillins.
- (2) Clinical presentation: Scaling, crusting erosions on erythematous base that appear on face, scalp, trunk, and back. No mucosal involvement. Lesions are more superficial than in pemphigus vulgaris.
- (3) Treatment: Systemic glucocorticoids or rituximab. There is currently a move away from systemic steroids due to good efficacy and safety data on rituximab.

- d. **Bullous pemphigoid:**

- (1) Pathogenesis: Autoantibodies to the epithelial basement membrane that results in an inflammatory cascade and causes separation of epidermis from dermis and epithelium from subepithelium.



Figure EC 8.A

Pemphigus vulgaris. (From Cohen BA. *Dermatology Image Atlas*; 2001. <http://www.dermatlas.org/>.)

**Figure EC 8.B**

Acute paronychia. (From Cohen BA. Disorders of the hair and nails. In: *Pediatric Dermatology*. 4th ed. China: Saunders Elsevier; 2013:211–239.)

- (2) Clinical presentation: Prodrome of inflammatory lesions that progresses into large (1 to 3 cm), tense, extremely pruritic bullae on trunk, flexural regions, and intertriginous areas. Few patients have oral mucosal lesions. Negative Nikolsky sign.
  - (3) Treatment: Immunosuppression (topical glucocorticoids, systemic glucocorticoids, glucocorticoid-sparing agents like methotrexate, mycophenolate, or azathioprine).
- e. **Dermatitis herpetiformis:**
- (1) Pathogenesis: Strong genetic predisposition and link to gluten intolerance/celiac disease. IgA deposits found in dermal papillae.
  - (2) Clinical presentation: Symmetric, intensely pruritic papulovesicles clustered on extensor surfaces.
  - (3) Treatment: Dapsone, strict gluten-free diet.

## B. Nail Disorders<sup>28</sup>

1. Acquired nail disorders
  - a. Paronychia: Red, tender swelling of proximal or lateral nail folds ([Figures EC 8.B and EC 8.C](#))
    - (1) Acute form: Caused by bacterial invasion after trauma to cuticle
      - (a) Clinical features: Exquisite pain, sudden swelling, and abscess formation around one nail.
      - (b) Treatment: Responds quickly to drainage of abscess and warm tap-water soaks; occasionally anti-staphylococcal antibiotics required.



**Figure EC 8.C**

Chronic paronychia. (From Cohen BA. Disorders of the hair and nails. In: *Pediatric Dermatology*. 4th ed. China: Saunders Elsevier; 2013:211–239.)

- (2) Chronic form: May involve one or several nails, history of frequent exposure to water or thumb-sucking; causative organisms *Candida* species, usually *C. albicans*.
  - (a) Clinical features: Mild tenderness, minimal purulence, nail may be discolored or dystrophic.
  - (b) Treatment: Resolves with topical antifungal agents and water avoidance; heals without scarring when thumb-sucking ends.
- b. Nail dystrophy: Distortion and discoloration of normal nail-plate structure; often traumatic or inflammatory causes ([Figures EC 8.D-EC 8.I](#)).
  - (1) Onychomycosis: A result of dermatophyte fungal infection, unusual before puberty. Oral and topical antifungals (terbinafine, itraconazole, ciclopirox) are used off-label with high cure rates and few adverse effects.<sup>29</sup>
  - (2) Subungual hematoma: Brown-black nail discoloration following crush injury. Usually resolves without treatment; large, painful blood collections may be drained. Must differentiate from melanoma and melanonychia.
- c. Nail changes and systemic disease ([Figures EC 8.J and EC 8.K](#))
  - (1) Clubbing: Complication of chronic lung or heart disease.
  - (2) Beau lines: Transverse, white lines/grooves that move distally with nail growth; due to growth arrest from systemic illness, medications, or toxins.
  - (3) Onychomadesis: Accentuated Beau lines often with separation of the nail from base of nail. Usually self-limited and very common following Coxsackie A6 hand, foot, and mouth disease.



**Figure EC 8.D**

Onychomycosis. (From Cohen BA. Disorders of the hair and nails. In: *Pediatric Dermatology*. 4th ed. China: Saunders Elsevier; 2013:211–239.)



**Figure EC 8.E**

Traumatic subungual hemorrhage. (From Cohen BA. Disorders of the hair and nails. In: *Pediatric Dermatology*. 4th ed. China: Saunders Elsevier; 2013:211–239.)



**Figure EC 8.F**

Acral melanoma. (From Cohen BA. Disorders of the hair and nails. In: *Pediatric Dermatology*. 4th ed. China: Saunders Elsevier; 2013:211–239.)



**Figure EC 8.G**

Melonychia. (From Cohen BA. Disorders of the hair and nails. In: *Pediatric Dermatology*. 4th ed. China: Saunders Elsevier; 2013:211–239.)



**Figure EC 8.H**

Nail psoriasis. (From Cohen BA. Disorders of the Hair and Nails. In: *Pediatric Dermatology*. 4th ed. China: Saunders Elsevier; 2013:211–239.)



**Figure EC 8.I**

Atopic nails. (From Cohen BA. Disorders of the hair and nails. In: *Pediatric Dermatology*. 4th ed. China: Saunders Elsevier; 2013:211–239.)



**Figure EC 8.J**

Nail clubbing. (From Cohen BA. Disorders of the hair and nails. In: *Pediatric Dermatology*. 4th ed. China: Saunders Elsevier; 2013:211–239.)



**Figure EC 8.K**

Beau lines. (From Cohen BA. Disorders of the hair and nails. In: *Pediatric Dermatology*. 4th ed. China: Saunders Elsevier; 2013:211–239.)

**2. Congenital/hereditary nail disorders****a. Isolated nail disorders (Figures EC 8.L and EC 8.M)**

(1) Congenital nail dystrophy: Clubbing and spooning (koilonychia), may be autosomal dominant with no other anomalies.

(2) Congenital ingrown toenails: Most self-limiting.

**b. Genodermatoses and systemic disease (Figures EC 8.N and EC 8.O)**

(1) Periungual fibromas: Arise in proximal nail groove, common finding in tuberous sclerosis.

(2) Congenital nail hypoplasia: Can occur with intrauterine exposure to anticonvulsants, alcohol, and warfarin.

**Figure EC 8.L**

Koilonychia. (From Cohen BA. Disorders of the hair and nails. In: *Pediatric Dermatology*. 4th ed. China: Saunders Elsevier; 2013:211–239.)

**Figure EC 8.M**

Congenital ingrown nails. (From Cohen BA. Disorders of the hair and nails. In: *Pediatric Dermatology*. 4th ed. China: Saunders Elsevier; 2013:211–239.)

**Figure EC 8.N**

Periumgual fibromas. (From Cohen BA. Disorders of the hair and nails. In: *Pediatric Dermatology*. 4th ed. China: Saunders Elsevier; 2013:211–239.)

**Figure EC 8.0**

Fetal alcohol syndrome with congenital hypoplastic and dysplastic nails. (From Cohen BA. Disorders of the hair and nails. In: *Pediatric Dermatology*. 4th ed. China: Saunders Elsevier; 2013:211–239.)

**Figure EC 8.P**

Café-au-lait spot. (From Cohen BA. Disorders in pigmentation. In: *Pediatric Dermatology*. 4th ed. China: Saunders Elsevier; 2013:48–68.)

### C. Disorders of Pigmentation<sup>30</sup>

#### 1. Hyperpigmentation

- a. Congenital melanocytic nevi (CMN): Melanocytic nevi that are either present at birth or appear within the first few months of life in 1% to 3% of neonates.<sup>31</sup>
  - (1) Appearance: Black or tan in color with irregular borders and often dark terminal hairs.
  - (2) Risks:
    - (a) Melanoma—At least 5% of large CMN greater than 20 cm with 70% of this cohort having cancerous transformation by 10 years of age.<sup>32</sup> The presence of approximately 20 satellite nevi (smaller congenital nevi) also increases risk of melanoma.
    - (b) Neurocutaneous melanosis—Children with large, multiple, satellite nevi, or lesions over the spine are at risk for lepto-meningeal involvement with symptoms that may include hydrocephalus and seizures that may require evaluation by gadolinium contrast MRI.<sup>33,34</sup>
- b. Epidermal melanosis: Most lesions appear tan or light brown
  - (1) Café au lait spots (**Figure EC 8.P**): Discrete tan macules that appear at birth or during childhood in 10% to 20% of normal individuals, sizes vary from freckles to patches, may involve any

**Figure EC 8.Q**

Acanthosis nigricans, axilla. (From Cohen BA. Disorders in pigmentation. In: *Pediatric Dermatology*. 4th ed. China: Saunders Elsevier; 2013:48–68.)

∞

**Figure EC 8.R**

Acanthosis nigricans, neck. (From Cohen BA. Disorders in pigmentation. In: *Pediatric Dermatology*. 4th ed. China: Saunders Elsevier; 2013:48–68.)

site on skin. May be diagnostic marker for Neurofibromatosis type 1 ( $\geq 6$  lesions, each greater than 5 mm in diameter in prepubertal, or greater than 15 mm in postpubertal child) or other syndromes.

- (2) Freckles (ephelides): Reddish-tan and brown macules on sun-exposed surfaces, usually 2 to 3 mm in diameter. Serve as an independent risk factor for skin cancers in adulthood and can be an added sign of the importance of photoprotection which may decrease additional lesions.
- (3) Acanthosis nigricans (**Figures EC 8.Q and EC 8.R**): Brown-to-black hyperpigmentation with velvety or warty skin in intertriginous areas, typically found in the skin folds of the neck and

**Figure EC 8.S**

Postinflammatory hyperpigmentation. (From Cohen BA. Disorders in pigmentation. In: *Pediatric Dermatology*. 4th ed. China: Saunders Elsevier; 2013:48–68.)

axilla. Most commonly occur in obese individuals with insulin resistance at risk for type II diabetes. Finding may decrease after puberty with weight reduction.

- c. Dermal melanosis: Slate-gray, dark brown, or bluish-green lesions.
  - (1) Post-inflammatory hyperpigmentation ([Figure EC 8.S](#)): Most common cause of increased pigmentation.
    - (a) Pathogenesis: Follows inflammatory processes in the skin (e.g., diaper dermatitis, insect bites, drug reactions, traumatic injuries).
    - (b) Clinical features: Localized lesions, follow distribution of resolving disorder. More prominent in darkly pigmented children.
    - (c) Treatment: Lesions typically fade over several months. Photoprotection is critical with protective clothing and sunscreen of at least SPF 30. Individuals should also avoid physical trauma to areas as well as medications that may worsen hyperpigmentation. Intervention with medication is not always required; however, when it is, hydroquinone is first-line therapy.<sup>35</sup>
  - (2) Acquired nevomelanocytic nevi (aka pigmented nevi or moles) ([Figure EC 8.T](#))
    - (a) Pathogenesis: Develop in early childhood as flat lesions called junctional nevi, then develop into compound nevi when nevus cells migrate into the dermis and lesions enlarge and become papular.
    - (b) Clinical features: Increase in darkness, size, and number during puberty; generally do not exceed 5 mm and retain regularity in color, texture, and symmetry; on sun-exposed areas.
    - (c) Treatment: Excision unnecessary, unless cosmetic concern.

**Figure EC 8.T**

Compound nevomelanocytic nevus. (From Cohen BA. Disorders in pigmentation. In: *Pediatric Dermatology*. 4th ed. China: Saunders Elsevier; 2013:48–68.)

- (d) Changes associated with development of melanoma: See ABCDEs of Melanoma as well as burning, itching, or redness.
- (3) Melanomas ([Figure EC 8.U](#))
  - (a) Pathogenesis: May occur de novo or within acquired or congenital nevi.
  - (b) Epidemiology: High lifetime risk in those with presence of multiple, large, and irregularly pigmented, bordered, textured nevi and family history of malignant melanomas.
  - (c) Management: Children in high-risk families must be carefully observed for atypical nevi development especially in adolescence. Changing nevi with unusual appearance or an “ugly duckling” (mole that is different from all other moles) must be considered for biopsy.
  - (d) ABCDEs of Melanoma: Criteria for older children and adults is as follows: **A**symmetric shape, **B**orders that are irregular, **C**olor that is variable throughout lesion, **D**iameter greater than the size of a pencil eraser (>6 mm), **E**volution (change is the most important factor in melanoma diagnosis).<sup>36</sup> Pediatric patients up to age 20 have their own ABCD criteria: **A**melanotic, **B**leeding, **B**ump, **C**olor uniformity, **D**e novo, any **D**iameter.<sup>37</sup>
- (4) Melanonychia (see [Figure EC 8.G](#)): Darkened nail pigment that most commonly is caused by melanin or hemosiderin deposits in the nail plate. Regular, organized longitudinal lines tend to be benign whereas irregularities are associated with nail melanoma in adults. However, nail matrix nevi in children often have features that would be considered red flags in the adult population; thus,



**Figure EC 8.U**

Melanoma. (From Cohen BA. Disorders in pigmentation. In: *Pediatric Dermatology*. 4th ed. China: Saunders Elsevier; 2013:48–68.)



∞

**Figure EC 8.V**

Pigmented spitz nevus. (From Cohen BA. Disorders in pigmentation. In: *Pediatric Dermatology*. 4th ed. China: Saunders Elsevier; 2013:48–68.)

these criteria may not be applied to children. There are no pediatric specific guidelines for management. Typically, this clinical finding is due to nail matrix nevi. Nail melanomas are very rare though children with this clinical finding warrant close follow up.<sup>38,39</sup>

- (5) Spitz nevus (aka spindle and epithelial cell nevus) ([Figure EC 8.V](#)): Innocent nevomelanocytic nevus often confused with malignant melanoma.
- (a) Clinical features: Rapidly growing, dome-shaped, red or reddish-brown papules or nodules on face or lower extremities that reach full size quickly.
  - (b) Management: Observe if features of innocent acquired nevus are present. Consider referral to pediatric dermatology if unusual atypical features present.

## 2. Hypopigmentation and depigmentation

### a. Localized hypopigmentation

- (1) Hypopigmented macules ([Figure EC 8.W](#))
- (a) Epidemiology: 0.1% to 0.5% of normal newborns have a single hypopigmented macule but it may be a marker for tuberous sclerosis as 70% to 90% of those affected have such macules on the trunk at birth.
  - (b) Clinical features: Trunk involvement is most common. Majority are lancet or ash-leaf shaped, but may be round, oval, dermatomal, segmental, or irregularly shaped. Vary from pinpoint confetti spots to large patches (>10 cm).

**Figure EC 8.W**

Congenital hypopigmented macule. (From Cohen BA. Disorders in pigmentation. In: *Pediatric Dermatology*. 4th ed. China: Saunders Elsevier; 2013:48–68.)

- (c) Diagnosis: Wood lamp helpful in lightly pigmented children.
  - (d) Management: In those where systemic disease is suspected, close observation for other cutaneous findings and systemic symptoms is indicated.
- (2) Post-inflammatory hypopigmentation ([Figure EC 8.X](#))
- (a) Pathogenesis: May appear after an inflammatory skin condition.
  - (b) Clinical features: Seen in association with primary lesions of underlying disorder (such as atopic dermatitis). Patches usually variable in size and irregularly shaped. Concomitant hyperpigmentation is common.
- b. Diffuse hypopigmentation
- (1) Albinism: Heterogeneous group of inherited disorders manifested by generalized hypopigmentation or depigmentation of skin, eyes, and hair. These individuals should undergo ophthalmologic examination to evaluate for various associated conditions. Sun protection is important as well as regular skin exams.
- 3. Dyspigmentation**
- a. Blaschkoid dyspigmentation<sup>40</sup>: Congenital hypopigmentation and hyperpigmentation along the lines of Blaschko ([Figure EC 8.Y](#)).
    - (1) Patterns of hyper- or hypopigmentation: Whorl shape on trunk, V-shape on the back, waves on the vertex scalp.
    - (2) Pathogenesis: Blaschko lines occur due to genetic mosaicism.
    - (3) Children unlikely to have or develop serious extracutaneous involvement.



**Figure EC 8.X**

Postinflammatory hypopigmentation. (From Cohen BA. Disorders in pigmentation. In: *Pediatric Dermatology*. 4th ed. China: Saunders Elsevier; 2013:48–68.)



**Figure EC 8.Y**

Blaschkoid dyspigmentation. (From Cohen BA. Disorders in pigmentation. In: *Pediatric Dermatology*. 4th ed. China: Saunders Elsevier; 2013:48–68.)

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**FIGURE 8.1**

Infantile hemangioma. (From Cohen BA. *Atlas of Pediatric Dermatology*. 3rd ed. St Louis: Mosby; 2005:126.)



**FIGURE 8.2**

Pyogenic granuloma. (From Cohen BA. *Dermatology Image Atlas*; 2001. <http://www.dermatlas.org/>.)



**FIGURE 8.3**

Molluscum contagiosum. (From Cohen BA. *Atlas of Pediatric Dermatology*. 4th ed. China: Elsevier Limited; 2013:131.)



**FIGURE 8.5**

Roseola. (From Cohen BA. *Atlas of Pediatric Dermatology*. 4th ed. China: Elsevier Limited; 2013:177.)



**FIGURE 8.6**

Herpetic gingivostomatitis. (Modified from Cohen BA. *Atlas of Pediatric Dermatology*. 4th ed. China: Elsevier Limited; 2013:106.)



**FIGURE 8.7**

Herpes zoster. (From Cohen BA. *Atlas of Pediatric Dermatology*. 4th ed. China: Elsevier Limited; 2013:110.)



**FIGURE 8.8**

Varicella. (From Cohen BA. *Atlas of Pediatric Dermatology*. 4th ed. China: Elsevier Limited; 2013:108.)



**FIGURE 8.9**

Measles. (From Cohen BA. *Atlas of Pediatric Dermatology*. 4th ed. China: Elsevier Limited; 2013:175.)



**FIGURE 8.10**

Fifth disease. (From Cohen BA. *Atlas of Pediatric Dermatology*. 4th ed. China: Elsevier Limited; 2013:176.)



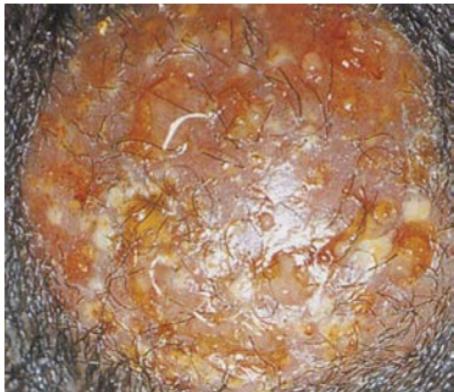
**FIGURE 8.11**

Scabies. (From Cohen BA. *Atlas of Pediatric Dermatology*. 3rd ed. St Louis: Mosby; 2005:126.)



**FIGURE 8.12**

Tinea capitis. (From Cohen BA. *Atlas of Pediatric Dermatology*. 3rd ed. St Louis: Mosby; 1993.)



**FIGURE 8.13**

Kerion. (From Cohen BA. *Atlas of Pediatric Dermatology*. 4th ed. China: Elsevier Limited; 2013:218c.)



**FIGURE 8.14**

Tinea corporis. (From Cohen BA. *Atlas of Pediatric Dermatology*. 4th ed. China: Elsevier Limited; 2013:96.)



**FIGURE 8.15**

Tinea pedis. (From Cohen BA. *Dermatology Image Atlas*; 2001. <http://www.dermatlas.org/>.)



**FIGURE 8.16**

Tinea versicolor. (From Cohen BA. *Atlas of Pediatric Dermatology*. 4th ed. China: Elsevier Limited; 2013:99.)



**FIGURE 8.17**

Scarlet fever. (From Cohen BA. *Dermatology Image Atlas*; 2001. <http://www.dermatlas.org/>.)



**FIGURE 8.18**

Telogen effluvium. (From Cohen BA. *Dermatology Image Atlas*; 2001. <http://www.dermatlas.org/>.)



**FIGURE 8.19**

Alopecia areata. (From Cohen BA. *Atlas of Pediatric Dermatology*. 4th ed. China: Elsevier Limited; 2013:219.)



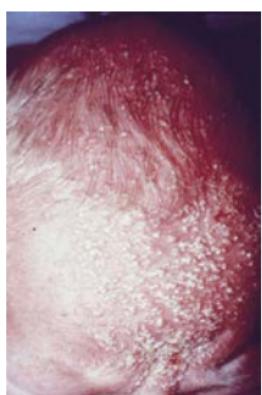
**FIGURE 8.20**

Traction alopecia. (From Cohen BA. *Atlas of Pediatric Dermatology*. 4th ed. China: Elsevier Limited; 2013:220.)



**FIGURE 8.22**

Erythema toxicum neonatorum. (From Cohen BA. *Pediatric Dermatology*. 2nd ed. St Louis: Mosby; 1999:18.)



**FIGURE 8.23**

Transient neonatal pustular melanosis. (From Cohen BA. *Atlas of Pediatric Dermatology*. 4th ed. China: Elsevier Limited; 2013:20.)



**FIGURE 8.24**

Hyperpigmentation from resolving transient neonatal pustular melanosis. (From Cohen BA. *Atlas of Pediatric Dermatology*. 4th ed. China: Elsevier Limited; 2013:20.)



**FIGURE 8.25**

Miliaria rubra. (From Cohen BA. *Atlas of Pediatric Dermatology*. 4th ed. China: Elsevier Limited; 2013:21.)



**FIGURE 8.26**

Milia. (From Cohen BA. *Atlas of Pediatric Dermatology*. 4th ed. China: Elsevier Limited; 2013:22.)



**FIGURE 8.27**

Neonatal acne. (From Cohen BA. *Atlas of Pediatric Dermatology*. 4th ed. China: Elsevier Limited; 2013:22.)



**FIGURE 8.28**

Seborrheic dermatitis. (From Cohen BA. *Atlas of Pediatric Dermatology*. 4th ed. China: Elsevier Limited; 2013:32.)



**FIGURE 8.29**

Seborrheic dermatitis. (From Cohen BA. *Atlas of Pediatric Dermatology*. 4th ed. China: Elsevier Limited; 2013:32.)



**FIGURE 8.30**

Diaper candidiasis. (From Cohen BA. *Atlas of Pediatric Dermatology*. 4th ed. China: Elsevier Limited; 2013:33.)



**FIGURE 8.31**

Allergic contact dermatitis. (From Cohen BA. *Atlas of Pediatric Dermatology*. 4th ed. China: Elsevier Limited; 2013:77.)



**FIGURE 8.32**

Poison ivy. (From Cohen BA. *Dermatology Image Atlas*; 2001. <http://www.dermatlas.org/>.)



**FIGURE 8.33**

Infantile eczema. (From Cohen BA. *Atlas of Pediatric Dermatology*. 3rd ed. St Louis: Mosby; 2005:79.)



**FIGURE 8.34**

Childhood eczema. (From Cohen BA. *Dermatology Image Atlas*; 2001. <http://www.dermatlas.org/>.)



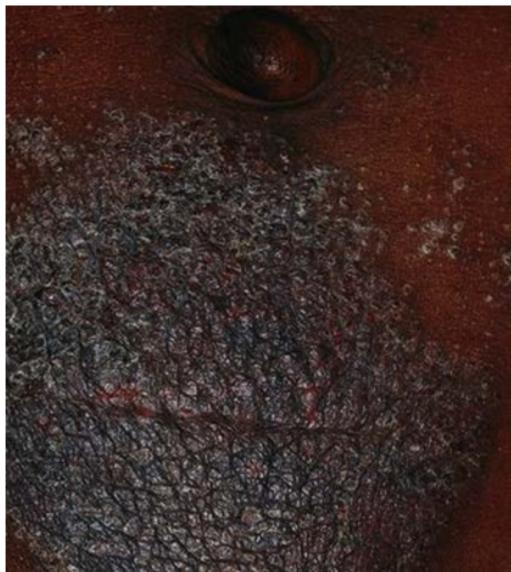
**FIGURE 8.35**

Nummular eczema. (From Cohen BA. *Atlas of Pediatric Dermatology*. 3rd ed. St Louis: Mosby; 2005:80.)



**FIGURE 8.36**

Follicular eczema. (From Cohen BA. *Atlas of Pediatric Dermatology*. 4th ed. China: Elsevier Limited; 2013:83.)



**FIGURE 8.37**

Childhood eczema with lesion in suprapubic area. (From Cohen BA. *Atlas of Pediatric Dermatology*. 3rd ed. St Louis: Mosby; 2005.)



**FIGURE 8.38**

Papular urticaria. (From Cohen BA. *Dermatology Image Atlas*; 2001. <http://www.dermatlas.org/>.)

# Chapter 9

## Development, Behavior, and Developmental Disability

Brittany Badesch, MD

 See additional content on Expert Consult

### I. DEVELOPMENTAL DEFINITIONS<sup>1,2</sup>

#### A. Developmental Streams

1. **Gross Motor Skills:** Descriptions of posture and locomotion—in general, how a child moves from one location to another.
2. **Fine-Motor and Visual-Motor Problem-Solving Skills:** Upper extremity and hand manipulative abilities and hand-eye coordination. These require an intact motor substrate and a given level of nonverbal cognitive ability.
3. **Language:** The ability to understand and communicate with another person. This is the best predictor of intellectual performance in the absence of a communication disorder or significant hearing impairment.
4. **Personal-Social Skills:** Communicative in origin; represent the cumulative impact of language comprehension and problem-solving skills.
5. **Adaptive Skills:** Skills concerned with self-help or activities of daily living.

#### B. Developmental Quotient (DQ)

1. A calculation that reflects the rate of development in any given stream; represents the percentage of normal development present at the time of testing.

$$DQ = \frac{\text{Developmental age}}{\text{Chronological age}} \times 100$$

2. Two separate developmental assessments over time are more predictive of later abilities than a single assessment.
3. In contrast to developmental quotient (DQ), intelligence quotient (IQ) has statistical reliability and validity.<sup>2</sup>

#### C. Abnormal Development

1. **Delay:** Performance significantly below average (DQ <70) in a given area of development. May occur in a single stream or several streams (“global developmental delay”).
2. **Deviancy:** Atypical development within a single stream, such as developmental milestones occurring out of sequence. Deviancy does not necessarily imply abnormality, but should alert one to the possibility that problems may exist.

*Example:* An infant who rolls at an early age may have abnormally increased tone.

3. **Dissociation:** A substantial difference in the rate of development between two or more streams.

*Example:* Increased motor delay relative to cognition seen in some children with cerebral palsy (CP).

## II. GUIDELINES FOR NORMAL DEVELOPMENT AND BEHAVIOR

### A. Developmental Milestones (**Table 9.1**)

Developmental assessment is based on the premise that milestone acquisition occurs at a specific rate in an orderly and sequential manner.

### B. Age-Appropriate Behavioral Issues in Infancy and Early Childhood:

See **Table 9.2.**

## III. DEVELOPMENTAL SCREENING AND EVALUATION OF DEVELOPMENTAL DISORDERS

### A. Developmental Surveillance and Screening Guidelines

1. **Developmental surveillance should be included in every well-child visit, and any concerns should be addressed immediately with formal screening.** This includes direct observation of the child and eliciting and attending to the parent's concerns.
2. **Standardized developmental screening should be administered at 9-month, 18-month, and 30-month well-child visits,** in the absence of developmental concerns. If a 30-month visit is not possible, this screening can be done at the 24-month visit.
3. See full American Academy of Pediatrics (AAP) guideline for developmental screening algorithm.<sup>3</sup>

### B. Commonly Used Developmental Screening and Assessment Tools:

See **Table 9.3**

### C. Identification of Developmental “Red Flags”: See **Table 9.4**

### D. Evaluation of Abnormal Development

1. Referral to developmental and appropriate subspecialists.
2. Referral to early intervention services for children aged 0 to 3 years (see **Section V**).
3. Medical evaluation as outlined in **Tables 9.5–9.7**.
4. Genetic evaluation (**Table 9.8**) is warranted for all children with developmental delay or intellectual disability (ID) if the cause is not known (e.g., previous traumatic brain injury or neurologic insult).

## IV. SPECIFIC DISORDERS OF DEVELOPMENT

### A. Overview

1. Mental and/or physical impairment(s) that cause significant limitations in functioning.
2. **Developmental diagnosis** is a functional description; identification of an etiology is important to further inform treatment, prognosis, comorbidities, and future risk.

**TABLE 9.1****DEVELOPMENTAL MILESTONES**

Age	Gross Motor	Visual–Motor/Problem-Solving	Language	Social/Adaptive
1 month	Raises head from prone position	Visually fixes, follows to midline, has tight grasp	Alerts to sound	Regards face
2 months	Holds head in midline, lifts chest off table	No longer clenches fists tightly, follows object past midline	Smiles socially (after being stroked or talked to)	Recognizes parent
3 months	Supports on forearms in prone position, holds head up steadily	Holds hands open at rest, follows in circular fashion, responds to visual threat	Coos (produces long vowel sounds in musical fashion)	Reaches for familiar people or objects, anticipates feeding
4 months	Rolls over, supports on wrists, shifts weight	Reaches with arms in unison, brings hands to midline	Laughs, orients to voice	Enjoys looking around
6 months	Sits unsupported, puts feet in mouth in supine position	Unilateral reach, uses raking grasp, transfers objects	Babbles, ah-goo, razz, lateral orientation to bell	Recognizes that someone is a stranger
9 months	Pivots when sitting, crawls well, pulls to stand, cruises	Uses immature pincer grasp, probes with forefinger, holds bottle, throws objects	Says “mama, dada” indiscriminately, gestures, waves bye-bye, understands “no”	Starts exploring environment, plays gesture games (e.g., pat-a-cake)
12 months	Walks alone	Uses mature pincer grasp, can make a crayon mark, releases voluntarily	Uses two words other than “mama, dada” or proper nouns, jargoning (runs several unintelligible words together with tone or inflection), one-step command with gesture	Imitates actions, comes when called, cooperates with dressing
15 months	Creeps up stairs, walks backward independently	Scribbles in imitation, builds tower of two blocks in imitation	Uses 4–6 words, follows one-step command without gesture	15–18 months: uses spoon and cup
18 months	Runs, throws objects from standing without falling	Scribbles spontaneously, builds tower of three blocks, turns two or three pages at a time	Mature jargoning (includes intelligible words), 7–10-word vocabulary, knows five body parts	Copies parent in tasks (sweeping, dusting), plays in company of other children

*Continued*

**TABLE 9.1—CONT'D****DEVELOPMENTAL MILESTONES**

<b>Age</b>	<b>Gross Motor</b>	<b>Visual-Motor/Problem-Solving</b>	<b>Language</b>	<b>Social/Adaptive</b>
24 months	Walks up and down steps without help	Imitates stroke with pencil, builds tower of seven blocks, turns pages one at a time, removes shoes, pants, etc.	Uses pronouns (I, you, me) inappropriately, follows two-step commands, 50-word vocabulary, uses two-word sentences	Parallel play
3 years	Can alternate feet going up steps, pedals tricycle	Copies a circle, undresses completely, dresses partially, dries hands if reminded, unbuttons	Uses minimum of 250 words, three-word sentences, uses plurals, knows all pronouns, repeats two digits	Group play, shares toys, takes turns, plays well with others, knows full name, age, gender
4 years	Hops, skips, alternates feet going down steps	Copies a square, buttons clothing, dresses self completely, catches ball	Knows colors, says song or poem from memory, asks questions	Tells "tall tales," plays cooperatively with a group of children
5 years	Skips alternating feet, jumps over low obstacles	Copies triangle, ties shoes, spreads with knife	Prints first name, asks what a word means	Plays competitive games, abides by rules, likes to help in household tasks

From Capute AJ, Biehl RF. Functional developmental evaluation: prerequisite to habilitation. *Pediatr Clin North Am.* 1973;20:3; Capute AJ, Accardo PJ. Linguistic and auditory milestones during the first two years of life: a language inventory for the practitioner. *Clin Pediatr.* 1978;17:847; and Capute AJ, Shapiro BK, Wachtel RC, et al. The Clinical Linguistic and Auditory Milestone Scale (CLAMS): identification of cognitive defects in motor delayed children. *Am J Dis Child.* 1986;140:694. Rounded norms from Capute AJ, Palmer FB, Shapiro BK, et al. Clinical Linguistic and Auditory Milestone Scale: prediction of cognition in infancy. *Dev Med Child Neurol.* 1986;28:762.

**TABLE 9.2****AGE-APPROPRIATE BEHAVIORAL ISSUES IN INFANCY AND EARLY CHILDHOOD**

Age	Behavioral Issue	Symptoms	Guidance
1–3 months	Colic	Paroxysms of fussiness/crying, ≥3 per day, ≥3 days/week, may pull knees up to chest, pass flatus	Crying usually peaks at 6 weeks and resolves by 3–4 months. Prevent overstimulation; swaddle infant; use white noise, swing, or car rides to soothe. Avoid medication and formula changes. Encourage breaks for the primary caregiver.
3–4 months	Trained night feeding	Night awakening	Comfort quietly, avoid reinforcing behavior (i.e., avoid night feeds). Do not play at night. Introducing cereal or solid food does not reduce awakening. Develop a consistent bedtime routine. Place baby in bed while drowsy and not fully asleep.
9 months	Stranger anxiety/ separation anxiety	Distress when separated from parent or approached by a stranger	Use a transitional object (e.g., special toy, blanket). Use routine or ritual to separate from parent. May continue until 24 months but can reduce in intensity.
	Developmental night waking	Separation anxiety at night	Keep lights off. Avoid picking child up or feeding. May reassure verbally at regular intervals or place a transitional object in crib.
12 months	Aggression	Biting, hitting, kicking in frustration	Say “no” with negative facial cues. Begin time out (1 minute/year of age). No eye contact or interaction, place in a nonstimulating location. May restrain child gently until cooperation is achieved.
	Need for limit setting	Exploration of environment, danger of injury	Avoid punishing exploration or poor judgment. Emphasize child-proofing and distraction.
18 months	Temper tantrums	Occur with frustration, attention-seeking rage, negativity/refusal	Try to determine cause, react appropriately (i.e., help child who is frustrated, ignore attention-seeking behavior). Make sure child is in a safe location.
24 months	Toilet training	Child needs to demonstrate readiness: shows interest, neurologic maturity (i.e., recognizes urge to urinate or defecate), ability to walk to bathroom and undress self, desire to please/imitate parents, increasing periods of daytime dryness	Age range for toilet training is usually 2–4 years. Give guidance early; may introduce potty seat but avoid pressure or punishment for accidents. Wait until the child is ready. Expect some periods of regression, especially with stressors.

**TABLE 9.2—CONT'D****AGE-APPROPRIATE BEHAVIORAL ISSUES IN INFANCY AND EARLY CHILDHOOD**

<b>Age</b>	<b>Behavioral Issue</b>	<b>Symptoms</b>	<b>Guidance</b>
24–36 months	New sibling	Regression, aggressive behavior	Allow for special time with parent, 10–20 min daily of one-on-one time exclusively devoted to the older sibling(s). Child chooses activity with parent. No interruptions. May not be taken away as punishment.
36 months	Nightmares	Awakens crying, may or may not complain of bad dream	Reassure child, explain that he or she had a bad dream. Leave bedroom door open, use a nightlight, demonstrate there are no monsters under the bed. Discuss dream the following day. Avoid scary movies or television shows.
	Night terrors	Agitation, screaming 1–2 hours after going to bed. Child may have eyes open but not respond to parent. May occur at same time each night	May be familial, not volitional. <i>Prevention:</i> For several nights, awaken child 15 min before terrors typically occur. Avoid overtiredness. <i>Acute:</i> Be calm; speak in soft, soothing, repetitive tones; help child return to sleep. Protect child against injury.

From Dixon SD, Stein MT. *Encounters With Children: Pediatric Behavior and Development*. St Louis: Mosby; 2000.

**TABLE 9.3****DEVELOPMENTAL SCREENING TESTS BY DIAGNOSIS**

Diagnosis Evaluated	Screening Test	Age	Completed by	Comments	Weblink
Cognitive and motor development	Ages and Stages Questionnaire (ASQ)	4–60 months	Parent	Increased time efficiency (can fill out while waiting) Documents milestones that are difficult to assess in the office	<a href="http://www.agesandstages.com">www.agesandstages.com</a>
Developmental and behavioral problems	Parents' Evaluation of Developmental Status (PEDS)	0–8 years	Parent	May also be useful as a surveillance tool	<a href="http://www.pedstest.com">www.pedstest.com</a>
Language, problem-solving development	Capute Scales: Clinical Linguistic and Auditory Milestone Scale (CLAMS), Clinical Adaptive Test (CAT)	3–36 months	Clinician	Give quantitative DQs for language (CLAMS) and visual-motor/problem-solving (CAT) abilities	<a href="http://www.brookespublishing.com/resource-center/screening-and-assessment/the-capute-scales/">http://www.brookespublishing.com/resource-center/screening-and-assessment/the-capute-scales/</a>
Autism spectrum disorders	Modified Checklist for Autism in Toddlers, Revised with Follow-Up (M-CHAT-R/F)	16–30 months	Parent	Positive screens require clinician follow-up	<a href="http://www.m-chat.org">www.m-chat.org</a>
	Communication and Symbolic Behavior Scales and Developmental Profile (CSBS DP; Infant Toddler Checklist)	6–24 months	Parent	The Infant Toddler Checklist is a one-page questionnaire that is part of a larger standardized screening tool (CSBS DP) Can be used in patients as young as 6 months	<a href="http://www.brookespublishing.com/checklist.pdf">www.brookespublishing.com/checklist.pdf</a>
	Childhood Autism Screening Test (CAST)	4–11 years	Parent	Only screening tool evaluated in preschool population	<a href="http://www.autismresearchcenter.com/project_9_cast">http://www.autismresearchcenter.com/project_9_cast</a>

Modified from American Academy of Pediatrics. Identifying infants and young children with developmental disorders in the medical home: an algorithm for developmental surveillance and screening. *Pediatrics*. 2006;118:405–420; American Academy of Pediatrics. Identification and evaluation of children with autism spectrum disorders. *Pediatrics*. 2007;120: 1183–1215; Robins DL, Casagrande K, Barton M, et al. Validation of the Modified Checklist for Autism in Toddlers, Revised with Follow-up (M-CHAT-R/F). *Pediatrics*. 2014;133:37–45.

**TABLE 9.4****DEVELOPMENTAL RED FLAGS**

Age of Patient	Red Flag Symptom
Any age	Loss of previously obtained developmental skills Parental or professional concerns about vision, fixing, or following an object or a confirmed visual impairment Hearing loss Persistently low muscle tone or floppiness Asymmetry of movements or other features suggestive of cerebral palsy, such as increased muscle tone Head circumference above the 99.6th percentile, below 0.4th percentile, or has crossed two major percentile lines (up or down)
5 months (corrected for gestation)	Not able to hold object placed in hand
6 months (corrected for gestation)	Not reaching for objects
12 months	Unable to sit unsupported
18 months	Not walking in male patients Not pointing at objects to share interest with others
24 months	Not walking in female patients
30 months and older	Unable to run Persistent toe walking

From Bellman M, Byrne O, Sege R. Developmental assessment of children. *BMJ*. 2013;346:31–36.

**TABLE 9.5****DEVELOPMENTAL EVALUATION: PERTINENT HISTORY AND PHYSICAL**

Prenatal and Birth History	Prenatal genetic screening Perception of fetal movement Pregnancy complications Toxins/teratogens Gestational age Birthweight Days in hospital/NICU admission Newborn screen results
Past Medical Problems	Trauma Infection Medication
Developmental History	Timing of milestone achievement Delayed skills Loss of skills (regression)
Behavioral History	Social skills Eye contact Affection Hyperactivity, impulsivity, inattention, distractibility Self-regulation Perseveration Worries/avoidance Stereotypies, peculiar habits

**TABLE 9.5—CONT'D****DEVELOPMENTAL EVALUATION: PERTINENT HISTORY AND PHYSICAL**

Educational History	Need for special services Grade retention Established educational plans
Family History	History of developmental disabilities, ADHD, seizures, tics, stillbirths, neonatal death, congenital malformations, mental illness, or recurrent miscarriages Family members who were late talkers or walkers Family member school performance Family pedigree (see Chapter 13)
General Exam	Height, weight, and head circumference Dysmorphic features Cardiac murmurs Midline defects Hepatosplenomegaly Skin exam
Age-directed Neuro Exam	Cranial nerves Tone and strength Postural reactions (Table EC 9.A) Functional abilities Reflexes [including primitive reflexes for infants (Table 9.6)]
In-Clinic Activities/Tests	Goodenough–Harris Draw-a-Person Test Gesell figures (Figure EC 9.A): Ask the child to copy various shapes Gesell block skills (Figure EC 9.B): Ask the child to reproduce block structures as built by the examiner

**TABLE 9.6****PRIMITIVE REFLEXES**

Primitive Reflexes	Elicitation	Response	Timing
Moro reflex (“embrace” response) of fingers, wrists, and elbows	<i>Supine</i> : Sudden neck extension; allow head to fall back about 3 cm	Extension, adduction, and then abduction of UEs, with semiflexion	Present at birth; disappears by 3–6 months
Galant reflex (GR)	<i>Prone suspension</i> : Stroking paravertebral area from thoracic to sacral region	Produces trunca l incurvature with concavity toward stimulated side	Present at birth; disappears by 2–6 months
Asymmetrical tonic neck reflex (ATNR, “fencer” response)	<i>Supine</i> : Rotate head laterally about 45–90 degrees	Relative extension of limbs on chin side and flexion on occiput side	Present at birth; disappears by 4–9 months
Symmetrical tonic neck reflex (STNR, “cat” reflex)	<i>Sitting</i> : Head extension/ flexion	Extension of UEs and flexion of LEs/ flexion of UEs and LE extension	Appears at 5 months; not present in most normal children; disappears by 8–9 months
Tonic labyrinthine supine (TLS)	<i>Supine</i> : Extension of the neck (alters relation of the labyrinths)	Tonic extension of trunk and LEs, shoulder retraction and adduction, usually with elbow flexion	Present at birth; disappears by 6–9 months

*Continued*

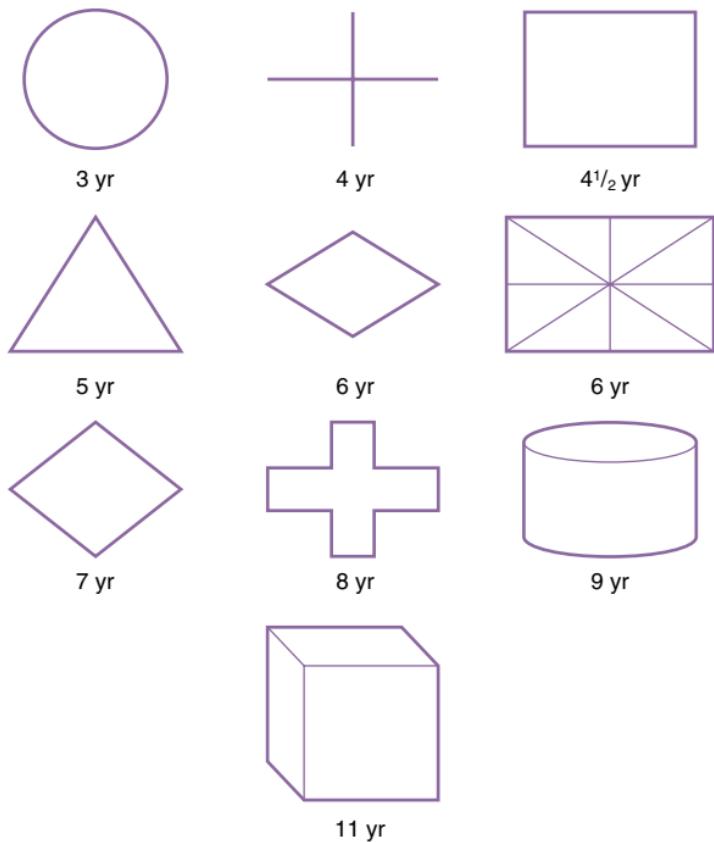
**TABLE EC 9.A****POSTURAL REACTIONS**

Postural Reaction	Age of Appearance	Description	Importance
Head righting	6 weeks–3 months	Lifts chin from table top in prone position	Necessary for adequate head control and sitting
Landau response	2–3 months	Extension of head, then trunk and legs when held prone	Early measure of developing trunk control
Derotational righting	4–5 months	Following passive or active head turning, body rotates to follow direction of head	Prerequisite to independent rolling
Anterior propping	4–5 months	Arm extension anteriorly in supported sitting	Necessary for tripod sitting
Parachute	5–6 months	Arm extension when falling	Facial protection when falling
Lateral propping	6–7 months	Arm extension laterally in protective response	Allows independent sitting
Posterior propping	8–10 months	Arm extension posteriorly	Allows pivoting in sitting

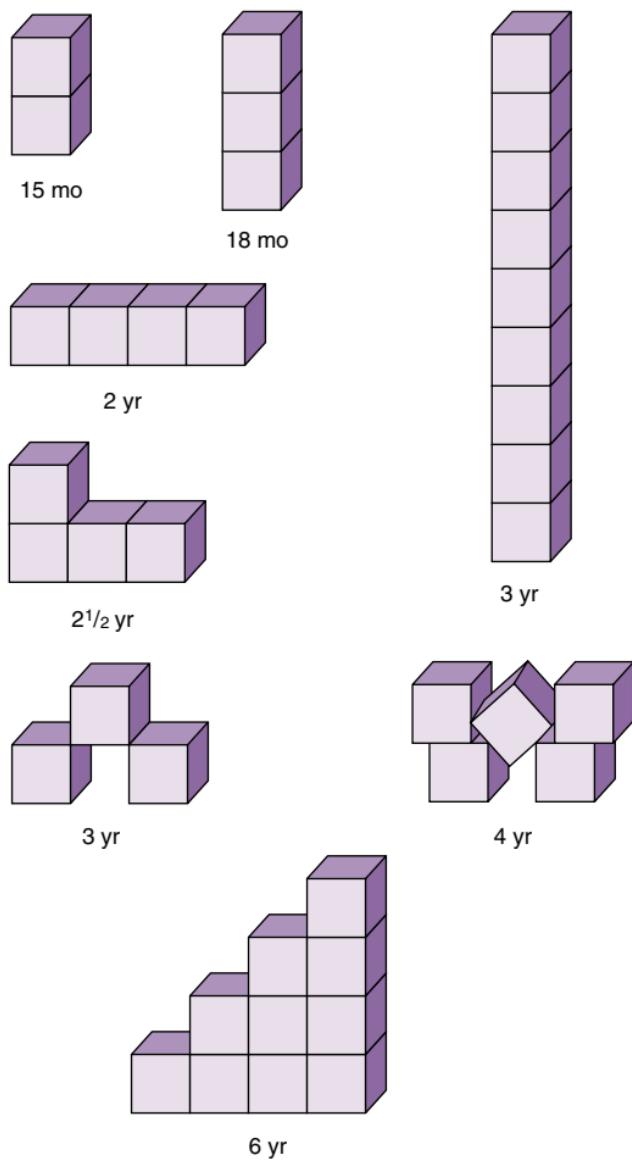
Modified from Milani-Comparetti A, Gidoni EA. Routine developmental examination in normal and retarded children. *Dev Med Child Neurol*. 1967;9:631–638; Capute AJ. Early neuromotor reflexes in infancy. *Pediatr Ann*. 1986;15:217–218, 221–223, 226; Capute AJ, Palmer FB, Shapiro BK, et al. Primitive reflex profile: a quantitation of primitive reflexes in infancy. *Dev Med Child Neurol*. 1984;26:375–383; and Palmer FB, Capute AJ. Developmental disabilities. In: Oski FA, ed. *Principles and Practice of Pediatrics*. Philadelphia: JB Lippincott; 1994.



15 months	Imitates scribble
18 months	Scribbles spontaneously
2 years	Imitates stroke
2½ years	Differentiates horizontal and vertical stroke

**FIGURE EC 9.A**

Gesell Figures. (From Illingsworth RS. *The Development of the Infant and Young Child, Normal and Abnormal*. 5th ed. Baltimore: Williams & Wilkins; 1972:229–232; and Cattell P. *The Measurement of Intelligence of Infants and Young Children*. New York: Psychological Corporation; 1960:97–261.)

**FIGURE EC 9.B**

Gesell Block Skills. (From Capute AJ, Accardo PJ. *The Pediatrician and the Developmentally Disabled Child: A Clinical Textbook on Mental Retardation*. Baltimore: University Park Press; 1979:122.)

**TABLE 9.6—CONT'D****PRIMITIVE REFLEXES**

Primitive Reflexes	Elicitation	Response	Timing
Tonic labyrinthine prone (TLP)	<i>Prone:</i> Flexion of the neck	Active flexion of trunk with protraction of shoulders	Present at birth; disappears by 6–9 months
Positive support reflex (PSR)	<i>Vertical suspension:</i> Bouncing hallucal areas on firm surface	<i>Neonatal:</i> momentary LE extension followed by flexion <i>Mature:</i> extension of LEs and support of body weight	Present at birth; disappears by 2–4 months Appears by 6 months
Stepping reflex (SR, walking reflex)	<i>Vertical suspension:</i> Hallucal stimulation	Stepping gait	Disappears by 2–3 months
Crossed extension reflex (CER)	<i>Prone:</i> Hallucal stimulation of LE in full extension	Initial flexion, adduction, then extension of contralateral limb	Present at birth; disappears by 9 months
Plantar grasp	Stimulation of hallucal areas	Plantar flexion grasp	Present at birth; disappears by 9 months
Palmar grasp	Stimulation of palm	Palmar grasp	Present at birth; disappears by 9 months
Lower extremity placing (LEP)	<i>Vertical suspension:</i> Rubbing tibia or dorsum of foot against edge of table top	Initial flexion, then extension, then placing of LE on table top	Appears at 1 day
Upper extremity placing (UEP)	Rubbing lateral surface of forearm along edge of table top from elbow to wrist to dorsal hand	Flexion, extension, then placing of hand on table top	Appears at 3 months
Downward thrust (DT)	<i>Vertical suspension:</i> Thrust LEs downward	Full extension of LEs	Appears at 3 months

LE, Lower extremity; UE, upper extremity.

**TABLE 9.7****DEVELOPMENTAL EVALUATION: INITIAL LABS AND OTHER STUDIES**

Hearing screening	Formal audiologic testing is indicated for all children with global developmental delay or any delay in communication or language
Neuroimaging	Consider if abnormal neurologic exam, concern about head circumference growth velocity, or global developmental delay present
Electroencephalogram	Consider if history of or concern for seizure disorder
Laboratory studies	Consider CBC, CMP, lead level, CK, TSH based on history and exam Confirm newborn screen results

CBC, Complete blood count; CK, creatine kinase; CMP, complete metabolic panel; TSH, thyroid stimulating hormone.

**TABLE 9.8****DEVELOPMENTAL EVALUATION: GENETIC WORK-UP**

Chromosomal microarray (CMA)	Considered first-tier diagnostic test in <b>all</b> children with GDD/ID. <sup>24</sup>
Fragile X testing	Should be performed in all boys <b>and</b> girls with GDD/ID of unknown cause. Of boys with GDD/ID of unknown cause, 2%–3% will have fragile X syndrome, as will 1%–2% of girls.

**TABLE 9.8—CONT'D****DEVELOPMENTAL EVALUATION: GENETIC WORK-UP**

Testing for X-linked conditions Consider genetic testing for X-linked genes in boys with GDD/ID after negative CMA and negative fragile X testing. Should be specifically in those patients whose pedigree is suggestive of an X-linked condition.

The tests discussed above do not require referral to a genetic specialist and can be ordered by the patient's pediatrician as a part of the evaluation of global developmental delay/intellectual disability (GDD/ID). If unrevealing and severe DD/ID present, refer to genetic specialist for consideration of additional testing such as metabolic testing or whole exome sequencing.

From Moeschler JB, Shevell M. Comprehensive evaluation of the child with intellectual disability or global developmental delays. *Pediatrics*. 2014;134(3):e903–e918.

3. School- and home-based programs are helpful interventions for all developmental disorders (see [Section V](#)).

**B. Intellectual Disability****1. Definition and Epidemiology**

- a. Deficits in general mental abilities
- b. Affects approximately 1% of the population<sup>4</sup>

**2. Clinical Presentation**

- a. Delay in milestones (motor, language, social)
- b. Academic difficulty
- c. Identifiable features of known associated genetic syndrome (e.g., Trisomy 21, fragile X, Rett syndrome)

**3. Diagnosis**

- a. Diagnostic criteria: (1) deficits in intellectual functioning, (2) deficits in adaptive functioning, (3) onset of these deficits during the developmental period
- b. Deficits in adaptive functioning must be in one or more domains of activities of daily living.
- c. ID is further categorized as mild, moderate, severe, or profound in the DSM-5 based on the degree of functional deficit ([Table EC 9.B](#)).

**4. Interventions/Treatment**

Support, employment, and recreational programs through resources such as The Arc ([www.thearc.org](http://www.thearc.org)).

**C. Communication Disorders****1. Definition**

- a. Deficits in communication, language, or speech
- b. Can be subdivided into<sup>5</sup>:
  - (1) Receptive/expressive language disorder
  - (2) Speech sound disorders
  - (3) Childhood-onset fluency disorder (stuttering)
  - (4) Social pragmatic communication disorder



*Continued***TABLE EC 9.B****SEVERITY LEVELS FOR INTELLECTUAL DISABILITY**

Severity Level	Conceptual Domain	Social Domain
Mild	<p>For preschool children, there may be no obvious conceptual differences. For school-aged children and adults, there are difficulties in learning academic skills involving reading, writing, arithmetic, time, or money, with support needed in one or more areas to meet age-related expectations. In adults, abstract thinking, executive function (i.e., planning, strategizing, priority setting, and cognitive flexibility), and short-term memory, as well as functional use of academic skills (e.g., reading, money management) are impaired. There is a somewhat concrete approach to problems and solutions compared with age mates.</p>	<p>Compared with typically developing age mates, the individual is immature in social interactions (e.g., difficulty in accurately perceiving peers' social cues). Communication, conversation, and language are more concrete or immature than expected for age. There may be difficulties regulating emotion and behavior in age-appropriate fashion; these difficulties are noticed by peers in social situations. There is limited understanding of risk in social situations; social judgment is immature for age, and the person is at risk of being manipulated by others (gullibility).</p>
Moderate	<p>All through development, the individual's conceptual skills lag markedly behind those of peers. For preschoolers, language and preacademic skills may develop slowly. For school-aged children, progress in reading, writing, mathematics, and understanding of time and money occurs slowly across the school years, and is markedly limited compared with that of peers. For adults, academic skill development is typically at an elementary level, and support is required for all use of academic skills in work and personal life. Ongoing assistance on a daily basis is needed to complete conceptual tasks of day-to-day life, and others may take over these responsibilities fully for the individual.</p>	<p>The individual shows marked differences from peers in social and communicative behavior across development. Spoken language is typically a primary tool for social communication, but is much less complex than that of peers. Capacity for relationships is evident in ties to family and friends, and the individual may have successful friendships across life and sometimes romantic relations in adulthood. However, individuals may not perceive or interpret social cues accurately. Social judgment and decision-making abilities are limited, and caretakers must assist the person with life decisions. Friendships with typically developing peers are often affected by communication or social limitations. Significant social and communicative support is needed in work settings for success.</p>

**Table EC 9.B—CONT'D****SEVERITY LEVELS FOR INTELLECTUAL DISABILITY**

<b>Severity Level</b>	<b>Conceptual Domain</b>	<b>Social Domain</b>
Severe	Attainment of conceptual skills is limited. The individual generally has little understanding of written language or of concepts involving numbers, quantity, time, and money. Caretakers provide extensive support for problem solving throughout life.	Spoken language is quite limited in terms of vocabulary and grammar. Speech may be single words or phrases, and may be supplemented through augmentative means. Speech and communication are focused on the here and now within everyday events. Language is used for social communication more than for explication. Individuals understand simple speech and gestural communication. Relationships with family members and familiar others are a source of pleasure and help.
Profound	Conceptual skills generally involve the physical world rather than symbolic processes. The individual may use objects in goal-directed fashion for self-care, work, and recreation. Certain visuospatial skills (e.g., matching and sorting based on physical characteristics) may be acquired. However, co-occurring motor and sensory impairments may prevent functional use of objects.	The individual has very limited understanding of symbolic communication in speech or gesture. He or she may understand some simple instructions or gestures. The individual expresses his or her own desires and emotions largely through nonverbal, nonsymbolic communication. The individual enjoys relationships with well-known family members, caretakers, and familiar others, and initiates and responds to social interactions through gestural and emotional cues. Co-occurring sensory and physical impairments may prevent many social activities.

Reprinted with permission from the American Psychiatric Association. *Diagnostic and Statistical Manual of Mental Disorders*. 5th ed. Arlington: APA; 2013.

- c. Differential diagnosis includes ID, hearing loss, significant motor impairment, or severe mental health difficulties.

## 2. Interventions/Treatment

Referrals to speech-language pathology (SLP), audiology

## D. Learning Disabilities<sup>4</sup>

### 1. Definition

A heterogeneous group of deficits in an individual's ability to perceive or process information efficiently and accurately.

### 2. Diagnosis

- a. Achievement on standardized tests that is substantially below expected for age, schooling, and level of intelligence in one or more of the following areas: basic reading skills, reading comprehension, reading fluency skills, oral expression, listening comprehension, written expression, mathematic calculation, and mathematic problem solving.
- b. There is no alternative diagnosis such as sensory impairment or ID.<sup>6,7</sup>

### 3. Interventions/Treatment

School-based services through IEPs and 504 plans tailored to specific learning needs.

## E. Cerebral Palsy (CP)

### 1. Definition and Epidemiology

- a. A group of disorders of the development of movement and posture attributed to *non-progressive* disturbances that occurred in the developing fetal or infant brain.<sup>8</sup>
- b. Prevalence: 2 to 3/1000 live births<sup>2</sup>

### 2. Clinical Presentation

- a. Delayed motor development, abnormal tone, atypical postures, persistent primitive reflexes past 6 months.
- b. History of known or suspected brain injury.
- c. Manifestations may change with brain maturation and development.

### 3. Diagnosis

- a. Classification is based on physiologic and topographic characteristics as well as severity ([Table 9.9](#)).<sup>9</sup>
- b. Brain imaging should be obtained with magnetic resonance imaging (MRI); abnormal in 70% to 90% of individuals with CP.<sup>10</sup>

### 4. Interventions/Treatment

- a. Baseline and ongoing medical subspecialty care, including developmental pediatrics, neurology, orthopedics, and neurosurgery.
- b. Interdisciplinary team involvement (see [Section V](#)).
- c. Equipment to promote mobility and communication, including Augmentative and Alternative Communication - any form of communication other than oral speech ([Table EC 9.C](#)).<sup>11</sup>
  - (1) Augmentative Communication: Communication supports/methods used by individuals who have some speech but limited use of their speech.

**TABLE EC 9.C****TYPES OF ALTERNATIVE AND AUGMENTATIVE COMMUNICATION**

Type of AAC	Description	Formats	Access Method
Low-tech AAC	Generally paper-based supports Messages represented by gestures, symbols/photos, objects, words, phrases, or spelling with letters	Basic signs, pencil/paper (writing), eye pointing board, communication board or book, Velcro/magnet/pull-off messages	Direct selection with upper extremities/stylus/laser pointer/head-stick/mouth-stick/eye pointing Indirect selection with partner assisted scanning
Mid-tech AAC	Generally non-computer-based devices with recordable/digitized speech Messages represented by symbols/photos, words, phrases No spelling options; device can't blend letters together to make a spoken word May require physical dexterity to change pages Generally, more limited vocabulary possibilities as compared to high-tech AAC	Single button with single message or multiple messages to scan through Multi-level devices with changeable paper overlay Single level device with non-changing vocabulary overlay	Direct selection with fingers/hand/stylus/laser pointer/head-stick/mouth-stick Indirect selection with switch scanning
High-tech AAC	Computer-based devices with synthesized or digitized speech Messages could be represented by symbols, photos, words, phrases, or spelling Over 40 different picture-based vocabulary setups are available on the market, to match to patients' language and access needs	Tablet/smartphone/smart watch with AAC application Dedicated speech generating devices, talking word processor Non-dedicated/integrated devices with computer access options	Direct selection with fingers/hand/stylus/laser pointer/head-stick/mouth-stick/eye pointing/mouse Indirect selection with switch scanning

From personal communication with Tooley, Lauren M.S., CCC-SLP, Kennedy Krieger Institute and from Augmentative and Alternative Communication. American Speech-Language-Hearing Association, [https://www.asha.org/PRPSpecificTopic.aspx?folderid=8589942773&section=Key\\_Issues](https://www.asha.org/PRPSpecificTopic.aspx?folderid=8589942773&section=Key_Issues).

**TABLE 9.9****CLINICAL CLASSIFICATION OF CEREBRAL PALSY<sup>9</sup>**

Type	Pattern of Involvement
<b>I. SPASTIC (INCREASED TONE, CLASPED KNIFE, CLONUS, FURTHER CLASSIFIED BY DISTRIBUTION)</b>	
Bilateral spasticity	Diplegia (legs primarily affected)
	Quadriplegia (all four extremities impaired; legs worse than arms)
Unilateral spasticity	Hemiplegia (ipsilateral arm and leg; arm worse than leg)
	Monoplegia (one extremity, usually upper; probably reflects a mild hemiplegia)
<b>II. DYSKINETIC (LEAD-PIPE OR CANDLE-WAX RIGIDITY, VARIABLE TONE, ± CLONUS)</b>	
Dystonic	Complex disorders often reflecting basal ganglia pathology, resulting in involuntary and uncontrolled movements. May be focal or generalized.
Choreoathetoid	
<b>III. OTHER</b>	
Ataxic	Movement and tone disorders often reflecting cerebellar origin
Hypotonic	Usually related to diffuse, often severe cerebral and/or cerebellar cortical dysfunction. May be axial, appendicular, or generalized.
Rigid	Muscle contraction, seen in rare neurogenetic diseases

From Graham HK, Rosenbaum P, et al. Cerebral palsy. *Nat Rev Disease Primers*. 2016;2(15082).



- (2) Alternative Communication: Communication supports/methods used by individuals who have no speech.
- d. Pharmacotherapy for spasticity (e.g., botulinum toxin injections, baclofen), dyskinesia, hypersalivation (e.g., glycopyrrolate, scopolamine patch).<sup>12</sup>
- e. In carefully selected patients: Intrathecal baclofen, selective dorsal rhizotomy, deep brain stimulation.

## F. Autism Spectrum Disorders

### 1. Definition and Epidemiology

- a. Encompasses previously named disorders of autistic disorder (autism), Asperger disorder, childhood disintegrative disorder, and pervasive developmental disorder not otherwise specified (PDD-NOS).
- b. Increasing prevalence: 1 in 59 children in the United States has an autism spectrum disorder (ASD) in 2018.<sup>13,14</sup>
- c. Almost five times more common in males than females.<sup>13</sup>

### 2. Screening

- a. **Formal screening for ASD recommended at the 18- and 24-month visits** (see the AAP practice guidelines for more detailed recommendations).<sup>15</sup>
- b. Recommendation upheld by the AAP despite a U.S. Preventive Services Task Force (USPSTF) draft recommendation statement citing insufficient evidence for screening.<sup>16,17</sup>
- c. Evaluate using screening tools such as the **Modified Checklist for Autism in Toddlers (M-CHAT-R/F)** and **Childhood Autism Screening Test (CAST)** (see Table 9.4)

### 3. Diagnosis

- a. Symptoms vary by age, developmental level, language ability, and supports in place.
- b. Diagnostic criteria include<sup>4</sup>:

- (1) **Impaired social communication and interaction.**

*Examples:* Lack of joint attention behaviors (e.g., showing toys, pointing for showing), diminished eye contact, no sharing of emotions, lack of imitation

- (2) **Restricted repetitive patterns of behavior, interests, or activities.**

*Examples:* Simple motor stereotypies (hand flapping, finger flicking), repetitive use of objects (spinning coins, lining up toys), repetitive speech (echolalia), resistance to change, unusual sensory responses

- (3) Presentation in early childhood and significant limitation of functioning.

### 4. Interventions/Treatment

- a. Educational interventions, visual supports, naturalistic developmental behavioral interventions (integrating behavioral and child-responsive strategies to teach developmentally appropriate skills in a more natural and interactive setting).<sup>16,17</sup>
- b. Referral to SLP, OT/sensory-based interventions.

### G. Attention Deficit/Hyperactivity Disorder: See Chapter 24

## V. LONGITUDINAL CARE OF CHILDREN WITH DEVELOPMENTAL DISORDERS AND DISABILITIES

### A. Interdisciplinary Involvement

1. Neurodevelopmental pediatrician, child neurologist, developmental/behavioral pediatrician, other medical subspecialties as indicated (e.g., orthopedics for CP can be very important).
2. Genetic counseling for families of children with a genetic condition.
3. Psychologists for formal testing, counseling.
4. Rehabilitation and therapists, including physical therapy (PT), occupational therapy (OT), and SLP.
5. Educators

### B. Relevant Laws and Regulation

1. The **Individuals with Disabilities Education Act (IDEA)** sets forth regulations in the following areas for states that receive federal funding<sup>6,18</sup>:
  - a. Entitles all children with qualifying disabilities to a free and appropriate public education in the least restrictive environment.
  - b. **Early intervention services:** Infants and toddlers younger than 3 years may be referred for evaluation to receive developmental services. Eligibility criteria vary by state; see The National Early Childhood Technical Assistance Center ([www.ectacenter.org](http://www.ectacenter.org)) for details.

- c. **Qualifying disabilities:** Children aged 3 to 21 years with autism spectrum disorder, ID, specific learning disability (LD), hearing or visual impairment, speech or language impairment, orthopedic impairment, traumatic brain injury, emotional disturbance, or other health impairment are eligible.
  - d. **Individualized Education Program (IEP):** Written statement that includes a child's current capabilities, goals and how they will be measured, and services required. A comprehensive team is needed to develop and implement the IEP.
  - e. **Transition Services:** School systems must provide transitions services that prepare students for post-secondary activities and IEPs must include a statement of transition service needs starting no later than age 14. The student must be included in the IEP process starting at age 14.
2. **Head Start and Early Head Start:** Programs instituted by the federal government to promote school readiness of low-income children aged 3 to 5 years and younger than 3 years, respectively, within their communities.<sup>19</sup>
3. **Section 504** of the Rehabilitation Act of 1973 and the Americans with Disabilities Act (ADA) prohibit discrimination against individuals with any disability, more broadly defined as an impairment that limits function.<sup>20</sup>



## VI. TRANSITIONS FROM PEDIATRIC TO ADULT CARE FOR YOUTH WITH DEVELOPMENTAL DISABILITIES

### A. The Need

Research reveals health disparities between adults with developmental disabilities and those without. Disparities include:

1. Increased ED utilization
2. Lack of identified adult provider
3. Worse self-perception of health<sup>21</sup>

### B. The Role of the Pediatric Provider

AAP Consensus Statement on Transitions<sup>22,23</sup>:

1. Identify a health professional as point person to work with the youth and family on transition process.
2. **Create health care transition plan by age 14** with the youth and family.
3. Apply same guidelines for primary and preventive care for all adolescents and young adults.
4. Ensure affordable, continuous insurance coverage.

### C. Transition Domains

Transitions for young adults with disabilities occur across many domains of life and warrant support from an interdisciplinary team ([Table 9.10](#)).

**TABLE 9.10****TRANSITION DOMAINS FOR YOUTH WITH DEVELOPMENTAL DISORDERS AND DISABILITIES**

Transition Domain	Common Issues	Necessary Support Personnel/Services
Physical/Emotional Health	Difficulty identifying adult providers, retained in pediatric care, lost to follow-up, increased ED usage, insurance difficulties	Pediatrician, adult PCP, sub-specialists
Education/ Employment	Education services through IDEA end at 21 years old. Subsequent difficulty finding and engaging in post-secondary education and/or employment opportunities.	Educational team members (teachers, therapists), vocational rehab specialists, college counselors, post-secondary education programs
Legal/Financial	Difficulties with issues of SSI, guardianship, conservatorship	Attorney, legal counsel, family advocate
Housing/ Transportation	Access to accessible housing and transportation, development and ongoing support of skills needs for independent living	Life skills courses, group homes, independent living supports/ aides, resources through state departments of disability and the US Department of Housing and Urban Development, state mobility services
Leisure Pursuits/ Respite Care	Decreased structure of leisure pursuits with termination of school services at 21, increased burden on caregivers	Day programs, social engagement programs (e.g., Best Buddies, Special Olympics), respite care services for caregivers
Sexuality	Romantic and sexual relationships, vulnerability, reproductive rights, contraception, parenthood, access to appropriate screening and health care	Education team members (sexual education while in school), family members; OB/Gyn providers, adult healthcare providers

*ED*, Emergency department; *IDEA*, Individuals with Disabilities Education Act; *OB/Gyn*, obstetrician/gynecologist; *PCP*, primary care physician; *SSI*, supplemental security income.

## VII. WEB RESOURCES

- Autism Speaks: [www.autismspeaks.org](http://www.autismspeaks.org)
- Bright Futures: [www.brightfutures.org](http://www.brightfutures.org)
- Cerebral Palsy Foundation: [yourcpf.org](http://yourcpf.org)
- Disability Programs and Services: <https://www.dol.gov/odep/topics/disability.htm>
- Got Transition: [www.gotransition.org](http://www.gotransition.org)
- Individuals with Disabilities Education Act (IDEA): [idea.ed.gov](http://idea.ed.gov)
- Intellectual Disability: [aidd.org](http://aidd.org)
- National Center for Learning Disabilities: [www.ncld.org](http://www.ncld.org)
- National Early Childhood Technical Assistance Center: [www.ectacenter.org](http://www.ectacenter.org)
- Reach Out and Read: [www.reachoutandread.org](http://www.reachoutandread.org)

## REFERENCES

A complete list of references can be found online at [www.expertconsult.com](http://www.expertconsult.com).



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# Chapter 10

## Endocrinology

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 See additional content on Expert Consult

### I. DIABETES

#### A. Diagnosis of Diabetes Mellitus<sup>1-3</sup>

Diagnostic criteria (must meet one of four):

1. Symptoms of diabetes (polyuria, polydipsia, weight loss, frequent yeast infections) and random blood glucose (BG)  $\geq 200$  mg/dL
2. Fasting plasma glucose (FPG = no caloric intake for at least 8 hours)  $\geq 126$  mg/dL
3. Oral glucose tolerance test (OGTT) with a 2-hour post-load plasma glucose of  $\geq 200$  mg/dL
4. Hemoglobin A<sub>1c</sub> (HbA<sub>1c</sub>)  $\geq 6.5\%$

**NOTE:** In the absence of symptoms of hyperglycemia, FPG or OGTT should be repeated on another day.

#### B. Definition of Increased Risk (Prediabetes)

1. FPG 100 to 125 mg/dL
2. 2-hour post-OGTT 140 to 199 mg/dL
3. HbA<sub>1c</sub> 5.7% to 6.4%

#### C. Interpreting Hemoglobin A<sub>1c</sub><sup>1,2</sup>

1. Estimates average BG for the past 3 months.
2. HbA<sub>1c</sub> of 6% approximately equals an average BG of 130 mg/dL; each additional 1%  $\approx$  30 mg/dL more.
3. Unreliable in patients with abnormal red cell lifespan or morphology (e.g., sickle cell disease, spherocytosis).
4. Although the HbA<sub>1c</sub> criterion has been accepted by the American Diabetes Association for the diagnosis of diabetes in adults, this criterion remains controversial in children, especially as it relates to type 2 diabetes mellitus (T2DM).

#### D. Etiology: Distinguishing Between Types of Diabetes Mellitus<sup>1,2</sup>

1. Type 1 (T1DM) versus T2DM (most common types, polygenic; [Table 10.1](#))
2. Other forms of diabetes<sup>4,5</sup>
  - a. Monogenic diabetes: 1% to 2% of diabetes mellitus (DM). Due to single-gene mutations, typically relating to insulin production or release. Identifying gene can have clinical significance.
    - (1) Suspect if autosomal dominant inheritance pattern of early-onset (<25 years) DM, insulin independence, absent T2DM phenotype (non-obese), or preservation of C-peptide.

**TABLE 10.1****CHARACTERISTICS SUGGESTIVE OF TYPE 1 VERSUS TYPE 2 DIABETES**

Characteristic	Type 1	Type 2
Onset	As early as 1-year-old through adulthood	Usually post-pubertal
Polydipsia and polyuria	Present for days to weeks	Absent or present for weeks to months
Ethnicity	Caucasian	African American, Hispanic, Asian, Native American
Weight	Weight loss	Obese (although weight loss is common in presentation with severe hyperglycemia)
Other physical findings		Acanthosis nigricans
Family history	Autoimmune diseases	Type 2 diabetes
Ketoacidosis	More common (1/3 at onset)	Less common (6% at onset)
Lab characteristics	Autoantibodies common; C-peptide generally should be unmeasurable >2 years after diagnosis	Autoantibodies less common, but sometimes present

(2) Well-described subtypes: MODY1 and MODY3, due to mutations in transcription factors for insulin production; responsive to sulfonylureas.

- b. Neonatal diabetes (NDM): Defined as DM onset <6 months of age.
  - (1) Rare: 1:160,000 to 260,000 live births, typically a de novo mutation
  - (2) May be transient (50% recur) or permanent
  - (3) Subset respond to sulfonylureas
- c. Cystic fibrosis-related diabetes (CFRD): OGTT rather than HbA<sub>1c</sub> is the recommended screening test.
- d. Other causes of DM: Diseases of exocrine pancreas due to pancreatitis, trauma, infection, invasive disease (e.g., hemochromatosis).

### E. Screening for Type 2 Diabetes Mellitus<sup>1,6</sup>

1. **Who to screen:** Children who are overweight [body mass index (BMI) >85th percentile] and have one or more of the following risk factors:
  - a. Maternal history of diabetes or gestational diabetes mellitus during child's gestation
  - b. Family history of T2DM in a first- or second-degree relative
  - c. Race/ethnicity: African American, Native American, Hispanic, Asian, or Pacific Islander
  - d. Signs associated with insulin resistance (acanthosis nigricans, hypertension, dyslipidemia, polycystic ovarian syndrome, or small-for-gestational-age birth weight)
2. **How to screen:** Fasting plasma glucose, OGTT, or HbA<sub>1c</sub>
3. **When to screen:** Begin at the age of 10 years or at the onset of puberty (whichever occurs first), and repeat at a minimum of every 3 years or more often if BMI is increasing.

**TABLE 10.2**  
**SUBCUTANEOUS INSULIN DOSING**

	Insulin	Dose Calculation	Sample Calculation for 24-kg Child	Dose
Total daily dose		0.5–1 unit/kg/day	$0.75 \times 24 = 18$ units/day	18 units
Basal	Glargine <i>OR</i>	1/2 daily total	$\frac{1}{2} \times 18 \text{ units} = 9$	9 units daily
	Detemir	1/2 daily total ÷ BID	$\frac{1}{2} \times 18 \text{ units} = 9$	4.5 units BID
Carbohydrate coverage ratio	Lispro, aspart <i>OR</i>	500 ÷ daily total	$500 \div 18 = 28$	1 unit: 28 g carbohydrate
	Regular	450 ÷ daily total	$450/18 = 25$	1 unit: 25 g carbohydrate
Correction factor	Lispro, aspart <i>OR</i>	1800 ÷ daily total	$1800 \div 18 = 100$	1 unit expected to drop BG by 100 mg/dL
	Regular	1500 ÷ daily total	$1500/18 = 83$	1 unit expected to drop BG by 83 mg/dL

## F. Additional Testing in New-Onset Diabetes

- Diabetes autoantibodies<sup>1,2</sup>:** Recommended for all children with suspected T2DM. No universal agreement regarding whether to test all patients.
  - Includes islet cell autoantibodies (ICAs) and antibodies to GAD (GAD65), insulin, and tyrosine phosphotases IA-2, IA-2 $\beta$ , ZnT8.
  - Suggestive of T1DM if present, though about 5% of T1DM will not have measurable ICAs, and some children with T2DM will have measureable ICAs.
- Screening for autoimmune diseases in T1DM<sup>6</sup>:**
  - Thyroid disease (present in 17% to 30% of patients with T1DM): Screen with TSH when clinically well and consider screening for thyroid antibodies. If TSH normal, recheck every 1 to 2 years or sooner if symptoms develop.
  - Celiac disease (present in 1.6% to 16.4% of patients with T1DM): Screen with tissue transglutaminase (TTG) IgA antibody and total IgA. Repeat within 2 years of diabetes diagnosis and again after 5 years. Repeat more frequently if there are symptoms or a first degree relative with celiac disease.

## G. Management of Diabetes<sup>6-8</sup>

- Diabetes medications FDA-approved for children:**
  - Insulin: See Tables 10.2 and 10.3 for calculations. Insulin doses are subsequently adjusted based on actual blood sugars.
  - Metformin: FDA-approved in children  $\geq 10$  years old, though sometimes used off-label in younger children. Main side effects are

**TABLE 10.3****TYPES OF INSULIN PREPARATIONS AND SUGGESTED ACTION PROFILES FOR SUBCUTANEOUS ADMINISTRATION<sup>54</sup>**

Insulin <sup>a</sup>	Onset	Peak	Effective Duration
Ultra rapid acting analog (faster aspart)	5–10 min	1–3 hr	3–5 hr
Rapid acting (lispro, aspart, glulisine)	10–20 min	1–3 hr	3–5 hr
Short acting (regular)	30–60 min	2–4 hr	5–8 hr
Intermediate acting (NPH)	2–4 hr	4–12 hr	12–24 hr
Long acting			
Glargine	2–4 hr	8–12 hr	22–24 hr
Detemir	1–2 hr	4–7 hr	20–24 hr
Degludec	30–90 min	No peak	>42 hr

<sup>a</sup>Assuming 0.1–0.2 U/kg per injection. Onset and duration vary significantly by injection site.

**NOTE:** Be aware that there are stronger concentrations of various types of insulin available (e.g., U-500 regular insulin, which is 5 times more concentrated than U-100 regular insulin; U-300 insulin glargine). There are also pre-mixed combinations of rapid or short AND intermediate acting insulin (e.g., 70% NPH/30% regular [Humulin 70/30]).

NPH, Neutral protamine Hagedorn.

Modified from The American Diabetes Association. *Practical Insulin: A Handbook for Prescribing Providers*. 2nd ed. Alexandria, VA: American Diabetes Association; 2007.

gastrointestinal and are often transient. Extended release option available for patients with GI side effects.

2. **T1DM management:** The majority of children with T1DM should be treated with intensive insulin regimens, either via multiple daily injections or continuous subcutaneous infusion.
3. **T2DM management:**
  - a. Lifestyle modification therapy (nutrition and physical activity) and metformin should be initiated at time of diagnosis.
  - b. Insulin therapy should be initiated if distinction between T1DM and T2DM is unclear, when  $\text{HbA}_{1c} \geq 8.5\%$ , when random BG  $\geq 250$ , or when patient with known T2DM is not meeting glycemic target with metformin and lifestyle modification alone. **NOTE:** If significant hyperglycemia (BG  $> 600$ ) or ketosis is present, patient should be evaluated for DKA/HHS.
4. **Goals of therapy:** A1c  $<7.5\%$  for T1DM and  $<7\%$  for T2DM in patients on metformin alone (individualized to avoid excessive hypoglycemia).
5. Interdisciplinary care team should include mental health provider and medical nutrition therapy with initial education and annual update. Regularly assess for eating disorders, disease-related coping, depression, and psychosocial stressors impacting diabetes management.

**H. Diabetes-Related Devices<sup>9,10</sup>**

1. Technology is rapidly changing, but general principles are described below.
2. **Insulin pumps:** Contain rapid acting insulin only and provide basal and bolus insulin. Doses can be programmed to vary throughout the day. Settings consist of:

- a. Basal rate—continuous insulin infusion
- b. Carbohydrate coverage—insulin to carbohydrate ratio
- c. Hyperglycemia correction—based on correction factor and target blood glucose

**NOTE:** There is risk for DKA with interruptions in insulin delivery (e.g., pump malfunction) given lack of long-acting insulin.

- 3. **Continuous glucose monitors (CGMs):** Measure glucose concentration in interstitial fluid continuously and provide alerts for high and low glucose levels.
- 4. **Sensor augmented insulin pump therapy:** Integration of continuous glucose monitor and insulin pump to adjust insulin delivery based on blood glucose.

## I. Monitoring<sup>6,8,9,11</sup>

### 1. Glycemic control:

- a. Assessment of blood glucose using glucometer or CGM—multiple times daily (before meals/snacks, at bedtime, prior to exercise, with symptoms of hypoglycemia, after treating for hypoglycemia, before driving, etc.)
  - b. HbA<sub>1c</sub> every 3 months
2. Urine ketones should be checked with persistent hyperglycemia, any illness (regardless of blood glucose level), or with nausea/vomiting.

### 3. Associated conditions or complications: See Table 10.4.

## J. Diabetic Emergencies<sup>12,13</sup>

### 1. Diabetic ketoacidosis (DKA)

- a. Definition: Hyperglycemia (or euglycemia in a patient with known diabetes), ketonemia, ketonuria, and metabolic acidosis ( $\text{pH} < 7.30$ , bicarbonate  $< 15 \text{ mEq/L}$ )
- b. BG reflects hydration status, pH reflects DKA severity
- c. Symptoms: Nausea, emesis, abdominal pain, fruity breath, altered mental status, Kussmaul respirations
- d. Precipitating factors: New-onset DM, known diabetes with missed insulin doses, insulin pump/infusion site malfunction, or physiologic stress due to acute illness
- e. Management: See Fig. 10.1. Because the fluid and electrolyte requirements vary greatly from patient to patient, guidelines are only a starting point and therapy must be individualized based on patient characteristics. **NOTE:** Initial insulin administration may cause transient worsening of the acidosis as potassium is driven into cells in exchange for hydrogen ions.
- f. Cerebral edema: Most severe complication of DKA. Overly aggressive hydration and rapid correction of hyperglycemia may play a role in its development. Risk factors include severe acidosis, evidence of renal insufficiency, young age and new onset, use of bicarbonate.

**TABLE 10.4****SCREENING FOR DIABETES-ASSOCIATED CONDITIONS AND COMPLICATIONS<sup>6,11</sup>**

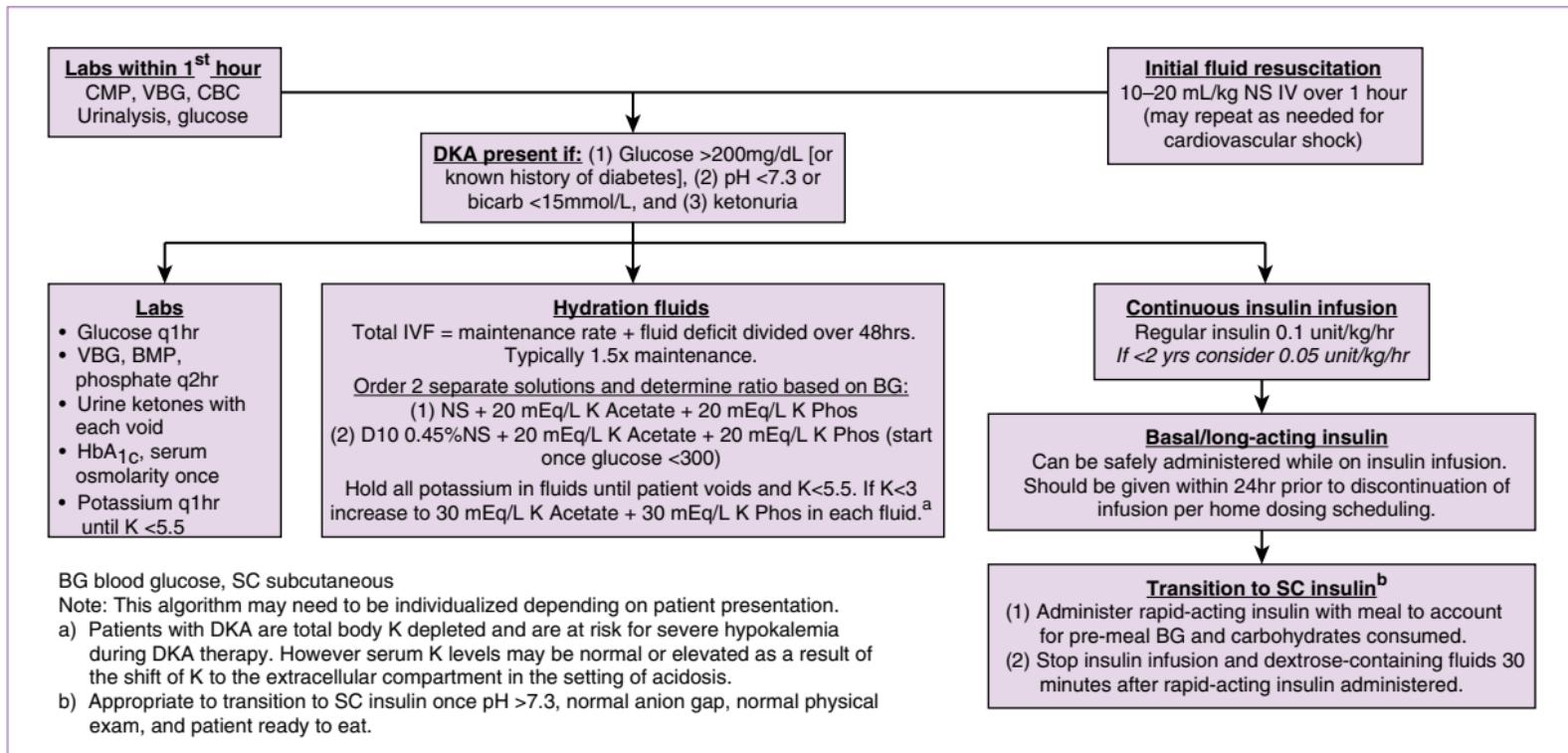
Type of Condition or Complication	Screening Test	Frequency
Hypertension	Blood pressure measurement	At every visit
Hyperlipidemia	Lipid profile	At diagnosis, then yearly if T2DM or T1DM and overweight; every 5 years if low-density lipoprotein [LDL] <100 mg/dL
Retinopathy	Dilated eye examination	T1DM: yearly after 3–5 years of diabetes, provided ≥ age 10; T2DM: at diagnosis, yearly
Diabetic nephropathy	Random spot urine microalbumin-to-creatinine ratio	T1DM: yearly after 5 years of diabetes, provided ≥ age 10; T2DM: at diagnosis, yearly
Neuropathy	Foot exam	T1DM: yearly after 5 years of diabetes, provided ≥ age 10; T2DM: at diagnosis, yearly
Nonalcoholic steatohepatitis (NASH)	ALT, AST	T2DM: at diagnosis, yearly
Obstructive sleep apnea (OSA)	Review of symptoms	T2DM: at every visit
Polycystic ovary syndrome (PCOS)	Menstrual history ± lab evaluation	T2DM: at every visit

ALT, Alanine amino transferase; AST, aspartate amino transferase; T1DM, type 1 diabetes mellitus; T2DM, type 2 diabetes mellitus.

- g. Once DKA is resolved, transition to subcutaneous (SQ) insulin. See Tables 10.2 and 10.3 for calculations or resume home insulin doses.

## 2. Hyperglycemic hyperosmolar state (HHS)

- Definition: Extreme hyperglycemia (BG >600 mg/dL) and hyperosmolarity (>320 mOsm/kg), **without significant ketosis or acidosis**.
- Characteristics of HHS: Gradually increasing polyuria and polydipsia leading to profound dehydration, altered consciousness.
- Management:
  - Fluids: Fluids alone will decrease BG due to dilution, promotion of glucosuria, and increased glucose uptake with improved circulation. Fluid replacement should be more rapid than in DKA with goal of gradual decline in serum sodium (about 0.5 mEq/dL/h) and osmolality. Bolus ≥20 cc/kg 0.9% saline and repeat until perfusion improved. Then start maintenance fluids plus deficit replacement over 24 to 48 hours using 0.45% to 0.75% saline (if perfusion inadequate, consider isotonic fluids). Urine output should also be replaced.
  - Insulin therapy: Start insulin (0.025 to 0.05 unit/kg/h) when BG no longer declining at least 50 mg/dL/h with fluids alone. Titrate insulin to decrease BG by 75 to 100 mg/dL/h.



BG blood glucose, SC subcutaneous

Note: This algorithm may need to be individualized depending on patient presentation.

- Patients with DKA are total body K depleted and are at risk for severe hypokalemia during DKA therapy. However serum K levels may be normal or elevated as a result of the shift of K to the extracellular compartment in the setting of acidosis.
- Appropriate to transition to SC insulin once pH >7.3, normal anion gap, normal physical exam, and patient ready to eat.

**FIGURE 10.1**

Management of Diabetic Ketoacidosis. (Modified from Cooke DW, Plotnick L. Management of diabetic ketoacidosis in children and adolescents. *Pediatr Rev.* 2008;29:431–436.)

**TABLE 10.5****AGE-BASED NORMAL VALUES FOR ROUTINE THYROID FUNCTION TESTS**

Test	Age	Normal Range	
TSH (mIU/L)	Birth–6 days	0.70–15.2	
	1 week–3 months	0.72–11.0	
	3 months–12 months	0.73–8.35	
	1–5 years	0.70–5.97	
	6–10 years	0.60–4.84	
	>10 years	0.45–4.50	
Free T <sub>4</sub> (ng/dL)	Birth–3 days	0.66–2.71	
	4–30 days	0.83–3.09	
	31 days–12 months	0.48–2.34	
	13 months–5 years	0.85–1.75	
	6–10 years	0.90–1.67	
	11–19 years	0.93–1.60	
	>19 years	0.82–1.77	
Total T <sub>4</sub> (mCg/dL)		Male	Female
	<1 months	4.5–17.2	4.5–17.2
	1–23 months	5.9–13.9	5.9–13.9
	2–12 years	5.7–11.6	5.7–11.6
	13–20 years	5.1–10.3	5.3–11.7
	>20 years	4.9–10.5	5.1–11.9

T<sub>4</sub>, Thyroxine; TSH, thyroid-stimulating hormone.

NOTE: If age-specific reference ranges are provided by the laboratory that is running the assay, please refer to those ranges.

TSH and Free T<sub>4</sub> reference ranges from Labcorp; Total T<sub>4</sub> reference range from Quest Diagnostics.

- (3) Electrolytes: Potassium, phosphate, and magnesium deficits greater than in DKA; monitor every 2 to 4 hours. Start potassium replacement with 40 mEq/L once K <5 mEq/L.

## II. THYROID GLAND<sup>14–16</sup>

### A. Thyroid Tests<sup>15,17,18</sup>

1. **Normal thyroid function values:** See reference values for age (Table 10.5). Preterm infants have different ranges (Table 10.6).
2. **Interpretation of abnormal thyroid function values:** See Table 10.7.
3. **Imaging studies:**
  - a. Thyroid ultrasound: Most useful in assessing thyroid nodules for features suspicious for malignancy.
  - b. Thyroid uptake scan: Measures uptake of Technetium (<sup>99m</sup>Tc) pertechnetate or radioactive iodine by metabolically active thyroid tissue, helping to identify etiology of hyperthyroidism.

### B. Hypothyroidism

1. **Types of hypothyroidism:** Can be either congenital or acquired and either primary or central. See Table 10.8 for details on identification and management.

**TABLE 10.6****MEAN TSH AND T<sub>4</sub> OF PRETERM AND TERM INFANTS 0–28 DAYS<sup>18</sup>**

Age ± SD	Cord (Day 0)	Day 7	Day 14	Day 28
<b>T<sub>4</sub> (mCg/DL)</b>				
23–27 <sup>a</sup>	5.44 ± 2.02	4.04 ± 1.79	4.74 ± 2.56	6.14 ± 2.33
28–30	6.29 ± 2.02	6.29 ± 2.10	6.60 ± 2.25	7.46 ± 2.33
31–34	7.61 ± 2.25	9.40 ± 3.42	9.09 ± 3.57	8.94 ± 2.95
>37	9.17 ± 1.94	12.67 ± 2.87	10.72 ± 1.40	9.71 ± 2.18
<b>FT<sub>4</sub> (NG/DL)</b>				
23–27	1.28 ± 0.41	1.47 ± 0.56	1.45 ± 0.51	1.50 ± 0.43
28–30	1.45 ± 0.43	1.82 ± 0.66	1.65 ± 0.44	1.71 ± 0.43
31–34	1.49 ± 0.33	2.14 ± 0.57	1.96 ± 0.43	1.88 ± 0.46
>37	1.41 ± 0.39	2.70 ± 0.57	2.03 ± 0.28	1.65 ± 0.34
<b>TSH (MIU/L)</b>				
23–27	6.80 ± 2.90	3.50 ± 2.60	3.90 ± 2.70	3.80 ± 4.70
28–30	7.00 ± 3.70	3.60 ± 2.50	4.90 ± 11.2	3.60 ± 2.50
31–34	7.90 ± 5.20	3.60 ± 4.80	3.80 ± 9.30	3.50 ± 3.40
>37	6.70 ± 4.80	2.60 ± 1.80	2.50 ± 2.00	1.80 ± 0.90

<sup>a</sup>Weeks gestational age.T<sub>4</sub>, Free thyroxine; T<sub>3</sub>, thyroxine; TSH, thyroid-stimulating hormone.

Data modified from Williams FL, Simpson J, Delahunty C, et al. Collaboration from The Scottish Preterm Thyroid Group:

Developmental trends in cord and postpartum serum thyroid hormones in preterm infants. *J Clin Endocrinol Metab*.

2004;89:5314–5320.

**TABLE 10.7****THYROID FUNCTION TESTS: INTERPRETATION**

Disorder	TSH	T <sub>4</sub>	Free T <sub>4</sub>
Primary hyperthyroidism	L	H	High N to H
Primary hypothyroidism	H	L	L
Hypothalamic/pituitary hypothyroidism	L, N, H <sup>a</sup>	L	L
TBG deficiency	N	L	N
Euthyroid sick syndrome	L, N, H <sup>a</sup>	L	L to low N
TSH adenoma or pituitary resistance	N to H	H	H
Subclinical hypothyroidism <sup>b</sup>	H	N	N

<sup>a</sup>Can be normal, low, or slightly high.<sup>b</sup>Treatment may not be necessary.H, High; L, low; N, normal; T<sub>4</sub>, thyroxine; TBG, thyroxine-binding globulin; TSH, thyroid-stimulating hormone.

2. **Subclinical hypothyroidism and obesity**<sup>19</sup>: Moderate elevations in thyroid-stimulating hormone (TSH [4 to 10 mIU/L]), with normal or slightly elevated triiodothyronine (T<sub>3</sub>) and thyroxine (T<sub>4</sub>) are seen in 10% to 23% of obese children. There does not appear to be a benefit to treating these individuals. Values tend to normalize with weight loss. Could consider testing for thyroid antibodies to further clarify whether there is true thyroid dysfunction.

**TABLE 10.8****HYPOTHYROIDISM<sup>55,56</sup>**

Clinical Symptoms	Onset	Etiology	Management	Follow-up
<b>PRIMARY/CONGENITAL</b>				
Large fontanelles, lethargy, constipation, hoarse cry, hypotonia, hypothermia, jaundice. Most often picked up on newborn screen.	Symptoms usually develop by 2 weeks; almost always by 6 weeks. Some infants may be relatively asymptomatic if not caused by absence of thyroid gland.	<b>Primary:</b> Defect of fetal thyroid development most common. Other causes include TSH receptor mutation or thyroid dyshormonogenesis. <i>OR</i> <b>Central:</b> Deficiency of TSH or thyrotropin-releasing hormone (TRH).	Replacement with L-thyroxine once newborn screen is positive, pending results of confirmatory testing. <sup>a</sup> Goal T <sub>4</sub> in upper half of normal range. In primary hypothyroidism, TSH should be kept in normal range for age.	Monitor T <sub>4</sub> and TSH 1–2 weeks after initiation and then every 2 weeks until TSH normalizes. Once levels are adequate follow per schedule listed below. Treated patients are still at risk for developmental delay.
<b>ACQUIRED</b>				
Growth deceleration, coarse brittle hair, dry skin, delayed tooth eruption, cold intolerance.	Can occur as early as 2 years old.	<b>Primary:</b> Can be caused by Hashimoto thyroiditis (diagnosis supported by + antithyroglobulin or antimicrosomal antibodies), head/neck radiation. <i>OR</i> <b>Central:</b> Caused by pituitary/hypothalamic insults including brain tumor.	Replacement with L-thyroxine. <sup>a</sup> Targets for TSH and T <sub>4</sub> same as for congenital hypothyroidism above.	Follow every 1–3 months during the first 12 months, every 2–4 months until 3 years, and then every 3–12 months until growth complete. Follow 4–6 weeks after any dose change.

<sup>a</sup>Because of the risk of inducing adrenal crisis if adrenocorticotrophic hormone (ACTH) deficiency is present, the treatment of central hypothyroidism *should not* be started until normal ACTH/cortisol function is documented.

**NOTE:** Thyroid hormone levels in premature infants are lower than those seen in full-term infants. Furthermore, the TSH surge seen at approximately 24 hours of age in full-term babies does not appear in preterm infants. In this population, lower levels are associated with increased illness; however, the effect of replacement therapy remains controversial.

L-thyroxine, Levothyroxine; TSH, thyroid-stimulating hormone.

**TABLE 10.9**  
**HYPERTHYROIDISM**

Presentation	Distinguishing Imaging/Lab Findings	Management
<b>GRAVES DISEASE</b>		
Typical symptoms of hyperthyroidism plus diffuse goiter, eye symptoms, localized dermopathy, and lymphoid hyperplasia	TSH is often undetectable. ↑ $^{99m}\text{Tc}$ -pertechnate uptake. Positive TSI.	First-line treatment in children is methimazole. Radioactive iodine ( $^{131}\text{I}$ ) or surgical thyroidectomy are options for initial treatment or refractory cases. Follow symptoms, $\text{T}_4$ , and TSH levels.
<b>HASHIMOTO THYROIDITIS</b>		
± Initial hyperthyroidism, followed by eventual thyroid burnout and hypothyroidism.	Often low but detectable TSH and less significant increase in $\text{T}_4$ . ↓ $^{99m}\text{Tc}$ -pertechnate uptake. Significant elevation of thyroglobulin and/or microsomal antibody.	Hyperthyroid phase is usually self-limited; patient may eventually need thyroid replacement therapy. Propranolol if symptomatic during hyperthyroid phase.

$^{99m}\text{Tc}$ , Technetium;  $\text{T}_4$ , thyroxine; TSH, thyroid stimulating hormone; TSI, thyroid stimulating immunoglobulin.

3. **Newborn screening for hypothyroidism**<sup>16,20</sup>: Mandated in all 50 states. Measures a combination of TSH and  $\text{T}_4$ , based on the particular state's algorithm; 1:25 abnormal tests are confirmed. Congenital hypothyroidism has prevalence of 1:3000 to 1:4000 U.S. infants. If abnormal results are found, clinicians should follow recommendations of the American College of Medical Genetics ACT Sheets and Algorithm for confirmation testing.

### C. Hyperthyroidism

- General features:**
  - Epidemiology: Prevalence increases with age, rare before adolescence; female-to-male predominance.
  - Etiology: Most common cause is Graves disease, followed by subacute thyroiditis. Less common causes are Hashitoxicosis, autonomously functioning thyroid nodule, factitious hyperthyroidism (intake of exogenous hormone), TSH-secreting pituitary tumor (rare), and pituitary resistance to thyroid hormone. See Table 10.9 for comparison of Graves and Hashimoto disease.
  - Laboratory findings: See Table 10.7. Further tests include TSH receptor-stimulating antibody, thyroid-stimulating immunoglobulin (TSI), antithyroglobulin and antimicrosomal (thyroid peroxidase) antibodies.

**2. Thyroid storm<sup>21</sup>:**

- a. Presentation: Acute onset of hyperthermia, tachycardia, and restlessness. May progress to delirium, coma, and death.
- b. Treatment: Admission to ICU. Emergent pediatric endocrinology consultation recommended. Therapy aimed at relieving symptoms (propranolol) and reducing peripheral conversion of T4 to T3 (hydrocortisone), thyroid hormone production (antithyroid drugs), release of hormone from thyroid gland (potassium iodide), and reabsorption from enterohepatic circulation (cholestyramine).

**3. Neonatal thyrotoxicosis:**

- a. Presentation: Microcephaly, frontal bossing, intrauterine growth restriction (IUGR), tachycardia, systolic hypertension leading to widened pulse pressure, irritability, failure to thrive, exophthalmos, goiter, flushing, vomiting, diarrhea, jaundice, thrombocytopenia, and cardiac failure or arrhythmias. Onset from immediately after birth to weeks.
- b. Etiology: Occurs exclusively in infants born to mothers with Graves disease. Caused by transplacental passage of maternal TSI. Occasionally, mothers are unaware they have Graves. Even if a mother has received definitive treatment (thyroidectomy or radiation therapy), passage of TSI remains possible.
- c. Treatment and monitoring: Propranolol for symptom control. Methimazole to lower thyroxine levels. Digoxin may be indicated for heart failure. Disease usually resolves by 6 months of age.

### III. PARATHYROID GLAND AND VITAMIN D<sup>22-24</sup>

#### A. Parathyroid Hormone Function

1. Increases serum calcium by increasing bone resorption
2. Increases calcium and magnesium reuptake and phosphorus excretion in the kidney
3. Increases 25-hydroxy vitamin D conversion to 1,25-dihydroxy vitamin D, in order to increase calcium absorption in the intestine

#### B. Distinguishing Between Abnormalities Related to Parathyroid Hormone and Vitamin D

See [Table 10.10](#).

#### C. Vitamin D Supplementation

Please see [Chapter 21](#) for additional information.

### IV. ADRENAL GLAND<sup>25-29</sup>

#### A. Adrenal Insufficiency

##### 1. Causes of adrenal insufficiency:

- a. Impaired steroidogenesis, as in congenital adrenal hyperplasia.
- b. Adrenal destruction or dysfunction as in primary adrenal insufficiency (AI) (Addison disease), autoimmune polyendocrine syndrome, or adrenoleukodystrophy.

**TABLE 10.10****DISTINGUISHING BETWEEN DISORDERS OF PARATHYROID GLANDS AND VITAMIN D REGULATION**

	Hypoparathyroidism	Hyperparathyroidism	PTH Resistance/ Pseudo-Hypoparathyroidism	Vitamin D Deficiency
PTH	↓ or inappropriately normal in the setting of hypocalcemia	↑	↑	-/↑
1,25-D	↓	↑	↓	-
25-OHD	-	-/↓	-	-/↓
Calcium	↓	↑	↓	-/↓
Phosphorus	↑	↓	↑	-/↓
Alkaline Phosphate	-/↓	-/↑	↑	↑
Common Causes	DiGeorge, autoimmune (APS), iatrogenic	Primary: Adenoma, hyperplasia Secondary: Renal failure, rickets	Genetic mutations	Nutritional deficiency
First line Rx	Calcium, calcitriol	Hydration with normal saline, surgical resection	Calcitriol	Vitamin D +/- calcium

1,25-D, 1,25 dihydroxy vitamin D; 25-OHD, 25-hydroxy vitamin D; APS, autoimmune polyendocrine syndrome; MEN, multiple endocrine neoplasia; PTH, parathyroid hormone; Rx, treatment.

**TABLE 10.11****17-HYDROXYPROGESTERONE, SERUM**

Age	Baseline (ng/dL)
Premature (31–35 weeks)	≤360
Term infants (3 days)	≤420
1–12 months	11–170
1–4 years	4–115
5–9 years	≤90
10–13 years	≤169
14–17 years	16–283
Males, Tanner II–III	12–130
Females, Tanner II–III	18–220
Males, Tanner IV–V	51–190
Females, Tanner IV–V	36–200
Male (18–30 years)	32–307
Adult female	
Follicular phase	≤185
Midcycle phase	≤225
Luteal phase	≤285

Reference ranges from Quest Diagnostics LC/MS assay (liquid chromatography/tandem mass spectroscopy).

For preterm infants or infants born small for gestational age, see Olgemöller B, Roscher AA, Liebl B, et al. Screening for congenital adrenal hyperplasia: adjustment of 17-hydroxyprogesterone cut-off values to both age and birth weight markedly improves the predictive value. *J Clin Endocrinol Metab.* 2003;88:5790–5794.

- c. Secondary AI caused by impaired circulating adrenocorticotrophic hormone (ACTH) due to hypothalamic or pituitary pathology.
- d. Acquired insufficiency secondary to long-term corticosteroid use leading to HPA suppression. **NOTE:** This is the most common cause seen in clinical practice and may also occur in setting of chronic high-dose inhaled corticosteroids.

### 2. Laboratory findings in adrenal insufficiency:

- a. In primary AI, there is deficient mineralocorticoid and glucocorticoid production. In central AI, there is only deficient glucocorticoid production, and mineralocorticoid production is normal.
- b. Primary AI: Elevated ACTH, elevated plasma renin activity, low cortisol, low aldosterone, hypoglycemia, hyponatremia, hyperkalemia.
- c. Central AI: Normal/low ACTH, normal plasma renin activity (no impairment of mineralocorticoid function), low cortisol, normal aldosterone, hyponatremia, hypoglycemia.
- d. In infants with congenital adrenal hyperplasia (CAH), 17-hydroxyprogesterone (17-OHP) is increased (see Table 10.11 for normal values by age).

### 3. Diagnosis of adrenal insufficiency:

- a. Initial screening with AM cortisol level, which may be drawn concomitantly with an ACTH level.
- b. See Table 10.12 for interpretation of AM cortisol results.

**TABLE 10.12****CORTISOL, 8 AM**

Interpretation	Cortisol (mCg/dL)
Suggestive of adrenal insufficiency	<5 mCg/dL
Indeterminate	5–14 mCg/dL
Adrenal insufficiency unlikely	>14 mCg/dL

**BOX 10.1****PERFORMANCE AND INTERPRETATION OF ACTH STIMULATION TEST****Standard Dose ACTH Stimulation Test**

Obtain initial baseline cortisol level

Give 250 mg IV ACTH

Measure cortisol at 30 min

Measure cortisol at 60 min

**Interpretation of Results****For evaluation of primary adrenal insufficiency:**

<18 mCg/dL: Highly suggestive of adrenal insufficiency

≥18 mCg/dL: Normal (rules out adrenal insufficiency)

**For evaluation of central adrenal insufficiency:**

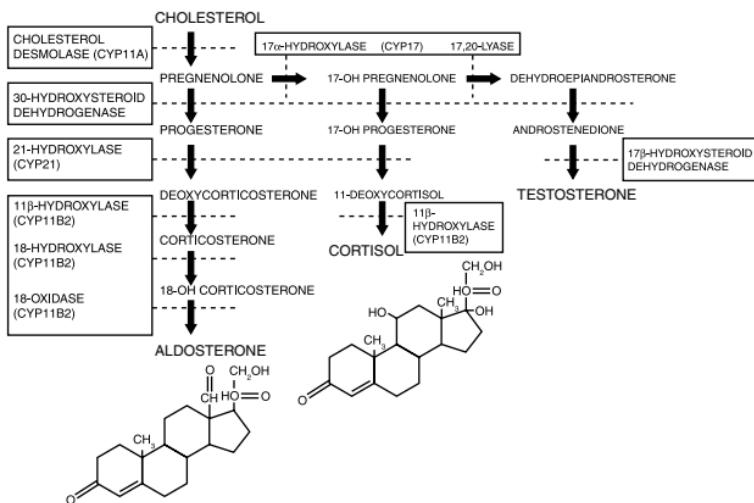
<16 mCg/dL: Highly suggestive of adrenal insufficiency

16–30 mCg/dL: Adrenal insufficiency less likely but not excluded

>30 mCg/dL: Normal (rules out adrenal insufficiency)

**NOTE:** No test for adrenal insufficiency has perfect sensitivity or specificity, so results must be interpreted in the individual clinical context. Measurement of serum ACTH is also beneficial (elevated in Addison's, low/normal in secondary insufficiency).

- c. Plasma ACTH elevation >100 pg/mL with concomitant hypocortisolism <10 µg/dL is consistent with glucocorticoid deficiency due to primary AI.
  - d. Standard dose ACTH stimulation test is used to confirm diagnosis.
4. **ACTH stimulation test:**
- a. In brief, with ACTH deficiency or prolonged adrenal suppression, there is an inadequate rise in cortisol after a single ACTH dose.
  - b. See [Box 10.1](#) for interpretation of ACTH stimulation test.
5. **Congenital adrenal hyperplasia (CAH):**
- a. See [Fig. 10.2](#).
  - b. Group of autosomal recessive disorders characterized by a defect in one of the enzymes required in the synthesis of adrenal hormones.
  - c. The enzymatic defect results in impaired synthesis of adrenal steroids beyond the enzymatic block and overproduction of the precursors before the block.
  - d. 21-hydroxylase deficiency accounts for 90% of cases.
  - e. Most common cause of ambiguous genitalia in females.

**FIGURE 10.2**

Biosynthetic Pathway for Steroid Hormones.

#### 6. Diagnosis of CAH on newborn screen:

- The test measures 17-OHP and is 2% specific, resulting in a 98% false-positive rate due to artificial elevations from prematurity, sickness, stress.<sup>27</sup>
- If 17-OHP is 40 to 100 ng/mL, repeat test.
- If 17-OHP is higher than 100 ng/mL, obtain electrolytes and serum 17-OHP. If evidence of hyperkalemia or hyponatremia, initiate treatment with hydrocortisone.
- In complete enzyme deficiency, adrenal crisis in untreated patients occurs at 1 to 2 weeks of age due to salt wasting.

#### 7. Diagnosis of CAH outside of newborn period:

- Suspect partial enzyme deficiency if evidence of androgen excess (premature adrenarche, hirsutism, irregular menses, acne, advanced bone age).
- Morning 17-OHP levels may be elevated.
- Diagnosis may require an ACTH stimulation test. A significant rise in the 17-OHP level 60 minutes after ACTH injection is diagnostic. Cortisol response may be decreased.

#### 8. Addison disease<sup>30</sup>:

- Primary AI due to autoimmune destruction of the adrenal glands.
- In children, it may be part of autoimmune polyendocrine syndrome type 1 (APS-1), which also includes hypoparathyroidism and chronic mucocutaneous candidiasis.
- Individuals with autoimmune Addison disease should also be screened for other endocrinopathies (T1DM, celiac disease, hypothyroidism, hypoparathyroidism).

**TABLE 10.13**  
**POTENCY OF VARIOUS THERAPEUTIC STEROIDS<sup>c</sup>**

Steroid	Glucocorticoid Effect <sup>a</sup> (in mg of Cortisol per mg of Steroid)	Mineralocorticoid Effect <sup>b</sup> (in mg of Cortisol per mg of Steroid)
Cortisol (hydrocortisone)	1	1
Cortisone acetate (oral)	0.8	0.8
Cortisone acetate (intramuscular)	0.8	0.8
Prednisone	4	0.25
Prednisolone	4	0.25
Methylprednisolone	5	0.4
$\beta$ -Methasone	25	0
Triamcinolone	5	0
Dexamethasone	30	0
9 $\alpha$ -Fluorocortisone (fludrocortisone)	15	200
Deoxycorticosterone acetate	0	20
Aldosterone	0.3	200–1,000

<sup>a</sup>To determine cortisol equivalent of a given steroid dose, multiply dose of steroid by corresponding number in column for glucocorticoid or mineralocorticoid effect. To determine dose of a given steroid based on desired cortisol dose, divide desired hydrocortisone dose by corresponding number in the column.

<sup>b</sup>Total physiologic replacement for salt retention is usually 0.1 mg fludrocortisone, regardless of patient size.

<sup>c</sup>Set relative to potency of cortisol.

Modified from Sperling MA. *Pediatric Endocrinology*. 3rd ed. Philadelphia: Elsevier; 2008:476.

**TABLE 10.14**  
**MAINTENANCE DOSING STEROIDS**

Adrenal Hormone	Dose
Glucocorticoid dosing	<ol style="list-style-type: none"> <li>PO hydrocortisone 6–18 mg/m<sup>2</sup>/day <math>\div</math> TID OR</li> <li>PO prednisone 1.5–3.5 mg/m<sup>2</sup>/day <math>\div</math> BID</li> </ol>
Mineralocorticoid dosing <sup>a</sup>	<ol style="list-style-type: none"> <li>PO fludrocortisone acetate 0.1 mg/m<sup>2</sup>/day OR</li> <li>If unable to take PO: IV hydrocortisone 50 mg/m<sup>2</sup>/day<sup>b</sup> PLUS</li> <li>Infants require an additional 1–2 g (17–34 mEq) of sodium supplementation daily</li> </ol>

<sup>a</sup>Required in salt losing forms of adrenal insufficiency.

<sup>b</sup>Synthetic steroids (e.g., prednisone, dexamethasone) do not have sufficient mineralocorticoid effect.

#### 9. Treatment of adrenal insufficiency:

- See Table 10.13 for relative steroid potencies.
- See Table 10.14 for maintenance glucocorticoid and mineralocorticoid dosing.
- Typically, lower doses are required for central AI, intermediate doses for primary AI, and higher doses for CAH.

**TABLE 10.15****STRESS DOSING STEROIDS**

Degree of Stress	Dose
Moderate Stress (minor illness, fever)	1. PO hydrocortisone 30–50 mg/m <sup>2</sup> /day ÷ TID OR 2. PO prednisone 6–10 mg/m <sup>2</sup> /day ÷ BID
Severe Stress (surgery, severe illness, compensated shock)	1. IV bolus of hydrocortisone 50 mg/m <sup>2</sup> then 50–100 mg/m <sup>2</sup> /day IV as continuous infusion or divided Q6 hr OR 2. IM injection of 25 mg/m <sup>2</sup> /dose Q6 hr

**BOX 10.2****RAPID APPROXIMATION OF STRESS DOSE STEROID REQUIREMENT****Infant:** 25 mg hydrocortisone**Small child:** 50 mg hydrocortisone**Large child/adolescent:** 100 mg hydrocortisone

10

**10. Stress dosing of steroids:**

- Hydrocortisone and cortisone are the only glucocorticoids that provide the necessary mineralocorticoid effects; prednisone and dexamethasone do not.
- See Table 10.15 for calculation of moderate and major stress dose steroid calculations.
- See Box 10.2 for rapid approximation of steroid dosing in the setting of acute adrenal crisis.

**11. Indications for stress dosing of steroids:**

- “Stress” is defined as systemic infection, febrile illness, diarrheal illness, trauma/fracture, burns, or surgery.
- Stress glucocorticoids should be given to patients:
  - With known primary or secondary AI
  - Following discontinuation of exogenous steroid (given for greater than 2 weeks at doses greater than physiologic replacement) until there is recovery of endogenous cortisol production (consider the need for 8am cortisol or ACTH stimulation test)
- Consider for hypotension refractory to fluid resuscitation in patients with suspicion for AI (even if not clinically diagnosed).

**B. Adrenal Cortex Hormone Excess<sup>29</sup>****1. Causes:**

- Hypercortisolism (Cushing syndrome):
  - Exogenous steroid use
  - Excess cortisol secretion from the adrenals
  - Excess ACTH production from ectopic ACTH producing tumor
  - Excess ACTH production from a pituitary tumor (known as Cushing disease)

- b. Hyperaldosteronism:
  - (1) Benign tumor of adrenal cortex (Conn syndrome)
  - (2) Overproduction by both adrenal glands (idiopathic hyperaldosteronism)
  - (3) Rarely glucocorticoid remediable aldosteronism
  - (4) Less common in children than hypercortisolism
  - (5) Lab findings include hypokalemia and hypernatremia

**2. Diagnosis of Cushing Syndrome<sup>31</sup>:**

- a. Step 1: Demonstrate hypercortisolism with two separate measurements. Multiple screening tests are available; specificity increases when they are used in combination:
  - (1) 24-hour urine cortisol ( $>90 \mu\text{g}/24 \text{ hour}$  consistent with hypercortisolism).
  - (2) Midnight salivary cortisol level ( $>0.13 \text{ mCg/dL}$  consistent with hypercortisolism).
  - (3) Overnight low dose dexamethasone suppression test: Give 1 mg dexamethasone at 11pm followed by an 8am serum cortisol the next morning (normal suppression  $<1.8 \text{ mCg/dL}$ ).
- b. Step 2: Determine etiology of hypercortisolism (ACTH-dependent vs. independent)
  - (1) Obtain plasma ACTH between 11pm–1am:  $>23 \text{ pg/mL}$  in a patient with hypercortisolism (as diagnosed above) indicates ACTH dependency (Cushing Disease vs. ectopic tumor).
  - (2) If cause is Cushing disease (ACTH-dependent), ACTH level will be  $>100x$  elevated.
  - (3) In ACTH-independent Cushing syndrome, level will be  $<5 \text{ pg/mL}$ .

**C. Adrenal Medulla Hormone Excess: Pheochromocytoma<sup>32-34</sup>**

**1. Clinical findings:**

- a. Extreme, sustained elevations in blood pressure (accounts for  $<1\%$  of pediatric hypertension).
- b. Associated with syndromes: multiple endocrine neoplasia (MEN) IIA and IIB, von Hippel-Lindau, neurofibromatosis (NF) 1, familial paraganglioma syndrome.

**2. Diagnosis:**

- a. Urine metanephrenes (see **Table EC 10.A** for age-specific normal values).
- b. Plasma metanephrenes (see **Table EC 10.B** for age-specific normal values).

**V. DISORDERS OF SODIUM AND WATER REGULATION<sup>35</sup>**

**A. Distinguishing Between Disorders of Sodium and Water Regulation:  
See **Table 10.16****

**B. Correction of Hypo- and Hypernatremia: See **Chapter 11**.**

**C. Conducting a Water Deprivation Test**

1. Begin test after a 24-hour period of adequate hydration and stable weight.

**TABLE EC 10.A****CATECHOLAMINES,<sup>a</sup> URINE**

Compound	3–8 Years	9–12 Years	13–17 Years	Adults
Dopamine (mCg/24 hr)	80–378	51–474	51–645	52–480
Epinephrine (mCg/24 hr)	1–7	≤8	≤11	2–14
Norepinephrine (mCg/24 hr)	5–41	5–50	12–88	15–100
Homovanillic acid (mg/24 hr)	0.5–6.7	1.1–6.8	1.4–7.2	1.6–7.5
Vanillylmandelic acid (g/24 hr)	≤2.3	≤3.4	≤3.9	≤6.0
	3 months–4 years	5–9 years	10–13 years	14–17 years
Metanephrines (mCg/24 hr)	25–117	11–139	51–275	40–189
Normetanephrines (mCg/24 hr)	54–249	31–398	67–503	69–531
				18–29 years
				25–222
				40–412

<sup>a</sup>Catecholamines are elevated in a variety of tumors, including neuroblastoma, ganglioneuroma, ganglioblastoma, and pheochromocytoma.

Data from JHH laboratories.

**TABLE EC 10.B****CATECHOLAMINES, PLASMA**

	Supine (pg/mL)	Sitting (pg/mL)
<b>EPINEPHRINE</b>		
3–15 years	≤464	Not determined
Adult	≤50	≤95
<b>NOREPINEPHRINE</b>		
3–15 years	≤1251	Not determined
Adult	112–658	217–1109
<b>DOPAMINE</b>		
3–15 years	≤60	Not determined
Adult	≤30	≤30

Data from Blondell R, Foster MB, Dave KC. Disorders of puberty. *Am Family Phys*. 1999;60:209–218; and JHH Laboratories.

**TABLE 10.16****DIFFERENTIATING BETWEEN DISORDERS OF SODIUM AND WATER REGULATION**

	SIADH	Cerebral Salt Wasting	DI
Serum Na <sup>+</sup>	<135 mEq/L	<135 mEq/L	>145 mEq/L <sup>a</sup>
Serum Osm	<280 mOsm/kg	<280 mOsm/kg	>300 mOsm/kg <sup>a</sup>
Urine Na <sup>+</sup>	>40 mEq/L	>40 mEq/L	< 40 mEq/L <sup>b</sup>
Urine Osm	>100 mOsm/kg (inappropriately concentrated)	>100 mOsm/kg (inappropriately concentrated)	<300 mOsm/kg (inappropriately dilute)
Volume Status	Euvolemia	Hypovolemia	Hypovolemia
Urine Output	Decreased	Increased	Increased
Other lab findings	High vasopressin	Low vasopressin	<ol style="list-style-type: none"> <li>1. Central: low vaso-pressin (&lt;0.5 pg/mL)</li> <li>2. Nephrogenic: high vasopressin</li> </ol>
Causes	Nausea, CNS and pulmonary pathology, surgery, certain medications	CNS disorders, hypersecretion of atrial natriuretic peptide	<ol style="list-style-type: none"> <li>1. Central: ADH secretion from posterior pituitary</li> <li>2. Nephrogenic: ADH resistance at the nephron collecting duct</li> </ol>
Treatment	<p>Fluid restriction and correction of underlying cause</p> <p>Treatment with sodium will cause diuresis</p>	<p>Replacement of urine volume with IV solutions ± salt replacement</p>	<ol style="list-style-type: none"> <li>1. Central: Intransal Desmopressin acetate (DDAVP)</li> <li>2. Nephrogenic: Access to free water, salt restriction, consider thiazide diuretics, indomethacin</li> </ol>

<sup>a</sup>Normal serum sodium and osmolality can be seen in compensated diabetes insipidus, and water deprivation test should be performed if clinical suspicion is high.

<sup>b</sup>Urine sodium generally low in diabetes insipidus, however this depends on solute intake.

ADH, Antidiuretic hormone; CNS, central nervous system; DI, diabetes insipidus; Na<sup>+</sup>, sodium; Osm, osmolarity; SIADH, syndrome of inappropriate ADH secretion; IV, intravenous.

2. Obtain a baseline weight after bladder emptying, as well as baseline urine and blood osmolality and electrolytes.
3. Restrict fluids (max 7 hours, 4 hours for infants).
4. Measure body weight and urine-specific gravity and volume hourly.
5. If urine specific gravity  $\geq 1.014$ , or weight loss approaching 5%, terminate test and obtain urine and blood for osmolality and electrolytes.

#### D. Interpretation of Water Deprivation Test Results: See Table 10.17

#### E. Differentiating Between Central Versus Nephrogenic Causes of Diabetes Insipidus

1. Administer vasopressin subcutaneously at end of water deprivation test. Assess urine output, urine specific gravity, and water intake.
2. See Table 10.18 for interpretation of vasopressin test.

**TABLE 10.17****RESULTS OF WATER DEPRIVATION TEST IN NORMAL VERSUS CENTRAL/NEPHROGENIC DIABETES INSIPIDUS**

	Normal (Psychogenic Polydipsia)	Central/Nephrogenic DI
Urine volume	Decreased	No change
Weight loss	No change	$\leq 5\%$
Urine osmolality (mOsm/L)	500–1400 ( $>1000$ generally excludes diagnosis of DI)	$<150$
Plasma osmolality (mOsm/L)	288–291	$>290$
Urine specific gravity	$\geq 1.010$	$<1.005$
Urine: plasma osmolality ratio	$>2$	$<2$

DI, Diabetes insipidus.

**TABLE 10.18****RESULTS OF VASOPRESSIN ADMINISTRATION IN EVALUATION OF DIABETES INSIPIDUS**

	Psychogenic Polydipsia	Central <sup>a</sup>	Nephrogenic
Urine volume	↓	↓	No change
Urine specific gravity	$\geq 1.010$	$\geq 1.010$	No change
Oral fluid intake	No change	↓	No change

<sup>a</sup>In central diabetes insipidus, urine osmolality increases by 200% or more in response to vasopressin administration.

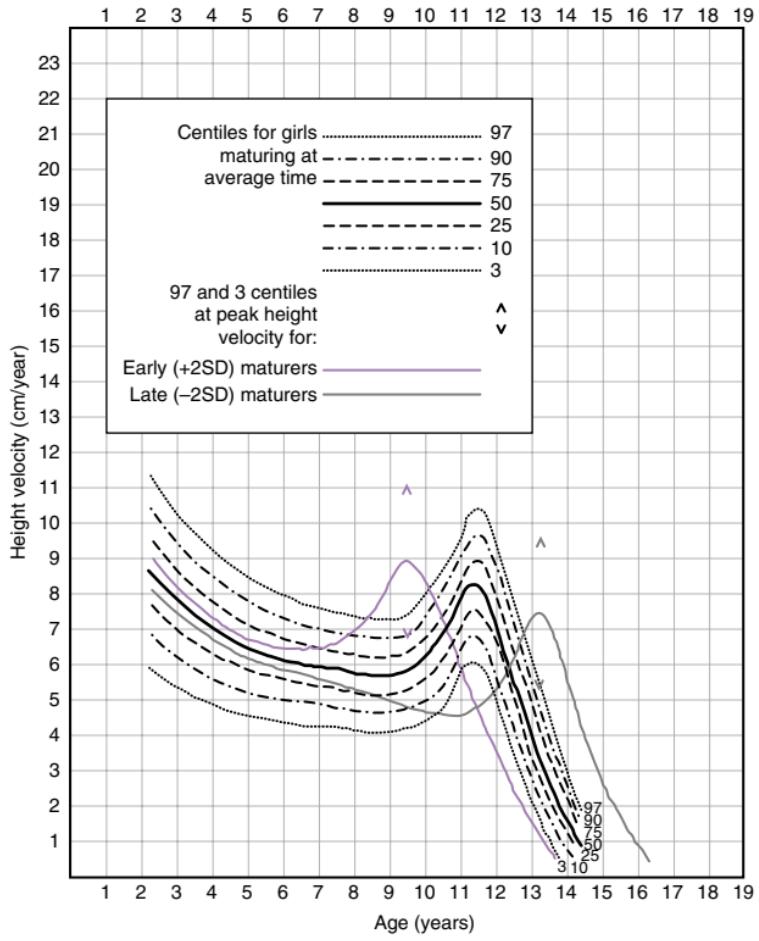
**TABLE 10.19****ESTIMATED GROWTH VELOCITY IN CHILDREN BASED ON AGE**

Age	Growth
Birth to 1 year old	25 cm/year
1 year old to 4 years old	10 cm/year
4 years old to 8 years old	5 cm/year
8 years old to 12 years old	5 cm/year <sup>a</sup>

<sup>a</sup>Rates may be considerably higher at later end of this age range when individuals have entered their pubertal growth spurt.

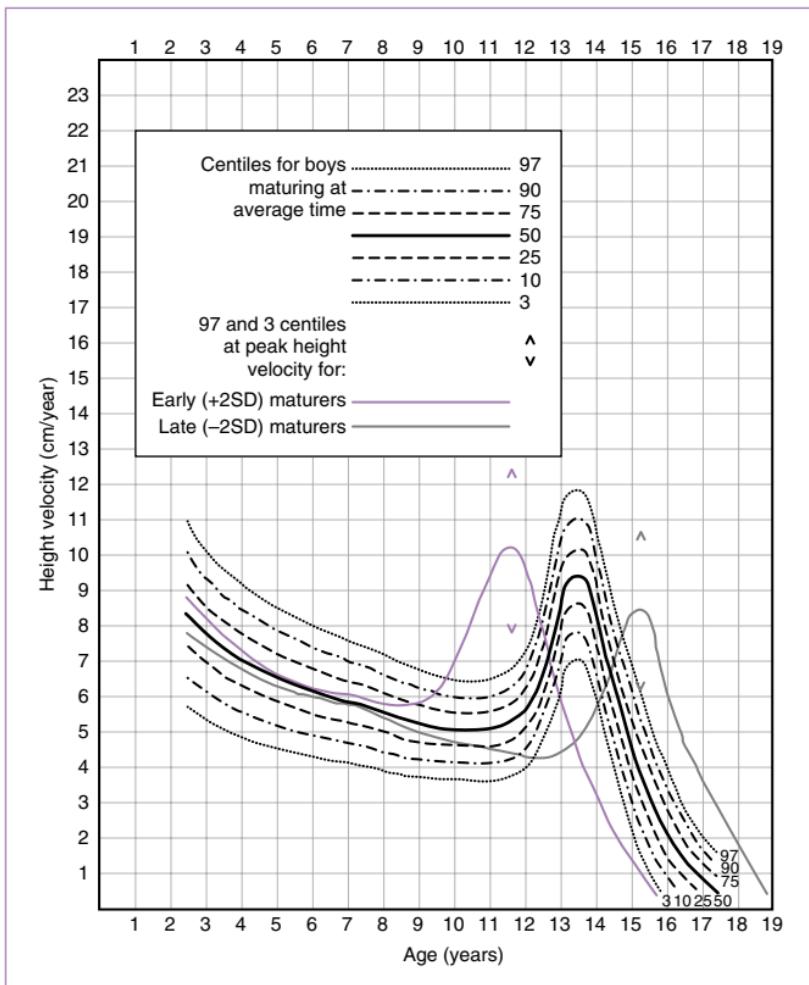
**VI. GROWTH<sup>35-37</sup>****A. Assessing Height**

- Mid-parental height and target height range:**
  - Mid-parental height for boys: (Paternal height + maternal height + 5 in or 13 cm)/2
  - Mid-parental height for girls: (Paternal height + maternal height – 5 in or 13 cm)/2
  - Target height range: Mid-parental height  $\pm 2$  SD (1 SD = 2 in or 5 cm)
- Determining average growth velocity:** See Table 10.19.
- See Figs. EC 10.A and EC 10.B for normal growth velocity curves for American females and males, respectively.



**FIGURE EC 10.A**

Height Velocity by Age for American Girls. (From Kelly A, Winer KK, Kalkwarf H, et al. Age-based references ranges for annual height velocity in US children. *J Clin Endocrinol Metab*. 2014;99:2104.)



**FIGURE EC 10.B**

Height Velocity by Age for American Boys. (From Kelly A, Winer KK, Kalkwarf H, et al. Age-based references ranges for annual height velocity in US children. *J Clin Endocrinol Metab*. 2014;99:2104.)

**TABLE 10.20****PATHOLOGIC VERSUS NONPATHOLGIC CAUSES OF SHORT STATURE**

	Familial Short Stature	Constitutional Growth Delay	Pathologic Causes (endocrine, genetic, etc.)
Growth velocity	Normal	Normal	Decreased
Onset of puberty	Normal	Delayed	Depends on cause
Family history	Short stature	Delayed puberty	+/-
Bone age	Normal	Delayed	Usually delayed (may be normal in genetic causes)
Eventual adult height	Short, near mid-parental height	Normal	Depends on cause

**B. Short Stature****1. Definition:**

- Short stature is height <2 SD below mean or <3<sup>rd</sup> percentile for age and sex.
- Growth failure is defined as height <2 SD below mid-parental height or height velocity <10<sup>th</sup> percentile for age resulting in a downward trend crossing height percentiles.
- Majority of children with short stature are healthy; true growth failure is typically pathologic and requires evaluation.

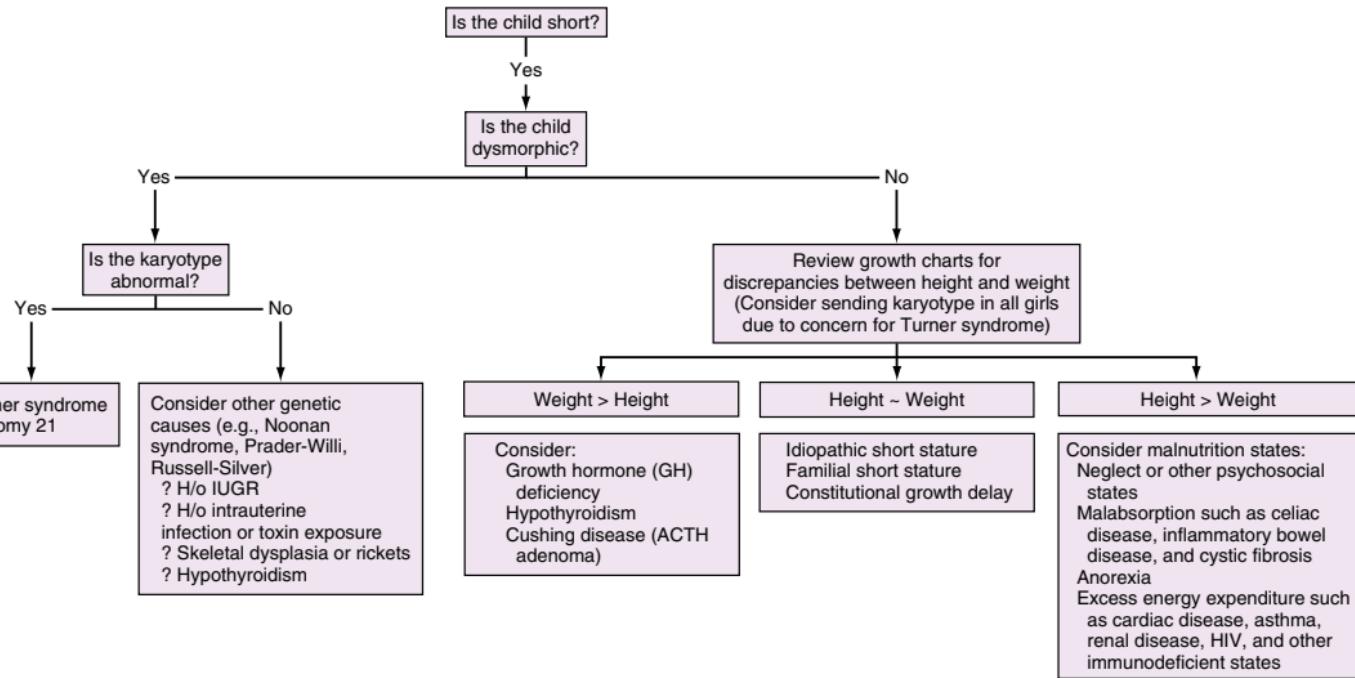
**2. Determining etiology:**

- See [Table 10.20](#) for approach to differentiating between pathologic and non-pathologic causes of short stature.
- Bone age is determined by radiographs of left wrist and hand.
- See [Fig. 10.3](#) for initial work up.
- A more extensive work-up can be guided by history and physical exam and could include:
  - TTG and IgA (celiac disease)
  - CBC with differential (anemia, malignancy, inflammation)
  - CRP/ESR (inflammation, infection)
  - CMP (renal/liver disorders, malnutrition, calcium disorders)
  - TSH, free T4 (hypothyroidism)
  - Karyotype or targeted gene testing (Turner syndrome, SHOX mutation)
  - IGF1, IGFBP-3 [proxy measurements for growth hormone (GH)]; IGFBP-3 has a higher specificity in children <10]; see [Table 10.21](#) and [Table EC 10.C](#) for normal values of IGF-1 and IGFBP-3, respectively

**3. Indications for growth hormone use<sup>38</sup>:**

The FDA has approved growth hormone for:

- Growth hormone deficiency
- Children born small-for-gestational-age (SGA) who between 2 and 4 years of age have shown inadequate catch-up growth or evidence of normal growth velocity with height < 2.5 SD below mean

**FIGURE 10.3**

Differential Diagnosis of Short Stature.

**TABLE 10.21****INSULIN-LIKE GROWTH FACTOR 1<sup>a</sup>**

Age (Years)	Male (ng/mL)	Females (ng/mL)
<1	≤142	≤185
1–1.9	≤134	≤175
2–2.9	≤135	≤178
3–3.9	30–155	38–214
4–4.9	28–181	34–238
5–5.9	31–214	37–272
6–6.9	38–253	45–316
7–7.9	48–298	58–367
8–8.9	62–347	76–424
9–9.9	80–398	99–483
10–10.9	100–449	125–541
11–11.9	123–497	152–593
12–12.9	146–541	178–636
13–13.9	168–576	200–664
14–14.9	187–599	214–673
15–15.9	201–609	218–659
16–16.9	209–602	208–619
17–17.9	207–576	185–551

<sup>a</sup>A clearly normal IGF-1 level argues against growth hormone (GH) deficiency, except in young children in whom there is considerable overlap between normal levels and those with GH deficiency.

Reference ranges from Quest Diagnostics LC/MS (liquid chromatography/tandem mass spectrometry) assay.

- c. Chronic kidney disease
- d. Turner syndrome, Noonan syndrome, Prader-Willi syndrome
- e. Short stature homeobox containing gene (SHOX) deficiency
- f. Children with idiopathic short stature (height <2.25 SD below mean and unlikely to attain normal adult height)

**VII. SEXUAL DEVELOPMENT<sup>39-45</sup>****A. Puberty**

- For normal pubertal stages, please see [Chapter 5](#).
- For definitions of precocious and delayed puberty, see [Table 10.22](#).

**B. Lab Evaluation**

- LH, FSH, estradiol, and testosterone (free and total), see [Tables 10.23–10.27](#) for normal values. **NOTE:** Early in puberty, LH production peaks overnight and is lower during the day, so consider obtaining levels in the early morning.
- GnRH stimulation test<sup>46</sup>:
  - Purpose: To evaluate for biochemical evidence of puberty when LH, FSH, and sex hormone testing is inconclusive.
  - Method: Give GnRH analog (Leuprolide) SQ, and measure LH and FSH levels at 0 and 60 minutes.

**TABLE EC 10.C****INSULIN-LIKE GROWTH FACTOR-BINDING PROTEIN (IGF-BP3)<sup>a</sup>**

Age	mg/L	Tanner Stage	Female (mg/L)	Male (mg/L)
0–7 days	≤0.7	Tanner I	1.2–6.4	1.4–5.2
8–15 days	0.5–1.4	Tanner II	2.8–6.9	2.3–6.3
16 days–1 years	0.7–3.6	Tanner III	3.9–9.4	3.2–8.9
2 years	0.8–3.9	Tanner IV	3.3–8.1	3.7–8.7
3 years	0.9–4.3	Tanner V	2.7–9.1	2.6–8.6
4 years	1.0–4.7			
5 years	1.1–5.2			
6 years	1.3–5.6			
7 years	1.4–6.1			
8 years	1.6–6.5			
9 years	1.8–7.1			
10 years	2.1–7.7			
11 years	2.4–8.4			
12 years	2.7–8.9			
13 years	3.1–9.5			
14 years	3.3–10.0			
15 years	3.5–10.0			
16 years	3.4–9.5			
17 years	3.2–8.7			
18 years	3.1–7.9			
19 years	2.9–7.3			

Adults continue to vary  
by age

<sup>a</sup>Levels below the 5th percentile suggest growth hormone deficiency. This test may have greater discrimination than the IGF-1 test in younger patients.

Data from Quest Diagnostics immunochemiluminometric assay (ICMA).

**TABLE 10.22****DEFINITIONS OF PRECOCIOUS AND DELAYED SEXUAL MATURATION**

	<b>Females</b>	<b>Males</b>
Precocious	Before age 8 years: Thelarche (may be benign or progressive as seen in precocious puberty) Adrenarche (may be isolated or a feature of precocious puberty)	Before age 9 years: Testicular enlargement Adrenarche (may be isolated or a feature of precocious puberty)
Delayed	No thelarche by 13 years or >5 years between thelarche and menarche. Primary amenorrhea: no menarche by age 16 years in the presence of secondary sexual characteristics, or no menarche and no secondary sexual characteristics by age 14 years.	No testicular enlargement by 14 years.

**TABLE 10.23****LUTEINIZING HORMONE**

<b>Age</b>	<b>Males (mIU/mL)</b>	<b>Females (mIU/mL)</b>
0–2 years	Not established	Not established
3–7 years	≤0.26	≤0.26
8–9 years	≤0.46	≤0.69
10–11 years	≤3.13	≤4.38
12–14 years	0.23–4.41	0.04–10.80
15–17 years	0.29–4.77	0.97–14.70
<b>Tanner Stages</b>	<b>Males (mIU/mL)</b>	<b>Females (mIU/mL)</b>
I	≤0.52	≤0.15
II	≤1.76	≤2.91
III	≤4.06	≤7.01
IV–V	0.06–4.77	0.10–14.70

Data from Quest Diagnostics immunoassay. For more information, see [www.questdiagnostics.com](http://www.questdiagnostics.com).

**TABLE 10.24****FOLLICLE-STIMULATING HORMONE**

<b>Age</b>	<b>Male (mIU/mL)</b>	<b>Female (mIU/mL)</b>
0–4 years	Not established	Not established
5–9 years	0.21–4.33	0.72–5.33
10–13 years	0.53–4.92	0.87–9.16
14–17 years	0.85–8.74	0.64–10.98

Data from Quest Diagnostics immunoassay. For more information, see [www.questdiagnostics.com](http://www.questdiagnostics.com).

**TABLE 10.25****ESTRADIOL<sup>a</sup>**

Age	Level (pg/mL)
Prepubertal children	<25
Men	6–44
Women	
Luteal phase	26–165
Follicular phase	None detected–266
Midcycle	118–355
Adult women on OCP	None detected–102

<sup>a</sup>Normal infants have elevated estradiol at birth, which decreases to prepubertal values during the first week of life. Estradiol levels increase again between age 1 and 2 months and return to pre-pubertal values by age 6–12 months.

Data from JHH Laboratories.

OCP, Oral contraceptive pill.

**TABLE 10.26****TESTOSTERONE, TOTAL SERUM<sup>a</sup>**

Age	Male (ng/dL)	Female (ng/dL)
Cord blood	17–61	16–44
1–10 days	≤187	≤24
1–3 months	72–344	≤17
3–5 months	≤201	≤12
5–7 months	≤59	≤13
7–12 months	≤16	≤11
1–5.9 years	≤5	≤8
6–7.9 years	≤25	≤20
8–10.9 years	≤42	≤35
11–11.9 years	≤260	≤40
12–13.9 years	≤420	≤40
14–17.9 years	≤1000	≤40
≥18 (adult)	250–1100	2–45
TANNER STAGE		
Stage I	≤5	≤8
Stage II	≤167	≤24
Stage III	21–719	≤28
Stage IV	25–912	≤31
Stage V	110–975	≤33

<sup>a</sup>Normal testosterone/dihydrotestosterone (T/DHT) ratio is <18 in adults and older children and <10 in neonates. A T/DHT ratio >20 suggests 5-α-reductase deficiency or androgen insensitivity syndrome.

Data from Quest Diagnostics LC/MS (liquid chromatography/tandem mass spectrometry) assay.

**TABLE 10.27****TESTOSTERONE, FREE**

Age	Male (pg/mL)	Female (pg/mL)
1–11 years	≤1.3	≤1.5
12–13 years	≤64.0	≤1.5
14–17 years	4.0–100.0	≤3.6
18–69 years	46.0–224.0	0.2–5.0

Data from Quest Diagnostics LC/MS (liquid chromatography/tandem mass spectrometry) assay.

- c. Interpretation: Prepubertal children should show no or minimal increase in LH and FSH in response to GnRH. A rise of LH to  $>3.3$  to  $5.0$  IU/L is evidence of central puberty.
- 3. **Delayed puberty**<sup>41,45,47</sup>: See Fig. 10.4 for information on evaluation and management of delayed puberty.
- 4. **Precocious puberty**<sup>42,47</sup>: See Fig. 10.5 for information on evaluation and management of precocious puberty.

### C. Polycystic Ovarian Syndrome<sup>48</sup>

#### 1. Clinical features in adolescents:

- a. Diagnostic criteria (must have features of both):
  - (1) Hyperandrogenism: Either clinical or biochemical
    - (a) Clinical: Hirsutism, acne, male pattern alopecia
    - (b) Biochemical characteristics: Elevated androgens including DHEA-S (see Table 10.28 for normal values), free or total testosterone
  - (2) Menstrual abnormalities: Amenorrhea or oligomenorrhea (chronic anovulation).

**NOTE:** Appearance of multiple ovarian cysts is a diagnostic criterion for adults, but not for adolescents, as this can be a normal finding in adolescent females.

- b. Common cause of female infertility.
- c. Often LH>FSH, but this is not required for diagnosis.
- d. Chronic anovulation and unopposed estrogen exposure increase risk for endometrial cancer.
- e. Associated with insulin resistance and increased risk of type 2 diabetes.

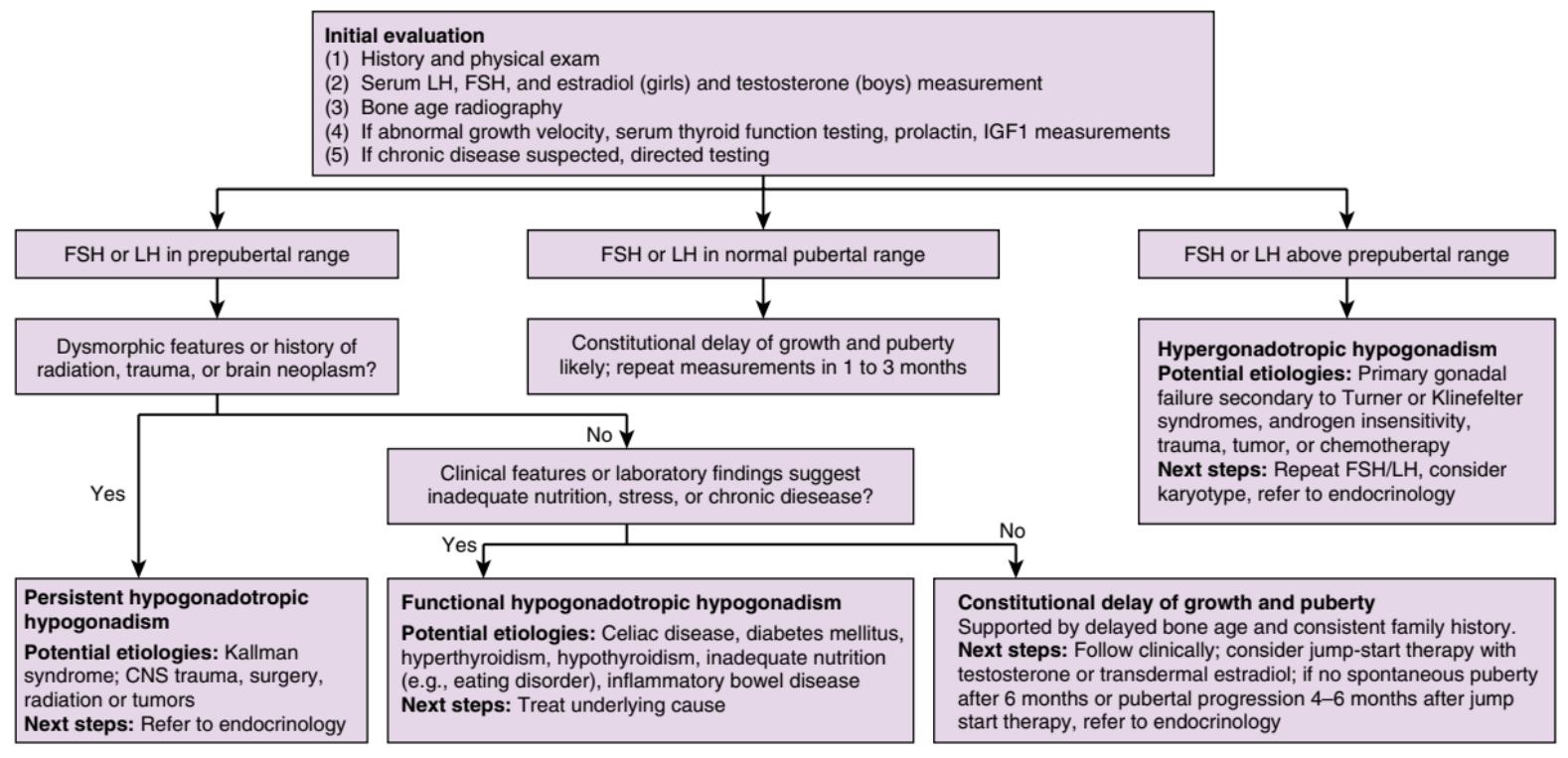
#### 2. Management:

- a. Combined oral contraceptives: First-line for management of menstrual abnormalities and hirsutism/acne. Increases SHBG (thus decreasing free testosterone), which may result in increased insulin sensitivity and restoration of ovulation.
- b. Anti-androgen therapy, such as spironolactone, to treat hirsutism.
- c. Weight reduction and other lifestyle changes.
- d. Metformin: Can be considered as possible treatment if goal is to treat insulin resistance.

### D. Ambiguous Genitalia<sup>49</sup>

#### 1. Clinical findings in a neonate suspicious for ambiguous genitalia:

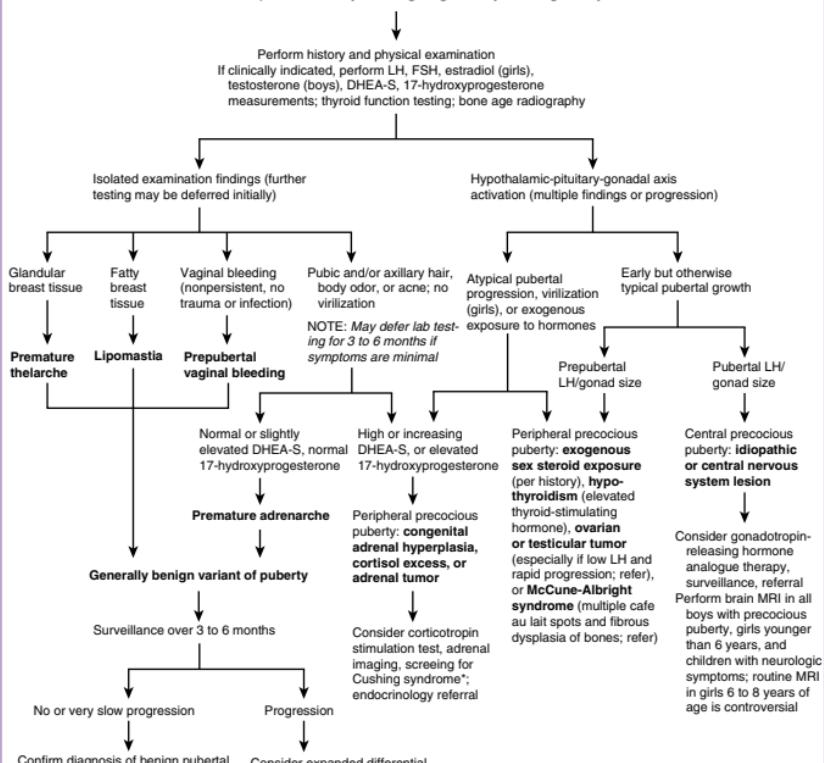
- a. Apparent female with clitoromegaly (length  $>1$  cm or width  $>6$  mm in term infant), inguinal or labial mass, or posterior labial fusion.
- b. Micropenis (stretched penile length that is  $-2.5$  SD below mean for age, see Table 10.29 for normal values).
- c. Non-palpable gonads in an apparent male.
- d. Hypospadias associated with separation of scrotal sacs or undescended testis.
- e. Discordance between prenatal karyotype and genital appearance.

**FIGURE 10.4**

An Approach to the Child Presenting With Delayed Puberty. CNS, Central nervous system; FSH, follicle-stimulating hormone; IGF1, insulin-like growth factor 1; LH, luteinizing hormone; MRI, magnetic resonance imaging. (From Klein DA, Emerick JE, Sylvester JE, Vogt KS. Disorders of puberty. An approach to diagnosis and management. *Am Fam Physician*. 2017;96(9):590–599.)

**Diagnostic approach to early pubertal development**

Pubertal development before 8 years of age in girls or 9 years of age in boys



\*24-hour urine free cortisol or midnight salivary cortisol measurement.

**FIGURE 10.5**

An Approach to the Child Presenting With Early Puberty. *DHEA-S*, Dehydroepiandrosterone sulfate; *FSH*, follicle-stimulating hormone; *LH*, luteinizing hormone; *MRI*, magnetic resonance imaging. (Reprinted with permission from Disorders of Puberty: An Approach to Diagnosis and Management, November 1, 2017, Vol 96, No 9, American Family Physician Copyright © 2017 American Academy of Family Physicians. All Rights Reserved.)

**TABLE 10.28****DEHYDROEPIANDROSTERONE SULFATE (DHEA-S)**

Age	Male (mCg/dL)	Female (mCg/dL)
<1 months	≤316	15–261
1–6 months	≤58	≤74
7–11 months	≤26	≤26
1–3 years	≤15	≤22
4–6 years	≤27	≤34
7–9 years	≤91	≤92
10–13 years	≤138	≤148
14–17 years	38–340	37–307

*continued*

**TABLE 10.28****DEHYDROPIANDROSTERONE SULFATE (DHEA-S)—CONT'D**

Age	Male (mCg/dL)	Female (mCg/dL)
<b>TANNER STAGES (AGES 7–17)</b>		
I	≤49	≤46
II	≤81	15–133
III	22–126	42–126
IV	33–177	42–241
V	110–370	45–320

Data from Quest Diagnostics assay. For more information see [www.questdiagnostics.com](http://www.questdiagnostics.com).

**TABLE 10.29****MEAN STRETCHED PENILE LENGTH (CM)<sup>a</sup>**

Age	Mean ± SD	-2.5 SD
<b>BIRTH</b>		
30 week gestation	2.5 ± 0.4	1.5
34 week gestation	3.0 ± 0.4	2.0
Full term	3.5 ± 0.4	2.5
0–5 months	3.9 ± 0.8	1.9
6–12 months	4.3 ± 0.8	2.3
1–2 years	4.7 ± 0.8	2.6
2–3 years	5.1 ± 0.9	2.9
3–4 years	5.5 ± 0.9	3.3
4–5 years	5.7 ± 0.9	3.5
5–6 years	6.0 ± 0.9	3.8
6–7 years	6.1 ± 0.9	3.9
7–8 years	6.2 ± 1.0	3.7
8–9 years	6.3 ± 1.0	3.8
9–10 years	6.3 ± 1.0	3.8
10–11 years	6.4 ± 1.1	3.7
Adult	13.3 ± 1.6	9.3

<sup>a</sup>Measured from the pubic ramus to the tip of the glans while traction is applied along the length of the phallus to the point of increased resistance.

SD, standard deviation.

Data from Feldman KW, Smith DW. Fetal phallic growth and penile standards for newborn male infants. *J Pediatr*. 1975;86:395.

## 2. Etiology:

- Due to undervirilization of male genitalia or virilization of female genitalia
- Most common cause is CAH
- Other causes by male versus female karyotype:
  - 46,XY karyotype: Testicular regression, androgen insensitivity, testosterone biosynthesis disorders, rare forms of CAH, absence of SRY
  - 46,XX karyotype: SRY+, classical (21-hydroxylase deficiency) or more rare forms of CAH
  - Other: Sex chromosome mosaicism (46,XY/46,XX, 46,XY/45,XO, etc.)

**3. Evaluation:**

- a. Labs: Timing of collection is important.
  - (1) Initial testing: LH, FSH, testosterone, dihydrotestosterone (DHT, see [Table EC 10.D](#)), anti-Müllerian hormone (AMH) and expedited determination of sex chromosomes (ask that resulting lab rush results of sex chromosomes)
  - (2) After 36 hours of life: 17-hydroxyprogesterone
  - (3) Daily electrolytes until salt-wasting CAH is ruled out
  - (4) Further testing as needed to evaluate for more rare forms of CAH: DHEA, 17-hydroxypregnolone, 11-deoxycortisol, cortisol, ACTH
- b. Imaging: Options include genitogram (contrast study of the urogenital sinus and internal duct structures) or voiding cysto-urethrogram (VCUG), pelvic and abdominal US, and pelvic magnetic resonance imaging (MRI) to evaluate internal anatomy.
- c. Care should be taken to avoid premature gender/sex designation, completion of birth certificate, and naming of infant.

**E. Cryptorchidism<sup>50</sup>****1. Epidemiology and clinical course:**

- a. Can be present at birth (congenital) or after birth (acquired). Congenital rate is 1% to 4.6% of males born >2.5 kg.
- b. Increased risk with preterm birth or low birthweight.
- c. About 1/3 to 1/2 of cryptorchid testicles descend spontaneously, usually by age 3 months.
- d. Neoplasm more common in males with cryptorchidism and may occur in contralateral testis; early orchidopexy decreases risk of malignancy.
- e. Males with bilateral cryptorchidism have higher risk for reduced fertility.
- f. There is a higher risk of testicular torsion prior to repair.

**2. Evaluation:**

- a. Providers should palpate testes for quality and position in all males at each well child visit.
  - b. Any phenotypic male newborn with bilateral, *nonpalpable* testes should undergo evaluation for CAH with karyotype and hormonal profile.
  - c. In those without CAH, distinguish between cryptorchidism and anorchia (absent testes) with serum Müllerian inhibiting substance and consider additional hormone testing (inhibin B, FSH, LH, and testosterone).
3. **Treatment:** Observe for 6 months, at which time if testis remains undescended, referral to specialist recommended. Orchidopexy between 6 and 18 months of age recommended.

**TABLE EC 10.D****DIHYDROTESTOSTERONE**

<b>Age</b>	<b>Males (ng/dL)</b>	<b>Females (ng/dL)</b>
Cord blood	<2–8	<2–5
1–6 months	12–85	<5
Prepubertal	<5	<5
Tanner stage II–III	3–33	5–19
Tanner stage IV–V	22–75	3–30

Data from Quest Diagnostics RIA (radioimmunoassay).

**TABLE EC 10.E****ANDROSTENEDIONE, SERUM**

<b>Age</b>	<b>Males (ng/dL)</b>	<b>Females (ng/dL)</b>
Premature (31–35 weeks)	≤480	≤480
Full-term infants	≤290	≤290
1–12 months	6–78	6–78
1–4 years	5–51	5–51
5–9 years	6–115	6–115
10–13 years	12–221	12–221
14–17 years	22–225	22–225
Tanner stage II–III	17–82	43–180
Tanner stage IV–V	57–150	73–220
Adult male (18–30 years)	50–220	
Female follicular phase		35–250
Female luteal phase		30–235

Data from Quest Diagnostics LC/MS (liquid chromatography/tandem mass spectrometry) analysis.

## VIII. NEONATAL HYPOGLYCEMIA EVALUATION<sup>51,52</sup>

### A. Definition

1. Serum glucose level insufficient to meet metabolic requirements. For practical purposes, value is defined as a point-of-care glucose (POCG) <45 to 50 mg/dL within first 48 hours of life and <70 mg/dL beyond this period.

**NOTE:** Bedside glucometer is a convenient tool to screen for hypoglycemia but can be inaccurate by 10 to 15 mg/dL when in the range of hypoglycemia. STAT plasma glucose must be sent to establish diagnosis of hypoglycemia.

### B. Treatment Goals

1. For neonates with suspected congenital hypoglycemia disorder and infants/children with confirmed hypoglycemia disorder, maintain plasma glucose >70 mg/dL.
2. For high risk neonates without a suspected congenital hypoglycemia disorder, maintain plasma glucose >45 to 50 mg/dL for those <48 hours of age and >60 mg/dL for those aged >48 hours.

### C. Management

See Chapter 18.

10

### D. Further Work-up

1. If serum glucose is consistently <70 mg/dL after 48 hours of life, at the time of hypoglycemia (serum glucose <45 to 50 mg/dL via glucometer), obtain STAT serum glucose, insulin, growth hormone, cortisol, free fatty acids, and  $\beta$ -hydroxybutyrate.
2. Consider **glucagon stimulation test**: Administer glucagon and obtain serum glucose levels Q10 min  $\times 4$ . Repeat growth hormone and cortisol levels 30 minutes after documented hypoglycemia.

### E. Interpretation of Results

1. A rise in glucose  $\geq 30$  mg/dL on glucagon stimulation test, along with elevated plasma insulin levels  $>2 \mu\text{U/mL}$  (absence of detectable insulin does not rule out hyperinsulinism, as insulin may be present below the lower limit of detection of the assay), low serum levels of free fatty acids ( $<1.5 \text{ mmol/L}$ ) and  $\beta$ -hydroxybutyrate ( $<2 \text{ mmol/L}$ ), and a persistent glucose requirement  $>8 \text{ mg/kg/min}$  suggests a diagnosis of hyperinsulinemia.
2. Hypoglycemia with midline defects and micropenis in a male suggest hypopituitarism, supported by low serum levels of growth hormone and cortisol at the time of hypoglycemia.

### F. Hyperinsulinemia

1. Hyperinsulinemia is the most common cause of neonatal hypoglycemia beyond the first 7 days of life and may be congenital or transient.
2. Congenital hyperinsulinism can be caused by dominant or recessive mutations in genes responsible for regulating insulin secretion from pancreatic  $\beta$  cells.

3. Transient hyperinsulinemia is commonly seen in infants of diabetic mothers and less commonly in the setting of perinatal asphyxia and intrauterine growth restriction.
4. Long-term management of persistent hyperinsulinism includes diazoxide, which inhibits pancreatic secretion of insulin by keeping  $\beta$ -cell ATP-sensitive potassium channels open; however, it has been rarely associated with pulmonary hypertension (black box warning<sup>53</sup>).

## IX. ADDITIONAL NORMAL VALUES

Normal values may differ among laboratories because of variation in technique and type of assay used.

See the following tables for normal values:

Table EC 10.A, Catecholamines, urine

Table EC 10.B, Catecholamines, plasma

Table EC 10.C, Insulin-like growth factor binding protein

Table EC 10.D, DHT

Table EC 10.E, Androstenedione, serum

## X. WEB RESOURCES

- A. Children with Diabetes ([www.childrenwithdiabetes.com](http://www.childrenwithdiabetes.com))
- B. American Diabetes Association ([www.diabetes.org](http://www.diabetes.org))
- C. International Society for Pediatric and Adolescent Diabetes ([www.ispad.org](http://www.ispad.org))
- D. Pediatric Endocrine Society ([www.lwpes.org](http://www.lwpes.org))
- E. The Endocrine Society ([www.endocrine.org](http://www.endocrine.org))
- F. American Thyroid Association ([www.thyroid.org](http://www.thyroid.org))

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A complete list of references can be found online at [www.expertconsult.com](http://www.expertconsult.com).

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# Chapter 11

## Fluids and Electrolytes

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 See additional content on Expert Consult

### I. INTRODUCTION

Intravenous fluids (IVFs) should be thought of as a medication by those who prescribe them. Since the late 1950s, IVF choice has been largely guided by Holliday and Segar's estimations of sodium requirements. Using the electrolyte composition of human milk, they calculated that the average child requires 3 mEq sodium (Na) and 2 mEq potassium (K) per 100 to 120 mL water ( $H_2O$ ).<sup>1</sup> According to their calculation, basic solute needs can be met by administering  $\frac{1}{4}$  normal saline (NS), a hypotonic fluid. While this estimation led to a long-standing tradition in pediatric maintenance IVF (MIVF) therapy, evidence published over the past few decades culminated in new American Academy of Pediatrics (AAP) guidelines recommending isotonic fluids as the maintenance fluid of choice for the majority of hospitalized children.<sup>2</sup>

### II. FLUID RESUSCITATION

#### A. Calculating Maintenance Fluid Volume

1. The Holliday-Segar method (Table 11.1 and Box 11.1) is the most widely used method to approximate maintenance fluid volume. This method estimates caloric expenditure in fixed-weight categories and assumes the average patient will require 100 mL of water for each 100 calories metabolized, with approximately 100 kcal burned per kg.<sup>1</sup>
2. NOTE: The Holliday-Segar method is not suitable for neonates <14 days old, because it generally overestimates fluid needs in neonates. (See Chapter 18 for neonatal fluid management.)

#### B. Calculating Fluid Loss

1. Total body water (TBW) is equal to **60% of a child's weight in kg (75% in infants)**.<sup>3</sup>

$$\text{EQUATION 11.1: } \text{TBW}^a = \text{weight (kg)} \times 0.6$$

<sup>a</sup>TBW uses preillness weight; 1 L water = 1 kg water

2. In a euvolemic child, 60% of TBW resides in the intracellular compartment [where potassium (K) concentration is 140 mEq/L and sodium (Na) is negligible], and 40% of TBW is in the extracellular compartment (where Na concentration is ~140 mEq/L and K is negligible).<sup>4-6</sup>
3. The most precise method of assessing fluid deficit uses weight loss:

$$\text{EQUATION 11.2: } \text{Fluid deficit (L)} = \text{preillness weight (kg)} - \text{illness weight (kg)}$$

**TABLE 11.1**  
**HOLLIDAY-SEGAR METHOD**

Body Weight	Fluid Volume	
	mL/kg/day	mL/kg/hr
First 10 kg	100	≈4
Second 10 kg	50	≈2
Each additional kg	20	≈1

**BOX 11.1****HOLLIDAY-SEGAR METHOD**

Example: Determine the correct fluid rate for an 8-year-old child weighing 25 kg:

$$\text{First 10 kg: } 4 \text{ mL/kg/hr} \times 10 \text{ kg} = 40 \text{ mL/hr} \quad 100 \text{ mL/kg/day} \times 10 \text{ kg} = 1000 \text{ mL/day}$$

$$\text{Second 10 kg: } 2 \text{ mL/kg/hr} \times 10 \text{ kg} = 20 \text{ mL/hr} \quad 50 \text{ mL/kg/day} \times 10 \text{ kg} = 500 \text{ mL/day}$$

$$\text{Each additional 1 kg: } 1 \text{ mL/kg/hr} \times 5 \text{ kg} = 5 \text{ mL/hr} \quad 20 \text{ mL/kg/day} \times 5 \text{ kg} = 100 \text{ mL/day}$$

$$\text{Answer: } 65 \text{ mL/hr} \quad \text{Answer: } 1600 \text{ mL/day}$$

**TABLE 11.2****CLINICAL OBSERVATIONS IN DEHYDRATION<sup>7</sup>**

	Older Child		
	3% (30 mL/kg)	6% (60 mL/kg)	9% (90 mL/kg)
	Infant		
	5% (50 mL/kg)	10% (100 mL/kg)	15% (150 mL/kg)
Dehydration Classification	Mild	Moderate	Severe
Mental status	Alert		Lethargic/obtunded
Fontanelle	Flat	Soft	Sunken
Eyes	Normal	Deep set	Sunken
Tears	Present	Reduced	None
Buccal mucosa/lips	Dry	Dry	Parched/cracked
Pulse rate	Normal	Slightly increased	Increased
Skin (touch)	Normal	Dry	Clammy
Skin turgor	Normal	Tenting	None
Capillary refill	Normal	≈2–3 seconds	>3 seconds
Pulse quality	Normal	Weak	Feeble/impalpable
Urine output	Normal/mild oliguria	Mild oliguria	Severe oliguria

4. Clinical assessment: If weight loss is not known, clinical observation may be used to approximate the percentage of dehydration (Table 11.2).<sup>7,8</sup>

**EQUATION 11.3:** % Dehydration =  $\frac{\text{fluid deficit}^a}{\text{preillness weight}} \times 100\%$

<sup>a</sup>1% dehydration = 10 mL/kg of fluid deficit;

<sup>a</sup>1 L of water = 1 kg of water

5. In a healthy child, insensible fluid volume loss is approximated as  $\frac{1}{3}$  of the Holliday-Segar MIVF per day. **NOTE:** This calculation is based on fluid requirements of healthy children. Many hospitalized children have increased insensible losses (e.g., secondary to fever or increased respiratory rate) that must be factored into fluid determinations.

### C. Maintenance Fluid Choice in Hospitalized Children

1. Based on a growing body of evidence, the AAP recommends isotonic fluid as the most appropriate MIVF therapy for the vast majority of hospitalized children between the ages of 28 days and 18 years.<sup>2</sup> See **Table 11.3** for isotonic fluid options.
2. Various disease states can lead to an increased secretion of antidiuretic hormone (ADH), which promotes the retention of free water, leading to hyponatremia.<sup>9,10</sup> See **Box 11.2** for examples.
3. Exceptions exist in certain patient populations, such as children with neuro-surgical disorders, congenital or acquired cardiac disease, hepatic disease, cancer, acute kidney injury, chronic kidney disease, nephrotic syndrome, diabetes insipidus, and voluminous watery diarrhea or severe burns.<sup>2</sup>
4. See **Table 11.3** and **Table 11.4** for electrolyte composition of various parenteral and enteral fluid replacement options.
5. Unless hyperkalemia is present or the child is in renal failure, maintenance potassium requirements (20 mEq/L of fluid) should be given.<sup>11</sup> Do not add potassium ( $K^+$ ) to fluids until urine output has been established.<sup>12,13</sup>

### D. Volume Replacement Strategy<sup>7,12,13</sup>

1. Volume resuscitation and deficit replacement should generally be completed over 24 hours.
2. See **Table 11.5** for a three-phase approach to fluid replacement.
3. Children with isonatremic hypovolemia can be repleted with isotonic fluid per AAP recommendations.<sup>2</sup> See **Box 11.3** for sample calculations in isonatremic hypovolemia.
4. If ongoing losses can be measured directly, they should be replaced 1:1 concurrently with maintenance fluid administration. If the losses cannot be measured, an estimate of 10 mL/kg body weight for each watery stool and 2 mL/kg body weight for each episode of emesis should be administered.<sup>3</sup> See **Table 11.6** for electrolyte composition of certain bodily fluids.
5. Oral intake is the preferred method for repletion and maintenance, if possible.

## III. ELECTROLYTE MANAGEMENT

See **Chapter 28** for age specific normal values of electrolytes.

### A. Serum Osmolality and Tonicity<sup>2,7,14</sup>

1. Fluids can be expressed in terms of their tonicity and their osmolality.

TABLE 11.3

## COMPOSITION OF FREQUENTLY USED PARENTERAL REHYDRATION FLUIDS

	D% CHO (g/100 mL)	Protein <sup>a</sup> (g/100 mL)	Cal/L	Na (mEq/L)	K <sup>+</sup> (mEq/L)	Cl <sup>-</sup> (mEq/L)	HCO <sub>3</sub> <sup>-b</sup> (mEq/L)	Mg <sup>2+</sup>	Ca <sup>2+</sup> (mEq/L)	mOsm/L
<b>HYPOTONIC</b>										
D <sub>5</sub> W	5	—	170	—	—	—	—	—	—	252
D <sub>10</sub> W	10	—	340	—	—	—	—	—	—	505
D <sub>5</sub> 1/4 NS (0.225% NaCl)	5	—	170	38.5	—	34	—	—	—	329
1/2 NS (0.45% NaCl)	—	—	—	77	—	77	—	—	—	154
<b>ISOTONIC</b>										
Lactated Ringer	0–10	—	0–340	130	4	109	28	—	3	273
Plamalyte	—	—	—	140	5	98	27	3	—	294
Ringer solution	0–10	—	0–340	147	4	155.5	—	—	≈4	—
NS (0.9% NaCl)	—	—	—	154	—	154	—	—	—	308
<b>HYPERTONIC</b>										
2% NaCl	—	—	—	342	—	342	—	—	—	684
3% NaCl	—	—	—	513	—	513	—	—	—	1027
8.4% sodium bicarbonate (1 mEq/mL)	—	—	—	1000	—	—	1000	—	—	2000
<b>COLLOID</b>										
Plasmanate	—	5	200	110	2	50	29	—	—	—
Amino acid 8.5% (Travasol)	—	8.5	340	3	—	34	52	—	—	880
Albumin 25% (salt poor)	—	25	1000	100–160	—	<120	—	—	—	300
Intralipid <sup>c</sup>	2.25	—	1100	2.5	0.5	4.0	—	—	—	258–284

<sup>a</sup>Protein or amino acid equivalent.<sup>b</sup>Bicarbonate or equivalent (citrate, acetate, lactate).<sup>c</sup>Values are approximate; may vary from lot to lot. Also contains < 1.2% egg phosphatides.CHO, Carbohydrate; HCO<sub>3</sub><sup>-</sup>, bicarbonate; NS, normal saline.

**BOX 11.2****CLINICAL SETTING OF INCREASED ADH RELEASE IN CHILDREN<sup>7,26</sup>**

Hemodynamic Stimuli for ADH Release (Decreased Effective Volume)	Nonosmotic and Nonhemodynamic Stimuli for ADH Release
Hypovolemia	CNS disturbances (infection, brain tumors, head injury, thrombosis)
Nephrosis	Pulmonary disease (pneumonia, asthma, bronchiolitis, PPV)
Cirrhosis	Cancer
Congestive heart failure	Medications (MDMA, AEDs, cytoxin, vincristine, opiates, TCAs, SSRIs)
Hypoaldosteronism	GI disturbances (nausea and emesis)
Hypotension	Pain or stress
Hypoalbuminemia	Postoperative state

*ADH*, Antidiuretic hormone; *AED*, antiepileptic drugs; *CNS*, central nervous system; *GI*, gastrointestinal; *MDMA*, 3,4-methylenedioxymethamphetamine (ecstasy); *PPV*; positive pressure ventilation; *SSRI*, selective serotonin reuptake inhibitor; *TCA*, tricyclic antidepressant.

11

2. Serum osmolality (285 to 295 mOsm/kg) is a measure of both permeable and nonpermeable solutes and is calculated using the following equation:

$$\text{EQUATION 11.4: Osmolality} = 2 \text{ Na} + \frac{\text{glucose (mg/dL)}}{18} + \frac{\text{BUN (mg/dL)}}{2.8}$$

3. Osmolality is measured as osmoles per weight (kg) versus osmolarity, which is measured as osmoles per volume (L).
4. Tonicity is effective osmolality. It is the net force on water across a semi-permeable membrane (e.g., the cell membrane) based on the osmotic pressures. It is relative and determined largely by sodium content. Substances that flow freely across membranes, such as urea, are ineffective osmoles and influence osmolality but not tonicity.

## B. Sodium

The equations within this section are **theoretical** and are not validated. They offer a starting point for calculation of electrolyte abnormalities, but clinical context is **ALWAYS** of the utmost importance and frequent monitoring is necessary. **Children with neurosurgical disorders, cardiac disease, hepatic disease, cancer, kidney disease, diabetes insipidus, and severe burns may require consultation with subspecialists before fluid choice and volume is administered.** When correcting dysnatremias, frequent lab monitoring (~2 to 4 hours) is indicated with adjustment of fluid type and rate as needed.

1. **Hyponatremia:** Excess Na loss ( $\text{Na} < 135 \text{ mEq/L}$ )
  - a. Clinical manifestations and differential diagnosis (**Table 11.7**)
  - b. Pseudohyponatremia etiologies:
    - (1) Increased serum osmolality: Hyperglycemia: Na artificially decreased 1.6 mEq/L for each 100-mg/dL rise in glucose
    - (2) Normal serum osmolality:
      - (a) Hyperlipidemia: Na artificially decreased by  $0.002 \times \text{lipid (mg/dL)}$

TABLE 11.4

## COMPOSITION OF ORAL REHYDRATION FLUIDS

	D% CHO (g/100 mL)	Na (mEq/L)	K <sup>+</sup> (mEq/L)	Cl <sup>-</sup> (mEq/L)	HCO <sub>3</sub> <sup>-</sup> <sup>b</sup> (mEq/L)	Ca <sup>2+</sup> (mEq/L)	mOsm/L
<b>ORAL FLUIDS</b>							
Pedialyte	2.5	45	20	35	30	—	250
WHO solution	2	90	20	80	30	—	310
Rehydralyte	2.5	75	20	65	30	—	310
<b>COMMONLY CONSUMED FLUIDS (NOT RECOMMENDED FOR ORAL REHYDRATION)<sup>a</sup></b>							
Apple juice	11.9	0.4	26	—	—	—	700
Coca-Cola	10.9	4.3	0.1	—	13.4	—	656
Gatorade	5.9	21	2.5	17	—	—	377
G2	4.7	20	3.2	—	—	—	—
Ginger ale	9	3.5	0.1	—	3.6	—	565
Milk	4.9	22	36	28	30	—	260
Orange juice	10.4	0.2	49	—	50	—	654
Powerade	5.8	18	2.7	—	—	—	264

<sup>a</sup>Electrolyte values are approximate.

<sup>b</sup>Bicarbonate or equivalent (citrate, acetate, lactate).

CHO, Carbohydrate; HCO<sub>3</sub><sup>-</sup>, bicarbonate; NS, normal saline; WHO, World Health Organization.

**TABLE 11.5****VOLUME REPLACEMENT STRATEGY**

Phase I	Phase II	Phase III
<b>Initial stabilization</b>	<b>Deficit repletion, maintenance volume, and ongoing losses</b>	<b>Recovery and ongoing losses</b>
Rapid fluid resuscitation with isotonic fluid. <sup>a</sup> 20 mL/kg represents only a 2% volume replacement	Replace half of the remaining deficit over the first 8 hr (this includes any fluid given in the initial stabilization phase). Replace the second half of deficit over the following 16 hr, making sure to also include maintenance fluid volume replacement during this time.	Continue maintenance fluid replacement, taking ongoing losses into consideration.

<sup>a</sup>Should be used in patients in need of rapid volume expansion.

See Box 11.3 for sample calculation

**BOX 11.3****SAMPLE CALCULATIONS: ISONATREMIC DEHYDRATION**

Example: A 15-kg (preillness weight) child with 10% dehydration and normal serum sodium

Requirement	Formula	Sample Calculation
Maintenance fluid requirements	Holliday–Segar formula	(100 mL/kg/day × 10 kg) + (50 mL/kg/day × 5 kg) = 1250 mL/24 hr = 52 mL/hr
Fluid deficit	Equation 11.2 or Equation 11.3	10 mL × 15 kg × 10% = 1500 mL

**Fluid Replacement Rate Over 24 hrs**

$\frac{1}{2}$  fluid deficit replaced in first 8 hrs       $750 \text{ mL}/8 \text{ hr} = 94 \text{ mL/hr} + 52 \text{ mL/hr maintenance} = 146 \text{ mL/hr}$

$\frac{1}{2}$  fluid deficit replaced over 16 hrs       $750 \text{ mL}/16 \text{ hr} = 47 \text{ mL/hr} + 52 \text{ mL/hr maintenance} = 99 \text{ mL/hr}$

**Note:** If patient received an initial 20 mL/kg bolus (300 mL):  $1500 \text{ mL} - 300 \text{ mL} = 1200 \text{ mL}$

$\frac{1}{2}$  fluid deficit in first 8 hrs:  $600 \text{ cc}/8 \text{ hr} = 75 \text{ mL} + 52 \text{ mL/hr maintenance} = 127 \text{ mL/hr}$

$\frac{1}{2}$  fluid deficit over next 16 hrs:  $600 \text{ cc}/16 \text{ hr} = 38 \text{ mL/hr} + 52 \text{ mL/hr maintenance} = 90 \text{ mL/hr}$

11

(b) Hyperproteinemia: Na artificially decreased by  $0.25 \times [\text{protein (g/dL)} - 8]$

c. Management

- (1) The traditional equation used to calculate the excess sodium deficit in hyponatremia is:

**EQUATION 11.5<sup>3</sup>:**

$$\text{Na deficit(mEq)}^a = [\text{Desired Na (mEq/L)} - \text{Serum Na (mEq/L)}] \times \text{TBW (L)}$$

<sup>a</sup>This represents the excess sodium deficit in hyponatremic dehydration. It must be added to the daily sodium requirement for hospitalized patients of ~14 mEq/100 mL fluid given.

- (2) Hyponatremia should be corrected **by no more than 10 to 12 mEq per 24 hr** to avoid rapid change of serum sodium, which can cause osmotic demyelination syndrome.<sup>6,13,15</sup>

**TABLE 11.6****ELECTROLYTE COMPOSITION OF VARIOUS FLUIDS**

Source of Fluid	$\text{Na}^+$ (mEq/L)	$\text{K}^+$ (mEq/L)	$\text{Cl}^-$ (mEq/L)
Gastric	20–80	5–20	100–150
Pancreatic	120–140	5–15	90–120
Small bowel	100–140	5–15	90–130
Bile	120–140	5–15	80–120
Ileostomy	45–135	3–15	20–115
Diarrhea	10–90	10–80	10–110
Skin with burns <sup>a</sup>	140	5	110
Sweat			
Normal	10–30	3–10	10–35
Cystic fibrosis <sup>b</sup>	50–130	5–25	50–110

<sup>a</sup>3–5 g/dL of protein may be lost in fluid from burn wounds.

<sup>b</sup>Replacement fluid dependent on sodium content.

Modified from Kliegman RM, Stanton B, St. Geme J, et al. *Nelson Textbook of Pediatrics*. 19th ed. Philadelphia: Saunders; 2011.

**TABLE 11.7****HYPONATREMIA<sup>7,14</sup>****CLINICAL MANIFESTATIONS**

Related to rate of change: Nausea, headache, muscle cramps, weakness, confusion, apnea, lethargy, seizure, coma, hypothermia, depressed DTRs

**ETIOLOGIES**

Hypovolemic	Euvolemic	Hypervolemic
<b>Renal Losses</b>	<b>Extrarenal Losses</b>	
Na-losing nephropathy	GI losses	SIADH (see Chapter 10)
Diuretics	Skin losses	Excess salt-free infusions
Juvenile nephronophtisis	Third spacing	Hypoalbuminemia
Hypoaldosteronism (CAH, pseudohypoaldosteronism, UTI/obstruction)	Cystic fibrosis	Desmopressin acetate
Cerebral salt-wasting syndrome		Water intoxication
Postobstructive diuresis		Hypothyroidism
ATN (polyuric phase)		Sepsis
		Primary polydipsia <sup>c</sup>
		Malnutrition <sup>c</sup>

**LABORATORY DATA**

↑ Urine Na (> 20 mEq/L)	↓ Urine Na (< 20 mEq/L)	↓ Urine volume	↓ Urine $\text{Na}^b$ (< 20 mEq/L)
↑ Urine volume		↑ Specific gravity	
↓ Specific gravity	↓ Urine volume	↑ Urine osmolality (> 100 mOsm/L)	↓ Urine volume
↓ Urine osmolality <sup>a</sup> (< 100 mOsm/L)	↑ Specific gravity		
	↑ Urine osmolality (> 100 mOsm/L)		

**MANAGEMENT**

Replace losses (see hypovolemic hyponatremia)

Restrict fluids

Address the underlying cause

<sup>a</sup>Minimum possible urine osmolality = 50 mOsm/kg

<sup>b</sup>Urine Na may be appropriate for the level of Na intake in patients with SIADH and water intoxication.

<sup>c</sup>Urine osmolality is <100 mOsm/L

ATN, Acute tubular necrosis; CAH, congenital adrenal hyperplasia; DTR, deep tendon reflex; GI, gastrointestinal; Na, sodium; SIADH, syndrome of inappropriate antidiuretic hormone secretion; UTI, urinary tract infection.

- (3) Witnessed onset of hyponatremia over the course of hours does not pose as great of a risk and can be corrected in a similar amount of time that it developed.<sup>7</sup>
- (4) If central nervous system (CNS) symptoms are present, hypertonic saline (HTS) should be administered over 3 to 4 hours to correct the hyponatremia by ~5 mEq/L.<sup>5,6,11</sup> Use [Equation 11.7](#) to determine rate of HTS.
- (5) To determine the sodium content of the fluid necessary for repletion:

**EQUATION 11.6:**

Na content (mEq / L) =

$$\frac{[\text{Na deficit} + (14 \text{ mEq} / 100 \text{ mL} \times \text{maintenance fluid volume [mL]})]}{\text{volume deficit}^a}$$

<sup>a</sup>Use daily maintenance volume requirements if euvolemic

- (6) Once the fluid type is determined, the starting rate can be calculated using the following:

**EQUATION 11.7:**

$$\text{Fluid rate (mL/hour)} = \frac{\text{Na deficit (mEq)} \times 1000 \text{ mL}}{\text{infuse Na (mEq)} \times \text{hours IVF will run in a day}}$$

- (7) See [Box 11.4](#) and [11.5](#) for sample calculations in hyponatremic dehydration.

**2. Hypernatremia:** Excess free water loss ( $\text{Na} > 145 \text{ mEq/L}$ )

- a. Clinical manifestations and differential diagnosis ([Table 11.8](#))
- b. Management

- (1) Hypernatremic hypovolemia occurs in scenarios in which free water is either unavailable/restricted or there is excessive loss of solute-free water (see [Table 11.8](#)).
- (2) Hypernatremia is dangerous because of complications from potential treatment sequelae, the most serious of which is cerebral edema.<sup>4,7</sup>
- (3) Plan to correct the serum Na by no more than 10 mEq/24 hours and correct the free water deficit over 48 hours to minimize the risk of cerebral edema.<sup>4,10,11,16</sup>
- (4) As with hyponatremia, witnessed onset of hypernatremia over the course of hours can be corrected rapidly; this is because the brain has not had time to produce idiogenic osmoles to adapt to the change in osmolality.<sup>7,11</sup>
- (5) Expert opinion recommends starting with D5 ½ NS.<sup>16</sup> However, the sodium and fluid needs can also be calculated.
- (6) The free water deficit is as follows:

**EQUATION 11.8<sup>4,6</sup>:**

$$\text{FWD (mL)} = \text{TBW (mL)} \times \left[ 1 - \frac{\text{Desired Na (mEq/L)}}{\text{Serum Na (mEq/L)}} \right]^a$$

<sup>a</sup>The difference in desired and serum Na should be no more than 10 mEq/L/day

**BOX 11.4****SAMPLE CALCULATIONS: HYponatremic Dehydration**

Example: A 15-kg (preillness weight) child with 10% dehydration and serum sodium 125 mEq/L without central nervous system symptoms

Requirement	Formula	Sample Calculation
Maintenance fluid requirements	Holliday-Segar formula	(100 mL/kg/d × 10 kg) + (50 mL/kg/d × 5 kg) = 1250 mL/24 hr = 52 mL/hr
Fluid deficit	<a href="#">Equation 11.2</a> or <a href="#">Equation 11.3</a>	10 mL × 15 kg × 10% = 1500 mL
<b>Fluid Replacement Rate Over 24 hrs</b>		
1500 mL/24 hr = 63 mL/hr + 52 mL/hr maintenance = 115 mL/hr		
<b>Calculations for Fluid Selection</b>		
Maintenance sodium requirements	3 mEq per 100 mL of maintenance fluid	3 mEq × (1250 mL/100 mL) = 38 mEq Na <sup>+</sup>
Isotonic sodium deficit	8–10 mEq Na <sup>+</sup> per each 100 mL of fluid deficit	10 mEq × (1500 mL/100 mL) = 150 mEq Na <sup>+</sup>
Sodium deficit	<a href="#">Equation 11.5</a>	(135 mEq – 125 mEq) × 9 = 90 mEq Na <sup>+</sup>
Total sodium content	<a href="#">Equation 11.6</a>	90 mEq + (14 mEq/100mL × 1250) = 265 mEq
Sodium required per L	Divide total sodium by fluid deficit in L	278 mEq/1.5 L = 185 mEq

**BOX 11.5****SAMPLE CALCULATIONS: SEVERE SYMPTOMATIC HYponatremic DEHYDRATION****Initial Fluid Replacement for Neurologic Stabilization**

Example: A 15-kg (preillness weight) child with altered mental status and serum sodium 110 mEq/L

Fluid to be used: 3% hypertonic saline (HTS)

Requirement	Formula	Sample Calculation
Sodium deficit	<a href="#">Equation 11.5</a>	5 mEq/L × 9 = 45 mEq Na <sup>+</sup>
Rate of administration	<a href="#">Equation 11.7</a>	[(45 mEq × 1000 mL) /513 mEq × 4 hrs] = 22 mL/hr of 3% HTS

- (7) The FWD is used to calculate the solute fluid deficit (SFD) (i.e., the amount of fluid that contains electrolytes).

**EQUATION 11.9:** SFD = Fluid Deficit <sup>a</sup> – FWD

<sup>a</sup>See equation 11.2 for fluid deficit calculations

- (8) Despite the hypernatremia, there is also a Na deficit that should be accounted for:

**TABLE 11.8****HYPERNATREMIA<sup>7,25</sup>****CLINICAL MANIFESTATIONS**

With hypernatremic hypovolemia, there is better preservation of intravascular volume compared to hypovolemic hyponatremia. Lethargy, weakness, altered mental status, irritability, coma, and seizures. High-pitched cry, thrombosis, brain hemorrhage, muscle cramps, hyperpnea, and respiratory failure.

**ETIOLOGIES**

Low urine osmolality	Elevated urine osmolality <sup>b</sup>	
	↓ Urine Na (< 20 mEq/L)	↑ Urine Na (> 20 mEq/L)
Diabetes insipidus (central and nephrogenic) (see Chapters 10 and 19)	GI losses Skin losses Respiratory <sup>a</sup>	Exogenous Na <sup>+</sup> (meds, infant formula) Mineralocorticoid excess (e.g., hyperaldosteronism)
Postobstructive diuresis	Increased insensible losses	
CKD	Adipsia	
Diuretic use		
Polyuric phase of ATN		

**MANAGEMENT**

Timeline of onset can mirror timeline for correction.

<sup>a</sup>This cause of hypernatremia is usually secondary to free water loss; therefore the fractional excretion of sodium may be decreased or normal.

<sup>b</sup>>1000 mosm/kg

ATN, Acute tubular necrosis; CKD, chronic kidney disease; GI, gastrointestinal; Na, sodium.

11

**EQUATION 11.10:**

$$\text{Na required (mEq)} = [\text{SFD (mL)} + \text{maintenance fluid volume (mL)}] \times \frac{14 \text{ mEq}}{100 \text{ mL}}$$

- (9) The amount of sodium is then divided by the total fluid deficit in addition to the maintenance fluid volume. This will help approximate the fluid tonicity required.

**EQUATION 11.11:**

$$\text{Na content of fluid (mEq / L)} = \frac{\text{Na required (mEq)}}{\text{Fluid Deficit (L)} + \text{maintenance fluid volume (L)}}$$

- (10) See Box 11.6 for sample calculations in hypernatremic dehydration.

- (11) If the fluid necessary contains >154 mEq of Na, then the following equation can be used to make a 1-L bag at the desired tonicity:<sup>16</sup>

**EQUATION 11.12:**

$$\text{mL of 3% saline} = 1000 \text{ mL} \times \frac{\text{desired Na (mEq / L)} - 154 \text{ (mEq / L)}}{513 \text{ (mEq / L)} - \text{desired Na (mEq / L)}}$$

- (12) This equation can also be used to calculate rate to run HTS with NS bolus in a severely hypernatremic child. See Box 11.7.

**BOX 11.6****SAMPLE CALCULATIONS: HYPERNATREMIC DEHYDRATION**

Example: A 15-kg (preillness weight) child with 10% dehydration and serum sodium 155 mEq/L

Requirement	Formula	Sample Calculation
Maintenance fluid requirements	Holliday-Segar formula	$(100 \text{ mL/kg/d} \times 10 \text{ kg}) + (50 \text{ mL/kg/d} \times 5 \text{ kg}) = 1250 \text{ mL/24 hr} = 52 \text{ mL/hr}$
Total fluid deficit	<a href="#">Equation 11.2</a> or <a href="#">Equation 11.3</a>	$10 \text{ mL} \times 15 \text{ kg} \times 10\% = 1500 \text{ mL}$
<b>Fluid Replacement Rate Over 24 hrs</b>		
$1500 \text{ mL/24 hr} = 63 \text{ mL/hr} + 52 \text{ mL/hr maintenance} = 115 \text{ mL/hr}$		
<b>Calculations for Fluid Selection</b>		
Free water deficit	<a href="#">Equation 11.8</a>	$4 \text{ mL/kg} \times 15 \text{ kg} \times (155 \text{ mEq/L} - 145 \text{ mEq/L}) = 600 \text{ mL}$
Solute fluid deficit	<a href="#">Equation 11.9</a>	$1500 \text{ mL} - 600 \text{ mL} = 900 \text{ mL}$
Total sodium required	<a href="#">Equation 11.10</a>	$(900 \text{ mL} + 1250 \text{ mL}) \times 14 \text{ mEq/100 mL} = 300 \text{ mEq Na}^+$
Na content of fluid	<a href="#">Equation 11.11</a>	$300 \text{ mEq} / (1.25 + 1.5 \text{ L}) = 110 \text{ mEq Na}$

**BOX 11.7****SAMPLE CALCULATIONS: SEVERE HYPERNATREMIC DEHYDRATION**
**Initial Fluid Resuscitation Strategy to Avoid Rapid Sodium Correction when Serum  $\text{Na}^+ > 175 \text{ mEq/L}$ <sup>16</sup>**

Example: A 3-kg (preillness-weight) breastfed neonate appearing severely dehydrated with serum sodium 185 mEq/L and hemodynamic instability

Resuscitation with normal saline (NS) may drop the serum  $\text{Na}^+$  too quickly. Plan to simultaneously run NS and 3% hypertonic saline (HTS), given rapidly together (i.e., over 5 minutes), to effectively give resuscitation fluid with a concentration no more than 15 mEq/L below the child's serum  $\text{Na}^+$ . Repeat the boluses as needed to achieve hemodynamic stability.

Requirement	Formula	Sample Calculation
Ideal bolus fluid concentration	Serum sodium (in mEq/L) – 15 mEq/L	$185 \text{ mEq/L} - 15 \text{ mEq/L} = 170 \text{ mEq/L}$
mL of HTS required per L of NS	<a href="#">Equation 11.12</a>	$1000 \text{ mL} \times (170 \text{ mEq/L} - 154 \text{ mEq/L}) / (513 \text{ mEq/L} - 170 \text{ mEq/L}) = 47 \text{ mL}$
Bolus NS amount in mL	$20 \text{ mL/kg} \times \text{wt (in kg)}$	$20 \text{ mL/kg} \times 3 \text{ kg} = 60 \text{ mL}$
Bolus amount HTS in mL	$\text{mL HTS required per L of NS} \times \text{NS bolus amount (in mL)} / 1000 \text{ mL}$	$47 \text{ mL} \times 60 \text{ mL} / 1000 \text{ mL} = 2.8 \text{ mL}$

Note: In clinical practice, one will often not have laboratory data available quickly enough to employ this strategy. However, severe hypernatremia should be suspected in the clinical scenario of a solely breastfed neonate who appears severely dehydrated.<sup>16</sup> STAT labs should be sent, and this strategy may be employed as soon as laboratory values are available.

3. Calculations pertaining to dysnatremias can be double-checked using the following equation:

**EQUATION 11.13:**<sup>4-6</sup>

$$\frac{\text{Change in Serum Na}}{1\text{L of parenteral fluid administration}} = \frac{(\text{Infuse Na} + \text{Infuse K}) - \text{Serum Na}}{\text{TBW} + 1}$$

**C. Potassium**

1. **Hypokalemia**

- a. Clinical manifestations and differential diagnosis ([Table 11.9](#))
- b. The transtubular potassium gradient (TTKG) can help differentiate between etiologies of hypokalemia, as noted in [Table 11.9](#):

**EQUATION 11.14:**<sup>7</sup>

$${}^7\text{TTKG}^a = \frac{[\text{K}]_{\text{urine}}}{[\text{K}]_{\text{plasma}}} \times \left( \frac{\text{plasma osmolality}}{\text{urine osmolality}} \right)$$

<sup>a</sup>The urine osmolality must be greater than the serum osmolality for the calculation to be valid

- c. Management: Potassium infusion rates generally should not exceed 1 mEq/kg/hr.<sup>3</sup>

2. **Hyperkalemia**

- a. Clinical manifestations and differential diagnosis ([Table 11.10](#))
- b. Management ([Fig. 11.1](#))

**D. Calcium**

1. **Hypocalcemia**

- a. Clinical manifestations and differential diagnosis ([Table 11.11](#))
- b. Special considerations:
  - i. Albumin readily binds serum calcium. Correction for albumin:  $\Delta$  of 1 g/dL changes the total serum calcium in the same direction by 0.8 mg/dL.
  - ii. pH: Acidosis increases ionized calcium.
  - iii. Symptoms of hypocalcemia refractory to calcium supplementation may be caused by hypomagnesemia.
  - iv. Significant hyperphosphatemia should be corrected before the correction of hypocalcemia because renal calculi or soft-tissue calcification may occur if total  $[\text{Ca}^{2+}] \times [\text{PO}_4^{3-}] \geq 70$ .<sup>7</sup>

2. **Hypercalcemia:** [Table 11.11](#)

**E. Magnesium**

1. Hypomagnesemia: [Table 11.12](#)
2. Hypermagnesemia: [Table 11.12](#)

**F. Phosphate**

1. Hypophosphatemia: [Table 11.13](#)
2. Hyperphosphatemia: [Table 11.13](#)

TABLE 11.9

HYPOKALEMIA<sup>7,25</sup>

## CLINICAL MANIFESTATIONS

Manifest at levels <2.5 mEq/L. Skeletal muscle weakness or ascending paralysis, muscle cramps, ileus, urinary retention, and cardiac arrhythmias.

Electrocardiogram (ECG) changes:

Delayed depolarization, flat T waves, depressed ST segment, and U waves.

## ETIOLOGIES

Decreased Stores					
Metabolic Alkalosis					
Hypertensive	Normotensive	Metabolic Acidosis	No Change in Serum pH	Extrarenal	Normal Stores <sup>a</sup>
Renovascular disease	Gittleman syndrome	RTA (type I and II)	Meds (amphotericin, cis-platin, aminoglycosides, penicillin or penicillin derivatives, diuretics)	Skin losses GI losses/laxative abuse/enema abuse Clay ingestion Kayexalate Interstitial nephritis	Acute metabolic alkalosis Hyperinsulinemia Leukocytosis (if sample sits at room temperature) Meds (adrenergic agonists, theophylline, toluene, cesium chloride, hydroxychloroquine, barium) Familial hypokalemic periodic paralysis Familial
Excess renin	Bartter syndrome	DKA			
Cushing syndrome	Hypoparathyroidism	Uretosigmoidoscopy			
CAH	Cystic fibrosis	Fanconi Syndrome			
Adrenal adenoma	EAST syndrome				
Licorice ingestion	Loop and thiazide diuretics				
Liddle syndrome	Emesis				

## LABORATORY DATA

TTKG > 4

TTKG ≤ 4

~ Urine K<sup>+</sup>

## MANAGEMENT

Acute	Calculate deficit and replace with potassium acetate or potassium chloride. Enteral replacement is safer when feasible. Follow K <sup>+</sup> closely. IV replacement generally should not exceed 1 mEq/kg given over 1 hr.
Chronic	Determine daily requirement and replace with potassium chloride or potassium gluconate.

<sup>a</sup>Blood pressure may vary.

CAH, Congenital adrenal hyperplasia; DKA, diabetic ketoacidosis; GI, gastrointestinal; K<sup>+</sup>, potassium; RTA, renal tubular acidosis; EAST, epilepsy, ataxia, sensorineural hearing loss, and tubulopathy; TTKG, transtubular potassium gradient.

**TABLE 11.10****HYPERKALEMIA<sup>7</sup>****CLINICAL MANIFESTATIONS**

Skeletal muscle weakness, fasciculations, paresthesias, and ascending paralysis.

The typical ECG progression with increasing serum K<sup>+</sup> values:

1. Peaked T waves
2. Prolonged PR and widening of QRS
3. Loss of P waves
4. ST segment depression with further widening of QRS
5. Bradycardia, atrioventricular (AV) block, ventricular arrhythmias, torsades de pointes, and cardiac arrest

**ETIOLOGIES**

<b>Increased total body K<sup>+</sup></b>		<b>Intracellular shifts (no change in total body K<sup>+</sup>)</b>
<b>Increased urine K<sup>+</sup></b>	<b>Decreased urine K<sup>+</sup></b>	
Transfusion with aged blood	Renal failure	Tumor lysis syndrome
Exogenous K <sup>+</sup>	Hypoaldosteronism	Leukocytosis (>200 x 10 <sup>3</sup> /μL)
Spitzer syndrome	Aldosterone insensitivity	Thrombocytosis (>750 x 10 <sup>3</sup> /μL) <sup>b</sup>
	↓ Insulin causing hyperglycemia and/or DKA	Metabolic acidosis <sup>a</sup>
	K <sup>+</sup> -sparing diuretics	Blood drawing (hemolyzed sample)
	Congenital adrenal hyperplasia	Rhabdomyolysis/crush injury
	Type IV RTA	Malignant hyperthermia
	Meds: ACE inhibitors, angiotensin II blockers, K sparing diuretics, calcineurin inhibitors, NSAIDs, heparin, TMX, spironolactone	Theophylline intoxication

**MANAGEMENT**

See Fig. 11.1.

<sup>a</sup>For every 0.1-unit reduction in arterial pH, there is approximately a 0.2–0.4 mEq/L increase in plasma K<sup>+</sup>.

<sup>b</sup>For every platelet increase of 100,000/μL, there is a 0.15 mEq/L increase in serum K<sup>+</sup>.

ACE, Angiotensin converting enzyme; DKA, diabetic ketoacidosis; ECG, electrocardiogram; K<sup>+</sup>, potassium; NSAIDs, nonsteroidal antiinflammatory drugs; RTA, renal tubular acidosis; TMX, trimethoprim.

**IV. ALGORITHM FOR EVALUATING ACID-BASE DISTURBANCES<sup>7,17,18</sup>****A. Determine the pH**

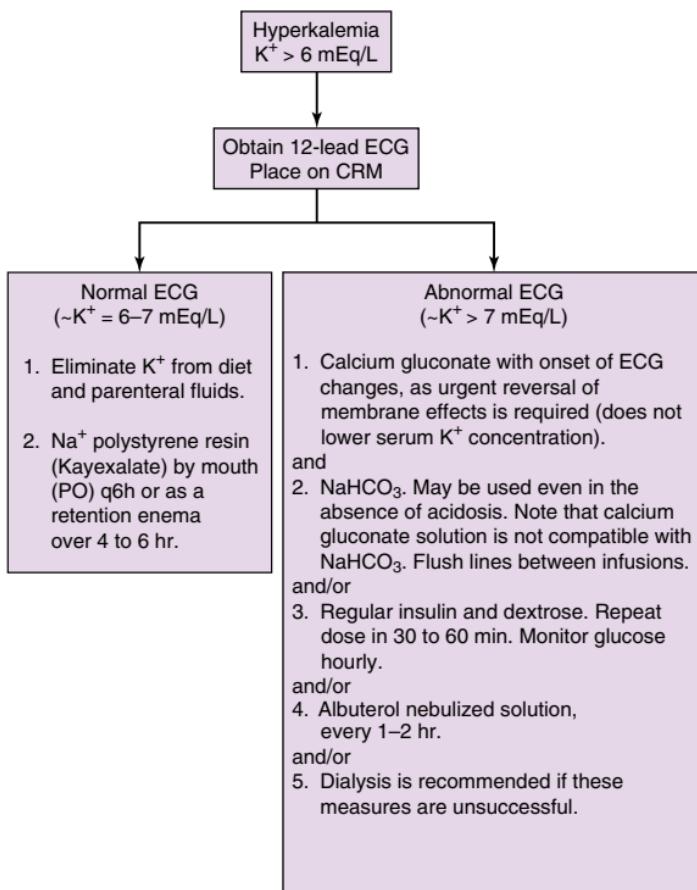
The body does not fully compensate for primary acid-base disorders; therefore the primary disturbance will shift the pH away from 7.40.

**1. Acidemia (pH < 7.35):**

- a. Respiratory acidosis: PCO<sub>2</sub> > 45 mm Hg
- b. Metabolic acidosis: Arterial bicarbonate < 20 mmol/L

**2. Alkalolemia (pH > 7.45):**

- a. Respiratory alkalosis: PCO<sub>2</sub> < 35 mm Hg
- b. Metabolic alkalosis: Arterial bicarbonate > 28 mmol/L

**FIGURE 11.1**

Algorithm for hyperkalemia. *CRM*, Cardiorespiratory monitor; *D25W*, 25% dextrose in water; *ECG*, electrocardiogram; *INH*, inhaled; *IV*, intravenous.

**TABLE 11.11****HYPOCALCEMIA AND HYPERCALCEMIA**

<b>Hypocalcemia</b>	<b>Hypercalcemia</b>
<b>CLINICAL MANIFESTATIONS</b>	
Tetany, neuromuscular irritability with weakness, paresthesias, fatigue, cramping, altered mental status, seizures, laryngospasm, and cardiac arrhythmias <sup>18,19</sup> :	Weakness, irritability, lethargy, seizures, coma, abdominal cramping, anorexia, nausea, vomiting, polyuria, polydipsia, renal calculi, pancreatitis, and ECG changes (shortened QT interval)
<ul style="list-style-type: none"> <li>• ECG changes (prolonged QT interval)</li> <li>• Troussseau's sign (carpopedal spasm after arterial occlusion of an extremity for 3 minutes)</li> <li>• Chvostek sign (muscle twitching on percussion of the facial nerve)</li> </ul>	
<b>ETIOLOGIES</b>	
Hypoparathyroidism	Hyperparathyroidism
Vitamin D deficiency	Vitamin D intoxication
Hyperphosphatemia	Excessive exogenous calcium administration
Pancreatitis	Malignancy
Malabsorption (malnutrition)	Prolonged immobilization
Drugs (anticonvulsants, cimetidine, aminoglycosides, calcium channel blockers)	Thiazide diuretics
Hypomagnesemia/hypermagnesemia	Subcutaneous fat necrosis
Maternal hyperparathyroidism (in neonates)	Williams syndrome
Ethylene glycol ingestion	Granulomatous disease (e.g., sarcoidosis)
Calcitriol (activated vitamin D) insufficiency	Hyperthyroidism
Tumor lysis syndrome	Milk-alkali syndrome
<b>MANAGEMENT</b>	
Acute	Consider IV replacement (calcium gluconate, calcium gluceptate, or calcium chloride [cardiac arrest dose])
Chronic	Consider use of oral supplements of calcium carbonate, calcium gluconate, calcium gluionate, or calcium lactate
	Increase UOP and $\text{Ca}^{2+}$ excretion: 1. If the glomerular filtration rate and blood pressure are stable, give NS with maintenance $\text{K}^+$ at 2-3 times the maintenance rate 2. Diuresis with furosemide Consider hemodialysis for severe or refractory cases Consider steroids in malignancy, granulomatous disease, and vitamin D toxicity to decrease vitamin D and $\text{Ca}^{2+}$ absorption Severe or persistently elevated $\text{Ca}^{2+}$ : Consider calcitonin or bisphosphonate

$\text{Ca}^{2+}$ , Calcium; ECG, electrocardiogram; UOP, urine output.

### B. Calculate the anion gap (AG)

1. **AG:** Represents anions other than bicarbonate and chloride required to balance the positive charge of Na. Normal:  $12 \text{ mEq/L} \pm 2 \text{ mEq/L}$ .

$$\text{EQUATION 11.15: } \text{AG} = \text{Na}^+ - (\text{Cl}^- + \text{HCO}_3^-)$$

**TABLE 11.12****HYPOMAGNESEMIA AND HYPERMAGNESEMIA<sup>7</sup>**

Hypomagnesemia		Hypermagnesemia	
CLINICAL MANIFESTATIONS			
Typically, dominant manifestations are caused by concurrent hypocalcemia ( <b>Table 11.11</b> )		Typically occur at levels >4.5 mg/dL: Hypotonia, hyporeflexia, paralysis, lethargy, confusion, hypotension, and prolonged QT, QRS, and PR intervals.	
Typically occur at levels <0.7 mg/dL: Anorexia, nausea, weakness, malaise, depression, nonspecific psychiatric symptoms, hyper-reflexia, ECG changes: flattening of T wave and lengthening of ST segment		Respiratory failure and cardiac arrest at >15 mg/dL	
ETIOLOGIES			
GI Disorders	Genetic	Medications	Miscellaneous
Diarrhea	Gitelman syndrome	Amphotericin	Decreased intake
Malabsorption diseases	Bartter syndrome	Cisplatin	Hungry bone syndrome
Short bowel	EAST syndrome	Cyclosporine	Exchange transfusion
Malnutrition	AD hypoparathyroidism	Loop and thiazide diuretics	Diabetes mellitus
Pancreatitis	Mitochondrial disorders	Mannitol	Steatorrhea
	Miscellaneous disorders	Pentamidine	Hyperaldosteronism
Renal Failure and Excessive Administration			
Status asthmaticus eclampsia/preeclampsia, cathartics, enemas, phosphate binders, laxatives, lithium ingestions, milk-alkali syndrome			
MANAGEMENT			
Acute		IV Magnesium sulfate	Stop supplemental Mg <sup>2+</sup>
Chronic		PO Magnesium oxide or magnesium sulfate	Diuresis Ca <sup>2+</sup> supplements, such as calcium chloride (cardiac arrest doses) or calcium gluconate

*AD*, Autosomal dominant; *Ca<sup>2+</sup>*, calcium; *EAST*, epilepsy, ataxia, sensorineural hearing loss, and tubulopathy; *ECG*, electrocardiogram; *GI*, gastrointestinal; *IV*, intravenous; *Mg<sup>2+</sup>*, magnesium; *PO*, by mouth.

2. The majority of unmeasured anions contributing to the AG in normal individuals are albumin and phosphate. Correcting the AG for albumin concentration increases the utility of the traditional method.<sup>19</sup>

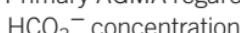
**EQUATION 11.16:** Corrected AG =

$$\text{Observed AG} + 2.5 \times (\text{Normal albumin} - \text{measured albumin})$$

AG > 15 : Anion gap metabolic acidosis (AGMA)

AG < 12 : Nonelevated anion gap metabolic acidosis (NAGMA)

AG > 20 mEq / L : Primary AGMA regardless of the pH or serum



**TABLE 11.13****HYPOPHOSPHATEMIA AND HYPERPHOSPHATEMIA<sup>7</sup>**

Hypophosphatemia	Hyperphosphatemia	
<b>CLINICAL MANIFESTATIONS</b>		
Symptomatic only at very low levels (<1 mg/dL). Acute: rhabdomyolysis, tremor, paresthesias, irritability, confusion, hemolysis, delirium, seizure, myocardial depression, and coma. Chronic: Rickets, proximal muscle weakness	Symptoms of resulting hypocalcemia and systemic calcification (i.e., deposition of phosphorus calcium salts in tissues).	
<b>ETIOLOGIES</b>		
Refeeding syndrome Insulin BMT Hungry bone Decreased intake Antacids Glucocorticoids Rickets Hyperparathyroidism Increased renal losses (e.g., renal tubular defects, diuretic use) McCune-Albright syndrome Epidermal nevus syndrome Fanconi syndrome Metabolic acidosis/respiratory alkalosis Glycosuria Volume expansion Sepsis	Tumor lysis syndrome Rhabdomyolysis DKA/lactic acidosis Hemolysis Renal failure Hypoparathyroidism Hyperthyroidism Excessive intake (enemas/laxatives and cow's milk) Vitamin D intoxication Familial tumoral calcinosis Acromegaly	
<b>MANAGEMENT</b>		
Acute Chronic	IV potassium phosphate or sodium phosphate PO potassium phosphate or sodium phosphate	Restrict dietary phosphate. Phosphate binders (calcium carbonate, aluminum hydroxide)

BMT, Bone marrow transplant; DKA, diabetic ketoacidosis. /V, intravenous; PO, by mouth.

**C. Calculate the delta gap (DG)<sup>20</sup>:**

If there is an AGMA, calculating the DG will help to determine if there is another, concurrent metabolic abnormality:

$$\text{EQUATION 11.17: } \text{DG} = (\text{AG} - 12) - (24 - \text{HCO}_3^-)$$

DG > 6: combined AGMA and metabolic alkalosis.

DG < -6: combined AGMA and NAGMA.

**D. Calculate the osmolar gap**

$$\text{EQUATION 11.18: Serum osmolar gap} = \text{calculated serum osmolality} - \text{laboratory measured osmolality}$$

**TABLE 11.14****CALCULATION OF EXPECTED COMPENSATORY RESPONSE<sup>7,20</sup>**

Disturbance	Primary Change	Expected Compensatory Response
Acute respiratory acidosis	$\uparrow \text{PaCO}_2$	$\uparrow \text{HCO}_3^-$ by 1 mEq/L for each 10 mmHg rise in $\text{PaCO}_2$
Acute respiratory alkalosis	$\downarrow \text{PaCO}_2$	$\downarrow \text{HCO}_3^-$ by 2 mEq/L for each 10 mmHg fall in $\text{PaCO}_2$
Chronic respiratory acidosis	$\uparrow \text{PaCO}_2$	$\uparrow \text{HCO}_3^-$ by 4 mEq/L for each 10 mmHg rise in $\text{PaCO}_2$
Chronic respiratory alkalosis	$\downarrow \text{PaCO}_2$	$\downarrow \text{HCO}_3^-$ by 4 mEq/L for each 10 mmHg fall in $\text{PaCO}_2$
Metabolic acidosis	$\downarrow \text{HCO}_3^-$	$\text{PaCO}_2 = 1.5 \times [\text{HCO}_3^-] + 8 \pm 2$
Metabolic alkalosis	$\uparrow \text{HCO}_3^-$	$\uparrow \text{PaCO}_2$ by 7 mmHg for each 10 mEq/L rise in $\text{HCO}_3^-$

- There is always a difference (<6) between calculated osmolality and measured osmolality.<sup>21</sup>
- A markedly elevated osmolar gap (>10) in the setting of an AG acidosis is highly suggestive of acute methanol or ethylene glycol intoxication.<sup>22–24</sup>

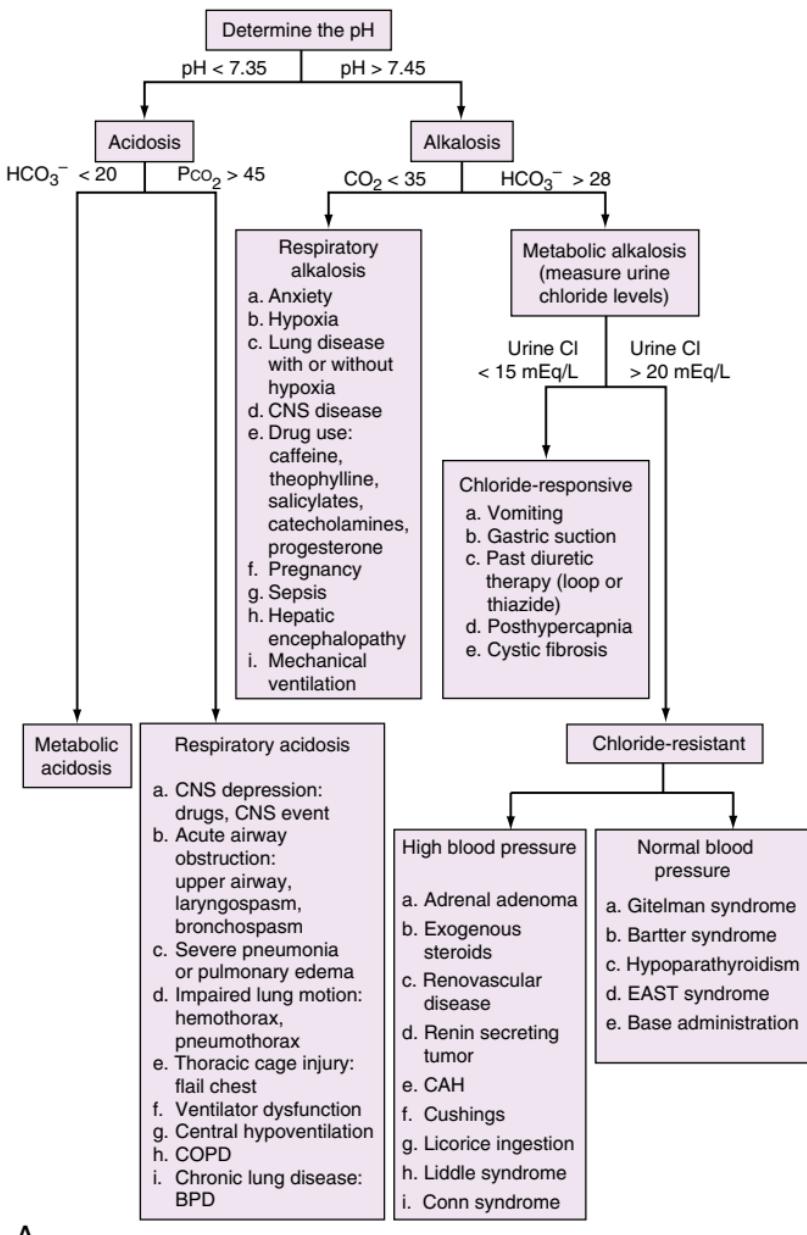
**E. Calculate expected compensatory response: (Table 11.14)**

- Pure **respiratory** acidosis (or alkalosis): 10 mmHg rise (fall) in  $\text{PaCO}_2$  results in an average 0.08 fall (rise) in pH.
- Pure **metabolic** acidosis (or alkalosis): 10 mEq/L fall (rise) in  $\text{HCO}_3^-$  results in an average 0.15 fall (rise) in pH.

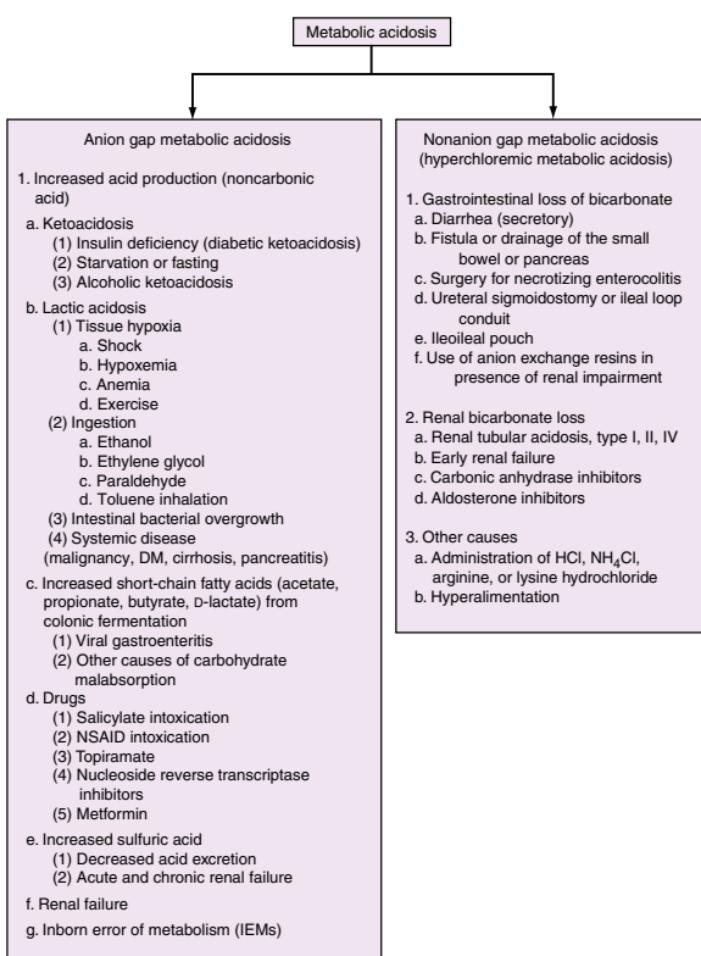
**F. Determine the likely etiology**

Check for appropriate compensation

**G. If there is not appropriate compensation, consider an additional acid-base derangement (Fig. 11.2)**

**FIGURE 11.2**

(A and B) Etiology of acid-base disturbances. *BPD*, bronchopulmonary dysplasia; *CAH*, congenital adrenal hyperplasia; *CNS*, central nervous system; *COPD*, chronic obstructive pulmonary disease; *EAST*, epilepsy, ataxia, sensorineural hearing loss, and tubulopathy; *NSAID*, nonsteroidal antiinflammatory drug.

**B****FIGURE 11.2, cont'd****REFERENCES**

A complete list of references can be found online at [www.expertconsult.com](http://www.expertconsult.com).

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# Chapter 12

## Gastroenterology

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 See additional content on Expert Consult

### I. GASTROINTESTINAL EMERGENCIES

#### A. Gastrointestinal Bleeding

1. **Presentation:** Blood loss from the gastrointestinal (GI) tract occurs in four ways: hematemesis, hematochezia, melena, and occult bleeding.
2. **Differential diagnosis of GI bleeding:** [Table 12.1](#)
3. **Diagnosis/Management**
  - a. Assess airway, breathing, circulation, and hemodynamic stability.
  - b. Perform full physical exam, verify bleeding with rectal examination, and testing of stool or emesis for occult blood. Notable exam findings include abdominal tenderness, guarding, rebound, hepatosplenomegaly, perianal skin tags, or fissures.
  - c. Obtain baseline laboratory tests. Complete blood cell count (CBC), coagulation studies, type and screen, reticulocyte count, complete metabolic panel (CMP), C-reactive protein (CRP), and erythrocyte sedimentation rate (ESR), and assess for disseminated intravascular coagulation (D-dimer, fibrinogen).
  - d. If concerned for hemodynamic instability, begin initial fluid resuscitation. Consider transfusion if there is continued bleeding, symptomatic anemia, and/or a hematocrit level <21%. Initiate intravenous (IV) proton pump inhibitor (PPI).
  - e. Further evaluation and therapy based on the assessment and site of bleeding:
    - (1) Upper GI Bleeding: Consider esophagogastroduodenoscopy (EGD) and testing for *Helicobacter pylori*.<sup>1</sup>
    - (2) Lower GI Bleeding: Consider abdominal radiograph, upper GI study ( $\pm$  small bowel follow-through), air-contrast barium enema, colonoscopy, Meckel scan, tagged red cell scan, computed tomography (CT), and magnetic resonance enterography (MRE). Consider stool cultures, stool ova and parasites, *Clostridium difficile* toxin, and stool calprotectin.

#### B. Acute Abdomen<sup>2</sup>

1. **Definition:** Severe abdominal pain that may require emergency surgical intervention.
2. **Differential diagnosis:** [Table 12.2](#)
3. **Diagnosis:**
  - a. **History:** Course and characterization of the pain, emesis, melena, hematochezia, diet, stool history, fever, travel history, menstrual

**TABLE 12.1****DIFFERENTIAL DIAGNOSIS OF GASTROINTESTINAL BLEEDING**

<b>Age</b>	<b>Upper Gastrointestinal Tract</b>	<b>Lower Gastrointestinal Tract</b>
Newborns (0–30 days)	Swallowed maternal blood Gastritis	Necrotizing enterocolitis Malrotation with midgut volvulus Anal fissure Hirschsprung disease
Infant (30 days–1 year)	Gastritis Esophagitis Peptic ulcer disease Pyloric stenosis	Anal fissure Allergic proctocolitis Intussusception Meckel diverticulum Lymphonodular hyperplasia Intestinal duplication Infectious colitis Hirschsprung disease
Preschool (1–5 years)	Gastritis Esophagitis Peptic ulcer disease Esophageal varices Epistaxis Mallory-Weiss tear	Juvenile polyps Lymphonodular hyperplasia Meckel diverticulum Hemolytic-uremic syndrome Henoch-Schönlein purpura Infectious colitis Anal fissure
School age and adolescence	Esophageal varices Peptic ulcer disease Epistaxis Gastritis Mallory-Weiss tear	Inflammatory bowel disease Infectious colitis Juvenile polyps Anal fissure Hemorrhoids

Modified from Pearl R. The approach to common abdominal diagnoses in infants and children. Part II. *Pediatr Clin North Am.* 1998;45:1287–1326.

**TABLE 12.2****ACUTE ABDOMINAL PAIN**

Gastrointestinal source	Appendicitis, pancreatitis, intussusception, malrotation with volvulus, inflammatory bowel disease, gastritis, bowel obstruction, mesenteric lymphadenitis, irritable bowel syndrome, abscess, hepatitis, perforated ulcer, Meckel diverticulitis, cholecystitis, choledocholithiasis, constipation, gastroenteritis, abdominal trauma, mesenteric ischemia, and abdominal migraine
Renal source	Urinary tract infection, pyelonephritis, and nephrolithiasis
Genitourinary source	Ectopic pregnancy, ovarian cyst/torsion, pelvic inflammatory disease, and testicular torsion
Oncologic source	Wilms tumor, neuroblastoma, rhabdomyosarcoma, and lymphoma
Other sources	Henoch-Schönlein purpura, pneumonia, sickle cell anemia, diabetic ketoacidosis, juvenile rheumatoid arthritis, and incarcerated hernia

**TABLE 12.3****DIFFERENTIAL DIAGNOSIS OF VOMITING**

Age	Typically Nonbilious	Typically Bilious
Newborn and infant (0 days–1 year)	Overfeeding, physiologic reflux, milk protein sensitivity, pyloric stenosis, necrotizing enterocolitis, metabolic disorder, infection (GU, respiratory, GI), esophageal/intestinal atresia/stenosis, and Hirschsprung disease	Malrotation ± volvulus, intestinal atresia/stenosis, intussusception, pancreatitis
Preschool (1–5 years)	Cyclic vomiting, infectious (GI, GU), toxin ingestion, diabetic ketoacidosis (DKA), CNS mass effect, eosinophilic esophagitis, post-tussive, peptic disease, and appendicitis	Malrotation, intussusception, incarcerated hernia, pancreatitis, intestinal dysmotility
School age and adolescence	Eating disorders, pregnancy, CNS mass effect, eosinophilic esophagitis, DKA, peptic disease, cyclic vomiting, toxins/drugs of abuse, infectious (GU, GI), and appendicitis	Peritoneal adhesions, malrotation, incarcerated hernia, pancreatitis, and intestinal dysmotility

CNS, Central nervous system; DKA, diabetic ketoacidosis; GI, gastrointestinal; GU, genitourinary.

12

history, vaginal/testicular symptoms, urinary symptoms, respiratory symptoms, and recent surgeries.

- b. **Physical Exam:** Rashes, arthritis, and jaundice. Abdominal tenderness on palpation, rebound/guarding, rigidity, masses, distention, or abnormal bowel sounds, rectal examination with stool hemoccult testing, pelvic examination (discharge, masses, adnexal/cervical motion tenderness), and genital examinations.
  - c. **Labs:** CBC, CMP, coagulation studies, lactate, type and screen, urinalysis, amylase, lipase, gonorrhea/chlamydia testing,  $\beta$ -human chorionic gonadotropin ( $\beta$ -hCG), ESR, and CRP.
  - d. **Imaging:** Two-view abdominal radiographs to assess for obstruction, constipation, free air, gallstones, and kidney stones. Consider chest radiograph to evaluate for pneumonia, abdominal/pelvic ultrasonography, and abdominal CT with contrast or magnetic resonance imaging (MRI).
4. **Management:** Ensure patient is NPO and begin IV hydration. Consider nasogastric decompression, serial abdominal examinations, surgical/gynecologic/GI evaluation, pain control, and antibiotics as indicated.

## II. CONDITIONS OF THE GASTROINTESTINAL TRACT

### A. Vomiting

1. **Definition:** Forceful oral expulsion of gastric contents can be bilious or nonbilious.
2. **Differential Diagnosis:** Table 12.3

**3. Diagnosis:**

- a. **History:** Diet, medications, timing (acute vs. chronic), exposures, character (bilious, bloody, projectile) and associated symptoms. Pay special attention to vomiting **without** concomitant diarrhea.
- b. **Physical Exam:** HEENT and neurologic exam with specific attention to mucus membranes, skin and dentition, as well as a thorough abdominal exam.
- c. **Labs:** Although not always necessary, consider CMP, CBC, UA,  $\beta$ -hCG, and lipase.
- d. **Imaging:** Plain abdominal radiograph with upright view (to rule out obstruction or free air), abdominal ultrasound (US), upper GI series. Consider neurologic imaging if indicated.

**4. Management:** Hydration. Gastric decompression if GI obstruction suspected. Antiemetic therapy can be used in the acute setting, avoid chronic use (see [Chapter 22](#) for discussion of antiemetic therapy). Consider surgical consultation if the vomiting is bilious.**B. Gastrointestinal Reflux Disease<sup>3</sup>**

1. **Definition:** Gastroesophageal reflux (GER) is physiologic passage of gastric contents into the esophagus, and gastroesophageal reflux disease (GERD) is defined as troublesome symptoms or complications of GER.
2. **Differential Diagnosis:** Dysmotility including achalasia, gastroparesis, ileus, and obstruction. Inflammatory conditions such as esophagitis, gastritis/dyspepsia, peptic ulcer disease. Anatomic abnormalities such as Zenker diverticulum, tracheoesophageal fistula, vascular ring, pyloric stenosis. Functional disorders including abdominal migraines and cyclical vomiting syndrome. Food allergies/intolerance in infants.

**3. Diagnosis:**

- a. **History:** Recurrent regurgitation, choking, vomiting, heartburn, chest pain, dysphagia, stridor or wheezing, cough, recurrent aspiration pneumonia, dental erosions, and sleep disturbances. In infants, GERD may present as irritability, weight loss, feeding refusal, or Sandifer syndrome. History is typically sufficient for diagnosis and to initiate management.
- b. **Testing:** Esophageal pH monitoring and esophageal impedance monitoring if diagnosis unclear.<sup>4</sup>

**4. Management:**

- a. **Lifestyle:** A prone or left-sided sleeping position and elevation of head of bed may improve GER symptoms in older children, but current studies for infants have been inconclusive. Infants up to 12 months should continue to sleep supine—risk of sleep-related infant death far outweighs benefit of prone or lateral sleeping in GERD. After feeds, infants should be kept upright and a trial of smaller more frequent feeds may be beneficial. Avoidance of second-hand smoke exposure.
- b. **Diet:** Milk-thickening agents can be beneficial for symptom relief. If severe and unresponsive to conservative management, consider 2- to 4-week trial of extensively hydrolyzed protein formula in infants

or elimination of cow's milk in maternal diet to eliminate milk protein sensitivity as a cause of unexplained vomiting.

- c. **Pharmacotherapy:** Medication is not recommended for "happy spitters" or infants with uncomplicated GER. Both PPIs and H2 receptor antagonists (H2RAs) are effective in relieving symptoms and promoting mucosal healing.<sup>5</sup> There is insufficient evidence to support routine use of prokinetic therapies (metoclopramide and erythromycin).

### C. Eosinophilic Esophagitis<sup>6,7</sup>

1. **Definition:** A chronic, immune/antigen-mediated disease characterized by symptoms of esophageal dysfunction with  $\geq 15$  eosinophils/high-power field (hpf) on esophageal biopsy.
2. **Diagnosis:**
  - a. **History:** Dysphagia, food impaction, chest pain, food refusal or intolerance, GER symptoms, emesis, abdominal pain, and failure to thrive. Majority of patients with EoE have concurrent atopic disorder.
  - b. **Diagnosis:** EGD with esophageal biopsies demonstrating at least 15 eos/hpf histologically with chronic symptoms of esophageal dysfunction; Must evaluate for other causes or contributions to esophageal eosinophilia. Importantly histologic evidence without clinical correlation is not diagnostic. Per the AGREE conference, a PPI trial is no longer needed for diagnosis. Consider obtaining allergy testing (see Chapter 15).
3. **Management<sup>8</sup>:**
  - a. **Dietary therapy:** 6-food elimination diet (milk, wheat, eggs, soy, peanuts/tree nuts, seafood), elemental diet, or targeted elimination diet determined by allergy testing.
  - b. **Pharmacotherapy:** Topical swallowed steroids delivered via inhaler are preferred as first line therapy to induce remission with limited side effects (6- to 8-week course of fluticasone or budesonide metered-dose inhaler administered orally **without** a spacer). PPI therapy can also be trialed for initial treatment. Systemic steroids for short-term use (e.g., dysphagia leading to dehydration or weight loss). No current evidence to support routine use of biologics.
  - c. **Complications:** Symptomatic strictures requiring esophageal dilation.

### D. Celiac Disease<sup>9</sup>

1. **Definition:** An immune-mediated inflammatory enteropathy caused by sensitivity to dietary gluten and related proteins (wheat, barley, and rye) in genetically susceptible individuals.
2. **Diagnosis:**
  - a. **History:** Presentation can be variable, and some patients are asymptomatic. Most common symptoms include diarrhea, vomiting, abdominal pain, constipation, distention, and failure to thrive. Non-GI symptoms include rash (dermatitis herpetiformis), osteoporosis, short stature, delayed puberty, and iron deficiency anemia that is resistant to oral iron. Increased occurrence in children with autoimmune disorders, Down syndrome, Turner syndrome, William syndrome, immunoglobulin A (IgA) deficiency, and in first-degree relatives of those with celiac disease.

- b. **Labs:** First line screening is IgA antibody to human recombinant tissue transglutaminase (TTG) and serum IgA. If known selective IgA deficiency with symptoms suggestive of celiac disease, testing with TTG IgG is recommended. CBC, iron studies, hepatic function panel, thyroid tests, calcium, and vitamin D are recommended. Additional antibody testing may be necessary for inconclusive clinical scenarios.
  - c. **Procedures:** Biopsy is “gold standard” for diagnosis. Intestinal biopsies showing villous atrophy supports diagnosis. Results dependent on adequate consumption of gluten prior to testing; ensure 6 to 8 weeks of gluten ingestion prior to endoscopy.
3. **Management:** Lifetime, gluten-free diet. Annual screening with TTG is recommended to monitor adherence to diet.
4. **Complications:** More often seen in adulthood but at risk for vitamin deficiencies and other autoimmune disorders. Higher risk of non-Hodgkin lymphoma, specifically enteropathy associated T-cell lymphoma.

### E. Inflammatory Bowel Disease (EoE)<sup>10,11</sup>

- 1. **Classification:**
  - a. **Crohn disease:** Transmural inflammatory process affecting any segment of the GI tract, most commonly terminal ileum. Commonly presents with abdominal pain, weight loss, diarrhea, and poor growth.
  - b. **Ulcerative colitis (UC):** Chronic, relapsing, inflammatory disease of the colon and rectum. Commonly presents with rectal bleeding and diarrhea.
- 2. **Diagnosis:**
  - a. **History:** Abdominal pain, weight loss, diarrhea, lethargy, nausea, vomiting, malnutrition, psychiatric symptoms, arthropathy, and rashes. Family history, exposure to infectious agents, or antibiotic treatment.
  - b. **Physical Exam:** Stomatitis, perianal skin tags, fissures, and fistulas. Assessment of hydration and nutritional status. Fever, orthostasis, tachycardia, abdominal tenderness, distention, or masses suggests moderate to severe disease and need for hospitalization.
  - c. **Labs:** CBC, CMP, ESR, CRP. Fecal calprotectin has been shown to be elevated in inflammatory bowel disease (IBD) and may serve as a sensitive, noninvasive test.<sup>12</sup> IBD often associated with anemia, hypoalbuminemia, thrombocytosis, and elevated inflammatory markers. Stool studies to exclude infectious process are necessary.
  - d. **Imaging:** MRE is the preferred imaging modality for diagnosis of pediatric IBD due to high diagnostic accuracy and no radiation exposure. CT and fluoroscopy are other alternative strategies if MRE unavailable.
  - e. **Procedures:** Diagnostic endoscopy with biopsies used to confirm diagnosis.

### 3. Management<sup>13-16</sup>:

#### a. Induction of remission:

- (1) Crohn: Exclusive enteral formula-based nutrition (80%–100% caloric need by liquid formula), 5-aminosalicylates, antitumor necrosis factor (TNF) agents, and, if indicated, antibiotics or surgery. Corticosteroids can be used if necessary.
- (2) UC: Corticosteroids, 5-aminosalicylates, TNF agents, and if indicated, antibiotics or surgery. Therapy guided by severity of illness.

#### b. Maintenance of remission:

Immunosuppression includes thiopurines, methotrexate, cyclosporine, tacrolimus, and anti-TNF monoclonal antibodies. Avoid prolonged steroid use.

#### c. Other:

Surgical intervention indicated only after medical management has failed in both Crohn's disease and UC. In Crohn disease, surgery is indicated for localized disease (strictures), abscess, or disease refractory to medical management.

## F. Constipation<sup>17</sup>

Normal stooling patterns by age: Infants 0 to 3 months, 2 to 3 bowel movements/day (breastfed infants may stool after every feed or go 5 to 7 days with no stool); 6 to 12 months, 1.8/day; 1 to 3 years, 1.4/day; >3 years, 1/day. If an exclusively breastfed <1 month old is not stooling regularly, it may be a sign of insufficient milk intake; monitor weight gain.

### 1. Definitions:

- a. **Constipation:** Delay or difficulty in defecation for 2 or more weeks. Functional causes of constipation are the most common. History and physical exam are often sufficient for diagnosis.
  - (1) Functional: Consider Rome IV Criteria ([Table EC 12.A](#))
  - (2) Nonfunctional: See [Table 12.4](#) for differential diagnosis.

### 2. Diagnosis:

- a. **History:** Age of onset, toilet training experience, frequency/consistency/size of stools, pain or bleeding with defecation, presence of abdominal pain, soiling of underwear, stool-withholding behavior, change in appetite, abdominal distention, allergies, dietary history, medications, developmental history, psychosocial history. Refer to Bristol Stool Form Scale for classification of stool history ([Fig. 12.1](#)). Delayed meconium, poor weight gain or weight loss, anorexia, nausea, vomiting, and family history (e.g., thyroid disorders, cystic fibrosis) would warrant further evaluation for nonfunctional causes.
- b. **Physical Exam:** External perineum, perianal examination. Fecal impaction may be palpated on abdominal or digital rectal examination. Plain abdominal single view radiography can be considered when physical examination is unreliable.

### 3. Management of functional constipation: [Box 12.1](#) and [Table EC 12.B](#).

#### a. Disimpaction:

- (1) Oral/Nasogastric Approach: Polyethylene glycol (PEG) solutions are effective for initial disimpaction. May also use other osmotic laxatives.

**TABLE EC 12.A****ROME CRITERIA FOR FUNCTIONAL CONSTIPATION**

In the absence of organic pathology, must include two or more of the following occurring at least once per week for a minimum of 1 month with insufficient criteria for a diagnosis of irritable bowel syndrome:

1.  $\leq 2$  defecations in the toilet per week in child of developmental age of at least 4 years
2. At least 1 episodes of fecal incontinence per week
3. History of retentive posturing or excessive volitional stool retention
4. History of painful or hard bowel movements
5. Presence of a large fecal mass in the rectum
6. History of large diameter stools that can obstruct the toilet

Modified from Constipation Guideline Committee of the North American Society for Pediatric Gastroenterology, Hepatology and Nutrition. Evaluation and treatment of constipation in infants and children: Evidence-Based Recommendations from ESPGHAN and NASPCHAN. *J Pediatr Gastroenterol Nutr.* 2014;58(2):261 and Rome IV Criteria.

TABLE 12.4

DIFFERENTIAL DIAGNOSIS OF CONSTIPATION<sup>a</sup>

Anatomic malformations	Anal stenosis, anterior displaced anus, imperforate anus, and pelvic mass (e.g., sacral teratoma)
Metabolic and gastrointestinal	Cystic fibrosis, diabetes mellitus, gluten enteropathy, hypercalcemia, hypokalemia, hypothyroidism, and multiple endocrine neoplasia type 2B
Neuropathic conditions	Neurofibromatosis, spinal cord abnormalities, spinal cord trauma, static encephalopathy, and tethered cord
Intestinal nerve or muscle disorders	Hirschsprung disease, intestinal neuronal dysplasia, visceral myopathies, and visceral neuropathies
Abnormal abdominal musculature	Down syndrome, gastroschisis, and prune belly
Connective tissue disorders	Ehlers-Danlos syndrome, scleroderma, and systemic lupus erythematosus
Drugs	Antacids, anticholinergics, antidepressants, antihypertensives, opiates, phenobarbital, sucralfate, and sympathomimetics
Other	Botulism, cow's milk protein intolerance, heavy metal ingestion (lead), and vitamin D intoxication

<sup>a</sup>Remember that functional constipation remains the most common cause.

Constipation Guideline Committee of the North American Society for Pediatric Gastroenterology, Hepatology, and Nutrition. Evaluation and treatment of constipation in infants and children: Evidence-Based Recommendations from ESPGHAN and NASPGHAN. *J Pediatr Gastroenterol Nutr.* 2014;58(2):258–274.

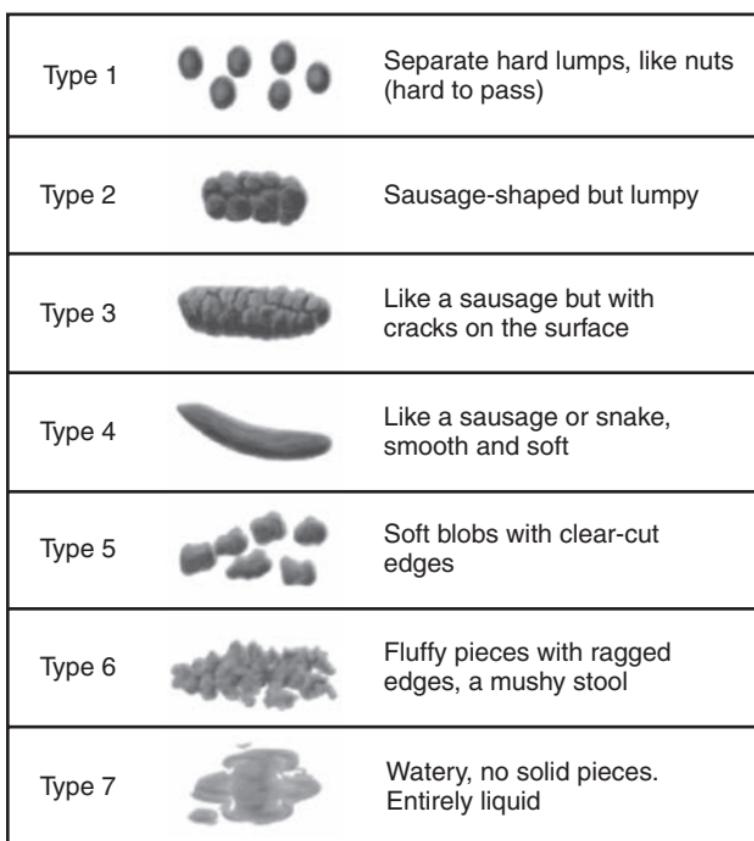


FIGURE 12.1

Bristol Stool Form Scale. (From Campbell-Walsh-Wein Urology. 12th ed. Philadelphia: Elsevier; 2020, Fig. 36.1.)

**BOX 12.1****MANAGEMENT OF CONSTIPATION****HOME CLEANOUT INSTRUCTIONS**

- Step 1: Take a stimulant laxative (bisacodyl, senna) with 8 oz of liquid, as per dosing instructions below. This should be done 6 hr prior to intended effect.
- Step 2: Drink polyethylene glycol (PEG). Mix with water or another clear noncarbonated liquid. Drink full amount in 2 hr. See below for dosing instructions.
- Step 3: 1–2 hr after finishing PEG, should begin passing formed/thick brown stool. The stool should become thinner and clearer stooling continues.
- Step 4: If not stooling or passing very thick stools 4 hr after the PEG is finished, drink 1 capful of PEG in 8 oz of liquid every hour until stools are clear.
- Step 5: Cleanout is finished when stool is mostly clear with very little sand-like material mixed in. Proceed to maintenance instructions below.

**DOSING INSTRUCTIONS**

Weight	Polyethylene Glycol (PEG) Dose	Stimulant Laxative Recommendation
8–10 kg	Mix 2.5 capfuls of PEG in 8 oz of clear drink	<2 years old: No stimulant laxative use
10.1–15 kg	Mix 3.5 capfuls of PEG in 16 oz of clear drink	2 years to <3 years old: Chewable senna (chocolate squares) <sup>a</sup>
15.1–20 kg	Mix 5 capfuls of PEG in 20 oz of clear drink	≥3 years old: Oral chewable senna (chocolate squares) until child can swallow pills, then oral bisacodyl laxative <sup>a</sup>
20.1–25 kg	Mix 6 capfuls of PEG in 24 oz of clear drink	
25.1–30 kg	Mix 7 capfuls of PEG in 28 oz of clear drink	
30.1–40 kg	Mix 9.5 capfuls of PEG in 40 oz of clear drink	
40.1–50 kg	Mix 12 capfuls of PEG in 48 oz of clear drink	
50.1 kg or more	Mix 14 capfuls of PEG in 56 oz of clear drink	

**DAILY MAINTENANCE THERAPY**

The day after colon cleanse, the patient should begin taking maintenance daily PEG for continued management of constipation.

Advise patient/family to mix PEG in clear noncarbonated drink or water at least once daily. See formulary for dosing. Advise to drink the entire solution in 30 min or less for it to work well. It is best to give the PEG after school and before dinner. Do not give PEG right before bedtime.

The goal of daily maintenance PEG is for the child to have 1 or 2 soft and easily passable bowel movements every day.

Advise to have child to sit on the toilet after every meal or whenever they feel the need to stool.

<sup>a</sup>See Formulary for dosing recommendations.

Modified from handout given to patients who visit the Johns Hopkins Children's Center Pediatric Chronic Constipation Center, as an example of constipation management; variations are found at other institutions.

**TABLE EC 12.B****PHARMACOLOGIC MANAGEMENT OF CONSTIPATION****OSMOTIC LAXATIVES**

Polyethylene Glycol (PEG)—oral	First line for disimpaction and maintenance
Lactulose—oral	If PEG not available, best and safest alternative (if >1 year of age)
Magnesium Hydroxide (Milk of Magnesia)—oral	

Sodium Phosphate—oral/enema	Risk of acute phosphate nephropathy
Glycerin—suppository/enema	Should not be used in children <2 years
	Suppository may be used in infants <1 year old

**STIMULANT LAXATIVES**

Bisacodyl—oral/enema/suppository	
Senna—oral	

**STOOL SOFTENERS**

Mineral Oil—oral/enema	
Sodium Docusate—oral/enema	

Modified from Management of Functional Constipation in Children: Therapy in Practice. *Paediatr Drugs.*

2015;17(5):349–360.

- (2) Rectal approach: Saline or mineral oil enemas effective. Avoid enemas in infants, glycerin suppositories may be used in infants less than 1 year.
- b. **Maintenance therapy** (usually 3 to 12 months): Goal is to prevent recurrence.
  - (1) **Dietary changes:** Evidence supporting dietary intervention is weak; however, increased intake of fruits, vegetables, whole grains, and fluids other than milk is recommended.
  - (2) **Behavioral modifications:** Regular toilet habits with positive reinforcement. Referral to a mental health specialist for motivational or behavioral concerns if soiling is an issue.
  - (3) **Medications:** Daily PEG. Lactulose as second line treatment. The use of stimulant laxatives and stool softeners may also be considered. Avoid prolonged use of stimulant laxatives. Discontinue therapy gradually only after return of regular bowel movements with good evacuation. Evidence does not support use of probiotics.
- c. **Special considerations in infants aged <1 year:** 2 to 4 oz of 100% fruit juice (e.g., prune or pear) recommended in younger infants. Glycerin suppositories may be useful. While use is off label, PEG is routinely used in children <1 year of age. Avoid mineral oil, stimulant laxatives, and phosphate enemas.

#### G. Diarrhea<sup>18</sup>

- 1. **Definition:** Acute diarrhea is more than three loose or watery stools per day. Chronic diarrhea is diarrhea lasting more than 2 to 4 weeks.
- 2. **Pathogenesis:** It can be infectious or malabsorptive with an osmotic or secretory mechanism.
  - a. **Osmotic diarrhea:** Water is drawn into intestinal lumen by maldigested osmotic compounds, as seen in celiac disease, pancreatic disease, or lactose intolerance. Stool volume depends on diet and decreases with fasting (stool osmolar gap  $\geq 100 \text{ mOsm/kg}$ ).
  - b. **Secretory diarrhea:** Water accompanies secreted or unabsorbed electrolytes into the intestinal lumen (e.g., excessive secretion of chloride ions caused by cholera toxin). Stool volume is increased and does not vary with diet (stool osmolar gap  $< 50 \text{ mOsm/kg}$ ).
  - c. Stool osmolar gap: The standard value is 290 mOsm/kg.<sup>19</sup>

$$\text{Stool osmolar gap} = \\ \text{Stool Osm} - \{2 \times [\text{stool (Na) mEq/L} + \text{stool (K) mEq/L}]\}$$

#### 3. Differential Diagnosis: Table 12.5

#### 4. Diagnosis:

- a. **History:** acute vs. chronic, travel history, recent antibiotic use, and immune status.
- b. **Labs:** CMP, CBC, stool hemoccult testing, stool culture, *C. difficile* toxin, ova and parasites, and viral antigens (see Chapter 17 for common bacterial and viral pathogens).

**TABLE 12.5****DIFFERENTIAL DIAGNOSIS OF COMMON CAUSES OF DIARRHEA**

Diagnosis	Major Clinical Features
Infectious colitis (viral, bacterial, protozoal)	Blood or mucous in stool, possible exposure history (e.g., travel)
Lactose malabsorption	Bloating, flatulence, abdominal pain, and elevated breath hydrogen concentration postlactose ingestion
Small bowel bacterial overgrowth	Abdominal discomfort and increased risk if ileocecal valve removed
Irritable bowel syndrome	Constipation and/or diarrhea and absence of laboratory or imaging findings
Allergic enteropathy	Growth failure, hypoalbuminemia, anemia, and may have elevated serum IgE
Hirschsprung disease	Distended abdomen, abnormal barium enema, absent ganglion cells on rectal biopsy
Cystic fibrosis	Decreased fecal elastase, steatorrhea, and poor growth
IBD and celiac disease	See sections III.D and III.E
Other: Hyperthyroidism, UTI, and encopresis	Dependent on etiology

IBD, Inflammatory bowel disease; IgE, immunoglobulin E; UTI, urinary tract infection.

Modified from Zella GC, Israel EJ. Chronic Diarrhea in Children. *Pediatrics in Review*. 2012;33(5):207–218.

## 5. Management

- Oral rehydration therapy (ORT)**<sup>20</sup>: Enteral hydration has proven superior in reducing the length of hospital stay and adverse events.<sup>21</sup> Parenteral hydration is indicated in severe dehydration, hemodynamic instability, or failure of ORT.
- Diet:** Restart regular diet as soon as tolerated.
- Pharmacotherapy:** No supporting evidence for use of nonspecific antidiarrheal agents, antimotility agents (e.g., loperamide), antisecretory drugs, and toxin binders (e.g., cholestyramine). Consider evidence-based antimicrobial therapy for infectious diarrhea (see Chapter 17). If malabsorptive (e.g., celiac disease or IBD), therapy should be tailored to disease process.
- Probiotics**<sup>22</sup>: Evidence supporting use of probiotics is limited; however, their efficacy has been demonstrated in the following circumstances: antibiotic-associated diarrhea, mild to moderate acute diarrhea, *C. difficile* diarrhea (severe recurrent disease only), hepatic encephalopathy, the prevention of atopic dermatitis, and possibly preventing necrotizing enterocolitis in premature infants.<sup>23</sup>

## III. CONDITIONS OF THE LIVER

### A. Liver Laboratory Studies: Table 12.6

- Synthetic/Metabolic function:** Albumin, prealbumin, international normalized ratio (INR), activated partial thromboplastin time (aPTT), cholesterol levels, bilirubin, and ammonia.

**TABLE 12.6****LIVER LABORATORY TESTS**

Enzyme	Source	Increased	Decreased	Comments
AST/ALT	Liver, heart, skeletal muscle, pancreas, RBCs, and kidney	Hepatocellular injury, rhabdomyolysis, muscular dystrophy, hemolysis, and liver cancer	Vitamin B <sub>6</sub> deficiency and uremia	ALT more specific than AST for liver, AST > ALT in hemolysis
Alkaline phosphatase	Osteoblasts, liver, small intestine, kidney, and placenta	Hepatocellular injury, bone growth, disease, trauma, pregnancy, and familial	Low phosphate, Wilson disease, zinc deficiency, hypothyroidism, and pernicious anemia	Highest in cholestatic conditions; must be differentiated from bone source
GGT	Renal tubules, bile ducts, pancreas, small intestine, and brain	Cholestasis, newborn period, and induced by drugs	Estrogen therapy, artificially low in hyperbilirubinemia	Not found in bone, increased in 90% of primary liver disease, specific for hepatobiliary disease in nonpregnant patient
Ammonia	Bowel flora and protein metabolism	Hepatic disease secondary to urea cycle dysfunction, hemodialysis, valproic acid therapy, urea cycle enzyme deficiency, organic academia, and carnitine deficiency		Converted to urea in liver

AST/ALT, Aspartate aminotransferase/alanine aminotransferase; GGT,  $\gamma$ -glutamyl transpeptidase; RBCs, red blood cells.

**TABLE 12.7****DIFFERENTIAL DIAGNOSIS OF ACUTE LIVER FAILURE**

Infection	Herpes simplex virus, hepatitis A, hepatitis B, adenovirus, cytomegalovirus, Epstein-Barr virus, enterovirus, human herpes virus 6, parvovirus B19, and Dengue fever
Vascular	Budd-Chiari syndrome, portal vein thrombosis, venoocclusive disease, and ischemic hepatitis
Inherited/Metabolic	Wilson disease, mitochondrial, tyrosinemia, galactosemia, hemochromatosis, fatty acid oxidation defect, and iron storage disease
Immune Dysregulation	Natural killer cell dysfunction (hemophagocytic lymphohistiocytosis), autoimmune, and macrophage activation syndrome
Drugs/Toxins	Acetaminophen, anticonvulsants, and chemotherapy
Other	Idiopathic and cancer/leukemia

- Liver cell injury:** aspartate aminotransferase (AST), alanine aminotransferase (ALT), and lactate dehydrogenase.
- Biliary system:** bilirubin (total and direct), urobilinogen,  $\gamma$ -glutamyltransferase, and alkaline phosphatase.

**B. Acute Liver Failure<sup>24,25</sup>**

- Definition:** Laboratory evidence of liver injury with no known history of chronic liver disease, the presence of coagulopathy not corrected by vitamin K administration, and an INR >1.5 if patient has encephalopathy or >2.0 if patient does not have encephalopathy.
- Differential Diagnosis:** [Table 12.7](#)
- Diagnosis:**
  - History:** Fatigue, nausea, vomiting, irritability, confusion, drowsiness, skin changes, medications, ingestion, illicit drug use, family history, developmental delay, transfusion history.
  - Physical Exam:** Neurologic status, skin exam, hepatosplenomegaly, nutritional status, growth, bruising, petechiae. Slit lamp exam if concern for Wilson disease. Findings of chronic liver disease include clubbing, palmar erythema, cutaneous xanthoma, ascites, and prominent abdominal vessels.
  - Labs:** Liver synthetic/metabolic function, liver cell injury, and biliary system tests (see earlier). BMP, magnesium, phosphorus, CBC with peripheral smear, reticulocyte count, ammonia, lipase. Factors V, VII (depleted first in ALF), VIII, and fibrinogen. A urine toxicology screen and a serum acetaminophen level should be obtained (see [Chapter 3](#)). Viral hepatitis studies, autoantibodies, and evaluation for metabolic syndromes must be considered.

**NOTE:** See [Chapter 17](#) for interpretation of serologic markers of hepatitis B.

- d. **Imaging:** Abdominal US with Doppler flow. Consider head CT scan to exclude hemorrhage/edema, and chest radiography.
- e. **Procedures:** Liver biopsy
- f. **Management:** Evaluate for underlying cause. Consider intensive care unit (ICU) level care with close monitoring of mental status, fluid balance, metabolic disturbances, hepatorenal syndrome, sepsis, and coagulopathies. Cerebral edema is life-threatening and may require intracranial pressure monitoring. Consider liver transplant when indicated.

#### C. Nonalcoholic Fatty Liver Disease<sup>26</sup>

- 1. **Definition:** Chronic liver disease from excessive fat accumulation in the liver, often secondary to insulin resistance and obesity. Most common liver disease in children in the United States.
- 2. **Diagnosis:** Screen between 9 and 11 years for obese children and overweight children with risk factors. ALT is the recommended test. If ALT persistently elevated >2 times upper limit of normal for >3 months, further evaluation is warranted. Must exclude alternative etiologies.
- 3. **Management:** Extensive lifestyle modifications, well-balanced healthy diet. No medications have proven benefit. Bariatric surgery can be considered if severe comorbidities. Screen for diabetes and other comorbid conditions.

#### D. Hyperbilirubinemia<sup>27-29</sup>

- 1. **Definition:** Bilirubin is the product of hemoglobin metabolism. There are two forms: direct (conjugated) and indirect (unconjugated). Hyperbilirubinemia is usually the result of increased hemoglobin load, reduced hepatic uptake, reduced hepatic conjugation, or decreased excretion. Direct hyperbilirubinemia is defined as a direct bilirubin >20% of the total bilirubin or a direct bilirubin of >2 mg/dL.
- 2. **Differential Diagnosis:** [Table 12.8](#)
- 3. **Management:** Dependent upon etiology. Evaluation and diagnosis should be guided by history; however, liver laboratory studies (see earlier) and USs are warranted in many patients. Refer to [Chapter 18](#) for evaluation and treatment of neonatal hyperbilirubinemia.

### IV. PANCREATITIS<sup>30-32</sup>

**Definition:** Inflammatory disease of the pancreas.

#### A. Acute Pancreatitis<sup>33</sup>

- 1. **Diagnosis:**
  - a. **History:** Abdominal pain, irritability, epigastric tenderness, nausea and vomiting. Multiple etiologies ([Table 12.9](#)). Per INSPIRE criteria, diagnosis of acute pancreatitis requires at least two of the following:
    - (1) Abdominal pain compatible with acute pancreatitis
    - (2) Serum amylase and/or lipase values >3 times upper limit of normal
    - (3) Imaging findings consistent with acute pancreatitis

**TABLE 12.8****DIFFERENTIAL DIAGNOSIS OF HYPERBILIRUBINEMIA**

<b>INDIRECT HYPERBILIRUBINEMIA</b>	
Transient neonatal jaundice	Breast milk jaundice and physiologic jaundice
Hemolytic disorders	Polycythemia and reabsorption of extravascular blood Autoimmune disease, blood group incompatibility, hemoglobinopathies, microangiopathies, red cell enzyme deficiencies, and red cell membrane disorders
Enteropathic recirculation	Cystic fibrosis, Hirschsprung disease, ileal atresia, and pyloric stenosis
Disorders of bilirubin metabolism	Acidosis, Crigler-Najjar syndrome, Gilbert syndrome, hypothyroidism, and hypoxia
Miscellaneous	Dehydration, drugs, hypoalbuminemia, sepsis, and panhypopituitarism
<b>DIRECT HYPERBILIRUBINEMIA</b>	
Biliary obstruction	Biliary atresia, choledochal cyst, fibrosing pancreatitis, gallstones or biliary sludge, inspissated bile syndrome, neoplasm, and primary sclerosing cholangitis
Infection	Cholangitis, cytomegalovirus, adenovirus, enterovirus, Epstein-Barr virus, herpes simplex virus, histoplasmosis, human immunodeficiency virus, leptospirosis, liver abscess, sepsis, syphilis, rubella, toxocariasis, toxoplasmosis, tuberculosis, urinary tract infection, varicella-zoster virus, and viral hepatitis
Genetic/metabolic disorders	$\alpha_1$ -Antitrypsin deficiency, Alagille syndrome, Caroli disease, cystic fibrosis, Dubin-Johnson syndrome, galactokinase deficiency, galactosemia, glycogen storage disease, hereditary fructose intolerance, hypothyroidism, Niemann-Pick disease, progressive familial intrahepatic cholestasis (PFIC), Rotor syndrome, tyrosinemia, and Wilson disease
Chromosomal abnormalities	Trisomy 18, trisomy 21, and Turner syndrome
Drugs	Acetaminophen, aspirin, erythromycin, ethanol, iron, isoniazid, methotrexate, parenteral nutrition, oxacillin, rifampin, steroids, sulfonamides, tetracycline, and vitamin A
Miscellaneous	Neonatal hepatitis syndrome, parenteral alimentation, and Reye syndrome

- b. **Labs:** CMP, GGT, CBC, amylase, lipase, calcium, and triglycerides.
- c. **Imaging:** Transabdominal US recommended. CT or MRI reserved for more complicated cases depending on etiology.

**2. Management:**

- a. **Analgesia:** Acetaminophen or NSAIDs as first line therapy; opiates for refractory pain.
- b. **Nutrition:** Aggressive IV fluid hydration within initial 48 hours. Early enteral feeding recommended (within 72 hours of presentation and hemodynamically stable) and associated with shorter hospitalization and decreases comorbidity.

**TABLE 12.9**  
**CONDITIONS ASSOCIATED WITH ACUTE PANCREATITIS**

<b>SYSTEMIC DISEASES</b>	
Infections	Coxsackie, CMV, cryptosporidium, EBV, hepatitis, influenza A or B, leptospirosis, mycoplasma, mumps, rubella, typhoid fever, and varicella
Inflammatory and vasculitic disorders	Collagen vascular diseases, hemolytic uremic syndrome, Henoch-Schönlein purpura, IBD, and Kawasaki disease
Sepsis/peritonitis/shock	
<b>IDIOPATHIC (UP TO 25% OF CASES)</b>	
<b>MECHANICAL/STRUCTURAL</b>	
Trauma	Blunt trauma, child abuse, and ERCP
Anatomic anomalies	Annular pancreas, choledochal cyst, pancreatic divisum, stenosis, and other
Obstruction	Parasites, stones, and tumors
<b>METABOLIC AND TOXIC FACTORS</b>	
Drugs/toxins	Salicylates, cytotoxic drugs ( $\text{l}$ -asparaginase), corticosteroids, chlorothiazides, furosemide, oral contraceptives (estrogen), tetracyclines, sulfonamides, valproic acid, azathioprine, and 6-mercaptopurine
Cystic fibrosis	
Diabetes mellitus	
Hypercalcemia	
Hyperlipidemia	
Hypothermia	
Malnutrition	
Organic academia	
Renal disease	

*CMV*, Cytomegalovirus; *EBV*, Epstein-Barr virus; *ERCP*, endoscopic retrograde cholangiopancreatography; *IBD*, inflammatory bowel disease.

- c. **Complications:** Multiorgan dysfunction, shock, pseudocysts, fluid collections, and necrosis. Antibiotics reserved for infected necrosis. Surgical consult as indicated.

## B. Chronic Pancreatitis<sup>34,35</sup>

### 1. Diagnosis:

- a. **History:** Abdominal pain consistent with pancreatic origin, pancreatic insufficiency; plus consistent imaging findings or biopsy with histopathologic features. Must be distinguished from acute recurrent pancreatitis (ARP), which is defined as at least two distinct episodes of pancreatitis with complete resolution of pain or normalization of laboratory levels.
- b. **Labs:** Same as acute pancreatitis. Normal amylase/lipase does not exclude diagnosis of chronic pancreatitis or ARP. Fecal elastase to screen for exocrine function and fat-soluble vitamins assessment. Consider genetic testing.

**TABLE 12.10****PROPOSED ETIOLOGIES OF CHRONIC PANCREATITIS IN CHILDHOOD**

Calcific	Cystic fibrosis, hereditary pancreatitis (e.g., PRSS1 and SPINK1 mutations), hypercalcemia, hyperlipidemia, idiopathic, and juvenile tropical pancreatitis
Obstructive (noncalcific)	Congenital anomalies, idiopathic fibrosing pancreatitis, renal disease, sclerosing cholangitis, sphincter of Oddi dysfunction, and trauma

Modified from Robertson MA. Pancreatitis. In: Walker WA et al, eds. *Pediatric Gastrointestinal Disease*. 3rd ed. New York: BC Decker; 2000:1321–1344; Werlin SL. Pancreatitis. In: McMillan JA et al, eds. *Oski's Pediatrics*. Philadelphia: Lippincott Williams & Wilkins; 2006:2010–2012.

c. **Imaging:** Repeat imaging recommended with US and/or MRCP.

**Note:** See Table 12.10 for proposed etiologies of chronic pancreatitis in childhood.

2. **Management:** (For acute exacerbations) same as management of acute pancreatitis. Maintenance to focus on nonmedication strategies, adequate nutrition for growth, nonopioids and planned opioids.

**V. WEB RESOURCES**

- North American Society for Pediatric Gastroenterology, Hepatology, and Nutrition: [www.naspghan.org](http://www.naspghan.org)
- Children's Digestive Health Information for Kids and Parents: [www.gikids.org](http://www.gikids.org)
- Celiac Disease Foundation: [clinical.celiac.org](http://clinical.celiac.org)
- Rome Foundation for Diagnosis and Treatment of Functional Gastrointestinal Disorders: [www.theromefoundation.org](http://www.theromefoundation.org)

12

**REFERENCES**

A complete list of references can be found online at [www.expertconsult.com](http://www.expertconsult.com).