CHAPTER 8

Anesthesiology

More than 50 percent of all operations performed in children are performed in ambulatory or same-day surgery settings. The success of outpatient pediatric surgery depends in part on the adequate preparation of both child and parents. Much of the preoperative and immediate postoperative care of these children, previously provided in the hospital by the anesthesiologist, is now being provided by the child's surgeon or pediatrician in an outpatient setting.

Infants and children who undergo anesthesia for orthopaedic procedures represent all stages of development, from birth to adolescence, and cover the health spectrum from an otherwise healthy child to the very sick child with multiple congenital anomalies, in whom operation is likely to pose special problems for the anesthesiologist (Appendix 8–1). The orthopaedic surgeon, by being aware of these potential problems, can alert the anesthesiologist.

The following section discusses those aspects of the child's history and physical examination and relevant laboratory tests that contribute to decreasing anesthetic risk.

Preoperative Evaluation of Children

The areas of primary concern in the child's history are related to respiratory, cardiovascular, neuromuscular, endocrine, hematologic, and oncologic diseases. In the physical examination the anesthesiologist is mainly interested in airway anatomy (which may suggest the possibility of a difficult intubation), the presence of stridor, wheezing (pre- and postoperative hypoxemia), murmurs, and any preexisting neurologic deficit, because anesthesia agents and medication can exacerbate the preexisting neuromuscular weakness.

Children taking seizure medication should take their morning dose with a sip of water. Because virtually all anesthetics produce cerebral vasodilation and therefore may increase intracranial pressure, children at risk of developing intracranial hypertension (e.g., those with hydrocephalus, blocked ventriculoperitoneal shunts, or brain tumors) must be identified before surgery. Any evidence of brain stem dysfunction (vocal cord paralysis, swallowing dysfunction, or aspiration) should be noted in the preoperative evaluation. In children with progressive diseases of nerve or muscle, hyperkalemia or malignant hyperthermia occur more commonly following succinylcholine administration; therefore the anesthesiologist should be informed of the nature and extent of the underlying condition.

The presence of an abnormal murmur, cyanosis, decreased exercise tolerance, poor weight gain, sweating, dedecreased femoral pulses, or a precordial heave necessitates a more thorough evaluation (hematocrit determination, electrocardiography [ECG], chest x-ray, oxygen saturation determination, and a cardiology consult).

Antibiotic prophylaxis to prevent bacterial endocarditis for children with congenital heart disease undergoing surgery is essential. (We follow the American Heart Association antibiotic regimen.) The first dose is usually given intravenously (IV) after initiation of anesthesia and before nasotracheal intubation or surgical incision. The parents must be alerted to the fact that the antibiotic prophylaxis must be continued postoperatively at home in cases of outpatient surgery.

Infants born prematurely have a significant risk of postoperative apnea that decreases as postconceptual age increases. Children with a history of bronchopulmonary dysplasia and those with obstructive sleep apnea are at greater risk for perioperative hypoxemia and acute right heart failure. Diabetic patients need to be closely evaluated by the anesthesiologist and the pediatrician jointly, and a plan for managing the diabetes must be decided on.

Patients on long-term corticosteroid therapy have suppression of the hypothalamic-pituitary-adrenal axis and therefore cannot manifest an appropriate stress response (Addisonian crisis). These children should receive prednisone, 0.5 mg/kg orally (PO), the night before, and hydrocortisone, 1 mg/kg IV, after induction of anesthesia.

In otherwise healthy children scheduled for elective surgery, routine laboratory tests, chest radiography, or urinalysis are rarely indicated. African-American children, however, who have not had a hemoglobin or hematocrit determination after 4 months of age should undergo sickle cell screening, and if they test positive, hemoglobin electrophoresis should be done to define the exact nature of the hemoglobinopathy.

Although the healthy child needs almost no preoperative laboratory testing, the situation is entirely different in children who have a history of an abnormality. For example, the hemoglobin level should be determined in the child with cardiac or sickle cell disease. A chest radiograph is helpful in a child with a history of chronic aspiration or lower airway disease. Children on digoxin therapy should have their serum sodium, potassium, and digoxin levels measured. An ECG is warranted in a child with congenital heart disease, obstructive sleep apnea, or bronchopulmonary dysplasia.

In the child with Down syndrome or diastrophic dysplasia, anteroposterior (AP) and lateral radiographs of the neck should be obtained to detect any instability of the cervical spine.

UPPER RESPIRATORY INFECTION

Upper respiratory infections (URI) are the most common illnesses affecting children under 5 years old. (The commonly reported incidence of URI in this age group is 24 percent.) Children under 1 year old have an average of 6.1 respiratory illnesses per year. Children between 1 and 5 years old have an average of 4.7 to 5.7 respiratory illnesses per year. In children with a URI there is an increased incidence of the following problems during and after surgery:

- Laryngospasm during induction of and emergence from anesthesia.
- Bronchospasm—A viral URI will initiate wheezing more commonly in children than adults, whether or not they have a history of asthma. Children under 5 years old with preexisting asthma or with respiratory syncytial virus infection are prone to develop bronchospasm.
- Coughing due to increased airway secretion and hyperreactivity. Coughing can infrequently cause silent regurgitation and aspiration.
- Reduction in oxygen saturation intra- and postoperatively due to the above factors and also due to lung atelectasis and a reduction in functional residual capacity (FRC).

Diagnosis of URI. In general, two of the following criteria must exist:

- Sore or scratchy throat
- Sneezing
- Rhinitis
- Fever (mild)
- Congestion
- Malaise
- Nonproductive cough
- Laryngitis

Sneezing and a runny nose do not necessarily indicate a URI, and when in doubt, it is helpful to ask the parents whether they feel the child has a URI. These symptoms are often due to allergic rhinitis, and one should ask whether there is a family history of allergies.

Routine surgery in the presence of URI should definitely be canceled if:

- The patient is under 12 months old
- The patient has a lower respiratory tract infection
- The patient has signs of overt viremia or bacteria

Elective surgery in patients with a suspected respiratory tract infection should be postponed for 1 to 2 weeks after cessation of symptoms for *upper* respiratory infection and at least 4 to 6 weeks after cessation of symptoms for *lower* respiratory infection (e.g., bronchiolitis, pneumonia).

ASTHMA

Medical therapy for asthma should be optimized preoperatively because children with reactive airway disease have a high likelihood of sustaining intraoperative bronchospasm, hypoxia, and hypercarbia acidosis, and their hospitalization is likely to be prolonged. In general, therapy should be escalated preoperatively for children with asthma, even if the asthma is well controlled, because many procedures routinely performed during anesthetics, such as laryngoscopy and intubation, are potential triggers for bronchospasm. For example, the child taking an inhaled beta-agonist, steroid, or oral medication on an as-needed (PRN) basis should be changed to regular administration for 3 to 5 days preoperatively. The child taking beta-agonist medication on a regular basis should have a steroid added. If the child is taking both a bronchodilator and a steroid chronically, the frequency of nebulizers should be increased or the steroid dosage increased (or both, if necessary).

A recent asthma exacerbation that required emergency admission or hospitalization within 6 weeks of surgery precludes elective surgery. The peak expiratory flow rate and FEV_1 (forced expired volume during first second) are decreased up to 6 weeks following an asthma attack. Elective surgery in asthmatic children who have a URI should be postponed for 6 weeks even if they do not have any evidence of wheezing on auscultation, as the incidence of bronchospasm increases (11-fold) in asthmatic compared with nonasthmatic children.

Corticosteroids are extremely effective in preventing perioperative wheezing. For children who have required steroid administration in the past year, those who are taking bronchodilators chronically, and those who are almost never wheeze-free, a short course of prednisone, 0.5 mg/kg PO once daily for 3 days prior to surgery and on the morning of surgery, is highly recommended.

NPO REQUIREMENTS IN INFANTS AND CHILDREN

In the hope of reducing the risk of perioperative aspiration pneumonitis, a period of fasting has become routine in the preoperative preparation of the surgical patient (Table 8–1).

TABLE 8–1 **Fasting (NPO) Guidelines for Children and Adults**

	Fasting Time (hr)	
Age	*Solids (Including Formula and Breast Milk)*	*Clear Liquids*
<6 mo	6 hr	2 hr
6–36 mo	6 hr for formula 8 hr for solids	2 hr
>36 mo	8 hr	2 hr

POSTOPERATIVE FLUID MANAGEMENT

During the immediate postoperative period, fluids are administered IV to replace

1. Maintenance fluid loss
2. Ongoing third space loss, depending on the extent of the surgical procedure (range, 1 to 10 mL/kg/hr)
3. Minor blood loss

HOURLY MAINTENANCE FLUID REQUIREMENT

Managing fluid balance is usually easier if fluid requirements are calculated on an hourly basis:

- 4 mL/kg/hr for the first 10 kg of body weight
- 2 mL/kg/hr for the next 10 kg of body weight
- 1 mL/kg/hr for any weight more than 20 kg (e.g., a 27-kg child will receive 67 mL/hr)

Sedation for the Pediatric Patient by Nonanesthesiologists

The safe sedation of children requires a network of trained personnel with common sense in selecting patients suitable for sedation, appropriate selection and dosage of drugs, ability to monitor vital signs, and airway management using appropriate equipment. Catastrophic outcomes such as seizures, respiratory arrest, and cardiac arrest have occurred in various practice settings when personnel were deficient in any one of these areas.

SEDATION AND ANALGESIA (CONSCIOUS SEDATION)

Conscious sedation allows a patient to tolerate unpleasant procedures while maintaining (1) protective reflexes, (2) a patent airway independently and continuously, and (3) the ability to respond to physical stimulation or verbal commands (e.g., "open your eyes").

DEEP SEDATION

Deep sedation is a medically controlled state of depressed consciousness or unconsciousness from which the patient is not easily aroused. It may be accompanied by a partial or complete loss of protective reflexes. The deeply sedated patient is unable to maintain a patent airway independently or to respond purposefully to physical stimulation or verbal commands.

Deep sedation can be an unintended consequence of conscious sedation. The latter term should be abolished, as it is often difficult to achieve conscious sedation without approaching deep sedation. The term "conscious sedation" has given many physicians the false hope that they can achieve satisfactory sedation for all kinds of painful and stressful procedures without adverse sequelae. Any physician who orders narcotics and sedatives for the purpose of inducing conscious sedation must be able to manage an obstructed airway or a depressed level of respiration.

TABLE 8–2 **American Society of Anesthesiologists Physical Status Classification**

Class I	A normally health patient
Class II	A patient with mild systemic disease
Class III	A patient with severe systemic disease
Class IV	A patient with severe systemic disease that is a constant threat to life
Class V	A moribund patient who is not expected to survive without the operation

PATIENT SELECTION AND EVALUATION

The relative health of the child can be readily assessed by using the same simple scale used by anesthesiologists when evaluating preoperative patients (Table 8–2). The nonanesthesiologist should restrict the choice of patient for conscious sedation to American Society of Anesthesiologists (ASA) class I and class II, and request assistance from the anesthesiology department for sedating the very sick patient (Table 8–3).

A child undergoing a procedure that is not painful (e.g., computed tomography) does not need a narcotic. Conversely, a child undergoing a painful procedure needs a narcotic or a combination of a narcotic and a sedative, but never a sedative alone.

It is of utmost importance that the child be observed in an appropriate recovery facility for an appropriate period of time prior to discharge.

TABLE 8–3 **Dosages of Drugs Commonly Used for Sedation**

Drug	Route	Dose (mg/kg)
Barbiturates		
Pentobarbital	Rectal	5–10
Benzodiazepines		
Diazepam	PO	0.1–0.3
	IV	0.1–0.3
	Rectal	0.2–0.3
Midazolam	PO	0.5–0.75
	Rectal	0.5–0.75
	Nasal	0.2–0.5
	Sublingual	0.2–0.5
	IV	0.05–0.15
	IM	0.05–0.15
Chloral hydrate	PO	50–100
Ketamine	PO	6–10
	Rectal	5–10
	IV	1–3
	IM	3–10
Opioids		
Morphine	IV	0.1–0.3
	IM	0.1–0.3
Meperidine	IV	1–3
	IM	1–3
Fentanyl	Oral transmucosal	0.015–0.030 (15–30 μg/kg)
	IV	0.001–0.005 (1–5 μg/kg) in increments of 0.5–1.0 μg/kg

Abbreviations: PO, per os; IV, intravenous; IM, intramuscular.

FIGURE 8–1 Wong-Baker FACES Pain Rating Scale. (Reproduced from Wong DL, Hockenberry-Eaton M, Wilson D, Winkelstein ML, Schwartz P: Wong's Essentials of Pediatric Nursing, ed 6, St. Louis, 2001, p. 1301. Copyrighted by Mosby, Inc. Reprinted by permission.)

Acute Pain Management

Acute pain refers to pain of short duration (usually 3 to 7 days) and is usually associated with surgery, trauma, or an acute illness. Historically, the management of pain in children has been neglected. The common misconception that neonates were unable to perceive pain and that all children were at excessive risk of respiratory depression after administration of opioids led to underdosing of children after surgery. A body of evidence now exists showing that pain activates neuroendocrine responses, which increases tissue catabolism and impairs tissue healing. Moreover, inadequate analgesia may further impair postoperative pulmonary function and increase the risk of morbidity or mortality after surgery.

ASSESSMENT OF PAIN IN CHILDREN

To treat pain effectively, the clinician must be able to assess and measure pain accurately. Because pain is a subjective experience, a self-assessment scale is always preferable to an observer's objective assessment and should be used whenever possible. Unfortunately, no fully satisfactory system has been devised for evaluating pain in preverbal or mentally impaired children, who must be assessed by an objective observer.

Self-Assessment. Simple self-assessment methods can be used in children 4 years old and older who can verbalize, and are the most reliable and effective methods of pain assessment. Younger children prefer to use the Faces Scale (Fig. 8–1).

Children 7 years old and older can frequently use a visual analogue scale or a word-graphic rating scale (Fig. 8–2).

Physiologic Measurement and Behavioral Observation. These methods of pain assessment are used in children with limited or no verbal skills. All objective rating systems rely on changes in heart rate, respiratory rate, and blood pressure, together with behavioral assessment. There is always some difficulty in separating behavior that is associated with pain from behavior that is caused by anxiety, fear, or hunger.

FIGURE 8–2 Example of a word-graphic rating scale.

TABLE 8–4 **Assessment of Pain Intensity (FLACC) Scale**

Behavior Category	0	1	2
Face	No particular expression or smiling	Occasional grimace or frown, withdrawn, disinterested	Frequent to constant clenched jaw, quivering chin
Legs	Normal position or relaxed	Uneasy, restless, tense	Kicking, or legs drawn up
Activity	Lying quietly, normal position, moves easily	Squirming, shifting back and forth, tense	Arched, rigid, or jerking
Cry	No crying—awake or asleep	Moans or whimpers, occasional complaint	Crying steadily, screams or sobs, frequent complaints
Consolability	Content, relaxed	Reassured by occasional touching, hugging, or "talking to," distractable	Difficult to console or comfort

One such scale is the FLACC (Face, Leg, Activity, Cry, Consolability) Scale (Table 8–4).

PAIN MANAGEMENT

Pain following orthopaedic surgery is often intense. Effective pain management always begins in preoperative discussions with children and their parents. Psychological preparation can favorably modify the amount of discomfort and anxiety experienced by the child. The child and parents should be informed that every effort will be made to have the child as pain free as possible, and that although some discomfort is inevitable, it will be minimized. Appropriate pain management techniques are discussed. Play therapy, both as a preoperative teaching tool and as a postoperative distraction technique, is also very helpful. A child life specialist is often enlisted for this purpose.

SYSTEMIC ANALGESIA

Nonopioid Analgesics

ACETAMINOPHEN. Acetaminophen is the nonopioid analgesic most commonly used in pediatrics. It should be used in preference to aspirin because of aspirin's greater frequency of side effects (e.g., gastritis, platelet dysfunction, and the rare but statistically significant association with Reye's syndrome). Acetaminophen is quite safe even in newborns.

Acetaminophen may be administered orally, in a dosage of 15 to 18 mg/kg every 4 hours, or rectally, in a dosage of 25 to 30 mg/kg every 4 hours.

NONSTEROIDAL ANTI-INFLAMMATORY DRUGS. NSAIDs inhibit the enzyme cyclo-oxygenase, resulting in decreased production of prostaglandin. It is believed that because prostaglandins are a major component of the inflammatory response, and because the inflammatory response contributes to pain, use

TABLE 8–5 **Commonly Used Oral Opiates**

Medication	Ingredients	Usual Dosage
Acetaminophen + codeine elixir (Tylenol with codeine elixir)	Acetaminophen, 120 mg Codeine, 12 mg/5 mL	Acetaminophen, 15 mg/kg Codeine, 1 mg/kg Dose PO every 4 hr PRN
Acetaminophen + codeine tablets (Tylenol No. 3)	Acetaminophen, 300 mg Codeine, 15 mg/tablet	1–2 tablets PO every 4 hr PRN
Morphine sulfate, oral solution	Morphine sulfate, 2 mg/mL	0.3 mg/kg PO every 4 hr PRN
Acetaminophen + oxycodone (Percocet)	Acetaminophen, 325 mg Oxycodone, 5 mg	1–2 tablets PO every 4 hr PRN
Acetaminophen + hydrocodone (Vicodin)	Acetaminophen, 500 mg/tablet Hydrocodone, 5 mg	1–2 tablets PO every 4 hr PRN
Acetaminophen + propoxyphene (Darvocet)	Acetaminophen, 650 mg Propoxyphene, 100 mg	1 tablet PO every 4 hr PRN
	Acetaminophen, 325 mg Propoxyphene, 50 mg	1–2 tablets PO every 4 hr PRN

Abbreviations: PO, per os; PRN, as needed.

of NSAIDs reduces pain. These drugs in combination with opioids will provide more effective analgesia in the short-term management of moderate to severe postoperative pain than either NSAIDs or opioids used alone.

Ketorolac may be administered IV in a dosage of 0.5 mg/kg (maximum, 30 mg/kg) every 6 hours for a maximum of 3 to 5 days. The risk of renal impairment increases if ketorolac is used longer than 3 to 5 days.

If the child is able to tolerate oral medication, ibuprofen, 8 to 10 mg/kg given four times a day, is recommended.

The side effects of NSAIDs include renal impairment (especially in patients in hypovolemic or cardiogenic shock), platelet dysfunction, a prolonged bleeding time, and gastrointestinal (GI) upset. Studies in adults have demonstrated that the prolonged bleeding time that occurs after ketorolac administration is probably of little clinical importance.

BENZODIAZEPINES. Benzodiazepines have sedative, anxiolytic, and amnesic properties but no analgesic properties. They are frequently used in conjunction with pain medication when muscle spasm is a component of pain. Benzodiazepine potentiates the respiratory depressant effects of opioids, and such combinations must be used with great caution.

Opioids. Opioids act on opioid receptors in the brain, spinal cord, and periphery (hence the rationale for administration of narcotics in joints). Four main pain receptors have thus far been identified, mu, delta, kappa, and sigma, in addition to a number of subgroups. Both endogenous substances (endorphins) and exogenous substances (opioid drugs) interact with these receptors to inhibit pain sensation.

Opioids are administered by IV, intramuscular (IM), oral, transmucosal, and transdermal routes. They have recently been added to neuraxial anesthesia administered via the epidural or spinal route.

The least reliable method of achieving a desired blood level of opioid is by oral administration, as most of the drug that is absorbed from the GI tract undergoes first-pass metabolism in the liver before reaching the opioid receptors. Commonly used oral opiates are listed in Table 8–5.

IM injections of opioids should be avoided in children because of variable and unpredictable absorption of the opioid via this route and because the fear of frequent injection may lead to the child's denying pain and refusing the much-needed analgesia.

IV bolus administration of opioids has the advantage of achieving a rapid onset of action with a high level of efficacy in terms of pain relief. The disadvantages of IV bolus administration include the relatively short duration of analgesic activity and the labor-intensive nature of this route of administration. Guidelines for administering morphine sulfate by IV bolus are given in (Table 8–6).

CONTINUOUS INTRAVENOUS INFUSION. Continuous IV infusion of an opioid (usually morphine sulfate) is used when patient-controlled analgesia (PCA) or regional anesthesia is not indicated. It is very important that a loading dose of the opioid be administered before the start of the continuous infusion. Patients on continuous infusion must be closely monitored because if excessive drug accumulates, respiratory depression will ensue. A reduction in the hourly dose must be considered if the child is to remain on continuous infusion for more than 24 hours. The hourly rate is usually reduced by 10 percent for each additional day that continuous infusion of opioid remains in effect. Guidelines for the continuous infusion of morphine sulfate are given in Table 8–7.

PATIENT-CONTROLLED ANALGESIA. PCA provides the patient with an element of control in pain management. The method is successful only after an adequate bolus injection of opioid has been given to provide adequate analgesia. Subsequently the patient will receive a predetermined dose of opioid when he or she pushes the button on the PCA device. The advantages of PCA include a high level of efficacy, fewer side effects, and high patient satisfaction. Children ages 6 to 7 years or older can use a PCA device effectively. Younger children may benefit from IV bolus or continuous infusion

TABLE 8–6 **Guidelines for IV Bolus of Morphine Sulfate**

For severe pain:	0.08 mg/kg IV every 2 hr PRN
For moderate pain:	0.05 mg/kg IV every 2 hr PRN
For mild pain:	0.025 mg/kg IV every 2 hr PRN

TABLE 8–7 **Guidelines for Continuous Infusion of Opioids (Morphine Sulfate)**

Concentration:	0.1 mg/mL
Loading dose:	0.05 mg/kg every 10 min until analgesia is achieved
Subsequent infusion:	
Ages 3–12 mo:	0.015 mg/kg/hr
Ages 1–7 yr:	0.025 mg/kg/hr

of opioids. In infants under 3 months old, great caution must be exercised in the use of opioids; titrating to effect is of paramount importance. Opioid is cleared very slowly in these infants compared with its rate of clearance in older children. In addition, there is less protein binding of opioid, resulting in higher levels of free, active opioids in the blood. Guidelines for implementing PCA are given in Table 8–8.

Regional Analgesia

Regional anesthesia for surgery and postoperative pain relief has been significantly better received in recent years. In no other specialty are the benefits of regional anesthesia more apparent than in orthopaedic surgery, where its intraoperative and postoperative advantages have been clearly documented.

Regional analgesia is an extremely effective method of pain control for most patients. Effective pain relief can be achieved with much smaller doses of narcotics and less sedation than with parenteral opioids. The addition of a very dilute local anesthetic agent will allow the patient to ambulate postoperatively. Motor blockade occurs uncommonly but has been observed even with dilute bupivacaine (0.1%); therefore, the patient must always be accompanied by a nurse when ambulating.

Contraindications to regional anesthesia include

- Sepsis
- Infection at or near the site of insertion
- Anatomic abnormalities (e.g., meningomyelocele, sacral dysgenesis)
- Coagulopathies and thrombocytopenia

TABLE 8–8 **Guidelines for Implementation of PCA (Program the PCA Pump as Follows)**

Drug:	Morphine, 1 mg/mL *or* Meperidine, 10 mg/mL
Loading dose:	Morphine, 0.05–0.1 mg/kg every 10 min until analgesia is achieved *or* Meperidine, 0.5–1 mg/kg until analgesia is achieved
PCA dose:	Morphine, 0.02–0.03 mg/kg *or* Meperidine, 0.2–0.3 mg/kg
Lockout interval or delay:	Usual range, 7–12 min
1 hr limit:	Morphine, 0.1 mg/kg *or* Meperidine, 1.0 mg/kg
1 hr limit for spinal fusion patients:	Morphine, 0.2 mg/kg *or* Meperidine, 2.0 mg/kg

It is advantageous to place the block at the beginning of the surgical procedure rather than the end. Patients given regional anesthesia are often more alert and oriented in the postanesthesia care unit, as regional blockade obviates the use of intraoperative IV opioids, and less anesthetic agent is needed to maintain anesthesia. Interruption of nociceptive pathways at the spinal cord level may prevent imprinting of painful stimuli on the sensory cortex. Hence patients who receive an intraoperative neural block may experience much less postoperative pain than those managed with general analgesia. It is well demonstrated that for certain operations, such as limb amputation, patients who have received a regional block have a decreased incidence of phantom pain.

Regional analgesia ranges from local infiltration of the wound edges to epidural or subarachnoid administration of local anesthetics and/or opioid along the neuraxis.

GENERAL RECOMMENDATIONS FOR EPIDURALS

Epidural anesthesia is the most versatile analgesic technique in children undergoing thoracic, abdominal, perineal, or lower extremity surgery. The epidural space can be approached at any level but is most frequently approached via a caudal or lumbar route. A caudal block is commonly used in younger children (6 to 7 years old, weight 20 kg or less). A lumbar epidural block is used in older children.

A catheter can be placed via the caudal or lumbar route, and a variety of local anesthetics or opioids (or combinations of both) can be administered to maintain analgesia postoperatively. Typically, 0.1% bupivacaine plus a lipophilic opioid such as fentanyl is used for operations on the lower limb; 0.1% bupivacaine with a hydrophilic opioid such as morphine sulfate or hydromorphone is used when a larger number of dermatones need to be blocked, as in scoliosis surgery. The potential side effects of opioid administration in the neuraxis include

- Respiratory depression
- Urine retention
- Nausea and vomiting
- Pruritus

The lipophilic agents (e.g., fentanyl) have less tendency to spread in rostrally, as they tend to bind to the pain receptors in the spinal cord adjacent to the site of drug administration. The hydrophilic agents (morphine sulfate and hydromorphone) tend to spread farther rostrally and can precipitate respiratory depression. The hallmarks of an impending overdose of epidural opioid are increasing sedation and decreasing depth of respiration before the respiratory rate decreases.

The most important monitor for the patient receiving epidural opioid is not an electronic device but the nurse who uses a stethoscope to detect a reduction in the depth of respiration and level of sedation. Therefore, educating the nursing staff is of critical importance in creating a safe and effective pain service.

Recommendations

1. Place epidurals at the start of the procedure whenever possible.
2. Give no more than 2 mg/kg of bupivacaine as an initial loading dose after a standard test dose of lidocaine 1.5% with epinephrine 1:200,000, 0.1 mL/kg

(maximum, 3 mL). Do not repeat the full loading dose intraoperatively.
3. After an initial loading dose, infuse no more than 0.5 mg/kg/hr (= 0.5 mL/kg/hr) of bupivacaine 0.1%.
4. Always use the loss-of-resistance technique with saline.
5. In general, the bolus for fentanyl is 1 μg/kg in 5 mL of preservative-free NS. The bolus for hydromorphone is 10 μg/kg in 5 mL of NS. Bupivacaine 0.1% and fentanyl, 2 μg/mL, usually start at 0.2 mL/kg/hr (1 μg/kg/hr).
6. Use Mastisol® to dress the catheter.

Intravenous Regional Anesthesia

Intravenous regional anesthesia of the upper extremity was first described by August Bier in 1908. In this method, a local anesthetic agent is injected IV into the involved limb while circulation to the limb is occluded. This form of regional block is often used for short surgical procedures (under 90 minutes) distal to the elbow. Surgery on the arm proximal to the elbow is best managed with a brachial plexus blockade.

An IV regional block is not suitable for lower extremity surgery because a large volume of local anesthetic solution is required.

INDICATIONS

Indications for IV regional anesthesia include
- Surgery on the arm proximal to the elbow
- Reduction of closed fracture of the forearm

CONTRAINDICATIONS

Contraindications to IV regional anesthesia include
- Sickle cell disease or trait (stasis of blood flow and acidosis promote sickling of red cells)
- Severe peripheral vascular disease or Raynaud's disease
- Cellulitis, infection, or venous thrombosis
- A history of heart block (severe bradycardia and asystole following tourniquet release have been reported even in patients with no myocardial conduction abnormality)
- A history of epilepsy or hepatic disease
- A full stomach (aspiration pneumonitis can occur if patient seizes after tourniquet release, owing to CNS toxic effects of local anesthetic agent)

TECHNIQUE

- Insert an IV catheter in the distal aspect of the limb involved in the surgical procedure. This usually means an IV catheter in the hand. Inadequate or patchy anesthesia is more likely to occur when proximal veins such as antecubital veins are used to administer the local anesthetic agent.
- Insert another IV catheter in the opposite extremity and have equipment available for immediate resuscitation in the event of an untoward reaction.
- Exsanguinate the surgical limb to ensure more complete analgesia. This can be achieved by placing an Esmarch bandage or elevating the arm for 2 minutes.
- Apply a double tourniquet to the arm proximal to the operative site and securely fasten it with an exterior bandage.
- Inflate the proximal cuff of the double tourniquet to 100 to 150 mm Hg above the patient's systolic blood pressure.
- Inject the local anesthetic agent slowly and without vasoconstrictors. Mottling of skin appears as the local anesthetic spreads. Muscular relaxation and complete analgesia usually develop within 10 minutes.
- Most patients tolerate the tourniquet pressure for 30 to 40 minutes. Once the proximal tourniquet becomes uncomfortable, the distal tourniquet is inflated to the appropriate pressure and the proximal cuff is deflated. Because the distal cuff is placed over an anesthetized segment of arm, the patient should again tolerate the tourniquet for another 30 to 40 minutes. The double cuff method, therefore, allows for approximately 60 to 90 minutes of relatively pain-free tourniquet time.
- Allow at least 30 minutes from the time of the local anesthetic injection and release of the tourniquet. Deflate the tourniquet slowly over 10 minutes. Following tourniquet deflation, sensation and motor function rapidly return to the anesthetized limb.
- Vigilant monitoring of the patient's pulse, blood pressure, ECG, and mental status is mandatory, especially after tourniquet release, to enable early detection of toxic reactions.

CHOICE OF LOCAL ANESTHETICS

- Prilocaine 0.5% in a dosage of 4 mg/kg (maximum dose, 500 mg) is the most rapidly metabolized amide and provides good anesthesia with few side effects. Theoretically, prilocaine could cause methemoglobinemia, especially if the total dose of prilocaine exceeds 600 mg in a 70-kg adult.
- Bupivacaine 0.25% is no longer recommended for use in IV regional anesthesia as bupivacaine has greater cardiotoxicity, and the affected patient may be difficult to resuscitate once toxic levels are attained.
- Lidocaine 0.5% has been extensively used for IV regional anesthesia in a dosage of 3 mg/kg. Side effects have included CNS toxicity, including dizziness, tinnitus, and convulsions. Cardiovascular toxic effects from lidocaine include hypotension, bradycardia, and ECG changes consisting of nodal rhythms, ventricular extrasystole, and cardiac arrest.

Recently, IV regional anesthesia with low-dose lidocaine (1 mg/kg) has been used for closed reduction of forearm fractures in children, with great success. To perform this block, dilute 1% lidocaine to 0.125%.

SIDE EFFECTS OF LOCAL ANESTHESIA

The side effects of local anesthesia include CNS toxic effects ranging from mild dizziness and tinnitus to convulsions. Also, cardiovascular toxic effects may occur, ranging from mild, transient bradycardia to cardiac arrest.

Toxic manifestations usually occur immediately after tourniquet deflation and correspond to the release of local anesthetic into the bloodstream. For this reason it is impor-

tant to ensure a minimum tourniquet inflation period of at least 30 minutes. Rapid reinflation of the tourniquet immediately after the initial deflation can result in a reduced peak plasma anesthetic level and a decreased incidence of side effects.

Strategies to Reduce Blood Transfusions

Scoliosis surgery is an extensive operative procedure that is often accompanied by substantial blood loss. Factors affecting blood loss during scoliosis surgery can be summarized into three groups:

- *Surgical factors*
  - Length of operation
  - Surgical technique
  - Extent of dissection and number of vertebrae to be fused
  - Site and size of bone graft and phase of operation in which it is obtained
  - Previous spinal fusions
- *Anesthetic factors*
  - Increased arterial pressure
  - Increased venous pressure
  - Intermittent positive pressure ventilation
- *Positioning factors*
  - Increased abdominal wall tension
  - Increased intra-abdominal pressure
  - Extrinsic pressure

Meticulous attention must be given to positioning the patient in the prone position so that the patient's abdomen is truly free of any pressure. There are two principal beneficial effects of a pressure-free abdomen. First, the pressure on the inferior vena cava is minimized, and hence venous congestion into the vertebral venous plexus is reduced. Second, avoiding abdominal pressure results in maintenance of FRC, which tends to prevent atelectasis and hypoxemia during surgery.

Complete relaxation of the diaphragm and the abdominal musculature decreases intra-abdominal pressure and hence decreases the pressure on the inferior vena cava. Straining, coughing, bucking, and airway obstruction all increase bleeding and should be avoided. To take advantage of gravity to help reduce blood pressure, the operative field should be the highest point of the patient's body. In scoliosis surgery this translates to a slight reversed Trendelenburg position.

INDUCED HYPOTENSION

The use of controlled hypotension to minimize blood loss during scoliosis surgery has decreased the need for blood replacement and total blood loss by an average of 40 percent. The induction of deliberate hypotension for pediatric spine surgery must not be taken lightly, however, as the risks and benefits vary with each patient and procedure and with the skill and knowledge of the surgeon and anesthesiologist.

The use of elective hypotension for spine surgery mandates that the reduction in blood pressure not interfere with the monitoring of spinal cord function. While reducing blood loss and improving operating conditions, it is essential to maintain adequate oxygen supply to the spinal cord. The goal is to achieve a moderate, sustained decrease in blood pressure and heart rate and to avoid hypoxia, hypercarbia, and acidosis during the course of the procedure.

A relatively dry operative field and improved operative conditions may not be achieved at a predetermined level of hypotension. The skilled anesthesiologist will consider many other factors, such as decreased heart rate, in order to achieve an improved operating condition.

One of the most important considerations of any hypotensive technique is the effect it will have on cerebral blood flow (CBF) and spinal cord blood flow. Studies in adults demonstrate that little change occurs in cerebral metabolism when mean arterial pressure (MAP) is maintained above 55 mm Hg. Brain ischemia in adults has been documented when a MAP of 55 mm Hg was combined with hypocarbia; however, no similar studies in children have been carried out. Maintenance of normal arterial carbon dioxide tension ($PaCO_2$) is vitally important to ensure adequate CBF and spinal cord blood flow during induced hypotension.

In the rare situation in which acute neurologic complications occur during scoliosis surgery, the spinal cord autoregulation is abolished. It is advisable to return the blood pressure to normal, and a blood transfusion may be administered to correct any significant anemia. At present, methylprednisolone is considered routine treatment in acute spinal cord injury.

Methylprednisolone, 30 mg/kg, followed by a continuous infusion of 5.4 mg/kg/hr for the next 23 hours, is administered. This drug must be started within 8 hours of injury to modify the secondary injury processes.

AUTOLOGOUS BLOOD DONATION

There are several techniques available to avoid the use of homologous blood. The complications of homologous blood transfusion include the transmission of such diseases as human immunodeficiency virus infection, hepatitis, and cytomegalovirus infection, as well as the possibility of hemolytic transfusion reaction, nonhemolytic reactions, and recipient alloimmunization. The three techniques of autologous blood donations are preoperative blood donation, intraoperative blood salvage, and acute normovolemic hemodilution. In each of these techniques the patient's own blood is reinfused, thereby eliminating the risks of transmission of infectious agents. Preoperative donation begins 4 to 6 weeks before surgery and can continue weekly until 72 hours before surgery. This time interval is necessary to allow plasma proteins to normalize and to restore intravascular volume. This blood has a shelf-life of 35 to 42 days, depending on the type of anticoagulant used in the storage bag. It may be frozen if surgery is postponed. Contraindications to autologous donation include a hematocrit less than 33 percent and bacteremia.

Recombinant erythropoietin in a dose of 600 U/kg administered subcutaneously, twice weekly, during the 4 weeks before surgery is recommended when relatively large donations of autologous blood are needed. Erythropoietin stimulates erythropoiesis by the bone marrow. Serum iron levels must be sufficient for erythropoiesis to be augmented. Therefore, patients receiving erythropoietin therapy must take iron supplements.

Autologous blood should not be given merely because it is available. Because of the risks of incorrect identification and possible bacterial contamination, it is prudent to avoid transfusion unless indicated.

Intraoperative blood salvage and reinfusion is now commonly used in orthopaedic surgery. Contraindications to the technique are usually related to contamination of the collected blood with infectious agents, cancer cells, or hemostatic agents such as protamine.

Acute normovolemic hemodilution involves withdrawing a calculated volume of the patient's blood after induction of anesthesia and simultaneously replacing it with crystalloid or colloid solutions. The patient's own fresh blood is reinfused near the end of the surgical procedure after major blood loss has ceased. There are two major advantages to acute normovolemic hemodilution. The blood lost during surgery has a low hematocrit, and fresh, whole, autologous blood is available for transfusion. Other advantages include a very cost-effective method of collecting autologous blood and no risk of clerical error if the blood is kept in the operating room after withdrawal. The amount of blood to be removed is calculated using the formula:

$$V_R = \frac{EBV \times (H_I - H_F)}{\frac{H_I + H_F}{2}}$$

where V_R = amount of blood to be withdrawn
H_I = initial hematocrit
H_F = final hematocrit
EBV = estimated blood volume

Combined deliberate hypotension and hemodilution will further reduce the need for homologous blood transfusion. However, the use of such combined techniques requires experience and vigilance, as oxygen delivery to critical organs may be at risk. It is advisable to limit hemodilution to a hematocrit of 28 to 30 and to maintain MAP at 55 to 60 mm Hg.

Latex Allergy

Perioperative anaphylactic reactions to latex are increasingly reported. A dramatic increase in reports of latex-induced allergic reactions began to appear in the late 1980s. This increase occurred shortly after the Centers for Disease Control and Prevention recommended the use of universal precautions, during which time the use of surgical gloves increased from 800 million annually to more than 20 billion.

The diagnosis of latex allergy is frequently made retrospectively. Latex may now be the most common cause of intraoperative sensitivity reactions.

IDENTIFICATION OF A HIGH-RISK PEDIATRIC GROUP

Children considered to be at high risk for latex allergy include the following:

1. Children with a history of multiple surgical procedures. This group includes patients with myelomeningocele, who have 500 to 1,000 times the risk of latex allergy, and those with congenital genitourinary tract anomalies.
2. Children with a history of a sensitivity reaction, such as urticaria, eye irritation, or bronchospasm, on exposure to balloons or other latex-containing objects.
3. Children with a history of allergic rhinitis, eczema, or asthma.
4. Children with a history of food allergy to tropical fruits such as avocado, kiwi, banana, or chestnut.

CLINICAL MANIFESTATIONS OF LATEX ALLERGY

A common manifestation of latex sensitivity is irritant contact dermatitis. Irritant contact dermatitis accounts for 80 percent of work-related reactions to gloves. This type of reaction results from a direct action of latex and other irritant chemicals found in latex gloves. Irritant contact dermatitis is not mediated by the immune system and is not a true allergy. However, the resulting deterioration in skin integrity may enhance absorption of latex protein and is believed to accelerate the onset of allergic reaction.

Another form of latex sensitivity is Type IV delayed hypersensitivity reaction. This reaction, also called allergic contact dermatitis, directly involves the immune system. The resulting skin rashes are similar to those caused by poison ivy. As with poison ivy reactivity, the skin rash usually appears 6 to 72 hours after initial contact and may progress from a mild dermatitis to oozing skin blisters.

The most dangerous form of latex allergy is Type I immediate hypersensitivity reaction. Also called IgE-mediated anaphylactic reaction, it can be very severe, even life-threatening. When an antigen (such as latex) induces the production of an antibody of the IgE class, re-exposure to the same antigen triggers a cascade of events, including the release of histamine, leukotrienes, and prostaglandins.

Reactions usually begin within minutes of exposure. Symptoms can be mild (skin erythema, hives, itching) to more severe (hoarse voice, chest tightness, swollen eyes, running nose), to life-threatening (bronchospasm and circulatory shock). The number of deaths reported in the literature that are directly attributed to Type I latex allergy is increasing.

SCREENING FOR LATEX ALLERGY

Preoperative screening to identify latex sensitivity is recommended for high-risk groups. The tests available are (1) patch testing, using a standardized patch test screen or a fragment of the offending latex product; (2) serologic tests, such as a radioallergosorbent test (RAST) and an enzyme allergosorbent test (EAST) for latex-specific IgE antibodies; and (3) skin prick tests.

These tests, however, lack specificity for predicting the likely occurrence of an anaphylactic reaction during anesthesia. The RAST test is no longer considered sufficiently specific, with 20 to 45 percent of patients having a negative RAST test but a positive skin test. A standardized latex skin prick test solution is not widely available, and the test itself can precipitate an anaphylactic reaction. It has been suggested that a RAST test be performed first, and if it is negative, a skin prick test can be performed. A positive history or findings on physical examination and one confirmatory test establish the diagnosis of latex sensitivity.

MANAGEMENT OF LATEX-SENSITIVE PATIENTS

Latex-sensitive patients must be managed with complete latex avoidance. This is not very easily achieved, as latex is prevalent in the health care environment, but exposure can be limited.

When possible, the patient should be scheduled for elective surgery as the first case of the day, when aerosolized latex antigens are at their lowest level in the operating room. The number of personnel entering the operating room should be limited, as they can be carriers of latex particles. Latex-free equipment is available, and guidelines for the management of anaphylaxis during anesthesia should be in place in all anesthetic settings.

Malignant Hyperthermia

Malignant hyperthermia is a devastating genetic condition that is almost exclusively related to general anesthesia. Two-thirds of susceptible patients manifest this syndrome during their first anesthetic exposure and the remaining one-third during subsequent anesthetic exposure.

The symptoms of malignant hyperthermia may present immediately (fulminant malignant hyperthermia) or up to many hours after induction of anesthesia and into the postoperative period. Malignant hyperthermia usually occurs in children and young adults, but it has been reported at all ages from 2 months to 70 years. The overall incidence of malignant hyperthemia is about 1 in 40,000 in adults and 1 in 15,000 in children. The incidence is higher when succinylcholine is used with other triggering agents. The triggering agents includes *all* potent volatile agents and the depolarizing muscle relaxants, such as succinylcholine.

The etiologic defect responsible for malignant hyperthermia has not yet been identified. A series of different mutations indicates a heterogeneous genetic basis for this syndrome. The Ryanodine receptor has been identified in humans and is responsible for release of calcium from the sarcoplasmic reticulum membrane. Malignant hyperthermia susceptibility may be related to mutations of the genes that modulate the Ryanodine receptor.

SIGNS AND SYMPTOMS

The first systemic effect of malignant hyperthermia is increased metabolism (increased oxygen consumption and carbon dioxide [CO_2] production). Therefore, the first clinically evident signs of this syndrome are tachycardia and tachypnea. Usually an increased end-tidal CO_2 precedes all other signs and symptoms and in this condition it is extremely difficult to bring the end-tidal CO_2 level to within normal range. Cardiac dysrhythmia, such as ventricular premature beats, bigeminy, and ventricular tachycardia, occur as a result of hyperkalemia and metabolic acidosis and their effects on the myocardium.

A mottled skin appearance, dark blood at the site of surgery, and muscle rigidity may also be present. Body temperature elevation is often a late manifestation of malignant hyperthermia. The diagnosis of malignant hyperthermia should not depend on an increase in body temperature.

Analysis of arterial and central venous blood will reveal respiratory and metabolic acidosis (pH of 7.15 to 6.80), arterial hypoxemia, hypercarbia (100 to 200 mm Hg), and marked central venous oxygen desaturation. Hyperkalemia may occur early in the course of the disease. The plasma CPK will be markedly elevated, although peak levels may not occur for 12 to 24 hours after an acute episode. Plasma and urine myoglobin concentrations are also elevated, and the urine has a pinkish red coloration. Late complications of untreated malignant hyperthermia include disseminated intravascular coagulopathy, acute renal failure, coma, and death.

TREATMENT

Immediate, aggressive therapy is absolutely essential as soon as malignant hyperthermia is diagnosed. The mainstay of treatment is dantrolene sodium. Dantrolene administered early during the episode decreases cellular metabolism and reverses the biochemical changes of malignant hyperthermia. The availability of IV dantrolene has reduced the mortality from malignant hyperthermia from 70 percent to less than 8 percent. The usual dose is 2 to 3 mg/kg IV, repeated every 5 to 10 minutes until signs of hypermetabolism subside. IV dantrolene should be continued postoperatively to prevent a possible recurrence of malignant hyperthermia. The following summarizes the treatment of malignant hyperthermia.

Specific Treatment

- Dantrolene (2 to 3 mg/kg IV) as an initial bolus, followed by repeat doses every 5 to 10 minutes until symptoms are controlled (rarely need a total dose in excess of 10 mg/kg).
- To prevent the recurrence of malignant hyperthermia: Dantrolene, 1 mg/kg IV every 6 hours for 72 hours.

Symptomatic Treatment

- Stop anesthesia and surgery immediately.
- Hyperventilate the lungs with 100% oxygen.
- Initiate active cooling (iced saline, 15 mL/kg IV every 10 minutes; surface cooling; gastric lavage with iced saline).
- Correct metabolic acidosis (sodium bicarbonate, 1 to 2 mEg/kg IV, based on arterial pH).
- Maintain urine output (hydration; furosemide, 1 mg/kg IV; mannitol, 0.25 g/kg IV).
- Treat hyperkalemia (glucose; insulin, 1 g of glucose per 1 unit insulin).
- Treat cardiac dysrhythmia (procainamide, 15 mg/kg IV).
- Monitor in an intensive care unit.

CONDITIONS ASSOCIATED WITH MALIGNANT HYPERTHERMIA

- Duchenne's muscular dystrophy
- Central core disease
- All muscular dystrophies
- Myotonia congenita
- King-Denborough syndrome
- Schwartz-Jampal syndrome

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APPENDIX 8–1 **Anesthetic Considerations for Pediatric Orthopaedic Syndromes**

Syndrome	Characteristics	Considerations
Amyotonia congenita (infantile muscular atrophy)	Noninherited, but sometimes familial, disease characterized by muscular atrophy involving anterior horn cell deterioration	Respiratory reserve compromise; difficulties in positioning; risk of aspiration due to pharyngeal muscles involvement Possible altering effects with neuromuscular-blocking anesthetics Succinylcholine contraindicated to reduce risk of hyperkalemia Increased sensitivity to thiopentone and respiratory depressants
Amyotrophic lateral sclerosis	Progressive degenerative disease involving muscular weakness; deterioration of motor neurons of spinal cord, medulla, and cortex	Succinylcholine contraindicated (induces hyperkalemia; possible cardiovascular collapse) Nondepolarizing relaxants may alter sensitivity and should be avoided. Reduced respiratory reserve; possible aspiration in presence of pharyngeal involvement
Apert's syndrome (acrocephalosyndactyly)	Congenital condition marked by irregular craniosynostosis, hypoplasia of midface, syndactyly, webbed fingers and toes; mental retardation; cervical spine stenosis; congenital heart disease	Difficult airway management and intubation due to subglottic stenosis Possibly increased intracranial pressure; postoperative ventilatory support may be required
Arthrogryposis multiplex congenita	Congenital multiple contractures of arms and legs; cardiac malformations; inguinal hernias; cleft palate; scoliosis; rigidity of temporomandibular joints	Ensure patient's ability to open mouth and control airway to determine difficulty of intubation and maintenance of airway requirements. Thiopentone requirement minimal, as muscles are replaced by fat
Beckwith-Wiedemann syndrome (infantile gigantism)	Macroglossia, omphalocele, macrosomatia, visceromegaly; neonatal polycythemia; hypoglycemia develops in 30%–50% of cases during early infancy; cardiovascular defects, including isolated cardiomegaly	Consider constant infusion of glucose administration to avoid severe rebound hypoglycemia. Possible difficulty securing airway and intubating
Carpenter's syndrome (craniosynostosis)	Premature closure of a skull suture; increased cranial pressure; hypoplastic mandible; associated congenital heart disease	Intubation may be difficult. Observation of intracranial compliance important for safe administration of anesthetic drugs Intraoperative venous air embolism risk with patient positioned in slight head-up position Corrective surgery associated with increased blood loss
Central core disease	Familial polymyopathy; nonprogressive muscle weakness; hypotonia; hip dislocation; kyphoscoliosis; funnel chest; mandibular hypoplasia; short neck; muscular dystrophy	Similar to amyotonia congenita Increased risk of malignant hyperthermia (MH)—follow all precautions to avoid MH.
Chotzen's syndrome	Congenital disorder marked by craniosynostosis; may lead to abnormal protrusion of eyeball; optic degeneration, blindness, increased intracranial pressure, seizures, and mental retardation; congenital cardiac defects; hydrocephalus	Possible difficult intubation Possibility of increased intracranial pressure and blood loss Monitor postoperatively for onset of hypotension or localizing neurologic signs.
Collagen vascular diseases (dermatomyositis, rheumatoid arthritis, systemic lupus erythematosus, polyarteritis nodosa, progressive systemic sclerosis, Wegener's granulomatosis)	Blood vessel diseases manifesting as generalized inflammation of connective tissue and blood vessels, frequently treated with steroids Variable systemic involvement	Pulmonary infiltration or fibrosis common; possible difficult airway management and intubation due to temporomandibular or cricoarytenoid arthritis Anemia common Fat embolism may occur after osteotomy, fracture, or minor trauma.
Conradi's syndrome (chondrodystrophy)	Cartilage formation defect at the epiphyses leading to dwarfism; saddle nose; asymmetric limb shortness; scoliosis; mild to moderate mental retardation; congenital heart diseases; renal anomalies	Associated renal and cardiac disease Possible difficult intubation; patients with small rib cage or severe lateral curvature of the spine at increased risk of pulmonary hypotension Increased risk of airway obstruction and inadequate ventilation suggest use of controlled ventilation.
Cri du chat syndrome	Chromosome 5-P abnormal Low birth weight; slow growth; catlike cry; microcephaly; micrognathia; congenital heart disease	Possible difficult airway management and intubation
Crouzon's disease	Congenital disease marked by shallow orbits and proptosis, premature craniosynostosis, maxillary hypoplasia, craniofacial dysostosis, deafness, beaked nose, hypoplastic maxilla, soft palate against posterior pharyngeal wall (making mouth breathing necessary), syndactyly associated with Apert's syndrome	Difficult tracheal intubation; possible severe blood loss during cranial operations; possible hypothermia; possible increased intracranial pressure; abrasions of cornea; possible requirement for postoperative mechanical ventilation

APPENDIX 8–1 **Anesthetic Considerations for Pediatric Orthopaedic Syndromes** *Continued*

Syndrome	Characteristics	Considerations
Down syndrome (mongolism)	Hypotonia; mental and physical retardation; hyperflexibility of joints; brachycephaly; mild microcephaly; small nasopharynx; congenital heart disease (CHD) in 50% of patients (endocardial cushion defeats, ventricular septal defects, tetralogy of Fallot, patent ductus arteriosus, atrial septal defect); abnormal cervical spine	Response to sedatives unpredictable Patency of upper airway (congenital subglottic stenosis) possibly difficult to maintain; difficult intubation; pathophysiology precautions for CHD; risk of laryngospasm (especially on extubation); increased risk of sleep apnea necessitating postoperative monitoring for respiratory complications
Duchenne's muscular dystrophy (pseudohypertrophic muscular dystrophy)	Progressive atrophy of muscles; kyphoscoliosis may develop; predisposition to long bone fractures; elevated plasma creatine kinase concentrations; compromised pulmonary function, often leading to pneumonia as a terminal event; sleep hypoxemia possible; degeneration of cardiac muscle leading to congestive heart failure	Respiratory insufficiency; compromised respiratory reserve; increased risk of aspiration; cardiomyopathy; difficulties in positioning; neuromuscular-blocking drugs altering effects Avoid use of succinylcholine (to reduce risk of life-threatening cardiac dysrhythmias and induced hyperkalemia). Increased sensitivity to thiopentone and respiratory depressants; avoid use of muscle relaxants. High susceptibility to malignant hyperthermia Postoperative mechanical ventilation may be necessary.
Ehlers-Danlos syndrome	Inherited elastic connective tissue disorder marked by hyperextensible skin, hypermobile joints, and excessive bleeding after minor trauma	Coagulation screen, including bleeding time, should be obtained preoperatively. Adequate blood products must be available. Difficult to maintain IV Increased risk of spontaneous pneumothorax, mitral regurgitation, and cardiac conduction abnormalities Susceptibility to excessive bleeding with even minor trauma suggests avoiding IM injections and instrumentation of nose and esophagus Patient must be carefully moved and positioned to avoid skin and skeletal traumas.
Epidermolysis bullosa	Hereditary disorder of the skin characterized by spontaneous subepidermal blistering followed by fluid accumulation	Preoperative assessment may reveal poor nutritional state, chronic anemia, respiratory infections, sepsis, or fluid and electrolyte disturbances, oral lesions or adhesions of tongue, renal disease, hypercoagulable state. Avoid trauma to the skin and mucous membranes (e.g., trauma from tape, adhesive electrodes, blood pressure cuffs, tourniquets, anesthetic face mask). Pressure points must be adequately padded during positioning. IV induction should be avoided (anticipate extremely difficult intubation). Face and chin should be protected prior to mask or finger contact. Perioperative supplemental corticosteroids may be indicated if patient has been on steroid therapy. Be aware of porphyria cutanea tarda.
Familial periodic paralysis	Intermittent, acute attacks of skeletal muscle weakness or paralysis; characterized as hypokalemic, normokalemic, or hyperkalemic	Determine electrolyte levels preoperatively. Avoid perioperative use of glucose- and sodium-containing IV fluid solutions; glucose may precipitate hypokalemia and potassium chloride may precipitate hyperkalemia in affected patients. ECG monitoring is recommended for patients exhibiting hyperkalemic characteristics. Avoid muscle relaxants. Perioperative temperature monitoring is critical. Prolonged monitoring postoperatively is indicated, even with an uneventful intraoperative course.
Friedreich's ataxia	Progressive degeneration of spinocerebellar and pyramidal tracts associated with cardiomyopathy and scoliosis	Increased risk of kyphoscoliosis may lead to cardiopulmonary failure. Care should be exercised in using cardiac depressant drugs.
Gardner's syndrome	Polyposis of colon associated with high risk of colon carcinoma developing; epidermal and/or sebaceous cysts; multiple osteomas and soft tissue tumors of the skin; fibromas	No unique anesthetic considerations in these patients
Goldenhar's syndrome (oculoauriculovertebral dysplasia)	Unilateral mandibular hypoplasia; cleft palate; associated anomalies include eye, ear and vertebral abnormalities; mental deficiency; micrognathia; 40% incidence of vertebral abnormality; 35% incidence of congenital heart disease (septal defects and tetralogy of Fallot)	Predictable upper airway obstruction and difficult tracheal intubation are considerations. Patients with chronic airway obstruction are at high risk for hypoxemia and possible development of pulmonary hypertension. During induction of anesthesia, spontaneous ventilation is desirable. Extubation should be delayed until patient is fully awake and alert. Associated congenital heart disease problems should be considered. Avoid muscle relaxants.

Appendix continued on following page

APPENDIX 8–1 **Anesthetic Considerations for Pediatric Orthopaedic Syndromes** *Continued*

Syndrome	Characteristics	Considerations
Guillain-Barré syndrome (acute idiopathic polyneuritis)	Acute polyneuritis with progressive muscular weakness or paralysis of extremities; difficulty swallowing; impaired respiration; autonomic nervous system dysfunction; high CSF protein content; cardiovascular system instability; impaired respiratory reserve; hypotension	Monitoring vital capacity and arterial blood gases will assist in managing ventilation. Avoid use of succinylcholine. Increased likelihood of the need for mechanical ventilation exists during perioperative and postoperative period; patient may need a tracheostomy. Autonomic nervous system dysfunction may lead to hyper- or hypotension and cardiac conduction disturbances. Special considerations include pharyngeal muscle weakness, sensory paresthesias, and pain.
Hand-Schüller-Christian disease (histiocytosis X)	Multisystem disease; lipid accumulation manifesting as histiocytic granulomas in bones, skin, and viscera; exophthalmos and diabetes insipidus sometimes present	Ulceration of palatal and gingival mucosa; laryngeal fibrosis; possible upper airway obstruction; possible mild anemia; pulmonary-diffuse or hilar infiltration; respiratory failure, cor pulmonale; liver involvement (hepatosplenomegaly and pancytopenia common); history of steroid use
Holt-Oram syndrome (heart-hand syndrome)	Upper limb defect, cardiac anomaly, narrow shoulders; mild congenital heart disease (70% of cases include atrial septal defect); atrial and ventricular cardiac rhythm disturbances; radial aplasia; hand abnormalities; possible sudden death from pulmonary embolus, coronary occlusion	Anesthetic management follows that for patients with cardiac defects. No other unique anesthetic problems are associated with these patients.
Hunter's syndrome (mucopolysaccharidosis II)	Coarse and mild facies; moderate dwarfism; possible retinitis pigmentosa; progressive deafness; nodular, ivory-colored skin lesions; hydrocephalus (occasionally severe); valvular and ischemic heart disease; stiff joints; hepatosplenomegaly; pectus excavatum; kyphoscoliosis; possible hypersplenism	Possible upper airway obstruction and difficult intubation Increased risk of pneumonia; valvular and ischemic heart disease frequently present
Hurler's syndrome (mucopolysaccharidosis I)	Large head, short neck; dwarfism; kyphoscoliosis; hepatosplenomegaly; cardiac valvular abnormalities; coronary artery disease; severe mental retardation; prone to respiratory infections; pulmonary hypertension; death before 10 years from respiratory and cardiac failure	Similar to Hunter's syndrome but more severe. Upper respiratory infection is frequent. Tracheobronchial cartilages are abnormal. Anticipate difficult airway management. Because severe coronary artery disease develops at an early age, be aware of valvular and myocardial abnormalities.
Klinefelter's syndrome (gonosomal aneuploidy)	Scoliosis; tall and slim stature; mental and physical impairment; microcephaly; cardiac disease	Position patient carefully. Otherwise, there are no unique anesthetic considerations in these patients.
Klippel-Feil syndrome	Short neck with low hairline and limited head movement; early development of cervical vertebrae; congenital fusion of cervical vertebrae; spinal canal stenosis or scoliosis; possible mandibular malformations and micrognathia; primary or secondary neurologic defects possible; congenital heart defects (ventricular septal defect most common); possible mental deficiency	Difficult airway management and intubation (preoperative lateral neck radiographs may be helpful); increased risk of neurologic damage during direct laryngoscopy if cervical spine instability is present
Klippel-Trénaunay-Weber syndrome (angio-osteohypertrophy)	Asymmetric limb hypertrophy; hemangiomas; various vascular malformations; arteriovenous fistula	Children who have an increased cardiac output will not tolerate anesthetic agents that decrease myocardial contractility.
Laurence-Moon-Biedl syndrome	Hypogenitalism; mental retardation; spastic paraplegia; possibility of congenital heart disease; obesity; retinitis pigmentosa; renal abnormalities; polydactyly	Follow cardiac or renal anesthetic precautions if abnormalities exist. Diabetes insipidus may be present.
Larsen's syndrome	Muliple joint dislocation; flat facies with depressed nasal bridge; prominent forehead; hypertelorism; connective tissue defect; deficient cartilage in rib cage, epiglottis, and arytenoids	Possible difficult intubation; increased care necessary during anesthesia owing to mobile arytenoid cartilage Possible cardiovascular disease and chronic respiratory problems
Lesch-Nyhan syndrome	Inherited metabolic disease that affects males only, characterized by absence of purine metabolism, mental retardation, autistic behavior, self-mutilation, seizures, hyperuricemia, and renal failure	Affected individuals are prone to regurgitation and vomiting. Consider preexisting renal dysfunction and/or impaired metabolism when planning anesthesia drug management. If a spastic skeletal muscle disorder is present, use succinylcholine cautiously. Use exogenous catecholamines cautiously.
Maffucci's syndrome	Dyschondroplasia; multiple cutaneous hemangiomas; variable early bowing of long bones; in 43% of cases, thrombosis of dilated blood vessels with phlebolith formation	Pathologic fractures; possible sensitivity to vasodilator drugs; GI bleeding from hemangiomas; orthostatic hypotension

APPENDIX 8–1 **Anesthetic Considerations for Pediatric Orthopaedic Syndromes** *Continued*

Syndrome	Characteristics	Considerations
Marfan's syndrome (arachnodactyly)	Hereditary condition of connective tissue, bones, muscles, ligaments, and skeletal structures. Thoracolumbar scoliosis; hyperextensibility; joint laxity; dilation with or without dissecting aneurysm of ascending aorta; mitral valve prolapse	If valvular heart disease is present, suggest prophylactic antibiotics. Transfer and position patient carefully to avoid joint stress and prevent intraoperative dislocations. Increased risk exists for cardiac failure, aortic aneurysm, and pulmonary complications (pneumothorax, decreased respiratory reserve, bronchogenic bullae); use myocardial depressant drugs cautiously; control hypertension aggressively.
Maroteaux-Lamy syndrome (mucopolysaccharidosis)	Coarse facies; short stature; stiff joints; cloudy corneas; possible hydrocephalus, macrocephaly, valvular disease; frequent upper respiratory tract infections; hepatosplenomegaly; kyphoscoliosis; skeletal dysplasia; possible odontoid hypoplasia; anemia; thrombocytopenia	Possible difficult intubation and airway management; poor respiratory reserve Use cardiac depressant drugs cautiously.
Möbius syndrome (congenital facial diplegia)	Congenital cranial nerve dysplasia resulting in face palsy; difficulty chewing and swallowing; micrognathia; recurrent pulmonary infections	Increased risk of pulmonary infections Possible difficult intubation and airway management
Morquio-Ullrich syndrome (mucopolysaccharidosis IV)	Partial deafness; short neck and trunk; progressive pectus carinatum; severe dwarfism; joint laxity; possible spinal cord compression and paralysis; aortic regurgitation; chronic pulmonary disease; cardiac complications	Possible spinal instability—laryngoscopy with excessive extension of head and neck should be avoided. Increased risk of pulmonary infections exists; valvular heart disease and presence of associated major organ disease (pulmonary, renal, hepatic) should be considered in planning anesthetic management.
Myasthenia congenita	Neuromuscular disorder characterized by weakness and fatigability of voluntary muscles; marginal respiratory function; pharyngeal and laryngeal muscle weakness; cardiomyopathy; hypothyroidism	Risk of aspiration is high. Use respiratory depressants and muscle relaxants cautiously; consider possible interactions between anticholinesterase drugs and anesthetic drugs. Cholinergic crisis is possible. Use narcotic drugs cautiously. Patient will probably need ventilatory support postoperatively.
Myositis ossificans (fibrodysplasia ossificans)	Inflammation of connective tissues, usually in elbow, hip, and knee muscles, leading to ectopic bone formation of skeletal muscle mass; limited joint movement	Restricted neck and jaw movement may make intubation difficult. Severe limitations of rib movement may lead to aspiration and asphyxia, pneumonia, and greatly diminished thoracic compliance.
Myotonia congenita (Thomsen's disease)	Nonprogressive disease, similar to myotonic dystrophy but more benign; difficulty in relaxing contracted muscles; poor swallowing, leading to aspiration; possible facial muscle weakness, ptosis, and arthrogryposis; may be worsened by exercise or cold	Possible difficult airway management High risk of sensitivity to muscle relaxants and respiratory depressants exists; succinylcholine is contraindicated. Close, continuous monitoring should be provided for patients given preoperative sedatives or analgesics. Perform close cardiac monitoring and avoid cold stress. There is a possibility of prolonged recovery time; ventilation and airway should be supported until patient has adequately recovered. Patient may be predisposed to pulmonary complications.
Myotonic dystrophy (myotonia dystrophia)	Hypotonia; facial diplegia; muscle weakness; mental deficiency; cataracts; ptosis; clubfeet; scoliosis; cranial hyperostosis; poor feeding; impaired deglutition; cryptorchidism; hypogonadism; conduction defects with arrhythmias; frontal baldness; diabetes mellitus; decreased thyroid function; hypersomnolence; central sleep apnea; respiratory insufficiency	Possible difficult airway management The presence of cardiomyopathy, respiratory muscle weakness, and abnormal responses to anesthetic drugs are considerations in the anesthetic management of these patients. Myocardial depression may be exaggerated. Succinylcholine is contraindicated. Patients are highly sensitive to respiratory depressants (barbiturates, opioids, diazepam). Monitor ECG. Postoperative ventilation may be required, and prolonged recovery from anesthesia should be anticipated. Patients may be predisposed to pulmonary complications.
Niemann-Pick disease (sphingomyelin-cholesterol lipidoses)	Hereditary disease characterized by sphingomyelin and cholesterol accumulation in and enlargement of liver and spleen; anemia; lymphadenopathy; progressive mental and physical deterioration; foam cells in bone marrow, spleen, or lymph nodes; CNS involvement; diffuse infiltration of lungs; epilepsy	Suggest rapid-sequence induction to reduce risk of regurgitation and pulmonary aspiration. High risk of pneumonia; possible thrombocytopenia
Noack's syndrome (craniosynostosis)	Congenital disorder with variety of deformities due to premature closure of the skull suture(s), which may lead to exophthalmos, optic degeneration, increased intracranial pressure, mental retardation, subaortic valvular hypertrophy, ventricular septal defect	Possible difficult intubation Increased risk of raised intracranial pressure; continuous arterial blood pressure monitoring is suggested. Intraoperative venous air embolism is possible, depending on patient position, and precautions should be taken. Blood loss may be high if corrective cranial surgery is performed.

Appendix continued on following page

APPENDIX 8–1 **Anesthetic Considerations for Pediatric Orthopaedic Syndromes** *Continued*

Syndrome	Characteristics	Considerations
Noonan's syndrome (male Turner's syndrome)	Webbing of the neck; short stature; low-set ears; mild to severe mental retardation; slight obesity; pectus excavatum; pulmonary valve stenosis; hypertension; micrognathia; hydronephrosis or hypoplasia of kidneys	Possible difficult airway management and intubation Cardiopulmonary complications should be considered. Administer renally excreted drugs cautiously if kidney complications are present.
Ollier's disease (enchondromatosis)	Multiple exostoses of growth of the epiphyses, metacarpals, and phalanges; prone to fractures	Precautions should be used in positioning patient.
Orofaciodigital syndrome	Oral frenula and clefts; hypoplasia of alae nasi; asymmetric shortening of digits; mental deficiency; hydrocephalus; seizures; polycystic renal disease; hypoplastic mandible and maxilla	Possible difficult intubation and airway management Administer renally excreted drugs cautiously if renal complications are present.
Osteogenesis imperfecta (fragilitas ossium)	I. *Congenita*—usually early mortality owing to fractures occurring in utero II. *Tarda*—blue sclera; fractures with minor trauma (changing a diaper, applying blood pressure cuff); scoliosis; discolored and brittle teeth; cardiovascular anomalies (aortic and mitral valve regurgitation, chordae tendineae abnormalities, aortic cystic medial necrosis)	In children with severe osteogenesis, extreme care should be taken in positioning the patient, monitoring, establishing IV access, and controlling airway. There is a high likelihood of restrictive lung disease and cardiovascular problems. Succinylcholine is contraindicated. Fragile vessels lead to subcutaneous hemorrhage. Dentin deficiency causes carious and easily broken teeth.
Pierre Robin syndrome	Mandibular hypoplasia; micrognathia and glossoptosis, often in combination with cleft palate; possible airway obstruction development within first 4 wk of life; possible associated congenital heart disease	There is a possibility of difficult intubation and airway management; newborns may have total airway occlusion due to posterior tongue displacement. Intensive nursing attention and maintaining patient in prone position may improve outcome. There is an increased likelihood of pulmonary artery hypertension, cor pulmonale, abrupt respiratory failure, aspiration, and infection. Muscle relaxants are contraindicated.
Prader-Willi syndrome	*Infant*—severe hypotonia, feeding problems, respiratory tract problems; absent reflexes *Second phase*—small, very obese stature; possible severe mental retardation; hyperphagia; hypotonia; behavior problems; possible development of diabetes mellitus; uncontrollable polyphagia	Monitor for hypoglycemia. Assess for possible upper airway obstruction. Ventilation may be impaired postoperatively due to extreme obesity; obesity may lead to cardiopulmonary failure.
Riley-Day syndrome (familial dysautonomia)	Inherited CNS disorder manifesting as abnormal autonomic nervous system, sensory, motor, and psychic dysfunction; characterized by sudden variations in blood pressure; emotional lability (patient vulnerable to unrecognized trauma or self-multilation); retarded growth; erratic temperature control; pulmonary aspiration; cardiovascular instability	These patients are unable to compensate for sudden acute blood loss, increased intra thoracic pressure, or sudden change in body position. Blood pressure should be closely monitored and hypotension should be aggressively managed. Anesthetic requirements may be reduced. Patients are highly sensitive to adrenergic and cholinergic drugs. Respiratory depressants should be avoided. Postoperatively, there is a high risk of vomiting and aspiration.
Sanfilippo's syndrome (mucopolysaccharidosis III)	Mildly coarse facies; mildly stiff joints; severe, progressive mental retardation, sleep disturbances, chronic respiratory tract infections; emotional disturbances; agitation	There are no unique anesthetic requirements in these patients.
Scheie's syndrome (mucopolysaccharidosis I S)	Broad mouth with full lips; coarse facial features; early corneal clouding; normal intelligence; deafness; aortic valvular disease; sleep apnea; decreased joint mobility; carpal tunnel syndrome; cardiomyopathy	Care should be taken in positioning and moving the patient to decrease the risk of joint dysplasia. Chronic pulmonary infections may complicate anesthetic management.
Scleroderma	Chronic disease causing hardening of the skin; visceral involvement includes gastrointestinal tract, lungs, heart, kidneys, peripheral nervous system, musculoskeletal system; diffuse pulmonary fibrosis; plastic surgery required for contractures and constrictions	Possible difficult airway management and intubation There is a high risk of cardiopulmonary complications; evaluation of cardiopulmonary, renal, and hepatic function should be completed preoperatively. Risk of arterial hypoxemia and peripheral vasoconstriction is high. Operating room should be kept warm. Postoperatively, ventilation may be required. Care in positioning is also suggested. Patient may have a history of steroid therapy.
Silver's syndrome	Prenatal onset of short stature; skeletal asymmetry; small facies; possible excess sweating; liability to fasting hypoglycemia in infants and young children; abnormal sexual development; café-au-lait spots	Micrognathia may predispose to difficult intubation.
Smith-Lemli-Optiz syndrome	Anteverted nostrils; ptosis of eyelids; microcephaly; micrognathia; moderate to severe mental retardation; hypotonia; males predisposed to cryptorchidism, hypospadias, and syndactyly	High probability of airway management and intubation difficulties High risk for pneumonia; increased susceptibility to infection

APPENDIX 8–1 **Anesthetic Considerations for Pediatric Orthopaedic Syndromes** *Continued*

Syndrome	Characteristics	Considerations
Sotos' syndrome (cerebral gigantism)	Large size, large hands and feet; delayed motor development; moderate to severe mental deficiency; macrocephaly; hypertelorism; prognathism; scoliosis; neonatal problems with respiration and feeding	Possible airway management and intubation problems
Stevens-Johnson syndrome	Inflammatory cutaneous syndrome; severe lesions of stomatitis and conjunctivitis; possible toxic epidermal necrolysis; possible high fever, tachycardia, tachypnea; possible hypersensitivity to external agents (drugs, infections, etc.)	There is a high risk of cardiovascular and/or respiratory involvement; there is a possibility of difficult airway management and intubation. If extensive oral mucosal lesions are present, avoid intubation or use of esophageal stethoscope. Patients are at high risk for pneumothorax, cardiac problems (myocarditis, pericarditis), and renal insufficiency. Avoid IM or SC injections and barbiturates. Monitoring is difficult because of skin lesions but essential.
Sturge-Weber syndrome	Congenital syndrome characterized by port-wine stain on face, mostly in a facial distribution; angiomas of leptomeninges and choroid; glaucoma; intracranial calcifications; epileptic seizures (grand mal); possible mental retardation	There are no unique anesthetic considerations in these patients.
Tay-Sachs disease (gangliosidosis)	Neurologic deterioration—mental and physical retardation; blindness; hypotonia; spasticity; convulsions; poor motor function; death usually by 4 years of age	Patients are at high risk for severe pulmonary disease and therefore at high risk for aspiration.
Thrombocytopenia with absent radius	Abnormal decrease in the number of blood platelets; possibility of spontaneous hemorrhages into the skin, mucous membranes, internal organs, and other tissues; epistaxis; may be precipitated by stress, infection, surgery; congenital heart disease in 30%; increased risk of intracranial hemorrhage	Surgery patients should be transfused with platelet concentrates as often as necessary to control hemorrhages. Minimize trauma to upper airway. Steroid augmentation may be needed. IM injections are contraindicated.
Treacher Collins syndrome (mandibulofacial dysostosis)	Hypoplasia of facial bones; micrognathia; macrostomia; cleft palate; abnormal dentition; malformation of the ear; conductive deafness; pronounced narrowing of the pharynx; respiratory problems; congenital heart disease (most commonly ventricular septal defect)	Probable upper airway obstruction and difficult intubation; not as severe as Pierre Robin syndrome
Tuberous sclerosis	Convulsive seizures; progressive mental disorder; adenoma sebaceum; intracranial calcification; tumors of the kidneys and brain with projection into cerebral ventricles; hamartomas in lungs, kidneys, heart	Because of kidney inflammation and renal failure, renally excreted drugs are contraindicated. There is an increased risk of pulmonary complications with rupture of lung cysts, and a risk of cardiac arrhythmia.
von-Recklinghausen's disease (neurofibromatosis)	Numerous tumors of nerve sheaths occurring in skin, subcutaneous tissue, bones, internal organs, and CNS (peripheral, spinal, and cranial nerve roots); café-au-lait spots; congenital pseudarthrosis; short stature; motor seizures; possible cardiorespiratory and neurologic compromise; possible renal artery compression or stenosis; possible mental impairment	Rule out obstructive airway symptoms. Pulmonary function tests should be performed preoperatively in patients with scoliosis. Hypertensive patients should be evaluated for pheochromocytoma, and laboratory tests should include urinary catecholamines, pituitary function, electrolytes, blood urea nitrogen, and creatinine. Anticipate possible tumors in larynx and right ventricle outflow tract. Renally excreted drugs should be considered cautiously if kidney disease is involved.
Welander's syndrome (muscular atrophy)	Peripheral muscles involved initially; prognosis good for life, poor for ambulation	Spinal fusion surgery may be required. Caution is advised in the use of thiopentone and muscle relaxants. Respiratory depressant drugs should be avoided.
Werdnig-Hoffmann disease	Progressive, hereditary infantile form of muscle atrophy caused by degeneration of anterior horn cells and cranial nerve motor nuclei occurring in first 3 years of life, characterized by hypotonia, flaccid paralysis, dysphagia leading to pulmonary aspiration, difficulty in feeding, inability to control emotional responses	Succinylcholine may induce hyperkalemia and lead to cardiovascular collapse. Nondepolarizing muscle relaxants and respiratory depressant anesthetics should be avoided. Patients are predisposed to pulmonary aspiration. Postoperative ventilatory support must be provided for patients unable to maintain airway or adequate ventilation. It may be difficult to wean patient from assisted ventilation.
Wolff-Parkinson-White syndrome	Cardiac rhythm abnormality evidenced by supraventricular tachycardia; abnormal conduction pathway between atria and ventricles; short P-R intervals, wide QRS complexes and delta waves; congenital heart diseases	Anesthetic management focuses on preventing atrial fibrillation, atrial flutter, and reciprocating tachycardia by treatment that controls ventricular rate, chiefly vagal maneuvers. Electrical cardioversion and artificial cardiac pacing are used to treat hemodynamically significant tachydysrhythmias.