Chapter 22: Anesthesia for Orthopedic Procedures

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INTRODUCTION

FOCUS POINTS

1. Somatosensory evoked potentials (SSEPs) indicate the integrity of the afferent pathways of the dorsal columns of the spinal cord.
2. Increased latency and decreased amplitude are indicators of potential injury to the spinal cord.
3. Motor evoked potentials (MEPs) are both more sensitive to anesthetic agents and spinal cord injury, but only function in the absence of neuromuscular paralysis.
4. Careful positioning is paramount to all procedures especially prone and those of long duration; it should be completed in collaboration with the surgeon, anesthesiologist, and nursing staff.
5. Postoperative visual loss (POVL) is a devastating complication of prone positioning and has been associated with hypotension, anemia, and direct external pressure.
6. Surgical wound infection prevention is the responsibility of all members of the health care team. It can be minimized by hand hygiene with feedback (monitoring of practitioners), frequent environmental cleaning, patient decolonization, improved line access methods, and infection surveillance.
7. Congenital and neuromuscular scoliosis patients tend to have significantly higher blood loss than patients with idiopathic scoliosis undergoing spinal fusion.
8. Maintaining a neutral cervical spine position and awareness of the potential for a difficult airway are the most important anesthetic considerations for patients with Klippel-Feil syndrome.
9. There are multiple forms of osteogenesis imperfecta (“brittle bone” disease), all of which require extreme care perioperatively to prevent fractures (padding, avoidance of frequent noninvasive blood pressures, etc.).
10. Patients with Marfan syndrome have skeletal, cardiovascular, and ocular abnormalities. The major cause of morbidity and mortality though is dilation of the aortic root leading to aortic dissection.
11. When an Ehlers-Danlos syndrome (EDS) child presents for surgery, particular attention should be given to bleeding tendencies with a low threshold to prepare blood products.
12. The most severe form of cerebral palsy is spastic quadriplegia with higher association of intellectual disability, seizures, and swallowing difficulties.
13. Cardiomyopathy is a major cause of death in patients with Duchenne muscular dystrophy (DMD) and all patients should undergo a cardiac evaluation with echocardiogram or cardiac MRI prior to an elective anesthetic.
14. Succinylcholine has been used without incident in spinal muscular atrophy (SMA) patients, but there is a potential for rhabdomyolysis and hyperkalemia and should be used with extreme caution.
15. Scoliosis repair requires careful planning that includes positioning, adequate access, invasive monitoring, and neuromonitoring (SSEP, MEP). The anesthetic plan should be tailored to the degree of surgical repair and the comorbidities of the patient.
16. The single most important risk factor for venous thromboembolism (VTE) in the pediatric population is the presence of a central venous catheter (CVC).

Anesthesia for orthopedic surgery in children is determined as much by the patient’s underlying health status and comorbidities as it is by the specific operation. The anesthetic plan varies depending on individual circumstances. In this chapter, we will outline many of the reasons patients present to the orthopedic operating room, both in elective and emergent circumstances. Conditions and syndromes most pertinent to orthopedic surgery will be described and particular anesthetic concerns will be highlighted.

Regional anesthesia is a critical component of pediatric orthopedic surgery. In pediatric anesthesia, regional procedures are often performed while the patient is under general anesthesia. Ideally, blocks are performed prior to surgery. Upper extremity surgeries are facilitated by blocks of the brachial plexus. Lower extremity surgeries are facilitated by blocks of the femoral and/or sciatic nerves. Epidural analgesia can be considered for any bilateral lower extremity procedure. Regional anesthesia is covered in more detail elsewhere in this text.

ANESTHETIC MANAGEMENT

Many aspects of anesthetic management for orthopedic procedures in children are similar to those for nonorthopedic procedures. This chapter highlights some of the issues most pertinent to the orthopedic operating rooms, including neuromonitoring, positioning, infection prevention, and tourniquet physiology.

Neuromonitoring

Intraoperative neurophysiological monitoring is an integral component of surgery and anesthesia for major spine procedures. Monitoring of somatosensory evoked potentials (SSEPs) and motor evoked potentials (MEPs) has made identification of intraoperative ischemia to the spinal cord possible, which may enable the team to address the problem before irreversible damage occurs. Spinal cord perfusion is determined by the mean arterial pressure (MAP) minus the cerebrospinal fluid (CSF) pressure and is generally autoregulated within the range of 60 to 150 mm Hg. Spinal cord perfusion pressure is influenced by hypoxia, hypercarbia, and temperature. Inadequate spinal cord perfusion puts the patient at risk of neurological injury. Combined, SSEPs and MEPs provide a highly sensitive and specific measure of spinal cord function.

SSEPs indicate the integrity of the afferent pathways of the dorsal columns of the spinal cord. SSEP monitoring measures the average electrical response at the cortex to a peripheral stimulus. Most commonly, the posterior tibial or peroneal nerves are used for monitoring lower extremities and the ulnar nerve is used for monitoring upper extremities. Limitations of SSEPs include the delay associated with data collection, signal averaging, and the fact that the anterior spinal artery does not directly supply the dorsal columns.[1] Upon stimulation of a peripheral nerve, amplitude and latency of the cortical responses are monitored. Increased latency and decreased amplitude require evaluation as they are indicators of potential injury to the spinal cord.

MEPs provide a more sensitive measure of spinal cord function. The efferent motor pathways occur along the anterior portion of the spinal cord. MEPs are both more sensitive to anesthetic agents and spinal cord injury, but only function in the absence of neuromuscular paralysis. A stimulus is generated over the motor cortex of the scalp and the electrical response of the corresponding peripheral muscles is monitored. Most often, the tibialis anterior is used for this purpose. Upper extremity motor function is also monitored and can serve as a benchmark for anesthetic-induced decrease versus true neurological compromise.

Historically, a wake-up test was utilized to rule out the possibility of a motor deficit prior to closure after spine surgery. Currently this test is still performed although by fewer surgeons and sometimes only when there is concern for neurological injury based on neurophysiological monitoring. A wake-up test involves lightening the anesthetic until the patient can follow commands. The end point is to have the patient move their fingers and toes upon request. Anesthesia is deepened as soon as the wake-up test is complete. In the event that a patient can squeeze their fingers upon command, but no lower extremity movement is noted, a neurological injury is assumed.

Performing a safe and effective wake-up test requires skill and advance planning to occur efficiently. When possible, the patient can be prepared ahead of time for the test. Very few patients remember the wake-up test, especially when they have been prepared for it. The idea of the wake-up test is somewhat terrifying for patients, but most are reassured to know that even in the unlikely event that they have recall of the wake-up, they will not feel pain.

Communication with the surgeon to clarify timing of the test can be very helpful. All medications are discontinued in anticipation of the wake-up test. The patient should be placed on 100% oxygen. The surgeon packs the field with wet lap sponges and covers the wound. This is to prevent venous air embolism with deep inhalation and maintain sterility of the site. One person should be under the drapes to feel for movement of the toes. Two people should be at the head of the bed. One person can hold a hand and place a hand on the patient's head to prevent sudden movement and dislodging of the endotracheal tube. The other person can hold the other hand and be ready to manage the ventilator or administer medications when the test is complete.

The wake-up test has the potential for disaster if the patient self-extubates in the prone position or comes off the operating table completely. Therefore, a stretcher should be in the room and ready and all members of the team available to respond in case of an emergency. A wake-up test is only helpful in patients who can understand and respond to the command to “wiggle your toes.” It can be performed in less than 5 minutes with planning, communication, and attention to the timing of any sedating medications that are long-acting.

Positioning

Careful positioning of pediatric patients is an important consideration in anesthesia care. It may be particularly challenging for patients with contractures for whom standardized operating room beds may not have adequate support. However, these same children may be specifically prone to positioning injury due to poor nutrition and chronic pressure points. Major orthopedic procedures may be associated with large volume blood loss and the resulting hypotension would place the patient at increased risk of pressure necrosis.

Peripheral nerve injury under anesthesia is thought to be related to stretching or compression of the nerve in addition to direct trauma caused by needle puncture or chemical toxicity. It is helpful to elicit a complete history of any underlying nerve damage during the preoperative interview, which can guide careful positioning. Additionally, during long procedures, pressure points, especially the face, can be periodically adjusted when the patient is positioned prone and the head when positioned supine. Fortunately, severe and permanent neurological dysfunction is rare from anesthesia and surgery.

Many orthopedic spine procedures are performed in the prone position. Confirmation of safe positioning prior to the start of the surgical procedure is the responsibility of the surgeon and circulating nurse in collaboration with the anesthesiologist. The face is usually in a foam headrest, which allows the ventilating circuit to exit without pressure or tension. The chest should be supported by chest-rolls placed carefully to avoid pressure on nipples. Shoulders should be abducted less than 90 degrees and the ulnar groove protected. Hips should be supported with particular attention to padding at the anterior superior iliac crests, knees should be clear of pressure, and feet should be supported on pillows, blankets, or pads. Male genitalia should be free of pressure as well. One benefit to SSEP monitoring is that there is potential for positioning injury to be identified prior to the occurrence of irreversible damage.

In addition to the importance of padding pressure points during prone positioning, there are some physiological changes pertinent to anesthesia in that position. Intraocular pressure is increased, which can cause decreased ocular perfusion pressure despite normal mean arterial pressures.[2] It is generally recommended that the bed be at a 15-degree reverse Trendelenburg position to protect the eyes. Postoperative visual loss (POVL) is a devastating complication during surgery and is poorly understood. Factors that may contribute include retinal ischemia, optic nerve ischemia, and optic vein engorgement in the setting of hypotension, anemia, and external pressure. However, POVL has occurred in the absence of any known risk factors.[3]

Prone positioning can compromise ventilation due to increased intra-abdominal pressure. Tables have been designed that allow the abdomen to hang free benefiting ventilation. Increased intra-abdominal pressure decreases venous return via compression of the inferior vena cava, thus resulting in decreased cardiac output and end-organ perfusion. Healthy patients may or may not demonstrate untoward effects from the physiological sequelae of prone positioning, but more fragile patients may develop hemodynamic and respiratory compromise.

Infection Prevention

Infections in orthopedic surgery patients can have far-reaching consequences due to the potential for hardware infection and infection deep in the tissues, joints, and bones, which can severely limit a patient’s mobility and quality of life. The anesthesiologist plays an important role in the prevention of surgical site infections (SSIs) by using aseptic techniques and timely administering prophylactic antibiotics.

According to the Centers for Disease Control and Prevention (CDC), health care–associated infections (HAIs) are present in 1 in 25 in-hospital patients on any given day.[4] SSIs are a major component of HAIs and not only increase patient morbidity and mortality but also reflect poorly on a hospital system and will increasingly be tied to lower reimbursement rates. The financial burden of HAIs is estimated to be 30 billion dollars annually.[5]

Based on data collected in 2010 by the American College of Surgeons (ACS) National Surgical Quality Improvement Program-Pediatric (NSQIP-P), the overall rate of pediatric SSIs was 1.8% and the rate of neonatal SSIs was 3%.[6] Neonates are thought to be at higher risk of SSI due to their immature immune systems. This data does not demonstrate the difference that may be seen in SSI in sicker patients with chronic illnesses versus healthy

patients presenting for straightforward outpatient operations.

According to a retrospective study of data collected over a 10-year period, children who did not receive antibiotic prophylaxis within the recommended 60-minute time frame were at a 1.7-fold increased risk of developing an SSI. Identified modifiable risk factors for development of SSIs included incorrect dosing and time of administration of antibiotics.[7]

Another retrospective study from a 9-year period of data covering 16,031 patients indicated a rate of SSI of 0.99% (159 patients). Risk factors identified by this study included young age (neonates), African American race, postoperative ICU admission, urinary catheters, and implantable device placement. In this study, wound classification and antibiotic administration were not independent predictors of SSI.[8]

In addition to patient and surgical risk factors, and antibiotic administration, the anesthesia team can impact the development of HAIs. In particular, recurrent access of central lines may increase the risk of a HAI. The mantra now in practice is as follows: one syringe, one patient, one time, indicating that a syringe should only be used for administration of one dose to one patient and should not be refilled even with the same medication for the same patient.

Keeping the anesthesia work environment clean between cases is also essential in decreasing HAI and SSI. In addition to thorough cleaning between cases, hand hygiene among the anesthesia personnel is critical. Filters in the patient circuit can also help prevent bacterial transfer. Stopcock contamination is a significant problem for patients in the operating room. A comprehensive method to minimize bacterial transmission to patients in the anesthesia work area includes excellent hand hygiene with feedback (monitoring of practitioners), frequent environmental cleaning, patient decolonization, improved line access methods, and infection surveillance.[9]

Tourniquets

A tourniquet is a compression device often used during orthopedic surgery on an extremity to limit blood loss and maintain a clear operating field. It is usually placed on the patient prior to the surgical prep but not inflated until just prior to incision. The limb can be exsanguinated with an Esmarch bandage or by gravity and the tourniquet is then inflated to 50 to 100 mm Hg above the pressure required to occlude the arterial supply to the limb. To prevent injury the cuff should have a width that is greater than one-half the limb's diameter and should be inflated to the lowest pressure possible to satisfy the surgical requirements. Inflation time is monitored closely.[10]

While there is controversy over the maximum time a tourniquet can safely stay inflated, the general recommendation is not to exceed 2 hours. This is based on the fact that cellular changes appear to be reversible within 2 hours of ischemia.[10] When the safe ischemic time is exceeded, patients can have damage to muscle and nerves and develop neuropathy as a result. In patients who are prone to neuropathy for other reasons, the tourniquet time should be decreased.

Tourniquet pain is a phenomenon that can occur both in patients who are awake and in those who are under general anesthesia. It begins approximately 45 minutes into the tourniquet time. Patients who are anesthetized with a regional block but otherwise awake will note a dull ache that eventually becomes extremely painful. Patients under general anesthesia will often have increased blood pressure and heart rate when the tourniquet pain starts to set in. The pain does not respond well to narcotics and the hemodynamics, when necessary, can be treated with labetalol. The pain resolves when the tourniquet is deflated.

Tourniquet inflation increases blood pressure and central venous pressure. Likewise, with deflation the blood pressure and central venous pressure decrease.[10] For patients who are hemodynamically fragile, this may be relevant. Most children, however, do not have a dramatic hemodynamic response to inflation or deflation. The patient's core temperature will generally increase during tourniquet inflation and should be closely monitored to maintain euthermia.

With release of the tourniquet, there is an increase in carbon dioxide, which is most noticeable as increased end-tidal CO_2 in patients under general anesthesia. There is also a transient metabolic acidosis due to washout of the anaerobic metabolic byproducts such as lactate. These changes are generally self-limiting assuming the tourniquet time has been reasonable.

Blood Conservation

Some orthopedic operations and in particular spine surgeries are associated with significant blood loss. Congenital and neuromuscular scoliosis patients tend to have significantly higher blood loss than patients with idiopathic scoliosis undergoing spinal fusion. Neuromuscular patients have an almost seven times higher risk of losing greater than half of their blood volume during scoliosis surgery.[11] Optimal methods of mitigating blood loss transfusion are ongoing areas of interest. Some patients, such as the Jehovah's Witness population, will not accept blood products regardless of consequence, including death.

Blood loss is important because loss of oxygen-carrying red blood cells results in decreased delivery of oxygen to end-organs such as the kidneys, heart, and brain. Additionally, blood loss in excess of half the patient's blood volume and fluid replacement can result in dilutional coagulopathies, which lead to further hemorrhage and the risk of exsanguination. Predictors of blood loss for spine surgery include operative time, preoperative kyphosis, male sex, and mean arterial pressure.[12]

The determination of transfusion thresholds is an actively debated topic and ultimately is an individualized decision based on the risk/benefit ratio for each clinical situation. Healthy patients can tolerate significant anemia without known untoward effect, but patients with known cardiac impairment or other chronic illnesses may require a lower transfusion threshold for optimization of long-term outcomes.

In the past, deliberate hypotension was a common method of conserving blood loss. Despite the fact that it is effective for this purpose, it is no longer in common use for long spine surgeries. Prolonged hypotension can negatively affect perfusion to the spinal cord, kidneys, and eyes increasing the risk of potentially devastating consequences.

Hemodilution is another method of blood conservation but should be used with caution due to similar risk of anemia and loss of oxygen-carrying capacity. Cardiac indices and oxygen extraction increase during hemodilution and systemic vascular resistance, oxygen delivery, and mixed venous saturation decrease. When oxygen delivery becomes critical, lactic acidosis develops; therefore, close monitoring of hemoglobin is warranted. Complications of hemodilution include postoperative pulmonary edema, anasarca, and prolonged postoperative mechanical ventilation.[3]

Cell saver is often used for spine surgeries as a method of returning the patient's own blood to oneself. This is generally acceptable for Jehovah's Witness patients provided the blood is kept in a continuous circuit in connection with the patient. Contraindications to cell saver include tumor operations and allergy to the anticoagulant. Potential complications of the use of salvaged blood include air embolism and hemolytic and bleeding complications from centrifugation, cellular debris, or anticoagulant overdosage.[3]

Antifibrinolytic medications are increasingly used in long surgeries associated with high-volume blood loss. Both ε-aminocaproic acid (EACA) and tranexamic acid (TXA) can decrease blood loss in spine surgery for idiopathic and neuromuscular scoliosis surgery. TXA is a synthetic antifibrinolytic that acts by competitive blockade of the lysine-binding sites of plasminogen, plasmin, and tissue plasminogen activator resulting in a reversible blockade that slows fibrinolysis and degradation of clots. ε-aminocaproic acid acts via a similar mechanism but is 6 to 10 times less potent than TXA.

In one prospective randomized double-blind study by Sethna et al. in 2005, blood loss was reduced by 41% in the TXA group as compared with placebo. However, the amount of blood transfused was not different.[13] TXA was administered with a 100 mg/kg bolus followed by an infusion of 10 mg/kg/h until skin closure. This study indicated three variables predictive of blood loss including preoperative platelet count, American Society of Anesthesiologists (ASA) physical status, and treatment with TXA. No adverse events related to the TXA were present. A retrospective study by Yagi et al. in 2012 demonstrated significantly less intraoperative blood loss as well as significantly fewer blood products transfused in patients who received TXA.[14]

EACA has been shown to significantly decrease postoperative wound drainage in a prospective, randomized, double-blind study by Florentino-Pineda and colleagues in 2004.[15] A retrospective case control study by Thompson and colleagues in 2008 demonstrated that EACA was highly effective in decreasing perioperative blood loss and transfusion requirements in patients with neuromuscular scoliosis undergoing spine surgery.[16] EACA can be administered by bolus of 100 mg/kg, not to exceed 5 g, over 15 minutes followed by an infusion of 10 mg/kg/h until wound closure.[17]

Based on a prospective, randomized, double-blinded comparison of TXA and EACA by Halanski et al. in 2014, TXA use was associated with lower allogenic transfusion requirements, less alteration in postoperative clotting studies, and a trend toward lower blood loss in pediatric posterior spinal fusion patients.[18] However, due to flaws in the study such as an unequal distribution of patients undergoing Ponte osteotomies, their data is not sufficient to consider TXA to be more effective than EACA. Further investigation is warranted, especially considering the significantly increased cost

associated with TXA over EACA.

Invasive arterial monitoring and frequent blood gas determinations are indicated for major orthopedic operations in order to keep close track of hemodynamics and blood loss. For any spine surgery, venous access in anticipation of large volume blood loss is essential. When peripheral access is inadequate, central access should be obtained prior to the start of the procedure since gaining access for line placement can be very difficult once the surgery has started. Significant bleeding can occur postoperatively, and patients should be monitored closely for the first few days after surgery.

SPECIFIC DISORDERS

While there are a multitude of genetic disorders, both inherited and sporadic, that have orthopedic and anesthetic implications, only a select few will be discussed in detail. Whenever one is faced with providing an anesthetic for a child with an unfamiliar disorder, at least a cursory literature review is indicated. Access to the basic components of syndromes and disorders is incredibly easy with access to the Internet. Some syndromes are known to have difficult airways, unstable cervical spines, or even predilection for malignant hyperthermia, and knowledge of the basics can be quite helpful. In many cases, parents have extensive knowledge of their child's rare condition and can be used as a resource where appropriate.

Arthrogryposis

Arthrogryposis is a term used to describe patients with joint contractures, which can result from multiple etiologies. The term derives from the Greek words "arthron" for "joint" and "gryposis" for "hooking." The joint contractures may be caused by muscular, neurological, or connective tissue anomalies. The "tes" are descriptive of patients who present with two or more congenital contractures of their joints. There are over 200 different disorders associated with arthrogryposis and approximately 1% of all births have associated joint contractures. The incidence of arthrogryposis is 1:5,000–10,000 live births and is equal between male and female infants.[19]

Arthrogryposis can be a result of intrauterine abnormalities such as fibroids or oligohydramnios and abnormalities of connective tissue development.[19] Neurological abnormalities are present in 70% to 80% of patients with arthrogryposis and are associated with patchy damage to the anterior horn cells of the spinal cord. Arthrogryposis is also associated with muscular dystrophies, intrauterine myositis, and mitochondrial diseases.

Classic arthrogryposis is called amyoplasia. It is a sporadic symmetric disorder where muscles are replaced by fibrous tissue. Multiple joints are involved including shoulders, elbows, hips, fingers, wrists, feet, and knees.[19] Distal arthrogryposis is a disorder of autosomal dominant inheritance primarily affecting the hands and feet. Typically, infants will have clenched fists with ulnar deviation of the fingers, medially overlapping fingers and club feet.[20]

Infants with arthrogryposis will generally have multiple joints involved and are best managed by a multidisciplinary team of therapists, surgeons, and primary physicians. Physical and occupational therapists can begin intervention early in the child's life with passive range of motion exercises and splinting and casting. Therapeutic goals include maximization of range of motion and function.[19]

Clubfoot deformities are common in patients with arthrogryposis. Treatment is initiated with serial casting followed by heel cord lengthening. Lifelong treatment is indicated to prevent recurrence of deformity. Treatment goals include maintenance of a stable, pain-free foot.[19]

Knee problems in arthrogryposis patients may be related to flexion, extension, subluxation, and stiffness. Occasionally, knee flexion can be associated with skin pterygiums and requires plastic Z lengthening procedures. Other flexion contractures may respond to hamstring lengthening with posterior knee capsular releases. The quadriceps muscle group is likely to be weak and easily fatigued. Quadriceps lengthening via release of the lateral and medial quadriceps and proximal detachment of the rectus femoris may need to be performed in the case of knee hyperextension.[19] Circular external fixation strategies or osteotomies and growth guidance may be used to manage knee flexion deformities. Hip dislocations are relatively common in patients with arthrogryposis. The hip joints are generally very stiff and require operative reduction about one year of age.[19]

Arthrogryposis of the upper extremities typically involves internally rotated arms, extended elbows, flexed wrists, and thumb in palm or clasp thumb deformities. Treatment begins early with splinting and occupational therapy. Surgery is generally performed between 1 and 12 months of age.[19] Maintaining elbow flexion is extremely important for independence with activities of daily living. Surgery may involve elbow release with reconstructive lengthening of the triceps and potentially muscle transfer to enable active elbow flexion. In cases where the wrist does not respond to hand therapy,

partial corpectomies may be performed. Thumb adduction may be treated with an adductor release with an opponensplasty.[19]

Scoliosis is common in children with arthrogryposis and is associated with hip dislocations and compensatory lumbar lordosis.[19] If the curve continues to progress despite thoracolumbar spinal orthosis (TLSO) bracing, surgery may be indicated. Care of spinal deformity in particular is individualized as independent mobility may depend on spine flexibility.

Children with arthrogryposis will generally present for multiple orthopedic surgeries beginning during infancy. Particular care must be taken to provide a positive experience for children and their families so as not to contribute to a negative association with the hospital and surgery experience. Children who are traumatized early in life by a forceful hold of a mask during inhalation induction may carry anxiety into every hospitalization and surgery.

In addition to the orthopedic issues, infants may have associated conditions. Of particular relevance to anesthesia are micrognathia, trismus, short neck, limited mandibular excursion, and fusion of cervical vertebrae, which can cause difficult airway intubation.[20] There is also a potential for musculoskeletal deformities that can compromise respiratory function and lead to increased risk of restrictive lung disease, postoperative respiratory insufficiency, and aspiration. In general, patients with isolated distal arthrogryposis are less likely to have a difficult airway.[20]

Intravenous access in arthrogryposis patients can present unique challenges. In addition to a relative paucity of veins, limb contractures can make mechanical access to a vein difficult. Contractures and rotational changes can also result in veins appearing in relatively unexpected areas of the extremities. The common practice of inhalation induction and subsequent intravenous cannulation in healthy children carries an increased risk of potential difficulty in arthrogryposis. Careful preoperative evaluation of venous and airway anatomy may help prevent intraoperative anesthetic complications.

Meticulous attention to positioning is important in patients with arthrogryposis because they are at increased risk of fracture from joint contractures. If the patient has an associated muscular dystrophy, succinylcholine should be avoided due to the risk of hyperkalemia. Patients with arthrogryposis are not considered to be at increased risk of malignant hyperthermia but will sometimes exhibit a hypermetabolic state under anesthesia.[20]

Juvenile Idiopathic Arthritis

Formerly known as juvenile rheumatoid arthritis, juvenile idiopathic arthritis (JIA) is the most common autoimmune disease of childhood. The worldwide incidence ranges from 0.8 to 22.6 per 100,000 children per year. Approximately 100,000 children in the United States have JIA. The subtypes of JIA include oligoarthritis, polyarthritis, and systemic JIA. While the etiology of JIA is not clearly understood, there are likely both immunological and environmental factors. Associated immunological abnormalities cause an inflammatory synovitis, which can result in joint destruction if not treated.[21]

Diagnosis of JIA is clinical, and the differential diagnosis of joint pain is vast. Generally, patients with JIA present with arthritis defined as intra-articular swelling with two of the following: limitation in range of motion, tenderness or pain with movement, and warmth.[21] The joints are generally not erythematous, although they are swollen and painful. Patients with chronic temporomandibular joint disease can develop micrognathia. Patients may have cervical spine involvement with decreased neck extension and risk of atlantoaxial subluxation. Both jaw inflammation and spine involvement can lead to a difficult airway and tenuous neck stability.

Patients with JIA may present with laboratory abnormalities including increased white blood cell count and decreased red blood cell count. Patients with systemic JIA can develop macrophage activation syndrome (MAS), a potentially fatal complication. MAS is associated with a falling platelet count, extreme hyperferritinemia, evidence of macrophage hemophagocytosis in the bone marrow, increased liver enzymes, falling leukocyte count, persistent, continuous fever greater than or equal to 38°C, hypofibrinogenemia, and hypertriglyceridemia.[21]

Children with JIA have osteopenia, as well as increased levels of cytokines, which regulate bone metabolism and can result in abnormalities of skeletal growth. These children may present to the orthopedic operating rooms for surgery for limb length discrepancy, scoliosis, joint arthroplasties, and synovectomies. They might also present for limb and mandibular osteotomies, and trauma-related fractures frequent in children. They may be at increased risk of bleeding if platelets are low or if they are on chronic nonsteroidal anti-inflammatory medications. When the disease flares, they may be treated with oral steroids, and a careful history can help elicit whether stress-dosing is indicated perioperatively.

Klippel-Feil Syndrome

Klippel-Feil syndrome is a cervical spine disorder that results in a short neck due to fusion of cervical vertebrae. The traditional clinical triad is a short neck, low posterior hairline, and limited cervical mobility, but most patients do not exhibit all three signs. The incidence of Klippel-Feil syndrome is approximately 1:42,000 with a slight predilection for girls. Klippel-Feil may also be a phenotypic presentation associated with several other disorders including syndromes such as Goldenhar, Mohr, VACTERL (vertebral defects, anal atresia, cardiac defects, tracheo-esophageal fistula, renal anomalies, and limb abnormalities), and fetal alcohol.[22,23] The cervical spine in Klippel-Feil syndrome may also have occipitocervical synostosis, odontoid abnormalities, and the proximal migration of the C2 vertebra known as basilar impression.[23]

In addition to the clinical triad, patients with Klippel-Feil syndrome may have sensorineural or conductive hearing loss, cleft lip, oligodontia, micrognathia, a webbed neck, and torticollis. They may have voice abnormalities due to malformed laryngeal cartilage. Congenital cardiac defects, including ventriculoseptal defect (VSD) and conduction delays, may be present (5% to 10% incidence). Many patients have winged scapula, vertebral anomalies including scoliosis, and renal abnormalities (double collecting systems, renal aplasia, and horseshoe kidney).[22-24]

Because the cervical spine becomes more immobile over time, the airway can become progressively more difficult to intubate. Therefore, a history of an uneventful anesthetic is not as reassuring as it would be in other patients with potentially difficult airways. Additionally, the cervical spine becomes hypermobile at levels above and below immobile joints causing increased risk of cervical spine injury with direct laryngoscopy. Seemingly minimal movement of the cervical spine under anesthesia can result in serious neurological sequelae and patients should be evaluated for neurological deficits and previously unidentified deficits should be investigated. For cooperative patients, the awake fiberoptic intubation has been the gold standard in airway management. However, most children are not able to tolerate the awake intubation and thus asleep fiberoptic intubation with spontaneous ventilation is appropriate. The widespread availability of the video laryngoscope in conjunction with in-line stabilization has enabled a view greater than that given by a traditional laryngoscope with minimal neck extension.

Postoperatively, patients with significant scoliosis have a higher risk of respiratory failure and may need to remain intubated in which case plans must be in place to maintain neutral neck position until the patient is fully awake. Patients with renal insufficiency may have a prolonged response to medications with renal clearance such as nondepolarizing neuromuscular blockers.

Skeletal Dysplasias

Achondroplasia and Dwarfism

Achondroplasia is the most common cause of short-limbed dwarfism.[25] It is an autosomal dominant disorder characterized by a disproportionately short stature. The incidence ranges between 1:15,000 and 1:40,000 births.[26] The clinical features result from abnormal cartilage formation especially at the epiphyseal growth plates. Patients with achondroplasia have a large head with a prominent forehead, depressed nasal bridge, prominent mandible, normal trunk, and short limbs. They can develop lordosis, thoracolumbar kyphosis, pelvic narrowing, and severe spinal stenosis. The spinal stenosis may present with cervical cord or thoracolumbar cord compression and cauda equina syndrome.[27]

Clinically, motor milestones are delayed in infants with achondroplasia due to hypotonia and the difficulty associated with balancing their disproportionately large heads. However, intelligence is normal in most achondroplastic dwarfs.

Patients with achondroplasia have comorbidities that include hydrocephalus, atlantoaxial instability, cleft palate, tracheomalacia, congenital heart disease, obstructive sleep apnea (OSA), pulmonary hypertension, and seizure disorders.[27] They may present to an orthopedic surgeon for repair of clubfoot, angular deformity, scoliosis, or spinal stenosis in addition to any of the common orthopedic complaints affecting children. Major preoperative issues include the potential for difficult airway management, cervical spine instability, abnormal respiratory mechanics, and OSA.

In patients with congenital heart disease, preoperative ECG, echocardiogram, and chest radiograph may be helpful. Patients with severe scoliosis are at risk for restrictive lung disease and pulmonary hypertension, further warranting a preoperative echocardiogram.

Intraoperatively, the anesthesiologist should take extreme care with positioning due to the potential for spinal cord injury and be cognizant of the potential for difficult airway and potential for subglottic stenosis, laryngomalacia, and tracheomalacia.

Postoperatively, patients are at risk for respiratory failure due to sleep apnea and the associated sensitivity to narcotic pain medications, as well as the altered pulmonary mechanics due to the lack of chest wall compliance. Nonopioid analgesics and regional anesthetics should be utilized whenever

possible. Local anesthetic volume should be reduced in patients with achondroplasia undergoing neuraxial blocks due to their shorter height.

Osteogenesis Imperfecta

Also known as "brittle bone disease," osteogenesis imperfecta (OI) is a disorder of collagen formation. The defect in collagen may be qualitative or quantitative, either of which results in bones that are extremely fragile. The disease ranges in severity from mild to severe and incompatible with life. Children with the mild form of the disease have presented with multiple fractures during their childhood and their presentation can be mistakenly identified as resulting from child abuse.

Osteogenesis Imperfecta	Disease Severity	Inheritance	Manifestations
Type I	Mild	Autosomal dominant	Blue sclerae, dentinogenesis imperfecta, childhood fractures
Type II	Severe	Autosomal dominant or recessive	Stillborn or death in first year of life
Type III	Severe	Autosomal dominant or recessive	Fractures in utero, "popcorn" calcifications
Type IV	Moderately severe	Autosomal dominant	Fractures in utero, bowing of long bones

Type I collagen is the basis of ligament, tendon, and bone formation. Defective structural collagen leads to compromised endochondral and intramembranous bone, poor ligament and tendon formation, and thin bone trabeculae.[27] Historically, OI was considered to be a triad of bone fragility, blue sclerae, and early deafness, but the complete triad does not hold true for most patients.[28]

OI type I is generally a mild form of the disease. In addition to recurrent fractures, blue sclerae, and early-onset hearing loss, children may exhibit hypermobile joints, easy bruising, thin skin, scoliosis, hernias, and mild short stature.[28] They may have dentinogenesis imperfecta, which is a disorder of tooth development resulting in discoloration and fragility of the dentition. Most patients with OI type I will have fractures in childhood but not as neonates. There is a decrease in fracture risk postpuberty but a resurgence in postmenopausal women.[29] Inheritance is autosomal dominant.

OI type II is generally a severe form of the disease and patients are either stillborn or die in the first year of life. Fractures may present in utero and the bones are extremely fragile. A small thorax contributes to respiratory insufficiency. There are abnormalities of the cerebral cortex including agyria, gliosis, and periventricular leukomalacia.[28] Inheritance may be either autosomal dominant or autosomal recessive.

Type III OI is the most severe, nonlethal form of the disease. Children will present at birth with fractures that occurred in utero. Postnatal fractures occur easily and heal poorly. There may be a "popcorn" appearance of the bones at the metaphyses.[28] Abnormalities of the thorax can result in respiratory insufficiency and patients can develop scoliosis and vertebral fractures. Patients will have an extreme short stature. They may have scleral changes, dentinogenesis imperfecta, hearing loss, and kyphoscoliosis.[28] Inheritance is either autosomal dominant or recessive.

Patients with type IV OI may also present with fractures in utero and bowing of the long bones. This type of OI is considered moderately severe and few patients will achieve independent ambulation.[28] Inheritance is autosomal dominant. OI types V to XI have been described and are beyond the scope of this text. They account for varying severity of bone fragility with and without the above-listed symptoms of blue sclerae, dentinogenesis imperfecta, ligamentous laxity, and rhizomelia.[28]

Clinically, patients will present with multiple fractures, may develop scoliosis due to decreased strength of the ligaments, compression fractures, osteoporosis, and spondylolisthesis,[27] and have associated congenital heart disease [aortic root dilation, patent ductus arteriosus (PDA), atrial septal defect (ASD), VSD, and valvular defects].[27,29]

Patients with OI may present for elective, urgent, or emergent procedures. When possible, the preoperative evaluation should include investigation of associated cardiac anomalies with an echocardiogram. Patients who have severe kyphoscoliosis or other abnormalities of the thorax should be evaluated for restrictive lung disease. In patients with severe thoracic abnormalities there is a risk of cor pulmonale. The possibility of basilar

impression, atlantoaxial instability, and cervical cord compression should not be overlooked.[27] Although not yet clearly characterized, there is a potential for platelet dysfunction.[29]

Intraoperatively, all pressure points must be padded, and great care must be taken to avoid fractures and injury to tendons and ligaments during positioning and throughout the surgery. Fractures from succinylcholine-induced fasciculations are possible and thus depolarizing neuromuscular blockade is relatively contraindicated. Fractures can also result from direct laryngoscopy and blood pressure cuffs. It may be appropriate to place an arterial catheter for longer operations and possibly avoid blood pressure cuff use for shorter operations given that the risk may outweigh the benefit. Patients with dentinogenesis imperfecta have a higher risk of dental damage.

Patients with OI are known to exhibit signs consistent with a hypermetabolic state such as hyperthermia but without the typical manifestations of malignant hyperthermia; to date there is no association with this potentially lethal disease.

Emergence of anesthesia should be as smooth as possible given the risk of fracture with agitation. This is of particular relevance in patients emerging from posterior spinal fusion due to the fact that they are at risk of dislodging hardware and injuring the spinal cord.

Osteopetrosis

Osteopetrosis is a bone disorder in which bones are sclerotic with increased density. It is also known as marble bone disease. It results from insufficient resorption of bone by osteoclasts. It can be inherited in an autosomal dominant or autosomal recessive manner.

The autosomal recessive form is severe and presents with an incidence of 1 in 250,000 births.[30] Generally, patients with this form will present in infancy with macrocephaly, hepatosplenomegaly, deafness, blindness, and severe anemia. There is bone sclerosis throughout and laboratory evaluation shows low calcium and phosphorus levels with elevated parathyroid hormone levels. Survival may extend into the second decade, but likely children will have progressive cranial neuropathies, anemia, pathological fractures, and dental problems.[30]

The autosomal dominant form is mild and is much more frequent with an incidence of 1 in 20,000 births.[30] The body's failure to remodel growing bone results in narrowing of cranial nerve foramina and a decrease in functional bone marrow. Clinically, children with develop cranial nerve palsies, anemia, and compensatory extramedullary hematopoiesis in the liver and spleen.[30] They are also prone to pathological fractures and for this reason may present with some frequency to the orthopedic operating room.

Preoperatively, hematocrit and platelet count should be evaluated due to the potential for pancytopenia and hemorrhage. A calcium level may be indicated as well due to the risk of hypocalcemia-related seizures or tetany. Patients may have abnormal cells of immunity (white blood cells and macrophages) and are prone to infection. In more severe cases, infection may be the cause of demise in childhood. Patients may be on chronic steroids and stress doses of steroids should be considered.

Patients with osteopetrosis are at risk of difficult airway due to limitation of the mandibular movement, and restricted size of the oropharynx and nasal pharynx. Nasal intubation may be contraindicated due to the risk of bone growth limiting the size of the nasal cavity.[31] The risk of dental damage is also increased due to poor formation of teeth and higher likelihood of osteomyelitis of the mandible. Patients are at a greater risk of obstructive sleep apnea and considerations should be taken for increased sensitivity to narcotics and higher incidence of postoperative respiratory complications. Positioning with great care is of extreme importance due to the risk of pathological fractures intraoperatively.

Connective Tissue Disorders

Marfan Syndrome

Marfan syndrome is a connective tissue disease resulting from a defect in fibrillin, a major component of the connective tissue. Inheritance is autosomal dominant with high penetrance and incomplete expressivity, which results in a great degree of variability in presentation.[32] The incidence is 1 in 10,000 live births with approximately one-quarter of the cases sporadic.[33] Mutation in fibrillin-1 on chromosome 15 is present in 66% to 91% of cases; therefore, the inability to detect an abnormality on fibrillin-1 does not exclude the diagnosis.[34]

Patients with Marfan syndrome generally have skeletal, cardiovascular, and ocular pathology. They are tall with joint laxity and may have a wingspan

greater than their height. They may have pectus excavatum or pectus carinatum. Their fingers are long and slender (arachnodactyly). Their heads are long and narrow (dolichocephaly) with enophthalmos, retrognathia or micrognathia, malar hypoplasia, a high arched palate, and downward slanting palpebral fissures.[33] They may have the "wrist sign," which is when the thumb overlaps the fifth finger when grasping the contralateral wrist, as well as the "thumb sign," which is when the thumb extends beyond the ulnar border of the hand when overlapped by the fingers. Other orthopedic considerations include scoliosis, kyphosis, chest asymmetry, hind foot deformity, severe flatfoot, acetabular protrusion, and reduced elbow extension.[34]

More than 50% of patients with Marfan syndrome develop scoliosis, but only 10% to 20% of those who develop it need treatment.[10] Generally, patients with Marfan syndrome have either atypical scoliosis or typical scoliosis with atypical features.[35] Atypical curve patterns include left thoracic curves, whereas right thoracic curves are more typical and thus more common. The natural history of scoliosis in patients with Marfan syndrome is more severe than in patients with adolescent idiopathic scoliosis (AIS).

The cardiovascular abnormalities associated with Marfan syndrome include mitral valve prolapse (MVP) with mitral insufficiency (MI), aortic insufficiency (AI), and a tendency toward aortic dissection and dilation of the ascending aorta. Cardiovascular symptoms may be present very early in life and any patient presenting for surgery should have a preoperative cardiac workup including a focused history, ECG, and echocardiography. Aortic dissection is the main cause of death in patients with undetected Marfan syndrome, and early diagnosis with treatment can prolong disease-free survival. Beta-blockers are an essential treatment because they have been shown to decrease the rate of aortic root dilation.[36]

From a pulmonary perspective, patients with Marfan syndrome are more prone to blebs and pneumothoraces. They have lower forced vital capacity due to the connective tissue abnormality and early airway closure. Chest deformities and scoliosis can impact vital capacity and total lung capacity and result in restrictive respiratory dysfunction. Pectus excavatum may result in cardiorespiratory dysfunction with dyspnea, decreased endurance, chest pain, and tachycardia.[37]

There is a high incidence of obstructive sleep apnea in patients with Marfan syndrome relative to the normal population. Untreated OSA may lead to an increased risk of aortic events, which may explain why patients with Marfan syndrome with OSA have a worse prognosis.[38] The etiology of the high incidence of OSA in these patients may be related to craniofacial abnormalities, tissue laxity, and high nasal airway resistance.[39] Multiple episodes of upper airway collapse followed by arousal cause additional stress on the aorta, which may lead to more rapid development of aortic dilation.[39]

Neurologically, patients are prone to dural ectasia, which is a widening of the dural sac. While the abnormality can occur anywhere along the spinal column, it is most likely to present in the lumbosacral region. There may be thinning of the cortex of the pedicles and laminae of the vertebrae and there may be an associated meningocele.[40] Essentially, there is a disparity between the diameters of the dural sac and the vertebral body of the associated spine segment. The possibility of dural ectasia must be considered when planning neuraxial anesthesia. One population-based study demonstrated a prevalence of 90% for dural ectasia in patients with Marfan syndrome.[41]

The orthopedic operations for which patients with Marfan syndrome are most likely to present include scoliosis procedures and operations for foot deformity, angular deformity, and patella instability. In addition to a complete history and physical, cardiac workup may be indicated, and beta-blockers should be continued perioperatively. The patient's neck should be evaluated clinically and radiographically for atlantoaxial instability.

Intraoperatively, stable hemodynamics are essential. Maintaining low heart rate and avoiding hypertension can protect against acute aortic dissection intraoperatively and myocardial ischemia. Patients may have a higher requirement for neuraxial local anesthetics due to their height. Positioning must be done with great care due to joint laxity and potential for injury.

Ehlers-Danlos Syndrome

Ehlers-Danlos syndrome (EDS) is a diagnosis applied to a heterogenous group of connective tissue disorders characterized by joint hypermobility, skin laxity, and potential fragility of vascular structures. The incidence is estimated to be between 1:10,000 and 1:25,000 with an equal distribution between sexes and different ethnicities.

While as many as 10 subtypes of EDS have been described, the most commonly used nosology delineates 6 categories of disease pattern.[42] Most cases appear to be inherited in an autosomal dominant fashion and some are caused by mutations in genes encoding collagen type, or proteins that regulate

collagen synthesis. Extreme variability in expression results in a broad range of clinical manifestations from insignificant to life-threatening. Table 22-1 provides an overview of the six categories and their characteristic profiles (modified from Weismann et al.).[43]

Table 22-1

Ehlers-Danlos Syndrome Types

Type	Major Criteria	Minor Criteria
Classic	Joint hypermobility Skin hyperextensibility Atrophic scars	Smooth, velvety skin Easy bruising Muscle hypotonia Motor delay Positive family history
Hypermobility	Generalized joint hypermobility Skin hyperextensibility	Recurring joint dislocations Chronic joint/limb pain Positive family history
Vascular	Translucent thin skin Arterial/intestinal rupture Extensive bruising Characteristic facies	Acrogeria Hypermobility of small joints Tendon and muscle rupture Club feet Arteriovenous, carotid-cavernous sinus fistula Pneumothorax/hemothorax Positive family history, sudden death in close relative
Kyphoscoliotic	Congenital and progressive scoliosis Congenital hypotonia Scleral fragility and rupture of the ocular globe	Tissue fragility with easy bruising Arterial rupture Marfanoid habitus Microcornea Osteopenia/porosis Positive family history
Arthrochalasis	Congenital bilateral hip dislocations Generalized joint hypermobility with recurrent subluxations	Skin hyperextensibility Tissue fragility Easy bruising Hypotonia Kyphoscoliosis Osteopenia/porosis
Dermatosparaxis	Severe skin fragility Sagging, redundant skin	Doughy skin texture Easy bruising Large hernias

The classic and hypermobility types combinedly account for approximately 75% of EDS cases. These patients suffer frequent complications of joint

hypermobility with recurrent dislocations as well as chronic joint and limb pain, which can result in the frequent need for pediatric orthopedic care. Vascular-type EDS children account for approximately 5% of cases. They commonly have club feet as well as tendon and muscle ruptures. The hallmark vascular and intestinal fragility can lead to catastrophic rupture. Children with kyphoscoliotic-type EDS frequently have congenital hypotonia and ocular fragility in addition to congenital and progressive scoliosis. Arthrochalasis-type EDS patients have an increased incidence of congenital bilateral hip dislocation and a propensity for recurrent subluxations. Dermatosparaxis-type patients have multiple skin-related features including redundancy, doughy texture, and hyperextensibility, but have fewer of the more severe traits seen in other EDS categories.

When an EDS child presents for surgery, particular attention should be given to bleeding tendencies, complications from past operations, and airway management issues. Further testing is dictated by the patient's history and the expected surgical risk. One should always have a low threshold for preparing blood products for EDS patients undergoing surgery. When formulating an anesthetic plan for an EDS patient, serious consideration of the risk/benefit ratio for every monitor and every type of access must be undertaken. Any procedure that requires a needle puncture carries with it a significantly increased risk of complication. Preemptive use of ultrasound for venous and arterial access is recommended as is utilizing the most experienced practitioner available. Even the interval time of automatic blood pressure devices and the tightness with which a pulse oximeter probe is applied can lead to excessive bruising or hemorrhage. Preoperative discussion with the surgical team should address the need for tourniquets, foley catheters, gastric tubes, and any other invasive device. The preprocedure time out should also be used to ensure that everyone in the operating room is aware of the risks unique to EDS surgical patients. Additionally, extreme care must be taken with positioning and padding of all areas of the patient since joints can easily be dislocated or hyperextended, bruising can result from minimal pressure, and ocular and globe rupture can occur in prone positioning.

General anesthesia with volatile anesthetic or propofol is acceptable for EDS patients. Drugs that affect platelets should be either avoided or used with caution. Neuraxial blocks should be performed with caution or avoided due to the increased risk of hematoma formation. Regional blocks are an acceptable adjuvant therapy as long as ultrasound guidance is employed. Infiltration of incision sites with local anesthetics as well as locally injected anesthetics for dental procedures have been reported to have reduced efficacy in EDS patients.[44,45] Desmopressin (DDAVP) can be used prophylactically for procedures where large volume blood loss is a possibility. While the mechanism of effect is uncertain, studies of small groups of patients have demonstrated significant reductions in blood loss and related complications when desmopressin was given.[46]

During emergence, padding, and positioning must be meticulously maintained. Postoperatively, patients with casts, splints, and external fixation devices must be closely observed for signs of compartment syndrome. Postoperative nausea and vomiting (PONV) should be aggressively prevented as esophageal rupture has been reported during the recovery period.[47] When possible, EDS patients should be cared for in specialized facilities and monitored for at least 24 hours after surgery.

Neuromuscular Diseases

Cerebral Palsy

Cerebral palsy (CP) is a disorder of motor function that results from an irreversible insult to the developing brain. The brain damage causing CP may be due to a multitude of causes including infection, ischemia, and metabolic and genetic perturbations. Less than 10% of cases of CP are due to birth trauma, contrary to former beliefs.[48] In addition to a disorder of movement, patients may have difficulty with sensation, perception, cognition, communication, and behavior though many patients are of normal intelligence and function at a high level.[49]

The incidence of CP ranges from 1.5 to more than 4 per 1,000 live births worldwide. Approximately 1 in 323 children has CP. Most children with CP have spastic CP and 58.7% of children with CP can walk independently. Forty-one percent of children have concurrent epilepsy and nearly 7% have concurrent autism spectrum disorders.[48]

Risk factors for CP include low birth weight and premature birth as well as multiple birth pregnancies.[48] Preterm infants with intracerebral hemorrhage and periventricular leukomalacia are also at increased risk. Boys are at a higher risk than girls and are more likely to have a more severe degree of impairment.[49]

Symptoms of CP are highly variable between individuals and manifest in early childhood or infancy. CP is nonprogressive, chronic, and not curable. Patients present for multiple orthopedic procedures due to spasticity, extremity contractures, scoliosis, and hip dislocation.

Different types of CP include spastic diplegia, spastic quadriplegia, spastic hemiplegia, and dyskinetic CP. The most severe form is spastic quadriplegia, which accounts for approximately 20% of cases.[49] There is a higher association of intellectual disability with this form as well as seizures and swallowing difficulties leading to recurrent aspiration pneumonias. Patients have increased tone and spasticity in all four extremities, decreased spontaneous movements, and brisk reflexes.

Spastic hemiplegia refers to patients with CP who have increased muscle tone and spasticity in the upper and lower extremities of either the left or right side of their bodies. Generally, the arm is more affected than the leg. Approximately one-third of patients with spastic hemiplegia have a seizure disorder and 25% have cognitive abnormalities.[49] Spastic hemiplegia may result from an intrauterine stroke or infection as well as other causes. Spastic diplegia is spasticity of both legs more so than the arms. It is associated with damage to the immature white matter in utero. Up to 70% of these children have periventricular leukomalacia.[49] The likelihood of seizure is far less than with spastic hemiplegia and these children are more likely to have normal intellect.

Dyskinetic CP, also known as athetoid, choreoathetoid, or extrapyramidal CP, accounts for approximately 20% of patients with CP. Infants are hypotonic with poor head control. Generally, the upper extremities are more affected than the lower extremities and patients will often have prominent tongue thrusting and drooling. Their movements may be athetoid (slow and writhing), choreoathetoid (jerky), or dystonic. These children are likely to have normal intellect but will experience difficulty with speech. Causes of dyskinetic CP include intrauterine or birth asphyxia, kernicterus, and metabolic disorders. There is cognitive impairment in approximately 50% of these children and 25% have a seizure disorder. In addition to chronic pain, they may develop hip displacement and contractures.[49]

Orthopedic procedures for which patients with CP will come to the operating room include injection of botulinum toxin (botox), release of the hip including adductor tenotomy or psoas transfer and release, hip reconstruction, rhizotomy, heel cord lengthening, scoliosis, and placement or change of intrathecal baclofen pumps.

There are no specific anesthesia contraindications for patients with CP. Succinylcholine is not more likely to cause hyperkalemia than in an otherwise healthy child and they are not at increased risk of developing malignant hyperthermia. Anesthesia will most often be dictated by the specific procedure, although many anesthesiologists have a low threshold for intubation in patients with copious oral secretions due to the increased risk of laryngospasm.

It is essential to remember that while many patients are limited in their ability to communicate with us, they are often not limited in their ability to understand. The caregiver is often a very helpful translator and can help identify anxiety and pain on behalf of the patient. Any anesthetic should be explained to the child in age-appropriate terms with reassuring tones. Many CP patients frequent the operating room and it is prudent to allow the patient and parent to have input into the anesthetic care, particularly with regard to intravenous placement prior to, or after, induction.

Short cases such as botox injections to spastic limbs often require only a mask anesthetic. In children who are otherwise healthy, this can safely be performed without the placement of an IV catheter. However, there are various reasons for placing an IV catheter in this situation, including signs of upper respiratory infection, excessive oral secretions, and concern for laryngospasm as well as access for medications such as ondansetron to help prevent PONV. Children with CP require a lower concentration of volatile anesthetic and have a longer emergence time than their healthy peers.[50]

Longer operations often require placing an endotracheal tube. Patients should be given their regular medications such as baclofen for spasticity and antiepileptics for seizure prevention preoperatively. Patients with CP have low pharyngeal tone and a high incidence of gastroesophageal reflux disease, and the endotracheal tube can help protect against aspiration. They have variable responses to neuromuscular relaxing drugs. Patients who take antiepileptic medications may be relatively insensitive to muscle relaxants due to induction of the cytochrome P-450 enzymes and other patients may have a prolonged recovery time. Extubation should be done with the patient completely reversed from neuromuscular blockade and as recovered from the anesthetic as possible.

Positioning for any procedure on a child with CP may be difficult due to multiple contractures. It is important to pad any bony surfaces to prevent pressure ulceration and nerve injury. The more painful orthopedic surgeries present difficulty with postoperative pain management. When indicated, regional anesthesia can be extremely helpful. Diazepam 0.1 mg/kg intravenously (or orally when tolerated postoperatively) treats muscle spasms and is an adjunct to pain management.

Duchenne Muscular Dystrophy

Muscular dystrophies are a heterogenous group of hereditary diseases of muscle, which present with progressive weakness. They are characterized by painless degeneration and atrophy of the skeletal muscles without denervation. Over time, muscle fibers are replaced with fibrous and fatty connective tissue. The defect is due to an absence of dystrophin, which results in failed integrity of the skeletal muscle membrane. This causes breakdown of the sarcolemma and an influx of extracellular calcium, activation of cellular proteases, inflammation, necrosis, and fibrotic infiltration.[51]

Duchenne muscular dystrophy (DMD) is the most common muscular dystrophy and has an incidence of approximately 1 in 3,500 births. It is an X-linked recessive disorder primarily affecting boys. There are documented cases in females with a range in severity of clinical course. Diagnosis is by proximal limb muscle biopsy with staining, which demonstrates an absence of dystrophin at the surface of the muscle fibers.[51]

Patients with DMD present with a waddling gait, frequent falls and difficulty climbing stairs, due to proximal muscle weakness. Calves hypertrophy, despite loss of functional muscle, is due to fatty infiltration. Combined with the proximal muscle weakness, pelvic girdle weakness causes “Gower’s sign,” which is when a child climbs up his legs to reach a standing position. Weakness in the shoulder girdle and thoracic muscles leads to thoracolumbar scoliosis for which these patients may require orthopedic surgery.

Generally, children will have normal development until 3 to 5 years of age at which point the muscle weakness becomes apparent and motor milestones may be lost. While the clinical course is variable, most children are unable to walk by 9 to 11 years of age.

Of importance in anesthesia is the development of dilated cardiomyopathy, which can occur at a very young age. Loss of dystrophin in the heart affects the L-type calcium channels resulting in increased intracellular calcium. This causes activation of proteases that degrade the contractile proteins leading to inflammation, myocardial cell death, and fibrosis. Cardiomyopathy is a major cause of death in patients with DMD and all patients should undergo a cardiac evaluation with echocardiogram or cardiac MRI prior to an elective anesthetic.[51] When patients present emergently, dilated cardiomyopathy with poor cardiac function should be presumed.

Patients with DMD are also at risk of postoperative respiratory failure. In severe cases, an intubation for an operation may even become a terminal intubation. All anesthetics should be performed at centers where pediatric intensive care is readily available. By the end of the first decade of life, pulmonary function tests (PFTs) will demonstrate decreases in inspiration, expiration, vital capacity, and total lung capacity, which may not be easily identified in the patient’s history given poor exercise tolerance overall. There is benefit to extubating to Bi-Pap or CPAP. Preoperative preparation with noninvasive positive pressure ventilation and cough-assist devices can help facilitate transition back to independent respiratory effort.

The other major anesthetic consideration with DMD patients is the risk of life-threatening hyperkalemia in response to succinylcholine administration. Any paralytic agent should be given with caution given the underlying weakness in patients with DMD because they are more likely to have an increased response to nondepolarizing muscle relaxants in both peak effect and duration of action. Traditionally, volatile anesthetics were avoided in patients with DMD due to potential of triggering malignant hyperthermia. However, they are not contraindicated and can be used provided the patient’s hemodynamics are maintained.

Spinal Muscular Atrophies

Spinal muscular atrophies (SMAs) are degenerative diseases of the anterior horn cells and lower brainstem nuclei that result in diffuse proximal muscle weakness. The degeneration begins in utero and continues to progress throughout infancy and childhood with varying degrees of severity. The incidence is approximately 1 in 6,000 to 1 in 10,000 people.[52]

SMA is characterized by hypotonia, hyporeflexia, and overall weakness especially in the lower extremities.

SMA[22]	Age of Onset	Disease Progression
Type 0	Neonatal period	Fatal during that period
Type 1: Werdnig-Hoffmann	Infantile	Respiratory failure, first year of life
Type 2	Late infantile	Most common (50% of SMA patients), slowly progressive
Type 3: Kugelberg-Welander	Juvenile	Chronic
Type 4	Adult	Slowly progressive

Most cases of SMA are autosomal recessive with a small minority of sporadic cases. They are caused by mutations or deletions of the survivor motor neuron 1 (*SMN-1*) gene on chromosome 5q. In some cases, survival motor neuron 2 (*SMN-2*) can take over function of the *SMN-1* gene, which may account for some of the variability in clinical severity of SMA.[22]

Treatment of SMA is mostly supportive involving chest physiotherapy, respiratory support, prevention of aspiration, and supplemental nutrition. Orthopedic intervention is often indicated for joint contractures including hip subluxations and dislocations as well as contractures and hypermobile joints of the upper extremities. Patients with SMA may present with spontaneous fractures due to osteopenia. Nearly all nonambulatory patients with SMA develop scoliosis with severe progression.[53]

Pulmonary compromise is present in children with SMA and patients are at increased risk of perioperative respiratory failure with decreased vital capacity and severe weakness of auxiliary respiratory muscles. Patients have difficulty clearing secretions and are prone to development of pulmonary infections, hypoventilation, and atelectasis. Some pulmonary function can be spared with early intervention for scoliosis.[53] Perioperatively, noninvasive positive pressure ventilation should be available and mechanical cough-assist devices may be helpful in preventing postoperative atelectasis and pneumonia.

Poor nutrition is a significant problem for patients with SMA and can increase perioperative complications. Some surgeons may opt to bolster nutritional status with gastric feedings or total parenteral nutrition (TPN) prior to major operations for scoliosis. Prolonged fasting should be avoided due to mitochondrial dysfunction with fatty acid oxidation.[53] Patients will also decondition very quickly over the course of a prolonged hospital stay, and physical therapists and occupational therapists are essential team members.

Anesthetics for patients with SMA should be considered at facilities with appropriate postoperative pediatric intensive care availability. Patients may require prolonged hospitalization especially after major scoliosis surgery. While there isn't a strong association between SMA and cardiomyopathy, preoperative echocardiogram may be indicated in patients with severe sleep apnea.

While most children do not present for surgery with advance directives in place, there is a potential for failed ventilatory wean after small operations for gastrostomy tube placement, which may result in a transition to comfort measures and hospice care.[54] Patients with SMA do not have an increased risk of cognitive disability and should be included in formulation of care plans, including the potential transition to comfort measures, as is age-appropriate. Avoiding hypoventilation and impairment of the respiratory drive is as critical as obtaining good pain control postoperatively.

In addition to narcotic and inhalation agent sensitivity, patients with SMA may have increased sensitivity to nondepolarizing neuromuscular blockers due to decreased choline acetyltransferase, which is a result of anterior horn cell degeneration.[22] This can result in a prolonged duration of action with otherwise short-acting muscle relaxants. Additionally, the nerve stimulator may not provide an accurate indication of recovery of neuromuscular function and thus muscle relaxants should be used with extreme caution. While succinylcholine has been used without incident in some patients with SMA, there is a potential for rhabdomyolysis and hyperkalemia due to the lower motor neuron denervation hypersensitivity and immobilization.[54]

The airway may be difficult to intubate in some patients with SMA due to atrophy of the masseter and other muscles of mastication. Regional anesthetics can be used, but paralysis of any muscles involved in respiration should be avoided. To decrease narcotic requirements postoperatively,

surgeons should be asked to infiltrate the field with local anesthetic when appropriate.

The use of nusinersen (Spinraza, intrathecal administration), an SMN-2 antisense oligonucleotide, has provided an option to modify the disease course in patients with SMA with improvement in motor function and survival. Its current price tag in the United States makes it prohibitive for some patients.[55]

Neurofibromatosis

Neurofibromatosis type 1 (NF-1), the most prevalent type of neurofibromatosis, may have associated orthopedic considerations. It is an autosomal dominant condition affecting 1 in 3,000 live births.[56] The hallmark characteristic is multiple café au lait spots. Diagnosis is made via clinical features or genetic testing.

While neurofibromas generally involve the skin, they can form in many other places including peripheral nerves, blood vessels, viscera, and bone. Plexiform neurofibromas can result in deformity or overgrowth of a bone. Patients may develop sphenoid dysplasia, which is an osseous lesion or cortical thinning of long bones that may be associated with pseudoarthrosis. Congenital pseudoarthrosis is a spontaneous fracture that progresses to nonunion. It most often involves the tibia and the radius and severity can range from asymptomatic to severe requiring limb amputation.[57]

Children with NF-1 may have central nervous system abnormalities including learning disabilities and seizure disorders. Approximately 3% of patients will go on to develop malignant neoplasms and 10% of patients will develop scoliosis.[56] There is some association of NF-1 with pheochromocytoma, although this is rarely relevant to children. More likely, hypertension may develop in response to renal artery stenosis. Children can develop Moyamoya syndrome as a result of cerebral artery dysplasia, which can result in cerebral infarction and hemorrhage.

Patients may develop airway compromise from neurofibromas of the laryngeal, cervical, or mediastinal regions. Further respiratory compromise can result from severe kyphoscoliosis that remains untreated. In addition to airway considerations, anesthetic management for patients with neurofibromatosis depends on the underlying condition of presentation. Additionally, any signs or symptoms of increased intracranial pressure should be evaluated due to the possibility of intracranial lesions. Patients may have a variable response to both depolarizing and nondepolarizing neuromuscular relaxants.[57]

Neurofibromatosis-2 (NF-2) is a distinct entity from NF-1 and involves chromosome 22, as opposed to chromosome 17. Most patients will present in the second or third decade of life with acoustic schwannomas resulting in hearing loss, vestibular symptoms, or facial weakness.

Selected Lower Extremity Conditions

Blount Disease

Blount disease is a form of idiopathic tibia vara, more commonly known as “bowlegs.” It is a result of abnormal endochondral ossification of the medial aspect of the proximal tibial physis leading to varus angulation and medial rotation of the tibia.[58] The infantile (age 1 to 3 years) type is most commonly seen in black females and the juvenile and adolescent types are more common in black males. Many patients are obese, although there has not been any causality described.

Young children with mild Blount disease are given a trial of orthotic management for up to one year. More severe disease and deformity that do not respond to noninvasive management are treated with surgery. The usual procedures are proximal tibial valgus osteotomies and fibular diaphyseal osteotomies. When the disease is severe, correction may also be indicated.

The anesthetic for patients with Blount disease may be complicated by the patient’s underlying obesity, making airway, intravenous access, and positioning a challenge. The surgeon will usually inject local anesthetic and opioids with nonopioid adjuncts supporting postoperative pain management.

Slipped Capital Femoral Epiphysis

Slipped capital femoral epiphysis (SCFE) is a disorder of the hip in which the femoral head is displaced through the growth plate. It is most likely to present in patients during rapid growth in adolescence. Associated pain is in the groin, hip, or knee. The incidence of SCFE varies geographically with a

much higher incidence in the northeastern United States (10 per 100,000) than in Japan (0.2 per 100,000). Obesity is a risk factor with approximately 60% of patients in the 90th percentile for weight.[59] Males are more likely than females to develop SCFE and there may be as many as 60% who have the disorder bilaterally, even when symptoms are only one-sided.[59]

SCFE may be acute, chronic, or acute-on-chronic. Acute SCFE occurs when a patient presents with 3 weeks or less of pain. It may be associated with a relatively minor injury. Patients with chronic SCFE present after a few months of pain, which may be vague in nature. Acute-on-chronic SCFE occurs in patients who have had greater than 3 weeks of prodromal symptoms and present with an acute exacerbation of the underlying pain. Radiographically, there is associated femoral neck remodeling with displacement of the capital epiphysis past the femoral neck.[59]

The most serious complications of SCFE are osteonecrosis and chondrolysis. Osteonecrosis is avascular necrosis and occurs due to injury to the blood supply of the femoral head. Chondrolysis is an acute loss of the articular cartilage in the hip. Patients who are unable to ambulate are considered to have unstable SCFE and are at greater risk of osteonecrosis of the hip. Those with stable SCFE ambulate and have a negligible risk of osteonecrosis.[59]

Treatment for SCFE involves pinning of the hip to prevent further progression of the slip and to stabilize the physis. The hip is not reduced due to the increased risk of osteonecrosis. Concerns of bilateral disease progression may lead to surgical intervention (prophylactically) on the contralateral side even with unilateral presentation.

Anesthesia for patients with SCFE may be urgent or emergent. Patients may have to be admitted to the hospital upon diagnosis and stabilized as soon as possible. Preoperative evaluation is dictated by whether or not the patient has any medical comorbidities. General anesthesia with an endotracheal tube is the preferred method of anesthesia to provide adequate muscle relaxation. Extubation at the end of the case, dictated by any additional comorbidities, is expected. Pain is usually managed with local anesthesia injected by the surgeon as well as intravenous pain medications both narcotic and adjuncts. Postoperative pain is generally not severe.

Clubfoot

Clubfoot, also known as congenital talipes equinovarus (CTEV), is a congenital deformity of the foot, which may be idiopathic or associated with many different conditions, including neuromuscular diseases and syndromes. The deformity involves forefoot adductus (medial deviation), midfoot cavus (high arch), hindfoot varus (back of the heel is rolled inward), and equinus (back of the heel is up). It is relatively common with an incidence of 1 in 1,000 births.[60]

Patients present for comprehensive release of clubfoot only after nonoperative treatment has failed. Most commonly, Ponseti casting is successful in correction of all but the equinus release, which can be performed via percutaneous tenotomy under local or general anesthesia.

When surgery is required, it is to provide a plantigrade, functional, and painless foot. Surgical release includes a comprehensive and systematic release of all the tight and contracted structures encountered. The term *posterior medial release* (PMR) describes the approach and may include lengthening of the Achilles tendon, toe flexors, plantar fascia, and abductor hallucis. Additionally, the joint capsules of the tibiotalar, calcaneal cuboid, and talar navicular will require releases as well. The subtalar will need to be addressed but with great care as medial and lateral releases can cause translation of this joint with significant pain and deformity. Throughout the entire procedure, the neurovascular structures are identified and protected. Pins and casting are often required for 6 to 8 weeks to hold the foot in a good position until healing has occurred.

Anesthesia for clubfoot surgeries in children is generally a combination of general with regional. General anesthesia can be either with an endotracheal tube or with a laryngeal mask airway depending on the patient's comorbidities and the length of the anticipated surgical procedure, as well as the preference of the surgeon and anesthesiologist. Some surgeons prefer prone positioning for the operation, which may impact the method of airway maintenance. While the anticipated surgical pain may be only moderate, young children may have a significant negative reaction to the full leg cast.

There are several options for regional anesthesia, including caudal, epidural, and peripheral nerve blocks. Generally, a caudal is performed after induction for young children, and popliteal and saphenous blocks are done in older children. Many children will need to return to the operating room for cast change under anesthesia approximately 2 weeks after the primary repair.

Developmental Dysplasia of the Hip

Developmental dysplasia of the hip (DDH) describes a phenomenon of joint laxity and instability that may result from multiple causes. There is a wide range in severity from mild dysplasia to dislocation. Some deformities are associated with underlying syndromes and others are isolated and idiopathic. Typical DDH describes the isolated deformity. Teratological DDH describes genetically influenced hip dysplasia.[61]

The incidence of DDH is highly variable and ethnically diverse. Caucasian newborns have approximately 1% incidence of DDH and 0.1% incidence of hip dislocation. The incidence is very high in Manitoba, Canada with 188.5 cases per 1000 babies. DDH is more common with a positive family history, breech presentation, or female sex, and in conditions such as oligohydramnios, large birth weight, and first pregnancy that may lead to intrauterine crowding.[61]

All neonates are screened for DDH because it is asymptomatic at birth and the joint will not develop appropriately if the hip is dislocated. As children get older, they may present with limited hip abduction and apparent asymmetry of the thigh and hip. Children who can walk may present with an awkward gait, a limp, or apparent leg-length discrepancy. Patients may develop an excessive secondary lordosis. Hip dysplasia, or failure to develop acetabular coverage, may be identified in teens. This scenario is more likely to occur when the dysphasia is bilateral since the newborn exam depends somewhat on asymmetry for diagnosis.

The goal in treatment of DDH is to maximize normal development of the hip joint, which requires that the femoral head be securely nestled within the acetabulum. Most children who are diagnosed with DDH as infants will respond to nonoperative interventions. Children who are older at diagnosis may come to the operating room for closed reduction. Once the hip is reduced, a spica cast is placed to maintain the reduction. If closed reduction fails or if the child is older than 2 years of age at diagnosis, the patient is likely to require an open reduction of the hip. Generally, a femoral shortening osteotomy is done at the same time due to the risk of osteonecrosis due to pressure on the proximal femur.[61]

Surgical complications include avascular necrosis of the femoral epiphysis, redislocation, residual subluxation, acetabular dysplasia, and wound infections.[61] Rare complications include bleeding or sciatic or peroneal nerve palsies.

Anesthesia for hip reduction, both open and closed, is usually general with or without epidural placement for postoperative pain management. Postoperative pain is moderate, and patients may be more distressed by the spica cast than by the incisions and hip reduction.

Leg-Length Discrepancy

While a difference in leg length of 1 cm is relatively common, people with a discrepancy of 2 cm or more are considered to have a pathological leg-length discrepancy. There are many congenital or acquired causes of a leg-length discrepancy. Patients will present with gait asymmetry. They may also develop secondary lumbar curvature.[62]

Treatment for leg-length discrepancy is variable depending on the amount of discrepancy, as well as the patient's comorbidities and preferences. For discrepancies of up to 2.5 cm, observation and use of shoe lift may be adequate. Operative procedures may involve limb-shortening or limb-lengthening techniques.

Epiphysiodesis is performed for patients with a discrepancy between 2 and 5 cm who are skeletally immature. It is a temporary or permanent growth cessation at the physis of the long leg. In percutaneous epiphysiodesis, the physis is permanently ablated with a drill and curette.[62] Alternatively, plates and screws can be inserted for temporary cessation of growth. Typically, the hardware would be removed when the legs have equalized.

Acute leg shortening may be necessary if the patient is already skeletally mature. This is usually performed at the femur as the risk of neurovascular compromise is increased when performed at the level of the lower leg.[62] Lengthening of the short leg is indicated when the leg-length discrepancy is greater than 5 cm.

The Ilizarov method is a common technique used for limb lengthening via distraction osteogenesis. The procedure involves placement of an external fixator. The bone is cut at the metaphyseal–diaphyseal junction and lengthening occurs gradually.[62] The major principles involve a low-energy osteotomy to preserve blood flow to the periosteum, a slow, incremental distraction to preserve soft-tissue blood supply, and continued full function of the extremity.[10]

The Ilizarov method can also be used in the treatment of acquired short limbs and defects from trauma, tumor excisions, fractures, and infections.[10]

Surgical complications of leg-discrepancy procedures include pin-tract infections, wound infections, hypertension, joint subluxation, muscle contracture, premature consolidation, delayed union, implant-related problems, and fractures.[62]

Anesthesia for leg-length discrepancy surgery is guided foremost by the patient's comorbidities. Usually a combined approach is appropriate. After inducing general anesthesia, the preferred regional technique can be performed to provide postoperative pain control. Regional anesthesia should be discussed with the surgeon because a major surgical complication can be nerve damage and local-anesthetic-induced numbness might confuse postoperative evaluation. Additionally, patients are started in physical therapy as early as possible postoperatively and a continued dense block could interfere with safe mobility.

Postoperative pain can be moderate to severe, so regional anesthesia with intravenous narcotics and adjuvant analgesics such as acetaminophen and ketamine should be considered. Many surgeons will prefer to avoid nonsteroidal anti-inflammatory drugs (NSAIDs) due to the potential for impaired bone healing based on a retrospective review, which demonstrated a significant increase in the rate of nonunion after spinal fusion with the use of ketorolac.[63] An additional review article investigating the effect of NSAIDs on bone healing indicated that they do inhibit or delay fracture healing.[64]

Generally, there are no major hemodynamic shifts associated with leg-length discrepancy surgeries and invasive monitoring is not required. A single IV catheter that runs well is usually adequate.

Selected Upper Extremity Conditions

The upper limbs develop over weeks 4 to 8 of gestation. Limb anomalies are classified according to principles of limb development using a system called the OMT (Oberg, Manske, and Tonkin) scheme. Limb malformations are one of the most common types of anomalies.[65]

Amniotic Band Syndrome

Amniotic band syndrome (ABS) is a condition of circumferential bands around the limbs or digits resulting in significant loss of function and cosmesis. While the etiology of ABS may be either syndromic or not, treatment is similar for all patients. The hands are reconstructed by release of fused digits with skin grafting and release of bands where possible. These children present for surgery at a young age and may come back for recurrent surgeries as they grow, and scars need revision.

Anesthesia for patients with ABS is determined largely by any underlying conditions relating to the patient. The procedures are usually done under general anesthesia, but in many situations, upper extremity regional anesthesia can be very helpful for postoperative pain control, especially when limitation of narcotics is beneficial.

Syndactyly

Syndactyly is a webbing of the digits that results from a failure of apoptosis during limb development. Complete syndactyly involves the entire length of the digit, whereas incomplete syndactyly does not cover the length of the entire digit. Complex versus cutaneous syndactyly describes bony involvement. There may also be anomalies of the nerves, tendons, and muscles. Anesthesia considerations are like those listed for ABS earlier in this chapter.

Spine Deformity

Scoliosis

Scoliosis is an abnormal curvature of the spine in the coronal plane. The term *scoliosis* refers to a lateral curvature of the spine and derives from the Greek word "skolios," which means bent or curved. Scoliosis is a three-dimensional deformity with both curvature and rotation. The term *kyphosis* refers to anterior flexion of the spine, which can be a component of spinal deformity requiring surgery as well. Most scoliosis cases are idiopathic, but a significant number of children have neuromuscular, congenital, or syndromic scoliosis.

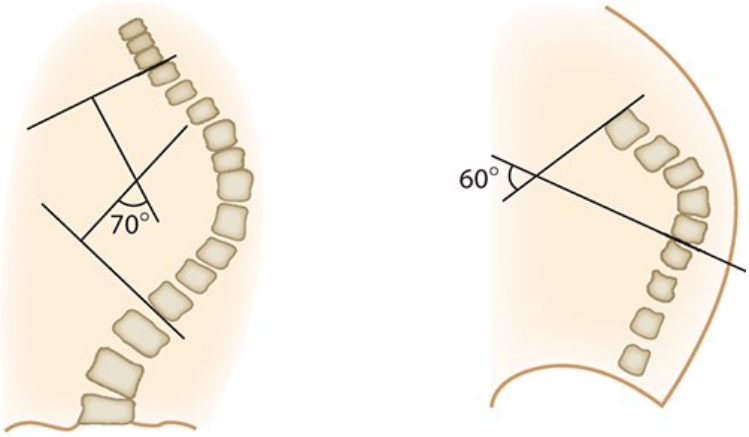
Idiopathic scoliosis may be infantile, juvenile, or adolescent with overall prevalence from 1% to 3% of skeletally immature patients.[66]

Scoliosis[66]	Age	Incidence (% of idiopathic cases)
Infantile	Birth to 3 years	0.5–4
Juvenile	3–10 years of age	8–16
Adolescent	≥11 years of age	70–80

Normal sagittal alignment includes cervical lordosis (concave curve), thoracic kyphosis (convex curve), and lumbar lordosis. Idiopathic scoliosis is a pathological exaggeration of the normal curvature of the spine, with additional rotation, and is a diagnosis of exclusion. Underlying intraspinal abnormalities such as tethered cord and syringomyelia must be ruled out. Evaluation of scoliosis begins clinically and is aided by radiographs. The curvature is defined by the Cobb angle (Figure 22-1). An angle of greater than 10 degrees is termed scoliosis.

Figure 22-1

Cobb angle. (Reproduced with permission, from Freeman BS, Berger JS: *Anesthesiology Core Review: Part 2, Advanced Exam*. 2016. https://accessanesthesiology.mhmedical.com. Copyright © McGraw Hill LLC. All rights reserved.)



Source: Herodotos Ellinas, Kai Matthes, Walid Alrayashi, Aykut Bilge: *Clinical Pediatric Anesthesiology*

Many patients with infantile idiopathic scoliosis will have spontaneous resolution of their spinal curvature. Those with developmental delay, presentation after 1 year of age, and larger curves are less likely to have this resolution. Far fewer patients with juvenile or AIS will have spontaneous resolution.

Congenital scoliosis refers to abnormality of the spine that is present from birth and usually results from abnormal development of the spine or ribs in utero. The clinical abnormality may not be immediately apparent at birth. Since the deformity results from an intrauterine insult, it is not surprising that many children will have other congenital abnormalities: 10% to 40% will have genitourinary abnormalities including unilateral renal agenesis, ureteral duplication, horseshoe kidney, and genital anomalies, 10% to 25% cardiac abnormalities, and 15% to 40% intraspinal anomalies such as tethered cord, teratomas, and closed spinal dysraphisms.[66]

Neuromuscular scoliosis is seen in children with underlying neuromuscular diseases and ones with spinal cord trauma. The scoliotic curve for these children results from muscular imbalance and sometimes spasticity. Neuromuscular scoliosis is positively associated with higher degrees of neurological impairment. It is present in more than 70% of patients with cerebral palsy and more than 90% of patients with Duchenne muscular dystrophy.[66] Virtually all patients who suffer a spinal cord injury prior to 10 years of age will develop scoliosis.

Many syndromes are strongly associated with scoliosis including Ehlers-Danlos, Marfan, Prader-Willi, neurofibromatosis, osteogenesis imperfecta, mucopolysaccharidosis, and rheumatoid arthritis.[10,66]

Specific treatment of scoliosis is based on the type of scoliosis, the rate of curve progression, the age of the patient, and the patient's goals. Otherwise healthy patients with scoliosis generally have a goal of maintaining the highest degree of function, with good cosmetic outcomes and as little risk as possible. Severely debilitated children may have a completely different end point such as prevention of curve progression and maintenance of alignment that best supports care of the child (ability to sit comfortably).

Preoperative evaluation for scoliosis surgery should be thorough given the potential for comorbidities and the fact that the operation is elective. In addition to a complete history and physical to elicit abnormalities and changes from baseline, radiographs, PFTs, electrocardiograms, echocardiograms, and coagulation studies are often helpful.

Chest radiographs are part of the surgical planning but are also relevant to the anesthesiologist. Thoracic curves can impact lung volumes and pulmonary compliance, even with a relatively small degree of curvature. Patients who have a curve of 65 degrees or more are likely to have restrictive lung disease based on PFTs and are at higher risk of postoperative respiratory complications. They may also have abnormalities in their central respiratory drive and upper airway function resulting in impaired clearance of secretions and recurrent infections putting them at risk of perioperative aspiration and need for postoperative mechanical ventilation.

Preoperative echocardiogram may be indicated even for patients with AIS. There is an increased incidence of mitral valve prolapse in patients with scoliosis and the study can provide valuable information on the degree of valve insufficiency. Patients with as little as 25 degrees of curvature can have increased pulmonary artery pressures, especially with exercise. Most patients will have pulmonary artery hypertension with exercise at a 70-degree curve and even at rest with a 110-degree curve.[3] Patients with pulmonary hypertension are at increased risk of perioperative right-sided heart failure and sudden death due to increased right ventricular (RV) afterload, hypoxemia, hypotension, and inadequate RV preload.[67] Tight hemodynamic parameters must be maintained for patients with pulmonary artery hypertension undergoing scoliosis surgery. Any child with a myopathy should have a preoperative electrocardiogram and echocardiogram due to the potential for associated cardiomyopathy.

Surgical goals in scoliosis surgery include stabilization of the spine and prevention of continued curve progression as well as improved cosmetic appearance and comfort. The approach may be anterior, posterior, or both. Anterior spinal surgery may be intended for complete treatment or for release of the spine to enhance the correction done posteriorly. Anterior spinal surgery is indicated for severe kyphosis. Thoracic curves are approached via thoracotomy or video-assisted thoracotomy on the convex side of the curve. Discectomies are performed in order to release the tension on the curvature. Thoracolumbar curves may be approached with a high subcostal incision and lumbar curves can be approached transabdominally or extraperitoneally.

Posterior spinal fusion is performed via bone grafting. The operation generally extends from one vertebra above the curve to the second vertebra below the curve. Raw bone is exposed via removal of the spinous processes. The bone graft is packed over the decorticated surface on the concave side. Instrumentation is performed to hold the spine stable during the healing of the bone resulting in spinal fusion.

Patients with neuromuscular scoliosis may require placement of growing rods at a young age. The hardware is placed and extended as the child grows. Newer rods have a magnetic extension system, which allows elongation of the rods without an operation. Anesthetic considerations for growing rod placement and removal are similar to those of posterior spinal fusion, although since the surgery is less involved, often an arterial line is not necessary, and the surgery may even be done as an outpatient procedure.

Spinal fusion for scoliosis is a major operation and requires general endotracheal anesthesia. The anesthesia team must be prepared for large volume blood loss and associated hemodynamic and electrolyte changes, complications of prone positioning, as well as rare complications such as pneumothorax, anaphylaxis, venous air embolism, and loss of motor or sensory evoked potentials.

Many patients presenting for scoliosis surgery will benefit from preoperative anxiolysis with midazolam intravenously or orally. Induction can be done via inhalation or intravenous routes. Standard of care for scoliosis surgery includes neuromonitoring and therefore the anesthetic plan will optimize this to the extent possible. An arterial line in addition to standard ASA monitors is indicated for hemodynamic monitoring and frequent blood sampling. A central line may be necessary if there is inadequate peripheral access for the anticipated blood loss or if medications or CVP monitoring are indicated by the patient's comorbidities.

Once the patient is asleep, the neuromonitoring technician will place electrodes for SSEP and MEP monitoring. This can occur simultaneously with placement of additional lines and the foley catheter. The whole team should be present for prone positioning. The surgeon can direct positioning for the operation and the anesthesia team can ensure head and neck remain neutral and supported. Pressure points are particularly vulnerable during this operation because of duration and possible hypotension.

A small dose of rocuronium or vecuronium given with induction drugs is generally metabolized by the time the patient is ready for MEP monitoring. It is useful to look at the anesthetic depth at the time of MEP baselines because if there are MEP changes associated with the surgery, one can determine the relationship to anesthetic changes from baseline. Many institutions will run 0.5 MAC of isoflurane in conjunction with a propofol drip and a narcotic infusion to maintain an adequate anesthetic/analgesic depth.

Blood loss can be quite significant during spine surgery, especially for patients with neuromuscular scoliosis who may have a higher tendency toward bleeding, fragile tissues, and a smaller circulating blood volume relative to healthy patients presenting with AIS. Cell saver is utilized and can decrease the amount of allogeneic blood transfusion required. Transfusion triggers should be discussed as a team. They are variable and should account for anticipated ongoing blood loss. Exposed bony surfaces are a source of blood loss throughout the operation.

At the conclusion of a spine surgery, patients are usually emerged from anesthesia and extubated awake. The surgeon will often prefer a motor exam in the operating room when the patient has the mental capacity to follow commands. In cases where the patient is not suitable for extubation postoperatively, she or he can be transferred to a pediatric intensive care unit (PICU) for further management. This may be indicated for patients with severely compromised pulmonary function or poor baseline functioning as well as those who may have had major blood loss and resuscitation.

Scoliosis surgery is painful and there are a number of ways to manage the pain. For patients who can do so, a PCA is appropriate. Adjunctive pain medicines including acetaminophen and ketamine may be helpful. Many surgeons prefer to avoid ketorolac or ibuprofen due to the potential for inhibition of bone healing.

In addition to blood loss and positioning injuries, complications of scoliosis surgery can include coagulopathy, postoperative visual loss (POVL), infection, pneumothorax, neurological injury including paralysis, and cardiovascular collapse as a result of hemorrhage, anaphylaxis, or air embolism.

When there is an emergency during spine surgery, the surgeon focuses on controlling bleeding and covering the incision to maintain sterility in preparation for a quick “flip” to the supine position. A stretcher should always be available either inside or just outside the operating room for this purpose. The anesthesia team focuses on resuscitation and the differential diagnosis. Once the patient has been stabilized a decision needs to be collaboratively made whether to abort the surgery. Factors that must be considered include ongoing hemodynamic changes, bleeding and risk of bleeding, spine instability, and neurological injury as indicated by neurophysiological monitoring.

Acute Presentations

Fractures

Pediatric fractures differ from adult fractures in several ways. Pediatric bones have periosseous cartilage, physes, and a thicker, stronger, more osteogenic periosteum, which produces new bone more rapidly and in greater amounts. They are also lower in density and are more porous. These differences result in lower elasticity and bending strength.[68] Children are prone to remodeling and potential overgrowth at the site of the fracture. Injuries to the physes can cause progressive deformities and this type of fracture should be observed over the long term.

Fractures can alert the health care team to the potential for child abuse because they are the second most common manifestation of abuse, after skin changes such as bruising, burns, and abrasions.[68] Fractures that should raise suspicion of abuse include femur fractures in nonambulatory children, distal femoral metaphyseal corner fractures, posterior rib fractures, scapular spinous process fractures, and proximal humeral fractures.[68] These fractures are not definite indicators of abuse and there are situations in which underlying disease can mimic the fracture pattern of abuse such as osteogenesis imperfecta and osteomyelitis. Suspected nonaccidental trauma needs to be reported and managed by a specially trained team. However, the operating room may be an opportune time to examine the child and document injuries.

Fractures around the elbow in children require aggressive management to protect against malunion or nonunion.[68] The fractures may be transcondylar, supracondylar, and epiphyseal. Displaced supracondylar fractures may have associated neurovascular injury that may warrant immediate intervention. Operative treatment for distal humeral fractures is closed reduction and pinning when possible. Inadequate reductions can

result in limitation of motion, cubitus varus or valgus, and nonunion or instability.[68]

Hip fractures in children are relatively uncommon but may present with surgical urgency. They result from high-impact trauma and are likely to have associated injuries. The rate of avascular necrosis is as high as 50% without urgent reduction, stable fixation, and spica casting.[68] In the urgent setting, precautions against aspiration of gastric contents can be taken with a rapid sequence intubation. Regional anesthesia via a subarachnoid block or continuous epidural is less practical in this situation given the potential concurrent injuries and the risk of a full stomach or ileus.

Surgery for repair of fractures is indicated for displaced physeal fractures, displaced intra-articular fractures, unstable fractures, multiple injuries, open fractures, failure to achieve or maintain adequate reduction, and pathological fractures.[68] Surgical goals include restoration of alignment and stability. External fixation may be indicated for open fractures or fractures associated with other injuries as well as fractures associated with vascular or nerve injury.

Complications from fractures include growth arrest, or premature physeal closure, misalignment avascular necrosis, and compartment syndrome. Other risks include infection and neurovascular compromise postoperatively. There can be limb deformities from partial or complete closure of the physis. Complex regional pain syndrome is a rare but potentially debilitating complication.[68]

PERIOPERATIVE COMPLICATIONS

Venous Thromboembolism

Venous thromboembolism (VTE), including the clinical conditions of deep vein thrombosis (DVT) and pulmonary embolism (PE), is increasingly relevant to pediatric patients. The increased incidence is thought to be due to better surveillance and diagnosis and longer survival of children with chronic diseases.

The incidence of VTE in children increased by 70% from 2001 to 2007 (from 34 to 58 cases per 10,000 hospital admissions in tertiary care facilities).[69] Of patients admitted with a diagnosis of VTE in the same cohort, the incidence of PE was 11% and mortality was 8%.[69] The mortality of children who suffer from PE is 2.2%.[70,71] Infants younger than 1 year of age and adolescents are more likely than other children to develop VTE.

A high index of suspicion for VTE as well as a clear understanding of risk factors are essential in diagnosing and preventing the potentially catastrophic complication of massive PE. The single most important risk factor for VTE in the pediatric population is the presence of a central venous catheter (CVC).[70] More than 50% of DVTs in children and 80% of DVTs in newborns occur in patients with indwelling central lines.[71] Infection, including osteomyelitis, septic arthritis, septicemia, and local infection, was the second most common risk factor for VTE.[70] Other common risk factors for pediatric VTE include surgery, malignancy, trauma, heart disease, and nephrotic syndrome.

A venous thrombus can form when there is hemostasis and hypercoagulability or damage to the vessel endothelium which occur during trauma and surgery. Pulmonary embolism is the result of a dislodged thrombus from a deep vein. The thrombus can travel through the venous system until it reaches the right atrium and from there occlude the pulmonary arteries. An otherwise healthy child may remain asymptomatic with up to 50% of the pulmonary circulation occluded. With a massive PE, cardiopulmonary collapse can result from an acute increase in RV afterload, resulting in RV dilation and increased RV and PA pressures. Ultimately, cardiac output can be compromised, and death can result.

Awake adolescent patients are most likely to complain of pleuritic chest pain with a new onset PE. Younger patients may present with unexplained tachypnea. Intraoperative manifestations of a PE may include the sudden onset of hypoxemia, hypertension, and loss of end-tidal carbon dioxide (which indicates a loss of cardiac output). The diagnosis can be supported by intraoperative transesophageal echocardiogram (TEE) when available. When a PE is suspected intraoperatively, the surgery should be completed or aborted as quickly as possible. The diagnosis can be confirmed by CT or MRI once the patient is stabilized. Once diagnosis is made, initial therapy is likely a heparin infusion or low-molecular-weight heparin injection. A hematologist should be consulted for evaluation of underlying procoagulant states. A postsurgical patient may be best treated with unfractionated heparin due to the shorter half-life of the medication in the event of postoperative bleeding. Children who have less of a bleeding risk may benefit from low-molecular-weight heparin since it is dosed subcutaneously and does not need to have drug levels closely monitored.

Other complications include major hemorrhage as well as recurrent thrombus formation. Children are at risk of developing long-term disability from postthrombotic syndrome, which is a result of damage to the venous valves causing swelling and pain due to venous insufficiency. Other clinical

manifestations of VTE may include cerebral sinovenous thrombosis, renal vein thrombosis, peripheral arterial thrombosis, and stroke as well as thrombotic storm, which is a rapidly progressive multifocal thrombosis that can result in multiorgan system dysfunction.[72]

Fat Embolism Syndrome

While fat embolism is nearly universal in patients with long bone fractures, fat embolism syndrome (FES) occurs with far less frequency. Fat emboli occur in as many as 90% of patients with long bone fractures, but symptomatic FES occurs in only 10% to 20% of patients with long bone or pelvic fractures. Symptomatic FES increases to 30% with multiple fractures.[10] The syndrome is characterized by a triad of hypoxemia, neurological changes, and a petechial rash. The onset of symptoms is approximately 48 to 72 hours after the injury and usually the first symptom is respiratory insufficiency. Most patients don't develop the complete triad of symptoms.

There are two theories of FES pathophysiology: mechanical and biochemical. The mechanical theory describes the direct damage caused by fat globules that are released into the pulmonary, neurological, and cutaneous systems. Essentially, fat globules are released at the site of injury and enter blood vessels. From the venous system, they can pass into the arterial system via the lungs or a patent foramen ovale (PFO). The biochemical theory describes the localized reaction of free fatty acids in the pulmonary microvasculature resulting in hemorrhage and inflammation. The resulting clinical syndrome is much like acute respiratory distress syndrome.[10]

Under anesthesia, the most likely presentation of FES is hypoxia without another explanation. The patient may exhibit a drop in the arterial oxygen concentration, hematocrit, platelet, and fibrinogen levels. Fulminant FES is most likely to occur 30 minutes after transfer to the operating room table. Clinically, the patient will exhibit progressive oxygen desaturation, hypotension, tachycardia, bradycardia, dysrhythmia, decreased lung compliance, pulmonary edema, and disseminated intravascular coagulation.[10] Treatment of FES is supportive. Patients may benefit from bronchoalveolar lavage to remove particles of fat and hemorrhage. This is also the most rapid and specific method of FES diagnosis. The mortality rate of FES in children is 33%.[10]

CONCLUSION

The variety of pediatric patients who present for orthopedic surgery is immense. Common themes which can improve care for patients intraoperatively include knowledge of the underlying disease process or pathophysiology, extreme care with positioning, attention to potential blood loss, and teamwork. Anesthesiologists can positively impact long-term outcomes by fostering a connection with each individual patient and family and leading the operative team through any crisis that may arise.

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