

# Other Surgeries

SECTION

# VII

## Orthopedic and Spine Surgery

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CHAPTER

# 30

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**ANESTHESIA FOR ORTHOPEDIC AND** spinal surgery provides a multitude of challenges. Children often present with concomitant diseases that affect cardiovascular and respiratory function. The ability to maintain a clear airway during anesthesia is not straightforward in some of these children (e.g., those with arthrogryposis multiplex congenita).<sup>1</sup> Operating times can be protracted. Significant blood loss can occur and requires strate-

gies for blood product management and transfusion reduction (see Chapter 10). Major trauma causing orthopedic injuries invariably involves other organ systems that may adversely interact with or compromise anesthetic management (see Chapter 39). The risks of aspiration of gastric contents into the lungs and the requisite fasting times, after even minor trauma involving an isolated forearm fracture, continue to be debated.

Fat embolus is uncommon in children with fractures of the long bones but should be sought in any child with hypoxia and altered consciousness.<sup>2</sup> Tumor surgery may be complicated by chemotherapy, altered drug disposition, or bone grafting considerations akin to plastic and reconstructive surgery (see Chapter 33).

Children with chronic illnesses present repeatedly for surgical or diagnostic procedures. A single bad experience can blight attitudes to anesthesia for a considerable time. These children should be managed with sensitivity and compassion. Positioning children on the operating table involves care, especially in those with limb deformities and contractures. Padding, pillows, and special frames are required to protect against damage from inadvertent pressure ischemia while achieving the best posture for surgery. Plaster application, particularly around the hip, should allow for bowel and bladder function, avoid skin breakdown due to pressure or friction, and allow access to epidural catheters. The postoperative management of casts on peripheral limbs must account for the possibility of compartment syndromes attributable to restrictive casts or compartment pathology. Regional techniques should not mask pressure effects under plaster casts or compartment syndrome, although epidural blocks may be ineffective against the discomfort of pressure.<sup>3,4</sup> Intraoperative temperature regulation (see Chapter 25) may be affected by tourniquet application owing to a combination of decreased heat loss from the ischemic limb and reduced heat transfer from the central to ischemic peripheral compartment. Some disease processes are associated with altered temperature regulation (e.g., osteogenesis imperfecta, arthrogryposis multiplex congenita). The use of radiology is common during orthopedic surgery, and precautions against radiation exposure during bony manipulations should not be neglected by the anesthesiologist.

Regional anesthesia (see Chapter 42) reduces anesthesia requirements intraoperatively and provides analgesia postoperatively. The use of ultrasound techniques to locate neural tissue improves success and reduces local anesthetic doses (see Chapters 42 and 43).<sup>5,6</sup> The recent introduction of ultrasound techniques has heralded a rapidly increasing use of peripheral nerve blockade rather than central blockade for unilateral lower limb surgery. Acetaminophen (paracetamol) and NSAIDs are the most common analgesics prescribed to children for moderate pain. The regular administration of acetaminophen and NSAIDs decreases the amount of systemic opioids administered,<sup>7</sup> but NSAIDs decrease osteogenic activity and may increase the incidence of nonunion after spinal fusion.<sup>8,9</sup> Intravenous administration of acetaminophen improves the early effectiveness of this drug before the child is able to tolerate oral intake, but this formulation is not available in all countries.<sup>10</sup> Long-term pain associated with limb-lengthening techniques (e.g., Ilizarov frame) may require oral opioids after hospital discharge.

## Scoliosis Surgery

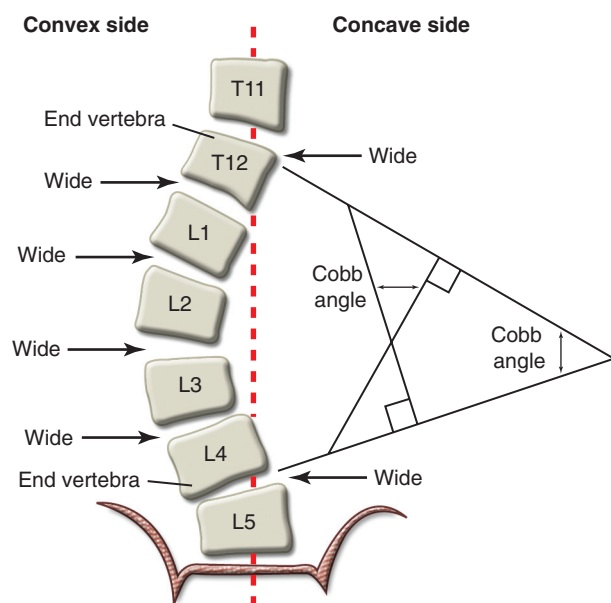
Children presenting for scoliosis surgery represent a spectrum from the uncomplicated adolescent to severely compromised children with neuromuscular disease, respiratory failure, and cardiac problems. The age range at presentation varies from infancy to young adulthood, and anesthetic approaches need to be tailored for each individual child.

## Terminology, History, and Surgical Development

Hindu literature (3500-1800 BC) describes Lord Krishna curing a woman whose back was “deformed in three places.”<sup>11</sup> The words “scoliosis” (crooked), “kyphosis” (humpbacked), and “lordosis” (bent backward) originated with the Greek physician Galen. *Scoliosis* is a lateral deviation of the normal vertical line of the spine, which when measured by x-ray, is greater than 10 degrees. There is a lateral curvature of the spine with rotation of the vertebrae within the curve. *Lordosis* refers to an anterior angulation of the spine in the sagittal plane, and *kyphosis* refers to a posterior angulation of the spine as evaluated on a side view of the spine. Curves may be simple or complex, flexible or rigid, and structural or nonstructural. Primary curves are the earliest to appear and occur most frequently in the thoracic and lumbar regions. Secondary (or compensatory) curves can develop above or below the primary curve and evolve to maintain normal body alignment. The varying combinations of curve types result in different pathophysiologic consequences.

The magnitude of the scoliosis curve is most commonly measured using the Cobb method.<sup>12</sup> Measurement is made from an anteroposterior radiograph and requires accurate identification of the upper and lower end vertebrae involved with the curve. These are the vertebrae that tilt most severely toward the concavity of the curve. The Cobb method of angle measurement is shown in Figure 30-1.

Hippocrates (circa 400 BC) developed treatments that relied primarily on manipulation and traction, using an elaborate traction table called a scamnum.<sup>13</sup> Nonsurgical treatments for spinal deformities persisted until 1839 when a surgical treatment in the form of a subcutaneous tenotomy and myotomy was described by the French surgeon Jules Guerin.<sup>14</sup> Posterior spinal fusion appears to have been first described by Russell Hibbs for tuberculous spinal deformity in 1911.<sup>15</sup> The original spinal instrumentation system was the Harrington rod system.<sup>16</sup> Modification of this technique allowing segmental fixation of the rods, and early mobilization followed.<sup>17</sup> These systems treated



**Figure 30-1.** Diagram of anteroposterior spinal radiograph showing Cobb method of scoliosis curve measurement.

the lateral curve but did not allow for correction of the axial rotation. Subsequent developments allowed both via cantilever maneuvers using Cotrel-Dubousset instrumentation.<sup>18</sup> Pedicle screws rather than hooks were the next advance. These were initially used with lumbar curves as a distal anchor and were found to enhance correction and stabilization, even when used with hooks for the more proximal curves (hybrid constructs).<sup>19</sup> Pedicle screw instrumentation techniques for total curve correction have been a recent development and have been shown to offer better curve correction than hook techniques<sup>20</sup> and the hybrid pedicle screw/hook technique.<sup>21</sup>

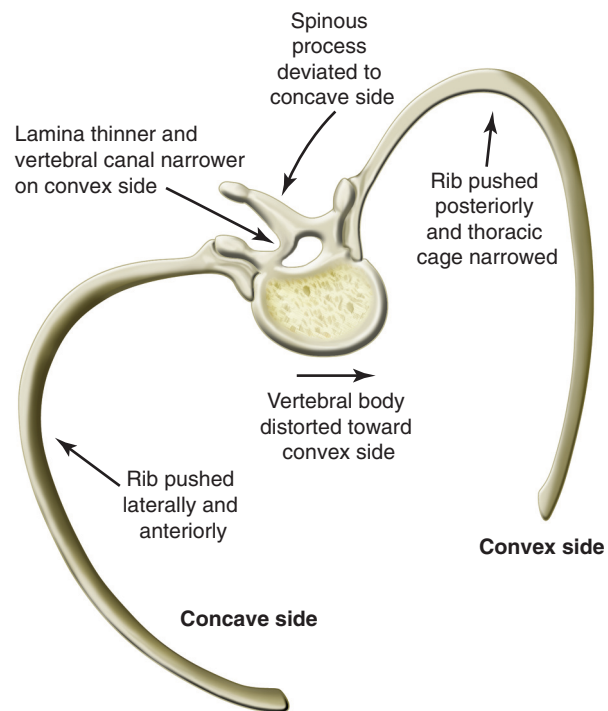
### Classification

Classification of scoliosis deformities is imperfect because the systems used are clinically rather than etiologically based. Most classifications are surgically based and relate to surgical decision-making. Curves can be described on the basis of age at onset, associated pathology, and anatomic configurations of the curve (e.g., single, double, or triple curves; amount of pelvic tilt; curve flexibility, as well as systems based on three-dimensional analysis).<sup>22</sup> From an anesthetic perspective, a classification that gives some idea of the risk of adverse outcome, in particular respiratory failure, would be of clinical benefit. Children with scoliosis of early onset (<5 years) or with independent cardiac or pulmonary disease appear to be at increased risk of respiratory failure, whereas children with idiopathic scoliosis in whom the curve develops at adolescence appear to have minimal risk.<sup>23</sup> A classification adapted from that proposed by the Scoliosis Research Society in 1973 remains relevant to anesthesiologists (Table 30-1).<sup>24</sup>

### Pathophysiology and Natural History

Vertebral rotation and rib cage deformity usually accompany any lateral curvature. With progression of the curve, the vertebral bodies in the area of the primary curve rotate toward the convex aspect of the curve and the spinous process rotates to the concave side. This vertebral rotation can be determined by measurement of the position of the pedicles from the midline (Moes method).<sup>25</sup> The vertebral bodies and the discs develop a wedge-shaped appearance with the apex of the wedge toward the concave side. On the convex side of the curve the ribs are pushed posteriorly, which narrows the thoracic cavity and causes the characteristic hump. On the concave side the same rotation forces the ribs laterally, with consequent crowding toward their lateral margins (Fig. 30-2). These changes result in an increasing restrictive lung defect. Exactly when this becomes a problem depends on the child's accompanying pathology. The thoracic and lumbar regions are the most common sites of the primary curve. In children in whom the primary curve is in the lumbar region, the rotation of the vertebral bodies and spinous processes should be taken into consideration when a spinal or epidural drug is to be administered.

The physical distortion in the thorax results in restriction of lung volumes and function. Ventilation depends on the mobility of the thoracic cage, the volume of each hemithorax, and the muscle power and elastic forces required to move the thorax. Children with idiopathic scoliosis with a mild decrease in vital capacity (VC) also have reduced forced expired volume at 1 second (FEV<sub>1</sub>), gas transfer factor, and maximum static expiratory airway pressures (PE<sub>max</sub>) (see Chapter 11). The predominant deformity of lateral flexion and vertebral rotation results in



**Figure 30-2.** Characteristic distortion of vertebrae and ribs in thoracic scoliosis. (Modified from Kleim HA: *Scoliosis*. Ciba Foundation Symposium, Summit, NJ, Ciba, 1978, vol 1, p 609 and Gregory GA [ed]: *Anesthesia for orthopaedic surgery*. In *Paediatric Anesthesia*, 3rd ed. Edinburgh, Churchill Livingstone, 1994.)

the lung on the concave side being able to achieve a near-normal end-expiratory position but not end-inspiratory position, whereas the lung on the convex side achieves a normal end-inspiratory position but cannot reach a normal end-expiratory position. The concave side will contribute less than normal at total lung capacity (TLC), resulting in a decrease in PE<sub>max</sub>. Similarly, because the convex side does not reach a normal end-expiratory position, the intercostal muscles and hemidiaphragm will be less efficient, resulting in a reduced maximum static inspiratory airway pressure (PI<sub>max</sub>), although this reduction may not be quite so marked.<sup>26</sup> The main effect of scoliosis on respiratory function is thought to be mechanical, with the anatomic changes in the chest wall causing impaired movement and reduced compliance. Potential long-term respiratory problems when these defects are left untreated include hypoxemia, hypercarbia, recurrent lung infections, and pulmonary hypertension.

### Idiopathic Scoliosis

Although adolescent idiopathic scoliosis is relatively common, severe morbidity, and even mortality is only seen in children with early onset (infantile or juvenile) idiopathic scoliosis.<sup>27</sup> Respiratory deterioration alone is seldom the reason for surgery in children who develop scoliosis after the age of 5 years.<sup>23</sup> This is probably because most of the multiplication, division, and development of alveoli in the lungs have occurred by this age.<sup>28,29</sup>

The scoliosis evolves during periods of development and physical growth, and curve progression occurs during growth spurts. This is not the case in children with neuromuscular

**Table 30-1.** Classification of Scoliosis with Associated Key Anesthetic Risk Factors

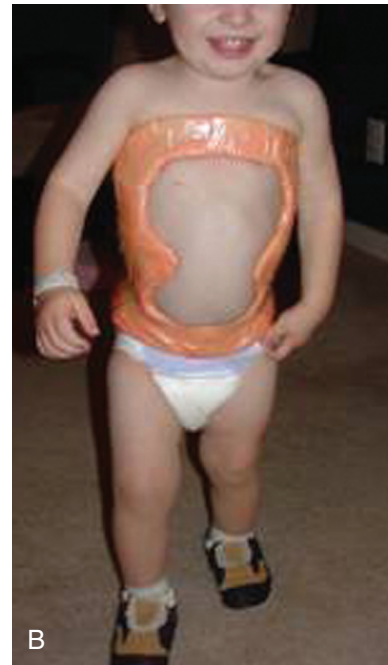
Classification	Associated Issues with Scoliosis Surgery	↑ K <sup>+</sup> with Succinylcholine	Expected High Blood Loss	Respiratory Complication/Ventilatory Support
<b>Idiopathic</b>				
Infantile <3 years	Repeat operations, small size		√	√
Juvenile 3-9 years				
Adolescent 9-18 years	Regarded as “cosmetic” by patient—perfect result expected			
<b>“Congenital”</b>				
Bony abnormalities	“Acute angle” deformity: high risk of spinal cord injury, genitourinary malformations			
Neural tube defects				
Meningocele, spina bifida, syringomyelia	Latex allergy, pressure sores, hydrocephalus, Arnold-Chiari malformation (avoid neck extension)			
<b>Neuromuscular</b>				
<b>Neuropathic</b>				
Upper motor neuron Cerebral palsy, cerebral hypoxia	Upper airway obstruction, recurrent pneumonia, postoperative pain management		√√	√√
Lower motor neuron Poliomyelitis				
<b>Myopathic</b>				
Progressive				
Duchenne muscular dystrophy	Cardiomyopathy, mitral valve prolapse, conduction abnormalities	√	√√	√√
Spinal muscular atrophy	Electrocardiographic abnormalities	√	√	√
Facioscapulohumeral muscular dystrophy	Hypertrophic cardiomyopathy, cardiac failure	√		
Other				
Friedreich ataxia		√		
<b>Neurofibromatosis</b>	Hypertension, other neurofibromas			
<b>Mesenchymal</b>				
Marfan syndrome	Mitral/aortic regurgitation			
Mucopolysaccharidoses (Morquio syndrome)	Atlantoaxial subluxation, difficult intubation			
Arthrogyrosis	Difficult intubation, severe contractures		√	
Osteogenesis imperfecta	Small size			
<b>Trauma</b>				
<b>Tumor</b>				

Adapted from Goldstein LA, Waugh TR: Classification and terminology of scoliosis. Clin Orthop Relat Res 1973; (93):10-22.

scoliosis, in whom progressive and relentless deterioration in respiratory function can occur throughout life. The earlier the age at onset and the more immature the bone growth at the time the process begins, the more adverse the outcome. The relentless progression of children with infantile-onset idiopathic scoliosis with rapidly deteriorating curves and lung function appears not to be improved by surgery. Treatment involving spinal instrumentation and anterior epiphysiodesis does not prevent the reappearance of the deformity or the decrease in pulmonary function.<sup>30</sup>

Nonoperative approaches to correcting early-onset scoliosis are being described, with infants and toddlers undergoing sequential body casts molded to correct the spinal deformity. Treatment may begin as early as 4 to 5 months of age or as soon as the diagnosis of scoliosis has been made. Improvement and, in some cases, resolution, has been achieved at 9-year follow-up.<sup>31</sup> After induction of anesthesia, the child is positioned on the frame (first described by Cottrell and Morel) with the pelvis secured to the caudal end of the frame and the head tethered via a chin strap to the rostral end. The spine is mildly distracted, but the main maneuver is to derotate the spine through the ribs (Fig. 30-3A). General anesthesia with tracheal intubation is required to facilitate positioning the child, stretching the spine and molding the body cast. Without intubation, hemoglobin desaturation frequently occurs when molding the cast to correct the spine deformity. An oral airway is also needed to prevent external compression of the airway after the chin strap is applied and tightened. Once the cast has hardened, it is cut back and trimmed to maintain the correction to the spine while facilitating breathing, gastrointestinal function, and activities of daily living (see Fig. 30-3B).

There is a direct correlation between pulmonary impairment and the magnitude of the thoracic curve. The severity of the scoliosis is the most accurate predictor of impaired lung function.<sup>32</sup> The morphology of the thoracic curve, the number of vertebrae in the major curve, and the rigidity of the curve are also factors associated with deteriorating pulmonary function.<sup>33</sup> The conventional wisdom has been that there is minimal impact on the vital capacity (VC) until the curve exceeds 60 degrees, with clinically relevant decreases in respiratory function occurring only after the thoracic scoliosis has progressed beyond 100 degrees.<sup>34</sup> More recently it has been shown that children with adolescent idiopathic scoliosis may have pulmonary impairment disproportionate to the severity of the scoliosis and that significant respiratory impairment can occur well before the curve reaches 100 degrees. One review suggests that FVC falls below the normal threshold (80%) once the magnitude of the thoracic curve exceeds 70 degrees; FEV<sub>1</sub> falls below the normal threshold once the main thoracic curve exceeds 60 degrees.<sup>35</sup> Twenty percent of children with a thoracic curve of 50 to 70 degrees have moderate or severe pulmonary impairment (<65% of predicted) (Fig. 30-4A).<sup>35</sup> Children with thoracic hypokyphosis are more likely to have moderate or severe pulmonary impairment; complex curves have a higher prevalence of moderate or severe pulmonary impairment, and the number of vertebrae in the thoracic curve is the most significant predictor of impaired respiratory function (see Fig. 30-4B).<sup>35</sup> These data indicate that children with a structural cephalad thoracic curve, a major thoracic curve spanning eight or more vertebral levels, or thoracic hypokyphosis are at increased risk for moderate to severe pulmonary impairment.

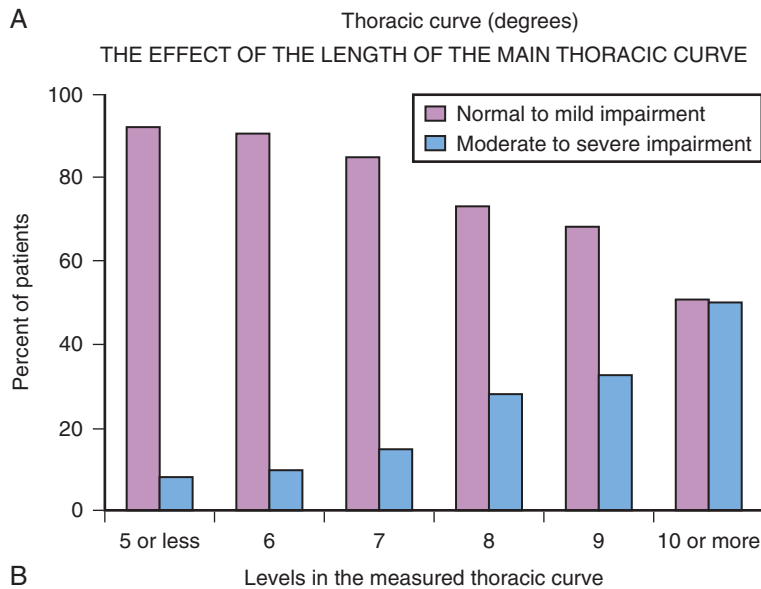
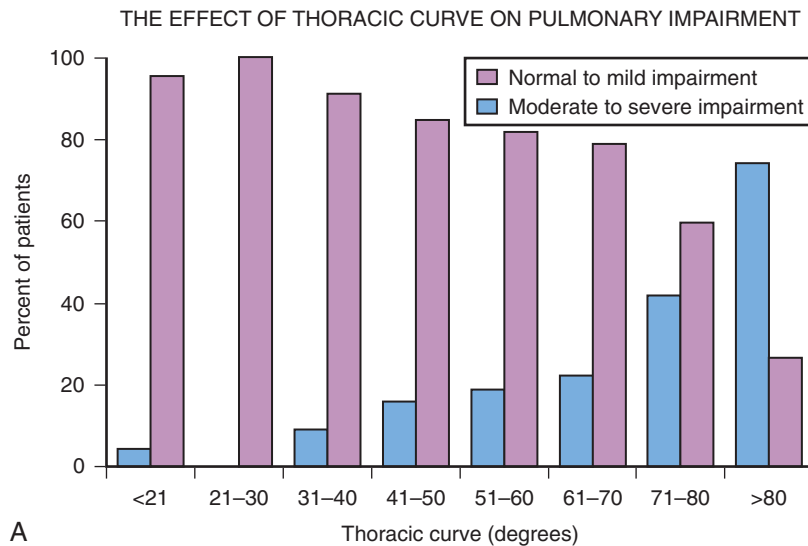


**Figure 30-3.** **A**, Nonoperative correction of scoliosis in infants and toddlers may be achieved with repeated casting. **B**, Cutting and trimming of the cast allows correction of the spine while facilitating day-to-day living.

### Neuromuscular Scoliosis

Children with neuromuscular scoliosis have the burden of deteriorating muscle function in addition to mechanical distortion. Crowding of the ribs on the concave side of the curve limits chest wall expansion and the sitting posture restricts diaphragmatic excursion. This inevitably leads to more rapid deterioration in both the curve and respiratory function. This group of children also has potential for rapid and unpredictable deterioration of the curve.<sup>36</sup> It is important to consider the natural history of the specific neuromuscular disease when trying to balance the risks of surgery against conservative management.

Children with Duchenne muscular dystrophy (DMD) suffer from progressive muscular weakness and increasing disability until death occurs, usually somewhere at the beginning of the third decade of life. Increasing disability with age causes difficulty walking that leads to becoming wheelchair bound at the



**Figure 30-4.** Bar graphs demonstrating increasing pulmonary impairment with increasing curve severity (A), increasing pulmonary impairment with increasing length of thoracic curve (B). (From Newton PO, Faro FD, Gollogly S, et al: Results of preoperative pulmonary function testing of adolescents with idiopathic scoliosis: a study of six hundred and thirty-one patients. *J Bone Joint Surg Am* 2005; 87:1937-1946.)

age of 8 to 10 years. The scoliosis then progresses with an acute deterioration during the growth spurt somewhere between the ages of 13 and 15 years, such that it becomes difficult or impossible to sit unaided. Once the lumbar curve exceeds 35 degrees, further progression becomes inevitable.<sup>37</sup>

## Risk Minimization and Improving Outcome from Surgical Intervention

### Respiratory Function

Children presenting for reconstructive spine surgery usually have some degree of respiratory impairment. Anesthesia for major surgery is associated with a number of changes in lung function that have the potential to cause problems postoperatively.

### Early Postoperative Period

Decreases in lung volumes and flow rates similar to thoracic and upper abdominal surgery occur after scoliosis surgery. The FVC and FEV<sub>1</sub> decrease with a nadir at 3 days and are about 60% of

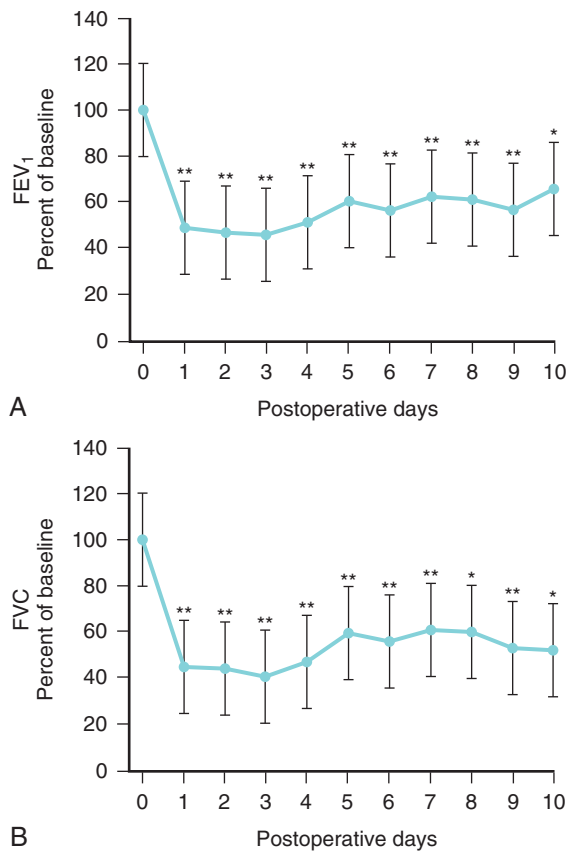
preoperative values 7 to 10 days after surgery (Fig. 30-5).<sup>38</sup> It is not until 1 to 2 months after surgery that pulmonary function tests reapproach baseline values. The magnitude of this decrease is not affected by the type of surgery performed (posterior spinal fusion versus combined anterior and posterior spinal fusion) or whether the scoliosis is due to an idiopathic or neuromuscular cause.<sup>38</sup>

Children with neuromuscular disease are more likely to require prolonged mechanical ventilation after spinal surgery because of more severe preoperative respiratory impairment.<sup>39</sup> The marked decrease in VC is undoubtedly related to the risk of postoperative complications, but determining when it is no longer safe to anesthetize children with a restrictive lung defect remains an imperfect science.

### Long-Term Changes

#### Idiopathic Scoliosis

Correction of idiopathic scoliosis improves the magnitude of the curve and cosmetic appearance, but improvements in pulmonary function are less impressive. Early studies in children with



**Figure 30-5.** **A**, Changes in FEV<sub>1</sub> during the 10 days after scoliosis surgery. **B**, Changes in FVC during the 10 days after scoliosis surgery. (From Yuan N, Fraire JA, Margetis MM, et al: The effect of scoliosis surgery on lung function in the immediate postoperative period. *Spine* 2005; 30:2182-2185.)

idiopathic scoliosis suggested that spinal fusion stabilized the respiratory dysfunction that existed preoperatively but failed to offer any improvement.<sup>40</sup> Improvements may be possible in certain subgroups of children with some surgical techniques, but it takes months to years for pulmonary function to improve. For example, children with a preoperative curve less than 90 degrees undergoing a posterior procedure had a greater than 10% increase in VC, maximum voluntary ventilation, and maximum respiratory mid flow rate after 2 years; this improvement was not seen in those who underwent anterior surgery.<sup>41</sup> Harrington rod instrumentation in children with idiopathic scoliosis has also been demonstrated to result in a small but statistically significant improvement in VC.<sup>42</sup> The newer instrumentation systems such as the Cotrel-Dubouset instrumentation that allow segmental realignment and approximation result in further improvements in pulmonary volumes.<sup>43</sup> A return to preoperative pulmonary function has been demonstrated within 3 months after the posterior approach using the newer instrumentation systems, with modest improvements occurring and being sustained at 2 years.<sup>44</sup> The use of pedicle screws in adolescent idiopathic scoliosis has been shown to result in a greater curve correction with a trend toward improved pulmonary function after 2 years when compared with other instrumentation techniques.<sup>21</sup>

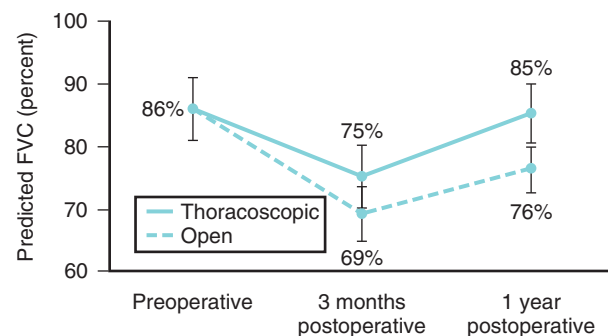
Chest cage disruption (thoracoplasty or anterior thoracotomy) is associated with reduced pulmonary function at 3 months with a 10% to 20% decrease in TLC and FVC. These values do not return to baseline until 1 to 2 years after surgery. Improvements in lung function with this approach are seldom seen.<sup>44,45</sup> A thoracoscopic approach to anterior release and instrumentation appears to result in less pulmonary morbidity and a smaller decrease in pulmonary function at 3 months. One year after surgery in children treated thoracoscopically values had returned to baseline, whereas this did not occur in those undergoing an open thoracotomy (Fig. 30-6).<sup>46</sup>

### Neuromuscular Scoliosis

Improvements in the scoliosis angle and the degree of pelvic obliquity are achieved after spinal instrumentation in children with neuromuscular disease. Significant improvement can usually be shown in the ability to sit unaided, particularly if children are unable to do so beforehand.<sup>47-51</sup> Some authors have also reported an increase in the quality of life perceived either by the child or the caregiver.<sup>49,51</sup>

The effects on lung function, however, are much more variable. There is little evidence for any improvement in respiratory function in this group of children. There may be a period of delay or even stabilization in the inevitable deterioration of respiratory function. One study demonstrated that respiratory function remains stable for 3 to 5 years after surgery in children with DMD, whereas an 8% decline per year occurs in those managed conservatively. Improved survival was also demonstrated in those undergoing surgery.<sup>52</sup> A slower rate of decline or lack of deterioration in respiratory function for 3 years after operation has also been reported.<sup>36,48</sup> These reports are countered by others that show either no difference in respiratory function after 5 years compared with those managed conservatively<sup>50,53</sup> or an early loss in vital capacity after surgery with a progressive decrease of 25% over 4 years with 66% of children requiring mechanical respiratory assistance by that time.<sup>49</sup>

Less morbidity has been claimed for the same day (one stage) surgery compared with the 2-staged approach in children with neuromuscular disease requiring anterior as well as posterior spinal surgery.<sup>54,55</sup> However, it would seem reasonable to try and avoid anterior thoracotomy on neuromuscular patients wherever possible in view of the poor respiratory function after chest cage disruption.<sup>44,45</sup>



**Figure 30-6.** Changes in percent of FVC for thoracoscopic versus open anterior instrumentation during first year after surgery. (From Faro FD, Marks MC, Newton PO, et al: Perioperative changes in pulmonary function after anterior scoliosis instrumentation: thoracoscopic versus open approaches. *Spine* 2005; 30:1058-1063.)

### Respiratory Complications

Respiratory complications after surgery in children with nonidiopathic scoliosis have a reported incidence five times that of idiopathic scoliosis.<sup>56</sup> The risk increases as the degree of curvature increases and the respiratory function decreases.<sup>57</sup> Anterior spinal procedures are associated with a greater incidence of complications than posterior spinal fusion, such that some consider this to be the main risk factor for postoperative respiratory complications.<sup>57</sup> Atelectasis, infiltrates, hemo/pneumothoraces, pleural effusions and prolonged intubation have the greatest incidence, whereas pneumonia, pulmonary edema, and upper airway obstruction occur less frequently. These problems occur more commonly when the scoliosis is associated with mental retardation and developmental delay. The greatest complication rate occurs in those with cerebral palsy.<sup>56</sup>

## Spinal Cord Injury during Surgery

### Etiology

Spinal cord injury can occur by four main mechanisms: direct contusion of the cord during surgical exposure; contusion by hooks, wires, or pedicle screws; distraction by rods or halo traction; and reduction in spinal cord blood flow.<sup>58</sup> Epidural hematoma should be included in the differential diagnosis of deficits occurring postoperatively. The areas of the spinal cord most vulnerable to ischemic injury are the motor pathways, which are supplied by the anterior spinal artery. This is fed in a segmental manner by the radicular arteries that arise from the vertebral, cervical, intercostal, lumbar, and iliolumbar arteries. The largest radicular artery is known as the artery of Adamkiewicz and arises between T8 and L4. A watershed area between T4 and T9 is prone to ischemia because the blood supply is poorest to this region of the cord.<sup>59</sup> Paraplegia is the most feared neurologic complication, but partial spinal cord injury resulting in areas of localized weakness and numbness as well as bladder and bowel disturbances have all been reported. The increasing use of pedicle screws in spine surgery also raises the possibility of increased risk to individual nerve roots.

### The Risk of Spinal Cord Injury and the Role of Spinal Cord Monitoring

Spine surgery is associated with a small incidence of neurological impairment. Combined surveys undertaken by the Scoliosis Research Society (SRS) and the European Society for Deformities of the Spine (EUSDS) suggested an incidence of 0.72% in 1975.<sup>60</sup> This incidence has decreased to less than half (0.3%) that reported 30 years ago, and all were partial cord lesions.<sup>58</sup> Children with curves greater than 100 degrees, congenital scoliosis, kyphosis, and post-radiation deformity appear to be at greatest risk for complications.

Monitoring spinal cord function is undertaken to ensure that the complication rate is as small as possible. The SRS has issued a position statement concluding that neurophysiologic monitoring can assist in the early detection of complications and can possibly prevent postoperative morbidity in children undergoing operations on the spine. For any monitoring technique to be effective, it needs to have a sensitivity and specificity that allows true changes to be recognized with a very low occurrence of both false-negative and false-positive results. Furthermore, such a test or technique must also produce its results in a timeframe that allows the problem to be reversed or prevented. Slight dif-

ferences and difficulties with reliable interpretation of these techniques explain why no one test has been universally adopted, although, increasingly, motor evoked potentials are becoming the predominant modality.

### Methods of Monitoring Spinal Cord Function

#### Wake-Up Test

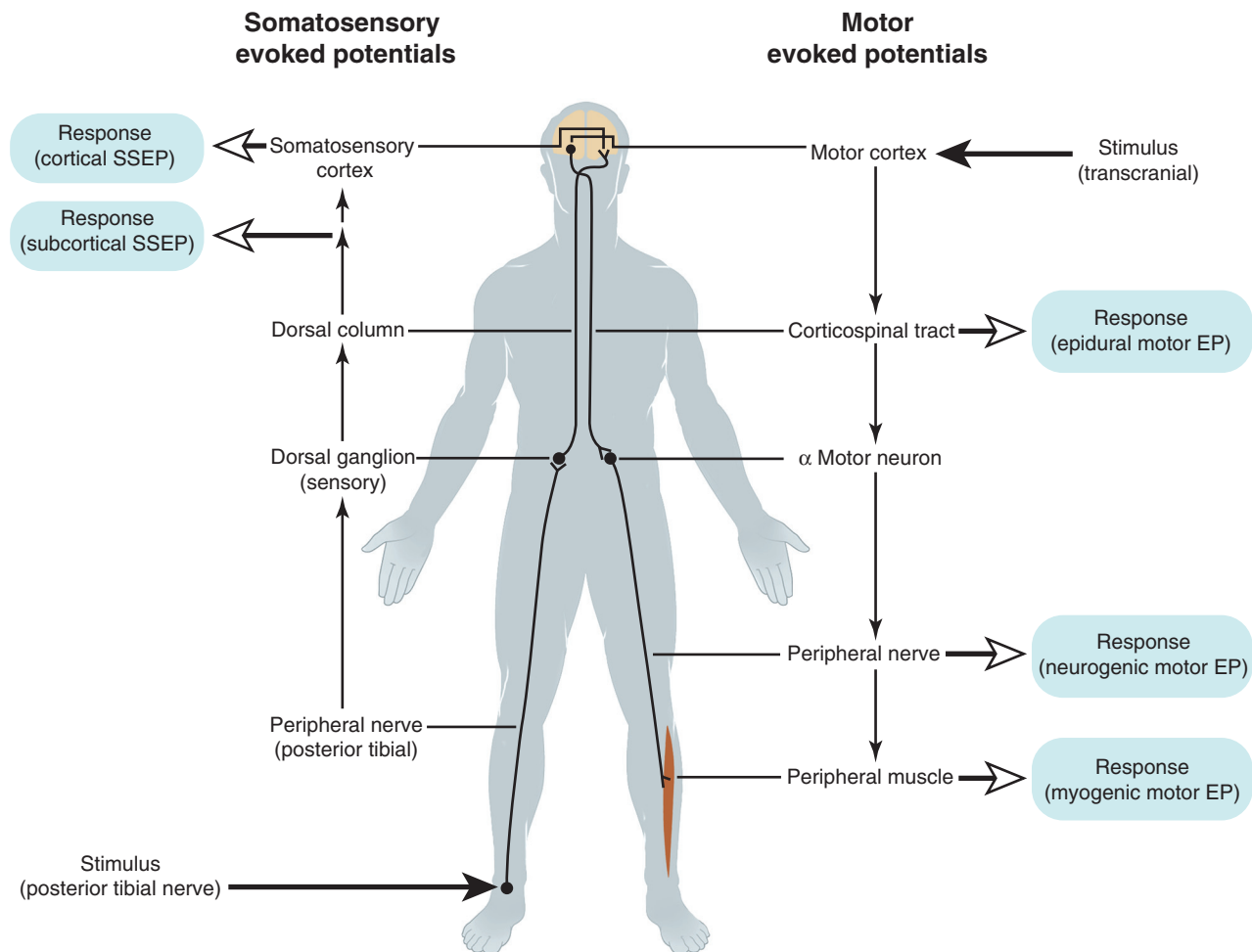
The wake-up test measures gross motor function of the upper and lower extremities. It has been in widespread use since it was first described.<sup>61</sup> The test consists of decreasing the depth of anesthesia almost to the point of wakefulness and asking the child to respond to verbal commands. Failure to move the feet and toes while being able to squeeze a hand suggests a problem with the spinal cord. The test requires the ability to limit or reverse muscle relaxation and lighten the anesthesia sufficiently to enable the child to follow commands. When the test was initially described, 3 of 124 children had a positive result (i.e., no movement) and were saved from paraplegia.<sup>61</sup> A major concern is that the test is conducted after maximal spinal correction, which may be a significant time after a neurologic insult has occurred. Removal or modification of the spinal instrumentation within 3 hours of the onset of the neurologic deficit has been reported to prevent the risk of permanent neurologic sequelae.<sup>62</sup> The wake-up test is unlikely to detect isolated nerve root injury or sensory changes. It is limited to neurologically normal children with an appropriate developmental age who can follow instructions. The use of somatosensory evoked potential (SSEP) and motor evoked potential (MEP) monitoring (Fig. 30-7) is now sufficiently developed that, in the absence of changes occurring intraoperatively, there is no need to perform the wake-up test.<sup>63</sup> It is still considered the standard by some surgeons and may be used to confirm changes demonstrated by SSEP or MEP monitoring.<sup>64</sup> Risks include accidental extubation, dislodgement of the instrumentation, intraoperative recall with subsequent psychological trauma, air embolism, and cardiac ischemia. If a wake-up test is to be performed, it is prudent to fill the wound with saline to reduce the potential for air embolization.

#### Ankle-Clonus Test

The ankle-clonus test uses the presence of clonus that occurs just before consciousness is regained during wakening from anesthesia. It is thought to be due to spinal reflexes returning while the higher centers remain inhibited by anesthesia and thus demonstrates an intact spinal cord. Inability to demonstrate clonus suggests spinal cord injury.<sup>65</sup> Like the wake-up test, it is a post hoc test rather than real-time monitoring. However, in a review of more than 1,000 patients undergoing spinal procedures in which six postoperative neurologic deficits occurred, this test identified all the deficits but produced three false-positive findings, giving a sensitivity of 100% and a specificity of 99.7%. In comparison, the wake-up test produced false-negative results in 4 of the 5 patients who developed deficits.<sup>65</sup>

#### Somatosensory-Evoked Potentials

Somatosensory-evoked potentials involve stimulating a peripheral nerve and measuring the response to that stimulation using scalp electrodes (cortical somatosensory-evoked potentials).<sup>66,67</sup> Alternatively, the response can be measured near the spinal cord (subcortical) by electrodes placed either in the epidural space, the interspinous ligament, or the spinous processes of the ver-



**Figure 30-7.** Comparison of pathways involved in SSEP and MEP monitoring. (Modified from de Haan P, Kalkman CJ: Spinal cord monitoring; somatosensory- and motor-evoked potentials. *Anesthesiol Clin North Am* 2001; 19:923-945.)

tebrae.<sup>68</sup> An intranasally placed pharyngeal electrode can act as a surrogate for these. The advantage of the subcortical evoked potential is that the responses are more stable, reproducible, and resistant to the effects of anesthesia. The signal produced with SSEP monitoring travels from the peripheral nerve via the nerve root and up the ipsilateral dorsal column. The impulses then cross over at the level of the brain stem and progress rostrally via the thalamus to the primary sensory cortex. The rationale for using SSEP as a monitor for concerns regarding motor deficits is based on the fact that the sensory tracts are in proximity to the motor tracts of the spinal cord. Injury to the motor tracts indirectly affects the sensory tracts and causes changes in the SSEP. When significant impairment of spinal cord function occurs there is usually an increase in latency and a decrease in amplitude in the SSEP, with eventual loss of signal. A 10% increase in latency of the first cortical peak (P1) or 50% decreases in the peak-to-peak amplitude (P1N1) constitute an indication for intervention.<sup>69,70</sup> Although SSEP signals primarily monitor transmission through the sensory dorsal columns there is a considerable body of evidence that demonstrates their effectiveness in detecting and therefore potentially reducing spinal cord injury.<sup>70</sup> SSEP monitoring is associated with a 50% decrease in the incidence of neurologic deficits. Although it is unusual for

motor tract injury to occur when SSEPs remain unchanged, a significant number of false positives and, more importantly, false negatives have been reported.<sup>70,71</sup> Seventy percent of the post-operative complications were detected by the monitor but 30% were not. These and other similar reports have led to the development of methods to monitor the motor tracts of the spinal cord.

#### Motor-Evoked Potentials

The motor pathways can be activated by transcranial stimulation of the motor cortex or spinal cord stimulation. Transcranial stimulation is achieved using electrical or magnetic stimulation applied to the scalp. Electrical stimulators are most commonly used in spine surgery and operate by applying high voltage pulses to the scalp using corkscrew, needle, or surface electrodes. The stimulation pulses can be applied as single stimuli or brief pulse trains with intervals between the pulse trains. Multiple stimuli result in a stronger signal with less variability due to temporal summation of the excitatory postsynaptic potential.<sup>72</sup> There is evidence that younger children require a higher stimulating voltage and pulse train frequency for MEP monitoring, which is thought to be due to immaturity of the CNS, specifically the descending corticospinal tracts.<sup>73</sup> Spinal

cord stimulation is achieved electrically and can be applied using electrodes placed either outside or inside the spinal cord rostral to the area of interest. Single stimuli rather than brief pulse trains are commonly used for spinal cord stimulation.<sup>74</sup>

Responses can be recorded anywhere distal to the area of interest. These have included the lower lumbar epidural space (epidural MEP), peripheral nerve (neurogenic MEP), and peripheral muscles using compound muscle action potential (CMAP) (see Fig. 30-7B).<sup>75</sup> Each recording site has its limitations regarding the accuracy of the information displayed and the susceptibility to anesthetic drug interference. Epidural MEPs are the least affected by muscle relaxants, but they only monitor conduction in the (cortico)spinal tract and provide no information about the anterior horn gray matter.<sup>76</sup> They have a much slower response to acute spinal cord ischemia when compared with myogenic responses (CMAP).<sup>77</sup> Neurogenic MEPs are also resistant to anesthetic interference but appear not to accurately measure motor conduction. Most of the spinally elicited peripheral nerve responses seen with neurogenic MEPs have been shown to occur via the dorsal columns in a retrograde fashion and are sensory rather than motor.<sup>78</sup> Furthermore, anterior spinal cord injury has occurred with normal neurogenic MEPs.<sup>79</sup> CMAPs after transcranial stimulation are believed to be exclusively generated via motor tract conduction and unlike epidural MEPs include the ischemia-sensitive anterior horn alpha motor neurons.<sup>75</sup> These responses are very sensitive to anesthetic agents. A total intravenous anesthetic (TIVA)  $\pm$  <0.5% MAC isoflurane may be requested. The responses obtained with CMAP after spinal cord stimulation also appear to contain signals that include transmission via the dorsal columns and may represent a mixed response.<sup>80</sup>

One outstanding problem with MEP monitoring is deciding when and how much change in the signal is significant and indicative of spinal cord ischemia. Some centers use the same criteria they have adopted for SSEP monitoring, whereas others require a greater degree of change, such as a 75% decrease in amplitude.<sup>81</sup> More recently, an amplitude decrease of 80% at one of six sites using transcranial myogenic MEP monitoring was demonstrated to have a sensitivity of 1.0 and a specificity of 0.91 when used as the sole monitor during spine surgery.<sup>82</sup> An alternative technique of measuring MEP has been described whereby a minimum threshold for producing a response is established and a significant increase in that threshold is used to indicate a problem.<sup>83</sup> Supportive data for this technique are lacking.

Whether MEP monitoring should replace SSEP monitoring, or whether the two should be used together, is a source of controversy at the time of writing. There is some evidence that the dorsal columns may be injured without involvement of the motor tract.<sup>84</sup> Occasionally, adverse changes in SSEP occur without changes in MEP but the association with neurologic deficit is unclear.<sup>74</sup>

## Preoperative Assessment and Postoperative Planning

### Respiratory Assessment and Planning for Postoperative Ventilatory Support

The preoperative pulmonary assessment should identify those children at increased risk from short-term postoperative pulmonary complications as well as long-term respiratory failure (or

failure to wean from a ventilator). Rigid statements regarding either issue are difficult because different surgical approaches, child and institutional variabilities, and author interest has resulted in heterogeneous reporting. Children with idiopathic scoliosis tend to have less decrease in pulmonary function with correspondingly low complication rates, so most studies have been directed at nonidiopathic children.<sup>56</sup> The rate of postoperative pulmonary complications correlates broadly with the decrease in VC.<sup>39,85,86</sup> VC less than 30% to 35% of predicted indicates marginal respiratory reserve and a level at which complications and a need for postoperative respiratory support are likely. Many children with these low vital capacities are unable to cough effectively, rendering them prone to postoperative atelectasis, pneumonia, and respiratory failure.

Studies involving a mixed population with a VC less than 40% (but with few neuromuscular patients) suggest that although short- and mid-term pulmonary complications occur, these children can be successfully managed to achieve discharge to home, although some will require prolonged postoperative ventilation.<sup>87,88</sup> Modest numbers of children with a VC less than 25% predicted are described in these studies and do not appear to have a higher complication rate than those with greater VC. Anterior or combined approaches increase the likelihood of respiratory complications, particularly due to pleural effusion.<sup>87,88</sup>

Children with neuromuscular scoliosis are at particular risk for postoperative ventilation that is often prolonged.<sup>39,86</sup> These children may also have abnormalities in the central control of breathing and impaired airway defense mechanisms. Impaired coordination of laryngeal and pharyngeal muscles may result in impaired swallowing and inadequate cough with increased risk of aspiration. Initial work suggested that as the VC decreased to less than 35% predicted, most children would need a brief period of postoperative ventilation.<sup>84</sup> Recent studies suggest that scoliosis surgery can be successfully undertaken in children with a VC of less than 35% predicted, often with no more than 24 hours of planned ventilation, followed by a period of noninvasive ventilation (e.g., bilevel positive airway pressure [BiPAP]).<sup>39,89,90</sup> The overall complication rate is similar whether the FVC is more or less than 30%, with an average hospital stay of approximately 3 weeks.<sup>90</sup> Again, the successful management of children with an FVC of less than 25% predicted is described. With the known changes in VC after surgery, it would seem reasonable to anticipate using noninvasive ventilator support for up to a week after spine stabilization surgery in these children. Children with neuromuscular scoliosis and early-onset respiratory failure requiring nocturnal noninvasive ventilation (BiPAP) have successfully undergone scoliosis surgery. A small group of children with a mean FVC of 20% predicted has been successfully managed with a short period of postoperative ventilation and transitioned to BiPAP within 48 hours without any respiratory complications.<sup>91</sup>

Whether a child should be denied surgery requires consideration of individual patient factors. The successful management of a few children with a VC of 15% to 20% predicted have been described, but the numbers are small.<sup>87-90</sup> Although the risk of an unsuccessful outcome will increase at this level, individual circumstances may justify that risk.

### Cardiovascular Assessment

Muscle disorders may affect the myocardium as well as the skeletal system. Children with DMD develop a cardiomyopathy

that may be difficult to evaluate because the child is wheelchair bound. Sinus tachycardia is an early manifestation, and evidence of decreased cardiac function increases in frequency and severity from early adolescence.<sup>92</sup> Over 90% of adolescents with DMD have subclinical or clinical cardiac involvement.<sup>93</sup> Echocardiography is an essential part of the evaluation of any wheelchair-bound child presenting for scoliosis surgery.

### Postoperative Pain Management

Scoliosis surgery is associated with severe pain that lasts for at least 3 days.<sup>94</sup> Analgesia minimizes postoperative respiratory complications by allowing deep breathing, chest physiotherapy, early ambulation, and rehabilitation. The two broad approaches to postoperative pain management are either systemic or epidural analgesics. A multimodal approach is likely to be most effective.

Intraoperative intrathecal morphine has been demonstrated to provide potent analgesia during the first 24 hours after spinal fusion in children.<sup>95</sup> Intrathecal morphine also decreases the amount of remifentanyl required intraoperatively and so may minimize the development of acute opioid tolerance that may be seen after the use of high-dose remifentanyl infusions.<sup>96,97</sup>

### Nonsteroidal Anti-inflammatory Drugs

NSAIDs, but not acetaminophen, impair fracture healing in animal models.<sup>98</sup> Cyclooxygenase-2 activity plays an important role in bone healing, and the use of NSAIDs decreases osteogenic activity that may increase the incidence of nonunion after spinal fusion.<sup>89</sup> The effect on osteogenic activity is dose dependent and reversible.<sup>99</sup> Similar effects have not been demonstrated in humans. Nonetheless, based on animal evidence, NSAIDs should be used with caution and in consultation with the surgeon during the first 3 to 5 days after scoliosis surgery.<sup>100</sup>

### Systemic Analgesics

Morphine is the mainstay of systemic analgesic regimens. Morphine infusions of 20 to 40  $\mu\text{g}/\text{kg}/\text{hr}$  are required during the first 48 hours after surgery. Achieving a balance of effective analgesia while avoiding incapacitating sedation can be difficult in children with neurodevelopmental delay. Regular evaluation of these children is important if complications are to be avoided. Patient-controlled analgesia (PCA) is appropriate for children older than the age of 6 to 7 years and can be used with a typical bolus dose of 20  $\mu\text{g}/\text{kg}$  and a lockout interval of 5 to 10 minutes. The use of a background morphine infusion may be effective in some children, although its inclusion is controversial.<sup>101,102</sup> Our preference is to add a night-time background infusion at 5- to 10- $\mu\text{g}/\text{kg}/\text{hr}$  but to use PCA alone during the day (see Chapter 44). Nurse- and parent-controlled analgesia have also been shown to be effective if the child is too young or unable to use PCA.<sup>103</sup> A low-dose ketamine infusion (0.05-0.2  $\text{mg}/\text{kg}/\text{hr}$ ) has been used as an adjunct to morphine infusions or PCA, although its role is debated<sup>104-109</sup>; its use is generally reserved for those with morphine-resistant pain. Ketamine may be initiated intraoperatively (an infusion of 5  $\mu\text{g}/\text{kg}/\text{min}$ , decreasing to 2  $\mu\text{g}/\text{kg}/\text{min}$  at the end of surgery) as part of the anesthetic technique to minimize the hyperalgesia reported after high-dose remifentanyl infusions.<sup>110,111</sup> If added to PCA, the optimal combination of morphine/ketamine is 1:1.<sup>107</sup>

### Epidural Analgesia

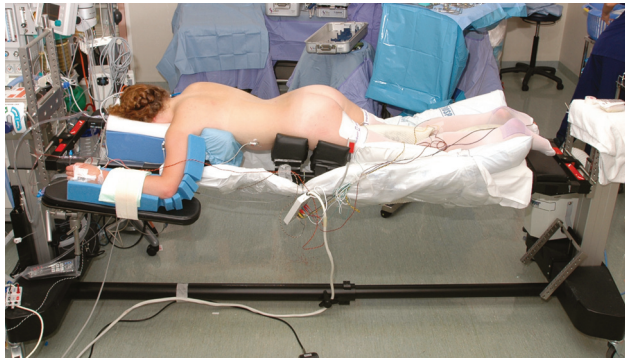
Continuous epidural analgesia, using both single- and double-catheter techniques, may provide effective analgesia after spine surgery. The single-catheter technique using bupivacaine-fentanyl and sited at T6-T7 for children undergoing a mean 12-level scoliosis surgery correction has been reported to provide similar analgesia to PCA. Bowel sounds returned earlier in the epidural group, but liquid intake and hospitalization were similar.<sup>112</sup> Similar results were reported with a bupivacaine-morphine combination in children undergoing 10-level spinal fusions. Full diet and discharge from hospital were achieved half a day earlier with the epidural technique than with PCA.<sup>113</sup> A retrospective review of more than 600 patients treated with either epidural analgesia or PCA for postscoliosis analgesia, in which an average number of segments fused was 8.5, confirmed the effectiveness of epidural analgesia.<sup>114</sup> In that study a bupivacaine-hydromorphone epidural combination was used and although pain management was effective, more complications occurred in the epidural group. Respiratory depression and transient neurologic changes were the most common complications observed. Thirteen percent of adolescents with an epidural catheter required discontinuation of the epidural, most commonly for inadequate pain relief.<sup>114</sup> Patient-controlled epidural analgesia (PCEA) has been successfully used in children older than age 5 years for orthopedic surgery and thoracotomies but experience after scoliosis surgery is limited.<sup>115</sup>

Double epidural techniques comprise an upper catheter positioned in the upper or mid thoracic segments and a lower catheter at the mid lumbar level. Initial reports of this technique involved a ropivacaine-hydromorphone mixture with catheters that were in situ for 5 postoperative days without adverse effects.<sup>116,117</sup> Ropivacaine, 0.3% 10 mL/hr, for nine-segment scoliosis surgery improved pain scores both at rest and with movement compared with a morphine infusion. Bowel activity returned earlier, and a decreased incidence of postoperative nausea and vomiting was observed in the epidural group.<sup>118</sup>

## Anesthetic and Intraoperative Management

### Positioning and Related Issues

It is essential that the child is positioned so that extreme pressure points are avoided, the limb positions are adjusted to prevent nerve injury, and the abdomen is free, to minimize venous congestion. This is usually achieved by the use of the Relton-Hall frame or a variant.<sup>119</sup> The frame comprises four well-padded supports arranged into V-shaped pairs with the upper pads supporting the thoracic cage and the lower pair supporting the anterolateral aspects of the pelvic girdle at the anterior iliac crests. The arms should not be abducted or extended greater than 90 degrees from their natural position, and the weight of the arms should be evenly distributed across the forearm to avoid pressure on the ulnar nerve at the elbow. This can present quite a challenge in children with severe deformities, and creative positioning may be required. In some centers, the nipples are covered with Tegaderm (3M, St. Paul, MN) and positioned free of direct pressure. It is also essential that the head is maintained in a neutral position and that pressure is evenly distributed between the forehead and face, avoiding direct pressure on the eyeballs. Care must be taken to avoid



**Figure 30-8.** Positioning on the OSI Jackson frame showing protected pressure points and underframe forced air warming blanket.

any direct pressure on the knees, with distribution of the child's weight spread throughout the lower limb (Fig. 30-8). Reston (3M, St. Paul, MN) may be used to pad the pelvic brim and knees.

Not all spinal tables and frames affect cardiac function in the same way. There is some evidence that the Jackson spine table or longitudinal bolsters have minimal effects on cardiac function, whereas Wilson, Siemens, and Andrews frames may negatively impact cardiac function.<sup>120</sup>

Postoperative visual loss is an uncommon, unpredictable, but devastating complication associated with spinal surgery. The incidence may be as great as 0.2% of cases; and although most of the reports involve adult patients, older children are not immune.<sup>121,122</sup> The most common cause is ischemic optic neuropathy, but the etiology remains obscure. Prolonged operating time (>6 hours) and increased or uncontrolled blood loss are a feature of most of the reports.<sup>123-126</sup> The phenomenon is unrelated to pressure on the globe and usually occurs without evidence of any other ischemia-related complications.<sup>126</sup> There are no data to support controlled hypotension or hemodilution as contributory factors despite occasional "expert" opinions to the contrary.<sup>123,126,127</sup>

### Temperature Regulation

The long preparation time and exposure of an undraped child on the spinal frame render children susceptible to hypothermia. Hypothermia is associated with hemodynamic instability and increased blood loss.<sup>128</sup> A threefold increase in surgical wound infection occurs with a 2°C decrease in core temperature.<sup>129</sup> Efforts should be made to increase the ambient temperature in the operating room while the child is prepared for surgery. Subsequent hypothermia will be minimized if the room temperature is maintained at 24°C during this period rather than 18 to 21°C as is often encountered during surgery.<sup>130</sup> Once the child has cooled during preparation and positioning, it may take several hours before the core temperature begins to return toward normal. Even with forced air warming systems, it is often difficult to restore normothermia because only a small amount of the child's body is exposed to these devices. It may be possible to position a warming blanket underneath the frame so that warming from below as well as from above occurs (see Fig. 30-8).

### Patient Monitoring

Patient monitoring needs to be tailored to the individual case, but, at a minimum, hemoglobin oxygen saturation, end-tidal CO<sub>2</sub>, systemic blood pressure, electrocardiographic findings, core temperature, and urine output should be recorded. In most cases invasive arterial and central venous pressures are monitored because of large blood losses, fluid shifts, and the risk of cardiovascular instability. Direct pressure by the surgeon during either dissection or curve correction may compromise cardiac function or filling. Central venous pressure is an accurate and valid measurement in the prone position, providing the zero is adjusted for the child's position on the spinal frame. Children with a significant kyphotic component are at increased risk of venous air embolism and should be monitored for this possibility. Depth of anesthesia monitoring should be considered, particularly when MEP monitoring limits the concentrations of anesthetic drugs. Care should be taken with positioning the head because pressure on the forehead by the sensor while the child is in the prone position for many hours can cause erythema, localized swelling, and possible tissue necrosis. Contact dermatitis from the adhesive has also been reported.<sup>131</sup> Transesophageal echocardiography can be useful for determining ventricular filling and function when hemodynamic compromise is identified or suspected preoperatively.

### Minimizing Blood Loss and Decreasing Transfusion Requirements

Scoliosis surgery involves exposure of a large wound over a considerable period of time. Positioning the child with the abdomen free to avoid venous compression is important to control and minimize blood loss. Increased intra-abdominal pressure attributable to positioning can double intraoperative blood loss.<sup>132</sup>

The reported estimated blood loss (EBL) for this type of surgery varies from institution to institution and from one surgical technique to another. Posterior spinal fusion procedures tend to lose more blood than anterior procedures. This loss is probably due to the greater number of vertebral levels fused with the posterior approach. Blood loss increases as the number of vertebrae included in the fusion increases. The EBL is 750 to 1500 mL in children with idiopathic scoliosis, which equates to 60 to 150 mL per vertebral segment fused. The blood loss is significantly greater in children with cerebral palsy—1300 to 2200 mL—which equates to 100 to 190 mL per level. Children with DMD have the greatest EBL of 2500 to 4000 mL, which equates to an EBL of 200 to 280 mL per vertebral level.<sup>133</sup>

Children with neuromuscular scoliosis demonstrate a prolonged prothrombin time and a decrease in factor VII activity intraoperatively, suggesting that consumption of clotting factors as well as dilution of clotting factors enhances the blood loss.<sup>134</sup> It has been postulated that children with DMD lack dystrophin in all muscle types and that the poor vascular smooth muscle vasoconstrictor response may be a factor in the increased blood loss.<sup>135</sup> Hypothermia exacerbates blood loss by decreasing platelet function, decreasing coagulation factor activity, and slowing vasoconstriction.<sup>128</sup>

### Hypotensive Techniques

Controlled hypotension has been used to minimize blood loss during scoliosis surgery. A greater than 50% decrease in blood loss with a decreased need for blood replacement, as well as a

reduced operating time, was demonstrated in early studies. Ganglion-blocking agents (pentolinium and trimethaphan) have been superseded by  $\beta$  blockers, direct arterial vasodilators, calcium-channel blockers, and  $\alpha_2$  agonists. Evidence from studies in which sodium nitroprusside and trimethaphan were compared suggests that the mean arterial pressure and not cardiac output determines intraoperative blood loss.<sup>136</sup> On the other hand, a study in which a  $\beta$  blocker and nitroprusside were compared suggests that a slower heart rate and hence cardiac output is associated with reduced blood loss.<sup>137</sup> A target mean arterial pressure (MAP) of 50 to 65 mm Hg has been recommended. Although this appears to be safe from published data and knowledge of the autoregulation of cerebral and spinal cord blood flow, there is no doubt that the margin of safety for cerebral and spinal cord ischemia is reduced by controlled hypotension. This is of particular concern when this target MAP is used in long operations, with the potential for periods of hypovolemic hypotension in addition to drug-induced (controlled) hypotension.<sup>138</sup> From this stems concern that low MAP may be associated with cerebral ischemia, spinal cord ischemia, and end-organ failure.<sup>139</sup> The incidence of these feared complications is fortunately very low with or without the use of controlled hypotension. Renal function appears well preserved even when hypotensive anesthesia is used during scoliosis surgery, but hypotension may cause decreased SSEPs used as a monitor and indicator of spinal cord function.<sup>140,141</sup> Because of these concerns and the concomitant use of hemodilution, less extreme degrees of hypotension are usually employed today. This can often be achieved without the use of specific vasoactive drugs. In most cases, adequate hypotension can be achieved using a remifentanyl infusion titrated to the desired blood pressure without concerns of prolonged blood pressure or sedation effects.<sup>142</sup>

Clonidine is a useful adjunct for reducing blood pressure in children.<sup>143</sup> We have found that using an anesthetic combination of less than 1 MAC of inhalation agent plus remifentanyl with clonidine (2  $\mu$ g/kg) results in controlled hypotension in most children without the need for any additional agents.

Although not considered a hypotensive agent, intrathecal morphine decreases blood loss and may facilitate blood pressure control, particularly with a remifentanyl infusion. At an analgesic dose of 5  $\mu$ g/kg, a decrease in EBL from 41 to 14 mL/kg occurred.<sup>95</sup> Again, using this technique, blood pressure control can often be achieved without any additional agents.

Nicardipine has been used to produce hypotension in children undergoing spinal surgery and is associated with less blood loss at the same mean arterial pressure when compared with sodium nitroprusside.<sup>144,145</sup> A slower return to baseline blood pressure (27 vs. 7 minutes) is the problem with this drug and may be the reason it is not widely used. If a hypotensive technique is to be used, then invasive arterial and central venous pressure catheters are essential for the safe conduct of anesthesia (see Chapter 10).

### Hemodilution

Decreasing the hemoglobin concentration by removing red cells and replacing the volume with a combination of crystalloid and albumin means that for a given volume loss there is less red cell loss (see Chapter 10). The decreased metabolic rate during anesthesia implies that oxygen delivery can be maintained with a lower hemoglobin concentration providing normovolemia is maintained. In many cases, deciding on the degree of hemodilu-

tion and establishing a threshold for transfusion is difficult. Many clinicians use a hematocrit below 20% to 25% (hemoglobin of 7 to 8 g/dL) as the trigger for transfusion. At this hemoglobin concentration, tachycardia and hemodynamic instability frequently first appear. More extreme degrees of hemodilution have been described, but the decrease in oxygen-carrying capacity reduces the margin of safety to prevent cerebral and spinal cord ischemia. Myocardial ischemia becomes a risk at hemoglobin concentrations less than 5 g/dL.<sup>146</sup> Cyanosis cannot occur at these levels because 5 g/dL of desaturated hemoglobin is required for cyanosis to be observed. Many children demonstrate tachycardia and circulatory instability at a hemoglobin in excess of this level, so extreme hemodilution techniques such as these are reserved for Jehovah's Witnesses and those who are opposed to blood transfusion. In one report, children were hemodiluted during scoliosis surgery to a hemoglobin concentration of 3 g/dL in the absence of any preexisting cardiac disease.<sup>147</sup> Presumably, adequate oxygen transport was achieved by an increase in oxygen extraction and an increase in cardiac output. Cardiac output increased by over 30% with only a modest increase in heart rate and decrease in blood pressure.<sup>147</sup> Although no cerebral sequelae were reported, *this degree of extreme hemodilution is not recommended.*

Hemodilution modeling in adult patients has suggested that as many as 5 units of blood need to be removed before there is a decrease in transfusion requirements.<sup>148</sup> This is significantly greater than the usual 2 to 3 units removed in most adult-sized children. Despite these theoretical concerns, a relatively conservative hemodilution strategy is commonly and readily employed in children with idiopathic scoliosis. Reduction to an initial hematocrit of 30% has been shown to be effective in reducing and minimizing transfusion requirements.<sup>149</sup> These numbers must be tailored for each child because a unit of blood in a 35-kg child is a much larger fraction of the circulating blood volume than in a 70-kg child.

### Autologous Pre-donation

Alternately, several other preoperative strategies may be used, including pre-donation of blood and preoperative red cell augmentation. In the latter case, sequestration of several units of blood after induction of anesthesia but before surgery can be achieved provided the child's hemoglobin concentration has been increased to 16 to 18 g/dL preoperatively with parenteral erythropoietin, oral iron, and vitamin C (see Chapter 10). Sequestration of several units of blood can be achieved (and returned to the circulation as needed) without decreasing the hemoglobin concentration to values that put the brain, spinal cord, and heart at risk for adverse sequelae.

### Antifibrinolytic Agents

The use of synthetic antifibrinolytic agents to decrease perioperative blood loss after scoliosis surgery has produced mixed results. It should be emphasized that for an antifibrinolytic to be most effective an effective plasma concentration should be established before skin incision.  $\epsilon$ -Aminocaproic acid (EACA) has been shown to decrease the EBL by 25% during the perioperative period,<sup>150</sup> mainly attributable to decreasing postoperative suction drainage.<sup>151</sup> In contrast, tranexamic acid, 10 mg/kg followed by an infusion of 1 mg/kg/hr, failed to significantly decrease blood loss in a small sample.<sup>152</sup> High-dose tranexamic acid (100 mg/kg loading dose followed by an infusion of 10 mg/kg/hr) did decrease blood loss by 40% but did not affect

transfusion requirements. Post hoc analysis in children with secondary (neuromuscular) scoliosis showed significant reduction in both blood loss and transfusion requirements.<sup>153</sup> Aprotinin,\* in a dose approximating the full dose “Hammersmith” regimen in adults (240 mg/m<sup>2</sup> loading dose followed by an infusion of 56 mg/m<sup>2</sup>/hr for children with body surface area (BSA) <1 m<sup>2</sup> or 280 mg loading dose followed by an infusion of 70 mg/hr for BSA >1 m<sup>2</sup>) has been associated with a 40% decrease in blood loss, which equated to a decrease from 76 to 38 mL per vertebral level fused. This decrease in blood loss with aprotinin also resulted in fewer units of blood transfused (2.2 [control] vs. 1.1 units [aprotinin]).<sup>154</sup> In adult patients undergoing complex spine surgery, using approximately one half “Hammersmith” dose, aprotinin effectively reduced the blood loss, blood component therapy after surgery, and was associated with a decrease in respiratory morbidity. This was not observed in patients treated with EACA.<sup>155</sup> Aprotinin may not be indicated for all children because it is expensive and carries allergic risks, but it may be appropriate for those at risk for postoperative pulmonary complications and for those undergoing complex curve corrections.

Desmopressin probably has no beneficial effect on decreasing blood loss associated with spinal surgery. Initial beneficial results<sup>156</sup> have not been reproduced in children with idiopathic scoliosis<sup>157,158</sup> or in those with neuromuscular scoliosis.<sup>159,160</sup>

#### Intraoperative Salvage of Shed Blood

Decisions concerning the use of intraoperative salvage of shed blood (cell saver) are dependent on the anticipated blood loss, size of the child, and use of other methods to minimize blood transfusion, such as pre-donation and hemodilution (see Chapter 10). The addition of a cell saver was found to be beneficial in less than 5% of adolescents with idiopathic scoliosis involved with either an autologous pre-donation program and/or modest intraoperative hemodilution.<sup>149</sup> The technique is beneficial in children with a smaller body weight and greater anticipated blood loss such as children with neuromuscular scoliosis undergoing extensive spinal fusion.<sup>161,162</sup>

#### Managing Blood Loss

Using autologous blood requires an organized schedule of donation with or without the administration of erythropoietin.<sup>163,164</sup> This may be the safest and most effective method of avoiding or minimizing the use of allogenic blood products in this group of children.<sup>165</sup> A pre-donation program was effective in minimizing blood exposure in idiopathic adolescents undergoing surgical correction for their scoliosis; a mean of 3.7 units of blood was donated by each child before surgery, and 97% of adolescents avoided the use of allogenic blood during and after surgery.<sup>163</sup>

Measurement of blood loss during scoliosis surgery is difficult. Accuracy is lost as measurements embrace blood suctioned from the operative field that includes irrigation fluid, weighing or estimating blood collected on swabs and sponges, approximations of blood on drapes and gowns, as well as consideration of evaporation from the wound.

The decision when to administer blood component therapy (i.e., non-red cell blood components) is often based on clinical

\*Aprotinin has recently been removed from all hospital pharmacies and warehouses and is not available for purchase in the United States. It is available as an investigational drug only under certain guidelines. Please see <http://www.fda.gov/CDER/DRUG/infopage/aprotinin/default.htm>

judgment. Dilutional thrombocytopenia would be expected only after several blood volumes have been lost and depends on the initial platelet count before surgery (see Chapter 10). Platelet concentrations should be measured after loss of one blood volume and at periodic intervals after this. Dilution of coagulation factors may also lead to surgical bleeding when packed red blood cells only are used to replace blood loss. Prolongation of prothrombin time and activated partial thromboplastin time may occur when the blood loss exceeds one blood volume and should be checked at this time. These coagulation tests are not usually associated with increased bleeding until values are greater than 1.5 times mean control values, at which time increased surgical bleeding can be effectively treated with fresh frozen plasma.<sup>166</sup> Platelet counts after one blood volume loss, whether associated with normal or abnormal clotting, were within the normal range.<sup>166</sup> Blood component therapy should probably be based on abnormal clotting tests, uncontrolled bleeding, or the absence of normal clotting in the surgical field. It is preferable to intervene with blood component therapy before uncontrolled bleeding develops. If pooled blood in a dependent part of the operative field fails to show evidence of clotting, it is time to transfuse with blood components starting with fresh frozen plasma and only administering platelets if this is not effective.<sup>166</sup>

Recombinant factor VIIa may be a useful therapy for children with a dilutional coagulopathy and who do not respond to blood component replacement therapy. Successful use has been described in two children with neuromuscular scoliosis, but its use for this indication remains unapproved.<sup>167</sup>

#### Anesthetic Agents: Effect on SSEP and MEP

Spinal cord monitoring is an integral part of providing care for children undergoing scoliosis surgery. Knowledge of the effects of drugs on evoked potentials is pivotal for developing a successful anesthetic regimen. Anesthetic agents produce their effects by either directly inhibiting synaptic pathways or indirectly changing the balance of inhibitory and excitatory influences.<sup>168,169</sup> In general terms, the greater the number of synapses and the more complex the neuronal pathway being monitored, the greater the potential impact from anesthetic agents. Most anesthetic agents depress the amplitude and increase the latency of both the SSEP and MEP. For this reason, cortical SSEPs are more sensitive than spinal cord or brainstem-measured SSEPs. MEPs are susceptible to anesthetic agents at three sites: the motor cortex, the anterior horn cell, and the neuromuscular junction. Consequently, transcranial stimulation with peripheral muscle detection (using CMAP) is most susceptible to anesthetic interference. Although inhalational anesthetics and most intravenous anesthetics markedly depress SSEP and MEP, ketamine and etomidate appear to enhance the amplitudes of both, possibly by attenuating inhibition.<sup>169</sup>

#### Inhalational Anesthetics

The inhalational anesthetics cause dose-dependent depression of the SSEP and MEP; myogenic MEP is affected to a greater degree than SSEP. This means that while inhalation agents can be used during SSEP monitoring, they often need to be used in subanesthetic doses during MEP monitoring. Adequate cortical SSEPs and subcortical SSEPs can be measured with up to 1 MAC of isoflurane, sevoflurane, and desflurane, although some increase in latency and decrease in amplitude may be

detected.<sup>170,171</sup> It is important to maintain constant end-tidal concentrations throughout the anesthetic once baseline measurements have been established. The concentrations of these anesthetics that allow adequate monitoring are significantly lower than was possible with halothane.<sup>172</sup>

Myogenic MEPs (CMAPs) are only recordable at low concentrations of inhalation anesthetics. The exact concentration depends on the system being used and is greatly influenced by the number of pulses in the stimulus. Single-pulse transcranial stimuli may be inhibited by end-tidal concentrations as low as 0.2 MAC and abolished by end-tidal concentrations as low as 0.5 MAC.<sup>173-175</sup> This suppression can be partially overcome by using higher-intensity stimuli with multiple-pulse stimulation of up to six pulses per stimulus. An increasing number of children lose recordable myogenic MEPs, even when multiple-pulse stimuli are used as the concentration of inhalational anesthetic exceeds 0.5 MAC. At end-tidal concentrations in excess of 0.75% isoflurane, monitoring conditions may become unacceptable.<sup>176-180</sup> Stimulus intensity as well as pulse train frequency is probably a factor in determining successful myogenic MEPs with inhalational anesthetics. Using direct stimulation of the cortex during craniotomy, CMAP was easily recordable at 1 MAC of both isoflurane and sevoflurane.<sup>181</sup> Similar results have been demonstrated with sevoflurane using transcranial stimulation.<sup>182,197</sup> Information regarding desflurane is limited; although it causes a dose-dependent depression, myogenic MEPs have been successfully recorded at 0.5 MAC.<sup>180,183</sup> With the use of a multiple-pulse stimulation technique, intraoperative recording of MEPs was equally successful during desflurane or propofol anesthesia.<sup>184</sup> In contrast to its effects on SSEPs, halothane depresses myogenic MEPs to a lesser extent than the newer inhalational anesthetics.<sup>182</sup>

### Nitrous Oxide

Nitrous oxide reduces the amplitude of the cortical SSEP, but comparisons with other inhalational anesthetics are limited. Nitrous oxide, 0.5 MAC, depresses SSEPs to a greater extent than isoflurane at a similar MAC.<sup>185</sup> Similarly, nitrous oxide, 66%, depressed SSEPs to a greater extent than propofol 6 mg/kg/hr (100 µg/kg/min).<sup>186</sup> Nitrous oxide also depresses myogenic MEPs.<sup>171</sup> The effect relative to inhalational anesthetics is difficult to determine. Nitrous oxide appears to affect CMAP amplitude to a lesser extent than isoflurane.<sup>187</sup> Multiple-pulse stimulus techniques can partially reverse nitrous oxide-induced depression of amplitude. Compared with a propofol infusion designed to maintain a target concentration of 3 µg/mL, 50% nitrous oxide decreased CMAPs with both single and paired stimuli to a lesser extent.<sup>188</sup> When 60% nitrous was added to low-dose propofol infusion at a target concentration of 1 µg/mL, adequate CMAPs were obtained using multiple-pulse transcranial stimulation.<sup>189</sup> Conversely, the addition of nitrous oxide to a variety of different total intravenous techniques significantly depressed the CMAP such that some were not recordable.<sup>190</sup>

With the widespread availability of remifentanyl and the variable but mostly negative effects of nitrous oxide on SSEP and MEP signals, it would seem that nitrous oxide is best avoided when spinal cord monitoring is used.

### Propofol

Propofol produces a decrease in amplitude of the cortical SSEP, but adequate signals can be recorded, even in the presence of nitrous oxide, at doses used for anesthesia (6 mg/kg/hr).<sup>191</sup> Pro-

propofol better preserves cortical SSEP amplitude and provides a deeper level of hypnosis as measured by processed electroencephalographic values than combinations of low-dose isoflurane/N<sub>2</sub>O or low-dose isoflurane or sevoflurane alone.<sup>192-194</sup>

Propofol depresses the amplitude of myogenic MEPs. In addition to its cortical effect, it also suppresses activation of the alpha motor neuron at the level of the spinal gray matter.<sup>195,196</sup> Low-dose propofol infusions have become popular as part of the anesthetic technique with MEP monitoring owing to the rapid improvement of signals when the drug is terminated and because multiple-pulse stimulation techniques can improve the response amplitude.<sup>177,178</sup> Propofol, even in combination with nitrous oxide, depresses multiple-pulse transcranial CMAP less than isoflurane.<sup>177</sup> Propofol, 5 mg/kg/hr, combined with 66% nitrous oxide produced satisfactory CMAP recordings in 75% of patients when a four-pulse stimulation sequence was used. In contrast, no recordings were possible with 1 MAC isoflurane.<sup>178</sup> The infusion rates or target concentrations that allow acceptable myogenic MEP recordings vary considerably and reflect different adjuvants (e.g., opioids, ketamine, and nitrous oxide), degrees of neuromuscular blockade, and transcranial pulse rates. Propofol at a target of 4 µg/mL or at an infusion rate of 6 mg/kg/hr produces acceptable signals with multiple-pulse stimuli.<sup>198,199</sup>

### α<sub>2</sub>-Adrenoreceptor Agonists: Clonidine and Dexmedetomidine

The cerebral effects of the α<sub>2</sub> agonists appear to be mainly at the locus coeruleus rather than by the more generalized inhibition of synaptic pathways, as in the case of general anesthetics.<sup>200</sup> Clonidine at intravenous doses of 2 to 5 µg/kg had minimal effects on cortical SSEPs when added to isoflurane.<sup>201-203</sup> In view of its lack of effect on SSEPs and its anesthetic sparing properties with both inhalational agents and propofol,<sup>203-205</sup> it seems reasonable to consider clonidine at a dose of 2 to 4 µg/kg as part of an anesthetic technique. Dexmedetomidine appears to have similar beneficial properties on SSEPs.<sup>206,207</sup> There are no published studies on the effects of clonidine or dexmedetomidine on MEPs, but one might speculate that they would improve signal recordings of MEPs by allowing lower concentrations of inhalational anesthetics or propofol.

### Opioids

Alfentanil, fentanyl, sufentanil, and remifentanyl produce minimal effects on SSEP and MEP signal recording.<sup>208,209</sup> Dose-dependent depression of CMAP does occur at doses of opioids that far exceed those used in clinical anesthesia.<sup>210,211</sup> Comparison of alfentanil, fentanyl, and sufentanil at doses sufficient to suppress noxious stimuli suggested that sufentanil exerted the least effect.<sup>210</sup> A similar study including remifentanyl showed that this drug had the least depressive effects, with CMAPs measurable at infusion rates of 0.6 µg/kg/min.<sup>211</sup> It is likely that larger doses can be used if clinically indicated.

### Ketamine and Etomidate

Ketamine enhances cortical SSEP amplitude and has a minimal effect on subcortical and peripheral SSEP responses.<sup>212</sup> It also produces minimal effects on the myogenic MEP responses, either as a bolus of 0.5 mg/kg<sup>213</sup> or when used in moderate doses (1-4 mg/kg/hr) as a supplement to a nitrous oxide/opioid anesthesia.<sup>213,214</sup> Experimental evidence suggests S(+)-ketamine modulates CMAP by a peripheral mechanism at or distal to the spinal alpha motor neuron.<sup>215</sup>

Etomidate, although capable of inducing general anesthesia, behaves more like ketamine in its effect on evoked potentials. It improves the quality of SSEPs and enhances the amplitude of MEPs.<sup>216</sup> It produces minimal changes in MEPs when compared with barbiturates or propofol.<sup>195</sup> Etomidate infusions (10–35 µg/kg/min) produce adequate MEP monitoring signals.<sup>213,217</sup> Concerns regarding adrenocortical depression with etomidate infusions remain and limit its widespread use.<sup>218</sup> Bolus doses of etomidate however, can transiently depress MEPs.<sup>213</sup>

### Midazolam

Intravenous midazolam (0.2 mg/kg) decreases the SSEP amplitude by 60%.<sup>219</sup> This does not seem to occur with subcortical SSEPs, in which a slight increase in latency but no change in amplitude has been demonstrated.<sup>220</sup> Although midazolam (0.5 mg/kg) caused marked depression of MEP in monkeys that persisted during awakening,<sup>221</sup> this does not hold true in human studies. MEP amplitude was not affected by a midazolam-ketamine infusion technique, in comparison with propofol-ketamine or propofol-alfentanil techniques.<sup>190</sup> Midazolam did not suppress myogenic MEP, even at doses sufficient to produce anesthesia<sup>211</sup>; effects were similar to those with etomidate.<sup>211</sup>

### Neuromuscular Blockade

Neuromuscular blocking drugs (NMBDs) exert little or no effect on the SSEP. They prevent or limit recording of CMAP during myogenic MEP recording because of their effects on the neuromuscular junction. Partial neuromuscular blockade, however, is commonly used during MEP monitoring because it improves conditions for surgery by providing adequate muscle relaxation when retraction of the tissues is required and limits any child movement during the stimulus generation. Partial muscle relaxation may also reduce noise caused by spontaneous muscle movement. It is important that constant neuromuscular blockade is maintained during the procedure. Many centers avoid neuromuscular blockade after intubation, the initial incision, and muscle dissection.

Two methods have been used to assess the degree of neuromuscular blockade for MEP monitoring. One is measurement of the amplitude of the CMAP produced by single supramaximal stimulation (T1) before use of an NMBD. When T1 is maintained between 20% and 50% of the baseline level, reproducible CMAP responses can be obtained with a degree of muscular blockade that allows surgery.<sup>217,222</sup> The other technique is to adjust the neuromuscular blockade based on the train-of-four responses. Comparison of the fourth twitch (T4) with that of first twitch (T1) suggests acceptable CMAP monitoring is possible when two of the four twitches remain (see Fig. 6-26).<sup>222-224</sup> Neuromuscular blockade should be evaluated in the specific muscle groups that are used for electrophysiologic monitoring because different muscle groups have different sensitivities to the NMBDs. Children with preoperative neuromuscular dysfunction tend to demonstrate greater reduction after partial neuromuscular blockade than children with normal preoperative motor function. It is appropriate to avoid neuromuscular blockade in most of these children.<sup>217</sup>

### Choosing the Optimal Combination of Anesthetic Drugs and Techniques

There is no one anesthetic technique suitable for evoked potential monitoring that is applicable to all children. The choice of anesthesia will depend on the child's pathology and the choice

of electrophysiologic monitoring planned during the operation. A marked increase in the use of MEPs and advances in MEP techniques have occurred in recent years. CMAP appear to provide the most useful data for minimizing the risk of spinal cord injury.

The key to success is to use a technique that allows a stable concentration of the “hypnotic” component of anesthesia. There is probably no difference between the inhalational anesthetics (<1 MAC) and propofol (<6 mg/kg/hr). Concentrations of the inhalational anesthetics approaching 1 MAC are now compatible with multiple-pulse MEP monitoring systems that did not appear possible several years ago. Short-acting medications offer greater flexibility should the monitored signals deteriorate. The use of a remifentanyl infusion allows a rapidly titratable “analgesic” component with minimal effect on spinal cord monitoring. Clonidine may be used to decrease the concentration of “hypnotic” drugs during SSEP monitoring and possibly during MEP monitoring. Ketamine may be added as an adjunct to improve MEP monitoring because it better preserves the MEP signals and also allows a reduced dose of other “hypnotic” agents to be used. If a processed electroencephalographic monitor is used to determine anesthetic depth, then the addition of ketamine may confound the processed electroencephalographic monitor reading (by increasing it).<sup>225,226</sup> This occurs despite a deepening in the level of hypnosis.<sup>225</sup> NMBD improves the SSEP monitoring and may be used in conjunction with MEP monitoring within the confines described earlier. However, even in children with idiopathic scoliosis, adequate operating conditions after the initial muscle dissection can be produced in the absence of neuromuscular blockade. In the absence of muscle relaxation, muscle contractions including the masseter muscles will occur during stimulation. In this situation, it would be prudent to insert a bite block to prevent obstructing the tracheal tube or to intubate the child nasally.

## Tourniquet

The tourniquet has been used since Roman times to control bleeding during amputation.<sup>227</sup>

### Indications

The arterial tourniquet is used during orthopedic procedures to reduce blood loss and provide good operating conditions, for intravenous regional blockade and sympathectomy, and for isolated limb perfusion in the management of localized malignancy.<sup>228</sup>

### Design

The word “tourniquet” is derived from the French verb *tourner*, meaning “to turn,” referring to the twisting or screwing action applied to the constricting bandage to tighten it. Von Eschmarck introduced the use of a flat rubber bandage wrapped repeatedly around a limb in 1873.<sup>227</sup> Although this rubber bandage is still used to render a limb bloodless, the pneumatic tourniquet, introduced by Cushing in 1904, has replaced the rubber bandage to maintain ischemia. Compressed gas (nitrogen or air) is used for inflation. The target pressure is preset and compensatory feedback mechanisms maintain that pressure during inflation. Curved and wider tourniquet cuffs, designed to fit conical limbs, are associated with lower arterial occlusion pressures than standard cuffs.<sup>229</sup> A soft dressing applied to the limb before tourni-

quet application helps prevent wrinkles and blisters that may occur when the skin is pinched.<sup>230</sup> Adequate exsanguination can also be achieved by elevation of the arm at 90 degrees or the leg at 45 degrees for 5 minutes.<sup>231,232</sup>

## Physiology

### Ischemia

Ischemia leads to tissue hypoxia and acidosis. The severity and consequences of the associated changes (e.g., increased capillary permeability, coagulation alteration, and cell membrane sodium pump activity) vary depending on the tissue type, duration of ischemia, and collateral circulation. Muscle is more susceptible to ischemic damage than nerves. Histologic changes are more pronounced in muscle beneath the tourniquet compared with muscle distal to the tourniquet.

### Reperfusion

Reperfusion removes toxic metabolites and restores energy supplies. There is a sudden release of lactic acid, creatine phosphokinase, potassium (peak increase 0.32 mEq/L), and carbon dioxide (peak increase 0.8-18 mm Hg) when the cuff is deflated suddenly. Metabolic changes are greater after a longer period of ischemia but return to baseline within 30 minutes. Muscle damage may result in the release of myoglobin that can collect in the collecting tubules of the kidney, precipitating renal failure. Systemic effects after deflation of the tourniquet include a shift of blood volume back into the limb with a transient decrease in blood pressure that is exacerbated by a postischemic reactive hyperemia in the limb. CO<sub>2</sub> release generates a transient increased minute volume. The rapid increase in CO<sub>2</sub> is also associated with a transient (8-10 minute) increase in cerebral blood volume that may affect children with raised intracranial pressure.<sup>228</sup>

Increased microvascular permeability of muscle and nerve tissue occurs with tourniquet release after 2 to 4 hours of ischemia. Interstitial and intracellular edema as well as capillary occlusion secondary to endothelial edema and leukocyte aggregates may take months to resolve.

### Ischemic Conditioning

Short periods of ischemia followed by reperfusion render muscle more resistant to subsequent ischemia. Such ischemic preconditioning improves skeletal muscle force, contractility, and performance and decreases fatigue of skeletal muscle. This preconditioning may enable prolongation of orthopedic and reconstructive procedures.<sup>233</sup>

## Complications

### Local

#### *Muscle Damage*

Histologic changes in the muscle beneath the tourniquet are present after 2 hours of tourniquet time (at 200 mm Hg, 26.7 kPa), but similar changes can occur after 4 hours of tourniquet in the distal ischemic muscle. Direct pressure and mechanical deformation contribute to increased severity of muscle damage under the cuff.<sup>228</sup> These changes include an increase in the number of inflammatory cells in the perifascicular space, focal fiber necrosis, and signs of hyaline degeneration.

The combination of muscle ischemia, edema, and microvascular congestion contributes to "post-tourniquet syndrome":

edema, stiffness, pallor, weakness without paralysis, and subjective numbness of the extremity without objective anesthesia. The common use of postoperative casts may conceal the true incidence of this syndrome. Recovery usually occurs over 7 days.<sup>234</sup>

#### *Nerve Damage*

Direct compression under the cuff rather than ischemia is thought to cause nerve injuries. Sheer forces that are maximal at the upper and lower edges of the tourniquet cause the most damage. These forces are greater with the Esmarch bandage than with the pneumatic tourniquet. The incidence is greater in the upper limb (1/11,000) than in the lower limb (1/250,000), with the radial nerve being the most vulnerable nerve in the upper extremity and the sciatic nerve in the lower extremity.<sup>235</sup>

#### *Vascular Damage*

Arterial injury is uncommon in children. It is an injury of adults with atheromatous vessels, and the tourniquet should be avoided in those patients with absent distal pulses, poor capillary return, a calcified femoropopliteal system, or a history of vascular surgery on the involved limb.<sup>236</sup>

#### *Skin Safety*

Pressure necrosis and friction burns may occur with poorly applied tourniquets, and some form of skin protection should be used routinely.<sup>237</sup> Chemical burns may result from antiseptic skin preparations that seep beneath the tourniquet and are then retained and compressed against the skin.

#### *Tourniquet Pain*

The tourniquet causes a vague dull ache that becomes intolerable after approximately 30 minutes.<sup>238</sup> This pain is associated with an increase in both heart rate and blood pressure that is not ameliorated by general anesthesia and neuraxial blockade.<sup>239</sup> This pain is transmitted by unmyelinated C-fibers. These fibers are normally inhibited by fast pain impulses transmitted by myelinated A-delta fibers, but mechanical compression causes reduced transmission in these larger fibers.<sup>239</sup>

## Systemic

#### *Temperature Regulation*

The combination of decreased heat loss from the ischemic limb and reduced heat transfer from the central to ischemic peripheral compartment increases core body temperature.<sup>240,241</sup> The temperature increase is greater with bilateral tourniquets compared with a unilateral tourniquet.<sup>241</sup> Children requiring intraoperative tourniquets should not be aggressively warmed during surgery.<sup>241</sup> Redistribution of body heat and the efflux of hypothermic venous blood from the ischemic area into the systemic circulation after deflation of the tourniquet decreases the core body temperature, which may switch off thermoregulatory vasodilation and cause a decrease in skin-surface temperature.<sup>242</sup>

#### *Deep Vein Thrombosis and Emboli*

The incidence of emboli after release of the tourniquet in children is uncertain. The tourniquet appears to have no influence on deep venous thromboembolism formation, but release of the tourniquet may be associated with an increased risk of embo-

lism in adults. Some authors have suggested that heparin be used during total joint arthroplasty in adults to prevent emboli formation,<sup>243</sup> although this practice is not routine in children. Some surgeons will use such therapy in adolescents.

#### *Sickle Cell Disease*

Hypoxia, acidosis, and circulatory stasis all contribute to the sickling of sickle cells in susceptible individuals. However, several institutions routinely use tourniquets in children with sickle cell disease while maintaining acid-base status and oxygenation throughout the procedure.<sup>244,245</sup> Each case must be assessed individually for the balance between the advantages of a bloodless field and the risks of precipitating sickling crises (see Chapter 9).

#### *Drug Effects*

Antibiotics given after the tourniquet is inflated will not produce effective blood and tissue concentrations of antibiotics in the ischemic limb. Inflation of the tourniquet should be delayed at least 5 minutes after administration of the antibiotics.<sup>246,247</sup> Medications administered before inflation of the tourniquet may be sequestered in the ischemic limb and then re-released into the systemic circulation when the tourniquet is deflated. The antibiotic effect will depend on the amount of antibiotic sequestered, the tissue binding, and the concentration-response relationship for the antibiotic, although the impact is minimal for most medications used in anesthesia. Volume of distribution may be reduced if the drug is administered after tourniquet inflation, but the plasma clearance remains unaffected.

#### **Recommended Cuff Pressures**

Tourniquets should generally remain inflated less than 2 hours, with most authors suggesting a maximal time of 1.5 to 2 hours. Techniques such as hourly release of the tourniquet for 10 minutes, cooling of the affected limb, and alternating dual cuffs may reduce the risk of injury.<sup>248</sup> Both nerve and muscle injuries that occur beneath the tourniquet cuff are related to the pneumatic pressure. Consequently, the lowest possible pressure that maintains ischemic conditions should be sought. Hypotensive anesthetic techniques have been used in adults to reduce the need for high cuff inflation pressures,<sup>249</sup> but there seems to be little need for this in children. One author has suggested that pediatric occlusion pressures should be measured by Doppler imaging and the tourniquet pressure set at 50 mm Hg above this value. The maximum mean pressures recommended for the upper and lower extremities are  $173.4 \pm 11.6$  mm Hg (range: 155-190 mm Hg) and  $176.7 \pm 28.7$  mm Hg (range: 140-250 mm Hg), respectively.<sup>250</sup> Wider cuffs exert less force per unit area and reduce the risk of local sequelae. Recommendations for adults suggest that the cuff should exceed the circumference of the extremity by 7 to 15 cm. This is difficult to achieve in infants in whom the proximal limb length is proportionally shorter than adults and wide cuffs would impinge on the surgical field.

## **Acute Bone and Joint Infections**

The mainstays of management for osteomyelitis and septic arthritis are antibiotics and surgical drainage. The incidence of these infections is increasing, particularly in immunocompromised children with human immunodeficiency virus (HIV) infection. Tuberculosis remains a scourge in many developing

countries. Mortality rates for both hospital-acquired staphylococcal disease in compromised children<sup>251</sup> and community-acquired disease in healthy children<sup>252,253</sup> range from 8% to 47% in those presenting with severe sepsis.<sup>254</sup> *Mycobacterium* and *Staphylococcus* organisms resistant to conventional antibiotics increase morbidity and mortality.

#### **Pathophysiology**

*Staphylococcus aureus* is the most common pathogen. Osteomyelitis develops after a bacteremia mostly in prepubertal children. Normal bone is highly resistant to infection, but *S. aureus* adheres to bone by expressing receptors for components of bone matrix, and the expression of collagen-binding adhesin permits the attachment to cartilage.<sup>255</sup> Once the microorganisms adhere to bone they express phenotypic resistance to antimicrobial treatment.<sup>255</sup>

The metaphyseal region around the growth plate is the predominant area of infection. Sluggish blood flow in the metaphysis predisposes to bacterial infection, and endothelial gaps in developing vessels allow bacteria to escape into the metaphysis. Subsequent abscesses may decompress into the joint or subperiosteally. Infection may involve adjacent tissue planes, and hematogenous spread causes multiple pathologic processes beyond the primary site of infection.

Septic arthritis is more common in neonates because transphyseal vessels link the metaphysis and epiphysis. Growth plate and epiphyseal destruction may both occur in this age group. Articular cartilage damage is attributable to the release of proteolytic enzymes from both the pathogen and activated neutrophils.

#### **Clinical Presentation**

The majority of children with staphylococcal disease present with musculoskeletal symptoms and fever, but those with disseminated disease can present critically ill (4%-10%) with severe sepsis and lung disease.<sup>252,253</sup> There is often a history of trauma.<sup>252,253</sup> It can be difficult to diagnose extracutaneous foci. One study<sup>251</sup> reported that 50% of extracutaneous foci of staphylococcal infection were not detected on hospital admission, and one third of these lesions were noted for the first time at autopsy. An absolute polymorphonuclear cell count of greater than  $10,000/\text{mm}^3$  or an absolute bandform count of greater than  $500/\text{mm}^3$ , or both, correlates with the presence of one or more inadequately treated sites of staphylococcal infection.<sup>251</sup> Tuberculosis is the great mimic and must always be suspected in endemic areas.

Diagnosis is confirmed by blood, bone, or joint aspirate culture. Radiologic procedures (plain radiographs, computed tomography, magnetic resonance imaging, radionuclide scans) are often required to identify foci, and the anesthesiologist is often requested to provide sedation/analgesia.

#### **Treatment Options**

Antibiotic therapy is the mainstay of treatment. Initial antibiotic choice is dictated by age, local pathogen, and sensitivity profiles. Antibiotic treatment should be extended to cover gram-negative enterococci in neonates and streptococci in older children. *Haemophilus influenzae* remains a pathogen in unvaccinated regions. Surgical decompression of acute osteomyelitis that is responding poorly to antimicrobial therapy may release intramedullary or subperiosteal pus and lead to clinical improvement. Pus within fascial planes also requires release. Venous

thrombosis attributable to pus in soft tissue planes around major joints was associated with a high mortality in one series.<sup>252</sup> Determining and eradicating the primary focus improves both mortality and recurrence rates.<sup>256</sup> An aggressive search for foci and surgical drainage of infective foci is required.

Highly active antiretroviral therapy (HAART) has positively altered the mortality rates in HIV-infected children. However, acute bone and joint infections still occur<sup>257</sup> and these drugs have the potential to cause significant morbidity secondary to changes in fat distribution, lipid profiles, glucose, homeostasis, and bone turnover.<sup>258</sup> Infarction may replace infection as the major cause of morbidity and mortality from HIV.<sup>258</sup> It is uncertain that HAART should be continued during acute osteomyelitis. Worsening cell-mediated immune function may occur during tuberculosis treatment if HAART is continued.<sup>259</sup> The combination of HIV infection and tuberculosis is potentially lethal in children, and antituberculous treatment is continued for 12 to 18 months.

### Anesthesia Considerations

Anesthesiology services are commonly required for sedation during diagnostic investigation, anesthesia for surgical exploration and release of pus or fixation of pathologic fractures, management of pulmonary complications (intercostal chest drain insertion, pleurodesis), central venous cannulation for long-term antibiotic treatment and analgesic modalities.

Children with disseminated staphylococcal disease may be critically ill with multisystem disease and require fluid volume augmentation, inotrope support, positive-pressure ventilation, extracorporeal renal support, and coagulation factor replacement. Others may appear clinically stable before anesthetic induction; the assessment of hypovolemia in children is subject to moderate to poor inter-rater agreement.<sup>260</sup> Intravenous access and rehydration are required before beginning anesthesia to avoid a precipitous blood pressure drop immediately after induction. Bacteremic showering during manipulation and drainage of pus causes further decompensation. Excessive bleeding due to altered coagulation status should also be anticipated.

The presence of a septic arthritis in the shoulder or neck may cause cervical ligamentous laxity predisposing to C1/C2 subluxation during intubation.<sup>261</sup> Pneumatoceles from staphylococcal pneumonia can rupture during positive-pressure ventilation. A spontaneous breathing mode, however, may be difficult to achieve because of laryngospasm, breath holding, increased secretions, and bronchospasm. The use of NMBDs and positive-pressure ventilation in these children with a low threshold to introduce inotropes to support the cardiovascular system is an easier option. Vigilance is required for the presence of an acute pneumothorax.

Myocarditis, pericarditis, and pericardial effusions compromise myocardial function; one author reported a 12% prevalence of infective endocarditis in children with hospital-acquired *S. aureus* bacteremia. This prevalence of infective endocarditis was frequently associated with congenital heart disease and multiple blood cultures.<sup>262</sup> The incidence of infective endocarditis among those children with community-acquired disease without preexisting cardiac abnormalities was low,<sup>252</sup> suggesting that echocardiography could be reserved for children with preexisting cardiac disease, suspicious clinical findings, those whose temperature fails to settle, or those who have prolonged bacteremia without an obvious source of infection.

### Pain Management

Morphine and acetaminophen are the analgesics commonly used for postoperative pain management. The use of tramadol in children is increasing as our understanding of the pharmacokinetics of this medication increases.<sup>263,264</sup> The low incidence of respiratory depression and constipation, fewer controls on use, and similar frequency of nausea and vomiting (10%-40%) compared with opioids make tramadol an attractive alternative.<sup>265-267</sup> NSAIDs are relatively contraindicated in the presence of coagulation disorders, altered renal function, and cyclooxygenase-2-mediated inhibition of osteogenesis.

The performance of regional blockade in children with acute bone or joint infection is controversial. There are no studies addressing the risk/benefit ratios of regional techniques in this population. It would seem reasonable to use these techniques only after 24 hours of appropriate antibiotic therapy in apyrexial children who show no signs of a coagulopathy.

## Common Syndromes

Children with some specific conditions present repeatedly for orthopedic procedures. It is worthwhile maintaining a database for these children detailing their anesthetic management. In addition, there should be 24-hour access to standard texts concerning anesthesia and uncommon pediatric diseases.

### Cerebral Palsy

#### Clinical Features

*Cerebral palsy* is an umbrella term that describes a group of nonprogressive, but often changing, motor impairment syndromes secondary to lesions or anomalies in the brain that occur during the early stages of its development.<sup>268</sup> It is the leading cause of motor disability during childhood, with a prevalence of approximately 2 per 1000 live births in developed countries.<sup>269</sup> Disorders include cognitive impairment, sensory loss (vision and hearing), seizures, and communication and behavioral disturbances. Systemic disorders resulting from cerebral palsy affect the gastrointestinal, respiratory, urinary tract and orthopedic systems. Cerebral palsy is divided into three broad categories: spastic (70%), dyskinetic (10%), and ataxic (10%) (see Chapter 22).<sup>268</sup> Those children suffering from spastic cerebral palsy commonly present for orthopedic procedures because of the development of contractures at major peripheral joints.<sup>270,271</sup> Functional improvement after surgery in children with spastic diplegia and spastic hemiplegia is better than for those suffering spastic quadriplegia.<sup>270</sup>

#### Orthopedic Considerations

Orthopedic manipulations form only part of treatments designed to either improve performance or improve the ease of care. Management includes orthopedic surgery, physical and occupational therapy, recreational therapy, orthotics, and assistive devices improve functional outcomes. Medical modalities such as intramuscular injections of botulinum toxin, and intrathecal administration of baclofen via an implanted pump may also be of benefit.<sup>272</sup> Selective dorsal rhizotomy has also been used to control spasticity.<sup>273,274</sup> The indications and timing of surgical interventions vary. Gait analysis increases the age of the first orthopedic surgical procedure, and treatment with botulinum toxin type A delays and reduces the frequency of surgical procedures on the lower extremities.<sup>275,276</sup> Bone and soft tissue

surgical procedures are designed to lengthen or weaken spastic muscles to give opposing muscles a chance to attain muscle balance.

### Anesthetic Considerations

Children presenting for orthopedic surgery often have previous experience of operating rooms. These children should be handled with sensitivity because communication disorders and sensory deficits may mask mild or normal intellect. They may be accompanied by their parent or a caregiver and/or be premedicated before induction of anesthesia. If there is a communication problem, then the parent or caregiver should be present before and after anesthesia.<sup>270,277</sup> Medical conditions (seizure control, respiratory function, gastroesophageal reflux) should be optimized preoperatively. Contracture deformities, spinal deformities, decubitus ulceration, and skin infection must be considered when positioning the child for anesthesia and surgery. Poor nutritional status affects postoperative wound healing and the risk of infection. Concurrent medications may influence the anesthetic considerations: cisapride for gastroesophageal reflux is associated with prolonged QT interval, sodium valproate can cause platelet dysfunction and affect drug metabolism, and anti-convulsant use increases resistance to NMBDs.<sup>278</sup> A history of latex allergy should be sought because of exposure to latex allergens from an early age.<sup>279</sup>

Intravenous access may be difficult. Drooling, a decreased ability to swallow secretions, and gastroesophageal reflux may dissuade some against an inhalational induction, but there is no evidence that rapid sequence induction is safer. Succinylcholine use is not associated with hyperkalemia because the muscles of these patients have never been denervated. Noncommunicative/nonverbal children with cerebral palsy require less propofol to obtain the same bispectral index values (i.e., 35-45) than do otherwise healthy children.<sup>280</sup> The MAC of halothane is 20% less in children suffering from cerebral palsy, whether or not they take anticonvulsant drugs (MAC, 0.62 and 0.71, respectively).<sup>281</sup> Intraoperative hypothermia is common in those children with disordered temperature regulation secondary to hypothalamic dysfunction, reduced muscle bulk, and fat deposits. Thermal homeostasis should be managed aggressively from the moment the child enters the operating room. Extensive plaster casting is an important component of bone and soft tissue surgical procedures. These casts may conceal blood loss, and limb swelling within the cast may contribute to compartment syndromes. Plaster jackets and hip spicas have been associated with mesenteric occlusion and acute gastric dilatation. Pain and spasm are regular features postoperatively. Epidural analgesia is particularly valuable when major orthopedic procedures are performed.<sup>270</sup> Occasionally, two epidurals at different spinal sites may be required for multilevel surgery. Systemic benzodiazepines, baclofen, dantrolene, and clonidine have been used to reduce muscle spasms. Selective dorsal rhizotomy is associated with severe pain, muscle spasms, and dysesthesia. Epidural and intrathecal morphine have been used to control this pain. Intravenous morphine and midazolam has also been successfully used.<sup>282</sup> Oral benzodiazepines may be required to reduce the incidence and severity of muscle spasms but should be used with caution if combined with opioid analgesia.

Pain assessment is difficult in these children, but there are a number of scoring systems available (see Chapter 44).<sup>283,284</sup> The opinions of parents and caregivers are extremely valuable in the

assessment of pain and discrimination from other factors such as irritability on anesthetic emergence, poor positioning, a full bladder, or nausea.

### Spina Bifida

Spina bifida is characterized by developmental abnormalities of the vertebrae and spinal cord that may be associated with changes in the cerebrum, brainstem, and peripheral nerves. The failure of fusion of the vertebral arches is commonly known as spina bifida. *Spina bifida occulta* refers to spina bifida that occurs when skin and soft tissues cover the defect. *Spina bifida aperta* is used to describe those lesions where the defect communicates with the outside as either a meningocele or a myelomeningocele (incidence 1:1,000 live births) (see also Chapter 23). The myelomeningocele sac contains nerve roots that do not function below the level of the lesion.

### Clinical Features

Nerve root dysfunction results in muscle paralysis as well as a neurogenic bowel and bladder. The majority (80%) of children develop hydrocephalus consequent to aqueductal stenosis (Arnold-Chiari type II malformation). Skeletal abnormalities such as clubfoot and congenital dislocation of the hip are common. Scoliosis may be due to either congenital vertebral abnormalities or, more commonly, abnormal neuromuscular control. Epilepsy and learning disorders can also occur, but most children have normal intelligence.

### Orthopedic Considerations

Denervation causes muscle imbalance that results in abnormalities at the hip, knee, and foot. The aims of surgery are to reduce flexor posture at the hip and knee and plantigrade feet. Clubfeet, hip subluxation, and scoliosis are common presentations for orthopedic correction.

### Anesthetic Considerations

The potential for infection of the central nervous system dictates early closure of the sac within the first few days of life. Subsequent surgical procedures and urinary catheterizations set the stage for sensitivity to latex.<sup>285</sup> Primary prophylaxis (avoiding all latex materials and a latex-free operating room) is recommended for prevention of latex allergy and anaphylaxis.<sup>286</sup>

Preoperative assessment should include motor and sensory deficits, respiratory and renal function, and functioning of the ventriculoperitoneal shunt, if present. Positioning on the operating table may require additional pillows for support of limbs with contractures. As a result of hypoesthesia in the lower extremities, intravenous cannulae can be inserted painlessly. However, venous access is usually poor in the lower extremities because of their limited use. The risk of endobronchial intubation is increased because of a short trachea (36%).<sup>287</sup> Kyphoscoliosis may distort tracheal anatomy. Renal dysfunction may dictate choice of NMBD and avoidance of NSAIDs. Succinylcholine does not cause hyperkalemia.<sup>288</sup> A reduced hypercapnic ventilation response means that these children should be closely observed in the recovery period.

### Osteogenesis Imperfecta

Osteogenesis imperfecta was believed to have afflicted Ivar the Boneless (Ivar Ragnarsson), a Viking chieftain who led a successful invasion of the East Anglia region of England in AD 865. He is reported to have had legs as soft as cartilage so that he

was unable to walk and had to be carried on a shield. Ivar's name is also associated with an early form of thoracoplasty. When the King of Northumbria, Aelle, was captured a few years later, he was subjected to the horrific "blood eagle" ordeal by the Vikings. His ribs were torn out and folded back to form the shape of an eagle's wings and his lungs were removed.

### Clinical Features

Osteogenesis imperfecta (OI) is a genetically determined disorder of connective tissue that is characterized by bone fragility. The disease state encompasses a phenotypically and genotypically heterogeneous group of inherited disorders that result from mutations in the genes that code for type I collagen.<sup>289</sup> The disorder is manifest in tissues in which the principal matrix protein is type I collagen (mainly bone, dentin, sclerae, and ligaments). Musculoskeletal manifestations are variable in severity along a continuum ranging from perinatal lethal forms with crumpled bones to moderate forms with deformity and propensity for fracture to clinically silent forms with subtle osteopenia and no deformity.<sup>289</sup> Classification (types I-IV) is based on the timing of fractures or on multiple clinical, genetic, and radiologic features. Type I is the most common (1:30,000 live births) and, together with type IV, has autosomal dominant inheritance patterns. These children have the classic triad of blue sclerae, multiple fractures, and conductive hearing loss in adolescence. Bowing of the lower limbs, genu valgum, flat feet, and scoliosis develop with age. Type IV is characterized by osteoporosis, leading to bone fragility without many of the other features of type I. Types II and III are more severe forms of OI and have autosomal recessive inheritance patterns. Molecular genetic studies have identified more than 150 mutations of the *COL1A1* and *COL1A2* genes, which encode for type I procollagen.<sup>289</sup>

### Orthopedic Considerations

The goals of treatment of OI are to maximize function, minimize deformity and disability, maintain comfort, achieve relative independence in activities of daily living, and enhance social integration. Physiotherapy, rehabilitation, and orthopedic surgery are the mainstay of treatment in moderate to severe forms of OI. Medical treatment with the antiresorptive bisphosphonates (e.g., pamidronate) can decrease pain, lower fracture incidence, and improve mobility. Initial investigations have demonstrated an acceptable safety profile for pamidronate. Currently, there is a lack of long-term follow-up data, which will be necessary for the development of responsible guidelines for therapy.<sup>290</sup> Medical therapies other than bisphosphonates, such as growth hormone and parathyroid hormone, currently contribute only a minor role. Gene-based therapy currently remains in the early stages of preclinical research.<sup>291,292</sup>

Operative intervention is indicated for recurrent fractures or deformity that impairs function.<sup>289</sup> Fractures in mild to moderately severe cases of OI type I are treated using the same methods as for children without OI. Realignment of deformed bones that are fracturing frequently followed by external or internal support is commonly performed. It is important in selecting various modes of treatment to consider the natural history of the particular type of OI and to set realistic goals.<sup>293-297</sup>

### Anesthetic Considerations

In common with other children suffering chronic disability, these children are veterans to the operating room. Chronic pain from frequent fractures complicates handling. Deafness may

hinder communication. Preoperative assessment centers on the chest wall deformity because this determines the severity of restrictive lung disease and subsequent cardiovascular compromise. Neck mobility, mouth opening, and dentition should also be assessed.

There is a risk of further fractures with positioning, tourniquet application, airway handling, and even use of a blood pressure cuff. Invasive pressure monitoring may be less traumatic than a blood pressure cuff for some children. If noninvasive blood pressure monitoring is used, less frequent monitoring of the blood pressure is recommended if possible. A laryngeal mask airway may avoid pressure from face masks. An individual history may help determine the risk-benefit ratio for each child. Succinylcholine has the theoretical potential to cause fasciculation-induced fractures.

Abnormal temperature homeostasis may result in intraoperative hyperthermia that may be severe and accompanied by tachycardia and metabolic acidosis. This response differs from that of malignant hyperthermia in that there is an absence of respiratory acidosis and muscle rigidity.<sup>298</sup> Surface cooling is usually effective in restoring thermal homeostasis.

## Duchenne Muscular Dystrophy

### Clinical Features

Duchenne muscular dystrophy is the most common of the progressive muscular dystrophies. It is an X-linked recessive disorder with an incidence of 3:10,000 births (see Chapter 22). The DMD gene (*Xp21*) codes for a large sarcolemmal membrane protein (dystrophin)-associated muscle cell membrane integrity. Dystrophin is missing or nonfunctional in DMD children. Children usually present before school age with a waddling gait and go on to develop a lumbar lordosis and difficulty climbing stairs. Children use their arms to assist standing up, owing to proximal weakness of the hip girdle. Distal muscles, such as the calves, appear hypertrophied. The disease process is progressive with increasing muscle weakness with age: often these children become wheelchair bound by 10 to 11 years of age. Respiratory weakness, often exacerbated by scoliosis and by difficulty swallowing secretions due to pharyngeal involvement, can progress to a terminal pneumonia in late teenage years.<sup>299</sup>

Most clinicians regard DMD as a static disease, but this could not be farther from the truth. In early childhood, skeletal muscle is constantly catabolized and unstable. The use of membrane destabilizing medications such as succinylcholine and potent inhalational anesthetics (halothane in particular) in these young children can result in hyperkalemia, rhabdomyolysis, and cardiac arrest (see later). However, once the children reach adolescence, the bulk of the skeletal muscle catabolism has arrested and membrane-destabilizing medications are left with no substrate. Indeed, at The Hospital for Sick Children in Toronto, both succinylcholine and potent inhalational anesthetics were used in adolescents with DMD who were undergoing scoliosis surgery and instrumentation without sequelae. In contrast, cardiac and smooth muscle consequences of DMD are minor and insignificant in childhood but may become life-threatening during adolescence and adulthood (see later). It is imperative to appreciate the developmental nature of DMD and to recognize the risks that may be associated with the use of succinylcholine and inhalational anesthetics in this group of children.

DMD can affect cardiac and smooth muscle as well as skeletal muscle. Right ventricular function may be compromised by nocturnal oxygen desaturation and sleep apnea contributing to pulmonary hypertension. Sinus tachycardia and arrhythmias may occur at an early age, but clinically apparent cardiomyopathy usually does not develop before 10 years of age. One third of children have a degree of intellectual impairment.

Clinical suspicion of DMD should arise in male preschool-aged children with delayed walking ability and serum creatine phosphokinase concentration measured as a screening tool. Corticosteroids are increasingly used for the management of DMD; it is believed that they increase muscle mass by decreasing protein breakdown.<sup>300</sup>

Becker muscular dystrophy (BMD) is a milder form of DMD with onset around puberty or later in the teenage years. Clinical expression is variable, but even adolescents presenting with mild or subclinical weakness can develop a cardiomyopathy with age progression. Death secondary to cardiac or respiratory failure does not usually occur until the fourth or fifth decade. Improvements in respiratory care have resulted in dilated cardiomyopathy as being the major cause of death.<sup>301</sup> This autosomal recessive myopathy is also due to mutations of dystrophin with a deletion of exons 11 to 13 in the *Xp21* gene.<sup>302</sup> Dystrophin exerts its effect at the voltage-gated chloride channel (CLCN1). Genetic analysis is an essential step in confirming the diagnosis. Additional electromyographic procedures may be of diagnostic value even when muscle biopsy may reveal no evidence of dystrophy.

Two thirds of children with mild or subclinical BMD have evidence of right ventricular dilatation and one third have evidence of LV dysfunction.<sup>303</sup> A thorough cardiac evaluation (similar to DMD) is recommended before scoliosis surgery.<sup>301</sup> Hyperthermia plus heart failure, mimicking malignant hyperthermia and hyperkalemia with rhabdomyolysis after inhalational agents, have both been reported with BMD.<sup>304,305</sup> Despite these reports, the relationship between BMD and malignant hyperthermia remains unclear.

### Orthopedic Considerations

Orthopedic surgery is indicated to improve or maintain ambulation and standing. Early treatment of contractures of the hips and the lower limbs prevents severe contractures and delays the progression of scoliosis.<sup>306</sup> Techniques designed to improve deformities and permit early postoperative mobilization include subcutaneous release of contracted tendons and percutaneous removal of cancellous bone with corrective manipulation of the feet. Maintenance of the upright posture extends the ability of these children to attend to their tasks of daily living.<sup>307</sup> Spinal deformities attributable to muscle imbalance or a collapsing spine are corrected to improve or maintain sitting posture. Spinal fusion may also decrease the rate of deterioration of respiratory function, although this has been questioned.<sup>308</sup>

### Anesthetic Considerations

Respiratory and cardiovascular compromise dominates preoperative assessment. Deformities and contractures of limb joints hinder vascular access, regional anesthetic techniques, and positioning on the operating table. Hypertrophy of the tongue may cause difficulty during intubation. Gastric motility is delayed with prolonged gastric emptying times.<sup>309</sup> Tracheobronchial tree compression has been described in a child positioned prone for spinal instrumentation.<sup>310</sup> These children have a tendency to

greater blood loss during surgery. The precise etiology remains unclear but may be because fat and connective tissue have replaced muscle or because of abnormalities in the blood vessels.<sup>135</sup>

Nondepolarizing NMBDs have a slow onset of action and prolonged duration of action.<sup>311-313</sup> All NMBDs should be monitored with a peripheral nerve stimulator.<sup>314</sup> *Succinylcholine is contraindicated in these children because of the risk of hyperkalemia, muscle rigidity, rhabdomyolysis, myoglobinuria, arrhythmias, and cardiac arrest.* There is no clear link between DMD and malignant hyperthermia.<sup>315,316</sup> The predominant candidate gene for malignant hyperthermia is located on the long arm of chromosome 19, whereas the gene for DMD is located on the short arm of the X chromosome.<sup>317</sup> Although volatile anesthetic agents continue to be used in young children with DMD, rhabdomyolysis and hyperkalemia have been reported in the recovery room after halothane, isoflurane, desflurane, and sevoflurane anesthesia.<sup>318-322</sup> *Potent inhalational anesthetics should be avoided in young children with DMD and supplanted with alternative anesthetics that do not trigger rhabdomyolysis and hyperkalemia.*<sup>323</sup>

Regional techniques such as epidurals may be technically more difficult due to kyphoscoliosis and obesity. The use of ultrasound-guided peripheral nerve blockade can improve the quality and reduce complications of neuronal blockade.<sup>6</sup> Opioids are not contraindicated in the postoperative period but should be used with caution in children with respiratory compromise. Tramadol is an effective alternative. Noninvasive ventilation support using bilateral or continuous airway pressure is sometimes required after major surgery or in those already receiving this treatment overnight.

## Arthrogryposis Multiplex Congenita

### Clinical Features

Arthrogryposis multiplex congenita is a syndrome of multiple persistent limb contractures often accompanied by associated anomalies, including cleft palate, genitourinary defects, gastroschisis, and cardiac defects.<sup>324</sup> The incidence is 1 in 3000 births. Joint contractures are present at birth and are a result of immobility in utero, commonly related to a neurogenic abnormality or myopathy.<sup>325</sup> These children have been likened to a "thin, wooden doll" because muscles connected with affected joints are atrophic and replaced by fibrous tissue and fat.<sup>326</sup> The temporomandibular joint may also be involved, causing restricted jaw opening and micrognathia. Scoliosis commonly develops. Restrictive lung disease, rib cage deformities, and pulmonary hypoplasia predispose to recurrent chest infections.

### Orthopedic Considerations

The aims of surgery are to improve function. The majority of surgery involves the soft tissues, tendons, and osteotomies of the lower limbs and hips.<sup>327</sup> Upper limb surgery is less common. The extension contracture of the elbow joint makes it impossible to reach the mouth or to perform hygienic necessities. Improvement in passive elbow flexion by capsulotomy or in active flexion by triceps transfer can increase independence and personal hygiene. When both arms are involved, consideration may be given to have one arm in flexion for reaching the head and mouth passively or even actively and one arm in extension for basic hygiene cares.<sup>328</sup>

### Anesthetic Considerations

Arthrogryposis multiplex congenita is commonly associated with other syndromes that may complicate anesthesia.<sup>324,329</sup> Venous cannulation is difficult because veins tend to be small and fragile. The concavity of joints is difficult to access. Care must be taken with positioning on the operating table and protecting skin overlying bony joints to prevent pathologic fractures. These children should be evaluated for a difficult airway because of temporomandibular joint limitation and micrognathia.<sup>330-332</sup> Fusion or underdevelopment of the first and second cervical vertebrae may further complicate laryngoscopy and tracheal intubation. Tracheal intubation may become progressively more difficult with age. During infancy, however, evaluating mouth opening may be difficult; it may be necessary to insert a tongue blade into the mouth to determine whether the mandible

can be distracted from the maxilla. Succinylcholine has been used without incident in these children, although teleologically the use of a depolarizing muscle relaxant in the presence of anterior horn cell disease is contentious. The response to non-depolarizing NMBDs should be monitored.

Hyperthermia and persistent tachycardia have both been reported during general anesthesia.<sup>324,333-335</sup> These signs occur irrespective of the anesthetic agent and are not associated with malignant hyperthermia. In this case, hyperthermia responds to simple cooling techniques.

Pulmonary dysfunction and an increased sensitivity to opioids dictate suitable monitoring postoperatively. Regional techniques may be difficult in the presence of contractures but, if successful, offer both intraoperative and postoperative analgesia.<sup>326</sup> Success can be improved using ultrasound-guided techniques.

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## References

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