Cerebral palsy (CP) is a group of nonprogressive, motor impairment patterns due to an insult to the developing encephalon. Clinical manifestations vary by the specific motor deformity, anatomically affected region, and location of the brain injury. Spasticity is common, resulting in skeletal muscle weakness and loss of fine motor control. Spasticity in a child undergoing skeletal maturation may precipitate joint contractures and dislocation. Long-term medical care is interventional. The therapeutic goals are to increase the person’s independence and improve the caretaker’s ability to provide daily care. Early medical intervention to control spasticity and prevent contractures may reduce the need for future orthopedic surgical intervention. CENTrally acting, tone-reducing medications may decrease spasticity but cause central nervous system side effects. Orthopedic surgical procedures may be necessary to remedy the chronic effects of increased tone on the muscles and bones of the extremities and spine. Anesthetic care of children and adolescents with CP is increasing. Thorough preoperative assessment facilitates preparation of an intraoperative care plan. Intellectual disability may attend CP and limit the person’s ability to participate in preoperative preparation. Perioperative complications include hypothermia, intravascular depletion, muscle spasm, limb contracture, and seizure control. Gastroesophageal reflux and poor respiratory function might complicate anesthetic management.

Key words: Baclofen, cerebral palsy, dorsal rhizotomy, general anesthesia, static encephalopathy.

Objectives
At the completion of this course the reader should be able to:

1. Detail the epidemiology and accompanying pathophysiology that accompanies cerebral palsy.
2. List the classifications and clinical manifestations of cerebral palsy.
3. Describe the potential side effects of oral anti-spasticity drugs, and list the advantages of intrathecal baclofen administration for patients with cerebral palsy.
4. Prepare an intraoperative anesthetic care plan for a patient with cerebral palsy.
5. Describe the pharmacodynamics of skeletal muscle relaxant and sedative-hypnotic administration in patients with cerebral palsy.

Introduction
Cerebral palsy (CP) is a descriptive clinical term referring to a symptom complex characterized by nonprogressive motor impairment that develops from an antenatal, perinatal, or postnatal insult to the developing encephalon. Accordingly, CP is not a diagnosis. Although CP is the most common cause of severe childhood disability, the inciting event is typically unknown. The clinical impact of the brain insult is diverse and may range from mild monoplegia to quadriplegia and normal intellect to mental retardation. Most people with CP survive to adulthood, yet mortality is greater in people with neurological dysfunction that impairs mobility and feeding. The United Cerebral Palsy Foundation estimated that more than 760,000 children and adults were identified as having CP in 2001 and as many as 8,000 infants and 1,200 to 1,500 preschool-aged children are newly diagnosed each year in the United States. The psychological, social, and medical implications of CP are staggering. The estimated US health-related expenses in 2002 were $8.2 billion.3

The medical care of disabled people has traditionally been supportive rather than interventional. Changes in societal attitude regarding the care of disabled people and advances in medical and surgical...
management have transformed care to an interventional approach. Contemporary care of people with CP is multidisciplinary, involving pediatricians, family practitioners, neurologists, orthopedists, physiatrists, neurosurgeons, anesthesiologists, general surgeons, and physical, occupational, and speech therapists. This Journal course will review the cause, epidemiology, classification, and medical, anesthetic, and orthopedic surgical management of people with CP.

Epidemiology and pathophysiology
In the 1860s, the English surgeon William John Little described muscle spasticity affecting the lower extremities in children during the first year of life. Little's original term was cerebral paresis. He observed that the majority of children with paresis were born prematurely or following complicated deliveries. Many interpreted his description to suggest the cause of cerebral paresis was birth asphyxia. Not only was this declaration scientifically unchallenged, it has been unequivocally accepted by physicians, the general public, and the legal profession for the past 100 years. If birth asphyxia is the genesis of CP, it follows that interventional obstetrical and neonatal care should reduce the overall incidence of CP. Despite reductions in the neonatal death rate, parallel decreases in the prevalence of CP have not been observed.

The central nervous system is one of the first organ systems to develop during embryogenesis, with continued maturation postnatally until the age of 2 years. Injury during this developmental period may produce a permanent, nonprogressive lesion of the cerebral motor cortex. This insult to the motor cortex results in spasticity and hypertonicity. Additional coexisting neurological disabilities include developmental delay, cognitive impairment, visual and auditory impairment, sensory impairment of the upper extremities, attention deficit, epilepsy, and autonomic dysfunction. Chronic organ system dysfunction also affects the gastrointestinal, respiratory, and musculoskeletal systems.

In the 1940s, an international group of American and British physicians and scientists formed the American Academy for Cerebral Palsy and Developmental Medicine to research and examine the treatment for CP. Yet, a consensus definition of CP was not adopted until 1964. A contemporary revised definition and classification of CP was published in 2005 following a review of pertinent scientific material at an international symposium:

Cerebral palsy (CP) describes a group of disorders of the development of movement and posture, causing activity limitation, that are attributed to non-progressive disturbances that occurred in the developing fetal or infant brain. The motor disorders of cerebral palsy are often accompanied by disturbances of sensation, cognition, communication, perception, and/or behavior, and/or by a seizure disorder.

Cerebral palsy registries from Western Australia, the United Kingdom, Sweden, and the United States report the incidence of CP to be between 2.0 and 2.5 per 1,000 live births. The literature has demonstrated a consistent relationship between an abbreviated gestation and low birth weight with the subsequent development of CP. Infants weighing less than 1,500 g have a 30 times greater risk for the development of CP than full-term infants weighing more than 2,500 g. There is an increased incidence of CP in very preterm (<33 weeks' gestation) and preterm infants (<37 weeks' gestation) with increasing severity of disability. The increasing incidence of CP may reflect improved neonatal intensive care management and accompanying survival rates, improvement in diagnosis and identification, and more accurate documentation via national registries.

Despite the historical focus on the perinatal period as the genesis of CP, contemporary literature suggests that perinatal asphyxia is responsible for only 6% to 10% of new CP cases. The etiological event producing the cerebral insult is thought to occur perinatally in as many as 75% of cases, whereas postnatal insults account for 10% to 18% of cases. The timing and proposed causes of CP are listed in the Figure.

**Classification and clinical manifestations of cerebral palsy**
Classification of CP is based on the specific motor deformity, the anatomically affected region, and the location of the brain injury. As illustrated in the Figure, the injury to the developing nervous system may occur during the antenatal, perinatal, or postnatal period, producing subtle motor impairment or profound motor dysfunction. Spastic CP (70% of cases) develops following injury to the cerebral motor cortex. The clinical manifestations include spastic diplegia (spasticity affecting the lower extremities, minimal upper extremity involvement), spastic hemiplegia (spasticity of the ipsilateral extremities), and spastic quadriplegia (spasticity of all extremities).

Spastic diplegia follows a ventricular hemorrhage within the periventricular white matter, producing periventricular leukomalacia. A patient with spastic diplegia has normal intellect. Spastic hemiplegia involves a single hemispheric insult occurring more frequently in term infants. Patients with spastic diplegia and spastic hemiplegia have the best prognosis for functional improvement. In people with spastic hemiplegia, the upper extremity is affected more than the lower extremity. On the other hand, spastic quadriplegia involves multiple central nervous system insults with accompanying severe neurological dysfunction and disability. Epilepsy is a common comorbidity in spastic quadriplegia.
Dyskinetic CP develops following injury to the basal ganglia. Dystonia (involuntary muscle contraction), chorea (irregular, rapid, and involuntary movements of the extremities), and athetosis (involuntary writhing movements of the upper extremities) are characteristic of basal ganglia injury. People with dyskinetic CP have impaired speech and drooling. Epilepsy occurs in approximately 25% of people with dyskinetic CP. Dyskinesia is commonly exhibited in CP.

Ataxic CP follows injury of the cerebellum, producing tremor, loss of balance, and difficulty with speech. Spasticity and dyskinesia may be exhibited when injury has occurred to the cerebral motor cortex and the cerebellum. People with ataxic CP have a poor prognosis for functional improvement.

The pyramidal motor system controls all voluntary movements. Insults within the pyramidal motor system precipitate neurological disability. The pyramidal system is a 2-neuron system consisting of upper motor neurons in the primary motor cortex and lower motor neurons in the anterior horn of the spinal cord. Injury to upper motor neurons decreases cortical input from the reticulospinal and corticospinal tracts, inhibiting operative motor units and resulting in abnormal muscle control and weakness. In addition, the loss of descending inhibitory input increases excitability of γ and α neurons, producing spasticity and involuntary muscle activity. Spasticity contributes to musculoskeletal pain, contractures, and joint subluxation.

Medical treatment of CP

The term static encephalopathy is often used synonymously with CP; this description is apt because it conveys the picture of a nonprogressive disorder that is the result of a brain lesion or malformation. Because there is no known repair for a damaged brain, there is no cure for CP. Spasticity results in skeletal muscle weakness and loss of fine motor control and is a grave problem for a person with CP. The presence of spasticity in a child undergoing skeletal maturation may precipitate joint contractures and dislocation. Early medical intervention to control spasticity and prevent contractures may reduce the need for future orthopedic surgical intervention.

The primary goal of medical therapy is to increase the person’s independence and improve the caretaker’s ability to provide daily general care. To meet this goal, medical therapy must focus on a general reduction of spasticity, minimizing joint contracture, improving and maintaining range of motion, and decreasing accompanying pain. This goal is accomplished through carefully planned physical therapy and pharmacological support. The most common pharmacological agents used for the treatment of spasticity include those that act peripherally (botulinum toxin [Botox A]) and those that act centrally (baclofen [Lioresal], diazepam [Valium], vigabatrin [Sabril], and tizanidine [Zanaflex]).

Treatments for CP are judged on the basis of their effect on 2 major life areas for patients and families: ease of ambulation (in diplegic and hemiplegic CP) and ease of caretaking and patient comfort (in quadriplegic CP). In some cases, tone-altering, centrally acting medications help achieve these outcomes. Examples include baclofen, diazepam, tizanidine, and dantrolene [Dantrium].

The centrally acting, tone-reducing medications act on a specific receptor site in the central nervous system and have clinically significant side effects (Table 1). The most common side effect of these agents is sedation, which is often intolerable enough at the effective dose required to reduce tone that patients and/or caretakers often discontinue the medication. However, for the minority of patients who can achieve meaningful tone reduction without major side effects, these oral agents can significantly improve function and quality of life.

A more recent clinical approach for tone reduction involves direct intrathecal administration of baclofen using a surgically implanted, programmable, battery-powered pump. Because the baclofen is administered directly into the spinal fluid, the dose needed for tone reduction is reduced to a small fraction of the oral dose, and side effects are minimized. This treatment
method has the risks inherent with any nonbiological implant, including infection and mechanical malfunction, so it is offered only in selected cases where the risk/benefit ratio is highly favorable.

A targeted intramuscular injection of botulinum toxin (Botox A) may be chosen for the peripheral management of spasticity. These injections into the gastrocnemius or hamstrings are undertaken with topical, local, or general anesthesia. Botox A produces a functional muscular denervation by inhibiting the presynaptic release of acetylcholine at the neuromuscular junction. The resulting paralysis is dose-dependent, and the duration of paralysis is dependent on the generation of new motor endplates. The onset of action following intramuscular injection is up to 1 week, and the expected duration of action approaches 3 months. The preoperative injection of Botox A may be helpful for relief of postoperative pain due to muscle spasm.27 Side effects include muscular weakness that prevents limb function and ambulation, leg cramps, pain at the injection site, and lethargy.28 Generalized muscle weakness as a result of systemic spread is rare.

**Surgical treatment of CP**

Surgical procedures for patients with CP are considered for 2 main reasons: to reduce tone in the extremities or the trunk and to correct the acquired skeletal deformities of long-standing spasticity. Traditionally, orthopedic surgery was the only surgical option available and was directed at the musculoskeletal complications of increased muscle tone, such as muscle and joint contractures, deformities of the feet and extremity long bones, and scoliosis. More recently, a neurosurgical treatment to reduce spasticity has been developed. This procedure, called selective dorsal rhizotomy (SDR), decreases muscle tone in the lower extremities. Finally, surgery to implant a computer-controlled pump for intrathecal baclofen (ITB) therapy has been the most recent surgical advance.

- **Selective dorsal rhizotomy.** Described by Fasano et al.29 in 1976 and popularized by Peacock and Arens,30 SDR is a neurosurgical procedure intended to reduce spasticity in the lower limbs. Because spasticity is an unregulated response of a muscle to stretch, interruption of the stretch reflex arc should theoretically decrease spastic muscle tone. The SDR accomplishes this by surgical division of the dorsal rootlets of the spinal nerves in the lumbar region. During the procedure, intraoperative evoked potentials identify the contributing rootlets responsible for lower extremity spasticity. These are subsequently divided while the rootlets that are noncontributory are left intact. Proper patient selection is crucial for a successful outcome because tone reduction in a patient with little underlying strength may result in less function postoperatively. The ideal candidate for SDR is a child with spastic diplegia who has good selective motor control and strength but tone that interferes with ambulation.

- **ITB therapy.** Baclofen has long been known to reduce tone following oral administration. In 1985, Dralle and colleagues31 reported successful administration of baclofen via the intrathecal route. Eight years later, Albright et al.32 documented the first series of patients who underwent implantation of a system consisting of an intrathecal catheter and a programmable, battery-operated pump for the continuous infusion of ITB. Orally administered baclofen does not cross the blood-brain barrier easily, so relatively high doses must be administered to achieve a concentration within the cerebrospinal fluid sufficient to provide the desired reduction in tone. Many if not most patients experience drowsiness at the usual oral doses. However, direct intrathecal administration of baclofen produces a superior clinical effect at a dose several hundred times lower than the oral dosing.

Most patients treated with ITB have quadriplegic CP. When a patient is identified as a potential candidate for ITB therapy, the family receives educational materials and the patient is scheduled for a single-

**Table 1. Commonly used oral antispasticity agents**

<table>
<thead>
<tr>
<th>Drug name</th>
<th>Receptor site</th>
<th>Side effects</th>
<th>Comments</th>
</tr>
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<tbody>
<tr>
<td>Baclofen</td>
<td>GABA&lt;sub&gt;B&lt;/sub&gt;</td>
<td>Sedation</td>
<td>Crosses blood-brain barrier poorly</td>
</tr>
<tr>
<td>Diazepam</td>
<td>GABA&lt;sub&gt;A&lt;/sub&gt;</td>
<td>Sedation</td>
<td>Potential for withdrawal if discontinued</td>
</tr>
<tr>
<td>Clorazepate</td>
<td>GABA&lt;sub&gt;A&lt;/sub&gt;</td>
<td>Mild sedation</td>
<td>Benzodiazepine analogue</td>
</tr>
<tr>
<td>Dantrolene sodium</td>
<td>Muscle cell membrane</td>
<td>Weakness and hepatotoxic effects</td>
<td>Not useful in ambulatory patients</td>
</tr>
<tr>
<td>Tizanidine</td>
<td>&lt;span&gt;α&lt;sub&gt;2&lt;/sub&gt;&lt;/span&gt; adrenergic</td>
<td>Sedation and hepatotoxic effects</td>
<td>Very sedating; used at bedtime</td>
</tr>
</tbody>
</table>

GABA indicates γ-aminobutyric acid.
bolus injection trial. The trial involves administration of a single intrathecal dose of baclofen via lumbar puncture. Subsequently, the patient is carefully monitored for a decrease in muscle tone during the next several hours as the baclofen effect peaks and fades. If the patient's tone is visibly less during the observation period, the patient is considered a responder to ITB therapy, and pump implantation may be offered. The patient will subsequently be admitted and scheduled for an intraoperative baclofen pump implantation. This procedure is generally facilitated in the lateral position during general anesthesia.

Surgical implantation of the baclofen pump and catheter is performed by a variety of practitioners; however, neurosurgeons and orthopedic surgeons implant the majority of ITB therapy pumps. The potential complications following ITB therapy pump implantation include infection, spinal fluid leak or fistula formation, and mechanical problems with the catheter. The complication rate reported in most case series is approximately 10%. Pump refills are performed percutaneously in the outpatient setting. The pump is interrogated and programmed with a small computer that communicates with the pump transcutaneously via radiofrequency.

**Orthopedic surgery**

The majority of surgery performed on children with CP is orthopedic to remedy the chronic effects of increased tone on the muscles and bones of the extremities and spine. The orthopedic procedures address the contractures and deformities that develop during the child’s growing years. Generally speaking, people with greater degrees of spasticity are more likely to require surgical intervention. Accordingly, a patient with spastic quadriplegia often requires repeated surgical intervention. Patients with spastic quadriplegia are at high risk for scoliosis and hip dislocation, both of which can threaten comfortable seating. Furthermore, a dislocated hip can become arthritic and painful during several years. Spasticity and resulting contractures of the adductors can interfere with the caretakers’ ability to provide perineal hygiene. Tight hips and knees are not conducive to positioning patients in standers and walkers. Ankle and foot deformities hamper shoe fitting and proper positioning of feet in wheelchairs. All of these problems have corresponding surgical solutions.

People with spastic diplegia have predominantly lower body involvement, so as a group, they also have many hip, knee, and foot procedures. They are at risk for hip dysplasia; so hip reconstructions may be necessary. Most patients have ambulatory potential, so surgeries are carefully planned and timed to maximize muscle balance and lower limb alignment for walking, tight hip flexors, hamstrings, adductors, or gastrocnemius may require lengthening. Osteotomies may be necessary to correct rotational deformities of the femur or tibia. Severe flat feet may result from muscle imbalance at the ankle level, which makes the foot unstable when body weight is imposed on it. A combination of tendon and tarsal bone procedures is usually needed.

Finally, spastic hemiplegia may produce surgical problems of upper or lower extremities. Virtually all children with spastic hemiplegia will walk, and their lower extremity dysfunction is at the level of the knee and below. Muscular correction involves Achilles and tibialis posterior tendon lengthening and skeletal correction of foot deformities. In contrast, upper extremity involvement is usually more severe than that in the lower extremities. Some patients undergo extensive multilevel surgical procedures for modest functional gains in fine motor skills.

A controversy exists in the orthopedic community about the ideal method of executing the numerous surgical procedures needed by so many patients with CP. During the past few decades, improved anesthetic and orthopedic techniques have enabled surgeons to perform more and more surgery in a single session. The result is the so-called single event multilevel surgery approach, in which virtually every significant muscle or bone problem in the limbs is corrected at one time. These undertakings may necessitate long anesthesia times, multiple surgeons, and repositioning of the patient from prone to lateral and supine positions during the stages of the operation. Opponents of single event multilevel surgery object to the prolonged anesthesia times and the intensive postoperative rehabilitation required. Neither approach has proven superior in prospective randomized trials.

**Preoperative anesthesia assessment**

A patient with CP may undergo a variety of repeated diagnostic interventions and surgical procedures to improve organ function and mobility (Table 2). A patient with dysarthria may be unable to participate in the preoperative assessment due to communicative difficulties. The person may have normal intelligence and complete understanding of the ongoing communication despite impaired communicative ability. The previous medical and surgical records, a thorough history from the primary care physician, and current information from the daily caretaker are essential. Not uncommonly, patients with CP have had repeated interventional diagnostic and surgical procedures, providing a historical record of care.

It is important to be sensitive to the concerns and requests of the patient and caretaker. For example, the caretaker may request that intravenous access be
placed in a specific extremity to facilitate postoperative care. The parent and/or caretaker may be an aggressive advocate for the person with CP. This aggressive advocacy arises from previous anesthetic and surgical experiences, frustration with the healthcare system, anger, and guilt.33

The preoperative interview must include a discussion of the medical and surgical history, current medications, drug and environmental allergies, and problems with general care. Multiple prior diagnostic and surgical procedures expose the person to latex allergens. Accordingly, a patient with CP has an increased risk for development of latex allergy.34 At our institution, the anesthetic environment is latex-free, and patients with CP are treated surgically as being latex-sensitive.

The physical examination provides time for the assessment of the degree of spasticity and limb contractures that may interfere with airway management, intravenous access, and surgical positioning. Observation of the general appearance of the skin for decubitus ulceration or infected sites is important before considering the use of regional anesthesia. Epidural analgesia may be considered for postoperative pain management.

Formal airway assessment may be difficult because of upper extremity or neck contracture, drooling, dental malocclusion, and repeated tongue thrusting in people with bulbar involvement. Dental caries, loose teeth, and accompanying temporomandibular joint ankylosis may make endotracheal intubation difficult. A review of previous anesthetic records is indispensable when planning airway management.

Commonly prescribed pharmacological agents include antiepileptic drugs, antispasmodics, anticholinergics, antacids, and laxatives. Antidepressant agents are not uncommonly prescribed for adolescents and adults with CP. A seizure history may be ascertained in as many as 25% to 30% of patients with CP. Antiepileptic drugs should be continued preoperatively and their scheduled administration continued postoperatively. Prescribed antiepileptic drugs include carbamazepine (Tegretol), valproic acid (Depakene), clonazepam (Klonopin), lamotrigine (Lamictal), phenytoin (Dilantin), and vigabatrin (Sabril). Carbamazepine and valproic acid are the most commonly prescribed antiepileptic drugs. Valproic acid has a variety of dose-dependent hematological side effects, including transient immune thrombocytopenia, red cell aplasia, acquired von Willebrand disease, and bone marrow failure. Acquired von Willebrand disease may increase intraoperative bleeding.35 These hematological side effects occur in as many as 55% of people, with valproic acid serum levels exceeding 100 µg/mL.35 Accordingly, the preoperative laboratory assessment of coagulation may be indicated in patients who have received long-term valproic acid.

### Table 2. Common surgical interventions for patients with cerebral palsy

<table>
<thead>
<tr>
<th>Intervention</th>
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<tbody>
<tr>
<td>Tracheostomy</td>
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<tr>
<td>Feeding gastrostomy</td>
</tr>
<tr>
<td>Remedy of gastroesophageal reflux</td>
</tr>
<tr>
<td>Diagnostic esophagoscopy or gastroscopy</td>
</tr>
<tr>
<td>Dental rehabilitation (restorative and dental extractions)</td>
</tr>
<tr>
<td>Corrective spinal surgery (for scoliosis)</td>
</tr>
<tr>
<td>Selective dorsal rhizotomy (relief of spasticity)</td>
</tr>
<tr>
<td>Corrective orthopedic procedures (tendon lengthening, contracture releases)</td>
</tr>
</tbody>
</table>

Gastrointestinal reflux and impaired pharyngeal function with pooling of oral secretions are common comorbidities that are medically managed with antireflux agents, antacids, and anticholinergics. Ultimately, patients may need Nissen fundoplication.36 Glycopyrrolate (Robinul) may be prescribed for the control of oral secretions. Intravenous administration will decrease pharyngeal pooling intraoperatively. However, this may desiccate bronchial secretions, increasing the risk of postoperative pulmonary dysfunction. Patients with impaired swallowing (bulbar palsy) accompanied by tongue thrusting and drooling may require definitive management with salivary duct reimplantation.

Respiratory morbidity is common as a result of ineffective cough and chronic aspiration of pooled pharyngeal secretions, precipitating the development of recurrent pneumonia. Respiratory dysfunction is the leading cause of death in CP.37 Chronic aspiration produces pulmonary mucosal damage leading to the development of reactive airway disease. A patient with scoliosis may develop restrictive pulmonary disease characterized by a reduction in total lung capacity, vital capacity, and resting lung volume.

Preoperative sedation may be offered to patients with CP but should not be administered indiscriminately. The majority of patients with CP have generalized hypotonia, decreasing airway tone and increasing the risk of aspiration. Accordingly, patients with CP may be particularly sensitive to the depressant effects of sedative-hypnotic agents. The parent or caretaker should be consulted regarding the patient’s tolerance to previous premedication. Vigilant observation with continuous pulse oximetry should accompany any premedication.

**Intraoperative anesthetic management**

Anesthetic induction may be accomplished with an intravenous or inhalation agent. An inhalation induction may be preferable because intravenous access...
may be difficult because of extremity contractures and scarring from previous intravenous access. Carefully monitored increases in anesthetic concentration are required because the minimum alveolar concentration for volatile agents is reduced in patients with CP.12 The minimal alveolar concentration for halothane is 20% lower in patients with CP (0.7% end-tidal in child with CP, 0.9% end-tidal in child without CP) and is reduced an additional 10% in people taking concomitant antiepileptic drugs.38 The bispectral index score is lower in patients with quadriplegic CP than in healthy patients during the administration of similar concentrations of sevoflurane.39

Airway management can be difficult in patients with marked upper extremity contractures and/or contractures of the musculature of the neck. Patients with CP may be managed with a laryngeal mask airway or endotracheal intubation. The specific airway management technique is dependent on the surgical procedure and the patient's underlying comorbidity (eg, gastroesophageal reflux).

With the presence of gastroesophageal reflux and increased oral secretions, anesthetists may select a rapid-sequence intravenous induction. Propofol and thiopental are acceptable choices. Although thiopental, unlike propofol, may be selected because of its lack of pain on injection, a number of patients with CP have reactive airway disease. The advantages of propofol administration include a known reduction of airway tone.40

Another consideration is the selection of a skeletal relaxant to facilitate airway management. Is succinylcholine clinically appropriate for patients with spastic CP? A 1985 study by Dierdorf and colleagues41 examined potassium release following succinylcholine administration and found no difference between children with and without CP. A hyperkalemic response following succinylcholine administration is a relatively rare event in children, and the aforementioned study may have failed to capture this event.

Denervation and immobilization are responsible for the appearance of acetylcholine receptors outside the confines of the neuromuscular junction, those responsible for the hyperkalemic response following succinylcholine administration. These extrajunctional acetylcholine receptors are responsible for an increased response to succinylcholine (lower dose required) and resistance (increased dose required) to nondepolarizing skeletal relaxants.42,43 Theroux and colleagues44 found that the effective dose of succinylcholine to depress the baseline twitch by 50% (ED50) and 95% (ED95) was reduced in patients with CP, suggesting that patients with CP have extrajunctional acetylcholine receptors. In a subsequent investigation via an examination of muscle biopsy specimens secured during spinal surgery in patients with CP, Theroux et al45 found that 30% of children with CP had extrajunctional acetylcholine receptors. This direct evidence suggests that the routine administration of succinylcholine should be avoided in patients with CP.

Patients with CP are resistant to nondepolarizing skeletal muscle relaxants (specifically, vecuronium) and require a greater initial dose and more frequent dosing intervals. Moorby and colleagues46 found that the time to recovery to 25% of control twitch height following an initial dose of 0.1 mg/kg of vecuronium required less than half the time required by children without CP. This finding supports the previously cited findings of Theroux et al45 of the presence of extrajunctional acetylcholine receptors. This effect may also be a consequence of acetylcholine receptor up-regulation as a result of immobilization and/or chronic antiepileptic drug therapy.47

Patients with CP typically have a chronically low fluid intake that, combined with a preoperative fast, results in relative hypovolemia at the time of anesthetic induction. We have found the preoperative hematocrit value (obtained following anesthetic induction) to be as high as 55%. Accordingly, patients with CP are at risk for prerenal and postoperative renal failure. Perioperative fluid homeostasis requires careful monitoring of administered fluid, urinary output, and the replacement of blood and fluid deficits. For prolonged surgical procedures, procedures with expected large blood and fluid losses, and patients with a known fluid deficit, the insertion of a urinary catheter should be considered.

Perioperative hypothermia is particularly problematic for patients with CP and is associated with increased surgical blood loss, postoperative wound infection, altered drug metabolism, increased duration of action of nondepolarizing muscle relaxants, and slowed recovery following inhalation agent administration.48 Prolonged extremity preparations, regional anesthetic procedures, insufflation of the chest or abdomen during laparoscopic procedures, and prolonged periods required for surgical positioning may quickly lead to hypothermia. Many patients with CP lack normal central thermoregulation. The operating room environment must be warmed before patient entry into the operating room. Additional strategies for avoiding perioperative hypothermia include the humidification of inspired gas, the use of low fresh gas flows, warming of intravenous fluids, and the use of an active warming method such as forced-air warming blankets. It is important to monitor core temperature during the perioperative period because the use of these active warming methods may result in unintended hyperthermia.
Patient positioning may be particularly difficult due to severe upper and lower extremity contractures. In some circumstances, this may preclude the use of the prone position. Extremity contractures may make extremity support by conventional methods difficult at best. Forceful positioning may result in skeletal and/or nerve injury. In addition, patients with CP are often malnourished, and, accordingly, the skin is at risk of injury from compressive and shearing forces of surgical positioning. Positioning on a malleable support or gel pad may minimize injury to the skin.

Following orthopedic procedures, extensive casting is used to maintain the corrected deformity. The application of a “tight” hip spica may decrease mesenteric blood flow and produce acute gastric dilatation. Cast application may mask significant blood loss. In extremity compartment syndromes, this is especially problematic following tibial osteotomy. The port or gelpad may minimize injury to the skin.

Surgical positioning. Positioning on a malleable surface may be challenging due to torsional deformity of the spine because there is an increased incidence of scoliosis in patients with CP. Continuous postoperative infusion of bupivacaine or ropivacaine, with hydromorphone or fentanyl, with the addition of clonidine monitored by an acute pain service is thought to provide better analgesia and fewer side effects than intermittent opioid boluses. The addition of clonidine may decrease postoperative muscle spasm.

Summary

Cerebral palsy is characterized by a nonprogressive motor impairment that develops from an antenatal, perinatal, or postnatal insult to the developing encephalon. Contemporary care of patients with CP is multidisciplinary and oriented to improve the quality of life. The challenge for anesthetists in the care of patients with CP is to understand the clinical manifestations, accompanying pathophysiology, and current medical therapy to facilitate a perioperative plan of care.

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