Lesson 244: PreAnesthetic Assessment of the Patient With Cerebral Palsy

C erebral palsy (CP) comprises a group of nonpro- gressive motor impairment syndromes secondary to lesions or anomalies of the brain that arise in the early stages of development. Lesions occur in single or multiple locations in the brain and result in motor and sensory deficits. Surgical intervention is one option for these patients. It is important that the clinical anesthetist be knowledgeable about available surgical procedures and the anesthetic implications in the treatment of CP.

LEARNING OBJECTIVES
At the end of this activity, the participant should be able to:
1. List the group of syndromes known as CP.
2. Describe the epidemiology of CP.
3. Identify the causes of CP.
4. Explain the pathophysiology of the disease.
5. Outline clinical manifestations in patients with CP.
6. Identify the associated diagnostic criteria for CP.
7. Discuss the anesthetic considerations of the patient with CP.
9. Identify currently available medical therapies.

CASE HISTORY
A 6-year-old Caucasian boy, weighing 18 kg, who had CP associated with bilateral spasticity of the limbs, was scheduled to undergo a selective dorsal rhizotomy to improve his ambulation. Physical examination findings revealed an undernourished, combative child with normal cardiac and pulmonary function. Preoperative vital signs and laboratory test results were within normal limits.

NEEDS STATEMENT
Cerebral palsy (CP) is a neurologic syndrome that results in dysfunction of the brain at an early age. Therapies are available for the treatment of patients with CP who may have motor and sensory deficits. Surgical intervention is one option for these patients. It is important that the clinical anesthetist be knowledgeable about available surgical procedures and the anesthetic implications in the treatment of CP.

Anesthesiologists should be prepared for the clinical evaluation and treatment of this patient population. Interventions to alleviate spasticity consist of oral medications, nerve blocks, selective dorsal rhizotomy, and intrathecal baclofen pumps. Other types of supportive treatment include physical therapy, orthopedic care, and other management strategies. In some cases, surgical procedures to cut nerves and reduce spasticity may be necessary to enable the patient to proceed with physical therapy.

Epidemiology
CP is a leading cause of childhood disability. The estimated incidence of CP is 1 to 2 cases per 1,000 live births. Interestingly, CP has been found to occur in all countries of the world and within all ethnic groups. Pre-maturity and low birth weight have been found to significantly increase the risk for developing CP. Advances in neonatal medicine and improved survival rates for premature infants are associated with an increased incidence of CP in low-birth weight children. Children with birth weights less than 1,500 g have a 9% chance of developing CP, compared with a 0.3% chance in infants weighing more than 2,500 g at birth. The distribution of CP cases in this population is estimated to be 85% for the congenital form and 15% for the acquired type.

Causative Factors
CP is an established symptom complex resulting from heterogeneous etiologies. Over the past 20 years, there has been significant progress in understanding these etiologies.

PREANESTHETIC ASSESSMENT
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2) You must achieve a score of 70% or better to earn CME credit.
3) The estimated time to complete this activity is 2 hours.

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Table 1. Comparison of Birth Weight and Incidence of Cerebral Palsy at Age 7

<table>
<thead>
<tr>
<th>Birth Weight, g</th>
<th>Incidence of Cerebral Palsy per 1,000</th>
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<tbody>
<tr>
<td>&gt;2,500</td>
<td>3.3</td>
</tr>
<tr>
<td>&lt;1,500</td>
<td>90.4</td>
</tr>
<tr>
<td>1,500–2,500†</td>
<td>22.9</td>
</tr>
<tr>
<td>1,000–1,500†</td>
<td>6.7</td>
</tr>
</tbody>
</table>

Small for gestational age.

*Appropriate for gestational age.


Table 2. Classification of Cerebral Palsy Based on Involvement in the Pathophysiology

<table>
<thead>
<tr>
<th>Type</th>
<th>Description</th>
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<tbody>
<tr>
<td>Spastic</td>
<td>DIPLEGIC: greater involvement of legs than arms</td>
</tr>
<tr>
<td></td>
<td>QUADRIPLEGIC: all 4 extremities equally involved</td>
</tr>
<tr>
<td></td>
<td>HEMIPLEGIC: one-sided involvement, greater involvement of arm than leg</td>
</tr>
<tr>
<td></td>
<td>DOUBLE HEMIPLEGIC: both sides; greater involvement of arms than legs</td>
</tr>
<tr>
<td>Dyskinetic</td>
<td>CHOREOATHETOID (HYPERKINETIC); snake-like writhing motion</td>
</tr>
<tr>
<td></td>
<td>DYSTONIC</td>
</tr>
<tr>
<td>Atlactic</td>
<td>MIXED</td>
</tr>
</tbody>
</table>


Pathophysiology

The underlying pathophysiological finding in the preterm infant with neurodevelopmental sequelae is periventricular leukomalacia (white matter damage), which encompasses a variety of lesions including germinal matrix hemorrhage, periventricular hemorrhage, intraventricular hemorrhage, periventricular hemorrhagic infarction, and periventricular leukomalacia (PVL).2 Preterm infants are prone to neuronal damage because cerebral circulation is sensitive to changes in blood pressure (autoregulatory mechanisms are not fully developed) and lacks supporting neuroglia in the germinal matrix.2

PVL in premature infants represents a major precursor for neurologic and intellectual impairment and the onset of CP in later life.2 PVL is associated with periventricular hemorrhage, basal ganglia, and cortical-subcortical damage comprising approximately one third of the cases.3 Factors that predispose to PVL include birth trauma, asphyxia and respiratory failure, cardiopulmonary defects, premature birth/low birth weight, associated immature cerebrovascular development, and lack of appropriate autoregulation of cerebral blood flow in response to hypoxic-ischemic insults.12 In the past few years, epidemiologic and experimental studies have implicated intrauterine infection and chorionicamnionitis as causative in the pathogenesis of PVL.

PVL is characterized by multifocal areas of necrosis deep in the cortical white matter. The lesions are often symmetrically placed adjacent to the lateral ventricle and the foramen of Monro. The damage to white matter is distinguished by a loss of oligodendrocytes with an increase in hypertrophic astrocytes. The loss of oligodendrocytes affects nerve cell growth, which in turn impairs myelination.2 In patients with PVL there is essentially a lack of myelinations, neuropil, and accessory axons that result in poor transmission of impulses.2,2 The intrinsic vulnerability of oligodendrocyte precursors is considered central to the pathogenesis of PVL.2 The damage can be mediated by cytokines (such as tumor necrosis alpha, interleukin-6, and free radicals that are released as a result of hypoxia and ischemia.2 The ensuing brain damage affects descending inhibitory neurons, causing an inadequate release of gamma-aminobutyric acid (GABA) and a relative excess of excitatory transmitters—chiefly glutamate. Excessive excitatory stimulation of alpha motor neurons leads to spasticity because of the simultaneous activation of agonist and antagonist muscle groups.2 The magnitude of the cerebral lesions correlates directly with the severity of associated clinical symptoms.2 Clinically, children with more spasticity have more extensive brain damage, and thus a modulation of drug dosages is required.

Clinical Manifestations

The clinical presentation of CP varies significantly (Table 2). The spectrum of symptoms ranges from mild monoplegia to severe hemiplegia and quadriplegia.2 The children with PVL have more than one localization of lesion, and thus the time to diagnosis can extend well beyond the neonatal period.12 In the past few years, epidemiologic and experimental studies have implicated intrauterine infection and chorionicamnionitis as causative in the pathogenesis of PVL.2

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The study findings demonstrated that such individuals possess a low bone mineral density that was associated with a significant risk for fracture. The underlying pathophysiology is complex, and includes factors such as limited weight-bearing ambulation during skeletal growth and poor nutrition.10 Positioning of these patients during anesthesia should be done carefully and checked frequently. In patients with CP, there are many secondary effects of the disease. Difficulties in parent-child interactions can arise from misconceptions. For example, an abnormal labyrinthine reflex can cause a baby to exhibit an exaggerated extensor posturing, often perceived by a parent as “stiffness” or “immobility.”15 The nerve damage and brain atrophy of children with CP are likely to affect the ability of children with CP to ambulate during skeletal growth and poor nutrition.10 Positioning of these patients during anesthesia should be done carefully and checked frequently. In patients with CP, there are many secondary effects of the disease. Difficulties in parent-child interactions can arise from misconceptions. For example, an abnormal labyrinthine reflex can cause a baby to exhibit an exaggerated extensor posturing, often perceived by a parent as “stiffness” or “immobility.”15

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Dyskinetic movements can be subdivided into 2 main types. The first type, dystonic, is characterized by abnormal variations in general muscle tone with movement. The second type, choreoathetoid, is characterized by the initiation of movement and the continuation of movements of other muscle groups. Individuals with this type of CP demonstrate slow, writhing movements in addition to involuntary, irregular “jerky” movements.2 Ataxic CP may be considered as a subtype of dystonic CP or even as a separate group. It is a result of cerebellar dysfunction and is characterized by dysmetria, a disturbance in the coordination of voluntary movements.2

Mental retardation, the most serious of the sequelae associated with CP, is found in only 30% to 50% of patients. It is important for the anesthesiologist to realize that although a child may appear to have severe neurologic damage, he or she may in fact have normal intelligence. Before speaking with the patient, the anesthesiologist should ascertain the patient’s level of comprehension. CP has also been linked to other significant neurologic findings. Children with dystonia and athetosis CP may have significant problems with speech and language development. Children with CP are prone to abnormal postures and, rarely, autism.13

A propensity for bone fractures (with minimal trauma) and decreased bone density is common in patients with moderate to severe CP.2 The study findings demonstrated that such individuals possess a low bone mineral density that was associated with a significant risk for fracture. The underlying pathophysiology is complex, and includes factors such as limited weight-bearing ambulation during skeletal growth and poor nutrition.10 Positioning of these patients during anesthesia should be done carefully and checked frequently. In patients with CP, there are many secondary effects of the disease. Difficulties in parent-child interactions can arise from misconceptions. For example, an abnormal labyrinthine reflex can cause a baby to exhibit an exaggerated extensor posturing, often perceived by a parent as “stiffness” or “immobility.”15 The nerve damage and brain atrophy of children with CP are likely to affect the ability of children with CP to ambulate during skeletal growth and poor nutrition.10 Positioning of these patients during anesthesia should be done carefully and checked frequently. In patients with CP, there are many secondary effects of the disease. Difficulties in parent-child interactions can arise from misconceptions. For example, an abnormal labyrinthine reflex can cause a baby to exhibit an exaggerated extensor posturing, often perceived by a parent as “stiffness” or “immobility.”15
Mechanism of Action

- **α**

Side Effects and Precautions

- **α**

Table 3. Medications for the Treatment of Spasticity in Children

<table>
<thead>
<tr>
<th>Drug</th>
<th>Mechanism of Action</th>
<th>Side Effects and Precautions</th>
<th>Pharmacology and Dosing</th>
</tr>
</thead>
<tbody>
<tr>
<td>Baclofen</td>
<td>Bonds to GABA (B) receptors in the spinal cord to inhibit reflexes reducing tone</td>
<td>Sedation, confusion, nausea, dizziness, muscle weakness, hypotonia, ataxia, and parenthesias. Can cause loss of seizure control. Withdrawal can produce seizures, rebound hypotonia, fever, and death.</td>
<td>Rapidly absorbed after oral dosing; mean half-life: 3.5 h; Excretion: primarily renal. Dosage: start at 2.5-5 mg/d, increase to 30 mg/d (in children aged 2.7 y) or 60 mg/d (in children aged 28 y)</td>
</tr>
<tr>
<td>Diazepam</td>
<td>Facilitates postsynaptic binding of GABA in the brain stem and spinal cord to inhibit reflexes reducing tone</td>
<td>CNS depression causing sedation, decreased motor coordination, impaired attention, and memory loss. Overdoses and withdrawal occur. Sedative effect generally limits use to severely affected children</td>
<td>Well absorbed after oral dosing; mean half-life: 20-80 h. Metabolism: primarily hepatic. Dosage: 0.12-0.80 mg/kg/d in divided doses</td>
</tr>
<tr>
<td>Clonidine</td>
<td>α2-agonist; acts in the brain and spinal cord to enhance presynaptic inhibition of reflexes decreasing tone</td>
<td>Bradycardia, hypotension, dry mouth, drowsiness, dizziness, constipation, and depression. Side effects are common and lead to disconltdiation of the medication by one half of patients.</td>
<td>Well absorbed after oral dosing; half-life: 5-19 h. Metabolism: half via liver, half via renal excretion. Dosage: start with 0.05 mg bid; titrate up until side effects limit tolerance; maximum, 36 mg/d</td>
</tr>
<tr>
<td>Clonazepam</td>
<td>α2-agonist; acts in the brain and spinal cord to enhance presynaptic inhibition of reflexes decreasing tone</td>
<td>Dry mouth, sedation, dizziness, visual hallucinations, elevated liver enzymes, insomnia, and muscle weakness.</td>
<td>Well absorbed after oral dosing; half-life: 2.5 h. Extensive first-pass metabolism in liver. Dosage: start with 2 mg qhs and increase until side effects limit tolerance; maximum, 36 mg/d</td>
</tr>
<tr>
<td>Dantrolene</td>
<td>Works directly on the muscle to decrease muscle force produced during contrac- tion. Little effect on smooth and cardiac muscles</td>
<td>Most important side effect is hepatotoxicity, which may be severe. Liver function must be initially monitored monthly, then several times per year. Other side effects include mild sedation, dizziness, diarrhea, and parenthesias.</td>
<td>Oral dose is absorbed approximately 70% in small intestine; half-life: 15 h. Metabolism: primarily hepatic. Dosage: 0.5-10 mg bid to maximum 3 mg/kg qd</td>
</tr>
<tr>
<td>Baclofen</td>
<td>Binds to GABA (B) receptors in the spinal cord to inhibit reflexes reducing tone</td>
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**Therapeutic Interventions**

The complete medical care of a child with CP requires the interdisciplinary efforts of physicians, nurses, and therapists. Appropriate treatment is essential to improve physiologic deficits, assist in motor progression, and maintain appropriate levels of physical fitness, thus increasing the individual’s level of participation in the activities of daily life. The treatment of CP is centered on repair of the brain lesion, if possible, and management of impairments and disabilities.

**Repair of the Injured Brain**

Unfortunately, no medical therapies currently exist for repair of damage to those regions of the CNS that control muscle coordination and movement. Repair strategies now being investigated include the replacement of injured brain cells through stem cell research, the repair of injured neuronal processes, and stimulation of the development of alternative neuronal pathways. Currently, there are several treatments that can reduce the degree of impairment produced by the brain lesions of CP.

**Physical and Occupational Therapy**

Physical and occupational therapy is the treatment of choice for children with CP who are less than 3 years of age. The primary goals are to improve function, decrease spasticity, and enhance the quality of life. In addition, occupational therapy provides an opportunity for parents to learn procedures to help the child develop the necessary skills for self-care. Occupational therapy is task-oriented and focuses on fine motor skills. These therapies can comprise the primary treatment or serve as an adjunct to the interventions discussed below.

**Botulinum Toxic**

Injections of botulinum toxin have been used for the past decade to decrease muscle tone in children with spastic CP. Botulinum toxin inhibits the release of synaptic vesicles at the neuromuscular junction and decreases the level of muscle contraction. Each injection produces relaxation of a specific muscle for 4 to 6 months. The number of muscles that can be injected is limited by the total dose that can be administered.

**Selective Dorsal Rhizotomy**

Selective dorsal rhizotomy involves the surgical transection of selected sensory nerves entering the lower spinal cord. The challenge with this procedure is to identify and separate the nerve rootlets responsible for abnormal motor responses, while leaving normal rootlets intact. The pathologic rootlets are identified by direct stimulation while monitoring muscle action potential via electromyography. The pathologic rootlets are then sectioned to decrease afferent nerve conduction, but retain sensory and proprioceptive fibers.

**Surgery**

Surgery is an invasive approach for reducing spasticity of the lower limbs. For improvement in lower limb functionality, the procedure must be followed by an intense program of physiotherapy. In a comparative study of the efficacy of selective dorsal rhizotomy versus orthopedic surgery, Buck- on et al demonstrated that self-care skills, mobility, and social function gains were seen earlier and with greater frequency in those patients who underwent selective dorsal rhizotomy. Brachial plexus dorsal rhizotomy has been shown to release spasticity in the hand and improve function. Movement speed and dexterity, and grasp and pinch strength, can be improved while preserving sensibility. The procedure is also beneficial in athetotic patients. Un’estimative side effects to bowel and bladder function sometimes occur. Anesthesia is necessary during the procedure, as the patient must remain still for extended periods.

**Orthopedic Intervention**

Early and continuous involvement by an orthopedist is key to providing the most realistic and appropriate surgical interventions for a child with CP. Deciding when and how to intervene is often challenging. Also, obtaining proper informed consent from the patient might be difficult. The anesthesiologist must ensure that the patient is competent or that the healthcare proxy or next of kin consents with the anesthetic plan.
extensively to operate is extremely important because of variability in growth.7 Orthopedic procedures repair skeletal misalignment and joint fixation resulting from spasticity, tendon force, and contractures. These procedures have been proven to be efficacious in the control or elimination of muscle spasticity.7 Orthopedic surgical intervention should be avoided until the gait of the patient is mature.2 Adjunct procedures include serial casting and the use of orthoses—techniques that have been shown to improve function by decreasing tone and improving range of motion.7 Orthopedic procedures can directly restore function and set the stage, anatomically, for physiotherapy.1

Oral Medications

The recent use of oral medications for the treatment of CP has generally been unsuccessful (Table 3, page 75).2 The drugs used often have only modest benefits, but many undesirable side effects.5 Many of the agents can reduce spasticity in patients with CP, but none improve coordination.7 Oral medication is not recommended in patients less than 3 years of age.7 Diazepam and oral baclofen facilitate the presynaptic effects of GABA. They also potentially inhibit the postsynaptic areas of the spinal cord, alleviating the chronic disinhibition associated with CP. These drugs are effective in reducing tone, but often cause significant sedation.18 Studies conducted by Hansel and colleagues indicate that baclofen may be used in patients less than 3 years of age.2

Dantrolene acts directly on skeletal muscle to decrease the force of contraction while generating minimal cognitive effects.21 It has not been proven to be effective in children.2 The most important side effect associated with dantrolene is hepatotoxicity, which occurs in 2% of patients.2

Tizanidine, an α2-agonist, acts on the CNS to increase presynaptic inhibition of spinal reflexes.2 The side effects of this drug are sedation, hallucinations, and liver damage.2 Although these drugs can reduce spasticity, the functional improvements associated with their use are often not significant enough to justify their adverse side effects.30 Sedation and weakness can occur even at therapeutic drug levels and are usually unacceptable to the patient.2 Some reports have indicated that oral medications in conjunction with physical therapy can improve ease of care for the patient.2

Management of the Case Presented

After administration of gastric prophylaxis, anesthesia was induced in the patient with sevoflurane and oxygen by mask. A 22-gauge IV catheter was inserted into the dorsal region of the patient’s right hand. Movacurium was administered and a 5.5-mm endotracheal tube was placed after visualization of the vocal cords with a Miller-2 blade. The procedure was uneventful and the patient was later discharged without incident.

Conclusion

It is important for anesthesiologists to be aware of the pathophysiology, clinical manifestations, and treatments associated with CP. There is no effective cure for CP. Only heightened awareness and understanding of the syndrome will enable safe and effective anesthetic management of this challenging patient population.

References


