

# Cerebral Palsy: An Overview

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The presentation of cerebral palsy can be global mental and physical dysfunction or isolated disturbances in gait, cognition, growth, or sensation. It is the most common childhood physical disability and affects 2 to 2.5 children per 1,000 born in the United States. The differential diagnosis of cerebral palsy includes metabolic and genetic disorders. The goals of treatment are to improve functionality and capabilities toward independence. Multispecialty treatment teams should be developed around the needs of each patient to provide continuously updated global treatment care plans. Complications of cerebral palsy include spasticity and contractures; feeding difficulties; drooling; communication difficulties; osteopenia; osteoporosis; fractures; pain; and functional gastrointestinal abnormalities contributing to bowel obstruction, vomiting, and constipation. Valid and reliable assessment tools to establish baseline functions and monitor developmental gains have contributed to an increasing body of evidenced-based recommendations for cerebral palsy. Many of the historical treatments for this ailment are being challenged, and several new treatment modalities are available. Adult morbidity and mortality from ischemic heart disease, cerebrovascular disease, cancer, and trauma are higher in patients with cerebral palsy than in the general population. (*Am Fam Physician* 2006;73:91-100, 101-2. Copyright © 2006 American Academy of Family Physicians.)

► **Patient information:**  
A handout on cerebral palsy, written by the author of this article, is provided on page 101.

Cerebral palsy is characterized by motor impairment and can present with global physical and mental dysfunction. In 2001, the United Cerebral Palsy Foundation estimated that 764,000 children and adults in the United States carried the diagnosis of cerebral palsy. In addition, an estimated 8,000 babies and infants, plus 1,200 to 1,500 preschool-age children are diagnosed with cerebral palsy every year in the United States.<sup>1</sup>

## Etiology

Cerebral palsy is a static neurologic condition resulting from brain injury that occurs before cerebral development is complete. Because brain development continues during the first two years of life, cerebral palsy can result from brain injury occurring during the prenatal, perinatal, or postnatal periods.<sup>1,2</sup> Seventy to 80 percent of cerebral palsy cases are acquired prenatally and from largely unknown causes. Birth complications, including asphyxia, are currently estimated to account for about 6 percent of patients with congenital cerebral palsy.<sup>3</sup> Neonatal risk factors for cerebral palsy include birth after fewer than 32 weeks' gestation, birth weight of less than 5 lb, 8 oz (2,500 g), intrauterine growth retardation, intracranial hemorrhage, and trauma. In about 10 to 20 percent of patients, cerebral palsy is acquired postnatally, mainly because

of brain damage from bacterial meningitis, viral encephalitis, hyperbilirubinemia, motor vehicle collisions, falls, or child abuse.<sup>3</sup>

## Diagnosis

Observation of slow motor development, abnormal muscle tone, and unusual posture are common initial clues to the diagnosis of cerebral palsy. Assessment of persistent infantile reflexes is important. In infants who do not have cerebral palsy, the Moro reflex is rarely present after six months of age, and hand preference rarely develops earlier than 12 months of age. Hand preference may occur before 12 months of age if spastic hemiplegia is present.<sup>3</sup> Progressive hereditary neurologic or metabolic disorders must be eliminated as the cause of observed abnormalities (*Table 1*<sup>2</sup>). The testing strategy is based on the clinical picture, pattern of development of symptoms, family history, and other factors influencing the probability of specific diagnoses. Targeted laboratory tests and cerebral imaging using computed tomography, magnetic resonance imaging, and ultrasound are useful physical diagnostic tools. Surveillance for associated disabilities such as hearing and vision impairment, seizures, perception problems with touch or pain, and cognitive dysfunction can help complete the clinical assessment and determine the diagnosis.

## SORT: KEY RECOMMENDATIONS FOR PRACTICE

<i>Clinical recommendation</i>	<i>Evidence rating</i>	<i>References</i>	<i>Comments</i>
Use neurodevelopmental treatment (i.e., the Bobath method) to provide immediate improvement in dynamic range of motion; however, it may not consistently change abnormal motor responses, slow or prevent contractures, or facilitate more normal motor development of functional motor activities.	B	17	A 2001 American Academy for Cerebral Palsy and Developmental Medicine (AAPDM) evidence report stated that there was insufficient evidence for or against the use of neurodevelopmental treatment. A review of 15 studies had severe problems with small numbers of participants, methodologic problems, and inconsistent reporting of results.
Use intrathecal baclofen (Lioresal) to reduce spasticity in the lower extremities (unclear effects on upper extremities).	B	30	A 2000 AAPDM evidence report reviewed 14 small studies. A tentative conclusion was reached because of small numbers and problems in study design and reporting.
Use gastrostomy to benefit selected patients with cerebral palsy.	B	42	A 2003 AAPDM evidence report reviewed 10 studies of 281 children and stated that evidence is weak with little control of threats to internal validity and that conclusions remain highly speculative.

*A = consistent, good-quality patient-oriented evidence; B = inconsistent or limited-quality patient-oriented evidence; C = consensus, disease-oriented evidence, usual practice, expert opinion, or case series. For information about the SORT evidence rating system, see page 17 or <http://www.aafp.org/afpsort.xml>.*

### Clinical Features

Seventy to 80 percent of patients with cerebral palsy have spastic clinical features. Affected limbs may demonstrate increased deep tendon reflexes, tremors, muscular hypertonicity, weakness, and a characteristic scissors gait with toe-walking.<sup>4</sup> The athetoid or dysknetic type of cerebral palsy, affecting 10 to 20 percent of patients, is characterized by abnormally slow, writhing movements of the hands, feet, arms, or legs that are exacerbated during periods of stress and absent during sleep.<sup>3</sup> The rarest form, ataxic cerebral palsy, affects 5 to 10 percent of patients and predominately impairs balance and coordination. These patients walk with a wide-based gait and have intention tremors that complicate performance of daily activities requiring fine-motor function.<sup>3</sup>

Intellectual impairment occurs in about two thirds of patients with cerebral palsy. About one half of pediatric patients have seizures. Growth problems are common, as well as neurologic abnormalities such as impaired vision or hearing and abnormal touch and pain perceptions (*Table 2*<sup>3,5-7</sup>).<sup>3</sup> By definition, cerebral palsy is nonprogressive; therefore, children who experience loss of previously acquired skills, or who show slowing of development, disappearance of reflexes, or unusual body odors should be evaluated for genetic, metabolic, muscular, or neuronal tumor disorders that precipitate neurodegenerative conditions (*Table 1*).<sup>2</sup>

### Assessment Instruments

Several assessment instruments are available to quantify and monitor developmental milestones and skills and to assess the quality of life of patients and their caregivers (*Table 3*<sup>8-14</sup>). Readily available and useful assessment instruments include the Child Health Questionnaire,<sup>10</sup>

the Wong-Baker FACES Pain Rating Scale,<sup>14</sup> and the Gross Motor Function Classification System for Cerebral Palsy.<sup>12</sup> Functional scales such as the Gross Motor Function Classification System for Cerebral Palsy (*Table 4*<sup>12</sup>) standardize self-initiated movements and measure change in gross motor function over time, and this particular scale is widely accepted and easy to administer in the primary care office.<sup>12,15</sup> Other functional scales include: the Pediatric Evaluation of Disability Inventory,<sup>13</sup> a judgment-based, standardized instrument using parent report through a structured interview measuring both fine- and gross-motor movements related to self-care and mobility; the Functional Independence Measure (FIM)<sup>16</sup> for adults and the WeeFIM<sup>11</sup> for children, which measure the amount of assistance a person would require to perform activities of daily living; and the Ashworth<sup>8,9</sup> and Modified Ashworth<sup>9</sup> scales, which grade muscle spasticity.

### Management

The goal of management of cerebral palsy is not to cure or to achieve normalcy but to increase functionality, improve capabilities, and sustain health in terms of locomotion, cognitive development, social interaction, and independence. The best clinical outcomes result from early, intensive management.<sup>3</sup> Optimal treatment in children requires a team approach (*Table 5*).<sup>3</sup> A modern team approach focuses on total patient development, not just on improvement of a single symptom. Treatment programs encompass physical and behavioral therapy, pharmacologic and surgical treatments, mechanical aids, and management of associated medical conditions. In physical, occupational, speech, and behavioral therapies, the goals include enhancing patient and caregiver interactions while providing family support.

**TABLE 1**  
**Differential Diagnosis of Cerebral Palsy**

<i>Diagnosis</i>	<i>Features</i>	<i>Diagnostic test</i>
Arginase deficiency	No neonatal onset; progressive spastic diplegia; subsequent dementia	Measurement of serum amino acids shows a dramatic increase in arginine concentrations; ammonia levels elevated (range: 85 to 170 mcg per dL [50 to 100 $\mu$ mol per L])
Glutaric aciduria type 1	Progressive dystonia; choreoathetosis; progressive movement disorder developing during the first one to two years	Serum or urine test for the presence of glutaric acid
Juvenile neuronal ceroid-lipofuscinosis (i.e., Vogt-Spielmeyer disease)	Onset after five years of age; progressive loss of cognitive abilities; progressive extrapyramidal symptoms (e.g., rigidity, seizure, visual failure)	Characteristic inclusion bodies in skin fibroblasts or blood lymphocytes
Juvenile variant of metachromatic leukodystrophy	Decline in school performance; slowly progressive paraparesis	Lysosomal enzyme aryl sulfatase
Lesch-Nyhan syndrome	X-linked disorder of purine metabolism; choreoathetosis; self-mutilation; uric acid or orange crystals in the urine; mental retardation (IQ lower than 60)	Assay for the enzyme hypoxanthine-guanine phosphoribosyltransferase
Mitochondrial disorders	Ataxia; neuropathy; retinitis pigmentosa	Point mutation of mitochondrial DNA at the 8993 location
Niemann-Pick disease type C	Autosomal recessive disorder of intracellular processing of cholesterol; 70 percent have late childhood or adolescent onset; loss of vertical eye movements; choreoathetosis; dystonia; tremors	Demonstration of impaired cholesterol esterification on skin fibroblasts; the gene is on chromosome 18
Pelizaeus-Merzbacher disease	Leukodystrophy classification; mixed pyramidal and extrapyramidal symptoms; X-linked; slow rate of progression; pendular nystagmus; choreoathetosis; microcephaly; spastic quadriparesis	Deficiency of proteolipid protein (a primary myelin protein)
Rett syndrome	Primarily in girls; autistic features; choreoathetosis; progressive spasticity; characteristic loss of purposeful hand function resulting in continuous hand wringing; slow progression	Clinical diagnosis

*Adapted from Bass N. Cerebral palsy and neurodegenerative disease. Curr Opin Pediatr 1999;11:504.*

## Treatments

The types of treatment for patients with cerebral palsy depend on the patient's specific symptoms and range from physical therapy to medication use and surgery.

### GLOBAL STRATEGIES

Neurodevelopmental treatment (i.e., the Bobath method) is a common cerebral palsy treatment strategy that aims to control sensorimotor components of muscle tone, reflexes, abnormal movement patterns, postural control, sensation, perception, and memory by utilizing specific handling techniques. A 2001 American Academy for Cerebral Palsy and Developmental Medicine (AACPD) evidence report<sup>17</sup> stated that, although patients with neurodevelopmental treatment did show some immediate improvement in dynamic range of motion, there was no consistent evidence that neurodevelopmental treatment changed abnormal motoric responses, slowed or prevented contractures, or facilitated more normal motor development of functional motor activities.

Another specific cerebral palsy treatment, conductive education, was reviewed by AACPD in 2003.<sup>18</sup>

Conductive education emphasizes an integrated model of education and rehabilitation rather than a medical approach. The panel concluded that the current literature base does not offer enough conclusive evidence for an opinion for or against conductive education as an intervention strategy.<sup>18</sup>

### PHYSICAL THERAPY

Muscle strengthening and fitness programs are popular interventions for cerebral palsy; however, advocates of neurodevelopmental treatment advise against the use of resistive exercise, because it is believed to increase spasticity.<sup>19</sup> Several recent studies<sup>19-21</sup> have examined the effectiveness of resistive exercise. A study<sup>19</sup> using the stretch reflex as measured by the pendulum test found that children with cerebral palsy did not demonstrate increased spasticity of the quadriceps femoris muscle immediately following strengthening exercises as compared with children without cerebral palsy. The study<sup>19</sup> also showed that resistive exercise could be beneficial in muscle strengthening when muscle weakness causes dysfunction. A 10-week progressive strength training program for adults

**TABLE 2**  
**Common Complications of Cerebral Palsy Conditions**

<i>Condition</i>	<i>Complications</i>	<i>Care</i>
Abnormal neurologic control	Lack selective control of muscle activity and anticipatory regulation	Caregivers and patients should protect the joints and related soft tissues during movement, including avoidance of head injury.
Abnormal sensation and perception	Some children have impaired sensations to touch and pain with or without astereognosis.	Mittens may be needed during teething to prevent damage to fingers and hands.
Gastrointestinal problems (e.g., vomiting, constipation, or bowel obstruction)	Caused by delayed gastric emptying, abnormal autonomic control of gastrointestinal mobility, immobilization, inadequate oral intake, and prolonged colonic transit	Use stool softeners with narcotic pain medications. Perform bowel hygiene. Increase fluids and fiber with or without laxatives.
Hearing and vision abnormalities	Children may present with strabismus or hemianopia.  Visual defects occur in 25 to 39 percent of adult patients.  Eight to 18 percent of adults with cerebral palsy have hearing problems.	Screen early and periodically.
Impaired oral-motor functions	Can cause hypoxemia, temporomandibular joint contractures, vomiting, and aspiration pneumonia associated with gastroesophageal reflux, poor nutrition, failure to thrive, drooling, and communication difficulties	For feeding difficulties, use special diets, positioning, new feeding techniques, gastrostomy, or nasogastric tube feeding.*  Medications, surgery, and biofeedback have been used to control drooling.  Speech therapy and the use of computer voice synthesizers can help impaired communication.
Markedly reduced bone mass in nonambulatory adults and children	Can cause osteopenia, osteoporosis, fracture, scoliosis, or pain	Assess clinical conditions by physical examination and radiographic studies.  Use medications, vitamins, and mineral supplementation to reduce bone loss.  Encourage exercise.  Ask about or use instruments to qualify and monitor pain.  Adequately treat pain.
Mental health	Cognitive impairment is present in two thirds of patients with cerebral palsy.  Neurosis and psychosis also can occur.	Encourage functionality and independence with living accommodations, transportation, exercise, mechanical aids, or employment opportunities.  Provide counseling for emotional and psychological challenges.  Monitor for needed medications.
Seizures	One half of children with cerebral palsy demonstrate seizure activity.	Monitor and control with medication.
Spasticity and contractures	Spasticity prevents the stretching of muscles and tendons. Consequently, they do not grow at the same rate as lengthening bones, forming contractures and difficulty with ambulation and fine- or gross-motor movements.  Pain is created by hip dislocations, repetitive use syndromes, and degenerative joint disease.	Prevent with physical therapy with or without orthotic devices.  Treat with drugs, surgery, or cerebral stimulation.  Assess clinical condition by physical examination and radiographic studies.  Ask or use instruments to qualify and monitor pain.  Adequately treat the pain.
Urinary incontinence	Caused by impaired control of bladder muscles	Special exercises Biofeedback Prescription medications Surgery Surgically implanted devices to replace or aid muscles Specially designed undergarments

\*—Long-term use of nasogastric tube feeding is associated with nasal discomfort, recurrent aspiration pneumonia, and decreased survival.  
Information from references 3 and 5 through 7.

TABLE 3

**Clinical Assessment Instruments for Child Development and Quality of Life**

<i>Name of instrument</i>	<i>Age range</i>	<i>Use</i>
Ashworth Scale <sup>8,9</sup> and Modified Ashworth Scale <sup>9</sup>	Any Age	Method of measuring muscle spasticity
Caregiver questionnaire <sup>10</sup>	Any age	Cerebral palsy-specific measure of the effect of interventions on personal care, positioning, transferring, comfort, integration, and communication from the perspective of the caregiver
Child Health Questionnaire <sup>10</sup>	Child form: five to 18 years Parent/caregiver form: any age	Assesses a child's physical, emotional, and social well-being from the perspective of the parent or guardian and, in some instances, from that of the child.
Functional Independence Measure (WeeFIM) for children <sup>11</sup>	Six months to seven years	Measures functional performance in three domains: self-care, mobility, and cognition
Gross Motor Function Classification System for Cerebral Palsy patients <sup>12</sup>	One to 12 years	Assesses self-initiated movements (e.g., walking and sitting) over time and categorizes patients based on functional abilities and limitations; useful for planning interventions and evaluating outcomes
Pediatric Evaluation of Disability Inventory <sup>13</sup>	Six months to seven years	Measures capability and performance of functional activities in three domains: self-care, mobility, and social function
Wong-Baker FACES Pain Rating Scale <sup>14</sup>	Three years and older	Assesses intensity of pain using drawings of six faces whose expressions represent varying degrees of pain

*Information from references 8 through 14.*

who had cerebral palsy with spastic diplegia focused on the lower extremities and resulted in improved muscle strength, walking velocity, and gross motor function when standing and walking without spasticity when compared with a control group of adult patients with spastic diplegia.<sup>20</sup> Training was found to increase strength and possibly improve motor activity in persons with cerebral palsy in a literature review<sup>21</sup> of 10 empirical studies and one review study. No adverse effects were noted. Because only one randomized controlled trial was identified, more rigorous studies are needed.

Transporting a child with cerebral palsy to regularly scheduled physical therapy sessions can cause significant family stress. One study<sup>22</sup> compared attendees of an intermittent program (two to four treatments per week over a four-week period with rest periods of eight-week duration) with patients completing four visits a week without rest periods. Greater improvement was evident in the higher intensity phases; however, in both scenarios it was shown that the children did not regress during the rest period.<sup>22</sup> Superior clinical results have been observed in children participating in functional physical therapy activities when compared with those emphasizing normalization of movement.<sup>23</sup>

#### MEDICATIONS

**Botulinum toxin.** Upper motor neuron syndrome often leads to common patterns of motor dysfunction and characteristic spasticity and contractures.<sup>24</sup> Botulinum toxin (Botox) is a formulation of botulinum toxin type A, derived from the bacterium *Clostridium botulinum*. This bacterium produces a protein that blocks the release

of acetylcholine and relaxes muscles. Several studies have supported the use of botulinum toxin type A in the treatment of equine spasticity during walking,<sup>25-28</sup> but a literature review<sup>29</sup> did not find strong evidence to support or refute its use for the treatment of leg spasticity in patients with cerebral palsy. All studies reviewed used at least two injection sites in each calf, targeting the medial and lateral heads of the gastrocnemius.<sup>29</sup> All but two of the studies reviewed utilized 3- to 8-mouse units (mu)-per-kg botulinum toxin injections. Botulinum toxin type A injections have equivalent effectiveness to serial casting; however, longer-lasting effects and patient preference were seen with injections.<sup>28</sup>

Botulinum toxin type A, administered by ultrasound-guided intrasalivary gland injections, has been investigated to reduce salivary flow rate and correct pediatric drooling associated with cerebral palsy. A total dose of 30 to 50 units was diluted in a volume of 1.0 to 1.5 mL of saline. The solution was divided over two sites per gland and injected with a 25-gauge needle via ultrasound.<sup>5</sup>

**Baclofen (Lioresal).** The AACPDM Treatment Outcomes Committee Review Panel's systematic literature review in 2000<sup>30</sup> evaluated the use of intrathecal baclofen for spastic and dystonic cerebral palsy. Summary results reported limited evidence for reduced spasticity in the lower extremities, with unclear effects for upper extremities. Complications of therapy included somnolence, hypotonia, headache, nausea, vomiting, infections, cerebrospinal fluid leaks, and seizure activity. Although medical complications were common, function and ability of caregivers to render care appeared to improve.<sup>30</sup> A subsequent study<sup>31</sup> found the use of

**TABLE 4**  
**Gross Motor Function Classification System for Cerebral Palsy**

**Before second birthday**

Level I	Infants move in and out of sitting and floor sit with both hands free to manipulate objects. Infants crawl on hands and knees, pull to stand, and take steps holding onto furniture. Infants walk between 18 months and two years of age without the need for any assistive mobility device.
Level II	Infants maintain floor sitting but may need to use their hands for support to maintain balance. Infants creep on their stomachs or crawl on hands and knees. Infants may pull to stand and take steps holding onto furniture.
Level III	Infants maintain floor sitting when the low back is supported. Infants roll and creep forward on their stomachs.
Level IV	Infants have head control but trunk support is required for floor sitting. Infants can roll to supine and may roll to prone.
Level V	Physical impairments limit voluntary control of movement. Infants are unable to maintain antigravity head and trunk postures in prone and sitting. Infants require adult assistance to roll.

**Between second and fourth birthdays**

Level I	Children floor sit with both hands free to manipulate objects. Movements in and out of floor sitting and standing are performed without adult assistance. Children walk as the preferred method of mobility without the need for any assistive mobility device.
Level II	Children floor sit but may have difficulty with balance when both hands are free to manipulate objects. Movements in and out of sitting are performed without adult assistance. Children pull to stand on stable surface. Children crawl on hands and knees with a reciprocal pattern, cruise holding onto furniture, and walk using an assistive mobility device as preferred methods of mobility.
Level III	Children maintain floor sitting often by "W-sitting" (sitting between flexed and internally rotated hips and knees) and may require adult assistance to assume sitting. Children creep on the stomach or crawl on hands and knees (often without reciprocal leg movements) as their primary methods of self-mobility. Children may pull to stand on a stable surface and cruise short distances. Children may walk short distances indoors using an assistive mobility device and adult assistance for steering and turning.
Level IV	Children floor sit when placed but are unable to maintain alignment and balance without use of their hands for support. Children commonly require adaptive equipment for sitting and standing. Self-mobility for short distances (within a room) is achieved through rolling, creeping on the stomach, or crawling on hands and knees without reciprocal leg movement.
Level V	Physical impairments restrict voluntary control of movement and the ability to maintain antigravity head and trunk postures. All areas of motor function are limited. Functional limitations in sitting and standing are not fully compensated for through the use of adaptive equipment and assistive technology. Children at level V have no means of independent mobility and are transported. Some children achieve self-mobility using a power wheelchair with extensive adaptations.

**Between fourth and sixth birthdays**

Level I	Children get into and out of, and sit in, a chair without the need for hand support. Children move from the floor and from chair sitting to standing without the need for objects for support. Children walk indoors and outdoors and climb stairs. Emerging ability to run and jump.
Level II	Children sit in a chair with both hands free to manipulate objects. Children move from the floor to standing and from chair sitting to standing but often require a stable surface to push or pull up on with their arms. Children walk without the need for any assistive mobility device indoors and for short distances on level surfaces outdoors. Children climb stairs holding onto a railing but are unable to run or jump.

*continued*

TABLE 4 (continued)

**Gross Motor Function Classification System for Cerebral Palsy****Between fourth and sixth birthdays (continued)**

Level III	Children sit on a regular chair but may require pelvic or trunk support to maximize hand function. Children move in and out of chair sitting using a stable surface to push or pull up on with their arms. Children walk with an assistive mobility device on level surfaces and climb stairs with assistance from an adult. Children commonly are transported when traveling for long distances or outdoors on uneven terrain.
Level IV	Children sit on a chair but need adaptive seating for trunk control and to maximize hand function. Children move in and out of chair sitting with assistance from an adult or a stable surface to push or pull up on with their arms. At best, children may walk short distances with a walker and adult supervision but have difficulty turning and maintaining balance on uneven surfaces. Children are transported in the community. Children may achieve self-mobility using a power wheelchair.
Level V	Same as between second and fourth birthday.

**Between sixth and twelfth birthdays**

Level I	Children walk indoors and outdoors and climb stairs without limitations. Children perform gross motor skills including running and jumping but speed, balance, and coordination are reduced.
Level II	Children walk indoors and outdoors and climb stairs holding onto a railing, but they experience limitations walking on uneven surfaces and inclines, and walking in crowds or confined spaces. Children have at best only minimal ability to perform gross-motor skills such as running and jumping.
Level III	Children walk indoors and outdoors on a level surface with an assistive mobility device. Children may climb stairs holding onto railing. Depending on upper limb function, children propel a wheelchair manually or are transported when traveling for long distances or outdoors on uneven terrain.
Level IV	Children may maintain levels of function achieved before six years of age or rely more on wheeled mobility at home, school, and in the community. Children may achieve self-mobility using a power wheelchair.
Level V	Same as between second and fourth birthdays.

**Distinctions between levels I and II**

Compared with children in level I, children in level II have limitations in the ease of performing movement transitions, walking outdoors and in the community, the need for assistive mobility devices when beginning to walk, quality of movement, and the ability to perform gross-motor skills such as running and jumping.

**Distinctions between levels II and III**

Differences are seen in the degree of achievement of functional mobility. Children in level III need assistive mobility devices and often need orthoses to walk, whereas children in level II do not require assistive mobility devices after four years of age.

**Distinctions between levels III and IV**

Differences in sitting ability and mobility exist, even allowing for extensive use of assistive technology. Children in level III sit independently, have independent floor mobility, and walk with assistive mobility devices. Children in level IV function in sitting (usually supported), but independent mobility is very limited. Children in Level IV are more likely to be transported or to use power mobility.

**Distinctions between levels IV and V**

Children in level V lack independence even in basic antigravity postural control. Self-mobility is achieved only if the child can learn how to operate an electronically powered wheelchair.

*Adapted with permission from Palisano R, Rosenbaum P, Walter S, Russell D, Wood E, Galuppi B. Development and reliability of a system to classify gross motor function in children with cerebral palsy. Dev Med Child Neurol 1997;39:221-2.*

## Cerebral Palsy

continuous intrathecal baclofen in children with functional gross motor levels IV and V (*Table 4*<sup>12</sup>) provided substantial pain and spasm relief, and improved sleep, independence, and ease of care.

### SURGICAL TREATMENTS

Selective dorsal rhizotomy is a procedure intended to minimize or eliminate spasticity by selectively cutting dorsal rootlets from spinal cord segments L1 to S2. Postoperatively, it can create proprioceptive loss, bladder or bowel dysfunction, prolonged marked hypotonia, persistent back pain, or spinal deformities. A meta-analysis<sup>32</sup> of three randomized controlled trials revealed a direct relationship between the percentage of dorsal root tissue transected and functional gross-motor function improvement.

Muscle imbalance caused by spasticity can lead to complete dislocation of hips. The incidence of hip dislocation in children with cerebral palsy has been reported to be as high as 59 percent.<sup>33</sup> Approximately one half of patients with frank hip dislocation report pain.<sup>33</sup> Surgical treatment options include noninvasive abduction bracing, soft-tissue releases, major reconstructive femoral and/or pelvic osteotomies, and salvage procedures such as proximal femoral resection. A common surgical procedure for the subluxating hip is the proximal femoral varus-producing osteotomy in combination with appropriate soft-tissue releases. Unilateral surgery for unilateral subluxation appears to be effective in reducing and stabilizing the spastic hip without inducing instability in the contralateral hip, pelvic obliquity, or scoliosis.<sup>34</sup> Although only studied in a retrospective review,<sup>34</sup> the results of this surgery are promising and should encourage further investigations. Subluxation was easier to treat than dislocation. The majority of fair and poor results were seen in patients with spastic quadriplegia. After surgery, patients were immobilized in a hip spica cast for a minimum of six weeks. Adverse events reported included decubitus ulcers, lower extremity fracture at cast removal, and technical complications related to the use of the hardware.

### EXTERNAL AIDS

Orthoses are commonly used in conjunction with physical therapy, botulinum toxin type A, baclofen, and neurosurgery or orthopedic surgery to prevent inappropriate joint movements. A literature review<sup>35</sup> reported poor evidence-based support for the use of lower limb orthoses to prevent deformities or improve activities in children. An investigation of the usefulness of bodysuits made from elastic material has demonstrated functional gains,<sup>36</sup> but only one of the 12 caregivers wanted to continue use of this treatment

**TABLE 5**  
**Multispecialty Management Team**  
**for Children with Cerebral Palsy**

<i>Specialist</i>	<i>Role</i>
Physician*	Team leader; synthesizes long-term, comprehensive plans and treatments
Orthopedist	Focuses on preventing contractures, hip dislocations, and spinal curvatures
Physical therapist	Develops and implements care plans to improve movement and strength, and administers formal gait analyses
Occupational therapist	Develops and implements care plans focused on activities of daily living
Speech and language pathologist	Develops and implements care plans to optimize the patient's capacity for communication
Social worker	Assists the patient's family in identifying community assistance programs
Psychologist	Assists the patient and patient's family to cope with the stress and demands of the disability
Educator	Develops strategies to address cognitive or learning disabilities

\*—Family physician or pediatrician, with support or direction from children's neurologist, or children's psychiatrist trained to help developmentally disabled children, if available.

Information from reference 3.

modality because of toileting and incontinence problems with the appliance. No evidence-based indication currently exists for the use of hyperbaric oxygen therapy in the management of patients with cerebral palsy.<sup>37</sup>

Several studies have investigated electrical stimulation. One study<sup>38</sup> found it a useful adjunctive therapy for pediatric upper limb dysfunction; however, two other studies<sup>39,40</sup> did not find a significant clinical effect when it was applied to lower limb dysfunction. Cerebral stimulation to the superior-medial cerebellar cortex by an implantable, controlled-current pulse generator apparently can reduce seizure activity and spasticity of primitive reflexes, increase muscle tone and co-contraction, and reduce athetoid movements in patients with cerebral palsy.<sup>41</sup>

### Secondary Conditions

Impaired oral-motor function occurs commonly in patients with cerebral palsy, precipitating hypoxemia, temporomandibular joint contractures, vomiting, and aspiration pneumonia associated with gastroesophageal reflux (*Table 2*<sup>3,5-7</sup>). These conditions can cause lengthy mealtimes and fatigue, contributing to malnourishment. Treatments for such conditions have included nasogastric



tube feeds. Limitations of long-term use of nasogastric tube feeds include nasal discomfort, recurrent aspiration pneumonia caused by tube displacement, and decreased survival. The AACPDM evidence reports of 2003<sup>42</sup> addressed gastrostomy as an option for long-term treatment. The limited published evidence consistently supports gastrostomy as beneficial to most, but not all, patients with cerebral palsy.<sup>42</sup>

Markedly reduced bone mass in nonambulatory children and adults with spastic quadriplegia has been reported to place these persons at risk of osteopenia, osteoporosis, and fracture.<sup>43</sup> The prevalence of hip pain in nonambulatory patients is reported to be 47.2 percent.<sup>33</sup> The most common pain identified was provoked pain (e.g., during mobilization, palpation, and weight bearing on the lower extremities). Only 13.6 percent of nonambulatory adolescent and young adult study patients experiencing pain reported receiving pain treatment.<sup>33</sup>

Mental health can be affected by the chronic pain, social isolation, and loss of functionality and independence associated with cerebral palsy. Care plans must decrease barriers to participation in work, school, and community activities by utilizing mechanical devices such as motorized wheelchairs, voice synthesizers, and community environment modification. Periodic functional assessments are necessary to ensure that the equipment matches the current needs of each patient. Patients must be given an adequate knowledge base concerning injury prevention, safety, and transfer strategies.<sup>44</sup> Cognitively intact adult patients should be offered instruction in the evaluation and hiring of quality personal aids and caregivers, and in self-advocacy skills.<sup>6</sup> Children with cerebral palsy are at high risk of peer rejection and social isolation. Their caregivers and family members are equally at risk of depression and isolation, even in the presence of mild cerebral palsy symptoms.<sup>45</sup> Consequently, care plans should be based on a biopsychosocial model of health care delivery that includes family dynamics and the well-being of all concerned.

### Preventive Care for the Adult with Cerebral Palsy

The number of adults with cerebral palsy is increasing because of increased survival of low birth weight infants and increased longevity of the adult population. When compared with the general population, patients with cerebral palsy have higher mortality from ischemic heart disease, cerebrovascular disease, and digestive disorders.<sup>46</sup> They also appear to be at increased risk of breast and brain cancer. Preventable deaths from drowning (e.g., in swimming pools, hot tubs, and bathtubs) and from motor vehicle crashes involving pedestrians occur more

often in persons with cerebral palsy than in the general population.<sup>46</sup> Medical efforts to reduce cardiovascular risk should include exercise with physical accommodations for disabilities.<sup>7</sup> Regular health maintenance, including routine breast and pelvic examinations for the sexually active, should be encouraged. Pelvic examinations can be offered in the left lateral position.<sup>6,7</sup>

Adult mobility and ability to perform activities of daily living should be routinely monitored as the patient ages. The ability of patients to access adaptive devices and services may be decreased as they survive into adulthood because of declining social services and aging caregivers.<sup>6</sup> Placement options, medical surrogate identification, living wills, and power of attorney issues should be explored to ensure continuity of care.

Members of various family medicine departments develop articles for "Practical Therapeutics." This article is one in a series coordinated by the Department of Family and Geriatric Medicine at the University of Louisville School of Medicine, Louisville, Ky. Coordinator of the series is James G. O'Brien, M.D.

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