

# Cerebral Palsy: Comprehensive Review and Update

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Cerebral palsy (CP) is a common pediatric disorder occurring in about 2 to 2.5 per 1000 live births. It is a chronic motor disorder resulting from a non-progressive (static) insult to the developing brain. CP is the clinical presentation of a wide variety of cerebral cortical or sub-cortical insults occurring during the first year of life. The commonest cause of CP remains unknown in 50% of the cases; prematurity remains the commonest risk factor. Children with CP suffer from multiple problems and potential disabilities such as mental retardation, epilepsy, feeding difficulties, and ophthalmologic and hearing impairments. Screening for these conditions should be part of the initial assessment. The child with CP is best cared for with an individualized treatment plan that provides a combination of interventions. This requires the provision of a number of family-centered services that make a difference in the lives of these children and their families. Management of spasticity can be challenging with a wide variety of possible therapeutic interventions. The treatment must be goal oriented, such as to assist with mobility, reduce or prevent contractures, improve positioning and hygiene, and provide comfort. Each member of the child's multidisciplinary team, including the child and both parents, should participate in the serial evaluations and treatment planning.

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Cerebral palsy (CP) was first described in 1862 by an orthopedic surgeon named William James Little. A motor disorder resulting from a non-progressive (static) insult to the developing brain,<sup>1</sup> CP is, in fact, a clinical presentation of a wide variety of cerebral cortical or sub-cortical insults occurring during the first year of life. Preterm infants are at the highest risk for developing CP. The vulnerable brain is harmed during a critical period of development primarily by known CNS complications of prematurity such as intraventricular hemorrhage (IVH) and periventricular leukomalacia (PVL). Children with CP suffer from multiple problems and potential disabilities that require the provision of family-centered services that make a difference in the lives of these children and their families.<sup>2</sup> The aim of this article is to provide an updated overview of CP and review the most recent advances in clinical and therapeutic interventions.

## Epidemiology

The worldwide incidence of CP is approximately 2 to 2.5/1000 live births. The incidence is strongly associated with gestational age, occurring in 1 of 20 surviving preterm infants. It is important to note that although prematurity is the commonest risk factor for developing CP, the majority of affected children are full-term. This can be explained by the fact that there are many more full-term than preterm infants

born at a given time.<sup>3</sup> Despite the reduction in the rate of birth asphyxia from 40/100 000 in 1979 to 11/100 000 in 1996, no associated reduction in the prevalence or incidence of CP was seen.<sup>4</sup> In fact, the prevalence of CP in the USA increased by 20% (from 1.9 to 2.3/1000 live births) between 1960 and 1986.<sup>5</sup> This increase is likely related to the survival of very low birth weight premature infants.<sup>5</sup> There is also evidence of an associated increase in the severity of the disability.<sup>6</sup> This emphasizes the need for more efforts to decrease the rate of prematurity in addition to decreasing the associated neurological injury among these infants.<sup>5</sup>

#### Clinical manifestations and classification schemes

Children with CP usually present with developmental delay and motor deficits. The distinction between a static (non-progressive) and progressive clinical course is very important. Classically, loss of previously acquired milestones (regression) marks the onset of most metabolic and neurodegenerative disorders (NDD). However, some NDD or metabolic disorders have a slow rate of progression and can be misdiagnosed as CP.<sup>7</sup> Therefore, clear developmental regression may not be evident, particularly in the early stages of the disease or at a younger age of onset. In addition, the neurological consequences of CP may be delayed for several months because of the immaturity of the nervous system.<sup>7</sup> Motor deficits of CP include negative phenomena such as weakness, fatigue, incoordination and positive phenomena such as spasticity, clonus, rigidity, and spasms. Spasticity is a velocity dependent increased muscle tone with hyperreflexia resulting from hyperexcitability of the stretch reflex. It can lead to muscle stiffness, functional impairment, and atrophy. If not treated, it can progress to muscle fibrosis, contractures, and subsequent musculoskeletal deformities. CP can be classified according to the severity of motor deficits as mild, moderate, or severe. Several other classification systems exist based on the pathophysiology, etiology, and distribution of motor deficits as follows.

#### Pathophysiologic classification

Insults resulting in neuronal loss can be 1) cortical (pyramidal), resulting in spasticity, 2) basal ganglial (extrapyramidal), resulting in abnormal movements such as choreoathetosis, 3) cerebellar, resulting in hypotonia, or 4) mixed. Spastic CP is the most common type, accounting for up to 75% of cases.<sup>8</sup>

A smaller percentage of children with CP demonstrate extrapyramidal (dyskinetic) features, including combinations of athetosis, chorea, and dystonia. The abnormal movements usually develop in the second year of life and become most apparent during volitional motor activities with associated speech impairments.<sup>8</sup> Most children with extrapyramidal CP have normal intelligence, but their abilities can be underestimated due to the severity of their motor and communication deficits. Kernicterus (bilirubin encephalopathy) is a leading cause of extrapyramidal CP. The affected neonate appears weak, listless, and hypotonic, with poor feeding. Over a period of months, hypertonia, opisthotonus, choreoathetosis, and sensorineural hearing loss develops.<sup>8</sup> Hypotonic cerebral palsy occurs rarely; however, most children progress to other CP subtypes. Mixed CP occurs when the child displays a combination of features, such as spasticity and choreoathetosis.

#### Etiologic Classification

Up to 50% of CP cases have no identifiable underlying etiology.<sup>3</sup> The etiologies can be classified according to the timing of the insult as prenatal (commonest), natal, or postnatal. Another etiologic classification system depends on the actual cause such as congenital (developmental, malformations, syndromic) or acquired (traumatic, infectious, hypoxic, ischemic, TORCH infections, and others). Perinatal asphyxia is a cause in only 8% to 15% of all cases.<sup>9</sup> Most of these children have clinical features of neonatal hypoxic ischemic encephalopathy (HIE) such as a disturbed level of consciousness, seizures, and other end organ dysfunction. Although a normal cord pH excludes HIE, a pH of <7.0 is associated with encephalopathy in only 15% of infants.<sup>10</sup> Similarly, apgar scores are predictive of mortality but not sensitive in predicting the neurological outcome. Chorioamnionitis and maternal infections have been shown to be risk factors for HIE and CP.<sup>11</sup> PVL is the strongest and most independent risk factor for the development of CP. Ultrasonographic abnormalities of persistent ventricular enlargement or persistent parenchymal echodensities carry a 50% risk for CP, and large bilateral periventricular cysts carry a risk of 85%.<sup>12</sup> In another study, CP occurred in 56% of infants with PVL and IVH.<sup>13</sup>

#### Classification of Motor Dysfunction

CP can be classified according to the topographic distribution of motor involvement. Motor deficits include monoplegia, diplegia, hemiplegia, triplegia,

quadriplegia, and double hemiplegia. Diplegia is present when the lower extremities are primarily affected, although the upper extremities are not completely spared. Spastic diplegia is the most common type of CP and is associated with prematurity. The periventricular germinal matrix, which is a region of active neuronal proliferation, is particularly susceptible to bleeding and hypoxic ischemic injury. The surrounding periventricular white matter contains pyramidal fibers that descend through the internal capsule to supply the lower limbs. More peripheral in the periventricular white matter are the pyramidal tracts of the upper limbs. Therefore, periventricular insult in preterm infants affects the lower limbs more than the upper limbs, resulting in spastic diplegia. Note that the term paraplegia should not be used in this context as it implies a spinal cord insult resulting in lower motor neuron lesion involving the lower limbs only, i.e. not cerebral in origin with completely normal arm function. Hemiplegia is characterized by involvement of one side of the body, with the arm typically more affected than the leg. This is because of larger cortical representation (motor homunculus) of the hand and arm compared to a smaller leg area. Monoplegia refers to single limb involvement. This is usually the result of very mild hemiplegia with arm deficits only. When all four limbs are involved, quadriplegia is the appropriate descriptive term. This is the most disabling, with 25% of the affected children requiring total care.<sup>1</sup> Double hemiplegia refers to the child with quadriplegia involving the arms more than the legs with side asymmetry. Triplegia is rare and usually results from milder and very asymmetric double hemiplegia (sparing one leg) or milder asymmetric diplegia (sparing one arm). These subtypes can be difficult to delineate clinically in some children, particularly since the degrees of disability can vary widely within these subtypes.<sup>8</sup>

#### Associate Manifestations and Complications

##### Mental Retardation

Not all children with CP are cognitively impaired. In fact, the commonest type (spastic diplegic CP) is characterized by normal cognition because the lesion is in the periventricular white matter, i.e. sparing the cortical grey matter. However, there is a relationship between the severity of CP and mental retardation.<sup>14</sup> Children with spastic quadriplegic CP have greater degrees of mental retardation than children with spastic hemiplegia.<sup>14</sup> Other factors associated with

increased cognitive impairment include epilepsy and cortical abnormalities on neuroimaging.

##### Epilepsy

Up to 36% of children with CP have epilepsy, with onset in the first year of life in 70%.<sup>15</sup> Focal seizures with or without secondary generalization are most common with frequently focal EEG abnormalities.<sup>16</sup> Every effort should be made to avoid sedation prior to EEG as this may affect the result of the test.<sup>17</sup> Epilepsy can be an indicator of the severity of neurological injury (quadriplegic CP) or cortical insult (hemiplegic CP).<sup>18</sup> Children with spastic diplegic CP are at a lower risk for epilepsy mainly because their pathology predominantly involves the periventricular white matter. Several new antiepileptic drugs have improved our ability to control the seizures in these children.<sup>19,20</sup>

##### Feeding, Nutrition, and Growth

These are the most common issues encountered in children with severe CP. About 30% are undernourished, and many show reduced linear growth below the third percentile.<sup>21</sup> Although growth delays appear to be multifactorial in origin, the leading cause appears to be poor nutrition secondary to pseudobulbar palsy. This is an upper motor neuron disorder resulting in poor coordination of sucking, chewing, and swallowing. In addition, gastroesophageal (GE) reflux results in regurgitation, vomiting, and possible aspiration. GE reflux can be a source of pain and food refusals in the difficult-to-feed child. Dystonic dyspepsia (Sandifer's syndrome) in children with severe GE reflux can be confused with tonic seizures. Early nasogastric (NG) or gastrostomy tube (GT) feedings can be solutions to these problems with improved growth and greater family satisfaction (22). NG tube feeding can be used for short-term nutritional support. However, on a long-term basis, NG feeding is not socially acceptable and can be associated with nasal discomfort, sinusitis, irritation of the larynx, and recurrent tube blockage or displacement. Surgically or endoscopically placed GT provides a long-term solution to the feeding disorder in conjunction with treating the associated GE reflux (21). Fundoplication may be indicated at the time of GT placement if medical treatment for GE reflux fails.

##### Bladder Dysfunction

Children with CP are at increased risk for urinary incontinence, urgency, and infections.<sup>23</sup> Spastic CP can be associated with spasticity of the detrusor muscles

resulting in small frequent voids and a low capacity irritable bladder. Primary incontinence has been reported in up to 23% of these children and correlates with lower cognition and severe motor deficits.<sup>24</sup> The communication skills and physical ability to go to the bathroom promptly and manage clothing influences the attainment of continence. Adapted toilet seats, handrails, and clothing modifications can increase toileting successes.

#### Bowel Dysfunction

Constipation is common in children with CP and results from multiple factors including poor feeding, reduced water intake and immobility. The long-term solution involves increased consumption of water, juices, fruits, and vegetables. Initiating bowel evacuation is recommended and requires a combination of laxatives (upper intestinal tract) and enemas or suppositories (lower tract). Afterward, a schedule of softening agents such as artificial powdered fiber or docusate sodium with dietary modifications can result in more regular and softer bowel movements. Sitting on the toilet daily after the main meal takes advantage of the gastro-colic reflex and may be further stimulated occasionally with glycerin suppositories.<sup>23</sup> With effective bowel management programs, many children can attain reasonably regular bowel movements.

#### Sleep Disturbances

Sleep disorders are common in children with CP, particularly those with visual impairment, occurring in up to 50% of cases.<sup>25</sup> These children often have disturbed sleep patterns with fragmented sleep and frequent nocturnal awakenings, which is highly disruptive to parents. Medications that improve the sleep-wake cycle may also decrease spasticity and improve daytime behavior.<sup>25,26</sup> Hypnotics are generally effective for short periods but lose their effect in a few days due to tolerance. Melatonin is a recently developed natural compound with a phase setting effect on sleep. It is the hormone of darkness as the detection of darkness by visual receptors drives the hypothalamus to stimulate the pineal gland via sympathetic pathways to increase melatonin secretion.<sup>25</sup> Visual impairment diminishes the ability of the child to perceive and interpret the multitude of cues for synchronizing their sleep with the environment. This makes these children susceptible to circadian sleep-wake cycle disturbances. Up to 80% of children had a dramatic response to a 3-mg melatonin dose at bedtime with a reduction in delayed sleep onset, nocturnal awaken-

ing, and early arousals.<sup>25</sup> The drug has minimal side effects and no tolerance or dependence.

#### Drooling

Drooling occurs in up to 30% of children with CP.<sup>27</sup> It is not usually related to increased production of saliva unless an irritating lesion is present, such as dental caries or throat infection. Drooling is usually secondary to mouth opening and/or swallowing difficulties due to pseudobulbar palsy. It is not socially acceptable and can lead to aspiration, skin irritation, and articulation difficulties.<sup>27</sup> Management for this difficult problem is not very effective. Anticholinergic medications, such as glycopyrrolate, decrease salivation by blocking parasympathetic innervation. Side effects include irritability, sedation, blurred vision, and constipation.<sup>28</sup> Scopolamine is another anticholinergic agent that is available as a skin patch. Surgical re-routing of salivary ducts is an option, but may lead to increased aspiration.<sup>28</sup> Recent studies suggest that botulinum toxin injection into the parotid and submandibular glands may be an effective in reducing excessive drooling.<sup>29</sup>

#### Hearing Loss

Certain etiologies, such as kernicterus, post-meningitis, and congenital rubella, increase the risk for hearing loss. If not diagnosed and treated early, hearing loss can interfere with developmental progress and rehabilitation, thereby contributing further to developmental delays. Screening is recommended, including behavioral audiometry, auditory-evoked brainstem responses (ABR), or transient evoked otoacoustic emissions. ABR should be performed before or shortly after discharge from the neonatal intensive care unit for every preterm. Hearing assessment is recommended routinely for any child with global developmental delay, particularly if language delay is present. The yield may reach 91% if hearing loss was suspected clinically.<sup>30</sup>

#### Visual Abnormalities

Children with CP, particularly preterm infants, are also at increased risk for visual impairment, including retinopathy of prematurity, myopia, strabismus, glaucoma, and amblyopia.<sup>31</sup> If not diagnosed and managed early, visual deficits can interfere with developmental progress and rehabilitation. Strabismus can lead to permanent monocular vision loss (amblyopia). Visual impairments can be cortical due to damage to the visual cortex of the occipital lobes. Screening is recommended including acuity, eye

movements, and funduscopy. Visually evoked potentials assess the integrity of the visual pathway from the optic nerve to the visual cortex. Serial ophthalmologic assessments are recommended routinely on any child with global developmental delay, particularly if vision loss is suspected. The yield is 13% to 25% for refractive errors and strabismus, and 20% to 50% for visual impairment.<sup>30</sup>

### Orthopedic Abnormalities

The developing bones grow in the direction of the forces placed upon them. Spasticity can lead to progressive joint contractures, shortened muscles, and hip or foot deformities. Other orthopedic complications that need to be watched for include scoliosis and fractures due to osteomalacia or osteoporosis. These manifestations are more common with severe motor disability and immobility, such as quadriplegia.

### Diagnosis

Diagnostic labels should not be taken for granted as misdiagnoses are not uncommon. Many times the term CP is loosely applied to children with various chronic neurological disorders.<sup>7</sup> A comprehensive history for risk factors and genetic background, complete physical and neurological examinations are mandatory for accurate diagnosis. Serial developmental evaluations may be necessary in the young child for proper diagnosis and follow up.<sup>32,33</sup> Perinatal complications such as prematurity, head injury, kernicterus, and meningitis are important risk factors for CP. On the other hand, a family history of neurological disorders and early or unexplained deaths indicates an undiagnosed inherited neurodegenerative disorder. Familial CP is a misdiagnosis that should not be made. Occasionally CP recurrence occurs due to similar perinatal risk factors; however, a family history of CP should always raise the suspicion of an undiagnosed NDD or metabolic disorder. An example is glutaric aciduria type 1, which is an autosomal recessive disorder that results in a clinical picture similar to dyskinetic CP. Another rare disorder that can be confused with CP is dopa-responsive dystonia (Segawa disease). In one series, up to 24% of patients with dopa-responsive dystonia had been misdiagnosed as CP.<sup>34</sup> Dystonic movements do not usually cause the wasting, contractures, and deformities that develop in spasticity.<sup>35</sup> Patients usually have a good muscle bulk because of the repeated dystonic contractions. The clue to the

diagnosis of dopa-responsive dystonia is the diurnal fluctuation with worsening of symptoms towards the end of the day, and lower limb onset. It is important to recognize this disorder because it responds dramatically to small dose L-dopa.<sup>35</sup> Early warning signs of CP include developmental delay, toe walking, persistent fisting, microcephaly, epilepsy, irritability, poor sucking, handedness before 2 years of age (indicating hemiparesis), and scissoring of the lower limbs.<sup>33</sup> In addition, persistence of primitive reflexes can be an early indicator. A multidisciplinary evaluation is recommended and may necessitate input from physiotherapy, occupational therapy, ophthalmology, audiology, orthopedics, radiology, neurology, genetics, developmental pediatrics, and social services. Metabolic and chromosomal analyses are not recommended routinely, but are indicated if the child has dysmorphic features, a family history of delay, or consanguinity. Brain CT may be abnormal in 63% to 73% of CP cases.<sup>30</sup> Brain MRI is more sensitive than CT, particularly in delineating the extent of white matter changes. If available, it should be obtained in preference to CT.<sup>30</sup> Once the diagnosis of CP is established, communicating such news to the parents is often both difficult and emotionally unwelcome.<sup>36</sup> In addition, most physicians do not feel comfortable dealing with children with neurological disorders such as CP.<sup>37</sup> At the same time, it is important that the transfer of such information is done well as the manner in which neurological bad news is conveyed to parents can significantly influence their emotions, beliefs, and attitudes towards the child, the medical staff, and the future.<sup>36</sup> Most families find the attitude of the newsgiver, combined with the clarity of the message and the newsgiver's knowledge to answer questions as the most important aspects of giving the news.

### Management

The primary care physician should provide anticipatory guidance, immunizations, and developmental surveillance. Additionally, the child's respiratory status should be carefully assessed, as bronchopulmonary dysplasia, reactive airway disease, aspiration, and recurrent chest infections are not uncommon.<sup>23</sup> All routine immunizations should be provided, including pertussis vaccine, even if the child has epilepsy. Progressive uncontrolled epilepsy indicates DT rather than DPT vaccine. Annual influenza vaccination should be provided for those with recurrent or chronic respiratory illnesses. Pneumococcal im-

munization is recommended for those with chronic or recurrent pulmonary illnesses, and for those at risk for infection with antibiotic resistant organisms, such as children in long-term care facilities and residential settings.<sup>28</sup>

Specific treatment options for children with CP include physical and occupational therapy, drug treatments for spasticity (local, intrathecal, systemic), and orthopedic and neurosurgical interventions. Most patients require combinations of these therapies, but physical therapy is always essential. Early institution of physical, occupational, and speech therapies are essential for proper developmental progress.<sup>38</sup> Therapeutic challenges include formulating an individualized treatment plan that is functional, goal-oriented, time-limited, and cost-effective. This treatment plan should be team delivered and hospital-home-rehabilitation center-based according to the needs of each child. The basic treatment goals include parent education, facilitation of normal motor development and function, prevention of secondary complications such as deformities and disabilities and improvement of functional acquisition, community integration, and family adjustment.<sup>39</sup> Thus, emphasis has shifted from a strict focus on impairments to a broader focus on the function of the child.<sup>40</sup> The key participants on any multidisciplinary treatment team are the child and family as well as physical and occupational therapists.<sup>41</sup> Physical therapists focus on gross motor skills, including sitting, standing, walking, wheelchair mobility, transfers, and community mobility. Wheelchairs can allow the children to keep up with peers in social, educational, and recreational activities and to develop independence.<sup>23</sup> Power wheelchairs are appropriate for children who lack the strength or coordination to operate a manual chair, but demonstrate the cognitive skills necessary for safe navigation.<sup>42</sup> Occupational therapists address the visual and fine motor skills that enable coordinated functions of activities of daily living such as dressing, toileting, eating, bathing, and writing.<sup>43</sup> Forced use or constraint-induced movement therapy can be an effective technique to increase the use of the affected arm in hemiplegic CP. Restraining the stronger arm forces the weaker arm to become more functional. Orthotic interventions are aimed at the prevention and/or correction of deformities, provision of support, facilitation of skill development, and improvement of gait.<sup>44</sup> The most common orthoses used is ankle-foot orthoses (AFO), designed to hold the heel and forefoot in optimal biomechanical position. The multidisciplinary team also includes

subspecialists, social workers, nutritionists, and educators, as indicated by the individual needs of the children and their families. This team setting allows for the most effective care delivery system.<sup>38</sup> There are a number of therapeutic interventions that have no scientific literature supporting their use in CP including patterning, conductive education, and hyperbaric oxygen therapy.<sup>45</sup> It is important to emphasize that each child with CP should have the right to comprehensive management, medical education, and environmental modifications that would improve their quality of life. Several Internet resources for physicians and parents dealing with children with CP are summarized in Table 1.

### Management of Spasticity

Spasticity often generates widespread and debilitating consequences for many children with CP including pain, spasm and subsequent contractures. While spasticity need not be treated in every case, physicians today have a wide variety of treatment options. However, tone reduction is indicated only if spasticity interferes with some level of function, positioning, care or comfort. Familiarity with the strengths and weaknesses of each treatment option is an important aspect of clinical decision-making. The impact of spasticity on function must be assessed as children may rely on lower limb extensor tone for stance and ambulation. Spasticity management therefore must be goal-specific, such as to assist with mobility, reduce or prevent contractures, improve positioning and hygiene, and provide comfort. Each member of the child's multidisciplinary team, including the parents, should participate in treatment planning and serial evaluations.<sup>46</sup> In some centers, spasticity clinics exist where a pediatric neurologist, clinical neurophysiologist, orthopedic surgeon, physiotherapist, and occupational therapist assess each child. Team assessment identifies each child's strengths and deficits, sets the goals with the family, and develops a comprehensive problem list. Objective spasticity measures (rating scales) help in documenting baseline deficits, progress, and response to various therapeutic modalities.<sup>47</sup> These rating scales are summarized in Table 2.

Systemic treatments for spasticity include baclofen, diazepam, dantrolene, and tizanidine, alone or in combinations. Baclofen is the most commonly used oral medication in children with generalized spasticity. Spasticity results from an inadequate release of gamma-aminobutyric acid (GABA), an

inhibitory neurotransmitter in the central nervous system. Baclofen is a structural GABA analog enhancing presynaptic inhibition.<sup>48</sup> It crosses the blood-brain barrier poorly. Therefore, high doses may be necessary to achieve clinical response. Side effects include fatigue, irritability, hypotension, drooling, impaired memory and attention, and a lowered seizure threshold. Slow drug titration may minimize these side effects. Abrupt withdrawal of baclofen results in rebound spasticity, irritability, and subsequently fever, hallucinations, and seizures.<sup>49</sup> Benzodiazepines, including diazepam, clonazepam, and clobazam, are also useful for generalized spasticity. They increase presynaptic neuronal inhibition through GABA pathways.<sup>50</sup> Sedation and tolerance are the most common adverse effects. Dantrolene exerts its action directly at the muscular level by inhibiting calcium release from sarcoplasmic reticulum and thereby uncoupling excitation and contraction.<sup>51</sup> Muscle weakness, hepatotoxicity, and fatigue are the main side effects, making it a less favorable option. Tizanidine is an alpha-2 adrenergic agonist that hyperpolarizes motoneurons and decreases the release of excitatory amino acids. Side effects include nausea, vomiting, hypotension, sedation, and hepatotoxicity.<sup>49</sup> Children with spasticity that are refractory or intolerant to oral medications may be candidates for intrathecal baclofen therapy. After a favorable response to an initial intrathecal test dose, baclofen is provided via a programmable, refillable pump, surgically implanted into a subcutaneous abdominal pocket. The pump is connected to a catheter system that delivers a continuous infusion of baclofen into the spinal canal with significant reduction in limb tone.<sup>48</sup> Complications are related to the medication or mechanical pump failure. Overdose typically caused by programming errors, leads to somnolence, hypotonia, and respiratory depression and may progress to loss of consciousness and respiratory failure.<sup>52</sup> Withdrawal can be life threatening, with severe hypertonicity progressing to seizures, hyperthermia, rhabdomyolysis, and multiorgan failure.<sup>53</sup>

Spasticity can be focal, or unequally distributed in the extremities. In such instances, botulinum toxin injections can be used before any surgical considerations.<sup>54</sup> Botulinum toxin blocks the release of acetylcholine at the neuromuscular junction with an onset of action of 3 to 10 days and an average therapeutic duration of 3 to 6 months.<sup>54</sup> With ongoing active physiotherapy, longer benefits from the injections can occur. Side effects are rare and include transient local pain, fever, and muscle weakness. The

**Table 1.** Internet resources for professionals and parents dealing with cerebral palsy.

Organization	Web site
National Library Service for the Blind and Physically Handicapped	<a href="http://www.loc.gov/nls">www.loc.gov/nls</a>
Children's Hemiplegia and Stroke Association	<a href="http://www.chasa.org">www.chasa.org</a>
United Cerebral Palsy Associations	<a href="http://www.ucpa.org">www.ucpa.org</a>
CP Parent Home Page	<a href="http://www.cpparent.org">www.cpparent.org</a>
The Cerebral Palsy Network	<a href="http://www.thecpnetwork.netfirms.com">www.thecpnetwork.netfirms.com</a>
National Parent Network on Disabilities	<a href="http://www.npnd.org">www.npnd.org</a>
Special Needs Advocate for Parents	<a href="http://www.snapinfo.org">www.snapinfo.org</a>
National Parent-to-Parent Network	<a href="http://www.netnet.net/mums">www.netnet.net/mums</a>
Disabled Sports	<a href="http://www.dsusa@dsusa.org">www.dsusa@dsusa.org</a>
Special Olympics International	<a href="http://www.specialolympics.org">www.specialolympics.org</a>
National Center on Accessibility	<a href="http://www.Indiana.edu">www.Indiana.edu</a>
Disability and Rehabilitation Research	<a href="http://www.resna.org">www.resna.org</a>
National Information Center for Children and Youth with Disabilities	<a href="http://www.nichy.org">www.nichy.org</a>
National Organization on Disability	<a href="http://www.nod.org">www.nod.org</a>
National Association of Developmental Disabilities Councils	<a href="http://www.naddc.org">www.naddc.org</a>
Disability Rights Education and Defense Fund	<a href="http://www.dredf.org">www.dredf.org</a>

drug is not useful if fixed contractures are present. Orthopedic procedures are best left as a last resort for children with severe spasticity and/or fixed contractures or deformities. Tendon lengthening procedures are used to reduce abnormal muscle activity. The timing of these procedures is critical and best planned after the development of a mature gait pattern (5-8 years of age). Rapid growth, postural maturation, and physiologic ligamentous tightening during the first few years of life contraindicate these procedures in the younger child. A lengthened muscle is also weakened, and postoperative rehabilitation is essential. Selective dorsal rhizotomy is a neurosurgical procedure that reduces lower limb spasticity.<sup>55</sup> It involves intraoperative electromyographic monitoring to identify the sensory rootlets from L2 to S2, which, when stimulated, result in abnormal motor responses. Approximately 50% of the stimulated rootlets are cut.<sup>55</sup> The ideal candidate for this procedure is the cooperative, motivated child with spastic diplegic CP who demonstrates good strength, balance, and range of motion in the lower limbs. The procedure reduces lower limb spasticity and improves joint range of motion, and gait.

**Table 2.** Objective spasticity measures (rating scales).

<b>1- Percent of Function Scale</b>	
Estimate the amount of function you had at your best. Rate as if 0% represents fully disabled with no functional ability and 100% represents normal functional ability	
No Function	Normal Function
0%—5—10—15—20—25—30—35—40—45—50—55—60—65—70—75—80—85—90—95—100%	
<b>2- Disability Scale</b>	
Use 0.5 increments to describe the patient's disability (e.g., 0.5, 1.5 etc)	
0	Absent
1	Mild, barely noticeable spasm, tremor, pain without functional impairment
2	Mild to moderate, spasm, tremor, pain with minimal functional impairment
3	Moderate spasm, tremor, pain with moderate functional impairment
4	Severe and disabling spasm, tremor, pain
<b>3- Spasm Frequency Scale</b>	
Number of spasms in the last 24 hours in affected muscles or extremity?	
0	No spasm
1	One spasm or less per day
2	Between 1-5 spasms / day
3	Between 5-9 spasms / day
4	Ten or more spasms / day
<b>4- Global Pain Scale</b>	
Rate the total amount of pain you have had in the last 24 hours	
No Pain	Maximum Pain
0%—5—10—15—20—25—30—35—40—45—50—55—60—65—70—75—80—85—90—95—100%	
<b>5- Modified Ashworth Scale</b>	
0	No increase in muscle tone
1	Slight increase in muscle tone (a catch and release or minimal resistance)
1+	A catch followed by resistance throughout the movement
2	Marked increase in muscle tone through most ROM, but the part is easily moved
3	Considerable increase in muscle tone, passive movement is difficult
4	Affected part is rigid in flexion, extension, abduction or adduction
<b>6- Adductor Tone Rating Scale</b>	
0	No increase in tone
1	Increased tone, hips easily abducted to 45° by one person
2	Hips abducted to 45° by one person with mild effort
3	Hips abducted to 45° by one person with moderate effort
4	Two people required to abduct the hips to 45°



## Prognosis

In general, children have an enhanced capacity for brain plasticity, resulting in a capacity to recover and improve from brain insults.<sup>56</sup> There are many possible theories and mechanisms for this brain plasticity. In simple terms, it implies that normal and less damaged areas of the brain have the ability to develop and mature with time to result in developmental progression and motor improvements. A common question asked by parents is whether the child will be able to walk independently. The ability to sit independently at 2 years of age is predictive of future ambulation. Most children with hemiplegic CP will be able to ambulate independently. Regarding life expectancy and mortality rates in children with CP, the type and severity of disability and feeding skills are major determinants.<sup>23</sup> Those who require NG feeding during the first year of life have a 5-times greater mortality rate than children with oral feeding. Overall, the probability of reaching the age of 20 years in a child with severe CP is 50%.<sup>57</sup> Respiratory infections, aspiration, and epilepsy are leading causes of death. Newer therapeutic

advances and the degree of parental care have a strong influence on the length of survival of these children.

## Conclusions

CP is a chronic motor disorder that various efforts failed to prevent its occurrence. In most cases, the cause is unknown and prematurity remains the commonest risk factor. Children with CP suffer from multiple problems and potential disabilities such as mental retardation, epilepsy, feeding difficulties, vision, and hearing impairments. Screening for these conditions should be part of the initial assessment. The child with CP is best cared for with an individualized treatment plan that provides a combination of interventions. This requires the provision of a number of family centered services. Management is not curative; however, if provided optimally it can improve the quality of life of these children and their families. Physicians, in cooperation with the child, family, and members of a multidisciplinary team, can coordinate a complex care system to the maximal benefit of each child.

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