CEREBRAL PALSY-PG DISCUSSION 2014

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KEY POINTS

- Cerebral Palsy describes a group of disorders of the development of movement and posture, causing activity limitations, that are attributed to non-progressive disturbances that occurred in the developing fetal or infant brain. The motor Disorders of cerebral palsy are often accompanied by disturbances of sensation, cognition, communication perception and/or by a seizure disorder
- The etiology of cerebral palsy can be thought of using the four P's: **prenatal**, **perinatal,postnatal, and prematurity**. Common prenatal, perinatal, and postnatal causes of cerebral palsy include

Perinatal Brain Injury
Hypoxia-ischemia
Neonatal stroke (arterial and venous)
Traumatic brain injury
Intracranial hemorrhage
Brain Injury Related to Prematurity
Periventricular leukomalacia
Intraventricular hemorrhage
Developmental Abnormality
Brain malformation
Genetic abnormality
Metabolic abnormality
Postnatal Brain Injury
Kernicterus
Central nervous system infection
Prenatal Risk Factors
Maternal chorioamnionitis
Intrauterine growth retardation
Toxin exposure
Congenital TORCH infections

- **CP is the most common and costly form of chronic motor disability** that begins in childhood, and recent data from the Centers for Disease Control and Prevention indicate that the incidence is 3.6/1000 with a male/female ratio of 1.4/1. The Collaborative Perinatal Project (CPP), in which approximately 45,000 children were regularly monitored from in utero to the age of 7 yr, found that most children with CP had been born at term with uncomplicated labors and deliveries.
- **In 80% of cases, features were identified pointing to antenatal factors causing abnormal brain development**. A substantial number of children with CP had congenital anomalies external to the central nervous system (CNS).
- ***Fewer than 10% of children with CP had evidence of intrapartum asphyxia***. Intrauterine exposure to maternal infection (chorioamnionitis, inflammation of placental membranes, umbilical cord inflammation, foul-smelling amniotic fluid, maternal sepsis, temperature >38°C during labor, urinary tract infection) was associated with a significant increase in the risk of CP in normal birthweight infants. Elevated levels of inflammatory cytokines have been reported in heelstick blood collected at birth from children who later were identified with CP.
- **Genetic factors may contribute to the inflammatory cytokine response, and a functional polymorphism in the interleukin-6** gene has recently been associated with a higher rate of CP in term infants.
- the gestational age at birth-adjusted prevalence of CP among 2 yr old former premature infants born at 20-27 wk of gestation has decreased over the past decade. The major lesions that contribute to CP in this group are **intracerebral hemorrhage** and **periventricular leukomalacia** (PVL). Although the incidence of intracerebral hemorrhage has declined significantly, PVL remains a major problem. PVL reflects the enhanced vulnerability of immature oligodendroglia in premature infants to oxidative stress caused by ischemia or infectious/inflammatory insults.
- **White matter abnormalities (loss of volume of periventricular white matter, extent of cystic changes, ventricular dilatation, thinning of the corpus callosum) present on MRI at 40 wk of gestational age among former preterm infants are a predictor of later CP.**
- In agreement with earlier data, antenatal infection was strongly associated with CP and 39.5% of mothers of children with CP reported having an infection during the pregnancy, with 19% having evidence of a urinary tract infection and 11.5% reporting taking antibiotics.

- **Multiple pregnancy was also associated with a higher incidence of CP** and 12% of the cases in the European CP study resulted from a multiple pregnancy, in contrast to a 1.5% incidence of multiple pregnancy in the study. Other studies have also documented a relationship between multiple births and CP, with a rate in twins that is 5-8 times greater than in singleton pregnancies and a rate in triplets that is 20-47 times greater.
- **Death of a twin in utero carries an even greater risk of CP that is 8 times** that of a pregnancy in which both twins survive and approximately 60 times the risk in a singleton pregnancy.
- **Infertility treatments are also associated with a higher rate of CP**, probably because these treatments are often associated with multiple pregnancies. Among children from multiple pregnancies, 24% were from pregnancies after infertility treatment compared with 3.4% of the singleton pregnancies in the study.
- **CP is more common and more severe in boys compared to girls and this effect is enhanced at the extremes of body weight**. Male infants with intrauterine growth retardation and a birthweight less than the 3rd percentile are 16 times more likely to have CP than males with optimal growth, and infants with weights above the 97th percentile are 4 times more likely to have CP.

How to dx cp?

- Despite advances in technology, cerebral palsy remains a clinical diagnosis, and represents a continuing role for the ‘‘art’’ of medicine. The essential findings include:
- Delayed motor milestones
- Abnormal muscle tone
- Hyperreflexia
- Absence of regression or evidence of a more specific diagnosis .

These clinical findings should be present to the degree that the child appears unlikely to ‘‘outgrow’’ these findings. The potential to ‘‘outgrow’’ the manifestations ofcerebral palsy is best recognized in the preterm population, who may have transient abnormalities in tone and reflexes that may seem to interfere with motor progress, but these clinical findings resolve by 1 to 2 years of age.7 Term infants as well sometimes ‘‘outgrow’’ the manifestations of cerebral palsy,8,9 so clinical judgment is needed for determining how long to ‘‘watch’’ before labeling

- Early diagnosis of CP is aided by a history of an abnormal pregnancy, labor, delivery, or neonatal period or by the occurrence of a serious acute illness and trauma. The diagnosis is further aided by evaluat- ing the child's primitive reflexes (eg, the asymmetrical tonic neck response), postural responses (righting the head when tilted to the side), muscle tone, motor mile- stones, and neurobehavioral responsiveness. 24 Persis- tent or exaggerated primitive reflexes, a delay in the emergence of postural reactions, hyperreflexia, asym- metry, and abnormal muscle tone all suggest the pos- sibility of CP. Other early signs that suggest CP include difficulty feeding because of abnormal oral-motor pat-terns (tongue thrusting, tonic bite, oral hypersensitiv- ity), irritability, and delayed milestones such as headcontrol.

CLASSIFICATION OF CEREBRAL PALSY AND MAJOR CAUSES

MOTOR SYNDROME (APPROX % OF CP)	NEUROPATHOLOGY/MRI	MAJOR CAUSES
Spastic diplegia (35%)	Periventricular leukomalacia Periventricular cysts or scars in White matter, enlargement of ventricles, squared of posterior ventricles	Prematurity
		Ischemia
		Infection
		Endocrine/metabolic (e.g., thyroid)
Spastic quadriplegia (20%)	Periventricular leukomalacia	Ischemia, infection
	Multicystic encephalomalacia Cortical malformations	Endocrine/metabolic, genetic/developmental
Hemiplegia (25%)	Stroke: in utero or neonatal Focal infarct or cortical, subcortical damage Cortical malformations	Thrombophilic disorders
		Infection
		Genetic/developmental
		Periventricular hemorrhagic infarction
Extrapyramidal (athetoid, dyskinetic) (15%)	Asphyxia: symmetric scars in putamen and thalamus Kernicterus: scars in globus	Asphyxia
		Kernicterus
		Mitochondrial

MOTOR SYNDROME (APPROX % OF CP)	NEUROPATHOLOGY/MRI	MAJOR CAUSES
	pallidus, hippocampus Mitochondrial: scaring globus pallidus, caudate, putamen, brainstem No lesions: ? dopa-responsive dystonia	Genetic/metabolic

Which all are the mimics of cp?

Mimics of cerebral palsy

Disorder	Clue
Familial spastic paraplegia	Family history
Transient toe walking	Normal deep tendon reflexes
Muscular dystrophy	Calf hypertrophy, positive Gower's sign
Metabolic disorders	Regression, lethargy, unusual vomiting
Sjogren-Larrson	Ichthyosis
Lesch-Nyhan	Severe self-mutilation
Mitochondrial disorders	Recurrent stroke, cardiomyopathy, hypoglycemia
Genetic disorders	Multiple anomalies
Miller-Dieker	Lissencephaly
Rett Syndrome	Acquired microcephaly, hand wringing

What are the types of cp?

Box 1
Types of cerebral palsy

Spastic
- Hemiplegia (unilateral involvement)
- Diplegia (disporprotionate lower extremity involvement)
- Quadriplegia (total body involvement)

Dyskinetic
- Choreoathetoid
- Dystonic

Hypotonic

Mixed

Common Cerebral Palsy Syndromes

Cerebral palsy can be classified by the predominant type of motor abnormality – spasticity, choreoathetotic, dystonic,hypotonic, ataxic, or mixed – as well as by the distribution of limb involvement – hemiplegia, quadriplegia, or diplegia.
The most common clinical cerebral palsy syndromes are discussed next.

Spastic Hemiplegia

- Although children may manifest obvious hemiplegia in the second year of life, specific difficulties may not be observed during the first 3–5 months of life. After a perinatal stroke, an infant may be neurologically normal until the development of pathologic handedness at approximately 4–6 months of age. **For unexplained reasons, the left hemisphere (right side of the body) is affected in two-thirds of patients, and perinatal stroke is more common on the left than on the right**
- During the examination, the child exhibits impaired gross and fine motor coordination, has difficulty moving the hand quickly, and frequently is **unable to grasp small items with a pincer grasp**. **The obligate palmar grasp reflex, which usually is absent by age 6 months and frequently rudimentary after age 4 months, may remain obligate**. Weakness of the wristand forearm often is associated with limitation of range of motion of supination. The range of elbow extension may berestricted. **Attempts at reaching for objects may be accompanied by athetotic posturing with**

flexion of the wrist and hyperextension of the fingers (avoidance reaction). Facialinvolvement is unusual. Only 10 percent of affected patients,including those with extensive hemiplegia, have homonymous hemianopia []. Childrenwith hemiparesis may have a circumductive gait with a variable degree of abnormality. **Most commonly, the child walks on the toes and swings the affected leg over a nearly semicircular arc during the courseof each step**. In contrast with the leg, the affected arm usually moves less than normal and does not participate in normal reciprocal motion during ambulation. An equinovarus positioning of the foot is seen; weakness and lack of full range of motion of dorsiflexion often are present. Further evidence of upper motor neuron involvement on the hemiplegic side includes hyperreflexia of the deep tendon reflexes, ankle clonus, and extensor toe signs.

- Growth retardation of the abnormal side, usually more prominent in the distal arm and hand or distal leg and foot, may be manifest. An indication of the presence of growth impairment may be obtained when the thumb and thumbnail of the affected side are compared with their normal opposite members and found to be smaller. Growth discrepancy of the leg may result in significant difficulties during walking, leading to orthopedic problems involving the proximal leg and the lower spinal vertebrae. Although frequently overlooked, corticosensory impairment and hemineglect of the affected side are common. Examination for the integrity of stereognosis and graphesthesia usually reveals varying degrees of compromise [Brown et al., 1987; Skatvedt, 1960].

Spastic Quadriplegia

- Little [1861] first described cerebral palsy. He used the term spastic rigidity in place of the modern term spasticity. As part of his original treatise, he wrote: “Spastic cerebral palsy has the characteristics of upper motor unit involvement, such as hyperreflexia and increased tone, often with ankle clonus, crossed adductor reflexes, and extensor toe signs. Both lower extremities are more or less generally involved. Sometimes the affection of one limb only is observed by the parent, but examination usually shows a smaller degree of affection in the limb supposed to be sound. The contraction in the hips, knees, and ankles is often considerable. The flexors and adductors of thighs, the flexors of knees, and the gastrocnemii,preponderate. In most cases, after a time, owing to structuralshortening of the muscles and of the articular ligaments, and perhaps to some changes of formof articular surface, the thighscannot be completely abducted or extended, the knees cannot be straightened, nor can the heels be properly applied to the ground. The upper extremities are

sometimes held down by preponderating action of pectorals, teres major and teres minor, and latissimus dorsi; the elbows are semiflexed, the wrists partially flexed, pronated, and the fingers incapable of perfect voluntary direction. Sometimes the upper extremities appear unaffected with spasm or want of volition, sometimes a mere awkwardness in using them exists."

- **Spastic quadriplegia is characterized by a generalized increase in muscle tone. The legs are involved more than the arms, and paucity of limb movement is characteristic. Opisthotonic posturing may be evident in early infancy and may persist through the first year of life**. Movement of the head often initiates forced extension of the arms and legs, resulting in a position similar to that in decerebrate rigidity. Accompanying supranuclear bulbarpalsy, the result of bilateral corticobulbar tract impairment,may produce difficulties with swallowing and articulation. The incoordination of the oropharyngeal muscles may predispose the patient to recurrent pneumonia during the first years of life.
- **Neurologic examination demonstrates marked spasticity and accompanying signs of corticospinal tract involvement, including hyperactive deep tendon reflexes, ankle clonus, and extensor toe signs. Weakness of dorsiflexion of the feet, associatedwith equinovarus deformities, is common**. Marked spasticity of the hip muscles may lead to subluxation of the femur and associated acetabular pathologic conditions. **Radiographs may be necessary to exclude the abnormal positioning of the head of the femur**. Flexion contractures of the wrists and elbows of various degrees and spasticity of the arm muscles are readily apparent. Ophthalmologic evaluation of children with spastic quadriplegia more commonly reveals visual impairment in thesechildren than in children with athetoid cerebral palsy [Preakey et al., 1974].
- **The incidence of auditory, visual, motor, and learning disability is much higher in children with spastic quadriplegia than in children with spastic** hemiplegia, spastic diplegia, and ataxic cerebral palsy [Robinson, 1973; Shevell et al., 2009].

Spastic Diplegia

- Spastic diplegia is characterized by bilateral leg involvement and, Commonly, some degree of upper extremity impairment. Preterm infants are particularly prone to spastic diplegia. **Approximately 80 percent of preterm infants who manifest motor abnormalities have spastic diplegia** [McDonald, 1963].In recent years, the survival of very small preterm infants has resulted in a larger group of more severely neurologically impaired survivors [Hagberg et al., 1989a, b].

- **Diffusion tensor imaging in preterm infants has demonstrated disruption of thalamocortical connections, as well as descending Corticospinal pathways** [Hoon et al., 2009].Some infants with spastic diplegia manifest ataxia after further maturation. These infants have a great increase of tone of the leg muscles and accompanying difficulties in coordination andstrength. Impairment may be asymmetric.
- **When a small child is held in the vertical position by the examiner and the plantar surfaces of the feet are lightly bounced on the examining table, adduction of the legs (scissoring) and obligatory extension (extensor thrust) are seen. The feet also are kept in an equinovarus posture. Further examination reveals weakness of dorsiflexion of the feet**.
- In older children, this same spasticity causes them to toe-walk. As expected, signs of upper motor unit involvement are easily demonstrable in the legs (e.g., hyperactive deep tendon reflexes, bilateral ankle clonus, extensor toe signs).
- **Striking spasticity of the hip muscles may lead to subluxation of the femur and associated acetabular pathologic conditions andfurther restriction of motion.** Radiographs may be necessaryto exclude the abnormal positioning of the femoral head.The arms may be affected but usually only to a mild degree.The child may hold the arms in unusual fixed postures, either extended or flexed during walking, and may have clumsy, reciprocating, swinging arm movements or hold both arms flexed at the elbows.
- Affected children may extend their arms, pronate their hands, and clench their fists during running. Associated athetosis makes this latter posturing more likely. Vasomotor instability, often manifested by cold extremities and variable and sometimes unpredictable patterns of sweating, may prove troublesome for the patient.
- **After a variable period, usually 18 months to 2 years inchildren with moderate involvement, spasticity is increasingly accompanied by contractures that maintain the hips in flexion, knees in flexion, and the feet** in an equinovarus position. For reasons that are unclear – either disuse or probably hemispheric (parietal) lobe dysfunction – marked retardation of linear growth of the legs may be a feature.

Extrapyramidal Cerebral Palsy

- **Extrapyramidal (dyskinetic) cerebral palsy can be divided arbitrarily into two primary clinical subtypes – choreoathetotic and dystonic**. Patients are unable to perform meaningful movements smoothly because of interfering movements and involvement of inappropriate agonist and

antagonist muscles Extrapyramidal cerebral palsy involves defects of posture and involuntary movement (e.g., athetosis, ballismus, chorea, dystonia); increased tone usually is associated with these conditions and is of the “lead pipe” or rigid variety. Children with this form of cerebral palsy have more severe cognitive and motor impairments than children withbilateral spastic quadriplegia [Himmelmann et al., 2009].

CHOREOATHETOTIC CEREBRAL PALSY

- **Choreoathetotic cerebral palsy is characterized by large amplitude,involuntary movements**. The most obvious and dominating movement component is athetosis. Chorea is present in variable degree. Tremor, myoclonus, and even some element of dystonia also may be evident.
- **Athetosis usually involves the distal limbs. Athetosis results in slow, writhing involuntary movements**. Other common features are finger and toe extension and rotation of the limb and its long axis. The resultant pattern of these movements culminates in bizarre transient positions of the limbs.
- Chorea may involve the face, limbs, and rarely the trunk. The choreiformmovements can be characterized as asymmetric, fleeting, incoordinated,involuntary contractions of individual muscle groups. Athetotic posturing may be evident in the first year of life when the child begins to reach for objects. The movements,as generally is true of most involuntary movements, are not present during sleep.
- Movements are more prominent during stress or illness, and their intensity changes from day to day. As expected from the pathologic findings, evidence of upper motor neuron unit impairment (e.g., hyperactive deep tendon reflexes, ankle clonus, positive extensor toe signs), as well as seizures, spasticity, and mental retardation, may be present.
- Children with choreoathetosis may have marked difficulty with speech, which is characterized by great variability in rate and explosive changes in volume.
- Ballismus, a movement disorder in which the arms and legs are violently flung about, may be an extreme form of choreoathetotic cerebral palsy. Most of the activity takes place at the shoulders and hips. Although patients with ballismus are said to have a shortened life expectancy and do not survive beyond the second decade, few data are available, and clinicopathologic correlation is undefined.

DYSTONIC CEREBRAL PALSY

- The dystonic form of cerebral palsy is uncommon. The extrapyramidal form of cerebral palsy is often, but not always, preceded by hypoxic-ischemic brain injury or kernicterus.Requirement for respiratory support and hypoxic-

ischemic encephalopathy at birth usually are documented in the patient history. The dystonic movements are not unlike those in other conditions associated with dystonia. **The trunk muscles and proximal portions of the limbs are predominantly affected.Movements may be slow and persistent, particularly of the head and neck, which may be pulled to one side or the other, or retrocollis may be present**. At times, the movements may consist of rapid and repetitive retractions of the head. The trunk may be literally twisted into many fixed positions that may appear bizarre.

- **Corticospinal tract findings may be evident in children who have choreoathetosis. On pathologic examination, the brains of these children may reveal large** areas of patchy necrosis of the cortical laminar pattern, venous congestion in the cortex, ventricular dilatation, and accompanying white-matter loss that may be related to demyelination and central necrosis.Fibrosis in the meninges also may be present. Both cortexand basal ganglia may be jointly involved in these patients.
- Cortical lesions associated with necrosis in areas adjacent to the ventricles may be the result of occlusion of the vein of Galen. Obstruction of this major vessel triggers a chain of events, including rupture of blood vessels, primarily veins, with ensuing multiple hemorrhages in the areas served by the branches of the internal cerebral veins. The hemorrhages result in subependymal necrosis and subsequent pathologic dilatation of the lateral ventricles and associated atrophy of the basal ganglia. If obstruction is extremely widespread, the internal capsule may be involved, and further symptoms and signs of corticospinal tract involvement arise.
- In one report, patients with severe **athetoid cerebral palsy originating perinatally were divided into two groups neuropathologically: the "globo-Luysian group" and the "thalamoputaminal group."** The major abnormal sites in the globo-Luysian group were the pallidum and subthalamicnucleus, and in the thalamoputaminal group, the thalamus and putamen. **The causative pathologic condition in the globo-Luysian group was primarily severe perinatal jaundice, and the cause in the thalamoputaminal group was predominantly neonatal asphyxia. The patients in the thalamoputaminalgroup demonstrated lower mental ability and suffered from more intractable convulsions than those in the globo- Luysian group. In the globo-Luysian group, rigidity and spasticity were frequently demonstrated, with fluctuation of athetoid movements, whereas in the thalamoputaminal group, various abnormalities of muscle tone and rather restricted athetosis were observed** [Hayashi et al., 1991]. MRI studies in 22 children with athetotic

cerebral palsy frequently revealed high-intensity areas in the thalamus and putamen in T2-weighted images. Of 16 children with known perinatal asphyxia, 14 had lesions in the basal ganglia, thalamus, and/or cerebral white matter. MRI findings were normal in 7 of the 22 children [Yokochi et al., 1991].

Hypotonic (Atonic) Cerebral Palsy

- Infants with hypotonic cerebral palsy almost always have associated leg weakness. Although hypotonic, the arms maymanifest near-normal strength and coordination. In the past,this combination of clinical findings led to the use of the term atonic diplegia to describe such children.
- **Diagnosis is difficult because of the plethora of diagnostic possibilities. Most children with generalized hypotonia have so-called central hypotonia, resulting from inadequate control of the motor pathways and subsequent disruption of gamma loop function**. **Others, with absent or hypoactive deep tendon reflexes, may have involvement of the lower motor neuron unit (i.e., anterior horn cell, peripheral nerve, neuromuscular junction, muscle). Extrapyramidal (choreoathetotic and dystonic) cerebral palsy may be preceded by a hypotonic phase**.
- Atonic cerebral palsy is relatively uncommon, compared with other forms of cerebral palsy; it often is associated with slow attainment of motor ilestones and the presence of normal or hyperactive deep tendon reflexes. Children with atonic cerebral palsy, when suspended while held under the arms, flex both legs at the hips (Fo¨rster's sign). Although, in the past, it has been thought that muscle tone almost always increases with maturation in this form of cerebral palsy [Ingram, 1964], experience has taught that in a sizable number of cases, spasticity does not develop, but the child remains hypotonic.
- The causes leading to this condition and the associated anatomic location of brain involvement are unknown. It is through their effect on the gamma motor neuron that portions of the central nervous system (e.g., motor cortex, thalamus, basal ganglia, vestibular nuclei, reticular formation, cerebellum)modify tone, with ensuing hypotonia.

Ataxic Cerebral Palsy

- The least common form of cerebral palsy is the ataxic form. It sometimes coexists with spastic diplegia [Hagberg et al., 1975].**This form usually is associated with other motor abnormalities; however, the diagnosis is applied only when the predominant manifestation is cerebellar dysfunction. Patients with ataxic cerebral palsy may have impairment of intellectual ability**,but they are rarely grossly delayed [Clement et al.,

1984].Motor difficulties often are not apparent until late in the first year of life.

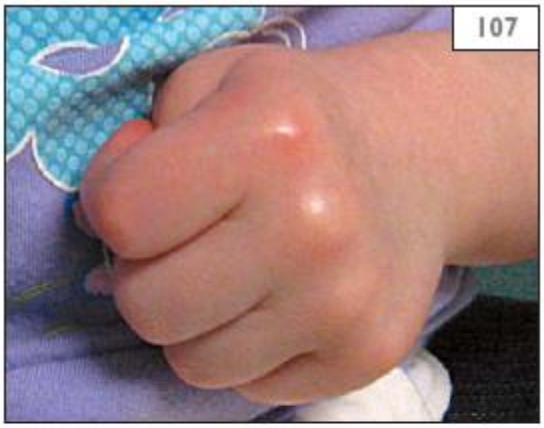
- **Early manifestations include hypotonia, truncal ataxia with sitting, dysmetria, and gross incoordination. The motor involvement results in delayed attainment of motor skills; independent walking may not occur until age 3 or 4 years, and then may be performed only with great difficulty andfrequent falling**. Compromise of writing skills and other skillsthat demand good fine motor coordination often adverselyaffects educational endeavors.
- Examination often reveals nystagmus, dysmetria, hypotonia,and a wide-based gait. The result on Romberg testing with the eyes open is positive. Likely sites of involvement are the cerebellum and adjacent brainstem.Because of the large number of conditions associated withataxia, the clinician must exclude conditions in which ataxia predominates in early childhood Ataxia,especially if accompanied by mental retardation, may not be properly included among cerebral palsy conditions but may be the result of one of many inherited conditions [
- The pathologic features of ataxic cerebral palsy are poorly defined and inconstant. Discussion of these features is confounded by the fact that total absence of the vermis may not give rise to cerebellar symptoms in certain congenital conditions, whereas aplasia of the vermis may be associated withnonprogressive ataxia Cerebellar hemispheric lesions may or may not be present in patients with ataxic cerebral palsy. The lack of correlation of evident structural changes with functional impairment is emphasized by CT studies. In one report, CT evaluation of patients with ataxic cerebral palsy revealed that the posterior fossa was normal in 38 percent and abnormal in 28 percent.
- By contrast, the cerebral hemispheres were abnormal in 55 percent of the patients [Miller and Cala, 1989].
- **Mixed Cerebral Palsy**

- Mixed cerebral palsy includes manifestations of both spastic and extrapyramidal types; often an ataxic component is present.Patients with predominantly spastic quadriplegia may have a mild to marked degree of choreoathetosis.
- Conversely, frequently patients in whom choreoathetosis predominates also may manifest upper motor neuron unit involvement.These patterns of motor impairment are the result of compromise of large areas of the brain, with sequelae of basal ganglia, cortex, and subcortical disruption..

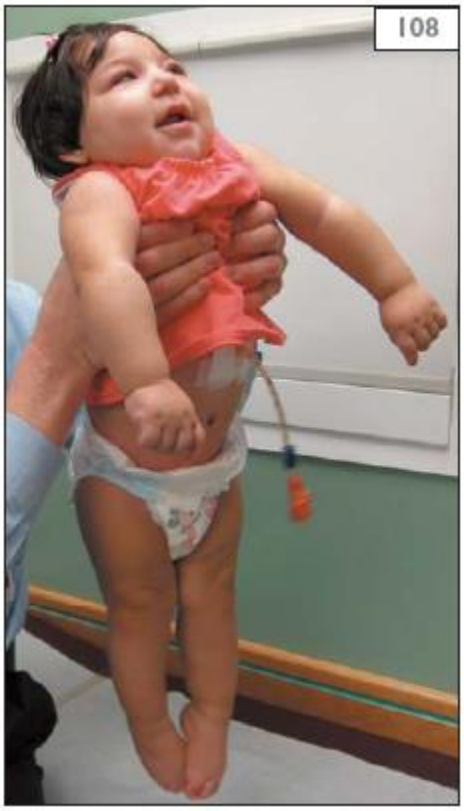
General Prognosis for Motor Function

- A number of factors affect the prognosis of the child with cerebral palsy: the clinical type of cerebral palsy; the degree of delay in meeting milestones noted at evaluation; the pathologic reflexes present; and the degree of associated deficits in intelligence, sensation, and emotional adjustment [Sala and Grant, 1995]. Cognitive level is difficult to assess in the youngchild with motor impairments but can be gauged even in the severely affected child [McCarty et al., 1986]. It is necessary to consider the cognitive level, despite the challenges posed in assessment, because the level of mental function may be the factor that really determines the quality of life the child will enjoy.
- **Children with hemiplegia but with no other major problems almost always walk by the age of approximately 2 years;**some benefit is gained from use of a short leg brace, often needed only for temporary assistance. The presence of a small hand on the hemiplegic side, with a thumbnail that isnarrower than that of the other thumb, may be associated with sensory dysfunction of parietal origin, and the sensory defect may limit the development of fine motor skills in that hand. **About 25 percent of children with hemiplegias have hemianopsia; recognition of this deficit allows the clinician to advise placing the affected child in an area of the classroom**that maximizes useful vision. Because most daily activities canbe accomplished with only one hand, using the affected hand only as a "helper," with small adaptations, such as shoes that do not require lacing and tying, hemiplegic children of reasonable intelligence can be expected to achieve independence in daily living. Seizures may be a problem in children with hemiplegia.
- **More than 50 percent of children with spastic diplegia learn to walk, commonly by the age of approximately 3 years, but gait often is abnormal**, and some children require assistance devices, such as crutches. Hand activities commonly are involved to some degree, although the impairment may be subtle. Abnormalities of extraocular movement are relatively common.
- **Of children with spastic quadriplegia, 25 percent require total care; approximately 33 percent walk, usually after the age of 3 years**. Intellectual function often is the most life-limiting concomitant problem, and involvement of the bulbar musculature may add further difficulties. Marked truncal hypotonia, with pathologic reflexes or persisting rigidity, is associated with an unfavorable outlook. A majority of such children have grave intellectual limitations.
- The prognosis for walking has been evaluated recently in large populations of children with cerebral palsy. The validated GMFCS can help predict

whether the child will eventually achieve the ability to walk 10 steps unsupported [Rosenbaum et al., 2002]. Furthermore, a child's motor function at age 2 years can be used to evaluate the prognosis for future ambulation at three levels of competency, with the help of ambulation chartsreflecting the combined experience with more than 5000 children with cerebral palsy



107 Cortical thumb indicating upper motor neuron dysfunction.



108 Scissoring (crossing) of the legs in a child with cerebral palsy and spastic quadriparesis.

Table 2
Gross motor functional classification system

Level	Function
I	Ambulatory in all settings
II	Walks without aides but has limitations in community settings
III	Walks with aides
IV	Mobility requires wheelchair or adult assist
V	Dependent for mobility

EVALUATION

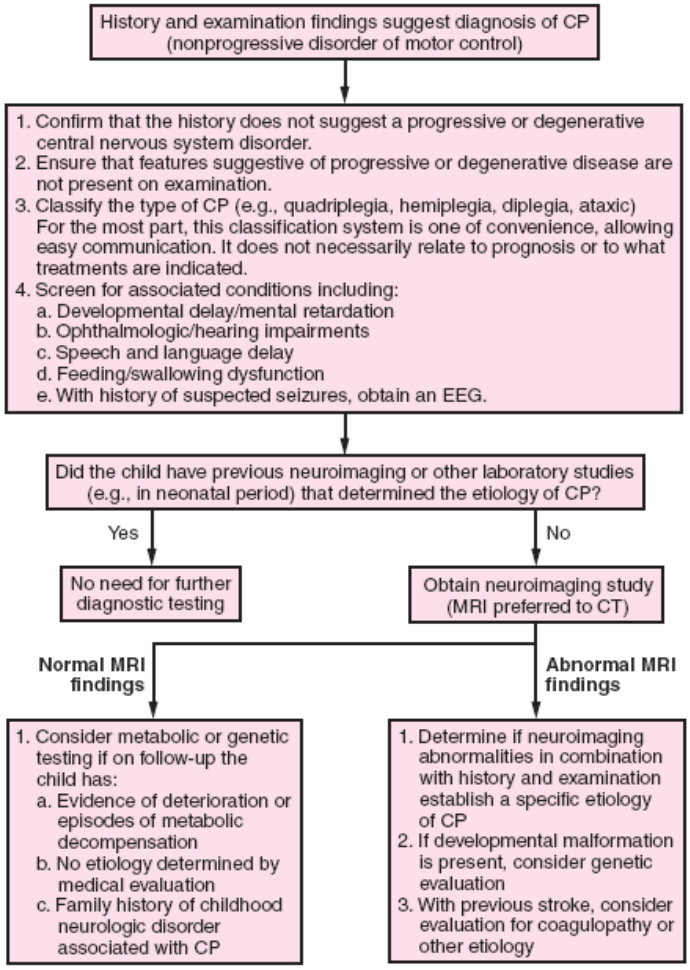


Fig. 69-1 Algorithm for the evaluation of the child with cerebral palsy (CP). Screening for associated conditions (mental retardation, vision/hearing impairments, speech and language delays, oral motor dysfunction, and epilepsy) is recommended. Neuroimaging (magnetic resonance imaging [MRI] is preferred to computed tomography [CT]) is recommended for further evaluation if the etiology has not been previously determined. In some children, additional metabolic or genetic testing may be indicated. EEG, electroencephalogram. *(From Ashwal S et al. Practice parameter: Diagnostic assessment of the child with cerebral palsy: Report of the Quality Standards Subcommittee of the American Academy of Neurology and the Practice Committee of the Child Neurology Society. Neurology 2004;62:851–863.)*

What is the management strategy?

MANAGEMENT TEAM

A large team of experts needs to work together to manage the many impairments associated with the primary neurologic lesion and the accompanying motor disorder seen in CP. The team consists of physicians; surgeons; and allied health professionals,such as physiotherapists, occupational therapists, and child development professionals, who are aware of all the needs and limitations of the child and know what all other members of the team are doing for the child.

Goal of Management

The ultimate goal is minimizing disability while promoting independence and full participation in society. All treatment efforts are directed to gain independence in activities of daily living, ability to go to school, earn a living, and a have a social life.Everyone is a child only once. The child should live a

childhood as close to normal as possible and grow up to be a happy and healthy adult.

Management strategy and rehabilitation of the child who has cerebral palsy

1. Support growth and nutrition
2. Therapy for vision: ophthalmologic therapy
Rehabilitation
Glasses
Surgery
3. Dental hygiene
4. Gastrointestinal problems
Medications for reflux
Gastrostomy
Antireflux surgery
5. Therapy for motor function
Physical therapy
Occupational therapy
Adaptive seating
Bracing
Wheeled mobility
Orthopedic surgery
Sports-recreation
6. Oromotor therapy
Chewing
Swallowing
Speech
7. Seizure prevention
8. Spasticity and dyskinesia
Medical treatment
Botulinum toxin
Intrathecal baclofen
Selective dorsal rhizotomy (SDR)

The gross motor classification system

Level	Ability
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 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 or outdoors on a level surface with an assistive mobility device. Children may climb stairs holding onto a 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 the age of 6 years or rely more on wheeled mobility at home, school, and in the community. Children may achieve self-mobility using a power wheelchair.
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. At level V, children have no means of independent mobility and are transported.

- Management between 0 and 2 years of age should include physiotherapy, infant stimulation, positioning, and parent education. Ways to decrease muscle tone should be encouraged during the age of 2 to 5 years when muscle tone becomes a problem and dyskinesias manifest themselves. Orthopedic intervention may be necessary from 5 years onward. Hygiene, seating, and care issues, in addition to preventing pain and discomfort

secondary to spasticity, become predominant during the teenage years

Table 3
Timing of rehabilitative measures

Age	Goal	Methods
Infancy	Supportive measures for prolonging and optimizing physical status and life	Nutritional support Infant stimulation and positioning
Childhood	Strategies to obtain maximum independent mobility	Medication to minimize spasticity Exercise Botulinum toxin Bracing
Preschooler	Maximum independent mobility	Minimize deformity Medication Exercise Botulinum toxin Bracing Orthopedics surgery
Adolescence	Education, vocation, and integration into the community	Schooling, sports, psychosocial support

SEIZURES

- **Approximately 30% of persons with cerebral palsy have a seizure disorde**r.30 Just like seizures in the general population, seizures in persons with cerebral palsy may come on at any age and be of any type. In other words, there is nothing ‘‘unique’’ about seizures in children with cerebral palsy. Choice of anticonvulsants, when required, is guided as usual by seizure type and side effect profile.
- **Also like other children, a child with cerebral palsy may be able to be tried off such medicines once the child has been seizure-free for at least 2 years**.31 What warrants special consideration in this population, however, is monitoring for anticonvulsant side effects, as these children may have other medical issues that could be exacerbated by side effects of anticonvulsant medications. **For example, a child with feeding problems and gastroesophageal reflux may suffer further nutritional compromise if she develops anorexia as a side effect of an anticonvulsant, such as topiramate**.32
- Likewise, children with associated learning problems may do better with some of the newer generation anticonvulsants, which appear to have less impact on cognition, although complete data are lacking.33

COGNITION/LEARNING

- **Roughly half of individuals with cerebral palsy have associated cognitive impairment, with others having overall cognitive skills in the normal range** but with specific deficits in learning or attention that could be classified as learning disabilities or attention-deficit/ hyperactivity disorder.30
- The risk of associated cognitive or learning difficulties varies somewhat with the type of cerebral palsy, with **those with quadriplegic cerebral palsy having the highest risk of cognitive impairment and those with hemiplegic cerebral palsy the lowest.** This makes sense given that children with spastic quadriplegia have had a more global brain insult, while those with hemiplegia have had a quite localized one. Those with other cerebral palsy subtypes have an intermediate risk of impaired cognition. What is perhaps more important for the clinician to remember is that there are clearly exceptions, and that it is crucial that persons with extensive physical involvement be given the opportunity to demonstrate their mental abilities.
- This may require the intervention of a skilled speech and language pathologist to appreciate nonverbal communication potential, and a willingness to listen to families who feel that their child understands more than he or she can demonstrate.
- **For individuals whose motor impairment hinders verbal communication, augmentative communication and other rehabilitation technologies can allow them to communicate and learn at their cognitive level**. The clinician may need to advocate forpatients with cerebral palsy to receive such technology, especially to have access to it in all settings—at home, in school, and within the community (Fig. 1).

NUTRITION AND GROWTH

- Children with cerebral palsy face multiple challenges to normal growth, and are more likely to be underweight and short compared with peers.34
- **Medical offices may lack scales for older children who cannot stand, and length measures may be difficult to reliably obtain in a child with joint contractures or a child who is in constant motion due to choreoathetosis.**
- **The latter can be addressed by using segmental measures, such as upper arm or tibial length, for which standards are available**.36 Use of such measures requires staff training, as even small errors in these measurements can result in misclassification of the child's growth status. In the past, these children's poor nutritional status was accepted as part of the condition, but it has been shown that these children can grow normally37

and benefit from being adequately nourished.34 Although being underweight is the most common nutritionalproblem among those with cerebral palsy, some with cerebral palsy are overweight.

- Excess weight in a person with cerebral palsy should be reduced to prevent respiratory compromise38 and to facilitate maximal mobility and ease of care.39 The evaluation of the underweight child with cerebral palsy begins with the diet history, which means looking at what foods and drinks are offered, how long meals take, and how the child eats.
- Children with cerebral palsy may have impaired control of oromotor musculature, impairing their ability to handle food and drink, resulting in prolonged mealtimes, increased spillage, or delayed transition to more calorically dense table foods.40 Some families restrict dairy products because of concerns about constipation or about the development of excessive mucous with milk.
- An occupationaltherapist may be able to offer suggestions and adaptations, such as improved seating and adaptive cups, to make eating and drinking more efficient.
- Interventions to increase the caloric density of the diet can help the child consume more calories and other nutrients. A registered dietitian can be very helpful in this regard, but, if this resource is not available, clinicians can recommend such strategies as adding extra butter or sauces to foods, incorporating high calorie beverages in the diet, such as milkshakes, drinkable yogurts, and commercially available complete nutrition drinks The accompanying step is a careful history and physical examination looking for medical factors that might interfere with intake.
- The possibilities are numerous and include sources of discomfort, such as constipation or hip pain, and abnormal losses, such as emesis due to gastroesophageal reflux or as a medication side effect.
- Medication side effects are common contributors to nutritional problems in this group of patients and should always be considered. It is also helpful to keep in mind uncommon presentations of common problems. For instance, practitioners may only consider the possibility of clinically significant gastroesophageal reflux when parents report chronic vomiting, but discomfort after meals, early satiety, or unexplained respiratory problems41may also be presenting signs.

SWALLOWING

- Persons with cerebral palsy are at increased risk for dysphagia, which can affect their health in several ways. **Dysphagia with aspiration may result in or contribute to recurrent pulmonary infections or reactive airway disease**. It can limit food choices and compromise the efficiency of intake,

contributing to impaired nutrition. However, just as in the discussion of gastroesophageal reflux above, dysphagia may not always present as clinicians might expect. **When documented to aspirate, most individuals with cerebral palsy do so silently, meaning they do not cough at the time of aspiration.42 Thus, practitioners must have a high index of suspicion for dysphagia andseek indirect signs and symptoms of aspiration, such as congestion or wet vocal quality with meals**. Prolonged feeding times are also associated with dysphagia.43

- **Dysphagia should also be sought in persons with recurrent pneumonia or reactive airway disease of unexplained cause or severity. Reliable assessment of swallowing coordination requires fluoroscopic visualizationof swallow during intake of barium-containing foods and beverages**. These studies are performed with the individual seated in a neutral position, and are done in conjunction with a speech pathologist or occupational therapist with feeding experience. The therapist reviews a videotape of the study before interpretation, as aspiration can be missed at the time in a busy fluoroscopic suite.
- Bedside assessments and upper gastrointestinal series studies cannot substitute for a formal videofluoroscopic study, and ultrasound assessment remains investigational.44 A thorough report will comment on risk factors for aspiration, such as residue in the valleculae and pyriform sinuses andlaryngeal penetration, as well as the presence and frequency of aspiration. It should also discuss what compensations, if any, were seen to be effective, such as thickness and rate of flow of liquids given. If no liquid consistency can be found to be safe, alternative modes of nutrition and hydration need to be considered.

GASTROINTESTINAL

- Just as control of the muscles of the limbs and trunk are affected in cerebral palsy, so is the smooth muscle of the gastrointestinal tract. Persons with **cerebral palsy have a high incidence of gastroesophageal reflux and constipation, and may have altered gut motility as well, resulting in delayed gastric emptying and intestinal dysmotility**.45 Disorders here may have an impact on many aspects of a child's life. Both gastroesophageal reflux and constipation can impair nutritional intake and be sources of pain. Reflux can contribute to acute and chronic pulmonary problems,46 and can bea source of blood loss leading to chronic anemia and to dental erosions.47**Gastroesophageal reflux in persons with cerebral palsy differs from those in the general population in incidence more than treatmen**t. Medications to decrease gastric acid production are the first-line therapies. Those that directly effect motility may alsobe helpful, but their use

is limited by their side affect profiles. Other interventions that can be considered include upright (but not slumped) position after meals, avoidance of reported dietary triggers and high osmolarity medications and supplements, andrelief of any constipation.

- Surgical treatment of gastroesophageal reflux with fundoplicationis considered in cases of clinically significant but medically refractorygastroesophageal reflux, or in conjunction with gastrostomy placement when gastroesophagealreflux is present or suspected (to be discussed further below).
- **Gastrostomy placement is contemplated in the setting of insufficient intake or excessivefeeding times despite intervention**, or in persons who, due to risk of aspiration,pose a danger to themselves if they eat and drink orally. This is an emotionally ladenstep for most families, as feeding one's child is intimately connected with the parentalrole, and the inability to meet this most basic need of the child can be seen as a failure.
- The gastrostomy is also a marker of how ''different'' the child is, and that the child'sdifferences are not going to go away. Families may need some time to make this decision,and may benefit greatly from talking with another parent who has faced a similarchoice. The ability of gastrostomy to facilitate more normal growth and to improve the child's quality of life is well documented, although its impact on family quality of life and the child's longevity is less clear.48 Finally, the circumstances under which the gastrostomy should be accompanied by a fundoplication to prevent reflux remain a matter of experience and opinion, with no controlled studies to provide guidance.49
- Children should be followed closely after gastrostomy placement to address issues offeeding intolerance promptly when they arise, and to prevent excessive weight gain.Constipation is estimated to occur in 80% of individuals with cerebral palsy. The cause is multifactorial and for any given child may include low fluid or fiber intake, decreased activity, and difficulty sitting comfortably on the toilet. There are likely skeletal muscle contributors as well, such as difficulty relaxing the pelvic floor musculature and with pushing. For many, intestinal motility is also abnormal.50 Constipation for these individuals is more than a nuisance. It can adversely affect appetite, continence, and comfort. Just as in typically developing individuals, it can predispose to urinary tract infections.51
- There are many potential interventions for constipation in this population as well. For some, the first step is getting them sitting on the toilet. For those already using the commode, better positioning on the toilet, including foot support, can help assessmentand intervention in fluid and fiber intake may

provide some avenues for improvement,keeping in mind that increased fiber in the face of low fluid intake is likely to make matters worse. For many, medications are needed. These include osmoticagents, such as polyethylene glycol, and laxatives, such as senna. Mineral oil should be used with caution, if at all, in this population because of the risk of lipoid pneumoniaif aspirated.52

RESPIRATORY

- **Respiratory compromise in children with cerebral palsy is a major source of morbidity and mortality, especially among those with severe motor involvement.**
- Cerebral palsy is associated with an increased risk of upper airway obstruction, which can cause chronic obstructive sleep apnea, as well as acute airway obstruction, especially when illness or sedation compound underlying airway issues.53 S
- noring in association with apneic pauses or gasping respirations is a clear indication for polysomnography, as is unexplained daytime somnolence.54 Parenchymal lung disease may be caused by chronic aspiration of food, secretions, or refluxed gastric contents. Severe scoliosis55 and obesity39 can both cause restrictive lung disease.

ORTHOPEDIC

- Orthopedic surgeons experienced in the care of children with cerebral palsy are integral members of the treatment team for all but those most mildly affected. **They can monitor for the secondary musculoskeletal complications of cerebral palsy, which include contractures, joint dislocations (especially at the hips), and bony deformities, such as scoliosis**. Tools at the disposal of the orthopedist include gait analysis, which can guide clinical decision-making by breaking down the gait into its components,indicating the muscle activity pattern and the alignment of the body in each phase of gait.
- Areas of particular concern in the child with cerebral palsy include the hips57 and spine.58 **A consensus statement published in Developmental Medicine and Child Neurology in 200659 recommends baseline hip films at 30 months of age in all children who can walk fewer than 10 steps, and that these films should be repeated every 6 to 12 months until age 7 years**. The statement also recommends spinal radiographs at ages 5 and 10 years in nonambulatory children with cerebral palsy, and otherwise as clinically indicated.
- Goals of orthopedic treatment include prevention of deformity, as well as treatment of deformity when such treatment will improve comfort and quality of live. Decisions about surgical interventions must take into account

the natural history of the disorder, about which we still know little. Children with dislocated hips may develop pain later, but which ones? The decision to surgically treat scoliosis in the most severely involved persons with cerebral palsy is a particularly complex one. Will the benefits for preventing progressive pulmonary compromise and loss of sitting outweigh the risks of a major surgery? This decision is best made by an experienced team with input from family and those who know the patient well.

- Thorough preoperative preparation can significantly decrease the risk of postoperative morbidity and mortality. This assessment should include a careful evaluation that looks for all the associated problems discussed in this issue. Particular emphasis should be placed on nutritional status and proactive management of gastroesophageal reflux and dysphagia, as poor nutrition, reflux, and aspiration are clearly shown60,61 to correlate with postoperative morbidity and mortality.
- Other pertinent areas to consider are the child's airway and pulmonary status, the child's risk for bleeding problems, and the child's dental health. Assessment of bone density should be considered in those who have had significant nutritional problems, have had recurrent or pathologic fractures, or who take medications that could interfere with bone health.62 Preliminary steps to improve bone density include ensuring optimal intake of calcium, phosphorus, and vitamin D. Use of such medications as the bisphosphonates has received preliminary study.63

ABNORMALTONE

- Spasticity affects most people with cerebral palsy, and a significant minority have dystonia, either in addition to or instead of the spasticity. Despite the clear difference in definitions, dystonia and spasticity are commonly confused in practice. Developingthe skills to differentiate the two is important for patient care, as different treatment approaches are often needed.
- Spasticity is defined as a velocity-dependent increase in tone. Dystonia involves involuntary muscle contractions that cause twisting, repetitive movement, and abnormal postures.64 The second major challenge to the clinician is determining where the abnormal tone fits in with the child's ''big picture,'' as we do not want to treat the tone abnormalities for their own sake, but to improve the child's function, comfort, or ease of care. Therapists working with the child have helpful input in this regard.
- **Interventions for spasticity usually start with passive stretching and other physical therapy interventions**.65 The choice of which steps to take next depends on whether the spasticity is generalized or localized. Localized

spasticity, such as in the heelcords or hamstrings, can respond well to injections of botulinum toxin66 or phenol.67

- **In more generalized spasticity, considerations include oral medications, such as baclofen;68 dorsal rhizotomy;69 and intrathecal baclofen by implanted pump.70 T**hese options are best explored by an experienced interdisciplinary team.
- For the generalist, what is most important is to realize that treatment options do exist and to help families find them when appropriate. Also important for those who care for children with intrathecal baclofen pumps is awareness of the signs and symptoms of both baclofen withdrawal and overdose, as both situations can be life threatening. Symptoms of baclofen withdrawal include fever, itching, and increased stiffness, while lethargy, seizures, and respiratory suppression can be seen with excessive doses. All patients with baclofen pumps are instructed to carry emergency procedure information with them at all times.
- **Dystonia is even more of a challenge to treat. In general, it responds less robustly and predictably to a narrower spectrum of medications, such as trihexyphenidyl.**71 Dystonia may also respond to intrathecal baclofen,72 but usually requires higher doses.73

VISION

- **Individuals with cerebral palsy have a high incidence of eye disorders, which range from refractive errors and strabismus to visual impairment**.74 There is some relationship to the etiology of the child's cerebral palsy, as prematurity is associated with retinal abnormalities that may affect vision75 and also cause a high incidence of severe myopia.76 **Those with a severe global central nervous system insult or disproportionate occipital involvement are at more risk of cortical visual impairment, and the severity of visual deficits increases with increasing severity of motor impairment.** Other lesions may be associated with optic nerve hypoplasia.77 Regardless of cause, ophthalmologic problems are important to identify, as they are often treatable. In situations where no direct treatment exists, such as optic nerve hypoplasia, a better understanding of the child's visual status will affect educational and therapy interventions.
- **All children with cerebral palsy should be evaluated by a pediatric ophthalmologist as soon as vision concerns are suspected. Many recommend such evaluation even if the child is asymptomatic**.78 Children with confirmed visual impairment qualify for and should receive specialized vision services through the schools.79

HEARING

- The incidence of hearing impairment in persons with cerebral palsy is estimated to be 10% to 15%.80 Just as in the case of vision, preterm infants are at increased risk, as are those who suffered a hypoxic or infectious insult. **Former premature infants, as well as infants treated with extracorporeal membrane oxygenations81 and those with intrauterine cytomegalovirus infections82 may experience late-onset or progressivehearing loss**, and thus require periodic hearing assessment over the first several years of life.

UROLOGIC

- **A poorly defined subset of individuals with cerebral palsy may experience voiding dysfunction**. 83 This possibility should be considered in those with recurrent urinary tract infections, unexplained irritability, or failure to achieve continence when otherwise expected based on the child's level of cognitive and motor functions. Studies have indicated that, on average, children with cerebral palsy achieve continence later than their peers,84 but the data were not controlled for cognitive level.

DENTAL

- **Persons with cerebral palsy are high risk for dental issues.85 These include malocclusion due to the abnormal forces in the oromotor musculature.86 Some children with cerebral palsy have a hyperactive gag reflex or oral aversion, which makes dental hygiene difficult**. The child's positioning needs may be difficult to accommodate in the usual dental office, and the individual may have difficulty cooperating with dental procedures because of motor or cognitive issues.87 Finally, dental care sometimes seems to ''get lost'' among the child's many other care needs. There is also the mistaken assumption that persons who receive all their nutrition by gastrostomy, because they are not eating, do not need regular dental cleaning. The converse is actually true: Persons who are exclusively fed via gastrostomy have a high risk for calculus deposition and gingivitis.88

SIALORRHEA

- Individuals with oromotor impairment may experience drooling past the age considered socially acceptable, or many do so to a degree that interferes with activities. **Complications of chronic drooling may include odor, chapping, and, rarely, pulmonary compromise**. Like many other issues, a spectrum of interventions exists from least to most invasive, with treatment decisions guided more by the degree to which sialorrhea is interfering with

the individual's life than by the amount or frequency of drooling. Options include oromotor therapy,89 intraoral appliances,90 such medications as glycopyrrolate, 91 botulinum injections,92 and surgery.93 The first two address deficits in mouth closure and swallowing frequency, while the latter interventions help indirectly by decreasing saliva production. Those who undergo surgery for drooling need careful dental follow up, as the incidence of caries appears to increase following surgery.94

PAIN

- Pain is a common problem for individuals with cerebral palsy, with more than half of adults95 and children96 with cerebral palsy reporting pain as an ongoing health concern. **Common sources are musculoskeletal and gastrointestinal. Particularly challenging is the nonverbal child with severe cerebral palsy who is excessively irritable or who cries with discomfort.** Parents are sometimes desperate to find someone who will take responsibility for working through the problem as opposed to ruling out the causes in their area of specialization. Often a thorough history and physical examination,with attention to the time course and temporal association (worse after meals or diaper changes, for example) can suggest a potential cause and intervention.Several possibilities may have to be explored and empiric interventions tried before the child finally becomes more comfortable. Parents are usually remarkably patientwith this ‘‘trial and error’’ process as long as it moves in a timely manner and they are kept involved and informed..

Table 3
Possible sources of pain

Systems	Source
Cranial	Increased intracranial pressure
	Migraines and other headaches
Ophthalmologic	Corneal abrasions
	Glaucoma
Dental	Abscesses
	Temporomandibular joint pain
Gastrointestinal	Gastroesophageal reflux
	Constipation
Muskoloskeletal	Patella alta
	Hip dislocation
	Scoliosis
Neuromuscular	Muscle spasms
Urologic	Urolithiasis
	Bladder spasms
Other	Decubitus ulcers

SLEEP

- children with cerebral palsy are prone to sleep problems, with an incidence recentlyreported to be 23%.97 **These difficulties may include difficulty falling asleep, frequent night awakening, and a sleep schedule that does not fit the needs of school or family.**For any given child, there are potential behavioral, neurologic, and physical causes. Some parents of children with special needs may have difficulty with the limit setting that is necessary for children to develop good sleep habits, such as falling asleep in their own bed and putting themselves back to sleep after a normal night awakening.
- In some children, the ‘‘biologic clock’’ or other neural mechanisms involved with sleep initiation and maintenance may be abnormal. Physical causes may include difficulty with position changes during the night as well as the myriad potential sources of discomfort mentioned in the previous section. When behavioral concerns and sources of discomfort have been addressed but sleep problems persists, medication may beconsidered. Studies with grade B evidence support the use of melatonin,98 particularly to help the child fall asleep, with some studies showing the additional benefit of fewer night awakenings and longer duration of sleep. Antihistamines, the melatonin receptor agonist ramelteon, and a variety of other medication are used, though they lack supportive research, and no medications approved by

the Food and Drug Administration for insomnia in adults have been studied adequately in children.

TRANSITION

- The vast majority of individuals with cerebral palsy live into adulthood and thus need to make the transition to adult medical care. Numerous barriers to this transition have been identified,99 including insufficient knowledge and training in childhood-onset disabilities among adult health care providers, as well as funding issues. The American Academy of Family Medicine, the American College of Physicians, and the American Academy of Pediatrics100 have agreed on steps to ease the transition of persons with special needs from pediatric to adult health care. One of these steps addresses the training issue. They also recommended three concrete steps that pediatric practitioners can take now to improve transition: Identify a health care provider who will take responsibility for working with the patient and family on transition; work with the individual and family to develop a written transition plan; and facilitate development of a portable health care summary. Fortunately, many transition checklists and medical summary templates are available to facilitate these tasks. Many are available through the National Center of Medical Home Initiatives for Children with Special Needs.

Movement disorder

- Spasticity is a velocity-depenydent resistance of a muscle to stretch. It is defined as havingone or both of the following signs: (1) resistance toexternally imposed movement increases withincreasing speed of stretch and varies with the direction of joint movement, and/or (2) resistanceto externally imposed movement rises rapidlyabove a threshold speed or joint angle.” Spasticity can vary depending on several factors such as thepatient’s level of activity, level of alertness,emotional state, and discomfort. On examination, upper motor neuron signs such as hyperreflexia,clonus, and Babinski response are commonlypresent.
- Spasticity is a result of injury to the centralnervous system that produces an upper motorneuron lesion. Secondary consequences of the injury occur all the way to the muscle fiber. Fibersmay have increased variability in size, and insome cases there is an increase in the number of type I muscle fibers and a deficiency of type IIbmuscle fibers.5 Spasticity is thought to be themost common tone abnormality in children withcerebral palsy, and it is certainly seen as a commonconsequence of injury to the white matter of thebrain, such as periventricular leukomalacia caused by the stretching of the pyramidal tracts from intraventricularhemorrhage.

- A child with evidence of increased tone on examination that is not improving with standard anti-spasticitytreatments may also be suffering from an underlying dystonia. The consensus-based definition formulated by the Taskforce on Childhood MotorDisorders is as follows4: “Dystonia in childhoodis a movement disorder in which involuntary sustained or intermittent muscle contractions causetwisting and repetitive movements, abnormal postures, or both.” Dystonia can cause hypertoniaif there is dystonic muscle contraction opposingpassive movement of the limb being tested by the examiner,4 which is referred to as dystonichypertonia. Dystonia can also be classified in relationto the region of the body that is affected.
- Focaldystonias affect a single body part, segmentaldystonias affect contiguous body parts, and multifocal dystonias affect 2 or more noncontiguousbody parts.3,7 On examination, dystonia can beobserved while the patient is at rest or is sometimes triggered by a voluntary task.8
- The physiologybehind dystonia involves cocontraction ofantagonist muscles, overflow of electromyographic activity onto uninvolved muscles duringvoluntary movement, and involuntary activationof muscles during passive shortening,9 which may be the result of damage to deeper structuresof the brain, including the basal ganglia, whichacts as a feedback loop for cortical activity.10 “(1) the resistance to externallyimposed joint movement is present at very low speeds of movement, does not depend onimposed speed, and does not exhibit a speed orangle threshold; (2) simultaneous co-contraction of agonists and antagonists may occur, and this isreflected in an immediate resistance to a reversalof direction of movement about a joint; (3) the limb does not tend to return toward a particularfixed posture or extreme joint angle; and (4) voluntary activity in distant muscle groups does notlead to involuntary movements about the rigidjoints, although rigidity may worsen.”

Oral medications for hypertonia

Medication	Mechanism of Action	Side Effects	Pediatric Dosing
Diazepam	Facilitates transmission at the $GABA_A$ receptors, one of the principal types of inhibitory synapses in the CNS	Sedation, decreased motor coordination, impaired attention and memory. Can cause overdose or withdrawal symptoms	0.12—0.8 mg/kg/d in divided doses or once daily at night
Baclofen	Binds to $GABA_B$ receptors in the spinal cord to inhibit reflexes that cause spasticity	Sedation, confusion, nausea, dizziness, hypotonia, muscle weakness, and ataxia. Can cause withdrawal symptoms	Start at 2.5—10 mg/d. Can taper up to a maximum of 40 mg/d divided tid or qid
Dantrolene	Acts at the site of skeletal muscle to inhibit calcium release from the sarcoplasmic reticulum	Sedation, diarrhea, and dizziness. Causes hepatotoxicity in 2%, so LFTs must be monitored	Start at 0.5 mg/kg/d. Can taper up to a maximum of 3 mg/kg administered qid
Tizanidine	α_2-Agonist that acts on the brain and spinal cord to decrease tone through hyperpolarization of motoneurons	Sedation, dizziness, dry mouth, elevated levels in LFTs, insomnia, muscle weakness	Start at 2 mg at bedtime. Can taper up to a maximum of 32 mg/d divided tid or qid

Intramuscular medications for hypertonia

Medication	Mechanism	Site of Injection	Onset and Duration	Disadvantages/Risks	Cost
Phenol	Chemical neurolysis causes denervation via axonal degeneration	Injected into the motor points of the involved muscle	Takes effect immediately. Can last 3—12 mo	Can be painful and may require anesthesia. Can cause dysesthesias, numbness, or hematoma	Very low cost
Botulinum Type A	Presynaptic block of acetylcholine release	Intramuscular	Takes effect in 5—7 d. Can last 3—6 mo	Lasts only 3—4 mo and cannot be repeated in shorter intervals. Can cause swallowing and respiratory difficulties when used in large quantities in the neck muscles	Up to $600 per 100-U vial

Oral Medications for the Treatment of Dystonia

- All patients with childhood dystonia should receivea trial of carbidopa/levodopa because it is highlyeffective in dopa-responsive dystonias and otherdisorders affecting dopamine synthesis.48 Thedrug is slowly titrated up to the maximum tolerateddose; central side effects such as

memory impairment,confusion, and hallucinations usually limitdosing. Effects are usually seen with levodopa, less than 300mg,when combined with carbidopa.49If dopaminergic agents are ineffective, anticholinergicsshould be tried. In children who havea secondary dystonia caused by cerebral palsy,

Table 3
Comparison of carbidopa/levodopa and trihexyphenidyl for dystonia treatment

Drug	Mechanism of Action	Side Effects	Pediatric Dosing	Cost
Carbidopa/ Levodopa	Inhibits peripheral dopamine decarboxylation. Serves as a dopamine precursor	Dyskinesia, bradykinesia, hypotension, memory impairment, confusion, and hallucinations	Start at 10/100 mg bid and titrate up to 25/100 mg tid	$90–$115 for a 1-mo supply
Trihexyphenidyl	Antagonizes acetylcholine receptors	Dry mouth, blurry vision, dizziness, nausea, anxiety, glaucoma, anhidrosis, neuroleptic malignant syndrome, and tardive dyskinesia	Start at 2.5 mg/d and titrate up slowly as needed. Maximum dose is 15 mg/d	$25–$70 for a 1-mo supply

COMPONENTS OF REHABILITATION

- Rehabilitation of the child who has CP consists of improving mobility, preventing deformity, helping the child to learn the skills he or she needs in daily life, and educatingthe parents about the child's problem. Rehabilitation should help to provideschooling, sports, and recreation for the child. Methods used in CP rehabilitationare physiotherapy, occupational therapy, bracing, assistive devices, adaptive technology,and sports and recreation.Physiotherapy aims to bring the child to an erect position, give the child independentmobility, and prevent deformity (Table 4). It begins in early infancy and continuesthroughout adolescence to facilitate normal

neuromotor development. With the help of

Table 4
Effects of physiotherapy

Physiotherapy Tries to Improve	
Postural control	Decreasing spasticity and contracture
Muscle strength	Increasing muscle elasticity and joint laxity
Range of motion	
Joint alignment	Increasing coordination/agility
Motor control	Balance
Muscular/cardiovascular endurance	Transitions
and mobility skills	Use of assistive devices

correct positioning, appropriate stimulation, and intensive exercise, the therapist triesto gain head control, postural stability, and good mobility in the child. This is possibleonly to the extent of the child's neurologic capacity. Therapeutic exercises help thechild to learn how to sit, stand, and walk and how to use his or her upper extremityfor function. The child also learns how to use his or her remaining potential to compensatefor the movements he or she cannot perform.

- Decreasing spasticity, gainingmuscle strength, and improving joint alignment decrease deformity. The educationof caregivers involves gently coaching them to set reasonable expectations for theirchild and teaching them to follow their child's exercises at home. Parents should encourage their children to participate in daily living activities by using the functionalskills they learned during therapy. Community and social support is another aspectof rehabilitation.18\

- All therapy methods should support the development of cognitive, visual, sensory, and musculoskeletal systems; involve play activities to ensure compliance; enhancesocial integration; and involve the family. The basic element is having fun. Occupational therapy aims to improve hand and upper extremity function in the child through play and purposeful activity. Sports and physical and recreational activitiesplay an important role in physical development, general fitness, and health of thechild who has CP. Physical and occupational therapy, combined with recreational activities or adapted physical education, increase the efficiency of rehabilitation andassist the disabled child to use his or her potential.19
- Management should prevent disability by minimizing the effects of impairments, preventing secondary disabilities, and maximizing motor function throughout the child's life. Functional goals change as the baby becomes a child and the child matures into an adult, but the principles

remain the same. Younger children focus mainly on mobility, whereas adults put more emphasis on communication and activities of daily living.

- The first 4 years are spent in physiotherapy and bracing, orthopedic surgical procedures are performed between 5 and 7 years of age, and education and psychosocial integration become the main issues in adolescents. Mobilization goals should be met by the time the child is ready to go to school. Aggressive physiotherapy is needed in the growth spurt period and after orthopedic surgery

Bracing

- Braces are devices that hold the extremities in a stable position. The goals of bracing are to increase function, prevent deformity, keep the joint in the functional position, stabilize the trunk and extremities, facilitate selective motor control, decrease spasticity, and protect the extremity from injury in the postoperative phase. Indications differ according to the age, selective motor control level, type of deformity, and functional prognosis of the child.21 Various kinds of ankle-foot orthoses (AFOs) are the most common braces used in CP (Fig. 2). Knee-immobilizing splints and hip abduction splints are prescribed for nonambulatory and ambulatory children. Compliance with night splints to prevent deformity is low. The indications of bracing in the upper extremity are limited.

Assistive Devices

- A child who has CP needs to move around, to explore his or her surroundings, and to interact with his or her peers so that his or her mental, social, and psychologic skills develop to the fullest. A variety of mobility aids and wheelchairs provide differing degrees of mobility to these children. Transfer aids, such as lift systems, assist the caregiver when performing transfers. Passive standing devices called standers allow the child to get accustomed to standing erect and provide therapeutic standing. Some ambulatory children use gait aids, such as walkers, crutches, and canes, in addition tobraces for efficient and safe ambulation. Gait aids mainly used to improve balance also decrease energy expenditure, decrease the loads on the joints, and improve posture and pain. Nonambulatory children use wheelchairs for moving around. Wheelchairs must be properly fitted with seating aids, cushions, and other positioningcomponents.

Spasticity Management

- Spasticity should be treated when it causes loss of function; produces contractures, deformities, pressure sores, or pain; or causes difficulty in positioning or caring for the total body–involved child. All treatment options

aim to modulate the stretch reflex. Basic measures, such as positioning, exercises, and bracing, may be combined with oral medications or botulinum toxin injections to improve outcome. Orthopedic surgery is an effective option in moderate to severe spasticity.

Oral management

- Systemic oral antispastic drugs are useful for total body–involved children who have severe spasticity, and for a short period after orthopedic surgery, but are not recommended for ambulatory children because of side effects, such as drowsiness, sedation, and generalized weakness.22

Botulinum toxin

- **Botulinum toxin inhibits acetylcholine release at the neuromuscular junction causingreversible chemodenervation. The general indication for botulinum toxin injection in CP is ‘‘the presence of a dynamic contracture, interfering with function, in the absence of a fixed muscular contracture**.’’23 If botulinum toxin injections are started at an early age and repeated as necessary, they can help to prevent the development of muscle contractures and bony deformities.23

Intrathecal baclofen

- Intrathecal administration introduces baclofen directly into the cerebrospinal fluid (CSF) through an implantable pump and catheter system, increasing efficacy and minimizing side effects. Intrathecal baclofen administration is useful for the severely involved spastic child, dystonic child, or child who has mixed disease to enable sitting in the wheelchair, make transfers easier, decrease spinal deformity, and increase the comfort level and ease of care.24

Selective dorsal rhizotomy

- SDR involves sectioning of the dorsal column rootlets to interrupt the spinal reflex arc, inhibiting afferent input from the muscle and tendons, and reducing efferent activity at the level of the spinal cord. There is a permanent and global muscle tone reduction in the lower extremities with loss of superficial and deep sensation. Hip instability and spinal deformity are more common after the procedure.25,26

Corrective casting

- Corrective casting is used for minor ankle equinus contracture that does not respond to physical therapy or botulinum toxin injections and for knee flexion deformities that involve more than just hamstring tightness. The compliance with serial casting is low because of the difficulties of repeated casting and cast removals.18 Some researchers propose that casting weakens

the already weak spastic muscles, creates atrophy, and does not allow the antagonist muscle to work.

ORTHOPEDIC SURGERY

- Orthopedic surgery is widely used in the management of children who have CP to prevent or correct certain musculoskeletal problems, such as muscle shortening and bony deformities. The goal of orthopedic surgery in a child with walking potential is to improve functional ambulation. For a good walking pattern in the ambulatory child, the feet should be plantigrade and stable, the hips should extend well and be stable, and the knees should have good extension. For nonambulatory children, the goal of orthopedic surgery is to ease the burden of care through facilitating sitting,improving hygiene, and preventing pain. Therefore, the spine should be straight, the pelvis horizontal, and the hips stable

Indications

- **The main indication for orthopedic surgery is to correct contractures and deformities of the spine and extremities that disturb sitting, standing, and walking. O**rthopedic surgery reduces muscle tone by lengthening the spastic muscles and decreasing the sensitivity of the stretch reflex. Balance is decreased immediately after surgery but improves in the long run because of the plantigrade stable feet providing a better base of support. Muscles usually get weak, but they can be strengthened. Tendon transfers change the direction of deforming forces that create muscle imbalance, thus preventing deformity and allowing the child to use his or her muscle strength more efficiently. Decreased spasticity and improved balance may indirectly improveselective motor control; however, primitive reflexes do not change after surgery.27

Surgical Methods

- Orthopedic surgical procedures used in CP are muscle releases and lengthenings, split tendon transfers, osteotomies, and arthrodeses. Muscle-tendon lengthening weakens spastic and shortened muscles and decreases the unopposed pull of spastic muscles, thereby balancing the forces acting on the joint. Even severe contractures can be treated effectively with muscle lengthening. **Osteotomy corrects varus and valgus deformities of the foot and flexion deformities in the lower extremity. Hip osteotomy stabilizes the subluxated or dislocated hip, preventing pelvic obliquity and pain, to improve sitting balance.** Rotational osteotomies correct the torsional deformities in the tibia or the femur and help to transfer the malaligned muscle force into the correct plane of movement to make it easier for the

child to walk. Arthrodesis corrects deformity and stabilizes the joint. Spinal fusion and instrumentation correct spinal deformity.

- he correction of joint alignment makes walking easier, and the child may stop using coping mechanisms and adaptive responses that he or she developed because of his or her contractures and deformities.
- **Timing of surgery depends on CNS maturation, ambulation potential, and rate of deformity development. The nervous system matures around the age of 4 to 6 years when the physician can assess muscle imbalance more accurately**; predict a functional prognosis; and make sure that no other abnormalities, such as athetosis or dystonia, are present. **Therefore, soft tissue procedures are generally performed between 4 and 7 years of age, hand surgery between 6 and 12 years of age, and bony procedures after 8 years of age although it is the needs of the child that determine the exact timing.** Exceptions are progressive hip instability and deformities and contractures interfering with function.18,27
- Orthopedic surgery is usually performed to correct pes equinus and pes varus in hemiplegia and jump, scissoring, and crouch gait in diplegia. In the child with total body involvement, spinal deformity and hip instability are treated with surgical methods. Certain patients benefit a lot from orthopedic surgery, whereas others may not benefit at all. Spastic diplegic and hemiplegic children improve more compared with spastic total body–involved, dyskinetic, and mixed types. Fewer operations are performed and the gains are limited in dyskinetic cases. Results are best in children who have a higher degree of selective motor control.
- Lack of balance, cognitive deficits, and visual impairments are not by themselves contraindications for orthopedic surgery and do not affect the surgical outcome unless they are extremely severe. Surgical procedures requiring postoperative intensive physiotherapy or long-term cast immobilization should be avoided in children who have severe cognitive deficits, however.28 Children who have sensory deficits are not candidates for upper extremity surgery for function.

Postoperative Care

- The focus in the immediate postoperative period is on analgesia and muscle relaxation. Usually, a combination of a narcotic analgesic and diazepam helps to control the immediate pain. Early mobilization and early weight bearing in addition to strengthening of the trunk and upper extremities should be encouraged. Casts, splints, plastic AFOs, or knee ankle foot orthosis (KAFO) are used depending on the age and cooperation of the child and surgical stability. Weight bearing is allowed on the second to fourth day after soft tissue surgery, whereas it depends on the quality of internal

fixation of the bones in combined soft tissue and bony operations. Adequate nutrition and skin care are necessary to prevent complications, such as pressure sores.

- Range-of-motion and strength exercises begin as early as possible after surgery. The lengthened muscles need to be strengthened with proper exercises. It usually takes approximately 3 months to regain the preoperative muscle strength after multilevel surgery.29 The child then begins to acquire new skills. Changes in function are not extremely obvious for up to 1 year after the operation. Postoperative physiotherapy should be combined with sports and purposeful play to increase the benefits of surgical intervention. The physician must also monitor for new dynamic or fixed contractures or orthotic problems, prescribe accurate braces, and provide guidance for adaptive equipment.

Complications of Surgery

- Common complications are pressure sores attributable to prolonged bed rest or cast, sciatic nerve traction injury after surgery to relieve knee flexion contracture, and fracture in the immobilized and osteoporotic children who are wheelchair-bound spastic quadriplegics. Heel cord and hamstring tendon overlengthening can lead to pes calcaneus and knee recurvatum, respectively.30

The ‘‘Birthday Syndrome’’

- One group of complications related to a chain of operations over the years is social isolation, loss of motivation, frustration, and psychosocial problems termed the birthday syndrome.31 Instead of performing one operation at a time over many years, all necessary surgical interventions should be done at the same time in a single setting if the child’s medical and social status permits. This spares the child the burden of multiple consecutive surgical interventions throughout his or her life. Treatment plans must be individualized for each child according to his or her specific needs

Limitations

- Surgery is not the single solution to the wide variety of problems caused by CP. It is only a momentary pause in the long journey of CP management. The need for rehabilitation measures, such as bracing, physiotherapy, sports, and antispastic medication, still remains after surgery.
- Because family cooperation is essential for the success of treatment, realistic goals should be set after a thorough evaluation of the expectations and limitations of the family. The expected functional gains after surgery must be thoroughly and realistically decided on, and surgery should not be

attempted unless one is certain that the operation is going to create considerable functional gains not obtainable by other means

PROBLEMS SPECIFIC TO THE TYPE OF CEREBRAL PALSY

Problems and Management in Hemiplegia

- Musculoskeletal problems of the upper extremity are shoulder abduction, internal rotation, elbow flexion and pronation, wrist and finger flexion, and thumb in palm. Typical lower extremity problems are hip flexion, internal rotation, knee flexion or extension, ankle plantar flexion and foot varus, although valgus may also be seen. The hemiplegic side may be slightly atrophic and short.\
- Physiotherapy may prevent contractures of the involved side and strengthen the weak muscles, thus enabling better use of the upper extremity and a more efficient gait pattern. Botulinum toxin injections reduce gastrocnemius-soleus and rectus femoris spasticity in the young child. Early relief of spasticity may prevent shortening of the gastrocnemius muscle, help to establish a close-to-normal gait, and delay or eliminate the need for surgical intervention.23 Injection to the upper extremity may minimize forearm pronation and relax wrist, finger, and thumb flexors so that the child may gain forearm supination and wrist stabilization. Hand splints in hemiplegia are used to prevent deformity and to improve function. The child's compliance with night splints is generally poor. Splints prevent sensory input in the already compromised hand. AFOs stabilize the ankle and foot and keep it in the plantigrade position for weight bearing.
- All fixed contractures must be corrected before applying braces. The usual indications for orthopedic surgery in the hemiplegic child are pes equinus,pes varus, and stiff knee. Thumb-in palm and wrist flexion deformity may also respond to surgery. Soft tissue procedures are usually performed in children around 5 to 6 years of age. Bone procedures are delayed until children are at least 8 years of age unless the deformity is causing a functional problem.32 Upper extremity surgery for function should be delayed until the child is mature enough to cooperate with rehabilitation.

Problems and Management in Diplegia

- Problems in maintaining balance; muscle imbalance; and spasticity leading to contractures and deformities of hips, knees, and ankles contribute to the specific posture and gait patterns typical for diplegic CP. The frontal plane pathologic condition is scissoring, which occurs as a result of persistent femoral anteversion and hip adductor and medial hamstring spasticity. A scissoring gait may accompany sagittal plane pathologic findings, such as

jump or crouch knee gait. Jump gait, defined as excessive hip flexion, knee flexion, and equinus in stance, is the most common sagittal plane pathologic finding in young diplegic children. The crouch gait that occurs in the older diplegic child is defined as excessive knee flexion and ankle dorsiflexion throughout stance. Common causes are short or spastic hamstrings, hip flexor tightness, and excessive ankle dorsiflexion. Stiff knee gait is characterized by limited or lack of knee flexion in swing as a result of rectus femoris spasticity or unopposed rectus femoris function after hamstring lengthening. Genu recurvatum, conversely, is hyperextension of the knee at midstance, which is generally associated with mild equinus caused by triceps surae spasticity, excessive spasticity in the quadriceps, weak hamstring muscles, or hip flexion contracture.32

- Positioning, strengthening, and stretching exercises preserve joint range of motion, increase strength, and help to improve gait. They should be combined with bracing, walking aids, and antispastic treatments. The risk for contracture formation in the more vulnerable biarticular hamstring and gastrocnemius muscles increases during the prepubertal growth spurt. Intensive physiotherapy and botulinum toxin injections to lengthen the spastic muscles may prevent contractures then. Exercises should be integrated into play, particularly in toddlers and in noncompliant children. Oral antispastic medications are valuable if spasticity interferes with sleep. Botulinum toxin is useful to relieve spasticity of the lower extremities and prevent contractures of the young diplegic child when it is too early for orthopedic surgery. It must be combined with physiotherapy and bracing. Most diplegic children need variations of the AFO to provide a stable base for standing and maintaining good joint alignment during walking. Resting and night splints are used to prevent knee and ankle contractures; however, the compliance is poor in the child who has severe spasticity.
- Most deformities of diplegics can be prevented or corrected with appropriate orthopedic intervention. The ideal age of surgery is between 5 and 7 years of age or when the child is able to cruise holding onto furniture or walk holding hands. Early surgery may be necessary in cases with hip instability; knee flexion contracture because of spastic hamstrings; and contracture of gastrocnemius-soleus unresponsive to physiotherapy, botulinum toxin, or serial casting. It is necessary to define clearly all the musculoskeletal problems of the lower extremities before surgery and address them in a single setting if possible to obtain a successful result.14,33 All orthopedic interventions should be timed so as not to interfere with the child's education and social life. Sports activities and play with peers are essential during school years. Swimming and horseback riding are beneficial for the

poorly developed balance reactions of the diplegic.34 These activities restore a sense of well-being and self-confidence in the child. Occupational therapy is helpful to improve hand function.

Problems and Management in Quadriplegia

- Spine and hip deformities, such as hip instability, pelvic obliquity, and scoliosis, are common and interfere with sitting balance. Knee and ankle deformities seen in hemiplegic and diplegic children may also exist. The incidence of lower extremity contractures increases with severity of the motor impairment. Elbow flexion-pronation contracture creates problems when using forearm crutches, and severe flexion contractures in the hand impair hygiene and cosmesis.
- Hip instability and spinal deformity of the nonambulatory quadriplegic child do not respond to conservative measures and generally require orthopedic surgery.

Scoliosis

- can develop as early as 5 years of age and continues to progress after skeletal maturity, especially if the curve exceeds 40_.35 Hyperkyphosis is common in the young child with weak spinal extensor muscles. The hips are usually normal during the first years of life. Progressive instability occurs later because of a combination of muscle imbalance, persistent primitive reflexes, faulty posture, and absence of weightbearing stimulation on bone to progressive instability.
- Dislocation can occur as early as 18 months of age, and most hips dislocate by the time the child is 6 years of age if they are going to do so.36,37 Hip dislocation affects hygiene, sitting, and gait of the total body–involved child. It causes pain by early adulthood. Secondary scoliosis and contralateral adduction deformity causing ‘‘windswept hips’’ further worsen the situation.
- The aim of rehabilitation in quadriplegic children is to obtain and maintain sitting balance. Infant stimulation, positioning, and parent education are important between 0 and 2 years of age. Ways to decrease muscle tone should be emphasized from the age of 2 years, when muscle tone becomes a problem and dyskinesias manifest themselves. In the nonambulatory child, orthopedic surgery is usually reserved for hips at risk and spinal deformity. Hygiene, seating, and care issues in addition to preventing pain and discomfort secondary to spasticity become predominant during the teenage years. The knee and ankle deformities should also be corrected in

mildlyinvolved quadriplegic children who have the potential to stand independently and take a few steps to enable efficient transfers and limited ambulation. The aim is to obtain a comfortable posture in lying, sitting, and the standing frame. The knee should flex to 90_ for sitting and extend to at least 20_ for transfers.

- Stretching and range-ofmotion exercises may prevent knee flexion deformity early on. Regular exercises, night splints, and standing in the stander to protect the range of motion gained by surgical intervention are necessary.

Problems and Management in Dyskinesia

- Hyperkinetic or choreoathetoid children have purposeless, often massive involuntary, movements that increase when the child is excited or frightened. Dystonic children manifest abnormal shifts of general muscle tone induced by movement, leading to an abnormal posture. Contractures are almost never seen. Degenerative hip disease and acetabular dislocation are common complications during the adolescent growth spurt, particularly in children who have athetoid CP.30,38 Scoliosis is common. The complication rate of spine surgery is high. The aim is to minimize muscle contractions and unwanted movements to ease the burden of care and the child's discomfort