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Cerebral Palsy

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Dr. Swaiman was the Chairman of the organizing committee and first President of the Child Neurology Society (CNS). He received the Hower award, the highest award of that society and the Founder’s Award at its 25th Anniversary meeting as well as The Lifetime Achievement Award for Neurologic Education by the American Academy of Neurology. As the chairman of the organizing committee of the Professors of Child Neurology, he was its first President and a prime mover and also first president of the Child Neurology Foundation. He was a member of the organizing committee of the International Child Neurology Association (ICNA), on many National Institutes of Health Study Sections and visiting professor and lecturer at medical schools in the United States and throughout the world including Canada, South America, Asia, Mexico, Europe, and Africa.

Dr. Swaiman has been the Editor and a primary contributor to the textbook titled Practice of Pediatric Neurology (2 editions) and Pediatric Neurology: Principles and Practice (5 editions). He is the founding editor and immediate past Editor-in-Chief of Pediatric Neurology, an international journal devoted to the basic and clinical aspects of the diagnosis of children with neurologic impairment. He has served as a member of the Editorial Boards of the Annals of Neurology, Brain and Development, Neuropediatrics, and the Chinese Journal of Pediatrics.

His investigative endeavors have included research into brain energy metabolism, the effect of malnutrition on the developing brain, and the metabolic effects of iron and other metals on metabolism of various portions of the brain. He has been particularly involved in studies of Neurodegeneration with Brain Iron Accumulation (NBIA) and other movement disorders of children.

SUMMARY

Cerebral palsy consists of a group of brain disorders involving movement and posture causing limitation of activity. The condition is most commonly evident in the early months of life, but usually dates back to the neonatal period. Although muscle tone (ranging from floppy to stiff) and abnormal postures (body and limb positions) may become more pronounced during early childhood, changes in type are uncommon. The condition is always non-progressive. There is a wide range of involvement, from mild to severe. Although life expectancy may be decreased, acute death is extremely rare. Intellectual, sensory, and behavioral difficulties may accompany cerebral palsy, but are not necessary for diagnosis.

Because cerebral palsy is an umbrella term that includes a wide range of movement dysfunction, causes are varied and are discussed later in this article. It is important to know that children with cerebral palsy often have accompanying intellectual delay, hearing impairment, and speech and language disorders. Additionally, epilepsy occurs in many children with cerebral palsy.

The definition of cerebral palsy in research studies often varies depending on the research plan; therefore, the definition varies depending on the citation. In spite of all the various definitions, designating cerebral palsy as a single entity is valuable because affected children commonly have similar needs for medical care, rehabilitation, and social services.

DESCRIPTION

Cerebral palsy can be classified by the following qualities:

- Movement abnormalities: Muscle tone abnormality (increased or decreased) and type of movement disorder (e.g., ataxia, dystonia, choreoathetosis)
- Severity of the handicap
- Anatomic: Parts of body affected (e.g., hemiplegia, diplegia, quadriplegia) See later in this article for classification

PREVALENCE

The precise number of affected individuals with cerebral palsy in a population is not known. Most likely, cerebral palsy occurs in 1.2 to 3.6 children per 1000 live births. Numerous cerebral palsy registries exist throughout the world. Widespread regional prevalence rates have remained consistent over several decades. Based on current figures, approximately 8,000 children with cerebral palsy are born annually in the United States.

SYMPTOMS

In most children with cerebral palsy, delay in attaining developmental milestones is the most distinctive symptom of the condition. A detailed history as well as thorough physical and neurologic examinations are critical in the diagnostic process. Records of the mother's pregnancy and delivery and records of the infant's early neonatal period can prove invaluable.

Physical and neurologic abnormalities may be subtle and professionals should be reserved about diagnostic statements unless the findings are unequivocal.

Signs and symptoms of birth asphyxia or neonatal encephalopathy are strongly predictive of cerebral palsy in a child. For instance, full-term infants whose immediate postpartum course is comprised of a 5-minute Apgar score of 5 or less, with continuing neurologic abnormalities and seizures in the first days of life, constitute a high-risk group for developing cerebral palsy.

PHYSICAL ABNORMALITIES

The initial clinical findings may change with maturation and severity may change, but distribution is only occasionally altered. A child with cerebral palsy who has been hypotonic may become hypertonic. Although unusual, some infants with mild abnormalities subsequently manifest a decrease or, in rare instances, a disappearance of motor dysfunction.

The clinical pattern should not include evidence of progressive disease or loss of previously acquired skills. Progression of disability (qualitatively and quantitatively) requires review of the diagnostic possibilities to explain the progression. History-taking should focus on identification of a specific cause and should particularly investigate familial or metabolic disease. Such information may have value for determining treatment or family counseling.

The diagnosis of cerebral palsy often is suggested by the persistence of primitive reflexes, the presence of pathologic reflexes elicited by the clinician, abnormal muscle tone, and the failure to develop maturational reflexes in a timely fashion. These maturational reflexes include the traction response and the parachute response. The use of standard developmental screening tests (e.g., the Denver Developmental Screening Test II) may provide quantitative evidence of motor delay, as well as evidence of delay in acquisition of other skills.

CAUSATION

Premature birth is the single most important risk factor for cerebral palsy. The risk of cerebral palsy in very-low-birth-weight infants (birthweight below 1500 grams) is as high as 4% to 10%, whereas the risk in term infants is only 1 to 1.5 per 1000 live births. Infants born at 24-26 weeks gestation may have as high as a 20% chance of developing cerebral palsy. In some pertinent studies, prematurity accounts for approximately half of all infants with cerebral palsy.

An increase in the prevalence rate of cerebral palsy among preterm infants during the mid-1980s was attributed to increased survival of low-birth-weight infants. Later studies suggest that the rate of cerebral palsy among preterm infants has not changed.

Nonetheless, it is noteworthy that term infants represent more than half of all cases of cerebral palsy. The prevalence of cerebral palsy among term infants, 1-1.5 per 1000 live births, has not changed during the past 3 decades. The common use of electronic fetal monitoring and the pronounced increase in births by cesarean section appear to have had little affect.

The unchanged prevalence is disappointing in view of the fact that perinatal deaths, stillbirths, and birth asphyxia as measured by low Apgar scores have dropped dramatically in recent decades.

Male infants have a greater risk of cerebral palsy than female infants. Twins, who constitute 2% of the population, also carry a higher risk and contribute 10-12% to the overall prevalence of cerebral palsy. The increased risk of cerebral palsy among multiple birth babies is in part the result of increased rate of prematurity; twins born at term may also have an increased risk for cerebral palsy .

Black children have an increased likelihood of cerebral palsy when compared with white children. Although this finding is due in part to an increased rate of prematurity, the risk of cerebral palsy among black infants born at term may also be elevated. In addition, studies suggest that infants born to mothers with lower socioeconomic status have an increased risk of cerebral palsy.

A wide range of causative disorders and risk factors have been identified for cerebral palsy. Table 1 lists some of the more common causal categories and risk factors broadly lumped into the following groups: perinatal brain injury, brain injury related to prematurity, developmental abnormalities, prenatal risk factors, and postnatal brain injury.

These factors may co-exist and interact in contributing to the causes of brain injury resulting in cerebral palsy.

Table 1

PERINATAL BRAIN INJURY

Intrapartum hypoxia-ischemia brain injury is a well-described cause of cerebral palsy, especially in the setting of an acute intrapartum event such as uterine rupture, placental abruption, or cord prolapse. The extent to which hypoxic-ischemic brain injury is responsible for cerebral palsy, however, has been a major source of controversy.

On the contrary, studies suggest that in a minority (between 6% and 28%) of affected children, cerebral palsy is due to perinatal asphyxia. The term perinatal asphyxia is confusing and deserves further clarification. Traditionally, this term has been defined by clinical signs and symptoms that include low Apgar scores, meconium-stained amniotic fluid, and low cord blood pH. The clinical findings used to define this term are not exclusive to hypoxic-ischemic brain injury nor is there a widely accepted evidence-based standard for determining when cerebral palsy is related to hypoxia-ischemia.

To the extent hypoxic-ischemic brain injury is an accepted cause for a case of cerebral palsy, the form of cerebral palsy most often associated with it is spastic quadriparesis (discussed later).

BRAIN INJURY RELATED TO PREMATURITY

Preterm infants have a more than expected share of the cases of spastic diplegia cerebral palsy but can manifest any cerebral palsy subtype. Both intraventricular hemorrhage and white matter necrosis seen in periventricular leukomalacia may occur before or after birth. The main pathogenetic mechanisms underlying periventricular leukomalacia are hypoxia-ischemia and inflammation, which are brain injuries associated with cerebral palsy (discussed above).

DEVELOPMENTAL ABNORMALITIES

Brain malformations originating from intrauterine maldevelopment may underlie the neurologic impairment seen in children with cerebral palsy. Children with cerebral palsy have a higher incidence of congenital malformations of the brain.

A genetic or metabolic disorder may be associated with a specific brain malformation that causes cerebral palsy. For example, polymicrogyria and lissencephaly, as well as other cortical malformations, may be seen on imaging studies. Some relatively rare metabolic diseases may result in the clinical picture of cerebral palsy.

Children with cerebral palsy who demonstrate either progressive decline, cerebral malformations, or who have a known family history should be tested for a causal genetic or metabolic disorder.

PRENATAL RISK FACTORS

Several maternal conditions have been associated with an increased risk of cerebral palsy. In particular, intrauterine inflammation, or chorioamnionitis, has received increasing attention as a potential risk factor. Studies suggest that maternal intrapartum fever, a clinical or histologic diagnosis of chorioamnionitis, and serologic markers of inflammation in the fetus all confer an increased risk of cerebral palsy. It is hypothesized that the fetal inflammatory response that occurs in the setting of an inflammatory intrauterine environment is responsible for brain injury leading to cerebral palsy, yet the mechanism by which intrauterine inflammation might cause cerebral palsy remains unproved.

In term infants, chorioamnionitis, often diagnosed by the presence of intrapartum maternal fever, is a particularly strong risk factor for spastic quadriplegia. Maternal fever during labor also increases the chance that the child will have low Apgar scores and characteristics of neonatal encephalopathy.

Other prenatal risk factors associated with an increased risk of cerebral palsy include: slower than expected intrauterine growth — especially in term infants, infants who are large for gestational age blood clotting abnormalities, and history of infertility.

Finally, a number of toxins have been reported to rarely cause cerebral palsy. Congenital infections, often referred to as TORCH infections (toxoplasmosis, rubella, cytomegalovirus, and herpes simplex infection), also can infect the fetus and produce serious brain inflammation with motor sequelae. Recent literature suggests that the risk of cerebral palsy may be modified by relatively common genetic distortions.

POSTNATAL BRAIN INJURY

Cerebral palsy may result from brain injury occurring during the neonatal period. For instance, neonatal infections such as meningitis from group B streptococcal may cause brain damage leading to cerebral palsy. Other types of postnatal brain injury that may cause cerebral palsy include hypoxia-ischemia and traumatic brain injury from nonaccidental trauma.

Bilirubin toxicity is a well-known cause of dyskinetic cerebral palsy and continues to be a significant problem despite vast improvements in hyperbilirubinemia therapy. While bilirubin levels below 25 mg/dL are rarely

associated with bilirubin-induced brain damage (kernicterus); bilirubin levels greater than 30 mg/dL are often responsible for kernicterus.

LABORATORY INVESTIGATIONS

An abnormality associated with cerebral palsy is often documented on head magnetic resonance imaging in the majority of MRI or CT images; therefore, all children with cerebral palsy should undergo a neuroimaging study and, where possible, an MRI study is best. Repeat imaging studies are infrequently needed. Because of the low incidence of metabolic and genetic disorders among children with cerebral palsy, testing for these disorders is indicated only in children for whom the history or clinical examination includes atypical features or if a specific diagnosis is not established with neuroimaging. Children with a brain malformation also warrant consideration for further testing to determine if an underlying abnormal gene or metabolic disorder is present.

Relatedly, the high incidence rates for mental retardation, epilepsy, ophthalmologic defects, speech and language disorders, and hearing impairment make it important that all children with cerebral palsy be screened for these problems.

PROGNOSIS

Generally, prognostic statements are not appropriate until after a number of serial examinations over an extended period of time. Even after repeated examinations, the results of physical therapy cannot be fully anticipated. The child's daily activities may be not limiting to severely limiting depending on mobility and associated conditions. The participation of family members is often essential to quality of daily living and improvement.

Life expectancy of patients with cerebral palsy is related to the pattern of involvement and the severity of motor disability. For example, severe quadriplegia has been associated with a decreased life expectancy. Other significant variables include associated disabilities and availability of quality medical care. The risk of death is highest in the first 5 years of life. As mortality data have become available, it is now clear that with reasonable medical attention, a majority of affected persons will survive into adult life.

SEVERITY OF THE HANDICAP

The Gross Motor Function Classification System (GMFCS) offers an easy and practical way to classify the severity of motor impairment into one of five defined levels of function provided below. Evidence suggests that family reporting may enhance the use of the GMFCS. In recent years, the GMFCS has been applied to develop five distinct motor development curves for use in prognostication. Many other scales and inventories are also in use. These include the modified Ashworth scale and passive range of motion measurements. Proficiency with everyday activities is best predicted by the child's ability to perform gross motor tasks.

COMMON CEREBRAL PALSY SYNDROMES

SPASTIC HEMIPLEGIA

Specific difficulties may not be observed during the first 4 to 6 months of life. Occasionally, children may manifest obvious involvement of one side of the body (hemiplegia) as late as the second year of life. For unexplained reasons, the left brain hemisphere (right side of the body) is affected in two-thirds of patients.

During the examination, the child exhibits impaired large and fine motor coordination, has difficulty moving the hand quickly, and frequently is unable to grasp small items with a thumb-finger pincer grasp. The involuntary palmar grasp reflex that is elicited by placing an object such as a finger or pencil against the palm which normally disappears by age 6 months may continue to be present. The range of elbow straightening (extension) may be decreased. Attempts at reaching for objects may be accompanied by movement marked by continous slow, sinuous, writhing movements that are more pronounced in the hands and performed involuntarily (athetosis) posturing with flexion of the wrist and hyperextension of the fingers (avoidance reaction). Facial involvement is uncommon.

Children with hemiparesis may have a gait that is characterized by leg swinging outward in a circular motion (circumductive gait) with each step. Most commonly the child walks on the toes. In contrast with the leg, the affected arm usually moves less than normal and does not swing in the normal reciprocal motion during walking. The feet are extended and are turned inward (equinovarus position). Weakness of the foot and lack of full range of motion of dorsiflexion often are present. Further evidence of brain movement 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 lower arm and hand or lower leg and foot, may be present. The presence of growth impairment may be demonstraed when the thumb and thumbnail of the affected side are compared with the normal side—the abnormal side is smaller. Discrepant growth of the leg may result in significant difficulties during walking, leading to orthopedic problems involving the upper leg and the lower spine.

SPASTIC QUADRIPLEGIA

Spastic quadriplegia is identified by a generalized increase in muscle tone in both legs and arms. The legs are involved more than the arms and limb movement is usually decreased. Posturing may be evident in early infancy and may persist through the first year of life. In severe dysfunction, movement of the head is accompanied by forced straightening (extension) of the arms and legs, with posturing of the trunk and extremities lying in rigid straightening (extension) with arms internally rotated at the shoulders with the knees and hips rigidly held extended with flexion of the ankles and toes.

Weakness of the speech and swallowing muscles may occur as a result of bilateral brain dysfunction and may produce difficulties with swallowing and articulation. The incoordination of the swallowing muscles may predispose the patient to recurrent pneumonia during the first years of life.

Neurologic examination demonstrates marked stiffness (spasticity) and accompanying signs of brain and spinal cord involvement, including overly brisk reaction when the tendon is struck with a reflex hammer, involuntary rapid up and down movements of the foot when the feet are sharply pushed up extending the Achilles tendon (clonus), and the reflex movement of the big

toe upward when the sole is stroked. Weakness of upward movement (dorsiflexion) of the feet associated with equinovarus deformities is common. Marked stiffness of the hip muscles may lead to displacement of the head of the femur in relation to the hip joint (acetabular joint). Imaging may be necessary to diagnose the abnormal positioning of the head of the femur in relation to the hip socket. 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 often reveals greater visual deficiencies in these children than in children with athetoid cerebral palsy. 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.

SPASTIC DIPLEGIA

Spastic diplegia is diagnosed by the presence of leg spasticity and often some degree of arm and hand involvement. Prematurity is a very frequent cause of spastic diplegia. Unfortunately, it appears that in recent years, the survival of very small preterm infants has resulted in more severely neurologically involved survivors. Preterm infants have a disproportionate share of the cases of spastic diplegia but may manifest any cerebral palsy subtype.
Some infants with spastic diplegia manifest failure of muscle coordination (ataxia) after further maturation. These infants have a great increase of muscle tone of the leg muscles and accompanying problems with coordination and strength; involvement may be asymmetric. When a small child is held in the vertical position by the examiner and the soles of the feet are lightly bounced on the examining table, the legs may be held together (scissoring) and reflex extension of the legs (extensor thrust) occur. The feet also are held in position with the foot extended and the feet turned inward (equinovarus). Further examination reveals weakness of bringing the foot to the shin (dorsiflexion). In older children, this same spasticity results in toe-walking.

As expected, neurologic examination discloses brain control involvement that is demonstrable in the legs (e.g., hyperactive deep tendon reflexes, bilateral ankle clonus, extensor toe signs)[see above]. Striking spasticity of the hip muscles may lead to slipping of the femur from the hip joint with associated breakdown of the hip joint and further restriction of motion. Imaging studies may be necessary to exclude the abnormal positioning of the head of the femur bone.

The arms may be mildly affected. The child may extend or flex the arms in unusual fixed positions while walking. The expected reciprocating arm swinging movements during walking may be clumsy or both arms may be held flexed at the elbows. Affected children also may extend their arms, rotate their hands inward, and clench their fists during running. Associated athetosis makes this latter posturing more likely. Superficial skin blood supply inadequacy, often manifested by cold extremities and variable and sometimes unpredictable patterns of sweating, may trouble the patient.

After a period, usually up to 2 years in moderately involved children, spasticity is increasingly associated with contractures that hold the hips in flexion, knees in flexion, and the feet in an equinovarus position. The feet also are held in position with the foot extended and the feet turned inward (equinovarus position). For reasons that are unclear because of faulty brain control, notable retardation of growth of leg length may occur.

EXTRAPYRAMIDAL CEREBRAL PALSY

Extrapyramidal cerebral palsy can be divided arbitrarily into two primary clinical subtypes—choreoathetotic and dystonic. Choreoathetotic movements are involuntary, rapid, jerky movements that continue for long periods. Dystonic movements are those that occur because of distortion of voluntary movements. In both types, patients are unable to smoothly perform meaningful movements because of interfering movements and involvement of inappropriate contractions of muscles that ordinarily oppose each other (agonist and antagonist muscles) and thus provide for smooth movements. Extrapyramidal cerebral palsy involves defects of posture and involuntary movement (e.g., athetosis, ballismus, chorea, dystonia); increased muscle tone usually is associated with these conditions and is of the “lead pipe” or rigid variety.

CHOREOATHETOTIC CEREBRAL PALSY

Choreoathetotic cerebral palsy is characterized by large-amplitude, involuntary movements. The most obvious and dominating movement component is athetosis. Athetosis results in slow, writhing involuntary movements and usually involves the lower (distal) limbs; chorea is present in variable degree. Tremor, myoclonus, and even some element of dystonia also may be evident.

Other common features are finger and toe extension and rotation of the limb along 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 choreiform movements can be characterized as asymmetric, fleeting, uncoordinated, involuntary contractions of individual muscle groups. The combination of athetoid and choreiform movements results in a pattern of distal extremity movement, ongoing increase of muscle tone (hypertonia), and rotary writhing movements of the limbs.

Athetotic posturing may be evident in the first year of life when the child begins to reach for objects. The movements, as is generally true of most involuntary movements, are not present during sleep. Movements are more prominent during stress or illness and their intensity varies from day to day.

As expected from the pathologic findings, evidence of defective brain control (upper motor neuron unit) (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 that is characterized by great variability in rate and explosive changes in volume.

Ballismus, a very rare movement disorder in which the arms and legs are violently flung about, is possibly 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

Dystonic movements are the result of disordered tonicity of muscle. 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 is 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; 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.

HYPOTONIC (ATONIC) CEREBRAL PALSY

Infants with hypotonic (extreme floppiness) cerebral palsy almost always have associated leg weakness. Although hypotonic, the arms may manifest 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 many possibilities. Most children with generalized hypotonia have so-called central hypotonia that results from brain dysfunction, resulting from inadequate control of the movement circuits. Others, with absent or hypoactive deep tendon reflexes, may have involvement of the lower motor neuron unit (i.e., cord neurons, nerve to the muscle, connection of the nerve to the muscle, and the muscle). Extrapyramidal (choreoathetotic and dystonic) cerebral palsy may be preceded by a hypotonic phase.

Atonic (greatly decreased muscle tone) cerebral palsy is relatively uncommon compared with other forms of cerebral palsy; it often is associated with slow development of motor milestones and the presence of normal or extremely reactive deep tendon reflexes.

Although in the past it has been thought that muscle tone almost always increases with maturation in this form of cerebral palsy, it has become clear 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 complex circuits controlled through the spinal cord that portions of the brain (e.g., motor cortex, thalamus, basal ganglia, vestibular nuclei, reticular formation, cerebellum) modify muscle tone, with ensuing hypotonia.

ATAXIC CEREBRAL PALSY

The least common form of cerebral palsy is the ataxic form. Ataxia is defined as failure of muscle coordination with irregularity of muscular action. It sometimes coexists with spastic diplegia. This form usually is associated with other movement abnormalities; however, the diagnosis is used only when the main finding is ataxia. Most commonly ataxia is the result of dysfunction of the cerebellum. 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 and frequent falling. Compromise of writing skills and other skills that demand good fine

motor coordination often adversely affects 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 with ataxia, 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 (a part of the cerebellum) may not give rise to cerebellar symptoms in certain congenital conditions, whereas lack of development of the vermis may be associated with nonprogressive 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 corroborated by CT studies.

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, it is common for patients in whom choreoathetosis predominates to also manifest changes that occur with injury to higher portions of the brain. These patterns of motor dysfunction are the result of compromise of large areas of the brain.

Characteristics of these patients are discussed in the sections describing individual types. Most patients can be categorized into the types discussed on the basis of most obvious manifestations.

THERAPEUTIC INTERVENTION

Most commonly, treatment of cerebral palsy will require a number of professionals to provide various treatments, ranging from medications to surgery to physical therapy.
Medications to relieve spasticity include baclofen (Gablofen), diazepam (Valium), and dantrolene (Dantrium). Baclofen may also be injected into the spinal cord canal after a surgical procedure provides access. There is no evidence that special diet or dietary supplementation is of value.
Injections of botulinum toxin A (Botox) in individual muscles may also make the muscle tone more normal. The effects of injections are not permanent and injections need to be repeated in approximately three months.
Children with severe contractures may benefit from surgery of the joints and bones to repositions arms and legs. Contractures may lead to shortening of the tendons or muscles and some operations may substantially lengthen the tendons or muscles.
The neurosurgical procedure in which nerves are cut, known as selective dorsal rhizotomy, may improve severe muscle spasticity.
Physical therapy is very often helpful. Various muscle exercises may improve strength, development and balance and decrease spasticity. Many of these exercises can be repeated at home to maintain therapy continuity.
Beneficial equipment is available. Walkers, braces, splints, canes, power

wheelchairs, and special seating apparatus may be of immense help. Occupational therapy may be valuable to increase skills that enhance abilities that allow independent participation in everyday activities.

PROGNOSIS

Life expectancy of patients with cerebral palsy is related to the type and severity of motor disability. Severe quadriplegia has been associated with a decreased life expectancy. Other significant variables include associated disabilities and availability of quality medical care. The risk of death is highest in the first 5 years of life. As mortality data have become available, it has become clear that with reasonable medical attention, a majority of affected persons will survive into adult life.
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 as described above, and the degree of associated deficits in intelligence, sensation, and emotional adjustment. Cognitive level is difficult to assess in the young child with motor impairments but can be gauged even in the severely affected. 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 and some benefit 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 is narrower 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% of children with hemiplegias also 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 can be 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% of children with spastic diplegia learn to walk, commonly by the age of approximately 3 years, but gait often is abnormal; some children require assistance devices such as crutches. Hand activities are often involved to some degree, although the impairment may be subtle. Abnormalities of extraocular movement are relatively common.

Of children with spastic quadriplegia, 25% require total care; approximately 33% walk, usually after the age of 3 years. Intellectual function is often 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 problems.

GLOSSARY/DEFINITIONS

- Ataxia – Failure of muscle coordination, irregularity of muscular action.
- Atonic – Lack of normal muscle tone.
- Asphyxia – Deleterious changes in tissue due to relative lack of blood oxygen and abnormal increase in blood carbon dioxide concentration.
- Athetosis – Dyskinesia marked by continual involuntary, slow, sinuous, writhing movements, especially severe in the hands.
- Ballismus – Very rare movement disorder in which the arms and legs are violently flung about,
- Chorea – Rapid, continuing, jerky, complex, dyskinetic movements that are involuntary.
- Dorsiflexion – Bending of the hand or foot toward the arm or leg.
- Dyskinesia (dyskinetic) – Distortion or impairment of voluntary movement.
- Dysmetria – Relative inability to estimate distance during movement; usually takes place in the arms and hands.
- Dystonia – Dyskinetic movements due to abnormal changes in tone of muscle.
- Encephalopathy – Disease of the brain.
- Equinovarus – Downward extension of the foot that is also turned inward (equinovarus position).
- Hemianopsia – Impairment of the visual field in both eyes. Patient cannot see images to the right or left in both eyes.
- Hyperreflexia – Abnormal increase in deep tendon reflexes. Deep tendon reflexes are elicited by the striking of the muscle tendon with a reflex hammer.
- Hypertonia – Increased muscle tone from expected (See muscle tone).
- Hypotonia – Decreased muscle tone from expected (See muscle tone).
- Hypoxic-ischemic brain injury – Brain injury that occurs when either oxygen supply or blood flow is curtailed, or both.
- Kernicterus – Abnormal neurologic findings in newborn associated with increased levels of bilirubin (jaundice) and resulting specific injuries of the brain.
- Lissencephaly – The absence of normal brain gyri due to congenital abnormality.
- Necrosis – Cell death and its accompanying changes.
- Perinatal – The time just before and just after birth.
- Polymicrogyria – Developmental abnormality manifested by many abnormally small gyri on the surface of the brain.
- Primitive reflexes – Reflexes in the newborn that are elicited with various maneuvers that are mediated by the brainstem and that disappear with maturation.
- Muscle Tone – Resistance of muscles to limb movement by examiner when patient is completely relaxed.
- Nystagmus – Rapid, rhythmic movements in any direction of the eyes that are involuntary.
- Parachute response – A primitive response that is elicited by thrusting the infant toward a flat surface with subsequent reflex straightening (extension) of the arms. The thrusting maneuver is terminated before the infant strikes the surface or when the arms are extended.
- Periventricular leukomalacia – Pathologic disssolution of the brain white matter adjacent to the ventricles in newborns.
- Polymicrogyria – A congenital abnormality of development that consists of

formation of multiple, smaller than normal brain gyri.

- Quadriparesis – Weakness of all four limbs, usually with greater involvement of the legs.
- Retrocollis – A condition in which the head is involuntarily extended backwards.
- Tone – See Muscle Tone
- Traction response – Primitive reflex that is elicited by grasping the supine infant's hands and beginning to pull the infant into the sitting position with subsequent flexion of the arms and trunk.

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