REHABILITATION MEDICINE Adding Life to Years

Rehabilitation in Cerebral Palsy

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Cerebral palsy is the most frequent physical disability of childhood onset. Over the past four decades, prevalence has remained remarkably constant at 2 to 3 per 1,000 live births in industrialized countries. In this article I concentrate on the rehabilitation and outcome of patients with cerebral palsy. The epidemiologic, pathogenetic, and diagnostic aspects are highlighted briefly as they pertain to the planning and implementation of the rehabilitation process.

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The clinical syndromes of cerebral palsy share three unifying diagnostic criteria: The leading symptom is a *movement disorder* resulting from *nonprogressive brain damage* that has affected the *immature central nervous system.*[1] The possibility of deficits in areas other than motor function is implicit in the underlying disorder. According to this definition, the time of brain damage may be prenatal, perinatal, or postnatal. Most cases are related to prenatal or perinatal antecedents or both.[1-3] Several large-scale studies indicate that in both term and preterm infants, prenatal factors have a more significant role than was previously recognized.[1-4]

In premature infants, the risk of cerebral palsy is inversely related to birth weight. A residual motor deficit following a central nervous system injury or illness sustained between 4 weeks and 5 years of age can be classified as postnatal cerebral palsy after the acute stage has passed.[1] Postnatal causes are responsible for 10% to 15% of cases. No cause is apparent in 20% to 30% of cases wherein symptoms occur early.[1]

Classification

The classification of cerebral palsy is based on the clinical signs of motor dysfunction.[1] Spasticity is the presenting sign in most cases (Table 1). Diplegia is the accepted classification when the lower extremities are affected and usually follows hemorrhagic or hypoxic periventricular lesions of prematurity.[2] As a rule, in quadriparetic distribution of spasticity there is a greater impairment of the legs. By convention, double hemiplegia designates quadriparesis with more involvement of the arms. Dyskinetic clinical types include several movement disorders; athetosis and dystonia are the most common. Preventive treatment of neonatal isoimmunization and bilirubin encephalopathy has reduced the occurrence of typical kernicteric athetosis and the relative frequency of dyskinetic cerebral palsy in general.[5] Other clinical types represent a small number of cases with predominant or mixed signs of various movement abnormalities. The possibility of familial and progressive neurologic disease must be ruled out when ataxia with or without spasticity is the leading sign.[1]

TABLE 1.—*Clinical Classification of Cerebral Palsy*

Clinical Type	*Relative Frequency, %*
Spasticity	75-85
Hemiparesis	10-15
Diplegia	35-40
Quadriparesis	25-30
Dyskinesia	5-10
Athetosis	5-8
Dystonia	2-3
Choreiform, ballistic, tremor	Rare
Other types	10-15
Spastic, athetoid	5-10
Rigid, spastic	3-5
Ataxic diplegia	Rare
Ataxia	Rare
Atonia (hypotonia)	Rare

Diagnosis

The early detection of cerebral palsy has some diagnostic uncertainties. Several independent long-term studies have shown that initial neurologic abnormalities may resolve and have demonstrated the need for caution about unjustified labeling of young infants as having cerebral palsy.[1,6,7] On the other hand, definitive signs sometimes do not become evident until volitional movement control begins to emerge. A suspicion of cerebral palsy is usually confirmed during the second half of the first year. Mild cases with subtle symptoms, however, may escape detection until the second year when delayed or abnormal standing and walking become noticeable.[8]

Signs and symptoms that should arouse suspicion are abnormalities of tone, posture, and infantile reflexes, delayed motor development, and atypical motor performance.[9] Hypotonia is a precursor of spasticity or athetosis. A long-standing hypotonic stage or definite spastic hypertonicity around the age of 6 months suggests a severe motor deficit. A fisted hand with thumb adduction, scissoring of the legs, and a tendency for opisthotonus are typical postural deviations. Persistent Moro and obligatory tonic neck reflexes are examples of infantile reflex abnormalities. Late sitting is often the first symptom of parental concern. Motor delay tends to increase with successive milestones. Hand preference during the first year is an indication of impaired coordination of the contralateral arm in hemiparesis. Crawling in a prone position rather than on hands and knees or toe walking are well-known signs of motor deficit in the lower limbs.

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TABLE 2.—*Disabilities Associated With Cerebral Palsy*

Type	Estimated Frequency, %	Clinical Types With Highest Relative Frequency
Dysarthria	20-30	Athetosis Spastic quadriparesis
Visual deficits	60	Spastic diplegia
Strabismus		Spastic quadriparesis
Hemianopsia		Spastic hemiparesis
Refractory errors		Spastic/dyskinetic (2:1)
Decreased visual acuity		
Blindness		
Hearing impairment	25-30	Athetosis (kernicterus 75%; other 25%)
Sensorineural		
Mental retardation	50	Atonic and rigid types Spastic quadriparesis
Perceptual dysfunction	40-50	
Learning disability		

Rehabilitation

The rehabilitation of children with cerebral palsy requires a developmentally oriented approach that is not limited to the care of the motor disorder.[10] The therapeutic team consists of the family, physicians, and other contributing professionals who must have a clear understanding of attainable goals. Taking advantage of functional assets, promoting compensatory adaptations, and preventing the secondary consequences of disability are principles of rehabilitation that are distinguished in concept but inseparable in practice. Changing concerns from childhood to adult life call for adjustments in treatment and priorities.

In addition to a movement disorder, other functional deficits may reflect central nervous system damage.[5] Table 2 shows the estimated overall rate of associated disabilities and their relative frequency in different clinical types. Comprehensive rehabilitation entails assessing all aspects of function and appropriate remediation.

Motor Disability

Early intervention programs address developmental deviations that result from associated central nervous system deficits or are imposed by impaired motor function.[9] Public Law 99-457 (Education of the Handicapped Act of 1986) is a federal mandate to provide such programs for high-risk infants.

Therapies for neuromuscular disorders in infants and young children incorporate handling, positioning, and play to challenge postural and movement responses that are prerequisites for gross and fine motor milestones. Feeding difficulties require the facilitation of oral motor coordination to improve swallowing.[11] Instructing the family in modified ways of physical handling extends therapeutic efforts to everyday activities at home.[11,12] Therapy becomes more structured as children learn to follow instructions and are able to cooperate. During the preschool years, the functional goal is to achieve the most effective mode of locomotion and self-care skills, with assistive devices if necessary.

Physical therapy is a widely used treatment method. Among the techniques proposed for patients with cerebral palsy, some remain controversial on both theoretic and clinical grounds.[11,13,14] Controlled trials on the effectiveness or comparative efficacy of various techniques are scarce.[15] The great diversity in physical and associated deficits and in the consequent natural course poses difficulties for designing research studies. Clinical experience indicates that no therapeutic method can produce striking changes in the functional potential determined by a central nervous system deficit. The contribution of physical therapy is to ameliorate the adverse effects of disuse, inexperience, and secondary complications. The importance of these benefits, however, should not be underestimated.

Compensatory functional training includes a range of rehabilitation possibilities that can be illustrated by several examples. Environmental modifications to ensure access, suitable clothing, and learning to use simple adapted implements for feeding, writing, and other daily tasks are practical approaches to independence that occupational therapy offers.[5,12,16] Although assistive devices such as crutches, walkerettes, and wheelchairs have been used for a long time, rehabilitation engineering has revolutionized the technology for disabled persons. The light weight and versatility of new wheelchair designs allow greater mobility, including participation in adapted sports. An important development for patients with cerebral palsy is the construction of wheelchair seats that incorporate biomechanical features to alleviate abnormal tone and posture.[5,9,17] When upper extremity impairment necessitates, motorized wheelchair models are available from pediatric to adult sizes. Children with adequate cognitive, visual, and spatial discrimination abilities can learn to operate a motorized device under adult supervision as early as 3 years of age. Electronic and computer technologies have created a range of possibilities to help patients overcome physical restrictions in activities of daily living, education, work, and leisure.[9,17] Economic constraints are the primary obstacle that limits the availability of such devices.

Preventing musculoskeletal complications, which can further compromise function, constitutes an additional aim of treatment.[9] Abnormal tone, movements, and posture create a risk of soft tissue contractures and bone deformities. The propensity for these complications is greatest during the rapid phases of growth in childhood, but an insidious progression may continue after skeletal maturity.[18] Anticipatory treatment is the best preventive measure. Options include range-of-motion exercises, splinting, braces, and surgical procedures as warranted by the clinical findings and course.

Therapy is most intensive from infancy to early school years because most children reach their expected functional capacity in ambulation and daily activity skills by school age.[19,20] As formal therapy is gradually tapered, consistent practice at home and school and in other functional situations must continue to increase motor proficiency, endurance, and the self-initiated acquisition of new skills. Adapted physical education and sports, aside from their benefits on motor abilities, provide enjoyment, companionship, and a sense of accomplishment, which are important for the development of confidence and self-image in all children and adolescents.[10] A periodic reinstitution of formal therapy may be necessary for defined reasons, such as to regain preoperative function after immobilization for orthopedic procedures or to learn new skills that have become appropriate and feasible for the child's developmental level and physical abilities.

Lower extremity orthoses or splints are used to improve function—for example, toe walking—or to prevent deformity in a child who does not ambulate.[5,17] Extensive bracing does not enable a child with severe motor disability to attain useful walking. Despite these considerations, the treatment team

might elect to use orthoses for passive standing or exercise ambulation in a bright, well-motivated youngster. The family, however, must understand clearly the limited purpose of these attempts.

Orthopedic surgery plays an important role in the treatment of physical disability.[5,21] Correcting abnormal posture and gait enhances the efficiency of walking. Nonambulatory children are more likely to need surgical procedures for preventive reasons, such as the release of spastic hip flexors, adductors, and femoral osteotomy for hip subluxation, which would otherwise progress to painful dislocation.

Selective posterior rhizotomy is a neurosurgical procedure suggested to decrease spasticity.[22] Phenol blocks of the motor points or motor nerve branches of selected muscles produce a reversible chemical neurolysis and temporary reduction of spastic hypertonia lasting four to six months.[23] Dantrolene sodium and diazepam are antispasticity drugs that are helpful in some patients.[23]

Associated Disabilities

Speech and language development need close monitoring because effective communication is one of the most valuable functional assets.[5,9] All children with cerebral palsy should have an audiologic examination and a timely correction of hearing impairments that may have detrimental effects on language acquisition. Sensorineural hearing loss is the characteristic auditory deficit and is particularly common in patients with kernicteric athetosis. Central language disorders are more likely related to cognitive dysfunction because young children show good recovery from aphasia after suffering damage to the dominant hemisphere. A subtle impairment of language competence may persist despite fluent speech, however, and also has been reported with congenital right hemiparesis.[9] Speech defects occur in athetosis and bilateral spastic paresis when pseudobulbar palsy affects the coordination of muscles that participate in phonation and articulation.[5,9] The resultant speech impairments range from mild articulation disorder to severe dysarthria with a total lack of functional verbal communication.

Therapy for speech and language deficits should commence as soon as the deficits are detected.[5] An important advance in the treatment of severe speech defects is nonverbal communication.[5,9,24] The simplest initial method is the simultaneous presentation of several objects or an improvised communication board of pictures from which a child can select the appropriate response by eye gaze, pointing, or any other consistent signal. When education and social interaction demand an expanded repertoire of communication skills, manually or electronically operated voice and printed word-symbol communicators offer various choices.[5,9,17,24] The potential for nonverbal communication should be explored when speech is delayed to identify children who have sufficient cognitive language ability to master this mode of interaction.

Feeding difficulties and salivation are additional signs of pseudobulbar palsy. Skillful feeding and carefully selected food textures facilitate oral motor function.[5,9,11,12] In almost all patients, dysphagia and drooling are reduced with time. In severe cases, inadequate nutritional intake or recurrent aspiration necessitates temporary or permanent feeding gastrostomy. For the socially embarrassing handicap of drooling, there are behavioral, pharmacologic, and surgical treatment methods.[5,9]

The possibility of a seizure disorder must be investigated in every patient.[5] Although seizures can occur in any clinical type of cerebral palsy, spastic hemiparesis and quadriparesis have the highest incidence rates. The selection of anticonvulsant medications depends on the clinical seizure manifestations, with consideration given to side effects on behavior and alertness.

A complete diagnostic assessment includes the evaluation of visual function.[5] Strabismus, most common in diplegia and quadriparesis, refractory errors, and retinopathy of prematurity require ophthalmologic treatment. Severe visual impairment or blindness has an adverse influence on motor development and ultimate level of physical function.

Patients with spastic quadriparesis, rigid or atonic clinical types, have the highest relative frequency of mental retardation and severe intellectual deficits.[5,9] In general, cognitive impairment is mild and occurs at a lower rate in those with hemiparesis and diplegia. Discrete lesions of the basal ganglia spare intellectual function in those with athetosis following kernicterus, but this may not be the case when hypoxic encephalopathy is a contributing factor.[5] Dysfunction of central perceptual processing may be a cause of learning disability despite satisfactory intellectual function.

Planning for appropriate educational placement begins before a child reaches school age and involves the cooperation of the family, educators, and the rehabilitation team.[5] Psychological assessment should be obtained before the child enters school. The psychologist must have experience with physical disabilities because the standard test batteries rely on motor items, verbal responses, and timed performance, which place children with impaired hand dexterity or speech at a disadvantage. In selecting suitable schooling, intellectual function is the deciding factor.[5,9] If necessary, adaptations in classroom environment should accommodate the physical disability. The Education for All Handicapped Children Act of 1975, PL 94-142, includes provisions for the least restrictive environment pertaining to educational goals. It must be stressed that education is a high priority for disabled children, whose future employment opportunities will be contingent on scholastic competence.

Psychosocial Issues

Initial counseling of the family revolves around diagnosis, outcome, and treatment.[5,9] Discussions about these issues and about special concerns of child rearing should be ongoing during the course of treatment. At the appropriate age, discussion and counseling should involve the child. Education and vocational preparation come into the foreground by school age. Concern with the physical disability should not distract attention from the emotional and social needs of childhood and adolescence. Disabled youngsters need the same variety of life experiences as all other children to develop emotional resilience, personal determination, and social skills.[5,9,10,18] These traits of behavior, though less tangible than the physical aspects of function, are essential for success in adulthood and originate from interaction between the child and family.[25]

Predicting Outcome

To predict the functional outcome and long-range achievement, the physician must weigh many diverse factors. Physical and intellectual abilities determine the highest possible function, but long-term studies show that the greatest

fulfillment of potential depends on psychosocial characteristics as well.[5,9,19,25]

Expectations in motor function, considering solely the physical disability, vary by the clinical type of cerebral palsy.[5,19] Children with hemiparesis walk between 1½ and 3 years of age. Although there are individual differences in assistive function of the affected hand, hemiparesis does not interfere with independence in usual daily activities. The outlook for ambulation is less uniform in children with bilateral lower extremity spasticity or dyskinesia. Independent sitting by the age of 2 years is a favorable sign for the eventual ability to walk, whereas an inability to sit at 4 years of age indicates a poor prognosis for ambulation.[5,19,20] Between 80% and 90% of children with diplegia, approximately half of those with quadriparesis, and three fourths of those with dyskinesia eventually are able to walk. The attainment of sitting by age 1½ to 2 years has a good correlation with community ambulation. The walking ability of children who learn to sit between the ages of 2 and 4 years probably will be limited to short distances outdoors or household ambulation with assistive devices or partial wheelchair use. The outlook for ambulation is generally good in children with ataxia. Independence in activities of daily living is expected of children with diplegia and those with quadriparesis who walk. Partial self-sufficiency in daily skills is a possibility for some children with quadriparesis who can use wheelchairs. Approximately 25% of children in this group require complete help. Dyskinetic and ataxic movement disorders may be more disabling in fine hand dexterity than in walking. The few patients with rigid or atonic clinical type have a poor overall functional prognosis. In general, mental retardation delays but does not exclude ambulation if the physical disability permits it. On the other hand, expectations in skills of daily life must be adjusted to a level commensurate with intellectual competence.

The estimated employment rate among persons with cerebral palsy, some in sheltered work, is approximately 50%.[5] One study compared vocational predictions at ages 7 to 16 with adult achievements.[25] Most young adults achieved or exceeded the predicted vocational status; only 28% did not. These results illustrate that vocational outcome depends on a complex interaction of many factors. Strong family support and personal determination were found to be characteristics common to those who did better than expected. Integrated education, vocational counseling, community-based independent living programs, and technologic advances were additional factors contributing to success according to this and other studies.[5,25]

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