EITI Newsletter

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Rose F. Kennedy Center ■ University Affiliated Program

CEREBRAL PALSY

Cerebral palsy continues to be the most frequent childhood motor disability. Cerebral palsy can be defined as a disorder of movement and posture which is a manifestation of a non-progressive lesion or an injury sustained during the period of brain growth or due to a developmental deficit of the brain.

The incidence of cerebral palsy has remained constant at a rate of 2-3/1000 live births during the last four decades. The brain lesions resulting in cerebral palsy may occur during prenatal, perinatal, and postnatal periods. These causes include maternal infection, congenital malforma-tions, trauma, exposure to teratogens, prematurity, and intracranial hemorrhages. The most common causes of cerebral palsy are prenatal in origin.

Cerebral palsy can be classified as spastic, dyskinetic, and mixed. The spastic type, which is the most common form, is further subdivided into: 1) spastic diplegia in which both lower extremities are involved; 2) spastic quadriparesis in which all extremities are spastic, lower extremities more severely; 3) spastic triplegia involving both lower extremities and one arm; 4) spastic hemiparesis, involving one side of the body, arm and the leg (arm is more involved) and; 5) spastic monoplegia involving one limb, usually mildly, and very often is a misdiagnosed hemiplegia.

Dyskinetic types of cerebral palsy are subdivided into: 1) athetosis, characterized by slow writhing movements of the face and distal extremities; 2) dystonia characterized by rhythmic twisting movements of trunk and proximal limbs with changes in muscle tone and; 3) chorea which are rapid irregular jerky movements of face and extremities (uncommon).

The mixed type of cerebral palsy is subdivided into spastic athetoid and spastic ataxic or ataxic type with unsteadiness, nystagmus, dyskinetic and uncoordinated movements. Other rare types of cerebral palsy include atonic and rigid types.

Diagnosis

Early symptoms and signs that should arouse suspicion of cerebral palsy are delayed motor development, abnormal muscle tone and posture, and abnormal movement patterns. Since motor control is acquired gradually during the first year of life, it is difficult to recognize motor deficits at birth or during the first few months of life, unless the abnormalities are significant. As the neuromuscular deficit and the abnormal movement patterns continue to evolve, the diagnosis can be usually confirmed at the end of the first year of life. In mild cases, however, the diagnosis may be overlooked until much later when the abnormalities in walking and significant developmental delays are noted.

The diagnostic work-up of cerebral palsy consists of a detailed and careful history, physical examination, laboratory studies, and neurodiagnostic imaging, as well as visual and auditory evoked potentials, EEGs and electrophysiological studies, when indicated. The history should include prenatal, perinatal, and developmental history, family history, feeding history, detailed medical history and a review of systems. The physical examination should include assessment of the musculoskeletal system, neurological examination, including muscle tone, posture, both normal and abnormal reflexes as well as gait, mobility and functional and developmental assessment. Blood and urine samples should be obtained to rule out metabolic and genetic diseases. Other tests should include thyroid function, chromosomes, organic and amino acids, lactate and pyruvates, when appropriate.

Neuroimaging techniques should be done (MRI, CT, Cranial Ultrasound) to obtain information regarding intracranial hemorrhages, congenital malformations and periventricular leukomalacia. Evoked potentials may provide information regarding visual and auditory pathways.

Clinical Course

The clinical course in cerebral palsy is diverse, depending on severity and the clinical manifestations. Clinical findings at the onset or at the time of diagnosis may change over the years due to therapeutic intervention (or lack of it) and due to growth and development of the child.

The child may be hypotonic initially and later become hypertonic or dyskinetic. The clinical picture may be influenced by secondary adverse musculoskeletal changes due to muscle imbalance, abnormal posture and tonal abnormalities. These secondary sequelae tend to occur earlier in children with moderate and severe disabilities and can further affect the functional outcome. Children with spastic hemiplegia may have loss of function in the affected hand, especially if there is sensory impairment in addition to either weakness or spasticity. This can lead to contractures and growth disturbance in the affected extremity. Most children with hemiplegia can be independent in self care but may need adaptive equipment for bimanual skills. Almost all children with hemiplegia walk. They may need orthoses to stretch tight ankles or to support a weak limb. Children may need nerve blocks or surgical intervention to decrease the spasticity and to release tight tendons.

Children with spastic diplegia may have deficits in hand function at the onset. With therapeutic intervention they usually achieve independence in ADL skills. They may achieve lower extremity gross motor skills such as standing and ambulation with intensive therapy, orthotic devices and assistor devices for mobility. Continued spasticity may lead

to contractures of major joints, and abnormalities in posture and gait. Hip dislocation may require surgical intervention. Often medications and surgical procedures are needed to control the spasticity.

Spastic quadriplegia presents with varying degrees of severity in motor deficits, which directly influence acquisition of motor skills and functional independence. Persistent increased muscle tone in the lower limbs may cause hip dislocation, pain, and scoliosis, especially in non-ambulatory children. Associated deficits such as mental retardation, seizures, and oromotor deficits can further compromise the acquisition of functional skills. One fourth of children with spastic quadriparesis are mildly involved with minimal or no functional limitations in ADL and walking. One half of the children have moderate involvement and need assistance in ADL and mobility. One fourth have severe deficits and need total care.

Children with dyskinetic cerebral palsy have tonal abnormalities which may be present for a long time (especially hypotonia) and persistence of primitive reflexes. These children develop athetoid movements in their hands and feet by 18 months which progress to dystonic movements with growth and maturation. Upper extremities are more involved than the lower extremities. About 50% attain independent walking, often after 3 years of age. These children develop adequate upper extremity control for ADL. They may develop scoliosis later on.

Prediction of long-term outcome in cerebral palsy is difficult in the first few years of life. The outcome is influenced by the severity of the motor deficit, presence of associated deficits and effect of intervention. Overall, 75% of children with cerebral palsy walk either independently or with assistance. Failure to achieve independent sitting by 2 years of age and the presence of primitive reflexes at 18 months of age has been shown to have a poor prognosis for walking. Ninety percent of children live to reach adulthood. The presence of mental retardation, seizure disorders and severe motor disability make functional independence less likely.

Management

Management of cerebral palsy consists of treatment of motor disabilities, treatment of associated deficits, promotion of good physical and emotional health, support of the family, provision of appropriate educational and vocational services, and integration of the child/young adult into the community.

Management requires cooperative efforts of the family, physician and other professionals involved in the care of the child. Realistic expectations and goals should be established. A mutual understanding of the problem and an agreement on the plans of intervention is essential. The main goals are to maximize functional skills, foster independence, and prevent or minimize potential complications.

The course of motor development and ultimate functional outcome varies in different types of cerebral palsy, and often within the same type. In many instances a definitive diagnosis cannot be established with certainty until much later. However, the intervention must be initiated when there is evidence of motor delays and or abnormalities of muscle tone.

Public Law 99-457, enacted in 1986, mandates that all states must establish early intervention programs for infants and toddlers at risk or with known developmental delays. These services are family focused and help establish effective parenting skills and effective infant caregiver interaction. Services can be either home based or center based as a part of a multidisciplinary approach. Parents are instructed on handling and positioning as well as feeding techniques. In addition, direct therapeutic intervention is provided to promote normal movement patterns and to enable young children with limited motor abilities to explore their environment. Psychosocial support is provided to improve parents' coping abilities.

Treatment of motor disabilities should utilize well-timed and appropriate therapeutic exercises, orthotic and assistive devices, and timely medical and surgical intervention. There are various types of therapeutic systems utilized in the treatment of motor deficits. There is no one single method that is suitable for all children; therefore the therapy regime should be individualized. In addition to therapeutic exercises, functional training should be an integral part of treatment to promote independence in self care, activities of daily living, and school related activities.

Orthotic devices are prescribed to improve function, to maintain joint ranges or to prevent soft tissue deformities, and are used as an adjunct to physical and occupational therapy. For example, an ankle foot orthosis is used to stretch a spastic and tight heel cord, to maintain the ankle in neutral position for passive standing or to improve the walking pattern in a child with ankle deformities. Orthotic devices should be reevaluated on a periodic basis to ensure the proper fit and to assess the need for continuation.

Assistive devices and durable medical equipment are used to attain functional self care, communication, mobility, and job skills which may not be otherwise possible. When choosing equipment it is important to consider the functional goals, prognosis, patient and family needs, and the cost effectiveness of the device.

Well timed and judicious use of orthopedic surgical procedures may be considered to improve function and physical appearance as well as to prevent or correct deformities. Soft tissue releases, tendon lengthening, and transfers are often used to decrease spastic muscle imbalance or to prevent secondary skeletal complications. Soft tissue release procedures are not always successful and soft tissue contractures may progress to bony abnormalities which may require surgery. Upper extremity orthopedic procedures are less frequent and are often considered for improved appearance rather than improvement of function.

An integral part of managing motor disabilities is treatment of spasticity and dystonias. There are many modalities available, including pharmacological agents, neuromuscular blocks, neurosurgery, use of therapeutic heat or cold, biofeedback, functional and therapeutic electrical stimulation, and inhibitory casts and splints.

The oral antispastic medications frequently used are benzodiazepines, oral Baclofen, Dantrolene Sodium and Tizanidine. They decrease spasticity but do not improve function. Long-term use may be limited due to the side effects.

Baclofen administered via a programmable pump implanted surgically in the anterior abdominal wall and connected with a catheter has been shown to improve function and reduce spasticity. The advantages are a very low dose requirement and easy administration of

medication directly into the spinal canal. The disadvantages are the prohibitive cost of the equipment and medication, and the possible complications such as infections, CSF leak and kinking of the catheter. Long-term efficacy needs to be established in children.

Intramuscular blocks using Botulinum Toxin A (Botox) reduces spasticity for a 3-4 month period. This improves range of motion and reduces deformities. It also allows therapeutic intervention to be effective in increasing muscle strength and motor control.

Several neurosurgical procedures have been used in the treatment of spasticity. Selective dorsal rhizotomy of L2-L5 spinal rootlets followed by intensive physical therapy has been successful in decreasing spasticity as measured by the modified Ashworth scale. In children who are walking, gait analysis has shown an increase in stride length and an improvement in hip and knee range of motion. In non-ambulatory children the decrease in spasticity results in easy management in self care skills and in appropriate positioning. Further studies are needed to assess the long-term efficacy of this procedure.

Management of cerebral palsy should also include periodic evaluation of children to assess their physical growth and visual and auditory functions. Management of associated conditions such as seizures, dental abnormali-ties and feeding difficulties, visual and hearing impairment is essential.

Appropriate educational placement should be obtained, with assistance and classroom adaptation to compensate for the disability. Counseling for emotional and social adjustment should be implemented. The ultimate goals should be maximizing all aspects of function and integration of the child into the community.

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