Cerebral Palsy–Definition, Classification, Etiology and Early Diagnosis

Chitra Sankar and Nandini Mundkur

Department of Developmental Pediatrics, Bangalore Children's Hospital, City Centre, Bangalore, India

Abstract: Cerebral palsy is a common neurodevelopmental condition encountered by pediatricians. The condition may present itself in many diferent clinical spectra. The etiological and risk factors are many and an awareness of the interplay of multiple factors in the causation of CP is crucial. In many cases, the cause of Cerebral palsy may not be apparent. Cerebral palsy is invariably associated with many deficits such as mental retardation, speech and language and oromotor problems. A thorough neurodevelopmental assessment of the child with Cerebral Palsy should include evaluation of associated deficits so that a comprehensive early intervention program an be planned and executed. **[Indian J Pediatr 2005; 72 (10) : 865-868]** *E-mail : bchrc@vsnl.com*

Key words: *Cerebral palsy; Spasticity; Hypotonia, Associated deficits; Infanib.*

Cerebral palsy is a common developmental disability first described by William Little in the 1840s. The condition poses considerable diagnostic and therapeutic challenges to the physician with degree of involvement ranging from mild with minimal disability to severe, associated with several co morbid conditions. It is one of the three most common life long developmental disabilities, the other two being autism and mental retardation causing considerable hardship to affected individuals and their families.

DEFINITION

Cerebral palsy is primarily a disorder of movement and posture. It is defined as an "umbrella term covering a group of non-progressive, but often changing, motor impairment syndromes secondary to lesions or anomalies of the brain arising in the early stages of its development".[1] It may be stated as a static encephalopathy in which, even though the primary lesion, anomaly or injury is static, the clinical pattern of presentation may change with time due to growth and developmental plasticity and maturation of the central nervous system.

INCIDENCE

CP is a common problem, the worldwide incidence being 2 to 2.5 per 1000 live births.[2] When Little first described CP, he attributed the cause of CP to birth trauma and this view has persisted for several decades. Recent advances in neonatal management and obstetric care have not shown a decline in the incidence of CP.[3] On the contrary, with a decline in infant mortality rate, there has actually been an increase in the incidence and severity of CP. The incidence in premature babies is much higher than in term babies. For the vast majority of term infants who develop CP, birth asphyxia or obstetric complications cannot be ascribed as the cause.[4,5]

Correspondence and Reprint requests : Dr. Chitra Sankar, Department of Developmental Pediatrics, Bangalore Children's' Hospital, Chitrapur Bhavan, 15th Cross, 18th Main, Malleswaram, Bangalore – 560055.

Etiology and Risk Factors for CP

The etiology of CP is very diverse and multifactorial. The causes are congenital, genetic, inflammatory, infectious, anoxic, traumatic and metabolic. The injury to the developing brain may be prenatal, natal or postnatal. As much as 75% - 80% of the cases are due to prenatal injury with less than 10% being due to significant birth trauma or asphyxia.[5] The most important risk factor seems to be prematurity and low birth weight with risk of CP increasing with decreasing gestational age and birth weight. Cerebral palsy is seen in 10 – 18 % of babies in 500–999 grams birth weight.[6] CP occurs more commonly in children who are born very prematurely or at term. Although term infants are at relatively low absolute risk, term births constitute the large majority of all births, as well as approximately half of all births of children with cerebral palsy. Prenatal maternal chorioamnionitis is also a significant risk factor accounting for as much as 12% of cerebral palsy in term infants and 28% in premature infants.[7,8] Cystic periventricular leukomalacia (CPVL) is a risk factor with 60%-100% of patients with CPVL developing CP.[8]

Prenatal risk factors include intrauterine infections, teratogenic exposures, placental complications, multiple births, and maternal conditions such as mental retardation, seizures, or hyperthyroidism. The incidence of CP is higher among twins and triplets than singletons.

Perinatal risk factors are infections, intracranial

hemorrhage, seizures, hypoglycemia, hyperbilirubinemia, and significant birth asphyxia. Perinatal arterial ischemic stroke has been identified as another probable cause which leads to hemiplegic CP in many infants.

Postnatal causes include toxic, infectious meningitis, encephalitis, traumatic such as drowning. There is also a relation between coagulopathies causing cerebral infarction and particularly hemiplegic type of CP. Postnatal events account for 12% – 21% of CP. But in a large number of cases, the cause of CP remains unknown.

Associated Deficits are Present in a Large Majority of Cases – (75%)

Mental retardation (MR) is common in CP in up to 60% of the cases. Singhi *et al* in a study in India report MR in 72.5% of affected children. Children with spastic quadriplegia have greater degree of cognitive impairment than children with spastic hemiplegia.

Visual impairments and disorders of ocular motility are common (28%) in children with CP. There is an increased presence of strabismus, amblyopia, nystagmus, optic atrophy, and refractive errors. Children whose CP is due to periventricular leukomalacia are also more likely to have visual perceptual problems.

Hearing impairment occurs in approximately 12% of children with CP. This occurs more commonly if the etiology of CP is related to very low birth weight, kernicterus, neonatal meningitis, or severe hypoxic-ischemic insults.

Epilepsy is common in children with CP. And 35% to 62% of children develop epilepsy. Children with spastic quadriplegia (50% to 94%) or hemiplegia (30%) have a higher incidence of epilepsy than patients with diplegia or ataxic CP (16 to 27%). In an Indian study, it was found that 35% had epilepsy. 66% of children with spastic hemiplegia, 43% of spastic quadriplegia and 16% of children with spastic diplegia had seizures as an associated feature.[9]

Speech and Language Disorders

Speech is affected in CP due to bilateral corticobulbar and oromotor dysfunctions. Both receptive and expressive language deficits are common and go hand-in-hand with mental retardation. Articulation disorders and impaired speech are present in 38% children with CP.

Oromotor problems with feeding difficulties, swallowing dysfunction and drooling are also present.[10] This can result in nutritional problems affecting physical growth.[11,12] Behavioral problems are also well documented. Abnormalities of proprioception and tactile sensations are common in children with CP. Psychiatric disorders such as anxiety, depression, conduct disorders and hyperkinesis and inattention were seen in 61% of 6%-10 year-old-children with hemiplegic CP.[13] The associated deficits may be more disastrous for the CP child than the motor problem.

CLASSIFICATION OF CP

The topographic classification of CP is monoplegia, hemiplegia, diplegia and quadriplegia; monoplegia and triplegia are relatively uncommon. There is a substantial overlap of the affected areas. In most studies, diplegia is the commonest form (30% – 40%), hemiplegiae is 20% – 30%, and quadriplegia accounting for 10% – 15%. In an analysis of 1000 cases of CP from India, it was found that spastic quadriplegia constituted 61% of cases followed by diplegia 22%.[13]

Quadriplegic CP

This is the most severe form involving all four limbs, and the trunk upper limbs are more severely involved than the lower limbs, associated with acute hypoxic intrapartum asphyxia. However, this is not the only cause of spastic quadriplegia.[5] Neuroimaging reveals extensive cystic degeneration of the brain – polycystic encephalomalacia and polyporencephalon MRI and a variety of developmental abnormalities such as polymicrogyria and schizencephaly. Voluntary movements are few; vasomotor changes of the extremities are common. Most children have psuedobulbar signs with difficulties in swallowing and recurrent aspiration of food material. Half the patients have optic atrophy and seizures. Intellectual impairment is severe in all cases.[14]

Hemiplegic CP

Spastic hemiparesis is a unilateral paresis with upper limbs more severely affected than the lower limbs. It is seen in 56% of term infants and 17% of preterm infants. Pathogenesis is multifactorial. Voluntary movements are impaired with hand functions being most affected. Pincer grasp of the thumb, extension of the wrist and supination of the forearm are affected. In the lower limb, dorsiflexion and aversion of the foot are most impaired. There is increased flexor tone with hemiparetic posture, flexion at the elbow and wrist, knees and equines position of the foot. Palmer grasp may persist for many years. Sensory abnormalities in the affected limbs are common. Sterognosis impaired most frequently. 2 point discrimination and position sense is also defective. Seizures occur in more than 50%. Visual field defects, homonymous hemianopia, cranial nerve abnormalities most commonly facial nerve palsies are seen.[14]

Diplegic CP

Spastic diplegia is associated with prematurity and low birth weight. Nearly all preterm infants with spastic diplegia exhibit cystic periventricular leukomalacia on neuroimaging. Periventricular leukomalacia (PVL) is the most common ischemic brain injury in premature infants. The ischemia occurs in the border zone at the end of arterial vascular distributions. The ischemia of PVL occurs in the white matter adjacent to the lateral ventricles. The diagnostic hallmarks of PVL are periventricular echo

densities or cysts detected by cranial ultrasonography. Diagnosing PVL is important because a significant percentage of surviving premature infants with PVL develop cerebral palsy, intellectual impairment, or visual disturbances due to site of injury affecting the descending corticospinal tracts and visual radiations. Premature infants have impaired cerebrovascular auto regulation and are susceptible to intracranial hemorrhage (ICH) as well as PVL. Many premature infants have both PVL and ICH detected on ultrasonography. Maternal chorioamnionitis or vasculitis, with the production of cytokines, leading to inflammatory damage to the periventricular area in the developing brain is another factor in the pathophysiology of PVL. An estimated 60% 100% of patients with cystic periventricular leukomalacia go on to develop CP. [7, 14]

In this condition, lower limbs are more severely affected then the upper limbs. Mild cases may present with toe walking due to impaired dorsiflexion of the feet with increased tone of the ankles. In severe cases, there is flexion of the hips, knees and to a lesser extent elbows. When the child is held vertically, rigidity of lower limbs is most evident and adductor spasm of the lower extremities causes scissoring of the legs. Seizures are common. Fixation difficulties, nystagmus, strabismus, and blindness have been associated with PVL.

CP is classified based on the type of neuromuscular deficit into (i) spastic (ii) dyskinetic (inclusive of choreo-athetoid and dystonic) (iii) ataxic (iv) hypotonic and (v) mixed. Spastic CP is the commonest and accounts for 70%-75% of all cases, dyskinetic – 10% to 15% and ataxic is less than 5% of cases.

Spastic types exhibit pyramidal involvement with upper motor neuron signs, weakness, hypertonia, hyperreflexia, clonus and positive Babinski. Dyskinesia is characterized by extra pyramidal involvement in which rigidity, chorea, choreoathetosis, athetoid and dystonic movements are seen. This type of CP is also associated with birth asphyxia.[5] The severity of dystonic postures may vary with body position, emotional state and sleep. Clonus and Babinski are absent. Primitive reflexes are more prominent and persist for a longer time in dyskinetic CP. These movement patterns are eliminated in sleep, with a decrease in tone of the affected limbs. There are also abnormalities of posture control and coordination. Those children who are hypotonic to start with may develop into this type by 1 to 3 yrs of age. In majority of this group, there is no cognitive impairment. Dysarthria, oromotor problems with drooling and swallowing difficulties are seen. 30% of children with CP have a mixed pattern of involvement. While contractures are common in spastic group, they are uncommon in the extra pyramidal group.

Hypotonic CP is characterized by generalized muscular hypotonia that persists beyond 2 to 3 yrs of age that does not result from a primary disorder of muscle or peripheral nerve. The deep tendon reflexes are normal or hyperactive, and the electrical reactions of muscle and nerve are normal. More than half the children develop frank cerebellar deficits with incoordination, ataxia and impaired rapid succession movements.[14]

The Gross Motor Function Classification System (GMFCS)

This is a recently developed system which has been found to be a reliable and valid system that classifies children with cerebral palsy by their age-specific gross motor activity. The GMFCS describes the functional characteristics in five levels, from I to V, level I being the mildest in the following age groups: up to 2 yrs, 2 – 4 yrs, 4 – 6 years and between 6 to 12 years. For each level, separate descriptions are provided. Children in level III usually require orthoses and assisting mobility devices, while children in level II do not require assisting mobility devices after age 4. Children in level III sit independently, have independent floor mobility, and walk with assisting mobility devices. In level IV, affected children function in supported sitting but independent mobility is very limited. Children in level V lack independence even in basic antigravity postural control and need power mobility.[15]

EARLY DIAGNOSIS

Cerebral palsy is a clinical diagnosis made by an awareness of risk factors, regular developmental screening of all high risk babies and neurological examination. As in all medical conditions, a systematic approach focusing on maternal, obstetric and perinatal histories, review of developmental milestones, and a thorough neurological examination and observation of the child in various positions such as supine, prone, sitting, standing, walking and running is mandatory.[16]

It is not possible to diagnose CP in infants less than 6 months except in very severe cases. The patterns of various forms of CP emerge gradually with the earliest clues being a delay in developmental milestones and abnormal muscle tone. In CP, the history is nonprogressive. Milestones once acquired do not show regression in CP. Tone may be hypertonic or hypotonia. Many of the early hypotonia change to spasticity or dystonia by 2 – 3 yrs of age. Early signs include presence of hand preference in the first year, prominent fisting, abnormalities of tone–either spasticity or hypotonia of various distribution, persistence of abnormal neonatal reflexes, delay in the emergence of protective and postural reflexes, asymmetrical movements like asymmetrical crawl and hyperreflexia. Primitive reflexes should gradually extinguish by 6 months of age. Among the most clinically useful primitive reflexes are Moro, Tonic labyrinthine and Asymmetric Tonic Neck Reflex (ATNR). In many cases a diagnosis of CP may not be possible till 12 months. Repeated examinations and observation over a period of time may be required in mild cases before a firm diagnosis can be made.[17, 18]

In the further evaluation of a child with CP, an EEG is obtained if there is history of epilepsy. Neuroimaging studies are carried out if they have not been done in the neonatal period that provided the etiology of CP. MRI studies is preferred to CT scans. Genetic and metabolic tests are carried out if there is evidence of deterioration or metabolic compensation, family history of childhood neurological disorder associated with CP. Tests to rule out coagulopathy in children with stroke is necessary.[19]

Complete evaluation of a child with CP should include an assessment of associated deficits like vision, speech and hearing, sensory profile, oromotor evaluation, epilepsy and cognitive functioning. Orthopedic evaluation is a must as muscle imbalance and spasticity cause subluxation / dislocation of the hips, equinus deformities, contractures, and scoliosis.

CONCLUSION

CP is a chronic condition with considerable impact on affected individuals. Overall prevention of CP has not been successful. Early diagnosis and a comprehensive management with a multidisciplinary approach involving developmental pediatrician or neurologist, orthopedic surgeon, speech and language therapist, physio and occupational therapist are required for the further satisfactory management of a child with CP.

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