



Mini Review

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Cerebral Palsy: A Brief Review



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Abstract

Cerebral palsy (CP) is a group of permanent movement disorders that appear in early childhood. Cerebral palsy is caused by abnormal development or damage to the parts of the brain that control movement, balance and posture. Risk factors include preterm birth, being a twin, certain infections during pregnancy such as toxoplasmosis or rubella, exposure to methylmercury during pregnancy, a difficult delivery and head trauma during the first few years of life, among others. About 2% of cases are believed to be due to an inherited genetic cause. A number of sub-types are classified based on the specific problems present. The treatment must be goal oriented, such as to assist with mobility, reduce or prevent contractures, improve positioning and hygiene, and provide comfort. Each member of the child's multidisciplinary team, including the child and both parents, should participate in the serial evaluations and treatment planning.

Definition

Cerebral palsy is defined as chronic disability of the CNS involving posture and tone, occurring early in life not the result of progressive neurological disease associated with visual, hearing, dental, behavioural with or without seizures. Cerebral palsy is the most common motor disability in childhood.

Incidence of Cerebral palsy in multiple births is 7.5/1000 live births and in Singletons it is 2.1/1000 live births and, more in 1500grams or less is 80/1000. 10% of the global population has some form of disability due to different causes, In India, it is 3.8% of the population. Nearly 15-20% of the total physically handicapped children suffer from Cerebral palsy. In India, the estimated incidence is around 3/1000 live births. In an analysis of 1000 cases of CP it was found that spastic quadriplegia constituted 61% of cases followed by diplegia 22% [1]. The prevalence of CP has increased somewhat as a result of the enhanced survival of very premature infants weighing less than 1000 grams who go on to develop CP at a rate of 15 per 100. Risk factors include preterm birth, being a twin, certain infections during pregnancy such as toxoplasmosis or rubella, exposure to methylmercury during pregnancy, a difficult delivery, and head trauma during the first few years of life, among others. About 2% of cases are believed to be due to an inherited genetic cause.

Classification

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%), hemiplegia is 20%-30%, and quadriplegia accounting for 10%-15%.

CP can also be classified based on the type of neuromuscular deficit into

- a. Spastic.
- b. Dyskinetic (inclusive of choreoathetoid and dystonic).
- c. Ataxic.
- d. Hypotonic.
- e. 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. 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 [2]. Dysequilibrium syndrome may be seen presenting as difficulty in maintaining an upright position and in experiencing the position of the body in space.

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 [3].

Early Diagnosis

Early signs of cerebral palsy include 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. It is not possible to diagnose CP in infants less than 6 months except in very severe cases. The Vojta technique can detect cerebral palsy earlier. Early indicators of Cerebral Palsy may include birth history of prematurity, seizures, low Apgars, intracranial haemorrhage, periventricular leukomalacia. History of delayed milestones especially noticed during the period where child sits up. Signs of abnormal motor performance early handedness in the first year of age, reptilian crawl, toe walking along with altered tone detected by Bailey scoring. Another early feature is persistence of primitive reflexes and abnormal posturing. Also present is 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). 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 hypotonic change to spasticity or dystonia by 2-3 years of age. 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 [4,5].

The further evaluation of a child with CP, is done with help of developmental Paediatrician, Neurologist, Physician, ENT surgeon, Ophthalmologist, Physiotherapist, occupational therapist. 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. 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 [6]. Common management Problems in Cerebral Palsy are feeding problems failure to suck, tongue trusting, gagging and choking, vomiting and regurgitation, dribbling, constipation, crying, screaming and sleep disturbances, chilblains and cold injury and growth.

Treatment

Treatment of Cerebral Palsy is done by a team of Physicians including neuro developmental paediatrician, paediatric neurologist, physical medicine and rehabilitation specialists as well as occupational and physical therapist speech pathologist, social workers, educators and developmental psychologist, physiotherapist and occupational therapist, is important to reduce abnormalities of movement and tone and to optimise normal psychomotor development. Parents should be taught how to

work with their child in daily activities such as feeding, carrying, dressing, bathing and playing in ways that limit the effects of abnormal muscle tones. Physiotherapy by Bobath method, Peto, Doman-Delacato methods. Surgeries for e.g. neurosurgery for Spasticity, movement disorder (dystonia) etc. Orthopedic surgery for contracture and dislocation. The Neurosurgical procedures SPR (Selective Posterior Rhizotomy), SMF (Selective Motor Fasciculotomy), Orthopedic procedures, STR (Soft Tissue Release) surgeries and Tendon transfer are to be considered. Management of Spasticity in Cerebral Palsy is by oral medications Baclofen, Diazepam, Tizanidine, Dantrolene, Intrathecal Baclofen by insertion of a drug delivery system for continuous infusion and use of Botulinum Toxin. Small doses of levodopa and Artane (trihexiphenidyl) is sometimes useful for treating dystonia. Reserpine or tetrabenazine can be useful for hyperkinetic movement disorders. Treatment of seizures according to the type is essential, Brain tonics for few months to few years following insult to brain are used. Spinal cord stimulation is another option of treatment. In hemidystonic cases and in a few diffuse cases stereo tactic thalamic-basal ganglionic stimulatory or ablative surgery can be considered. The nano technology in future is likely to be quite promising in this group of patients. Both physical and mental exercises (Physiotherapy, occupational therapy, cognitive therapy, special education) are essential during most of the period. Neurostimulation method of the therapy, indeed, stimulates brain and improves neuronal functions of the existing brain cells. Brain has tremendous potential power which can be brought to light by stimulating therapies. Cooling term infants with hypoxic ischemic encephalopathy to 33.3 °C for three days, starting with in six hours of birth, in perinatal asphyxia affected new borns reduces the risk of dyskinetic or spastic quadriplegia. Following asphyxia starting very early treatment will give the best results is to be

stressed. Assistive technology by equipment or devices to improve independence e.g. walking frames, wheelchairs, adapted computer access is being used. Communication skills may be enhanced by the use of Bliss symbols, talking typewriters, electronic speech generating devices. Prevention of CP is by efficient antenatal neonatal care, early detection and early maternal advice, Immunization and screening, genetic counselling and health education [7]. Ultimate goal of the management of cerebral palsy patients is to provide them as well as their families a comfortable life and to make them as much productive as possible.

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