Cerebral Palsy: An overview

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Introduction

The developmental disabilities are a group of disorders differentiated by the pattern of delay among developmental streams. The four streams of development include language, problem solving, motor and social. Cerebral palsy (CP) is a disorder of development in which motor function abnormalities are a key feature. It is a most common developmental disorder of children first described by William little in the 1861. Severity varies from mild with minimal disability in which affected person leads almost normal life to severe, associated with several comorbid conditions.

Definition

Cerebral palsy is a static encephalopathy. It is a disorder of posture and or movement. 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]. Although the brain lesion is a nonprogressive, the neurologic features change over a period of time and the clinical presentation may change accordingly due to growth and maturation of CNS.

Epidemiology

The incidence of cerebral palsy in developed world is 2-2.5/1000 live births [2]. However, with the increasing survival of VLBW premature infants, the prevalence appears to increase slightly. Although the risk of cerebral palsy is increased 30 fold in prematures compared to term infants, the vast majority of children who are VLBW do not have cerebral palsy. Life expectancy of patients with cerebral palsy is related to the type of involvement and pattern of motor disability. Severe quadriplegia and profound intellectual retardation have been associated with limitation of life expectancy.

It is increasingly obvious that with reasonable care the majority of affected persons will survive into adult life [3]. Studies in developed countries have indicated that the prevalence of cerebral palsy has not declined in any birth weight group. With the increasing survival of LBW infants who are at high risk of cerebral palsy the cerebral palsy rate of such population is yet to be determined [4].

Etiology and Risk Factors

Etiology of cerebral palsy is diverse. According to collaborative perinatal project, most of children with cerebral palsy are born at term with uncomplicated labor and deliveries. In 80% of cases features were identified pointing to antenatal factors causing abnormal brain development. Fewer than 10% had evidence of intrapartum asphyxia [5]. Various congenital anomalies, metabolic disorders, genetic defects, infections, birth trauma can produce cerebral palsy.

Prematurity and LBW are significant risk factors for cerebral palsy. Preterm infants who are small for gestational age are even at greater risk than preterm appropriate for gestational age babies, risk of cerebral palsy increases with
decreasing birth weight and gestational age. In one study 10-18% with birth weight between 500-999 grams developed cerebral palsy. Cerebral palsy in preterm population is primarily as a result of periventricular leukomalacia [6].

Chorioamnionites is a risk factor for cerebral palsy for all birth weights as many as 80% of preterm births have been associated with placental infection [7]. Intrauterine and neonatal infections such as Rubella, cytomegalovirus, toxoplasmosis and other infectious agents can infect the fetus or neonate and can produce severe encephalitides with motor sequelae [8].

Maternal conditions like unusual menstrual periods, thyroid disorders and estrogen administration have also been associated with cerebral palsy [9]. Term infants who experience an immediate postpartum course that is comprised of a 5-minute Apgar score of 5 or less, continuing neurological abnormalities and seizures in the first day of life constitute a group at high risk for chronic motor disability (55%) and for death and disability (70%) [10].

Besides above mentioned risk factors there are other risk factors acting in prenatal, perinatal, postnatal periods that increase chances of cerebral palsy like multiple births, maternal conditions like mental retardation, seizures, hyperthyroidism etc. intracranial hemorrhage, hyperbilirubinemia, perinatal arterial ischemic to hemiplegic cerebral palsy in many infants. Postnatal causes include – meningitis, encephalitis, trauma, coagulation disorders.

Classification

The modified Swedish classification is the wisely used classification which is based on tone, number and distribution of affected limbs (1). The changing nature of symptoms and signs make the clinical classification difficult in the first years of life:

a) Clinical classification


In a study of Franco and Andrews [ ], spastic Diplegia occurred in 53% of the cerebral palsy cases and was most common clinical type regardless of birth weight and gestational age spastic quadriplegia was present in 28% of the patients and 10% with spastic Hemiplegia other types comprised only 9%.

In an analysis of 1000 cases of cerebral palsy from north India it was found that spastic quadriplegia constituted 61% of cases followed by Diplegia 22% [11].

b) Functional Classification

Classification based on an estimate of degree of severity and patient’s ability to perform normal activity has been proposed

Class I-Practical limitation of activity

Class II -Slight to moderate limitation of activity

Class III -Moderate to great limitation of activity.

Class IV-Inability to carryout any useful physical activity.

This type of classification of cerebral palsy has obvious advantage of describing the patients’ functional abilities without regard to specific pathologic conditions or symptomatology. It is useful over long periods to gauge the progression or regression of symptoms or the effect of drug, physical and environmental therapy.

Associated deficits
Cerebral palsy is commonly associated with a spectrum of developmental disabilities including mental retardation, epilepsy, visual, speech, hearing, cognitive, and behavioral abnormalities. The motor handicap may be the least of the child’s problems. Mental retardation is the commonest associated problem in children with cerebral palsy [12].

Ocular abnormalities are also common associated problem. In a study of 117 children with cerebral palsy only 20% had normal eyes. Refractive errors were present in 50% of the patients. Concomitant squint in 37% and paralytic squint in 14%. In general patients with spastic diplegia tend to have the most ocular abnormalities followed in order by spastic quadriparesis and spastic hemiparesis. The high percentage of ocular abnormalities clearly requires that virtually all patients with cerebral palsy have an ophthalmoscopic examination (12).

Hearing problems occurs approximately in 12% of children with cerebral palsy. Children with hearing impairment, associated congenital heart disease and microcephaly should be looked for other stigmata of TORCH infection. In cases with dyskinetic cerebral palsy presence of hearing impairment may point to kernicterus as a cause of cerebral palsy. Sensorineural hearing loss is a prominent feature of cerebral palsy due to iodine deficiency in endemic areas. Hearing problems may also be associated with cerebral palsy due to perinatal asphyxia and meningitis.

Epilepsy is common in children with cerebral palsy. It occurs in almost 50% of children with spastic quadriplegia [13], almost 1/3 of children with spastic diplegia. Mental retardation is highly correlated with epilepsy in hemiplegic cerebral palsy [14]. Epilepsy is present in 30% of patients with spastic hemiplegia.

Speech and Language - the production of speech language and gesture for communication is often affected by cerebral palsy [15]. It may be due to bilateral corticobulbar, oromotor, hearing impairment, cognitive deficits. These children need to be assessed for speech and language by a speech therapist.

Oromotor problems and feeding difficulties – 30-80% of disabled individuals feed with difficulty. In a study by Gangil et al [16] oromotor dysfunction was observed in all cases.

**Clinical features**

1. Spastic quadriplegia – is characterized by generalized increase in muscle tone. The legs are involved more than the arms and there is paucity of limb movements. The in coordination of oropharyngeal muscles may predispose the patient to recurrent pneumonias during the first years of life. It is usually associated with acute hypoxic intrapartum asphyxia. However, this is not the only cause of spastic quadriplegia. Half the patients have optic atrophy and seizures. Intellectual impairment is severe in all case [17].

The most common lesion seen on pathological examination or on MRI are severe periventricular leukomalacia and multi cystic cortical encephalomalacia and a variety of developmental abnormalities like polymicrogyria and schizencephaly.

2. Spastic hemiplegia – Although affected child may manifest obvious hemiplegia in the 2nd year of life their difficulties may not be observed during the first 3-5 months of life. Arm is usually affected more than the leg and hypotonia may be the most prominent feature. Parents may observe that the child has developed prematurely right or left handedness during the first 2 years of life. This type is seen in 50% of term and 17% preterm population. Palmer grasp which is usually absent after 6 months may be obligate. Weakness of wrist and forearm is often associated with limitation of range of motion of supination, range of elbow extension may be restricted. These children have circumductive gait that is present in varying degrees. Most commonly the child walks on the toes and swings the affected leg over a nearly semicircular arc during the course of each step.

Seizures occurs approximately in 30% of patients, visual field defects, homonymous hemianopia, cranial nerve abnormalities most commonly facial nerve palsy are seen.

3. Diplegia - Spastic diplegia is characterized by bilateral leg involvement and commonly some degree upper extremity impairment. Preterm infants are particularly prone to spastic diplegia. Approximately 80% of preterm infants who manifest motor abnormalities have spastic diplegia [18]. In recent years the survival of very small preterm infants has led to larger group of more severely neurologically impaired survivors.
4. Chorioathetoid or extra pyramidal cerebral palsy is less common than spastic cerebral palsy. Affected infants are characteristically hypotonic with poor head control and marked head lag and develop increased variable tone with rigidity and dystonia over several years. Feeding may be difficult and tongue thrust and drooling may be prominent. Speech is typically affected because oropharyngeal muscles are involved. Generally upper motor neuron signs are not present. Seizures are uncommon and intellect is preserved in many cases. This type is most commonly associated with perinatal asphyxia. Extra pyramidal cerebral palsy secondary to acute intrapartum near total asphyxia is associated with bilaterally symmetric lesions in the posterior putamen and ventrolateral thalamus. These lesions appear to be the correlate of the neuropathologic lesion called "status marmoratus."

**Diagnosis**

Diagnosis of cerebral palsy requires a complete history, physical examination and ancillary investigations. The history should include a detailed account of gestation, and perinatal events and documentation of attainment of developmental milestones. In addition to a general physical inspection, the examination should assess station (Pelvic and leg alignment during stance), spinal alignment, gait, active and passive range of motion of joints, sensibility, motor power, muscle tone type, extent of movement disorders and presence of limb deformity.

Early detection of cerebral palsy is important to improve functional abilities of affected child. Development of human brain is far from finished at birth and is greatly influenced by extensive environmental stimuli as well as "programmed" genetic factors. Postnatal brain growth continues in adulthood but it is more rapid and pliable in the early years of life. Increasing evidence suggests that the children are never too young to learn. Furthermore, the younger brain is better able to compensate for static injury than is the older or adult brain. So by early detection of cerebral palsy we may be able to improve child’s chances by modifying his environment.

It is often difficult to diagnose cerebral palsy in infants less than 6 months except in very severe cases. The pattern of various forms of cerebral palsy emerges gradually with the earliest signs being delay in developmental milestones and abnormal muscle tone. In cerebral palsy history is non progressive, no regression of milestones. Tone may be hypertonic or hypotonic. A cluster of certain behavioral symptoms and deviation from normal motor patterns are suggestive of cerebral palsy.

Early signs of cerebral palsy are

- Presence of hand preference in the first year.
- Prominent fisting of hands beyond 2 months
- Tone abnormalities
- Persistence of abnormal neonatal reflexes.
- Abnormal asymmetric tonic neck reflex.
- Delay in emergence of protective and postural reflexes.
- Feeding Problems
- Paucity of movement or excessive or disorganize movement.

In many cases diagnosis of cerebral palsy may not be possible till first year. Repeated examination and observation over a period of time may be required in mild cases before a firm diagnosis can be made [19]. Cranial ultrasonography, MRI, CT scan head and other specialized tests are used to assess the extent of CNS insult, currently functional MRI and trans cranial magnetic stimulation are used only for research purposes in the children with cerebral palsy.

**Treatment**

No single brain abnormality is responsible for cerebral palsy and no two patients are identically impaired. Children also differ in their genetic make up, intellect, personality and family environment; consequently each child’s treatment must be individualized to meet his or her needs. The pediatrician plays a key role in organizing the team of professionals necessary to care for children with cerebral palsy; this includes the pediatrician, physical, occupational, speech therapist, neurologist, neurosurgeon, orthopedician, psychiatrist, educational expert, councilors and others.
All children with cerebral palsy must have full evaluation of their motor, sensory, cognitive functions, hearing, vision, speech, general health, feeding and nutrition, behavioral problems and should be treated appropriately.

I. Vision and hearing

Overall 11% of patients with cerebral palsy have severe visual impairment (20) and 12% have hearing problem. Routine assessment of vision and hearing is therefore required in all patients with cerebral palsy. Early diagnosis of strabismus and amblyopia is necessary to get optimal visual potential. Furthermore assessment of vision and hearing is important to determine rehabilitative programme.

II. Speech and language

The production of speech, language and gesture for communication is often affected by cerebral palsy. These may be due to oromotor dysfunction, hearing impairment. So in a comprehensive management programme of cerebral palsy speech therapy has a pivotal role, all such patients should be assessed by speech therapist.

III. Feeding and nutrition

Feeding problems are quite common in children with cerebral palsy. 30-80% of disabled individuals feed with difficulty they are especially at risk because of oral, pharyngeal or esophageal dysphasia and due to oromotor dysfunction. Furthermore because of communication difficulties many of them are unable to request for food and drink. Pressure of seizures worsens the intake as a result they do not receive adequate nutrition resulting in growth retardation, reported in as many as 48% of children with neurodevelopment handicaps [22]. So a nutritional rehabilitation can be rewarding in these children.

The important cause of failure to thrive in these patients in addition to above which is often neglected is gastroesophageal reflux disease (GERD). In one study 72% of patients had symptoms suggestive of GERD. Failure to thrive with feeding difficulties was the commonest presenting symptom [23].

While treating such patients due care to be given regarding feeding and nutrition. Regular assessment of growth of patient, vitamin or micronutrient deficiency should be looked for; complete dietary advice is essential component of rehabilitation programme.

IV. Behavior and emotional problems:

Often overlooked problem these patients are prone to have anxiety, depression, Conduct disorders, hyperkinesis inattention and poor self esteem. Assessment of behavior and emotional problems and their treatment by a child psychiatrist must be included in the comprehensive programme in the management of patient with cerebral palsy.

V. Orthopedic problems

Patients of cerebral palsy are prone to develop subluxation of hip, equines deformity, contractures of hamstring muscles, reduced bone density and increased tendency to fractures [24] and scoliosis. A thorough musculoskeletal system examination needs to be performed in children with cerebral palsy. Observation of gait provides useful information about coordination and musculoskeletal abnormalities if a child is ambulatory. Long established orthopedic surgical procedures are designed to lengthen contracted myotendinous units, balance joint forces, transfer motor power, biomechanical alignment, reduce joint subluxation and dislocation to improve joint congruency, diminish painful spasticity and maintain, restore or stabilize spinal deformity. The surgical procedures should be individually adjusted according to the patient’s age, disease severity, underlying pathology, co morbidities, and overall wellbeing. The natural course of a specific musculoskeletal deformity should be factored into the surgical decision making process.

VI. Epilepsy

As already mentioned epilepsy is commonly associated with cerebral palsy. Furthermore epilepsy in such patients is often refractory to treatment. Any type of epilepsy can be associated with cerebral palsy. Most children with cerebral
Palsy have onset of seizures in 1st or 2nd year of life. These patients often require polytherapy to control their seizures. Management of severe epilepsy is best done in consultation with pediatric neurologist.

**VII. Spasticity**

Spasticity refers to constellation of clinical findings, including increased tone, weakness, hyperactive reflexes and poor coordination. In the pediatric population, spasticity is most commonly encountered in the setting of cerebral palsy. Objective methods to measure spasticity can be used like modified Ashworth scale but the reliability of these scales are not up to the mark:

**Modified Ashworth scale of spasticity:**

<table>
<thead>
<tr>
<th>Score/Grade</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>1.</td>
<td>Hypotonia Normal, Mild, slight increase in tone, minimal</td>
</tr>
<tr>
<td>2.</td>
<td>Resistance to movement through less than One half of range of movement (ROM)</td>
</tr>
<tr>
<td>3.</td>
<td>Moderate, more increase in tone through most of ROM but affected parts easily moved.</td>
</tr>
<tr>
<td>4.</td>
<td>Severe, considerable increase in muscle tone; passive movement difficult.</td>
</tr>
<tr>
<td>5.</td>
<td>Extreme, affected parts rigid in flexion or extension</td>
</tr>
</tbody>
</table>

Various non pharmacologic, pharmacologic, surgical methods can be used to reduce spasticity.

**A. Physiotherapy and occupational therapy**

Early and aggressive physiotherapy and occupational therapy have been shown to be very effective and may obviate the later need for other therapeutic interventions to reduce spasticity [25]. In addition to their importance in maximizing patient function, the success of other medical and surgical interventions for the treatment of spasticity is highly dependent on rehabilitative exercises to optimize results. Therapists provide range of motion exercises, strengthening regimens and facilitatory inhibitory exercise programs. The patients daily function may also be enhanced by the provision of adaptive devices such as seating systems and transfer benches.

**B. Pharmacotherapy**

a) Oral:

When a generalized reduction of body tone is desired, the use of orally administered medications for the treatment of spasticity can be very effective in selected patients. Despite the usefulness of these medications many patients will sustain excessive sedation prior to achieving appropriate modulation of their spasticity. These medications frequently exert their effect by inhibiting excitatory neurotransmitters at the level of spinal cord. In general the use of oral medications is limited to children with generalized spasticity who may benefit from mild reduction of spasticity. Higher doses are associated with systemic side effects like sedation, weakness, behavior changes and other central side effects limiting their usefulness:

b) Local drug injections: Botulinum toxin type A, alcohol, phenol. Local injections of these drugs cause interruption of impulse transmission. Botulinum toxin acts by neuromuscular blockade [26]. Alcohol and phenol exert their effect by causing chemodenervation.

i) Botulinum toxin type A:

Botulinum toxin A can be injected intramuscularly to produce selective and reversible chemodenervation at the neuromuscular junction. Compared with phenol injections it is associated with fewer complications, greater reversibility of effects, and ease of drug administration [27]. Botulinum toxin A has been used clinically in the management of spasticity associated with cerebral palsy since 1988 [28]. Botulinum toxin is the toxin produced by the bacterium Clostridium Botulinum, which when consumed produces the general paralysis associated with botulism. It acts by blocking release of acetylcholine presynaptically, thereby weakening the force of contraction. In clinical trials Botulinum toxin has been effective in reducing limb tone, reducing painful muscle spasm and in improving limb function. Contraindications in the use of botulinum toxin are few, the most important being the presence of fixed contractures.
Injection protocols for botulinum toxin should begin with obtaining informed consent, including discussion of potential side effects. Side effects are rare but may include pain during injection, infection, bleeding, a cool feeling in injected limbs, rash allergic reactions, flu-like symptoms, excessive weakness and fatigue.

The doses are individualized for each patient based on their functional limitations. Dose also depends on type of commercial preparation used. Injection of botulinum toxin needs to be repeated every 3-6 months to maintain optimal therapeutic effectiveness. In selected patients botulinum injections may have to be repeated. Failure to respond to these injections may indicate the presence of antibodies against botulinum toxin which develops in some 5% of patients.

ii) Alcohol and Phenol:

Alcohol and phenol are non selective proteolytic agents and produce selective denervation when injected into motor nerves or muscles. Because diffusion of both is limited, the area of effective denervation extends a few millimeters from the injection site. The duration of denervation associated with alcohol injection is 3-6 months, whereas phenol denervation lasts 4-8 months. Side effects of alcohol and phenol injection include pain on injection, non selective protein denaturation, possible permanent muscle fibrosis, and dysaesthesias lasting for several weeks [29].

C). Intrathecal Baclofen:

Intrathecal Baclofen was initially used in 1980 as a way of controlling the severe muscle spasms in patients with spinal cord injury. Since the early 1990s its use in the “correct” cerebral palsy patients had been demonstrated to be efficacious. Intrathecal Baclofen allows higher concentration into the CSF while reducing systemic side effects associated with oral administration [30]. Drug is delivered with the help of pump implanted subcutaneously in the abdomen. Prior to the pump installation a trial dose of 50-100 micrograms of Baclofen is administered by lumbar puncture. If this produces improved tone in the absence of significant side effects, the patient is deemed a candidate for pump implantation. Complications associated with pump include CSF leakage, catheter infections and meningitis.

Table 2: Various drugs in this category are:

<table>
<thead>
<tr>
<th>Drug</th>
<th>Dose</th>
<th>Mechanism of action</th>
<th>Side effect</th>
</tr>
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<tbody>
<tr>
<td>Diazepam</td>
<td>1-10 mg/dose QID</td>
<td>Potentates effect of GABA in the spinal cord</td>
<td>Sedation, drooling, ataxia behavioral disturbances</td>
</tr>
<tr>
<td>Baclofen</td>
<td>2.5-25 mg/dose TID. Max. dose 80-120 mg/day</td>
<td>Decreases release of excitatory Neurotransmitters</td>
<td>Headache, dizziness, weakness, insomnia, hypotension, nausea, constipation</td>
</tr>
<tr>
<td>Dantrolene</td>
<td>1-3 mg/kg/dose QID</td>
<td>Decreases release of calcium from sarcoplasmic reticulum in muscle cells</td>
<td>weakness, hepatitis, diarrhea, drowsiness, dizziness</td>
</tr>
<tr>
<td>Tizandine &amp; Clonidine</td>
<td>Tizandine 0.3-0.5 mg/kg/day in 4 divided doses Clonidine – starting</td>
<td>α2 adrenergic agonist inhibits release of excitatory neurotransmitters</td>
<td>Sedation, hypotension, especially with Clonidine, depression, drymouth, dizziness</td>
</tr>
</tbody>
</table>
C. Selective Dorsal Rhizotomy:

The use of selective dorsal rhizotomy for spasticity management was first reported in cerebral palsy in 1981 [31]. It decreases spasticity by balancing spinal cord mediated inhibitory control. Because transient muscle weakness is observed after the procedure, many patients require orthotics and long term physiotherapy to achieve the best outcome. Patients with normal intelligence, pure spasticity, diplegia, no fixed contractures, good strength, postural stability have been identified as the best candidates for selective dorsal rhizotomy, although the exact indications are difficult to define from the existing evidence [32].

D. Hyperbaric oxygen

Hyperbaric oxygen therapy is used in some patients despite the lack of scientific validation of its efficacy. The largest randomized, placebo controlled trial of this therapy found that hyperbaric oxygen did not improve the condition of children with cerebral palsy compared with slightly pressurized air [33]. In another double blind, placebo controlled trial in 75 children, 100% oxygen at 1.75 atmospheres absolute for 40 sessions was compared with air at 1.30 atmospheres absolute for 40 sessions [34]. No significant difference was found between the groups in the results of neuropsychological testing. Hyperbaric oxygen therapy is associated with various risks including, ear pain, damage to the ear, pneumothorax, myopia, convulsions, and fire and explosions associated with the equipment. On the basis of current peerreviewed scientific data, hyperbaric oxygen therapy should be considered an experimental intervention until further randomized, placebo controlled; doubleblind trials are completed [35].

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