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Reference Review And Discussion On Prostheses & Cerebral Palsy: Dyskinetic

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Reference Review And Discussion On Prostheses

Cerebral Palsy: Dyskinetic

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Description

Dyskinetic cerebral palsy (CP) is one of the most disabling forms of CP and is characterized by a predominance of stereotyped, involuntary movements that are accentuated with effort.

Etiology/Types

Seen with injury to the extrapyramidal system (mainly the basal ganglia and thalamus) while the more common spastic form is associated with pyramidal tract involvement. Classified as given below:

■ _Dystonic
■ _Hyperkinetic
• Slow movements: Athetosis
• Fast movements: Chorea, Ballismus, Tremors

Five percent of children with CP may have ataxic type of CP. This is seen with injury to the cerebellum or cerebellar pathways and children are often hypotonic. This chapter will focus primarily on the dyskinetic form and the spastic and ataxic forms will be covered elsewhere

Epidemiology

- _Lack of a standard classification system makes it difficult to determine the exact worldwide prevalence
- _The prevalence of dyskinetic CP per 1000 live births increased from 0.08 in the 1970s to 0.14 in the 1990s
- _3% to 15% of children with CP have dyskinetic type
- _Dyskinetic CP is more commonly seen in term infants with only a third occurring in preterm infants

Pathogenesis

- _Outstanding neuropathological feature of dystonia is bilateral sclerosis of the globus pallidus
- _Dystonia is seen with lesions involving the thalamus and basal ganglia, particularly the striatopallidal tracts
- _Athetosis due to asphyxia is seen with lesions of the caudate nucleus and putamen
- _Athetosis from kernicterus is seen in lesions of the globus pallidus and subthalamic nuclei as well as cranial nerve nuclei in the floor of the fourth ventricle

Risk Factors

- _Perinatal adverse events account for more than two-thirds of those with dyskinetic CP; prenatal events in 20%
- _Term infants and those weighing >2500 g at birth
- _Low Apgar scores (0–3) at 1 and 5 min. The lower the score, the more severe the functional impairments noted
- _Neonatal jaundice and kernicterus are risk factors for dyskinetic CP, particularly athetosis, but are not as common now as in the days of Rh incompatibility

Clinical Features

■ _Dystonia: abnormal postures from sustained muscle contractions; usually combined with some spasticity
■ _Athetosis: slow writhing movements
■ _Chorea: rapid, jerky, and dancing movements
■ _Athetosis and chorea are usually seen together as choreo-athetoid CP
■ _While visual impairments are common findings in spastic CP, hearing impairments may be seen more frequently with dyskinetic CP
■ _Motor control, communication, and learning may be affected
■ _The motor limitations and dysarthria from dyskinetic CP may cause individuals to appear as if they are cognitively impaired even when in reality they may be of higher than normal intelligence
■ _Movements may be accentuated by effort and excitement, and are frequently abolished by sleep
■ _Hands are well developed and appear relatively large as compared to children with predominantly spastic forms of CP

Natural History

■ _Infants at risk for the development of dyskinetic CP show fewer spontaneous “fidgety” movements than normal in the first few months. They show arm movement patterns that differentiate them from those at risk for spastic CP
■ _Postural impairments of head and trunk control are the earliest signs; dyskinesias develop by the end of the first year or later 8
■ _Dyskinesias may worsen over time

Diagnosis

Differential diagnosis

- _Glutaric aciduria type I (a condition where the infant is usually normal at birth, may have sudden onset of vomiting, hypotonia, and neurological problems after a period of normal development, may have intracranial bleeds and is often mistaken for child abuse) and other amino acid disorders
- _Primary and dopa responsive dystonia, which shows a diurnal variation in gait disturbance
- _Metabolic disorders including mitochondrial disorders and biopterin deficiency
- _Lipid disorders such as metachromatic leukodystrophy
- _Inherited disorders such as neurodegeneration with brain iron accumulation (NBIA) previously known as Hallervorden-Spatz disease and Rett syndrome

History

- _Perinatal adverse event history suggests CP
- _Age at onset of concerns and at onset of dyskinesia may help differentiate between CP and glutaric aciduria I (GTA1 macrocephaly)
- _No relationship of onset of symptoms to illnesses (as seen in Sydenham’s chorea)
- _No response to a trial of low dose Levodopa in CP (as seen in dopa responsive dystonia)
- _Cognitive regression goes against a diagnosis of CP
- _Consanguinity or Jewish ancestry

Pitfalls

■ _Missing a treatable cause such as DOPA responsive dystonia or glutaric aciduria I

Red Flags

■ _Neck pain with progressive weakness in an individual with dyskinetic CP may indicate compression of the cervical nerves or cord from disc degeneration, listhetic instability, or cervical stenosis.

Treatment

Medical

■ _Trihexyphenidyl—an anticholinergic antiparkinsonian agent that is useful in treatment of dystonia

■ _Levodopa—particularly useful in dopa responsive dystonia

■ _Levetiracetam—may be helpful in the management of choreoathetosis

■ _Tetrabenazine—is a dopamine depleting agent and is reportedly useful in the treatment of hyperkinesias, particularly chorea associated with Huntington's disease

Exercises

■ _General strengthening and stretching of the dystonic muscles

■ _Truncal strengthening for stability in sitting

Injection

- _Focal botulinum toxin injections
- _Phenol injections

Surgical

- _Deep brain stimulation
- _Intrathecal baclofen therapy is helpful in treating dystonia

Complications

- _Cervical disc degeneration starts earlier and progresses more rapidly, often starting in late adolescence or early adulthood and is generally present in over 97% of patients beyond 35 years of age
- _Listhetic instability and narrowing of the cervical canal are a common occurrence and combined with the disc degeneration 9
predisposes individuals with athetoid CP to rapidly progressive devastating neurological deficits

Prognosis

- _Although the brain lesion is considered “nonprogressive” in CP, dyskinesia may progressively worsen, especially in late adosecence or adulthood.