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Reference Review And Discussion On Prostheses

&

Cerebral Palsy: Gross Motor Function Classification System IV–V

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Cerebral Palsy: Gross Motor Function Classification System IV–V

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Description

Cerebral Palsy (CP) is a group of disorders affecting the development of movement and posture that are generally nonprogressive and affect the developing fetal or infant brain. Gross motor classification IV–V individuals have very limited functional mobility. 11

Etiology/Types

- _GMFCS IV—some evidence of head and trunk control, powered mobility possible
- _GMFCS V—Very limited head and trunk control, usually no independent mobility
- _Topology—Most are quadriplegic, one side of the body can be more affected
- _Tone disorder—Spasticity usually combined with some dystonia/dyskinesia

Pathogenesis

- _Damage is more global and anoxia is a frequent cause

Risk Factors

- _Extreme prematurity
- _Multiple pregnancy, especially with twin-twin transfusion
- _Intrauterine infection
- _Birth trauma resulting in anoxia (i.e., placental abruption and severe pre-eclampsia)
- _Postbirth trauma, that is, traumatic brain injury and shaken baby, early episodes of meningitis

Clinical Features

- _Often floppy at birth
- _Characterized by persistent primitive reflexes (i.e., asymmetric tonic neck reflex, exaggerated and persistent startle reflex, and persistent palmar/plantar grasp reflexes)
- _Motor impairment—spasticity usually in the flexor muscle groups of the extremities; may have hypotonia in the trunk; lack of selective motor control; dystonia is common
- _Sensory impairment—involving all types of sensation, sight is also commonly impaired
- _Cognitive impairment—common but by no means universal

Natural History

- _Nonprogressive, (although some question about “early aging”)
- _Growth is commonly associated with contracture, joint dislocation, and scoliosis, leading to increased functional deficit and discomfort if not treated
- _There are a number of medical issues that can result in early death

Diagnosis

Differential diagnosis

- _Brain tumor
- _Dopamine dependent dystonia
- _Familial spastic paraparesis
- _Muscular dystrophies
- _Genetic disorders
- _Metabolic disorders

History

- _Premature birth with complications
- _Maternal or infant infection
- _Severe delay in gross motor and fine motor skills
- _History of a seizure disorder, failure to thrive, decreased pulmonary function, strabismus, constipation, and dysphagia

Testing

- _Cranial ultrasound in newborn often shows grade III–VI intraventricular hemorrhage
- _Magnetic resonance imaging of the brain commonly shows diffuse damage and evidence of atrophy
- _Metabolic and thyroid workup to look for treatable causes
- _Genetic testing to look for cause, expected course, and future risk

Red Flags

- _Changing neurologic picture—NOT CP

Treatment

Medical/Surgical

■ _Spasticity

• Medications commonly used include baclofen, dantrolene, zanaflex, and valium

• Intrathecal baclofen useful for increasing comfort and ease of care, but complication rate increased in children who undergo multiple hip and spine surgeries

• Injection therapy with botulinum toxin and/or phenol can be targeted to improve function of the less involved arm to allow for powered mobility, or to improve ease of care for dressing and hygiene

• Selective dorsal rhizotomy surgery can be used in this population

• ■ _Seizures

• Often difficult to control, requiring multiple medications

• Ketogenic diet often used and can be an easier option for tube fed children

• Vagal nerve stimulator also used

• ■ _Pulmonary issues

• Reactive airway disease, especially with children who were premature

• Obstructive lung disease also common and can lead to sleep apnea

• Restrictive lung disease develops in children with scoliosis or severely decreased chest wall expansion due to truncal hypotonia

• Pneumonia is a frequent cause of death in severely involved individuals

■ _Gastrointestinal issues

– Failure to thrive develops in infants due to poor oral motor function and often necessitates G-tube

– Constipation requires management with stool softeners, laxatives, suppositories, and/or enemas

■ _Orthopedic issues

• Joint contractures treated with tendon releases

• Hip subluxation initially treated with tendon releases and bony reconstruction, persistent subluxation may be treated with girdlestone procedure

• Scoliosis usually treated surgically when the curve reaches 60°. Thoracic lumbosacral orthosis (TLSO) bracing not usually helpful in decreasing curvature. Vertical expandable prosthetic titanium rib (VEPTR) is an option for very young children with bad curves by expanding and supporting a deformed thorax using telescoping rods

Exercises

■ _Range of motion is the mainstay of therapy
■ _Strengthening may be possible
■ _Weight bearing in standers or limited ambulation in supportive gait trainers
■ _Developmental stimulation
■ _Speech and language therapy for adaptive communication
■ _Swallowing therapy

Equipment

■ _Small children
• adaptive seating systems can often be transferred from an immobile base for use at home to a stroller for use in the community
• adaptive high chairs
• adaptive car seats
• standers
• gait trainers
• bracing including ankle-foot orthoses, thumb splints, and wrist extension splints
• trunk supports such as neoprene vests or TLSOs to improve trunk stability

■ _Older children
• wheelchairs—manual or electric
• seating either with customized lateral supports or custom molded seating
• must be safe to tie down for transportation on school bus
• wheelchair ramps and accessible vans
• lifts

Consults 13

- _Orthopedic surgery
- _Neurosurgery
- _Ophthamology for strabismus
- _Gastroenterology/surgery
- _Neurology
- _Sleep specialist
- _Pulmonology
- _ENT
- _Psychology

Prognosis

- _Survival into adulthood is the norm, with improved survival over the past 20 years.
- _Early death often due to pneumonia, intractable seizures