

Möbius Syndrome

- Described by Möbius as “Congenital Facial Diplegia” with bilateral abducens palsy in 1888
- Diplegia= symmetrical bilateral palsy

Cause

- Pathogenesis is unclear
- Deletion/translocation in long arm of chromosome 13 in a few families
- Timing of insult 4-6 weeks gestation when cranial nerve nuclei are rapidly developing
- Trauma, illness or toxic exposure

Clinical Features

- 6th and 7th nerve palsies
 - Usually bilateral but may be asymmetric
 - Esotropia most common
 - “Mask-like” facies
 - If incomplete palsy- upper division of facial nerve involved

Other Ocular features:

- Small palpebral fissures
- Epicanthal folds
- Hypertelorism
- Exposure or neurotrophic keratitis
- Situs inversus of retinal vessels
- Entropion
- Ptosis
- Head tilt
- Amblyopia
- Gaze palsy

Other Systemic Features

- Extremities
 - Syndactyly, polydactyly, brachydactyly, agenesis of digits, clubfoot
- Swallowing and speech abnormalities from cranial V, IX and X palsies
- Craniofacial abnormalities
 - Micrognathia, Microstomia, Ear abnormalities, Bifid uvula, cleft palate
- Dextrocardia

- Defective musculature
 - Missing pectoral and trapezius muscles
 - Absence of sternal head of pectoralis major
- Rib defects
- Tongue hypoplasia
- Mild Mental retardation

First signs:

- Difficulty sucking, drooling, incomplete closure of eyelids
- Lack of smiling response
- Other cranial nerve abnormalities:
 - V, IX, X and XII can be involved
- Craniofacial abnormalities

Evaluation

- Strabismus (Esotropia)
- Amblyopia
- Corneal health
- Pediatric Genetics evaluation
 - Craniofacial abnormalities in differential diagnosis
 - Nager syndrome (acrofacial dysostosis)
 - Neuromuscular disease in differential diagnosis
 - Facioscapulohumeral (FSH) muscular dystrophy- shoulders primarily affected
 - Congenital or infantile myotonic dystrophy (slow relaxation of muscles after contraction)
 - Muscle wasting, cataracts, heart conduction defects, endocrine abnormalities
 - Charcot-Marie-Tooth disease
 - progressive loss of muscle and touch sensation in extremities
 - Usually in late childhood or early adulthood
 - Often first present with foot drop and claw toe

Strabismus Management

- Abnormal Extraocular muscles
 - Hypoplasia, aplasia and fibrous bands
- Forced duction testing
- Vertical Rectus transposition
- Medial rectus botox (more effective if muscle not tight)
- Medial rectus recession

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