Moebius Syndrome

Synonym: Moebius sequence, Mobius sequence/syndrome, congenital facial diplegia

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This is a complex developmental disorder of the brainstem, where there is a congenital facial palsy (usually bilateral) in association with impairment in abduction of the eyes (also usually bilateral). As well as involving the facial (VII) and abducens nerves (VI), other cranial nerves can be involved.

It was described by Paul Julius Moebius, a German neurologist who lived from 1853 to 1907.

Pathogenesis

This has been found to include cranial nerve and brain stem hypoplasia. Most cases are sporadic mutations, but an autosomal dominant transmission has been described (chromosome 13).

Risk factors

Misoprostol, ergotamine, and chorionic villus sampling have been suggested as possible environmental factors.

Associated abnormalities

Orofacial dysmorphism

- Micrognathia (64% of patients), ocular hypertelorism (25%), epicantal folds (89%), external ear defects (47%), high-arched palate (61%), tongue hypoplasia (77%), teeth defects (37%)

Limb malformations

- Syndactyly, oligodactyly, brachydactyly, arthrogryposis, absent trapezius or pectoral muscles, lower limb deformities in 69% eg club foot

Mental retardation

- IQ is low in 50%. Disturbances in psychomotor and speech development are very common. The incidence of autistic spectrum disorders is 30% in patients with Moebius sequence.

Neurological sequelae

- Hypotonia, pharyngeal weakness, feeding difficulties.
- Respiratory difficulties in infancy.
- Delayed developmental milestones.

Recognition

Because early recognition of Moebius syndrome can lead to early diagnosis and treatment, education of nurses in perinatal, paediatric, midwifery, and neonatal specialties is crucial. Nurses can offer anticipatory guidance and provide resources to parents of children with this condition.

Management

This is supportive and will include physical, psychological and educational resources:

- Infants sometimes require special bottles (ie Special Needs or Pigeon Feeder) or feeding tubes to maintain sufficient nutrition.
- Children with Moebius syndrome usually benefit from physical and speech therapy to improve their gross motor skills and coordination, and to gain better control over speaking and eating.
- Occupational and sensory integration therapies are also beneficial.

Surgical
Strabismus is usually correctable with surgery.
Limb and jaw deformities may often be improved through surgery.
Plastic reconstructive surgery of the face can offer benefits in individual cases, in that surgery, nerve and muscle transfers to
the corners of the mouth have been performed to provide an ability to smile.

Prognosis

- Children may crawl and walk later. Most children with Moebius Syndrome eventually catch up.
- Speech problems often respond to therapy, but may persist due to impaired mobility of the tongue and/or mouth.
- As children get older, the lack of facial expression and an inability to smile may become the dominant visible symptoms.

Further reading & references

- Moebius Syndrome Foundation
  1. Moebius Syndrome; MBS, Online Mendelian Inheritance in Man (OMIM)

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