Strabismus surgery in congenital cranial dysinnervation disorders

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Congenital cranial dysinnervation disorders

- Result from abnormal development and/or connectivity of cranial motor neurons
- Extraocular muscles lack normal innervation
- Extraocular muscles misinnervated

CCDDs

CCDDs

- Duane syndrome
- Moebius syndrome
- Monocular Elevation Deficiency with Marcus Gunn Jaw Winking

Case 1: Duane syndrome

Exam

- Vcc: 20/20 OD and OS
- Gross stereopsis in right face turn
- Cret:
  - OD - 8.75 + 0.50 x 090
  - OS - 7.00 DS

37 year-old male with 15 degree right face turn
Left exotropia and hypertropia in forced primary gaze

Globe retraction and lid fissure narrowing on adduction

Limitation to abduction

Duane syndrome

Decreased normal LR innervation

Anomalous LR innervation

Anomalous LR innervation

- Globe retraction and lid fissure narrowing on adduction
- Limitation to abduction

Decreased normal LR innervation

Surgical goals

- Improve face turn
- Improve alignment in primary gaze
- Improve globe retraction
- Eliminate upshoot
- Maintain ocular rotations

Surgical plan

- #1 Weaken the lateral rectus
#1 Fixation of lateral rectus to orbital wall

#2 Create a new abducting force

- Partial tendon vertical rectus transposition to lateral rectus insertion with posterior fixation

Exotropia improved, left hypertropia eliminated
Globe retraction improved
Abduction deficit unchanged

Upshoot eliminated; ocular rotations preserved
Case 2: Moebius syndrome

Moebius Syndrome
Facial weakness (VII nerve or nucleus)
+
Abduction deficit or horizontal gaze palsy (VI nerve or nucleus)

Moebius syndrome
- 16 y o male
- Bilateral CN 6 and 7 palsies
- >90 PD V pattern esotropia
- bilateral MR recession at age 2 years
- bilateral MR disinsertion at age 6 years
OD +3.00 +1.50 x 090  20/30-
OS -1.00 +2.50 x 075  20/30-

Left face turn

Surgical goals

- Improve left face turn
- Improve esotropia
Surgery

- Forced duction tests: marked restriction to abduction OU
- Both MR muscles found 16 mm post to the limbus

Surgery

- RMR free tenotomy*
- LMR recessed an additional 4 mm
- Mitomycin C OU
- Recession conjunctiva/tenon’s OU
- Amniotic membrane graft OU

Postoperative exam

- Face turn eliminated
- RXT 30 PD
- Transient diplopia with ARC

Postoperative exam

- 0
- 0
- 0
- 0
- 0
- 0

-4
-5
-4
-4

preop
post op
preop
post op
Case 3:  
Monocular elevation deficiency + MGJW ptosis

Marcus Gunn Jaw Winking Syndrome

- Ptosis and hypotropia due to deficient normal innervation of the levator and SR by the superior division of CN3
- Jaw wink due to synkinesis between superior division of CN3 and V3

18 mos old boy with MGJW
Clinical features

- MGJW RUL ptosis
- 25Δ R hypotropia and R supraduction deficit
- 30Δ R esotropia
- Ambylopia OD→ patching & spectacles (+2.50 OU)

Surgical goals

- Address the strabismus
- Correct the (MGJW) ptosis of the right upper eyelid

Surgery

- Right Hypotropia
  - FD test negative for restriction to elevation of right eye
  - Right IR recession
  - Transposition of half of the right MR & LR to SR insertion
  - Posterior fixation sutures
- Esotropia
  - Left MR recession & Left LR resection
- MGJW RUL Ptosis
  - Right frontalis suspension
Postop

- 5-10° RXT, 5° RHoT
  - Discontinued hyperopic spectacles
- Improved ptosis

Summary

- Congenital cranial dysinnervation disorders
  - Duane syndrome
  - Moebius syndrome
  - MGJW w monocular elevation deficiency

Surgical approach to treating strabismus due to CCDDs
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- Surgical tables do not apply
- Relieve restrictions
- Don’t operate on muscles that have no innervation (consider transpositions)
- Don’t operate on too many rectus muscles at once
Surgical approach to treating strabismus due to CCDDs

- Surgical tables do not apply
- Relieve restrictions
- Don’t operate on muscles that have no innervation (consider transpositions)
- Don’t operate on too many rectus muscles at once
- Avoid free tenotomies

Thank you