

Chronic Progressive External Ophthalmoplegia: Diagnosis and Management

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CPEO - Purpose

- Present clinical and diagnostic aspects of 3 patients with chronic progressive external ophthalmoplegia
- Describe the results of strabismus surgery in 2 of these patients

CPEO - Introduction

- First described by Von Graefe in 1868
- Mitochondrial encephalomyopathy
- Onset: age 11-82 years
- Progressive bilateral ptosis
- Progressive paresis of motility with exotropia
 - Diplopia rare

CPEO – Introduction con't:

Systemic Involvement – CPEO plus

- Facial, limb muscles (60-90%)
- Endocrine (67%)
- Cardiac conduction disorders (26%)
- Ataxia and tremor (39%)
- Polyneuropathy (23%)
- Dementia/ other CNS abnormalities (13%)
- Vestibular dysfunction / hearing loss

Methods

- 3 patients with chronic, progressive, EOM weakness and strabismus referred for possible strabismus surgery
- Complete ophthalmologic, neuroophthalmic and neurologic examinations were done
- Neuro-imaging, cardiograms, audiograms, DNA studies and muscle biopsy were performed when possible
- Surgery was performed in 2 patients to reduce the exotropia and correct the head turn

Case #1 (JD) – 32 yr old woman

Ophthalmologic exam

- Marked bilateral ptosis
- Frontalis overaction
- Chin-up position
- Poor levator function
- Pigmentary retinopathy



Case #1 - EOM's and Alignment



- **Adduction:** -5 OU
- **Abduction & Vertical gaze:** -3 OU
- **Saccadic velocity:** Poor OU
- **Alignment:** XT/XT'60, mild hypotropia OU. *No diplopia*

Case #1 Video





Case #1 – Systemic findings

- Heart block
- Mild sensory-neural hearing loss
- Proximal muscle weakness
- Ragged red fibers
- Decreased COX activity
- Elevated CSF protein

Kearns-Sayre Syndrome

Case #2: (JF) – 66 yr old woman

- Marked bilateral ptosis
- Frontalis overaction
- Chin-up position
- Exposure keratitis



Case #2: EOM's & Alignment Pre-Op



- **Adduction:** OD -6, OS -5
- **Abduction:** OD -3, OS -2
- **Vertical gaze:** Elevation & Depression -4 OU
- **Saccadic velocities:** Poor OU
- **Alignment:** XT/XT' 50, mild hypotropia OU

Case #2 - Video



Case #2: Surgery

- Bilateral Lateral Rectus Recession
 - OD 8mm
 - OS 7mm
- Bilateral Medial Rectus Resection 7mm
- Adjustable x4

Case #2: Post-op



- **Alignment:** XT/T' 5-10
- **Motility:** poor vertical and horizontal ductions and versions

Pre-Op



Post-Op



Improved head position

Case #3 (GM) – 49 yr old man

- Previous strabismus surgery age 12 yrs
- Amblyopia OS
- Progressive LXT since age 25 yrs
- Constant diplopia



Case #3 - EOM's & Alignment Pre-Op



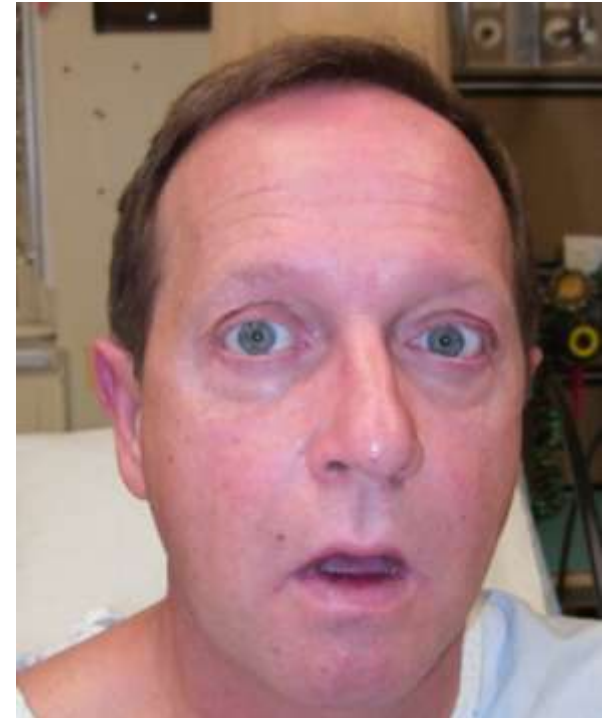
- **Adduction:** OD -4, OS -2
- **Abduction:** OD -1.5, OS -2.5
- **Vertical gaze:** Elevation -3 OU, Depressions Normal
- **Saccadic velocities:** Poor OU
- **Alignment:** XT/XT' 20, mild Hypotropia OU

Case #3 - Video



Case #3: Surgery #1

- Bilateral Medial Rectus Recession
 - OD: 5.5mm
 - OS: 6.5mm
- Left Lateral Recession from 13 to 16 mm from limbus
- Post-op
 - Head turn to left
 - LET/T' 15
 - Abduction: OS -4



Case #3: Surgery #2

(L) Med Recess from 7 to 9 mm from limbus



Post-op #2



- **Adduction:** OD -4, OS -2
- **Abduction:** OD -1, OS -4
- **Vertical gaze:** Elevation -3 OU, Depressions Normal
- **Saccadic velocities:** Poor OU
- **Alignment:** Ortho, mild Hypotropia OU

Discussion

- CPEO is a common presenting sign of a potential multi-system disorder
 - Ptosis and XT are the most common ocular manifestations
 - Cardiac workup critical
- Surgery possible for strabismus
 - Recess/Resect most common procedure
 - Large corrections usually necessary

Discussion cont. – Surgery

- Case 2
 - For cosmesis
 - Required large corrections
 - Ptosis still an issue
- Case 3
 - Complicated by previous surgery
 - Had diplopia
- Both used recess/resect procedures on adjustable sutures

References

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