Special forms of Strabismus

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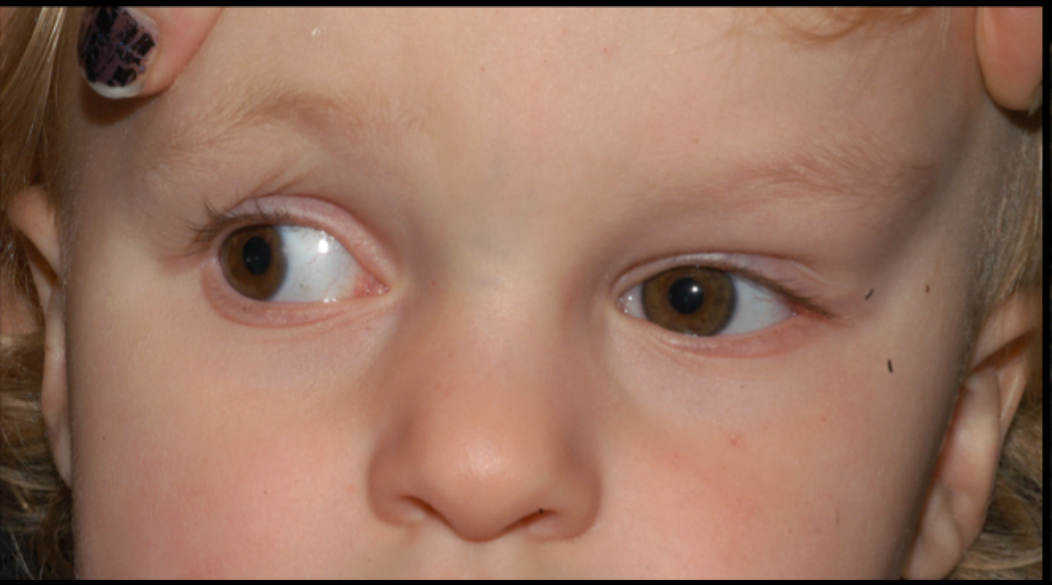

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Duane Syndrome

- Congenital abnormality of the 6th cranial nerve
  - 4th-8th week of gestation
  - Missing nucleus in most cases (4th week)
- Abnormal innervation from 3rd cranial nerve

Type	Abduction	Adduction	Deviation	Frequency
I	Poor	Normal	Ortho- or Esotropia	70%
II	Normal	Poor	Ortho- or Exotropia	15%
III	Poor	Poor	Ortho-, Eso- or Exotropia	15%

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Duane Syndrome

- Clinical Characteristics
  - Most sporadic, 5% autosomal dominant inheritance
  - More common in females than males.
  - OS affected more often than OD.
  - 80% of cases are unilateral, when bilateral it is usually asymmetric.

Female	Male	OS	OD	OU
58%	42%	59%	23%	18%

Gender and Laterality in 835 cases of Duane Syndrome
DeRespinis PA, Wagner RS, Guo S; Duane's Retraction Syndrome. Surv Ophthalmol 38:257, 1993

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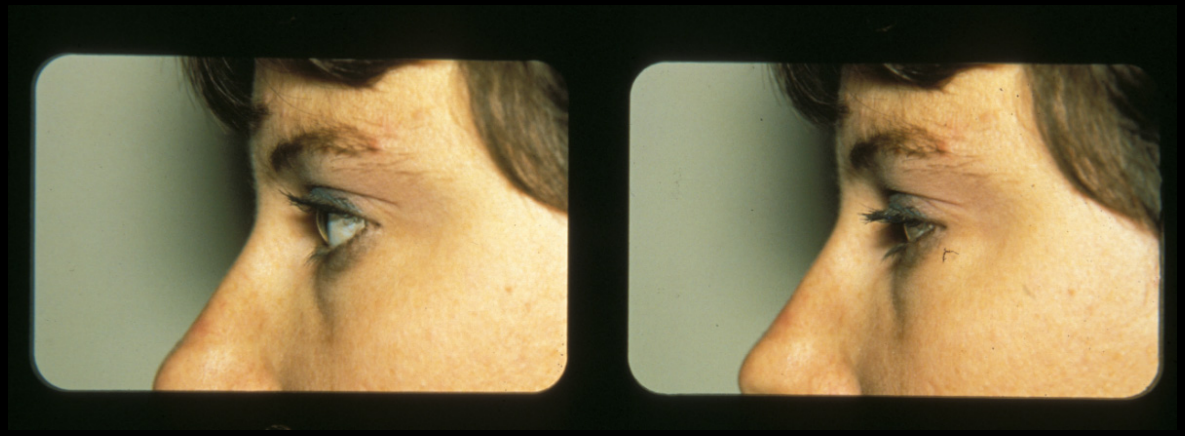
Duane Syndrome

- Exam findings:
  - Narrowing of the palpebral fissure and retraction of the globe of the involved eye on attempted adduction.
  - The amount of limitation depends on the amount of co-contraction of the medial rectus and lateral rectus.
  - Abnormal firing of the lateral rectus is found with EMG testing.



Duane Syndrome

- Exam Findings:
  - Angle of deviation almost always less than 30D , usually less than 15D .
  - Face turn toward the side of limited movement- permits fusion.
  - Anisometropic Ambylopia 10-30%
  - Ipsilateral congenital hearing loss 5%

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Duane Syndrome

- Selected Associated Syndromes:
  - Most cases are isolated
  - **Goldenhar syndrome** (hemifacial microsomia, ocular dermoids, ear abnormalities, preauricular skin tags, eyelid colobomas)
  - Wildervanck syndrome (sensorineural hearing loss, Klippel-Feil anomaly- shortness of neck with fused or missing vertebrae)

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Duane Syndrome

- Treatment
  - Treat Refractive Error
  - Consider treating Hyperopia ≧ +2.00 if ET
  - Treat Amblyopia

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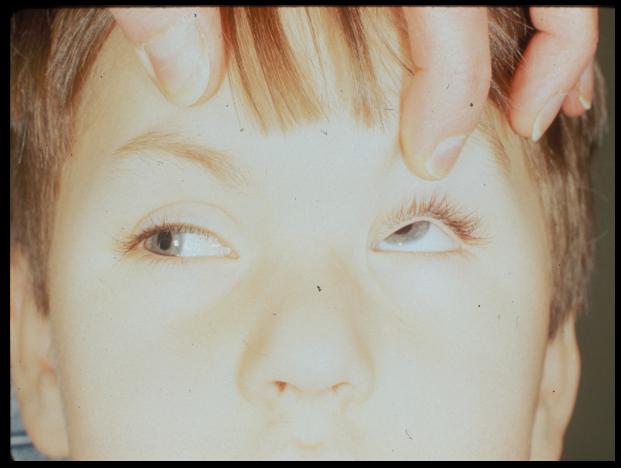
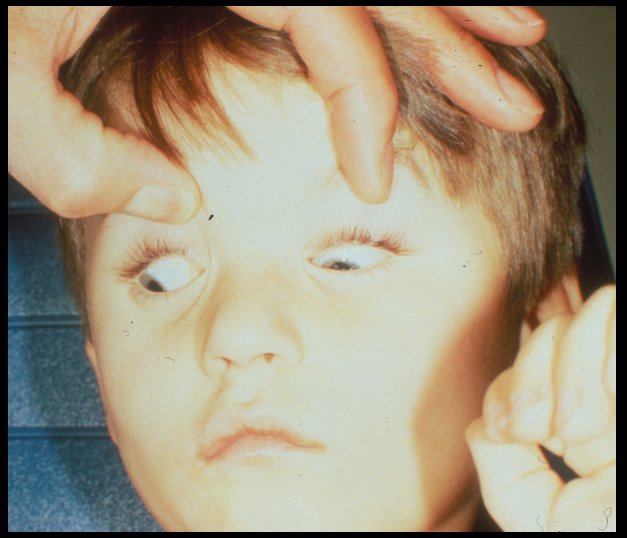
Duane Syndrome

- Goals of Surgery:
  - Improve primary position alignment
  - Improve head turn
  - Decrease up-shoot or down-shoot





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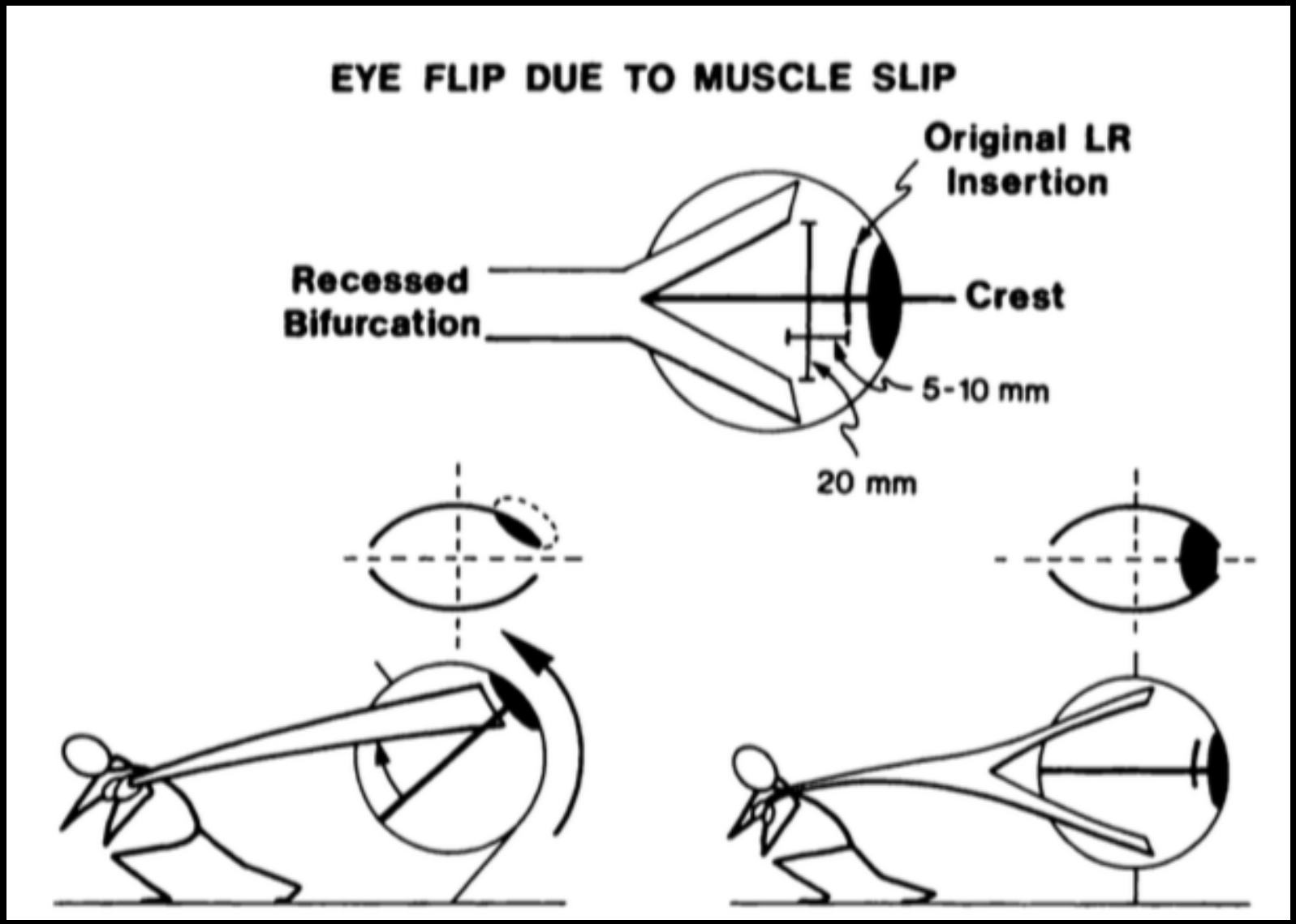
Duane Syndrome

- Surgical Treatment Possibilities
  - Eso-Duanes
    - Medial Rectus (MR) Recession
    - Bilateral MR Recessions
    - Some advocate:
      - SR and IR transposition to LR
      - SR transposition alone
  - Exo-Duanes
    - Lateral Rectus (LR) Recession(s)
  - Up-shoots/Down-shoots
    - Recession with Y split or posterior fixation of LR



Rosenbaum and Santiago eds. *Clinical Strabismus Management* p.336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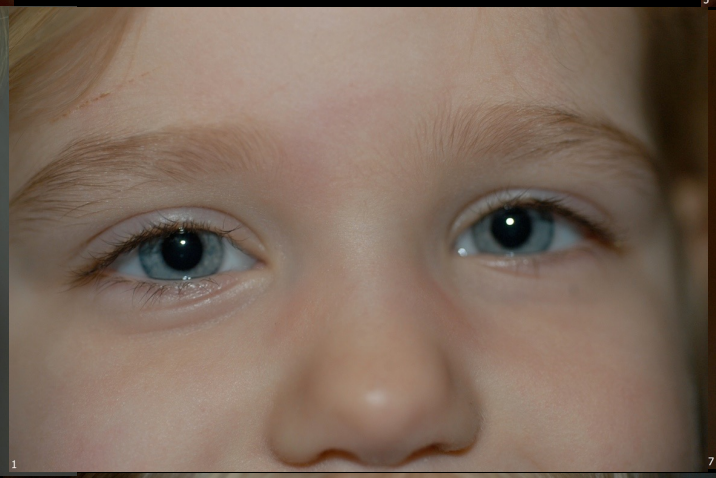
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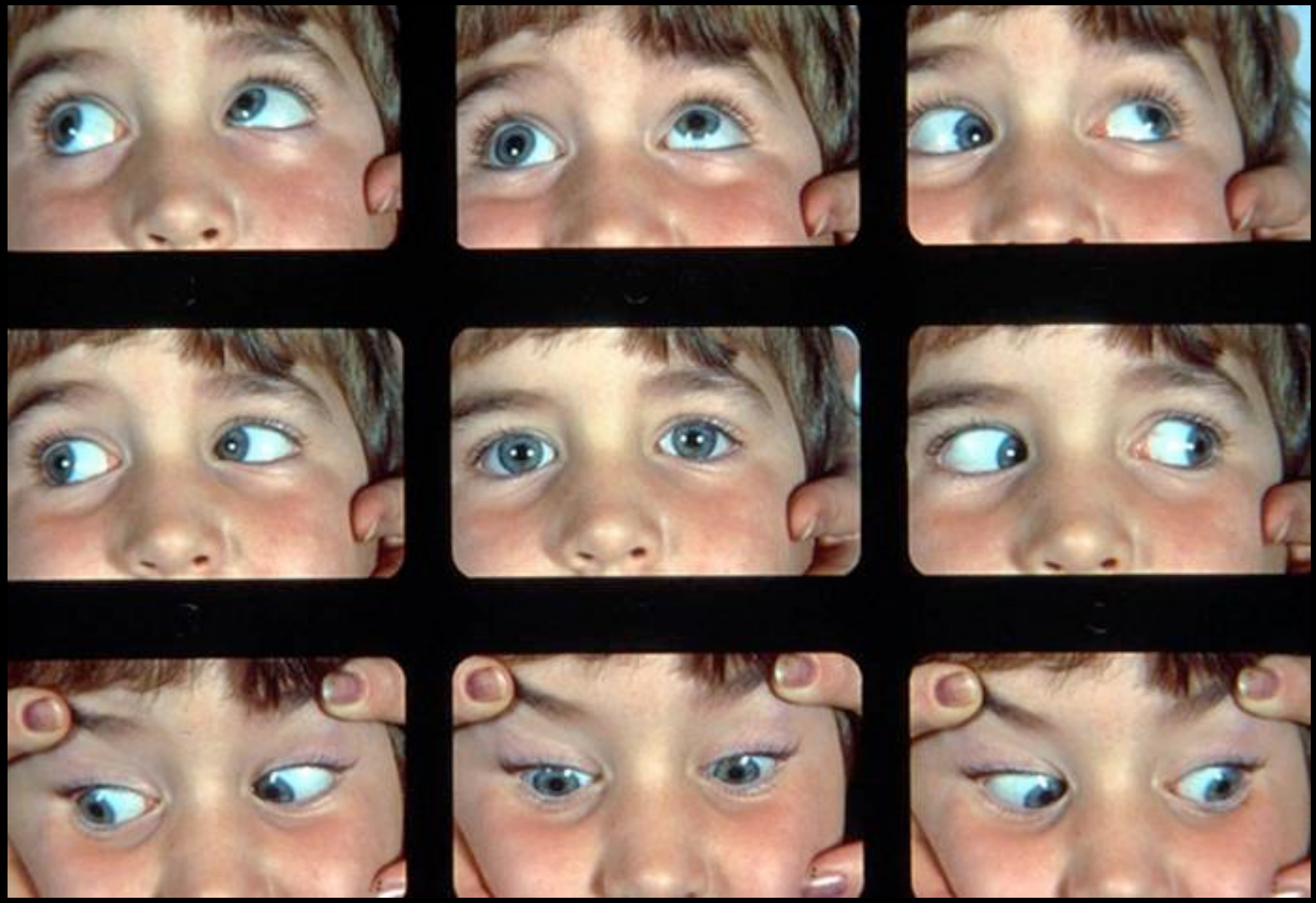

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Brown Syndrome

- Abnormality of the superior oblique tendon
- Limitation of elevation in adduction

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Brown Syndrome

- Clinical Characteristics
  - Deficient elevation in adduction that improves in abduction
  - Hypotropia
  - Chin up head position and/or face turn away from affected eye
  - Forced ductions show restriction to elevation in adduction that is worse with retropulsion*
  - V pattern*
  - Superior oblique function normal*

*Helps distinguish from Inferior oblique palsy

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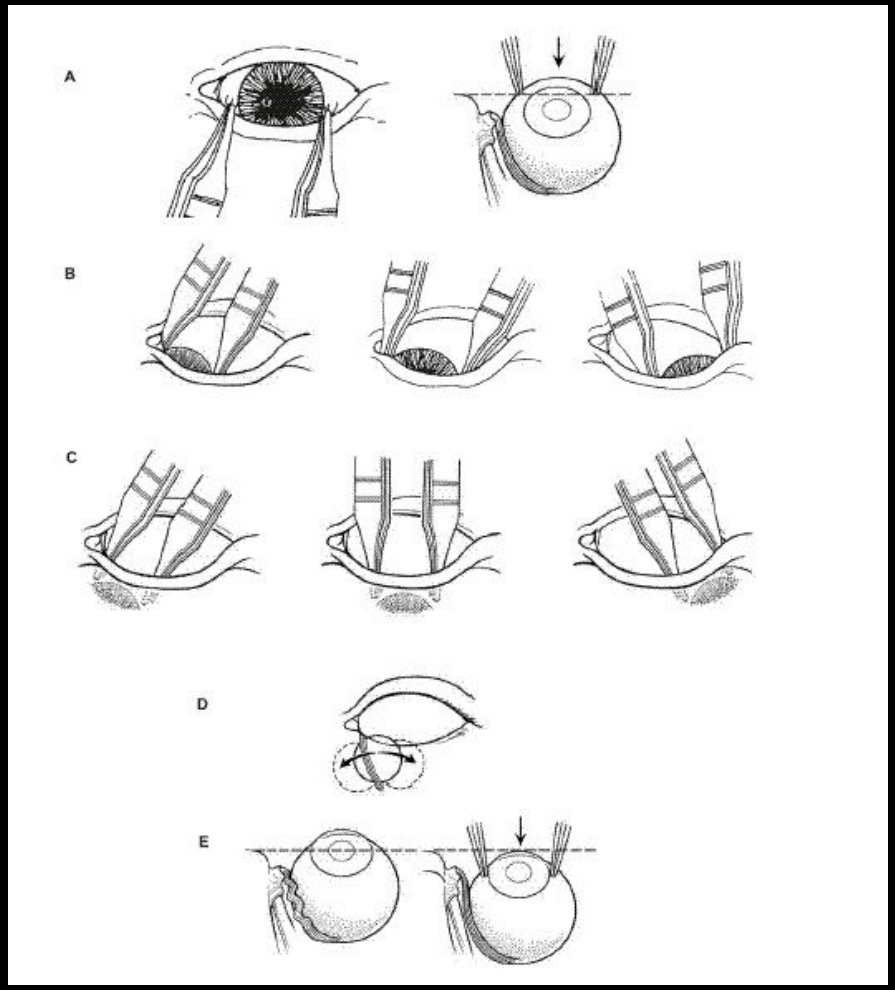
SO Traction Test



Surgical Management of Strabismus. Ch 5, 2005. Helveston

The superior oblique traction test (viewed from above the patient's head)

A. The eye is grasped at the 2 o'clock and 10 o'clock position (right eye from above)

B. The eye is pushed back into the orbit and is guided from nasal to temporal. As it goes over the normal superior oblique tendon, the eye 'pops' up.

C. With a lax or loose tendon the cornea disappears and remains hidden behind the upper lid as the eye is rotated.

D. The relative path of the globe as it passes over a normal tendon.

E. A lax superior oblique tendon allows the globe to be pushed backward into the orbit.

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Brown Syndrome

- Etiologies
  - Congenital tendon or trochlear abnormalities
  - Acquired
    - Trauma
    - Inflammatory
      - Sinusitis
      - Systemic inflammatory diseases: Rheumatoid arthritis

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Brown Syndrome

- Differential Diagnosis
  - **Isolated inferior oblique paresis**
  - Monocular Elevation Deficiency
    - Limited elevation and abduction and adduction
    - Ptosis or pseudoptosis
  - Congenital Fibrosis Syndrome
    - Restriction to elevation in abduction and adduction
    - Esotropia in limited upgaze more common
  - Blow-out Fracture
    - Limited elevation in adduction and abduction
    - Trauma, enophthalmos, infraorbital paresthesia, radiographic findings
  - Thyroid Eye Disease
    - Elevation limited in adduction and abduction (unless SO most affected- RARE)
    - Esotropia and typical clinical appearance
  - Fat Adherence Syndrome
    - Previous surgery history, Elevation limited more in Abduction

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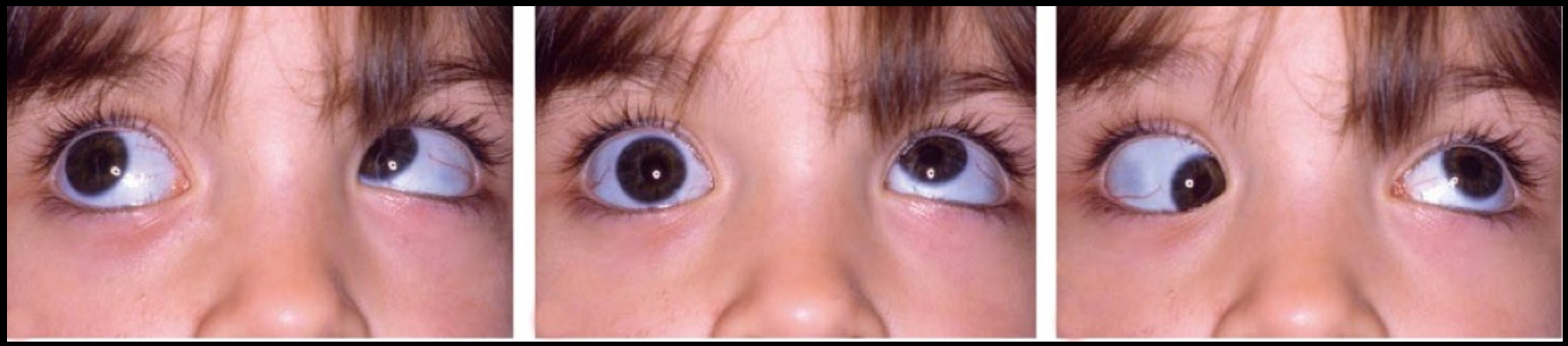
Brown Syndrome

- Treatment Goals
  - Improve Hypotropia in primary position
  - Improve chin up head position and face turn if present



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Brown Syndrome

- Treatment
  - Treat underlying inflammatory disease if present
    - Steroid injection into trochlear area
    - Oral non-steroidal anti-inflammatory agents
- U of Iowa Data suggests:
  - Most congenital cases remain stable

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Brown Syndrome

- Surgical Options
  - Superior Oblique tenotomy/tenectomy
    - Usually doesn't give SO palsy
    - Combined with IO recession
  - Superior Oblique tendon spacer
    - Silicone
    - Suture ("Chicken Suture")

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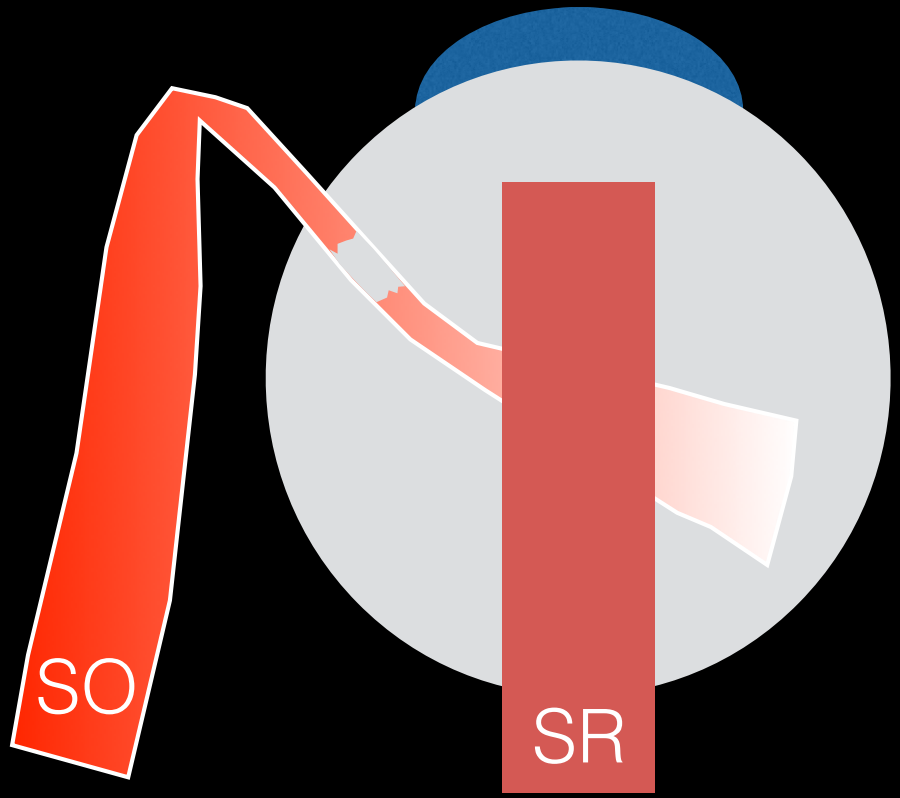
Superior Oblique Intra-sheath Tenotomy



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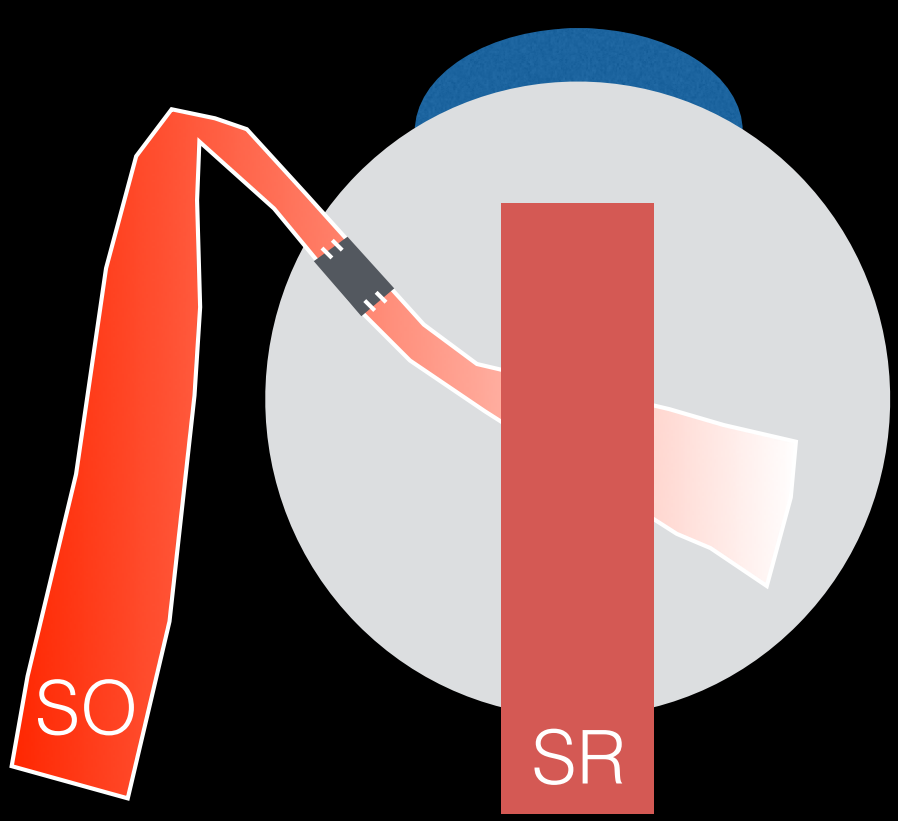
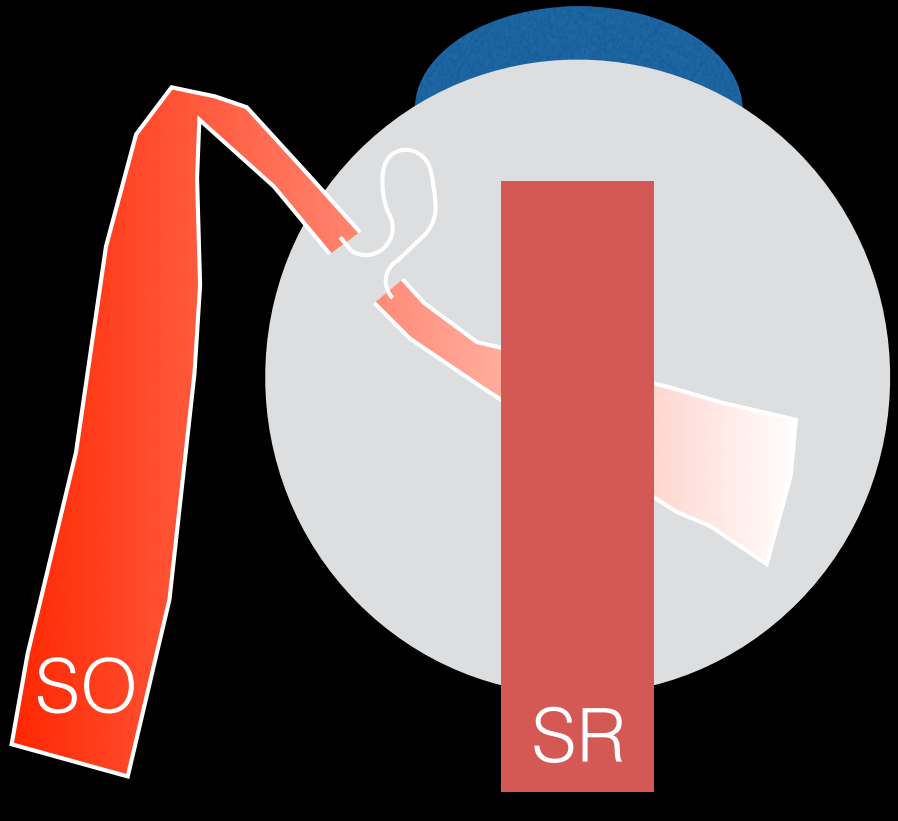
Superior Oblique "Guarded Tenotomy"



Silicone Spacer



Chicken Suture

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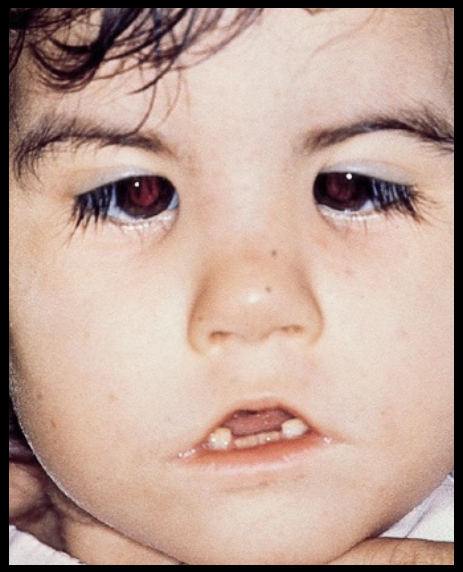
Möbius Syndrome

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



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Möbius Syndrome

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

Möbius Syndrome

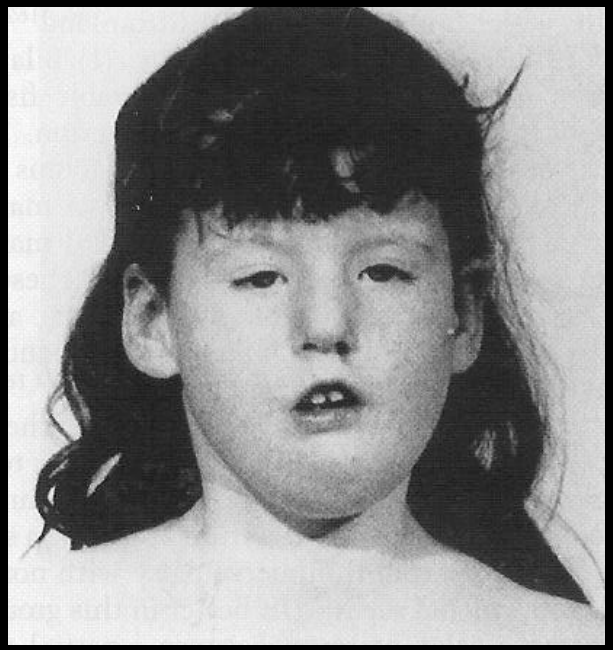
- Clinical Features
  - 6th and 7th nerve palsies
    - Usually bilateral but may be asymmetric
    - Esotropia most common
    - “Mask-like” facies
    - If incomplete palsy- upper division of facial nerve involved

Möbius Syndrome

- Clinical Features
  - 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

Möbius Syndrome

- 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



From Pediatric Ophthalmology and Strabismus. Wright and Spiegel. 2002

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Möbius Syndrome Systemic features

- Extremities
  - Syndactyly, polydactyly, brachydactyly, agenesis of digits, clubfoot
- Swallowing and speech problems
- 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

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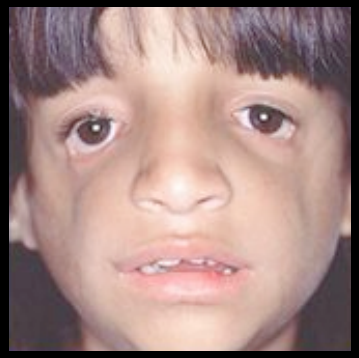
Möbius Syndrome

- Evaluation
  - Strabismus (Esotropia)
  - Amblyopia
  - Corneal health
- Pediatric Genetics evaluation
- Distinguish from other Craniofacial syndromes
  - Nager syndrome (acrofacial dysostosis)



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Möbius Syndrome

- Strabismus Management
  - Abnormal Extraocular muscles
    - Hypoplasia, aplasia and fibrous bands
    - Forced duction testing
  - Vertical Rectus transposition
  - Medial / lateral rectus surgery- incomplete

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Congenital Fibrosis Syndrome

- Non-progressive restrictive **ophthalmoplegia** often associated with **blepharoptosis**
- Severely **limited vertical gaze** and less limited horizontal gaze
  - Fibrous tissue replaces muscle
- **Cranial nerve dysfunction**
  - dysinnervation and aberrant innervation

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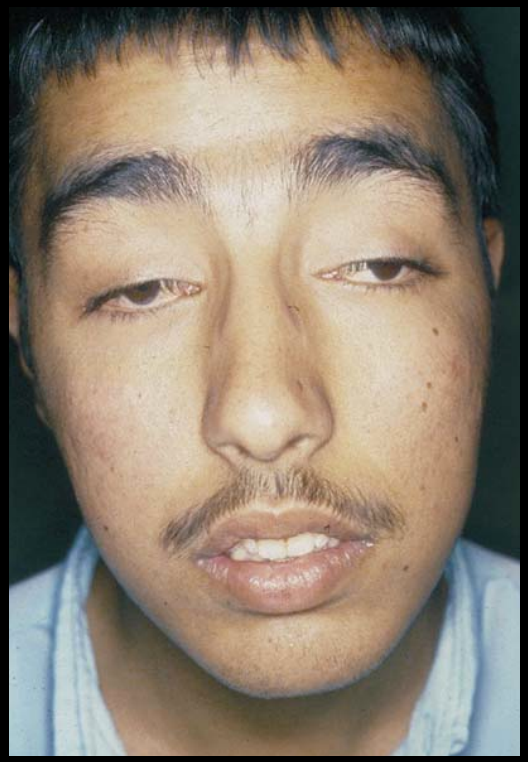
Congenital Fibrosis Syndrome

- **Type 1 "Classic" form**
  - Congenital non-progressive bilateral **external ophthalmoplegia**
    - Inability to elevate above the horizontal midline
  - Horizontal eye restriction is variable and may be severe
    - May have be orthotropic, esotropic or exotropic
    - Binocular vision usually absent
  - **Aberrant eye movements** are common
    - Eyes turn inward on attempted upgaze
    - Marcus Gunn Jaw winking
  - Congenital non-progressive bilateral **ptosis**
  - Frequently high astigmatism



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Congenital Fibrosis syndrome

- **Type 2**
  - **More severe restriction** to horizontal eye movements (exotropic)
  - Aberrant innervation less common
  - **Pupil abnormalities**
- **Type 3**
  - More variable presentation (unilateral or bilateral)
    - **Ptosis may be absent**
    - Restriction is more variable horizontally
    - Other systemic findings
- **Tukel Syndrome**
  - Type 3 plus postaxial oligodactyly or oligosyndactyly of hands

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Congenital Fibrosis Subtypes

Type	Laterality	Inheritance	Gene	Details
CFEOM 1A	Bilat	AD	KIF21A	Classic Form Aberrancy common
CFEOM 1B	Bilat	AD	TUBB3	Like classic Form
CFEOM 2	Bilat	AR	PHOX2A	Typically XT Horizontal EOM severe Less aberrancy
CFEOM 3A	Uni or Bilat	AD	TUBB3	Intellectual and social disabilities Progressive senorimotor axonal polyneuropathy Dysgenesis of Corpus Callosum, cortical spinal tracks, basal ganglia
CFEOM 3B	Uni or Bilat	AD	KIF21A	
CFEOM 3C	Uni or Bilat	AD	translocation	Single family
Tukel Synd	Uni or Bilat	AR	22q	Like type 3 with hand abnormalities

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Congenital Fibrosis syndrome

- Diagnosis
  - Clinical findings
  - Clinical testing for KIF21A, TUBB3 and PHOX2A
- Ddx
  - 3rd nerve palsy
  - chronic progressive external ophthalmoplegia
  - myasthenia gravis

Binocular Vision and Ocular Motility: Theory and Management of Strabismus. 6th ed p. 476 by von Noorden and Campos

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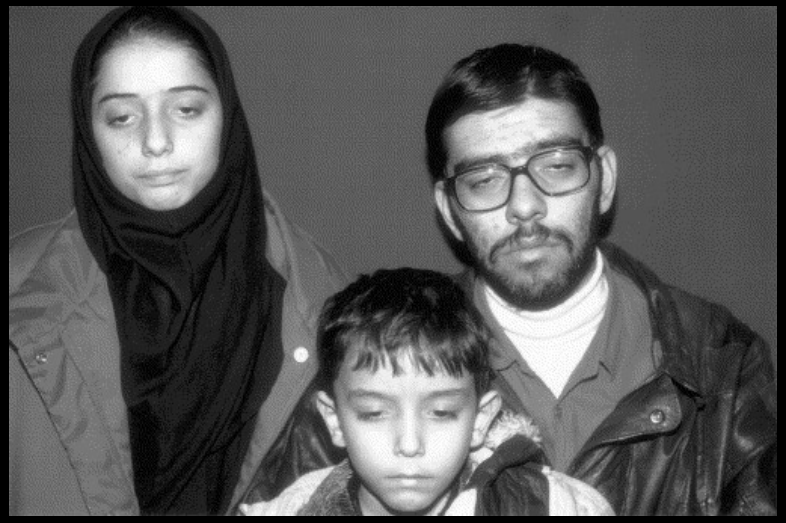
Congenital Fibrosis syndrome

- Treatment
  - Correct Refractive Error
  - Treat Amblyopia



Yazdani & Traboulsi. Ophthalmology. May 2004.111(5);1035-1042

- Surgical
  - Strabismus surgery before ptosis surgery
  - Goals: primary position alignment, improve chin up

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Congenital Fibrosis syndrome

- Intraoperative forced duction testing
- Large recessions or “free tenotomy”
- Recession of conjunctiva and tractions sutures
- Resection has been used in select cases


CHEVROLET

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Superior Oblique Myokymia

- Uniocular paroxysms of small amplitude, high frequency rotary nystagmus.
- Etiology: unknown
- Described in:
  - Regeneration of damage to trochlear nerve- described in 2 patients
  - Tumor of midbrain (other neurological symptoms)

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Superior Oblique Myokymia

- Clinical Features
  - Intermittent bursts
  - Vertical and or torsional diplopia
  - “Shimmering” vision
  - Episodes last seconds, minutes or longer
  - Exacerbated by stress and fatigue

Superior Oblique Myokymia

- Examination findings
  - Normal eye movements in absence of symptoms
  - Rapid small- amplitude torsional eye movement
  - May only be visible at slit-lamp

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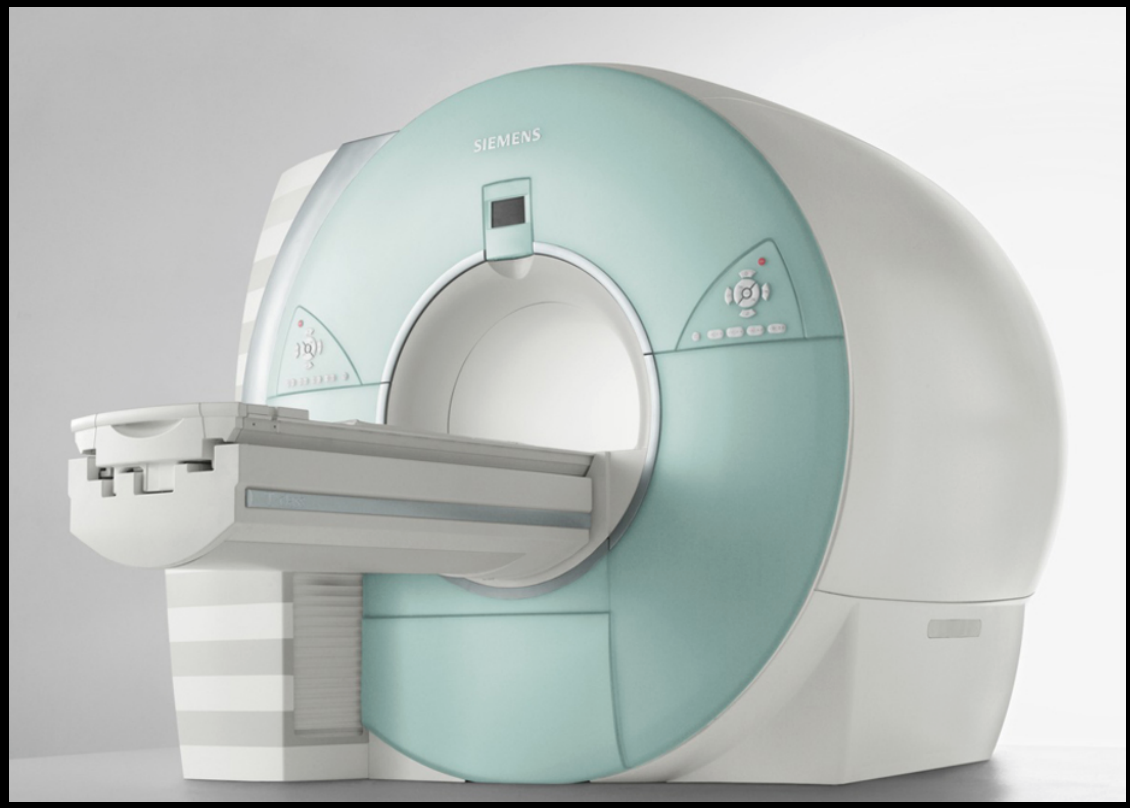
Superior Oblique Myokymia

- Work-up
  - None unless neurologic signs present



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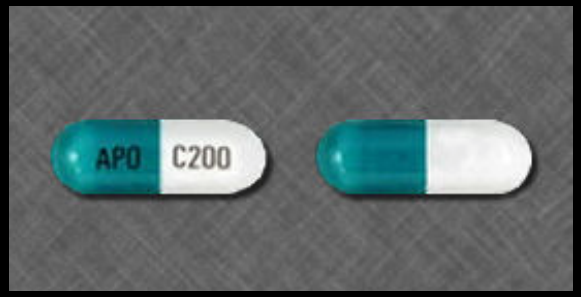
Superior Oblique Myokymia

- Treatment
  - Topical betaxolol BID
  - Carbamazepine
  - Phenytoin
  - Propranolol
  - Propranolol plus Valproic acid
  - Baclofen
  - Gabapentin
- Often Disappointing
- Side effects



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Superior Oblique Myokymia

- Surgical treatment
  - Tenectomy of Superior Oblique- large
  - Plus: Inferior oblique weakening

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Cyclic Heterotropia

- Manifest Strabismus that follows a regular rhythm of occurrence and disappearance
  - 48 hour cycles most common
  - First cases reported had esotropia for 2 days followed by orthotropia for 2 days- repeated.
    - *circadian, periodic, alternate-day esotropia*

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Cyclic Heterotropia

- Onset in early infancy
- On orthotropic days:
  - No abnormalities of binocular vision!
  - Heterophoria or refractive error are minimal if present
- On strabismic days:
  - Sensory abnormalities present (suppression)
  - Fusional amplitudes absent

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Cyclic Heterotropia

- Cycle continues for months to years
  - Progress to constant strabismus
- 1-4 day cycles possible
- Adult onset possible
- Cyclic esotropia most common

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Cyclic Heterotropia

- Cause Unknown
- Case Reports:
  - after removal of **third ventricle astrocytoma**
  - starting with the onset of **epileptic disorder**
  - after recovery from **closed head trauma and 6th nerve palsy**
  - after **surgery for intermittent exotropia**
  - after **uniocular traumatic aphakia**- resolved with secondary IOL implantation

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Cyclic Heterotropia

- Some have family history of “run-of-the-mill” strabismus
- Not associated with
  - Fatigue, accommodation, or disruption of fusion
- Contrast to “periodic alternating esotropia”
  - 1-2 minute cycles where eyes alternate fixation and alignment alternates between esotropia and orthotropia
    - Cerebellar atrophy

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Cyclic Heterotropia

- Cycle eventually breaks -> strabismus
- Surgery for the full amount of heterotropia present on strabismic days.