

Review of Strabismus

Revised 2002

Frederick M. Wang, M.D.

Clinical Professor, Ophthalmology and Visual Science
The Albert Einstein College of Medicine
Attending Surgeon, The New York Eye & Ear Infirmary

ESODEVIATIONS

CONGENITAL ESOTROPIA

- Characteristics

1. Onset confirmed by six months
2. Large angle (esp $> 40\Delta$)
3. Constant
4. Cross-fixation
5. Pursuit asymmetry
6. Occlusion nystagmus
7. Low plus refraction
8. Inferior oblique overaction

- Goals

1. Prevent amblyopia
2. Fusion (peripheral)
3. Cosmesis

- Management

1. Surgery
2. Amblyopia
3. Refractive

OTHER COMITANT ESOTROPIA

- 1. **Accommodative** (High plus vs. High AC/A)
- 2. **Sensory** (Poor vision one or both eyes)
- 3. **Nystagmus - Blockage syndrome** (convergence blocks a true nystagmus)
- 4. **Dyskinetic** (esp with cerebral palsy)
- 5. **Spasm of near reflex** (esp hysterical or post head trauma)

ESODEVIATIONS

INCOMITANT ESOTROPIA

1. Lateral Rectus Weakness
 - a. VIth nerve paresis (isolated)
 - Non-localizing CNS - increased pressure
 - Specific CNS - lesion along route
 - Demyelinating - post-viral, MS
 - Microvascular: diabetesRx: Wait 6-12 months for surgery. Consider botox before
 - b. Möbius
 - Bilateral VIth and VIIth nerve palsies
 - Gaze palsy
 - Midline cranial nerve (IX, XII, occ III) palsies
 - Defects chest and neck muscles
 - c. Duane's Types I & III
 - VIth N. palsy with aberrant lateral rectus innervation with branch from III rd. nerve
2. Medial Restriction
 - a. Thyroid
 - b. Medial wall fracture
 - c. Medial fat and fibrous trauma
 - d. Status post excessive resection

INCOMITANT HORIZONTAL STRABISMUS

- The horizontal deviation changes with horizontal gaze.
- May be:
 1. Paralytic
 2. Restrictive
 3. Co-contraction (esp. Duane's)
- To differentiate

	<u>Paralytic</u>	<u>Restrictive</u>
Forced Traction:	Free	Restricted
Forced Generation:	Weak	Normal
Lid Fissure in field of limited gaze	Widens	Narrows
IOP in field of limited gaze	Stable	Increases
Saccadic velocity:	Slow	Normal
Excursion:	Duction of paretic eye into paretic field is greater than the version movement of the paretic eye in the same field (the innervation to the paretic eye in version movement is controlled by the non-parietic eye)	Duction = Version

COMITANT EXODEVIATIONS

- **Etiologies**
 - Innervation
 - Topographic (with orbital anomaly)
 - Sensory

- **Types**
 - Intermittent vs Constant
 - AC/A ratio considerations:
 - divergence excess/basic/convergence insufficiency
 - Tenacious proximal fusion

- **Evaluation**
 - Observe proportion of time that exotropia is present
 - Cover testing
 - Measure with + 3.00 and -2.00
 - Patch test
 - Sensory status

- **Treatment**
 - Surgery
 - Refractive (esp over minus)
 - Occlusion
 - Prisms

INCOMITANT EXODEVIATION

- **Paralytic**
 - IIIrd N. paresis:
 - a. Children: congenital, traumatic, inflamm, migraine, aneurysm, tumor
 - b. Adults: diabetes, aneurysm, trauma, neoplastic
 - Lost medial

- **Restriction:** Fibrosis

- **Duane's II** (? VIth intact with IIIrd N. to MR and LR)

“A” AND “V” PATTERNS

- **Definition:** A differential in horizontal alignment between up and down gaze. For “A” greater than 10 diopters for “V” greater than 15 diopters.

- **Etiology:**
 - Oblique dysfunction
 - Overaction superior oblique = “A”
 - Overaction inferior oblique = “V”

 - Topographic -
 - Possible vertical offset horizontal muscles
 - Possible orbital pulley issue

- **Treatment:** **Surgery**
 - Weaken/strengthen obliques
 - Vertical offset of horizontal muscles:
Move medials to “point” of pattern and laterals to base of pattern
(i.e., for “V” move medials down/laterals up -
this is true for recess or resect of any muscle)

VERTICAL DEVIATIONS

- NOTE:**
- Vertical fusional amplitudes are small (in range of 8 diopters).
 - Once dissociated, any horizontal phoria becomes manifest

DIAGNOSTIC TESTS

1. Observe if abnormal head posture: implies fusion.
2. Three-step test (Parks)
Understand this test and why it works!
(Memory help: If tilt toward side of hyper makes hyper worse - it's an oblique)
3. Versions - obliques/vertical recti
4. Sensory - fusion
torsion (double Maddox rod)
5. Disc/fovea relation -
Fovea normally 0.3 ± 0.3 DD below disc center
6. Tests of paralysis/restriction

SUPERIOR OBLIQUE PALSY

- **Etiology**
 1. Congenital: IVth n. vs. lax tendon
 2. Closed head trauma (often misdiagnosed as orbital floor fracture)
 3. Other: demyelinating, diabetes, tumor, etc.
- **Monocular vs. Bilateral Palsy**
 1. 3-step if gaze or tilt reverses primary hyper anywhere - think bilateral
 2. If greater than 10 degrees of excyclotorsion - think bilateral
 3. If significant "V" pattern (esp. 20 diopters) - think bilateral

- **Treatment** - Think about field of action and cyclo effect of each operated muscle.
 1. Weaken antagonist ipsilateral inferior oblique: anterior transposition /recession
 2. Tuck or advance superior oblique
 3. Recess contralateral inferior rectus

DISSOCIATED VERTICAL DEVIATION

- **Characteristics**
 1. Eye drifts up (also abducts and excyclotorts) without comitant elevation of fixing eye.
 2. Usually bilateral (may be asymmetric)
 3. Brought on by inattention, occlusion, low illumination
 4. Occurs esp. with congenital ET (but may not appear for years) and poor binocular fusion status
- **Evaluation**
 1. Difficult to measure - varies, builds. Try simultaneous prism cover test
 2. Differentiate from IO overaction which
 - a. produces a true hyper
 - b. is worse in adduction
 3. To measure true hypertropia when DVD also present: Place increasing base down prism over the hypertropic eye until no up movement of opposite eye is noted on alternate covertest on uncovering the opposite eye.
- **Treatment**
 1. Nothing
 2. Surgery
 - a. Large recess SR (range 8-10 mm)
 - b. Recess SR 4 mm + Posterior Fixation Suture (Faden)

Note: Principle of Faden: Point of posterior fixation is new insertion for action.
As antagonist entire muscle acts. Therefore limits muscle only in field of action.

DOUBLE ELEVATOR PALSY - CONGENITAL

- Characteristics

1. Usually Monocular
2. Limited elevation across all fields
3. May have primary hypotropia
4. Chin elevation may be present for fusion
5. May have ptosis or pseudo ptosis (pseudo lessens with affected eye fixation)

- Etiology - unknown - multiple - theories

1. Paretic SR + IO
2. Paretic SR
3. Inferior restriction (primary vs. secondary)
4. Central
5. Combination

- Treatment: Surgery

- With inferior restriction: free IR (recess usually 6-8 mm)
Lid considerations - cut lower lid retractors or reattach them
- With no restriction: Knapp procedure - Transpose MR & LR to SR (corrects 35 prism diopters of hypo- can grade).
- Combination paretic and restriction - do both.

SPECIAL FORMS OF STRABISMUS

DUANE'S RETRACTION SYNDROME

- All types have in common that not only does IIIrd N. innervate its normal route but a branch goes to the lateral rectus producing co-contraction of the MR and LR on attempted adduction.
- TYPE I: Absent VI = limited abduction often ET less than 30 P.D.
- TYPE II: ? Present VI = can abduct (may be weak) usually XT
- TYPE III: Larger III n. branch to LR = limited abduction and adduction
- More females 85%, left eyes 85%, bilat 15%
- Head turns
- Up or down leash
- Associated: Goldenhar-Gorlin, deafness, vertebral abnormalities Thalidomide embryopathy
- Treatment:
 - a. Refractive errors (esp. Anisometropia)
 - b. For amblyopia
 - c. Surgery: Head Posture/Strabismus
 - 1) Type I: Recess MR \pm recess ipsilateral LR +/- recess contralateral medial rectus
 - 2) Type II: Recess LR +/- recess MR +/- recess contralateral lateral rectus
 - 3) Type III: Recess MR + Recess LR
- 4) Use of Faden in Duane's
 - a) To limit normal eye
 - b) To prevent leash (on affected LR)

BROWN'S SYNDROME

Characteristics

1. Limitation of elevation maximum in adduction.
Near normal elevation in abduction.
2. Limitation of elevation is restrictive: +
forced traction test
Duction = Version
3. Eye dips down in adduction
4. V pattern with XT in upgaze
5. No or minimal S.O. overaction
6. Widening palpebral fissure on adduction
- g. May have head turn or chin-elevation to achieve fusion

Differential Diagnosis

Brown's Syndrome

- a. Version = duction
- b. V pattern
- c. (+) Forced Traction Test
- d. No or little S.O. overaction

I.O. Paresis

- a. Duction greater than version
- b. A pattern
- c. (-) Forced Traction Test
- d. S.O. overaction

Etiology

- a. Congenital: Taut S.O. Tendon. Rarely other restrictive bands
- b. Acquired: Trauma
S.O. tenosynovitis
- arthritis

Treatment

- Congenital -
 - Indications:
abnormal head posture,
hypotropia in primary,
cosmesis.
 - Surgery
 1. Tenotomy S.O. \pm weaken ipsilateral IO
 2. Silicone spacer (6-7 mm) placed into S.O.
- Acquired:
 - Treat inflammation
non-steroidals,
steroid injection
 - ? surgery

SKEW DEVIATION

- Vertical deviation which may or may not be laterally comitant
- May simulate a specific muscle palsy
- Caused by brainstem or cerebellar disease
- Diagnosis by associated signs

THYROID MYOPATHY

- Restrictive myopathy
 - Esp.
 - a. Inferior rectus
 - b. Medial rectus
 - c. Superior rectus
 - d. Lateral rectus
- Rarely primary true paresis SR
- Beware post-op inflammation
- With Graves' disease - often euthyroid
- Muscle infiltrated (large), edematous, fibrotic
- Surgery: Relieve restriction. Try to avoid resections.

ORBITAL FLOOR FRACTURE

Clinical Dx:

- a. History
- b. Ocular muscle dysfunction
- c. Hypesthesia infraorbital nerve distribution
- d. Orbital emphysema

Imaging

Characteristics of “entrapment”

Note: Rarely find muscle trapped in fracture site but rather other tissue with septal connection to the muscle sheaths

- a. IR - hypertropia in primary increases with upgaze. Restrictive. May have ipsilateral hypertropia on extreme downgaze
- b. MR - restrictive esotropia

Rx entrapment:

Surgery - how long to wait is controversial.

Advocates vary immediate to 6 months

CONGENITAL FIBROSIS SYNDROME

A spectrum of conditions of replacement by fibrous tissue of the extra ocular muscles with resultant restriction and weakness. The spectrum ranges from isolated involvement of one muscle to involvement of all (including the levator).

- Types:
- a. General fibrosis syndrome
 - b. Congenital fibrosis of the inferior rectus muscle
 - c. Strabismus fixus (horizontal)
 - d. Vertical retraction syndrome (SR fibrosis)
 - e. Congenital unilateral fibrosis (general on one side)

- Etiology:
- a. Possible neurogenic
 - b. Possible myopathic

MYASTHENIA GRAVIS

- A disorder of neuromuscular transmission

Types:

	<u>Juvenile</u>	<u>Transient Neonatal</u>	<u>Persistent Neonatal</u>
ACR-AB	Often present	Transient after birth	Absent
Age at onset	Any age	Typically just after birth	First 12 months
Duration of symptoms	Some spontaneous remission	2-4 weeks	Life-long

Approaches to Rx

Anticholin- esterase Corticosteroid Thymectomy Plasmapheresis	Anticholinesterase Support: Resp & feeding Exchange transfusion	Anticholinesterase
---	---	--------------------

- May be purely ocular
- Esp. ptosis (fatigues) but may simulate any EOM problem at any one time. Variability is key to diagnosis. Diplopia frequent.
- a. Sleep test/ice test
- b. Edrophonium chloride (Tensilon) test/Neostigmine test
- c. Fatigue on EMG

PROGRESSIVE EXTERNAL OPHTHALMOPLEGIA

- Progressive EOM weakness. Not acutely variable. Little C/O diplopia.
- Esp. ptosis and vertical muscles
- Associated with pigment retinopathy and beware heart block
- Usually maternal mitochondrial inheritance.

PRESCRIBING ASTIGMATIC LENSES TO CHILDREN

PRINCIPALS

- A. If no strabismus or asthenopia -
Patient should leave your office accommodating the same amount as they walked in.
 - 1. Do cycloplegic refraction -
 - 2. Give each eye full cylinder -
 - 3. Make dominant eye a cross-cylinder, i.e., zero spherical equivalent power.
Rx fellow eye to maintain anisometric difference

- B. If Accommodative ET:
 - 1. Do cycloplegic refraction
 - 2. Give each eye full cylinder
 - 3. Give full plus to each eye maintaining anisometric difference

- C. If XT and desire over minus:
 - 1. Do cycloplegic refraction
 - 2. Give each eye full cylinder
 - 3. Take 2D off sphere of each eye

EXAMPLE #1

Cycloplegic Refraction

O.D. +6.00 -6.00 X 180

O.S. +5.00 -2.00 X 180

Spherical Equivalent*

+ 3.00 S

+ 4.00 S

* The Spherical Equivalent is the amount the patient was accommodating (if it is a positive number) before obtaining glasses. The amount of accommodation is determined by the dominant eye and then both eyes accommodate equally.

a) For patients **Without Strabismus:**

1. If OD Dominant Then Rx:

OD + 3.00 -6.00 X 180

OS + 2.00 - 2.00 X 180

This is full cycloplegic refraction minus the 3D (spherical equivalent OD) that this patient was accommodating before glasses

2. If OS Dominant Then Rx:

OD + 2.00 -6.00 X 180

OS + 1.00 -2.00 X 180

This is full cycloplegic refraction minus the 4D (spherical equivalent OS) that the patient was accommodating before glasses

b) For patients with **Accommodative Esotropia:**

Rx = Full Cycloplegic Refraction

OD + 6.00 -6.00 X 180

OS + 5.00 -2.00 X 180

With these glasses, patient does not accommodate (whether OD or OS dominant)

c) For patients with **Exotropia** if want to overminus by 2 diopters more than patient was accommodating without glasses

1. OD Dominant Rx**

OD + 1.00 -6.00 X 180

OS plano -2.00 X 180

Full cycloplegic refraction minus 3D spherical equivalent OD minus 2D more

2. OS Dominant Rx**

OD plano -6.00 X 180

OS -1.00 0 -2.00 X 180

Full cycloplegic refraction minus 4D spherical equivalent OS minus 2D more

(**Note: This amount of accommodation may not be tolerated)

EXAMPLE #2

Cycloplegic Refraction (Fc)

O.D. +1.00 -4.00 X 180

O.S. +3.00 -4.00 X 180

Spherical Equivalent

+ 1.00 S

+ 1.00 S

a) For patients **Without Strabismus:**

1. If OD Dominant Then Rx:

OD + 1.00 -4.00 X 180

OS + 3.00 - 4.00 X 180

Rx = Fc as patient OD dominant was not accommodating before glasses

2. If OS Dominant Then Rx:

OD + plano -4.00 X 180

OS + 2.00 -4.00 X 180

This is full cycloplegic refraction minus the 1D (spherical equivalent OS). This patient was accommodating before glasses

b) For patients with **Accommodative Esotropia:**

Rx = Full Cycloplegic Refraction

OD + 1.00 -4.00 X 180

OS + 3.00 -4.00 X 180

With these glasses, patient does not accommodate (whether OD or OS dominant)

c) For patients with **Exotropia** if want to overminus by 2 diopters more than patient was accommodating without glasses

1. If OD Dominant Rx

OD - 1.00 -4.00 X 180

OS +1.00 -4.00 X 180

Full cycloplegic refraction minus 2D as patient was not accommodating without glasses

2. If OS dominant then Rx

OD - 2.00 -4.00 X 180

OS plano -4.00 X 180

This is full cycloplegic refraction minus the 1D (spherical equivalent OS).

The patient was accommodating without glasses minus 2D more. We wish patient to accommodate.