Handout for Demystifying Pediatric and Adult Strabismus

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1. **Basic Terms:**
   - Ipsilateral – Same side
   - Contralateral – Opposite side
   - Antagonist – Action opposes that of the contracting muscle that moves the eye.
   - Yoke – One of two muscles that move the eyes in the same direction

2. **Basic Ocular Movements:**
   - Ductions – The movement of one eye
   - Versions – (See below)
   - Vergences – Convergence, Divergence
   - Supra nuclear movements – Saccades and smooth pursuit

3. **Ductions refer to the movement of the one eye, and consist of:**
   - Abduction – Movement away from the nose
   - Adduction – Movement toward the nose
   - Elevation – Movement upward
   - Depression – Movement downward

4. **Muscles Responsible For Ocular Ductions (OD):**

5. **Versions** - Binocular movements, in which the two eyes move synchronously and symmetrically in the **same** direction (Yokes):

6. **Basic Principles:**
   - *Hering’s Law* – A bilateral phenomenon. Equal innervation to synergistic yokes: Innervation to one extra-ocular muscle (EOM) to contract generates equal innervation to contract its yoke muscle.
   - *Sherrington’s Law* – A unilateral phenomenon. As one EOM receives an impulse to contract, its ipsilateral antagonist receives an impulse to relax

7. **Functions of the EOMs: 1º, 2º, 3º**
   - LR Abduction
   - MR Adduction
   - IO Excyclotorsion, elevation, abduction
   - SO Incyclotorsion, depression, abduction
   - SR Elevation, incyclotorsion, adduct
   - IR Depression, excyclotorsion, adduction

8. **Innervation of the EOMs:**
   - CN III SR, IR, IO, MR, Lids, Pupils
   - CN IV SO
   - CN VI LR
   - Remember: LR

9. **Strabismus – Definition:**
   - Eye misalignment or eyes that do not move normally, caused by extra-ocular muscle imbalance.
   - One fovea is not directed at the same object as the other.
   - Only 30% of neonates are orthophoric!
10. **Basic Types of Strabismus:**

- **Ortho** - Greek for “straight” or “correct”
- **Eso** - IN
- **Exo** - Out
- **Hyper** - Up
- **Hypo** - Down
- **Cyclo-rotary** - Rotation around the Y axis of Fick (see diagram below)

11. **Classified According to Fusion Status:**

- **Phoria** - A “latent” tendency of the eyes to deviate; controlled by fusion
- **Tropia** - Greek, “turn in,” a manifest deviation exceeding fusional control
- **Intermittent** - Fusional control sometimes present

12. **Classified According to Fixation:**

- **Alternating** - spontaneous alternation of fixation
- **Monocular** - Constant, definitive preference for fixation with one eye.

13. **Classified According to Age of Onset:**

- **Congenital** - Documented early in infancy (<1 year old), term ‘infantile’ more appropriate.
- **Acquired** - Later Onset, after a period of apparently normal visual development

14. **Classified According to Type of Deviation:**

- **Horizontal** - Eso or Exo deviation
- **Vertical** - Hyper or Hypo deviation
- **Torsional** - Incyclo or excyclo-deviation
- **Combined** - Horizontal, vertical, and/or torsional deviation

15. **Overaction and Underaction:**

- Overaction, LIO
- Underaction, LIO

16. **Symptoms of Strabismus:**

- Diplopia in Adults - (children suppress)
  - Monocular (resolves w/ pinhole, likely refractive)
  - Binocular (Implies strabismus)
- Blurred Vision
- Headache - PM onset

17. **Assessment - Tools:**

- W4D and stereoacuity
- Single Maddox Rod Occluder
- Loose or Bar prism
- Muscle light

18. **Assessment - Techniques for Measuring Strabismus:**
Cover / Uncover – Fusion preserved; distinguish between phoria and tropia

Simultaneous Prism and Cover Test (SPCT) – Measures a manifest deviation without disrupting fusion

Alternate Prism and Cover test (APCT) – Measures a total deviation by disrupting fusion

Corneal Light Reflex Tests – Hirschberg, Krimsky, Bruckner

The Hirschberg Test:  The Krimsky Test:

19. **Quickly, Prism optics:**

   A prism bends light and real images toward it’s base

   Virtual images (patient’s perspective) are bent toward the apex

   Displacement in degrees, is approximately 50% of the prism’s power

20. **So Then, Measuring Strabismus:**

   Observation of face / head position

   Identify the deviation → cover and cover-uncover

   Measure the deviation:

   Orient prism apex toward deviation, like an arrow

   Begin APCT, and increase until reversal or no shift in either eye

   At both distance(6M) and near(1/3M) fixation

21. **Measuring Distance Deviation in 9 diagnostic positions:**

   Determines incomitance

   Measures degree of paresis in CN palsies

   Establishing what muscles are involved in paralysis

   Example:

   Assessing Ocular Versions: Very subjective:

22. **Taking a History:**

   *Begin w/ Family History*

   Distant relatives (uncles, aunts, cousins), g-parents, parents, siblings

   Ask about strabismus, glasses

   Be careful with “lazy eye”

   Any strabismus surgery? Results may be predictive
**Personal History**

Pregnancy and birth events

Prematurity, birth weight, labor complications

Hypoxia

Trauma, instruments used

Age of strabismus onset

Which eye? (Have parent point)

Always the same eye?

Constant or Intermittent?

Worse with fatigue, illness?

Worse Distance or Near?

Worse while focusing or daydreaming?

Any unusual ocular behaviors?

Squinting (bilateral = refractive error, unilateral = strabismus)

Onset related to convulsions, disease, immunizations, trauma?

**Developmental History**

Age appropriate milestones, mental and physical development (motor, speech, hearing, etc.)

**Any treatment for the strabismus?**

Glasses, patching, exercises, surgery

**Any diplopia in adults or where acute onset?**

Constant, intermittent, binocular, monocular, distance, near?

(Rule out blurred vision and "ghosting" vs distinct double)

Visible in photos, to strangers, to other relatives?

Do not dismiss an Intermittent Hx that does not present at the first exam!

Observe the patient during the history for signs of strabismus, preferred eye fixation, anomalous head posture (tilt or turn)

23. **Common Forms of Strabismus:**

**Esotropia - Types**

Congenital:

Onset of ET within the first year of life

Often requires several surgeries

Accommodative:
Hyperopes able to clear the blurred distance vision with accommodation, causes excessive convergence.

Usually resolves with full hyperopic correction from CR.

**Partially Accommodative:**

Residual ET after Full cycloplegic spectacle Rx

If ortho @ dist and 'ET @ near, treat with Bifocal

If ET distance AND near → surgery

**Sensory:**

Secondary to early-onset, unilateral blindness, cataract, aphakia

**Pseudo ET - The appearance of strabismus due to common childhood facial features:**

- Extra Fold of Skin at the inner eyelid (epicanthus)
- Wide, Flat Nose
- Eyes that are Close Together
- By SPCT or APCT, there is no eso-shift

A child will outgrow false strabismus as the face grows and changes.

<table>
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<tr>
<th>Pseudo ET</th>
<th>True ET</th>
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**Esotropia - Treatment**

Full hyperopic spectacle Rx in younger children

Bifocals if ortho @ dist and ET @ near

Patching for any amblyopia / fixation preference

Surgery if constant ET distance and near: BMRc

**Exotropia - Types**

**Basic:**

Intermittent most common, onset age 2-4 ½

**Convergence Insufficiency Type:**

Common across all age groups

Blurred VA, headache, aesthenopia

N>D by 15Δ, but may just be exophoria

Remote NPC

Treat w/ orthoptic exercises

**Congenital (Infantile):**

Rare - associated w/ organic disease, craniofacial abnormalities, and neurological problems
Consecutive XT

After surgical correction for ET - may be an overcorrection, or possible slipped or lost muscle

**Exotropia - Presentation**

Closing one eye in bright sunlight

Headaches

Only ¼ of the X(T)’s will improve or are stable

Adults c/o diplopia while driving

**Exotropia - Treatment**

Full myopic Rx

Base-in prism

Alternate occlusion as anti-suppression measure to avoid amblyopia

Surgery if tropia > 50% of the time. Delay until 2 ½-4 y/o to promote fusion: BLRc aiming for ET 10-20 ∆ early on.

24. **Cranial nerve palsies:**

Congenital - III (rare)

IV (common)

VI (less common)

Acquired -

Trauma (6th n. most common)

Post-Viral / post-inoculation

CNS disease / neoplasm

Idiopathic

25. **Third Nerve Palsy (Cranial Nerve III)**

Levator palpebrae superioris, pupillary sphincter & ciliary body, MR, IR, SR, IO muscles.

“Complete” 3rd nerve palsy “down and out” eye position secondary to unopposed LR & SO muscles

Usually sudden-onset diplopia - horizontal & vertical components

Ptosis can act as an occluder, eliminating diplopia!

May be painful

Common etiologies: Vascular (DM, and HBP) and ICA aneurysm due to proximity of 3rd nerve to ICA.

If pupils involved → MRI indicated

Resolution in 3-6 months w/ pupil-sparing and underlying cardiovascular Hx.

Fresnel prisms may not be helpful due to complexity of the strabismus: horizontal, vertical, torsional

26. **Fourth Nerve Palsy (Cranial Nerve IV)**
Most common cause of vertical diplopia

CN IV longest intra-cranial course, susceptible to trauma
Innervates only SO muscle, so paresis results in unopposed antagonist (IO) and hyper deviation
Pts present with head tilt (torticollis), maybe hypertropia

“Fallen Eye Syndrome” Fixing with the paretic eye induces contralateral hypo deviation and pseudo-ptosis

Measure deviation in all 9 diagnostic fields, if possible

Three-Step Test:
1. Identify Hyper eye, R or L
2. Worse in L or R gaze?
3. Worse in L or R tilt (Bielschowsky head tilt test)?

Example: R hyper > L gaze > R tilt

Presence of Subjective excyclotorsion:
Measure with Double Maddox rod (DMR) lenses in trial frames and muscle light at near

Presence of Objective excyclotorsion:
Affected eye fundus will appear excyclotorted compared to contralateral eye – based on position of O.N. relative to the horizontal midline.
Will always be present if true 4th nerve palsy, even in the absence of subjective torsion

Congenital Etiology:
Long-standing tilt
Rare amblyopia
Facial asymmetry, affected side fuller.

Acquired Etiology:
Trauma, CVA, sinusitis, tumor, ischemia in older pts.

Bilateral Fourth Nerve Palsy:
Uncommon – Usually due to trauma. Hyper may reverse in different gaze positions

27. Sixth Nerve Palsy (Cranial Nerve VI):
Most commonly affected CN after trauma secondary to unique intra-cranial course through Dorello’s canal

Binocular horizontal diplopia D>N
Clinically, abduction deficit past the midline
Adults: Vascular infarct secondary to DM, HBP, demyelinating disease, trauma, or iatrogenic – improves
- No CT / MRI

Children: Viral syndrome, trauma, occasional tumor, increased ICP –
Congenital, idiopathic – Improves, but do CT / MRI

Prognosis for full recovery good in adults w/ vascular disease – allow 3-6 months→ Interim patch or Fresnel

28. **Duane’s Syndrome:**

Co-contraction of MR and LR secondary to a congenital miswiring of the VIth nerve splitting to innervate both the LR and MR

Usually unilateral, rarely bilateral

Historically three types:

- Type I: Poor ABduction
- Type II: Poor ADduction
- Type III: Poor ABduction AND Adduction

Compensatory face turn common – *usually toward the affected eye*

Co-contraction causes globe retraction and characteristic palpebral fissure narrowing with attempted ADduction

Palpebral fissure widening with attempted ABduction secondary to LR dysfunction / paresis and inhibition of the MR tone by Sherrington’s

Anisometric amblyopia common

Strabismus surgery indicated to correct the face turn

29. **Brown’s Syndrome:**

Little or no elevation in ADduction, most always unilateral (Bilateral in 1/10)

Caused by a short / abnormal SO tendon sheath

No overaction of antagonist!

V-pattern in upgaze, ortho in 1°

1° position Hypo

Anomalous head posture: chin elevation and / or face-turn
Restricted forced ductions

Acquired Cases:

- Blunt local trauma
- Inflammatory – Grave’s, lupus, RA, Sjogren’s
- Iatrogenic (after certain types of strabismus surgery, scleral buckling procedures)

Treatment:
Observe for possible spontaneous improvement
Surgery indicated for cosmetically unacceptable head posture
High incidence post-op complications

30. **Dissociated Vertical Deviation (DVD):**
Elevation, abduction and excyclotorsion of a non-fixing eye with the cover-uncover test without apparent movement of the fixing eye

Appears to disregard Hering’s Law. Recent work using electronic analysis of ocular movements suggests that this is not the case.
Usually bilateral and asymmetric, spontaneous or w/ occlusion

Strong association w/ early-onset strabismus and latent nystagmus

31. **Blowout Orbital Fracture**
Orbital floor fracture secondary to blunt trauma to the inferior orbital rim

Results in a downward herniation of the orbital fat and the IR muscle into the maxillary sinus with entrapment of the inferior rectus muscle, inferior oblique muscle, or surrounding tissue, in the fracture trap door.

Hypotropia in the primary position that increases w/upgaze and possibly decreases or becomes a hypertropia in downgaze

Symptoms: Diplopia (Occasionally painful) in vertical gaze immediately following the trauma, hypesthesia

Clinical signs: Ecchymosis, enophthalmos, limited elevation and depression positive forced ductions

Treatment: Repair the fracture first then eventually repair the strabismus by releasing the restricted extraocular muscle(s)

32. **Internuclear Ophthalmoplegia (INO):**
Ipsilateral adduction deficit with a contralateral abducting nystagmus

Normal convergence

Attributable to a brainstem lesion associated w/ demyelinating disease, typically MS.

An abnormality of the inter-neuron pathway connecting the 6th nerve nucleus with the third nerve nucleus – responsible for conjugate (coordinated) horizontal gaze

Neurons of the 6th nerve nucleus “yoke” the LR movements with the contralateral MR

In young adults – Etiology is demyelinating disease (MS)

In older adults – Vascular infarct
33. **Treatment of Strabismus:**

- Optical / correction of refractive error
- Press-on prisms (Fresnel)
- Ground-in spectacle prism Rx
- Surgical
  - Weakening → Recessions
  - Strengthening → Resections
  - Transpositions

34. **Post-Operative Care:**

Close follow-up is important

Complications

- Induced refractive error changes
- Diplopia
- Perforation of the sclera w/ surgical needle
- Anterior segment ischemia
- Lost and slipped muscles
- Changes in eyelid position
- Postoperative infections
- Foreign body granuloma and allergic reaction
- Conjunctival scarring and cysts
- Adherence Syndrome
- Nausea and vomiting
- Oculocardiac reflex
- Malignant hyperthermia

**References / Further Study:**


Helpful Internet Sites:

University of Illinois: The Online Eye Manual
http://mail.ml.usoms.poznan.pl/eyemanual/emstrabismus.htm

The Binocular Vision Tutor
http://www.city.ac.uk/optics/BVTutor/home.html

Thamburaj.com
www.thamburaj.com/ocular_movements.htm