OKAP REVIEW OF PEDIATRIC OPHTHALMOLOGY AND STRABISMUS

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DEVELOPMENT OF VISION
THE SENSORY SYSTEM
AMBYOPIA
ANATOMY OF THE EXTRAOCULAR MUSCLES

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OUTLINE

1. Sensory physiology and pathology
   
   A. Development of normal binocular vision
      1) Normal retinal correspondance
      2) Vieth-Muller circle
      3) Empirical horopter
      4) Fusion
      5) Stereopsis
   
   B. Pathology of sensory function
   
   C. Abnormal Retinal Correspondance
   
   D. Diplopia
      1) Physiologic diplopia
      2) Confusion
   
   E. Supression
   
   F. Monofixation syndrome
   
   G. Tests of sensory anomalies
      1) Worth four dot
      2) Bagolini test
      3) Afterimage test
      4) Synoptophore
      5) Amblyoscope
      6) Titmus test

2. Development of the visual system
   
   A. Amblyopia
   
   B. Assessment of amblyopia
   
   C. Treatment of amblyopia
   
   D. Eccentric fixation

3. Anatomy of the extraocular muscles
   
   A. Origin
   
   B. Course
   
   C. Insertion
   
   D. Action
   
   E. Vascular supply
   
   F. Orbital and facial relationships
      1) Lockwoods ligament

4. Summary
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OKAP REVIEW 2008
Pediatric Ophthalmology and Strabismus
I. Development of the visual system

OKAP REVIEW 2008
Pediatric Ophthalmology and Strabismus
II. Sensory anomalies

III. Amblyopia

OKAP REVIEW 2008
Pediatric Ophthalmology and Strabismus
IV. Anatomy of the EOM's
Summary, review and quiz

Sensory system
COLOR

Sensory system
FORM

Sensory system
LOCATION visual space

Sensory system + Motor system
= Sensorimotor system

visual sensations precipitate a chain of motor responses that move the eyes
Visual development
Vision requires:
1) intact optical system
2) photo-pigment-mediated transformation of light into wave action potentials

Visual development
Vision requires:
3) synapses with feedback
4) precise binocular mapping of the environment onto the retina, lateral geniculate body and occipital cortex

Neurophysiology
Anterior visual system
> stimulus received by retinal photoreceptors
> optic nerve
> optic tract
> optic chiasm

Neurophysiology
Lateral Geniculate Body
> LGN or LGB
> thalamus

Neurophysiology
Lateral Geniculate Body
> receives afferent fibers from the anterior visual pathway
> relays information to primary visual cortex
> mechanism unknown

Neurophysiology
Lateral Geniculate Body
> organized in 6 layers
> 6 - outermost
crossed, contralateral - 1, 4 6
uncrossed, ipsilateral - 2, 3, 5
Pcells
> color, two point discrimination

Lateral Geniculate Body
> important clinically
> must know for BOARDS:
2 cell types:
  • Magnocellular neurons
  • Parvocellular neurons

Vision
Neurophysiology - LGN
M CELLS
Magnocellular neurons
Parvo - small
WHERE
parafoveal, peripheral
WHAT

Lateral Geniculate Body
> important clinically
> must know for BOARDS:
2 cell types:
  • Magnocellular neurons
  • Parvocellular neurons
Neurophysiology
Occipital lobe

- also called:
  striate cortex
  Brodmann’s area 17

Development of normal binocular vision

- must understand concepts of
  - visual space
  - visual direction

Visual space

- Objective
  - objects in physical space outside of and independent of our visual system
- Subjective
  - conscious awareness of objects and perception by our brain

Development of normal binocular vision

- stimulation of any retinal area results in visual sensation from a subjective visual direction

Retinal correspondence

- retinal areas in the two eyes share a common subjective visual direction

Visual direction

- normally
  - fovea = visual axis = straight ahead

Retinal correspondence

- corresponding retinal points are located on the same meridian and at the same distance from the fovea in each eye
Empirical horopter
- Clinically defined area (3D space) where all points are seen singly

Vieth-Müller circle
- Model based upon assumption that eye is a perfect sphere

Panum’s area
- Area around horopter where non-corresponding retinal points are stimulated without diplopia
  - Based upon ability to fuse slightly disparate images
  - Stereopsis exists
  - Single binocular vision

Stereopsis
- Relative localization of visual objects in depth
  - Limited by distance (<20 feet)
  - Remember: monocular clues - important in interpretation of depth

Titmus stereo test
- Allows stereopsis by presenting disparate images
  - Fly = 4000 sec of arc
  - 9/9 circles = 40 sec

Random-dot stereogram
- No monocular clues

Sensory adaptations to strabismus
- Suppression
- ARC (Anomalous Retinal Correspondence)
- Diplopia
- Confusion
Diplopia

Confusion
- rare
- perception of 2 images superimposed

Suppression
- strabismus
- confusing images originating from the retina
- central inhibition to avoid diplopia
- typically seen in children
- clinically, can give clues about etiology and age of onset of strabismus
- dense amblyopia

ARC fusion
- simultaneous perception of similar images - stimulate retinal points which normally do not correspond

Sensory testing
Why bother?
- can give insight into etiology of strabismus
- can help with surgical plan
- helps to prepare the patient post-op
  - i.e. diplopia

Tests for retinal correspondence
- Based upon principle that images that are less "alike" are harder to fuse
- Tests that make images less alike (red filter) are more dissociating and therefore reveal suppression easier
Tests for retinal correspondence

- Bagolini lenses
- Synoptophore
- Red Filter
- Worth four-dot
- Afterimage test

Tests for Retinal Correspondence

- Bagolini lenses
  - Striated lenses produce streaks before the R and L eye at 45 and 135 degrees

Tests for Retinal Correspondence

- Synoptophore
  - major amblyoscope
Tests for Retinal Correspondence

Worth four-dot

- Test for fusion
- Very dissociating

WORTH FOUR DOT

NRC - fusion eyes aligned
ARC - fusion eyes deviated

WORTH FOUR DOT

FUSION

SUPPRESSION

OD

OS

Patient’s view

Patient’s view
Tests for Retinal Correspondence

Afterimage test
- must have central fixation
- each eye tested separately
- streak of light
  - horizontal image on fixing eye
  - vertical image on deviated eye
- result independent of eye position

Afterimage test
- central cross represents direction of fovea
- results independent of alignment

Right 6th nerve palsy
- Patient’s view

WORTH FOUR DOT
- Patient’s view

Medial rectus trauma
Clinical example

5 year old - BMR age 2 for esotropia

- "holds" in small angle (10 PD) ET
- ACT - builds to ET 20
- what may sensory testing reveal?
- would you operate?

Bagolini lenses

ARC - peripheral fusion

Bagolini lenses

OS

OD

Suppression OD

SENSORY TESTING

DOUBLE MADDOX ROD

- Test for torsion
- Streaks are placed vertically and perceived horizontally

Fixation

Eccentric Fixation
**ANGLE KAPPA**

Angle between "line of sight" and the corneal/pupillary axis

- **Positive**: + patient looks \( \times \text{T} \)
- **Negative**: – patient looks \( \text{E} \text{ T} \)

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**Eccentric fixation**

- fixation is not at the fovea

**Angle kappa**

- fixation is at the fovea
- must use ophthalmoscope

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**Amblyopia**

- MOST COMMON CAUSE OF UNILATERAL POOR VISION IN CHILDREN
- Prevalence 2-4%
- PREVENTABLE!!!

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**Amblyopia**

Amblyos - (Greek) “dullness of vision”
opacity - from ops (Greek) vision

- Has come to refer to decreased vision in the setting of a "normal exam"
- Accepted definition: > 2 lines difference in acuity between the eyes
Hubel and Weisel
1970’s

- Nobel Prize winners
- identified “sensitive period” for development of normal binocular vision
- Discovered that suturing lids of kittens resulted in atrophy of cell bodies in the LGN

Amblyopia - pathogenesis

- Lateral Geniculate Body
  - Clinical relevance - amblyopia
    - Amblyopia
      - M and P cell mal-development
      - severe sensory deprivation causes reduced cell size

Amblyopia - neurophysiology

- Lateral geniculate

- abnormal early visual experience
- profound effect on neural function
- occipital cortex
- lateral geniculate
- receptive fields of neurons become large
- monocular and binocular cells affected

Amblyopia-Functional

- reversible
- strabismic
- anisometropic
- occlusion
- primarily defect of central vision

Functional Amblyopia

- Child with high myopia
  - right eye
  - Normal left eye
- Secondary exotropia
- Must always correct refractive error and improve vision before considering strabismus surgery

Lateral geniculate
**Amblyopia - Organic**
- typically refers to ocular anomalies preventing optimal acuity
- abnormality may be subtle or undetectable
- “irreversible”
- may be diagnosed after failure to respond to occlusion therapy
- must remember that organic amblyopia may have superimposed functional amblyopia

**Occlusion Amblyopia**
- Organic cause
- Can result from patching

**Bilateral Amblyopia**

**Amblyopia - diagnosis**
- Pre-verbal
  - fixation preference
  - vertical prism test (8-10 PD)
  - acuity
    - OKN, FPL, Teller, VEP
- Verbal
  - Allen pictures, numbers, letters
  - > 2 lines difference

**Amblyopia - Strabismus**

**Fixation**
- In classic “textbook” congenital esotropia infants alternate their fixation and do not become amblyopic
Amblyopia
Crowding phenomenon

ACUITY
ISOLATED > LINEAR
in amblyopes

Amblyopia - Treatment
- correct refractive errors
- treat ocular problems - i.e. cataract, ptosis
- occlusion
- penalization (i.e. atropine)
- follow closely
- occlusion amblyopia - always check Va in "better" eye

Essentials of treatment
- Patient and family understanding and involvement
- Motivation/rewards
- Realistic goals
- Make it fun and easy
- Know when to stop

Essentials of treatment
- Must take treatment seriously
- Often make contract with older kids
- Capitalize on their interests
- Visual challenging is essential

Pediatric Eye Disease Investigator Group
- Amblyopia Treatment Study Group
- Looking at measurement of visual acuity
  - Efforts to standardize
  - Effects of treatment on child and family
- Comparison of
  - Drops and patching
  - Shorter vs. longer occlusion

Amblyopia Study Group Patching Regimens
- Archives of Ophthalmology 2003;121:603-611
  - 189 children<7 y w/ moderate amblyopia (20/40-20/80)
  - Randomized to 2h/d vs. 6h/d patching
  - Both groups performed > 1 h per day of near visual activities
  - Compliance consistent with other studies (poor)
  - 4 m follow up
  - Similar improvement in 2 groups

Anatomy of the EOM's
- Ocular alignment is determined by the extraocular muscles and their surrounding tissues
  - PRIMARY POSITION
    - the eye and head are directed straight ahead
    - medial walls are parallel
    - lateral walls are 45° from medial walls
    - in primary position: SO, IO - 51°
    - SR, IR - 23°
    - MR, LR - 90°
EOM’s Origin
Annulus of Zinn

- oval fibrous ring at the orbital apex
- EOMs originate at annulus

EXCEPT:
- inferior oblique
- superior oblique
- levator palpebrae superiors

Origin of the EOM

Inferior Oblique
- maxillary bone, adjacent to lacrimal fossa, posterior to orbital rim

Superior Oblique
- orbital apex above annulus (functional origin at trochlea of LPS)

LPS
- orbital apex above annulus

Insertions of the EOM’s
Spiral of Tillaux

- Rectus muscles insert into sclera anterior to the equator via tenons
- Their anatomic relationship at the insertions forms the Spiral of Tillaux
- Thinnest sclera ~ .3mm just posterior to the insertion

Insertions of the EOM’s Oblique muscles

- insert posterior to the equator
- SO longest tendon, courses inferior to SR
- IO shortest tendon, courses inferior to IR

Cardinal positions and yolk muscles

Cardinal positions and yolk muscles
EOM’S function
- Function dependent upon position of the globe
- 1st, 2nd, 3rd action in the primary position:
  - Medial, lateral recti - adduct, abduct
  - Sup, inf oblique’s - elevate, depress, intort, extort, abduct
  - Sup, inf recti - elevate, depress, intort extort, adduct

EOM’S function
- Clinical Correlation
  - A and V patterns due to oblique overaction
  - ET in thyroid patients with tight inferior recti

EOM - Innervation
- CN III LPS, SR upper
  - MR, IR, IO lower
- CN IV SO
- CN VI LR

Anatomy of the EOM’s Vascular supply
- Major: muscular branches of the ophthalmic artery
- Additional: Lacrimal artery to LR
  - Infraorbital to IO, IR

Vascular supply
- Most Often
  - Recti muscles - contain 2 anterior ciliary arteries
  - Exception - Lateral rectus contains 1

Vascular supply
- Clinical Relevance
  - Surgery on multiple recti muscles contraindicated due to risk of anterior segment ischemia

Summary NRC
- Horopter
  - Based upon NRC
    - Points stimulate corresponding retinal elements
    - Single vision and fusion exists
- Panum’s area
  - Slightly disparate points
  - Allows stereopsis
  - Outside - have physiologic diplopia

Normal Retinal Correspondence
- Empirical horopter
  - TL, NR, FL, FR, NL, TR
  - Cyclopean eye
### Summary

#### Retinal correspondence
- Retinal correspondence
- ARC  NRC
- Binocular

#### Amblyopia
- Major preventable cause of visual loss
- Strabismus, anisometropia, high hyperopia, and myopia at risk
- Maximum FT occlusion = 1 week/year of life
- "Critical period" for development of binocular vision
- Possibly "at risk" until 10 years of age

#### Anatomy
- Anterior segment circulation from 4 recti
  - All have 2 except LR has one ACA
- All EOM orig. from annulus except IO, SO, LPS
- Recti insert anterior to equator, obliques-posterior
- Inferior-extorters, Superior-intort
- Recti - adduct, Obliques - abduct
- Obliques course inferior to recti

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GOOD LUCK!!!!

QUIZ
1. Which of the following does not arise from the annulus of Zinn?
   1. superior oblique.  
      a. 1, 2, and 3. 
      b. 1 and 3  
   2. levator palpebrae.  
      c. 2 and 4  
      d. 4 only  
   3. inferior oblique.  
      e. 1, 2, 3, and 4  
   4. superior rectus.  

2. T or F -- The superior oblique tendon passes between the superior rectus muscle and the globe on the way to its insertion.  
   ANS = TRUE

3. Match each set of action from primary position listed in the left-hand column with the appropriate muscle in the right-hand column.
   a. intorsion, depression, abduction. =  
      1. Inferior rectus.  
   b. extorsion, elevation, abduction. =  
      2. Superior rectus.  
   c. depression, extorsion, adduction. =  
   d. elevation, intorsion, adduction. =  
      4. Inferior oblique.

4. T of F-- Physiologically, any point not lying on the empirical horopter will be perceived doubly by the human visual system.  
   ANS =

5. T or F-- If simultaneous stimulation of retinal areas in two eyes leads to the perception of one image, normal retinal correspondence is said to exist.  
   ANS = FALSE

6. T or F-- For fusion to exist, there must be simultaneous stimulation of corresponding retinal areas with normal retinal correspondence.  
   ANS =

7. T or F-- The most important visual clues for depth perception require binocular vision.  
   ANS =

8. T or F-- Diplopia occurs when the two foveas of a singe patient each contain a distinct retinal image.  
   ANS =

9. T or F-- If a patient with manifest strabismus does not complain of diplopia, then suppression must be active.  
   ANS =

10. Which of the following regarding amblyopia is/are true?
    1. The incidence in general population is approximately 2 to 3 %.
    2. The presence of an afferent pupillary defect clearly establishes an organic etiology for visual loss, rather than amblyopia.
    3. Patient with amblyopia will frequently perform better with single-symbol acuity test targets than with line targets ("crowded stimuli")
    4. A neutral density filter placed over an amblyopic eye will generally cause a greater decrement in visual acuity than the same filter placed over an eye with maculopathy.
    a. 1, 2, and 3  
    b. 1 and 3  
    c. 2 and 4  
    d. 4 only  
    e. 1, 2, 3, and 4.  
    ANS =

11. T or F-- The proper guideline for intervals between examinations for a child undergoing full-time occlusion therapy is 1 week for every month of age.  
    ANS =

12. T or F-- Testing with Bagolini striated glasses for retinal correspondence requires preparation with cover-uncover testing and assessment of fixation behavior.  
    ANS =

13. When tested with a Maddox rod held over the affected eye with its cylinders running horizontally, a patient with exyclotropia will perceive:
    a. a horizontal line.  
    b. a vertical line.  
    c. an oblique line running superotemporal to inferonasal.  
    d. an oblique line running superonasal to inferotemporal.  
    e. a curved line concave toward the nose.  
    ANS =
14. Broad nasal bridges with abnormally large angle kappa may lead to an error in the diagnosis of strabismus with which of the following methods?
   1. alternate-cover tests. a. 1, 2, and 3.
   2. Maddox rod testing. b. 1 and 3.
   3. Cover-uncover testing. c. 2 and 4
   4. Hirschberg testing. d. 4 only
   e. 1, 2, 3, and 4.

ANS = D

15. T or F—Negative angle kappa simulates esotropia

ANS -

16. Compared to magnocellular cells, parvocellular neurons are more sensitive to
   a. Low-medium spatial frequencies
   b. Fine two-point discrimination
   c. Direction, motion, and speed
   d. Flicker stereopsis

ANS =

17. The vertical prism or induced tropia fixation test is useful
   a. To measure cyclovertical deviations
   b. To detect amblyopia in preverbal children without strabismus
   c. To assess binocular cooperation
   d. To measure vertical fusional vergences

ANS =

18. A 34-year-old man sustained closed head trauma and now complains of objects appearing tilted. The degree of tilting can be quantified by which of the following tests?
   a. Simultaneous prism-cover test
   b. Double Maddox rod test
   c. Careful analysis of ductions and versions together
   d. Lateroverision reflex test

ANS =

19. Paradoxic diplopia observed after strabismus surgery in a formerly esotropic patient is most likely caused by which one of the following?
   a. Surgical undercorrection
   b. Eccentric fixation
   c. Surgical overcorrection
   d. Persistence of abnormal retinal correspondence

ANS =

20. Which of the following cannot be used to test for ARC?
   a. Worth four-dot test
   b. Major amblyoscope test
   c. Titmus stereo test
   d. Cuppers monocular afterimage test

ANS =

21. A patient presents with a left superior oblique muscle paresis and the following measurements. Which of the operations listed is the most appropriate?

Right head tilt: LHT = 5

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Left head tilt: LHT = 15

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<th></th>
<th>Right gaze</th>
<th>Left gaze</th>
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</thead>
<tbody>
<tr>
<td></td>
<td>a. Tuck LSO</td>
<td>b. Tuck LSO, recess LIO</td>
</tr>
<tr>
<td></td>
<td>c. Recess LIO</td>
<td>d. Recess RSR and LIR</td>
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ANS =