

OKAP REVIEW OF PEDIATRIC OPHTHALMOLOGY AND STRABISMUS

April 1, 2008

DEVELOPMENT OF VISION

THE SENSORY SYSTEM

AMBLYOPIA

ANATOMY OF THE EXTRAOCULAR MUSCLES

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OKAP REVIEW OF PEDIATRIC OPHTHALMOLOGY AND STRABISMUS

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 ablyopia
- 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

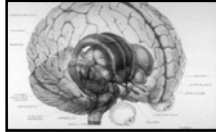
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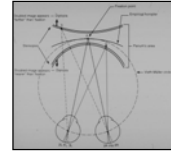
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I. Development of the visual system



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II. Sensory anomalies



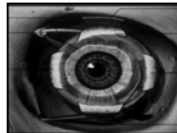
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III. Amblyopia



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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

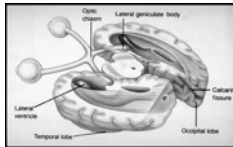
Neurophysiology Anterior visual system

- > stimulus received by retinal photoreceptors
- > optic nerve
- > optic tract
- > optic chiasm



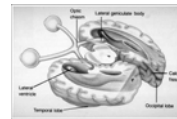
Neurophysiology Lateral Geniculate Body

- > LGN or LGB
- > thalamus



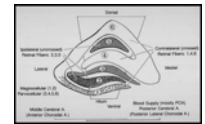
Neurophysiology Lateral Geniculate Body

- > 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
 - 1 - innermost
- uncrossed, ipsilateral - 2, 3, 5
- crossed, contralateral - 1, 4, 6



Lateral Geniculate Body

- > important clinically
- > must know for BOARDS:

2 cell types:

- Magnacellular neurons
- Parvocellular neurons

Vision Neurophysiology - LGN

M CELLS

Magno-large

WHERE

parafoveal, peripheral

Neurophysiology - LGN

P cells

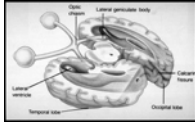
Parvo - small

WHAT

color, two point discrimination

Neurophysiology Occipital lobe

- > also called:
striate cortex
Brodman'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

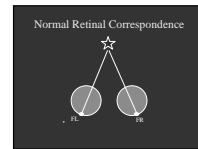
- > stimulation of any retinal area results in visual sensation from a subjective visual direction

Retinal correspondence

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

Visual direction

- > normally
fovea = visual axis = straight ahead

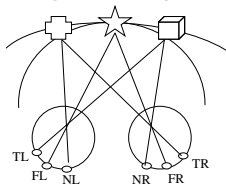


Retinal correspondence

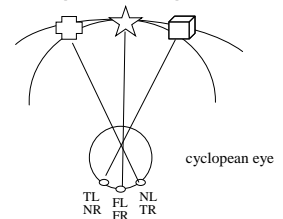
NRC

- > corresponding retinal points are located on the same meridian and at the same distance from the fovea in each eye

Normal Retinal Correspondence Empirical Horopter



Normal Retinal Correspondence Empirical Horopter



Empirical horopter

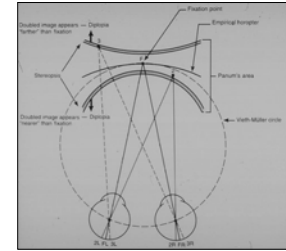
- > clinically defined area (3D space) where all points are seen singly

Vieth-Muller circle

- > model based upon assumption that eye is a perfect sphere

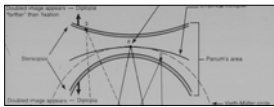
Empirical horopter

- > fusion exists
- > all points are seen singly
- > requires NRC by definition
- > NO stereopsis



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



Stereopsis

Titmus stereo test

- > allows stereopsis by presenting disparate images
- > fly = 4000 sec of arc
- > 9/9 circles = 40 sec

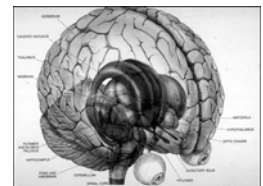


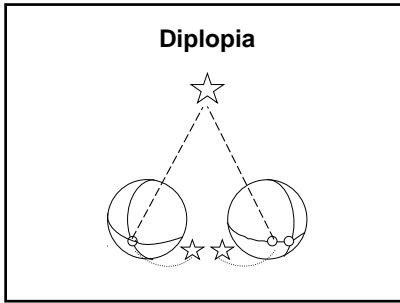
Random-dot stereogram

- > no monocular clues

Sensory adaptations to strabismus

- > suppression
- > ARC
- > diplopia
- > confusion

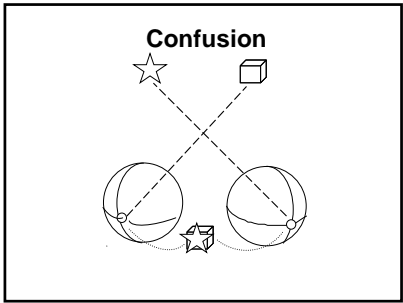




Sensory adaptations to strabismus

confusion

- rare
- perception of 2 images superimposed

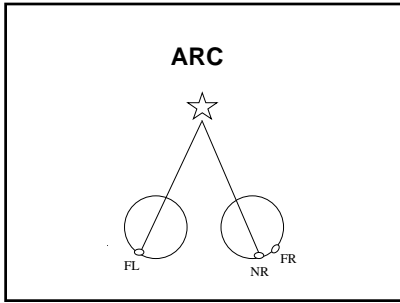


Suppression

- > 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

Sensory adaptations to strabismus

- > 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

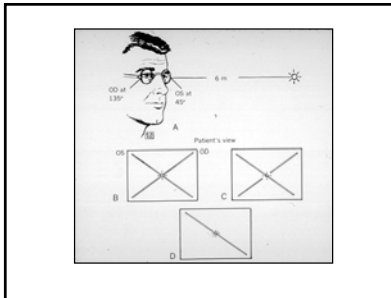
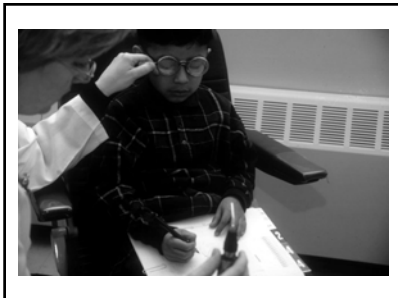
**Tests for retinal
correspondence**

- DISASSOCIATION ↓
- Bagolini
 - Synoptophore
 - Red Filter
 - Worth four-dot
 - Afterimage test

**Tests for Retinal
Correspondence**

bagolini lenses

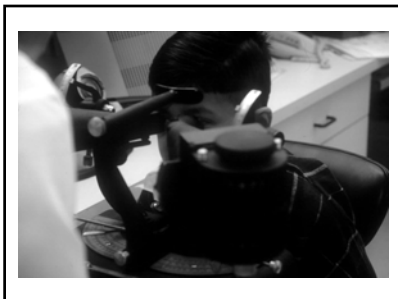
- Striated lenses produce streaks before the R and L eye at 45 and 135 degrees



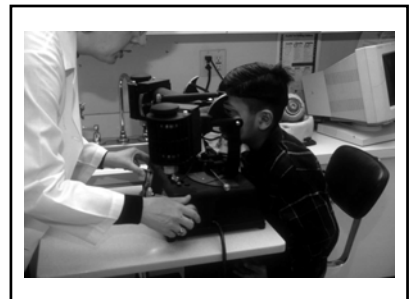
**Tests for Retinal
Correspondence**

Synoptophore

- major amblyoscope



synoptophore

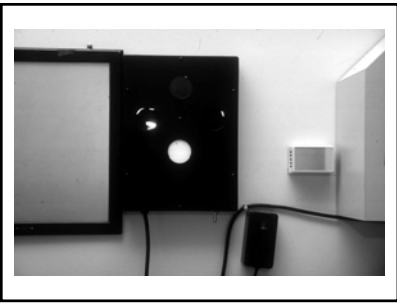
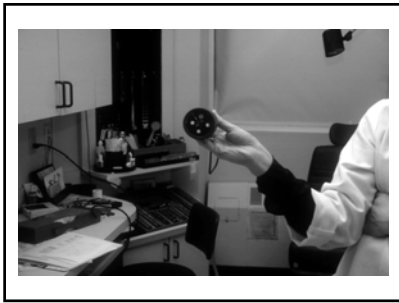




Tests for Retinal Correspondence

Worth four-dot

- > Test for fusion
- > Very dissociating



WORTH FOUR DOT
Patient's view

FUSION

WORTH FOUR DOT

NRC - fusion eyes aligned

ARC - fusion eyes deviated

WORTH FOUR DOT
Patient's view

SUPPRESSION

OD

WORTH FOUR DOT
Patient's view

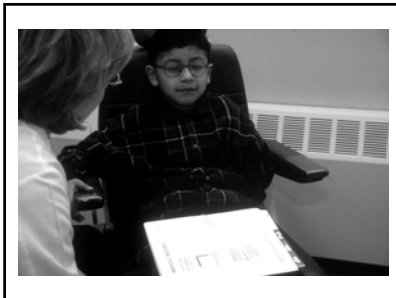
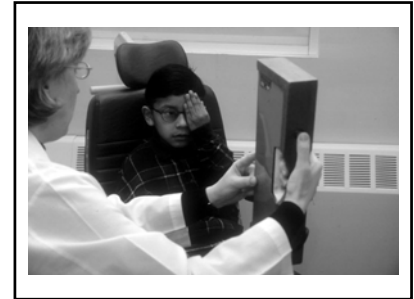
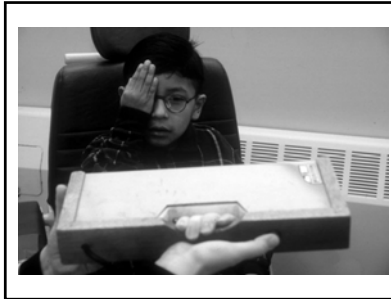
SUPPRESSION

OS

Tests for Retinal Correspondence

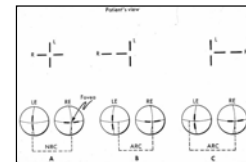
Afterimage test

- 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

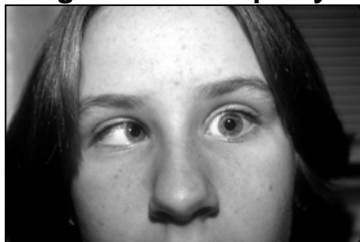


Afterimage test

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

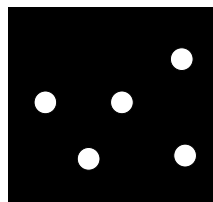


Right 6th nerve palsy



WORTH FOUR DOT Patient's view

- ESOTROPIA
- UNCROSSED
DIPLOPIA
- HOMONYMOUS



Medial rectus trauma



WORTH FOUR DOT
Patient's view

EXOTROPIA
CROSSED
DIPLOPIA
HETERONYMOUS

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

OS OD

Bagolini lenses

OS Suppression OD

WORTH FOUR DOT
Patient's view

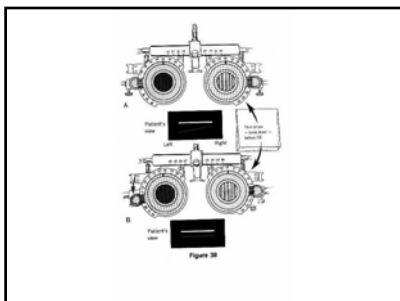
ARC
FUSION

Peripheral Fusion

SENSORY TESTING

DOUBLE MADDOX ROD

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



➢ Fixation

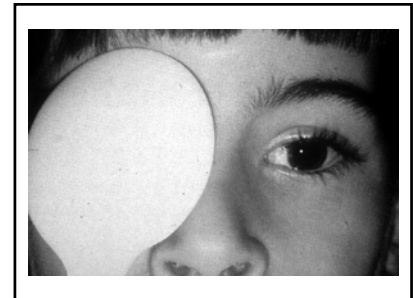
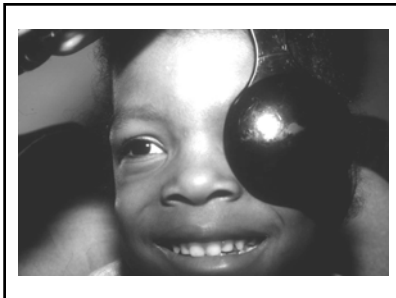
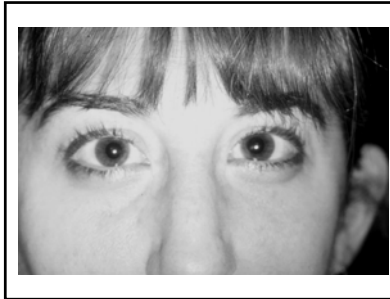
Eccentric Fixation

ANGLE KAPPA

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

> Positive + patient looks XT

> Negative - patient looks ET



Eccentric fixation

- fixation is not at the fovea

Angle kappa

- fixation is at the fovea

- must use ophthalmoscope

Amblyopia

> MOST COMMON CAUSE OF UNILATERAL POOR VISION IN CHILDREN

> Prevalence 2-4%

> PREVENTABLE!!!!



Amblyopia

Amblyos - (Greek) "dullness of vision"
opia - 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

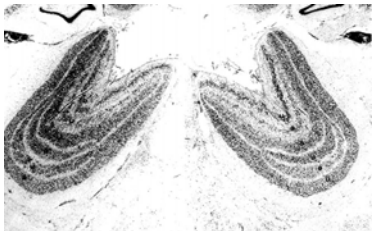
Amblyopia - pathogenesis



Lateral Geniculate Body Clinical relevance - amblyopia

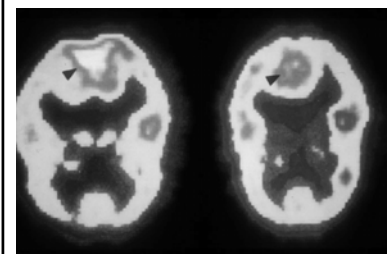
- > Amblyopia
 - M and P cell mal-development
 - severe sensory deprivation causes reduced cell size

Lateral geniculate



Amblyopia neurophysiology

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

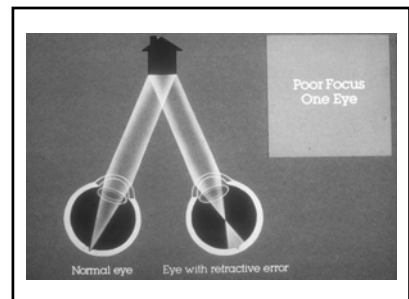


Amblyopia-Functional

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

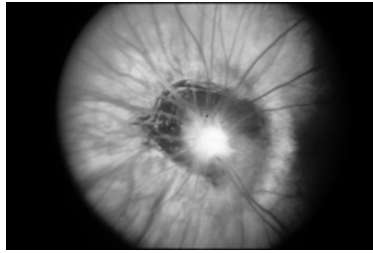
Functional Amblyopia

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



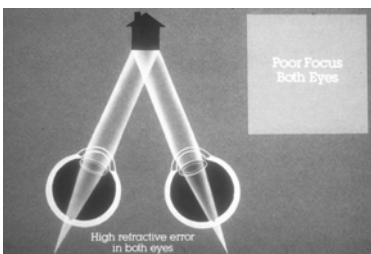
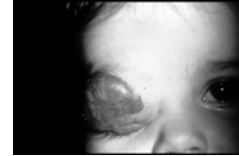
Amblyopia - Organic

- > 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



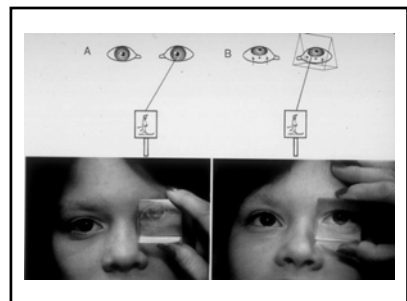
Amblyopia - diagnosis

- 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

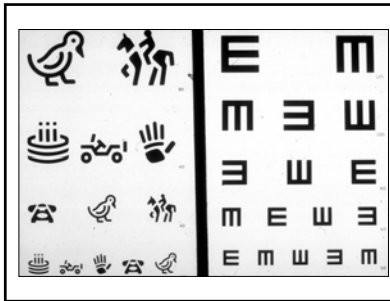
Fixation

- > 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

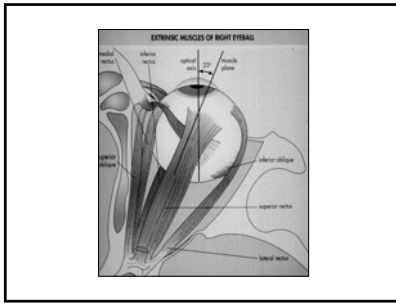
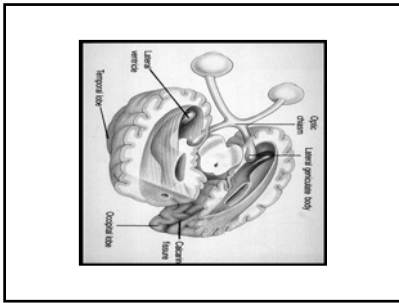
- > Amblyopia Treatment Study Group
- > Looking at measurement of visual acuity
 - Efforts to standardize
- > Effects of treatment on child and family
- > Comparison of
 - Drops and patching
 - Shorter vs. longer occlusion

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°



MUSCLE	ORIGIN	INNERVATION	SIZE (mm)	ACTING ON	WHICH MUSCLE POSITION	INNERVATION
Superior rectus	Annulus of Zinn	3.5 mm from lateral border	107	4.5	7	Abduction CN III
Lateral rectus	Annulus of Zinn	6.5 mm from lateral border	97	7	12	Adduction CN VI
Inferior rectus	Annulus of Zinn	7.2 mm from superior border	23	4	4.5	Elevation Upper Adduction CN III
Medial rectus	Annulus of Zinn	6.5 mm from medial border	23	7	6.5	Depression Adduction CN III
Superior oblique	Orbit apex above annulus of Zinn	Functional origin at trochlea	31	26	7.8	Intorsion Depressor
Inferior oblique	Orbit apex above annulus of Zinn	Functional origin at the trochlea	31	1	13	Extorsion Elevator
Levator palpebrae superioris	Orbit apex above annulus of Zinn	Functional origin at the trochlea	14-20	—	—	Eyelid elevation

*Numbers in small text in previous position

EOM'S Origin Annulus of Zinn

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

EXCEPT:

- inferior oblique
- superior oblique
- levator palpebrae superioris

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)

LPS

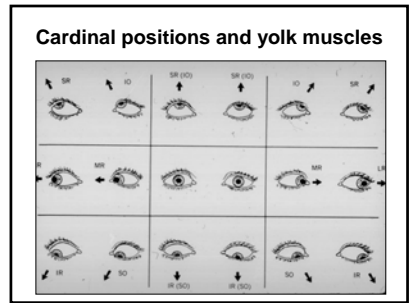
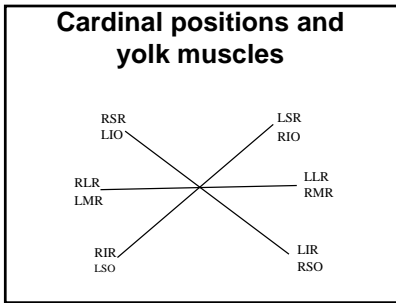
- orbital apex above annulus

Insertions of the EOM's Spiral of Tillaux

- Rectus muscles insert into sclera anterior to the equator via tendons
- 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



EOM'S function

- function dependent upon position of the globe
- 1°, 2°, 3° 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

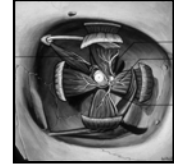
ET in thyroid patients with tight inferior recti

EOM - Innervation

- CN III LPS, SR upper
- MR, IR, IO lower

- CN IV SO

- CN VI LR

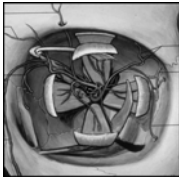


Anatomy of the EOM's Vascular supply

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

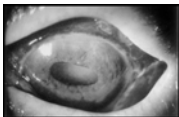
Recti muscles - perfuse anterior segment via anterior ciliary arteries

Oblique muscles - do not contribute to anterior segment circulation

Vascular supply

➤ Clinical Relevance

- Surgery on multiple recti muscles contraindicated due to risk of anterior segment ischemia



Summary NRC

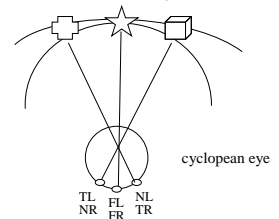
➤ Horopter

- 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



**Summary
Retinal correspondence**

➤ Retinal correspondence

ARC NRC

Binocular

**Summary
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

**Summary
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

GOOD LUCK!!!!



QUIZ

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.
 2. Maddox rod testing.
 3. Cover-uncover testing.
 4. Hirschberg testing.
- a. 1, 2, and 3. b. 1 and 3.
c. 2 and 4 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. Lateroversion reflex test
- ANS =

19. **Paradoxical 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

Left head tilt: LHT = 15

30	15	0
15	LHT = 10	0
0	0	0

Right gaze

Left gaze

- a. Tuck LSO
 - b. Tuck LSO, recess LIO
 - c. Recess LIO
 - d. Recess RSR and LIR
- ANS =