# Amblyopia and Strabismus In The Pediatric Patient

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## Prior to Beginning This Lecture On Amblyopia and Strabismus In The Pediatric Patient…

- Pediatric eye exam will be outlined (to supplement Basic Eye Examination lecture)
- NO recorded lecture accompanies these initial slides on the Pediatric Eye Exam

## Pediatric Ocular History

- A history of any of the following may warrant Ophthalmology evaluation:
  - Glasses
  - Amblyopia (reduced best-corrected vision not fully attributable to a structural issue in the eye)
  - Strabismus (eye misalignment)
  - Ptosis (droopy eyelid)
  - Leukocoria (pale pupillary red reflex on casual observation or in photos)
  - Ocular tearing, irritation/redness, rubbing
  - Eye trauma
  - Previous eye surgery
  - Parental concern about the eyes
  - Family history of eye disorder (especially one presenting in infancy or childhood)

## External Inspection

- Inspection of the head, face, lids, orbits may identify:
  - Epiphora (tearing)
  - Blepharospasm (blinking)
  - Photophobia (sensitivity to light)
  - Eyelid issue (mass, malposition, asymmetry, or droop)
  - Proptosis (forward “bulging” of eye(s))
  - Facial asymmetry
  - Nystagmus (jiggle of the eyes)
  - Head position (head tilt or turn, chin up or down)

## External Inspection

- Eye or orbital abnormality on external inspection often requires Ophthalmology evaluation
- The clinical triad of photophobia, tearing, and blepharospasm in an infant or child may be signs of Infantile or Juvenile Glaucoma and require prompt evaluation with Ophthalmology
- Penlight inspection of the anterior segment
  - Abnormality or asymmetry of the anterior segment of the eye requires ophthalmology evaluation
  - Discussed in Basic Eye Examination

## Red Reflex Test

- To identify leukocoria (pallor or whitening of the pupillary red reflex)
- Dim the room lights
- From 1 meter away, view both eyes of child through the direct ophthalmoscope paying attention to the red reflex
  - Look for symmetry, and a full regular reflex
  - Dark spots, asymmetric color, white reflex, absent reflex, diminished reflex are of concern
- Abnormal or absent red reflex requires ophthalmology evaluation
- Discussed further in lecture that follows

## Corneal Light Reflex Test

- Objective assessment of ocular alignment
  - May suggest an ocular misalignment
  - Helpful in evaluation of pseudostrabismus (appearance of strabismus with normal ocular alignment)
- Hold penlight at 33 cm (fixation target)
- Compare position of corneal light reflex in both eyes
  - Normal: symmetric corneal light reflex located slightly nasal to center of both pupils
  - Asymmetric corneal light reflex may indicate ocular misalignment
    - ***Must evaluate further with cover testing to confirm
- Discussed further in lectures:
  - Basic Eye Examination
  - Lecture that follows

## Pupils

- Swinging flashlight test
- Pupils should be equal in size, round, and equally react to light
- There should NOT be an afferent pupillary defect
- Assess for:
  - Symmetric constriction to light
    - Sluggish or poor reactivity, asymmetric size, or afferent pupillary defect (Marcus Gunn Pupil) require Ophthalmology evaluation
- Discussed further in Basic Eye Examination

## Strabismus Tests

- Corneal light reflex test
  - A guesstimate that may suggest ocular misalignment
  - Evaluate further with cover testing
- Cover testing
  - Movement of either eye on cover-uncover test is suggestive of strabismus
- Abnormality on corneal light reflex test or cover testing is suggestive of strabismus and requires ophthalmology evaluation

## Vision Testing

- Divided primarily by age
  - Infancy
  - Preverbal
  - Verbal
- MONOCULAR test
- Tested with glasses in place
- Visual acuity improves after birth with visual maturation, so referral criteria are age dependent

## Vision Testing: Infants and Preverbal Children

- Infancy (first 2 to 3 months of life)
  - Assess blink to light in each eye
  - Muscle light or direct ophthalmoscope on high and swing light over eye
    - Blinks to light vision (normal)
    - Poor blink to light (abnormal) --> requires ophthalmology evaluation
- Preverbal (age 2 to 3 months to around 3 years of age)
  - Check each eye for fixation and ability to follow
  - Interesting object or toy held in front of one eye
  - Cover the other eye with your hand)
  - Move toy in various directions noting the child's fixation and ability to maintain fixation and follow the toy
  - Assess for:
    - Preference of one eye --> requires ophthalmology evaluation
    - Inability to fix or follow with one or both eye(s) -->requires ophthalmology evaluation
  - Discussed further in lecture that follows

## Vision Testing: Verbal Child

- Best corrected monocular vision with glasses in place
  - May sometimes need to encourage child to guess
  - Use a patch or the parent's hand to cover one eye (children peek)
- Use the highest cognitive test the child is able to perform (child must be able to identify the tested symbols)
  - Snellen letters (highest cognitive difficulty)
  - Snellen numbers
  - Tumbling E
  - HOTV
  - Allen pictures or LEA symbols (least cognitive difficulty)
- Test vision with ROW of symbols that the child understands
  - Can have parent point to each symbol along the line if child has difficulty progressing along the row

## Vision Testing: Verbal Child

- Age based referral criteria ****
  - Age less than or equal to 5 years: two line difference between the eyes and/or less than 20/40 vision in one or both eyes
  - Age 6 years and older: two line difference between the eyes and/or less than 20/30 vision in one or both eyes
- Discussed further in lecture that follows

## Extraocular Motility

- Examined in 9 positions of gaze
- Head is stabilized in primary position
- Move interesting object/toy into one position of gaze and then return to primary position
- Note overactions (excess movement) and underactions (lagging movement) of muscles
- May identify nerve palsy or restriction of eye(s)
- Discussed further in lectures:
  - Basic Eye Examination
  - Lecture that follows

## And now the lecture...

- Please proceed to the recorded Lectures which include images on slideshow

## Amblyopia and Strabismus In The Pediatric Patient

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## Objectives

- Obtain examination skills to screen for amblyopia and strabismus
- Identify clinical signs of amblyopia inducing risk factors, and clinical characteristics of amblyopia and strabismus
- Understand importance of early identification and treatment of amblyopia and strabismus
- Become familiar with ophthalmology treatment protocol for amblyopia and strabismus

## Amblyopia and Strabismus

- Amblyopia
  - Reduced best-corrected vision not fully attributable to a structural issue in the eye
- Strabismus
  - Eye misalignment
- "Lazy eye"
  - Lay term for amblyopia, strabismus, or ptosis
- Amblyopia can result in strabismus, AND strabismus can result in amblyopia...

## Amblyopia

- Decreased best-corrected vision in one (or both) eyes
- Reduced vision is not directly attributable to a structural issue of the eye or posterior vision pathways
  - i.e., if structural abnormality is corrected the amblyopic eye's vision would still be sub-normal (needs treatment)
- Develops PRIOR to maturity of the visual system
- Generally unilateral (can be bilateral)
  - Difference in best-corrected vision of more than 2 lines between the two eyes on vision testing
- Common vision problem in children
  - Responsible for more vision loss in children than combination of all other causes
- If detected early and treated early, can be correctable


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## Amblyopia

- Secondary to:
  - Strabismus
    - Most common
  - Refractive error
    - Second most common
  - Stimulus deprivation
    - Least common…. More about these later…

## Amblyopia Onset

- Amblyopia develops PRIOR to maturity of the visual system
- Normal visual development:
  - Continuous process beginning at birth progressing through age 8 to 9 years
    - Most vulnerable period is during the first 2 years of life, particularly the first 3 months after birth
  - During the vision "learning" period the visual pathways are "plastic"
  - Visual development requires ONGOING stimulation of vision receptors in the brain, from birth onward

## Development of Amblyopia

- Visual stimulation/input is DISRUPTED in an eye during immature visual system period
  - Earlier the age of onset the worse the potential amblyopia if not corrected
- Eye with an amblyopia-inducing risk factor sends a distorted or blurred image to brain →
- Brain visual cortex selectively ignores/suppresses the distorted/blurred input from that eye →
- Brain prefers/favors other eye with better vision→
- Vision pathways from amblyopic eye do not develop normally (anatomic changes occur in the brain)
- Ultimately, this interruption of the normal developmental process results in amblyopia

## Three Major Types of Amblyopia

- Strabismic amblyopia
  - Ocular misalignment
- Refractive amblyopia
  - Glasses need
    - Anisometropic (unequal glasses need)
    - Isometropic (high glasses need in both eyes)
- Stimulus deprivation amblyopia
  - Occlusion of visual axis… More about these later…

## Diagnosis of Amblyopia

- IDENTIFICATION of amblyopia inducing risk factor(s) and reduced best-corrected vision
- Diagnosis through vision screening
- PCP vision screening
  - Red reflex
  - Fixation pattern and visual acuity (identify monocular preference)
  - Strabismus evaluation (cover testing)
  - +/- Photoscreening to identify risk factors

## Amblyopia and Strabismus

- Amblyopia can result in strabismus, AND strabismus can result in amblyopia…
- Early detection allows for timely treatment
- Early identification by the primary care provider allows for further early evaluation with pediatric ophthalmology (including a full eye exam) with subsequent initiation of treatment in a timely manner
- Without treatment in a timely basis, visual impairment may not be correctable and could persist for a lifetime

## Examination

- Red reflex and corneal light reflex
- Pupils
- Visual acuity
- Cover testing
- Head position
- Extraocular motility
- Fundus exam

## Red Reflex Test

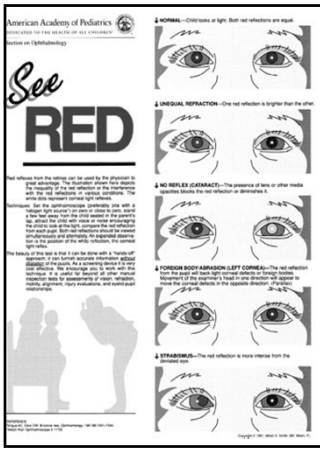
- = Bruckner test
- With room lights dimmed (pupils will dilate allowing improved view of larger reflex)
- View both eyes of child through the direct ophthalmoscope paying attention to the red reflex
  - Look for symmetry, and a full regular reflex
  - Dark spots, asymmetric color, white reflex, absent reflex, diminished reflex are of concern

## Leukocoria

- Red reflex test:
  - Screening evaluation to identify sign of visual axis opacity or posterior segment anomaly
    - Focus direct ophthalmoscope onto pupils from 1 meter away
  - Normal red reflex:
    - Symmetric
  - Abnormal red reflex:
    - Asymmetric
    - Irregular
    - Pale/whitened (leukocoria)

## Corneal Light Reflex Test

- Objective assessment of ocular alignment
  - May suggest an ocular misalignment (must further assess with cover testing to confirm)
- Penlight held at 33 cm (fixation target)
- Compare position of corneal light reflex in both eyes
  - Normal alignment suggested by symmetric corneal light reflex located slightly nasal to center of the pupil
  - Asymmetric corneal light reflex may indicate ocular misalignment
    - Fixing "normal" eye
      - corneal light reflex is slightly nasal to central pupillary axis
    - Strabismic eye
      - corneal light reflex is deviated either nasally or temporally….
    - ***Must evaluate further with cover testing to confirm




## "See Red"

- Alfred G. Smith, MD
- Red Reflex Examination in Neonates, Infants, and Children. American Academy of Pediatrics, Section on Ophthalmology, American Association for Pediatric Ophthalmology and Strabismus, American Academy of Ophthalmology, and American Association of Certified Orthoptists. Pediatrics December 2008; 122:6 1401-1404; doi:10.1542/peds.2008-2624

## Red Reflex

- ***Criterion for pediatric ophthalmology referral:***
  - ***Abnormal red reflex***
  - ***Absent red reflex***
  - ***Concern for leukokoria (white pupil)***
  - ***Family history of high-risk eye issues (even if normal red reflex):***
    - ***Retinoblastoma***
    - ***Infantile cataract***
    - ***Infantile or pediatric retina problem***
    - ***Infantile or pediatric glaucoma***
    - ***Other concerning eye issue***

## Vision Testing

- Normal ocular alignment does not mean normal vision in both eyes
- Monocular visual acuity testing is necessary to assess for signs of amblyopia

## Vision Testing

- If preverbal, check for fixation preference
  - Cover each eye and check fixation and following for other eye
  - Assess ease of one eye following an interesting object (e.g., toy)
  - Assess for objection to cover --> suggests preference of covered eye
    - Child may less consistently follow object
    - Child may move head to peek around cover
    - Child may cry with covering of one eye

## Vision Testing

- If verbal, optotype vision testing
- Ensure child knows the images being tested
- Best corrected vision (test WITH glasses)
- One eyed testing
  - ***Tape a patch over non-tested eye (no peeking)
- LINE vision
  - Not single optotypes (over-estimates vision in amblyopia)

## Vision Testing

- ***Criterion for pediatric ophthalmogy referral:***
  - ***Poor blink to light in preverbal infant or child***
  - ***Fixation preference during vision testing***
  - ***Difference in best-corrected vision between the two eyes of 2 lines or more***
  - ***Poor performance on vision testing in one or both eyes (age-based criteria)***
  - Age based referral criteria ****
    - Age less than or equal to 5 years: two line difference between the eyes and/or less than 20/40 vision in one or both eyes
    - Age 6 years and older: two line difference between the eyes and/or less than 20/30 vision in one or both eyes

## Cover Testing

Eyes appear to be straight

Cover testing → Left eye covered

And uncovered → Left Exotropia

- Cover-uncover test:
  - Performed with glasses in place
  - Fixate on distant interesting accommodative object, and then repeat with a near target (interesting colorful noise-making toy or eye chart)
  - Cover test:
    - Occluder is used to cover the right eye and watch for refixation movement of the uncovered left eye
    - Then cover the left eye …
    - On covering of one eye any movement of non-occluded other eye indicates tropia
  - Uncover test:
    - Cover the right eye and watch for refixation movement of the right eye when the cover is removed
    - Then cover the left eye …
    - Movement of occluded eye on removal of cover from that eye indicates phoria, or intermittent tropia

## Cover-Uncover Test

- ***Movement of either eye on cover-uncover test is suggestive of strabismus and requires pediatric ophthalmology evaluation***

## Anomalous Head Position (Torticollis)

- May assume head position…
  - Head tilt and/or
  - Face turn
- To compensate for:
  - Strabismus
    - Horizontal muscle palsy → head turn into field of paralytic muscle
    - Fourth nerve palsy → head tilt towards opposite side of paresis
    - Eyes straight on anomalous head position, with strabismus noted on head straightening (check with cover test)
  - Nystagmus (jiggling of eyes)
  - Ptosis (drooping of eyelid)
  - Refractive error (glasses need)
- Non-ocular etiologies include:
  - Bony malformations of the cervical vertebrae
  - Abnormalities of the sternocleidomastoid

## Torticollis

- ***Consider eye etiology, as this may be a compensatory mechanism for amblyopia inducing issue such as strabismus, refractive error, or ptosis, which would warrant pediatric ophthalmology evaluation***

## Extraocular Motility

H, And up, down

Motility evaluation to assess
for signs of over-/under-action
Of the extraocular muscles

Check versions,
Then ductions PRN

## Continued on Next Lecture

## Three Major Types of Amblyopia

- **Strabismic amblyopia**
  - **Ocular misalignment**
- Refractive amblyopia
  - Glasses need
    - Anisometropic (unequal glasses need)
    - Isometropic (high glasses need in both eyes)
- Stimulus deprivation amblyopia
  - Occlusion of visual axis

## Strabismic Amblyopia

- Ocular misalignment
  - Most common form of amblyopia
  - Unilateral
- Two eyes are not simultaneously directed at the object of regard -->
- Brain cannot fuse two different images →
- During the visually immature period brain will compensate by ignoring the image from one eye →
- Brain develops a preference to input of fixing eye with reduced responsiveness to input from misaligned eye
  - Cortical suppression
  - Adaptation of ignoring the image from the misaligned eye also results in interruption of the ongoing visual stimulation required for normal visual development --> amblyopia

## Strabismus (a few definitions)

- Orthophoria = straight eyes
- Strabismus = misalignment of the eyes
  - Affects about 4% of US population
  - Direction of deviation (in, out, up, down)
    - Horizontal: Eso (= In), Exo (= Out)
    - Vertical: Hyper (= Up), Hypo (= Down)
  - Frequency (phoria, tropia)
    - Phoria = *Latent tendency toward eye deviation*
      - Controlled by fusion under binocular viewing (both eyes open)
      - Present when covering one eye
    - Tropia = *Manifest eye deviation*
      - Present under binocular viewing (both eyes open)
        - Intermittent = present part of the time
        - Constant = always present
  - Amount of the deviation (measured in prism diopters)
  - Etiology of the strabismus
    - Paralytic → neurologic cause
    - Non-paralytic
      - A neuromuscular abnormal control of eye movement (majority of strabismus)
    - Special forms and restrictive

## 6 Extraocular Muscles

(plus Levator Palpebrae)

- Horizontal rectus muscles
  - Medial rectus (CN III)
    - Adduction
  - Lateral rectus (CN VI)
    - Abduction
- Vertical rectus muscles
  - Superior rectus (CN III)
    - Elevation, incyclotorsion, adduction
  - Inferior rectus (CN III)
    - Depression, excyclotorsion, adduction
- Vertical oblique muscles
  - Superior oblique (CN IV)
    - Incyclotorsion, depression, abduction
  - Inferior oblique (CN III)
    - Excyclotorsion, elevation, abduction
- Innervation:
  - "LR 6, SO 4 (rest are 3)"

## Some Strabismus Risk Factors

- Prematurity
- Positive family history
- Hydrocephalus
- Developmental delay
  - Incl. Cerebral palsy, Down Syndrome
- Certain neurological disorders
  - Eg., brain tumors, stroke
- Certain genetic disorders
- Certain systemic disorders
- Trauma
  - Incl. orbital fracture, traumatic brain injury, shaken baby syndrome

## Strabismus Deviations

- Esotropia
  - Convergent deviation → inward crossing of eye(s)
  - "cross-eyes"
- Exotropia
  - Divergent deviation → outward drifting of eye(s)
  - "wall-eyes"
- Hypertropia
  - Elevated eye
- Hypotropia
  - Depressed eye

## Pseudostrabismus

- In the differential of true strabismus
- "Appearance" of strabismus BUT eyes are not truly misaligned
  - Note the normal corneal light reflex
  - ***May have superimposed true strabismus (***need to always check with cover testing to rule-out true strabismus***)
- Commonly illusion of esotropia due to skin covering of normally visible nasal sclera
- Most commonly secondary to
  - Prominent epicanthal skin folds of the eyelids
  - Wide and flat nasal bridge

## Pseudostrabismus

**Pseudostrabismus** **Versus True Esotropia**

Note the corneal light reflex
***Must perform cover testing to confirm***

## Esotropia

Convergent ocular deviation

## Infantile/ Congenital Esotropia

- Onset < 6 months age
- Familial
- Large constant convergent deviation
- Alternate eye fixation
- Differential includes Cranial Nerve VI palsy
- Treatment:
  - Refractive correction PRN
  - Amblyopia treatment PRN
  - **Strabismus surgery
    - EARLY: *Around 6 months of age or earlier*
- Recurrent (70% require additional surgery)

## Accommodative Esotropia

- = Refractive Esotropia
- Onset 6 mos. to 7 yrs.
- Convergent ocular deviation associated with effort of focusing
- Moderate to high hyperopia
- Background (Normal):
  - Focus at near → lenticular accommodation & simultaneous normal amount of ocular convergence
- Accommodative Esotropia:
  - Work of focusing (accommodating) --> results in over-convergence through multiple mechanisms
- Treatment:
  - Refractive correction -->relax focusing effort → decrease over-convergence
    - Eyes often still cross without glasses
    - +/- Bifocals if more crossing at near
  - Amblyopia treatment PRN
  - Strabismus surgery PRN

## Acquired Esotropia

- = Non-refractive Esotropia
- Onset > 6 months age
- Deviation:
  - EQUAL at distance & near
- Symptoms:
  - Diplopia
- Refractive error:
  - Insignificant hyperopia
- Differential:
  - CNS lesion
  - Cranial Nerve VI Palsy
- Treatment:
  - Consider neuro work-up
  - Refractive correction PRN
  - Amblyopia treatment PRN
  - Strabismus surgery PRN

## Exotropia

Divergent ocular deviation

## Intermittent Exotropia

At times the eyes are straight
And then…

Right eye out

Left eye out

- Most common exodeviation (except exophoria at near)
- Onset usually < 5 years
- Deviation:
  - Initially intermittent and greater at distance --> progresses...
  - Apparent during visual inattentiveness, fatigue, stress (often more notable later in the day)
- Signs:
  - Squinting
  - Closing one eye in bright sunlight
  - Rubbing of the eyes
- Treatment:
  - Refractive correction PRN
  - Amblyopia treatment PRN
  - Monitor with frequent exams
    - Watch for increased angle & frequency (signs of deterioration)
  - Strabismus surgery PRN

## Intermittent Exotropia

Eyes straight

- Remember to perform cover testing***

Cover testing → Left eye covered

And uncovered → Left Exotropia

## Intermittent Exotropia

**Intermittent Exotropia →** **May Progress to Exotropia**

## Exotropia

Divergent ocular deviation

## Congenital Exotropia

- Rare
- Onset < 6 months age
- Deviation:
  - Large constant exotropia
- Often associated with:
  - **Neurologic impairment
  - Craniofacial disorders
- Treatment:
  - Work-up
  - Refractive correction PRN
  - Amblyopia treatment PRN
  - Strabismus surgery
    - As early as 6 months of age

## Hypertropia

Left Hypertropia
OR
Right Hypotropia
(Depends on fixing eye)

## Incomitant Strabismus

- Ocular misalignment varies with gaze
  - Extraocular muscle issue
  - Extraocular nerve issue
  - Mechanical restriction issue
- Some etiologies
  - Restrictive strabismus
  - Orbital disease
  - Paralytic strabismus
  - Systemic disease

## Third Nerve Palsy

- Cranial Nerve III innervates medial/superior/inferior rectus and inferior oblique
- Exotropia usually with hypotropia
  - Paretic eye is down and out
  - If complete palsy → associated ptosis and pupillary dilation
- Etiology:
  - In children, congenital etiology is more common than acquired
- However may still require work-up, especially if pupil involvement or other

## Fourth Nerve Palsy

- CN IV innervates superior oblique
- Hypertropia of paretic eye
  - →+/- head tilt towards opposite side of paretic eye
- Etiology:
  - Congenital
  - Acquired (head trauma)
  - May require work-up

## Sixth Nerve Palsy

- Cranial nerve VI innervates lateral rectus (limited abduction)
- → paretic eye is esotropic, especially at distance and in gaze towards the affected eye
- May assume a head turn toward the paretic eye to avoid need for abduction of that eye
- Etiology:
  - Usually acquired (trauma or CNS lesion/elevated ICP)
  - Congenital is rare
- Requires work-up

## Strabismus

- ***Criterion for pediatric ophthalmology referral:***
  - ***Any ocular misalignment still present after one (to three) months of age***
    - ***Unless:***
      - ***Large angle deviation → refer earlier***
      - ***Constant deviation → refer earlier***
      - ***Preference of one eye --> refer***
      - ***Associated with abnormal red reflex or other ocular anomaly/signs/symptoms→ refer on presentation***
      - ***Poor blink to light (infancy) or poor vision --> refer***
      - ***If uncertain --> refer***
      - ***Other (based on history, associated medical condition) --> refer***
      - ***Remember, strabismus may be a sign of a more serious condition (e.g., optic nerve glioma, CNS tumor, elevated ICP, retinoblastoma….) and may result in amblyopia***

## Strabismus Treatment

Post-operative orthophoria

- All patients require a full eye exam (including dilation)
- Refractive correction PRN
- Amblyopia treatment PRN
  - Occlusion therapy of preferred eye
    - Patching
    - Atropine
- Strabismus surgery PRN
  - Recession (weakening procedure)
  - Resection (strengthening or tethering procedure)
  - Other

## Three Major Types of Amblyopia

- Strabismic amblyopia
  - Ocular misalignment
- Refractive amblyopia
  - Glasses need
    - Anisometropic (unequal glasses need)
    - Isometropic (high glasses need in both eyes)
- Stimulus deprivation amblyopia
  - Occlusion of visual axis

## Refractive Amblyopia

- Anisometropic Refractive Amblyopia (unequal glasses need)
  - Second most common form of amblyopia
  - Unilateral
  - Unequal amount of refractive error in two eyes
    - Eye with less refractive error --> clear(er) image
    - Eye with higher refractive error --> constantly out of focus with image distortion
  - Eye with clearer vision is "favored," and other eye is susceptible to amblyopia

## Refractive Amblyopia

- Isometropic Amblyopia (high glasses need in both eyes)
  - Blurred image for both eyes
  - Bilateral

## Refractive Amblyopia Treatment

- All patients require a full eye exam (including dilation)
- Refractive correction (glasses) full-time
- Amblyopia treatment PRN
- Usually more responsive to treatment, HOWEVER often diagnosed much LATER while no external signs of issue
- Identification is often dependent on vision screening

## Three Major Types of Amblyopia

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  - Ocular misalignment
- Refractive amblyopia
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    - Anisometropic (unequal glasses need)
    - Isometropic (high glasses need in both eyes)
- Stimulus deprivation amblyopia
  - Occlusion of visual axis

## Stimulus Deprivation Amblyopia

- Visual axis occlusion or obstruction disrupts visual development in the involved eye(s)
  - Least common form of amblyopia
  - Often most difficult to treat
  - May be unilateral or bilateral
  - Secondary to:
    - Media opacity (cornea, lens, vitreous, retina)
      - Cataract
      - Corneal opacity/scar
      - Vitreous hemorrhage
      - Retinal lesion
    - Ptosis (congenital, secondary to lid lesion such as a hemangioma)

## Stimulus Deprivation Amblyopia

- Stimulus deprivation amblyopia is most commonly secondary to a congenital or acquired cataract
- Congenital cataract(s) will blur the image to one or both eyes during most visually critical period after birth--> so early detection is critical to prompt surgical management and early visual rehabilitation
- Red reflex test
  - Look for leukocoria

## Stimulus Deprivation Amblyopia

- Congenital cataracts account for 10% of all visual loss in children worldwide
  - Incidence: 1/250 newborns (50% U/L, 50% B/L)
  - Variety of hereditary & etiologic factors
- Visual significance:
  - Size, location & density of opacity
  - Any central opacity or surrounding lenticular distortion 3 mm + in size is visually significant
- Treatment:
  - Early lensectomy +/- Intraocular lens implantation
  - Post-op refractive correction
    - Contact lens
    - Glasses
  - Aggressive post-op amblyopia treatment (occlusion therapy) for 10 years
- Visual outcome
  - Best visual acuity if surgery prior to 2 months age, followed by compliance with amblyopia management
  - Bilateral aphakes do better than unilateral secondary to competition (amblyopia)

## Amblyopia --> Sensory Strabismus

U/L strabismus secondary to decreased vision/amblyopia in affected eye

H/O Congenital Cataract right eye,
Secondary glaucoma right eye,
Dense amblyopia right eye,
Sensory exotropia right eye

## Sensory Esotropia or Exotropia

H/O Congenital cataract left eye,
Aphakic left eye,
Non-compliant with glasses,
Dense amblyopia left eye,
Sensory esotropia left eye

- Unilateral strabismus secondary to decreased monocular visual acuity in affected eye
  - Develop preference for other eye & amblyopia in affected eye, and then strabismus in amblyopic eye
- Etiology (examples):
  - Refractive error/anisometropia
  - Corneal opacities (scar)
  - Lenticular opacities (cataract)
  - Optic nerve atrophy/hypoplasia
  - Macular lesions
  - Intraocular tumor
  - Retinal detachment (chronic)
- Treatment:
  - Management of primary disorder
  - Refractive correction PRN
  - Amblyopia treatment
  - Strabismus surgery PRN

## Organic Amblyopia

- Amblyopia in presence of ADDITIONAL vision loss from uncorrectable structural abnormality of the eye
  - Eye has poorer vision because of structural issue and secondarily develops amblyopia
- Optic nerve anomaly
  - Hypoplasia
  - Coloboma
- Retinal lesion/anomaly
  - Scar
  - Coloboma
- Trial of amblyopia treatment is “ALWAYS” indicated

## Organic Amblyopia

Abnormal reflex
Left eye

- Check for leukocoria
- Red reflex test
  - Screening evaluation to identify sign of visual axis opacity or posterior segment anomaly
    - Focus direct ophthalmoscope onto pupils from 1 meter away
  - Normal red reflex:
    - Symmetric
  - Abnormal red reflex:
    - Asymmetric
    - Irregular
    - Pale/whitened

## And Then...

Total
Retinal
Detachment

- Retinoblastoma
  - Most common malignant ocular tumor of childhood
    - Incidence: 1/15,000
  - Diagnosed by age
    - 1 year in familial or B/L cases
    - 1 – 3 years in sporadic U/L cases
  - Signs:
    - **Leukocoria (most common)
    - +/- Strabismus (25%)
    - +/- Vitreous hemorrhage, hyphema, inflammation, glaucoma, proptosis, hypopyon
  - Requires aggressive emergent treatment
    - Prompt treatment can be life-saving

## Amblyopia Treatment

- Treatment is more successful prior to development of mature visual system
  - Early (earlier) treatment is best
    - Generally, the younger the age the more successful the treatment
  - Less substantial improvement in vision often after the age of 8 to 9 years
- Optimize the retinal image in the amblyopic eye
  - Clear the visual axis by clearing any obstacle to vision
  - Clear the image that the eye presents to the brain by correcting any significant refractive error (glasses)
- Force use of the amblyopic eye (occlusion therapy)
  - Requires "work" by family; compliance can be difficult

## Amblyopia Treatment

- May require occlusion therapy of preferred eye to force brain to use image from the amblyopic eye
  - Enhances stimulus to the visual cortex and strengthens the pathways
    - Patching of preferred eye forces child to fix with amblyopic eye
      - Adhesive eye patches, NOT pirate patch
      - More effective when performing near activities during patching
    - Pharmacologic penalization with atropine eye drops to blur image in the preferred eye
- "Vision therapy" does not treat amblyopia or strabismus, and may in fact delay initiation of appropriate treatment
- If strabismus, treat amblyopia first (strabismus surgery PRN)
- Close follow-up is necessary to assess response to amblyopia treatment and compliance
  - GOAL is best-corrected 20/20 vision in either eye, or Snellen acuity vision that differs by one line or less between the two eyes
  - ***Window of opportunity for treatment (earlier is better)
- Amblyopia may recur and sometimes requires maintenance treatment

## Strabismus Treatment

- Treat any co-existing conditions
- Refractive correction PRN
- Amblyopia treatment PRN
- Strabismus surgery PRN
- Always best to intervene when strabismus is intermittent, PRIOR to decompensating to a constant deviation with development of suppression

## Conclusions

- Goal: EARLY IDENTIFICATION of amblyopic risk factors
  - If congenital etiology there is a short window for intervention (first few weeks of life)
- Goal: EARLY DIAGNOSIS of amblyopia and strabismus
- Goal: EARLY INTERVENTION
  - Allowing for more successful treatment
- Criterion for complete ophthalmology evaluation
  - Amblyopia
  - Strabismus
  - Leukocoria...
- If dense amblyopia or amblyopia treatment is not successful, must PROTECT the preferred eye
  - Polycarbonate glasses full-time and sports goggles

## Thank you

Great websites for browsing:
http://www.aapos.org
http://www.telemedicine.orbis.org