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

**Strabismus Tests**
- Corneal light reflex test
  - A guesstimate that may suggest ocular misalignment
  - Evaluate further with cover testing
- Cover testing
  - Movement of one 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 blinks 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 eyes(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

**Pupils**
- Swinging flashlight test
- Pupils should be equal in size, round, and equally react to light
- There should NOT be an afferent pupillary defect
- Must evaluate further:
  - Symmetric constriction to light
  - Sluggish or poor reactivity, asymmetric size, or afferent pupillary defect (Marcus Gunn Pupil) require ophthalmology evaluation

**Discussed further in lectures:**
- Basic Eye Examination
- Lecture that follows
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

- 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...

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 In The Pediatric Patient

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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 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

Three Major Types of Amblyopia
- Strabismic amblyopia
  - Ocular misalignment
- Refractive amblyopia
  - Glasses needed
    - Anisometropic (unequal glasses need)
    - Isometropic (high glasses need for both eyes)
- Stimulus deprivation amblyopia
  - Occlusion of visual axis... More about these later...

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

Secondary to:
- Strabismus
  - Most common
- Refractive error
  - Second most common
- Stimulus deprivation
  - Least common... 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

- **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

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

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)

- **Criterion for pediatric ophthalmology 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

- 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 straigtening (check with cover test)
  - Nystagmus (jiggling of eyes)
  - Ptosis (drooping of eyelid)
  - Refractive error (glasses needed)
- 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

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

**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: Exo (= 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)
  - Tropia = Manifest eye deviation
    - Present when covering one eye
    - 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

**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

**Strabismic Amblyopia**
- Ocular misalignment
  - Most common form of amblyopia
- 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

**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

**6 Extraocular Muscles**
(plus Levator Palpebrae)
- Horizontal rectus muscles
  - Medial rectus (CN III)
    - Abduction
  - 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)
    - Ircyclotorsion, depression, abduction
  - Inferior oblique (CN IV)
    - Eycyclotorsion, elevation, abduction
- Innervation:
  - "LR 6, 5O 4 (rest are 3)"
### 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 illusion of esotropia due to prominent epicanthal skin folds of the eyelids
  - Wide and flat nasal bridge

### 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

### Note the corneal light reflex

***Must perform cover testing to confirm***
Exotropia

Divergent ocular deviation

Intermittent Exotropia

- 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

At times the eyes are straight
And then...

Right eye out

Left eye out

Intermittent Exotropia

- Remember to perform cover testing***

Eyes straight

Cover testing  Left eye covered

And uncovered  Left 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
- 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 hypertropia
  - 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

- 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 → clearer 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

- 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

- 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
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

- 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 or less is visually significant
  - Treatment:
    - Early lens extraction +/- 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

- 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

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

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**

- 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...**

- 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

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**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

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**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

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**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