Ophthalmology Lecture 6: What not to miss in Pediatric Ophthalmology

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What not to miss in Pediatric ophthalmology

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OUTLINE

- Introduction
- The problem of Childhood blindness
- Common pediatric eye disorders
  - Amblyopia
  - Strabismus
  - Cataract
  - Congenital glaucoma
  - Retinoblastoma
  - Red eyes
- Eye Screening for children and Prevention of childhood blindness
INTRODUCTION

- Although there are many more blind adults than blind children, a blind child will have their whole life ahead of them rather than just their older years.
- Newborns, infants, and young children can suffer from many of the same external and internal eye disorders as adults, including those often associated with old age, such as cataracts and glaucoma.
- Some disorders, however, are more common or even unique to young patients, such as retinoblastoma, squint, and retinopathy of prematurity.
WHY DOES IT MATTER?

• The most important difference between the treatment of adult patients and young patients, especially those under the age of two years, relates to the timing of treatment.

• In adults, for conditions that reduce or distort the passage of light to the retina, such as refractive error, squint, ptosis, pterygium, and cataract, delaying treatment usually will not adversely affect the outcome of treatment.

• Progressive conditions, such as infections, glaucoma, and diabetic retinopathy, clearly, must be treated as quickly as possible, to minimize further damage to the eye.
WHY DOES IT MATTER?

• In pediatric patients, however, any condition that reduces the amount of light passing through the eye, stimulating the retina, and sending signals to the visual cortex will negatively affect the brain’s ability to learn to see and result in permanent vision loss.

• Progressive conditions, if not treated quickly, will permanently reduce the child’s vision.

• Therefore, it is essential to treat all pediatric eye disorders as quickly as possible.
Childhood Blindness - Definition

• A group of diseases and conditions occurring in childhood or early adolescence, which, if left untreated, result in blindness or severe visual impairment that are likely to be untreatable later in life (WHO)

• Blindness in a child

• Child is < 16 years (UNICEF)

• Blindness = presenting visual acuity (VA) <3/60 in the better eye
  = loss of walk-about vision or independent navigation

Common Causes- Worldwide

1. Corneal scarring
   - Vitamin A deficiency
   - Measles
   - Ophthalmia neonatorum
   - Harmful eye practices
   - Infective corneal ulcer
2. Congenital cataract
3. Uncorrected Refractive errors
4. Retinopathy of prematurity
5. Congenital glaucoma
6. Others
   - Optic nerve disease
The WHO estimates that there are about 19 million visually impaired children in the world and 1.4 million who are irreversibly blind. However, this is almost certainly a serious underestimation of the problem for several reasons:

- Many blinding conditions like xerophthalmia are associated with a high mortality.
- Some statistics are obtained from blind schools. These often underestimate the burden as many blind children never even get to school.
- Children with multiple handicaps, such as blindness and learning delay are often not recorded either, as they may never leave their remote village.
• In low-income countries with high under-5 mortality rates, the prevalence may be as high as 1.5 per 1000 children, while in high-income countries with low under-5 mortality rates, the prevalence is around 0.3 per 1000 children

Prevalence in Ghana
• 0.75/1000 children

Magnitude in Ghana
• 8,700 blind children
• 70% of causes → avoidable
Avoidable blindness

Preventable Causes

- Corneal scarring
- Intrauterine factors: Rubella, Toxoplasmosis, teratogens (alcohol).
- Perinatal factors: ROP, Birth hypoxia.
- Hereditary diseases
  - Risk counselling for dominant diseases

Treatable Causes

- Cataract
- Glaucoma
- Retinopathy of prematurity
- Uveitis
- Corneal disease (corneal ulcers and opacity)
ALL 3 CHILDREN IN THE FAMILY HAVE SEVERE VISUAL IMPAIRMENT
COMMON CHILDHOOD EYE DISORDERS
AMBLYOPIA: DEFINITION

- Abnormal visual development
- Decreased best-corrected visual acuity
- Unilateral or bilateral
- Apparently normal physical exam, but may also result from recognizable structural abnormalities
Learning to see

- It is easy to think we see with our eyes. In fact, we see with our brain.
Amblyopia (Lazy Eye)

- If one eye is working properly, but the other eye is not, the brain will learn to pay attention to the good eye and ignore the eye that is not working properly.
- This condition is known as Amblyopia or “Lazy Eye.” In fact, it is not the eye that is lazy, but the part of the brain, where this eye sends its image, that is not developing and functioning properly.
- Amblyopia can be caused by any eye problem that blocks or distorts the light passing from the eye to the brain.
- This includes: ptosis, pterygium, cataract, glaucoma, refractive errors that are more severe in one eye, and squint (strabismus).
Why treat amblyopia?

- First, it is important to treat the problem that is causing the eye not to see properly.
- Then, it is necessary to teach the brain, on the affected side, to see properly. If this is not done, the brain may never learn to see and the child may never have good vision on the affected side.
AMBLYOPIA: EARLY DETECTION

- Assess red reflex
- Determine visual acuity
- Evaluate ocular alignment
The ophthalmologist treats amblyopia, but the primary care physician detects amblyopia.
Predisposing factors
SOS:
Spectacles, Occlusion, Strabismus

• Refractive error - very high error in both eyes or unequal between the two (anisometropia)
• Poor media clarity: cataract, lid ptosis
• Strabismus: Misaligned eyes
Poor Focus One Eye
Poor Focus Both Eyes

High refractive error in both eyes
Poor Aim
Amblyopia treatment

- Clear the media
- Spectacles: focus the image
- Patching the good eye: force use of the poor eye
OCCLUSION THERAPY: PURPOSE

- Improves visual acuity
- Does not eliminate strabismus
ATROPINE PENALIZATION THERAPY

- Morning drop of atropine 1% in non-amblyopic eye
- Atropinized eye cannot accommodate for near vision
- Child can still use better-seeing eye for distance
- Child switches fixation at near to amblyopic eye
Strabismus

- Definition: misalignment of the eyes
- Literally means “to squint or look obliquely”
- Affects 4% of children < 6 years old
- 30-50% of cases result in amblyopia (vision loss)
- Can also have detrimental psychological impact
Strabismus Testing

• Corneal light reflex testing

• Cover testing:
  • Requires cooperation from the child and fairly good vision in each eye
  • Cover-uncover test: as the child looks at a distant object, the examiner covers one eye and watches for movement in the other eye
  • Alternate cover test: cover and uncover each eye, looking for eye movement when the cover is shifted to the other eye
Corneal light reflex testing
Pseudostrabismus
Cover-uncover test or alternate cover test shows movement of an eye when fixating on a single target.

When the left eye is covered, the right eye moves outward to pick up the fixation.
(Esotropia, OD)
Vertical strabismus (less common)

Hypertropia

Hypotropia
Head position variability

Eyes appear aligned with head tilt and turn.

Misalignment revealed when head is straightened.
Strabismus: special considerations

- Sudden onset is not uncommon with benign forms of strabismus
- New onset of nystagmus or cranial neuropathy should be referred immediately
- Diplopia is not a feature of strabismus in children as they suppress an image.
- If present may mean serious disorder-refer promptly
Cataracts

- Opacity of the crystalline lens of the eye
- One third hereditary, one third associated with other disorders, one third idiopathic
- If congenital, must remove in first three months or the amblyopia is irreversible
A white pupil is a sign of serious eye disease. All children with a white pupil should be referred to an eye specialist immediately!
Congenital Cataracts

- Unilateral develops amblyopia more rapidly than bilateral
- Bilateral should have systemic work-up
Congenital cataracts
Congenital ptosis

- Bilateral cases may have chin up position to see.
- Unilateral cases more worrisome for amblyopia development
Congenital glaucoma

The eyes look big, with the corneas blurry and gray.
Orbital Tumors

- Can closely resemble orbital cellulitis

- Signs:
  - Proptosis
  - Impairment of eye movement
  - Possibly a palpable mass
  - Ptosis, optic atrophy, vision loss
  - Bruit, visible pulsation of globe in vascular tumors

- Evaluation: ultrasound, MRI, CT

- Benign
  - Most are vascular (primarily hemangiomas)
  - Dermoid tumors

- Malignant
  - Rhabdomyosarcoma, lymphomas, metastatic neuroblastoma
  - Also optic gliomas and retinoblastomas that extend into the orbit
Retinoblastoma

- Most common malignant ocular tumor of childhood
- Unilateral or bilateral
- Onset generally before 4 years of age
- Most present with leukocoria (white pupil reflex)
- 25% present with strabismus
- Tumor generally outgrows vascular supply and necrotic areas calcify
- Get CT scan to look for calcifications
- Defect is in tumor suppressor gene
- Treatment with radiation, chemo, cryo, laser, enucleation
Leukocoria

Decreased Vision → Retinoblastoma?
Retinoblastoma
Retinoblastoma

Early
Late
Very late
The Red Eye

- Blepharitis
- Nasolacrimal duct obstruction
- Chalazion
- Preseptal and Orbital Cellulitis
Blepharitis
Nasolacrimal duct obstruction

- Present in about 5% of newborns
- 90% will spontaneously resolve by 12 months
- Due to obstruction in the nose
- 90-95% success rate with nasolacrimal probe
Nasolacrimal Duct
Dacryocystitis

- A possible complication of dacryostenosis
- More common in neonates
- Infection of the nasolacrimal sac
- Red, edematous sac area
- Possibly fever, irritability
- Tx: massage, antibiotics
Dacryocystitis
**Chalazion or hordeolum**

- Obstruction of meibomian glands
- Hordeolum has injection of blocked gland
- Chalazion is more chronic Granulomatous inflammation
- They may coexist
- Hygiene, antibiotics and hot compresses
- Role of systemic antibiotics in acute infection with significant preseptal swelling
Preseptal Cellulitis

- Skin infection around the eye
- Anterior to the orbital septum, an anatomic landmark
Orbital cellulitis

- Infection posterior to anatomic septum
- Presents fever, orbital swelling, redness, pain with extraocular movements, headache
- Often associated with sinus disease
- Can progress to sepsis, secondary meningitis if untreated
Conjunctivitis

- Neonatal Conjunctivitis/Ophthalmia neonatorum
- Allergic conjunctivitis
- Bacterial and viral conjunctivitis
Neonatal Conjunctivitis

- **Causes:**
  - Neisseria gonorrhoeae
  - Chlamydia trachomatis
  - Chemical
  - Hemophilus influenzae
  - Staph aureus
  - Strep pneumoniae
  - Strep viridans
  - Herpes simplex
Neonatal Conjunctivitis/Ophthalmia Neonatorum

- **Chlamydia trachomatis:**
  - Usually presents 5-14 days after birth
  - Usually infected when passing through birth canal
  - At risk for Chlamydia pneumonia

- **Neisseria gonorrhoeae:**
  - Often presents 2-4 days after birth
  - Hyperacute, copious purulent exudate
  - Corneal perforation in up to 10%
Allergic Conjunctivitis

- Allergic often associated with chemosis and itching, FBS
- Watery discharge
- Often papillae
**Subconjunctival hemorrhage**
- Usually painless, but frightened patients may describe irritation
- Secondary to cough or valsalva
- Check platelets, coags if recurrent
- Can be traumatic (kids)
- Resolves spontaneously in 2 weeks
PEDIATRIC EYE SCREENING
Pediatric Eye Screening

• History:
  • symptoms/signs/worries
  • Premature? Normal pregnancy?
  • Normal delivery/development
  • Family history

• Vision- Infants toddlers- central steady and maintained? young children- pictures, Tumbling E, HOVT

• Red Reflex: Equal? White reflex (leukocoria)?

• Eyes aligned: Equal corneal light reflex? Cover-uncover test
Screening Importance

• Early detection may treat and reverse some visual loss and prevent permanent visual loss (amblyopia)
• Life threatening disorders may detected (retinoblastoma)
• Improving vision early even with glasses alone helps functioning
• Screening is the primary care providers responsibility
Symptoms/signs such as eye rubbing
Parental Concern: Leukocoria

Parental Concern: Enlarged Cornea
Asymmetric Red Reflex
Screening tools
Light reflex is not in same place in both corneas
Equal red reflex and corneal light reflex
Vision testing in children
Acuity in Infancy

- Reaction to light
- Simply a pupil response or lid closure. Used when normal vision is expected in very young infants
NORMAL INFANT VISION

- Good visual function
- Fixate and follow with each eye
- Steady fixation
Acuity in Preverbal Child

- Fix and follow
- Looking at target or face and following it
- Usually begins at 2 months of age
- Can fool you with unilateral visual problems
Verbal Visual Acuity

- Allen
- Tumbling E
- Snellen
Ophthalmologist can quantify refractive error in infants.

Children usually do well with eyeglasses.
<table>
<thead>
<tr>
<th>Condition</th>
<th>Prevention</th>
<th>Treatment</th>
</tr>
</thead>
<tbody>
<tr>
<td>Genetic disorders</td>
<td>Genetic counselling. Education about the risks of marrying close relatives</td>
<td>Very few genetic diseases can be treated</td>
</tr>
<tr>
<td>Congenital rubella*</td>
<td>Maternal vaccination</td>
<td>None</td>
</tr>
<tr>
<td>Ophthalmia neonatorum*</td>
<td>Good antenatal care and treatment of maternal genital infection</td>
<td>Urgent treatment of neonates with purulent conjunctivitis</td>
</tr>
<tr>
<td>Retinopathy of prematurity</td>
<td>High quality care of premature infants</td>
<td>Retinal laser or cryotherapy. Possibly intravitreal anti-VEGF injection</td>
</tr>
<tr>
<td>Congenital cataract</td>
<td>Not preventable except from rubella</td>
<td>Cataract surgery and specialist paediatric ophthalmology care</td>
</tr>
<tr>
<td>Congenital glaucoma</td>
<td>Not preventable</td>
<td>Glaucoma surgery and specialist paediatric ophthalmology care</td>
</tr>
<tr>
<td>Condition</td>
<td>Treatment</td>
<td>Notes</td>
</tr>
<tr>
<td>---------------------------------</td>
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</tr>
<tr>
<td>Xerophthalmia*</td>
<td>Vitamin A supplements Nutritional advice</td>
<td>Vitamin A supplements</td>
</tr>
<tr>
<td>Measles*</td>
<td>Vaccination</td>
<td>Vitamin A supplements will minimise the extent of measles corneal ulcers if vitamin A deficiency also present</td>
</tr>
<tr>
<td>Toxic traditional eye medication (TEM)</td>
<td>Education</td>
<td>Thorough wash out with water or saline if the child is seen soon after the TEM has been used</td>
</tr>
<tr>
<td>Amblyopia*</td>
<td>Treat causative pathology, e.g. cataract, ptosis, large chalazions</td>
<td>Patching or atropine for better eye</td>
</tr>
<tr>
<td>Refractive error*</td>
<td>Not possible</td>
<td>Carefully prescribed glasses</td>
</tr>
</tbody>
</table>

* Should be fairly easily treated and it is a great failing of governance and health systems if they are not.
HELPING TO KEEP OUR CHILDREN’S EYES HEALTHY FOR THEY ARE OUR FUTURE
REFERENCES

- Eye Diseases in Hot Climates, 5th Edition, Saul Rajak, John Sandford-Smith
- BCSC, Pediatric Ophthalmology and Strabismus, American Academy Of Ophthalmology, 2019
- Previous Lecture to ELWA Residents, Pediatric Ophthalmology for the Family Physician
Thank You