Pediatric Ophthalmology Update

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Outline

- Strabismus and Binocular Vision
- Tearing in Infancy
- The Differential Diagnosis of the White Pupil
  - Retinoblastoma
  - Conjunctivitis
  - Childhood Cataracts

Strabismus and Binocular Vision
Strabismus

- Strabismus is defined as any form of ocular misalignment or tendency toward misalignment
- Also called squint
- Occurs in 4% of children in the U.S.A.
- Occurs in 1% of adults
- One of the leading causes of amblyopia

Pseudo-Strabismus

- Very common
- Wide nasal bridge
- Epicanthal folds
- Look for symmetry of light reflex in pupils
  - Hirshberg test
  - Bruchner test
- Confirm using cover test
- If in doubt refer to ophthalmologist

Binocular Vision

- The eyes are aligned on the object.
- Images fall on the foveas
- The brain merges the images allowing the perception of depth
- In strabismus there is suppression of part of the visual field under binocular conditions
- This leads to under-stimulation of the visual cortex and to amblyopia
Types of Strabismus

- **Phoria**
  - Suffix meaning the deviation is latent
  - Underlying tendency for the eyes to cross

- **Tropia**
  - Suffix meaning the deviation is constant
  - The deviation will be observable

- **Intermittent Tropia**

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Treatment of Strabismus

- **Goals**
  - Enable or restore binocularity in children
  - Eliminate diplopia and restore fusion and normal ocular movements in adults
  - Eliminate abnormal head positions
  - Correct errors of refraction
  - Treat amblyopia in children
  - Realign visual axes using prisms or surgery
Tearing in Infancy

Anatomy of the Lacrimal System

Nasolacrimal Duct Obstruction

- 2% - 6% of newborns
- Bilateral in 1/3 of cases
- Tearing and crusting of one or both eyes starting at 2 - 3 weeks of age
- Superimposed conjunctivitis
- Etiology
  - Developmental 61%
  - Infectious 24%
  - Trauma 12%
  - Dysfunction 3%
**Tearing in Infancy**

- NLD obstruction
  - Most common
- Punctal agenesis
- Dacryocystocele
  - Rare
- Fistula of lacrimal sac onto skin
- Canalicular laceration
- **Congenital glaucoma**
  - Do not miss

**NLD Obstruction Management**

- 80% - 90% resolve spontaneously or with non-surgical management in the first year of life
- **Medical**
  - Crigler massage – **IMPORTANCE OF PROPER TECHNIQUE**
  - Antibiotic ointment **ONLY** with conjunctivitis
- **Surgical**
  - Probing +/- repeat +/- infracture of inferior turbinate
  - Probing with silicone intubation
  - Dacryocystorhinostomy

**Leukocoria**

The Differential Diagnosis of Retinoblastoma
Leukocoria
White Pupil

- The differential diagnosis of Retinoblastoma
- The abnormal pupillary light reflex
  - White or tan
  - Depends on lighting conditions
  - Depends on direction of gaze

Retinoblastoma

- 1/13,000 live births
- 25% Bilateral : 75% Unilateral
- 40% Heritable (Germline)
- 60% Non-Heritable (Sporadic)

- Age at diagnosis
  - <12 months in bilateral cases
  - >15 months in unilateral cases
  - 95% of cases diagnosed before age 5 years
  - 5/760 cases at AFIP were > 10 years old

Retinoblastoma Genetics

- Autosomal dominant trait
- Recessive tumor suppressor gene on 13q14
- Heritable - One germinal and one somatic mutation
  - Bilateral
  - Multifocal
  - Familial - 5%
  - Second tumors
- Sporadic - Two somatic mutations
  - Unilateral unifocal
Retinoblastoma: Clinical Presentation

- Leukocoria
- Strabismus
- Persistent red eye
- Hypopyon – WBCs in the anterior chamber
- Other
- Importance of early referral from pediatrician

Retinoblastoma: Diagnosis

- > 90% diagnosed by ophthalmoscopy, ultrasonography and computed tomography
- The importance of calcifications

Retinoblastoma Differential Diagnosis

- Cataract
- Persistent Hyperplastic Primary Vitreous
  - Small eye
  - Cataract
  - Persistent fetal vasculature
  - Retrolental membrane
- Coats' disease
  - Vascular malformation
Retinoblastoma Differential Diagnosis

- Toxocariasis
- Myelinated nerve fibers
- Coloboma
- Morning Glory Disc Anomaly
- ROP
- Child abuse with vitreous hemorrhage and retinal detachment

Retinoblastoma Treatment

- Cure in 98% of cases if given in a timely fashion
- Enucleation in advanced cases
- Chemotherapy
  - Chemo-reduction
- Focal treatment
  - Cryotherapy
  - Laser
  - Radiocative plaques

Retinoblastoma Second Malignant Neoplasms

- Result from mutations in Rb Gene
- Only in heritable cases
- 25%-30% of patients
- Up to 30 or 40 years later
- Osteosarcomas - majority
- Cutaneous malignant melanoma - 7%
- Role of radiation therapy
Conjunctivitis

- Viral
  - Preauricular lymph node
  - URI
- Bacterial
  - Purulent discharge
- Allergic
  - Itching
  - Other allergies
- Chlamydia
  - Neonatal
  - Persistent or recurrent
- Toxic/Chemical
  - History
- Immune
  - Subconjunctival scarring

Conjunctivitis Treatment

- Bacterial: Antibiotic drops or unguent
- Viral: Observe +/- antibiotic drops
- Chlamydia: Systemic antibiotic
- Gonococcal: Systemic antibiotics
- Allergic: Cold compresses; anti-allergy antihistamines
- Chemical: Irrigate; steroids
- Immune: Steroid drops
**Ophthalmia Neonatorum**

- First day: Chemical
- 2-4 days: Gonococcal
- Any other time: Bacterial
- Chlamydia: up to 40% - 70% of cases
- Pseudomonas: Very aggressive

**Ophthalmia Neonatorum Treatment**

- Prophylaxis
  - Tetracycline or erythromycin ointment
- Isolate infant
- Gram stain and cultures
- Identify source and carriers
- Antibiotics
  - Topical
  - Systemic

**Childhood Cataracts**
Cataracts in Infants and Children

- Concerns
  - Amblyopia
  - Sign of systemic disease
  - Associated ocular abnormalities
- Management
  - Determine significance
  - Extract early
  - Manage refractive error and amblyopia

Cataracts in Systemic Disease

- Myotonic Dystrophy
- Galactosemia
- Wilson Disease
- Fabry Disease
- Mysotonic Dystrophy

Infantile Cataracts Work-up

- Examine both parents for lens opacities
- IgG and IgM titers
  - Toxoplasmosis
  - Rubella
  - CMV
  - HSV
  - Varicella
- VDRL on mother and baby
Infantile Cataracts
Work-up

- Urine for reducing substances after a milk feeding or assay for GALT, Gal-1-P and Galactokinase
- Ca and P to r/o hypoparathyroidism
- Ferritin levels
- Cholesterol and dehydroxycholesterol levels

Galactosemia

Infantile Cataracts
Work-up

- Mevalonic acid in blood and urine
  - mevalonic aciduria
- Urine amino acid levels, especially in the presence of glaucoma
  - Lowe syndrome
- G6PD levels, especially in the presence of anemia and jaundice
- Consider trauma

Thank You
Question 1

- NLD obstruction can be differentiated from congenital glaucoma by
  - The presence of lacrimation in NLD but not glaucoma
  - The occurrence of NLD obstruction in younger and glaucoma in older infants
  - Clouding and enlargement of the cornea in glaucoma but not NLD obstruction
  - Mode of inheritance

Question 2

- The differential diagnosis of a white pupillary reflex includes all of the following except
  - retinoblastoma
  - Toxocariasis
  - Coats’ disease
  - Optic nerve glioma
  - Persistent Hyperplastic Primary Vitreous

Answer 1

- NLD obstruction can be differentiated from congenital glaucoma by
  - Clouding and enlargement of the cornea in glaucoma but not NLD obstruction
Answer 2

- An optic nerve glioma does not cause a white reflex in the pupil. It is located in the retrobulbar part of the optic nerve.