The Imposter: Recognizing ocular disease in kids
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Disclosure Statement:
• Nothing to disclose

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Challenges diagnosing disease in primary care settings
• Broad range of problems present
• Decisions in general practice different from those in specialist settings
• Many symptoms - not specific for unique condition
• Diagnostic labels - less important than determining course of action
• Diagnoses often involve two option decisions
  – serious vs. benign
  – common vs. rare
  – stable vs. progressive
  – treat vs. don’t treat
  – refer vs. don’t refer


Specific challenges in diagnosing kids
• History
  – Age, emotional and cognitive development vary
  – Complexity of the family unit
  – Chief complaints/symptoms – history may evolve or be minimally available
• Testing
  – Children are moving targets
  – Limited time before your child fatigues or resists
  – Diagnostic test selection – can kids do the test?

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Case
• CC 10 yo black male; no complaints
• Ocular Hx, Med Hx unremarkable
• Meds, Allergies none
• Refraction VA
  OD -3.00-0.50 x 89 20/20
  OS -2.00 sph 20/20
• Cover Test c Rx
  – Orthophoria at distance
  – 4 exophoria at near
• Visual Field by confrontation normal OD, OS
• Pupils 6 mm OU RRL-APD
• SLX unremarkable
• IOP OD 14 mm Hg OS 15 mm Hg

EVERYTHING NORMAL ON EXAM UP TO FUNDUS EXAM

Fundus
• Disc Margins - ill-defined
• Tortuous vessels
• No Spontaneous Venous Pulsation seen

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Differential Diagnosis

Optic disc drusen
- Buried
- Visible

Disc Swelling/Papilledema
Grade I
Grade IV

Fundus Evaluation

<table>
<thead>
<tr>
<th></th>
<th>ODE</th>
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<tbody>
<tr>
<td>Disc</td>
<td>Distinct margins?</td>
<td>Pushed from behind?</td>
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<td>Color</td>
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<td>Blood Vessels</td>
<td>Obscured at NRR?</td>
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<td>Splinter Hemes?</td>
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Pediatric Use of OCT

- Underutilized in dx pediatric ocular dz
  - Many ped ocular dz extramacular
  - Need for refinement
  - Spectral-Domain OCT - 50-100x faster than Stratus OCT; eye tracking technology

- Pediatric norms for ONH analysis – status
  - Some studies; variety of OCTs, populations
  - Used to measure peripapillary RNFL

Diagnostic Testing in Kids

- Optical Coherence Tomography (OCT)
- B-scan
- Visual Field
- Autofluorescence

STRENGTHS AND LIMITATIONS IN KIDS?

OCT

- Optic Disc Drusen (ODD)
  - elevated ONH
  - “lumpy-bumpy” internal contour
  - abrupt end hyporeflective space between sensory retina and RPE

- Optic Disc Edema (ODE)
  - elevated ONH with smooth internal contour
  - subretinal hyporeflective space - Lazy V pattern
  - Mean RNFL thickness >all quadrants than in ODD
  - Nasal RNFL thickness >78.0 mm - 80.0% sensitivity and 88.9% specificity for ODE

**Can OCT distinguish Optic Disc Drusen from Optic Disc Edema?**

- **YES**
  - Stratus OCT images of 20 ONHD, 20 ODE, 20 controls
  - Qualitative, quantitative (nasal RNFL)
- **NO**
  - compared 16 buried ONHD to 9 eyes to 12 equal to Frisén grade 2 papilledema
  - no difference in RNFL thickness
  - not clinically reliable; 50-64% accuracy


**B scan**

- B scan
- calcification on the optic nerve head
- not as readily and widely available in all clinical settings as OCT

**Summary**

- Small disc; Disc margins?
- Vessels – tortuous, but distinct at disc margin;
  - (--) Hemes;
  - (--) HVF

**Visual Field**

- Pediatric Use > 8 years
- Visual Field
  - Normal
  - ODD
  - ODE


**Case 2**

- 8-year old male seeks care visiting from Latin America
- CC: blurred vision OD, OS; can’t see blackboard at school
- Ocular Hx, Family Hx: unremarkable
- Pupils, Confrontation fields, EOMs: within normal limits
- Cover Test
  - Distance: ortho (without correction)
  - Near: 8 exophoria (without correction)
- Color Vision: *HRR Color Vision Test*
  - OD, OS: fail
Case 2

Refraction  Best corrected VA
OD  -0.25 -2.75 x 005  20/100
OS  +0.50 -3.00 x 180  20/100

Additional Hx
Patient and parent say that he wears his glasses, current pair is one month old; VA hasn’t improved over one last month

Visual Fields
- Goldmann visual fields performed
  - Two isopters - II4e for periphery and I3e for central, OD and OS
  - Fixation losses – WNL
  - Peripheral vision normal
  - Centrally-small relative defect inferior central OD
- Amsler Grid Test

OCT
- Use in pediatric dx vitreoretinal diseases
- Clearer understanding – differences within retina, on retinal surface, bt retina and vitreous
- Produces and analyze quantitative data
  - Reflective interfaces bt layers, change in optical density

OCT Findings
Dif dx: Stargardt’s, cone, cone rod dystrophy

Other testing
- Electrodiagnostics Services
  - fERG – intact rod population
  - mfERG – diffuse loss central cone OU
- Results of fluorescein angiography and fundus autofluorescence
  Diagnosis Stargardt’s disease; Visual Impairment
Does amblyopia contribute to the reduced VA?
• OCT - link of structural differences and amblyopia
  – Strabismic amblyopes - no difference in RNFL thickness in amblyopic eyes compared to fellow eyes
  – Anisometropic amblyopia w/ unilateral high myopia - thicker fovea, thinner inner and outer macula in amblyopic eye than fellow eye
More studies are needed to macular changes differences in high myopia, amblyopia, or combination of both
Link to treatment results?
• Some amblyopes who failed to achieve normal VA after tx showed thickened RNFL in macular area with no fovea

Amblyopia with ocular disease – what’s to be gained by tx?
• 3 children with severe pathologies
  – Unilateral macular scar secondary to retinoblastoma ->20/25
  – Severe optic nerve atrophy secondary to hemangioma -> 20/40
  – Unilateral large optic nerve coloboma-> XT shifted unilateral to alternating

Case 3 – Candy Crush
• 15 BM came in after being hit in the eye at school.....
• VA sc 20/20 OD/OS/OU
• EOM, CF, Pupils unremarkable
• Ocular Health Evaluation
  – OD: Unremarkable
  – OS: 1-2+ injection, hyperemia, 1+ cell, posterior segment health unremarkable

Case Presentation – Is it Pink Eye?
• 5 BF presents for “pink eye” OD
• She has been treated approximately 4 times in the past year for conjunctivitis
VA
  – OD: 20/200
  – OS: 20/25
Pupils – Poorly reactive OD>OS
EOMs and Confrontation Fields - Unremarkable
Considerations in Diagnosis

- Children often do not complain of symptoms
- Examination may be difficult
- History – how long has the patient had symptoms and/or signs?
- Unilateral or bilateral
- Anterior only?
  - Granulomatous or Non granulomatous

Causes of Pediatric Uveitis—Systemic

Granulomatous

- Sarcoidosis
- Syphilis
- Herpetic Uveitis
- Lyme Disease
- Tuberculosis
- Inflammatory Bowel Disease
- Vogt-Koyanagi-Harada

Causes of Pediatric Uveitis—Systemic

Non Granulomatous

- Juvenile Idiopathic Arthritis
- Lupus
- Behcet’s disease
- HIV
- Lyme Disease
- Leukemia
- Herpetic Uveitis
- Kawasaki Disease

Causes of Pediatric Uveitis—Non-Systemic

- Idiopathic
- Traumatic
- Fuch’s Heterochromic Cyclitis

Masquerade Syndromes

- Mimic uveitis or cause uveitis
  - Retinoblastoma
  - Leukemia
  - Lymphoma
  - Juvenile Xanthogranuloma

Diagnostic Testing

- Ocular Evaluation
- Systemic Evaluation
  - Bloodwork
    - CBC, ESR, HLA-B27, ACE, ANA, VDRL, FTA-ABS
  - Chest X-Ray
  - PPD skin test
Management

- Cycloplegic Agent
- Topical, Injected and Oral Corticosteroids
- Treat any underlying bacterial or viral condition
- NSAIDS
- Immunomodulatory agents

Complications of Pediatric Uveitis

- Amblyopia
- Cataract
- Glaucoma
- Retinal Detachment
- Band Keratopathy
- Macular Edema

Management

- Treat any underlying bacterial or viral condition
- Cycloplegic Agent
- Topical, Injected and Oral Corticosteroids
- NSAIDS
- Immunomodulatory agents
- Biologic Agents

Case 4

Blurry vision at near....

- 9 yo BM, complains of blur at near
  - Wearing his grandmother’s glasses helps

Medical history
- Anemia per grandmother

Ocular history
- Unremarkable; first eye exam

Family medical history
- Glaucoma (great grandfather)
- DM and HTN (aunts and uncles)

Examination

VA 20/20 OD, OS, OU at both distance and near
Pupils, EOM, CF unremarkable
Anterior Segment Health Unremarkable
Posterior Segment See photo
1 Year Later……

- Patient returned after failing a school screening
- VA 20/20 at D and N with +0.12 sph OU
- Preliminary testing unremarkable
- Anterior Segment health unremarkable
- Posterior Segment see photos

More History…….

- Mother has sickle cell anemia
- Patient may have the sickle cell trait…. 
Sickle Cell Disease

- Point mutation in Beta Hb chain
  - 6th aa changes from glutamic acid to valine
- HbSS
- HbSC**
  - Ocular manifestations are more common
- HbSThal
- HbAS

Systemic Complications

- Stroke
  - Intracranial ICA and MCA
- Acute Chest Syndrome
- Dactylitis
- Pulmonary Hypertension
- Renal Disease
  - Splenic sequestration
  - Pain crisis

Ocular Complications

- Sickled blood cells occlude small vessels
- Proliferative vs. Non proliferative Retinopathy
  - Non proliferative – Salmon patch hemorrhages, black sunbursts
  - Proliferative – Sea fan neovascularization
- Central Retinal Artery or Arteriole Occlusion
- Angioid Streaks
- Orbital Compression Syndrome
- Comma shaped conjunctival vessels

Diagnostic Testing

- Blood work to determine quantity of HbS
- Mutation analysis

Recommendations for Ocular Health Evaluation

- Age 9-10
  - 9 for SC, 13 for SS
  - Eye exam every 2 years
  - FA if suspect proliferative disease
- Over age 20
  - Eye examination yearly
- More frequent monitoring...
  - Pain crisis
  - Splenic sequestration
  - Pulmonary hypertension

Management Considerations

- Hydroxyurea Therapy
- Red blood cell transfusion
- Yearly testing
  - CXR, liver, pulmonary, renal function, CBC/reticulocyte count, urinalysis, abdominal ultrasound
- Transcranial Doppler studies of arterial blood flow velocity
- Avoid circumstances that cause sickling
- Retinopathy
  - Laser photocoagulation
  - Cryotherapy
  - Observation
Summary

• Treatment of pediatric ocular disease presents unique challenges not encountered in adult patients.
• Spectral domain OCT
  – noninvasive and fast
  – assists in pediatric diagnosis of ocular diseases, but more research on pediatric norms is needed
• Ocular health evaluation can reveal serious systemic and ocular conditions even in children presenting without symptoms.

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