The Imposter: Recognizing ocular disease in kids
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Disclosure Slide
• Coulter – none
• Jenewein - Research support/salary support
  – Nevakar, Inc.
  – NIH

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

Specific Challenges in Diagnosing Ocular Disease in Kids
• History
  – Age, emotional and cognitive development vary
  – Complexity of the family unit
  – Chief complaints/symptoms – history may evolve or be minimally available

Specific Challenges in Diagnosing Ocular Disease in Kids
• Testing
  – Children are MOVING targets
  – Limited time before your child fatigues or resists
  – Diagnostic test selection – can kids do the test?
    • Visual Acuity
    • Ocular Health
    • OCT
    • Visual Field
Managing Pediatric Patients

- After diagnosis is made...
  - Choose the best management plan
    - Routine care/monitoring
    - Additional testing
    - Referral for specialty care
    - Urgent referral

Optic Nerve Anomalies

Case 1

- **CC:** 10 yo black male; no complaints
- **Ocular Hx, Med Hx:** unremarkable
- **Meds, Allergies:** none
- **Refraction**
  - OD -3.00-0.50 x 89 20/20
  - OS -2.50 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

Fundus

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

Differential Diagnosis

**Optic disc drusen**
- Buried
- Visible

**Disc Swelling/Papilledema**
- Grade I
- Grade IV

Fundus Evaluation

<table>
<thead>
<tr>
<th>ODE</th>
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<tbody>
<tr>
<td>Disc</td>
<td>Distinct margins?</td>
</tr>
<tr>
<td></td>
<td>Hyperemic?</td>
</tr>
<tr>
<td>Color</td>
<td>Yellow?</td>
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<tr>
<td>Blood Vessels</td>
<td>Obscured at NRR?</td>
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<tr>
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<td>Splinter Hemes?</td>
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<td>SVP absent?</td>
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Spontaneous Venous Pulsation (SVP)

- Present in 87.6% of 146 unselected subjects 20 to 90 years of age and absent in 100% of 33 patients with raised intracranial pressure without papilledema and ten patients with papilledema.
- Caused by variation in the pressure gradient along the retinal vein as it traverses the lamina cribrosa.
- Cessation of the spontaneous venous pulsation is a sensitive marker of raised intracranial pressure.
- Best viewed with Direct Ophthalmoscopy

Diagnostic Testing in Kids

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

STRENGTHS AND LIMITATIONS IN KIDS?

Optic Disc Drusen - OCT

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

Optic Disc Edema - OCT

- 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

Pediatric Use of OCT

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

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STRENGTHS AND LIMITATIONS IN KIDS?
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)
    - SHYPS, alpha angle

- **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


[S. Lee et al. Differentiation of ONHD and ODE with SD OCT. Ophthalmology. 2011;18:5.]

[Ultrasonography – B scan]

<table>
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<th>Edema</th>
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<tr>
<td>Elevation</td>
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<tr>
<td>High reflectivity</td>
<td>Enlarged ON sheath diameter; Crescent sign</td>
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[Crescent Sign]

- 50 patients diagnosed with papilledema on MRI
  - 92% had crescent sign

[Bhosale et al. World Journal of Methodology. 2017;7(3)]

[Ultrasonography – 30 Degree Test]

- A-scan
- Measure width of optic nerve in primary gaze and 30 degrees in abduction
- Increased ICP
  - Smaller measurement at 30 degrees
    - Nerve and sheath are stretched
    - Subarachnoid fluid evenly distributed over nerve

[Fluorescein Angiography]

- Papilledema
  - Leakage of ON
- Optic Disc Drusen
  - Buried drusen – no hyperfluorescence
  - Superficial drusen – nodular staining

- 97% accurate for differentiating between ODD and ODE


Chang et al. Ophthalmology, 2017]
**ONH Drusen vs. Disc Edema**

- ONH Drusen
  - (+) elevated nerve
  - (-) vessel obscuration, (+) SVP
  - (+) pink ONH, (+) distinct margins
  - (+) high reflectivity on B-scan
- Disc Edema
  - (+) elevated nerve
  - (+) vessel obscuration, (-) SVP
  - Enlarged optic nerve on B-scan

**Case #2 – Elevated ONH**

- 16 year old patient
- All previous examinations were **unremarkable**

**Case #3 – Abnormal Nerves and Diplopia**

- 7 year old patient
- Previously normal ocular health
- Normal systemic health
- Complaint of intermittent diplopia

**Summary**

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

**B Scan**

**Diagnosis? ONH Drusen**

**Diagnosis? Papilledema**

**Three Different Cases, Three Different Diagnoses**
Diagnosing ONH Elevation in Children

- Look for signs of ODE
- Look for signs of ONHD
- Accompanying Neurological Signs?
- B-Scan
- OCT
- FAF
- If unsure, neurological work-up

Is This Amblyopia?

Case #1

- 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 #1

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<th>Refraction</th>
<th>Best corrected VA</th>
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<td>OD</td>
<td>-0.25 -2.75 x 005</td>
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<tr>
<td>OS</td>
<td>+0.50 -3.00 x 180</td>
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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

What is Amblyopia?

- Unilateral or bilateral condition
- BCVA is poorer than 20/20
- No structural or pathologic anomalies
- One or more of the following conditions occurring before 6-8 years of age:
  - Significant refractive error
  - Constant, unilateral strabismus
  - Form vision deprivation
What is **Significant** Refractive Error?

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<th>Anisometropic</th>
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**Tarczy-Hornoch et al. (2013) Ophthalmology. 120:1220-1226.**

**Form Deprivation**
- 0.1% of the general population*
- Severe amblyopia
- Obstruction of the line of sight
  - Prolonged blepharospasm/ptosis
  - Corneal opacity
  - Hyphema
  - Cataract
  - Vitreous opacity


**Is It Really Amblyopia??**
- Amblyogenic factor must be present
- Must rule-out any underlying ocular or neurological pathology that may explain a decrease in VA
- Ocular pathology may co-exist with amblyopia

**Normal Findings in Patients with Amblyopia**
- Pupils
- Amsler Grid
- Visual Field
- Color Vision

If any of these are abnormal, carefully consider your diagnosis!!!!!

**Back to Our Patient....**
- Fundus evaluation - mild thinning of the macula with associated granular deposits OU

Abnormal macular findings; High astigmatism OU
Visual Fields

- Goldmann visual fields performed
  - Two isopters: I14e 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

- Stargardt’s disease
- 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

Is this Amblyopia?

- 16 HM c/o of blurry vision OD for “many years”
- History significant only for possible corneal abrasion > 8-9 years ago
- First eye exam

Is this Amblyopia?

- DVA (sc)
  - OD: 20/200 at 2 feet
  - OS: 20/100
- Retinoscopy
  - OD: +6.50-2.50x010 (20/300)
  - OS: -2.50-1.50x180 (20/20)
Is this Amblyopia?

- Color Vision
  - OD: Fail / OS: Pass

- Pupils
  - OD: PERRL(+APD) / OS: PERRL(-)APD

- Red Cap Desaturation
  - Patient reports cap is “brown” OD and red OS

- Amsler Grid
  - Patient notes part of the grid is missing OD

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 Presentation

- 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

Pediatric Uveitis
Case Presentation

- Anterior Segment Health
  - **Adnexa:** Mild periorbital swelling OD
  - **L/L:** Clean and clear
  - **Conjunctiva:** 1+ injection OD, white and quiet OS
  - **Cornea:**
    - OD: 3-4+ edema, with muttonfat KPs inferiorly, inferior pannus
    - OS: Inferior pannus
  - **Anterior Chamber:**
    - OD: 3+ Cell/Flare, granuloma inferiorly
    - OS: Trace cell
  - **Iris:**
    - OD: Posterior synechiae 360 degrees
    - OS: Posterior synechiae at 3, 6 and 11 o’clock

Pediatric Uveitis

- Uncommon but potentially blinding condition
- Higher rate of complications and vision loss in children than in adults
  - severe vision loss in 25% to 30% of pediatric uveitis patients
- Diagnosis requires a good history, a thorough review of systems, a complete examination, and a focused laboratory workup

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
- Need to distinguish between *infectious* and *non-infectious* causes -> treatment pathways differ

<table>
<thead>
<tr>
<th>Infectious</th>
<th>Non-Infectious Causes</th>
</tr>
</thead>
<tbody>
<tr>
<td>Toxoplasmosis</td>
<td>Juvenile Idiopathic Arthritis</td>
</tr>
<tr>
<td>Toxocariasis</td>
<td>Blau syndrome</td>
</tr>
<tr>
<td>Herpes Simplex Virus</td>
<td>Tubulointerstitial Nephritis</td>
</tr>
<tr>
<td>Varicella Zoster Virus</td>
<td>Sarcoidosis</td>
</tr>
<tr>
<td>HIV</td>
<td>Behcet disease</td>
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<tr>
<td>Cytomegaloivirus</td>
<td>Pars planitis</td>
</tr>
<tr>
<td>Rubella Syphilis</td>
<td>Vogt-Koyanagi-Harada</td>
</tr>
</tbody>
</table>

### History
- Review of Systems
- Previous infectious exposures
  - Pets?
  - Travel?
  - Camping, hiking?
  - Food?

### Previous Exposure – Other symptoms
- Bulls eye pattern rash

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

### Juvenile Idiopathic Arthritis
- Most common systemic association of pediatric uveitis

#### Questions for History
- Arthritis present for at least 6 weeks?
- Onset associated with morning stiffness and arthralgia during day?
- Complaints of joint pain?
- History of school absences or limited ability to participate in physical education classes?
- Spiking fevers occurring at about the same time of day?
- Dermatologic symptoms:
  - Rash on the trunk and extremities
  - Psoriasis or more subtle dermatologic manifestations
Complications of Pediatric Uveitis

• Cataract
• Glaucoma
• Amblyopia
• Retinal Detachment
• Band Keratopathy

Management

• Early aggressive treatment improves outcomes
• Cycloplegic Agent
• Treat any underlying bacterial or viral condition

Management

• Topical, Injected and Oral Corticosteroids
  – Topical therapies may be enough for ant uveitis
  – Post, Pan uveitis often requires stronger meds
    (injection, iv implant)
• NSAIDS
• Immunomodulatory agents
• Biologic Agents

Medications Ped Use Approval

• Few medications with approval in pediatric uveitis
• Methotrexate approved for children with JIA
• Adalimumab approved for patients > 4 yrs
• Newer therapies —safety, efficacy studies for children not done

Case Presentation

• 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

Considerations in Traumatic Uveitis

• Trauma leading cause of uveitis in children who do not have JIA*
• Rule out other complications of trauma
  – Orbital Blow-Out Fracture
    • Diplopia?
• Evaluate patient for retinal breaks
• Future evaluation - Gonioscopy

Sickle Cell Anemia

Blury 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

- Who has sickle cell disease?
  - Primarily affects children of African descent and Hispanics of Caribbean ancestry
  - Also occurs in children of Middle-Eastern and Indian descent
- Clinical Picture
  - highly variable
    - 6 episodes a year to rarely occurring
    - Difficult to predict at an early age

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

Sickle Cell Disease

- Mechanism
  - Red blood cells cluster together ->
  - Clusters create blockages in small blood vessels ->
  - Cut off oxygen ->
  - Lead to sudden pain, damage to body tissues and organs over time

Ocular Complications

- Sickled blood cells occlude small vessels
- Proliferative vs. Non proliferative Retinopathy
  - Non proliferative – Salmon patch hemorrhages, black sunbursts
  - Proliferative – Sea fan neovascularization
### Ocular Complications

- 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
  - Identify mutations of HBB (S, C, Thalassemia)

### 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
- Gene Therapy
  - clinical trials underway
  - “Flip the switch” in SCD RBC back to fetal hemoglobin to prevent sickling

### Management Considerations

- Yearly testing
  - CXR, liver, pulmonary, renal function, CBC/reticulocyte count, urinalysis, abdominal ultrasound
- Transcranial Doppler studies of arteriole 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.
Questions???

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