Not So Atypical Pediatric Cases

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• 5 y.o AM referred in for RXT eval and amblyopia
  – Parents interested in VT for XT
  – Parents noticed the eye turn 1.5 years ago
  – Has worn glasses q1year
  – H/o patching 5-60min/day OS daily for 1.5 years
• BHx: Unremarkable
• DHx: Unremarkable
• Ehx: Unremarkable
• Meds: None
• FHx: Unremarkable

Lensometry: OD: +0.75-2.75x175  OS: plano-3.00x180
VAcc:  D: 20/50 OD, 20/50 OS with HOTV , poor attention
Vacc:  N: 20/50 OD,OS
Pupils, EOMS – normal
CVF: pt uncooperative
Nystagmus: unilateral (OD), high frequency, low amplitude horizontal nystagmus; dampens in left gaze and convergence
Color – pt uncooperative

CTcc: Primary gaze: 40 XT, 8RHT @ D (comitant), except no hyper on L tilt
NPC: poor
Stereo: RDS: none
Dry ret:
  OD: +1.50-1.50x180  OS: plano-2.00x180

• SLE- unremarkable
• IOPs: soft and equal : digital
Cyclo ret:
  OD: +3.00-2.50x180  OS: +1.50-2.75x180
DFE: Retinoschisis OD>OS
A: X linked juvenile retinoschisis
P: Consult with Dr. Brooks, Pedi OMD Stat
Anisometropic Amblyopia?

- 11 y-o male
- BCVA 20/20 OD and 20/50 OS
- Rx: +3.50 -3.50 X 175 OD
  -5.00 -2.00 X 180 OS

- Typically males
- VA 20/60-20/120
- Requires Retinal consult
- Low Vision Referral
- Avoid Contact Sports
- Tx: topical dorzolamide or oral azetazolamide
- In the pipeline gene replacement therapy
Another Anisometropic Amblyope?

- 12 year-old male
- BCVA 20/40 OD and 20/20 OS
- Rx: -6.00 OD, Plano OS
Myelinated Nerve Fibers, Dysplastic Discs and Myopia

- Developmental anomaly
- Affected macular integrity from birth
- Resulting myopia?
- Degree of myelination directly proportional to poorer prognosis of visual improvement with therapy


Nystagmus

- Nystagmus occurs in 0.4% of the clinical population.
- Frequency range of 2-5 Hertz (Hz) with an amplitude of 1-5 degrees. Usually horizontal in direction, although they may include a small vertical or rotary component.
- They cause sensory deficits such as reduced contrast sensitivity, visual acuity, and stereo acuity can have profound psychological effects resulting from the unusual cosmetic

Case 1 Nystagmus

- 7 month old
- F&F
- OD +1.25-0.50x180
- OS +1.25
- Pendular Nystagmus
- CAXT (Right eye preferred fixation)

- 3YO
- VA Cardiff OU 20/50
- EOM full OA IO
- Intermittent LXT

- 5yo
- 20/60 OD 20/32 OS
- EOM full
- NO tropia
Infantile/Congenital

- F – fixation
- U-upgaze
- N-null point
- B- bilateral
- L-latent
- O-OKN inverse response
- C-convergence
- S-symptomless

THE 5

- Aniridia
- Achromatopsia
- Lebers Amarosis
- Albinism
- Optic Atrophy

Nystagmus

Rutner Treatment of Choice

- less optical aberrations
- enlarged retinal image (in refractive myopes)
- increased peripheral visual field.
- Any or all of the above would improve the quality and/or extent of the retinal image, and hence provide a visual input of higher fidelity for fusion and subsequent visual information processing.

Nystagmus Case 2

- An 18-year-old female with non-PAN, congenital jerk nystagmus, who had never worn contact lenses, was evaluated. Her refraction and best corrected visual acuity with spectacles was OD -4.00D/-2.75D x 170 (20/120), and OS -4.00D/-3.00D x 025 (20/200). She had strabismus surgery two years earlier to correct a constant esotropia, with a residual 15 prism diopters of intermittent alternating esotropia. Ocular health examination revealed ocular albinism; all else was within normal limits.
- After a comprehensive eye examination to assess refractive, binocular, and ocular health factors, the patient was fit with CooperVision Preference Toric soft contact lenses (SCL). Lens parameters were: 8.7mm base curve, 14.4mm diameter, 0.09mm center thickness, OD -3.00D/-2.25D x 180, and OS -2.00D/-2.25D x 010.

Nystagmus

<table>
<thead>
<tr>
<th>Condition</th>
<th>Amplitude (degrees)</th>
<th>Frequency (Hz)</th>
<th>High Contrast Visual Acuity</th>
<th>Low Contrast Visual Acuity</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hot lens One</td>
<td>3.60</td>
<td>1.25</td>
<td>20/30 OD 20/200 OS</td>
<td></td>
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<tr>
<td>SCL</td>
<td>1.72</td>
<td>1.05</td>
<td>20/200 OD 20/200 OS 20/300 OS</td>
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<tr>
<td>SCL with anesthetic 1 week later</td>
<td>3.30</td>
<td>1.95</td>
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<tr>
<td>Hot lens One</td>
<td>2.98</td>
<td>1.80</td>
<td>20/60 OD 20/60 OS 20/600 OS</td>
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<tr>
<td>SCL one week later</td>
<td>9.72</td>
<td>5.00</td>
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Nystagmus

- Glasses
- Prism
- Contact lenses
- Biofeedback
  - Visual
  - Auditory
- Surgery
3rd Case

- 4 month old with nystagmus
- Healthy, FT
- No family history of ocular disease
- Parents not related
- Mild photophobia
- Nystagmus noted age 8 weeks

Examination Findings

- Blinks to light OU
- Tracking inconsistent
- Pupils equal, normal reactivity
- Conjugate, horizontal, pendular nystagmus with small amplitude and high frequency
- Cycloplegic refraction: +4.50 sph OU

Differential Diagnosis

- Congenital motor nystagmus/infantile nystagmus syndrome
- Albinism
- Retinal dystrophy (eg. LCA, rod-dome dystrophy, CSNB)
- Optic nerve hypoplasia
- Coloboma
Key Examination Points

- Assess visual function
- Iris transillumination
- Check for paradoxical pupil response
- Funduscopic examination

New Onset Nystagmus in a 2 year old

- Healthy 2 yo F
- No prior illness
- Sudden onset of ataxia and nystagmus
- Lethargy, irritability
- Myoclonic jerks

Examination Findings

- Rapid, conjugate, multi-directional jerky eye movements
- Central fixation, disrupted by jerky eye movements
- Normal pupils

Examination Findings

- Normal anterior segment and funduscopic examination
- Full range of EOM’s
- Cycloplegic refraction +2.00sph OU

Diagnosis

- OMA (opsoclonus-myoclonus-ataxia)
  - Post-encephalitic (viral)
  - Para-neoplastic (50%)
  - Idiopathic
- This is NOT nystagmus

Opsoclonus-Myoclonus-Ataxia

- Rare
- Requires urgent neurological evaluation
- Neuroblastoma most frequently implicated neoplasm
- Opsoclonus secondary to autoimmune attack of purkinje cells in the cerebellum
- Recovery often incomplete
5 year old with R face turn

- Healthy 5 yo M
- Noted by parents to turn habitually turn face to the right when watching TV
- Nystagmus noted age 4 months
- Good vision, asymptomatic
- Family history negative

Examination Findings

- Va 20/60 OD, 20/70 OS, 20/30 OU open, 20/25 OU open with face turn to R
- Conjugate, horizontal, nystagmus with fast phase to L in primary position
- Normal pupils and EOM’s

Examination Findings

- No iris transillumination
- No photophobia
- No strabismus
- Nystagmus decreases in right gaze, increases in left gaze
- Normal anterior segment and fundus exam
- Cycloplegic refraction: plano OU

Differential Diagnosis

- Congenital motor nystagmus/infantile nystagmus syndrome
- Manifest-latent nystagmus/fusion maldevelopment nystagmus syndrome
- Periodic alternating nystagmus
- Cerebellar lesion
- Vestibular lesion
- Retinal dystrophy

Interpretation of Examination

- Congenital motor nystagmus with null point in left gaze (causing right face turn)
- Visual acuity improved with OU open (latent component to nystagmus)
- Visual acuity improved in left gaze (null point)

Management

- Surgery to shift null point
- Surgery to reduce (dampen) nystagmus
- Prism glasses
- Imaging?? Neurology consultation??
- Follow-up
6 month old with OS “shaking”

- Healthy, FT
- Parents note that left eye seems to intermittently shake or jiggle
- Normal development
- No FH of ocular disease

Examination Findings

- Normal appearing baby
- Central, steady fixation with each eye
- Normal pupils
- OS intermittently shows very low amplitude, high frequency, pendular nystagmus
- Child seems to tilt head to left and shake head intermittently

Examination Findings

- Normal anterior segment OU
- Normal funduscopic examination OU
- Full EOM’s
- Cycloplegic refraction
  OD: +1.75+1.75x50
  OS: +1.75+1.50x130

Differential Diagnosis

- Spasmus nutans
- Sensory-loss nystagmus
- Retinal dystrophy
- Idiopathic
- Leukoencephalopathy/Leukomalacia

Interpretation of Exam

- Likely spasmus nutans

Management

- Neuro-imaging
- Close observation
- Correction of refractive error?
- Patching to prevent amblyopia?
8 month old with esotropia

- Parents have noted ET “since birth”
- Healthy, FT
- Family history positive for ET in mother and a cousin
- Slower to crawl than older sibling

Examination Findings

- Normal appearing baby
- Constant esotropia 40 PD by Krimsky testing
- Holds fixation well with either eye
- Mild limitation of abduction OU

Examination Findings

- Normal pupils
- Normal anterior segment and fundus
- No nystagmus
- Normal versions
- Cycloplegic refraction
  OD: +1.75 sph
  OS: +1.75 sph

Differential Diagnosis

- Congenital esotropia
- Infantile accommodative esotropia
- Sensory loss
- EOM fibrosis syndrome
- Duane syndrome
- Nystagmus blockage syndrome

Interpretation of Examination

- Large angle constant esotropia
- No amblyopia
- Normal vision for age, no evidence of ocular defects
- Essentially full ductions
- Minimal hyperopia, normal for age
Interpretation of Examination

• Likely congenital esotropia without amblyopia
• Accommodative component unlikely

Management

• Glasses??
• Patching??
• Vision therapy??
• Surgery?? Timing??

5 yo M with new onset Left ET

• Healthy
• No FH of ocular disease
• Normal development
• Occasional headaches recently

Examination Findings

• Va 20/25 OD, 20/25 OS
• Pupils normal
• Normal orbits
• Anterior segment: OD palpebral fissure narrower than OS

Examination Findings

• EOM: Full OD, mod limitation of Abduction OS
• No nystagmus

Interpretation of Findings

• Incomitant esotropia
• Limited ductions for OS
• No amblyopia
• Palpebral fissure asymmetry
Differential Diagnosis
• EOM fibrosis syndrome
• Duane Syndrome
• Myasthenia gravis
• Orbital lesion
• Sixth n. palsy (multiple possible causes)

3 year old with red eye OD
• Healthy
• Redness and photophobia OD for 10 days
• Not responsive to topical antibiotics
• Similar episode 6 months ago
• Tearing

Examination Findings
• Va 20/50 OD, 20/25 OS (allen pics)
• OD mild ptosis
• OD 2+injection, small epi defect and opacity paracentrally with pannus
• No adenopathy

Examination Findings
• No hypopyon
• No FB
• Eyelid margins clear
• Unable to measure IOP
• Fundus normal
Interpretation of Findings

- Keratitis OD with corneal neovascularization
- Unresponsive to topical antibiotics
- Prior episode in same eye

Differential Diagnosis

- Recurrent corneal erosions
- Occult FB
- Staph marginal keratitis
- HSV

3 year old with head tilt

- Healthy
- Head tilt to R noted age 10 months
- Normal development
- Family history negative for ocular disease

Examination Findings

- Va Fixes and Follows well each eye
- Tilts head approx. 10 degrees to the right
- Resists having head tilted to the left
- Normal pupils
- Normal anterior segment and optic disc OU

Examination Findings

- Left inferior oblique overaction
- Full ductions OU

Three Step Test

- Measure hypertropia in primary gaze
- Measure hypertropia in R and L gaze
- Measure hypertropia in R and L head tilt
Example: LHT secondary to LSO palsy
Step 1 (LHT in primary)

Example: LHT secondary to LSO palsy
Step 2 (LHT worse in right gaze)

Example: LHT secondary to LSO palsy
Step 3 (LHT worse in left tilt)

Additional Testing

- Patch test
- Fundus exam
- Review old photos
Diagnosis

• Left superior oblique palsy

8 month old with R ET

• Healthy, FT
• Normal development
• Family history of strabismus in a 1st cousin

Examination Findings

• Va Blinks to light, poor fixation OD, Fixes well OS
• Pupils appear normal
• Anterior segment and red reflex clear OU

Examination Findings

• Possible nystagmus OD
• 25 RET
• Full ductions

Interpretation of Findings

• Esotropia
• Decreased vision OD
• Normal ocular motility
• Concern for vision loss OD leading to ET
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COATS

- Abnormal development of Blood Vessels in the Retina: dilated tortuous and leaky
- Males to females 3:1
5 STAGES

• 1. abnormal blood vessels are seen in the retina but these vessels are not yet leaky.
• 2. leakage from the vessels into the retina. Effect on VA variable
• 3. Retinal detachment
• 4. Complicated with glaucoma (raised
• 5. End stage blind painful eye.

Treatment

• Referral for cyro
• Laser
• Surgical

Not So Simple Tropia

• 3 year old referred by pediatrician for second opinion for strabismus surgery

Previous Hx

• H/O strabismus surgery secondary to double elevator palsy
• Previous diagnosis of bilateral ambylopia
• Born at 34 wks
• 4lbs 8 oz at birth
• +developmental delays with +ST, OT, PT
• Long standing ptosis of right eye

Monocular elevation Deficiency

• Monocular Elevation Deficiency, also known by the older term Double Elevator Palsy, is an inability to elevate one eye, usually resulting in one eye that is pointed downward relative to the other eye
• The apparent paralysis of both elevators (superior rectus and inferior oblique) of one eye that results in a rather large hypotropia on the affected side is uncommon. The levator palpebrae may or may not be involved, and Bell’s phenomenon may be present but is usually absent; if it is present, a supranuclear lesion is implied. The pupil is normal, as are horizontal rotations.
• The eyelid on the involved side is droopy (ptosis) 25% of the time while 75% of cases have pseudoptosis. In this case, the pseudoptosis is the appearance of ptosis caused by the eye being hypotropia (downward deviation).
• 25% of those with Monocular Elevation Deficiency and Congenital Ptosis have a phenomenon called Marcus Gunn jaw-winking. This a condition in which the cranial nerve that usually controls eyelid movement is mis-wired with the cranial nerve that controls chewing or sucking thus creating a “wink” when chewing or sucking
Cycloplegic Refraction

- VA
  - OD 20/70 current Rx pl-0.75x5
  - OS 20/60 current Rx +0.25-0.75x158

- OD +1.00-1.00x180
- OS +1.00-2.00x150

Cover Test

- With correction
- 15CLHT, 15 CLXT equal distance and near
- With a V pattern
- EOM full with over action of the IO of the left eye

Slit Lamp

- Normal findings
- No ptosis noted

Dilation

- FLN
Neurofibromatosis

- NF 1
  - Lisch Nodules
  - Café au lait spots (>6)
  - Learning Disabilities
  - Large Head
  - Bone deformities
  - ONH glioma

- NF 2
  - Hearing loss
  - Schwannomas
  - Numbness
  - Weakness
  - Cataract

- Schwannomatosis
  - Numbness or weakness in various parts of your body
  - Loss of muscle
  - PAIN

- Patient Demographics: 7 year old Hispanic female

- Chief Complaint: Returning to the clinic for a comprehensive examination with no ocular complaints. Has been moderately compliant with spectacle wear

Ocular History:
- Has been followed since at the UEC x 5 years and LEE x 3 years.
- Hyperopia and astigmatism OU, full time wear glasses
- OMD
- Physiological cupping OU

Medical History:
- LME: 10-2013 with blood work: unremarkable;
- Asthma, Seizures;
- Father reported pt. was scratched by a cat a few days prior with no treatment sought; reports no fever or myalgia

Milestones: Slightly delay in speech

Birth and Educational History are all unremarkable

Family Medical and Ocular History:
- Mother (+) Progressive MS, Arthritis, Glaucoma

Pertinent Findings

Visual Acuity: Snellen
- OD: 20/20
- OS: 20/20

Glasses RX:
- OD: +0.25 -1.00 x180
- OS: +0.50 -1.50 x065

Extra-ocular motilities, color vision, stereo, pupils, cover test at distance and near, near point of convergence, accommodation: all unremarkable

IOPs:
- 12mmHg OD, OS at 5:20pm

SLE: Unremarkable OU

Dilated Fundus Exam:
- C/D: 0.55V/0.50H; 0.55V/0.60H, pink, distinct, healthy rim margins
- Vessels: 2/3, normal caliber
- Macula: flat and clear (+) FR OU
- Periphery: Bilateral vitreal snow balling I/IT/IN OS>OD and vitreous heme OS adjacent to the area of snow banking

Additional testing:
- Fundus photos (unable to capture snowbanking with photos)
- Lymphadenopathy (unable to capture swelling with photos)
Intermediate uveitis, or pars planitis, is an insidious, chronic, relapsing inflammation of the anterior vitreous and pars plana. While uveitis occurs in 5-10% of the pediatric population, pars planitis accounts for 8-33% of all uveitis in pediatric patients.

Clinical presentation includes snowballs, snowbanking, vitritis, and peripheral retinal vasculitis. Vitreous hemorrhages occur in 16.7% of pediatric patients. It is typically bilateral and asymmetric and patients often remain asymptomatic.

Although pars planitis is often idiopathic (96%) in pediatric patients, inflammatory diseases must be ruled out as a possible etiology.

Systemic Associations
- **Idiopathic:** Most common
- **Sarcoidosis:** relatively uncommon, can precede systemic disease
- **Lyme disease** associated with severe anterior uveitis.
- **Tuberculosis:** uncommon
- **Toxoplasmosis**
- **EBV or HTLV-1 infections**
- **Neurological concomitants:** Vogt–Koyanagi–Harada syndrome,
  Behçet syndrome, AIDS, primary CNS lymphoma, herpes virus infections,
  syphilis, acute posterior multifocal placoid pigment epitheliopathy
  and Whipple disease.
- **Multiple sclerosis:** 15% of the patients with pars planitis developed
  MS after five years of follow-up

3 year old with red eye OD
- **Healthy**
- **Redness and photophobia OD for 10 days**
- **Not responsive to topical antibiotics**
- **Similar episode 6 months ago**
- **Tearing**

Examination Findings
- **VA 20/50 OD, 20/25 OS (allen pics)**
- **OD mild ptosis**
- **OD 2+injection, small epi defect paracentrally with pannus**
- **No adenopathy**

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- **No hypopyon**
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- **Eyelid margins clear**
- **Unable to measure IOP**
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- Left inferior oblique overaction
- Full ductions OU

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Tilt R: ortho  Tilt L: 12 LHT

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- Normal ocular motility
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