2018 Victoria Conference
Workbook Part 2

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Course Description
This course will review the types of corneal ectasias and their possible etiologies and treatment options with an emphasis on keratoconus. Cases will be used to highlight diagnosis and management options.

Course Learning Objectives
After participating in this course, the participant should be able to:

- Be familiar with the difference between a corneal dystrophy and degeneration
- Be able to identify the different ectasias with corneal topography
- Identify keratoconus and the various treatment options
- Be familiar with the corneal crosslinking procedure and criteria for referral
- Be familiar with the contact lens management options for ectasias

Course Outline
- Corneal Ectasia
  - Non-inflammatory
  - Bilateral thinning of the central, paracentral or peripheral cornea
- Dystrophy vs. Degeneration
  - Controversial classification
  - Case for dystrophy
    - Genetic basis
      - Family history in 10% of cases
        - Found in twins / families with 2 or more generations
      - Increased prevalence in 1st degree relatives
      - Increased prevalence in trisomy 21 (Down Syndrome)
  - Case against dystrophy
    - Sporadic (in an individual only)
    - Causative associations with eye rubbing and atopy
    - Insufficient evidence of clear genetic basis
- Genetics of Keratoconus
  - Found in identical twins / families with 2 or more generations
  - Prevalence in 1st degree relatives
    - 15-67 times higher than the general pop.
  - Associated genes
    - Chromosome 21, chromosome 17 (Leber’s congenital amaurosis), chromosome 13
  - Autosomal dominant inheritance with variable penetration
  - In the Japanese, human major histocompatibility complex (HLA) antigens
- HLA-A26, B40 and DR9 associated with early onset

- Corneal Ectasia
  - Keratoconus
  - Keratoglobus
  - Pellucid Marginal Degeneration
  - Posterior Keratoconus
  - Post-LASIK ectasia
  - Terrien’s Marginal Degeneration

- Keratoconus
  - The incidence of keratoconus is approximately 1 in 2000
  - What we know about keratoconus
    - The condition occurs in every country throughout the world
    - Occurs equally in men and women?
    - Usually begins between the ages of 12 and 32
    - Condition of unknown etiology
  - Associated Conditions
    - Atopic disease
      - Oxidative stress?
    - Eye rubbing
    - Hereditary
    - CL wear?
      - CLEK study
      - Do CL stop or slow the progression?
  - Only 10% of People with KC Undergo Corneal Transplant Surgery
  - Unilateral keratoconus
  - Hallmarks of Keratoconus
    - Decline in visual acuity
    - Changes in cylindrical power and axis
      - Scissor reflex on retinoscopy
    - Topographical changes
    - Squinting of the eyelids, artificially creating a pinhole effect
    - Appearance of halos around street lights
  - Morphological Changes in Keratoconus
    - Epithelium
      - Basal cells degenerate - grow towards Bowman’s layer
    - Stroma
      - Reduction and disorganization of lamellae
    - Decrease number of keratocytes
    - Descemet’s membrane
      - Hydrops
    - Endothelium
      - Change in cell shape with cells pointing towards the cone
  - Clinical Changes in Keratoconus
    - Corneal nerves more visible
    - Scissor reflex with retinoscopy
    - Charleaux oil drop
- Fleisher’s ring
  - Iron deposition at the base of the cone
- Straie
  - Compression of Descement’s membrane
- Corneal thinning
- Corneal scarring
- Hydrops
- Munson’s sign
- Rizutti’s sign

- Puberty Onset Keratoconus
  - Begins in early adolescence approximately age 12 to 16
  - Usually bilateral with one eye affected worse than the other
  - The younger the patient, the more severe the condition.

- Late Onset Keratoconus
  - Usually begins in late 20’s or early 30’s
  - Both eyes can be affected the same
  - The incidence of progression reduces greatly with the age of onset

- Keratoconus Fruste
  - A mild non-progressive form KC
  - Can occur anytime throughout life
  - No positive slit lamp findings associated with KC
  - Normal corneal thickness
  - Often diagnosed with topography

- National Keratoconus Foundation

- Classification
  - Morphology
    - Nipple
      - Cone is ≤ 5 mm round near the central or paracentral cornea
    - Oval
      - >5 mm and paracentral or peripheral
    - Keratoglobus
      - Cone is throughout 75% of the cornea
  - Disease evolution
    - Four stages
  - Index-based
    - Topography
    - OCT
    - Aberrometry

- Treatment Options
  - Spectacles
    - Need more regular cornea to be a good option
  - Corneal GP lenses
    - Potential for great acuity
    - Adaptation to comfort
    - Case example of GP contact lens fit
- **Soft contact lenses**
  - Potential for similar (or better) acuity as spectacles
  - Stability of vision
    - Consider large astigmatism correction
- **Custom soft contact lenses**
  - No CL vs. over CL
  - Initial base curve selection
    - Mean K + 1.00 mm
  - Select fitting curve
    - 8.3 mm, 8.6, mm 8.9 mm
  - Select overall diameter
    - 10.0 mm to 17.0 mm
  - Material
    - Hydrogel
    - Silicone hydrogel
      - Lathe
      - Reproducibility
  - Case example for custom SCL
    - A 23-year-old presents with a chief concern of vision that has gotten worse OD>OS for the past 2 years and is seeking a second opinion regarding refractive surgery
    - Exam findings
      - Decreased best corrected acuity, distortion with keratometry
      - Anterior segment is unremarkable
      - Topography – inferior steepening with superior/nasal flattening
    - Treatment
      - Custom SCL with increased center thickness
- **Scleral GP lenses**
  - Potential for great acuity
  - Less adaptation to comfort
  - Cost
  - Anatomy of a Scleral Lens
    - Central zone, peripheral zone, limbal zone, and scleral zone
  - Fitting Goals
    - Clearance across the central cornea
    - Increase limbal clearance
      - Bright ring of fluorescein at the limbus
    - Scleral alignment
      - All pressure, weight and bearing of the lens should be on the sclera
  - What is the Appropriate Apical Clearance?
    - 200 to 400 microns
  - Limbal Clearance Zone
- Appropriate scleral landing zone
- Case example for scleral GP fit

### Implantable Intracorneal Rings Segments
- Flatten the cornea and decrease irregular astigmatism
- Need minimum of 450 um at incision site
- Removable

### Corneal Cross-Linking
- Hope to stabilize / slow progression
- Generally still needs some type of vision correction
- Concept first introduced in 1998
- Strengthen tissue by photosensitization and chemical cross-linking
- Exposed to measured dose of UVA radiation
  - Free radicals are produced covalent bonds between collagen molecules and glycosaminoglycan molecules
  - Exact mechanism is not understood
- April 2016 - FDA granted approval to Avedro Inc. for its corneal cross-linking system to treat patients with:
  - Progressive Keratoconus
  - Post LASIK Ectasia
- Dresden Protocol
  - Epithelial debridement over the central 9mm
  - Topical riboflavin is instilled every 2 minutes x 30 minutes
    - Saturation of the stromal with riboflavin
  - UVA (365nm) irradiance of 3.5 mW/cm² x 30 minutes
  - Riboflavin every 2 minutes x 30 minutes
  - Antibiotic and steroid drops + bandage CL
    - Removed when epithelium heals
- Normal Corneal Anatomy = 540 um
- In Corneal Crosslinking the Epithelium is Removed
- Minimal stromal thickness is 400 um
- Most of UV energy dissipated in the anterior 400 um
  - Energy level at the endothelium 0.18 J/cm²
  - Half of the endothelial damage threshold
- Transepithelial Cross-linking
  - Epithelial is not removed
  - Thinner corneas
  - New formulations of riboflavin and/or longer pre-op loading time (45-60 minutes) may contribute to improved stromal uptake
  - Possibly higher rates of progression vs. Dresden
    - Shallower treatment depth
- Accelerated Cross-linking (Law of Reciprocity)
  - Twice the level of irradiance (30mW/cm² x 3 minutes)
- Effects on the cornea
  - Early apoptosis of keratocytes to 300 um
Keratocyte repopulation after 3-12 months, contributes to post-op haze

- Stromal edema
- Loss of sub-epithelial nerve pelxus
- Loss of midstromal nerve fibers
- Increased reflectivity in the mid-stroma
  - Contributes to post-op haze in the anterior stroma
  - Plateau in 1-3 months then declines
  - Generally does not affect acuity
- Increased corneal stability
  - Increase in collagen fiber diameter (12.2% anterior stroma and 4.6% posterior stroma)
  - Resistance to enzymatic degradation
  - Increase in corneal rigidity by 328% (Wollensack, Spoerl and Seiler)
- Visual Acuity
  - Worsening acuity up to 1 month
  - Improvement 6 months to 1 year
- Keratometry
  - Steepening up to 1 month
  - Flattening 6 months to 1 year
- CXL Complications
- Patient selection
  - Progressive keratoconus
    - Changes to keratometric (Sim K) measurements
      - Increase in maximum value of 1.00 D in a year
    - Change in refraction?
    - Change in CL base curve
    - Clinical judgment
      - Minimal stromal thickness of 400um
- Contraindication
  - History of herpetic eye disease
  - Pregnant or nursing
  - Central corneal opacity
  - Stroma thickness less than 400um

- Treatment Options
  - Protect against oxidative damage
    - Use UV filters
    - NSAID
      - Placibo effect?
    - Preservative-free artificial tears
    - Ophthalmic antihistamines / mast cell stabilizers
    - Minimize corneal microtrauma
- Pellucid Marginal Degeneration
  - Rare
- Thinning of the inferior cornea
- Onset during 4th and 5th decade
- Slowly progressive band of thinning down to 1 mm from the limbus
- Topography - Generally high ATR astigmatism
  - Early to moderate stages can be corrected with a toric SCL
  - Case example of PMD SCL fit
- Posterior Keratoconus
  - Affects the posterior cornea and may not be neutralized with a contact lens
  - Posterior corneal imaging
- Post-LASIK ectasia
  - Treatment and management similar to keratoconus
  - Screening for ectasia prior to refractive surgery
- Terrien’s Marginal Degeneration
  - Rare
  - Unknown etiology
  - Mostly asymptomatic
  - More common in males >40 years old
  - Early
    - Stromal opacification superiorly
  - Late
    - Thinning superiorly and circumferentially
  - Perforation is uncommon but may occur spontaneously or with trauma
  - Topography - ATR or Oblique astigmatism
Learning Objectives

1. Which cause of Chiari malformation is most common? What are the presenting symptoms?
2. Which cause of Chiari malformation is not evident with imagining, but may have ocular signs and symptoms?
3. What are the differential diagnoses for Chiari syndrome?
4. What are the surgical and non-surgical treatments for Chiari malformation?

Definition of Chiari Malformation

- Chiari malformation is traditionally defined as the cerebellar tonsils being located 3mm-5mm or more below the foramen magnum as measured on an MRI.

Problems with the Traditional Chiari Definition

- Some people have large herniations with no symptoms.
- Others have only small herniations, but are severely symptomatic.

Variable Symptoms of Chiari Malformation

Chiari Symptoms
Chiari Signs

Brainstem Compression in Chiari Malformation

Causes of Chiari: Increased CSF Pressure
- Chiari doesn’t require obstruction, just higher CSF pressure
- This is sometimes called Chiari Type 0, and resembles idiopathic intracranial hypertension (IIH), and may be indistinguishable from it

Causes of Chiari: Small Posterior Fossa
- This is the default “anatomical determinism” theory
- When this occurs, transient but recurrent hydrocephalus can result
- In these cases, surgery may be the best option

Causes of Chiari: Connective Tissue Disorders
- There is one published large study that has looked at the association between type 1 Chiari and EDS
- This was undertaken by Milhorat et al. in 2007 at The Chiari Institute in New York
- They looked at 2813 patients with a known diagnosis of type 1 Chiari malformation
- They found that 357 (12-13%) had features of EDS
Causes of Chiari: Tethered Cord (?

- "Tethered cord is a relatively new entity, medically speaking, and as such there is still quite a bit of controversy surrounding it.
- There is even occult TCS since the tethering is not apparent on MRI.
- A second area of controversy involves the relationship, if any, between tethered cord and Chiari and/or syringomyelia."

How Common is Chiari? (and does IIH mimic it?)

- The US Association of Neurological surgeons performs 10K Chiari surgeries per year.
- Chiari may be as common as 1:1000 patients in the US. Also...
  - "Cerebellar tonsil position in patients with IIH was significantly lower than that in age-matched controls, often times peg-like, mimicking Chiari."

How Common is Chiari? (and does IIH mimic it?)

- "Cerebellar tonsil position in patients with IIH was significantly lower than that in age-matched controls, often times peg-like, mimicking Chiari."

Gender and Ethnic Trends in Chiari Malformation

- Chiari is often diagnosed in young adults.
- Women get Chiari more often than men.
- It affects all ethnicities.

Three Types of Chiari

1. Chiari Malformation, Type I
2. Arnold-Chiari Malformation
3. Chiari Malformation, Type III

Three Types of Chiari

1. Chiari Malformation, Type I
2. Arnold-Chiari Malformation
3. Chiari Malformation, Type III

Classical Chiari Type I

Chiari malformation happens when the cerebellar tonsils protrude through the foramen magnum, pinch the medulla, and block CSF flow.
**Congenital Chiari Type I**

- Much less common than Type I, this birth defect leads to fourth ventricle hydrocephalus
- Expect childhood-onset with more severe symptoms
- These patients are usually born with paraplegia due to a defect (hole) in the spine and back called a myelomeningocele

**Arnold-Chiari (Type II)**

- This terrible birth defect is characterized by cerebellar herniation outside the skull cavity, called an encephalocele
- This happens when the neural tube does not close during the first trimester of gestation
- Various teratogens like arsenic can cause this usually fatal condition

**Chiari, Type III**

- Visual Symptoms of Chiari
  - Blurred Vision
  - Diplopia
  - Nystagmus
  - Photophobia
  - Strabismus
Symptom Profiles That Mimic Chiari

<table>
<thead>
<tr>
<th>Set of Symptoms</th>
<th>Conditions with similar symptoms</th>
</tr>
</thead>
<tbody>
<tr>
<td>Headache</td>
<td>Multiple Sclerosis</td>
</tr>
<tr>
<td>Dizziness</td>
<td>Fibromyalgia</td>
</tr>
<tr>
<td>Weakness in arms/hands</td>
<td>Chronic Fatigue</td>
</tr>
<tr>
<td>Numbness in arms/hands</td>
<td>Lupus</td>
</tr>
<tr>
<td>Leg Weakness</td>
<td>Migraines</td>
</tr>
<tr>
<td>Fatigue</td>
<td>Carpal Tunnel ALS &amp; Low Gehrig</td>
</tr>
</tbody>
</table>

Chiari Obscured by Morning Glory Disc

Chiari Malformation and Eye Movements
- Milder cases of Chiari may present as intermittent diplopia
- Often, this presents as esotropia at far of the divergence insufficiency type
- When this occurs, suspect CN VI palsy

Pursuits and Saccades in Chiari Malformation

Eye Movements in Chiari

Syrinxes and Syringomyelia
- A syrinx is a fluid-filled cyst in the spinal cord
- This can cause spinal cord edema, compromising neuronal function
- Permanent nerve damage can result
Normal vs. Chiari Cerebellum & Syrinx

Spinal Cord Syringes and Chiari Symptoms

Syrinxes and Chiari Surgery

Surgical Treatments: Posterior Fossa Decompression

Craniectomy for Chiari

Opening the Dura in Chiari
Dura Patch for Chiari

Surgical Time Course for Posterior Fossa Decompression

Surgical Time Course for Posterior Fossa Decompression

Surgical Outcomes for Posterior Fossa Decompression

Case #1: Chiari Surgery Relieves Cluster-Like Headache
Case #2: Pre-Op Signs of Chiari in 13 YOM with ↑BMI

A pre-operative sagittal T1-weighted MRI demonstrating CMI with tonsillar herniation 5 mm below the foramen magnum.

Case #2: Post-Op Signs of Chiari in 13 YOM with ↑BMI

A sagittal T2-weighted MRI performed 6 months after suboccipital decompression demonstrating the creation of a normal-sized cisterna magna for sufficient CSF flow.

Case #3: Chiari Causing Esotropia Pre-Op in 6 YOF

Case #3: Chiari Causing Esotropia Post-Op in 6 YOF

Non-Surgical Chiari Tx: Acetazolamide and Lasix

Future Directions in Chiari

- Researchers are looking for a new way to measure Chiari severity
- Focus areas include advanced MRI and engineering techniques to quantify cerebrospinal fluid (CSF) flow, crowding and compliance

GIF Animation of CSF flow in Chiari


https://www.mayfieldclinic.com/PE-Chiari.htm

http://www.mayfieldclinic.com/PE-Chiari.htm
Questions? Thank you!

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Reading and References

For more information, see
Eye Movement Disorders by
Agnes Wong
(2007, see chapter 10 on
the cerebellum)
Evidence Based Optic Nerve
Anthony DeWilde, OD FAAO
Kansas City VAMC

No financial disclosures

Goals
1. Understand Diagnostic Strategies
2. Learn Nuances of GCA
3. Develop Referral Strategy

Diagnosis
1. Possible
2. Probable
3. Prognostic
4. Pragmatic

Optic Nerve Edema
V - Vascular
O - Ophthalmic
I - Inflammatory
C - Compressive

Optic Nerve Edema
Vascular
AION
NAION
Optic Nerve Edema

Ophthalmic

Drusen
CRVO
Hypotony

Optic Nerve Edema

Inflammatory

Infectious (syphilis, etc)
Non-infectious (collagen vascular)

Optic Neuritis

Optic Nerve Edema

Compressive

Tumor
Graves’ Disease
Chiasmal

Optic Nerve Edema

Probable

Based on Age
40 and younger -- Optic Neuritis
50-60 -- mostly atypical
60+ -- NAION

Optic Nerve Edema

1. Possible
2. Probable - based on age
3. Prognostic
4. Pragmatic

Optic Nerve Edema

1. Possible
2. Probable
3. Prognostic
4. Pragmatic
Optic Nerve Edema

1. Possible
2. Probable
3. Prognostic - GCA
4. Pragmatic - GCA

What is GCA?
Immune-Mediated Vasculitis
Focal arteritic lesions — Ischemia
Affects medium, large arteries
~18 per 100,000


What is GCA?
Immune-Mediated Vasculitis
Focal arteritic lesions — Ischemia
Affects medium, large arteries
~18 per 100,000

How does the Temporal Artery connect to the eye?

Why is it so important?
Profound Vision Loss
Bilateral in 14 days in 1/3 if Untreated
Systemic Complications
Treatable
**Ophthalmic**

Anterior Ischemic Optic Neuropathy (AION)
Central Retinal Artery Occlusion (CRAO)
Amaurosis Fugax
Diplopia


**Systemic**

Headache
Jaw Claudication
Scalp Tenderness
Neck Pain
Anorexia/Weight Loss

**A-AION**

Sudden, Painless Vision Loss
Amaurosis Fugax
Occurs ≥ 50 years of age
1 out of 10

**Ocular Symptoms**

Vision loss
Amaurosis Fugax
Amaurosis Fugax

From 7% to 50% of patients with GCA

Hayreh found 30%

In sharp contrast to NAION (2.5%)

Transient ischemia to ONH


Less Predictable

Headache

Fever

Scalp Tenderness

Malaise


Systemic Symptoms

Jaw Claudication (Odds Ratio 9.0)

Neck Pain (Odds Ratio 3.4)

Anorexia (Odds Ratio 2)


Headache

A-AION - 46% had Headache

NA-AION – 54% had Headache

Could Mislead

Average Number of Symptoms = 3

Occult GCA
Between 5 and 38% of cases
No systemic symptoms


Contralateral Eye
Important for 2 reasons
1. Gives us clues about diagnosis
2. Make sure other eye stays healthy

C/D Ratio
Average C/D in Population = 0.4

Contralateral C/D in NA-AION
• 75% are ≤ 0.3
• 33% are ≤ 0.15

C/D Ratio
A-AION
• ≤0.3 = 50/725 = 1/15
• ≥0.4 = 50/275 = 1/5
C/D Ratio

Some evidence says:
90% of C/D in NA-AION is \( \leq 0.3 \)

Then…
\( \leq 0.3 = \frac{50}{860} = \frac{1}{17} \)
\( \geq 0.4 = \frac{50}{140} = \frac{1}{3} \)

Testing

Labs – ESR, CRP, CBC, Platelets
Fluorescein Angiography
Ultrasound, PET, MRI – Limited Benefit
Temporal Artery Biopsy

FA

Management of ischemic optic neuropathies

Labs

ESR
\( \geq 33 \text{ mm/h} \)
Sensitivity 92%
Specificity 92%


Labs

CRP
\( \geq 2.45 \text{ mg/dl} \)
Sensitivity 100%
Specificity 82%


Labs

CBC includes
WBC
RBC
Platelets

ESR + CRP

<table>
<thead>
<tr>
<th>Sensitivity 100%</th>
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<tbody>
<tr>
<td>Specificity 97%</td>
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</table>

Platelets

Odds Ratio:
- ESR > 47 mm/hr = 1.5
- CRP > 2.45 mg/dL = 5.3
- Platelets > 400,000/μL = 4.2
- All 3 elevated = 8


Additional Testing

Temporal Artery Biopsy

- Gold Standard
- Case by Case
- Side Effects
  - Necrosis, Infection, Nerve Damage
  - Bilateral?

If case is equivocal
- ESR + CRP +
- ESR + CRP -
- ESR - CRP +
- ESR - CRP -

Clinical Picture

Unilateral Optic Disc Edema

Age

Systemic Symptoms

Labs (ESR, CRP, Platelets)

Other Eye

Temporal Artery Biopsy

American College of Rheumatology

Need 3 of the following 5
1. Over 50 years of age
2. New onset of Headache
3. Scalp tenderness
4. ESR > 50 mm/h
5. (+) Temporal Artery Biopsy
American College of Rheumatology

Meeting this criteria yields:

94% Sensitivity
91% Specificity

NOT good enough with swollen optic nerve

Study of ACR Criteria

112 Patients in Neuro-Ophthalmology Clinic
25% with + TAB missed by ACR
28% with - TAB met criteria


Case

62 Year Old Male
Sudden Vision Loss
Unilateral Optic Nerve Edema
What now?

More Information

Case

62 Year Old Male
Sudden Vision Loss
Unilateral Optic Nerve Edema
What now?

More details….

• Other eye = C/D 0.5
• Systemic symptoms = Headache, Neck Pain
• Labs: ESR = 70 mm/h
  CRP = 3.2 mg/dl

What If...

More details….

• Other eye = C/D 0.2
• Systemic symptoms = Headache
• Labs: ESR = 20 mm/h
  CRP = 0.8 mg/dl

Treatment

Oral Steroid
80-100+ mg
VERY long taper

75% reached 5 mg/day at year
Treatment

IV Steroid no better
Limited Evidence for Immune Modulators
TNF Blockers
Methotrexate

Clinical Picture

Unilateral Optic Nerve Edema
Systemic Symptoms
Lab Results
Other Eye

Referral

If AION suspect
Labs
Case History
Possible referral

Referral

Ophthalmology
Rheumatology
Neurologist
Urgent!!!

Giant Cell Arteritis

Affects Eyes
ION, CRAO, Diplopia, Amaurosis Fugax
Affects Systemic
Elevated Labs, Symptoms
Emergency!

MR. A

63 y/o White Male
New onset headache, temporal pain, neck pain
NO vision complaints
EOM full
NO APD
MR. A Labs
ESR = 56
CRP = 1.37
Pending Temporal Artery Biopsy

MR. A

Started on 40 mg Pred
When he tapers, headaches return
What about vision?
20/70 and VF reduction
MR. A

When Pred resumed, vision returned to 20/25
Visual Field improved

MR. A

Has been on low dose Prednisone for 4 years!

MR. T

63 Year Old White Male
Presents to ER with vision loss OS
Progressed over the day
MR. T

20/20 OD
NLP OS

MR. T

Anterior Segment normal
0.2 C/D OD
Edematous OS

MR. T

Scalp Tenderness
NO headache, jaw claudication, neck pain
Intermittent diplopia a month ago

MR. T

ESR = 96 mm/h
CRP = 6.9 mg/dL

MR. T

Admitted and started on IV corticosteroids
Temporal Artery Biopsy +

MR. T

12 years later on 5 mg/day Prednisone
If tapers off, symptoms start OD
NAION

Sudden, Painless Vision Loss

3/4 Wake up with Vision Loss

Occurs 50+ years of age

9 out of 10

NAION

Systemic Risk Factors

HTN

DM

Nocturnal Hypotension

Hyperlipidemia

NAION

More common in small optic cup

≤ 0.15 in 33%

≤ 0.3 in 75%

Small C/D NOT primary factor

NAION

“Disk at Risk”

“Disk at Risk”

Average C/D = 0.4

Incidence NAION = 10 per 100,000
NAION

20/20 in 33%
≥ 20/40 in 50%
≤ 20/200 in 20%

NAION

Treatment?
Referral?

Case

64 Year Old White Male
C/O Blur OD
S/P PCIOL OU

nature.com
Case

+DM
+HTN

Only symptoms is neck pain

Case

ESR = 32
CRP = 0.22
Platelets = 180,000

Case

What’s the plan?

Case

Get opinion from Rheumatology
Treated with Corticosteroids
No benefit noted - tapered quickly

Ophthalmic

Hypotony
CRVO
Drusen
Consider lab work/imaging with:

- Age
- Systemic health
- Symptoms

Infectious vs. noninfectious

- Spirochete - Syphilis
- Viral
- Cat scratch
Syphilis

- If suspected, it’s tertiary
- Need to evaluate CSF
- Treated with infused antibiotics

Inflammatory

- Non-Infectious
- Sarcoidosis
- Other connective tissue disorders

Optic Neuritis

- Consider if 40 years old or younger
- 1/3 are swollen
- 2/3 retrobulbar

Lhermitte symptom

- Uhthoff phenomenon
- Numbness/weakness
- Tingling
Optic Neuritis

25% Optic Neuritis initial manifestation of MS

Optic Neuritis

MRI
At 10 years
Overall rate of developing MS was 38%
If no lesions on MRI, risk of MS was 20%
If 1 or more lesion, 56%

Optic Neuritis

MRI
At 15 years
If no lesions on MRI, risk of MS was 25%
If 1 or more lesion, 75%

Optic Neuritis

Treatment - current attack
Steroids
Plasmapheresis

Optic Neuritis

Treatment - prevention
Beta Interferon (Avonex, etc)
Copaxone
Immune suppression - monoclonal antibodies

Optic Neuritis

Treatment
IV steroid followed by oral steroid sped recovery (4 days compared to 15 days)
Reduced recurrence of Multiple Sclerosis by 2 years
Optic Neuritis

Treatment
Oral steroids alone did not help
Increased recurrence

CHAMPS Trial (Interferon)
50% of placebo progressed to MS in 3 yrs
35% of interferon treated progressed

Optic Neuritis

Referral
Neurology or Neuro-ophthalmology
Order MRI if available

Graves’ Disease
80% are Hyperthyroid
Sweat, tremor, weight loss
10% are Hypothyroid
Cold, weight gain, hair loss
10% are Euthyroid

Graves’ Disease
30-50% of Graves’ patients have orbitopathy
2-5% serious complications
(Compressive Optic Neuropathy)
Graves’ Disease

Dry eye
Injection
Eyelid retraction
Diplopia
Compressive Optic Neuropathy

Graves’ Disease

Increase in
Fibroblasts
Hyaluronic Acid
Collagen
Adipose

Orbitopathy


Graves’ Disease

Exophthalmos

Hertel
Asian upper limit = 18
White upper limit = 21
Black upper limit = 24

Graves’ Disease

Free T3 and T4
TSH
Anti-thyroglobulin (TSI)
Thyrotropin-Binding Inhibitory Immunoglobulin (TBII)
Thyroid Peroxidase (TPO)
Graves’ Disease

CT allows measurement of
- Orbital fat
- Lacrimal gland
- Extraocular muscles
MRI for serial imaging

Graves’ Disease

Treatment
- Quiet inflammation - Steroid
- Stabilize Thyroid

Graves’ Disease

Stabilize Thyroid
- Medication
- Surgery
- Radioiodine

Graves’ Disease

Treatment
- Ocular Comfort
- Prism
- Surgery

Graves’ Disease

Surgery
- Orbital Decompression
- Strabismus
- Eyelid
- Cataract
Graves’ Disease

50 year old male
Complains of:
  Diplopia
  Swollen Eyelid OS
  Red Eye OS

Exam

20/25 OD and OS
IOP 18/18
No APD
Diplopia in lateral and downgaze
Pain in lateral gaze

Exophthalmos OS
Hertel 19/23
Lagophthalmos
Conjunctival edema and injection
Eyelid edema

Smoking makes disease worse
Smoking makes treatment less effective
Labs

TSH = 0.003 (normal = 0.47-5.00)
T4 = 20.3 (normal = 4.5-12)

Referral

Endocrine
Oculoplastics
Inform PCP of findings

5 months later

IOP 18/24
? APD OS
Start IOP Timolol 0.5%
Start Oral Pred (40 mg)

6 months later

IOP as high as 38/28
Oral Pred now 80 mg
+ APD OS
IOP 19/19 on Travatan, Cosopt, Alphagan
Refer for Orbital Decompression
After Orbital Decompression

Develops Diplopia
But...IOP 12/14 on meds

Last exam

Now S/P:
Orbital Decompression
Strabismus Surgery
Eyelid Retraction
Now has 20/80 cataract

History

72 year old African American male

Blur OD x 3 months

Last eye exam 10 years ago

Ocular History

Mixed Mechanism Glaucoma
S/P LPI OU
Was on Xalatan qhs OU – no longer taking
Blunt trauma OD

Medical History

HTN  Amlodipine
Anemia  Atenolol
CVA x 2  HCTZ
Hyperlipidemia  Simvastatin
Kidney Disease  ASA

Exam

BCVA OD: 20/320, OS: 20/25
+APD OD
Anterior Segment Normal
Except Mild NS OU
Gonio: Narrow with old PAS
S/P LPI OU
IOP 14/14
Posterior Segment

Optic Nerve

- 0.75 OD - Pallor
- 0.90 OS

No maculopathy
No vasculopathy
Peripheral retina normal

Pallor Vs. Excavation
**Differential**

- Glaucoma
- Other Optic Atrophy
- Traumatic
- Compressive
- Inflammatory

**What Tests?**

- VF
- OCT

**VF - OD**

**VF - OS**

**New Differential**

- Glaucoma
- Compressive Lesion
- CVA
**Diagnosis**

Pituitary Adenoma – 2.5 x 1.6 cm

**Treatment**

Monitor only

Due to other health factors

Patient reports vision is fine

**Treatment Goals**

- Normalize hormone levels
- Pituitary gland function
- Reduce signs/symptoms of tumor

**Treatment**

Medication (Micro)

- Bromocriptine – Dopamine agonist
- Hormone stabilization

Surgery (Macro)

- Transsphenoidal
- Transcranial
Take Home

- Check both eyes
- Pituitary vs. Glaucoma
- Pallor vs. Excavation
- Urgency

Optic Nerve Edema

1. Possible
2. Probable
3. Prognostic
4. Pragmatic

Goals

1. Understand Diagnostic Strategies
2. Learn Nuances of GCA
3. Develop Referral Strategy

Thank You!

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v
Macular Cherry-Red Spots: Causes and Consequences

2018 Victoria Conference
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Pacific University College of Optometry
Financial Disclosures: Nothing to Disclose
https://en.wikipedia.org/wiki/Cherry-red_spot

Learning Objectives
Today, we will explore the varied causes and consequences of cherry red spots on the macula:
1. To cover some congenital and systemic causes behind central retinal artery occlusion (CRAO)
2. To review prevention and treatment of CRAO
3. To explore the varied presentation of the genetically-inherited lysosomal storage diseases, including Tay-Sachs, Niemann-Pick, Gaucher, and Sandhoff diseases
4. To differentially diagnose traumatic causes of cherry red macular spots, like commotio retinae

Causes of Cherry Red Spots
1. Central Retinal Artery Occlusion
2. Tay-Sachs disease
3. Niemann-Pick disease
4. Other Causes (Gaucher, Commotio Retinae, Sandhoff disease)

CRAO: Diagnosis and Treatment
1. CRAO: Objective “Cattle Trucking”
2. CRAO: Fluorescein Angiography

https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4612404/
https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4764311/
CRAO: Cilioretinal Artery Sparing with Acute RNFL Defects

https://www.ncbi.nlm.nih.gov/pmc/article/PMC3822202/

CRAO: RNFL Improvement and Fluorescein Angiography

https://www.ncbi.nlm.nih.gov/pmc/article/PMC3822202/

CRAO Triggers: Cataract Surgery Retrobulbar Injection

https://www.ncbi.nlm.nih.gov/pmc/article/PMC5433131/

CRAO Triggers: Cataract Surgery Retrobulbar Injection

https://www.ncbi.nlm.nih.gov/pmc/article/PMC5433131/

CRAO Triggers: Chiropractic Manipulation

https://www.ncbi.nlm.nih.gov/pmc/article/PMC3325618/

CRAO Triggers: Chiropractic Manipulation

https://www.ncbi.nlm.nih.gov/pmc/article/PMC3325618/
CRAO Triggers: Chung-Strauss Syndrome

https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3714957/

CRAO Triggers: Chung-Strauss Syndrome

https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3714957/

CRAO Triggers: Pediatric Pneumonia

https://www.ncbi.nlm.nih.gov/pmc/articles/PMC5148912/

CRAO Triggers: Pediatric Pneumonia

https://www.ncbi.nlm.nih.gov/pmc/articles/PMC5148912/

CRAO Triggers: Congenital Single Heart Atrium

https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4612404/

CRAO DDx: Sickle Cell Anemia

https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4244145/
2. Tay-Sachs Disease

- One of the gangliosides, diseases of sialic acid-containing material found in neural tissue like gray matter
- If lysosomes don’t break these down efficiently, they accumulate in the brain and produce a spectrum of disorders, including a cherry-red spot in the macula
- But Tay-Sachs disease is simply the most famous of the gangliodisoses

http://en.wikipedia.org/wiki/Tay-Sachs

Tay-Sachs and the Gangliodisoses

- Tay-Sachs Disease (TSD), or acute infantile GM2, is often fatal by age 5
- It is named for British ophthalmologist Waren Tay and American neurologist Bernard Sachs first described the cellular appearance of the disease in the 1880’s
- It has a classic cherry-red macular spot that is pathognomonic in children

http://en.wikipedia.org/wiki/Bernard_Sachs

Tay-Sachs Cherry-Red Spot

- The red spot is essentially a choroidal window at the fovea compared to the fatty accumulation elsewhere on the retina
- The spot causes poor vision, that leads in turn to poor fixation
- Poor fixation commonly results in nystagmus because of larger-than-normal tremors of the eye
- Larger-than-normal drifts lead to strabismus as the eye goes to its phoric position in TSD


Ethnic Predilection and Late-Onset Tay-Sachs

- Recessive carriers of Tay-Sachs are found in at least 1 in 30 in each of the following ethnic groups:
  - Eastern European (Ashkenazi) Jews (eradicated in the US?)
  - French Canadians
  - Louisiana Cajuns
  - Irish Americans have a 1 in 50 chance of being a carrier
  - In the general population, the incidence of carriers is 1 in 300

Other Clinical Signs of Tay-Sachs

- Demyelination of optic nerve, chiasm and tracts
- Optic atrophy
- Progressive loss of vision
- Blindness by age 2
- Flatline VEP early, with normal ERG late
- Oculomotor ataxia

Degenerated Cherry Red Spot
Figure: 7.5B, page 374

Stalling Saccades in Tay-Sachs Disease

Stalling Saccades video from Leigh & Zee, 4th edition

Bottom Line on Tay-Sachs

- Some of these metabolic disorders are always fatal, others are not.
- All are autosomal recessive, meaning both the children of carrier parents are carriers themselves.
- The full disease in this family of conditions penetrate as often as about 1 in 4 of certain populations.
- Tear enzyme assay someday?

http://www.slideshare.net/aggabriel1/tay-sachs-disease-32430703

3. Niemann-Pick Disease

This loosely-knit group of metabolic diseases are characterized by abnormal accumulation of fats and cholesterol in visceral and neural tissue.

This autosomal recessive disease happens due to missing enzymes to break down body fats which accumulate in the spleen, liver, and eyes.

There are three identified types:
- Types A and B are caused by lipid buildup in myelin sheaths of nerve cells.
- Type C is caused by cholesterol accumulation.

Diagnosing Niemann-Pick Disease (NPD - VIDEO)

- Preschool children with NPD Type A first show “failure to thrive.”
- Next comes progressive vision loss and neurological deterioration.
- As in Tay-Sachs disease, NPD is characterized by cherry-red macular spots and eye movement disorders (shown here).

Diagnosing Niemann-Pick Disease (NPD - VIDEO)

http://bcove.me/oe9b02h6

The Five Known Types of Niemann-Pick Disease

- The ophthalmic hallmark of NPD Type C is progressive supranuclear vertical gaze palsy.
- Look for hard blinks and head thrusts with vertical eye movements especially.
- Oculomotor and other striated muscle ataxia is common, as are learning disabilities.

http://en.wikipedia.org/wiki/Tay-Sachs
Other Ocular Manifestations of Niemann-Pick Disease, Type A

- Mild corneal haze
- Fine lenticular deposits
- Cherry-red spot (50%)
- Retinal "haze" that extends far beyond the fovea
- Central vision loss, occurring later in the disease (age 2)

http://imagebank.asrs.org/file/8649/niemann-pick-disease-type-b

Niemann-Pick Disease: Type B vs. Type C

- Type B is mostly respiratory, has less neurological involvement, and survival into adulthood
- Expect orbital congestion due to increased orbital fat
- A macular halo may be seen instead of a red spot in Type B
- Not usually associated with vision loss (normal BCVA)

Type B is mostly respiratory, has less neurological involvement, and survival into adulthood in Type C, cholesterol-laden "foam cells" accumulate, classically leading to:

- Hepatosplenomegaly, all starting in late childhood
- Progressive dementia or intellectual disability
- Ataxia
- Dystonia
- Vertical gaze paresis

CNS Effects of Types B and C Niemann-Pick Disease

- In type B, patients can develop psychosis due to accumulation of myelin in the central nervous system
- Since Type B patients survive well into adulthood, when these mental health disorders emerge, they have to be managed, sometimes surgically
- Seen here is the loss of gray matter in the brain of a Niemann-Pick Type C patient

https://neurowiki2012.wikispaces.com/Niemann-Pick+Disease

Signs and Symptoms of Niemann-Pick Disease, Type C

- In terms of eye effects, you can expect at least 4 out of 5 Niemann-Pick patients to exhibit:
  - Oculomotor ataxia
  - Vertical gaze palsy
  - Learning disabilities and visual-perceptual problems
  - Some have mental health concerns

https://neurowiki2012.wikispaces.com/Niemann-Pick+Disease

Treatments for Niemann-Pick Disease

- Like Tay-Sachs, there is currently no known treatment for any of the three types of Niemann-Pick
- Types B and C have much greater longevity, and may be managed pharmacologically or surgically in some cases
- These diseases can be slowly progressive and take a while to diagnose

http://imagebank.asrs.org/file/8648/niemann-pick-disease-type-b

4. Other Lysosomal Storage Diseases

https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4802848/
Other Causes of Macular Cherry Red Spots

- Gaucher disease
- Trauma (Commotio Retinae)
- Sandhoff disease
- Others: Sialidosis

https://youtu.be/7zmbMM07M6Q

Other Causes of Cherry Red Spots: Gaucher Disease

https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3831135/

Other Causes of Cherry Red Spots: Commotio Retinae

https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4185168/

Other Causes of Cherry Red Spots: Sandhoff Disease

Other Causes of Cherry Red Spots: Sandhoff Disease


Other Causes of Cherry Red Spots: Sialidosis

A 53-year-old man, with non-consanguineous parents, presented to our hospital with a history of progressive decrease of visual acuity since the age of 26.

At 36, he developed generalized myoclonus and ataxic gait.

He showed low visual acuity, ataxic gait, dysarthria and difficulty in writing.

https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4202095/

Other Causes of Cherry Red Spots: Sialidosis

https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4802848/

Summary: Cherry Red Spots

https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3864975/

Questions? Thank You!

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Readings and References

- For more information on Niemann-Pick and Tay-Sachs diseases, see chapter 7 of Wright’s Handbook of Pediatric Eye and Systemic Disease.
Recognizing “Red Flag” Headaches

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Various Classification Systems

• Vascular Headache
• Myogenic H.A. (“muscle contraction”, “muscle tension”)
• Cervicogenic H.A.
• Tractional H.A.
• Inflammatory H.A.

International Classification of Headache Disorders, 2nd ed. (ICHD-II)
-- International Headache Society
-- Cephalgia. 2004;24:1-160

“Any good references, Doc?”

• CEPHALGIA
  – Journal
  – Online

Source 2

• Tension H.A.
• Migraine H.A.
• Ictal H.A.
• “Brain Freeze” H.A.
• Thunderclap H.A.

• Vascular H.A.
• Coital Cephalgia
• Sinus H.A.
• Rebound H.A.
• Red Wine H.A.

PRIMARY vs. SECONDARY H.A.’s

• PRIMARY
  – USUALLY “safe”
  – Principally NEUROCHEMICAL cause

• SECONDARY
  – MAY indicate a significant problem
  – Associated with “pulling”, “pushing”, inflammation, or ischemia of a Pain-sensitive structure
Most Headaches are Harmless

- Most headaches are self-limiting
- Most headaches remain “idiopathic”
- Most headache sufferers get more than one form of headache

The “Pearls”

- “….Headache by itself tells you nothing…."
- “What is the company that headache is keeping???”
- “What else is going on???”

Assessing Severity

- “Scale of 1 to 10…..”
- What Can't You Do Because of this Headache?
- NOT USEFUL: “Do you get relief from over-the-counter pain medications?”

New Form of Headache

- In elderly patients….
  R/O CRANIAL ARTERITIS

BEWARE!!!!!

…..BUT I NEVER, EVER HAD A HEADACHE LIKE THIS ONE BEFORE…..

BEWARE !!!!!!

…..AND ALONG WITH MY HEADACHE (XXX) ISN'T WORKING RIGHT, OR (YYY) HAS STARTED TO HAPPEN TO ME…..”
Quiz:

The most important assessment for an Optometrist to make on a Headache patient is?

Examine the Eye(s) !!!!!

- OD and OS
- OU
- The Oculo-Cranial Nerves

RED FLAG!

- Very recent onset…
- Acutely explosive pain…
- Intense, “Thunderclap-like…”
- “Unlike anything ever before…”
  - Check Pupils!
  - Check Motilities?
  - Check Vitreous?

Quiz 2

List the Cranial Nerves most often affected by elevations in Intracranial Pressure (elevated C.S.F. pressure)

- C#2
- C#3
- C#6
- C#8

Mechanism of Headache

<table>
<thead>
<tr>
<th>P</th>
<th>A</th>
<th>I</th>
<th>N</th>
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<tbody>
<tr>
<td>Threshold</td>
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Mechanism of Primary Headaches

Threshold lowered

Mechanism of Secondary Headaches

Neuro Input Raised

Primary Headache’s Componants

- Neurochemical
- Neurovascular
- Vascular

CLINICAL EVALUATION of the Headache Patient

#1: “The History is Where It’s At” J. Lawton Smith, MD

#2: Ancillary Testing is Indicated for All Suspected Secondary Headache Cases

#3: Imaging Studies are considered Standard Level of Care in most areas
   → MINIMUM: CT of the Head without enhancement

#4: Blood Tests are considered Standard Level of Care in most areas
   → ESPECIALLY FOR PATIENTS OVER AGE 50!!!

“Pearls” for H.A. Workups

- Check blood pressure in ALL cases
- H.A. or Face Pain is Giant Cell Arteritis in all patients > 50….until proven otherwise!
- Ocular evaluation should always include:
  - assessment of corneal sensitivities
  - IOP
  - gonioscopy
  - evaluation for cell&flare
  - careful assessment of both optic nerves
    -- always look for spontaneous venous pulsations

What About Visual Fields?????

- Sometimes helpful; Oftentimes not
- If you run them—make them simple
- They never finalize a lesion’s location/
  They are not a substitute for imaging
QUIZ: What % of Headache Patients have normal Visual Fields on initial fields testing?

A. Less than 25%
B. 25 – 49%
C. 50 – 74%
D. 75 – 90%

QUIZ: What % of Brain Tumors become associated with Visual Field defects at some point during their development?

A. Less than 10%
B. 20 – 30%
C. 40 – 50%
D. 75 – 85%

Always Check Pupils

• “Blown” Neurologic Pupil
• Sudden-onset, painful, Horner’s Pupil

Always Check Motilities

• Large Amplitude, Rapid Horizontal Saccades
  – ABdutional ability of each eye (6th nerve)
  – ADductional ability of each eye (3rd nerve & M.L.F.)
    • Test horizontal gaze ADduction
    • Test convergence ADduction

Always Check Motilities

• Test horizontal gaze ADduction
• Test convergence ADduction

Try to determine Temperature

• Thermometer
• Patient’s Report
  – Signs & Symptoms

Check Neck Flexibility

• “Very Stiff and Painful” = Need to Rule-out:
  – Neck trauma
  – Meningitis
  – Subarachnoid Hemorrhage

• “Blown” Neurologic Pupil
• Sudden-onset, painful, Horner’s Pupil
Palpate Temporal Arteries (if cranial arteritis not ruled-out)
- Swollen?
- Hard?
- Tender?

→ “Hurt to comb your hair?”
→ “Sore at base of head?”
→ “Jaw claudication?”
→ “Unexpected weight loss?”
→ “Night sweats?”

May want to add:
- Tick Panel
- Sickle Cell Test
- Drug Screen – Blood
- Urine Toxicology

Additional Testing (when the Hx says, “Keep looking”)
- All Highly Acute Presentations
- All Rapidly Progressing Cases
- All Cases of Disc Edema
- All Cases with Neurologic Findings

CT Scan of the Head
- 1st: WITHOUT contrast
  - Best for “fresh blood”
  - Best for hydrocephalus
- 2nd: WITH contrast
  - Enhances soft tissue lesions
  - Recommend obtaining blood urea nitrogen and serum creatinine tests in “at risk” kidney patients!!!

Blood Tests
- Complete Blood Count with Differential
- Platelet Count
- Erythrocyte Sedimentation Rate
- C-reactive Protein
- Serum Fibrinogen (?)

MRI of the Brain (and orbits?)
- What sequence to order???
  - MRI-brain, Stroke Protocol
  - MRI-brain, Parenchymal Study
  - MRI-brain, Vascular Study
  - MRI-brain, Cranial Nerve (#) Emphasis
  - T-1 with Fat Suppression for orbital exam
  - T-2 with FLAIR for exam of periventricular tissues (vital in M.S. workup)
Lumbar Puncture
• Cerebrospinal fluid opening pressure
• With C.S.F. Analysis

KEYPOINT:
Imaging indicated prior to Punctures!!!

r/o INTRACRANIAL MASSES
r/o ARNOLD-CHIARI SYNDROME

Brain & Neck M.R.A.
• When arterial lesions are suspected, but M.R.I.-brain was negative/inconclusive

Brain & Neck M.R.V. indicated if venous disease is suspected
– Differentiating Indiopathic Intracranial Hypertension from Cerebral Venous Thrombosis

Well-guided Referral
• Neurologist
• Neurosurgeon
• Neuro-ophthalmologist
• Family Practice Physician
  – Comprehensive physical exam
  – No “frightening findings”
The Critical Clinical Tool:
CASE HISTORY

“What Else is Going On?”
• Any Triggering Events?
• Any Trigger Points?
• Other Painful Areas
• Changes in Head or Body Functions?
• NAUSEA? VOMITING?
• CHANGES IN VISION?

The “Big Three”
• MODE of Onset
  – Sudden, Acute vs. Progressive
  – Sudden “Explosion” vs. Prolonged Buildup
• WHEN & HOW 1ST Noticed
  – Initial features
• TIMELINE (Progression over Time)
  – Episodic & Intermittent vs. Constant

The Useful “Fourth”
• INTENSITY
  – “How bad on a scale of 1-10?"
  – “How disruptive is it?”
  – “What can’t you do because of your headache? (Face pain, eye pain, etc.)”

Many Migraine Patients Demonstrate RAYNAUD’S PHENOMONON

Helpful “Fifth”
• CONSTANT vs. INTERMITTENT
  – “How long does each attack last?”
  – “How long is each episode?”
  – “Are the attacks Similar or Different?”
  – “Do you know when an attack is coming on?”
Other Useful Questions

• Experiencing Visual Phenomena?
• Light Sensitivity?
• Odor Sensitivity?
• Sensitive to Noise/Sounds?
• Any Ringing in the Ears?

Treatments Tried?

• Did they work?
• Do they still work?
• Who prescribed them?

REBOUND HEADACHE

• “Medication Overuse (MOU) Headache”
• May involve Addiction
• Does involve Dependency
• MANAGEMENT
  – Patient education
  – Do NOT “cold turkey”
  – DO Taper off…..

RED FLAG!

• Very recent onset…
• Acutely explosive pain…
• Intense, “Thunderclap-like…”
• “Unlike anything ever before…”
  – Check Pupils!
  – Check Motilities?
  – Check Vitreous?

RED FLAGS!

• HEADACHE + NECK PAIN + SHOULD PAIN + STIFF NECK
• HEADACHE/FACE PAIN + SHOULD PAIN + ARM PAIN + HEAVY SWEATING
• SUDDEN-ONSET, HYPER-INTENSE “THUNDERCLAP” HEADACHE (especially following a valsalva maneuver"

RED FLAGS!

• HEADACHES ASSOCIATED WITH SEIZURES or FOCAL SEIZURES
• HEADACHES ASSOCIATED WITH LOSS OF CONSCIOUSNESS or WITH CONFUSION
Ictal Headaches
- "Headache Associated with Seizures"
- Rarely occur simultaneously
  - Pre-ictal HA
  - Post-ictal HA
- Intensity varies by patient, and by episode
- Some patients experience hallucinations
  - Unusual thoughts
  - Unusual sensations
  - "Neurological Aura"

Orange-Red Flag!
- Steadily building…
- Increasing intensity…
- Steadily progressing…
- Steadily more intrusive…
- “Started several days (or weeks) ago…relentless. Just won’t stop.”

Post-Migraine Headache
- Exhaustion
- Mood Alteration
- Cognitive Alteration
→ Usually resolves in 24-48 hours

Yellow Flag
- Episodic pain…
- Definite pain-free intervals…
- “Has been going on for awhile…”
- Stereotypical for that patient
Coatings and Treatments and Agents Oh My!
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Course Description
The course will review the state of the contact lens industry and how contact lens dropout affects the growth of the contact lens modality. Minimizing contact lens discomfort may be key in reducing dropout rates. The role of contact lens treatments and coating will be explored to see if this may be the key to keeping patients comfortable in contact lenses.

Course Learning Objectives
After participating in this course, the participant should be able to:
• Be familiar with the growth and dropout rates in the different contact lens modalities
• Understand the role of contact lens dropout and what fit and material factors may contribute to dropout
• Understand coefficient of friction and how the industry is trying to improve contact lens discomfort
• Be familiar with how lens care products may help or hinder the comfort of a contact lens
• Understand the distinction between plasma treatment and a true coating of a contact lens

Course Outline
• State of the Industry
  o 40.9 million Americans (16.7% of the US adult population) wear contact lenses
  o Soft contact lens (SCL) market is growing
  o US CL market $2.7 billion
    ▪ Steady growth but slowing?
  o Contact lens wearers make up 36% of a typical practice
    ▪ Gross revenue: 32%
    ▪ Net Profit: 27%
  o The CL industry is healthy
    ▪ Most practitioners (67%) believe that CL will grow in their practice
  o What are we fitting?
    ▪ Soft contact lenses: 88%
    ▪ Gas permeable: 9%
    ▪ Other: 3%
  o Replacement Schedule
• Dropout
  o A sustained or permanent discontinuation of CL wear
  o New fits = 29%
  o Mean dropout in the USA ~16%
  o Higher in other parts of the world
• Improving
  • Primary reason for discontinuing wear is discomfort

  • A Contact Lens and The Eye
    • A well fit soft contact lens
    • The average eyelid

  • Soft Contact Lenses
    • Materials
      ▪ 29% Hydrogel
      ▪ 71% Silicone hydrogel
    • Hydrogel (polyhydroxyethylmethacrylate or pHEMA)
      ▪ Start as a cured, hard plastic lens that absorbs water and forms a soft, flexible material
      ▪ Oxygen permeability dependent on the water content
    • Silicone hydrogel
      ▪ Combines hydrogel monomers with silicone
      ▪ Oxygen permeability dependent on the silicone content

  • Interactions of the CL and the Eye
    • Lid
      ▪ Lid Wiper Epitheliopathy
      ▪ Contact Lens Papillary Conjunctivitis
    • Cornea
      ▪ Epithelial
      ▪ Stromal – edema
      ▪ Endothelial changes – Morphologic and density changes
    • Conjunctiva
      ▪ Bulbar

  • What Is CL Discomfort
    • Difficult to define
    • Lack of clinical signs that correspond with symptoms
    • No pre-existing dry eye condition
    • Approximately 50% of CL wearers
    • TFOS Definition
      ▪ A condition characterized by episodic or persistent adverse ocular sensations related to lens wear, either with or without visual disturbance, resulting from reduced compatibility between the contact lens and the ocular environment, which can lead to decreasing wearing time and discontinuation of contact lens wear.

    • Multifactorial
    • Symptoms
    • Identifying the Problem
      ▪ Contact lens related
      ▪ Patient related
      ▪ Environmental

  • Contact Lens Related
    • Coefficient of friction*
    • Oxygen permeability
- Wettability
- Surface modification
- Modulus
- Dehydration
- Replacement frequency
- Design
- Thickness
- Edge design

- Contact Lens Related
  - Improved comfort with…
    - Low water content
    - More frequent replacement
    - Thin designs
    - Thin, tapered edges
    - Less lens movement on blink
  - Deposits
    - Active lysozymes (major tear protein) had a significant positive correlation with subjective comfort
    - Tear dried material can change the friction force
  - Coefficient of Friction
    - Force that prevents one surface from sliding over another
    - Tribology
      - Microtribometer
      - Atomic force microscopy method
      - Inclined plane method
      - Finger lubricity method
    - Factors impacting measurements
      - Substrate choice – should mimic the chemistry, roughness and softness of the ocular surface
      - Dynamic contact
      - Force pressure
      - Sliding speed – generally test at a lower speed (CoF is usually highest)
      - Sample preparation
      - Lubrication fluid – try to mimic the tears but no standard composition
    - Lubricity
      - Visual acuity
      - Physiological
      - Treat the lens
      - Submerge the lens
  - Treat the Lens
    - Silicone hydrogel naturally hydrophobic
      - Surface treatment – gas plasma technique
        - Plasma coating (25nm) – N&D
• Plasma oxidation - PV
• Plasma coating + oxidation - Miru
  • Internal wetting agent
  • Inherent lens properties - Biofinity
    • Bind to water molecules
  • Submerge the Lens
    • Lens Care Products
      • Preservative – increases disinfection efficacy
      • Surfactant – improve cleaning
      • Viscosity – buffer the ocular tissue from the preservative and surfactant
      • MPS vs. H₂O₂
        • Poor compliance
• Compliance - Replacement Schedule
• Compliance – Lens Care Products
• What LCP are we recommending?
  • Topping off
  • Rub vs. No Rub
• Patient related
  • Non-modifiable
  • Modifiable?
• Environmental
  • Few well controlled studies
  • Low humidity (RH <30%)
  • Low humidity + Temperature
  • Wind and airflow – exacerbate evaporation
  • Occupation
• The Eye or the Lens?
  • Effects of Three Interventions on Contact Lens Comfort in Symptomatic Wearers: A RCT (Navascues-Cornago 2015)
    • Three interventions after 5 hours of CL wear
    • None of the interventions had a meaningful impact on end-of-day comfort
• Plasma Treatment
  • Gas permeable lenses
  • Not a coating
  • Front surface of the lens is sterilized via exposure to cold plasma gas in a reaction chamber
    • Removes all residue = clean and wettable lens surface
  • Improved initial comfort
    • Wears out with time
    • Avoid abrasive cleaners
• A True Coating
  • Hydra-PEG (Ocular Dynamics – Menlo Park, CA)
  • Tear Film
- Lubricates, moisturizes, oxygenates, cleans and protects the ocular surface
- Mucin
  - Hydra-PEG Study
    - Habitual SCL wearers with self-reported symptoms of CLD
- Contact Lens Comfort
  - Personalize prescribing
  - Innovations in the area of material, design, lens care
  - Novel approaches to contact lens comfort
**Rethinking Gonioscopy**  
Fundamentals and Future Tech  

Anthony DeWilde, O.D.

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**Goals**

- Misconceptions about angle closure
- Prognosis/Treatment
- New Technology

---

**Indications**

- Elevated IOP
- Asymmetric IOP
- Vascular  
  - CRVO, DM, OIS
- Trauma
- Every Glaucoma patient!!

---

**Contraindications**

- Hyphema?
- Open Globe
- Compromised Cornea

---

**Indications**

- Every Glaucoma patient!!
- Role of IOP = assess risk
- Role of gonioscopy = determine treatment

---

**Underutilized**

- Only 50% of Glaucoma patients have gonio recorded
- 74% of referred patients had no angle status

Am J Ophthalmol 2018;188:16–29
Underutilized

- Don’t understand value
- Difficult to handle equipment
- Difficult to interpret

Subacute

- Most primary angle closure is subacute
- Spend months to years asymptomatic
- May not catch during exam
- Acute closure is uncommon

Risk of closure

- Not all angle closure is acute
- Not all acute glaucoma is angle closure


Not all angle closure is acute
- Most is subacute

Not all acute glaucoma is angle closure
- Rubeosis
- Uveitis
Difficult

Technically difficult to handle
Practice
Few good references

gonioscopy.org
eyecalcs.com

Difficult

How Do Angles Close?

Risk Factors

Age
Race (Asian, Eskimo)
Shorter Axial Length
Shallow Anterior Chamber
Lens

Accuracy

Classify type of glaucoma
Leads to better treatment
Accuracy

POAG
NTG
Angle Closure
Rubeotic
Uveitic
Pigmentary/Pseudoexfoliation

Accuracy

Many different methods
Van Herrick
Shaffer
Spaeth

Why Change?

Gonioscopy should grade occludability

Why Change?

Spaeth tells us
Oculudability
Relationship of Iris to TM
Easier to monitor for change

Spaeth Method
Spaeth Method

**Normal Angle - Video**

Video from gonioscopy.org - Used with written permission

---

**Angle of Insertion**

---

**grade the angle**

---

---
Steep or Regular

Go to most narrow angle
A = Anterior to TM
B = Behind TM
C = Scleral Spur
D = Deep (Ciliary Body)
E = Very Deep

Compression

If angle is narrow, may need compression
How far back does it go?
Check for PAS
Compression

Compression Gonioscopy - Video

Video from gonioscopy.org - Used with written permission

Occludable

Who is occludable?
30 and 40 are not
10 and 20 are

Not all occludable angles will occlude

Who needs treatment?

No controlled clinical trials

Not everyone needs treatment

Only 10% of occludable angles

Who needs treatment?

Increased IOP

ONH and/or VF progression

PAS - current or aborted

Symptoms

Other eye
Provocative Tests

Dark Room
Water
Dilation
Prone

All test suffer from
Poor specificity
Tedious

Provocative Tests

Dark Room
Gonio (after 1.5 hours)
66% sensitivity
80% specificity

OCT (after 3 minutes)
90% sensitivity
57% specificity

J Glaucoma 2012 Mar;21(3):155-9

Provocative Tests

Friedman - AJO 1972

Sensitivity
Dark room 48%
Prone 71%
Pharmacologic 58%
Treatment

- Laser Peripheral Iridotomy
- Argon Laser Iridoplasty
- Cataract Surgery

Acute Treatment

- Diamox not fast enough
- Isosorbide and Osmoglycin not available
- Paracentesis
- Cannot do this if angle is narrow

Acute Treatment

- Cornea is edematous
  - Underestimate by as much as 20%
- If cornea is clearing, IOP is improving

Acute Treatment

Meds!
- Consider Iopidine

Quigley

- Two factors influence risk of closure
  - Iris proximity to TM
  - Iris volume
Possible mechanisms:

- Iris volume increase on dilation
- Choroidal expansion

**Advanced Tech**

Ultrasound Biomicroscopy (UBM)
Scheimpflug photography
Anterior Segment OCT (ASOCT)

*Ophthalmology 2013;120:1985-1997*

**Benefits**

- Good visualization of angle
- Documentation
- Patient education

**UBM**

- Similar to B-Scan
- Uses higher frequency
- Images anterior segment

**Pros**

- Quantitative/Qualitative view of ACA
- Correlates well with gonioscopy
- Plateau Iris
- Confirm efficacy of LPI
UBM

Cons
Best imaging requires coupling with eye bath
Inconvenient
May be difficult to interpret

Scheimpflug

Anterior Segment images from slit lamp
Noncontact optical system
Common system is Pentacam

Scheimpflug

Pros
Inexpensive
Good correlation with gonioscopy

Scheimpflug

Cons
Not as good as OCT or UBM
No view of angle structures

ASOCT
Objective measures
- Iris curvature
- Lens vault
- Iris volume
- Anterior chamber depth
- Anterior chamber width

**Pros**
- Can do in dark room
- Good sensitivity
- Many doctors familiar with OCT

**Con**
- May overestimate risk
- Questionable specificity
- Cannot visualize behind iris

**Pros**
- Good visualization - even with corneal pathology
- Good on uncooperative patients

**Con**
- Uses scleral spur as landmark, not TM
- Cost
ASOCT

Am J Ophthalmol 2018;188:16–29

Imaging

All technology is a complement to gonio
Cannot visualize angle as well as gonio
Cannot compress

Imaging

Need prospective trials
LPI vs Monitoring
None can predict which angles close

anthony.dewilde@va.gov
Eclipses, Climate Change, & the Eye

2018 Victoria Conference
Pacific University
College of Optometry

James Kundart OD
MEd FAAO FCOVD-A

Financial Disclosures:
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https://en.wikipedia.org/wiki/Photokeratitis

Learning Objectives
After this course, the practicing optometrist will know how to prevent, differentially diagnose, and treat (when possible):
1. Heat-induced cataract and exfoliation
2. UV-induced pterygia
3. Climatic droplet keratopathy
4. Retinal snowblindness
5. Solar and eclipse retinopathy

Ultraviolet Radiation and Climate Change

<table>
<thead>
<tr>
<th>UV band</th>
<th>Wavelength (nm)</th>
<th>Availability</th>
<th>% absorbed by cornea</th>
<th>% absorbed by aqueous</th>
<th>% absorbed by lens</th>
</tr>
</thead>
<tbody>
<tr>
<td>UV-A</td>
<td>315-400</td>
<td>Primarily % V6A absorbed</td>
<td>45 (at 220 nm)</td>
<td>16 (at 300 nm)</td>
<td>26 (at 310 nm)</td>
</tr>
<tr>
<td>UV-B</td>
<td>280-315</td>
<td>Intermittent corneal absorbed</td>
<td>37 (at 540 nm)</td>
<td>14 (at 350 nm)</td>
<td>6 (at 300 nm)</td>
</tr>
<tr>
<td>UV-C</td>
<td>100-280</td>
<td>About all absorbed by cornea</td>
<td>100</td>
<td>0</td>
<td>0</td>
</tr>
</tbody>
</table>

https://www.ncbi.nlm.nih.gov/pmc/articles/PMC5820813/

Which Climate Variables Affect Health Care? UV!

Heat-Induced (Glassblower’s) Cataract and True Exfoliation Case #1

UV Effects: Basal Cell Carcinoma of the Eyelid


Heat-Induced (Glassblower’s) Cataract and True Exfoliation Case #1

https://www.jstage.jst.go.jp/article/jnms/74/1/74_1_55/_pdf/-char/en

UV Effects: Pterygia

The Chesapeake Bay Study (1989) of 838 watermen in Maryland found those with the highest quartile of UV-A and UV-B exposure had a 3x higher odds ratio for pterygia


Treating Pterygia with Topical Azasite Case #1


Heat-Induced (Glassblower’s) Cataract and True Exfoliation Case #2

https://www.jstage.jst.go.jp/article/jnms/74/1/74_1_55/_pdf/-char/en

Treating Pterygia with Topical Azasite Case #2

Treating Pterygia with Topical Azasite Case #3

Treating Pterygia with Azasite $185 a bottle with coupon

UV Effects: Climatic Droplet Keratopathy

Climatic Droplet Keratopathy #1

Climatic Droplet Keratopathy #2

Climatic Droplet Keratopathy #3
How Other Species Prevent Snowblindness

1. The concentration of vitamin C in the corneal epithelium of reindeer is 2x higher than that found in humans.

2. Reindeer live in higher elevations (>100m above sea level), where there is higher UV exposure.


Case #1: 58 YOM with UV Retinopathy (Snowblindness)

https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4801146/

Case #1: 58 YOM with Snowblindness Macular OCT 5-Line Raster

https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4801146/

Case #1: 58 YOM with Snowblindness Autofluorescence

https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4801146/

Erythropsia in Mountain Climbers

https://julbo-canada.ca/blogs/news/33320936-the-most-common-eye-disorders-found-in-climbers

Prevention and Treatment of Snowblindness

http://www.mrcophth.com/ophthalmologyonstamps/phototoxicity/c.html
Phototoxicity and the Retina
M A Conti, M M Benedetto, M L Quintana-Quintana, and M E Guido
Light pollution: the possible consequences of excessive illumination on retina
https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4763120/

Case #2: Solar Retinopathy from Bipolar Disorder in 45 YOF
https://www.ncbi.nlm.nih.gov/pmc/articles/PMC1880187/

Case #2: Time-Domain OCT
https://www.ncbi.nlm.nih.gov/pmc/articles/PMC1880187/

Case #3: Baseline Sungazing Retinopathy
(A) Right eye at initial presentation showing a retinal pigment epithelial defect at the fovea
(B) Left eye at initial presentation showing a retinal pigment epithelial defect at the fovea
https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3604317/

Sungazing Retinopathy: One Month Follow-Up
(A) Right eye at 1 month: persistent retinal pigment epithelial (RPE) changes at the fovea
(B) Left eye at 1 month: persistent RPE changes at the fovea
https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3604317/

Sungazing Retinopathy: Six Month Follow-Up
Right eye at 6 months: improvement in foveal changes and
(B) left eye at 6 months: improvement in foveal changes
https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3604317/
Sungazing Retinopathy: One Year Follow-Up

(A) Right eye at 1 year; almost full resolution of retinal pigment epithelial (RPE) changes
(B) Left eye at 1 year; almost full resolution of RPE changes

Case #4: 38 YOWF Solar Retinopathy
Without Abnormal Exposure, Baseline and Six Months Later

Case #5: 21 YOM Solar Retinopathy OD

Case #5: 21 YOM Solar Retinopathy Fluorescein Angiography

Case #5: 21 YOM Solar Retinopathy Time-Domain OCT OD
Case #5: 21 YOM Solar Retinopathy
Spectral-Domain OCT OD
https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3149141/

Pediatric Case #6: 10 YOM
Solar Retinopathy OS
https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3149141/

Case #6: 10 YOM Solar Retinopathy
Fluorescein Angiography OS
https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3149141/

Case #6: 10 YOM Solar Retinopathy
Time-Domain OCT OS
https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3149141/

Eclipse Retinopathy Case #7: 29 YOM
https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4213866/
Eclipse Retinopathy Case #8: 29 YOF

Pediatric Eclipse Case #9: 13 YOF

Pediatric Eclipse Case #10: 14 YOF

Case #11: 63 YOF Eclipse Retinopathy

Case #11: Eclipse Retinopathy Auto-fluorescence

Case #11: Eclipse Retinopathy Baseline Macular OCT
Case #11: Retinopathy Epiretinal Membrane 1 Mo. Later
https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3545138/

Case #11: Eclipse Retinopathy Vitrectomy 4 Mo. Later
https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3545138/

Pediatric Case #12: Eclipse Retinopathy 10 YOM
https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3545138/

Pediatric Case #13: Eclipse Retinopathy in 8 YOF
https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3545138/

Pediatric Case #14: Eclipse Retinopathy in 11 YOF
https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3545138/
Treating Solar Retinopathy

There is no standardized treatment for solar retinopathy, but antioxidant vitamins and micronutrients similar to AREDS (vitamin A, C, E, lutein, zeaxanthin, and zinc) have shown promise. Oral prednisolone has been tried, but it is a risk factor for central serous chorioretinopathy.


THANKS FOR THE MEMORIES, MS. JEANNE OLIVER!