Slide 1

AN OCULAR GRAND ROUNDS
TOUR OF ORLANDO

John Spalding, OD, FAAO
Orlando, FL

NO FINANCIAL DISCLOSURES!

Slide 2

CASE

• 38 / AA / F
• CC
• No changes in vision, no ocular comfort problems
• Oc Hx
• LEE 18 mos, unremarkable
• Med Hx
• DM (gestational, still on meds)
• HTN, anemia
• Fam Hx
• unremarkable
• Soc Hx
• - tobacco, alcohol

BVA
2020 OD: 455-079-6625
2020 OS: 455-079-6135
PRELIMS
ETEC OD OS, FROM
Normal Pupils, No APD
SLE:
OD corneal scar, OS normal
IOP:
18/18 mm Hg @ 1p
DFE
See Photos

Slide 3

DDX OF ONH EDEMA

<table>
<thead>
<tr>
<th>CONDITION</th>
<th>UNILATERAL / BILATERAL</th>
</tr>
</thead>
<tbody>
<tr>
<td>OCULAR DISEASE (uveitis, hypotony, vein occlusion)</td>
<td>UNILATERAL / BILATERAL</td>
</tr>
<tr>
<td>INFLAMMATION (papillitis, neuroretinitis, papillophlebitis)</td>
<td>UNILATERAL</td>
</tr>
<tr>
<td>INFILTRATIVE (lymphoma, reticuloendothelial)</td>
<td>UNILATERAL</td>
</tr>
<tr>
<td>DISC TUMORS (hemangioma, glioma, metastatic)</td>
<td>UNILATERAL</td>
</tr>
<tr>
<td>VASCULAR (HUN, arteritis)</td>
<td>UNILATERAL</td>
</tr>
<tr>
<td>ORBITAL TUMORS (meningioma, glioma, cysts, retrobulbar mass)</td>
<td>UNILATERAL</td>
</tr>
<tr>
<td>SYSTEMIC DISEASE (anemia, hypoxemia, HTN)</td>
<td>UNILATERAL OR BILATERAL</td>
</tr>
<tr>
<td>METABOLIC (dysthyroidism, type 1 DM, proliferative retinopathies)</td>
<td>BILATERAL</td>
</tr>
<tr>
<td>ELEVATED INTRACRANIAL PRESSURE (mass, HTN, IIH, etc.)</td>
<td>BILATERAL, MAY BE ASYMMETRIC</td>
</tr>
<tr>
<td>LEBER'S HEREDITARY OPTIC NEUROPATHY</td>
<td>BILATERAL OR NORMAL</td>
</tr>
<tr>
<td>CONGENITAL DISC ANOMALY (tilted ONH, hypoplastic)</td>
<td>BILATERAL, RARELY UNILATERAL</td>
</tr>
<tr>
<td>PSEUDOPAPILLEDEMA (hyperopia, ONH drusen)</td>
<td>BILATERAL, MAY BE ASYMMETRIC</td>
</tr>
<tr>
<td>ELEVATED INTRACRANIAL PRESSURE (mass, HTN, IIH, etc.)</td>
<td>BILATERAL, MAY BE ASYMMETRIC</td>
</tr>
</tbody>
</table>
Slide 4

WHAT’S NOT ON THE LIST?

- “PAPILLEDEMA”
- A TERM FOR...
  - BILATERALLY SWOLLEN OPTIC NERVES
  - WITHOUT THE WORK-UP…
  - YOU CAN’T CALL IT THAT YET

Slide 5

INCREASED INTRACRANIAL PRESSURE

- CAUSES
  - INTRACRANIAL MASS LESION
  - CEREBRAL EDEMA
  - DECREASED CSF ABSORPTION
  - OBSTRUCTIVE HYDROCEPHALUS
  - OBSTRUCTION OF VENOUS OUTFLOW
  - MALIGNANT HYPERTENSION
  - IDIOPATHIC INTRACRANIAL HYPERTENSION
  - PSEUDOTUMOR CEREBRI

Slide 6

EYE CLINIC WORK-UP

- EYE CLINIC
  - TESTING
    - PHOTOS
    - BSCAN, OCT
    - VF
  - BLOOD PRESSURE
    - 139/83
  - CHART REVIEW
    - 5’3”, 179 lbs
    - BMI = 31.8 (obese)
EMERGENCY ROOM WORK-UP

- MRI LOOKING FOR
  - INTRACRANIAL MASS LESION
  - OBSTRUCTION OF VENOUS OUTFLOW
  - OBSTRUCTIVE HYDROCEPHALUS
  - DECREASED CSF ABSORPTION
  - INCREASED CEREBROSPINAL FLUID
- CT IF MRI DELAYED OR CONTRAINDICATED

EMERGENCY ROOM WORK-UP

- MRV LOOKING FOR
  - VENOUS OBSTRUCTION IN DURAL SINUSES OR NECK
- IF ALL NORMAL (NOT BEFORE)
  - LUMBAR PUNCTURE
    - OPENING PRESSURE
      - IF > 25 CM (250 MM H2O) THAT’S ELEVATED
      - 20-25 IS EQUIVOCAL
    - CSF ANALYSIS
      - LOOKING FOR SIGNS OF INFECTION

ER RESULTS

- BP
  - 132/77, 157/104
- MRI / MRV
  - SAME DAY AT LOCAL HOSPITAL
    - “NORMAL” PER THE PATIENT
    - NO ACTUAL WRITTEN REPORT
    - DISCHARGED BY HOSPITAL NEXT DAY
    - RX: ANOTHER BP MED
  - NO LUMBAR PUNCTURE WAS DONE
Slide 10

6 DAYS LATER

- VA NEUROLOGY CLINIC
- BP 159/90
- LUMBAR PUNCTURE
  - OPENING PRESSURE 39 CM
  - > 25 IS HIGH
- LAB ANALYSIS OF CSF = NORMAL

- SO THE DIAGNOSIS IS...

---

Slide 11

DIAGNOSIS: IIH

- IDIOPATHIC INTRACRANIAL HYPERTENSION
  - PREVIOUSLY KNOWN AS PSEUDOTUMOR CEREBRI
- INCIDENCE
  - 1-2/100,000
  - HIGHER IN WOMEN AGE 15-44
  - 4-8X MORE LIKELY THAN MEN
  - MEDIAN AGE IS 30

---

Slide 12

IDIOPATHIC INTRACRANIAL HYPERTENSION (IIH)

- RISK FACTORS
  - WOMEN OF CHILDBEARING AGE
  - OVERWEIGHT
    - > 10% OVER IDEAL BODY WEIGHT
    - > 20% OVER IDEAL BODY WEIGHT
    - > 30% = 9X MORE LIKELY
  - MEDICATIONS
    - BCPS, TETRACYCLINES, HORMONE REPLACEMENT, ACCUTANE, VITAMIN A
    - HGH, ORAL STEROIDS, SYNTHROID, ISONIAZID, LITHIUM, NITROGLYCERIN
  - FAMILY HISTORY?
IDIOPATHIC INTRACRANIAL HYPERTENSION (IIH)

- **SYMPTOMS**
  - ASYMPTOMATIC
  - OR
  - HEADACHE 84-92%
  - LATERALIZED, THROBBING, PULSATILE, WORSE WITH POSTURE CHANGES
  - TRANSIENT VISUAL OBSCURATIONS 68-72%
  - INTRACRANIAL NOISES (PULSATILE TINNITUS) 52-60%
  - PHOTOPSIA 48-54%
  - BACK PAIN 53%
  - RETROBULBAR PAIN 44%
  - DIPLOPIA 18-38%
  - SUSTAINED VISUAL LOSS 26-32%

IDIOPATHIC INTRACRANIAL HYPERTENSION (IIH)

- **SIGNS**
  - SWOLLEN OPTIC NERVES
    - BILATERAL, ASYMMETRIC OR UNILATERAL
  - VISUAL LOSS
    - VA < 20/20 10-29%
  - VF LOSS
    - TYPICALLY PERIPHERAL, CENTRAL LATER
  - 6th NERVE PALSY
    - UNILATERAL OR BILATERAL
  - OTHER CN INVOLVEMENT
    - I, III, IV, V, VII, VIII

IDIOPATHIC INTRACRANIAL HYPERTENSION (IIH)

- **POSSIBLE RADIOLOGIC SIGNS**
  - ABSENCE OF ANOTHER CAUSE
  - EMPTY SELLA
  - ENLARGED OPTIC NERVE SHEATH
  - OPTIC NERVE TORTUOSITY
  - PROTRUSION OF THE OPTIC NERVE HEAD
  - CONCAVITY OR FLATTENING OF THE POSTERIOR GLOBES
  - OTHERS
    - NARROWING OF MECKEL'S (TRIGEMINAL) CAVE
    - REDUCTION IN DIAMETER OF CAVERNOUS SINUSES

IDIOPATHIC INTRACRANIAL HYPERTENSION (IIH)

- MECHANISM (WHAT CAUSES IT)
  - NOT REALLY KNOWN
- THEORIES
  1. INCREASED PRODUCTION OF CSF
  2. INCREASED BLOOD FLOW TO THE BRAIN OR INCREASE IN THE BRAIN TISSUE
  3. RESTRICTED VENOUS DRAINAGE FROM BRAIN

IIH TREATMENT

- MEDICAL
  - WEIGHT LOSS
  - CARBONIC ANHYDRASE INHIBITORS
    - ACETAZOLAMIDE
  - DIURETICS
    - FUROSEMIDE
  - CORTICOSTEROIDS
  - SERIAL LUMBAR PUNCTURES

IIH TREATMENT

- SURGERY PURSUED IF
  - SEVERE VISION LOSS
  - PROGRESSIVE VISION LOSS
  - INTRACTABLE HEADACHES
- OPTIONS
  - LUMBOPERITONEAL SHUNT
  - VENTRICULOLOPERITONEAL SHUNT
  - VENTRICULOATRIAL SHUNT
  - OPTIC NERVE SHEATH FENESTRATION
• OPTOMETRY 6 MOS LATER
  • PATIENT IS ON DIAMOX
  • 20/20 OD AND OS, VF UNCHANGED
  • ONH / OCT NFL APPEAR RELATIVELY UNCHANGED
• NEUROLOGY 3 MOS LATER
  • PATIENT HAS BEEN POORLY COMPLIANT WITH DIAMOX
  • FT DECLINES AFTER LP
  • OPTOMETRY 3 MOS LATER
  • 20/20 OD AND OS, VF UNCHANGED
  • ONH APPEARS SLIGHTLY BETTER
  • NEUROLOGY PENDING

• NEUROLOGY APPT
  • DUE TO NO / MINIMAL IMPROVEMENT AND NO ACTUAL IMAGING REPORT
  • REPEATED MRI / MRV
  • REPEAT MRI RESULTS
  • SMALL POSTERIOR FOSSA
  • OPTIC NERVE SHEATHS ARE TORTUOUS
  • CEREBELLAR TONSILS 12 MM BELOW FORAMEN MAGNUM

DIAGNOSIS:
CHIARI MALFORMATION

• EPIDEMIOLOGY
  • TRUE FREQUENCY UNKNOWN, MORE SINCE 1985 (MORE MRIS)
  • TYPE I PREVALENCE 0.1 TO 0.5%
• ETIOLOGY
  • USUALLY CONGENITAL
  • NOT NECESSARILY INHERITED
  • MUTATION OR DELETION OR EXOGENOUS TERATOGEN
  • EXACT CAUSE IS UNKNOWN BUT THERE ARE THEORIES
  • SMALL POSTERIOR FOSSA (CROWDING THEORY)
  • MOLECULAR GENETIC THEORY
  • HYDRODYNAMIC PULSION THEORY
  • OLIGO-CEREBROSPINAL FLUID THEORY
### CHIARI MALFORMATIONS

<table>
<thead>
<tr>
<th>Classification</th>
<th>Finding</th>
</tr>
</thead>
<tbody>
<tr>
<td>Type I (only type that can be acquired)</td>
<td>Cerebellar tonsils into foramen magnum</td>
</tr>
<tr>
<td>Type II (Arnold-Chiari)</td>
<td>Cerebellum and brainstem tissue into foramen, Vermis is partially complete or absent</td>
</tr>
<tr>
<td>Type III (most serious)</td>
<td>Occipital encephalocele</td>
</tr>
<tr>
<td>Type IV</td>
<td>Lack of cerebellar development</td>
</tr>
</tbody>
</table>

### HOW DIAGNOSED

- Opiostion-Basion Line
- Lower limit of posterior fossa
- Healthy adults
- Cerebellar tonsils rarely 3mm below foramen magnum
- Chiari Type I
  - Tonsils 5mm below

### NORMAL vs CHIARI vs CASE

- **NORMAL**
- **CHIARI**
- **CASE**
A DIFFERENT VIEW

CHIARI TYPE I

- SYMPTOMS
  - ASYMPTOMATIC
  - INCURABLE
  - HEADACHE, NECK PAIN
  - BALANCE PROBLEM
  - DIZZINESS
  - MUSCLE WEAKNESS
  - NUMBNESS
  - DIFFICULTY SWALLOWING
  - RINGING/DIZZINESS IN EARS
  - HEARING LOSS
  - MUSCLE WEAKNESS
  - NUMBNESS
  - DIFFICULTY SWALLOWING
  - RINGING/DIZZINESS IN EARS
  - HEARING LOSS

- OCULAR PROBLEMS
  - DRY EYES
  - PHOTOPHOBIA
  - BLURRED VISION
  - Nystagmus
  - PAIN BEHIND THE EYES
  - MAY WORSEN WITH PHYSICAL ACTIVITY
  - COUGHING
  - LAUGHING
  - SNEEZING

CHIARI

- COMPLICATIONS
  - HERNIATED TISSUES COMPRESS BRAINSTEM
  - BLOCKS FLOW OF NORMAL CSF
  - SYRINGOMYELIA
  - BUILDUP OF FLUID IN SPINAL CORD
  - SYRINGOMYELIA
  - BUILDUP OF FLUID IN SPINAL CORD
  - SYRINGOMYELIA
  - BUILDUP OF FLUID IN SPINAL CORD
  - VOMITING
  - DIFFICULTY SWALLOWING
  - RINGING/DIZZINESS IN EARS
  - HEARING LOSS

- HYDROCEPHALUS
  - BUILDUP OF FLUID IN THE BRAIN
CHIARI TYPE I

- MANAGEMENT
  - ASYMPTOMATIC
    - MONITORING WITH MRI
    - 6 MOS THEN YEARLY
    - MAY RESOLVE
  - SYMPTOMATIC / LARGE / PROGRESSIVE SYRINX
    - SURGERY
    - POSTERIOR FOSSA DECOMPRESSION
    - SYMPTOM IMPROVEMENT
    - 80-95% SYMPTOM IMPROVEMENT
    - 20% COMPLICATION RATE
    - SPINAL LAMECTOMY
    - SHUNTING PROCEDURES

A FEW QUESTIONS REMAIN...

IS IT POSSIBLE THE LP CAUSED THE CHIARI MALFORMATION?

- 2 CASES OF 30 YEAR OLD WOMEN WITH IIH
- 4 MM TONSILAR DESCENT ON MRI PRIOR TO LP
- RESULTS
  - 12 MM CEREBELLAR Herniation AFTER 1 LP
  - TONSILAR Herniation AFTER MULTIPLE LPS DUE TO PERSISTENT HEADACHES
Slide 31

HOW DO IIH AND CHIARI RELATE TO EACH OTHER?

• RELATIONSHIP IS POORLY UNDERSTOOD
• ELEVATED INTRACRANIAL PRESSURE IN IIH MAY CAUSE CEREBELLAR TONSILS TO herniate through FORAMEN MAGNUM
• OR
• PATIENTS WITH CHIARI TYPE I HAVE ABNORMAL CSF DYNAMICS WHICH MAY predispose patients to ELEVATED INTRACRANIAL PRESSURE AND IIH

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SO… WHICH CAME FIRST?

Slide 33

CASE

• 45 / W / F
• CC: loss of vision and floaters in left eye hours after visiting a theme park
• OCULAR HX:
  • LDE 1yr, lattice degeneration, no holes
• MED HX: unremarkable OU
• MEDS: none
• FAM HX: unremarkable OU
• SOCIAL HX: -etoh, -tobacco
• BVA:
  • cc 20/20 - 650DS
• cc 20/200, ph N I-675DS
• SLIT LAMP:
  • unremarkable OU
• IOP: 15/16 @ 815a
• DFE:
  • OD lattice degeneration, a few holes, no tears/ detachments
  • OS see photo
Differential Diagnosis

- Posterior Vitreous Detachment
- Retinal Hole
- Retinal Tear
- Retinal Detachment

Diagnosis: Retinal Detachment

- Result of separation of neurosensory retina from the RPE
- Ocular emergency
- Sight threatening
- Incidence 1 in 10,000
- Risk is highest in age 55-70 year olds
- Risk in second eye
  - 3.5-5.8% in year one
  - 9-10% in 4 yrs

Types of Retinal Detachment

- Rhegmatogenous
- Tractional
- Exudative
- Combined
- Tractional Rhegmatogenous
**Slide 37**

**RHEGMATOGENOUS RETINAL DETACHMENT**

- BREACH IN NEUROSENSORY RETINA
- SEEPAGE OF FLUID INTO SUBRETINAL SPACE

**Slide 38**

**RISK FACTORS**

- LATTICE DEGENERATION
- PERIPHERAL RETINAL BREAKS
- MYOPIA
  - UP TO 1D IS 4X RISK, 2D IS 6X RISK
- HISTORY OF OCULAR SURGERY
  - UNCOMPPLICATED CILIAR RISK IS 1/1000, 4 YEARS AFTER RISK IS 7X
- TRAUMA
  - 0.2/10000
- PREVIOUS RETINAL DETACHMENT
- FAMILY HISTORY
- OTHER
  - CONTACT SPORTS, HIGH RISK ACTIVITIES

**Slide 39**

**HIGH RISK ACTIVITY?**

SHAIKH S. RETINAL CASES AND BRIEF REPORTS. SPRING 2011. VOL 5, ISSUE 2, PP 128-129.
HIGH RISK ACTIVITIES

- WITHOUT MUCH EVIDENCE...
- THESE ALSO ARE CONSIDERED HIGH RISK
  - SKYDIVING
  - BUNGEE JUMPING
  - DIVING INTO WATER
- THEORIES
  - SUDDEN CHANGES IN PRESSURE
  - SUDDEN STARTING / STOPPING

AMUSEMENT PARK ACCIDENTS

- MOST COMMON
  - HEAD, NECK, BACK INJURIES
  - DEATH
  - STROKE
  - TRAUMATIC BRAIN INJURY
  - BRAIN ANEURYSMS
  - LACERATIONS, BROKEN BONES, TORN LIGAMENTS
  - DROWNING

AMUSEMENT PARK ACCIDENTS

- CAUSES
  - NEGLIGENCE BY GUEST
    - PASSENGER MISUSE
    - FAILURE TO FOLLOW INSTRUCTIONS
  - NEGLIGENCE BY PARK
    - MECHANICAL FAILURE OF RIDE
    - IMPROPER OPERATION OF RIDE
  - DIAGNOSED / UNDIAGNOSED PRE-EXISTING MEDICAL CONDITIONS
  - INHERENT NATURE OF RIDE
PREVIOUSLY REPORTED ROLLER COASTER SEQUELAE

- NEUROLOGIC
  - SUBDURAL HEMATOMA
  - SUBARACHNOID HEMORRHAGE
  - INTRAPARENCHYMAL HEMORRHAGE
  - CERVICOCEPHALIC ARTERIAL DISSECTION
  - CAROTID ARTERY THROMBOSIS
  - POSTTRAUMATIC MIGRAINE

- OCULAR
  - BLURRED VISION
  - LENS DISLOCATION
  - SUBLUXATION IN MARFAN’S LEAD TO GLAUCOMA
  - INTRAOCULAR LENS DISLOCATION
  - REPOSITIONING AFTER ROLLER COASTER
  - RETINAL / MACULAR HEMORRHAGE
  - RETINAL ARTERY OCCLUSION

MACULAR HEMORRHAGE

- 26 YO FEMALE
- SMALL AREA OF VISUAL DISTORTION OD X 2 DAYS
- NO MEDICAL HISTORY, BCP ONLY
- RODE ROLLER COASTER 13X THREE DAYS BEFORE
- THEORETICAL CAUSE
  - HIGH HYDROSTATIC PRESSURE FROM G FORCES
  - Sudden Increased Cerebral Pressure
  - Leads to Increased Retinal Venous Pressure

**Slide 46**

**Retinal Artery Occlusion**
- 35 yo female
- Right neck pain, frontal headache after riding roller coaster
- No prior medical history
- Visual field defect OD of vision for 2 days
- At hospital: 20/200 OD, 20/20 OS

**Theoretical Cause**
- Rapid acceleration and deceleration leads to whiplash-like head movements
- This may cause excessive force on the vitreous that causes shear stress exceeding the strength of retinal adhesions
- Additionally, that may also damage the neural parenchyma and vasculature

[Representative photo](http://webeye.ophth.uiowa.edu/eyeforum/atlas/pages/BRAO-OD.html)

**Case From:**

**Representative Photo**

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**Slide 47**

**Roller Coaster RD**

**Mechanism Theories**
- Rapid acceleration and deceleration leads to whiplash-like head movements
- This may cause excessive force on the vitreous that causes shear stress exceeding the strength of retinal adhesions
- Additionally, that may also damage the neural parenchyma and vasculature

[Representative photo](http://webeye.ophth.uiowa.edu/eyeforum/atlas/pages/BRAO-OD.html)

**Slide 48**

**RD Treatment Options**

- **1950s Scleral Buckle**
  - Sponge or encircling band
  - Reduces traction
  - RPE absorbs the SRF

- **1970s Vitrectomy**

- **1986 Pneumatic Retinopexy**
  - Air
  - Argon or diode laser
  - Special silicon oil (SF6, C2F6, C3F8)

- **Silicone Oil**
  - For PVR, may stay in long-term

- **Combination**
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**HOW TO TREAT RHEGMATOGENOUS RD?**

- **2013 METAANALYSIS**
- **7 REPORTS REVIEWED**
- **836 EYES PPV, 670 EYES SB**

**RESULTS**

- **PHAKIC**
  - **NO SIGNIFICANT DIFFERENCES IN REATTACHMENT**
  - **SB EYES HAD BETTER VISION (LESS CATARACTS)**

- **PSEUDOPHAKIC**
  - **NO DIFFERENCES IN REATTACHMENT VS BCVA AT 6 MONTHS**

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**Slide 50**

**RD TREATMENT INDICATIONS / CONTRAINDICATIONS**

- **Ahmad BU, Shah G, Binder K. Review of Ophthalmology. 4/05/13**

**COMPLICATIONS OF TREATMENT**

- **SCLERAL BUCKLE**
  - **CHANGE IN REFRACTION**
  - **DOUBLE VISION**
  - **INFECTION**
  - **MOTION PROBLEMS**
  - **VITRECTOMY**
  - **77% CATARACT FORMATION WITHIN 1YR**
  - **HOLE CREATION IN 17%**
  - **RARE VITREOUS HEME, ENDOPHTHALMITIS**
SUPPLEMENTAL TREATMENT

- LASER AND / OR CRYO
- DONE FOR HOLES AND TEARS IN THE RD EYE
- DONE IF SYMPTOMATIC IN THE OTHER EYE
- WHAT ABOUT THE OTHER EYE...
  - LATTICE (FROM THE AAO)
  - NO CONSENSUS GUIDELINES
  - RD IN OTHER EYE 5%
  - UP TO 25% IN HIGH MYOPES WITH EXTENSIVE LATTICE
  - LASER REDUCES RISK 3X

FOLLOW-UP

CHANGING TIMES

- IMPROVE SAFETY
- RIDER EDUCATION
- BETTER RESTRAINTS
- SHORTENING THE RIDE
- REDUCES TIME AN INJURED PERSON SUFFERS
- ORLANDO
  - SEAWORLD'S MAKO
    - 750 FT LONG, 135 MPH, 3.5 SEC
    - SKYSCRAPER
      - COMING TO ORLANDO 2017
      - HEIGHT / DISTANCE / SPEED?
WHAT SHOULD OPTOMETRISTS DO?

- Educate at risk patients

- Advise potentially predisposed patients about their ocular risks of high risk activities

- Have eyes checked immediately if sudden flashes of light

- Floating spots

- Appearance of curtain in front of the vision

- Loss of peripheral vision

OTHER AREA “ATTRATIONS”

- Options

- Orlando Eye, Gatorland, Holy Land, Fun Spot, Old Town

- Dining / Entertainment Areas

- Disney Springs, City Walk, Downtown, College Park, Winter Park, Lake Mary, etc.

- Water Parks

- Typhoon Lagoon, Blizzard Beach, AQUATICA, Wet N Wild (until 12/16)

- Shows

- Cirque du Soleil, Pirate’s Dinner Adventure, Hoop Dee Doo Revue, Blue Man Group

CASE

- 89 / W / M

- CC: distance blur x 1yr, no near problems, no comfort problems

- Oc Hx:

  - LEE 2yrs: trauma OS, h/o strab surgery, pseudophakic with scleral OS

- Oc Meds: none

- Med Hx:

  - HTN, Heart (A fib)

- Soc Hx:

  - Alcohol, Tobacco

- BVA: cc 20/25 - 2, cc 20/40+1

- Prelim: FROM, FTFC OD OS, surgical pupil OS, no APD

- S.E. see photos

- D.E.: see photos
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WHAT DO I MEAN?

HE APPEARS BLUE.

NO, NOT SAD.

HE’S BLUE.

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DIFFERENTIAL DIAGNOSIS

- CYANOSIS
  - Lack of oxygen
  - Central or peripheral
- MEDICATIONS
  - Antimalarials, minocycline, amiodarone, chlorpromazine
- HEAVY METALS
  - Gold, mercury, bismuth
- MELANOSIS
  - From metastatic melanoma
- HEMOCHROMATOSIS
- POLYCYTHEMIA VERA
- ADDISON’S DISEASE
- BLUE NEVUS
- OCHRONOSIS
- ARGYRIA

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DIAGNOSIS: ARGYRIA

- SIGNS
  - Hyperpigmentation in sun-exposed areas of skin
  - Forehead, nose, hands
  - Entire skin may be slate blue-gray color
  - Sclera, nail beds, mucous membranes may be hyperpigmented
  - Viscera a high blue discoloration
  - Spleen, liver, gut
ARGYRIA

- CAUSE
  - PROLONGED CONTACT WITH OR INGESTION OF SILVER

HOW DOES IT HAPPEN

- COLORLESS SILVER
  - REDUCED BY SUNLIGHT TO BROWNISH-BLACK SILVER SULPHIDES AND SELENIDE WITHIN TISSUE
- SILVER
  - STIMULATES TYROSINASES ACTIVITY OF MELANOCYTES TO PRODUCE MORE MELANIN
- THEREFORE
  - MOST PROMINENT IN SUN-EXPOSED AREAS

RISK FACTORS

- WHO IS AT RISK?
  - INDIVIDUAL VARIABILITY
  - DUE TO LENGTH OF EXPOSURE AND TOTAL DOSE NEEDED
- INCREASED RISK
  - WORKERS
    - SILVER MINERS / REFINERS
    - SILVERWARE / GLASS / CHINA MANUFACTURING
    - PHOTOGRAPHIC PROCESSING
  - NOT AS MUCH ANYMORE
RISK FACTORS

- INCREASED RISK
- SILVER SALT IRRIGATION
- URETHRAL / NASAL MUCOUS MEMBRANES
- EYE DROPS
- WOUND DRESSING
- SMOKING REMEDY
- SURGICAL AND DENTAL PROCEDURES
- ACUPUNCTURE
- EARRING SITES
- DIETARY SUPPLEMENTS
- COLLOIDAL SILVER

WHAT IS COLLOIDAL SILVER?

- ONCE AVAILABLE IN OTC DRUG PRODUCTS
- 1999 FDA RULED NOT SAFE AND EFFECTIVE
- STILL BEING SOLD
- AS HOMEOPATHIC REMEDIES AND DIETARY SUPPLEMENTS
- MAKE YOUR OWN

http://www.webmd.com/vitamins-supplements/
SERIOUS COMPLICATIONS

- GI PROBLEMS
- TISSUE WASTING
- UREMIA
- ALBUMINURIA
- FATTY DEGENERATION
- LIVER, KIDNEY, HEART
- THROMBOCYTOPENIA
- FLUIDITY OF THE BLOOD
- CHRONIC BRONCHITIS
- LOSS OF COORDINATION
- DECREASED NIGHT VISION
- TASTE DISTURBANCE
- VESTIBULAR IMPAIRMENT
- GRAND MAL SEIZURES
- PARALYSIS OF RESPIRATORY SYSTEM
- DEATH

OCULAR COMPLICATIONS

- CONJUNCTIVAL ARGYRIA
- BLACK TEARS RARELY OCCUR
- SCLERAL HYPERPIGMENTATION
- DECREASED NIGHT VISION

TREATMENT OPTIONS

- DEPIGMENTATION PREPARATIONS
  - SOME REPORT 5% HYDROQUININE MAY WORK
- LASER
  - TO REMOVE PIGMENTED SKIN/CELLS
- CHELATION
  - UNSUCCESSFUL
- SUNSCREEN
  - MAY PREVENT WORSENING
- PROGNOSIS
  - PERMANENT, IRREVERSIBLE METALLIC TINGE
...BACK TO THE PATIENT

- HOW DID HE TURN BLUE?
- DURING WWII, PENICILLIN WAS IN SHORT SUPPLY
- HE WAS TREATED WITH SILVER WOUND DRESSING
- THAT WAS 70 YEARS AGO
- UPON QUESTIONING...
- WHY?
- HE TREATED HER TOO
- "THANKS DEAR"

- FYI...HIS WIFE WAS BLUE TOO

JF

- 65/W/M
- CC: no changes in vision, blue top drop stings, dry mouth from purple dyes
- Oc Hx: LEE 4 mos, dx/tx Glaucoma x 13yrs, highest IOP off meds 26/27, h/o Lasik OU, dry eyes OU, DM without Ret OU, Early Cataracts OU
- Oc Meds: Latanoprost qhs OU, Cosopt q12h OU, Brimonidine 0.2 q12h OU
- Med Hx: +DM, +HTN, +Chol
- Fam Hx: unremarkable
- Soc Hx: -etoh, -tobacco
- BVA: cc 20/25 -2, cc 20/25 -1+2
- Prelim: FROM, FTFC OD OS, No APD
- SLE: see photos
- IOP: 14/14@ 625a, pachym: 547/548
- Gonio: OU open to CBB 360, no PAS/recess/nv, trace pigment
- DFE: see photos

YOUR PATIENT HAS GLAUCOMA.
• ONCE THE DIAGNOSIS OF GLAUCOMA HAS BEEN MADE, THE MOST IMPORTANT REMAINING QUESTION IS…

• WHETHER THE DISEASE IS STABLE AND THE THERAPY / COMPLIANCE ARE SUFFICIENT OR

• WHETHER THE DISEASE IS PROGRESSIVE AND THE THERAPY IN RELATION TO LIFE EXPECTANCY HAS TO BE INTENSIFIED

• CURRENTLY, NO SPECIFIC TEST CAN BE REGARDED AS THE PERFECT STANDARD FOR DETERMINATION OF PROGRESSION

• BOTH ONH STRUCTURE AND FUNCTION SHOULD BE EVALUATED FOR DETECTION OF PROGRESSION

• YOUR PATIENT IS GETTING WORSE.
Slide 76

**QUESTION**

WHAT ARE YOU GOING TO DO NOW?

Slide 77

**VISIT THE OUTLETS**

- IOP LOWERING OPTIONS
  - MECHANISM
  - RESEARCH OUTLOOK
  - METHODS
  - LAMBS
  - ALT / SLT
  - EMBERS
  - EXTRACT SYSTEM
  - VERS
  - TRABECULAR IRRIGATION
  - CYLINDRICAL
  - TRACTION ACTUATION
  - SPIKE METHOD
  - THEIR IRRIGATION
  - PHA
  - CYCLOPLASTY
  - CANALOPLASTY
  - VISCOCANALOSTOMY
  - ETC.

1. Lake Buena Vista
2. Premium Outlets International Dr
3. Premium Outlets Vineland Ave

From Forbes.com on 1/20/12

Slide 78

**LASER TRABECULOPLASTY**

- ALT OR SLT
  - MECHANISM IS NOT CLEAR
    - MECHANICAL
    - CELLULAR
    - BIOCHEMICAL
  - PROS
    - ALT AS GOOD AS
    - TIMOLOL
  - CONS
    - IOP SPIKE, A/C REACTION
    - SLT NEEDS DEEP ANGLE,
      PIGMENTED TM
    - NOT EFFECTIVE ON
      EVERYONE
    - MAY WEAR OFF
SLT IN CLINICAL TRIALS

- SLT VS ALT
  - SIMILAR EFFECT
  - EFFICACY
    - 20-30% IOP REDUCTION
  - UNCONTROLLED OAG
    - MAX MEDS, PRIOR FAILED ALT
    - 70% DECREASED 3 months
    - 24% SPIKED > 5 mmHg

- SLT MED STUDY
  - Katz LJ, et al. 2011
  - SLT (58 EYES)
    - 100 APPLICATIONS, 360 DEGREES (REPEATED IF ABOVE TARGET)
    - 26.4% IOP REDUCTION
  - MEDICATION (36 EYES)
    - MED CHANGED IF ABOVE TARGET
    - 27% IOP REDUCTION
    - ADDITIONAL TOP TREATMENT NEEDED IN MED GROUP

CATARACT SURGERY

- 32 STUDIES REVIEWED
  - POAG, PXG, PACG
  - POAG / NTG (ON 1-2 MEDS)
    - REDUCES IOP 13%, MEDS BY 12%
  - COMPLICATIONS
    - 17.5% CHANCE OF IOP SPIKE
  - OHTS RESULTS
    - 17% IOP REDUCTION 3YRS AFTER
  - OTHER STUDIES
    - 2-4 mmHg IOP REDUCTION

CATARACT SURGERY

- BALTIMORE VAMC 2006-2008
  - RETROSPECTIVE STUDY, 115 PTS
  - RESULTS
    - HIGHER PREOPERATIVE IOP ASSOCIATED WITH GREATER IOP LOWERING
    - PHACO TIME ASSOCIATED WITH IOP REDUCTION
  - MECHANISM
    - NOT WELL UNDERSTOOD
    - THEORIES
      - HYPOSECRETION OF AQUEOUS DUE TO FREE RADICALS
      - IMPROVED TM OUTFLOW DUE TO DEEPENING ANTERIOR CHAMBER
      - STRESS REMODELING FROM THE ULTRASONIC VIBRATIONS ACTIVATES CYTOKINE PATHWAY

Int Ophthal. 11 January 2016
MIGS
MINIMALLY INVASIVE GLAUCOMA SURGERY

- FROM THE AMERICAN GLAUCOMA SOCIETY
- REDUCE IOP BY IMPROVING OUTFLOW
- APPROACHED FROM INSIDE (AB INTERNO) OR OUTSIDE (AB EXTERNO)
- LIMITED SURGICAL MANIPULATION OF THE SCLERA
- LITTLE MANIPULATION OF THE CONJUNCTIVA
- DOES NOT PRECLUDE POSSIBILITY OF TRADITIONAL SURGERY

- BENEFITS
  - HIGHER SAFETY PROFILE VS TRABECULECTOMY
  - SHORTER SURGERY TIME
  - FEWER COMPLICATIONS
  - FASTER RECOVERY TIME
  - DECREASE IOP AND/OR PATIENT'S NEED FOR MEDICATION

WHO IS A CANDIDATE?
- MILD-MODERATE OPEN-ANGLE GLAUCOMA
- PSEUDOEXFOLIATION GLAUCOMA
- PIGMENTARY GLAUCOMA
- UNCONTROLLED GLAUCOMA ON MAXIMUM TREATMENT OR BARRIERS TO PREVENT ADEQUATE DOSE
  - SIDE EFFECTS
  - CATARACT?
- >18 CATARACT?

CONTRAINDICATIONS
- ANGLE CLOSURE
- MODERATE-ADVANCED SECONDARY GLAUCOMA
- SEVERELY UNCONTROLLED GLAUCOMA
- PREVIOUS GLAUCOMA SURGERY
- PREVIOUS REFRACTIVE PROCEDURES
- MONOCULAR PATIENTS

MIGS CURRENTLY APPROVED
- TRABECTOME
- ISTENT
- CYPASS MICROSTENT
**Slide 85: TRABECTOME**
- FDA APPROVED 2004
- MECHANISM: REDUCES RESISTANCE TO OUTFLOW
- PROCEDURE: MICROLELECTROCAUTERY ABLATION OF PART OF TM AND INNER WALL OF SCHLEMM'S CANAL
- PROS
  - CAN BE PERFORMED AT SAME TIME AS CATARACT SURGERY
- CONS
  - IOP DOES NOT REACH SINGLE DIGITS (LIMITED BY EVP)
  - Healed IOP spikes in 5.8%
- STUDY RESULTS
  - LIMITED STUDY DATA
  - FDG REPORT: 1127 CASES
  - DECREASED IOP 32% AT 5 YEARS
  - MEDS REDUCED BY 39%
- **Slide 86: iSTENT**
- FDA APPROVED 2012
- MECHANISM: BYPASSES THE TRABECULAR MESHWORK
- PROCEDURE:
  - 120 um DIAMETER TITANIUM IMPLANT
  - PRELOADED APPLICATOR
  - INSERTED INTO SCHLEMM'S CANAL TO ALLOW AQUEOUS TO DRAIN DIRECTLY FROM AC
  - 2ND/3RD GENERATIONS IN TRIALS
- PROS
  - NO BLEB THEREFORE PRESERVES CONJ For TRAB
  - NO IRIDECTOMY NEEDED
- CONS
  - IOP DOES NOT REACH SINGLE DIGITS (LIMITED BY EVP)
  - SURGICAL COMPLICATIONS
    - TOUCHING THE IRIS
    - FAILURE TO IMPLANT
    - ENDOTHELIUM CONTACT
    - MALPOSITION
  - $1000 PER PIECE
- STUDY RESULTS
  - FDA REPORT: 116 ISTENT WITH CATARACT SURGERY VS 123 CATARACT SURGERY ALONE
  - RESULTS
    - 68% COMBINED IOP <21
    - 50% CATARACT ONLY
- **Slide 87: iSTENT WITH CATARACT SURGERY**
- PROS
  - NO BLEB THEREFORE PRESERVES CONJ For TRAB
  - NO IRIDECTOMY NEEDED
  - NO SURGICAL COMPLICATIONS
    - TOUCHING THE IRIS
    - MALPOSITION
  - LESS EXPENSIVE
- STUDY RESULTS
  - FDA REPORT: 116 ISTENT WITH CATARACT SURGERY VS 123 CATARACT SURGERY ALONE
  - RESULTS
    - 68% COMBINED IOP <21
    - 50% CATARACT ONLY
ISTENT ALONE

- 2015 ARMENIAN STUDY
- 119 PATIENTS FOLLOWED 18 MONTHS
- 38 (1 STENT) VS 41 (2 STENTS) VS 40 (3 STENTS)
- IOP 15.9 vs 14.1 vs 12.2
- UNMEDICATED 64.9% VS 85.4% VS 92.1%
- HEALTH INSURANCE GENERALLY ONLY REIMBURSES FOR ONE STENT
- FDA HAS APPROVED STENT IN CONJUNCTIONS WITH CATARACT SURGERY BUT NOT AS STAND ALONE PROCEDURE

CYPASS MICRO-STENT

- FDA APPROVED AUGUST 2016
- FROM TRANSCEND MEDICAL
- 6.35 mm LONG TUBE, 510 um
- MECHANISM
- INSERTED THROUGH THE ANGLE BETWEEN SCLERAL SPUR AND THE CILIARY BODY INTO THE SUPRACILIARY SPACE
- NEGATIVE PRESSURE GRADIENT BETWEEN SUPRACHOROIDAL SPACE AND ANTERIOR CHAMBER
- AQUEOUS MOVES TO SUPRACHOROIDAL SPACE

COMPASS STUDY

- 505 PATIENTS
- EITHER CATARACT OR CATARACT AND CYPASS STENT
- AT 24 MONTHS
  - 73% ACHIEVED IOP DECREASE OF 20%
  - 61% IN THE TARGET RANGE (6-18 mHg)
- DONE WITH CATARACT SURGERY
Slide 91

FUTURE MIGS?

- XEN GEL STENT
  (AQUESYS)

- HYDRUS IV
  (IVANTIS)

- MICROSHUNT
  (INNFOCUS)

Slide 92

COMMENTS ON MIGS

- WHAT'S NOT KNOWN?
- LONGTERM SAFETY OR EFFECTIVENESS DATA

http://www.aao.org/eyenet/article/two-approaches-to-migs-istent--trabectome

Slide 93

Follow-up (years)

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<td>Percent of Visits with IOP Less Than 18 mm Hg</td>
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<td>75-99% of visits</td>
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<td>50-74% of visits</td>
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<td>0-49% of visits</td>
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AGIS 7
Sustained IOP below 18 mm Hg:
Positive Correlation with Stability of Visual Field

12.3
14.7
16.9
20.2
GLAUCOMA SURGERY

- TRABECULECTOMY
  - First described in 1968
  - Mechanism
    - Creates a drainage channel (a hole in the eye) from the anterior chamber to external surface of the eye under the conjunctiva
    - Aqueous seeps into bleb which is slowly absorbed

GLAUCOMA SURGERY

- Tube / Shunt (Glaucoma Drainage Device)
  - Mechanism
    - Placement of tube / valve to facilitate aqueous outflow
  - Types
    - Little resistance to aqueous (Baerveldt, Molteno)
    - Unidirectional valve (Ahmed, Krupin)

TUBE vs TRAB (TVT) STUDY

- Prospective study (17 centers, 212 eyes of 212 patients)
  - 107 in Tube Group, 105 in Trab / MMC Group
  - Patients
    - Uncontrolled glaucoma, s/p CE / IOL and / or failed Trab
  - 5 Year Results
    - 15.3 ± 3.9 mmHg vs 12.8 ± 5.8 mmHg
    - Probability of failure: 26% at 5 years in Tube vs 45% in Trab (p = 0.002)
    - Late complications: 34% in Tube vs 37% in Trab (p = 0.67)
    - Endophthalmitis / blebitis: 0% in Tube vs 4.8% in Trab
  - Conclusions
    - Tube shunts are a good alternative in those who have had prior surgery
    - Total costs of tube were higher than trab
    - Was not a study on “fresh” patients
PATIENTS IN TROUBLE
• RECOGNIZE PATIENT
  • PATIENT PRESENTING WITH SEVERE DAMAGE
  • BASED ON OPTIC NERVE, NERVE FIBER LAYER, VISUAL FIELD
  • PATIENT WHO IS YOUNG
  • PATIENT WHO IS AFRICAN AMERICAN
  • PATIENT WHO IS AN IOP SPIKER
  • PATIENT WITH A THINNER CORNEA
  • LOW CORNEAL HYSTERESIS
  • PATIENT WHO SHOWED PROGRESSION OF DISEASE EVEN THOUGH TREATED

BACK TO THE PATIENT...
• HE HAS HAD SLT OS
• WAITING TO DETERMINE HOW MUCH IOP RESPONSE HE HAD
• NEED TO BE VERY CAREFUL
• OS IS ALREADY IN BAD SHAPE
• OD IS AT GREAT RISK
  • NEED CLOSE MONITORING
  • QUICKER ADVANCEMENT OF TREATMENT

CASE
• 55 / B / M
• CC: no vision complaints, would like new NVOS, some dryness
• OCULAR HX:
  • LEE 1 yr, trauma to left eye while fishing
• MED HX: +DM, +Chol
• FAM HX: unremarkable OU
• SOCIAL HX: -etoh -tobacco
• BVA:
  • sc 20/20-1, sc h/2 at 2 feet
• PRELIMS:
  • TPR 4+ (50 ml), +h/2, +m/2
  • ROUND IRREGULAR
• NO APD BY REVERSE
• SLIT LAMP:
  • OD unremarkable, OS hazy photo
• IOP: 20/18 @ 9:45a
• DFSE:
  • OD unremarkable, OS no view
OCULAR TRAUMA

- **UNITED STATES**
- 2.5 MILLION OCULAR INJURIES PER YEAR
- 40,000-60,000 CASES OF BLINDNESS PER YEAR
- 25% RELATED TO SPORTS / RECREATION
- ~100,000 SPORTS RELATED EYE INJURIES / YR WARRANT VISIT TO ER OR DOCTOR’S OFFICE

ACUTE COMPLICATIONS OF SPORTS RELATED OCULAR INJURIES

- LID EDEMA / CONTUSION / LACERATION
- CONJUNCTIVAL HEMORRHAGE / LACERATION
- CORNEAL ABRASION / LACERATION / PENETRATION
- HYPhEMA, IRITIS
- IRIS Prolapse
- VITREOUS HEMORRHAGE
- COMMOTIO RETINAE
- RETINAL HEMORRHAGE
- RETINAL BREAK / DETACHMENT
- CHOROIDAL HEMORRHAGE
- ORBITAL FRACTURE

LATE COMPLICATIONS OF SPORTS RELATED OCULAR INJURIES

- TRAUMATIC MYDRIASIS
- CATARACT
  - TRAUMATIC
  - SUBSTAINED LENS
- SECONDARY GLAUCOMA
- CHOROIDAL RUPTURE
  - CNVM
- HYPOTONY
- PHTHIS
- DIPLOPIA
FISHING-RELATED OCULAR TRAUMA STUDY

- CAUSES
  - FIshhooks, weights, lures, rods

- DEMOGRAPHICS
  - 6-68 years old, 39% < 19 yo
  - 79% male

- OTHER DATA
  - 12.48% bystanders impacted
  - 2% involved alcohol

- PROGNOSIS
  - 21% < 20/200
  - 57% closed globe > 20/40
  - 11% open globe > 20/40
FISHING-RELATED TRAUMA

FISHING-RELATED OCULAR TRAUMA

CORNEAL PENETRATION ONLY

Ophthalmic Surg Lasers Imaging 2008;39:137-139

Pre and Post-OP
QUESTION
WHAT HAPPENED IN THE CASE PRESENTED?

PRESUMED CORNEAL PENETRATION / LACERATION AND ANTERIOR CAPSULAR DISRUPTION

LENS TRAUMA
• POSSIBILITIES
  • BLUNT TRAUMA
  • PENETRATING TRAUMA
  • CATARACT FORMATION
  • TRAUMA TO LENS CAPSULE OR INJURY TO LENS PUMP
  • INCREASED PERMEABILITY
  • INFLUX OF SODIUM AND WATER INTO AQUEOUS
  • INTRACELLULAR / EXTRACELLULAR SWELLING
  • LENS PROTEOLYSIS, AGGREGATION, CONFORMATIONAL CHANGES
• LENS OPACIFICATION

TRAUMATIC LENS COMPLICATIONS

QUESTION
WOULD THE PATIENT BENEFIT FROM CATARACT SURGERY?

REVIEW OF THE CHART...

• OPHTHALMOLOGY
  • SAID SURGERY WAS AN OPTION
  • PATIENT AGREED
  • DAY OF SURGERY
  • SURGERY STARTED
  • THEN STOPPED
  • WHY?
  • ZONULAR SUPPORT OF THE LENS WAS DEEMED TO BE POOR

THINGS THEY CONSIDERED

- Trauma Considerations
  - Zonular Dialysis
  - Posterior Capsular Tear
  - Iris Trauma
  - Corneal Trauma
  - Poor visualization from media haze

SURGICAL CONSIDERATIONS

- Synechiae
- Non-Dilating Pupil
- Intraoperative Dialysis
- Nucleus Drop
- Vitreous Loss
- Hypoesthesia

AGARWAL A, ET AL. CURRENT OPTION IN OPHTHALMOLOGY 2010, 21: 65-70

QUESTION
WHAT WERE THEY MOST WORRIED ABOUT?

ZONULAR DEHISCENCE

- What Causes It
  - Inherited or Acquired
  - High Myopia
  - Pseudoxfoliation, Retinitis Pigmentosa, Marfan Syndrome, Homocystinuria
  - S/P Multiple Vitrectomies, Silicone Oil
  - Blunt and Penetrating Trauma
  - Idiopathic
Slide 118

**OPHTHALMOLOGY**

- RETINA
- LENSECTOMY
- PPV
- ENDO LASER
- WHY?

---

Slide 119

**SHOULDA / COULDA / WOULDA…**

Recommendation is for cataract surgery to be done at time of laceration.

---

Slide 120

**WHAT SHOULD OPTOMETRISTS DO?**

- EMPHASIZE PREVENTION
- KNOW YOUR PATIENTS
- DEMOGRAPHIC
- HOBBIES
- RECOMMEND
- PROTECTIVE LENSES
  - POLYCARB OR TRIVEX
  - REDUCES INJURY RISK BY 90%
- OTHER: UV, POLARIZED
- IF FISHING RELATED INJURY…
  - BARE FIST AID
  - DON’T REMOVE FISH HOOK
  - GET PATIENT TO SPECIALIST

SEPTEMBER IS SPORTS EYE SAFETY MONTH

preventblindness.org
BACK TO THE PATIENT...

• NO NEW PROBLEMS
• WEAR RX FULL-TIME FOR PROTECTION
• PATIENT EXPRESSES UNDERSTANDING AND AGREES

CASE

• 45 / W / F
• CC: floaters os, no ocular comfort problems, no vision problems
• OCULAR HX: LEE few years ago, no surgery, no trauma/cataract/glaucoma
• MED-HX: unremarkable
• MEDS: none
• FAM HX: unremarkable
• SOCIAL HX: unremarkable
• BVA: sc 20/20, sc 20/20
• PRELIMS: ETDC 00-00, FROM
• NORMAL PUPILS, NO APD
• SLIT LAMP: unremarkable
• IOP: 12/13 @ 719a
• DFE: see photos

DIFFERENTIAL DIAGNOSIS

• CHOROIDAL NEVUS
• RPE HYPERPLASIA
• RPE HYPERTROPHY (CONGENITAL OR NOT)
• MELANOCYTOMA OF CHOROID
• METASTATIC CARCINOMA
• MALIGNANT MELANOMA
• HAMARTOMA
• ADENOMA
• ADENOCARCINOMA
DIAGNOSIS: RPE HYPERTROPHY

- APPEARANCE
  - FLAT, ROUND OR OVAL
  - WELL DERMACATED OR SCALLOPED MARGINS
  - UNILATERAL > BILATERAL
  - 88% PIGMENTED
  - ½ BROWN / 1/3 BLACK / GRAY / RARELY NO PIGMENT
  - ½ HAVE LACUNAE (ATROPHIED WINDOW DEFECTS)
  - PIGMENTED OR NONPIGMENTED HALO
  - LACK OF LIPOFUSCIN OR DRUSEN
  - PROGRESSIVE LOSS OF OVERLYING PHOTORECEPTORS (SEEN ON OCT)

RPE HYPERTROPHY

- PATHOLOGY
  - FOCAL AREA OF TALLER RPE CELLS
  - MORE DENSELY PACKED WITH MELANOSOMES
  - MELANOSOMES ARE LARGER AND SPHERICAL COMPARED TO NORMAL RPE (SMALLER, ELLIPTICAL)
  - INTENSELY HYPERTROPHIED RPE CELLS

- PROGRESSION
  - 75-80% SLOWLY GROW IN DIAMETER
  - MAY DEVELOP AN ELEVATED NODULE
  - REPRESENTS ADENOMA OR ADENOCARCINOMA

RPE HYPERTROPHY

- BACKGROUND
  - PREVELANCE 1.2-4.4% IN NORMALS
  - FEMALES > MALES 2:1
  - 2-3 SUBTYPES
    - SOLITARY
    - GROUPED (BEAR TRACKS)
    - MULTIPLE

http://imagebank.asrs.org/file/1791/congenital-hypertrophy-of-rpe
http://fighteyecancer.com/the-eye/intraocular/retinal-pigment-epithelium-rpe/
BEAR TRACKS

- A RPE HYPERTROPHY VARIATION
- APPEARANCE
- CLUSTER OR GROUPS OF LESIONS
- SMALL, SHARPLY CIRCUMSCRIBED
- POSSIBLY MORE ELLIPTICAL

BEAR TRACKS EXAMPLE

FROM: RETINA QUIZ. REVIEW OF OPTOMETRY 3/15/12
BY MARK T. DUNBAR, OD

QUESTION

IS THERE ANYTHING TO WORRY ABOUT?
**FAMILIAL ADENOMATOUS POLYPOSIS (FAP)**

- **CHARACTERISTICS**
  - ADENOMATOUS POLYPOSIS OF LARGE AND SMALL INTESTINE
  - HUNDREDS TO THOUSANDS OF POLYPS
  - HAMARTOMAS OF SKELETON
  - HAMARTOMAS OF SKELETAL TUMORS
  - AUTOSOMAL DOMINANT
  - INCIDENCE 0.4-1.3 /10000
  - PREVALENCE 1 / 35000

- **POTENTIAL SIGN**
  - MULTIPLE RPE HYPERTROPHY LESIONS IN BOTH EYES
  - DIFFERENT THAN RPE HYPERTROPHY?
  - SOME THINK SO
  - PIGMENTED OCULAR FUNDUS LESIONS OF FAP (POFLs)
  - DEVELOPMENT OF NEW BILATERAL LESIONS
  - STRONGLY ASSOCIATED WITH FAP
  - SENSITIVITY 65-84%, SPECIFICITY >94%
  - ANOTHER REASON TO HAVE EYES DILATED

- **VARIANTS**
  - ATTENUATED FAP
    - MILDER FORM, LATER AGE / CANCER
  - GARDNER SYNDROME
    - RPE HYPERTROPHY LIKE LESIONS IN 66-75%
  - COLONIC ADENOMATOUS POLYPOSIS
    - BONE CYSTS, HAMARTOMAS
  - SOFT TISSUE TUMORS (DESMOID TUMORS)
  - RARE
    - TURCOT SYNDROME
      - MEDULLOBLASTOMAS AND/OR ASTROCYTOMAS
**FAMILIAL ADENOMATOUS POLYPOSIS (FAP)**

- Fatality rate is 100% if untreated by age 50
- 1% of all colorectal cancers
- 5% of familial colorectal cancer
- Prevalence: 1 in 11,300 to 1 in 37,600
- Manifests by late teens to twenties
- Male = Female

Familial adenomatous polyposis is a genetic disorder characterized by the development of hundreds to thousands of adenomatous polyps in the colon and rectum. These polyps have a high risk of developing into colorectal cancer. The disease is autosomal dominant, meaning that a single copy of the mutated gene, inherited from either parent, is sufficient to cause the disorder. If left untreated, the disease is usually fatal by age 50 due to the significant risk of colorectal cancer.

**Slide 134**

**FAMILIAL ADENOMATOUS POLYPOSIS (FAP)**

- No symptoms until polyps are large
- 0-10 years from time of discovery
- Symptoms:
  - Rectal bleeding
  - Iron deficiency anemia
  - GI complaints
    - Change in bowel habits
    - Constipation
    - Flatulence
    - Abdominal pain
  - If symptomatic, 2/3 already have carcinoma of colon or rectum

**Slide 135**

**MULTIPLE CHRPE AND / OR FAMILIAL ADENOMATOUS POLYPOSIS (FAP)**

- How to identify 80% of gene carriers
  - If 4 or more small lesions
  - Bilateral, multiple lesions
- Management:
  - Probe patient history and/or family history
  - About polyps, colorectal cancer, colon surgery
  - Refer to gastroenterologist
  - Endoscopy/colonoscopy
  - Genetic testing
  - Refer for genetic testing
- Evaluate family members for RPE hypertrophy

The management of FAP includes surveillance and intervention strategies to reduce the risk of colorectal cancer. These strategies involve regular endoscopic examinations to detect and remove polyps, which can then be monitored for progression to cancer. Genetic testing is also available to identify individuals at risk, allowing for early intervention and monitoring.
• SHOULD OUR PATIENT BE WORKED UP FOR FAP?
  • ONLY 2 LESIONS IN ONLY ONE EYE
  • NOT AT RISK OF FAP
  • IF NO FAMILY HISTORY
  • IF NO CHANGE IN SIZE
  • IF NO NEW LESIONS DEVELOP
  • MONITOR YEARLY