

Oops Now what: Say this, not that. Optometric cases that turned out to be Neuro-Op

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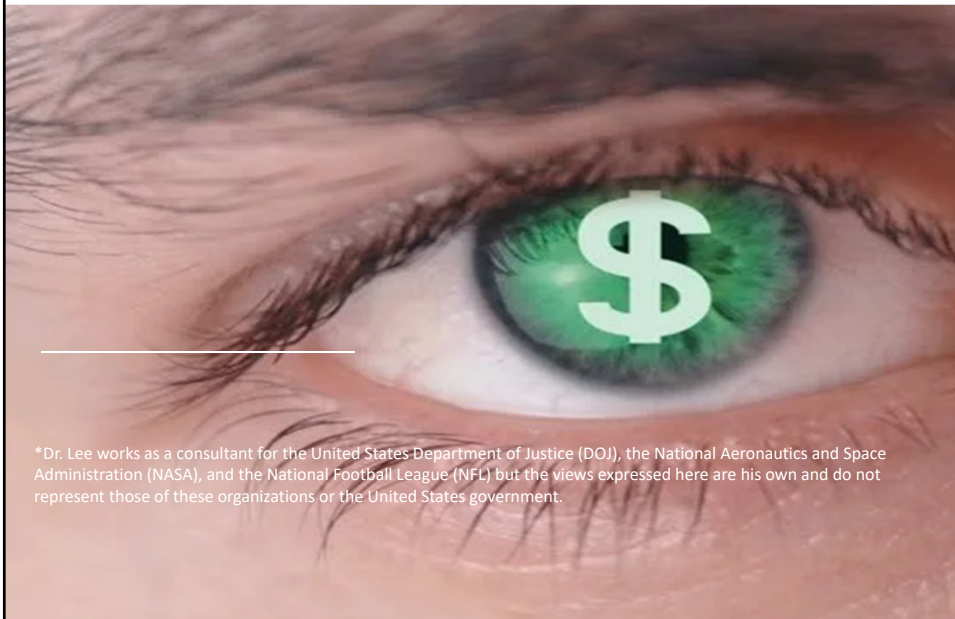


Weill Cornell Medical College



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## DISCLOSURES



\*Dr. Lee works as a consultant for the United States Department of Justice (DOJ), the National Aeronautics and Space Administration (NASA), and the National Football League (NFL) but the views expressed here are his own and do not represent those of these organizations or the United States government.

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THE FOLLOWING **PREVIEW** HAS BEEN REVIEWED FOR  
**ALL AUDIENCES**

**This talk might require a behavior change that  
might put you outside of your comfort zone.**

<b>PG-13</b>	PROJECT MANAGERS STRONGLY CAUTIONED
Some Material May Be Inappropriate for CIOs Under 13	
INTENSE METHODOLOGY MATERIAL, SCENES OF VIOLENCE	

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## Say this....not that

1. Check the medication list
2. Check pupil in light & dark (Don't use abbreviation "PERRLA" alone: Pupils, equal, round, reactive, light & accommodation)
3. Letting only tech only check pupil
4. Not taking "Blurred disc margins" seriously  
Misusing the term "papilledema"
5. Writing "Dysconjugate gaze" or "EOMI"
6. Thinking "optic atrophy" is a diagnosis

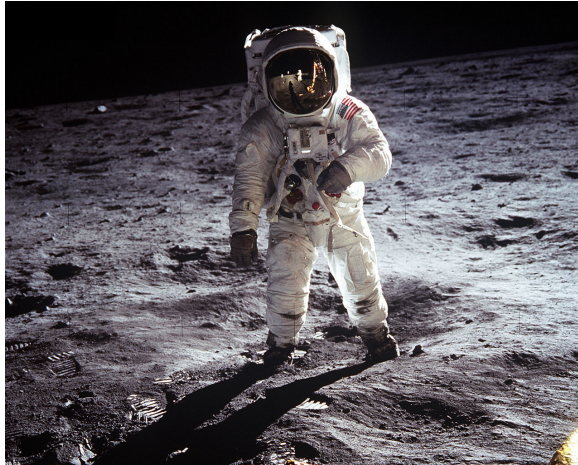
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On July 20, 1969, I was 5 years old, the moon landing was on tv....



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“Houston” was the first word spoken  
from the moon



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Texas Medical Center



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## Texas Medical Center

- Annual Patient Visits: 7.1 million
- Employees: 92,500 & Full-time Students: 34,000 & Volunteers: 12,000
- Residents and Fellows: 4,000
- Visiting Scientists, Researchers: 7,000
- Total Hospital Beds: 6,900 beds
- Annual Surgeries: 350,000

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## I care about feedback....

- Yes, fill out your evaluations
- Yes, say how great it was
- But I care more about you and your patients....
- Stop me at AAO & tell me how you saved someone by using the Force (Neuro-OP)
- That's powerful feedback

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1978: I wanted to be a doctor...2<sup>nd</sup>  
choice Jedi knight



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## Objectives: Meds that I H.A.T.E. in neuro-op clinic

1. **H**ydroxychloroquine/chloroquine retinopathy
2. **A**midarone optic neuropathy: Anterior ischemic optic neuropathy
3. **T**etracycline: pseudotumor cerebri
4. **E**thambutol optic neuropathy

The Erectile dysfunction agents (Viagra): Anterior ischemic optic neuropathy

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## Inappropriate medication lists....

- “See list
- “Some type of lung medicine”
- “heart medicine”
- “Some kind of antibiotic”



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# Our frenemy the EMR

**Xbialidocious, Fuzzy** Age: 5 yrs. M DOB: 1/1/06 Vitals: 70/45/92/25 Allergies: Penicillin, Nuts, Cats, M. TEST PAT: MILLER, JAMIE CAMBRIDGE On

**Patient Snapshot**

**Problem List**

- DM (diabetes mellitus)
- bhn
- Down's syndrome
- Adjustment disorder with depressed mood
- ENROLLED - COPO PROG (NOT DX, FOR PROB LIST ONLY)
- ANTICOAGULANT LONG-TERM USE
- ANTERIOR CHAMBER IMPLANTATION CYSTS
- GENETIC SUSCEPTIBILITY TO HEMACHROMOTOSIS
- GENETIC SUSCEPTIBILITY TO HEMACHROMOTOSIS
- Family planning, emergency contraceptive counseling and prescription
- Rheumatoid arthritis
- Paronychia or onychia of finger
- Down's syndrome
- UNSPECIFIED BACKACHE - lower back
- ENROLLED - ANTICOAGULATION SVC (NOT DX, FOR PROB LIST ONLY)
- GENETIC SUSCEPTIBILITY TO HEMACHROMOTOSIS

**Health Maintenance**

HEARING SCREENING (4 YEARS)	Completed
(HEDIS) HEPATITIS B (0-18 YEARS)	Completed
(HEDIS) DIPHTHERIA-TETANUS- PERTUSSIS	Completed
(HEDIS) POLIOVIRUS	Completed
(HEDIS) MEASLES/MUMPS/RUBELLA (1-5 YEARS)	Completed
(HEDIS) VARICELLA (1-18 YRS)	Completed

**Patient Lists**

- ASTHMA BTRPDS [552]
- TEST [1616]
- TEST [1183]
- TEST [3841]
- TEST [82]

**Allergies/Contraindications**

- PENICILLINS (PENICILLINS) Hives
- NUTS (TREE NUTS)
- CATS (CATS)
- NSAIDS Anaphylactoid reaction
- AMOXICIL-CLARITHROMY-  
LANSOPRAZ
- SOLFADIAZINE Hives

**Medications**

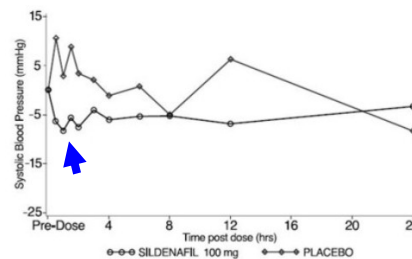
Insulin NPH & Regular Human (HUMULIN 50/50) 100	None Entered
Unifol (50-50) Subcutaneous Suspension	test
Insulin NPH & Regular Human (HUMULIN 50/50) 100	1 ml
unifol (50-50) Subcutaneous Suspension	1 vial
Insulin Glargine (LANTUS) 100 unit/mL Subcutaneous	15 tubes
Solution	testing refreshable
Fluoridolone 0.025 % Topical Cream	testing
Clonazepam (KLONOPIN) 0.125 mg Oral Tablet, Rapid	pm for pain
Dissolve	testing DO NOT FILL
Ceftriaxone (ZYRTEC) 1 mg/mL Oral Solution	TESTING DO NOT FILL - XL
Acetaminophen (CHILDREN'S TYLENOL MELTAWAYS) 80	Designation
mg Oral Tablet, Rapid Dissolve	Take 1 tablet daily
Bupropion HCl 300 mg Oral Tablet Sustained Release 24 hr	1 capsule daily; do not stop
Bupropion HCl XL 300 mg Oral Tablet Sustained Release 24	without consulting clinician
hr	pery; c
Lisinopril 40 mg Oral Tablet	Take 1 tablet every evening
Lorazepam 0.5 mg Oral Tablet	for cholesterol
Fluoxetine (PROZAC) 10 mg Oral Capsule	Take 1-2 puffs every 4 to 6
Lisinopril 20 mg Oral Tablet	hours as needed
Simvastatin 10 mg Oral Tablet	Epipen is chosen not twin
Albuterol Sulfate (PROAIR HFA) 90 mcg/actuation Inhalation	
HFA Aerosol Inhaler	
Epinephrine (EPIPEN) 0.3 mg/0.3 mL Intramuscular Pen	

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## Viagra

Biologically plausible mechanism: SBP drop 5-10 mm Hg at 2-4 hours after dose

- Non-arteritic AION
  - Hypotension
  - Hypoperfusion
- ED agents
  - Hypotension
  - Sympathomimetics
- My take: There is a weak but biologically plausible mechanism for NAION in ED agents



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## NAION & phosphodiesterase type 5 inhibitors (sildenafil)

- J Sexual Medicine 2015;12:139-51
- 103 centers (US and Europe)
- 43 definite NAION cases with PDE5i exposure in prior 30 days (five half lives)
- **OR = 2.15** (95% CI: 1.06, 4.34)
- Possible NAION cases included (n = 64) OR = 2.36 (95% CI: 1.33, 4.19)



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Bottom line:  
Someone needs to tell the patient....



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What will happen if you don't tell them  
about ED agents & NAION....



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*"The trouble with quotes on the  
Internet is that you can never know if  
they are genuine."*

*Abraham Lincoln*



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## How about amiodarone optic neuropathy?

- Macaluso et al
  - 73 optic neuropathy patients on amiodarone
  - Insidious onset, slow progression
  - Bilateral & protracted disc swelling
  - Resolved within several months after discontinuing
- Nagra et al
  - Three patients
  - Loss of visual acuity & visual field
  - Bilateral disc swelling slowly improved after discontinuation

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## Amiodarone optic neuropathy

- Amiodarone saves lives (stopping drug may kill people)
- Not dose-dependent phenomenon
- Seen in minority of patients on drug
- No proven pathogenic mechanism
- Can look just like non-arteritic anterior ischemic optic neuropathy
- Patients with other vasculopathic risk factors
- May not resolve after discontinuation of drug

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## Randomized prospective double masked trial showed no AION

- Mindel et al. Am Heart J. 2007;153(5):837-842
- Amiodarone (n = 837) vs placebo (n = 832)
- Median follow-up 45.5 months
- End point = bilateral vision loss
- No subject was removed from study because of bilateral vision loss
- Conclusion: Bilateral vision loss from amiodarone toxic optic neuropathy occurs infrequently if at all

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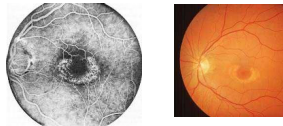
## What to tell the patient

- There is a risk of amiodarone optic neuropathy
- The risk factors for taking amiodarone overlap with the vasculopathic risk factors for NAION
- I will call your cardiologist about your medicine
- You need to make a risk benefit decision with cardiology

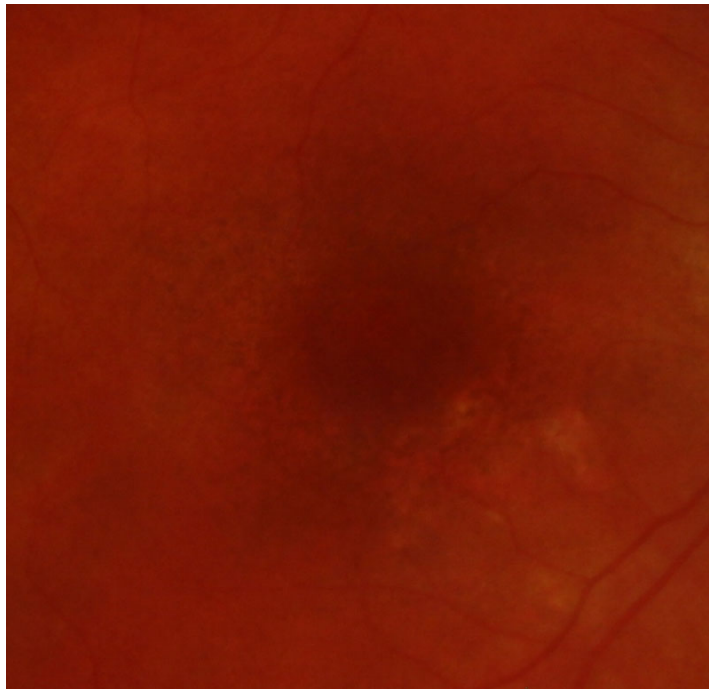
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## Hydroxychloroquine toxicity

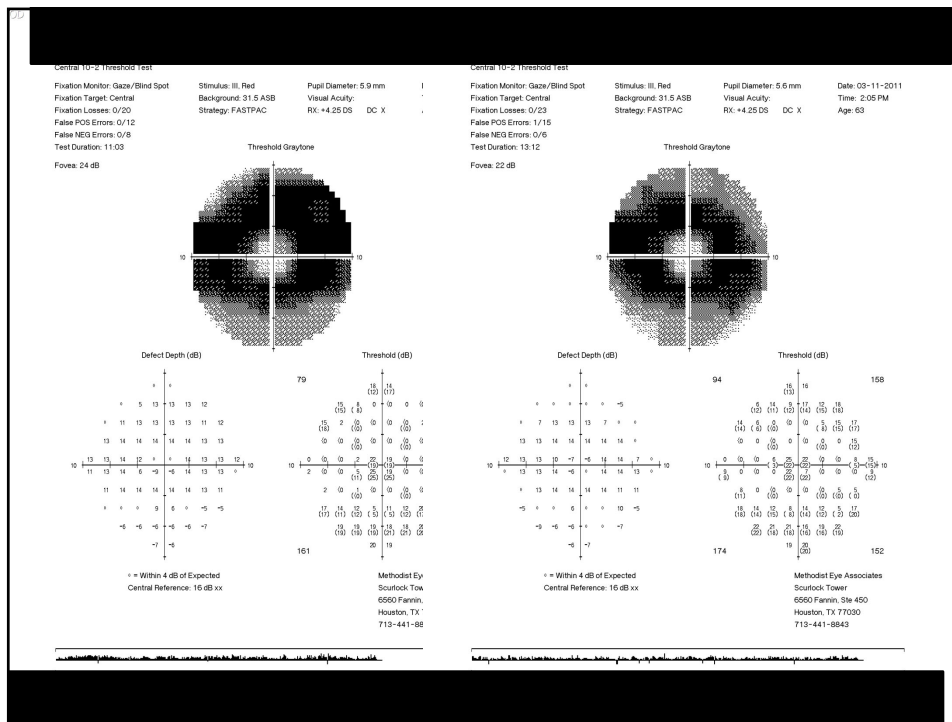
- Toxicity mechanism poorly understood
- Ring-shaped perifoveal zone spares central 2-3 degrees & extends out to 10 degrees
- “Bull’s eye” halo around the fovea
- Bilateral & often irreversible
- May progress even after drug discontinued



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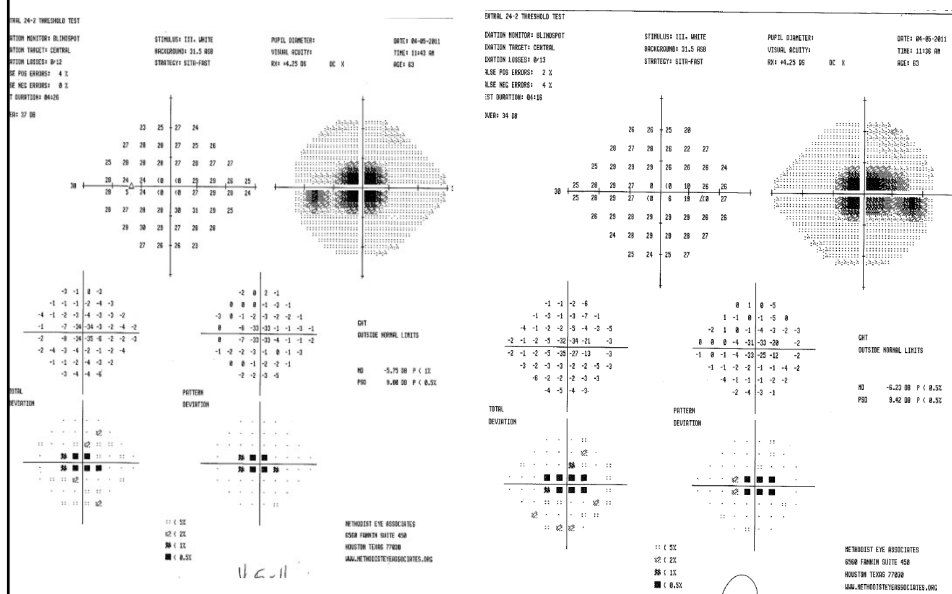


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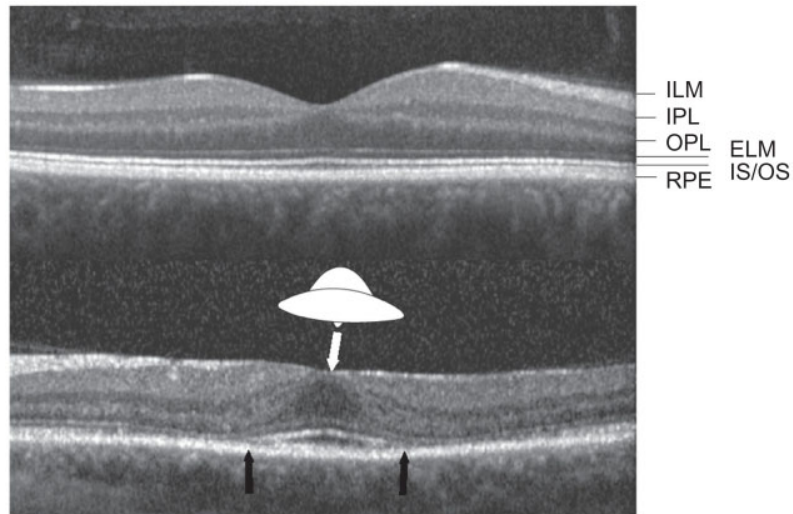
## HVF 24-2 (especially Asians)



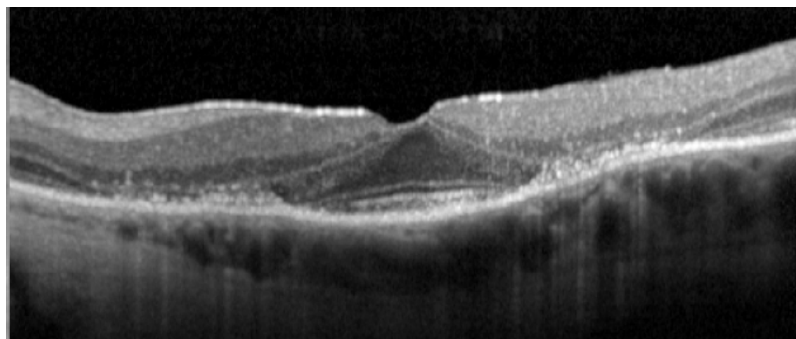
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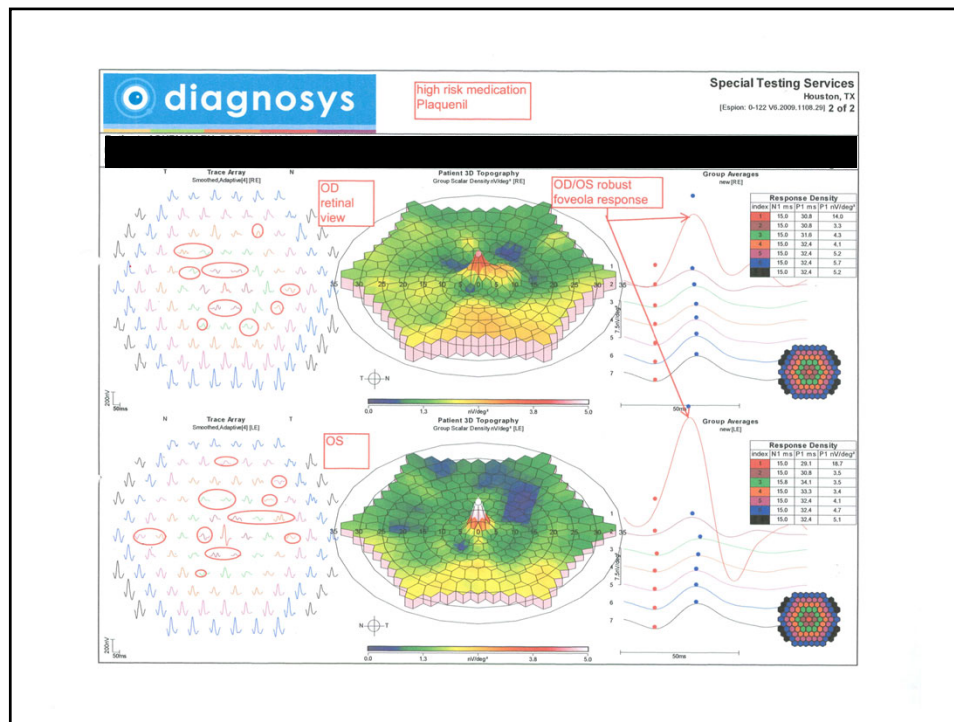
## HCQ (Plaquenil) screening SD OCT



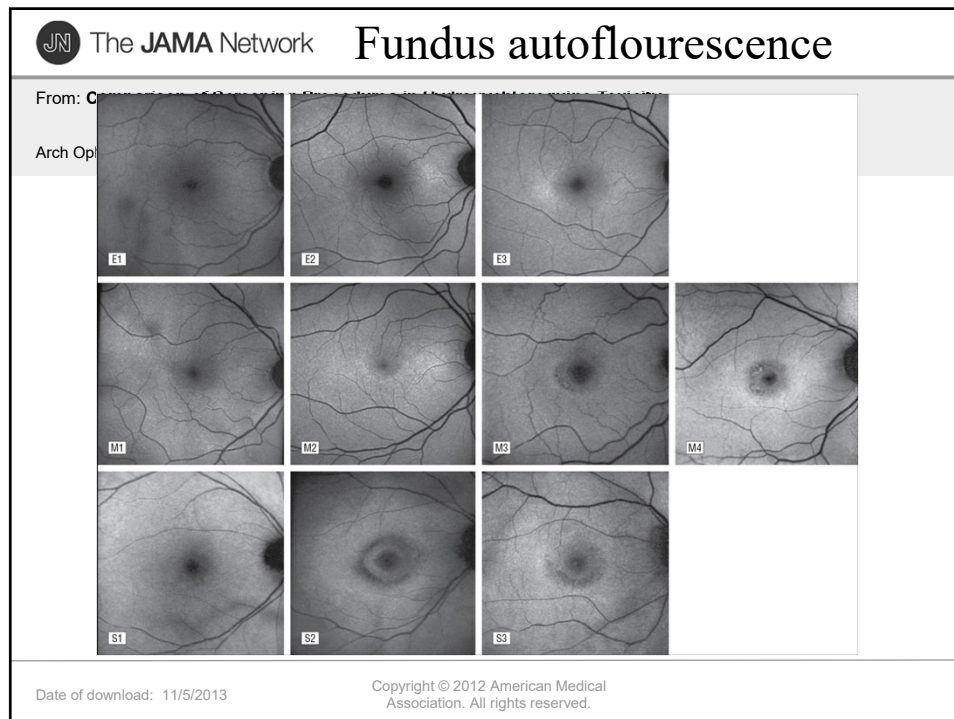
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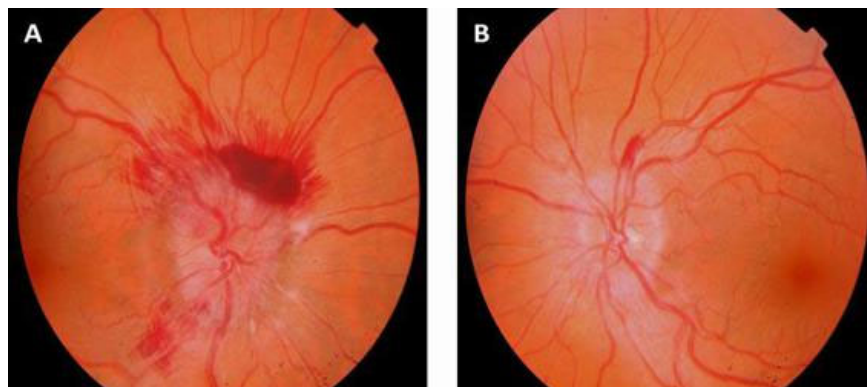
## Some recommendations

Marmor et al. Ophtahlmol 2011: 118:415-422.

- MERG, SD-OCT, FAF more sensitive than VF
- 10-2 HVF + one of the above
- If 10-2 HVF abnormal then complaints “should be taken seriously” (Asians 24-2)
- MERG may be “used in place of VF”
- Amsler grid no longer recommended
- Beware Tamoxifen

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A 15 y/o thin, male with HA, blurred vision OU and....



Which of the following medications can cause this finding?

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## Accutane (vitamin A analog) & tetracycline

- Pseudotumor Cerebri warning
- “Accutane use has been associated with a number of cases of pseudotumor cerebri”  
Some cases involved concomitant use of tetracyclines
- Concomitant treatment with tetracyclines should therefore be avoided “

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## Ethambutol toxicity

1. Painless, progressive, bilateral visual acuity loss
2. Proven causality, dose related optic neuropathy
3. Color loss (e.g. blue-yellow dyschromatopsia)
4. Central or cecocentral scotomas
5. Initially normal optic nerve (retrobulbar) followed by temporal optic disc pallor OU

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## Ethambutol screening

- Screen high risk patients (high dose > 15 mg/kg/day, renal failure, long duration):  
Follow q month (longer for low risk)
- Warn patients about toxicity
- Baseline exam & visual field
  - Color testing
  - Dilated fundus exam
  - Automated 24-2 or 10-2/Amsler (self check)
  - If any change come in right away

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## Homeland security risk stratification for ethambutol toxicity: Dose! Dose! Dose!



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### Do not confuse the screening strategies

- “Imp: No evidence of EMB toxicity
- Plan: 1 year”
- This is NOT the correct screening strategy
- PS: Weight loss will change dose (s/p lung transplant or MAI or TB patients lose weight over time unintentionally)

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- (e) During medical consultations in the course of anti-TB treatment including EMB, all patients should be ~~assessed~~ clinically for symptoms of visual disturbance. Enquiring monthly about visual symptoms is advisable.
- (f) Directly observed treatment (DOT), apart from ensuring treatment adherence, also allows health care workers to monitor the patients closely for such symptoms.

**Table 2. British Thoracic Society Guidelines - Chemotherapy and management of tuberculosis in the United Kingdom: recommendations 1998\*.**

**Special precautions and pretreatment screening point (1)**

Because of the possible (but rare) toxic effects of ethambutol on the eye, it is recommended that visual acuity should be tested by Snellen chart before it is first prescribed. The drug should only be used in patients who have reasonable visual acuity and who are able to appreciate and report visual symptoms or changes in vision. The notes should record that the patient has been told to stop the drug immediately if such symptoms occur, and to report to the physician. The general

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You are going to see more ethambutol  
toxicity  
22 countries have 80% of TB



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## Scary math: 100,000 blind

[J Neuroophthalmol](#). 2008 Dec;28(4):265-8. doi: 10.1097/WNO.0b013e31818f138f.

**Ethambutol optic neuropathy: how we can prevent 100,000 new cases of blindness each year.**

[Sadun AA](#), [Wang MY](#).

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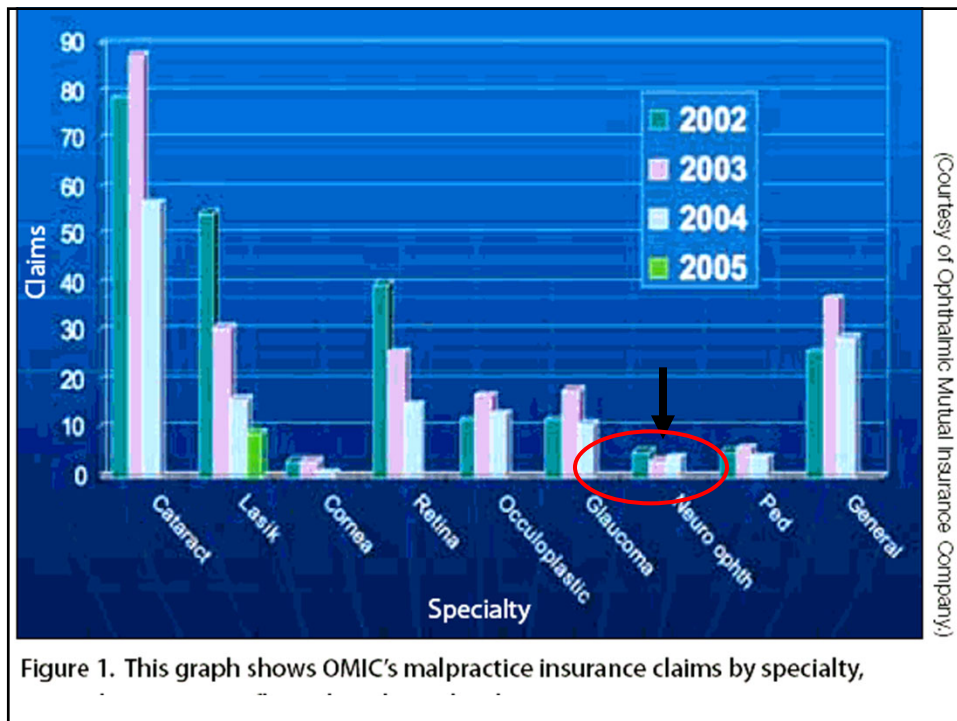
PS: The Erectile dysfunction agents (Viagra): Anterior ischemic optic neuropathy

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## Say this....not that

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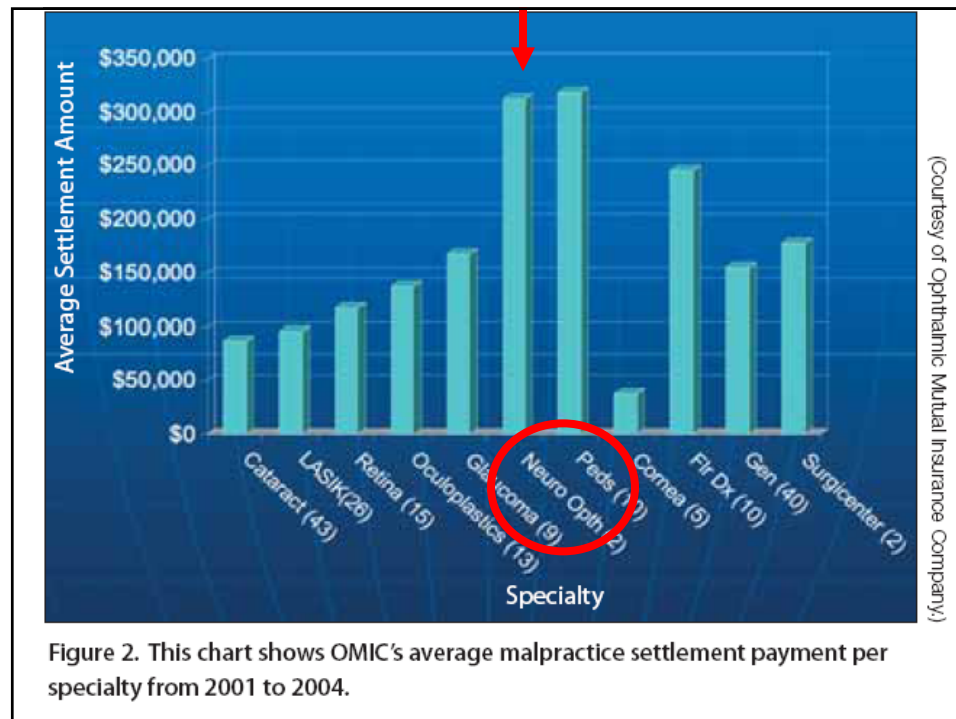


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## The big five

- Refractive/cataract surgery (missed endophthalmitis)
- Diabetic retinopathy
- Glaucoma
- Delayed diagnosis of brain tumor
- Retinal detachment

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## A case from the optometric literature

- 12 y.o. girl (routine eye exam)
- Well respected, well trained, optometrist
- BCVA: 20/40 OD and 20/20 OS
- Normal eye exam
- Confrontation visual field normal OU
- No RAPD
- Normal fundus
- No strabismus, no anisometropia
- Dx: “Amblyopia OD”
- Plan: “RTC 1 year”

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## One year later

- Vision 20/200 OD and 20/20 OS
- Visual field: Junctional scotoma
- Referred to neuro-op: MRI: Large sellar tumor (craniopharyngioma)
- Hydrocephalus
- Neurosurgery: Difficult resection
- NLP OU in post-operative recovery room
- Family and doctor are “devastated”

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## Litigation follows

- OD: Insurance company refuses to settle
- Case goes to trial
- 9.2 million dollar judgment AGAINST optometrist
- No real winners here
  - Sad, blind girl
  - Angry parents
  - Sad and angry doctor
- Optometrist closes practice after 30 years

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## Jury decides against optometrist, awards \$9.2 million for missed tumor diagnosis

Primary Care Optometry News, March 1997



COMMENT



[+ ADD TOPIC TO EMAIL ALERTS](#)

FT. LAUDERDALE, Fla. — A six-member jury here awarded a \$9.2 million judgment to a teen-age girl and her family after deciding an optometrist's diagnosis of amblyopia delayed treatment of a brain tumor near the girl's optic nerve.

The patient, now age 16, underwent two operations for the tumor and was left totally blind, according to Andrew B. Yaffa, an attorney for the family. "This case stands for taking a full and adequate patient history, doing a complete exam and learning what the patient's problems are before settling on a benign diagnosis of amblyopia," Mr. Yaffa said.

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## Amblyopia definition

- Unilateral (or, less often, bilateral)
- Reduction of best-corrected visual acuity (BCVA)
- Otherwise normal eye exam
- Developmental disorder of CNS
- Deprivation amblyopia has structural abnormality involving the eye or visual pathway but BCVA loss cannot be attributed only to the effect of the structural abnormality

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## You must document an amblyogenic etiology

- Strabismic
- Refractive
- Anisometropia
- High bilateral refractive (isoametropic)
- Visual deprivation
  - Media opacities
  - Ptosis
  - Occlusion (reverse)

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## Beware abnormal exam findings other than decreased visual acuity

- No RAPD (Less than 0.3 log unit can be seen)
- No optic atrophy
- No visual field defect
- No other neurologic signs/symptoms

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## Document stability over time

- Amblyopia does NOT worsen in adulthood
- Patient may not notice visual loss in amblyopic eye
- Amblyopia patients get brain tumors

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## Back to the case from optometric literature

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- Plan: “RTC 1 year”

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## Ask why

- Why no RAPD?
- Why was confrontation visual field normal?
- Why was fundus exam normal?
- Why was the normal eye exam a BAD thing for amblyopia diagnosis?
- Why was the OD dx of “amblyopia” incomplete?
- Why was the plan: “RTC in 1 year” inappropriate?

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## Take home messages for amblyopia in adult

- Behavior change: Don't be lazy: ("POH: lazy eye OD")
- Define amblyogenic etiology (strabismic, anisometropic, deprivation)
- Document, document, document (see something, say something)
- Must be stable over time (document BCVA)
- Normal eye exam can be deceiving (bilateral = no RAPD, retrobulbar = no optic atrophy initially)
- Perform an OCT and a formal visual field (HVF) in every patient with unexplained visual loss

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## 55 yo WF referred for ptosis secondary to levator dehiscence OU



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Tacoma Narrows Bridge:  
Small holes matter (like the pupil)



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Checking pupils in ambient light easily  
misses Horner syndrome  
("PERRLA"  $\neq$  NORMAL)



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## Here's a pearl don't use "PERRLA"

- Pupils equal, round, reactive to light and accommodation (PERRLA)
- Pupils can be equal, round & reactive to light and accommodation and have a HORNER syndrome
- PERRLA only checks PNS pathway



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## The Perils of PERRLA

### The Perils of PERRLA

Leonard A. Levin, MD, PhD

[\[+\] Article and Author Information](#)

*Ann Intern Med.* 17 April 2007;146(8):615-616

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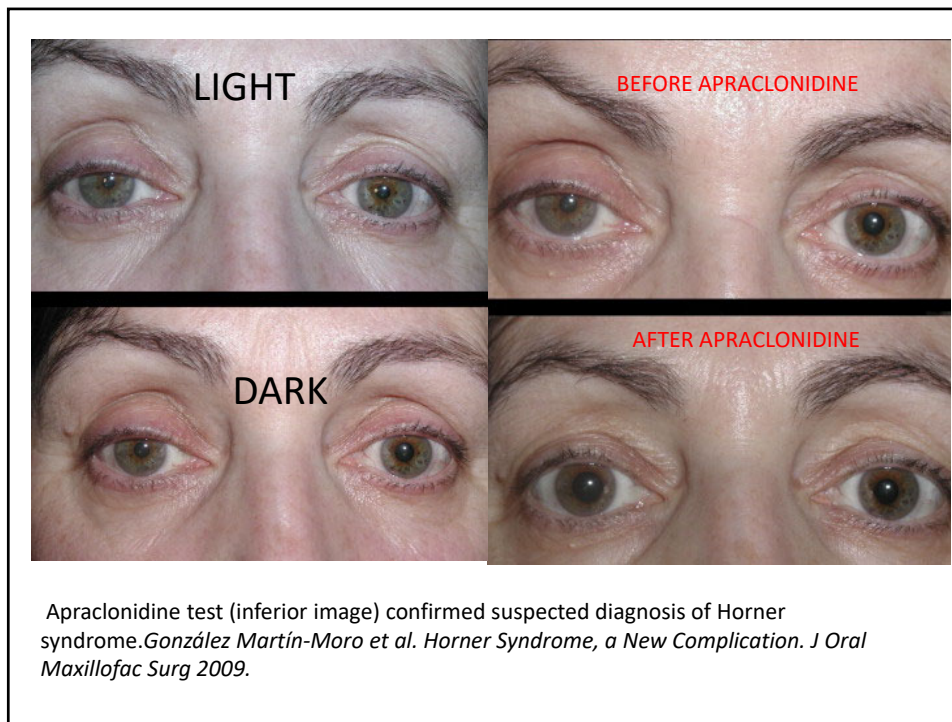
Article

References

Comments

*Background:* The pupillary examination is arguably the single most useful clinical test of the nervous system in the conscious or unconscious patient. When properly done with a few swings of a light, the examination can assess the functional status of the retina, optic nerve, contralateral optic tract, cranial nerve III, dorsal midbrain, and sympathetic chain from the hypothalamus down to the lower cervical spinal cord and up to the cavernous sinus and orbit. The results of normal pupil testing are frequently documented with PERRLA (pupils equal, round, reactive to light, and accommodation). But PERRLA is not only incorrect and inexact—it is also, more seriously, incomplete.

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## Horner syndrome

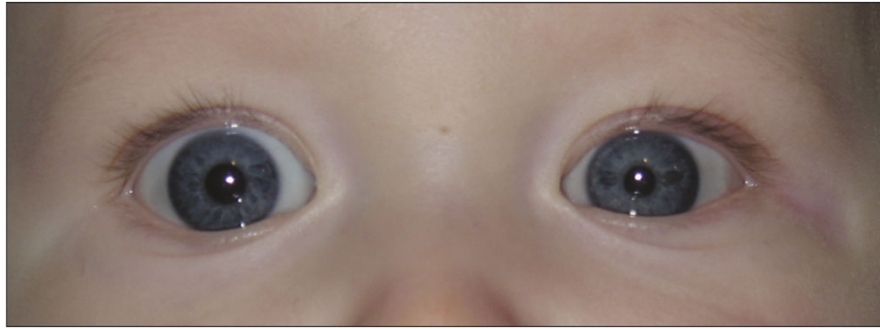
- Anisocoria is greater in the dark
- If pupil tested in light only then easy to miss subtle anisocoria: PERRLA can miss Horner pupil
- Ptosis is always mild in HS (some cases: no ptosis)
- Book Horner does not look like real world sometime



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From: **Atypical Acquired Pediatric Horner Syndrome**

Arch Ophthalmol. 2010;128(7):937-940. doi:10.1001/archophthalmol.2010.119



**Figure Legend:**

Case 1 showing miosis with only a trace of ptosis of the left upper eyelid. Reverse ptosis of the left lower eyelid is present.

Date of download: 12/2/2012

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From: **Atypical Acquired Pediatric Horner Syndrome**

Arch Ophthalmol. 2010;128(7):937-940. doi:10.1001/archophthalmol.2010.119



**Figure Legend:**

Case 3 with subtle miosis of the left pupil and no ptosis.

Date of download: 12/2/2012

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## How is your tech checking the pupil?

- Behavior change(s)
- If the chief complaint is diplopia or ptosis, let me check the pupil before dilation
- If you have to lift a droopy eyelid before putting in the drops come & get me
- If you have a question about an afferent pupillary defect (??RAPD) come & let me check it too

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## Now what: Say this, not that

- Not PERRLA
- (or if using PERRLA then PERRLDA and no RAPD)
- Say this: Pupils, round: 5 mm OU (dark) 3mm OU (light), no RAPD

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## Horner syndrome RE “Normal MRI head”



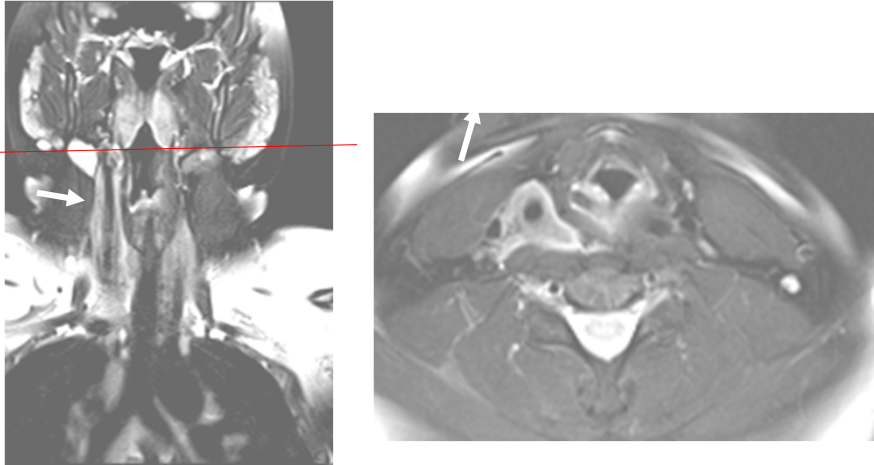
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## Oops..Now what Behavior change regarding tumors

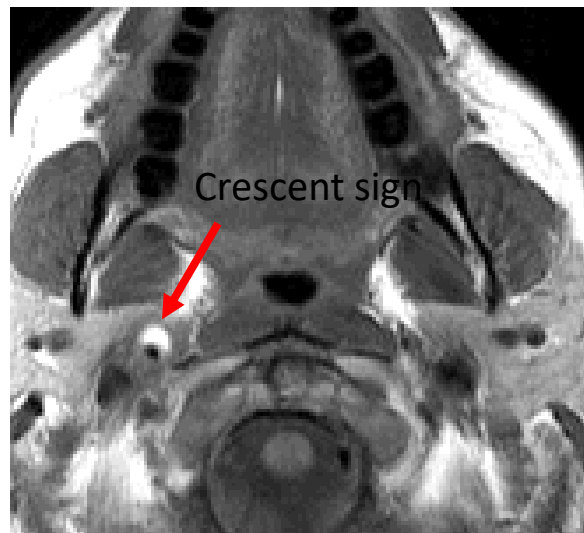
- Old: “PMH: breast cancer”
- New: “PMH: Stage 4 breast cancer with distant metastasis” or “Stage 0 DCIS”

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Imaging head alone is false sense of security: Pericarotid biopsy proven sarcoid



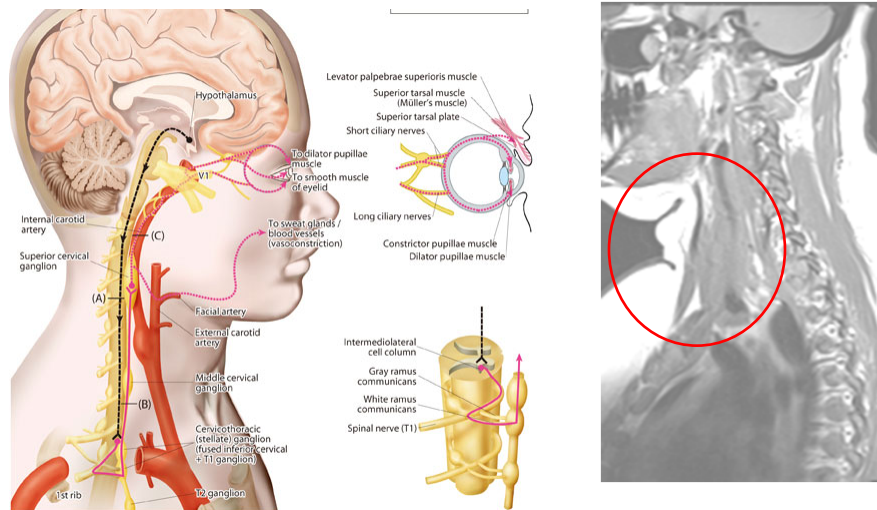
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Life threatening cause of Horner syndrome  
= Carotid dissection is extracranial in NECK

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## MRI head to neck T2 level



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## You can image the entire pathway with one MRI scan in Horner syndrome

- You could do many overlapping expensive studies
  - MRI head
  - MRI neck
  - CT neck
  - CXR with apical views
  - CT chest
- Or....you could do one scan (MR head to apex of lung (T2 level in chest))
- Sagittal & parasagittal imaging on the SIDE of the lesion
- A Horner protocol MRI

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## “PERRLA” failure #2

- There is no assessment of the relative afferent pupillary defect (RAPD) in PERRLA
- Proper format
  - OD: Dark 5 mm→ Light 3 mm No RAPD
  - OS: Dark 6 mm→ Light 3 mm



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Who's fault is it if the resident/fellow/technician doesn't check the pupil properly?



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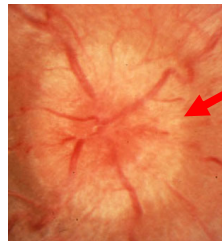
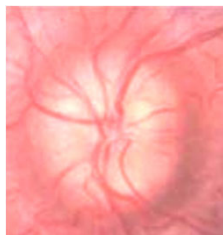
## Now what: The behavior change

- You: Strike PERRLA from your lexicon & your encounter forms, check tough ones personally
- Your tech: Don't use PERRLA, call the doctor for the tough ones

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## “Blurred disc margin” is a worthless description of what you already know

- Does NOT differentiate pseudopapilledema from true papilledema (Both have “blurred disc margins”)
- Is disc margin blurred because of something above (peripapillary nerve layer) or below (deeper like drusen?)



LOOK HERE

Obscuration of peripapillary NFL (blurred VESSELS)

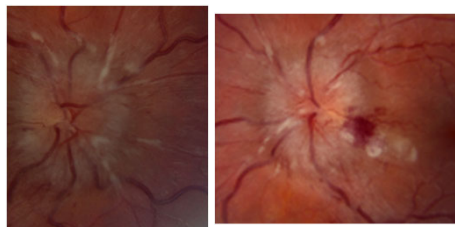
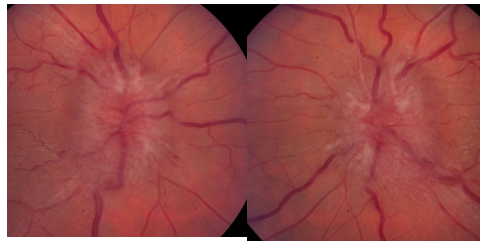
80

## Never underestimate optic disc edema: When to call neuro-op!

- “Next available” optic disc edema
  - Unilateral nonarteritic AION
  - Unilateral optic neuritis
  - Unilateral “neuroretinitis”
- Not “next available” (pick up the phone)
  - Arteritic AION
  - Bilateral optic disc edema (including “neuroretinitis”)
  - Severe visual loss with disc edema
  - Chronic atrophic papilledema
  - Optic disc edema in elderly (rule out giant cell)

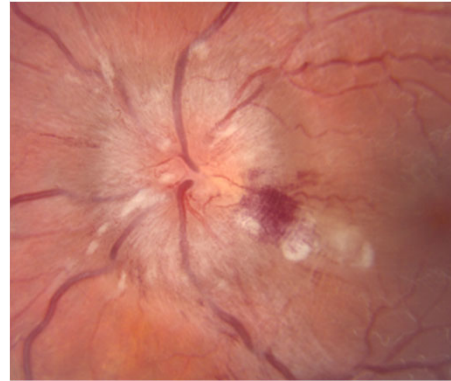
81

## Which is tumor & which is pseudotumor cerebri?



82

Don't use "blurred disc margin"



83

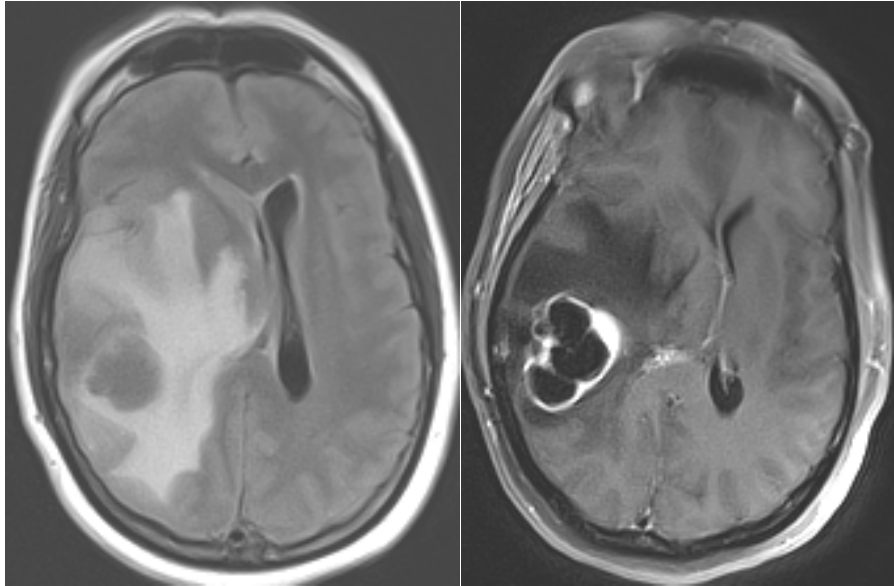
Little edema



84

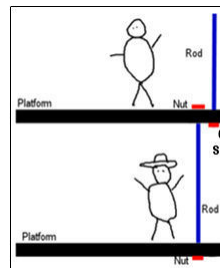
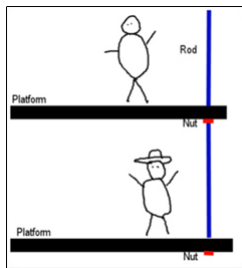


## Little edema can be Big problem



85

## Little findings can be big problems Hyatt Regency Kansas City



Little change  
Big Problem

86



## No what: The behavior change

- You: Don't write "papilledema" or use word "papilledema" (increased ICP) when you mean optic disc edema
- Your tech: Don't let your doctor write "papilledema" for optic neuritis, NAION, pseudopapilledema, funny discs, etc.

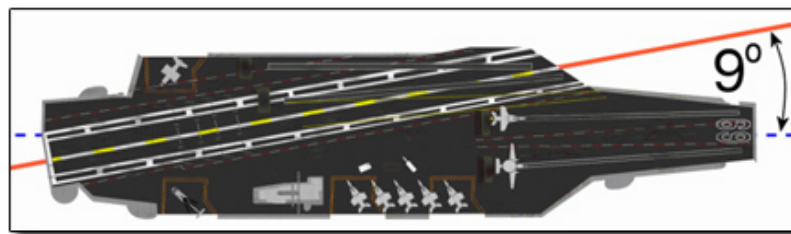
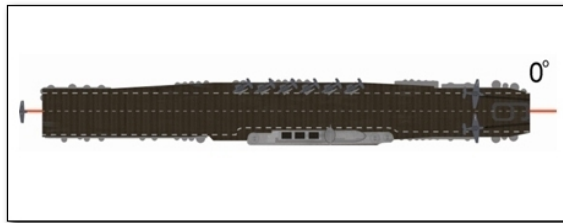
87

## Don't use "EOMI" as your sole documentation of motility exam

- Not "EOMI"= Extraocular muscles intact
- Primary position deviation will be missed if no cover-uncover testing performed
- Small incomitant deviation will be missed if cover-uncover test not performed in diagnostic positions of gaze
- Record the angle of deviation: (e.g., 15 ET in primary, 30 in left gaze, 0 in right gaze)

88

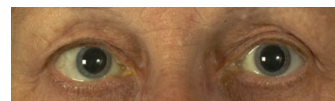
Angled not straight on aircraft carrier  
Time to stop and start matter



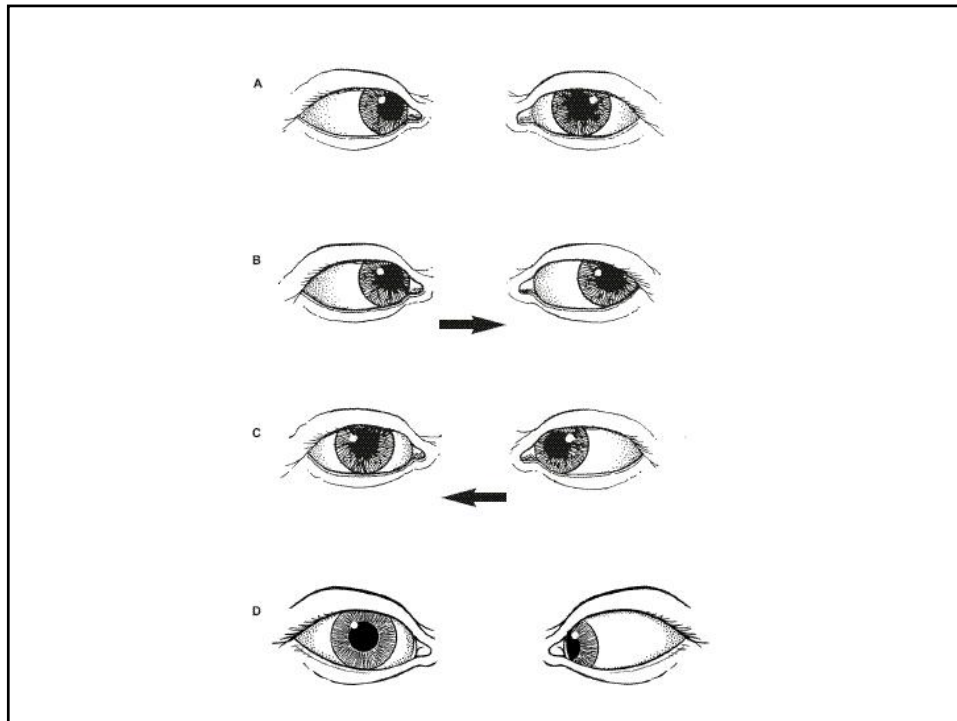
89

## Sixth nerve palsy can be “EOMI”

- Need cover-uncover test in diagnostic positions of gaze to find small esotropia
- Ductions-versions can be normal in patients with ocular motor cranial neuropathies
- Book sixth=complete abduction deficit: 50 ET!
- Real world 6<sup>th</sup> = small incomitant ET in right gaze only



90



91

## Cocoanut Grove fire, Boston 1942: Location and direction matter



Only one exit  
Inward swinging doors



92

## Now what: Behavior change

- You: Do cover/uncover test for patients with diplopia & test in diagnostic positions of gaze
- Don't use symptom (i.e., "diplopia") or sign (i.e., hypertropia) as diagnosis or impression
- Your tech: Tell eyeMD if diplopia is problem, don't let people leave clinic without a diagnosis
- Diplopia is NOT a diagnosis

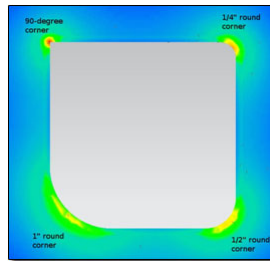
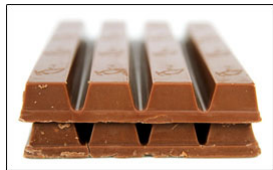
93

## Why are airplane windows round?



94

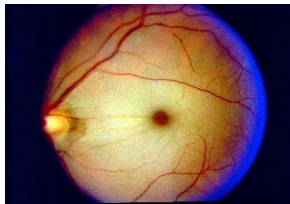
## Round not square: Comet airplane Looks good and is good are not the same



**Radomile C. 6 Small  
Math Errors That  
Caused Huge Disasters**

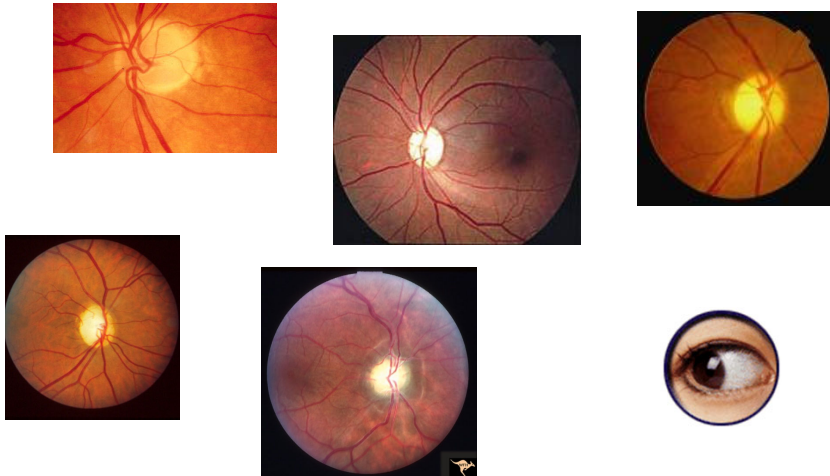
95

## Most of your diagnoses are obvious! (Augenblick)



96

## Optic atrophy is NOT a diagnosis! Not Augenblick!



97

Optic atrophy



98

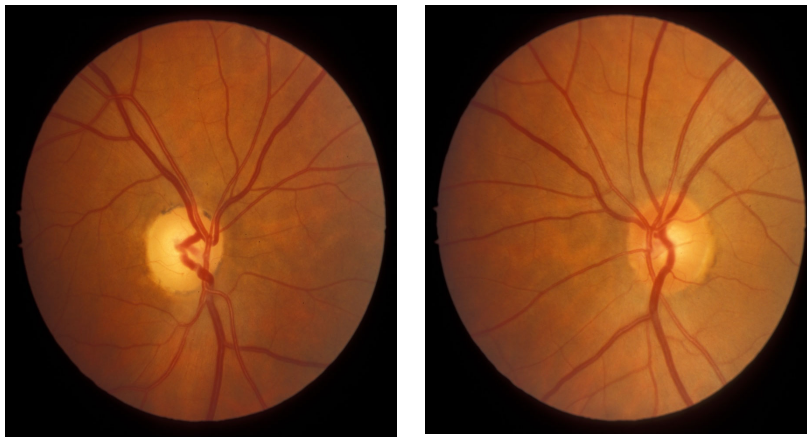
Is this nerve pale? Mild pallor? Temporal pallor? Optic atrophy?



Look for clinical signs of optic neuropathy  
(RAPD, visual field, fellow eye, OCT)

99

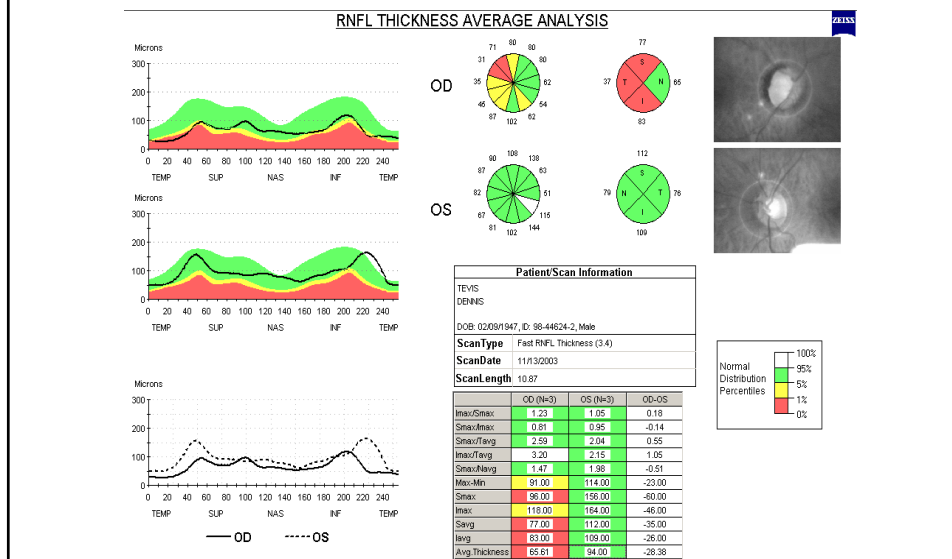
### Determination of Pallor vs No Pallor



100



# OCT can see better than me



101

## Am I pale?



102



Am I pale?



103

Common things are common

- Is it old AION
- Is it old optic neuritis?



104

## Dad's rules of DDx



- If it sounds like a duck, looks like a duck, & acts like a duck then it's a duck
- Is it old AION?
  - Disc edema
  - Vasculopath
  - Older patient
  - Static course
- Is it old ON?
  - Younger
  - Recovered
  - MS history



105

When making your differential diagnosis...think horses



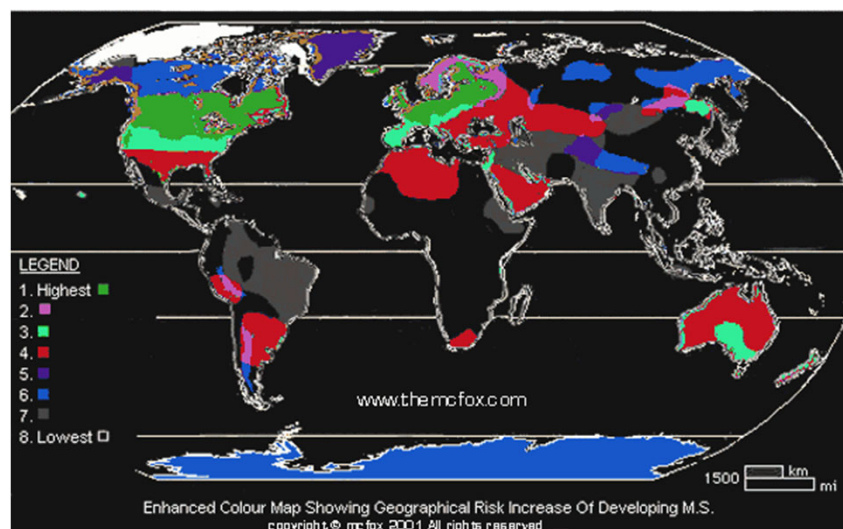
106

## Uncommon presentations of common diseases are COMMON



107

## MS: Old optic neuritis?



108

## If not AION or ON then more history & exam

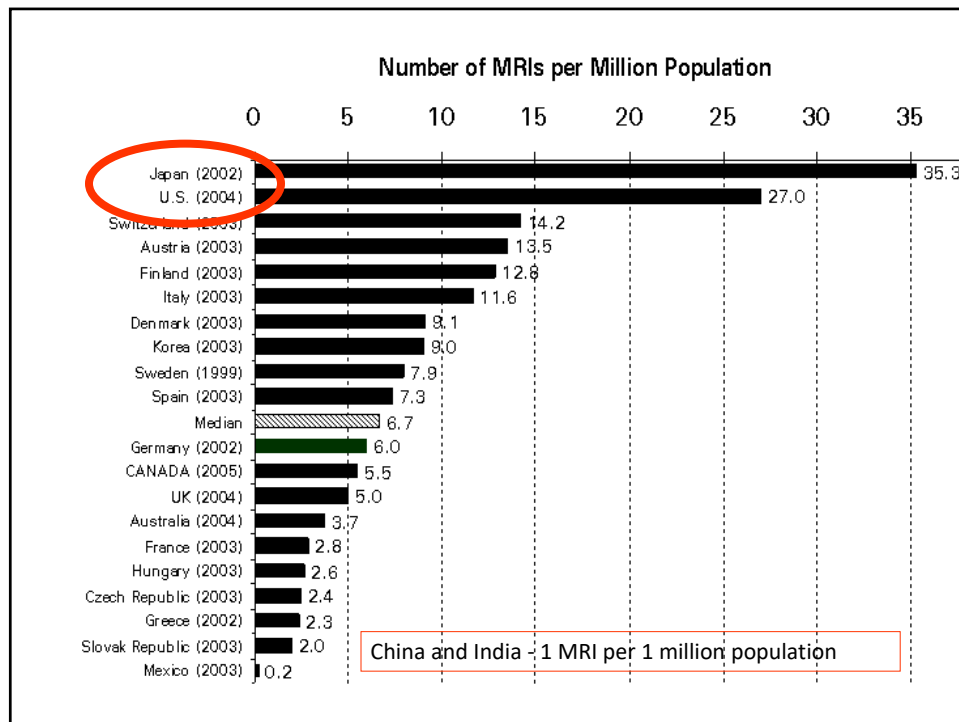
- Bilateral progressive central-cecocentral scotoma => B12/folate/Leber's hereditary optic neuropathy/ethambutol toxicity
- Chronic progressive optic neuropathy => compressive lesion (get formal fields)
  - Bitemporal hemianopsia: Chiasmal
  - Homonymous hemianopsia: Optic tract
- Uveitis (old or new) Sarcoid, syphilis

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## If history & exam come up short then image unexplained optic atrophy

- MRI head/orbit fat suppression and gadolinium (optic nerve protocol)
- If suspicion low for compression or cost is an issue in your part of the world you might choose observation (if static, old NAION then no imaging) or CT scan with contrast
- Optic atrophy can always be a tumor!

110



111

## Directed evaluation vs. shotgun

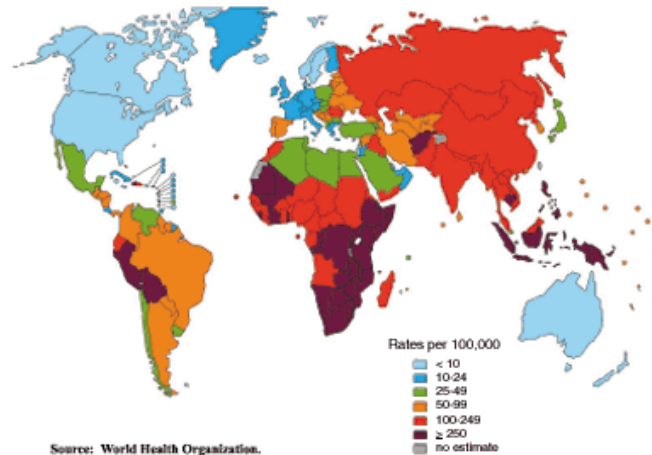


www.myweapons.nl

112

## Tuberculosis worldwide

Estimated Rates of New Cases of Tuberculosis, 1997



113

## Syphilis worldwide



114



## Round up the usual suspects



115

## Optic atrophy is NOT a diagnosis

- Impression equals diagnosis = most of your clinic day!
  - Cataract
  - CRVO
  - CRAO
  - RD
- Optic atrophy is NOT a diagnosis
  - Could be a compressive lesion
  - Image if unexplained optic atrophy (don't write “? Mild”)
  - If not imaging document WHY (e.g., “I believe that this is old NAION and I am following this patient”)
  - Document RATIONALE for decision making

116

## Behavior change

- You: Don't use "optic atrophy" as a diagnosis
- Your techs: Don't let people leave clinic with a photo or OCT or chart that says "optic atrophy" and has NO etiologic diagnosis
- Reschedule patient or refer
- You need time to start and stop (or you will crash in neuro-ophthalmology)

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## Five behavior changes TODAY

1. Check pupil in light & dark (not "PERRLA")
2. Don't let technician be only pupil exam for tough ones
3. Avoid "Blurred disc margins" & take the finding seriously (i.e., "? papilledema")
4. Don't use vague motility terms like "dysconjugate gaze" or "extraocular muscles intact ("EOMI")
5. Remember: "optic atrophy", "diplopia", "esotropia" are not diagnoses (PS: Neither are "ptosis" or "blurred vision" or "unexplained visual loss")

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## Summary: Behavior changes

### Say this...

1. Pupils round, 5 mm OU (D) and 3 mm OU (L); No RAPD
2. Grade x Frisen scale optic disc edema
3. 15 ET in primary, 30 ET in left gaze, 0 in right gaze
4. Sector optic atrophy secondary to prior NAION

### ....Not that

1. "PERRLA"
2. "Blurred disc margins"
3. "EOMI"/"Dysconjugate gaze"
4. "Optic atrophy"

119

There is a difference between data and information

- DATA  
–28 17 26 80 81
- INFORMATION  
–(281) 726-8081
- If you have questions call me or email me [AGLee@tmhs.org](mailto:AGLee@tmhs.org)

120

End with a philosophical question & two really quick cases.  
Why are you here... because you believe as we all do that you  
can....?



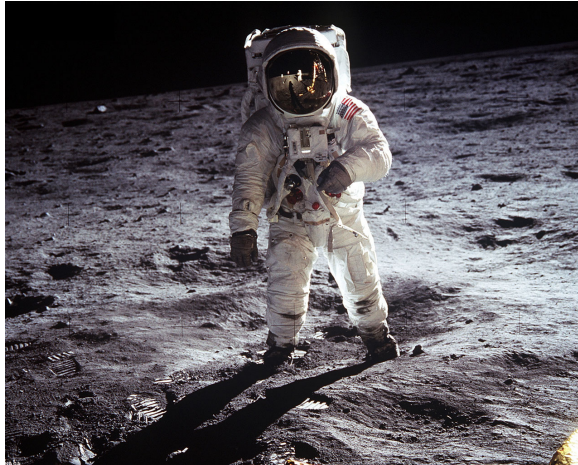
121

On July 20, 1969, I was 5 years old, the moon  
landing was on tv....



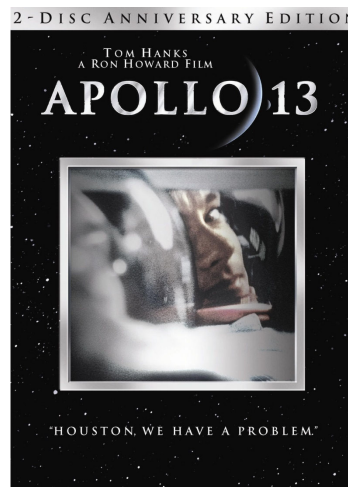
122

“Houston” was the first word spoken  
from the moon



123

April 1970: “Houston, we’ve had a  
problem”—Jim Lovell



124

## Jim Lovell



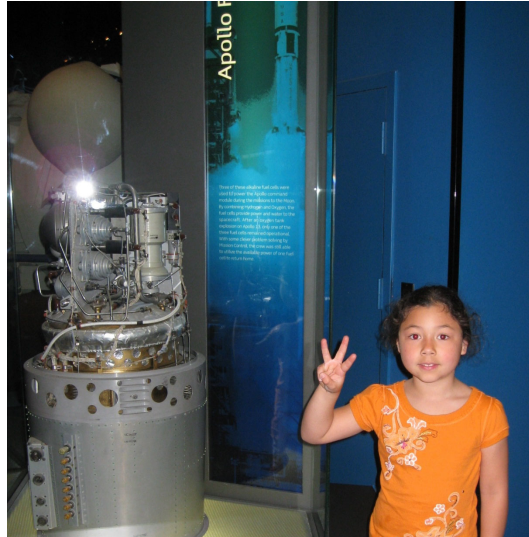
125

## Half the spacecraft panel lost



126

## Apollo fuel cell



127

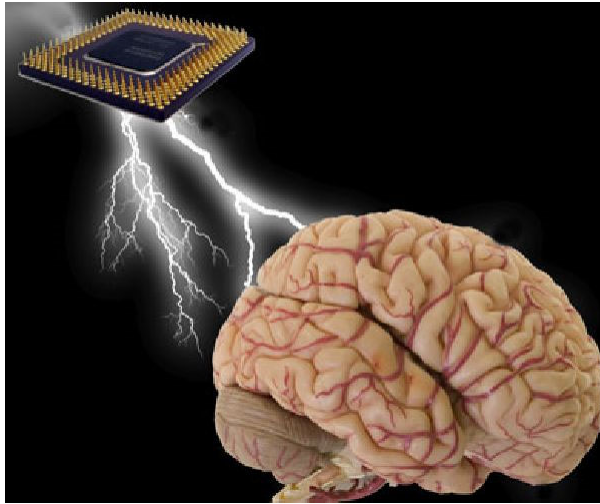
## Most of the computing power was human brains at NASA



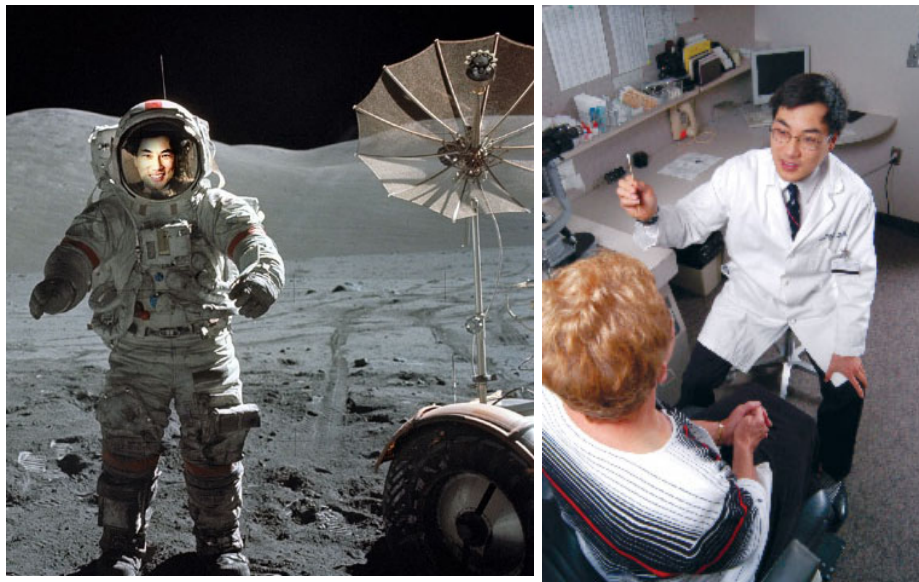
128



It was human brain power that brought  
Apollo 13 home....



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Today, I get to work with real rocket scientists at  
NASA in Houston



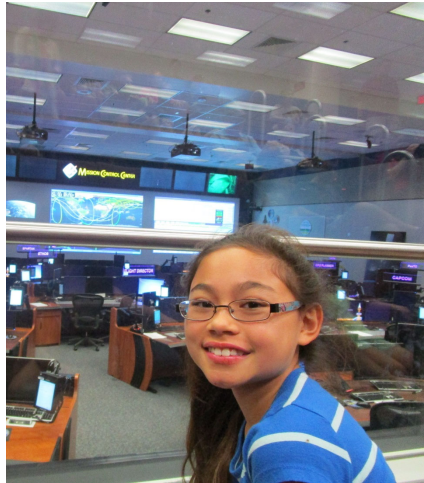
131

Houston is home to JSC NASA



132

This is what mission control looks like  
now



133

Go out and...



134



Thank you for your time & attention



135

## Chief complaint: NONE

- 73-year-old WF
- Chief complaint: NONE now (2010)
- PMH: Paraneoplastic optic neuropathy, recovered
- CXR: Small cell carcinoma of lung
- Resected, chemotherapy, radiation in 1997
- Published: Luiz JE, Lee AG, Keltner JL, Thirkill CE, Lai EC. Paraneoplastic optic neuropathy and autoantibody production in small-cell carcinoma of the lung. *J Neuroophthalmol.* 1998;18:178–181.

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## Follow up 2010

- Pt: "You don't remember me do you Dr. Lee?"
- Me: "Well,...I um....sure...maybe"
- Pt: "I had lung cancer & you found it thru my eye"
- Me: "Really"
- Pt: "Yeah, you wrote it up in a journal"
- Me: "Oh, yeah, sure, now I remember. How are you, why are you coming today?"
- Pt: "I just wanted to tell you that I was still alive and it is been 14 years, so thanks."

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## Longest known survivor

### Long-Term Survivor of Paraneoplastic Optic Neuropathy

**S**mall cell lung cancer carries a very poor long-term prognosis. In a survey performed at the Mayo Clinic from 1997 to 2003, the 5-year survival rate was only 9% (1). In addition, to our knowledge, the longest published survival duration for paraneoplastic optic neuropathy secondary to small cell lung cancer has been 8 years (2). We wish to provide an update on a patient previously reported by one of us (A.G.L.) in this Journal in 1998 (3) who returned 14 years later without evidence of tumor recurrence and believed to be in clinical remission. The earlier detection of the tumor from her neuro-ophthalmologic examination followed by timely systemic treatment may have contributed to her favorable outcome. To the best of our knowledge, she is the longest survivor of paraneoplastic optic neuropathy secondary to small cell lung cancer. At the time of her diagnosis, she underwent surgery, chemotherapy, and radiation therapy and was believed to be in remission at the last follow-up.

The patient, a 73-year-old white woman, was last seen in the neuro-ophthalmology clinic on July 20, 2010. She was complaining of blurred vision in the left eye that had worsened since sustaining a fall on March 1, 2010. She was seen by her neurologist who obtained a brain MRI that showed no focal lesions.

in March 2010 showed no evidence of recurrent or metastatic disease. The patient returned to The Methodist Hospital after 10 years of follow-up to specifically report on her progress and survival from small cell carcinoma of the lung.

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**Sushma Yalamanchili, MD**

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Iowa City, Iowa*  
*Department of Ophthalmology, UTMB-Galveston  
Galveston, Texas*

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## His name is Andrew....

- 33-year-old WM
- Transient dizziness, blurry vision, followed by loss of consciousness after watching bungee jumpers at Iowa St. Fair
- On regaining consciousness, bilateral ptosis, exotropia: Noncontrast cranial CT in ER was “normal” MRI with contrast : “normal”
- About to be discharged

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## MRI head negative



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## Course

- Top of the basilar syndrome
- Intravascular tPA
- Locked in syndrome
- Recovered slowly
- Rehab, walked out of hospital
- Writing a book about his experience called “One Fine Day” ....

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### Years later...

- Receive a phone call from this patient
- *"Hey, Dr. Lee...you don't remember me probably but I had a stroke at age 33 and you helped me at Iowa"*
- Me: *"Sure, I remember you"*
- *"I was just calling to let you know that I went back to college, I got married, and now I have a new baby, his name is Andrew"*
- Me: *"That is so great, congratulations"*
- *"No, Dr. Lee you don't understand...his name is ANDREW!"*

143

ONE PERSON  
CAN MAKE A  
DIFFERENCE,  
AND EVERYONE  
SHOULD TRY

-JOHN F. KENNEDY-

144

## Thanks for your time and attention

- Andrew G. Lee, MD
- Chair Ophthalmology, **Houston Methodist Hospital**, Professor of Ophthalmology, Neurology, & Neurosurgery, Weill **Cornell** Medical College; Clinical Professor, **UTMB** Galveston; **UT MD Anderson Cancer Center**; Adjunct Professor, **Baylor** COM, U. **Iowa** & U. **Buffalo**, SUNY



Weill Cornell Medical College

