Ophthalmology Grand Rounds
31 March 2016
SUNY Downstate Medical Center

Benjamin Whigham, M.D.

Presentation

- 34 F “History of uveitis”
- 2012: peripheral vision “greyed-out” + temporal flashes of light “like a strobe light” (no other symptoms)
- Involved both eyes, progressed over 2 weeks
- Improved on prednisone
- “Blood tests negative”, visual field test “stable”, “nothing” on MRI
- Prednisone tapered, methotrexate started but later self-discontinued
- Uncomplicated pregnancy

Presentation

- Stable, chronic symptoms
  - Occasional flashes of light (worse with rain)
  - Occasional floaters (worse with rain)
  - Intermittent mild photosensitivity
- Same before/after stopping methotrexate
- Same during pregnancy

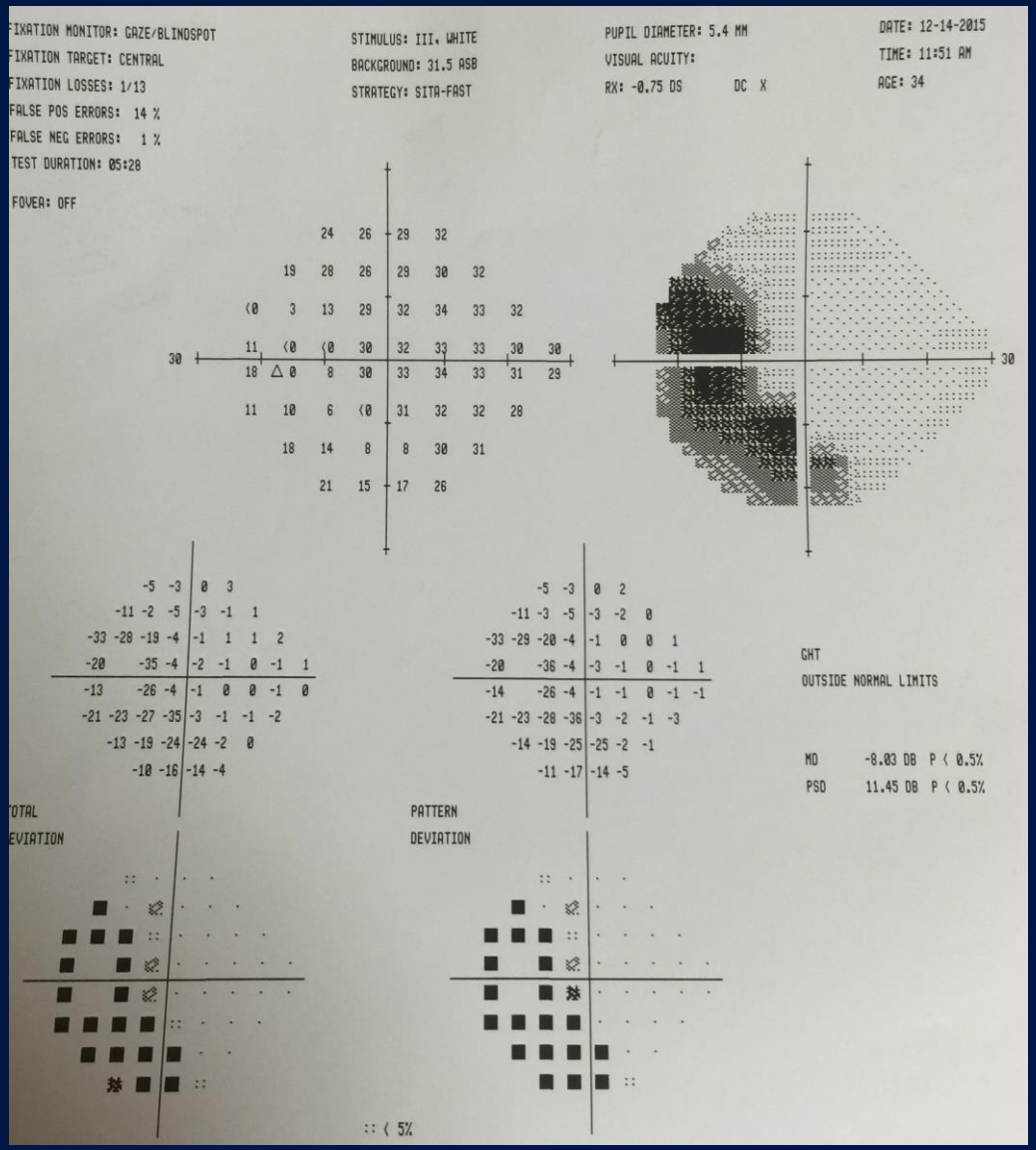
Additional History

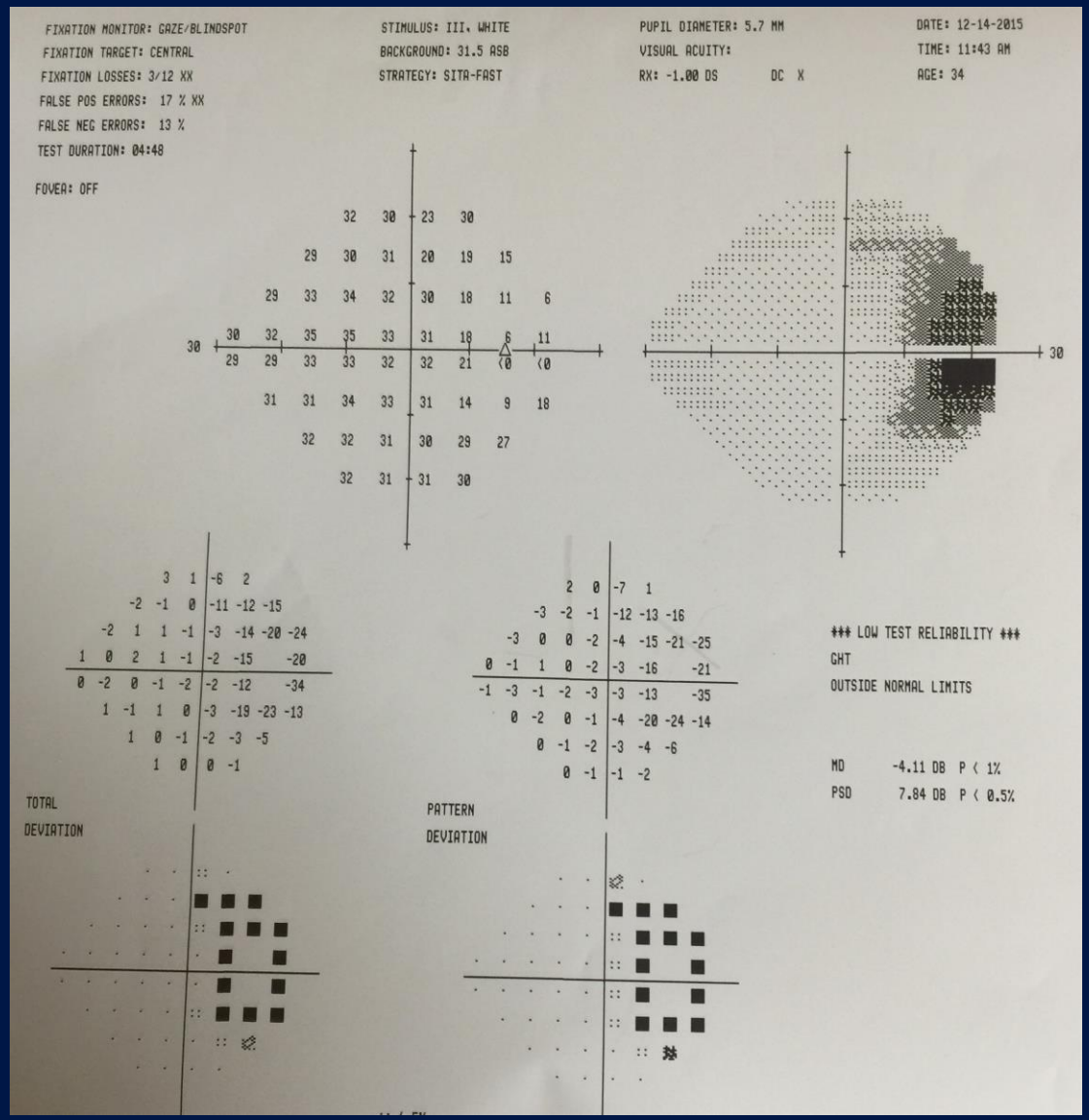
- ROS: Negative.
- POHx (pre 2012): Myopia (-4.00 OU)
- PMHx: Bilateral carpal tunnel syndrome, Migraines (rare), Sinus infections (3-4/year), HLD, Pregnancy x 2 (first c/b mild preeclampsia)
- PSHx: Appendectomy
- Birth Hx: Premature (3lb, 13oz)
- Allergies: None
- Meds: Multivatimin
- Family: Mother glaucoma (Diagnosed 5th decade); Half-sister with lupus
- Social: Social alcohol, works as clerk

Exam

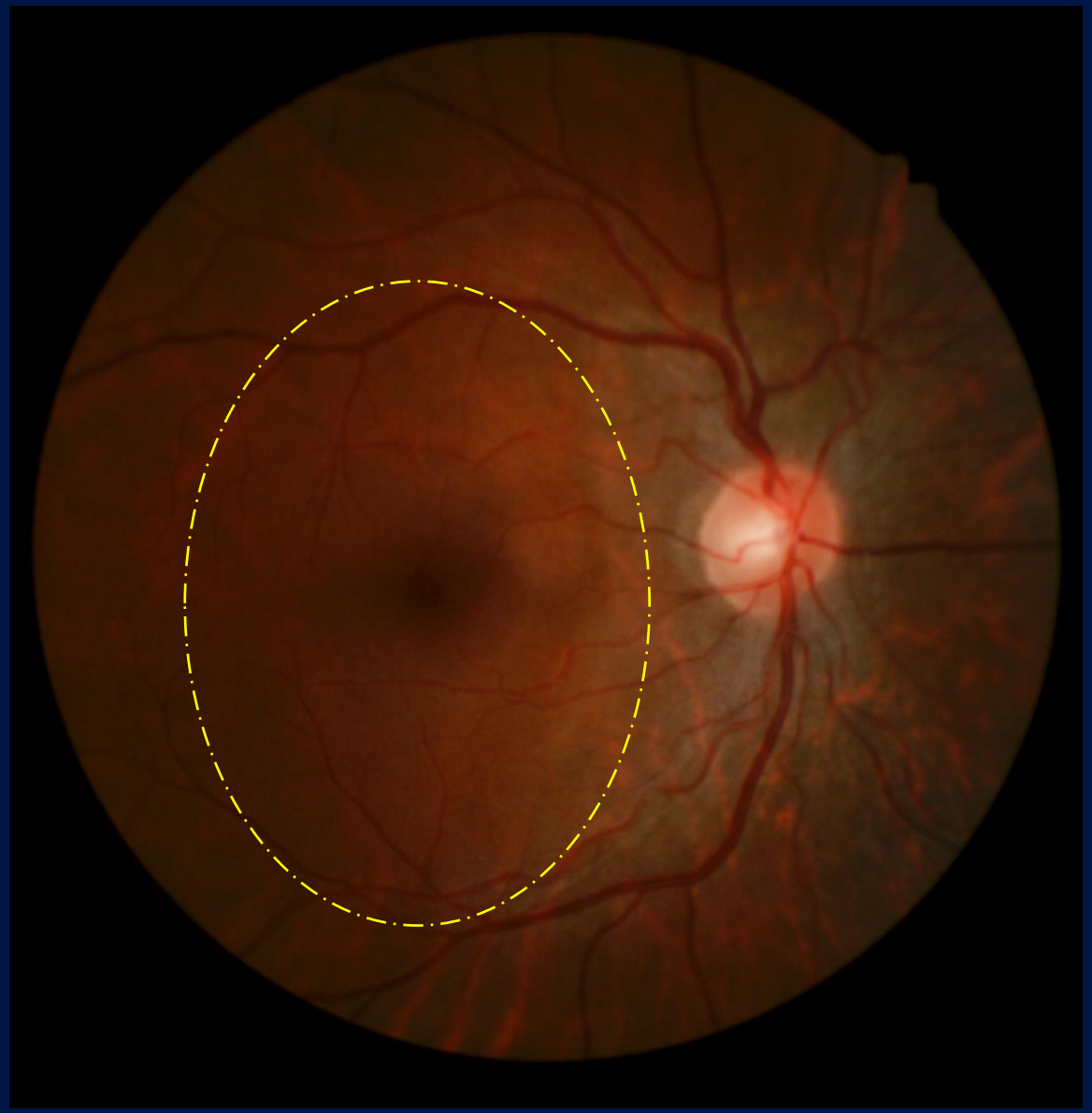
- DVAcc 20/20-2 (PHNI) OD, 20/20 OS
- Pupils 7-3 OU, no RAPD
- EOM full
- Color Plates: 12.5/14 OD, 13/14 OS
- Ta: 17, 18 @ 1145
- SLE: DTBUT OU

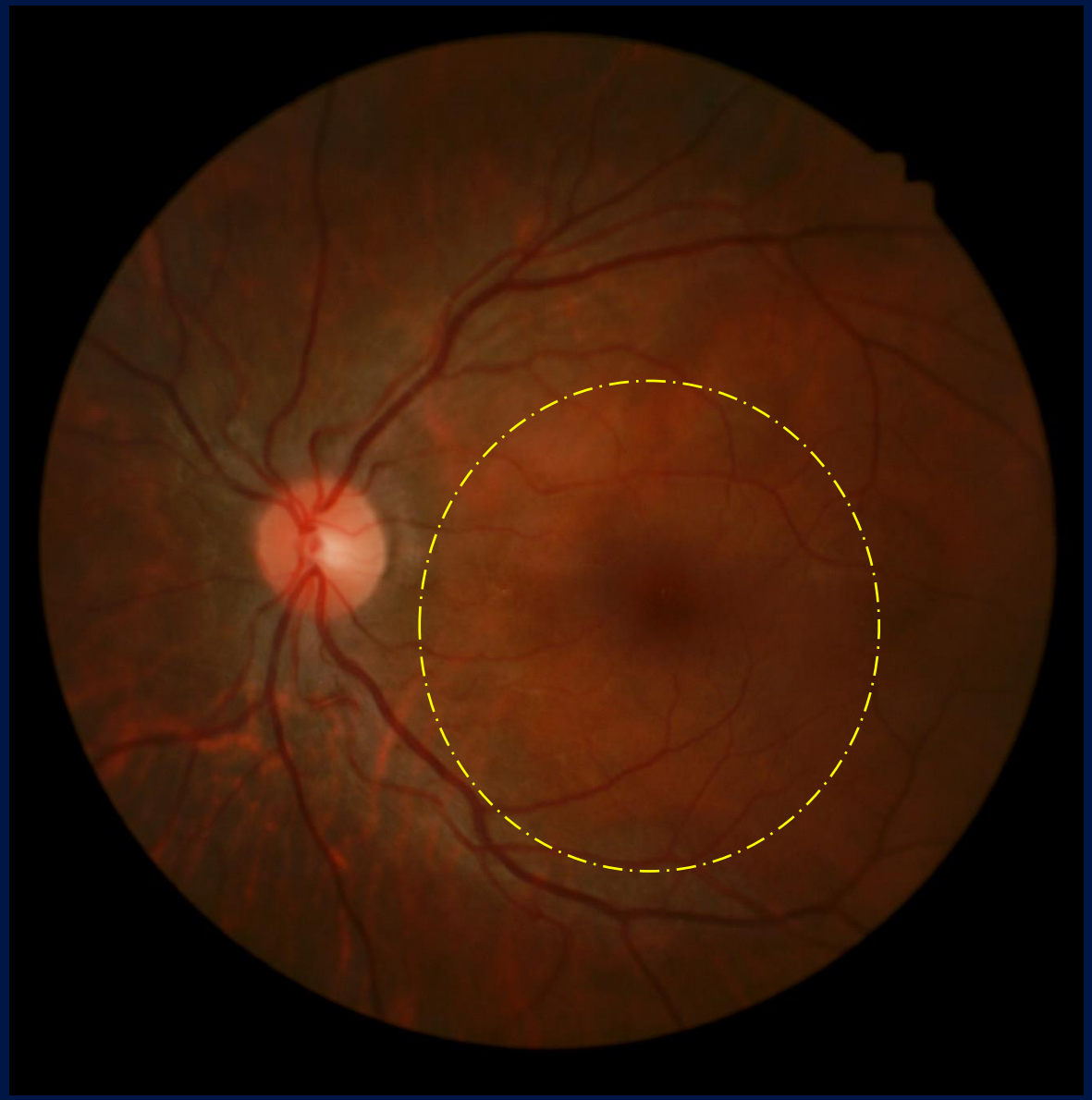
HVF 24-2


FIXATION MONITOR: GAZE/BLINDSPOT
FIXATION TARGET: CENTRAL
FIXATION LOSSES: 1/13
FALSE POS ERRORS: 14 %
FALSE NEG ERRORS: 1 %
TEST DURATION: 05:28
FOVEA: OFF
STIMULUS: III, WHITE
BACKGROUND: 31.5 ASB
STRATEGY: SITA-FAST
PUPIL DIAMETER: 5.4 MM
VISUAL ACUITY:
RX: -0.75 DS DC X
DATE: 12-14-2015
TIME: 11:51 AM
AGE: 34
GHT
OUTSIDE NORMAL LIMITS
MD -8.03 DB P < 0.5%
PSD 11.45 DB P < 0.5%
TOTAL DEVIATION
PATTERN DEVIATION
:: < 5%

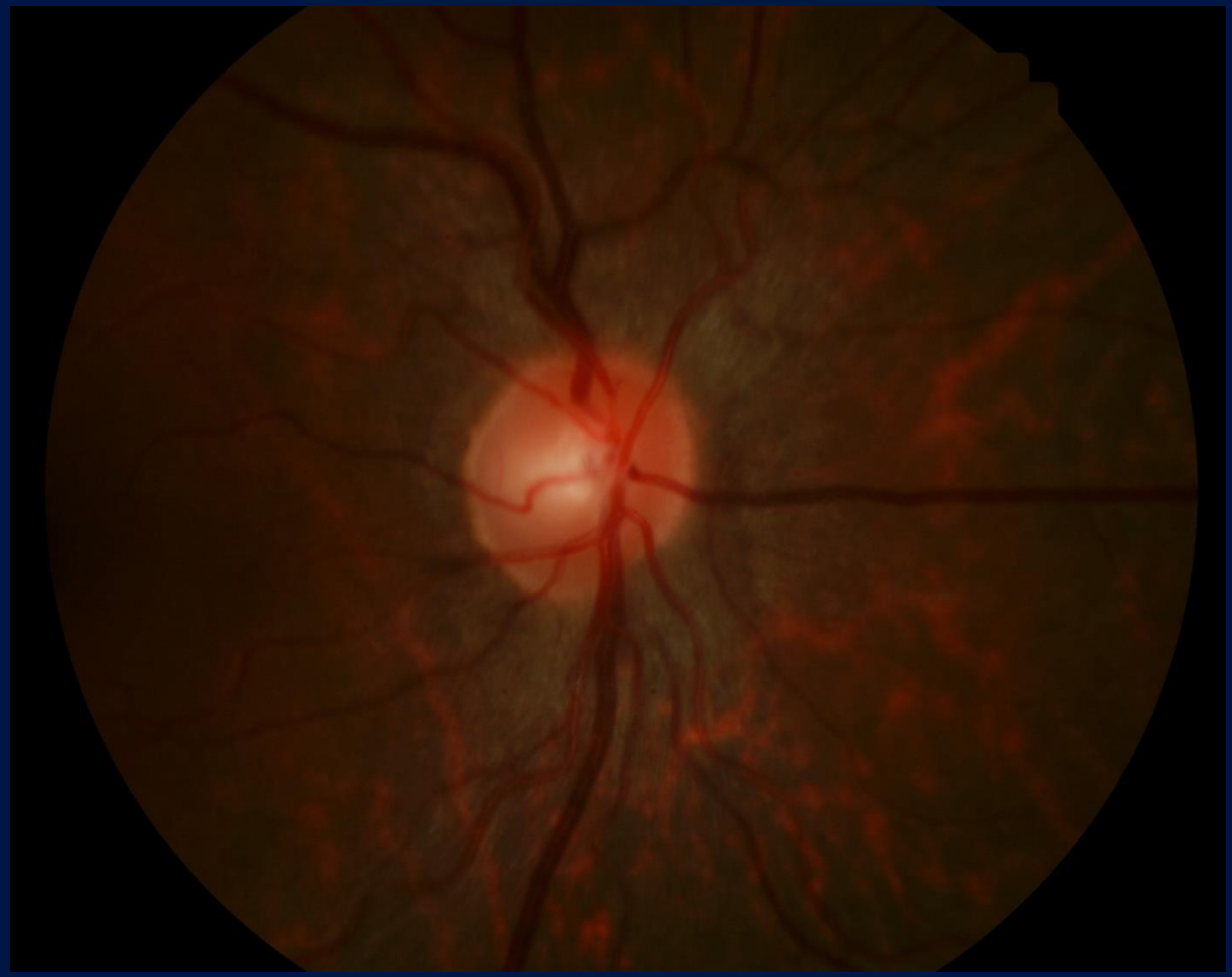

FIXATION MONITOR: GAZE/BLINDSPOT
FIXATION TARGET: CENTRAL
FIXATION LOSSES: 3/12 XX
FALSE POS ERRORS: 17 % XX
FALSE NEG ERRORS: 13 %
TEST DURATION: 04:48
FOVEA: OFF
STIMULUS: III, WHITE
BACKGROUND: 31.5 ASB
STRATEGY: SITA-FAST
PUPIL DIAMETER: 5.7 MM
VISUAL ACUITY:
RX: -1.00 DS DC X
DATE: 12-14-2015
TIME: 11:43 AM
AGE: 34
*** LOW TEST RELIABILITY ***
GHT
OUTSIDE NORMAL LIMITS
MD -4.11 DB P < 1%
PSD 7.84 DB P < 0.5%
TOTAL DEVIATION
PATTERN DEVIATION

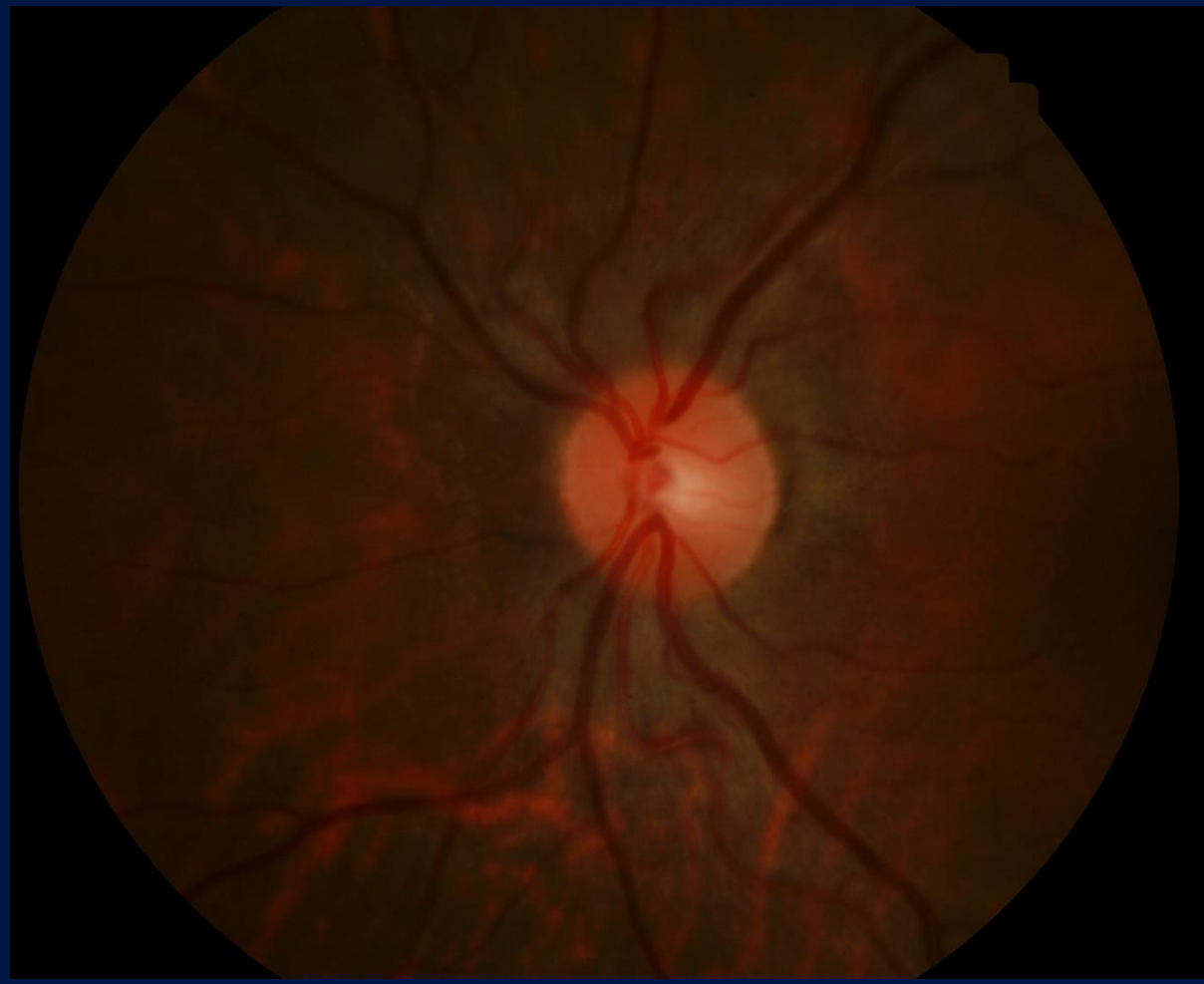
Fundus photos





Optic nerves





Differential?

- Inflammatory chorioretinopathies of unknown etiology (White Dot Syndromes)
  - Acute idiopathic blind spot enlargement
  - Multiple evanescent white dot syndrome
  - Acute zonal occult outer retinopathy
- Optic neuritis
- Pituitary mass lesion

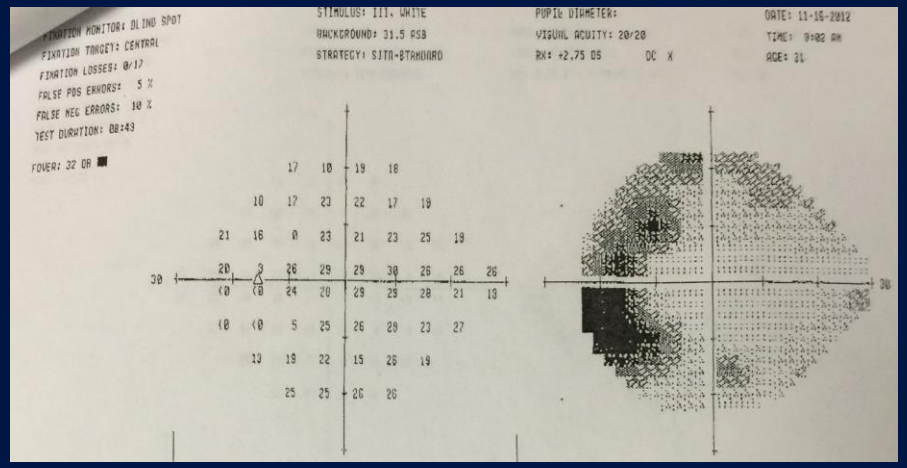
Obtain medical records

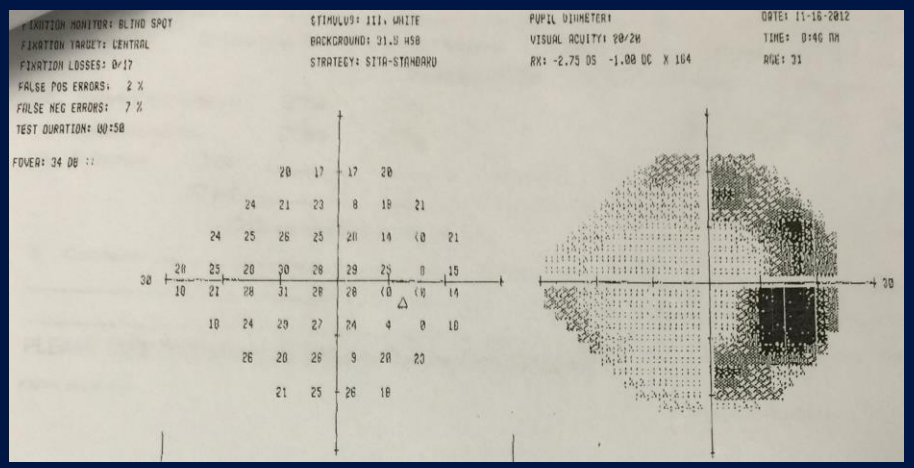
- 2012 Presentation
  - Floaters, grey blind spot x 2 weeks, temporal photopsias
  - 20/20 – 20/30 OU; Bilateral VF defects
  - Vitreous haze
  - FA: delayed filling, hyperintensities around vessels OS > OD, “vasculitis OU”
  - OCT: increased RNFL thickness
  - ESR 49 (high), RPR (-), ANA (-), MPO (-), ANCA (-), MHATP (-)
  - MRI brain normal

Medical records

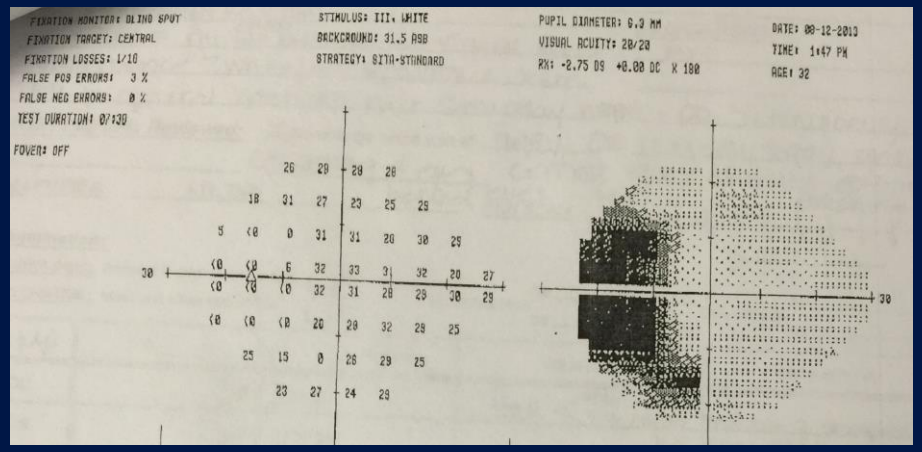
- 11/2012 Initial treatment: Prednisone 60mg
  - Photopsias improved, vision stable, 20/20 at 2 weeks
  - Taper slowly due to recurring photopsias
- 2/2013 Started Methotrexate + Weaned off Pred
  - Photopsias stable on MTX
  - Stable HVF 24-2 (temporal defects OU)
- 5/2014 Stopped MTX

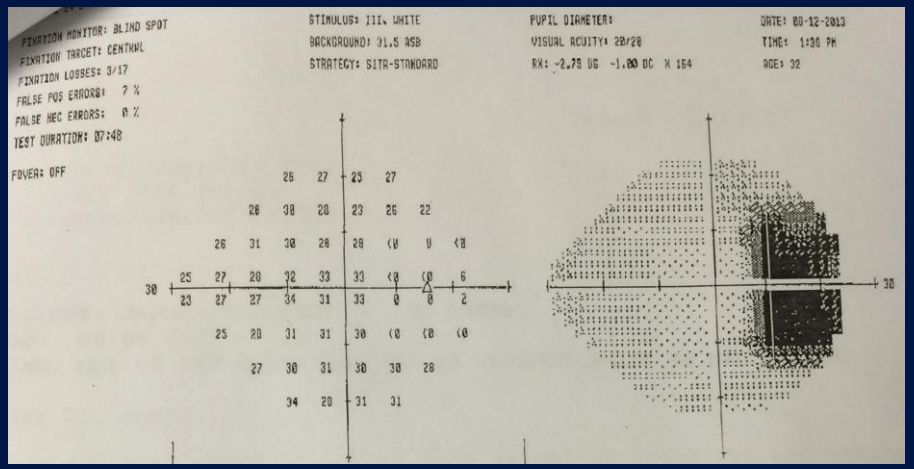
11/2012



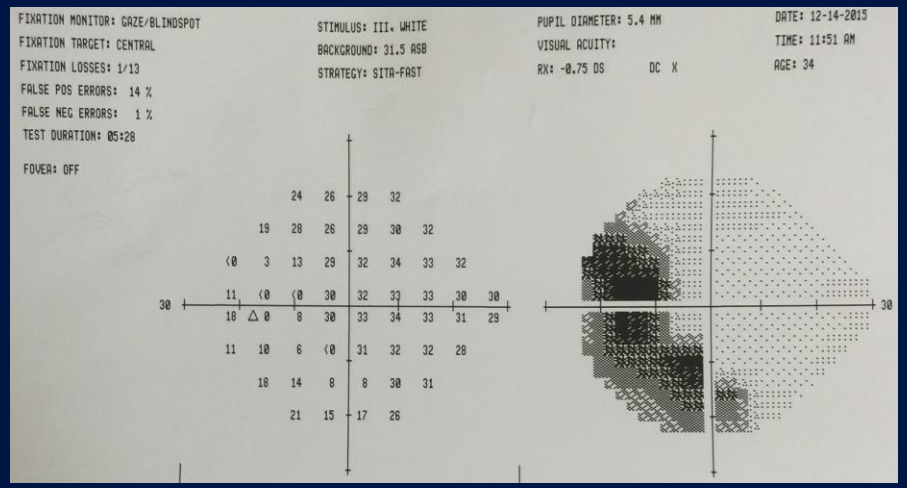


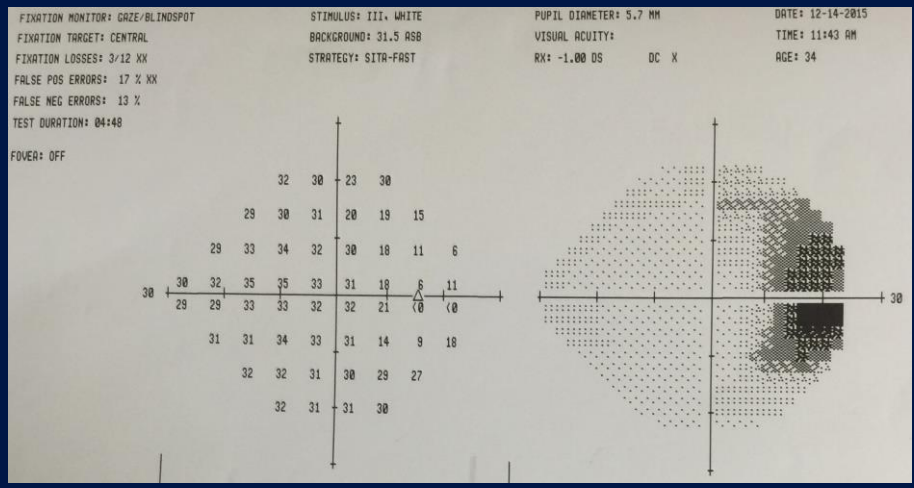
08/2013





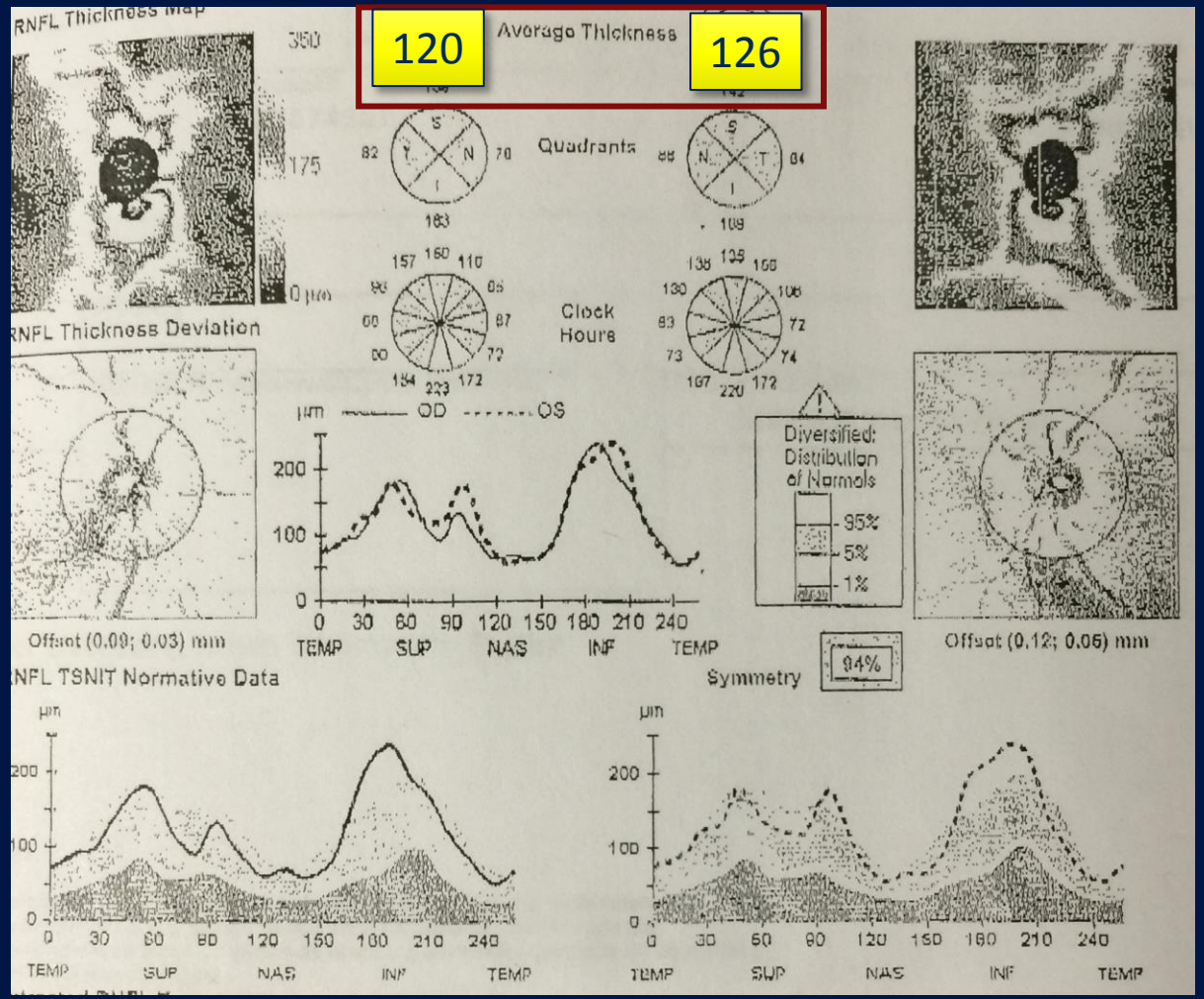
12/2015



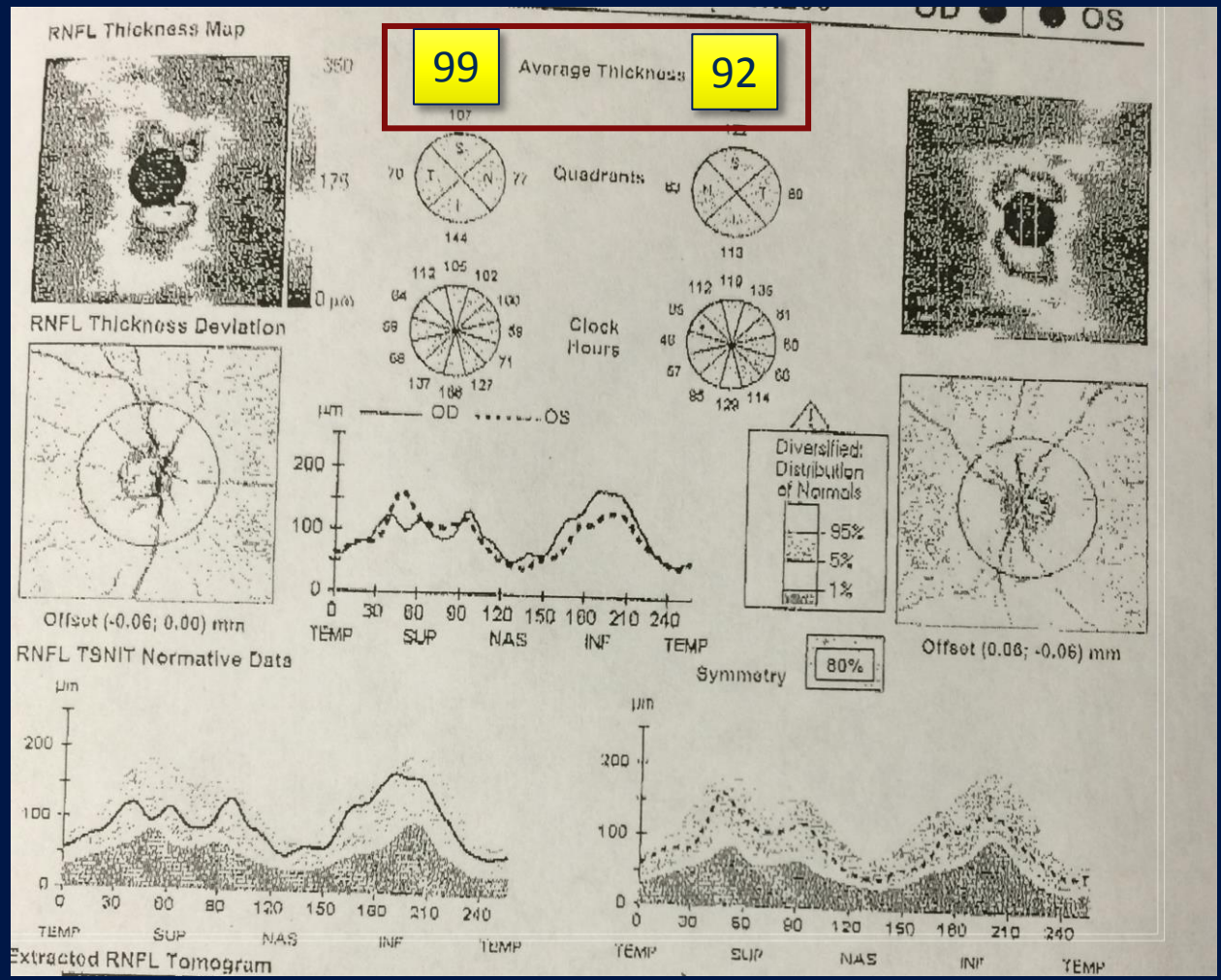


OCT

11/2012



05/2013



Differential

- Inflammatory chorioretinopathies of unknown etiology (White Dot Syndromes)
  - Acute idiopathic blind spot enlargement
  - ~~Multiple evanescent white dot syndrome~~ (FA negative, no white dots)
  - Acute zonal occult outer retinopathy

White dot syndromes

AZOOR Complex Diseases

1. Acute Zonal Occult Outer Retinopathy
   Acute Annular Outer Retinopathy
2. Multifocal Choroiditis and Panuveitis
   Punctate Inner Choroidopathy
3. Multiple Evanescent White Dot Syndrome
   Acute Idiopathic Blind-Spot Enlargement Syndrome

- Acute Posterior Multifocal Placoid Pigment Epitheliopathy
- Serpiginous Choroiditis
- Birdshot Retinopathy

Darrell Baskin, “White Dot Syndromes”, 2015.

Acute Idiopathic Blind Spot Enlargement Syndrome (AIBSE)

- "Spectrum of peripapillary retinal disease in women with abnormal parafoveal ERG results, some evidence of optic neuropathy, and limited recovery."
- 27 patients collected 1989 to 1997
- Female, Ages 19 – 53
- Predominately unilateral

Table 1. Initial Symptoms of 27 Patients With Acute Idiopathic Blind Spot Enlargement

Symptom	No. of Patients
Loss of vision	25
Blurring	9
Awareness of darkened area or missing vision	9
Spots in vision	3
"Looking through film"	4
Positive visual phenomena	23
Photopsia (sparkles, flashes, flickering)	16
Swirling	1
Movement within scotoma	3
Colored light	1
After "flash bulb"	2

(Volpe, Rizzo, & Lessell, 2001)

AIBSE Spectrum

Table 2. Measured Visual Dysfunction in 27 Patients With Acute Idiopathic Blind Spot Enlargement

Examination Finding	No. of Patients
Visual acuity	
20/20	16
20/25-20/50	10
20/200	1
Dyschromatopsia	9
Afferent pupil defect	8
Visual field defects (largest diameter of scotoma, degrees)	
<15	12
15-30	12
>30	3

Table 3. Opthalmoscopic Findings in 27 Patients With Acute Idiopathic Blind Spot Enlargement Syndrome*

Ophthalmoscopic Finding	No. of Patients
Optic nerve swelling, hyperemia, or staining on fluorescein angiography	12
Normal examination	8
Peripapillary RPE/choroidal abnormality	6
White dots	5
Peripapillary subretinal grayish discoloration	4
Peripheral RPE changes	4
Vitritis	3
Macular pigment mottling/granularity	2
Macular edema	1

** Ten patients had more than 1 finding. RPE indicates retinal pigment epithelium.*

ERG: Full field ~ normal (8 of 9 patients)
Nasal parafoveal ERG abnormal (8 of 9 pts)

Photopsias resolve; VF defects remain stable

(Volpe, Rizzo, & Lessell, 2001)

AIBSE

- Treatments: None

- Prognosis: Good
  - Only 6 of 27 patients with recurrence (2 in opposite eye)
  - Blind spot enlargement does not reverse (or progress)
  - Photopsias decrease in all patients

Acute Zonal Occult Outer Retinopathy (AZOOR)

- 131 reported cases / 205 eyes
- Average age 36.7 years; F:M of 3:1
- VA > 20/40 in 75%
- Fundus unremarkable in 75% + Mild vitritis possible
- Blind spot enlarged (± other VF deficits) in 75%
- ERG abnormalities ≥ 99%
- Occasional RPE disturbances develop
- Good prognosis; VF loss stops by 6 months; immunosuppression ineffective

(Monson & Smith, 2011)

AZOOR

- Second case series with 51 Patients (37 F : 14 M)
- 39% bilateral
- 88% Photopsias
- ≥ 20/40 in 76%
- 100% VF defects + ERG amplitude depression
- 48% affected eyes had RPE changes
- 16 of 51 patients had recurrences (total of 23 recurrences)
- Chronic photopsias

(Gass, Agarwal, & Scott, 2002)

AIBSE or AZOOR?

- AIBSE: "… resolution of photopsia with stable, persistent visual field defects" + unilateral

  AZOOR: "… chronic photopsia…progression of visual field loss during weeks or months…second eye involvement, and late RPE atrophy."

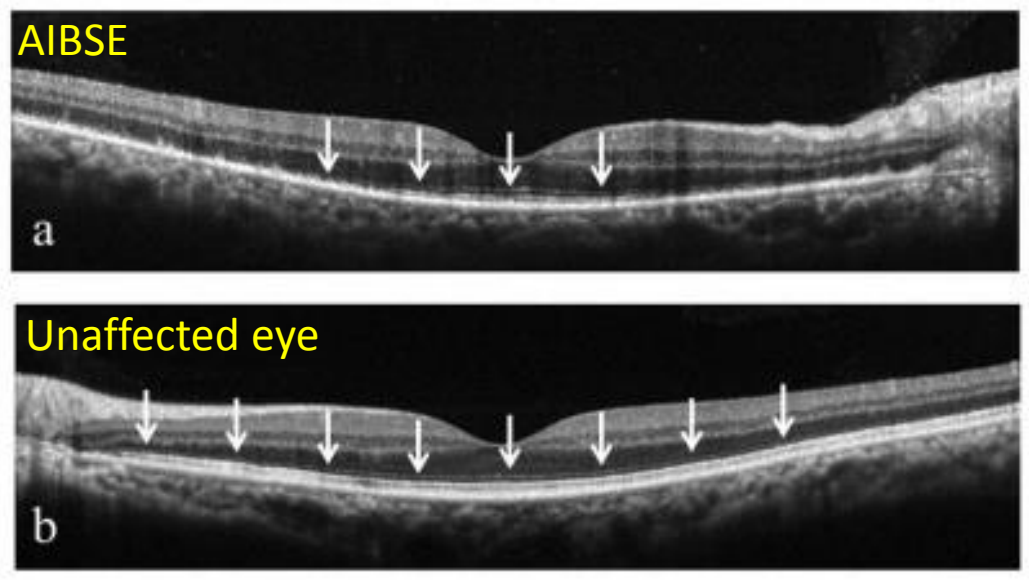
- Either way: No treatment indicated + similar rates of recurrences

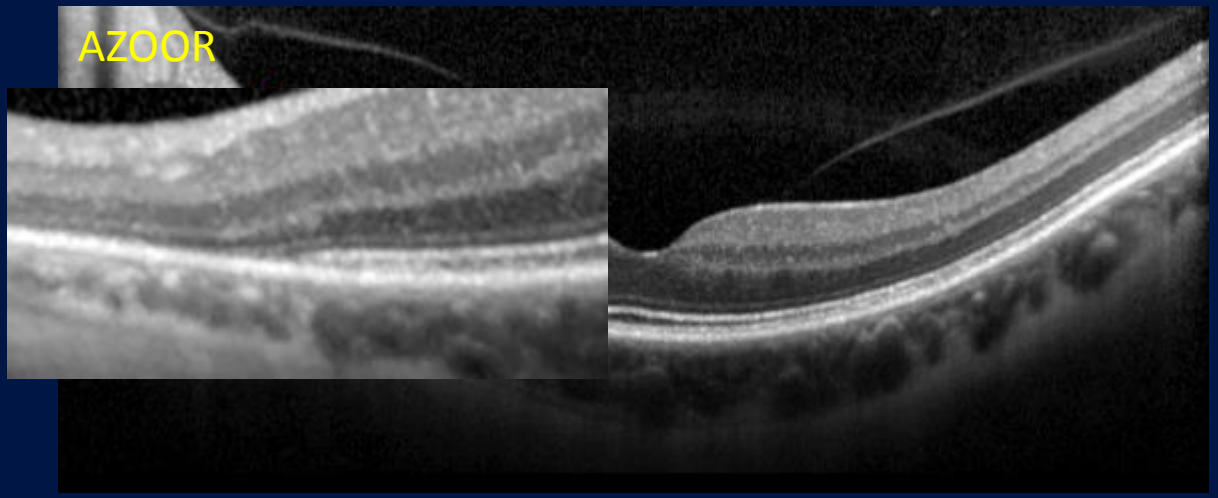
- MEWDS: Dots + Generally recover VF

(Volpe, Rizzo, & Lessell, 2001)

Theories on etiology/pathophysiology

- Both AIBSE and AZOOR are unknown
- AIBSE: Likely outer segment dysfunction
- AZOOR: “retinal pigment epithelium cell death with lipofuscin-laden cells at the border of the expanding lesion and associated atrophy of the underlying choriocapillaris”





(Spaide, 2004)
(Volpe, Rizzo, & Lessell, 2001)
http://westcoastretina.com/july-2012.html
http://ispub.com/IJOVS/11/1/14787

Summary

- AZ**O**OR: OCCULT (normal fundus at presentation)
- A**I**BSE: IDIOPATHIC (diagnosis of exclusion)
- Lots of overlap between inflammatory chorioretinopathies

References

- Gass, J. D., Agarwal, A., & Scott, I. U. (2002). Acute zonal occult outer retinopathy: a long-term follow-up study. *Am J Ophthalmol, 134*(3), 329-339. Retrieved from http://www.ncbi.nlm.nih.gov/pubmed/12208243
- Monson, D. M., & Smith, J. R. (2011). Acute zonal occult outer retinopathy. *Surv Ophthalmol, 56*(1), 23-35. doi:10.1016/j.survophthal.2010.07.004
- Newcomb, R. D. (2000). Acute, idiopathic blind spot enlargement syndrome. *Optom Vis Sci, 77*(4), 178-187. Retrieved from http://www.ncbi.nlm.nih.gov/pubmed/10795800
- Spaide, R. F. (2004). Collateral damage in acute zonal occult outer retinopathy. *Am J Ophthalmol, 138*(5), 887-889. doi:10.1016/j.ajo.2004.06.001
- Volpe, N. J., Rizzo, J. F., 3rd, & Lessell, S. (2001). Acute idiopathic blind spot enlargement syndrome: a review of 27 new cases. *Arch Ophthalmol, 119*(1), 59-63. Retrieved from http://www.ncbi.nlm.nih.gov/pubmed/11146727

Thank you

- Dr. Valerie Elmalem
- Dr. Joseph Tseng