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Seeing Stars: A review of the white dot syndromes

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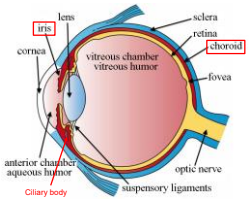
Objectives

At the end of this talk, audience members should be able to...

1. Appreciate phenotypic differences among the white dot syndromes
2. Possess a cursory understanding of treatment approaches for the white dot syndromes
3. Understand the importance of distinguishing white dot syndromes from infectious masquerades

Introduction

- **White dot syndromes** = a heterogenous collection of posterior uveitides characterized by **white spots and dots** in the retina/choroid
- **Posterior uveitis** involves inflammation in the retina and/or choroid and is generally much more vision-threatening than anterior or intermediate uveitis
- Approximately 30 000 cases of posterior uveitis per year in the US



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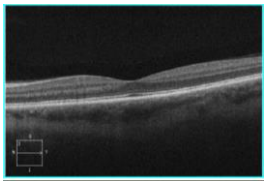
OVERVIEW OF IMAGING MODALITIES

Optical coherence tomography (OCT)

Cross-sectional, anatomic/structural view of retina and choroid

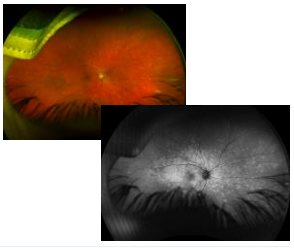
- Uses near-IR light to measure reflectance from tissue layers
- Provides high-resolution intraretinal detail and some choroidal detail
- Limited utility for vascular imaging
- Limited field of view (central macula or occasionally mid-periphery)



Fundus photography and autofluorescence (FAF)

Wide-angle two-dimensional views of posterior segment structures

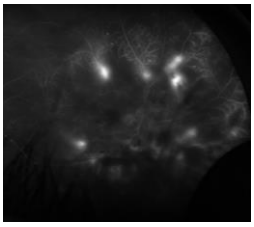
- Optos photos are not true-color (lack a blue channel so image is biased towards red/green)
- Autofluorescence detects RPE, metabolic activity, lipofuscin
- Autofluorescence can highlight lesions that are not readily apparent on photos



Fluorescein angiography (FA)

Great for imaging retina and vasculature, provides less information on choroid

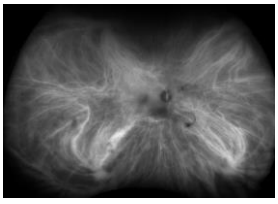
- Light-sensitive, light-emitting dye that illuminates the path of blood flow through the retinal vessels
- Dark hypofluorescent findings
  - Blockage from vitreous hemorrhage
  - Areas of ischemia and capillary non-perfusion
- Bright hyperfluorescent findings
  - Staining of scars and lesions
  - Leakage in areas of vasculitis, retinal edema, inflammation



Indocyanine green angiography (ICGA)

Great for choroidal vascular imaging

- ICG binds to proteins and is less "leaky" than fluorescein so better for imaging choroidal vessels
- Excitation/emission wavelengths are different from FA and result in less interference from the overlying RPE
- Can show lesions in the choroid as areas of hypocyanescence
- Few side effects than fluorescein



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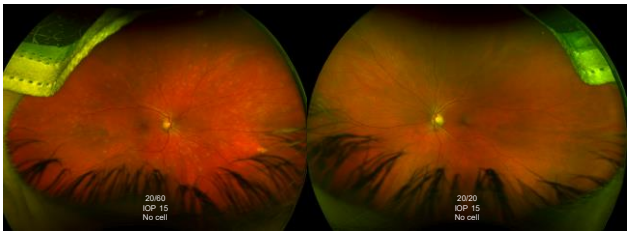
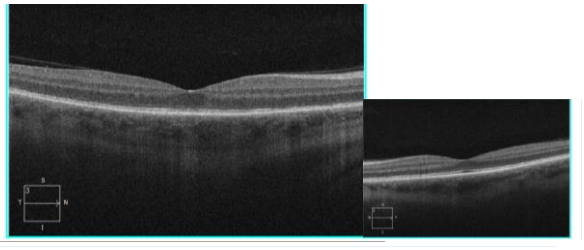
THE WHITE DOT SYNDROMES

Case 1

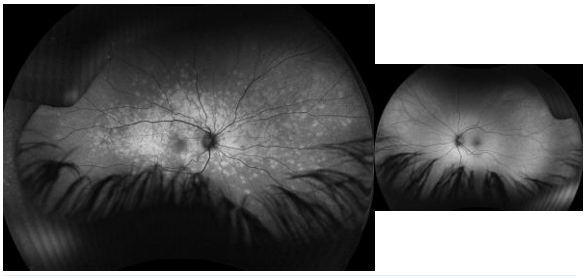
41F w/ new onset "speckles of light" in right eye only



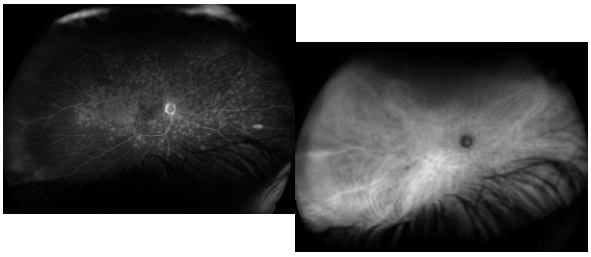
Case 1 OCT



Case 1 FAF



Case 1 FA and ICGA

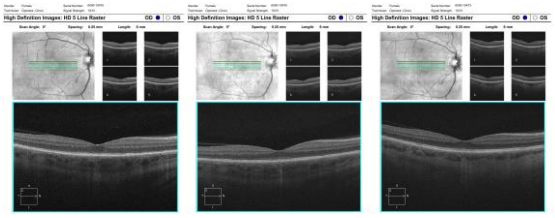


Multiple evanescent white dot syndrome (MEWDS)

- Acute onset, multiple small gray-white dots in deep retina and RPE
- 80% of cases are unilateral and there is a F > M predominance
- Foveal granularity on exam and loss of EZ integrity are characteristic
- Most cases are self-limited and vision recovers well
- Steroids can hasten recovery

- From the neurology literature: AKA idiopathic enlargement of blind spot syndrome

Case 1 Follow-up



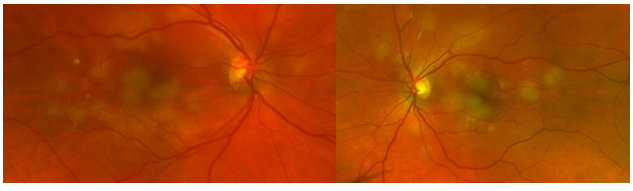
Presentation
VA 20/60

2 weeks
VA 20/40

6 weeks
VA 20/20

Case 2

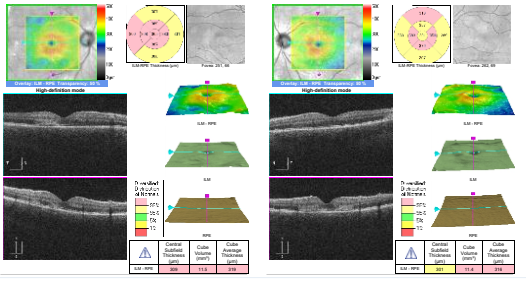
24M w/ headaches and new vision changes OU



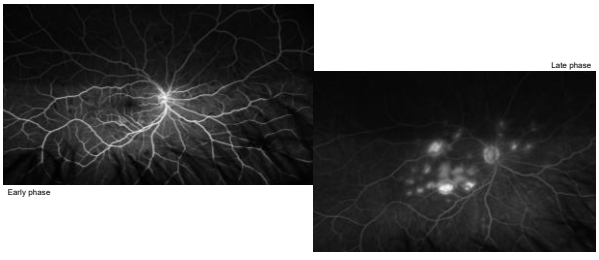
20/30
IOP 9

20/25
IOP 9

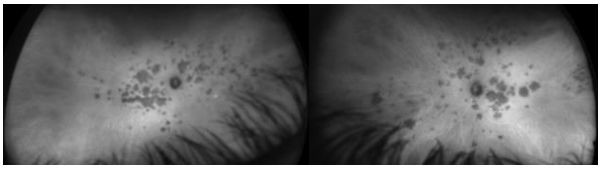
Case 2 OCT



Case 2 FA



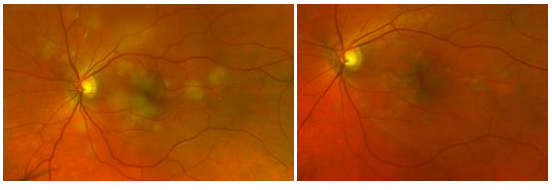
Case 2 ICGA



Acute posterior multifocal placoid pigment epitheliopathy (APMPPE)

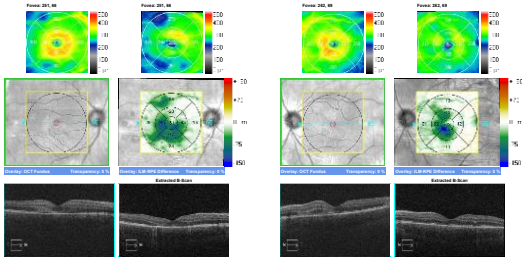
- Sequential, bilateral onset in young patients (2nd – 3rd decade)
- Male predominance
- Characteristic lesions on FA; "blocks early, hyperfluoresces late"
  - However this is not specific
- Treatment is steroids or observation
- DDx to consider: serpiginous, "ampiginous" (relentless APMPPE)
- Watch for neurologic signs (headaches)
  - Can develop strokes in very rare cases within days to weeks of eye diagnosis
  - Most commonly occur as steroids are being tapered or IMT is being changed

Case 2 Follow-up (2 weeks later)



20/30 20/25

Case 2 Follow-up (2 weeks later)



Case 3

67F with new onset flashes and floaters OU

Visual Acuity (Snellen - Linear)

	Right	Left
Dist cc	20/20 -2	20/30 +-2
Dist ph cc		20/30 +2

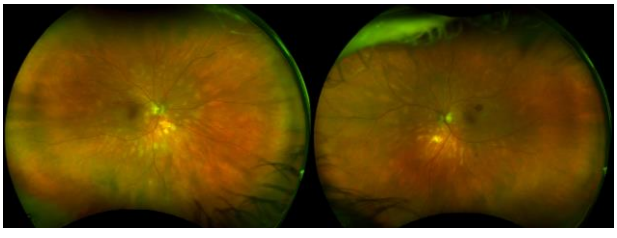
Tonometry (Tonopen, 1:57 PM)

	Right	Left
Pressure	18	12

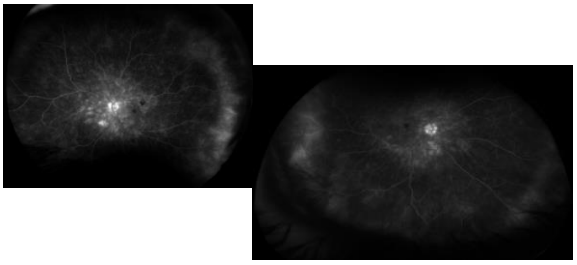
Slit Lamp Exam

	Right	Left
Lids/Lashes	Normal lids, lashes, lacrimal glands, and lacrimal drainage	Normal lids, lashes, lacrimal glands, and lacrimal drainage
Conjunctiva/Sclera	White and quiet	White and quiet
Cornea	Normal epithelium, stroma, endothelium, and tear film	Normal epithelium, stroma, endothelium, and tear film
Anterior Chamber	Deep and quiet	Deep and quiet
Iris	dilated	dilated
Lens	1+ NS, 2+ CC	1+ NS, 2+ CC
Vitreous	PVD, 1+ ant vit cell	PVD, trace ant vit cell

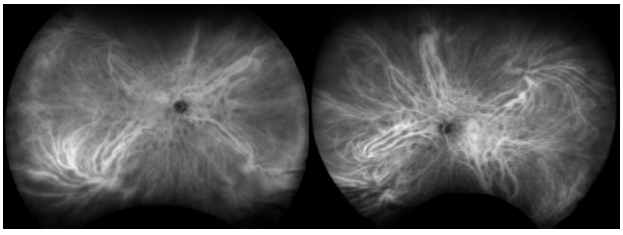
Case 3 Photos



Case 3 FA



Case 3 ICGA



Birdshot retinochoroidopathy

- Characteristic **deep, ovoid, creamy lesions** best seen on **ICGA**
- Chronic, **bilateral,** and often recurrent if untreated
- Not generally associated with systemic disease (ocular inflammation only)
- **Female >** male predominance
- Average age at diagnosis in **5 – 6th decade of life**
- **Photopsias, nyctalopia, color vision loss**
- Inflammation (vitritis, disc leakage, vasculitis) → **immunosuppression**

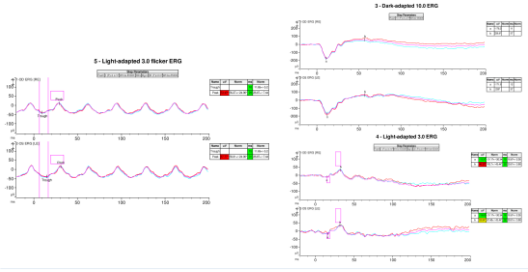
Birdshot retinochoroidopathy

- HLA-A29 is highly sensitive but not specific for birdshot
  - About 8 – 10% of the population is HLA-A29 (+)
  - Only 1 in 10 000 HLA-A29 (+) patients have birdshot
- Also has a retinal degeneration-like component that is often chronic and progressive → follow with exam, imaging, HVF, ERG

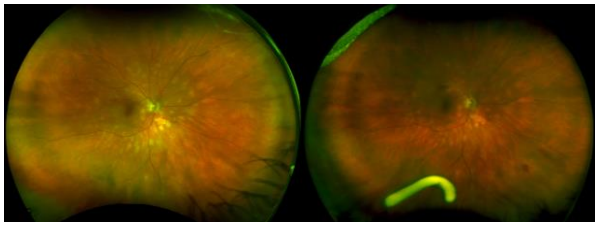
HLA-A29 testing in birdshot retinochoroidopathy

If you are HLA-A29 positive, does this mean you have birdshot?

Case 3 ERG



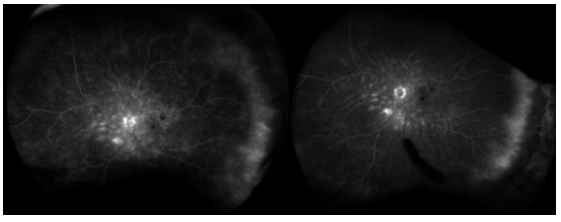
Case 3 Follow-up



Presentation
20/20

Month 2
20/20

Case 3 ERG

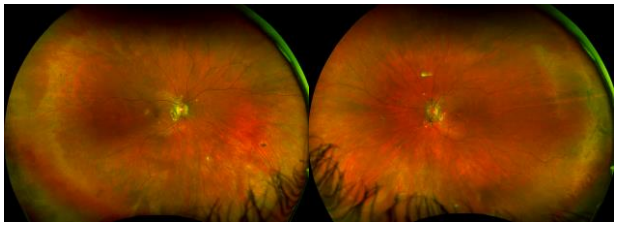


Presentation
20/30

Month 2
20/20

Case 4

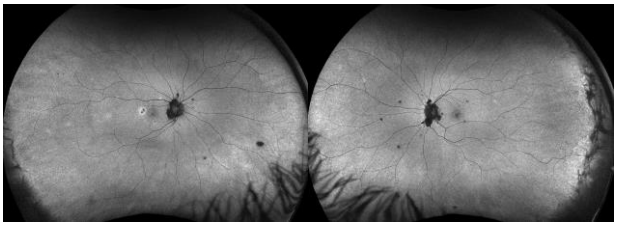
32F with h/o uveitis presents with increased photopsias OD



20/20
IOP 15

20/20
IOP 16

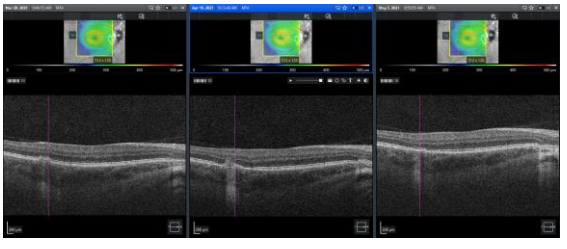
Case 4 FAF



Punctate inner choroidopathy (PIC)

- **Young, myopic women in 3rd – 4th decade of life**
- Characteristic photopsias
- **Acute/recurrent inflammatory lesions (but <u>no vitritis</u>)**
  - PO steroids, intravitreal steroids, IMT, implants (Yutiq, Retisert)
- **CNVM sequelae**
  - Anti-VEGF

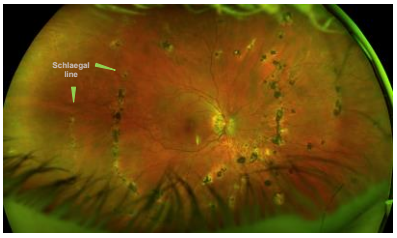
Case 4 Follow-up



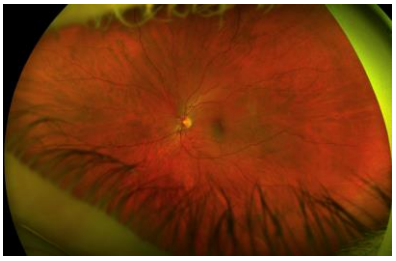
Case 5

43M with blurry VA, floaters, and photopsias OD

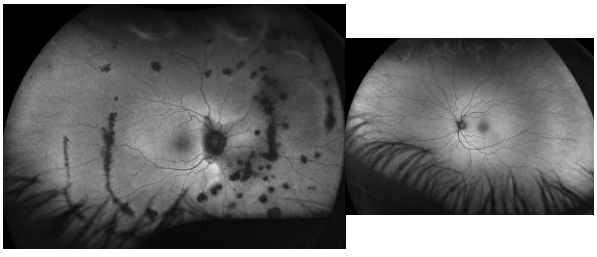
20/30
IOP 20
Pigment vit cell and debris



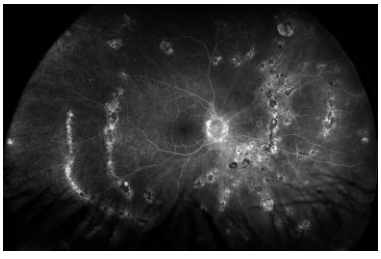
Case 5 Photo of normal left eye



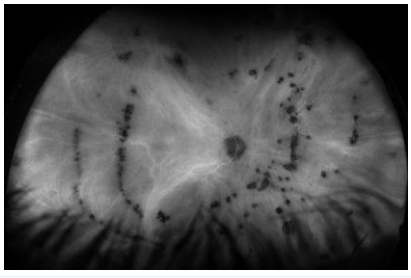
Case 5 FAF



Case 5 FA



Case 5 ICGA

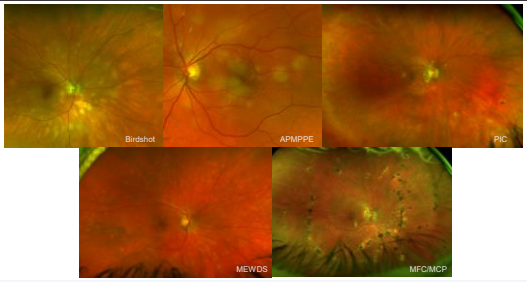


Multifocal choroiditis with panuveitis (MCP)

- Creamy active choroidal lesions that progress to pigmented and punched out chorioretinal scars
- Lesions may be slightly **bigger**, more **numerous**, and more **confluent** than those seen in PIC
- Can have anterior and/or vitreous inflammation (MCP) but may not
- Like PIC, can have macular CNVM complications that cause VA loss

- Schlaegal line- equatorial curvilinear streaks of CR lesions

Pattern recognition is important...

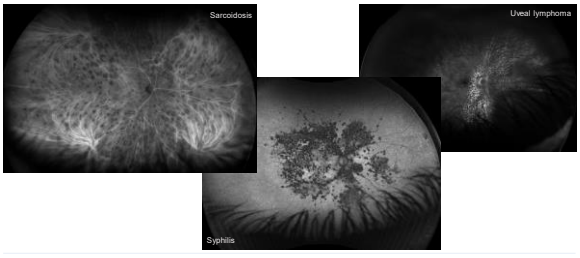


...but watch for masquerades!

SYPHILIS

Systemic disease can mimic white dot syndromes

...and should be identified and treated



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TREATMENT MODALITIES

Acute phase treatments

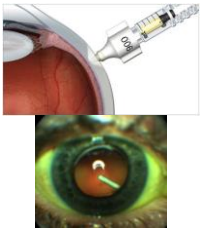
Initial treatments to rapidly control disease at onset

- Oral prednisone
  - High dose 1 mg/kg up to 60mg daily
  - Taper weekly
  - Extensive long-term side effects affecting nearly every organ system
  - Safe in pregnancy
  - Generally highly effective at controlling inflammation
- Topical steroid drops are less effective as disease is mostly posterior

Acute phase treatments

Intravitreal injection options

- Ozurdex (dexamethasone implant)
- Xipere (triamcinolone suprachoroidal)
- Anti-VEGF intravitreal injections
  - Only for CNVM in PIC or MCP



Long-term treatments

Systemic immunosuppression and long-term steroid options

- Iluvien (fluocinolone implant)
  - Lasts for 3 years
- Retisert (surgical implant)
  - Lasts for 5+ years
  - ...but being discontinued
- Systemic immunosuppression
  - Methotrexate (once a week pills)
  - Mycophenolate (daily pills)
  - Adalimumab (biweekly injections)
  - Infliximab, tocilizumab (monthly or bimonthly infusion)

Treatments for pregnant patients

Many white dot syndromes affect women of child-bearing age

- Many but not all treatment options are safe in pregnancy
- Oral and injection **steroids** are generally always safe
- **Adalimumab** and **infliximab** and **azathioprine** (less used) also safe to use

- Anti-VEGF injections generally avoided in pregnancy
- Methotrexate and mycophenolate contraindicated in pregnancy

Conclusions and summary

- The various white dot syndromes differ in the size and distribution of lesions, patient demographics, and prognosis/outcomes
- Multimodal imaging (OCT, FAF, FA, ICGA) is essential in fully characterizing the different presentations of the white dot syndromes
- Oral steroids are a mainstay of treatment; chronic conditions such as birdshot may require long-term immunosuppression; PIC and MCP can cause secondary CNVM which may require anti-VEGF injections
- Patients w/ white dot syndromes require a full uveitis work-up to rule out masquerade conditions which may require a different treatment approach

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Thank you for your attention!

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