# What do you need to know about posterior uveitis

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# Disclosures

- Off label treatments
- Paid advisory board Bayer
- Paid research support Allergan (makers of Ozurdex)
- Paid research support B and L (makers of Retisert)
- Paid research support Novartis

My aim: to give you an easy diagnostic approach to posterior uveitis

# Your aims of assessment

- Make a descriptive anatomical diagnosis
- Recognise specific named uveitis entities
- Diagnose significant underlying systemic inflammatory/infective disease
- Diagnose  intra-ocular/systemic infections/masquerade syndromes
- Diagnose the cause of visual loss

# Diagnostic possibilities in intermediate or posterior uveitis

- Isolated ocular disease
  - Idiopathic
  - Named
  - Infectious
- Ocular disease as part of systemic disease
  - Non-infectious
  - Infectious
  - Malignant
- Masquerade syndromes

- Attempt to make an anatomic and descriptive diagnosis
- Use recognised nomenclature

# PERSPECTIVES

## Standardization of Uveitis Nomenclature for Reporting Clinical Data. Results of the First International Workshop

THE STANDARDIZATION OF UVEITIS NOMENCLATURE (SUN) WORKING GROUP

• PURPOSE: To begin a process of standardizing the methods for reporting clinical data in the field of uveitis.
• DESIGN: Consensus workshop.
• METHODS: Members of an international working group were surveyed about diagnostic terminology, inflammation grading schema, and outcome measures, and the results used to develop a series of proposals to better standardize the use of these entities. Small groups employed nominal group techniques to achieve consensus on several of these issues.
• RESULTS: The group affirmed that an anatomic classification of uveitis should be used as a framework for subsequent work on diagnostic criteria for specific uveitic syndromes and that the classification of uveitis entities should be on the basis of the location of the inflammation and not on the presence of structural complications. Issues regarding the use of the terms "intermediate uveitis," "pars planitis," "pauvuveitis," and descriptors of the onset and course of the uveitis were addressed. The following were adopted: standardized grading schema for anterior chamber cells, anterior chamber flare, and for vitreous haze; standardized methods of recording structural complications of uveitis; standardized definitions of outcomes, including "inactive" inflammation, "improvement" and "worsening" of the inflammation, and "corticosteroid sparing," and standardized guidelines for reporting visual acuity outcomes.
• CONCLUSIONS: A process of standardizing the approach to reporting clinical data in uveitis research has begun, and several format have been standardized. (Am J Ophthalmol 2005;140:509–516. © 2005 by Elsevier Inc. All rights reserved.)

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A listing of members of the Standardization of Uveitis Nomenclature (SUN) Working Group appears in the Appendix.
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THE FIELD OF UVEITIS DEALS WITH MULTIPLE DISEASE entities, some of which are caused directly by infectious agents, several culture of which appear to be immune-mediated. Many uveitic entities are associated with systemic immune-mediated diseases, such as sarcoidosis, the HLA-B27–associated spondyloarthropathies and Behçet's disease, whereas others are limited to the eye. Although attempts have been made to standardize some aspects of uveitis,[1] in general there is limited standardization of classification criteria, inflammation grading schema, and outcomes.[2] Standardization would enhance greatly the comparability of clinical research from different centers, permit meta-analysis, and assist in the development of a more complete and meaningful picture of the clinical course of these diseases and their response to treatment.

The American College of Rheumatology (ACR) has developed classification criteria for many of the rheumatic diseases such as rheumatoid arthritis and systemic lupus erythematosus.[3,4] These criteria have been developed through a standard process and validated against large databases, in an effort to maximize sensitivity and specificity. In the field of uveitis, provisional criteria have been developed for a limited number of disorders (acute retinal necrosis, progressive outer retinal necrosis, Vogt-Koyanagi-Harada disease, and tubulointerstitial nephritis with uveitis),[5–9] and they still await validation. Additionally, there are criteria for the systemic portion of three diseases in which uveitis is an important feature (ankylosing spondylitis, juvenile idiopathic arthritis, and Behçet's disease),[9–11] but not for the uveitis in these diseases. Like the ACR classifications of arthritis and vasculitis, which are on the basis of the anatomic patterns of the disease, the most widely used classification of uveitis is the one devised by

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doi:10.1016/j.ajo.2005.03.057

# Anatomical classification of uveitis

- Anterior uveitis
- Intermediate uveitis
- Posterior uveitis
- Panuveitis

# Anatomical classification of uveitis

- Anterior uveitis
  - Inflammation limited mainly to the anterior segment inc
    - Iritis
    - Anterior cyclitis
    - Iridocyclitis
  - Can be acute or chronic

# Anatomical classification of uveitis

- Intermediate uveitis
  - Inflammation limited mainly to the vitreous but may include anterior retina, retinal vasculitis or macular oedema inc
    - Pars planitis

# Anatomical classification of uveitis

- Posterior uveitis
  - Inflammation limited mainly to the retina, choroid or optic nerve inc
    - Focal, diffuse or multifocal choroiditis
    - Chorioretinitis
    - Neuroretinitis
    - Serous detachment

# Anatomical classification of uveitis

- Panuveitis
  - Inflammation involving the whole eye without preference for anterior or posterior segment

# Different clinical pictures in intermediate/posterior/panuveitis

- Diffuse uveitis/vitritis
- Unifocal chorioretinitis
- Multifocal chorioretinitis
- Confluent chorioretinitis
- Retinal vasculitis
- Optic papillitis
- Serous retinal detachment

# Different clinical pictures in intermediate/posterior/panuveitis

- Diffuse uveitis/vitritis
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- Retinal vasculitis
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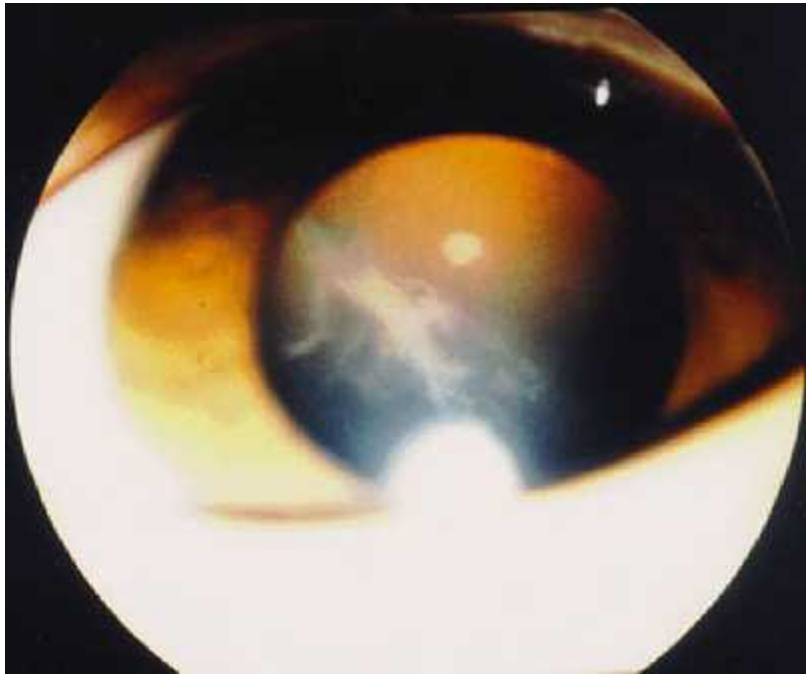
- Serous retinal detachment

- Isolated ocular disease
  - Idiopathic
  - Named
  - Infectious
- Ocular disease as part of systemic disease
  - Non-infectious
  - Infectious
  - Malignant
- Masquerade syndromes

# Non specific/diffuse uveitis

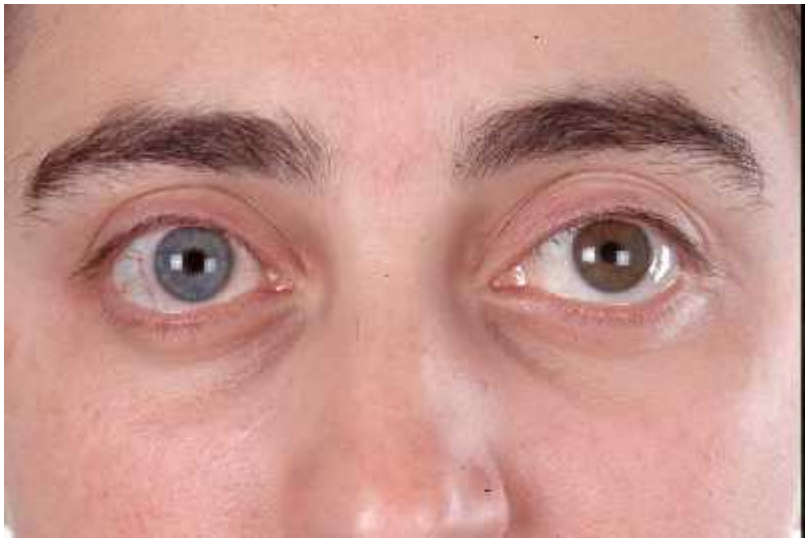
- Vitritis
- +/- macular oedema
- +/- retinal vasculitis
- No focal signs



# Intermediate uveitis without localizing features

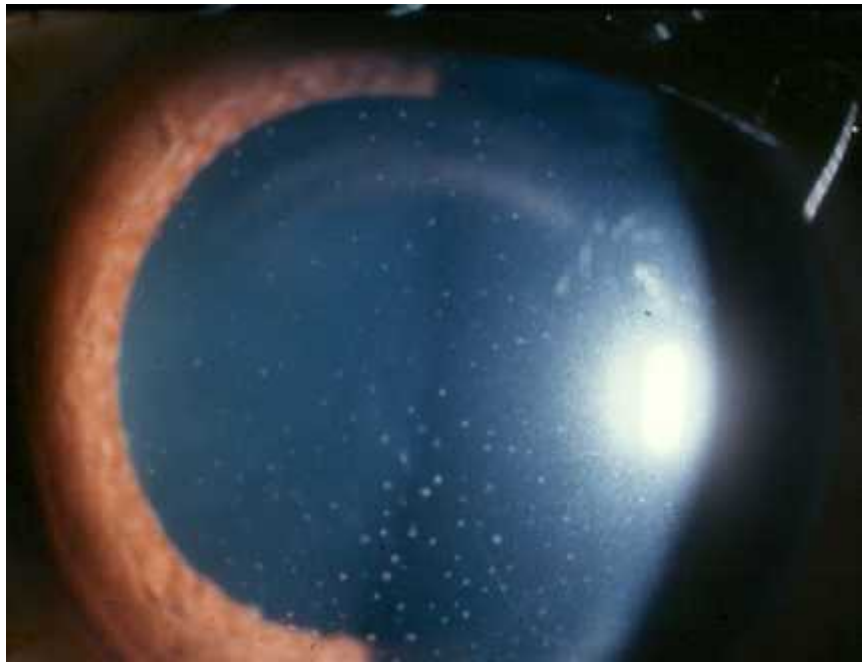
- Ocular diagnoses
  - Intermediate uveitis
  - Sympathetic ophthalmia
  - Fuch's heterochromic iridocyclitis
- Systemic diagnoses
  - Often idiopathic!
  - Sarcoidosis
  - Behcet's
  - MS
  - Oculo-cerebral lymphoma
  - Syphilis
  - TINU

# Fuch's heterochromic iridocyclitis

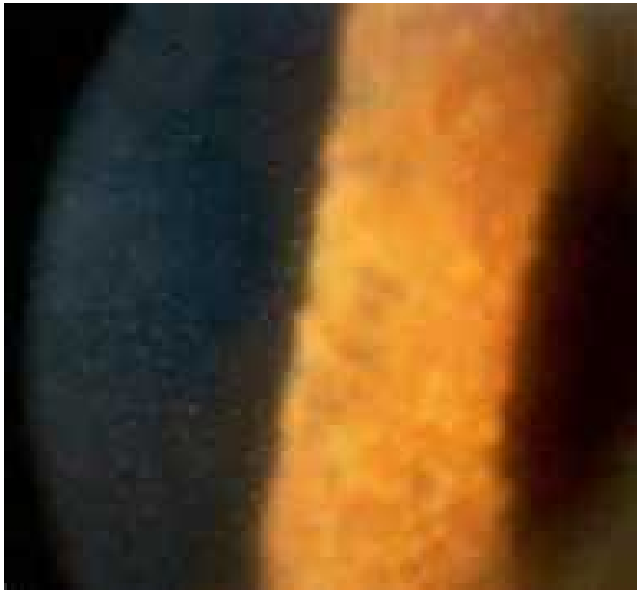


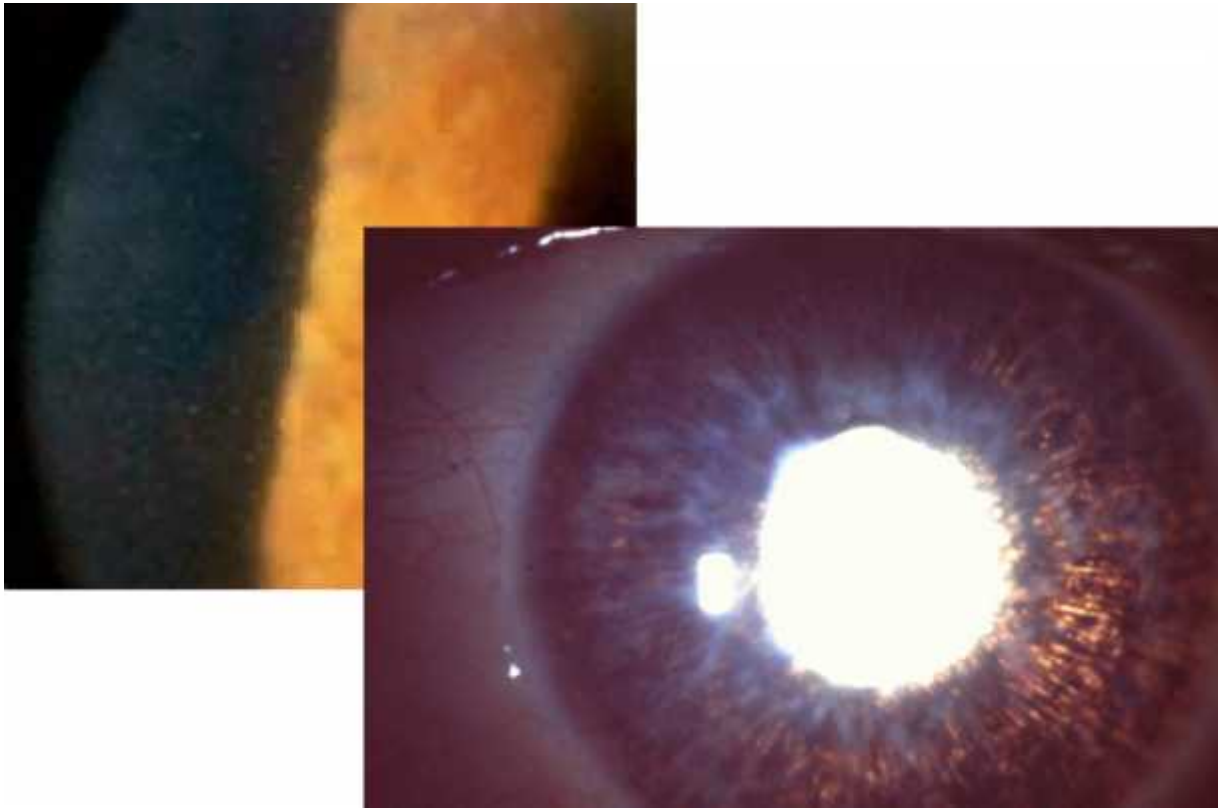
- Chronic unilateral uveitis
- Iris changes
  - Atrophy
  - Heterochromia
- Widespread non pig KP
- Anterior vitritis
- High incidence of cataract and glaucoma
- Poor response to topical steroids

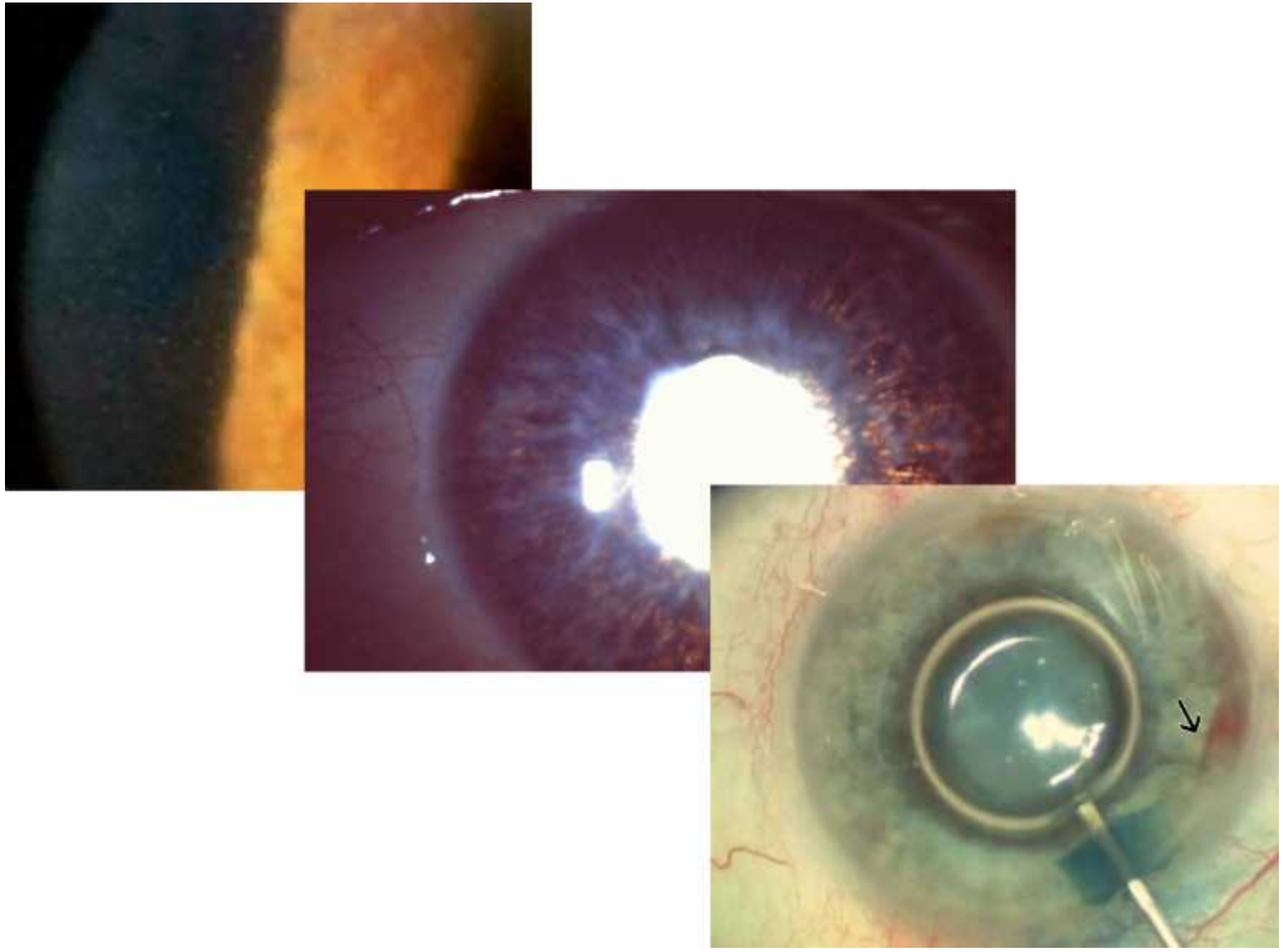
# Fuch's

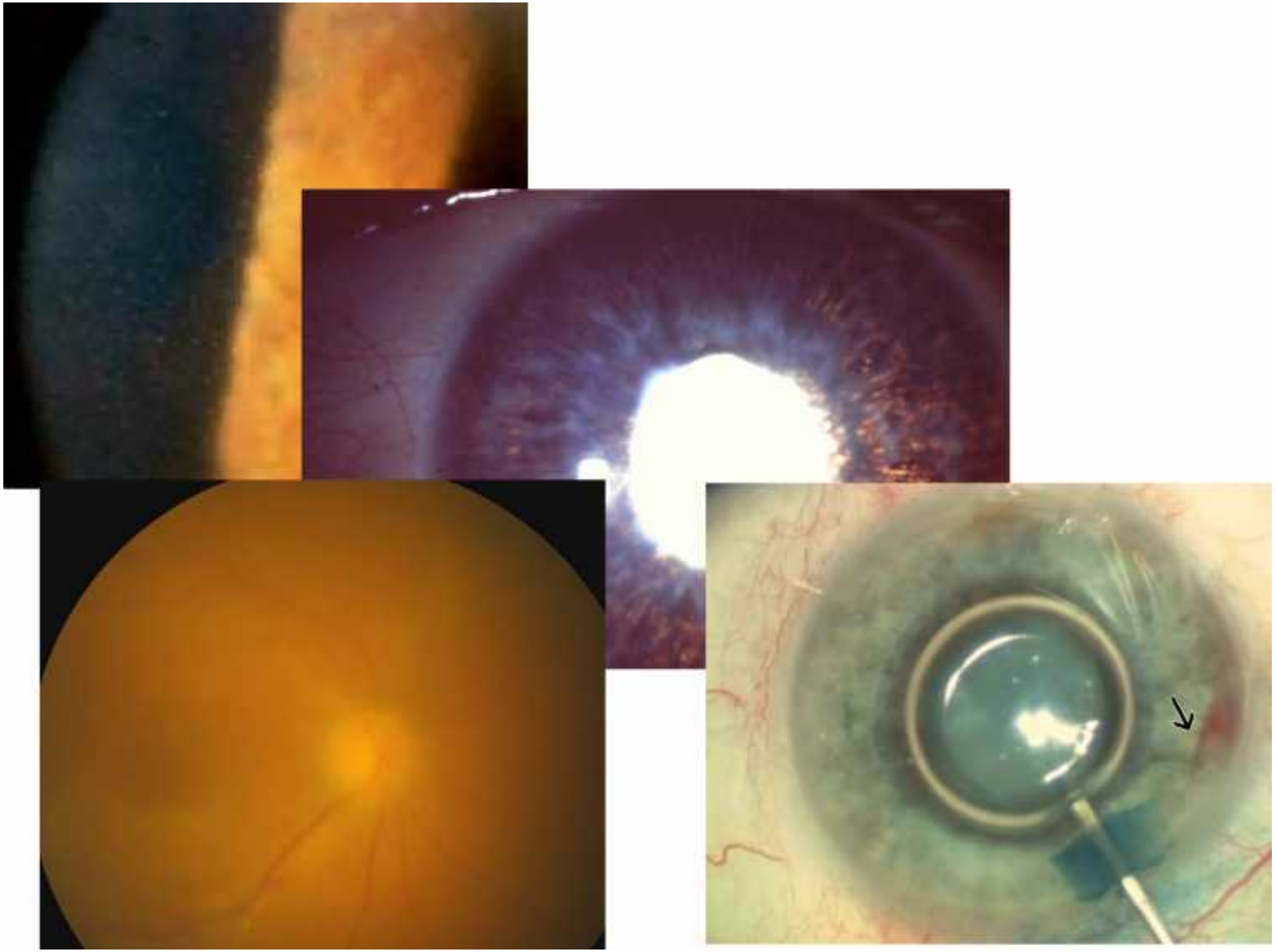


- NB
  - Characteristic KP
    - Widespread, non pigmented, stellate
  - Vitritis is common and may be severe
  - May not have heterochromia
  - 5-10% bilateral

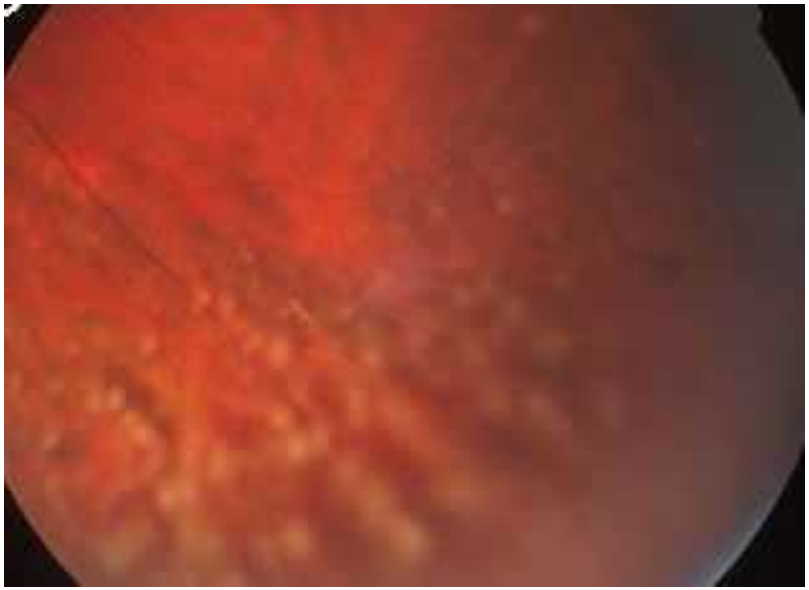








# Intermediate uveitis



- Common
- Young and middle aged
- +/- macular oedema
- +/- retinal vasculitis
- +/- inferior snow balls
- +/- snow bank

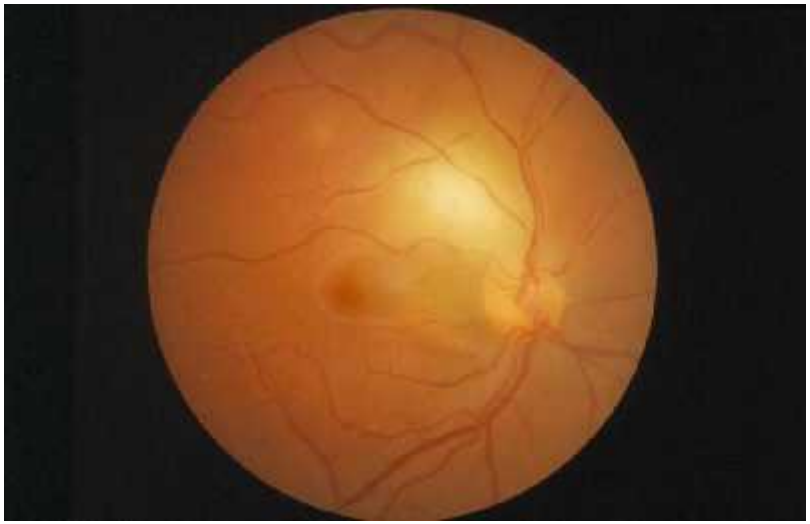
- 1/3 require topical treatment only
- 1/3 intermittent orbital or systemic therapy
- 1/3 more than steroids

# Investigation of intermediate uveitis without localising features

- History

- FBE
- U&E, LFT
- Syph serology
- ACE
- If indicated
  - MR brain
  - LP

# Unifocal chorioretinitis

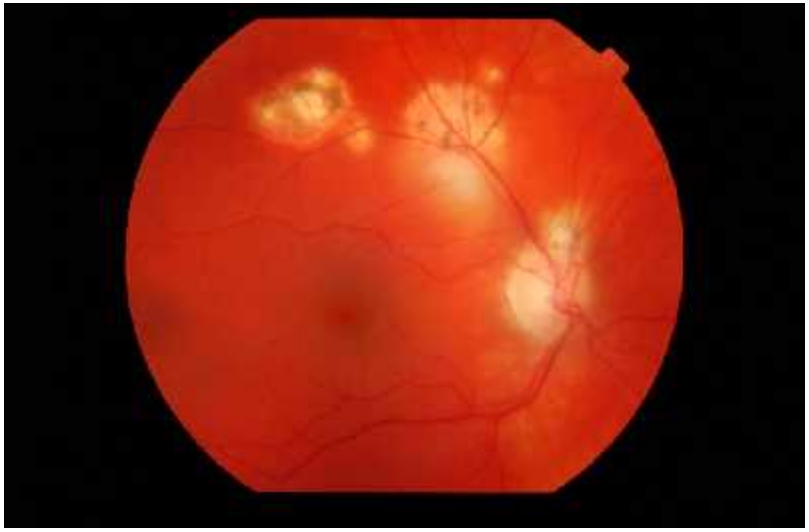


- Uveitis
- Single focus of retinal or chorioretinal inflammation

# Unifocal chorioretinitis

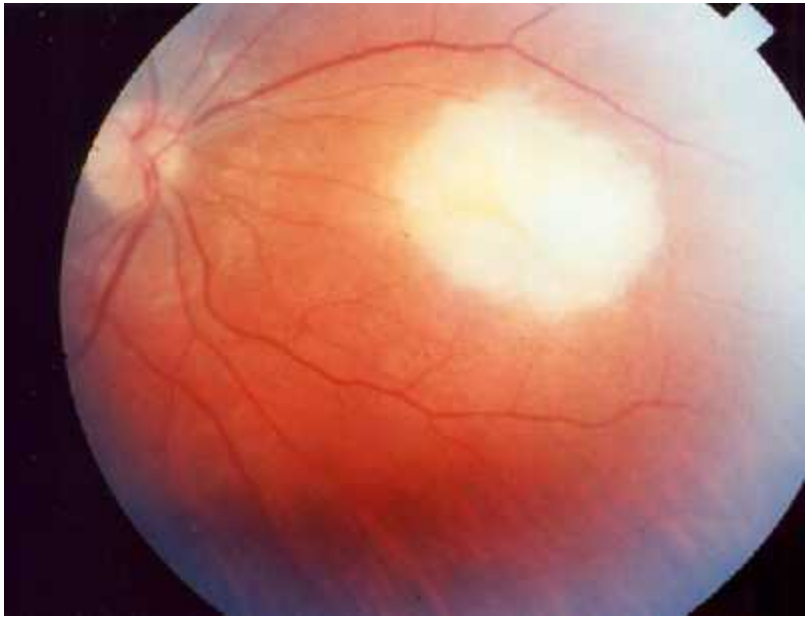
- Ocular diagnoses
  - Toxoplasmosis
  - Toxocara
  - Candida
- Systemic diagnoses
  - Sarcoidosis
  - Candida
  - (other fungal)

# Toxoplasmosis



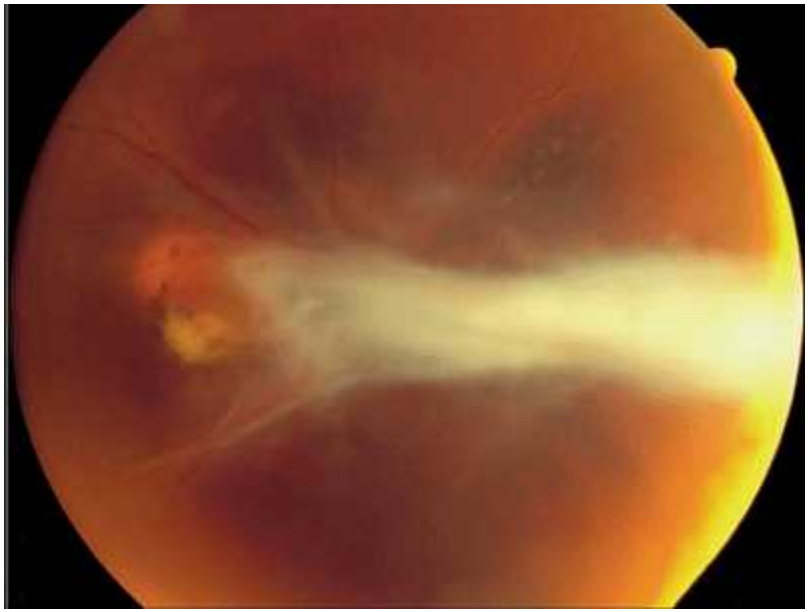
- Unilateral uveitis
- Active focus of chorioretinitis – usually adjacent to an old scar
  - AC activity
  - Raised pressure
- Otherwise well patient
- Clinical diagnosis!
- Can be confirmed by vitreous PCR if needed

# Candida



- Unilateral uveitis
- Single (or multiple) focus of chorioretinitis with no adjacent scar
- May have vitreous puffballs or preretinal lesions
- At risk patient
  - Recent IV access
  - IVDU
  - Sick inpatients
- Diagnosed clinically and confirmed with systemic cultures or vit tap/Vx

# Toxocara



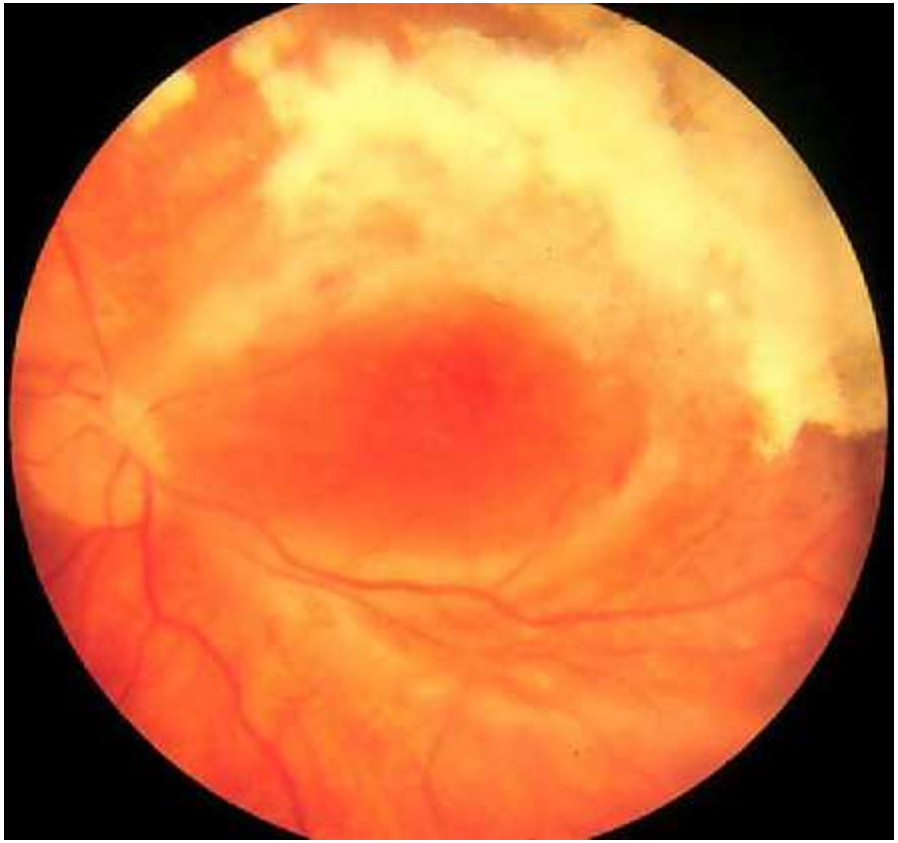
- Uncommon cause of unilateral uveitis and focal chorioretinitis
- Mainly young children
  - Focal chorioretinitis
    - Peripheral
    - Posterior pole
  - Rarely endophthalmitis

# Investigation of unifocal chorioretinitis

- History

- Toxoplasmosis serology
- FBE
- ACE
- If indicated
  - Toxocara serology
  - Blood/urine cultures
  - Vit tap
    - Toxoplasma pcr
    - Fungal cultures

# Diffuse/confluent chorioretinitis

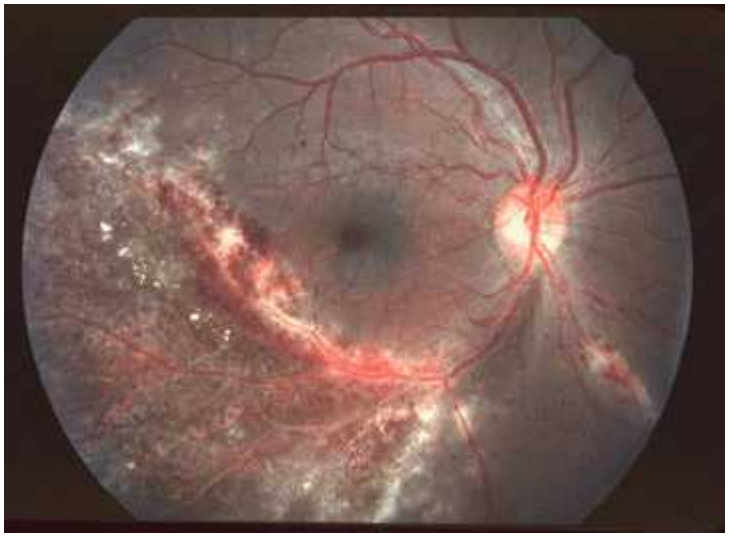


- Uveitis
- Widespread confluent area of chorioretinal inflammation
- Often with
  - Retinal vasculitis
  - Optic nerve swelling

# Diffuse chorioretinitis

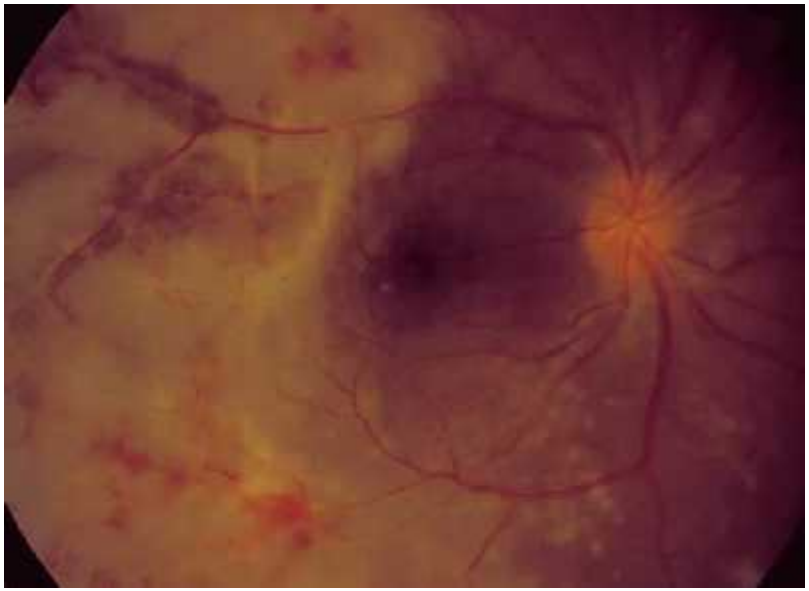
- Ocular diagnoses
  - Viral retinitis
  - Serpiginous choroidopathy
  - Fungal retinitis
- Systemic diagnoses
  - Oculo-cerebral lymphoma
  - Syphilis
  - Immuno-suppression/AIDS (and viral retinitis)

# CMV retinitis



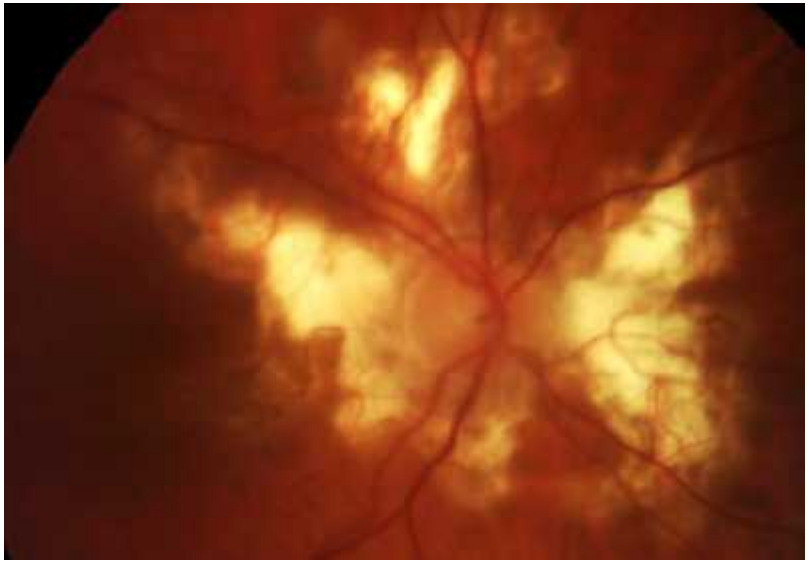
- Confluent retinitis
- Haemorrhage and progressive scarring
- Mild vitritis
- HIV positive with low CD4 count
- Treated with IV or intravitreal antivirals and with immune restoration

# Acute retinal necrosis



- Rapidly progressive peripheral retinitis
- Well patients
- VZV or HSV
- High incidence of retinal detachment
- Treated with IV/intravitreal antivirals

# Serpiginous choroiditis



- Relapsing remitting slowly progressive chorioretinitis with quiet vitreous
- Typical serpentine shape
- Posterior pole
- Beware TB

# Serpiginous in Australia

- ¼ had TB


Original Article

**Retrospective analysis of the natural history and management of serpiginous choroiditis in Australia and New Zealand**

Jason Tonello MBBS, Anthony J Hall FRANZCO, Jo Sims FRANZCO, Samantha Fraser Bell FRANZCO, Jane Khan FRANZCO, Christine Younan FRANZCO, Brian Kent Smith FRANZCO, Stephanie Young FRANZCO, Lidho Paul MSc, Lyndell L Lim FRANZCO

# Investigation of diffuse chorioretinitis

- History

- FBE, U&E, LFT
- HIV viral load, CD4 count
- CMV viral load
- Consider
  - Quantiferon
  - Vit tap
    - Herpes pcr
    - Fungal cultures
  - MRI

# Multifocal choroiditis

- Multifocal choroiditis is common in many forms of uveitis and often does not carry diagnostic significance

# Multifocal choroiditis

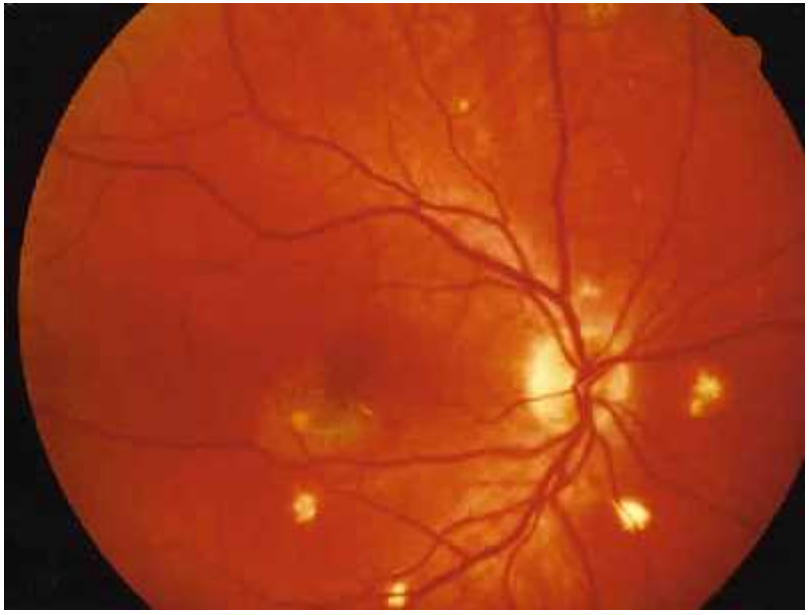
- Ocular diagnoses
  - No inflammation
  - Mild inflammation
  - Significant inflammation
- Systemic diagnoses
  - Sarcoidosis
  - Behcet's
  - Oculo-cerebral lymphoma
  - Infection
    - TB
    - Cryptococcus
    - PCP
    - syphillis

# An approach to multiple white retinal lesions

- No inflammation
  - Flavimaculatus
  - Punctata albescens
  - PIC
  - POHS
- Min inflammation
  - AMPPE
  - MEWDS

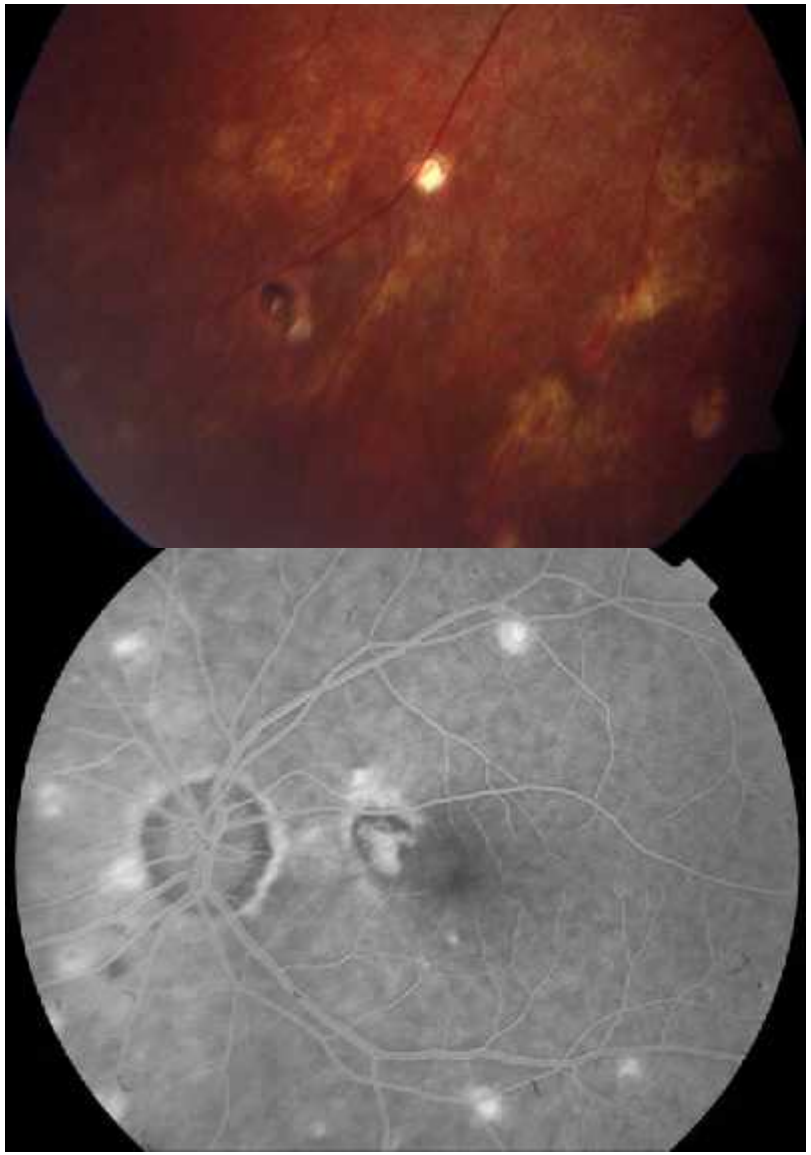
- Lots of inflammation
  - Multifocal choroiditis
  - Sympathetic
  - Birdshot
  - Sarcoidosis
  - Infections

# Punctate inner choroidopathy (PIC)



- Myopic women
- Small pale atrophic lesions around the posterior pole
- Minimal or no vitritis
- CNVM common

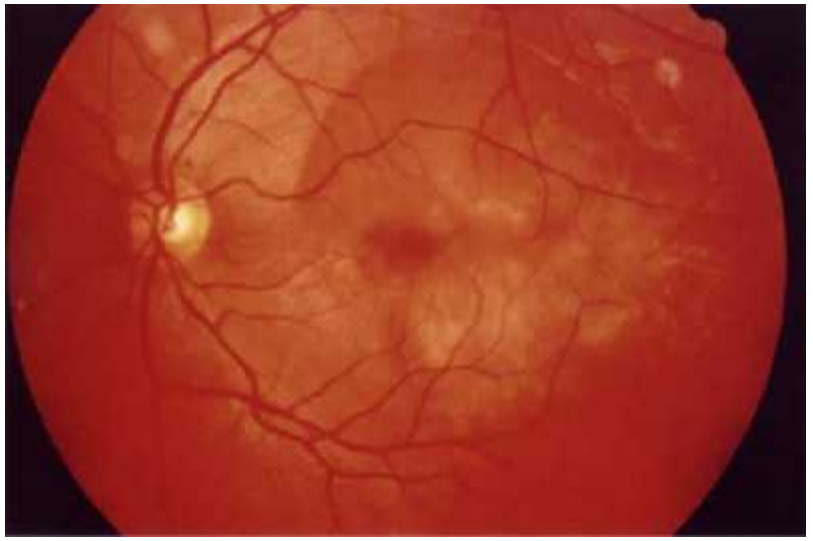
# POHS



- Atrophic peripheral scars and streaks
- Peripapillary atrophy
- Macular CNVM very common
- More common in the USA

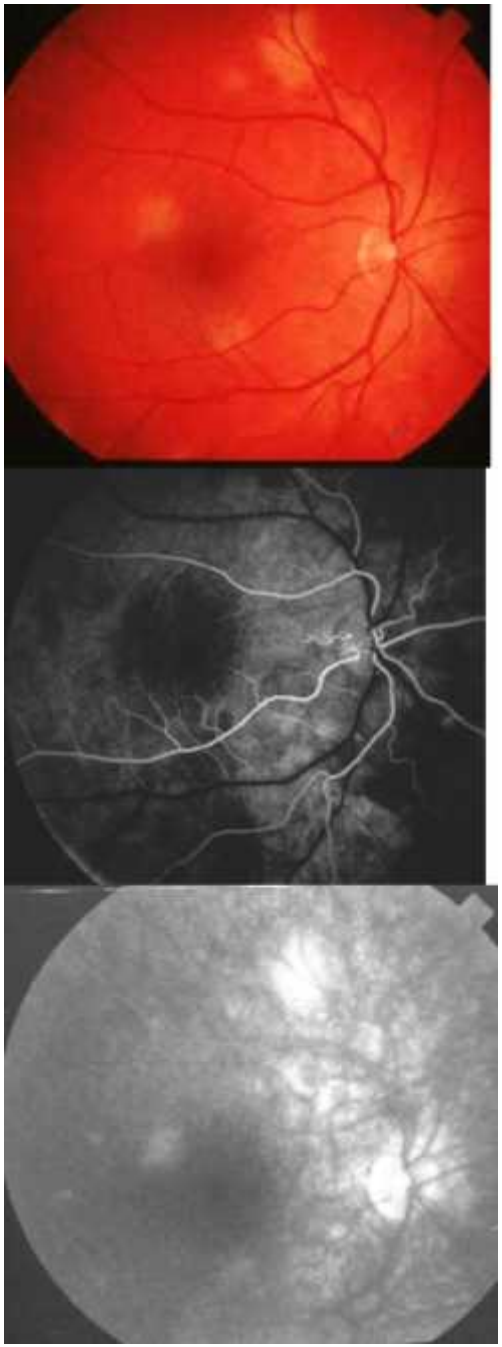
# AMPPE

- Young adults
- acute decrease in central visual acuity in association with subretinal yellow-white placoid lesions of the posterior pole
- Mild AC and vitreous activity
- spontaneous recovery

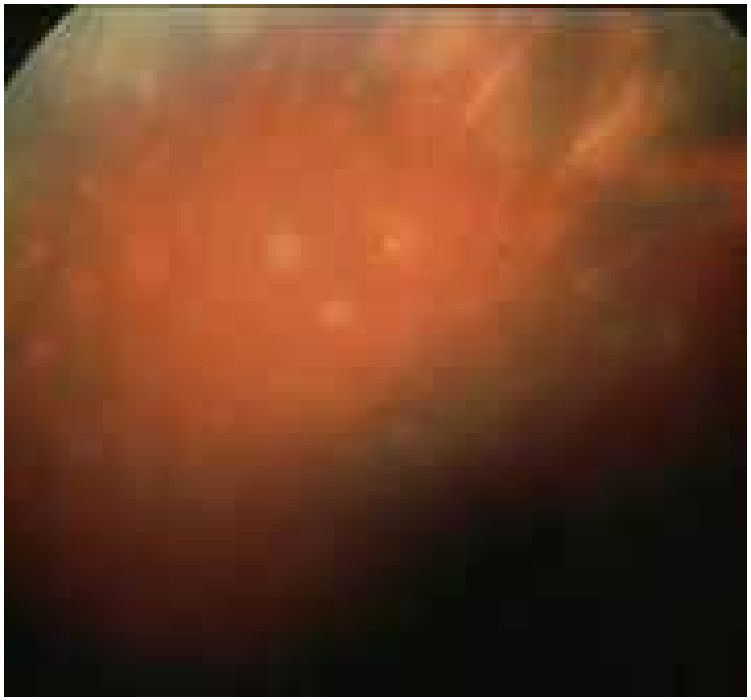


# MEWDS

- Young women
- Sudden onset unilateral visual loss with photopsia
- Small pale evanescent white outer retinal dots
- Mild AC and vitreous activity
- Spontaneous recovery

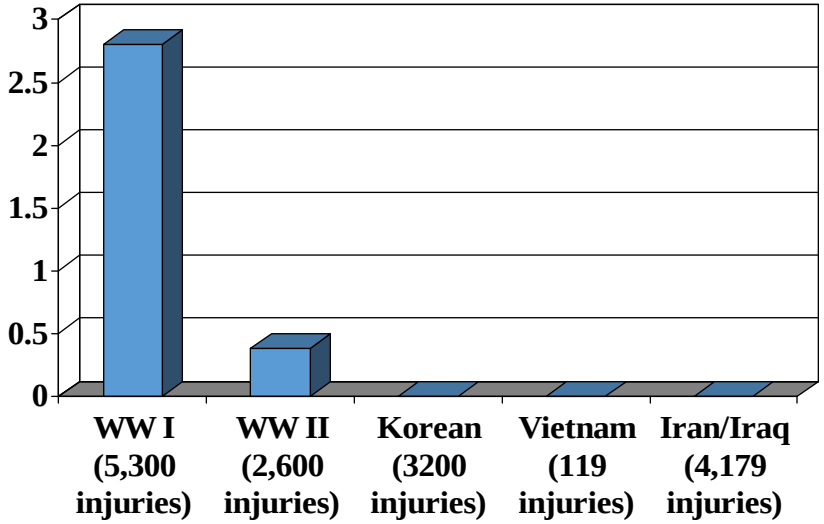


# Sympathetic ophthalmia

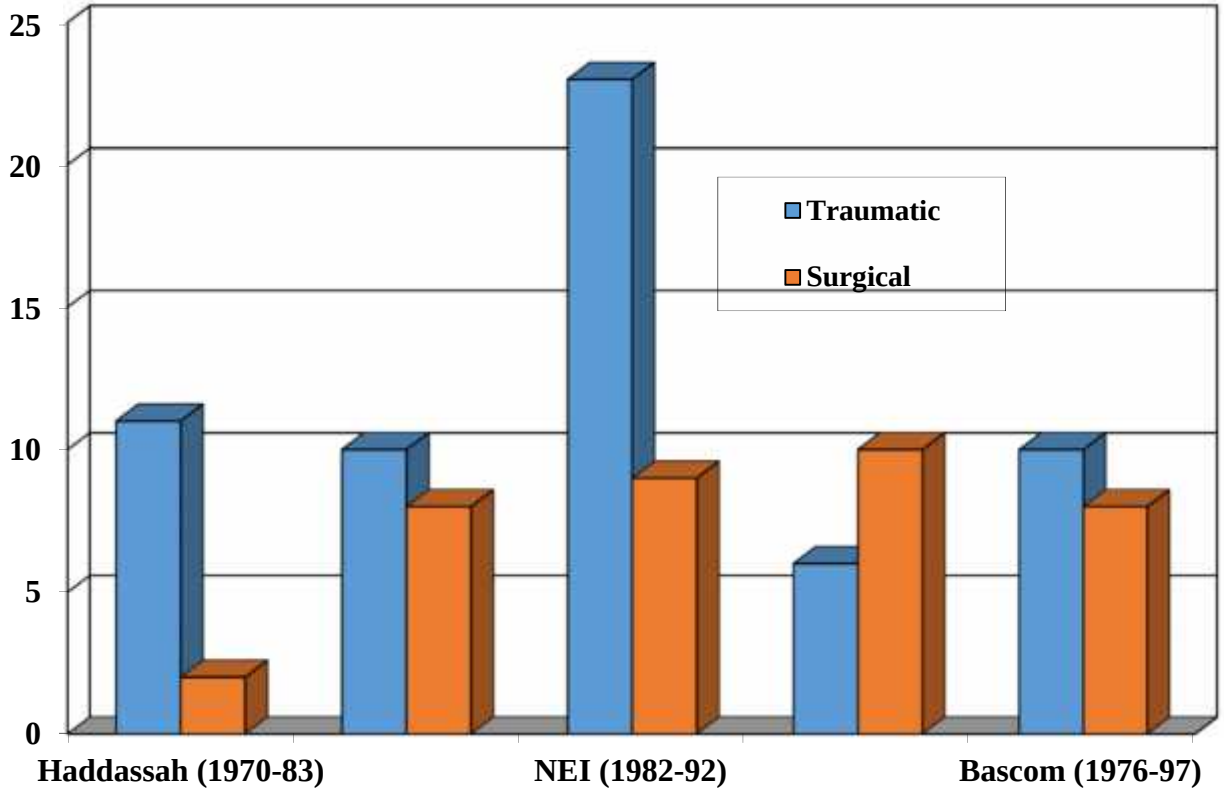


- Bilateral uveitis following penetrating trauma
  - Surgery
  - Accidental trauma
- Uveitis with
  - multifocal choroiditis
- Diagnosis of exclusion

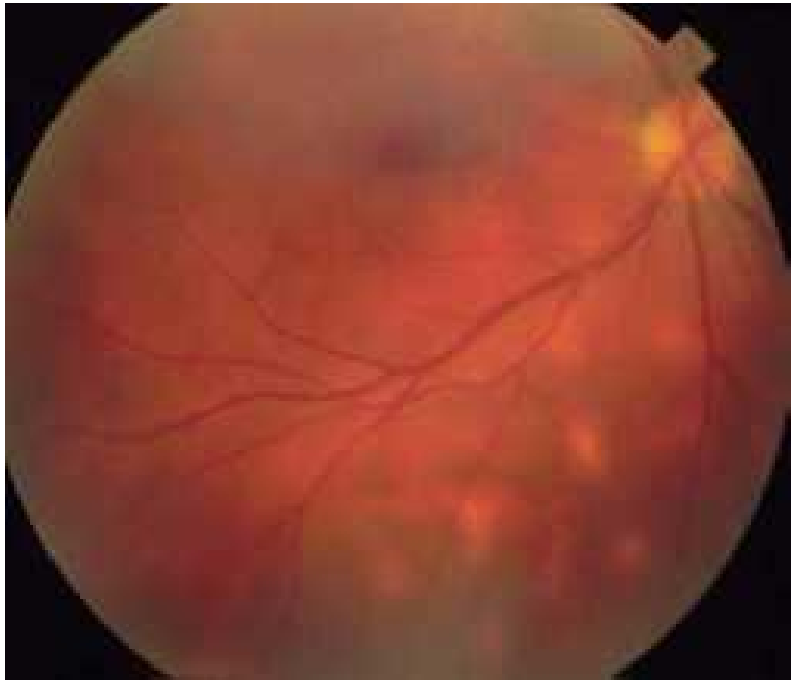
# Incidence of sympathetic ophthalmia after penetrating trauma (cases/1,000 injuries)



# Aetiology of sympathetic ophthalmia

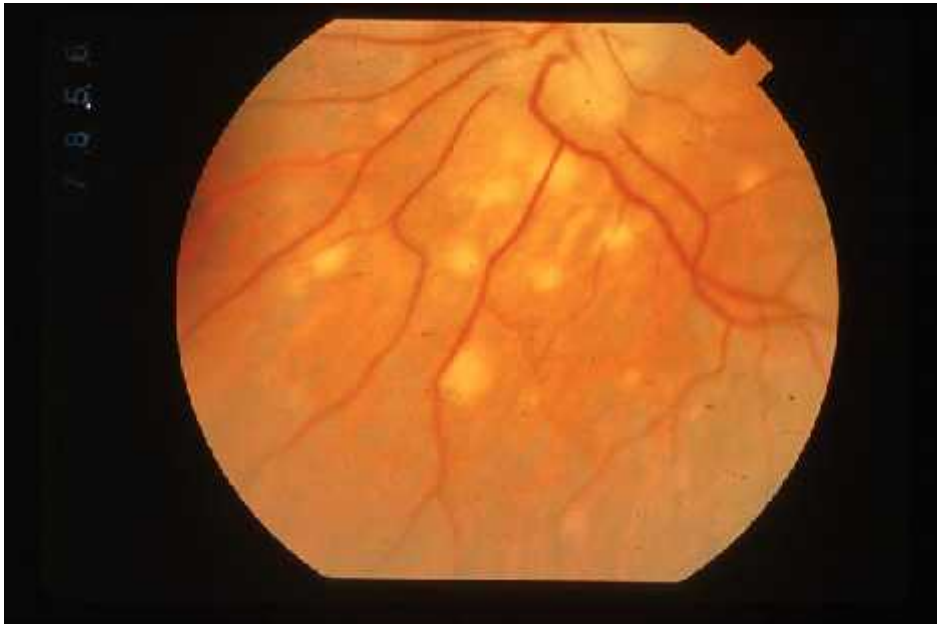


# Birdshot chorioretinopathy



- Middle aged women > men
- HLA A 29 +ve
- Bilateral widespread pale retinal lesions with minimal pigmentation
- +/- retinal vasculitis
- +/- macular oedema

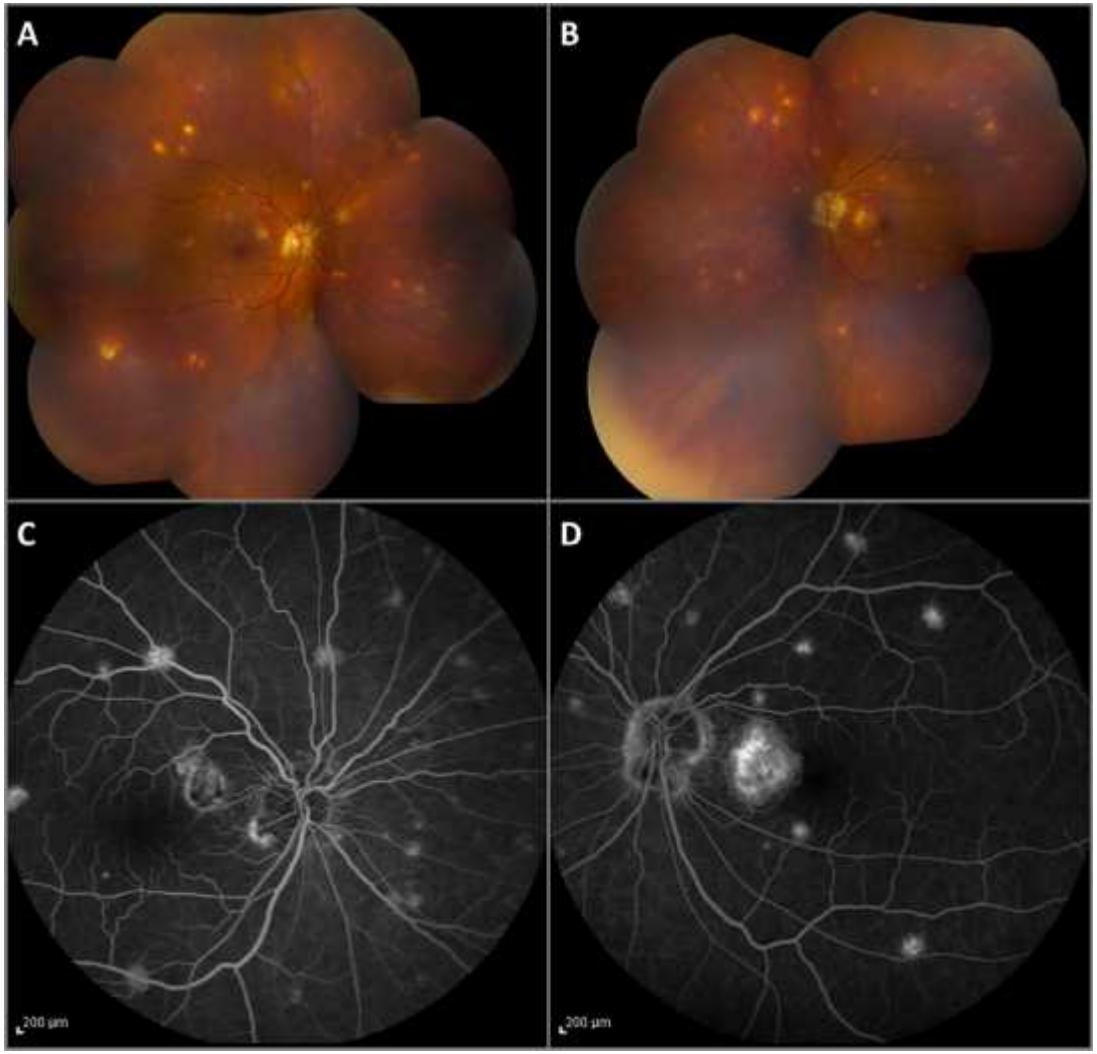
# Non infectious multifocal choroiditis



- Is still generally idiopathic
- Think of
  - Sarcoid
  - Behcet's
  - SO

# Infectious multifocal choroiditis

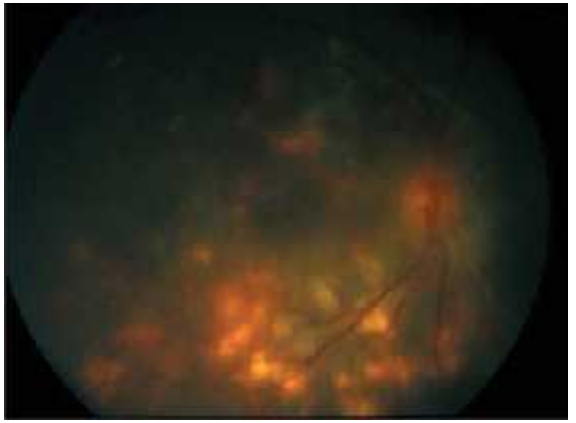
- Aetiology
  - Mycobacterial
    - Tuberculous
    - Non-tuberculous
  - Cryptococcus and other fungus
  - Pneumocystis carinii



- Pneumocystis choroiditis
  - Exclusively in AIDS patients on inhaled prophylaxis for Pneumocystis pneumonia

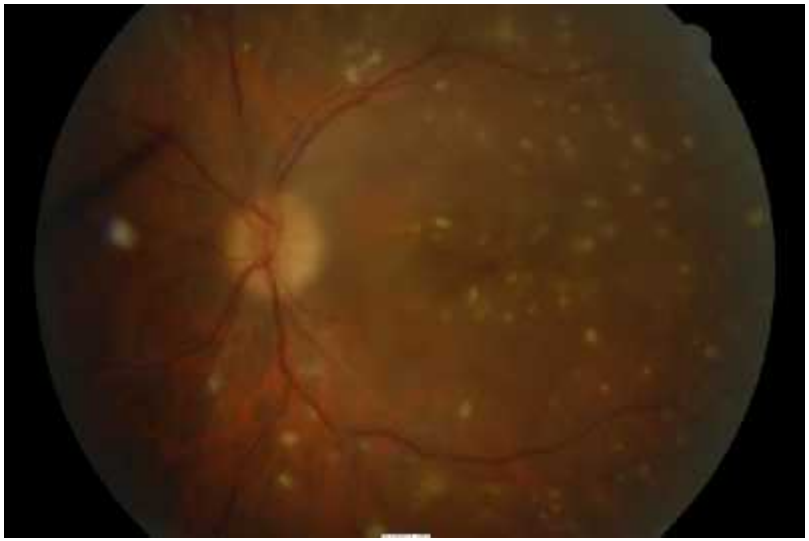
- Cryptococcal choroiditis
  - occurs in patients with cryptococcal meningitis

- Multifocal mycobacterial choroiditis
  - Uncommon
  - Very sick patients with disseminated TB
  - Non TB mycobacterium



# Syphilitic retinitis

- Punctate inner retinitis
- Secondary syphilis

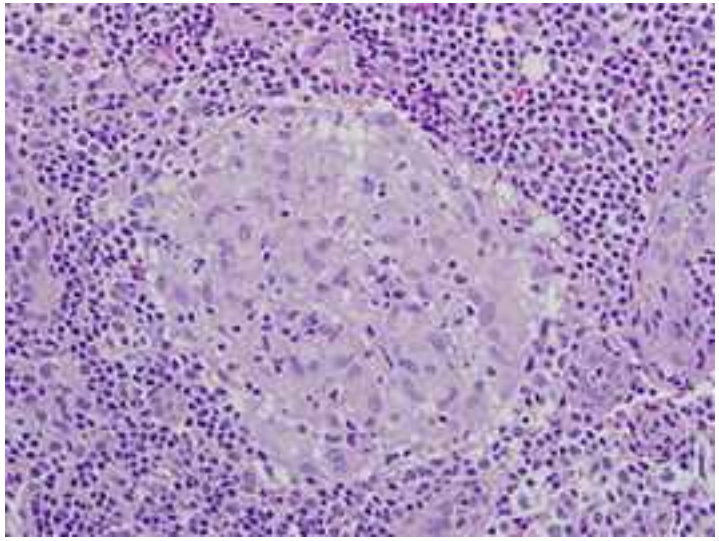


**Syphilitic Punctate Inner Retinitis in Immunocompetent Gay Men**

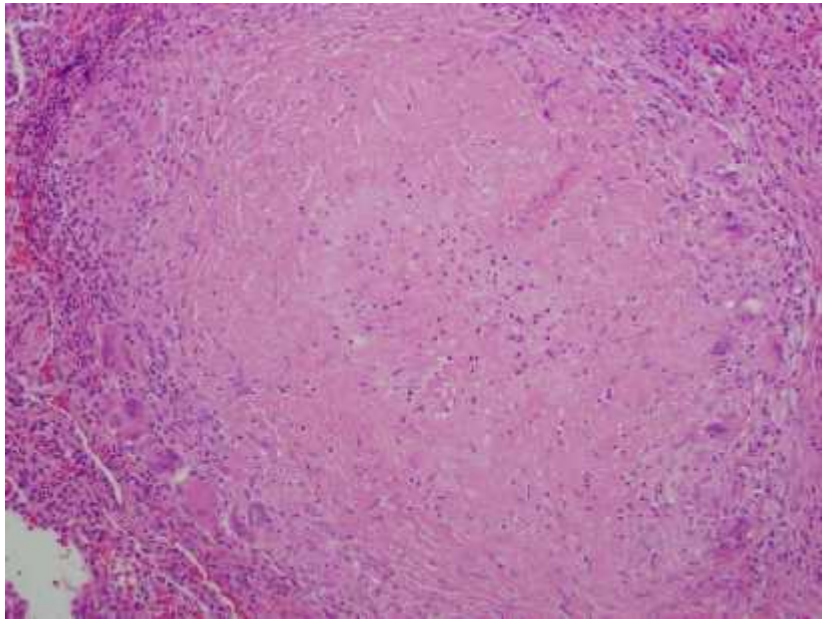
Sangeeta Wadhwani, FRCOphth, FRANZCO,[1,2] Cecilia Lau, FRANZCO,[1] Richard Stawell, FRANZCO, FRACS,[1] Jonathan Yeoh, FRANZCO,[1] Anthony Hall, MD, FRANZCO,[3] Paul Turner, MD, FRANZCO[3]

# Sarcoidosis

- Focal granulomatous inflammation
- Non necrotising granulomas
- Generalised immunosuppression



# “Caseating” granulomas



- Usually infectious
  - TB
  - Cat scratch

# Sarcoidosis

- Focal granulomatous inflammation
- Generalised immunosuppression

- Lung
- Skin
- Nodes
- Eyes
- Liver
- Heart
- CNS
- kidneys

# Investigations for sarcoidosis

- FBE
  - Lymphopenia
- U and E, LFT, Ca
- ACE
  - Beware ACE inhibitors
- Lysozyme
- CT chest
  - Adults with pulmonary symptoms

| | Sensitivity | Specificity | Youden Index | Error |
|---|---|---|---|---|
| ACE | 0.53 | 0.88 | 0.41 | 0.25 |
| sIL-2R | 0.91 | 0.53 | 0.45 | 0.19 |
| Lysozyme | 0.62 | 0.58 | 0.20 | 0.40 |
| ACE/sIL-2R | 0.80 | 0.29 | 0.50 | 0.17 |
| ACE/lysozyme | 0.43 | 0.29 | 0.41 | 0.23 |
| sIL-2R/lysozyme | 0.73 | 0.71 | 0.44 | 0.29 |
| ACE/sIL-2R/lysozyme | 0.75 | 1.00 | 0.45 | 0.13 |

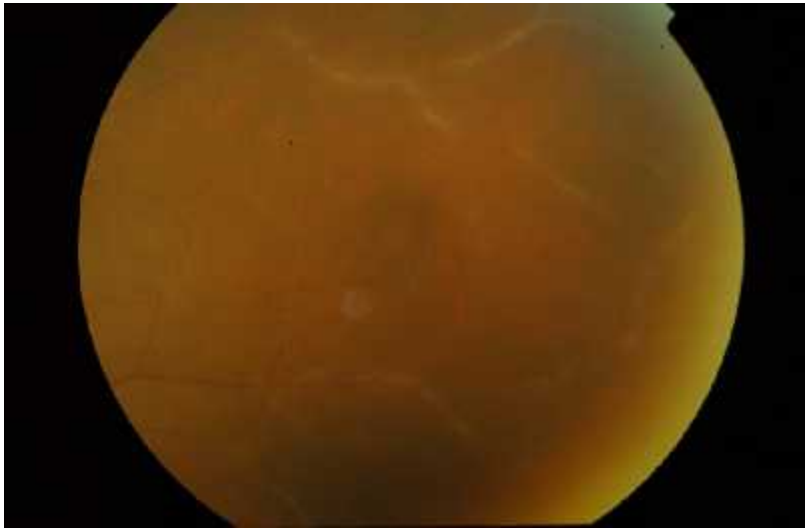
Based on the Between-the-negative rule (BN rule, Shen 2018), sensitivity, specificity, Youden index and classifical discrimination errors are shown for possible combinations of ACE, sIL-2R and lysozyme regarding the predictive value for pulmonary involvement. For this, the combination of ACE and sIL-2R is the most reliable predictor (Youden Index 0.50). Youden Index is a measure combining sensitivity and specificity (= sensitivity + specificity - 1).

# Investigation of multifocal chorioretinitis

- History

- FBE, U&E, LFT
- ACE
- If indicated
  - HLA A 29
  - Quantiferon
  - Blood/urine cultures
  - FA
  - Syph serology

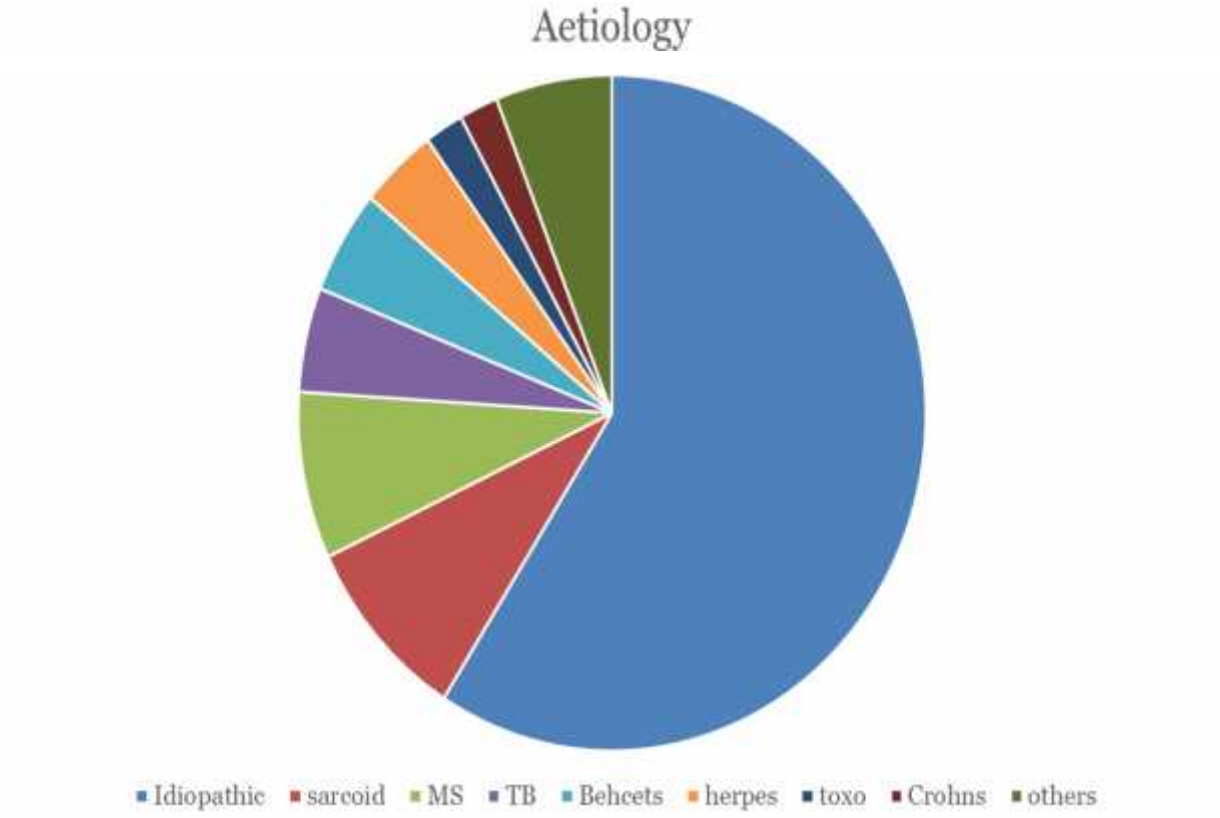
# Retinal vasculitis



- Uveitis
- Inflammation localised around blood vessels (esp veins)

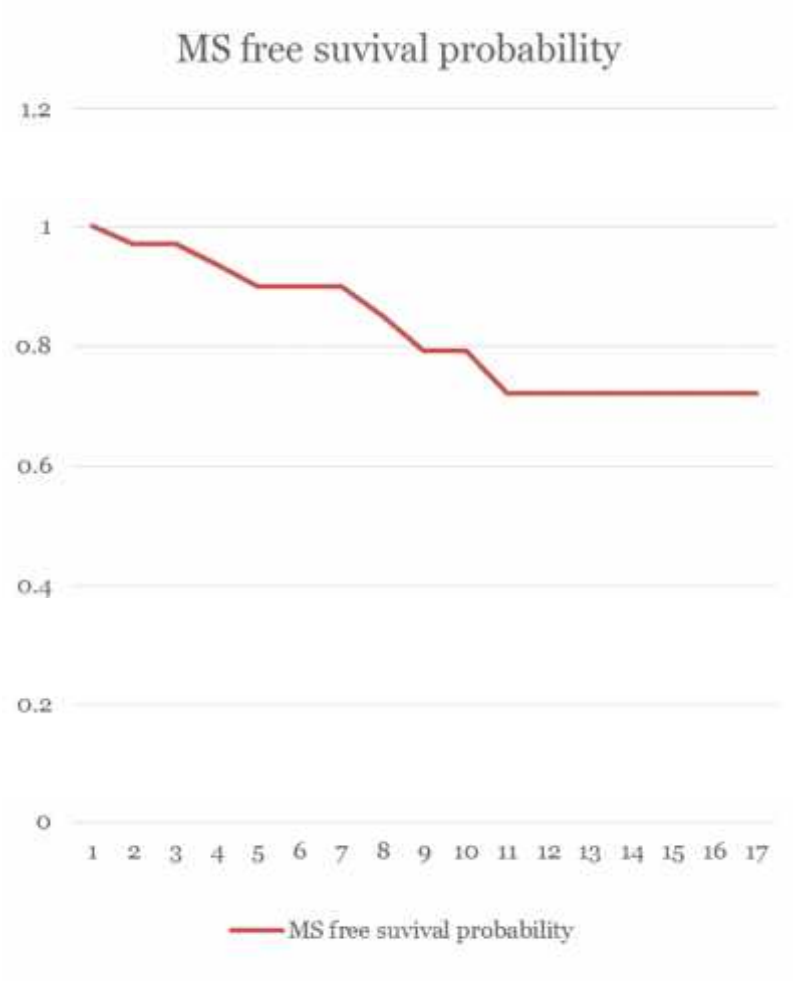
# Retinal vasculitis - aetiology

- Aetiology in 103 patients in an Australian uveitis clinic



# Development of MS in intermediate uveitis (with or without vasculitis)

- At 10 yrs the chance of MS is 28%



# Uveitis in MS treatment trials

- All patients with MS on treatment were followed regularly
- 4735 unique patients in 6 different trials
- All with reg ophthalmic follow up
- 34/4735 (0.7%) developed uveitis

Vivien Li, MBBS
Jack Kane, MBBS
Helen H.L. Chan, MBBS
Anthony J. Hall, MD*
Helmut Butzkueven, PhD*

*Neurol Neuroimmunol Neuroinflammation*
2014;1:e13; doi: 10.1212/NXI.0000000000000013

CONTINUING FINGOLIMOD AFTER DEVELOPMENT OF MACULAR EDEMA: A CASE REPORT

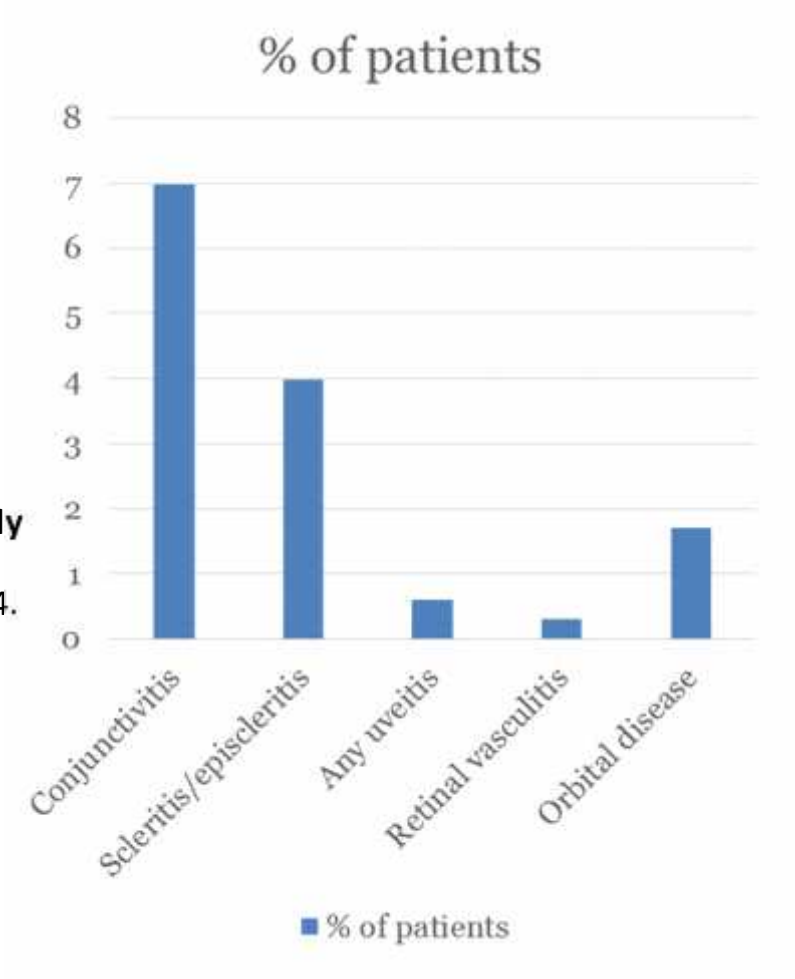
OPEN

Fingolimod is the first effective oral agent in widespread use for relapsing-remitting multiple sclerosis (MS), but it can cause macular edema (ME) as an uncommon complication. ME may be mild and asymptomatic, but it can also produce visual impairment. The mechanism of fingolimod-associated ME (FAME) is thought to be through sphingosine-1-phosphate receptor antagonism, affecting endothelial integrity and increasing the risk of microvascular leaks.[1]

# Retinal vasculitis as part of systemic vasculitis

- 1286 patients with systemic vasculitis
  - PAN (393)
  - GPA (343)
  - MPA (280)
  - EGPA/CSS (270)

- Rothschild PR[1], Pagnoux C, Seror R, Brézin AP, Delair E, Guillevin L. **Ophthalmologic manifestations of systemic necrotizing vasculitides at diagnosis: a retrospective study of 1286 patients and review of the literature.** Semin Arthritis Rheum 2013 Apr;42(5):507-14.



# Investigation of “typical” retinal vasculitis

- History
- Examination

- Tests
  - ACE (CXR CT chest)
  - Quantiferon
  - T P IgG
  - Consider MRI

# Investigation of “Atypical” retinal vasculitis – acute retinal microangiopathy

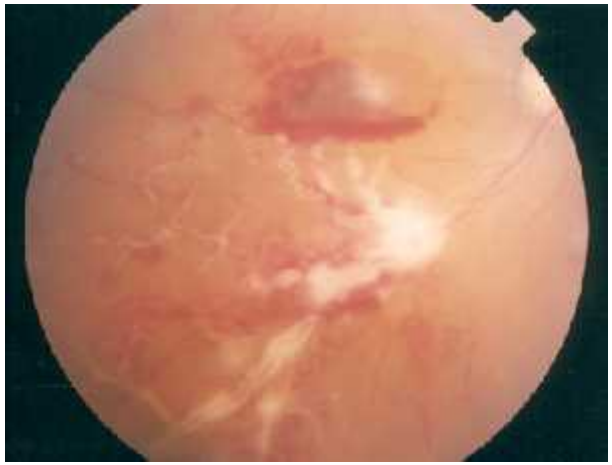
- History
- Examination

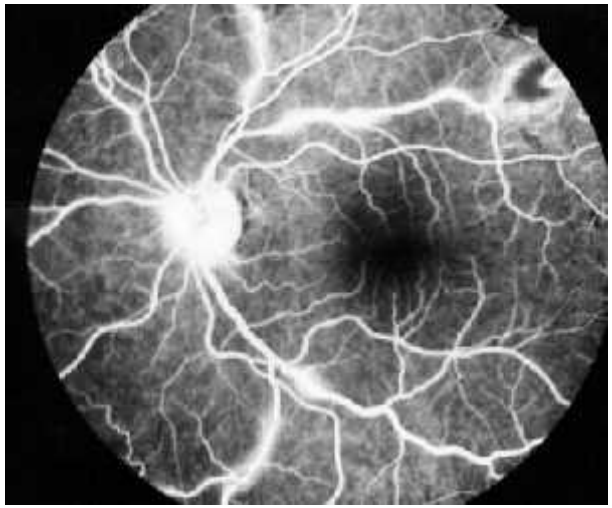
- Tests
  - FBE
  - U&E, LFT,
  - ANA
    - Lupus anticoagulant
    - Anti-cardiolipin antibodies
  - Se Igs and cryoglobulins
  - Se  viscosity

# Retinal vasculitis - aetiology

- Ocular diagnoses
  - Intermediate uveitis
  - Secondary to other uveitis
    - Especially retinal infections
    - Toxo
    - Viral retinitis
  - Eale's disease
- Systemic diagnoses
  - Often idiopathic
  - Sarcoidosis
  - Behcet's
  - MS
  - Rarely systemic vasculitis

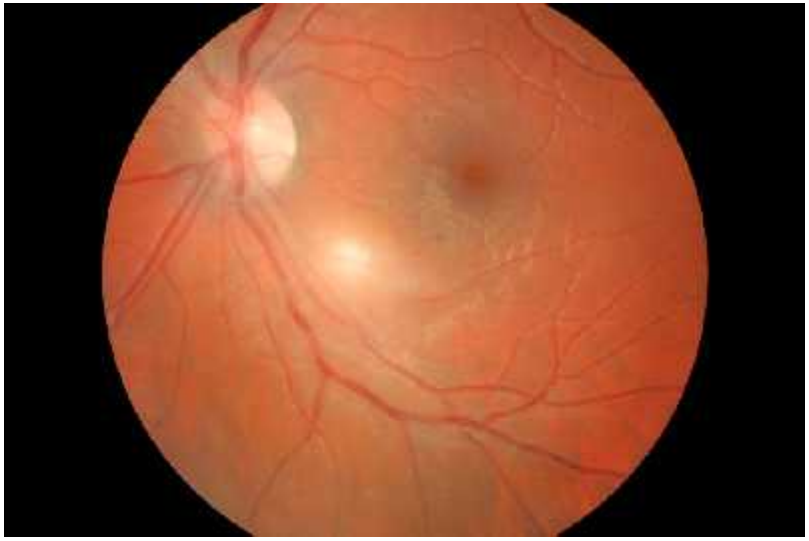
# Eale's disease





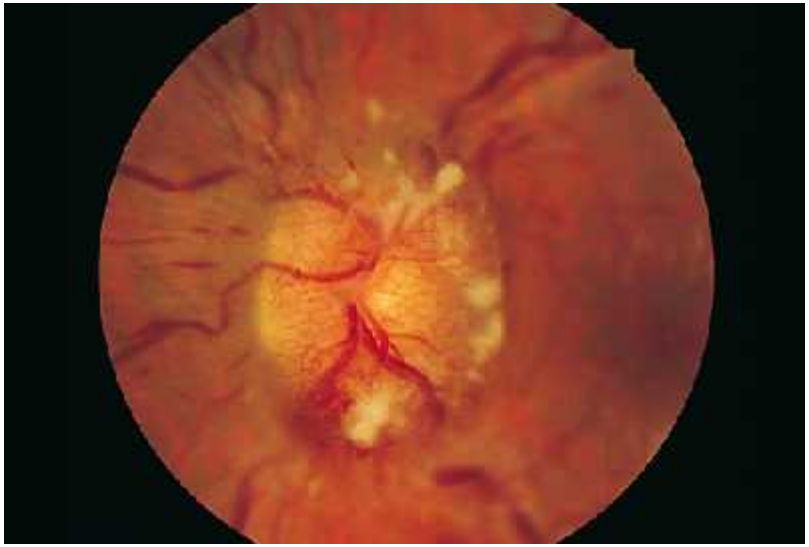
- Occlusive retinal vasculitis
- Veins > arteries
- Secondary neovascularisation and vitreous haemorrhage
- Most common in patients from south Asia
- Diagnosis of exclusion

# Retinal vasculitis secondary to retinal infections



- Often seen with
  - Toxo
  - Viral retinitis

# Optic papillitis

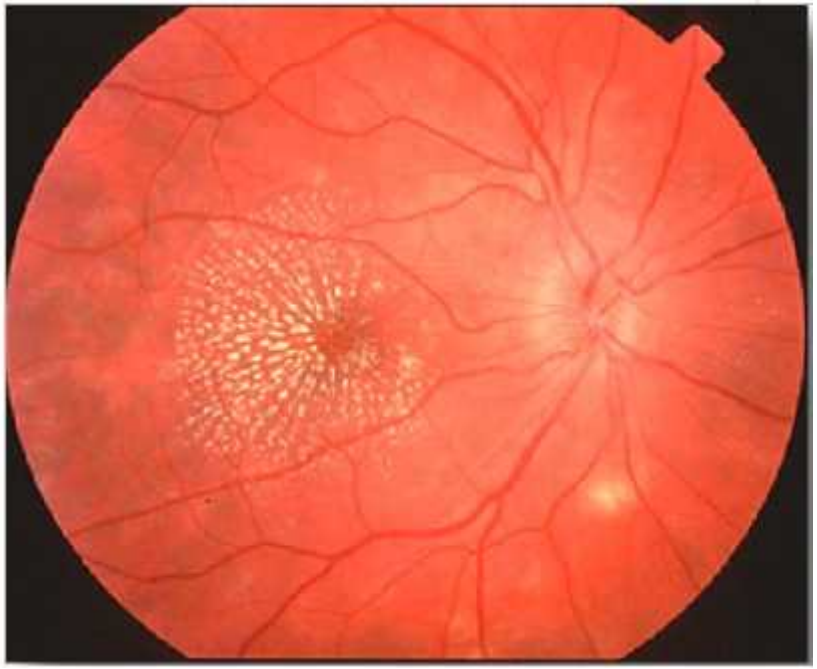


- Uveitis
- Inflammation centred on the optic nerve
- Note: mild disc swelling commonly accompanies many forms of uveitis and is not necessarily of diagnostic significance

# Optic papillitis

- Ocular diagnoses
  - Associated with other uveitis
  - Leber's stellate neuroretinitis
  - Bartonella
- Systemic diagnoses
  - MS
  - Sarcoidosis
  - VKH
  - Bartonella
  - Lyme disease
  - Syphilis

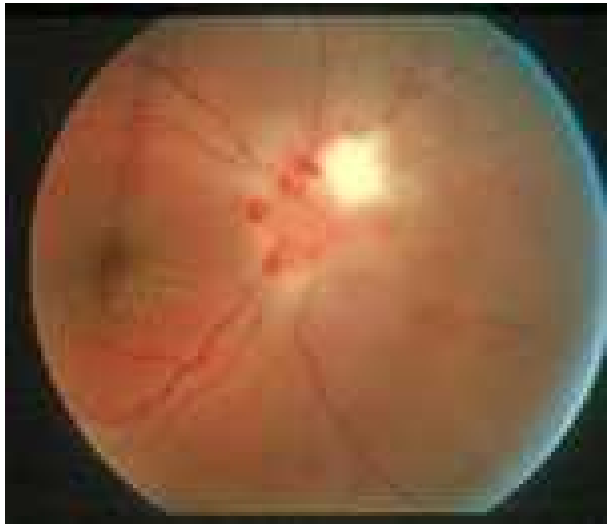
# Bartonella

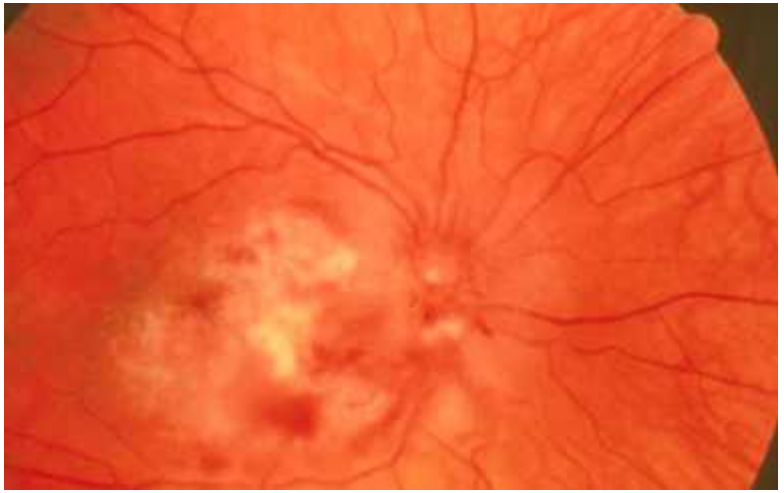


- Systemic illness following cat scratch with lymphadenopathy
- Disc swelling
- Stellate maculopathy

# Papillitis and adjacent retinitis

- Immunocompetent
  - Toxo
- Immunosuppressed
  - CMV



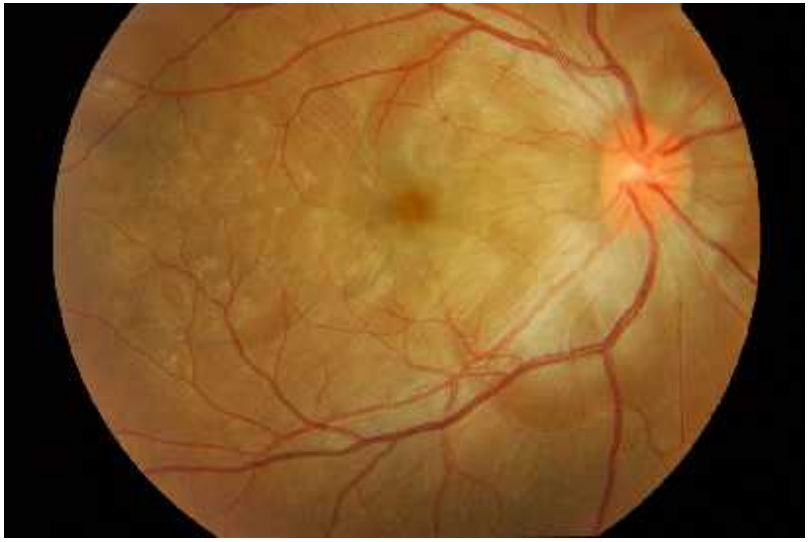


# Investigation of optic papillitis

- History

- Syph serology
- Bartonella serology
- ACE
- MRI
- If indicated
  - Lyme serology
  - Toxo serology
  - Quantiferon
  - Lebers mitochondrial gene testing

# Uveitis and serous retinal detachment



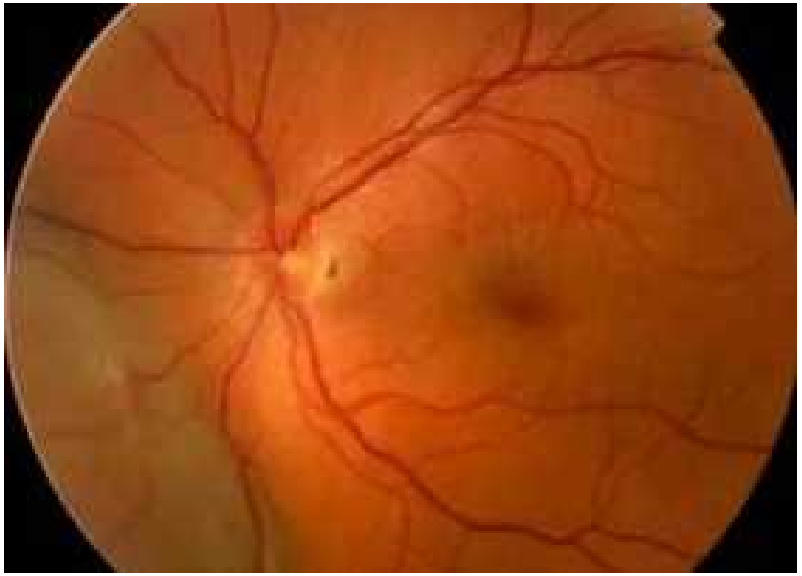
- Serous detachment is uncommon in uveitis
- Usually caries diagnostic significance

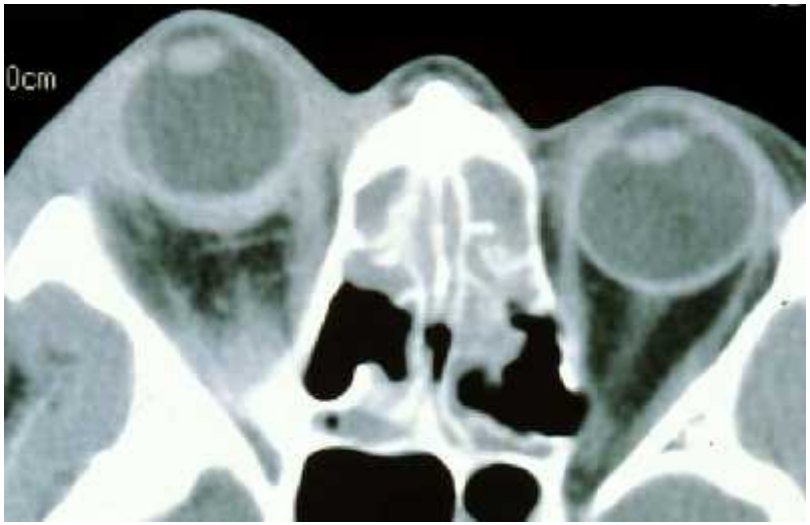
# Uveitis and serous retinal detachment

- Ocular diagnoses
  - Posterior scleritis
  - Sympathetic ophthalmia
- Beware
  - Steroid induced CSR
  - CNVM
  - Uveal effusion syndrome
- Systemic diagnoses
  - VKH

# Posterior scleritis

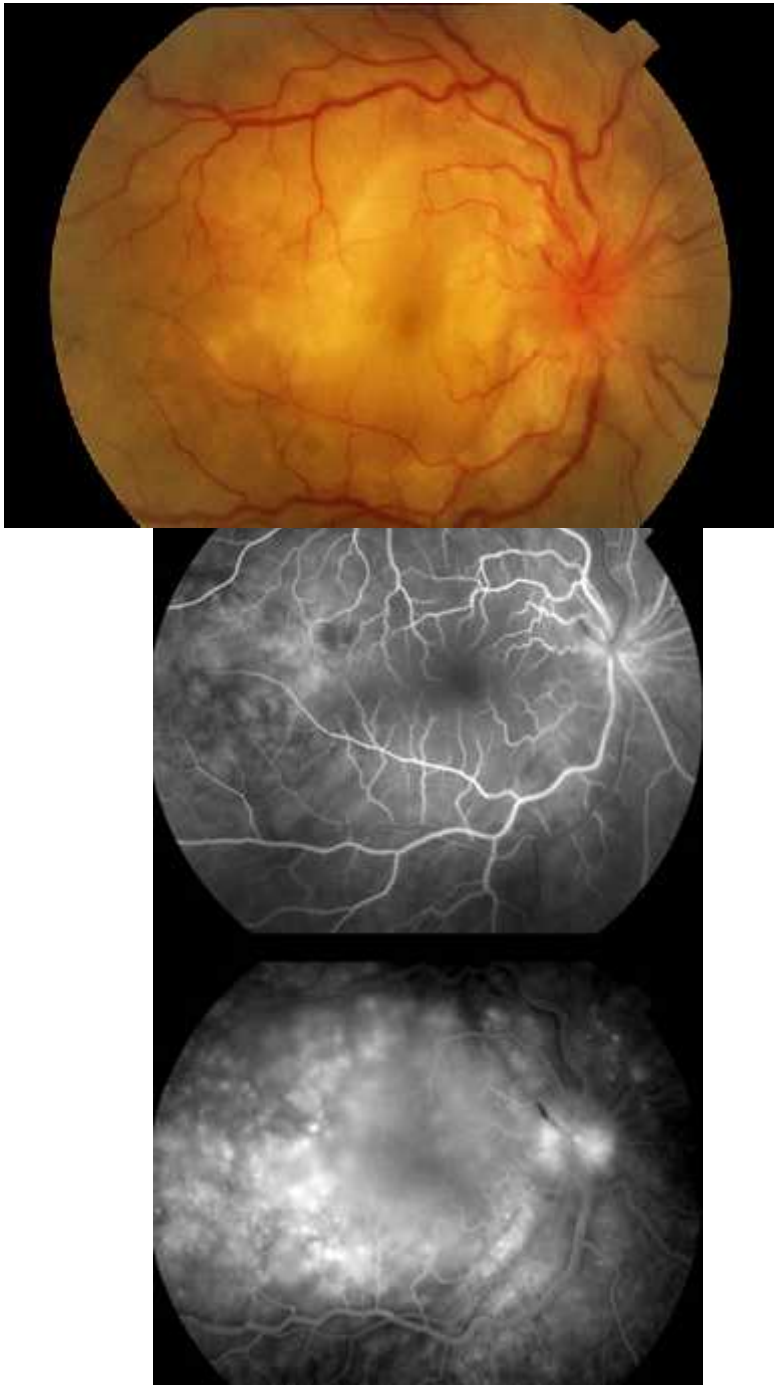
- Posterior scleritis
  - Pain
  - Disc swelling
  - Choroidal folds
  - Serous detachment
  - Minimal uveitis





# VKH

- Headaches
  - CSF lymphocytosis
- Hearing changes
- Uveitis
  - Serous detachment
  - Disc swelling
  - Multifocal leak and disc swelling on FA

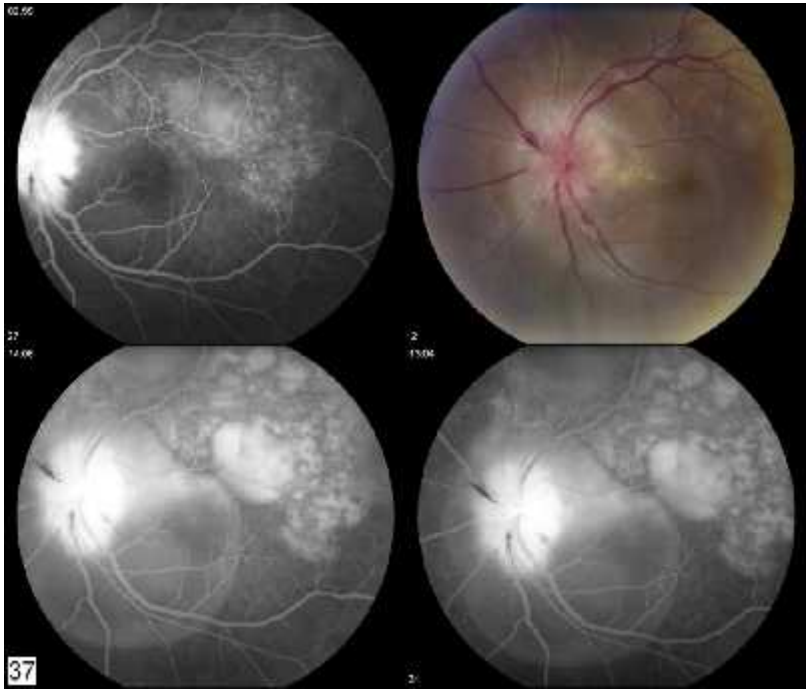


# VKH

- 3 out of
- Bilateral anterior uveitis
- Bilateral posterior uveitis with
  - Serous ret det
  - Disc swelling
  - RPE changes
- CNS involvement
  - Pleocytosis
  - HA
  - Tinnitus/CN involvement
- Skin involvement

# VKH

- 3 phases
- Meningo-encephalitic phase
- Uveitis/auditory phase
- Skin/hair phase



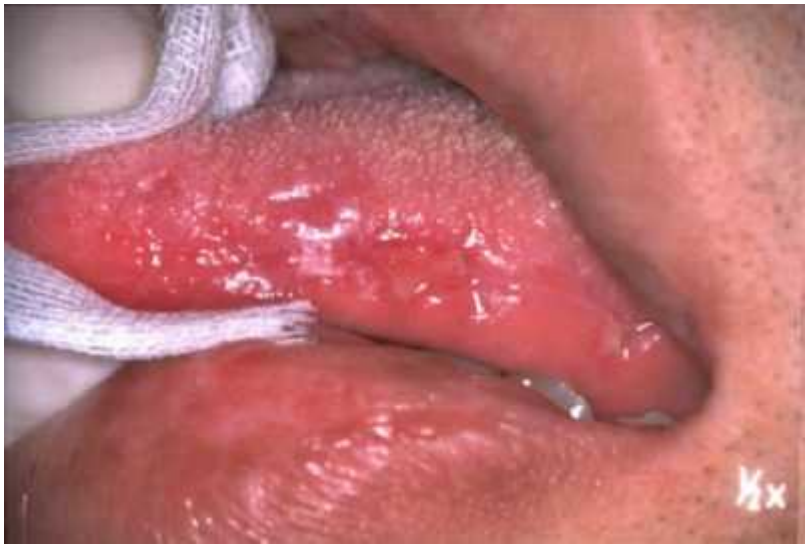
| Type of posterior uveitis | Ocular diagnoses | Systemic diagnoses |
|---|---|---|
| Vitritis/diffuse uveitis | Fuch's heterochromic iridocyclitis<br>Intermediate uveitis/pars planitis | Idiopathic<br>MS, Behçet's, sarcoidosis, lymphoma, syphilis, TINU |
| Unifocal chorio-retinitis | Toxoplasmosis, candida, toxocara | Sarcoidosis, candida |
| Multifocal chorio-retinitis | No inflammation: PIC POHS<br>Min inflammation: AMPPE, MEWDS<br>Inflammation: Sympathetic, birdshot, infections | Idiopathic, sarcoidosis, Behçet's, lymphoma, syphilis, infections |
| Confluent chorio-retinitis | Viral retinitis (CMV,HSV, VZV)<br>Serpiginous, fungal retinitis | Immunosupression/AIDS (+ viral retinitis), sarcoidosis, lymphoma |
| Retinal vasculitis | Associated with other uveitis (especially infection), Eale's disease | Sarcoidosis, Behçet's, MS, TB, rarely other systemic vasculitis |
| Optic neuritis/papillitis | Isolated (non MS related) optic neuritis, Leber's stellate neuroretinitis, toxo, CMV | MS, syphilis, cat scratch, VKH, Lyme disease |
| Serous retinal detachment | Posterior scleritis<br>Steroid induced CSR | VKH, sympathetic ophthalmia |

# Important concepts in making a diagnosis in posterior uveitis

- Limited number of diagnostic possibilities for each type of posterior uveitis and a limited number of systemic diseases seen commonly in posterior uveitis
- Most posterior uveitis is **not** associated with systemic disease
- A good history is more important than untargeted investigation
- Many more mistakes are made in the treatment of uveitis than in its diagnosis
- The underlying diagnosis **rarely** guides treatment

- Handful of important systemic diagnoses
  - Sarcoidosis
  - Behcets
  - MS
  - Syphilis
  - Oculo-cerebral lymphoma

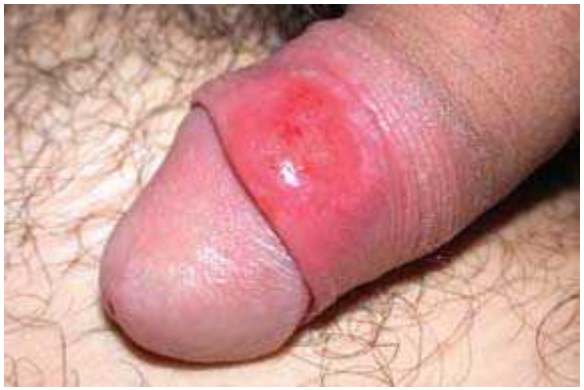
# Behcet's



- Recurrent oral ulceration plus
- Two of
  - Genital ulceration
  - Uveitis
  - Skin lesions
  - pathergy

# Syphilis

| Stage | Clinical features |
|---|---|
| Primary | Chancre |
| Secondary | Skin<br>Liver<br>Joints<br>Eyes (2.5-5%) |
| Latent | |
| Tertiary | Neuro-syphilis<br>Cardiac |



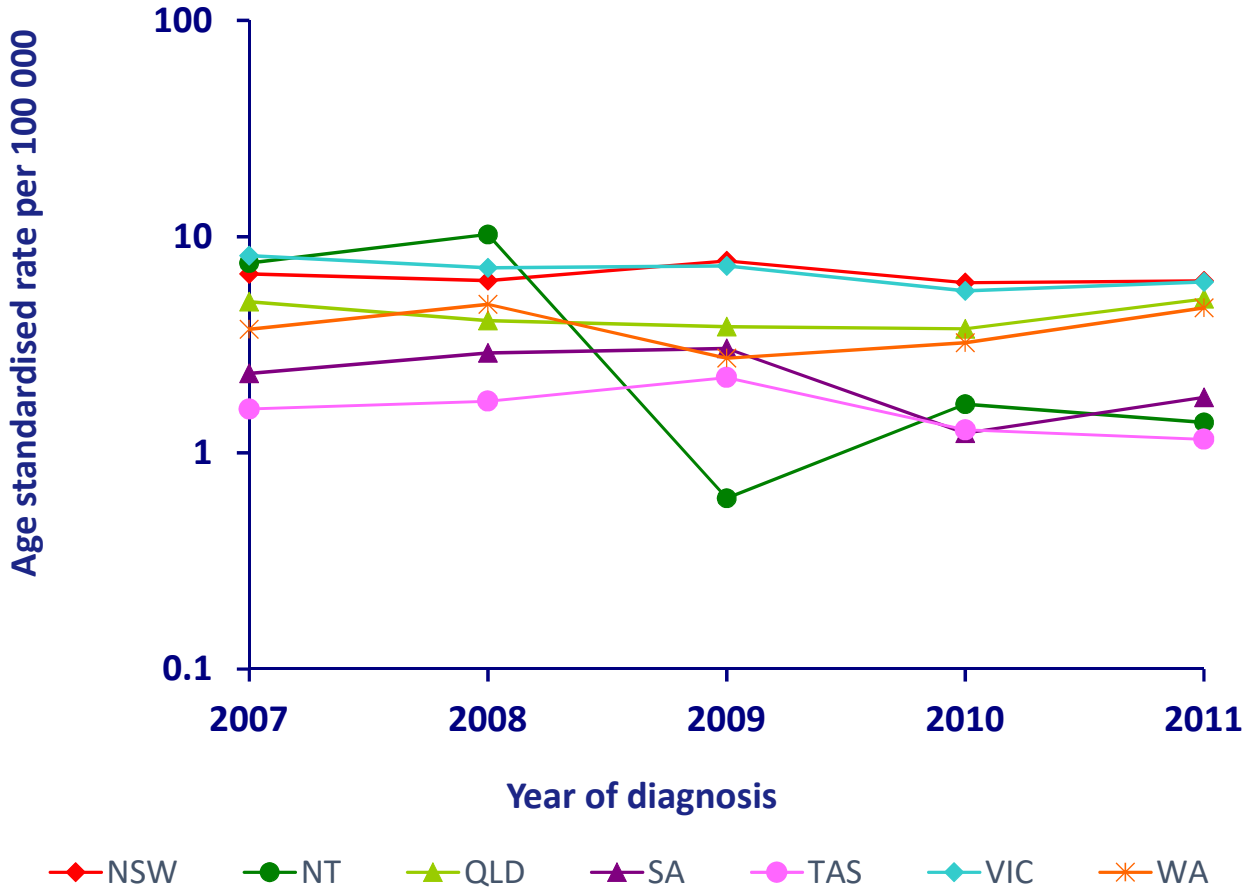
# Ocular syphilis

- 

| | Anterior uveitis | Posterior Uveitis | Panuveitis | Total patients |
|---|---|---|---|---|
| HIV positive | 27 (96.4%) | 48 (60.8%) | 18 (50%) | 93 (65%) |
| HIV negative | 1 (3.6%) | 31 (39.2%) | 18 (50%) | 50 (35%) |
| Total | 28 (20%) | 79 (55%) | 36 (25%) | 143 |

Amaratunge BC Camuglia JE Hall AJ **Syphilitic Uveitis: A review of clinical manifestations and treatment outcomes of syphilitic uveitis in HIV positive and HIV negative patients. Clin Exp Ophth in press**

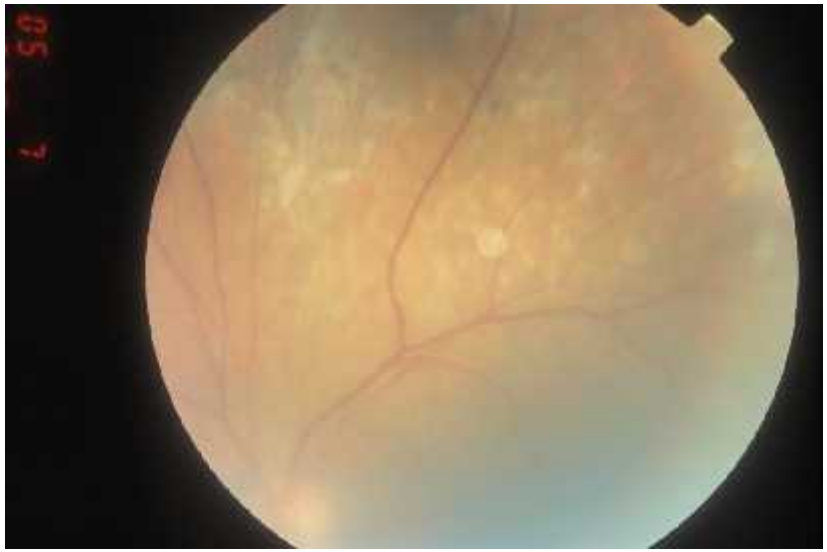
# Infectious syphilis, 2007 – 2011, by State/Territory and year




Source: National Notifiable Diseases Surveillance System

# Primary ocular lymphoma

- Elderly patients (>50 yo)
- Persistent and progressive and treatment resistant uveitis with:
  - Prominent vitritis
  - +/- multifocal choroiditis, retinitis, retinal vasculitis



# Diagnosis

- Systemic
  - MRI
  - LP
  - Brain biopsy
- Ocular
  - Vit biopsy
  - +/- retinal biopsy

**Intraocular lymphoma: a series of 14 patients with clinicopathological features and treatment outcomes.**
Hoffman PM, McKelvie P, Hall AJ, Stawell RJ, Santamaria JD.
Eye 2003 May;17(4):513-21.

# Ocular history taking in uveitis

- Trauma/surgery
  - Infection/SO
- Persistent unilaterality
  - infection

# Systemic history taking in uveitis

| System | Possible diseases |
|---|---|
| Joints | B27, Sarcoid, RA, Lupus… |
| Respiratory | Sarcoid, GPA, TB, … |
| GIT | IBD, Behcet's, Whipples… |
| GUT | Behcet's, Reactive arthritis, GPA, syphilis, TINU… |
| Skin | Ps arthritis, syphilis, Sarcoid, SLE… |
| Mouth | Behcet’s, HSV… |
| CNS | Lymphoma, MS, neuro-Behcets, infections, VKH… |
| Drugs | Bisphosphonates, Rifabutin, vaccines, tattoos. |

# Investigation in uveitis

- Limited untargeted investigations for when there are no specific features to suggest a diagnosis
  - FBE, U&E, LFT, RBG, ACE, CXR, VDRL
- More intensive/expensive/ invasive investigations only when clinically indicated

| Underlying disease | Useful investigations |
|---|---|
| Sarcoidosis | Most useful: ACE, CR, HRCT chest, targeted biopsy, BAL<br>Less useful: Gallium scan, blind biopsy |
| Systemic vasculitis | ANA, ENA, ANCA, Rh Factor, MSU, HLA A29, Hep serology |
| Birdshot | HLA A29 |
| VKH | LP |
| MS | MRI and LP |
| Primary oculo-cerebral lymphoma | MRI brain, LP, brain biopsy, vitrectomy +/- retinal biopsy |
| Ocular syphilis (usually 2ry) | RPR, TP IgG |
| Toxocara | Toxocara serology |
| Lyme disease | Borelia serology |
| Cat scratch disease | Bartonella serology |
| Brucellosis | Brucella serology |
| Viral retinitis | Most useful: AC or vitreous PCR, systemic viral load<br>Consider HIV serology/CD4 count<br>Less useful: viral serology (may be useful if negative) |
| TB | Most useful: IFN gamma release assay (Quantiferon), CXR<br>Less useful: mantoux |
| West Nile | West Nile virus serology |
| Metastatic endophthalmitis | Blood cultures, culture site of suspected primary infection eg catheter tips, central lines, chest, urine, etc. Vitreous tap |

# Summary

- Make a descriptive diagnosis of the inflammation
- Determine clinically if possible the cause of the patients visual loss
- Take a targeted history

# Summary

- Investigate aggressively any features identified on history
- If no clues on history then a limited number of investigations targeting the likely causes and possible side effects of treatment

Thank you