Dr Mei Tan
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Specialist Eye Surgeons- Jacana, Moonee Ponds.
Royal Melbourne Hospital.
Royal Victorian Eye and Ear Hospital.
Senior Clinical lecturer, University of Melbourne
RS, 68 year old lady

Referred with reduced vision in right eye following cataract surgery.
No improvement despite 8 weeks treatment with Prednefrin Forte and Acular.

BCVA: 6/18 RE 6/12 LE
Corneas: clear
AC: deep and quiet OU
Lens: PCIOL RE Grade 2 NS LE
Fundus: Thickening of macula RE; moderate epiretinal membrane LE
OCT and FFA
Diagnosis: pseudophakic cystoid macular oedema
Treatment: orbital floor kenocort RE
Macular oedema improved, BCVA 6/9 RE

At review 2 months later, c/o slight deterioration of vision in RE

<table>
<thead>
<tr>
<th>6/12</th>
<th>6/12</th>
</tr>
</thead>
<tbody>
<tr>
<td>Fine stellate KPS</td>
<td>Clear cornea</td>
</tr>
<tr>
<td>Trace cell in AC</td>
<td>Quiet and deep AC</td>
</tr>
<tr>
<td>IOP 27 mmHg</td>
<td>IOP 11 mmHg</td>
</tr>
<tr>
<td>PCIOL clear and centered</td>
<td>Grade 2 NS</td>
</tr>
<tr>
<td>Quiet vitreous</td>
<td>Quiet vitreous</td>
</tr>
<tr>
<td>Few peripheral intraretinal haemorrhages,</td>
<td>Stable epiretinal membrane</td>
</tr>
<tr>
<td>Patch of granular retinal whitening</td>
<td></td>
</tr>
<tr>
<td>Paucity of retinal vessels peripheral in</td>
<td></td>
</tr>
<tr>
<td>retina</td>
<td></td>
</tr>
</tbody>
</table>
Blood investigation:
FBC, dWCC, ESR, CRP, ACE, ANA, syphillis serology, HSV VZV and CMV serology
Aqueous tap for viral PCR

Virus serology :
Negative for CMV and HSV
IgG positive for VZV
Aqueous tap : **positive for CMV**

Admitted to Royal Mebourne Hospital:
Further vitreous biopsy – also positive for CMV
Intravitreal foscarnet at Royal Melbourne Hospital
Oral Valganciclovir.

Further history from family........
Autoimmune hepatitis – on long term prednisolone and methotrexate past 5 years!
Low CD4 count – under investigation
HIV serology negative
2 months later…..
• At review, still on oral valganciclovir
• Area of retinitis improved
• But developed inferior nasal retinal detachment in right eye
• Underwent right vitrectomy, endolaser and silicone oil tamponade
• Macular oedema recurred 3 months after surgery under silicone oil……
Introduction

_Uveitis_ is broadly defined as inflammation (i.e., _-itis_) of the uvea (from the Latin _uva_, meaning "grape")

Thorough history and review of systems is the essential first step

Thorough ocular examination and pertinent organ systems

Determine the anatomical classification and subcategorize

Laboratory investigations

Treatment
## Table 5-1 The SUN Working Group Anatomical Classification of Uveitis

<table>
<thead>
<tr>
<th>Type</th>
<th>Primary Site of Inflammation</th>
<th>Includes</th>
</tr>
</thead>
<tbody>
<tr>
<td>Anterior uveitis</td>
<td>Anterior chamber</td>
<td>Iritis</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Iridocyclitis</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Anterior cyclitis</td>
</tr>
<tr>
<td>Intermediate uveitis</td>
<td>Vitreous</td>
<td>Pars planitis</td>
</tr>
<tr>
<td>Posterior uveitis</td>
<td>Retina or choroid</td>
<td>Posterior cyclitis</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Hyalitis</td>
</tr>
<tr>
<td>Panuveitis</td>
<td>Anterior chamber, vitreous,</td>
<td>Focal, multifocal, or diffuse choroiditis</td>
</tr>
<tr>
<td></td>
<td>and retina or choroid</td>
<td>Chorioretinitis</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Retinochoroiditis</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Retinitis</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Neuroretinitis</td>
</tr>
</tbody>
</table>

## Classification

### Table 5-2: The SUN Working Group Descriptors in Uveitis

<table>
<thead>
<tr>
<th>Category</th>
<th>Descriptor</th>
<th>Comment</th>
</tr>
</thead>
<tbody>
<tr>
<td>Onset</td>
<td>Sudden</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Insidious</td>
<td></td>
</tr>
<tr>
<td>Duration</td>
<td>Limited</td>
<td>≤3 months’ duration</td>
</tr>
<tr>
<td></td>
<td>Persistent</td>
<td>&gt;3 months’ duration</td>
</tr>
<tr>
<td>Course</td>
<td>Acute</td>
<td>Episode characterized by sudden onset and limited duration</td>
</tr>
<tr>
<td></td>
<td>Recurrent</td>
<td>Repeated episodes separated by periods of inactivity without treatment ≥3 months’ duration</td>
</tr>
<tr>
<td></td>
<td>Chronic</td>
<td>Persistent uveitis with relapse in &lt;3 months after discontinuing treatment</td>
</tr>
</tbody>
</table>

Epidemiology

![Epidemiology Bar Chart]

- **Anterior**: 15% Infectious, 80% Non-infectious
- **Intermediate**: 1% Infectious, 88.8% Non-infectious
- **Posterior**: 38.8% Infectious, 57.1% Non-infectious
- **Panuveitis**: 10.8% Infectious, 66.7% Non-infectious
Anterior Uveitis

Prevalence

Anterior uveitis is the most common form of intraocular inflammatory disease – accounts for 75-90% of all cases of uveitis.

Represents a wide spectrum of disease- may be isolated, part of a panuveitis, or part of a systemic disease.

Incidence of 8-12 per 100,000 population per year

Risk factors :
Associated systemic conditions
Surgery
Blunt trauma
Previous history
Idiopathic AAU

- 50% of patients with AAU, occurs in isolation
- Affects any age, biphasic peaking at 30y and 60y.
- Affects both sexes equally
- Almost always unilateral, but may affect both eyes sequentially
- Recurrences common, rarely may become persistent.
Anterior Uveitis

Symptoms:
• Acute
  • Redness
  • Pain
  • Photophobia
  • Excessive tearing
  • Decreased vision

• Chronic
  • No symptoms
  • Decreased vision due to band keratopathy, CMO or cataract
  • Exacerbations and remissions with few acute symptoms
Rubeosis iridis
Peripheral anterior synechiae
Pigment dispersion

Cells and flare in AC
Pupillary miosis
Fibrin
Hypopyon
Spillover cells in anterior vitreous
IOP may be elevated
Anterior Uveitis

Circumlimbal injection
Anterior Uveitis
Anterior Uveitis
Anterior Uveitis
Anterior Uveitis

Medium to small KPs

Large mutton fat KPs in patient with sarcoidosis
Anterior Uveitis

Keratic precipitates:
- Fine KP ("Stellate"; typically covers entire corneal endothelium):
  - Herpetic
  - FHIC
  - CMV retinitis
- Small, non-granulomatous KP:
  - HLA B27 associated uveitis
  - Trauma
  - Masquerade
  - JIA
  - Posner-Schlossman syndrome
- Granulomatous KP (large, greasy mutton fat; mostly on inferior cornea):
  - Sarcoidosis
  - Syphilis
  - Tuberculosis
  - Sympathetic ophthalmia
  - VKH
Anterior Uveitis

Other signs:
- Low IOP
- Elevated IOP: Herpetic, lens-induced, FHIC, Hypertensive uveitis
- Fibrin: HLA B27, endophthalmitis
- Hypopyon: HLA B27, Behcet disease, infectious endophthalmitis, rifabutin, tumor
- Iris nodules: Sarcoidosis, syphilis, tuberculosis
- Iris atrophy: herpetic
- Iris heterochromia: FHIC
- Iris synechiae: HLA B27, sarcoidosis
- BSK: JIA, any chronic uveitis in older

**Uveitis in quiet eye: consider JIA, FHIC, masquerade syndromes**
Anterior Uveitis

Management:

• All patients - check IOP, esp if raised needs urgent referral

• Acute episode, previously undiagnosed – urgent referral to eye specialist or hospital.

• Recurrent acute episode – topical treatment may be initiated with urgent referral to eye specialist or hospital

• Chronic AAU – early referral if not being managed or under medical care
Anterior Uveitis

Why test.....

• Not all uveitis is inflammatory – infections and masquerades.
• Diagnosis predicts natural history, response to therapy, and associated disorders.
• Knowing is better than not.....

When not to test?

• Simple anterior uveitis.
• Diagnosis known
When to test?

- Idiopathic AAU constitutes approx 50% of cases- significant proportion of patients will require investigation.
- Anything other than simple anterior uveitis
  - Recurrent
  - Severe
  - Chronic
  - Granulomatous
  - Suggestive signs or symptoms
  - Not responsive to initial corticosteroid therapy
Anterior Uveitis

Suggestive signs or symptoms
• History
• Review of systems
  • Constitutional – fever, night sweats, weight loss (TB)
  • Rheumatologic – joint pain or swelling (HLA-B27)
  • Dermatologic – rash, alopecia (syphilis)
  • Neurologic – headache, tinnitus, hearing loss (VKH)
  • Gastroenterologic – frequency, diarrhoea, blood (IBD)
  • Urological – pain, frequency, ulcer (Reiter’s)
  • Respiratory – cough, SOB (sarcoidosis)
Anterior Uveitis

- Investigations at baseline includes FBC, ESR, CRP, ACE, ANA, syphilis serology, quantiferon gold. Chest X-ray, Urinalysis.

- Selective investigations - ANCA, Toxoplasma serology, toxocara ELISA, HLA B27, HLA A29, HLA B51, Borrelia serology.

- Conjunctival biopsy, PCR of intraocular fluid from AC tap, vitreous biopsy, choroidal biopsy.

- Lumbar puncture, MRI
Anterior Uveitis

HLA-B27 associated AAU

• Up to 50% of patients with AAU are HLA-B27 positive
• Peak at 30 y of age
• Commoner in males and positive family history
• Associated with ankylosing spondylitis, Reiter’s disease, psoriasis or inflammatory bowel disease
• Almost always unilateral but may affect both eyes sequentially
• Often more severe and recurrences more frequent than idiopathic AAU
<table>
<thead>
<tr>
<th></th>
<th>HLA-B27 positive</th>
<th>HLA-B27 negative</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age at onset (y)</td>
<td>32–35</td>
<td>39–48</td>
</tr>
<tr>
<td>Gender</td>
<td>♂:♀ 1.5–2.5:1</td>
<td>1:1</td>
</tr>
<tr>
<td>Eye involvement</td>
<td>Unilateral 48–59%</td>
<td>Bilateral 21–64%</td>
</tr>
<tr>
<td></td>
<td>Alternating 29–36%</td>
<td></td>
</tr>
<tr>
<td>Pattern of uveitis</td>
<td>Acute in 80–87%</td>
<td>Chronic in 43–61%</td>
</tr>
<tr>
<td>Recurrence</td>
<td>Frequent</td>
<td>Uncommon</td>
</tr>
<tr>
<td>KP</td>
<td>Mutton fat KP in 0–3%</td>
<td>Mutton fat KP in 17–46%</td>
</tr>
<tr>
<td>Fibrin in AC</td>
<td>25–56%</td>
<td>0–10%</td>
</tr>
<tr>
<td>Hypopyon</td>
<td>12–15%</td>
<td>0–2%</td>
</tr>
<tr>
<td>Associated systemic disease</td>
<td>48–84%</td>
<td>1–13%</td>
</tr>
<tr>
<td>FH</td>
<td>Yes</td>
<td>No</td>
</tr>
<tr>
<td>PS</td>
<td>40.4%</td>
<td>18.7%</td>
</tr>
<tr>
<td>Cataract</td>
<td>12.9%</td>
<td>13.6%</td>
</tr>
<tr>
<td>OHT</td>
<td>11.4%</td>
<td>11.4%</td>
</tr>
<tr>
<td>Glaucoma</td>
<td>4.4%</td>
<td>6.6%</td>
</tr>
<tr>
<td>CMO</td>
<td>11.7%</td>
<td>1.0%</td>
</tr>
</tbody>
</table>

Table published in Albert & Jakobiec's Principles and Practice of Ophthalmology (Third Edition) by Albert and Miller, p 1139. Copyright Elsevier; reproduced by kind permission.
Topical treatment is mainstay of management.

Topical corticosteroid therapy
- Prednefrin Forte, Maxidex, betnesol ointment
- ½ hourly - 1 hourly depending on severity; reassess within 1 week.
- Titrate according to disease activity
- Reduce inflammation, reduce capillary permeability and exudate leakage
- Beware of steroid responders.

Cycloplegia
- Atropine, cyclopentolate
- Relieves pain and ciliary spasm
- Prevents posterior synechiae
Anterior Uveitis

Treatment

• Subconjunctival injection of betamethasone and atropine, or triamcinolone may be needed in severe cases.

• If not improving after 48 hours of half-hourly drops, may require consideration of systemic steroids.

Systemic NSAID (aspirin, ibuprofen, voltaren)

Systemic steroid therapy
A 6-year-old girl presented to her GP with a red left eye.

No pain or irritation. Blurring of vision in the affected eye.

Onset of neck stiffness with limitation in neck flexion and rotation. Worse in the mornings.

No associated fevers, rashes, gastrointestinal or genitourinary symptoms.

She had been well in the past, no medications, and her vaccinations were up to date.

Her mother had type 1 diabetes mellitus. A maternal aunt had been diagnosed with rheumatoid arthritis 4 years earlier.

mild restriction in the range of neck rotation and flexion.

GP diagnosed pt with conjunctivitis and intermittently prescribed chloramphenicol eye drops over 3 months with no improvement.

Referred to ophthalmologist.
• VA RE 6/7.5 ; LE 6/12
• Band keratopathy present in the region of the visual axis
• Posterior synechiae. Cells 1+
• The child’s vision continued to deteriorate to 6/24 in the left eye despite treatment with topical dexamethasone 0.1% and atropine 1%. She was referred to the rheumatology service.
• Rheumatology – bloods investigations
• Elevated erythrocyte sedimentation rate of 43 mm/hour (normal range, 0–20 mm/hour),
• C-reactive protein concentration was within normal limits at 4 mg/L (normal range, 0–10 mg/L).
• The ANA titer was strongly positive at over 1:2560.

• Treatment with non-steroidal anti-inflammatory drugs resulted in little improvement, so therapy with oral prednisolone and pulse methylprednisolone was commenced.
JUVENILE IDIOPATHIC ARTHRITIS

• Most common cause of arthritis (and uveitis) in children

• Prevalence 1/1000 children in USA (M:F 1:3)

• 80% of childhood uveitis (typically anterior)

• Greatest risk of uveitis in first 2 years after diagnosis (risk falls off dramatically at 8 y)
UVEITIS IN CHILDREN

• Overall prevalence 30/100,000 in North America and Europe

• Incidence of uveitis increases with age in the childhood population (particularly acute uveitis)

• Girls affected more than boys, likely due to the gender predilection of JIA

• Approximate anatomic distribution:
  • Anterior 30%
  • Intermediate 20%
  • Posterior 40%
  • Panuveitis 10%
UVEITIS IN CHILDREN

• Most common causes of endogenous uveitis in children:
  
  • JIA (ant.) - most common identifiable cause (most cases of chronic anterior uveitis in children are idiopathic)
  
  • Post-traumatic uveitis

• Less commonly:
  
  • HLA-B27 associated uveitis (ant.)
    ▪ Ankylosing spondylitis
    ▪ Reiter’s syndrome
    ▪ IBD
    ▪ Psoriatic arthritis
  
  • Fuchs’ uveitis syndrome (ant.)
  
  • Behcet’s disease (pan)
  
  • VKH (pan)
  
  • Sarcoid (any)
  
  • Sympathetic ophthalmia
  
  • Infectious
    ▪ HSV, VZV (ant.)
    ▪ Toxoplasmic retinochoroiditis (post.)
Complications of uveitis in children

- Band keratopathy (may lead to amblyopia)
- Secondary glaucoma - 42% of JIA pt.
- Posterior synechiae
- Cataract (steroids / inflam.) - 18% of JIA pt.
- Inflammatory membranes (ciliary membranes may lead to detachment)
- Macular edema
- Hypotony
- Phthisis
Four major complications exist:

• Cataract
• Secondary glaucoma
• Band keratopathy
• Cystoid macula oedema

Check patient with associated systemic symptoms for underlying conditions which may be asymptomatic.

Bilateral involvement, recurrent acute episodes X3 - warrants a full systemic workup for associated systemic disease.
Intermediate Uveitis

• Accounts for around 10% of all cases of uveitis.

• Bimodal; commonest in young adults and middle-age/elderly.

• Both sexes equally affected.

• Bilateral in 80%, but often asymptomatic.

• Investigations in all patients – FBC, U+E, ESR, syphilis serology, urinalysis, CXR.

• OCT/FFA for CMO.
Intermediate Uveitis

Symptoms:
Painless floaters
DOV
Minimal photophobia
Minimal external inflammation
Usually, 15 to 40 years of age
Bilateral

Signs:
Vitreous inflammatory cells
Snowball opacities
Snowbank
Vitreal strands
Chronic uveitis may be associated with cyclitic membrane formation, secondary CB detachment and hypotony
Intermediate Uveitis

Complications:
- CMD
- Cataract
- Secondary glaucoma
- Cyclitic membrane
- TRD
- ERM
- VMT
- Retinal neovascularization
# Intermediate Uveitis

## Table 11.11 Associations of intermediate uveitis

<table>
<thead>
<tr>
<th>Group</th>
<th>Cause</th>
<th>Consider</th>
</tr>
</thead>
<tbody>
<tr>
<td>1° ocular</td>
<td>Idiopathic/pars planitis</td>
<td>After exclusion of other associations&lt;br&gt;&gt;70%</td>
</tr>
<tr>
<td>2° systemic</td>
<td>MS</td>
<td>MRI brain, LP</td>
</tr>
<tr>
<td></td>
<td>Sarcoïd</td>
<td>ACE, Ca, CXR, CT thorax</td>
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<tr>
<td></td>
<td>IBD</td>
<td>Bowel studies, biopsy</td>
</tr>
<tr>
<td></td>
<td>CNS/intraocular lymphoma</td>
<td>MRI brain, LP</td>
</tr>
<tr>
<td>2° infective</td>
<td>Toxocara</td>
<td>Serology</td>
</tr>
<tr>
<td></td>
<td>Lyme disease</td>
<td>Serology</td>
</tr>
<tr>
<td></td>
<td>HTLV-1</td>
<td>Serology</td>
</tr>
</tbody>
</table>
Intermediate Uveitis

Management:

- Topical corticosteroid with or without mydriatics – significant AC activity.
- Orbital floor/sub-Tenon steroid injection – triamcinolone acetonide.
- Intravitreal triamcinolone injection – risk of elevated IOP, cataract
- Sustained-release devices – risk of cataract and elevated IOP
  - Ozurdex (dexamethasone implant)
  - Retisert (fluocinolone acetonide)
- Systemic treatment
  - Corticosteroids – oral prednisolone
  - Immunosuppressants – reserved for bilateral or resistant disease, intolerable steroid side effects, inability to reduce steroid dose.
  - Biologics – anti-TNF (contraindicated in patients with MS.)
Intermediate Uveitis

Management:

• Surgical – management of complications and sequelae.
  • Cataract extraction with lens implant
  • Vitrectomy
    • for vitreous opacities
    • CMO
    • vitreomacular traction
    • ERM
    • RD
    • can be combined with cataract extraction with lens implant.
• Glaucoma surgery
Posterior Uveitis

Symptoms:
Blurred vision
Floaters

Pain, redness and photophobia typically absent (unless anterior segment inflammation)

Signs:
Retinal or choroidal inflammatory infiltrates
Inflammatory sheathing of arteries or veins
Exudative, tractional or rhegmatogenous RD
RPE hypertrophy or atrophy
Atrophy or swelling of retina, choroid or optic nerve head
Pre-retinal or subretinal fibrosis
Retinal or choroidal neovascularization
Posterior Uveitis

• The front of the eye is richly innervated with pain fibers. Therefore common symptoms from the “anterior segment” of the eye include pain, redness, and discharge.

• These symptoms have no relevance to infection/inflammation in the retina.

• Symptoms of disease in the retina include scotoma, flashes and floaters.

• Also, if one eye has normal vision the brain will favor the good eye and the patient may not complain of any symptoms.
Posterior Uveitis

- **Infectious**
  - Viral – HSV, VZV, CMV, WNV, EBV, HTLV-1
  - Bacterial – TB, syphillis, Borrelia (Lyme disease), Bartonella (cat-scratch), Brucella, leptospira
  - Parasitic – Toxoplasma, toxocara, nematodes, onchocerciasis (river blindness)
  - Fungal – candida, aspergillus, histoplasma.

- **Non infectious**
  - Idiopathic
  - Sarcoidosis
  - White dot syndromes – MEWDS, Birdshot, APMPPE
  - VKH
  - Behcet’s disease

- **Masquerades**
  - Intraocular lymphoma
  - Leukaemia
  - Retinoblastoma
  - Intraocular FB
  - Amyloidosis
  - Sympathetic ophthalmia
Necrotizing Herpetic Retinopathy

- Usually caused by the varicella zoster virus (VZV) and less commonly by other herpes virus’, HSV 1 and 2.
- Visually devastating in most cases.
- Typical setting in an elderly/immunosuppressed patient with HZO (shingles affecting the ophthalmic branch of trigeminal nerve).
- 2 Clinical presentations
  - Acute retinal necrosis (ARN)
  - Posterior outer retinal necrosis (PORN)
- Investigations
  - AC tap +/- vitreous biopsy with PCR to identify viral DNA
ARN

- Rare, incidence 1 per 2 million population per year
- Healthy and immunocompromised pts
- VZV commonest cause
- Usually unilateral
- Predominantly peripheral disease characterised by occlusive arteritis, necrotizing retinitis
- Marked vitiritis and AC activity, some pain
- **Second eye involvement in 30%**
PORNO

- Rare

- Only in the context of immunosuppression (HIV, CD4+ T cell < 50/mm3)

- Unilateral or bilateral

- Painless, rapid VA

- Starts centrally, early macular involvement

- Rapidly coalescing white areas of outer retinal necrosis

- Minimal vitritis, vasculitis or retinitis.
## ARN vs PORN

<table>
<thead>
<tr>
<th></th>
<th>ARN</th>
<th>PORN</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Appearance</strong></td>
<td>One or more foci of full-thickness retinal necrosis with discrete borders</td>
<td>Multiple foci of deep retinal opacification which may be confluent</td>
</tr>
<tr>
<td><strong>Location</strong></td>
<td>Peripheral retina (usually adjacent/outside temporal arcades)</td>
<td>Peripheral retina</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Macular involvement</td>
</tr>
<tr>
<td><strong>Progression</strong></td>
<td>Rapid (but usually responds to treatment)</td>
<td>Extremely rapid</td>
</tr>
<tr>
<td><strong>Direction</strong></td>
<td>Circumferential</td>
<td>No consistent direction</td>
</tr>
<tr>
<td><strong>Vessels</strong></td>
<td>Occlusive vasculopathy (arterial)</td>
<td>No vascular inflammation</td>
</tr>
<tr>
<td><strong>Inflammation</strong></td>
<td>Prominent AC and vitreous inflammation</td>
<td>Minimal or none</td>
</tr>
<tr>
<td><strong>Suggestive features</strong></td>
<td>Optic neuropathy/atrophy Scleritis Pain</td>
<td>Perivascular clearing of retinal opacification</td>
</tr>
</tbody>
</table>
Treatment

- Intravitreal foscarnet/ganciclovir as initial treatment (threatening optic n. or fovea)

- Intravenous aciclovir followed by oral aciclovir for 12 weeks; oral valaciclovir for 8-12 weeks.

- Systemic corticosteroids (inflammation)

- Aspirin (arterial occlusion)

- Extremely high rates of retinal detachment – vitrectomy, endolaser with silicone oil tamponade.

- In HIV patients – need to coordinate management with HIV physician.
Toxoplasmosis retinochoroiditis

- Acute primary infection – characterized by focal white chorioretinitis

- Secondary toxoplasmosis is characterized by similar focal white chorioretinitis + an adjacent pigmented scar (site of prior primary infection)

- Human infection - contact with cat faeces/contaminated soil/contaminated water, ingestion of undercooked meat, transplacentally.

- Primarily clinical diagnosis, PCR of intraocular samples

- Treatment – prednisolone with clindamycin, sulfadiazine or co-trimoxazole
A.P. : a case of primary toxoplasmosis

Dense retinal whitening with no adjacent pigmented scar. A case of toxo that might be mistaken for syphilis
A.P., an example of “secondary” toxoplasmosis

- Pigmented scar from acute episode 12/07
- New focus of acute dense white retinal necrosis with overlying vitritis.
Ocular Syphilis

- Infection with spirochaete Treponema pallidium
- Transmitted by sexual contact or transplacentally.
- May occur during secondary and tertiary stages.
- Bilateral in 50%
- Increasing in incidence
- Anterior uveitis is commonest
  - Variable severity
  - Roseolae (vascular fronds on iris)
  - Iris nodules
  - Granulomatous or non-granulomatous
Posterior syphilitic uveitis

- Variable presentation
- Unilateral or bilateral
- Uni- or multifocal
- Choroiditis or chorioretinitis
- Retinal vasculitis
- Neuroretinitis
- Serous retinal detachment

Rx – high dose penicillin
Sarcoidosis

- Relatively common, affects 0.1% of population. Cause unknown.
- Granulomatous multisystem disorder, may be life-threatening.
- Commoner in females, African Caribbeans, Irish, Scandinavians.
- Eye is affected in 25% of patients:
  - Anterior uveitis in 60%
  - Posterior segment disease in 25%
Sarcoidosis

Variety of posterior segment lesions in sarcoidosis

Treatment: systemic corticosteroids +/- immunosuppressives. New roles for biologics eg. anti-TNF
White dot syndromes

- A group of disorders characterized by multiple whitish-yellow inflammatory lesions located at the level of the outer retina, retinal pigment epithelium, and choroid

- Frequently includes anything that gives white ‘dots’ in the fundus

- They present important diagnostic and therapeutic challenges

- History and examination findings are extremely important

- Look for the clues
White dot syndromes

Associated features

- Ocular
  - Uveitis (presence or absence of)
  - Vitreous cells/abnormalities
  - Retinal vascular changes
  - Macular changes
  - Disc changes

- Non-ocular
  - CNS signs and symptoms
### White dots syndrome

<table>
<thead>
<tr>
<th>Syndrome</th>
<th>Age</th>
<th>Sex</th>
<th>Laterality</th>
<th>Vitritis</th>
<th>Lesion size</th>
<th>Prognosis</th>
</tr>
</thead>
<tbody>
<tr>
<td>PIC</td>
<td>20–40</td>
<td>&gt;♀♂</td>
<td>Bilateral</td>
<td>−</td>
<td>1/10DD</td>
<td>Guarded</td>
</tr>
<tr>
<td>POHS</td>
<td>20–50</td>
<td>=♀♂</td>
<td>Bilateral</td>
<td>−</td>
<td>1/3DD</td>
<td>Guarded</td>
</tr>
<tr>
<td>MEWDS</td>
<td>20–40</td>
<td>&gt;♂♀</td>
<td>Unilateral</td>
<td>+</td>
<td>1/5DD</td>
<td>Good</td>
</tr>
<tr>
<td>APMPPE</td>
<td>20–40</td>
<td>=♀♂</td>
<td>Bilateral</td>
<td>+</td>
<td>1DD</td>
<td>Good</td>
</tr>
<tr>
<td>Serpiginous choroidopathy</td>
<td>30–60</td>
<td>=♀♂</td>
<td>Bilateral</td>
<td>+</td>
<td></td>
<td>Poor</td>
</tr>
<tr>
<td>Birdshot chorioretinopathy</td>
<td>23–79</td>
<td>&gt;♀♂</td>
<td>Bilateral</td>
<td>++</td>
<td>1/4–1/2DD</td>
<td>Guarded</td>
</tr>
<tr>
<td>Multifocal choroiditis with panuveitis (MCP)</td>
<td>30–60</td>
<td>&gt;♀♂</td>
<td>Bilateral</td>
<td>++</td>
<td>1/10DD</td>
<td>Guarded</td>
</tr>
</tbody>
</table>
APMPPE

Birdshot chorioretinopathy

MEWDS

Multifocal choroiditis with panuveitis
Management

• Some conditions have good prognosis and spontaneous recovery can be expected within 2-3 months eg. APMPPE, MEWDS
• Corticosteroids +/- immunosuppressives in progressive conditions eg. Serpiginous, birdshot, MCP, PIC
• Intravitreal anti-VEGF for secondary CNVM
• Local ‘rescue’ steroid for CMO
Posterior uveitis therapeutics – a double edge sword

- Multitude of side effects and adverse effects from systemic treatment of posterior uveitis
- Corticosteroids – osteoporosis, peptic ulcer, diabetes, hypertension, psychosis
- Immunosuppressants – bone marrow suppression, secondary malignancies, hepatotoxicity, nephrotoxicity, sterility.
- Increased risk of infections with biologics
  - Anti-TNF – risk of TB, herpes zoster reactivation, CMV retinitis
- Monitoring of therapies is imperative
  - Regular BP, weight, BM, urinalysis
  - Regular monthly blood investigations – FBP, U+E, LFT
Posterior Uveitis

Take home message

• **Rule out infectious diseases**

• Prevent iatrogenic disease

• Monitor regularly

• Knowledge of immunology will allow better treatment