Principles of uveitis evaluation and therapy

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National Eye Institute
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Warren Grant Magnuson Clinical Center

2000
• 5 year old boy
• CC: difficulty reading
• Pediatrician did “color blind test”, said child was color blind but otherwise okay
• No ophthalmic consultation

November 2002
• 7 years old
• CC: “2 spots in right eye”
• Retinal specialist: OD 20/30, no inflammation, macular ERM, peripheral intraretinal fibrotic lesion 5x5x2 mm
• “Inactive toxocara” vs. regressed retinoblastoma vs. astrocytoma
• Follow every 3 months

November 2002-October 2003
• Steady decline in vision and development of inflammation
• PPD, HIV, FTA, toxocara ab--negative per history
• No treatment
November 4, 2003: NEI

- POHx: unremarkable
- PMHx: cleft tongue surgery at age 2
- FamHX: unremarkable
- SocHX: no well water, livestock, unpasteurized milk, TB exposures, overseas travel, or tick bites. Sandbox at home. Races dirt bikes.
- Pets: cats, dogs, frogs, iguana, python, bearded dragon
- ROS: negative

November 4, 2003: NEI

- OD 20/200 (laser acuity 20/150), OS 20/16
- IOP OD 10, OS 12
- 1.5 log unit right RAPD
- Ishihara OD 0/14, OS 3/14
- SLE: OD trace AC cell, otherwise anterior segment normal OU
- Gonioscopy unremarkable OU
Chronic unilateral granulomatous posterior uveitis

- Toxocara canis, toxocara cati
- Sarcoidosis
- Tuberculosis
- Syphilis
- Bartonella
- Masquerade syndrome
  - Regressed retinoblastoma
  - Astrocytoma

• Chem 20
• CBC
• Bartonella antibody
• Lyme antibody
• Toxoplasma antibody
• FTA

• ACE 82 u/L (0-52)

• EUA: no other lesions in right eye. Aqueous fluid aspirate, could not aspirate vitreous fluid

• MRI brain: arachnoid cyst right middle cranial fossa with commensurate hypoplasia of the anterior temporal lobe

• CXR: normal

Aqueous fluid

• IL-1 alpha
• IL-1 beta
• IL-6
• TNF alpha
• Below detectable levels

• Cytology: cellular debris, non-diagnostic
Some help from the CDC…

• Serum ELISA 1:128
• Aqueous fluid ELISA 1:256

“McDonald’s protocol”

• Albendazole 400 mg po bid x1 week, off x1 week, then repeat Albendazole for 1 week
• French fries, Happy Meal®
• Prednisone 30 mg po q day

Uveitis epidemiology

• 10% of the visual handicap in the Western World
• 30,000 new cases of legal blindness in the United States per year
• 4th leading cause of blindness in U.S.
Uveitis impact

- Vision
- Quality of life
- Socioeconomic

How uveitis decreases vision

- Cataract
- Glaucoma
- CME
- Fibrosis
- Ischemia
- Band K
- CNV
- Neovascularization
- Photoreceptor degeneration
Anatomic site

<table>
<thead>
<tr>
<th></th>
<th>Community-based</th>
<th>University-referral</th>
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</thead>
<tbody>
<tr>
<td>Anterior</td>
<td>90.6</td>
<td>60.6</td>
</tr>
<tr>
<td>Intermediate</td>
<td>1.4</td>
<td>12.2</td>
</tr>
<tr>
<td>Posterior</td>
<td>4.7</td>
<td>14.6</td>
</tr>
<tr>
<td>Panuveitis</td>
<td>1.4</td>
<td>9.4</td>
</tr>
</tbody>
</table>

McCannel et al., 1996

- 6 Kaiser medical centers in northern California
- 732,000 patients
### Historical data: incidence and prevalence

<table>
<thead>
<tr>
<th>Study</th>
<th>Location</th>
<th>Study Period</th>
<th>Size of Population</th>
<th>Incidence per 100,000 persons/year</th>
<th>Prevalence per 100,000 persons</th>
</tr>
</thead>
<tbody>
<tr>
<td>Darrel et al.</td>
<td>Rochester, Minnesota</td>
<td>1945-1954</td>
<td>23,988</td>
<td>17</td>
<td>264*</td>
</tr>
<tr>
<td>Gritz et al.</td>
<td>Northern California</td>
<td>1988</td>
<td>471,254</td>
<td>52.0</td>
<td></td>
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<tr>
<td>Friedman</td>
<td>Northwestern Finland</td>
<td>1969</td>
<td>613,426</td>
<td>75.4</td>
<td></td>
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<tr>
<td>Freedman</td>
<td>South Africa</td>
<td>1971-1973</td>
<td>652,259</td>
<td>19.6</td>
<td></td>
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<tr>
<td>Saari et al.</td>
<td>Southwestern Finland</td>
<td>1980-1982</td>
<td>459,515</td>
<td>22.6</td>
<td>75.4</td>
</tr>
<tr>
<td>Saari et al.</td>
<td>Northern California</td>
<td>1998-1999</td>
<td>731,895</td>
<td>52.0</td>
<td>119.3</td>
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</tbody>
</table>

*10 year prevalence

Adapted from Gritz et al., 2004

### Uveitis Incidence

<table>
<thead>
<tr>
<th>Category</th>
<th>All N=731,898</th>
<th>Male N=382 (52.4%)</th>
<th>Female N=350 (47.6%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Anterior</td>
<td>269 (36.8%)</td>
<td>128 (36.4%)</td>
<td>142 (37.4%)</td>
</tr>
<tr>
<td>Intermediate</td>
<td>11 (1.5%)</td>
<td>4 (1.1%)</td>
<td>7 (1.9%)</td>
</tr>
<tr>
<td>Posterior</td>
<td>8 (1.1%)</td>
<td>7 (2.0%)</td>
<td>3 (0.8%)</td>
</tr>
<tr>
<td>Panuveitis</td>
<td>19 (2.6%)</td>
<td>6 (1.7%)</td>
<td>13 (3.4%)</td>
</tr>
<tr>
<td>Indeterminate</td>
<td>72 (9.9%)</td>
<td>25 (7.1%)</td>
<td>47 (12.5%)</td>
</tr>
</tbody>
</table>

(*=incidence rate per 100,000 person-years

Adapted from Gritz et al., 2004

### Uveitis Prevalence

<table>
<thead>
<tr>
<th>Category</th>
<th>All N=731,898</th>
<th>Male N=336 (45.9%)</th>
<th>Female N=395 (54.1%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Anterior</td>
<td>598 (70.9%)</td>
<td>249 (74.1%)</td>
<td>349 (68.7%)</td>
</tr>
<tr>
<td>Intermediate</td>
<td>28 (3.4%)</td>
<td>11 (3.3%)</td>
<td>18 (3.5%)</td>
</tr>
<tr>
<td>Posterior</td>
<td>39 (3.6%)</td>
<td>15 (4.5%)</td>
<td>15 (3.0%)</td>
</tr>
<tr>
<td>Panuveitis</td>
<td>60 (7.1%)</td>
<td>19 (5.7%)</td>
<td>41 (8.1%)</td>
</tr>
<tr>
<td>Indeterminate</td>
<td>177 (15.0%)</td>
<td>42 (12.5%)</td>
<td>88 (16.9%)</td>
</tr>
</tbody>
</table>

Adapted from Gritz et al., 2004

### Clinical evaluation

#### Symptoms

- Floaters
- Blurred or decreased vision
- Pain
- Redness
- Photophobia

### History or present illness—critical elements to elicit

- Onset
- Pattern of recurrence
- Chronicity
- Response to previous therapy
History: “Patterns”

- Alternating unilateral (B27 associated)
- Explosive onset and rapid resolution (Behcet’s, B27 associated)

Past medical history

- Rheumatologic disease (RA, Sarcoidosis, Lupus, Behcet’s disease, inflammatory bowel disease)
- Malignancy
- Infection (lyme, syphilis, tuberculosis, parasitic disease)
- Neurological disease (multiple sclerosis)

Past ophthalmic history

- Trauma or penetrating injury (intraocular foreign body, infection, sympathetic ophthalmia)
- Previous surgery (P. acnes, haptic rub, sympathetic ophthalmia)
Social history
- Race, ethnicity
- Geographic residence
- Diet
- Animal exposure
- Travel
- Social behavior
- Sexual habits

ROS
- General health
- Skin
- Neurologic
- Ears, nose, throat
- Respiratory
- Gastrointestinal
- Bones and joints
- Vascular
- Genitourinary

SLE: conjunctiva and cornea
- Conjunctival nodules (sarcoidosis)
- Keratic precipitates: granulomatous versus nongranulomatous
- Interstitial keratitis (syphilis, Cogan’s syndrome)
- Stromal edema and/or fibrosis (HSV)

Ocular examination
- Visual acuity
- Pupils (optic neuritis, Behcet’s, sarcoidosis)
- Visual fields
- Extraocular muscle movements
- Muscle balance
- Corneal sensation
- IOP (hold on fluorescein until after AC exam)
- Lacrimal gland
SLE: anterior chamber

<table>
<thead>
<tr>
<th>Grade</th>
<th>Cells</th>
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</thead>
<tbody>
<tr>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Trace</td>
<td>1-5</td>
</tr>
<tr>
<td>1+</td>
<td>6-15</td>
</tr>
<tr>
<td>2+</td>
<td>16-25</td>
</tr>
<tr>
<td>3+</td>
<td>26-50</td>
</tr>
<tr>
<td>4+</td>
<td>&gt;50</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Grade</th>
<th>Degree of light scattering</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>absent</td>
</tr>
<tr>
<td>1</td>
<td>Very slight</td>
</tr>
<tr>
<td>2</td>
<td>Moderate (iris and lens clear)</td>
</tr>
<tr>
<td>3</td>
<td>Marked (iris and lens hazy)</td>
</tr>
<tr>
<td>4</td>
<td>Intense (fibrin)</td>
</tr>
</tbody>
</table>

SLE: anterior chamber angle

- Foreign body
- PAS

SLE: iris and lens

- Synechiae
- Pupillary fibrovascular membrane
- Iris nodules
- Transillumination defects (look before dilation)
- Cataract
- Inflammatory aggregates against posterior capsule
Vitreous cell

<table>
<thead>
<tr>
<th>Cells in retroilluminated field</th>
<th>Description</th>
<th>Grade</th>
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</thead>
<tbody>
<tr>
<td>0-1</td>
<td>Clear</td>
<td>0</td>
</tr>
<tr>
<td>2-20</td>
<td>Few opacities</td>
<td>Trace</td>
</tr>
<tr>
<td>21-50</td>
<td>Scattered opacities</td>
<td>1</td>
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<tr>
<td>51-100</td>
<td>Moderate opacities</td>
<td>2</td>
</tr>
<tr>
<td>101-250</td>
<td>Many opacities</td>
<td>3</td>
</tr>
<tr>
<td>&gt;251</td>
<td>Dense opacities</td>
<td>4</td>
</tr>
</tbody>
</table>

Vitreous haze

Retina

- Macula: cystoid and noncystoid edema
- Vessels: sheathing, narrowing, obliteration
- Periphery: fibrosis, hemorrhages, cotton wool spots, retinal atrophy, retinitis, focal chorioretinal lesion, pigment epithelial stippling, retinal neovascularization
Choroid

- Choroidal neovascularization
- Choroidal inflammatory nodules
- Fibrosis

Pars plana

- Snowbank
Optic nerve

- Disc hyperemia
- Neovascularization
- Rim pallor
- Glaucomatous atrophy

Acute versus chronic

<table>
<thead>
<tr>
<th>ACUTE</th>
<th>CHRONIC</th>
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<tbody>
<tr>
<td>S.27 associated</td>
<td>Juvenile rheumatoid arthritis</td>
</tr>
<tr>
<td>Fuchs' heterochromic iridocyclitis</td>
<td>Birdshot choroidopathy</td>
</tr>
<tr>
<td>Vogt-Koyanagi-Harada Syndrome</td>
<td>Serpiginous choroidopathy</td>
</tr>
<tr>
<td>Toxoplasmosis</td>
<td>Tuberculous uveitis</td>
</tr>
<tr>
<td>White dot syndromes, AMPPE, MEWDS</td>
<td>Sympathetic uveitis</td>
</tr>
<tr>
<td>Acute retinal necrosis</td>
<td>Sarcoidosis</td>
</tr>
<tr>
<td>Postsurgical bacterial infection</td>
<td>Postsurgical infection (P. acnes, fungal)</td>
</tr>
<tr>
<td>Trauma</td>
<td>Intermediate uveitis/pars planitis</td>
</tr>
<tr>
<td></td>
<td>Intraocular lymphoma</td>
</tr>
</tbody>
</table>

Refining a differential

- Acute or chronic
- Unilateral or bilateral
- Granulomatous or nongranulomatous
- Anatomic classification
- Patient demographics
- Review of systems
- Response to previous therapy

Causes of unilateral uveitis

- Sarcoidosis
- Postsurgical
- Intraocular foreign body
- Trauma
- Parasitic disease
- Acute retinal necrosis
- Behcet’s disease
- HSV, VZV

Causes of granulomatous inflammation

- Sarcoidosis
- Vogt-Koyanagi-Harada Syndrome
- Syphilitic uveitis
- Intraocular foreign body
- Lens-induced uveitis
- Uveitis associated with multiple sclerosis
- Syphilis
- Tuberculosis
- Other infectious agents
Anatomic localization

- Anterior
- Intermediate
- Posterior
- Panuveitis

Modifiers
- +/- cornea (keratouveitis)
- +/- sclera (sclerouveitis)
- +/- retinal vasculature (retinal vasculitis)

Causes of anterior uveitis

- Masquerade syndrome
- Posner-Schlossman glaucomatocyclitic crisis
- Syphilis
- Behçet’s disease
- Sarcoidosis
- Inflammatory bowel disease
- Psoriatic arthritis
- B27 associated juvenile rheumatoid arthritis
- Fuchs heterochromic iridocyclitis

Causes of intermediate uveitis

- HIV
- Lyme disease
- Idiopathic
- Sarcoidosis
- Sarcoidosis/pars planititis subtype
- Idiopathic

Causes of posterior uveitis

- Focal retinitis
- Syphilis
- Toxoplasmosis
- HSV
- CMV
- Focal choroiditis
- Toxocariasis
- Vogt-Koyanagi-Harada syndrome
- Nocardiosis
- Sarcoidosis

Causes of intermediate uveitis

- Inflammatory bowel disease
- Multiple sclerosis
- Lyme disease
- HIV
- Idiopathic
- Idiopathic/pars planititis subtype

Causes of posterior uveitis

- Focal choroiditis
- Toxocariasis
- Tuberculosis
- Nocardiosis
- Masquerade

- Focal choroiditis
- Toxocariasis
- Vogt-Koyanagi-Harada syndrome
- Histoplasmosis
- Sarcoidosis
- Serpigious choroidopathy
- Birdshot choroidopathy
- Masquerade
Causes of panuveitis

- Sarcoidosis
- Vogt-Koyanagi-Harada syndrome
- Behcet’s disease
- Syphilis
- Tuberculosis
- Infectious endophthalmitis

Masquerade syndromes

- Intraocular lymphoma and leukemia
- Retinoblastoma
- Choroidal melanoma
- Metastatic carcinomas
- Systemic vascular diseases
- Pigment dispersion syndrome
- Retinal detachment
- Hereditary/Degenerative retinal diseases
- Trauma and intraocular foreign bodies
### Patient demographics: age

<table>
<thead>
<tr>
<th>Age (in yrs)</th>
<th>Disease risks</th>
</tr>
</thead>
<tbody>
<tr>
<td>&lt;5</td>
<td>Leukemia, Postviral neuroretinitis, Toxocariasis, Sarcoidosis, JRA</td>
</tr>
<tr>
<td>5-15</td>
<td>Acute retinal necrosis, Pars Planitis, Sarcoidosis, Toxocariasis</td>
</tr>
<tr>
<td>16-25</td>
<td>Serpiginous choroidopathy, Behcet’s disease, Idiopathic retinal vasculitis, Idiopathic intermediate uveitis</td>
</tr>
<tr>
<td>25-45</td>
<td>Acute retinal necrosis, Serpiginous choroidopathy, Behcet’s disease, Idiopathic retinal vasculitis, Idiopathic intermediate uveitis, Birdshot retinochoroiditis</td>
</tr>
<tr>
<td>&gt;65</td>
<td>Masquerade syndromes, Post-surgical (P. acnes), Sarcoidosis</td>
</tr>
</tbody>
</table>

### Associated symptoms and signs

<table>
<thead>
<tr>
<th>Symptom or sign</th>
<th>Possible condition</th>
</tr>
</thead>
<tbody>
<tr>
<td>Headaches</td>
<td>Sarcoidosis, VKH</td>
</tr>
<tr>
<td>Neurosensory deafness</td>
<td>VKH, Sarcoidosis</td>
</tr>
<tr>
<td>CSF pleocytosis</td>
<td>VKH, Sarcoidosis, AMPPE, Behcet’s</td>
</tr>
<tr>
<td>Paresthesias, weakness</td>
<td>Multiple sclerosis, Behcet’s</td>
</tr>
<tr>
<td>Psychosis</td>
<td>VKH, Behcet’s, Lupus</td>
</tr>
</tbody>
</table>

### Patient demographics

<table>
<thead>
<tr>
<th>Factor</th>
<th>Disease risks</th>
</tr>
</thead>
<tbody>
<tr>
<td>African American</td>
<td>Sarcoidosis</td>
</tr>
<tr>
<td>Native American</td>
<td>Vogt-Koyanagi-Harada syndrome</td>
</tr>
<tr>
<td>Asian</td>
<td>VKH, Behcet’s</td>
</tr>
<tr>
<td>Mediterranean</td>
<td>Behcet’s</td>
</tr>
<tr>
<td>American midwest</td>
<td>Ocular histoplasmosis</td>
</tr>
<tr>
<td>Central &amp; South America</td>
<td>Cysticercosis, onchocerciasis</td>
</tr>
<tr>
<td>West African</td>
<td>Onchocerciasis</td>
</tr>
</tbody>
</table>

### Associated symptoms and signs

<table>
<thead>
<tr>
<th>Symptom or sign</th>
<th>Possible condition</th>
</tr>
</thead>
<tbody>
<tr>
<td>Vitiligo, poliosis</td>
<td>VKH</td>
</tr>
<tr>
<td>Erythema nodosum</td>
<td>Behcet’s, sarcoidosis</td>
</tr>
<tr>
<td>Skin nodules</td>
<td>Sarcoidosis, onchocerciasis</td>
</tr>
<tr>
<td>Alopecia</td>
<td>VKH</td>
</tr>
<tr>
<td>Skin rash</td>
<td>Behcet’s, sarcoidosis, post-viral, Lyme Disease, syphilis, zoster, psoriatic arthritis</td>
</tr>
<tr>
<td>Pathergy</td>
<td>Behcet’s</td>
</tr>
</tbody>
</table>

### Patient demographics

<table>
<thead>
<tr>
<th>Factor</th>
<th>Disease risks</th>
</tr>
</thead>
<tbody>
<tr>
<td>IV drug use</td>
<td>HIV, fungal infection</td>
</tr>
<tr>
<td>Multiple sexual partners</td>
<td>HIV, syphilis</td>
</tr>
<tr>
<td>Hiking in wooded areas</td>
<td>Lyme</td>
</tr>
<tr>
<td>Unpasteurized milk consumption</td>
<td>Brucellosis</td>
</tr>
<tr>
<td>Exposure to puppies</td>
<td>Toxocariasis</td>
</tr>
<tr>
<td>Cats</td>
<td>Toxoplasmosis</td>
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</tbody>
</table>

### Associated symptoms and signs

<table>
<thead>
<tr>
<th>Symptom or sign</th>
<th>Possible condition</th>
</tr>
</thead>
<tbody>
<tr>
<td>Oral ulcers</td>
<td>Behcet’s, inflammatory bowel disease</td>
</tr>
<tr>
<td>Genital ulcers</td>
<td>Behcet’s, Reiter’s syndrome, sexually transmitted disease</td>
</tr>
<tr>
<td>Genital ulcers</td>
<td>Behcet’s, Reiter’s syndrome, sexually transmitted disease</td>
</tr>
</tbody>
</table>
### Associated symptoms and signs

<table>
<thead>
<tr>
<th>Symptom or sign</th>
<th>Possible condition</th>
</tr>
</thead>
<tbody>
<tr>
<td>Salivary or lacrimal gland swelling</td>
<td>Sarcoidosis, lymphoma</td>
</tr>
<tr>
<td>Lymphadenopathy</td>
<td>Sarcoidosis, lymphoma, HIV</td>
</tr>
</tbody>
</table>

### Associated symptoms and signs

<table>
<thead>
<tr>
<th>Symptom or sign</th>
<th>Possible condition</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sinusitis</td>
<td>Wegener’s granulomatosis</td>
</tr>
<tr>
<td>Cough, shortness of breath, wheezing</td>
<td>Sarcoidosis, tuberculosis, malignancy</td>
</tr>
<tr>
<td>Diarrhea</td>
<td>Inflammatory bowel disease</td>
</tr>
<tr>
<td>Chemotherapy or other immunosuppression</td>
<td>CMV retinitis, Candida retinochoroiditis, opportunistic infection</td>
</tr>
</tbody>
</table>

### Laboratory evaluation

- FTA
- ACE (not as useful in children)
- Chest X-ray
- PPD

Do not shotgun!

### HLA associations

<table>
<thead>
<tr>
<th>Disease</th>
<th>Antigen</th>
<th>Relative Risk</th>
</tr>
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<tbody>
<tr>
<td>Acute anterior uveitis</td>
<td>HLA B27 (W)</td>
<td>10</td>
</tr>
<tr>
<td>Ankylosing spondylitis</td>
<td>HLA B27 (W)</td>
<td>100</td>
</tr>
<tr>
<td>Behcet’s disease</td>
<td>HLA B51 (O, W)</td>
<td>4-6</td>
</tr>
<tr>
<td>Behcet’s retinochoroidopathy</td>
<td>HLA A29 (W)</td>
<td>49</td>
</tr>
<tr>
<td>Reiter’s syndrome</td>
<td>HLA A27 (W)</td>
<td>40</td>
</tr>
<tr>
<td>Rheumatoid arthritis</td>
<td>HLA DR4 (W)</td>
<td>11</td>
</tr>
<tr>
<td>Sympathetic ophthima</td>
<td>HLA A11</td>
<td>4</td>
</tr>
<tr>
<td>Vogt-Koyanagi-Harada disease</td>
<td>HLA DR4 (O)</td>
<td>15</td>
</tr>
<tr>
<td>Ankylosing spondylitis</td>
<td>HLA DR3-3 (O)</td>
<td>74</td>
</tr>
</tbody>
</table>

Adapted from Nussenblatt and Whitcup, 2004.

### Ancillary studies

- Classify
- Diagnose
- Treat
Fluorescein angiography

OKAP chant

• “block early, stain late”
Prednisone and cyclosporine
Laser interferometry visual acuity

- 3 line or better improvement in visual acuity correlates with improved vision after treatment with immunosuppressive therapy, even in setting of cystoid macular edema
B Scan

UBM

Functional Testing

- Initial evaluation
- Response to therapy

- Fields
- Electrophysiologic testing
**Biopsy**
- PCR
- Antibodies
- Cytokines
- Communicate with pathologist!

**Steroids**
- Start at 0.75 to 1.0 mg/kg per day for active inflammation.
- Start at 0.5 mg/kg for CME
- Do not taper too soon or too fast.
- Start tapering from high dosages after one month while phasing in a steroid sparing agent if needed

**Therapy: Principles**
- Define why vision is down.
- Each patient receives an individualized regimen that will evolve over time.
- Two to three months must pass to evaluate full response to a particular regimen.
- Weigh risks and side effects versus potential benefit.
- Phase in steroid sparing agents over time.
- Do not expect to eliminate systemic steroids totally.
Steroids: monitor and supplement

- Blood pressure, weight, glucose, lipids
- Bone mineral density

Supplement with Calcium 1500 mg daily and vitamin D 800 IU daily +/- Fosamax if indicated.
Tapering steroids

- Listen to the patient: he or she often knows the “flare dose.”
- Do not expect to get to zero steroid in every patient.

<table>
<thead>
<tr>
<th>Current dose per day</th>
<th>Tapering regimen</th>
</tr>
</thead>
<tbody>
<tr>
<td>&gt;40 mg</td>
<td>Decrease by 10 mg every 1-2 weeks</td>
</tr>
<tr>
<td>20-40 mg</td>
<td>Decrease by 5 mg every 1-2 weeks</td>
</tr>
<tr>
<td>10-20 mg</td>
<td>Decrease by 1 to 2.5 mg every 1-2 weeks</td>
</tr>
<tr>
<td>0-10 mg</td>
<td>Decrease by 1 to 2.5 mg every 1-4 weeks</td>
</tr>
</tbody>
</table>

Periocular steroid
Cyclosporine

- 3-5 mg/kg/day
- Monitor BP, serum creatinine, liver enzymes
- ?Check peak and trough

Other agents

- Azathioprine
- Mycophenolate
- Methotrexate
- Cyclophosphamide
Perioperative management

- Quiet for 3 months
- Start prednisone 0.75 to 1 mg per kg two days before surgery
- Hold at that dose for 1 week, then start taper

Future horizons

- Infliximab
- Daclizumab
- Biologics
Daclizumab

- Autoaggressive Th1 cells express IL-2R
- Chimeric antibody
- Targets alpha or “Tac” subunit
- Not good for anterior disease
Inflammation and other retinal conditions

- AMD
- Diabetic retinopathy
- Immunological manipulation of eye
57 year old woman

- LASIK July 2000
- Enhancement OD October 2000—still no improvement
- Steady decline OD 2001 and 2002

Retinal consultation spring 2003

- Clinical exam, angiography, and ERG -> “Atypical retinitis pigmentosa”

2nd retinal consultation

- Atypical retinitis pigmentosa
- Vitamin A supplementation

POHX: unremarkable
PMHx: skin cancer cheek 1993
FamHx: sister pars planitis
SocHx: Dutch ancestry
Diffuse thickening in macula 400 microns

78 ETDRS

160 microns at fovea
Aqueous fluid OS

- IL 10: 175 pg/mL
- IL 6: undetectable
- Acellular specimen

Lumbar puncture

- VDRL negative
- IL 10: 775 pg/mL
- IL 6 undetectable
- Few large lymphoid cells with prominent nuclei
- IgH gene rearrangement
- bcl-2 gene translocation

Core vitreous specimen

- Large B cell primary central nervous system lymphoma with parenchymal, leptomeningeal, and bilateral ocular involvement
“Vitritis” without RPE infiltrates
- Clumps of cells along detached posterior hyaloid
- Absence of CME
- Age
- Fluorescein changes at level of RPE
- Vision better than expected for vitreous cellularity
- Minimal anterior segment activity

Primary CNS Lymphoma
- Diffuse large B cell
- Burkitt’s
- Intravascular large B cell
- T cell and anaplastic large cell
- Hodgkins
- Diffuse large B cell, HIV associated (PCR on CSF for EBV)

MRI
- Parenchymal disease: T1 post gadolinium
- Solid mass that enhances homogenously, often in periventricular white matter or basal ganglia
- Leptomeningeal: FLAIR with fat suppression and gadolinium

Diagnostic vitrectomy
- Fragile cells
- 20 minutes versus 60 minutes

LP
- Flow cytometry to detect occult CNS disease

- Atypical retinitis pigmentosa
- Retinal degeneration of unknown etiology
Retinal degeneration of unknown etiology vs. Atypical retinitis pigmentosa

“Atypical something is usually typical something else.”

Warren Grant Magnuson Clinical Center