Uveitis

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Uveal tissue. Note the deep purple hue
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**Identifying a uveitis is, in essence, a pattern-recognition task.** The uveitides do not present in random fashion; rather, they ‘select’ their victims based on pt demographics. Likewise, the nonocular manifestations of those 2ndry to a systemic condition tend to follow specific patterns as well. Each tends to affect the eye in a characteristic manner.
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Other important demographic factors include:
- Geographic history, ie, where they have lived or visited
- Social history (eg, sexual behaviors; dietary habits; IVDU)
- Vocational/avocational activities (eg, exposure to farm animals)

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**Uveitis**

What needs to be covered in the ROS? **Everything!** The following list is by no means complete:

- **Constitutional:** Fever; night sweats; weight loss
- **Neuro:** HA; cranial neuropathies; hearing loss; cognitive changes
- **ENT:** Oral ulcers; sinusitis; ear or nose deformities
- **Skin:** Rashes; poliosis, madarosis; vitiligo; erythema nodosum
- **Pulmonary:** SOB/DOE; cough; hemoptysis
- **Cardiac:** Arrhythmias; pericarditis symptoms
- **GI:** Diarrhea; ulcers
- **GU:** Genital lesions; discharge; nephritis symptoms
- **Musculoskeletal:** Arthralgias; low back pain

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Let’s drill down on the different ways uveitis manifests within the eye. One very important manifestation issue is **location**, ie, the portion or segment of the eye that’s involved.

- Each tends to affect the eye in a characteristic manner.
What are the four basic anatomic locations for uveitis?
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In anterior uveitis, the primary location of inflammation is the two words and/or two words.
In **anterior uveitis**, the primary location of inflammation is the **anterior chamber** and/or **anterior vitreous**.
Uveitis

Anterior uveitis

*If cell is located...*

Exclusively in the AC

*It is called:*

? 

In **anterior uveitis**, the primary location of inflammation is the anterior chamber and/or anterior vitreous
Uveitis

Anterior uveitis

If cell is located…

Exclusively in the AC

It is called:

Iritis

In anterior uveitis, the primary location of inflammation is the anterior chamber and/or anterior vitreous
Uveitis

In **anterior uveitis**, the primary location of inflammation is the anterior chamber and/or anterior vitreous.
Uveitis

Anterior uveitis

If cell is located…

- Exclusively in the AC
  - It is called: Iritis

- Exclusively in the AVit
  - It is called: Anterior cyclitis

In anterior uveitis, the primary location of inflammation is the anterior chamber and/or anterior vitreous.
Uveitis

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Uveitis

The onset, duration and course of the uveitis:
- **Onset:** Sudden vs insidious
- **Duration:** Limited vs persistent
- **Course:** Acute vs recurrent vs chronic

The severity of the uveitis:
- **AC cell grade**
- **AC flare grade**
- **Vitreous haze score**

Anterior uveitis

If cell is located...
- Exclusively in the AC
  - It is called: **Iritis**
- In the AC and the anterior vitreous
  - It is called: **Iridocyclitis**
- Exclusively in the AVit
  - It is called: **Anterior cyclitis**

In **anterior uveitis**, the primary location of inflammation is the anterior chamber and/or anterior vitreous
In **intermediate uveitis**, the primary location of inflammation is the **main vitreous cavity**, +/- the **peripheral retina**.
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**Intermediate uveitis**

*If condition is…*

- **Idiopathic**
  - *It is called:*
  - **Pars planitis**

- **Not idiopathic**

*How we divvy up IU*
In intermediate uveitis, the primary location of inflammation is the main vitreous cavity, +/- the peripheral retina.
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Intermediate uveitis

If condition is...

Idiopathic

- It is called: Pars planitis

Not idiopathic

- It is called: Intermediate uveitis
In posterior uveitis, the site of inflammation is the retina and/or choroid (the optic nerve head can be involved too).
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Uveitis

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**Posterior uveitis**

*If inflammation is located...*

- Exclusively in the choroid
  - *It is called:* Choroiditis
- In both the choroid and the retina
  - *It is called:* ?
- Exclusively in the retina
  - *It is called:* Retinitis
In posterior uveitis, the site of inflammation is the retina and/or choroid (the optic nerve head can be involved too)
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Uveitis

Posterior uveitis

If inflammation is located...

- Exclusively in the choroid
  - It is called: Choroiditis

- In both the choroid and the retina
  - It is called: Chorioretinitis or Retinochoroiditis

- Exclusively in the retina
  - It is called: Retinitis

- Exclusively in the retina and ONH
  - It is called: Neuroretinitis

In posterior uveitis, the site of inflammation is the retina and/or choroid (the optic nerve head can be involved too)
In panuveitis, all three locations are equally involved.
Many experts endorse a *profiling and meshing* approach to diagnosing uveitis.
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*Profiling* refers to identifying germane aspects of the pt’s personal history (age, ethnicity, occupation, etc); nonocular signs and symptoms associated with the uveitis (eg, skin findings; CNS involvement); and key features of the inflammation itself (ie, location, duration, etc).
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*Meshing* refers to matching the profile of the pt with the known proclivities of specific uveitic entities. In this way, a DDx is generated: ‘*VKH* affects adults of ME descent, produces a bilateral granulomatous panuveitis, and is associated with CNS manifestations, especially tinnitus’

(*VKH* = *Vogt-Koyanagi-Harada dz*. You’ll come to know it well.)
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Once a set of potential diagnoses have been identified via profiling and meshing, lab and other studies are obtained to identify the offending condition…After which the appropriate treatment can be instituted.
Let’s drill down on **anterior uveitis**. Specifically, let’s look at how the BCSC organizes it by presentation.
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*What is meant by the term ciliary flush?*
Anterior uveitis is by far the most common form encountered clinically. The classic symptoms are **pain** and **photophobia**, along with some degree of **reduced vision**. Patients will also complain of **surface injection** (which presents often in a so-called *ciliary flush* pattern).

*What is meant by the term ciliary flush?*  
It refers to dilated deep conjunctival and episcleral vessels adjacent and circumferential to the corneal limbus.
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Ciliary body just deep to the limbal region

Injection in anterior uveitis, aka *ciliary flush*

In surface disorders (eg, conjunctivitis), redness is either distributed uniformly across the eye, or it tapers off near the limbus. In contrast, redness associated with anterior uveitis is usually most intense at and just behind the limbus, and may taper away from it. This is because this area overlies the inflamed ciliary body (hence the term *ciliary flush* for this presentation).
Anterior uveitis is by far the most common form encountered clinically. The classic symptoms are pain and photophobia, along with some degree of reduced vision. Patients will also complain of surface injection (which presents often in a so-called ciliary flush pattern). At the slit lamp, the classic signs of anterior uveitis are WBCs and inflammatory proteins in the AC (cell and flare).
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Uveitis

Aqueous cells and flare

cells

flare
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Granulomatous Nongranulomatous

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Uveitis

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Granulomatous Nongranulomatous

These are the common entities that can produce a granulomatous anterior uveitis. (Note: For some of these, the granulomatous anterior findings are part of an overall panuveitic presentation, ie, they typically do not present as an *isolated* anterior uveitis.)
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In contrast, nongranulomatous KP are smaller, lighter in color, and do not look greasy. (Note: If no KP are present, the inflammation is considered nongranulomatous.)
Uveitis

Nongranulomatous KP
The rest of the anterior-uveitis classification tree concerns nongranulomatous dz.
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The rest of the anterior-uveitis classification tree concerns nongranulomatous dz. The first branch-point divides the etiologies into those that produce **acute dz** vs those producing **chronic dz**.

**Acute uveitis** comes on suddenly and resolves fairly quickly.
The rest of the anterior-uveitis classification tree concerns nongranulomatous dz. The first branch-point divides the etiologies into those that produce acute dz vs those producing chronic dz.

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Anterior Uveitis

Granulomatous

Nongranulomatous

Acute

Chronic

Unilateral

Bilateral

Take a good look at this—it represents how you should think about anterior uveitides encountered in the clinic or on the OKAP. It wouldn’t be a bad idea to commit this to memory at this juncture.

Finally, the acute uveitides are divided into those that present unilaterally vs those that tend to present bilaterally.
Just as an FYI, these are the anterior uveitides that are covered in detail in the *Uveitis* book. *Don’t try to memorize all this now!* (They will stick better if you learn them in their naturally-occurring groupings.)
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One fact you **should** memorize is that the vast majority of anterior-uveitis cases—about 80%—are infectious vs non-infectious.
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One fact you **should** memorize is that the vast majority of anterior-uveitis cases—about 80%—are *noninfectious*. 
Note that syphilis, sarcoid and TB show up everywhere on the tree. This is because all three can manifest in so many different ways.
Acute Chronic

Unilateral Bilateral

Nongranulomatous

Granulomatous

Anterior Uveitis

HSV

Syphilis

Sarcoid

TB

HLA-B27 dz

Posner-Schlossman

HSV/VZV

Syphilis

Sarcoid

TB

Acute

Bilateral

TINU

Behçet

Drug rxn

Leptospirosis

IBD/PA

Syphilis

Sarcoid

TB

Rule of thumb:

Syphilis, sarcoid and TB are on the DDx for every pt with any form of uveitis!

Note that syphilis, sarcoid and TB show up everywhere on the tree. This is because all three can manifest in so many different ways.
Syphilis, sarcoid and TB are on the DDx for every pt with any form of uveitis!
Next let’s look at intermediate uveitis
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The hallmark of **intermediate uveitis** (IU) is inflammation in the **anterior vitreous** that involves the **vitreous base**. The vitreous base is the primary attachment point of the vitreous; it forms a ~5 mm-wide band that straddles the **ora serrata** (the location where the anterior-most retina meets the posterior-most portion of the ciliary body).
Uveitis

The vitreous base

- Ciliary body
- Peripheral retina
The hallmark of intermediate uveitis (IU) is inflammation in the anterior vitreous that involves the vitreous base. The vitreous base is the primary attachment point of the vitreous; it forms a ~5 mm-wide band that straddles the ora serrata (the location where the anterior-most retina meets the posterior-most portion of the ciliary body). AC cell is typically present.
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Snowballs in intermediate uveitis
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IU tends to be a dz of young people—teens through 40 or so. It is bilateral in most (80%) cases (although it can be quite asymmetric).
As indicated previously, IU is divvied up into two categories.
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As indicated previously, IU is divided into two categories. If the inflammation is associated with an identifiable condition, the uveitis is called **IU**. If it is idiopathic, i.e., if no cause can be identified, it is called **pars planitis**.
The entities most likely to produce IU are:

- Pars planitis
- Intermediate uveitis

The diagram illustrates the relationships between anterior, intermediate, and posterior uveitis, with intermediate uveitis being a subset of anterior and posterior uveitis.
Uveitis

- Anterior
- Intermediate
- Posterior
- Panuveitis

Intermediate

- Pars planitis
- Intermediate uveitis

The entities most likely to produce IU

- MS
- Lyme
- Toxocariasis
- Sarcoid
- Syphilis
- TB
Uveitis

Anterior

Posterior

Intermediate

Pars planitis

Intermediate uveitis

Panuveitis

MS

Lyme

Syphilis

Sarcoid

TB

Note the appearance of these three on the IU DDx as well.
Let’s take a closer look at **posterior uveitis**
Uveitis

Posterior uveitis

*If inflammation is located…*

- Exclusively in the choroid
  - *It is called:* Choroiditis

- In both the choroid and the retina
  - *It is called:* Chorioretinitis or Retinochoroiditis

- Exclusively in the retina
  - *It is called:* Retinitis

As noted previously, here are the ways posterior uveitis can manifest.
As noted previously, here are the ways *posterior uveitis* can manifest. Also noted previously was the condition *neuroretinitis*—inflammation involving both the retina and optic nerve.
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While 80% of anterior uveitis cases are noninfectious in origin, the opposite is true for posterior uveitis: most cases are infectious—weirdly, also about 80%.
**Posterior uveitis**

*If inflammation is located…*

- Exclusively in the choroid
  - It is called: **Choroiditis**

- In both the choroid and the retina
  - It is called: **Chorioretinitis** or **Retinochoroiditis**

- Exclusively in the retina and ONH
  - It is called: **Retinitis**
  - **Neuroretinitis**

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**Toxoplasmosis** is a common, classic cause of posterior uveitis. It is infectious, the bug being an obligate intracellular parasite. **Cats** are its definitive host. *T gondii* has a worldwide distribution; an estimated one billion people are infected. Humans usually acquire the parasite via **consumption of unwashed produce or undercooked meat**. Another crucial mechanism of transmission is transplacentally, which leads to devastating congenital manifestations in affected infants (it is one of the TORCH syndrome etiologies).
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Uveitis

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Posterior uveitis

If inflammation is located...

- Exclusively in the choroid: Choroiditis
- In both the choroid and the retina: Chorioretinitis or Retinochoroiditis
- Exclusively in the retina and ONH: Retinitis

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Uveitis

Toxoplasma gondii: Three infectious forms

Oocyst form
--Found in GI tract of cat (shed in feces)
--Acquired via ingestion of unwashed produce

Tachyzoite form
--Found in circulatory system of infected mother
--Responsible for transplacental infection

Tissue cyst
--Found in tissue of infected livestock
--Acquired via consumption of undercooked meat
Posterior uveitis

If inflammation is located…

- Exclusively in the choroid
  - It is called: Choroiditis
- In both the choroid *and* the retina
  - It is called: Chorioretinitis *or* Retinochoroiditis
- Exclusively in the retina *and* ONH
  - It is called: Retinitis

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Toxoplasmosis typically manifests as a retinochoroiditis accompanied by a dense overlying vitritis. Taken together, the appearance has been likened to a 'four words'.
**Uveitis**

**Posterior uveitis**

*If inflammation is located…*

- Exclusively in the choroid
  - *It is called:* Choroiditis

- In both the choroid *and* the retina
  - *It is called:* Chorioretinitis or Retinochoroiditis

- Exclusively in the retina *and* ONH
  - *It is called:* Retinitis
    - Neuroretinitis

---

Toxoplasmosis is a common, classic cause of posterior uveitis. It is infectious, the bug being *Toxoplasma gondii*, an obligate intracellular parasite. Cats are its definitive host. *T. gondii* has a worldwide distribution; an estimated one billion people are infected. Humans usually acquire the parasite via consumption of unwashed produce or undercooked meat. Another crucial mechanism of transmission is transplacentally, which leads to devastating congenital manifestations in affected infants (it is one of the TORCH syndrome etiologies).

Toxoplasmosis typically manifests as a retinochoroiditis accompanied by a dense overlying vitritis. Taken together, the appearance has been likened to a ‘**headlight in the fog**.’
Uveitis

Ocular toxoplasmosis: ‘Headlight in the fog’
Let’s take a closer look at panuveitis.
To qualify as a panuveitis, all compartments of the eye—the AC, vitreous, and retina/choroid—must be equally involved in the inflammatory process.
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The panuveitides are divvied into **noninfectious** and **infectious** causes.
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- Sarcoid
- Sympathetic ophthalmia
- Vogt-Koyanagi-Harada
- Behçet syndrome
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- Leptospirosis
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- Note that these three appear yet again
- Syphilis
- TB
- Lyme
- Leptospirosis
- Whipple dz
Endophthalmitis

Now we’ll change gears and look at endophthalmitis
Endophthalmitis

The *Uveitis* book defines **endophthalmitis** as an inflammatory process involving both the AC and vitreous cavities that is secondary to bacterial or fungal infection.
Endophthalmitis

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Endophthalmitis can be **posttraumatic**, **postoperative** or **endogenous**.
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**Endogenous endophthalmitis** involves spread of infection from a remote location to the eye. It is uncommon, mechanism common vs not.
Endogenous Endophthalmitis

The *Uveitis* book defines endophthalmitis as an inflammatory process involving both the AC and vitreous cavities that is secondary to bacterial or fungal infection. Endophthalmitis can be posttraumatic, postoperative or endogenous.

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The *Uveitis* book defines **endophthalmitis** as an inflammatory process involving both the AC and vitreous cavities that is secondary to bacterial or fungal infection. Endophthalmitis can be **posttraumatic, postoperative** or **endogenous**.

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Note that the fact that the route is hematogenous indicates a nidus of infection is present somewhere in the body, and it is incumbent upon the pt’s care team to find and treat it!
Endogenous Endophthalmitis

The *Uveitis* book defines **endophthalmitis** as an inflammatory process involving both the AC and vitreous cavities that is secondary to bacterial or fungal infection. Endophthalmitis can be **posttraumatic, postoperative or endogenous**.

**Endogenous endophthalmitis** involves hematogenous spread of infection from a remote location to the eye. It is uncommon, accounting for less than 10% of all cases of endophthalmitis. **Individuals at increased risk of endogenous endophthalmitis include those with general state**, those who recently underwent an invasive medical procedure, and those subjected to chronic and/or repeated breaching of the body’s outer barrier.
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**Endogenous endophthalmitis** involves hematogenous spread of infection from a remote location to the eye. It is uncommon, accounting for less than 10% of all cases of endophthalmitis. **Individuals at increased risk of endogenous endophthalmitis include those with impaired immune status**, those who recently underwent an invasive medical procedure, and those subjected to chronic and/or repeated breaching of the body’s outer barrier.
Endogenous Endophthalmitis

- Bacterial
- Fungal

As mentioned, endogenous endophthalmitis can be bacterial or fungal.
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Some classic bacterial pathogen associations in endogenous endophthalmitis:
--Endocarditis: ?
--Skin infections
--IVDU
--Liver abscess
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Bacterial    Fungal

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Some classic bacterial pathogen associations in endogenous endophthalmitis:
-- *Endocarditis*: Strep
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Endogenous Endophthalmitis

Bacterial

Fungal

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Some classic bacterial pathogen associations in endogenous endophthalmitis:

--*Endocarditis*: Strep
--*Skin infections*: Staph
--*IVDU*: Bacillus
--*Liver abscess*: Klebsiella
Endogenous Endophthalmitis

Bacterial

Fungal

In contrast to the bacterial version, endogenous **fungal** endophthalmitis tends to be more insidious in onset.
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Endogenous Endophthalmitis

In contrast to the bacterial version, endogenous **fungal** endophthalmitis tends to be more insidious in onset. It generally progresses in a particular fashion. First, isolated metastatic lesions appear.
In contrast to the bacterial version, endogenous fungal endophthalmitis tends to be more insidious in onset. It generally progresses in a particular fashion. First, isolated choroidal metastatic lesions appear.
Endogenous Endophthalmitis

*Candida* endophthalmitis: Choroidal lesions
Endogenous Endophthalmitis

- Bacterial
- Fungal

In contrast to the bacterial version, endogenous **fungal** endophthalmitis tends to be more insidious in onset. It generally progresses in a particular fashion. First, isolated choroidal metastatic lesions appear. With time, these break through to involve the retina.
In contrast to the bacterial version, endogenous **fungal** endophthalmitis tends to be more insidious in onset. It generally progresses in a particular fashion. First, isolated choroidal metastatic lesions appear. **With time, these break through** Bruch’s membrane to involve the retina.
Endogenous Endophthalmitis

*Candida* endophthalmitis: Retinal involvement
Endogenous Endophthalmitis

In contrast to the bacterial version, endogenous fungal endophthalmitis tends to be more insidious in onset. It generally progresses in a particular fashion. First, isolated choroidal metastatic lesions appear. With time, these break through Bruch’s membrane to involve the retina. Eventually, the bug reaches the vitreous, and (if still unchecked) the anterior segment.
Endogenous Endophthalmitis

Candida endophthalmitis: Classic three words vitreous involvement
Endogenous Endophthalmitis

*Candida* endophthalmitis: Classic ‘string of pearls’ vitreous involvement
Endogenous Endophthalmitis

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Some classic fungal pathogen associations in endogenous endophthalmitis:

--Chronic indwelling lines/catheters: ?
--HIV/AIDS
--Hx liver transplantation
--San Joaquin valley
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Some classic fungal pathogen associations in endogenous endophthalmitis:
--Chronic indwelling lines/catheters: Candida
--HIV/AIDS: Cryptococcus
--Hx liver transplantation: Aspergillus
--San Joaquin valley: Coccidioides
Uveitis

Next let’s take a look at Scleritis
Scleritis is an inflammatory condition characterized by painful infiltrative scleral edema and congestion of the deep episcleral plexus.
Scleritis is an inflammatory condition characterized by painful infiltrative scleral edema and congestion of the deep episcleral plexus.
Uveitis

Anatomical depiction of the conjunctiva, episclera, and scleral stroma, and the approximate location of the conjunctival, superficial episcleral, and deep vascular plexi.

Scleritis: Deep episcleral plexus edema
Scleritis is an inflammatory condition characterized by painful infiltrative scleral edema and congestion of the deep episcleral plexus. It can be extremely painful, and can lead to blindness and loss of the eye.
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Uveitis

Scleritis

Scleritis is an inflammatory condition characterized by painful infiltrative scleral edema and congestion of the deep episcleral plexus. It can be extremely painful, and can lead to blindness and loss of the eye. Women are more likely to be affected than are men. It is rare in children.

To make matters worse, scleritis can herald the presence or worsening of a systemic condition that may be potentially lethal. About % of scleritis pts have an identifiable systemic inflammatory condition, the most common of which is
Scleritis

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To make matters worse, scleritis can herald the presence or worsening of a systemic condition that may be potentially lethal. About 40% of scleritis pts have an identifiable systemic inflammatory condition, the most common of which is rheumatoid arthritis (RA).
Uveitis

Scleritis is divvied up with respect to whether the...
Scleritis is divvied up with respect to whether the…

Anterior sclera is affected, vs the Posterior sclera.
There are three classic signs of anterior scleritis:
--Scleral
--
There are three classic signs of anterior scleritis:
--Scleral edema
--
--
Anterior scleritis: Scleral edema. Note the thickening of the limbal sclera (b) in comparison to the unaffected fellow eye (a).
There are three classic signs of anterior scleritis:

- Scleral edema
- Sclera has a violaceous hue
There are three classic signs of anterior scleritis:
--Scleral edema
--Sclera has a violaceous hue
--
Uveitis

Anterior scleritis: Violaceous hue
There are three classic signs of anterior scleritis:
--Scleral edema
--Sclera has a violaceous hue
--Inflamed vasculature has a pattern
There are three classic signs of anterior scleritis:
--Scleral edema
--Sclera has a violaceous hue
--Inflamed vasculature has a criss-cross pattern
‘Criss-cross’ injection of the deep vasculature in anterior scleritis. (To see it, you have to ‘look past’ the brighter injection of the inflamed overlying conj vessels)
Uveitis

Scleritis

Anterior

Posterior

Anterior scleritis comes in three forms:
Anterior scleritis comes in three forms: *Diffuse, nodular and necrotizing*
Uveitis

Diffuse anterior scleritis
Uveitis

Nodular anterior scleritis
Necrotizing anterior scleritis comes in two forms:
Necrotizing anterior scleritis comes in two forms: 
*With* and *without inflammation*.
Uveitis

Necrotizing anterior scleritis \textit{with} inflammation
Uveitis

Scleritis

Anterior

• Diffuse
• Nodular

Posterior

• Necrotizing
  - w/ inflammation
  - w/o inflammation

Contrary to the implications of the name, inflammation is present in necrotizing scleritis w/o inflammation.
Contrary to the implications of the name, inflammation is present in *necrotizing scleritis w/o inflammation*. It is so named because, unlike its ‘with inflammation’ cousin, it is typically painless, and the eye does not appear inflamed.
Uveitis

Necrotizing anterior scleritis without inflammation
Contrary to the implications of the name, inflammation is present in necrotizing scleritis w/o inflammation. It is so named because, unlike its ‘with inflammation’ cousin, it is typically painless, and the eye does not appear inflamed. Necrotizing scleritis w/o inflammation is also known as "scleromalacia perforans". It is strongly associated with RA.
Contrary to the implications of the name, inflammation is present in **necrotizing scleritis w/o inflammation**. It is so named because, unlike its ‘with inflammation’ cousin, it is typically painless, and the eye does not appear inflamed. **Necrotizing scleritis w/o inflammation** is also known as **scleromalacia perforans**. It is strongly associated with RA.
Unlike anterior scleritis, posterior scleritis does not present with a red eye, and nodules are not present. Instead, posterior scleritis presents with:

- Proptosis
- Disc edema
- Motility disorders
- Retinal/choroidal findings
Unlike anterior scleritis, **posterior scleritis** does not present with a red eye, and nodules are not present. Instead, posterior scleritis presents with:

--**Proptosis**

---Please note---
Uveitis

Posterior scleritis: Proptosis
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Unlike anterior scleritis, **posterior scleritis** does not present with a red eye, and nodules are not present. Instead, posterior scleritis presents with:

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- **Disc edema**

**Uveitis**

**Scleritis**

- **Anterior**
- **Posterior**

**Diffuse**

w/ inflammation  w/o inflammation
Uveitis

Posterior scleritis OD: Optic nerve edema
Scleritis

Unlike anterior scleritis, **posterior scleritis** does not present with a red eye, and nodules are not present. Instead, posterior scleritis presents with:

- **Proptosis**
- **Disc edema**
- Retinal/choroidal findings

Anterior

Posterior

Diffuse

w/ inflammation

w/o inflammation
Uveitis

Posterior scleritis producing retinal folds
Unlike anterior scleritis, posterior scleritis does not present with a red eye, and nodules are not present. Instead, posterior scleritis presents with:

--Proptosis
--Disc edema
--Retinal/choroidal findings
--Motility disorders

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-- **Proptosis**
-- Disc **edema**
-- Retinal/choroidal findings
-- **Motility** disorders

w/ inflammation  w/o inflammation
The pain of posterior scleritis has three characteristics that should alert you to the diagnosis:

- The pain awakens the pt at night
- If you encounter descriptions such as this in the clinic (or on the OKAP), think posterior scleritis!
The pain of posterior scleritis has three characteristics that should alert you to the diagnosis:

--The pain radiates to the ____________

--

--
Uveitis

Scleritis

Anterior

Posterior

Diffuse

Nodular

w/ inflammation

The pain of posterior scleritis has three characteristics that should alert you to the diagnosis:
--The pain radiates to the **brow**
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--The pain radiates to the brow
--The pain is aggravated by
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w/ inflamm

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If you encounter descriptions such as this in the clinic (or on the OKAP), think posterior scleritis!
An easy-to-obtain imaging study for confirming the diagnosis of posterior scleritis is B-scan ultrasonography.
Uveitis

Scleritis

- Anterior
  - Diffuse
  - Nodular

- Posterior

An easy-to-obtain imaging study for confirming the diagnosis of posterior scleritis is *B-scan ultrasonography*. 

- w/ inflammation
- w/o inflammation
Scleritis

Uveitis

An easy-to-obtain imaging study for confirming the diagnosis of posterior scleritis is **B-scan ultrasonography**. B-scan will reveal choroidal thickening and sub-Tenon's edema. When sub-Tenon's edema involves the space around the optic nerve, the classic T sign finding will result.
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Uveitis

Posterior scleritis: Choroidal thickening; sub-Tenon’s edema
Scleritis

Anterior

Posterior

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W/ inflammation  
W/o inflammation
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Uveitis

Posterior scleritis: T-sign (advance to next slide if you don’t see it)
Uveitis

Posterior scleritis: T-sign
Scleritis requires systemic treatment. Diffuse scleritis might respond to PO NSAIDs, so try them first if not contraindicated.
Uveitis

Scleritis

Anterior

Diffuse

Nodular

Necrotizing

w/ inflammation

w/o inflammation

Posterior

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In the next section we will go through the criteria regarding how one classifies/describes a uveitis with respect to its ocular findings. (Some of this will be a recapitulation of material we’ve already covered.)
In the context of uveitis, what does the acronym SUN stand for?
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The extent of the uveitis, the onset, duration and course of the uveitis, the severity of the uveitis.
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The **location** of the uveitis

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---?
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The **onset, duration** and **course** of the uveitis

The **severity** of the uveitis

What are the four locations?
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The **location** of the uveitis  
--Anterior  
--Intermediate  
--Posterior  
--Panuveitis

The **onset**, **duration** and **course** of the uveitis  
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The **onset, duration** and **course** of the uveitis

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With respect to uveitis: Where is the primary location of inflammation in… **Anterior** uveitis?
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---Onset: Sudden vs insidious
---Duration: Limited vs persistent
---Course: Acute vs recurrent vs chronic

The **severity** of the uveitis
---AC cell grade
---AC flare grade
---Vitreous haze score

A pt has dense AC cell, scant anterior vitreous cell, and cystoid macular edema. Given all three locations are involved, this pt has panuveitis, yes?

With respect to uveitis, where is the primary inflammation located in…

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A pt has dense AC cell, scant anterior vitreous cell, and cystoid macular edema. Given all three locations are involved, this pt has panuveitis, yes?
No. The description clearly suggests the pt has an anterior uveitis with spillover of cell into the anterior vitreous, along with cystoid macular edema caused by the anterior uveitis.

With respect to uveitis, where is the primary inflammation located in…

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No. The description clearly suggests the pt has an anterior uveitis with spillover of cell into the anterior vitreous, along with cystoid macular edema caused by the anterior uveitis. In other words, despite the fact that all three locations are involved, the primary location is anterior, making this an anterior uveitis.

With respect to uveitis, where is the primary inflammation located in…
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The location of the uveitis

- Anterior
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- Panuveitis

The onset, duration and course of the uveitis

- Onset: something vs something
- Duration
- Course

The severity of the uveitis

How are onset, duration and course delineated?
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<td>0.5+</td>
<td>1-5</td>
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<td>1+</td>
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Where/what is Arlt's triangle?

It's a triangle with its apex at the corneal center and base near the inferior border of the cornea.
Uveitis

Diffusely distributed KP

KP concentrated in Arlt’s triangle
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It can be tough to distinguish between nongranulomatous and stellate KP. Here is another distinguishing characteristic:

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Always…

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Always…in Arlt’s triangle
Always…diffuse
Uveitis

Stellate KP

Granulomatous KP
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--Laterality (i.e., whether it is uni- vs bilateral)
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--Iris changes

Broadly, what sorts of iris changes must be looked for?
--Atrophy
--Synechiae
--Nodules
--Heterochromia
# Uveitis

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- Diffuse
- Sectoral
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Sectoral

Diffuse

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Synechiae typically are found in one of two locations--where?

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Uveitis
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**Uveitis**

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Uveitis

Iris synechiae

Posterior

Peripheral anterior
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Iris nodules typically are found in one of three locations--where?
- At the pupillary margin
- The mid-iris
- At the angle
Uveitis

Marginal

Mid-iris

Iris nodules

Near the angle
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In a pt with uveitis and heterochromia, which iris is more likely to be the abnormal one— the lighter iris, or the darker?
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The lighter
Uveitis

Heterochromia iridis in a uveitis pt
Finally, we will look at masquerade syndromes
Masquerade Syndrome refers to entities that mimic immune-mediated dz.
**Uveitis**

**Masquerade Syndrome** refers to entities that mimic immune-mediated dz.

*In other words, these are pts who look like they have a uveitic condition, but do not.*
In other words, these are pts who look like they have a uveitic condition, but do not. It is very important that masquerade syndromes be recognized as such, because their prolonged (mis)diagnosis as uveitis may result in a devastating delay in the treatment of the actual underlying condition.
Uveitis

Masquerade Syndrome refers to entities that mimic immune-mediated dz.

The entities can be broadly divided into nonneoplastic and neoplastic causes.
Uveitis

*Masquerade Syndrome* refers to entities that mimic immune-mediated dz.

The entities can be broadly divided into Nonneoplastic and Neoplastic causes.
Masquerade Syndrome

Neoplastic

Nonneoplastic

Uveitis

Neoplastic causes are subdivided into those stemming from hematologic vs solid neoplasms.
Neoplastic causes are subdivided into those stemming from *hematologic* vs *solid* neoplasms.
Uveitis

Masquerade Syndrome

Neoplastic

Nonneoplastic

Solid

Hematologic cases are subdivided into lymphoid and leukemic causes
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Virtually all PVRLs are non-Hodgkin B-cell lymphomas. The typical PVRL pt is an adult in their 50s-60s. They usually present with complaints of decreased vision and/or floaters. Importantly, many will also manifest evidence of CNS involvement, the most common being changes in behavior or personality. Other, more obvious S/S include seizures, cerebellar signs, hemiparesis and cranial nerve palsies. Confusion, weakness, and memory loss may also occur.

DFE in PVRL typically reveals subretinal infiltrates described as "creamy yellow" in color. The infiltrates can mimic the findings of other, more common conditions (eg, toxoplasmosis). PVRL is diagnosed by finding 'big blue cells' on vitreous biopsy.
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Vitreous biopsy reveals 'big blue cells'.

DFE typically reveals subretinal infiltrates described as "creamy yellow" in color.

Other symptoms include:
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Uveitis

PVRL: Typical white-yellow subretinal infiltrates
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Uveitis

Typical cytology of PVRL cells from the vitreous showing several atypical lymphoid cells with basophilic cytoplasm and large prominent irregular nuclei.
Other hematologic neoplasias can masquerade as well, but are far less common than PVRL.
Uveitis

Masquerade Syndrome

Neoplastic

Nonneoplastic

Solid-tumor cases are subdivided into primary vs metastatic tumors
Uveitis

Masquerade Syndrome

Neoplastic

Solid

Primary

Solid-tumor cases are subdivided into *primary* vs *metastatic* tumors

Nonneoplastic

Hematologic

Leukemic

Primary vitreoretinal lymphoma

Secondary to systemic lymphoma

Lymphoproliferative dz

Lymphoid

Leukemia

Mets
Uveitis

Masquerade Syndrome

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Leukemia

Primary

? ?

Nonneoplastic

The two ocular primaries most likely to masquerade as uveitis are probably not surprising:

- Primary vitreoretinal lymphoma
- Secondary to systemic lymphoma
- Lymphoproliferative dz
The two ocular primaries most likely to masquerade as uveitis are probably not surprising:
Uveitis

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Leukemic

Lymphoid

Primary vitreoretinal lymphoma

Secondary to systemic lymphoma

Lymphoproliferative dz

Leukemia

Primary

Uveal melanoma

Rb

Mets

Likewise, the two most common primaries involved in ‘metastatic masquerade’ are probably not surprising either:
Likewise, the two most common primaries involved in ‘metastatic masquerade’ are probably not surprising either:
Lung and breast masquerades commonly present with bilateral multifocal lesions mimicking uveitis.
Lung and breast masquerades commonly present with bilateral multifocal choroidal lesions mimicking choroiditis.
Uveitis

Bilateral metastatic lung cancer
Uveitis

Bilateral metastatic breast cancer
**Uveitis**

**Masquerade Syndrome**

**Neoplastic**

**Solid**

**Hematologic**

**Lymphoid**

- Primary vitreoretinal lymphoma
- Secondary to systemic lymphoma
- Lymphoproliferative dz

**Nonneoplastic**

- Lung
- Breast

*Lung and breast masquerades commonly present with bilateral multifocal choroidal lesions mimicking choroiditis. An overlying vitritis may be present.*
Uveitis

Masquerade Syndrome

Nonneoplastic

Neoplastic

Solid

Mets

Lung

Breast

Hematologic

Lymphoid

Leukemia

Primary vitreoretinal lymphoma

Secondary to systemic lymphoma

Lymphoproliferative dz

Leukemic leukemia

Primary lymphoma

Lung and breast masquerades commonly present with bilateral multifocal choroidal lesions mimicking choroiditis. An overlying vitritis may be present.
These are the nonneoplastic masquerade entities discussed in the *Uveitis* book.
Uveitis

Masquerade Syndrome

Nonneoplastic

Neoplastic

Hematologic

Lymphoid

Leukemic

Leukemia

Primary vitreoretinal lymphoma

Secondary to systemic lymphoma

Lymphoproliferative dz

Primary

Uveal melanoma

Rb

Mets

Lung

Breast

Retinitis pigmentosa

Ocular ischemic syndrome

Chronic rhegmatogenous RD

Intraocular foreign body

Pigment dispersion syndrome

Juvenile xanthogranuloma

These are the nonneoplastic masquerade entities discussed in the *Uveitis* book.
Ocular ischemic syndrome (OIS) is a constellation of ocular abnormalities stemming from chronic hypoperfusion of the globe. The classic cause is carotid stenosis ipsilateral to the eye in question. The typical pt is an elderly vasculopathic male.

Four findings, common in OIS, can (mis)lead one to conclude the pt has uveitis:
- AC cell and flare
- Low IOP
- Neovascularization of the iris and/or angle
- Cataract more advanced on that side
Uveitis

**Masquerade Syndrome**

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The typical pt is an *age, health status, gender*. 

- **Leukemic**
  - **Primary vitreoretinal lymphoma**
  - **Secondary to systemic lymphoma**
- **Lymphoproliferative dz**
- **Solid**
- **Primary vitreoretinal lymphoma**
- **Secondary to systemic lymphoma**
- **Lymphoproliferative dz**
- **Uveal melanoma**
- **Rb**

**RP**

- **Chronic rhegmatogenous RD**
- **Intraocular foreign body**
- **Pigment dispersion syndrome**
- **Juvenile xanthogranuloma**
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--?
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**Uveitis**

**Masquerade Syndrome**

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Four findings, common in OIS, can (mis)lead one to conclude the pt has uveitis:

--AC…*cell and flare*

--IOP is…

--?

--?

---

**Nonneoplastic**

- RP
- Chronic rhegmatogenous RD
- Intraocular foreign body
- Pigment dispersion syndrome
- Juvenile xanthogranuloma

**Lymphoid**

- Leukemic
  - Leukemia
- Rb
- Primary vitreoretinal lymphoma
- Secondary to systemic lymphoma
- Lymphoproliferative dz
Uveitis

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--IOP is…low
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Masquerade Syndrome

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Lymphoid

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--IOP is…**low**

--Neovascularization of the…**iris and/or angle**

--Ipsilateral cataract is…
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--AC…cell and flare
--IOP is…low
--Neovascularization of the…iris and/or angle
--Ipsilateral cataract is…more advanced
The hallmark of PDS is the liberation of pigment from the posterior aspect of the iris. This pigment subsequently migrates into the anterior chamber, where the pigment granules can be mistaken for inflammatory cells. Typically, retroillumination of the iris will reveal transillumination defects with a radial orientation.
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Uveitis

PDS: Radial transillumination defects