Before you begin: This is a big topic, and big topics beget big slide-sets. There’s a natural break around slide 210; I placed a break time! slide at that point.
Uveitis

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Think of the eye as being composed of three layers or ‘tunics.’ The [Fibrous tunic] and [Sclera] and [Cornea] comprise the tough, outer tunic.
Think of the eye as being composed of three layers or ‘tunics.’ The sclera and cornea comprise the tough, outer tunic.
Uveitis

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Think of the eye as being composed of three layers or ‘tunics.’ The sclera and cornea comprise the tough, outer tunic. The RPE and neurosensory retina comprise the innermost ‘retinal’ tunic. In between these two is the highly vascular, highly pigmented tunic known as the uvea. (The word uvea derives from the Greek word for ‘grape’—an acknowledgement of the deep-purple color characteristic of most of the uvea.)
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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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With respect to uveitis, the most important aspects of a pt’s demographics are three things:
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With respect to uveitis, the most important aspects of a pt’s demographics are *age*, *gender*, and *ethnicity*.
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With respect to uveitis, the most important aspects of a pt’s demographics are **age**, **gender**, and **ethnicity**.

Other important demographic factors include:

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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)

With respect to uveitis, the most important aspects of a pt’s demographics are age, gender, and ethnicity.
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**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

**Anterior uveitis**

*If cell is located…*

- Exclusively in the AC: *It is called:*  
  - Iritis
- Exclusively in the AVit: *It is called:*  
  - ?

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Anterior uveitis

*If cell is located…*

- Exclusively in the AC
  - It is called: Iritis
- Exclusively in the AVit
  - It is called: Anterior cyclitis
Uveitis

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

**Anterior uveitis**

*If cell is located…*

- **Exclusively in the AC**
  - *It is called:* Iritis
- **In the AC and the anterior vitreous**
  - *It is called:* ?
- **Exclusively in the AVit**
  - *It is called:* Anterior cyclitis
**Uveitis**

**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 **three words** +/- the **two words**.
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
- Not idiopathic

*How we divvy up IU*
Intermediate uveitis

If condition is...

- Idiopathic
- Not idiopathic

How we divvy up IU

It is called:

? 

In intermediate uveitis, the primary location of inflammation is the main vitreous cavity, +/- the peripheral retina.
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Uveitis

Posterior uveitis

If inflammation is located…

- Exclusively in the choroid
- In both the choroid and the retina
- Exclusively in the retina

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

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
  - Retinitis
  - and ONH

In posterior uveitis, the site of inflammation is the retina and/or choroid (the optic nerve head can be involved too)
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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**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

Chorioretinitis or Retinochoroiditis

Neuroretinitis and ONH
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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‘**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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‘**Lyme uveitis** is granulomatous, and CNS manifestations are very common.’
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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…
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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.
In surface disorders (eg, conjunctivitis), redness is either distributed uniformly across the eye, or it tapers off near the limbus.
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Uveitis

Aqueous cells and flare

- cells
- flare
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The *Uveitis* book employs an organizational tree on which it hangs the common causes of anterior uveitis. The first branch point in this tree is whether the inflammation is **granulomatous** or **nongranulomatous**.
Granulomatous Nongranulomatous

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Uveitis

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*This is key:* In clinical use the term granulomatous refers to the slit-lamp appearance of the KP, not to the underlying histology of the condition.
Uveitis

Anterior Uveitis

Granulomatous

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, i.e., they typically do not present as an isolated anterior uveitis.)

Nongranulomatous
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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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(FYI: If a uveitis eventually relapses but is quiescent off-treatment for longer than three months, it is termed a **recurrent** uveitis.)
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Finally, the acute uveitides are divided into those that present unilaterally vs those that tend to present bilaterally.
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.
Acute Chronic

Unilateral Bilateral

Granulomatous
- TB
- Sarcoid
- Syphilis
- HSV
- VKH
- Toxoplasmosis
- Lyme

Nongranulomatous

Acute
- HLA-B27 dz
- Posner-Schlossman
- HSV/VZV
- Sarcoid
- Syphilis
- TB

Bilateral
- TINU
- Behçet
- Drug rxn
- Leptospirosis
- IBD/PA
- Sarcoid
- Syphilis
- TB

Chronic
- JIA
- FHI
- IBD/PA
- Sarcoid
- Syphilis
- TB

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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.
Rule of thumb:
Syphilis, sarcoid and TB are on the DDx for every pt with any form of uveitis!
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Next let’s look at intermediate uveitis
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Uveitis

The vitreous base

- Ciliary body
- Peripheral retina

Vitreous base
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IU tends to be a disease of young people—teens through 40 or so. 

**Uveitis**

- **Anterior**
- **Intermediate**
- **Posterior**
- **Panuveitis**
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IU tends to be a dz of young people—teens through 40 or so. It is uni- or bilateral in most (80%) cases.
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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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Uveitis

- Anterior
- Intermediate
  - Pars planitis
  - Intermediate uveitis
- Posterior
- Panuveitis

The entities most likely to produce IU:
Uveitis

- Anterior
- Intermediate
  - Pars planitis
  - Intermediate uveitis

- Posterior
- Panuveitis

The entities most likely to produce IU:
- MS
- Lyme
- Toxocariasis
- Sarcoid
- Syphilis
- TB
Uveitis

- Anterior
  - Pars planitis
  - Intermediate uveitis

- Intermediate
  - MS
  - Lyme
  - Toxocariasis
  - Sarcoid
  - Syphilis
  - 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.
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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%.
**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**

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**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**.
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Uveitis

**Posterior uveitis**

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

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

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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 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 *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).
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
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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 'headlight in the fog.'
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Uveitis

Ocular toxoplasmosis: ‘Headlight in the fog’
Uveitis

- Anterior
- Intermediate
- Posterior

Panuveitis

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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- Sympathetic ophthalmia
- Vogt-Koyanagi-Harada
- Behçet syndrome
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**Noninfectious**
- Sarcoid
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**Infectious**
- Syphilis
- TB
- Lyme
- Leptospirosis
- Whipple dz
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Note that these three appear yet again

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- Sympathetic ophthalmia
- Vogt-Koyanagi-Harada
- Behçet syndrome
- 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 **causal event A**, **causal event B**, or **causal event C**.
The *Uveitis* book defines **endophthalmitis** as an inflammatory process involving both the AC and vitreous cavities that is 2ndry to bacterial or fungal infection. **Endophthalmitis** can be **posttraumatic**, **postoperative** or **endogenous**.
Endogenous Endophthalmitis

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**Endogenous endophthalmitis** involves hematogenous spread of infection from a remote location to the eye. It is uncommon.
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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 2ndry to bacterial or fungal infection. Endophthalmitis can be **posttraumatic, postoperative** or **endogenous**.

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Endogenous Endophthalmitis

Bacterial

Fungal

As mentioned, endogenous endophthalmitis can be bacterial or fungal. **Bacterial endophthalmitis** presents with the expected ocular signs of **pain**, **redness**, and **decreased vision**. Additional ocular signs include **periorbital/lid edema**, **a dense AC reaction** (often with **hypopyon**), and **vitreous inflammation**.
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Some classic bacterial pathogen associations in endogenous endophthalmitis:

--Endocarditis: *
--Skin infections
--IVDU
--Liver abscess
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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

*Candida* endophthalmitis: Choroidal lesions
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Endogenous Endophthalmitis

*Candida* endophthalmitis: Retinal involvement
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
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Some classic fungal pathogen associations in endogenous endophthalmitis:

--Chronic indwelling lines/catheters: ?
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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
(This is a good point in the set to take a break)
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.
Uveitis

Scleritis

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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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...
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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...
Uveitis

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
- 
- 

Uveitis

Scleritis

Anterior

Posterior
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
--
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--Sclera has a violaceous hue
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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*
Diffuse anterior scleritis
Nodular anterior scleritis
Uveitis

Scleritis

Anterior

Diffuse

Nodular

Necrotizing

Posterio

Necrotizing anterior scleritis comes in two forms:
Scleritis

Anterior

- Diffuse
- Nodular

Posterior

Necrotizing

- w/ inflammation
- w/o inflammation

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

Necrotizing anterior scleritis with inflammation
Contrary to the implications of the name, inflammation is present in necrotizing scleritis w/o inflammation.
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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**
- **Disc edema**

w/ inflammation  w/o inflammation
Posterior scleritis: Proptosis
Uveitis

Scleritis

Anterior

Diffuse

Nodular

w/ inflammation

w/o inflammation

Posterior

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

--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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**Scleritis**

Anterior

Posterior

Diffuse

Nodular Necrotizing

w/ inflammation

w/o inflammation
Posterior scleritis OD: Optic nerve edema
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
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:

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

---

**Disorders**

**Function**
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
The pain of posterior scleritis has three characteristics that should alert you to the diagnosis:

--
--
--
The pain of posterior scleritis has three characteristics that should alert you to the diagnosis:

--The pain radiates to the **brow**
--
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 of posterior scleritis has three characteristics that should alert you to the diagnosis:
--The pain radiates to the **brow**
--The pain is aggravated by **two words**
--
The pain of posterior scleritis has three characteristics that should alert you to the diagnosis:
--The pain radiates to the **brow**
--The pain is aggravated by **eye movements**

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

--The pain radiates to the **brow**
--The pain is aggravated by **eye movements**
--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 **brow**
--The pain is aggravated by **eye movements**
--The pain **awakens the pt at night**
The pain of posterior scleritis has three characteristics that should alert you to the diagnosis:

- The pain radiates to the brow
- The pain is aggravated by eye movements
- The pain awakens the pt at night

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-ultrasound**. B-ultrasound 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.
Uveitis

Scleritis

Anterior

Posterior

Diffuse

Nodular

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

Scleritis

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Anterior

Posterior

Diffuse

Nodular

w/ inflammation

w/o inflammation
Uveitis

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

Anterior

Posterior

Diffuse

Nodular

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An easy-to-obtain imaging study for confirming the diagnosis of posterior scleritis is \textit{B-scan ultrasonography}. \textit{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 \textit{T sign} finding will result.
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.
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**Scleritis**

- **Anterior**
  - Diffuse
  - Nodular
- **Posterior**
  - Necrotizing
    - w/ inflammation
    - w/o inflammation

**Scleritis requires systemic treatment.** Diffuse scleritis might respond to PO NSAIDs, so try them first if not contraindicated. For the others, PO steroids are usually the first-line med, although NSAIDs may be tried. Immunomodulatory therapy is often required.
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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.)
Uveitis

In the context of uveitis, what does the acronym SUN stand for?
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Standardization of Uveitis Nomenclature, a working group appointed by the International Ocular Inflammation Society
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To standardize uveitis nomenclature

The SUN classification system is based on three sets of criteria. What are they?

The [location], The [onset, duration, and course], and The [severity] of the uveitis
In the context of uveitis, what does the acronym SUN stand for? Standardization of Uveitis Nomenclature, a working group appointed by the International Ocular Inflammation Society

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The location of the uveitis
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

The **severity** of the uveitis

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- Anterior?
  - Intermediate
  - Posterior
  - Panuveitis

The onset, duration and course of the uveitis

The severity of the uveitis

With respect to uveitis: Where is the primary location of inflammation in…
Anterior uveitis?
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--- Anterior
--- Intermediate
--- Posterior
--- Panuveitis

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The severity of the uveitis

With respect to uveitis: Where is the primary location of inflammation in…

Anterior uveitis? The anterior chamber (although cell 'spillover' into the anterior vitreous may occur)
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--Anterior
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The onset, duration and course of the uveitis

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- Anterior
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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? The anterior chamber (although cell ‘spillover’ into the anterior vitreous may occur)
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- Intermediate
- **Posterior**?
- Panuveitis

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

**The severity of the uveitis**

With respect to uveitis: Where is the primary location of inflammation in…

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--- Anterior  
--- Intermediate  
--- Posterior  
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The **onset**, **duration** and **course** of the uveitis

The **severity** of the uveitis

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  - Anterior
  - Intermediate
  - Posterior
  - Panuveitis?

- **Onset, duration** and course of the uveitis
- **Severity** of the uveitis

*With respect to uveitis: Where is the primary location of inflammation in…*
- **Anterior** uveitis? The anterior chamber (although cell ‘spillover’ into the anterior vitreous may occur)
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- **Panuveitis**? All three locations are equally involved
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- Anterior
- Intermediate
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The **onset, duration** and **course** of the uveitis

The **severity** of the uveitis

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…

**Anterior** uveitis? The anterior chamber (although cell 'spillover' into the anterior vitreous may occur)

**Intermediate** uveitis? The vitreous, peripheral retina and/or pars plana

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**The location of the uveitis**
- Anterior
  -- Intermediate
  -- Posterior
- Intermediate
- Posterior
- Panuveitis

**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

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…

**Anterior uveitis**? The anterior chamber (although cell 'spillover' into the anterior vitreous may occur)
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- AC cell grade
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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. 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…
**Anterior uveitis?** The anterior chamber (although cell 'spillover' into the anterior vitreous may occur)
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- The location of the uveitis
  -- Anterior
  -- Intermediate
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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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How are onset, duration and course delineated?
**Uveitis**

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---Anterior
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**With respect to uveitis, what is the difference between…**
Sudden vs insidious onset?
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With respect to uveitis, what is the difference between…

Sudden vs insidious onset? **Sudden-onset uveitis** presents with abrupt development of symptoms (pain, photophobia) and signs (injection); **insidious uveitis** is largely sign- and symptom-free
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Limited vs persistent duration?
Uveitis

In the context of uveitis, what does the acronym SUN stand for?
Standardization of Uveitis Nomenclature, a working group appointed by the International Ocular Inflammation Society

What was the task of the SUN Working Group?
To standardize uveitis nomenclature

The SUN classification system is based on three sets of criteria. What are they?
The location of the uveitis
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--Intermediate
--Posterior
--Panuveitis

The onset, duration and course of the uveitis
--Onset: Sudden vs insidious
--Duration: Limited vs persistent
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By counting the number of WBCs visible within a # x # mm slit-lamp beam.
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- Diffuse
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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

In addition to location, onset/duration/course, and severity, other characteristics of the uveitis are also important.
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Diffusely distributed KP

KP concentrated in Arlt’s triangle
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- The character of keratic precipitates (KP) if present

Broadly, what two characteristics of KP are important?
- Their appearance and their distribution

It can be tough to distinguish between nongranulomatous and stellate KP. Here is another distinguishing characteristic:

The appearance of KP is most often described with one of three terms. What are they?
- Granulomatous
- Nongranulomatous
- Stellate

Always…
Always…
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Always... in Arlt's triangle
Always... diffuse
Uveitis

Stellate KP

Granulomatous KP
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**Uveitis**

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In addition to location, onset/duration/course, and severity, what other aspects of presentation are important in diagnosing uveitis?

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Broadly, what is the character of atrophic changes?

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The character of atrophic changes is often described with one of two terms. What are they?

--Diffuse
--Sectoral
Uveitis

Sectoral

Diffuse

Iris atrophy
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Laterality (ie, laterality)

**Broadly, what sorts of iris changes must be looked for?**

---

**Synechiae**

---

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---

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--The character of keratic precipitates (KP) if present
--Iris changes

Broadly, what sorts of iris changes must be looked for?

--Atrophy
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**Synechiae typically are found in one of two locations--where?**
--At the pupillary margin (these are called *posterior synechiae*)
--At the angle (these are called *peripheral anterior synechiae*).
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**Broadly, what sorts of iris changes must be looked for?**

- Iris changes
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Iris synechiae

Posterior

Peripheral anterior

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--Laterality (ie, laterality)
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Broadly, what sorts of iris changes must be looked for?

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Iris nodules typically are found in one of three locations--where?

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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
Heterochromia iridis in a uveitis pt
Uveitis

Masquerade Syndrome

Finally, we will look at masquerade syndromes
Uveitis

*Masquerade Syndrome* refers to entities that mimic immune-mediated dz.
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In other words, these are pts who look like they have a uveitic condition, but do not.
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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.
The entities can be broadly divided into Masquerade Syndrome, which refers to entities that mimic immune-mediated dz. and Neoplastic and Nonneoplastic causes.
Uveitis

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

The entities can be broadly divided into *Nonneoplastic* and *Neoplastic* causes.
Neoplastic causes are subdivided into those stemming from hematologic vs solid neoplasms.
Uveitis

Masquerade Syndrome

Neoplastic

Solid

Hematologic

Neoplastic causes are subdivided into those stemming from *hematologic* vs *solid* neoplasms.
Hematologic cases are subdivided into lymphoid and leukemic causes.
Hematologic cases are subdivided into *lymphoid* and *leukemic* causes.
The most common entity to masquerade as intraocular uveitis is **primary vitreoretinal lymphoma (PVRL)**.
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Virtually all PVRLs are non-Hodgkin B-cell lymphomas.

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.
The most common entity to masquerade as intraocular uveitis is **primary vitreoretinal lymphoma** (PVRL).

**Primary vitreoretinal lymphoma**

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.

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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.
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Uveitis

PVRL: Typical white-yellow subretinal infiltrates
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PVRL is diagnosed by finding three words on vitreous biopsy.

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The most common entity to masquerade as intraocular uveitis is primary vitreoretinal lymphoma (PVRL).
Uveitis

Typical cytology of PVRL cells from the vitreous showing several atypical lymphoid cells with basophilic cytoplasm and large prominent irregular nuclei.
Uveitis

**Masquerade Syndrome**

- Neoplastic
  - Solid
  - Hematologic
    - Leukemic
      - Leukemia
    - Lymphoid
      - Primary vitreoretinal lymphoma
      - Secondary to systemic lymphoma
      - Lymphoproliferative dz

Other hematologic neoplasias can masquerade as well, but are **far less common** than PVRL.
Uveitis

Masquerade Syndrome

Nonneoplastic

Neoplastic

Solid

??

Solid-tumor cases are subdivided into primary vs metastatic tumors

Hematologic

Leukemic

Leukemia

Lymphoid

Primary vitreoretinal lymphoma

Secondary to systemic lymphoma

Lymphoproliferative dz
Masquerade Syndrome

Neoplastic

Solid-tumor cases are subdivided into primary vs metastatic tumors

Hematologic

Primary vitreoretinal lymphoma
Secondary to systemic lymphoma
Lymphoproliferative dz

Leukemic

Leukemia

Primary

Mets

Nonneoplastic

Solid
Masquerade Syndrome

Neoplastic

Solid

Mets

Nonneoplastic

Hematologic

Leukemic

Primary

Primary vitreoretinal lymphoma

Secondary to systemic lymphoma

Lymphoproliferative dz

Leukemia

The two ocular primaries most likely to masquerade as uveitis are probably not surprising:
The two ocular primaries most likely to masquerade as uveitis are probably not surprising:
Likewise, the two most common primaries involved in ‘metastatic masquerade’ are probably not surprising either:
Uveitis

Masquerade Syndrome

Neoplastic

Nonneoplastic

Hematologic

Solid

Primary

Uveal melanoma

RB

Mets

Lung

Breast

Likewise, the two most common primaries involved in ‘metastatic masquerade’ are probably not surprising either:

Primary vitreoretinal lymphoma
Secondary to systemic lymphoma
Lymphoproliferative dz
Uveitis

**Masquerade Syndrome**

Neoplastic

Solid

Mets

- Lung
- Breast

Hematologic

Lymphoid

Leukemic

Primary vitreoretinal lymphoma

Secondary to systemic lymphoma

Lymphoproliferative dz

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

Bilateral metastatic lung cancer
Uveitis

Bilateral metastatic breast cancer
Lung and breast masquerades commonly present with bilateral multifocal choroidal lesions mimicking choroiditis. An overlying vitritis may be present.
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.

- **Hematologic**
  - Leukemic
    - Leukemia
  - Lymphoid
    - Primary vitreoretinal lymphoma
    - Secondary to systemic lymphoma
    - Lymphoproliferative dz

- **Primary**
  - Uveal melanoma
  - Rb

- **Mets**
  - Lung
  - Breast
These are the nonneoplastic masquerade entities discussed in the *Uveitis* book.

- **Hematologic**
  - Leukemic
    - Leukemia
- **Lymphoid**
  - Primary vitreoretinal lymphoma
  - Secondary to systemic lymphoma
  - Lymphoproliferative dz
- **Neoplastic**
- **Nonneoplastic**
  - Retinitis pigmentosa
  - Ocular ischemic syndrome
  - Chronic rhegmatogenous RD
  - Intraocular foreign body
  - Pigment dispersion syndrome
  - Juvenile xanthogranuloma

**Mets**
- Lung
- Breast

**Primary**
- Uveal melanoma
- Rb

**Masquerade Syndrome**
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
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**Primary vitreoretinal lymphoma**

**Secondary to systemic lymphoma**

**Lymphoproliferative dz**

**Leukemia**

**Leukemic**

**Ocular melanoma**

**Uveal melanoma**

**Rb**

**RP**

**Chronic rhegmatogenous RD**

**Intraocular foreign body**

**Pigment dispersion syndrome**

**Juvenile xanthogranuloma**

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--?
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**OIS**

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

**Lymphoid**

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

**Leukemic**

- Leukemia

**Leukemic**

**Nonneoplastic**

**Masquerade Syndrome**
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**Leukemic**
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**Lymphoid**
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- Secondary to systemic lymphoma
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**Nonneoplastic**
- RP
- Chronic rhegmatogenous RD
- Intraocular foreign body
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**Masquerade Syndrome**

**Uveitis**
**Uveitis**

**Masquerade Syndrome**

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--Ipsilateral cataract is...
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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... 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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- **Lymphoid**
  - Primary vitreoretinal lymphoma
  - Secondary to systemic lymphoma
  - Lymphoproliferative dz

- **Leukemic**
  - Leukemia

- **Leukemic**

- **Neoplastic**

- **Nonneoplastic**

- **RP**
- **OIS**
- Chronic rhegmatogenous RD
- IOFB
- **Pigment dispersion syndrome**
- Juvenile xanthogranuloma

- **Polypoidal choroidal vasculopathy**

- **Uveal melanoma**

- **Rb**

- **Uveitis**

- **Masquerade Syndrome**
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Uveitis

PDS: Radial transillumination defects