

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.

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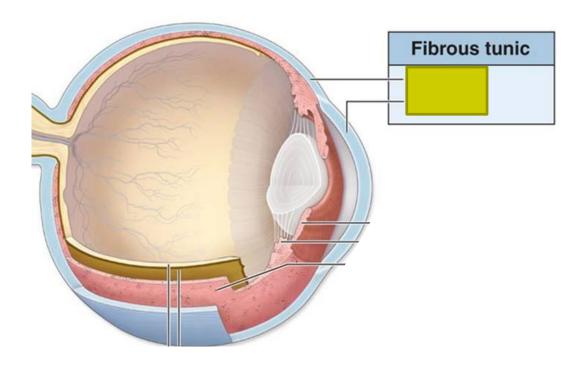


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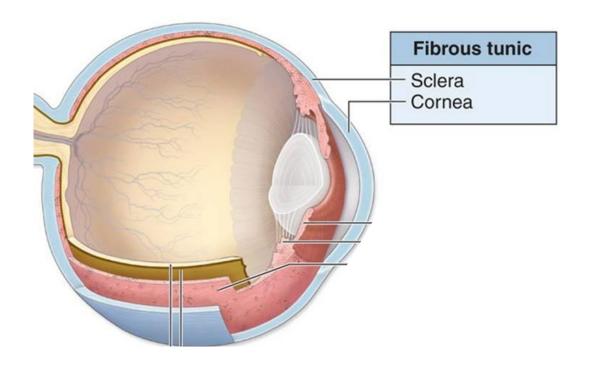






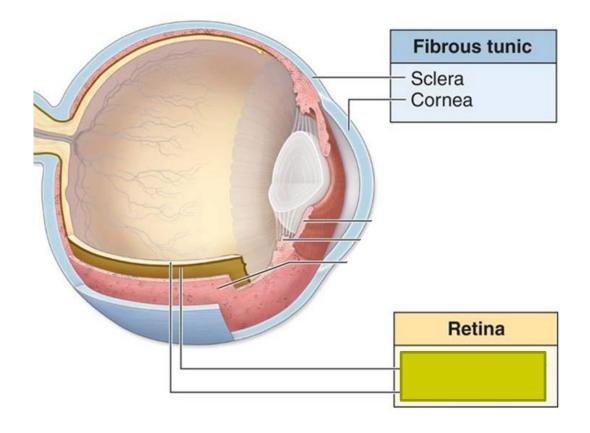
Think of the eye as being composed of three layers or 'tunics.' The comprise the tough, outer tunic.





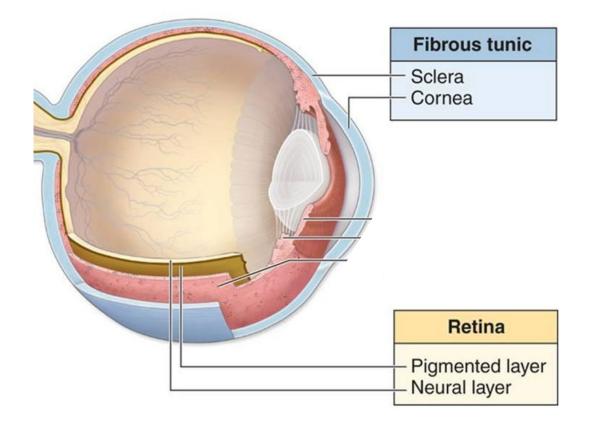


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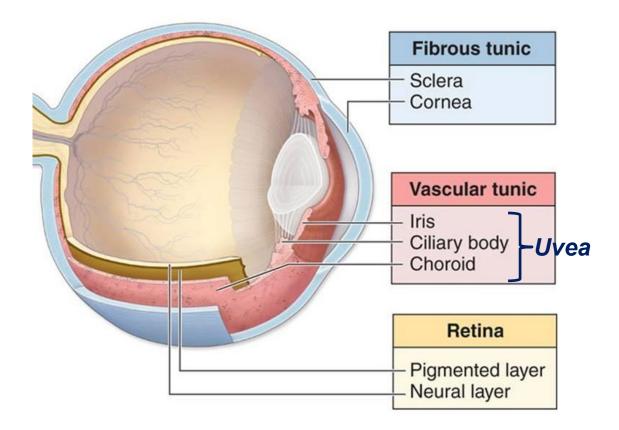


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Think of the eye as being composed of three layers or 'tunics.' The sclera and cornea comprise the tough, outer tunic. The RPE and neurosensory retina comprise the innermost 'retinal' tunic. In between these two is the highly vascular, highly pigmented 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.)







Uveal tissue. Note the deep purple hue

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I say 'technically speaking' because in clinical practice, *uveitis* refers to inflammatory processes involving most **non**-uveal segments of the eye as well, including (but not limited to) the retina, optic nerve, sclera, etc.



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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:

- Because of the --Geographic history, ie, where they have lived or visited
- learning how --Social history (eg, sexual behaviors; dietary habits; IVDU)
  - ---Vocational/avocational activities (eg, exposure to farm animals)

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

--Constitutional: Fever; night sweats; weight loss

--Neuro: HA; cranial neuropathies; hearing loss; cognitive changes I say 'te

-- ENT: Oral ulcers: sinusitis: ear or nose deformities

inflamm --Skin: Rashes; poliosis, madarosis; vitiligo; erythema nodosum as

well, inc --Pulmonary: SOB/DOE; cough; hemoptysis

--Cardiac; Arrythmias; pericarditis symptoms

In many --GI: Diarrhea; ulcers

disease --GU; Genital lesions; discharge; nephritis symptoms

of a sys -- Musculoskeletal: Arthralgias; low back pain

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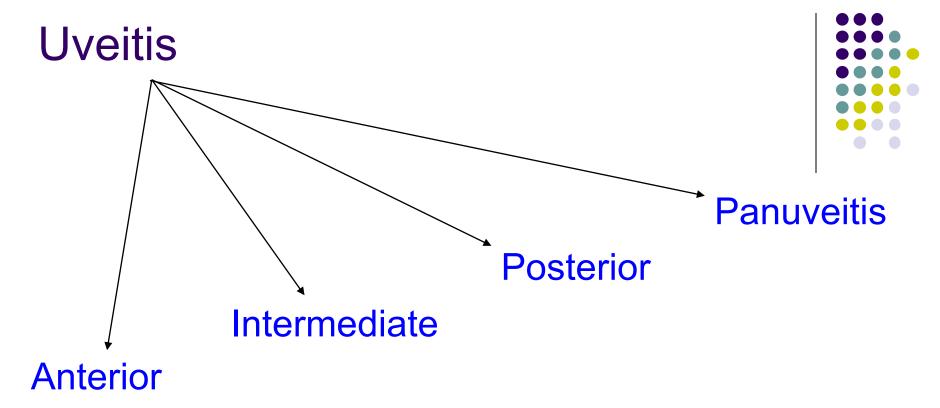
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Let's drill down on the different ways uveitis manifests within the eye. One very important manifestation issue is location, ie, the portion or segment of the eye that's involved.

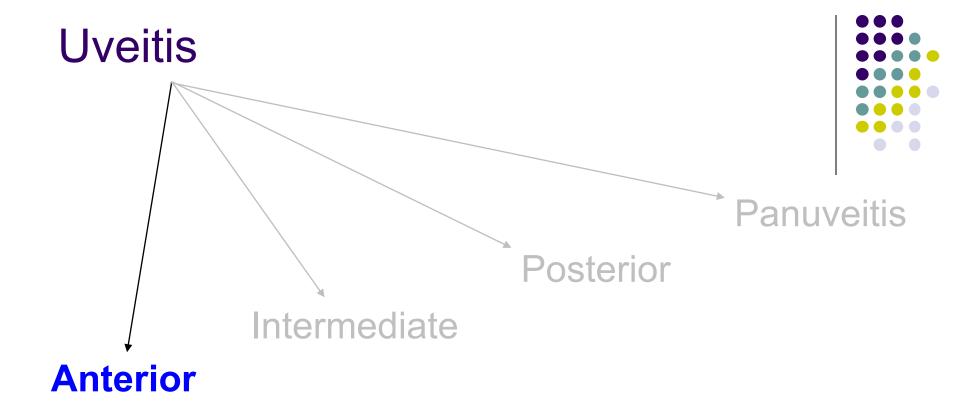
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# Uveitis ?

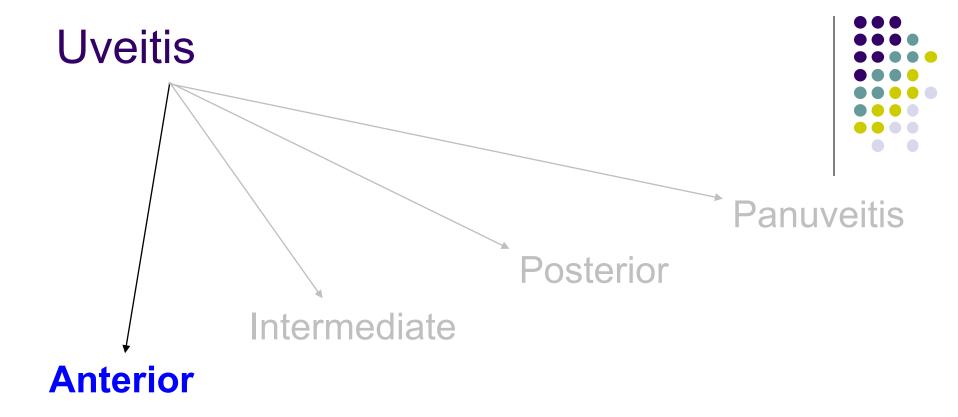
What are the four basic anatomic locations for uveitis?

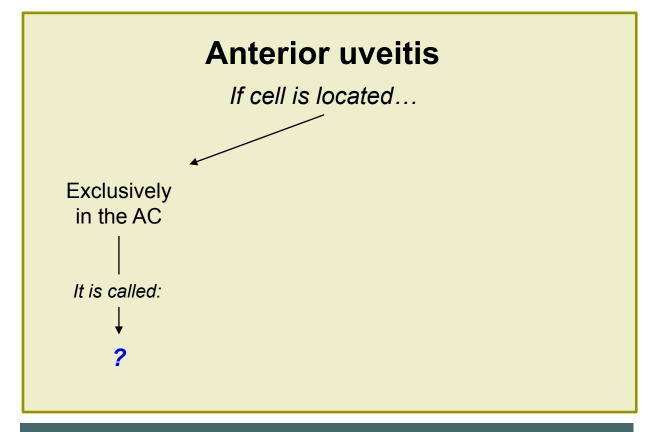


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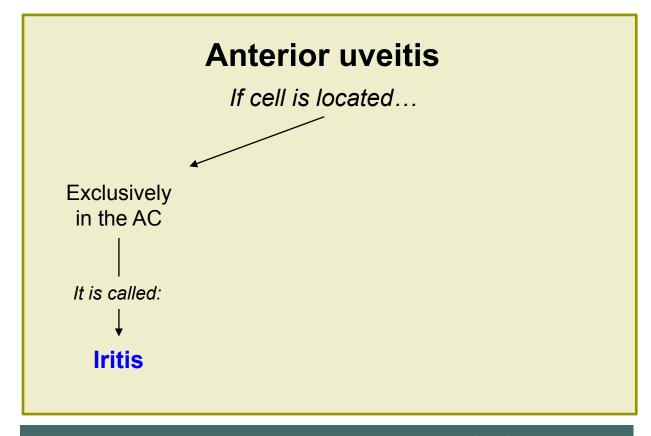
In **anterior uveitis**, the primary location of inflammation is the two words and/or two words

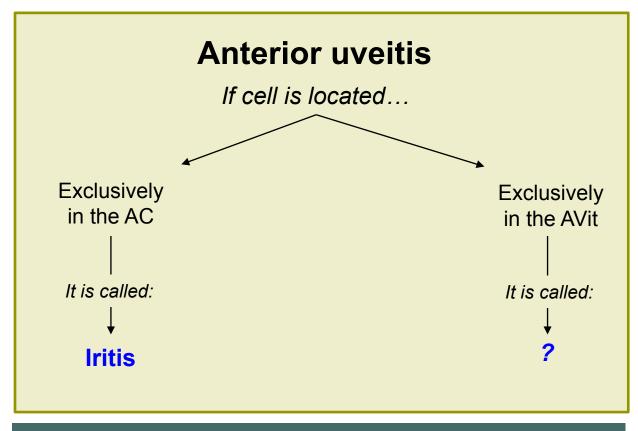




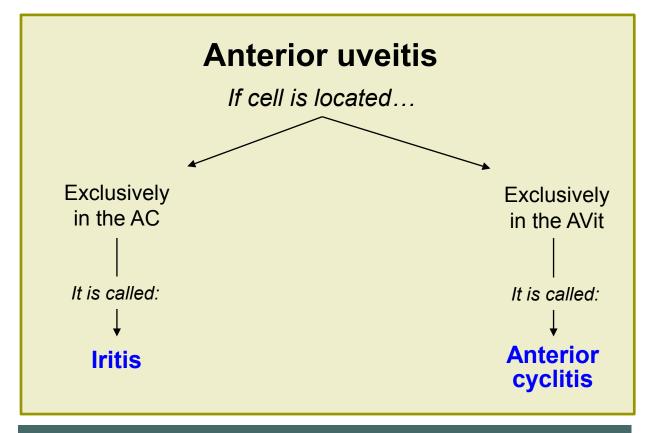




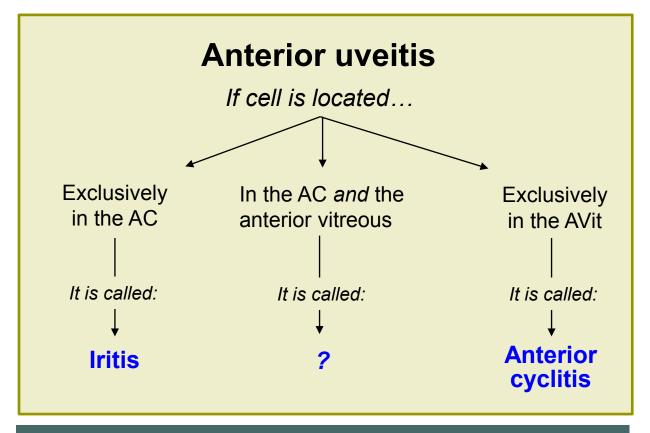




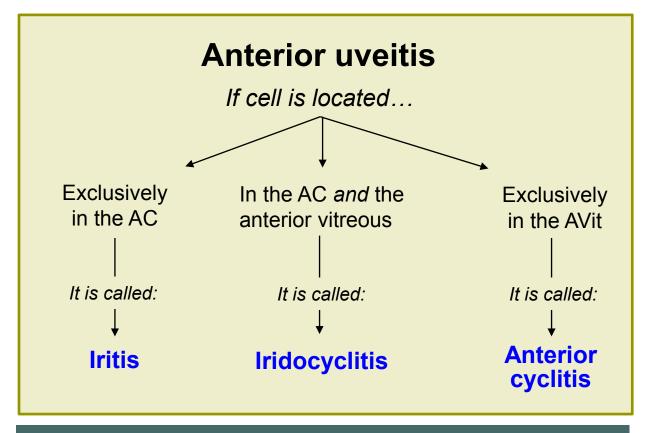




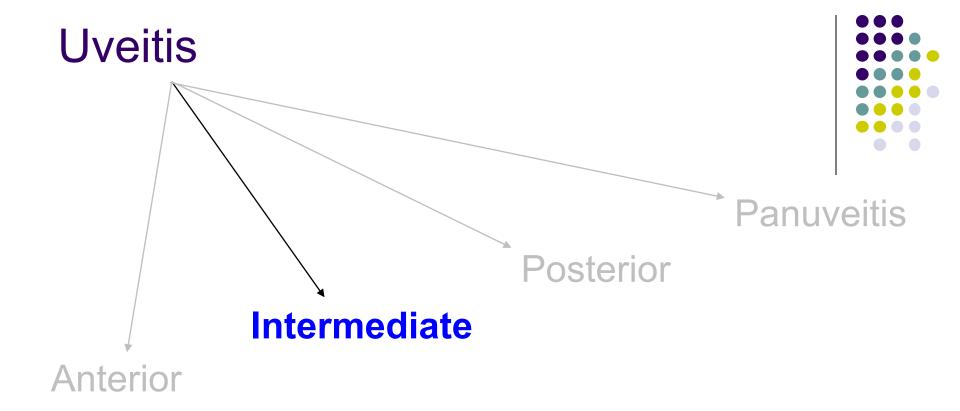




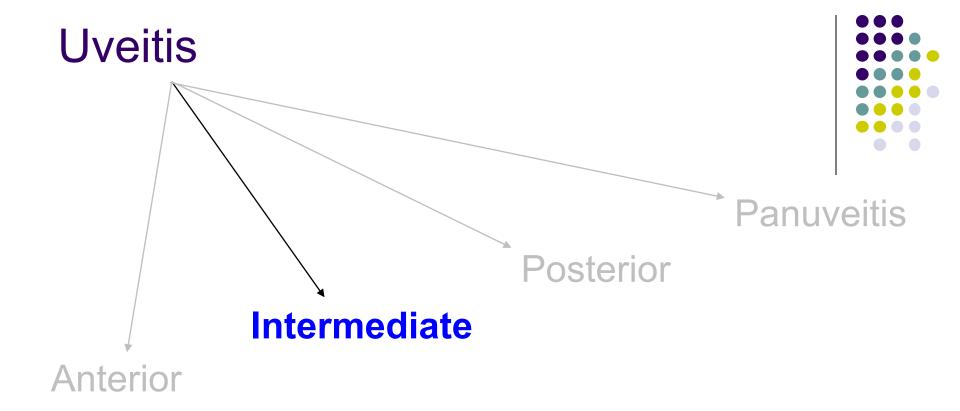


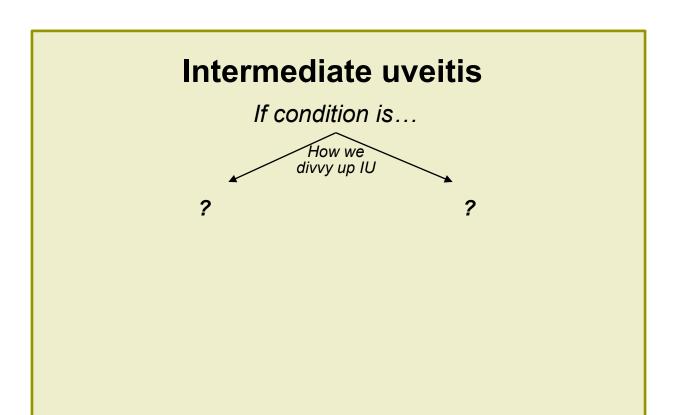






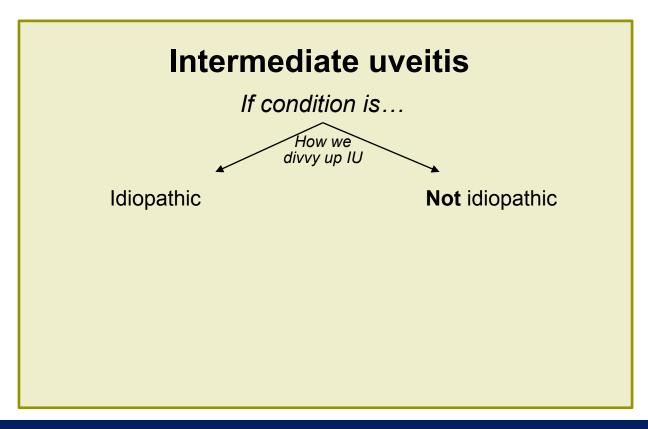
In **intermediate uveitis**, the primary location of inflammation is the three words +/- the two words

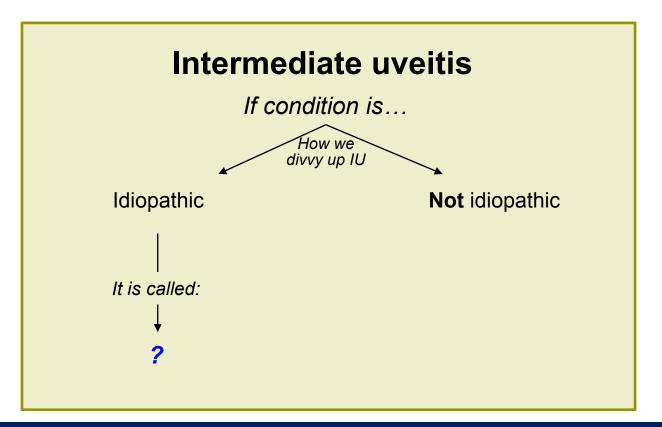




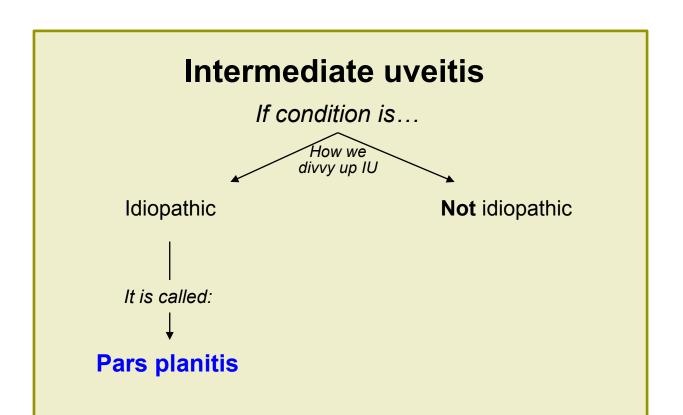




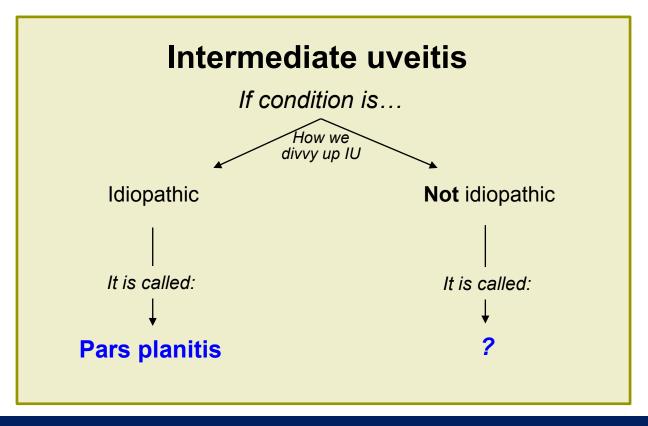




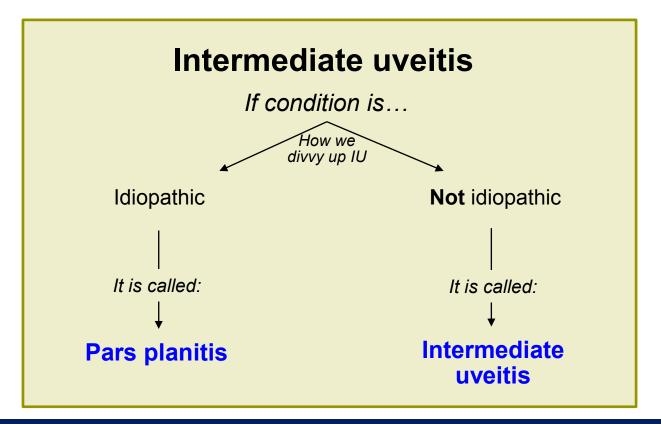




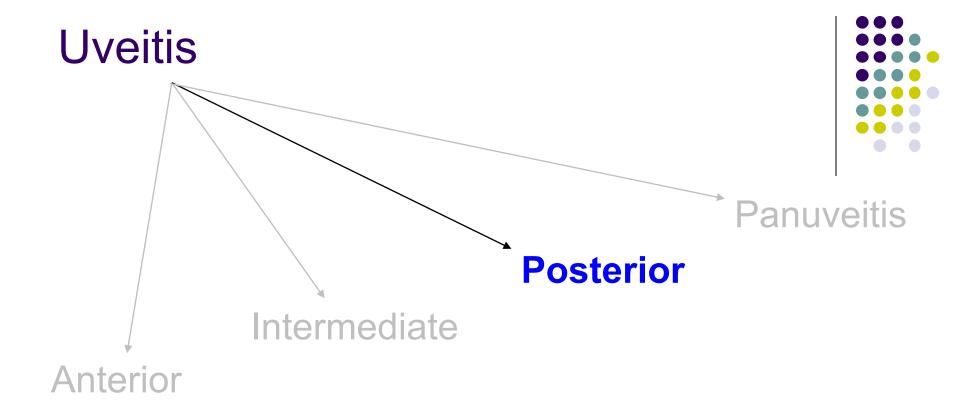




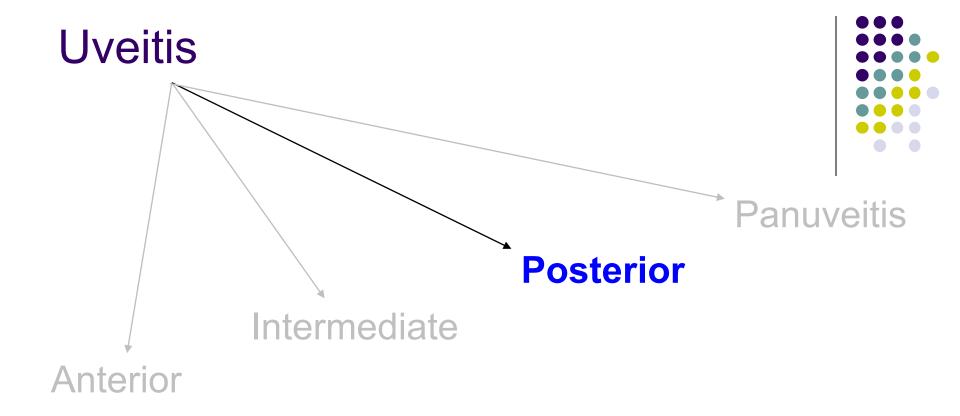




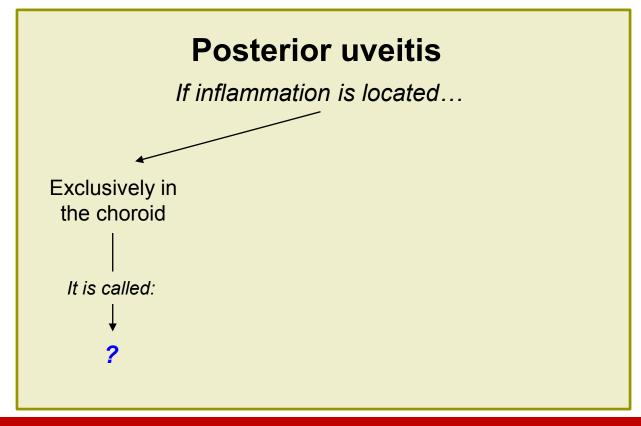




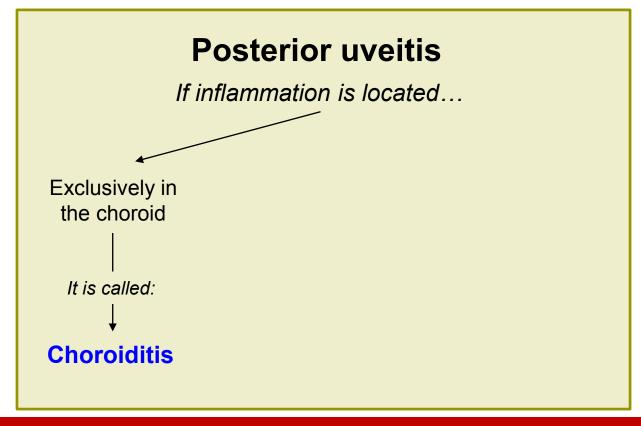
In **posterior uveitis**, the site of inflammation is the and/or (the three words can be involved too)



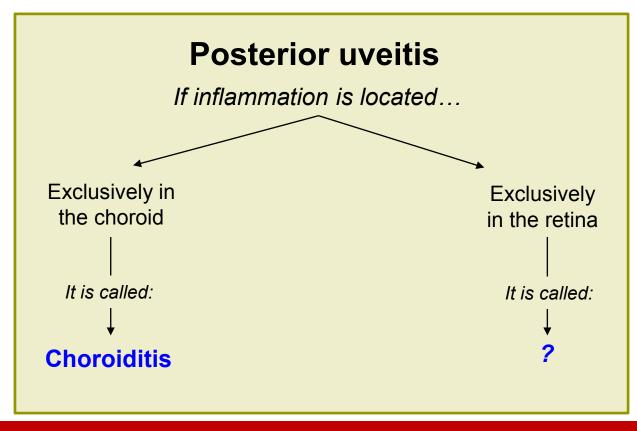




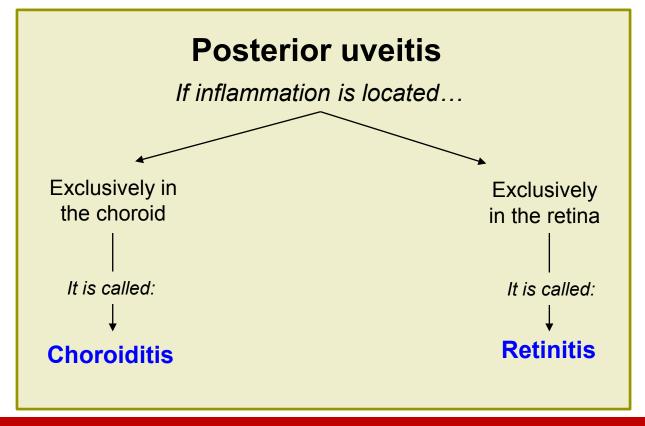


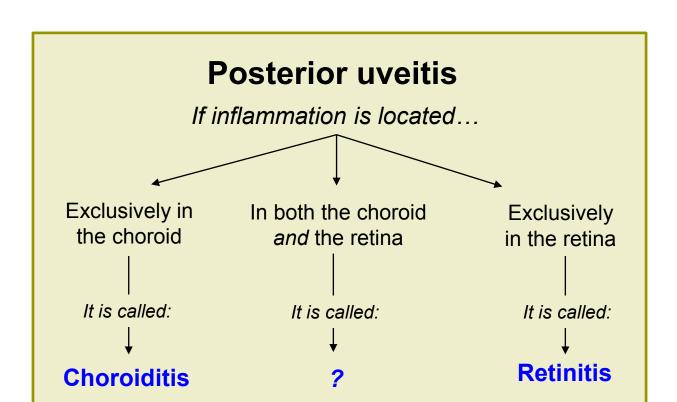




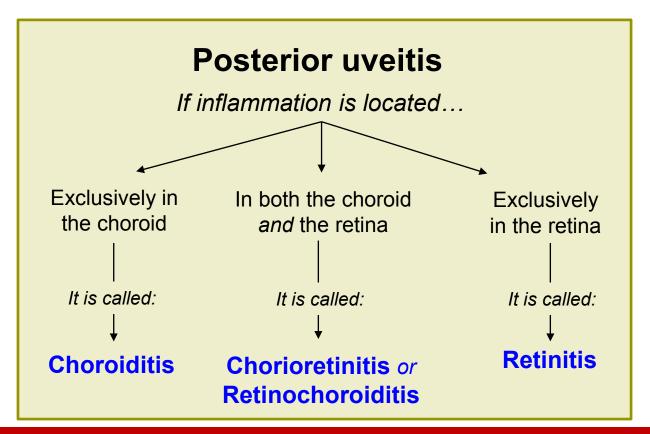




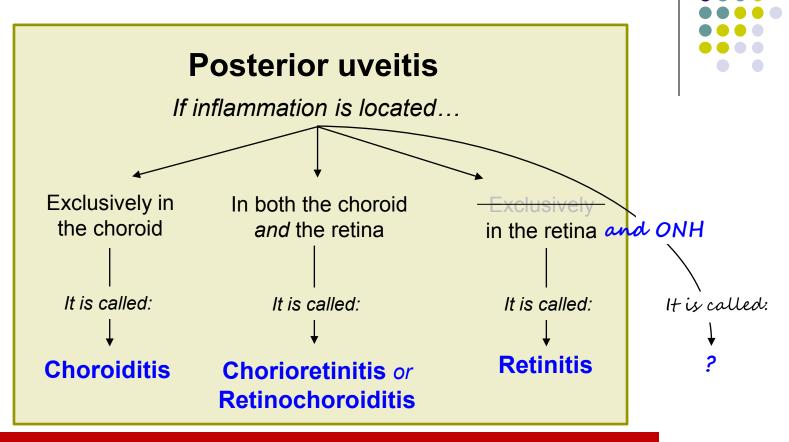




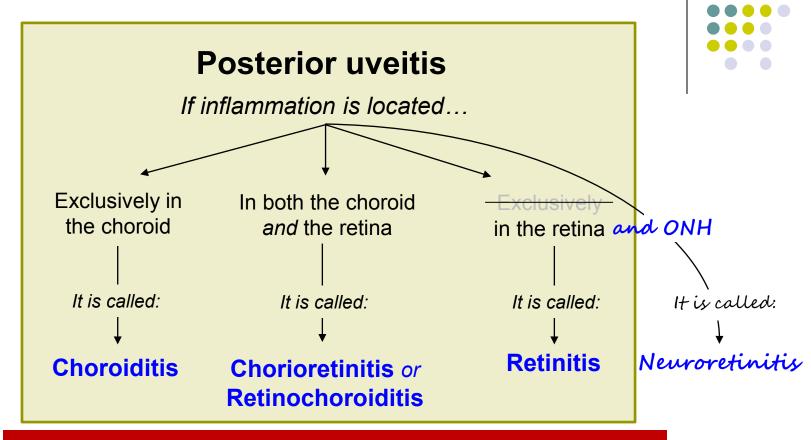




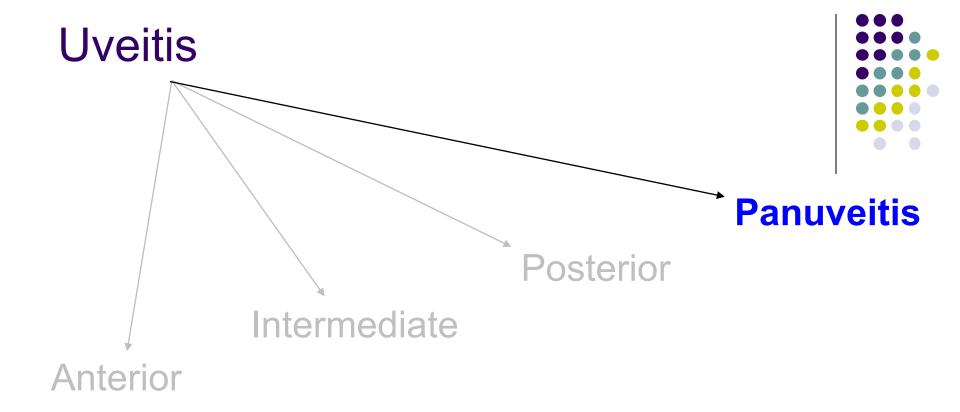




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In panuveitis, all three locations are equally involved



Many experts endorse a profiling and meshing approach to diagnosing uveitis.

1) The uveitis is profiled



Many experts endorse a profiling and meshing approach to diagnosing uveitis.

*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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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). The goal of the profiling process is to generate a concise declarative statement that captures the important features of the case; eg, 'Ms. Jones is a 40 y.o. female of Middle-Eastern descent who presents with a chronic bilateral granulomatous panuveitis associated with headache and tinnitus.'

- The uveitis is profiled
   The profiled case is meshed



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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:

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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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'Behçet is prevalent among people of 'Silk Road' ancestry. It is notorious for bilateral panuveitis and CNS symptoms. It tends not to be granulomatous, however.'

- 1) The uveitis is profiled
- 2) The profiled case is meshed
- 3) A differential diagnosis list is generated
- 4) Studies are obtained to identify the etiology



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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...

- 1) The uveitis is profiled
- 2) The profiled case is meshed
- 3) A differential diagnosis list is generated
- 4) Studies are obtained to identify the etiology
- 5) Treatment appropriate for the etiology is initiated



Many experts endorse a *profiling and meshing* approach to diagnosing uveitis.

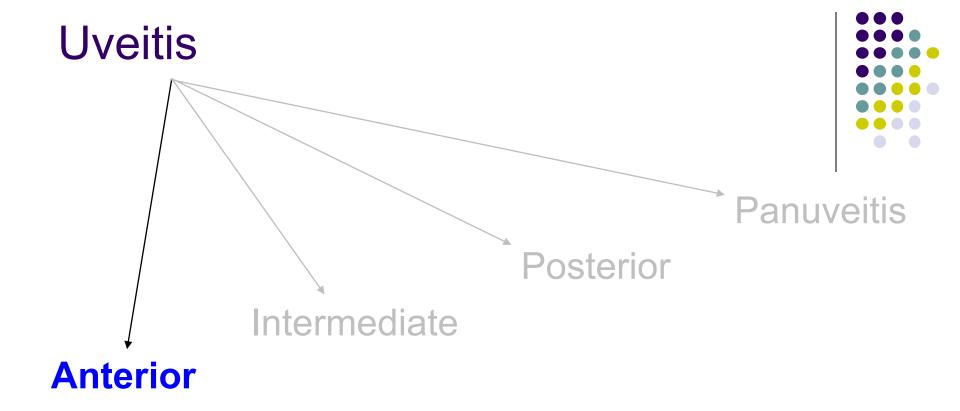
*Profiling* refers to identifying germane aspects of the pt's personal history (age, ethnicity, occupation, etc); nonocular signs and symptoms associated with the uveitis (eg, skin findings; CNS involvement); and key features of the inflammation itself (ie, location, duration, etc). The goal of the profiling process is to generate a concise declarative statement that captures the important features of the case; eg, 'Ms. Jones is a 40 y.o. female of Middle-Eastern descent who presents with a chronic bilateral granulomatous panuveitis associated with headache and tinnitus.'

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'

'Lyme uveitis is granulomatous, and CNS manifestations are very common.'
'Behçet is prevalent among people of 'Silk Road' ancestry. It is notorious for bilateral panuveitis and CNS symptoms. It tends not to be granulomatous, however.'

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



Anterior uveitis is by far the most common form encountered clinically. The classic symptoms are and and along with some degree of two words



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**.



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 two words (which presents often in a so-called two diff words pattern).



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).



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What is meant by the term ciliary flush?



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What is meant by the term ciliary flush? It refers to dilated deep conjunctival and episcleral vessels adjacent and circumferential to the corneal limbus





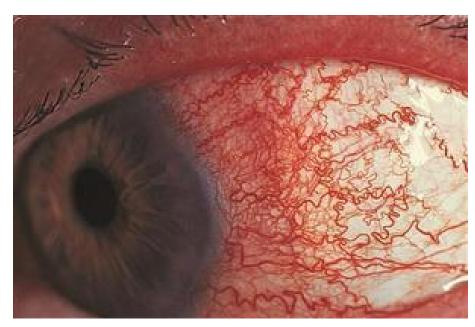
Injection in conjunctivitis

In surface disorders (eg, conjunctivitis), redness is either distributed uniformly across the eye, or it tapers off near the limbus.





Injection in conjunctivitis



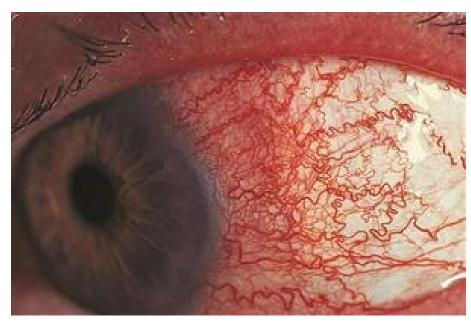
Injection in anterior uveitis, aka ciliary flush

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.





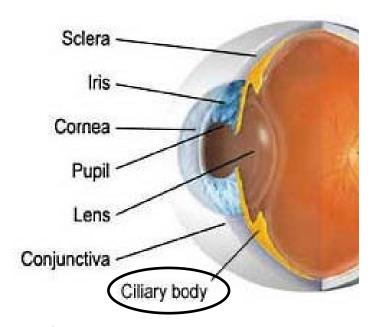
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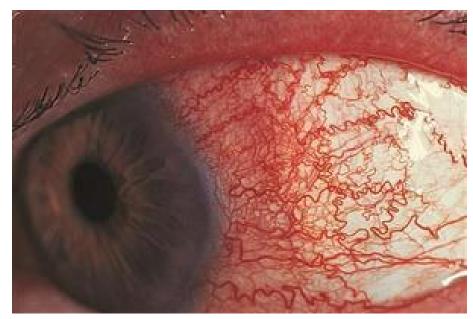
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Ciliary body just deep to the limbal region



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).

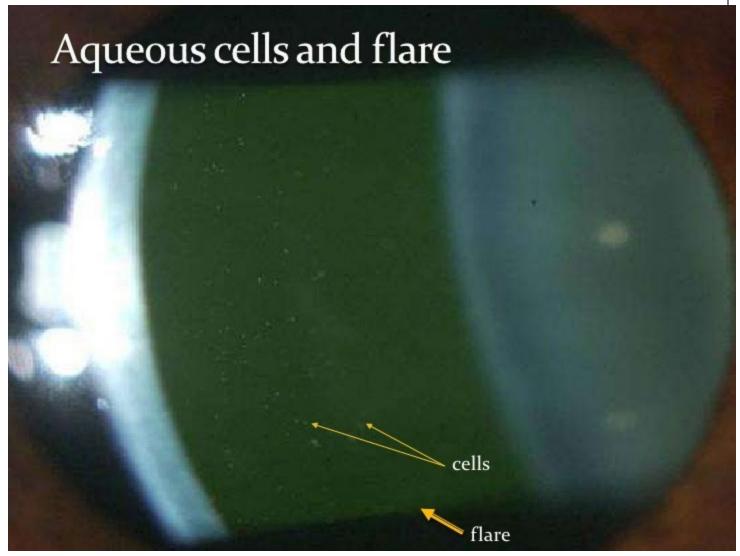


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At the slit lamp, the classic signs of anterior uveitis are WBCs and inflammatory proteins in the AC ( word and word ).



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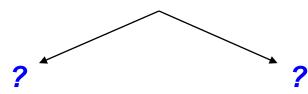






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#### Anterior Uveitis



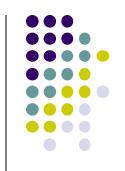


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The *Uveitis* book employs an organizational tree on which it hangs the common causes of anterior uveitis. The first branch point in this tree is whether the inflammation is word or non-word.

# Uveitis Anterior Uveitis Granulomatous



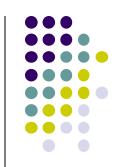
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# Uveitis Anterior Uveitis



Granulomatous Nongranulomatous

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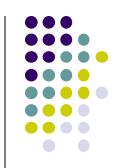
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Granulomatous KP

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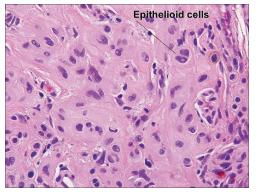


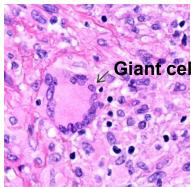
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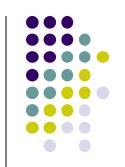
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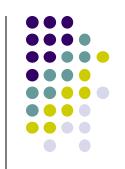
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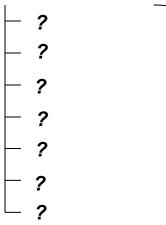
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This is key: In clinical use the term *granulomatous* refers to the slit-lamp appearance of the KP, **not** to the underlying histology of the condition.

#### **Anterior Uveitis**

#### 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.)

#### **Anterior Uveitis**

#### Granulomatous

TB

Sarcoid

**Syphilis** 

HSV

VKH

Toxoplasmosis

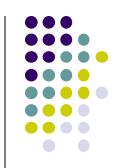
Lyme

#### Nongranulomatous

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# Uveitis Anterior Uveitis Granulomatous

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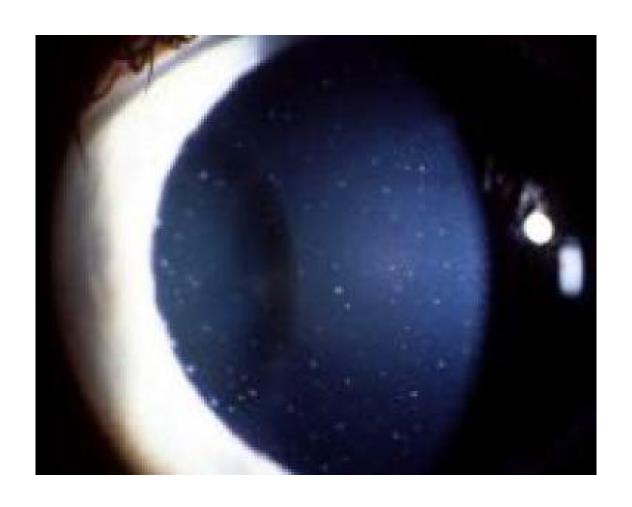
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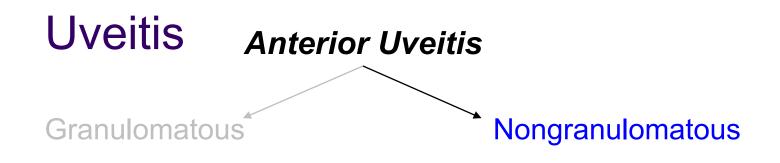
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In contrast, *nongranulomatous KP* are smaller, lighter in color, and do not look greasy. (Note: If no KP are present, the inflammation is considered nongranulomatous.)



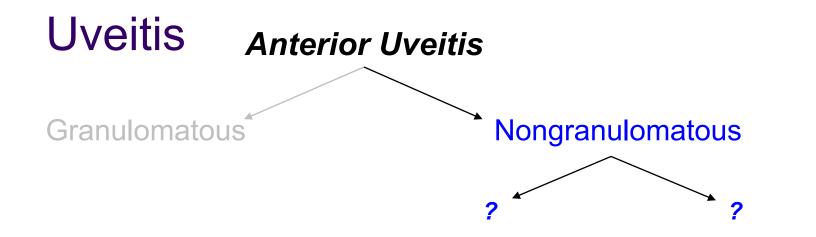


Nongranulomatous KP

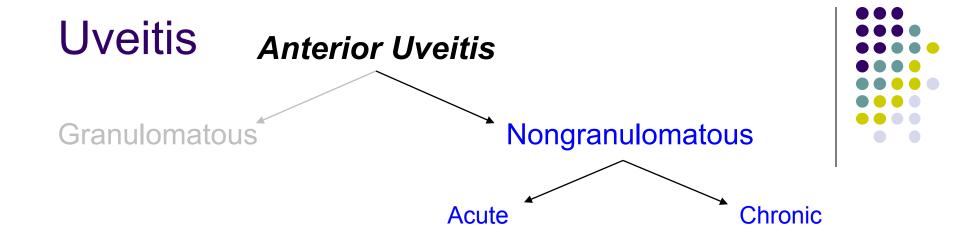


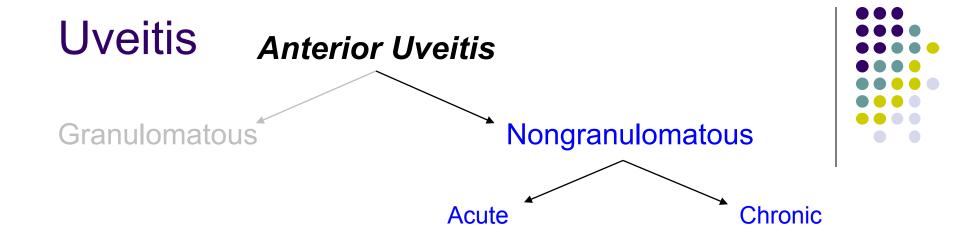


The rest of the anterior-uveitis classification tree concerns **nongranulomatous** dz.

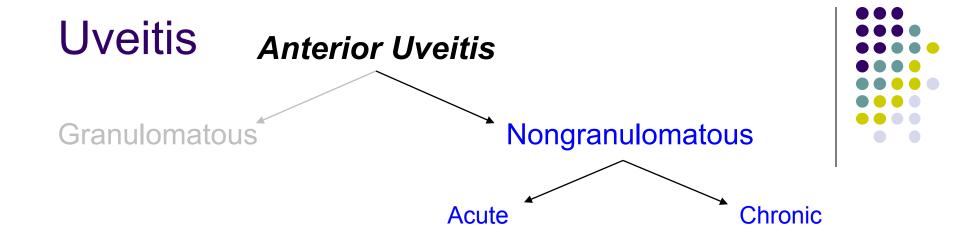






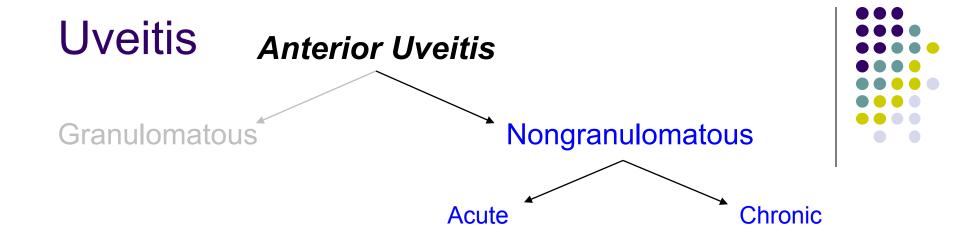


Acute uveitis comes on suddenly and resolves fairly quickly.

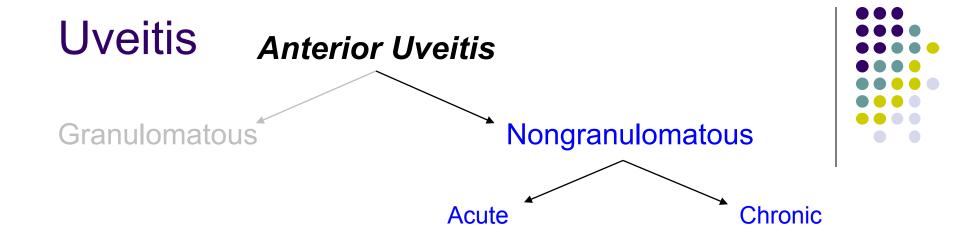


Acute uveitis comes on suddenly and resolves fairly quickly.

Chronic uveitis also resolves, but once treatment is withdrawn, it relapses within amount of time.

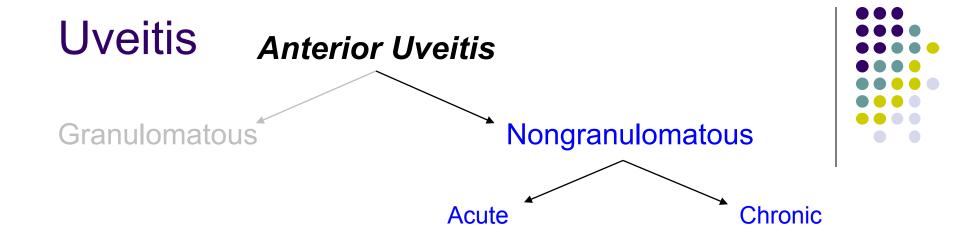


Acute uveitis comes on suddenly and resolves fairly quickly. Chronic uveitis also resolves, but once treatment is withdrawn, it relapses within three months.



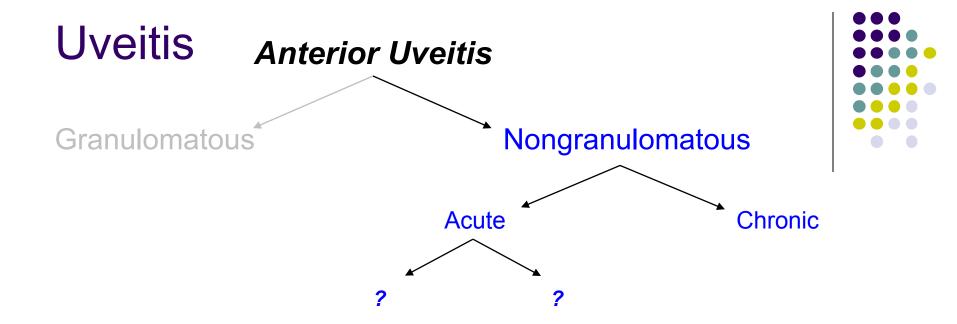
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(FYI: If a uveitis eventually relapses but is quiescent off-treatment for **longer** than three months, it is termed a uveitis.)

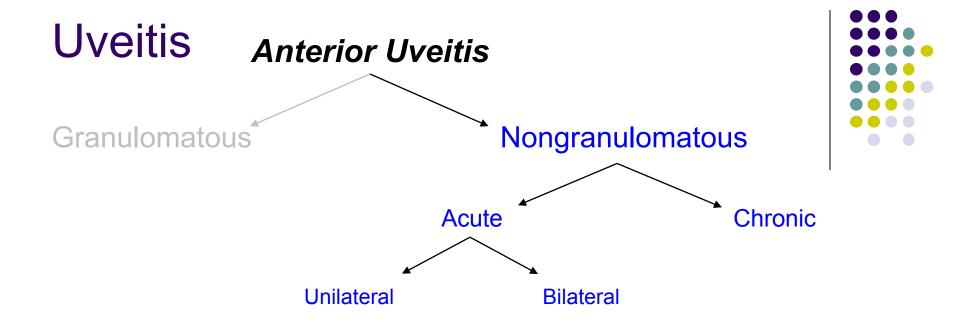


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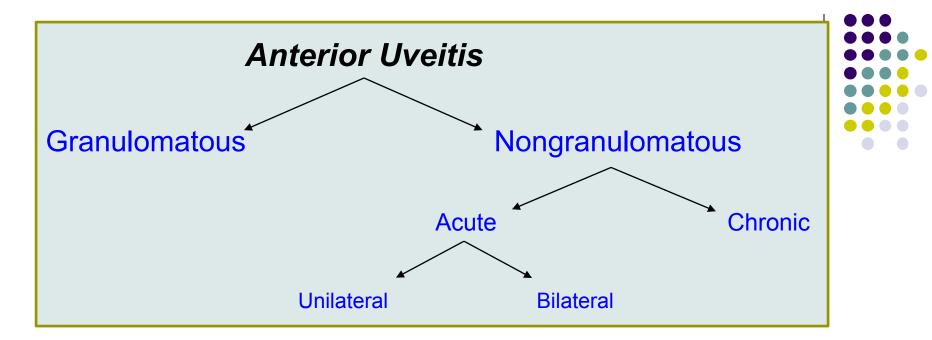
(FYI: If a uveitis eventually relapses but is quiescent off-treatment for **longer** than three months, it is termed a *recurrent* uveitis.)



Finally, the acute uveitides are divided into those that present vs those that tend to present



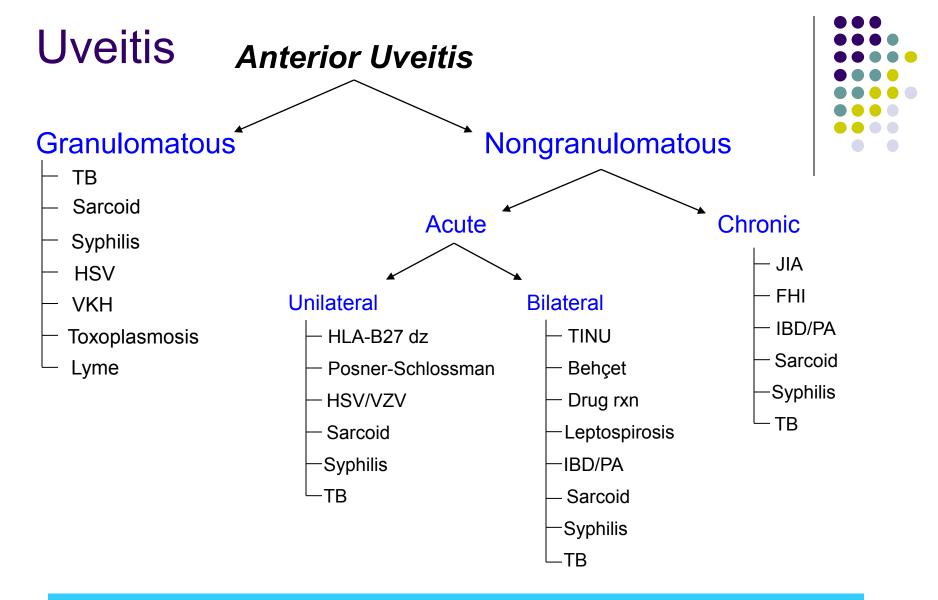
Finally, the acute uveitides are divided into those that present **unilaterally** vs those that tend to present **bilaterally**.



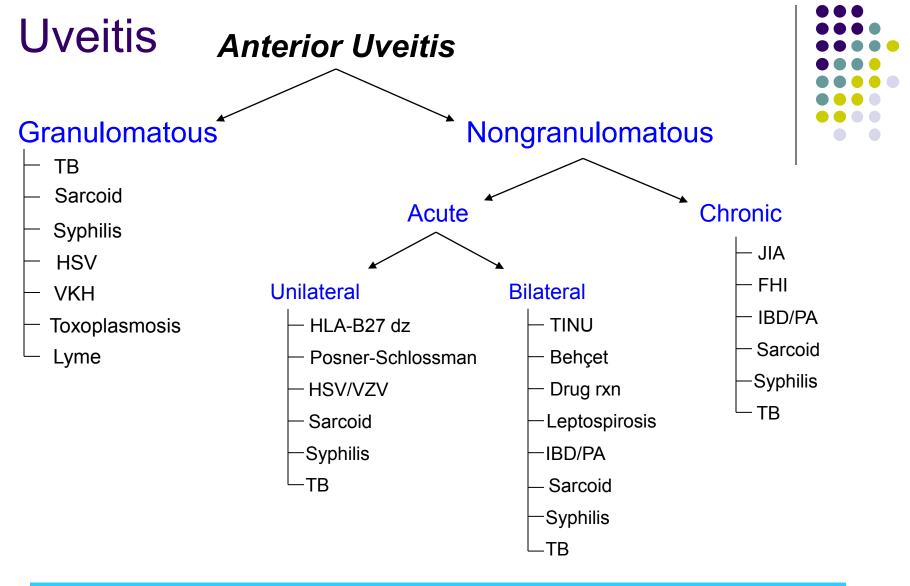
Take a good look at this—it represents how you should think about anterior uveitides encountered in the clinic or on the OKAP. It wouldn't be a bad idea to commit this to memory at this juncture.

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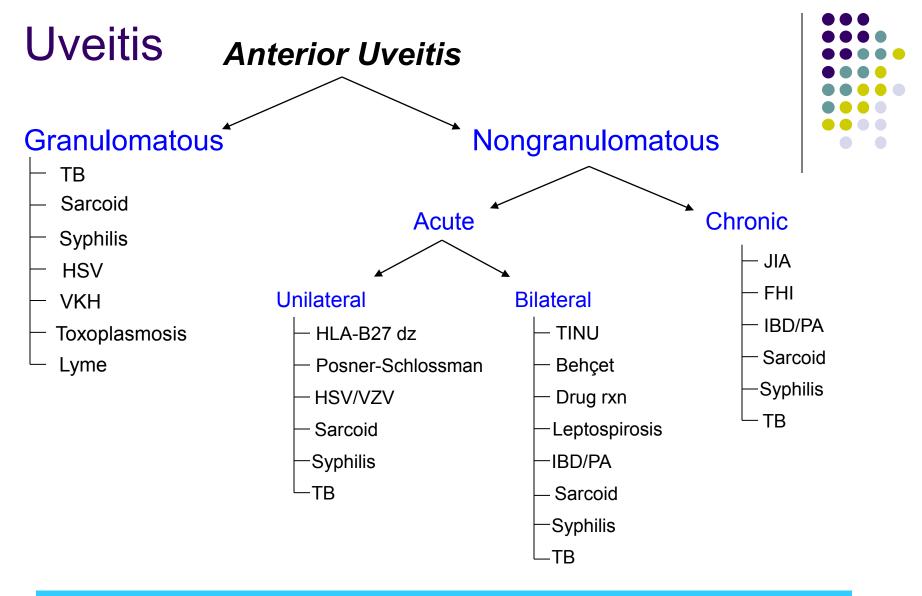
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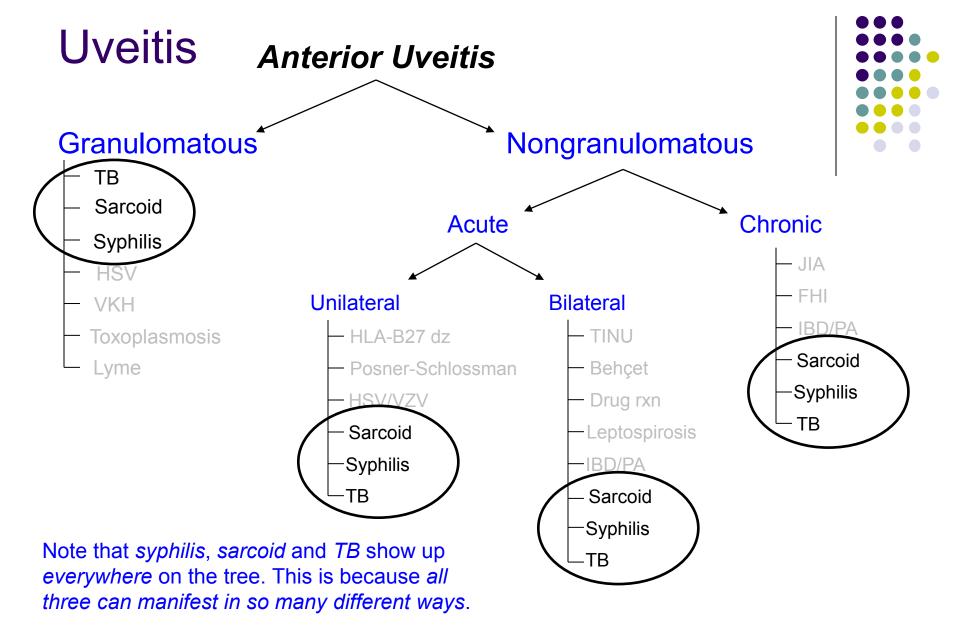
Just as an FYI, these are the anterior uveitides that are covered in detail in the *Uveitis* book. **Don't** *try to memorize all this now!* (They will stick better if you learn them in their naturally-occurring groupings.)

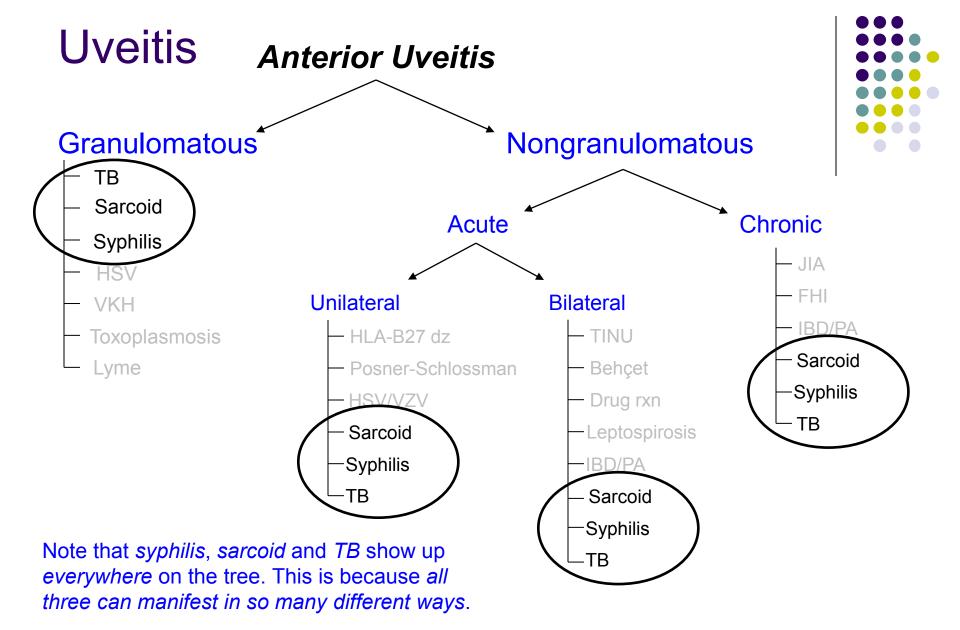


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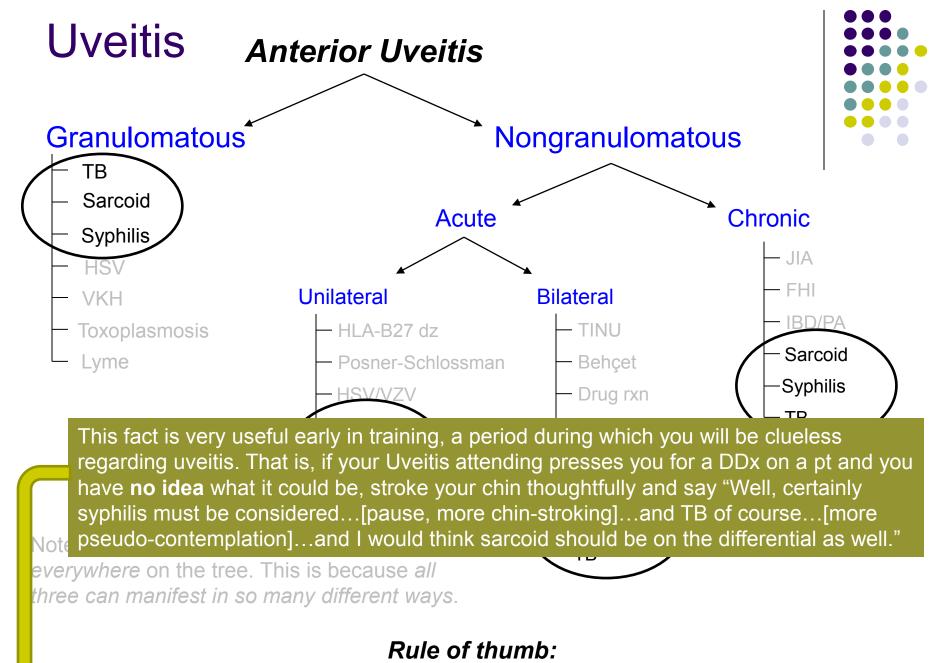
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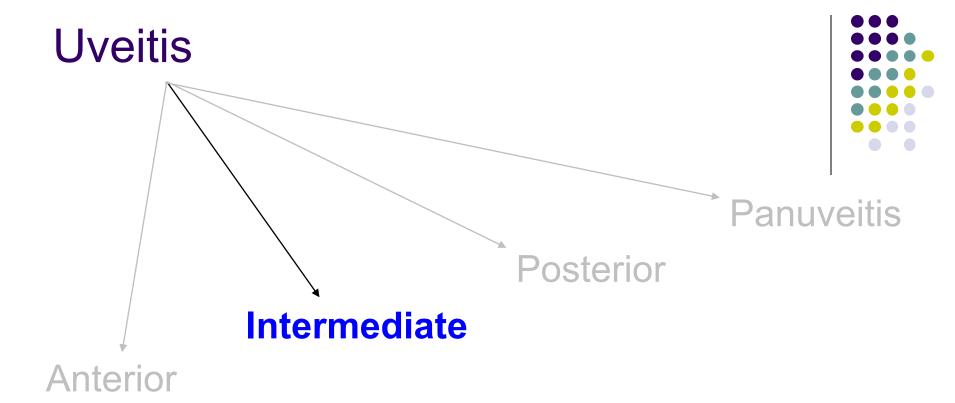


#### Rule of thumb:

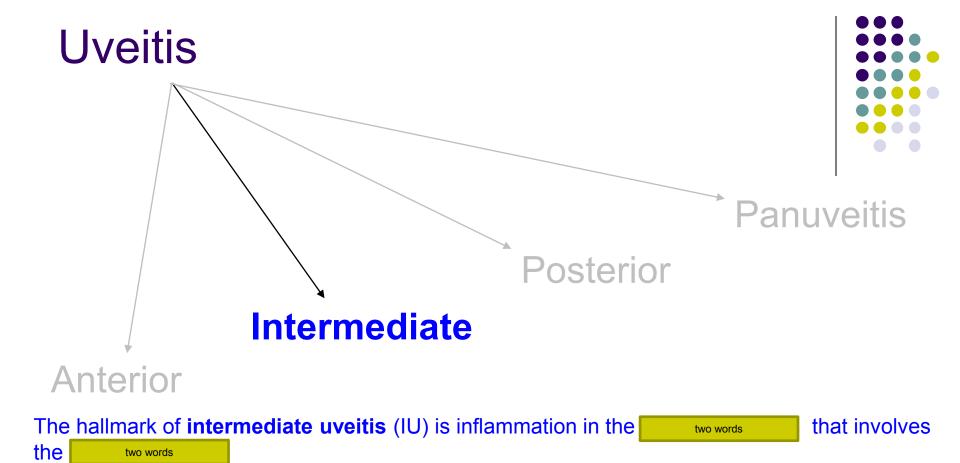
Syphilis, sarcoid and TB are on the DDx for every pt with any form of uveitis!



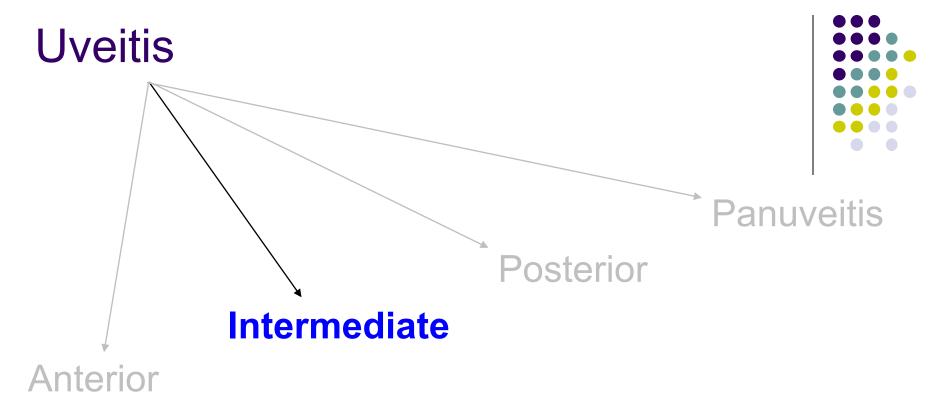
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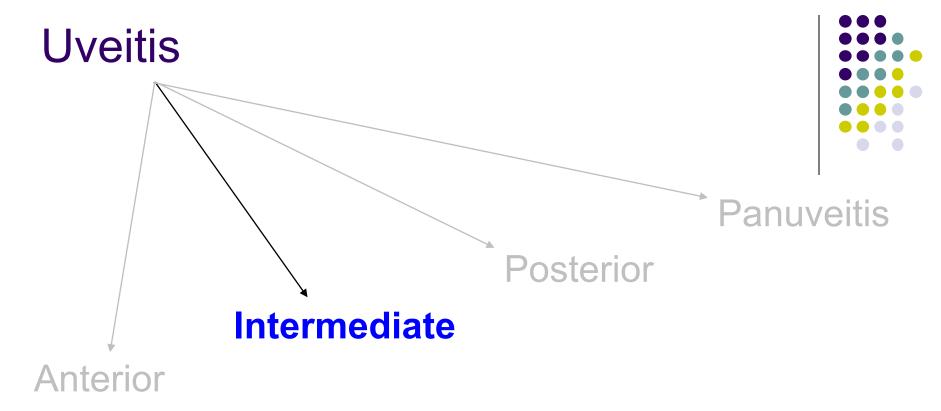
Next let's look at intermediate uveitis



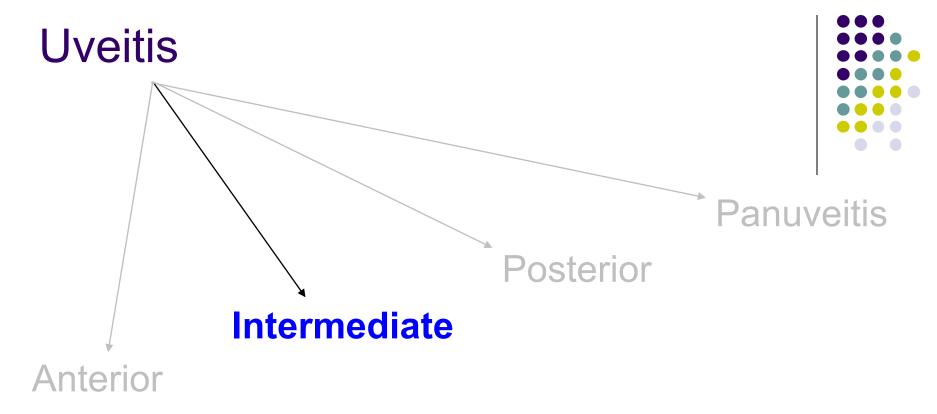
two words



The hallmark of **intermediate uveitis** (IU) is inflammation in the anterior vitreous that involves the vitreous base.

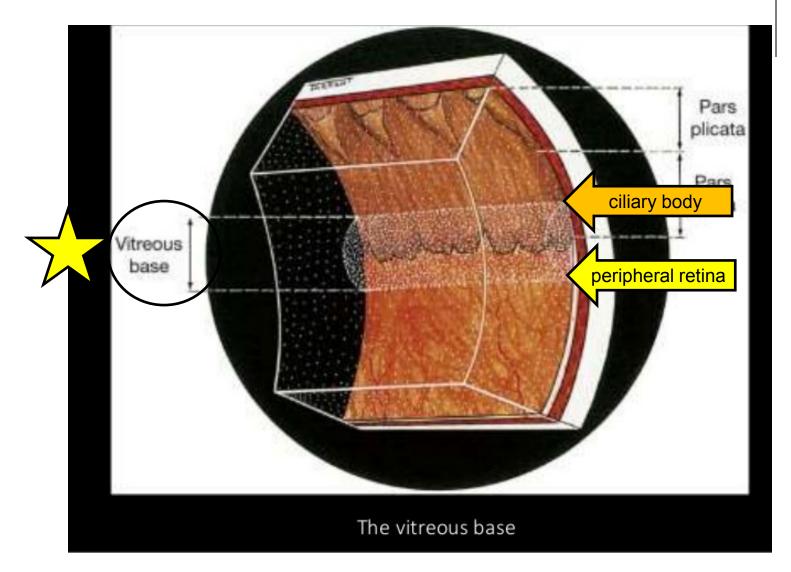


The hallmark of **intermediate uveitis** (IU) is inflammation in the anterior vitreous that involves the vitreous base. The vitreous base is the primary attachment point of the vitreous; it forms a ~5 mm-wide band that straddles the two words (the location where the anterior-most retina meets the posterior-most portion of the ciliary body).

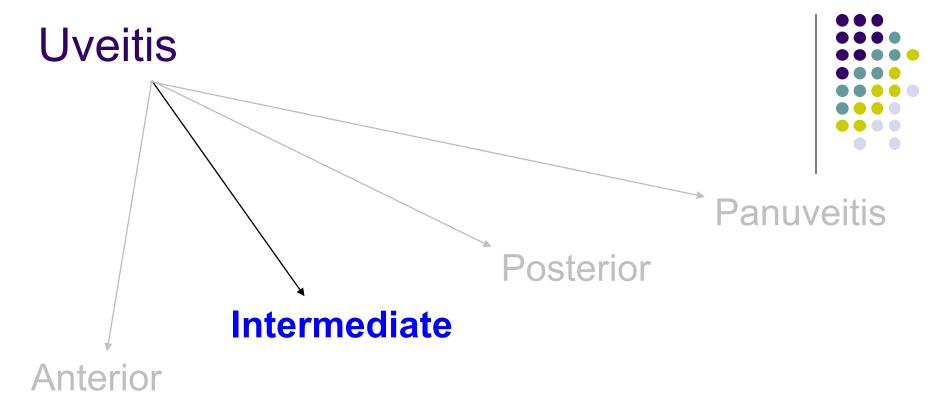


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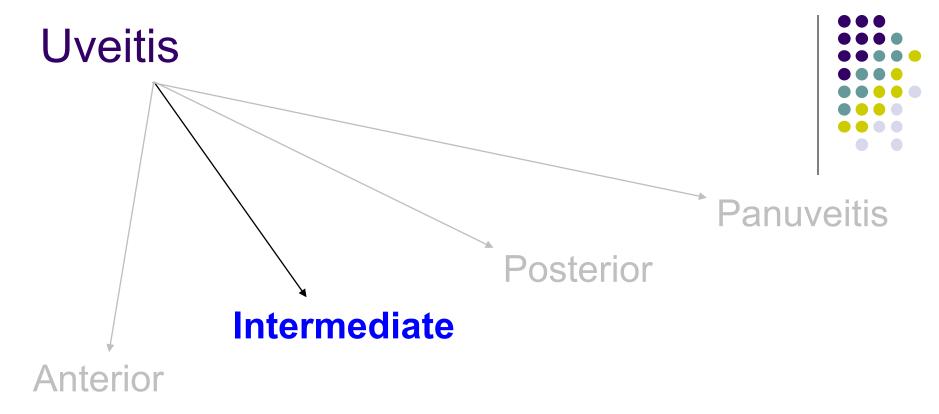
# **Uveitis**



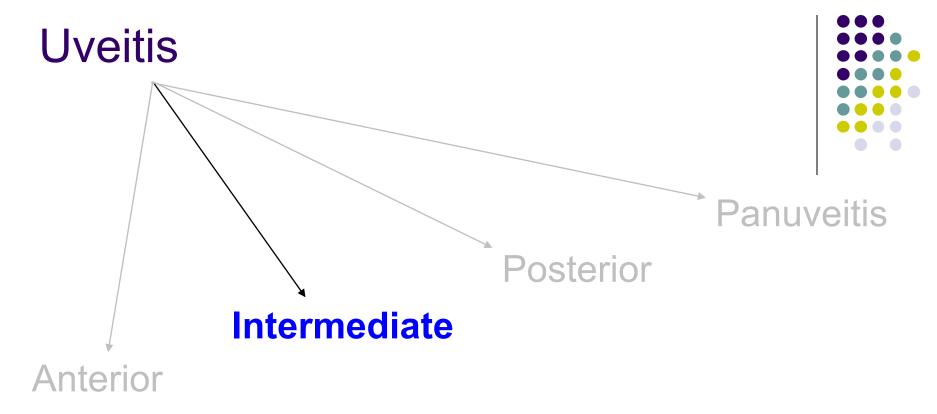




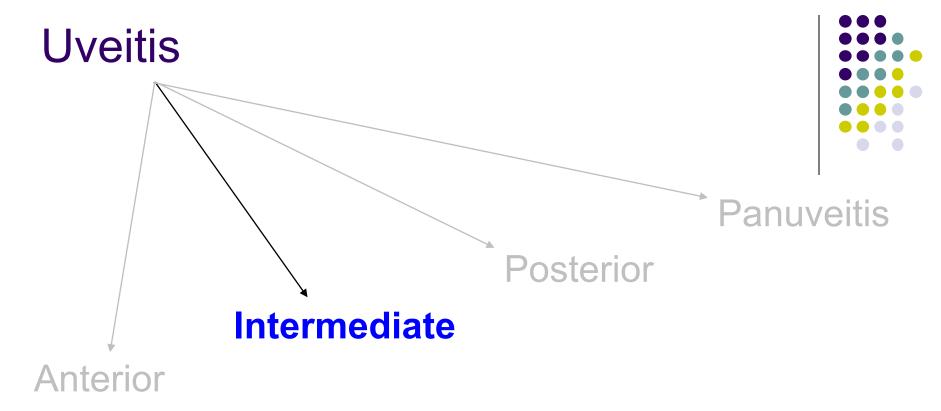
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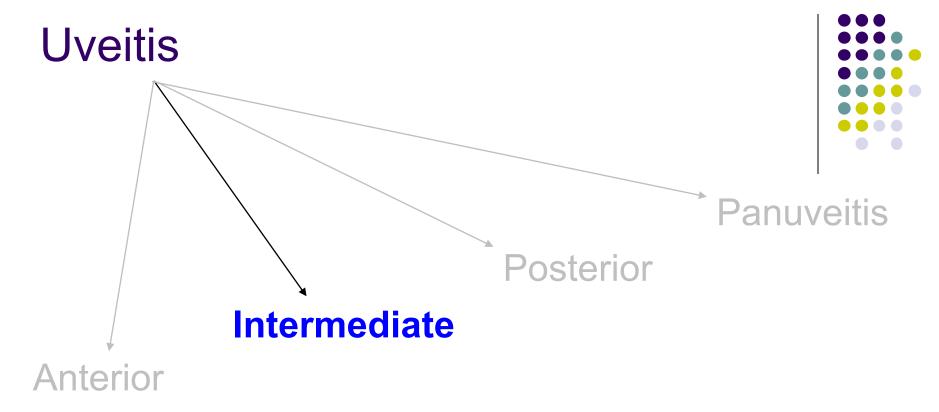


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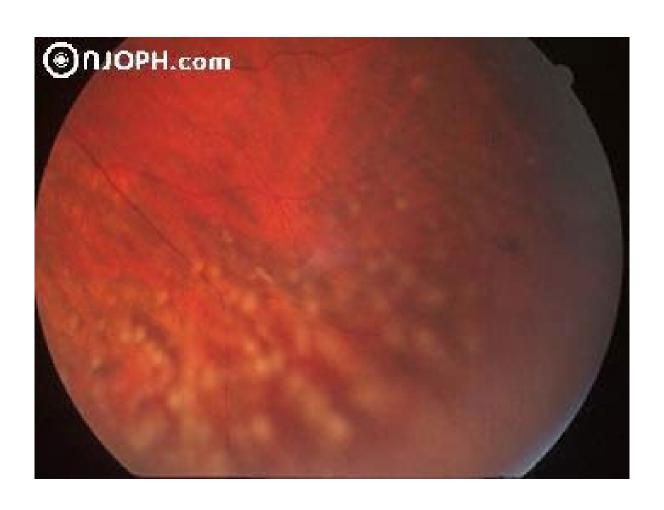
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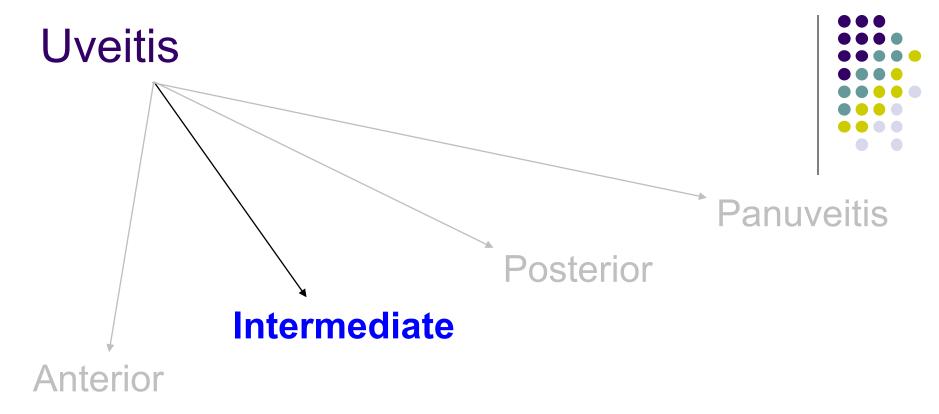


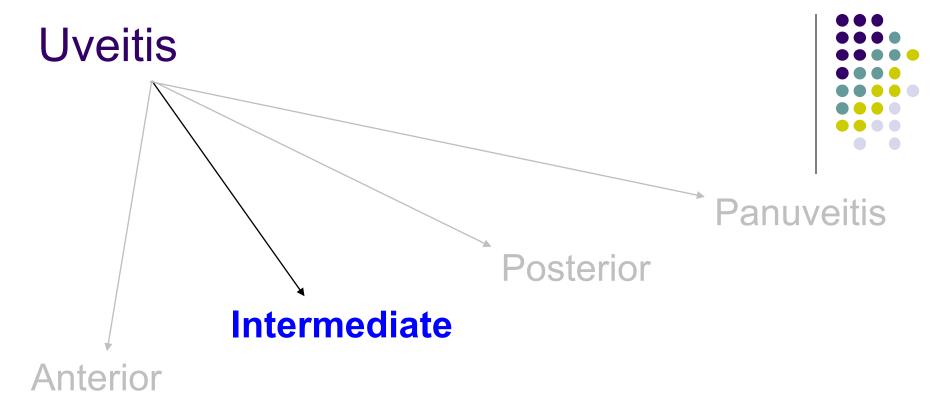
## **Uveitis**





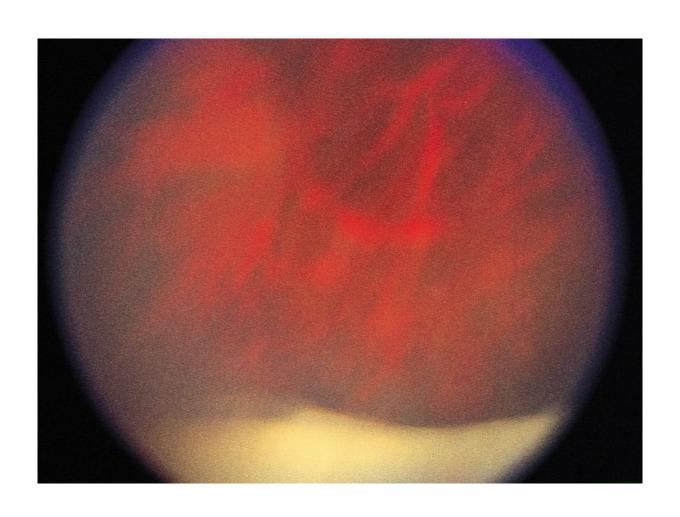
Snowballs in intermediate uveitis



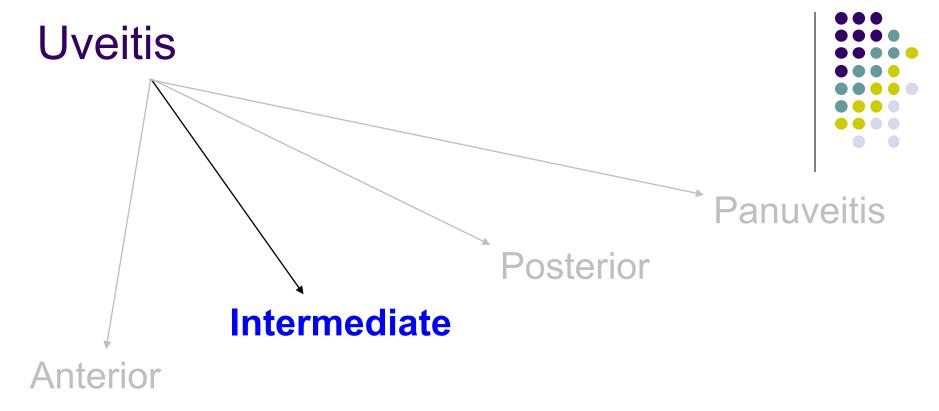


# **Uveitis**

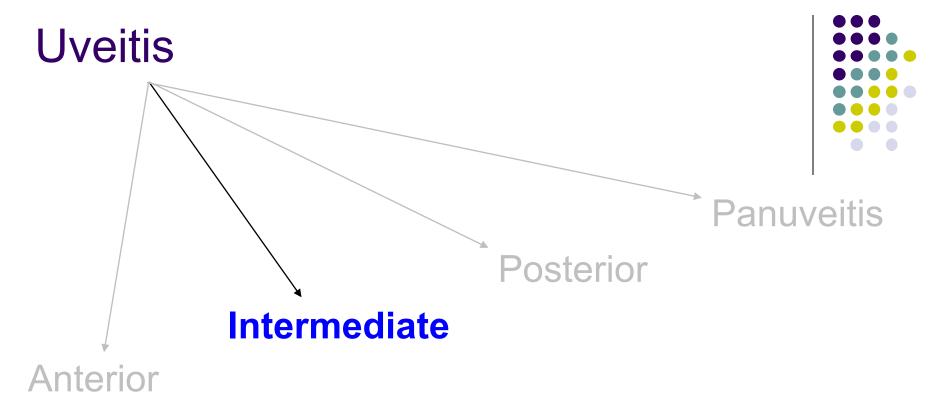




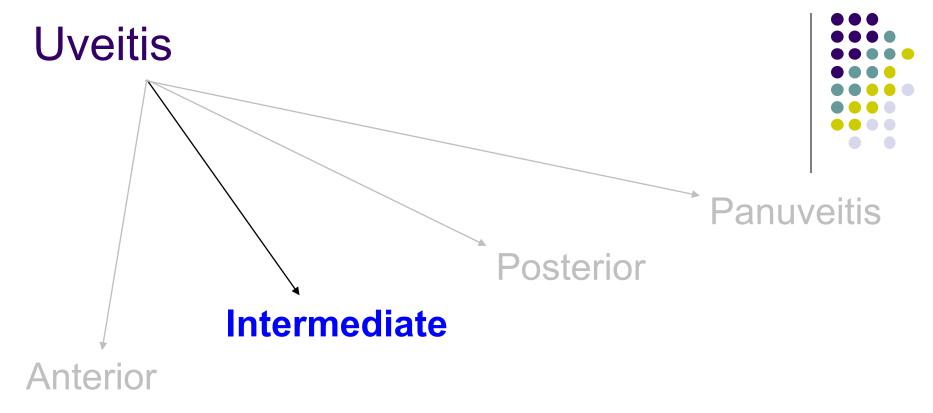
Snowbanking in intermediate uveitis



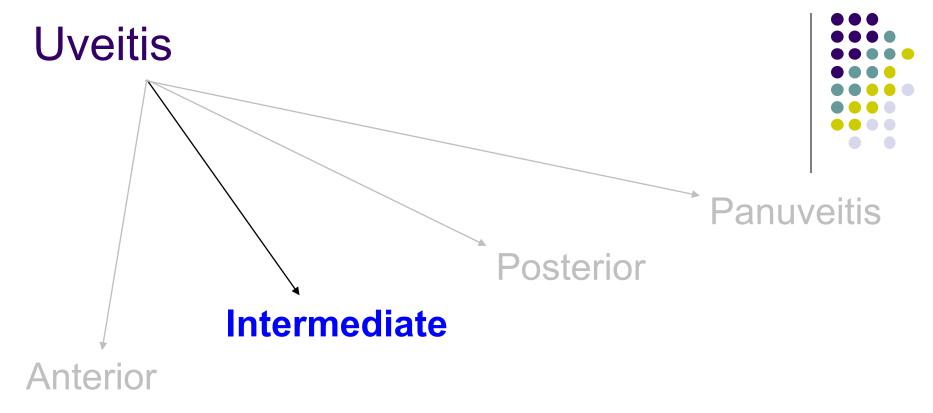
IU tends to be a dz of young vs older people— [age range]



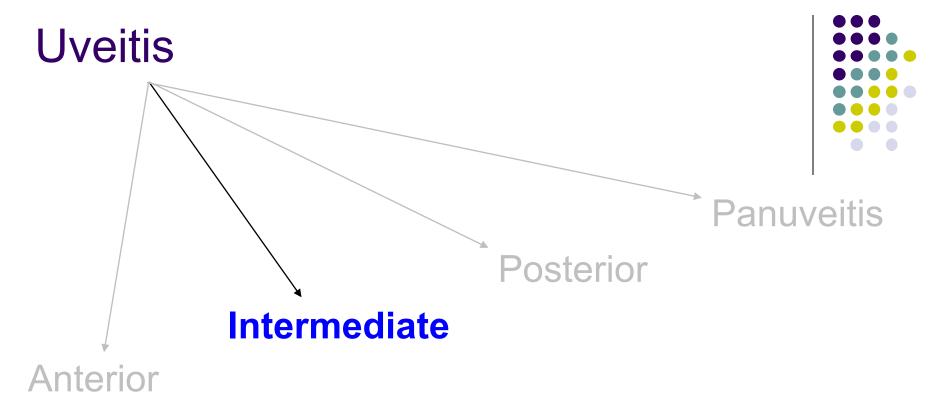
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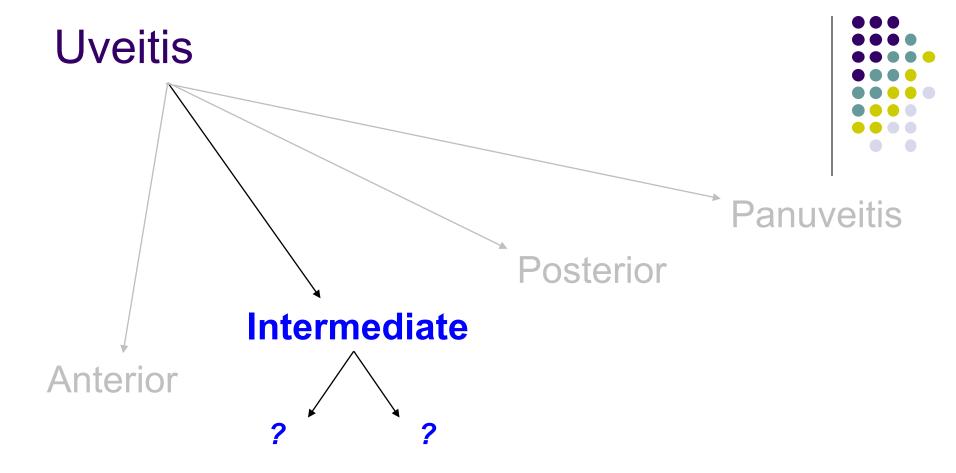
IU tends to be a dz of young people—teens through 40 or so. It is uni- v bilateral in most (80%) cases



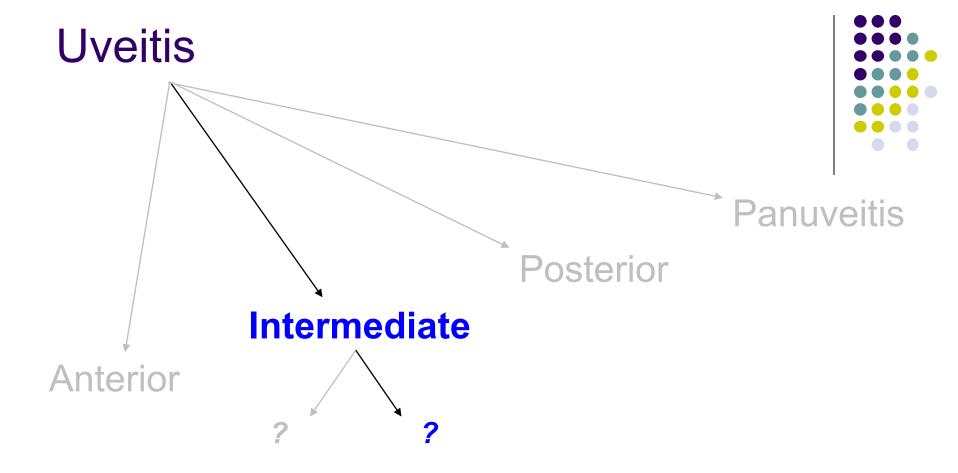
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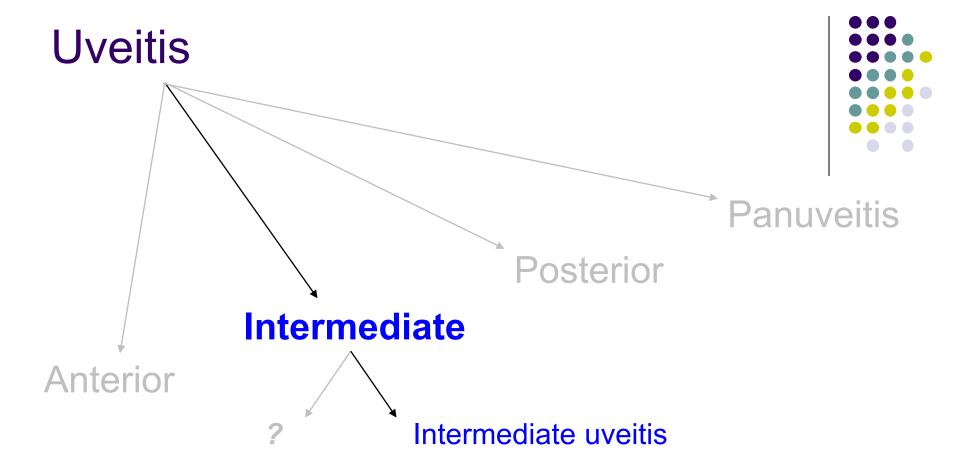
IU tends to be a dz of young people—teens through 40 or so. It is bilateral in most (80%) cases (although it can be quite asymmetric).



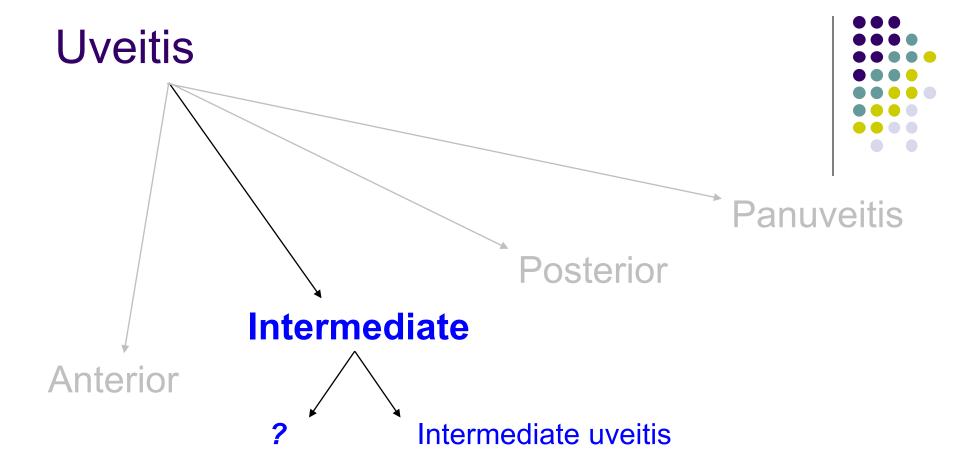
As indicated previously, IU is divvied up into two categories.



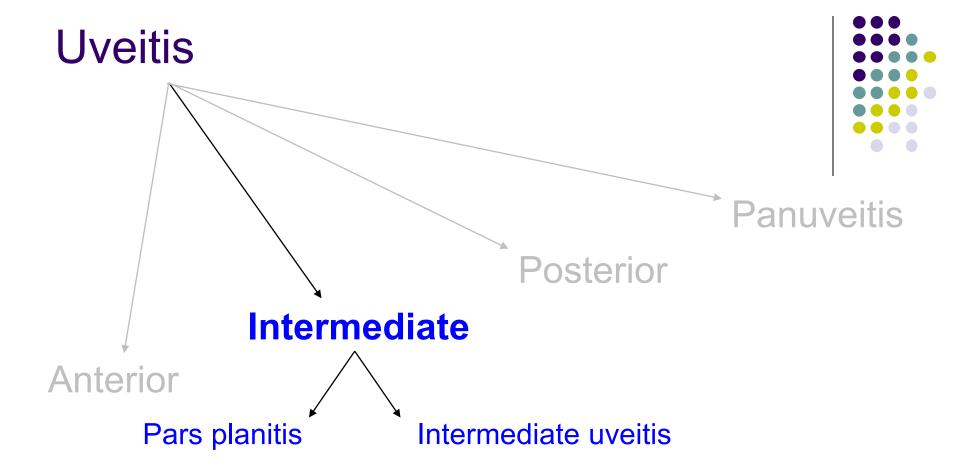
As indicated previously, IU is divvied up into two categories. If the inflammation is associated with an identifiable condition, the uveitis is called abb.



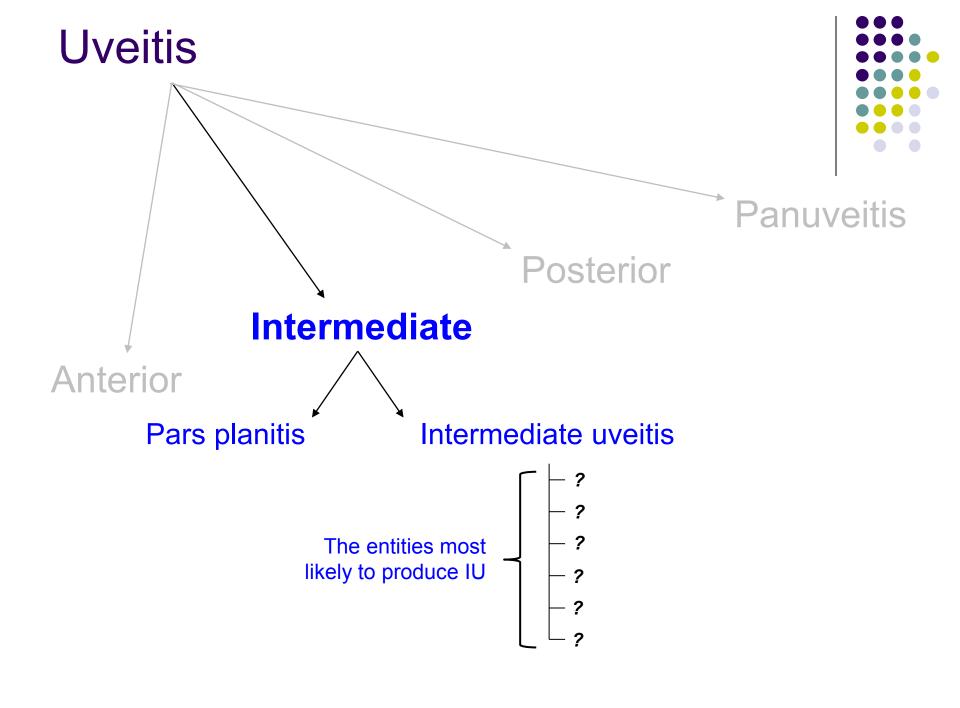
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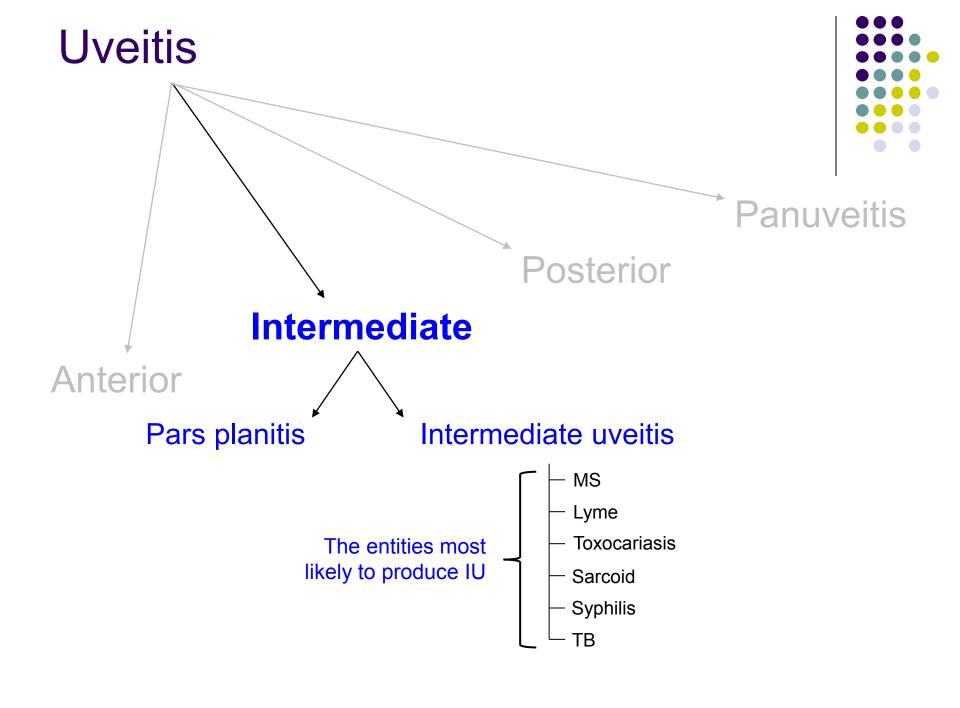


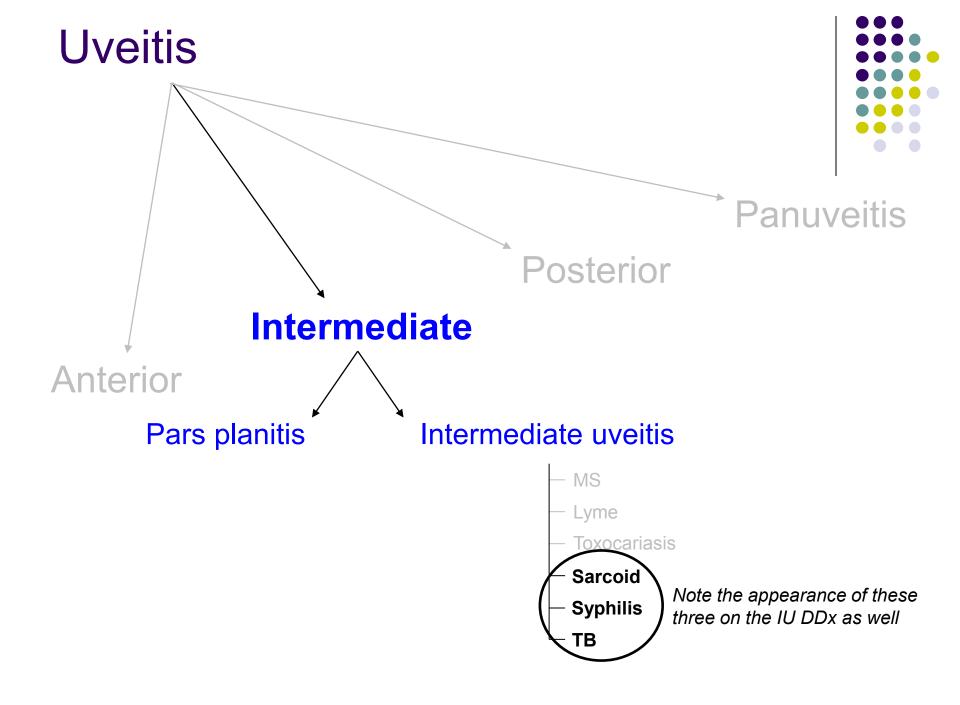
As indicated previously, IU is divvied up into two categories. If the inflammation is associated with an identifiable condition, the uveitis is called IU. If it is idiopathic, ie, if no cause can be identified, it is called

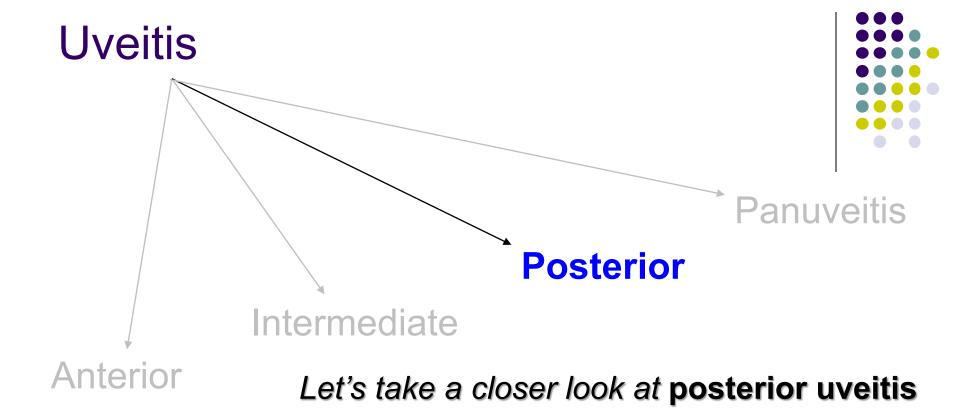


As indicated previously, IU is divvied up into two categories. If the inflammation is associated with an identifiable condition, the uveitis is called IU. If it is idiopathic, ie, if no cause can be identified, it is called pars planitis.





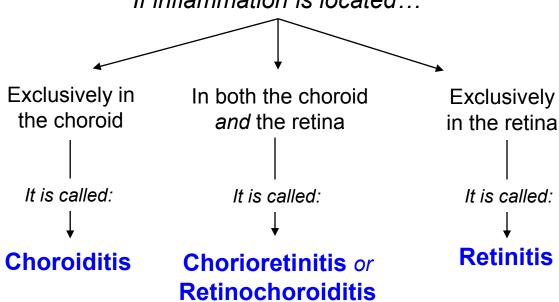




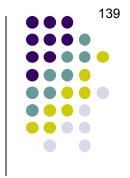
### **Uveitis**

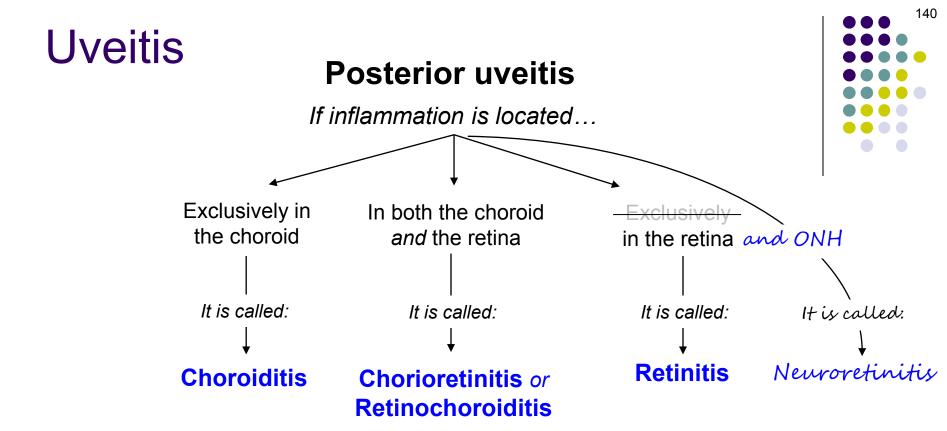
#### **Posterior uveitis**

If inflammation is located...

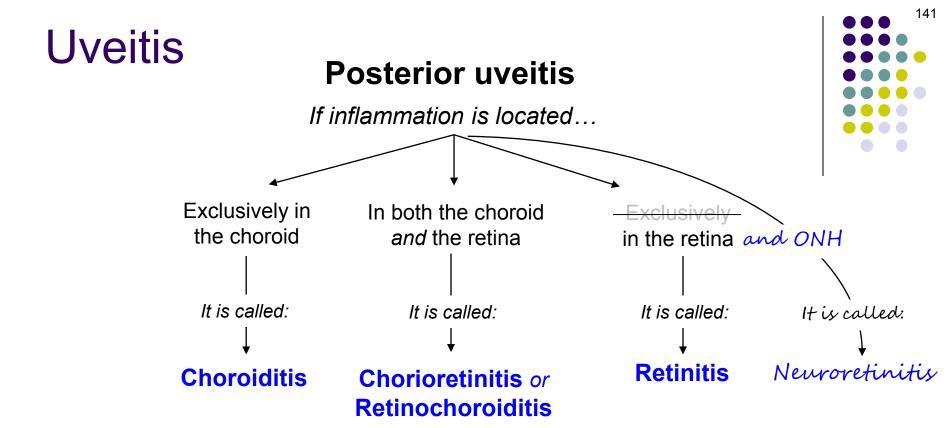


As noted previously, here are the ways **posterior uveitis** can manifest.



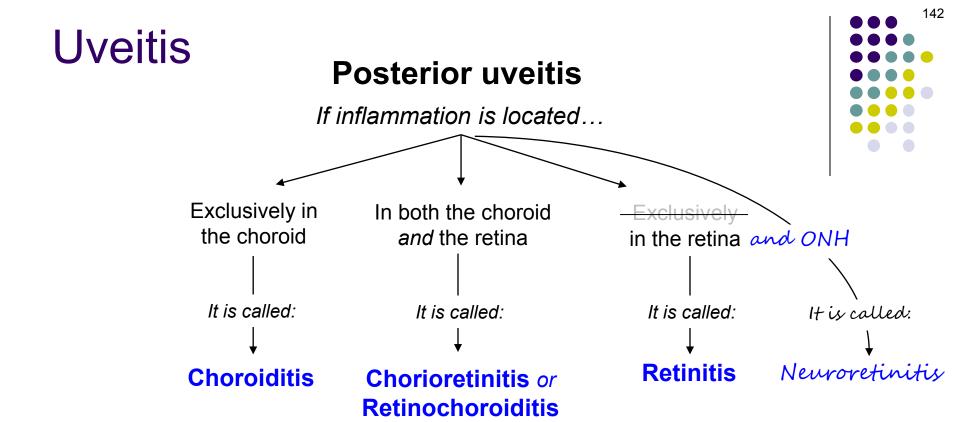


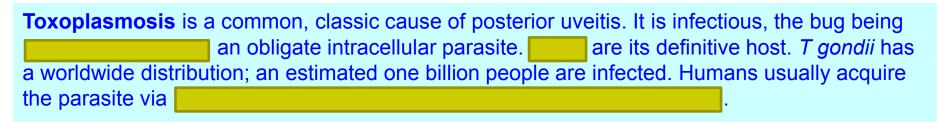
As noted previously, here are the ways **posterior uveitis** can manifest. Also noted previously was the condition *neuroretinitis*—inflammation involving both the retina and optic nerve.

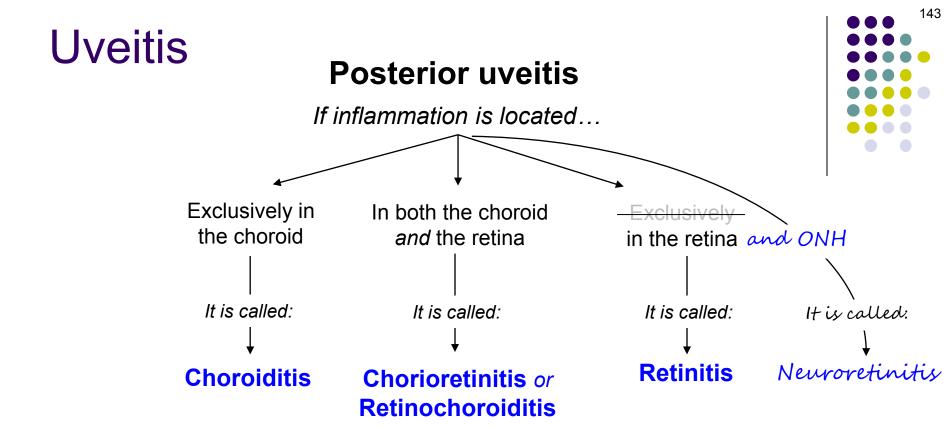


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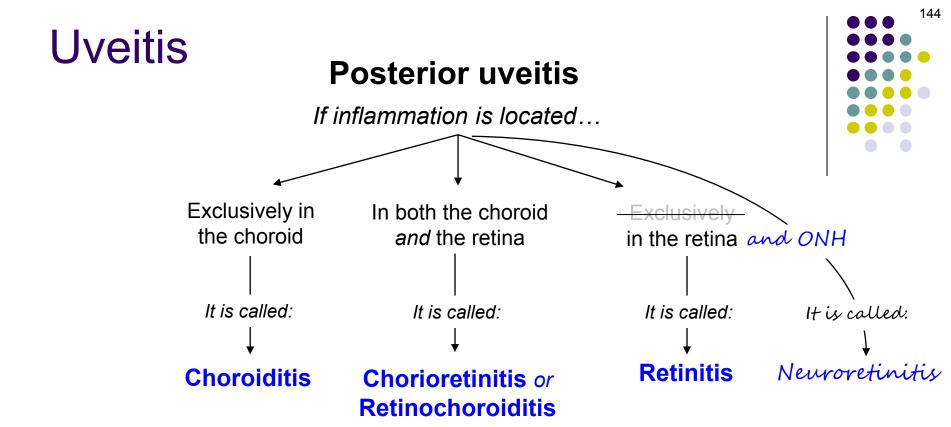
While 80% of anterior uveitis cases are noninfectious in origin, *the opposite is true for posterior uveitis*: most cases are infectious—weirdly, also about 80%.



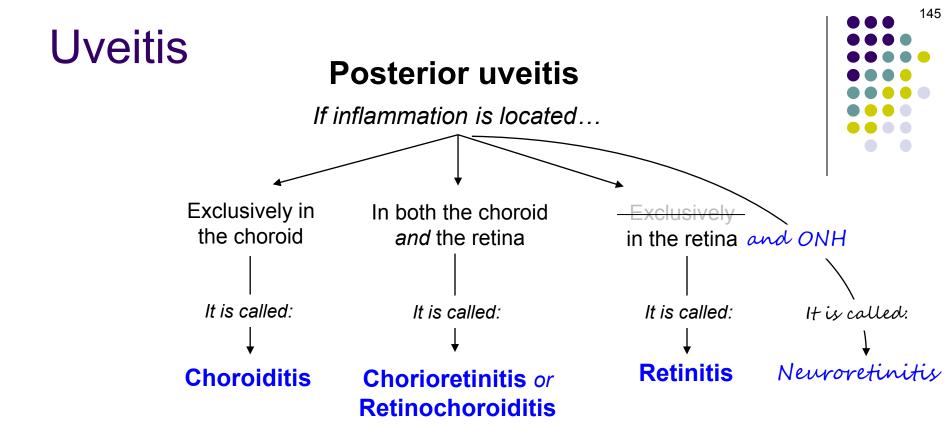




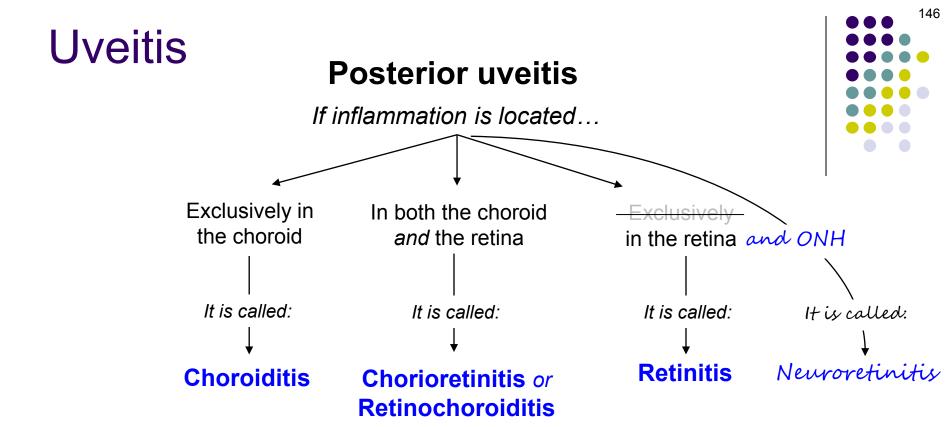
**Toxoplasmosis** is a common, classic cause of posterior uveitis. It is infectious, the bug being *Toxoplasma gondii*, an obligate intracellular parasite. Cats are its definitive host. *T gondii* has a worldwide distribution; an estimated one billion people are infected. Humans usually acquire the parasite via consumption of unwashed produce or undercooked meat.



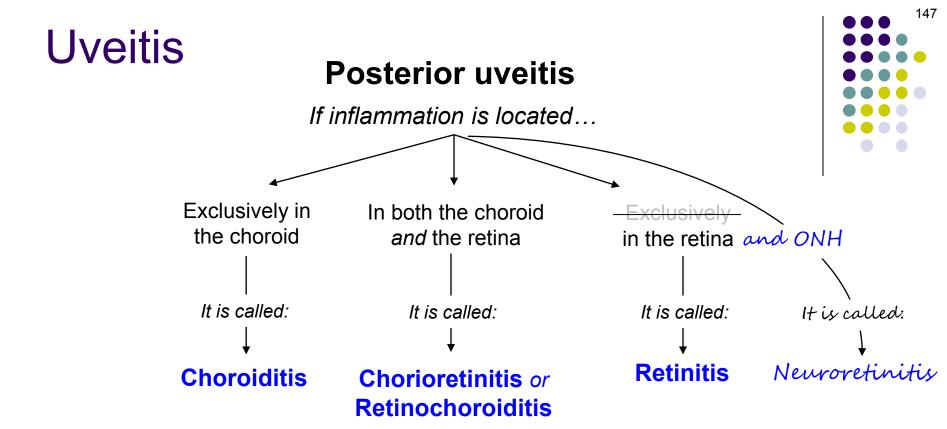
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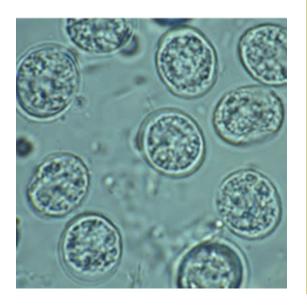
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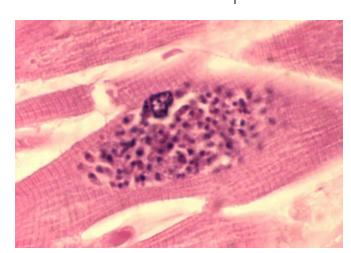


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#### **Uveitis**







Oocyst form

Tachyzoite form

Tissue cyst

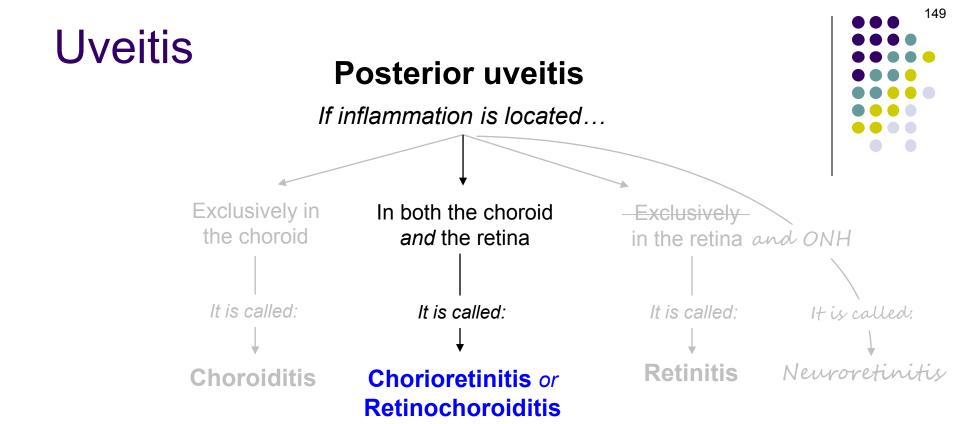
--Found in GI tract of cat (shed in feces)

--Acquired via ingestion of unwashed produce

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

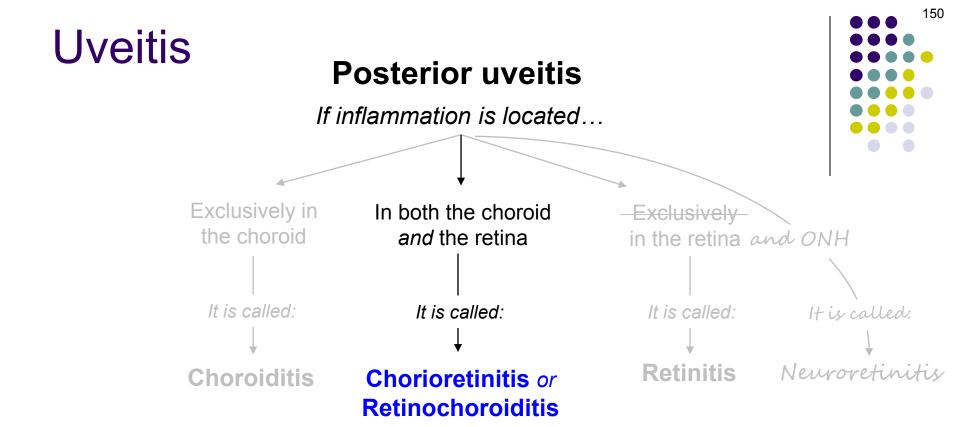
--Found in tissue of infected livestock
--Acquired via consumption of
undercooked meat

Toxoplasma gondii: Three infectious forms



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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

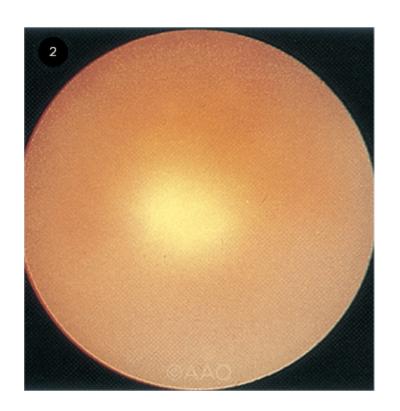


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Toxoplasmosis typically manifests as a retinochoroiditis accompanied by a dense overlying vitritis. Taken together, the appearance has been likened to a 'headlight in the fog.'

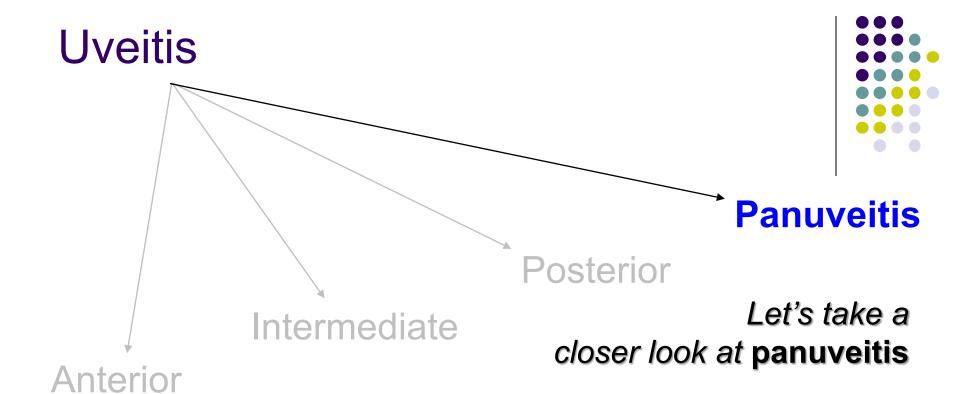
#### **Uveitis**

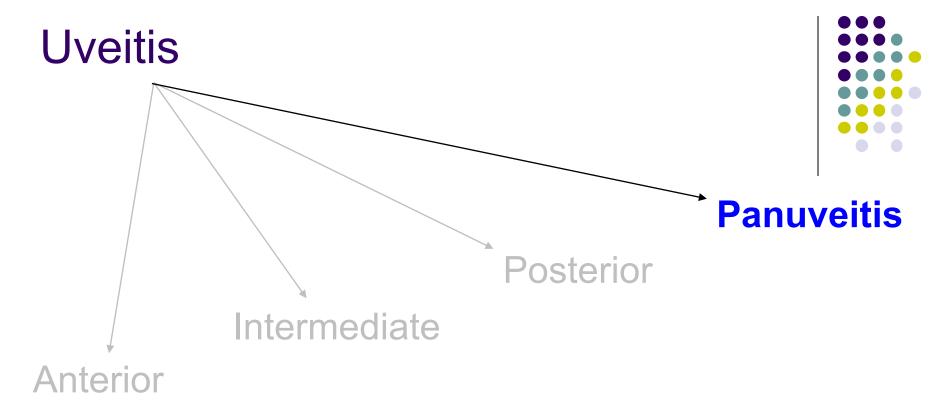




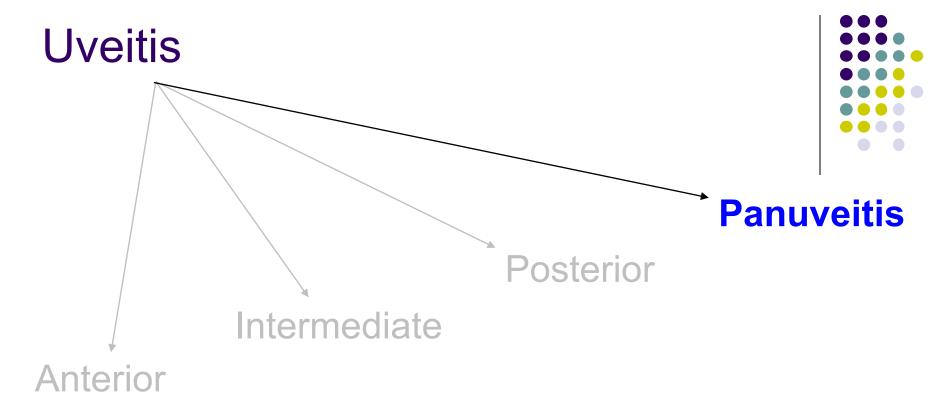


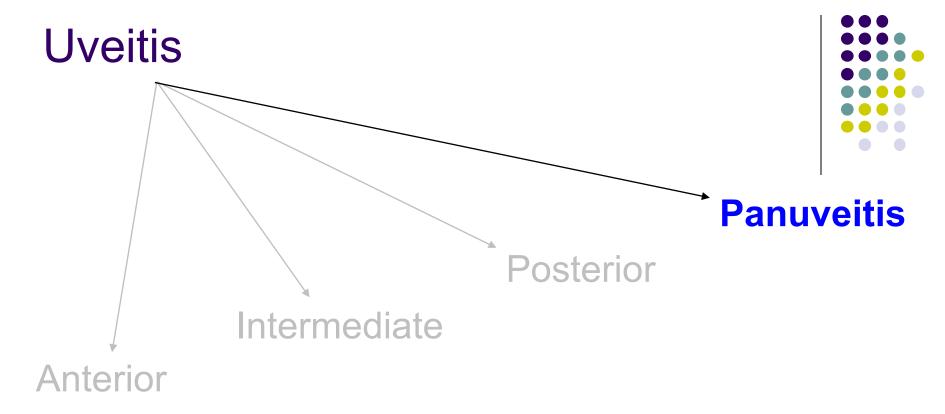
Ocular toxoplasmosis: 'Headlight in the fog'

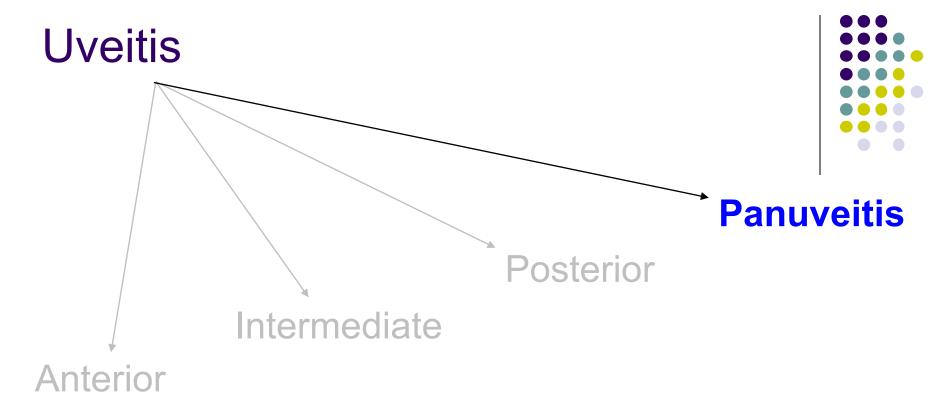


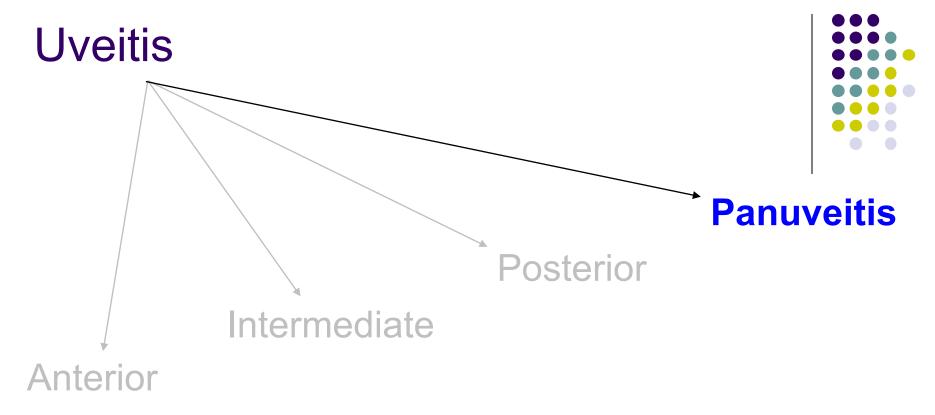


To qualify as a **panuveitis**, all compartments of the eye—the AC, vitreous, and retina/choroid—must be equally involved in the inflammatory process.

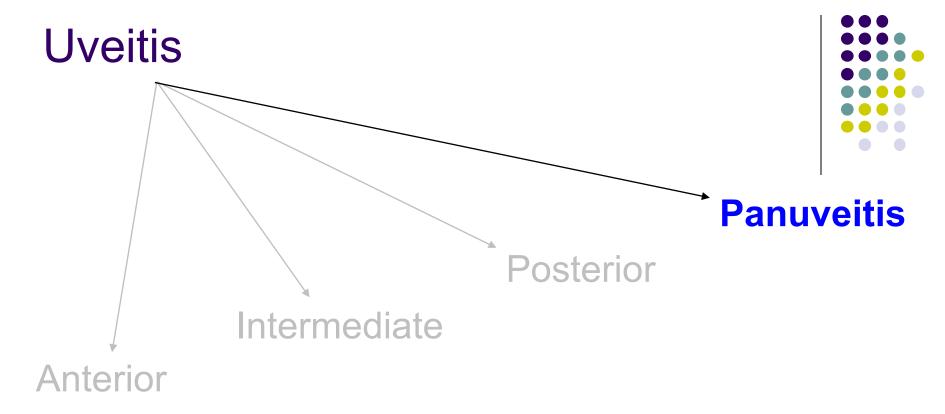


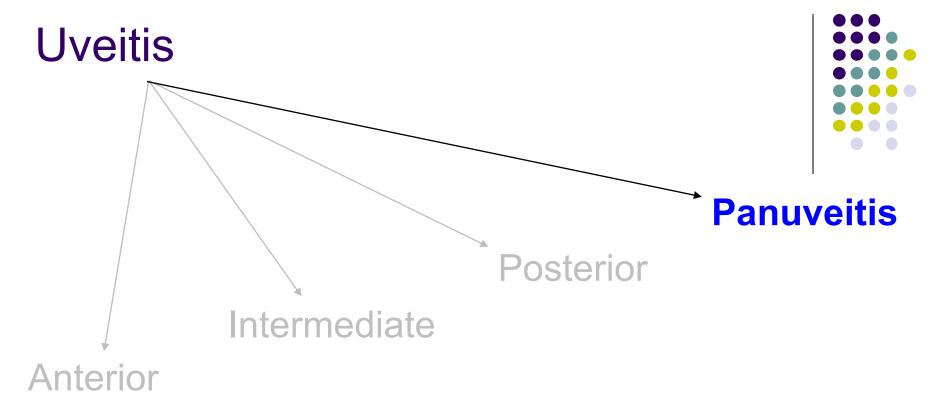


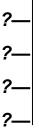


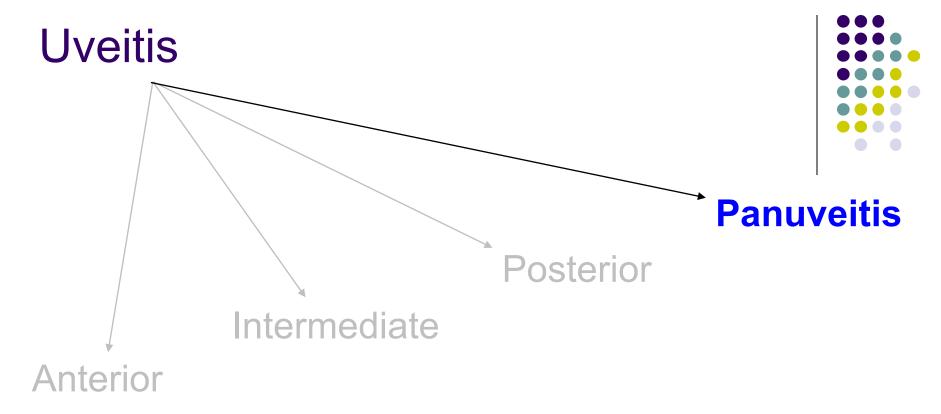


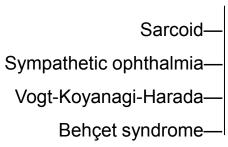
The panuveitides are divvied into \_\_\_\_\_ and \_\_\_\_ causes.

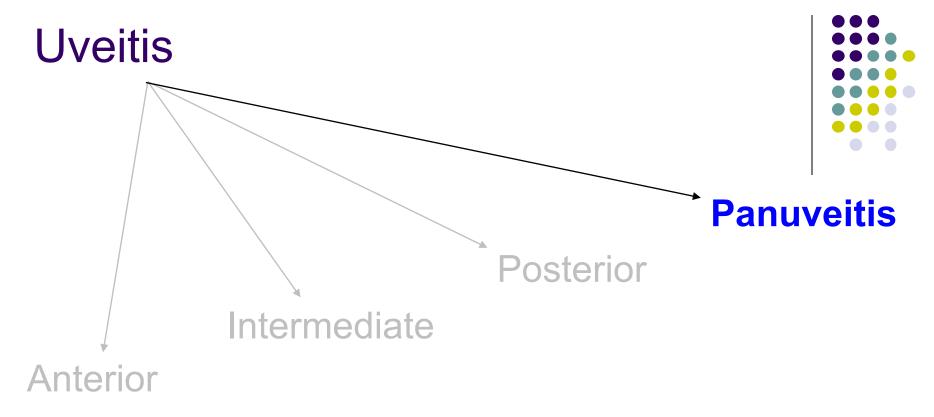


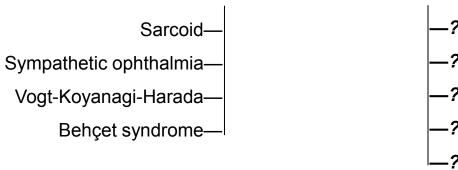


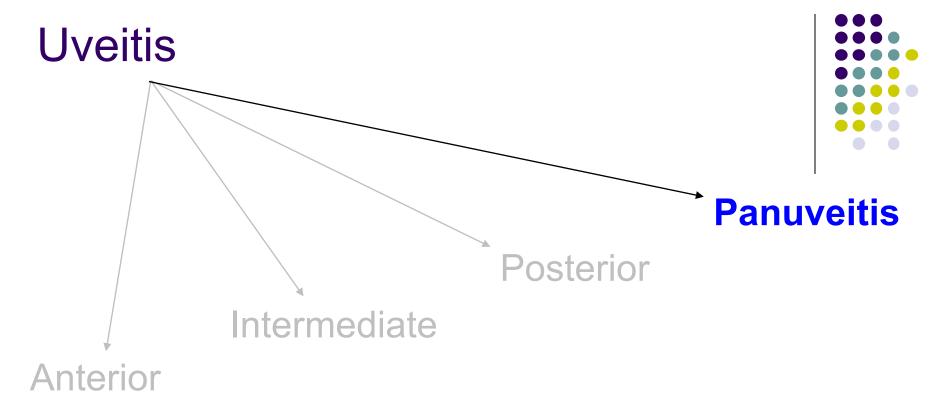


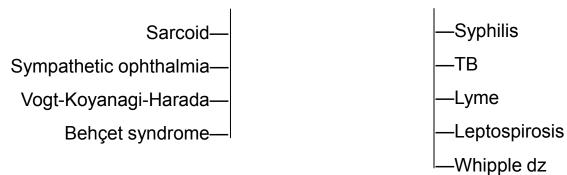


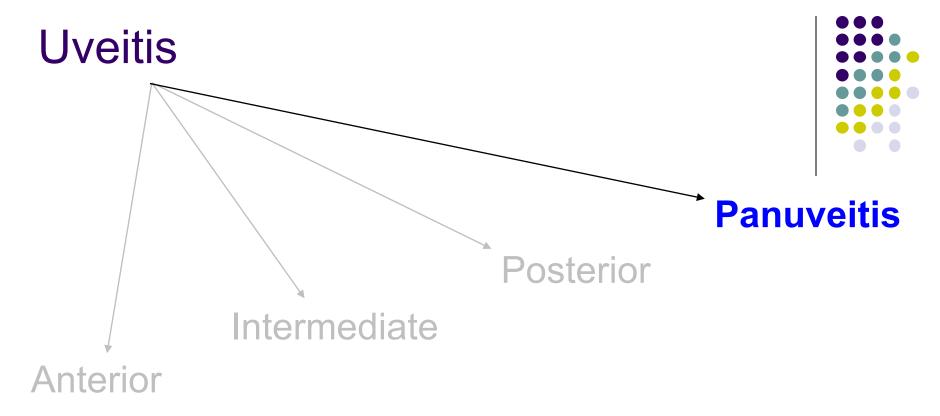












Sarcoid— Sympathetic ophthalmia—	Note that these three appear yet again	—Syphilis —TB
Vogt-Koyanagi-Harada—		—Lyme
Behçet syndrome—		—Leptospirosis
		—Whipple dz



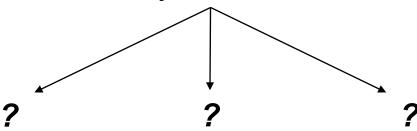
Now we'll change gears and look at endophthalmitis

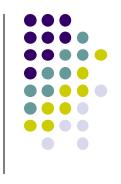


The *Uveitis* book defines **endophthalmitis** as an inflammatory process involving both the AC and vitreous cavities that is 2ndry to or infection.



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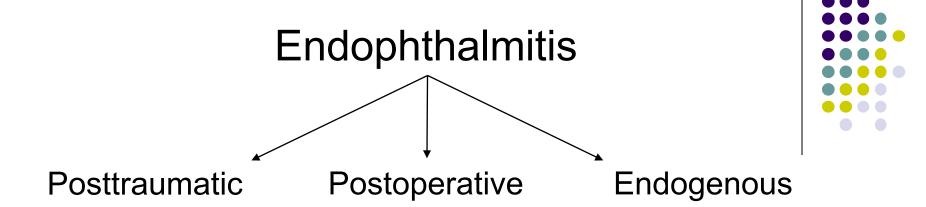
Endophthalmitis can be

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**.



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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!

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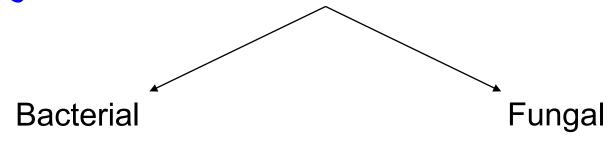
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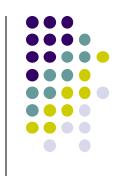
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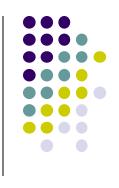
As mentioned, endogenous endophthalmitis can be bacterial or fungal.



As mentioned, endogenous endophthalmitis can be bacterial or fungal. **Bacterial** endophthalmitis presents with the expected ocular signs of \_\_\_\_\_, and

two words





As mentioned, endogenous endophthalmitis can be bacterial or fungal. **Bacterial endophthalmitis** presents with the expected ocular signs of pain, redness, and decreased vision.

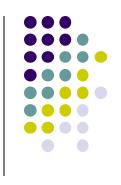


**Bacterial** 

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 two words, a dense three words (often with ), and two words.





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 .

Endog











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. **Retinal** 

may be present, including two-words hemorrhages (aka two words . Systemic findings of infection— , two-words , malaise, etc—will likely be present as well.





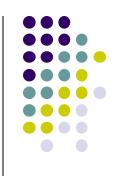
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- --IVDU
- --Liver abscess





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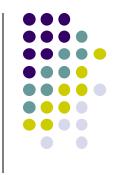
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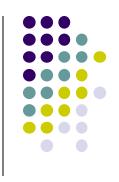
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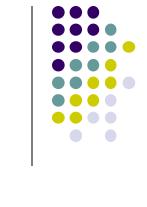
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Bacterial Fungal

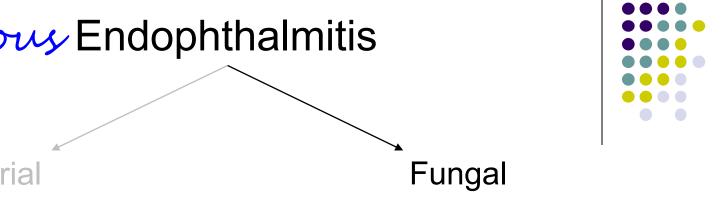
In contrast to the bacterial version, endogenous **fungal** endophthalmitis tends to be more in onset.





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





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Candida endophthalmitis: Choroidal lesions





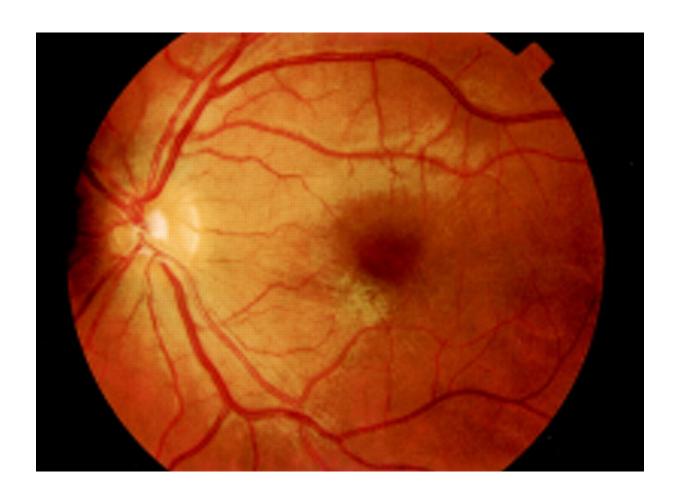
In contrast to the bacterial version, endogenous **fungal** endophthalmitis tends to be more insidious in onset. It generally progresses in a particular fashion. First, isolated choroidal metastatic lesions appear. With time, these break through to involve the retina





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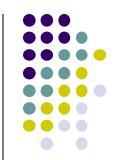


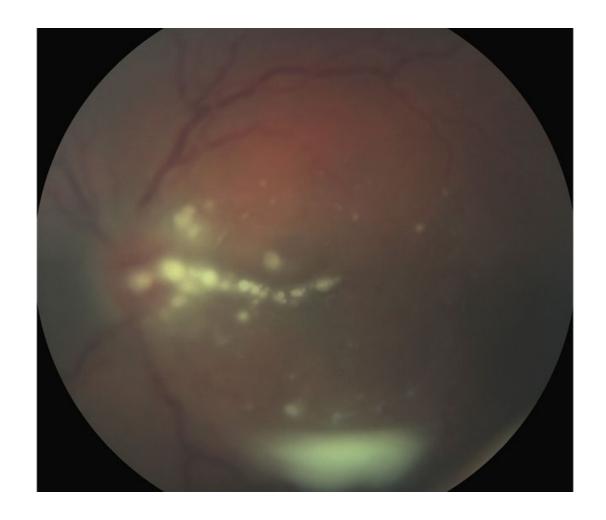
Candida endophthalmitis: Retinal involvement





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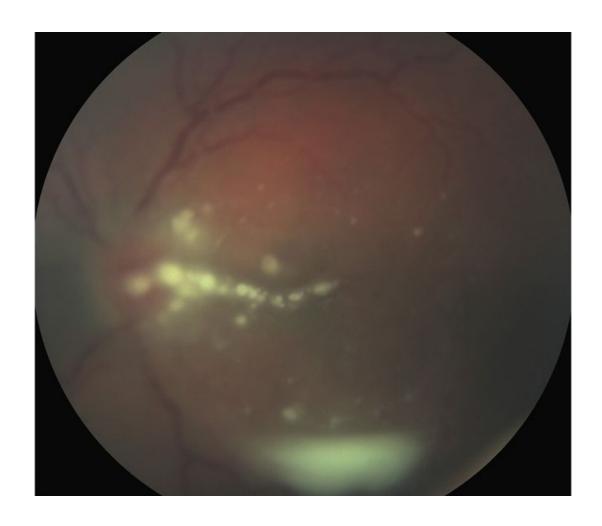


Candida endophthalmitis: Classic

three words

vitreous involvement



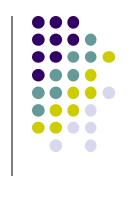


Candida endophthalmitis: Classic 'string of pearls' vitreous involvement



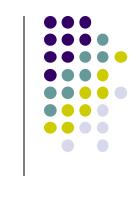
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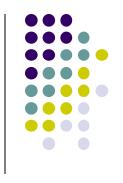
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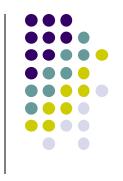




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- --HIV/AIDS
- --Hx liver transplantation
- --San Joaquin valley

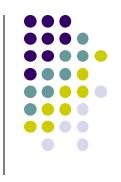




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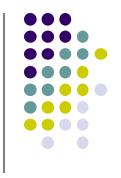




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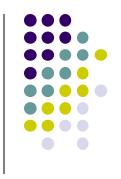




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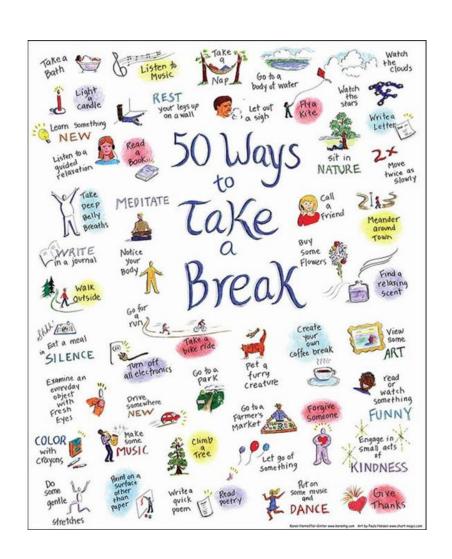
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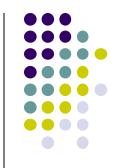
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(This is a good point in the set to take a break)



# Next let's take a look at Scleritis



#### **Scleritis**

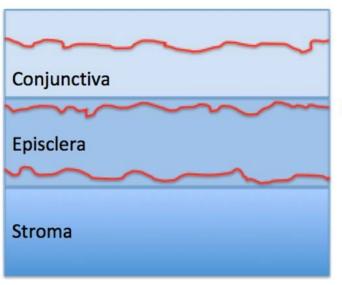
Scleritis is an inflammatory condition characterized by painful infiltrative scleral edema and congestion of the wow words plexus.



#### **Scleritis**

Scleritis is an inflammatory condition characterized by painful infiltrative scleral edema and congestion of the deep episcleral plexus.





Conjunctival Plexus

Superficial Episcleral Plexus

> Deep Vascular Plexus

> > EyeRounds.org

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**



#### **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 of scleritis pts have an identifiable systemic inflammatory condition, the most common of which is

two words



#### **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).



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.



## **Scleritis**



There are three classic signs of anterior scleritis:
--Scleral
---



## **Scleritis**

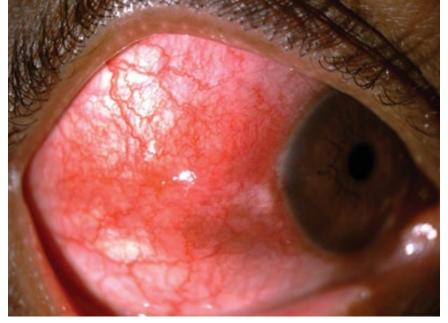


There are three classic signs of anterior scleritis:

--Scleral edema

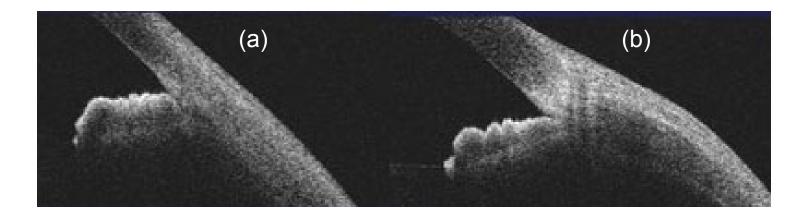
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-





Anterior scleritis: Scleral edema



Anterior scleritis: Scleral edema. Note the thickening of the limbal sclera (b) in comparison to the unaffected fellow eye (a)



## **Scleritis**



There are three classic signs of anterior scleritis:

- --Scleral edema
- --Sclera has a hue

--



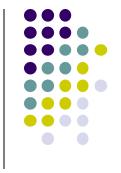
## **Scleritis**



There are three classic signs of anterior scleritis:

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Anterior scleritis: Violaceous hue



## **Scleritis**



There are three classic signs of anterior scleritis:

- --Scleral edema
- --Sclera has a violaceous hue
- --Inflamed vasculature has a pattern

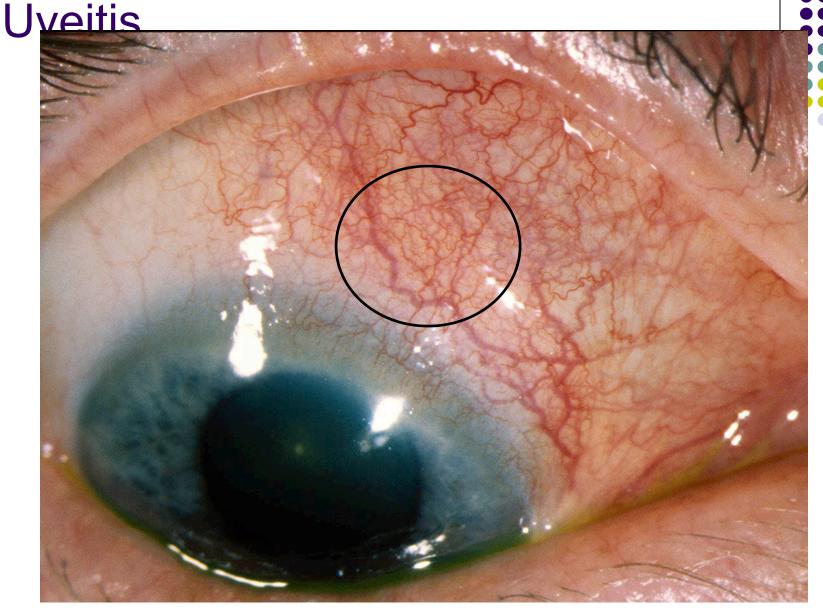






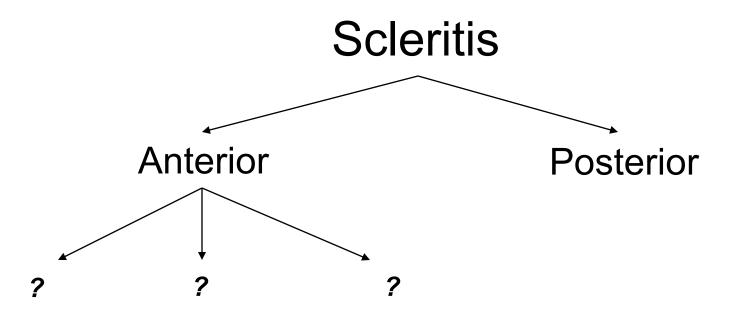
There are three classic signs of anterior scleritis:

- --Scleral edema
- --Sclera has a violaceous hue
- --Inflamed vasculature has a **criss-cross** pattern



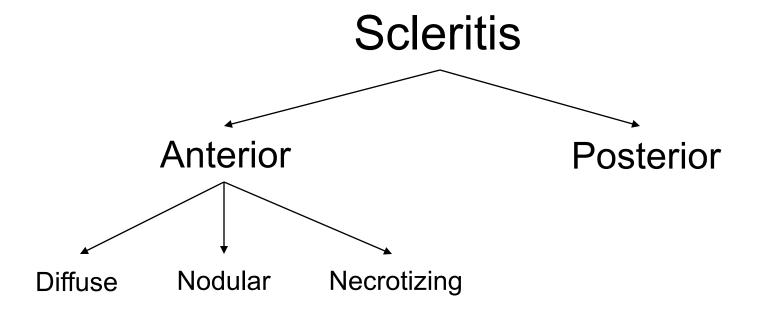
'Criss-cross' injection of the deep vasculature in anterior scleritis. (To see it, you have to 'look past' the brighter injection of the inflamed overlying conj vessels)





Anterior scleritis comes in three forms:



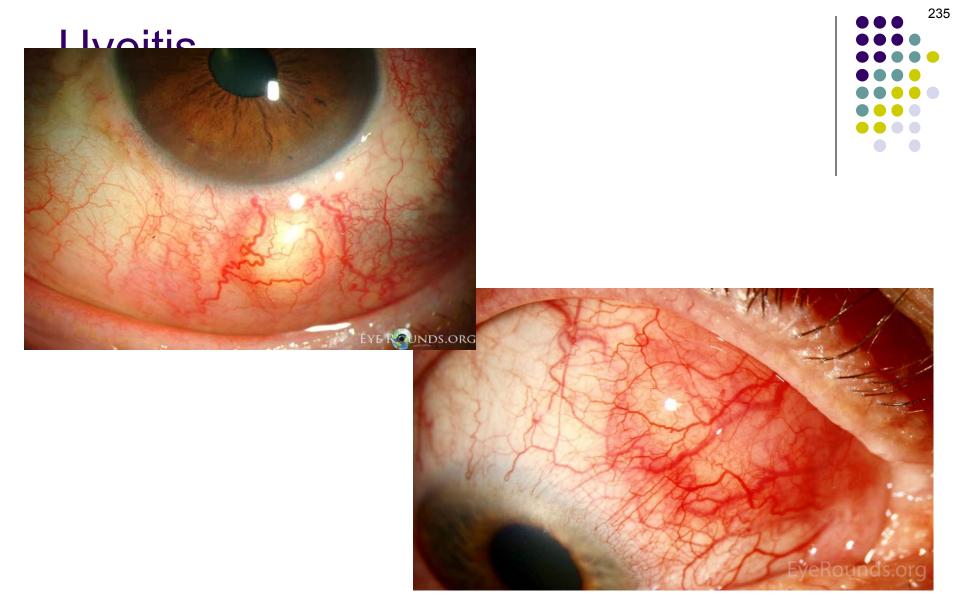


Anterior scleritis comes in three forms: *Diffuse, nodular* and *necrotizing* 



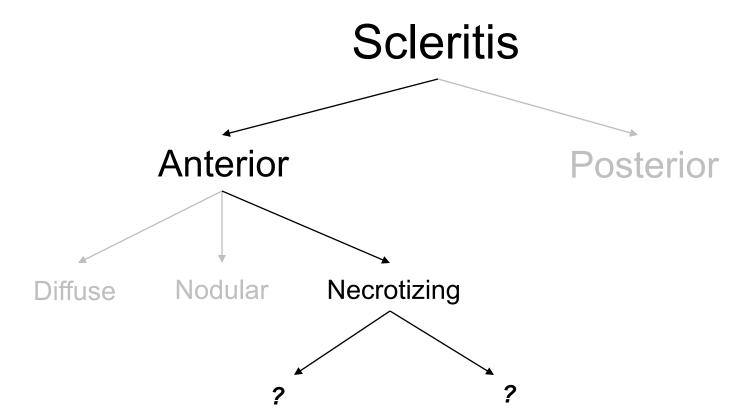


Diffuse anterior scleritis



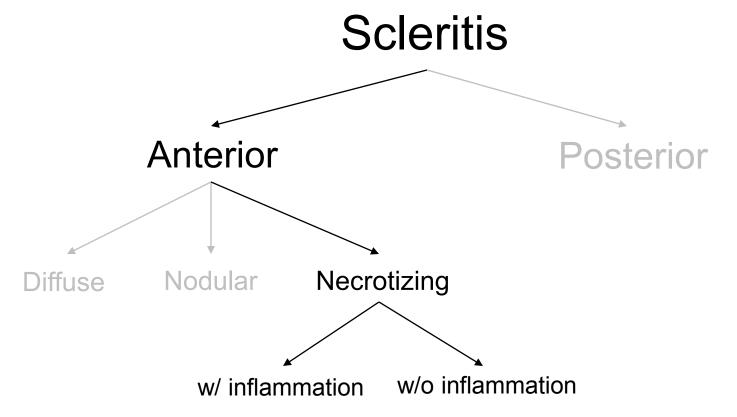
Nodular anterior scleritis





Necrotizing anterior scleritis comes in two forms:





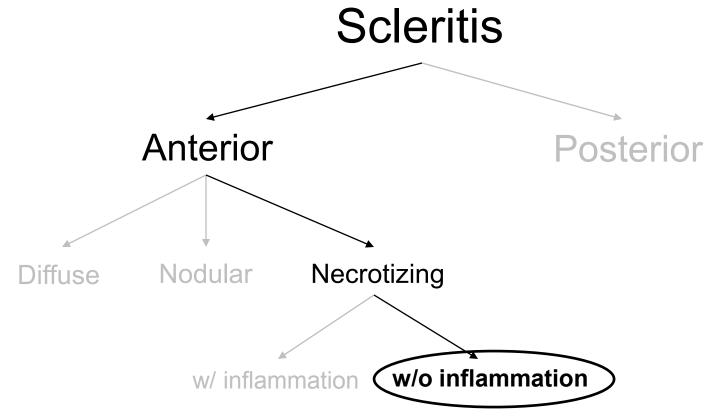
Necrotizing anterior scleritis comes in two forms: With and without inflammation



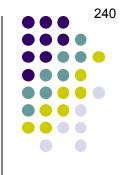


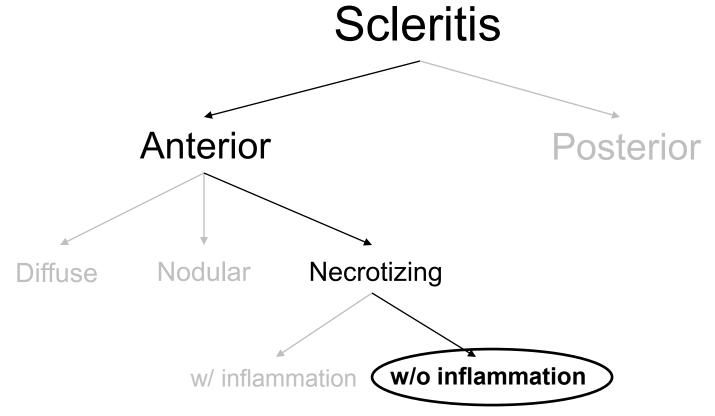
Necrotizing anterior scleritis with inflammation





Contrary to the implications of the name, inflammation **is** present in *necrotizing scleritis w/o inflammation*.





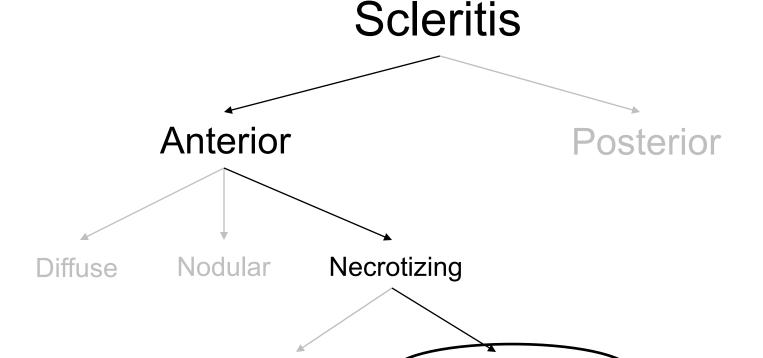
Contrary to the implications of the name, inflammation **is** present in *necrotizing scleritis w/o inflammation*. It is so named because, unlike its 'with inflammation' cousin, it is typically painless, and the eye does not *appear* inflamed.





Necrotizing anterior scleritis without inflammation





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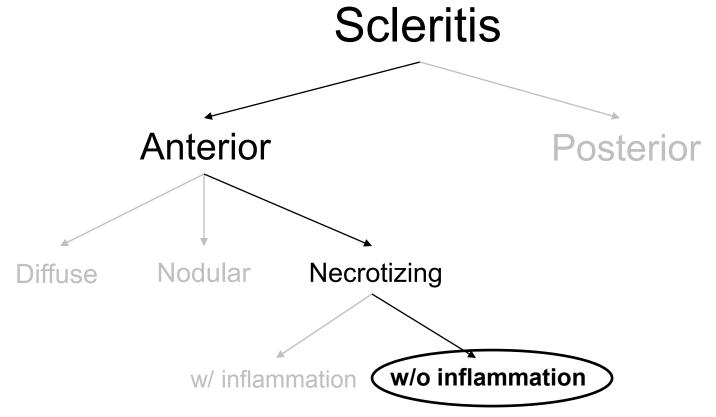
inflammation is also known as strongly associated with

w/ inflammation

two words

. It is

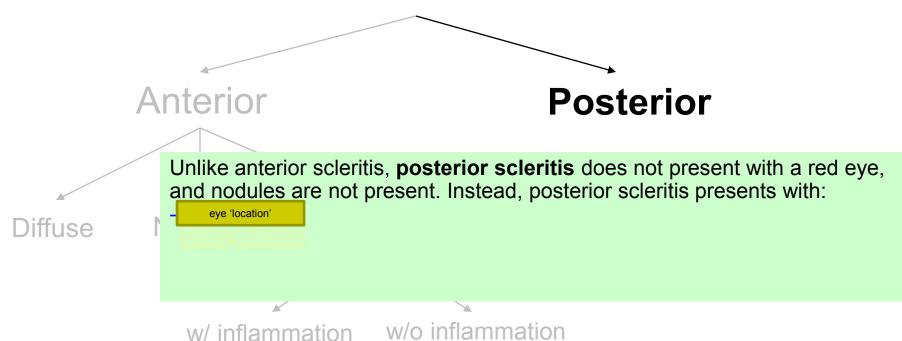




Contrary to the implications of the name, inflammation is present in *necrotizing scleritis w/o inflammation*. It is so named because, unlike its 'with inflammation' cousin, it is typically painless, and the eye does not *appear* inflamed. Necrotizing scleritis w/o inflammation is also known as **scleromalacia perforans**. It is strongly associated with RA.

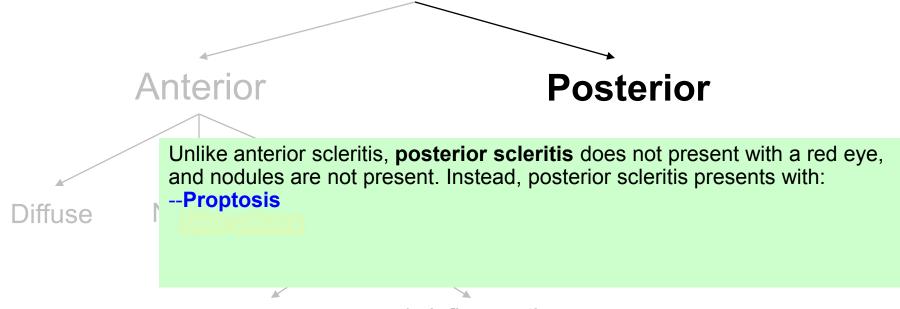


## **Scleritis**



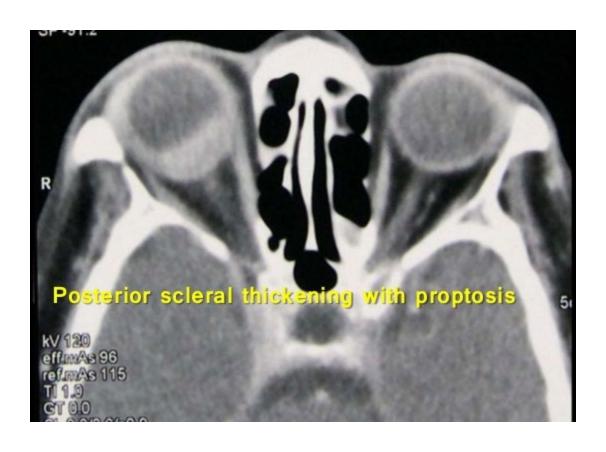


#### **Scleritis**



w/ inflammation w/o inflammation

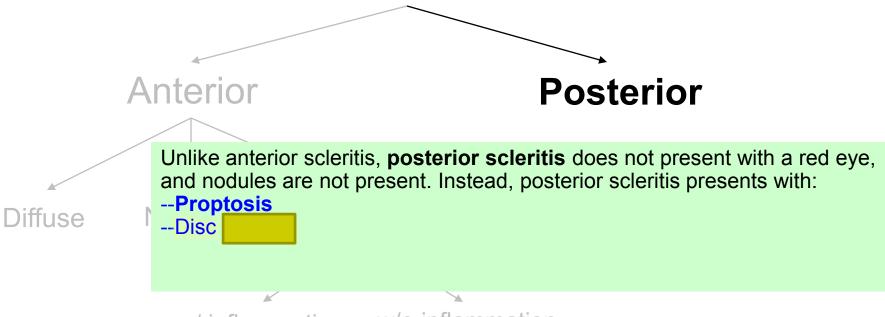




Posterior scleritis: Proptosis



#### **Scleritis**

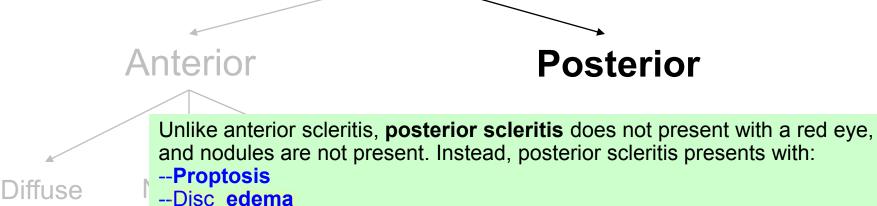


w/ inflammation w/o i

w/o inflammation



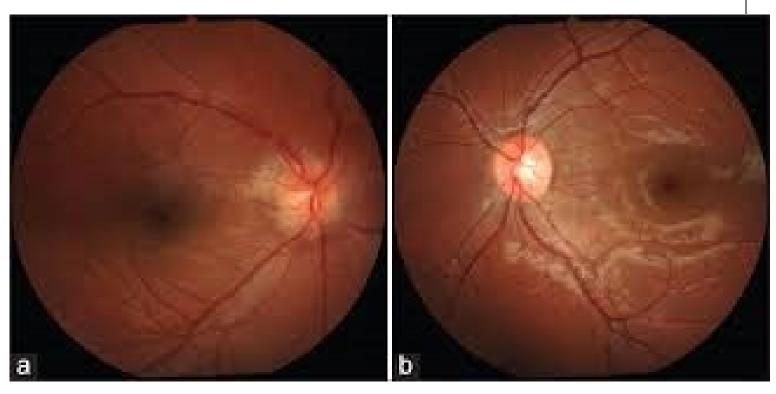




w/ inflammation

w/o inflammation





Posterior scleritis OD: Optic nerve edema



#### **Scleritis**



#### **Posterior**

and nodules are not present. Instead, posterior scleritis presents with: -- Proptosis Diffuse

--Disc edema

--Retinal/choroidal findings

w/ inflammation

w/o inflammation

Unlike anterior scleritis, posterior scleritis does not present with a red eye,

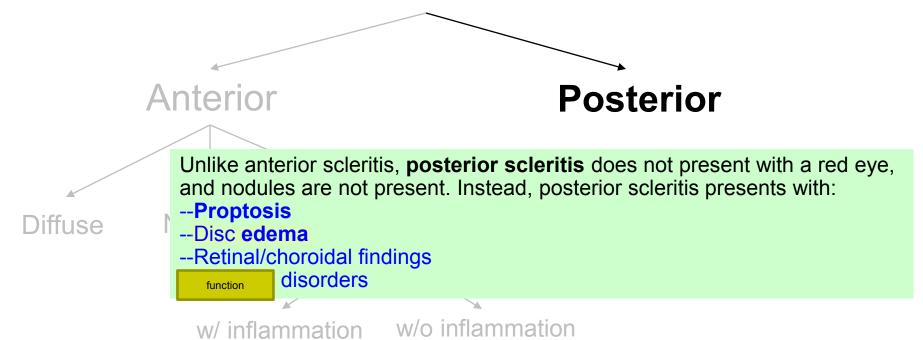




Posterior scleritis producing retinal folds



#### **Scleritis**





#### Scleritis



#### **Posterior**

Diffuse

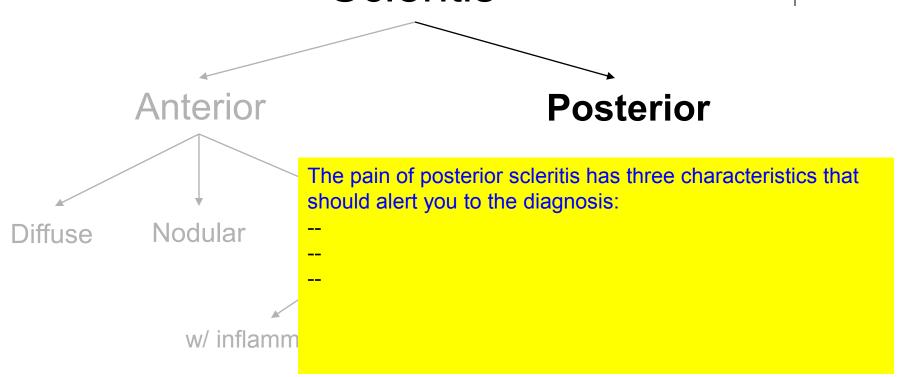
Unlike anterior scleritis, **posterior scleritis** does not present with a red eye, and nodules are not present. Instead, posterior scleritis presents with:

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

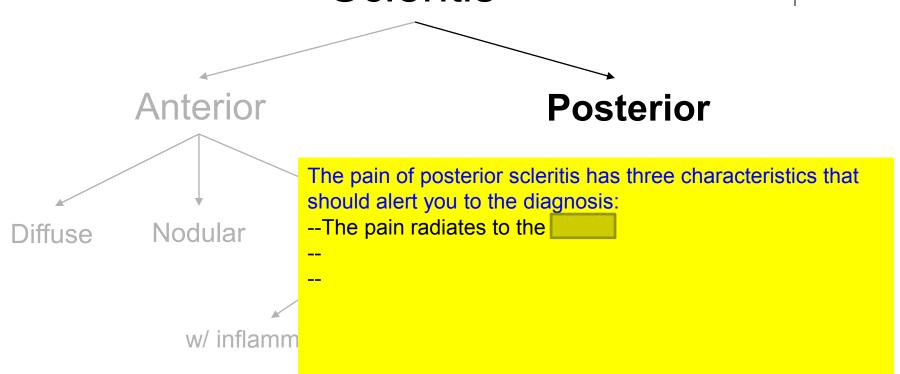
w/ inflammation

w/o inflammation

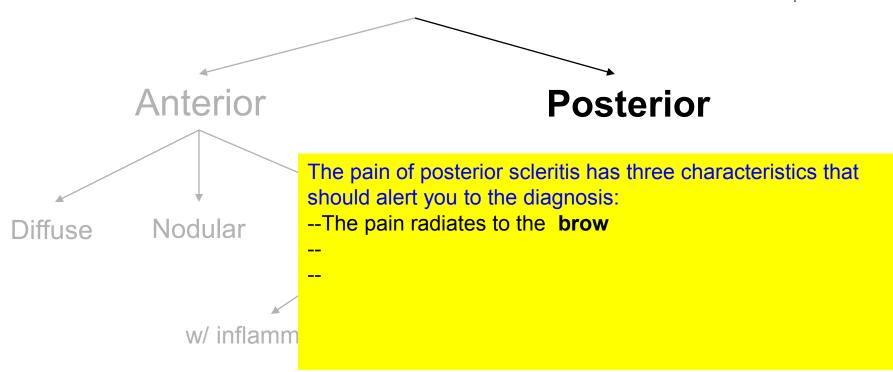




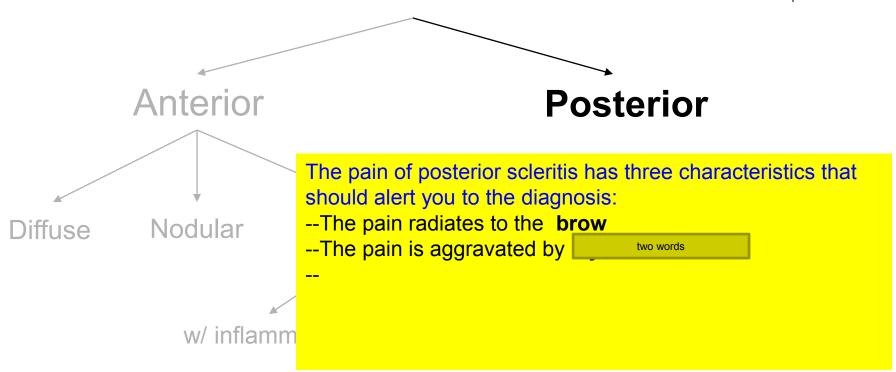




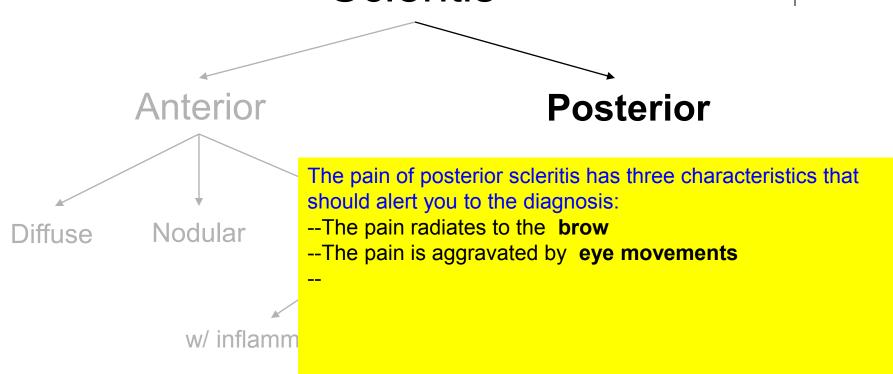




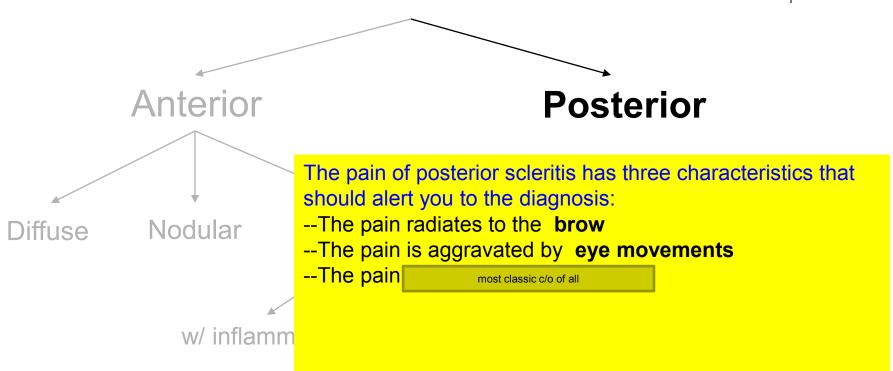




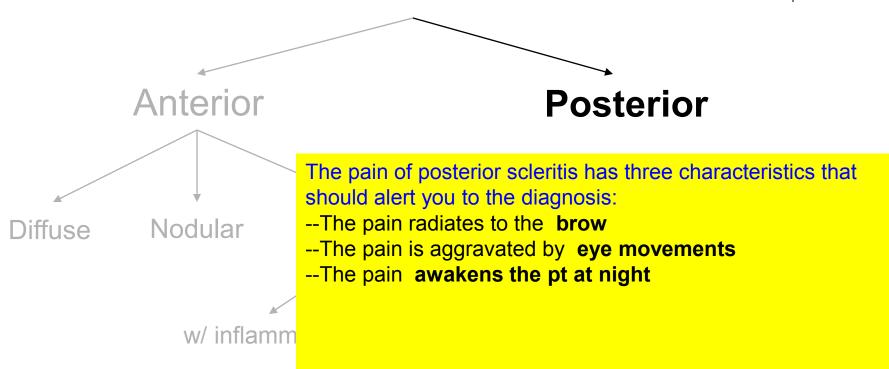






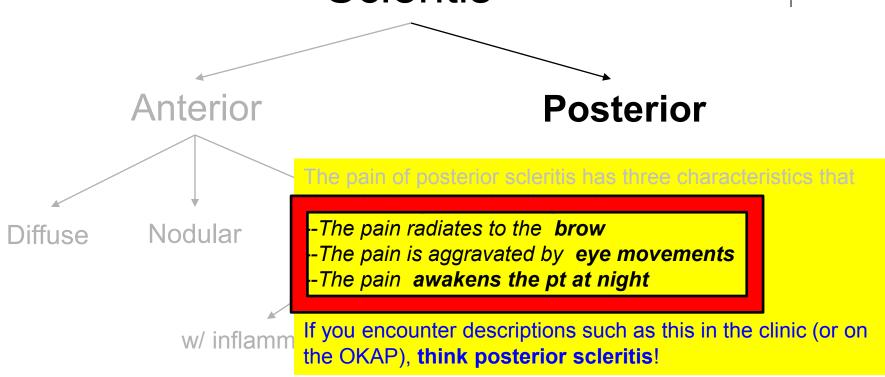




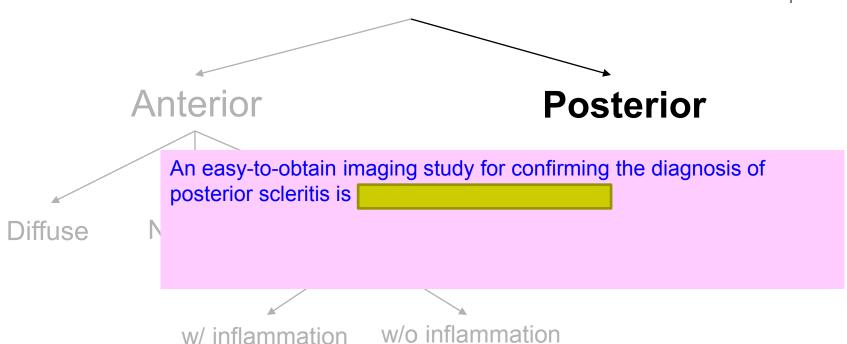




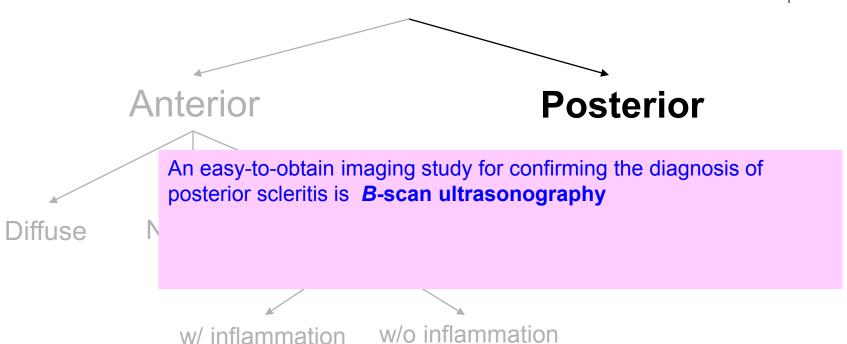






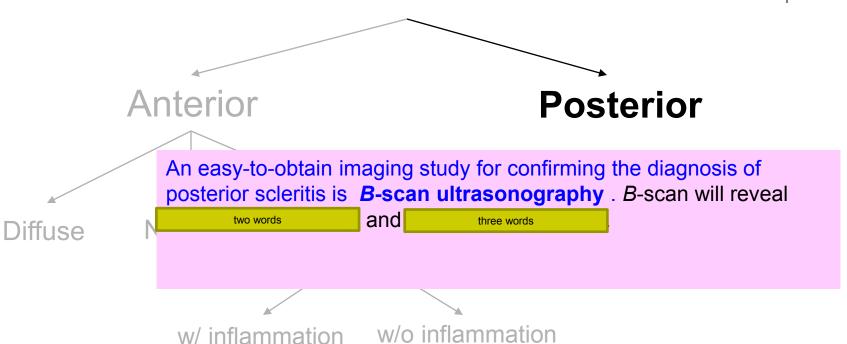












Diffuse



#### **Scleritis**



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.

w/ inflammation w/o inflammation





Posterior scleritis: Choroidal thickening; sub-Tenon's edema



#### **Scleritis**



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 involves the space around the optic nerve, the classic finding will result.

w/ inflammation w/o inflammation



#### **Scleritis**

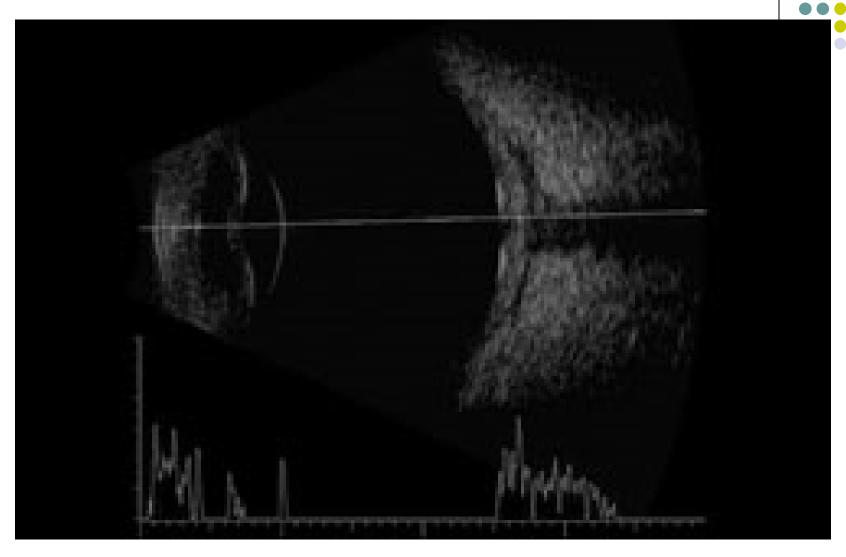


Diffuse

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 involves the space around the optic nerve, the classic **T sign** finding will result.

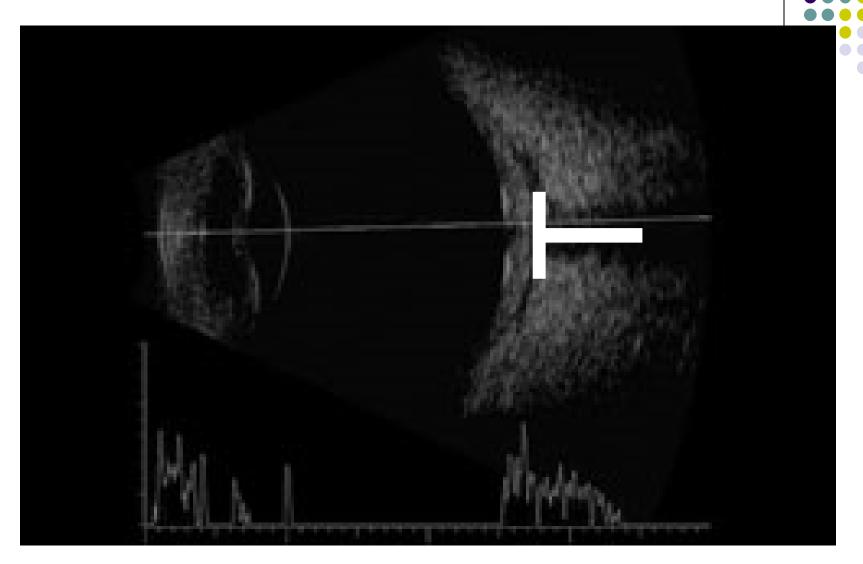
w/ inflammation

w/o inflammation



269

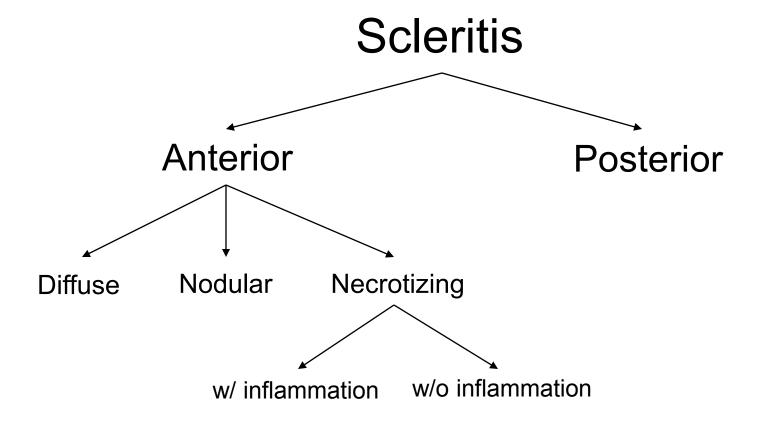
Posterior scleritis: T-sign (advance to next slide if you don't see it)



270

Posterior scleritis: T-sign

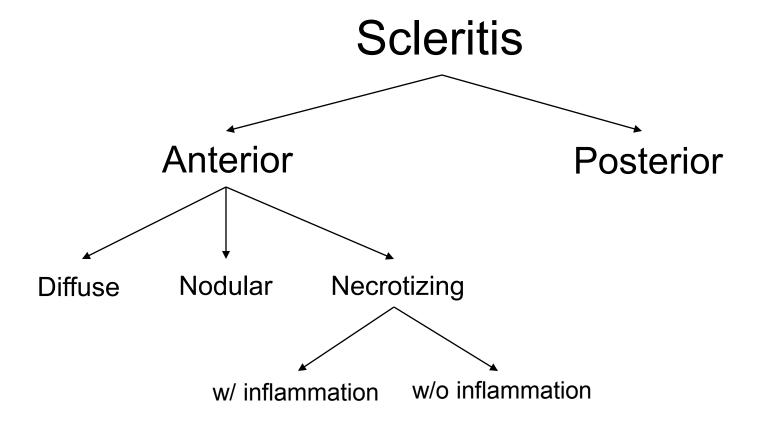




**Scleritis requires systemic treatment**. Diffuse scleritis might respond to so try them first if not contraindicated.

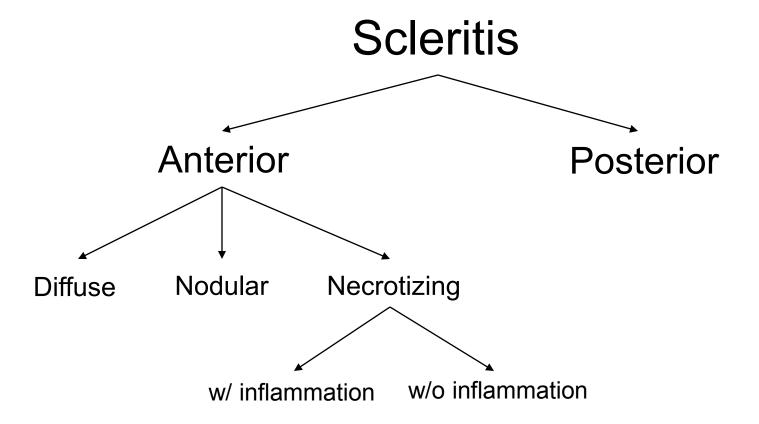
abb.





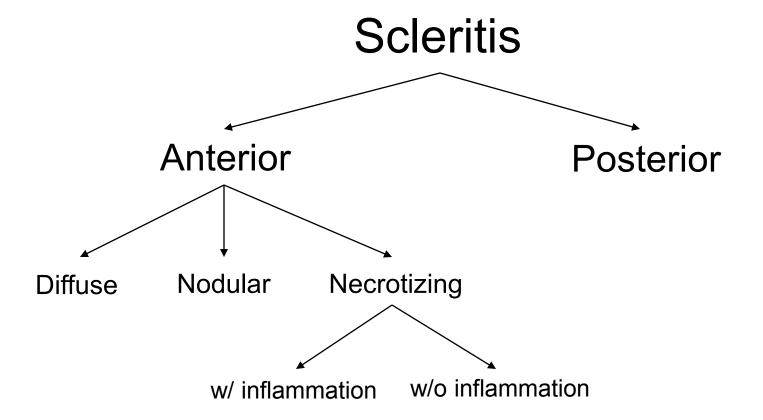
**Scleritis requires systemic treatment**. Diffuse scleritis might respond to PO NSAIDs, so try them first if not contraindicated.





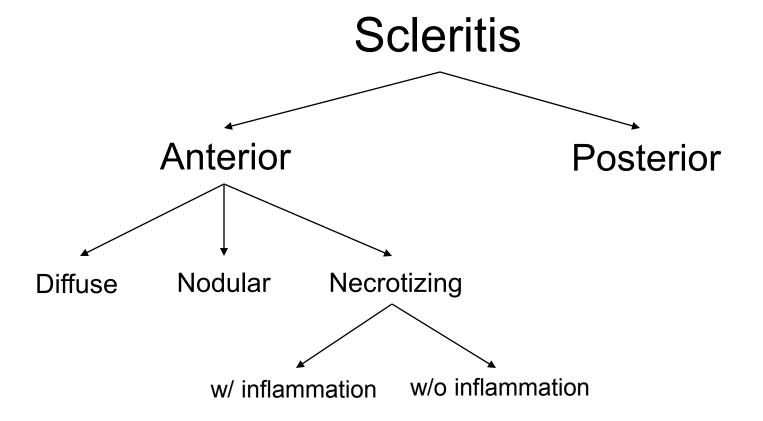
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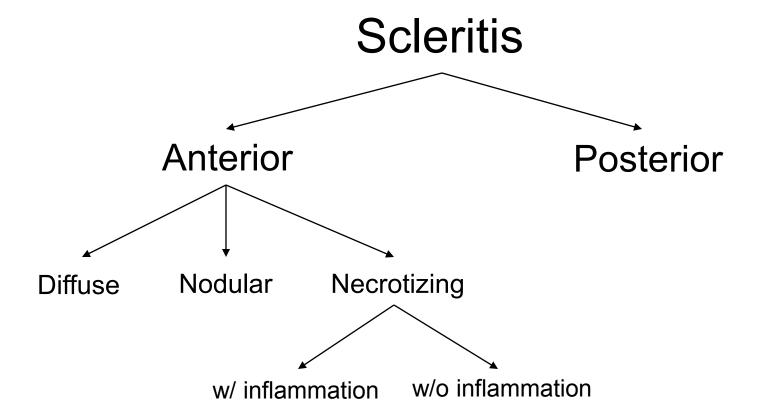




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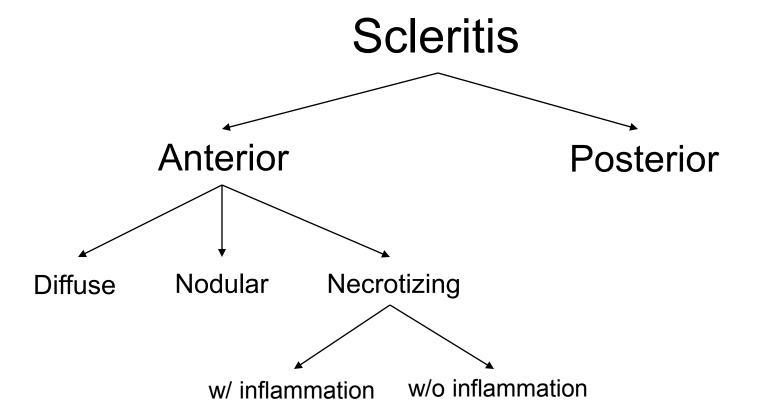
two words is often required.





**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.

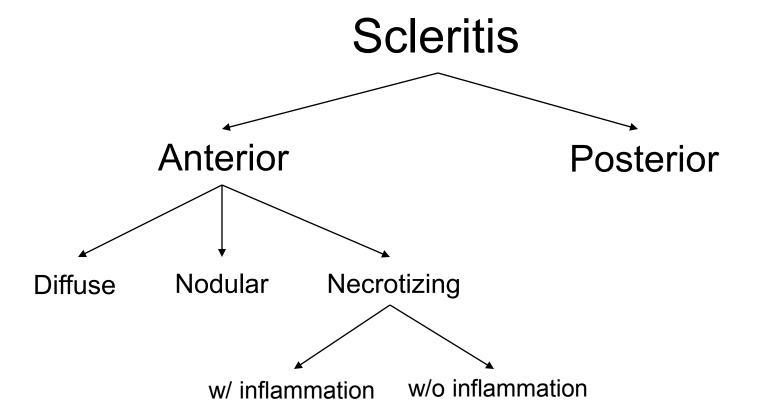




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steroids, long considered contraindicated, have recently gained wide acceptance as a treatment option.





**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.

Subconj depot steroids, long considered contraindicated, have recently gained wide acceptance as a treatment option.



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?



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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What was the task of the SUN Working Group?



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



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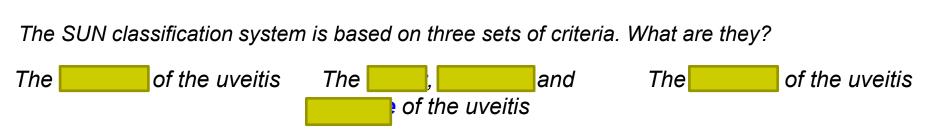
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course of the uveitis

The location of the uveitis The onset, duration and The

The **severity** of the uveitis



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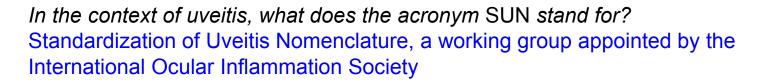
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The **location** of the uveitis --?

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

What are the four locations?



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

The onset, duration and course of the uveitis

The **severity** of the uveitis

- --Anterior
- --Intermediate
- --Posterior
- --Panuveitis

What are the four locations?





What was the task of the SUN Working Group?
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The SUN classification system is based on three sets of criteria. What are they?

The location of the uveitis --Anterior?

The onset, duration and course of the uveitis

The **severity** of the uveitis

- --Intermediate
- --Posterior
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With respect to uveitis: Where is the primary location of inflammation in...

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- --Intermediate
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- --Panuveitis

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 uveitis?



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

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Intermediate uveitis? The main vitreous cavity, +/- the peripheral retina and/or pars plana

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

The **severity** of the uveitis

--Anterior

--Intermediate



--Panuveitis

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Posterior uveitis? The retina, choroid, and/or optic nerve head

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The **location** of the uveitis

The onset, duration and course of the uveitis

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--Anterior

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**Panuveitis?** 





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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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The location of the uveitis The onset, duration and

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

--Anterior
--Intermediate
--Pasterior
--Panuveitis

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

**Posterior** *uveitis?* The retina and/or choroid

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

Th<del>e location</del> of the uveitis

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

-Anterior

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)

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

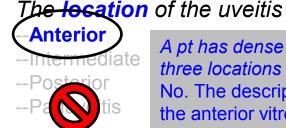
Posterior uveitis? The retina and/or choroid

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the uveitis The onset, duration and

The **onset**, **duration** and 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?

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 <u>cell</u> 'spillover' into the anterior vitreous may occur)

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#### The **location** of the uveitis

- --Anterior
- --Intermediate
- --Posterior
- --Panuveitis

The onset, duration and course of the uveitis

--Onset:

something vs something

- -- Duration
- --Course

How are onset, duration and course delineated?





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The onset, duration and course of the uveitis

- -- Onset: Sudden vs insidious
- --Duration
- --Course

The **severity** of the uveitis

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# The onset, duration and course of the uveitis

- --Onset: Sudden vs insidious
- -- Duration: Limited vs persistent
- --Course

How are onset, duration and course delineated?





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

- -- Onset: Sudden vs insidious
- -- Duration: Limited vs persistent
- --Course: Acute vs recurrent vs chronic

How are onset, duration and course delineated?

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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

With respect to uveitis, what is the difference between... Sudden vs insidious onset?





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The **severity** of the uveitis

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); <u>insidious uveitis</u> is largely sign- and symptom-free

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- --Intermediate
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Limited vs persistent duration? <u>Limited</u> lasts < 3 months; <u>persistent</u> > 3 months Acute, recurrent vs chronic course? Acute uveitis comes on suddenly and resolves fairly quickly

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- --Anterior
- --Intermediate
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- -- Onset: Sudden vs insidious
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- --Course: Acute vs recurrent vs
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Limited vs persistent duration? Limited lasts < 3 months; persistent > 3 months

Acute, recurrent vs chronic course? Acute uveitis comes on suddenly and resolves fairly quickly.

Recurrent uveitis eventually relapses, but is quiescent off-treatment for at least 3 months.

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Limited vs persistent duration? Limited lasts < 3 months; persistent > 3 months

Acute, recurrent vs chronic course? Acute uveitis comes on suddenly and resolves fairly quickly.

<u>Recurrent uveitis</u> eventually relapses, but is quiescent off-treatment for at least 3 months. <u>Chronic uveitis</u> also relapses, but its quiescent periods off-treatment last **less** than 3 months.



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How is severity determined?





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To standar	Grade	Number of Cells/Field	
The SUN of The location Anterior Intermedia Posterior Panuveitis	?	?	ts of criteria. What are they?  on and  The severity of the uveitis AC cell grade
	?	?	
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	3+	26-50	current vs
	4+	>50	

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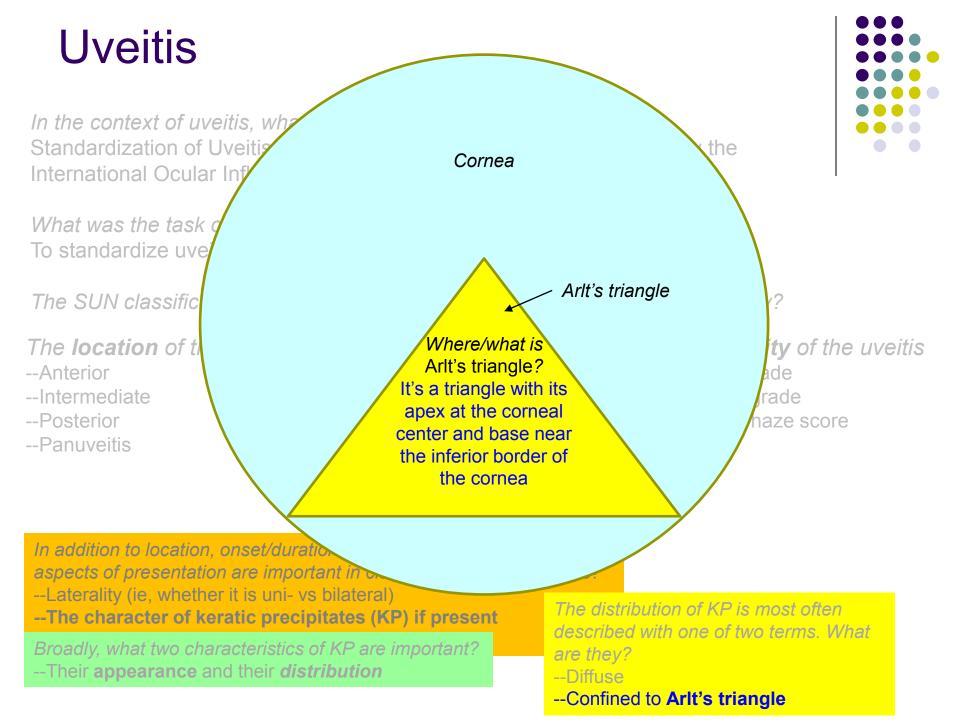
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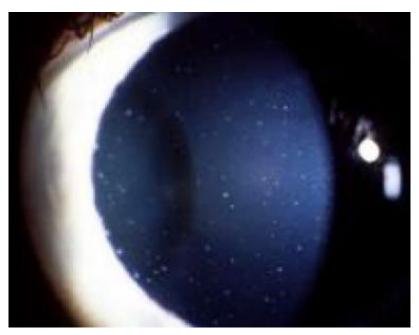
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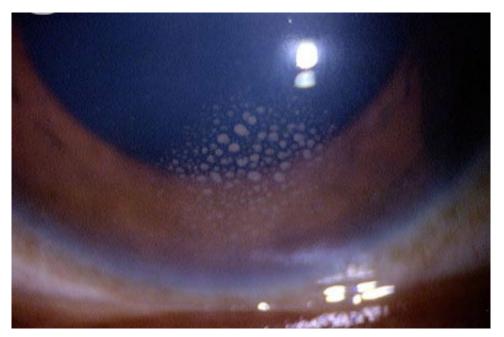
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Diffusely distributed KP



KP concentrated in Arlt's triangle

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- --Nongranulomatous
- --Stellate

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- Always...

Always...

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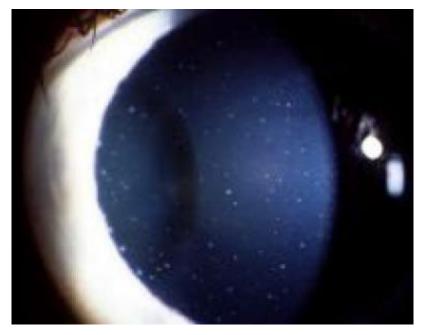
--Nongranulomatous

--Stellate

Always...in **Arlt's triangle** 

Always...diffuse







Stellate KP

Granulomatous KP

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- --Synechiae
- --Nodules
- --Heterochromia





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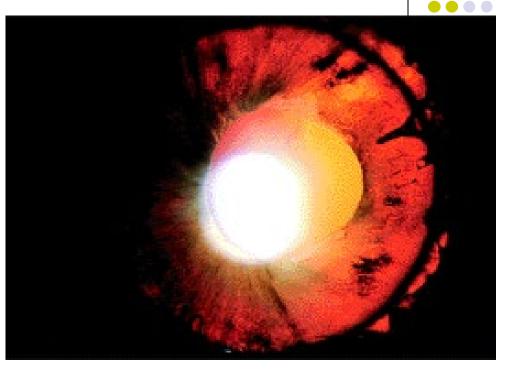
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Sectoral



Diffuse

Iris atrophy



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--At the pupillary margin (these are called

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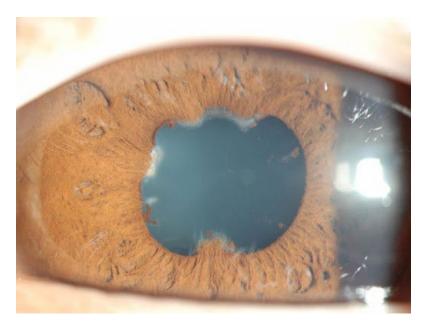
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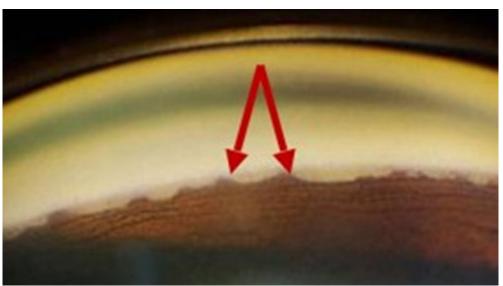
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Posterior

Peripheral anterior

Iris synechiae

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#### The **severity** of the uveitis

- --AC cell grade
- --AC flare grade
- --Vitreous haze score

-- Iris changes

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

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

- --Synechiae
- --Nodules

--Heterochro --

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

- --Anterior
- --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
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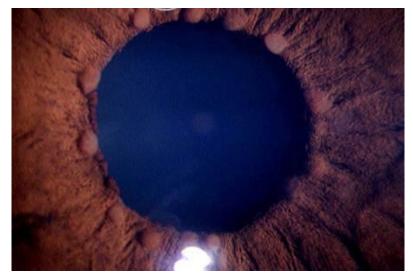
--At the pupillary margin

--Nodules

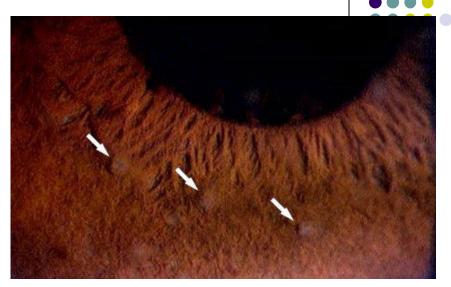
-- The mid-iris

--Heterochro

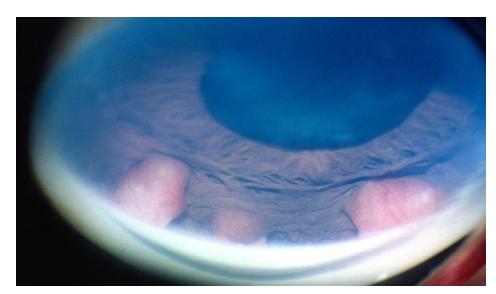
--At the angle



Marginal



Mid-iris



Near the angle

Iris nodules





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

--Laterality (ie

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

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- --Synechiae
- --Nodules

--Atrophy

--Heterochromia

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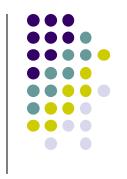
In a pt with uveitis and heterochromia, which iris is more likely to be the abnormal one--the lighter iris, or the darker? The lighter





Heterochromia iridis in a uveitis pt

# Masquerade Syndrome



Finally, we will look at masquerade syndromes

# Masquerade Syndrome refers to entities



that mimic immune-mediated dz.

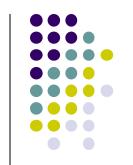
# Masquerade Syndrome refers to entities



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In other words, these are pts who look like they have a uveitic condition, but do not.

# Masquerade Syndrome refers to entities



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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.

# Masquerade Syndrome refers to entities



that mimic immune-mediated dz.

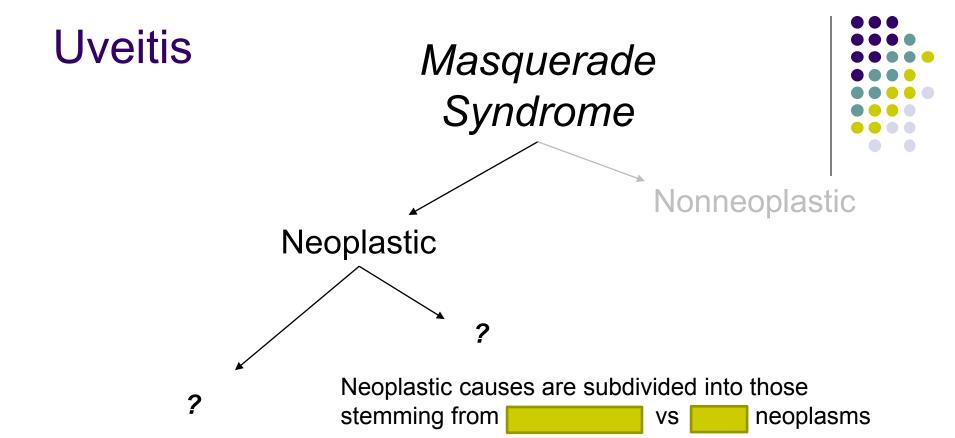
The entities can be broadly divided into			
and		causes.	

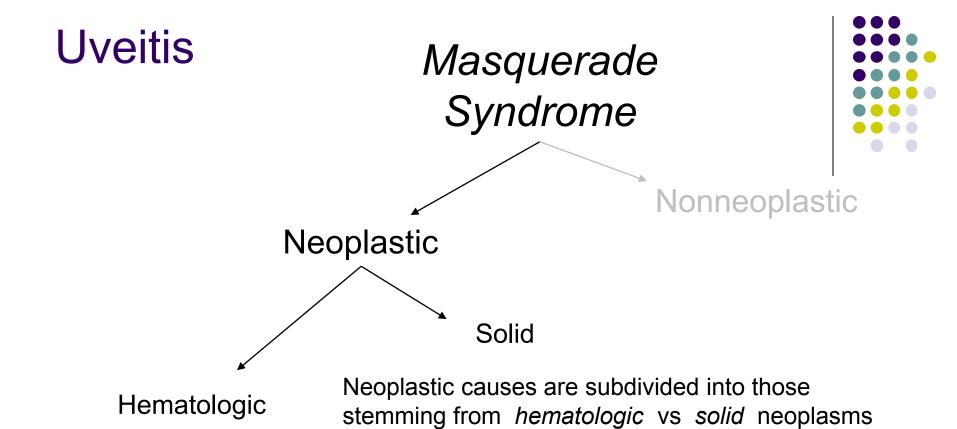
# Masquerade Syndrome refers to entities

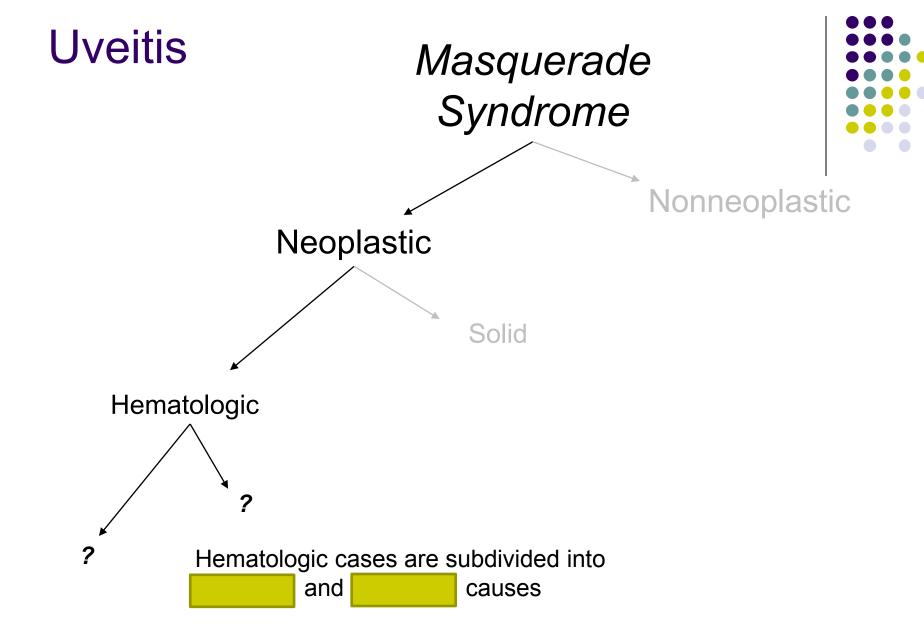


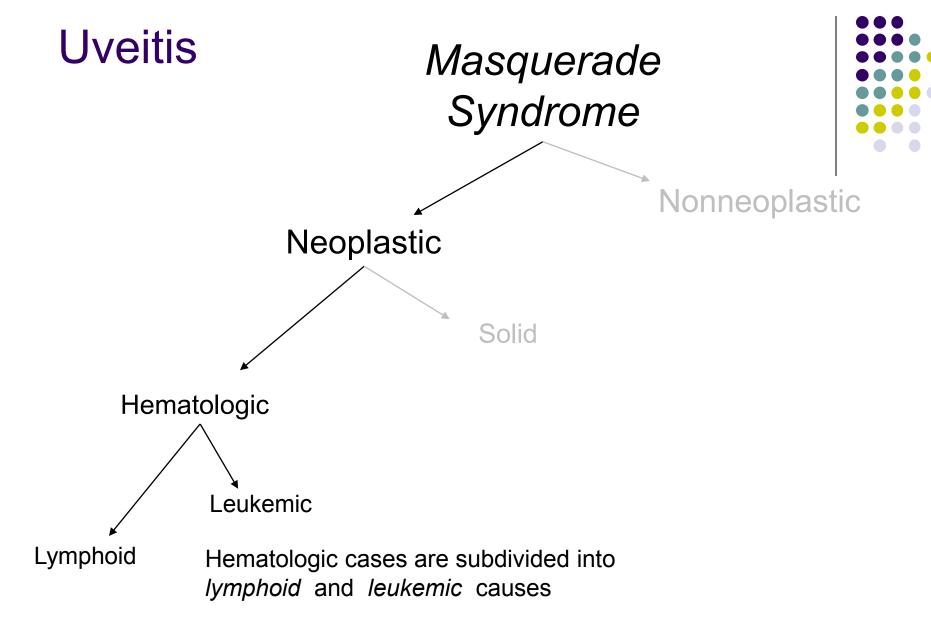
that mimic immune-mediated dz.

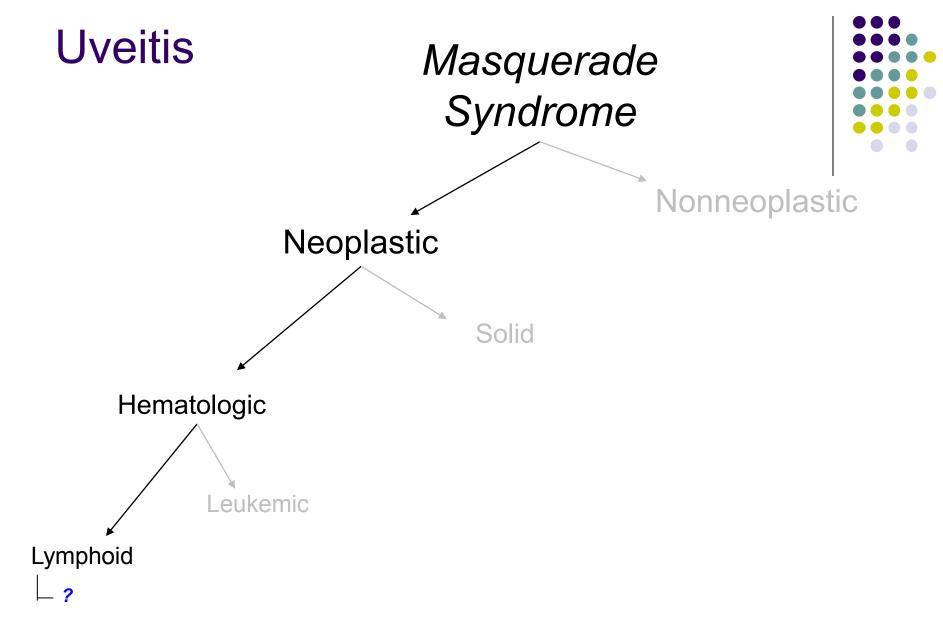
The entities can be broadly divided into Nonneoplastic and Neoplastic causes.



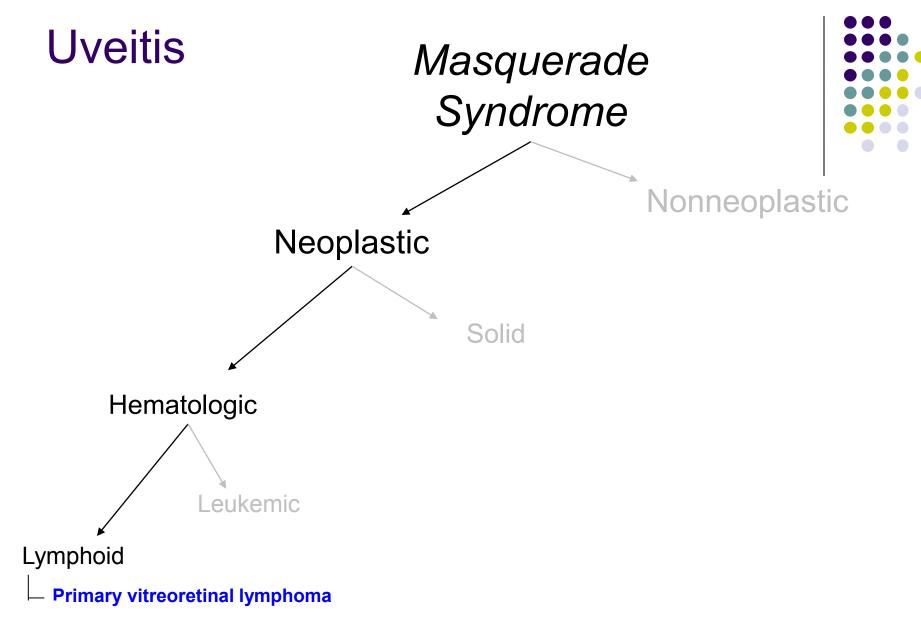


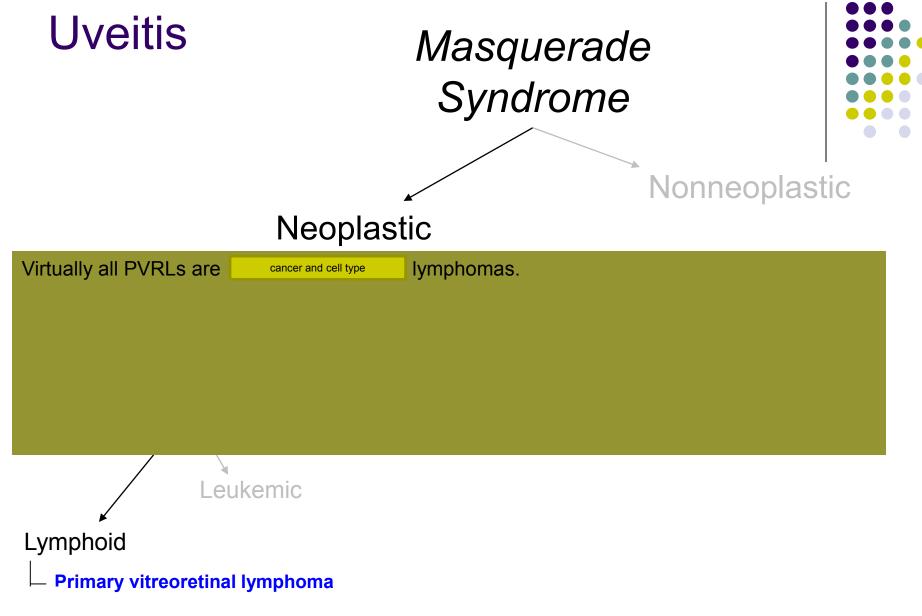






The most common entity to masquerade as intraocular uveitis is





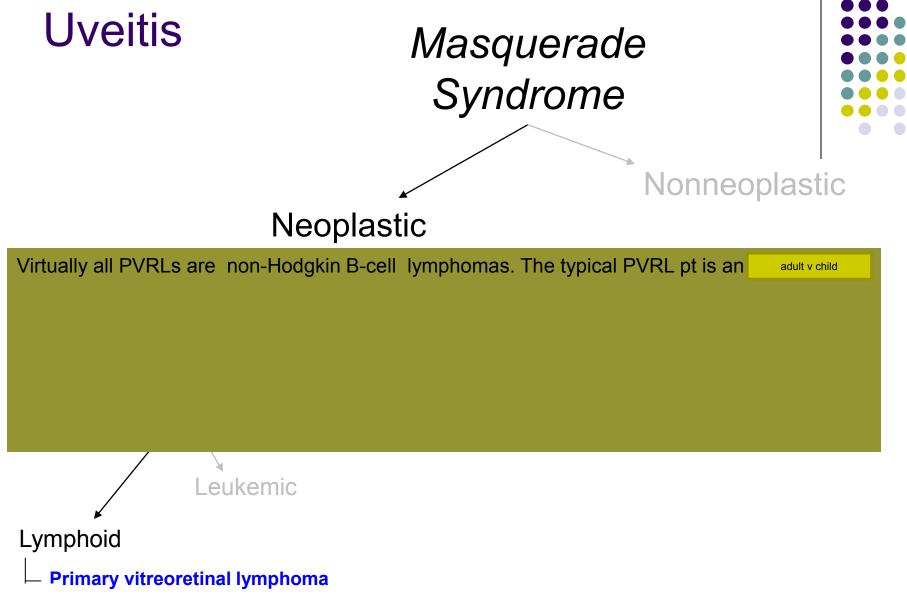
# **Uveitis** Masquerade Syndrome Nonneoplastic Neoplastic Virtually all PVRLs are non-Hodgkin B-cell lymphomas.

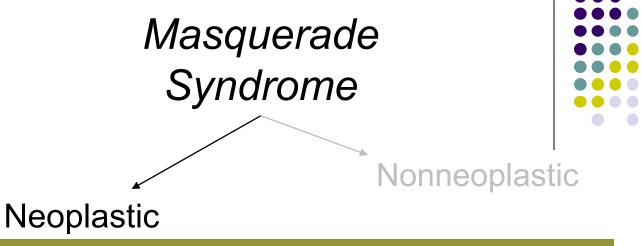
The most common entity to masquerade as intraocular uveitis is **primary vitreoretinal lymphoma** (PVRL)

Leukemic

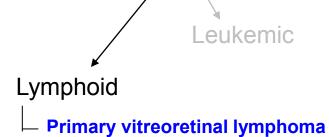
**Primary vitreoretinal lymphoma** 

Lymphoid





Virtually all PVRLs are non-Hodgkin B-cell lymphomas. The typical PVRL pt is an adult

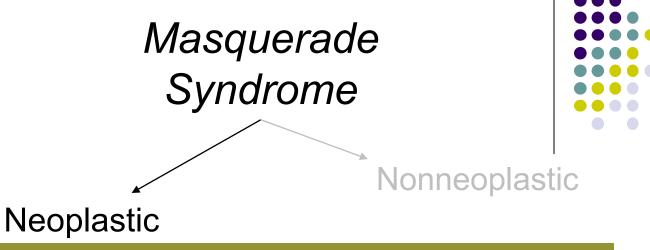


## **Uveitis** Masquerade Syndrome Nonneoplastic Neoplastic Virtually all PVRLs are non-Hodgkin B-cell lymphomas. The typical PVRL pt is an adult in their age range Leukemic

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

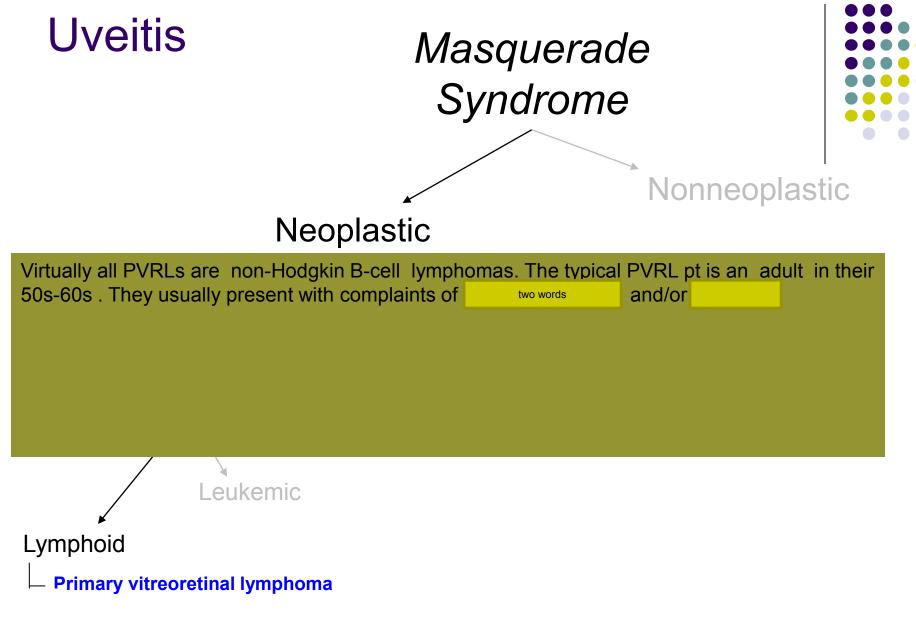
Lymphoid

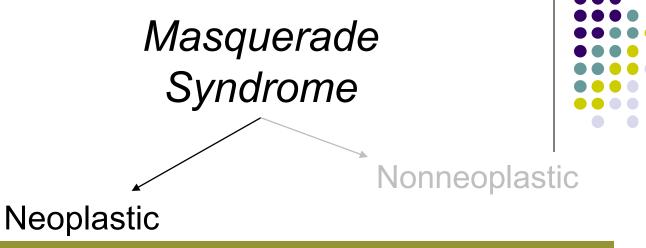
**Primary vitreoretinal lymphoma** 



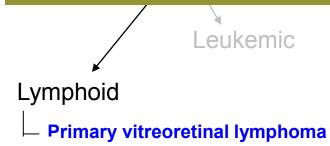
Virtually all PVRLs are non-Hodgkin B-cell lymphomas. The typical PVRL pt is an adult in their 50s-60s.

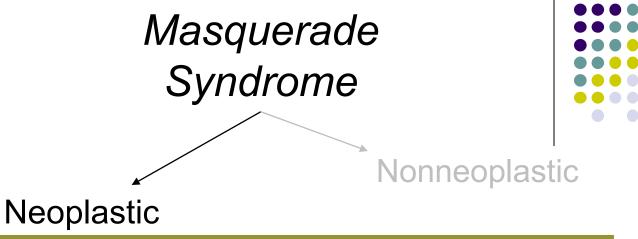






Virtually all PVRLs are non-Hodgkin B-cell lymphomas. The typical PVRL pt is an adult in their 50s-60s. They usually present with complaints of decreased vision and/or floaters.



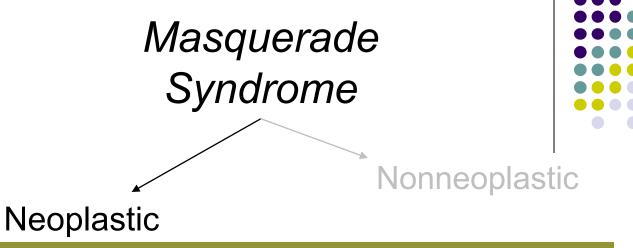


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Importantly, many will also manifest evidence of

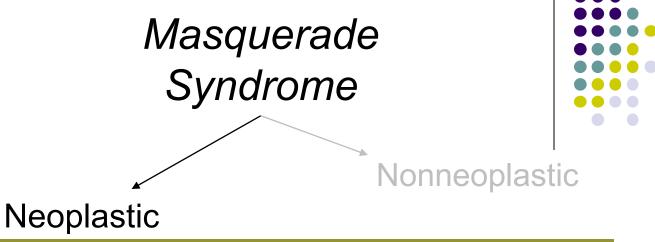
abb. + word





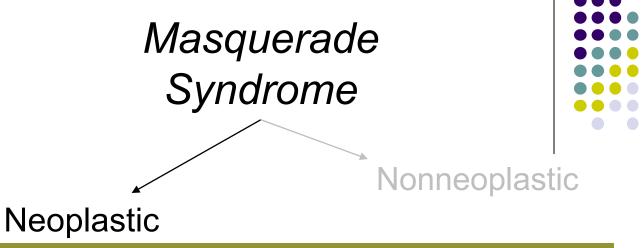
Virtually all PVRLs are non-Hodgkin B-cell lymphomas. The typical PVRL pt is an adult in their 50s-60s. They usually present with complaints of decreased vision and/or floaters. Importantly, many will also manifest evidence of CNS involvement





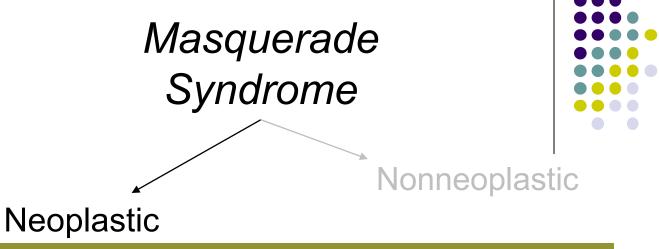
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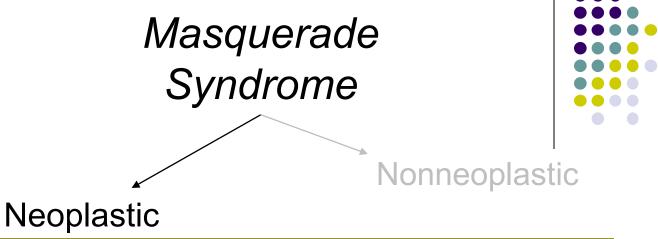
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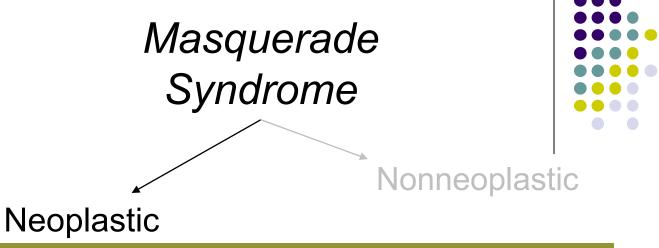


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Leukemic

Lymphoid

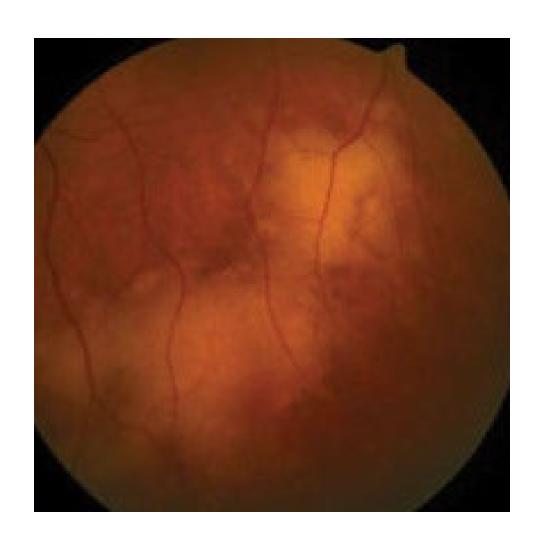
Primary vitreoretinal lymphoma



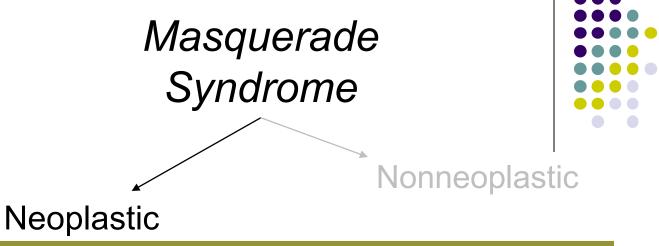
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PVRL: Typical white-yellow subretinal infiltrates



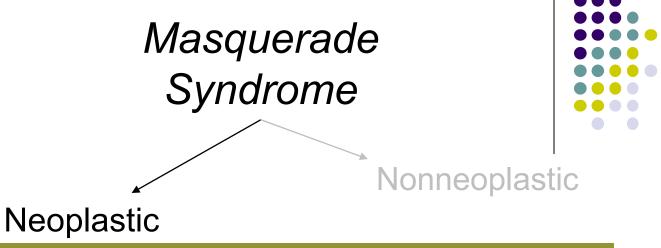
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PVRL is diagnosed by finding three words on vitreous biopsy.

Leukemic

Lymphoid

Primary vitreoretinal lymphoma



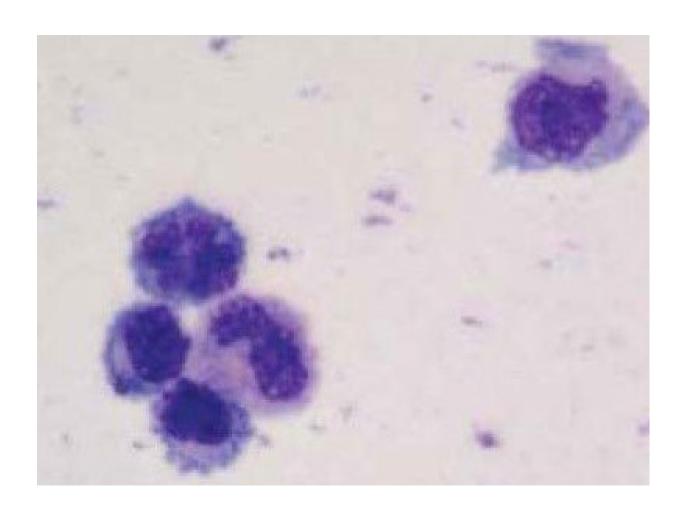
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Leukemic

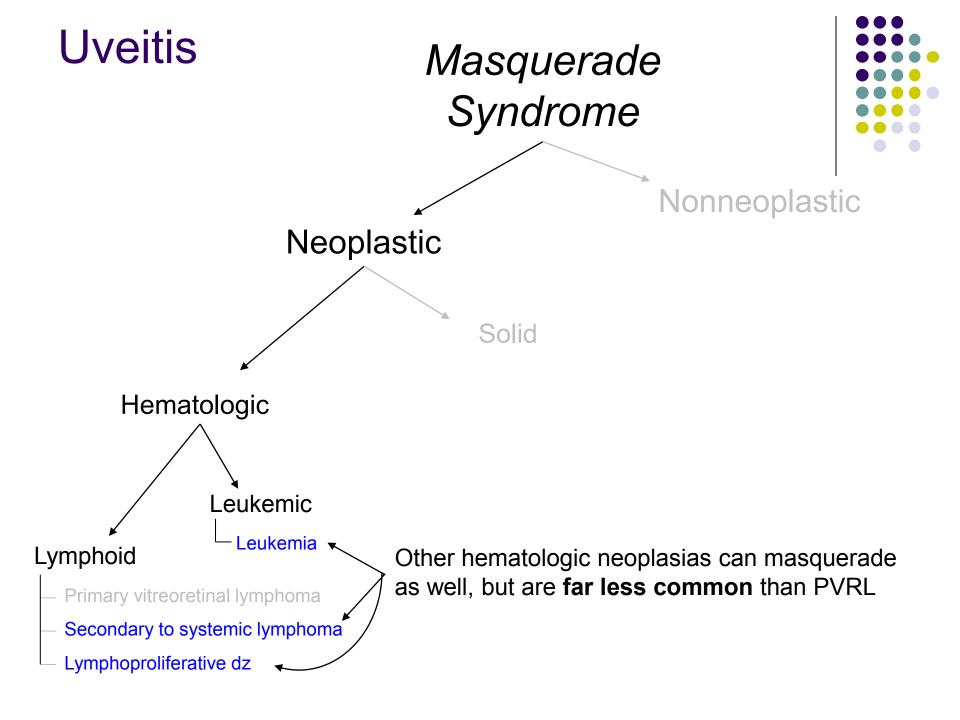
Lymphoid

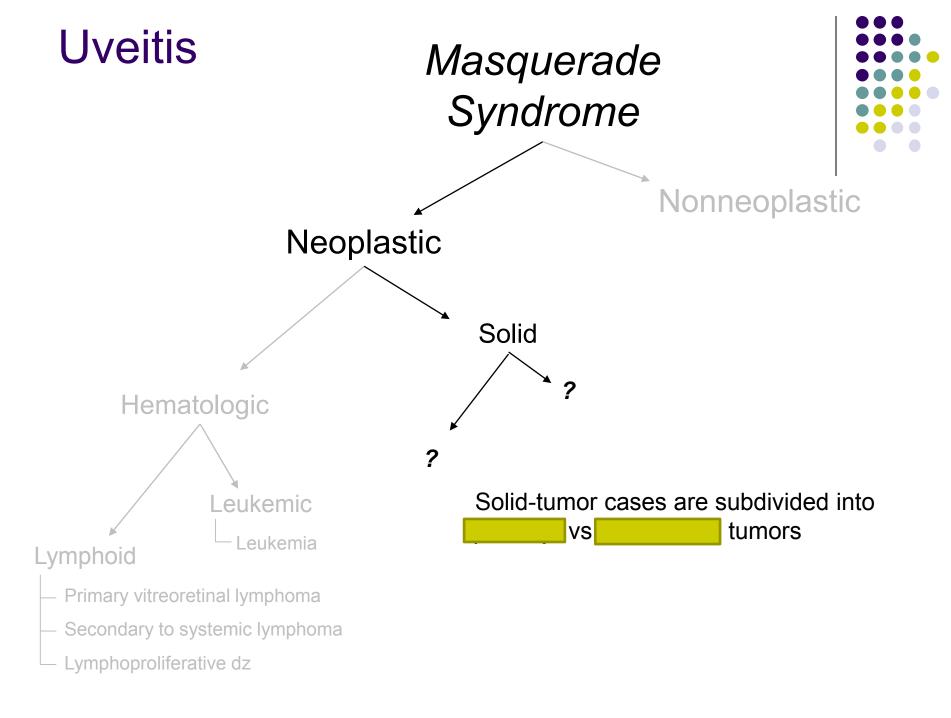
Primary vitreoretinal lymphoma

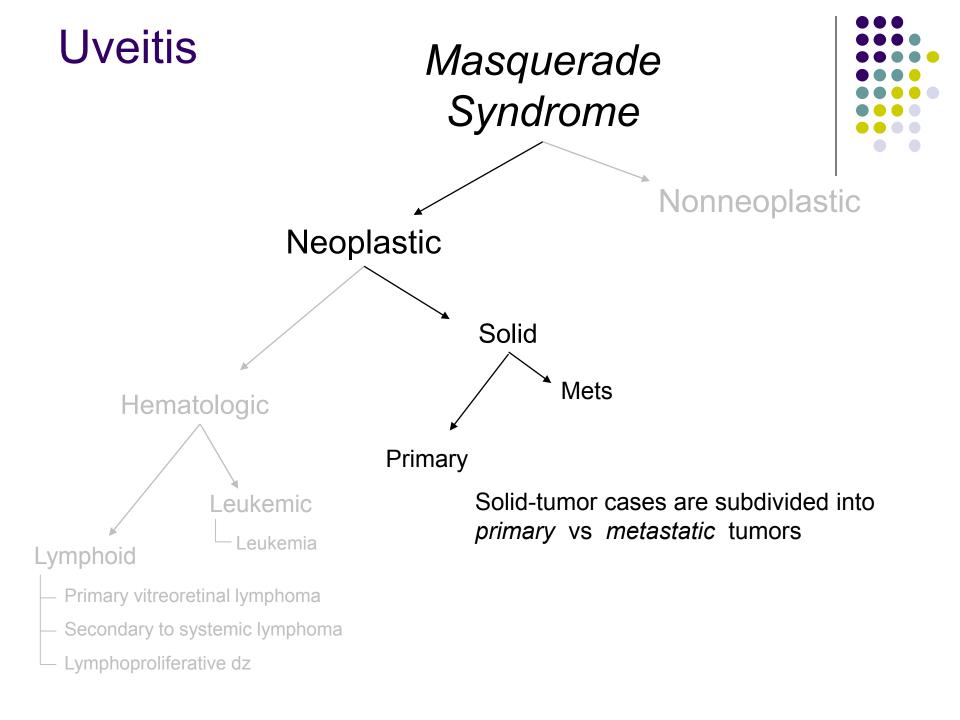


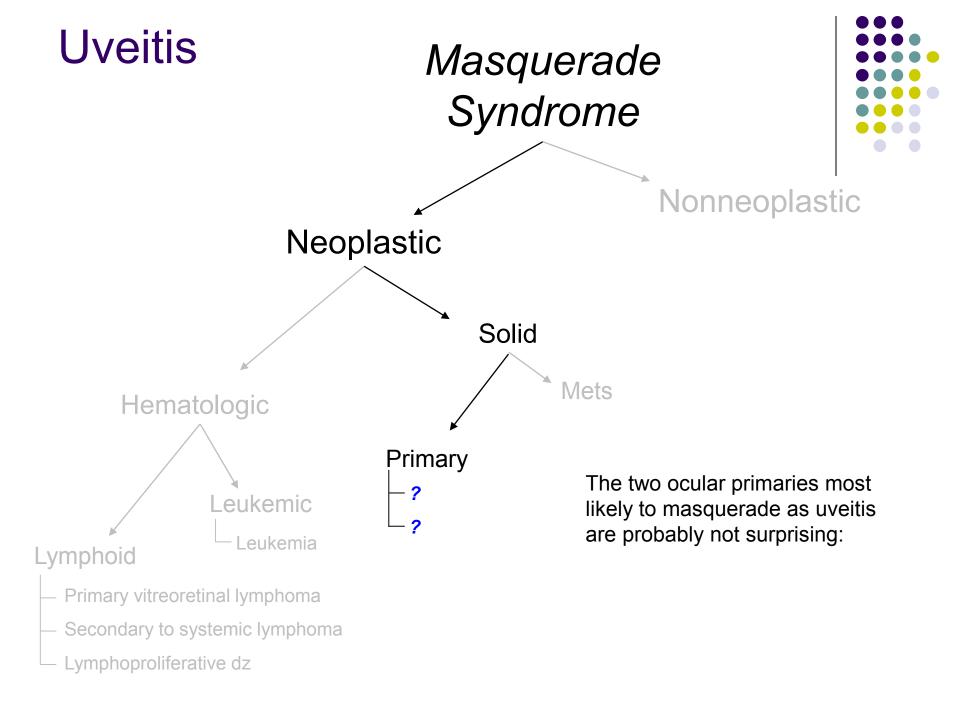


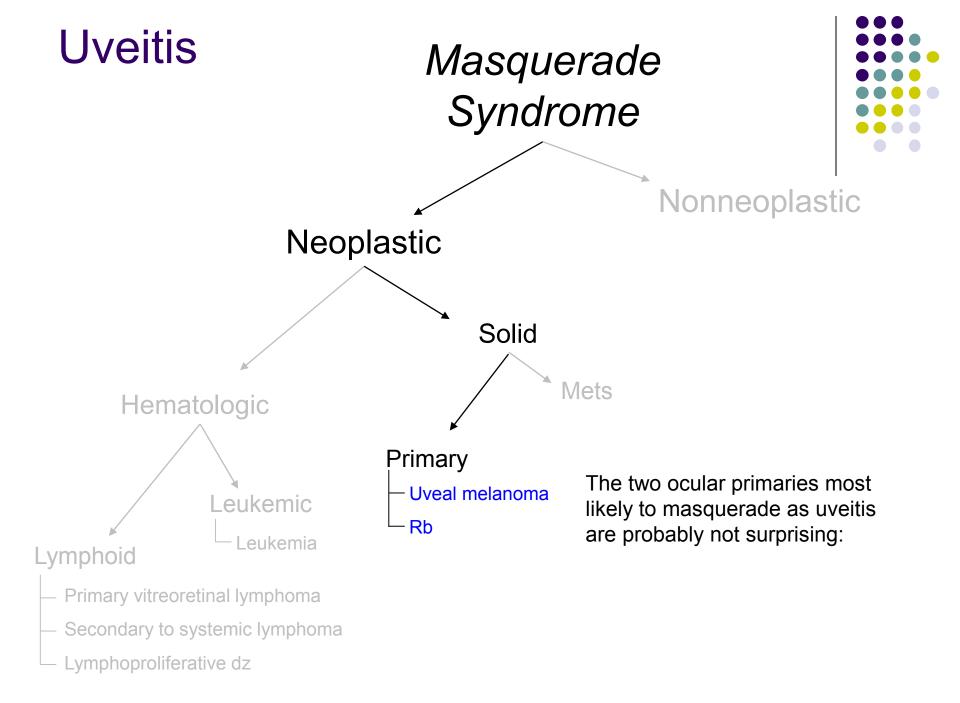
Typical cytology of PVRL cells from the vitreous showing several atypical lymphoid cells with basophilic cytoplasm and large prominent irregular nuclei

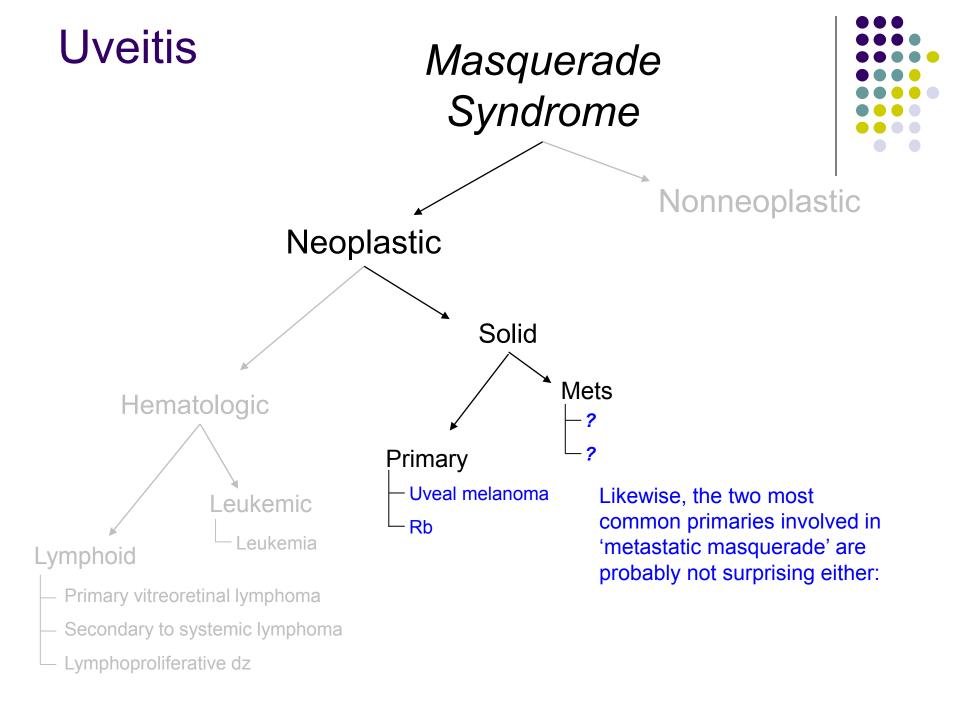


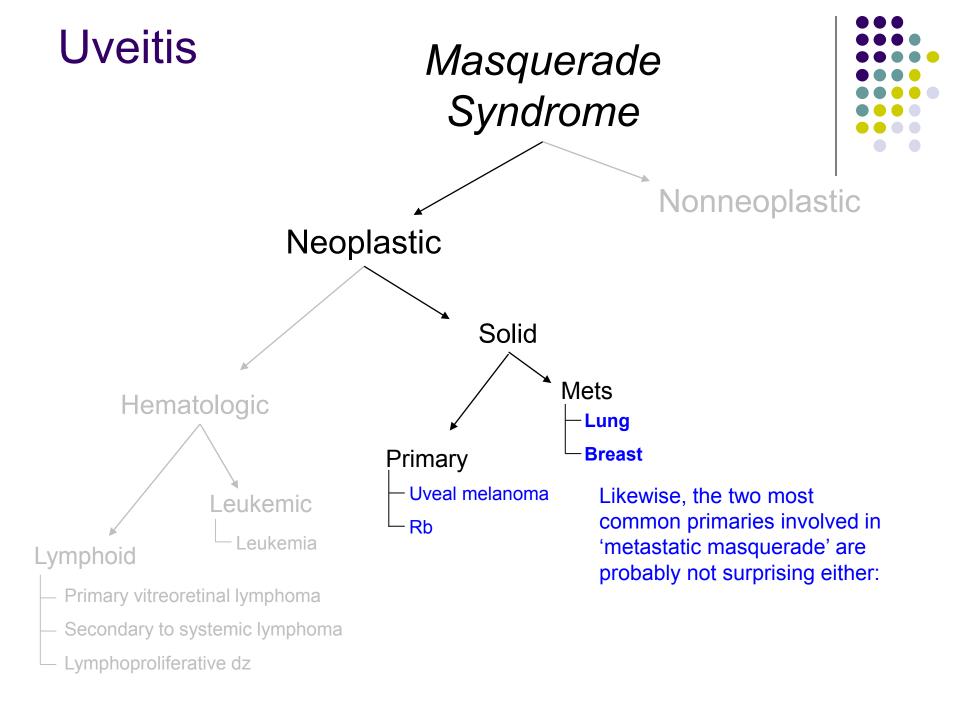


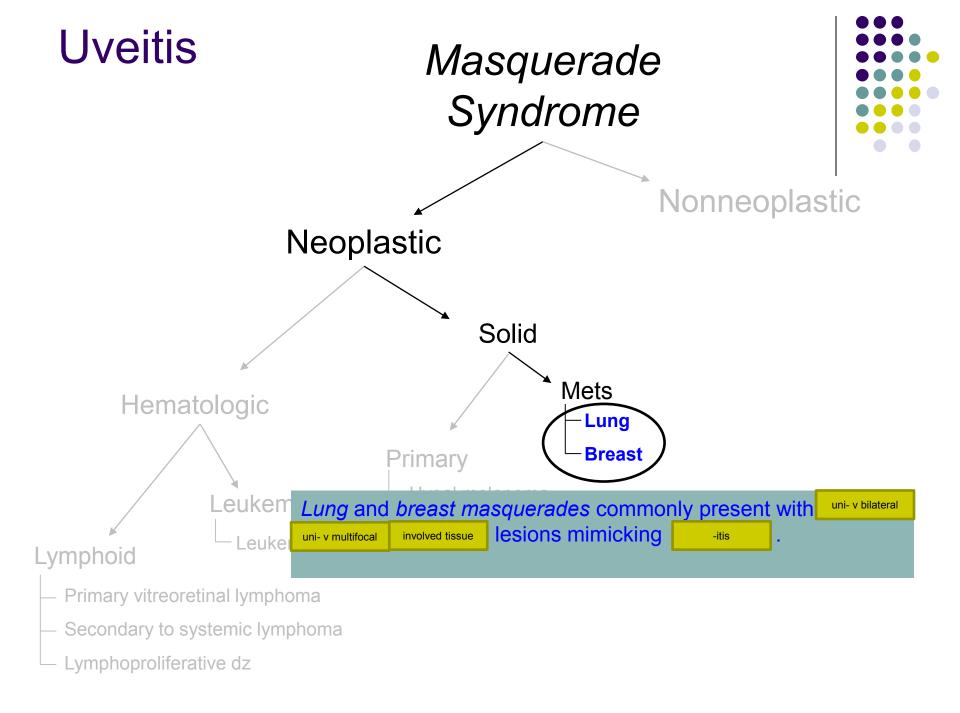


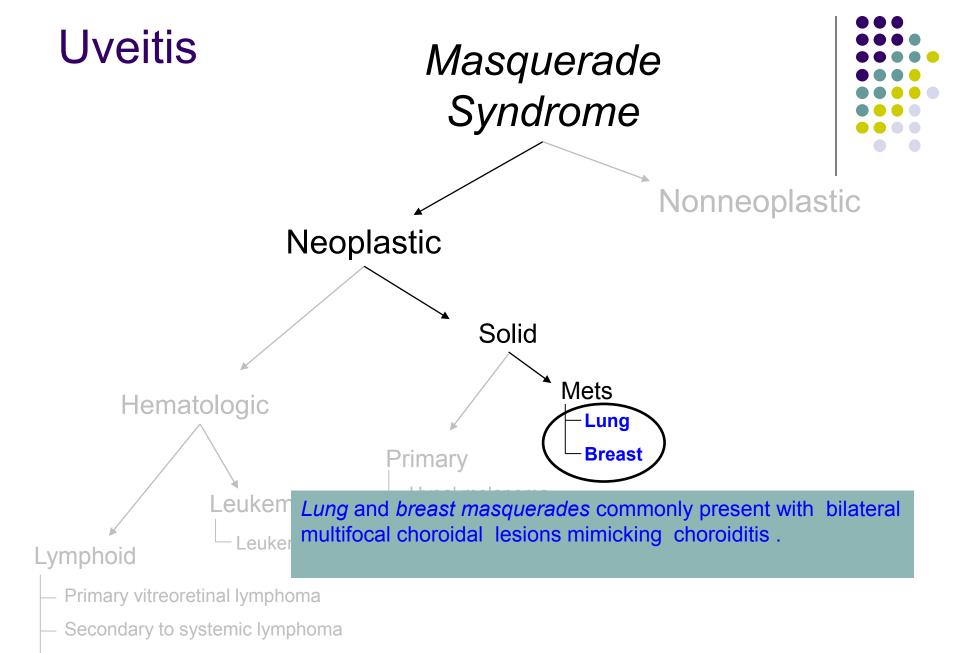








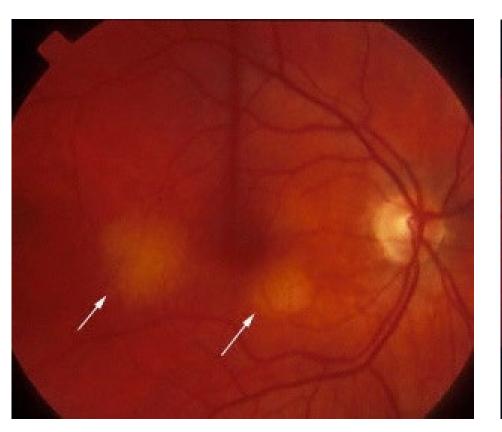


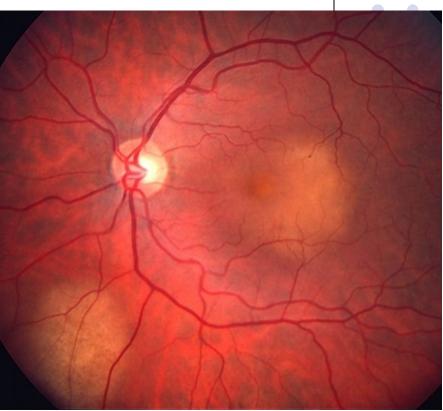


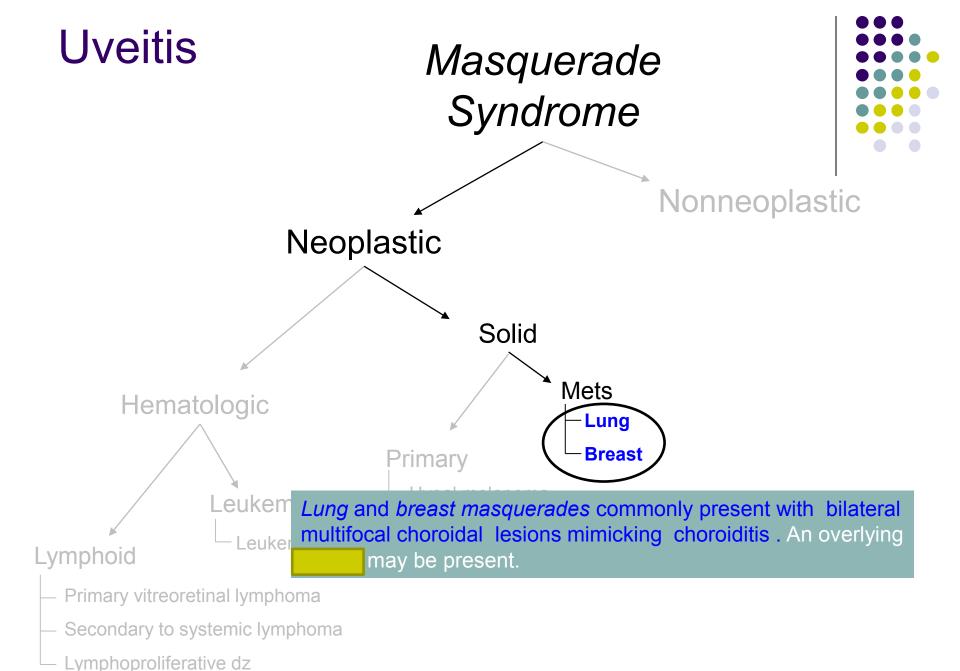
Lymphoproliferative dz

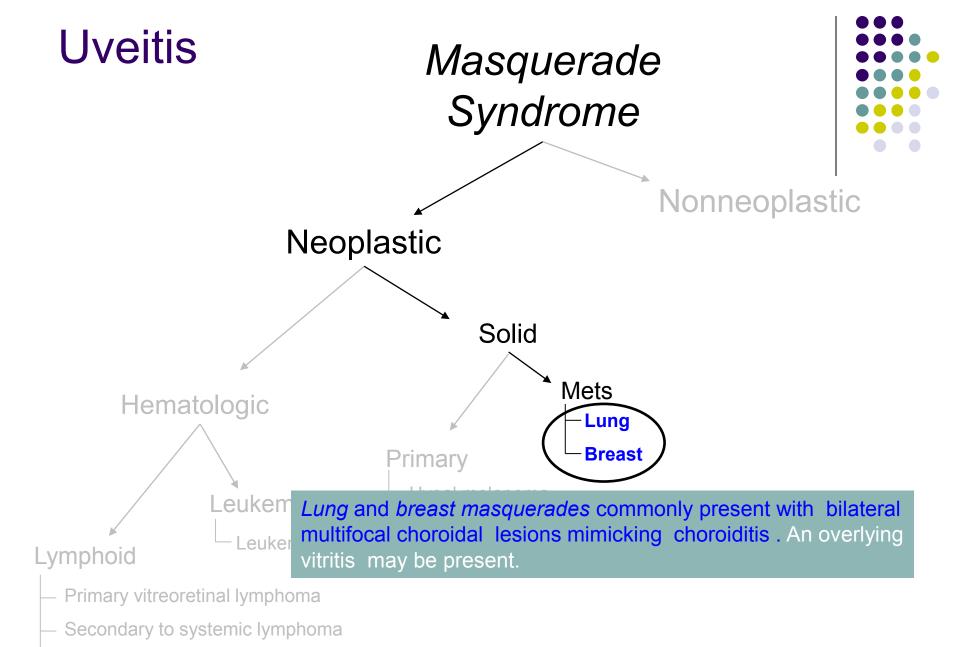




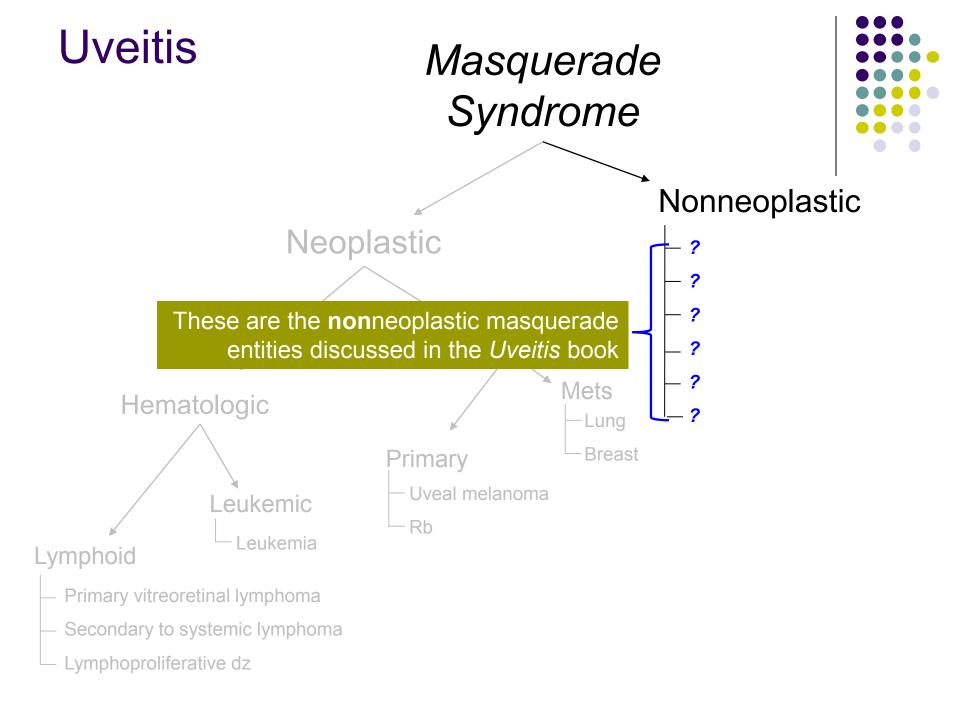


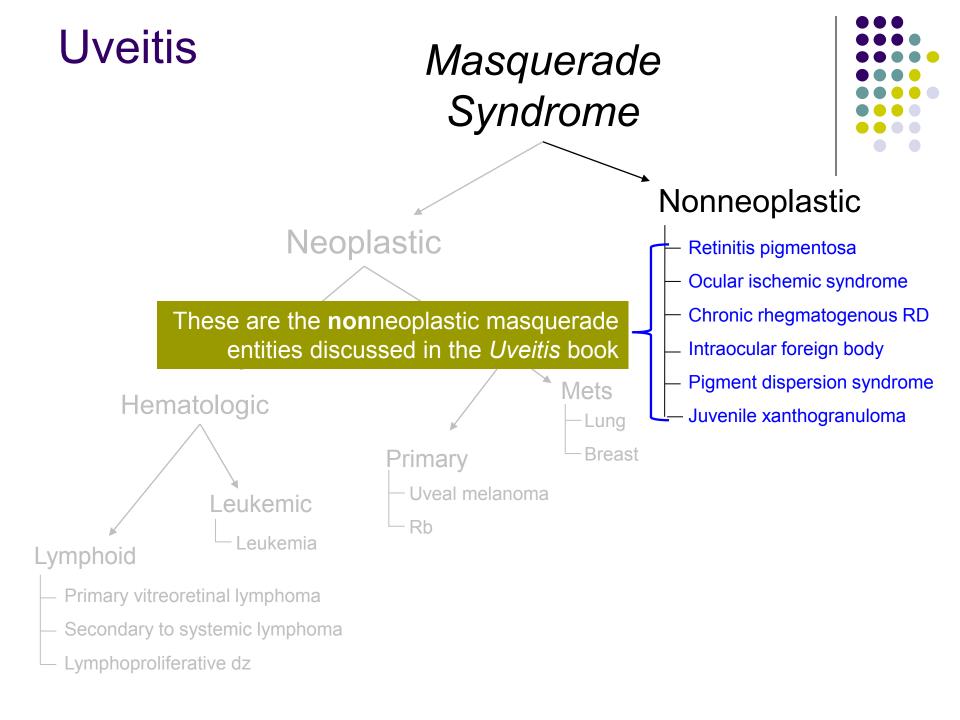






Lymphoproliferative dz





## Masquerade Syndrome



Ocular ischemic syndrome (OIS) is a constellation of ocular abnormalities stemming from chronic of the globe.

Nonneoplastic

OIS

S

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east

Chronic rhegmatogenous RD

Intraocular foreign body

Pigment dispersion syndrome

- Juvenile xanthogranuloma

Leukemic

Leukemia

- Rb

Lymphoid

Primary vitreoretinal lymphoma

Secondary to systemic lymphoma

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

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--?

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- --IOP is...low

--? --?

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.eukemic

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Rb

- --AC...cell and flare
- --IOP is...low
- --Neovascularization of the...iris and/or angle

.eukemic

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Secondary to systemic lymphoma

## 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
- --IOP is...low
- --Neovascularization of the...iris and/or angle
- -- Ipsilateral cataract is...

Leukemic

Leukemia

Rb

Lymphoid

Primary vitreoretinal lymphoma

Secondary to systemic lymphoma

Lymphoproliferative dz

#### Nonneoplastic

OIS

S

ıng

east

Chronic rhegmatogenous RD

Intraocular foreign body

Pigment dispersion syndrome

- Juvenile xanthogranuloma

## 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...low
- --Neovascularization of the...iris and/or angle
- -- Ipsilateral cataract is...more advanced

Leukemic

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Rb

Lymphoid

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#### Nonneoplastic

OIS

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#### **Uveitis** Masquerade Syndrome Nonneoplastic Neoplastic RP OIS The hallmark of PDS is the liberation of pigment from the Chronic rhegmatogenous RD \_ IOFB **Pigment dispersion syndrome** Juvenile xanthogranuloma Uveal melanoma eukemic Rb Leukemia Lymphoid Primary vitreoretinal lymphoma Secondary to systemic lymphoma Lymphoproliferative dz

# Masquerade Syndrome



Neoplastic

The hallmark of PDS is the liberation of pigment from the posterior aspect of the iris.

RP

OIS

Chronic rhegmatogenous RD

— IOFB

**Pigment dispersion syndrome** 

Juvenile xanthogranuloma

eukemic

Leukemia

Uveal melanoma

Rb

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# Masquerade Syndrome



Neoplastic

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.

- RP

- OIS

Chronic rhegmatogenous RD

\_ IOFB

**Pigment dispersion syndrome** 

Juvenile xanthogranuloma

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eukemic

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TITTICET y

Uveal melanoma

Rb

# Masquerade Syndrome

# Nonneoplastic

- RP

- OIS

Chronic rhegmatogenous RD

\_ IOFB

- Pigment dispersion syndrome

Juvenile xanthogranuloma

### Neoplastic

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

Leukemic
Leukemia

with a orientation.

Uveal melanoma

Rb

#### Lymphoid

Primary vitreoretinal lymphoma

Secondary to systemic lymphoma

# Masquerade Syndrome



### Neoplastic

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.

Uveal melanoma
Rb

#### Lymphoid

Primary vitreoretinal lymphoma

Secondary to systemic lymphoma

eukemic

\_eukemia

Lymphoproliferative dz

#### Nonneoplastic

RP

- OIS

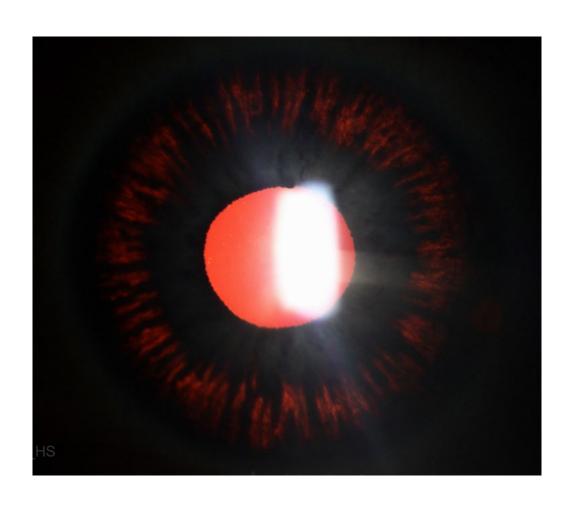
Chronic rhegmatogenous RD

– IOFB

**Pigment dispersion syndrome** 

Juvenile xanthogranuloma





PDS: Radial transillumination defects