Sarcoid: Overview

In broadest terms, how would you describe sarcoid?

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histology (two words)

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What is the classic CXR finding? Bilateral hilar adenopathy



Sarcoid: Bilateral hilar adenopathy

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parotitis and facial palsy. What is the eponymous name for this condition? Heerfordt syndrome

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A lot, actually. Recall that, upon exiting the skull, the facial nerve dives into the parotid gland before branching. Thus, it is not surprising that inflammation of the parotid gland could produce facial nerve palsy.

parotitis and facial palsy. What is the eponymous name for this condition? Heerfordt syndrome

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What percentage of sarcoid pts have ocular involvement? About 50

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Of pts with ocular sarcoid, how many will present with an anterior uveitis?

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Most with either a posterior uveitis or panuveitis

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For what population is the incidence of ocular involvement far higher--around 90%?

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Which other organ systems can be involved? Pretty much all of them, especially the CNS, biliary, skin, heart, lymphatics, joints. And eyes of course.

What percentage of sarcoid pts have ocular involvement? About 50

Who is the typical sarcoid pt in the US? An African-American

During what age-range does dz onset typically occur?

### Sarcoid: Overview

1) The uveitis is profiled

2) The profiled case is meshed

3) A differential diagnosis list is generated

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5) Treatment appropriate for the etiology is initiated

In broadest terms, how would you describe sarcoid?
It is a multi-organ disease characterized by the presence of noncaseating granulomas

What is the cause?
As of this writing, it is unknown

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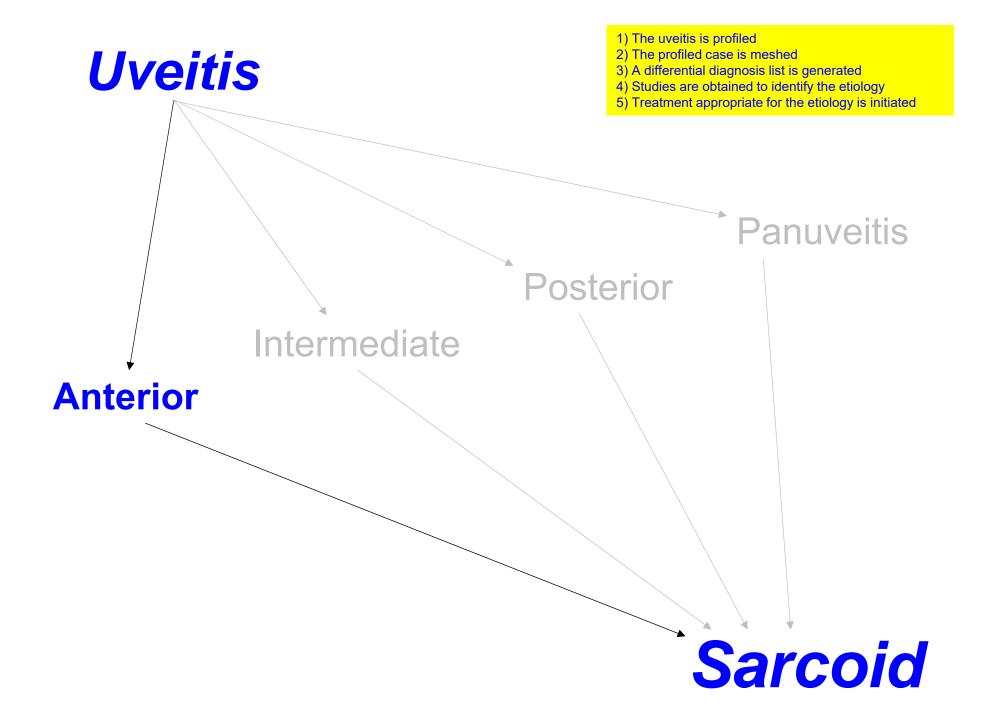
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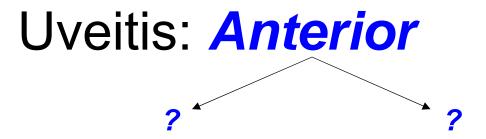
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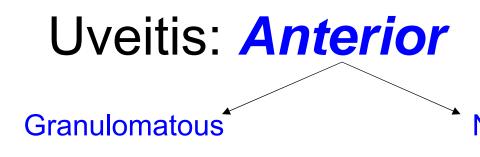
Is there a gender predilection for sarcoid?
Female (but only slightly, so don't consider this when meshing)





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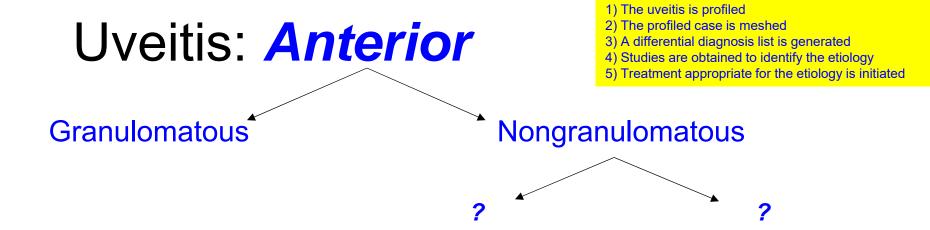
Per the Uveitis book, what are the two basic forms of anterior uveitis?



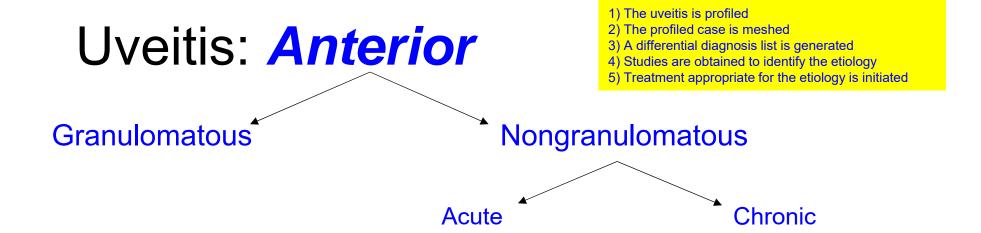
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Nongranulomatous

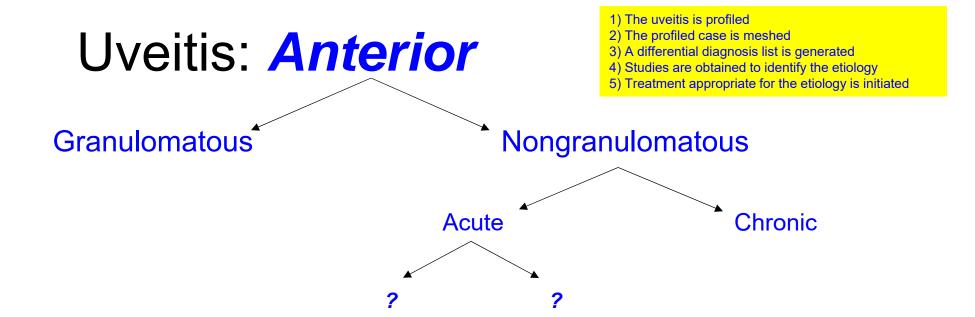
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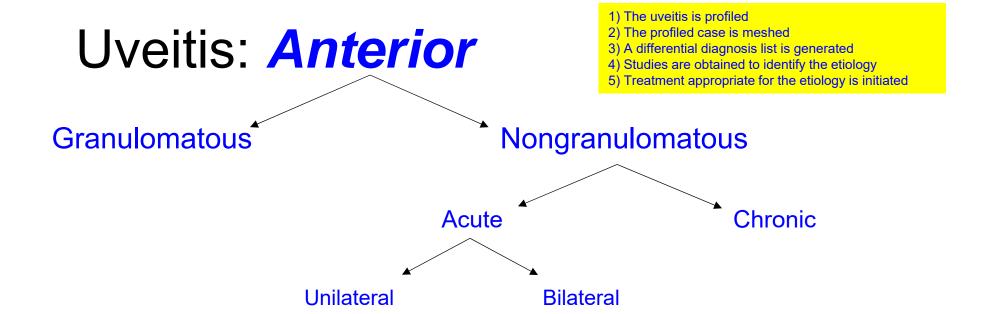
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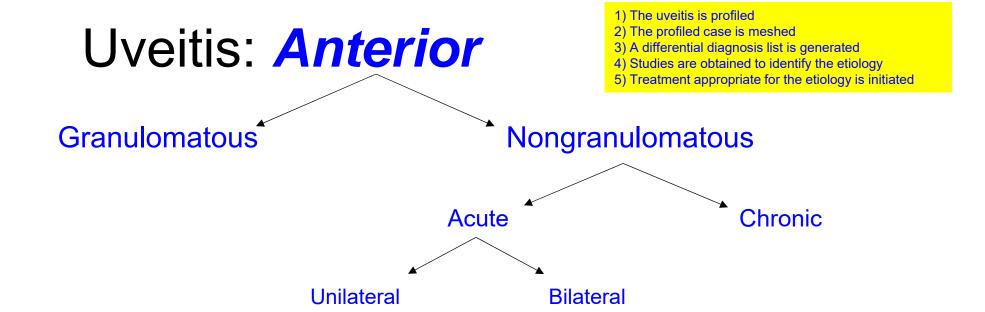
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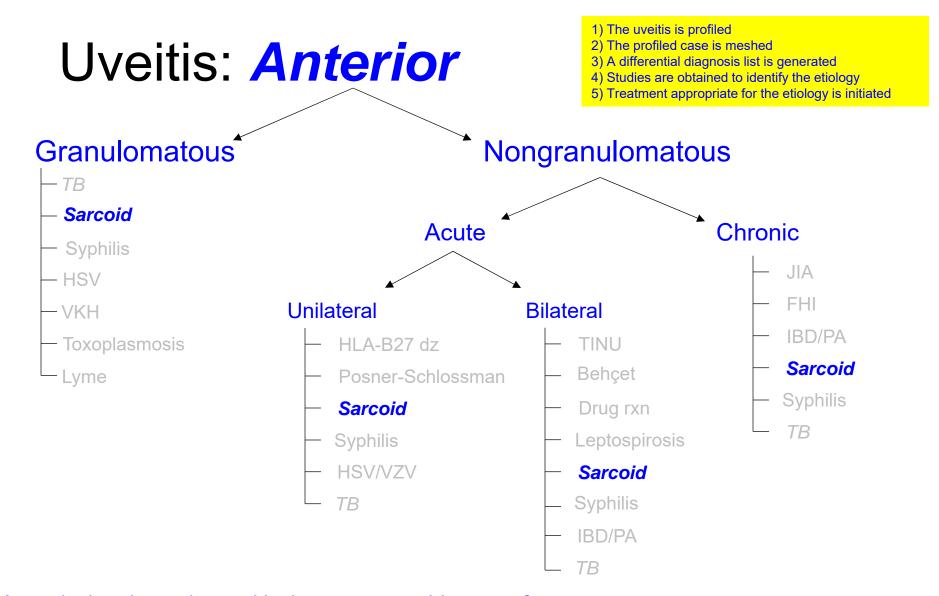
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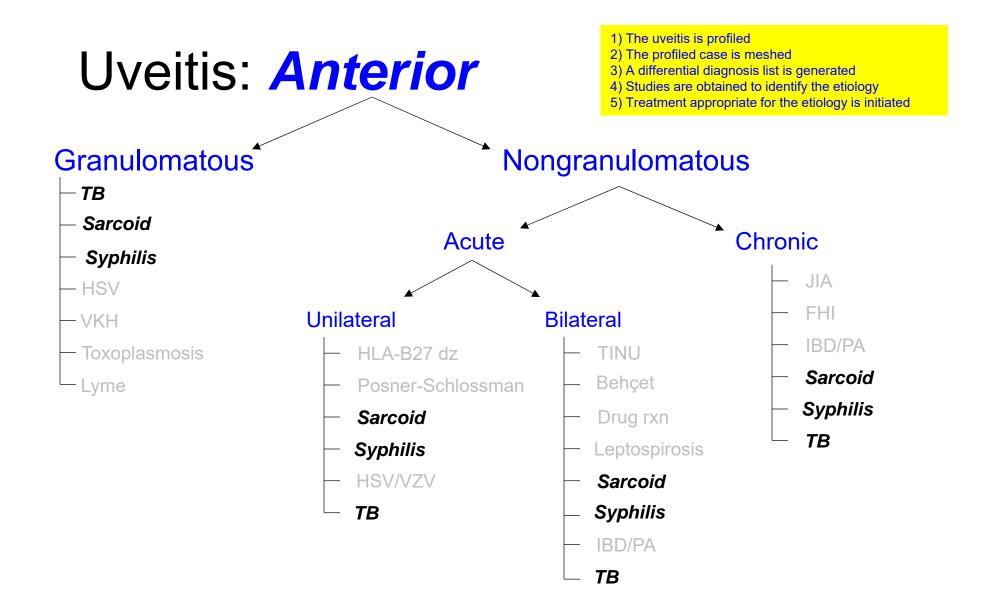
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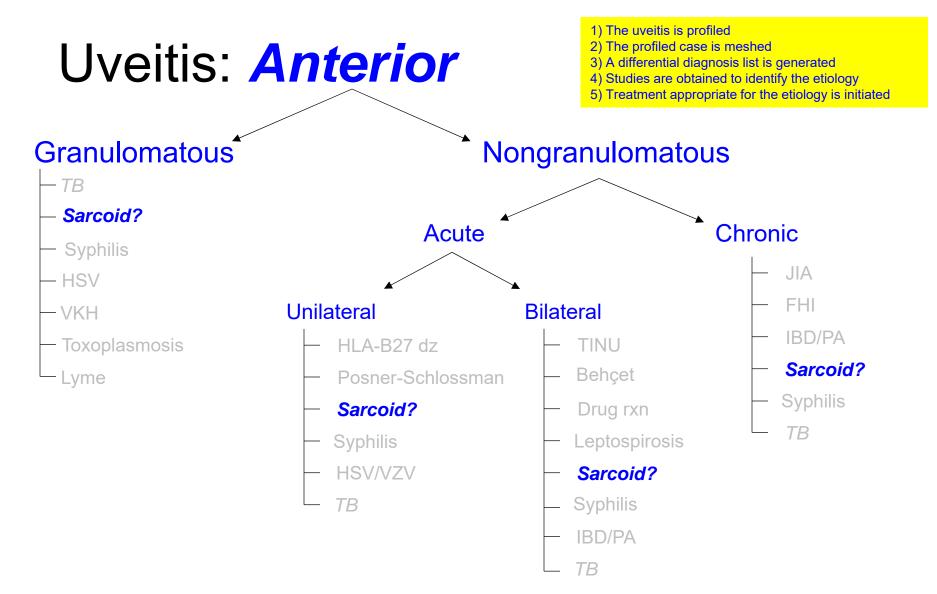
As an isolated anterior uveitis, how can sarcoid present?



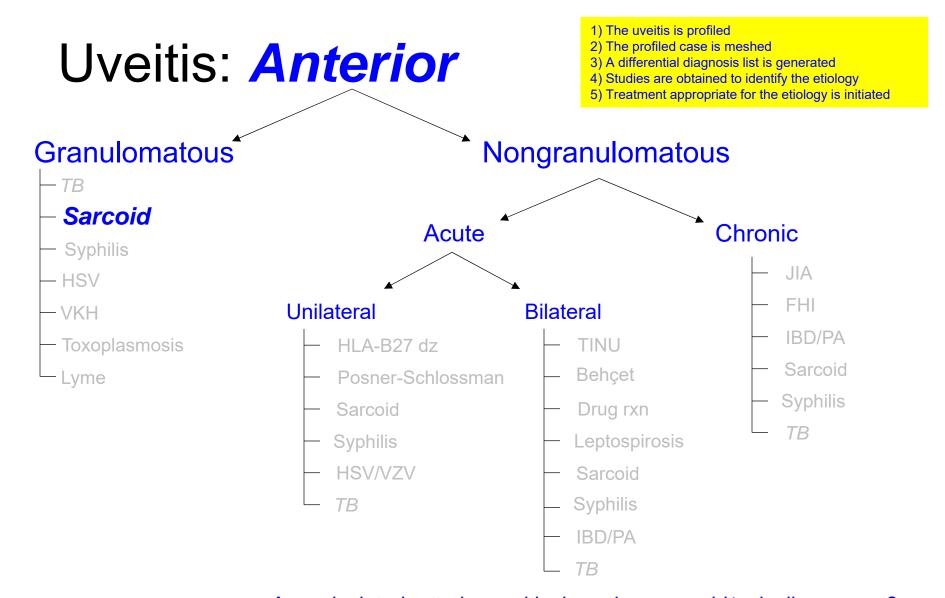
As an isolated anterior uveitis, how <u>can</u> sarcoid present? **As anything!** 



Remember: Sarcoid, syphilis and TB can manifest as any sort of uveitis!



As an isolated anterior uveitis, how does sarcoid typically present?



As an isolated anterior uveitis, how does sarcoid typically present?

As a granulomatous uveitis

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### **Granulomatous**

Nongranulomatous

- Sarcoid

Syphilis

- HSV

– VKH

Toxoplasmosis

Lyme

Histologically speaking, what makes an inflammatory condition 'granulomatous'?

The presence of epithelioid and glant cells

*In clinical ophtho-speak, to what does the term* granulomatous *refer?*To a particular slit-lamp appearance of KP in uveitis

*What do granulomatous KP look like?* They are large, grayish, and look 'greasy

A pt has granulomatous KP. If a KP was scraped and examined microscopically, would it be chock full of epithelioid and/or giant cells?

Not necessarily. While significant overlap exists between the two, it is **not** the

− IBD/PA − *TB* 

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Throughout the *anterior uveitis* slides, the term *granulomatous* refers to the slit-lamp appearance of the KP, not to the histology of the condition

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What three structures manifest important clinical signs in a typical case of sarcoid anterior uveitis?

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-- Cornea (one finding): Large granulomatous KP

What two-word term is often used to describe the appearance of KP in sarcoid?

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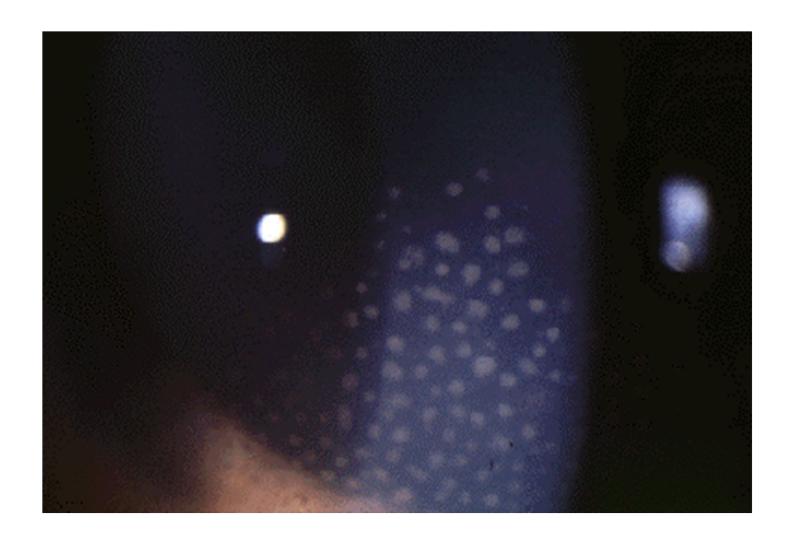
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Mutton-fat KP

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ArIt's triangle



KP in Arlt's triangle

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There are three eponymous iris nodules in sarcoid. What are they...

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- --?
- --?

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- --Koeppe nodules
- --Busacca nodules
- --Berlin nodules

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- --Busacca nodules: ?
- --Berlin nodules: ?

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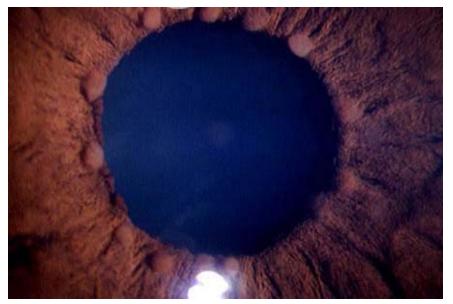
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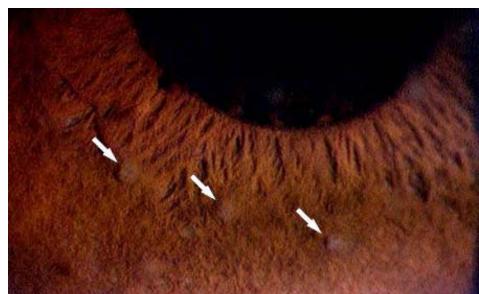
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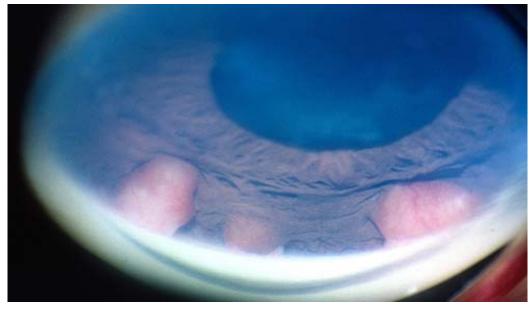
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- --Berlin nodules: The iris angle





Koeppe Busacca



Berlin

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Nongranulomatous

TB

Sarcoid

Syphilis

HSV

What three structures manifest important clinical signs in a typical case of sarcoid anterior uveitis? The cornea, iris and (to a lesser extent) anterior vitreous

What are the findings for each?

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There are three eponymous iris nodules in sarcoid. What are they...and where on the iris are they found?

- --Koeppe nodules: Pupillary margin
- --Busacca nodules: Mid-iris
- --Berlin nodules: The iris angle

Are these pathognomonic for sarcoid?

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They can be posterior, peripheral-anterior (ie, PAS), or both

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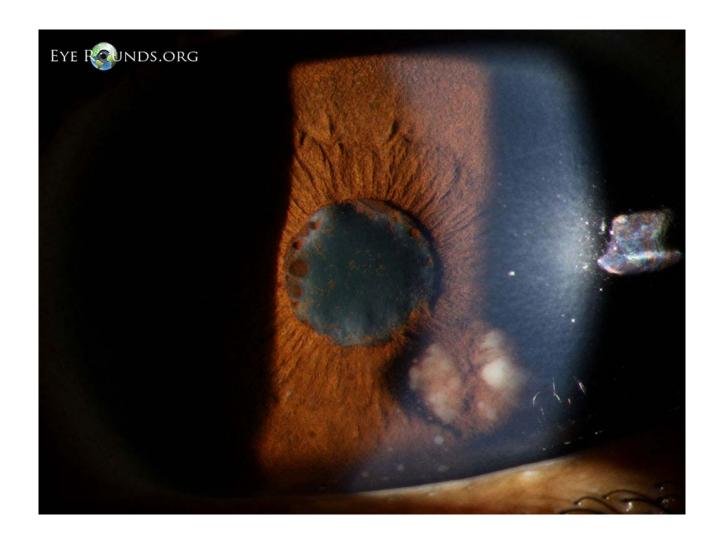
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Posterior synechiae (and the world's largest Busacca nodule) in sarcoid

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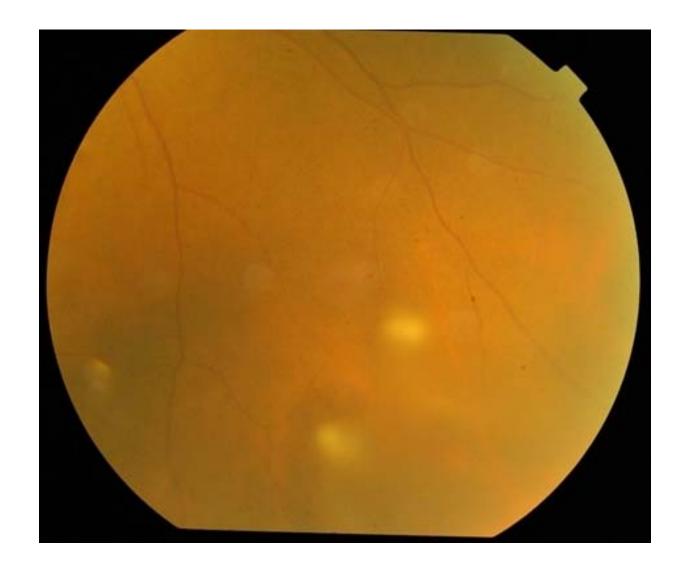
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Clumps of inflammatory cells floating in the vitreous are known as what? 'Snowballs'



Snowballs

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### Granulomatous

Nongranulomatous

- *TB*?

Sarcoid

Syphilis?

- HSV?

- VKH?

– Toxoplasmosis?

- Lyme?

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Nongranulomatous

- **TB** 

- Sarcoid

**Syphilis** 

HSV

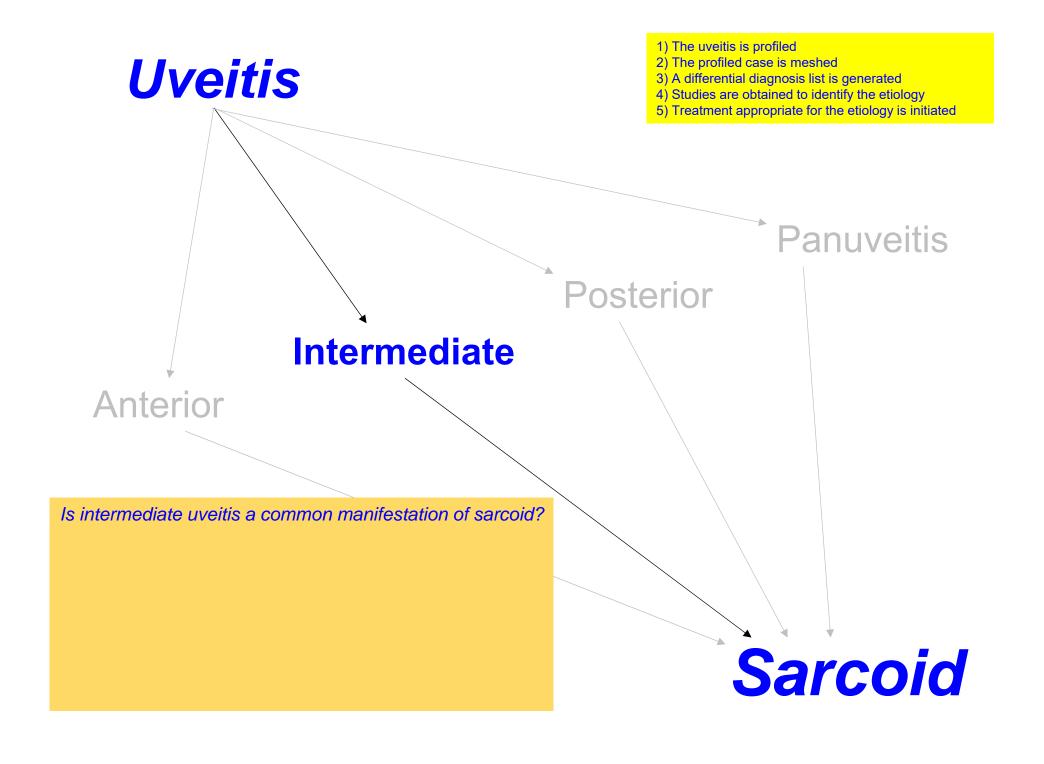
VKH

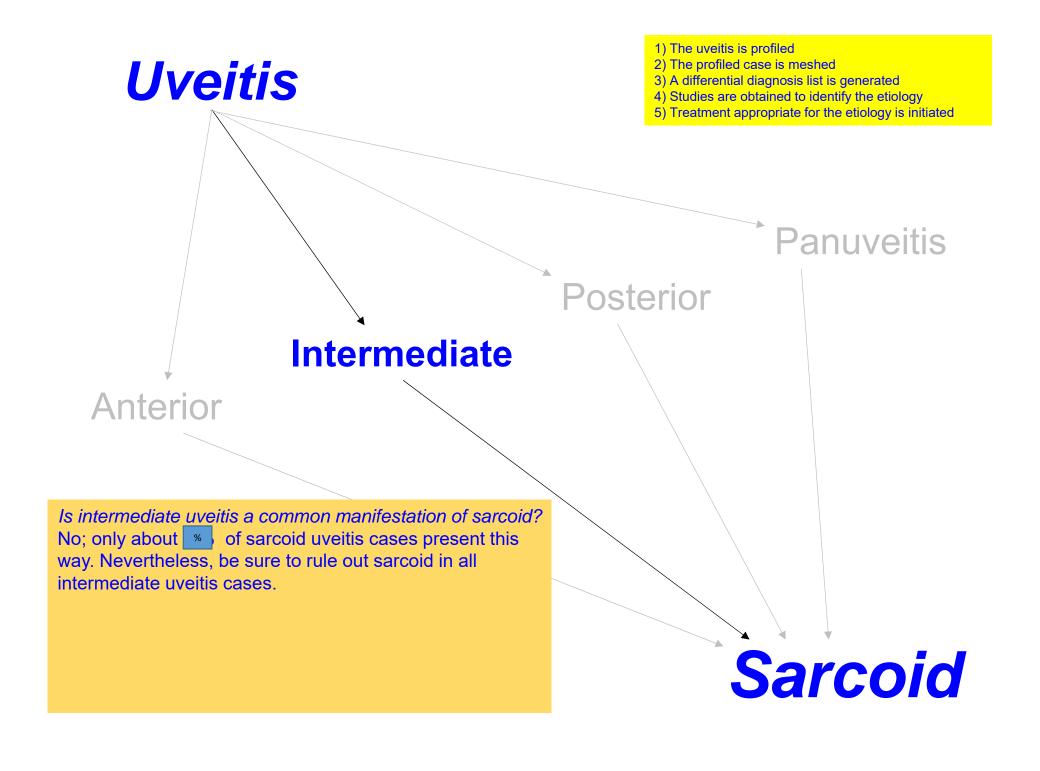
Toxoplasmosis

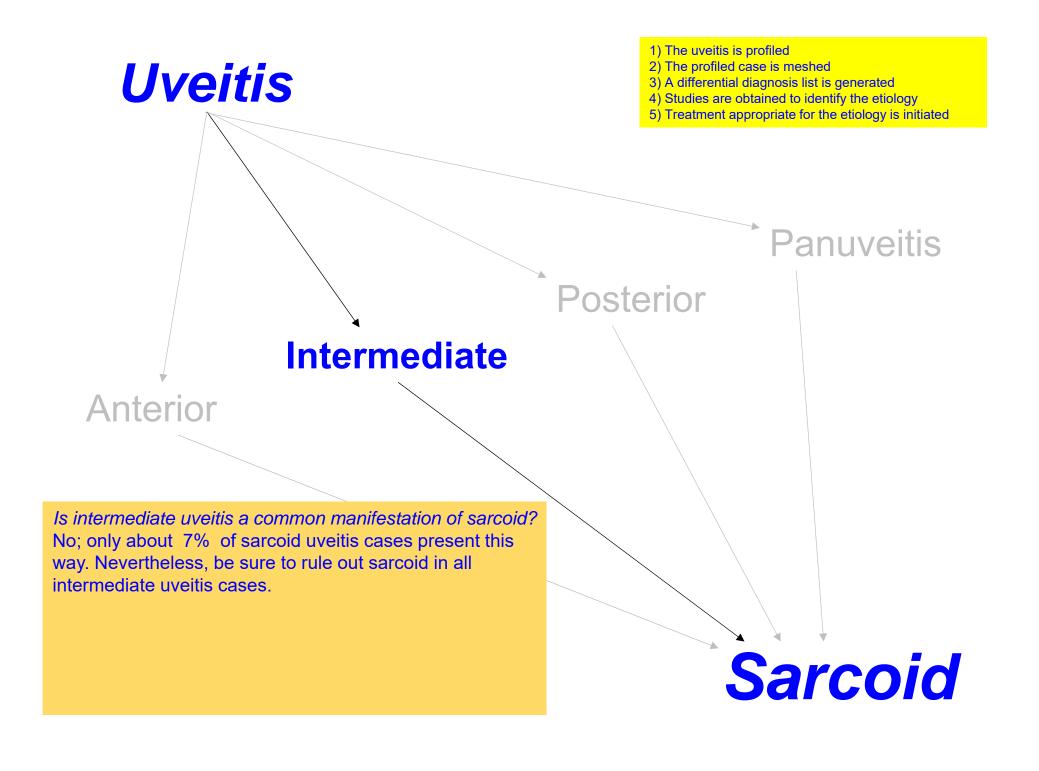
Lyme

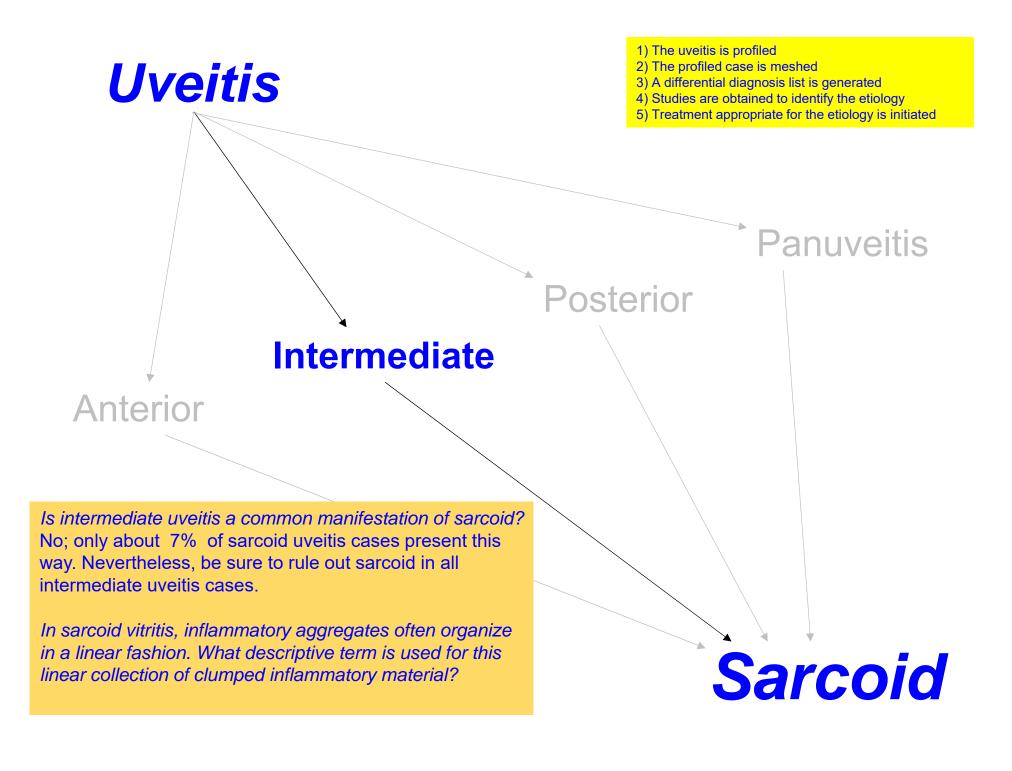
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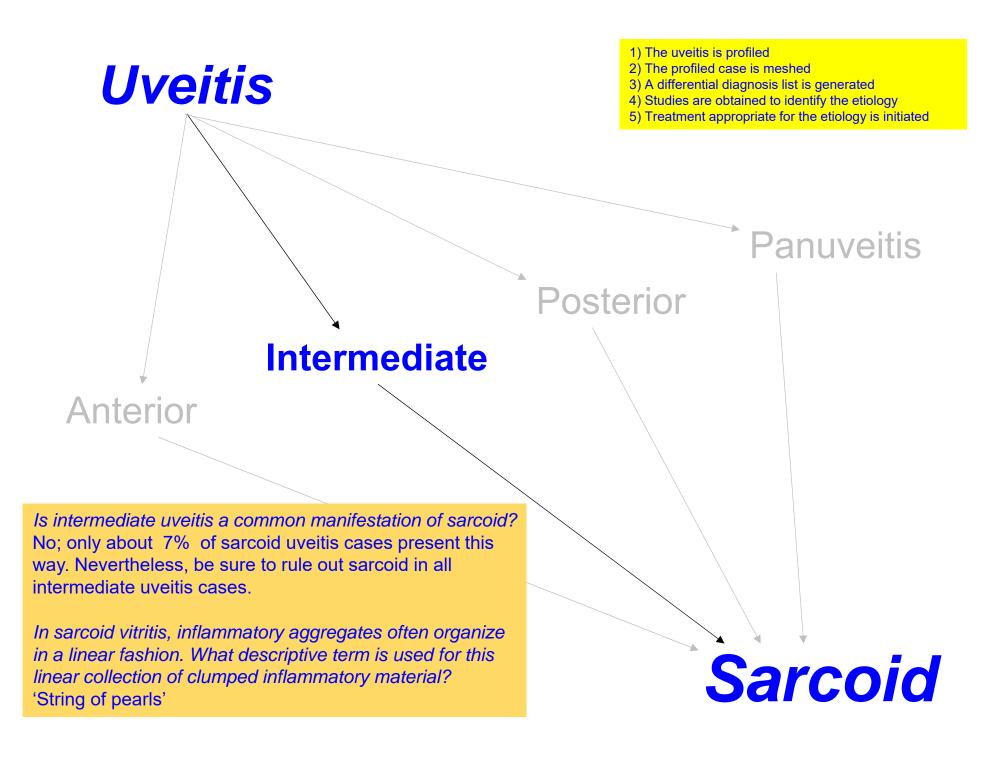
**TB** and **syphilis**. (Remember, TB, sarcoid and syphilis can present in multiple ways!)

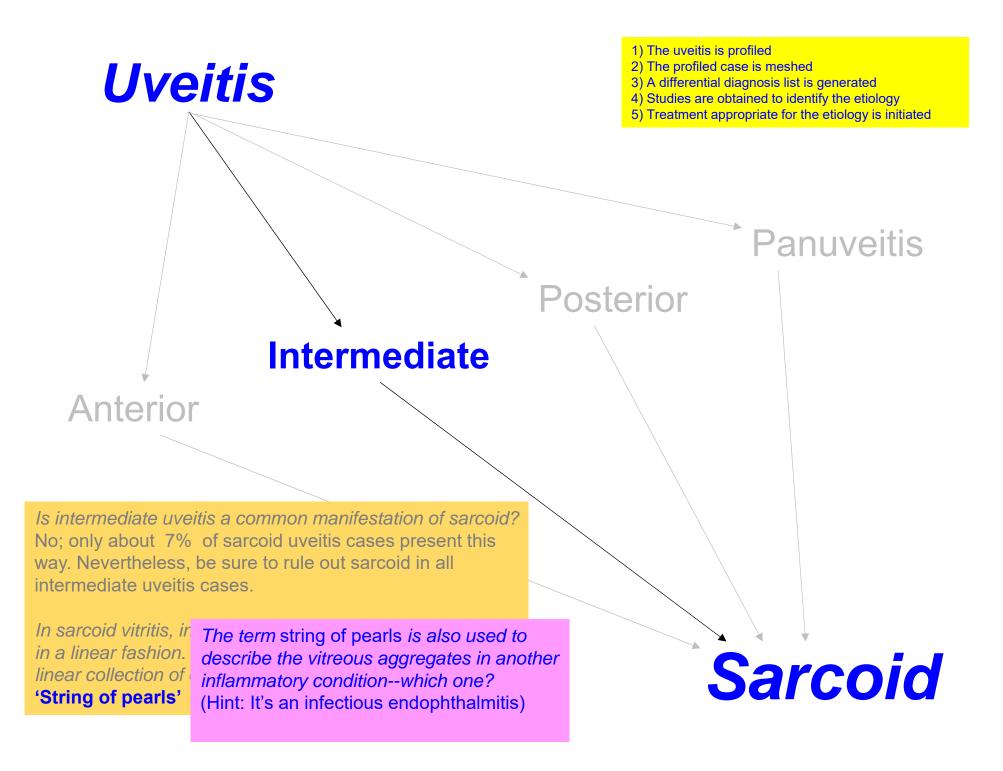


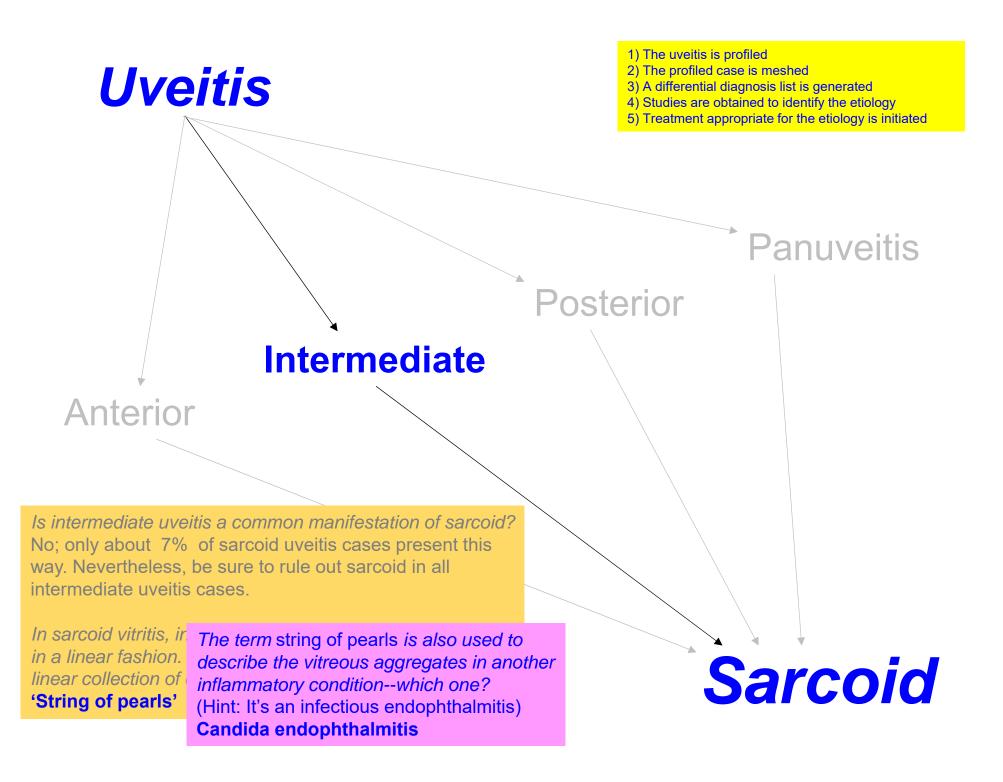


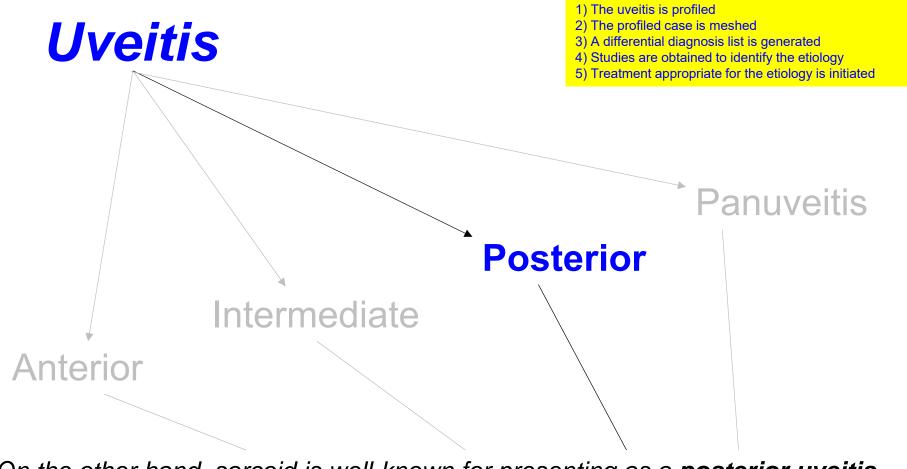






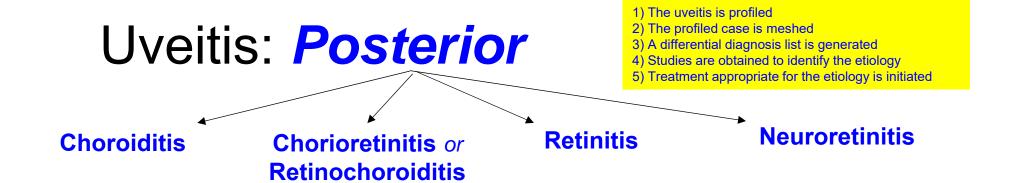




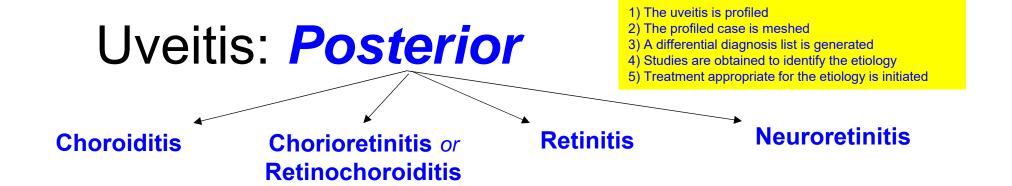


On the other hand, sarcoid is well-known for presenting as a posterior uveitis.

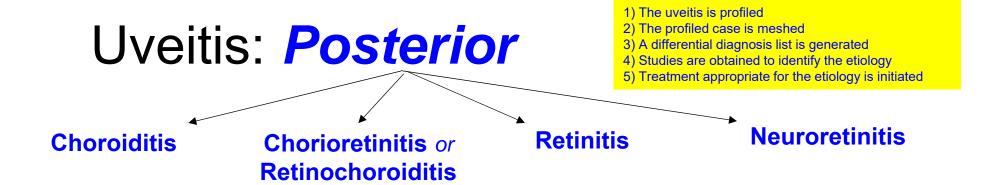




What percentage of sarcoid uveitis pts have posterior manifestations?



What percentage of sarcoid uveitis pts have posterior manifestations? About 20

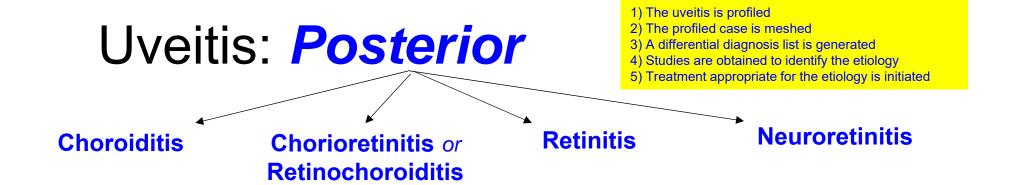


What percentage of sarcoid uveitis pts have posterior manifestations? About 20

What are the two most common posterior manifestations?

-

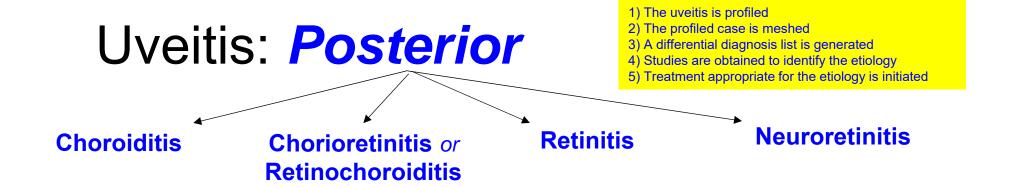
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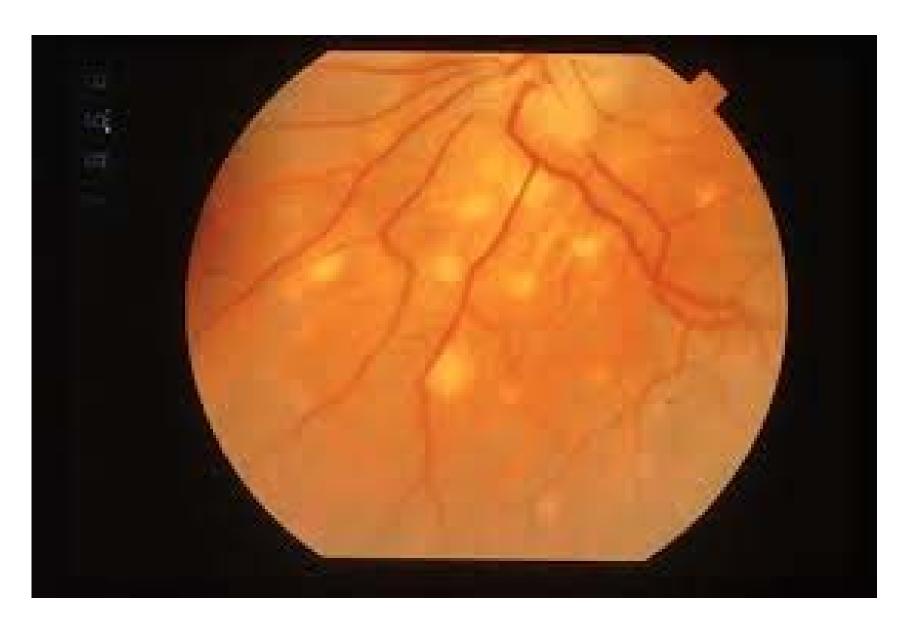
What are the two most common posterior manifestations?

- --Granulomas of the and/or
- --Retinal



What are the two most common posterior manifestations?

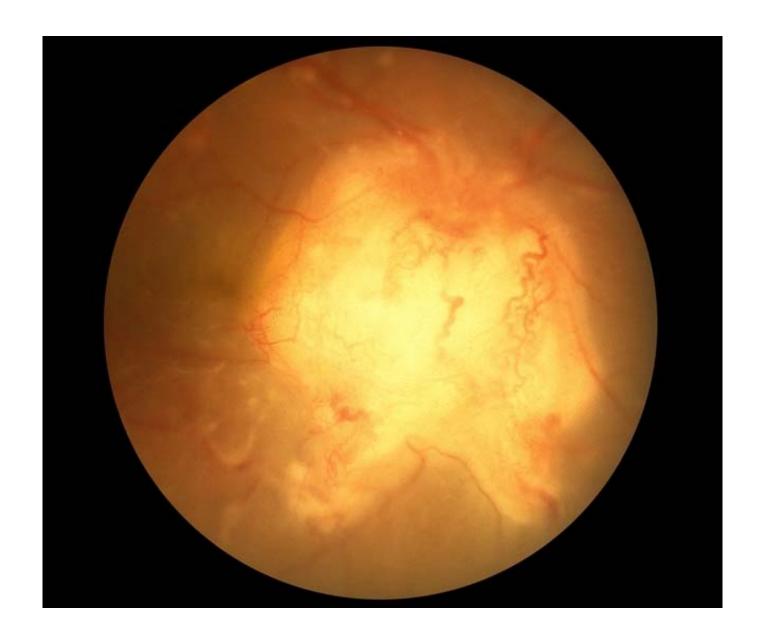
- --Granulomas of the choroid, retina and/or optic nerve
- --Retinal vasculitis



Choroidal granulomas in sarcoid



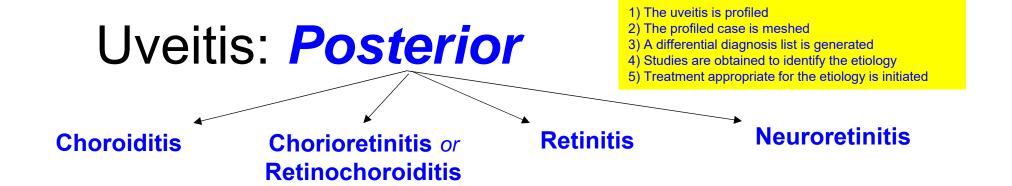
Choroidal granulomas in sarcoid. Note the 'punched out' appearance



ONH granuloma in sarcoid

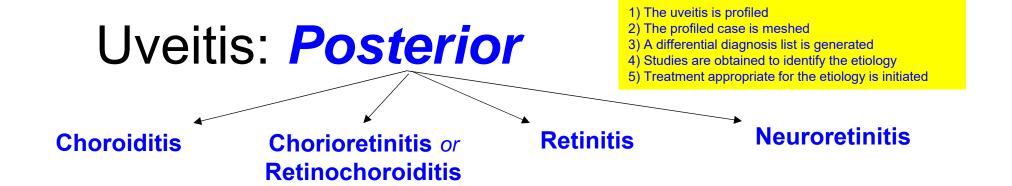


ONH granuloma with neuroretinitis



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--Granulomas of the choroid, retina and/or optic nerve
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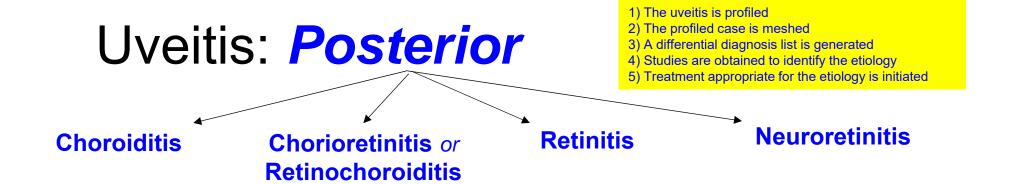
Is the vasculitis primarily an arteritis, a phlebitis, or both?



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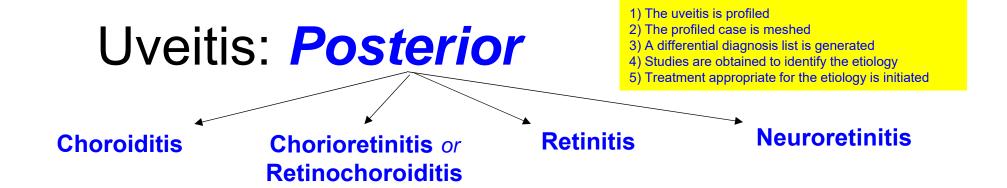
Sarcoid tends to cause a phlebitis (more specifically, a \_\_\_\_\_\_\_)



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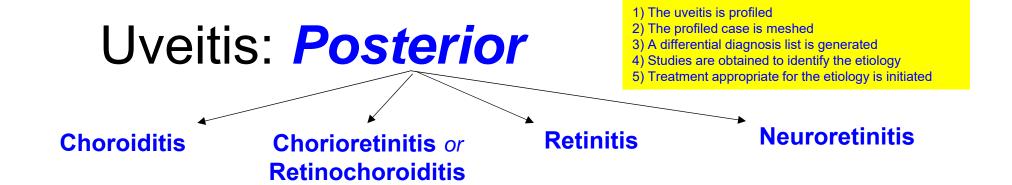


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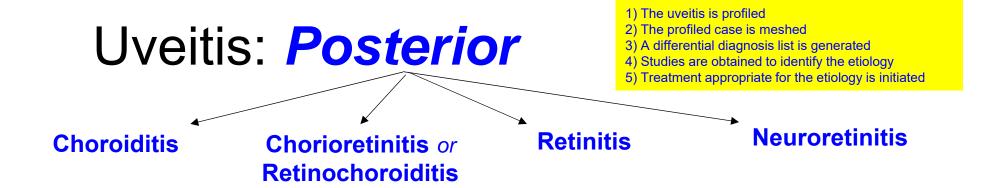
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Small yellowish-white nodular granulomas accumulate along venules in a pattern referred to as three words



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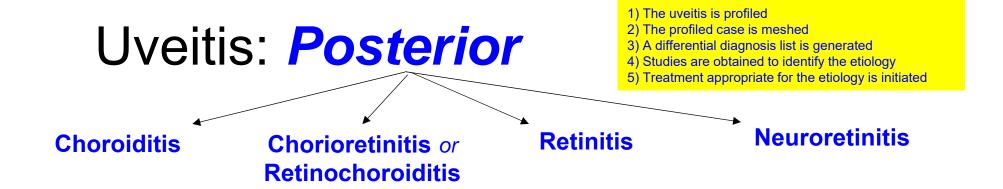
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Sarcoid periphlebitis: 'candle-wax drippings'



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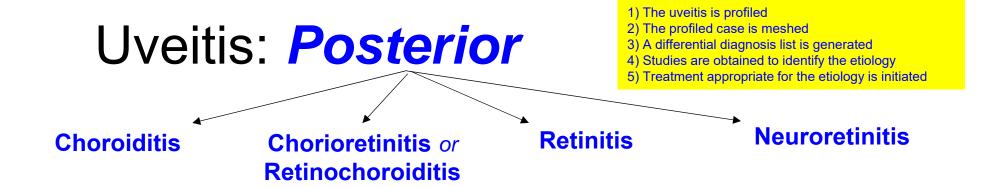
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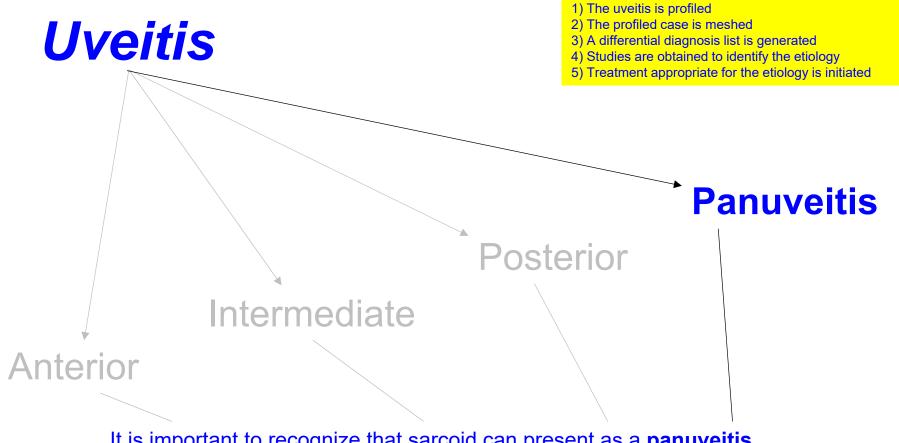
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Taches de bougie



It is important to recognize that sarcoid can present as a **panuveitis**, with primary inflammation occurring in the anterior, intermediate and posterior segments of the eye simultaneously!

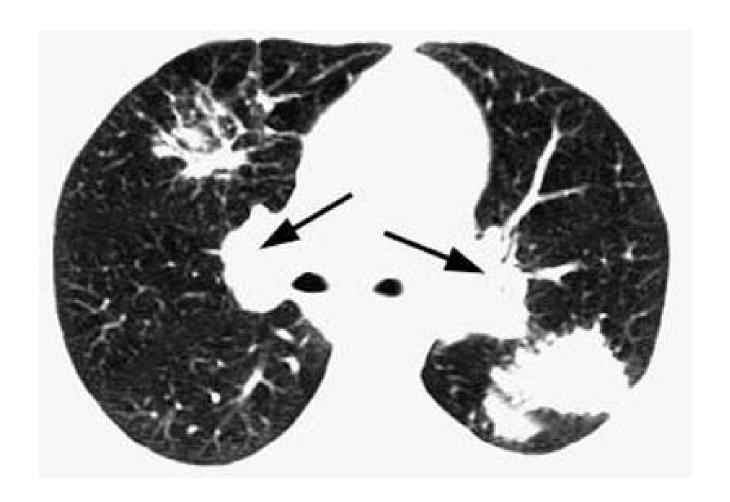




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CT chest in sarcoid: dense focal nodular infiltrates; bilateral hilar lymphadenopathy



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If both CXR and CT fail to reveal evidence of sarcoid, what other imaging test could be considered?



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Whole-body gallium scan (good luck getting **that** approved)



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Thin cut CT of the What does ACE stand for?



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As mentioned previously, CXR is the best screening test for sarcoid. If CXR is read as normal, but clinical suspicion for sarcoidosis remains high, what other imaging modality should be considered?

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Angiotensin converting enzyme

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Briefly, what does the renin-angiotensin system do? Help regulate blood pressure and fluid status

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What role does ACE play in the renin-angiotensin system? It converts angiotensin I to angiotensin II



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With what functional system/unit is this enzyme associated? The 'innate' immune system



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In sarcoidosis, are ACE and lysozyme levels expected to be abnormally high, or low?



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Why are ACE levels elevated in sarcoid?

In sarcoidosis, are **ACE** High





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What two lab tests should be considered? Serum ACE, and lysozyme Why are ACE

In sarcoidosis, ar ACE High

Why are ACE levels elevated in sarcoid?

The epithelioid and giant cells found in the granulomas are believed to secrete it



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No, it is elevated in many systemic conditions

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What common class of HTNive med will artificially lower the ACE level?

ACE inhibitors. Be sure to know whether your pt takes an ACEI before ordering a level!

What is the only way to definitively diagnose sarcoidosis?



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What is the only way to definitively diagnose sarcoidosis? Via biopsy demonstrating the presence of noncaseating granulomas



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What nonocular structures are typically biopsied?

What is the (The lung (transbroncial); enlarged lymph nodes; skin lesions (biopsy)



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What do you have to worry about if you biopsy the main lac gland? That you'll inadvertently bag the ductules, thereby decreasing secretion and causing a dry eye

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If the conj has no obvious nodules, is it reasonable to perform a 'blind biopsy;' ie, of normal-appearing conj?

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What other treatment is important to include, and why?

Cycloplegia (if there is a significant anterior component), both for comfort and to prevent synechiae

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