

Uveitis: *Behçet*

Basics

In one word, what sort of condition is Behçet disease (BD)?

- 1) The uveitis is profiled
- 2) The profiled case is meshed
- 3) A differential diagnosis list is generated
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Turkey, in which the prevalence may be as high as **#** per 100,000 individuals

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Turkey, in which the prevalence may be as high as 300 per 100,000 individuals

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Is BD common in the US? What is the prevalence here?

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Is BD common in the US? What is the prevalence here?

It is an uncommon disease in the US, with a prevalence of about # per 100,000

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Turkey, in which the prevalence may be as high as 300 per 100,000 individuals

Is BD common in the US? What is the prevalence here?

It is an uncommon disease in the US, with a prevalence of about 0.4 per 100,000

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Is there a gender predilection?

This is controversial. The literature seems to suggest a male preponderance; however, many researchers feel this represents sampling bias, and argue the prevalence is roughly equal between men and women.

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Is there a gender predilection?

This is controversial. The literature seems to suggest a male preponderance; however, many researchers feel this represents sampling bias, and argue the prevalence is roughly equal between **men and women**. That said, men **are** more likely to present with ocular manifestations, and are more likely to suffer severe/sight-threatening ocular disease. Thus, thinking of BD as having a male preponderance *might* be useful when meshing a uveitis case for the OKAP and/or Boards.

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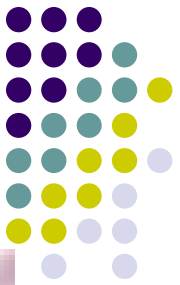
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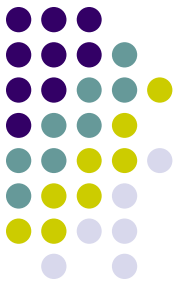
Recurrent oral aphthous ulcers. In some classification schemes, you can't diagnose BD without them.

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Behçet, multiple oral aphthae



Behçet, single oral aphthus



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Are the oral ulcers painful?

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Where is the classic location in BD (and other diseases)?

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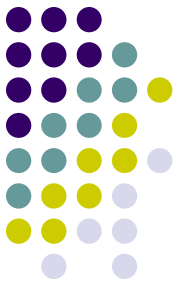
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Where is the classic location in BD (and other diseases)?

The pre-tibial region

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Erythema nodosum in Behçet

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What do the genital ulcers look like?

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What do the genital ulcers look like?

Like the oral ulcers--small, with raised margins

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Behçet: Ulcer

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What do the genital ulcers look like?

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Are they painful?

In men--always; in women--sometimes

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What joint is classically affected in BD?

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What joint is classically affected in BD?

The knee

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Uveitis: *Behçet*

Basics

In one word, what sort of condition is Behçet disease (BD)?

A **vasculitis**

Who is the typical BD pt?

An individual of 'Silk Road' descent age 25-35

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CNS (aka '*Neuro-BD*') carries a significant mortality risk

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Yes; estimates of ocular involvement run as high as

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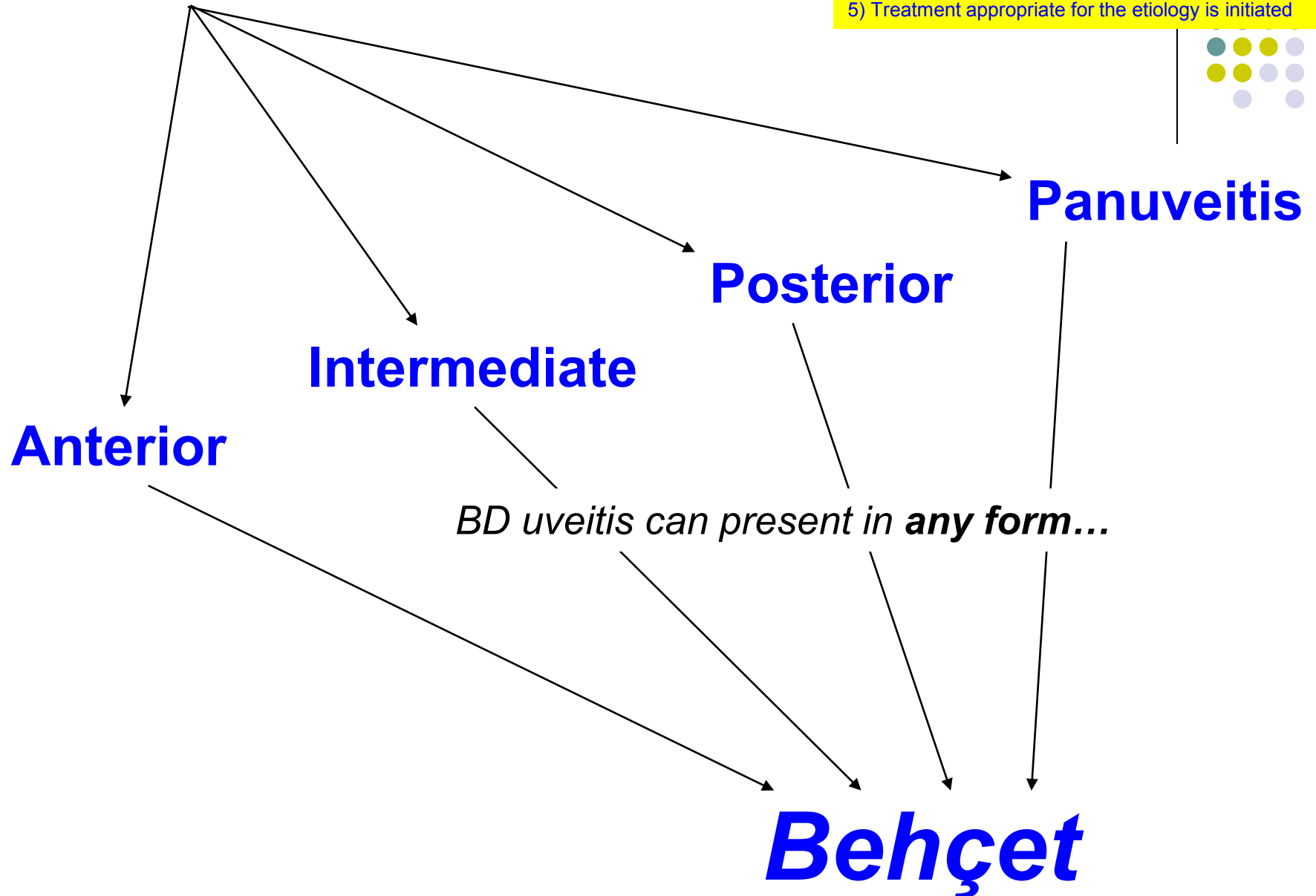
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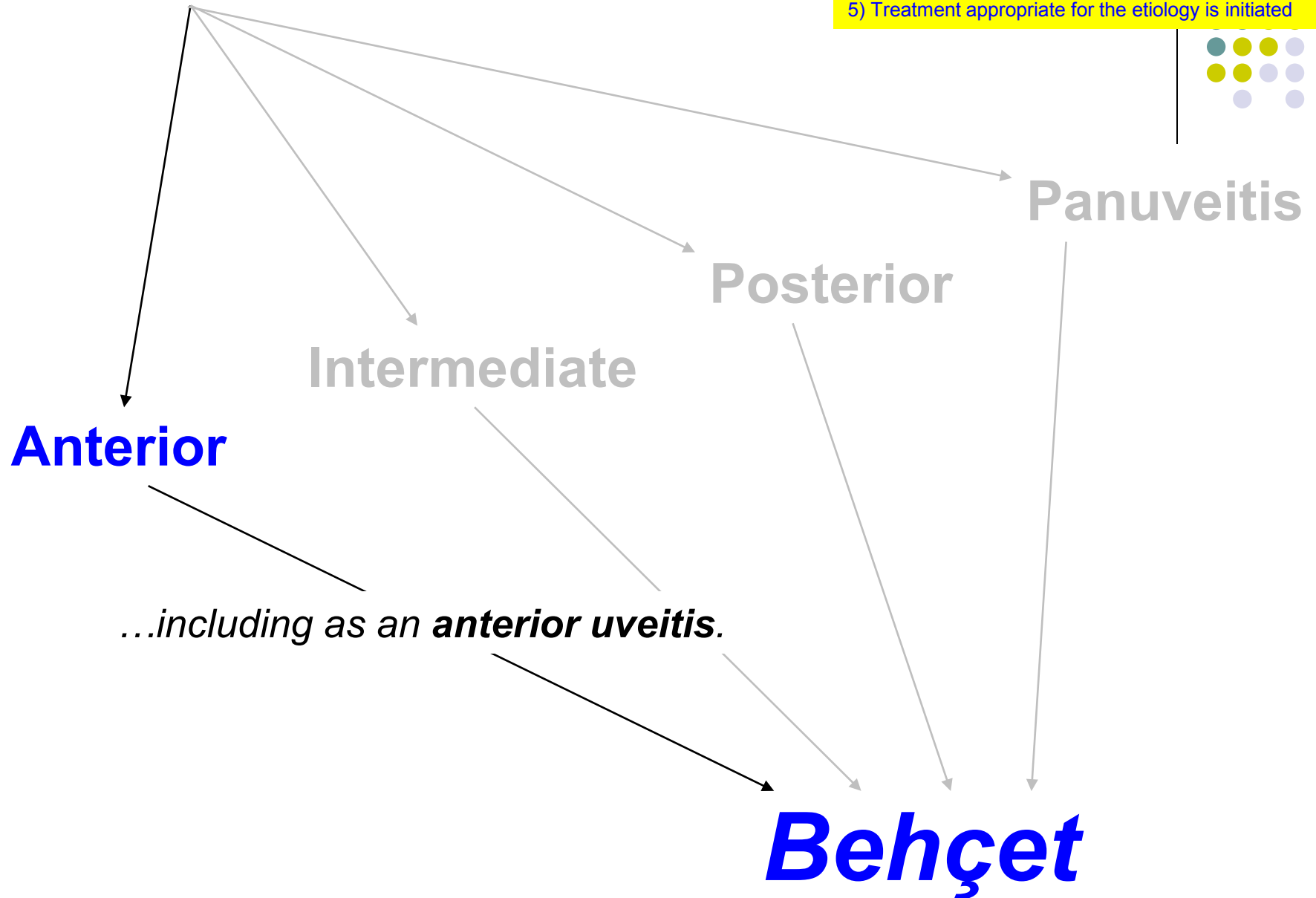
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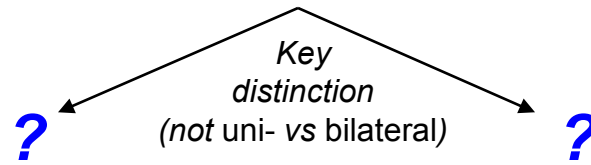


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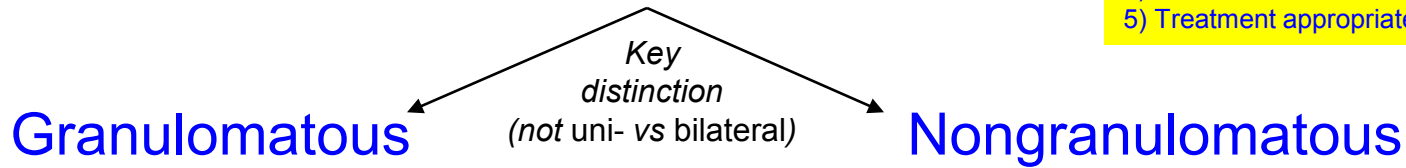
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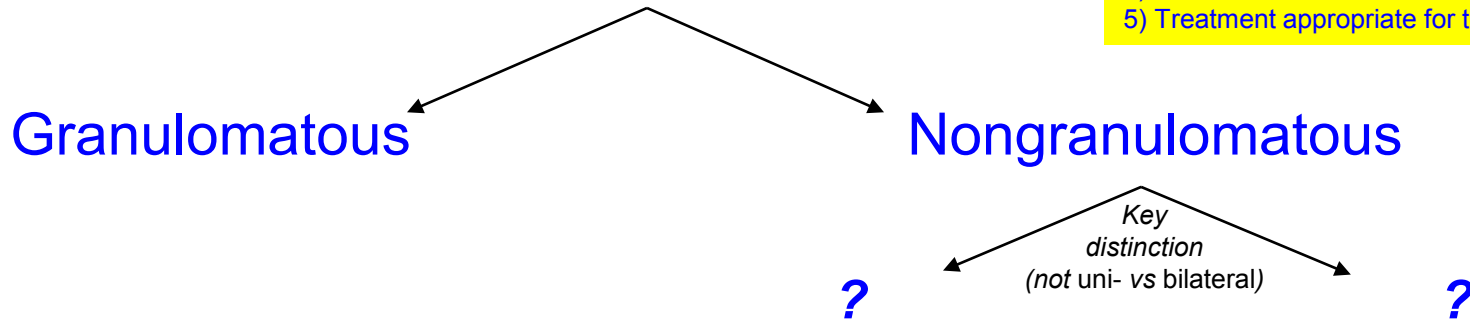
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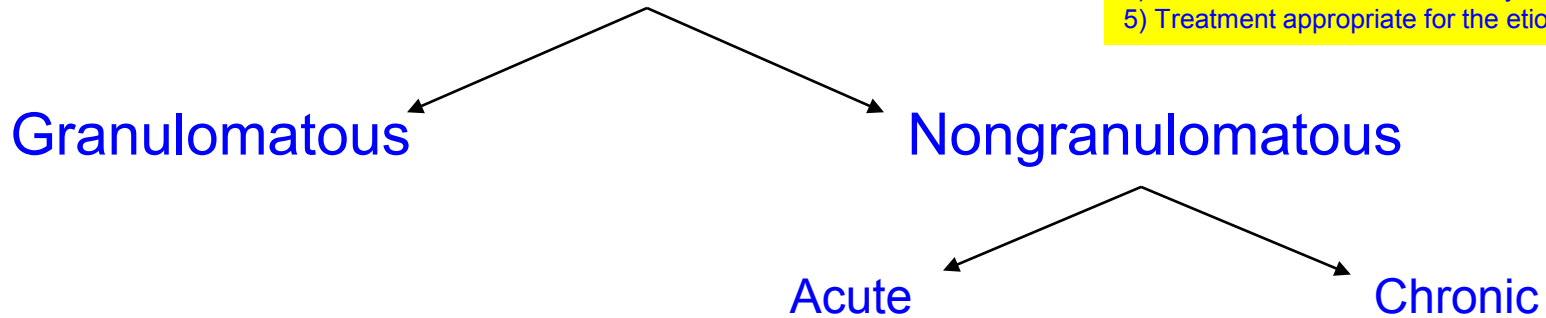
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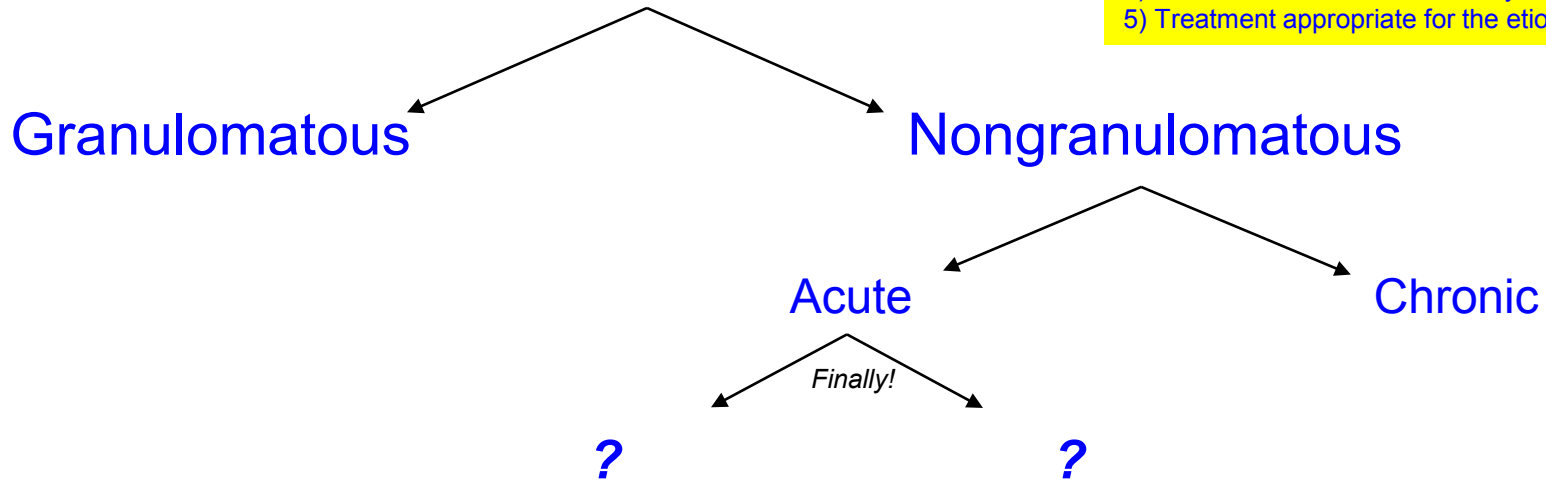
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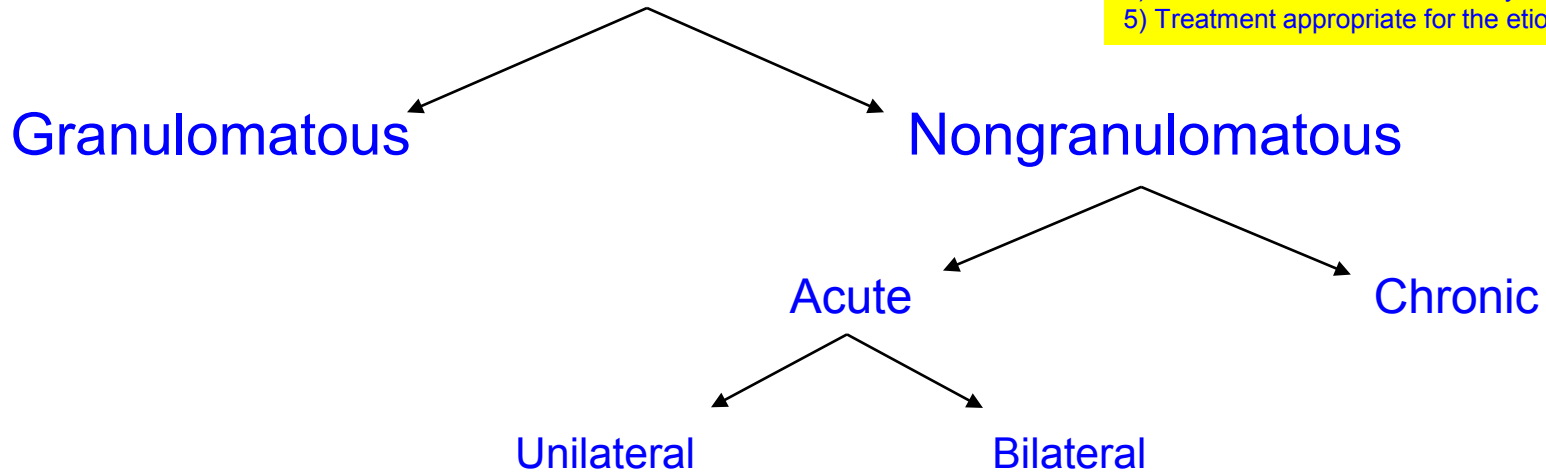
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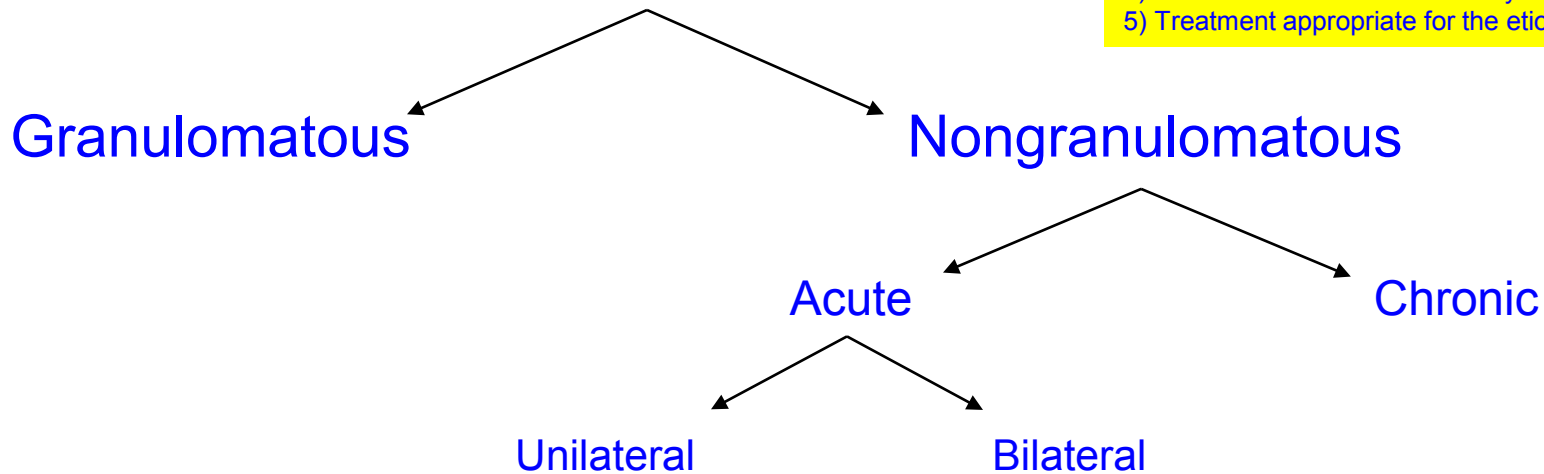
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When BD presents as an anterior uveitis, in which form is it most likely to occur?

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Granulomatous

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

Nongranulomatous

Acute

Unilateral

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

Bilateral

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

Chronic

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

When BD presents as an anterior uveitis, in which form is it most likely to occur?
As an **acute bilateral nongranulomatous uveitis**

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In those cases wherein BD presents as an acute anterior uveitis, what is the classic presentation?

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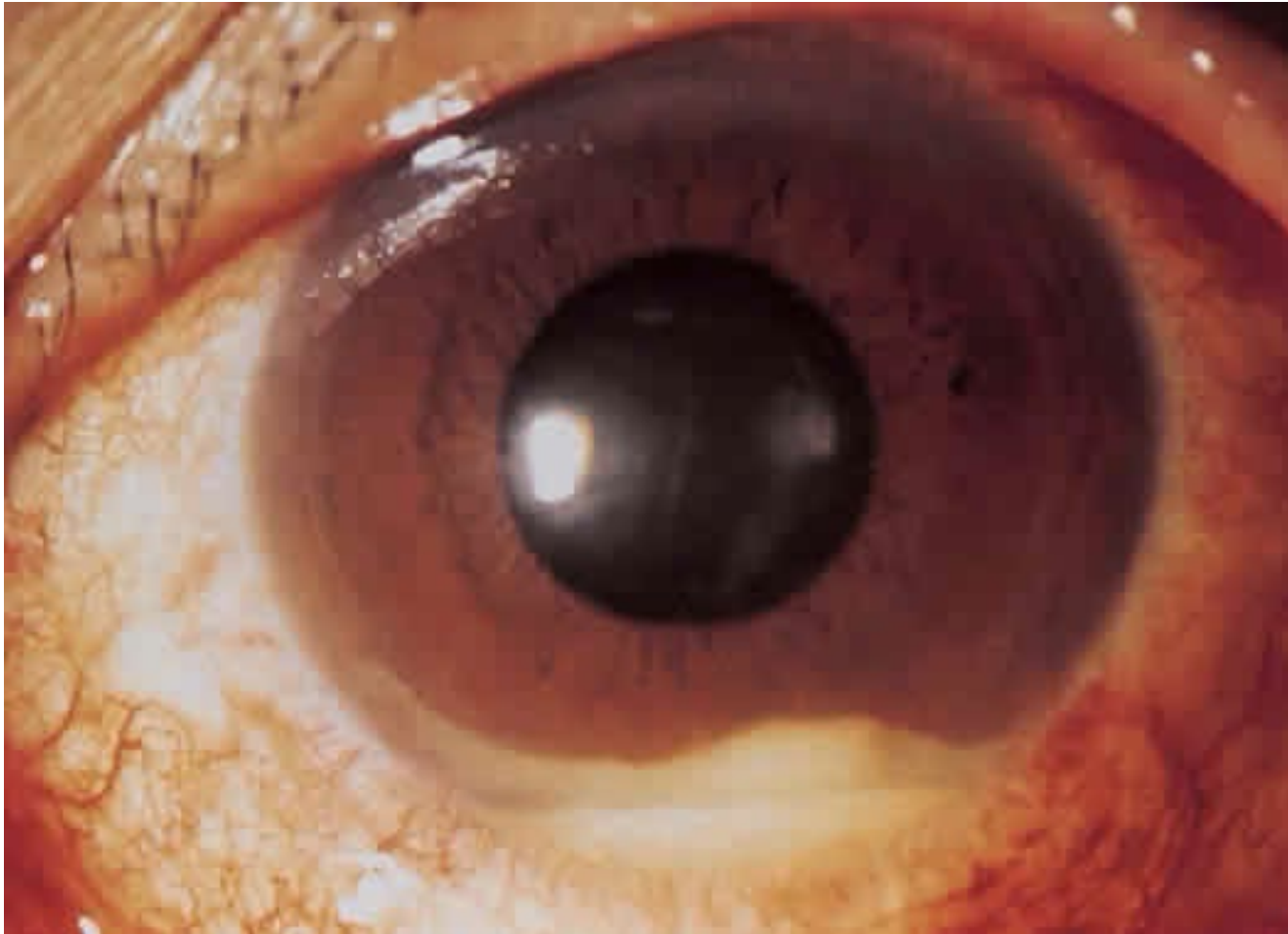
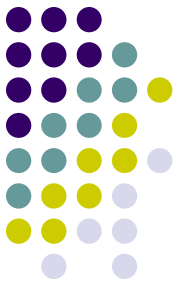
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A dense, bilateral, nongranulomatous iritis, occasionally with hypopyon



BD: Hypopyon

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Granulomatous

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

Nongranulomatous

Acute

Unilateral

- HLA-B27 dz
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Bilateral

- TINU
- **Behçet**
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Retinoblastoma presents with one of three growth patterns—what are they?

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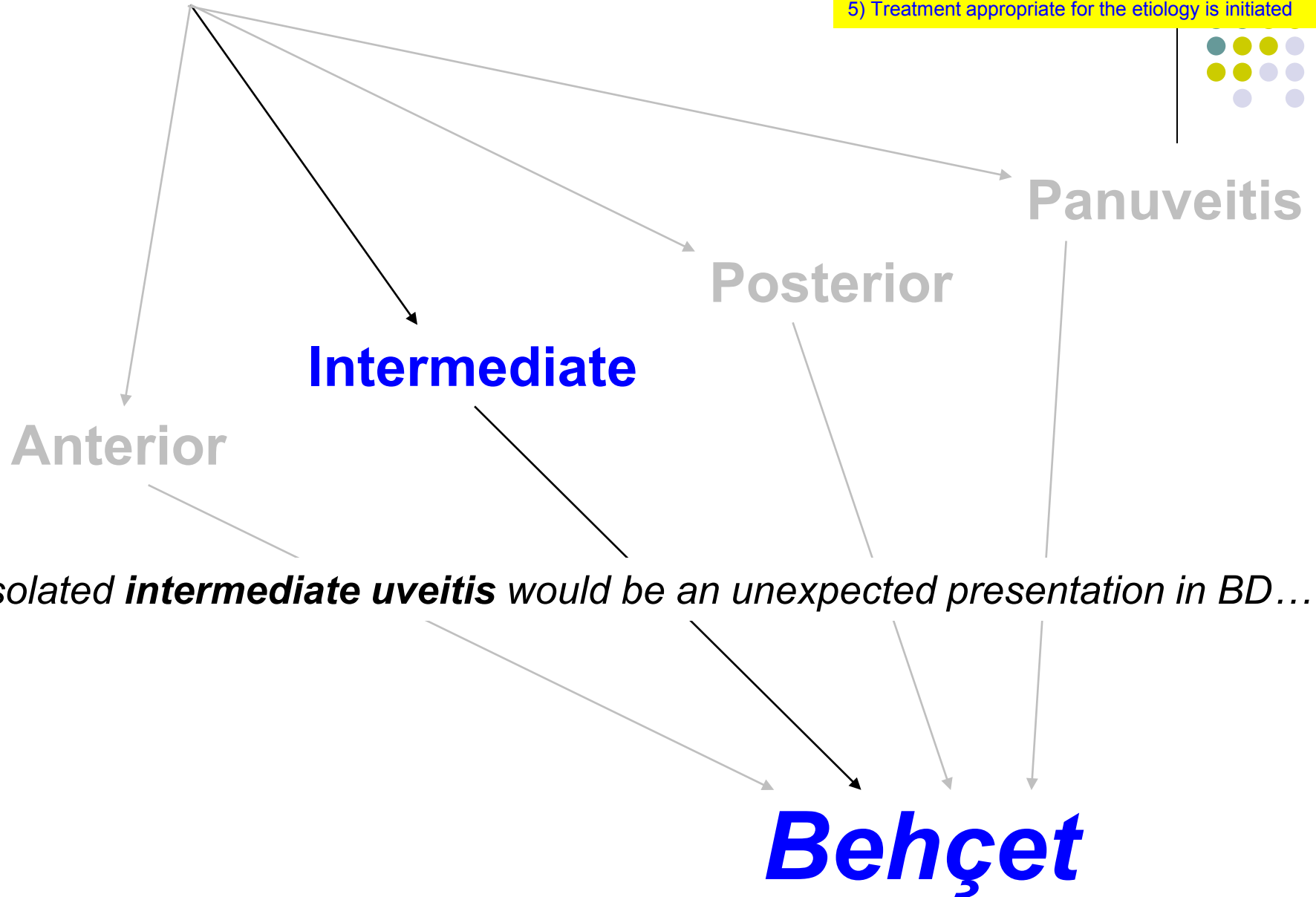
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Explosive! In BD, an eye can go from 'quiet' to severely inflamed in a matter of hours

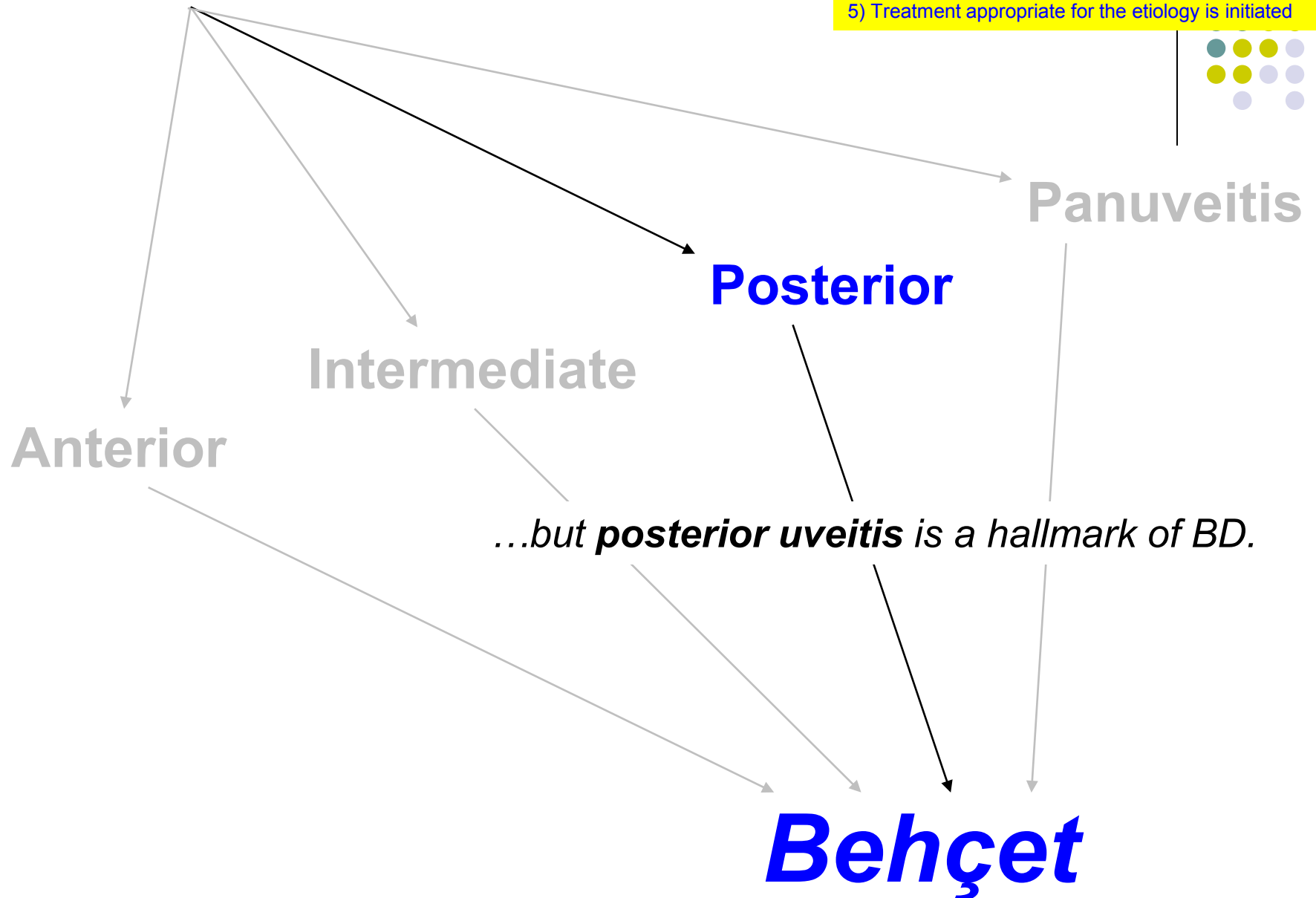
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Choroiditis

**Chorioretinitis or
Retinochoroiditis**

Retinitis

Neuroretinitis



What is the classic posterior manifestation of BD?

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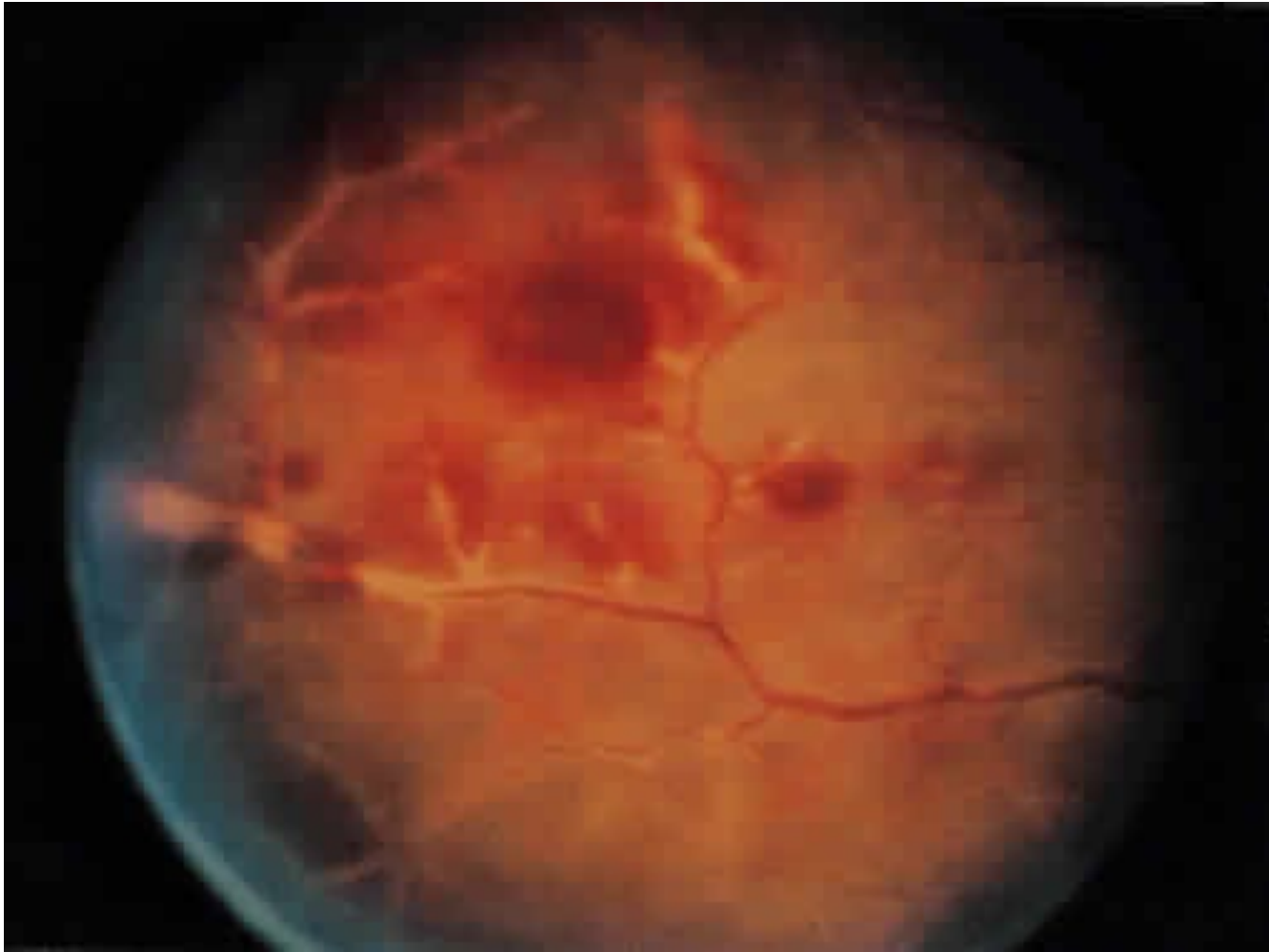
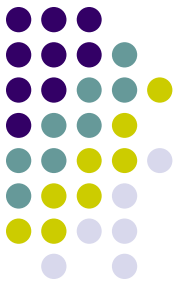
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BD: Vasculitis

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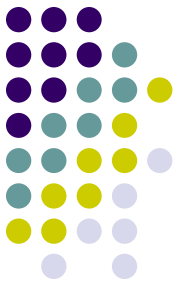
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BD: Occlusive vasculitis

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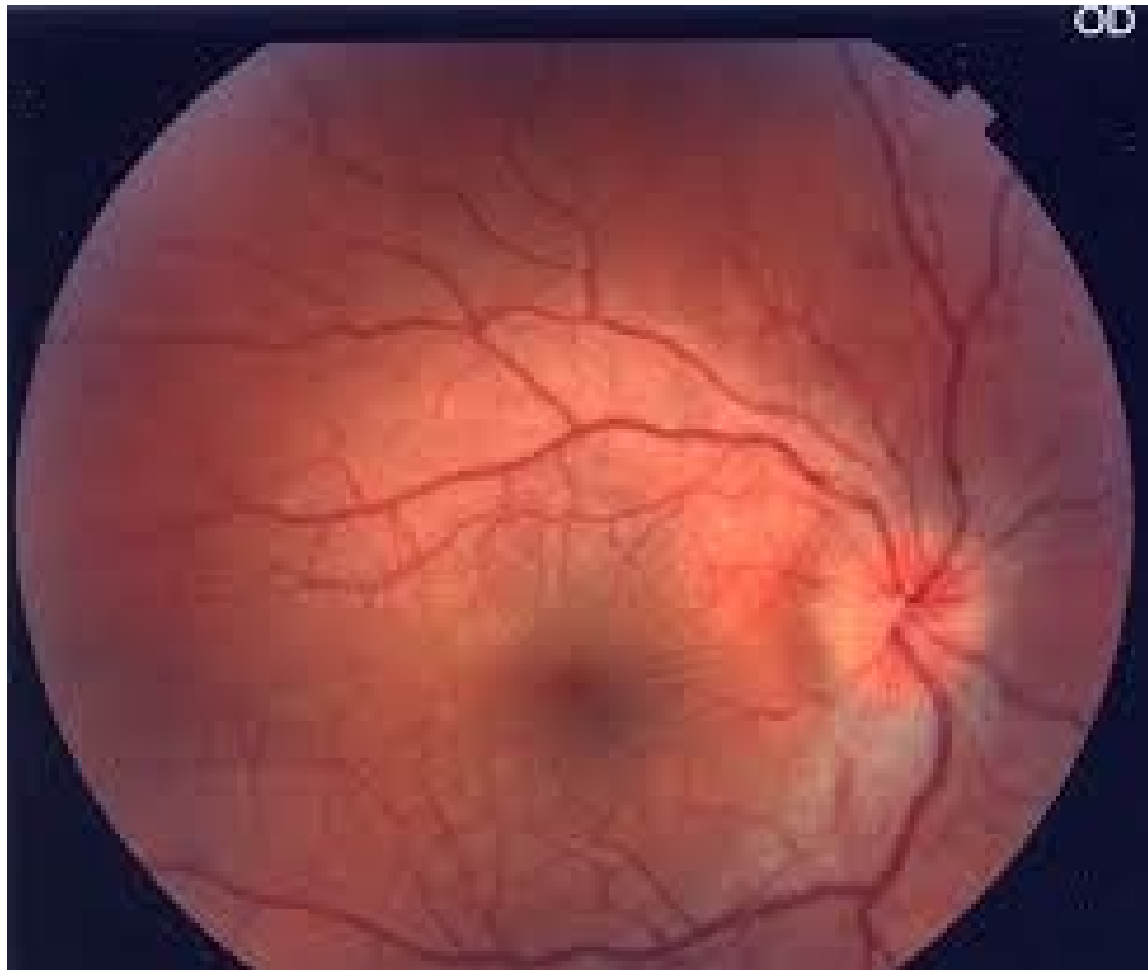
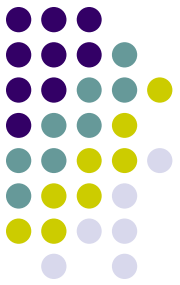
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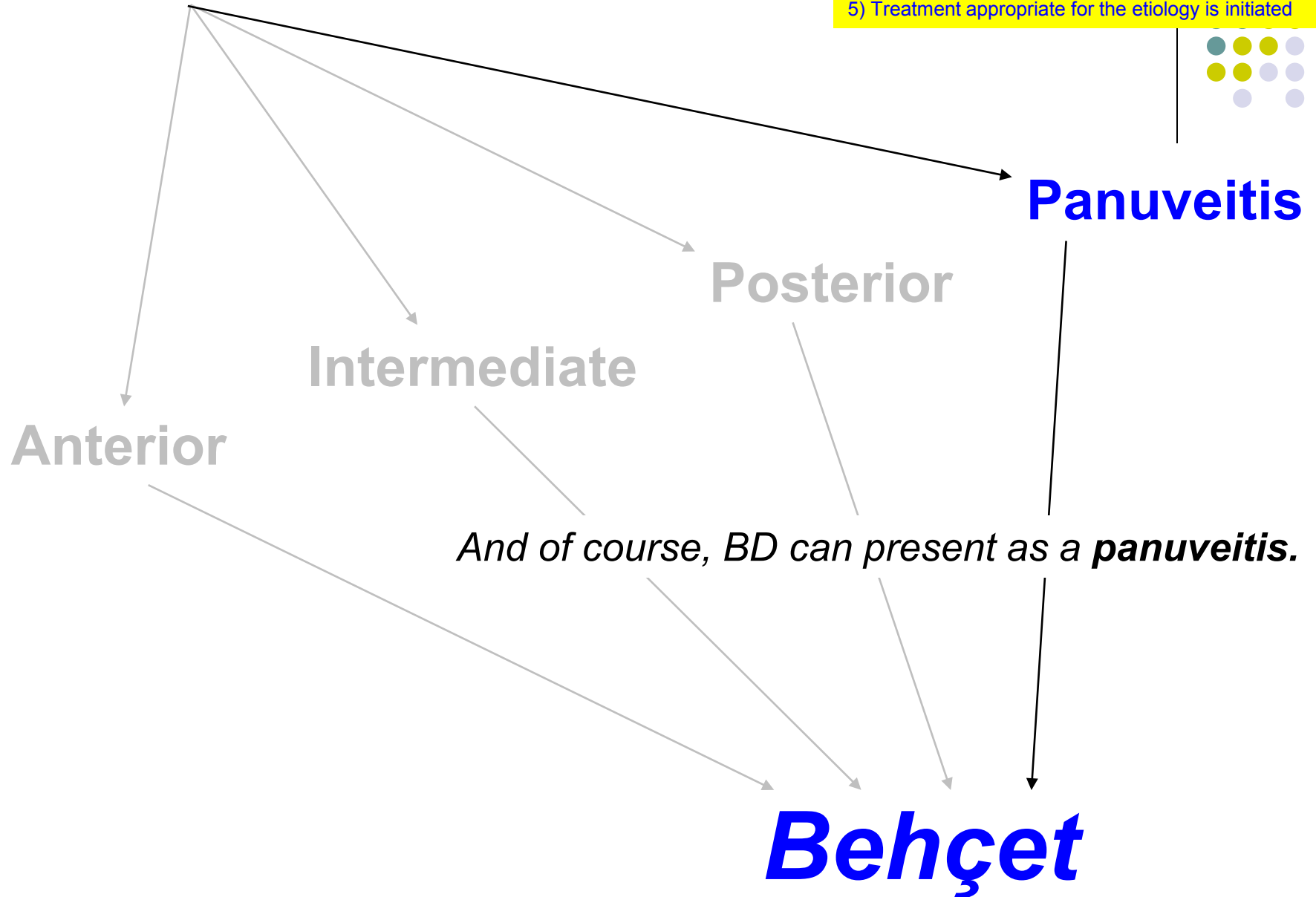
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BD: Papillitis

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Uveitis: *Behçet*

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How is the diagnosis of BD made?

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The BCSC *Uveitis* book contend it is “of little value”

In the context of BD, what is cutaneous pathergy testing?

A test for the presence of pathergy, which is the formation of sterile pustules in response to skin trauma

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Uveitis: *Behçet*

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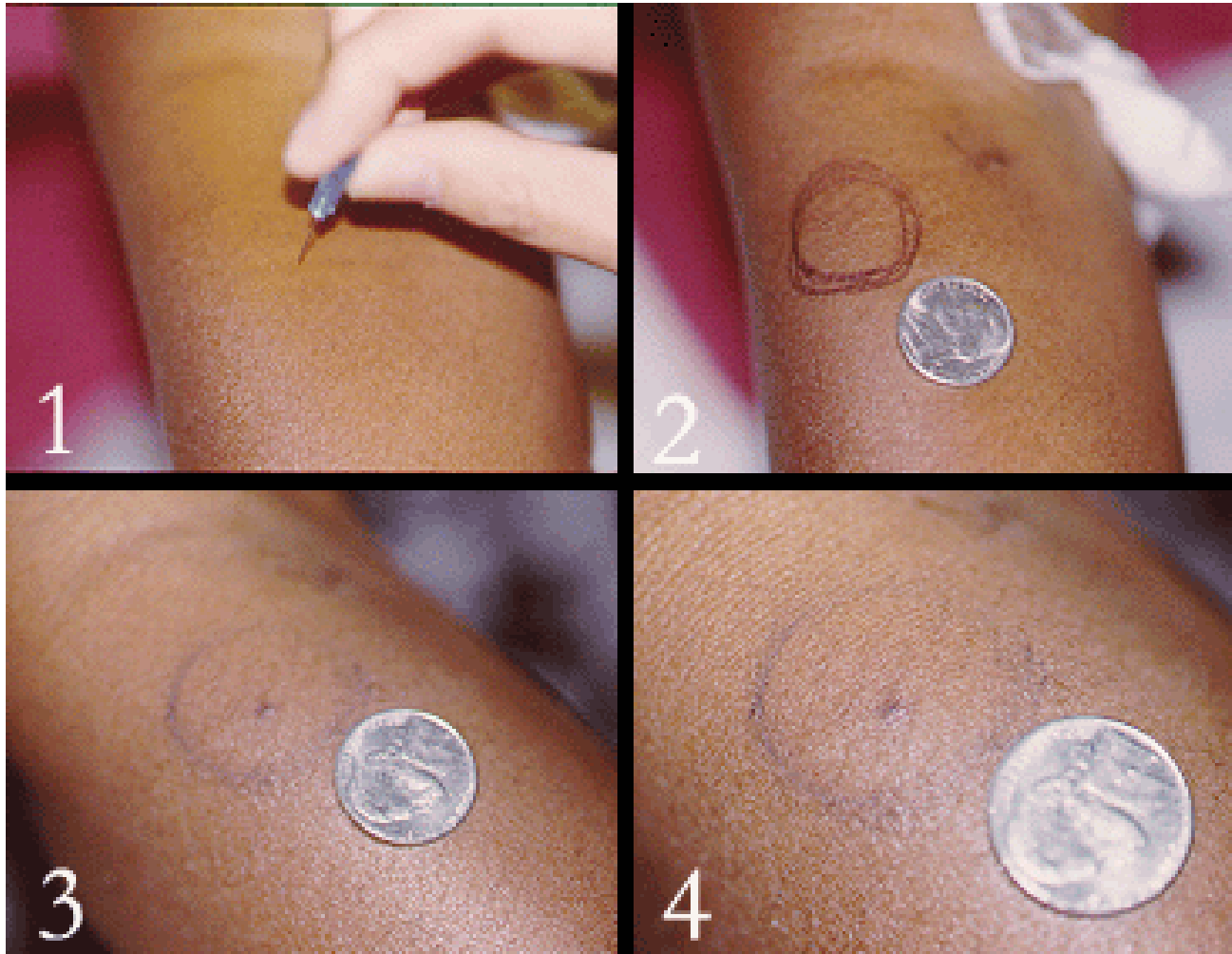
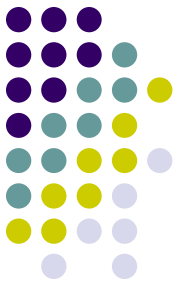
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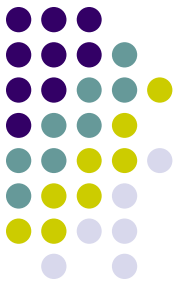
A hypodermic needle is used to prick the skin. In some variations of the test, a small amount of isotonic saline is injected. The area is then assessed for the development of pustules.

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The pathergy test. 1) taken at the time when the patient was “stuck” with the sterile needle; 2) shows the area immediately after the stick; 3) & 4) show the area one day and two days after the needle stick, respectively.



Pathergy test: Positive results

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Is the pathergy response pathognomonic for BD?

No

So, is cutaneous pathergy testing useful in diagnosing BD?

It's not clear whether the BCSC authors consider pathergy testing worthwhile. They say it is “of little value,” but in the same paragraph, assert that BD should be diagnosed per one of two sets of clinical criteria, one of which incorporates pathergy testing!

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Uveitis: *Behçet*

Treatment

What are the two goals in the management of BD?

- 1) Control/resolve acute inflammation
- 2) Suppress chronic inflammation/reduce the risk of recurrences

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Uveitis: *Behçet*

Treatment

What are the two goals in the management of BD? Broadly, how is each achieved?

- 1) Control/resolve acute inflammation with class of med
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Uveitis: *Behçet*

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Treatment

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- 1) Control/resolve acute inflammation **with corticosteroids**
- 2) Suppress chronic inflammation/reduce the risk of recurrences **via**

different class of meds

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Sterility. Pts wishing to have biological children should be encouraged to consider sperm/embryo banking.

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Infliximab is effective, but its side-effect profile renders it best used as a short-term induction agent