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

Basics

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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**Basics**

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

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*Who is the typical BD pt?*
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Who is the typical BD pt?
An individual of ‘Silk Road’ descent age 25-35
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*What is/was the Silk Road?*
An ancient trading route extending from the eastern Mediterranean to the Far East
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What regions/countries are found along the Silk Road?
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Is there a gender predilection?
**Uveitis: Behçet**

**Basics**

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

*Who is the typical BD pt?*
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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.
**Uveitis: Behçet**

**Basics**

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

A vasculitis

*Who is the typical BD pt?*

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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.
Uveitis: Behçet

**Basics**

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*What the classic nonocular complaint in BD?*
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What the classic nonocular complaint in BD?
Recurrent oral aphthous ulcers. In some classification schemes, you can’t diagnose BD without them.
Behçet, multiple oral aphthae
Behçet, single oral aphthus
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Are the oral ulcers painful?
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What is the typical complaint in BD?
Recurrent oral aphthous ulcers. In some classification schemes, you can’t diagnose BD without them.

What are three other findings that are particularly common?
Skin rash, genital ulcers and arthralgias

What is the classic rash?
Reddish nodules, painful, in the pre-tibial region
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What is the classic rash?
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Is it painful?
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**Skin rash**

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*What does erythema nodosum look like?*
Reddish nodules

*Is it painful?*
Yes
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What is the classic rash?
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What does erythema nodosum look like?
Reddish nodules

Is it painful?
Yes

Where is the classic location in BD (and other diseases)?
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Is it painful?
Yes

Where is the classic location in BD (and other diseases)?
The pre-tibial region
Erythema nodosum in Behçet
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What do the genital ulcers look like?
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In addition to uveitis and aphthous ulcers, what three other findings are particularly common?
Skin rash, genital ulcers, and arthralgias

What do the genital ulcers look like?
Like the oral ulcers--small, with raised margins
Behçet: Ulcer
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What do the genital ulcers look like?
Like the oral ulcers--small, with raised margins

Are they painful?
In men--always; in women--sometimes
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What joint is classically affected in BD?

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Besides the above-mentioned manifestations and ocular inflammation, what other organ systems can be involved?
Any organ-system can be affected; however, involvement of the following systems is classic:

Cardiac: Inflammation of the coronary arteries and/or cardiac tissue itself (eg, endocarditis; pericarditis)
GI: Ulcers (often multiple) of the pre-colonic GI tract
Lung: Pulmonary arteritis
CNS: Motor conues; CVAs; CN palsies; confusion; increased ICP → pseudotumor cerebri-type presentation

Of these, which is considered to be the most ominous?
CNS (aka 'Neuro-BD') carries a significant mortality risk
**Uveitis: Behçet**

**Basics**

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**Uveitis: Behçet**

### Basics

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Uveitis: *Behçet*

**Basics**

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Is uveitis usually an early manifestation of BD?
In one word, what sort of condition is Behçet disease (BD)?
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Is uveitis usually an early manifestation of BD?
No; it usually follows the onset of disease by several years
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Is ocular involvement common in BD?
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Recurrent oral aphthous ulcers. In some classification schemes, you can’t diagnose BD without them.

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Skin rash, genital ulcers and arthralgias

Is uveitis usually an early manifestation of BD?
No; it usually follows the onset of disease by several years

Is ocular involvement common in BD?
Yes; estimates of ocular involvement run as high as %
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

What the classic nonocular complaint in BD?
Recurrent oral aphthous ulcers. In some classification schemes, you can’t diagnose BD without them.

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Is ocular involvement common in BD?
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Does BD pose a significant threat to long-term ocular health, and/or vision?
In one word, what sort of condition is Behçet disease (BD)?
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Does BD pose a significant threat to long-term ocular health, and/or vision?
Most definitely. Severe vision loss results in as many as % of ocular BD cases
In one word, what sort of condition is Behçet disease (BD)?
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Does BD pose a significant threat to long-term ocular health, and/or vision?
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Does BD pose a significant threat to long-term ocular health, and/or vision?
Most definitely. Severe vision loss results in as many as 25% of ocular BD cases

What is the cause of BD?
In one word, what sort of condition is Behçet disease (BD)?
A vasculitis

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An individual of ‘Silk Road’ descent age 25-35

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Is ocular involvement common in BD?
Yes; estimates of ocular involvement run as high as 70%

Does BD pose a significant threat to long-term ocular health, and/or vision?
Most definitely. Severe vision loss results in as many as 25% of ocular BD cases

What is the cause of BD?
It is unknown at this time
Uveitis

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

Anterior

Posterior

Intermediate

Panuveitis

Behçet

BD uveitis can present in any form
Uveitis

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

...including as an anterior uveitis.

Behçet
Uveitis: *Anterior*

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

Key distinction

*(not uni- vs bilateral)*
Uveitis: Anterior

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

Key distinction:
(not uni- vs bilateral)

Granulomatous       Nongranulomatous
Uveitis: Anterior

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

Granulomatous → Nongranulomatous

Key distinction
(not uni- vs bilateral)
Uveitis: **Anterior**

- Granulomatous
- Nongranulomatous
  - Acute
  - Chronic

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
Uveitis: Anterior

- Granulomatous
- Nongranulomatous
  - Acute
  - Chronic

Finally!

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
Uveitis: **Anterior**

Granulomatous

Nongranulomatous

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

Acute

- Unilateral

Chronic

- Bilateral
When BD presents as an anterior uveitis, in which form is it most likely to occur?
When BD presents as an anterior uveitis, in which form is it most likely to occur? As an **acute bilateral nongranulomatous uveitis**
In those cases wherein BD presents as an acute anterior uveitis, what is the classic presentation?
In those cases wherein BD presents as an acute anterior uveitis, what is the classic presentation? A dense, bilateral, nongranulomatous iritis, occasionally with hypopyon.
BD: Hypopyon
Uveitis: Anterior

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

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

Nongranulomatous

Acute
- Unilateral
  - HLA-B27 dz
  - Posner-Schlossman
  - Sarcoid
  - Syphilis
  - HSV/VZV
- Bilateral
  - TINU
  - Behçet
  - Drug rxn
  - Leptospirosis
  - Sarcoid

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

In those cases wherein BD presents as an acute anterior uveitis, what is the classic presentation?
A dense, bilateral, nongranulomatous iritis, occasionally with hypopyon

What percent of cases develop hypopyon?
In those cases wherein BD presents as an acute anterior uveitis, what is the classic presentation? A dense, bilateral, nongranulomatous iritis, occasionally with hypopyon.

What percent of cases develop hypopyon? Roughly 25
In those cases wherein BD presents as an acute anterior uveitis, what is the classic presentation? A dense, bilateral, nongranulomatous iritis, occasionally with hypopyon. What percent of cases develop hypopyon? Roughly 25. The hypopyon in BD has a fairly unique property—what is it? Unlike most hypopyons, it tends to shift with changes in head position.
Uveitis: *Anterior*

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

**Nongranulomatous**

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

**Chronic**
- TINU
- Drug rxn
- Leptospirosis
- Sarcoid

In those cases wherein BD presents as an acute anterior uveitis, what is the classic presentation? A dense, bilateral, nongranulomatous iritis, occasionally with hypopyon.

What percent of cases develop hypopyon? Roughly 25

The hypopyon in BD has a fairly unique property—what is it? Unlike most hypopyons, it tends to shift with changes in head position.
In those cases wherein BD presents as an acute anterior uveitis, what is the classic presentation?
A dense, bilateral, nongranulomatous iritis, occasionally with hypopyon.

What percent of cases develop hypopyon?
Roughly 25

The hypopyon in BD has a fairly unique property—what is it?
Unlike most hypopyons, it tends to shift with changes in head position.

A hypopyon that shifts with head movements is characteristic of the pseudohypopyon associated with what masquerade syndrome?

Retinoblastoma, diffuse-infiltrating type
In those cases wherein BD presents as an acute anterior uveitis, what is the classic presentation? A dense, bilateral, nongranulomatous iritis, occasionally with hypopyon.

What percent of cases develop hypopyon? Roughly 25%

The hypopyon in BD has a fairly unique property—what is it? Unlike most hypopyons, it tends to shift with changes in head position.

A hypopyon that shifts with head movements is characteristic of the pseudohypopyon associated with what masquerade syndrome? Retinoblastoma.
**Uveitis: Anterior**

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2. The profiled case is meshed
3. A differential diagnosis list is generated
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**Granulomatous**
- TB
- Syphilis
- Sarcoid
- HSV
- VKH
- Toxoplasmosis
- Lyme

**Nongranulomatous**

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

**Chronic**
- Bilateral
  - TINU
  - Drug rxn
  - Leptospirosis
  - Sarcoid

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

**What word is often used to characterize the onset of BD anterior uveitis?**
Explosive! In BD, an eye can go from 'quiet' to severely inflamed in a matter of hours

**What percent of cases develop hypopyon?**
Roughly 25

**The hypopyon in BD has a fairly unique property—what is it?**
Unlike most hypopyons, it tends to shift with changes in head position

A hypopyon that shifts with head movements is characteristic of the pseudohypopyon associated with what masquerade syndrome?

**Retinoblastoma**
Uveitis: **Anterior**

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

**Nongranulomatous**

**Acute**
- Unilateral
  - HLA-B27 dz
  - Posner-Schlossman
  - Sarcoid
  - Syphilis
  - HSV/VZV
- Bilateral
  - TINU
  - Behçet
  - Drug rxn
  - Leptospirosis
  - Sarcoid

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

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**Retinoblastoma**

- Presents with one of three growth patterns—what are they?
  - Exophytic
  - Endophytic
  - Diffuse infiltrating

**In those cases wherein BD presents as an acute anterior uveitis, what is the classic presentation?**
- A dense, bilateral, nongranulomatous iritis, occasionally with hypopyon

**What percent of cases develop hypopyon?**
- Roughly 25%

**What word is often used to characterize the onset of BD anterior uveitis?**
- Explosive! In BD, an eye can go from ‘quiet’ to severely inflamed in a matter of hours

**The hypopyon in BD has a fairly unique property—what is it?**
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**A hypopyon that shifts with head movements is characteristic of the pseudohypopyon associated with what masquerade syndrome?**
- Retinoblastoma

---

**Retinoblastoma**

- Presents with one of three growth patterns—what are they?
  - Exophytic
  - Endophytic
  - Diffuse infiltrating
Uveitis: **Anterior**

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

**Nongranulomatous**

**Acute**
- Unilateral
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- Bilateral
  - TINU
  - Behçet
  - Drug rxn
  - Leptospirosis
  - Sarcoid
  - TB

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

---

**Retinoblastoma presents with one of three growth patterns—what are they?**
- Exophytic
- Endophytic
- Diffuse infiltrating

**Which type may present with a pseudohypopyon?**

---

**In those cases wherein BD presents as an acute anterior uveitis, what is the classic presentation?**
- Dense, bilateral, nongranulomatous iritis, occasionally with hypopyon

**What percent of cases develop hypopyon?**
- Roughly 25%

**What word is often used to characterize the onset of BD anterior uveitis?**
- Explosive! In BD, an eye can go from 'quiet' to severely inflamed in a matter of hours

**The hypopyon in BD has a fairly unique property—what is it?**
- Unlike most hypopyons, it tends to shift with changes in head position

---

**Posner-Schlossman Syndrome**

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**Retinoblastoma**
Uveitis: **Anterior**

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

Nongranulomatous

Acute
- Unilateral
  - HLA-B27 dz
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  - Drug rxn
  - Leptospirosis
  - Sarcoid

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

**Retinoblastoma** presents with one of three growth patterns—what are they?
- Exophytic
- Endophytic
- Diffuse infiltrating

Which type may present with a pseudohypopyon? Diffuse infiltrating

Retinoblastoma

What word is often used to characterize the onset of BD anterior uveitis?
**Explosive!** In BD, an eye can go from 'quiet' to severely inflamed in a matter of hours

The hypopyon in BD has a fairly unique property—what is it?
Unlike most hypopyons, it tends to shift with changes in head position

A hypopyon that shifts with head movements is characteristic of the **pseudohypopyon** associated with what masquerade syndrome?

Retinoblastoma presents with one of three growth patterns—what are they?
- Exophytic
- Endophytic
- Diffuse infiltrating

What percent of cases develop hypopyon?
Roughly 25

In those cases wherein BD presents as an acute anterior uveitis, what is the classic presentation?
A dense, bilateral, nongranulomatous iritis, occasionally with hypopyon

What is the classic presentation of BD with hypopyon?
In those cases wherein BD presents as an acute anterior uveitis, what is the classic presentation? A dense, bilateral, nongranulomatous iritis, occasionally with hypopyon.

What percent of cases develop hypopyon? Roughly 25.

What word is often used to characterize the onset of BD anterior uveitis?
**Uveitis: Anterior**

1) The uveitis is profiled
2) The profiled case is meshed
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- TB
- Syphilis
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- VKH
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- Acute
- Unilateral
  - HLA-B27 dz
  - Posner-Schlossman
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  - Syphilis
  - HSV/VZV
- Bilateral
  - TINU
  - Behçet
  - Drug rxn
  - Leptospirosis
  - Sarcoid

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

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**In those cases wherein BD presents as an acute anterior uveitis, what is the classic presentation?**
A dense, bilateral, nongranulomatous iritis, occasionally with hypopyon

**What percent of cases develop hypopyon?**
Roughly 25

**What word is often used to characterize the onset of BD anterior uveitis?**
Explosive! In BD, an eye can go from ‘quiet’ to severely inflamed in a matter of hours
Isolated *intermediate uveitis* would be an unexpected presentation in BD…
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2) The profiled case is meshed
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4) Studies are obtained to identify the etiology
5) Treatment appropriate for the etiology is initiated

Uveitis

Anterior

Intermediate

Posterior

Panuveitis

…but posterior uveitis is a hallmark of BD.

Behçet
What is the classic posterior manifestation of BD?
What is the classic posterior manifestation of BD?
A retinal vasculitis
Uveitis: *Posterior*

- Choroiditis
- Chorioretinitis or Retinochoroiditis
- Retinitis
- Neuroretinitis

1) The uveitis is profiled
2) The profiled case is meshed
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5) Treatment appropriate for the etiology is initiated

*What is the classic posterior manifestation of BD?*
A retinal vasculitis

*Does it affect the arterioles, venules, or both?*
What is the classic posterior manifestation of BD?
A retinal vasculitis

Does it affect the arterioles, venules, or both?
Both
BD: Vasculitis
Uveitis: **Posterior**

- Choroiditis
- Chorioretinitis or Retinochoroiditis
- Retinitis
- Neuroretinitis

---

**What is the classic posterior manifestation of BD?**
A retinal vasculitis

**Does it affect the arterioles, venules, or both?**
Both

**Can it affect the main arterioles and venules, leading to BRAOs and/or BRVOs?**
What is the classic posterior manifestation of BD?
A retinal vasculitis

Does it affect the arterioles, venules, or both?
Both

Can it affect the main arterioles and venules, leading to BRAOs and/or BRVOs?
Yes. In fact, these can occur simultaneously
What is the classic posterior manifestation of BD?
A retinal vasculitis

Does it affect the arterioles, venules, or both?
Both

Can it affect the main arterioles and venules, leading to BRAOs and/or BRVOs?
Yes. In fact, these can occur simultaneously

Is it necrotizing, or non-necrotizing?
What is the classic posterior manifestation of BD?
A retinal vasculitis

Does it affect the arterioles, venules, or both?
Both

Can it affect the main arterioles and venules, leading to BRAOs and/or BRVOs?
Yes. In fact, these can occur simultaneously

Is it necrotizing, or non-necrotizing?
Necrotizing. BD retinal vasculitis can appear similar to acute retinal necrosis (ARN)
What is the classic posterior manifestation of BD?
A retinal vasculitis

Does it affect the arterioles, venules, or both?
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Yes. In fact, these can occur simultaneously

Is it necrotizing, or non-necrotizing?
Necrotizing. BD retinal vasculitis can appear similar to acute retinal necrosis (ARN)

Can BD retinal vasculitis lead to retinal ischemia with subsequent neovascularization?
What is the classic posterior manifestation of BD?
A retinal vasculitis

Does it affect the arterioles, venules, or both?
Both

Can it affect the main arterioles and venules, leading to BRAOs and/or BRVOs?
Yes. In fact, these can occur simultaneously

Is it necrotizing, or non-necrotizing?
Necrotizing. BD retinal vasculitis can appear similar to acute retinal necrosis (ARN)

Can BD retinal vasculitis lead to retinal ischemia with subsequent neovascularization?
Yes--it is an occlusive vasculitis, so ischemia is a common occurrence
BD: Occlusive vasculitis
What is the classic posterior manifestation of BD?
A retinal vasculitis

Does it affect the arterioles, venules, or both?
Both

Can it affect the main arterioles and venules, leading to BRAOs and/or BRVOs?
Yes. In fact, these can occur simultaneously

Is it necrotizing, or non-necrotizing?
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Can BD retinal vasculitis lead to retinal ischemia with subsequent neovascularization?
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Can the optic nerve be involved?
What is the classic posterior manifestation of BD?
A retinal vasculitis

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Necrotizing. BD retinal vasculitis can appear similar to acute retinal necrosis (ARN)

Can BD retinal vasculitis lead to retinal ischemia with subsequent neovascularization?
Yes--it is an occlusive vasculitis, so ischemia is a common occurrence

Can the optic nerve be involved?
Yes--in fact, it frequently is. Papillitis or optic atrophy occurs in about [x]% of ocular BD pts
**Uveitis:** Posterior

- Choroiditis
- Chorioretinitis or Retinochoroiditis
- Retinitis
- Neuroretinitis

What is the classic posterior manifestation of BD?
A retinal vasculitis

*Does it affect the arterioles, venules, or both?*
Both

*Can it affect the main arterioles and venules, leading to BRAOs and/or BRVOs?*
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Necrotizing. BD retinal vasculitis can appear similar to acute retinal necrosis (ARN)

*Can BD retinal vasculitis lead to retinal ischemia with subsequent neovascularization?*
Yes--it is an occlusive vasculitis, so ischemia is a common occurrence

*Can the optic nerve be involved?*
Yes--in fact, it frequently is. Papillitis or optic atrophy occurs in about 25% of ocular BD pts
Uveitis

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

Anterior

Posterior

Intermediate

Panuveitis

And of course, BD can present as a panuveitis.

Behçet
Uveitis: Behçet

How is the diagnosis of BD made?
Uveitis: Behçet

Diagnosis

How is the diagnosis of BD made?
It is a clinical diagnosis—there are no specific tests for it
How is the diagnosis of BD made?
It is a clinical diagnosis—there are no specific tests for it

What is the HLA association for BD?
Uveitis: Behçet

**Diagnosis**

*How is the diagnosis of BD made?*
It is a clinical diagnosis—there are no specific tests for it

*What is the HLA association for BD?*
There are several; HLA-B51 is associated with ocular BD
Uveitis: Behçet

**Diagnosis**

*How is the diagnosis of BD made?*
It is a clinical diagnosis—there are no specific tests for it

*What is the HLA association for BD?*
There are several; HLA-B51 is associated with ocular BD

*So, is HLA testing helpful in diagnosing ocular BD?*
**Uveitis: Behçet**

**Diagnosis**

*How is the diagnosis of BD made?*
It is a clinical diagnosis—there are no specific tests for it

*What is the HLA association for BD?*
There are several; HLA-B51 is associated with ocular BD

*So, is HLA testing helpful in diagnosing ocular BD?*
The BCSC *Uveitis* book contend it is “of little value”
**Uveitis: Behçet**

**Diagnosis**

*How is the diagnosis of BD made?*
It is a clinical diagnosis—there are no specific tests for it

*What is the HLA association for BD?*
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*In the context of BD, what is cutaneous pathergy testing?*
Uveitis: **Behçet**

**Diagnosis**

*How is the diagnosis of BD made?*
It is a clinical diagnosis—there are no specific tests for it

*What is the HLA association for BD?*
There are several; HLA-B51 is associated with ocular BD

*So, is HLA testing helpful in diagnosing ocular BD?*
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
How is the diagnosis of BD made?
It is a clinical diagnosis—there are no specific tests for it.

What is the HLA association for BD?
There are several; HLA-B51 is associated with ocular BD.

So, is HLA testing helpful in diagnosing ocular BD?
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.

How is the cutaneous pathergy test performed?
Diagnosis

How is the diagnosis of BD made?
It is a clinical diagnosis—there are no specific tests for it

What is the HLA association for BD?
There are several; HLA-B51 is associated with ocular BD

So, is HLA testing helpful in diagnosing ocular BD?
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

How is the cutaneous pathergy test performed?
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.
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
**Uveitis: Behçet**

**Diagnosis**

*How is the diagnosis of BD made?*
It is a clinical diagnosis—there are no specific tests for it

*What is the HLA association for BD?*
There are several; HLA-B51 is associated with ocular BD

*So, is HLA testing helpful in diagnosing ocular BD?*
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

*Is this something BD pts experience?*
Uveitis: Behçet

**Diagnosis**

*How is the diagnosis of BD made?*
It is a clinical diagnosis—there are no specific tests for it.

*What is the HLA association for BD?*
There are several; HLA-B51 is associated with ocular BD.

*So, is HLA testing helpful in diagnosing ocular BD?*
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.

*Is this something BD pts experience?*
About 40% do.
Uveitis: Behçet

Diagnosis

How is the diagnosis of BD made?
It is a clinical diagnosis—there are no specific tests for it.

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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!
Uveitis: Behçet

Treatment

What are the two goals in the management of BD?
1) 
2)
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
What are the two goals in the management of BD? Broadly, how is each achieved?
1) Control/resolve acute inflammation with a specific class of medication
2)Suppress chronic inflammation/reduce the risk of recurrences
Uveitis: Behçet

**Treatment**

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

1) Control/resolve acute inflammation with corticosteroids
2) Suppress chronic inflammation/reduce the risk of recurrences
What are the two goals in the management of BD? Broadly, how is each achieved?
1) Control/resolve acute inflammation with corticosteroids
2) Suppress chronic inflammation/reduce the risk of recurrences via different class of meds
Uveitis: Behçet

Treatment

What are the two goals in the management of BD? Broadly, how is each achieved?
1) Control/resolve acute inflammation with corticosteroids
2) Suppress chronic inflammation/reduce the risk of recurrences via immunomodulatory therapy (IMT).
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By what route are steroids administered?
Uveitis: Behçet

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By what route are steroids administered?
Systemic steroids are generally necessary (and are mandatory in cases of posterior involvement)
Uveitis: Behçet

**Treatment**

What are the two goals in the management of BD? _Broadly, how is each achieved?_
1) Control/resolve acute inflammation _with_ corticosteroids
2) Suppress chronic inflammation/reduce the risk of recurrences _via_ immunomodulatory therapy (IMT).

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Systemic steroids are generally necessary (and are mandatory in cases of posterior involvement)

_What IMT meds are useful in BD?_
**Uveitis: Behçet**

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

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What IMT meds are useful in BD?
A number have been studied. Antimetabolite (+ steroids) has been shown to be effective, and is probably the most widely accepted first-line treatment.
Uveitis: Behçet

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Uveitis: Behçet

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What advantage do the alkylating agents have over azathioprine?
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

Uveitis: Behçet

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Pts can usually be tapered off steroids; ie, they are effective as single-agent regimens
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What side effect of the alkylating agents is most feared?

They carry a significant risk of malignancy

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

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