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

Basics

In one word, what sort of condition is Behçet disease (BD)?
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Who is the typical BD pt?
Uveitis: *Behçet*

**Basics**

*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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What is/was the Silk Road?

Basics

Uveitis: Behçet

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

In general terms, people from which two portions of the Silk Road are at greatest risk?
People from its endpoints; ie, those of Eastern Mediterranean descent, and those of Far East descent

Which specific country has the highest rate of BD?
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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What is/was the Silk Road?
An ancient trading route extending from the eastern Mediterranean to the Far East

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The Middle East; Turkey; China; Korea; Japan
**Uveitis: Behçet**

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**Basics**

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*Who is the typical BD pt?*

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

*Is there a gender predilection?*
**Uveitis: Behçet**

**Basics**

*In one word, what sort of condition is Behçet disease (BD)?*
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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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What the classic nonocular complaint in BD?
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Are the oral ulcers painful?
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What is the classic rash?

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

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- Skin rash
- Genital ulcers
- Arthralgias

What is the classic rash?
Erythema nodosum

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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What does erythema nodosum look like?
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Is it painful?
Yes

Where is the classic location in BD (and other diseases)?
The pre-tibial region
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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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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?

Basics

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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?
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Of these, which is considered to be the most ominous?
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Of these, which is considered to be the most ominous?
CNS (aka ‘Neuro-BD’) carries a significant mortality risk
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Is uveitis usually an early manifestation of BD?
**Uveitis: Behçet**

**Basics**

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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?
Uveitis: Behçet

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

*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)?
A vasculitis

Who is the typical BD pt?
An individual of ‘Silk Road’ descent age 25-35

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

In addition to uveitis and aphthous ulcers, what three other findings are particularly common?
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 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 [ ]% of ocular BD cases
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Most definitely. Severe vision loss results in as many as 25% of ocular BD cases

What is the cause?
**Behçet**

**Basics**

*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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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
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

Intermediate

Posterior

Panuveitis

Behçet

BD uveitis can present in any form...
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: \textit{Anterior}

- Key distinction
  - \textit{not uni- vs bilateral}
Uveitis: **Anterior**

- Granulomatous
- Nongranulomatous

**Key distinction**

*not uni- vs bilateral*

- 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

1) The uveitis is profiled
2) The profiled case is meshed
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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
  - Acute
    - Unilateral
  - Chronic
    - Bilateral
Uveitis: **Anterior**

- Granulomatous
- Nongranulomatous
  - Acute
    - Unilateral
  - Chronic
    - Bilateral

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

**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**
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.
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
**Uveitis: Anterior**

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

**Nongranulomatous**

**Acute**
- Unilateral
  - HLA-B27 dz
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- 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? 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**
- 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?*
Roughly 25%

*The hypopyon in BD has a fairly unique property--what is it?*
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Uveitis: **Anterior**

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

**Nongranulomatous**

**Acute**
- Unilateral
  - HLA-B27 dz
  - Posner-Schlossman
  - Sarcoid
  - Syphilis
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  - Behçet
  - Drug rxn
  - Leptospirosis
  - Sarcoid

**Chronic**
- JIA
- FHI
- IBD/PA
- Behçet
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---

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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?*
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
Uveitis: **Anterior**

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

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

Chronic
- JIA
- FHI
- IBD/PA
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What percent of cases develop hypopyon?
Roughly 25

What word is often used to characterize the onset of BD anterior uveitis?
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…
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

...but posterior uveitis is a hallmark of BD.

Anterior → Intermediate → Posterior → Panuveitis

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

Chorioretinitis or Retinochoroiditis

Choroiditis

Retinitis

Neuroretinitis

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

Does it affect the arterioles, venules, or both?
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

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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
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?
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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

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

Can the optic nerve be involved?
Uveitis: \textit{Posterior}

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2) The profiled case is meshed
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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 \% of ocular BD pts
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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 25% of ocular BD pts
Uveitis

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

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.
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?*
Uveitis: \textit{Behçet}

\textbf{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?
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What is the HLA association for BD?
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So, is HLA testing helpful in diagnosing ocular BD?
The BCSC Uveitis book contend it is “of little value”
Uveitis: Behçet

**Diagnosis**

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

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

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

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Is this something BD pts experience?
About 40% do

Is the pathergy response pathognomonic 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?*
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How is the diagnosis of BD made?
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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

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!
Uveitis: Behçet

Treatment

What are the two goals in the management of BD?
1)
2)
Uveitis: Behçet

What are the two goals in the management of BD?
1) Control/resolve acute inflammation
2) Suppress chronic inflammation/reduce the risk of recurrences
Uveitis: Behçet

What are the two goals in the management of BD? Broadly, how is each achieved?
1) Control/resolve acute inflammation with class of med.
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
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 a different class of meds.

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

**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).
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).

By what route are steroids administered?
**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).

*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).

*By what route are steroids administered?*

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

*What IMT meds are useful in BD?*
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).

*By what route are steroids administered?*
Systemic steroids are generally necessary (and are mandatory in cases of posterior involvement).

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

*By what route are steroids administered?*
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Uveitis: Behçet

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