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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

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

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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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Is there a gender predilection?
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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Behçet, multiple oral aphthae
Behçet, single oral aphthus
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Are the oral ulcers painful?
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What is the classic rash?
Erythema nodosum
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What does erythema nodosum look like?
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Skin rash

What is the classic rash?
Erythema nodosum

What does erythema nodosum look like?
Reddish nodules

Basics

Uveitis: Behçet

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Is it painful?
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Skin rash

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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**Skin rash**

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)?
The pre-tibial region
Erythema nodosum in Behçet
In one word, what sort of condition is Behçet disease (BD)?
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What do the genital ulcers look like?
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Skin rash, genital ulcers, and arthralgias

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

Uveitis: Behçet

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
5) Treatment appropriate for the etiology is initiated
Behçet: Ulcer
In one word, what sort of condition is Behçet disease (BD)?
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Like the oral ulcers--small, with raised margins

Are they painful?
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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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What joint is classically affected in BD?
The knee
Uveitis: Behçet

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

Of these, which is considered to be the most ominous?
CNS (aka ‘Neuro-BD’) carries a significant mortality risk
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No; it usually follows the onset of disease by several years
**Uveitis: Behçet**

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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.

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 \( \% \)
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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Does BD pose a significant threat to long-term ocular health, and/or vision?
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What is the cause?
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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

Uveitis

Anterior

Posterior

Intermediate

Behçet

Panuveitis

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

Anterior

Posterior

Intermediate

Panuveitis

...including as an anterior uveitis.

Behçet
Uveitis: **Anterior**

- 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*

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**

- Granulomatous
- Nongranulomatous

**Key distinction**

- (not uni- vs bilateral)
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
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

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

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

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

- Nongranulomatosus:
  - 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
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
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    - Bilateral
      - TINU
      - *Behçet*
      - Drug rxn
      - Leptospirosis
      - Sarcoid
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    - 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%

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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
    - Bilateral
      - TINU
      - Behçet
  - Chronic

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

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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?
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.

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...
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.
Uveitis: **Posterior**

- Choroiditis
- Chorioretinitis or Retinochoroiditis
- Retinitis
- Neuroretinitis

What is the classic posterior manifestation of BD?
What is the classic posterior manifestation of BD?
A retinal vasculitis
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
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?
Uveitis: \textit{Posterior}

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

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\textit{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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Is it necrotizing, or non-necrotizing?
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Can BD retinal vasculitis lead to retinal ischemia with subsequent neovascularization?
What is the classic posterior manifestation of BD?
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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

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Can it affect the main arterioles and venules, leading to BRAOs and/or BRVOs?  
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Can the optic nerve be involved?
What is the classic posterior manifestation of BD?
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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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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
BD: Papillitis
Uveitis

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5) Treatment appropriate for the etiology is initiated

Anterior

Intermediate

Posterior

Panuveitis

And of course, BD can present as a panuveitis.

Behçet
Uveitis: *Behçet*

**Diagnosis**

*How is the diagnosis of BD made?*

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

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: *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**

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

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

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

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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?
Uveitis: **Behçet**

**Diagnosis**

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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.
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

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

**Diagnosis**

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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?**
About 40% do
Uveitis: **Behçet**

**Diagnosis**

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It is a clinical diagnosis--there are no specific tests for it

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About 40% do

*Is the pathergy response pathognomonic for BD?*
How is the diagnosis of BD made?
It is a clinical diagnosis--there are no specific tests for it

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No
Uveitis: **Behçet**

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So, is cutaneous pathergy testing useful in diagnosing BD?
Uveitis: Behçet

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

**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
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*
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
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 different class of meds.
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).
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?*
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 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. Antimetabolite (+ 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?*
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. Azathioprine (+ 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).

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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. Azathioprine (+ steroids) has been shown to be effective, and is probably the most widely accepted first-line treatment. alkylating agent 1 and alkylating agent 2 are effective as well.
Uveitis: **Behçet**

**Treatment**

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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