Retinal Vasculitis

Per Stephen Foster MD: When confronted with a retinal vasculitis, the first step is to determine to which of these categories it belongs.
Per Stephen Foster MD: When confronted with a retinal vasculitis, the first step is to determine to which of these categories it belongs.
Retinal Vasculitis

Retinal Vasculitis

- Primarily Arterial
- Both Arterial and Venous
- Primarily Venous

(Note: The BCSC Retina book asserts that simultaneous involvement of both retinal arterioles and venules is the rule, and that isolated disease of either is distinctly uncommon)
Retinal Vasculitis

Primarily Arterial:
Think of these 3 diseases:

Both Arterial and Venous

Primarily Venous
Retinal Vasculitis

Primarily Arterial: Think of these 3 diseases:
- SLE (systemic lupus erythematosus)
- PAN (polyarteritis nodosa)
- Churg-Strauss

Both Arterial and Venous

Primarily Venous
Retinal Vasculitis

Primarily Arterial:
Think of these 2 diseases:

SLE
PAN
Churg-Strauss

Both Arterial and Venous

Primarily Venous

Neither the *Retina* nor *Uveitis* book addresses Churg-Strauss, so with all due respect to Dr. Foster, we will not consider it further. Just file it away as a very rare cause of retinal arteritis.
Retinal Vasculitis

Primarily Arterial:
Think of these 2 diseases:
- SLE
- PAN

Both Arterial and Venous

Primarily Venous:
Think of these 3 diseases:
- ?
- ?
- ?
Retinal Vasculitis

Primarily Arterial:
Think of these 2 diseases:
- SLE
- PAN

Both Arterial and Venous

Primarily Venous:
Think of these 3 diseases:
- Sarcoid
- Multiple sclerosis
- Birdshot
Retinal Vasculitis

Primarily *Arterial*: Think of these 2 diseases:
- SLE
- PAN

Both *Arterial* and *Venous*:

Primarily *Venous*: Think of these 2 diseases:
- Sarcoid
- Multiple sclerosis
- Birdshot

Birdshot *is addressed in detail in the set entitled* White Dot Syndromes
Retinal Vasculitis

Primarily **Arterial**: Think of these 2 diseases:
- SLE
- PAN

Both **Arterial** and **Venous**: Think of these 3 diseases:
- ?
- ?
- ?

Primarily **Venous**: Think of these 2 diseases:
- Sarcoid
- Multiple sclerosis
Retinal Vasculitis

- Primarily Arterial: Think of these 2 diseases:
  - SLE
  - PAN

- Both Arterial and Venous: Think of these 3 diseases:
  - Behçet’s
  - Wegener’s
  - Toxoplasmosis

- Primarily Venous: Think of these 2 diseases:
  - Sarcoid
  - Multiple sclerosis
Retinal Vasculitis

Primarily Arterial: Think of these 2 diseases:
- SLE
- PAN

Both Arterial and Venous: Think of these 2 diseases:
- Behçet’s
- Wegener’s
- Toxoplasmosis

Primarily Venous: Think of these 2 diseases:
- Sarcoid
- Multiple sclerosis

Toxoplasmosis is addressed in detail in its own slide-set
Retinal Vasculitis

Primarily Arterial:
Think of these 2 diseases:
- SLE
- PAN

Both Arterial and Venous:
Think of these 2 diseases:
- Behçet’s
- Wegener’s

Primarily Venous:
Think of these 2 diseases:
- Sarcoid
- Multiple sclerosis

The term ‘Wegener’s granulomatosis’ has fallen out of favor. What term is preferred in its place?
Retinal Vasculitis

Primarily Arterial: Think of these 2 diseases:
- SLE
- PAN

Both Arterial and Venous: Think of these 2 diseases:
- Behçet’s
- Granulomatosis with polyangiitis

Primarily Venous: Think of these 2 diseases:
- Sarcoid
- Multiple sclerosis

The term ‘Wegener’s granulomatosis’ has fallen out of favor.
What term is preferred in its place?
‘Granulomatosis with polyangiitis’
Retinal Vasculitis

Primarily Arterial:
Think of these 2 diseases:
- SLE
- PAN

Both Arterial and Venous:
Think of these 2 diseases:
- Behçet’s
- Granulomatosis with polyangiitis

Primarily Venous:
Think of these 2 diseases:
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The term ‘Wegener's granulomatosis’ has fallen out of favor.

What is the preferred name?

Why is the name ‘Wegener’s granulomatosis’ no longer preferred?
Retinal Vasculitis

Primarily Arterial:
Think of these 2 diseases:
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- Granulomatosis with polyangiitis

Primarily Venous:
Think of these 2 diseases:
- Sarcoid
- Multiple sclerosis

The term ‘Wegener’s granulomatosis’ has fallen out of favor.

What is ‘Wegener’s granulomatosis’ no longer preferred? Because Dr. Wegener was a Nazi
Primarily Venous:
Think of these 3 diseases:
- Sarcoid
- Multiple sclerosis
- Retinal Vasculitis

Primarily Arterial:
Think of these 3 diseases:
- PAN
- SLE
- MS

Both Arterial and Venous:
Think of these 3 diseases:
- Behçet's
- MS
- Wegener's

Who is the classic lupus pt?
- A woman of childbearing age
- Is there a racial predilection?
  - Yes, black and Hispanic women are at greater risk than are white women
 What proportion of lupus pts manifest retinal findings?
- About 5-10%
Why is lupus-associated retinal arteritis such an ominous finding?
- It heralds CNS and/or renal involvement with the disease, and carries a high risk of mortality
Given its dire systemic significance, how should lupus-associated retinal arteritis be managed?
- With the big dogs: Plasmapheresis + IV cyclophosphamide acutely

Retinal Vasculitis
Primarily Venous: Think of these 3 diseases:
- Sarcoid
- Multiple sclerosis
- Retinal Vasculitis

Primarily Arterial: Think of these 3 diseases:
- PAN
- SLE
- MS (multiple sclerosis)

Both Arterial and Venous: Think of these 3 diseases:
- Wegener’s
- Behçet’s
- MS (multiple sclerosis)

Who is the classic lupus pt?
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Multiple sclerosis
Retinal Vasculitis

Primarily Arterial:
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PAN
SLE
MS (multiple sclerosis)

Both Arterial and Venous:
Think of these 3 diseases:
Behçet's
Wegener's

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Sarcoid
Multiple sclerosis
Retinal Vasculitis

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Think of these 3 diseases:
PAN
SLE
Wegener's

Both Arterial and Venous:
Think of these 3 diseases:
Behçet's
MS
MS (multiple sclerosis)

Who is the classic lupus pt?
A woman of childbearing age

Is there a racial predilection?
Yes, and women are at greater risk than are women

About 5-10% of lupus pts manifest retinal findings.

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Given its dire systemic significance, how should lupus-associated retinal arteritis be managed?
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Lupus is a multisystem dz with protean manifestations. Which two are most common, found in up to 80% of pts?

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Primarily Venous:
Think of these 3 diseases:
- Sarcoidosis
- Multiple sclerosis
- Retinal Vasculitis

Primarily Arterial:
Think of these 3 diseases:
- PAN
- SLE
- MS

Both Arterial and Venous:
Think of these 3 diseases:
- Behçet's
- Wegener's
- MS

Who is the classic lupus pt?
A woman of childbearing age

Lupus is a multisystem dz with protean manifestations. Which two are most common, found in up to 80% of pts?
-- Skin lesions (eg, the classic malar rash)
-- Arthralgias

What proportion of lupus pts manifest retinal findings?
About 5-10%

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Lupus is a multisystem dz with protean manifestations. Which two are most common, found in up to 80% of pts?
-- Skin lesions (eg, the classic malar rash)
-- Arthralgias

What proportion of pts will manifest ophthalmic findings?
About half

Lupus' ophthalmic manifestations are protean. Which two are most common?
-- Lid-skin involvement in a rash
-- Sjögren syndrome
Retinal Vasculitis

SLE: Malar rash
**Primarily Venous:**
Think of these 3 diseases:
- Sarcoidosis
- Multiple sclerosis
- Retinal Vasculitis

**Primarily Arterial:**
Think of these 3 diseases:
- PAN
- SLE
- Wegener's

**Both Arterial and Venous:**
Think of these 3 diseases:
- Behçet's
- MS
- MS (multiple sclerosis)

Who is the classic lupus pt?
A woman of childbearing age

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Primarily Venous:
Think of these 3 diseases:
- Sarcoid
- Multiple sclerosis
- Retinal Vasculitis

Primarily Arterial:
Think of these 3 diseases:
- PAN
- SLE
- MS (multiple sclerosis)

Both Arterial and Venous:
Think of these 3 diseases:
- Behçet's
- MS
- Wegener's

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Primarily Arterial: Think of these 3 diseases:
- PAN
- SLE
- Wegener's

Arterial and Venous: Think of these 3 diseases:
- Behçet's
- MS (multiple sclerosis)
- Lupus

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Multiple sclerosis
Retinal Vasculitis

Primarily Arterial:
Think of these 3 diseases:
PAN
SLE
MS
Wegener’s

Both Arterial and Venous:
Think of these 3 diseases:
Behçet’s
Multiple sclerosis
Wegener’s

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Multiple sclerosis
Retinal Vasculitis

Primarily Arterial:
Think of these 3 diseases:
PAN
SLE
MS (multiple sclerosis)

Both Arterial and Venous:
Think of these 3 diseases:
Behçet’s
MS
Wegener’s

Who is the classic lupus pt?
A woman of childbearing age

Is there a racial predilection?
Yes, black and Hispanic women are at greater risk than are white women

What proportion of lupus pts manifest retinal findings?
About 5-10%

Why is lupus-associated retinal arteritis such an ominous finding?
It heralds CNS and/or renal involvement with the disease, and carries a high risk of mortality

Given its dire systemic significance, how should lupus-associated retinal arteritis be managed?
With the big dogs: Plasmapheresis + IV cyclophosphamide acutely
Who is the classic lupus pt?
A woman of childbearing age

Is there a racial predilection?
Yes, black and Hispanic women are at greater risk than are white women

What proportion of lupus pts manifest retinal findings?
About 3-30%
Retinal Vasculitis

Who is the classic lupus pt?
A woman of childbearing age

Is there a racial predilection?
Yes, black and Hispanic women are at greater risk than are white women

What proportion of lupus pts manifest retinal findings?
About 3-30%

This range represents a compromise between inconsistencies in the BCSC books:
--Rate of lupus retinopathy per the Retina book: 3-10%
--Rate per the Uveitis book: 3-29%
(I rounded to 30 to make it easier to remember)
Primarily Venous:
Think of these 3 diseases:
- Sarcoid
- Multiple sclerosis
- Retinal Vasculitis

Primarily Arterial:
Think of these 3 diseases:
- PAN
- SLE
- MS

Both Arterial and Venous:
Think of these 3 diseases:
- Behçet’s
- MS
- Wegener’s

Who is the classic lupus pt?
A woman of childbearing age

Is there a racial predilection?
Yes, black and Hispanic women are at greater risk than are white women

What proportion of lupus pts manifest retinal findings?
About 3-30%

What retinal findings may occur?
- Occlusive events: Asymptomatic cotton wool spots if 'lucky,' an infarcted macula if not

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Primarily Venous:
Think of these 3 diseases:
Sarcoid
Multiple sclerosis
Retinal Vasculitis

Primarily Arterial:
Think of these 3 diseases:
PAN
SLE
MS (multiple sclerosis)
Wegener's

Both Arterial and Venous:
Think of these 3 diseases:
Behçet's
MS
Wegener's

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**Retinal Vasculitis**

*Who is the classic lupus pt?*
A woman of childbearing age

*Is there a racial predilection?*
Yes, black and Hispanic women are at greater risk than are white women

*What proportion of lupus pts manifest retinal findings?*
About 3-30%

*CWS are the classic manifestation of lupus retinopathy!*

Occlusive events: Asymptomatic cotton wool spots if ‘lucky,’ an infarcted macula if not

*(No question—proceed when ready)*
Retinal Vasculitis

SLE: CWS
Who is the classic lupus pt?
A woman of childbearing age

Is there a racial predilection?
Yes, black and Hispanic women are at greater risk than are white women

What proportion of lupus pts manifest retinal findings?
About 3-30%

CWS are the classic manifestation of lupus retinopathy!

Occlusive events: Asymptomatic cotton wool spots if ‘lucky,’ an infarcted macula if not

Is macular infarction also a classic manifestation of lupus retinopathy?
**Primarily Venous:**
Think of these 3 diseases:
- Sarcoid
- Multiple sclerosis
- 2

**Primarily Arterial:**
Think of these 3 diseases:
- PAN
- SLE
- 2

**Both Arterial and Venous:**
Think of these 3 diseases:
- Behçet’s
- MS
- (multiple sclerosis)
- Wegener’s

Who is the classic lupus pt?
A woman of childbearing age

Is there a racial predilection?
Yes, black and Hispanic women are at greater risk than are white women

What proportion of lupus pts manifest retinal findings?
About 3-30%

CWS are the classic manifestation of lupus retinopathy!

Occlusive events: Asymptomatic cotton wool spots if ‘lucky,’ an infarcted macula if not

Is macular infarction also a classic manifestation of lupus retinopathy?
Thankfully no—it is quite rare
Retinal Vasculitis

SLE: Macular infarction
Primarily Venous:
Think of these 3 diseases:
- Sarcoid
- Multiple sclerosis
- Retinal Vasculitis

Primarily Arterial:
Think of these 3 diseases:
- PAN
- SLE
- Behçet's

Both Arterial and Venous:
Think of these 3 diseases:
- MS (multiple sclerosis)
- Wegener's
- Both

Who is the classic lupus pt?
A woman of childbearing age

Is there a racial predilection?
Yes, black and Hispanic women are at greater risk than are white women

What proportion of lupus pts manifest retinal findings?
About 3-30%

Why is lupus-associated retinal arteritis such an ominous finding?
It heralds CNS and/or renal involvement with the disease, and carries a high risk of mortality

Given its dire systemic significance, how should lupus-associated retinal arteritis be managed?
With the big dogs: Plasmapheresis + IV cyclophosphamide acutely

What retinal findings may occur?
Occlusive events: Asymptomatic cotton wool spots
if ‘lucky,’ an infarcted macula if not

Is macular infarction also a classic manifestation of lupus retinopathy?
Thankfully no—it is quite rare

CWS are the classic manifestation of lupus retinopathy!
Retinal Vasculitis

Primary Arterial:
Think of these 3 diseases:
PAN
SLE
Both Arterial and Venous:
Think of these 3 diseases:
Primarily Venous:
Think of these 3 diseases:
Sarcoid
Toxoplasmosis
Birdshot Behçet's
MS (multiple sclerosis)
Wegener's

Who is the classic lupus pt?
A woman of childbearing age

Is macular infarction also a classic manifestation of lupus retinopathy?
Thankfully no—it is quite rare

What is the fundamental difference between the pathophysiology of CWS and macular infarction?
It’s the size of the vessel involved. CWS result from infarction of tiny end-arterioles of the arterial tree, whereas macular infarction involves the relatively ginormous central retinal artery, or an immediate branch thereof.

CWS are the classic manifestation of lupus retinopathy!

Occlusive events: Asymptomatic cotton wool spots
if ‘lucky,’ an infarcted macula

Why is lupus-associated retinal arteritis such an ominous finding?
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Retinal Vasculitis

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It’s the size of the vessel involved. CWS result from infarction of tiny end-arterioles of the arterial tree, whereas macular infarction involves the relatively ginormous central retinal artery, or an immediate branch thereof.

How does this explain the fact that CWS are vastly more common than macular infarction?

CWS are the classic manifestation of lupus retinopathy!

Occlusive events: Asymptomatic if ‘lucky,’ an infarcted macula if not.

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Thankfully no—it is quite rare.
Retinal Vasculitis

Primarily Arterial:
Think of these 3 diseases:
- PAN
- SLE
- Behçet's

Both Arterial and Venous:
Think of these 3 diseases:
- Sarcoid
- Toxoplasmosis
- MS
- Wegener's

Primarily Venous:
Think of these 3 diseases:
- Birdshot
- Lupus

Who is the classic lupus pt?
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Yes, black and Hispanic women are at greater risk than are white women

What proportion of lupus pts manifest retinal findings?
About 3-30%

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How does this explain the fact that CWS are vastly more common than macular infarction? It's because lupus is a microvasculopathy, and thus much more likely to affect end-arterioles of the arterial tree.

How does this explain the fact that CWS are vastly more common than macular infarction? It's because lupus is a microvasculopathy, and thus much more likely to affect end-arterioles of the arterial tree.
**Retinal Vasculitis**

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**What proportion of lupus pts manifest retinal findings?**
About 3-30%

**Why is lupus-associated retinal arteritis an ominous finding?**

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- Multiple sclerosis
- Retinal Vasculitis

Primarily Arterial:
Think of these 3 diseases:
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- SLE
- Wegener’s

Both Arterial and Venous:
Think of these 3 diseases:
- Behçet’s
- MS
- MS (multiple sclerosis)

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- PAN
- SLE
- Multiple sclerosis

Both Arterial and Venous: Think of these 3 diseases:
- Behçet’s
- MS
- Wegener’s

In a nutshell, what is the pathophysiology of PAN?

Subacute episodes of focal necrotizing inflammation of arteries

Is it a common, or uncommon condition?

Uncommon

Who is the typical PAN pt?

A male between 40 and 60 years old

Is there a racial predilection?

No

PAN is strongly associated with seropositivity for what virus?

Hepatitis B
Primarily Venous:
Think of these 3 diseases:
Sarcoid
Multiple sclerosis
Retinal Vasculitis

Primarily Arterial:
Think of these 3 diseases:
PAN
SLE
MS (multiple sclerosis)

Both Arterial and Venous:
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PAN is strongly associated with seropositivity for what virus?
Hepatitis B
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Once considered a single entity, PAN is now subdivided into two conditions—what are they?

Classic PAN affects medium and small arteries, whereas MP affects smaller arteries.

Retinal Vasculitis
Primarily Venous:
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Sarcoid
Multiple sclerosis
Retinal Vasculitis

Primarily Arterial:
Think of these 3 diseases:
PAN
SLE
MS

Arterial and Venous:
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MS (multiple sclerosis)
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‘Classic’ PAN, and microscopic polyangiitis (MP)

Retinal Vasculitis

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Primarily Venous:
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Multiple sclerosis
Vasculitis
Primarily Arterial:
Think of these 3 diseases:
PAN
SLE
2
Both Arterial and Venous:
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‘Classic’ PAN, and microscopic polyangiitis (MP)

What is the difference between the two?
Classic PAN affects medium and small arteries, whereas MP affects smaller arteries
Primarily Venous: Think of these 3 diseases:
- Sarcoid
- Multiple sclerosis
- Retinal Vasculitis

Primarily Arterial: Think of these 3 diseases:
- PAN
- SLE
- MS

Both Arterial and Venous: Think of these 3 diseases:
- Behçet’s
- MS
- Wegener’s

In a nutshell, what is the pathophysiology of PAN?
Subacute episodes of focal necrotizing inflammation of arteries

Once considered a single entity, PAN is now subdivided into two conditions—what are they? ‘Classic’ PAN, and microscopic polyangiitis (MP)

What is the difference between the two?
Classic PAN affects medium and small arteries, whereas MP affects smaller arteries

What’s the difference between a ‘small’ artery and a ‘smaller’ artery?
Primarily Venous:
Think of these 3 diseases:
- Sarcoid
- Multiple sclerosis
- Retinal Vasculitis

Primarily Arterial:
Think of these 3 diseases:
- PAN
- SLE
- (multiple sclerosis)

Both Arterial and Venous:
Think of these 3 diseases:
- Behçet’s
- MS
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Retinal Vasculitis

In a nutshell, what is the pathophysiology of PAN?
Subacute episodes of focal necrotizing inflammation of arteries

Once considered a single entity, PAN is now subdivided into two conditions—what are they?
'Classic' PAN, and microscopic polyangiitis (MP)

What is the difference between the two?
Classic PAN affects medium and small arteries, whereas MP affects smaller arteries

What’s the difference between a ‘small’ artery and a ‘smaller’ artery?
Rule of thumb: Classic PAN only affects arteries large enough to be named, whereas MP only affects vessels smaller than that
Primarily Venous:
Think of these 3 diseases:
- Sarcoid
- Multiple sclerosis
- Retinal Vasculitis

Primarily Arterial:
Think of these 3 diseases:
- PAN
- SLE
- MS (multiple sclerosis)

Both Arterial and Venous:
Think of these 3 diseases:
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- MS
- Wegener's

Retinal Vasculitis

In a nutshell, what is the pathophysiology of PAN?
Subacute episodes of focal necrotizing inflammation of arteries

Is it a common, or uncommon condition?

Uncommon

Who is the typical PAN pt?
A male between 40 and 60 years old

Is there a racial predilection?
No

PAN is strongly associated with seropositivity for what virus?
Hepatitis B
Primarily Venous: Think of these 3 diseases: Sarcoid, Multiple sclerosis, Retinal Vasculitis

Primarily Arterial: Think of these 3 diseases: PAN, SLE, Behçet's

Both Arterial and Venous: Think of these 3 diseases: MS, Wegener's, MS

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Think of these 3 diseases:
PAN
SLE
MS (multiple sclerosis)

Both Arterial and Venous:
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Think of these 3 diseases:
- PAN
- SLE
- Behçet’s

Both Arterial and Venous:
Think of these 3 diseases:
- MS
- Wegener’s
- Multiple sclerosis

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Primarily Arterial: Think of these 3 diseases: PAN, SLE, Behçet’s

Both Arterial and Venous: Think of these 3 diseases: MS, Wegener’s, In a nutshell, what is the pathophysiology of PAN?
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- 2

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- PAN
- SLE
- 2

Both Arterial and Venous:
Think of these 3 diseases:
- Behçet's
- MS
- (multiple sclerosis)
- Wegener’s

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Retinal Vasculitis

Primarily Arterial:
Think of these 3 diseases:
- PAN
- SLE
- 2

Both Arterial and Venous:
Think of these 3 diseases:
- Behçet's
- MS (multiple sclerosis)
- Wegener's

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In a nutshell, what is the pathophysiology of PAN?
Subacute episodes of focal necrotizing inflammation of arteries

What is the typical presentation of PAN? (Note: It’s not ocular.)
Constitutional symptoms: Fever, fatigue, weight loss. Renal vasculitis resulting in secondary HTN is common.

Is there a racial predilection?
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SLE
MS (multiple sclerosis)

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What proportion of PAN pts develop ocular involvement?
About 20%

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What is the 5 year survival rate for untreated PAN?
A miserable 10%

What is the 5 year survival rate of treated PAN?
A robust 80%

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SLE
Behçet’s

Both Arterial and Venous:
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MS (multiple sclerosis)
Wegener’s

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What proportion of PAN pts develop ocular involvement?
About 20%

What is the typical ocular presentation of PAN? (Head's up: It's probably not what you think.)
Peripheral ulcerative keratitis (PUK). It can cause scleritis (both anterior and posterior) as well.
Primarily Venous: Think of these 3 diseases:
- Sarcoid
- Multiple sclerosis
- Retinal Vasculitis

Primarily Arterial: Think of these 3 diseases:
- PAN
- SLE
- MS (multiple sclerosis)

Both Arterial and Venous: Think of these 3 diseases:
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- MS
- Wegener's

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Peripheral ulcerative keratitis (PUK). It can cause scleritis (both anterior and posterior) as well.

Retinal Vasculitis
Retinal Vasculitis

PUK (full disclosure: I don’t think it’s 2ndry to PAN)
Retinal Vasculitis

Scleritis (same disclosure)
Primarily Venous:
Think of these 3 diseases:
Sarcoid
Multiple sclerosis
Vasculitis

Primarily Arterial:
Think of these 3 diseases:
PAN
SLE
MS
Wegener’s

Both Arterial and Venous:
Think of these 3 diseases:
Behçet’s
MS
Wegener’s

In a nutshell, what is the pathophysiology of PAN?
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What proportion of PAN pts develop ocular involvement?
About 20%

What is the typical ocular presentation of PAN? (Head’s up: It’s probably not what you think.)
Peripheral ulcerative keratitis (PUK). It can cause scleritis (both anterior and posterior) as well.

OK fine. What is the typical posterior-pole presentation of PAN?
Primarily Venous: Think of these 3 diseases: Sarcoidosis, Multiple sclerosis, Vasculitis

Primarily Arterial: Think of these 3 diseases: PAN, SLE, Wegener’s

Both Arterial and Venous: Think of these 3 diseases: Behçet’s, MS, Wegener’s

In a nutshell, what is the pathophysiology of PAN?
Subacute episodes of focal necrotizing inflammation of arteries

What is the typical presentation of PAN? (Note: It’s not ocular.)
Constitutional symptoms: Fever, fatigue, weight loss. Renal vasculitis resulting in secondary HTN is common.

What proportion of PAN pts develop ocular involvement?
About 20%

What is the typical ocular presentation of PAN? (Head’s up: It’s probably not what you think.)
Peripheral ulcerative keratitis (PUK). It can cause scleritis (both anterior and posterior) as well.

OK fine. What is the typical posterior-pole presentation of PAN?
In a manner consistent with severe HTN
Primarily Venous:
Think of these 3 diseases:
Sarcoid
Multiple sclerosis
Retinal Vasculitis

Primarily Arterial:
Think of these 3 diseases:
PAN
SLE
MS (multiple sclerosis)

Both Arterial and Venous:
Think of these 3 diseases:
Behçet’s
MS
Wegener’s

In a nutshell, what is the pathophysiology of PAN?
Subacute episodes of focal necrotizing inflammation of arteries

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What proportion of PAN pts develop ocular involvement?
About 20%

Is there a racial predilection?
No

PAN is strongly associated with seropositivity for what virus?
Hepatitis B

What is the typical ocular presentation of PAN? (Head’s up: It’s probably not what you think.)
Peripheral ulcerative keratitis (PUK). It can cause scleritis (both anterior and posterior)
OK fine. What is the typical posterior pole presentation of PAN?
In a manner consistent with severe HTN
What process mediates damage caused by acute, severe HTN?
Acute, severe HTN

What process mediates damage caused by acute, severe HTN?

Vasospasm of arterioles

*Retinal Vasculitis*
What process mediates damage caused by acute, severe HTN?

Vasospasm of arterioles

What categories of pathology can result?

?  ?  ?
Acute, severe HTN

What process mediates damage caused by acute, severe HTN?

Vasospasm of arterioles

What categories of pathology can result?

Retinopathy  Choroidopathy  Optic Neuropathy
Acute, severe HTN

What process mediates damage caused by acute, severe HTN?

Vasospasm of arterioles

What categories of pathology can result?

- Retinopathy
- Choroidopathy
- Optic Neuropathy
Acute, severe HTN

What process mediates damage caused by acute, severe HTN?

Vasospasm of arterioles

What categories of pathology can result?

Retinopathy
- Cotton-wool spots
- Retinal hemorrhages
- Macular star

Choroidopathy

Optic Neuropathy
Retinal Vasculitis

HTN retinopathy
Acute, severe HTN

What process mediates damage caused by acute, severe HTN?

Vasospasm of arterioles

What categories of pathology can result?

Retinopathy
- Retinal hemorrhages
- Cotton-wool spots
- Macular star

Choroidopathy
- Leads to…
  - ?

Optic Neuropathy
- Leads to…
  - ?
Acute, severe HTN

What process mediates damage caused by acute, severe HTN?

Vasospasm of arterioles

What categories of pathology can result?

Retinopathy
- Cotton-wool spots
- Retinal hemorrhages
- Macular star

Choroidopathy
- Leads to...
  - Choroidal infarction
    - Leads to...
      - Exudative RD

Optic Neuropathy
Retinal Vasculitis

HTN choroidopathy with exudative RD
Acute, severe HTN

What process mediates damage caused by acute, severe HTN?

Vasospasm of arterioles

What categories of pathology can result?

Retinopathy
- Cotton-wool spots
- Retinal hemorrhages
- Macular star

Choroidopathy
- Leads to...
- Choroidal infarction
- Leads to...
- Exudative RD

Optic Neuropathy
- Leads to...
- ?
Acute, severe HTN

What process mediates damage caused by acute, severe HTN?

Vasospasm of arterioles

What categories of pathology can result?

Retinopathy
- Cotton-wool spots
- Retinal hemorrhages
- Macular star

Choroidopathy
- Leads to...\[Choroidal infarction\]

Optic Neuropathy
- Leads to...\[Disc edema, hemorrhages\]

- Exudative RD
Retinal Vasculitis

HTN optic neuropathy
Retinal Vasculitis

Who is the typical Bechet’s disease (BD) pt?

- Behçet’s
- Granulomatosis
- SLE
- PAN
- Sarcoid
- Multiple sclerosis

- PAN
- SLE
- Granulomatosis

- Both Arterial and Venous:
- Primarily Arterial: Think of these 2 diseases:
- Venous:
- Primarily Venous: Think of these 2 diseases:
Retinal Vasculitis

Who is the typical Bechet’s disease (BD) pt?
An individual of ‘Silk Road’ descent age 25-35
Retinal Vasculitis

Primarily Arterial:
Think of these 2 diseases:
- SLE
- PAN

Both Arterial and Venous:
Think of these 2 diseases:
- Behçet's
- Granulomatosis

Primarily Venous:
Think of these 2 diseases:
- Sarcoid
- Multiple sclerosis

Who is the typical Behçet's disease (BD) pt?
An individual of ‘Silk Road’ descent age 25-35

What is/was the Silk Road?
Retinal Vasculitis

- Primarily Arterial: Think of these 2 diseases:
  - SLE
  - PAN

- Both Arterial and Venous: Think of these 2 diseases:
  - Behçet's
  - Granulomatosis

- Primarily Venous: Think of these 2 diseases:
  - Sarcoid
  - Multiple sclerosis

Who is the typical Behçet's disease (BD) pt?
An individual of ‘Silk Road’ descent age 25-35

What is/was the Silk Road?
An ancient trading route extending from the eastern Mediterranean to the Far East
Retinal Vasculitis
Retinal Vasculitis

Primarily Arterial: Think of these 2 diseases:
- SLE
- PAN

Both Arterial and Venous: Think of these 2 diseases:
- Behçet’s
- Granulomatosis

Primarily Venous: Think of these 2 diseases:
- Sarcoid
- Multiple sclerosis

Who is the typical Behçet’s disease (BD) pt?
An individual of ‘Silk Road’ descent age 25-35

What is/was the Silk Road?
An ancient trading route extending from the eastern Mediterranean to the Far East

What regions/countries are found along the Silk Road?
Retinal Vasculitis

Primarily Arterial:
Think of these 2 diseases:
- SLE
- PAN

Both Arterial and Venous:
Think of these 2 diseases:
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- Granulomatosis

Primarily Venous:
Think of these 2 diseases:
- Sarcoid
- Multiple sclerosis

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

What is/was the Silk Road?
An ancient trading route extending from the eastern Mediterranean to the Far East

What regions/countries are found along the Silk Road?
The Middle East; Turkey; China; Korea; Japan
Retinal Vasculitis

- Primarily Arterial: Think of these 2 diseases:
  - SLE
  - PAN

- Primarily Venous: Think of these 2 diseases:
  - Behçet’s
  - Granulomatosis

- Both Arterial and Venous: Think of these 2 diseases:
  - PAN
  - SLE

Who is the typical Behçet’s disease (BD) pt?
An individual of ‘Silk Road’ descent age 25-35

What is/was the Silk Road?

Which of these areas has the highest rate of BD?

The Middle East; Turkey; China; Korea; Japan

What regions/countries are found along the Silk Road?
Retinal Vasculitis

Primarily Arterial: Think of these 2 diseases:
- SLE
- PAN

Both Arterial and Venous: Think of these 2 diseases:
- Behçet’s
- Granulomatosis

Primarily Venous: Think of these 2 diseases:
- Sarcoid
- Multiple sclerosis

Who is the typical Behçet’s disease (BD) pt?
An individual of ‘Silk Road’ descent age 25-35

What is/was the Silk Road?

Which of these areas has the highest rate of BD?
Turkey

What regions/countries are found along the Silk Road?
The Middle East, Turkey, China, Korea, Japan
Retinal Vasculitis

Both *Arterial* and *Venous*:
Think of these 2 diseases:
- Behçet's
- Granulomatosis

Who is the typical Bechet’s disease (BD) pt?
An individual of ‘Silk Road’ descent age 25-35

What the classic nonocular complaint in BD?
Retinal Vasculitis

Primarily Arterial:
Think of these 2 diseases:
- SLE
- PAN

Both Arterial and Venous:
Think of these 2 diseases:
- Behçet’s
- Wegener’s

Primarily Venous:
Think of these 2 diseases:
- Granulomatosis
- Sarcoid
- Multiple sclerosis

Who is the typical Bechet’s disease (BD) pt?
An individual of ‘Silk Road’ descent age 25-35

What the classic nonocular complaint in BD?
Recurrent oral ulcers
Retinal Vasculitis

Behçet: Oral aphthae
Retinal Vasculitis

Primarily Arterial: Think of these diseases:
- SLE
- PAN

Both Arterial and Venous: Think of these diseases:
- Behçet’s
- Lupus
- Wegener

Primarily Venous: Think of these diseases:
- Sarcoid
- Multiple sclerosis

Who is the typical Bechet’s disease (BD) pt?
An individual of ‘Silk Road’ descent age 25-35

What the classic nonocular complaint in BD?
Recurrent oral ulcers

Are the oral ulcers painful?
Retinal Vasculitis

Primarily Arterial:
Think of these 2 diseases:
- SLE
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Who is the typical Behçet’s disease (BD) pt?
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What the classic nonocular complaint in BD?
Recurrence oral ulcers

Are the oral ulcers painful?
Yes, very
Retinal Vasculitis

Primarily Arterial:
Think of these 2 diseases:

SLE
PAN

Behçet’s

Both Arterial and Venous:
Think of these 2 diseases:

PAN
SLE

Wegener’s

Primarily Venous:
Think of these 2 diseases:

Behçet’s

Granulomatosis

Sarcoid
Multiplesclerosis

Who is the typical Bechet’s disease (BD) pt?
An individual of ‘Silk Road’ descent age 25-35

What the classic nonocular complaint in BD?
Recurrent oral ulcers

In addition to oral ulcers, what three other nonocular findings are common?

--
--
--
Retinal Vasculitis

Primarily Arterial:
Think of these 2 diseases:
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Both Arterial and Venous:
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What the classic nonocular complaint in BD?
Recurrent oral ulcers

In addition to oral ulcers, what three other nonocular findings are common?
--Skin rash
--Genital ulcers
--Arthralgias
Retinal Vasculitis

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  - SLE
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What the classic nonocular complaint in BD?
Recurrent oral ulcers

In addition to oral ulcers, what three other nonocular findings are common?
- Skin rash
- Genital ulcers
- Arthralgias

What is the classic rash?
Erythema nodosum
The pre-tibial region
Retinal Vasculitis

Primarily Arterial:
Think of these 2 diseases:
- SLE
- PAN

Both Arterial and Venous:
Think of these 2 diseases:
- Behçet's
- Granulomatosis

Primarily Venous:
Think of these 2 diseases:
- Sarcoid
- Multiple sclerosis

Who is the typical Bechet’s disease (BD) pt?
An individual of ‘Silk Road’ descent age 25-35

What the classic nonocular complaint in BD?
Recurrent oral ulcers

In addition to nonocular findings are common?

What is the classic rash?
Erythema nodosum

- Genital ulcers
- Arthralgias
**Retinal Vasculitis**

- **Primarily Arterial:**
  Think of these 2 diseases:
  - SLE
  - PAN

- **Both Arterial and Venous:**
  Think of these 2 diseases:
  - Behçet’s
  - Granulomatosis

- **Primarily Venous:**
  Think of these 2 diseases:
  - Sarcoid
  - Multiple sclerosis

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**Who is the typical Behçet’s disease (BD) pt?**
An individual of ‘Silk Road’ descent age 25-35

**What the classic nonocular complaint in BD?**
Recurrent oral ulcers

**In addition to the ocular symptoms, what nonocular findings are common?**

- **Skin rash**
  - Erythema nodosum
- Genital ulcers
- Arthralgias

**What is the classic rash?**

**Where is the classic location?**
Retinal Vasculitis

Primarily Arterial: Think of these 2 diseases:
- SLE
- PAN

Both Arterial and Venous: Think of these 2 diseases:
- Behçet’s
- Giant cell arteritis
- Wegener’s

Primarily Venous: Think of these 2 diseases:
- Sarcoid
- Multiple sclerosis

Who is the typical Bechet’s disease (BD) pt? An individual of ‘Silk Road’ descent age 25-35

What the classic nonocular complaint in BD? Recurrent oral ulcers

In addition to skin rash, what other nonocular findings are common?

What is the classic rash? Erythema nodosum

Where is the classic location? The pre-tibial region
Erythema nodosum in Behçet
Retinal Vasculitis

Both Arterial and Venous:
Think of these 2 diseases:

- Behçet's
- Granulomatosis

Primarily Arterial:
Think of these 2 diseases:

- SLE
- PAN

Primarily Venous:
Think of these 2 diseases:

- Sarcoid
- Multiple sclerosis

Who is the typical Behçet's disease (BD) pt?
An individual of 'Silk Road' descent age 25-35

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

What the classic nonocular complaint in BD?
Recurrent oral ulcers

In addition to oral ulcers, what three other nonocular findings are common?
--Skin rash
--Genital ulcers
--Arthralgias
Retinal Vasculitis

Primarily Arterial: Think of these 2 diseases:
- SLE
- PAN

Both Arterial and Venous: Think of these 2 diseases:
- Behçet's
- Granulomatosis

Primarily Venous: Think of these 2 diseases:
- Sarcoid
- Multiple sclerosis

Who is the typical Behçet's disease (BD) pt?
An individual of 'Silk Road' descent age 25-35

What is the classic nonocular complaint in BD?
Recurrent oral ulcers

In addition to oral ulcers, what three other nonocular findings are common?
-- Skin rash
-- Genital ulcers
-- Arthralgias

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

- Primarily Arterial:
  Think of these 2 diseases:
  - SLE
  - PAN
  
- Both Arterial and Venous:
  Think of these 2 diseases:
  - Behçet’s
  - Granulomatosis

- Primarily Venous:
  Think of these 2 diseases:
  - Sarcoid
  - Multiple sclerosis

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 ulcers

In addition to oral ulcers, what three other nonocular findings are common?
-- Skin rash
-- Genital ulcers
-- Arthralgias

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 def-- severe vision loss results in as many as 25% of ocular BD cases
Retinal Vasculitis

Primarily Arterial: Think of these 2 diseases:
- SLE
- PAN

Both Arterial and Venous: Think of these 2 diseases:
- Behçet’s
- Granulomatosis

Primarily Venous: Think of these 2 diseases:
- Sarcoid
- Multiple sclerosis

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

In addition to oral ulcers, what three other nonocular findings are common?
-- Skin rash
-- Genital ulcers
-- Arthralgias
Retinal Vasculitis

- Primarily Arterial: Think of these 2 diseases:
  - SLE
  - PAN

- Both Arterial and Venous: Think of these 2 diseases:
  - Behçet’s
  - Granulomatosis

- Primarily Venous: Think of these 2 diseases:
  - Sarcoid
  - Multiple sclerosis

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

What the classic nonocular complaint in BD?
Recurrent oral ulcers

In addition to oral ulcers, what three other nonocular findings are common?
--Skin rash
--Genital ulcers
--Arthralgias

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?
No.
Granulomatosis
Retinal Vasculitis

Primarily Arterial:
Think of these 2 diseases:
- PAN
- SLE

Primarily Venous:
Think of these 2 diseases:
- Behçet's
- Granulomatosis

Both Arterial and Venous:
Think of these 2 diseases:
- Wegener's
- Sarcoid

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 ulcers

In addition to oral ulcers, what three other nonocular findings are common?
-- Skin rash
-- Genital ulcers
-- Arthralgias

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?
Mos def--severe vision loss results in as many as 25% of ocular BD cases
Granulomatosis<br>Retinal<br>Vasculitis

Both *Arterial* and *Venous*:
Think of these 2 diseases:

- SLE
- PAN
- Behçet's
- Granulomatosis
- Sarcoid
- Multiple sclerosis

**What is the most common form of ocular involvement?** (It’s not retinal vasculitis.)

**What the Recurrence**

Does BD pose a significant threat to long-term ocular health, and/or vision?
Mos def--severe vision loss results in as many as 25% of ocular BD cases

*In addition to oral ulcers, what three other nonocular findings are common?*
--Skin rash
--Genital ulcers
--Arthralgias
Granulomatosis Retinal Vasculitis

Retinal Vasculitis

- Primarily Arterial: Think of these 3 diseases:
  - SLE
  - PAN
  - Behçet's

- Primarily Venous: Think of these 3 diseases:
  - Sarcoid
  - Multiple sclerosis
  - Multiple sclerosis

- Both Arterial and Venous: Think of these 2 diseases:
  - Behçet's
  - Granulomatosis

- Acute vs Chronic
- Granulomatous vs Nongranulomatous

What is the most common form of ocular involvement? (It’s not retinal vasculitis.)
- Anterior uveitis, often with a classic finding

What does BD pose a significant threat to long-term ocular health, and/or vision?
- Mos def--severe vision loss results in as many as 25% of ocular BD cases

In addition to oral ulcers, what three other nonocular findings are common?
- Skin rash
- Genital ulcers
- Arthralgias
Retinal Vasculitis

- Primarily Arterial: Think of these 2 diseases:
  - PAN
  - SLE

- Behçet’s

- Primarily Venous: Think of these 2 diseases:
  - Sarcoid
  - Multiple sclerosis

- In addition to oral ulcers, what three other nonocular findings are common?
  - Skin rash
  - Genital ulcers
  - Arthralgias

- What is the most common form of ocular involvement? (It’s not retinal vasculitis.)
  - Acute nongranulomatous anterior uveitis, often with a hypopyon

- Does BD pose a significant threat to long-term ocular health, and/or vision?
  - Mos def--severe vision loss results in as many as 25% of ocular BD cases
Retinal Vasculitis

BD: Hypopyon
Granulomatosis Retinal Vasculitis

Primarily Arterial: Think of these 2 diseases:
- SLE
- PAN

Both Arterial and Venous: Think of these 2 diseases:
- Behçet's
- Granulomatosis

Primarily Venous: Think of these 2 diseases:
- Sarcoid
- Multiple sclerosis

Who is the typical BD patient?
- An individual of 'Silk Road' descent, age 25-35

What is the classic nonocular complaint in BD?
- Recurrent oral ulcers

In addition to oral ulcers, what three other nonocular findings are common?
- Skin rash
- Genital ulcers
- Arthralgias

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?
- Yes--severe vision loss results in as many as 25% of ocular BD cases

What is the most common form of ocular involvement? (It’s not retinal vasculitis.)
- Acute nongranulomatous anterior uveitis, often with a hypopyon

Can BD retinal vasculitis lead to retinal ischemia with subsequent neovascularization?
- Yes--it is an occlusive vasculitis, so ischemia is a common occurrence, and a significant cause of ocular morbidity and vision loss.
Retinal Vasculitis

Primary Arterial:
Think of these 2 diseases:
- PAN
- SLE

Primary Venous:
Think of these 2 diseases:
- Behçet’s
- Sarcoid

Both Arterial and Venous:
Think of these 2 diseases:
- Granulomatosis
- Multiple sclerosis

What is the most common form of ocular involvement? (It’s not retinal vasculitis.)
Acute nongranulomatous anterior uveitis, often with a hypopyon

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

What is the classic nonocular complaint in BD?
Recurrent oral ulcers

In addition to oral ulcers, what three other nonocular findings are common?
-- Skin rash
-- Genital ulcers
-- Arthralgias

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?
Mos def—severe vision loss results in as many as 25% of ocular BD cases

Can BD retinal vasculitis lead to retinal ischemia with subsequent neovascularization?
Yes—it is an occlusive vasculitis, so ischemia is a common occurrence, and is a significant cause of ocular morbidity and vision loss
Retinal Vasculitis

BD: Occlusive vasculitis
Retinal Vasculitis

Primarily Arterial:
Think of these 2 diseases:
- PAN
- SLE

Both Arterial and Venous:
Think of these 2 diseases:
- Behçet’s
- Wegener’s

Primarily Venous:
Think of these 2 diseases:
- Sarcoid
- Multiple sclerosis

Is ocular involvement common in BD?

What is the most common form of ocular involvement? (It’s not retinal vasculitis.)
Acute nongranulomatous anterior uveitis, often with a hypopyon

Can BD retinal vasculitis lead to retinal ischemia with subsequent neovascularization?
Yes—it is an occlusive vasculitis, so ischemia is a common occurrence, and is a significant cause of ocular morbidity and vision loss

How is the diagnosis of BD made?

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

What is the classic nonocular complaint in BD?
Recurrent oral ulcers

In addition to oral ulcers, what three other nonocular findings are common?
- Skin rash
- Genital ulcers
- Arthralgias

Does BD pose a significant threat to long-term ocular health, and/or vision?
Yes; estimates of ocular involvement run as high as 70%

Severe vision loss results in as many as 25% of ocular BD cases
**Retinal Vasculitis**

- **Primarily Arterial:** Think of these 2 diseases:
  - PAN
  - SLE

- **Both Arterial and Venous:** Think of these 2 diseases:
  - Behçet’s
  - Wegener’s

- **Primarily Venous:** Think of these 2 diseases:
  - Sarcoid
  - Multiple sclerosis

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**Questions:**

1. What is the typical BD pt?
   - An individual of 'Silk Road' descent age 25-35

2. What is the classic nonocular complaint in BD?
   - Recurrent oral ulcers

3. In addition to oral ulcers, what three other nonocular findings are common?
   - Skin rash
   - Genital ulcers
   - Arthralgias

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

5. Does BD pose a significant threat to long-term ocular health, and/or vision?
   - Yes—it is an occlusive vasculitis, so ischemia is a common occurrence, and is a significant cause of ocular morbidity and vision loss

6. What is the most common form of ocular involvement? (It's not retinal vasculitis.)
   - Acute nongranulomatous anterior uveitis, often with a hypopyon

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**Additional Information:**

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

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

- **Can BD retinal vasculitis lead to retinal ischemia with subsequent neovascularization?**
  - Yes—it is an occlusive vasculitis, so ischemia is a common occurrence, and is a significant cause of ocular morbidity and vision loss
Granulomatosis Retinal Vasculitis

Primarily Arterial: Think of these 3 diseases:
- PAN
- SLE
- Behçet’s

Primarily Venous: Think of these 3 diseases:
- Sarcoid
- Multiple sclerosis
- Macular degeneration

Both Arterial and Venous: Think of these 3 diseases:
- Wegener’s
- Granulomatosis
- Multiple sclerosis

Retinal Vasculitis

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

What is the most common form of ocular involvement? (It’s not retinal vasculitis.) Acute nongranulomatous anterior uveitis, often with a hypopyon.

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.

Can BD retinal vasculitis lead to retinal ischemia with subsequent neovascularization? Yes—it is an occlusive vasculitis, so ischemia is a common occurrence, and is a significant cause of ocular morbidity and vision loss.
**Retinal Vasculitis**

**Primarily Arterial:**
Think of these 2 diseases:
- PAN
- SLE

**Both Arterial and Venous:**
Think of these 2 diseases:
- Behçet’s
- Wegener’s

**Primarily Venous:**
Think of these 2 diseases:
- Sarcoid
- Multiple sclerosis

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**What is the most common form of ocular involvement?** (It’s not retinal vasculitis.)
Acute nongranulomatous anterior uveitis, often with a hypopyon.

**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.

**Can BD retinal vasculitis lead to retinal ischemia with subsequent neovascularization?**
Yes—it is an occlusive vasculitis, so ischemia is a common occurrence, and is a significant cause of ocular morbidity and vision loss.
Retinal Vasculitis

Primarily Arterial: Think of these 2 diseases:
- PAN
- SLE
- Behçet's
- Wegener's
- Granulomatosis

Both Arterial and Venous: Think of these 2 diseases:
- Retinal Vasculitis
- Multiple sclerosis

Primarily Venous: Think of these 2 diseases:
- Sarcoid
- Acute retinal necrosis

For more on Behçet’s, see the self-titled slide-set

Acute nongranulomatous anterior uveitis, often with a hypopyon

What 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.

Can BD retinal vasculitis lead to retinal ischemia with subsequent neovascularization?
Yes—it is an occlusive vasculitis, so ischemia is a common occurrence, and is a significant cause of ocular morbidity and vision loss.
Retinal Vasculitis

Primarily Arterial: Think of these 2 diseases:
- SLE
- PAN

Both Arterial and Venous: Think of these 2 diseases:
- Behçet’s
- Granulomatosis with polyangiitis (GwP)

Primarily Venous: Think of these 2 diseases:
- Sarcoid
- Multiple sclerosis

If you’re having trouble remembering that granulomatosis with polyangiitis (GwP) is the entity formerly known as Wegener’s…
Retinal Vasculitis

Primarily Arterial:
Think of these 2 diseases:
- SLE
- PAN

Both Arterial and Venous:
Think of these 2 diseases:
- Behçet's
- Granulomatosis with polyangiitis

Primarily Venous:
Think of these 2 diseases:
- Sarcoid
- Multiple sclerosis

granulomatosis with polyangiitis (GwP)

If you’re having trouble remembering that granulomatosis with polyangiitis (GwP) is the entity formerly known as Wegener’s… think of the little ‘w’ as standing for ‘Wegener’s’
Retinal Vasculitis

Primarily Arterial: Think of these 2 diseases:
- SLE
- PAN

Both Arterial and Venous: Think of these 2 diseases:
- Behçet’s
- Granulomatosis with polyangiitis

Primarily Venous: Think of these 2 diseases:
- Sarcoid
- Multiple sclerosis

What is the classic triad of granulomatosis with polyangiitis (GwP)?
Retinal Vasculitis

Primarily Arterial:
Think of these 2 diseases:
- SLE
- PAN

Both Arterial and Venous:
Think of these 2 diseases:
- Behçet’s
- Granulomatosis with polyangiitis

Primarily Venous:
Think of these 2 diseases:
- Sarcoid
- Multiple sclerosis

What is the classic triad of granulomatosis with polyangiitis (GwP)?
Necrotizing vasculitis of:
--the upper and lower respiratory tract
--the kidneys
--small and medium-sized arteries and veins
Retinal Vasculitis

Primarily Arterial: Think of these 2 diseases:

Both Arterial and Venous: Think of these 2 diseases:

- Behçet’s
- Granulomatosis with polyangiitis

Primarily Venous: Think of these 2 diseases:

- Sarcoid
- Multiple sclerosis

What is the classic presentation of upper respiratory tract involvement?

- Sinusitis with bloody discharge

What is the classic triad of granulomatosis with polyangiitis (GwP)? Necrotizing vasculitis of:
- the upper and lower respiratory tract
- the kidneys
- small and medium-sized arteries and veins
Retinal Vasculitis

Primarily Arterial: Think of these 2 diseases:

- PAN
- SLE

Both Arterial and Venous: Think of these 2 diseases:

- Behçet’s
- Granulomatosis with polyangiitis

Primarily Venous: Think of these 2 diseases:

- Sarcoid
- Multiple sclerosis

What is the classic triad of granulomatosis with polyangiitis (GwP)? Necrotizing vasculitis of:
-- the **upper** and lower respiratory tract
-- the kidneys
-- small and medium-sized arteries and veins

What is the classic presentation of upper respiratory tract involvement? Sinusitis with bloody discharge
Retinal Vasculitis

Primarily Arterial:
Think of these 2 diseases:

Both Arterial and Venous:
Think of these 2 diseases:

Primarily Venous:
Think of these 2 diseases:

What is the classic triad of granulomatosis with polyangiitis (GwP)?
Necrotizing vasculitis of:
--the upper and lower respiratory tract
--the kidneys
--small and medium-sized arteries and veins

What is the classic presentation of upper respiratory tract involvement?
Sinusitis with bloody discharge

PAN
SLE
Behçet’s
Granulomatosis with polyangiitis
Sarcoid
Multiple sclerosis
Primarily Venous: Think of these 3 diseases:
- Sarcoid
- Multiple sclerosis
- Retinal Vasculitis

Primarily Arterial: Think of these 3 diseases:
- PAN
- SLE
- Behçet's

Both Arterial and Venous: Think of these 3 diseases:
- Granulomatosis with polyangiitis
- What is the classic triad of granulomatosis with polyangiitis (GwP)? Necrotizing vasculitis of:
  -- the **upper** and **lower** respiratory tract
  -- the kidneys
  -- small and medium-sized arteries and veins

What is the classic presentation of upper respiratory tract involvement?
- Sinusitis with bloody discharge

What is the classic presentation of lower respiratory tract involvement?
- Behçet's
- Granulomatosis with polyangiitis

Retinal Vasculitis
Primarily Venous: Think of these 3 diseases:
- Sarcoid
- Multiple sclerosis
- Retinal Vasculitis

Primarily Arterial:
Think of these 3 diseases:
- Behçet’s
- Granulomatosis with polyangiitis
- PAN

What is the classic presentation of upper respiratory tract involvement? Sinusitis with bloody discharge

What is the classic presentation of lower respiratory tract involvement? A variety of pulmonary S/S: Cough (+/- hemoptysis), wheezing, etc

What is the classic triad of granulomatosis with polyangiitis (GwP)? Necrotizing vasculitis of:
- the upper respiratory tract
- the kidneys
- small and medium-sized arteries and veins

Retinal Vasculitis
Primarily Venous: Think of these 3 diseases:
- Sarcoid
- Multiple sclerosis
- 2

Primarily Arterial: Think of these 2 diseases:
- Retinal Vasculitis
- 2

What is the classic presentation of upper respiratory tract involvement? Sinusitis with bloody discharge

What is the classic presentation of lower respiratory tract involvement? A variety of pulmonary S/S: Cough (+/- hemoptysis), wheezing, etc

What is the classic triad of granulomatosis with polyangiitis (GwP)? Necrotizing vasculitis of:
- the upper and lower respiratory tract
- the kidneys
- small and medium-sized arteries and veins

What specific pathologic renal condition is caused by GwP?

Retinal Vasculitis
Primarily Venous:
Think of these 3 diseases:
- Sarcoid
- Multiple sclerosis
- 2

Primarily Arterial:
Think of these 2 diseases:
- Retinal Vasculitis
- 2

What is the classic triad of granulomatosis with polyangiitis (GwP)?
Necrotizing vasculitis of:
- the upper and lower respiratory tract
- the kidneys
- small and medium-sized arteries and veins

What is the classic presentation of lower respiratory tract involvement?
A variety of pulmonary S/S: Cough (+/- hemoptysis), wheezing, etc

What is the classic presentation of upper respiratory tract involvement?
Sinusitis with bloody discharge

What specific pathologic renal condition is caused by GwP?
Glomerulonephritis

Retinal Vasculitis
Primarily Arterial: Think of these 3 diseases:

Primarily Venous: Think of these 2 diseases:

What is the classic presentation of upper respiratory tract involvement?

Sinusitis

What is the classic presentation of lower respiratory tract involvement?

A variety of pulmonary S/S: Cough (+/- hemoptysis), wheezing, etc

What is the classic triad of granulomatosis with polyangiitis (GwP)?

Necrotizing vasculitis of:
--the upper and lower respiratory tract
--the kidneys
--small and medium-sized arteries and veins

What specific pathologic renal condition is caused by GwP?

Glomerulonephritis

Of the three, which does the Uveitis book describe as “the most characteristic clinical feature” of GwP?

Retinal Vasculitis

Sarcoid

Multiple sclerosis

SLE

PAN

What is the classic presentation of GwP?

Necrotizing vasculitis of:
--the upper and lower respiratory tract
--the kidneys
--small and medium-sized arteries and veins

What is the classic presentation of upper respiratory tract involvement?

Sinusitis with bloody discharge

What is the classic presentation of lower respiratory tract involvement?

- Behçet’s
- Granulomatosis with polyangiitis
Retinal Vasculitis

Of the three, which does the Uveitis book describe as “the most characteristic clinical feature” of GwP?

Sinus dz

Primarily Arterial:
Think of these 2 diseases:

What is the classic presentation of upper respiratory tract involvement?
Sinusitis with bloody discharge

What is the classic presentation of lower respiratory tract involvement?
A variety of pulmonary S/S: Cough (+/- hemoptysis), wheezing, etc

Granulomatosis with polyangiitis

Behçet’s

Primarily Venous:
Think of these 2 diseases:

Sarcoid
Multiple sclerosis

What is the classic triad of granulomatosis with polyangiitis (GwP)?
Necrotizing vasculitis of:
--the upper and lower respiratory tract
--the kidneys
--small and medium-sized arteries and veins

What specific pathologic renal condition is caused by GwP?

Glomerulonephritis
Primarily Venous: Think of these 3 diseases:
- Sarcoid
- Multiple sclerosis
- Retinal Vasculitis

Primarily Arterial: Think of these 3 diseases:
- PAN
- SLE
- Behçet's

Both Arterial and Venous: Think of these 3 diseases:
- Granulomatosis with polyangiitis

What is the classic triad of granulomatosis with polyangiitis (GwP)?
Necrotizing vasculitis of:
--the **upper** and **lower** respiratory tract
--the kidneys
--small and medium-sized arteries and veins

What is the classic presentation of upper respiratory tract involvement?
Sinusitis with bloody discharge

What is the classic presentation of lower respiratory tract involvement?
A variety of pulmonary S/S: Cough (+/- hemoptysis), wheezing, etc

What specific pathologic renal condition is caused by GwP?
Glomerulonephritis

What is the classic finding on CT?
Bony destruction of the sinuses

Retinal Vasculitis
Primarily Venous:
Think of these 3 diseases:
- Sarcoid
- Multiple sclerosis
- Behçet’s

Primarily Arterial:
Think of these 2 diseases:
- PAN
- SLE

Both Arterial and Venous:
Think of these 3 diseases:
- Granulomatosis with polyangiitis

What is the classic finding on CT?
Bony destruction of the sinuses

Arterial:
Think of these 2 diseases:
- Sinusitis with bloody discharge
- What is the classic presentation of upper respiratory tract involvement?
  - Granulomatosis with polyangiitis

What is the classic presentation of lower respiratory tract involvement?
A variety of pulmonary S/S: Cough (+/- hemoptysis), wheezing, etc

What is the classic triad of granulomatosis with polyangiitis (GwP)?
Necrotizing vasculitis of:
- the upper and lower respiratory tract
- the kidneys
- small and medium-sized arteries and veins

What is the classic presentation of lower respiratory tract involvement?
Sinusitis with bloody discharge

What is the classic triad of granulomatosis with polyangiitis (GwP)?
Necrotizing vasculitis of:
- the upper and lower respiratory tract
- the kidneys
- small and medium-sized arteries and veins

What specific pathologic renal condition is caused by GwP?
Glomerulonephritis

What is the classic finding on CT?
Bony destruction of the sinuses

What is the classic presentation of lower respiratory tract involvement?
Sinusitis with bloody discharge
A) CT scan of the sinus in a patient with GwP, showing nasal septum perforation (arrow) and maxillary sinusitis with some degree of maxillary sinusitis and atrophy, with osteosclerosis and bony thickening of the paranasal sinuses. B) CT scan of the sinus in a patient with GwP showing sinusitis and sinus wall erosion leading to the formation of a communication (fistula) between right maxillary sinus, nasal cavity and orbit (double-headed arrow)

GwP: Sinusitis
Primarily Venous:
Think of these 3 diseases:
- Sarcoid
- Multiple sclerosis

Primarily Arterial:
Think of these 3 diseases:
- PAN
- SLE
- Behçet’s

Both Arterial and Venous:
Think of these 3 diseases:
- Granulomatosis with polyangiitis

What is the classic presentation of upper respiratory tract involvement?
Sinusitis with bloody discharge

What is the classic presentation of lower respiratory tract involvement?
A variety of pulmonary S/S: Cough (+/- hemoptysis), wheezing, etc

What is the classic triad of granulomatosis with polyangiitis (GwP)?
Necrotizing vasculitis of:
- the upper and lower respiratory tract
- the kidneys
- small and medium-sized arteries and veins

What specific pathologic renal condition is caused by GwP?
Glomerulonephritis

What is the classic finding on CT?
Bony destruction of the sinuses

What is the classic finding on CT?
Cavitary lung lesions
**Primarily Venous:**
Think of these 3 diseases:
- Sarcoid
- Multiple sclerosis
- 2

**Primarily Arterial:**
Think of these 3 diseases:
- Retinal Vasculitis
- PAN
- SLE

**Arterial:**
Think of these 2 diseases:
- Behçet's
- Granulomatosis with polyangiitis

What is the classic presentation of lower respiratory tract involvement?
- Sinusitis with bloody discharge

What is the classic triad of granulomatosis with polyangiitis (GwP)?
- Necrotizing vasculitis of:
  - The upper and lower respiratory tract
  - The kidneys
  - Small and medium-sized arteries and veins

What is the classic presentation of upper respiratory tract involvement?
- Cough (+/- hemoptysis), wheezing, etc

What specific pathologic renal condition is caused by GwP?
- Glomerulonephritis

What is the classic finding on CT?
- Bony destruction of the sinuses
- Cavitary lung lesions

What is the classic finding on CT?
- Retinal Vasculitis

Retinal Vasculitis

GwP: Cavitary lung lesions
Primarily Venous:
Think of these 3 diseases:

- Sarcoid
- Multiple sclerosis
- 2

Retinal Vasculitis

Primarily Arterial:
Think of these 3 diseases:

- PAN
- SLE
- 2

Both Arterial and Venous:
Think of these 3 diseases:

- Granulomatosis with polyangiitis

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Necrotizing vasculitis of:
- the upper respiratory tract
- the lower respiratory tract
- the kidneys
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What is the classic finding on CT?
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What is the classic finding on CT?
Bony destruction of the sinuses

What is the classic finding on CT?
Bony destruction of the sinuses

What is the classic presentation of lower respiratory tract involvement?
A variety of pulmonary S/S: Cough (+/- hemoptysis), wheezing, etc

What is the classic presentation of upper respiratory tract involvement?
Sinusitis with bloody discharge

What lab findings result?

Hematuria and proteinuria

What specific pathologic renal condition is caused by GwP?
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Next Q
**Primarily Venous:**
Think of these 3 diseases:
- Sarcoid
- Multiple sclerosis
- Retinal Vasculitis

**Primarily Arterial:**
Think of these 3 diseases:
- PAN
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What lab findings result?
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What is the classic triad of granulomatosis with polyangiitis (GwP)?
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What lab findings result?
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Two other manifestations are common as well—what are they?
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Think of these 2 diseases:
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Arterial:
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Both Arterial and Venous:
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What is the classic triad of granulomatosis with polyangiitis (GwP)?
Necrotizing vasculitis of:
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--the kidneys

What is the classic presentation of *upper* respiratory tract involvement?
Sinusitis with bloody discharge

What is the classic finding on CT?
Bony destruction of the sinuses

What lab findings result?
Hematuria and proteinuria

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What is the classic presentation of *lower* respiratory tract involvement?
A variety of pulmonary S/S: Cough (+/- hemoptysis), wheezing, etc

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Cavitary lung lesions

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Primarily Arterial: Think of these 3 diseases:
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Both Arterial and Venous: Think of these 3 diseases:
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- Wegener’s Granulomatosis
- Churg-Strauss Syndrome

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**Retinal Vasculitis**

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**How does retinal vasculitis manifest?**

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What is the prognosis for untreated GwP?
Dismal—the 1-yr mortality rate is 80%

What is the treatment of choice?
Steroids + cyclophosphamide

Retinal Vasculitis
**Primarily Venous:**

Think of these 3 diseases:
- Sarcoid
- Multiple sclerosis
- Retinal Vasculitis

**Primarily Arterial:**

Think of these 3 diseases:
- PAN
- SLE
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**Arterial and Venous:**

Think of these 3 diseases:
- Granulomatosis with polyangiitis

What is the classic triad of granulomatosis with polyangiitis (GwP)?
Necrotizing vasculitis of:
- the upper and lower respiratory tract
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What lab findings result?
- Hematuria and proteinuria

Two other manifestations are common as well—what are they?
- Rash
- Arthralgias

OK then, what proportion of GwP pts manifest vasculitis?
About 10%

How does retinal vasculitis manifest?
As CWS, or arterial occlusions (branch or central)

(It’s not retinal.)

Orbital inflammation

What is the next most common manifestation?
Scleritis (including PUK)

What is the classic presentation of upper respiratory tract involvement?
Sinusitis with bloody discharge

What is the classic presentation of lower respiratory tract involvement?
A variety of pulmonary S/S: Cough (+/- hemoptysis), wheezing, etc

What specific pathologic renal condition is caused by GwP?
Glomerulonephritis

What is the classic finding on CT?
- Bony destruction of the sinuses
- Cavitary lung lesions

What is the classic finding on CT?

What is the prognosis for untreated GwP?
Dismal—the 1-yr mortality rate is 80%

What is the treatment of choice?
Steroids + cyclophosphamide

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Cyclophosphamide is an extremely harsh drug—why not tx with steroids alone?
Because steroid monotherapy for GwP is associated with an increased risk of mortality

Steroid monotherapy? No!

Retinal Vasculitis
In broadest terms, how would you describe sarcoid?

Primarily

Venous:
Think of these 2 diseases:

Sarcoid

Multiple sclerosis
Retinal Vasculitis

In broadest terms, how would you describe sarcoid? It is a multi-organ disease characterized by the presence of noncaseating granulomas. What is the cause? As of this writing, it is unknown. What organ/system is most commonly involved in sarcoid? The lungs. Which other organ systems can be involved? Pretty much all of them. What percentage of sarcoid pts have ocular involvement? About 50%. Who is the typical sarcoid pt in the US? An African-American. During what age-range does dz onset typically occur? Usually 20 to 50.
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Immunologically, how would you describe sarcoid?

Primarily Arterial

Think of these 3 diseases:

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