Persistent Fetal Vasculature

In a nutshell: What is the issue in PFV?
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*By what name was PFV formerly known?*
In a nutshell: What is the issue in PFV?
Portions of the posterior fetal vasculature of the lens (aka the primary vitreous) continue to exist (i.e., it persists) past the developmental stage at which it should have regressed.

By what name was PFV formerly known?
Persistent hyperplastic primary vitreous (PHPV)
Persistent Fetal Vasculature

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What are the three vitreouses, ie, what do they form?
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What is the hyaloid system?
**Persistent Fetal Vasculature**

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*What is the hyaloid system?*

The fetal vasculature that derives from the hyaloid artery
Persistent Fetal Vasculature

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The fetal vasculature that derives from the hyaloid artery

Where does the hyaloid artery run?
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What is the hyaloid system?
The fetal vasculature that derives from the hyaloid artery

Where does the hyaloid artery run?
From the optic nerve head to the back of the lens
(we will have much more to say about the hyaloid artery)
In a nutshell: What is the issue in PFV?
Portions of the posterior fetal vasculature of the lens (aka the primary vitreous) continue to exist (ie, it ‘persists’) past the developmental stage at which it should have regressed.

Is PFV primarily sporadic, or hereditary (or are both equally likely)?
**Persistent Fetal Vasculature**

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Does PFV usually present unilaterally, or bilaterally (or are both equally likely)?
It is in over % of cases.
Persistent Fetal Vasculature

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Is PFV primarily sporadic, or hereditary (or are both equally likely)?
The vast majority of cases are sporadic.

Does PFV usually present unilaterally, or bilaterally (or are both equally likely)?
It is unilateral in over 90% of cases.
The vascular supply encapsulating the developing lens is called the **tunica vasculosa lentis**.
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The vascular supply encapsulating the developing lens is called the **tunica vasculosa lentis**. *It has three sections:*

1) The **three words** derives from the **two words** arteries

2)

3)
The vascular supply encapsulating the developing lens is called the **tunica vasculosa lentis**.

*It has three sections:*

1. The **anterior vascular capsule** derives from the **long ciliary arteries**
2. 
3. 

---

**Persistent Fetal Vasculature**
The vascular supply encapsulating the developing lens is called the **tunica vasculosa lentis**. **It has three sections:**

1. The **anterior vascular capsule** derives from the **long ciliary** arteries.
2. The **Posterior vascular capsule** arises from the **hyaloid** artery.
3. **Persistent Fetal Vasculature**

![Diagram of Tunica Vasculosa Lentis](image)
The vascular supply encapsulating the developing lens is called the **tunica vasculosa lentis**. 

It has three sections:

1) The *anterior vascular capsule* derives from the long ciliary arteries
2) The *posterior vascular capsule* arises from the hyaloid artery
3)
The vascular supply encapsulating the developing lens is called the **tunica vasculosa lentis**. **It has three sections:**

1. The **anterior vascular capsule** derives from the long ciliary arteries
2. The **posterior vascular capsule** arises from the hyaloid artery
3. The **capsulopupillary portion** anastomoses the anterior and posterior sections of the tunica

![Diagram of Tunica Vasculosa Lentis](image-url)
The vascular supply encapsulating the developing lens is called the **tunica vasculosa lentis**.

**It has three sections:**

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Persistent Fetal Vasculature

Tunica vasculosa lentis
In the figure of this very premature infant, the tunical vasculosa lentis surrounds the lens (arrows 1) and is contiguous with the hyaloid vascular system (arrow 2). Notice the glial sheath of the hyaloid artery (arrow 3).
The vascular supply encapsulating the developing lens is called the tunica vasculosa lentis. **It has three sections:**

1. The **anterior vascular capsule** derives from the long ciliary arteries
2. The **posterior vascular capsule** arises from the hyaloid artery
3. The **capsulopupillary portion** anastomoses the anterior and posterior sections of the tunica

The hyaloid artery also send branches to the developing retina
What is the normal natural history of the hyaloid system specifically, and the tunica vasculosa lentis in general?
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The hyaloid system regresses in the 8th month, whereas the anterior portion hangs on until the 9th month.
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What is the name of the intra-vitreal space, running from the ONH to the back of the lens, that is left behind by the regression of the hyaloid artery?
What is the normal natural history of the hyaloid system specifically, and the tunica vasculosa lentis in general?
The hyaloid system regresses in the 8th month, whereas the anterior portion hangs on until the 9th.

What is the name of the intra-vitreal space, running from the ONH to the back of the lens, that is left behind by the regression of the hyaloid artery?
Cloquet’s canal
Persistent Fetal Vasculature

Cloquet canal

Cloquet’s canal
Persistent Fetal Vasculature

Single loop of a persistent hyaloid artery extending anteriorly within Cloquet’s canal to insert on the posterior capsule of the lens.
In a nutshell: What is the issue in PFV?

Hearkening back to a question from a previous slide as a refresher: What did we say was the issue in PFV?
In a nutshell: What is the issue in PFV?
Portions of the posterior fetal vasculature of the lens (aka the *primary vitreous*) continue to exist (ie, it ‘persists’) past the developmental stage at which it should have regressed.

Hearkening back to a question from a previous slide as a refresher:
What did we say was the issue in PFV?
We said the issue was a *failure of the hyaloid system to fully regress*. 
Persistent Fetal Vasculature

PFV comes in two forms. What are they?
Persistent Fetal Vasculature

PFV comes in two forms. What are they? Anterior and posterior
Persistent Fetal Vasculature

PFV comes in two forms. What are they?
Anterior and posterior

What is the basic nature of each?
**Persistent Fetal Vasculature**

Anterior  

Posterior

*PFV comes in two forms. What are they?*
Anterior and posterior

*What is the basic nature of each?*
Anterior PFV involves pathology… [location]
Posterior PFV involves pathology…

Anterior PFV:
Persistent Fetal Vasculature

PFV comes in two forms. What are they?
Anterior and posterior

What is the basic nature of each?
Anterior PFV involves pathology…in the retrolental space
Posterior PFV involves pathology…

Anterior PFV:
Retrolental space
**Persistent Fetal Vasculature**

**Anterior**

**Posterior**

PFV comes in two forms. What are they?
Anterior and posterior

What is the basic nature of each?
Anterior PFV involves pathology…in the retrolental space
Posterior PFV involves pathology… [anatomy issue]

Anterior PFV:
Retrolental space

Posterior PFV:
Anterior PFV involves pathology...in the retrolental space

Posterior PFV involves pathology...related to the retinal branches of the hyaloid artery
Persistent Fetal Vasculature

Anterior

Posterior

Can both anterior and posterior PFV be present in the same eye?

Anterior PFV: Retrolental space

Posterior PFV: Retinal branch remnants
Can both anterior and posterior PFV be present in the same eye?
Yes

Anterior PFV: Retrolental space

Posterior PFV: Retinal branch remnants
Anterior Posterior Persistent Fetal Vasculature

Anterior

Posterior

Shouldn’t a failure to regress by the anterior vascular capsule constitute anterior PFV?
Shouldn’t a failure to regress by the **anterior** vascular capsule constitute **anterior** PFV? No, because the anterior vasc capsule derives from the long ciliary arteries, not the hyaloid system.
Speaking of: Two remnants of a failed-to-completely-regress anterior vascular capsule are commonly encountered at the slit lamp. What are they?
--?
--?

Shouldn’t a **failure to regress by the anterior vascular capsule** constitute anterior PFV?
No, because the anterior vasc capsule derives from the long ciliary arteries, not the hyaloid system
Speaking of: Two remnants of a failed-to-completely-regress anterior vascular capsule are commonly encountered at the slit lamp. What are they?
--Epicapsular star
--Persistent pupillary membrane

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--- Epicapsular star
--- Persistent pupillary membrane

How does an epicapsular star present?

Anterior vascular capsule

**Persistent Fetal Vasculature**
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How does an epicapsular star present?
As a tiny, star-shaped schmutz on the anterior capsule
Persistent Fetal Vasculature

Epicapsular star
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Is it common?

Never
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How does a persistent pupillary membrane present?

- As fibers stretching across the pupillary aperture
- It varies significantly, from a few barely-visible strands to a vast and intricate web
- Very common
- Rarely visually significant
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How extensive is it?
It varies significantly, from a few barely-visible strands to a vast and intricate web.

Is it common?
Very

Is it visually significant?
Rarely
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Persistent Fetal Vasculature

Anterior

Posterior

OK, back to PFV. Take note:

Anterior PFV: Retrolental space

Posterior PFV: Retinal branch remnants
Persistent Fetal Vasculature

Anterior

Mild ‘dz’ ↔ continuum ↔ Severe dz

Posterior

Mild ‘dz’ ↔ continuum ↔ Severe dz

Anterior and posterior PFV both exist on a ‘continuum of severity’ ranging from clinically insignificant to clinically devastating

OK, back to PFV. Take note:

Anterior PFV: Retrolental space

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Persistent Fetal Vasculature

Anterior

Mild ‘dz’ ← continuum → Severe dz

Posterior

Mild ‘dz’ ← continuum → Severe dz

Why is the word disease in ‘quotes’ here?
Why is the word disease in ‘quotes’ here? Because mild PFV is so clinically insignificant it doesn’t really count as a disease *per se*.
What is the eponymous name of the classic manifestation of mild, clinically insignificant anterior PFV?
What is the eponymous name of the classic manifestation of mild, clinically insignificant anterior PFV?

A) Mittendorf dot

Anterior PFV: Retrolental space
What is the eponymous name of the classic manifestation of mild, clinically insignificant anterior PFV?
A Mittendorf dot
What is the eponymous name of the classic manifestation of mild, clinically insignificant anterior PFV?
A Mittendorf dot
How does a Mittendorf dot present?

As a minute white dot on the back of the lens, slightly inferonasal to center.

How does it relate to the hyaloid system?

It is a vestige of the attachment of the hyaloid artery to the back of the lens.

Is it common?

Very.

Is it visually/medically significant?

Never.

What is the eponymous name of the classic manifestation of mild, clinically insignificant anterior PFV?

A Mittendorf dot.

How does a Mittendorf dot present?

Persistent Fetal Vasculature

Anterior

Mild ‘dz’ ← continuum → Severe dz

Posterior

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A Mittendorf dot.

How does a Mittendorf dot present?
As a minute white dot on the back of the lens, slightly inferonasal to center.

How does it relate to the hyaloid system?
How does a Mittendorf dot present?
As a minute white dot on the back of the lens, slightly inferonasal to center

How does it relate to the hyaloid system?
It is a vestige of the attachment of the hyaloid artery to the back of the lens

What is the eponymous name of the classic manifestation of mild, clinically insignificant anterior PFV?
A Mittendorf dot
How does a Mittendorf dot present?
As a minute white dot on the back of the lens, slightly inferonasal to center

How does it relate to the hyaloid system?
It is a vestige of the attachment of the hyaloid artery to the back of the lens

Is it common?

Very

Is it visually/medically significant?

Never

What is the eponymous name of the classic manifestation of mild, clinically insignificant anterior PFV?
A Mittendorf dot

How does a Mittendorf dot present?
As a minute white dot on the back of the lens, slightly inferonasal to center

How does it relate to the hyaloid system?
It is a vestige of the attachment of the hyaloid artery to the back of the lens

Is it common?
How does a Mittendorf dot present?
As a minute white dot on the back of the lens, slightly inferonasal to center

How does it relate to the hyaloid system?
It is a vestige of the attachment of the hyaloid artery to the back of the lens

Is it common?
Very

What is the eponymous name of the classic manifestation of mild, clinically insignificant anterior PFV?
A: **Mittendorf dot**
How does a Mittendorf dot present?
As a minute white dot on the back of the lens, slightly inferonasal to center.

How does it relate to the hyaloid system?
It is a vestige of the attachment of the hyaloid artery to the back of the lens.

Is it common?
Very.

Is it visually/medically significant?
Never.
**Mittendorf dot**

How does a Mittendorf dot present?
As a minute white dot on the back of the lens, slightly inferonasal to center.

How does it relate to the hyaloid system?
It is a vestige of the attachment of the hyaloid artery to the back of the lens.

Is it common?
Very.

Is it visually/medically significant?
Never.

**Persistent Fetal Vasculature**

What is the eponymous name of the classic manifestation of mild, clinically insignificant anterior PFV?
Mittendorf dot.
What is the eponymous name of the classic manifestation of mild, clinically insignificant posterior PFV?
What is the eponymous name of the classic manifestation of mild, clinically insignificant posterior PFV? A

two words

Persistent Fetal Vasculature

Anterior

Mild ‘dz’  continuum  Severe dz

Posterior

Mild ‘dz’  continuum  Severe dz

Posterior PFV: Retinal branch remnants

Mild ‘dz’  continuum  Severe dz
What is the eponymous name of the classic manifestation of mild, clinically insignificant posterior PFV?

A Bergmeister papilla
**Persistent Fetal Vasculature**

Anterior

- Mild ‘dz’
- Severe dz

Posterior

- Mild ‘dz’
- Severe dz

What is the eponymous name of the classic manifestation of mild, clinically insignificant posterior PFV?

A  Bergmeister papilla

Posterior PFV: Retinal branch remnants
**Persistent Fetal Vasculature**

**Anterior**
- Mild ‘dz’
- Severe dz

**Posterior**
- Mild ‘dz’
- Severe dz

What is the eponymous name of the classic manifestation of mild, clinically insignificant posterior PEV?

**Bergmeister papilla**

How does a Bergmeister papilla present?

- As a small tuft of glial tissue in the V that emanates from the ONH
- It is a vestige of the origin of the hyaloid artery at the ONH
- Yes
- Never
How does a Bergmeister papilla present?
As a small tuft of glial tissue in the V that emanates from the ONH.

How does it relate to the hyaloid system?
It is a vestige of the origin of the hyaloid artery at the ONH.

Is it common?
Yes

Is it visually/medically significant?
Never

What is the eponymous name of the classic manifestation of mild, clinically insignificant posterior PFV?
Bergmeister papilla
Persistent Fetal Vasculature

Bergmeister papillae
What is the eponymous name of the classic manifestation of mild, clinically insignificant posterior PFV?

Bergmeister papilla

How does a Bergmeister papilla present?
As a small tuft of glial tissue in the V that emanates from the ONH

How does it relate to the hyaloid system?
**Persistent Fetal Vasculature**

- **Anterior**
  - Mild ‘dz’
  - Severe ‘dz’

- **Posterior**
  - Mild ‘dz’
  - Severe ‘dz’

**What is the eponymous name of the classic manifestation of mild, clinically insignificant posterior PFV?**

- **Bergmeister papilla**

**How does a Bergmeister papilla present?**
- As a small tuft of glial tissue in the V that emanates from the ONH

**How does it relate to the hyaloid system?**
- It is a vestige of the origin of the hyaloid artery at the ONH
Persistent Fetal Vasculature

Anterior

Mild ‘dz’ ↔ continuum → Severe dz

Posterior

Mild ‘dz’ ↔ continuum → Severe dz

What is the eponymous name of the classic manifestation of mild, clinically insignificant posterior PFV?

Bergmeister papilla

How does a Bergmeister papilla present?
As a small tuft of glial tissue in the V that emanates from the ONH

How does it relate to the hyaloid system?
It is a vestige of the origin of the hyaloid artery at the ONH

Is it common?
Never
Persistent Fetal Vasculature

Anterior

Mild ‘dz’ ← continuum → Severe dz

Posterior

Mild ‘dz’ ← continuum → Severe dz

What is the eponymous name of the classic manifestation of mild, clinically insignificant posterior PFV?

Bergmeister papilla

How does a Bergmeister papilla present?
As a small tuft of glial tissue in the V that emanates from the ONH

How does it relate to the hyaloid system?
It is a vestige of the origin of the hyaloid artery at the ONH

Is it common?
Yes
**Persistent Fetal Vasculature**

- **Anterior**
  - Mild ‘dz’ ↔ continuum → Severe dz

- **Posterior**
  - Mild ‘dz’ ↔ continuum → Severe dz

What is the eponymous name of the classic manifestation of mild, clinically insignificant posterior PFV?

**Bergmeister papilla**

**How does a Bergmeister papilla present?**
As a small tuft of glial tissue in the V that emanates from the ONH

**How does it relate to the hyaloid system?**
It is a vestige of the origin of the hyaloid artery at the ONH

**Is it common?**
Yes

**Is it visually/medically significant?**
Never
**Persistent Fetal Vasculature**

Anterior

Mild 'dz' ← continuum → Severe dz

Posterior

Mild 'dz' ← continuum → Severe dz

What is the eponymous name of the classic manifestation of mild, clinically insignificant posterior PEV?

**Bergmeister papilla**

How does a Bergmeister papilla present?
As a small tuft of glial tissue in the V that emanates from the ONH

How does it relate to the hyaloid system?
It is a vestige of the origin of the hyaloid artery at the ONH

Is it common?
Yes

Is it visually/medically significant?
Never
How does a Bergmeister papilla present?
As a small tuft of glial tissue in the V that emanates from the ONH.

How does it relate to the hyaloid system?
It is a vestige of the origin of the hyaloid artery at the ONH.

Is it common?
Yes

Is it visually/medically significant?
Never

I saw a Bergmeister papilla that was perfused. Is this a still-viable hyaloid artery remnant?

What is the eponymous name of the classic manifestation of mild, clinically insignificant posterior PFV?
Bergmeister papilla
What is the eponymous name of the classic manifestation of mild, clinically insignificant posterior PFV?

**Bergmeister papilla**

How does a Bergmeister papilla present?
As a small tuft of glial tissue in the V that emanates from the ONH.

How does it relate to the hyaloid system?
It is a vestige of the origin of the hyaloid artery at the ONH.

Is it common?
Yes

Is it visually/medically significant?
Never
**Persistent Fetal Vasculature**

**Anterior**
- Mild ‘dz’
- Severe ‘dz’

**Posterior**
- Mild ‘dz’
- Severe ‘dz’

What is the eponymous name of the classic manifestation of mild, clinically insignificant posterior PFV?
- **Bergmeister papilla**

I saw a Bergmeister papilla that was perfused. Is this a still-viable hyaloid artery remnant?
- You’d think so, but no. It’s a **prepapillary vascular loop**.

What is a prepapillary vascular loop?
- It is a normal retinal vessel that ‘loops’ into a Bergmeister papilla, then returns.

How does a Bergmeister papilla present?
- As a small tuft of glial tissue in the V that emanates from the ONH.

How does it relate to the hyaloid system?
- It is a vestige of the origin of the hyaloid artery at the ONH.

Is it common?
- Yes

Is it visually/medically significant?
- Never
**Persistent Fetal Vasculature**

**Anterior**
- Mild ‘dz’
- Severe dz

**Posterior**
- Mild ‘dz’
- Severe dz

What is the eponymous name of the classic manifestation of mild, clinically insignificant posterior PFV?

**Bergmeister papilla**

How does a Bergmeister papilla present?
As a small tuft of glial tissue in the V that emanates from the ONH

How does it relate to the hyaloid system?
It is a vestige of the origin of the hyaloid artery at the ONH

Is it common?
Yes

Is it visually/medically significant?
Never

---

I saw a Bergmeister papilla that was perfused. Is this a still-viable hyaloid artery remnant?
You’d think so, but no. It’s a **prepapillary vascular loop**.

What is a prepapillary vascular loop?
It is a normal retinal vessel that ‘loops’ into a Bergmeister papilla, then exits.
Persistent Fetal Vasculature

Prepapillary vascular loop
Persistent Fetal Vasculature

Prepapillary vascular loop with a figure-of-eight configuration
**Persistent Fetal Vasculature**

Anterior

Mild ‘dz’ \(\xrightarrow{\text{continuum}}\) Severe dz

Posterior

Mild ‘dz’ \(\xleftarrow{\text{continuum}}\) Severe dz

*What is the main manifestation of severe anterior PFV?*
Persistent Fetal Vasculature

Anterior

Mild ‘dz’ Severe dz

Posterior

Mild ‘dz’ Severe dz

What is the main manifestation of severe anterior PFV?

A white, vascularized retrolental membrane
What is the main manifestation of severe anterior PFV?
A white, vascularized retrolental membrane
Persistent Fetal Vasculature

PFV: Retrolental membrane
Persistent Fetal Vasculature

Anterior

Mild ‘dz’  ↔  continuum  ↔  Severe dz

Posterior

Mild ‘dz’  ↔  continuum  ↔  Severe dz

What is the main manifestation of severe anterior PFV?
A white, vascularized retrolental membrane

What finding—described in the BCSC as “classic” for anterior PFV—is associated with the peripheral aspect of the retrolental membrane?
Persistent Fetal Vasculature

Anterior

Mild ‘dz’ ↔ continuum ↔ Severe dz

Posterior

Mild ‘dz’ ↔ continuum ↔ Severe dz

What is the main manifestation of severe anterior PFV?
A white, vascularized retrolental membrane

What finding—described in the BCSC as “classic” for anterior PFV—is associated with the peripheral aspect of the retrolental membrane?
Elongated are visible through the pupil when dilated
**Persistent Fetal Vasculature**

Anterior

- Mild ‘dz’
- **Severe dz**

Posterior

- Mild ‘dz’
- **Severe dz**

What is the main manifestation of severe anterior PFV?
A white, vascularized retrolental membrane

What finding—described in the BCSC as “classic” for anterior PFV—is associated with the peripheral aspect of the retrolental membrane?
Elongated ciliary processes are visible through the pupil when dilated
Persistent Fetal Vasculature

PFV: Retrolental membrane. Note the ciliary processes (arrow)
What is the main manifestation of severe anterior PFV?
A **white**, vascularized retrolental membrane

Three other classic manifestations of severe anterior PFV:
--The eye is…
--The lens is…
--The anterior chamber is…
What is the main manifestation of severe anterior PFV?
A white, vascularized retrolental membrane

Three other classic manifestations of severe anterior PFV:
--The eye is... microphthalmic
--The lens is...
--The anterior chamber is...
What is the main manifestation of severe anterior PFV?
A white, vascularized retrolental membrane

How prevalent is microphthalmia in severe anterior PFV?

Three other classic manifestations:
--The eye is... microphthalmic
--The lens is...
--The anterior chamber is...
What is the main manifestation of severe anterior PFV?
A white, vascularized retrolental membrane

Three other classic manifestations:
--The eye is microphthalmic
--The lens is...
--The anterior chamber is...

How prevalent is microphthalmia in severe anterior PFV?
Very. In fact, if an eye with apparent PFV isn’t microphthalmic, you should question the diagnosis.
Persistent Fetal Vasculature

PFV: Microphthalmia
**Persistent Fetal Vasculature**

Anterior

Mild ‘dz’ 乐 Severe dz

Posterior

Mild ‘dz’ 乐 Severe dz

What is the main manifestation of severe anterior PFV?
A **white**, vascularized retrolental membrane

Three other classic manifestations of severe anterior PFV:
--The eye is…**microphthalmic**
--The lens is…[two key attributes]
--The anterior chamber is…
**Persistent Fetal Vasculature**

Anterior

Mild ‘dz’ ↔ continuum → **Severe dz**

Posterior

Mild ‘dz’ ← continuum → **Severe dz**

*What is the main manifestation of severe anterior PFV?*

A **white**, vascularized retrolental membrane

*Three other classic manifestations of severe anterior PFV:*

--The eye is…**microphthalmic**
--The lens is…**small** and cataractous
--The anterior chamber is…
Persistent Fetal Vasculature

**Anterior**
- Mild ‘dz’
- **Severe dz**

**Posterior**
- Mild ‘dz’
- **Severe dz**

What is the main manifestation of severe anterior PFV?
A **white**, vascularized retrolental membrane

Three other classic manifestations of severe anterior PFV:
-- The eye is... **microphthalmic**
-- The lens is... **small** and **cataractous**
-- The anterior chamber is...

Where does anterior PFV rank as a cause of unilateral cataract in kids?
What is the main manifestation of severe anterior PFV?
A white, vascularized retrolental membrane

Three other classic manifestations of severe anterior PFV:
-- The eye is... microphthalmic
-- The lens is... small and cataractous
-- The anterior chamber is...

Where does anterior PFV rank as a cause of unilateral cataract in kids?
#1
Persistent Fetal Vasculature

PFV: Cataract
**Persistent Fetal Vasculature**

**Anterior**
- Mild ‘dz’
- Severe dz

**Posterior**
- Mild ‘dz’

**continuum**

What is the main manifestation of severe anterior PFV?
A **white**, vascularized retrolental membrane

*Three other classic manifestations of severe anterior PFV:*
-- The eye is **microphthalmic**
-- The lens is **small** and **cataractous**
-- The anterior chamber is **[ominous status]**
What is the main manifestation of severe anterior PFV?
A white, vascularized retrolental membrane

Three other classic manifestations of severe anterior PFV:
--The eye is...microphthalmic
--The lens is...small and cataractous
--The anterior chamber is...shallow
Persistent Fetal Vasculature

PFV: Shallow AC
What is the main manifestation of severe anterior PFV?
A) white, vascularized retrolental membrane

Why refer to the shallow-ness of the anterior chamber as ‘ominous’?
--The lens is small and cataractous
--The anterior chamber is shallow
What is the main manifestation of severe anterior PFV?
A white, vascularized retrolental membrane

Why refer to the shallow-ness of the anterior chamber as ‘ominous’?
-- Because it places the eye at significant risk for angle-closure glaucoma
-- The lens is small and cataractous
-- The anterior chamber is shallow
Persistent Fetal Vasculature

Anterior

Mild ‘dz’ ↔ continuum Severe dz

Posterior

Mild ‘dz’ ↔ continuum Severe dz

What is the main manifestation of severe anterior PFV?
A white, vascularized retrolental membrane

Three other classic manifestations of severe anterior PFV:
--The eye is...microphthalmic
--The lens is...small and cataractous
--The anterior chamber is...shallow

What is the natural hx of severe anterior PFV?
Relentless progression of the cataract and AC shallowing, resulting in blindness and glaucoma

Is severe PFV amenable to surgical intervention?
Yes. The cataract and retrolental membrane can be removed, and this usually precludes development of secondary glaucoma. However, the post-op course is usually fraught, and the visual prognosis is guarded.
What is the main manifestation of severe anterior PFV?
A white, vascularized retrolental membrane

Three other classic manifestations of severe anterior PFV:
--The eye is...microphthalmic
--The lens is...small and cataractous
--The anterior chamber is...shallow

What is the natural hx of severe anterior PFV?
Relentless progression of the cataract and AC shallowing, resulting in blindness and glaucoma
Persistent Fetal Vasculature

Anterior

Mild ‘dz’  continuum  Severe dz

Posterior

Mild ‘dz’  continuum  Severe dz

What is the main manifestation of severe anterior PFV?
A white, vascularized retrolental membrane

Three other classic manifestations of severe anterior PFV:
--The eye is microphthalmic
--The lens is small and cataractous
--The anterior chamber is shallow

What is the natural hx of severe anterior PFV?
Relentless progression of the cataract and AC shallowing, resulting in blindness and glaucoma
Persistent Fetal Vasculature

PFV: Note the advanced cataract and severely shallow AC. (Note also the thick white retrolental membrane, and the persistent hyaloid artery)
**Persistent Fetal Vasculature**

**Anterior**

- Mild ‘dz’
- **Severe dz**

**Posterior**

- Mild ‘dz’
- **Severe dz**

---

**What is the main manifestation of severe anterior PFV?**

A **white**, vascularized retrolental membrane

**Three other classic manifestations of severe anterior PFV:**

---

- The eye is... **microphthalmic**
- The lens is... **small** and **cataractous**
- The anterior chamber is... **shallow**

---

**What is the natural hx of severe anterior PFV?**

Relentless progression of the cataract and AC shallowing, resulting in blindness and glaucoma

**Is severe PFV amenable to surgical intervention?**
Persistence Fetal Vasculature

Anterior
Mild ‘dz’  continuum  Severe ‘dz’

Posterior
Mild ‘dz’  continuum  Severe ‘dz’

What is the main manifestation of severe anterior PFV?
A white, vascularized retrolental membrane

Three other classic manifestations of severe anterior PFV:
--The eye is...microphthalmic
--The lens is...small and cataractous
--The anterior chamber is...shallow

What is the natural hx of severe anterior PFV?
Relentless progression of the cataract and AC shallowing, resulting in blindness and glaucoma

Is severe PFV amenable to surgical intervention?
Yes. The cataract and retrolental membrane can be removed, and this usually precludes development of secondary glaucoma. However, the post-op course is fraught, and the visual prognosis guarded.
At what age does severe anterior PFV present?

Mild ‘dz’
Severe dz

What is the main manifestation of severe anterior PFV?
A white, vascularized retrolental membrane

Three other classic manifestations of severe anterior PFV:
--The eye is…microphthalmic
--The lens is…small and cataractous
--The anterior chamber is…shallow
At what age does severe anterior PFV present?
Birth

What is the main manifestation of severe anterior PFV?
A white, vascularized retrolental membrane

Three other classic manifestations of severe anterior PFV:
--The eye is...microphthalmic
--The lens is...small and cataractous
--The anterior chamber is...shallow
Persistent Fetal Vasculature

Mild 'dz'

Severe 'dz'

What is the main manifestation of severe anterior PFV?
A white, vascularized retrolental membrane

Three other classic manifestations of severe anterior PFV:
-- The eye is... microphthalmic
-- The lens is... small and cataractous
-- The anterior chamber is... shallow

At what age does severe anterior PFV present?
Birth

What is the classic clinical finding that alerts the pediatrician to the presence of a significant ocular issue?

Leukocoria. Anterior PFV is very high on the DDx for it.

What is the most feared dx on the leukocoria DDx?
Retinoblastoma (Rb)
At what age does severe anterior PFV present?
Birth

What is the classic clinical finding that alerts the pediatrician to the presence of a significant ocular issue? Leukocoria. Anterior PFV is very high on the DDx for it.

Mild ‘dz’

What is the main manifestation of severe anterior PFV?

- A white, vascularized retrolental membrane

Three other classic manifestations of severe anterior PFV:

-- The eye is... microphthalmic
-- The lens is... small and cataractous
-- The anterior chamber is... shallow

Severe dz
At what age does severe anterior PFV present? Birth

What is the classic clinical finding that alerts the pediatrician to the presence of a significant ocular issue? Leukocoria. Anterior PFV is very high on the DDx for it.

What is the most feared dx on the leukocoria DDx? Retinoblastoma (Rb)

What is the main manifestation of severe anterior PFV? White, vascularized retrolental membrane

Three other classic manifestations of severe anterior PFV:
--The eye is...microphthalmic
--The lens is...small and cataractous
--The anterior chamber is...shallow
At what age does severe anterior PFV present? Birth

What is the classic clinical finding that alerts the pediatrician to the presence of a significant ocular issue? Leukocoria. Anterior PFV is very high on the DDx for it.

What is the most feared dx on the leukocoria DDx? Retinoblastoma (Rb)

What is the main manifestation of severe anterior PFV? White, vascularized retrolental membrane

Three other classic manifestations of severe anterior PFV:
--The eye is...microphthalmic
--The lens is...small and cataractous
--The anterior chamber is...shallow
At what age does severe anterior PFV present?
Birth

It’s crucial that you be able to distinguish between anterior PFV and Rb, both in the clinic and on the OKAP/Boards. So let’s see how Rb stacks up against what we’ve learned about anterior PFV.

What is the most feared dx on the leukocoria DDx?
Retinoblastoma (Rb)

What is the main manifestation of severe anterior PFV?
A white, vascularized retrolental membrane

Three other classic manifestations of severe anterior PFV:
--The eye is... microphthalmic
--The lens is... small and cataractous
--The anterior chamber is... shallow
What is the main manifestation of severe anterior PFV?
A white, vascularized retro lent al membrane

Three other classic manifestations of severe anterior PFV:
--The eye is…microphthalmic
--The lens is…small and cataractous
--The anterior chamber is…shallow

At what age does severe anterior PFV present?
Birth

What is the classic clinical finding that alerts the pediatrician to the presence of a significant ocular issue?
Leukocoria. Anterior PFV is very high on the DDx for it.

What is the most feared dx on the leukocoria DDx?
Retinoblastoma (Rb)

Does Rb present with leukocoria at birth?
No
What is the main manifestation of severe anterior PFV?

Retinoblastoma (Rb)

Three other classic manifestations of severe anterior PFV:

--The eye is... microphthalmic
--The lens is... small and cataractous
--The anterior chamber is... shallow

At what age does severe anterior PFV present?

Birth

What is the classic clinical finding that alerts the pediatrician to the presence of a significant ocular issue?

Leukocoria. Anterior PFV is very high on the DDx for it.

What is the most feared dx on the leukocoria DDx?

Retinoblastoma (Rb)

Does Rb present with leukocoria at birth? No
At what age does severe anterior PFV present?
Birth

What is the classic clinical finding that alerts the pediatrician to the presence of a significant ocular issue? Leukocoria. Anterior PFV is very high on the DDx for it.

What is the most feared dx on the leukocoria DDx?
Retinoblastoma (Rb)

Does Rb present with leukocoria at birth? No

Does this mean Rb can’t be present at birth?
No, it can. It just isn’t obvious, ie, it isn’t causing leukocoria (or any other outward sign).

Mild ‘dz’

Persistent Fetal Vasculature

Anterior
Posterior

Mild ‘dz’

Severe dz

continuum

What is the main manifestation of severe anterior PFV?
White, vascularized retrolental membrane

Three other classic manifestations of severe anterior PFV:
--The eye is...microphthalmic
--The lens is...small and cataractous
--The anterior chamber is...shallow
At what age does severe anterior PFV present?
Birth

What is the classic clinical finding that alerts the pediatrician to the presence of a significant ocular issue? Leukocoria. Anterior PFV is very high on the DDx for it.

What is the most feared dx on the leukocoria DDx?
Retinoblastoma (Rb)

Does Rb present with leukocoria at birth? No

Does this mean Rb can’t be present at birth? Oh no, it mos def can. It just isn’t obvious, ie, it isn’t causing leukocoria (or any other outward sign).

Mild ‘dz’ continuum

Severe dz continuum

Mild ‘dz’

Severe ‘dz’

continuum

What is the main manifestation of severe anterior PFV?
white, vascularized retrolental membrane

Three other classic manifestations of severe anterior PFV:

--The eye is...microphthalmic
--The lens is...small and cataractous
--The anterior chamber is...shallow
Persistent Fetal Vasculature

Anterior

Persistent Fetal Vasculature (PFV) is a congenital condition where the fetal circulatory system does not regress postnatally. It can present with various manifestations, both anterior and posterior.

Mild ‘dz’

Severe ‘dz’

continuum

What is the main manifestation of severe anterior PFV?

A white, vascularized retrolental membrane

Three other classic manifestations of severe anterior PFV:

--The eye is... microphthalmic
--The lens is... small and cataractous
--The anterior chamber is... shallow

At what age does severe anterior PFV present?

Birth

What is the classic clinical finding that alerts the pediatrician to the presence of a significant ocular issue?

Leukocoria. Anterior PFV is very high on the DDx for it.

What is the most feared dx on the leukocoria DDx?

Retinoblastoma (Rb)

Are Rb eyes microphthalmic?

No

Does Rb present with leukocoria at birth?

No
At what age does severe anterior PFV present?
Birth

What is the classic clinical finding that alerts the pediatrician to the presence of a significant ocular issue? Leukocoria. Anterior PFV is very high on the DDx for it.

What is the most feared dx on the leukocoria DDx? Retinoblastoma (Rb)

Does Rb present with leukocoria at birth? No

Mild ‘dz

What is the main manifestation of severe anterior PFV?
A white, vascularized retrolental membrane

Three other classic manifestations of severe anterior PFV:
--The eye is microphthalmic
--The lens is...small and cataractous
--The anterior chamber is...shallow

Are Rb eyes microphthalmic? No
At what age does severe anterior PFV present? Birth

What is the classic clinical finding that alerts the pediatrician to the presence of a significant ocular issue? Leukocoria. Anterior PFV is very high on the DDx for it.

What is the most feared dx on the leukocoria DDx? Retinoblastoma (Rb)

Does Rb present with leukocoria at birth? No

Are Rb eyes microphthalmic? No

Do Rb eyes have small/cataractous lenses?

Mild ‘dz’ continuum

Severe dz continuum

What is the main manifestation of severe anterior PFV? A white, vascularized retrolental membrane.

Three other classic manifestations of severe anterior PFV:
--The eye is microphthalmic.
--The lens is small and cataractous?
--The anterior chamber is shallow.
At what age does severe anterior PFV present?
Birth

What is the classic clinical finding that alerts the pediatrician to the presence of a significant ocular issue? Leukocoria. Anterior PFV is very high on the DDx for it.

What is the main manifestation of severe anterior PFV?
White, vascularized retrolental membrane

Three other classic manifestations of severe anterior PFV:
--The eye is microphthalmic
--The lens is small and cataractous
--The anterior chamber is shallow

Does Rb present with leukocoria at birth? No

Are Rb eyes microphthalmic? No

Do Rb eyes have small/cataractous lenses? No
Persistent Fetal Vasculature

Anterior

Posterior

Mild ‘dz’

Severe ‘dz’

continuum

What is the main manifestation of severe anterior PFV?

Retinoblastoma (Rb)

What is the classic clinical finding that alerts the pediatrician to the presence of a significant ocular issue?

Leukocoria. Anterior PFV is very high on the DDx for it.

At what age does severe anterior PFV present?

Birth

What is the classic clinical finding that alerts the pediatrician to the presence of a significant ocular issue?

Leukocoria. Anterior PFV is very high on the DDx for it.

Does Rb present with leukocoria at birth? No

Are Rb eyes microphthalmic? No

Do Rb eyes have small/cataractous lenses? No

Do Rb eyes have shallow ACs?
At what age does severe anterior PFV present?
Birth

What is the classic clinical finding that alerts the pediatrician to the presence of a significant ocular issue?
Leukocoria. Anterior PFV is very high on the DDx for it.

What is the main manifestation of severe anterior PFV?
White vascularized retrolental membrane

Three other classic manifestations of severe anterior PFV:
--The eye is microphthalmic
--The lens is small and cataractous
--The anterior chamber is shallow

Do Rb eyes have shallow ACs? No

Are Rb eyes microphthalmic? No

Does Rb present with leukocoria at birth? No

Do Rb eyes have small/cataractous lenses? No
What is the main manifestation of severe anterior PFV?

A white, vascularized retrolental membrane

Three other classic manifestations of severe anterior PFV:

-- The eye is microphthalmic
-- The lens is small and cataractous
-- The anterior chamber is shallow

At what age does severe anterior PFV present?

Birth

What is the classic clinical finding that alerts the pediatrician to the presence of a significant ocular issue?

Leukocoria. Anterior PFV is very high on the DDx for it.

What is the most feared dx on the leukocoria DDx?

Retinoblastoma (Rb)

Are Rb eyes microphthalmic?

No

Anterior PFV Rb Leukocoria at birth?

Yes

No

Microphthalmic?

Yes

No

Cataract present?

Yes

No

AC shallow?

Yes

No

(Review slide—no question, proceed when ready)
### Persistent Fetal Vasculature

#### Anterior PFV

<table>
<thead>
<tr>
<th>What is the main manifestation of severe anterior PFV?</th>
<th>A white, vascularized retrolental membrane</th>
</tr>
</thead>
</table>
| Three other classic manifestations of severe anterior PFV: | -- The eye is microphthalmic
-- The lens is small and cataractous
-- The anterior chamber is shallow |

### At what age does severe anterior PFV present? Birth

### What is the classic clinical finding that alerts the pediatrician to the presence of a significant ocular issue? Leukocoria. Anterior PFV is very high on the DDx for it.

### What is the most feared dx on the leukocoria DDx? Retinoblastoma (Rb)

### Are Rb eyes microphthalmic? No

### Clinically obvious at birth? Yes

### Microphthalmic? Yes

### Cataract present? Yes

### AC shallow? Yes

### Next, we will change gears and consider clinically significant posterior PFV

<table>
<thead>
<tr>
<th>AC shallow?</th>
<th>Yes</th>
<th>No</th>
</tr>
</thead>
</table>

(Review slide—no question, proceed when ready)
In severe PFV…

Is a retrolental membrane present? No

A white, vascularized retrolental membrane

Persistent Fetal Vasculature

Anterior

Mild ‘dz’  Mild ‘dz’

Severe dz  continuum  Severe dz

Posterior

In severe posterior PFV…

The eye is…microphthalmic
The lens is…small and cataractous
The anterior chamber is…shallow
Persistent Fetal Vasculature

Anterior

Mild ‘dz’  Continuum  Severe dz

Posterior

Mild ‘dz’  Continuum  Severe dz

What is the main manifestation of severe anterior PFV?

A white, vascularized retrolental membrane?

In severe posterior PFV…
Is a retrolental membrane present?

The eye is…microphthalmic
The lens is…small and cataractous
The anterior chamber is…shallow
Persistent Fetal Vasculature

Anterior

Mild ‘dz’  Continuum  Severe dz

Posterior

Mild ‘dz’  Continuum  Severe dz

What is the main manifestation of severe anterior PFV?
A white, vascularized retrolental membrane?

In severe posterior PFV...
Is a retrolental membrane present? No

The eye is... microphthalmic
The lens is... small and cataractous
The anterior chamber is... shallow
In severe PFV...

- Is a retrolental membrane present?
- No

 Persistent Fetal Vasculature

Anterior

Mild ‘dz’ ← continuum → Severe dz

Posterior

Mild ‘dz’ ← continuum → Severe dz

What is the main manifestation of severe anterior PFV?
- A white, vascularized retrolental membrane?

Three other classic manifestations of severe anterior PFV:
- The eye is microphthalmic?
- The lens is small and cataractous
- The anterior chamber is shallow

In severe posterior PFV...
Is the eye microphthalmic?

The eye is... *microphthalmic*?
- The lens is small and cataractous
- The anterior chamber is shallow
In severe PFV...

Is a retrolental membrane present?
No

Mild ‘dz’ ↔ continuum ↔ Severe dz

Anterior

Persistent Fetal Vasculature

Posterior

Mild ‘dz’ ↔ continuum ↔ Severe dz

What is the main manifestation of severe anterior PFV?
A white, vascularized retrolental membrane?

In severe posterior PFV...

Is the eye microphthalmic? Yes

The eye is... microphthalmic!
The lens is... small and cataractous
The anterior chamber is... shallow
**Persistent Fetal Vasculature**

**Anterior**
- Mild ‘dz’
- Severe dz

**Posterior**
- Mild ‘dz’
- Severe dz

---

A white, vascularized retrolental membrane?

*In severe posterior PFV... Is the lens small/cataractous?*

- The eye is...microphthalmic!
- The lens is...*small* and cataractous?
- The anterior chamber is...shallow

---

*Hyaloid artery nerve*
In severe PFV...
Is a retrolental membrane present? No

Anterior
Mild ‘dz’ ↔ continuum Severe dz

Posterior
Mild ‘dz’ ↗ continuum Severe dz

What is the main manifestation of severe anterior PFV?
A white, vascularized retrolental membrane?

The eye is... microphthalmic!
The lens is... small and cataractous.
The anterior chamber is... shallow

In severe posterior PFV...
Is the lens small/cataractous? No
In severe PFV...

Is a retrolental membrane present?

No

Anterior

Mild ‘dz’ <-> continuum <-> Severe dz

Posterior

Mild ‘dz’ <-> continuum <-> Severe dz

The main manifestation of severe anterior PFV is a white, vascularized retrolental membrane?

Three other classic manifestations of severe anterior PFV:

- The eye is microphthalmic!
- The lens is small and cataractous
- The anterior chamber is shallow?

In severe posterior PFV...

Is the AC shallow?
In severe PFV…
Is a retrolental membrane present? No

1. Persistent Fetal Vasculature
   - Anterior
     - Mild ‘dz’ to Severe dz
   - Posterior
     - Mild ‘dz’ to Severe dz

What is the main manifestation of severe anterior PFV?
A white, vascularized retrolental membrane?

The eye is…microphthalmic!
The lens is…small and cataractous.
The anterior chamber is…shallow.

In severe posterior PFV…
Is the AC shallow? No

---

Hyaloid artery
Optic nerve
What is the main manifestation of severe anterior PFV? A white, vascularized retrolental membrane?

What is the defining feature of severe posterior PFV?

The eye is... microphthalmic!
The lens is... small and cataractous.
The anterior chamber is... shallow.
**Persistent Fetal Vasculature**

**Anterior**
- Mild ‘dz’
- **continuum**
- Severe dz

**Posterior**
- Mild ‘dz’
- **continuum**
- Severe dz

---

**What is the main manifestation of severe anterior PFV?**

A white, vascularized retrolental membrane?

---

**What is the defining feature of severe posterior PFV?**

*The eye is... microphthalmic!*

*The lens is... small and cataractous.*

*The anterior chamber is... shallow.*

---

Persistent hyaloid branches emanating from the ONH, running atop (and causing) a [two words]
**Persistent Fetal Vasculature**

**Anterior**
- Mild ‘dz’
- Severe dz

**Posterior**
- Mild ‘dz’
- Severe dz

**What is the main manifestation of severe anterior PFV?**
A white, vascularized retrolental membrane?

**What is the defining feature of severe posterior PFV?**

*The eye is... microphthalmic!*

*The lens is... small and cataractous.*

*The anterior chamber is... shallow.*

Persistent hyaloid branches emanating from the ONH, running atop (and causing) a retinal fold.
Persistent Fetal Vasculature

Posterior PFV with a fibrovascular stalk
**Persistent Fetal Vasculature**

**Anterior**
- Mild 'dz'
- Severe 'dz'

**Posterior**
- Mild 'dz'
- Severe 'dz'

**What is the main manifestation of severe anterior PFV?**
- A white, vascularized retrolental membrane?

**Three other classic manifestations of severe anterior PFV:**
- The eye is...
- The lens is...
- The anterior chamber is...

**What is the defining feature of severe posterior PFV?**
- Persistent hyaloid branches emanating from the ONH, running atop (and causing) a retinal fold

**What conditions top the DDx for posterior PFV?**
- ?
- ?
- ?

- The eye is...
- The lens is...
- The anterior chamber is...shallow
**Persistent Fetal Vasculature**

### Anterior
- Mild ‘dz’
- Severe ‘dz’

### Posterior
- Mild ‘dz’
- Severe ‘dz’

What is the main manifestation of severe anterior PFV? A white, vascularized retrolental membrane?

Three other classic manifestations of severe anterior PFV:
- The eye is microphthalmic.
- The lens is small and cataractous.
- The anterior chamber is shallow.

What is the defining feature of severe posterior PFV?
- Persistent hyaloid branches emanating from the ONH, running atop (and causing) a retinal fold.

What conditions top the DDx for posterior PFV?
- ROP
- FEVR
- Toxocariasis
What are the main manifestations of severe anterior PFV?

- A white, vascularized retrolental membrane?
- The eye is microphthalmic!
- The lens is small and cataractous
- The anterior chamber is shallow

What is the defining feature of severe posterior PFV?

- Persistent hyaloid branches emanating from the ONH, running atop (and causing) a retinal fold

What conditions top the DDx for posterior PFV?

- ROP
- FEVR
- Toxocariasis

What does ROP stand for here?
**Persistent Fetal Vasculature**

**Anterior**
- Mild ‘dz’
- Severe dz

**Posterior**
- Mild ‘dz’
- Severe dz

**What is the main manifestation of severe anterior PFV?**
- A white, vascularized retrolental membrane?

**Three other classic manifestations of severe anterior PFV:**
- The eye is microphthalmic!
- The lens is small and cataractous
- The anterior chamber is shallow

**What is the defining feature of severe posterior PFV?**
- Persistent hyaloid branches emanating from the ONH, running atop (and causing) a retinal fold

**What conditions top the DDx for posterior PFV?**
- --ROP
- --FEVR
- --Toxocariasis

**What does ROP stand for here?**
- Retinopathy of prematurity (it has its own slide-set if you’re interested)
**Persistent Fetal Vasculature**

Anterior

- Mild ‘dz’
- Continuum
- Severe dz

Posterior

- Mild ‘dz’
- Continuum
- Severe dz

What is the main manifestation of severe anterior PFV?
- A white, vascularized retrolental membrane?

Three other classic manifestations of severe anterior PFV:
- The eye is... microphthalmic!
- The lens is... small and cataractous
- The anterior chamber is... shallow

What is the defining feature of severe posterior PFV?
- Persistent hyaloid branches emanating from the ONH, running atop (and causing) a retinal fold

What conditions top the DDx for posterior PFV?
- ROP
- FEVR
- Toxocariasis

How about FEVR?
Persistent Fetal Vasculature

Anterior

Mild 'dz' ↔ continuum ➔ Severe dz

Posterior

Mild 'dz' ➔ continuum ➔ Severe dz

What is the main manifestation of severe anterior PFV?

A white, vascularized retrolental membrane?

Three other classic manifestations of severe anterior PFV:

-- The eye is microphthalmic!
-- The lens is small and cataractous
-- The anterior chamber is shallow

What is the defining feature of severe posterior PFV?

Persistent hyaloid branches emanating from the ONH, running atop (and causing) a retinal fold

What conditions top the DDx for posterior PFV?

-- ROP
-- FEVR
-- Toxocariasis

How about FEVR?

Familial exudative vitreoretinopathy (covered in slide-set R3)
Persistent Fetal Vasculature

Anterior

Mild ‘dz’ ↔ continuum → Severe dz

Posterior

Mild ‘dz’ ↔ continuum → Severe dz

What is the main manifestation of severe anterior PFV?

A white, vascularized retrolental membrane?

Three other classic manifestations of severe anterior PFV:

--The eye is... microphthalmic
--The lens is... small and cataractous
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What is the defining feature of severe posterior PFV?

Persistent hyaloid branches emanating from the ONH, running atop (and causing) a retinal fold

What conditions top the DDx for posterior PFV?

--ROP
--FEVR
--Toxocariasis

In a word, what sort of bug is Toxocara?

Toxocariasis

The eye is...
--The lens is...
--The anterior chamber is... shallow

optic nerve
**Persistent Fetal Vasculature**

- **Anterior**
  - Mild ‘dz’
  - Severe dz

- **Posterior**
  - Mild ‘dz’
  - Severe dz

**What is the main manifestation of severe anterior PFV?**
- White, vascularized retrolental membrane?

**Three other classic manifestations of severe anterior PFV:**
- The eye is small and microphthalmic!
- The lens is small and cataractous
- The anterior chamber is shallow

**What is the defining feature of severe posterior PFV?**
- Persistent hyaloid branches emanating from the ONH, running atop (and causing) a retinal fold

**What conditions top the DDx for posterior PFV?**
- ROP
- FEVR
- Toxocariasis

**In a word, what sort of bug is Toxocara?**
- A roundworm (it has its own slide-set)
The next section is a brief recapitulation of the high points for severe anterior PFV
Anterior PFV: Basics

- classic descriptors (three words)
- membrane present behind lens
Anterior PFV: Basics

- White vascularized fibrous membrane present behind lens
**Anterior PFV: Basics**

- White vascularized fibrous membrane present behind lens
- Associated with:
  - eye size
Anterior PFV: Basics

- White **vascularized fibrous** membrane present behind lens
- Associated with:
  - **Microphthalmos**
Anterior PFV: Basics

- White vascularized fibrous membrane present behind lens
- Associated with:
  - Microphthalmos
  - Leukocoria

Persistent Fetal Vasculature
Anterior PFV: Basics

- White *vascularized fibrous* membrane present behind lens
- Associated with:
  - Microphthalmos
  - Leukocoria *at birth*
Anterior PFV: Basics

- White vascularized fibrous membrane present behind lens
- Associated with:
  - Microphthalmos
  - Leukocoria at birth
  - Deep vs shallow anterior chamber
Anterior PFV: Basics

- White vascularized fibrous membrane present behind lens
- Associated with:
  - Microphthalmos
  - Leukocoria at birth
  - Shallow anterior chamber
Anterior PFV: Basics

- White vascularized fibrous membrane present behind lens
- Associated with:
  - Microphthalmos
  - Leukocoria at birth
  - Shallow anterior chamber
  - Long visible around small lens
Anterior PFV: Basics

- White vascularized fibrous membrane present behind lens
- Associated with:
  - Microphthalmos
  - Leukocoria at birth
  - Shallow anterior chamber
  - Long ciliary processes visible around small lens
Anterior PFV: Basics

- White **vascularized fibrous** membrane present behind lens
- Associated with:
  - **Microphthalmos**
  - Leukocoria *at birth*
  - **Shallow** anterior chamber
  - Long **ciliary processes** visible around small lens
- Dehiscence of posterior capsule → lens → cataract and 2o angle closure glaucoma
**Anterior PFV: Basics**

- White **vascularized fibrous** membrane present behind lens
- Associated with:
  - **Microphthalmos**
  - Leukocoria **at birth**
  - **Shallow** anterior chamber
  - Long **ciliary processes** visible around small lens
- Dehiscence of posterior capsule → lens **intumescence** → cataract and **2º angle closure** glaucoma
Anterior PFV: Basics

- White vascularized fibrous membrane present behind lens
- Associated with:
  - Microphthalmos
  - Leukocoria at birth
  - Shallow anterior chamber
  - Long ciliary processes visible around small lens
- Dehiscence of posterior capsule $\rightarrow$ lens intumescence $\rightarrow$ cataract and 2$^\circ$ angle closure glaucoma

Anterior PFV: Management

- Surgical procedure and another surgical procedure to prevent angle closure
Anterior PFV: Basics

- White vascularized fibrous membrane present behind lens
- Associated with:
  - **Microphthalmos**
  - Leukocoria at birth
  - **Shallow** anterior chamber
  - Long ciliary processes visible around small lens
- Dehiscence of posterior capsule → lens intumescence → cataract and 2º angle closure glaucoma

Anterior PFV: Management

- **Lensectomy** and **membranectomy** to prevent angle closure