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Can Coats occur in adulthood?

Yes
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- **RD**: Retinal Detachment

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**Can Coats occur in adulthood?** Yes

**What is adult-onset Coats disease called?** It is called 'adult-onset Coats disease'
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_Coats Disease vs Retinoblastoma_
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**RD?** - Retinal Detachment

**b-scan of sub-RD space reveals…** Sub-RD space empty Calcifications

**CT reveals…** RD RD + Ca 2+

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**Coats Disease vs Retinoblastoma**

**Age (yrs) at presentation**

- Coats: 6-8 yrs
- Rb: 1-2 yrs

**Sex**

- Coats: Male in 75-85%
- Rb: M = F

**% unilateral**

- Coats: 75 - 95%
- Rb: 60%

**Vitreous involvement?**

- Coats: No
- Rb: Frequently (Seeding)

**Cataract?**

- Coats: No
- Rb: Only very late in advanced cases

**Exudate?**

- Coats: Yes
- Rb: No

**RD?**

- Coats: Yes
- Rb: Yes
# Coats Disease vs Retinoblastoma

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- **RD**: Retinal Detachment
- **Ca2+**: Calcium Deposits
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- **RD + Ca**: Retinal Detachment with Calcifications
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## Coats Disease vs Retinoblastoma

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*Let’s drill down on Coats dz…*
Inheritance pattern?

Coats Disease vs Retinoblastoma
Inheritance pattern? None
Coats Disease vs Retinoblastoma

Inheritance pattern? None

Systemic associations?

Coats disease
Coats Disease vs Retinoblastoma

Inheritance pattern? None

Systemic associations? None
Inheritance pattern? None

Systemic associations? None

Neovascularization is... [rare vs common]
Coats Disease vs Retinoblastoma

Inheritance pattern? None

Systemic associations? None

Neovascularization is... rare
Coats Disease vs Retinoblastoma

Inheritance pattern? None

Systemic associations? None

Neovascularization is... rare

Why ask this question? That is, why might neovascularization be expected?
Inheritance pattern? None

Systemic associations? None

Neovascularization is... rare

*Why ask this question? That is, why might neovascularization be expected?* Because retinal nonperfusion is a common finding
Neovascularization is... rare

*Why ask this question? That is, why might neovascularization be expected?* Because retinal nonperfusion is a common finding

*Why doesn’t nonperfusion in Coats dz lead to neo?*
Inheritance pattern? None

Systemic associations? None

Neovascularization is...rare

Why ask this question? That is, why might neovascularization be expected? Because retinal nonperfusion is a common finding

Why doesn’t nonperfusion in Coats dz lead to neo? It is unknown at this time
Coats Disease vs Retinoblastoma

Inheritance pattern? None

Systemic associations? None

Characterized by 4 vascular anomalies:
1) Telangiectasias
2) Venous dilation
3) Microaneurysms
4) Capillary dilation

Neovascularization is... rare
Inheritance pattern? None

Systemic associations? None

Neovascularization is... rare

Coats disease vs Retinoblastoma

Characterized by 4 vascular anomalies:
1) Telangiectasias
2) Venous dilation
3) Microaneurysms
4) Capillary dilation

The vascular anomalies lead to...
Inheritance pattern? None
Systemic associations? None

Characterized by 4 vascular anomalies:
1) Telangiectasias
2) Venous dilation
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4) Capillary dilation

The vascular anomalies lead to subretinal exudates

Neovascularization is... rare
Coats Disease vs Retinoblastoma

Inheritance pattern? None

Systemic associations? None

Characterized by 4 vascular anomalies:
1) Telangiectasias
2) Venous dilation
3) Microaneurysms
4) Capillary dilation

The vascular anomalies lead to…

White-yellow subretinal exudates

Neovascularization is…rare
Coats Disease vs Retinoblastoma

Inheritance pattern? None

Systemic associations? None

Characterized by 4 vascular anomalies:
1) Telangiectasias
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The vascular anomalies lead to...

White-yellow subretinal exudates

Neovascularization is... rare

...and the subretinal exudates lead to...
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Systemic associations? None

Characterized by 4 vascular anomalies:
1) Telangiectasias
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3) Microaneurysms
4) Capillary dilation

The vascular anomalies lead to...
White-yellow subretinal exudates
...and the subretinal exudates lead to...
Exudative RD

Neovascularization is... rare

Usual presentation? +/-
**Coats Disease vs Retinoblastoma**

**Inheritance pattern?** None

**Systemic associations?** None

**Usual presentation?** Leukocoria +/- strabismus

Coats Disease is characterized by 4 vascular anomalies:
1) Telangiectasias
2) Venous dilation
3) Microaneurysms
4) Capillary dilation

The vascular anomalies lead to white-yellow subretinal exudates, and the subretinal exudates lead to exudative RD.

Neovascularization is rare.

**Coats disease**
Coats Disease vs Retinoblastoma

Inheritance pattern? None

Systemic associations? None

Characterized by 4 vascular anomalies:
1) Telangiectasias
2) Venous dilation
3) Microaneurysms
4) Capillary dilation

Neovascularization is... rare

The vascular anomalies lead to...

Coats disease

White-yellow subretinal exudates

...and the subretinal exudates lead to...

Exudative RD

Usual presentation? Leukocoria +/- strabismus
--places it high on the differential for... [bad dz]
Coats Disease vs Retinoblastoma

Inheritance pattern? None

Systemic associations? None

Characterized by 4 vascular anomalies:
1) Telangiectasias
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The vascular anomalies lead to...

White-yellow subretinal exudates

...and the subretinal exudates lead to...

Exudative RD

Neovascularization is... rare

Usual presentation? Leukocoria +/- strabismus
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Coats Disease vs Retinoblastoma

Inheritance pattern? None

Systemic associations? None

Usual presentation? Leukocoria +/- strabismus --places it high on the differential for... Rb

Characterized by 4 vascular anomalies:
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The vascular anomalies lead to... White-yellow subretinal exudates

Neovascularization is rare

Coats Disease vs Retinoblastoma... Speaking of differentials... Besides Rb, what is on the DDx for Coats?

Note: These are the only conditions included on the Coats DDx in both the Peds and Retina books
Inheritance pattern? None
Systemic associations? None

Usual presentation? Leukocoria +/- strabismus
--places it high on the differential for…Rb

Coats Disease vs Retinoblastoma

Characterized by 4 vascular anomalies:
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Coats disease

The vascular anomalies lead to…
White-yellow subretinal exudates

Speaking of differentials…Besides Rb, what is on the DDx for Coats?
--FEVR
--Retinopathy of prematurity (ROP)
--Persistent fetal vasculature (PFV)

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Usual presentation? Leukocoria +/- strabismus
--places it high on the differential for…Rb
Inheritance pattern: None

Systemic associations: None

Usual presentation: Leukocoria +/- strabismus places it high on the differential for...

Rb White-yellow subretinal exudates Exudative RD Neovascularization is rare Coats disease Characterized by 4 vascular anomalies:
1) Telangiectasias
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3) Microaneurysms
4) Capillary dilation

The vascular anomalies lead to...

and the subretinal exudates lead to...

Coats Disease vs Retinoblastoma
Speaking of differentials…Besides Rb, what is on the DDx for Coats? -- FEVR -- Persistent Fetal Vasculature (PFV) -- Retinopathy of prematurity (ROP)

In this context, what does FEVR stand for?

Familial exudative vitreoretinopathy

FEVR is characterized by the failure of something to happen--what?

Failure of the peripheral retina to vascularize

‘Failure of the peripheral retina to vascularize’—that sounds like what condition?

ROP
Coats Disease vs Retinoblastoma

Inheritance pattern? None

Systemic associations? None

Usual presentation? Leukocoria +/- strabismus
--places it high on the differential for…Rb

Characterized by 4 vascular anomalies:
1) Telangiectasias
2) Venous dilation
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The vascular anomalies lead to...
and the subretinal exudates lead to...

Coats Disease vs Retinoblastoma

Speaking of differentials…Besides Rb, what is on the DDx for Coats?
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In this context, what does FEVR stand for?
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Retinopathy of prematurity (ROP)
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Neovascularization is rare
**Coats Disease vs Retinoblastoma**

Inheritance pattern? **None**

Systemic associations? **None**

Usual presentation? **Leukocoria +/- strabismus** – places it high on the differential for… **Rb**

Characterized by **4 vascular anomalies**: 1) Telangiectasias 2) Venous dilation 3) Microaneurysms 4) Capillary dilation

The vascular anomalies lead to…

White-yellow subretinal exudates

Exudative RD

Neovascularization is… rare

Coats disease is characterized by **4 vascular anomalies**:
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White-yellow subretinal exudates

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Neovascularization is… rare

Coats Disease vs Retinoblastoma

Speaking of differentials… **Besides Rb, what is on the DDx for Coats?**

**FEVR** – Familial exudative vitreoretinopathy

FEVR is characterized by the failure of something to happen--what?

In this context, what does FEVR stand for?

Familial exudative vitreoretinopathy

Speaking of differentials… **Besides Rb, what is on the DDx for Coats?**

**FEVR** – Retinopathy of prematurity (ROP)

-- Persistent fetal vasculature (PFV)

Usual presentation? **Leukocoria +/- strabismus** – places it high on the differential for… **Rb**
Inheritance pattern? **None**

Systemic associations? **None**

**Usual presentation?** Leukocoria +/- strabismus -- places it high on the differential for...Rb

---

**Coats Disease vs Retinoblastoma**

Characterized by **4 vascular anomalies:**
1) Telangiectasias
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The vascular anomalies lead to...

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**White-yellow subretinal exudates**

Exudative RD

Neovascularization is rare

**Coats disease** is characterized by **4 vascular anomalies**:
1) Telangiectasias
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The vascular anomalies lead to...

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**In this context, what does FEVR stand for?**
Familial exudative vitreoretinopathy

**FEVR is characterized by the failure of something to happen--what?**
Failure of the peripheral retina to vascularize

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Speaking of differentials...Besides Rb, what is on the DDx for Coats?

**FEVR**

-- Retinopathy of prematurity (ROP)
-- Persistent fetal vasculature (PFV)

---

Usual presentation? Leukocoria +/- strabismus -- places it high on the differential for...Rb
Coats Disease vs Retinoblastoma

Inheritance pattern? None

Systemic associations? None

Usual presentation? Leukocoria +/- strabismus --places it high on the differential for...Rb

Characterized by 4 vascular anomalies:
1) Telangiectasias
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Speaking of differentials...Besides Rb, what is on the DDx for Coats?
FEVR
--Retinopathy of prematurity (ROP)
-- Persistent fetal vasculature (PFV)

55
Inheritance pattern? None

Systemic associations? None

Usual presentation? Leukocoria +/− strabismus -- places it high on the differential for...

Rb

White-yellow subretinal exudates

Exudative RD

Neovascularization is rare

Coats disease

Characterized by 4 vascular anomalies:

1) Telangiectasias
2) Venous dilation
3) Microaneurysms
4) Capillary dilation

The vascular anomalies lead to...

subretinal exudates lead to...

in this context, what does fevr stand for?

FEVR

Retinopathy of prematurity (ROP)

Persistent fetal vasculature (PFV)

Speaking of differentials... Besides Rb, what is on the DDx for Coats?

FEVR

Retinopathy of prematurity (ROP)

Persistent fetal vasculature (PFV)

In this context, what does FEVR stand for?

Familial exudative vitreoretinopathy

Failure of the peripheral retina to vascularize

FEVR is characterized by the failure of something to happen - what?

Coats Disease vs Retinoblastoma

Speaking of differentials... Besides Rb, what is on the DDx for Coats?

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Familial exudative vitreoretinopathy

Failure of the peripheral retina to vascularize

FEVR is characterized by the failure of something to happen - what?

Coats Disease vs Retinoblastoma

Speaking of differentials... Besides Rb, what is on the DDx for Coats?

FEVR

Retinopathy of prematurity (ROP)

Persistent fetal vasculature (PFV)

In this context, what does FEVR stand for?

Familial exudative vitreoretinopathy

Failure of the peripheral retina to vascularize

FEVR is characterized by the failure of something to happen - what?

Coats Disease vs Retinoblastoma

Speaking of differentials... Besides Rb, what is on the DDx for Coats?

FEVR

Retinopathy of prematurity (ROP)

Persistent fetal vasculature (PFV)

In this context, what does FEVR stand for?

Familial exudative vitreoretinopathy

Failure of the peripheral retina to vascularize

FEVR is characterized by the failure of something to happen - what?

Coats Disease vs Retinoblastoma
Inheritance pattern? None

Systemic associations? None

Usual presentation? Leukocoria +/- strabismus -- places it high on the differential for Rb

Characterized by 4 vascular anomalies:
1) Telangiectasias
2) Venous dilation
3) Microaneurysms
4) Capillary dilation

In this context, what does FEVR stand for? Familial exudative vitreoretinopathy

FEVR is characterized by the failure of something to happen -- "Failure of the peripheral retina to vascularize" -- that sounds like what condition? Retinopathy of prematurity (ROP)

Do FEVR pts develop neovascularization a la ROP? Yes

Do FEVR pts get tractional retinal detachments a la ROP? Yes

Speaking of differentials... Besides Rb, what is on the DDx for Coats? - FEVR - Retinopathy of prematurity (ROP) - Persistent fetal vasculature (PFV)

White-yellow subretinal exudates lead to neovascularization, which leads to tractional retinal detachments.

Neovascularization is rare in Coats disease but is more common in Retinoblastoma (Rb).

Coats Disease vs Retinoblastoma
Coats Disease vs Retinoblastoma

Inheritance pattern? None
Systemic associations? None

Usual presentation? Leukocoria +/- strabismus
--- places it high on the differential for...Rb

Coats Disease
Characterized by 4 vascular anomalies:
1) Telangiectasias
2) Venous dilation
3) Microaneurysms
4) Capillary dilation

The vascular anomalies lead to...
Subretinal exudates lead to...

Speaking of differentials... Besides Rb, what is on the DDx for Coats?
- FEVR
- Retinopathy of prematurity (ROP)
- Persistent fetal vasculature (PFV)

Do FEVR pts develop neovascularization a la ROP? Yes

In this context, what does FEVR stand for?
Familial exudative vitreoretinopathy

FEVR is characterized by the failure of something to happen---
Failure of the peripheral retina to vascularize

‘Failure of the peripheral retina to vascularize’---that sounds like what condition?
ROP

Do FEVR pts develop neovascularization a la ROP? Yes
Do FEVR pts get tractional retinal detachments a la ROP? Yes
Inheritance pattern? **None**

Systemic associations? **None**

Usual presentation? **Leukocoria +/- strabismus** -- places it high on the differential for... **Rb**

Characterized by **4 vascular anomalies**:  
1) Telangiectasias  
2) Venous dilation  
3) Microaneurysms  
4) Capillary dilation

The vascular anomalies lead to... subretinal exudates

Do FEVR pts develop neovascularization a la ROP? **Yes**

Do FEVR pts get tractional retinal detachments a la ROP? **Yes**

Speaking of differentials... Besides Rb, what is on the DDx for Coats?  
- FEVR  
- Retinopathy of prematurity (ROP)  
- Persistent fetal vasculature (PFV)

In this context, what does FEVR stand for?  
Familial exudative vitreoretinopathy

FEVR is characterized by the failure of something to happen -- what?  
Failure of the peripheral retina to vascularize

‘Failure of the peripheral retina to vascularize’ -- that sounds like what condition? **ROP**

Coats Disease vs Retinoblastoma
Inheritance pattern? **None**

Systemic associations? **None**

Usual presentation? **Leukocoria +/- strabismus** -- places it high on the differential for... **Rb**

**Coats Disease vs Retinoblastoma**

**Inheritance pattern? None**

**Systemic associations? None**

**Usual presentation? Leukocoria +/- strabismus -- places it high on the differential for... Rb**

**Characterized by 4 vascular anomalies:**
1) Telangiectasias
2) Venous dilation
3) Microaneurysms
4) Capillary dilation

The vascular anomalies lead to...

In this context, what does FEVR stand for?
Familial exudative vitreoretinopathy

Do FEVR pts develop neovascularization a la ROP?
Yes

Do FEVR pts get tractional retinal detachments a la ROP?
Yes

Speaking of differentials... Besides Rb, what is on the DDx for Coats?
- **FEVR**
- **Retinopathy of prematurity (ROP)**
- Persistent fetal vasculature (PFV)

In this context, what does FEVR stand for?
Familial exudative vitreoretinopathy

FEVR is characterized by the failure of something to happen -- what?
Failure of the peripheral retina to vascularize

‘Failure of the peripheral retina to vascularize’ -- that sounds like what condition?
Retinopathy of prematurity (ROP)

Do FEVR pts develop neovascularization a la ROP?
Yes

Do FEVR pts get tractional retinal detachments a la ROP?
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Neovascular anomalies lead to...

**Speaking of differentials... Besides Rb, what is on the DDx for Coats?**
- **FEVR**
- **Retinopathy of prematurity (ROP)**
- Persistent fetal vasculature (PFV)
Coats Disease vs Retinoblastoma

Inheritance pattern? None

Systemic associations? None

Usual presentation? Leukocoria +/- strabismus -- places it high on the differential for...Rb

Characterized by 4 vascular anomalies:
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Coats Disease
Characterized by 4 vascular anomalies:
1) Telangiectasias
2) Venous dilation
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4) Capillary dilation

The vascular anomalies lead to...and the subretinal exudates lead to...

Speaking of differentials...Besides Rb, what is on the DDx for Coats?
- FEVR
- Retinopathy of prematurity (ROP)
- Persistent fetal vasculature (PFV)

Do FEVR pts develop neovascularization a la ROP? Yes
Do FEVR pts get tractional retinal detachments a la ROP? Yes

In this context, what does FEVR stand for? Familial exudative vitreoretinopathy

FEVR is a failure of the peripheral retina to vascularize

‘Failure of the peripheral retina to vascularize’—that sounds like what condition? ROP

So, is FEVR just an ROP variant? Definitely not, for two reasons:
- It is an inherited condition
- It occurs in full term infant who are of normal respiratory status
Coats Disease vs Retinoblastoma

Inheritance pattern? None

Systemic associations? None

Usual presentation? Leukocoria +/- strabismus --places it high on the differential for...Rb

Characterized by 4 vascular anomalies:
1) Telangiectasias
2) Venous dilation
3) Microaneurysms
4) Capillary dilation

The vascular anomalies lead to neovascularization and the subretinal exudates lead to retinal detachment.

Speaking of differentials...Besides Rb, what is on the DDx for Coats?
- FEVR
- Retinopathy of prematurity (ROP)
- Persistent fetal vasculature (PFV)

In this context, what does FEVR stand for?
Familial exudative vitreoretinopathy

FEVR is Failure of Failure to vascularize

So, is FEVR just an ROP variant? Definitely not, for two reasons:
- It is an inherited condition
- It occurs in infant who are full term and of normal respiratory status

Do FEVR pts develop neovascularization a la ROP? Yes

Do FEVR pts get tractional retinal detachments a la ROP? Yes
Inheritance pattern? None
Systemic associations? None

**Usual presentation?** Leukocoria +/- strabismus

---places it high on the differential for... Rb

Characterized by 4 vascular anomalies:
1) Telangiectasias
2) Venous dilation
3) Microaneurysms
4) Capillary dilation

The vascular anomalies lead to... and the subretinal exudates lead to...

--- places it high on the differential for... Rb

Coats Disease vs Retinoblastoma

--- places it high on the differential for... Rb

In this context, what does FEVR stand for?
Familial exudative vitreoretinopathy

Do FEVR pts develop neovascularization a la ROP? **Yes**

Do FEVR pts get tractional retinal detachments a la ROP? **Yes**

So, is FEVR just an ROP variant?
Definitely not, for two reasons:
-- It is an inherited condition
-- It occurs in infant who are... and of... full term and of normal respiratory status

Speaking of differentials... Besides Rb, what is on the DDx for Coats?
- FEVR
- Retinopathy of prematurity (ROP)
- Persistent fetal vasculature (PFV)

In this context, what does FEVR stand for?
Familial exudative vitreoretinopathy

FEVR is Failure of peripheral retina to vascularize

‘Failure of peripheral retina to vascularize’—that sounds like what condition?
Retinopathy of prematurity (ROP)

Do FEVR pts develop neovascularization a la ROP?
**Yes**

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So, is FEVR just an ROP variant?
Definitely not, for two reasons:
-- It is an inherited condition
-- It occurs in infant who are... and of... full term and of normal respiratory status
Inheritance pattern? None
Systemic associations? None

Usual presentation? Leukocoria +/- strabismus
--places it high on the differential for...Rb

Characterized by 4 vascular anomalies:
1) Telangiectasias
2) Venous dilation
3) Microaneurysms
4) Capillary dilation

Coats Disease vs Retinoblastoma

Speaking of differentials...Besides Rb, what is on the DDx for Coats?
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Yes

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subretinal exudates lead to...
Inheritance pattern?
None

Systemic associations?
None

Usual presentation?
Leukocoria
+/– strabismus
--places it high on the differential for… Rb

White-yellow subretinal exudates
Exudative RD

Coats disease Characterized by 4 vascular anomalies:
1) Telangiectasias
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4) Capillary dilation

The vascular anomalies lead to…
… and the subretinal exudates lead to…

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Coats Disease vs Retinoblastoma

Speaking of differentials… Besides Rb, what is on the DDx for Coats?
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-- Retinopathy of prematurity (ROP)
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In this context, what does FEVR stand for?
Familial exudative vitreoretinopathy

FEVR is characterized by the failure of something to happen—what?
Failure of the peripheral retina to vascularize

‘Failure of the peripheral retina to vascularize’—that sounds like what condition?
ROP

Do FEVR pts develop neovascularization a la ROP?
Yes

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Definitely not, for two reasons:
-- It is an inherited condition
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The fact that FEVR is inherited implies what about its management?
It implies two things re diagnosing FEVR:
Family history can cinch the diagnosis in difficult-to-diagnose cases, as can
Examining family members
It also implies that family members should be carefully examined to determine… whether they too require treatment for impending sequelae of FEVR
Inheritance pattern?

None

Systemic associations?

None

Usual presentation?

Leukocoria +/- strabismus

--places it high on the differential for…

Rb

White-yellow subretinal exudates

Exudative RD

Neovascularization is… rare

Coats disease

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Coats Disease vs Retinoblastoma

Speaking of differentials… Besides Rb, what is on the DDx for Coats?

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Persistent fetal vasculature (PFV)

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Coats Disease vs Retinoblastoma

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In this context, what does FEVR stand for? Familial exudative vitreoretinopathy

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Neovascularization is…rare
White-yellow subretinal exudates

Characterized by 4 vascular anomalies: 1) Telangiectasias 2) Venous dilation 3) Microaneurysms 4) Capillary dilation The vascular anomalies lead to… and the subretinal exudates lead to…

In this context, what does FEVR stand for? --Failure of the peripheral retina to vascularize 'Failure of the peripheral retina to vascularize'—that sounds like what condition? ROP

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Inheritance pattern?

None

Systemic associations?

None

Usual presentation?

Leukocoria +/- strabismus

--places it high on the differential for…

Rb

White-yellow subretinal exudates

Exudative RD

Neovascularization is rare

Coats disease

Characterized by 4 vascular anomalies:

1) Telangiectasias
2) Venous dilation
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4) Capillary dilation

The vascular anomalies lead to…

and the subretinal exudates lead to…

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Coats Disease vs Retinoblastoma

Speaking of differentials…Besides Rb, what is on the DDx for Coats?

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Do FEVR pts develop neovascularization a la ROP?

Yes

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FEVR is...

Failure...

Failure of the peripheral retina to vascularize

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Coats Disease vs Retinoblastoma

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69
Inheritance pattern? None
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Rb
White-yellow subretinal exudates
Exudative RD
Neovascularization is... rare
Coats disease
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It also implies that...
family members should be carefully examined to determine...whether they too require treatment for impending sequelae of FEVR

What imaging modality might need to be employed in assessing family members of a FEVR pt?
Widefield FA

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Coats Disease vs Retinoblastoma

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4) Capillary dilation

The vascular anomalies lead to...
White-yellow subretinal exudates
...and the subretinal exudates lead to...
Exudative RD

Usual presentation? Leukocoria +/- strabismus
--places it high on the differential for...Rb

Management: Treat vascular anomalies with [two modalities]
Coats Disease vs Retinoblastoma

Inheritance pattern? **None**

Systemic associations? **None**

Characterized by **4 vascular anomalies**:
1) Telangiectasias
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The vascular anomalies lead to...

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...and the subretinal exudates lead to...

Exudative RD

Neovascularization is... **rare**

Usual presentation? **Leukocoria +/- strabismus**
--places it high on the differential for... **Rb**

Management: Treat vascular anomalies with **cryo, photocoagulation**