

Q

ROP: True or false



- Patients with a PDA are at increased risk of ROP

A

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- Patients with a PDA are at increased risk of ROP **True**



Q

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- Patients with a **PDA** are at increased risk of ROP True

Why would a Personal Digital Assistant put someone at increased risk of ROP?*

**Do y'all know what a Personal Digital Assistant is?*



Q

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Why would a Personal Digital Assistant put someone at increased risk of ROP?
In this context, PDA stands for

quack

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ROP: True or false



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Why would a Personal Digital Assistant put someone at increased risk of ROP?
In this context, PDA stands for **patent ductus arteriosus**



Q

ROP: True or false

- Patients with a PDA are at increased risk of ROP **True**
- Birth weight is a greater predictor for ROP than O₂ exposure

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- Patients with a PDA are at increased risk of ROP **True**
- Birth weight is a greater predictor for ROP than O₂ exposure **True; LBW is #1 risk factor**
(low birth weight)



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BW (gm)	Risk of <i>severe</i> ROP
<750	?%
750-999	
1000-1250	



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1/2
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Note the pattern...



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Is exposure to supplemental O2 a risk factor at all?



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Is exposure to supplemental O2 a risk factor at all?
Yes



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There is another risk factor that is tied for #1 with LBW—what is it?

- Patience
- Birth weight is a greater predictor for ROP than O2 exposure True; **LBW is #1 risk factor** along with...?



A

ROP: True or false

- Patient Infant age, ie, prematurity (probably not surprising that prematurity is a risk factor for retinopathy **of prematurity**)
- Birth weight is a greater predictor for ROP than gestational age exposure True; **LBW is #1 risk factor** along with...Prematurity



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- Patient **Infant age?** premature, prematurity (probably not surprising that prematurity is a risk factor for retinopathy of prematurity) true
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OK, but which variable is the best predictor of **when** an infant will develop significant ROP?

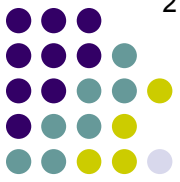


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*Can you be more specific? That is, **which** age is the best predictor--postmenstrual, gestational or chronologic? (And what are these different ages anyway?)*



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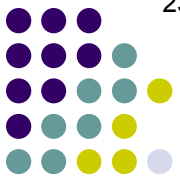
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Development of significant ROP correlates best with the infant's **postmenstrual** age.

Postmenstrual age equals one way of measuring infant age + another age.



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Postmenstrual age equals gestational age at birth + chronologic (postnatal) age.



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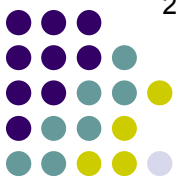
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What does this indicate about the relationship between timing of ROP development and an infant's chronologic age?



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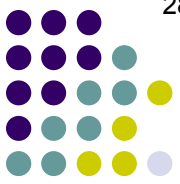
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It indicates that younger preemies take longer to develop significant ROP than do older preemies.



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It indicates that younger preemies take longer to develop significant ROP than do older preemies. Consider two infants, one born at gestational age 24 weeks, the other at 27. Neither is expected to develop ROP before postmenstrual age 31 weeks. Thus, the 24-weeker needs to be examined at chronologic age 7 weeks ($24+7=31$), whereas the 27-weeker should be examined at chronologic age 4 weeks ($27+4=31$). (We'll have more to say later about ROP screening, and its timing.)



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Which of the following are demonstrated risk factors for developing ROP?

- Paternal age
- Sepsis
- Receiving a blood transfusion (the infant, not mother)
- Poor postnatal weight gain
- Fever (w/o sepsis)
- Intraventricular hemorrhage



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*What does the term **Everest in utero** have to do with ROP?*

The term highlights the fact that the gestational environment is profoundly hypoxic compared to life ex utero. Oxygen levels in utero are about what they are at the 26,000 ft level on Everest—the so-called ‘death zone.’ It is under these O₂ conditions that the retinal vasculature is supposed to develop.



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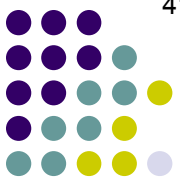
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When the preemie experiences normal ex utero O₂ levels, further development of the retinal vasculature is suppressed. This leaves more peripheral retinal areas with inadequate oxygenation. **As they mature, these hypoxic retinal cells do what hypoxic retinal cells tend to do—they produce VEGF. The result is the now-familiar cascade of neovascularization, bleeding, and tractional retinal detachment.**



ROP: True or false

- Patients with a PDA are at increased risk of ROP True
- Birth weight is a greater predictor for ROP than O_2

In other words, ROP is a **biphasic disease**:

(No question yet—keep going)

strong evidence that excess P_aO_2 is not causative

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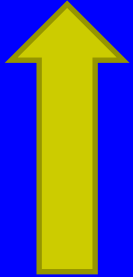


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--First, premature birth (+/- supplemental O_2) exposes the immature retina to vastly higher-than-normal O_2 levels, leading to **downregulation of VEGF**. This causes the immature retinal vascular tree to **stop proliferating**.

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(What happens later?)

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--Later, the (unvascularized) peripheral retina becomes metabolically active. The lack of vascularization renders the peripheral retina hypoxic, leading to **upregulation of VEGF**. This causes the vascular tree to **start proliferating again**.

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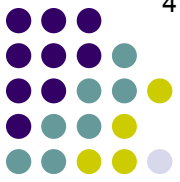
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- Whites have a greater risk of ROP than blacks



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M vs F



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- Exposure to ambient light has a small but significant effect on ROP development **False; the *Light-ROP* study found no relationship**
- Infants with a R→L cardiac shunt (and subsequent low O₂ sat) are protected from ROP **False, and this provides strong evidence that excess P_aO₂ is not causative**
- Whites have a greater risk of ROP than blacks **True**
- The sexes have roughly equal ROP incidence rates **False; ROP is significantly more common in males**



Q

ROP: True or false

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- Once the ROP process starts, it usually progresses to an advanced level



Q/A

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A

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- Whites have a greater risk of ROP than blacks **True**
- The sexes have roughly equal ROP incidence rates **False; ROP is significantly more common in males**
- Once the ROP process starts, it usually progresses to an advanced level **False; roughly 80% of ROP arrests spontaneously, without significant sequelae**

Q

- *ROP classification*: Based on pathology criteria
(called...), another criterion (called...), and another criterion (two words) status:

A

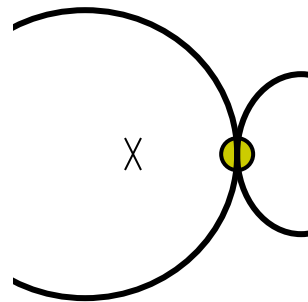
- *ROP classification*: Based on pathology **location** (**zone**), **appearance** (**stage**), and **plus disease** status:

Q

- *ROP classification*: Based on pathology **location** (**zone**), **appearance** (*stage*), and **plus disease** status:

- **Location**

- Zone 1:
- Zone 2
- Zone 3



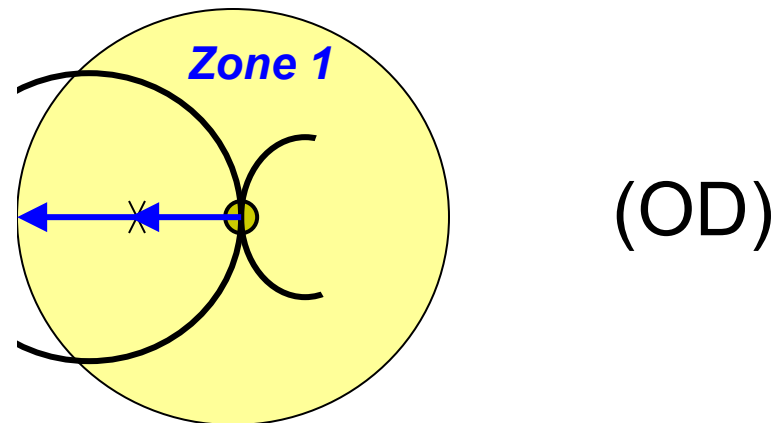
(OD)

A

- *ROP classification*: Based on pathology **location** (**zone**), **appearance** (*stage*), and **plus disease** status:

- **Location**

- *Zone 1*: Circle around ONH w/ radius 2x disc-fovea distance
- *Zone 2* (ONH = optic nerve head)
- *Zone 3*

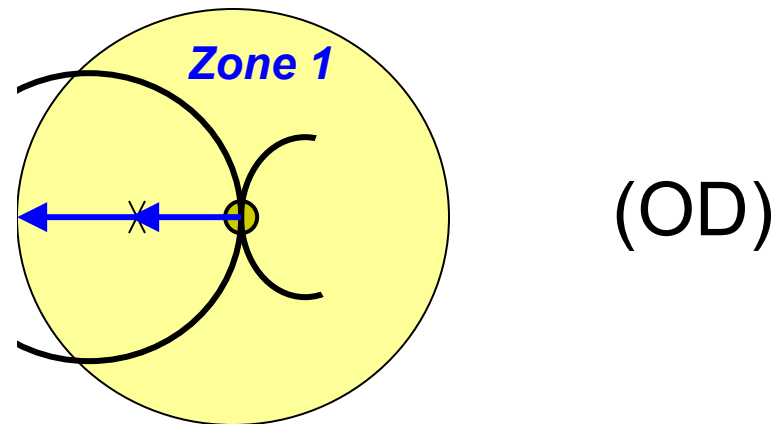


Q

- *ROP classification*: Based on pathology **location** (**zone**), **appearance** (*stage*), and **plus disease** status:

- **Location**

- Zone 1: Circle around ONH w/ radius 2x disc-fovea distance
- Zone 2:
- Zone 3

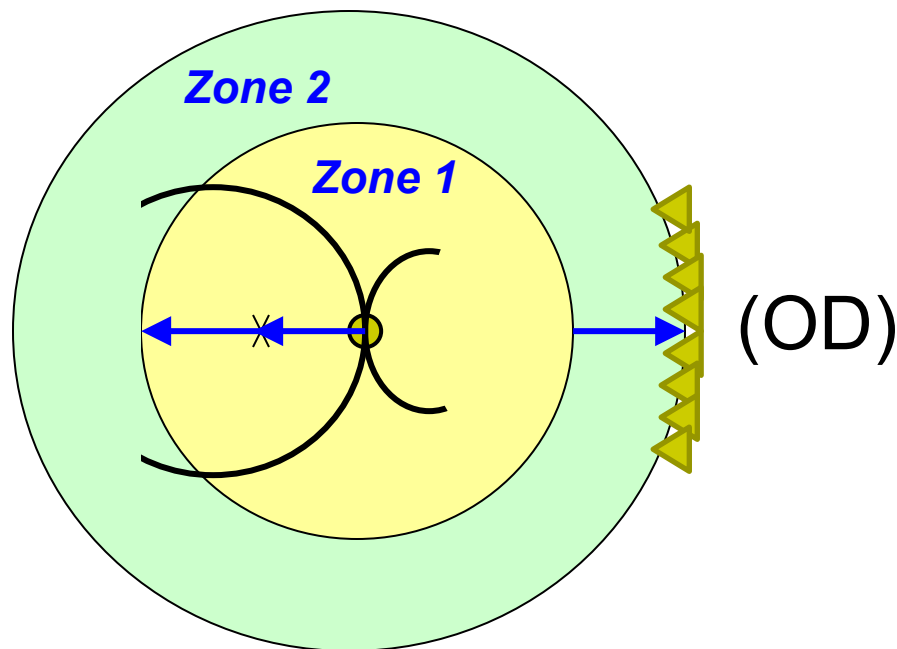


A

- *ROP classification*: Based on pathology **location** (**zone**), **appearance** (*stage*), and **plus disease** status:

- **Location**

- *Zone 1*: Circle around ONH w/ radius 2x disc-fovea distance
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- *Zone 3*

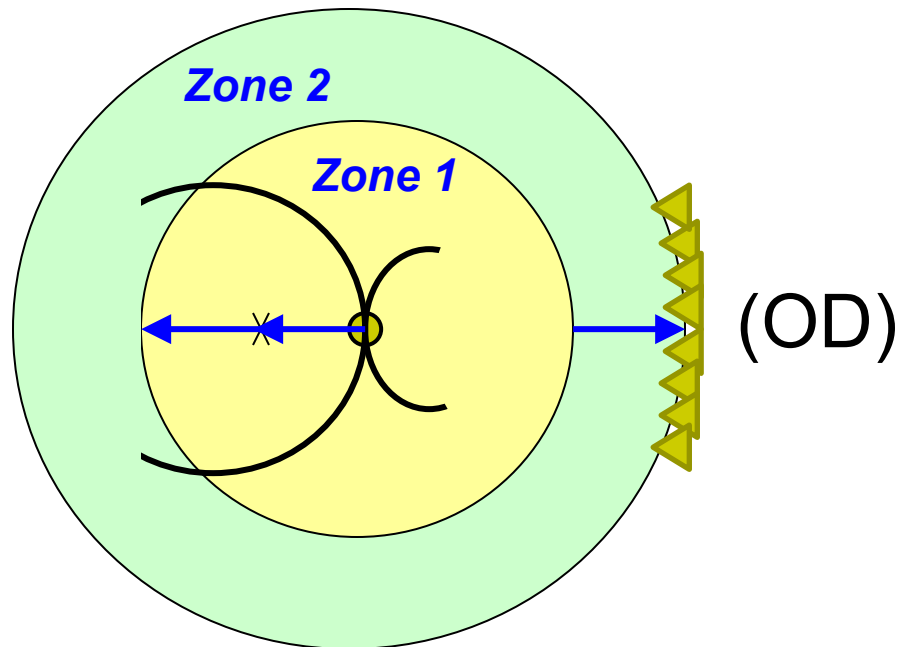


Q

- *ROP classification*: Based on pathology **location** (**zone**), **appearance** (*stage*), and **plus disease** status:

- **Location**

- Zone 1: Circle around ONH w/ radius 2x disc-fovea distance
- Zone 2: Edge of Zone 1 to nasal ora, and around temporally
- Zone 3:

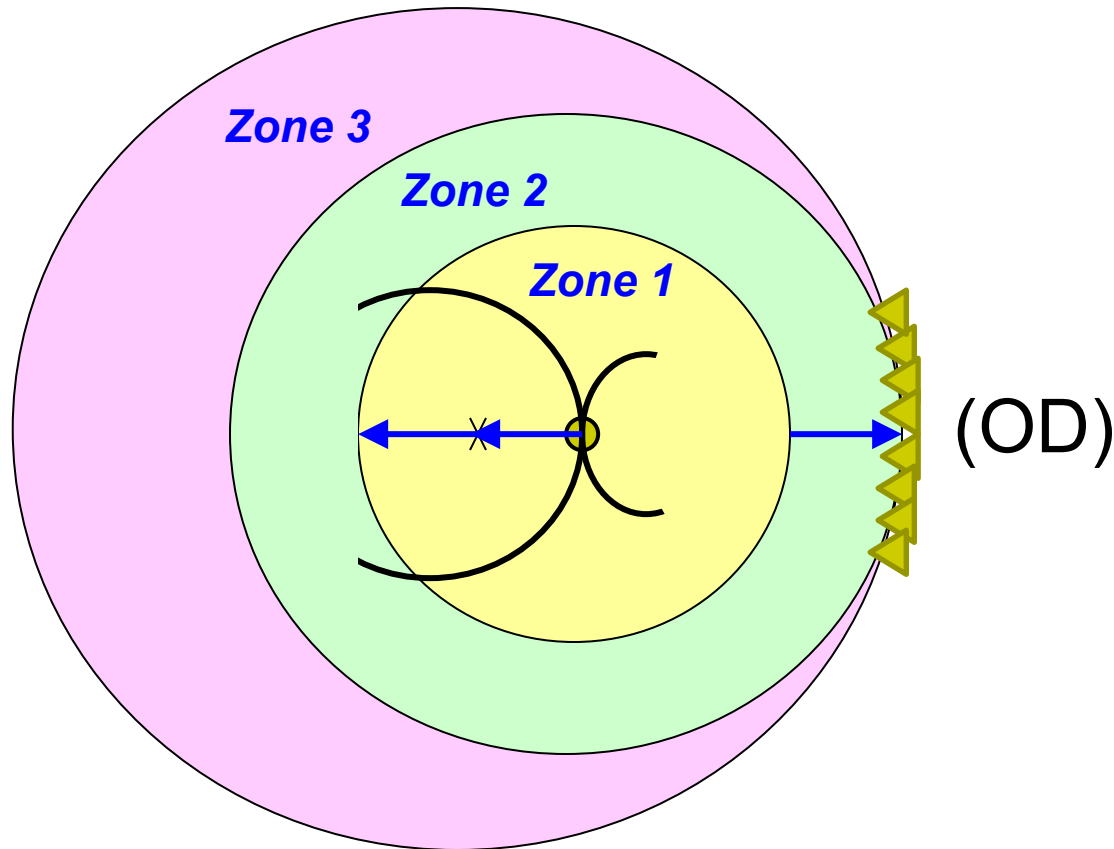


A

- *ROP classification*: Based on pathology **location** (**zone**), **appearance** (**stage**), and **plus disease** status:

- **Location**

- *Zone 1*: Circle around ONH w/ radius 2x disc-fovea distance
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- *Zone 3*: Residual crescent anterior to Zone 2



Q

- **ROP classification**: Based on pathology **location** (**zone**), **appearance** (**stage**), and **plus disease** status:

- **Location**

- *Zone 1*: Circle around ONH w/ radius 2x disc-fovea distance
- *Zone 2*: Edge of Zone 1 to nasal ora, and around temporally
- *Zone 3*: Residual crescent anterior to Zone 2

- **Appearance**

- *Stage 1*: 
- *Stage 2*
- *Stage 3*
- *Stage 4*
- *Stage 5*

A

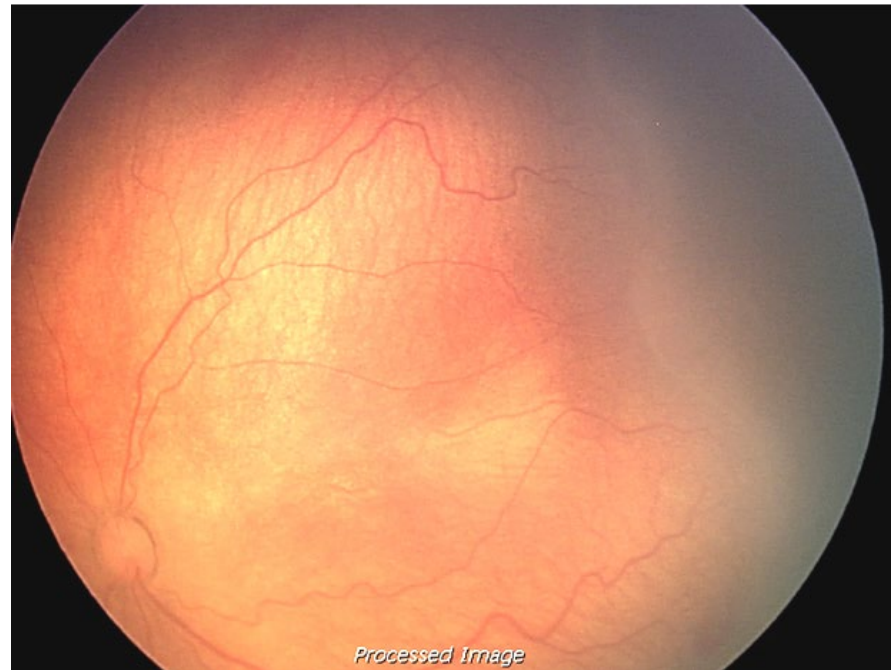
- **ROP classification**: Based on pathology **location** (**zone**), **appearance** (**stage**), and **plus disease** status:

- **Location**

- *Zone 1*: Circle around ONH w/ radius 2x disc-fovea distance
- *Zone 2*: Edge of Zone 1 to nasal ora, and around temporally
- *Zone 3*: Residual crescent anterior to Zone 2

- **Appearance**

- *Stage 1*: Demarcation line
- *Stage 2*
- *Stage 3*
- *Stage 4*
- *Stage 5*



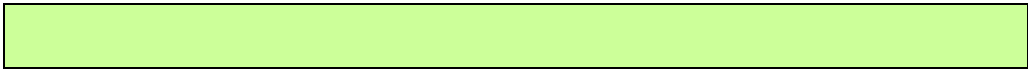
Q

- *ROP classification*: Based on pathology **location** (**zone**), **appearance** (**stage**), and **plus disease** status:

- **Location**

- *Zone 1*: Circle around ONH w/ radius 2x disc-fovea distance
- *Zone 2*: Edge of Zone 1 to nasal ora, and around temporally
- *Zone 3*: Residual crescent anterior to Zone 2

- **Appearance**

- *Stage 1*: Demarcation line
- *Stage 2*: 
- *Stage 3*
- *Stage 4*
- *Stage 5*

A

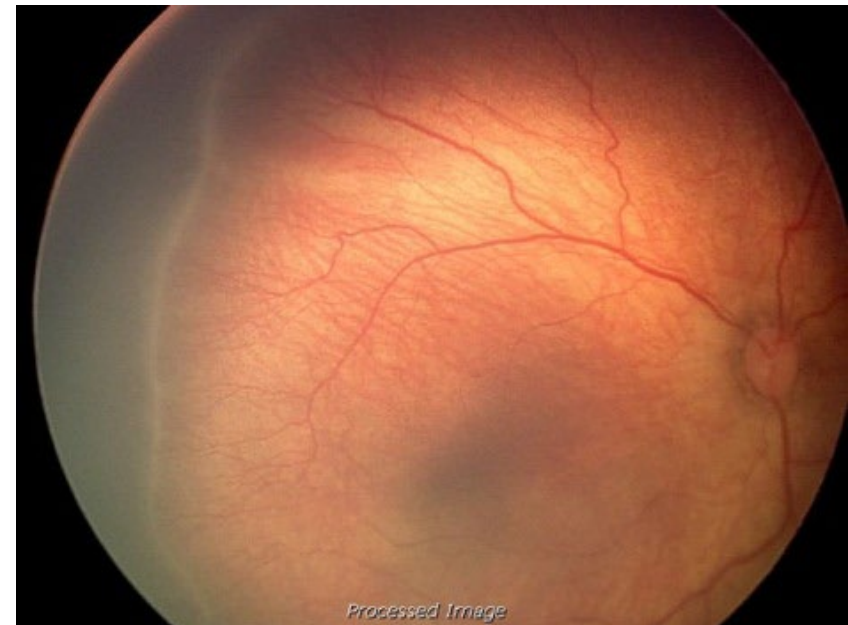
- **ROP classification**: Based on pathology **location** (**zone**), **appearance** (**stage**), and **plus disease** status:

- **Location**

- **Zone 1**: Circle around ONH w/ radius 2x disc-fovea distance
- **Zone 2**: Edge of Zone 1 to nasal ora, and around temporally
- **Zone 3**: Residual crescent anterior to Zone 2

- **Appearance**

- **Stage 1**: Demarcation line
- **Stage 2**: Elevated line ('ridge') +/- small tufts of neo




Q

- *ROP classification*: Based on pathology **location** (**zone**), **appearance** (**stage**), and **plus disease** status:

- **Location**

- *Zone 1*: Circle around ONH w/ radius 2x disc-fovea distance
- *Zone 2*: Edge of Zone 1 to nasal ora, and around temporally
- *Zone 3*: Residual crescent anterior to Zone 2

- **Appearance**

- *Stage 1*: Demarcation line
- *Stage 2*: Elevated line ('ridge') +/- small tufts of neo
- *Stage 3*: 
- *Stage 4*
- *Stage 5*

A

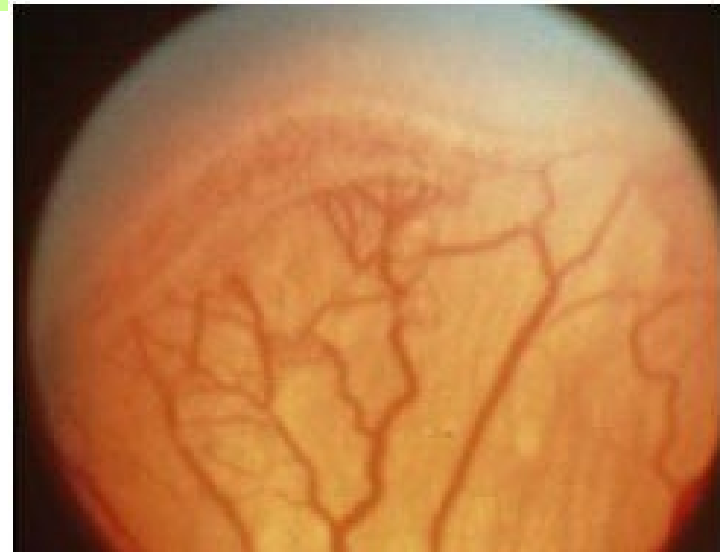
- **ROP classification**: Based on pathology **location** (**zone**), **appearance** (**stage**), and **plus disease** status:

- **Location**

- **Zone 1**: Circle around ONH w/ radius 2x disc-fovea distance
- **Zone 2**: Edge of Zone 1 to nasal ora, and around temporally
- **Zone 3**: Residual crescent anterior to Zone 2

- **Appearance**

- **Stage 1**: **Demarcation line** (ILM = internal limiting membrane)
- **Stage 2**: **Elevated line ('ridge') +/- small tufts of neo**
- **Stage 3**: **Ridge with extensive neo growing through ILM**



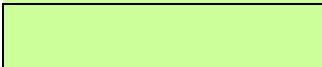
Q

- **ROP classification**: Based on pathology **location** (**zone**), **appearance** (**stage**), and **plus disease** status:

- **Location**

- *Zone 1*: Circle around ONH w/ radius 2x disc-fovea distance
- *Zone 2*: Edge of Zone 1 to nasal ora, and around temporally
- *Zone 3*: Residual crescent anterior to Zone 2

- **Appearance**

- *Stage 1*: Demarcation line
- *Stage 2*: Elevated line ('ridge') +/- small tufts of neo
- *Stage 3*: Ridge with extensive neo growing through ILM
- *Stage 4*: 
- *Stage 5*

A

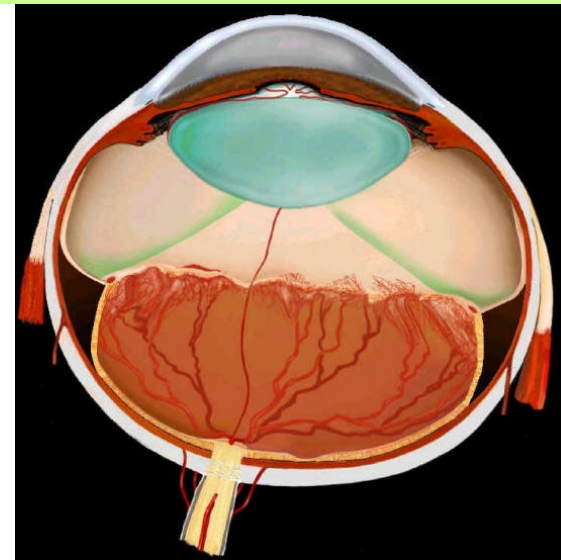
- **ROP classification**: Based on pathology **location** (zone), **appearance** (stage), and **plus disease** status:

- **Location**

- *Zone 1*: Circle around ONH w/ radius 2x disc-fovea distance
- *Zone 2*: Edge of Zone 1 to nasal ora, and around temporally
- *Zone 3*: Residual crescent anterior to Zone 2

- **Appearance**

- *Stage 1*: Demarcation line
- *Stage 2*: Elevated line ('ridge') +/- small tufts of neo
- *Stage 3*: Ridge with extensive neo growing through ILM
- *Stage 4*: Subtotal RD
- *Stage 5*



Q

- **ROP classification**: Based on pathology **location** (**zone**), **appearance** (**stage**), and **plus disease** status:

- **Location**

- *Zone 1*: Circle around ONH w/ radius 2x disc-fovea distance
- *Zone 2*: Edge of Zone 1 to nasal ora, and around temporally
- *Zone 3*: Residual crescent anterior to Zone 2

- **Appearance**

- *Stage 1*: Demarcation line
- *Stage 2*: Elevated line ('ridge') +/- small tufts of neo
- *Stage 3*: Ridge with extensive neo growing through ILM
- *Stage 4*: **Subtotal RD**
- *Stage 5*



Stage 4 is divided into two substages:

4a: RD with macula...

4b: RD with macula...

A

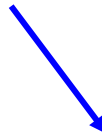
- **ROP classification**: Based on pathology **location** (**zone**), **appearance** (**stage**), and **plus disease** status:

- **Location**

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- **Appearance**

- *Stage 1*: Demarcation line
- *Stage 2*: Elevated line ('ridge') +/- small tufts of neo
- *Stage 3*: Ridge with extensive neo growing through ILM
- *Stage 4*: **Subtotal RD**
- *Stage 5*



Stage 4 is divided into two substages:

*4a: RD with macula...**on***

*4b: RD with macula...**off***

Q

- **ROP classification**: Based on pathology **location** (**zone**), **appearance** (**stage**), and **plus disease** status:

- **Location**

- *Zone 1*: Circle around ONH w/ radius 2x disc-fovea distance
- *Zone 2*: Edge of Zone 1 to nasal ora, and around temporally
- *Zone 3*: Residual crescent anterior to Zone 2

- **Appearance**

- *Stage 1*: Demarcation line
- *Stage 2*: Elevated line ('ridge') +/- small tufts of neo
- *Stage 3*: Ridge with extensive neo growing through ILM
- *Stage 4*: Subtotal RD
- *Stage 5*:

A

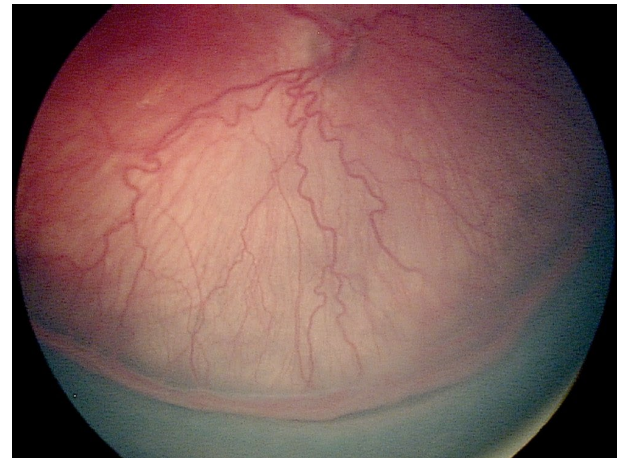
- **ROP classification**: Based on pathology **location** (**zone**), **appearance** (**stage**), and **plus disease** status:

- **Location**

- *Zone 1*: Circle around ONH w/ radius 2x disc-fovea distance
- *Zone 2*: Edge of Zone 1 to nasal ora, and around temporally
- *Zone 3*: Residual crescent anterior to Zone 2

- **Appearance**

- *Stage 1*: Demarcation line
- *Stage 2*: Elevated line ('ridge') +/- small tufts of neo
- *Stage 3*: Ridge with extensive neo growing through ILM
- *Stage 4*: Subtotal RD
- *Stage 5*: Total RD



Q

- **ROP classification**: Based on pathology **location** (**zone**), **appearance** (**stage**), and **plus disease** status:

- **Location**

- **Zone 1**: Circle around ONH w/ radius 2x disc-fovea distance
- **Zone 2**: Edge of Zone 1 to nasal ora, and around temporally
- **Zone 3**: Residual crescent anterior to Zone 2

- **Appearance**

- **Stage 1**: Demarcation line
- **Stage 2**: Elevated line ('ridge') +/- small tufts of neo
- **Stage 3**: Ridge with extensive neo growing through ILM
- **Stage 4**: Subtotal RD
- **Stage 5**: **Total RD**

What description is usually applied to the Stage 5 total RD?

A

- **ROP classification**: Based on pathology **location** (**zone**), **appearance** (**stage**), and **plus disease** status:

- **Location**

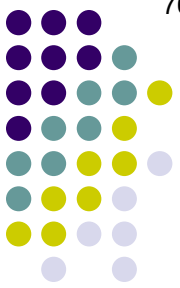
- **Zone 1**: Circle around ONH w/ radius 2x disc-fovea distance
- **Zone 2**: Edge of Zone 1 to nasal ora, and around temporally
- **Zone 3**: Residual crescent anterior to Zone 2

- **Appearance**

- **Stage 1**: Demarcation line
- **Stage 2**: Elevated line ('ridge') +/- small tufts of neo
- **Stage 3**: Ridge with extensive neo growing through ILM
- **Stage 4**: Subtotal RD
- **Stage 5**: **Total RD**

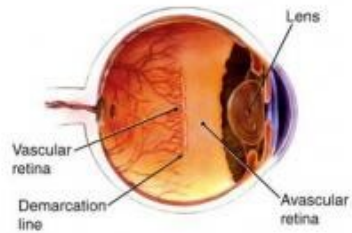
What description is usually applied to the Stage 5 total RD?

It is described as a 'funnel' RD

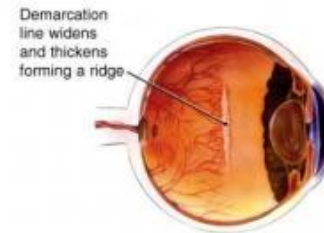


RETINOPATHY OF PREMATURITY

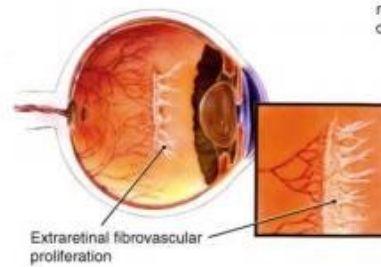
STAGE ONE



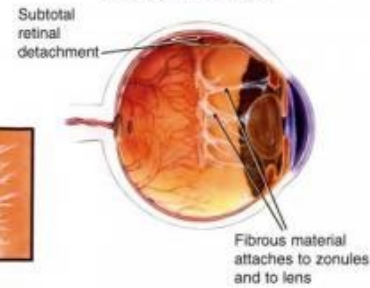
STAGE TWO



STAGE THREE



STAGE FOUR



STAGE FIVE RETINOPATHY "Retrolental Fibroplasia"



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ROP stages

Q

- *ROP classification*: Based on pathology **location** (**zone**), **appearance** (**stage**), and **plus disease** status:

- **Location**

- *Zone 1*: Circle around ONH w/ radius 2x disc-fovea distance
- *Zone 2*: Edge of Zone 1 to nasal ora, and around temporally
- *Zone 3*: Residual crescent anterior to Zone 2

- **Appearance**

- *Stage 1*: Demarcation line
- *Stage 2*: Elevated line ('ridge') +/- small tufts of neo
- *Stage 3*: Ridge with extensive neo growing through ILM
- *Stage 4*: Subtotal **RD**
- *Stage 5*: Total **RD**

What are the three basic types of retinal detachment (generally speaking; not specific to ROP)?

A

- *ROP classification*: Based on pathology **location** (**zone**), **appearance** (**stage**), and **plus disease** status:

- **Location**

- *Zone 1*: Circle around ONH w/ radius 2x disc-fovea distance
- *Zone 2*: Edge of Zone 1 to nasal ora, and around temporally
- *Zone 3*: Residual crescent anterior to Zone 2

- **Appearance**

- *Stage 1*: Demarcation line
- *Stage 2*: Elevated line ('ridge') +/- small tufts of neo
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- *Stage 4*: **Subtotal RD**
- *Stage 5*: **Total RD**

What are the three basic types of retinal detachment (generally speaking; not specific to ROP)?

Rhegmatogenous, exudative and tractional

Q

- *ROP classification*: Based on pathology **location** (**zone**), **appearance** (**stage**), and **plus disease** status:

- **Location**

- *Zone 1*: Circle around ONH w/ radius 2x disc-fovea distance
- *Zone 2*: Edge of Zone 1 to nasal ora, and around temporally
- *Zone 3*: Residual crescent anterior to Zone 2

- **Appearance**

- *Stage 1*: Demarcation line
- *Stage 2*: Elevated line ('ridge') +/- small tufts of neo
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- *Stage 4*: **Subtotal RD**
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What are the three basic types of retinal detachment (generally speaking; not specific to ROP)?

Rhegmatogenous, exudative and tractional

Which sort of RD occurs in ROP?

A

- *ROP classification*: Based on pathology **location** (**zone**), **appearance** (**stage**), and **plus disease** status:

- **Location**

- *Zone 1*: Circle around ONH w/ radius 2x disc-fovea distance
- *Zone 2*: Edge of Zone 1 to nasal ora, and around temporally
- *Zone 3*: Residual crescent anterior to Zone 2

- **Appearance**

- *Stage 1*: Demarcation line
- *Stage 2*: Elevated line ('ridge') +/- small tufts of neo
- *Stage 3*: Ridge with extensive neo growing through ILM
- *Stage 4*: **Subtotal RD**
- *Stage 5*: **Total RD**

What are the three basic types of retinal detachment (generally speaking; not specific to ROP)?

Rhegmatogenous, exudative and tractional

Which sort of RD occurs in ROP?

Tractional RD (TRD)

Q

- **ROP classification**: Based on pathology **location** (**zone**), **appearance** (**stage**), and **plus disease** status:

- **Location**

- *Zone 1*: Circle around ONH w/ radius 2x disc-fovea distance
- *Zone 2*: Edge of Zone 1 to nasal ora, and around temporally
- *Zone 3*: Residual crescent anterior to Zone 2

- **Appearance**

- *Stage 1*: Demarcation line
- *Stage 2*: Elevated line ('ridge') +/- small tufts of neo
- *Stage 3*: Ridge with extensive neo growing through ILM
- *Stage 4*: Subtotal RD
- *Stage 5*: Total RD

- **Presence/absence of plus disease**

- *Plus disease* = two/words retinal vessels

A

- **ROP classification**: Based on pathology **location** (**zone**), **appearance** (**stage**), and **plus disease** status:

- **Location**

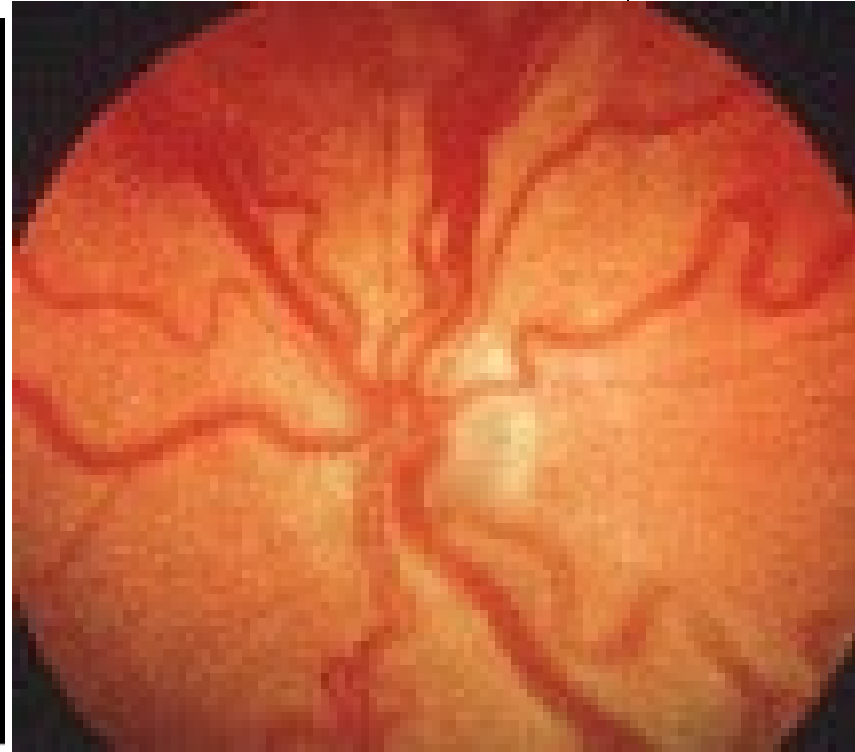
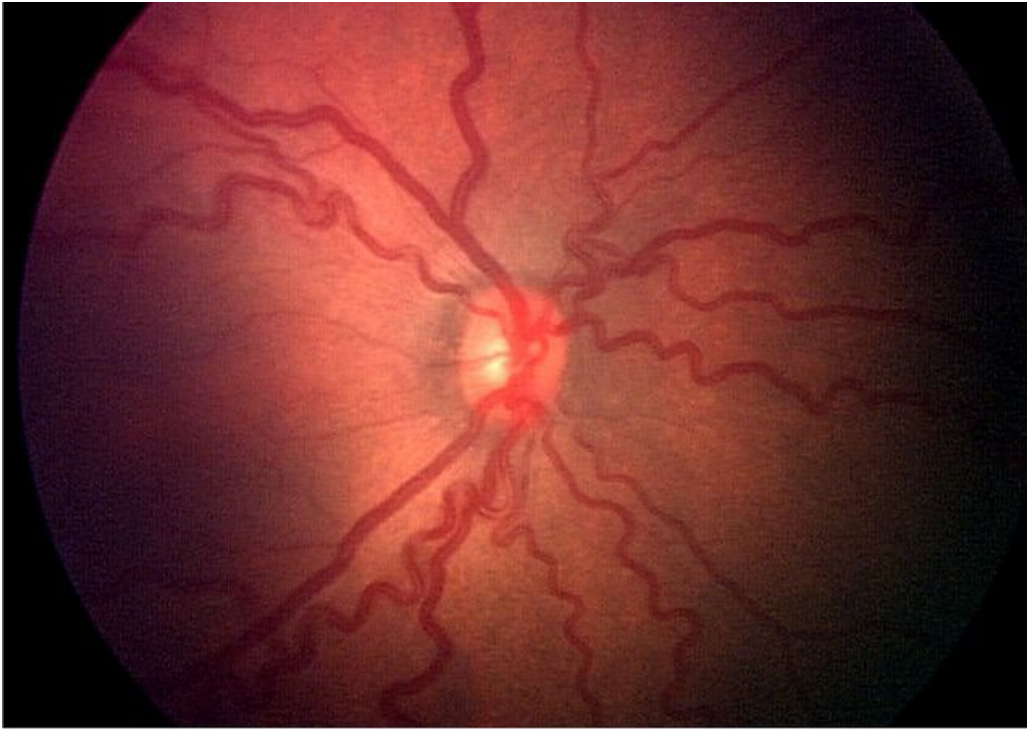
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- **Appearance**

- *Stage 1*: Demarcation line
- *Stage 2*: Elevated line ('ridge') +/- small tufts of neo
- *Stage 3*: Ridge with extensive neo growing through ILM
- *Stage 4*: Subtotal RD
- *Stage 5*: Total RD

- **Presence/absence of plus disease**

- *Plus disease* = Dilated/tortuous retinal vessels



ROP: *Plus* disease

Q

- **ROP classification**: Based on pathology **location** (**zone**), **appearance** (**stage**), and **plus disease** status:

- **Location**

- *Zone 1*: Circle around ONH w/ radius 2x disc-fovea distance
- *Zone 2*: Edge of Zone 1 to nasal ora, and around temporally
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- **Appearance**

- *Stage 1*: Demarcation line
- *Stage 2*: Elevated line ('ridge') +/- small tufts of neo
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- *Stage 4*: Subtotal RD
- *Stage 5*: Total RD

- **Presence/absence of plus disease**

- *Plus disease* = Dilated/tortuous retinal vessels

How dilated/tortuous do the vessels need to be to qualify as plus disease?

A

- **ROP classification**: Based on pathology **location** (**zone**), **appearance** (**stage**), and **plus disease** status:

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- *Zone 1*: Circle around ONH w/ radius 2x disc-fovea distance
- *Zone 2*: Edge of Zone 1 to nasal ora, and around temporally
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- **Appearance**

- *Stage 1*: Demarcation line
- *Stage 2*: Elevated line ('ridge') +/- small tufts of neo
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- *Plus disease* = Dilated/tortuous retinal vessels

How dilated/tortuous do the vessels need to be to qualify as plus disease?

A standardized photo exists indicating the 'official' amount needed

Q

- **ROP classification**: Based on pathology **location** (**zone**), **appearance** (**stage**), and **plus disease** status:

- **Location**

- **Zone 1**: Circle around ONH w/ radius 2x disc-fovea distance
- **Zone 2**: Edge of Zone 1 to nasal ora, and around temporally
- **Zone 3**: Residual crescent anterior to Zone 2

- **Appearance**

- **Stage 1**: Demarcation line
- **Stage 2**: Elevated line ('ridge') +/- small tufts of neo
- **Stage 3**: Ridge with extensive neo growing through ILM
- **Stage 4**: Subtotal RD
- **Stage 5**: Total RD

- **Presence/absence of plus disease**

- **Plus disease** = Dilated/tortuous retinal vessels

How dilated/tortuous do the vessels need to be to qualify as plus disease?

A standardized photo exists indicating the 'official' amount needed

What if the vessels are definitely dilated/tortuous, but not to the extent indicated in the standardized photo?

A

- **ROP classification**: Based on pathology **location** (**zone**), **appearance** (**stage**), and **plus disease** status:

- **Location**

- **Zone 1**: Circle around ONH w/ radius 2x disc-fovea distance
- **Zone 2**: Edge of Zone 1 to nasal ora, and around temporally
- **Zone 3**: Residual crescent anterior to Zone 2

- **Appearance**

- **Stage 1**: Demarcation line
- **Stage 2**: Elevated line ('ridge') +/- small tufts of neo
- **Stage 3**: Ridge with extensive neo growing through ILM
- **Stage 4**: Subtotal RD
- **Stage 5**: Total RD

- **Presence/absence of plus disease**

- **Plus disease** = Dilated/tortuous retinal vessels

How dilated/tortuous do the vessels need to be to qualify as plus disease?

A standardized photo exists indicating the 'official' amount needed

What if the vessels are definitely dilated/tortuous, but not to the extent indicated in the standardized photo?
This is referred to as **Pre-Plus disease**

Q

- **ROP classification**: Based on pathology **location** (**zone**), **appearance** (**stage**), and **plus disease** status:

- **Location**

- *Zone 1*: Circle around ONH w/ radius 2x disc-fovea distance
- *Zone 2*: Edge of Zone 1 to nasal ora, and around temporally
- *Zone 3*: Residual crescent anterior to Zone 2

- **Appearance**

- *Stage 1*: Demarcation line
- *Stage 2*: Elevated line ('ridge') +/- small tufts of neo
- *Stage 3*: Ridge with extensive neo growing through ILM
- *Stage 4*: Subtotal RD
- *Stage 5*: Total RD

- **Presence/absence of plus disease**

- *Plus disease* = Dilated/tortuous retinal vessels
 - Indicates two words is taking place

A

- **ROP classification**: Based on pathology **location** (**zone**), **appearance** (**stage**), and **plus disease** status:

- **Location**

- *Zone 1*: Circle around ONH w/ radius 2x disc-fovea distance
- *Zone 2*: Edge of Zone 1 to nasal ora, and around temporally
- *Zone 3*: Residual crescent anterior to Zone 2

- **Appearance**

- *Stage 1*: Demarcation line
- *Stage 2*: Elevated line ('ridge') +/- small tufts of neo
- *Stage 3*: Ridge with extensive neo growing through ILM
- *Stage 4*: Subtotal RD
- *Stage 5*: Total RD

- **Presence/absence of plus disease**

- *Plus disease* = Dilated/tortuous retinal vessels
 - Indicates arteriovenous shunting is taking place

Q

- **ROP classification**: Based on pathology **location** (**zone**), **appearance** (**stage**), and **plus disease** status:

- **Location**

- *Zone 1*: Circle around ONH w/ radius 2x disc-fovea distance
- *Zone 2*: Edge of Zone 1 to nasal ora, and around temporally
- *Zone 3*: Residual crescent anterior to Zone 2

- **Appearance**

- *Stage 1*: Demarcation line
- *Stage 2*: Elevated line ('ridge') +/- small tufts of neo
- *Stage 3*: Ridge with extensive neo growing through ILM
- *Stage 4*: Subtotal RD
- *Stage 5*: Total RD

- **Presence/absence of plus disease**

- *Plus disease* = Dilated/tortuous retinal vessels
 - Indicates arteriovenous shunting is taking place
 - Strong indicator that disease one word is occurring

A

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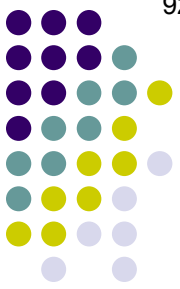
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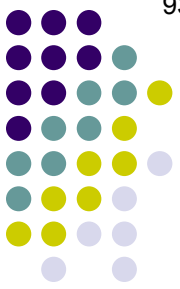
ROP: Treatment Considerations



- This is the **outdated** definition of when to treat ROP (so-called *Threshold disease*):
 - 5 contiguous clock hours or 8 noncontiguous hours of Stage 3 disease (or worse) in Zone I or II, associated with plus disease

Q

ROP: Treatment Considerations

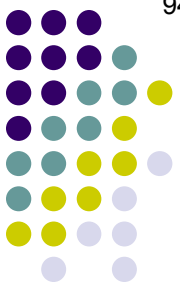


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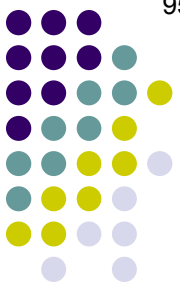
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The **CRYO-ROP** study

Q

ROP: Treatment Considerations

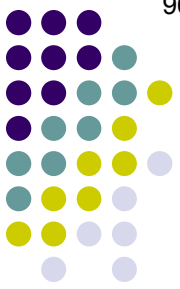


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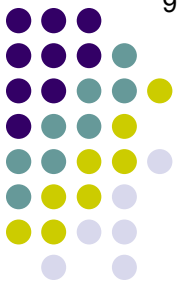
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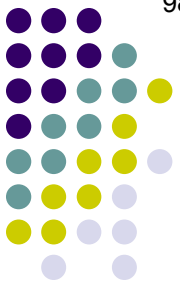
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Treatment is indicated if the ROP meets one of three criteria:

- 1.
- or
- 2.
- or
- 3.

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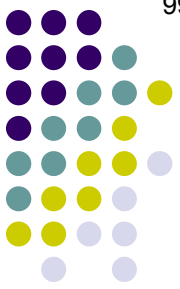
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What was the name of the study from which these treatment guidelines were developed?

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1. Zone 1, any Stage, with Plus disease
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What was the name of the study from which these treatment guidelines were developed?
 The **ET-ROP** (Early Treatment of Retinopathy of Prematurity) study



ROP: Treatment Considerations

- This is the **outdated** definition of when to treat ROP (so-called *Threshold disease*):
 - 5 contiguous clock hours or 8 noncontiguous hours

The motivating factor behind the **ET-ROP** was to see whether earlier intervention could improve upon these dismal results

What's wrong with these criteria for treatment? Why don't we use them anymore?

Research indicated **that less than 13% of children treated via these criteria went on to have 20/40 or better vision in treated eyes!** That's not a very good outcome, so these criteria have been revised.

What are the new criteria?

Treatment is indicated if the ROP meets one of three criteria:

1. Zone 1, any Stage, with Plus disease ('Rush disease')
- or
2. Zone 1, Stage 3, with or without Plus disease
- or
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Per the ET-ROP, disease meeting these criteria are known as what 'type' of ROP?

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Type I



ROP: Treatment Considerations

- This is the *outdated* definition of when to treat ROP (so-called *Threshold disease*):

- 5 contiguous clock hours or 8 noncontiguous hours of Stage 3 disease (or worse) in Zone I or II, associated with plus disease**

Note that disease meeting ET-ROP criteria for treatment would not have met threshold under CRYO-ROP criteria. For this reason, the new criteria are sometimes referred to as '*pre-threshold Type I ROP*'

What are the new criteria?

Treatment is indicated if the ROP meets one of three criteria:

- 1. Zone 1, any Stage, with Plus disease**
- or*
- 2. Zone 1, Stage 3, with or without Plus disease**
- or*
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By what special name is 'Zone 1 + Plus disease' known?

1. Zone 1, any Stage, with Plus disease

or

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any more?
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Why is it called Rush disease?

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Why is it called Rush disease?

Because these eyes are at especially high risk of very rapid progression to TRD

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Which infants are at particular risk for developing Rush disease?

Those weighing under 1000 grams

1. Zone 1, any Stage, with Plus disease

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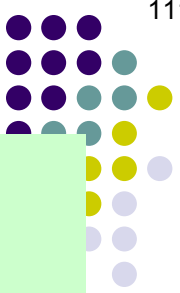
or

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ROP: Treatment Considerations



Another term is used for aggressive posterior ROP--what is it?

By what special name is 'Zone 1 + Plus disease' known?

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Why is it called Rush disease?

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Which infants are at particular risk for developing Rush disease?

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ROP: Treatment Considerations

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Is APROP simply another name for Rush disease?

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While the terms are sometimes used interchangeably, the clinical appearance and behavior of APROP can differ from that of Rush dz. APROP is characterized by the presence of neovascular fronds lying flat on the retinal surface (ie, without a ridge) in Zone 1 or posterior Zone 2. Active A-V shunting is the rule.

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- 1)
- 2)
- 3)
- 4)

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- 1) Progressing directly from Stage 1 to Stage 3 disease;
- 2) very rapid progression--Stage 1 to 3 (or even 4) in a matter of days;
- 3) a proclivity to recur despite seemingly adequate treatment; and
- 4) a less-than-robust response to conventional laser treatment

By what special name is 'Zone 1 + Plus disease' known?

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Why is it called Rush disease?

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What's wrong with these criteria for treatment? Why don't we use them anymore?
 Research indicated that less than 13% of children treated via these criteria went on to have 20/40 or better vision in treated eyes! That's not a very good outcome, so these criteria have been replaced.

What is the conventional treatment for ROP?

What are the new criteria?
Treatment is indicated if

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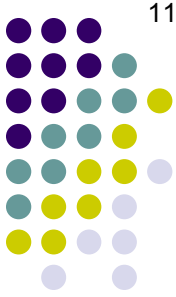
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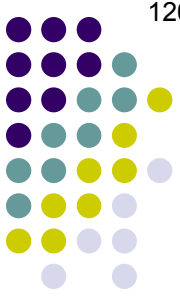
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Which is preferred, cryo or laser?

A/Q

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Which is preferred, cryo or laser?

Most clinicians prefer laser; it is less traumatic to the eye, and less risky (5% of infants undergoing cryo for ROP will have

bad complication (2 words)

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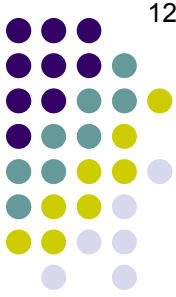
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Q

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What are the other advantages of laser over cryo?

--Less bad thing to tissue

--

--

--

Treatment is indicated if

1. Zone 1, any Stage, with
or

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or

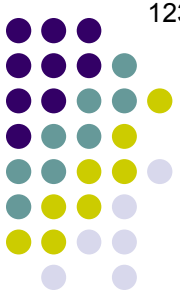
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What are the other advantages of laser over cryo?

- Less trauma to tissue
- Easier to treat posterior locations
-
-

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- or
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- This is the **outdated** definition of when to treat ROP (so-called *Threshold disease*):
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- Less trauma to tissue
- Easier to treat posterior locations
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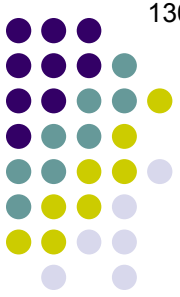
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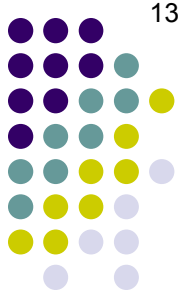
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Although it must be stressed that laser treatment is not wholly benign—issues with intra-operative apnea and/or adverse cardiac events have been reported, as have sequelae including cataract and glaucoma.

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 Research indicated that less than 13% of children treated via these criteria went on to have 20/40 or better vision in treated eyes! That's not a very good outcome, so these criteria have been abandoned.

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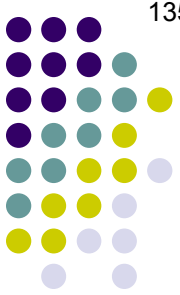
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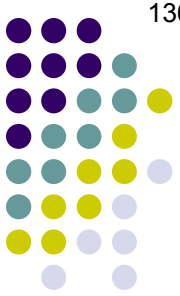
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As mentioned previously, APROP is notorious for its poor response to convention laser tx (CLT)

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ROP: Treatment Considerations

Another term is used for aggressive posterior ROP--what is it?

It is called **Aggressive Posterior ROP (APROP)**

Is APROP simply another name for Rush disease?

While the terms are sometimes used interchangeably, the clinical appearance and behavior of APROP can differ from that of Rush dz. APROP is characterized by the presence of neovascular fronds lying flat on the retinal surface (ie, without a ridge) in Zone 1 or posterior Zone 2. Active A-V shunting is the rule. APROP is notorious for four unfortunate tendencies:

- 1) Progressing directly from Stage 1 to Stage 3 disease;
- 2) very rapid progression--Stage 1 to 3 (or even 4) in a matter of days;
- 3) a proclivity to recur despite seemingly adequate treatment; and
- 4) **a less-than-robust response to conventional laser treatment**

Recall this info from
a previous slide

What's wrong with these criteria for treatment? Why don't we use them any more?

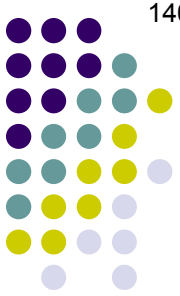
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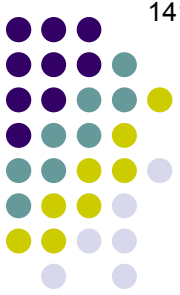
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Pts received a single intravitreal injection of 0.625 mg bevacizumab (note that this is ½ the usual adult dose), or CLT

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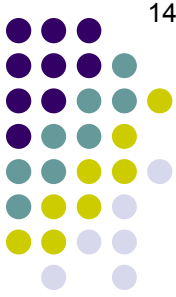
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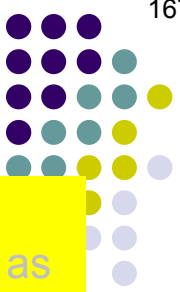
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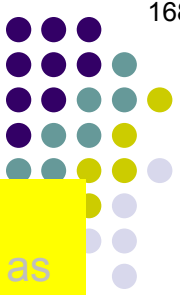
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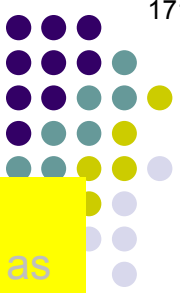
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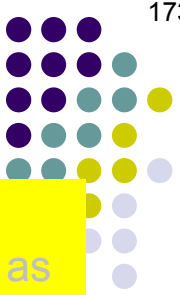
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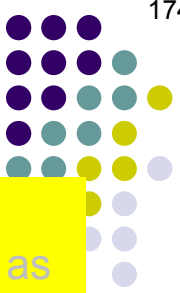
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What is perhaps the main concern re giving bevacizumab to neonates?

intravitreal bevacizumab was only 6%.

What about in eyes with posterior Zone 2 APROP?

The recurrence rates did not differ statistically

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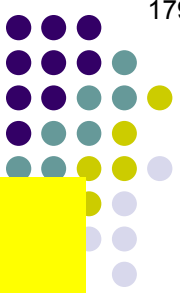
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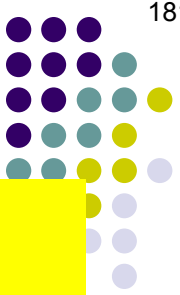
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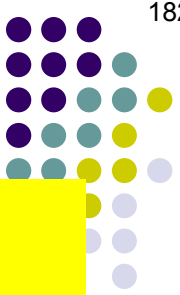
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On the other hand, there is no reason to think CLT has any long-term effects outside the eye.
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- This is the **outdated** definition of when to treat ROP (so-called *Threshold disease*):
 - 5 contiguous clock hours or 8 noncontiguous hours of Stage 3 disease (or worse) in Zone I or II, associated with plus disease

What's wrong with these criteria for treatment? Why don't we use them anymore?
 Research indicated that less than 13% of children treated via these criteria went on to have 20/40 or better vision in treated eyes! That's not a very good outcome, so these criteria have been replaced.

What if the pt develops a TRD--how is that managed?

What are the new criteria?

Treatment is indicated if t

1. Zone 1, any Stage, with or

2. Zone 1, Stage 3, with or

3. Zone 2, Stage 2 or 3, with Plus disease

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PPV and/or scleral buckle

(PPV = Pars plana vitrectomy)

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Is it effective?

Not so much. Only 30% of cases achieve anatomic reattachment; of these, only 25% are still attached at 5 years, and only 10% have ambulatory vision



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What's the new definition?
 Treat

Once a decision to treat has been made, how long can it be deferred?

1. Zone 1, any Stage, with Plus disease

or

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or

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 - 5 contiguous clock hours or 8 noncontiguous hours of Stage 3 disease (or worse) in Zone I or II, associated with plus disease

What's wrong with these criteria for treatment? Why don't we use them anymore?
 Research indicated that less than 13% of children treated via these criteria went on to have 20/40 or better vision in treated eyes! That's not a very good outcome, so these criteria have been revised.

What's the new definition?
 Treat

Once a decision to treat has been made, how long can it be deferred?
When possible, treatment should be initiated within 72 hours

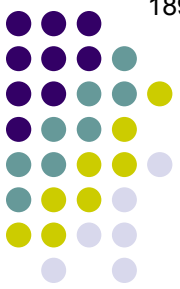
1. Zone 1, any Stage, with Plus disease

or

2. Zone 1, Stage 3, with or without Plus disease

or

3. Zone 2, Stage 2 or 3, with Plus disease



Q

ROP: Screening and Follow-Up

- ***ROP screening***

- ***Who?***

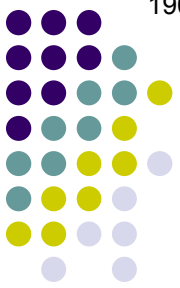
- Screen all infants...

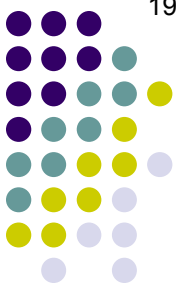
- ...with a birth weight of less than # gm

A

ROP: Screening and Follow-Up

- ***ROP screening***
 - ***Who?***
 - Screen all infants...
 - ...with a birth weight of less than 1500 gm





Q

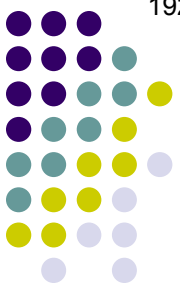
ROP: Screening and Follow-Up

- ***ROP screening***

- ***Who?***

- Screen all infants...

- ...with a birth weight of less than 1500 gm, ***and/or***
- ...whose gestational age at birth was # weeks or less



A

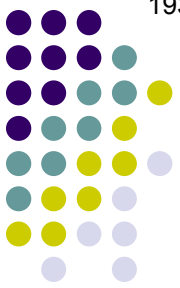
ROP: Screening and Follow-Up

- ***ROP screening***

- ***Who?***

- Screen all infants...

- ...with a birth weight of less than 1500 gm, ***and/or***
- ...whose gestational age at birth was 30 weeks or less



Q

ROP: Screening and Follow-Up

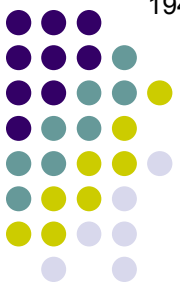
- *ROP screening*

- *Who?*

- Screen all infants...

- ...with a birth weight of less than 1500 gm, *and/or*
- ...whose gestational age at birth was 30 weeks or less

What about infants >1500 gm and/or with gestational age >30 weeks? Should they be screened?



A

ROP: Screening and Follow-Up

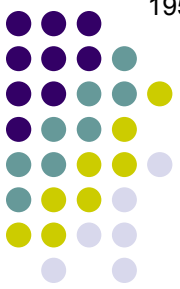
- *ROP screening*

- *Who?*

- Screen all infants...

- ...with a birth weight of less than 1500 gm, *and/or*
- ...whose gestational age at birth was 30 weeks or less

What about infants >1500 gm and/or with gestational age >30 weeks? Should they be screened?
Not as a general rule. However, the guidelines state that such infants should be screened if/when their neonatologist feels it is indicated



Q

ROP: Screening and Follow-Up

- **ROP screening**

- **Who?**

- Screen all infants...

- ...with a birth weight of less than 1500 gm, **and/or**
- ...whose gestational age at birth was 30 weeks or less

- **When?**

- Timing of first screen is a function of pt

A

• **ROP screening**

• **Who?**

• Screen all

- ...with
- ...whom

• **When?**

- Timing of first screen is a function of pt **age** (see *table*)

<i>Gestational age</i>	<i>Postmenstrual age</i>	<i>Chronologic age at time of first ROP screening</i>
22	31	9
23	31	8
24	31	7
25	31	6
26	31	5
27	31	4
28	32	4
29	33	4
30	34	4
31	35	4
32	36	4

Q

- **ROP screening**

- **Who?**

- Screen all

- ...with

- ...whom

- **When?**

<i>Gestational age</i>	<i>Postmenstrual age</i>	<i>Chronologic age at time of first ROP screening</i>
22	31	9
23	31	8
24	31	7
25	31	6
26	31	5
27	31	4
28	32	4
29	33	4
30	34	4
31	35	4
32	36	4

- Timing of first screen is a function of pt **age** (see table)

- Serious ROP rare before postmenstrual age **#** weeks, so this is the youngest age that requires screening

A

• ROP screening

• Who?

• Screen all

- ...with
- ...whom

• When?

<i>Gestational age</i>	<i>Postmenstrual age</i>	<i>Chronologic age at time of first ROP screening</i>
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27	31	4
28	32	4
29	33	4
30	34	4
31	35	4
32	36	4

- Timing of first screen is a function of pt **age** (see *table*)
 - Serious ROP rare before postmenstrual age **31** weeks, so this is the youngest age that requires screening

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27	31	4
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30	34	4
31	35	4
32	36	4

- Timing of first screen is a function of pt **age** (see *table*)
 - Serious ROP rare before postmenstrual age **31** weeks, so this is the youngest age that requires screening

Don't try and memorize the table! Instead, here is first-screen timing in a nutshell:

- **ROP screening**

- **Who?**

- Screen all

- ...with

- ...whom

- **When?**

Gestational age	Postmenstrual age	Chronologic age at time of first ROP screening
22	31	9
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- Timing of first screen is a function of pt **age** (see table)

- Serious ROP rare before postmenstrual age **31** weeks, so this is the youngest age that requires screening

Don't try and memorize the table! Instead, here is first-screen timing in a nutshell:
 If the infant's gestational age at birth was 27 weeks or younger, perform first screen at **postmenstrual age 31 weeks**,
 or

- **ROP screening**

- **Who?**

- Screen all

- ...with

- ...whom

- **When?**

Gestational age	Postmenstrual age	Chronologic age at time of first ROP screening
22	31	9
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30	34	4
31	35	4
32	36	4

- Timing of first screen is a function of pt **age** (see table)

- Serious ROP rare before postmenstrual age **31** weeks, so this is the youngest age that requires screening

Don't try and memorize the table! Instead, here is first-screen timing in a nutshell:

If the infant's gestational age at birth was 27 weeks or younger, perform first screen at

postmenstrual age 31 weeks,

or

If the infant's gestational age at birth was 28 weeks or older, perform first screen at

chronologic age 4 weeks



Q

ROP: Screening and Follow-Up

- **ROP screening**

- **Who?**

- Screen all infants...
 - ...with a birth weight of less than 1500 gm, **and/or**
 - ...whose gestational age at birth was 30 weeks or less

- **When?**

- Timing of first screen is a function of pt age (see table)
 - Serious ROP rare before postmenstrual age 31 weeks, so this is the youngest age that requires screening

- **How Often?**

- A single screening exam is sufficient if the retina is

three words



A

ROP: Screening and Follow-Up

- ***ROP screening***

- ***Who?***

- Screen all infants...

- ...with a birth weight of less than 1500 gm, ***and/or***
- ...whose gestational age at birth was 30 weeks or less

- ***When?***

- Timing of first screen is a function of pt age (see table)
 - Serious ROP rare before postmenstrual age 31 weeks, so this is the youngest age that requires screening

- ***How Often?***

- A single screening exam is sufficient if the retina is fully vascularized OU



Q

ROP: Screening and Follow-Up

● **ROP screening**

● **Who?**

- Screen all infants...
 - ...with a birth weight of less than 1500 gm, **and/or**
 - ...whose gestational age at birth was 30 weeks or less

● **When?**

- Timing of first screen is a function of pt age (see table)
 - Serious ROP rare before postmenstrual age 31 weeks, so this is the youngest age that requires screening

● **How Often?**

- A single screening exam is sufficient if the retina is fully vascularized OU
- Otherwise, ##, time period follow-up is indicated (depending upon exam findings)



A

ROP: Screening and Follow-Up

- ***ROP screening***

- ***Who?***

- Screen all infants...

- ...with a birth weight of less than 1500 gm, ***and/or***
- ...whose gestational age at birth was 30 weeks or less

- ***When?***

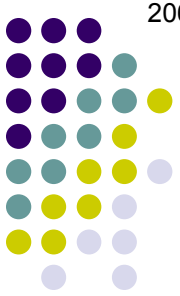
- Timing of first screen is a function of pt age (see table)
 - Serious ROP rare before postmenstrual age 31 weeks, so this is the youngest age that requires screening

- ***How Often?***

- A single screening exam is sufficient if the retina is fully vascularized OU
- Otherwise, 1 - 3 week follow-up is indicated (depending upon exam findings)

Q

ROP: Screening and Follow-Up



- ***Long-term follow-up:*** A child with ROP needs periodic follow-up beyond the newborn period because...

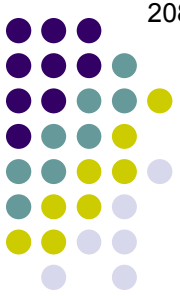
- pathology can lead to RD in decade(s) of life

A

ROP: Screening and Follow-Up



- ***Long-term follow-up:*** A child with ROP needs periodic follow-up beyond the newborn period because...
 - **Vitreoretinal traction** can lead to RD in **1st or 2nd decade**



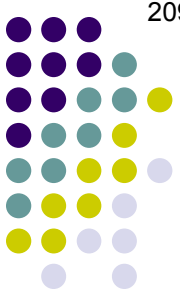
Q

ROP: Screening and Follow-Up

- ***Long-term follow-up:*** A child with ROP needs periodic follow-up beyond the newborn period because...
 - Vitreoretinal traction can lead to RD in 1st or 2nd decade
 - Amblyopia can result from pathology, and/or refractive problem, macular EOM problem

A

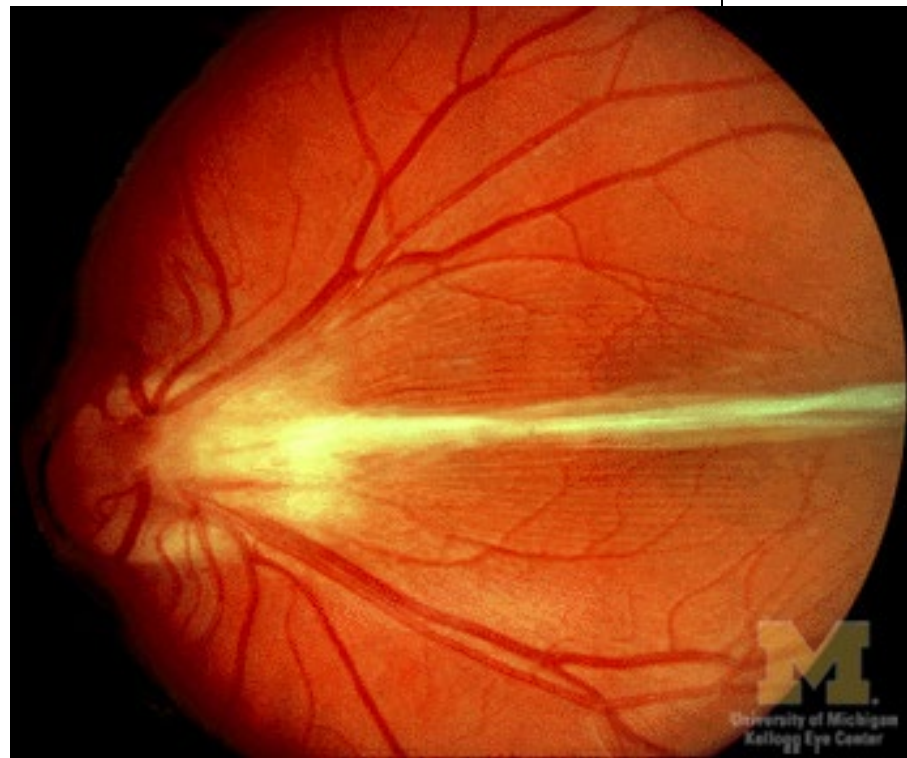
ROP: Screening and Follow-Up



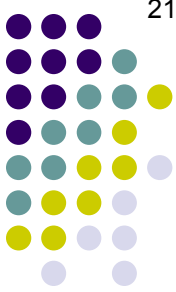
- ***Long-term follow-up:*** A child with ROP needs periodic follow-up beyond the newborn period because...
 - Vitreoretinal traction can lead to RD in 1st or 2nd decade
 - Amblyopia can result from high myopia, macular dragging, and/or strabismus



ROP: Screening and Follow-Up



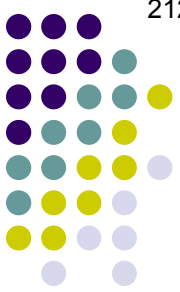
ROP: Macular dragging



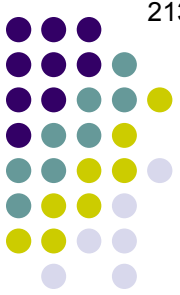
Q

ROP: Screening and Follow-Up

- ***Long-term follow-up:*** A child with ROP needs periodic follow-up beyond the newborn period because...
 - Vitreoretinal traction can lead to RD in 1st or 2nd decade
 - Amblyopia can result from high myopia, macular dragging, and/or strabismus
 - Macular dragging can produce pseudo EOM problem



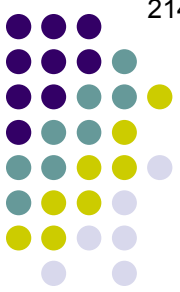
- ***Long-term follow-up:*** A child with ROP needs periodic follow-up beyond the newborn period because...
 - Vitreoretinal traction can lead to RD in 1st or 2nd decade
 - Amblyopia can result from high myopia, macular dragging, and/or strabismus
 - Macular dragging can produce pseudostrabismus



Q

ROP: Screening and Follow-Up

- ***Long-term follow-up:*** A child with ROP needs periodic follow-up beyond the newborn period because...
 - Vitreoretinal traction can lead to RD in 1st or 2nd decade
 - Amblyopia can result from high myopia, macular dragging, and/or strabismus
 - Macular dragging can produce pseudostrabismus
 - Will have positive exam finding in pseudo-EOM problem, but no exam finding on exam maneuver



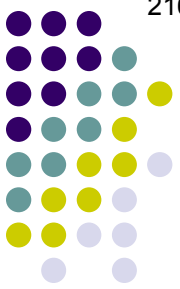
- ***Long-term follow-up:*** A child with ROP needs periodic follow-up beyond the newborn period because...
 - Vitreoretinal traction can lead to RD in 1st or 2nd decade
 - Amblyopia can result from high myopia, macular dragging, and/or strabismus
 - Macular dragging can produce pseudostrabismus
 - Will have positive angle kappa, but no shift on cover testing



ROP: Screening and Follow-Up


- ***Long-term follow-up:*** A child with ROP needs periodic follow-up beyond the newborn period because...
 - Vitreoretinal traction can lead to RD in 1st or 2nd decade
 - Amblyopia can result from high myopia, macular dragging, and/or strabismus
 - **Macular dragging** can produce pseudostrabismus
 - Will have positive angle kappa, but no shift on cover testing

Speaking of macular dragging...



Q

ROP: Screening and Follow-Up

- In addition to a dragged macula, ROP pts often have a dragged 



A

ROP: Screening and Follow-Up

- In addition to a dragged macula, ROP pts often have a dragged **disc**.



Q

ROP: **DDx**

- In addition to a dragged macula, ROP pts often have a dragged **disc**. *What three other clinical entities can give a similar picture?*

3 things that look like ROP:

- 1) ?
- 2) ?
- 3) ?

Hints forthcoming...



Q

ROP: DDx

- In addition to a dragged macula, ROP pts often have a dragged **disc**. *What three other clinical entities can give a similar picture?*

3 things that look like ROP:

1)

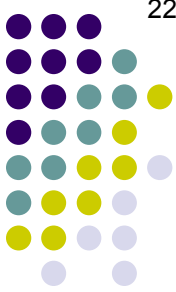
Hint: A phakomatosis (buzzterm: 'Splashed paint')

2) ?

3) ?

A

ROP: **DDx**



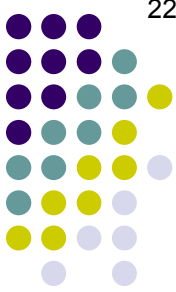
- In addition to a dragged macula, ROP pts often have a dragged **disc**. *What three other clinical entities can give a similar picture?*

3 things that look like ROP:

- 1) Incontinentia pigmenti
- 2) ?
- 3) ?

Q

ROP: DDx



- In addition to a dragged macula, ROP pts often have a dragged **disc**. *What three other clinical entities can give a similar picture?*

3 things that look like ROP:

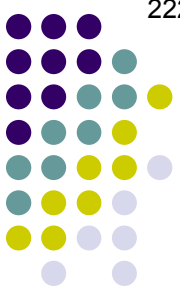
1) Incontinentia pigmenti

2) *Hint: A dz of the vitreoretinal interface*

3) ?

A

ROP: **DDx**

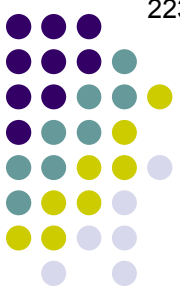


- In addition to a dragged macula, ROP pts often have a dragged **disc**. *What three other clinical entities can give a similar picture?*

3 things that look like ROP:

- 1) Incontinentia pigmenti
- 2) Familial exudative vitreoretinopathy (FEVR)
- 3) ?

Q

ROP: **DDx**

- In addition to a dragged macula, ROP pts often have a dragged **disc**. *What three other clinical entities can give a similar picture?*

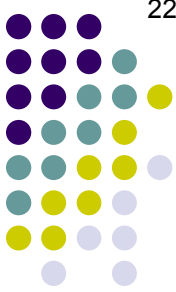
3 things that look like ROP:

- 1) Incontinentia pigmenti
- 2) Familial exudative vitreoretinopathy (FEVR)
- 3)

Hint: Can also look like Rb

A

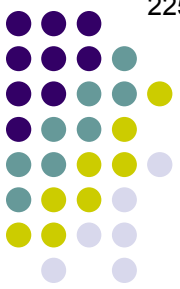
ROP: **DDx**



- In addition to a dragged macula, ROP pts often have a dragged **disc**. *What three other clinical entities can give a similar picture?*

3 things that look like ROP:

- 1) Incontinentia pigmenti
- 2) Familial exudative vitreoretinopathy (FEVR)
- 3) *Toxocara* chorioretinitis



Q

ROP: **DDx**

What is the eponymous name for IP?

and macula, ROP pts
disc. *What three other
e a similar picture?*

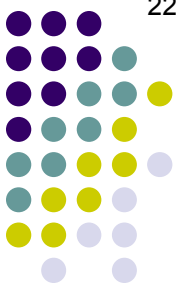
things that look like ROP:

Incontinentia pigmenti

Familial exudative vitreoretinopathy (FEVR)
Toxocara chorioretinitis

A

ROP: **DDx**



What is the eponymous name for IP?
Bloch-Sulzberger syndrome

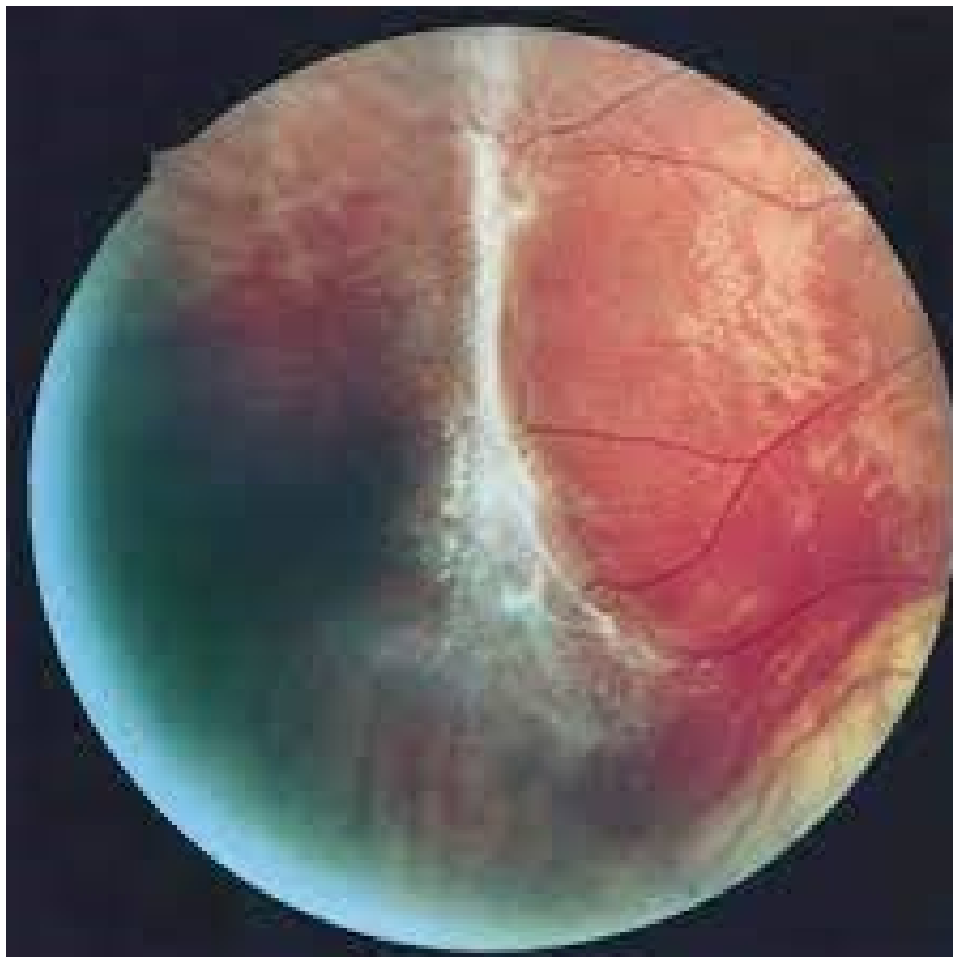
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disc. *What three other
e a similar picture?*

things that look like ROP:

Incontinentia pigmenti

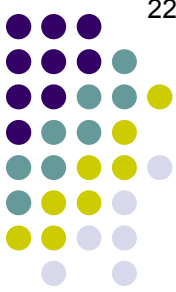
Familial exudative vitreoretinopathy (FEVR)
Toxocara chorioretinitis

ROP: **DDx**



Incontinentia pigmenti





Q

ROP: **DDx**

What is the eponymous name for IP?
Bloch-Sulzberger syndrome

What is the inheritance pattern of IP?

and macula, ROP pts
disc. *What three other
e a similar picture?*

things that look like ROP:

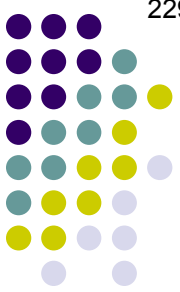
Incontinentia pigmenti

Familial exudative vitreoretinopathy (FEVR)
Toxocara chorioretinitis

A

ROP: **DDx**

229



What is the eponymous name for IP?
Bloch-Sulzberger syndrome

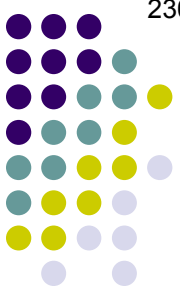
What is the inheritance pattern of IP?
X-linked dominant

and macula, ROP pts
disc. *What three other
e a similar picture?*

things that look like ROP:

Incontinentia pigmenti

Familial exudative vitreoretinopathy (FEVR)
Toxocara chorioretinitis



Q

ROP: **DDx**

What is the eponymous name for IP?
Bloch-Sulzberger syndrome

What is the inheritance pattern of IP?
X-linked dominant

What does this pattern portend for its demographics?

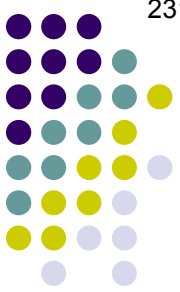
and macula, ROP pts

disc. *What three other
e a similar picture?*

things that look like ROP:

Incontinentia pigmenti

Familial exudative vitreoretinopathy (FEVR)
Toxocara chorioretinitis



What is the eponymous name for IP?
Bloch-Sulzberger syndrome

What is the inheritance pattern of IP?
X-linked dominant

What does this pattern portend for its demographics?
Males die in utero, so almost all cases will be females

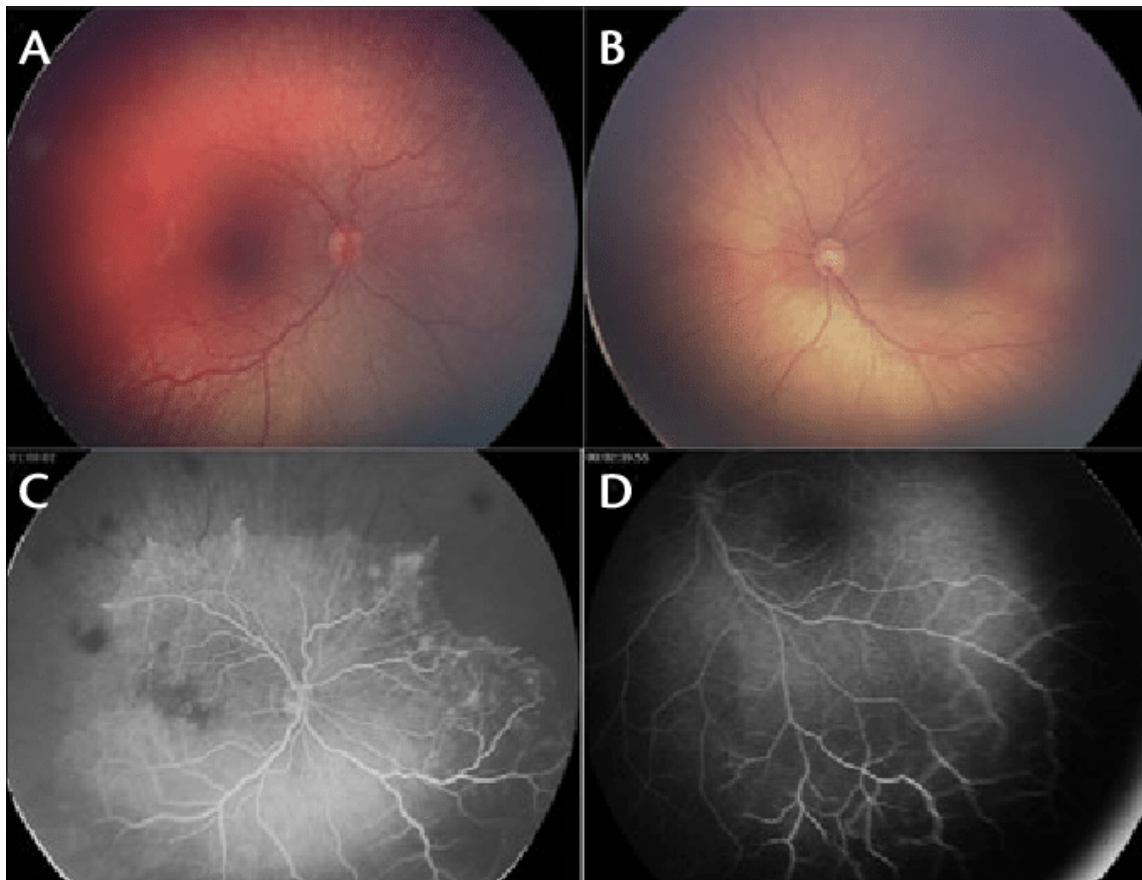
and macula, ROP pts

disc. What three other
e a similar picture?

things that look like ROP:

Incontinentia pigmenti

Familial exudative vitreoretinopathy (FEVR)
Toxocara chorioretinitis

ROP: **DDx**

A 4-month old girl with incontinentia pigmenti was admitted for seizures and intracranial hemorrhage. It may be difficult to appreciate the peripheral nonperfusion with RetCam photography alone (A-B), but the findings become clear with RetCam FA



Q

ROP: **DDx**

What is the eponymous name for IP?
Bloch-Sulzberger syndrome

What is the inheritance pattern of IP?
X-linked dominant

What does this pattern portend for its demographics?
Males die in utero, so almost all cases will be females

We noted that IP is a phakomatosis. By what more on-the-nose term are phakomatoses known?

and macula, ROP pts

disc. *What three other things have a similar picture?*

things that look like ROP:

Incontinentia pigmenti

Familial exudative vitreoretinopathy (FEVR)
Toxocara chorioretinitis



What is the eponymous name for IP?
Bloch-Sulzberger syndrome

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We noted that IP is a phakomatosis. By what more on-the-nose term are phakomatoses known?
'Neurocutaneous syndromes.'

and macula, ROP pts

disc. What three other
e a similar picture?

things that look like ROP:

Incontinentia pigmenti

Familial exudative vitreoretinopathy (FEVR)
Toxocara chorioretinitis



What is the eponymous name for IP?
Bloch-Sulzberger syndrome

What is the inheritance pattern of IP?
X-linked dominant

What does this pattern portend for its demographics?
Males die in utero, so almost all cases will be females

We noted that IP is a phakomatosis. By what more on-the-nose term are phakomatoses known?
'Neurocutaneous syndromes.' Most present with multiple lesions in two or more organ systems, usually including the CNS and skin (hence the name).

and macula, ROP pts
disc. What three other
e a similar picture?

things that look like ROP:

Incontinentia pigmenti

Familial exudative vitreoretinopathy (FEVR)
Toxocara chorioretinitis



ROP: **DDx**



What is the eponymous name for IP?
Bloch-Sulzberger syndrome

What is the inheritance pattern of IP?
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Males die in utero, so almost all cases will be females

We noted that IP is a phakomatosis. By what more on-the-nose term are phakomatoses known?
'Neurocutaneous syndromes.' Most present with multiple lesions in two or more organ systems, usually including the CNS and skin (hence the name).

To what does the buzzterm splashed paint refer?

and macula, ROP pts

disc. *What three other things have a similar picture?*

things that look like ROP:

Incontinentia pigmenti

Familial exudative vitreoretinopathy (FEVR)
Toxocara chorioretinitis

Q/A

ROP: **DDx**



What is the eponymous name for IP?

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X-linked dominant

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Males die in utero, so almost all cases will be females

We noted that IP is a phakomatosis. By what more on-the-nose term are phakomatoses known?

'Neurocutaneous syndromes.' Most present with multiple lesions in two or more organ systems, usually including the CNS and skin (hence the name).

To what does the buzzterm splashed paint refer?

The appearance of the infant's skin after erythema and bullae develop at age ~

and macula, ROP pts

disc. *What three other things have a similar picture?*

things that look like ROP:

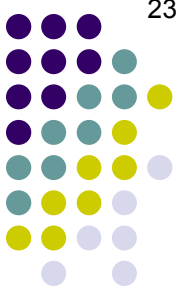
Incontinentia pigmenti

Familial exudative vitreoretinopathy (FEVR)

Toxocara chorioretinitis

A

ROP: **DDx**



What is the eponymous name for IP?

Bloch-Sulzberger syndrome

What is the inheritance pattern of IP?

X-linked dominant

What does this pattern portend for its demographics?

Males die in utero, so almost all cases will be females

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'Neurocutaneous syndromes.' Most present with multiple lesions in two or more organ systems, usually including the CNS and skin (hence the name).

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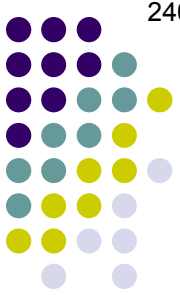


ROP: **DDx**



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Incontinentia pigmenti: Splashed-paint appearance



Q

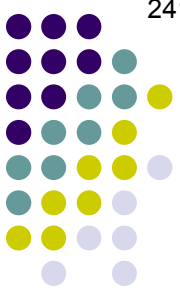
ROP: **DDx**

In a nutshell, what sort of condition is FEVR?

- Pa... ed fovea
an... er clinical
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2) **Familial exudative vitreoretinopathy (FEVR)**

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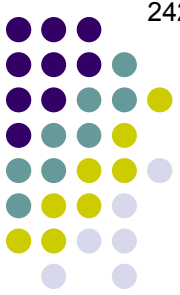
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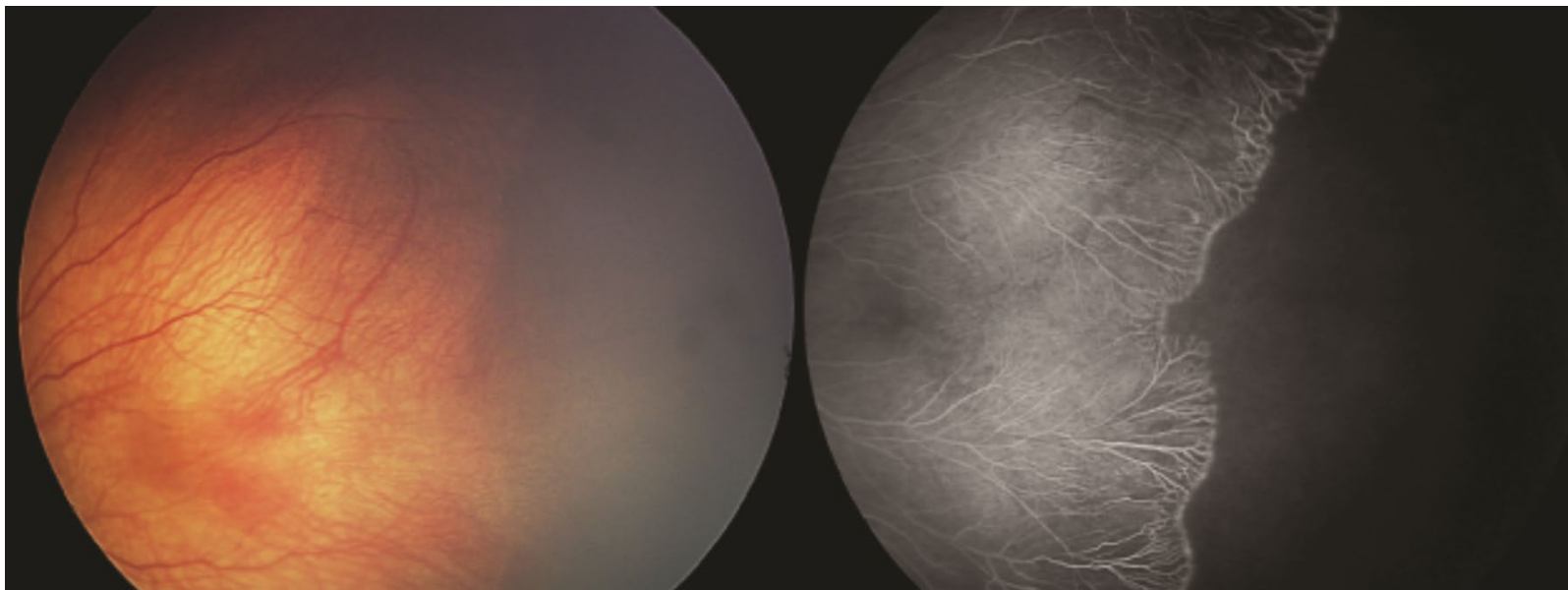
What is the basic retinal problem in FEVR?

The temporal retina fails to vascularize

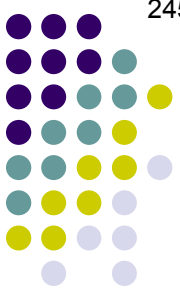
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ROP: **DDx**



FEVR: Fundus photo and FA



Q

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'The temporal retina fails to vascularize'—that sounds like ROP. In what two ways will FEVR neonates differ from ROP neonates?

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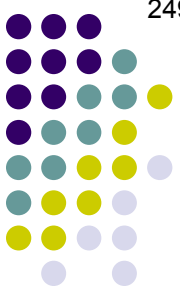
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AD, AR and X-linked forms all exist

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- Patients s/p ROP often have a dragged fovea and/or dragged disc. *What three other clinical entities can give a similar picture?*

What sort of bug is Toxocara?

Things that look like ROP:

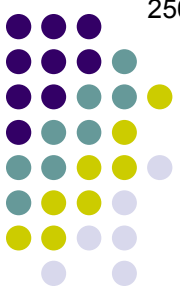
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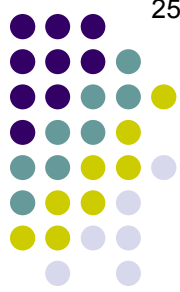
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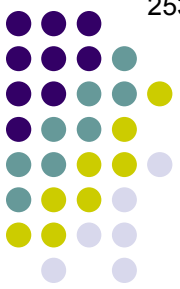
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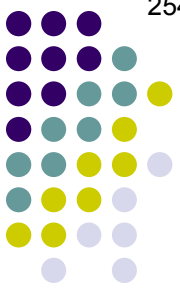
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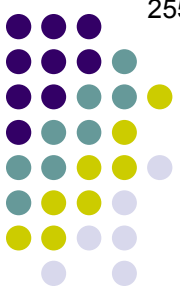
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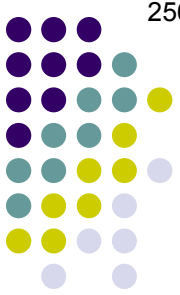
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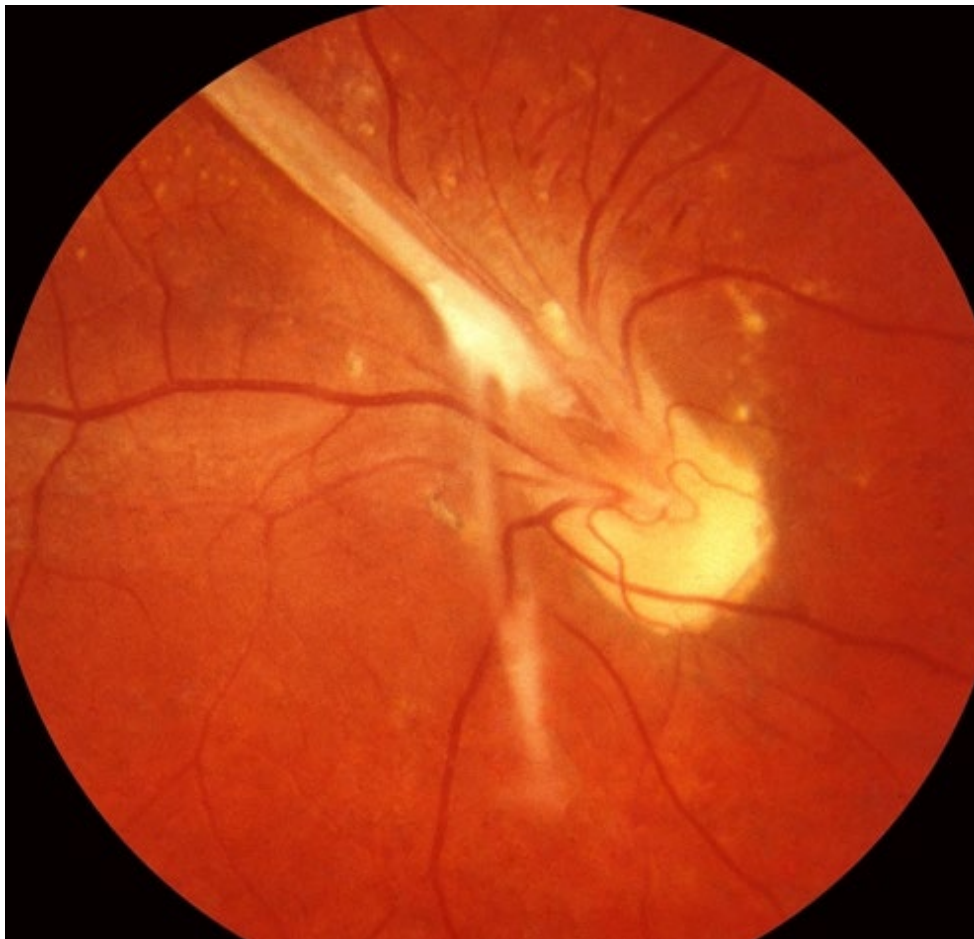
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What percent of ocular toxocariasis pts have the ROP-like presentation?

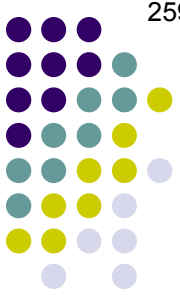
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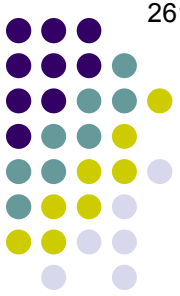
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Mechanism(s) for disc/foveal dragging in each?

3 things that look like ROP:

These two share a common mechanism

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This one has a different mechanism

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Disc/foveal dragging due
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