Q

- Concerning Rb, which of the following are true?

(Retinoblastoma)

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  - The incidence is roughly 1/100,000 live births

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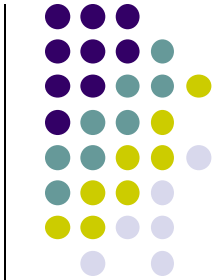

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How many new cases are there every year in North America?

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How many new cases are there every year in North America?
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--Whether there is a family hx of Rb
--Laterality; ie, whether the child has unilateral disease vs bilateral disease

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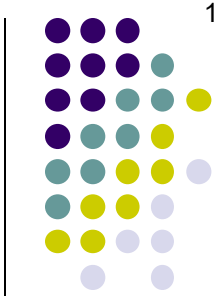

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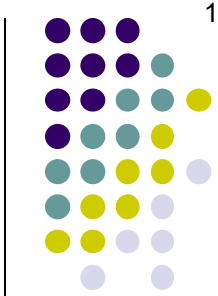

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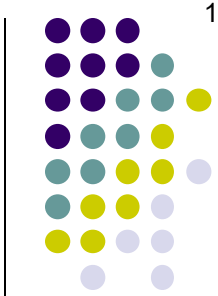
Is the rate in developing nations higher or lower?



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Which two areas of the world have the highest Rb rates?
Africa and **India**

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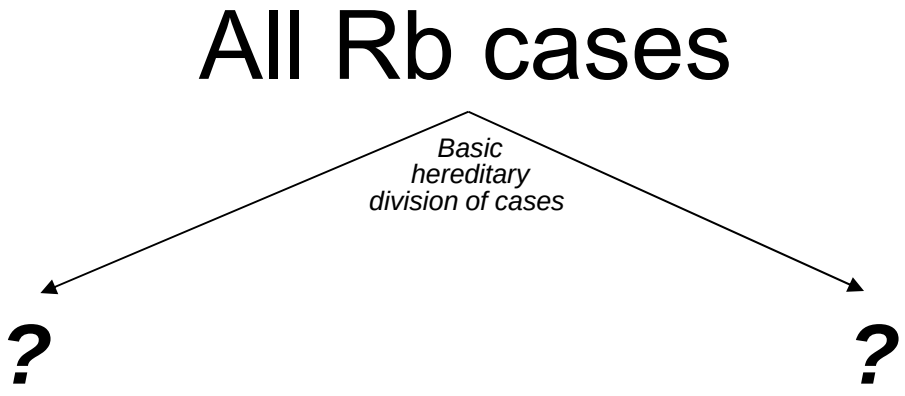
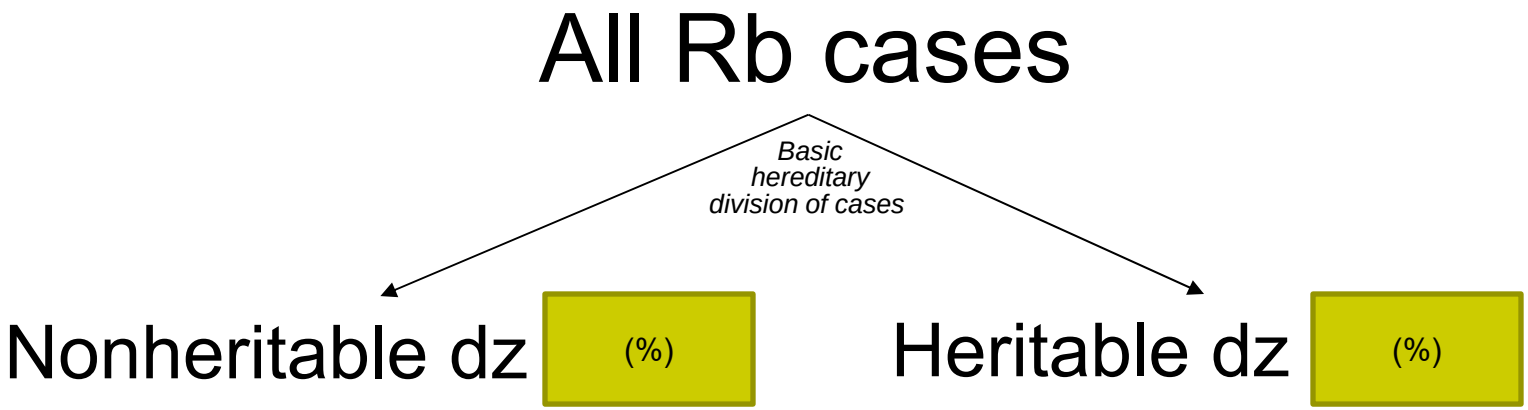


Genetic basics of Rb: tl;dr



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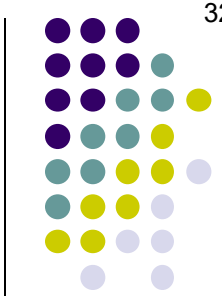
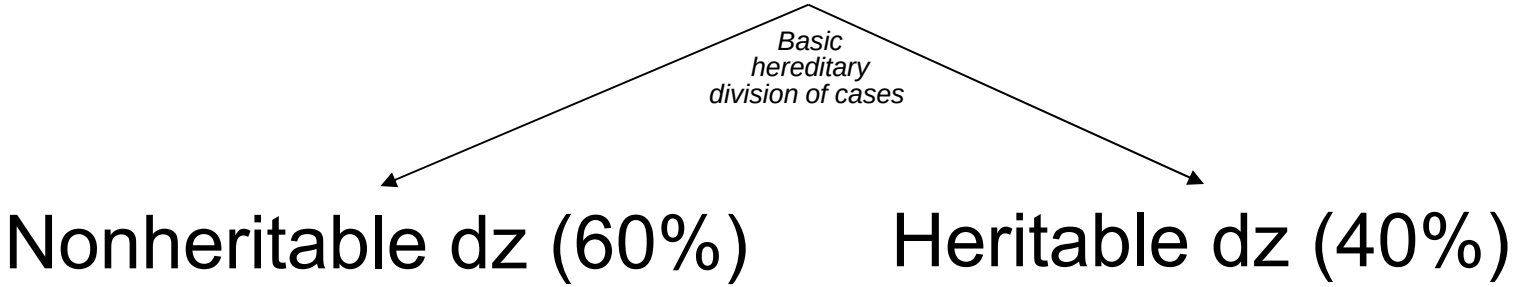


All Rb cases



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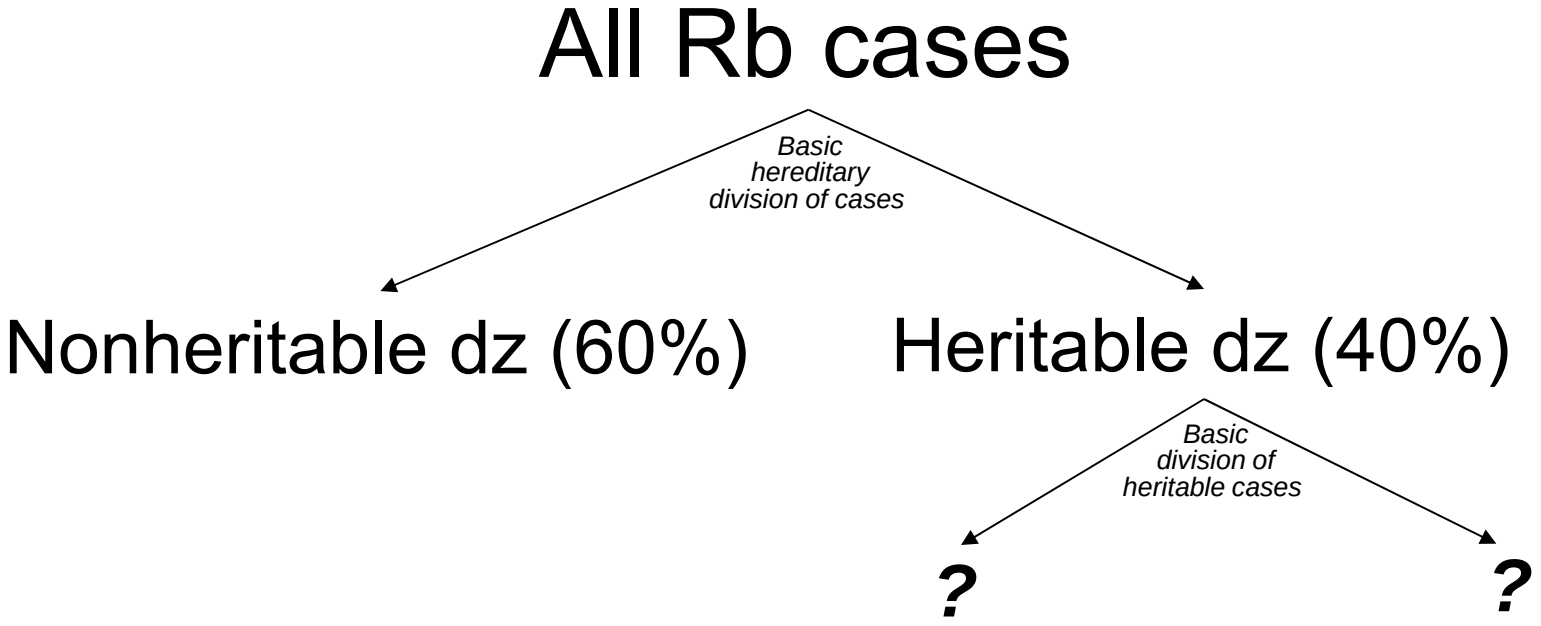

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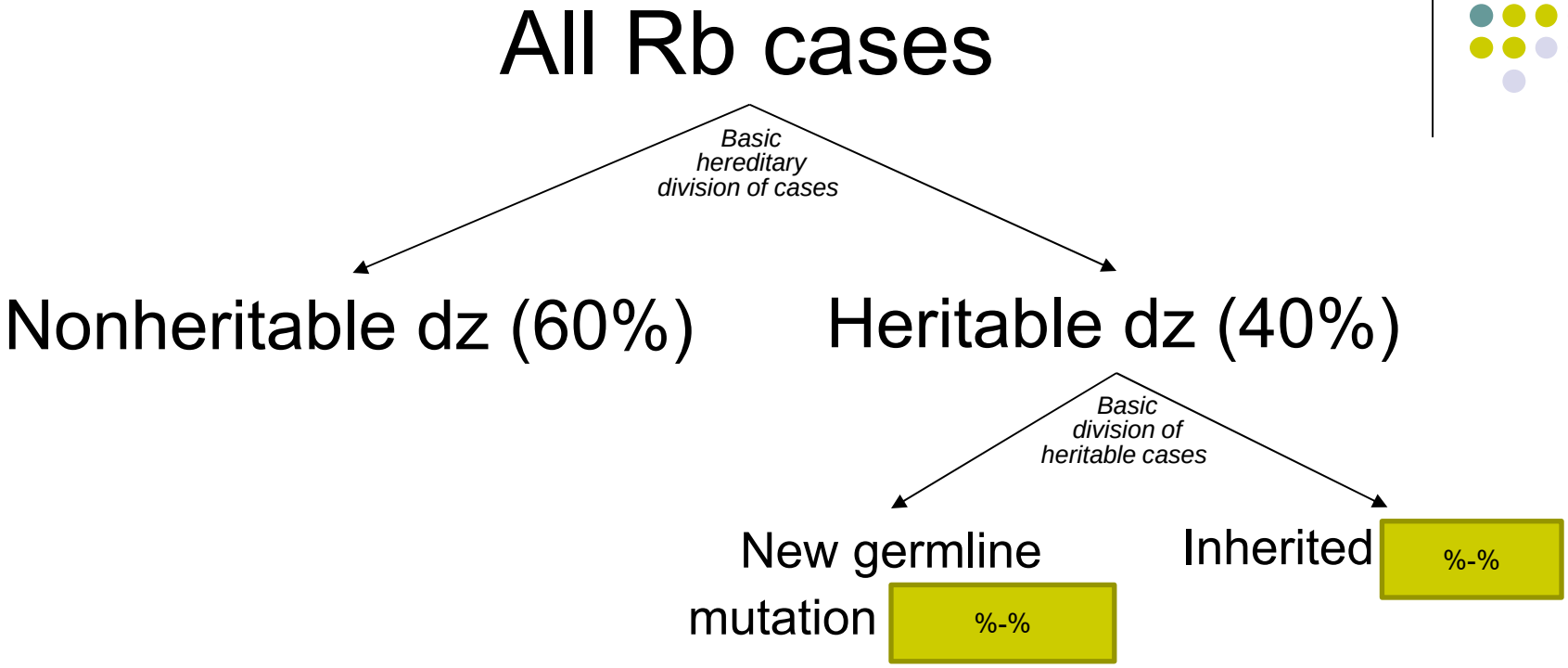

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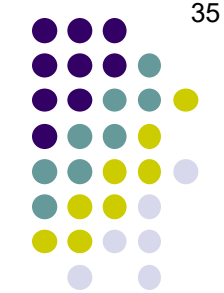

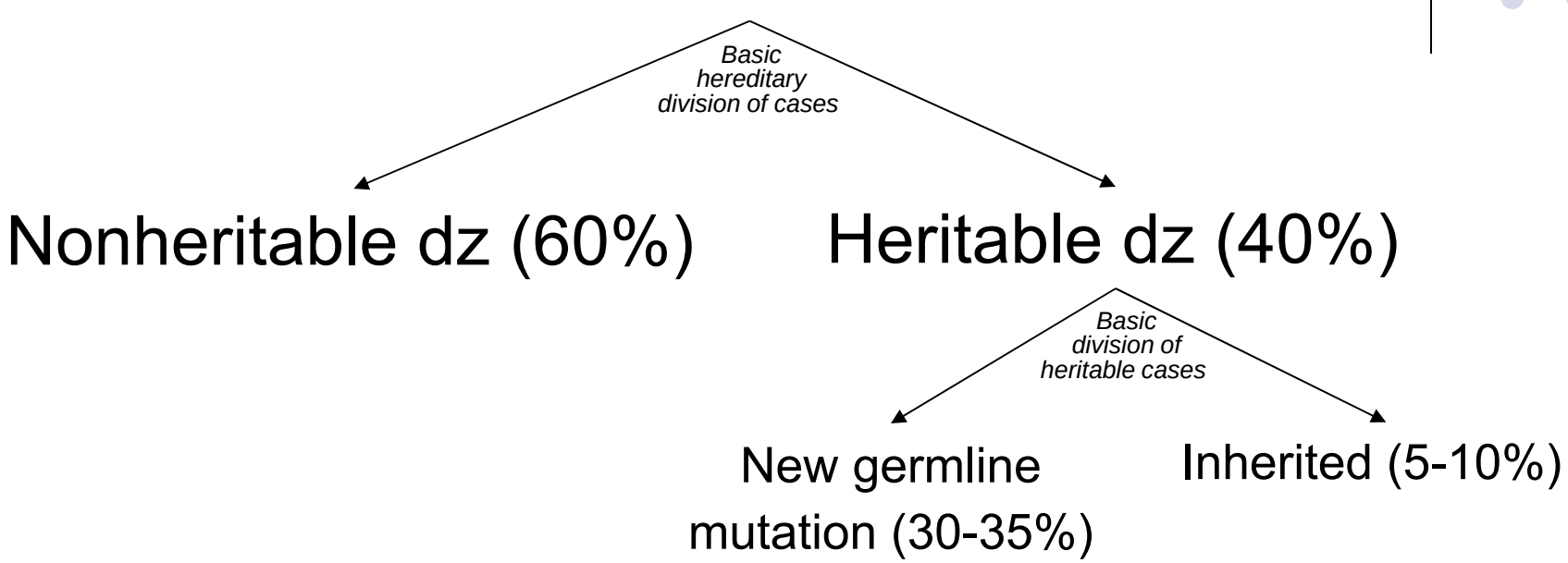

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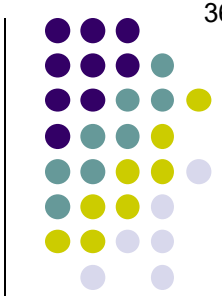
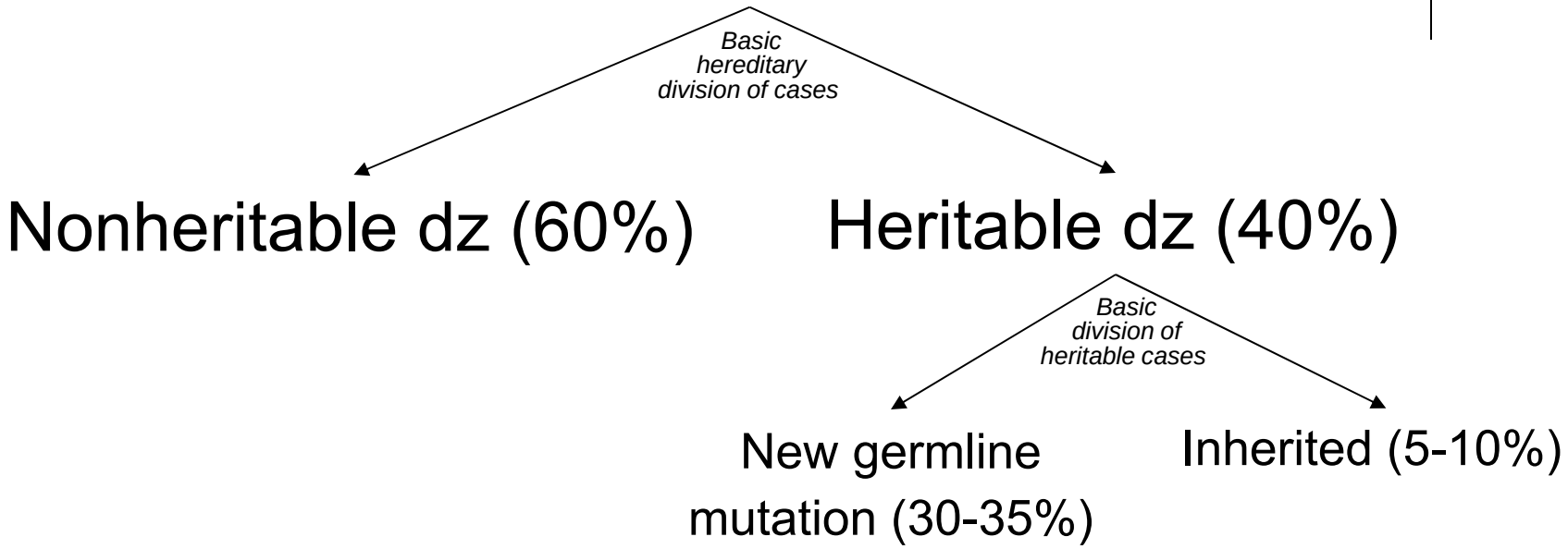
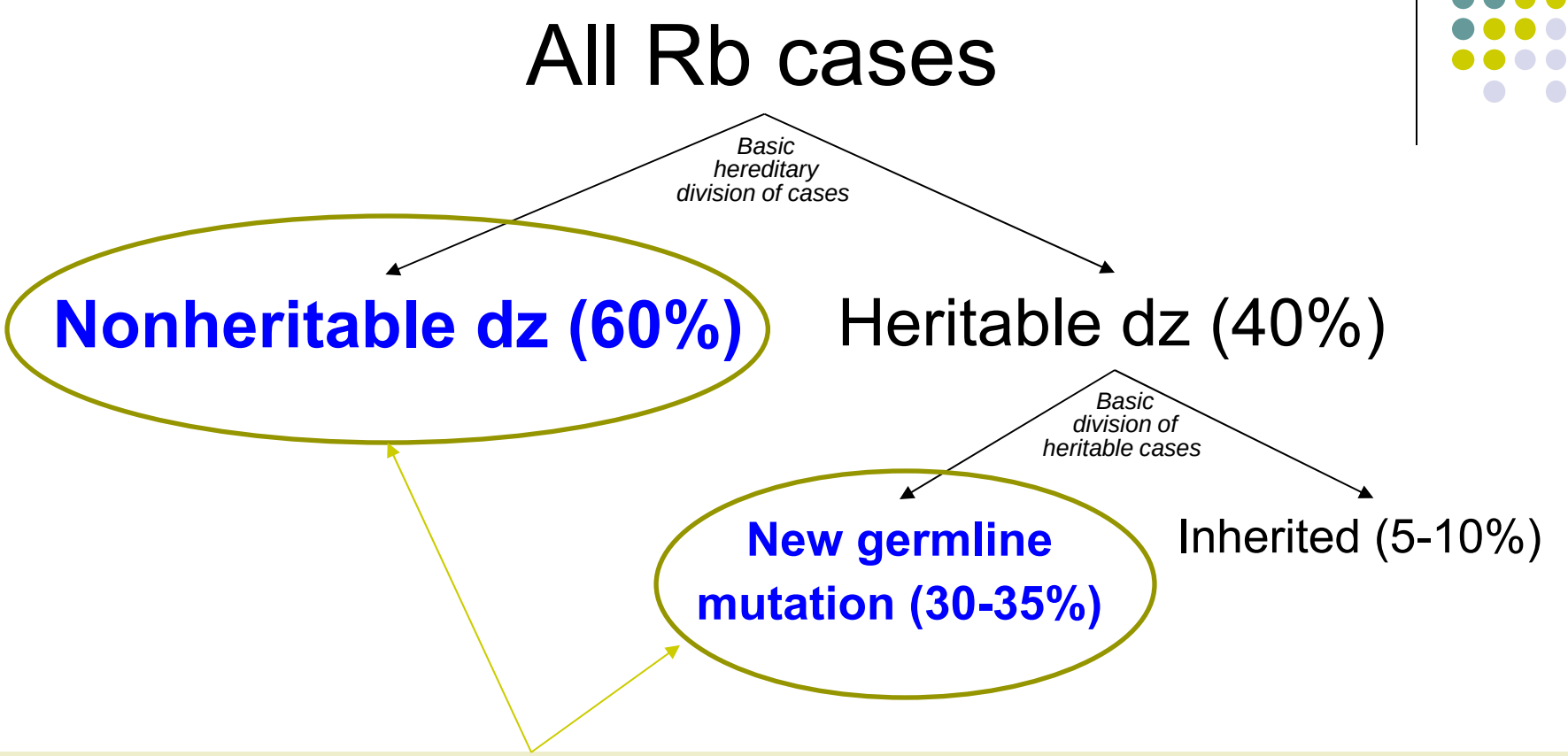




Which form(s) is/are sporadic?

Genetic basics of Rb: tl;dr



Which form(s) is/are sporadic? **Both of these.**
Because sporadic cases occur in the absence of a family history, it is often assumed (incorrectly) that all sporadic cases are nonheritable. To the contrary, fully 30-35% of Rb cases are both sporadic *and* heritable.

Genetic basics of Rb: tl;dr

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OK, so some Rb pts have heritable disease, and others don't. Other than implications for genetic counseling regarding having children, does it really matter?

How can a disease be heritable *if it's not* inherited?
A heritable disease is one that is coded for in germline cells. A heritable disease enters the germline in one of two ways--either it is inherited (ie, present at conception), or occurs as a new, post-conception germline mutation. In Rb, the 40% of pts with heritable (ie, germline) disease can be divided into 5-10% who inherited the disease, and the 30-35% who possess a new germline mutation.

What about the 60% with nonheritable disease?
In these case, mutagenesis occurred later, in non-germline (ie, somatic) cells

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(98% of the time. We'll talk about the other 2% later)

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In addition to bilaterality, what other sort of presentation is associated with the heritable form of Rb?

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Multifocal disease; ie, multiple tumors within the same eye

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Note that this means 15% of heritable Rb pts have *unilateral* disease. Thus, unilateral disease is **not** pathognomonic for nonheritable Rb.

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What finding would strongly suggest that a child with unilateral Rb harbors a germline mutation?

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If s/he had **multifocal dz** within the affected eye

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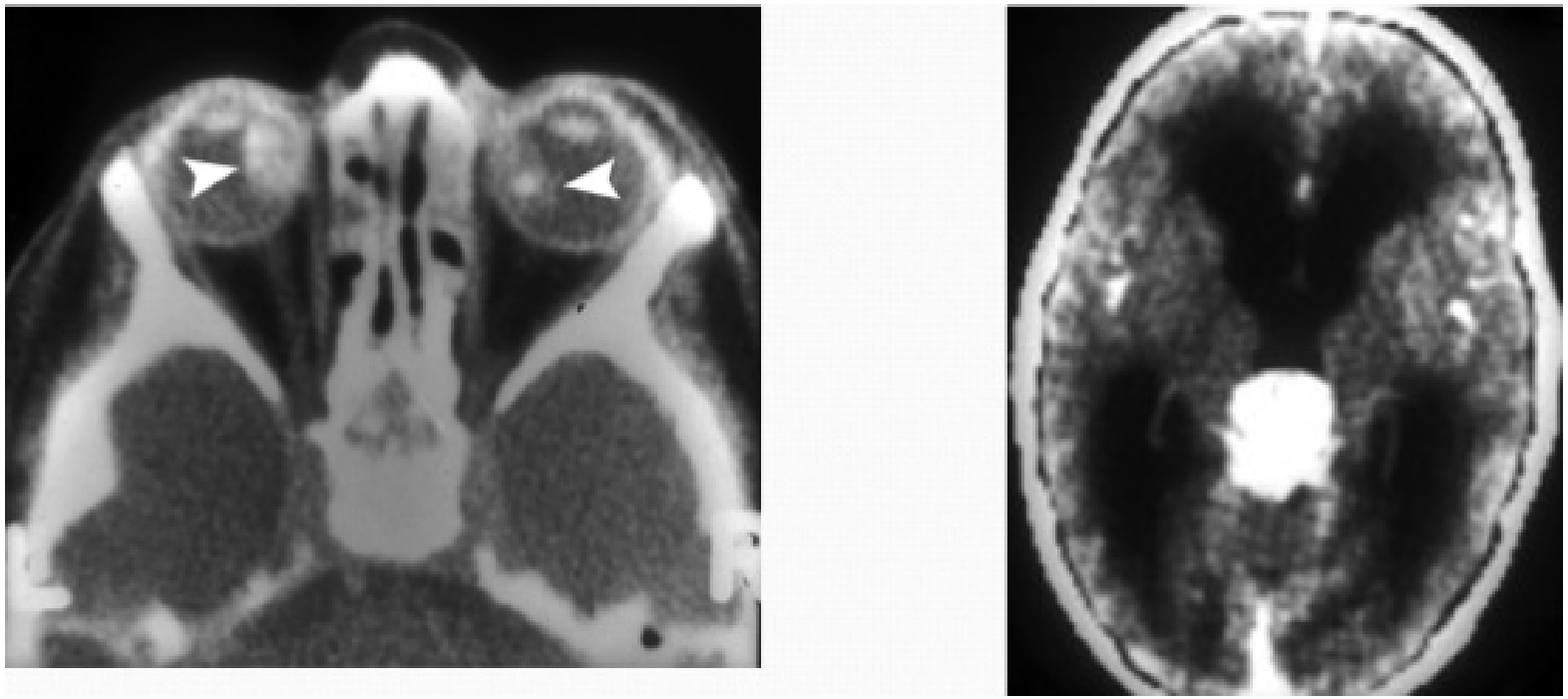

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Yes--about % of bilateral Rb is somatic/nonheritable in origin

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The inheritance pattern is consistent with autosomal dominant inheritance

So Rb is an AD disease, then?
No, it is unquestionably an autosomal recessive disease. **Both** copies of the responsible gene (*RB1*)

Given the plethora of potential mutations and the large number of retinoblasts in which they have the opportunity to occur, what percent of pts with germline loss of one RB1 *gene will lose the other one in at least one cell (and therefore will develop at least one tumor)?*
An astonishing 90-95%!

certain to occur in at least one retinoblast:

- **1) the number of different mutations that can occur is substantial, and**
- **2) the sheer number of retinoblasts provides many opportunities for such a mutation to take place (remember, all that need happen for a tumor to develop is that ONE retinoblast lose its sole functioning copy of *RB1*).**

What percent of Rb pts have a positive family hx for the disease?

This also explains the relatively high rate of nonheritable (somatic) Rb. In a nonheritable disease pt, every retinoblast starts off with two intact copies of *RB1*. **In order for such a person to develop Rb, at least one retinoblast must undergo mutations to *both* copies of RB1.** For most AR diseases, the chances of this happening are very low. However, as mentioned above, the combination of a high number of potential mutations,

Could this happen in **both** *eyes of the same child? That is, can nonheritable Rb present bilaterally?*
Yes--about **2%** of bilateral Rb is somatic/nonheritable in origin

inherited the disease, and the 30-35% who possess a new germline mutation.

What about the 60% with nonheritable disease?
In these case, mutagenesis occurred later, in non-germline (ie, somatic) cells

Pedigree analysis reveals what sort of inheritance pattern in families with inherited Rb?
The inheritance pattern is consistent with autosomal dominant inheritance

So Rb is an AD disease, then?

OK, so some Rb pts have heritable disease, and others don't. Other than implications for genetic counseling regarding having children, does it really matter?
Yes, very much. **Pts with nonheritable Rb have unilateral disease**, and once cured, have a lifetime cancer risk essentially identical to that of their non-Rb cohort. In contrast, a strong majority of pts with heritable Rb have bilateral disease. Further, they are strongly predisposed to develop a host of different primary cancers throughout life.

(98% of the time. We'll talk about the other 2% later)

Recall this statement from an earlier slide—we're now talking about "the other 2%"

However, as mentioned above, the combination of a high number of potential mutations,
Could this happen in ***both*** *eyes of the same child? That is, can nonheritable Rb present bilaterally?*
Yes--about 2% of bilateral Rb is somatic/nonheritable in origin
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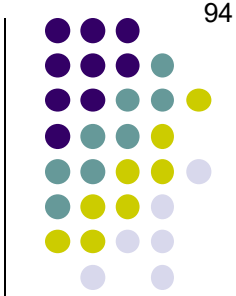

Q

- Concerning Rb, which of the following are true?
  - The incidence is roughly 1/~~100,000~~ 14K - 20K live births ***F***
  - About 60% represent nonheritable mutations ***T***
  - To diagnose a case as 'heritable,' family history ~~must~~ need not be positive ~~(but not necessarily a 1º relative)~~ ***F***
  - The *exophytic* type looks like Coats disease

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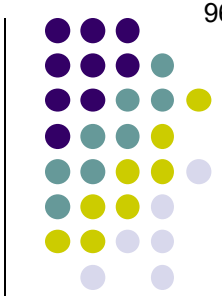

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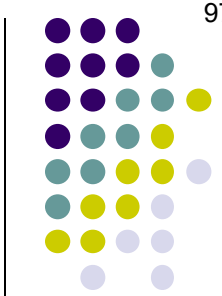
In a nutshell, what is Coats disease*?*



Q/A

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  - ~~(but not necessarily a 1º relative)~~ ositive *F*
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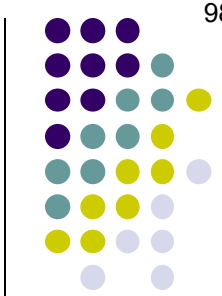
In a nutshell, what is Coats disease?
A congenital (usually) retinal vascular condition characterized by [redacted] that can be severe enough to result in [two words]



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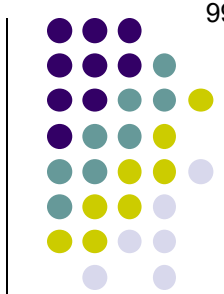

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that can be severe enough to result in
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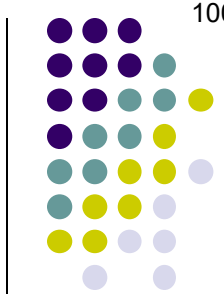

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What is the inheritance pattern in Coats?
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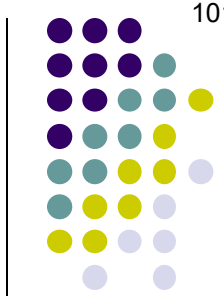

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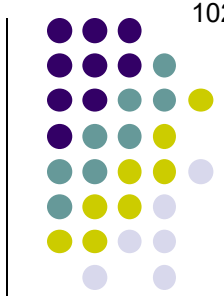

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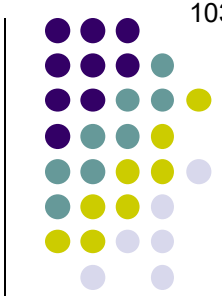

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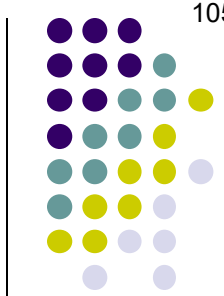

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Is there a gender predilection?
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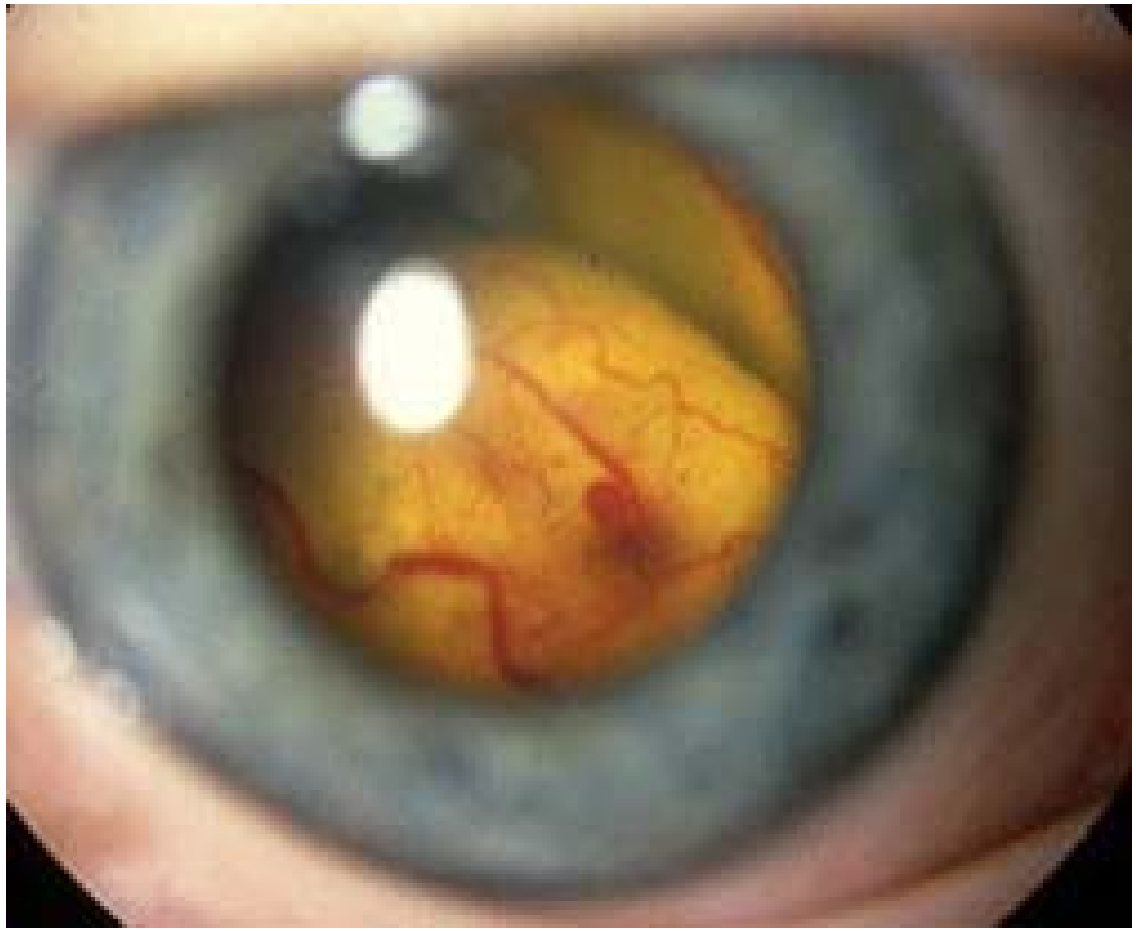
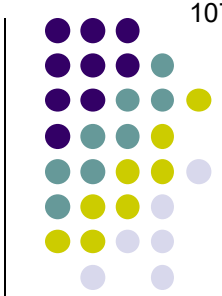


Coats dz



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In a nutshell, what is Coats disease?
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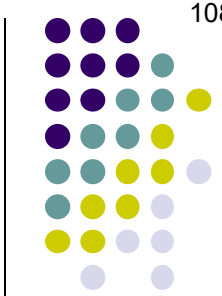
The three presentation types are…
--Exophytic
--***?***
--***?***



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The three presentation types are…
--Exophytic
--Endophytic
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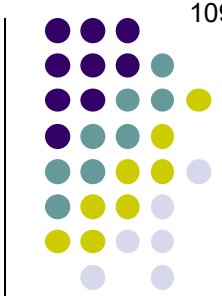

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The three presentation types are…and their respective growth patterns are…
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--Endophytic
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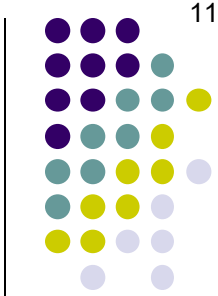

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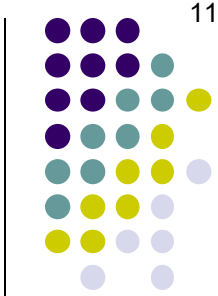
The three presentation types are…and their respective growth patterns are…
--Exophytic: Subretinal growth
--Endophytic
--Diffuse infiltrating



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What does an exophytic tumor look like on DFE?

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The three presentation types are…and their respective growth patterns are…
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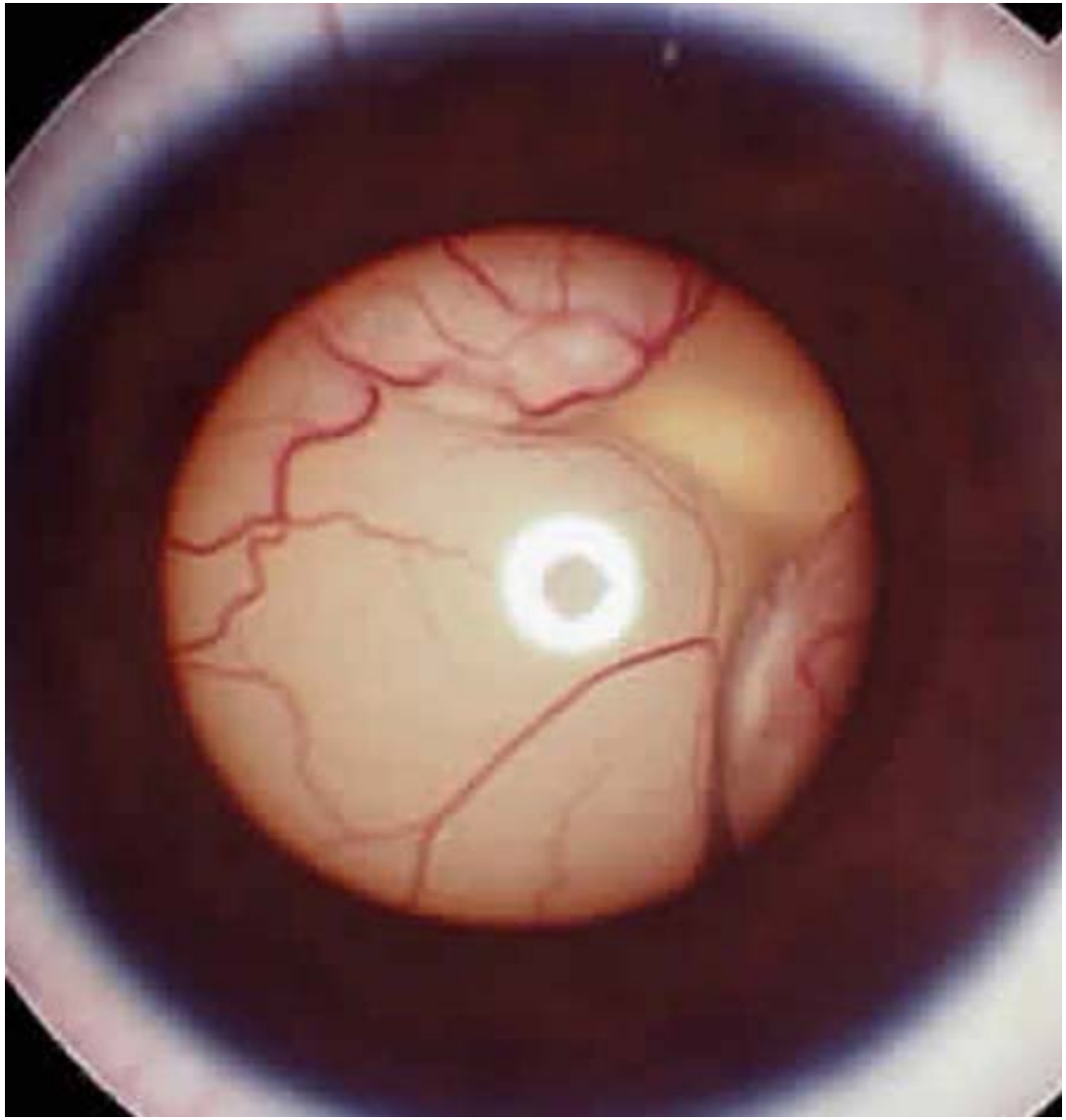
What does an exophytic tumor look like on DFE?
A white mass with retinal vessels coursing over it



Rb: Exophytic growth pattern

Q

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A white **mass** with retinal vessels coursing over it

*But Coats disease is an **exudative** process—no mass involved. Given this, how could an exophytic Rb be mistaken for Coats?*

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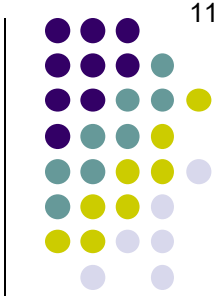
*But Coats disease is an **exudative** process—no mass involved. Given this, how could an exophytic Rb be mistaken for Coats?*
Because exophytic Rb is itself frequently associated with exuberant subretinal fluid, the effect of which can be to obscure the tumor mass



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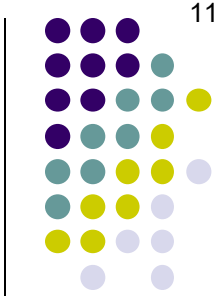
So, Coats disease is an exudative vascular condition…

(No question yet—proceed when ready)



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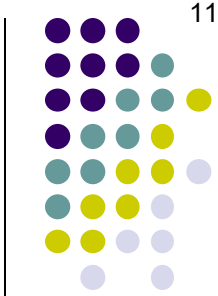
So, Coats disease is an exudative vascular condition…and Rb is a malignant neoplasm of retinal progenitor cells.

(No question yet—proceed when ready)



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So, Coats disease is an exudative vascular condition…and Rb is a malignant neoplasm of retinal progenitor cells. These two things don't sound anything alike, so why are we devoting so much space to Coats dz in a slide-set about Rb?

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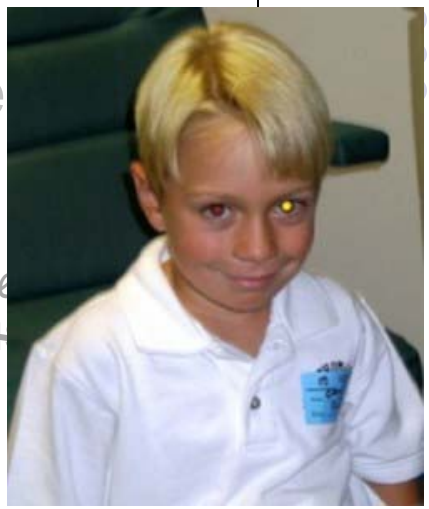
Because both are very high on the DDx for [one word] in the pediatric age group

A



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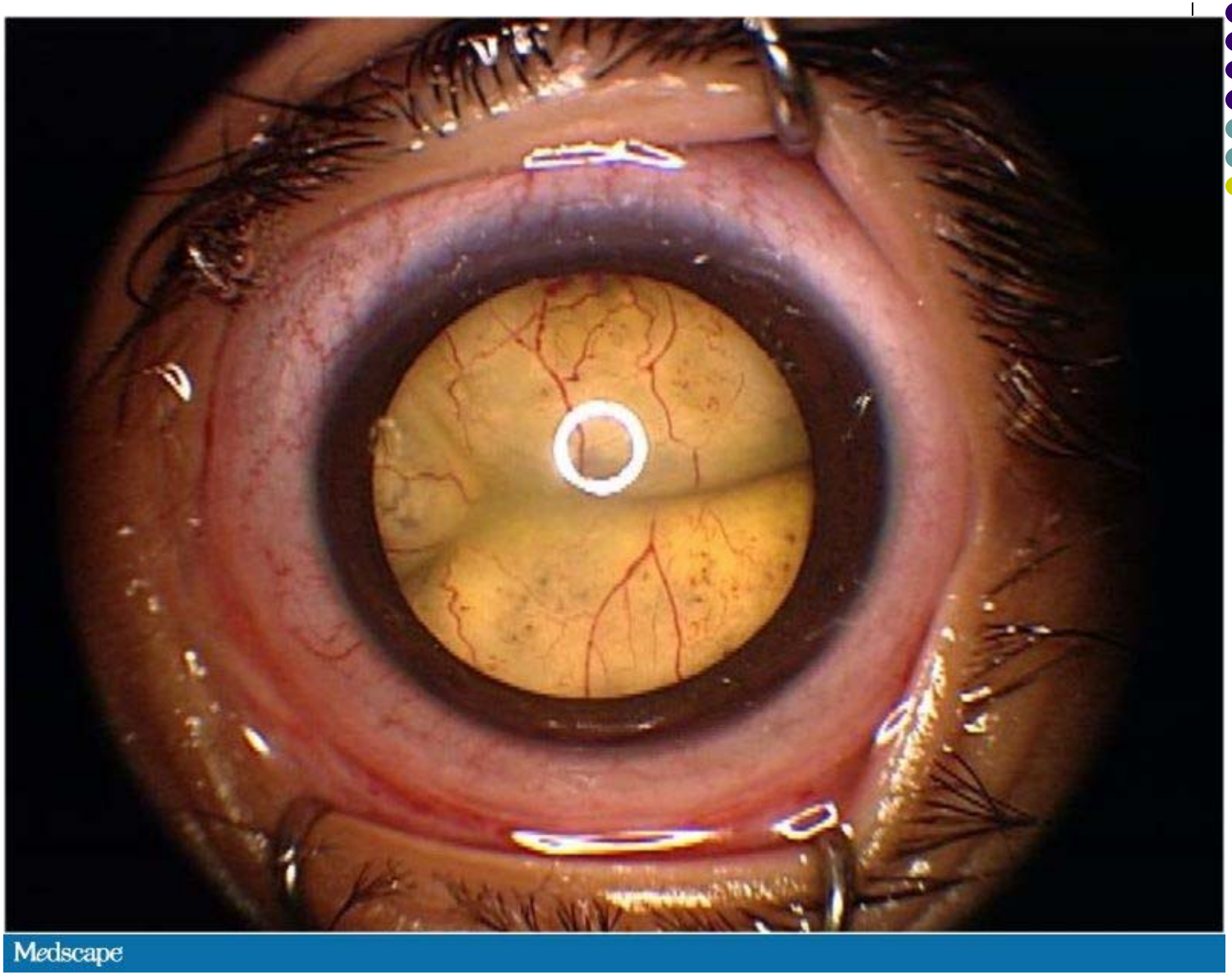
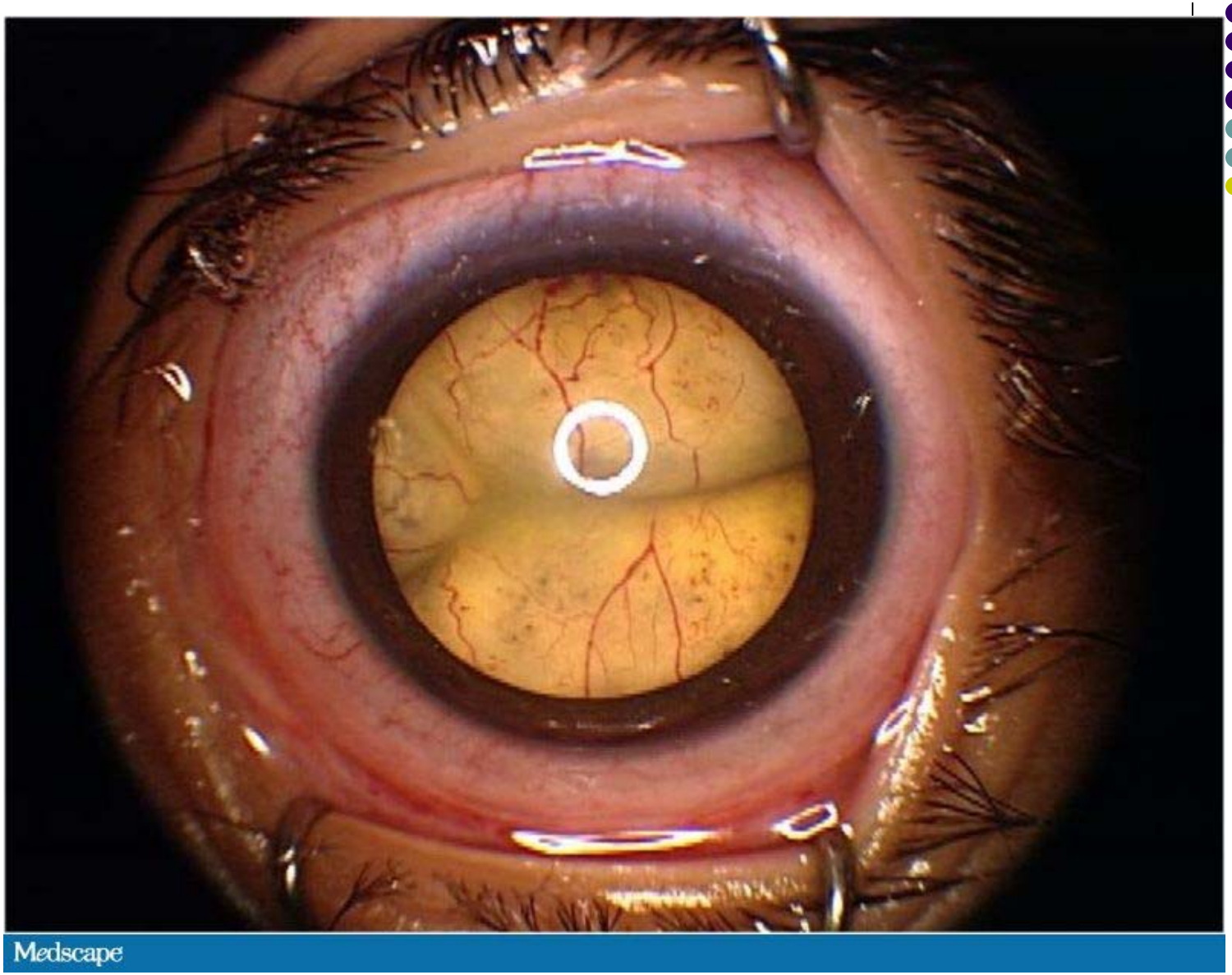
Because both are very high on the DDx for **leukocoria** in the pediatric age group

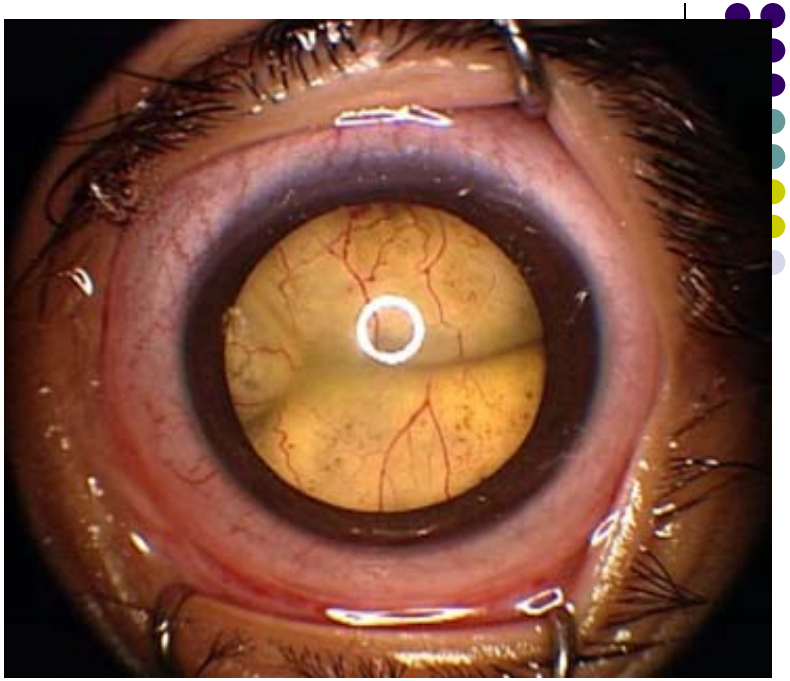


OK, quiz time. Is it Coats, or exophytic Rb?



To figure it out, look at the vasculature

In Coats, the retinal vessels are dilated, with microaneurysms and telangiectasias. (Further, the appearance often has a yellow hue.)



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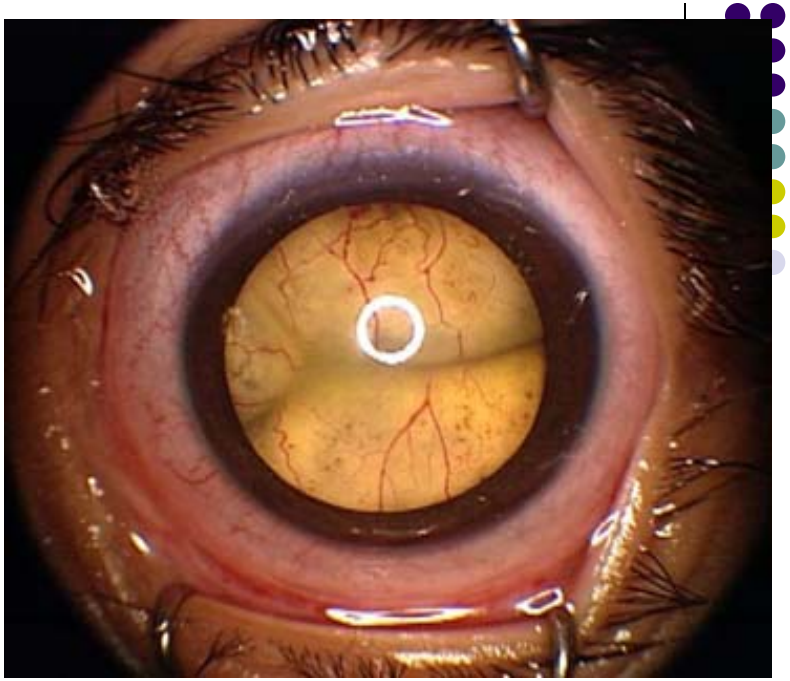
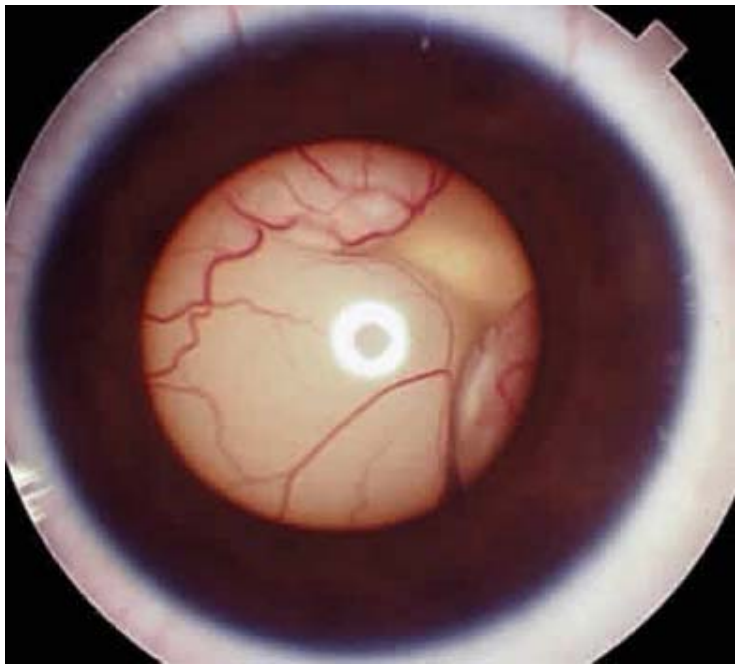
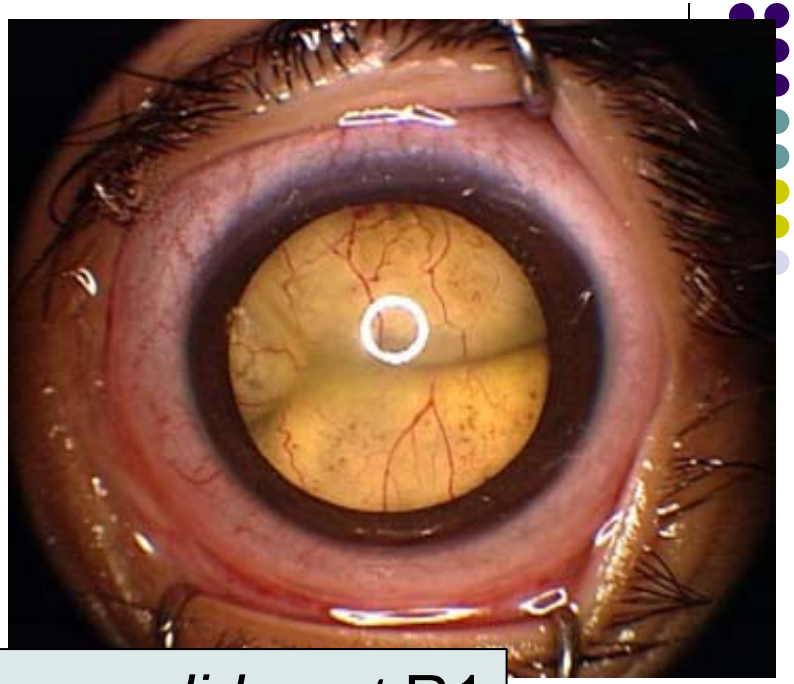


Contrast with Rb, in which the retinal vessels are *normal* in appearance. (And the hue tends to be white.)

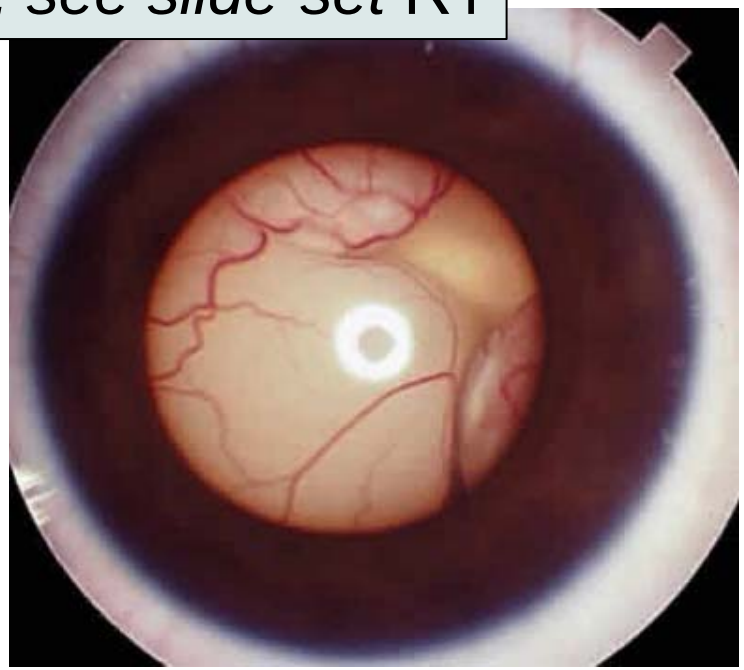


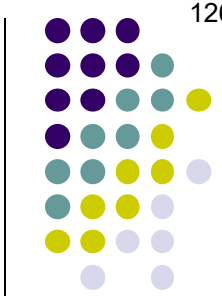
In Coats, the retinal vessels are dilated, with microaneurysms and telangiectasias. (Further, the appearance often has a yellow hue.)



For more on Coats vs Rb, see slide-set R1

Contrast with Rb, in which the retinal vessels are *normal* in appearance. (And the hue tends to be white.)





Q

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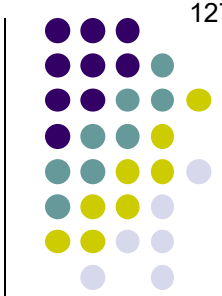
--Exophytic: Subretinal growth
--Endophytic:
--Diffuse infiltrating

Next Q: Growth pattern of endophytic Rb type



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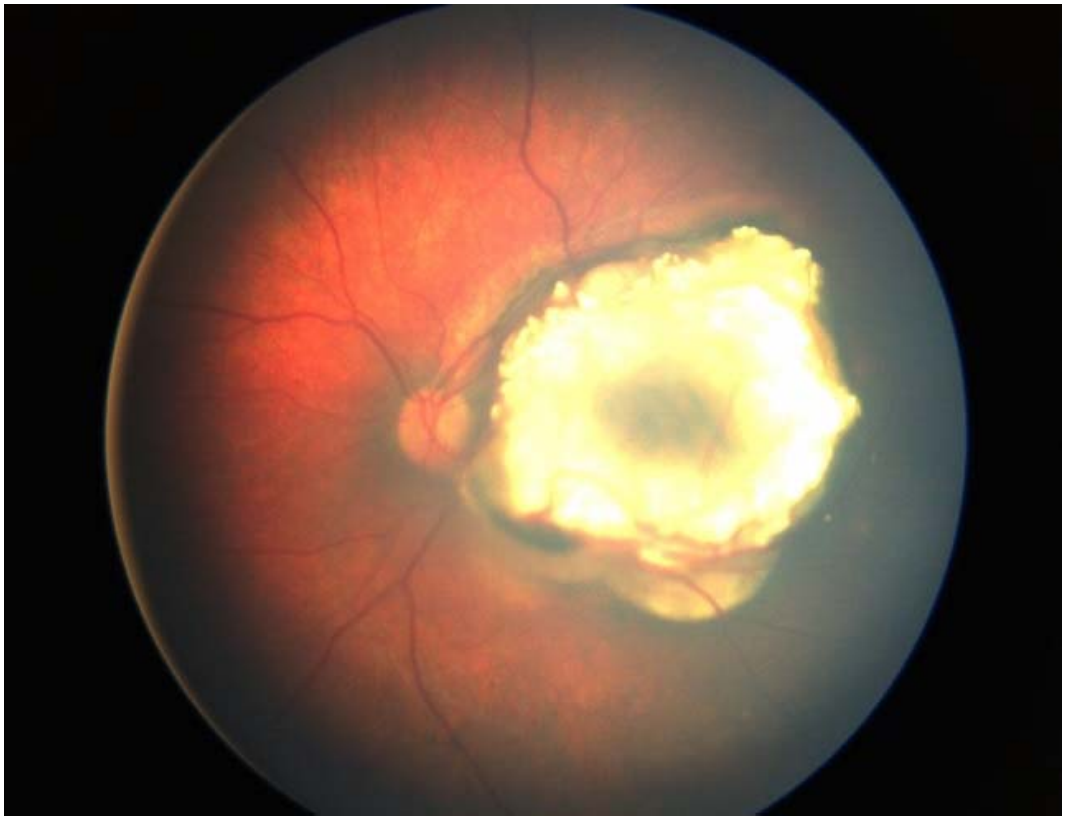
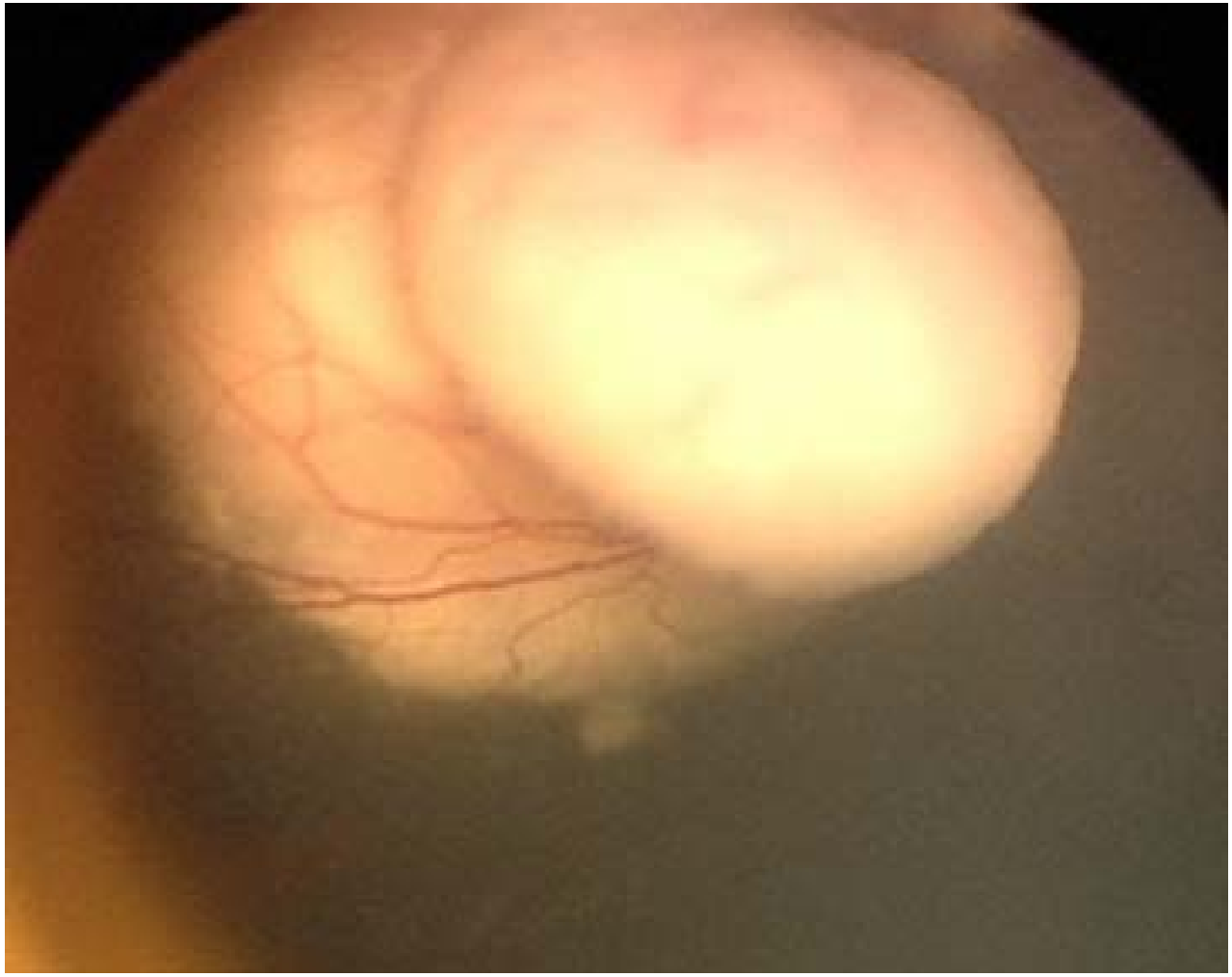
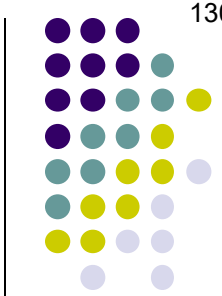


Rb: Endophytic growth pattern



Rb: Exophtic *and* endophytic growth pattern



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What does 'into the vitreous' indicate about the relationship between the tumor and the retina?
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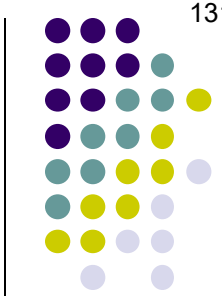

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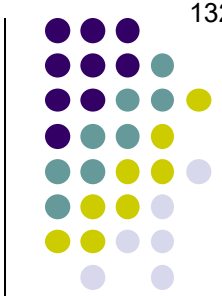
What does 'into the vitreous' indicate about the relationship between the tumor and the retina?
It indicates that the tumor has broken through the internal limiting membrane



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--It is much less v more common (% of all Rb) than the other two forms

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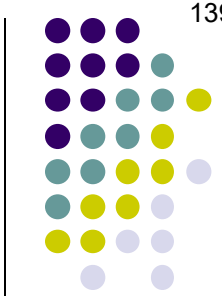

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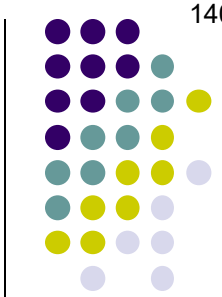

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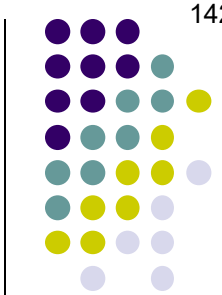

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--It grows at a much slower v faster rate than the other two forms

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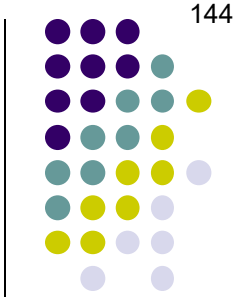

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--No distinct two words is present (hence its name)

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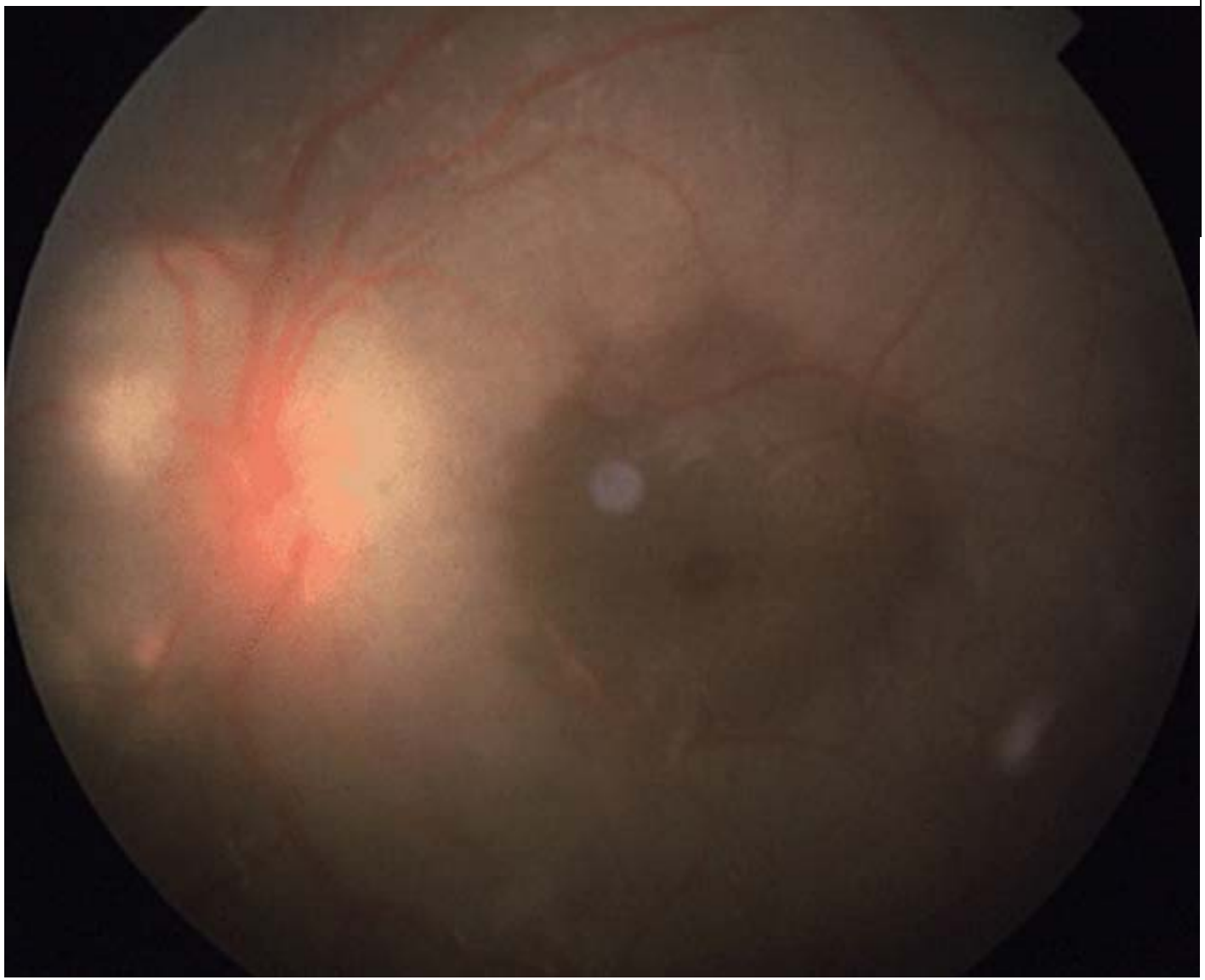
--No distinct **tumor mass** is present (hence its name)



Rb: Diffuse infiltrating growth pattern

Q

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--It grows at a much **slower** rate than the other two forms
--No distinct **tumor mass** is present (hence its name)
--Important Rb finding is usually absent

A

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--**Calcification** is usually absent

Q

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--No distinct **tumor mass** is present (hence its name)
--**Calcification** is usually absent
--It presents with an AC finding and V chamber finding

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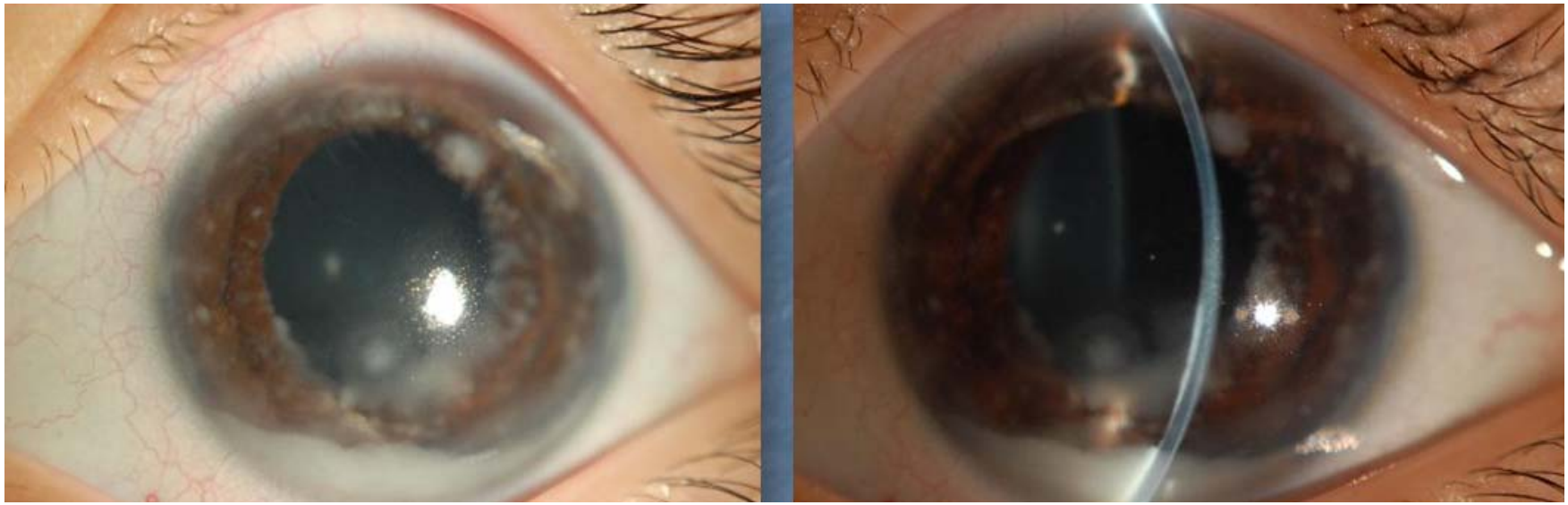
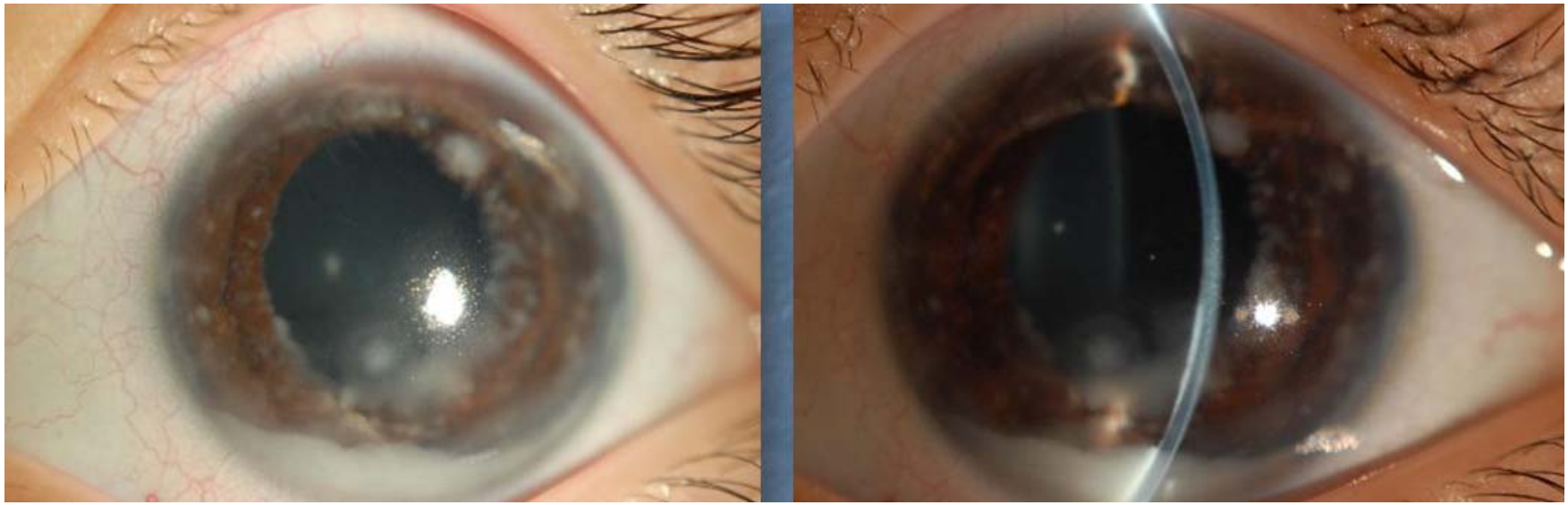
--**Calcification** is usually absent
--It presents with an **AC cell/pseudohypopyon** and **clumped vitreous cells**

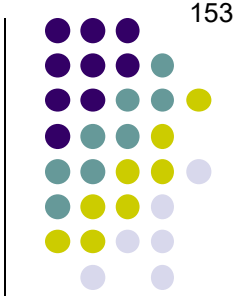


Pseudohypopyon in diffuse infiltrating Rb



Pseudohypopyon in diffuse infiltrating Rb

Diffuse infiltrating Rb *is the form that can present as a uveitis 'masquerade syndrome' with vitritis and a pseudohypopyon*



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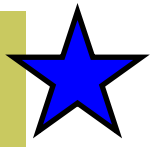
How does the pseudohypopyon of diffuse infiltrating Rb differ from a true inflammatory hypopyon?
--
--

Diffuse
--It is n
--It stri
--It is virtually always unilateral
--It is virtually
--It g
--No d
--**Calcification** is usually absent
--It presents with an **AC cell/pseudohypopyon** and **clumped vitreous cells**

Diffuse infiltrating Rb *is the form that can present as a uveitis 'masquerade syndrome' with vitritis and a pseudohypopyon*



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The three presentation types are…and their respective growth patterns are…
--Exophytic: Subretinal growth
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--Unlike a hypopyon, the pseudohypopyon will shift easily with changes in head position
--The pseudohypopyon is **snow-white**, as opposed to the yellowish tinge of a true hypopyon

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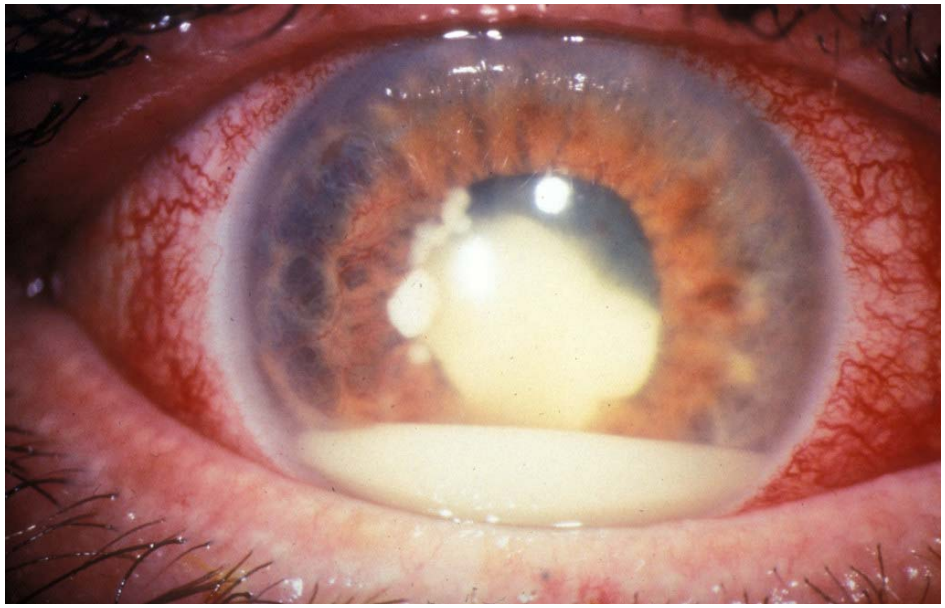
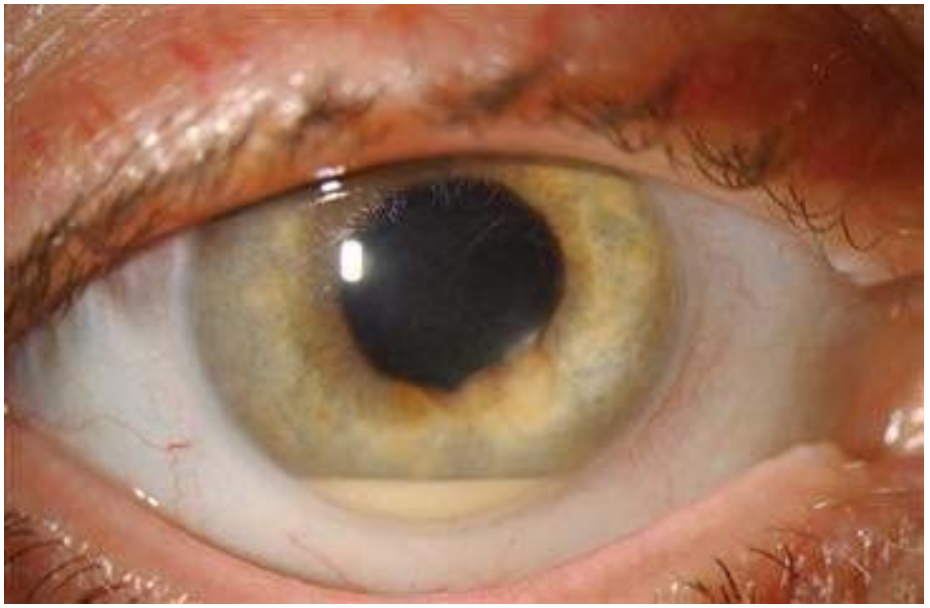
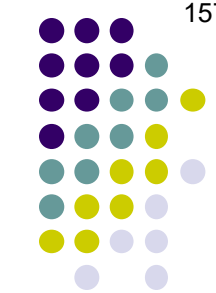


Pseudohypopyon in diffuse infiltrating Rb



Hypopyon in uveitis



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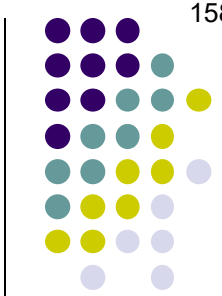

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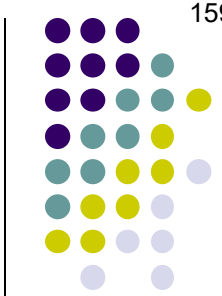

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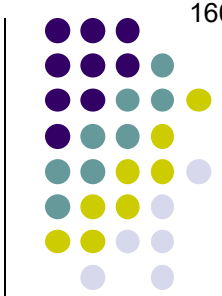

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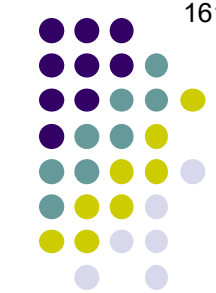
Because it incurs a significant risk of disseminating tumor cells



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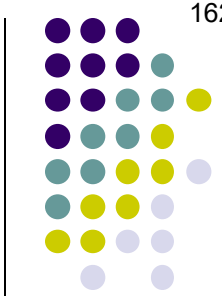

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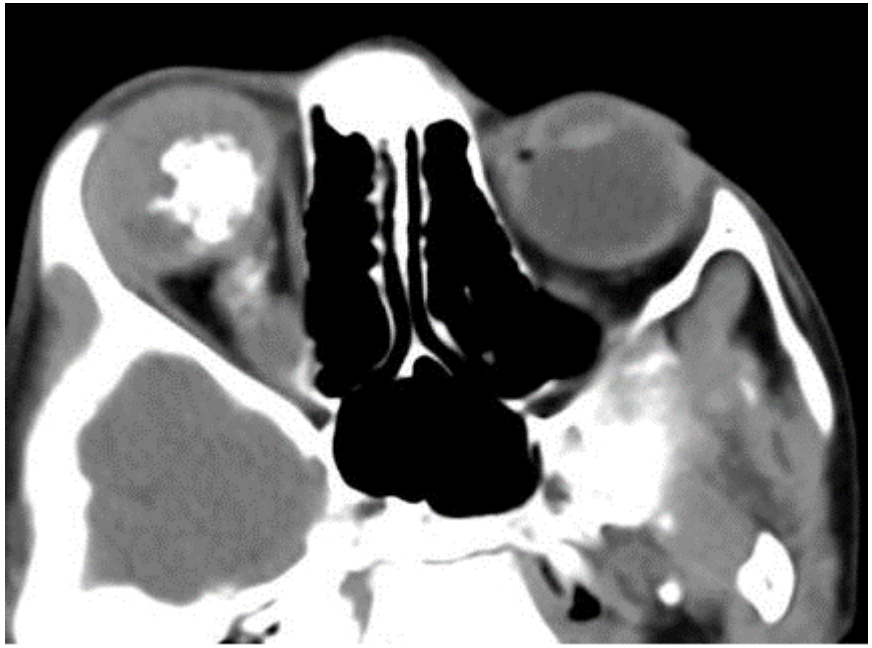
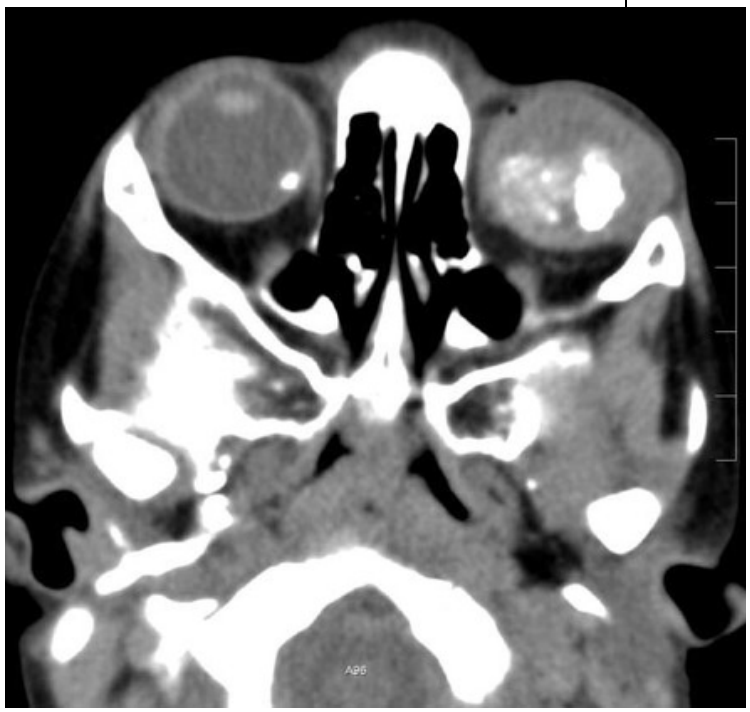
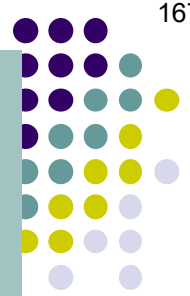




Rb: Calcifications on CT



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CT is better able to detect **intralesional calcifications**

What commonly-available alternative imaging technique is also effective for demonstrating intralesional calcifications?

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What commonly-available alternative imaging technique is also effective for demonstrating intralesional calcifications?
B-scan ultrasonography

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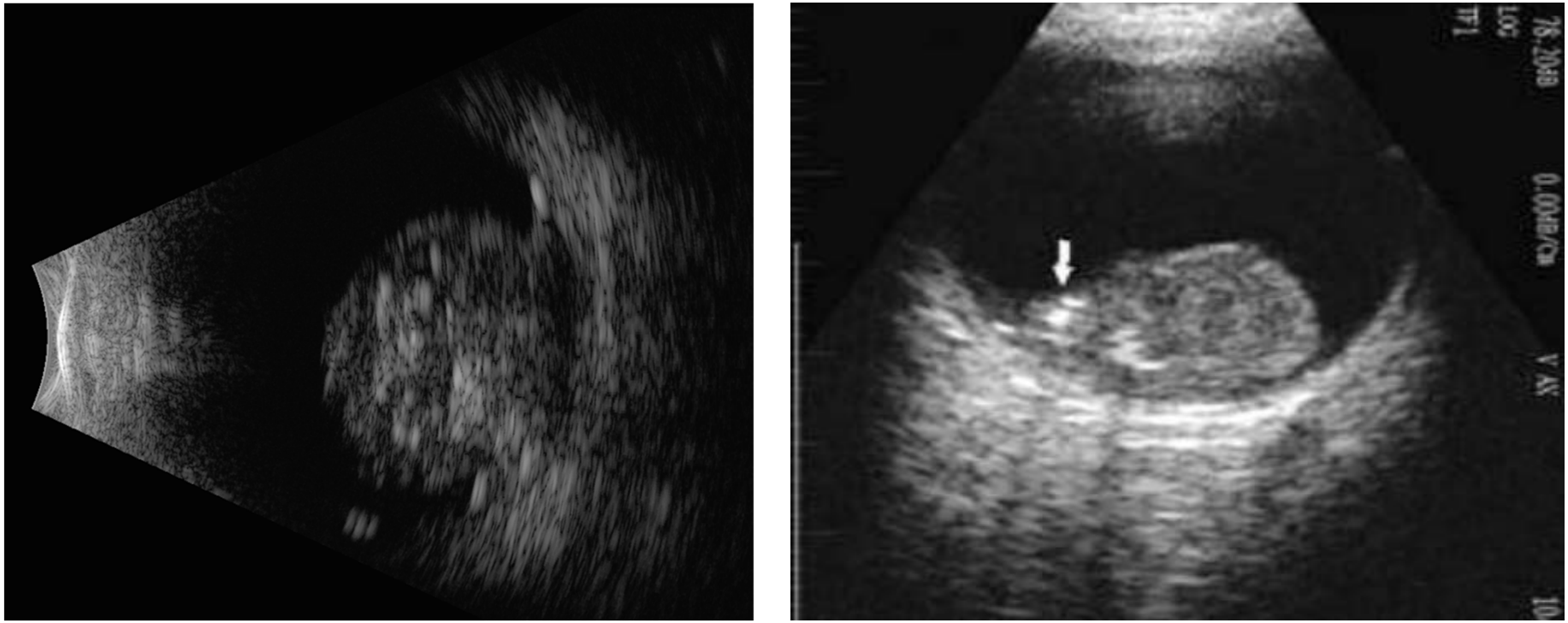
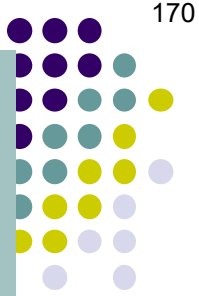


B-scan ultrasound of retinoblastoma. Note the intralesional calcifications



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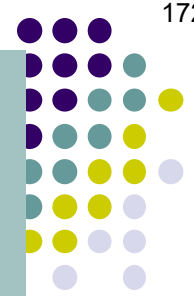

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For this reason, MRI is now preferred over CT in the workup of Rb!

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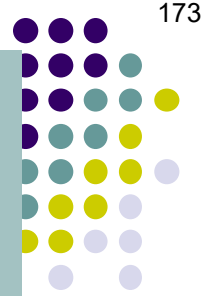

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What three specific findings are you looking for on imaging?
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What three specific findings are you looking for on imaging?
--Extraocular extension
--Optic nerve invasion
--A pinealoma (ie, 'trilateral disease')

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What needs to be undertaken if imaging reveals extraocular extension and/or optic nerve invasion?

- C
  - *What three specific findings*
  - **--Extraocular extension**
    --Optic nerve invasion
  - --A pinealoma (ie, 'trilateral
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What needs to be undertaken if imaging reveals extraocular extension and/or optic nerve invasion?
A metastatic workup

What does this entail?
--Bone scan
--Lumbar puncture to check for tumor cells in the CSF
--Bone marrow biopsy to check for tumor cells

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What does this entail?
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What is the most common mechanism by which Rb escapes the eye?

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What does this entail?
--Bone scan

What three specific findings
--Extraocular extension
Optic nerve invasion

What is the most common mechanism by which Rb escapes the eye?
Direct extension via the optic nerve, which allows access to the subarachnoid space

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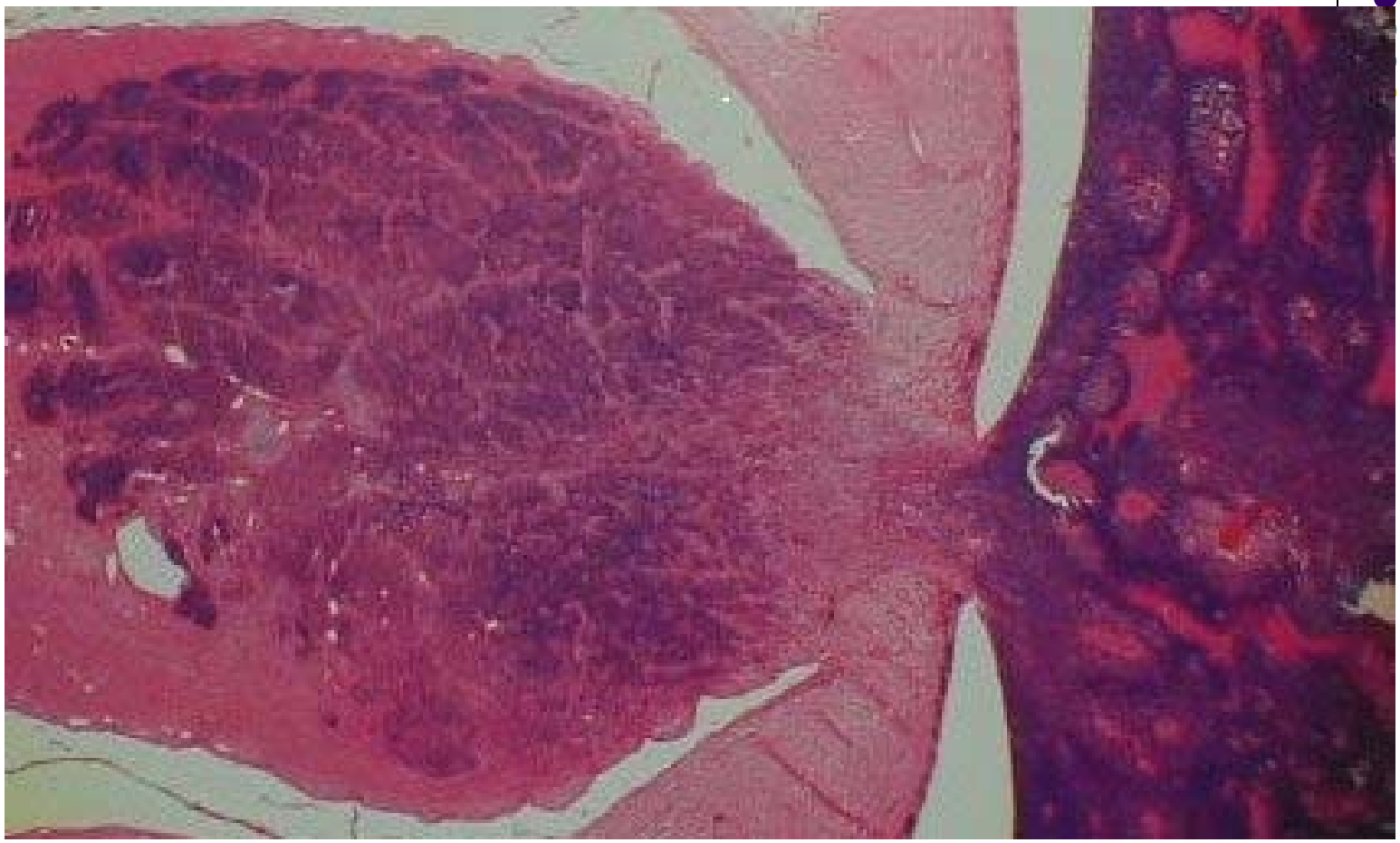
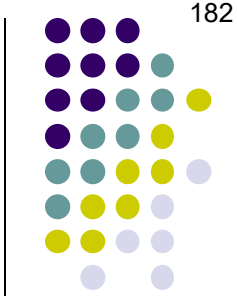


Rb: Optic nerve extension



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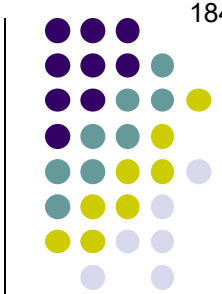

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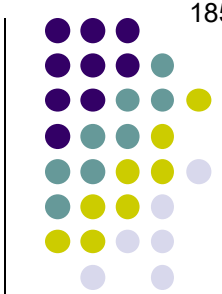
Why isn't periodic MRI surveillance for midline intracranial tumors warranted?



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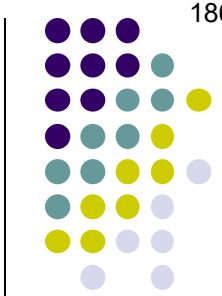
Why isn't periodic MRI surveillance for midline intracranial tumors warranted?
Because early detection has not been shown to prolong survival



Q

- Concerning Rb, which of the following are true?
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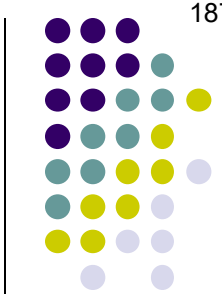
What is the average life expectancy after diagnosis of such a tumor?



A

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Why isn't periodic MRI surveillance for midline intracranial tumors warranted?
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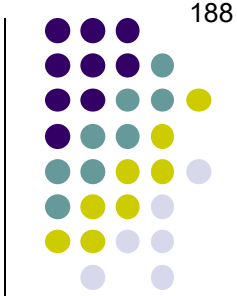
What is the average life expectancy after diagnosis of such a tumor?
About 9 months



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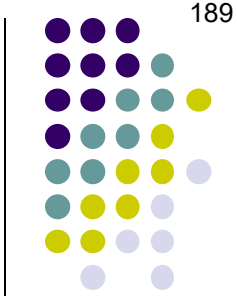

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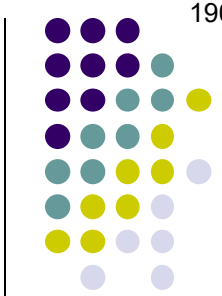

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A/Q

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14K - 20K

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The Reese-Ellsworth system was based on the assumption that the primary treatment modality was four words, and their abb. Now that the abb. is no longer the first-line treatment for most cases of Rb, the Reese-Ellsworth system is not as useful.

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no longer the

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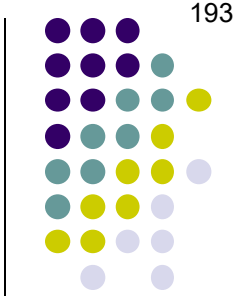

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Did XBRT fall out of favor because it was ineffective?

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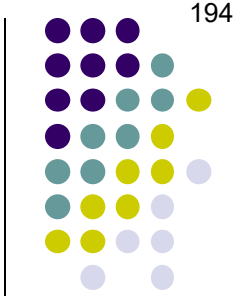

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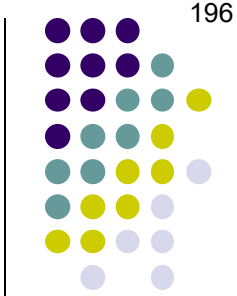

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Because it significantly increases the risk of secondary malignancies later in life

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Q

What classification system has replaced the outmoded Reese-Ellsworth system?

- Cor
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A

What classification system has replaced the outmoded Reese-Ellsworth system?
The International Classification for Intraocular Retinoblastoma (ICIR)

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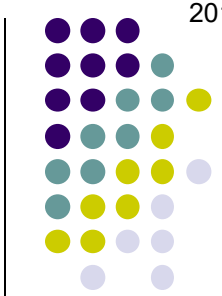

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How are the five groups in the ICIR defined?

- Cor
  - T

Group	***Description***	*Risk of losing the eye if tx'd with chemo only*
A	*(Start here)*	
B		
C		
D		
E		

sh

A

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B		
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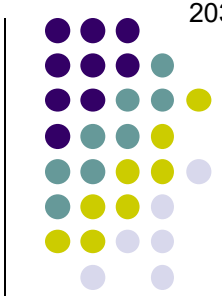
D		
E		



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B		
C		
D		
E		

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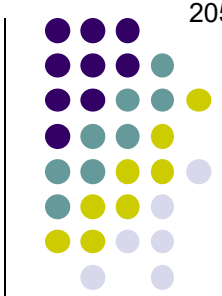

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B	*(Next)*	
C		
D		
E		

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C		
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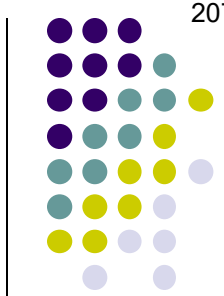

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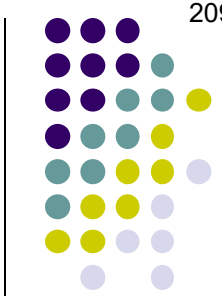

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D		
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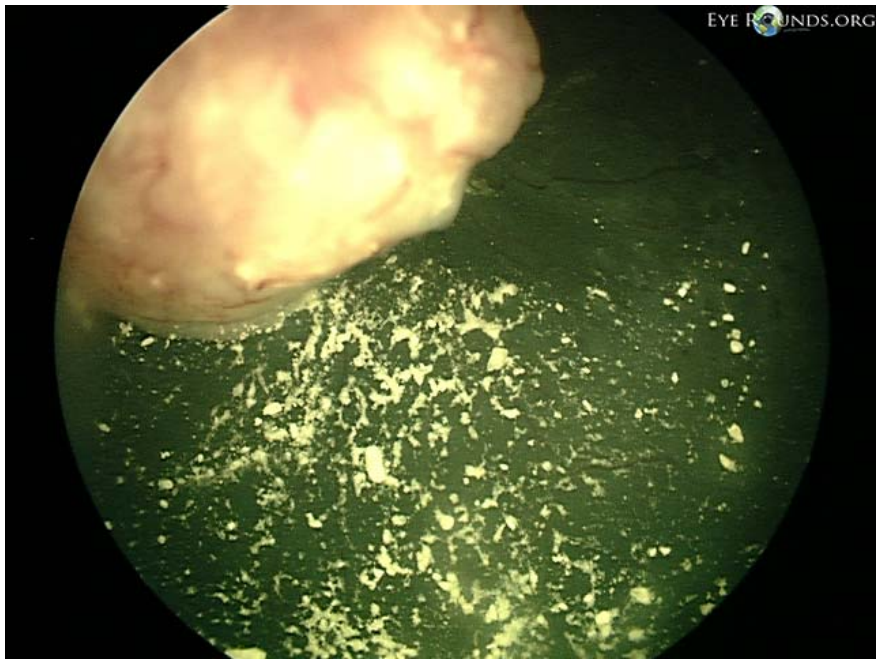
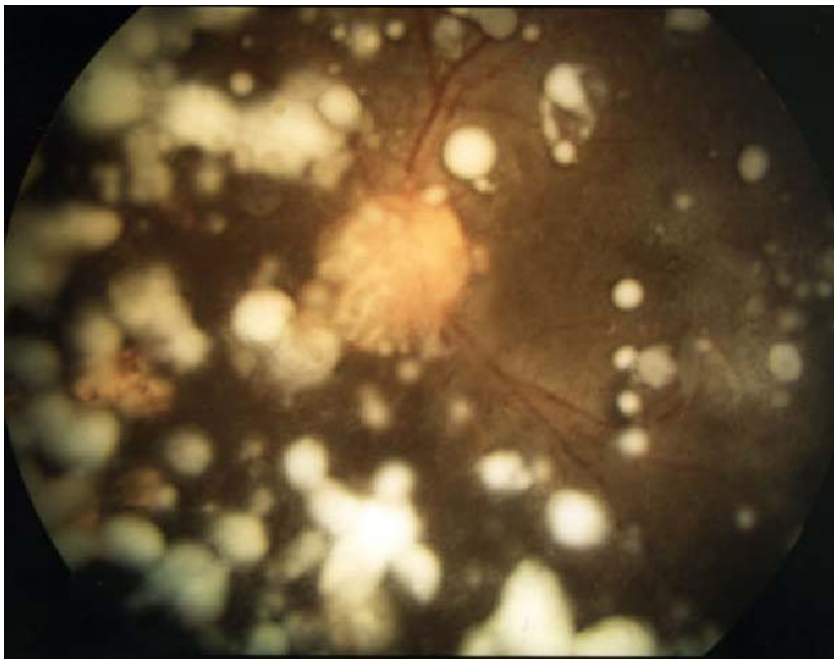
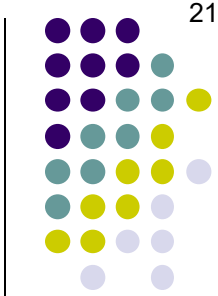
D		
E		

Coats Disease vs Retinoblastoma





Rb: Vitreous seeding



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E		

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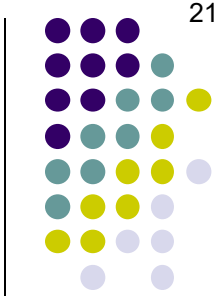
D	Extensive extraretinal spread	
E		



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Group	*Description*	***Risk of losing the eye if tx'd with chemo only***
A	Tumor(s) confined to retina, small, and far from the foveola and ONH	Very low
B	Tumor(s) confined to retina; otherwise fail to qualify for Group A	Low
C	Local extraretinal spread (ie, subretinal fluid and/or vitreous seeding)	Moderate
D	Extensive extraretinal spread	How low/high?
E		

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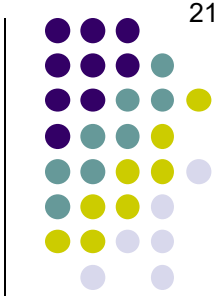

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E	*(Next)*	

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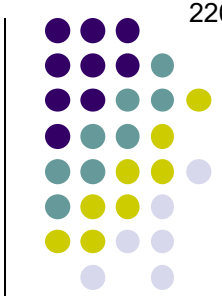

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Q

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How are the five groups in the ICIR ***treated?***

Note the change! We're talking about tx now

- Cor
  - T

Group	*Description*	***Preferred Treatment*** *(in unilateral Rb)*
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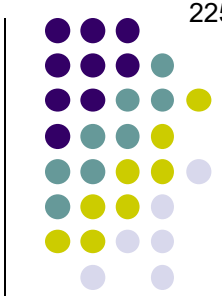
(Chemoreduction is usually not needed for these small, discrete tumors)



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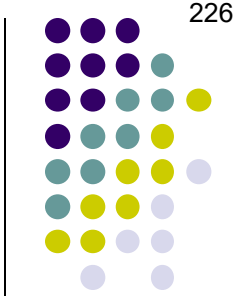

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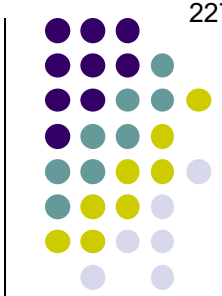

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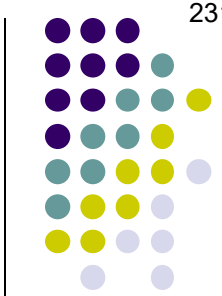

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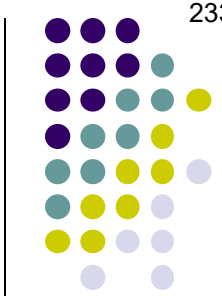

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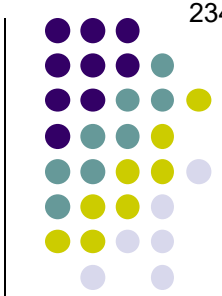
Unilateral Rb TLDR:

(Advance when ready)



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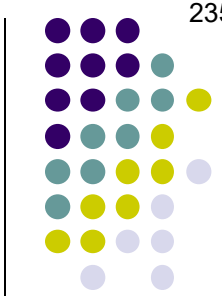
Unilateral Rb TLDR:
For relatively localized tumors: **'Chemoreduction with focal consolidation'**



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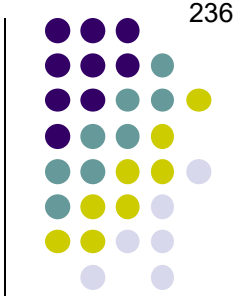
Unilateral Rb TLDR:
For relatively localized tumors: **'Chemoreduction with focal consolidation'**
For advanced tumors: **Enucleation**



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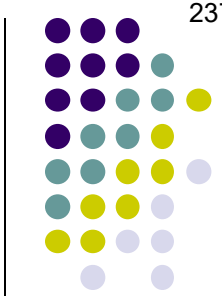
B	Tumor(s) confined to retina; otherwise fail to qualify for Group A	***?***
C		***?***
D	Extensive extraretinal spread	***?***
E	Profoundly compromised eye (eg, NVG, tumor-lens touch, vitreous hemorrhage, aseptic orbital cellulitis secondary to tumor necrosis, etc)	***?***

sh

As for treatment decisions in ***bilateral*** Rb…



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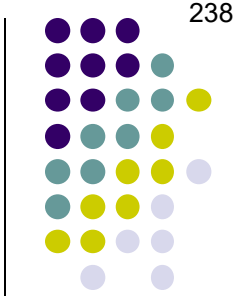
sh

As for treatment decisions in ***bilateral*** Rb… these are considerably more complex, and are beyond the scope of this review.



Q

- Concerning Rb, which of the following are true?
  - The incidence is roughly 1/~~100,000~~ 14K - 20K live births ***F***
  - About 60% represent nonheritable mutations ***T***
  - To diagnose a case as 'heritable,' family history ~~must~~ need not be positive ~~(but not necessarily a 1º relative)~~ ***F***
  - The *exophytic* type looks like Coats disease ***T***
  - Tissue diagnosis via fine-needle aspiration (FNA) or vitreous tap ~~must~~ should not be obtained if enucleation is being contemplated ***F***
  - CT orbits has a role in purely intraocular Rb (i.e., even if no orbital extension is present) ***F***
  - Periodic MRI brain is ~~warranted~~ not to detect early 'trilateral' disease ***F***
  - The Reese-Ellsworth classification system is ~~the current~~ no longer the preferred method for staging Rb ***F***
  - Patients with Rb are more likely to die of a second malignancy than of Rb itself

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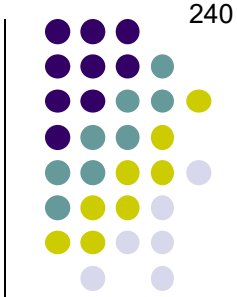

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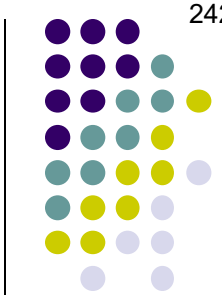
  - The Reese-Ellsworth classification system is ~~the current~~ no longer the preferred method for staging Rb ***F***
  - Patients with Rb are more likely to die of a second malignancy than of Rb itself ***T***
  - The histologic hallmark is the Homer Wright rosette

A

- Concerning Rb, which of the following are true?
  - The incidence is roughly 1/~~100,000~~ (14K - 20K) live births ***F***
  - About 60% represent nonheritable mutations ***T***
  - To diagnose a case as 'heritable,' family history ~~must~~ (need not) be positive ~~(but not necessarily a 1º relative)~~ ***F***
  - The *exophytic* type looks like Coats disease ***T***
  - Tissue diagnosis via fine-needle aspiration (FNA) or vitreous tap ~~must~~ (should not) be obtained if enucleation is being contemplated ***F***
  - CT orbits has a role in purely intraocular Rb (i.e., even if no orbital extension is present) ***F***
  - Periodic MRI brain is (not) warranted to detect early 'trilateral' disease ***F***
  - The Reese-Ellsworth classification system is ~~the current~~ (no longer the) preferred method for staging Rb ***F***
  - Patients with Rb are more likely to die of a second malignancy than of Rb itself ***T***
  - The histologic hallmark is the ~~Homer Wright~~ (Flexner-Wintersteiner) rosette ***F***



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With respect to Rb histology, the term 'rosette' is used in three contexts. What are they?

two-words rosettes

prefix rosettes

two different words rosettes

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'Fleurette'

Q

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  - Abo
  - To
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  - Tiss
  - CT
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14K - 20K

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What is the characteristic appearance of a Flexner-Wintersteiner rosette?

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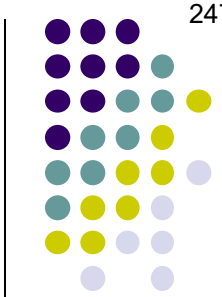
Flexner-Wintersteiner



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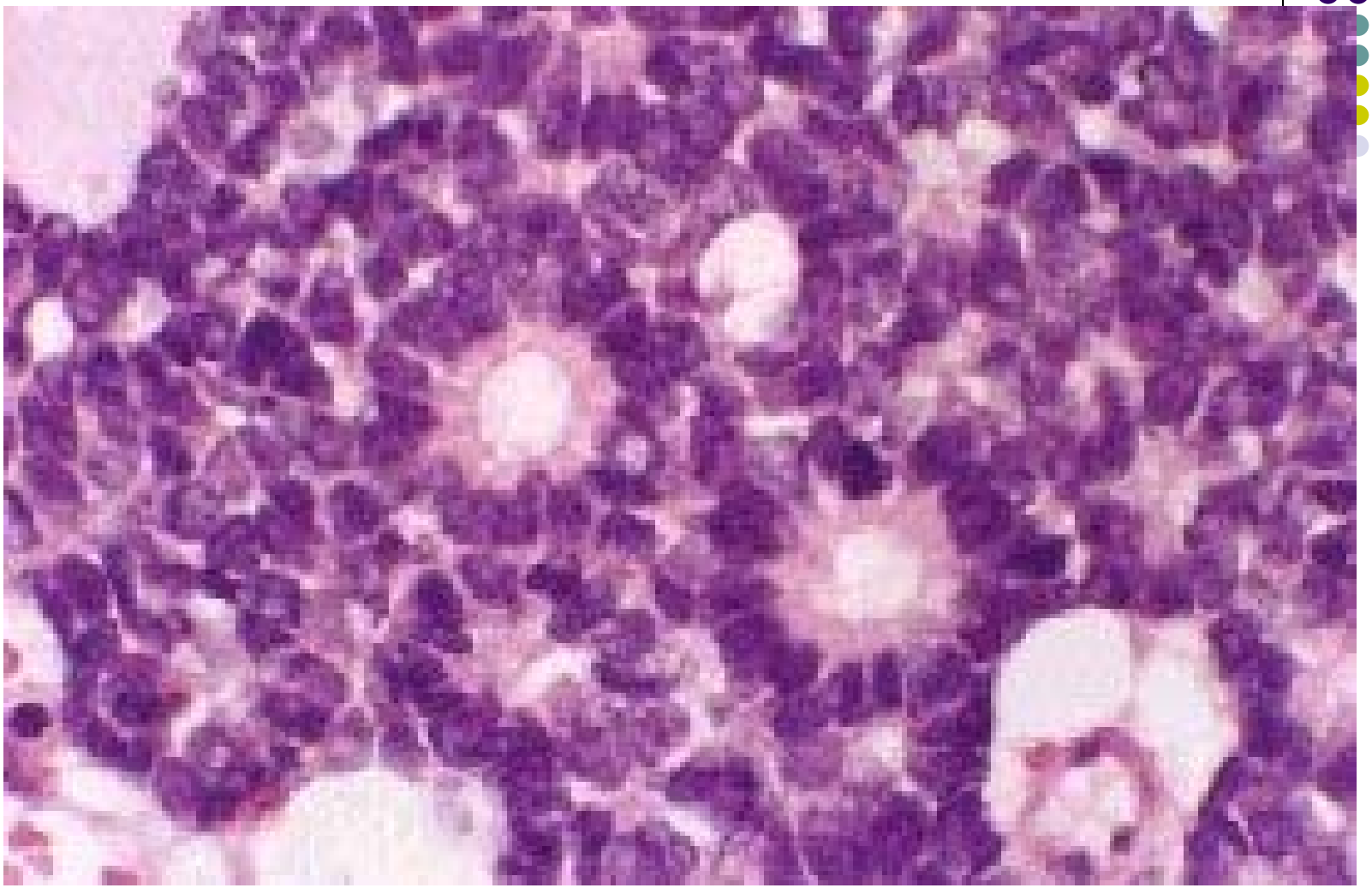

Rb: Flexner-Wintersteiner rosettes

Q

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Yes, but it is lined by a structure often described as one word

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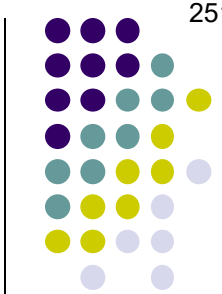

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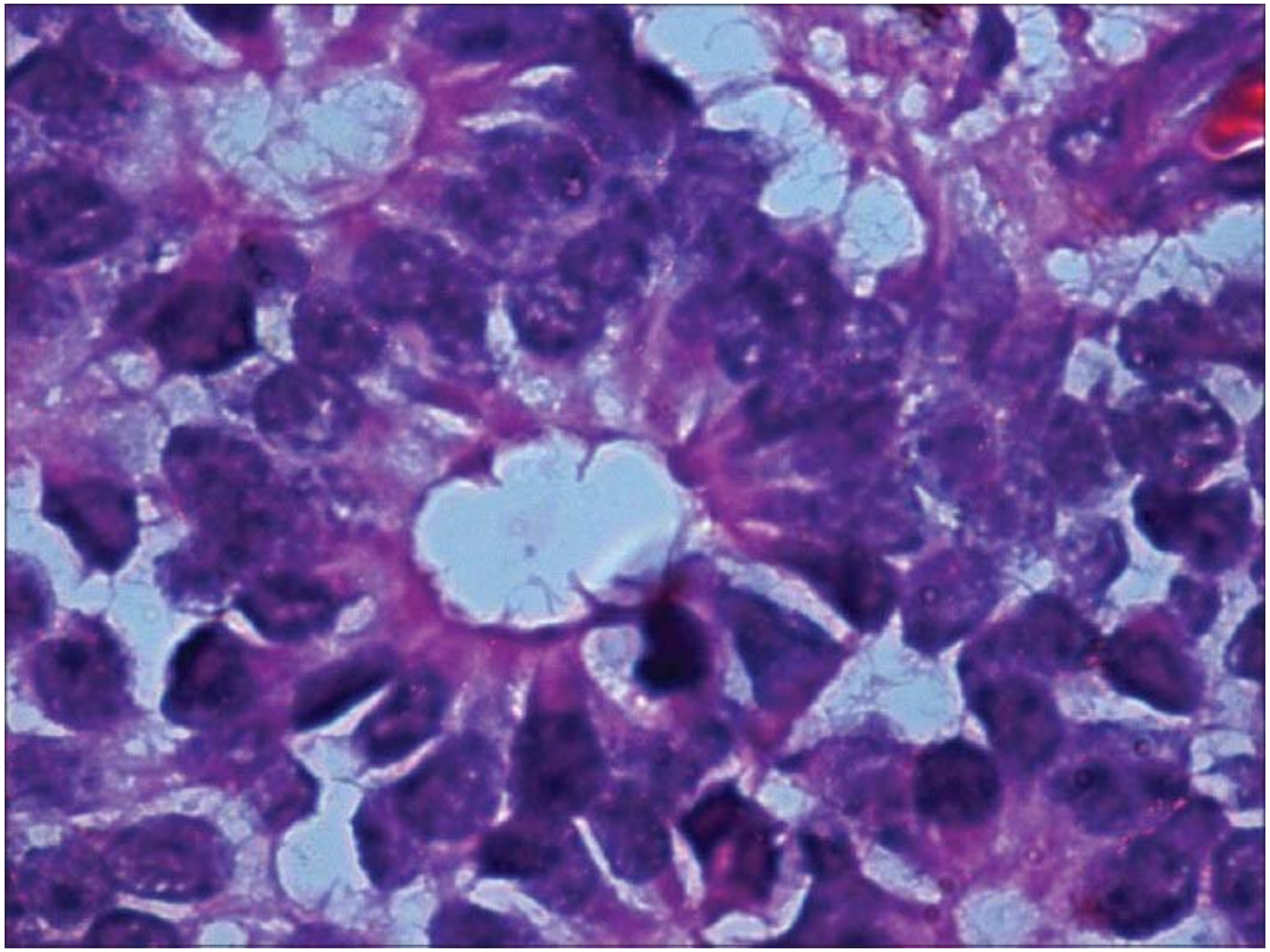
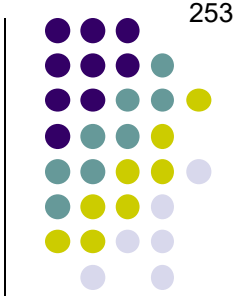


Flexner-Wintersteiner rosette. Note the empty, lined lumen



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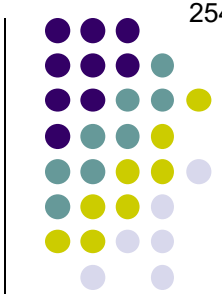

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What normal retinal structure correlates with this refractile lining?
The retinal outer membrane

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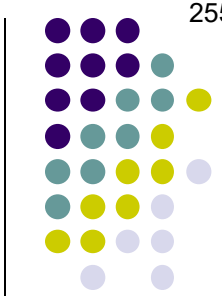

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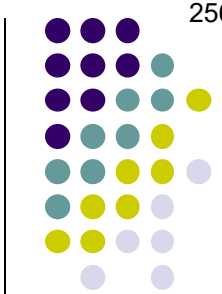

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They represent an attempt at differentiation into retinal structures

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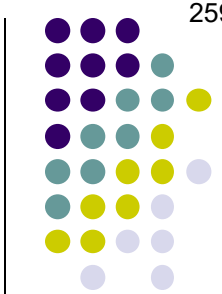

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  - ‘Fleurette’
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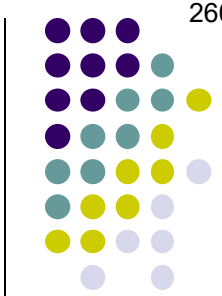
What are pseudorosettes?

Again with respect to Rb histology, another ‘-ette’ term is key. What is it?



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  - [illegible] ey?
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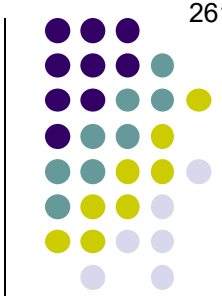
What are pseudorosettes?
A description of the histologic appearance of the tumor with respect to how it organizes around blood vessels



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How does the tumor tend to organize with respect to blood vessels?

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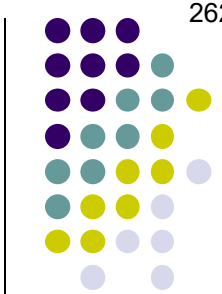

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How does the tumor tend to organize with respect to blood vessels?
Like other fast-growing tumors, Rb has a tendency to ‘outgrow’ its blood supply. That is, tumor cells frequently end up so far from a blood vessel that they are unable to have their metabolic needs met. Cells in these areas die, and subsequently necrose.

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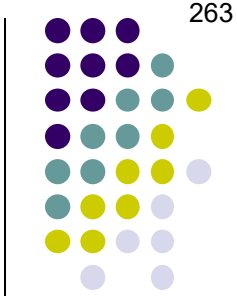

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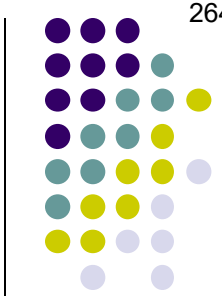

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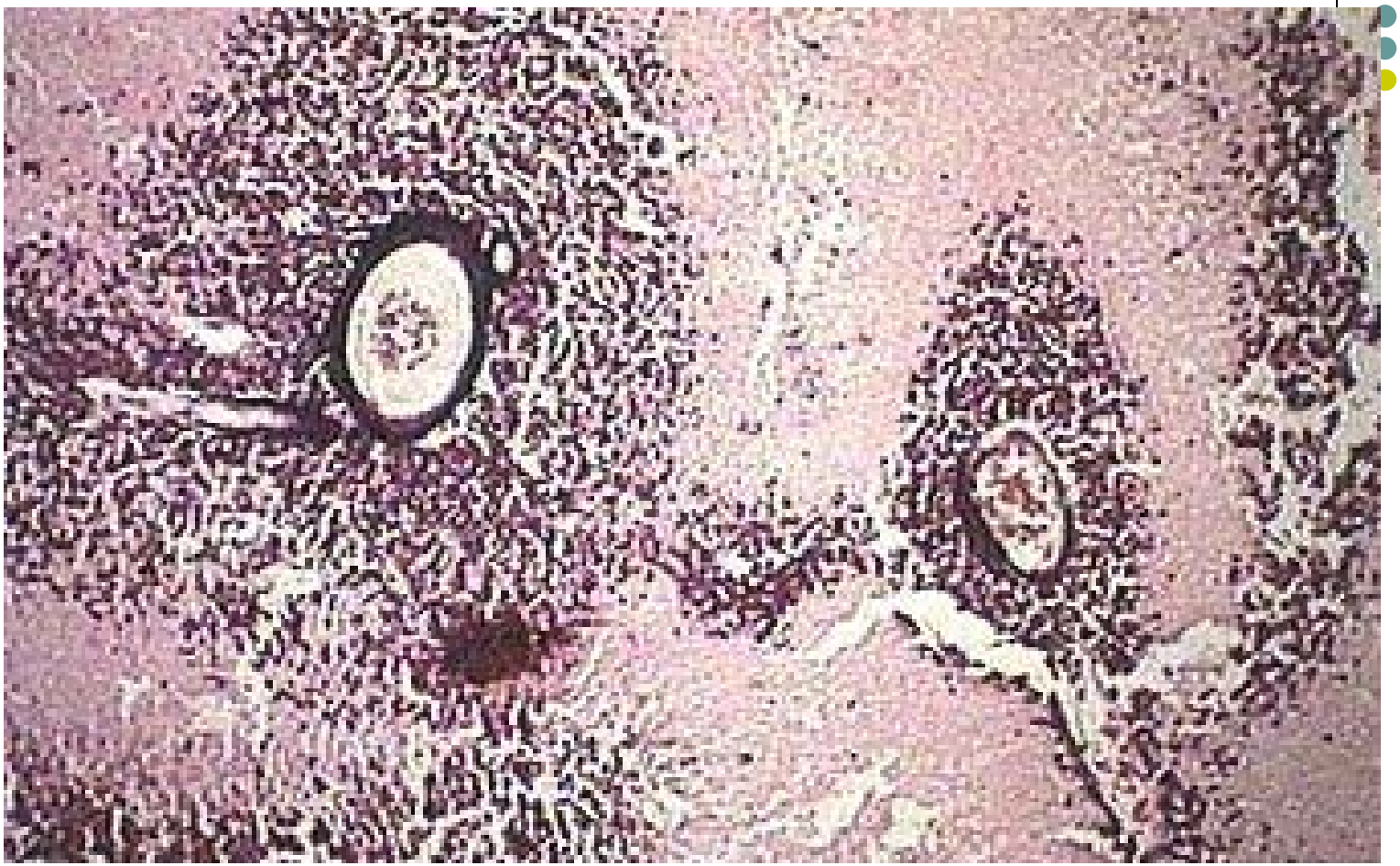
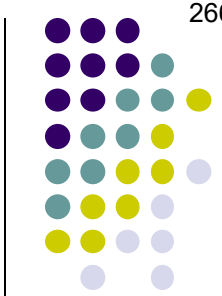
Again with respect to Rb histology, another '-ette' term is key. What is it?
'Fleurette'

  - of Rb itself *T*
  - The histologic hallmark is the ~~Homer Wright~~ Flexner-Wintersteiner rosette *F*



Rb: Pseudorosettes



Q

- Concerning Rb, which of the following are true?
  - The incidence is roughly 1/~~100,000~~ 14K - 20K live births *F*
  - About 60% represent nonheritable mutations *T*
  - an Rb tumor will be characterized by cuffs of living cells surrounding blood vessels, with the cuffs in turn being surrounded by **areas of necrosis** **A pseudorosette is the blood vessel along with its cuff of viable cells.**
  - --Flexner-Wintersteiner rosettes
  - **--Pseudorosettes**
  - --Homer Wright rosettes
  - ‘Fleurette’
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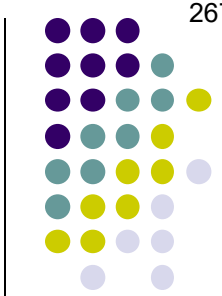
The areas of necrosis are characterized by the presence of a substance of some import. What is it?

Again with respect to Rb histology, another ‘-ette’ term is key. What is it?



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Calcium

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Flexner-Wintersteiner

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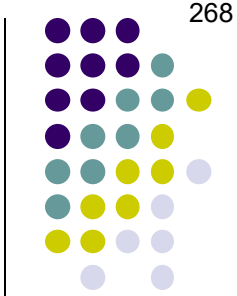

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The areas of necrosis are characterized by the presence of a substance of some import. What is it?
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Why is the presence of calcium within areas of necrosis important?

Again with respect to Rb histology, another '-ette' term is key. What is it?
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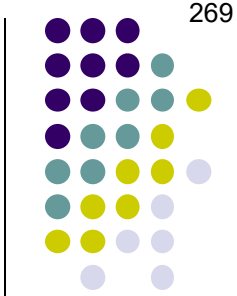

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The areas of necrosis are characterized by the presence of a substance of some import. What is it?
Calcium

Why is the presence of calcium within areas of necrosis important?
It is this calcium that shows up on imaging, thus providing an important diagnostic clue that one is dealing with Rb

Again with respect to Rb histology, another '-ette' term is key. What is it?
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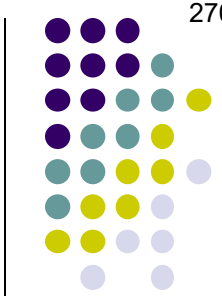

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What is the characteristic appearance of a Homer Wright rosette?

...are they?

--Flexner-Wintersteiner rosettes
--Pseudorosettes
--**Homer Wright rosettes**

Again with respect to Rb histology, another '-ette' term is key. What is it?
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What is the characteristic appearance of a Homer Wright rosette?
Like a Flexner-Wintersteiner rosette, it is composed of a number of retinoblasts organized in a circle around a lumen

… are they?
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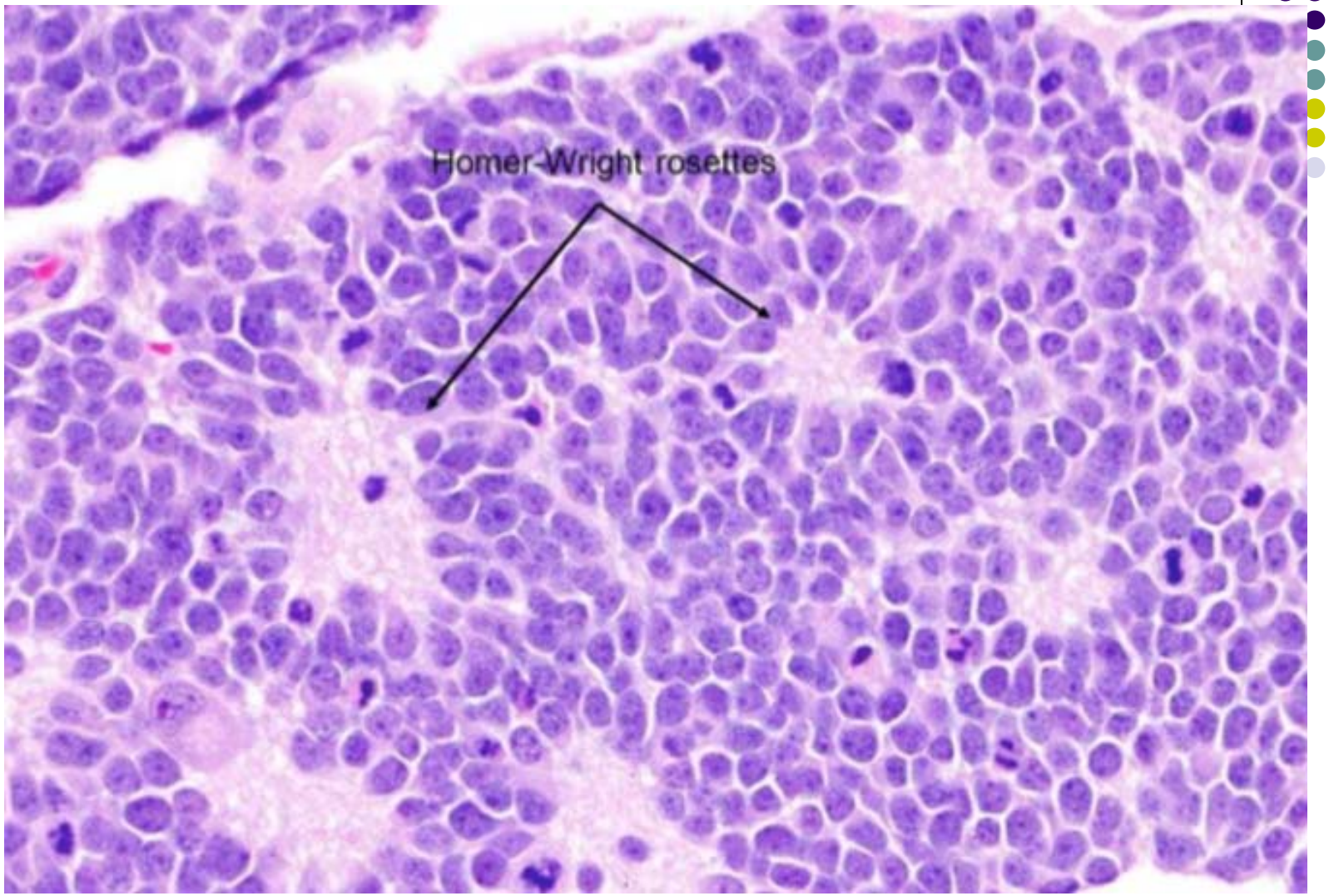
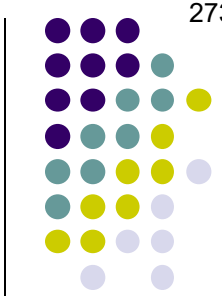


Rb: Homer Wright rosettes



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Is the lumen empty?

are they?
--Flexner-Wintersteiner rosettes
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Again with respect to Rb histology, another '-ette' term is key. What is it?
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Q/A

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What is the characteristic appearance of a Homer Wright rosette?
Like a Flexner-Wintersteiner rosette, it is composed of a number of retinoblasts organized in a circle around a lumen

Is the lumen empty?
No, it contains an eosinophilic structure called a [two words]

shou...

...are they?
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--**Homer Wright rosettes**

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What is the characteristic appearance of a Homer Wright rosette?
Like a Flexner-Wintersteiner rosette, it is composed of a number of retinoblasts organized in a circle around a lumen

Is the lumen empty?
No, it contains an eosinophilic structure called a 'neurofibrillary tangle'

are they?
--Flexner-Wintersteiner rosettes
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Again with respect to Rb histology, another '-ette' term is key. What is it?
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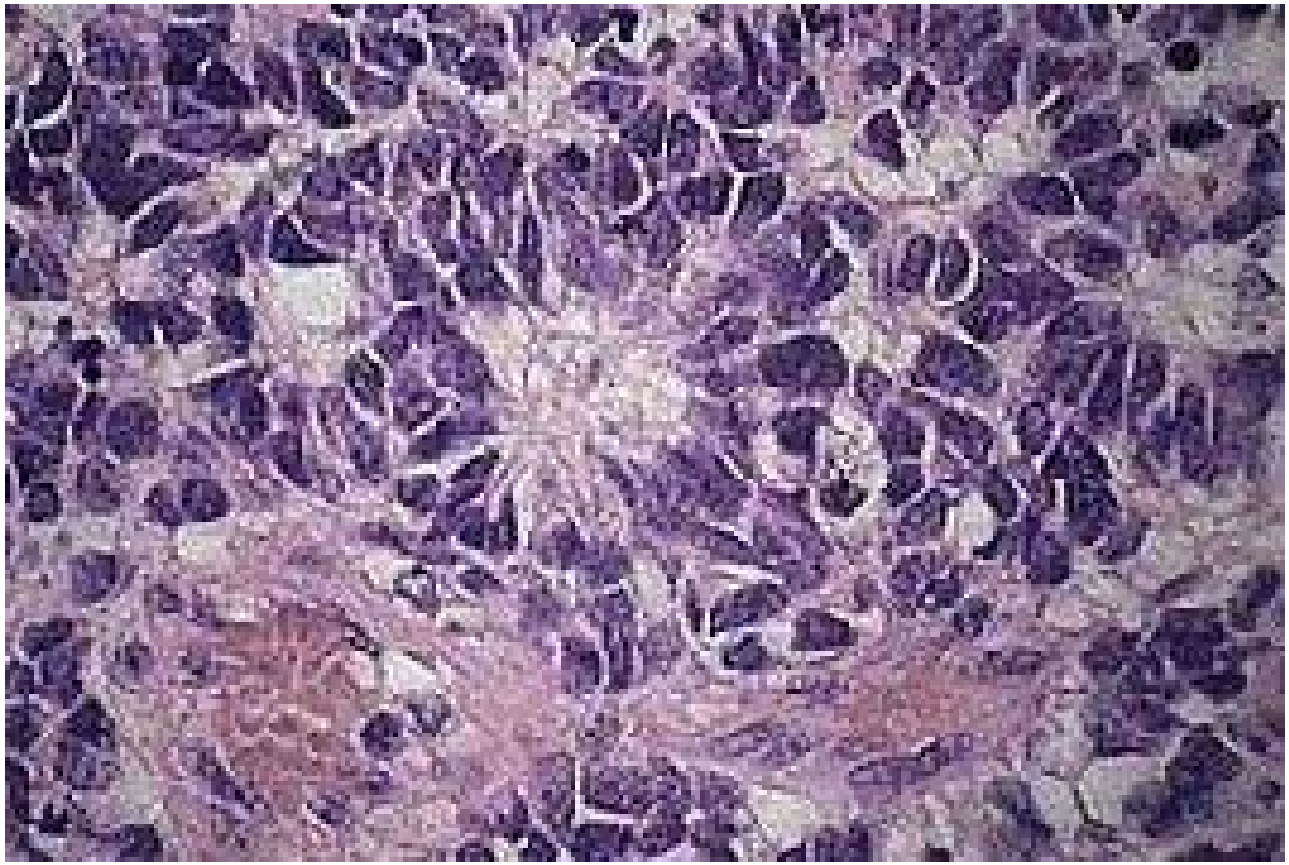

Rb: Homer Wright rosettes. Note the neurofibrillary tangle in the lumen of the rosette

Q

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Is the Homer Wright rosette pathognomonic for Rb?

are they?
--Flexner-Wintersteiner rosettes
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Is the lumen empty?
No, it contains an eosinophilic structure called a 'neurofibrillary tangle'

Is the Homer Wright rosette pathognomonic for Rb?
No. It is not always encountered in Rb, and is commonly present in other tumors

...are they?
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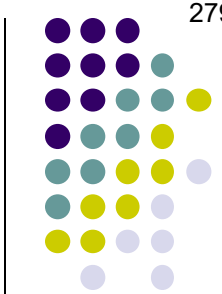
'Fleurette'



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  - ‘Fleurette’
  - of Rb itself *T*
  - The histologic hallmark is the ~~Homer Wright~~ Flexner-Wintersteiner rosette *F*

What is a fleurette*?*

*Again with respect to Rb histology, another ‘-*ette*’ term is key. What is it?*

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  - ~~Homer Wright rosettes~~ Flexner-Wintersteiner rosettes
  - ~~Again with respect to Rb histology, another '-ette' term is key. What is it?~~ **'Fleurette'**
  - of Rb itself *T*
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What is a fleurette*?*
A small cluster of Rb cells that have differentiated into photoreceptor-like structures

Q

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What does it look like?

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A/Q

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What is a fleurette*?*
A small cluster of Rb cells that have differentiated into photoreceptor-like structures

What does it look like?
It is a curvilinear structure, with extensions described as one word

Again with respect to Rb histology, another '-ette' term is key. What is it?

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- of Rb itself *T*
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What is a fleurette*?*
A small cluster of Rb cells that have differentiated into photoreceptor-like structures

What does it look like?
It is a curvilinear structure, with extensions described as 'bulbous'

vitreous tap
d *F*
n if no orbital

hat are they?
e *F*
ed

Again with respect to Rb histology, another '-ette' term is key. What is it?

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Flexner-Wintersteiner

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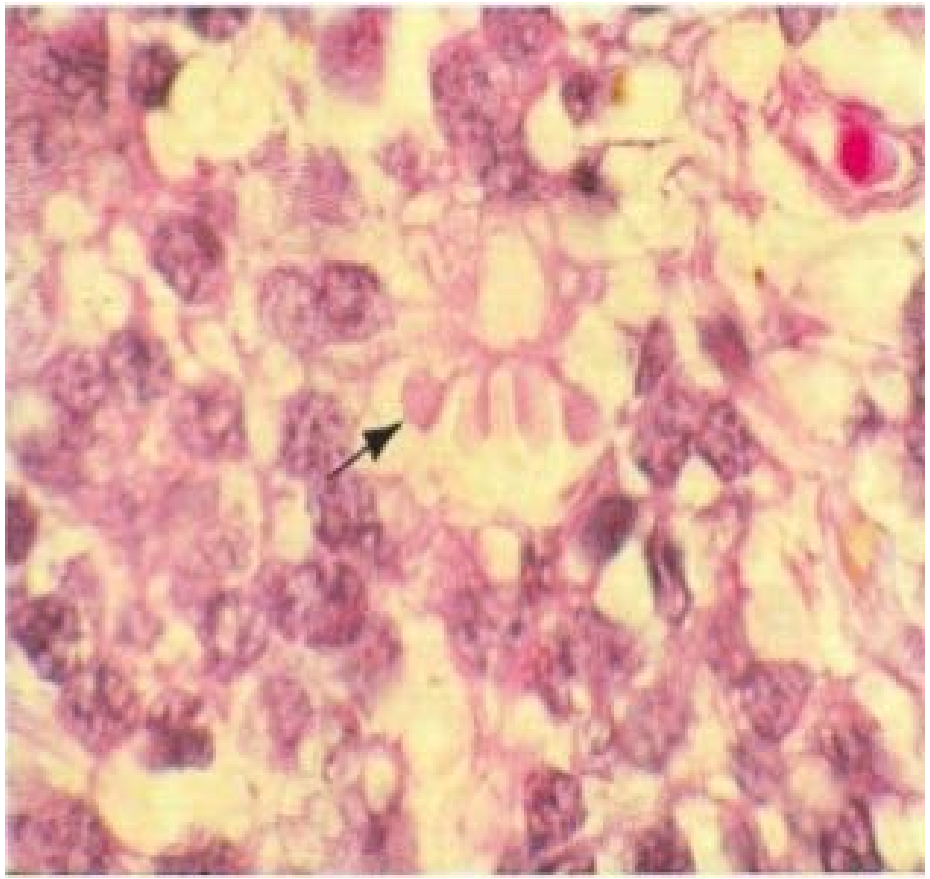

Rb: Fleurette. Note the bulbous extensions (arrow)

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What does it look like?
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Are fleurettes more, or less common that Flexner-Wintersteiner rosettes?

What are they?

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Are fleurettes more, or less common that Flexner-Wintersteiner rosettes?
As fleurettes represent a more advanced form of tumor-cell differentiation, it should come as no surprise that they are more v less commonly encountered than are Flexner-Wintersteiner rosettes

Again with respect to Rb histology, another '-ette' term is key. What is it?

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Are fleurettes more, or less common that Flexner-Wintersteiner rosettes?
As fleurettes represent a more advanced form of tumor-cell differentiation, it should come as no surprise that they are less commonly encountered than are Flexner-Wintersteiner rosettes

vitreous tap
d *F*
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hat are they?
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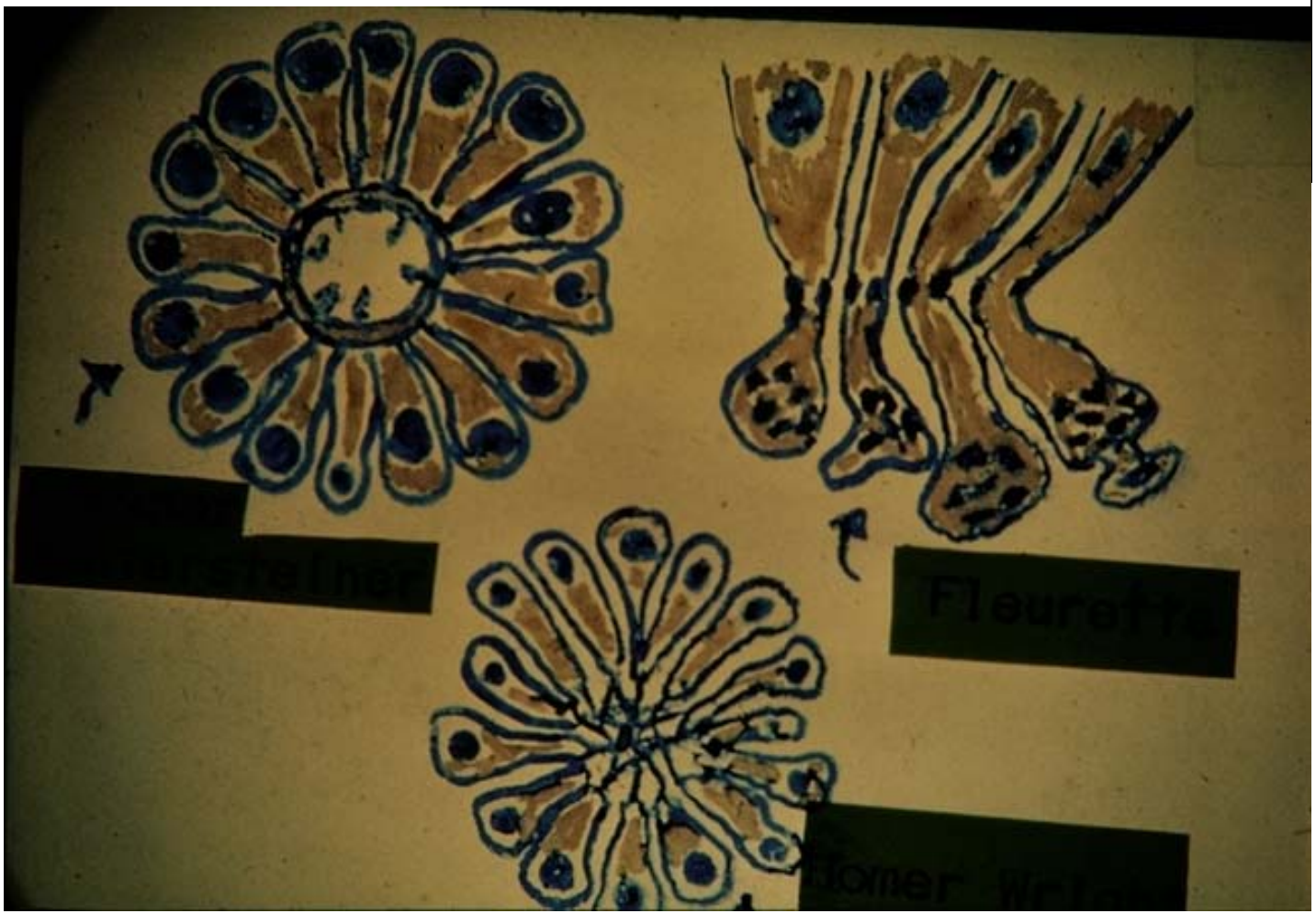

Not a great pic, but the best I could find for comparing and contrasting *F-W rosettes*, *Homer Wright rosettes*, and *fleurettes*