Is it ICE, PPMD or Fuchs?

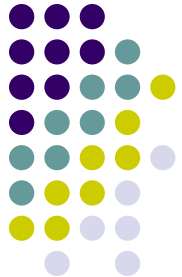
ICE =

PPMD =

Fuchs =

What do ICE *and* PPMD *stand for, and which Fuchs are we likely talking about?*

Is it ICE, PPMD or Fuchs?



ICE = Iridocorneal endothelial syndrome

PPMD = Posterior polymorphous dystrophy

Fuchs = Fuchs endothelial dystrophy

Is it ICE, PPMD or Fuchs?
Assign each statement to the proper condition

ICE
--

PPMD
--

Fuchs
--

Laterality: Which is…
--Essentially always unilateral
--Essentially always bilateral
--Bilateral, but can be so asymmetric as to appear unilateral

Is it ICE, PPMD or Fuchs?

Assign each statement to the proper condition

ICE
--Essentially always unilateral

Laterality: Which is…
--Essentially always unilateral
--Essentially always bilateral
--Bilateral, but can be so asymmetric as to appear unilateral

PPMD
--Bilateral, but can be so asymmetric as to appear unilateral

Fuchs
--Essentially always bilateral

Is it ICE, PPMD or Fuchs?
Assign each statement to the proper condition

ICE
--Essentially always unilateral
--

Etiology: Which is…
--Autosomal dominant (2 of them)
--Nonfamilial

PPMD
--Bilateral, but can be so asymmetric as to appear unilateral
--

Fuchs
--Essentially always bilateral
--

Is it ICE, PPMD or Fuchs?
Assign each statement to the proper condition

ICE
--Essentially always unilateral
--**Nonfamilial**

Etiology: Which is…
--Autosomal dominant (2 of them)
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PPMD
--Bilateral, but can be so asymmetric as to appear unilateral
--**Autosomal dominant**

Fuchs
--Essentially always bilateral
--**Autosomal dominant**

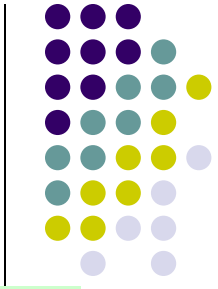
Is it ICE, PPMD or Fuchs?
Assign each statement to the proper condition

ICE
--Essentially always unilateral
--Nonfamilial
--

Gender predilection: Which is…
--Female > male
--Female > male
--Female = male

PPMD
--Bilateral, but can be so asymmetric as to appear unilateral
--Autosomal dominant
--

Fuchs
--Essentially always bilateral
--Autosomal dominant
--



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Assign each statement to the proper condition

ICE
--Essentially always unilateral
--Nonfamilial
--**Female > male**

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--Female > male
--Female = male

PPMD
--Bilateral, but can be so asymmetric as to appear unilateral
--Autosomal dominant
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Fuchs
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Is it ICE, PPMD or Fuchs?
Assign each statement to the proper condition

ICE
--Essentially always unilateral
--Nonfamilial
--Female > male
--

Classic endothelial finding: Which has…
--Guttae
--Bands, vesicles, variable opacities
--'Hammered silver' appearance

PPMD
--Bilateral, but can be so asymmetric as to appear unilateral
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--Female = male
--

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Is it ICE, PPMD or Fuchs?
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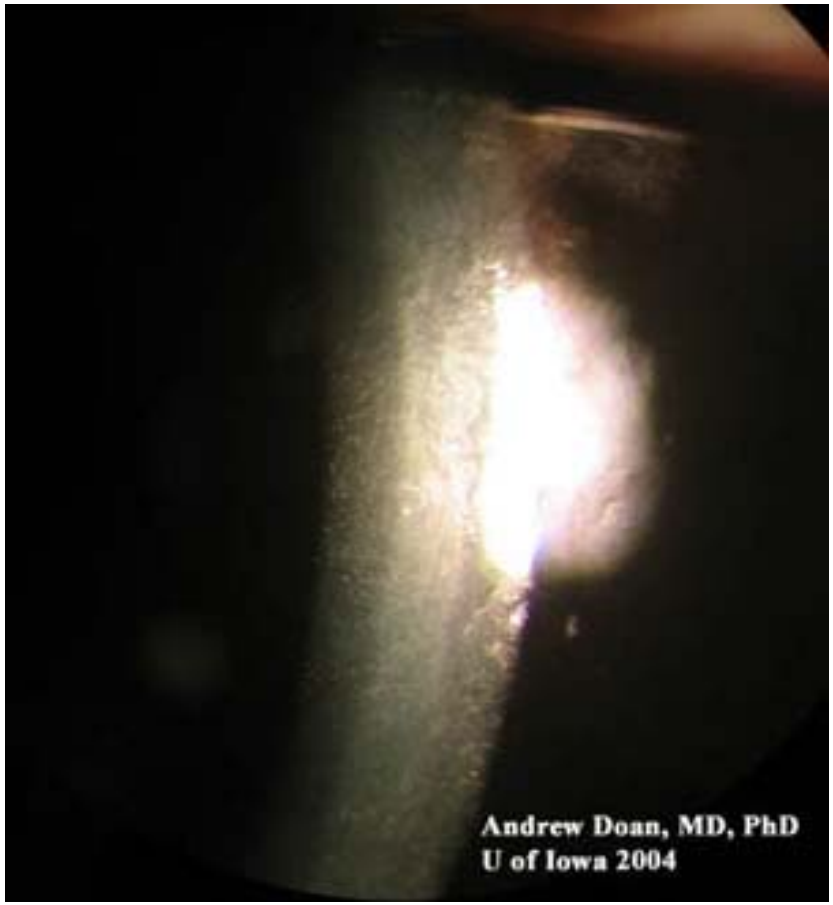
Is it ICE, PPMD or Fuchs?
Assign each statement to the proper condition



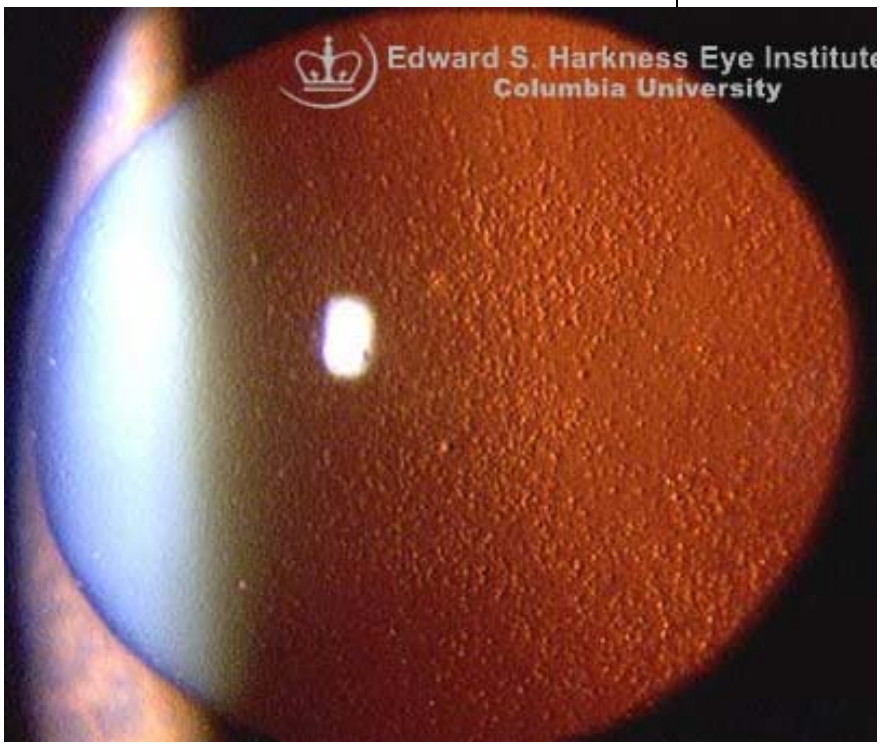
Low res

High res

ICE: ‘Hammered silver’ corneal endothelium

Is it ICE, PPMD or Fuchs?
Assign each statement to the proper condition

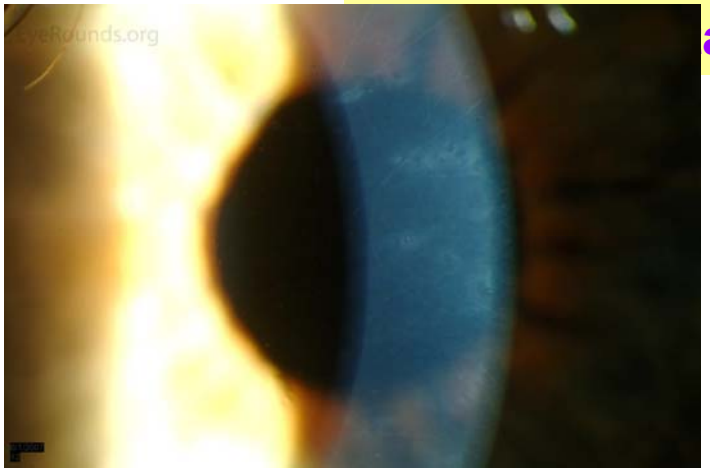
Direct illumination



Retroillumination

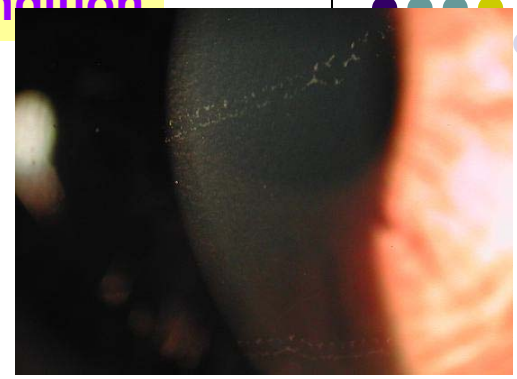
Fuchs: Cornea guttata

Is it ICE, PPMD or Fuchs?

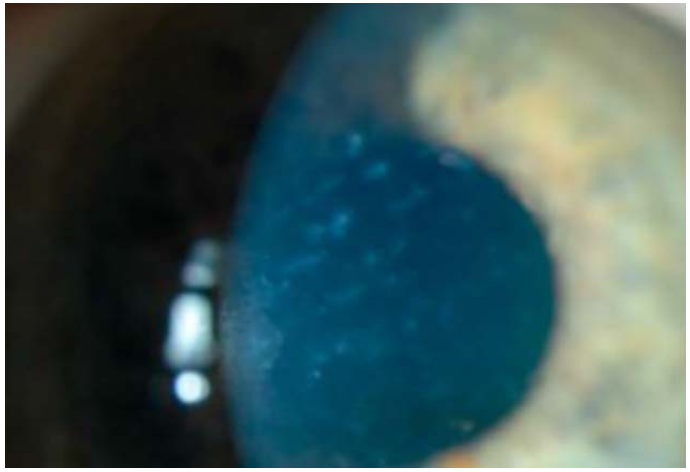


Vesicular lesions

Vesicular lesions

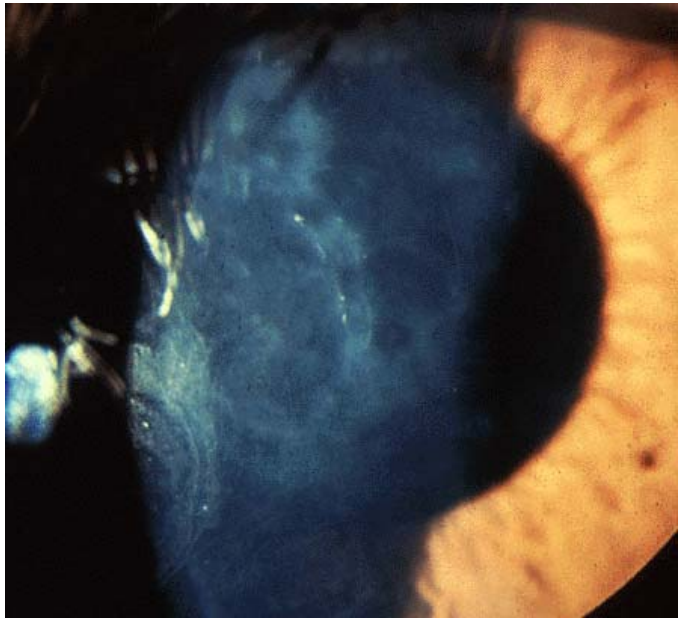


Snail or *railroad tracks*

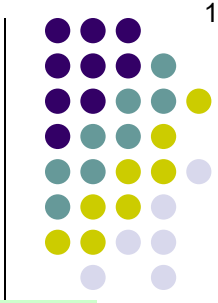


Endothelial plaque-like lesions

PPMD



The clinical appearance of PPMD is highly variable



Is it ICE, PPMD or Fuchs?
Assign each statement to the proper condition

ICE
--Essentially always unilateral
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Glaucoma status: Which has a…
--Strong association with glaucoma
--?No association with glaucoma?
--Modest association with glaucoma

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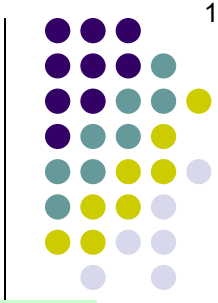

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Cornea status: For which is it the case that…
--Corneal edema common
--Corneal edema can occur, but is not a hallmark
--Corneal edema a defining feature of some forms

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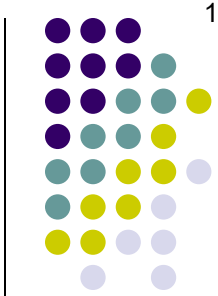

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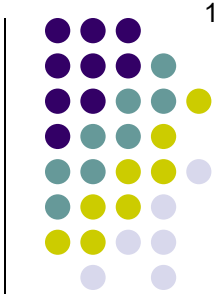
(Summary/review slide—no question, proceed when ready)



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What are the different subtypes of ICE*?*

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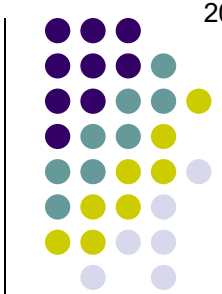

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Two are well established and accepted; they are:
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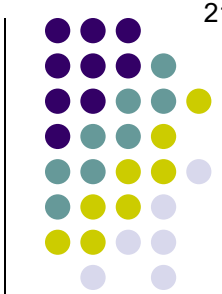

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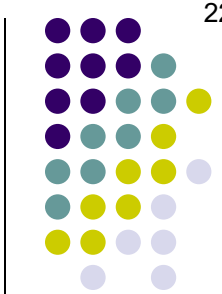

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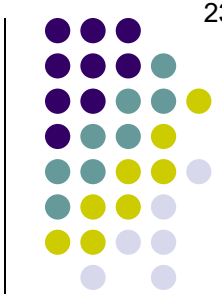

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--Iris nevus syndrome
(See the ICE slide-set, *K26*, for more details.)

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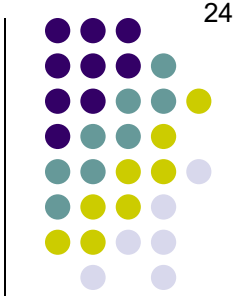

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ICE
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--'H
--S
--C

For the OKAP, I recommend thinking of them as the BCSC does:
Iris nevus syndrome (aka Cogan-Reese syndrome)
Chandler syndrome
Essential iris atrophy

PP
--B
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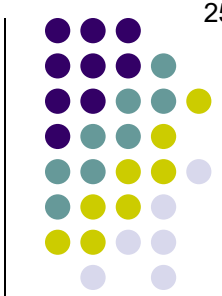

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C*handler syndrome*
E*ssential iris atrophy*

Note that doing this facilitates a very useful mnemonic for remembering the ICE subtypes!

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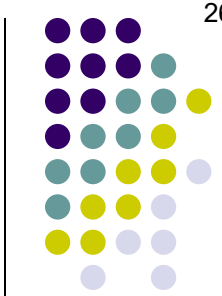

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For which form is the presence of significant corneal edema a defining feature?

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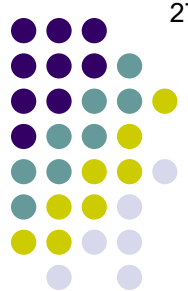

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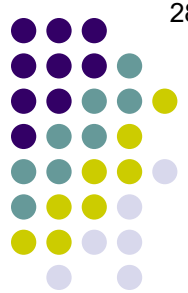

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PPMD

What is abnormal about the endothelial cells in PPMD?

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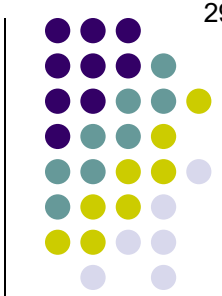

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What is abnormal about the endothelial cells in PPMD?
In PPMD, the endothelial cells 'behave' like epithelial cells and/or fibroblasts; ie, they **proliferate**, form **multiple layers**, and **migrate**

Fuchs
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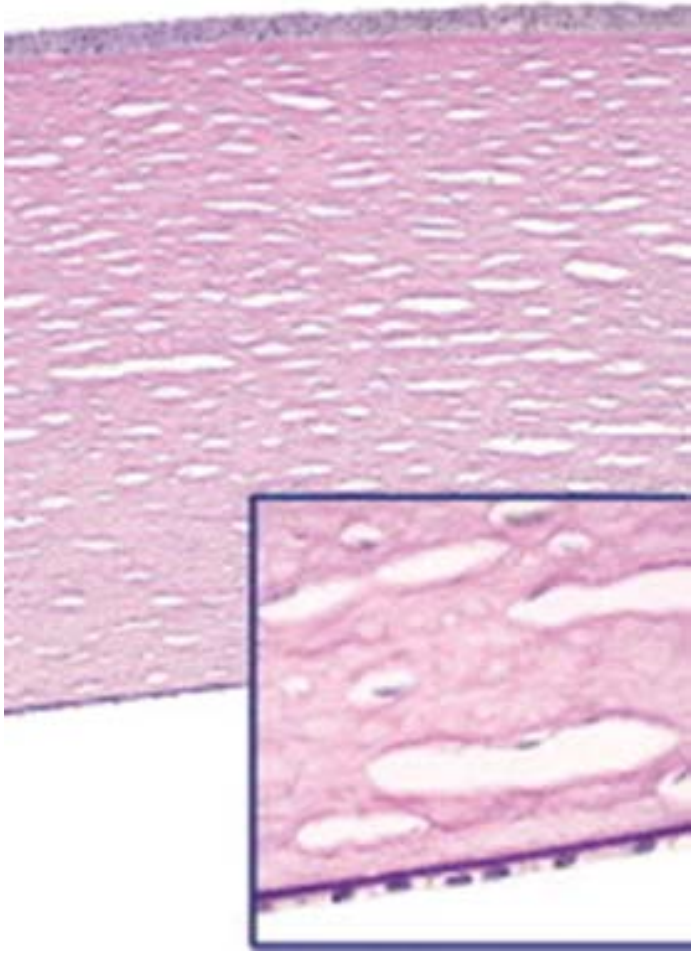

Normal cornea. Note the single-cell-thick nature of the endothelial cells.

Is it ICE, PPMD or Fuchs?
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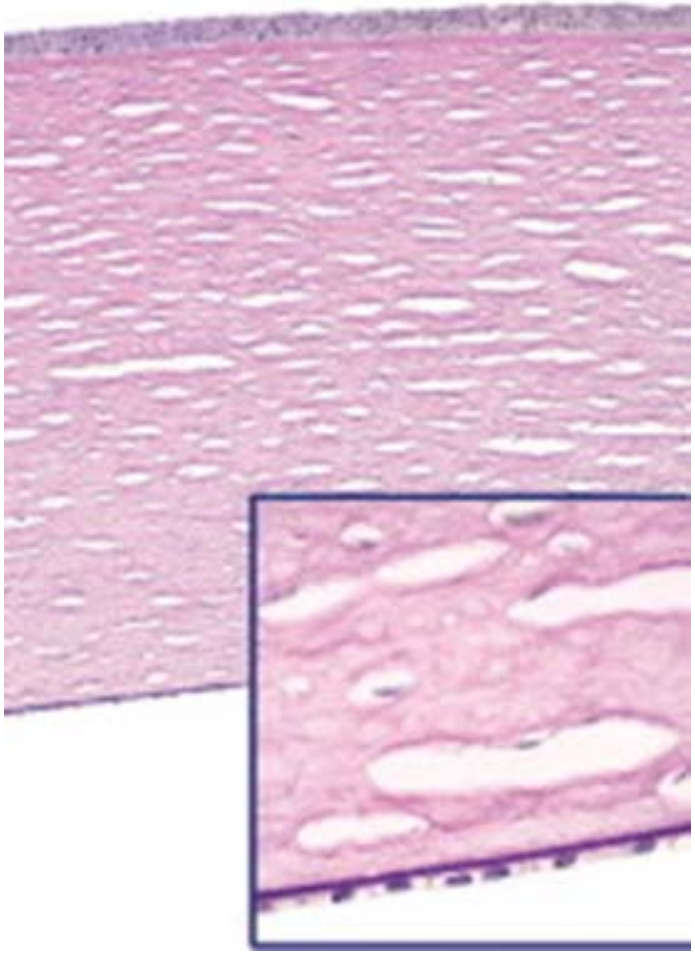

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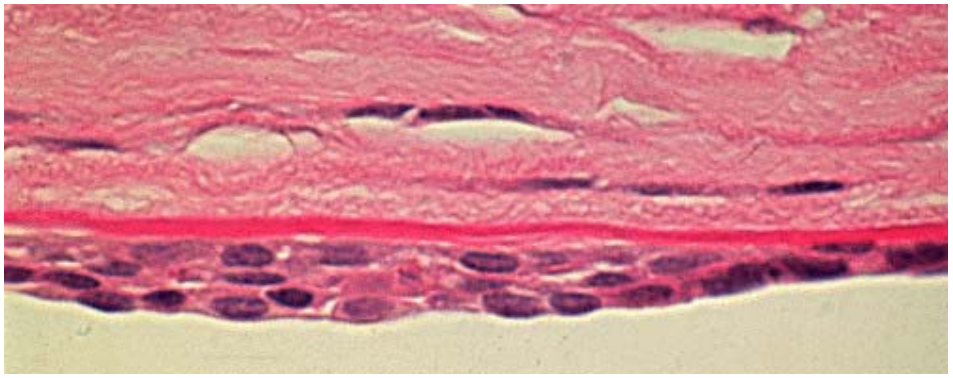
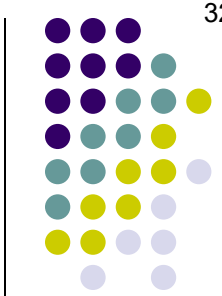


PPMD. Instead of being lined by cells with the attributes of corneal endothelium, the posterior cornea is covered by cells with **epithelial-** or **fibroblast-like** features.



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What condition manifests findings that render it easily confusable with PPMD?

--?No association with glaucoma?
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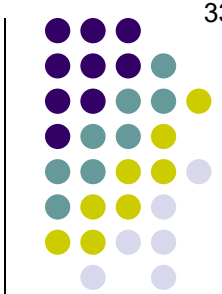

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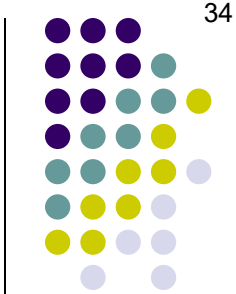

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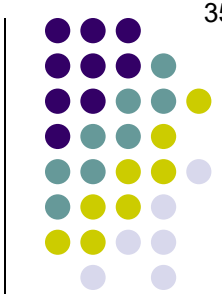

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--Unlike PPMD, ICE is always…
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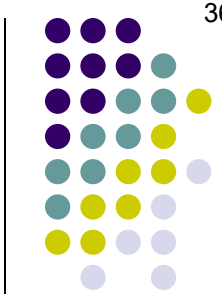

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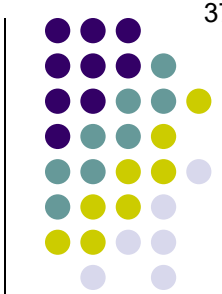

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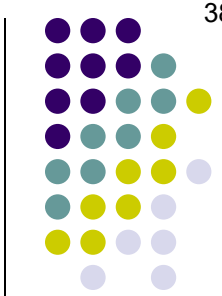
Iridocorneal endothelial (ICE) syndrome

How can you tell if a pt has PPMD vs ICE?
--Unlike PPMD, ICE is always…**unilateral**
--Unlike PPMD, ICE is always…**sporadic**
--?No association with glaucoma?
--Corneal edema common



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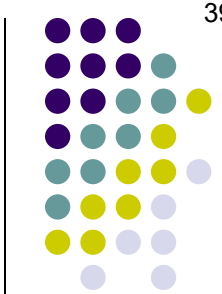

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Wartlike excrescences on Descemet's membrane

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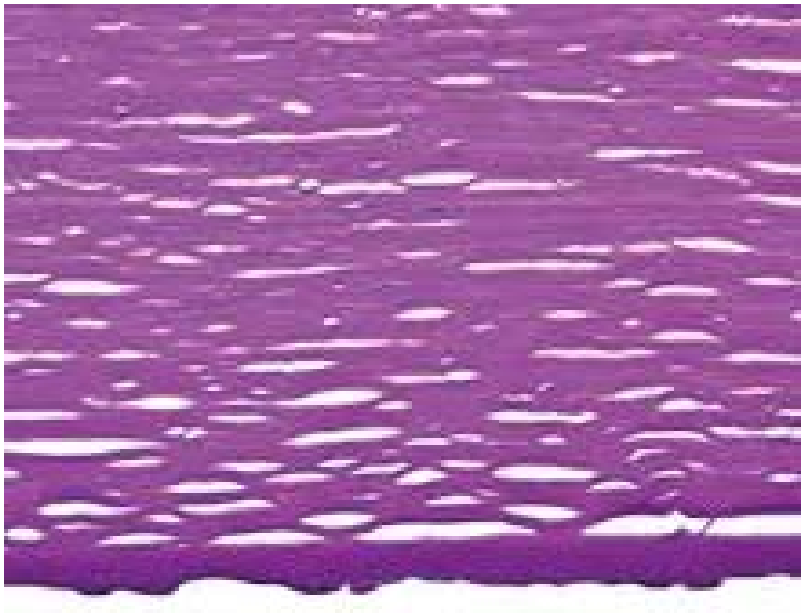
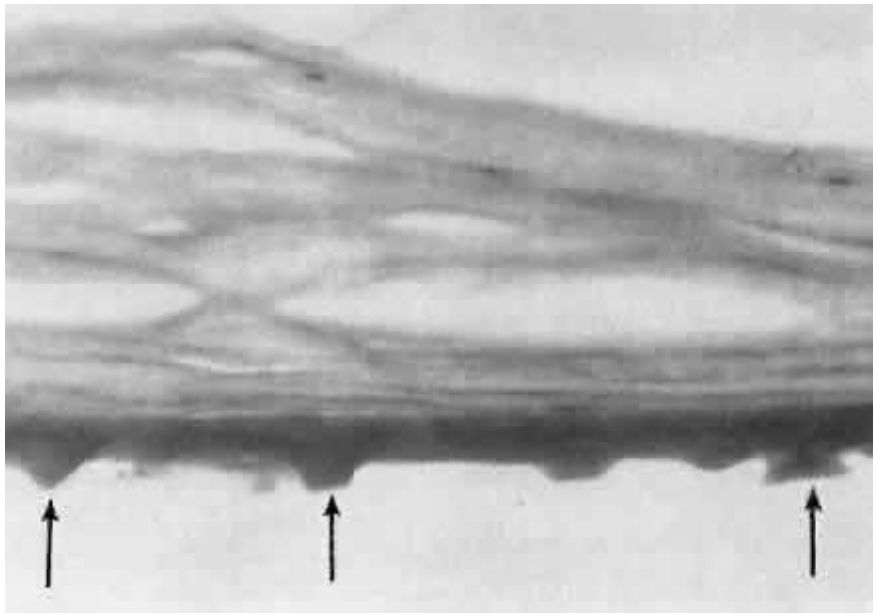
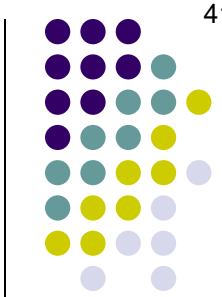




Fuchs endothelial corneal dystrophy. Light microscopy: cornea guttata in the form of focal excrescences at the level of the endothelium.



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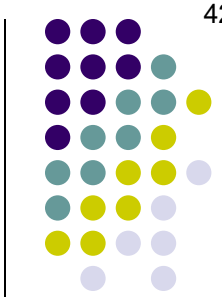

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'Beaten bronze'

Is it ICE, PPMD or Fuchs?
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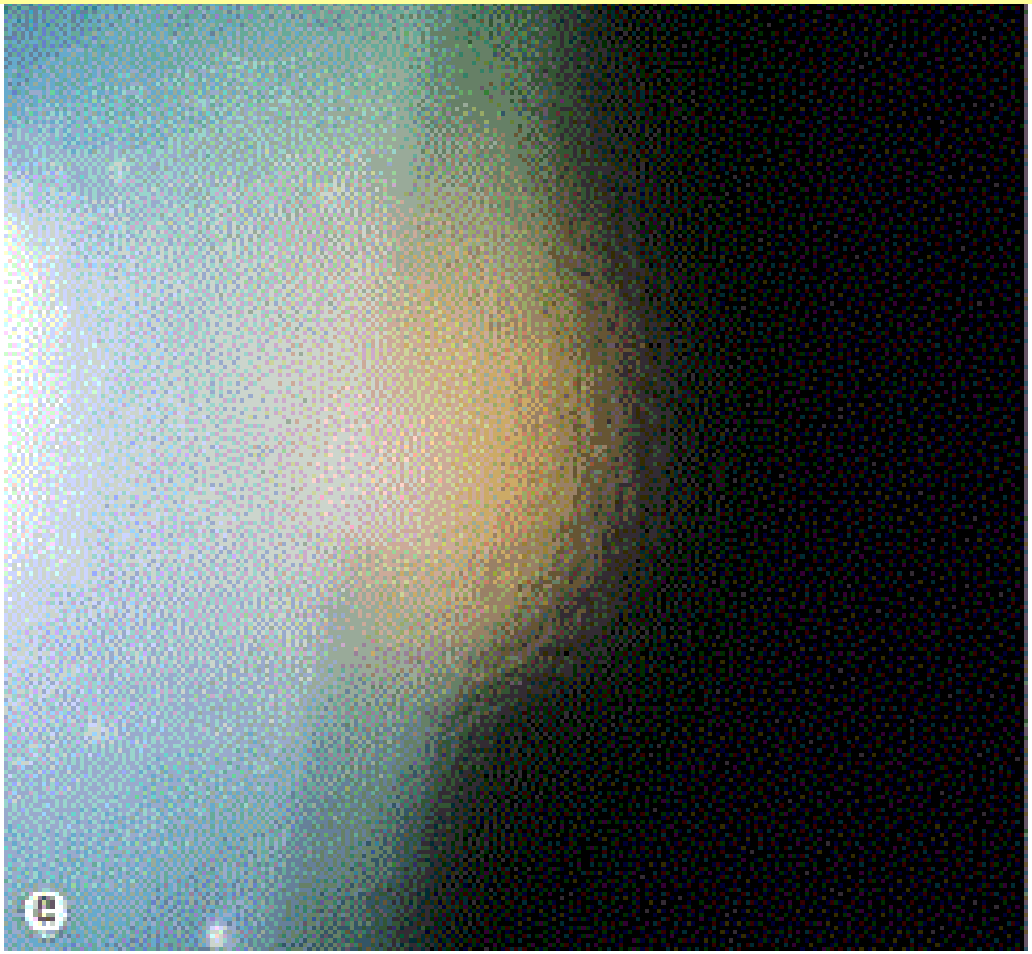
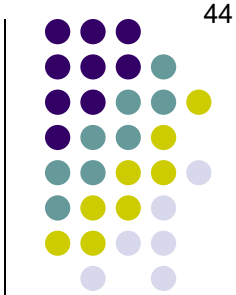


Fuchs endothelial corneal dystrophy. The appearance wrought by dense guttata has been likened to that of '**beaten bronze**.'



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W[illegible] are guttae?
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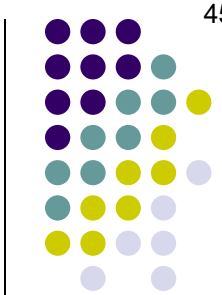
Don't confuse *beaten bronze* with *hammered silver!*



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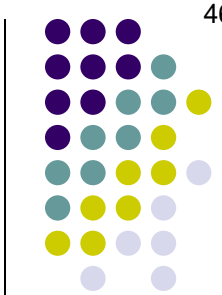

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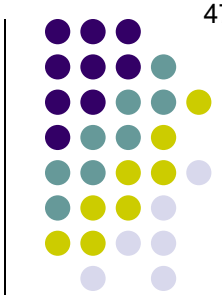

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--The presence of cells that are much too large ( )
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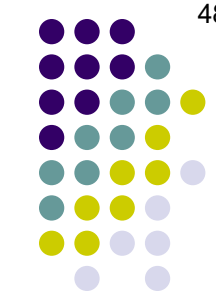

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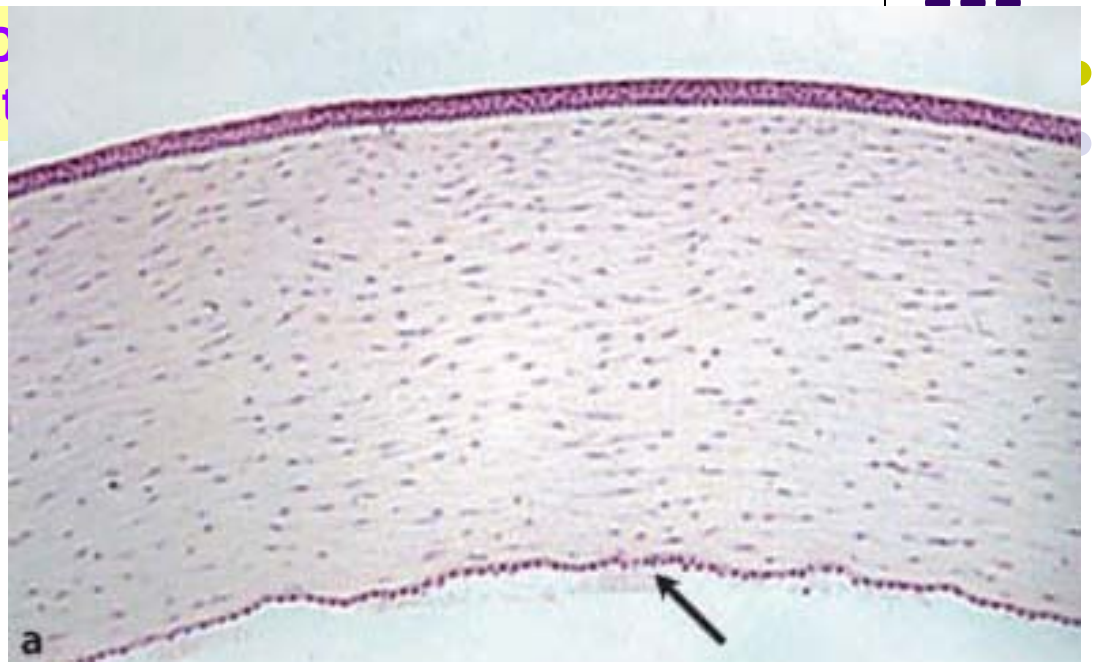
Endothelial cell abnormalities including:
--The presence of guttata (duh)
--Decreased cell density
--The presence of cells that are much too large (*polymegathism*)
--An increase in cell-to-cell variability in size (*pleomorphism*)

Is it IC
Assign each stat



Normal human cornea. Note numerous endothelial cell nuclei lining the posterior surface (*arrow*).

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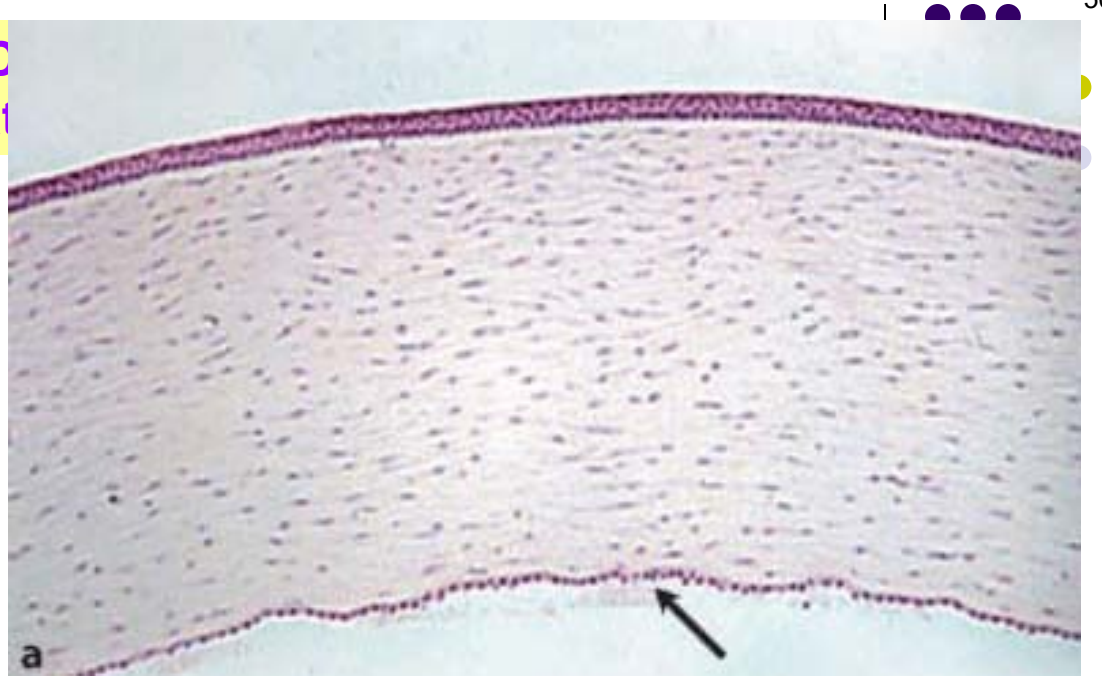

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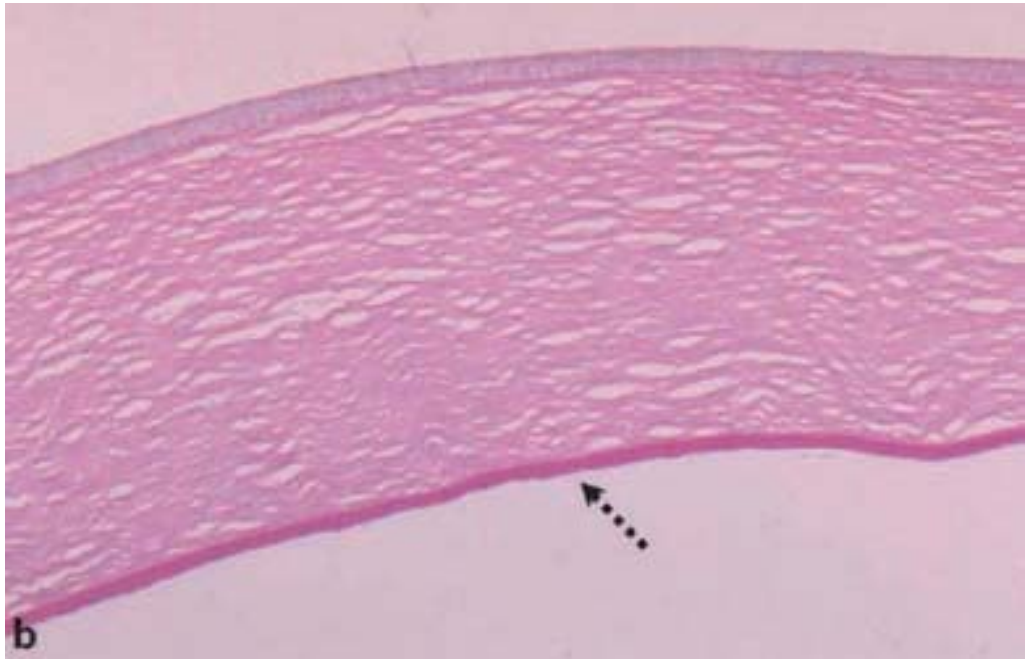
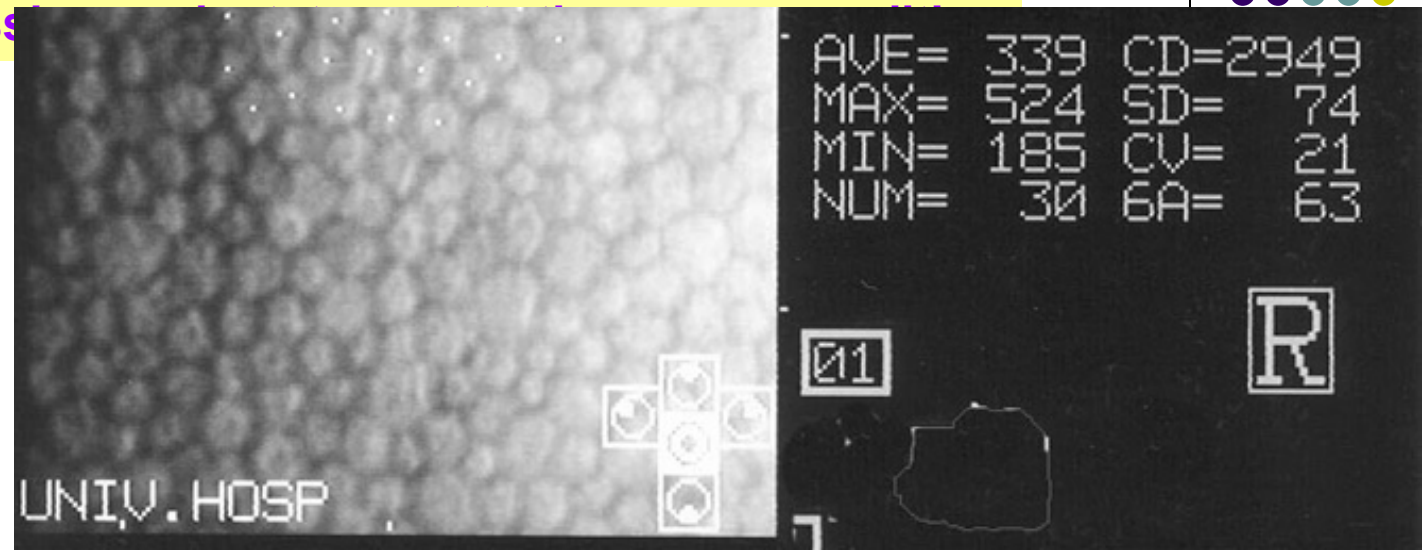


Fuchs endothelial corneal dystrophy. Light microscopy section of FED cornea. Note the markedly thickened Descemet's membrane and the absence of endothelial cell nuclei on the posterior surface (*dashed arrow*).

Is it ICE, PPMD or Fuchs?

Specular microscopic image, **normal cornea**. Note the plethora of polygonal cells of uniform size, and the absence of empty spaces. The cell density is 2949/mm^2 (nl 2-3K, avg ~2400)



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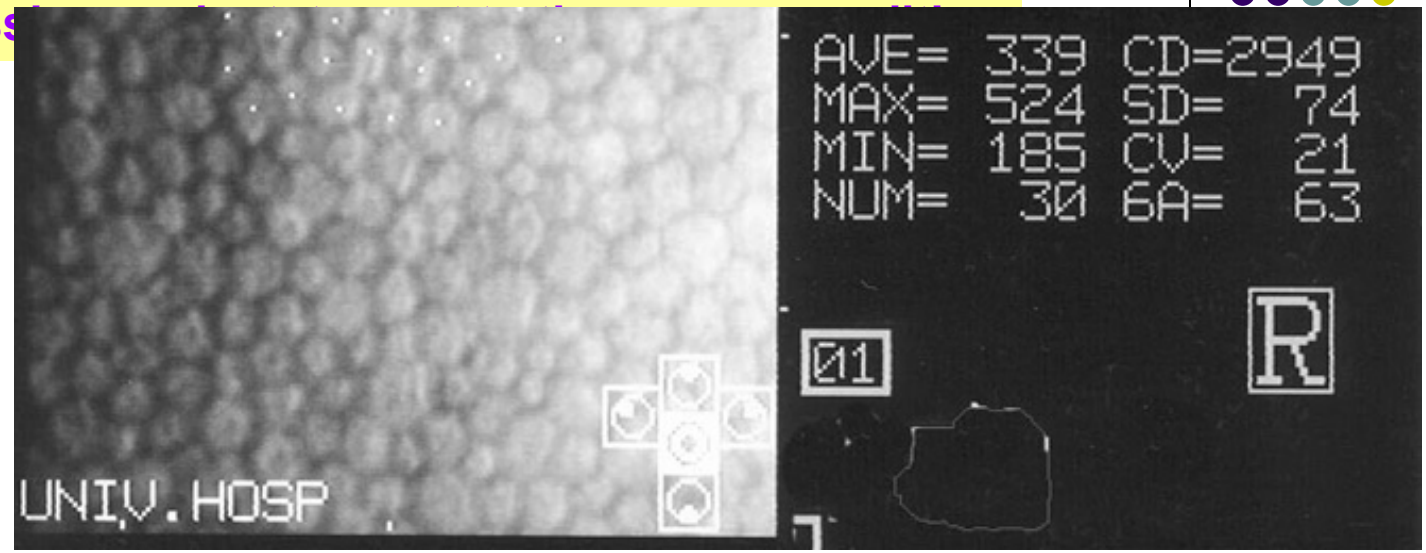
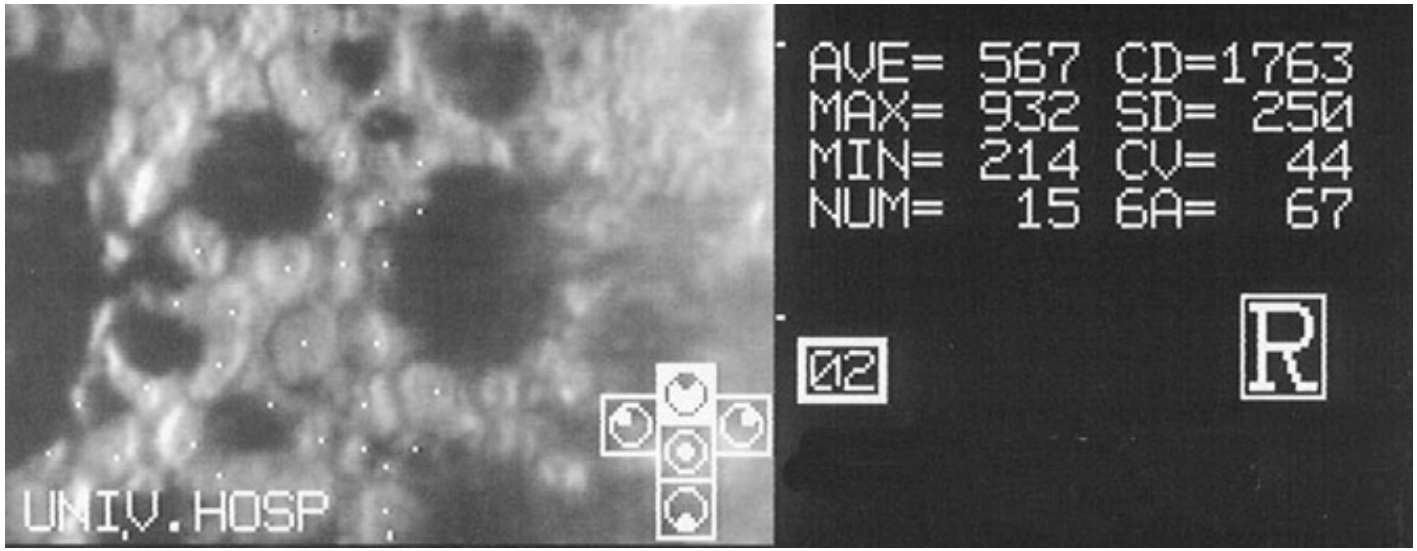
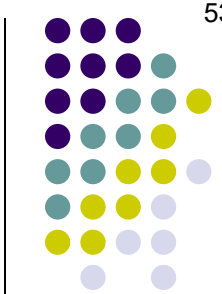


Specular microscopic image, **Fuchs**. Note the polymegathism and polymorphism, and the empty spaces (= guttata). Note that the cell density is only 1763/mm^2.





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How strong is this association?

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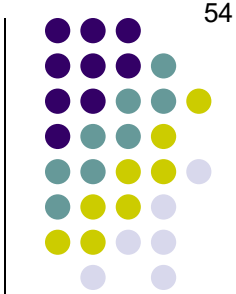

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How strong is this association?
Very—80 to 100% of ICE pts have glaucoma

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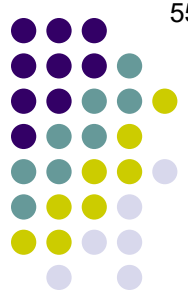

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~25% of PPMD pts have glaucoma

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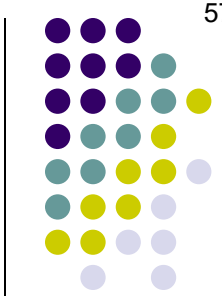

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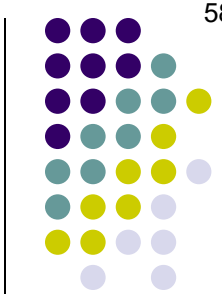
--Corneal edema common

Why the equivocation?



Is it ICE, PPMD or Fuchs?
Assign each statement to the proper condition

ICE
--Essentially always unilateral
--Nonfamilial
--Female > male
--'Hammered silver' appearance
--Strong association with glaucoma
--Corneal edema a defining feature of some forms

PPMD
--Bilateral, but can be so asymmetric as to appear unilateral
--Autosomal dominant
--Female = male
--Bands, vesicles, variable opacities
--Modest association with glaucoma
--Corneal edema can occur, but is not a hallmark

Fuchs
--Essentially always bilateral
--Autosomal dominant
--Female > male
--Guttae
--?No association with glaucoma?
--Corneal edema common

Why the equivocation?
Experts disagree whether an association exists between Fuch's and glaucoma