Is it ICE, PPMD or Fuchs?

ICE =

PPMD =

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

Fuchs =
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

**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

**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
--

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

**Fuchs**
--Essentially always bilateral
--

**Etiology:** Which is…
--Autosomal dominant (2 of them)
--Nonfamilial
Is it ICE, PPMD or Fuchs?
Assign each statement to the proper condition

ICE
--Essentially always unilateral
--Nonfamilial

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

Fuchs
--Essentially always bilateral
--Autosomal dominant

Etiology: Which is…
--Autosomal dominant (2 of them)
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Is it ICE, PPMD or Fuchs?
Assign each statement to the proper condition

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

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

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

**Fuchs**
--Essentially always bilateral  
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Is it ICE, PPMD or Fuchs?
Assign each statement to the proper condition

**ICE**
--Essentially always unilateral
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**Classic endothelial finding**: Which has…
--Guttae
--Bands, vesicles, variable opacities
--’Hammered silver‘ appearance
Is it ICE, PPMD or Fuchs?
Assign each statement to the proper condition

**ICE**
--Essentially always unilateral
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--Guttae

**Classic endothelial finding:** Which has…
--Guttae
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Is it ICE, PPMD or Fuchs?
Assign each statement to the proper condition

ICE: ‘Hammered silver’ corneal endothelium
Is it ICE, PPMD or Fuchs?
Assign each statement to the proper condition

Fuchs: Cornea guttata
Is it ICE, PPMD or Fuchs?

Assign each statement to the proper condition:

1. Vesicular lesions
2. Snail or railroad tracks
3. Endothelial plaque-like lesions
4. 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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--Female > male
--’Hammered silver’ appearance
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--Autosomal dominant
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**Fuchs**
--Essentially always bilateral
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**Glaucoma status:**
--Strong association with glaucoma
--?No association with glaucoma?
--Modest association with glaucoma
Is it ICE, PPMD or Fuchs?
Assign each statement to the proper condition

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--Essentially always unilateral
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--Essentially always bilateral
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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
Is it ICE, PPMD or Fuchs? 
Assign each statement to the proper condition

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--Essentially always unilateral
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(Summary/review slide—no question, proceed when ready)
Is it ICE, PPMD or Fuchs?
Assign each statement to the proper condition

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*What are the different subtypes of ICE?*
Is it ICE, PPMD or Fuchs?
Assign each statement to the proper condition

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What are the different subtypes of ICE?
Two are well established and accepted; they are:
--Chandler syndrome
--Essential (progressive) iris atrophy
The status of the other two are subject to debate; they are:
--Cogan-Reese syndrome
--Iris nevus syndrome
(See the ICE slide set for more details.)
Is it ICE, PPMD or Fuchs?
Assign each statement to the proper condition

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(See the ICE slide-set, K26, for more details.)
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--?No association with glaucoma?
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For the OKAP, I recommend thinking of them as the BCSC does:

**Iris nevus syndrome** (aka Cogan-Reese syndrome)

**Chandler syndrome**

**Essential (progressive) iris atrophy**

The status of the other two are subject to debate; they are:

- Cogan-Reese syndrome
- Iris nevus syndrome

Note that doing this facilitates a very useful mnemonic for remembering the ICE subtypes!
What are the different subtypes of ICE?

Two are well established and accepted; they are:--

Chandler syndrome
Essential (progressive) iris atrophy

The status of the other two are subject to debate; they are:

Cogan-Reese syndrome
Iris nevus syndrome

(See the ICE slide set, K26, for more details.)

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

Chandler syndrome
Essential (progressive) iris atrophy

The status of the other two are subject to debate; they are:

Cogan-Reese syndrome
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(See the ICE slide set, K26, for more details.)
Is it ICE, PPMD or Fuchs?
Assign each statement to the proper condition

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

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What is abnormal about the endothelial cells in PPMD?

Fuchs
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Is it ICE, PPMD or Fuchs?
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--No association with glaucoma?
--Corneal edema common

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

Is it ICE, PPMD or Fuchs?
Assign each statement to the proper condition.
Normal cornea. Note the single-cell-thick nature of the endothelial cells.

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

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

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

--?No association with glaucoma?
--Corneal edema common
Is it ICE, PPMD or Fuchs?
Assign each statement to the proper condition

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Iridocorneal endothelial (ICE) syndrome

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*What condition manifests findings that render it easily confusable with PPMD?*
Iridocorneal endothelial (ICE) syndrome

*How can you tell if a pt has PPMD vs ICE?*

--?No association with glaucoma?
--Corneal edema common
Is it ICE, PPMD or Fuchs?
Assign each statement to the proper condition

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*What condition manifests findings that render it easily confusable with PPMD?*
Iridocorneal endothelial (ICE) syndrome

*How can you tell if a pt has PPMD vs ICE?*
--Unlike PPMD, ICE is always…
--Unlike PPMD, ICE is always…

--?No association with glaucoma?
--Corneal edema common
Is it ICE, PPMD or Fuchs?
Assign each statement to the proper condition

ICE
--Essentially always unilateral
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What condition manifests findings that render it easily confusable with PPMD?
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…

--!?No association with glaucoma?
--Corneal edema common
Is it ICE, PPMD or Fuchs? Assign each statement to the proper condition

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What condition manifests findings that render it easily confusable with PPMD? 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
Is it ICE, PPMD or Fuchs?
Assign each statement to the proper condition

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--Essentially always bilateral
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--Guttae
--No association with glaucoma?
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What are guttae?

Wartlike excrescences on Descemet's membrane

What is the colorful description of the appearance of the endothelium in Fuchs?

'Beaten bronze'

What is the histologic hallmark of Fuchs on confocal microscopy?

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

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What are guttae?
Wartlike excrescences on Descemet’s membrane
Fuchs endothelial corneal dystrophy. Light microscopy: cornea guttata in the form of focal excrescences at the level of the endothelium.
Is it ICE, PPMD or Fuchs?
Assign each statement to the proper condition

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What are guttae?
Wartlike excrescences on Descemet’s membrane

What is the colorful description of the appearance of the endothelium in Fuchs?
‘Beaten bronze’
Fuchs endothelial corneal dystrophy. The appearance wrought by dense guttata has been likened to that of ‘beaten bronze.’
Is it ICE, PPMD or Fuchs?
Assign each statement to the proper condition

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--Appearance? ‘Beaten bronze’

What are guttatae?
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What is the colorful description of the appearance of the endothelium in Fuchs?
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Don’t confuse beaten bronze with hammered silver!
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--?No association with glaucoma?
--Corneal edema common

What are guttae?
Wartlike excrescences on Descemet’s membrane

What is the colorful description of the appearance of the endothelium in Fuchs?
‘Beaten bronze’

What is the histologic hallmark of Fuchs on confocal microscopy?
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)
Normal human cornea. Note numerous endothelial cell nuclei lining the posterior surface (arrow).
Is it ICE, PPMD or Fuchs?

Assign each statement to the proper condition.

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

**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*).
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/mm² (nl 2-3K, avg ~2400)
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/mm² (nl 2-3K, avg ~2400)

Specular microscopic image, **Fuchs**. Note the polymegathism and polymorphism, and the empty spaces (= guttata). Note that the cell density is only 1763/mm².
Is it ICE, PPMD or Fuchs?
Assign each statement to the proper condition

ICE
--Essentially always unilateral
--Nonfamilial
--Female > male
--’Hammered silver’ appearance
[Blue]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

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How strong is this association?
Very—80 to 100% of ICE pts have glaucoma
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How modest is this one?
~25% of PPMD pts have glaucoma
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Why the equivocation?
Is it ICE, PPMD or Fuchs?
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Why the equivocation?
Experts disagree whether an association exists between Fuch’s and glaucoma