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

ICEm

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

PPMD

Fuchs

--
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
--

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
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--Bilateral, but can be so asymmetric as to appear unilateral
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**Fuchs**
--Essentially always bilateral
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**Gender predilection:** Which is…
--Female > male
--Female > male
--Female = male
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
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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

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Glaucoma status: Which has a…
--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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**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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What are the different subtypes of ICE?

(See the ICE slide set for more details.)
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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.)
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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 iris atrophy

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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 iris atrophy

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

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

The status of the other two are subject to debate; they are:
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What is abnormal about the endothelial cells in PPMD?
--Bands, vesicles, variable opacities
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PPMD
--Bilateral, but can be so asymmetric as to appear unilateral
What is abnormal about the endothelial cells in PPMD?
In PPMD, the endothelial cells ‘behave’ like epithelial cells and/or fibroblasts; i.e., they proliferate, form multiple layers, and migrate
--Bands, vesicles, variable opacities
--Modest association with glaucoma
--Corneal edema can occur, but is not a hallmark

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

--?No association with glaucoma?
--Corneal edema common
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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?
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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?
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How can you tell if a pt has PPMD vs ICE?
--Unlike PPMD, ICE is always…unilateral
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--No association with glaucoma?
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*What condition manifests findings that render it easily confusible 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?
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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:
--Decreased cell density, the presence of cells that are much too large (polymegathism) and an increase in cell-to-cell variability in size (pleomorphism)
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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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How strong is this association?
Very—80 to 100% of ICE pts have glaucoma
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--?No association with glaucoma?
--Corneal edema common
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
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How modest?
~25% of PPMD pts have glaucoma
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**Why the ?equivocation?**
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*Why the ?equivocation?*
Experts disagree whether an association exists between Fuch’s and glaucoma