Iridocorneal Endothelial (ICE) Syndrome

ICE: Signs

What are the classic signs of ICE syndrome?
Iridocorneal Endothelial (ICE) Syndrome

**ICE: Signs**

- Iris Changes
- Cornea Changes
- PAS (Peripheral anterior synechiae)
- Unilateral ↑ IOP

What are the classic signs of ICE syndrome?
Iridocorneal Endothelial (ICE) Syndrome

**ICE: Signs**

- Iris Changes
- Cornea Changes
- PAS
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What are the classic signs of ICE syndrome?

Who is the typical patient?
What are the classic signs of ICE syndrome?

Who is the typical patient?
A young-to-middle-aged adult female
Iridocorneal Endothelial (ICE) Syndrome

ICE: Signs

- Iris Changes
- Cornea Changes
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What are the classic signs of ICE syndrome?

Who is the typical patient?
A young-to-middle-aged adult female

How will a pt with ICE present? (On the OKAP, that is.)
-- The pt will be an adult female
--
What are the classic signs of ICE syndrome?

Who is the typical patient?
A young-to-middle-aged adult female

How will a pt with ICE present? (On the OKAP, that is.)
--The pt will be an adult female
--Pt will complain of 1) changes in the eye's appearance, 2) pain, and/or 3) decreased VA
--Pt will have elevated IOP in that eye +/- glaucomatous ONH damage
--The cornea of the affected eye will have abnormal endothelium, and may be edematous
--The fellow eye will be essentially normal, with the possible exception of subtle changes to the corneal endothelium
Iridocorneal Endothelial (ICE) Syndrome

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--The cornea of the affected eye will have two words, and may be
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What ‘pertinent negative’ will be elicited when taking a history?
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What ‘pertinent negative’ will be elicited when taking a history?
She will deny any family history of similar eye findings
What are the subtypes of ICE syndrome?

Iridocorneal Endothelial (ICE) Syndrome

- Chandler syndrome
- Essential iris atrophy
  (Note: Some authors call this variant Progressive iris atrophy, or essential progressive iris atrophy)

What's up with the status Cogan-Reese and iris nevus syndrome?

Here’s the thing. Some sources (my version of the BCSC Cornea book, for example) treat these terms as synonyms, implying they are in fact one and the same condition, or at least closely related. Other sources contend that these are two separate and distinct subtypes of ICE. Still others argue that iris nevus syndrome is a separate disease entirely, ie, not a subtype of ICE at all!
Iridocorneal Endothelial (ICE) Syndrome

What are the subtypes of ICE syndrome? This is a difficult question, and the answer depends in part on who you ask. Let’s start with the easy ones. The identities of two subtypes are not in dispute. These are:

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Chandler Syndrome
Essential Iris Atrophy
Iris Nevus Syndrome
Cogan-Reese Syndrome
Chandler Syndrome
Essential Iris Atrophy

ICE: Subtypes
Iridocorneal Endothelial (ICE) Syndrome

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Iridocorneal Endothelial (ICE) Syndrome

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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*
Iridocorneal Endothelial (ICE) Syndrome

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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!

Iris Nevus Syndrome aka Cogan-Reese Syndrome
Chandler Syndrome
Essential Iris Atrophy

ICE: Subtypes
What is the fundamental pathology (ie, the basic underlying problem) in ICE?

Iridocorneal Endothelial (ICE) Syndrome

ICE: Signs

- Iris Changes
- Cornea Changes
- PAS
- Unilateral ↑ IOP
What is the fundamental pathology (ie, the basic underlying problem) in ICE? A subset of endothelium maldifferentiates, resulting in a cohort of abnormal cells that migrate across the angle and onto the iris, laying down a membrane (histologically similar to Descemet’s) as it goes.
What is the fundamental pathology (ie, the basic underlying problem) in ICE? A subset of endothelium maldifferentiates, resulting in a cohort of abnormal cells that migrate across the angle and onto the iris, laying down a membrane (histologically similar to Descemet’s) as it goes. These abnormal, migrating endothelial cells and associated membrane account for all of the signs found in ICE.
Iridocorneal Endothelial (ICE) Syndrome

ICE: Signs

- Iris Changes
- Cornea Changes
- PAS
- Unilateral ↑ IOP

What are the specific findings?
Iridocorneal Endothelial (ICE) Syndrome

ICE: Signs

Iris Changes
--Atrophy
--Corectopia
--Holes
--Ectropion uveae
--Nodules
--Nevi?

Cornea Changes

PAS

Unilateral ↑IOP

What are the specific findings?
Iridocorneal Endothelial (ICE) Syndrome

ICE: Signs

Iris Changes
- Atrophy
- Corectopia
- Holes
- Ectropion uveae
- Nodules
- Nevi?

Cornea Changes

PAS

Unilateral ↑ IOP
Iridocorneal Endothelial (ICE) Syndrome

ICE: Signs

Iris Changes
- Atrophy
- Corectopia
- Holes
- Ectropion uveae
- Nodules
- Nevi?

Cornea Changes

PAS

Unilateral ↑ IOP

What is corectopia?
Iridocorneal Endothelial (ICE) Syndrome

ICE: Signs

Iris Changes
- Atrophy
- Corectopia
- Holes
- Ectropion uveae
- Nodules
- Nevi?

Cornea Changes

PAS

Unilateral ↑ IOP

What is corectopia?
Displacement of the pupil
Corectopia
Iridocorneal Endothelial (ICE) Syndrome

ICE: Signs

Iris Changes
-- Atrophy
-- Corectopia
-- Holes
-- Ectropion uveae
-- Nodules
-- Nevi?

Cornea Changes

PAS

Unilateral ↑ IOP
Iridocorneal Endothelial (ICE) Syndrome

ICE: Signs

Iris Changes
- Atrophy
- Corectopia
- Holes
- Ectropion uveae
- Nodules
- Nevi?

Cornea Changes

PAS

Unilateral ↑ IOP

What is ectropion uveae?
Iridocorneal Endothelial (ICE) Syndrome

ICE: Signs

- Iris Changes
  - Atrophy
  - Corectopia
  - Holes
  - Ectropion uveae
  - Nodules
  - Nevi?
- Cornea Changes
- PAS
- Unilateral ↑ IOP

What is ectropion uveae?
The presence of posterior pigmented epithelium at the pupil margin and adjacent anterior iris surface
Ectropion uveae
Iridocorneal Endothelial (ICE) Syndrome

**ICE: Signs**

- Iris Changes
  - Atrophy
  - Corectopia
  - Holes
  - Ectropion uveae
  - Nodules
  - Nevi?

- Cornea Changes

- PAS

- Unilateral ↑ IOP

**What is ectropion uveae?**
The presence of posterior pigmented epithelium at the pupil margin and adjacent anterior iris surface

**How does it get there?**
Iridocorneal Endothelial (ICE) Syndrome

ICE: Signs

- Iris Changes
  - Atrophy
  - Corectopia
  - Holes
  - Ectropion uveae
  - Nodules
  - Nevi?

- Cornea Changes

- PAS

- Unilateral ↑ IOP

---

**What is ectropion uveae?**
The presence of posterior pigmented epithelium at the pupil margin and adjacent anterior iris surface

**How does it get there?**
It is pulled around the margin of the pupil by the contracting membrane on the surface of the iris
Iridocorneal Endothelial (ICE) Syndrome

ICE: Signs

Iris Changes
--Atrophy
--Corectopia
--Holes
--Ectropion uveae
--Nodules
--Nevi?

Cornea Changes

PAS

Unilateral ↑ IOP

What one-word description is applied to the nodules? 'Pedunculated'
Iridocorneal Endothelial (ICE) Syndrome

ICE: Signs

Iris Changes
- Atrophy
- Corectopia
- Holes
- Ectropion uveae
- Nodules
- Nevi?

Cornea Changes

PAS

Unilateral ↑ IOP

What one-word description is applied to the nodules? ‘Pedunculated’
Iris nodules
Iridocorneal Endothelial (ICE) Syndrome

**ICE: Signs**

- Iris Changes
  - Atrophy
  - Corectopia
  - Holes
  - Ectropion uveae
  - Nodules
  - Nevi?

- Cornea Changes

- PAS

- Unilateral ↑IOP

*Are the ‘nevi’ actually nevi (i.e., collections of melanocytes)?
Iridocorneal Endothelial (ICE) Syndrome

ICE: Signs

- Iris Changes
  - Atrophy
  - Corectopia
  - Holes
  - Ectropion uveae
  - Nodules
  - Nevi?

- Cornea Changes

- PAS

- Unilateral ↑ IOP

Are the ‘nevi’ actually nevi (ie, collections of melanocytes)?
No, they are pseudo-nevi--comprised in this case of focal areas of compressed iris stroma
Iris nevi
Iridocorneal Endothelial (ICE) Syndrome

ICE: Signs

- Iris Changes
  - Atrophy
  - Corectopia
  - Holes
  - Ectropion uveae
  - Nodules
  - Nevi?

- Cornea Changes
- PAS
- Unilateral ↑ IOP

Are the ‘nevi’ actually nevi (ie, collections of melanocytes)?
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What is the etiology of the iris-stroma compression?
Iridocorneal Endothelial (ICE) Syndrome

ICE: Signs

Iris Changes
- Atrophy
- Corectopia
- Holes
- Ectropion uveae
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No, they are pseudo-nevi--comprised in this case of focal areas of compressed iris stroma

What is the etiology of the iris-stroma compression?
Contraction of the ICE membrane
Iridocorneal Endothelial (ICE) Syndrome

ICE: Signs

Iris Changes
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-- Holes
-- Ectropion uveae
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-- Nevi?

Cornea Changes

PAS

Unilateral ↑ IOP

What are the specific findings?
Iridocorneal Endothelial (ICE) Syndrome

ICE: Signs

Iris Changes
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Cornea Changes
-- Abnormal endothelium
-- Edema

PAS

Unilateral ↑ IOP

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Cornea Changes
--Abnormal endothelium
--Edema

PAS

Unilateral ↑ IOP

What is the classic two-word description of the endothelium’s slit-lamp appearance in ICE?
Iridocorneal Endothelial (ICE) Syndrome

ICE: Signs

Iris Changes
-- Atrophy
-- Corectopia
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-- Ectropion uveae
-- Nodules
-- Nevi?

Cornea Changes
-- Abnormal endothelium
-- Edema

PAS

Unilateral ↑ IOP

What is the classic two-word description of the endothelium’s slit-lamp appearance in ICE?
‘Hammered silver’
‘Hammered silver’ corneal endothelium
Iridocorneal Endothelial (ICE) Syndrome

**ICE: Signs**

- **Iris Changes**
  - Atrophy
  - Corectopia
  - Holes
  - Ectropion uveae
  - Nodules
  - Nevi?

- **Cornea Changes**
  - Abnormal endothelium
  - Edema

- **PAS**

- **Unilateral ↑ IOP**

What is the classic two-word description of the endothelium’s slit-lamp appearance in ICE?

‘Hammered silver’

By way of comparison, what’s the classic two-word description of the endothelium’s slit-lamp appearance in Fuch’s endothelial dystrophy?
Iridocorneal Endothelial (ICE) Syndrome

ICE: Signs

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- Abnormal endothelium
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‘Hammered silver’

By way of comparison, what’s the classic two-word description of the endothelium’s slit-lamp appearance in Fuch’s endothelial dystrophy?
‘Beaten bronze’
Study guide: Iridocorneal Endothelial (ICE) Syndrome

ICE: Signs

- Iris Changes
  - Atrophy
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  - Holes
  - Ectropion uveae
  - Nodules
  - Nevi?

- Cornea Changes
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Edema
Iridocorneal Endothelial (ICE) Syndrome

ICE: Signs

Iris Changes
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--Nevi?

Cornea Changes
--Abnormal endothelium
--Edema

PAS

Unilateral ↑ IOP

Two words are commonly used to describe the appearance of the PAS in ICE. What are they?
Iridocorneal Endothelial (ICE) Syndrome

ICE: Signs

- Iris Changes
  - Atrophy
  - Corectopia
  - Holes
  - Ectropion uveae
  - Nodules
  - Nevi?

- Cornea Changes
  - Abnormal endothelium
  - Edema

- PAS
  - Two words are commonly used to describe the appearance of the PAS in ICE. What are they? ‘Broad’ and ‘high’

- Unilateral ↑ IOP
Iridocorneal Endothelial (ICE) Syndrome

ICE: Signs

**Iris Changes**
- Atrophy
- Corectopia
- Holes
- Ectropion uveae
- Nodules
- Nevi?

**Cornea Changes**
- Abnormal endothelium
- Edema

**PAS**

**Unilateral ↑ IOP**

Two words are commonly used to describe the appearance of the PAS in ICE. What are they? ‘Broad’ and ‘high’

What does high mean in this context?
Iridocorneal Endothelial (ICE) Syndrome

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  -- Atrophy
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  -- Holes
  -- Ectropion uveae
  -- Nodules
  -- Neví?

- **Cornea Changes**
  -- Abnormal endothelium
  -- Edema

- **PAS**

- **Unilateral ↑ IOP**

Two words are commonly used to describe the appearance of the PAS in ICE. What are they? ‘Broad’ and ‘high’.

What does high mean in this context? That the PAS extend beyond Schwalbe’s line (SL).
Iridocorneal Endothelial (ICE) Syndrome

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--Corectopia
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Cornea Changes
--Abnormal endothelium
--Edema

PAS
Two words are commonly used to describe the appearance of the PAS in ICE. What are they? ‘Broad’ and ‘high’

Unilateral ↑IOP

What does high mean in this context?
That the PAS extend beyond Schwalbe’s line (SL)

Why is this considered ‘high’?
Iridocorneal Endothelial (ICE) Syndrome

ICE: Signs

- Iris Changes
  - Atrophy
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  - Nodules
  - Nevi?

- Cornea Changes
  - Abnormal endothelium
  - Edema

- PAS
  - Abnormal endothelium
  - Edema

- Unilateral ↑ IOP

Two words are commonly used to describe the appearance of the PAS in ICE. What are they? ‘Broad’ and ‘high’

What does high mean in this context? That the PAS extend beyond Schwalbe’s line (SL)

Why is this considered ‘high’? Because PAS secondary to NVI/NVA end at SL
Iridocorneal Endothelial (ICE) Syndrome

ICE: Signs

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Cornea Changes
-- Abnormal endothelium
-- Edema

PAS

Unilateral ↑ IOP

Two words are commonly used to describe the appearance of the PAS in ICE. What are they? ‘Broad’ and ‘high’

What does high mean in this context?
That the PAS extend beyond Schwalbe’s line (SL)

Why is this considered ‘high’?
Because PAS secondary to NVI/NVA end at SL

Why does neo-related PAS end at SL?
Iridocorneal Endothelial (ICE) Syndrome

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Cornea Changes
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--Edema

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Unilateral ↑IOP

Two words are commonly used to describe the appearance of the PAS in ICE. What are they? ‘Broad’ and ‘high’

What does high mean in this context?
That the PAS extend beyond Schwalbe’s line (SL)

Why is this considered ‘high’?
Because PAS secondary to NVI/NVA end at SL

Why does neo-related PAS end at SL?
Because neo can’t grow over ‘normal’ endothelium
Iridocorneal Endothelial (ICE) Syndrome

ICE: Signs

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- Nevi?

Cornea Changes
- Abnormal endothelium
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PAS

Unilateral ↑ IOP

How common is ↑ IOP in ICE?
Iridocorneal Endothelial (ICE) Syndrome

ICE: Signs

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--Nodules
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Cornea Changes
--Abnormal endothelium
--Edema

PAS

Unilateral ↑ IOP

How common is ↑ IOP in ICE? Very—estimates range from 80-100% of cases
Iridocorneal Endothelial (ICE) Syndrome

ICE: Signs

- Iris Changes
  - Atrophy
  - Corectopia
  - Holes
  - Ectropion uveae
  - Nodules
  - Nevi?

- Cornea Changes
  - Abnormal endothelium
  - Edema

- PAS

- Unilateral ↑ IOP

- Iris Nevus Syndrome

- Cogan-Reese Syndrome

- Chandler Syndrome

- Essential Iris Atrophy

Now let’s look at how the ICE subtypes manifest the various ICE signs…

ICE: Subtypes
Iridocorneal Endothelial (ICE) Syndrome

ICE: Signs

Iris Changes
-- Atrophy
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-- Holes
-- Ectropion uveae
-- Nodules
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Cornea Changes
-- Abnormal endothelium
-- Edema

PAS

Unilateral ↑ IOP

All subtypes demonstrate, to varying degrees, three findings:

ICE: Subtypes

Iris Nevus Syndrome

Cogan-Reese Syndrome

Chandler Syndrome

Essential Iris Atrophy

ICE: Subtypes
Iridocorneal Endothelial (ICE) Syndrome

ICE: Signs

Iris Changes
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Cornea Changes
-- Abnormal endothelium
-- Edema

PAS
-- Abnormal endothelium
-- PAS
-- Unilateral ↑ IOP

Unilateral ↑ IOP

Iris Nevus Syndrome
-- Abn. endothelium
-- PAS
-- Unilateral ↑ IOP

Cogan-Reese Syndrome
-- Abn. endothelium
-- PAS
-- Unilateral ↑ IOP

Chandler Syndrome
-- Abn. endothelium
-- PAS
-- Unilateral ↑ IOP

Essential Iris Atrophy
-- Abn. endothelium
-- PAS
-- Unilateral ↑ IOP

All subtypes demonstrate, to varying degrees, **three findings:**

ICE: Subtypes
Iridocorneal Endothelial (ICE) Syndrome

ICE: Signs

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--Nodules
--Nevi?

Corneal Changes
--Abnormal endothelium
--Edema

PAS
--Abnormal endothelium
--PAS

Unilateral ↑ IOP

Iris Nevus Syndrome
--Abn. endothelium
--PAS
--Unilateral ↑ IOP

Cogan-Reese Syndrome
--Abn. endothelium
--PAS
--Unilateral ↑ IOP

Chandler Syndrome
--Abn. endothelium
--PAS
--Unilateral ↑ IOP

Essential Iris Atrophy
--Abn. endothelium
--PAS
--Unilateral ↑ IOP

One subtype tends to have more modest IOP elevation than the others—which one?
Iridocorneal Endothelial (ICE) Syndrome

ICE: Signs

Iris Changes
--Atrophy
--Corectopia
--Holes
--Ectropion uveae
--Nodules
--Nevi?

Cornea Changes
--Abnormal endothelium
--Edema

PAS
--Abnormal endothelium
--PAS
--Unilateral ↑ IOP

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--Abn. endothelium
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Essential Iris Atrophy
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One subtype tends to have more modest IOP elevation than the others—which one? Chandler syndrome
Iridocorneal Endothelial (ICE) Syndrome

**ICE: Signs**

**Iris Changes**
- Atrophy
- Corectopia
- Holes
- Ectropion uveae
- Nodules
- Nev? (question mark)

**Cornea Changes**
- Abnormal endothelium
- Edema

**PAS**

**Unilateral ↑ IOP**

However, each has a **key finding** that ‘makes’ the subtype:

**Iris Nevus Syndrome**

**Cogan-Reese Syndrome**

**Chandler Syndrome**

**Essential Iris Atrophy**

**ICE: Subtypes**
Iridocorneal Endothelial (ICE) Syndrome

ICE: Signs

Iris Changes
--Atrophy
--Corectopia
--Holes
--Ectropion uveae
--Nodules
--Nevi?

Cornea Changes
--Abnormal endothelium
--Edema

PAS

Unilateral ↑IOP

However, each has a key finding that ‘makes’ the subtype:

Iris Nevus Syndrome
--Iris nevi (duh)

Cogan-Reese Syndrome
--Iris nodules

Chandler Syndrome
--Corneal edema predominates

Essential Iris Atrophy
--Iris atrophy/holes predominate

ICE: Subtypes
Iris nevus/Cogan-Reese syndrome

Chandler syndrome

Essential iris atrophy
Iridocorneal Endothelial (ICE) Syndrome

ICE: Signs

- Iris Changes
  - Atrophy
  - Corectopia
  - Holes
  - Ectropion uveae
  - Nodules
  - Nevi?

- Cornea Changes
  - Abnormal endothelium

- PAS
  - Edema

- Unilateral ↑IOP

Of the four categories of signs (ie, iris changes, cornea changes, PAS and elevated IOP), which one is most important in differentiating among the ICE subtypes?

ICE: Subtypes

- Iris Nevus Syndrome
  - Iris nevi (duh)

- Cogan-Reese Syndrome
  - Iris nodules

- Chandler Syndrome
  - Corneal edema predominates

- Essential Iris Atrophy
  - Iris atrophy/holes predominate
Iridocorneal Endothelial (ICE) Syndrome

**ICE: Signs**

**Iris Changes**
- Atrophy
- Nevi?

**Cornea Changes**
- Abnormal endothelium

**PAS**

**Unilateral ↑ IOP**

Of the four categories of signs (ie, iris changes, cornea changes, PAS and elevated IOP), which one is most important in differentiating among the ICE subtypes? The clinical status of the iris is most important.

(Iris Nevus Syndrome)
- *Iris nevi* (duh)

(Cogan-Reese Syndrome)
- *Iris nodules*

(Chandler Syndrome)
- Corneal edema predominates
- Relatively nl iris

(Essential Iris Atrophy)
- Iris atrophy/holes predominate

(Note that one of the hallmarks of Chandler syndrome is the relative absence of iris changes, which makes iris status important in its diagnosis too.)
Iridocorneal Endothelial (ICE) Syndrome

What are the main management issues in ICE?
1) IOP/glaucoma control
2) Minimizing corneal edema
Iridocorneal Endothelial (ICE) Syndrome

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- Abn. endothelium
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What are the main management issues in ICE?
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2) Minimizing corneal edema

Which meds are most effective?
- Aqueous suppressants, which are:
  - β blockers
  - Carbonic anhydrase inhibitors
  - α agonists

If medical management is inadequate, should filtering surgery be attempted? Yes

ICE: Subtypes

- Iris Nevus Syndrome
  - Abn. endothelium
  - PAS
  - Unilateral ↑ IOP
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- Cogan-Reese Syndrome
  - Abn. endothelium
  - PAS
  - Unilateral ↑ IOP
  - Iris nodules

- Chandler Syndrome
  - Abn. endothelium
  - PAS
  - Unilateral ↑ IOP
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  - Abnormal endothelium
  - PAS
  - Unilateral ↑ IOP
  - Iris nodules

- Chandler Syndrome
  - Abnormal endothelium
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ICE: Subtypes
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Yes

Which meds are most effective?
Hypertonic saline solutions

ICE: Subtypes

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- Cogan-Reese Syndrome
  - --Iris nodules
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Which meds are most effective?
Hypertonic saline solutions

If medical management is inadequate, should PK be attempted?

Iris Nevus Syndrome
--Iris nevi (duh)

Cogan-Reese Syndrome
--Iris nodules

Chandler Syndrome
--Corneal edema predominates
--Relatively nl iris

Essential Iris Atrophy
--Iris atrophy/holes predominate

Ice: Subtypes
What are the main management issues in ICE?
1) IOP/glaucoma control
2) Minimizing corneal edema

Which meds are most effective?
- β blockers
- Carbonic anhydrase inhibitors
- α agonists

If medical management is inadequate, should filtering surgery be attempted?
Yes

Which meds are most effective?
Hypertonic saline solutions

If medical management is inadequate, should PK be attempted?
Yes

ICE: Subtypes

**Iris Nevus Syndrome**
- Iris nevi (duh)

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- Iris nodules

**Chandler Syndrome**
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