Iridocorneal Endothelial (ICE) Syndrome

ICE: Signs

- Changes in eye's appearance
- FBS-type pain
- Elevated IOP +/− glaucomatous damage to the ONH
- Abnormal endothelium, may be edematous
- Essentially normal fellow eye, with possible subtle changes to the corneal endothelium

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
- Unilateral \( \uparrow \) IOP

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
What are the classic signs of ICE syndrome?

Who is the typical patient?
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How will a pt with ICE present? (On the OKAP, that is.)
--The pt will be an adult female
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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**
--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**

**What 'pertinent negative' will be elicited when taking a history?**
She will deny any familial history of similar eye findings
Iridocorneal Endothelial (ICE) Syndrome

**ICE: Signs**

- Iris Changes
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Iridocorneal Endothelial (ICE) Syndrome

What are the subtypes of ICE syndrome?

ICE: Subtypes

- Chandler syndrome
- Essential iris atrophy
- Progression iris atrophy
- 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:

- Chandler syndrome
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**ICE: Subtypes**

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Iris Nevus Syndrome
Cogan-Reese Syndrome
Chandler Syndrome
Essential Iris Atrophy
(aka progressive iris atrophy, aka essential progressive iris atrophy)

ICE: Subtypes
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

Still others argue that iris nevus syndrome is a separate disease entirely, ie, not a subtype of ICE at all!

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

ICE: Subtypes
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- 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!
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?
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

*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
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
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

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- Holes
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- Nodules
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Cornea Changes

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Unilateral ↑ IOP

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- Cornea Changes

- PAS

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Are the ‘nevi’ actually nevi (ie, 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
Iridocorneal Endothelial (ICE) Syndrome

**ICE: Signs**

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

- Cornea Changes

- PAS

- Unilateral ↑IOP

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*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

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

ICE: Signs

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

Cornea Changes

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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

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

ICE: Signs

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

Cornea Changes
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PAS
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Unilateral ↑ IOP

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

ICE: Signs

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

Cornea Changes
--Abnormal endothelium
--Edema

PAS

Unilateral ↑IOP

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Cornea Changes
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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
--Holes
--Ectropion uveae
--Nodules
--Nevi?

Cornea Changes

PAS
--Abnormal endothelium
--Edema

Unilateral ↑IOP

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

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Iris Changes
--Atrophy
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--Nodules
--Nevi?

Cornea Changes
--Abnormal endothelium
--Edema

PAS

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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’
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?
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’
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?

Because neo can’t grow over ‘normal’ endothelium
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**

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

ICE: Signs

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

Cornea Changes:
- Abnormal endothelium
- Edema

PAS:
- Abnormal endothelium
- Edema

Unilateral \(\uparrow\) 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'?
Iridocorneal Endothelial (ICE) Syndrome

ICE: Signs

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

- Cornea Changes
  - Abnormal endothelium
  - Edema

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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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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?
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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
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- Nodules
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- Abnormal endothelium
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**Unilateral ↑ IOP**

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

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Cornea Changes
- Abnormal endothelium
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PAS

Unilateral ↑ IOP

How common is ↑ IOP in ICE? Very—estimates range from 80-100% of cases
Now let’s look at how the ICE subtypes manifest the various ICE signs…
Iridocorneal Endothelial (ICE) Syndrome

ICE: Signs

Iris Changes
-- Atrophy
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Cornea Changes
-- Abnormal endothelium
-- Edema

PAS

Unilateral ↑ IOP

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

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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--Nevi?

Cornea Changes

PAS
--Abnormal endothelium
--Edema

Unilateral ↑ IOP

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

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

Chandler Syndrome
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--PAS
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--PAS
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ICE: Subtypes
Iridocorneal Endothelial (ICE) Syndrome

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--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
  -- Nevus?

- Cornea Changes
  -- Abnormal endothelium
  -- Edema

- 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
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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
  - Nevi?

- Cornea Changes
  - Abnormal endothelium
  - Edema

- PAS

- Unilateral ↑ IOP

ICE: Subtypes

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

However, each has a **key finding** that ‘makes’ the subtype:
Iridocorneal Endothelial (ICE) Syndrome

ICE: Signs

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

Iris Nevus Syndrome
- Iris nevi (duh)

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ICE: Subtypes
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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)
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Iridocorneal Endothelial (ICE) Syndrome

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.

(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)
What are the main management issues in ICE?

1) IOP/glaucoma control
2) Minimizing corneal edema

ICE: Subtypes

Iris Nevus Syndrome
--Iris nevi (duh)

Cogan-Reese Syndrome
--Iris nodules

Chandler Syndrome
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--Relatively nl iris

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What are the main management issues in ICE?
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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

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Yes

**Which meds are most effective?**

Hypertonic saline solutions

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Which meds are most effective?
Hypertonic saline solutions

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

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