What are the classic signs of ICE syndrome?

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

- The pt will be an adult female
- She will complain of 1) changes in her eye’s appearance, 2) FBS-type pain, or both
- She will have elevated IOP in that eye +/- glaucomatous damage to the ONH
- 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.
What are the classic signs of ICE syndrome?

Who is the typical patient?

-Iris Changes
-Cornea Changes
-PAS
-Unilateral IOP
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?
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?

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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
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What are the subtypes of ICE syndrome?

ICE: Subtypes
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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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
-- Essential iris atrophy (Note: Some authors call this variant progressive iris atrophy, or essential progressive iris atrophy)

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

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What's up with the status of Cogan-Reese and iris nevus syndrome?
Here’s the thing: The BCSC books treat these terms as synonyms. 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, i.e., not a subtype of ICE at all.
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(Note: Some authors call this variant *progressive iris atrophy* or *essential progressive 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*

![Diagram of ICE subtypes](image)
What are the subtypes of ICE syndrome?
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Note that this creates a very apropos mnemonic for remembering the ICE subtypes!
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Which subtype is most common?

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

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- Chandler syndrome
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Which subtype is most common?
Chandler syndrome—it accounts for about ___% of all cases.
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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For the OKAP, I recommend thinking of them as the BCSC does:

- **Iris nevus syndrome** (aka Cogan-Reese syndrome)
- **Chandler syndrome**
- **Essential iris atrophy**

**Which subtype is most common?**

Chandler syndrome—it accounts for about half of all cases.

---

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

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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.
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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 \( \uparrow \) IOP

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

ICE: Signs

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

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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

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
Iridocorneal Endothelial (ICE) Syndrome

ICE: Ectropion uveae
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’
Iridocorneal Endothelial (ICE) Syndrome

ICE: Iris nodules (note also the ectropion uveae)
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)?
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
ICE: Iris nevi (and ectropion uveae)
**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)?*

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
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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?
Contraction of the ICE membrane
Iridocorneal Endothelial (ICE) Syndrome

ICE: Signs

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- Holes
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- 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
  - Abnormal endothelium
  - Edema

- PAS

- Unilateral ↑ IOP

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

Hammered silver

Another two-word description of ICE endothelium’s is beaten bronze.

In what other condition is this term use to describe the endothelium?

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

ICE: ‘Hammered silver’ corneal endothelium
Iridocorneal Endothelial (ICE) Syndrome

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

Cornea Changes
--Abnormal endothelium
--Edema

PAS

Unilateral ↑IOP

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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

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: PAS
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?
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

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?
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

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
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ICE: Signs

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--Atrophy
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--Ectropion uveae
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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

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? 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

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

Cornea Changes
- Abnormal endothelium
- Edema

PAS

Unilateral IOP

How common is glaucoma in ICE?
Iridocorneal Endothelial (ICE) Syndrome

ICE: Signs

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

Cornea Changes
-- Abnormal endothelium
-- Edema

PAS

Unilateral IOP

How common is glaucoma in ICE?
Quite—about 50% 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
- Corectopia
- Holes
- Ectropion uveae
- Nodules
- Nevus?

Cornea Changes
- Abnormal endothelium
- Edema

PAS

Unilateral ↑ IOP

All subtypes demonstrate, to varying degrees, three findings:

ICE: Subtypes

Iris Nevus Syndrome
aka

Cogan-Reese Syndrome

Chandler Syndrome

Essential Iris Atrophy
Iridocorneal Endothelial (ICE) Syndrome

ICE: Signs

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

Cornea Changes
--Abnormal endothelium
--Edema

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

Unilateral ↑ IOP

All subtypes demonstrate, to varying degrees, three findings:

Iris Nevus Syndrome aka

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

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

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

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**

**Iris Nevus Syndrome**
- Abnormal endothelium
- PAS
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**Cogan-Reese Syndrome**
- Abnormal endothelium
- PAS
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**Chandler Syndrome**
- Abnormal endothelium
- PAS
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**Essential Iris Atrophy**
- Abnormal endothelium
- PAS
- Unilateral ↑ IOP

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

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

- **Iris Nevus Syndrome**
  - aka Cogan-Reese Syndrome

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**ICE: Subtypes**
Chandler Syndrome

Essential Iris Atrophy

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Iris Changes

Cornea Changes

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ICE: Signs

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

**ICE: Subtypes**

- Iris Nevus Syndrome
  - Iris nevi /nodules

- Cogan-Reese Syndrome
  - Iris nevi /nodules

- Chandler Syndrome
  - Corneal edema predominates

- Essential Iris Atrophy
  - Iris atrophy/holes predominate

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Iridocorneal Endothelial (ICE) Syndrome

Iris nevus/Cogan-Reese syndrome

Chandler syndrome

Essential iris atrophy
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?
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)
Iridocorneal Endothelial (ICE) Syndrome

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

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- Aqueous suppressants, which are:
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  - α agonists

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