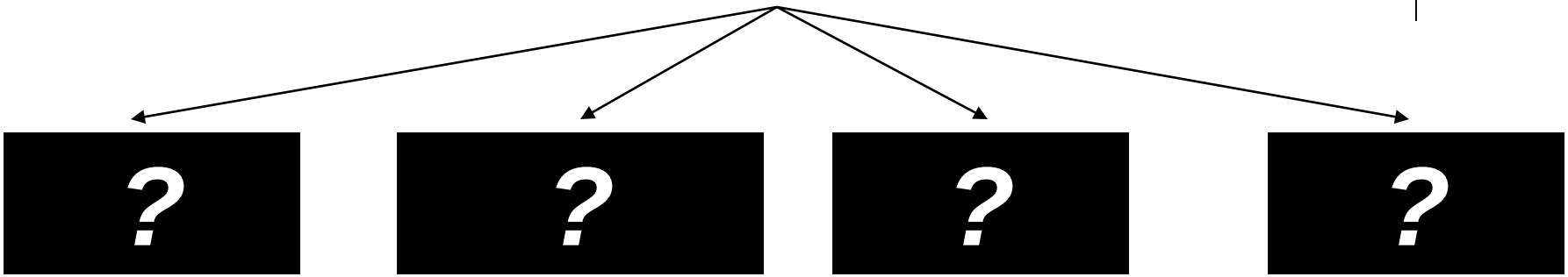
# Iridocorneal Endothelial (ICE) Syndrome

## 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

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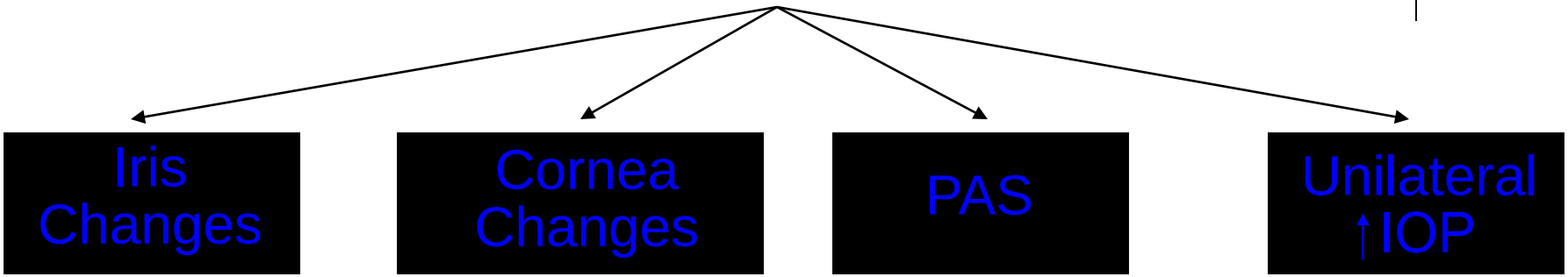


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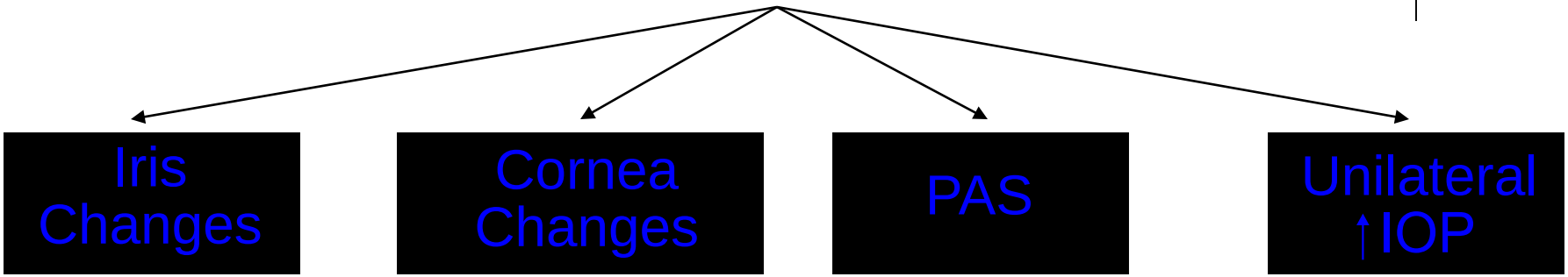


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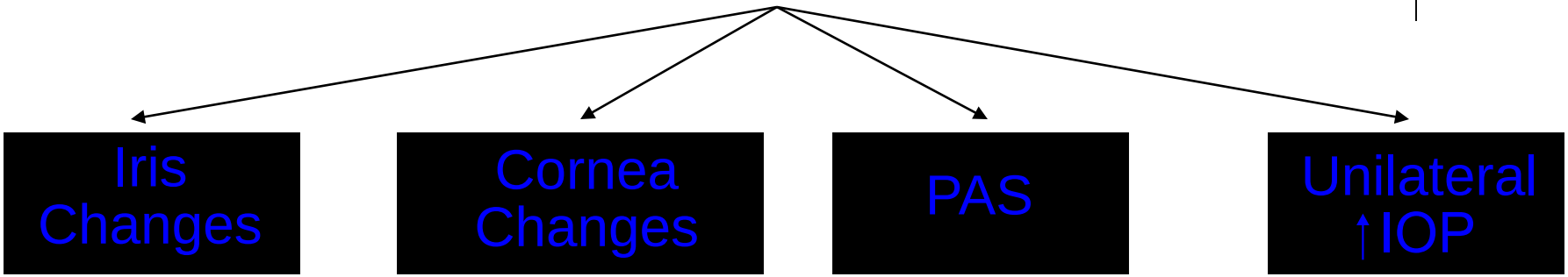


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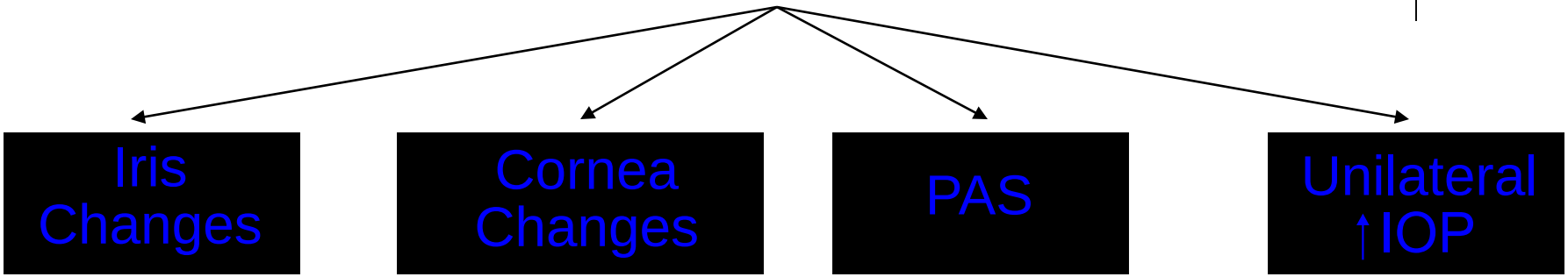


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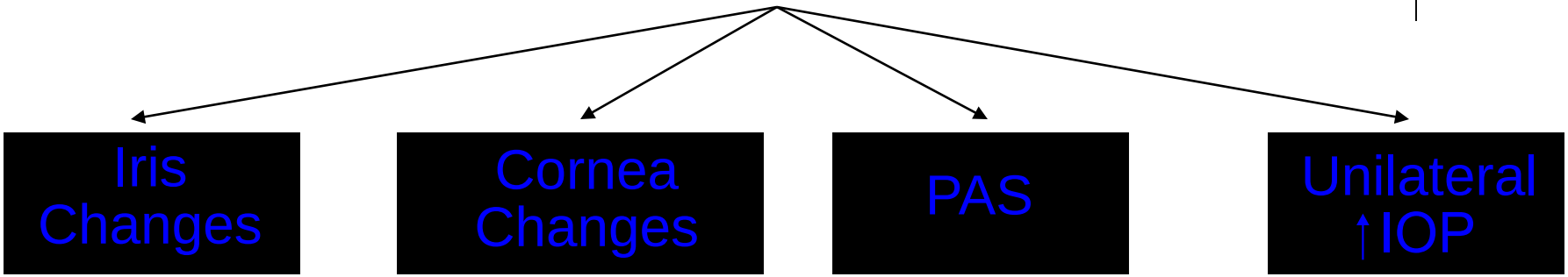


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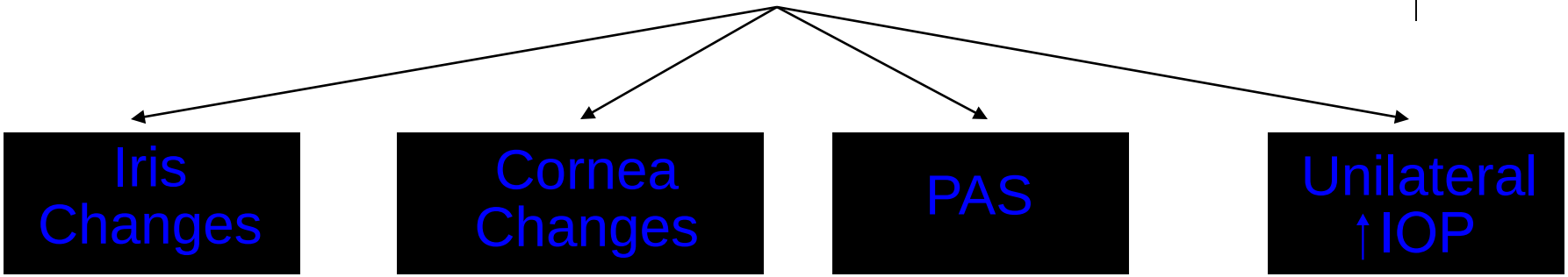


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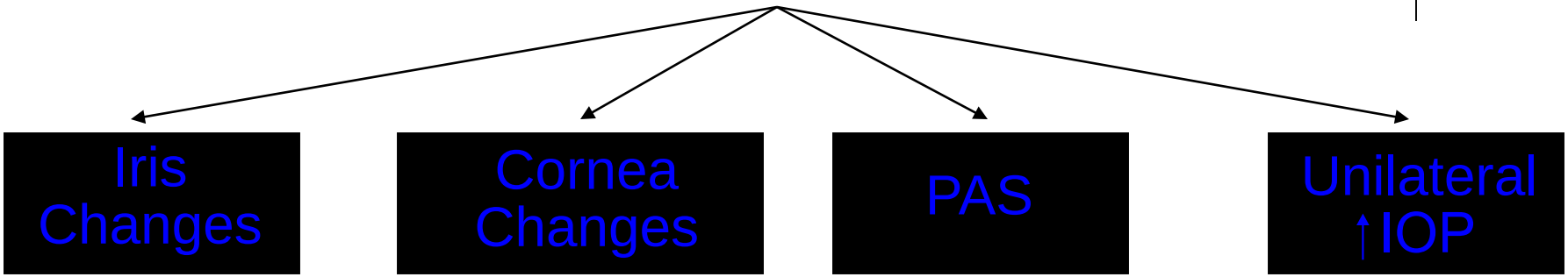


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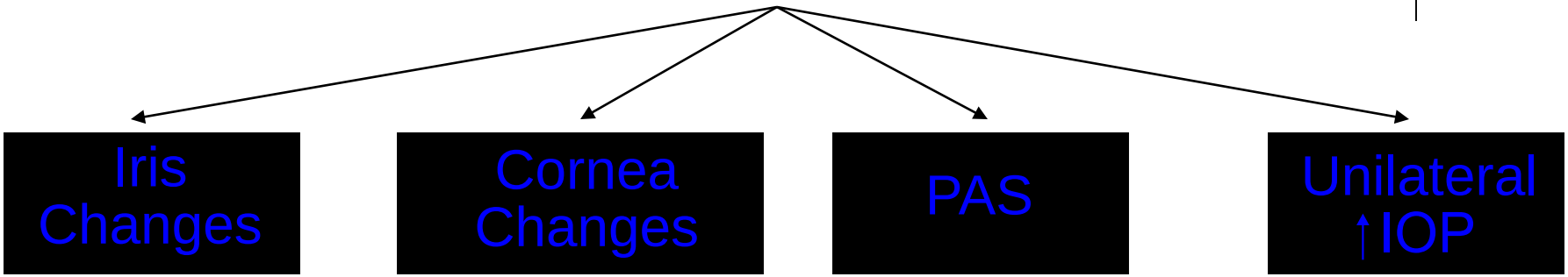


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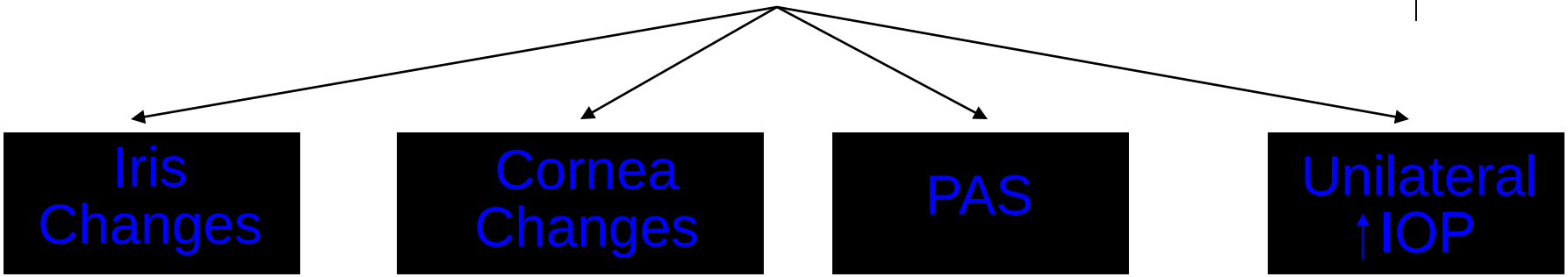


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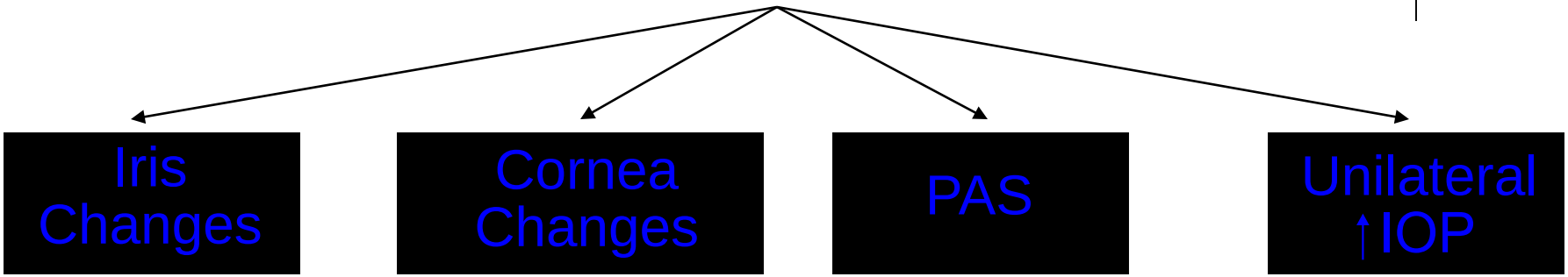


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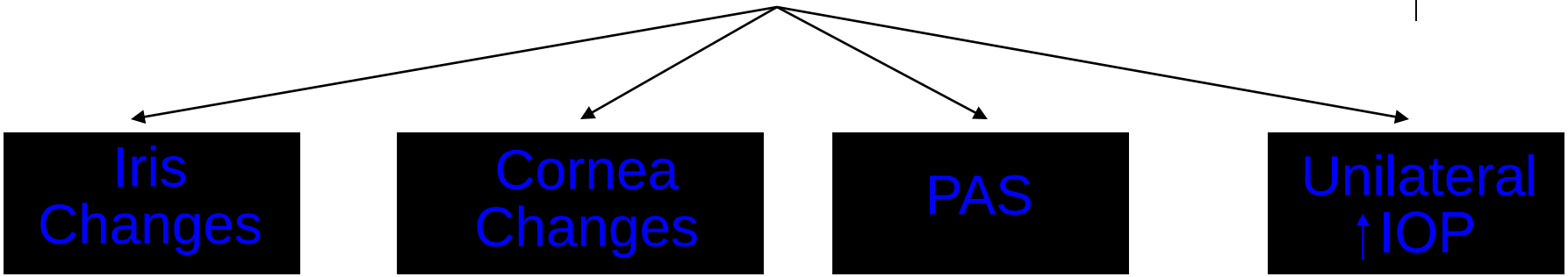


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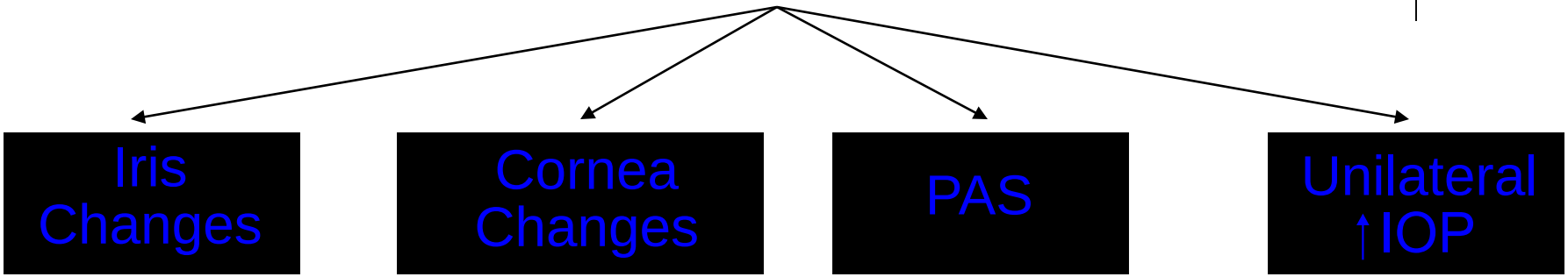


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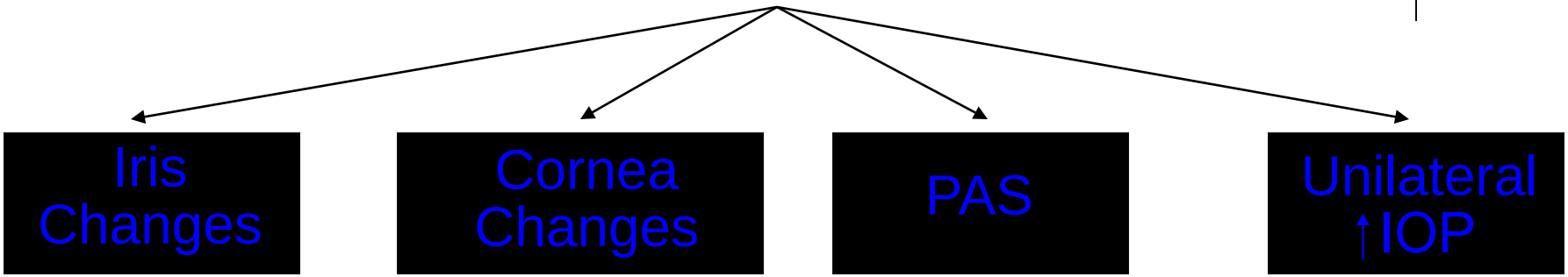


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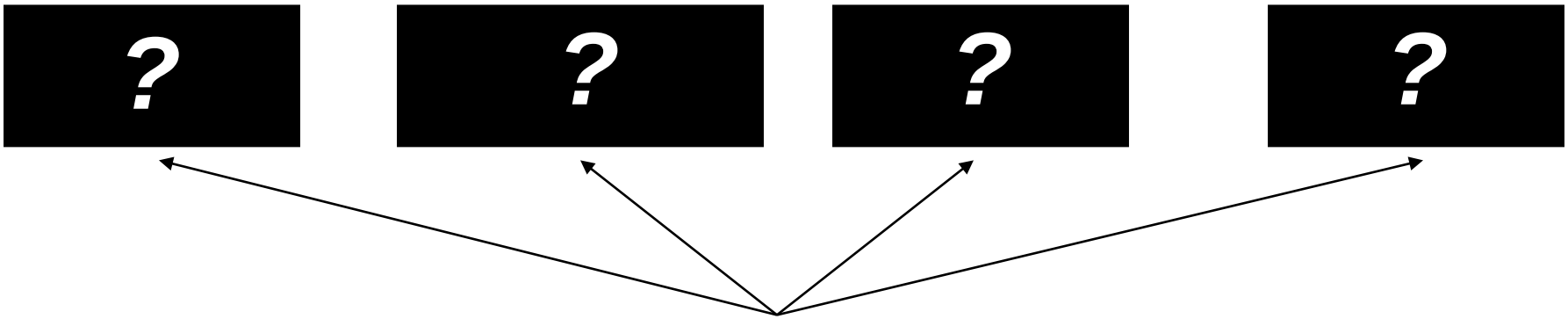
*What 'pertinent negative' will be elicited when taking a history?*
She will deny any family history of similar eye findings

# Iridocorneal Endothelial (ICE) Syndrome

*What are the subtypes of ICE syndrome?*




ICE: *Subtypes*

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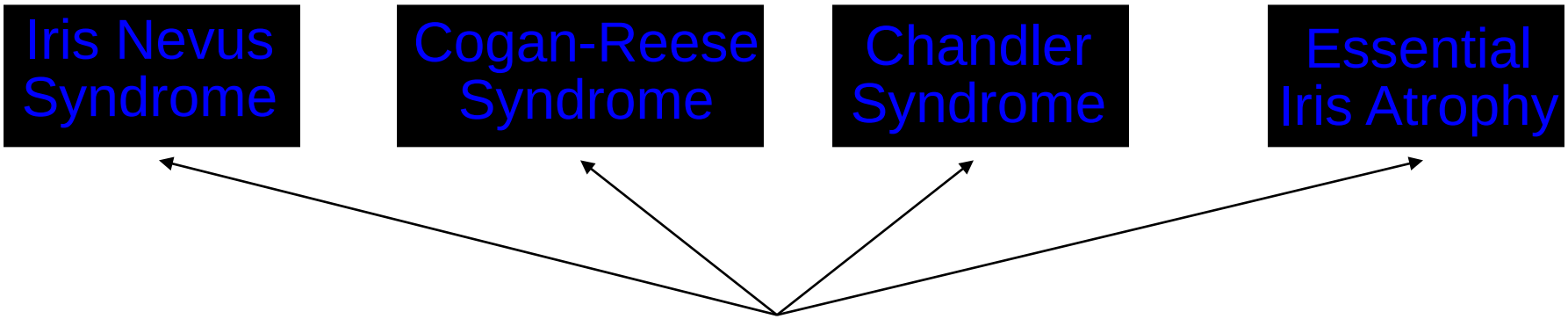


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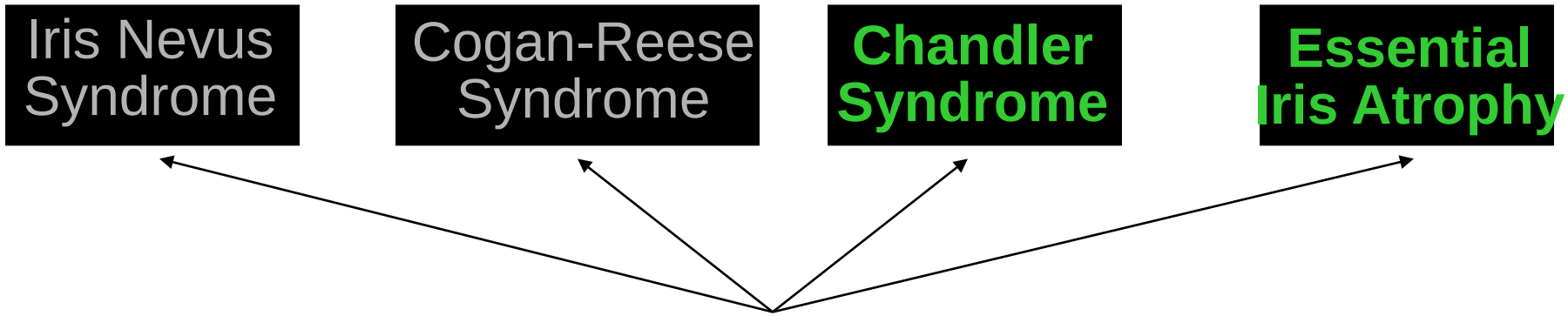


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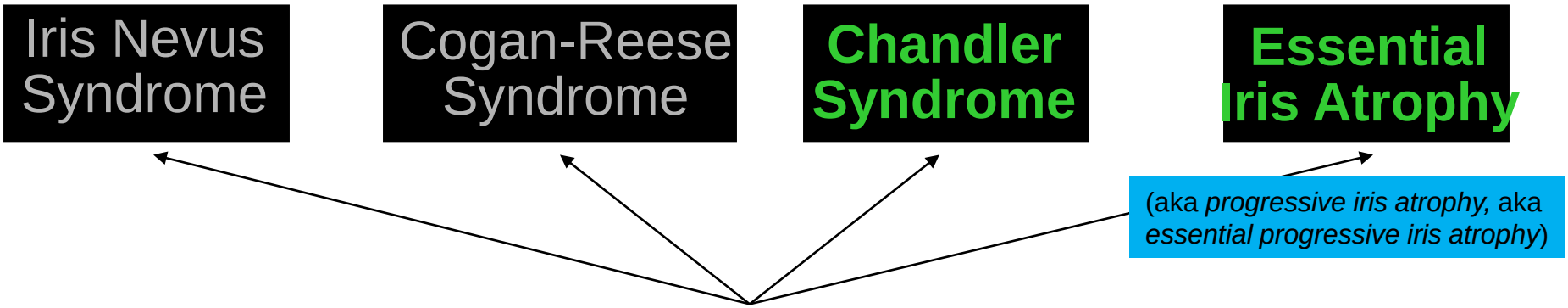


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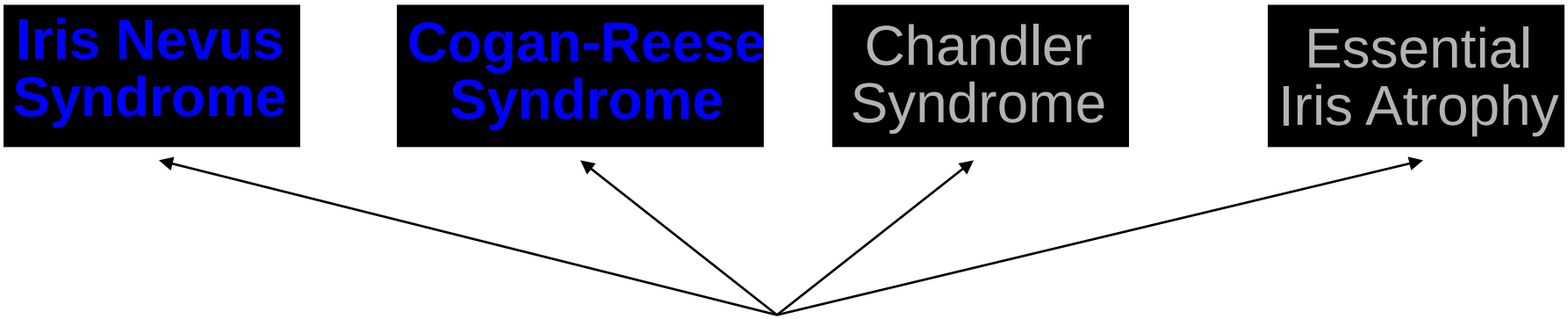
*What's up with the status of Cogan-Reese and iris nevus syndrome?*

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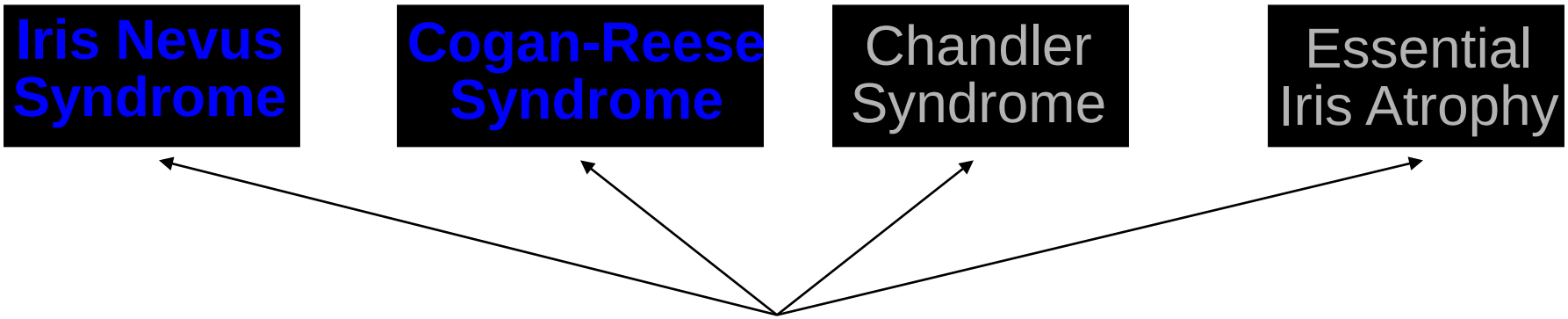
*What's up with the status of Cogan-Reese and iris nevus syndrome?*
Here's the thing: The BCSC books treat these terms as synonyms. Other sources contend that these are two separate and distinct subtypes of ICE. Still others argue that iris nevus syndrome is a separate disease entirely, ie, not a subtype of ICE at all.

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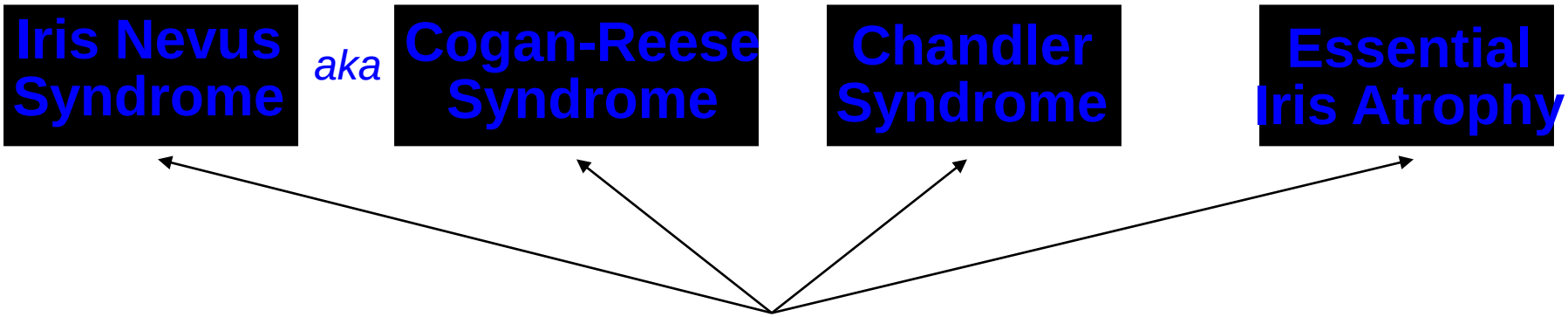


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**C***handler syndrome*

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Note that this creates a very apropos mnemonic for remembering the ICE subtypes!

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

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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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*Which subtype is most common?*
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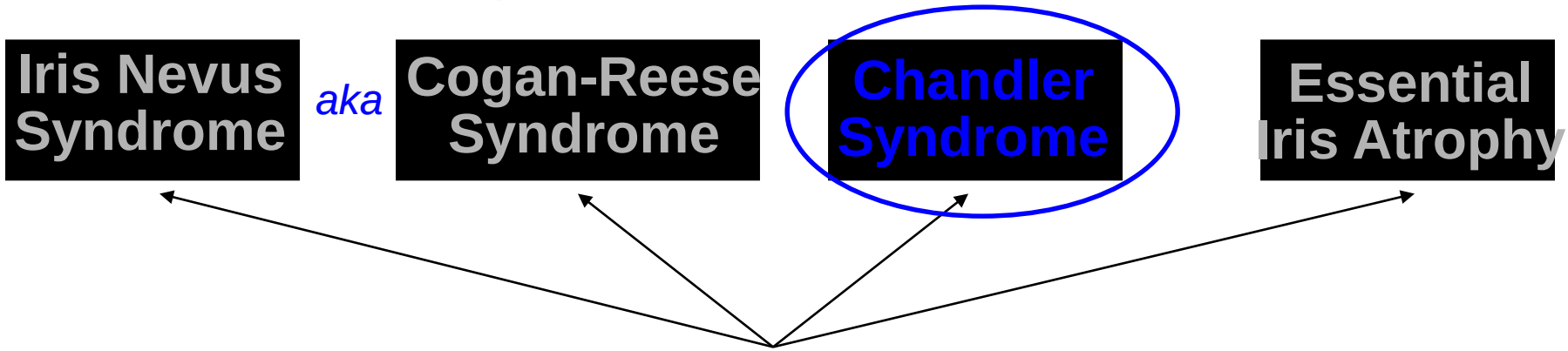


ICE: *Subtypes*

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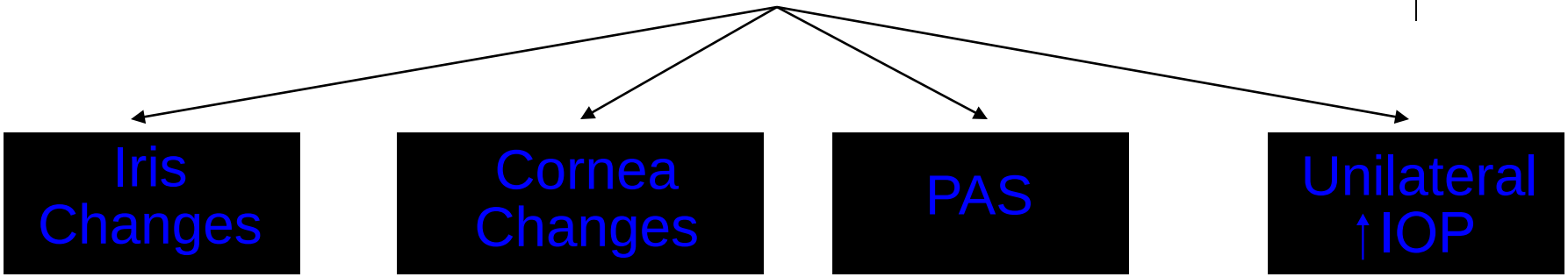


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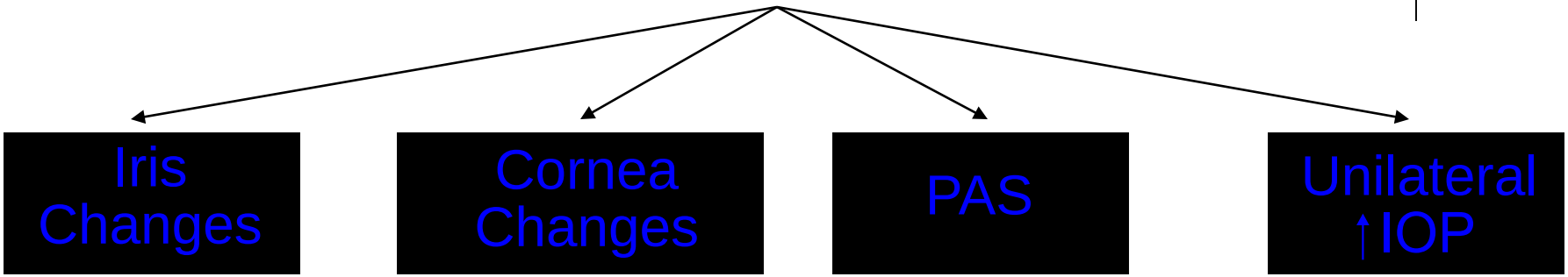


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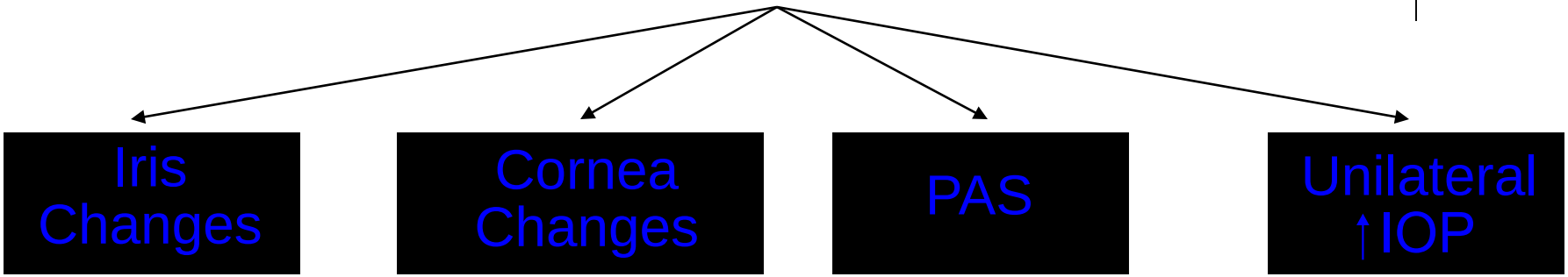


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## ICE: *Signs*

- 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**
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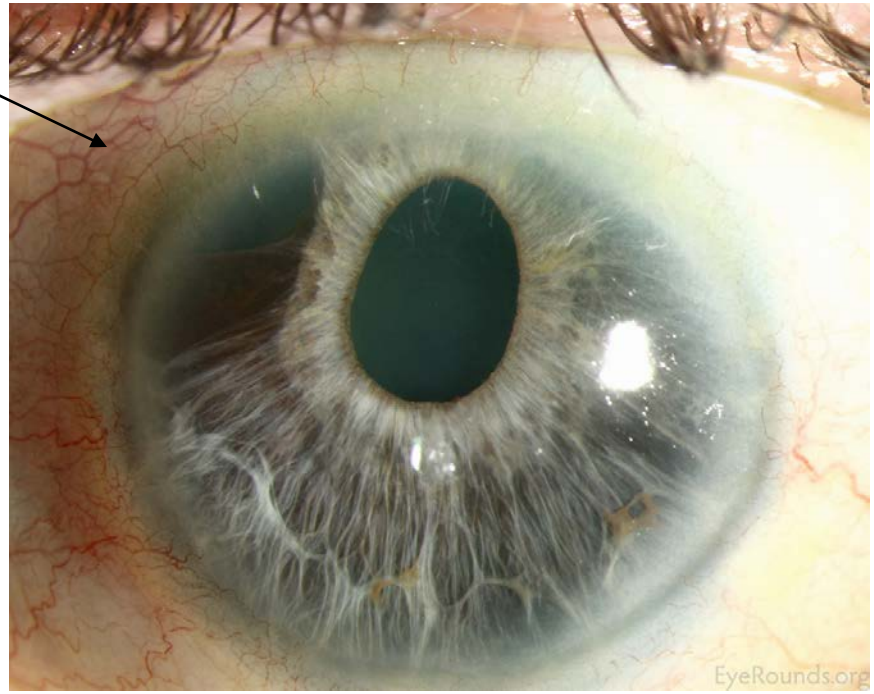

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*What is* corectopia*?*
Displacement of the pupil

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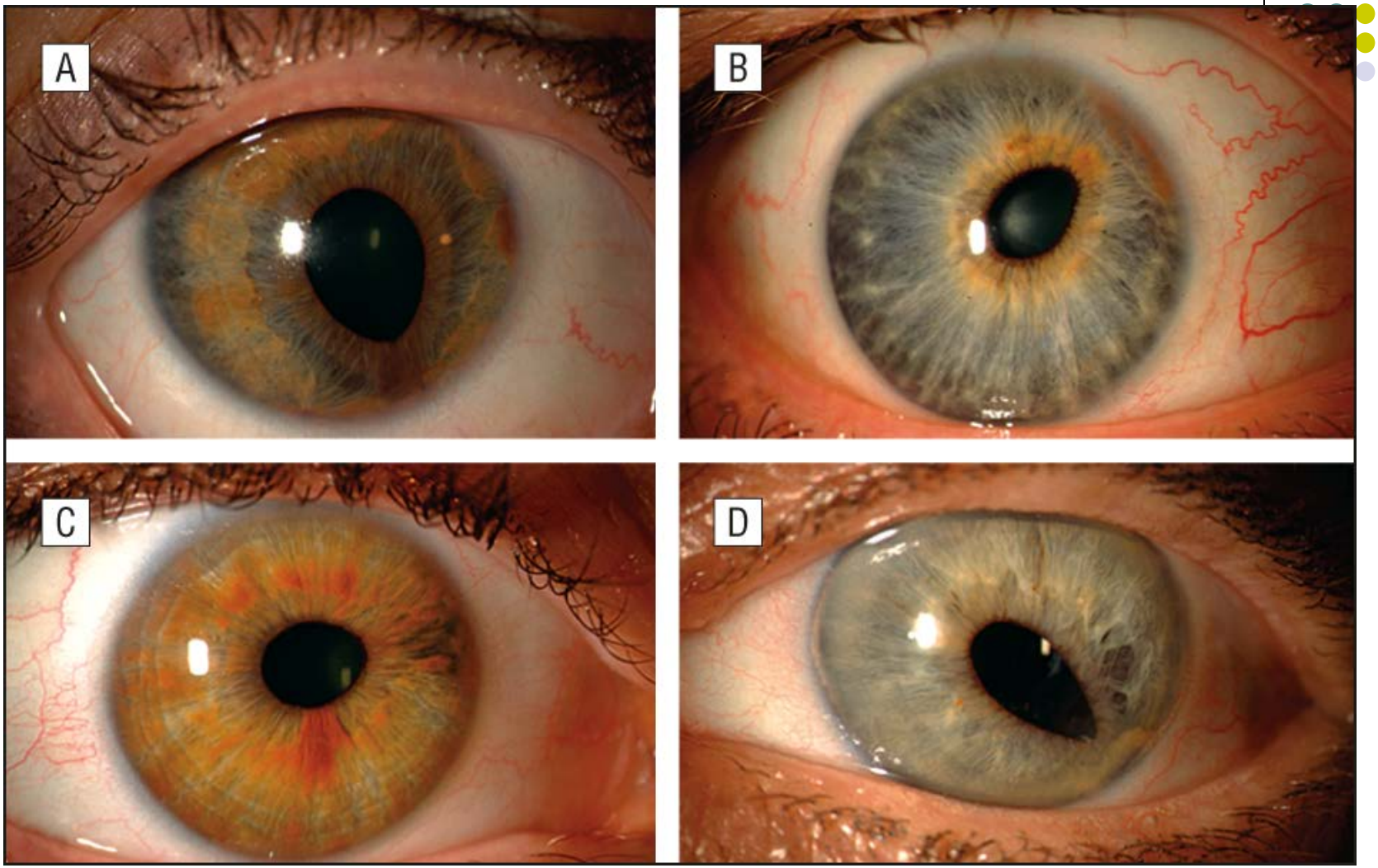


Corectopia

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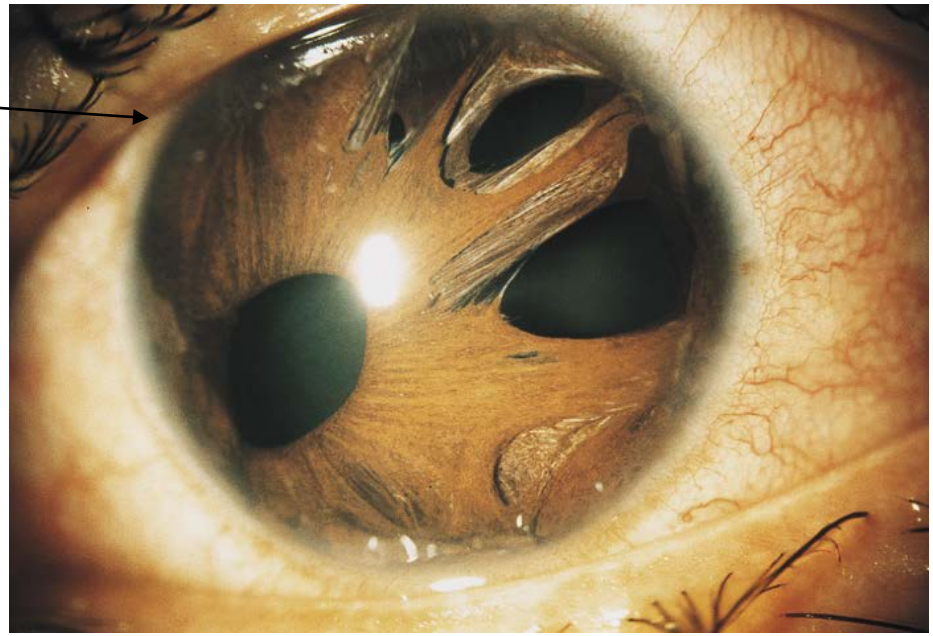

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*What is* ectropion uveae*?*

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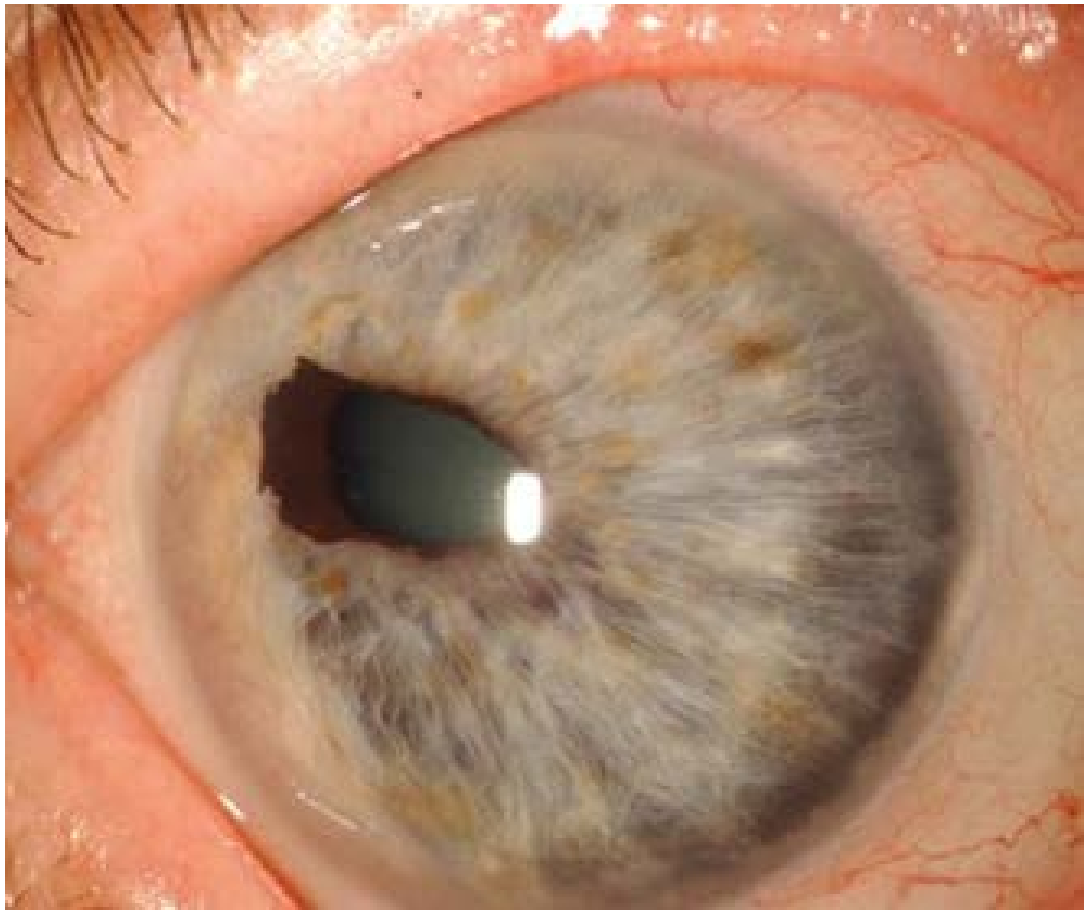
  - --Nevi?
- **Cornea Changes**
- **PAS**
- **Unilateral ↑IOP**

*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

# Iridocorneal Endothelial (ICE) Syndrome

## ICE: *Signs*

- **Iris Changes**
  - --Atrophy
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  - --Holes
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*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

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*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**
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*What one-word description is applied to the nodules?*

# Iridocorneal Endothelial (ICE) Syndrome

## ICE: *Signs*

- **Iris Changes**
  - --Atrophy
  - --Corectopia
  - --Holes
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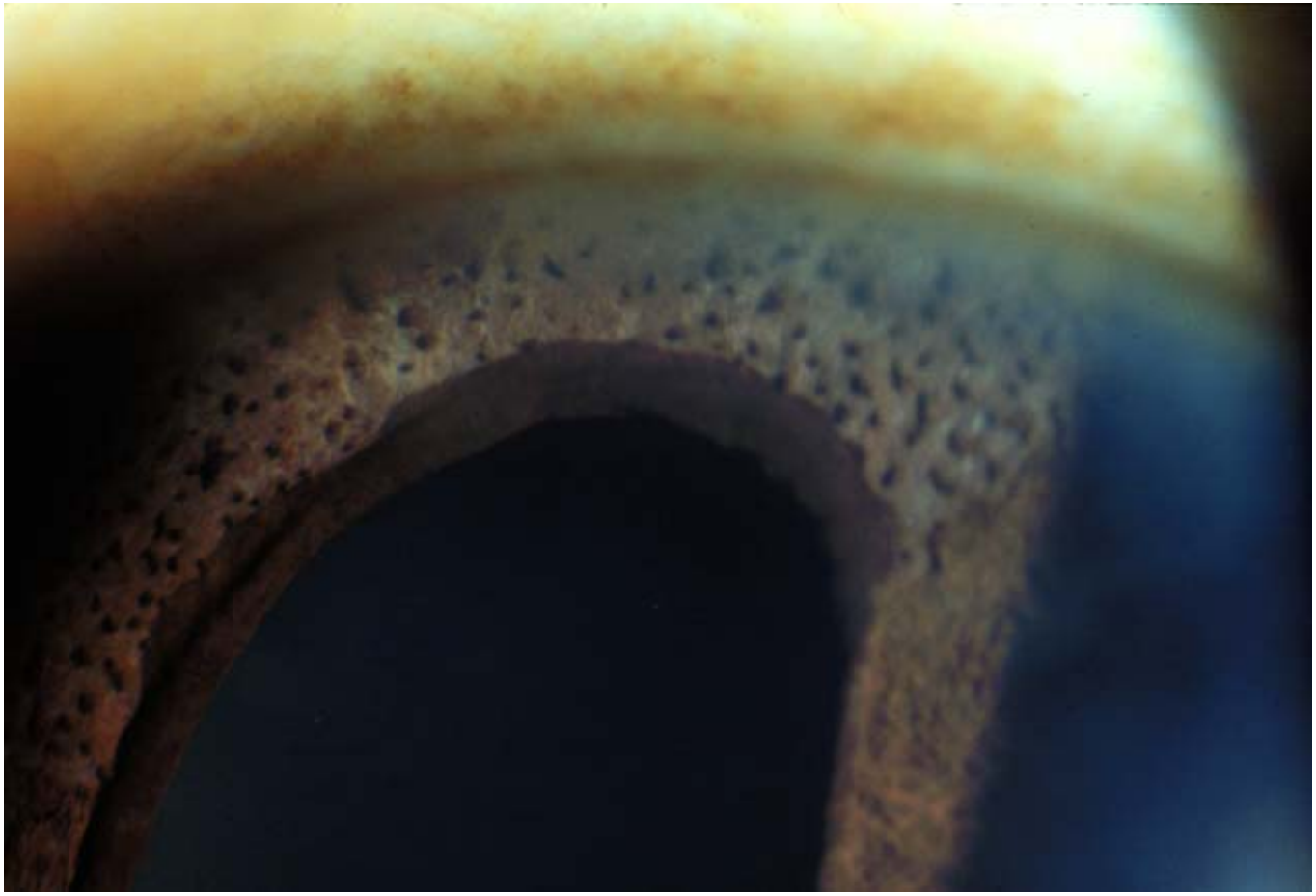
  - --Nevi?
- **Cornea Changes**
- **PAS**
- **Unilateral ↑IOP**

*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**
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*Are the 'nevi' actually nevi (ie, collections of melanocytes)?*

# Iridocorneal Endothelial (ICE) Syndrome

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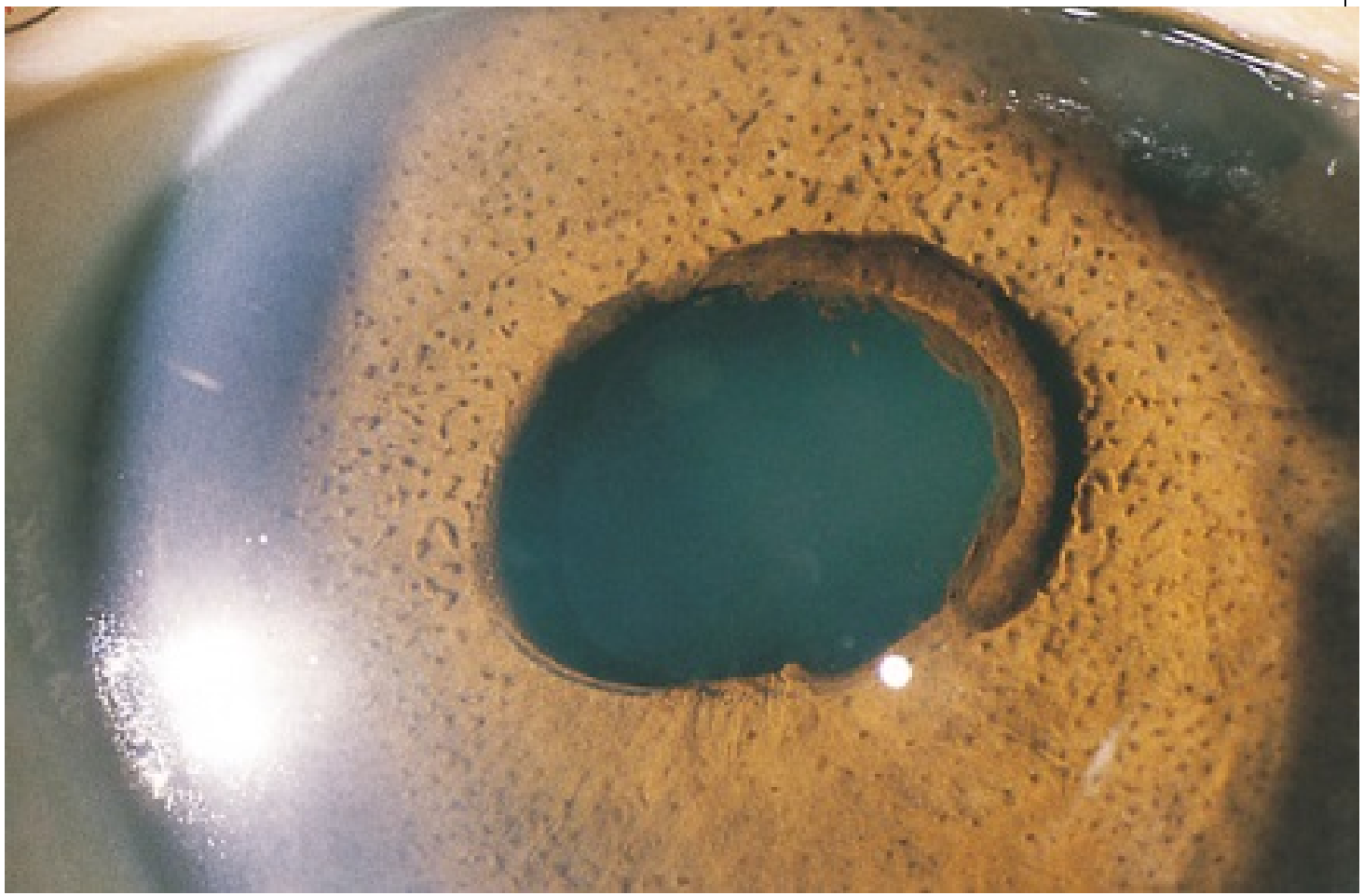
No, they are pseudo-nevi--comprised in this case of focal areas of compressed iris stroma

# Iridocorneal Endothelial (ICE) Syndrome



ICE: Iris nevi (and ectropion uveae)

# Iridocorneal Endothelial (ICE) Syndrome

## ICE: *Signs*

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

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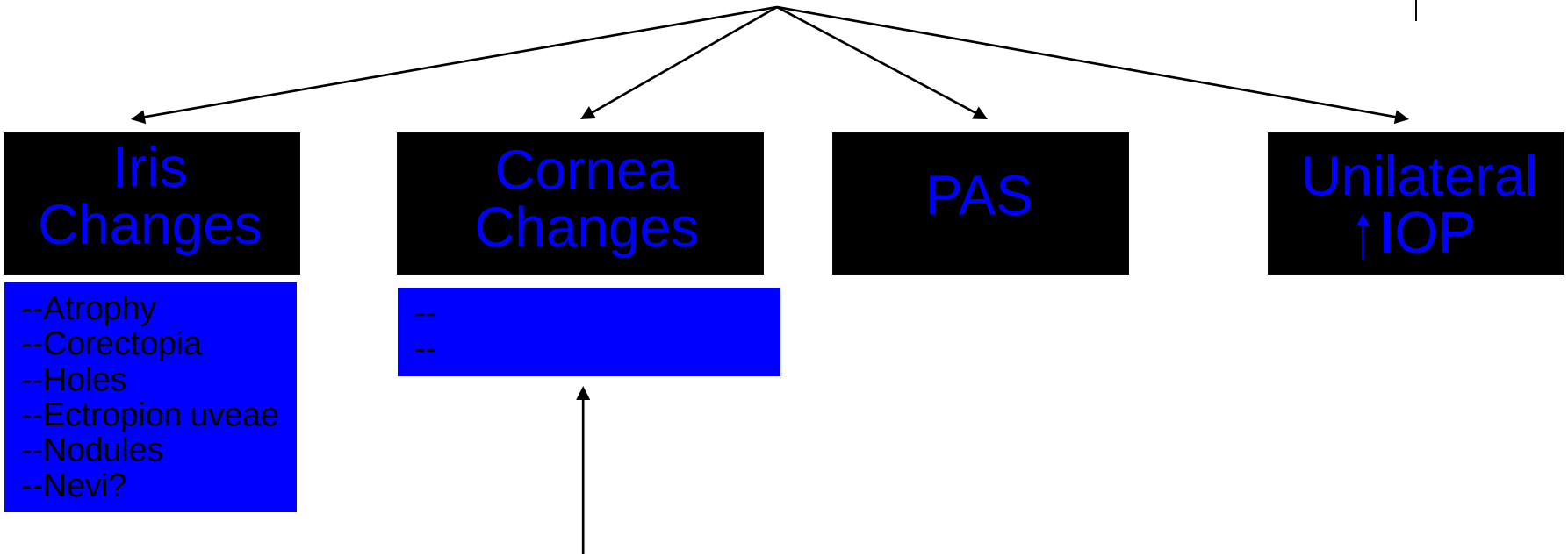
- **Cornea Changes**
- **PAS**
- **Unilateral ↑IOP**

*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

*What is the etiology of the iris-stroma compression?*
Contraction of the ICE membrane

# Iridocorneal Endothelial (ICE) Syndrome

## ICE: *Signs*



# Iridocorneal Endothelial (ICE) Syndrome

## ICE: *Signs*

- **Iris Changes**
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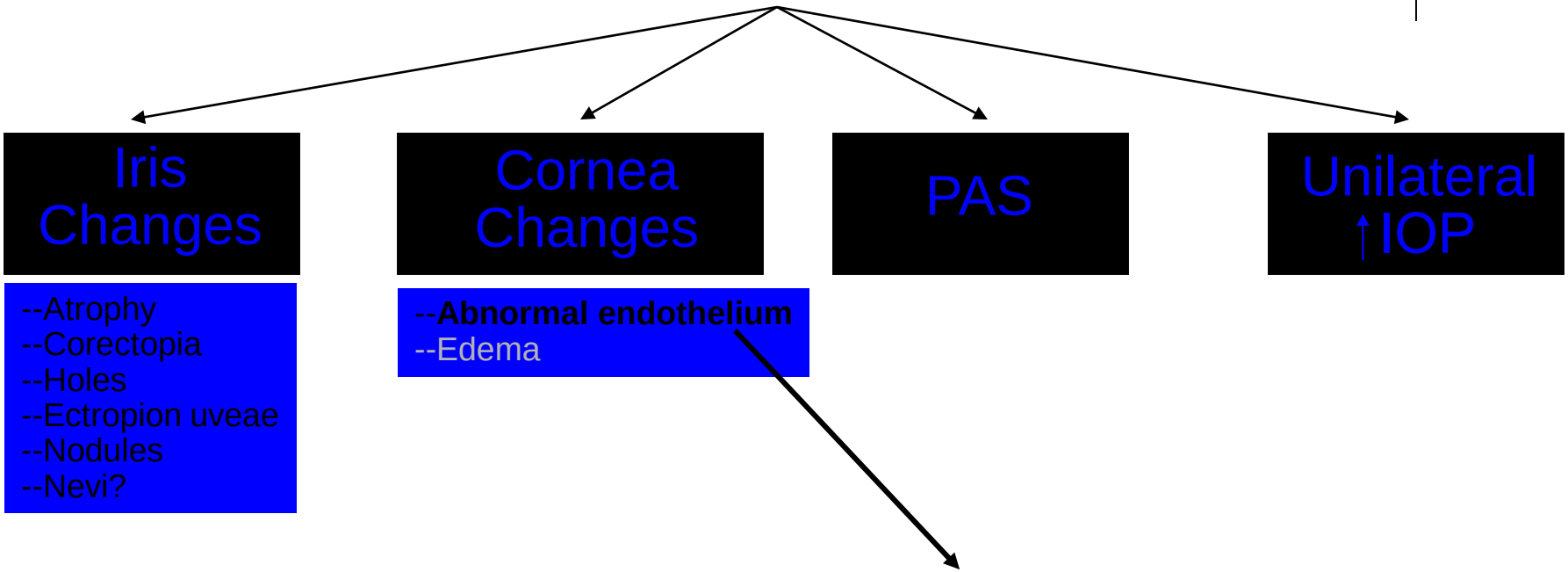
  - --Corectopia
  - --Holes
  - --Ectropion uveae
  - --Nodules
  - --Nevi?
- **Cornea Changes**
  - --Abnormal endothelium
  - --Edema
- **PAS**
- **Unilateral ↑IOP**

*What are the specific findings?*

# Iridocorneal Endothelial (ICE) Syndrome

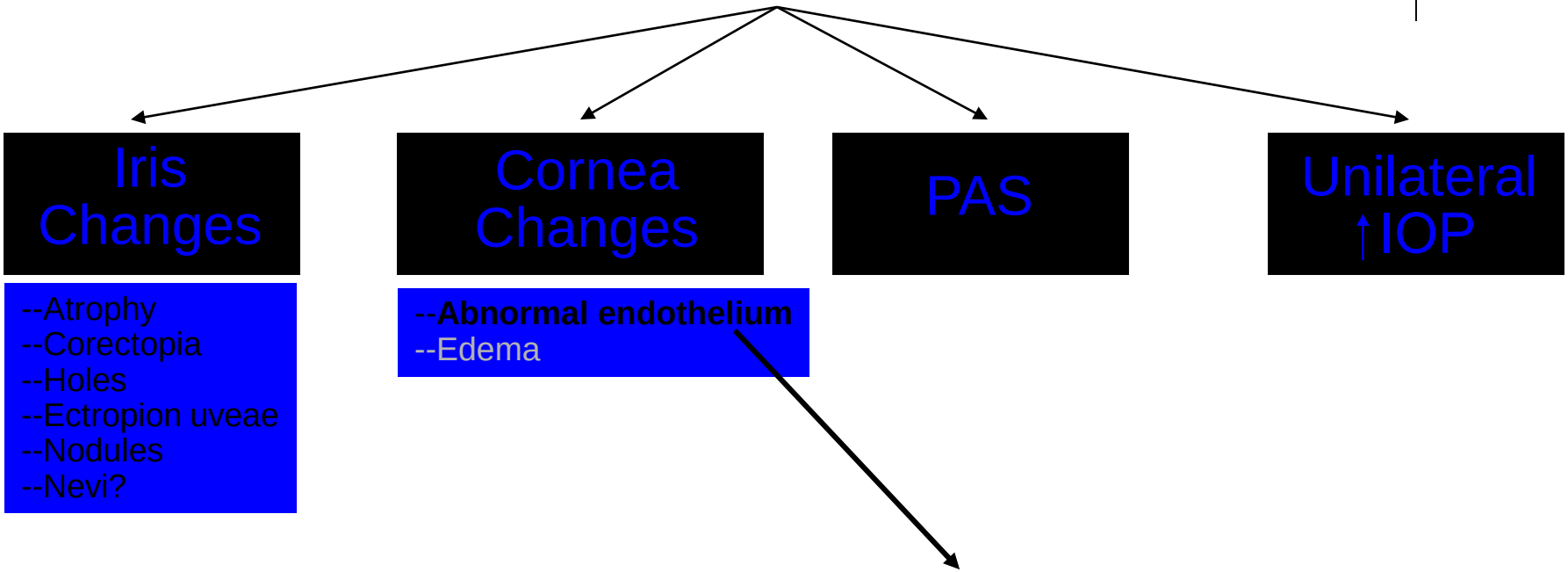
## ICE: *Signs*




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

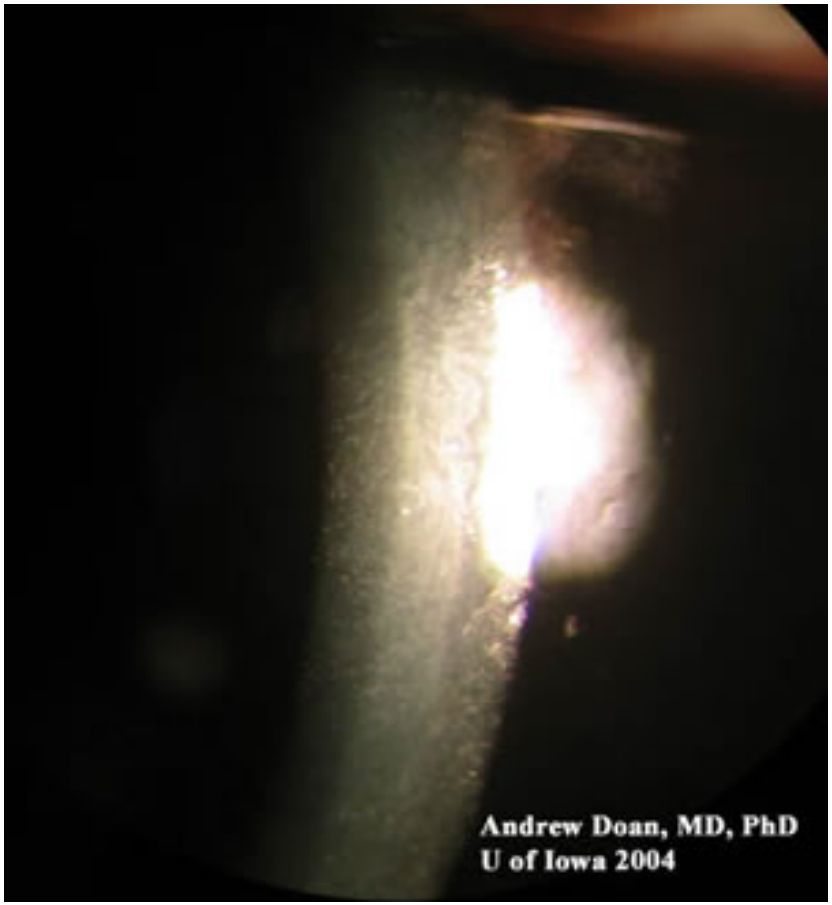
# Iridocorneal Endothelial (ICE) Syndrome

## ICE: *Signs*




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

# Iridocorneal Endothelial (ICE) Syndrome




Low res


High res

ICE: ‘Hammered silver’ corneal endothelium

# Iridocorneal Endothelial (ICE) Syndrome

## ICE: *Signs*

- **Iris Changes**
  - --Atrophy
  - --Corectopia
  - --Holes
  - --Ectropion uveae
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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?*
**'Hammered silver'**

*Another two-word descriptor for ICE endothelium is **beaten bronze.** In what other condition is this term used to describe the endothelium?*

# Iridocorneal Endothelial (ICE) Syndrome

## ICE: *Signs*

- **Iris Changes**
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**'Hammered silver'**

*Another two-word descriptor for ICE endothelium is **beaten bronze.** In what other condition is this term used to describe the endothelium?*
Fuch's endothelial dystrophy

# Iridocorneal Endothelial (ICE) Syndrome

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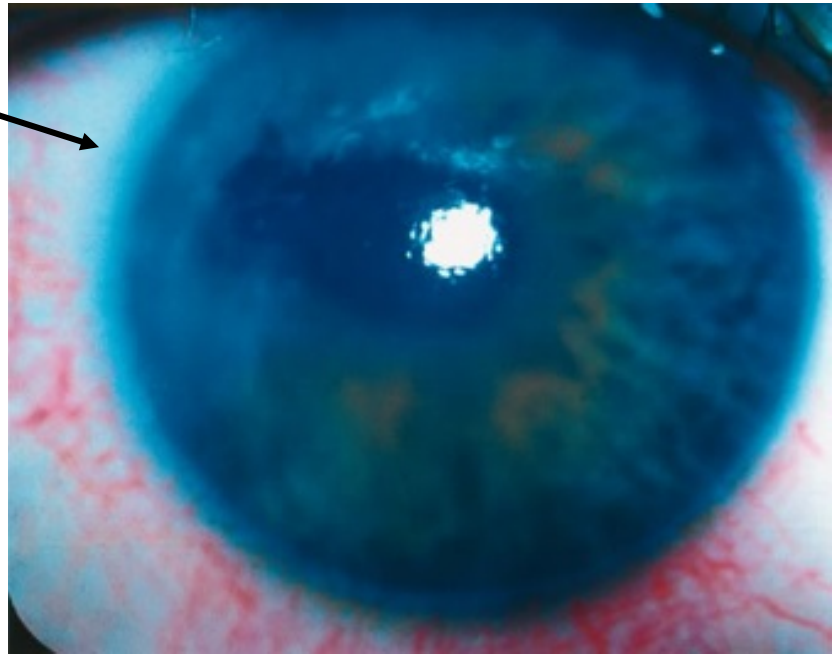
--Nodules
--Nevi?

**Cornea Changes**

--Abnormal endothelium
--**Edema**

**PAS**

**Unilateral ↑IOP**



# 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?*

**Unilateral ↑IOP**

# Iridocorneal Endothelial (ICE) Syndrome

## ICE: *Signs*

**Iris Changes**

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

**Cornea Changes**

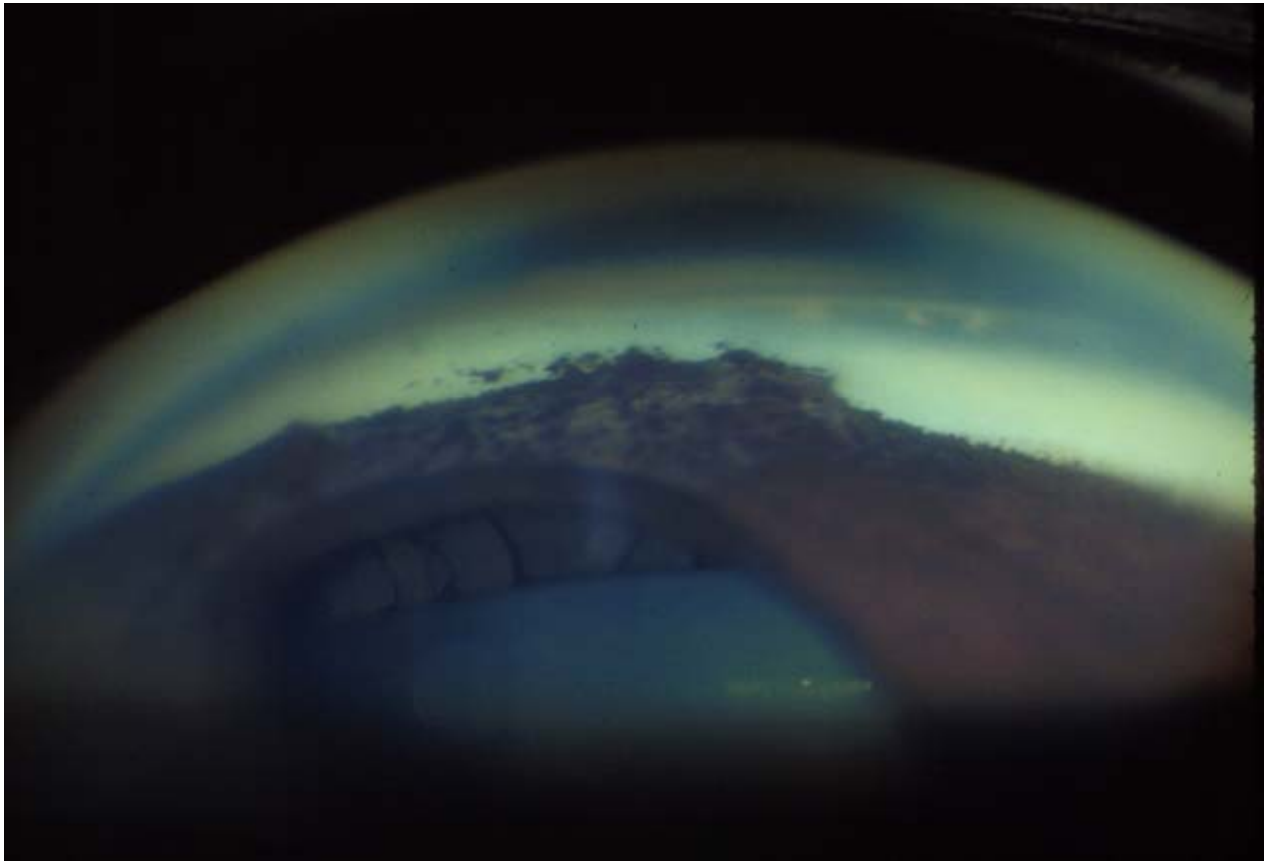
--Abnormal endothelium
--Edema

**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: PAS

# 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?*

# Iridocorneal Endothelial (ICE) Syndrome

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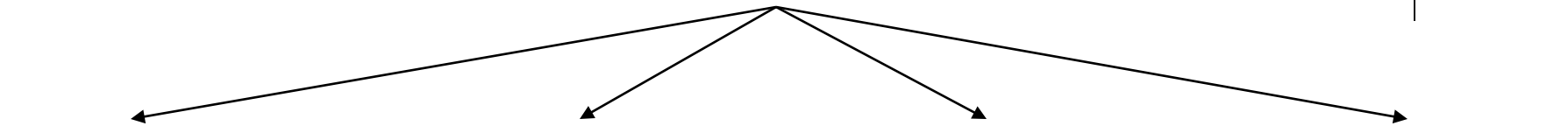

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--Nodules
--Nevi?

**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**'

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

**Unilateral ↑IOP**

# Iridocorneal Endothelial (ICE) Syndrome

## ICE: *Signs*

**Iris Changes**

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

**Cornea Changes**

--Abnormal endothelium
--Edema

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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'?*

**Unilateral ↑IOP**

# Iridocorneal Endothelial (ICE) Syndrome

## ICE: *Signs*

**Iris Changes**

--Atrophy
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--Nevi?

**Cornea Changes**

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**Unilateral ↑IOP**

*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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--Abnormal endothelium
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*Why does neo-related PAS end at SL?*

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**PAS**

*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

**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**

*How common is glaucoma in ICE?*

# Iridocorneal Endothelial (ICE) Syndrome

## ICE: *Signs*

**Iris Changes**

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--Nevi?

**Cornea Changes**

--Abnormal endothelium
--Edema

**PAS**

**Unilateral ↑IOP**

*How common is glaucoma in ICE?*
Quite—about 50% of cases

# Iridocorneal Endothelial (ICE) Syndrome

## ICE: *Signs*

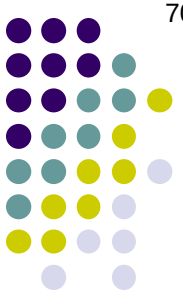
| Iris Changes | Cornea Changes | PAS | Unilateral ↑IOP |
|---|---|---|---|
| --Atrophy<br>--Corectopia<br>--Holes<br>--Ectropion uveae<br>--Nodules<br>--Nevi? | --Abnormal endothelium<br>--Edema | | |

| Iris Nevus Syndrome | *aka* | 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



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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**

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--Corectopia
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--Ectropion uveae
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--Nevi?

**Cornea Changes**

--**Abnormal endothelium**
--Edema

**PAS**

**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**

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**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**
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**Cornea Changes**
--**Abnormal endothelium**
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**Chandler syndrome**

# Iridocorneal Endothelial (ICE) Syndrome

## ICE: *Signs*

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

## ICE: *Subtypes*

# Iridocorneal Endothelial (ICE) Syndrome

## ICE: *Signs*

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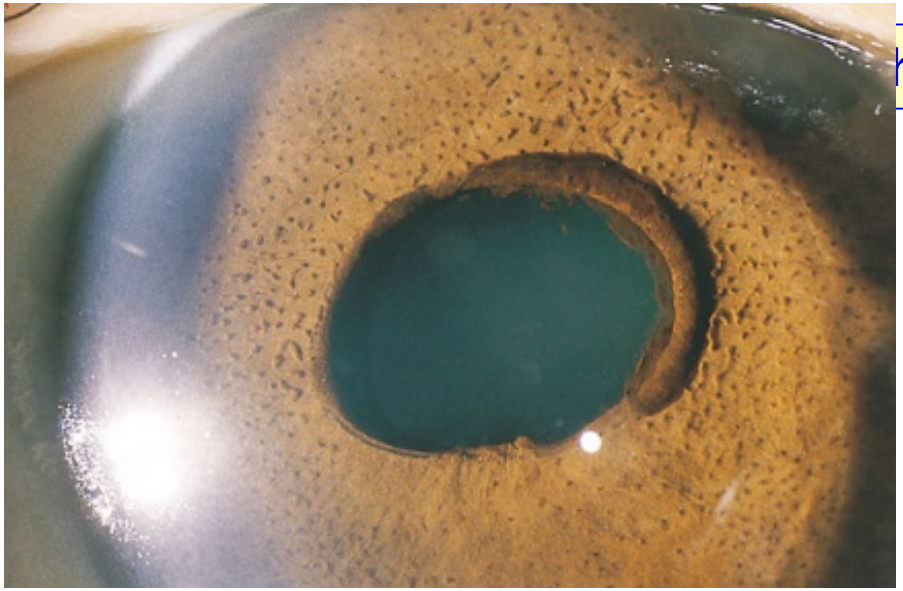
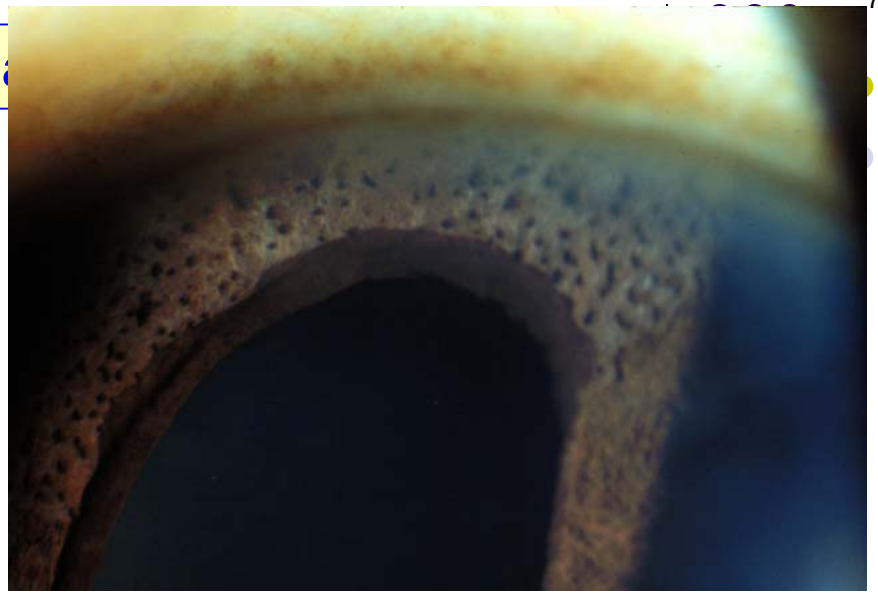
However, each has a **key finding** that 'makes' the subtype:

## ICE: *Subtypes*

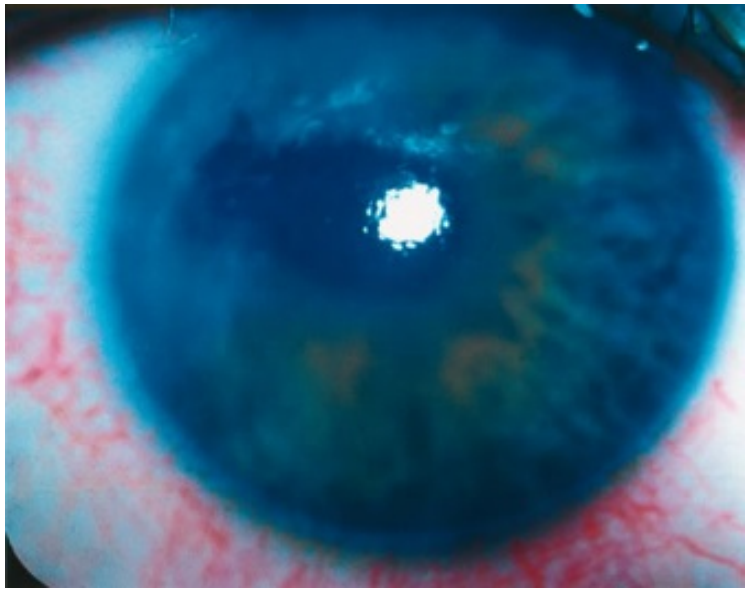
- **Iris Nevus Syndrome** (***aka*** **Cogan-Reese Syndrome**)
  - **--Iris nevi /nodules**
- **Chandler Syndrome**
  - **--Corneal edema predominates**
- **Essential Iris Atrophy**
  - **--Iris atrophy/holes predominate**



Iris nevus/Cogan-Reese syndrome



Iris nevus/Cogan-Reese syndrome



Chandler syndrome

Essential iris atrophy

# 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?*

## ICE: *Subtypes*

- **Iris Nevus Syndrome** (***aka*** **Cogan-Reese Syndrome**)
  - **--Iris nevi /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 *aka* Cogan-Reese Syndrome

***--Iris nevi /nodules***

Chandler Syndrome

**--Corneal edema predominates**
***--Relatively nl iris***

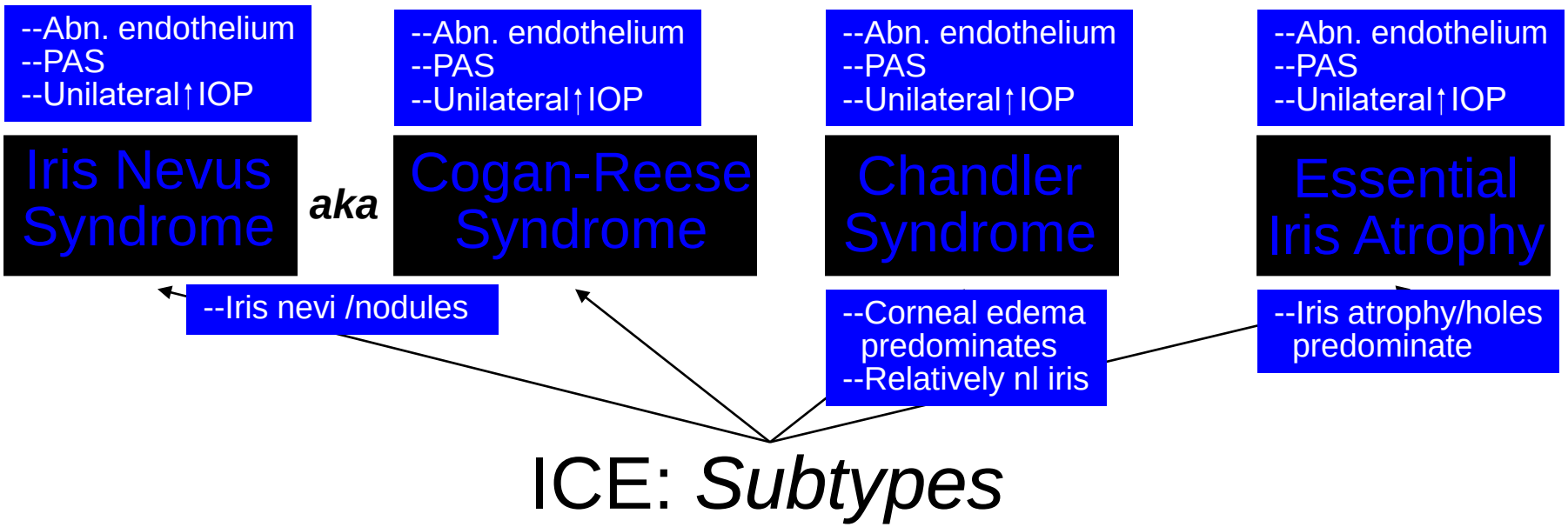
Essential Iris Atrophy

***--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

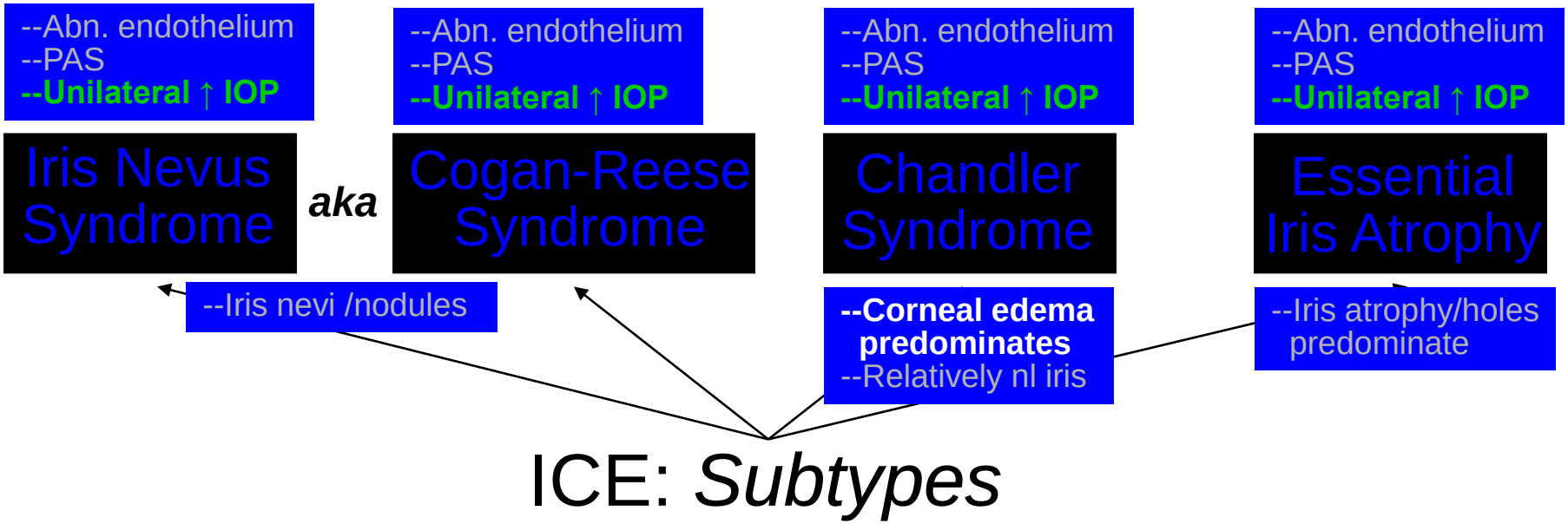
*What are the main management issues in ICE?*
***1)***
***2)***




ICE: *Subtypes*

# Iridocorneal Endothelial (ICE) Syndrome

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



## Iridocorneal Endothelial (

*Which meds are most effective?*

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

# ICE: *Subtypes*

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

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

--Iris nevi /nodules

**Chandler Syndrome**

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

**--Corneal edema predominates**
--Relatively nl iris

**Essential Iris Atrophy**

--Abn. endothelium
--PAS
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--Iris atrophy/holes predominate

# Iridocorneal Endothelial (

*Which meds are most effective?*
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--α agonists

*What are the main management issues in ICE?*
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***2)*** **Minimizing corneal edema**

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**Iris Nevus Syndrome**

--Iris nevi /nodules

***aka***

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**Chandler Syndrome**

**--Corneal edema predominates**
--Relatively nl iris

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## ICE: *Subtypes*

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