

Ernst Fuchs (1851-1930)





Rhetorical question at this juncture—proceed





Seen mainly by pathologists



Fuchs adenoma

Seen mainly by pathologists



Fuchs adenoma

Seen mainly by pathologists





Seen every day at the slit lamp

Fuchs adenoma

Seen mainly by pathologists



Fuchs crypts

Seen every day at the slit lamp

Fuchs adenoma

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Fuchs crypts

Seen every day at the slit lamp

Fuchs

two words-itis

An uncommon cornea condition

Fuchs adenoma

Seen mainly by pathologists



Fuchs crypts

Seen every day at the slit lamp

Fuchs marginal keratitis

An uncommon cornea condition

Fuchs adenoma

Seen mainly by pathologists



Fuchs crypts

Seen every day at the slit lamp

superficial Fuchs marginal keratitis

Ernst Fuchs (1851-1930)

Note that it is also known (including in the *Cornea* book) as Fuchs *superficial* marginal keratitis

Fuchs adenoma

Seen mainly by pathologists



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Dalén - Fuchs nodule

Fuchs adenoma

Seen mainly by pathologists

Fuchs something-oma

Has an interesting VF finding



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Fuchs coloboma

Has an interesting VF finding



Fuchs crypts

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Fuchs marginal keratitis

An uncommon cornea condition

Ernst Fuchs (1851-1930) Fuchs one word

IMHO, the most obscure of them all

Dalén - Fuchs nodule

Fuchs adenoma

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Fuchs coloboma

Has an interesting VF finding



Fuchs crypts

Seen every day at the slit lamp

Fuchs marginal keratitis

An uncommon cornea condition

Ernst Fuchs (1851-1930) Fuchs spot

IMHO, the most obscure of them all

Dalén - Fuchs nodule

Ernst Fuchs

(1851-1930)

Fuchs adenoma

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Has an interesting VF finding



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Fuchs marginal keratitis

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Fuchs spot

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Has an interesting VF finding



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Fuchs marginal keratitis

An uncommon cornea condition

Fuchs spot

IMHO, the most obscure of them all

Fuchs endothelial dystrophy

IMHO, the least obscure of them all

Dalén - Fuchs nodule

An occasional DFE finding

Fuchs two words

Of interest to the most subspecialties

Ernst Fuchs

(1851-1930)

Fuchs adenoma

Seen mainly by pathologists

Fuchs coloboma

Has an interesting VF finding



Fuchs crypts

Seen every day at the slit lamp

Fuchs marginal keratitis

An uncommon cornea condition

Fuchs spot

IMHO, the most obscure of them all

Fuchs endothelial dystrophy

IMHO, the least obscure of them all

Dalén - Fuchs nodule

An occasional DFE finding

Fuchs heterochromic iridocyclitis

Of interest to the most subspecialties

Fuchs adenoma

Fuchs coloboma

What are the two noneponymous names for Fuchs adenoma?

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--

Fuchs crypts

Fuchs spot

Fuchs marginal keratitis

Ernst Fuchs (1851-1930)

Fuchs endothelial dystrophy

Dalén - Fuchs nodule



Fuchs coloboma

Fuchs adenoma

What are the two noneponymous names for Fuchs adenoma?

- --Pseudoadenomatous hyperplasia of the ciliary body
- -- Coronal adenoma

Fuchs crypts

Fuchs spot

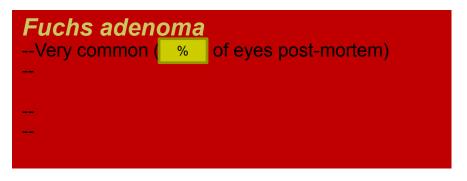
Fuchs marginal keratitis

Ernst Fuchs (1851-1930)

Fuchs endothelial dystrophy

Dalén - Fuchs nodule





Fuchs coloboma



Fuchs crypts

Fuchs spot

Fuchs marginal keratitis

Ernst Fuchs (1851-1930)

Fuchs endothelial dystrophy

Dalén - Fuchs nodule

Fuchs adenoma

--Very common (~25% of eyes post-mortem)

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Fuchs coloboma

Fuchs crypts

Fuchs spot

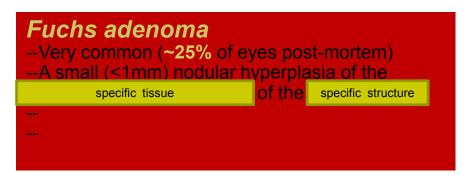
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Ernst Fuchs (1851-1930)

Fuchs endothelial dystrophy

Dalén - Fuchs nodule





Fuchs coloboma



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Fuchs adenoma

- --Very common (~25% of eyes post-mortem)
- --A small (<1mm) nodular hyperplasia of the nonpigmented epithelium of the ciliary body
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- -

Fuchs coloboma



Fuchs crypts

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Ernst Fuchs (1851-1930)

Fuchs endothelial dystrophy

Dalén - Fuchs nodule

Fuchs adenoma

--Very common (~25% of eyes post-mortem)

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Embryologically speaking: From what tissue does the nonpigmented epithelium of the ciliary body derive?

Fuchs coloboma



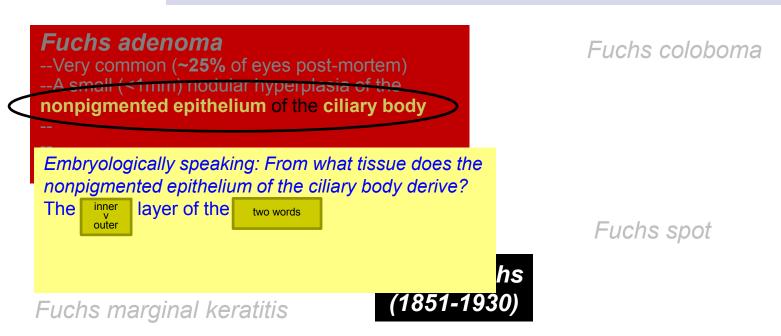
Fuchs spot

ns (1851-1930)

Fuchs marginal keratitis

Fuchs endothelial dystrophy

Dalén - Fuchs nodule



Fuchs endothelial dystrophy

Dalén - Fuchs nodule



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Embryologically speaking: From what tissue does the nonpigmented epithelium of the ciliary body derive? The inner layer of the optic cup

Fuchs coloboma



Fuchs spot

n: (1851-1930)

Fuchs marginal keratitis

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Dalén - Fuchs nodule

(1851-1930)

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Embryologically speaking: From what tissue does the nonpigmented epithelium of the ciliary body derive? The inner layer of the optic cup

What is this layer called?

Fuchs marginal keratitis



Fuchs spot

Fuchs endothelial dystrophy

Dalén - Fuchs nodule



Fuchs adenoma

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--A small (<1mm) nodular hyperplasia of the nonpigmented epithelium of the ciliary body

Embryologically speaking: From what tissue does the nonpigmented epithelium of the ciliary body derive? The inner layer of the optic cup

What is this layer called? The medullary epithelium

Fuchs marginal keratitis

Fuchs coloboma

Fuchs spot

(1851-1930)

Fuchs endothelial dystrophy

Dalén - Fuchs nodule



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Embryologically speaking: From what tissue does the nonpigmented epithelium of the ciliary body derive? The inner layer of the optic cup

What is this layer called?

The medullary epithelium

Fuchs marginal keratitis

This factoid is trivial—it gets a passing mention in one book only (*Path*). It's not even mentioned during the detailed coverage of eye embryology in *Fundamentals*. I feel 100%

Fuchs coloboma

certain you will never be asked *What's the name of the inner layer of the optic cup?* So why (you're asking yourself) mention it here?

Dalén - Fuchs nodule

Fuchs coloboma



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Dalén - Fuchs nodule

Fuchs adenoma

--Very common (~25% of eyes post-mortem)

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What extremely important function is served by the nonpigmented epithelium of the ciliary body?

Fuchs coloboma



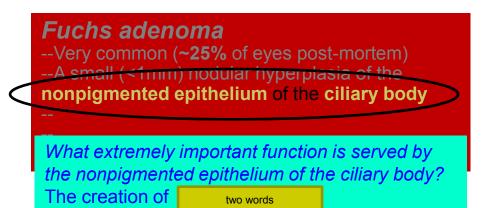
Fuchs spot

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Fuchs endothelial dystrophy

Dalén - Fuchs nodule



Fuchs coloboma



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What extremely important function is served by the nonpigmented epithelium of the ciliary body? The creation of **aqueous humor**

Fuchs coloboma



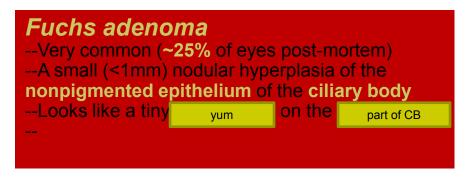
Fuchs spot

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Dalén - Fuchs nodule



Fuchs coloboma



Fuchs crypts

Fuchs spot

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Fuchs spot

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Ernst Fuchs (1851-1930)

Fuchs endothelial dystrophy

Dalén - Fuchs nodule







Fuchs adenoma—'a tiny pearl onion on the pars plicata'

Fuchs adenoma

- --Very common (~25% of eyes post-mortem)
- --A small (<1mm) nodular hyperplasia of the nonpigmented epithelium of the ciliary body
- --Looks like a tiny pearl onion on the pars plicata
- --Benign, but can cause two words be mistaken for a ciliary-body

and/or

Fuchs crypts

Fuchs spot

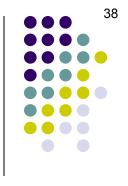
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- --Benign, but can cause **sectoral cataract** and/or be mistaken for a ciliary-body **melanoma**

Fuchs coloboma



Fuchs crypts

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There's another tumor related to the nonpigmented epithelium of the ciliary body that is not nearly as benign as Fuchs adenoma. What is it?

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Fuchs spot

hs 0)

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Medulloepithelioma (aka

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Fuchs spot

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Why are medulloepitheliomas called medulloepitheliomas? (You know this!)

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Because they derive from the inner layer of the optic cup, aka the **medullary epithelium** (this is what the earlier foreshadowing was about)

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So, why isn't Fuchs adenoma aka medulloepithelioma?

Fuchs coloboma



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So, why isn't Fuchs adenoma aka medulloepithelioma? Because it arises from the 'mature' epithelium, not its embryologic precursor a la medulloepithelioma

Fuchs coloboma



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hs endothelial dystrophy

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Medulloepithelioma (aka diktyoma)

Why are medulloepitheliomas called medulloepitheliomas? (You know this!)

Because they derive from the inner layer of the optic cup,

Because it arises from primordial/embryologic tissue, this would imply that medulloepithelioma must be present at birth, ie, congenital. Is this the case?

embryologic precursor a la medulloepithelioma

Fuchs coloboma



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embryologic precursor a la medulloepithelioma



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hs endothelial dystrophy

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Fuchs adenoma

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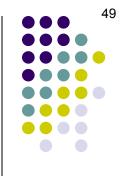
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Fuchs coloboma



In what sense is a medulloepithelioma 'not benign'? Does that mean it's a malignancy?

chs spot

othelial dystrophy

Dalén - Fuchs nodule

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Not exactly. While it's the case that the histology of the tumor *can* include malignant characteristics, clinically the tumor tends not to act malignantly in the sense that it only rarely metastasizes.

Fuchs coloboma

chs spot

thelial dystrophy

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It means that medulloepitheliomas tend to be locally very aggressive

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How 'locally aggressive' are we talking about here?

Dalén - Fuchs nodule

Fuchs coloboma

-uciis colobollia

53

chs spot

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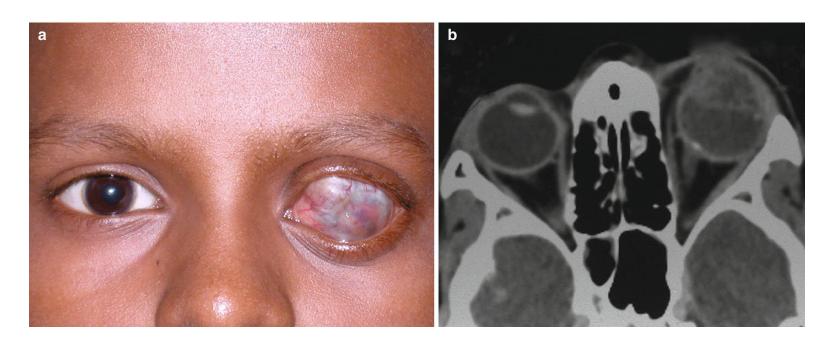
How 'locally aggressive' are we talking about here? Aggressive enough to be fatal

Dalén - Fuchs nodule

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thelial dystrophy





Fuchs adenoma

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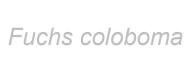
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Medulloepithelioma (aka diktyoma)

Is it common, or rare?



Fuchs spot



Fuchs endothelial dystrophy

Dalén - Fuchs nodule



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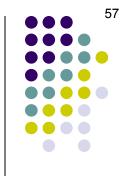
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Is it common, or rare?

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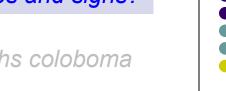
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Is it common, or rare? Rare

Does medulloepithelioma present post-mortem?

Fuchs coloboma



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Fuchs spot



Fuchs endothelial dystrophy

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Is it common, or rare?
Rare

Does medulloepithelioma present post-mortem? No, it presents in childhood



Fuchs spot



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Is it common, or rare? Rare

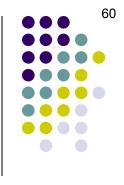
Does medulloepithelioma present post-mortem? No, it presents in childhood, age range, and unit

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Medulloepithelioma (aka diktyoma)

Is it common, or rare?
Rare

Does medulloepithelioma present post-mortem? No, it presents in childhood, age 4-12 years Fuchs coloboma



Fuchs spot

hs 0)

Fuchs endothelial dystrophy

Dalén - Fuchs nodule

Fuchs adenoma

--Very common (~25% of eyes post-mortem)

-- A small (<1mm) nodular hyperplasia of the

nonpigmented epithelium of the ciliary body

--Looks like a tiny nearl onion on the pars pilcata

There's another tumor related to the nonpigmented epithelium of the ciliary body that is not nearly as benign as Fuchs adenoma. What is it?

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Age 4-12? But it's congenital. What gives?



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> Age 4-12? But it's congenital. What gives? While present at birth, it doesn't become clinically apparent until age 4-12 or so

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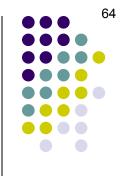
How does it present?

Fuchs coloboma

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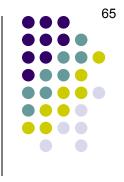
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How does it present? Usually as an iris mass Fuchs coloboma

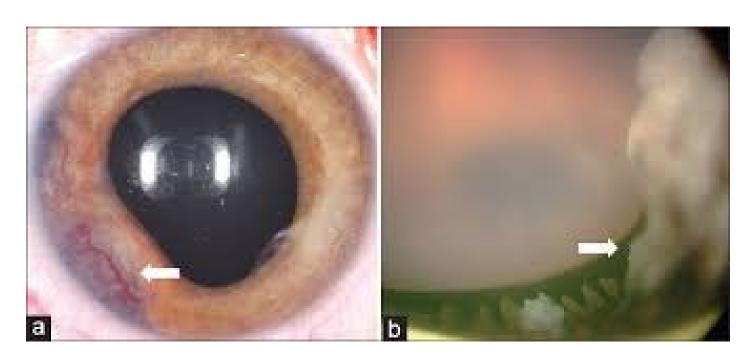
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How does it present?

Usually as an iris mass along with one or more of the following:

- --?
- --?

Dalén - Fuchs nodule

Fuchs coloboma

67

Fuchs spot

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dulloepithelioma present post-mortem? it presents in childhood age 4-12 years

How does it present?

Usually as an iris mass along with one or more of the following:

- --Glaucoma
- --Hyphema
- --Sectoral cataract

Dalén - Fuchs nodule

Fuchs coloboma

68

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How is it managed? Enucleation is usually required



70

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Fuchs crypts

--Craterlike openings in surface of the anterior

Fuchs coloboma



Fuchs spot

Fuchs marginal keratitis

Ernst Fuchs (1851-1930)

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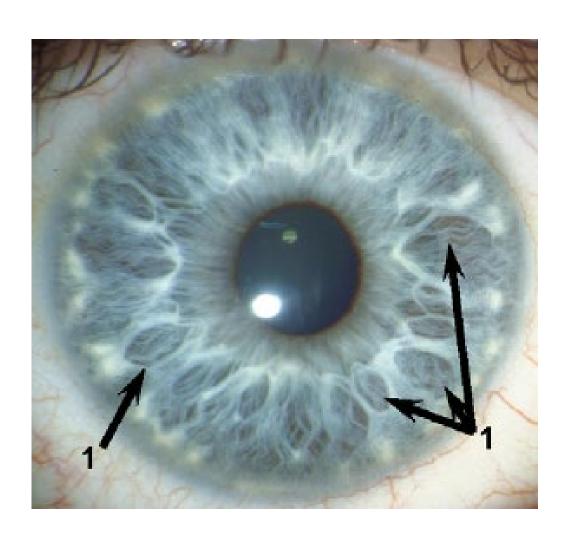
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--Peripheral non- vs ulcerative keratitis

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Fuchs marginal keratitis

- --Peripheral **nonulcerative** keratitis
- --Produces progressive thinning, can result in...[bad sequelae]

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- --Peripheral nonulcerative keratitis
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- --Resembles

Eponym + word + word

Fuchs endothelial dystrophy

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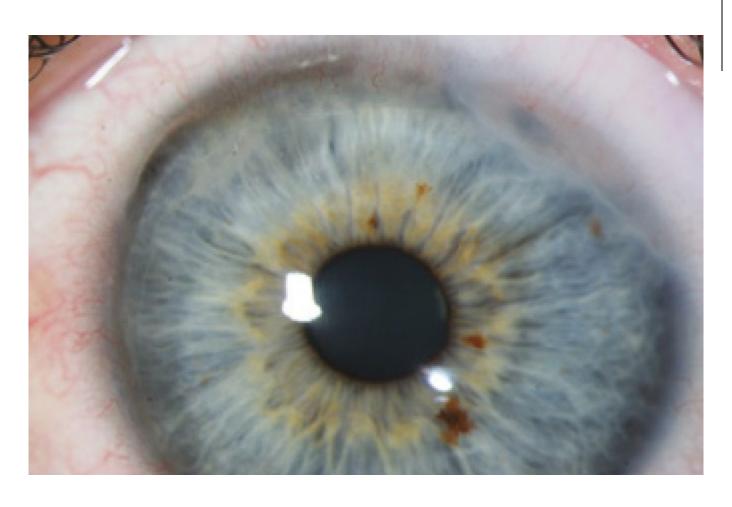
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What is the fundamental difference between Fuchs marginal keratitis and Terriens?

Dalén - Fuchs nodule

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What is the fundamental difference between Fuchs marginal keratitis and Terriens? Fuchs marginal is an inflammatory condition, whereas Terriens is **noninflammatory**

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Dalén - Fuchs nodule

--Highly focal aggregate of inflammatory cells beneath the... [specific tissue]

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--Highly focal aggregate of inflammatory cells beneath the...**RPE**

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- --Seen in granulomatous conditions, esp...[bad dz]

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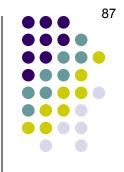
Fuchs marginal keratitis

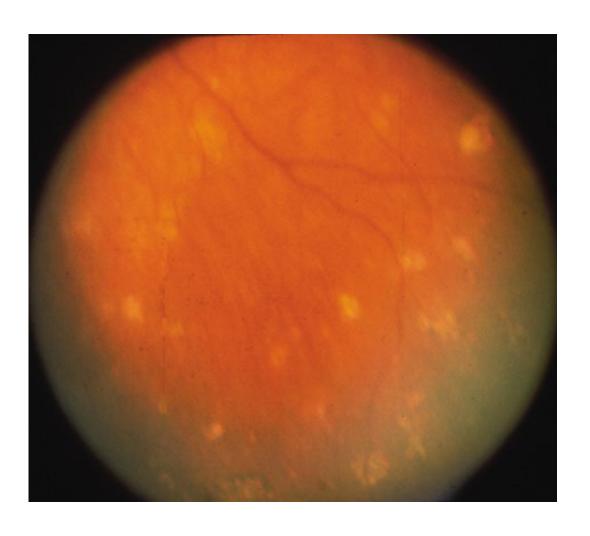
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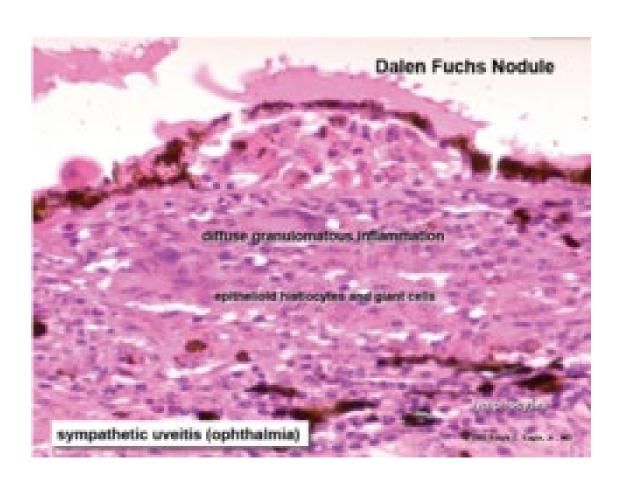
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- --Seen in granulomatous conditions, esp...sympathetic ophthalmia (SO)





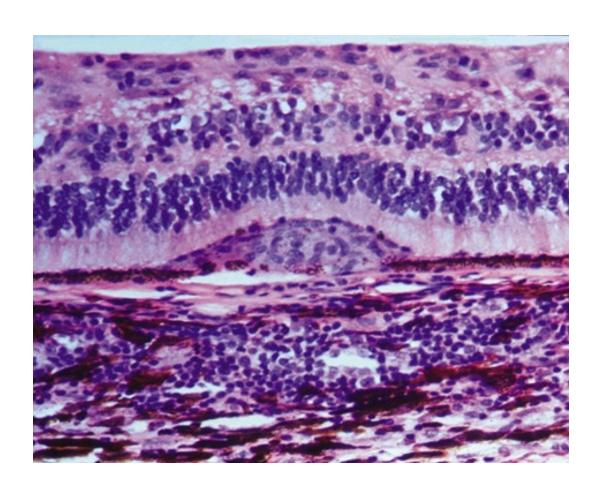
Dalén-Fuchs nodules





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The classic test-association for Dalén-Fuchs nodules is SO. That said, whenever SO is on the DDx, another condition should be as well. What is it?

cens beneath the...RFE

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Vogt-Koyanagi-Harada (VKH) dz. These two diagnoses are like salt-and-pepper shakers—they go everywhere together.

Cells Defleath the...KFE

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There is another condition—vastly more common than either SO or VKH—in which Dalén-Fuchs nodules are seen as well. So, whereas during an exam your first response to the term Dalén-Fuchs nodule should be SO/VKH, what condition should come to mind first if you encounter them in the clinic?

cells belleath the...KPE

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cells belleath the...KFE

Ernst Fuchs

(1851-1930)

96

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Fuchs coloboma --aka... two words syndrome

Fuchs spot

Fuchs marginal keratitis

- --Peripheral nonulcerative keratitis
- --Produces progressive thinning, can result in...perforation
- --Resembles Terrien marginal degeneration

Fuchs endothelial dystrophy

Dalén - Fuchs nodule

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- --Seen in granulomatous conditions, esp...sympathetic ophthalmia

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--Craterlike openings in surface of the anterior iris

Fuchs coloboma
--aka...tilted disc syndrome

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Fuchs coloboma --aka...tilted disc syndrome --The ONH is elevated direction , depressed opposite direction

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Fuchs coloboma

- --aka...tilted disc syndrome
- -- The ONH is elevated **superotemporal**, depressed inferonasal

Fuchs spot

Ernst Fuchs (1851-1930)

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be mistaken for a ciliary- Why is the ONH tilted in this manner?

Fuchs coloboma

aka...tilted disc syndrome

--The ONH is elevated **superotemporal** depressed **inferonasal**

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be mistaken for a ciliary- Why is the ONH tilted in this manner?

Abnormal closure of the embryonic optic fissure at the optic-nerve/globe junction results in an oblique (read: tilted) orientation of the ONH. The abnormal closure also creates a modest staphyloma in the inferonasal region of the globe.

Fuchs crypts

--Craterlike openings in s

Ernst Fuchs (1851-1930)

Fuchs endothelial dystrophy

Fuchs coloboma

depressed inferonasal

aka...tilted disc syndrome

--The ONH is elevated superotemporal

Dalén - Fuchs nodule

- cells beneath the...RPE
- esp...sympathetic ophthalmia

Ernst Fuchs

(1851-1930)

102

Fuchs adenoma

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--Craterlike openings in surface of the anterior iris

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- -- Associated with...

two words

Fuchs spot

Fuchs marginal keratitis

- --Peripheral nonulcerative keratitis
- --Produces progressive thinning, can result in...perforation
- --Resembles Terrien marginal degeneration

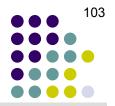
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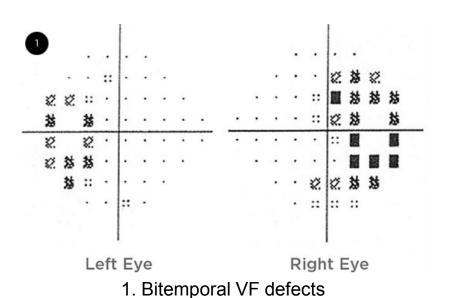
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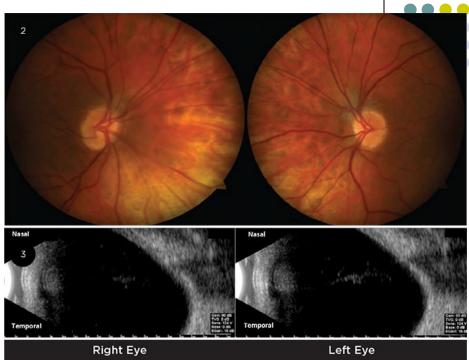
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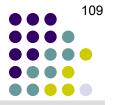
Herr Doktor Fuchs left quite an ophthalmic legacy. Can you identify these eponymous diseases and signs?





- 2. Note the tilted insertions of the optic nerves and diffuse atrophy of the nasal RPE
- 3. Ultrasound B-scan confirmed the oblique insertion of the optic nerves

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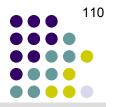
bitemporal superior loss

OK, but why on earth do these pts get a VF defect?

--Per --Pro result --Res

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inferior bitemporal superior loss

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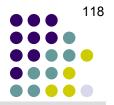
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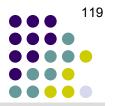
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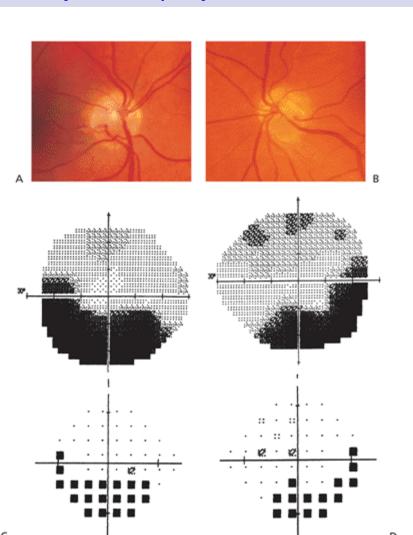
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Pretty much what you would expect based on the name--a normal-appearing nerve save for a thin superior rim, with associated thinning of the superior nerve fiber layer

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What is the classic causal association? (Hint: It concerns the status of the pt's mother.)

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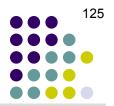
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What is the classic causal association? (Hint: It concerns the status of the pt's mother.) Maternal diabetes (It's one of the '4 Ds of optic-nerve hypoplasia')

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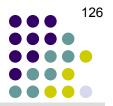
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Fut OK, but why o There is another optic-nerve condition that presents with nonprogressive --Crailt's actually pre bitemporal inferior loss that doesn't respect the vertical midline. What is it? As indicated Superior segmental optic nerve hypoplasia (SSONH) inferonasally is longer man it is in orner regions, because or mis extra axial length, the remacti correcti What is the appearance of the nerve head in SSONH2 staphyle What are the 4 Ds of optic-nerve hypoplasia? Pretty much what yo save for a thin super -- Diabetes Res This im Is SSONH an acquir Indeed Congenital --d This im What is the classic causal association? (Hint: It concerns the status of the pt's mother.)

Maternal diabetes (It's one of the '4 Ds of optic-nerve hypoplasia')

--Highly focal aggregate of inflammatory cells beneath the...**RPE**

--Seen in granulomatous conditions, esp...sympathetic ophthalmia



Fuchs adenoma

- --Very common (~25% of eyes post-mortem)
- --A small (<1mm) nodular hyperplasia of the nonpigmented epithelium of the ciliary body
- --Looks like a tiny **pearl onion** on the **pars plicata**
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-- Diabetes

- --Drink (ie, heavy maternal EtOH consumption during gestation)
- -- Drugs, especially Dilantin (ie, maternal use during gestation)
- --de Morsier syndrome

Res This im

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--Craterlike openings in surface of the anterior iris

Ernst Fuchs (1851-1930)

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Fuchs spot

--Area of RPE hyperplasia overlying regressed CNVM in two words

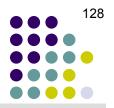
Fuchs marginal keratitis

- --Peripheral nonulcerative keratitis
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- --Resembles Terrien marginal degeneration

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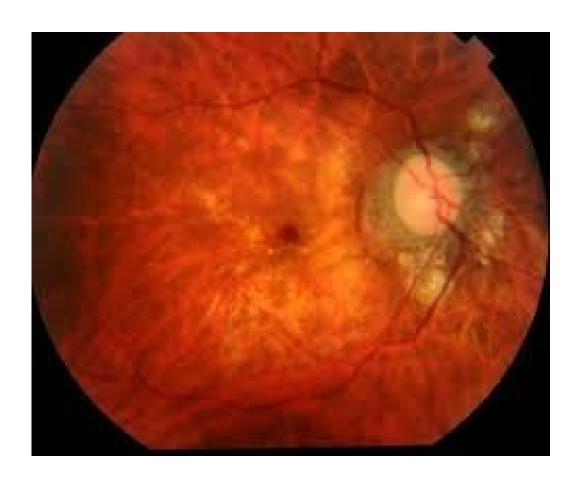
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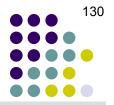
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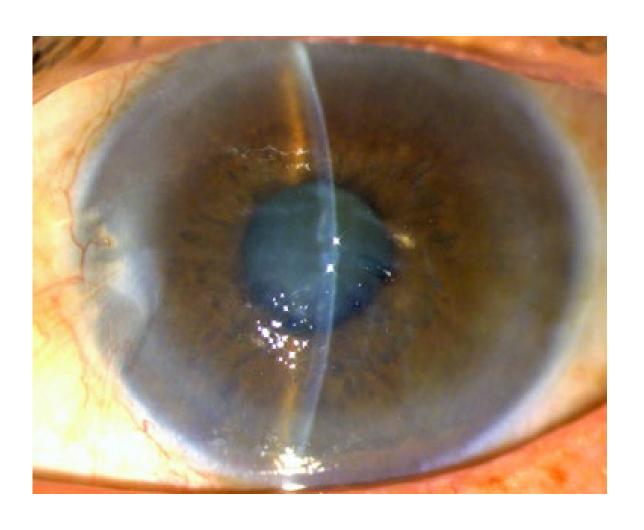
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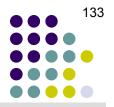
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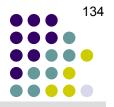
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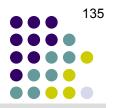
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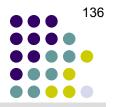
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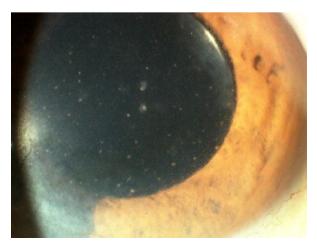




Online Journal of Ophthalmology www.onjoph.com

Heterochromia iridis

Unilateral cataract



Stellate KP



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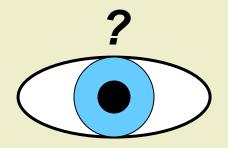
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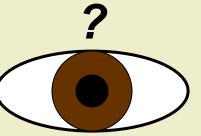
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rloss

Fuchs cryp

--Craterlike ope





plasia overlying pathologic myopia

Fuchs marg

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Is the affected eye the **darker** eye or the **lighter** eye?

trophy
um characterized
progressive

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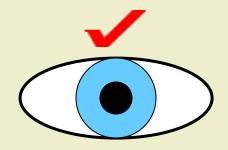
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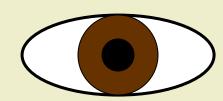
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- --Resembles Te

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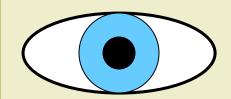
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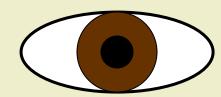
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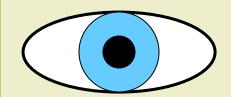
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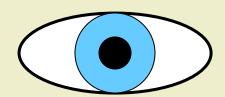
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- esp...sympathetic ophthalmia

- unilateral uveitis --Uncommon car
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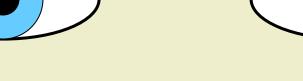
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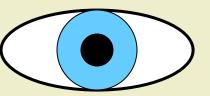
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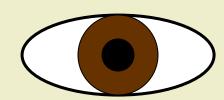
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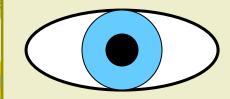
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FHI: 'Moth eaten' iris. Note the smooth stromal architecture and loss of iris crypts



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Fuchs spot

Besides being stellate, what else is unusual about the KP in FHI?

1)

2

Fuchs margina

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- -- Produces progres
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trophy m characterized progressive

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- 1) They can be interconnected via very fine tendrils or filaments
- 2) They are evenly distributed across the entire endothelium

piasia overiying **pathologic myopia**

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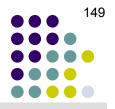
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- --aka...tilted disc syndrome
- --The ONH is elevated **superotemporal** , depressed **inferonasal**
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Besides being stellate, what else is unusual about the KP in FHI?

- 1) They can be interconnected via very fine tendrils or filaments
- 2) They are evenly distributed across the entire endothelium

How are KP usually distributed?
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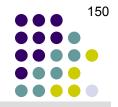
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Dalén - Fuch

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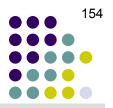
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- 2) Herpes zoster
- 3) CMV retinitis
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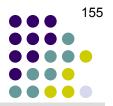
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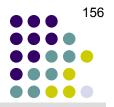
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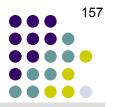
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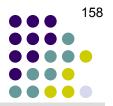
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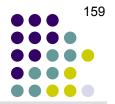
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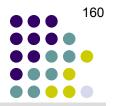
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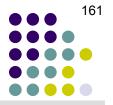
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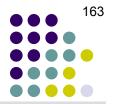
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770 docociated Williams



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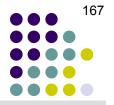
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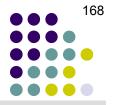
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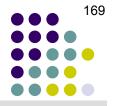
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--Craterlike openings in surface of the anterior iris

Ernst Fuchs (1851-1930)

Fuchs coloboma

- --aka...tilted disc syndrome
- --The ONH is elevated **superotemporal**, depressed **inferonasal**
- --Associated with...high myopia
- --Characteristic VF deficit is...
 bitemporal superior loss

Fuchs spot

 --Area of RPE hyperplasia overlying regressed CNVM in pathologic myopia

Fuchs marginal keratitis

- --Peripheral **nonulcerative** keratitis
- --Produces progressive thinning, can result in...perforation
- --Resembles Terrien marginal degeneration

Fuchs endothelial dystrophy

--Disease of corneal endothelium characterized by the presence of **guttae** and progressive **corneal edema**

Dalén - Fuchs nodule

- --Highly focal aggregate of inflammatory cells beneath the...**RPE**
- --Seen in granulomatous conditions, esp...sympathetic ophthalmia

While not confirmed (as of this writing), one possible cause for FHI is considered especially likely. What is it?

Chronic infection with the virus

--25-50% associated with secondary...glaucoma

-- Cause is unknown; is a diagnosis of exclusion



Fuchs adenoma

- --Very common (~25% of eyes post-mortem)
- --A small (<1mm) nodular hyperplasia of the **nonpigmented epithelium** of the **ciliary body**
- --Looks like a tiny pearl onion on the pars plicata
- --Benign, but can cause **sectoral cataract** and/or be mistaken for a ciliary-body **melanoma**

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While not confirmed (as of this writing), one possible cause for FHI is considered especially likely. What is it?

Chronic infection with the **rubella** virus

--25-50% associated with secondary...glaucoma

-- Cause is unknown; is a diagnosis of exclusion