Ernst Fuchs
(1851-1930)

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

Fuchs

Seen mainly by pathologists
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**Fuchs adenoma**

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*Seen every day at the slit lamp*

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Fuchs two words-itis

An uncommon cornea condition
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**Fuchs marginal keratitis**

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\textit{superficial} Fuchs marginal keratitis

\textbf{Ernst Fuchs (1851-1930)}

Note that it is also known (including in the \textit{Cornea} book) as Fuchs \textit{superficial} marginal keratitis
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- **Ernst Fuchs**
  - (1851-1930)

- An occasional DFE finding
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*Seen mainly by pathologists*

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**Dalén - Fuchs nodule**

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**Fuchs something-oma**
has an interesting VF finding

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**Fuchs**
- Of interest to the most subspecialties
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Fuchs heterochromic iridocyclitis
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Fuchs adenoma

What are the two noneponymous names for Fuchs adenoma?

- Pseudoadenomatous hyperplasia of the ciliary body
- Coronal adenoma

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

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

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

**Medulloepithelioma (aka diktyoma)**
- Rare
- Does not present post-mortem
- Presents in childhood
- Usually requires enucleation

**Fuchs coloboma**

**Fuchs endothelial dystrophy**

**Fuchs heterochromic iridocyclitis**

**Dalén - Fuchs nodule**

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- Presents in childhood
- Enucleation is usually required

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

Not exactly. While 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.

OK then, what is meant here by describing it as ‘not nearly as benign’ as a Fuchs adenoma?

It means that medulloepitheliomas tend to be locally very aggressive.

How ‘locally aggressive’ are we talking about here?

Aggressive enough to be fatal.
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Medulloepithelioma/diktyoma
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*Is it common, or rare?*

**Fuchs endothelial dystrophy**

**Fuchs heterochromic iridocyclitis**

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Is it common, or rare?
Rare

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Medulloepithelioma (aka diktyoma)

*Is it common, or rare?*
- Rare

*Does medulloepithelioma present post-mortem?*

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*Is it common, or rare?*
Rare

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

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- Looks like a tiny pearl onion on the pars plicata

There's another tumor of the nonpigmented epithelium of the ciliary body that is not nearly as benign as Fuchs adenoma. What is it?
Medulloepithelioma (aka diktyoma)

Is it common, or rare?
Rare

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

How does it present?

- Glaucoma
- Hyphema
- Sectoral cataract

**Fuchs coloboma**

**Fuchs endothelial dystrophy**

**Fuchs heterochromic iridocyclitis**

**Fuchs spot**

**Fuchs coloboma**

**Dalén - Fuchs nodule**
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As an iris mass

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No, it presents in childhood

*How does it present?*
As an iris mass along with one or more of the following:
- 
- 
- 

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**Fuchs coloboma**

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

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*How is it managed?*
Enucleation is usually required

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Fuchs crypts

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- Craterlike openings in surface of the anterior

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**Fuchs marginal keratitis**

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Ernst Fuchs (1851-1930)
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**Fuchs marginal keratitis**
--Peripheral non- vs ulcerative keratitis
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**Ernst Fuchs**
(1851-1930)

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--Craterlike openings in surface of the anterior **iris**

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

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-- Produces progressive thinning, can result in... **perforation**
-- Resembles **Eponym + word + word**

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**Ernst Fuchs**
(1851-1930)

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**Ernst Fuchs**
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**Ernst Fuchs** (1851-1930)

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

**Dalén - Fuchs nodule**

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Fuchs marginal is an inflammatory condition, whereas Terriens is noninflammatory
Fuchs endothelial dystrophy
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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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Dalén - Fuchs nodule
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--Seen in granulomatous conditions, esp...[bad dz]
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Dalen-Fuchs nodules
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Dalen-Fuchs Nodule

diffuse granulomatous inflammation
epithelioid histiocytes and giant cells
sympathetic uveitis (ophthalmia)

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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?
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- Craterlike openings in surface of the anterior iris

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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? Vogt-Koyanagi-Harada (VKH) dz. These two diagnoses are like salt-and-pepper shakers—they go everywhere together.
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Are Dalén-Fuchs nodules associated with VKH?
You betcha

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Cells beneath the...RPE
--Seen in granulomatous conditions, esp...sympathetic ophthalmia (SO)
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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?

Fuchs spot

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**Sarcoid**  
--Seen in granulomatous conditions, esp…sympathetic ophthalmia (SO)
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- Craterlike openings in surface of the anterior **iris**

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

**Fuchs coloboma**
- aka... **two words** syndrome

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

**Fuchs spot**

**Fuchs endothelial dystrophy**

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- aka... tilted disc syndrome

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

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**Is the myopia refractive, or axial?**
- Axial

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1. Bitemporal VF defects

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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Superior segmental optic nerve hypoplasia (SSONH)

What is the appearance of the nerve head in SSONH?

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

Is SSONH an acquire, or congenital condition?

Congenital

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

Maternal DM
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**Fuchs coloboma**

--aka...tilted disc syndrome
--The ONH is elevated superotemporal, depressed inferonasal
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This implies that the VF defect will resolve if the 'proper' refractive correction is employed. Does it?
- Indeed it does

This implies also that the bitemporal VF loss shouldn't respect the vertical midline. Does it?
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There is another optic-nerve condition that presents with nonprogressive bitemporal inferior loss that doesn't respect the vertical midline. What is it?
- Superior segmental optic nerve hypoplasia (SSONH)

What is the appearance of the nerve head in SSONH?
- 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

Is SSONH an acquire, or congenital condition?
- Congenital

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

**What are the 4 Ds of optic-nerve hypoplasia?**

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

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- **Heterochromia iridis**
- **Unilateral cataract**
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Is the affected eye the darker eye or the lighter eye?
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Fuchs coloboma
--Aka tilted disc syndrome
--The ONH is elevated superotemporal
--Associated with high myopia
--Depressed inferonasal plica overlying pathologic myopia

Fuchs coloboma
--Histologically, a cleft-like space in the iris
--Clinical feature is the affected eye the darker eye or the lighter eye?

What is the exception, i.e., under what circumstances is the darker eye the one with FHI?

The lighter eye with one exception

In individuals with light-blue eyes…iris atrophy stemming from the FHI process will make visible the darkly-pigmented epithelium of the posterior iris, thus making the eye appear darker

Ernst Fuchs (1851-1930)

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--Produces progressive thinning, can result in perforation
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--Area of RPE hyperplasia overlying regressed CNVM in pathologic myopia

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--Uncommon cause of chronic unilateral uveitis
--Classic triad: Heterochromia iridis, cataract, stellate KP
--In individuals with light-blue eyes...

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--Characteristic VF deficit is… bitemporal superior loss

**Fuchs spot**
--Area of RPE hyperplasia overlying regressed CNVM in pathologic myopia

**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?

**Fuchs coloboma**
--25-50% associated with secondary…*glaucoma*
--Cause is unknown; is a diagnosis of exclusion
Herr Doktor Fuchs left quite an ophthalmic legacy. Can you identify these eponymous diseases and signs?

**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

**Fuchs crypts**
- Craterlike openings in surface of the anterior iris

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

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**Ernst Fuchs (1851-1930)**

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