--Secondary to pathological process of central portion of location of path process
Lenticonus

--Secondary to "ectasia" of central portion of "lens surface"
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Lenticonus

When you hear the term oil droplet applied to the appearance of the lens, three conditions should come to mind. Two are anterior and posterior lenticonus—what is the third?

Galactosemia

What is galactosemia?

It is an inborn error of metabolism in which galactose cannot be converted to glucose.

How does this lead to an oil droplet lenticular appearance?

An inert byproduct of galactose metabolism (galactitol) accumulates in lens fibers, creating an osmotic gradient that draws in aqueous. Swelling of the fibers damages them, leading to the changes that produce an oil droplet.

So, there's a fundamental difference between the cause of the oil-droplet appearance in lenticonus (= increased central lenticular power) vs that of galactosemia (= the accumulation of galactitol and fluid in the central lens).

What are the systemic manifestations of galactosemia?

--Failure to thrive
--Hepatomegaly with jaundice
--Impaired cognitive development
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What is the long-term prognosis of galactosemia?

The severe form (which is also the most common) is uniformly fatal if not treated.

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What are the systemic manifestations of galactosemia?

--Failure to thrive
--Hepatomegaly with jaundice
--Impaired cognitive development
---Secondary to *ectasia* of central portion of *lens surface*

---Typical clinical course:

---Early: Manifests as *refractive issue*
Lenticonus

--Secondary to **ectasia** of central portion of lens surface

--Typical clinical course:
  --Early: Manifests as **myopia**
Lenticonus

--Secondary to **ectasia** of central portion of **lens surface**

--Typical clinical course:
  --Early: Manifests as **myopia**
  --Next: Manifests as different refractive issue
Lenticonus

--Secondary to **ectasia** of central portion of **lens surface**

--Typical clinical course:
  --Early: Manifests as **myopia**
  --Next: Manifests as **irregular astigmatism**
--Secondary to **ectasia** of central portion of **lens surface**
--Typical clinical course:
  --Early: Manifests as **myopia**
  --Next: Manifests as **irregular astigmatism**
  --Later: The **lens layer** opacifies
A

--Secondary to **ectasia** of central portion of **lens surface**
--Typical clinical course:
  --Early: Manifests as **myopia**
  --Next: Manifests as **irregular astigmatism**
  --Later: The **cortex** opacifies
Lenticonus

--Secondary to **ectasia** of central portion of **lens surface**

--Typical clinical course:
  --Early: Manifests as **myopia**
  --Next: Manifests as **irregular astigmatism**
  --Later: The **cortex** opacifies
  --At age **#.#** years: Capsule → **two words**
Lenticonus

--Secondary to **ectasia** of central portion of **lens surface**
--Typical clinical course:
  --Early: Manifests as **myopia**
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Lenticonus comes in two (very) basic forms—what are they?
--Secondary to ectasia of central portion of lens surface.
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  --At age 4-5 years: Capsule ruptures → total opacification

**Lenticonus comes in two (very) basic forms—what are they?**
**Anterior** lenticonus and **posterior** lenticonus, referring to involvement of the anterior and posterior capsules respectively.
Lenticonus

Anterior lenticonus
Lenticonus

Posterior lenticonus
Just for fun: An eye with both anterior *and* posterior lenticonus
Lenticonus

--Secondary to **ectasia** of central portion of **lens surface**
--Typical clinical course:
  --Early: Manifests as **myopia**
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### Lenticonus

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--- **Anterior lenticonus**
- Usually **bilateral**
- Less common

--- **Posterior lenticonus**
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**Lenticonus**

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--Secondary to *ectasia* of central portion of lens surface
--Typical clinical course:
  --Early: Manifests as *myopia*
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*Note that these factoids ‘go together,’ ie, syndromic conditions usually affect both eyes…*
--Secondary to **ectasia** of central portion of lens surface

--Typical clinical course:

--Early: Manifests as **myopia**

--Next: Manifests as **irregular astigmatism**

--Later: The **cortex** opacifies

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...and likewise, these two go together as well (sporadic conditions usually affect only one)

90% **unilateral**

More common (but still rare)

Usually **sporadic** (unless bilateral)
--Secondary to ectasia of central portion of lens surface

--Typical clinical course:

--Early: Manifests as myopia

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--Bilateral cases are associated with two of the familial oculorenal syndromes
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- Next: Manifests as *irregular astigmatism*
- Later: The *cortex* opacifies
- At age 4-5 years: Capsule ruptures → *total opacification*

**Bilateral** cases are associated with two of the *familial oculorenal* syndromes (Alport syndrome; Lowe syndrome)

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Anterior lenticonus in Alport syndrome
Lenticonus

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**How prevalent is lenticonus in Alport and Lowe syndromes?**

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How prevalent is lenticonus in Alport and Lowe syndromes?
Anterior lenticonus is not a major component of Alport syndrome, being present in only 25% of cases

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Bilateral cases can be associated with the familial oculorenal syndromes (e.g., Alport syndrome, Lowe syndrome).

**How prevalent is lenticonus in Alport and Lowe syndromes?**

Anterior lenticonus is not a major component of Alport syndrome, being present in only 25% of cases. In contrast, posterior lenticonus is a defining feature of Lowe syndrome.

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*Alport Syndrome* is X-linked with hematuria in childhood, high nerve deafness, and nephritis. *Lowe Syndrome* is also X-linked with hematuria in infancy, microsphero-phakia, MR, rickets, and hypotonia.
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Lenticonus
Lenticonus

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**For completeness’ sake:** *There are four more familial oculorenal syndromes mentioned in the BCSC books. All are the same type of disorder. What type are they?*
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*For completeness’ sake: There are four more familial oculorenal syndromes mentioned in the BCSC books. All are the same type of disorder. What type are they?** **Ciliopathy**
What is a ciliopathy?

A ciliopathy is an inherited condition marked by abnormal structure and/or function of cilia. Cilia are ubiquitous organelles, and ciliopathies primarily affect three organs:

- The eyes
- The brain
- The kidneys

For completeness’ sake: There are four more familial oculorenal syndromes mentioned in the BCSC books. All are the same type of disorder. What type are they? **Ciliopathy**
Lenticonus

What is a ciliopathy?
An inherited condition marked by abnormal structure and/or function of cilia

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**Lenticonus**

**What is a ciliopathy?**
An inherited condition marked by abnormal structure and/or function of cilia

*Cilia are ubiquitous organelles. That said, ciliopathies primarily affect three organs--what are they?*

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Cilia are ubiquitous organelles. That said, ciliopathies primarily affect three organs--what are they? The eyes, brain and kidneys

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What is a ciliopathy?
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Cilia are ubiquitous organelles. That said, ciliopathies primarily affect three organs—what are they?

The eyes??!! Which part of the eye contains cilia wiggling about?
None. Remember, cilia come in two basic flavors: Motile, and nonmotile. It is the nonmotile type which is ubiquitous in the eye.

Lenticonus

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**What is a ciliopathy?**
An inherited condition marked by abnormal structure and/or function of cilia.

Cilia are ubiquitous organelles. That said, ciliopathies primarily affect three organs:
- The eyes, brain
- The kidneys

**What are the four ciliopathies emphasized in the BCSC books?**
- Note: You have heard of at least several of these, but may not have thought of them as a group, i.e., as all being members of the *oculorenal syndrome* family. It's important that you make this connection!

**For completeness’ sake:** There are **four more** *familial oculorenal syndromes* mentioned in the BCSC books. All are the same type of disorder. What type are they? **Ciliopathy**

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An inherited condition marked by abnormal structure and/or function of cilia.

**Cilia are ubiquitous organelles.**
The eyes, brain, and kidneys are particularly affected.

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**What are the four ciliopathies emphasized in the BCSC books?**
- Joubert syndrome
- Alström syndrome
- Bardet-Biedl syndrome
- Senior-Løken syndrome

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**Bilateral cases are associated with two of the familial oculorenal syndromes**
(Alport syndrome; Lowe syndrome, and the...ciliopathies)

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**For completeness’ sake:** There are **four more** familial oculo-renal syndromes mentioned in the BCSC books. All are the same type of disorder. What type are they? **Ciliopathy**

---

**What is a ciliopathy?**

An inherited condition marked by abnormal structure and/or function of cilia.

Cilia are ubiquitous organelles. The eyes, brain, and kidneys are primarily affected by ciliopathies.

**What are the four ciliopathies emphasized in the BCSC books?**

- Joubert syndrome
- Alström syndrome
- Bardet-Biedl syndrome
- Senior-Løken syndrome

**Do you have a mnemonic for remembering the ciliopathies?**

Indeed I do--picture a cilia that JABS you in the eye.

---

**Cilia are ubiquitous organelles.**

- Bilateral cases are associated with two of the familial oculo-renal syndromes (**Alport** syndrome; **Lowe** syndrome, and the...ciliopathies)
Lenticonus

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Ciliopathy
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- Eyes
- Brain
- Kidneys

What are the four ciliopathies emphasized in the BCSC books?
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- Bilateral cases are associated with two of the familial oculorenal syndromes (Alport syndrome; Lowe syndrome, and the...ciliopathies)

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Cilia are ubiquitous organelles. That said, ciliopathies primarily affect three organs—what are they?

- The eyes
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What are the four ciliopathies emphasized in the BCSC books?
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Bilateral cases are associated with two of the familiar oculorenal syndromes (Alport syndrome; Lowe syndrome, and the...ciliopathies)

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Are lenticous or other lens findings a feature of the ciliopathies?
### What is a ciliopathy?
An inherited condition marked by abnormal structure and/or function of cilia.

### Cilia are ubiquitous organelles.
The eyes, brain, and kidneys are primarily affected by ciliopathies.

#### What are the four ciliopathies emphasized in the BCSC books?
- Joubert syndrome
- Alström syndrome
- Bardet-Biedl syndrome
- Senior-Løken syndrome

#### Bilateral cases are associated with two of the familial oculorenal syndromes (Alport syndrome; Lowe syndrome, and the...ciliopathies)

### Alport Syndrome
- Location: Anterior
- Inheritance: Most are X-linked
- Classic presentation (nonocular): Hematuria in childhood
- Associated findings: High-f nerve deafness; nephritis
- Another lens finding: Microsphero-phakia

### Lowe Syndrome
- Location: Posterior
- Inheritance: X-linked
- Classic presentation (nonocular): Hematuria in infancy
- Associated findings: MR; rickets; hypotonia
- Another lens finding: Microsphero-phakia

### Ciliopathies
- Location: None
- Inheritance: None
- Classic presentation (nonocular): None
- Associated findings: No--unlike Alport and Lowe syndromes, ciliopathies are not associated with lens abnormalities
- Another lens finding: None

---

**Lenticonus**

- **Location**: Inheritance Classic
- **Presentation**: Associated findings: Another lens finding

---

**What is a ciliopathy?**
An inherited condition marked by abnormal structure and/or function of cilia.

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The eyes, brain, and kidneys.

**What are the four ciliopathies emphasized in the BCSC books?**
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**Unlike the X-linked Alport and Lowe syndromes, ciliopathies are inherited AR.**

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- The eyes, brain
- The kidneys

What are the four ciliopathies emphasized in the BCSC books?
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Bilateral cases are associated with two of the familial oculorenal syndromes (Alport syndrome; Lowe syndrome, and the...ciliopathies)

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An inherited condition marked by abnormal structure and/or function of cilia. 

### Cilia are ubiquitous
The eyes, brain, and kidneys, et cetera.

#### What are the four ciliopathies emphasized in the BCSC books?---what are they?
- Joubert syndrome
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#### Bilateral cases are associated with two of the familial oculorenal syndromes (Alport syndrome; Lowe syndrome, and the…ciliopathies)

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**Lenticonus**

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Pigmentary retinopathy in Bardet-Biedl syndrome
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The eyes, brain, kidneys

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**Lenticonus**

- Secondary to ectasia of central portion of lens surface
- Typical clinical course:
  - Early: Manifests as myopia
  - Next: Manifests as irregular astigmatism
  - Later: The cortex opacifies
  - At age 4-5 years: Capsule ruptures → total opacification

- Bilateral cases are associated with two of the familial oculorenal syndromes (Alport syndrome; Lowe syndrome, and the...ciliopathies)

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The eyes, brain, and kidneys are affected by ciliopathies.

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**What are the four ciliopathies emphasized in the BCSC books?**
- Joubert syndrome
- Alström syndrome
- Bardet-Biedl syndrome
- Senior-Løken syndrome

---

**Alport Syndrome**
- Anterior
- Most are X-linked
- Hematuria in childhood
- High-f frequency nerve deafness
- Microspherophakia

---

**Lowe Syndrome**
- Posterior
- X-linked
- Hematuria in infancy
- MR; rickets; hypotonia
- Microspherophakia

---

**Ciliopathies**
- None
- AR
- No hematuria
- Pigmentary retinopathy with flat ERG
- None

---

Leber’s congenital amaurosis is an age-related variant of retinitis pigmentosa.

---

‘Pigmentary retinopathy with flat ERG in an infant’ sounds like what disease?

Leber’s congenital amaurosis. Therefore, before a child is diagnosed with LCA, one must consider the diagnosis of a ciliopathy.
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Leber’s congenital amaurosis is an age-related variant of... **retinitis pigmentosa**

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‘Pigmentary retinopathy with flat ERG in an infant’ sounds like what disease? Leber’s congenital amaurosis. Therefore, before a child is diagnosed with LCA, one must consider the diagnosis of a ciliopathy.
Familial Oculorenal Syndromes *tl;dr*

- One sort
- The other sort
Familial Oculorenal Syndromes

tl;dr

Ciliopathies

Not Ciliopathies
Familial Oculorenal Syndromes *tl;dr*

Lenticonus

Ciliopathies

Not Ciliopathies

Q
Familial Oculorenal Syndromes *tl;dr*

- Ciliopathies
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  - Alström syndrome
  - Bardet-Biedl syndrome
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- **Not Ciliopathies**
  - Alport syndrome
  - Lowe syndrome

**Key Features**

- Renal failure
- Hematuria
Familial Oculorenal Syndromes *tl;dr*

**Ciliopathies**
- Joubert syndrome
- Alström syndrome
- Bardet-Biedl syndrome
- Senior-Løken syndrome

**Not Ciliopathies**
- Alport syndrome
- Lowe syndrome

**Renal failure**
- *without* hematuria
- *with* hematuria

**Key Features**

**Lenticonus**
Familial Oculorenal Syndromes *tl;dr*

- **Ciliopathies**
  - Joubert syndrome
  - Alström syndrome
  - Bardet-Biedl syndrome
  - Senior-Løken syndrome

- **Not Ciliopathies**
  - Alport syndrome
  - Lowe syndrome

**Key Features**

- Renal failure *without* hematuria
  - Classic eye finding: [Lenticonus](#)

- Renal failure *with* hematuria
  - Classic eye finding:
Familial Oculorenal Syndromes

Ciliopathies
- Joubert syndrome
- Alström syndrome
- Bardet-Biedl syndrome
- Senior-Løken syndrome

Not Ciliopathies
- Alport syndrome
- Lowe syndrome

Renal failure

without hematuria

Classic eye finding:
Pigmentary retinopathy

Renal failure

with hematuria

Classic eye finding:
Lenticonus
Familial Oculoorenal Syndromes

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- Alport syndrome
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Renal failure
- without hematuria
- Renal failure with hematuria

Classic eye finding:
- Pigmentary retinopathy
- Lenticonus

Inheritance:

Key Features
Familial Oculorenal Syndromes

Ciliopathies
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- Bardet-Biedl syndrome
- Senior-Løken syndrome
- Alport syndrome
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Renal failure without hematuria
Classic eye finding: Pigmentary retinopathy
Inheritance: AR

Not Ciliopathies
- Alport syndrome
- Lowe syndrome

Renal failure with hematuria
Classic eye finding: Lenticonus
Inheritance: X-linked

Key Features