Re microspherophakia…which of the following are true?

- Due to faulty development of 2° lens fibers
Re microspherophakia…which of the following are true?

- Due to faulty development of 2° lens fibers  T
Re microspherophakia…which of the following are true?

- Due to faulty development of $2^\circ$ lens fibers  \( T \)
- Associated with high hyperopia
Re microspherophakia…which of the following are true?

- Due to faulty development of 2° lens fibers  **T**
- Associated with high hyperopia  **F**
Re microspherophakia...which of the following are true?

- Due to faulty development of $2^\circ$ lens fibers  **T**
- Associated with high hyperopia  **F**
Re microspherophakia…which of the following are true?
- Due to faulty development of 2° lens fibers  \(\text{T}\)
- Associated with high hyperopia  \(\text{F}\)

Does microspherophakia actually cause the high myopia with which it is associated?
Re microspherophakia…which of the following are true?

- Due to faulty development of 2° lens fibers  **T**
- Associated with high hyperopia  **F**

Does microspherophakia actually cause the high myopia with which it is associated? **Yes**
Re microspherophakia...which of the following are true?

- Due to faulty development of 2° lens fibers  T
- Associated with high hyperopia  F T

Does microspherophakia actually cause the high myopia with which it is associated?  Yes

How does it cause high myopia?
Re microspherophakia…which of the following are true?

- Due to faulty development of 2° lens fibers  
- Associated with high hyperopia  
  
Does microspherophakia actually cause the high myopia with which it is associated? Yes

How does it cause high myopia?
The surface of the spherical lens is far more curved than that of a normal lens, and thus possesses significantly more converging power.
Re microspherophakia...which of the following are true?

- Due to faulty development of $2^\circ$ lens fibers  \( \text{T} \)
- Associated with high myopia  \( \text{T} \) does microspherophakia actually cause the high myopia with which it is associated?

- Yes

How does it cause high myopia?
The surface of the spherical lens is far more curved than that of a normal lens, and thus possesses significantly more converging power

How does this differ from 'run of the mill' high myopia?
Re microspherophakia…which of the following are true?
- Due to faulty development of 2° lens fibers  ✓
- Associated with high hyperopia  ❌

Does microspherophakia actually cause the high myopia with which it is associated? Yes

How does it cause high myopia?
The surface of the spherical lens is far more curved than that of a normal lens, and thus possesses significantly more converging power.

How does this differ from ‘run of the mill’ high myopia?
Most cases of high myopia are due to excessive length of the optical axis (so-called ‘axial myopia’).
Q

Re microspherophakia...which of the following are true?

- Due to faulty development of 2° lens fibers  \( T \)
- Associated with high hyperopia  \( F \)
- Can cause pupillary block with subsequent angle closure glaucoma  \( T \)

Microspherophakia
Re microspherophakia…which of the following are true?

- Due to faulty development of 2° lens fibers \( T \)
- Associated with high hyperopia \( F \)
- Can cause pupillary block with subsequent angle closure glaucoma \( T \)
Microspherophakia

Re microspherophakia...which of the following are true?

- Due to faulty development of 2° lens fibers  T
- Associated with high hyperopia  F T
- Can cause pupillary block with subsequent angle closure glaucoma  T

What is the mechanism by which microspherophakia can lead to pupillary block and subsequent angle-closure glaucoma?
Re microspherophakia…which of the following are true?

- Due to faulty development of $2^\circ$ lens fibers  **T**
- Associated with high hyperopia  **F**  **T**
- Can cause pupillary block with subsequent angle closure glaucoma  **T**

What is the mechanism by which microspherophakia can lead to pupillary block and subsequent angle-closure glaucoma?

If zonular laxity is present, the lens may be able to drift far enough forward to block the pupillary aperture, leading to acute angle closure.
Re microspherophakia...which of the following are true?

- Due to faulty development of 2° lens fibers **T**
- Associated with high hyperopia **F**
- Can cause pupillary block with subsequent angle closure glaucoma **T**
- Angle closure can be successfully prophylaxed with miotics **T**
Re microspherophakia…which of the following are true?

- Due to faulty development of $2^\circ$ lens fibers **T**
- Associated with high hyperopia **F**
- Can cause pupillary block with subsequent angle closure glaucoma **T**
- Angle closure can be successfully prophylaxed with miotics **F**
Re microspherophakia…which of the following are true?

- Due to faulty development of 2° lens fibers **T**
- Associated with high hyperopia **F**
- Can cause pupillary block with subsequent angle closure glaucoma **T**
- Angle closure can be successfully prophylaxed with miotics **F**
Q

Re microspherophakia…which of the following are true?

- Due to faulty development of 2º lens fibers **T**
- Associated with high hyperopia **F T**
- Can cause pupillary block with subsequent angle closure glaucoma **T**
- Angle closure can be successfully prophylaxed with miotics **F T**

What surgical maneuvers are sometimes used to prophylax against angle closure?
Re microspherophakia...which of the following are true?

- Due to faulty development of 2° lens fibers  **True**
- Associated with high hyperopia  **False**
- Can cause pupillary block with subsequent angle closure glaucoma  **True**
- Angle closure can be successfully prophylaxed with miotics  **False**

What surgical maneuvers are sometimes used to prophylax against angle closure? Iridotomy, or lensectomy
Re microspherophakia...which of the following are true?

- Due to faulty development of 2° lens fibers  **T**
- Associated with high hyperopia  **F**
- Can cause pupillary block with subsequent angle closure glaucoma  **T**
- Angle closure can be successfully prophylaxed with miotics  **F** **T**

What surgical maneuvers are sometimes used to prophylax against angle closure?

Iridotomies or lensectomy

Some surgeons argue that two iridotomies 180° apart should be created to preclude pupillary blockage by a subluxed microspherophakic lens!
Re microspherophakia...which of the following are true?

- Due to faulty development of 2° lens fibers  T
- Associated with high hyperopia  F T
- Can cause pupillary block with subsequent angle closure glaucoma  T
- Angle closure can be successfully prophylaxed with miotics  F T
- Cycloplegics should be avoided, as they can close an already crowded angle  T
Re microspherophakia...which of the following are true?

- Due to faulty development of $2^\circ$ lens fibers $\sqrt{T}$
- Associated with high hyperopia $\sqrt{F}$
- Can cause pupillary block with subsequent angle closure glaucoma $\sqrt{T}$
- Angle closure can be successfully prophylaxed with miotics $\sqrt{F}$
- Cycloplegics should be avoided, as they can close an already crowded angle $\sqrt{F}$
Re microspherophakia…which of the following are true?

- Due to faulty development of $2^\circ$ lens fibers  **T**
- Associated with high hyperopia  **F**
- Can cause pupillary block with subsequent angle closure glaucoma  **T**
- Angle closure can be successfully prophylaxed with miotics  **F**
- Cycloplegics should be avoided, as they can close an already crowded angle  **F**
Re microspherophakia...which of the following are true?

- Due to faulty development of 2° lens fibers, T
- Associated with high hyperopia, F T
- Can cause pupillary block with subsequent angle closure glaucoma, T
- Angle closure can be successfully prophylaxed with miotics, F T
- Cycloplegics should be avoided, as they can close an already crowded angle, F T

Pilo is used to manage pupillary-block angle-closure glaucoma. Why shouldn’t it be used in cases secondary to microspherophakia, and why is cycloplegia employed therein?
Re microspherophakia…which of the following are true?

- Due to faulty development of 2° lens fibers  **T**
- Associated with high hyperopia  **F**
- Can cause pupillary block with subsequent angle closure glaucoma  **T**
- Angle closure can be successfully prophylaxed with miotics  **F**
- Cycloplegics should be avoided, as they can close an already crowded angle  **F**

Pilo is used to manage pupillary-block angle-closure glaucoma. Why shouldn’t it be used in cases secondary to microspherophakia, and why is cycloplegia employed therein?

Pilo will cause the lens to move farther forward, and will likely worsen the pupillary block. Cycloplegics will pull the lens posteriorly, away from the pupil.
Re microspherophakia…which of the following are true?

- Due to faulty development of 2° lens fibers: **T**
- Associated with high hyperopia: **F**
- Can cause pupillary block with subsequent angle closure glaucoma: **T**
- Angle closure can be successfully prophylaxed with miotics: **F**
- Cycloplegics should be avoided, as they can close an already crowded angle: **F**
- Strongly associated with Marfan syndrome: **T**
Re microspherophakia...which of the following are true?

- Due to faulty development of 2° lens fibers  \( \text{T} \)
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- Strongly associated with Marfan syndrome  \( \text{F} \)
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- Angle closure can be successfully prophylaxed with miotics  F  T
- Cycloplegics should be avoided, as they can close an already crowded angle  F  T
- Strongly associated with Marfan syndrome  F  T

What are the findings in Weill-Marchesani?

Patients with Weill-Marchesani have:
- short stature
Re microspherophakia...which of the following are true?

- Due to faulty development of 2° lens fibers T
- Associated with high hyperopia F T
- Can cause pupillary block with subsequent angle closure glaucoma T
- Angle closure can be successfully prophylaxed with miotics F T
- Cycloplegics should be avoided, as they can close an already crowded angle F T
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What are the findings in Weill-Marchesani?

Patients with Weill-Marchesani have:

- short stature

Weill-Marchesani syndrome
Re microspherophakia...which of the following are true?

- Due to faulty development of 2º lens fibers \( \text{T} \)
- Associated with high hyperopia \( \text{F} \)
- Can cause pupillary block with subsequent angle closure glaucoma \( \text{T} \)
- Angle closure can be successfully prophylaxed with miotics \( \text{F} \)
- Cycloplegics should be avoided, as they can close an already crowded angle \( \text{F} \)
- Strongly associated with Marfan syndrome \( \text{T} \)

What are the findings in Weill-Marchesani?

Patients with Weill-Marchesani have:

- Short stature
- Short fingers
- Stiff joints

(Think of it as the opposite of Marfan syndrome)
Re microspherophakia…which of the following are true?

- Due to faulty development of 2º lens fibers **T**
- Associated with high hyperopia **F**
- Can cause pupillary block with subsequent angle closure glaucoma **T**
- Angle closure can be successfully prophylaxed with miotics **F**
- Cycloplegics should be avoided, as they can close an already crowded angle **F**
- Strongly associated with Marfan syndrome **F**

*What are the findings in Weill-Marchesani?*

Patients with Weill-Marchesani have:

- ...short stature
- ...short fingers

*Weill-Marchesani syndrome*
Q

Re microspherophakia…which of the following are true?

- Due to faulty development of 2° lens fibers  \( \text{T} \)
- Associated with high hyperopia  \( \text{F} \)
- Can cause pupillary block with subsequent angle closure glaucoma  \( \text{T} \)
- Angle closure can be successfully prophylaxed with miotics  \( \text{F} \)
- Cycloplegics should be avoided, as they can close an already crowded angle  \( \text{F} \)
- Strongly associated with Marfan syndrome  \( \text{F} \)

What are the findings in Weill-Marchesani?

Patients with Weill-Marchesani have:

- \( \text{\textbf{...short}} \) stature
- \( \text{\textbf{...short}} \) fingers
- \( \text{\textbf{...short}} \) joints

Weill-Marchesani syndrome
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- Due to faulty development of 2° lens fibers **T**
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What are the findings in Weill-Marchesani?

Patients with Weill-Marchesani have:

...short stature
...short fingers
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Weill-Marchesani syndrome
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What are the findings in Weill-Marchesani?
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- **short** stature
- **short** fingers
- **stiff** joints

(Think of it as the opposite of Marfan syndrome)
Re microspherophakia…which of the following are true?

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- Cycloplegics should be avoided, as they can close an already crowded angle F T
- Strongly associated with Marfan syndrome T

What are the findings in Weill-Marchesani?

Patients with Weill-Marchesani have:

- short stature (Tall stature)
- short fingers (Long fingers)
- stiff joints (Lax joints)

(Think of it as the opposite of Marfan syndrome)
Microspherophakia

Re microspherophakia...which of the following are true?

- Due to faulty development of 2° lens fibers: **True**
- Associated with high hyperopia: **False**
- Can cause pupillary block with subsequent angle closure glaucoma: **True**
- Angle closure can be successfully prophylaxed with miotics: **False**
- Cycloplegics should be avoided, as they can close an already crowded angle: **True**
- Strongly associated with Marfan syndrome: **False**

What is the formal term for:
- Abnormally short fingers: **Brachydactyly**
- Abnormally long fingers: **Arachnodactyly**

What are the findings in Weill-Marchesani?
- Patients with Weill-Marchesani have:
  - Short stature
  - Short fingers
  - Stiff joints

(Think of it as the opposite of Marfan syndrome)
Re microspherophakia…which of the following are true?

- Due to faulty development of 2° lens fibers  **T**
- Associated with high hyperopia  **F**  **T**
- Can cause pupillary block with subsequent angle closure glaucoma  **T**
- Angle closure can be successfully prophylaxed with miotics  **F**  **T**
- Cycloplegics should be avoided, as they can close an already crowded angle  **F**  **T**
- Strongly associated with Marfan syndrome  **F**  **T**

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- Short stature (Tall stature)
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Weill-Marchesani syndrome
Re microspherophakia…which of the following are true?

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- Associated with high hyperopia  **F**
- Can cause pupillary block with subsequent angle closure glaucoma  **T**
- Angle closure can be successfully prophylaxed with miotics  **F**  **T**
- Cycloplegics should be avoided, as they can close an already crowded angle  **F**  **T**
- Strongly associated with Marfan syndrome  **F**  **T**

Weill-Marchesani is strongly associated with microspherophakia. With what conditions is microspherophakia occasionally associated?
Re microspherophakia…which of the following are true?

- Due to faulty development of $2^\circ$ lens fibers $\text{T}$
- Associated with high hyperopia $\text{F}$ $\text{T}$
- Can cause pupillary block with subsequent angle closure glaucoma $\text{T}$
- Angle closure can be successfully prophylaxed with miotics $\text{F}$ $\text{T}$
- Cycloplegics should be avoided, as they can close an already crowded angle $\text{F}$ $\text{T}$
- Strongly associated with Marfan syndrome $\text{F}$ $\text{T}$

Weill-Marchesani is strongly associated with microspherophakia. With what conditions is microspherophakia occasionally associated? Lowe syndrome, Alport syndrome, Marfan syndrome, Peters anomaly and congenital rubella.
Re microspherophakia…which of the following are true?

- Due to faulty development of 2° lens fibers  **T**
- Associated with high hyperopia  **F**
- Associated with high myopia  **T**
- Can cause pupillary block with subsequent angle closure glaucoma  **T**
- Angle closure can be successfully prophylaxed with miotics  **F**
- Cycloplegics should be avoided, as they can close an already crowded angle  **F**
- Strongly associated with Marfan syndrome  **F**

**Ruby LAMP** is a mnemonic for the other conditions associated with microspherophakia:

- **Ruby** = Rubella
- **Lowe syndrome**
- **Alport syndrome**
- **Marfan syndrome**
- **Peters anomaly**

Weill-Marchesani is strongly associated with microspherophakia. With what conditions is microspherophakia **occasionally** associated?

Lowe syndrome, Alport syndrome, Marfan syndrome, Peters anomaly and congenital rubella
Re microspherophakia...which of the following are true?

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- Associated with high hyperopia **F**
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- Strongly associated with Marfan syndrome **F**

Ruby LAMP is a mnemonic for the other conditions associated with microspherophakia:

- Ruby = Rubella
- Lowe syndrome
- Alport syndrome
- Marfan syndrome
- Peters anomaly

In three words (including syndrome), what are Lowe and Alport syndromes?

- Familial oculorenal syndromes
- Hematuria

Weill-Marchesani is strongly associated with microspherophakia. With what conditions is microspherophakia occasionally associated?

- Lowe syndrome, Alport syndrome
- Marfan syndrome, Peters anomaly and congenital rubella
Re microspherophakia...which of the following are true?

- Due to faulty development of 2° lens fibers \( \text{T} \)
- Associated with high hyperopia \( \text{F} \) \( \text{T} \)
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- Cycloplegics should be avoided, as they can close an already crowded angle \( \text{F} \)
- Strongly associated with Marfan syndrome \( \text{F} \)

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Ruby = Rubella

Lowe syndrome, Alport syndrome

Marfan syndrome

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In three words (including syndrome), what are Lowe and Alport syndromes?

Familial oculorenal syndromes

Weill-Marchesani is strongly associated with microspherophakia. With what conditions is microspherophakia occasionally associated?

Lowe syndrome, Alport syndrome, Marfan syndrome, Peters anomaly and congenital rubella
Re microspherophakia...which of the following are true?

- Due to faulty development of 2° lens fibers  
  - True
- Associated with high hyperopia  
  - False
- Can cause pupillary block with subsequent angle closure glaucoma  
  - True
- Angle closure can be successfully prophylaxed with miotics  
  - False
- Cycloplegics should be avoided, as they can close an already crowded angle  
  - False
- Strongly associated with Marfan syndrome  
  - False

Microspherophakia

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- Rubella
- Lowe syndrome
- Alport syndrome
- Marfan syndrome
- Peters anomaly

In three words (including syndrome), what are Lowe and Alport syndromes?

- Familial oculorenal syndromes

What is their classic (nonocular) presenting sign?

- Hematuria

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Lowe syndrome, Alport syndrome
Marfan syndrome, Peters anomaly and congenital rubella.
Re microspherophakia… which of the following are true?

- Due to faulty development of 2° lens fibers T
- Associated with high hyperopia F T
- Can cause pupillary block with subsequent angle closure glaucoma T
- Angle closure can be successfully prophylaxed with miotics F
- Cycloplegics should be avoided, as they can close an already crowded angle F
- Strongly associated with Marfan syndrome F

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- Ruby = Rubella
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In three words (including syndrome), what are Lowe and Alport syndromes?
Familial oculorenal syndromes

What is their classic (nonocular) presenting sign?
Hematuria

Microspherophakia is not the classic lens finding in the oculorenal syndromes (and should not be the first one out of your mouth if pimped about them). What is?

Weill-Marchesani is strongly associated with microspherophakia. With what conditions is microspherophakia occasionally associated?
- Lowe syndrome, Alport syndrome
Microspherophakia

Re microspherophakia…which of the following are true?

- Due to faulty development of 2ⁿ lens fibers  ✔
- Associated with high hyperopia  ✗
- Can cause pupillary block with subsequent angle closure glaucoma  ✔
- Angle closure can be successfully prophylaxed with miotics  ✗
- Cycloplegics should be avoided, as they can close an already crowded angle  ✗
- Strongly associated with Marfan syndrome  ✗

Ruby LAMP is a mnemonic for the other conditions associated with microspherophakia:
- Ruby = Rubella
- Lowe syndrome
- Alport syndrome
- Marfan syndrome
- Peters anomaly

In three words (including syndrome), what are Lowe and Alport syndromes?
Familial oculorenal syndromes

What is their classic (nonocular) presenting sign?
Hematuria

Microspherophakia is not the classic lens finding in the oculorenal syndromes (and should not be the first one out of your mouth if pimped about them). What is?
Lenticonus

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Lowe syndrome, Alport syndrome, Marfan syndrome, Peters anomaly and congenital rubella.
Re microspherophakia…which of the following are true?

- Due to faulty development of $2^\circ$ lens fibers **T**
- Associated with high hyperopia **F**
- Can cause pupillary block with subsequent angle closure glaucoma **T**
- Angle closure can be successfully prophylaxed with miotics **F**
- Cycloplegics should be avoided, as they can close an already crowded angle **T**
- Strongly associated with Marfan syndrome **F**
- Occurs as part of *ectopia lentis et pupillae* **T**
Re microspherophakia…which of the following are true?

- Due to faulty development of $2^\circ$ lens fibers
- Associated with high hyperopia
- Can cause pupillary block with subsequent angle closure glaucoma
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Re microspherophakia...which of the following are true?

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- Associated with high hyperopia
- Can cause pupillary block with subsequent angle closure glaucoma
- Angle closure can be successfully prophylaxed with miotics
- Cycloplegics should be avoided, as they can close an already crowded angle
- Strongly associated with Marfan syndrome
- Occurs as part of ectopia lentis et pupillae

Microspherophakia

What is ectopia lentis et pupillae?

- A genetic condition the hallmark of which is the displacement of the pupil (microspherophakic) lens
- It is very rare
- Bilateral
- In opposite directions—pupils inferotemporal, lenses superonasal
- The pupils typically have two further abnormalities—what are they?
  - Are very miotic, and dilate poorly
  - They are slit-like in shape
Re microspherophakia...which of the following are true?

- Due to faulty development of lens fibers
- Associated with high hyperopia
- Can cause pupillary block with subsequent angle closure glaucoma
- Angle closure can be successfully prophylaxed with miotics
- Cycloplegics should be avoided, as they can close an already crowded angle
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Microspherophakia

*What is ectopia lentis et pupillae?*

A genetic condition the hallmark of which is the displacement of the pupil and (microspherophakic) lens

Microspherophakia

**A**
Re microspherophakia...which of the following are true?

- Due to faulty development of lens fibers  
  T
- Associated with high hyperopia  
  F
- Can cause pupillary block with subsequent angle closure glaucoma  
  T
- Angle closure can be successfully prophylaxed with miotics  
  F
- Cycloplegics should be avoided, as they can close an already crowded angle  
  F
- Strongly associated with Marfan syndrome  
  F
- Occurs as part of ectopia lentis et pupillae  T

Microspherophakia

What is ectopia lentis et pupillae?
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How common is it?
Re microspherophakia…which of the following are true?

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A genetic condition the hallmark of which is the displacement of the pupil and (microspherophakic) lens

How common is it?
It is very rare

Is it unilateral, or bilateral?
Bilateral
Re microspherophakia…which of the following are true?

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A genetic condition the hallmark of which is the displacement of the pupil and (microspherophakic) lens

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It is very rare

Is it unilateral, or bilateral?
Bilateral
Re microspherophakia…which of the following are true?

- Due to faulty development of lens fibers
- Associated with high hyperopia
- Can cause pupillary block with subsequent angle closure glaucoma
- Angle closure can be successfully prophylaxed with miotics
- Cycloplegics should be avoided, as they can close already crowded angle
- Strongly associated with Marfan syndrome
- Occurs as part of ectopia lentis et pupillae

What is ectopia lentis et pupillae?
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Bilateral

In what direction are the pupils and lenses displaced?
- The pupils are miotic, and dilate poorly
- They are slit-like in shape

Microspherophakia
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  - True

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  - False

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