

# Q

## Microspherophakia

- *Re microspherophakia...which of the following are true?*
  - Due to faulty development of 2ndry lens fibers

# A

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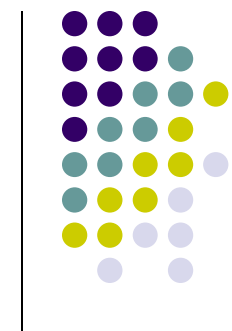
T



# Q

## Microspherophakia

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

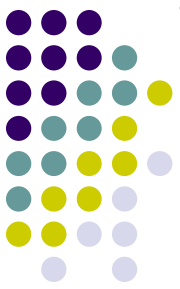


T

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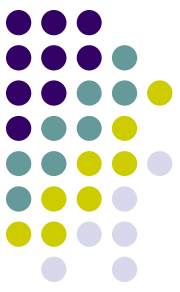
- *Re microspherophakia...which of the following are true?*
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## Microspherophakia

- *Re microspherophakia...which of the following are true?*
  - Due to faulty development of 2ndry lens fibers **T**
  - Associated with high <sup>myopia</sup>~~hyperopia~~ **F T**





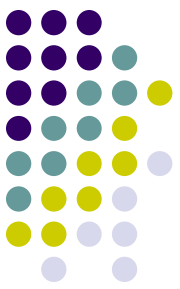
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## Microspherophakia

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

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- Associated with **high** <sup>myopia</sup> hyperopia F T

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



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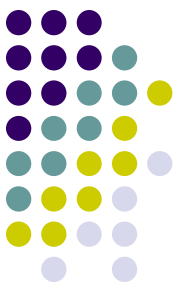
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Yes



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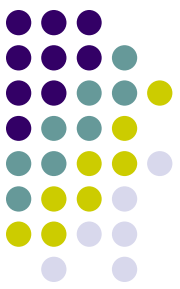
- Due to faulty development of 2ndry lens fibers **T**
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*Does microspherophakia actually cause the high myopia with which it is associated?*

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*How does it cause high myopia?*





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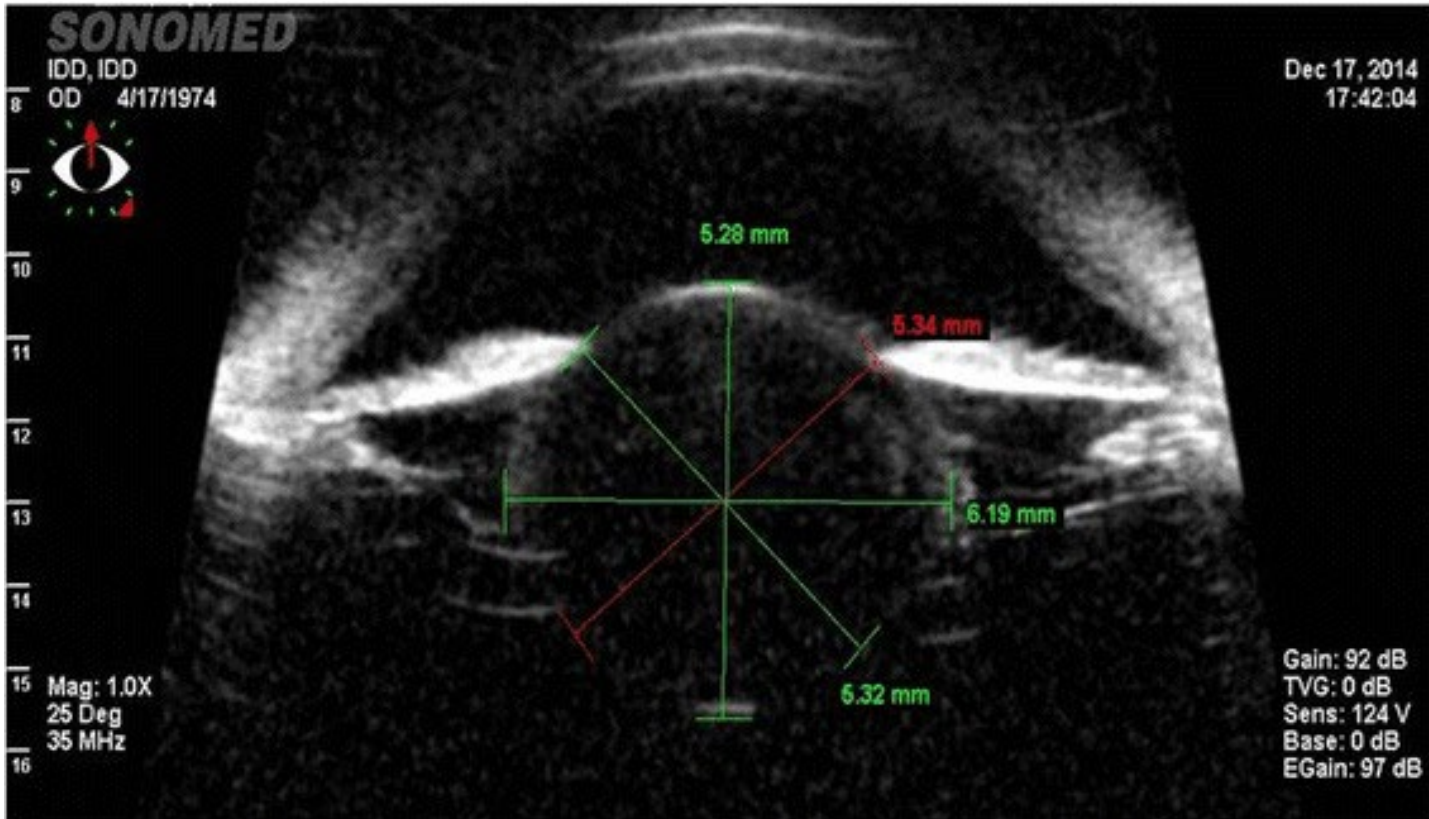
*Does microspherophakia actually cause the high myopia with which it is associated?*

Yes

*How does it cause high myopia?*

The surface of the spherical lens is much more curved than that of a normal lens, and thus possesses significantly more converging power

# Microspherophakia



Microspherophakia. Note the extreme curvature of the lens



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*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 much 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?*



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

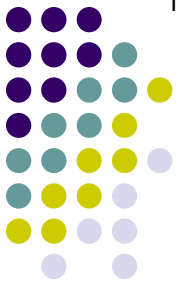
Yes

*How does it cause high myopia?*

The surface of the spherical lens is much 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?*

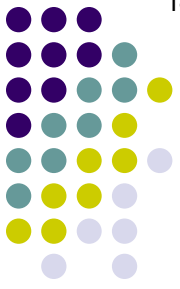
Most cases of high myopia are due to excessive length of the optical axis (so-called 'axial myopia')



# Q

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  - Can cause pupillary block with subsequent angle closure glaucoma



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



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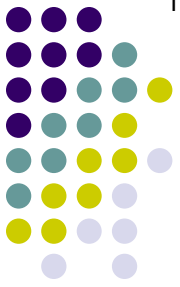
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  - Can cause **pupillary block** with subsequent angle closure glaucoma T

*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



## Microspherophakia



Microspherophakia. Lens is able to fit through the pupillary aperture with mydriasis

# Microspherophakia



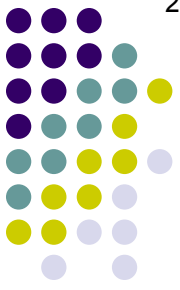
Microspherophakia with pupillary block leading to shallow AC



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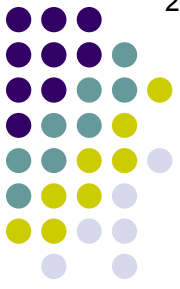


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*What surgical maneuvers are sometimes used to prophylax against angle closure?*



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*What surgical maneuvers are sometimes used to prophylax against angle closure?*  
Iridotomy, or lensectomy



## Microspherophakia

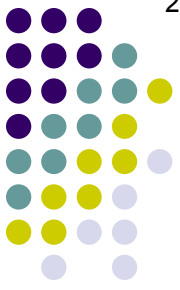
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*What surgical maneuvers are sometimes used to prophylax against angle closure?*

Iridotom<sup>ies</sup>y or iridectomy

Some surgeons argue that **two** iridotomies 180° apart should be created to insure against pupillary blockage by a subluxed microspherophakic lens!

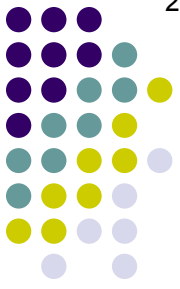




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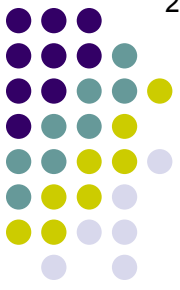
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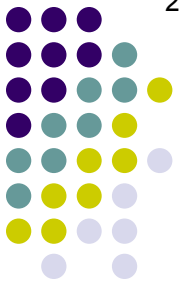
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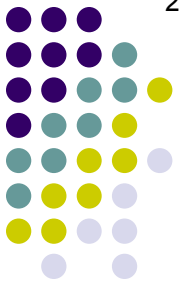


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*Pilo is used to manage pupillary-block angle-closure glaucoma. Why shouldn't it be used in cases secondary to microspherophakia, and why is cycloplegia employed instead?*



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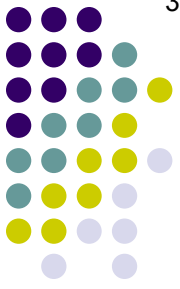
Pilo will cause the lens to move farther **forward**, and will likely worsen the pupillary block. Cycloplegics will pull the lens *posteriorly*, away from the pupil.



# Q

## Microspherophakia

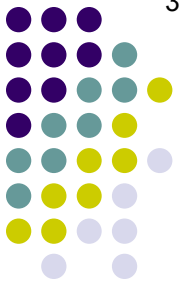
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  - Strongly associated with Marfan syndrome



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**This word** is what makes the statement false. Marfan is indeed associated with microspherophakia, but the relationship is by no means a strong one!





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  - **Strongly** associated with ~~Marfan~~ <sup>?</sup> syndrome ~~F~~ **T**



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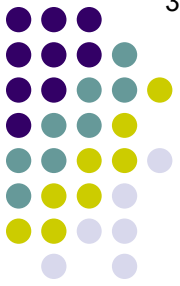
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# Microspherophakia



Microspherophakia in Weill-Marchesani syndrome



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- Angle closure can <sup>no</sup> be treated with miotics ~~F~~ **T**
- Cycloplegics should be avoided, as they can <sup>lock</sup> close an already crowded angle ~~F~~ **T**
- Strongly associated with <sup>Weill-Marchesani</sup>Marfan **syndrome** ~~F~~ **T**

*What are the findings in Weill-Marchesani?*  
 Patients with Weill-Marchesani have:

stature

*Weill-Marchesani*



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*What are the findings in Weill-Marchesani?*  
 Patients with Weill-Marchesani have:  
 ...**short stature**

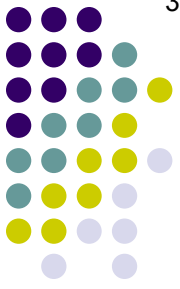
*Weill-Marchesani*



## Microspherophakia



Weill-Marchesani syndrome: Short stature



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*What are the findings in Weill-Marchesani?*

Patients with Weill-Marchesani have:

...**short** stature

**brachydactyly** fingers

*Weill-Marchesani*



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 Patients with Weill-Marchesani have:  
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# Microspherophakia



Weill-Marchesani syndrome: Short fingers



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Patients with Weill-Marchesani have:

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*What are the findings in Weill-Marchesani?*

Patients with Weill-Marchesani have:

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(Think of it as the opposite of      syndrome)

*Weill-Marchesani*

**syndrome**



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*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)

*Weill-Marchesani*

**syndrome**

# Microspherophakia



Weill-Marchesani syndrome



Marfan syndrome



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● Can cause pupillary block  
closure glaucoma

What is the formal term for:  
--Abnormally short fingers?

● Angle closure can  
with miotics **F T**

... short stature  
... **short fingers**

(Tall stature)  
(Long fingers)

... stiff joints

(Lax joints)

● Cycloplegics should be avoided, as they can  
close an already crowded angle **F T**

(Think of it as the opposite of Marfan syndrome)

● Strongly associated with <sup>Weill-Marchesani</sup> Marfan **syndrome** **F T**

Weill-Marchesani



# A

## Microspherophakia

- *Re microspherophakia...which of the following are true?*
  - Due to faulty development of 2ndry lens fibers **T**
  - Associated with high ~~hyperopia~~ <sup>myopia</sup> **F T**
  - Can cause pupillary block closure glaucoma
  - Angle closure can <sup>no</sup> be prevented with miotics **F T**
  - Cycloplegics should be avoided, as they can close an already crowded angle **F T**
  - Strongly associated with ~~Marfan~~ <sup>Weill-Marchesani</sup> **syndrome** **F T**

What is the formal term for:

--Abnormally short fingers? **Brachydactyly**

... short stature

... **short fingers**

... stiff joints

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

(**Tall** stature)

(**Long** fingers)

(**Lax** joints)

lock





# Q

## Microspherophakia

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

● Due to faulty development of 2ndry lens fibers **T**

● Associated with high <sup>myopia</sup> hyperopia **F T**

● Can cause pupillary block closure glaucoma

● Angle closure can <sup>no</sup> be treated 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 is the formal term for:

--Abnormally short fingers? **Brachydactyly**

--Abnormally long fingers?

...short stature

...short fingers

...stiff joints

(Think of it as the opposite of Marfan syndrome)

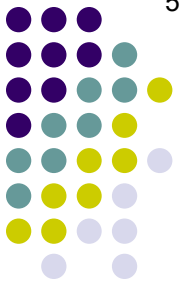
(Tall stature)

(Long fingers)

(Lax joints)

*Weill-Marchesani*

**syndrome**



# A

## Microspherophakia

- *Re microspherophakia...which of the following are true?*
  - Due to faulty development of 2ndry lens fibers **T**
  - Associated with high ~~hyperopia~~ <sup>myopia</sup> **F T**
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  - Angle closure can <sup>no</sup> be prevented with miotics **F T**
  - Cycloplegics should be avoided, as they can close an already crowded angle **F T**
  - Strongly associated with ~~Marfan~~ <sup>Weill-Marchesani</sup> **syndrome** **F T**

What is the formal term for:

--Abnormally short fingers? **Brachydactyly**

--Abnormally long fingers? **Arachnodactyly**

... ~~short~~ stature

(~~Tall~~ stature)

... ~~short~~ fingers

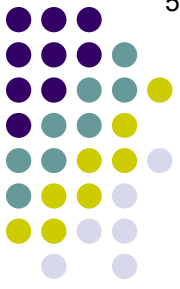
(**Long** fingers)

... ~~stiff~~ joints

(~~Lax~~ joints)

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

lock

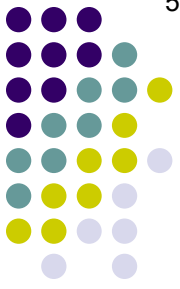


# Q

## Microspherophakia

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  - Due to faulty development of 2ndry lens fibers **T**
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  - Strongly associated with <sup>Weill-Marchesani</sup> ~~Marfan~~ **syndrome** **F T**

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



# A

## Microspherophakia

- *Re microspherophakia...which of the following are true?*
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  - Associated with high <sup>myopia</sup>hyperopia ~~**F**~~ **T**
  - Can cause pupillary block with subsequent angle closure glaucoma **T**
  - Angle closure can <sup>not</sup> be successfully prophylaxed with miotics ~~**F**~~ **T**
  - Cycloplegics should be <sup>used</sup> ~~avoided~~, as they can <sup>reduce the risk of pupillary block</sup> ~~close an already crowded angle~~ ~~**F**~~ **T**
  - Strongly associated with <sup>Weill-Marchesani</sup> ~~Marfan~~ **syndrome** ~~**F**~~ **T**

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

Low syndrome, Alport syndrome, Marfan syndrome, Peters anomaly and congenital rubella

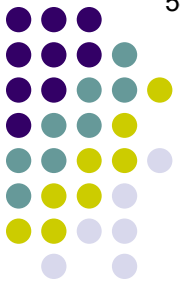


## Microspherophakia

- *Re microspherophakia...which of the following are true?*
  - Due to faulty development of 2ndry lens fibers **T**
  - Associated with high ~~hyperopia~~ <sup>myopia</sup> **F T**
  - **Ruby LAMP** is a mnemonic for the other conditions associated with microspherophakia:
    - Ruby** = Rubella
    - L**owe syndrome
    - A**lport syndrome
    - M**arfan syndrome
    - P**eters anomaly
  - ~~close an already crowded angle~~ **F T**
  - Strongly associated with Weill-Marchesani ~~Marfan syndrome~~ **F T**

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

Lowey syndrome, Alport syndrome, Marfan syndrome, Peters anomaly and congenital rubella



# Q

## Microspherophakia

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

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clo Ruby = Rubella

- An **Lowe syndrome**  
wit **Alport syndrome**

Marfan syndrome

- Cy Peters anomaly

close an already

- Strongly associa

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

two words

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



# A

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Ruby = Rubella

**Lowe syndrome**  
**Alport syndrome**

Marfan syndrome

Peters anomaly

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

- Strongly associated

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

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# Q

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

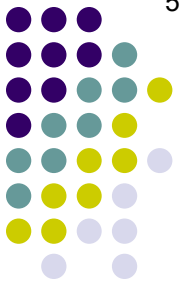
Familial oculorenal syndromes

*What is their classic (nonocular) presenting sign?*

*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





# A

## Microspherophakia

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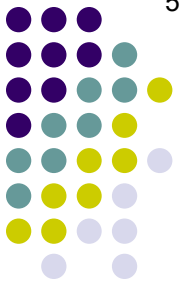
*What is their classic (nonocular) presenting sign?*

Hematuria

- Strongly associated

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

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# Q

## Microspherophakia

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close an already

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

Familial oculorenal syndromes

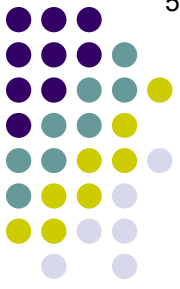
*What is their classic (nonocular) presenting sign?*

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**, Marfan syndrome, Peters anomaly and congenital rubella



# A

## Microspherophakia

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

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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

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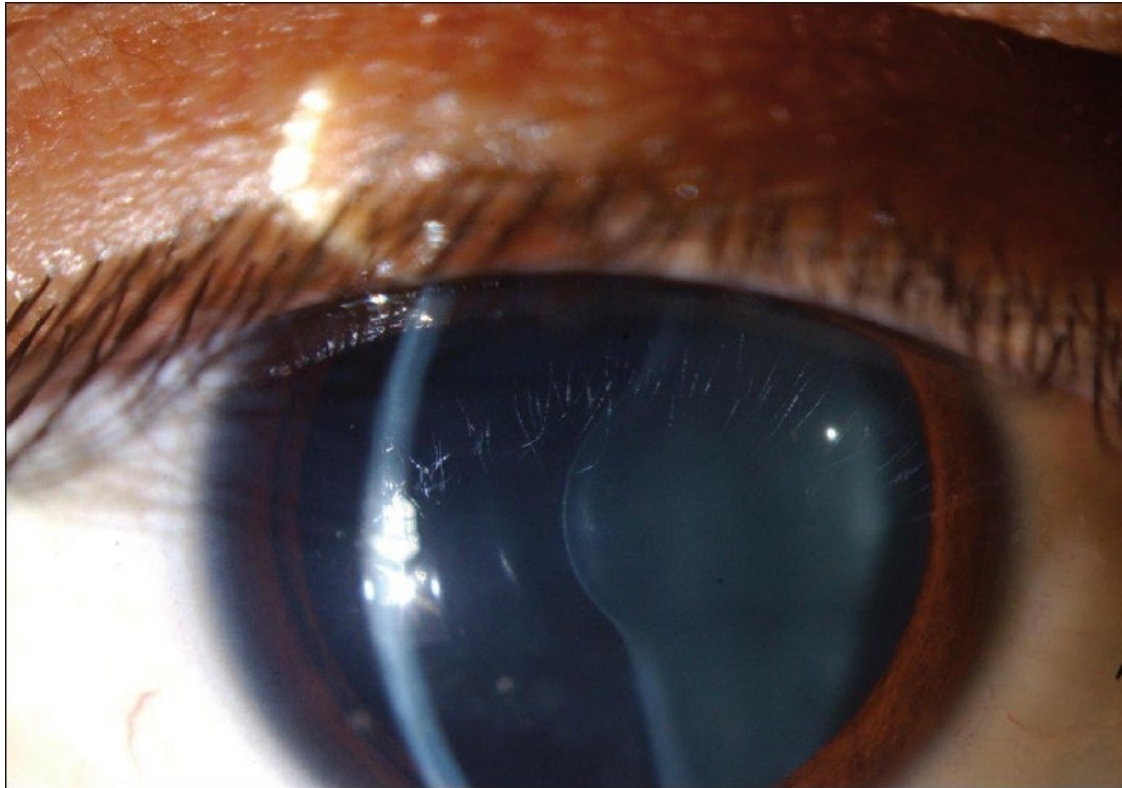
**Lenticonus**

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

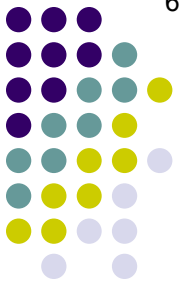
**Lowe syndrome, Alport syndrome**, Marfan syndrome, Peters anomaly and congenital rubella



## Microspherophakia



Anterior lenticonus in Alport syndrome



# Q

## Microspherophakia

- *Re microspherophakia...which of the following are true?*
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  - Associated with high ~~hyperopia~~ <sup>myopia</sup> **F T**
  - Can cause pupillary block with subsequent angle closure glaucoma **T**
  - Angle closure can <sup>not</sup> be successfully prophylaxed with miotics **F T**
  - Cycloplegics should be <sup>used</sup> ~~avoided~~, as they can <sup>reduce the risk of pupillary block</sup> ~~close an already crowded angle~~ **F T**
  - Strongly associated with ~~Marfan~~ <sup>Weill-Marchesani</sup> syndrome **F T**
  - Occurs as part of *ectopia lentis et pupillae*



# A

## Microspherophakia

- *Re microspherophakia...which of the following are true?*
  - Due to faulty development of 2ndry lens fibers **T**
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  - Strongly associated with ~~Marfan~~ <sup>Weill-Marchesani</sup> syndrome **F T**
  - Occurs as part of *ectopia lentis et pupillae* **T**



## Microspherophakia

Q

- Re microspherophakia...which of the following

What is ectopia lentis et pupillae?

- Due to
- Associa
- Can ca  
closure
- Angle o  
with mi
- Cyclop  
close-a
- Strongly
- Occurs as part of **ectopia lentis et pupillae** T



# A

## Microspherophakia

- *Re microspherophakia...which of the following*

*What is ectopia lentis et pupillae?*

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

- Due to
- Associated
- Can cause angle closure
- Angle of
- with mi
- Cyclop
- close a
- Strongly
- Occurs as part of ***ectopia lentis et pupillae*** **T**





# Q

## Microspherophakia

- Re microspherophakia...which of the following

*What is ectopia lentis et pupillae?*

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

- Due to

- Associated

*How common is it?*

- Can cause angle closure

- Angle of vision with mi

- Cyclopia close a

- Strongly

- Occurs as part of **ectopia lentis et pupillae** **T**



# A

## Microspherophakia

- *Re microspherophakia...which of the following*

*What is ectopia lentis et pupillae?*

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

- Due to

- Associated

*How common is it?*

It is very rare

- Can cause
- closure

- Angle of
- with mi

- Cyclop
- close a

- Strongly

- Occurs as part of ***ectopia lentis et pupillae*** **T**



# Q

## Microspherophakia

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- Due to

- Associated

- Can cause

closure

- Angle of

with mi

- Cyclop

close a

- Strongly

- Occurs as part of **ectopia lentis et pupillae** **T**

*What is ectopia lentis et pupillae?*

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?*



# A

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Bilateral

- Angle of

with mi

- Cyclop

close-a

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# Q

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Bilateral

*In what direction are the pupils and lenses displaced?*



# Q/A

## Microspherophakia

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- Cyclop

close-a

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Bilateral

*In what direction are the pupils and lenses displaced?*

In opposite directions—pupils                     , lenses



# A

## Microspherophakia

- *Re microspherophakia...which of the following*

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

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closure

*Is it unilateral, or bilateral?*

Bilateral

- Angle of

with mi

*In what direction are the pupils and lenses displaced?*

In opposite directions—pupils inferotemporal , lenses superonasal

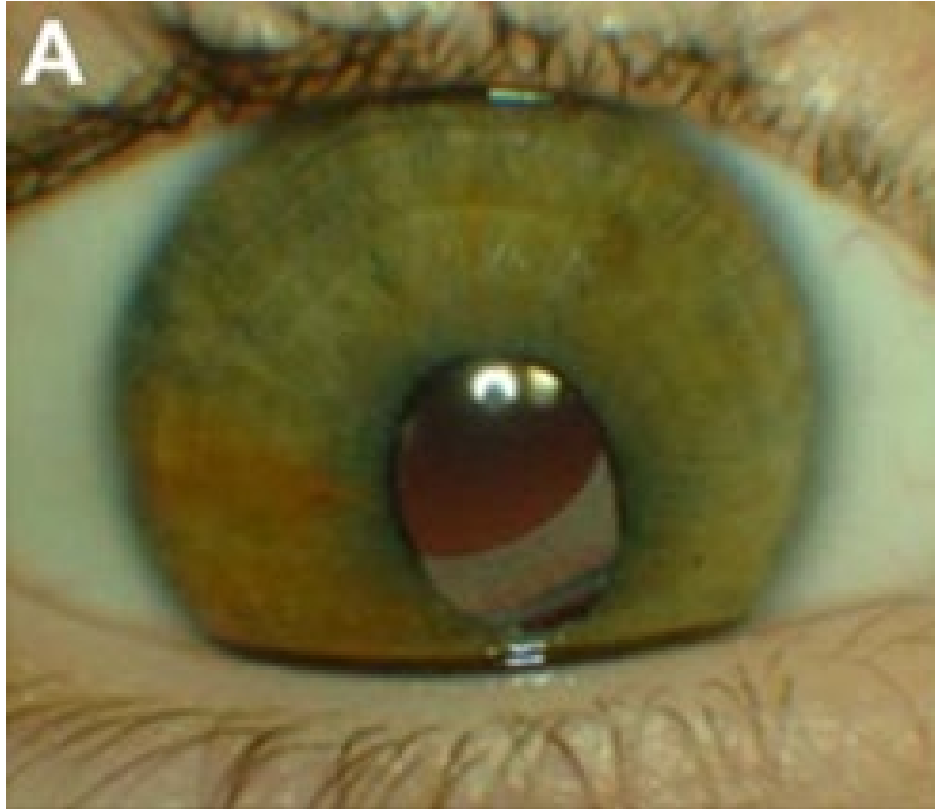
- Cyclop

close-a

- Strongly

- Occurs as part of **ectopia lentis et pupillae** **T**

## Microspherophakia



Ectopia lentis et pupillae: Pupil displaced inferonasal; lens, superotemporal





# Q

## Microspherophakia

- Re microspherophakia...which of the following

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Bilateral

*In what direction are*

In opposite direction

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*Can the pupils and lenses be so displaced that the lens isn't located within the pupillary aperture?*



# A

## Microspherophakia

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In opposite direction

**—pupils inferotemporal , lenses superonasal**

- Cyclop

close a

*Can the pupils and lenses be so displaced that the lens isn't located within the pupillary aperture?*

They can indeed

- Strongly

- Occurs as part of **ectopia lentis et pupillae** **T**



# Q

## Microspherophakia

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*In what direction are the pupils and lenses displaced?*

In opposite directions—pupils inferotemporal , lenses superonasal

- Cyclop

close-a

*The pupils typically have two further abnormalities—what are they?*

--?

--?

- Strongly

- Occurs as part of **ectopia lentis et pupillae** **T**



# Q/A

## Microspherophakia

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In opposite directions—pupils inferotemporal , lenses superonasal

*The pupils typically have two further abnormalities—what are they?*

--They are very miotic vs dilated , and dilate well vs poorly

--?



# A

## Microspherophakia

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--?

# Microspherophakia

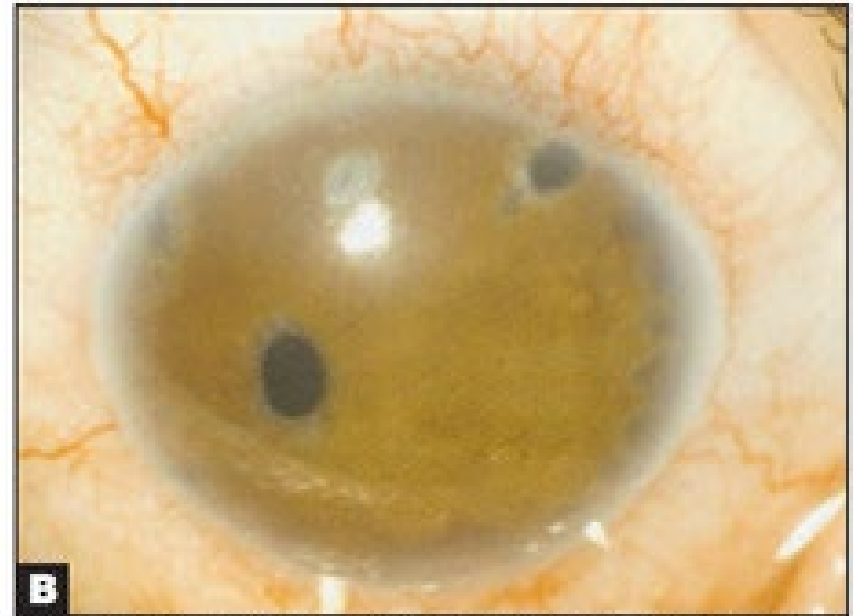


Figure 1. Preoperative view of both eyes in a patient with bilateral ectopia lentis et pupillae syndrome showing several lesions on the iris due to previous laser therapy. (A) Right eye. (B) Left eye.

Ectopia lentis et pupillae: Itty-bitty pupils (ignore the LPIs)



# Q

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*In what direction are the pupils and lenses displaced?*

In opposite directions—pupils inferotemporal , lenses superonasal

*The pupils typically have two further abnormalities—what are they?*

--They are very miotic , and dilate poorly

--They are 'something -like' in shape



# A

## Microspherophakia

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- Associated

- Can cause

closure

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In opposite directions—pupils inferotemporal , lenses superonasal

*The pupils typically have two further abnormalities—what are they?*

--They are very miotic , and dilate poorly

--They are slit-like in shape



# Microspherophakia



*Ectopia lentis et pupillae*: Bilateral inferonasal displacement of slit(ish) pupils