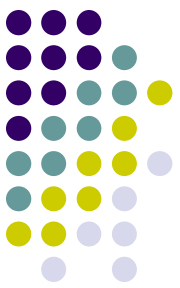


Q



# Lens luxation/subluxation

*What do these terms mean?*

*--Sublux(at)ed:*

*--Lux(at)ed*

A

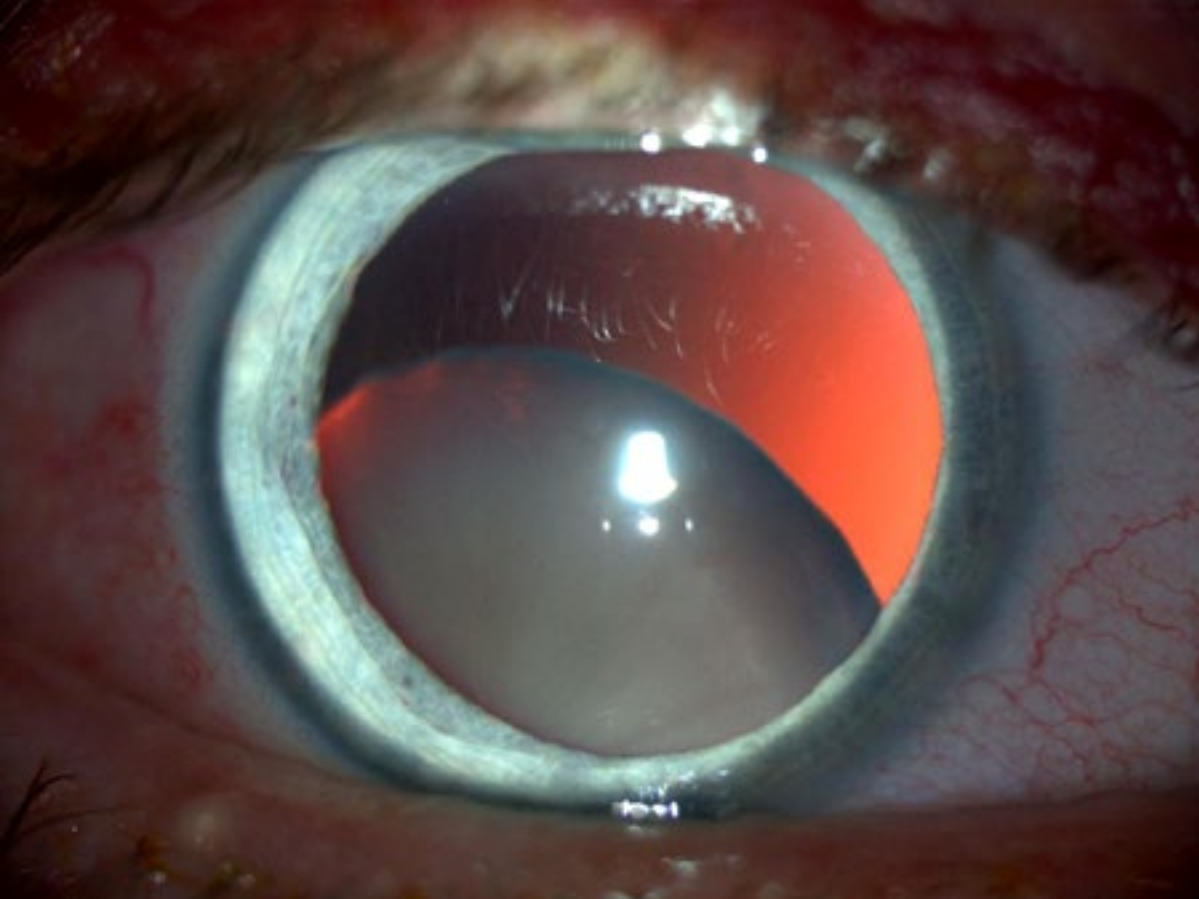
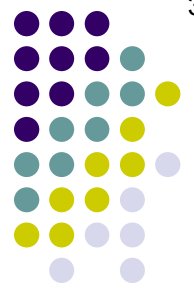


## Lens luxation/subluxation

*What do these terms mean?*

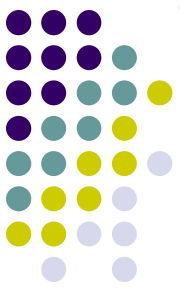
--*Sublux(at)ed*: The lens is partially displaced, but remains in the 'general area'

--*Lux(at)ed*



Subluxed lens

Q



## Lens luxation/subluxation

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--*Lux(at)ed*:

# A

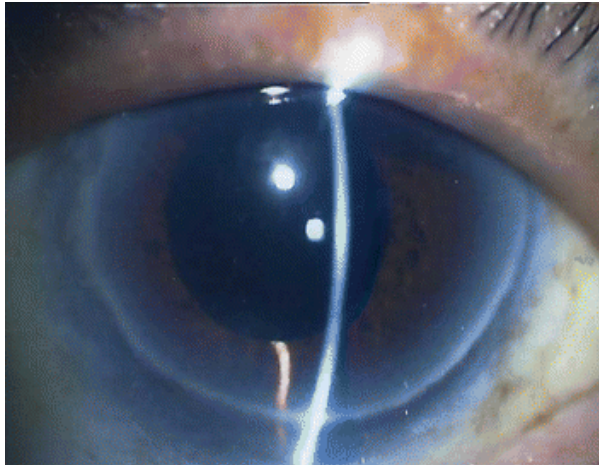
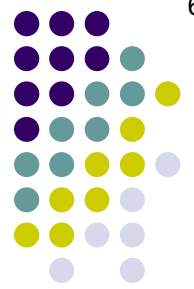


## Lens luxation/subluxation

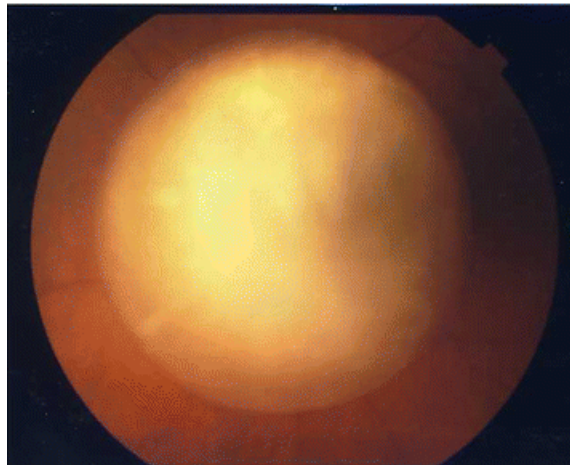
*What do these terms mean?*

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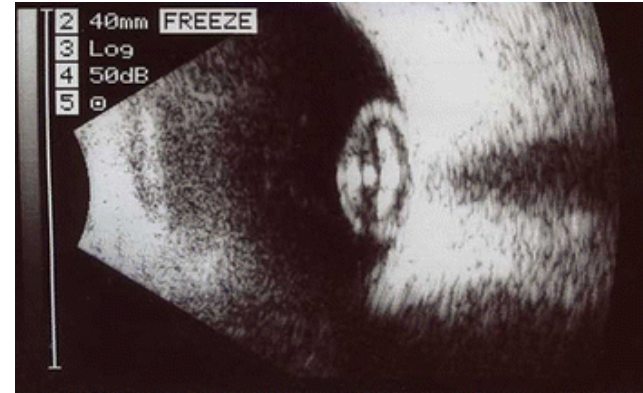
--*Lux(at)ed*: The lens is dislocated--completely removed from the pupillary aperture. All zonular attachments have been disrupted.



Aphakic



Lens resting on the retina



b-scan: lens on ONH

Luxated lens

# Q



- Name 7 causes of lens luxation/subluxation in the pediatric population:



(Yeah, I know there are only 6 dots...)



*Mnemonic forthcoming...*

# Q

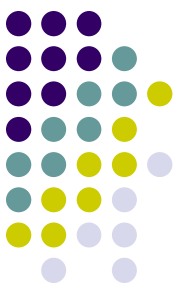


- Name 7 causes of lens luxation/subluxation in the pediatric population:
  - **T**
  - **H**
  - **A**
  - **M**
  - **E**
  - **S** (two causes)

*Think of countless displaced crystalline lenses floating on the Thames river in London...*



# A



- Name 7 causes of lens luxation/subluxation in the pediatric population:
  - **T**rauma
  - **H**omocystinuria
  - **A**niridia
  - **M**arfan syndrome
  - **E**ctopia lentis et pupillae
  - **S**imple ectopia lentis; **S**yphilis (congenital)

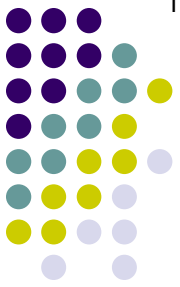
*Think of countless displaced crystalline lenses floating on the Thames river in London...*



# Q

- Name 7 causes of lens luxation/subluxation in the pediatric population:
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  - Aniridia
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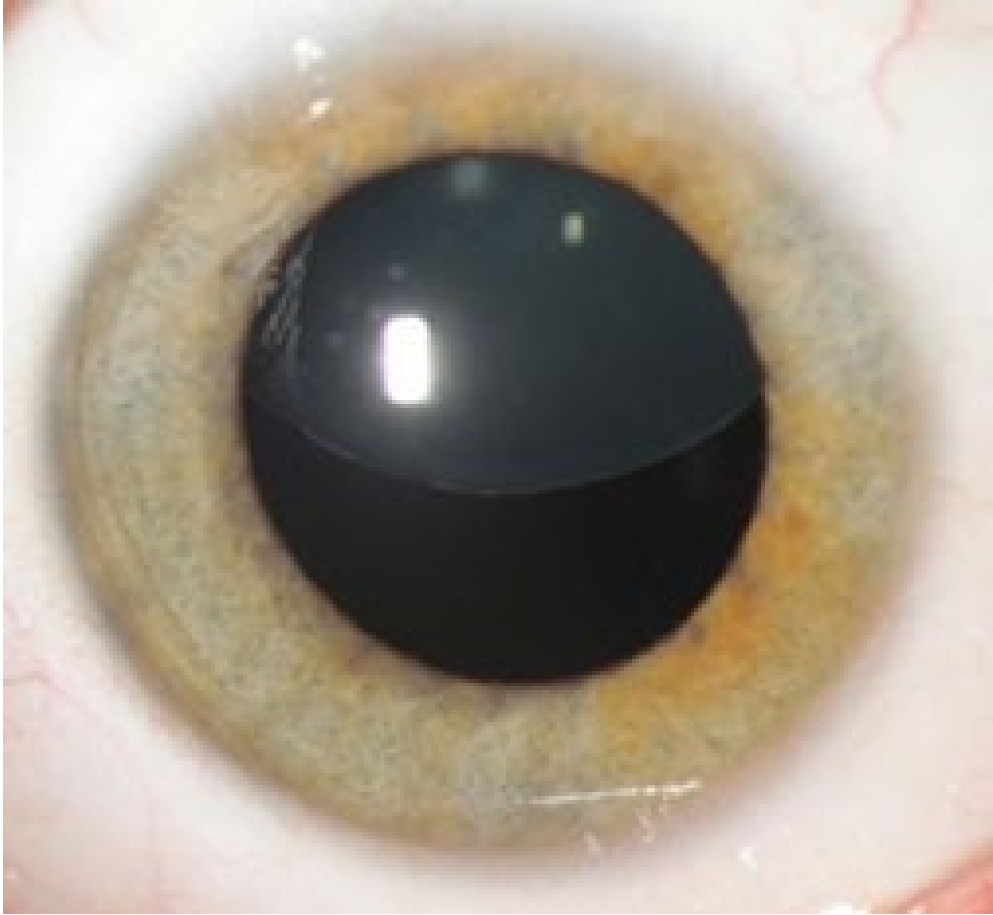
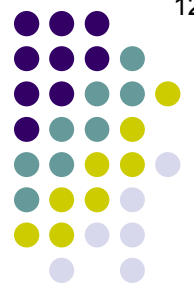
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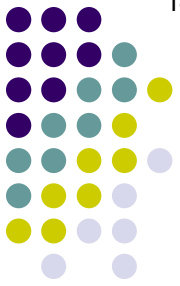
Marfan: Dislocated lens



# Q

- Name 7 causes of lens luxation/subluxation in the pediatric population:
  - Trauma
  - Homocystinuria *Which is the #1 cause of dislocating the lens into the anterior chamber?*
  - Aniridia
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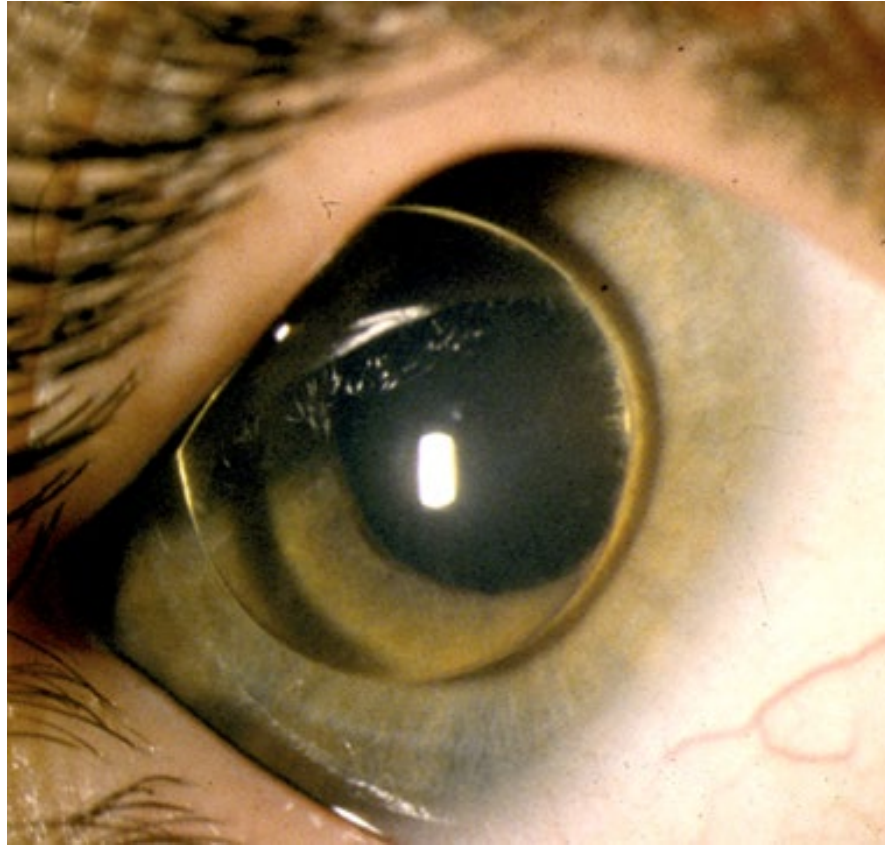
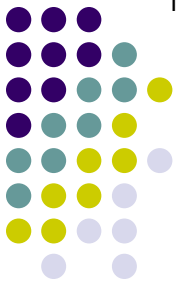
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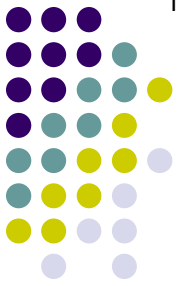
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*Think of countless displaced crystalline lenses floating on the Thames river in London...*



Homocystinuria: Lens in the AC



# Q

- Name 7 causes of lens luxation/subluxation in the pediatric population:

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*Which is the #1 cause of dislocating the lens into the anterior chamber?*

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- Aniridia

- Marfan syndrome

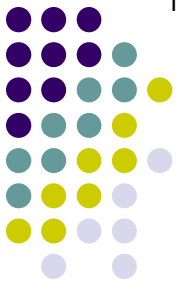
*What percentage of homocystinuria patients will have subluxation...  
--in infancy?*

- Ectopia lentis et pup

- Simple ectopia lentis; Syphilis (congenital)

*Think of countless displaced crystalline lenses floating on the Thames river in London...*





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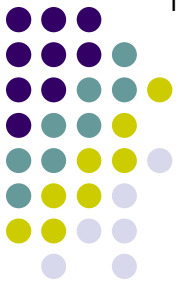
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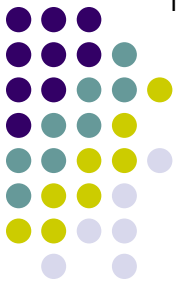
*--in infancy? **30***

*--by age 15?*

- Ectopia lentis et pup

- Simple ectopia lentis; **Syphilis** (congenital)

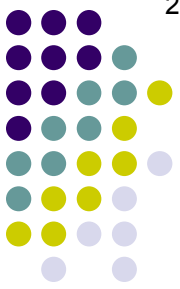
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--in infancy? **30**  
--by age 15? **80!***
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# Q

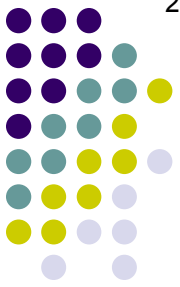
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*Identify key differences between Marfan syndrome and homocystinuria with respect to...*

**--zonule status:**

*Think of countless displaced crystalline lenses floating on the Thames river in London...*

# Q/A

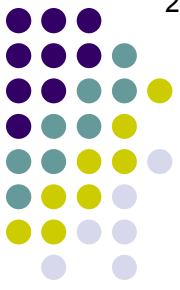


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*Think of countless displaced crystalline lenses floating on the Thames river in London...*



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- **Marfan syndrome**

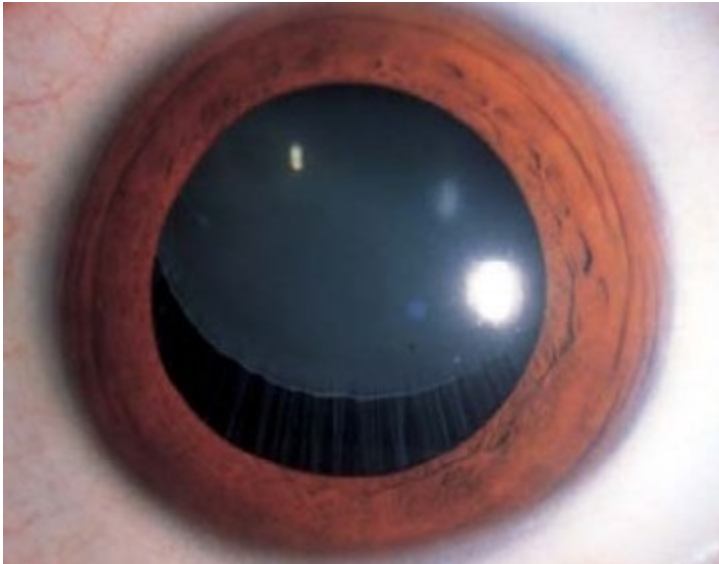
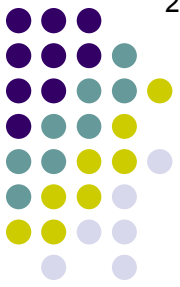
- Ectopia lentis et pupillae

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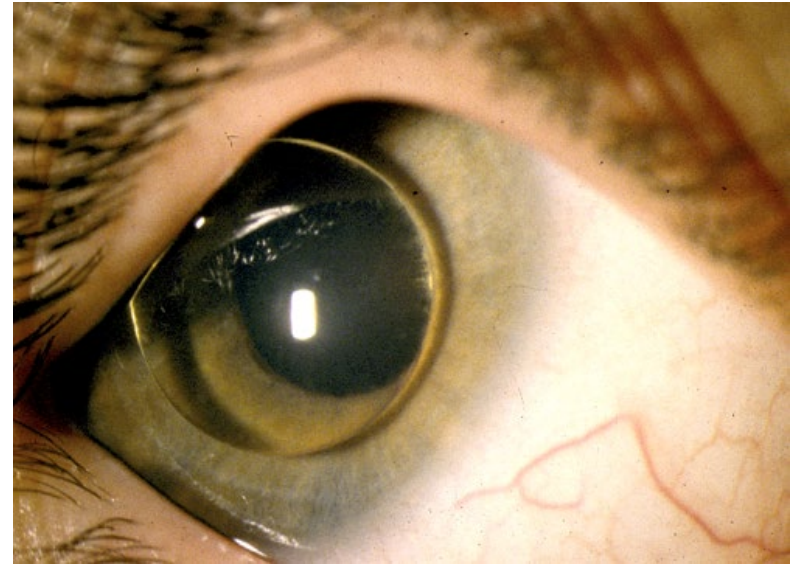
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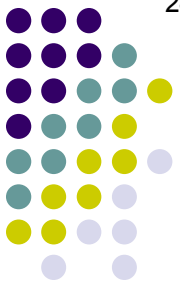
*Think of countless displaced crystalline lenses floating on the Thames river in London...*



Marfan: Stretched zonules



Homocystinuria: Broken zonules



# Q

- Name 7 causes of lens luxation/subluxation in the pediatric population:

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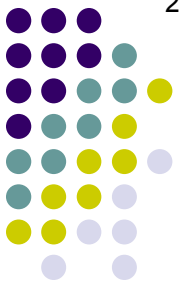
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# Q/A



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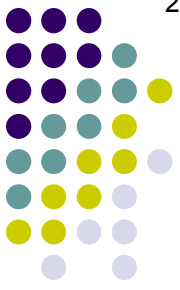
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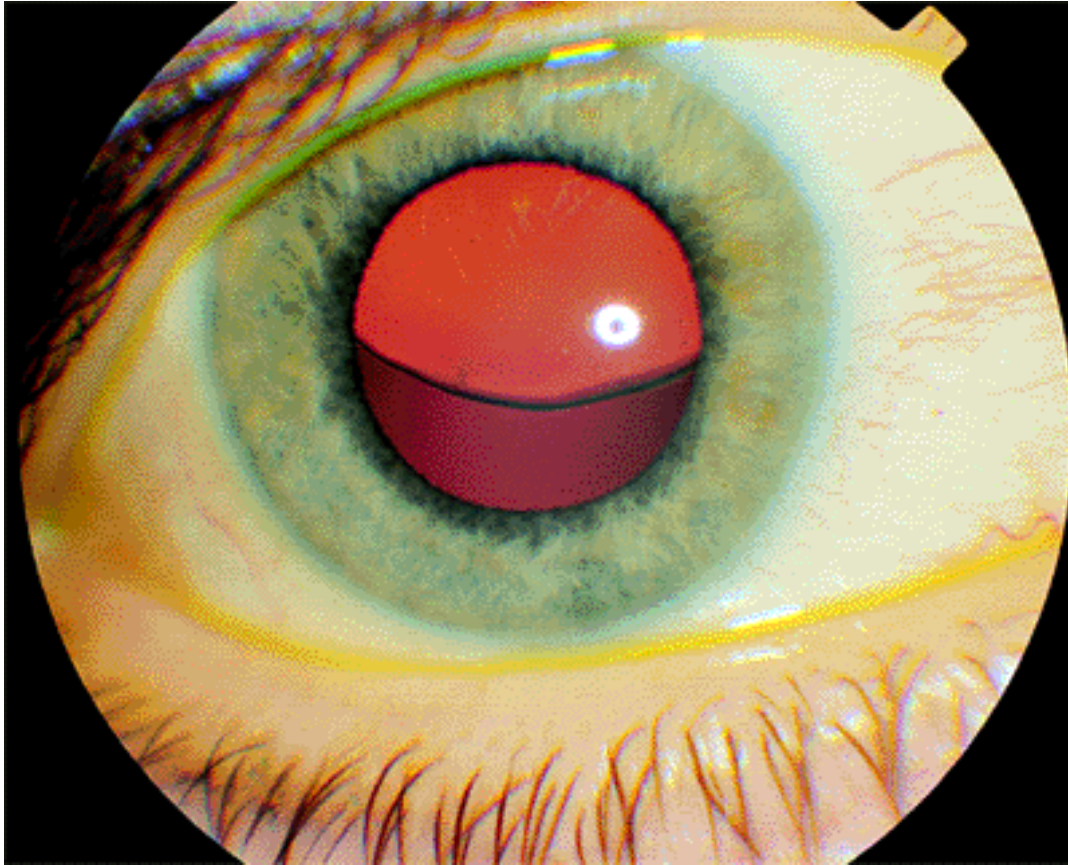
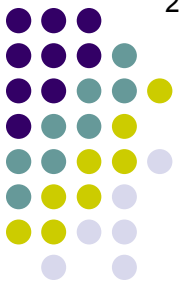
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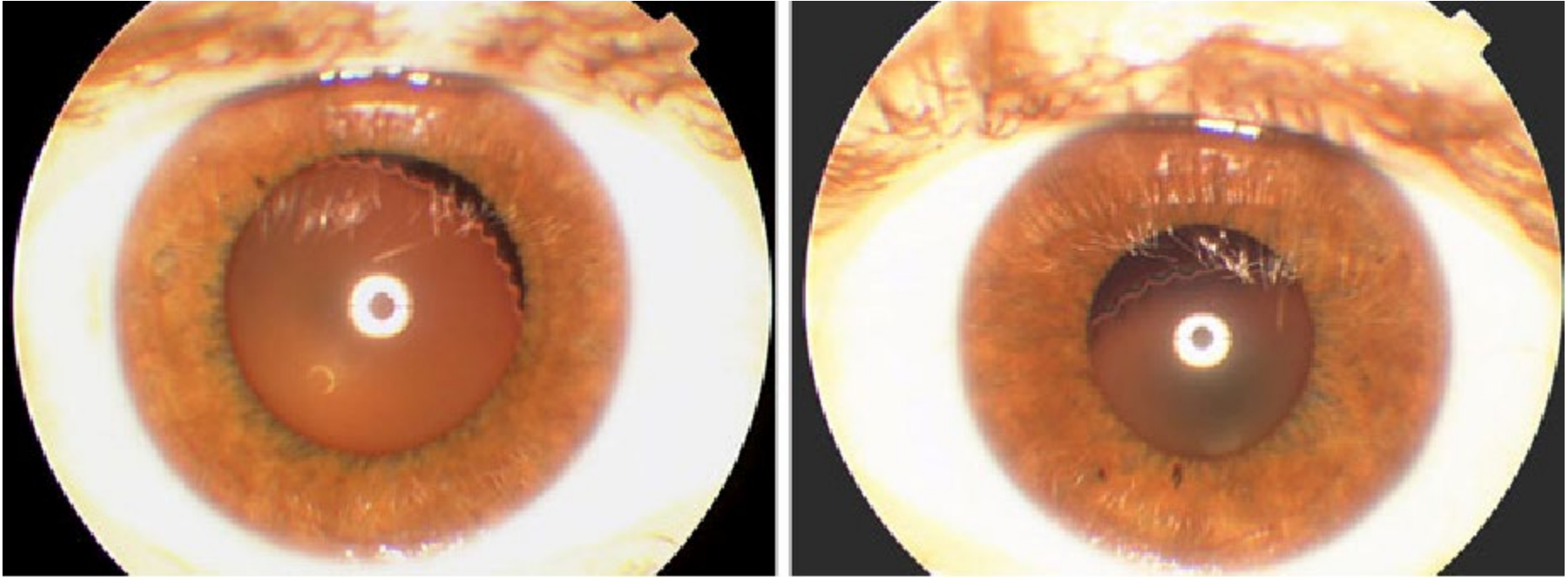
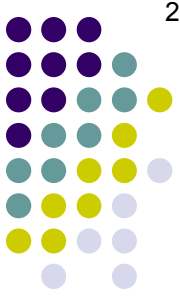
*Identify key differences between Marfan syndrome and homocystinuria with respect to...*

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*Think of countless displaced crystalline lenses floating on the Thames river in London...*



Marfan: Lens subluxed upward



Homocystinuria: Lens subluxed downward

Why are zonules broken in homocystinuria?

Q

● Name 7 causes in the pediatric p

● Trauma

● **Homocystinuria**

● Aniridia

● Marfan syndrome

● Ectopia lentis et pupillae

● Simple ectopia lentis; Syphilis (congenital)

Syndrome and homocystinuria with respect to...

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Think of countless displaced crystalline lenses floating on the Thames river in London...

# A

*Why are zonules broken in homocystinuria?*

Homocystinuria involves abnormal methionine metabolism, which ultimately results in a deficiency of cysteine.

- Name 7 causes in the pediatric population

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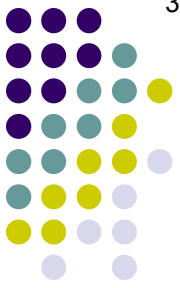
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Homocystinuria

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*What other problems manifest in homocystinuria?*

--M  
--O  
--S  
--T

**MOST** homocystinurics have these problems

*Syndrome and homocystinuria with respect to...*

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--Mental retardation

--Osteoporosis

--Seizures

--Thrombosis of the systemic vasculature

**MOST** homocystinurics have these problems

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- Name 7 causes in the pediatric p

***For more on homocystinuria, see slide-set FELT14***

- H**omocystinuria
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*Think of countless displaced crystalline lenses floating on the Thames river in London...*

What percentage of Marfan patients develop ectopia lentis?

Q

• M  
ir

- Homocystinuria
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- Ectopia lentis et pupillae
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*syndrome and homocystinuria with respect to...*

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*Think of countless displaced crystalline lenses floating on the Thames river in London...*



What percentage of Marfan patients develop ectopia lentis?

50 - 80

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--? [lens-related]

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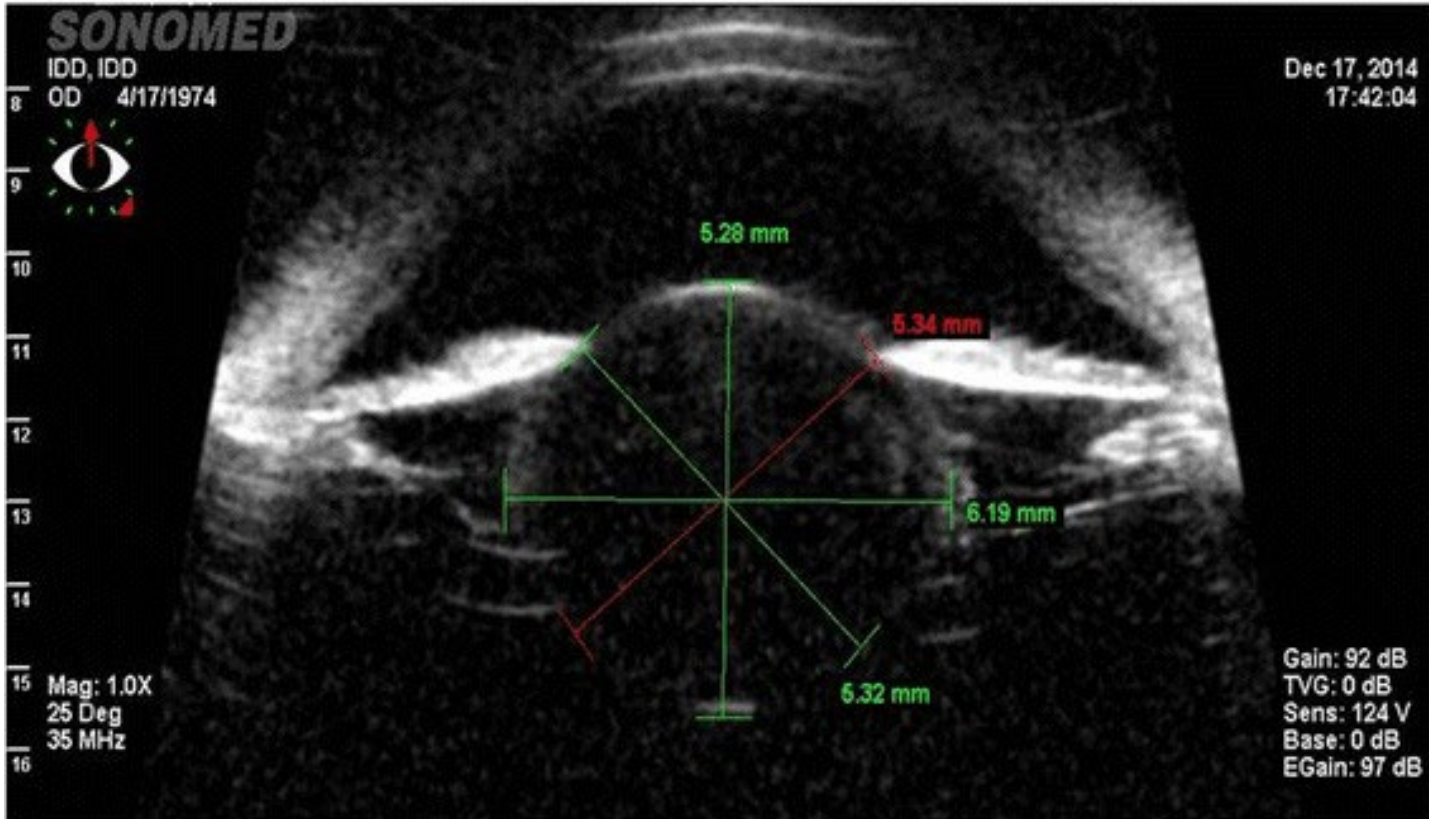
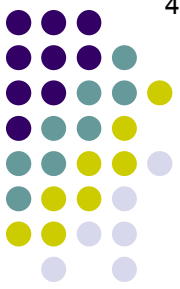
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Microspherophakia. Note the small size, extreme curvature of the lens

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- Increased axial length

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- Increased axial length → axial myopia
- Microspherophakia → increased lens curvature → lenticular myopia
- Lens subluxation → looking through highly curved peripheral lens → lens periphery myopia
- Increased corneal curvature?*

What about the cornea in Marfan pts--does it contribute to their high myopia?

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Give three reasons why **Marfan patients tend to be high myopes**: *spherical-aberration myopia*

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--Increased corneal curvature? *No*

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No—the cornea in Marfan tends to be **flatter** than normal

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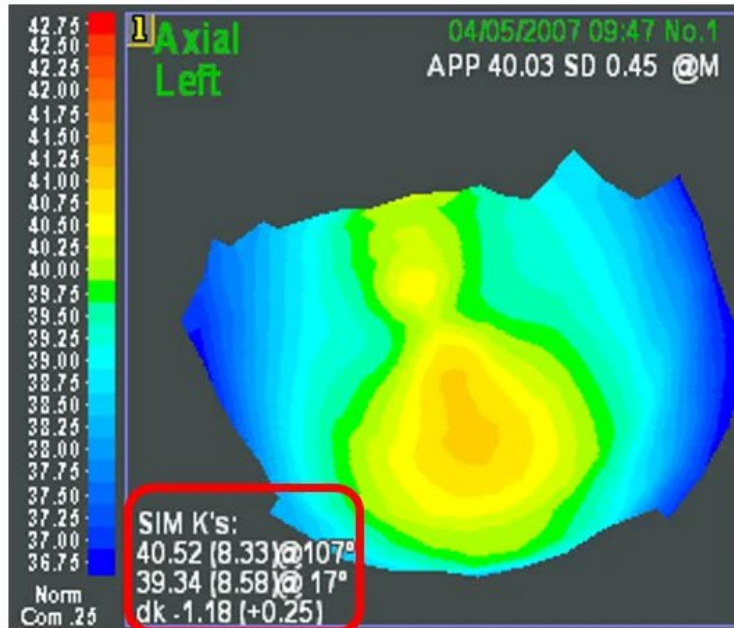
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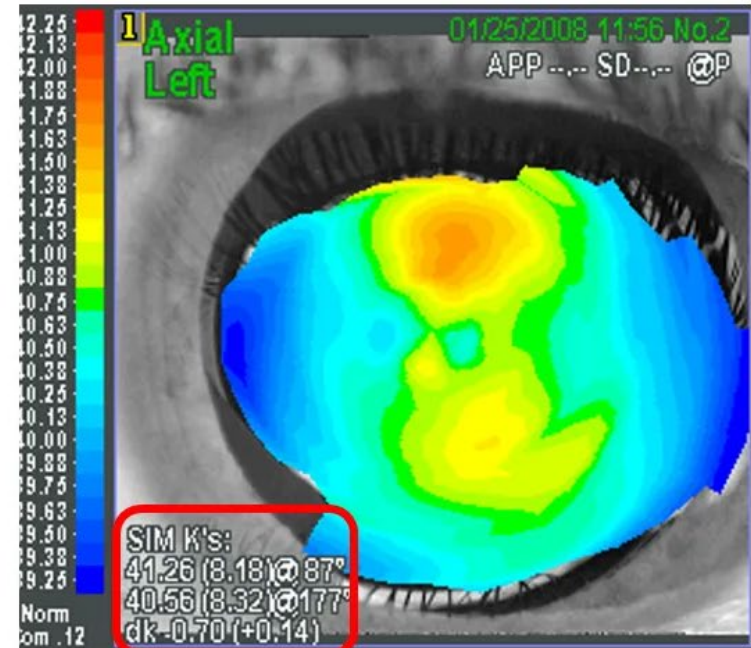
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**FLAT** KERATOMETRY



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Marfan topography. Note the corneas are rather flat



Q

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Is the cornea flatter because it has a smaller-than-normal diameter (ie, microcornea)?

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Is the cornea flatter because it has a smaller-than-normal diameter (ie, microcornea)?  
No—in fact, if anything, corneal diameter in Marfan's tends to be **larger** than normal (ie, megalocornea)

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A spectacle-corrected Marfan patient c/o difficulty reading—why?

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Why not just bust up in there and do a clear lens extraction with IOL?

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Stretched zonules impair accommodation, so bifocals may be needed at an early age

Why not just bust up in there and do a clear lens extraction with IOL?

Because Marfan eyes are at very high risk for vitreous loss as well as retinal detachment

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**For more on Marfan, see slide-set FELT14**

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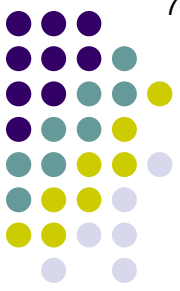
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So how can pupillary block in microspherophakia be pharmacologically prophylaxed?

With **cycloplegics**, though it may seem counterintuitive. By relaxing the ciliary body, cycloplegics tighten the zonules and pull the lens away from the pupil, thereby decreasing the risk of block

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# Q

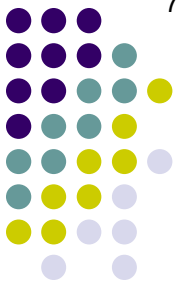
*What is ectopia lentis et pupillae?*

tion

- **Ectopia lentis et pupillae**
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*Think of countless displaced crystalline lenses floating on the Thames river in London...*





# A

*What is ectopia lentis et pupillae?*

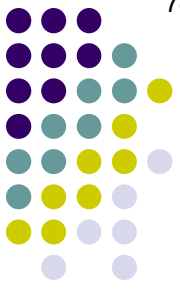
A genetic condition the hallmark of which is the displacement of the pupil and (microspherophakic) lens



tion

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Q

What is ectopia lentis et pupillae?

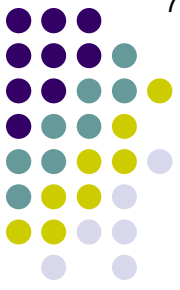
A genetic condition that causes the lens and (microspherophakic) iris to be displaced from the pupil

How is it inherited, ie, is it AR, AD, X-linked, etc?

on

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# A

What is ectopia lentis et pupillae?

A genetic condition that causes the lens and (microspherophakic) iris to be displaced from their normal positions.

How is it inherited, ie, is it AR, AD, X-linked, etc?

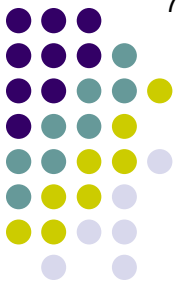
It is AR

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# Q

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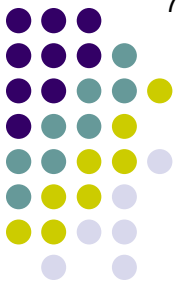
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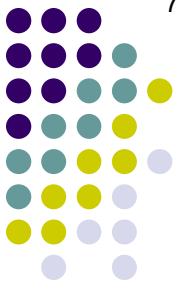
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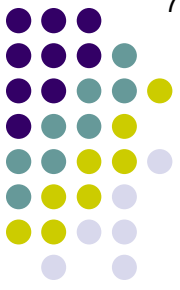
*Is it unilateral, or bilateral?*

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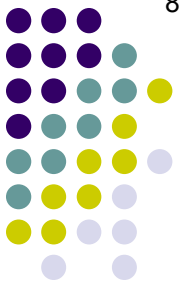
Bilateral

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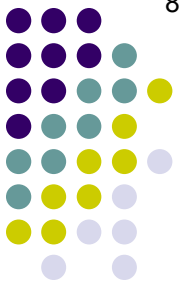
*In what direction are the pupils and lenses displaced?*

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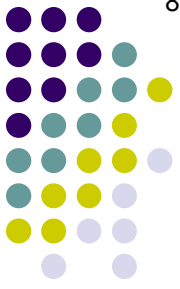
In opposite directions—pupils                     , lenses                     

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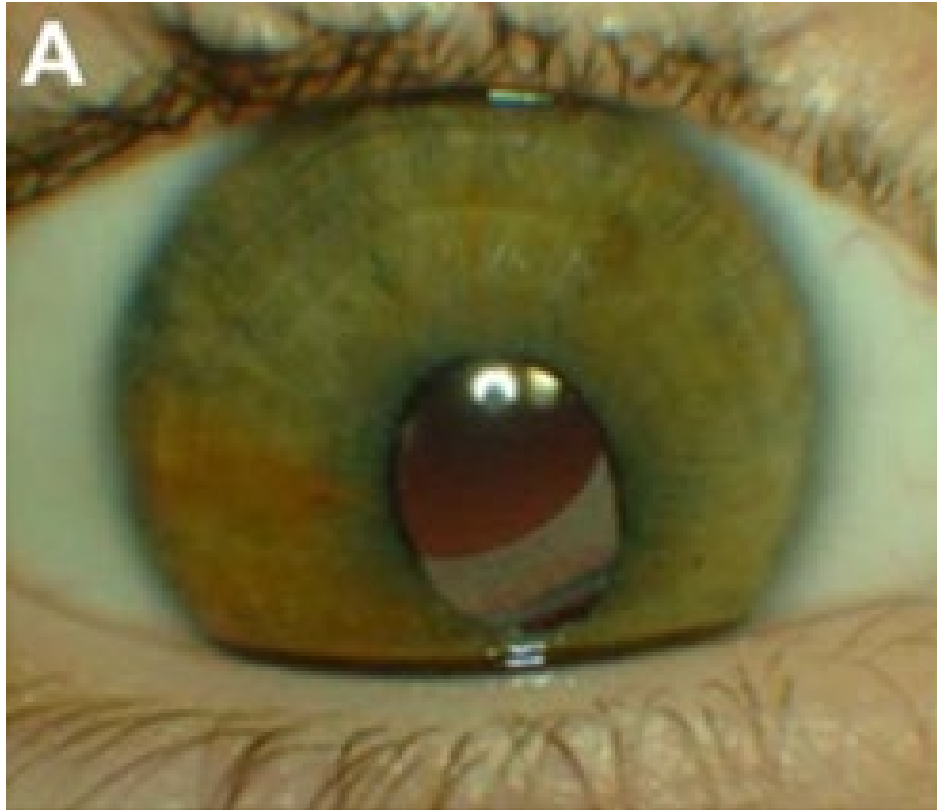
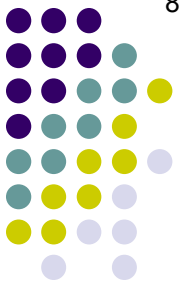
In opposite directions—pupils inferotemporal , lenses superonasal

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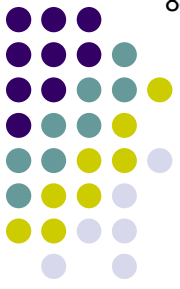
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Ectopia lentis et pupillae: Pupil displaced inferonasal; lens, superotemporal



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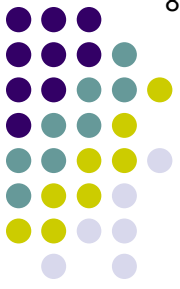
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--They are very miotic vs dilated , and dilate well vs poorly

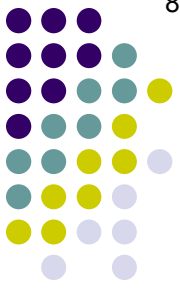
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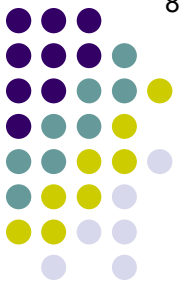
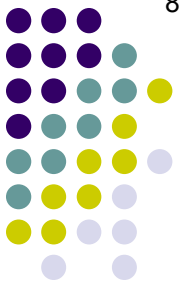


Figure 1. Preoperative view of both eyes in a patient with bilateral ectopia lentis et pupillae syndrome showing several lesions on the iris due to previous laser therapy. (A) Right eye. (B) Left eye.

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



# Q

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In opposite directions—pupils inferotemporal , lenses superonasal

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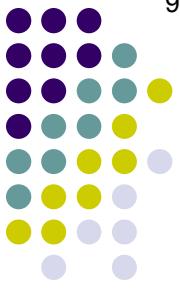
--They are slit-like in shape

- **Ectopia lentis et pupillae**

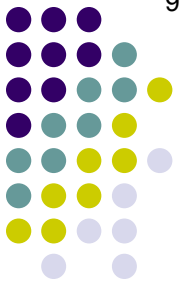
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Ectopia lentis et pupillae: Bilateral inferonasal displacement of slit(ish) pupils



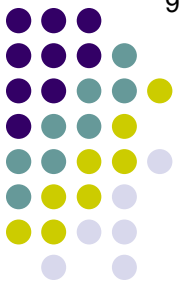
# Q

What is simple ectopia lentis?

xation

- Aniridia
- Marfan syndrome
- Ectopia lentis et pupillae
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# A

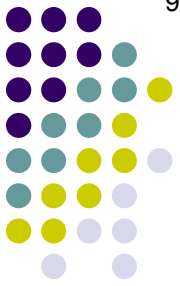
*What is simple ectopia lentis?*

A genetic condition the hallmark of which is subluxation of the lens

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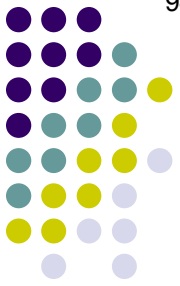
What is simple ectopia lentis?

**A genetic condition** that *How is it inherited, ie, is it AR, AD, X-linked, etc?*

Examination

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# A

What is simple ectopia lentis?

**A genetic condition**

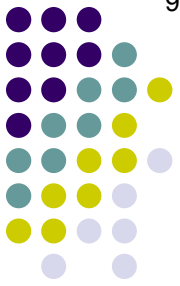
How is it inherited, ie, is it AR, AD, X-linked, etc?

It is AD

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# Q

*What is simple ectopia lentis?*

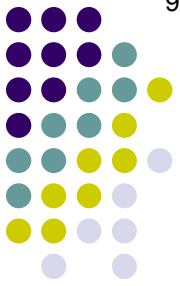
A genetic condition the hallmark of which is subluxation of the lens

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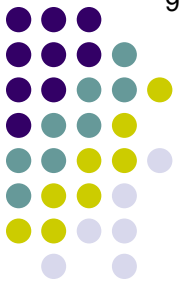
It is uncommon

xation

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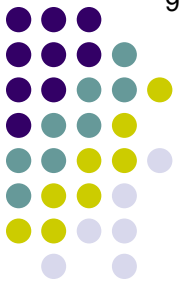
It is uncommon

*Is it unilateral, or bilateral?*

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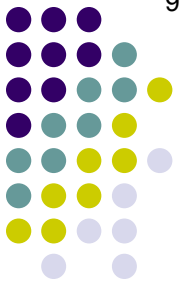
*Is it unilateral, or bilateral?*

Bilateral

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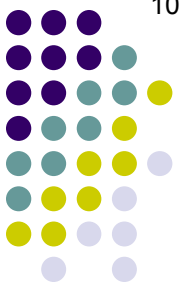
Bilateral

*In what direction are the lenses displaced?*

- Aniridia
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xation

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Superotemporal

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