Lens luxation/subluxation

What do these terms mean?
--Sublux(ate)ed:

--Lux(ate)ed
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
Lens luxation/subluxation

What do these terms mean?
--Sublux(ate)d: The lens is partially displaced, but remains in the ‘general area’
--Lux(ate)d:
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*: The lens is dislocated--completely removed from the pupillary aperture. All zonular attachments have been disrupted.
Luxated lens

Aphakic

Lens resting on the retina

b-scan: lens on ONH
Name 7 causes of lens luxation/subluxation in the pediatric population:

- Tsunamis
- Hurricanes
- Allergic reactions
- Medications
- Sudden trauma
- Mnemonic forthcoming…

Yeah, I know there are only 6 dots…

Mnemonic forthcoming…
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…
- Name 7 causes of lens luxation/subluxation in the pediatric population:
  - Trauma
  - Homocystinuria
  - Aniridia
  - Marfan syndrome
  - Ectopia lentis et pupillae
  - Simple ectopia lentis; Syphilis (congenital)

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

Which is the #1 cause of lens dislocation?
Name 7 causes of lens luxation/subluxation in the pediatric population:

- Trauma
- Homocystinuria
- Aniridia
- Marfan syndrome
- Ectopia lentis et pupillae
- Simple ectopia lentis; Syphilis (congenital)

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

Which is the #1 cause of lens dislocation? Marfan
Marfan: Dislocated lens
Name 7 causes of lens luxation/subluxation in the pediatric population:

- Trauma
- Homocystinuria
- Aniridia
- Marfan syndrome
- Ectopia lentis et pupillae
- Simple ectopia lentis; Syphilis (congenital)

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

Which is the #1 cause of dislocating the lens into the anterior chamber?
Name 7 causes of lens luxation/subluxation in the pediatric population:

- Trauma
- Homocystinuria
- Aniridia
- Marfan syndrome
- Ectopia lentis et pupillae
- Simple ectopia lentis; Syphilis (congenital)

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

Which is the #1 cause of dislocating the lens into the anterior chamber? Homocystinuria
Homocystinuria: Lens in the AC
Name 7 causes of lens luxation/subluxation in the pediatric population:

- Trauma
- Homocystinuria
- Aniridia
- Marfan syndrome
- Ectopia lentis et pupillae
- Simple ectopia lentis; Syphilis (congenital)

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

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

What percentage of homocystinuria patients will have subluxation…in infancy?
● Name 7 causes of lens luxation/subluxation in the pediatric population:
  ● **Trauma**
  ● **Homocystinuria**
  ● **Aniridia**
  ● **Marfan syndrome**
  ● **Ectopia lentis et pupillae**
  ● **Simple ectopia lentis; Syphilis (congenital)**

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

What percentage of homocystinuria patients will have subluxation… in infancy? **30**

Think of countless displaced crystalline lenses floating on the Thames river in London…
Name 7 causes of lens luxation/subluxation in the pediatric population:

- Trauma
- Homocystinuria
- Aniridia
- Marfan syndrome
- Ectopia lentis et pupillae
- Simple ectopia lentis; Syphilis (congenital)

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

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

What percentage of homocystinuria patients will have subluxation…

-- in infancy? **30**
-- by age 15? **80**
19

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

- Trauma
- Homocystinuria
- Aniridia
- Marfan syndrome
- Ectopia lentis et pupillae
- Simple ectopia lentis; Syphilis (congenital)

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

What percentage of homocystinuria patients will have subluxation…
--in infancy? 30
--by age 15? 80!

Think of countless displaced crystalline lenses floating on the Thames river in London…
Name 7 causes of lens luxation/subluxation in the pediatric population:

- Trauma
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- Aniridia
- Marfan syndrome
- Ectopia lentis et pupillae
- Simple ectopia lentis; Syphilis (congenital)

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

Identify key differences between Marfan syndrome and homocystinuria with respect to…
--zonule status:
Q/A

- Name 7 causes of lens luxation/subluxation in the pediatric population:
  - Trauma
  - **Homocystinuria**
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  - Ectopia lentis et pupillae
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Think of countless displaced crystalline lenses floating on the Thames river in London…

Identify key differences between Marfan syndrome and homocystinuria with respect to…

- **Zonule status:** Zonules are stretched in Marfan, but broken in homocystinuria
Name 7 causes of lens luxation/subluxation in the pediatric population:

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

--- **zonule status**: Zonules are stretched in Marfan, but broken in homocystinuria

Think of countless displaced crystalline lenses floating on the Thames river in London...
Marfan: Stretched zonules

Homocystinuria: Broken zonules
Name 7 causes of lens luxation/subluxation in the pediatric population:

- **Trauma**
- **Homocystinuria**
- **Aniridia**
- **Marfan syndrome**
- **Ectopia lentis et pupillae**
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Identify key differences between Marfan syndrome and homocystinuria with respect to...

- **zonule status:** Zonules are stretched in Marfan, but broken in homocystinuria
- **lens displacement:** The lens tends to be displaced upwards in Marfan syndrome, but downwards in homocystinuria

Think of countless displaced crystalline lenses floating on the Thames river in London…
Name 7 causes of lens luxation/subluxation in the pediatric population:

- **Trauma**
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-- **zonule status**: Zonules are stretched in Marfan, but broken in homocystinuria

-- **lens displacement**: The lens tends to be displaced upwards in Marfan syndrome, but downwards in homocystinuria

Think of countless displaced crystalline lenses floating on the Thames river in London…
Marfan: Lens subluxed upward
Homocystinuria: Lens subluxed downward
Name 7 causes of lens luxation/subluxation in the pediatric population:

- Trauma
- Homocystinuria
- Aniridia
- Marfan syndrome
- Ectopia lentis et pupillae
- Simple ectopia lentis; Syphilis (congenital)

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

Why are zonules broken in homocystinuria?

---

**-zonal status:** Zonules are stretched in Marfan, but **broken in homocystinuria**

---

**-lens displacement:** The lens tends to be displaced **upwards** in Marfan syndrome, but **downwards** in homocystinuria
Name 7 causes in the pediatric population:

- Trauma
- Homocystinuria
- Aniridia
- Marfan syndrome
- Ectopia lentis et pupillae
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Think of countless displaced crystalline lenses floating on the Thames river in London...

Why are zonules broken in homocystinuria?
Homocystinuria involves abnormal methionine metabolism, which ultimately results in a deficiency of cysteine. Zonules are high in cysteine content, so abnormalities of cysteine metabolism tend to produce brittle zonules—and brittle zonules tend to break.

---

Syndrome and homocystinuria with respect to...

- **Zonule status**: Zonules are stretched in Marfan, but **broken in homocystinuria**
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Name 7 causes in the pediatric population:

- Trauma
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What is the classic body habitus of a homocystinuric? Hair color?

---

**zonule status:** Zonules are stretched in Marfan, but broken in homocystinuria

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

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

*Why are zonules broken in homocystinuria?*
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*What is the classic body habitus of a homocystinuric? Hair color?*
Similar to Marfan: Tall, thin, long-limbed. Additionally, they tend to be fair-haired.

---

Syndrome and homocystinuria with respect to…

- **zonule status**: Zonules are *stretched* in Marfan, but *broken in homocystinuria*
- **lens displacement**: The lens tends to be displaced *upwards* in Marfan syndrome, but *downwards* in homocystinuria
Homocystinuria
Name 7 causes in the pediatric population:

- **Trauma**
- **Homocystinuria**
- **Aniridia**
- **Marfan syndrome**
- **Ectopia lentis et pupillae**
- **Simple ectopia lentis; Syphilis (congenital)**

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

---

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Homocystinuria involves abnormal methionine metabolism, which ultimately results in a deficiency of cysteine. Zonules are high in cysteine content, so abnormalities of cysteine metabolism tend to produce brittle zonules—and brittle zonules tend to break.

**What is the classic body habitus of a homocystinuric? Hair color?**
Similar to Marfan: Tall, thin, long-limbed. Additionally, they tend to be fair-haired.

**What other problems manifest in homocystinuria?**

---

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

**MOST homocystinurics have these problems**

---

**--zonule status:** Zonules are stretched in Marfan, but **broken in homocystinuria**

**--lens displacement:** The lens tends to be displaced **upwards** in Marfan syndrome, but **downwards** in homocystinuria
Name 7 causes in the pediatric population:
- Trauma
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What is the classic body habitus of a homocystinuric? Hair color?
Similar to Marfan: Tall, thin, long-limbed. Additionally, they tend to be fair-haired.

What other problems manifest in homocystinuria?
- Mental retardation
- Osteoporosis
- Seizures
- Thrombosis of the systemic vasculature

MOST homocystinurics have these problems

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Homocystinuria involves abnormal methionine metabolism, which ultimately results in a deficiency of cysteine. Zonules are high in cysteine content, so abnormalities of cysteine metabolism tend to produce brittle zonules—and brittle zonules tend to break.

- Zonule status: Zonules are stretched in Marfan, but broken in homocystinuria
- Lens displacement: The lens tends to be displaced upwards in Marfan syndrome, but downwards in homocystinuria

Think of countless displaced crystalline lenses floating on the Thames river in London…
Name 7 causes in the pediatric population:

- **Trauma**
- **Homocystinuria**
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- **Ectopia lentis et pupillae**
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*What is the classic body habitus of a homocystinuric? Hair color?*
Similar to Marfan: Tall, thin, long-limbed. Additionally, they tend to be fair-haired.

*What other problems manifest in homocystinuria?*
- Mental retardation
- Osteoporosis
- Seizures
- Thrombosis of the systemic vasculature

*Most homocystinurics have these problems*

For more on homocystinuria, see slide-set FELT14
Name 6 causes of lens luxation/subluxation in the pediatric population:

- Trauma
- Homocystinuria
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- Zonule status: Zonules are stretched in Marfan, but broken in homocystinuria
- Lens displacement: The lens tends to be displaced upwards in Marfan syndrome, but downwards in homocystinuria

What percentage of Marfan patients develop ectopia lentis? 50 - 80

What other ocular structural abnormalities might be present?
- Microspherophakia
- Increased axial length

Give three reasons why Marfan patients tend to be high myopes:
- Increased axial length → axial myopia
- Microspherophakia → increased lens curvature → lenticular myopia
- Lens subluxation → looking through highly curved peripheral lens → spheraberration myopia

What ocular functional abnormality might be present?
- The stretched zonules make accommodation difficult—might need a reading add at an early age

Why not just bust up in there and do a clear lens extraction with IOL?
- Marfan patients are at very high risk for vitreous loss as well as RD.
Name 6 causes of lens luxation/subluxation in the pediatric population:

- Trauma
- Homocystinuria
- Aniridia
- Marfan syndrome
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What other ocular structural abnormalities might be present?

- Microspherophakia
- Increased axial length

Give three reasons why Marfan patients tend to be high myopes…

- Increased axial length → axial myopia
- Microspherophakia → increased lens curvature → lenticular myopia
- Lens subluxation → looking through highly curved peripheral lens → spherical aberration myopia

What ocular functional abnormality might be present?

The stretched zonules make accommodation difficult—might need a reading add at an early age

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

Marfan patients are at very high risk for vitreous loss as well as RD
Name 6 causes of lens luxation/subluxation in the pediatric population:
- Trauma
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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

What other ocular structural abnormalities might be present?
- Microspherophakia
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Give three reasons why Marfan patients tend to be high myopes:
1. Increased axial length → axial myopia
2. Microspherophakia → increased lens curvature → lenticular myopia
3. Lens subluxation → looking through highly curved peripheral lens → sphéral aberration myopia

What ocular functional abnormality might be present?

The stretched zonules make accommodation difficult—might need a reading add at an early age.

Why not just bust up in there and do a clear lens extraction with IOL?
Marfan patients are at very high risk for vitreous loss as well as RD.
A

- Name 6 causes of lens luxation/subluxation in the pediatric population:
  - Trauma
  - Homocystinuria
  - Aniridia
  - Marfan syndrome
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Think of countless displaced crystalline lenses floating on the Thames river in London…

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**What other ocular structural abnormalities might be present?**
--Microspherophakia
--Increased axial length

- **Homocystinuria**
- **Aniridia**
- **Marfan syndrome**
- **Ectopia lentis et pupillae**
- **Simple ectopia lentis; Syphilis (congenital)**

---

**Homocystinuria and Marfan syndrome with respect to…**
--zonule status: Zonules are *stretched* in Marfan, but *broken* in homocystinuria
--*lens displacement*: The lens tends to be displaced *upwards* in Marfan syndrome, but *downwards* in homocystinuria

---

Think of countless displaced crystalline lenses floating on the Thames river in London…
Microspherophakia. Note the small size, extreme curvature of the lens
Name 6 causes of lens luxation/subluxation in the pediatric population:

- Trauma
- Homocystinuria
- Aniridia
- Marfan syndrome
- Ectopia lentis et pupillae
- Simple ectopia lentis; Syphilis (congenital)

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

What percentage of Marfan patients develop ectopia lentis? 
50 - 80

What other ocular structural abnormalities might be present? 
- Microspherophakia
- Increased axial length

Give three reasons why Marfan patients tend to be high myopes: 
- Increased axial length → axial myopia
- Microspherophakia → increased lens curvature → lenticular myopia
- Lens subluxation → looking through highly curved peripheral lens → lens periphery myopia

What ocular functional abnormality might be present? 
The stretched zonules make accommodation difficult—might need a reading add at an early age

Why not just bust up in there and do a clear lens extraction with IOL? 
Marfan patients are at very high risk for vitreous loss as well as RD

Homocystinuria
- Zonule status: Zonules are stretched in Marfan, but broken in homocystinuria
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Name 6 causes of lens luxation/subluxation in the pediatric population:

- Trauma
- Homocystinuria
- Aniridia
- Marfan syndrome
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- Simple ectopia lentis; Syphilis (congenital)

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

Name 6 causes of lens luxation/subluxation in the pediatric population:
- Trauma
- Homocystinuria
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- Ectopia lentis et pupillae
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Think of countless displaced crystalline lenses floating on the Thames river in London…

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- Give three reasons why Marfan patients tend to be high myopes:
  -- Increased axial length → **axial** myopia
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- What other ocular structural abnormalities might be present?
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  -- Increased axial length → **axial** myopia
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  -- Lens subluxation → looking through highly curved peripheral lens → **lens periphery** myopia

- Marfan syndrome and homocystinuria with respect to…
  -- Zonule status: Zonules are **stretched** in Marfan, but **broken** in homocystinuria
  -- **Lens displacement**: The lens tends to be displaced **upwards** in Marfan syndrome, but **downwards** in homocystinuria

- Simple ectopia lentis; Syphilis (congenital)

I made this term up; a better name is probably **spherical-aberration myopia**
Name 6 causes of lens luxation/subluxation in the pediatric population:

- Trauma
- Homocystinuria
- Aniridia
- Marfan syndrome
- Ectopia lentis et pupillae
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What about the cornea in Marfan pts—does it contribute to their high myopia?

Homocystinuria

Aniridia

Marfan syndrome

Ectopia lentis et pupillae

Simple ectopia lentis; Syphilis (congenital)

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Marfan syndrome and homocystinuria with respect to…
- Zonule status: Zonules are stretched in Marfan, but broken in homocystinuria
- Lens displacement: The lens tends to be displaced upwards in Marfan syndrome, but downwards in homocystinuria

- Increased corneal curvature?

Increased corneal curvature?

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

- The stretched zonules make accommodation difficult—might need a reading add at an early age

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

Marfan patients are at very high risk for vitreous loss as well as RD

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

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

I made this term up; a better name is probably spherical-aberration myopia
Name 6 causes of lens luxation/subluxation in the pediatric population:
- Trauma
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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

Increased corneal curvature? No

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Marfan patients are at very high risk for vitreous loss as well as RD

What about the cornea in Marfan pts—does it contribute to their high myopia?
No--the cornea in Marfan tends to be flatter than normal
Marfan topography. Note the corneas are rather flat.
Name 6 causes of lens luxation/subluxation in the pediatric population:

- Trauma
- Homocystinuria
- Aniridia
- Marfan syndrome
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What about the cornea in Marfan pts - does it contribute to their high myopia?
No--the cornea in Marfan tends to be flatter than normal

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

- Homocystinuria
- Aniridia
- Marfan syndrome
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Think of countless displaced crystalline lenses floating on the Thames river in London...
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What percentage of Marfan patients develop ectopia lentis?
50 - 80

What other ocular structural abnormalities might be present?
--Microspherophakia
--Increased axial length

Give three reasons why Marfan patients tend to be high myopes:
--Increased axial length → axial myopia
--Microspherophakia → increased lens curvature → lenticular myopia
--Lens subluxation → looking through highly curved peripheral lens → lens periphery myopia

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

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- Homocystinuria
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**Marfan syndrome** and homocystinuria with respect to…

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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

See slide-set FELT14 for more on Marfan as well
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What percentage of Marfan patients develop ectopia lentis?

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What other ocular structural abnormalities might be present?

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Marfan syndrome
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Can it be effectively prophylaxed with miotics, like some other forms of pupillary-block glaucoma?

No. By stimulating the ciliary body, miotics allow the lens to move farther forward and thereby increase the risk of pupillary block

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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Marfan syndrome

Ectopia lentis et pupillae

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What is ectopia lentis et pupillae?
A genetic condition the hallmark of which is the displacement of the pupil and (microspherophakic) lens

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

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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

  In what direction are the pupils and lenses displaced?
  
  In opposite directions—pupils inferotemporal, lenses superonasal

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A genetic condition.

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