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

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

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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%

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

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--in infancy? 30
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*Identify key differences between Marfan syndrome and homocystinuria with respect to…*

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Why are zonules broken in homocystinuria?

- Zonule status: Zonules are stretched in Marfan, but broken in homocystinuria
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Why are zonules broken in homocystinuria?

- Zonules are broken in homocystinuria due to abnormalities in cysteine metabolism, resulting in brittle zonules.

What is the classic body habitus for a homocystinuric?

- Similar to Marfan: Tall, thin, long-limbed. Additionally, they tend to be fair-haired.

What other problems manifest in homocystinuria?

- Seizures
- MR
- Osteoporosis
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**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.

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

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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%

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 → sphericaberration 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?

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Homocystinuria and Marfan syndrome and homocystinuria with respect to…

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I made this term up; a better name is probably spherical-aberration myopia

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

What other ocular structural abnormalities might be present?
--Microspherophakia

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 $\rightarrow$ axial myopia
--Microspherophakia $\rightarrow$ increased lens curvature $\rightarrow$ lenticular myopia
--Lens subluxation $\rightarrow$ looking through highly curved peripheral lens $\rightarrow$ lens periphery myopia

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

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

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I made this term up; a better name is probably spherical-aberration myopia

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

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

Q

Name six causes of lens luxation/subluxation in the pediatric population:  
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Identify key differences between Marfan syndrome and homocystinuria with respect to…  
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A spectacle-corrected Marfan patient c/o difficulty reading—why?
Stretched zonules impair accommodation, so bifocals may be needed at an early age

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Homocystinuria

Aniridia

Marfan syndrome

Ectopia lentis et pupillae

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Are zonules stretched in Marfan syndrome and homocystinuria with respect to…
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**Think of countless displaced crystalline lenses floating on the Thames river in London…**

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

**Aniridia**

**Marfan syndrome**

**Ectopia lentis et pupillae**

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- 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

**Q**

- Besides refractive issues, what is the other major ophthalmic sequelae of microspherophakia?
  - Angle-closure glaucoma

- What is the mechanism?
  - The small spherical lens can block the pupil

- Can it be effectively prophylaxed with miotics, like some other forms of pupillary-block glaucoma?
  - No
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- So how can pupillary block in microspherophakia be pharmacologically prophylaxed?
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A list of causes of lens luxation/subluxation in the pediatric population:

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I made this term up, but the idea is solid

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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.

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 *supernasal*.

The pupils typically have two further abnormalities—what are they? They are very miotic, and dilate poorly; they are slit-like in shape.

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

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

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

What is simple ectopia lentis?
Simple ectopia lentis is a condition where the lens is displaced but the pupil remains in its normal position.

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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What is Marfan syndrome?
A genetic disorder characterized by skeletal, connective tissue, and cardiovascular abnormalities.

What is aniridia?
A developmental disorder characterized by the absence of the iris in the eye.
A

What is ectopia lentis et pupillae?
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What is simple ectopia lentis?
- A genetic condition

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

How common is it?
- Uncommon

Is it unilateral, or bilateral?
- Bilateral

In what direction are the lenses displaced?
- Superotemporal

How is it inherited, ie, is it AR, AD, X-linked, etc?
- AD
Name 6 causes of lens luxation/subluxation in the pediatric population:

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How common is it?
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