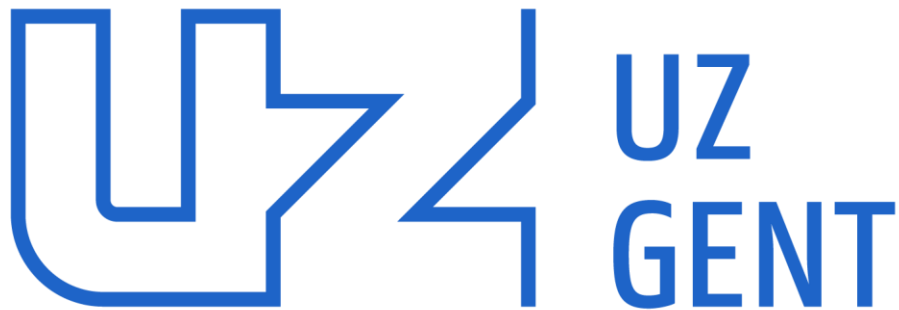
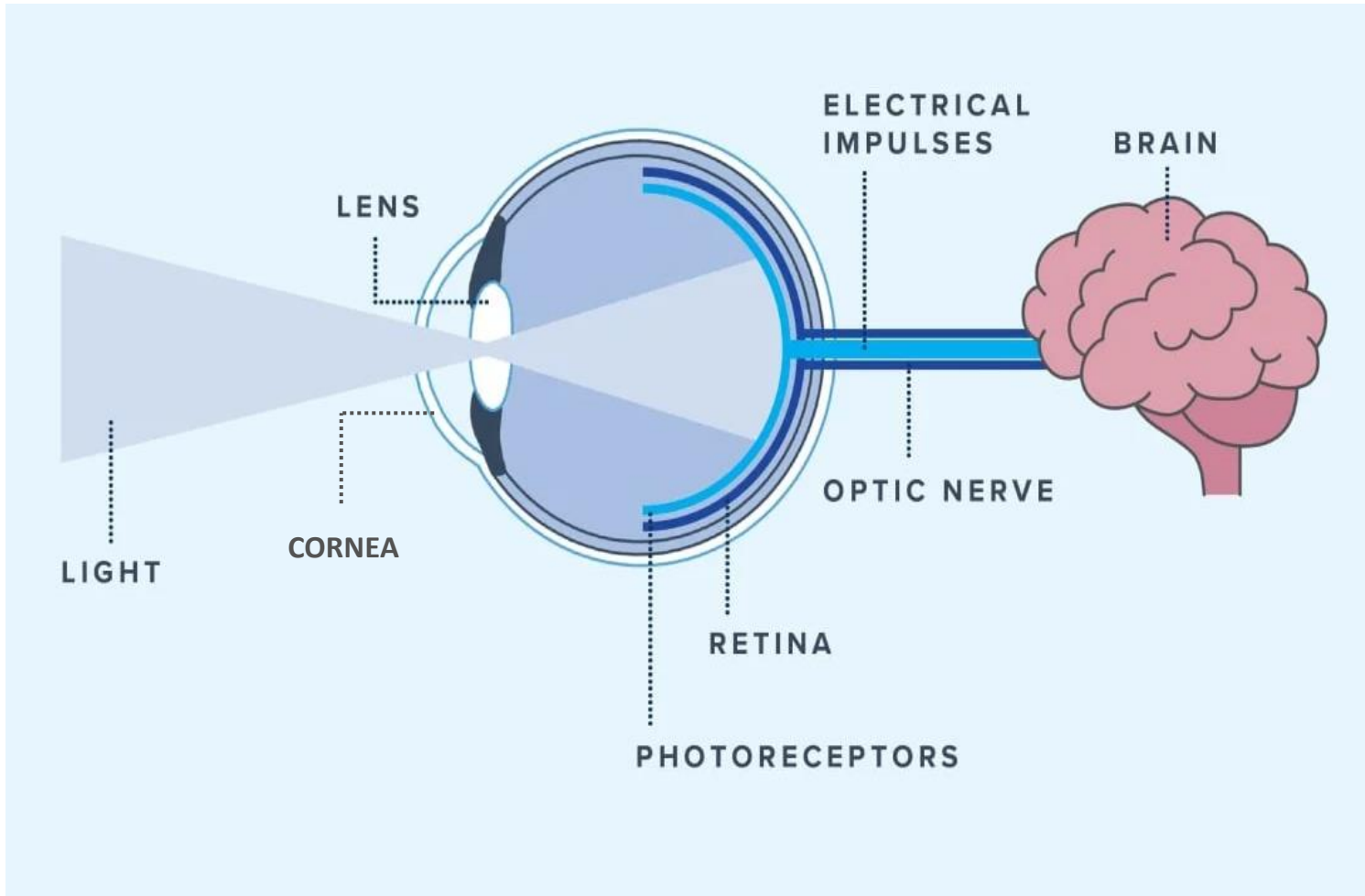


Ocular Manifestations in Cutis Laxa

Dr. L. Hoebeke



HOW VISION WORKS



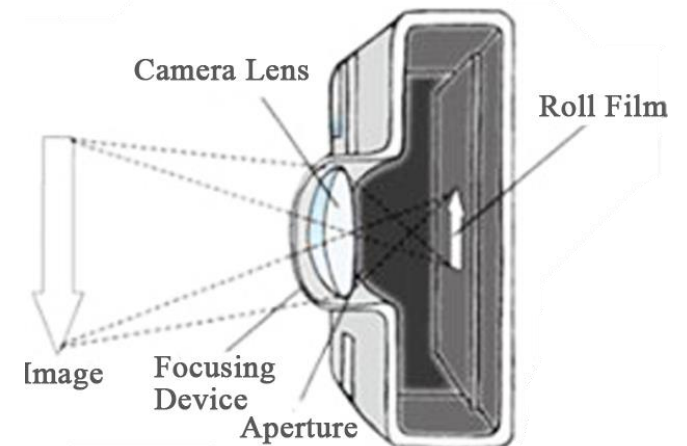
Key structural components

1/ Cornea: Clear outer layer that bends light

2/ Lens: Focuses the light onto the retina

3/ Retina: Light-sensitive tissue at the back of the eye

4/ Optic Nerve: Connects the eye to the brain



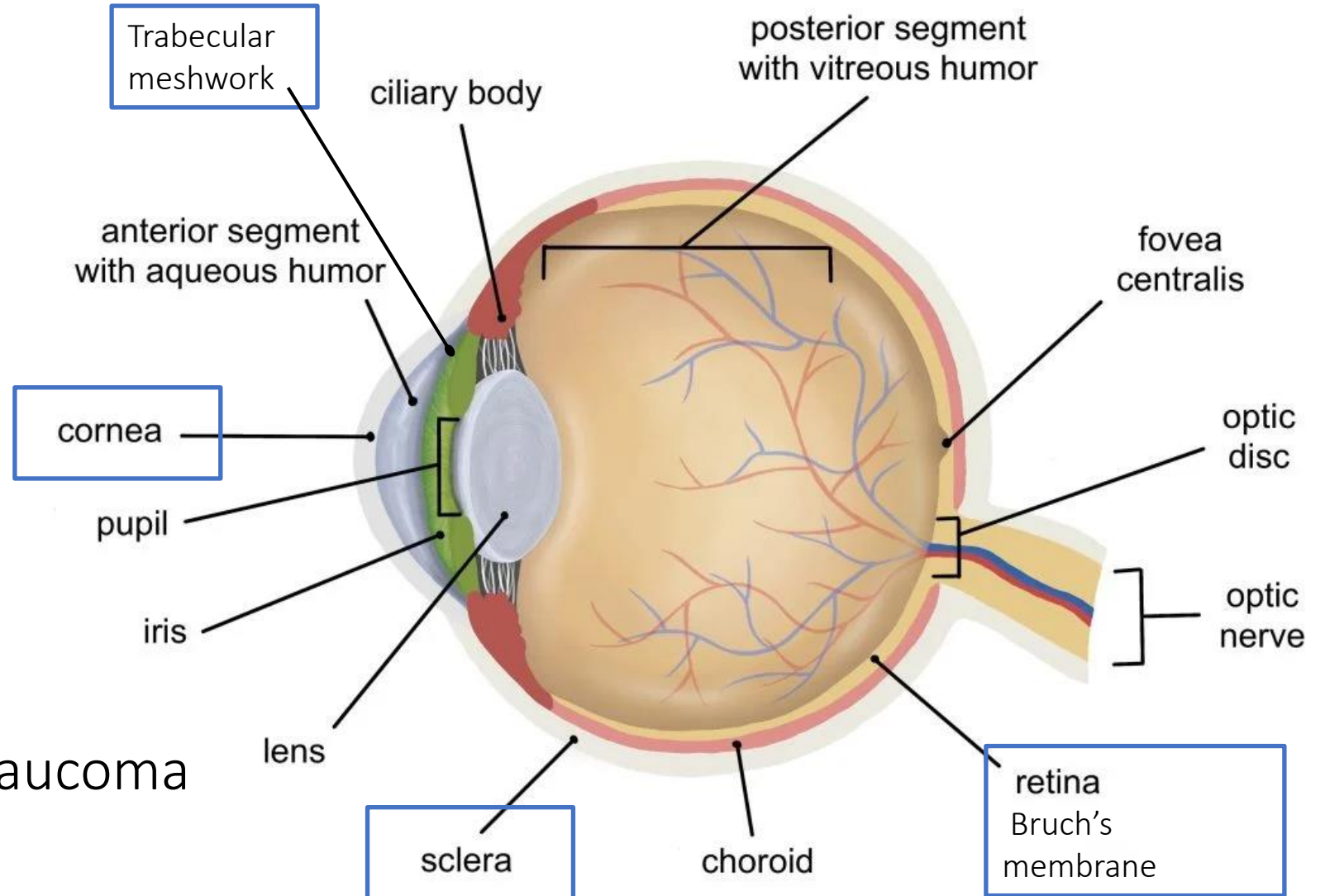
Why does cutis laxa affect the eye?



ELASTIC FIBERS contribute to ocular tissue stability

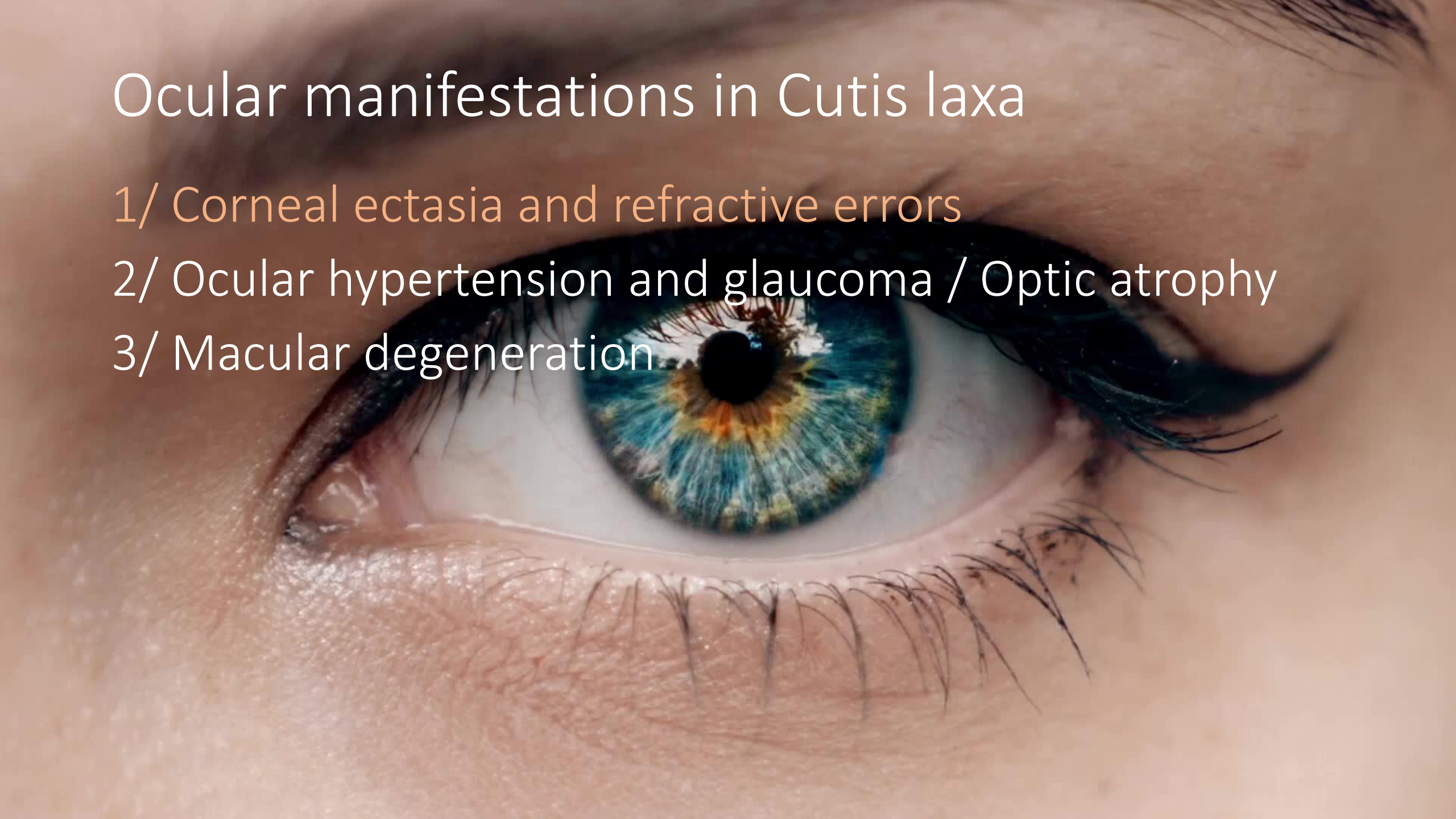
Elastic fiber
fragmentation
in cutis laxa

- 1/ Corneal ectasia
- 2/ Ocular hypertension / glaucoma
- 3/ Macular degeneration



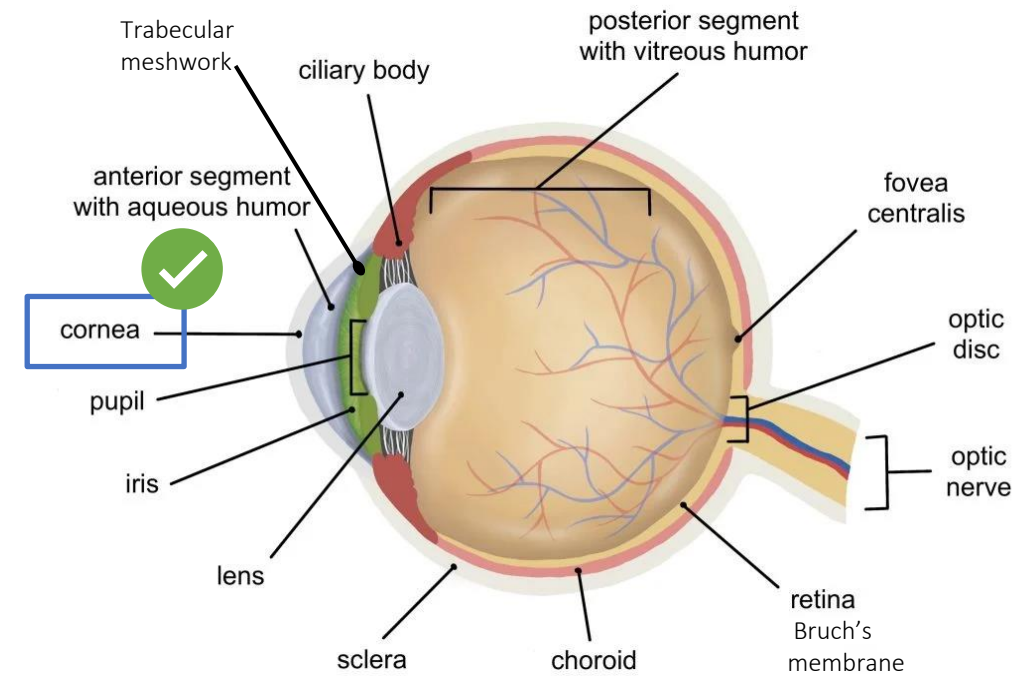
Ocular manifestations in Cutis laxa

- 1/ Corneal ectasia and refractive errors
- 2/ Ocular hypertension and glaucoma / Optic atrophy
- 3/ Macular degeneration



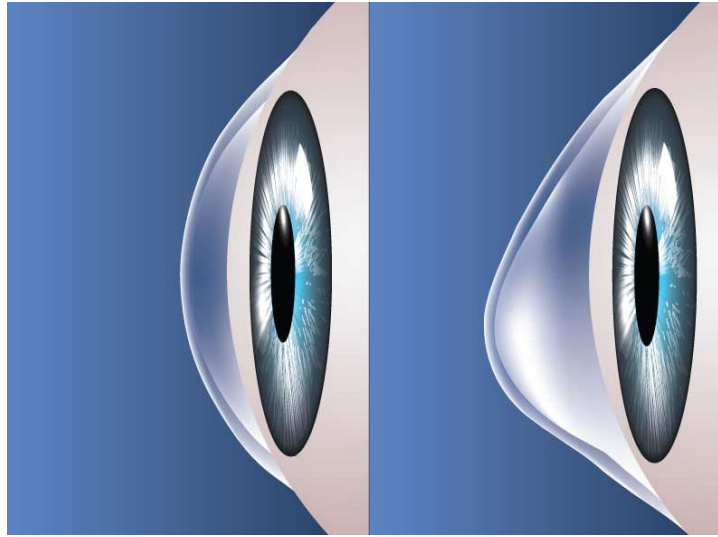
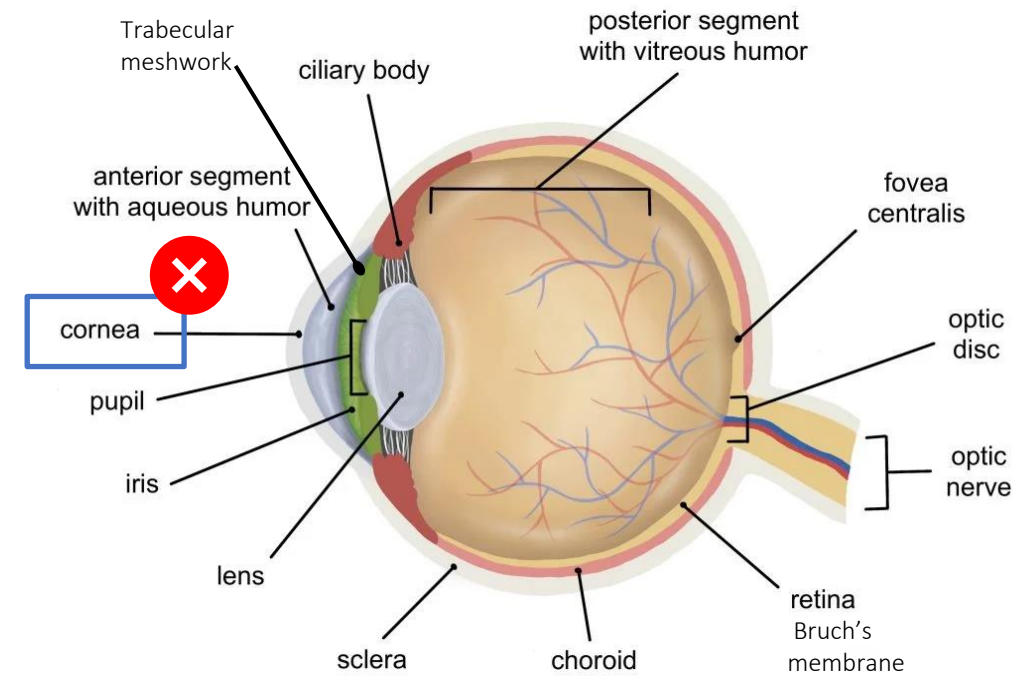
CORNEA

- > Outermost, transparent, dome-shaped layer
- > Primary refractive surface, providing two-thirds of the eye's total focusing power



CORNEA

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- > Primary refractive surface, providing two-thirds of the eye's total focusing power

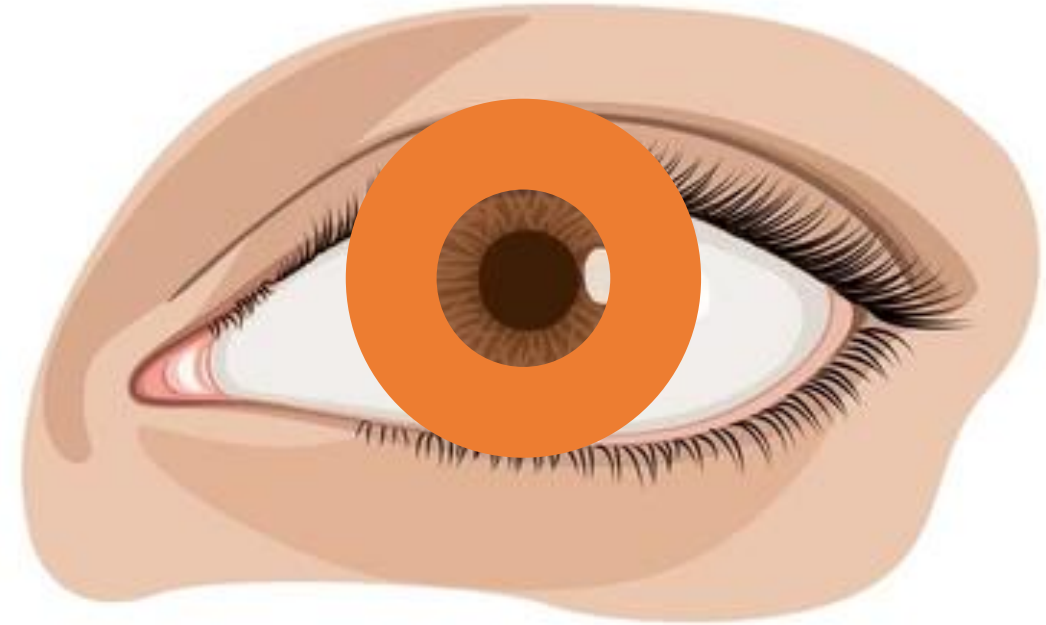


CORNEA

ELASTIC FIBRES maintain corneal shape



- True elastic fibers in the limbus and neighboring peripheral cornea
- Elaunin (elastic fibers with reduced elastin content)
- MBs (fibrillin-rich microfibril bundles with little or no elastin content)
- Limbus



CORNEA

ELASTIC FIBRES maintain corneal shape

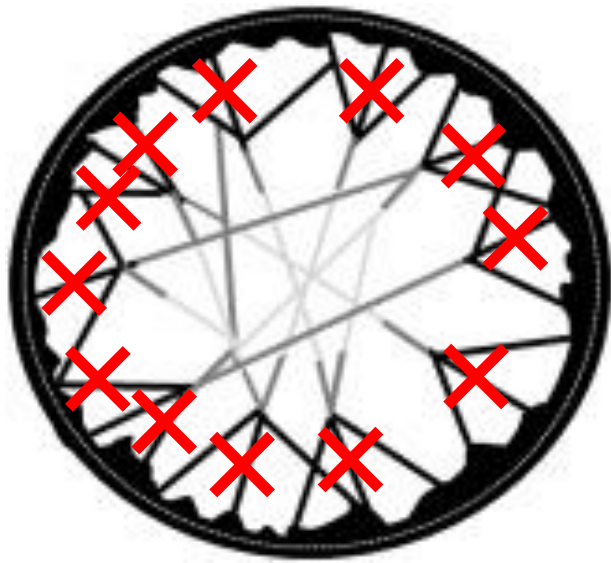


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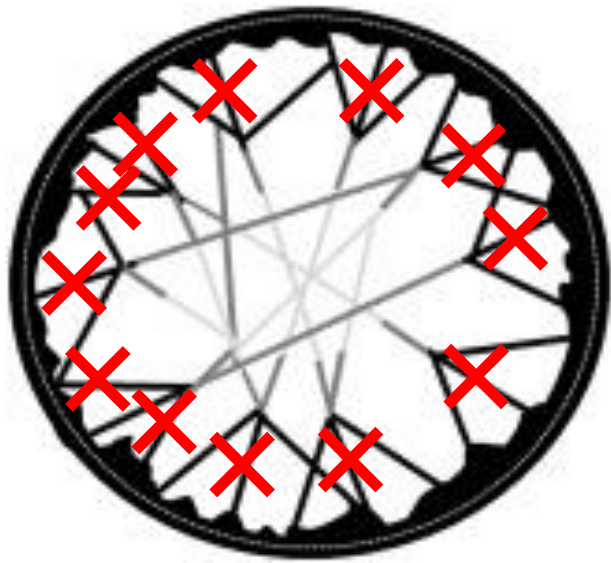


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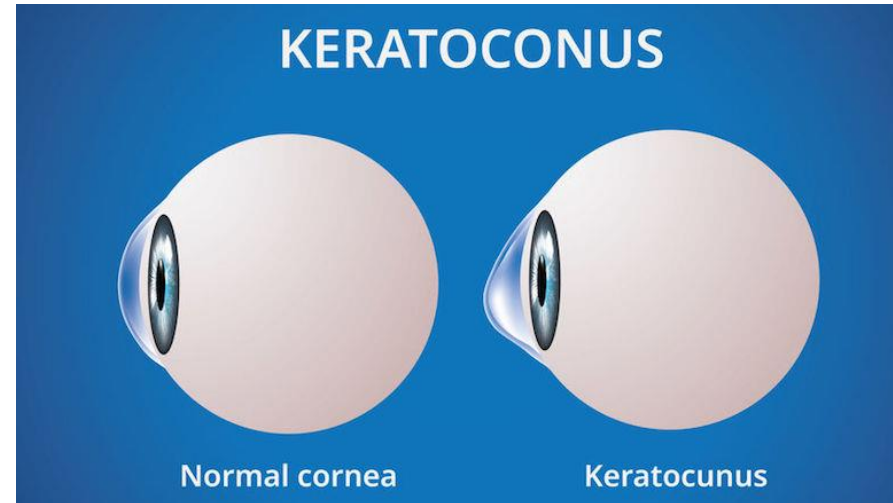
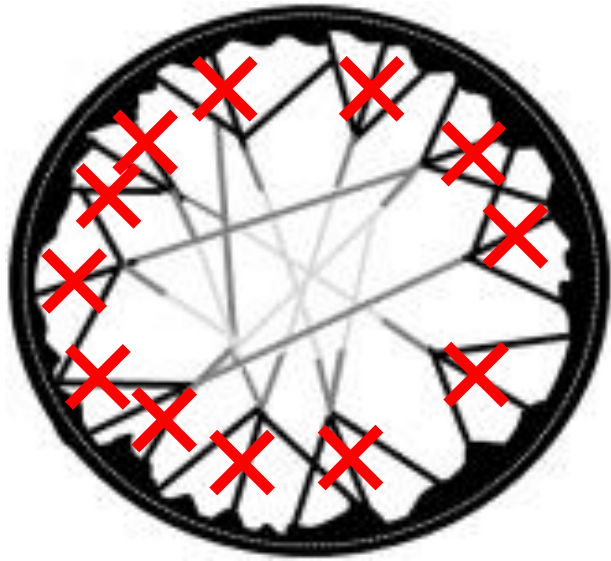
CORNEA

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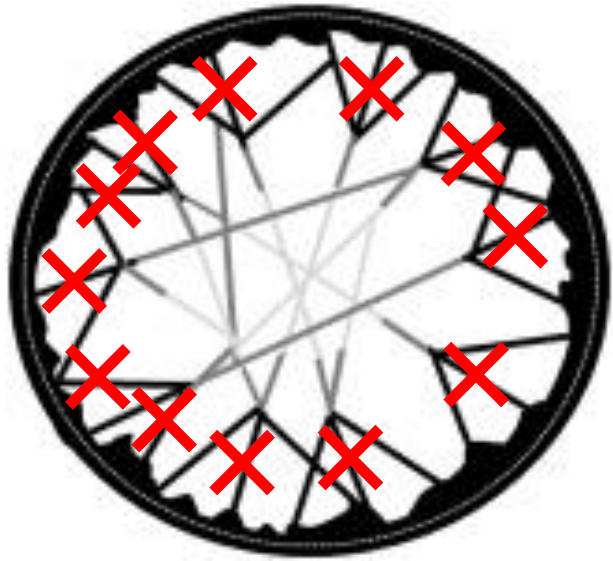
CORNEA

ELASTIC FIBRES maintain corneal shape



CORNEA

ELASTIC FIBRES maintain corneal shape



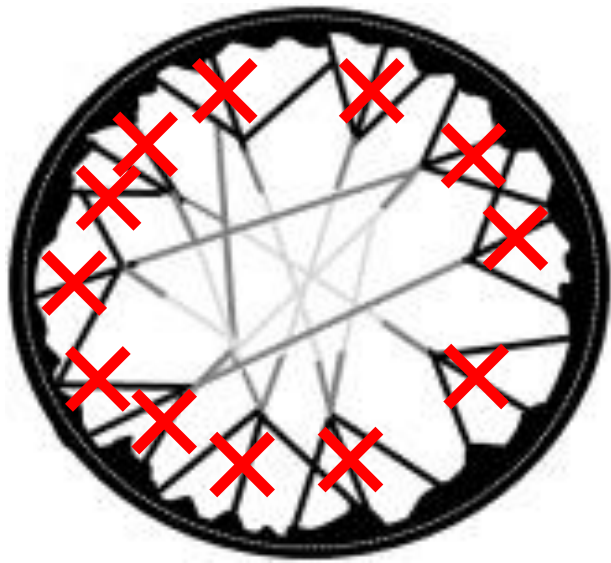
Normal eye

Keratoconic eye

Cornea plana

CORNEA

ELASTIC FIBRES maintain corneal shape



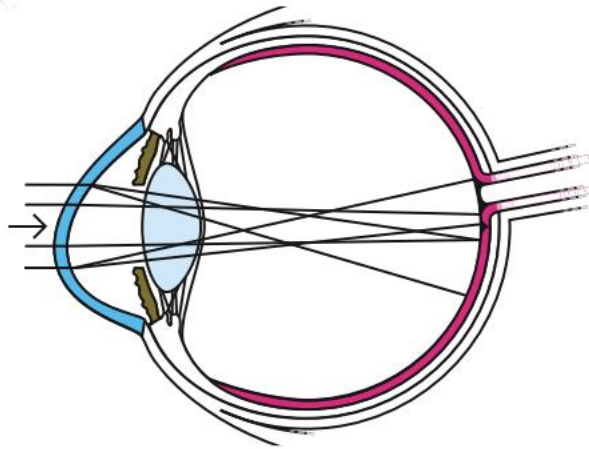
No
Astigmatism



Regular
Astigmatism



Irregular
Astigmatism



Without astigmatism



With astigmatism



Ocular Manifestations in Congenital Cutis Laxa: A Case Series

Arnaud Van Slycken, MD,* Aude Beyens, MD,†‡ Bert Callewaert, MD, PhD,†‡ and Elke O. Kreps, MD, PhD*

> In 7 of 13 patients with Cutis laxa significant alterations in corneal curvature (steep cornea, diffuse thinned or flattened cornea'), high grade astigmatisme, keratoconus was noted.

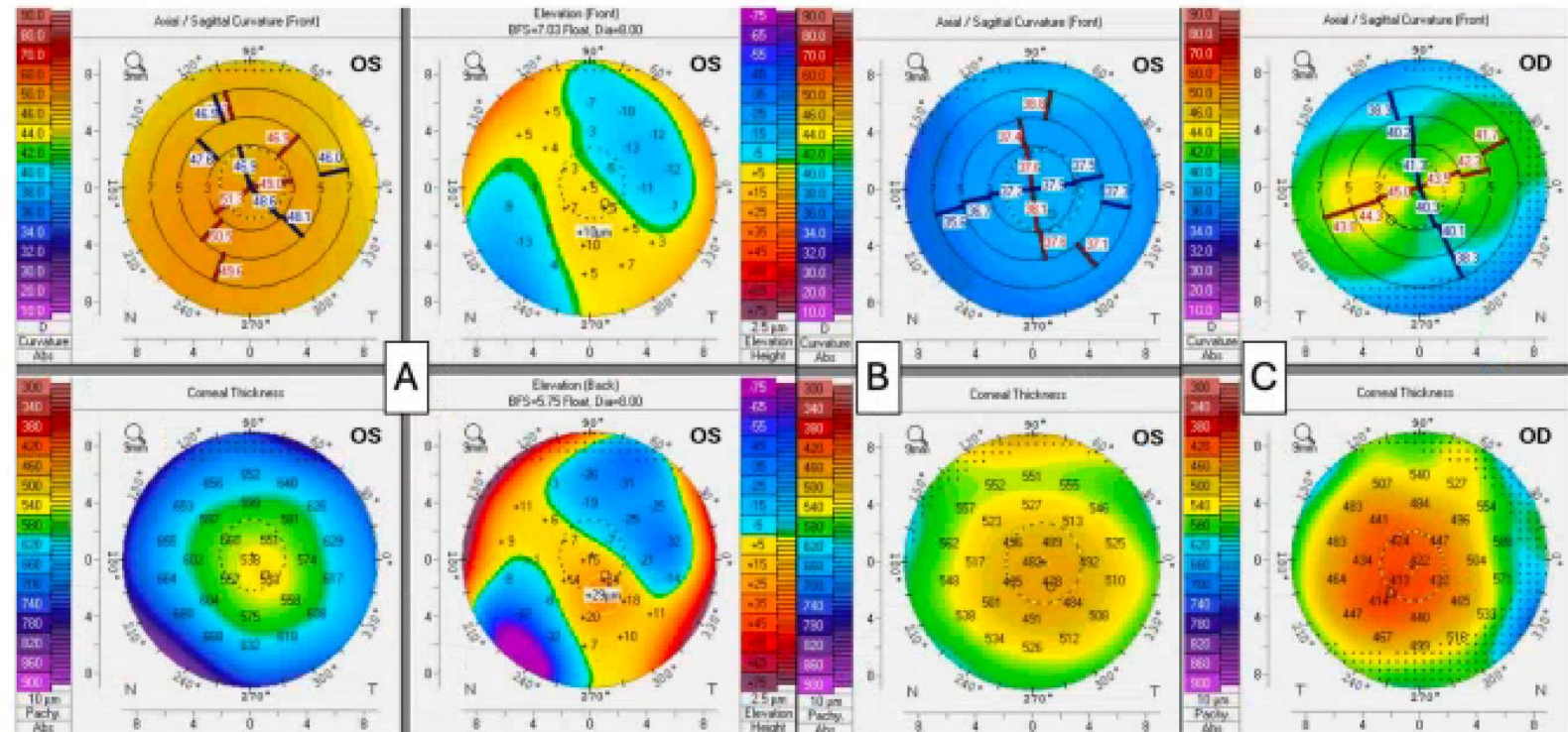


FIGURE 1. Pentacam HR measurements in patients with CL. A, The left eye of case 1 (*FBLN5*-related CL) with diffuse corneal steepening with focal corneal thinning and abnormal posterior elevation, indicative of keratoconus. B and C, Represent 2 patterns of corneal curvature findings in *ATP6V0A2*-related CL: B, the left eye of case 5 demonstrating a very flat and thin cornea. C, The right eye of case 6 is shown, with against-the-rule astigmatism and corneal thinning.

CORNEAL TREATMENT

1/ Glasses or soft contact lenses



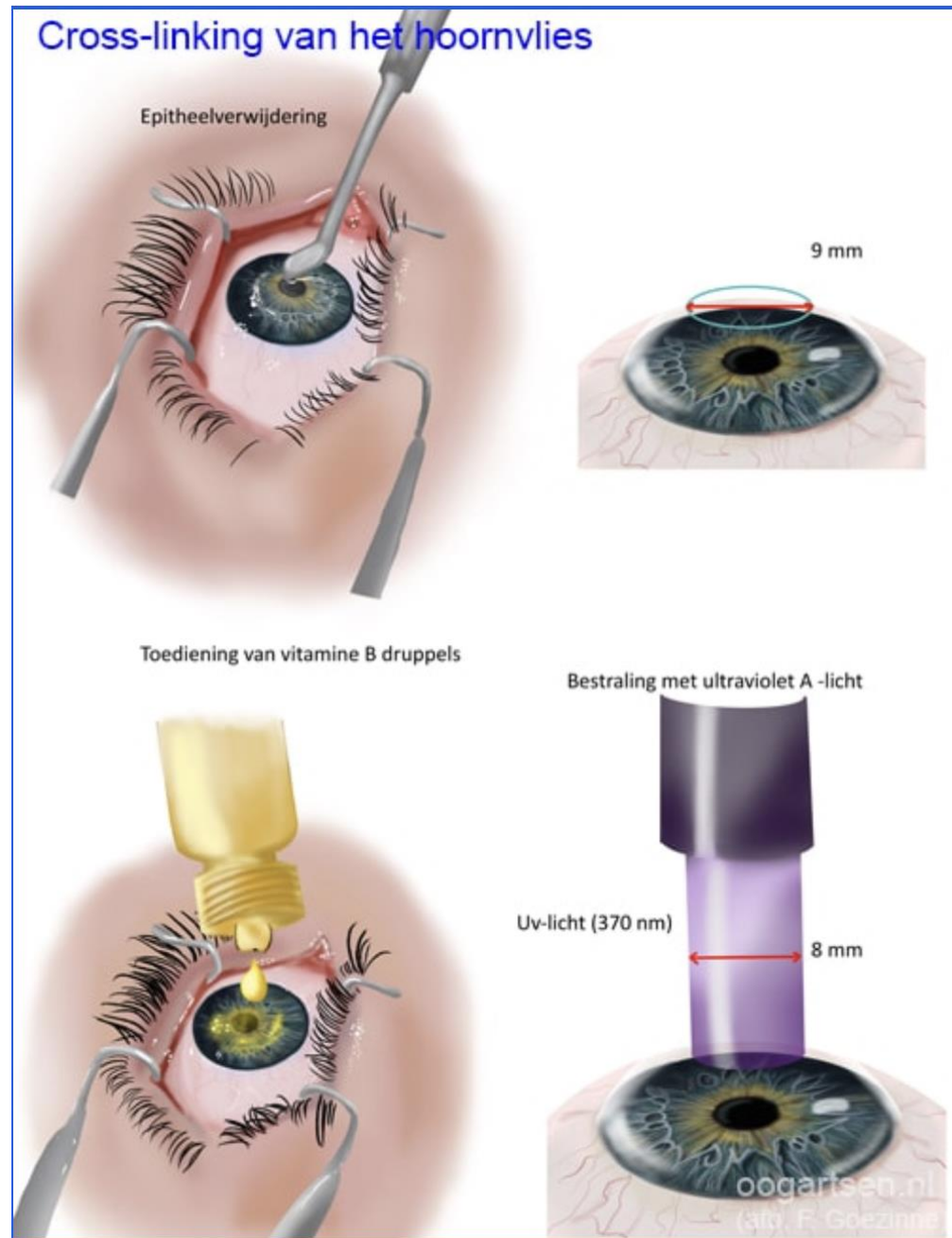
CORNEAL TREATMENT

- 1/ Glasses or soft contact lenses
- 2/ Hard contact lenses



CORNEAL TREATMENT

- 1/ Glasses or soft contact lenses
- 2/ Hard contact lenses
- 3/ UV Cross-linking



CORNEAL TREATMENT

- 1/ Glasses or soft contact lenses
- 2/ Hard contact lenses
- 3/ UV Cross-linking
- 4/ Corneal transplantation



CORNEAL TREATMENT

- 1/ Glasses or soft contact lenses
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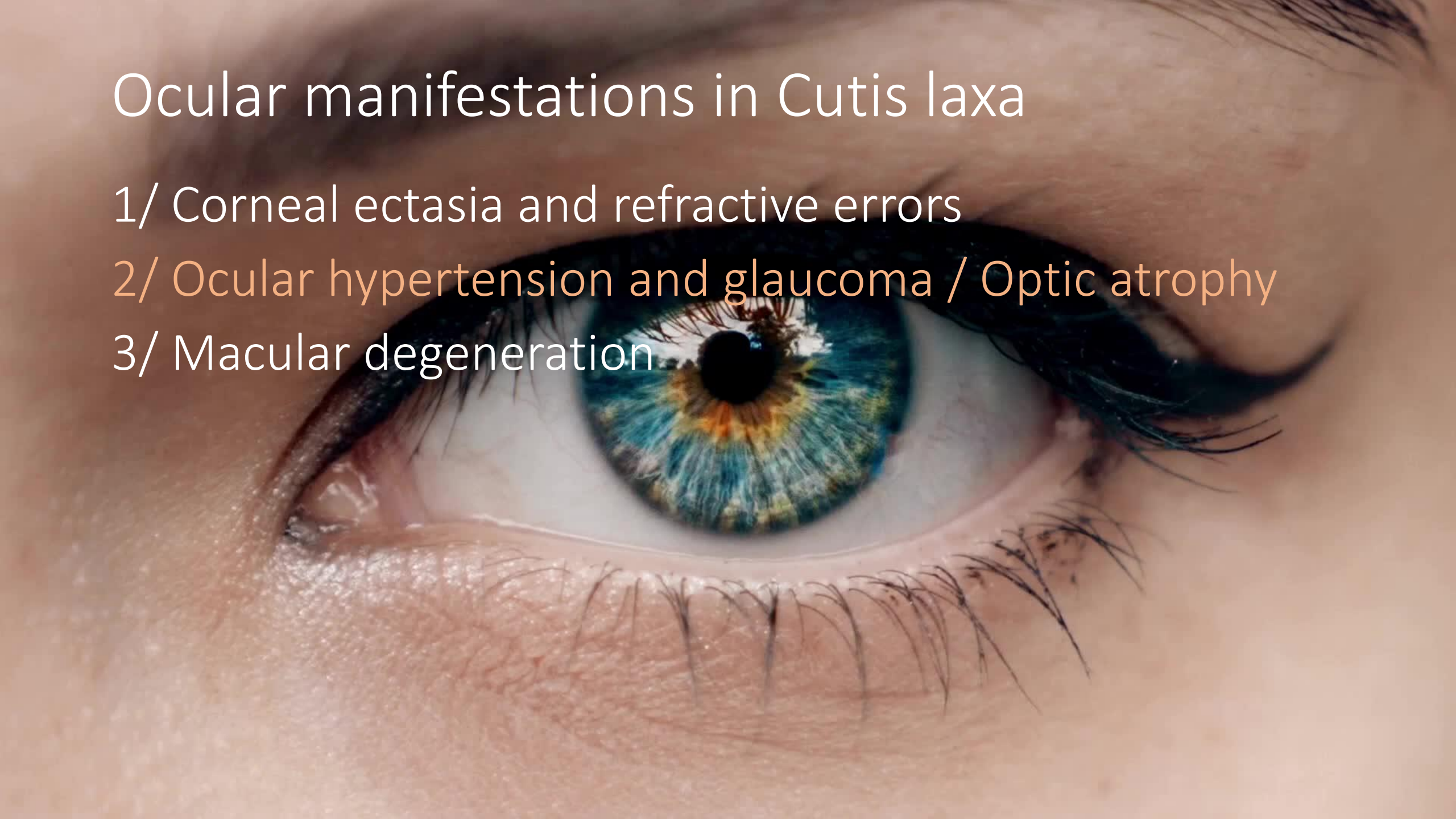
Don't rub your eyes!

Ocular manifestations in Cutis laxa

1/ Corneal ectasia and refractive errors

2/ Ocular hypertension and glaucoma / Optic atrophy

3/ Macular degeneration



Trabecular meshwork

> Complex network of connective tissue located between the iris and cornea

> Crucial for draining aqueous humor to regulate intraocular pressure

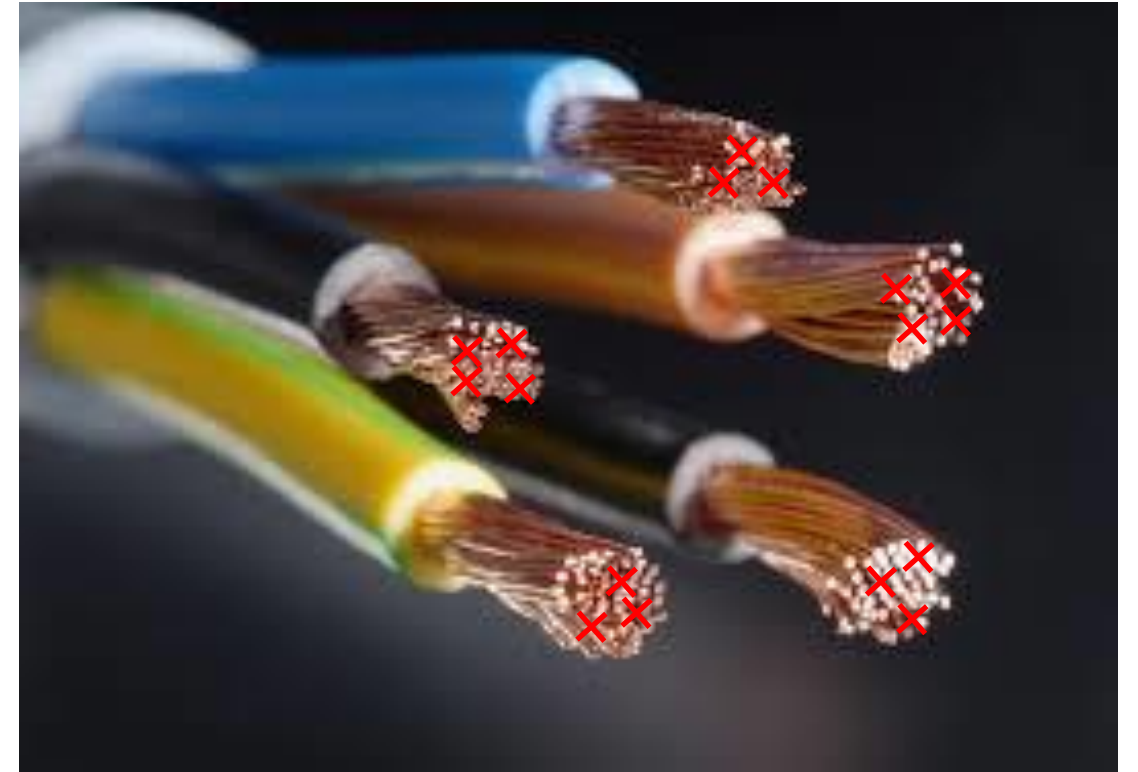
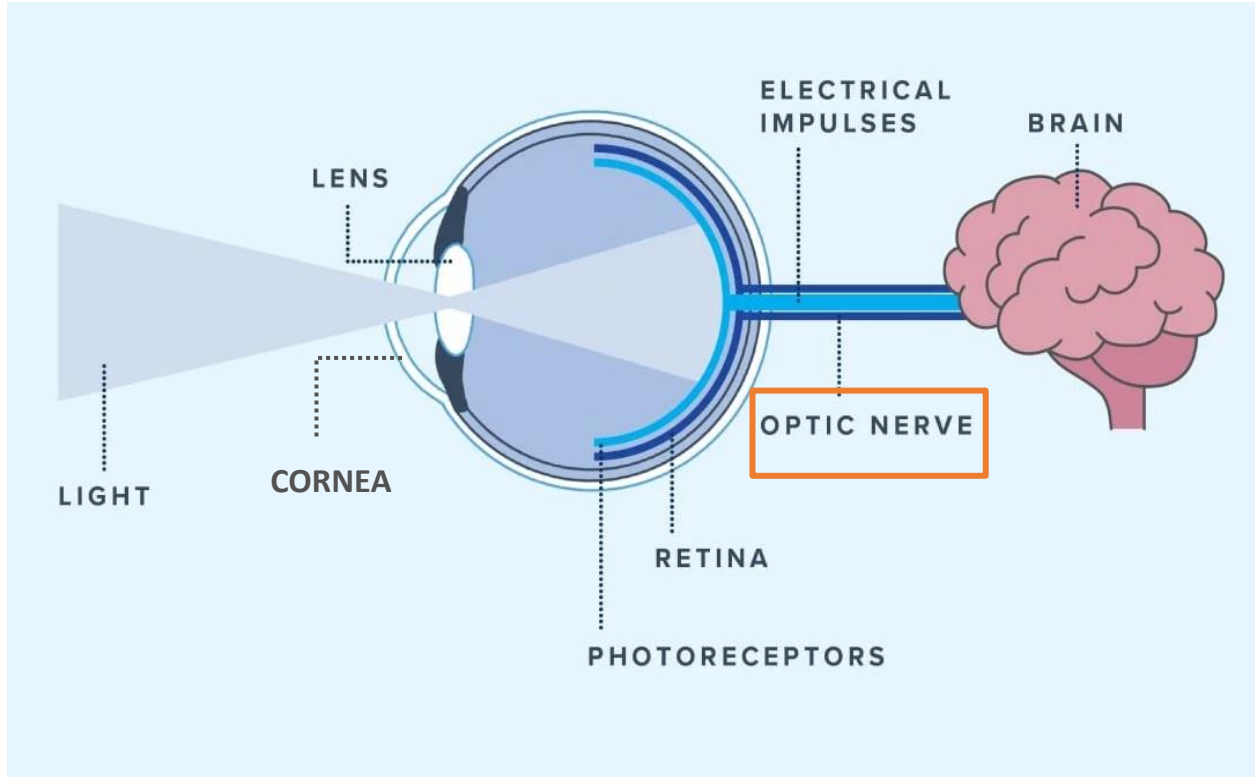
Elastic fiber
fragmentation in
Cutis laxa



Rise in intraocular pressure
Glaucoma /optic atrophy



Glaucoma



Glaucoma

Dysfunction of trabecular meshwork



Intra-ocular pressure rise



Slowly damages the optic nerve



Loss of peripheral visual field



Glaucoma treatment

- ❌ Damage is irreversible
- ✅ Prevent / slow further progression by lowering ocular pressure
1/ Drops

} Early detection is key!

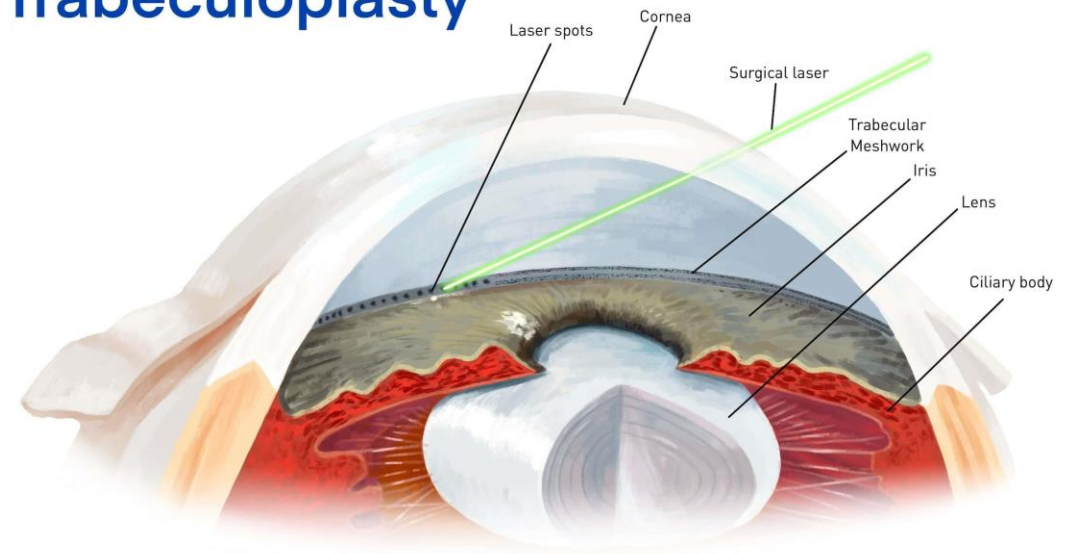


Glaucoma treatment

- ❌ Damage is irreversible
- ✅ Prevent / slow further progression by lowering ocular pressure
 - 1/ Drops
 - 2/ Laser therapy

Early detection is key!

Selective Laser Trabeculoplasty

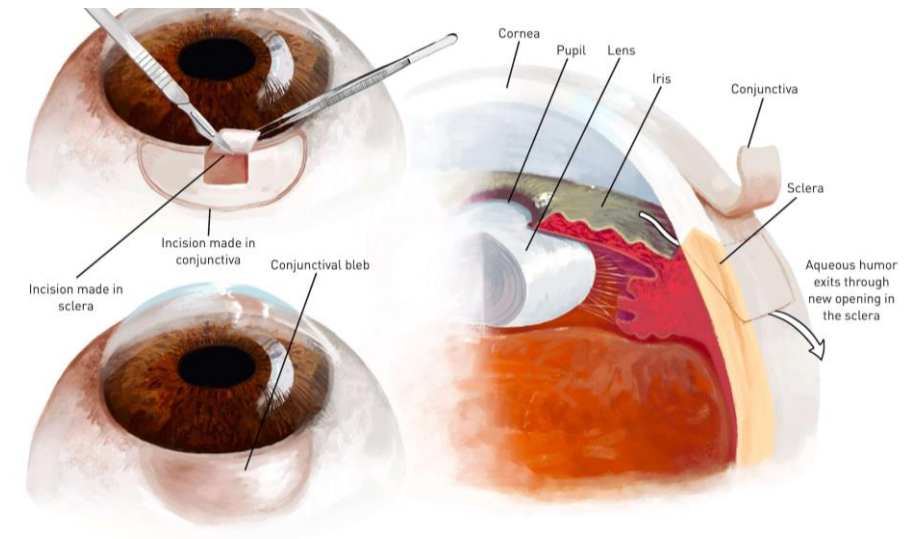
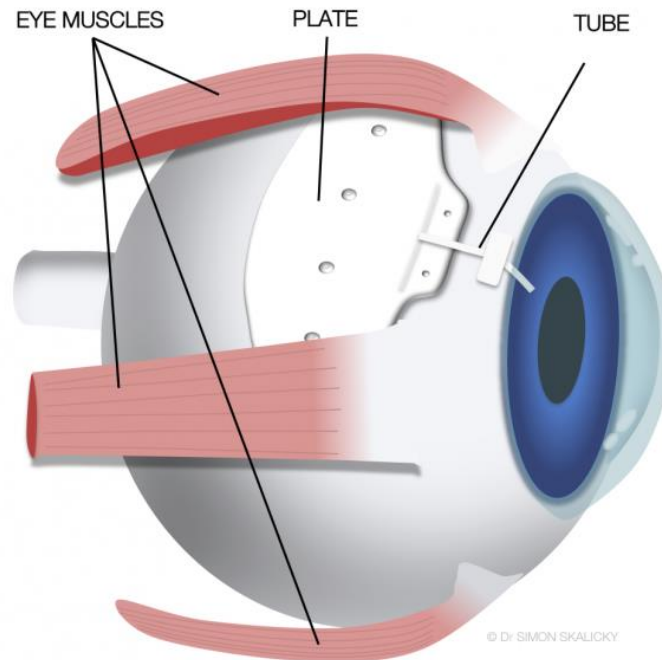


Glaucoma treatment

- ❌ Damage is irreversible
- ✅ Prevent / slow further progression by lowering ocular pressure

Early detection is key!

- 1/ Drops
- 2/ Laser therapy
- 3/ Surgery



MAIN TAKEAWAYS

- > Elastic fibers play a important role in ocular function
- > Patients with cutis laxa therefore have more risks of some ocular complications, e.g. changes in corneal shape, glaucoma, macular degeneration

Preschool ocular screening in patients with cutis laxa with a baseline ophtalmic review. Repeat eye examination every 3 years in the absence of significant findings at baseline.

ANY QUESTIONS ?



Ocular manifestations in Cutis Laxa

Dr. L. Hoebeke



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