

Case Presentation



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Patient Presentation

- CC: Fixed, dilated pupil after penetrating keratoplasty for scarred cornea, OS
- HPI: 45yo female presents with a fixed, dilated left pupil, associated with significant photosensitivity OS, 5 days after PK for her extensively scarred left cornea. The patient had a history of herpetic keratitis in the left eye which caused severe scarring, rendering poor VA at 20/300. Therefore, PK was done.

Patient Presentation (cont)

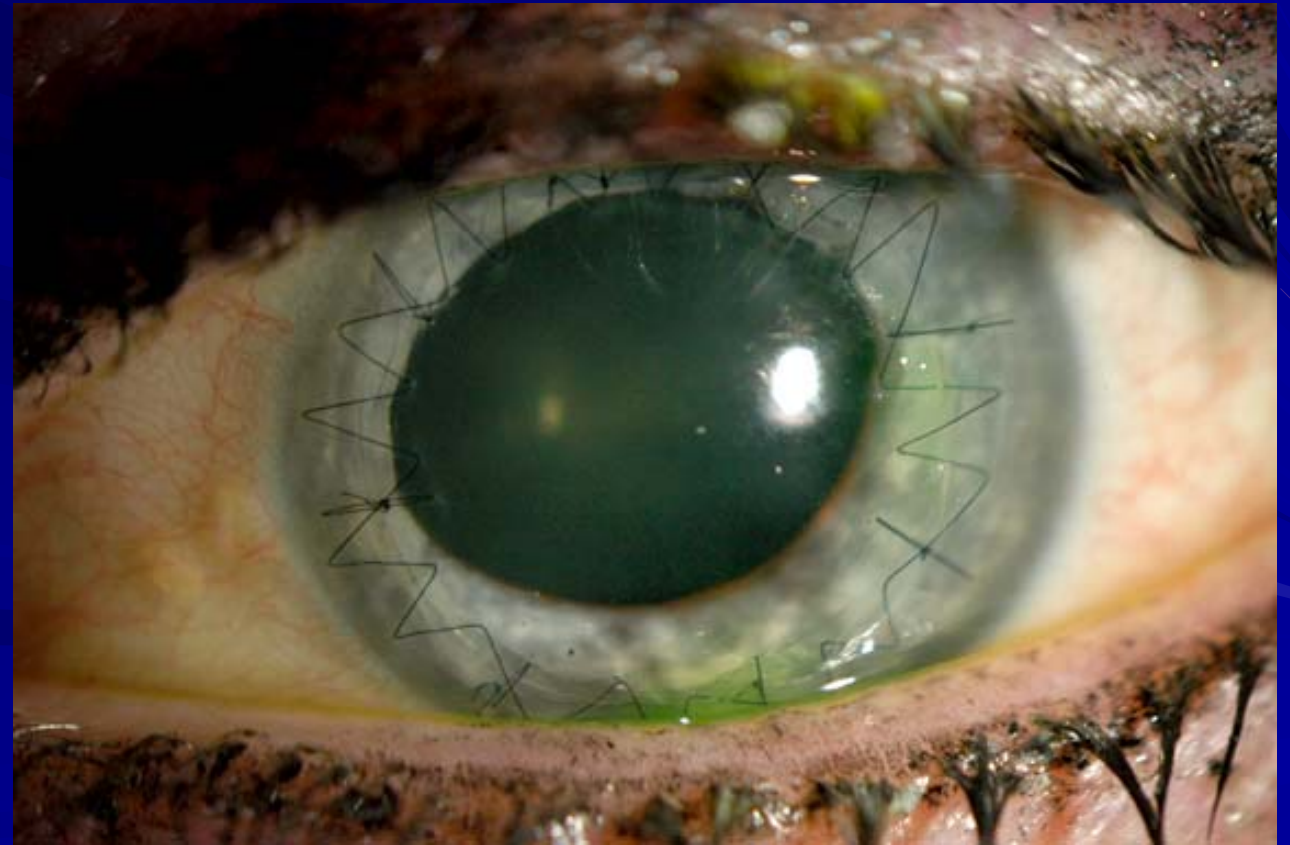
- The transplant was successful without complications. Topical steroids and antibiotics were used both pre-op and post-op. No mydriatic was used. Both continuous and interrupted sutures were employed to center the graft.

Patient Presentation (cont)

- On POD1, Pt presented with conjunctival injection 2+, corneal graft edema 2+, AC cell 2+, PERRLA; no wound leakage. VA 20/150, IOP 19. She was maintained on Pred Forte q2h and Zymar QID.

Patient Presentation (cont)

- On POD7, Pt returned with a complaint of increasing light sensitivity and 8/10 pain, OS. She also noticed in the mirror that her left pupil had been dilated over the past 2 days.
- Exam findings:
 - VA 20/125 (20/100ph)
 - IOP 46 mmHg: drops were initiated
 - Conj injection 1+, graft edema 1+, AC cell 1+
 - OS pupil diameter was fixed at 8mm, while OD pupil was PERRLA
 - Wound intact, all sutures buried



Patient Presentation (cont)

- On POD8, Pt returned with persistent light sensitivity, though the pain was considerably better. Her vision remained blurry, and her pupil was still fixed and dilated.
- Exam findings:
 - Same as POD7, except OS IOP is down to 30 mmHg: antihypertensive regimen was continued.

Patient Presentation (cont)

- POD14: Pt returned with slightly better light sensitivity and mild pain. All the post-operative parameters were improving. The only exception was the fixed, dilated pupil in the left eye, unresponsive to pilocarpine. Steroid taper began.
- Exam findings:
 - VA 20/100 (20/80ph)
 - IOP 17mmHg

What is with that Pupil? (Differentials)

■ Dilatation

- Sympathetic (excitement, fear)
- Anti-parasympathetic (ex. drugs)
- Midbrain damage (ex. grand mal)
- Increased IOP (iris ischemia)

What is with that Pupil? (cont)

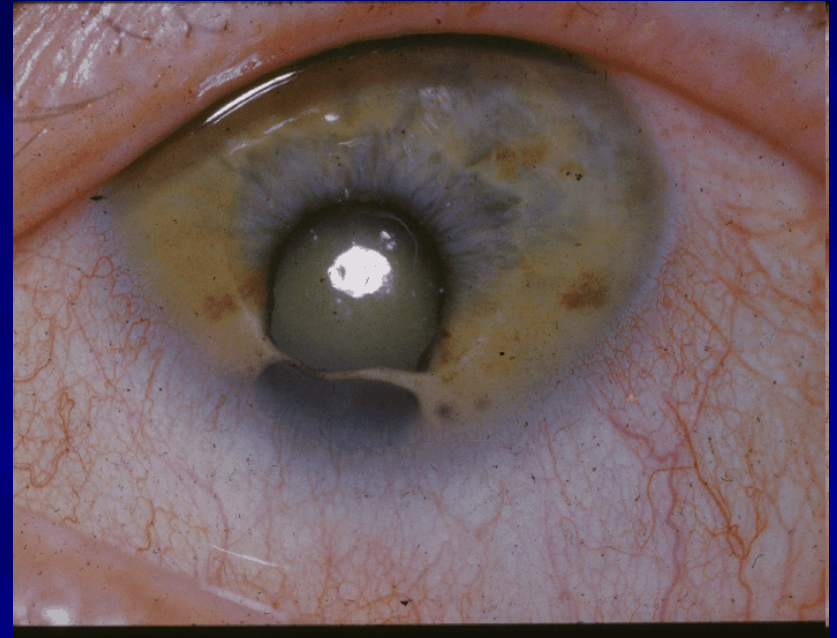
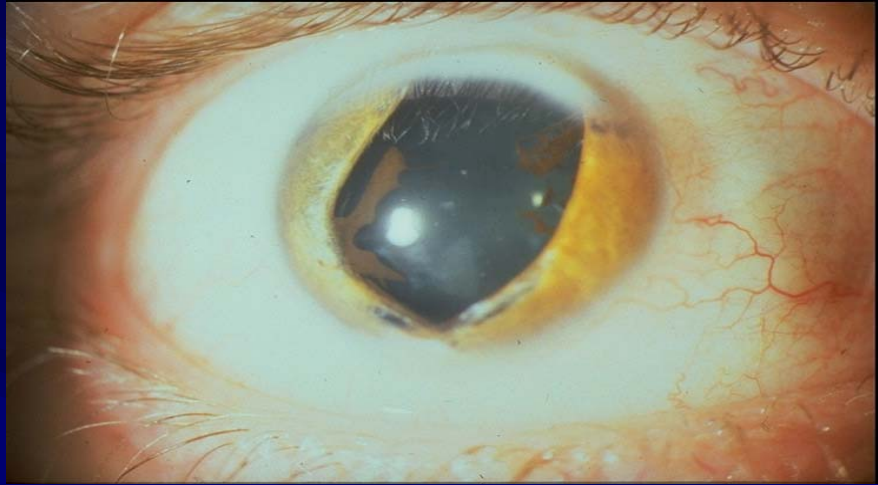
■ Constriction

- Autonomic nervous system (sleep)
- Narcotics
- Pilocarpine/phospholine iodide
- Pontine hemorrhage
- Aging

■ Irregular shape/size

- Trauma
- Inflammation
- Neovascularization
- Coloboma
- Aniridia
- s/p CE (most common)
- Tadpole pupil (migraine)
- Midbrain damage





Urrets-Zavalía Syndrome (UZS)

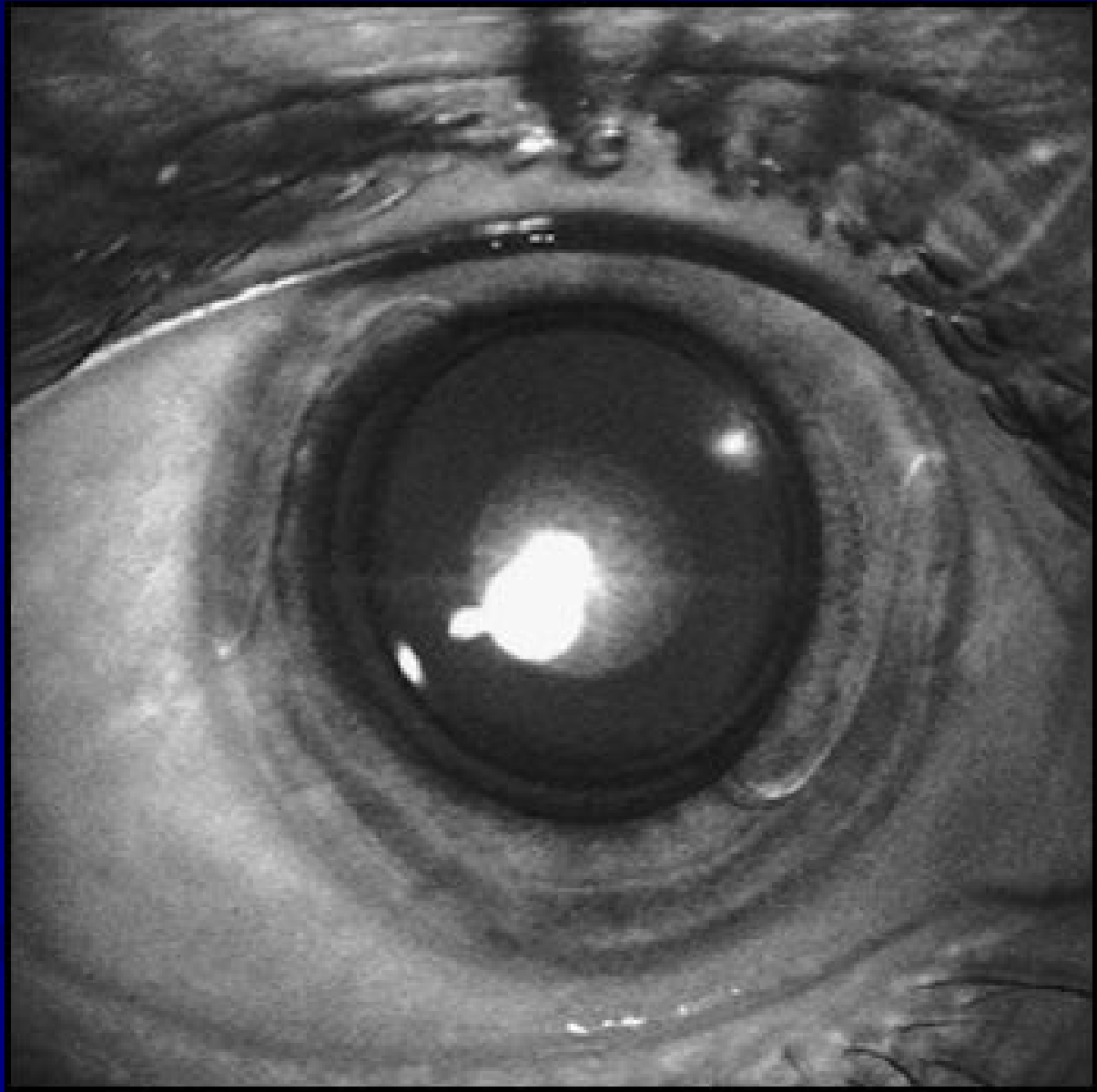
- An uncommon post-operative complication in which a pupil remains **fixed and dilated**, accompanied by **iris atrophy** and occasionally **secondary glaucoma**. These pupils are **unresponsive to miotic agents**.
- The exact features are in fact quite variable among published reports/series.

History

- First described by Dr. Urrets-Zavalía in 1963, after PK for keratoconus. He associated that with the post-op mydriatic treatment. He attributed this discovery to Dr. Castroviejo in the 1940s.
 - Dr. U-Z: 6 patients had the syndrome for up to 6 weeks, but all resolved spontaneously.
 - In 1967, Uribe et al described the syndrome in PK patients without post-op mydratics.

History

- Classically linked to PK for keratoconus
- Subsequent reports suggest that UZS can also be seen after:
 - Deep lamellar keratoplasty for keratoconus
 - Descemet stripping endothelial keratoplasty for Fuch's endothelial dystrophy
 - Argon laser peripheral iridoplasty
 - Surgical trabeculectomy
 - Phakic ACIOL implantation



Epidemiology

- Incidence: 2.2% – 17.7%
- This seems to be decreased in the recent years; some even question its continued existence.
- Likely due to improved surgical techniques and/or different diagnostic standards.

Variable Degrees of Dilatations

- Typically, 3 types of pupil dilatations can be seen in UZS:
 - At least 1.5mm larger than the fellow unoperated eye, but responds to miotic agents (90%)
 - Unreactive and paretic, but slowly returns to normal after time (some up to 1 yr)
 - Irreversible dilatation with iris atrophy

Possible Mechanisms

- Strong intra-op mydriasis brings iris into contact with peripheral cornea, producing peripheral anterior synechiae and glaucoma (Dr. U-Z)
- Relative pupillary block
- Pre-existing pupillary abnormalities
 - Hyperreaction to mydriatics, as seen in Keratoconus and Down's Syndrome
 - Iris plataeu syndrome
- Intraoperative trauma causing strangulation of iris vessels

Possible Mechanisms (cont)

- Intra-op IOP elevation causing occlusion of scleral vessels which leads to iris ischemia
- Damage to ciliary ganglion during sub-tenon injection of anesthetics
- Abnormalities of the sympathetic nervous system
- Overuse of general anesthesia
- Intra-op exposure to other toxins

Treatment/Prevention

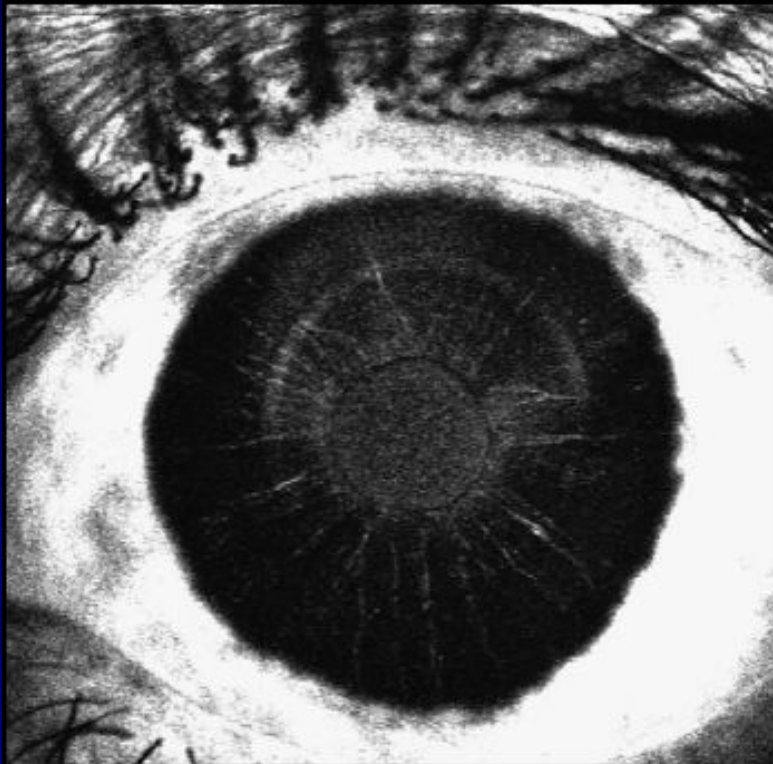
- Avoid using mydriatic agents following surgery in vulnerable eyes (keratoconic).
- Laqoutte (1983) proposed a regimen of sympatholytic drop (guanethidine) q4h for 1 day, followed by pilocarpine 2% for several days.
- Naumann (1997) recommended performing PI in all phakic patients undergoing PK.
- Special surgical techniques of PK have also been proposed to prevent iris damage (Loden and Price, 1998).

What Happened to the Patient?

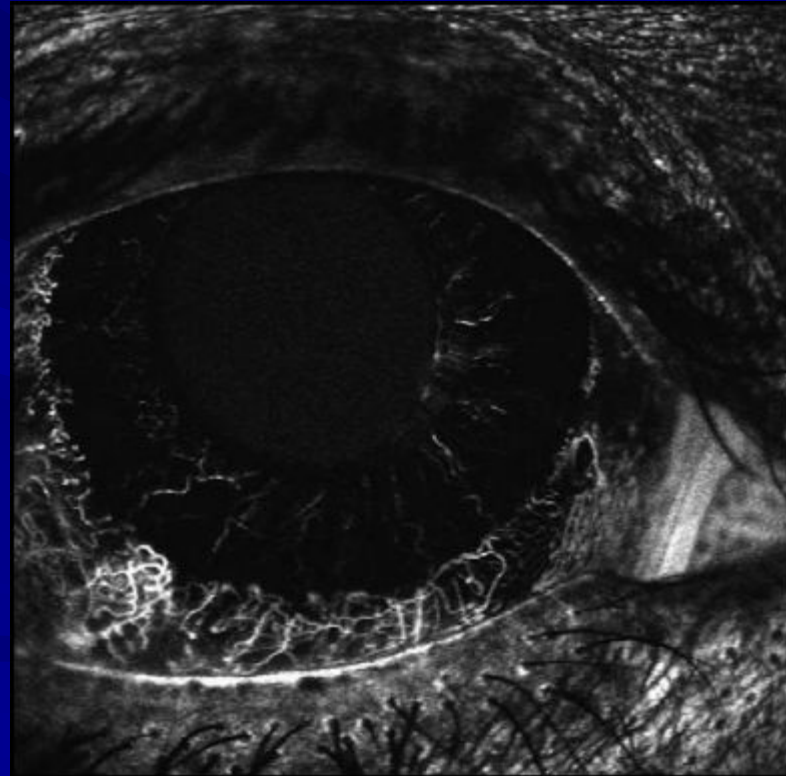
- After 6 months, Pt returned with OS VA of 20/40. There was no evidence of inflammation or graft failure. IOP had remained in the normal range, so anti-glaucoma drops were discontinued.
- However, the patient had significant glare (no photophobia or pain), as the OS pupil was still fixed and dilated, with no improvement.

What Happened to the Patient? (cont)

- Iris fluorescein angiography disclosed large area of non-perfusion in the left iris.



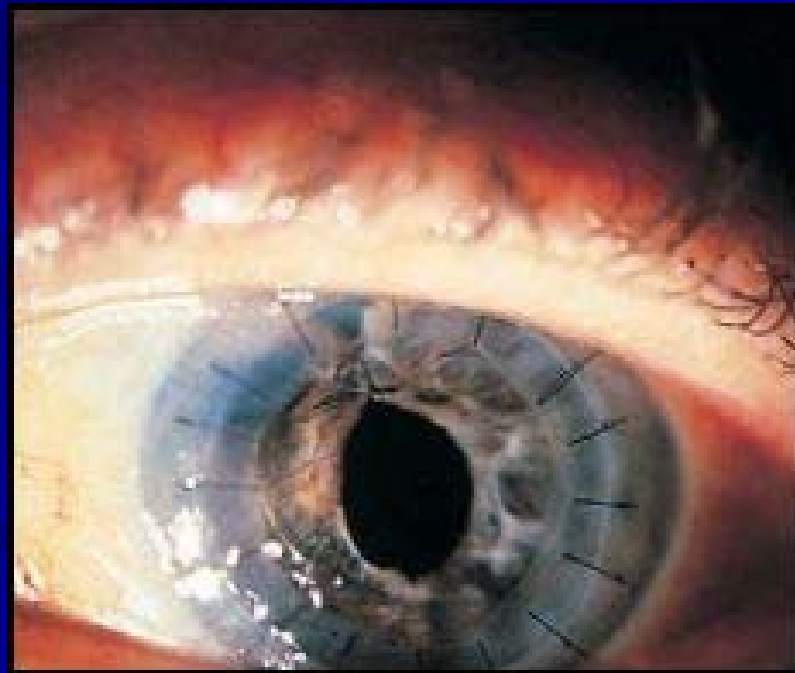
OD



OS

End of the Case

- Finally, decision was made to reconstruct the iris. Afterwards, her left eye had no more glares.



References

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