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.
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.
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.
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
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
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-Zavalia 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.
End of the Case

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


