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Cracking the ICE: Insights into the Enigmatic Syndrome

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Introduction

- Rare, acquired, unilateral endothelial disorder Typically affects young to middle-aged women
- Characterized by abnormal corneal endothelium, iris atrophy, and secondary angle-closure glaucoma

Pathophysiology

- Metaplastic endothelium migrates over the **trabecular meshwork** and **iris**, forming a **contractile membrane**
- Causes **peripheral anterior synechiae (PAS)**, iris distortion, and progressive IOP rise

Clinical variants

Variant

Hallmark Feature

Progressive Iris Atrophy (PIA)

Stromal loss, corectopia, iris holes

Cogan–Reese Syndrome (CRS)

Iris nodules (pigmented)

Chandler's Syndrome

Corneal edema predominant

A 29-Year-Old Female with Unilateral Glaucoma

Chief complaint: Blurred vision and mild photophobia (left eye)

History: No trauma, inflammation, or previous surgery

Visual acuity: Normal OD / slightly reduced OS

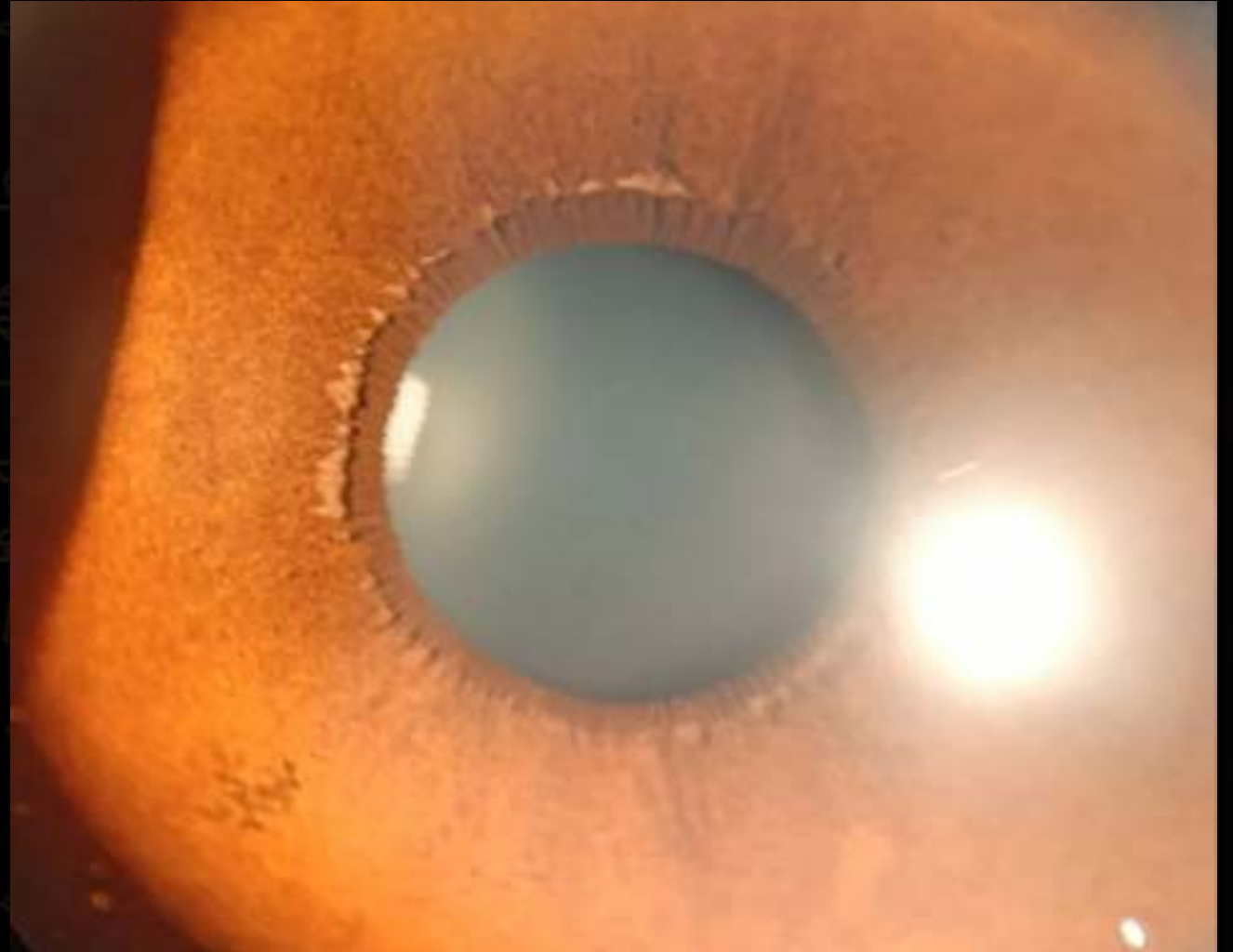
Initial IOP: 40 mmHg (OS)

After topical therapy: 20 mmHg on triple drops

Anterior segment: Clear cornea, early patchy iris atrophy, some PAS

Posterior segment: Cupping OS, normal disc OD

Fellow eye: Completely normal



Slit-Lamp and Gonioscopic Features of the Affected Eye

Clear cornea — no guttae or edema

High iris insertion and patchy stromal atrophy around the pupil

Early **corectopia** toward **PAS** formation

Multiple focal **peripheral anterior synechiae** (3–9 o'clock)

Fellow eye completely normal



Ultrasound Biomicroscopy (UBM) of the Left Eye

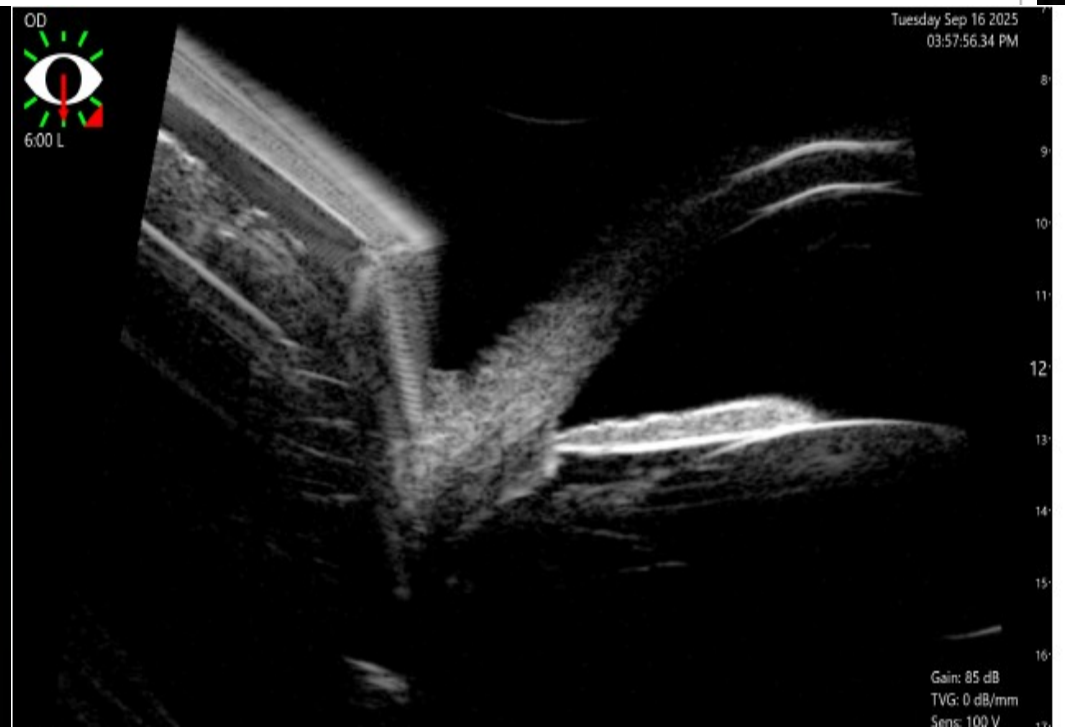
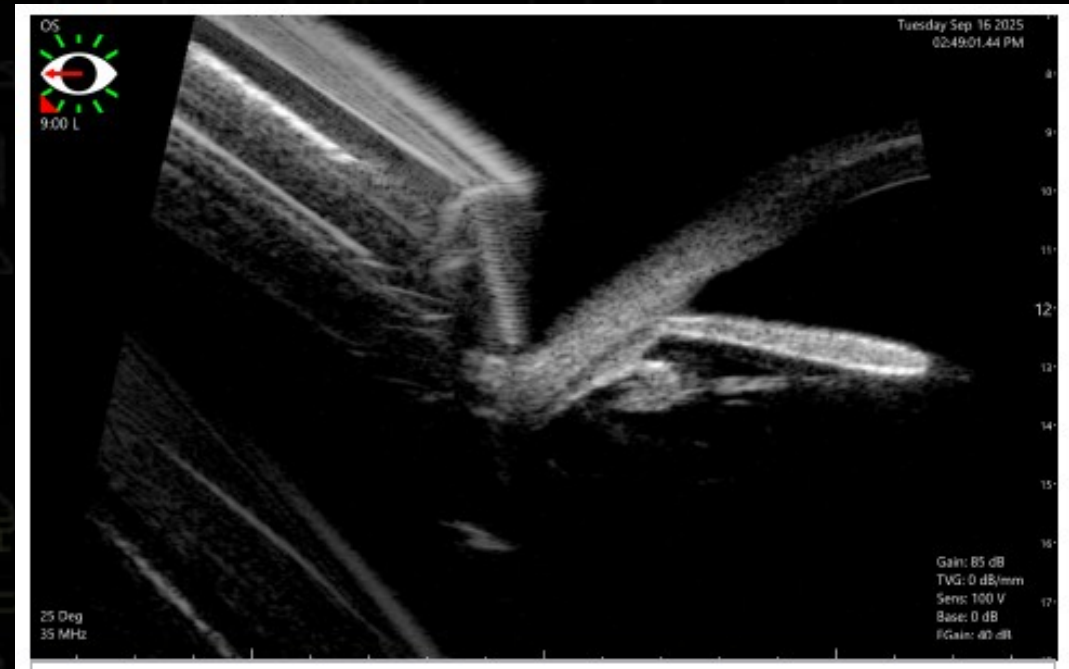
Deep central anterior chamber

High iris insertion on the anterior surface of the ciliary body

Broad peripheral anterior synechiae (PAS) inferiorly and temporally

No ciliary body cysts or plateau iris configuration

Fellow eye: completely normal anatomy



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Patient: MONIR, LOREN
Physician:
Operator:
Disease:

Watany Eye Hospital

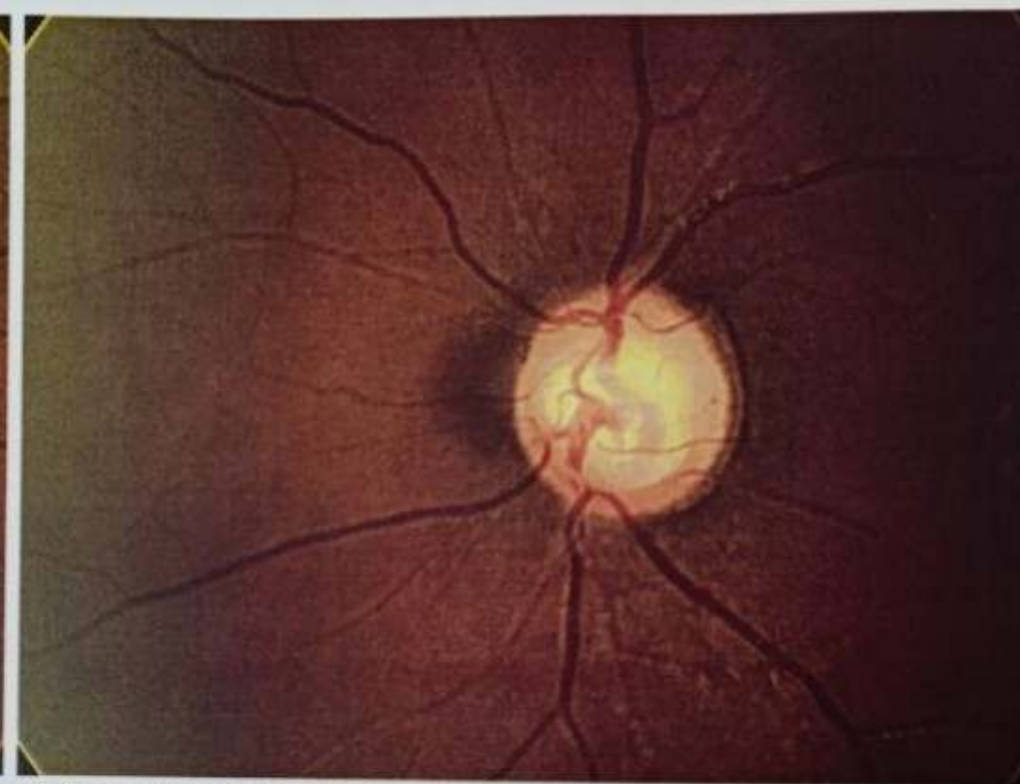
Gender: Female
ID: 1329437

Exam Date: 09/25/2024
DOB(age): 07/13/2004 (20)
Ethnicity: Caucasian
Algorithm Ver:

Multi Photo View



OD Disc 09/25/2024 15:01:51



OS Disc 09/25/2024 15:04:08



☐ Overlay

Mouse Buttons
Drag Left : Brightness / Contrast
Drag Right : Pan Image
Scroll Wheel : Zoom Image

Differentiating ICE from Axenfeld–Rieger Spectrum

Feature

Laterality

Inheritance

Age of onset

Iris changes

Cornea

Angle

Systemic findings

ICE Syndrome

Usually *unilateral*

Sporadic, acquired

Early to middle adulthood

Progressive atrophy, corectopia,
PAS

Abnormal endothelium
(metaplastic)

Progressive PAS from endothelium

None

Axenfeld–Rieger Spectrum

Typically *bilateral*

Autosomal dominant

Congenital / childhood

Hypoplasia, holes, peripheral
strands

Posterior embryotoxon

Developmental anomaly

Craniofacial / dental / periumbilical
defects

Final Diagnosis

- **Early Progressive Iris Atrophy (PIA) Variant of Iridocorneal Endothelial (ICE) Syndrome**
- – Unilateral (Left Eye)
 - Secondary Angle closure Glaucoma
 - Currently Controlled on Triple Topical Therapy

Current Management and Counseling

- **Topical IOP control:** triple therapy (prostaglandin analogue, beta-blocker, CAI)
- **Current IOP:** 20 mmHg, stable for >12 months
- **Optic nerve & corneal monitoring:** every 6 months
- **Gonioscopy:** monitor for PAS progression
- **Specular microscopy:** baseline and annual
- **Counseling:** nature of ICE, risk of progression, surgical options if uncontrolled

Surgical Approaches and Outcomes in ICE-Related Glaucoma

Study	Year	Surgical Technique	Key Outcome
İmamoğlu et al.	2017	Trabeculectomy vs Tube Surgery	Tube success rate 71% vs 29% for trab ($p < 0.05$); long-term IOP control better with tubes
Mao et al.	2021	AGV with MMC (enhanced protocol)	Improved 5-year survival vs conventional AGV
Zhang et al.	2023	DSAEK + <i>Spokewise Iridotomy</i>	↓ PAS recurrence, better graft survival & BCVA
Zhou et al.	2024	<i>Retrocorneal Membrane Interception–Enhanced Canaloplasty</i>	92% qualified success (1 yr), safe in open/small PAS ICE
Sun et al.	2024	<i>Combined Phaco + AGV + IOL</i>	72% survival at 5 yrs, stable vision, fewer reoperations

Surgical Pearls and Practical Lessons in ICE-Related Glaucoma

1. Tube Position Matters

In pseudophakic eyes → **insert tube via the ciliary sulcus**, not anterior chamber

Minimizes **corneal decompensation** and **iris trauma**, especially in extensive PAS (Sun et al., 2024)

♥ 2. Interrupt the Membrane's Progression

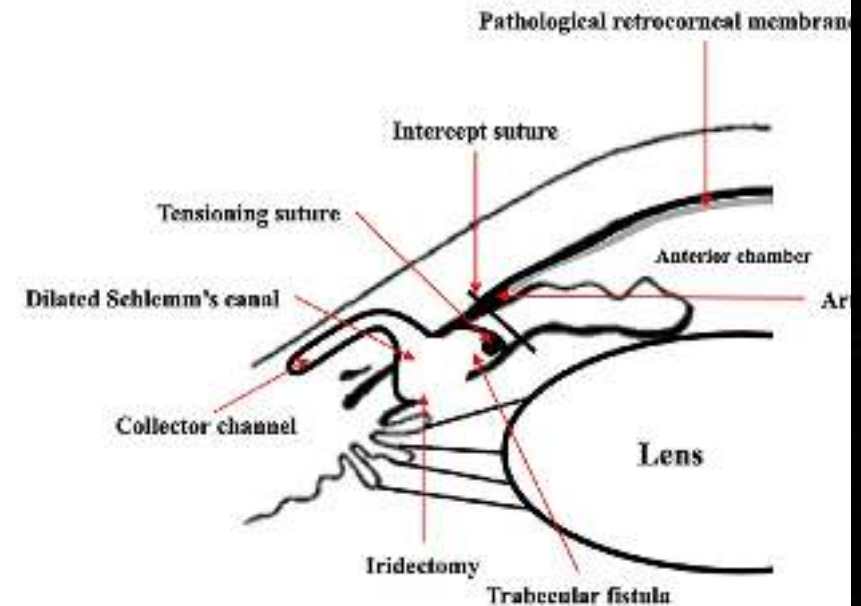
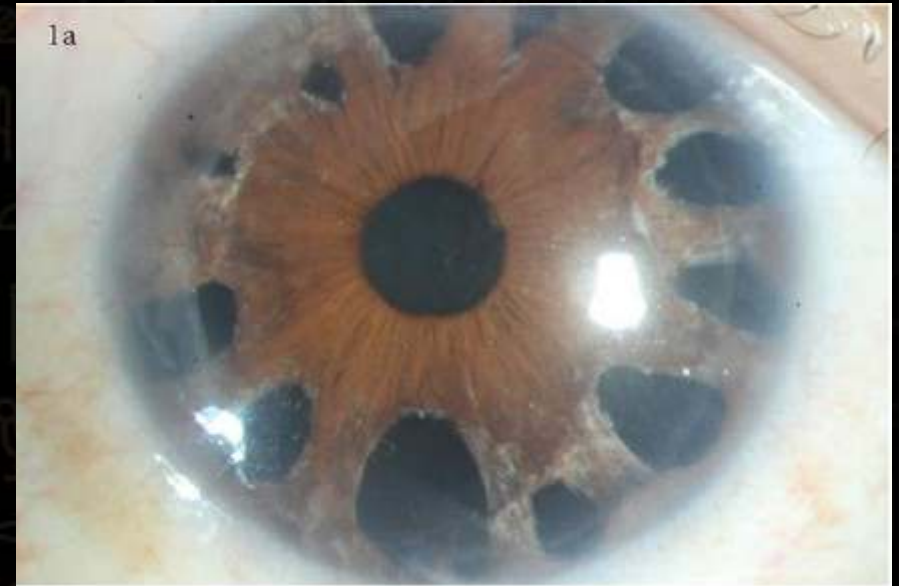
Retrocorneal membrane interception—enhanced canaloplasty (Zhou et al., 2024) prevents endothelial overgrowth over new outflow sites

Ideal for early / small PAS ICE with open angles

♥ 3. Protect the Cornea During EK

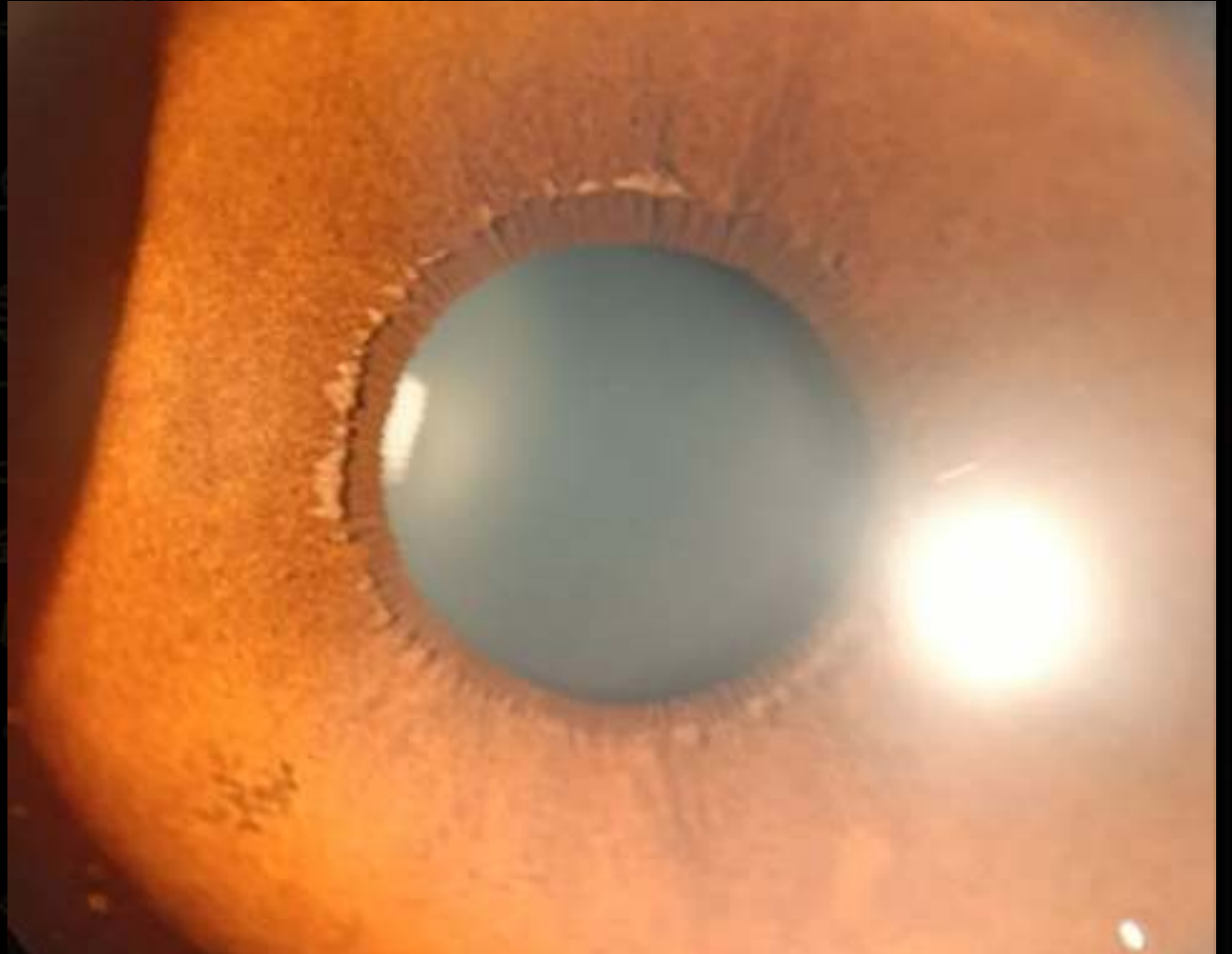
Spokewise iridotomy (Zhang et al., 2023) during DSAEK reduces PAS recurrence and postoperative IOP spikes

Improves graft survival and BCVA



When Diagnosis Brings Clarity

Now I know what I have... I can live with it."



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Key Takeaways — Cracking the ICE

- **ICE syndrome is unilateral, acquired, and deceptive.**
Always suspect it in young patients with unilateral glaucoma and iris changes.
- **Early recognition saves both cornea and optic nerve.**
Subtle iris atrophy or focal PAS may be your only clue.
- **Surgical success depends on respecting the endothelium.**
Sulcus tube placement, membrane interception, and adjunctive iridotomy all minimize damage.
- **Patient understanding matters as much as pressure control.**
A clear diagnosis transforms anxiety into adherence.

Thank you

- *Cracking the ICE: Insights into the Enigmatic Syndrome*
Dr. Khaled El Boushi, FRCed
- **References :**
- **İmamoğlu S, Sevim MS, Yıldız HE, Vural ET, Bardak H, Bardak Y. 2017** *Surgical outcomes of patients with iridocorneal endothelial syndrome: a case series. Int Ophthalmol...*
- **Mao AJ et al. Eye. 2021.** Mitomycin-C enhanced AGV outcomes in ICE.
- **Zhang J et al. Front Med (Lausanne). 2023; 10:1187009.** Spokewise Iridotomy with DSAEK.
- **Zhou M et al. Int Ophthalmol. 2024.** Retrocorneal Membrane Interception Canaloplasty.
- **Sun Y et al. BMC Ophthalmol. 2024; 24:476.** Combined AGV + Phaco in ICE.