HH-ASOCT in SECONDARY PEDIATRIC GLAUCOMA

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Evolution of OCT

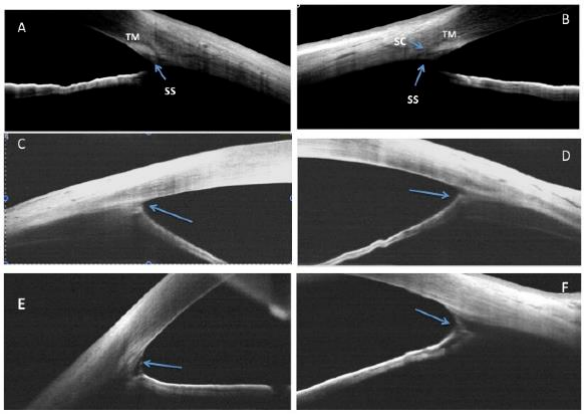
Time-domain prototype in 1991.

Limited by low-image resolution, the details of the angle structures could not be clearly visualized.

The use of **spectral-domain** OCT for anterior segment imaging was first described in 2001.

The introduction **HH-SDOCT** is considered a breakthrough specially in pediatric ophthalmology.

HH-OCT HELPED US DIFFERENTATAITE NORMAL ACA FROM PCG



Can HH-ASOCT HELP US IN THE DIAGNOSIS OF SECONDARY PEDIATRIC GLAUCOMA?

IMAGING CASES

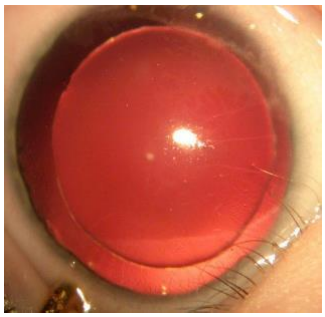
OPTOVUE IVUE SD-OCT.

The instrument has wavelength : 840±10nm , scan length : 2-8 mm, an axial resolution of approximately 15µm , scan speed is 26,000 A- scan/second , image frame takes 256 to 1024 A-scan/Frame.

I. Aniridia

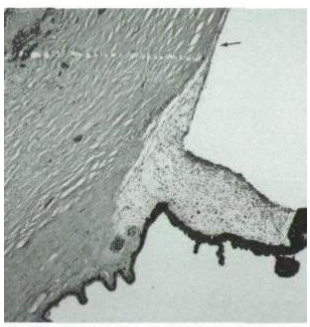
GLAUCOMA IN ANIRIDIA

- Glaucoma occurs in early adulthood.
- Occurs in infants and toddlers
- Incidence range **6% to 75%**



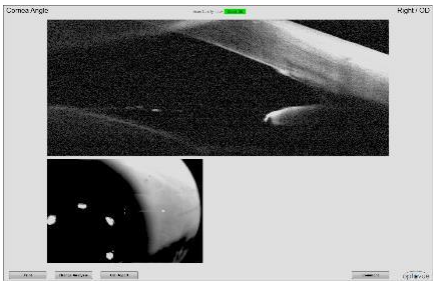
Aniridia

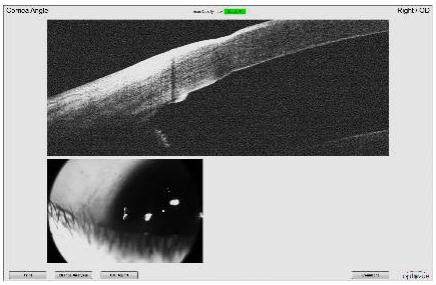
- **HISTOPATHOLOGY**
- All cases had an iris stump
- The TM and ciliary processes: visible posterior to the stump.
- If glaucoma develops:
  - irregular strands from the iris stroma attaching it to the angle wall.
  - These attachments become thicker, move forward, causing obscuration of the TM, SS and the CB.
  - The iris stump tilt and the angle gradually closed.



URE 3: The angle is markedly abnormal. The interr cus is filled with iris tissue. The anterior border layer ends forward to Descemet's membrane (arrow). The cil elatively more hypoplastic than other cases in this st pressed appearance of the ciliary muscles is conside ifact (hematoxylin-eosin, X 63, AFIP Neg 82-8512).

ANIRIDIA
OD NASAL AND TEMPORAL ANGLE





II. Anterior segment dysgenesis (ASD)

Anterior segment dysgenesis (ASD)

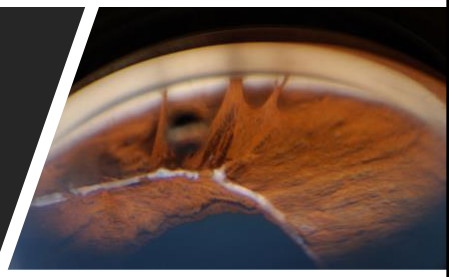
- A spectrum of developmental anomalies
- Resulting from abnormalities of neural crest migration and differentiation during embryologic development.
- INCLUDE:
- 1-Axenfeld-Rieger anomaly /syndrome
- 2-Peters anomaly
- 3-Posterior keratoconus
- 4-Iridoschisis.

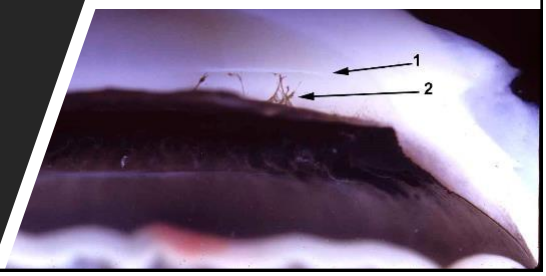
1-AXENFELD-REIGER ANOMALY/SYNDROME

- An autosomal dominant disorder
- A spectrum of anomalies.
- Ranging from isolated bilateral ocular defects to a fully manifested systemic disorder.
- There is at least a **50%** risk of developing glaucoma.

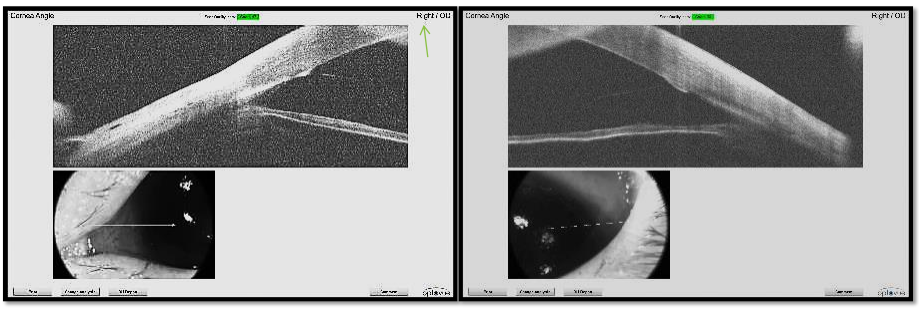
Ocular manifestations of Axenfeld-Rieger anomaly/syndrome

- Posterior embryotoxon (a thickened and anteriorly displaced Schwalbe line
- Iris strands adherent to the Schwalbe line
- iris hypoplasia, corectopia and polycoria
- A maldeveloped or "fetal" anterior chamber angle.





Axenfeld-Reiger anomaly



2-PETERS' ANOMALY

- Most common gene mutations include PAX6 and FOXC1
- Classified into two subtypes: Type 1, and Type 2.
- Glaucoma occurs in up to **90%** of the cases.

Types of Peters' anomaly

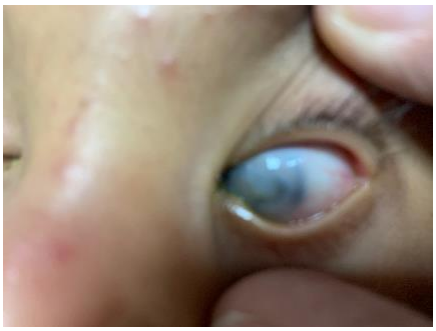
Type 1

- 80% of cases present bilaterally
- Central and paracentral corneal opacification.
- The cornea is avascular.
- Iris strands extend from the collarette
- Systemic abnormalities are not present.

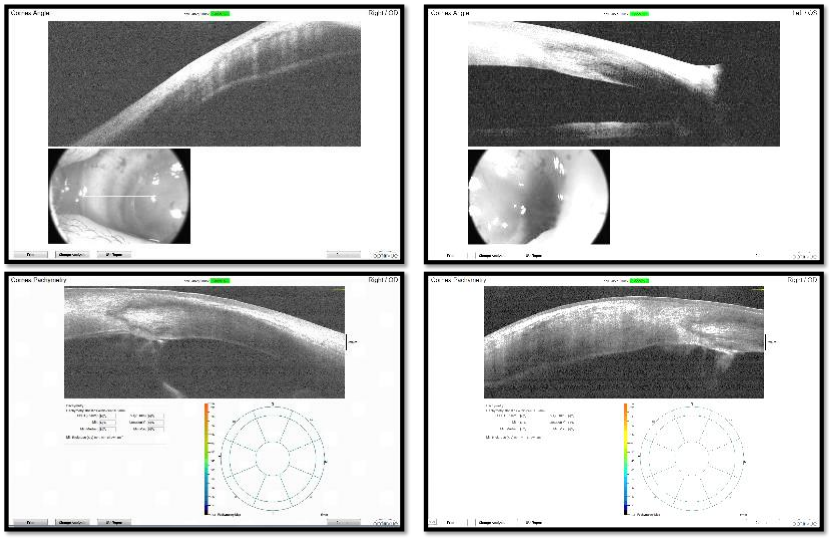
Type 2

- Cases are commonly bilateral
- Denser corneal opacification
- **Juxtaposition** of the lens
- iris strands may or may not be present.
- The posterior stroma and Descemet membrane is classically malformed.
- Systemic abnormalities are more common.

Types of Peters' anomaly

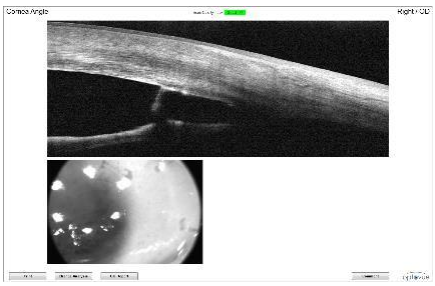


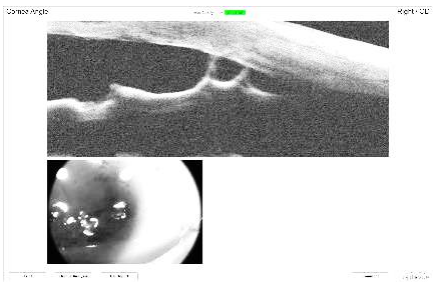
Peters' Anomaly
Case 1



- Malformed posterior stromal corneal fibres.
- Malformed interrupted DM.

Peters' anomaly TYPE 1 case 2 (OD temporal angle)

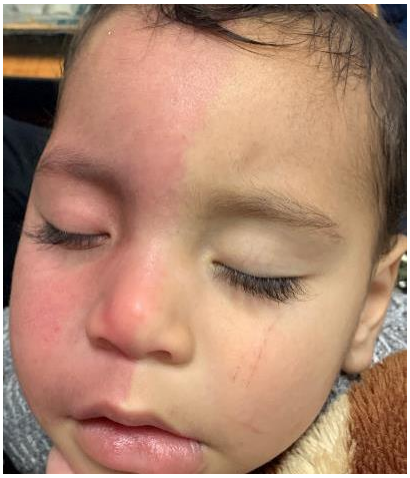




III. Phakomatoses

1-Sturge weber syndrome

2-Neurofibromatosis



1-STURGE-WEBER SYNDROME (SWS)

- About **1/3 to ½** of patients with SWS will develop glaucoma
- MECHANISM OF GLAUCOMA:

(early-onset glaucoma) congenital angle abnormality and elevated episcleral venous pressure

Elevated episcleral venous pressure (late-onset glaucoma).

Histopathological and Goniosopic findings in SWS

- **In infancy and childhood:**

Clinical and histopathological features of ACA are similar to PCG.

- **On gonioscopy:**

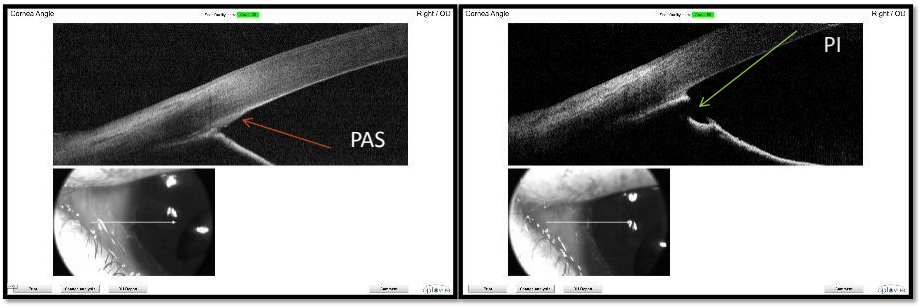
❑ The angle structures indistinct, with a high iris insertion.

❑ An anteriorly displaced iris root

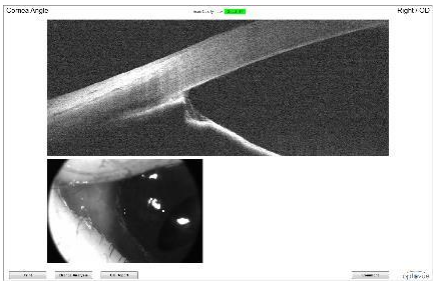
❑ Poorly developed scleral spur.

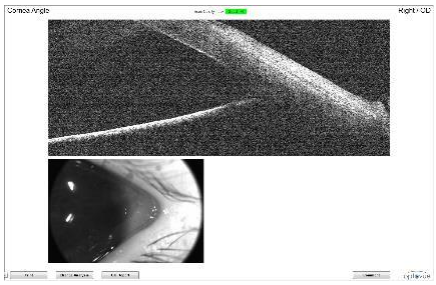
❑SWS patients with early-onset glaucoma present with typical signs of congenital glaucoma.

Sturge-Weber syndrome (SWS) case 1 (OD)

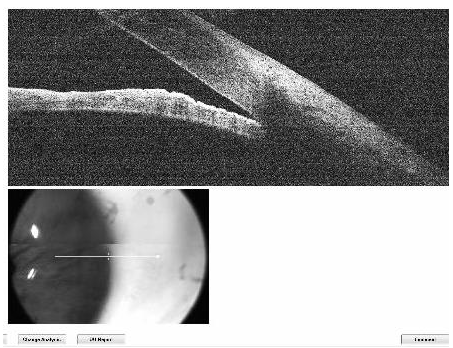


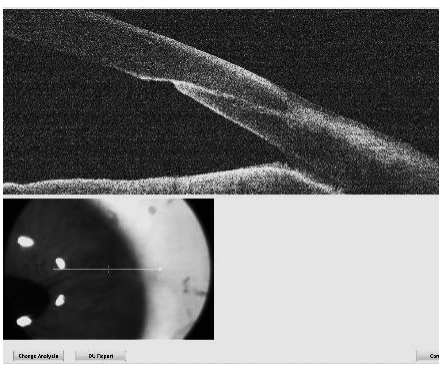
Sturge-Weber syndrome (SWS) case 1 (OD) temporal and nasal angle





STURGE-WEBER SYNDROME case 2

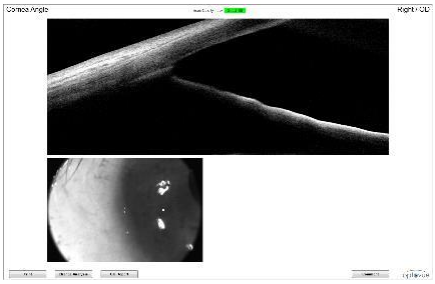




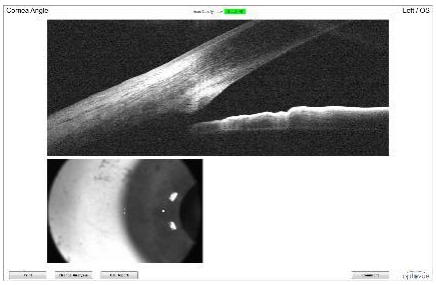
2-Neurofibromatosis

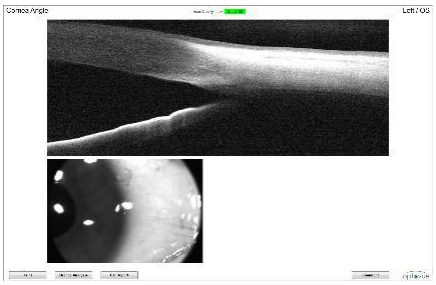
- Glaucoma occurs if the eyelids have neurofibromas.
- The mechanism:
  - Infiltration of the angle with neurofibromatous tissue
  - Angle closure caused by nodular thickening of the ciliary body and choroid
  - Failure of the anterior chamber to develop.

Neurofibromatosis
OD nasal and temporal angle



Neurofibromatosis
OS nasal and temporal angle (normal)





Is it useful to image the ACA using AS-OCT in secondary pediatric glaucoma?

- Non-contact.
- Requires no anesthesia.
- Helps in confirming the diagnosis.
- Follow up the possible changes (postoperative).
- Aids in the choice of the proper surgical method.


Thank You