Swept Source OCT and Wide-Field Fundus Autofluorescence in Paediatric Macular Dystrophies

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Imaging in the Paeds Clinic

- Illustrate the application of novel imaging modalities
- To demonstrate what we learn about our patients using these tools and their value in a tertiary paediatric retinal service
Wide-Field Fundus Autofluorescence

• Visualises disease specific distribution of lipofuscin in the retina that may not yet be visible/obvious on clinical examination

• WF-AF is helpful for the differential diagnosis of retinal dystrophies and to monitor the clinical course over time

• Up to 200 degrees retinal imaging in single image
Advantages by Swept-source

2x Faster imaging speed
(100,000 A-line/s)

Uniform image quality

Improved Vitreous visualization

Advantages by longer wavelength
(1,050nm)

Increased penetration and visibility of choroid and sclera

12mm (40 deg) long scans
Relevance in Children

Imaging gives:
- Documentation
- Help parents understand condition
- Improved ability to monitor/assess progression

Benefits over clinical examination
- Pick up pathology not clinically visible
- May significantly exceed what is seen clinically, esp in young children
  Single shot image up to 200 degrees view of retina (colour and AF)

SS-OCT
- Rapid imaging <3seconds
- Invisible scanning light \(\rightarrow\) reduced eye movements
Clinical Cases
Cone Dystrophy

- Autosomal-recessive, predominantly cone-mediated dystrophy
- CNGA3 mutation
- ERG: cone>>rod dysfunction

- **WF-AF**
  - Used to diagnose and monitor the monitor condition
Minimal change detectable clinically
WF-AF:

- RPE status imaging
- Extent of FAF correlates with severity of functional impairment
- Useful for monitoring for progression

Minimal change detectable clinically

VA: 6/15 BE
CONE DYSTROPHY –
WHAT DOES SS-OCT ADD?
CONE DYSTROPHY – WHAT DOES SS-OCT ADD?

- Extensive examination of retina to assess for changes in centre and periphery

Outer retinal disruption including ellipsoid zone centrally
Normal peripheral outer retina
X-Linked Retinoschisis

• **Widefield imaging** (Optos® and SS-OCT):
  – Monitor for peripheral schisis/vitreous haemorrhage/detachment

• **12mm SS-OCT line scans**
  – Used to delineate and monitor retinoschisis
  – Greater quality vitreous imaging
13yrs old. RE: 6/12, LE: 6/9
Abnormal tapeto-retinal sheen
12mm scans SS-OCT demonstrate extensive schisis and greater detail of layers involved
X-Linked Retinitis Pigmentosa

- 14-yr old male
- 7 yrs f/up, stable VAs 6/12 OD, 6/9 OS)

Deterioration in vision reported (6/19 OD):
- Clinically no change in macular appearance
- OCT – NO CMO

WF-AF: used to assess RPE status
X-Linked RP
6/12 → 6/19
AF – Central RPE stress explains the vision loss
Conclusion

• Multi-modal imaging is the now and the future in paediatric retinal conditions

• Imaging can give new information beyond the clinical picture and be invaluable for diagnosis, management, observation and prognosis

• Choose your imaging modalities to answer the clinical question
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- The extent of abnormal FAF reflects the severity of functional impairment in patients with cone-dominant retinal dystrophies. Fundus autofluorescence measurements are useful for predicting retinal function in these patients.