



Investigation of a child with a unilateral retinal lesion.

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An 11 year old female was referred for investigation after reduced vision of the left eye (hand movements) was detected at a routine optician appointment.

Vision and examination of the right eye was normal.

C/o: History of poor LVA for a few years initially appeared as a “black dot” and progressed.

Rx: Tried mild myopic glasses to improve left VA but these were discarded.

FH: Grandmother poor vision from glaucoma and Father has a lazy eye.

GH: Good, no medications, no other problems, vaccinations up to date, normal development.

VA: RE -0.100 LE No Perception of Light (NPL)

Pupils: RE normal LE mild RAPD

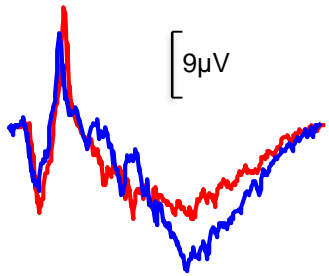
Refraction: No Rx

Anterior segment: R+L normal

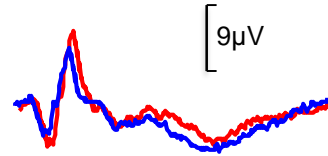
IOP: RE 14mmHg LE 10mmHg (Goldmann)

Fundus: RE normal. LE significant peripapillary hyperpigmented area extending over the macular.

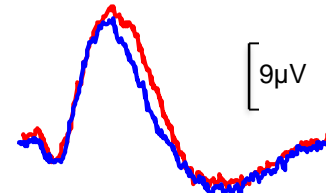
Visual Electrophysiology: Flash Electroretinogram (skin electrodes)



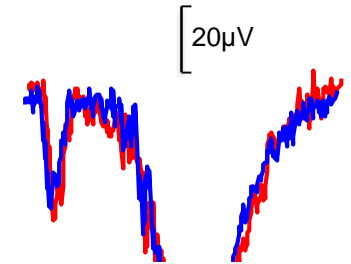
Binocular Gr 4 Scotopic



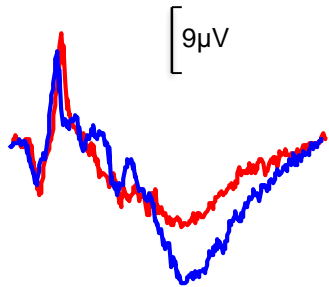
Red Gr 4 Scotopic



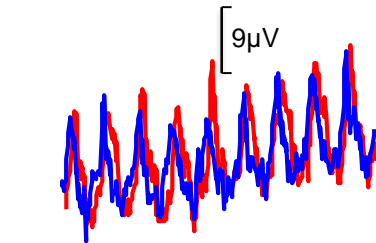
Blue Gr 1 Scotopic



Single Gr 16 Scotopic



White Gr 4 Photopic

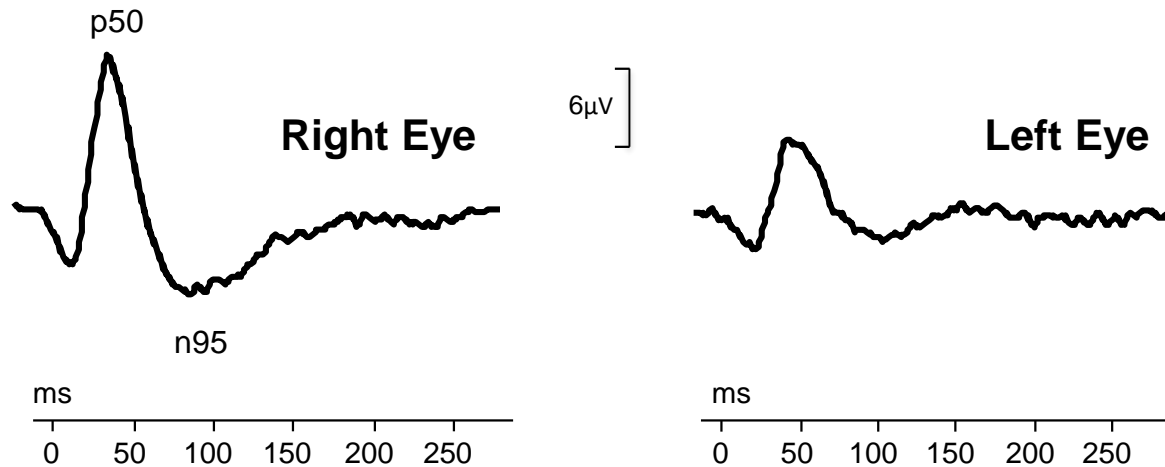


30Hz Flicker Photopic

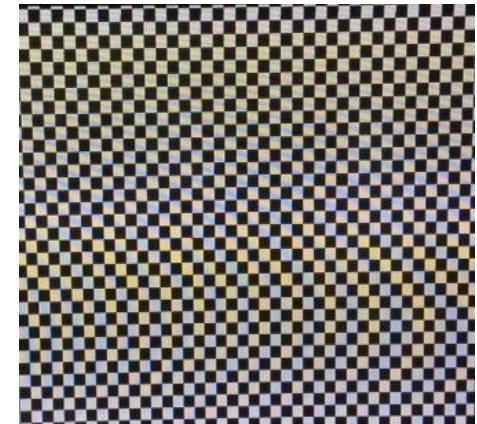
Grand average ERG responses from the “**Right eye**” and “**Left eye**”.

Responses are comparable for each eye and within normal limits indicating normal generalised rod and cone function.

Visual electrophysiology: Pattern Electroretinogram



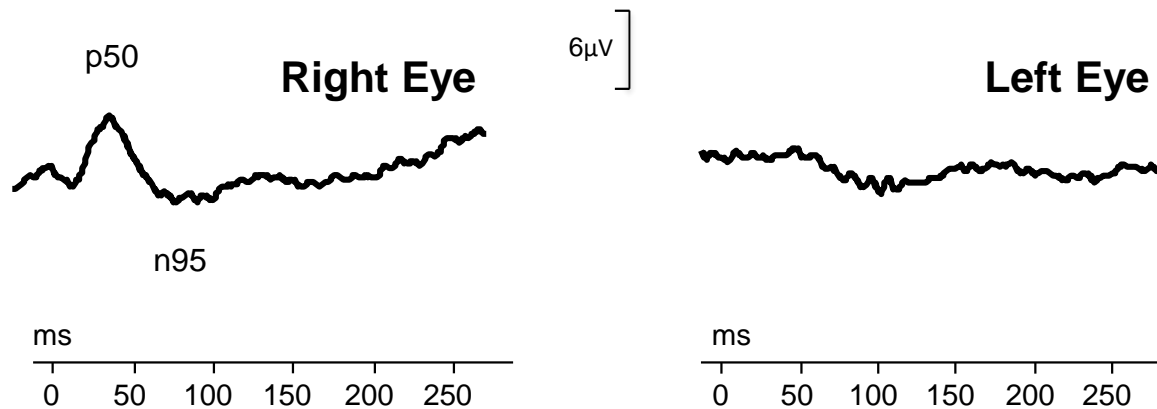
Stimuli = 30 Degrees



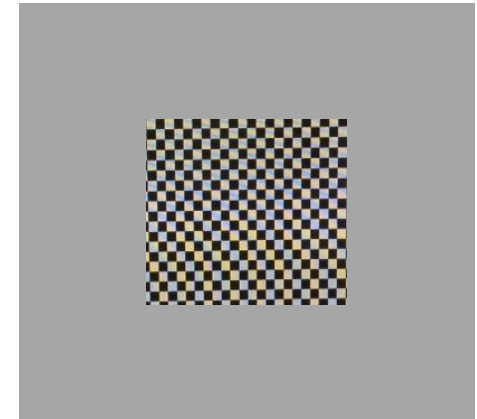
The p50 and n95 components of the response from the right eye are normal indicating normal distal macular and ganglion cell function of the central 30 degrees.

The response from the left eye is much smaller indicating significant distal macular and ganglion cell dysfunction affecting the central 30 degrees.

Visual electrophysiology: Pattern Electroretinogram



Stimuli = 15 Degrees

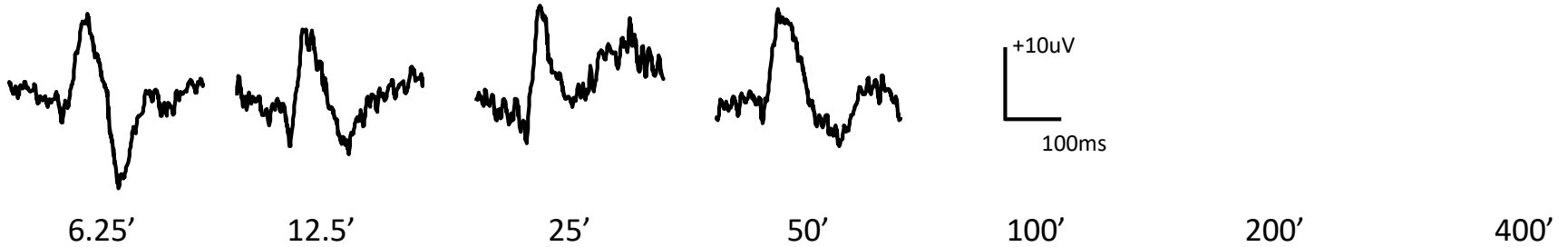


The p50 and n95 components of the response from the right eye remain normal. The response is smaller due to the smaller field of stimulation.

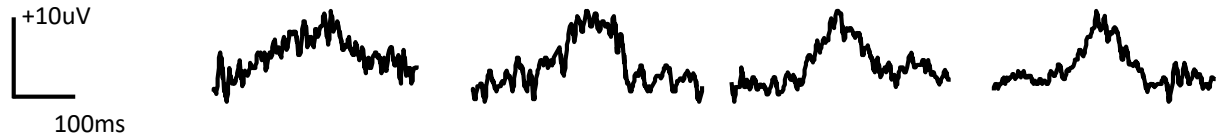
The response from the left eye becomes degraded indicating the major sight of the dysfunction is the central 15 degrees.

Visual electrophysiology: Pattern reversal Visual Evoked Potentials

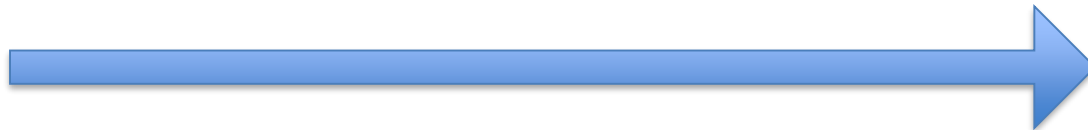
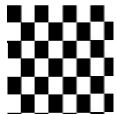
RIGHT EYE



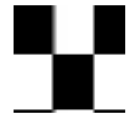
LEFT EYE



Smallest
Test
checks

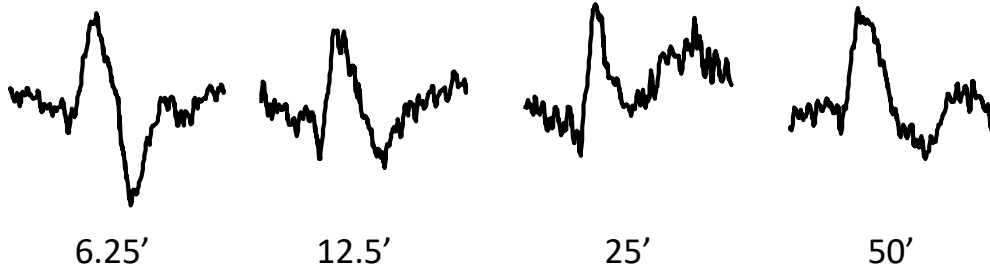


Largest
Test
checks



Visual electrophysiology: Pattern reversal Visual Evoked Potentials

RIGHT EYE

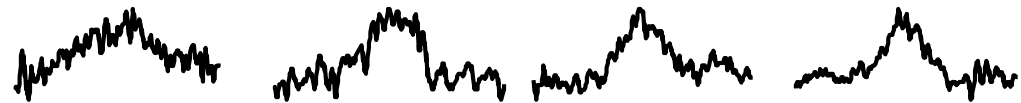


RE = Normal macular pathway function with well defined responses to the smallest test checks.

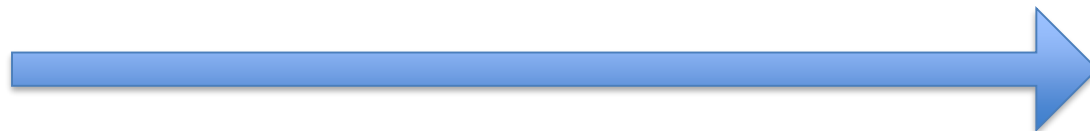
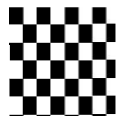
100' 200' 400'

LEFT EYE

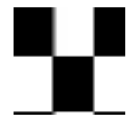
LE = Significant macular pathway dysfunction with preserved responses to the largest test checks.



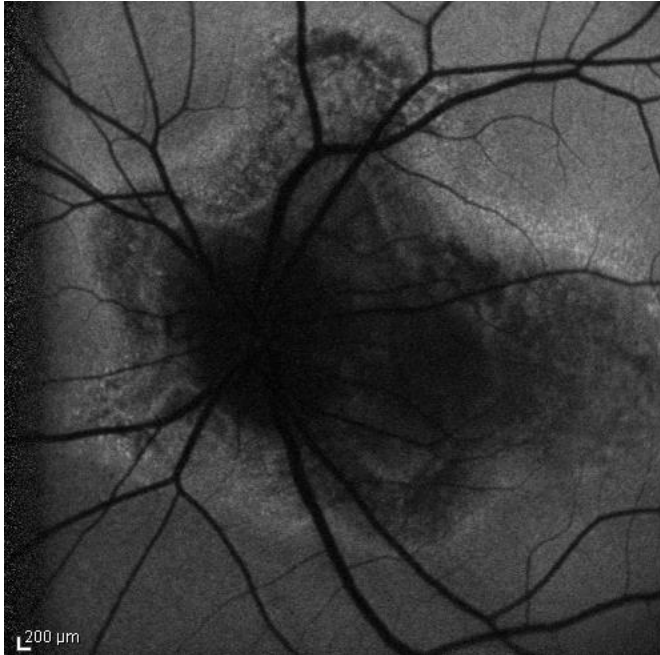
**Smallest
Test
checks**



**Largest
Test
checks**



Retinal imaging of the left eye (right eye not shown - normal)

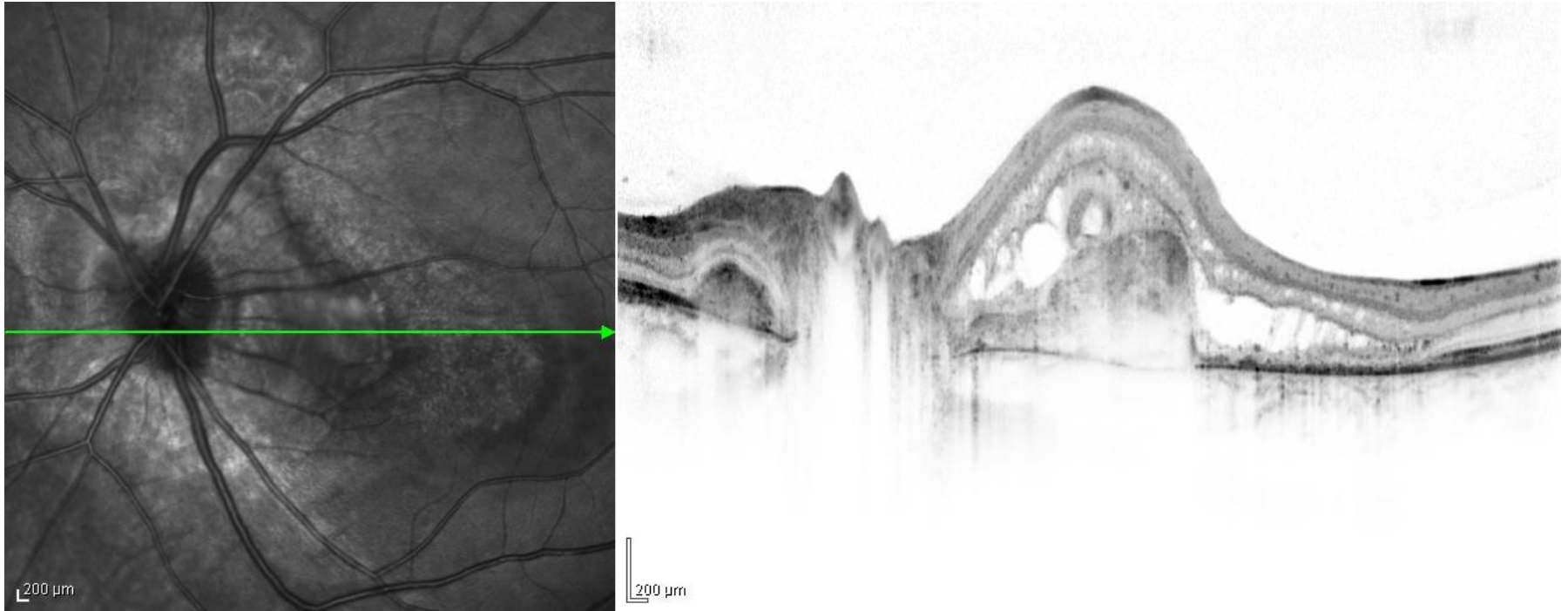


Fundus Auto-fluorescence



Colour Fundus Photograph

Retinal imaging of the left eye (right eye not shown - normal)



Infra-red image and Optical Coherence Tomography

Right Eye – Normal.

Left Eye – One very small linear drusen.

What should be done next?

-
- ① Of all the investigations undertaken which did you find the most useful?
 - a) Clinical examination
 - b) Retinal imaging (color photos OCT and Autofluoresence)
 - c) Pattern Visual Evoked Potentials
 - d) Pattern Electroretinogram
 - e) Flash Electroretinogram
 - f) Ultrasound

 - ② Would you request any additional investigation, if so what would be your priority?
 - a) Fluorescein angiography
 - b) Blood tests (ANA, ESR, U&E, FBC, ANCA, toxoplasma screen and complement function)
 - c) MRI
 - d) None

 - ③ Would you offer any treatment?
 - a) Intravitreal injection of bevacizumab
 - b) Intravitreal injection of ranibizumab
 - c) Intravitreal injection of a steroid
 - d) Nothing - observe

Could our experts answer these questions in addition to those on the previous slide?

1. What is your differential diagnosis?
2. The child is consistently adamant her vision in the left eye is no perception of light. Do you agree?
How would you try to prove/disprove this?