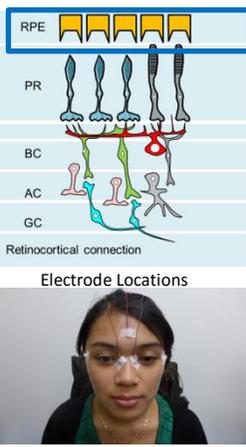
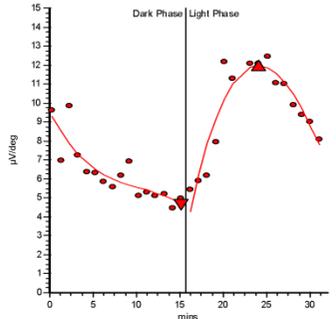
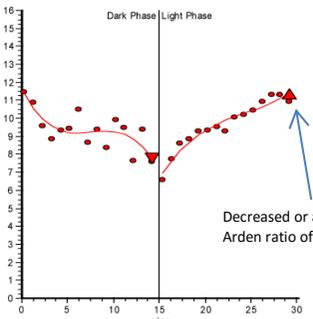
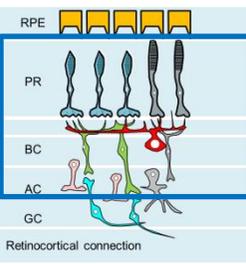
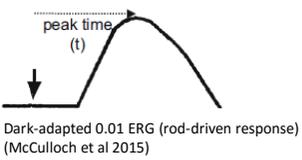
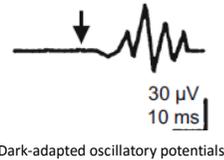
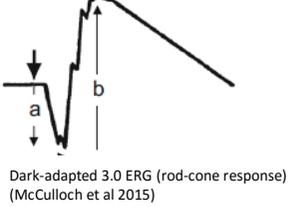
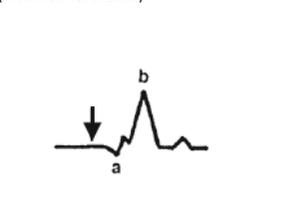
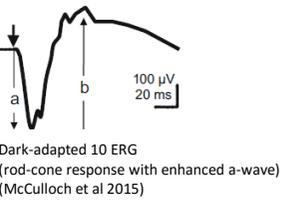
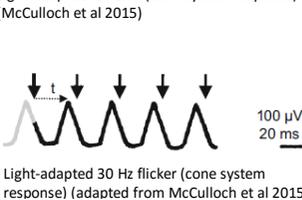
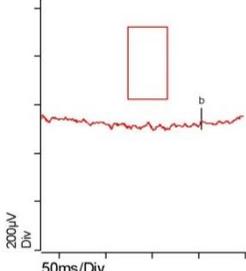
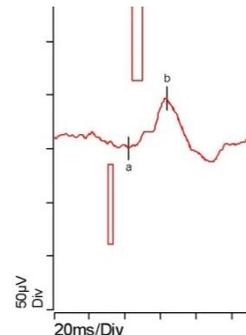


Electrophysiological tests may be considered in cases of: unexplained vision loss, symptoms and/or family history suggestive of an inherited eye disease, and use of potentially retino toxic medication. These tests are not primary tests but are performed on indication based on the results from an initial assessment typically consisting of imaging and functional tests.

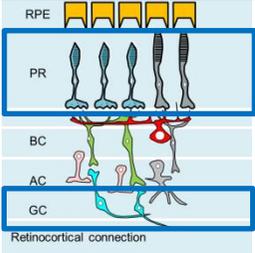
Technique	Clinical applications	Abnormal results and associated conditions
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Electro-oculography (EOG) – assessment of retinal pigment epithelium function		
 <ul style="list-style-type: none"> An eye movement generated voltage recorded by placing electrodes either side of the eyes. Created by the corneofundal potential (aka standing potential: the cornea is 6-10 μV positive relative to the RPE.) The corneofundal potential: <ul style="list-style-type: none"> is mainly derived from the RPE changes in response to background levels of retinal illumination Results are collected under scotopic and photopic conditions Usually expressed as light/dark or Arden ratio A ratio of the maximum (peak) amplitude in the light (LP) to the minimum (trough) amplitude in the dark (DT) 	 <p>An EOG in a healthy individual showing an obvious light peak (Arden ratio of 2.58)</p> <p>Mainly useful for subretinal diseases: It is most specific for Best disease</p> <ul style="list-style-type: none"> Arden ratios: <ul style="list-style-type: none"> >2.0 generally normal 1.5 - 2.0 borderline <1.5 generally abnormally low 	 <p>EOG with multifocal Best disease (multifocal vitelliform lesions)</p> <p>Decreased or absent light peak with Arden ratio of 1.46</p> <p>Typical patterns of loss Mainly used for Best disease however other conditions that involve the RPE will also show abnormal responses.</p>

Full-field electroretinography (ERG) – assessment of the function of photoreceptors and cells in the inner nuclear layer of the global retina		
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 <ul style="list-style-type: none"> Records the massed electrical response of the retina to a flash of light. The waveform of a typical full-field ERG has: <ul style="list-style-type: none"> Initial negative a-wave: Photoreceptors (receptor potential) Positive b-wave: ON-bipolar cells Oscillatory potentials (smaller waves): Amacrine cell activity There are two broad categories of full-field ERG: <ul style="list-style-type: none"> Light-adapted (photopic) ERG - Flash and 30Hz Flicker Dark-adapted (scotopic) ERG - 3 flash brightness levels and oscillatory potentials  <p>Diagnosys ERG unit</p> <p>DTL electrode (silver/nylon fibre electrode) sitting on the margin of the lower lid</p>	<p>Widely used for the differential diagnosis of conditions including:</p> <ul style="list-style-type: none"> Inherited retinal disease Retinal vascular disease Opaque media or trauma Unexplained visual loss Infant assessment Albinism Toxic and nutritional eye disease  <p>Dark-adapted 0.01 ERG (rod-driven response) (McCulloch et al 2015)</p>  <p>Dark-adapted oscillatory potentials (McCulloch et al 2015)</p>  <p>Dark-adapted 3.0 ERG (rod-cone response) (McCulloch et al 2015)</p>  <p>Light-adapted 3.0 ERG (cone system response) (McCulloch et al 2015)</p>  <p>Dark-adapted 10 ERG (rod-cone response with enhanced a-wave) (McCulloch et al 2015)</p>  <p>Light-adapted 30 Hz flicker (cone system response) (adapted from McCulloch et al 2015)</p>	 <p>Dark-adapted 0.01 ERG with RP: undetectable rod response (Box represents normative range)</p> <p>Typical patterns of loss</p> <p>Retinitis pigmentosa (RP)</p> <ul style="list-style-type: none"> Scotopic ERG is more reduced compared to photopic ERG with both worsening with time. <p>Cone dystrophy</p> <ul style="list-style-type: none"> Photopic ERG is reduced. <p>Cone-rod dystrophy</p> <ul style="list-style-type: none"> Photopic ERG is more reduced than scotopic ERG. <p>Choroideremia</p> <ul style="list-style-type: none"> Scotopic ERG is more reduced compared to photopic ERG in the early stages with both worsening with time.  <p>Light-adapted flash ERG with RP: detectable but reduced cone response (Boxes represent normative ranges)</p>
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Pattern electroretinography (PERG) – assessment of ganglion cell and central retinal function

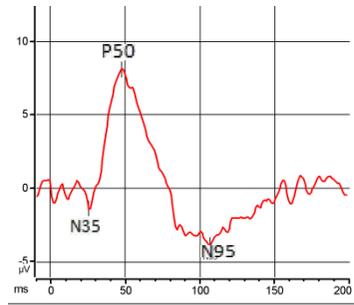


- A local retinal response evoked by a contrast-reversing pattern (black and white checkerboard)
- Arises largely in the **ganglion cells**
- Two major components:
 - P50: related to ganglion cell and partially pre-ganglionic activity
 - an indication of macular function
 - N95
 - related to ganglion cell function (numbers correspond to milliseconds)

Electrode locations



Checkerboard stimulus for pattern ERG

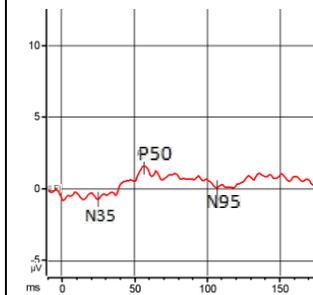


A normal pattern ERG

Pattern ERG may be useful in:

- Retinal vascular disease
- Unexplained visual loss
- Toxic and nutritional eye disease
- Stargardt disease
- Cone dystrophies
- Macular dystrophies
- Hydroxychloroquine (Plaquenil) retinopathy
- Glaucoma
- Optic neuropathies
- Primary ganglion cell diseases ([e.g. Leber Hereditary Optic Neuropathy (LHON) and Kjer-type dominant optic atrophy (DOA)])

It is also used when pattern VEP is abnormal to determine the location of the abnormality (ie central retinal or optic nerve)

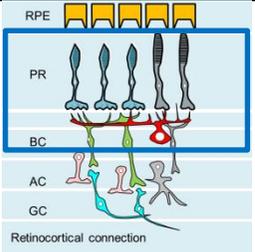


Pattern ERG in Stargardt disease
Reduced P50 and N95 amplitudes.

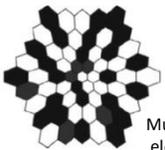
Typical patterns of loss

- **Stargardt disease**
 - P50 reduction
- **Optic nerve demyelination**
 - mainly N95 involvement
- **Acute optic neuritis**
 - Reduced P50 which usually recovers within a few weeks
- **Ischaemic optic neuropathy**
 - more frequently P50 involvement
- **Lebers Hereditary Optic Neuropathy**
 - Normal P50 but very poor N95 (at acute presentation)
- **Kjer-type dominant optic atrophy**
 - N95 reduction

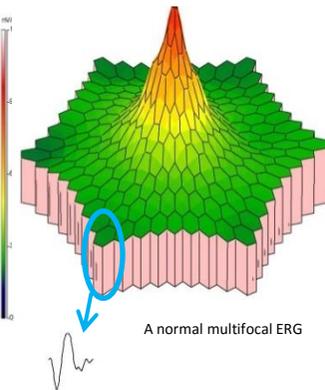
Multifocal Electroretinography (mfERG) – assessment of local retinal function



- Provides a topographical assessment of retinal function by testing local retinal function
 - typically 61 or 103 locations (40-50°)
- Recorded from the cone system by stimulating the retina with an array of hexagons
- Largely influenced by **bipolar cells** with some contribution from **photoreceptors and inner retinal cells**
- Each individual waveform is not an individual ERG waveform but a mathematical extraction from a single continuous ERG recording.



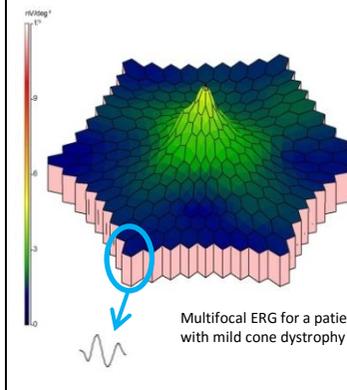
Multifocal ERG stimulus with 61 hexagonal elements (adapted from Hood et al 2012)



A normal multifocal ERG

Useful in assessing localised abnormalities in the **macular, paramacular, or discrete peripheral retina in conditions such as**

- Cone dystrophies
- Cone-rod dystrophies
- Macular dystrophies
- Hydroxychloroquine (Plaquenil) retinopathy
- Retinitis pigmentosa
- Fundus albipunctatus
- Stargardt disease

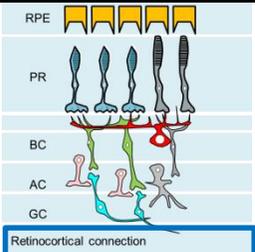


Multifocal ERG for a patient with mild cone dystrophy

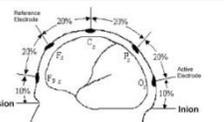
Typical patterns of loss

- Reduction in waveforms correspond to the retinal locations of the pathology
- 1. **Cone and Cone-Rod dystrophies** - reduced or undetectable
- 2. **Hydroxychloroquine (Plaquenil) retinopathy** - decreased parafoveal waveform amplitudes
- 3. **Stargardt disease** - depressed foveal peak
- 4. **Best disease** - often abnormal with amplitudes correlating to visual acuity

Visually evoked potentials (VEP) – assessment of the integrity of the visual pathway from the eye to the occipital cortex



- Generated by the **occipital visual cortex in response to stimulation of the retina** by either light flashes or pattern stimuli using different check sizes
- If abnormal, cause can be located anywhere between the retina and the cortex.
- Typical waveform of the pattern-reversal VEP consists of N75, P100, and N135 peaks (correspond to milliseconds)
- Analysis primarily looks at the peak time and amplitude of P100



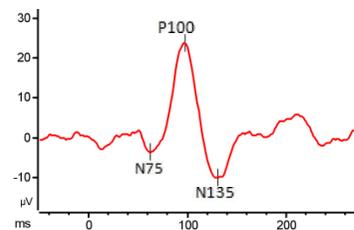
Electrode locations (adapted from Odum et al 2010)



Small checks



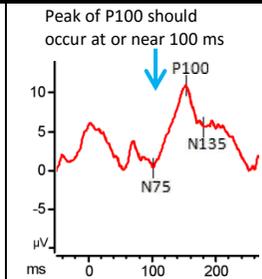
Large checks



A normal pattern-reversal VEP

Mainly used to confirm the diagnosis of **optic neuropathy and other demyelinating disease**

- Other uses include but not limited to:
- Unexplained visual loss
 - Toxic and nutritional eye disease
 - Suspected intracranial lesion



Pattern-reversal VEP showing a delayed peak time (P100) at 160 ms due to a chiasmal lesion

Typical patterns of loss

- **Optic nerve demyelination**
 - Delayed peak time with amplitude being less affected
- **Acute optic neuritis**
 - Reduced amplitude with peak time being less affected
 - amplitude recovers with time (associated with resolving oedema)
 - peak time becomes delayed (associated with demyelination).

This chair-side reference was designed to assist optometrists in private practice when interpreting results from electrophysiological tests covered by the ISCEV Standard. It provides general information only and does not include non-standard electrophysiological tests.

Disclaimer: Although every care has been taken to ensure that this document is free from any error or inaccuracy, CFEH does not make any representation or warranty regarding the currency, accuracy or completeness of these Guidelines.

Abbreviations

EOG	Electro-oculography
RPE	Retinal pigment epithelium
PR	Photoreceptors
BC	Bipolar cells
AC	Amacrine cells
GC	Ganglion cells
ERG	Electroretinography
ISCEV	International Society for Clinical Electrophysiology of Vision
VEP	Visually evoked potentials

Acknowledgements

The schematic retinal presentation figure is adapted from Figure 1 in Whatham AR and Nguyen V et al. The value of clinical electrophysiology in the assessment of the eye and visual system in the era of advanced imaging. *Clinical and Experimental Optometry* 2014;97:99-115

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