

Visual Electrodiagnostics: Clinical Cases, Diagnostic Indications, and New Developments for Use in Neuro-eye and Retinal Disorders.

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Key Words: Electrophysiology, Retina/Retinal disease, Neurologic disease

With the development of multifocal electrodiagnostic techniques it is now possible to record responses from localized areas of the retina and visual field. Using a series of case studies, the advances and clinical indications for visual electrodiagnostic testing, including full-field and multifocal electroretinography (fERG and mfERG), pattern and multifocal visual evoked potentials are presented.

I. Overview

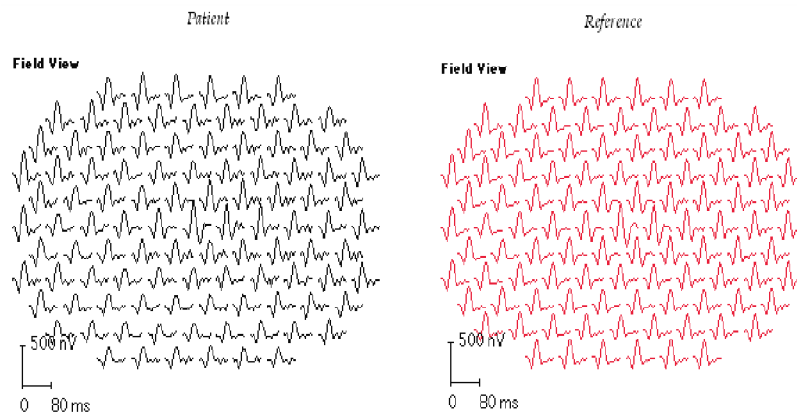
- A. Electrodiagnostic tests
 - 1. Electroretinogram (fERG, mfERG, pattern ERG)
 - 2. Electro-Oculogram (EOG)
 - 3. Visual Evoked Potential (flash/pattern/sweep VEP, mfVEP)

II. Electroretinogram (ERG)

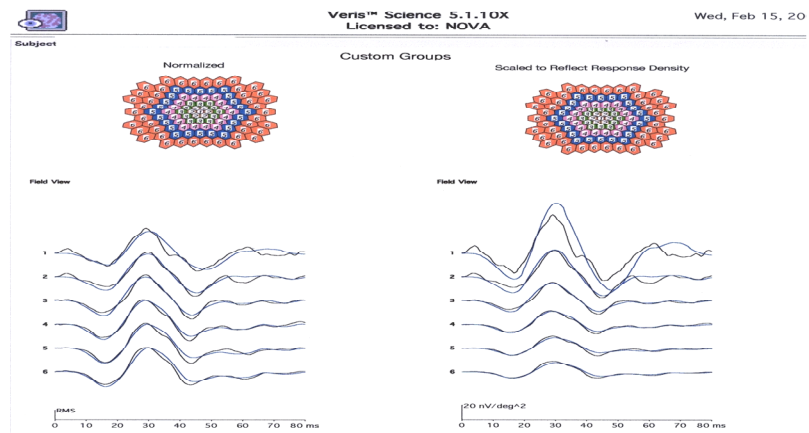
- A. Flash/traditional ERGs
 - 1. A mass potential used to assess both the rod-driven retina and the cone-driven retina
 - 2. Equipment and electrodes
 - 3. Procedure (VIDEO)
- B. Origins of the a-wave, b-wave, c-wave, OPs
- C. Diagnostic indications (with common clinical cases presented)
 - 1. Tapetoretinal degenerations
 - 2. Cone dysfunction
 - 3. Leber's congenital amaurosis
 - 4. Congenital stationary night-blindness
 - 5. Flecked retinal syndromes
 - 6. White dot syndromes
 - 7. Non-organic visual loss
 - 8. Other (Cancer associated retinopathy, early hyperthyroid disease)
- D. Pros
 - 1. Can measure both eyes at the same time
 - 2. Amplifier separates right eye from left eye
 - 3. Gives characteristic results for certain diseases
 - 4. Does not require attentive fixation
- Cons
 - 1. May miss "small" problems as the test only gives the overall potential of the entire retina. Patient with a small area of retinal disease may have a normal flash ERG
 - 2. Does not measure inner retinal disease

E. Multifocal ERGs (mfERG)

1. Overview
 - mfERG provides a topographic map of local retinal electrophysiological activity
2. Comparison to traditional ERG
 - a) Advantages
 - Can be used to detect damage in local regions of retina by spatial variations of mfERG responses
 - Trace arrays can be compared with the visual field results to confirm the relation between visual field defects and retinal dysfunction
 - b) Disadvantages
 - Tests the cone-driven retina under photopic conditions
 - Needs refractive correction for near
 - Requires good central fixation
 - May need advanced analysis for interpretation
3. How it works
 - a) The m-sequence (VIDEO)
 - b) Derivation of response kernels
 - Local response is extracted by correlating the recorded continuous ERG signal with the stimulus sequence
 - Each local response is associated with one of the scaled hexagonal elements (typically 61 or 103) of the display
4. Clinical information it provides
 - a) 1st -order kernel reflects the retinal response to the flash
 - b) 2nd -order kernel reflects the influence of the preceding flash
5. How it's interpreted
 - a) Waveform analysis
 - Trace array
 - Typical measurement for amplitude and timing
 - P1 amplitude of the 1st order kernel
 - P1 implicit time of the 1st order kernel

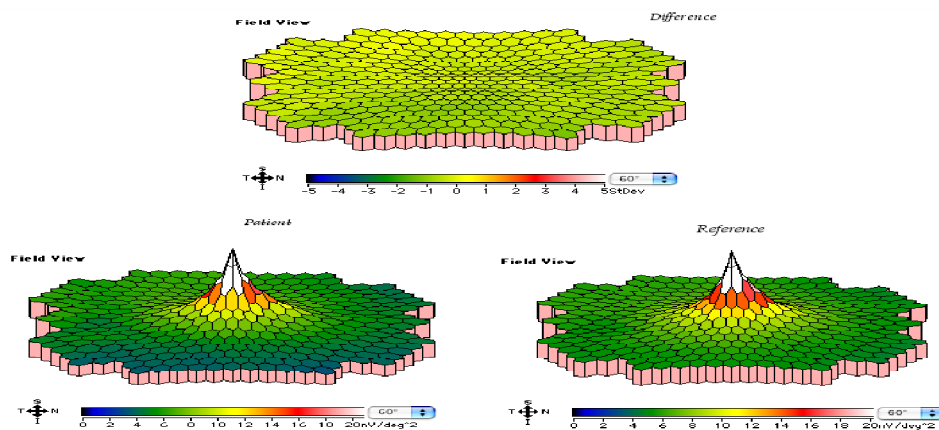


- Custom grouping
 - for various quadrants, hemiretinal areas, or eccentric rings from center to periphery



- Ring ratios
 - The ratios of these ring values to one another can be used to detect abnormalities such as parafoveal deficits

b) Topographic (3-D) response density plots



E. Specific Clinical Applications (with common clinical cases presented)

1. “Typical” retinitis pigmentosa
2. Cone dystrophy (vs. Stargardts disease)
3. Toxic retinopathy
 - e.g. Plaquenil retinopathy
 - mfERG can detect changes prior to Amsler grid, color vision, or fundusoscopic manifestation
4. Ischemic retinopathy
5. Non-organic visual loss
6. White dot syndromes
7. Distinguish diseases of the outer retina from diseases of ganglion cells, or/and optic nerve

8. Other (Cancer associated retinopathy, early hyperthyroid disease)

III. Visual Evoked Potential (VEP)

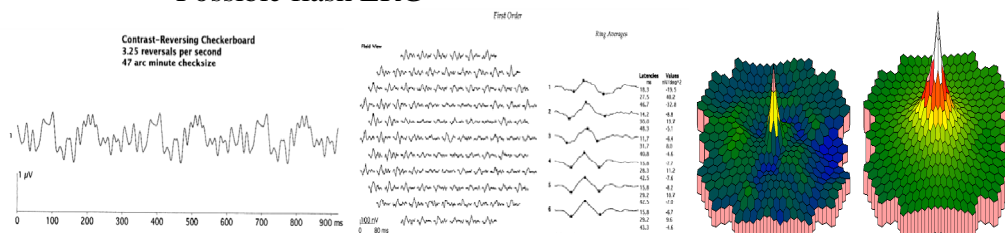
- A. Assesses integrity of visual pathway from retina, optic nerve to the visual cortex
- B. Assess spatial visual processing abilities in pre-verbal and non-verbal individuals
- C. Diagnostic indications
 1. Optic nerve disorders
 2. Demyelinating disease
 3. Spatial acuity estimates
- D. Specific traditional VEP protocols
flash and pattern VEPs
- E. Multifocal VEP (mfVEP)
 1. Comparisons to
 - a) Traditional flash and pattern VEPs
 - mfVEP provides topographic mapping of VEP response
 - b) Multifocal ERG
 - The mfVEP can be abnormal in diseases of the outer retina, as well as in diseases of the optic nerve and/or higher hierarchy levels
 2. How it's recorded
 - a) Electrode configuration - variable
 - b) Stimuli (VIDEO)

Radial scaled sectors of alternating checkerboard patterns
 3. New clinical indications for mfVEP

Evaluating patients (e.g. glaucoma patients) with unreliable or questionable VFs or those that cannot perform VFs
- F. Clinical VEP cases

IV. Electrodiagnostics & Neuro-eye Patients

- A. DDx optic nerve vs. retina disease
 - mfERG and pVEP
 - Possible flash ERG



- B. Optic neuritis, etc (occult presentations)
 - pVEP
 - mfVEP
- C. Vision loss unknown etiology
 - Functional vision loss vs retinal/visual pathway
 - The kitchen sink approach

V. Recommended Reading

1. Heckenlively JR and Arden GB. Principles and Practice of Clinical Electrophysiology of Vision, 2nd Ed. Cambridge: MIT Press; 2006.
2. Miyake Y. Electrodiagnosis of Retinal Disease. New York: Springer; 2006.
3. Lam B. Electrophysiology of Vision: Clinical Testing and Applications. New York: Taylor & Francis; 2005.
4. Fishman GA, Birch DG, Graham EH, Brigell MG. Electrophysiologic Testing in Disorders of the Retina, Optic Nerve, and Visual Pathway, 2nd Ed. San Francisco: The Foundation of American Academy of Ophthalmology; 2001

Online:

Webvision – Basic science and clinical ERGs: <http://webvision.med.utah.edu>