Multifocal Visual Evoked Potential in Nonorganic Visual Field Loss

Emma C Massicotte, MD; Linda Semela, MD; Thomas R. Hedges III, MD

Objective: To evaluate the use of multifocal visual evoked potentials in the diagnosis of nonorganic visual field loss.

Methods: Five patients with unexplained visual field loss underwent full neuro-ophthalmic examination, Humphrey visual field testing, and multifocal visual evoked potential testing using the Accumap objective perimeter (ObjectiVision Pty Ltd, Sydney, Australia).

Results: In all 5 cases, the results of the ophthalmic examination did not correlate with the degree of visual field loss seen on Humphrey visual field testing. Multifocal visual evoked potentials testing showed essentially normal tracings.

Conclusions: Multifocal visual evoked potentials may be useful in cases of difficult to prove functional visual field loss or in cases in which objective documentation of normal function is needed.


FunctionaL Visual Loss can be identified frequently by thorough clinical evaluation. However, uncertainty about the diagnosis may remain after the ophthalmologic examination. Furthermore, in some cases, functional disease coexists with organic disease. In these situations objective measurements may prove helpful in confirming suspected functional visual loss. Early identification of patients with a functional disorder is crucial to avoiding unnecessary and costly testing or invasive procedures.

Standard visual evoked potential (VEP) has been used to aid in the diagnosis of functional visual loss in the past and may be useful in cases in which the functional deficit is decreased central visual acuity. However, because bright flash VEP stimulates the entire anterior visual pathway and pattern evoked VEP reflects macular function, localized visual field defects may be missed owing to the averaging of the abnormal responses with the normal ones. The multifocal visual evoked potential (mfVEP) offers the advantage of obtaining signals from up to 60 sectors across 30º of the visual field without reliance on patient responses.

We evaluated 5 patients with presumed functional visual field deficits in which mfVEP was useful in establishing the diagnosis. In one case, invasive surgical optic nerve sheath decompression was avoided.

METHODS

The Tufts–New England Medical Center Institutional Review Board approval was obtained prior to conducting this study. Informed consent was obtained from all patients and the study complied with Health Insurance Portability and Accountability Act of 1996 regulations at all times. The patients underwent full neuro-ophthalmic examination, Humphrey visual field (HVF) testing (program 24-2, SITA Fast or Standard; Allergan, Irvine, Calif), and mfVEP testing using the Accumap perimeter (ObjectiVision Pty Ltd, Sydney, Australia)

Multifocal VEP testing was performed in each eye by technicians trained in the technique. Subjects were seated in a chair in front of the computer screen, then 4 gold disk electrodes were positioned on the scalp with the use of a custom-designed occipital cross-electrode holder. The visual stimulus was generated on the computer screen with 56 closely packed segments consisting of 16 alternating black-and-white checks within each segment arranged in a dartboard configuration. The checks alternated in pseudorandom sequences and VEP responses were recorded for each segment by the cross-correlation method described by Sutter. The duration of testing was approximately 8 minutes per eye with an average of 8 recordings per eye to provide a good signal-noise ratio.
RESULTS

CASE 1

A 56-year-old woman was evaluated 5 months after she suddenly noticed a left temporal field deficit after seeing a “thunderbolt” of bright light with an area of visual dimming temporally in her left visual field. She had had a history of narrow anterior chamber angles treated with bilateral laser peripheral iridectomies and normal visual fields years before the episode described herein. Medical history included migraine headaches for which she took a combination of acetaminophen, caffeine, and butalbital (Fioricet). The patient expressed concern that the field deficit was interfering with her ability to perform at work as a medical secretary.

Snellen visual acuities were 20/20 OU with normal color vision and no afferent pupillary defect. Her optic discs and retinas appeared normal. Optical coherence tomography (OCT) revealed normal and symmetric nerve fiber layer thicknesses. Magnetic resonance imaging (MRI) showed mild sinus mucosal thickening and some small, apparent, ischemic changes consistent with migraine.

Humphrey visual field testing showed a temporal hemianopic deficit in the left eye with greater involvement of the inferior quadrant (Figure 1A). Multifocal VEPs were entirely normal (Figure 1B).

She returned 1 year later. The visual field defect previously noted in the left eye had disappeared, but there was a new, right, temporal, visual field defect.

CASE 2

A 32-year-old woman was evaluated because of bilateral constriction of visual fields with reports of transient episodes of blurred vision and difficulty with night vision for the past 3 years. She stated that the visual problems caused her to leave her job as a cashier 3 to 4 months earlier. She had an ocular history of strabismus, amblyopia in the left eye, and congenital nystagmus in both eyes. Medical history included occasional frontal headaches and low back pain for which she took naproxen regularly. The findings from a review of the systems were otherwise unremarkable. She had had several computed tomographic scans in the recent past that also showed no abnormalities.

Examination revealed best-corrected Snellen visual acuities of 20/60 OD and 20/400 OS, normal color vision in the right eye, and near-normal color vision in the left eye, missing only the dimmest American Optical Hardy-Ritter-Rand (AOHRR) color plate. The remainder of her examination was notable for very fine irregular nystagmus on right and left gaze with a slight torsional component, an 18-diopter esodeviation, which was essentially unchanged in all gaze positions, and somewhat irregular pursuit movements. The right optic nerve and both retinas appeared normal, while the left optic nerve showed mild tilting.

Automated visual field testing with Humphrey program 24-2 showed generalized constriction of both visual fields to 5° on the right and 10° on the left (Figure 2A). Multifocal VEP recordings displayed some signal decrease in a small area superotemporally in the right eye, and some decrease in amplitude with increased latency superiorly that could be explained by eyelid artifact (Figure 2B). These changes did not correspond to the degree of visual loss that was seen on HVF testing.

CASE 3

A 35-year-old woman with a 9-year history of pseudotumor cerebri was referred for evaluation and consideration for possible nerve sheath fenestration. During the
previous 9 years, she had undergone 55 spinal taps and was treated with variable doses of acetazolamide and furosemide. Her weight fluctuated between 81 and 153 kg. Most recently she weighed 126 kg and was taking between 1500 and 2000 mg/d of acetazolamide. Her condition had been relatively stable until 10 months earlier when she began to have worsening headaches, syncopal episodes, transient visual obscurations, difficulty with peripheral vision, and horizontal diplopia. The opening pressure of her latest spinal tap was 290 mm H2O.

She had polycystic ovarian disease, diabetes mellitus, asthma, depression, and nephrolithiasis requiring 2 previous surgical procedures. Medications included acetazolamide, furosemide, potassium chloride, metformin hydrochloride, famotidine, budesonide, salmeterol, gabapentin, clonazepam, citalopram, and trazodone hydrochloride.

Neuro-ophthalmic examination showed Snellen visual acuities of 20/25 OD and 20/30 OS, normal color vision with the AOHR plates, and normal intraocular pressures in both eyes. Findings from the remainder of the examination were normal with the exception of the optic nerves, which had somewhat irregular optic disc margins consistent with probable previous swelling, but very little, if any, swelling at the time of the examination. The retinal nerve fiber layer appeared intact bilaterally. Optical coherence tomography showed evidence of mild diffuse nerve fiber layer thinning with mean readings of 73 µm OD and 86 µm OS (normal, 100 µm ±10 µm).

Humphrey 24-2 visual field testing showed constricted visual fields, more on the left than the right (Figure 3A). Multifocal VEP was essentially normal with a few abnormal tracings superiorly in both eyes (Figure 3B).

CASE 4

A 29-year-old woman was initially evaluated because of visual field loss in the left eye 1 year after being diagnosed as having pseudotumor cerebri, based on headaches, obesity, and several elevated intracranial pressure measurements. She had a medical history of asthma, depression, and polycystic kidney disease. Medications included oral acetazolamide, 500 mg twice a day; fluticasone propionate, zafirlukast, albuterol sulfate citalopram, loratadine, and an oral contraceptive. Results of the ophthalmic examination showed no evidence of optic disc swelling or nerve fiber layer thinning. However, there was inferonasal depression on HVF testing in the left. Acetazolamide therapy was continued and weight loss was encouraged.

During the next few years she had frequent hospital admissions for possible seizures, syncope, headaches, nausea, and vomiting. She underwent lumboperitoneal shunting, requiring 4 revisions. During this time her ophthalmic examination continued to show an inferonasal visual field defect in the left eye without papilledema or nerve fiber layer thinning. Then, 3 years after her initial manifestation, she began to report visual field changes in the right eye. Again, there was no papilledema and nerve fiber layer thicknesses were stable by OCT measurements. However, HVF testing revealed progression of a visual field deficit in the left eye, sparing only the superotemporal quadrant, and a new inferonasal defect in the right eye (Figure 4A). Multifocal VEP was normal in both eyes, except for a few areas with slight increased latency paracentrally in both eyes (Figure 4B).

CASE 5

A 14-year-old girl was initially seen in the emergency department reporting eyelid swelling and decreased vision in the left eye for 1 day. She had a medical history of migraine, asthma, attention-deficit/hyperactivity disorder, and depression.
On ophthalmic examination, visual acuity was 20/20 OD and 20/300 OS. She was, however, able to see 8 circles on the Titmus stereo vision test. Findings from the remainder of the examination were normal with the exception of complete loss of visual field in the left eye when tested by confrontation and Humphrey 24-2 automated perimetry (Figure 5A). Multifocal VEP showed a full field of normal amplitudes with random areas of increased latency on both sides (Figure 5B).

Thorough clinical examination is usually sufficient to diagnose nonorganic disease. However, a particularly savvy functional patient can make distinguishing a true field deficit from a functional field deficit very difficult. Thompson et al2 found that with minimal instruction, some study subjects could easily imitate and reproduce visual field defects on both automated and manual perimetry. In addition, experienced technicians were actually deceived by the subjects more often than the inexperienced technicians owing to their systematic approach to perimetry.

The use of multifocal VEP in diagnosing functional visual loss was reported by Miele et al3 in 2000. In their patient with an inferior bitemporal quadrantopia respecting the horizontal meridian, HVF testing showed normal multifocal VEP tracings in the area of visual field defects providing further evidence of the suspected functional loss.5

Multifocal VEP has been studied primarily for use in the investigation of glaucomatous visual field deficits. It has been shown to be 95% to 97% sensitive for glaucomatous scotoma detection in clinical trials. In contrast with HVF testing, the objective perimetry provided by multifocal VEP is less affected by patient performance or learning curve.6

The technique evaluates the pathway from the retinal receptors to the occipital cortex and assesses the visual field out to 30º using a multifocal pattern VEP stimulus, with multichannel recording. The rapidly alternating checkerboard pattern derives which signal response in the brain came from which location in the field by cross-correlating the signal recorded with the pattern reversal on the screen.2

In this case series, multifocal VEP helped to confirm 5 suspected cases of functional visual loss, initially seen as unilateral temporal visual field loss (patient 1), unilateral complete visual field loss (patient 2), 2 cases of bilaterally constricted fields (patients 3 and 4), and 1 case of a peculiar field deficit sparing only the superotemporal quadrant in the left eye, with an inferonasal defect in the right eye (patient 5). In patient 3, the diagnosis of functional visual field loss allowed the patient to avoid optic nerve sheath fenestration.

In 1986, Gittinger7 reported 4 cases of functional monocular temporal hemianopsia suggesting that this type of deficit, as seen in patient 1 of our series, is a relatively common and characteristic pattern of functional visual loss. When bilaterally constricted fields seen on automated perimetry (patients 3 and 4) fail to expand at increased testing distance when tested manually, functional visual loss is the likely diagnosis.7 Unlike multifocal VEP, however, tangent screen and Goldmann visual field reliability are often dependent on the skill and experience of the examiner.

The multifocal VEP was found to be particularly helpful in the 3 patients with coexisting ocular pathology; in patient 2 who had strabismus, amblyopia in the left eye, and congenital nystagmus; and in patients 3 and 4 who had pseudotumor cerebri. The high frequency of such cases may be related to the suggestion that a patient with a history of an ocular pathologic condition is predisposed to developing functional loss at a later time.8 This study suggests that multifocal VEP may be useful in cases of difficult to prove functional visual loss or in cases in which objective documentation of normal function is needed.

Submitted for Publication: December 30, 2003; final revision received July 23, 2004; accepted July 23, 2004.

Correspondence: Thomas R. Hedges III, MD, 750 Washington St, Box 450, Boston, MA 02111 (thedges@tufts-nemc.org).

REFERENCES