

Increasing Diagnostic Potential in Pediatric Optometry by Electrophysiological Methods

J.V. Lovasik*
M.E. Woodruff**

Abstract

With the advent of modern-day technology light-induced electrical activity in the retina and visual cortex can be measured clinically to assess visual function. The Electroretinogram (ERG) and the Visually Evoked Response (VER) are now routine clinical tests used to evaluate the physiological integrity of retinal and visual cortical mechanisms respectively. The theory, and application of these non-invasive, objective tests in the evaluation of various aspects of vision in infants and children are discussed. Numerous case reports are presented to illustrate the extended diagnostic capability afforded the pediatric optometric practitioner by the ERG and VER.

Abrégé

La technologie moderne nous permet de mesurer le potentiel électrique de la rétine et du cortex induit par des éclairs contrôlés de lumière et ainsi vérifier l'intégrité de la fonction visuelle. Le ERG et le VER sont des tests routiniers utilisés pour vérifier l'intégrité physiologique des structures de la rétine et du cortex visuelle respectivement. La théorie et l'application de ces procédures "non-chirurgicales" et objectives dans l'enquête même sur la vision des enfants sont expliquées. On présente plusieurs histoires de cas pour illustrer comment la capacité diagnostique du praticien en pédiatrie est rehaussée par le ERG et VER.

Introduction

Vision literature abounds with statements on the necessity of early detection and remediation of visual anomalies to prevent amblyopia, and irreversible sensory deficits.¹⁻⁵ In a series of papers, Ingram⁶⁻⁸ has reported changes in refraction in children between one and three years of age which indicate the necessity for assessment during the first year of a child's life, as well as continuing frequent surveillance if amblyopia is to be prevented. Thomas and Mohindra⁹ have shown that most infants respond quickly to amblyopia therapy lending further support to the necessity of early correction. Despite this overwhelming evidence the majority of children under five years of age remain without vision care until they enter school or are found, often inadvertently, to have a vision problem. This situation can be corrected by Optometrists becoming politically more active in the implementation of preschool screening programs^{10,11} and educating parents and persons of child bearing age to seek optometric care for their children at a very early age. Another area where optometrists must spend time is in educating family physicians and pediatricians of the necessity for an early vision examination.

The University of Waterloo Pediatric clinic patient load provides evidence that the activity in preschool screening programs results in a large number of children being referred for further optometric care. As the majority of this clinical population is made of children under five, a significant portion of the oculovisual assessment necessarily

involves objective testing procedures which have been described elsewhere.¹²

While standard clinical procedures such as retinoscopy, keratometry, and cover tests, can yield accurate assessments of ocular refraction and binocularity, the practitioner until recently has had to rely on behavioral responses of the child over a period of weeks to months to evaluate the effectiveness of any applied optical therapy. Subtle irregularities in the appearance of the ocular fundi in the absence of visual symptoms have often been classified "within normal limits" by optometrists and ophthalmologists alike. This was largely because accurate measurements of visual function could not be carried out in a majority of children under three by the standard optometric means. However, the advent of non-invasive electrodiagnostic tests of visual function has greatly expanded and improved the diagnostic capability of the optometric practitioner as well as providing him with a powerful clinical tool with prognostic value. The clinical feasibility of electrodiagnostic tests has added broad, new dimensions to the visual examination of infants and non-verbal populations.

It is the purpose of this paper to describe briefly the utility of two electrodiagnostic procedures in the detection of abnormalities in the visual system of infants and children. Whereas a systems examination¹² provides valid and reliable data on which a practitioner can originate therapy, the ability to access an electrodiagnostic service can reassure the practitioner that therapy

*B.Sc., M.Sc., O.D., Ph.D., F.A.A.O.

**O.D., Ph.D., F.A.A.O.
School of Optometry
University of Waterloo

based on both data sources will optimize binocular visual input. The practitioner who obtains such information can thus be more effective in prescribing, counseling, and reassuring the child's parents that the diagnosis and prognosis are sound since he has direct physiological evidence of the value of corrective lenses or prisms which form the basis of the therapy recommended.

The two main electrodiagnostic tests of visual function employed at the University of Waterloo, Electrodiagnostic Clinic, are 1) Electroretinography (ERG) and 2) the Visually Evoked Response (VER). A brief description of these tests together with case examples illustrating the usefulness of these procedures in evaluating various aspects of the visual system in infants, children and adults follows.

Electroretinography

Diffuse Flash ERG's

Electroretinography¹³⁻¹⁵ is an electrodiagnostic procedure that assesses objectively the functional integrity of outer and inner layers of the retina. The testing procedure involves the painless application of the recording electrode close to the eyeball near the lid margin (periorbital electrodes) or directly on the cornea (corneal electrodes) and measurement of electrical changes occurring within the eye in response to quantified diffuse flashes of light. (See Fig. 1-A). A normally functioning retina is identified by electrical responses with characteristic polarities and waveforms. A classical ERG is composed of "a", "b", "c" and "d" waves. The clinically useful components of the ERG pattern are the "a" and "b" waves. (See Fig. 1-B). Animal experimentation and clinical studies suggest that the cornea-negative "a" wave is generated by the photoreceptors (rods and cones) while the cornea-positive "b" wave is thought to be produced primarily by the Bipolar and/or Mueller cells. While it is generally accepted that the ganglion cell layer and the optic

nerve do not contribute to the flash ERG, the role of ganglion cell activity in the generation of the "pattern ERG"¹⁶ is at present an unsettled issue.^{17,18}

A. Scotopic ERG's

The functional integrity of the rod type retinal photoreceptors can be evaluated by dark adapting the patient and measuring the retinal response to specially selected light stimuli. The dark adaptation procedure maximizes the sensitivity of the rods thereby allowing an assessment of the physiological responsiveness of those retinal elements primarily subserving night-time vision. When fully dark adapted, depending on patient compliance, either periorbital or corneal ERG electrodes are used to record the ERG. Light flashes are presented within a ganzfeld stimulator¹³ which provides uniform retinal illumination. Photo-induced retinal responses to several brief light flashes are then

averaged by a computer of averaged transients. These test flashes ordinarily consist of scotopically matched blue and red light, as well as white light. These test flashes elicit the "rod isolated" (exclusively rod responses), "rod dominated" (primarily rods with some cone contribution), and "mixed" ERGs (both rod and cone responses) respectively. The waveform of the ERG representing the average of a preselected number of consecutive ERGs is analyzed by measurement of the "a" and "b" wave latencies, amplitudes and implicit times (peak times). These parameters are then compared to established norms to arrive at a diagnosis concerning retinal function.

Figure 2 illustrates scotopic ERGs obtained with periorbital electrodes for a patient with normal rod-cone function, and a thirteen year old girl (LB) who was diagnosed as having retinitis pigmentosa sine pigmento. LB presented with symptoms of impaired vision at night-time with

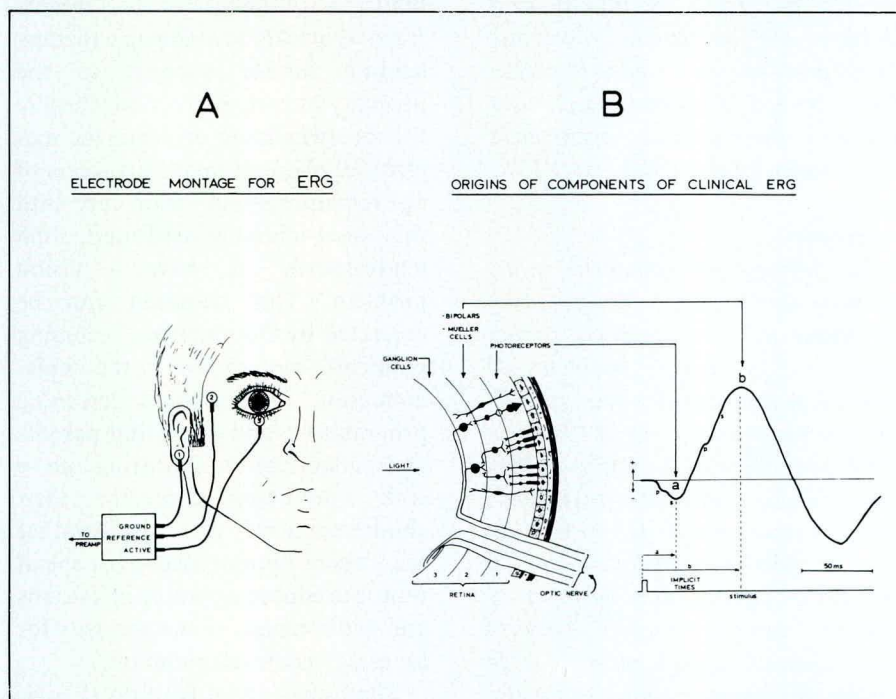


Fig. 1

A) Schematic diagram showing the relative positions of the active electrode (3), reference electrode (2), and ground electrode (1) used to record the ERG. Each surface electrode is held in place by small strips of adhesive tape. Good electrical contact between the skin and the electrodes is insured by an electroconductive gel placed on the contact surface of the electrodes. The periorbital positioning of these

electrodes allows ERG recordings in apprehensive adults and children.

B) Sagittal view of the retina showing the first (photoreceptors) second (bipolars) and third (ganglion cells) order neurons in the visual system. An idealized ERG with the presumed neural generators of its primary components is shown to the right of the retina. Note that the ganglion cells do not contribute to the ERG.

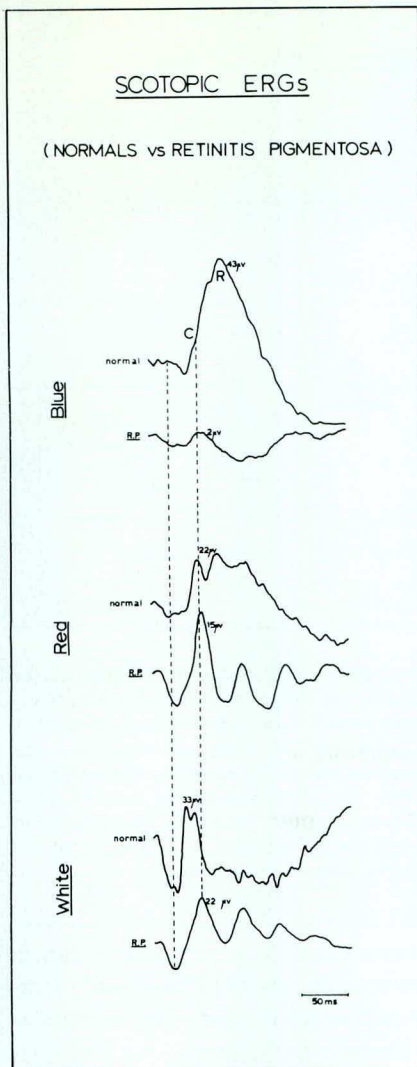


Fig. 2

Scotopic ERGs elicited from a normal adult (top traces for each type of light flash) and a 13 year old girl with retinitis pigmentosa (bottom traces for each flash). Each record is the average of twenty, 200 msec responses recorded with periorbital electrodes positioned as illustrated in Fig. 1A. The vertical dashed lines facilitate comparison of the salient temporal characteristics of the ERGs for the normal and RP patient. R and C designate the rod and cone contribution to the 'b' wave respectively. Note that the ERG of the retinitis pigmentosa patient is lacking a rod component and therefore is primarily a cone dominated response.

daytime vision and visual acuity being "normal". An ophthalmoscopic examination did not reveal any peripheral pigmentary changes characteristic of retinitis pigmentosa. In the case of the normal observer, a blue flash elicited an ERG with a prominent "b" wave. A red flash elicited an ERG with a double-peak "b" wave. The first peak in the "b" wave corresponded to the contribution of the faster reacting cone

photoreceptors to the scotopic ERG while the second peak represented the input of the slower rod photoreceptors. A white flash elicited a large amplitude ERG with a prominent "a" and "b" wave. Note that the peak of this "b" wave occurred earlier than the peak of the "b" wave in the rod dominated ERG elicited by the red flash, which in turn occurred before the peak of the rod isolated ERG "b" wave elicited by the blue flash. In the case of the youngster with retinitis pigmentosa, a blue flash failed to elicit any significant response from the rods. Note that the blue light elicited a small "a" wave and a small portion of the "b" wave. The implicit time of this small peak coincided with the photopic (cone) contribution to the scotopic ERG elicited by a red flash. A red flash elicited a prominent "a" wave and "b" wave with a waveform that was distinctly different from that seen in the normal observer. The peak of the "b" wave coincided with the first peak in the "b" wave of the ERG for the normal observer. This indicated a strong response from cone photoreceptors without a response from the rods. A number of oscillations of decreasing amplitudes followed the cone dominated "b" wave. A white flash elicited a prominent ERG, with the peak of the "b" wave coinciding in time with the peak of the "b" wave elicited by the red and blue light flashes. The implicit time of the "b" wave in this case was somewhat delayed when compared to the normal response. The nature of the ERG responses described above indicated a pathological dysfunction of rod photoreceptors with normally functioning cones. This example illustrates that retinal physiology may be severely impaired even when the ocular fundus appears normal on ophthalmoscopic examination and emphasizes the need to evaluate retinal physiology before ruling out retinal disease. The evaluation of retinal physiology is equally important when fundus signs suggest retinal disease (e.g. pseudo-retinitis pigmentosa) but where no functional impairment exists.

B. Photopic ERGs

Retinal cone function can be readily assessed by light adapting the patient for about a five minute period to a 10 ft. L, white light background within a ganzfeld. This light adaptation procedure maximizes the sensitivity of the cone photoreceptors thereby allowing an assessment of the physiological responsiveness of those retinal elements primarily subserving daytime vision. Then the same stimulus and analysis procedures described above for scotopic

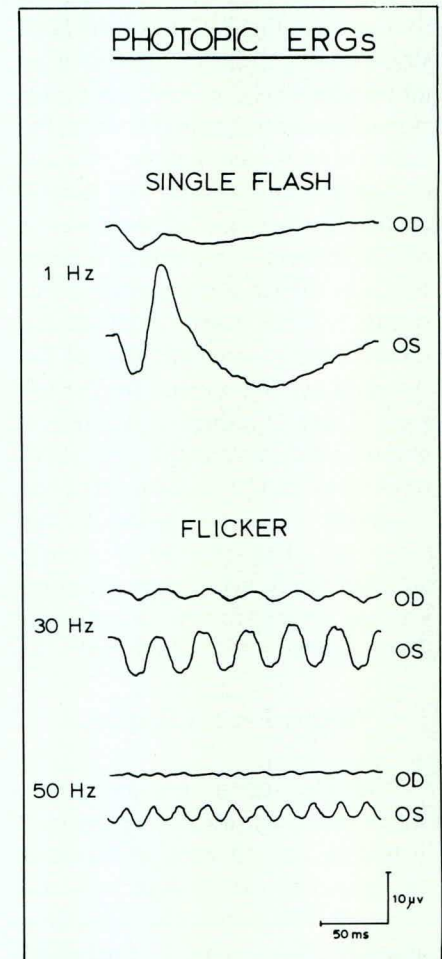


Fig. 3

Photopic ERGs elicited for the right and left eye of an adult patient. Each trace is the average of twenty, 200 msec. responses. Single flash ERGs elicited by a white flash indicated an abnormal response for the right eye. A selective minification of the 'b' wave characterized the ERG for the right eye. Flicker ERGs elicited by flashes delivered at a frequency of at 30 Hz and 50 Hz assumed a typical sinusoidal waveform corresponding to a series of connected 'b' waves. Under light adaptation conditions, and high flash frequencies, the ERG is generated exclusively by cones. The reduced amplitude of the flicker ERG for the right eye 30 Hz and extinguished response at 50 Hz indicated a gross abnormality of retinal physiology.

ERGs are repeated for the light adapted observer. Cone function can be assessed further by eliciting "flicker" ERGs with red light flashes delivered at a high flash frequency, typically 30-60 Hz. The combined conditions of light adaptation, high flash frequencies and the use of light with a spectral composition restricted to the long wavelength portion of the visible light spectrum insure that the responses are generated exclusively by the cones. Failure to elicit either a single flash photopic ERG or the flicker ERG indicates a gross cone dysfunction. Figure 3 illustrates both single flash and flicker ERGs for an adult who likely suffered an occlusion of the central retinal artery in the right eye. It is seen that the "b" wave of the photopic ERG was greatly decreased while the "a" wave was of nearly normal amplitude. Flicker ERGs at 30 Hz were detectable but greatly reduced in size. At 50 Hz the flicker ERG's were extinguished for the right eye but normal for the left eye. These findings indicated a pathological abnormality of retinal physiology and provided a very poor prognosis for normal vision. In this case, the diagnosis of a poorly functioning retina prevented unrewarding surgery for the removal of a dense cataract in the right eye.

Visually Evoked Responses

The electroretinographic procedures described above are useful in detecting disturbances of vision at the retinal level by flash scotopic and photopic ERGs. It is also possible to assess the function of the final stages of the "input" or sensory component of the visual system by a relatively easy to perform but technically sophisticated technique called the Visually Evoked Response (VER).¹⁹ This technique measures the functional integrity of the neural pathways originating at the macular area of the retina and terminating in Brodmann's area 17 of the visual cortex. The VERs are therefore indirect measures of macular function and when combined with flash ERGs are useful in identifying

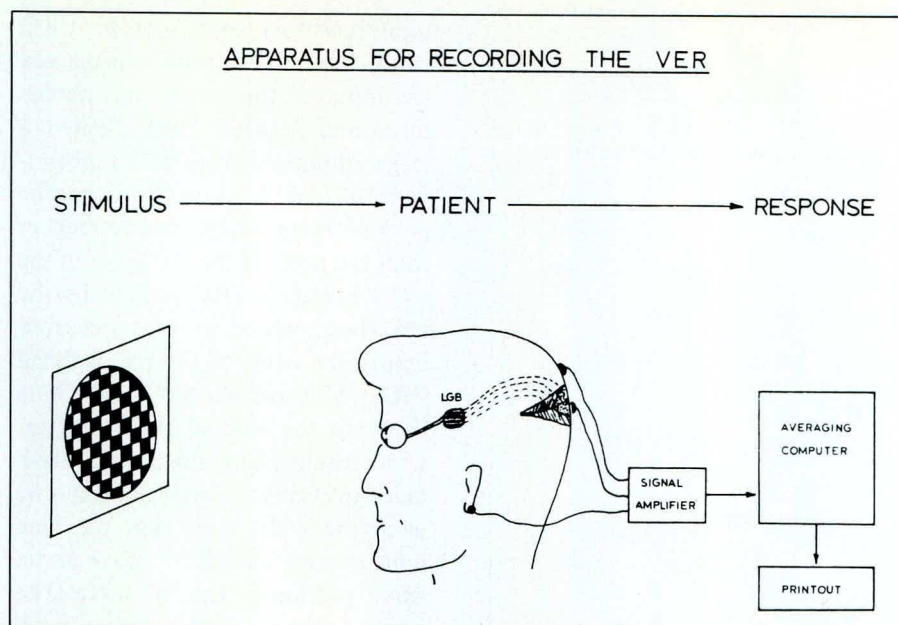


Fig. 4

Schematic illustration of the recording setup for the Visually Evoked Response (VER). The stimulus consists of a checkerboard made up of black and white checks temporally modulated in counterphase. Cortical responses to the checkerboard stimulus are recorded by surface electrodes positioned over the cortical projection of macular areas in the retina. Visually related electrical activity

(microvolt range) within the occipital cortex detected by the surface electrodes is differentially amplified and subsequently processed by a computer of averaged transients. The resulting waveform approximates a sinusoid when the checkerboard is reversed at a frequency greater than 4Hz. The number of peaks in the sinusoid is a function of the checkerboard reversal frequency.

retrobulbar disturbances in afferent visual fibers.

The recording of VERs involves placing scalp electrodes over the macular projection to the occipital cortex and computer averaging of the cortical activity elicited by a specially designed visual stimulus. Since the macular area of the retina projects onto the peripheral aspects of area 17 within the calcarine fissure, an electrode positioned 1-2 cms above the inion will record the neural activity resulting from visual excitation of the macular and foveal areas of the retina. (See Fig. 4).

The VER can be elicited by diffuse flashes of light to assess light perception and the integrity of frequency channels in the visual system, or patterned stimuli to assess spatial information channels. The visual stimulus evoking maximal cortical activity for the assessment of form vision is known as the "reversing checkerboard". This stimulus consists of a checkerboard pattern that can be electronically generated on a black and white television screen. The size of each check can be

varied and adjacent squares can be alternately shifted ("reversed") from black to white, and white to black at selected frequencies. At any one time, half of the total number of checks visible are black while the other half are white. This arrangement creates a stimulus with distinct pattern reversals but no net change in luminance. Since the average luminance of such a target remains constant, the foveal elements are responding exclusively to the presence of the check pattern within the stimulus and not luminance changes. This type of pattern also is very effective in eliciting strong responses from the visual cortex because most cells resident in the visual cortex are maximally stimulated by complex patterns with sharp borders.

The frequency at which the checkerboard target is "reversed" (checks alternated from white to black to white) determines the waveform of the evoked potentials. At low reversal frequencies (1-3 Hz) visual neurons are activated by the abrupt reversal of the checkerboard pattern resulting in a "Transient

VER" that is characterized primarily by a small negative wave (N-1) preceding a large positive wave (P-1) which has an implicit time (peak time) of approximately 105 milliseconds (SD \pm 5 milliseconds). The amplitude and implicit time of the P-1 component of Transient VERs are important parameters in the clinical evaluation of the functional integrity of macular-cortical pathways. The right hand side of Figure 5 illustrates the Transient VER waveform elicited under monocular and binocular

of the nervous system (multiple sclerosis).

When the checkerboard is reversed at high frequencies (8-16 Hz) the continuous oscillation of the target causes a rhythmical discharge of visual neurons such that the recorded potentials approximate a sinusoid with the number of peaks in the sinusoid being directly related to the checkerboard reversal frequency. The cortical response pattern elicited by a continuously reversing target is shown as the "Steady-State VER".

temporal characteristics, the patient retained a 20/20 level of visual acuity. This latter observation highlights the fact that visual acuity measurements should be considered as only coarse measurements of visual function and ocular health.

Various aspects of vision such as visual acuity, refractive error, and binocular function can be examined objectively by varying the VER stimulus determinants. In addition, VERs can be used as sensitive indicators of sub-clinical and manifest ocular diseases affecting the macular area of the retina, or retrobulbar components of the visual system. The utility of the VER in assessing ocular health and the above mentioned visual functions in children and adults is illustrated in the following sections by case examples.

Objective Assessment of Visual Acuity

The following example illustrates the utility of the patterned VER in assessing the resolution function of the human visual system. A four month old infant (LL) underwent a standard oculovisual assessment. The results of the examination revealed a normal appearance of the ocular tissues and adnexa. An ocular motility assessment revealed that the eyes moved freely into all cardinal positions of gaze, and an occasional symmetrical convergence could be induced. The Hirschberg corneal reflex test indicated a non-strabismic condition for near and far fixation. An evaluation of ocular health indicated brisk direct and consensual pupillary light reflexes, clear ocular media, with normal appearance of the optic nerve heads and ocular fundi. An evaluation of ocular refraction indicated a low hyperopic refractive error (+0.75 DS) in each eye. Altogether, there were no indications of any abnormalities within the infant's visual system that could preclude normal visual development. In order to quantify the infant's visual resolution function, she was referred to the Electrodiagnostic Clinic for patterned VER testing.

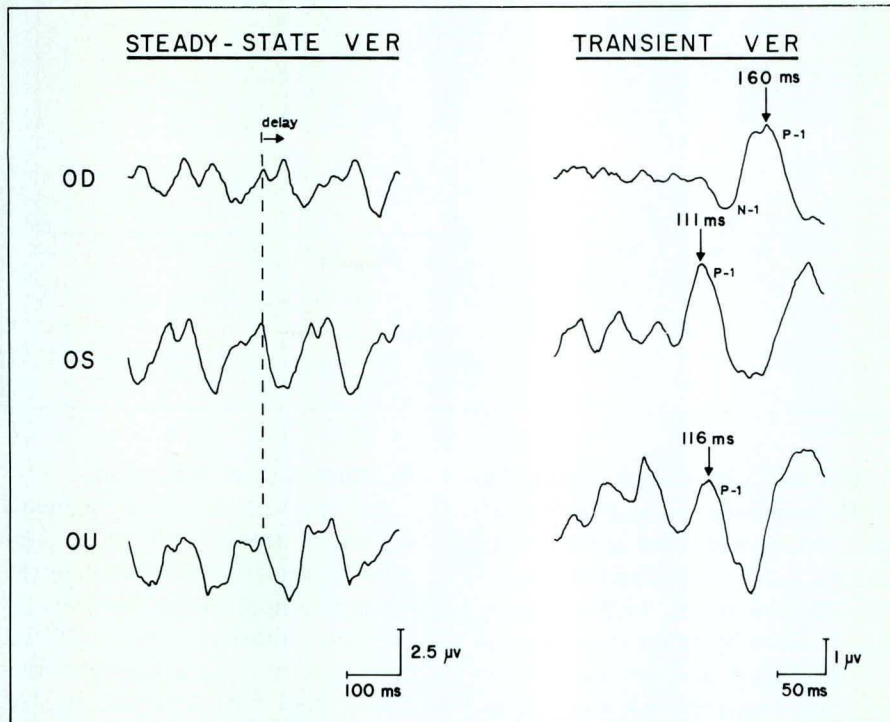


Fig. 5

Sample records illustrating the waveforms of the steady-state and transient VERs obtained for a 42 year old male with multiple sclerosis. Steady-state VERs were elicited by a 6° checkerboard composed of 14 minutes of arc black and white checks reversed at a frequency of 8 Hz. Transient VERs were elicited by the same checkerboard pattern reversed at a frequency of 2 Hz. Each trace is the average of 30 responses. The steady-state VER for the right eye was reduced in amplitude and had a slightly irregular

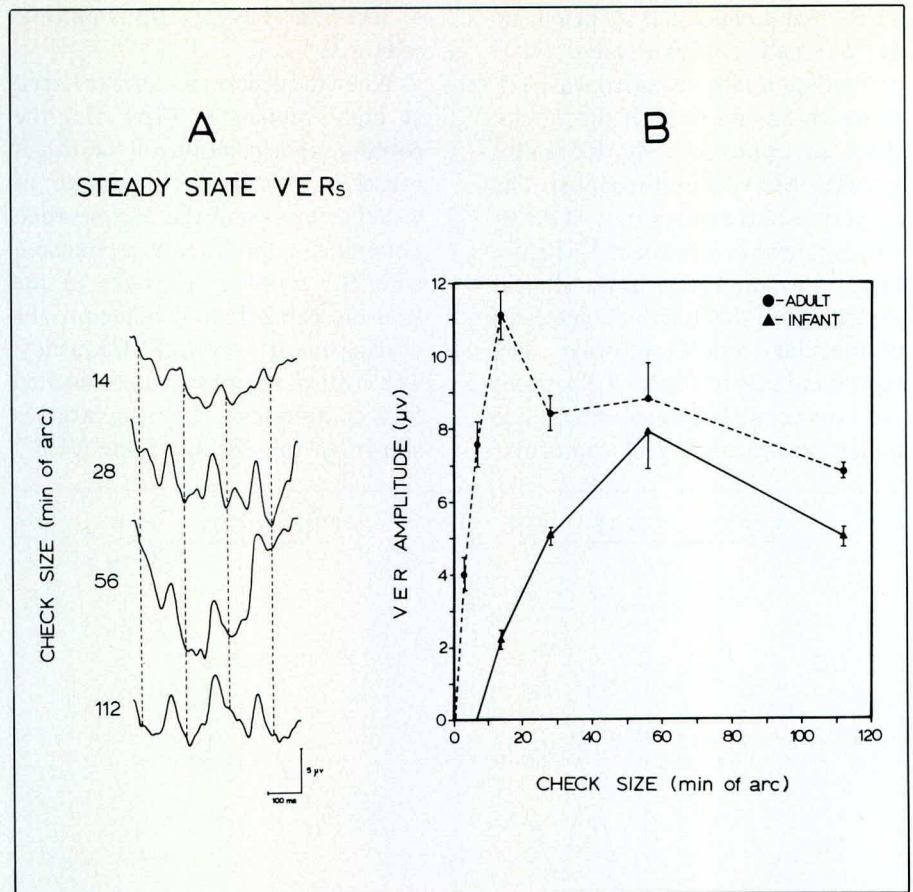
waveform compared to the VER for the left eye. A comparison of the time of occurrence of the peaks in the steady-state VERs for the right and left eye indicates a delayed response for the right eye (small horizontal arrow). The difference in retinal-cortical transmission times (approximately 49 msec) was also seen in the transient VERs. The implicit time of P-1 for the left eye was within normal limits. The binocular transient VER was slightly degraded in amplitude compared to the monocular VERs. This suggested some negative interaction between the cortical input from each eye.

viewing conditions. It is noted in this example that the implicit times of the P-1 component for the right eye (160 msec) and left eye (111 msec) differ greatly with the right eye showing the much slower response. In this example, the very significant delay in retinal-cortical neural transmission was the ocular manifestation of a generalized demyelination disorder

The left hand side of Figure 5 presents steady-state VERs recorded from the same patient. Note the clear sinusoidal waveform elicited for the left eye. The VER waveform was slightly degraded and delayed for the right eye likely due to multiple sclerotic lesions in the right optic nerve. However, despite this alteration of the VER waveform and its

Fig. 6

- A) Binocular steady-state VERs elicited from a 4 month old infant using a 12° checkerboard with 14, 28, 56 and 112 minutes of arc checks reversed at a frequency of 8 Hz. Each record is the average of 100, 500 msec responses. The vertical dashed lines isolate the peaks within the VERs. Shifts in the baseline were caused by the infant's head movements. Despite these irregularities in the recordings, the peaks within each trace were clearly visible.
- B) Graph illustrating the averaged amplitude of the peaks in the VER as a function of check size for the infant whose VERs are illustrated in A. The vertical bars through each data point represent ± 1 SEM (standard error of the mean) for VER amplitudes. The VER amplitude for this infant peaked for check sizes subtending 56 minutes of arc. For comparison, the same function is illustrated for the infant's thirty-four year old father whose visual acuity was 20/20. As is typical for most adult observers having a visual acuity of 20/30 or better, the VER peaked for check sizes of about 15 minutes of arc.²⁹ Thus a check size about 4 times as large as that eliciting the maximal VER in the adult is required to elicit the maximal VER in the infant. In this case, one can therefore predict a potential visual acuity of approximately 20/80 or better for the infant.



The results of the electrodiagnostic assessment are illustrated in Figure 6A. Binocular steady-state VERs were successfully recorded from the infant using the standard checkerboard target positioned one meter away from the infant's facial plane. The ambient light in the examination room was reduced to a minimum to induce the infant to fixate the only luminous object in the room, the checkerboard pattern. The averaging computer was triggered manually whenever the reflex of the checkerboard pattern was judged to be within the infant's pupillary area. One hundred, 500 millisecond responses were averaged in order to obtain a good signal-to-noise ratio. The results indicated that there was a recordable but poorly defined VER pattern in response to a checkerboard target composed of 14 minutes of arc black and white checks. The amplitude of the binocular VER increased as the checkerboard check size was increased to approximately 56 minutes of arc, and subsequently decreased slightly as the checkerboard check size was increased

further still. Figure 6B illustrates the VER amplitude as a function of check size for the infant, as well as the infant's father. This graph illustrates that the maximum VER response was obtained by different check sizes for the infant and the adult. This indicated that the adult was able to resolve a target with finer check size than the infant. Furthermore, a check size four times as great as that eliciting the maximal VER response from the adult was required to obtain the maximal VER response from the infant. Since the father's best visual acuity was 20/20, the VER measurement suggested a potential visual acuity of approximately 20/80, or better, for the infant. This prediction of a resolution capability equivalent to 20/80 or better was consistent with the reports by Sokol,^{20,21} and Marg,²² who also estimated the visual acuity of four month old infants to be between 20/80 and 20/40. In this case, therefore, the VER analysis extended the findings obtained in the standard pediatric oculovisual assessment to include a quantitative evaluation of visual acuity.

B. Physiological Refraction

An evaluation of a patient's refractive status by retinoscopy provides valuable information on the limitations imposed on visual resolution by refractive errors. Optical correction of any existing refractive errors does not, however, predict normal visual acuity since there may be limitations imposed on vision by neural disorders at the retinal level or higher visual centers. The prediction of resolving ability becomes even more problematical when dealing with infants and non-communicating adults. It is possible, however, to determine with greater certainty whether an ophthalmic prescription will improve visual function in non-communicating patients by means of the VER. It is well known that the amplitude and waveform of the VER is influenced by the sharpness of the retinal image,²³ as well as the functional integrity of the macular cortical pathways.²⁴⁻²⁶ Thus by monitoring the "strength" of the neural impulses arriving at the visual cortex, one can determine the likely benefit, if any, that a patient will

derive from an ophthalmic prescription which modifies the sharpness or stability of the retinal image. Thus a lens enhancing the cortical response would be considered necessary, while a lens either decreasing the cortical signal or having no significant effect on it would be considered unnecessary. The following three cases will illustrate the usefulness of the patterned VER in determining the need for an ophthalmic prescription on a totally objective basis.

Case No. 1: Assessing Ocular Refraction and Binocularity in an Infant

Patient CD, a nine month old girl was brought to the pediatric clinic on referral from the family physician. She had been a healthy child from birth but at three months of age the parents noticed an intermittent esotropia, that was manifest mainly when the child was fatigued or under stress. The infant had been assessed by an ophthalmologist who prescribed atropine drops over a two week period. This provided no relief of the intermittent esotropia and no further action was taken. The ocular assessment of the child showed the external physical health of each eye to be normal. She responded to an optokinetic nystagmus drum normally. She also passed the Bock candy bead test with either eye with the beads held at 25 centimeters. Fundus evaluation was unremarkable. The strabismus became manifest during the evaluation of binocular function and was approximated to be 30^Δ by observation of the corneal reflexes by the Hirschberg technique. The eyes were aligned on visual axis with 25 prism diopters using a prism bar. There was no paresis or hinderance to vergence or version movements of either eye. The refractive error was determined by retinoscopy both with and without cycloplegia (1% cyclopentolate hydrochloride) as +2.50 D of hyperopia. With the correction in place, the esotropia was not apparent.

The child was referred to the Electrodiagnostic Clinic for an objective assessment of the effect of plus lenses on visual function. The results

of VER testing are illustrated in Figure 7. These records indicated that the VER was recordable but irregular when a checkerboard composed of 14 minutes of arc was used to elicit the VER. When a correction for most of the hyperopia (in "loaner" glasses) was applied bilaterally, the VER improved significantly for the same checkerboard

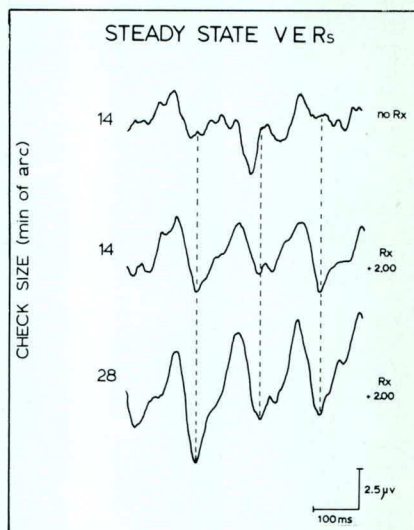


Fig. 7

Steady-state binocular VERs for a 9 month old girl (CB) with an intermittent left esotropia. VERs were elicited by a 12° checkerboard target composed of 14 and 28 minutes of arc black and white checks reversed at a frequency of 8 Hz. Each trace is an average of 50, 500 msec responses. The binocular VER elicited by a checkerboard with 14 minutes of arc checks without a correction for hyperopia (top trace) was recordable but irregular in waveform. This VER waveform was markedly enhanced by the addition of +2.00 DS to each eye (middle trace). Doubling the checkerboard check size increased the amplitude of the VER still further (bottom trace), thereby confirming that the previous response was not spurious and that afferent neural signals to the visual cortex were enhanced by the lenses neutralizing the hyperopia measured by retinoscopy.

target. This observation indicated that the plus lenses did not impair visual resolution, but rather facilitated the visual process. A similar improvement in the VER was obtained when the patient's attention was directed to a checkerboard target composed of 28 minutes of arc checks. This observation indicated the reliability and repeatability of the findings with the prescription for hyperopia in place. In view of the improved cortical activity resulting from the application of corrective lenses, and the greatly decreased

frequency of the esotropia, a prescription to neutralize the hyperopic refractive error in this child was strongly indicated. In a subsequent communication with the parents a few weeks later, they indicated that their child was happily wearing her glasses and the esotropia was eliminated.

Case No. 2: Evaluating the Potential Benefit of Ophthalmic Lenses for Children with Learning Problems

Patient FM, was a seven year old boy whose mother reported him in good health. The teacher in school requested that he be evaluated for vision, hearing, allergy tests and psychological aspects as he was behind his class peers academically. Examination revealed that his visual acuity, ocular motility, accommodative amplitudes and facility, external and internal ocular health were all normal. The refractive error determined by retinoscopy was +1.25 DS, OU. The patient's habitual distance and near phorias were 1^Δ and 2^Δ esophoria respectively. The addition of a +1.00 DS changed the near phoria to 4^Δ of exophoria. Perceptual testing showed the child's responses as being quick, accurate and well within normal limits. There was no indication from the maternal or teacher's history of an abnormal reading posture, or any of the signs and symptoms of accommodative or convergence failure. With the modest hyperopic error which shifted a small esophoria at near, it would be rational to try a lens application and have the parents and teachers observe if classroom behavior was altered in a positive direction. However, with a VER capability, one can be more informed before such a decision is made since visual cortex responses must form the basis for visual perceptions. Therefore FM was referred for an electrodiagnostic evaluation of the necessity for the correction for a modest amount of hyperopia.

The results of the electrodiagnostic assessment by means of the VER are indicated in Figure 8. The VER records obtained monocularly and



THE FAST-ACTION FIZZ!

**NEW, IMPROVED HYDROCARE® PROTEIN REMOVER
TABLETS WITH FIZZY ACTION CLEAN SOFT LENSES FAST.**

**SAFELY REMOVES AND PREVENTS
BOUND PROTEIN, PROLONGS LENS LIFE.**

When protein binds to the lens and denatures, blurred vision and discomfort can result. Used weekly, fizzy Hydrocare Protein Remover Tablets can effectively remove bound protein and prevent its buildup. For your patients, the benefits are clear — greater comfort, extended wearing time, and prolonged lens life.

**COMPATIBLE WITH ALL SOFT
AND GAS PERMEABLE LENSES.**

New fizzy tablets can be used with all soft and gas permeable contact lenses and are compatible with both chemical and heat disinfection systems. Convenient, too, because there's no need for patients to buy unwieldy jugs of distilled water. Tablets dissolve in any saline solution. Weekly protein removal is a necessity and Hydrocare Protein Remover Tablets make it simple.

**HYDROCARE PROTEIN REMOVER TABLETS WITH FIZZY ACTION.
FAST AT WORK PROVIDING CLEAR, COMFORTABLE LENS WEAR.**

binocularly, with and without the correction for hyperopia were very similar in waveform and amplitude. Since the cortical input was not improved when the patient's hyperopia was neutralized, it was concluded that glasses would not be beneficial for this patient.

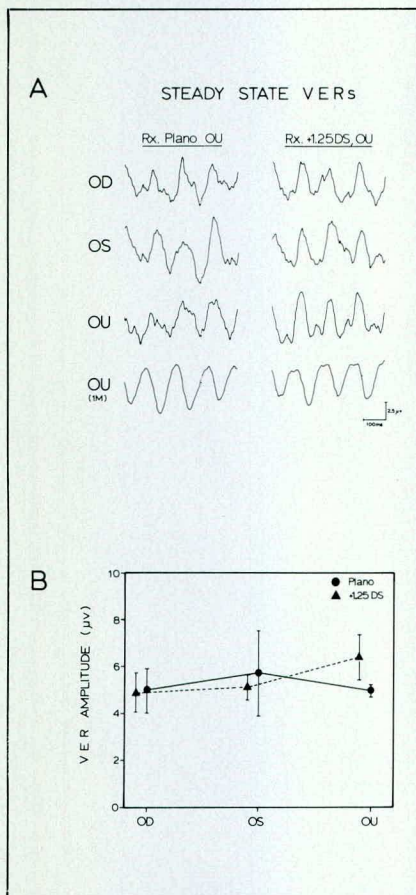


Fig. 8
A) Steady-state VERs for a seven year old boy, FM elicited by a 6° checkerboard target composed of 14 minutes of arc checks reversed at a frequency of 8 Hz. Each record is the average of 50, 500 msec responses. Monocular VERs with or without a correction for low hyperopia (+1.25 DS) were approximately equal in amplitude and had similar waveforms. Binocular VERs appeared marginally sharper and larger when the patient wore a correction for hyperopia. When the VER target was located at a 1 M viewing distance, the binocular VERs were similar in waveform and amplitude with or without the correction for hyperopia.
B) Graph showing the average amplitude the peaks in the steady-state VERs elicited under monocular and binocular conditions, with and without a correction for hyperopia. The vertical bars through each data point are ± 1 SEM for VER amplitudes. This graph illustrates that there was no statistically significant improvement in the VERs when the patient's hyperopia was corrected. Therefore, a prescription for hyperopia was not indicated.

Case No. 3: Assessing the Benefit of Glasses for Low Hyperopia

In direct contrast to the results of the electrodiagnostic assessment obtained for the patient described above, the following case illustrates that visual function was improved in a 7 year old boy (DM) by correction for a modest amount of hyperopia. Patient DM demonstrated good ocular health and good ocular motility with a low esophoric condition at 40 cm. The esophoria (4 prism diopters) was completely neutralized with a +1.0 DS correction for hyperopia in each eye. The results of the VER testing for this patient are shown in Figure 9. Monocular and binocular VERs without a prescription were seen to be of nearly equal amplitude and waveform. However, application of the correction for the modest amount of hyperopia greatly enhanced the binocular VER. In view of the improved cortical response when the patient wore the correction for hyperopia, a prescription was given to the patient to be worn for all near-point visual tasks. A re-examination one month after receiving his glasses indicated that the patient showed a distinct preference for wearing his glasses whenever he was required to do near-point visual work such as reading or writing. This type of purposeful behaviour in this patient who was born with mild cerebral palsy was interpreted as indicating a facilitation of the visual process derived from wearing the glasses.

C. Objective Evaluation of Binocular Function

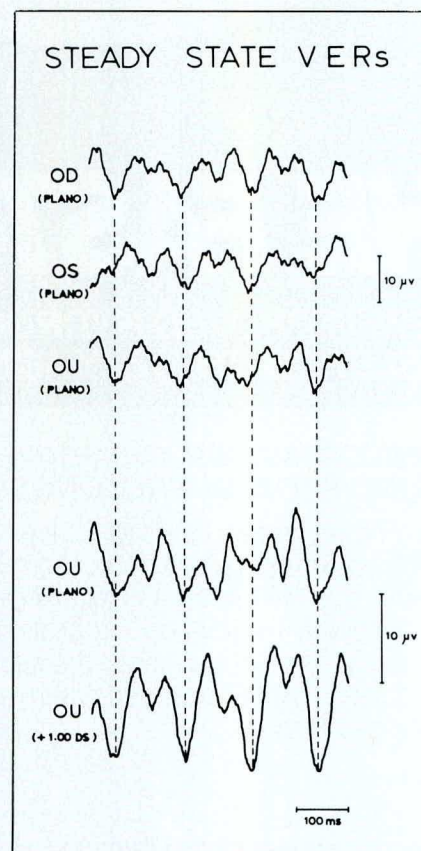
In addition to determining the efficacy of an ophthalmic prescription in improving visual func-

Fig. 9 ▶
 Steady-state VERs elicited from a seven year old boy, DM, born with cerebral palsy. The VER recording parameters were the same as those described in Figure 7A. The VER waveforms were slightly irregular because of poor patient cooperation. No significant differences were noted between the monocular and binocular VERs without glasses for low hyperopia. However, a very significant enhancement in the binocular VER was noted when a correction for hyperopia (+1.00 DS,OU) was applied. In this case, a prescription to neutralize the hyperopic refractive error was strongly indicated.

tion at a neural level, the binocular VER can provide an indication as to the quality of binocular co-ordination.²⁷ The following three cases illustrate various aspects of binocular function that can be evaluated objectively by the binocular VER.

Case No. 1: Evaluation of Binocularity in High Ametropia

A three year old non-communicating girl (BS A) was brought in for an oculovisual assessment largely because of a turned eye. The examination revealed a large angle alternating esotropia that was manifest at all observation distances. Ocular refraction indicated a high hyperopic refractive error in each eye (OD +9.00 -0.75 X 157, OS +8.25 DS). As the patient refused to provide any verbal indication concerning the status of her vision, she was referred to the Electrodiagnostic Clinic to determine whether the correction of the hyperopia detected by retinoscopy would provide an improvement in visual function both in terms of visual resolution and binocularity. The results of the electrodiagnostic assessment are shown in Figure 10. The VER records obtained under



monocular and binocular viewing indicated a very significant enhancement when the correction for hyperopia was worn. However, the VER for the left eye was distinctly smaller than that elicited for the right eye. In addition, as is illustrated in Figure 10B, the peaks in the VERs elicited for each eye were approximately 90° out of phase. Under binocular viewing conditions, the VER closely resembled the VER obtained for the right eye. This neural response suggested that the left eye likely had inferior vision to that of the right eye. Furthermore, since the binocular VER resembled the VER obtained for the right eye, there was a strong

possibility that the left eye was suppressed in binocular viewing conditions. The result of this electrodiagnostic assessment prompted the initiation of amblyopia therapy for the left eye together with a prescription for the entire amount of hyperopia as detected by retinoscopy. A follow-up evaluation of the patient's visual performance after she wore her glasses for approximately a 6 month period revealed improved VERs for the left eye, and a significant reduction in the phase difference in the monocular VERs.

Another follow-up examination 6 months later, revealed that the eyes were almost always straight, and that

the amplitude and waveform of the VER had improved. This was interpreted as indicating continued improvement in visual function which at the time of the second re-examination still could not be verified subjectively. The parent did report, however, that there were no apparent visual or motoric problems with the glasses, and that the patient assumed a normal posture while reading or writing.

Case No. 2: Assessment of Binocularity in the Absence of a Significant Refractive Error

Figure 11A shows the monocular and binocular VERs for a seven year old patient whose oculovisual assessment indicated normal function of all components of the visual system. In this case, the binocular VER waveform illustrates the phenomenon of "binocular facilitation" wherein the binocular VER amplitude is greater than either monocular VER. The occurrence of binocular facilitation usually indicates excellent binocular coordination and normal sensory integration of afferent visual stimuli. In contrast to this example where the binocular VER indicates a high degree of binocular integration, Figure 11B provides two examples (type 1 and type 2) of binocular VERs encountered in patients with various degrees of binocular dysfunction. The "type 1" abnormality in the binocular VER is characterized primarily by the appearance of secondary peaks (see small inverted arrows) unrelated to the checkerboard reversal rate. In addition, there may be a slight phase lag in the binocular VERs when compared to the monocular VERs. The "type 2" abnormality in binocular VERs is highlighted by the absence of a regular pattern under binocular viewing conditions, while under monocular fixation the VERs have both a regular waveform and normal amplitude. The specific binocular anomalies that were eventually detected by standard optometric procedures in these patients illustrating the "type 1" and "type 2" VER pattern were poor ocular coordination for

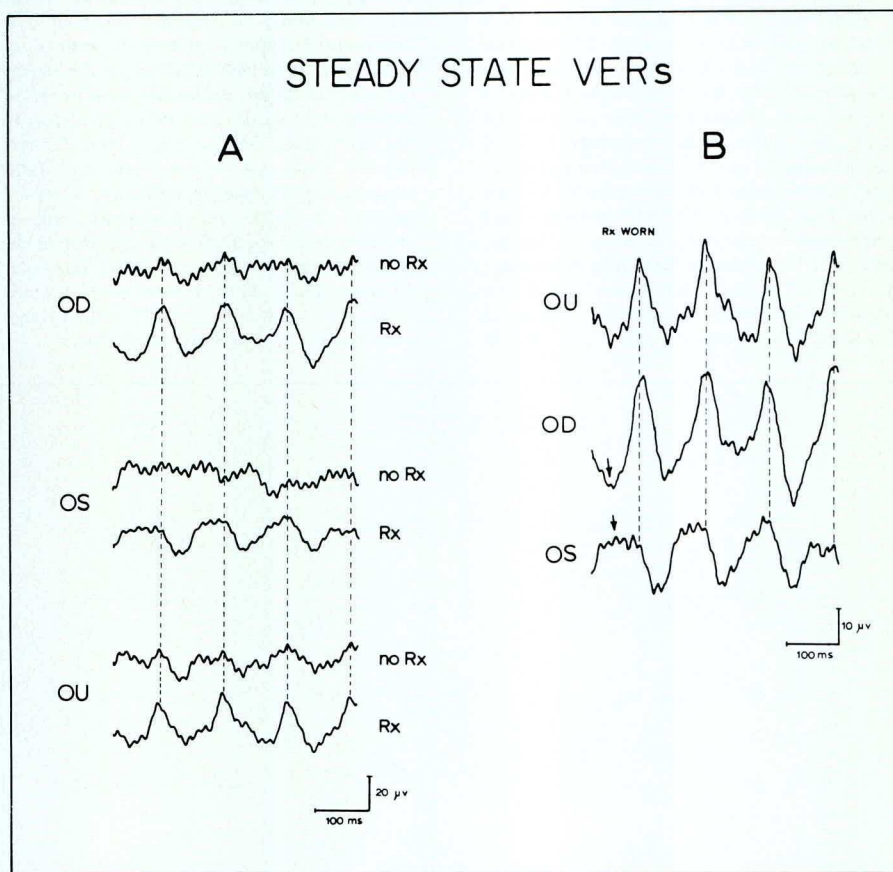


Fig. 10

A. Steady-state VERs for a non-communicating three year old girl with a large hyperopic refractive error and an alternating esotropia of approximately 35Δ at near. VERs were elicited by a 6° checkerboard target with 14 minutes of arc black and white checks reversed at a frequency of 8 Hz. Each trace is an average of 30, 500 msec. responses. An electrodiagnostic evaluation was performed to evaluate the effect of a large correction for hyperopia (OD +9.00 -0.75X157, OS +8.25 DS) on visual resolution and binocularity. The results indicated that the VER waveform and amplitude was greatly enhanced in each eye by the correction for hyperopia. However, the VER for the left eye

was of lower amplitude and had a flatter waveform than the VER for the right eye.

B. Enlargement of the steady-state VERs under monocular and binocular viewing conditions while the patient wore the correction for hyperopia. The vertical broken lines indicated that the monocular VERs were out of phase (see portions of traces identified by small inverted arrows). The binocular VERs were similar in waveform and had the same temporal characteristics as the VER elicited from the right eye. This observation suggested a suppression of the left eye in binocular viewing. A prescription for the hyperopia determined by retinoscopy combined with amblyopia therapy for the left eye was indicated.

versions, vergences and smooth pursuits in one patient, and uncompensated phorias in the second patient. The case examples provided in Figure 11 therefore illustrate that the binocular VER can be used to glean information concerning the degree of binocular coordination in a pediatric population. Inasmuch as these patients were seen in the Electrodiagnostic clinic prior to a general oculovisual assessment, these examples also illustrate that the binocular VER can be used as an objective screening procedure for detecting binocular anomalies. The occurrence of binocular VERs showing a degraded waveform when compared to that obtained for either eye alone indicates the need for a more detailed evaluation of binocular functions.

D. Objective Assessment of Ocular Health

The functional integrity of various components of the visual system can be assessed objectively by means of the pattern VER. The following examples will illustrate how the VER was used to assess neural function at the retinal level, optic nerve, and cortical levels.

Since the pattern VER is an indirect measure of macular function at the retinal level, it can be used to assess the functional integrity of macular-cortical pathways in patients with ophthalmoscopically visible irregularities in the macular areas. Figure 12 presents the steady-state VERs for three siblings of one family elicited by a checkerboard with check sizes varying from 14 minutes of arc to 112 minutes of arc. The sixteen year old boy demonstrated no abnormalities of visual function, normal appearance of the ocular fundus, and normal VER waveforms in response to all check sizes (VERs in the middle column). The VER responses for the 18 year old girl however were completely extinguished for all check sizes. This patient demonstrated fundus signs resembling Stargardt's disease and was classified as legally blind. With the aid of a 6 x 16 monocular telescope

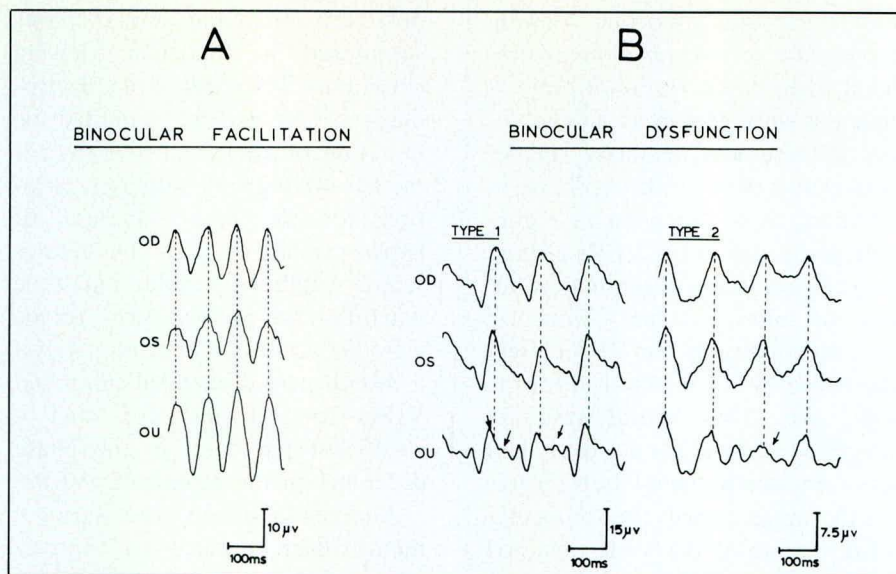


Fig. 11

Steady-state VER responses elicited by a checkerboard target composed of 14 minutes of arc, black and white checks reversed at a frequency of 8 Hz. Each trace is the average of 30, 500 msec. responses. These records illustrate the utility of the binocular VER in determining the quality of binocular coordination. (A) binocular and monocular VERs for a seven year old boy (AZ) with normal visual acuity and binocularity. It is noted that the binocular VER is larger than either monocular VER. This "binocular facilitation" serves as a physiological index of a high degree of binocular coordination. (B) VER records

showing two types of binocular VERs indicating some non-specific binocular dysfunction. A type 1 pattern is seen most frequently. It is characterized by the occurrence of secondary peaks (see small inverted arrows) and a small phase shift with respect to the monocular VERs. These records were derived from an 8 year old boy (RS) complaining of reading difficulty. A type 2 pattern is seen less frequently and is characterized by a marked degradation in the amplitude and regularity of the binocular VER waveform. These records were obtained from a 14 year old boy (JP) with a large uncompensated exophoria at 40 cms.

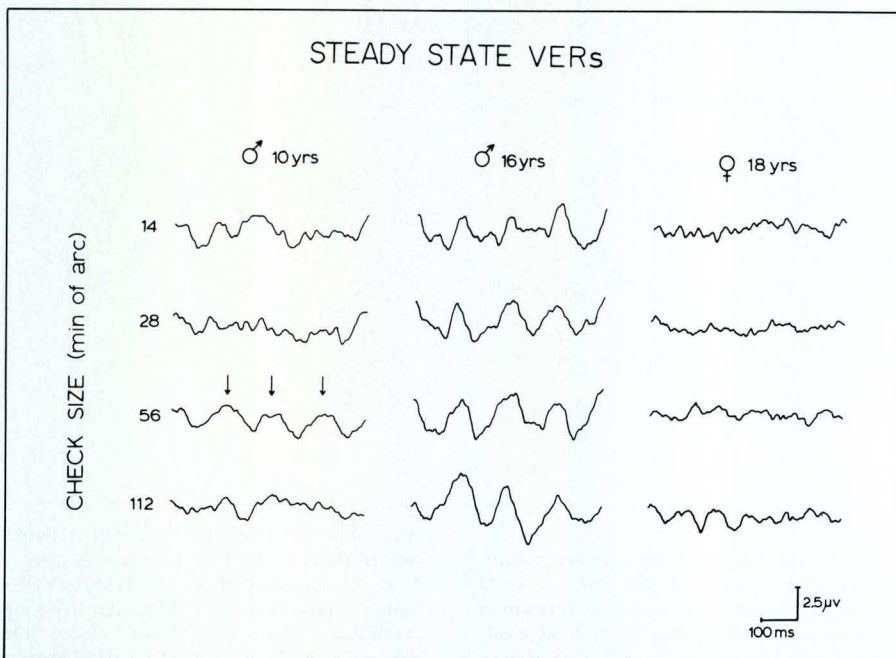


Fig. 12

Steady-state VERs elicited from three siblings using a checkerboard with black and white checks varying from 14 to 112 minutes of arc reversed at a frequency of 8 Hz. The two boys had normal visual acuity (20/20) while the eighteen year old girl had bilateral central scotomas reducing vision to approximately 20/125 in each eye. In her case, the pattern VER was extinguished for all check sizes. The sixteen year old boy demonstrated normal

VER waveforms for all check sizes. The ten year old boy demonstrated abnormal VER responses to all checkerboard check sizes except 56 minutes of arc. The irregularity in the waveforms indicated a significant functional disturbance of vision, even though the measured visual acuity was 20/20 in each eye. Ophthalmoscopically, this young boy demonstrated macular features similar to those seen in his sister's eyes.

Retrobulbar disorders of visual function can be identified by evaluating both the retinal and cortical responses to controlled light stimuli. Figure 13 presents the results of an electrodiagnostic evaluation for a fourteen year old boy who claimed only dim light preception in both eyes. Flicker ERGs and VERs were performed simultaneously for each eye with the results indicating normal flicker ERGs but a total absence of light-induced responses at the cortical level. This indicated grossly

The following case illustrates that the pattern VER can be used to access indirectly various cortical

cortical pathways indicated extinguished flicker VERs for light flashes presented at 20 and 40 Hz. These findings indicated a pathological disorder in retrobulbar components of the visual system subserving form vision. A tentative diagnosis of bilateral optic atrophy was supported by the pale appearance of the optic nerve heads.

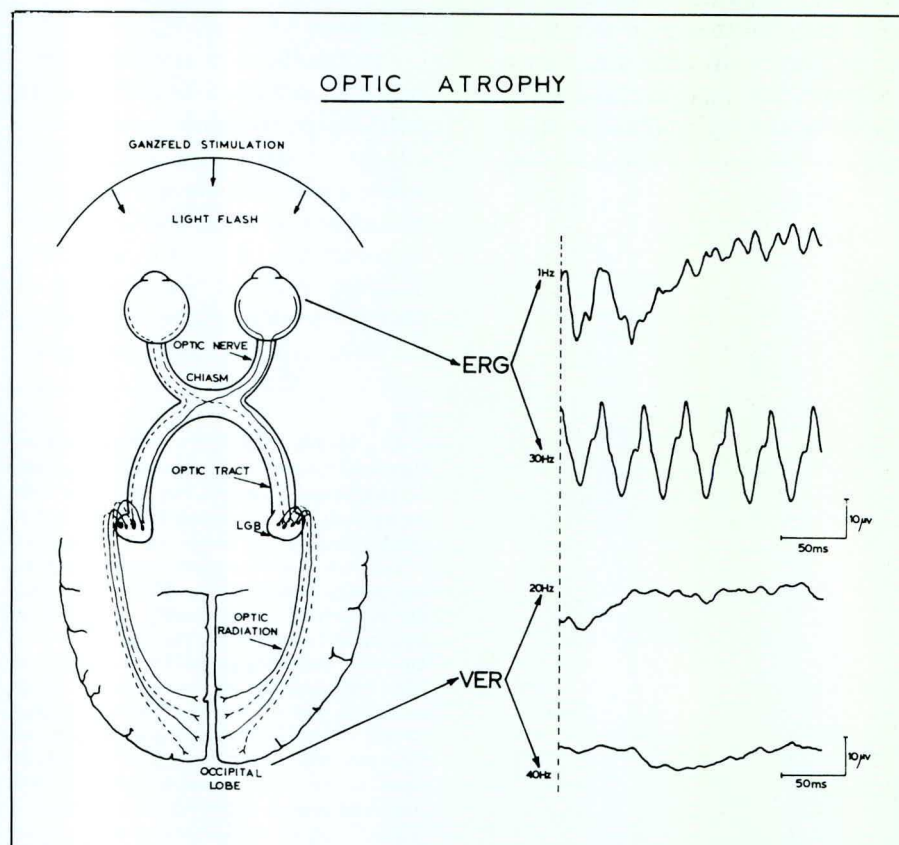


Fig. 13
Figure illustrating how flash ERGs and VEPs were combined to diagnose a bilateral post-retinal lesion in the visual pathways. In the case illustrated here, the fourteen year old boy (DB) claimed only dim perception of light. Single flash and flicker ERGs were normal in each eye. However an assessment of macular

refractive errors in each eye (OD +1.50 DS, OS +1.00 DS). JR could not respond to the Bock candy bead test for an approximation of visual acuity. An assessment of visual fields by confrontation did not reveal any gross abnormalities. The patient gave normal jerk-type nystagmus responses for each eye to the optokinetic nystagmus drum, but the left eye gave a noticeably weaker beat. The general conclusion of the oculo-visual assessment was that JR was developing normally with respect to his vision, and that glasses were not required. However, because the patient was totally non-communicating, it was not possible to quantify in any meaningful way the degree of visual resolution, or provide an explanation for the unusual visual behaviour reported by the mother.

The results of an electrodiagnostic evaluation by means of the patterned VER are illustrated in Figure 14. The VER patterns shown in this figure are of good amplitude and regular waveform for each eye and under binocular viewing conditions. Note-

ably, however, the amplitude of the VER for the left eye was considerably reduced when compared to that for the right eye. This indicated that the visual acuity in the left eye was likely inferior to that for the right eye, even though the refractive error as determined by retinoscopy indicated less hyperopia for that eye. Based on the check size used to elicit the VER, a visual acuity of at least 20/60 was predicted for the right eye with inferior vision in the left eye. Most significantly, the amplitude of the binocular VER was greater than either monocular VERs. This indicated that some level of binocular integration occurred under binocular viewing conditions a function that is compromised in patients with corpus calosum lesions.²⁴ This information concerning binocular function was not obtainable by any other clinical method since the patient was entirely non verbal.

Discussion

The results of a standard optometric examination usually provide sufficient information for the practitioner to define the visual capabilities and limitations of a particular patient and provide remedial therapy if required. There are instances, however, when in the presence of patient-reported vision difficulties, the results of a fundamental oculo-

visual assessment do not identify any abnormalities or allow a definitive diagnosis to be made. Such is the case when the optometrist is confronted with malingerers, noncommunicating patients, or cooperative patients providing paradoxical responses in various visual tests.

The primary objective of this paper is to make the optometric practitioner aware of the technology that is now available to assess the physiological integrity of various portions of the visual system and help identify sub-clinical visual disorders that may explain a patient's symptoms or identify them as psychogenic in origin. The electrodiagnostic techniques described in this paper illustrate their use in providing information concerning a patient's visual acuity, refractive status, binocular function, and ocular health when these functions cannot be assessed readily by the methods employed in a standard eye examination. The results of these tests are of considerable value in arriving at a final diagnosis concerning visual function in a patient, as well as providing prognostic information on specific visual anomalies. It should be emphasized, that these specialized tests neither have an application for every patient, nor are they likely to replace the diagnostic test procedures currently employed in optometric practice. Instead, these neurophysiological tests are intended to supplement the findings of a thorough oculo-visual assessment in aid of a well-founded diagnosis and disposition of clinical cases.

With the passage of time, a greater number of uses for the ERG and VER test procedures described in this paper will likely evolve. The authors have found these techniques extremely useful in providing practical solutions to various visual disorders common to a pediatric population. In addition, these electrodiagnostic techniques have provided new information concerning fundamental aspects of vision physiology and clinical correlates of neurophysiological mechanisms that until now could only be examined in

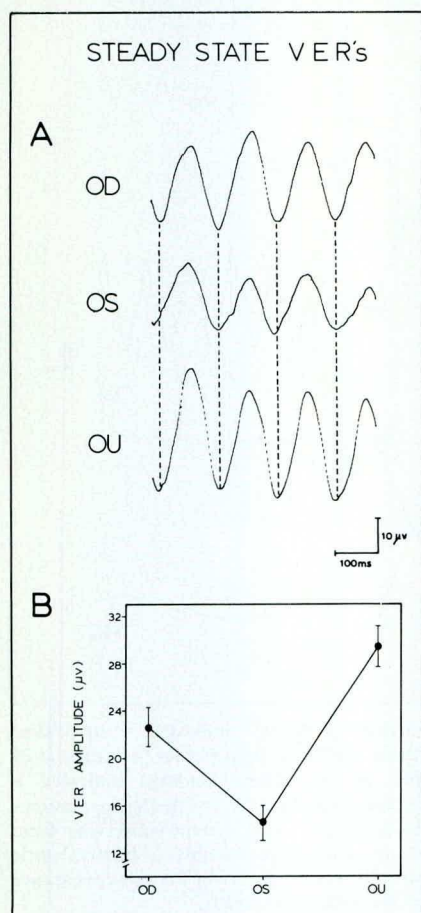


Fig. 14

A) - Steady-state VERs elicited from an eighteen month old, noncommunicating, developmentally delayed, boy (JR) born with an agenesis of the corpus callosum. A 12° checkerboard target composed of 28 minutes of arc black and white checks reversed at 8 Hz was used to elicit the VERs. The VER elicited for the left eye was notably decreased in amplitude compared to the right eye. The binocular cortical response, however, showed some binocular facilitation inasmuch as its amplitude was greater than either monocular VERs. This indicated some degree of binocular coordination, a function which tends to be compromised in patients with complete sections of the corpus callosum.

B) - Graphical presentation of the averaged amplitude of the peaks in the monocular and binocular steady-state VERs recorded for JR. The vertical bars through each data point represent ± 1 SEM for VER amplitudes. This graph emphasizes that the overall cortical response for the left eye was significantly less than that for the right eye, and that the binocular response was greater than either eye alone.

acute animal experiments. The ever increasing pool of knowledge concerning visual mechanisms derived from animal experimentation and clinical studies ultimately provide better diagnostic and treatment procedures for the delivery of vision care services. It is hoped that the information provided in this paper will encourage optometric practitioners to originate, utilize, and develop more fully electrodiagnostic resources accessible to them in the interest of providing a more comprehensive vision care service to their patients.

Acknowledgement

The authors thank the Canadian Optometric Education Trust Fund for its support of a significant portion of the work presented in this paper.

We also gratefully acknowledge the excellent photographic assistance by Robert Kalbfleisch, and the secretarial support of Joan MacLean, Diane Marleau and Gwen Smith.

References

- Woodruff ME: The Visually "At Risk" Child. *J Amer Optom Assoc* 44(2), February, 130-34, 1973.
- Observations on the Visual Acuity of Children During the first five years of life. *Amer J Optom and Arch of Amer Acad* 49(3), 205-213, 1972.
- Woodruff ME: Preventive Vision Care — Emphasizing Detection and Treatment in Early Life. *J Amer Optom Assoc* 46(10), 997-1003, 1975.
- Von Noorden GK: Application of basic research data to clinical amblyopia. *Ophth AAOO* 85: 496-504, 1978.
- Von Noorden GK: New clinical aspects of stimulus deprivation amblyopia. *Amer J Ophthalmol* 92: 416-21, 1981.
- Ingram RM, Traynor MJ, Walker C, Ivison JM: Screening for Refractive Error at Age 1 year - a Pilot Study. *Brit J Ophth* 63: 243-50, 1979.
- Ingram RM, Walker C: Refraction as a means of predicting squint or amblyopia in Preschool Siblings of Children known to have these defects. *Brit. J Ophth* 63: 238-42, 1979.
- Ingram RM, Walker C: Refraction as a basis for screening children for squint amblyopia. *Brit J Ophth* 61: 8-15, 1977.
- Thomas J, Mohindra I, Held R: Strabismic amblyopia in infants. *Amer J Opt Physiol Optics*. 56: 197-201, 1979.
- Woodruff ME: Vision Consultant to Department of Health, Government of New Brunswick, personal communication.
- Woodruff ME: Manitoba Optometrical Association Report on Manitoba Vision Screening Program.
- Woodruff ME: A systems examination of the infant's visual system. *J Amer Opt Assoc* 45(3), 410-415, 1973.
- Gouras Peter: Electroretinography: some basic principles. *Invest Ophthal* 9: 557-64, 1970.
- Norden Lyman: The ERG, concepts and clinical applications. *J Amer Opt Assoc*. 50: (1) 81-87, 1979.
- Chatrian Gean E et al. Computer assisted quantitative electroretinography. 1: A standardized method. *Am J EEG Techno* 20: 57-77, 1980.
- Sokol S, Bloom BH: Macular ERG's elicited by checkerboard pattern stimuli. *Doc Ophthalmol Proceedings Ser* 13: 299, 1977.
- Arden GB, Carter RM, Hogg CR, Powell DJ and Vaegan: Reduced pattern electroretinogram suggest a preganglionic basis for non-treatable human amblyopia. *J Physiol* 82: 308, 1980.
- Fiorentini A, Maffei L, Pirchio M, Spinelli D and Porciatti V: The ERG in response to alternating gratings in patients with diseases of the peripheral visual pathway. *Invest Ophthalmol Vis Sci* 21: 490-3, 1981.
- Sokol S: Visually evoked potentials: Theory, techniques and clinical applications. *Surv Ophthalmol* 21: 18-43, 1976.
- Sokol S, Dobson V: Pattern reversal visually evoked potentials in infants. *Invest Ophthalmol* 15(1): 58-62, 1976.
- Sokol S: Measurement of infant visual acuity from pattern reversal evoked potentials. *Vision Res* 18: 33, 1978.
- Marg E, Freeman DN: Visual acuity development in human infants: Evoked potential measurements. *Invest Ophthalmol* 15: 150-153, 1976.
- Millodot M, Riggs LA: Refraction Determined Electrophysiologically. *Arch Ophthalmol* 84: 272-278, 1970.
- Sherman J. Visual evoked potential (VER): basic concepts and clinical applications. *J Amer Opt Assoc*: 50(1): 19-30, 1979.
- Lovasik JV, Sorbora G: Asynchronous Cortical input and Asthenopia. *Amer J Opt Physiol Optics* 59(7): 573-579, 1982.
- Lovasik JV: An Electrophysiological Investigation of the Macular Photostress Test. *Invest Ophthalmol Vis Sci*. In press, 1983.
- Amigo G, Fiorentini A, Pirchio M, Spinelli D: Binocular Vision tested with visual evoked potentials in children and infants. *Invest Ophthalmol Visual Sci* 17: 910-915, 1978.
- Mitchell DE, Blakemore C: Binocular Depth perception and the corpus callosum. *Vision Res* 10: 49-54, 1970.
- Galloway NR: The Visually-Evoked Potential. *Brit Orthopt J* 36: 1-6, 1979.

Erratum

In the article, "Goodwill — what is it worth in the market?", which appeared in our March issue (Vol. 45, No. 1, Pp 35-39), the following chart was inadvertently omitted. We apologize for the error.

