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Annotations

Visually evoked responses

A number of electrophysiological responses to various sensory stimuli can now be recorded predictably and used as an aid in clinical diagnosis and in the study of different aspects of central nervous system (CNS) function. Prominent among these have been the visually evoked responses which are reviewed in this annotation as they relate to paediatric and paediatric neurological practice.

Background

Some form of electroretinogram (ERG) has been known to exist for over 100 years, with the recognition that a bright flash of light will elicit a distinct electrical response from the human eye. The development of a contact lens electrode in the 1940s brought this earlier observation towards being a predictable and measureable electrophysiological phenomena. In more recent years a greater sophistication of electronic systems has taken this investigation out of the laboratory and into clinical ophthalmology, neurology, and paediatrics. The ERG may provide objective information on several aspects of retinal function—obviously of interest to the ophthalmologist, and in a number of clinical situations of interest to the paediatrician.

Techniques in which a cortical response is recorded with scalp electrodes in response to a number of sensory stimuli have been developed during the last two decades. The VER or visually evoked potential is the cortical response originating primarily from the cones of the central retina in response to light stimulation. The VER is recorded with electrodes fixed to the skull close to the external occipital protuberance. With greater sophistication of electronic equipment and particularly the development of computer 'averaging' facilities the VER can be recorded relatively easily and employed in a number of clinical situations.

Characteristics of the ERG and VER

The ERG has two major and a number of minor components. The first major component is a negative downgoing 'a' wave and the second a larger positive upgoing 'b' wave. The ERG reflects rod function principally, since rods outnumber cones in a ratio of approximately 20:1. Nevertheless, by varying the state of light and dark adaptation of the subject's eyes and the intensity and frequency of light stimulation it is possible to record an ERG that reflects to a greater extent rod or cone function.

It is difficult to ascribe exactly the various components of the ERG to particular retinal layers, although it has long been established that the ERG originates from the retina and specifically from the superficial or outer layers.[3] It is possible to state that the principal responses reflect the activities of the bipolar cells, the photoreceptor cells (rods and cones), and the pigment epitheelium; but not the ganglion cells or the nerve fibres that constitute the inner retinal layers.

The VER has three principal components—one initial positive upgoing peak, a negative peak, and a second positive peak at approximately 150–200 ms. It is because the cones of the central retina, including the macula, have an almost one to one relation to the corresponding bipolar and ganglion cells and project onto about half the visual cortex that the VER recorded from electrodes over the occiput reflects central retinal function. Abnormalities of the VER may occur, however, as a result of a lesion of either the ganglion cell layers of the retina, the optic pathways, or the visual cortex.

Recording techniques

The recording of the VER may be undertaken in an EEG department equipped with apparatus appropriate for providing visual stimuli and recording responses. Some form of 'averaging' facility is essential. VER may also be performed using adapted EMG equipment preferably, but not essentially, in a room set aside for the purpose.

In many centres the ERG is still recorded using the contact lens electrode which can be inserted after corneal anaesthesia. With most children, however, the necessary cooperation is lacking and a general anaesthetic is given. The more recent development of a gold foil electrode that is placed on the lower eyelid may be satisfactory for many

recording purposes: it is easier to insert than the contact lens electrode, but can still be difficult to place and again a certain amount of cooperation and tolerance is required on the part of the subject. Perhaps of greatest interest for the successful recording of the ERG in children is the observation made by Harden that the ERG can be recorded from an electrode placed between the eyes on the bridge of the nose.[1] The amplitudes of the recordings are small and 'averaging' techniques are needed to distinguish them from the background activity. Although this technique of recording may lack some of the finer details sought from ophthalmologists in the diagnosis of retinal disease, when carried out in conjunction with the recording of the VER and EEG, valuable information on visual and CNS function can be obtained.[2] Furthermore, sedation is not required.

The visual stimulus for the VER can be either a bright flash of light in front of the eye as employed for the ERG, or any of a number of patterned visual stimuli—particularly the checkerboard pattern displayed on the screen of a television monitor. With this latter stimulus the child fixes on the central spot on the screen and the checkerboard squares reverse from black to white and back again at a selected alteration rate. The screen maintains a constant average luminace on the retina.[3] The patterned stimulus is certainly the 'ideal' in that more consistent wave forms are obtained[4] but concentration for fixation by the young or uncooperative child may not be possible. By using flash stimuli it is possible to record simultaneously not only the VER but the ERG.[1] With time being allowed for fixation of the electrodes these combined recordings can be undertaken within an hour.

Clinical applications

Recording of the VER may often be of diagnostic value in paediatrics. As with most (if not all) electrophysiological measurements the findings should be viewed as clinical signs, to be evaluated alongside other clinical phenomena. But it is equally important to realise that significant abnormalities may be present in the ERG before there are ophthalmoscopical changes or any clinically obvious deterioration of visual function. Conversely the VER (and also sometimes the ERG) may still be retained, although they will seldom be normal where there is clinically obvious visual impairment. It is generally preferable to attempt to record both the ERG and the VER at the same examination and often at the same time as the EEG.

Abnormalities, including the absence of the ERG will be found even in the preclinical stages of disorders in which deterioration of the superficial layers of the retina is a feature; particularly retinitis pigmentosa. In Leber's amaurosis (congenital retnal blindness, not to be confused with Leber's optic atrophy) the ERG will be either very small or absent even before there are any changes seen in the fundi. ERG abnormalities may serve as an aid to diagnosis in conditions in which there is an associated deterioration in the superficial layers of the retina such as Lawrence-Moon-Biedl syndrome; in some forms of autosomal dominant progressive ataxia, Freidrich's ataxia, Refsum's disease; α and β lipoproteinaemia; and the different forms of childhood onset Batten's disease (neuronal ceroid lipofuscinosis.)

Special mention should be made of the group of neurodegenerative disorders still referred to as the amaurotic familial idiocies. In both the Tay Sachs disease and the infantile Neimann-Pick disease the abnormal storage of material takes place in the ganglion cell layers of the retina, and for this reason abnormalities may be found in the VER but not in the ERG in the early stages of the disease. The opposite will be true in childhood onset Batten's disease which affects the superficial retinal layers. In the late infantile variety of Batten's disease, however, (Jansky-Bielchowsky disease), Pampiglione and Harden have noted not only the abnormalities in the ERG but also that the amplitude of the VER is greatly increased in parallel with similar findings in the occipital leads of the EEG after slow photic stimulation.[5]

Abnormalities of the VER will occur as a result of any lesion from the ganglion cell of the retina to the occipital cortex. Included may be some of the neurodegenerative disorders referred to above, demyelinating lesions of any sort,[3] and compressive lesions of or damage to the optic pathways. With so called cortical blindness, abnormalities of the VER are usual but not invariable, while with hysterical blindness normality is to be expected. Refractive errors will produce minor changes in the VER and studies have been undertaken in infants and young children to determine visual acuity using different checkerboard stimulations.[4]

Future developments

There are a number of clinical situations in which the recording of the VER may be of assistance in diagnosis and evaluation. While greater simplification of the recording apparatus and techniques has made this form of non-invasive study more useful in paediatric practice, we are still really only at the beginning. There is an urgent need to increase experience in this field of study and produce greater

standardisation of recording techniques, and the potential within the field of neurodevelopmental paediatrics has yet to be realised. The VER (in the same way as brain stem auditory response and the somatosensory responses) reflects CNS maturity[6] and integrity[7] and should be considered as a form of measurement that can be employed in the much needed follow up studies of the at risk neonate and infant.

References

[1] Harden A. Non-corneal electroretinogram. Parameters in normal children. *Br J Ophthalmol* 1974;**58**:811–6.

[2] Harden A, Pampiglione G. Visually evoked potential, electroretinogram and electroencephalogram studies in progressive neurometabolic 'storage' disease of childhood. In: Desmedt JE, ed. *Visual evoked potentials in Man: new developments*. Oxford: Clarendon Press, 1977:470–80.

[3] Halliday AM, McDonald WI, Mushin J. Visually evoked responses in the diagnosis of multiple sclerosis. *Br Med J* 1973;**iv**:661–4.

[4] Sokol S. Measurement of infant visual acuity from pattern reversal evoked potentials. *Vision Res* 1978;**18**:33–9.

[5] Pampiglione G, Harden A. Neurophysiological identification of a late infantile form of 'neuronal lipidosis'. *J Neurol Neurosurg Psychiatry* 1973;**36**:68–74.

[6] Harden A. Maturation of the visual evoked potentials. In: Chiarenza GA, Papakostopoulos D, eds. *Clinical application of cerebral evoked potentials in paediatric medicine*. (International congress series no 595). Amsterdam: Excerpta Medica, 1982:41–59.

[7] Marcus MM. Visual evoked potentials to flash and pattern in normal and high risk infants. In Desmedt JE, ed. *Visual evoked potentials in Man: new developments*. Oxford: Clarendon Press, 1977:490–9.

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Further reading

Galloway NR. *Ophthalmic electrodiagnosis*. 2nd ed. (Vol 1 in series: *Major problems in ophthalmology*, Trevor-Roper PD, ed.) London: Lloyd-Luke Medical Books, 1981.

Carr RE, Siegel IM. *Visual electrodiagnosis testing. A practical guide for the clinician*. Baltimore and London: Williams and Wilkins, 1982.

Halliday AM, ed. *Evoked potentials in clinical testing*. (Vol 3 in series: *Clinical neurology and neurosurgery monographs)*. London and New York: Churchill Livingstone, 1982.