Neurological applications of surface-recorded electrocochleography

A. R. D. THORNTON AND C. H. HAWKES

*From the Audiology Group, Institute of Sound and Vibration Research,
University of Southampton, and the Wessex Neurological Centre,
Southampton General Hospital, Southampton*

SYNOPSIS A new outpatient technique, involving the recording of auditory-evoked responses from scalp electrodes, is described. Responses are obtained from the cochlea, cochlear nerve, and auditory brain stem nuclei. The effects of various disorders on these responses are illustrated and the procedure is assessed as a diagnostic technique.

Electrocochleography is a technique that involves the recording of small amplitude ($<0.5 \mu\text{V}$), short latency ($<10 \text{ ms}$) auditory evoked responses using time-domain averaging techniques to extract these small responses from the larger amplitude electrical activity caused by the ongoing electroencephalogram (EEG) and spontaneous myogenic potentials. The method most generally used is the transtympanic approach described by Ruben (1967) and by Portmann and Aran (1971). This involves passing a needle electrode through the tympanum to make contact with the bony promontory. Two responses may be obtained from this recording site. The first is the end-organ potential or cochlear microphonic (CM). This is a

sensory potential, generated within the cochlea, which follows the time-history of the stimulus pressure wave. The second response is the cochlear nerve gross action potential (N_1).

In 1967 Sohmer and Feinmesser reported the recording of the cochlear nerve response and series of later responses using disc electrodes placed on the skin surface. Figure 1 shows the set of five neurogenic responses (N_1 to N_5) recorded from a normal subject. Lev and Sohmer (1972) investigated the generating sites of these responses in the cat by a comparison of recordings obtained from intracranial and from surface electrodes. They concluded that the N_1 response is generated at the cochlear nerve, N_2 at the cochlear nucleus, N_3 at the superior olive, and

FIG. 1 Cochlear nerve and brain stem evoked responses from a normal subject. The trailing edge of the inset pulse marks the time of acoustic stimulation. Normal responses: X axis— 1.5 ms/div . Y axis— 32 nV/div .

N₄ and N₅ at the inferior colliculus. Similar response complexes, recorded from humans, have been described by Coles and Thornton (1973), Terkildsen *et al.* (1973), and by Picton *et al.* (1974). The possible application of these measures in the assessment of peripheral and central neurological disorders has been described earlier (Thornton, 1975a). Here the neurological, radiological, and audiological findings are compared with electrocochleographic results in order to evaluate the use of this technique. Four selected cases will be used to illustrate the findings.

METHOD

The details of electrode derivation, test facilities, and experimental technique have been described earlier (Thornton, 1975b). Briefly, the patient lies on a bed in an electromagnetically screened anechoic room. Silver-chlorided EEG disc electrodes are used and the signals from the active electrodes at each mastoid process are amplified, passed to the adjacent control room, filtered, and fed to the analogue-to-digital converters of a PDP-12 computer. The computer carries out the averaging of 2000 stimulus presentations in two channels with a 30 ms window. The averaged data are stored on digital magnetic tape for subsequent retrieval and analysis. Click stimuli are presented via earphones or loudspeakers at a rate of 10/second and the test session lasts for about 30 minutes. The patients are not sedated but are instructed to relax as much as possible.

NORMATIVE DATA Normative data were obtained from six normal subjects who were tested four times at each of four stimulus levels. The response measurement parameters have been studied in detail (Thornton, 1975b) and it was concluded that, for clinical purposes, the data are best represented by the peak-to-peak amplitude, taken from a negative peak to the succeeding positive peak, and by the latencies of the negative peaks. The statistical properties of these response measures have also been evaluated by Thornton (1975c), who showed that the normal values at each stimulus level may be expressed as a graph of response amplitude by latency as shown in Fig. 2. Each ellipse corresponds to one of the responses N₁ to N₅ and is centred on the mean values of amplitude and latency obtained from the normal subjects for that response. The ellipse itself represents an equal probability contour that corresponds to one standard deviation of latency on the horizontal axis and one standard deviation of amplitude on the vertical axis. Such a presentation provides a convenient format in which to assess the values obtained from patients and to detect abnormal changes in latency and in amplitude. Approximately 90% of the results from normal subjects should lie within 1½ standard deviations from the mean value.

It is realized that this small number of subjects will not provide definitive norms and, when the final details of the optimum recording parameters have been determined, larger numbers will be tested. However, these data do provide a preliminary diagnostic baseline and allow an initial evaluation of this technique to be made.

FIG. 2 Normal values of amplitude and latency shown as standard deviation ellipses for 80 dB sensation level.

RESULTS

CASE 1 DIAGNOSIS: CONGENITAL END-ORGAN DEAFNESS A nine year old girl presented because of longstanding right-sided deafness and a six month history of rotary vertigo. Clinical examination revealed a severe perceptive deafness in the right ear but no other abnormality. Audiometrically there was no measurable hearing in the affected ear.

Electronystamography showed a first degree nystagmus to the left and caloric testing revealed absent vestibular function on the right side. A diagnosis of right-sided end-organ dysfunction was made. The cochlear microphonic results, shown in Fig. 3, support this viewpoint. Binaural stimulation was used and the CM from the left side shows a reasonable reproduction of the stimulus waveform, whereas no similar response is visible in the record obtained simultaneously from the right side.

FIG. 3 Cochlear microphonic (CM) responses from the patient with end-organ hearing loss (AG2806). Trace A: CM from the right affected side. Trace B: CM simultaneously recorded from the left unaffected side. Trace C: Pressure waveform of the acoustic stimulus. Y axis: 20 nV/div. X axis: 58 μ s/div.

CASE 2 DIAGNOSIS: ACOUSTIC NEUROMA A fifty-four year old lady was admitted with a two month history of progressive headache, ataxia, dysarthria, and right-sided deafness. Neurological examination revealed papilloedema, nystagmus, severe perceptive deafness on the right side, and truncal ataxia. A

ventriculogram showed symmetrical enlargement of the lateral and third ventricles; the aqueduct and the fourth ventricle were displaced to the left and there was a slight forward shift of the aqueduct. At operation a typical acoustic neuroma was found on the right side, expanding backwards into the cerebellar hemisphere.

Electrocochleography was performed before operation and the cochlear microphonic results are shown in Fig. 4. There are no appreciable differences

FIG. 4 Cochlear microphonic responses obtained simultaneously from both sides from the patient with an acoustic neurofibroma. X axis: 90 μ s/div. Y axis: 39 nV/div.

in the amplitude or in the waveshape between the simultaneously recorded responses from the left and right sides. This implies that there remain some functional hair-cells and that the vascular supply to the cochlea is intact. The neurogenic responses, obtained with binaural stimulation and simultaneous recording from left and right sides, are shown in Fig. 5. The responses on the left side are all within normal limits, as are the responses N₂ to N₅ (cochlear nucleus to inferior colliculus) from the right. The right side N₁ (cochlear nerve) response has a significantly smaller amplitude than normal.

Thus, the cochlear microphonic measurements indicate normal end-organ function and the brain stem responses give normal values, leaving only the abnormal cochlear nerve response, which implies a lesion at this level.

CASE 3 DIAGNOSIS: PONTINE GLIOMA A seven year old boy was admitted with a six week history of

FIG. 5 Cochlear nerve and brain stem response values from each side from the patient with a right side acoustic neurofibroma. Binaural stimulation was used. Responses from mastoid: left ●, right ○.

vomiting and morning headaches. Initial examination revealed left cerebellar signs and right hemiparesis followed three weeks later by a left V and VII cranial nerve palsy. Audiometrically, pure tone thresholds were normal on both sides. There was no apparent difficulty in understanding conversational speech but simultaneous binaural loudness balance tests showed some abnormality at high intensity levels. A left pontine mass was demonstrated at ventriculography. A suboccipital craniectomy was performed which revealed a tumour lifting up and displacing backwards the left side of the floor of the fourth ventricle, which was markedly distended. There was also a massive expansion of the brain stem on the left, with a large excrecence from the

region of the olive. The latter area was biopsied and found to contain a protoplasmic astrocytoma of the pons.

The electrocochleographic results obtained from binaural stimulation are shown in Fig. 6. The responses from the right are all within normal limits. The left side N_2 , N_3 , and N_4 responses show reduced amplitude, whereas the N_1 and N_5 response amplitudes are normal. These results indicate that there is a disorder on the left which has affected the cochlear nucleus, the superior olivary complex, and, possibly by interference with the lateral lemniscal tract, the first of the responses from the inferior colliculus. These findings are in close agreement with the operative evidence of the site of the glioma

FIG. 6 Cochlear nerve and brain stem response values from each side from patient (R.W.) with a left pontine glioma. Binaural stimulation was used. Responses from mastoid: left ●, right ○.

CASE 4 DIAGNOSIS: MULTIPLE SCLEROSIS A fifty-four year old lady was admitted with a 20 year history of multiple sclerosis. This commenced with numbness of the left arm which resolved but was followed five years afterwards by numbness below the waist and weakness of the legs. This last attack never resolved and subsequently she became paraplegic with urinary retention. Examination revealed bilateral optic atrophy, nystagmus, and spastic quadriparesis with cerebellar signs in the upper limbs.

Audiometric testing showed normal pure tone thresholds and good speech discrimination. Alternate and simultaneous binaural loudness balance tests gave normal results.

The electrocochleographic results recorded from each side were very similar and the record obtained from the left side with monaural stimulation of the left ear is shown in Fig. 7. The N_1 response is within normal limits but the N_2 , N_3 , and N_4 responses, while having normal amplitude, show a marked increase in latency. The N_5 response shows a decrease in amplitude as well as an increase in latency.

DISCUSSION

The results presented here are preliminary but represent findings typical of the cases studied to date. The clinical value of electrocochleography depends on a number of technical and interpretative factors.

TECHNICAL ASPECTS The present technique involves the far-field recording of extremely small responses by extensive time-domain averaging

in which response characterization and measurement are greatly aided by computer methods (Thornton, 1975a). With careful technique repeatable results have been obtained for normal subjects (Fig. 2). More data are necessary to provide more definitive normal values and to investigate variation with age and other factors.

Theoretically, it is to be expected that the evoked response from a brain stem nucleus on one side would propagate to the recording electrodes on both sides. However, additional studies on normal subjects (Thornton, 1975d) and results from patients with known lesions suggest that the responses recorded from one side are due predominantly to the ipsilateral nuclei.

It might then be expected that a lesion that reduced one response—say, N_2 —would lead to a reduction of the subsequent responses (N_3 to N_5) on that side. When binaural stimulation is used in such a case this does not occur, presumably because of the bilateral representation of the brain stem auditory pathways. For example, in Fig. 6 the N_5 response is preserved on the left, because it has been activated by the right auditory pathway. When this patient was tested using monaural stimulation of the left ear the responses N_2 to N_5 showed diminished amplitudes.

LOCALIZATION OF LESIONS The presumed origin of the five responses is based on the work of Lev

FIG. 7 Cochlear nerve and brain stem response values from patient (J.H.) with multiple sclerosis.

and Sohmer (1972) in cats. The feline and human brain stems are structurally similar and data obtained so far indicate lesion sites which correlate well with the known or suspected pathology, especially in the case of compressive lesions (Figs 5 and 6).

On occasion, it is very difficult, using conventional techniques, to distinguish end-organ from cochlear nerve deafness. In cases of total hearing loss there are few if any applicable standard tests, and in cases of subtotal loss the phenomenon of recruitment, for example, is not exclusive to end-organ lesions (Priede and Coles, 1974). The impaired cochlear microphonic response (case 1) and diminished N_1 response with normal cochlear microphonics (case 2) in the absence of any other abnormality unequivocally indicate the site of the disorder in each instance.

NATURE OF LESION In patients with compressive or vascular disease of the brain stem, the characteristic electrocochleographic finding has been a reduction of response amplitude at the appropriate neurological level (Fig. 6), while the latency has been altered little. In patients with multiple sclerosis involving the brain stem (Fig. 7) the characteristic change has been a diffuse increase in latency with a variable degree of amplitude reduction. It is well recognized that, in demyelinating peripheral neuropathy, a slowing of conduction velocity and reduced amplitude may be found (Gilliat and Sears, 1958) and it is of particular interest that demyelination of central pathways should produce a similar alteration of conduction. Comparable observations have been made by Halliday *et al.* (1973) with visual evoked responses and by Robinson and Rudge (1975) with auditory evoked responses. Clearly, the presence of marked slowing in the electrocochleographic response of a patient with suspected multiple sclerosis but no brain stem signs would be a valuable diagnostic aid. Furthermore, the latency measures could be used to monitor objectively the response to treatment and to compare treatments.

CONCLUSIONS Several tests, such as loudness balance, tone decay, short increment sensitivity index, etc., are used to distinguish lesions of the

cochlea from disturbances of its nerve but such tests can give equivocal results. Conversely, there are few, if any, dependable tests which probe the brain stem auditory pathways. The electrocochleographic examples given here, together with additional information about patients, indicate that this technique can offer both otologists and neurologists a non-invasive outpatient procedure which to date has yielded reliable information about the site and nature of auditory brain stem disorders.

This work was supported by the Medical Research Council, grant number 970/512/C.

REFERENCES

- Coles, R. R. A., and Thornton, A. R. D. (1973). Audiology research in the I.S.V.R.'s first decade. *Journal of Sound and Vibration*, **28**, 313–332.
- Gilliat, R. W., and Sears, T. A. (1958). Sensory nerve action potentials in patients with peripheral nerve lesions. *Journal of Neurology, Neurosurgery, and Psychiatry*, **21**, 109–118.
- Halliday, A. M., McDonald, W. I., and Mushin, J. (1973). Visual evoked response in diagnosis of multiple sclerosis. *British Medical Journal*, **173**, 661–664.
- Lev, A., and Sohmer, H. (1972). Sources of averaged neural responses recorded in animal and human subjects during cochlear audiometry (electro cochleography). *Archiv für klinische und experimentelle Ohren-Nasen-und Kehlkopfheilkunde*, **201**, 79–90.
- Picton, T. W., Hillard, S. A., Krausz, H. I., and Galambos, R. (1974). Human auditory evoked potentials. I. Evaluation of components. *Electroencephalography and Clinical Neurophysiology*, **36**, 179–190.
- Priede, V. M., and Coles, R. R. A. (1974). Interpretation of loudness recruitment tests—some concepts and criteria. *Journal of Laryngology and Otolaryngology*, **88**, 641–662.
- Portmann, M., and Aran, J.-M. (1971). Electrocochleography. *Laryngoscope*, **81**, 899–910.
- Robinson, K., and Rudge, P. (1975). Auditory evoked responses in multiple sclerosis. *Lancet*, **1**, 1164–1169.
- Ruben, R. J. (1967). Cochlear potentials as a diagnostic test in deafness. In *Sensorineural Hearing Processes and Disorders*, pp. 313–337. Edited by A. B. Graham. Little-Brown: Boston.
- Sohmer, H., and Feinmesser, M. (1967). Cochlear action potentials recorded from the external ear in man. *Annals of Otolaryngology, Rhinology and Laryngology*, **76**, 427–435.
- Terkildsen, K., Osterhammel, P., and Huis in't Veld, F. (1973). Electrocochleography with a far field technique. *Scandinavian Audiology*, **2**, 141–148.

- Thornton, A. R. D. (1975a). The diagnostic potential of surface-recorded electrocochleography. *British Journal of Audiology*, **9**, 7–13.
- Thornton, A. R. D. (1975b). The measurement of surface-recorded electrocochleographic responses. *Scandinavian Audiology*, **4**, 51–58.
- Thornton, A. R. D. (1975c). Statistical properties of surface-recorded electrocochleographic responses. *Scandinavian Audiology*, **4**, 91–102.
- Thornton, A. R. D. (1975d). Bilaterally recorded early acoustic responses. *Scandinavian Audiology*, **4**, 173–181.