Electronystagmographic criteria in neuro-otological diagnosis

1 Peripheral lesions

STAVROS KORRES

From the Medical Research Council Hearing and Balance Unit, The National Hospital, Queen Square, London

SUMMARY Direct current electronystagmographic recordings of spontaneous nystagmus, in the light, in darkness, and with eye closure were carried out on 34 patients with peripheral lesions of the vestibular system with a view to determining their specificity. In 60% of patients the responses were characterised by an enhancement of the nystagmus both with eye closure and in darkness. The remainder exhibited no nystagmus under any recording condition.

Despite the fact that the electronystagmographic examination of disordered eye movements of one kind or another is being increasingly promoted as a diagnostic tool in neuro-otology, review of the literature shows that there exists as yet no consensus of opinion concerning either the method of administration of the test procedure or the diagnostic criteria which may be applied to the interpretation of the results.

Thus, for example, Aschan *et al.* (1956) maintain that when recording is carried out with eyes closed spontaneous nystagmus is revealed in 40% of patients with Ménière's disease and most patients with vestibular neuronitis, while Pastor and Gaona (1974) report spontaneous nystagmus to be present with eyes closed in 78% of their patients with Ménière's disease. By contrast, Jongkees *et al.* (1962) found spontaneous or positional nystagmus during eye closure in all of 54 patients suffering from Ménière's disease but in only one of 85 normal subjects. Jongkees and Philipszoon (1964) make the additional point that horizontal nystagmus is always pathological in origin when present in more than two positions of gaze.

Of more immediate concern to the subject of electronystagmography are the striking changes a number of authors (Jung and Kornhuber, 1964; Hood, 1968; Rubin, 1968; Belkahlia *et al.* 1970; Padovan *et al.*, 1975) have commented upon in the

character of the nystagmus when optic fixation is removed either by closing the eyes or recording with eyes open in darkness. Perhaps the most serviceable approach to the establishment of diagnostic criteria is to be found in Hood's studies of spontaneous nystagmus resulting from lesions at various levels of the nervous system (Hood, 1968). On the basis of certain characteristic changes occurring as the result of the removal of optic fixation either by eye closure or recording in darkness, Hood claimed to be able to recognise three distinct patterns of response associated with (a) peripheral lesions, (b) vestibular nuclear lesions, and (c) high brain stem lesions. Thus in peripheral lesions the nystagmus is enhanced if present, or made manifest if not, by both eye closure and darkness; in vestibular nuclear lesions eye closure abolishes the nystagmus although it persists in an identifiable modified form in darkness; in high brain stem lesions the nystagmus is abolished by both eye closure and darkness. Hood, however, provided no indication of the generality of his findings. A study has, therefore, been carried out on three selected groups of patients to determine the specificity of the respective electronystagmographic responses to each, particularly in respect of their reliability in terms of localisation. The present paper is concerned with the findings in the first group.

Method

The technique adopted closely followed that described by Hood. The subject sat in an erect posi-

Address for reprint requests: Dr J. D. Hood, MRC Hearing and Balance Unit, Institute of Neurology, National Hospital, Queen Square, London WC1N 3BG, England.

Accepted 27 September 1977

tion with his chin in a rigidly mounted chin rest, head movement being restrained by means of a hinged support brought forward onto the occiput. In this position he was asked to direct his gaze to a fixation mark carried by a vertical bar to his front which could be moved laterally by the examiner seated behind him over an angle of 30° to left and right. Recordings were carried out with direct current amplification using an improved form of electrode of high stability together with a form of electrode attachment which isolated the electrodes from any mechanical movement which might otherwise have given rise to spurious potentials.

Both direction and magnitude of vestibular spontaneous nystagmus depend, in accordance with Alexander's law, upon gaze direction. In practice, therefore, nystagmus was examined not only in the primary position but also with gaze deviated 30° to left and right. In this way the complete pattern of the nystagmic response is fully specified on the recording. Apart from the eye-recording pen two additional pens completed the assembly. The first was stationary and provided a baseline made to correspond with the primary position of gaze. The second pen, operated by a potentiometer connected to the bar deviating the fixation mark, provided a recording of the fixation mark superimposed upon that of the eyes, upwards to the right, downwards to the left. Since its excursions were fixed at 10 mm for every 10° of movement, adjustment of the gain of the eye-recording pen to bring the two traces into coincidence served additionally as the calibration.

The fixation mark tracing was of particular importance when recordings were carried out in darkness or with eyes closed. For this purpose the patient was asked to direct his gaze in the light to the fixation mark, and then, with eyes closed or in darkness, to continue to endeavour to maintain the same gaze deviation. In this way the effect of the removal of optic fixation could be directly referred to the nystagmus associated with any particular gaze deviation. The presence of the fixation mark tracing also enabled pathological departures from the intended gaze direction to be determined with some accuracy.

Apart from the spontaneous nystagmus, recordings were also carried out of following movements elicited by smooth lateral movement of the fixation mark and also of the optokinetic responses.

Results

The results of our studies on 34 patients with peripheral lesions of the vestibular system are considered here. In this context the peripheral

system is defined as the labyrinth and nerve fibres up to the vestibular nuclei, but excluding the central involvement of any space-occupying lesion in the cerebellopontine angle. The 34 patients consisted of 16 with Ménière's disease, 11 with vestibular neuronitis, and seven with other disorders of the peripheral vestibular system. Of these, 20 patients presented with a characteristic pattern of spontaneous nystagmus conforming to that described by Hood and exemplified by the typical tracings from one of the patients with Ménière's disease shown in Figs. 1, 2, and 3. The remainder had complete absence of spontaneous nystagmus in any eye position with or without optic fixation. The results are summarised separately in respect of the three groups in Tables 1, 2, and 3.

The localising value of spontaneous nystagmus was considered in respect of its direction and the results are summarised in Table 4. In agreement with Jongkees *et al.* (1962) and Downie and Simmons (1975), no consistent relationship could be established between the side of the lesion and the direction of the nystagmus. Thus, as will be seen, the nystagmus sometimes beats towards the affected and sometimes towards the healthy side. On one occasion electronystagmographic studies carried out upon a patient immediately after an attack of Ménière's disease showed a spontaneous nystagmus of small amplitude and low frequency beating towards the affected side.

Examination of each patient's history showed that the appearance of spontaneous nystagmus to-

Fig. 1 Electronystagmographic recordings of spontaneous nystagmus in a case of Ménière's disease first, second, and third degree nystagmus in the presence of optic fixation.

The superimposed full line curves indicate the intended direction of gaze.

Fig. 2 Electronystagmographic recordings of spontaneous nystagmus in a case of Ménière's disease—effect of eye closure on spontaneous nystagmus. Note enhancement of nystagmus in all directions of gaze.

The superimposed full line curves indicate the intended direction of gaze.

Fig. 3 Electronystagmographic recordings of spontaneous nystagmus in a case of Ménière's disease—effect of darkness on spontaneous nystagmus. Note enhancement of spontaneous nystagmus in all directions of gaze.

The superimposed full line curves indicate the intended direction of gaze.

Table 1 *Ménière's disease*

Case number		Eyes open, fixating	Eyes closed	Eyes open in darkness
3	Probably bilateral Ménière's disease	1st, 2nd, and 3rd° nystagmus to left	Enhanced	Enhanced
11	Left Ménière's disease	1st° nystagmus to left	Enhanced? Some 2nd° nystagmus to left	Enhanced
12	Bilateral Ménière's disease	1st° nystagmus to left	Enhanced	Enhanced
14	Left Ménière's disease	None	Some 1st° nystagmus to left	Clear 1st° nystagmus to left
19	Right Ménière's disease	None	1st° nystagmus to left	1st° nystagmus to left enhanced
28	Right Ménière's disease	None	None	1st° nystagmus to right. Also some 2nd° to right
30	Left Ménière's disease	None	1st, 2nd, and 3rd° nystagmus to left	1st, 2nd, and 3rd° nystagmus to left
32	Right Ménière's disease	None	Some 1st° nystagmus to left	Clear 1st° nystagmus to left
6	Right Ménière's disease	None	1st, 2nd, and 3rd° nystagmus to right	1st, 2nd, and 3rd° nystagmus to right
31	Left Ménière's disease	None	1st° nystagmus to left	1st° nystagmus to left
2		None	None	None
4				
10				
29				
33				

Table 2 *Vestibular neuronitis*

Case number		Eyes open, fixating	Eyes closed	Eyes open in darkness
20	Left vestibular neuronitis	1st° nystagmus to right	Enhanced	Enhanced 2nd and ? 3rd° nystagmus to right
21	Right vestibular neuronitis	None	Some 1st and 2nd° nystagmus to left	Clear 1st and 2nd° nystagmus to left
22	Right vestibular neuronitis	None	1st, 2nd, and 3rd° nystagmus to left	1st, 2nd, and 3rd° nystagmus to left enhanced compared with eyes closed
26	Right vestibular neuronitis	Fine 1st° nystagmus to left	1st and 2nd° nystagmus to left	1st and 2nd° nystagmus to left
27	Right vestibular neuronitis	1st° nystagmus to left	Gaze not maintained at left position	Gaze maintained at left position, 1st° nystagmus to left, enhanced 2nd° nystagmus to left
15		None	None	None
18				
23				
24				
25				

Table 3 *Miscellaneous*

Case number		Eyes open, fixating	Eyes closed	Eyes open in darkness
1	Right lesion ? vestibular neuronitis ? unusual Ménière's disease ? labyrinthine otosclerosis	None	Fine 1st° nystagmus to left	Clear 1st° nystagmus to left
5	Left 8th nerve neuropathy	None	None	1st° nystagmus to left
16	Left division of 8th nerve	None	None	1st and fine 2nd° nystagmus to right
7	Right serious labyrinthitis	Some 1st° nystagmus to right	1st, 2nd, and 3rd° nystagmus to right	1st, 2nd, and 3rd° nystagmus to right
8	Left traumatic labyrinthitis	None	1st° nystagmus to right	1st° nystagmus to right
13		None	None	None
17				

Table 4 *Relationship of direction of nystagmus to side of lesion*

Peripheral cases	Number	Nystagmus towards damaged side	Nystagmus opposite damaged side
Ménière's disease	8	6	2
Vestibular neuronitis	5	—	5
Miscellaneous	5	2	3
Total number	18	8	10

gether with its amplitude was directly related to the frequency and severity of the attacks of vertigo. By contrast the electronystagmographic findings were negative in patients with Ménière's disease (cases 2, 4, 9, 10, 29, 33—Table 1) when (a) caloric responses were normal or showed only slight paresis and (b) attacks of vertigo were very mild, or in vestibular neuronitis patients (cases 15, 18, 23, 24, 25, 34—Table 2) when (a) there was a

longstanding history and (b) caloric responses were reduced bilaterally.

Discussion

The electronystagmographic pattern of response found to be uniquely associated with peripheral disorders of the vestibular system can be summarised as (a) the appearance of a unidirectional nystagmus with eye closure or in darkness in the absence of any nystagmus in the light, or (b) a striking enhancement of nystagmus present in the light by eye closure or in darkness. This pattern was found in almost 60% of the cases studied. In the remainder, in whom the electronystagmographic findings were negative, the clinical evidence was indicative of a mild or inactive lesion. To this extent, therefore, these studies have provided ample confirmation of a pattern of response pathognomonic of peripheral vestibular pathology and one, moreover, which appears to correlate well with the severity of the symptoms.

One final comment, however, is called for on the relative effects of eye closure or darkness. Both result in an enhancement of nystagmus from the removal of the inhibitory effects of optic fixation which normally tends to hold nystagmus in check. Nevertheless, certain differences are apparent. Thus in three cases nystagmus was absent in the light and with eye closure but present in darkness, and in another six cases absent in the light and present with both eye closure and darkness but of appreciably greater amplitude in the latter. In other words, enhancement of spontaneous nystagmus by darkness is systematically greater than that by eye closure. The suggestion is that different physiological mechanisms may be involved and, in particular, that the act of closing the eyes brings into play certain reticular mechanisms associated with sleep which are not present in darkness.

While, therefore, both procedures are effective in revealing peripheral vestibular pathology, recording in darkness would seem to provide the most sensitive measure.

References

- Aschan, G., Bergstedt, M., and Stahle, J. (1956). Nystagmography. *Acta Otolaryngologica (Stockholm)*, Supplement **129**, 1-103.
- Belkahlia, A., Andrianjatovo, J., and Maspétiol, R. (1970). Etude electronystagmographique de sclérose en plaques à la phase de début. *Annales d'Otolaryngologie (Paris)*, **87**, 573-580.
- Downie, D. B., and Simmons, F. B. (1975). Spontaneous nystagmus direction does not indicate laterality. *Archives of Otolaryngology*, **101**, 358-360.
- Hood, J. D. (1968). Electronystagmography. *Journal of Laryngology and Otology*, **82**, 167-183.
- Jongkees, L. B. W., Maas, J. P. M., and Philipszoon, A. J. (1962). Clinical nystagmography. A detailed study of electronystagmography in 341 patients with vertigo. *Practica Otorhinolaryngologica (Basel)*, **24**, 65-93.
- Jongkees, L. B. W., and Philipszoon, A. J. (1964). Electronystagmography. *Acta Otolaryngologica (Stockholm)* Supplement **189**, 1-111.
- Jung, R., and Kornhuber, H. H. (1964). Results of electronystagmography in man. The value of optokinetic, vestibular and spontaneous nystagmus for neurologic diagnosis and research. In *The Oculomotor System*, pp. 428-488. By M. B. Bender. Harper and Row: New York.
- Padovan, I., Pansini, M., Ledinski, G., Ribaric, K., and Necovetic, L. (1975). Electronystagmography in the pathology of the central nervous system. *Acta Otolaryngologica (Stockholm)*, **79**, 319-324.
- Pastor Bartual, J., and Gaona Roquette, J. (1974). Electro-nystagmographic findings in Ménière's disease. *Anales Otorrinolaringológicos Ibero-Americanos*, **1**, 36-58.
- Rubin, W. (1968). Nystagmography. *Archives of Otolaryngology*, **87**, 266-271.