Electronystagmographic criteria in neuro-otological diagnosis

2 Central nervous system 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 light, in darkness, and with eye closure have been carried out on (a) 33 patients with acoustic neurinomata pressing on the brain stem, and (b) 10 patients with lesions involving the brain stem at a high level. Patterns of response characteristics of each group have been identified in 21 out of 28 cases of group a, the spontaneous nystagmus present in light was abolished by eye closure and inhibited in darkness, nystagmus being absent in the remaining five; in group b the greater proportion (70-80%) of patients presented with spontaneous nystagmus in light which was abolished by both eye closure and darkness.

Spontaneous nystagmus can result from lesions affecting the vestibular pathways at any point between the periphery and the cerebral cortex. Characteristically the nystagmus takes on a saw-tooth waveform with a fast component in one direction followed by a slow component in the other, and on direct observation of the eyes is, in general, indistinguishable in appearance irrespective of the underlying pathology. An important exception stems from the fact that, since nystagmus of peripheral origin is consistently unidirectional, any nystagmus that changes direction with change in gaze deviation must be central in origin. By contrast a spontaneous nystagmus beating in only one direction can be either of peripheral or central origin. As long ago as 1917, however, Holmes noted that removal of optic fixation had a striking effect upon the character of the nystagmus according to the level of the causative lesion, and used this to good effect in the differential diagnosis of labyrinthine and cerebellar lesions.

More recently, with the advent of electronystagmography which has facilitated the accurate recording of eye movements either with eye closure or in total darkness, various authors have elaborated on Holmes's observations in respect of the identification of spontaneous nystagmus result-

ing from lesions at various levels of the central nervous system. Hood (1968), in particular, has described three distinct patterns of response which he maintains are pathognomonic of peripheral lesions, brain stem lesions affecting the vestibular nuclei, and high brain stem lesions. These are summarised in Table 1 in respect of the relative effects of eye closure or darkness upon spontaneous nystagmus observed in the light.

Table 1 *Relative effects of eye closure and darkness on spontaneous nystagmus*

Type of lesion	Eye closure	Darkness
Peripheral	Enhanced	Enhanced
Vestibular nuclei	Abolished	Inhibited
High brain stem	Abolished	Abolished

The preceding paper (Korres, 1978) has dealt with and confirmed the finding of a characteristic electronystagmographic pattern of response associated with lesions of the peripheral vestibular system. The present communication is concerned with the establishment of similar criteria in terms of localisation in central vestibular lesions, with a view to determining in particular their generality in selected groups of patients. Observations were also carried out upon optokinetic nystagmus and following movements.

Two groups of patients were selected for study: (a) 33 unselected cases of acoustic neuromata con-

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

firmed by histological examination, and (b) 10 cases in which the disease involved, among other structures, the brain stem at a high level. In both groups the pattern of spontaneous nystagmus with and without optic fixation was examined in relation to the clinical and operative findings including a full neuro-otological examination. Details of method are given in the earlier paper (Korres, 1978). The findings in the two groups will be considered separately.

Acoustic neuromata

RESULTS

The cases were classified in four groups according to the pattern of electronystagmographic responses in the light, in the dark, and with eye closure.

Group 1 This group included 21 patients whose pattern of response corresponded closely to that described by Hood (1968) as being characteristic of brain stem lesions affecting the vestibular nuclei—namely, abolition of the nystagmus with eye closure and inhibition in respect of slow component velocity in darkness. Seventeen of these exhibited, with eyes open in light, a first degree nystagmus on right and left lateral gaze, well-sustained and of regular rhythm. Second degree nystagmus was present in only six of these. The remaining four patients in the group had nystagmus on only left or right lateral gaze.

With eyes closed the nystagmus disappeared in 17 cases, while in four it was markedly inhibited. In lateral positions the gaze appeared particularly unsteady. In darkness the nystagmus reappeared, exhibiting a reduction in the velocity of the slow component. Occasionally it was of larger amplitude but less frequent and irregular. The following example is typical of the patients in this group.

A 65 year old male had a sudden onset of right sided deafness approximately 10 years ago. Recently, he had developed unsteadiness of gait, nausea, and vomiting. During clinical examination the sensation over the right trigeminal nerve territory was found to be diminished, and there was spasm of the right facial muscles. Sensation of the palate and pharynx was diminished on the right. Finally, tremor of the left upper limb was present. Neuro-otological examination showed absent hearing and absence of caloric responses on the right. The spontaneous nystagmus in this case is illustrated in Fig. 1. On right and left lateral gaze, first degree nystagmus to the right and left respectively can be seen (Fig. 1a). Figure 1b shows the effect of eye closure. The nystagmus disappeared in both lateral positions and there was a tendency for the eyes to deviate towards the midline. Figure 1c illustrates the results obtained in

darkness. The nystagmus reappeared but was irregular, less frequent, of larger amplitude but much inhibited in respect of the slow component velocity.

Group 2 Five atypical cases are included in the second group, each presenting with a different electronystagmographic pattern. Two showed first degree nystagmus to the left and right, which remained equally vigorous or even increased with the eyes closed and in darkness. The others had little in common. Occasionally unilateral nystagmus occurred in light. With the eyes closed and in darkness unilateral nystagmus persisted or became bilateral.

Group 3 The third group contained two cases with all the characteristics of a peripheral lesion. One of these, a 50 year old patient, presented with a history of sudden onset of hearing loss in the left ear four and a half years previously, with recent deterioration. Neurological examination was normal. Neuro-otological examination showed severe hearing loss on the left of 70–80 dB and absence of caloric responses on the same side. A large tumour invading the pons was removed. Electronystagmographic studies were carried out two months before the operation. Figure 2a shows the tracing obtained in the light. No nystagmus is evident in the straight ahead position or on lateral deviation of the eyes. The effect of eye closure is shown in Fig. 2b. In the primary position of gaze a regular second degree nystagmus to the right made its appearance. Gaze deviation to the right was poorly sustained, and the eyes had a tendency to deviate towards the midline. The left lateral gaze was not well-sustained and the eyes returned to the midline. Nystagmus to the right reappeared but was less regular. Figure 2c shows the results obtained in darkness. The nystagmus was less vigorous in all three positions in comparison with Fig. 2b. By contrast lateral gaze was well-sustained. The second case presented similar features.

Group 4 The last group included five cases in whom no nystagmus was apparent during the course of the electronystagmographic examination.

Direction of spontaneous nystagmus

In Table 2 the cases have been divided into groups according to the direction of the nystagmus in light. A clear preponderance to the opposite side of the lesion was a prominent feature. A similar distribution has been reported by Dix and Hallpike (1966).

Size of tumour

All the cases presenting with spontaneous nystagmus were found to have large tumours pressing upon the brain stem. In the only patient in whom

Fig. 1 Electronystagmographic recordings of spontaneous nystagmus in a case of acoustic neuroma—
 (a) 1st degree nystagmus to the left and right in the presence of optic fixation; (b) effect of eye closure upon spontaneous nystagmus. The nystagmus disappeared in both lateral positions; (c) effect of darkness upon spontaneous nystagmus. The nystagmus reappeared. Note inhibition in respect of the slow component velocity.

Full line tracings in this and subsequent figures indicate position of fixation mark.

Fig. 2 Electronystagmographic findings in a case of acoustic neuroma characteristic of a peripheral lesion: (a) no nystagmus can be seen in the presence of optic fixation; (b) and (c) effect of eye closure and darkness upon spontaneous nystagmus, 1st, 2nd, and 3rd degree nystagmus appeared beating to the right.

Table 2 *Direction of nystagmus relative to side of the tumour*

Type of nystagmus	Number
No nystagmus	8
1st° nystagmus to the same side	1
1st and 2nd° nystagmus to the same side	0
1st and 2nd° nystagmus to the same side. 1st° nystagmus to the opposite side	1
1st° nystagmus to the same side. 1st° nystagmus to the opposite side	12
1st° nystagmus to the same side. 1st and 2nd° nystagmus to the opposite side	6
1st and 2nd° nystagmus to the opposite side	1
1st° nystagmus to the opposite side	4

the tumour was confined to the meatus (10–12 mm in diameter) no spontaneous nystagmus was present.

Optokinetic nystagmus and following movements

None of the five patients in group 4 without spontaneous nystagmus presented with either abnormal optokinetic nystagmus or following movements. By contrast all subjects with spontaneous nystagmus in light showed abnormal optokinetic and following movements. This, in turn, seemed to be related to the amplitude and frequency of the former. Thus, Fig. 3a illustrates an example of a left acoustic neuroma showing first degree nystagmus to the left and right. The waveform of the optokinetic response was grossly deranged. The example shown in Fig. 3b is of a patient with a right acoustic neuroma, again with first degree nystagmus to right and left but of smaller amplitude than in the previous example. The optokinetic nystagmus is clearly less abnormal in this patient. Finally, when directional preponderance of optokinetic nystagmus was present it was consistently directed towards the side of the nystagmus of larger amplitude present in light, as seen in the example shown in Fig. 3b.

DISCUSSION

The pattern of spontaneous nystagmus described by Hood (1968)—that is to say present in light, abolished by eye closure, and inhibited in darkness—was present in 21 out of 28 cases, nystagmus being absent in five.

In the four cases in group 1 in whom some nystagmus was present with eye closure, nystagmus in the light was particularly prominent. In the two patients mentioned in group 2, with first degree nystagmus to the left and right, which remained equally vigorous or even increased with the eyes closed and in darkness, it is perhaps significant that the tumour extended below the level of the vestibular nuclei. Suga and Lindsay (1976) have recently

presented clear histological evidence of degenerative changes in the inner ear, including fibrosis and ossification of the semicircular canals, in three cases of acoustic neurinoma which they attribute to chronic partial obstruction of blood circulation by the tumour in the internal auditory meatus. In this event it may not be unexpected that a small proportion of acoustic neurinomata should present with a peripheral pattern of spontaneous nystagmus which in fact is a true reflection of end organ involvement in these cases. A further possibility that needs to be considered, however, is that if the growth of the tumour is such that nerve fibre involvement is of an unusually abrupt onset, central compensation will be incomplete and here, too, the spontaneous nystagmus will manifest as a peripheral variety.

The origin of spontaneous nystagmus in acoustic neurinomata has been considered by Dix and Hallpike (1966). They suggest that two basic mechanisms are involved in evoking spontaneous nystagmus. One is related to the balance of the vestibular tonus elements, and the other to the balance of so-called voluntary deviation maintenance elements, the latter being located more rostrally in the brain stem. They postulate that a left acoustic neuroma presses first upon and affects the left vestibular tonus elements. This results in a spontaneous nystagmus to the right. At this time or a little later, the second mechanism—that for left voluntary gaze—is affected, and this results in a spontaneous nystagmus to the left. In either event the nystagmus results from the tonic deviation of the eyes (the slow component) and the corrective movements (the fast components) required to maintain eccentric deviation of the eyes.

In support of their hypothesis these authors describe two cases of acoustic neurofibromata in which the operative findings indicated an unusually high extension in the brain stem. Both presented with ipsilateral nystagmus, the presumption being that this resulted from involvement of the voluntary deviation maintenance elements. Little support for these notions, however, is to be found in the material presented here. Thus in the nine cases shown in Table 3, the tumour, according to the neurosurgical report, extended above the level of the vestibular nuclei. In seven of these, nystagmus both to left and right was present, greater in the ipsilateral direction in three and in the contralateral direction in two. In the remaining two cases only first degree contralateral nystagmus was recorded.

Concerning the mechanism of the nystagmus, examination of the electronystagmographic tracings of the patients in this group revealed no

Fig. 3 Relation between amplitude and frequency of spontaneous nystagmus in the presence of optic fixation, and derangement of optokinetic nystagmus. Note the larger amplitude and higher frequency of the spontaneous nystagmus in (a) gives rise to a greater derangement of optokinetic nystagmus than in (b).

Table 3 Spontaneous nystagmus in tumours extending high in the brain stem

Case number	Side of tumour	Level	Nystagmus
1	Right	High	1st° to left and right
4	Right	High	1st° to left and right. Right > left. 2nd° to right
5	Right	High	1st° to left and right. Right > left
6	Right	High	1st° to left and right
8	Right	High	1st° to left and right. Left > right
9	Left	High	1st° to left and right. 2nd° to right
14	Left	Very high	1st° to left and right. Left > right
16	Left	Very high	1st° to right
17	Right	High	1st° to left

systematic correlation between eye deviation and nystagmus as is to be expected from Dix and Hallpike's hypothesis. The first example of this is a patient with a left acoustic neuroma who developed a first degree nystagmus to the left in the light as shown in Fig. 4. According to Dix and

Hallpike (1966) one would expect deviation of the eyes towards the right when fixation was removed. In fact, with the eyes closed a sudden and vigorous deviation to the left was observed from the right lateral position. From the left lateral position sudden but less vigorous deviation of the eyes further to the left occurred.

The second example is of a patient with a right acoustic neuroma who developed a large first degree nystagmus to the left in the light, as shown in Fig. 5. Again, according to Dix and Hallpike (1966), the eyes should be deviated, predominantly towards the right when closed and in darkness. However, as can be seen, a left deviation was observed in darkness, with inability to sustain deviation to the right.

The conclusion from these two examples is that tonic deviation of the eyes is not by itself the primary cause of spontaneous nystagmus in vestibular nuclei lesions.

Fig. 4 Incoordination of spontaneous nystagmus and eye deviation. The latter is recorded in the absence of optic fixation.

Fig. 5 Similar example as in Fig. 4.

High brain stem lesions

Precise localisation of lesions above the level of the vestibular nuclei is not easy in view of the fact that numerous structures are usually involved. In consequence, a somewhat different approach has been adopted in which a number of cases were selected presenting with the electronystagmographic pattern described by Hood (1968)—that is, spontaneous nystagmus in light abolished both with eye closure and darkness. Careful examination was then made of the available clinical and other data to try to assess the site of the causative lesion. From among these cases seven showed clear evidence of organic lesion in the upper brain stem as assessed by neurological examination.

In Table 4 the neurological diagnoses of the subjects studied are presented. Most suffered from widespread disease of the central nervous system. The presence or absence of clinical abnormalities related to the upper brain stem is given in the right hand column. Cases 1, 2, 4, 5, 7, and 10 present evidence of involvement of the brain stem above the level of the vestibular nuclei. In case 9 the existence of a longstanding lesion of the same anatomical area might be acceptable. In case 8, despite the presence of a functional overlay, the demonstration of up-beat nystagmus can be taken to justify the presence of a high brain stem lesion caused by drug intoxication. Cases 3 and 6 were on anticonvulsant treatment.

RESULTS

A summary of the findings on spontaneous nystagmus, following movements, and optokinetic nystagmus is given in Table 5. Typical findings are shown in Fig. 6: (a) shows spontaneous nystagmus with optic fixation in light, (b) and (c) the

Table 4 *Diagnosis and indications of high brain stem pathology*

Case number	Diagnosis	Upper brain stem abnormalities
1	Vascular lesion of midbrain	Diplopia, squint, and bilateral ptosis. Bilateral involvement of third cranial nerve muscles
2	Multiple sclerosis	Inequality of pupils. Right pupil reacted less in light. Diplopia in almost all directions of gaze. Inability to obtain full abduction of left eye
3	Epilepsy due to trauma	—
4	Multiple sclerosis	Signs of internuclear ophthalmoplegia with 1st° vestibular nystagmus upwards
5	Degenerative disease of brain stem and peripheral nerves and cord	Lid retraction and some lid lag
6	Focal Jacksonian epilepsy	—
7	Familial cerebellar atrophy	Left pupil irregular, unreactive to direct and consensual light on convergence. Left partial ptosis. Some limitation of upward gaze. Left eye not elevated as well as the right. 1st° vestibular nystagmus upwards
8	Hysteroepilepsy and drug intoxication	1st° vestibular nystagmus upwards
9	Degenerative or demyelinating disease, polyneuritis recently	Defect of conjugate gaze to left, right, and vertically. Bilateral ptosis
10	Multiple sclerosis	Weakness in abduction and adduction both eyes. Inconstant 1st° vestibular nystagmus upwards

results on eye closure and in darkness, and (d) following movements and optokinetic nystagmus.

Following movements and optokinetic nystagmus
Following movements and optokinetic nystagmus were always deranged. However, while in the case of acoustic neurinomata the degree of derangement of both following movements and optokinetic

Fig. 6 Electronystagmographic recordings of spontaneous nystagmus in a patient with multiple sclerosis involving, among other structures, the upper brain stem: (a) 1st degree nystagmus to the left and right ($L > R$) in the presence of optic fixation; (b) and (c) abolition of spontaneous nystagmus with eye closure and in darkness; (d) deranged following movements and sluggish optokinetic responses together with directional preponderance to the left.

Table 5 Summary of electronystagmographic findings in 10 patients with high brain stem lesion

Case number	Light	Eyes closed	Darkness	Following movements	Optokinetic nystagmus
1	1st° VN* to left and right	Abolished	Abolished	Not done	Not done
2	1st° VN* to left and right. Some 2nd° to right	Abolished	Abolished	Deranged	Deranged. Some DP† to right
3	1st° VN* to left. Some 2nd° to left	Abolished	Abolished	Deranged	Deranged. DP‡ to left
4	1st° VN* to left and right	Abolished	Abolished	Deranged	Very sluggish. Slight DP‡ to left
5	1st° VN* to left and right	Abolished	Abolished	Deranged	Not done
6	Small 1st° VN* to left	Abolished	Abolished	Not done	Not done
7	1st° VN* to left and right	Abolished	Abolished	Deranged	Deranged
8	1st° VN* to left and right	Abolished	Abolished	Not done	Not done
9	1st° VN* to left	Abolished	Abolished	Deranged	Not done
10	1st° VN* to left and right	Abolished	SN† to right abolished SN† to left much inhibited	Deranged	Deranged. DP‡ to left

*VN = vestibular nystagmus; †SN = spontaneous nystagmus; ‡DP = directional preponderance.

nystagmus matched the magnitude of the spontaneous nystagmus, this was not always so with high brain stem lesions in which spontaneous nystagmus of small amplitude was at times accompanied by severe derangement of both following movements and optokinetic nystagmus.

DISCUSSION

It is apparent from these results and those described in the previous paper (Korres, 1978) that the electronystagmographic studies of spontaneous nystagmus applied in the manner described reveal patterns of response characteristic of the level of the causative lesion with a highly acceptable degree of confidence; 60% in peripheral lesions, 64% in acoustic neuromata with brain stem involvement, and 70% in high brain stem lesions.

The particular patterns associated with each group and dependent upon the different effects of eye closure or darkness have been summarised in Table 1. The striking finding is the systematic way in which the removal of optic fixation, either by eye closure or darkness, differentially affects the nystagmic response according to the level of the lesion from enhancement at the periphery to total abolition at a high level of the brain stem.

It is interesting that the effects of eye closure are more immediate and profound than those of darkness. This may, in part, be attributed to the intervention of certain reticular mechanisms associated with sleep and brought into activity by eye closure. The surprising hierarchical order described would seem to find even further extension in the recent publications of Dix (1974) and Hood (1976). These authors have shown that, in certain rare cases of tumours situated in the posterior part of the temporal lobe and presenting with spontaneous nystagmus beating towards the affected side, the actual direction of the nystagmus

may be reversed by removal of fixation. Of perhaps more general interest is the remarkable parallel in the findings of Ledoux and Demanez (1967), Corvera *et al.* (1968), and Haciska (1973) in induced nystagmus; thus it now seems to be well established that caloric-induced nystagmus is much enhanced in the absence of optic fixation in peripheral lesions but substantially less so, or even inhibited, in central lesions. Furthermore, Carmichael *et al.* (1961) have shown that in temporal lobe lesions a directional preponderance of caloric or rotational nystagmus towards the side of the lesion is consistently reduced or even reversed in direction when the tests are carried out in darkness. In other words, the level of the lesion exerts the same influence with respect to the removal of fixation upon induced nystagmus as it does upon spontaneous nystagmus. While it is difficult at present to specify the anatomical mechanisms involved, which clearly involve the interaction of the central pathways subserving optic fixation and the vestibular system at different levels of the central nervous system, there can be little doubt that systematic studies of spontaneous nystagmus of the kind described can make a significant contribution to neuro-otological diagnosis.

References

- Carmichael, E. A., Dix, M. R., Hallpike, C. S., and Hood, J. D. (1961). Some further observations upon the effect of unilateral cerebral lesions on caloric and rotational nystagmus. *Brain*, **84**, 571-584.
- Corvera, J., Somonte, R., Torres, G., and Lopez, G. (1968). Une nouvelle méthode dans le diagnostic différentiel des lésions périphériques et des lésions centrales des voies vestibulo-oculomotrices. *Revue de Laryngologie (Bordeaux)*, **89**, 617-642.
- Dix, M. R. (1974). The vestibular acoustic system. In *Handbook of Clinical Neurology*. Edited by P. J. Vinken and B. W. Bruyn. Vol. 16, pp. 301-340.

- North-Holland Publishing Company: Amsterdam, American Elsevier Publishing Co.: New York.
- Dix, M. R., and Hallpike, C. S. (1966). Observations on the clinical features and neurological mechanisms of spontaneous nystagmus resulting from unilateral acoustic neuro-fibromata. *Acta Otolaryngologica (Stockholm)*, **61**, 1–22.
- Haciska, D. T. (1973). The influence of drugs on caloric-induced nystagmus. *Acta Otolaryngologica (Stockholm)*, **75**, 477–484.
- Holmes, G. (1917). The symptoms of acute cerebellar injuries due to gunshot injuries. *Brain*, **40**, 461–535.
- Hood, J. D. (1968). Electro-nystagmography. *Journal of Laryngology and Otology*, **82**, 67–183.
- Hood, J. D. (1976). The use of electro-nystagmography in neuro-otological diagnosis. In *Disorders of Auditory Function*. Edited by S. D. G. Stephens, pp. 215–218. Academic Press: London, New York, San Francisco.
- Korres, S. (1978). Electronystagmographic criteria in neuro-otological diagnosis I Peripheral lesions. *Journal of Neurology, Neurosurgery, and Psychiatry*, **41**, 249–253.
- Ledoux, A., and Demanez, J. P. (1967). Nystagmographie des yeux ouverts ou fermés au cours de l'épreuve calorique. *Acta Oto-Rhino-Laryngologica, Belgica*, **21**, 31–40.
- Suga, F., and Lindsay, S. R. (1976). Inner ear degeneration in acoustic neuroma. *Annals of Otology, Rhinology and Laryngology*, **85**, 343–358.