

# Hearing loss in brainstem disorders

LINDA M LUXON

*From the Department of Otoneurology and the Medical Research Council, Hearing and Balance Unit, National Hospital for Nervous Diseases, London*

**SUMMARY** A retrospective study of 309 unselected patients with brainstem disorders was carried out to establish the incidence and degree of hearing loss in this group as a whole and in each of three separate pathologies: multiple sclerosis, vertebro-basilar ischaemia and brainstem tumours. Pure tone audiograms were corrected for age and sex and upon analysis, thereafter, 59% and 26% of patients were found to have hearing thresholds in excess of 10dB and 30dB respectively. The hearing loss tended to be slight to moderate, 87% of those with a deficit were in the range 11–59dB, and tended to involve the higher frequencies. No characteristic audiometric configuration emerged. 75% of those with a hearing loss suffered a bilateral deficit. Considering the multiple sclerosis, vertebro-basilar insufficiency and tumour groups individually, similar findings were observed with the mildest losses occurring in multiple sclerosis and the most severe in the tumour group.

Auditory derangements in brainstem disorders were first reported at the end of the last century<sup>1,2</sup> and subsequently conflicting reports have appeared in the literature covering various aspects of the topic.<sup>3–9</sup> Many of these are open to criticism on a number of counts: the selected nature of the patient groups, the small numbers of patients studied and neglect of the influence of age and sex on pure tone hearing levels.

The present study was undertaken to investigate the incidence of hearing loss as judged by age and sex corrected pure tone audiograms in a large series of unselected patients with a variety of brainstem pathologies.

## Method

In order to evaluate the incidence and degree of hearing loss in patients with brainstem disorders, a retrospective analysis of 367 patients, investigated in the Department of Otoneurology at the National Hospital, Queen Square, London, between 1941 and 1976 was undertaken. No selection criteria were applied to this group of patients and they consisted of the first sequential 367 patients on the retrieval system for "Brainstem Disorders." Twenty-seven patients

with a diagnosis of cerebello-pontine angle tumour or acoustic neuroma were excluded, as deafness in this group was more likely to result from a nerve fibre rather than a brainstem lesion. In addition, a further 31 patients with deafness clearly attributable to some other cause for example otosclerosis, otitis media, trauma, noise, were also excluded. The remaining 309 patients, comprised 158 males and 151 females between the ages of five and 78 years (table 1).

The diagnosis of brainstem pathology was made on clinical (neurological and otological) grounds in all patients, and confirmed at operation or post-mortem examination in 108 patients (35%). The pure tone audiograms were reviewed and threshold sensitivities at each frequency were corrected for age and sex according to Hinchcliffe.<sup>10</sup>

As a first step in the analysis, all the age corrected audiograms with hearing levels under 10dB at all frequencies were removed. Three parameters of hearing loss were then studied in the remaining audiograms: the incidence, the severity and the audiometric configuration. These results were studied in the group as a whole, and separately in each of three major brainstem disorders: multiple sclerosis (MS), vertebro-basilar insufficiency (VBI) and malignant tumours (NG). A fourth group comprised the remaining brainstem pathologies.

Address for reprint requests: Dr LM Luxon, The National Hospital, Queen Square, London WC1N 3BG.

Accepted 7 February 1980

Table 1 Age and sex distribution of 309 patients

Age (yr)	0-9	10-19	20-29	30-39	40-49	50-59	60-69	70-79
Male	9	18	17	22	34	33	22	3
Female	3	9	16	23	37	30	29	4
Total	12	27	33	45	71	63	51	7
%	3.8	8.7	10.7	14.6	23.0	20.4	16.5	2.3

## Results

Of the 309 patients studied, 52 (16.8%) had multiple sclerosis, 57 (18.4%) had vertebral basilar insufficiency, 84 (27.2%) had malignant tumours and 116 (37.5%) had various other brainstem disorders including syringobulbia, angiomas, abscesses, encephalomyelitis, collagen vascular disorder, myelopathies, familial degenerative conditions, tuberculosis and benign cysts. Twenty six patients (8.4%) complained of hearing loss at the time of presentation. One hundred eighty one (58.6%) patients had hearing levels in excess of 10dB at one or more frequencies on the age and sex corrected pure tone audiograms (table 2). The incidence of hearing loss within the three major sub-groups varied between 57.7% to 64.3%. The degree of deficit was classified according to the most severely affected frequency after correction for age and sex: slight (11-29dB), moderate (30-59dB), severe (60-89dB) and complete (>89dB). Using this classification, slight deafness was present in 33% of all patients studied, moderate in 18%, severe in 5% and complete in 3%. Similar trends were apparent in all the sub-groups. However, more patients (25%) with vertebral basilar insufficiency were afflicted with moderate loss than patients with multiple sclerosis or malignant tumour (17% and 15% respectively). Severe and total hearing loss were more commonly seen in the tumour group.

Table 3 illustrates that the hearing loss was bilateral in approximately three quarters of the patients, but in only two thirds of the patients

with multiple sclerosis. In addition, the deafness was symmetrical at the two ears in 71% of the 132 patients with bilateral hearing loss.

The hearing levels averaged in respect of the total number of patients with (a) deafness greater than 10dB and (b) deafness greater than 30dB are shown in fig (a) and (b). Consideration was given to the individual audiometric configurations of the hearing loss adopting a modified form of the Carhart classification<sup>11</sup> namely high tone loss, low tone loss, dome loss, flat loss and island loss. No clear pattern emerged which could be associated with any particular group. However, of the 181 patients, 79% had hearing loss solely, or inclusive of, the higher frequencies.

## Discussion

The constellation of symptoms and signs resulting from involvement of multiple cranial nerve nuclei, long tract and cerebellar connections is characteristic of brainstem disease. The presence of audiological symptoms has previously been considered unusual<sup>8,12</sup> since destructive lesions in this region often produce rapidly fatal symptoms. This view has, however, been disputed.<sup>9</sup> In the present study, 59% of 309 unselected patients with brainstem disorders suffered some degree of hearing loss as judged by age and sex corrected pure tone audiograms, although only 8% of them complained of hearing loss, which was related to the presenting brainstem disorder. It should be emphasised that

Table 2 Distribution of severity of hearing loss in brainstem disorders (expressed as a percentage of the total number of patients in each group)

	Whole study		MS		VBI		NG		Others	
	No	%	No	%	No	%	No	%	No	%
Slight HL (11-29 dB)	101	32.8	19	36.5	20	35.1	29	34.5	33	28.5
Moderate HL (30-59 dB)	56	18.1	9	17.3	14	24.6	13	15.5	20	17.2
Severe HL (60-89 dB)	15	4.8	2	3.8	1	1.7	7	8.3	5	4.3
Complete HL (90 dB)	9	2.9	0	0	1	1.7	5	5.9	3	2.6
Total with HL	181	58.6	30	57.7	36	63.2	54	64.3	61	52.6
No HL	128	41.4	22	42.3	21	36.8	30	35.7	55	47.4

HL = hearing loss

Table 3 *Distribution of hearing loss*

	<i>No deaf</i>		<i>Bilateral</i>		<i>Unilateral</i>	
	<i>No</i>	<i>%</i>	<i>No</i>	<i>%</i>	<i>No</i>	<i>%</i>
All patients	181		132	72.9	49	27.1
MS	30		18	60.0	12	40.0
VBI	36		30	83.3	6	16.7
NG	54		41	75.9	13	24.1
Others	61		43	68.9	19	31.1

a higher percentage of auditory disturbances might well have been detected by more specialised audiometric tests.<sup>13 14</sup>

Loudness recruitment tests were available in 99 of the patients with deficit and in 78% of these recruitment was present. Loudness recruitment, of course, has historically been associated with lesions of the cochlear end organs and in consequence it could be argued that the hearing loss identified in these patients was not brainstem in origin. In this respect it is pertinent to recall that these were all patients with brainstem lesions in whom hearing loss was not a presenting symptom and the coincidental occurrence of a cochlear lesion in such an extremely high proportion can be deemed highly improbable. There remains the unlikely possibility that brainstem lesions can in some way give rise to end organ pathology. However, there is now a powerful body of opinion<sup>15-18</sup> which holds the view that recruitment is a feature not only of end organ but also of brainstem pathology. Although it has to be admitted that the issue cannot be resolved with certainty, the balance of probability in the light of the available evidence favours brainstem pathology as the cause of deafness.

Recruitment was absent in only 18 patients and of these, 11 had a bilateral deficit (4 with

malignant tumours, 2 with multiple sclerosis, 2 familial degenerative disorders, 1 vertebro basilar insufficiency, 1 myelopathy). In 1965, Dix<sup>19</sup> observed that deafness due to a lesion of the cochlear nerve fibres, whether attributable to tumour pressure upon the neurilemmal portion or demyelination involving the neuroglial portion, resulted in the absence of loudness recruitment. Later, Jerger and Jerger<sup>9</sup> commented that extra-axial brainstem pathologies (for example inflammatory viral and degenerative lesions) might produce deafness by involvement of the VIII nerve trunk rather than by involvement of the central auditory pathways. On this basis, the occasional occurrence of sensori-neural deafness with absence of loudness recruitment can be explained in terms of an extension of the lesion to the cochlear nerve fibres.

In small selected groups of patients, pure tone sensitivities in brainstem lesions have been variously reported as normal,<sup>6</sup> mildly impaired in the high frequency range<sup>3</sup> and severely impaired in one or both ears.<sup>20</sup> The present study of 309 unselected patients revealed that, although all degrees of deficit may occur, of those with hearing loss the majority (87%) suffer only slight to moderate impairment (that is, 11-59 dB).

Numerous attempts have been made to relate the distribution of hearing loss in brainstem disorders with the ear or ears affected and the levels and side of the lesion. Considering the anatomical configuration of the auditory pathways in the brainstem the high incidence (73% of those with hearing loss) of bilateral hearing loss observed in this study is readily explicable. Dix and Hood,<sup>15</sup> assuming a bilateral tonotopic organisation of auditory fibres above the coch-

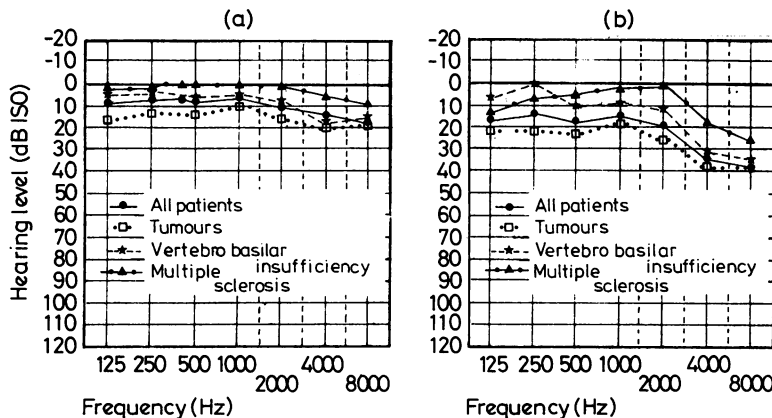


Figure Age and sex corrected hearing levels averaged in respect of the total number of patients with hearing thresholds greater than (a) 10 dB and (b) 30 dB at the most severely affected frequency.

lear nuclei, have postulated that a lesion above this level results in bilateral deafness, which is symmetrical in respect of the frequency distribution at both ears. In the present study, 71% of patients with bilateral deficit suffered symmetrical loss of this type. As noted earlier, certain pathologies may directly involve the cochlear nuclei or nerve trunk and may, therefore, give rise to asymmetrical bilateral hearing loss or, indeed, a unilateral deficit and in these instances loudness recruitment may be absent.

Previous authors, in limited studies of patients with brainstem disorders, have observed a tendency for greater hearing loss in the high frequency range. This trend was also apparent in the present study, in which 79% of patients with hearing loss were affected in the 4096-8192 Hz range.

Considering the individual audiometric configuration it was clear that no characteristic pattern emerged. Five different patterns were identified, with "high" frequency loss occurring most commonly in patients with bilateral deficit. Of the 52 patients affected by "island" loss (both bilateral and unilateral), 25% suffered the deficit at 4096 Hz and the possibility of noise trauma is, therefore, raised. While this explanation cannot be ruled out, the histories obtained gave no cause of suspicion on this point.

It should be stressed that the average configuration of hearing loss in brainstem disorders shown in the fig merely reflects the frequent occurrence of reduction of pure tone sensitivities in the high frequency range rather than the prevalence of the "high tone" configuration in individual patients.

#### *Multiple sclerosis*

Auditory disturbances in multiple sclerosis have been the subject of numerous studies,<sup>21-26</sup> but these too, have suffered the disadvantage of small numbers, patients selection and inadequate correction of audiograms for age. Vestibular symptoms and nystagmus are well documented in multiple sclerosis, but hearing loss has been reported to occur rarely. Indeed, only two patients in the present study complained of deafness at the time of presentation, but analysis of corrected pure tone audiograms revealed that 58% of this group of patients suffered some detectable reduction of threshold sensitivities.

Conraux and Collard<sup>27</sup> have reported that, although the hearing deficit in multiple sclerosis is variable, it is usually in the 20-40 dB range. This correlated well with the present finding that

the majority of patients with multiple sclerosis and hearing loss suffered slight deficit and a further 17% suffered moderate loss (30-59 dB). Compared with other groups, more patients with multiple sclerosis suffered unilateral deficit (40%) and this fact is well documented in the literature.<sup>28</sup> The frequency distribution of the hearing loss, revealed that 60% of patients with a deficit suffered loss in the high frequency range, as with other brainstem lesions. The individual audiometric configurations revealed no characteristic pattern of hearing loss in multiple sclerosis, but in both bilateral and unilateral deafness "island" loss occurred most commonly.

Hearing loss in multiple sclerosis has usually been attributed to lesions of the central auditory pathways in the brainstem,<sup>26</sup> but may be attributable to encroachment upon the cochlear nuclei.<sup>29</sup> A few reports have ascribed symptoms to lesions within the peripheral eighth nerve,<sup>30</sup> but these have not been supported by histological evidence and have been attempts to localise lesions on the basis of clinical symptoms. However, the present study would support this latter explanation as 40% of patients with a hearing loss attributed to multiple sclerosis were affected unilaterally.

#### *Vertebro-basilar insufficiency*

In 1894, Siebermann<sup>31</sup> described the blood supply to the inner ear from the basilar system. Consequent upon this distribution, deafness may result from reversible or irreversible ischaemic changes in the central auditory pathways, the eighth nerve or the inner ear. The present study confines itself to patients in whom there was neurological and otological involvement of the brainstem. Thirty-five per cent of patients were found to suffer hearing loss and this correlates well with previous reports.<sup>32-34</sup> Vertebro-basilar insufficiency tends to occur in the older patient and it is, therefore, reemphasised that in the present study pure tone audiograms were corrected for age and sex.

Upon consideration of the severity of the hearing loss in vertebro-basilar insufficiency as with other brainstem disorders, the loss was not marked. One third of the patients had slight hearing loss and one quarter, moderate loss. Only one out of 57 patients had a severe deficit.

Eighty three per cent of the patients with deafness suffered bilateral loss and in 83% of these patients, symmetrical hearing loss was present, as may be expected from ischaemia of the central auditory pathways. Six patients (17%)

suffered unilateral loss and of these four showed an "island" loss in the high frequency range. There was no characteristic audiometric configuration seen in vertebro-basilar ischaemia, but the "high tone" loss was most commonly seen in patients with bilateral deficit. Loss in the high frequency range again occurred most commonly both in the bilateral and unilateral deafness groups.

#### Tumours

Two thirds of all patients with malignant tumours of the brainstem suffered hearing loss (>10 dB) in the present study. Deafness has been reported in association with brainstem tumours,<sup>35-39</sup> contrary to the views of Liden<sup>17</sup> and Korsan-Bengsten<sup>40</sup> that fatal symptoms supervene before the development of audiological symptoms. Moreover, in the present study 11% of the patients with brainstem malignancies complained of deafness, temporally related to their illness.

The occurrence of moderate and severe hearing loss was more common in this group than in other brainstem pathologies (table 2) and this would be compatible with the aggressive and destructive nature of the disease process. In approximately 75% of patients affected, the deficit was bilateral suggesting intra-axial destruction of the central auditory pathways and indeed in 62% of these the deficit was symmetrical. Twenty-five per cent of patients with a hearing loss were affected unilaterally, suggesting destruction of the cochlear nuclei ipsilaterally, or involvement of the eighth nerve, either by direct infiltration, compression, or secondary ischaemia.

Reviewing the distribution of hearing loss, the high frequency levels were most commonly affected and indeed the "high tone" loss configuration was most commonly seen in bilateral hearing loss. In unilateral deficit, the "island" loss configuration (again usually affecting the high frequency range) was seen most commonly.

The author acknowledges the most kind help and encouragement throughout this work of Dr JD Hood, and the advice of Professor R Hinchcliffe.

#### References

- Weinland E. Ueber einem Tumor der Vierhügel gegend und über die Beziehungen der hinteren Vierhügel zu Gehörstörungen. *Arch Psychiatr Nervenkr* 1894; **26**:363-80.
- Siebertmann F. Ueber die zentrale Hörbahn und über ihre Schädigung durch Geschwülste des Mittelhirns speciell der Vierhügel gegend und der Haube. *Z Ohrenheilkunde* 1896; **29**:28-91.
- Antonelli A, Calearo C, DeMetri T. On auditory function in brainstem diseases. *Int Audiology* 1963; **2**:55-61.
- Benitez JT, Covera J, Novoa V. Auditory manifestations of cochlear and retrocochlear lesions in humans. *Ann Otol Rhinol and Laryngol* 1966; **75**:149-61.
- Calearo C, Antonelli AR. Audiometric findings in brainstorm lesions. *Acta Otolaryngol (Stockh)* 1968; **66**:305-19.
- Parker W, Decker RL, Richards NG. Auditory function and lesions of the pons. *Arch Otolaryngol* 1968; **87**:228-40.
- Haguenaer JP. Deafness caused by brainstem lesions. *Rev Otoneuroophthalmol* 1973; **45**:69-74.
- Liden G, Korsan-Bengsten M. Audiometric manifestations of retrocochlear lesions. *Adv Otorhinolaryngol* 1973; **20**:271-87.
- Jerger J, Jerger S. Auditory findings in brainstem disorders. *Arch Otolaryngol* 1974; **99**:342-50.
- Hinchcliffe R. Correction of pure tone audiograms for advancing ages. *J Laryngol Otol* 1959; **73**:830-2.
- Carhart R. Improved method for classifying audiograms. *Laryngoscope* 1945; **55**:640-62.
- Bocca E, Calearo C. *Central hearing processes. Modern Development in Audiology*. New York: Academic Press, 1963: 337.
- Antonelli A. *Sensitized tests: results in brainstem lesions and diffusive central nervous system disease*. In: Rojskaer C, ed. *Speech Audiometry*, 2nd Danavox Symposium, 1970: 130-9.
- Morales-Garcia C, Poole JP. Masked speech audiometry in central deafness. *Acta Otolaryngol* 1972; **74**:307-16.
- Dix MR, Hood JD. Symmetrical hearing loss in brainstem lesions. *Acta Otolaryngol* 1973; **75**:165-77.
- Liden G, Engström H, Hall J. Audiological and morphological assessment of effect of noise on cochlear and brainstem in cat. *Acta Otolaryngol (Stockh)* 1973; **75**:325-8.
- Liden G. The scope and application of current audiometric tests. *J Laryngol Otol* 1969; **83**:507-20.
- Carhart R. *Deafness in childhood*. In: Freeman McConnell, Paul Ward, eds. *Vanderbilt University Press*, 1967: 231.
- Dix MR. Observations upon the nerve fibre deafness of multiple sclerosis with particular reference to the phenomenon of loudness recruitment. *J Laryngol Otol* 1965; **79**:695-706.
- Eickel BS, Hedgecock LD, Williams HL. A review of the literature on the audiological aspects of neuro-otological diagnosis: report of five cases in which abnormality and function is associated with brainstem lesions. *Laryngoscope* 1966; **76**:1-29.

- 21 Von Leden H, Horton BT. Auditory nerve in multiple sclerosis. *Arch Otolaryngol* 1948; **48**: 51-7.
- 22 Bentzen O, Jernes K, Thygesen P. Acoustic and vestibular function in multiple sclerosis. *Acta Psychiatr Neurol Scand* 1951; **26**:265-95.
- 23 Simpkins WT. An audiometric profile in multiple sclerosis. *Arch Otolaryngol* 1961; **73**:557-64.
- 24 Antonelli AR, DeMitri T. Reperti Audiometrici Nella Sclerose A Placche. *Sisterna Nervosa* 1963; **15**:138-45.
- 25 Dayal VS, Swisher LP. Pure tone thresholds in multiple sclerosis. A further study. *Laryngoscope* 1967; **77**:2169-77.
- 26 Noffsinger D, Olsen WO, Carhart R, Hart CW, Sahgal V. Auditory and vestibular aberrations in multiple sclerosis. *Acta Otolaryngol* (Suppl) **1972**: 303.
- 27 Conraux C, Collard M. Les troubles cochléaires de la sclérose en plaques. *Rev Otoneuroophthalmol* 1969; **41**:442-3.
- 28 Schuknecht HF. *Pathology of the Ear*. Cambridge, Mass: Harvard University Press, 1974: 341.
- 29 McAlpine D, Compston N, Lumsden C. *Multiple Sclerosis*. Edinburgh: E and S Livingstone Ltd, 1955.
- 30 Citron L, Dix MR, Hallpike CS, Hood JD. A recent clinicopathologic study of cochlear nerve degeneration resulting from tumour pressure with particular reference to the findings of normal threshold sensitivity for pure tones. *Acta Otolaryngol* 1963; **56**:330-7.
- 31 Siebermann F. *Die Blutgefäße in Labyrinth des Menschlichen*. Wiesbaden: Bergmann, 1894.
- 32 Basseres F, Bonnieu A, Guerrier Y. L'exploration audio-vestibulaire. A propos de 48 cas de sténoses de l'artère vertébrale opérées. *Cah otorhinolaryngol* 1974; **9**:463-72.
- 33 Decroix G, Vaneecloo FM, Julliot JP, Quandalle P. Surdit  et insuffisance vertebrobasilaire. *J Fr Otorhinolaryngol* 1975; **24**:503-9.
- 34 Carre J, Huber B. A propos d'une surdit  par troubles vasculaires. *J Fr Otorhinolaryngol* 1975; **24**:621.
- 35 Horrax G. Differential diagnosis of tumours; primarily pineal, primarily pontine. *Arch Neurol Psychiatr* 1927; **17**:179-92.
- 36 Nysten CO. Otoneurological diagnosis of tumours of the brain. *Acta Otolaryngol* (Suppl) 1939; **33**: 1-151.
- 37 Sloan P, Persky A, Saltzman M. Midbrain deafness: tumour of the mid-brain producing sudden complete deafness. *Arch Neurol Psychiatr* 1943; **49**:237-43.
- 38 White HH. Brainstem tumours occurring in adults. *Neurol (Minneapolis)* 1963; **13**:292-300.
- 39 Shapiro I. Progression of auditory signs in pontine glioma. *Laryngoscope* 1969; **79**:201-12.
- 40 Korsan-Bengsten M. In: Rojskaer C, ed. *Audiometric patterns in diseases of the acoustic nerve, brainstem and cortex*. Speech Audiometry, 2nd Danavox Symposium, 1970: 184-90.