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A clinical, genetic and audiological study of patients and families with unilateral vestibular schwannomas. II. Audiological findings in 93 patients with unilateral vestibular schwannomas

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Abstract

Ninety-three patients with histologically or radiologically confirmed unilateral vestibular schwannomas were recruited. Audiological testing for retrocochlear pathology was undertaken. Patients' hospital records were examined for previous audiological and radiological results.

The audiometric configuration was designated as one of the following: normal, sloping, low frequency, peak, trough or flat. A sloping sensorineural audiometric configuration was present in 68 per cent of cases. No significant correlation was found between tumour size and average pure tone threshold 500 Hz to 4000 Hz, optimum discrimination score or interaural differences for wave V. Ninety-one per cent of cases had abnormalities on auditory evoked potential; 92 per cent of cases showed abnormalities on stapedial reflex testing.

The limitations of audiological testing in the investigation of patients with suspected unilateral vestibular schwannomas are discussed. A protocol for the investigation of such patients is presented.

Key words: Vestibular schwannoma; Neurofibromatosis Type 2; Audiology

Introduction

The management of vestibular schwannomas has undergone a number of changes during the past 35 years. The possibility of early diagnosis of tumours of the eighth nerve became feasible with advances in the fields of diagnostic audiology and radiology. The availability of electric response audiometry was a most significant event in the history of diagnostic audiology and neuro-otology. The use of auditory evoked potential (AEP) testing facilitated the screening of patients suspected of being affected with cerebellopontine angle (CPA) lesions (Selters and Brackmann, 1977; Glasscock *et al.*, 1979). The definitive diagnosis of a CPA lesion was made with appropriate radiological imaging techniques. In recent years the use of magnetic resonance imaging (MRI) with gadolinium diethylene-triamine-pentaacetic acid (Gd-DTPA) enhancement has been shown to be a safe, well-tolerated and effective method for the diagnosis of small tumours of the eighth nerve (Stack *et al.*, 1988). The marked improvements in mortality and morbidity, particularly to the facial nerve have not merely resulted from the evolution of microsurgery as an essential skill for the otologist and neurosurgeon, but have mainly been the consequence of the identification of small vestibular schwannomas and early referral to the otological-neurosurgical team.

We have carried out audiological testing for retrocochlear pathology in 91 patients with unilateral vestibular schwannomas, and their first degree relatives. In the case of the patients, the purpose of testing was to demonstrate the type of audiometric configuration in unilateral vestibular schwannomas, and to note which tests were of most assistance in suggesting the possibility of retrocochlear pathology. In patients who had undergone surgery, the purpose was to determine the hearing status in the contralateral ear, and to exclude the possibility of retrocochlear pathology on the contralateral side. In the case of the relatives, the purpose of testing was systematically to screen the relatives for audiological changes suggestive of retrocochlear pathology. In cases where audiological test results were suggestive of retrocochlear pathology, a referral was made for appropriate radiological imaging.

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Materials and methods

The study was carried out between October 1991 and July 1994. Ninety-three patients with histologically or radiologically confirmed unilateral vestibular schwannomas, together with 94 first degree relatives of patients, were recruited. The patients had been under the care of the otological-neurosurgical team at the Manchester Royal Infirmary during the period 1975 to 1994. Ninety-one patients were visited at home, when a genetic pedigree was compiled and a questionnaire was completed. The questionnaire was designed to search for those symptoms and signs which previous workers have described as being associated with neurofibromatosis type 1 (NF1) and neurofibromatosis type 2 (NF2). It was also designed to elicit other causes of deafness, and to compile a full medical and surgical profile on each patient. Two patients were deceased, and medical information was obtained from the Hospital records.

At Manchester, patients and first degree relatives were examined audiologically according to the following protocol.

(1) Routine audiological tests, none of which are invasive or unpleasant for the individual being tested. Those tests which are helpful in indicating the possibility of retrocochlear pathology are marked by * below:

Pure tone audiometry, air conduction and bone conduction thresholds, with masking if necessary; Speech audiometry with closed circuit speech using AB word lists* (Priede and Coles, 1976); Impedance studies; stapedial reflexes* (Chiveralls, 1977; Neary *et al.*, 1993); stapedial reflex decay*; tone decay* (Rosenberg, 1958).

(2) AEP testing*.

Testing was carried out in a sound-treated room, with a constant background noise level of less than 35 dBA.

Pure tone audiometry

Hearing thresholds were measured using recommended techniques (BSA, 1981). Air conduction thresholds were measured at octave intervals 250 Hz to 8000 Hz using standard headphones (TDH 39 in MX 41 AR cushions). Bone conduction thresholds were measured at octave intervals 250 Hz to 4000 Hz with the use of a bone oscillator (Radio Ear B71). Masking, with narrow bands of noise centred around the test frequency presented to the non-test ear, was carried out as necessary. The equipment was subject to regular calibration (BSI, 1969).

Hearing thresholds were considered to be within the normal range if the thresholds were 20 dBHL or less.

As pure tone testing was carried out in 5 dBHL steps, a hearing loss was considered to be present if hearing thresholds of 25 dBHL or more were indicated.

Hearing thresholds were measured at 250 Hz, 500 Hz, 1000 Hz, 2000 Hz, 4000 Hz and 8000 Hz. The mean hearing thresholds for 500 Hz, 1000 Hz, 2000 Hz and 4000 Hz were calculated for each ear separately.

Hearing loss was classified as sensorineural, conductive or mixed sensorineural and conductive.

Hearing loss was considered to be progressive if there was a deterioration of 15 dBHL or more in at least two frequencies, or a deterioration of 10 dBHL or more for an average of four frequencies, 500 Hz to 4000 Hz.

The test frequencies were classified as follows:
Low frequencies, i.e. 250 Hz and 500 Hz.
Mid frequencies, i.e. 1000 Hz and 2000 Hz.
High frequencies, i.e. 4000 Hz and 8000 Hz.
All frequencies, i.e. 250 Hz to 8000 Hz.

The audiograms were designated as one of six different audiometric configurations as follows:

(1) Normal — i.e. Hearing thresholds 20 dBHL or better at octave intervals 250 Hz to 8000 Hz.
(2) Sloping — i.e. ⩾15 dBHL difference between 500 Hz and 4000 Hz.
(3) Low frequency — i.e. 15 dBHL difference from the better to the poorer hearing threshold at the low frequencies to the mid frequencies.
(4) Peak — i.e. ⩾15 dBHL difference between the best hearing threshold and the poorer hearing thresholds (best hearing threshold at highest point on the audiogram).
(5) Trough — i.e. ⩾15 dBHL difference between the poorest hearing threshold and the better hearing thresholds (poorest hearing threshold at lowest point on the audiogram).
(6) Flat — i.e. <15 dBHL difference between 500 Hz and 4000 Hz.

Speech audiometry

Speech audiometry was carried out using isophonemic word lists (AB word lists), employing masking with wide band noise to the contralateral ear as necessary (Boothroyd, 1968). The optimum discrimination score (ODS), the score measured at the intensity level corresponding to the peak of the speech discrimination curve, was recorded. The half peak level (HPL) was estimated as the intensity level at which the speech discrimination would be half of that corresponding to the peak of the curve. The half peak level elevation (HPLE) was thus estimated as the difference between the measured HPL and the normal HPL for the particular test material and equipment. Following the recommendations of Coles *et al.* (1973) a smooth curve was drawn between the data points to outline the most likely shape of the speech discrimination curve. The ODS was plotted against the average of the best two pure tone hearing thresholds, 500 Hz, 1000 Hz and 2000 Hz. Corrections were made in taking the best two pure tone hearing thresholds in the manner defined by Coles *et al.* (1973) as follows: if the thresholds at 4000 Hz was 11–20 dB poorer than the best two average, 1 dB was added; if 21–30 dB poorer, 2 dB was added; if 31–41 dB poorer, 3 dB was added; if more than 40 dB poorer, 4 dB was added. For low tone hearing

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losses, factors of 3 dB or 10 dB were added to the best two average if the threshold at 500 Hz was 11 to 20 dB or 21 to 30 dB, respectively worse than the best two average. On the criteria for cochlear deafness described by Priede and Coles (1976), the cases were designed cochlear, retrocochlear or equivocal. Careful attention was made to the proper use of masking in carrying out speech audiometry. Priede and Coles (1976) had emphasized that if masking of the non-test ear was omitted, or was inadequate, a case with a unilateral hearing loss due to a neural lesion was likely to be misdiagnosed as a cochlear one.

Tone decay

Tone decay was measured at 2000 Hz using Rosenberg's modification of the Carhart technique (Rosenberg, 1958). A neural pattern was considered to be present at levels in excess of 20 dBHL.

AEP testing

AEP testing was carried out using Nicolet Pathfinder 11 apparatus. Alternating clicks were delivered monaurally through standard headphones (TDH 39 in MX 41 AR cushions) at suprathreshold levels of 75 dBnHL to 100 dBnHL. Clicks were presented at four repetition rates, varying from 11.1 clicks/sec to 44.1 clicks/sec, averaging 2048 responses for each repetition rate. Masking noise was applied to the contralateral ear as necessary.

Patients who had not undergone surgery for a unilateral vestibular schwannoma had AEP testing carried out on both sides. Patients who had undergone surgery for a unilateral vestibular schwannoma had AEP testing carried out on the contralateral ear. The AEP traces were labelled according to the classification of Jewett *et al.* (1970). The absolute latencies for waves I, III and V and the central conduction times (wave I to wave III interpeak latency differences and wave I to wave V interpeak latency differences) were compared with the normal range for adults. The diagnostic parameters for the normal range in adults had been determined previously following an 'in house' calibration of the equipment (Vidler, 1987). In the absence of a recognizable wave I, the interaural differences for the absolute latencies for wave V (IT5) were calculated, the possibility of a conductive component contributing to the latency delay having been excluded previously by the demonstration of normal tympanograms. An IT5 of 0.2 m.sec was regarded as suggestive of retrocochlear pathology, and 0.3 m.sec was regarded as 'highly significant' (Selters and Brackmann, 1977; Terkildsen and Thomsen, 1983).

Evaluation of the vestibular system

Evaluation of the vestibular system with bithermal caloric testing was considered, but rejected. Caloric testing is a traditional screening procedure for vestibular schwannoma suspects, with an abnormal response being reported in 82 per cent. However, in the case of small tumours, the caloric response has been reported to be reduced in slightly fewer than 50 per cent (Linthicum and Churchill, 1968).

Radiological imaging

In patients where the clinical or audiological findings in the contralateral ear were suggestive of retrocochlear pathology, and in patients where wave forms could not be identified on AEP traces of the contralateral ear, a referral was made to the Department of Diagnostic Radiology. In relatives where the findings on audiological testing were suggestive of retrocochlear pathology, or where wave forms could not be identified on AEP traces, a referral was made for appropriate radiological imaging. MRI studies were performed on a Picker International 0.26 Telsa super-conducting magnetic system using a 30 cm field of view head receiver coil, before and after administration of Gd-DTPA (0.1 m mol/Kg).

Results

The diagnosis of vestibular schwannoma had been histologically confirmed in 80 of the 93 patients. The diagnosis had been radiologically indicated in a further six patients, who were awaiting surgery. In seven patients the tumour was being monitored by means of serial scans, because of the patients' age, or contra-indications to surgery.

Of the 93 patients, 42 were male and 51 were female. The schwannoma was right-sided in 45 cases and left-sided in 48 cases.

The mean age of onset of symptoms in 93 patients with unilateral vestibular schwannomas was 44.42 years (range 14 years to 73 years). The mean age of diagnosis was 49.74 years (range 15 years to 78 years). The age of onset of symptoms in different age groups of patients affected with unilateral vestibular schwannomas is shown in Figure 1.

The earliest symptoms in 93 patients with unilateral vestibular schwannomas were reviewed. Hearing loss and tinnitus predominated as the earliest symptoms. In 44 patients (48 per cent) the earliest symptom was a unilateral hearing loss, in 14 patients (15 per cent) tinnitus, and in 10 patients (11 per cent), a unilateral hearing loss and tinnitus. At the time of diagnosis 92 patients were affected with a hearing loss. One patient was hearing normally. Eighty-five patients (91 per cent) gave a history of a gradually progressive hearing loss. Seven patients (eight per cent) had noticed sudden onset of a hearing loss.

Tumour size

The size of the tumours in 93 patients with unilateral vestibular schwannomas, diagnosed during the period 1975 to 1994 were reviewed. The tumours were assigned to five groups according to the classification proposed by Tos and Thomsen (1992):

Intrameatal (no size)			
Extrameatal size	0.1–1.0 cm	. . .	small
	1.1–2.5 cm	. . .	medium
	2.6–4.0 cm	. . .	large
	>4.0 cm	. . .	extra large

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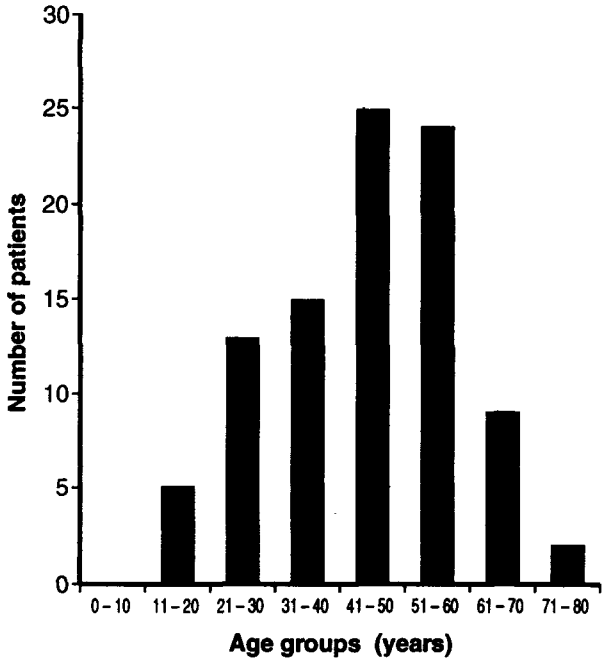

FIG. 1

Age in years of onset of earliest symptom(s) in 93 patients with unilateral vestibular schwannomas.

The number of tumours in the groups classified according to size was noted for successive five year periods. Intrameatal tumours were grouped together with tumours of extrameatal size 0.1–1.0 cm in Figure 2. Among the patients diagnosed during the period 1975 to 1979, the proportion of patients with extra large or large tumours predominated, and no patients with small tumours were represented. During the period 1980 to 1984, the proportion of patients with medium sized tumours predominated, and again no patients with small tumours were represented. During the periods 1985 to 1994, patients with intrameatal and small tumours were increasingly represented, although the proportion of patients with medium-sized tumours still predominated.

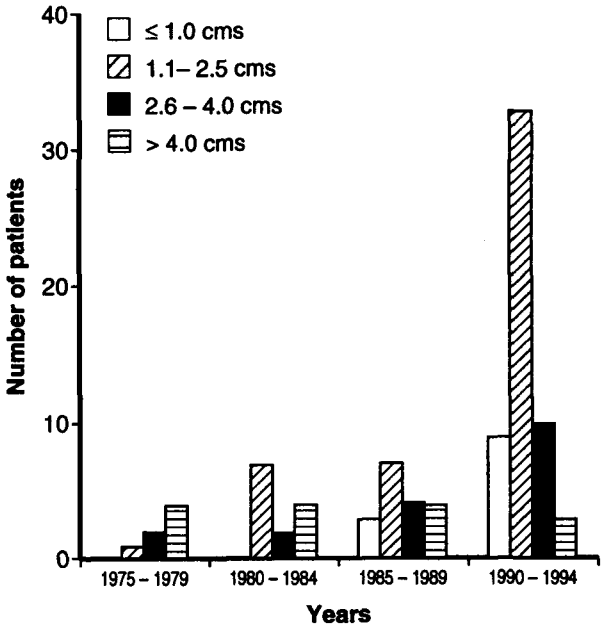

FIG. 2

Tumour size in 93 patients with unilateral vestibular schwannomas diagnosed during period 1975 to 1994.

Audiological findings in 93 patients with unilateral vestibular schwannomas

In those patients who had undergone surgery for a unilateral vestibular schwannoma, the audiological results at the time of diagnosis were obtained from the Hospital records.

Pure tone audiometry on the affected side

Seventy-eight patients with unilateral vestibular schwannomas were affected with a unilateral sensorineural hearing loss at the time of diagnosis. One patient had normal hearing on the affected side. The audiometric configurations are presented in Table I. Eleven patients had no response at the limits of the audiometer on the affected side at the time of diagnosis. The results of pure tone audiometry at the time of diagnosis were not available in three cases.

A sloping configuration was seen in 68 per cent of cases. In nine per cent the audiogram was trough shaped. In a further nine per cent of cases the audiogram was flat; eight per cent had a low-frequency configuration. Five per cent of cases had a peak shaped configuration. One per cent of cases had a normal configuration.

Consideration was given to the possibility of a relationship between the average pure tone hearing threshold, 500 Hz to 4000 Hz, and the size of the tumour in 79 cases of unilateral vestibular schwannomas. Extrameatal tumour size was plotted on the horizontal axis. Intrameatal tumours were considered to have no size, and were included in the scattergram (Figure 3).

There was no significant correlation (Spearman rank correlation: correlation coefficient, $r_s = 0.0008$) between the average pure tone hearing threshold and tumour size. It was noted that one patient with an extra large tumour had normal hearing, and several patients with intrameatal and small tumours had significant hearing losses.

Pure tone audiometry on the contralateral ear

Normal thresholds for hearing at octave intervals 250 Hz to 8000 Hz were indicated on testing the contralateral ear in 29 individuals.

In 64 individuals, testing of the contralateral ear indicated a sensorineural hearing loss. The results of

TABLE I

THE CONFIGURATION OF THE AUDIOGRAM ON THE AFFECTED SIDE IN 79 PATIENTS WITH UNILATERAL VESTIBULAR SCHWANNOMAS

Audiometric configuration	Number of cases	Percentage of cases
Sloping	54	68
Trough	7	9
Flat	7	9
Low frequency	6	8
Peak	4	5
Normal	1	1

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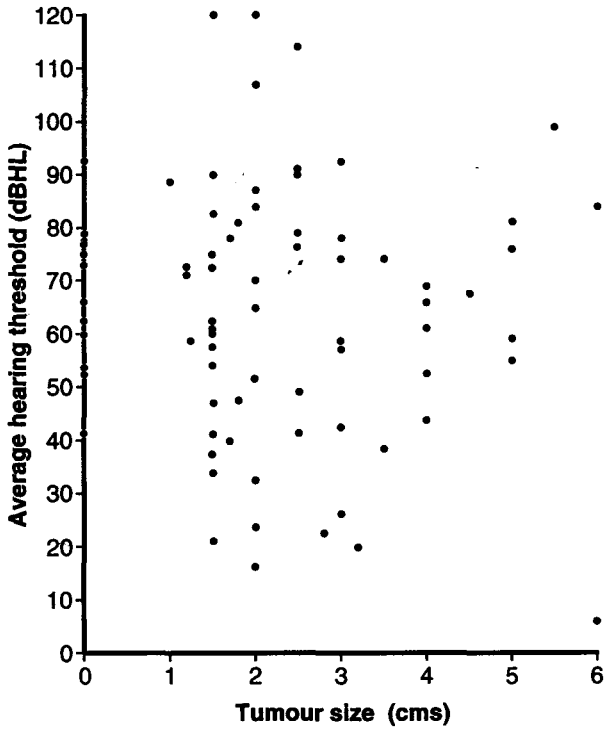

FIG. 3

Average pure tone hearing threshold (500 Hz to 4000 Hz) of affected ears in 79 patients with unilateral vestibular schwannomas plotted against tumour size.

pure tone audiometry obtained during the time of the study were compared with the results obtained at the time of the diagnosis, to determine whether significant progression of the hearing loss had occurred.

Speech audiometry on the affected side

Speech audiometry results were examined in 51 cases. ODS was plotted as a function of hearing sensitivity, measured as the average of the best two pure tone hearing thresholds 500 Hz, 1000 Hz and 2000 Hz, using the corrections for high-tone hearing loss specified by Coles *et al.* (1973) (Figure 4). The

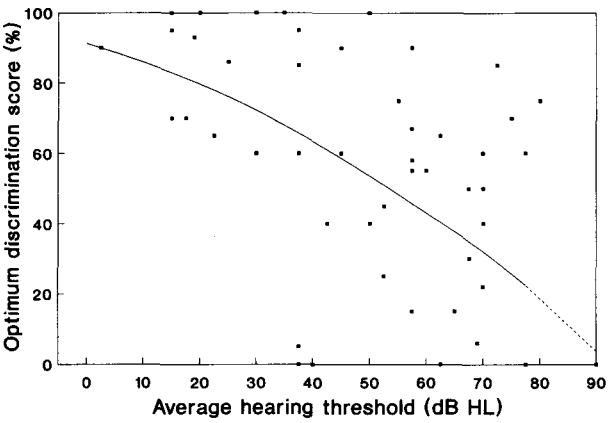

FIG. 4

Optimum discrimination score as a function of hearing sensitivity in 51 patients with unilateral vestibular schwannomas.

solid curve represented the criterion which divides primarily cochlear hearing loss (above line) from primarily retrocochlear hearing loss (below line). The data for the criterion in the publication of Coles *et al.* (1973) referred to the use of Fry's PB word lists. The data for the criterion with the use of AB(s) word lists was specified in the publication of Priede and Coles (1976). The criterion for AB(s) word lists used in the present study was redrawn from the work carried out at Southampton, with the permission of Coles. Cases were designated cochlear, retrocochlear or equivocal.

In a group of 51 proven unilateral vestibular schwannomas, the presence of 47 per cent of the tumours was indicated on the results of speech audiometry. Fifty-one per cent of tumours gave negative findings on speech audiometry. Two per cent of tumours were associated with equivocal results on speech audiometry.

Consideration was given to the possibility of a relationship between the ODS and the size of the tumour in 57 patients with unilateral vestibular schwannomas. Extrameatal tumour size was plotted on the horizontal axis. Intrameatal tumours were considered to have no size, and were included in the scattergram (Figure 5). There was no significant correlation (Spearman rank correlation; correlation coefficient $r_s = 0.2328$) between the ODS and the size of the tumour.

Speech audiometry on the contralateral side

Normal speech audiograms were obtained in the 29 individuals with normal hearing on the contralateral side.

Speech audiometry was carried out on 62 of the 64 individuals who were affected with a sensorineural hearing loss in the contralateral ear. All 62 speech audiograms had a cochlear pattern.

Stapedial reflexes

The results for stapedial reflex testing were available for 51 patients with unilateral vestibular schwannomas.

Ninety-two per cent of patients with unilateral vestibular schwannomas had abnormalities on testing stapedial reflex thresholds, 39 per cent having significant asymmetry of the stapedial reflexes and 53 per cent having absent stapedial reflexes. Normal stapedial reflex thresholds were seen in eight per cent of this group.

The results for testing for pathological stapedial reflex decay were available for 16 patients. Sixty-three per cent of patients with unilateral vestibular schwannomas showed pathological stapedial reflex decay at 1000 Hz.

Testing of ipsilateral stapedial reflexes in the unaffected ear in patients who had undergone surgery

In 45 of 80 patients, who had undergone surgery, normal results were obtained on testing ipsilateral stapedial reflexes. In nine patients ipsilateral stapedial reflexes were absent. No results for ipsilateral stapedial reflexes were obtained in 26 patients,

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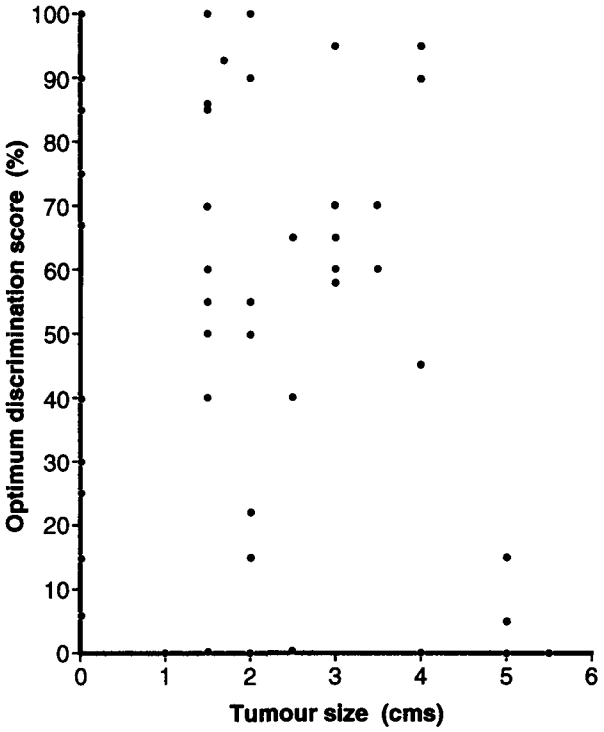

FIG. 5

Optimum discrimination score plotted against tumour size in 57 patients with unilateral vestibular schwannomas.

because of difficulties with obtaining a seal with the probe tip.

Tone decay

The results for testing of tone decay were available for 41 patients with unilateral vestibular schwannomas. Tone decay was found in 13 patients, 32 per cent of those tested.

Testing for tone decay in the contralateral ear.

There was no evidence of tone decay on testing the contralateral ear of 79 patients affected with a unilateral vestibular schwannoma. Sixty of these patients were affected with a sensorineural hearing loss in the contralateral ear. Tone decay was seen in four patients affected with a sensorineural hearing loss in the contralateral ear.

AEP testing

Ninety-one per cent of patients with unilateral vestibular schwannomas were found to have abnormalities on AEP testing. Fifty-eight per cent of patients had an IT5 greater than 0.2 m.sec. In 33 per cent of patients, no recognizable wave forms were recorded. Two patients with intrameatal tumours, and one patient with a small tumour had normal results on AEP testing. One patient with a tumour measuring 1.5 cm had normal results on AEP testing. Consideration was given to the possibility of a relationship between IT5 and the size of the tumour in 14 patients. The intrameatal measurement was included in the tumour size (Figure 6). There was no significant correlation (Spearman rank correlation; correlation coefficient $r_s = 0.4872$) between the IT5 and the size of the tumour.

AEP testing of the contralateral ear

AEP testing was carried out on the contralateral ear of 54 patients who had undergone surgery. There was no evidence of retrocochlear pathology in any patient.

Relatives

Audiological testing for retrocochlear pathology, followed by appropriate radiological imaging where indicated, was carried out on 92 first degree relatives of patients with unilateral vestibular schwannomas. There was no evidence of vestibular schwannomas in 87 relatives. Five relatives were referred for appropriate radiological imaging, because of the presence of abnormalities suggestive of the possibility of retrocochlear pathology, or inconclusive results on audiological testing. There was no evidence of vestibular schwannomas on radiological imaging (four MRI with Gd-DTPA and one CT scan).

The information that two relatives of patients were also affected with unilateral CPA lesions was obtained on taking the family history. These two relatives were not examined personally.

Sensitivity and specificity of audiological tests in screening for retrocochlear pathology

The sensitivity of audiological tests in screening for retrocochlear pathology in patients with unilateral vestibular schwannomas was calculated. For various reasons not all the patients had undergone all the audiological test procedures. Eleven patients had presented with total hearing loss and were unsuitable for audiological testing. The diagnosis in these patients was made with appropriate radiological imaging. Specificity was calculated as the

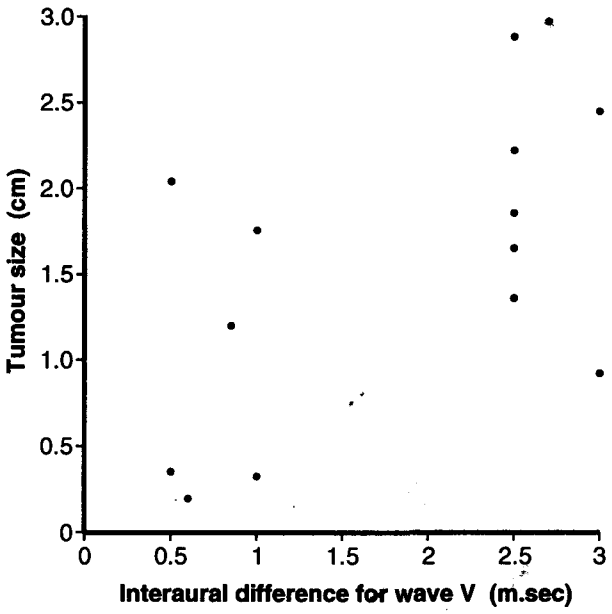

FIG. 6

Tumour size plotted against interaural difference for wave V in 14 patients with unilateral vestibular schwannomas.

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TABLE II
SENSITIVITY AND SPECIFICITY OF AUDIOLOGICAL TESTS IN SCREENING FOR RETROCOCHLEAR PATHOLOGY

Audiological test	No. (1)	No. (2)	S, % (3)	S, % (4)	Pos. % (5)	Eff. % (6)
SRT	51	81	92	96	94	95
AEP testing	43	82	91	92	85	91
SRD	16	73	63	100	100	93
Speech audiometry	51	92	47	100	100	81
Tone decay	41	92	32	99	93	78

Key to Table II:
SRT = stapedial reflex threshold testing (includes significant asymmetry of the stapedial reflexes, and absent stapedial reflexes).
AEP testing = auditory evoked potential testing.
SRD = stapedial reflex decay.
(1) = Number of patients with unilateral vestibular schwannomas tested.
(2) = Number of relatives tested.
(3) = Sensitivity.
(4) = Specificity.
(5) = Positive predictive value.
(6) = Efficiency.

percentage of non-tumour relatives who gave normal test results. For completeness, positive predictive value and efficiency of the audiological tests were also calculated. The derived sensitivity and specificity values, positive predictive value and efficiency are presented in Table II.

Discussion

The mean age of onset of the first symptoms of unilateral vestibular schwannomas in the present study, 44.42 years is in line with that reported in previous series. Pool and Pava (1957) reported that the mean age of onset of first symptoms and the mean age of diagnosis of patients with unilateral vestibular schwannomas was between the fourth and sixth decade. Revilla (1947) calculated that an average of 4.5 years elapsed between the mean age of onset of symptoms and the time of admission to hospital. In the present study, an average of 5.32 years had elapsed between the mean age of onset of symptoms and the mean age at diagnosis.

Consideration of the earliest symptoms of unilateral vestibular schwannomas in this study indicated that progressive hearing loss and tinnitus predominated, in accordance with the findings reported in other series (Ojemann *et al.*, 1972; Ellis and Wright, 1974; Thomsen, 1976; Hart and Davenport, 1981; and Guyot *et al.*, 1992). Ninety-two per cent of hearing impaired patients in the present study gave a history of a progressive hearing loss at the time of diagnosis. Eight per cent of patients reported a sudden hearing loss. These findings are in accordance with the observations of Hirsch and Anderson (1980), who found that 7.3 per cent of patients presented with a sudden hearing impairment, and Thomsen and Tos (1988), who reported that seven per cent of their series presented with a sudden loss of hearing.

Consideration of the sizes of the tumours diagnosed during the period 1975 to 1994 indicated the larger proportion of intrameatal, small and medium-sized vestibular schwannomas diagnosed during the last decade. These figures partly reflect the increased awareness of clinicians regarding the possibility of the presence of a vestibular schwannoma, the advances in diagnostic audiology, and the advent of sensitive radiological imaging techniques, in particular MRI with Gd-DTPA enhancement.

On consideration of the audiological test results at the time of diagnosis of the unilateral vestibular schwannomas in the present study, abnormalities of the stapedial reflex thresholds and the rate of stapedial reflex decay together with abnormalities on AEP testing, were the most valuable pointers to the presence of retrocochlear pathology.

A sloping sensorineural pattern predominated as the audiometric configuration on pure tone testing. However, pure tone audiometry was not of assistance in the differentiation of cochlear from retrocochlear pathology. Furthermore, there was no significant correlation between the degree of the hearing loss, and the size of the tumour.

A retrocochlear pattern of speech audiometry was indicated in 47 per cent of cases with unilateral vestibular schwannomas in the present study. This figure was similar to the figure of 45 per cent reported by Hirsch and Anderson (1980) for the results of speech audiometry in a series of 97 proven tumours of the eighth nerve. There was no significant correlation between the ODS and the size of the tumour in the present study.

Testing of tone decay in the present study yielded poor results. A retrocochlear pattern was indicated in only 32 per cent of patients. Hirsch and Anderson (1980) reported a retrocochlear pattern on tone decay testing in 53 per cent of patients in their series.

Ninety-two per cent of patients with unilateral vestibular schwannomas showed abnormalities on testing stapedial reflex thresholds in the present study. Sixty-three per cent showed pathological stapedial reflex decay. The value of the combined test of stapedial reflex threshold and decay has been confirmed by many studies (King *et al.*, 1976; Chiveralls, 1977). However, the stapedial reflex is abolished by a cochlear loss greater than 60–75 dBHL. Furthermore, it is not possible to carry out measurements of the stapedial reflexes in the presence of middle ear fluid.

Ninety-one per cent of patients in the present study had abnormalities on AEP testing. Selters and Brackmann (1977) reported a sensitivity of 96 per cent for AEP testing, in the diagnosis of vestibular schwannomas. They reported that the main limitation of AEP testing related to patients with hearing losses greater than about 75 dBHL. They reported that these losses produced a higher rate of false-positive findings, either no response or a delayed response in the case of severe high frequency loss. In the present study two patients with intrameatal tumours, one with a small tumour, and one with a tumour measuring 1.5 cm had normal results on AEP testing. Selters and Brackmann (1977) had suggested that the larger the tumour, the greater the pressure on the nerve, and the longer the delay of the response on AEP testing. They reported that for 18 vestibular schwannomas in their series, the IT5 was approximately proportional to the size of the tumour. In the present study there was no significant correlation between the IT5 and the size of the tumour.

Five relatives of patients were referred for appropriate radiological imaging, because of the presence of abnormalities suggestive of the possibility of retrocochlear pathology, or inconclusive test results on audiological testing. The results of imaging did not reveal any evidence of vestibular schwannomas. Thomsen and Tos (1992) emphasized the fact that AEP testing tests the function of the auditory system rather than being specific for vestibular schwannomas. They reported that it could be expected that many positive tests would be seen without a schwannoma being the cause. These tests would not be false positives in the proper sense, but would represent correct testing of a variety of diseases giving rise to a retrocochlear hearing impairment. Thomsen and Tos emphasized the necessity of cases with abnormal results on AEP testing, or cases with hearing worse than 70–80 dBHL, being referred on for appropriate radiological imaging.

Ramsden and Moffat (1994) have suggested the factors to be considered in deciding upon a treatment policy for an individual patient, now that MRI has provided the means of imaging vestibular schwannomas when the tumours are tiny. They proposed that the following factors should receive consideration: tumour growth characteristics; age of patient; general health of patient; the hearing level in the tumour ear; the hearing level in the contralateral ear; the risks of death or of serious morbidity from surgery; the cost in financial and human terms of prolonged follow-up.

Ramsden and Moffat concluded that the correct treatment for the majority of small vestibular schwannomas is prompt removal by an experienced neuro-otologist. They have suggested that neuro-otologists should press for the routine use of MRI as a first line investigation of unilateral audiovestibular failure of unknown aetiology.

Conclusions

As a result of this study, it is suggested that the following protocol should be undertaken for the evaluation of a patient with a suspected vestibular schwannoma. Initially a thorough history should be taken, including a detailed family history regarding other close family members affected with vestibular schwannomas, and other tumours of the central nervous system. The physical examination should include a careful examination for any signs of NF2. It should include the examination of the cranial nerves, of the skin for café-au-lait macules and peripheral neurofibromas, and a slit-lamp examination of the lenses through dilated pupils, for posterior subcapsular and cortical opacities.

In centres with ready access to MRI, it is recommended that the optimal approach towards achieving an early diagnosis of a vestibular schwannoma is the prompt referral for MRI with Gd-DTPA enhancement of all patients with an undiagnosed unilateral audiovestibular failure. MRI with Gd-DTPA enhancement permits virtually certain diagnosis, without the risk of irradiation, of intrameatal vestibular schwannomas 0.2 to 0.3 cm in size. Both IAC may be visualized on MRI, facilitating the diagnosis of bilateral vestibular schwannomas (NF2), as opposed to a unilateral vestibular schwannoma. MRI has also made possible the ability to diagnose with ease the small tumours in the brain and spinal cord, which are associated with NF2. In Manchester all patients attending the otology clinic have pure tone and speech audiometry, and stapedial reflex studies carried out on arrival. However, the routine use of other conventional audiological tests for retrocochlear pathology, and AEP testing, has been discontinued, patients being referred for MRI with Gd-DTPA enhancement at the earliest possible stage. The financial implications of the Manchester approach may be questioned in discussions regarding the allocation of resources for health care. It should be emphasized that with the increasing use of MRI, the difference in cost between MRI and a full battery of audiovestibular tests including AEP testing is not presently as great as was the case at the introduction of MRI (Saeed *et al.*, 1995). The financial savings to society from the resultant early diagnosis and decreased operative morbidity should be given careful consideration. Furthermore, it may also be relevant to consider the medicolegal responsibility which otologists assume by failing to employ the most sensitive tool at their disposal.

There are still many centres where facilities for MRI are not readily available. In these centres reliance will continue to be placed on an audiological evaluation. Such an evaluation should include an audiological test battery with the use of pure tone and speech audiometry, testing of stapedial reflexes and tone decay, and AEP testing, to select patients for CT scanning. In patients with hearing thresholds better than 60–75 dBHL, the importance of testing stapedial reflex thresholds and AEP testing in screening for retrocochlear pathology must be underlined. In the present study sensitivity and specificity values of 92 per cent and 96 per cent respectively were calculated for testing of the stapedial reflex thresholds. The calculated sensitivity

and specificity values for AEP testing were 91 per cent and 92 per cent respectively. It must be emphasized that a small number of patients with negative results for retrocochlear pathology on stapedial reflex threshold testing and AEP testing may be affected with a vestibular schwannoma. Furthermore, false negative results may be obtained with post contrast CT in the case of a vestibular schwannoma less than 10 mm in size.

Further management of the patient diagnosed as being affected with a unilateral vestibular schwannoma should be undertaken by an experienced neuro-otologist. Patients and families with NF2, however, present many complex considerations. It is suggested that a team, including specialists from the fields of otolaryngology, neurosurgery, audiological medicine, radiology and clinical genetics should be available for the management and treatment of such individuals.

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