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Audio-vestibular manifestations of Chiari malformation and outcome of surgical decompression: A case report

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Abstract
Sensorineural hearing loss, tinnitus, dizziness and ataxia are recognised symptoms associated with Chiari malformations but they are rarely the presenting complaints. Patients with such symptoms are frequently referred to otolaryngologists and audiological physicians. We report a case of a 13-year-old girl who presented complaining of tinnitus and impaired hearing, and was subsequently diagnosed as having a type I Chiari malformation. Pure tone audiogram showed a mild hearing impairment on the left side and the speech audiogram was normal. Auditory brain stem responses and the electronystagmography were abnormal. The patient underwent posterior fossa decompression following which her tinnitus disappeared, the hearing problem recovered and some of the abnormal electrophysiological parameters were corrected.

Key words: Arnold-Chiari deformity; Tinnitus; Hearing loss, sensorineural; dizziness

Introduction

John Cleland in 1883 first described a rare congenital anomaly of the brain stem where the medulla was elongated, the fourth ventricle extended into the cervical canal, and the inferior vermis distorted caudally. In 1891 Chiari described two main types of brain stem malformations; in the type I only the cerebellar tonsils protruded into the cervical canal without any involvement of the medulla, but in the type II there was a displacement of the lower part of the vermis, pons and medulla oblongata with elongation of the fourth ventricle into the cervical canal. Arnold's name was added to the type II malformation in 1907 by Schwalbe and Gredig and the type II Chiari anomaly of the brain-stem became to be known as Arnold-Chiari malformation (as cited in Carmel and Markesbery, 1972).

Chiari malformations are associated with audio-vestibular symptoms but such symptoms are rarely the presenting complaint and were first reported by Rydell and Pulec (1971). Hendrix *et al.* (1992) reviewed 226 consecutive patients with asymmetrical sensorineural hearing loss and found three patients with Chiari type I malformation out of 32 patients with retrocochlear pathology. Compression of the vestibular and cochlear nuclei by the herniated cerebellar tonsils, stretching of the eighth nerve and ischaemia caused by the distortion of the posterior inferior cerebellar artery or one of its branches (Rydell and Pulec, 1971; Bertrand *et al.*, 1973) have been suggested to account for the otoneurological features of the malformations. The audio-vestibular symptoms in Chiari malformations are given in Table I. We present the case of a 13-year-old girl with the type I malformation who presented with tinnitus and hearing impairment.

Case report

A 13-year-old girl was referred to the Audiological Medicine department with a six-month history of a buzzing noise in the left ear. The tinnitus was non-pulsatile and intermittent lasting several minutes at a time and occurring several times a day. It seemed that the tinnitus occurred

TABLE I
THE AUDIO-VESTIBULAR SYMPTOMS IN CHIARI MALFORMATIONS

Authors	Rydell and Pulec (1971)	Mohr *et al.* (1977)	Chait and Barber (1979)	Paul *et al.* (1983)
Number of patients with Chiari malformation studied:	130	40	5*	71
Number of patients with audio-vestibular symptoms:	29	not quoted	5*	not quoted**
Deafness:	13	2	1	2
Tinnitus:	not quoted**	1	1	5
Dizziness:	not quoted**	0	5*	2

*Chait and Barber found five cases of Chiari malformation amongst patients attending a "Dizzy unit".
**Exact figure not cited in the papers, and some of the patients had more than one audio-vestibular symptom.

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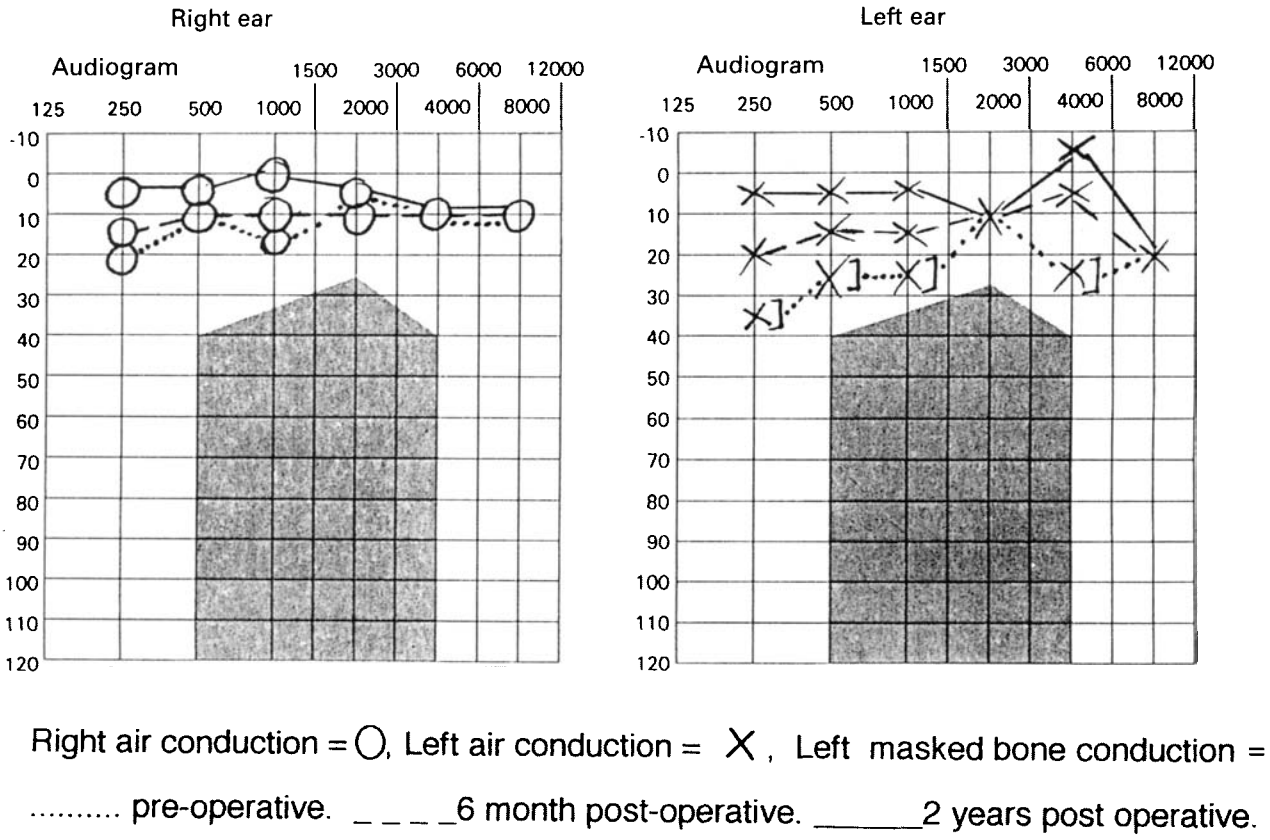

FIG. 1
Pure tone audiogram of the patient.

mostly during lessons at school, French lessons in particular, and while playing with other children at school or with her brothers or sisters at home. On a few occasions she thought that she had woken up from sleep due to the noise in her left ear. She also complained of impaired hearing both at school and at home in the presence of background noise. She was found to turn the television louder and also asked her mother to speak more loudly. There was no history of ear ache or discharge, head injury nor surgery to the ears. She never complained of dizziness or unsteadiness. The perinatal history was uneventful and the developmental milestones were normal. She had suffered from measles, mumps and German measles in early childhood but had not had any significant illness (e.g. meningitis) and had never had any ototoxic medications. She comes from a big family, with eight children in all, and the father was mostly away on business except on weekends. Her teachers had felt that she was attention seeking; and the referring physician felt that there could be a psychogenic element to the problem.

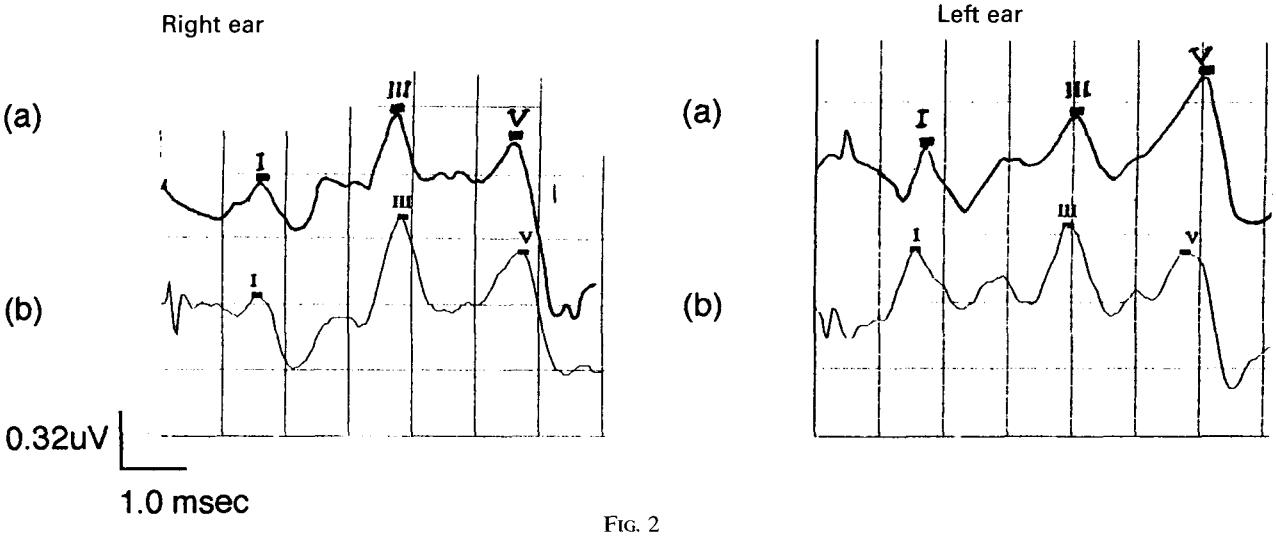

FIG. 2
Auditory brainstem responses, a) pre-operative, b) post-operative

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TABLE II
LATENCY VALUES OF AUDITORY BRAINSTEM RESPONSE

Waves	Pre-operative latencies Left	Pre-operative latencies Right	Post-operative latencies Left	Post-operative latencies Right	Normative data 95% C.I.
I	1.72	1.60	1.56	1.56	1.35–1.81
III	3.96	3.80	3.94	3.84	3.52–3.93
V	5.92	5.52	5.82	5.74	5.35–5.75
I–III	2.24	2.20	2.38	2.28	1.87–2.47
III–V	1.96	1.72	1.88	1.90	1.57–2.07
I–V	4.20	3.92	4.26	4.18	3.77–4.27

During the consultation she was able to carry out conversation at normal voice level without any difficulty. The physical examination of the ear, nose and throat did not reveal any abnormality. The Romberg and Unterberger tests were normal and there was no clinical evidence of abnormal cerebellar function. Clinically the cranial nerves were all intact and there was no spontaneous nystagmus. The pure tone audiogram (see Figure 1) showed a pure tone average of 10 dB HL on the right and 21 dB HL on the left at the frequencies of 0.5, 1, 2 and 4 kHz respectively for both air and bone conduction. The speech audiometry using Arthur Boothroyd word list corresponded well to the pure tone audiometric findings. In view of the relative asymmetry of pure tone thresholds auditory brain stem responses (ABR) were carried out.

A Nicolet Spirit evoked potential system was used which delivered unfiltered alternating rarefaction and condensation clicks, with a click duration of 100 μs, at a rate of 11.1/sec, through TDH-49 ear phones. The intensity of the stimuli was 80 dB nHL in the test ear with a masking noise of 40 dB nHL in the contralateral ear. The amplifier band pass filter was 100–3000 Hz and the analysis time was 10 ms from presentation of stimulus. For each ear a sum of three averages, each of 1000 sweeps, was obtained.

The result showed that the latencies for waves III and V on the left were outside the 95 per cent confidence interval of the normative data used in the department. The wave forms (Figure 2 and Table II) indicated that the I–V wave inter-peak latency was longer on the left ear by 0.28 milliseconds compared to the right, a value of 0.2 ms being considered in this department to be the normal maximum limit for the interaural I–V wave inter-peak latency difference. The latency figures for the wave forms are given in Table II. The increase in the I–V wave inter-peak latency of the left ear appears to originate between the peaks of waves III and V indicating a brain stem dysfunction as the possible source of the delay. In view of the large inter-aural I–V wave inter-peak latency difference a magnetic resonance image (MRI) scan was arranged which showed features suggestive of Chiari malformation. Figure 3 shows the abnormal MRI scan demonstrating the protrusion of the cerebellar tonsil through a distorted foramen magnum. In the scan the anterior margin of the foramen magnum is in a high position due basilar invagination. Figure 4 shows the normal foramen magnum with the normal position for the cerebellar tonsil.

Electronystagmography (ENG) was carried out and only the horizontal component of the movements of both the eyes were recorded using bitemporal electrodes. A horizontal nystagmus alternating direction at regular intervals- 'periodic alternating nystagmus' was found without eye fixation in the dark. Saccade, smooth pursuit and optokinetic nystagmus were not tested. No asymmetry was noted between the ears to irrigation with water at 30°C and 44°C in the bi-thermal water caloric test and the caloric nystagmus was found to be suppressed by optic fixation.

Finally the patient was referred to the neurosurgical department. Surgery was carried out and the cervicomedullary junction was exposed via a wide suboccipital craniectomy combined with removal of the arch of the atlas and the laminae and spinous process of the C2 vertebra. Adhesion and a tight band of dura around the cervicomedullary junction were divided to expose the cerebellar tonsils. These were found to be displaced caudally to the level of the upper border of the C2 laminae and the cervicomedullary junction was almost at right angles. A fascial graft was interposed between the edges of the dural opening to provide decompression. Post-operatively the patient made a rapid recovery with

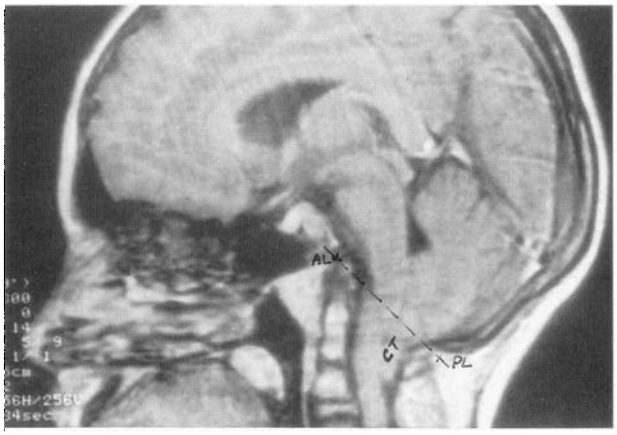

FIG. 3
Sagittal cut MRI scan showing the protrusion of the cerebellar tonsil (CT) below the line drawn through the anterior lip (AL) and the posterior lip (PL) of the foramen magnum. The position of the anterior lip in this patient is high compared to the normal scan in Figure 4 due to basilar invagination.

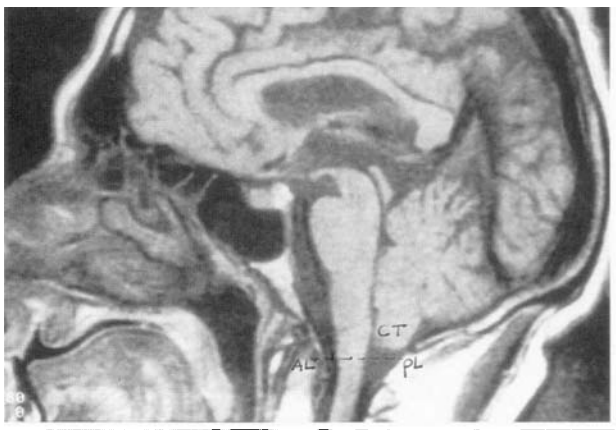

FIG. 4
Sagittal cut MRI scan showing the normal position of the cerebellum and lower brain stem. The cerebellar tonsil is above the line joining the anterior lip (AL) and the posterior lip (PL) of the foramen magnum.

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complete relief of her tinnitus and her hearing impairment.

The relief of her auditory symptoms was maintained, at the time of writing this paper, two years after the surgical decompression. Repeat pure tone audiogram post-operatively showed an improvement in pure tone average of the left ear at the frequencies of 0.5, 1, 2, and 4 kHz from 21 dBHL to 11 dBHL after six months and to 4 dB HL two years after the surgery. There were no significant changes noted in the right ear. Repeat ABR two years later by the same consultant audiological scientist using the same protocol and the same system showed the inter-aural I–V wave inter-peak latency difference returning to within the normal limits accepted by the department. Electronystagmography repeated one year after the surgery showed no evidence of the periodic alternating nystagmus noted in the pre-operative test.

Discussion

Although audio-vestibular symptoms are uncommon presenting features of Chiari malformations, detailed audio-vestibular investigation is indicated to assist in the early diagnosis of such cases so that appropriate surgical management can be carried out sooner.

Post-operative recovery of pre-operative symptoms has been reported by Paul *et al.* (1983) in their patients but there is no mention of the improvement in the hearing loss or tinnitus. Chait and Barber (1979) reported a case of unilateral sensorineural hearing loss with tinnitus and dizziness where surgical decompression relieved the dizziness and the hearing loss was unchanged but no note was made about the tinnitus. Rydell and Pulec (1971) documented the post-operative hearing status in three patients and found that hearing was unchanged in one, a partial improvement in one and a deterioration in one. A partial improvement of a unilateral hearing loss has been reported in a patient by Albers and Ingels (1993). In our patient both the tinnitus and hearing loss recovered completely soon after surgery and this was confirmed by normal and symmetrical thresholds on both sides on pure tone audiograms at six months and two years after the surgery.

Otoneurologist, otolaryngologist, audiological physicians and others dealing with patients with hearing impairment, tinnitus and dizziness, either on its own or in any other combination, should be aware of Chiari malformations as a differential diagnosis. Our case has demonstrated the justification of investigating such patients thoroughly. Even though an MRI scan is essential to establish the diagnosis of a Chiari malformation to be confirmed at surgery, audio-vestibular tests such as pure tone audiogram, speech audiogram, electronystagmography, caloric test and auditory brain stem responses are important initial investigations in those who present with audio-vestibular symptoms.

Review of the literature shows that apart from periodic alternating nystagmus (Barber and Stockwell, 1980), which was documented in our patient in the pre-operative stage, various other types of nystagmus are associated with Chiari malformations. These included: up-beat nystagmus (Rydell and Pulec, 1971), down-beat nystagmus (Cogan, 1968; Bertrand *et al.*, 1973; Chait and Barber, 1979; Faria *et al.*, 1980; Yee *et al.*, 1984; Bronstein *et al.*, 1987; Yeow and Tjia, 1989), bilateral gaze nystagmus, rebound nystagmus, asymmetric optokinetic nystagmus (Longridge and Mallinson, 1985), see-saw nystagmus (Zimmerman *et al.*, 1986), torsional nystagmus (Bronstein *et al.*, 1987), convergence nystagmus (Mossman *et al.*, 1990; Camarda *et al.*, 1991) and rebound caloric nystagmus (Kumar *et al.*, 1993). Internuclear ophthalmoplegia has also been reported (Arnold *et al.*, 1990). However down-beating vertical nystagmus on lateral gaze indicating a lower medullary or cervicomedullary lesion is more characteristic (Cogan, 1968; Faria *et al.*, 1980; Yeow and Tjia, 1989) Chait and Barber (1979) found this type of nystagmus in one out of three patients. The bithermal caloric test is usually normal, but can show canal paresis with, or without, directional preponderance. Waber and Cass (1993) reviewed the symptoms, signs and vestibular test findings of a series of patients with Chiari type I malformation and indicated that the patients fell in two groups of vestibular test profiles. The group with marked and advanced symptoms showed oculomotor dysfunction, central vestibular nystagmus and abnormal visuo-vestibular interaction. The other group where the Chiari malformation was diagnosed incidentally on MRI scan showed features of peripheral vestibulopathy. Brain stem auditory evoked potentials (ABR) show change in Chiari malformations, although a normal ABR does not rule it out (Longridge and Mallinson, 1985; Albers and Ingels, 1993). Mori *et al.* (1988) studied 16 cases of Arnold-Chiari malformation and found abnormal ABR responses in 75 per cent of cases. All the cases had prolonged III–V wave inter-peak latency while the I–III wave inter-peak latency was prolonged in only 36 per cent of cases. Holliday *et al.* (1985) on the other hand in a study of symptomatic and asymptomatic cases with Arnold-Chiari malformation, between the ages of three months and six years, showed that I–III wave inter-peak latency was more likely to be abnormal than the III–V inter-peak latency. A prolonged inter-peak latency would suggest a stretching of the cochlear nerve while a prolonged III–V inter-peak latency would suggest an abnormality in the brainstem. A prolonged I–III inter-peak latency was also noted by others (Faria *et al.*, 1980). Absence of wave III has also been found (Stone *et al.*, 1983). Mori *et al.* (1988) in their 16 cases with Arnold-Chiari malformation, who varied in age from one day to 33 years, found that the latencies of waves I and V were normal after the age of eight years and suggested that it was due to maturation of the central nervous system. However, ABR changes are found in cases with Chiari malformation above the age of eight years of age. Stone *et al.* (1983) reported a 16-year-old boy with Arnold-Chiari malformation and found inter-peak latency prolongations with significant I–V inter-aural inter-peak latency difference. Faria *et al.* (1980) reported a 30-year-old man with Arnold-Chiari malformation who had a prolonged I–III inter-peak latency. The ABR at presentation in our 13-year-old girl showed prolonged latencies of waves III and V on the left, and the III–V and I–V inter-peak latencies in both the ears individually were within the 95 per cent confidence interval of the normative data. However the I–V inter-peak latencies on the right and left sides showed a significant difference which indicates that the brainstem distortion was not symmetrical (Stone *et al.*, 1983).

Improvement in ABR responses after decompressive surgery for Chiari malformation have been documented. Stone *et al.* (1983) were first to show a gradual normalisation of the latencies of ABR wave forms over a period of six months after surgery. Holliday *et al.* (1985) reported a four-week-old child with Arnold-Chiari malformation and myelomeningocele where the I–III and III–V wave inter-peak latencies reverted to normal after decompression.

Mori *et al.* (1988) found in their cases that decompression resulted in shortening of the brain conduction time even though the latency of each wave was still prolonged compared to the normal. It was suggested that in addition to maturation of the nervous system and compression of the neural elements by the dilated ventricles some other

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intrinsic defect in the brain played a part in prolonging the latencies in such cases.

Following surgery, our patient recovered completely from her audiological symptoms, the tinnitus stopped and her hearing returned to normal subjectively and shown objectively by the total recovery of the pure tone threshold on the left. The electronystagmography six months after surgery showed recovery of the periodic alternating nystagmus. The ABR performed two years after surgery showed a reduced latency for waves III and V on the left, but despite the shortening the latencies were still prolonged compared to the normative data. However the inter-aural I–V wave inter-peak latency difference returned to within the accepted normal limit.

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