The Journal of Laryngology & Otology (2006), ***120***, *343–352*.

doi:10.1017/S0022215106000569
Printed in the United Kingdom
First published online 2 March 2006

Clinical Record

Rare cases of Ménière's disease in children

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Abstract

Classical Ménière's disease is rarely found in children and literature regarding it is scarce. In general, the frequency of Ménière's disease in children is only 0.4–7.0 per cent of that in adults. The progression pattern of Ménière's disease in children is not known well. Here, we report three cases of Ménière's disease in children less than 15 years old, treated over nine years. The three cases comprise 14- and 13-year-old boys and a nine-year-old girl. Two of the three patients initially complained only of recurrent bouts of vertigo, without any tinnitus, ear fullness or hearing impairment. In all three cases, the early pure tone audiograms showed only high tone frequency loss, regardless of subjective hearing loss, and the decrease in the hearing threshold was observed one to eight years after the dizziness attacks began. The hearing threshold was usually decreased to a level of mild or moderate hearing impairment. After diuretic treatment, vertigo was generally well controlled, and some cases showed improvement in hearing. Of the total number of patients with Ménière's disease who visited our department over nine years, 2.6 per cent (3/114) were children, and the overall incidence of Ménière's disease in children with vertigo was 2.0 per cent (3/147). In conclusion, Ménière's disease in children rarely develops and may have characteristics of high tone loss in initial audiograms.

Key words: Ménière's Disease; Child; Vertigo

Introduction

Since Prosper Ménière[1] first reported a patient with vertigo and hearing impairment in 1861, Ménière's disease has become a relatively common otologic diagnosis. Although the number varies from country to country, in general, the incidence of Ménière's disease has been reported as 7.5–157 cases per 100 000, and it occurs commonly regardless of race.[2] Ménière's disease usually occurs in the fifth and sixth decades of life and so is known as a disease affecting middle-aged patients. The classical type of Ménière's disease is rarely found in children and related reports are hard to find.[3] In 1962, Harrison[4] reported that five out of 16 children with dizziness had Ménière's disease, and, in 1978, Stahle *et al.*[5] observed that only one of 257 patients with Ménière's disease was a child and thus the frequency was 0.4 per cent. However, Hausler *et al.*[6] reported that nine out of 598 children complaining of dizziness had Ménière's disease, equating to an incidence rate of 1.5 per cent. In 2003, Choung *et al.*[7] reported that, of 55 cases of dizzy children (for whom otitis media had been excluded), two were diagnosed with Ménière's disease. Reviewing the literature, we found a very low reported incidence rate of Ménière's disease in children, and the progression pattern of Ménière's disease in children is not known well.

Over a nine-year period, we treated three cases of Ménière's disease in patients less than 15 years old, and we have analysed the incidence and the characteristic progression of Ménière's disease in children.

Case reports

The diagnosis of Ménière's disease was made according to the criteria proposed by the Committee on Hearing and Equilibrium of the American Academy of Otolaryngology–Head and Neck Surgery (AAO–HNS) in 1995.[8] Over nine years, the incidence of children with Ménière's disease, compared with the total number of patients with Ménière's disease, was 2.6 per cent (3/114), and the incidence of Ménière's disease in children with vertigo was 2.0 per cent (3/147).

Case one

A 14-year-old boy presented to our hospital on 3 November 1997 with left-sided tinnitus and dizziness that had developed over the previous two months. A review of his past history and the family history revealed no abnormal findings. The patient first experienced severe vertigo when he was 11 years old; this episode lasted three hours. After an asymptomatic period, he developed vertigo again, on 1 September 1997, and thereafter suffered recurrent bouts once every two to three days until presentation. The vertigo characteristically comprised dizziness with a spinning sensation and lasted for four hours. The patient also complained of left-sided ear fullness, tinnitus, nausea and vomiting. At that time, he did not have any hearing loss.

Physical examination showed that both tympanic membranes were intact and no nystagmus was seen. Pure tone audiometry on admission showed that right-sided hearing was normal; however, the left ear showed a peak-type audiogram with thresholds of 5–10 dB at 1, 2 and 3 kHz and 30 dB at 0.25, 0.5, 6 and 8 kHz (Figure 1a). The tympanogram was normal and speech discrimination was 100 per cent in both ears (40 dB threshold). The bithermal caloric test was performed on the fourth day of admission

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Accepted for publication: 18 October 2005.

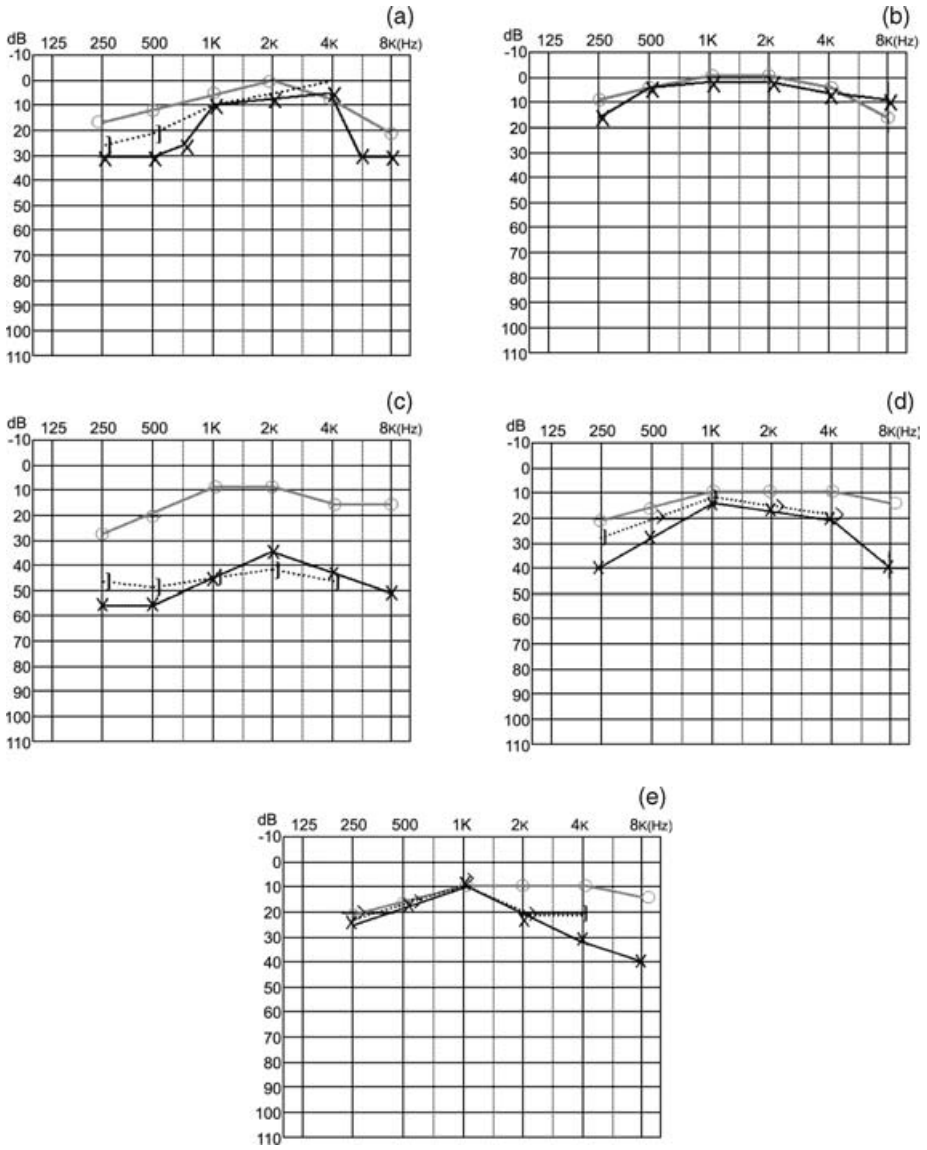

FIG. 1

Pure tone audiograms for case 1: (a) the initial audiogram at age 14 years (3 November 1997); (b) 5 January 1998 (2 months later); (c) 10 June 2002 (5 years later); (d) 2 April 2003 (6 years later); (e) 12 March 2005 (8 years later).

(META-4, Micro Medical Technologies, Chatham, Illinois, USA) and 33 per cent canal paresis was observed in the left ear (Figure 2). The rotational chair test showed a decrease in gain of the vestibulo-ocular reflex (VOR) and an increase in gain of the visual vestibulo-ocular reflex (VVOR) (Figure 2b).

With a provisional diagnosis of Ménière's disease, the patient was given dimenhydrinate (Dramamine®) for one week then flunarizine (Sibelium®) and ginkgo biloba extract (Tanamin®) for two months. His dizziness decreased and hearing impairment resolved (Figure 1b). Although the severity and frequency of the patient's dizziness decreased significantly, the left ear fullness recurred three to four times a month, so, one year after his first visit, he was given hydrochlorothiazide (Dichlozid®) diuretic for three months. The patient's symptoms improved greatly after diuretic treatment.

The patient re-presented to our hospital on 10 June 2002, five years after his first attack, due to recurrent bouts of vertigo, left-sided hearing impairment and tinnitus. The pure tone audiogram at this time showed the left-sided hearing threshold to be 45 dB (Figure 1c). Hydrochlorothiazide was again administered, this time for one month, while gingko biloba extract and trimetazidine (Vastinan®) were given for six months. Thereafter, dizziness subsided and hearing improved (Figure 1d). At his most recent visit, eight years after his first visit, the patient did not complain of dizziness, tinnitus or hearing loss (Figure 1e).

Case two

A 13-year-old boy presented to our hospital on 2 July 1999 due to bouts of vertigo suffered one and three days earlier. He had previously visited our department a year earlier due to right-sided hearing impairment, and, as the pure tone audiogram performed at that time showed high tone hearing loss (Figure 3a), he had been followed up regularly.

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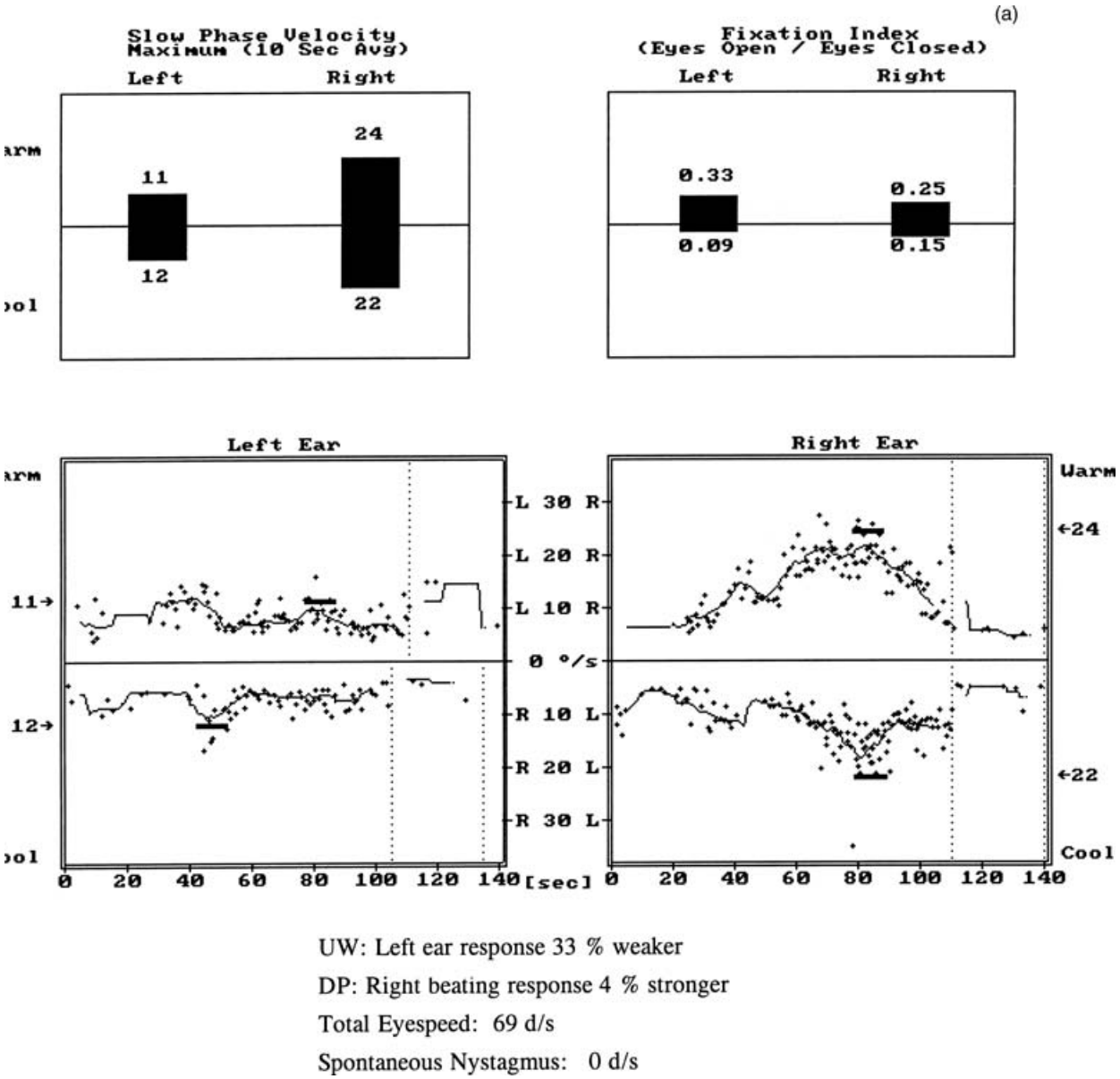

UW: Left ear response 33 % weaker

DP: Right beating response 4 % stronger

Total Eyespeed: 69 d/s

Spontaneous Nystagmus: 0 d/s

FIG. 2

Vestibular function tests for case 1: (a) caloric test shows 33% weakness of left ear; (b) rotation chair test shows decreased gain of vestibular ocular reflex and increased gain of visual vestibular ocular reflex. UW = unilateral weakness; DP = directional preponderance.

A review of the family history showed no abnormal findings. The vertigo characteristically comprised dizziness with a spinning sensation and lasted one hour and 30 minutes. The patient also complained of nausea, vomiting, right-sided ear fullness, tinnitus and hearing loss.

Physical examination showed that both tympanic membranes were intact, and no nystagmus was seen. The pure tone audiogram was similar to that performed a year previously (Figure 3b). However, the bithermal caloric test resulted in a 45 per cent unilateral weakness in the right ear (Figure 4a), and the rotational chair test showed a decrease in gain in the lower frequencies and a phase lead on VOR (Figure 4b).

Under a provisional diagnosis of Ménière's disease, the patient was given hydrochlorothiazide, ginkgo biloba extract and trimetazidine. Dizziness decreased, but pure tone audiometry performed one month later showed a peak type with a moderate degree of hearing impairment (Figure 3c). However, after two months of medication, the patient's hearing improved (Figure 3d). Hydrochlorothiazide was given for a total of 18 months, until January 2001. Overall, the frequency of the patient's dizziness decreased to two to three times every six months, and his hearing recovered to a mild hearing impairment level (after falling to a moderate level once, in August 2000) (Figure 3e). At last presentation (March 2005), the patient did not complain of hearing loss or dizziness (Figure 3f).

Case three

A nine-year-old girl presented to our hospital on 18 March 2003 complaining of bouts of dizziness lasting for 12 hours. She had experienced recurrent attacks of dizziness since she

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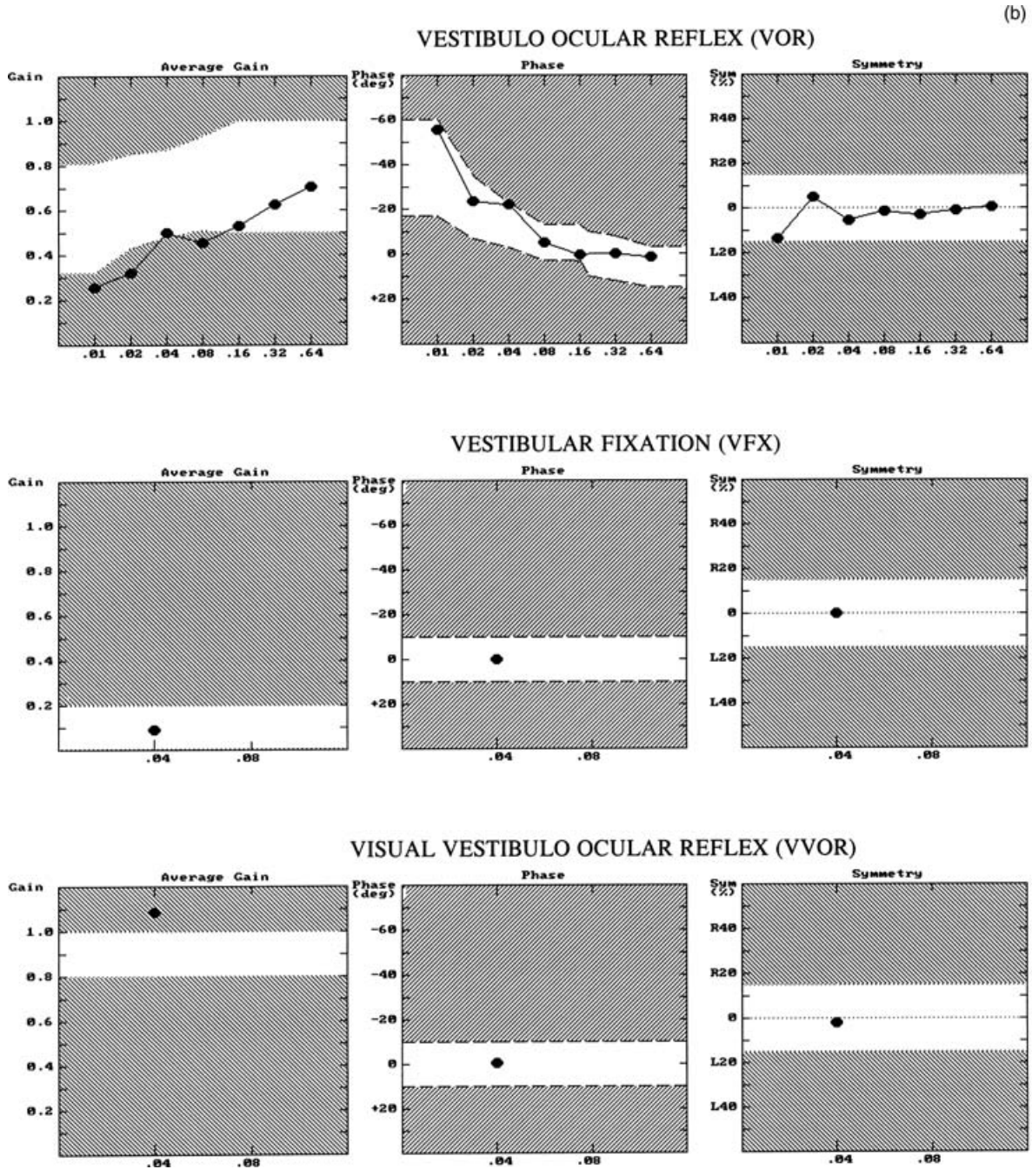

FIG. 2
Continued

was four years old and had been treated at another hospital five months prior due to vertigo lasting more than 10 hours. The bouts of dizziness were of a true vertigo nature. Although the patient complained of nausea, there were no auditory symptoms or headache. The bouts occurred about once every two weeks, regardless of position. There were no abnormal findings in the routine laboratory tests or on brain magnetic resonance imaging.

Physical examination showed that both tympanic membranes were intact, and no nystagmus was seen. Pure tone audiometry revealed a downward-type audiogram, with thresholds of 30 dB and 45 dB at 6 and 8 kHz, respectively (Figure 5a). However, the patient did not notice any hearing impairment. The bithermal caloric test resulted in a 53 per cent unilateral weakness in the left ear (Figure 6a), and the rotational chair test showed a phase lead on VOR and left symmetry in the lower frequencies (Figure 6b).

As there was no initial complaint of auditory symptoms, the patient was treated under a differential diagnosis of either benign paroxysmal vertigo of childhood (BPVC) or possible Ménière's disease and was given flunarizine and gingko biloba extract. The symptoms subsided initially; however, one month later, the patient presented again due

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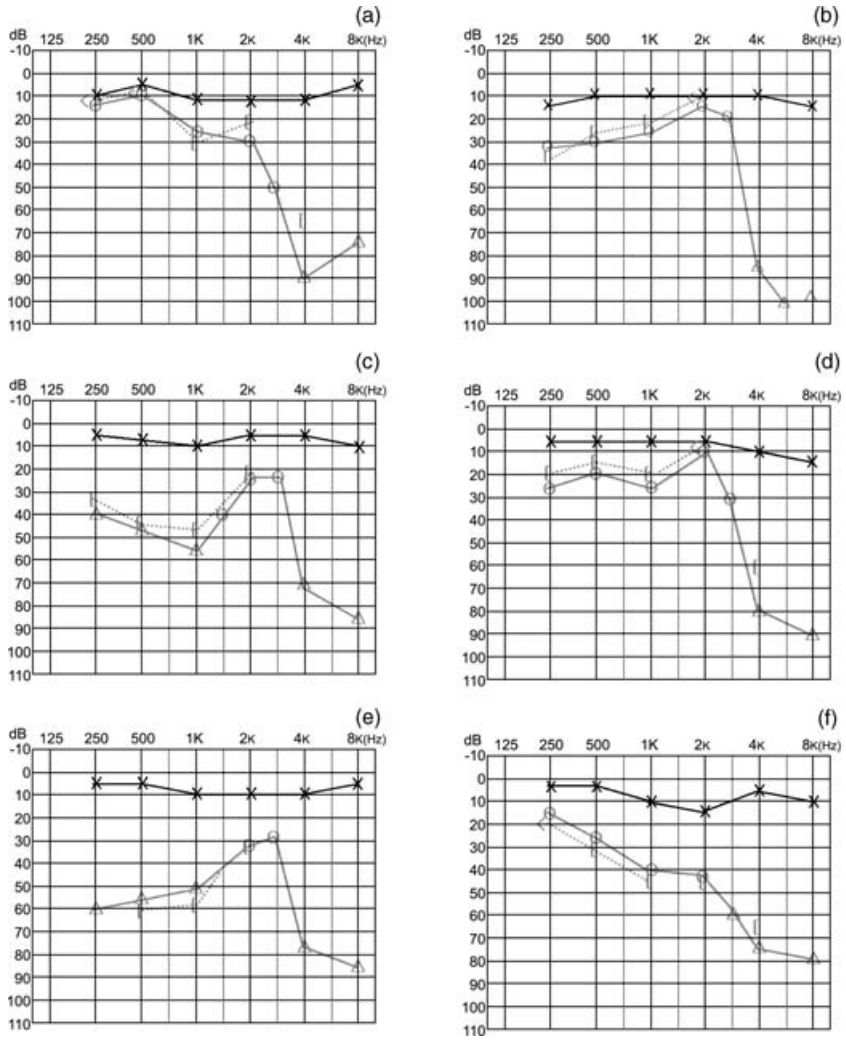

FIG. 3

Pure tone audiograms for case 2: (a) the initial audiogram at age 12 years (21 August 1998); (b) 2 July 1999; (c) 6 August 1999; (d) 10 September 1999; (e) 18 August 2000; (f) 5 January 2003 (same PTA on 8 March 2005).

to vertigo. Nausea and vomiting were severe and right-beating nystagmus was observed. With a provisional diagnosis of Ménière's disease, the patient was given the diuretic spironolactone (Aldactone®). The frequency of dizziness decreased to once every one to two months and the vertigo bouts were mild.

However, eight months later, in November 2003, the frequency of vertigo bouts increased to four every two weeks, and tinnitus, ear fullness and hearing loss developed. Pure tone audiometry performed at that time showed a mild to moderate level of hearing impairment in all frequencies (Figure 5b). After diuretic treatment, the patient's hearing recovered to a near normal range (except for some high frequency impairment seen in the pure tone audiogram performed in October 2004) (Figure 5c). Following treatment, the patient was given diuretics continuously for 17 months; she did not experience dizziness or hearing impairment over the following five months.

Discussion

Compared with adults, children who complain of dizziness tend to be overlooked. This is thought to be because children cannot express themselves well and also because the cause of dizziness is not easily defined. Bower and Cotton[9] reported that more than 50 per cent of cases of dizziness in children were due to otitis media, BPVC, migraine and vestibular neuritis, while Choung *et al.*[7] reported BPVC and migraine as the most common causes. Although some studies conclude that peripheral stimulation, such as otitis media, is more commonly the cause of childhood dizziness,[10] other reports state that systemic effects via central nervous system stimulation is the more common cause.[11]

Ménière's disease is rarely found in children, in contrast to adults. Reviewing the literature to date, compared with adults, the frequency of childhood Ménière's disease has been variously reported by Stahle *et al.*,[5] Meyerhoff *et al.*, [12] Filipo and Barbara,[13] and Hausler *et al.*[6] as being 0.4 per cent (1/257), 3 per cent, 7 per cent and 1.1 per cent (10/936), respectively. The incidence of Ménière's disease in children who complain of dizziness has been variously reported as 1.5 per cent (99/598), 2.8 per cent (2/72), 0 per cent (0/121), 4 per cent (1/25) and 2.9 per cent (3/103), by Hausler *et al.*,[6] Yoshimoto,[14] Yanagida,[11] Bower and Cotton,[9] and Akagi *et al.*,[15] respectively.

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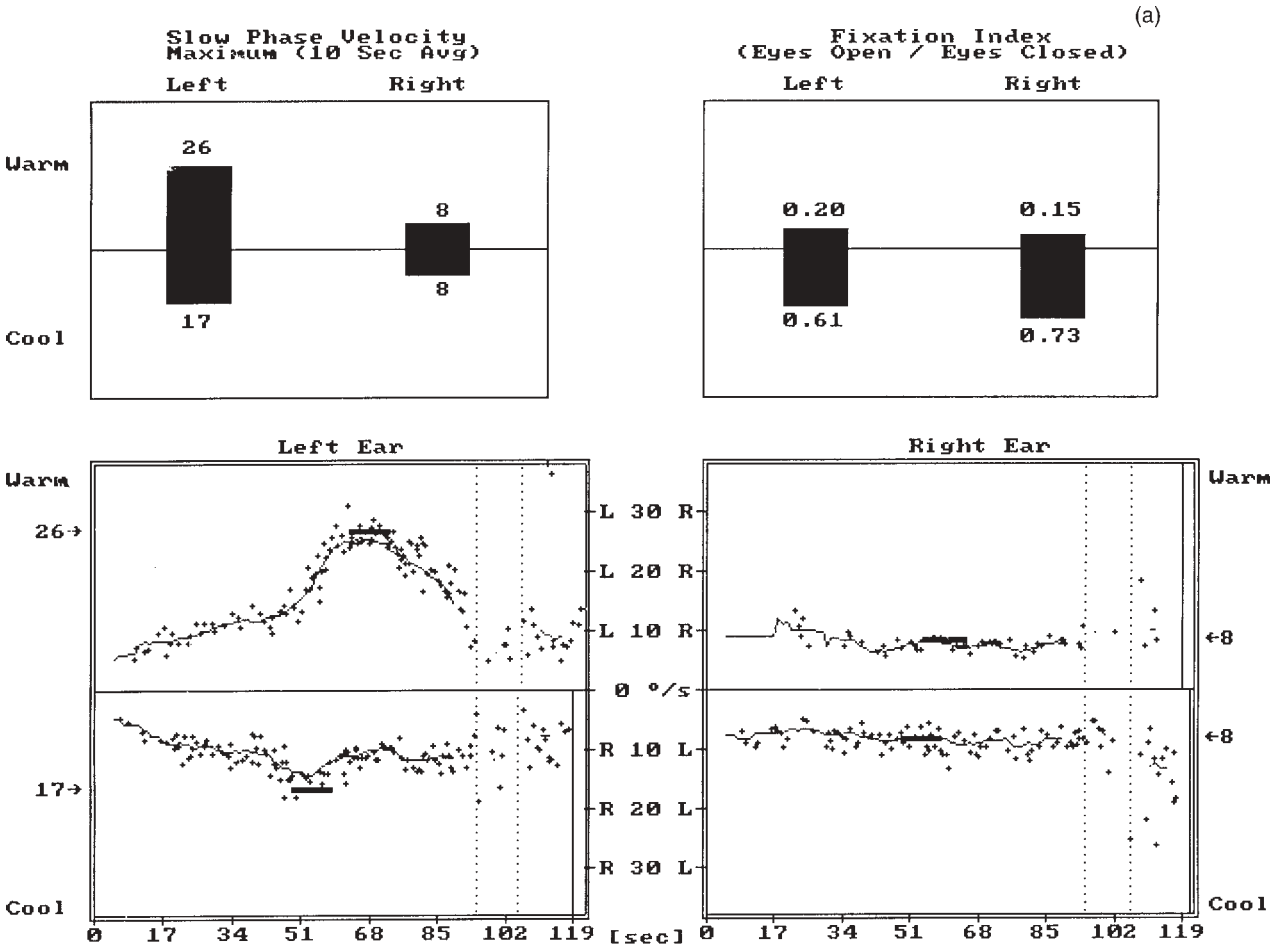

FIG. 4

Vestibular function tests for case 2: (a) caloric test shows 45% weakness of right ear; (b) rotation chair test shows decreased gain and phase lead of vestibular ocular reflex. UW = unilateral weakness; DP = directional preponderance.

In the current report, of the 147 patients 15 years old or younger who visited our hospital from January 1995 to April 2003, there were three cases of Ménière's disease (2.0 per cent). During the same period, 114 patients (adults and children) presented to our hospital with definite symptoms and signs of Ménière's disease. Of these 114 cases, only three were patients 15 years old or less, making the incidence 2.6 per cent. These results are similar to those of other reports.

Another characteristic of childhood Ménière's disease is difficulty in establishing a diagnosis in the early stages. Such a diagnosis requires the patient to have hearing impairment. According to Schknecht,[16] hearing loss in Ménière's disease usually develops within a year of the onset of recurrent vertigo; however, Goodman[17] describes hearing loss as a late-stage symptom, occurring years after the onset of vertigo. In general, vertigo, tinnitus, hearing loss and ear fullness develop relatively early in adult Ménière's disease, making it easier to diagnose. However, the aural symptoms in children with Ménière's disease seem to develop over a long period of time, so that the diagnosis may be made when patients are in their early 20s.[18] Of our three patients, two initially complained only of dizziness, and the diagnosis could only be made six to eight years later when the other classical signs of Ménière's disease had developed. Thus, Ménière's disease should also be suspected in children with one to four hours of severe vertigo, even if there are no other aural symptoms.

There are few reports of the early symptoms and characteristics of Ménière's disease in children. However, it is known that hearing impairment is generally found in the low frequencies in the early stages, showing as an ascending-type audiogram. This type of audiogram later becomes a horizontal or peak type.[19] However, our three cases all showed high frequency loss whether or not the child complained of hearing loss, with two cases having descending-type audiograms and one case having

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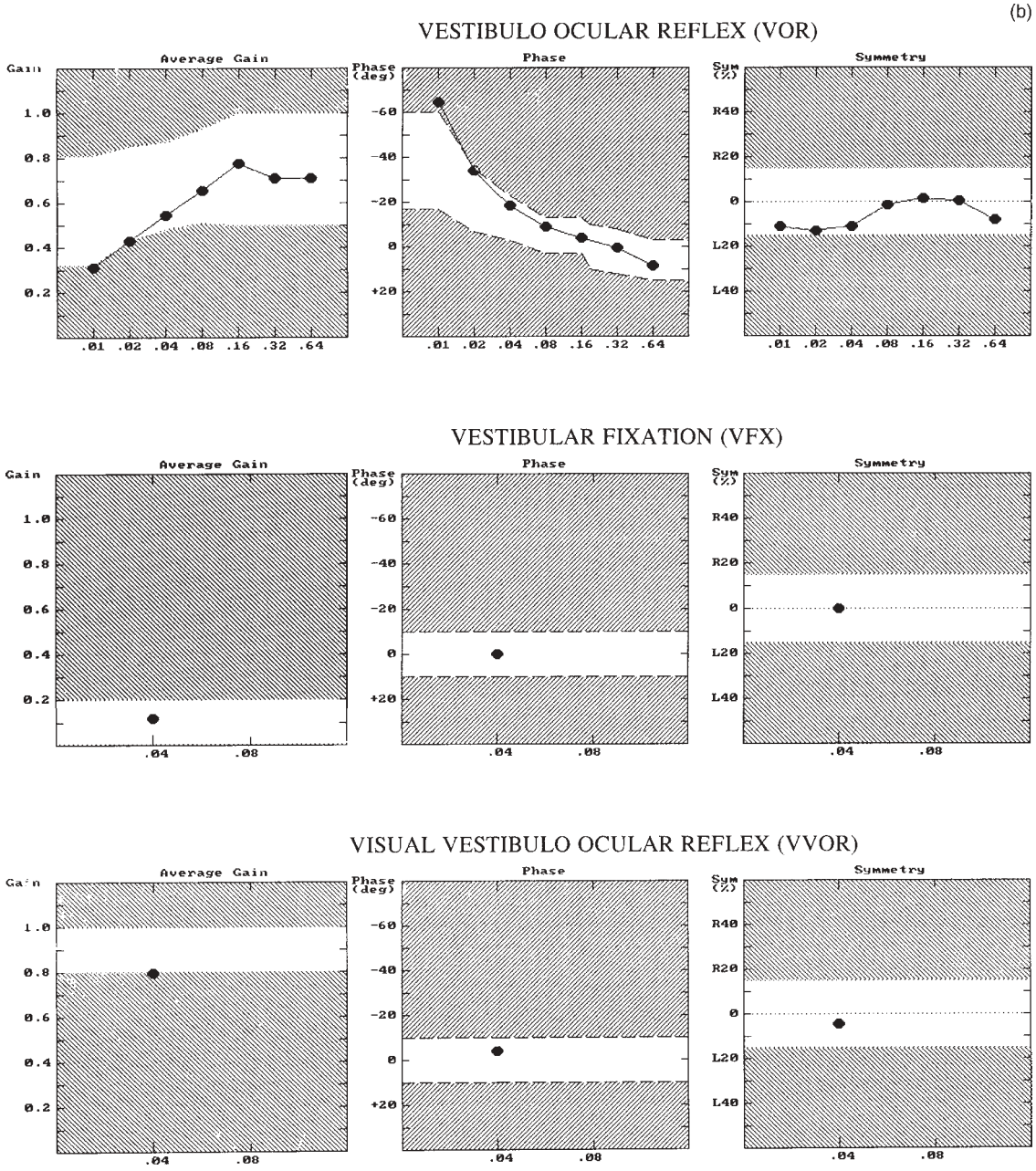

FIG. 4
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a peak-type audiogram. The decrease of hearing threshold was seen one to eight years after vertigo had developed, and hearing impairment was usually of a moderate degree. This study found a characteristic descending-type audiogram in our child patients, compared with the ascending type seen in adults. We believe that this finding can help establish an earlier diagnosis of childhood Ménière's disease.

In addition, in our study, all cases were treated with diuretics. After administration of diuretics, vertigo was well controlled and all three cases showed improvement of hearing loss. The hearing in two cases recovered to nearly normal level, however one case showed limited hearing improvement within a mild hearing impairment level. In conclusion, we believe that diuretics should be used if Ménière's disease is suspected in children, and that this will help prevent development of hearing impairment.

- **Classical Ménière's disease is rarely found in children. In general, the frequency of Ménière's disease in children is only 0.4–7.0 per cent of that in adults**
- **This paper reports three cases of Ménière's disease in children less than 15 years old, treated over a nine year period. Two of the three cases initially complained only of recurrent bouts of vertigo, without any tinnitus, ear fullness or hearing impairment**
- **In all three cases, the early pure tone audiograms showed only high tone frequency loss, regardless of subjective hearing loss. After diuretic treatment, vertigo was generally well controlled, and some cases showed improvement in hearing**

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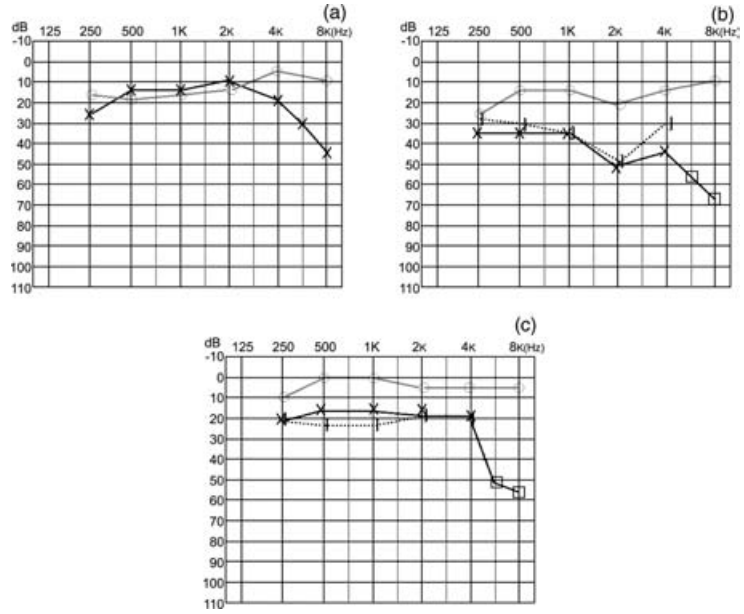
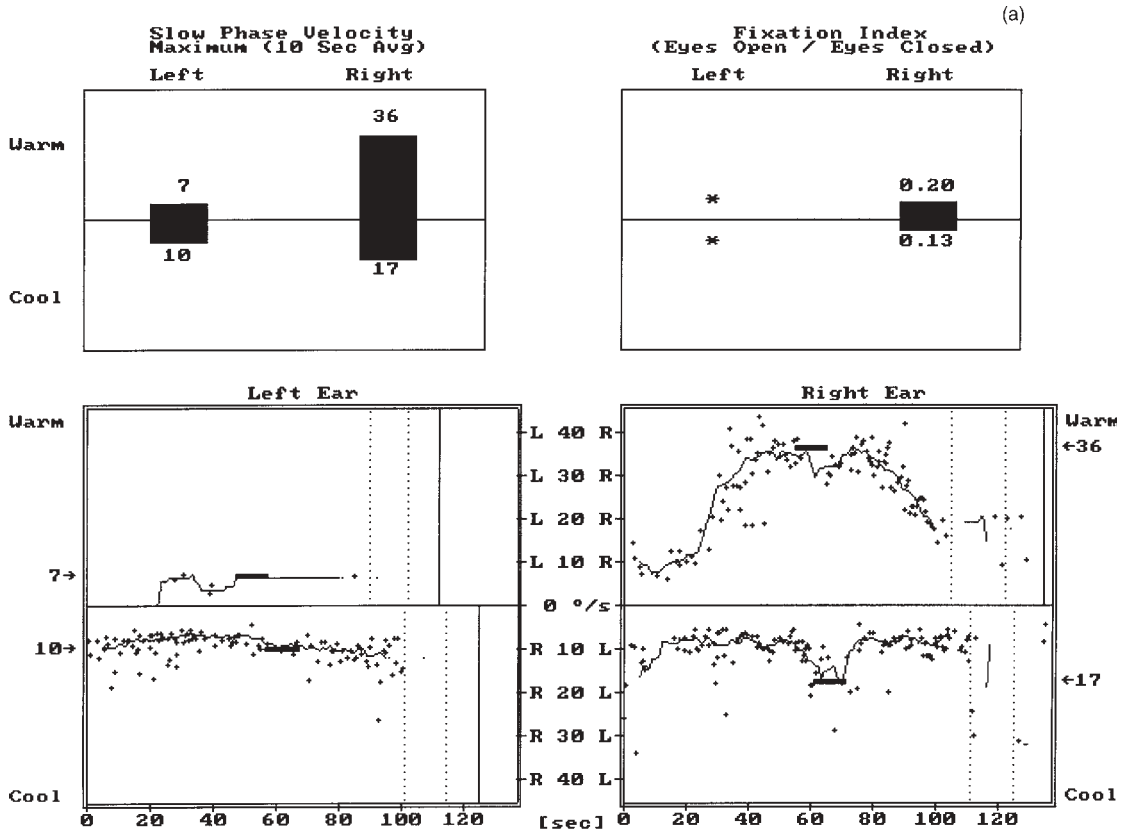


FIG. 5
Pure tone audiograms for case 3: (a) the initial audiogram at age 9 years (18 March 2003); (b) 26 November 2003; (c) 1 October 2004.



UW: Left ear response 53 % weaker
DP: Right beating response 31 % stronger
Total Eyespeed: 70 d/s
Spontaneous Nystagmus: 0 d/s

FIG. 6
Vestibular function tests for case 3: (a) caloric test shows 53% weakness of left ear; (b) rotation chair test shows phase lead and asymmetry of vestibular ocular reflex. UW = unilateral weakness; DP = directional preponderance.

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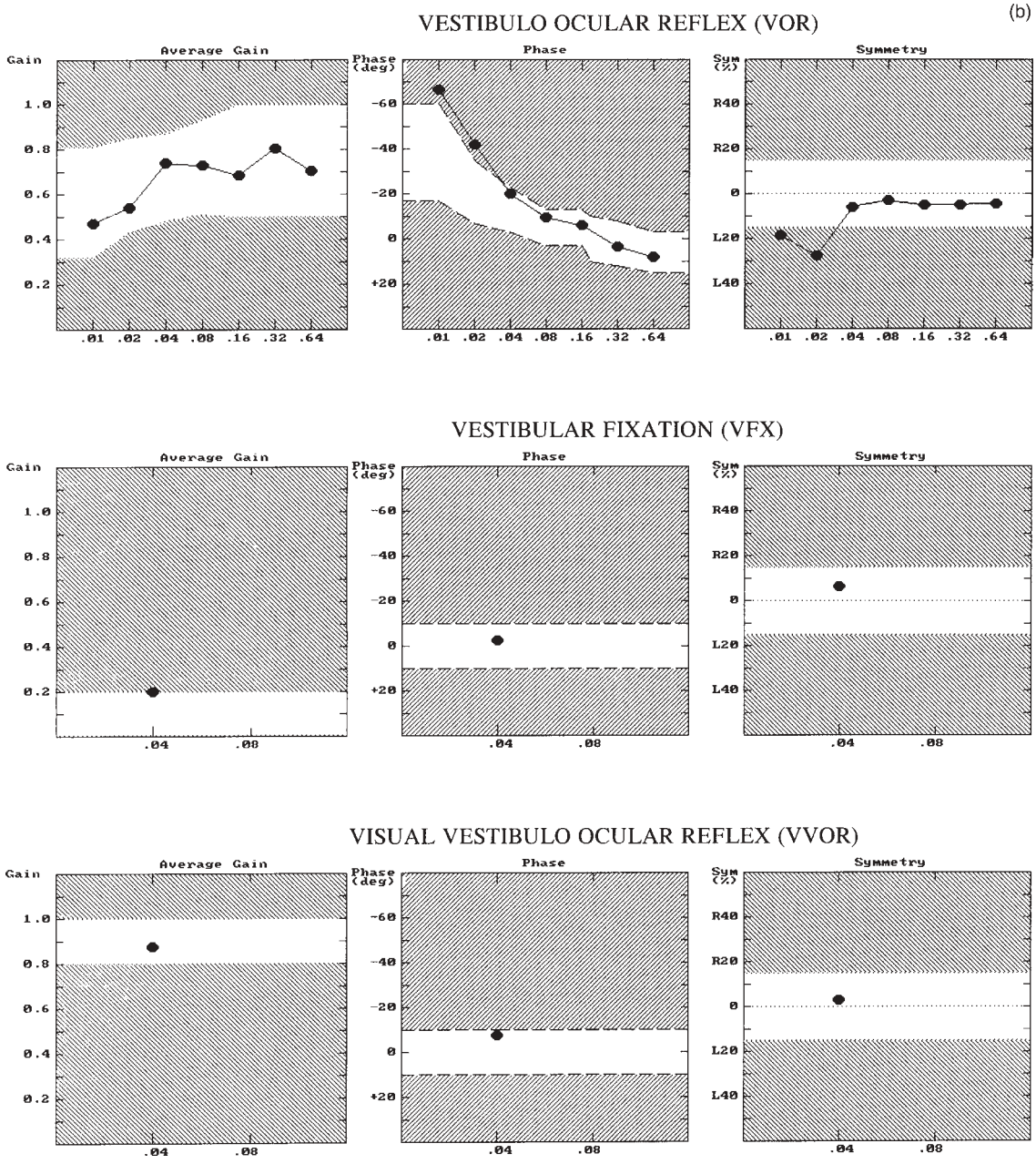

FIG. 6
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Acknowledgement

The authors thank Mi-Yoo Koh for technical assistance with the vestibular function tests.

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Dr Yun-Hoon Choung takes responsibility for the integrity of the content of the paper.
Competing interests: None declared