myelination in the developing neonate and infant. Although most of these radiological reports did not mention the brainstem auditory nuclei and pathway, a few papers about MRI study mentioned about brainstem auditory pathway. Only lateral lemniscus and inferior colliculus which is a part of the auditory pathway were mentioned by Martin et al. [21] and Counsell et al. [35] (Table 1). According to Martin's study inferior colliculus myelinated before 40 fetal weeks, and Counsell mentioned the lateral lemniscus is myelinated at 26 fetal weeks (T1-weighted) and 27 fetal weeks (T2-weighted) and the inferior colliculus is myelinated at 25 fetal weeks (T2-weighted).

Our results and Martin's study showed solidarity. On the other hand, Counsell's study showed earlier myelination period than our study or Martin's study, and reflects more closely previous histological study results. However, Serena's study is performed by 1.0 T MRI which is not so high-resolution than Martin's study (2.35 T) or our study (1.5 T), in addition, imaging slices were used for visual evaluation which is not so objective method, so it was considered difficult to evaluate correctly the small structure of very early brain (their subjects were 26 preterm infants with a median fetal age of 28 weeks). The reason of time lag between Serena's study and Martin's study or our study is considered in performance of the MRI unit or in methodology of evaluation (visual evaluation and ROI analysis).

4.3. The difference of T1- and T2-weighted imaging

Our study shows that in the cochlear nucleus, superior olivary nucleus, and inferior colliculus, myelinational intensity change did not show a significant difference in T1-weighted images and myelinational intensity change was identified only in T2-weighted images, although in the lateral lemniscus, T1-weighted images showed the change of intensity earlier than in T2-weighted imaging. T2-weighted sequences were superior to T1-weighted sequences in demonstrating the contrast between gray matter nucleus and surrounding white matter and were therefore more suitable for evaluating gray matter nucleus [35]. This might be explained by the fact that in a high-field-strength system, the difference in the values of the T1 relaxation times of gray matter nucleus and white matter is not sufficiently large [35]. T2-weighted MRI was superior at showing myelin in deep gray matter nuclei and T1-weighted MRI was better at showing myelin in white matter tracts. This could be due to the characteristics of the anatomic area; other MRI studies dealing with brainstem regions also confirm this observation [34,35]. These results suggest that partially modified protocols may be useful for assessing myelination in the brainstem.

4.4. Myelination progress from other aspects

We have also taken into account physiological study results to consider the myelination progress of the brainstem auditory nuclei and pathway. In the visual system, the duration of functional maturation (spatiotemporal vision) correlates with the duration of myelination of the optic radiation [33,39,40]. Moore et al. [4] mentioned that the time of onset of myelination coincides with the onset of acousticomotor reflexes or auditory startle reaction. Auditory function is also considered to correlate with myelination of the brainstem auditory nuclei and pathway. The myelin sheath surrounding an axon is composed of multiple segments of myelin [34]. Each segment is a modified plasma membrane that originates as an extension of an oligodendroglial cell process [34]. Small segments of bare axon are situated between the myelin sections, exposed to the interstitial space. These segments are called nodes of Ranvier. When the axonal membrane receives an action potential, the electrical impulse is unable to propagate through the high-resistance myelin sheath; therefore, the impulse "jumps" to the next node, which might be 1 mm or farther away [34]. Because of the low capacitance of the sheath, the remaining axonal membrane between the nodes depolarizes with little energy requirement and markedly increased speed. The increasing speed of fiber conduction involving the auditory brainstem nuclei and pathway can be measured by auditory brainstem response (ABR). Many researchers [5–9] have described a decrease in peak and interpeak latencies of ABR in early infancy and attributed the phenomenon to myelination of the brainstem auditory nuclei and pathway. The ABR wave I comes from the distal end of the auditory nerve, the generation of wave II is from the cochlear nucleus, wave III involves the superior olivary nucleus, wave IV is from the lateral lemniscus, and wave V is from the inferior colliculus. These ABR studies indicate that the speed of axonal conduction is low in children under 1 year of age and rapid axonal conduction gradually develops in the brainstem auditory nuclei and pathways. The increase of myelin density is likely to be a factor in the steady decrease in ABR interval between wave I and waves III–V.

5. Conclusion

Regarding the brainstem auditory nuclei and pathway, 1.5 T MRI revealed the signal intensity change by myelination at an average of 3.5 months (11–18 weeks) later than those reported in the histological literature. This time lag suggests that apart from histological research (comparing with histological work is not correct when evaluate the maturation of central auditory pathway using MRI because of time lag as shown in this study), the necessity for the new milestones of brainstem auditory pathway maturation using MRI is suggested. We suggest the new milestone of MRI used evaluation for brainstem auditory pathway in this study.

Myelination does not take place suddenly but happens gradually, so definite myelination, with a full change of myelin sheath ingredients (loss of water and gain of lipids), is needed to be detectable by MRI. This study shows the progress pattern of myelination in the brainstem auditory nuclei and pathway on MRI. These results can be used to assess with MRI the auditory system maturation of infants.

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Original Article

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Key Words
Congenital deafness
Vestibular failure
Central vestibular
Compensation
Caloric test
Rotation test
Vestibular myogenic potential

Abbreviations
ABR: Auditory brainstem response
COR: Conditioned orientation reflex audiometry
ENG: Electronystagmography
VEMP: Vestibular myogenic potentials

Vestibular failure in children with congenital deafness

Abstract
Congenitally deaf infants and children commonly suffer vestibular failure in both ears, and impairment of postural control, locomotion, and gait. The development of gross motor functions, such as head control, sitting, and walking is likely to be delayed, but fine motor function is usually preserved unless disorders of the central nervous system are present. These children can eventually catch up with their normal peers in terms of development and growth as a result of central vestibular compensation. The visual and somatosensory systems, pyramidal and extrapyramidal motor system (cerebellum, basal ganglia, cerebrum) and intellectual development, compensate for vestibular failure in infants and children with congenitally hypoactive or absent function of the semicircular canals and otolith organs.

Sumario
Los niños con sordera congénita comúnmente sufren falla vestibular en ambos oídos y discapacidad del control postural, la locomoción y la marcha. El desarrollo de las funciones motoras gruesas tales como el control cefálico, la sedestación y la marcha suele presentar retraso, pero la motricidad fina generalmente está conservada a menos que coexista algún desorden del sistema nervioso central. Estos niños usualmente alcanzan el nivel normal en términos de crecimiento y desarrollo como resultado de la compensación central. Los sistemas visual y somatosensorial; piramidal y extrapiramidal (cerebelo, ganglios basales y cerebro) y el desarrollo intelectual compensan la falla vestibular en los niños con hipoactividad o ausencia de la función de los canales semicirculares y órganos otolíticos.

With the introduction of new born hearing screening, hearing problems are often detected at the neonatal stage and in early infancy, and holistic management including not only auditory, but also vestibular function is needed.

In children, vestibular function plays an important role in gross motor development, and, therefore, otologists and audiologists should recognize and understand the high incidence of vestibular dysfunction in congenitally deaf children and be prepared to undertake appropriate evaluation. However, there have been very few studies that have investigated all aspects of vestibular function in congenitally deaf infants and young children. In infants and young children with hypoactive labyrinths, the loss of postural control is much more common and the development of gross motor function is delayed (Rapin, 1974). Kaga and colleagues have documented hypofunction or loss of vestibular function using rotational chair testing in congenitally deaf infants, in whom head control and independent walking were delayed. They have also emphasized vestibular involvement in the development of gross motor function and the significance of vestibular assessments of congenitally deaf infants (Kaga et al, 1981, 1988; Kaga, 1999).

The cochlear and vestibular organs are closely related anatomically and phylogenetically, and the relationship of vestibular function with hearing impairment has been discussed, since the 1950s. Several studies have been performed to establish the incidence of vestibular pathology in children with congenital and acquired deafness (Arnvig, 1955; Goldstein 1958; Everberg, 1960).

Although these studies have employed the caloric test for the assessment of horizontal semicircular canal function, most of them did not evaluate the other two semicircular canals or otolithic organs, whose functions may have more influence on the development of postural control and locomotion than the horizontal semicircular canals alone. In this paper, we report our studies on central vestibular compensation in deaf children with congenital vestibular failure demonstrated by the caloric test, the damped rotation test, and the vestibular myogenic potential (VEMP).

Methods to assess vestibular function

Three procedures were used to assess vestibular failure in congenitally deaf children:

Ice-water caloric test
The ice-water (4°C and 2 ml) caloric test was performed, irrigating the external auditory meatus to induce a thermal gradient across the horizontal semicircular canal of one ear. The duration of induced nystagmus was measured and evaluated statistically ($p < 0.05$) for asymmetry and hypofunction of the

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horizontal semicircular function, comparing with age-matched controls. Horizontal and vertical eye movements were recorded using standard electronystagmography (ENG) electrodes.

Damped rotational chair test

For the damped rotational chair test, a rotational chair (Nagashima Co. Ltd, S-II) was accelerated to a maximum rotational velocity of 160°/s^2, then decayed by 4°/s^2. The test was performed once in a clockwise direction and once in counter-clockwise direction in total darkness. Eye movements were recorded by ENG and the duration and number of beats of perrotatory nystagmus were calculated to evaluate semicircular canals and otolith organ function in both ears (Figure 1).

Vestibular myogenic potentials (VEMP)

VEMPs were recorded using conventional VEMP recording procedures in our hospital (Jin, 2006). Sound stimuli of clicks (0.1 ms, 95 dB nHL) were presented to each side of the ear through headphones (Figure 2). Electromyographic signals from the stimulated side of the sternocleidomastoid muscle were amplified. The stimulation rate was 5 Hz, the band-pass filter intensity was 20–2000 Hz, and the analysis time was 50 ms. VEMPs in response to 100 stimuli were averaged twice. VEMPs were considered significant when there was a reproducible short-latency biphasic wave (p13-n23). An absolute VEMP ratio of <0.5 was considered to indicate significant asymmetry. If the amplitude of p13-n23 was <50 μV on both sides, the subject was considered to have hypofunction bilaterally. VEMPs are responses of the sternocleidomastoid muscle via the saccules to clicks. Developmental changes of VEMPs are shown in Figure 3.

Incidence of vestibular failure revealed by caloric test, damped rotation test, and VEMP test in congenitally deaf children

Twenty children (11 boys, 9 girls; age range 31–97 months, mean age 54.2 months) with a severe congenitally profound hearing impairment, fitted with hearing aids, and who planned to

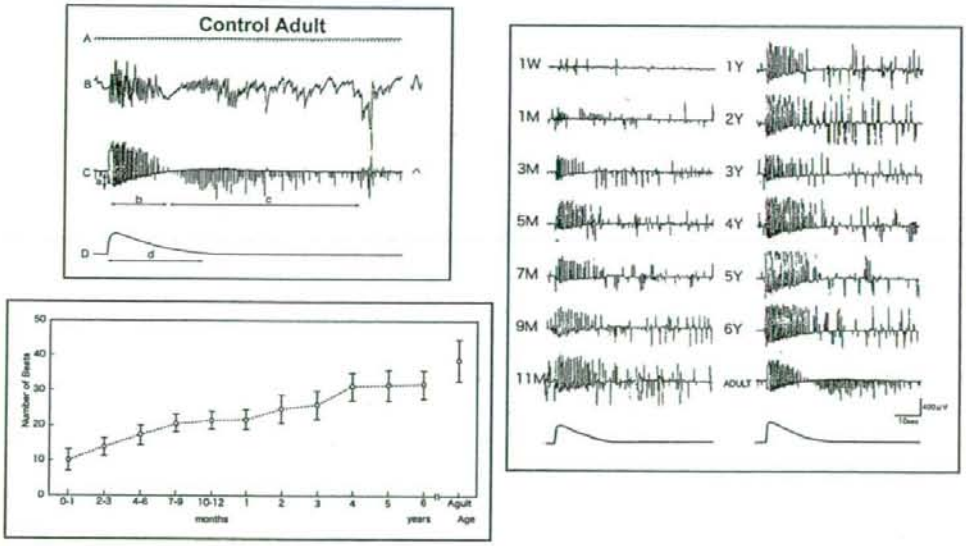

Figure 1. ENG recording of the damped rotation chair test.
1. A typical recording in an adult.
(A) Time scale (one division per second).
(B) Angular displacement of eyes (time constant, 0.3 s; calibration signal, 10°).
(C) Rotational velocity eyes (time constant, 0.003 s; calibration signal, 20°/s):
a. maximum slow-phase velocity during rotations;
b. duration of perrotatory nystagmus;
c. duration of postotatory nystagmus.
(D) Angular velocity of rotating chair;
d, duration of rotation.
2. Developmental recordings with age (W = weeks, M = months, Y = years).
The damped rotation test to the right from the normal neonates, infants, children and adults.
3. Developmental changes of number of beats in perrotatory nystagmus with age.

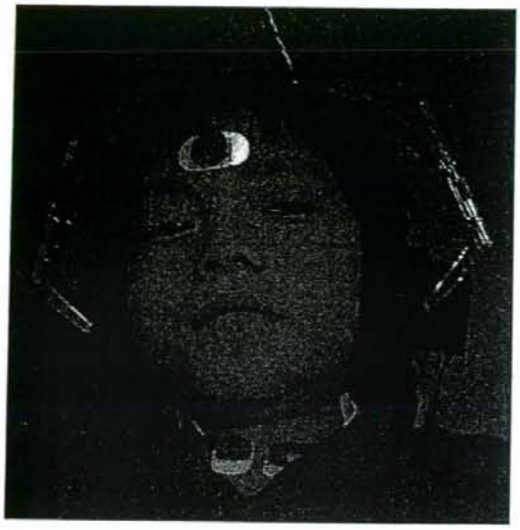

Figure 2. Electrode placement for VEMP recording, a relatively new method for measuring the vestibulo-spinal reflex.

undergo cochlear implantation (CI) at the University of Tokyo Hospital were studied. Before CI, their auditory and vestibular functions were evaluated.

Their hearing levels ranged from 87.5 dB to unrecordable, and all revealed a severe hearing impairment bilaterally.

Caloric test

Three children (15%) showed normal responses bilaterally. Seven children (35%) showed asymmetrical response, and two children (10%) showed hypo-reactions in both ears. Eight children (40%) showed no response bilaterally.

Damped rotational chair test

Fourteen children (70%) showed normal responses during rotation in both directions. One child showed normal responses during rotation in one direction, but poor responses in the opposite directions. Two children (10%) showed poor response in both directions, and three children (5%) showed no responses in any direction.

VEMP test

Ten children (50%) showed normal responses bilaterally. Six children (30%) showed asymmetrical responses, and four children (20%) showed no responses bilaterally.

Summary of vestibular assessment

These 20 severely hearing-impaired children were classified into four groups. Group A: only three (15%) showed normal responses in the caloric test, rotational chair test, and VEMP recording bilaterally. Group B: seven (35%) showed responses asymmetrically in the caloric test, despite normal responses in the rotational chair test and VEMP recording bilaterally. Group C: five (25%) showed hyporeflexia or areflexia in the caloric test bilaterally, but showed normal responses in the rotational chair test and normal reproducible or decreased VEMPs. Group D: five (25%) showed no responses at all in the caloric test, rotational chair test, and VEMP recording. In Figures 5 and 6, three typical recordings of vestibular assessment in two cases of A and D are shown.

Among the children, 85% showed abnormal responses in at least one test. Regarding the ice-water caloric test, 10 children (50%) showed caloric hypofunction or areflexia. The proportion of caloric hypo- or areflexia was slightly higher than those in several previous studies (Arnvig, 1955; Goldstein, 1958; Everberg, 1960), which ranged from 20% or 40%. It was considered that some of the children had lost vestibular hair cells in association with the congenital abnormality. According to recent research (Ito, 1998; Buckman, 2004), nearly 70% of cochlear implant children showed absent or reduced responses on caloric irrigation before operation.

As regard the rotational chair test, our study demonstrated that 70% of the children showed normal responses. That is, some children (Group C, 25%) showed normal responses in the rotational chair test despite their hypo- or areflexia in the caloric test. This result is inconsistent with the study of Tribukait and co-workers (2004), in which subjects showing no caloric responses did not show any nystagmus on rotation. The rotational chair test at higher frequencies may stimulate the semicircular canals and otolith organs in both ears simultaneously, whereas the caloric test stimulates only one horizontal semicircular canal at a time at very low frequency. The rotational chair rest may possibly stimulate labyrinths, including three semicircular canals and otolith organs on both sides, more strongly than the caloric test; thus, it will be able to detect the remaining weak vestibular function.

In the VEMP test, assessing saccular function, 50% of children showed normal responses bilaterally, 30% asymmetrical responses, and 20% no responses. There are few comparable data in the literature and this high prevalence of VEMPs may be related to earlier embryological differentiation of the sacculus as opposed to the semicircular canals.

However, these results are very similar to those of the VEMP study of Tribukait and co-workers (2004). Despite the difference in the children's ages, the proportions in that study showing normal responses bilaterally, asymmetrical responses, and no responses were 58%, 17%, and 25%, respectively. According to these results, saccular function would appear to be mature even at this early age of childhood (Shinjo, 2007).

Developmental milestones in children with congenital vestibular failure

Head control, balance, locomotion, and gait are observed as part of the neurovestibular examination of developmental milestones to define when these children catch up with normal children. Figure 4 shows a diagram (checklist) to assess primitive reflex and motor development which was presented by this author (Kaga, 1980).

Delay and acquisition of gross motor and balance function are compared in patients with congenital vestibular failure, without mental retardation.

Group 1a

Vestibular hypoactivity with normal intelligence and without inner-ear anomalies: Age of acquisition of gross motor functions

is shown in Table 1. Generally, all gross motor functions are delayed but are eventually attained.

Group 2a

Vestibular hypoactivity with normal intelligence and without semicircular canals, because of inner-ear anomaly. Age of acquisition of gross motor functions is shown in Table 1. The ages of acquisition in this group were close to those in group 1a (Kaga, 1981, 1988, 1999) (Figure 7).

Children with congenital vestibular failure dysfunction eventually compensate well, whereas adult patients with acquired vestibular loss often compensate poorly. This difference indicates that the plasticity of the developing brain in children with congenital vestibular failure can overcome delay of gross motor and balance function.

The pathophysiology of this delayed motor development could be caused by congenital loss of discharges of sensory cells from semicircular canals and otolithic organs through medial

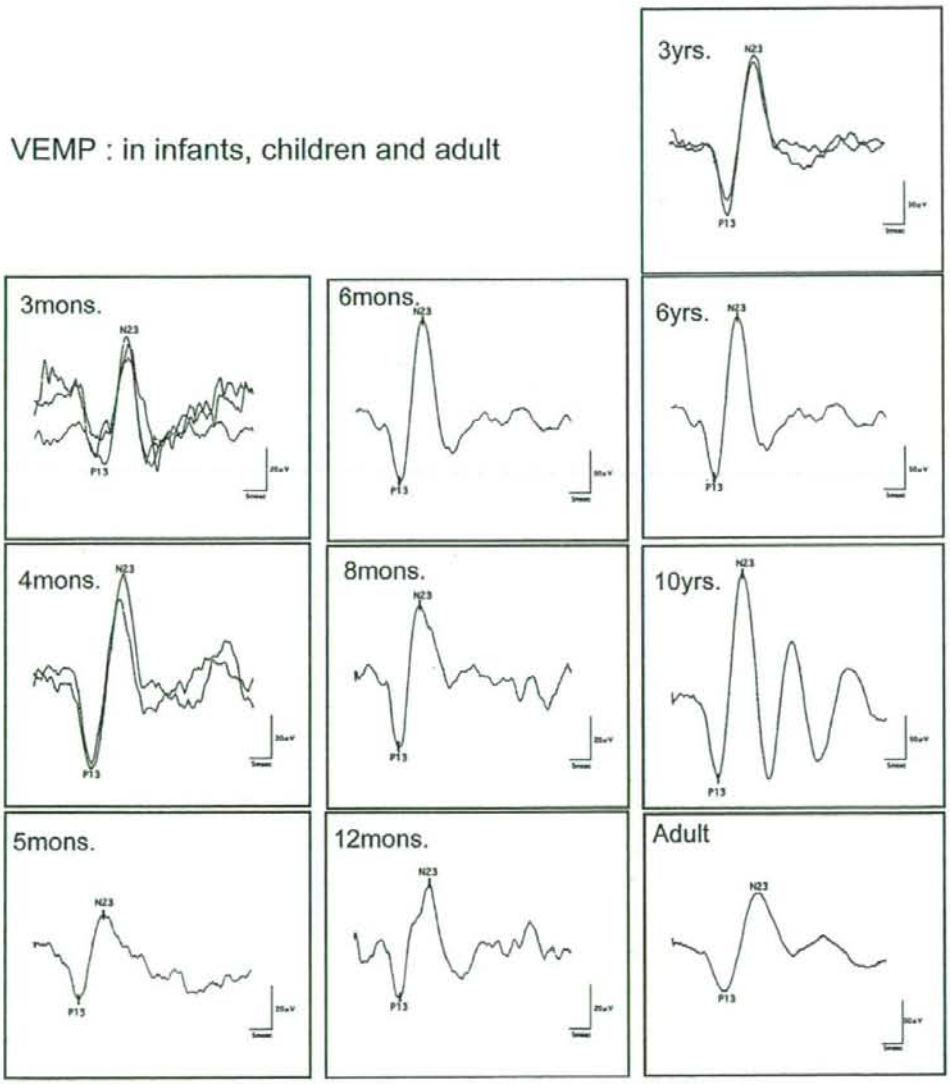

Figure 3. Developmental changes of VEMPs.

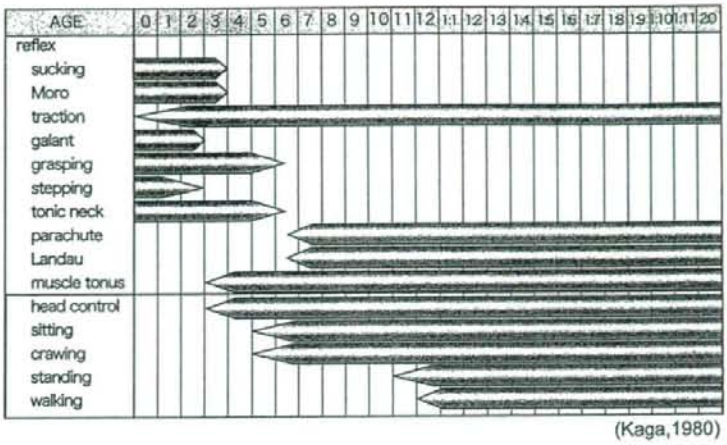

Figure 4. A chart to check developmental milestones of postural reflex and motor development. In early infancy primitive postural reflexes appear and soon disappear and new postural reactions appear with age.

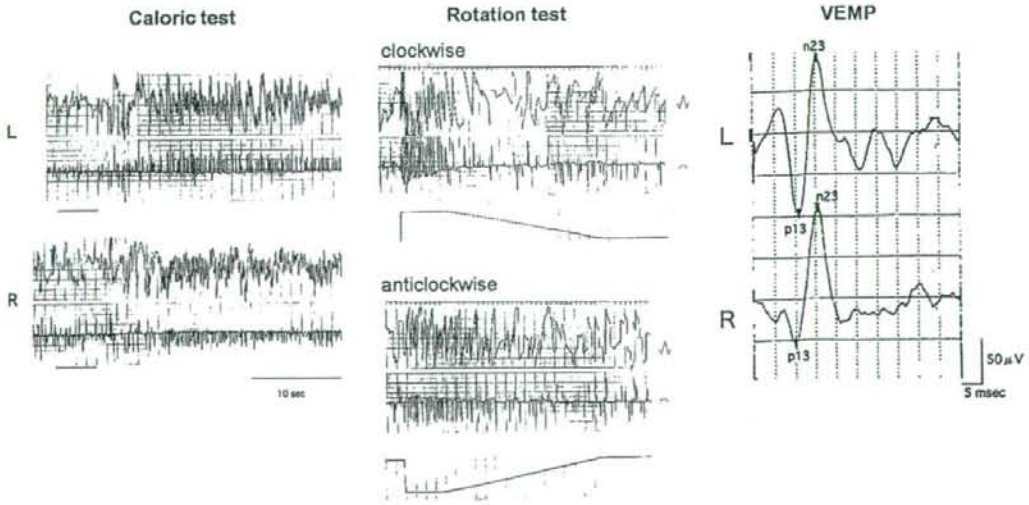

Figure 5. Recordings of a typical case with normal responses.

Table 1. Age of acquisition of motor milestones in infants and 3 to 10-year-old children with hypoactivity of vestibular end organs.

Subjects	*Head control (months)*	*Crawling (months)*	*Standing with support (months)*	*Independent walking (months)*
Control (n = 6)	3–4	7–10	10–11	10–12
Group 1a (n = 8)	4–8	7–14	9–16	17–27
Group 1b (n = 7)	4–16	12–28	15–348	24–48
Group 2a (n = 4)	3–7	8–24	9–20	12–33
Group 2b (n = 4)	8–24	15–30	27–36	44–54

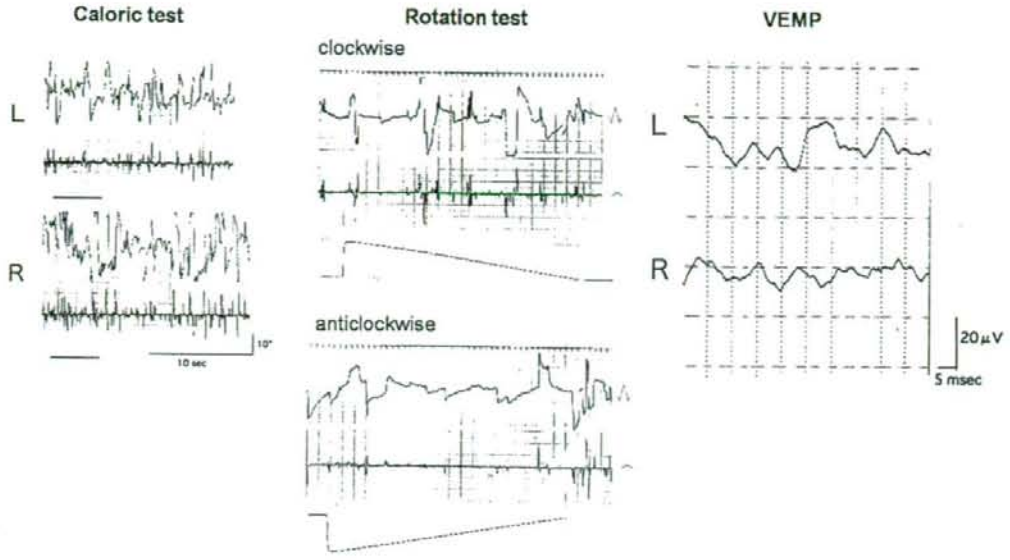

Figure 6. Recordings of a typical case with absent responses.

and lateral vestibulospinal tracts (Figure 8). However, other sensory organs and development of central motor systems in the brain compensate for this loss of peripheral vestibular function.

Influence of mental retardation on developmental milestones in children with congenital vestibular failure

Figure 9 shows the influence of mental retardation on locomotion in congenitally deaf infants with hypoactive and absent vestibular function. The bar chart shows average ages of deaf infants with normal intelligence, and a dot shows distribution of each case with mental retardation. Figure 9 reveals that mental retardation is correlated with a higher incidence and greater degree of delayed gross motor development.

Group 1b

Vestibular hypoactivity with mental retardation and without inner-ear anomalies: The age of acquisition of gross motor functions is compared with group 1a (Table 1). It is clear that the age of acquisition in this group is markedly older than in group 1a.

Group 2b

Vestibular hypoactivity with mental retardation and without semicircular canals because of inner-ear anomaly. The age of

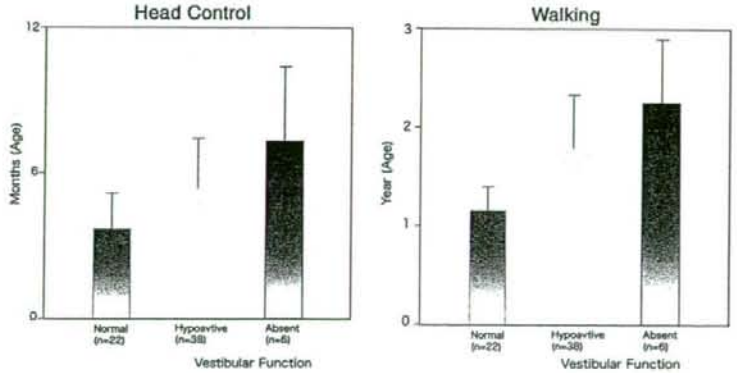

Figure 7. Age of acquisition of head control and walking in congenitally deaf infants with hypoactive and absent vestibular functions compared with controls.

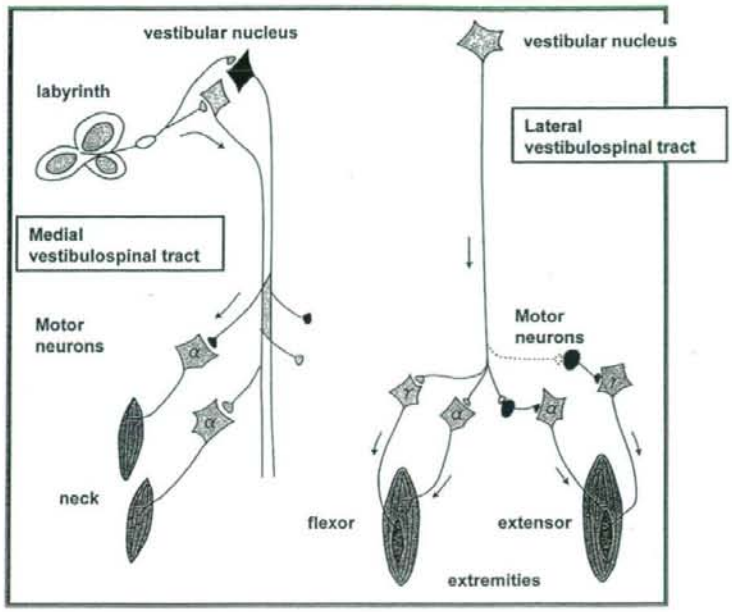

Figure 8. Neuronal circuits of labyrinthine vestibular reflex in brainstem.

acquisition of gross motor functions is compared in Table 1 and is shown to be markedly older in this group than in group 2a, but close to that in group 1b.

This comparison reveals that intellectual development is another important factor facilitating central vestibular compensation of infants with congenital vestibular failure (Kaga, 1988, 1999).

Case study I. Influence of blindness on congenital vestibular failure

Vision plays a very important role in stabilizing gaze and maintaining balance of the body. Patients with low vision or blindness, in addition to vestibular failure, have great difficulty in acquiring central vestibular compensation and both in darkness and in light.

Two congenitally deaf infants illustrate the influence of blindness on development of the postural reflex.

Case 1 is a two-year-old boy with opisthotonus-like persistent retroflexion of the head with congenital anophthalmia and deafness. It was not possible to perform the caloric test. In this case, VEMP revealed that saccular function was well preserved (Figure 10). It is possible that semicircular canal function was lost because of long-lasting opisthotonus-like head retroflexion. Central compensation was not good because he could not maintain body balance and did not walk.

Case 2 is a two-year-old girl with congenital blindness and deafness with loss of vestibular function which was revealed by the caloric test, the damped rotation test, and VEMP (Figure 11). In this case, long-lasting opisthotonus-like head retroflexion continued and delayed motor development was marked, with slow central vestibular compensation.

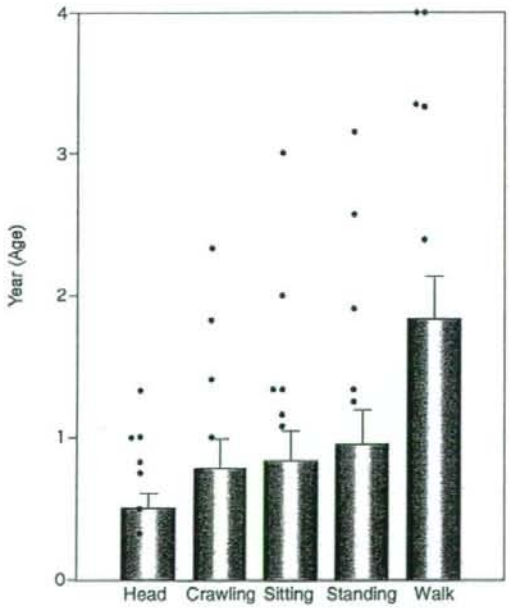

Figure 9. Influence of mental retardation (illustrated by dots) on age of acquisition of head control and walking. The bars show average ages of head control and walking in cases of congenital deafness and vestibular hypofunction with normal intelligence.

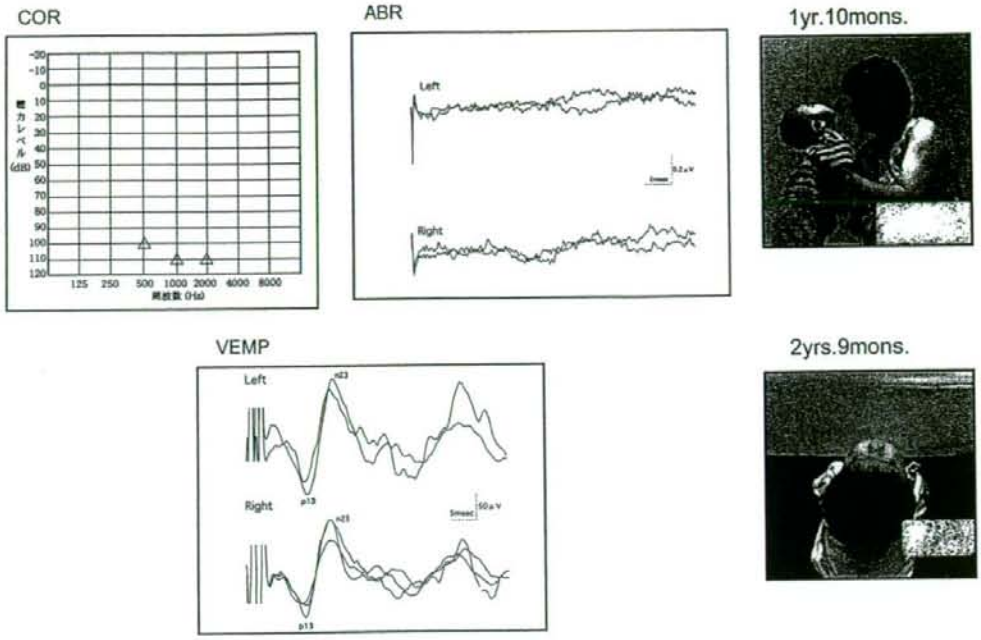

Figure 10. Case 1. A two-year-old deafblind boy.
COR: Conditioned orientation reflex audiometry.
ABR: Auditory brainstem response.
VEMP: Vestibular evoked myogenic potential.

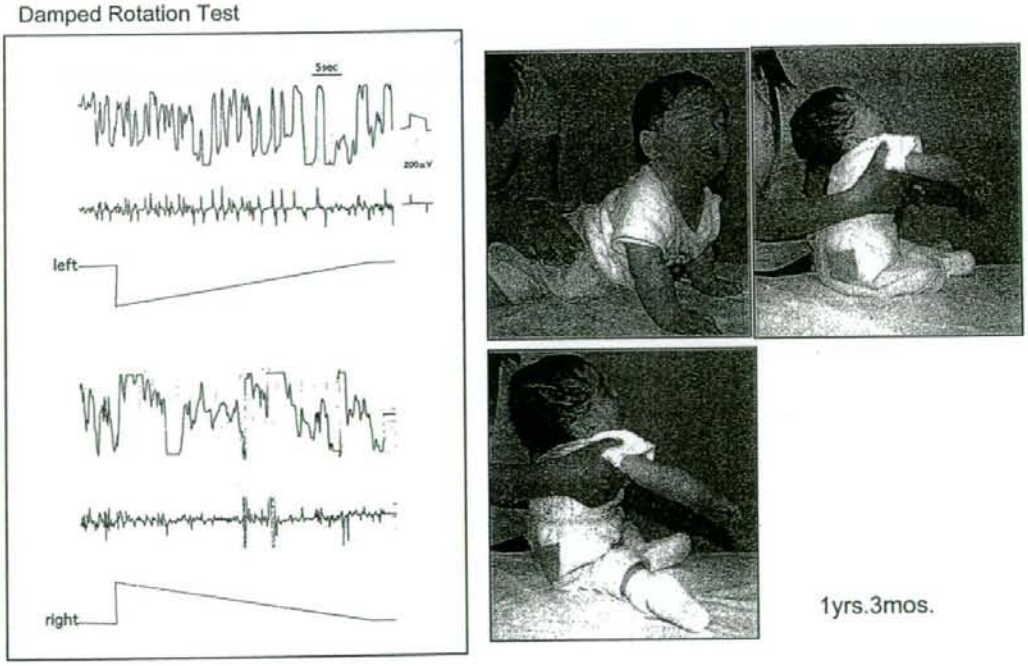

Figure 11. Case 2. A two-year-old deafblind girl with vestibular failure.

In these cases, vision is shown to play a very important role in acquiring central vestibular compensation in congenitally deaf infants with vestibular failure.

Case Study II. Final acquisition of sports activities after overcoming congenital vestibular failure. Skillfulness of exercises and sporting activities in young adults with congenital vestibular failure

In this final section, the ability of congenitally deafened young adults with vestibular failure to learn and acquire most physical and sporting skills, as a result of central vestibular compensation in development and growth, is emphasized.

We show photographs of exercises and sporting activities in two boys after central vestibular compensation.

Case 1

This boy has vestibular loss due to congenital inner-ear anomaly with common cavity only. In infancy, his head control and walking age were markedly delayed. However, in adolescence, he learned to swim and dive under water in spite of the absence of vestibular function. Moreover, he could play and enjoy most physically taxing sports (Figure 12).

Case 2

A congenitally deafened boy with vestibular loss in childhood. He was misdiagnosed as 'brain damaged' because his motor development was delayed in the first and second year of his life.

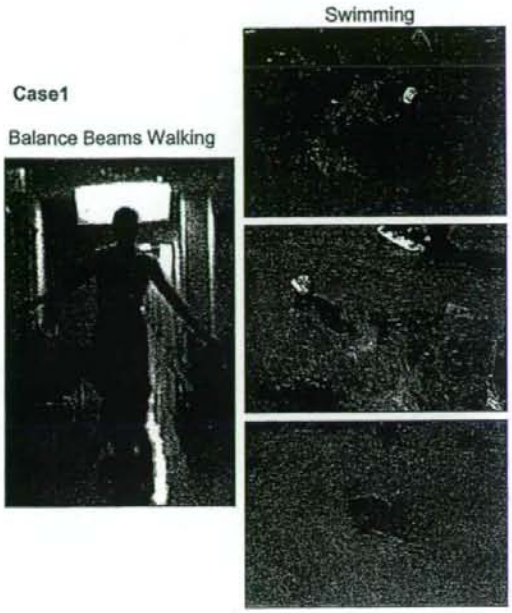

Figure 12. Case 1. A young adult with congenital deafness and vestibular loss due to congenital inner-ear anomaly swimming, and walking on a beam.

Figure 13. Case 2. A young adult with congenital deafness and vestibular failure riding a bicycle without holding the handlebars, skiing, and ski jumping.

However, he could play various sports at school age. By the age of twenty, he can enjoy skiing, ski jumping, and riding bicycles without holding on with both hands, which requires higher balance functions (Figure 13).

Eventually, these two cases overcame vestibular failure.

The evidence to date would suggest that the ceiling for central compensation in cases with congenital vestibular failure may be unlimited, but more longitudinal follow-up studies must be undertaken.

Conclusion

In general, congenitally deaf children with hypoactive or absent vestibular function are frequently misdiagnosed with psychomotor retardation or brain damage. However, in our studies, we conclude that in congenitally deaf children with vestibular loss, central vestibular compensation can overcome delayed motor function very well, such that the children may develop exercises and sports activities with great skill.

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Declaration of interest: The authors report no conflicts of interest. The authors alone are responsible for the content and writing of the paper.

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ORIGINAL ARTICLE

Vestibular evoked myogenic potentials evoked by multichannel cochlear implant – influence of C levels

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Abstract

Conclusions. This study showed that vestibular evoked myogenic potentials (VEMPs) evoked by cochlear implant (CI), could be related to the comfortable level (C level), particularly in the channels that are closer to the apical turn of the cochlea. *Objective.* The purpose of this study was to investigate the correlation between VEMPs and C level of each channel. *Subjects and methods.* We investigated 24 children who underwent cochlear implantation. VEMPs were recorded from the operated ears with the CI switched 'off' or 'on'. To investigate the correlation between VEMPs and C level, we selected 13 patients with Nucleus 24 (SPrint), and divided them into group A (normal VEMPs) and B (absence of VEMPs). In these children, all the 22 electrodes were active, and were mapped in the same frequency range for each channel. *Results.* Twenty children (83%) showed no VEMPs with the CI 'off'. Among them, 10 elicited VEMPs with the CI 'on', but the other 10 did not. In all channels, the mean C levels of CI were higher in group A than in group B. The p values in channels 1–12 were >0.10, in channels 13–16 were 0.06–0.09, and in channels 17–22 were 0.05–0.06, which were lower but not statistically significant.

Keywords: *Children, cochlear implant (CI), VEMPs, electrical stimulation, inferior vestibular nerve, C level*

Introduction

In patients with a multichannel cochlear implant (CI), electrical stimulation may not only stimulate the cochlear nerve, but may also partially spread to the facial nerve [1,2] or vestibular nerve [3]. Vestibular evoked myogenic potentials (VEMPs) are considered to be useful for evaluating the functions of the saccular [4], inferior vestibular nerve [5] and the tonus of the sternocleidomastoid muscle (SCM). In our previous study, we reported that there were no VEMPs during CI stimulation in eight children, but there were VEMPs in the other four children [3]. However, the differences in VEMPs in that study were still unclear.

The surgical procedures, CI devices, current intensity, stimulation schemes and testing paradigms used may be important for eliciting VEMPs. In this study, we selected 13 children with Nucleus 24 (SPrint), all the 22 electrodes of which were active and mapped in the same frequency range, and investigated the correlation between VEMPs and the C level of each channel.

Subjects and methods

Subjects

The patients were 24 children who underwent CI surgery at the University of Tokyo Hospital. The mean age at VEMP recording was 5.4 ± 3.0 years (range 2–14 years). All the patients had normal inner ear structures, as demonstrated by computed tomography (CT) of the temporal bone. All the children showed improved hearing after surgery.

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Procedures and VEMP recordings

The children were placed in the supine position. The active electrode was placed on the upper half of SCM, the reference electrode on the upper sternum and the ground electrode on the midline of the forehead. During VEMP recording, the children were instructed to lift their head up or to turn their head to the contralateral side to induce hypertonicity in the SCM.

The electromyography signal from the stimulated side was amplified and averaged using a Neuropack evoked-potential recorder (Nihon Kohden Co. Ltd, Tokyo, Japan). Electromyographic activities at a constant level were recorded for each child. The head-phone was placed over the microphone of the CI behind the ear. Rarefaction clicks (0.1 ms; 95 dB normal hearing level) were presented through the head-phone (type DR-531; Elega Acous Co. Ltd, Tokyo, Japan) and were used to evoke the VEMPs. The stimulation rate was 5 Hz, the band-pass filter intensity was in the range of 20–2000 Hz and the analysis time was 50 ms. VEMPs in response to 100 stimuli were averaged twice. After the CI surgery, the VEMPs of all the 24 children were recorded from the operated ear with the CI switched 'off' or 'on'.

Definition of VEMP

We classified VEMP recordings into three types as follows. Type 1, normal: the amplitudes of VEMPs were >50 μV. Type 2, small: the amplitudes of VEMPs were higher than 0 μV, but lower than 50 μV. Type 3, absent. Types 2 and type 3 were regarded as abnormal VEMPs.

Characteristics of CI device

The characteristics of CI devices are summarized in Table I.

CI device. All patients received multichannel co-chlear implants; 21 children had Nucleus 24 and 3 children had Nucleus 22.

Speech coding strategy. All patients with Nucleus 24 were coded with ACE and all patients with Nucleus 22 were coded with SPEAK.

Speech processor. Two patients used ESPrit, and one child used Spectra. Among patients with Nucleus 24, 18 patients used SPrint, one used ESPrint and 2 used ESPrit 3G. Among patients with Nucleus 24, the electrodes of all the channels were active in

Table I. Profile of patients.

Patient no.	Sex	Age (years)	CI type	Speech processor	Strategy	Active electrode number	VEMP	
							Off	On
1	M	2	24M	SPrint	ACE	22	–	+
2	F	2	24M	SPrint	ACE	22	–	+
3	F	4	24M	SPrint	ACE	22	Small	+
4	F	5	24M	SPrint	ACE	22	–	+
5	F	7	24M	SPrint	ACE	22	–	+
6	M	3	24M	SPrint	ACE	22	Small	+
7	M	3	24M	SPrint	ACE	22	Small	Small
8	M	2	24M	SPrint	ACE	22	Small	Small
9	M	6	24M	SPrint	ACE	22	–	–
10	F	4	24M	SPrint	ACE	22	–	–
11	M	6	24M	SPrint	ACE	22	–	–
12	F	4	24M	SPrint	ACE	22	–	–
13	F	5	24M	SPrint	ACE	22	–	–
14	F	3	24M	SPrint	ACE	22	–	–
15	F	3	24M	SPrint	ACE	22	–	–
16	F	7	24M	SPrint	ACE	22	–	–
17	M	4	24M	SPrint	ACE	21	–	–
18	F	3	24M	SPrint	ACE	18	–	+
19	F	11	24M	ESPrit	ACE	20	–	+
20	M	7	24M	ESPrit 3G	ACE	20	–	+
21	M	7	24M	ESPrit 3G	ACE	20	–	–
22	F	9	22M	ESPrit 22	SPEAK	18	–	+
23	M	14	22M	ESPrit 22	SPEAK	17	–	+
24	F	8	22M	Spectra	SPEAK	16	–	+

16, and electrodes of a part of channels were active in 5 patients.

Correlation between VEMP and C level

To investigate the correlation between VEMP and C level, we selected patients with the same CI device, coded with the same strategy and programmed with the same frequencies in each channel. All of these patients had Nucleus 24 CI devices (SPrint) and were coded with ACE strategy, and programmed with the same frequency ranges in each channel. In these patients, the electrodes of all the channels were active. We divided these patients into group A (normal VEMP; $n=6$) and group B (absence of VEMP; $n=7$) when the CI was switched on. Groups A and B were compared in terms of the C levels of each channel. However, two patients with small VEMPs were excluded, and one patient who used a different stimulation rate was also excluded from this study.

Procedures for obtaining C level data

MAPs were created by audiologists for each subject using Cochlear Corporation's Nucleus R126 ver.2.1. All MAP data of Nucleus 24 were created using ACE of monopolar stimulation at a duration of 25 μs/phase. The default stimulus for the ACE processor is a 500 ms burst of 250 Hz biphasic pulse train. The 'current level' used in both the NRT software (for masker and probe levels) and the R126 programming software were represented in units that vary from 1 to 255 and span a nominal range of approximately 10.2 μA to 1.75 mA, respectively. To avoid confusion between actual current level of microamps and Cochlear Corporation's 'current level' units, we refer to the stimulus level in 'programming units', which correspond to Cochlear Corporation's 'current level' units [6].

The child would indicate threshold and the maximum stimulus level that was comfortable for each electrode stimulated. For younger children (approximately aged 2–6 years), minimal response levels were obtained using play audiometry. These levels were used to measure T level. T level would be set at a level that evoked a consistent change in behaviours, such as quieting or head turn. At connection, C levels were typically set from above T level uncomfortable level for each of the three or four MAPs loaded onto the speech processor. The child was sent home wearing the softest MAP, and the parents were instructed to work their child through each of the progressively louder programs during the first few weeks of implant use until they returned for the next programming session. This was done to ensure that the initial MAP would be comfortable for the first day of use. To investigate the correlation between VEMP and C level, we selected the data of C level at the same time as VEMP recording.

Statistical analysis

Data were analysed statistically using unpaired t test. A difference was considered to be statistically significant at $p<0.05$.

Results

VEMPs appearance rate with CI off and on

When patients were tested with CI off, VEMPs were absent in 20/24 and small in 4/24 children. When patients were tested with CI on, VEMPs were normal in 12/24, small in 2/24 and absent in 10/24 children (Figure 1).

Comparison of VEMPs in the same patients with CI off and on

The comparison showed the following results: 10/24 children among those with absent VEMPs with CI off showed VEMPs with CI on (Figure 2); 10/24

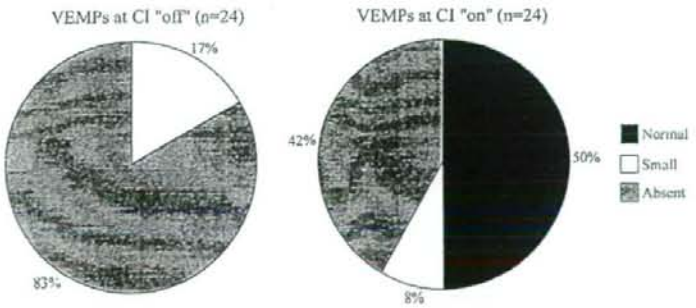

Figure 1. VEMPs appearance rate with CI off and on ($n=24$). Left: VEMPs with CI off ($n=24$). Right: VEMPs with CI on ($n=24$).

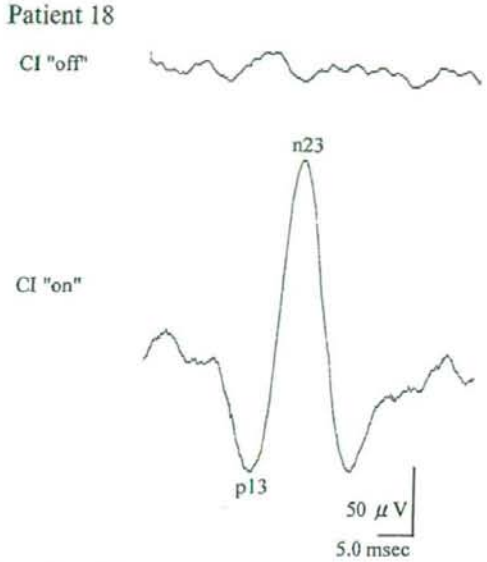

Figure 2. VEMPs of patient 18. This patient was fitted with a Nucleus 24 (SPrint), coded with ACE strategy. Electrode numbers 1–4 were not active, but numbers 5–22 were active. Top: absent VEMPs with CI off. Bottom: normal VEMPs with CI on.

among those with absent VEMPs with CI off showed absent VEMPs with CI on; 2/24 among those with small VEMPs with CI off showed VEMPs with CI on; 2/24 among those with small VEMPs with CI off showed small VEMPs with CI on.

The changes in VEMP amplitude before and after turning CI on are summarized in Figure 3. The mean amplitude with CI off and CI on were 6.5± 15.4 and 83.9±102.9 μv, respectively.

Effect of C level on VEMPs

The mean time lapse after CI surgery in groups A ($n=6$) and B ($n=7$) were 13.3±17.7 and 11.1± 10.1 months, respectively. There was no significant difference in the time lapse after CI surgery between groups A and B ($p=0.7843$).

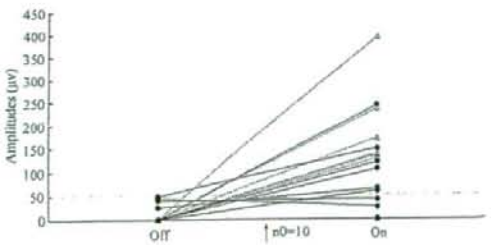

Figure 3. Changes in VEMP amplitude with the CI on ($n=24$). Solid circle, patients have Nucleus 24 and all the 22 electrodes were active; empty triangle, other kinds of patients. n0 =10: absent VEMPs with both CI off and on ($n=10$).

The VEMPs in groups A and B are illustrated in Figure 4.

The lower and upper frequencies in each channel in group A (normal VEMP) and group B (absence of VEMP) are illustrated in Figure 5a.

Mean C levels in each channel in group A and B are illustrated in Figure 5b. In all channels, each mean C level was higher in group A than in group B. The p values in channels 1–12 were >0.10, in channels 13–16 were 0.06–0.09, and in channels 17–22 were 0.05–0.06. Then, it was revealed that the C level that elicited VEMPs in group A was higher than in group B. The C level was higher than among those who had normal VEMPs (with CI on) and those who had no VEMPs, due to differences in channels 17–22.

Discussion

As regards saccular function after CI surgery, the insertion of an intrascalar electrode array causes immediate damage to the inner ear [7,8] and over time may cause additional changes, which can interfere with neuronal stimulation [9,10]. The postoperative dysfunction of the vestibular system in multichannel CI patients has been reported, such as dizziness [11], and reduction of caloric responses [12], and a vestibule-ocular response to rotation chair [13]. A decrease in VEMPs after CI surgery has also been reported [3,14]. In our previous study, we compared the VEMPs of the operated ear before and after CI surgery, and found that in 7 of the 12 children (58%), the VEMPs were abolished postoperatively without CI stimulation [3]. Ernst et al. demonstrated that VEMPs induced by bone-conducted acoustic stimuli are absent in 36% of patients before implantation, and that this percentage increases to 78% after the CI surgery [14]. These reports suggested that the saccule of these children must have been damaged after CI surgery in the operated ear.

In this study, we found that 20 patients (83%) did not show VEMPs without CI stimulation after surgery. This result is in agreement with those of previous studies [3,14]. In a pathological study of the human temporal bone, saccule collapse occurred in the operated ear [15,16], but the hair cell densities were the same as those in the non-operated ear [15]. Direct damage to the ductus reunions [17] or cochlear duct as a result of it being blocked externally by fibrous tissue or bone and internally by debris may result in saccule collapse [15]. Our

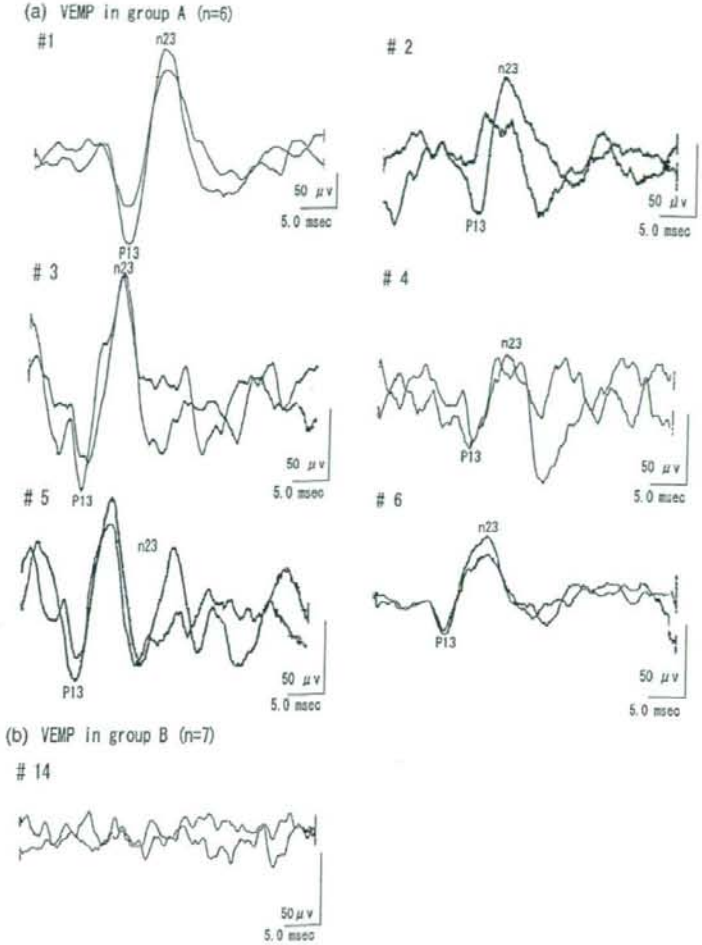

Figure 4. VEMPs in groups A and B. (a) VEMPs in group A ($n=6$); (b) VEMPs in group B ($n=7$).

results also revealed that the functions of the saccule are lost after CI surgery.

Galvanic stimulation of the mastoid can stimulate the vestibular nerve [18,19], and elicit a sway response [20], eye movement [21] and VEMPs (galvanic VEMPs) [22,23]. Watson et al. reported that galvanic VEMPs are abolished by selective vestibular nerve section [22]. These studies suggest that galvanic VEMPs can stimulate the vestibular nerve and vestibular nucleus and elicit myogenic potentials in the SCM.

In our study, 20 children (83%) showed no VEMP response with the CI off, but 10 of these patients showed normal VEMP responses with the CI switched on. With the CI off, children with a CI were given only acoustic stimulation, but with the CI on, the acoustic stimulation changed to electrical stimulation. The VEMPs evoked by CI must be caused by the electrical stimulation of CI, which probably stimulates the inferior vestibular nerve. A human temporal bone pathological study demonstrated that Scarpa's ganglion cell count in the operated ear was the same as that in the non-operated ear [15]; however, collapse was only seen in the operated ear [15,16]. It is suggested that the vestibular nerve must be intact, although the functions of the saccule were lost in CI patients.

The basilar membrane of the inner ear is responsible for analysing the input signals into different frequencies. Low-frequency sounds create travelling waves in the fluid of the cochlea that cause the basilar membrane to vibrate, with the largest

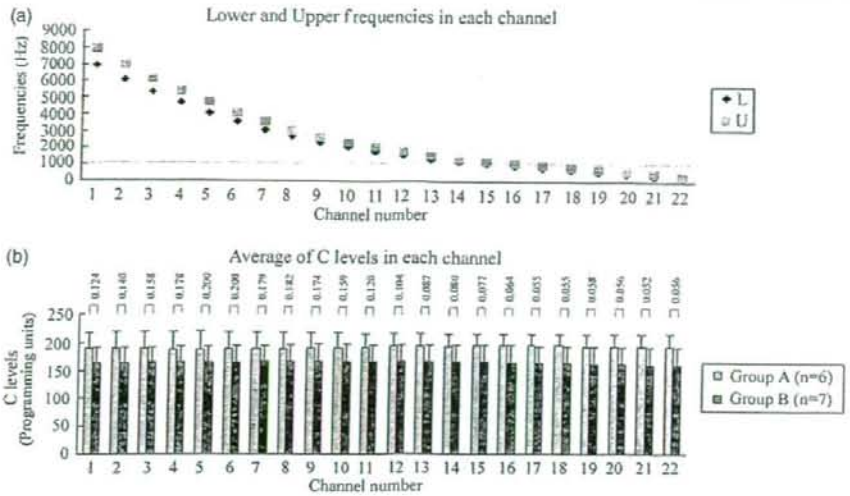

Figure 5. Average of C levels in each channel in group A ($n=6$) and B ($n=7$). (a) Lower and upper frequencies in each channel; L, lower frequency; U, upper frequency. (b) Average of C levels in each channel in group A ($n=6$) and B ($n=7$).

amplitude of displacement occurring at the apex of the basilar membrane. On the other hand, high frequency sounds create travelling waves with the largest amplitude of displacement occurring at the base of the basilar membrane. If the signals are composed of multiple frequencies, then the resulting travelling wave will show its maximum displacement at different points along the basilar membrane. This place theory for coding frequencies has motivated scientists to consider studying multichannel CIs and VEMPs. In multichannel CIs, the electrodes are located at different distances from the round window, and every channel shows different sensitivities to frequencies.

This study demonstrated that the C levels of CI were higher in group A than in group B. VEMPs stimulated by clicks are more sensitive to higher intensities, and the amplitudes of VEMPs increase with stimulus intensity [24,25]. The amplitudes of galvanic VEMPs also increase with current intensity [26]. It is suggested that the amplitudes of VEMPs are related to the current intensity of the C level.

In this study, the C level was higher among those who had VEMP (with CI on) than among those who had no VEMP, because of differences in channels 17–22 ($0.05 < p < 0.06$). The frequencies of channels 17–22 were 188–938 Hz. This difference in frequency range revealed that VEMPs evoked by CIs are easily elicited in the low frequency range. McCue and Guinan [27] reported that sound-sensitive vestibular afferents showed a broad, V-shaped tuning curve, with the ideal frequencies in the range of 500–1000 Hz. VEMPs evoked by short tone bursts (STBs) of 500 Hz, are used in clinical tests. These suggest that the saccular macula has its preferred stimulus frequency in the low frequency range. Our results suggest that the inferior vestibular nerve could be more sensitive to lower frequency ranges. The anatomic factor should be also considerable. Our results demonstrated that the p values in channels 1–12 were >0.10, in channels 13–16 were 0.06–0.09, and in channels 17–22 were 0.05–0.06. Channels 17–22 and channels 13–16 were further away from the inferior vestibular nerve endings than channels 1–13, so it may be that higher current intensities are needed to stimulate the inferior vestibular nerve in channels 17–22. This suggests that VEMPs evoked by CIs were correlated with C level, especially in the channels that are closer to apical turn of the cochlea.

In our study, 10 of the 24 patients showed no VEMP responses with the CI on. These patients may require higher current intensities to elicit VEMP responses. However, in children, it is difficult to increase the current intensity. Children feel pain or facial nerve stimulation when the current intensities are increased higher than those of the C level.

In conclusion, the presence or absence of VEMPs in children with CIs depends on the larger current intensities of the C level, particularly in the channels that are closer to the apical turn of the cochlea.

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