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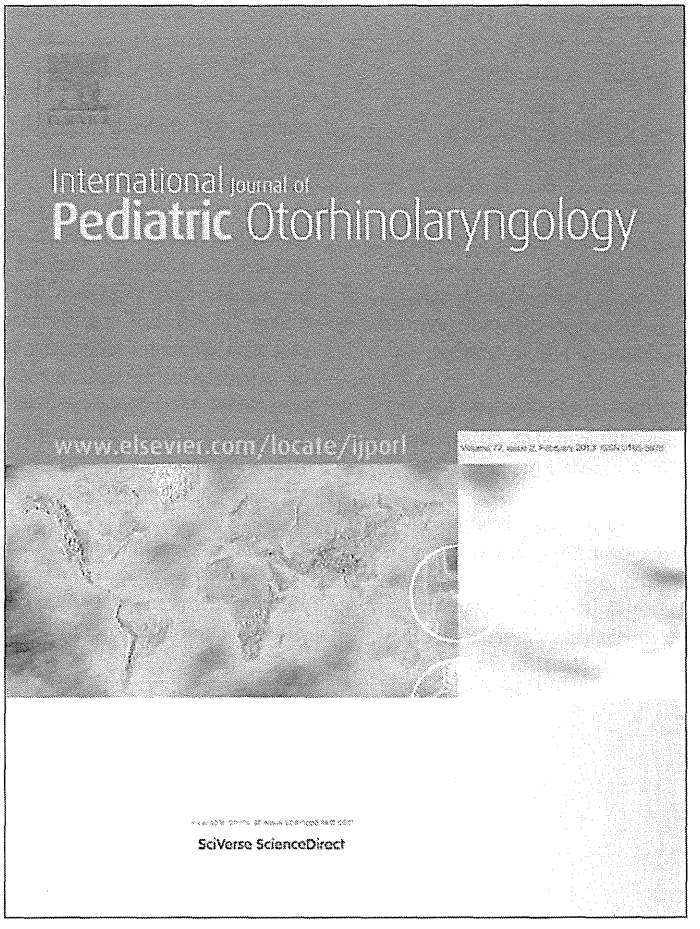

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High prevalence of inner-ear and/or internal auditory canal malformations in children with unilateral sensorineural hearing loss

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ABSTRACT

Objective: Radiological and genetic examination has recently advanced for diagnosis of congenital hearing loss. The aim of this study was to elucidate the prevalence of inner-ear and/or internal auditory canal malformations in children with unilateral sensorineural hearing loss (USNHL) for better management of hearing loss and genetic and lifestyle counseling.

Methods: We conducted a retrospective study of charts and temporal bone computed tomography (CT) findings of 69 consecutive patients 0–15 years old with USNHL. In two cases, genetic examination was conducted.

Results: Of these patients, 66.7% had inner-ear and/or internal auditory canal malformations. The prevalence of malformations in infants (age <1 year) was 84.6%, which was significantly higher than that in children 1–15 years old (55.8%; $p < 0.01$). Almost half of the patients (32; 46.4%) had cochlear nerve canal stenosis; 13 of them had cochlear nerve canal stenosis alone, and in 19 it accompanied other malformations. Internal auditory canal malformations were observed in 22 subjects (31.8%), 14 (20.3%) had cochlear malformations, and 5 (7.2%) had vestibular/semicircular canal malformations. These anomalies were seen only in the affected ear, except in two of five patients with vestibular and/or semicircular canal malformations. Two patients (2.9%) had bilateral enlarged vestibular aqueducts. Mutations were found in *SLC26A4* in one of the two patients with bilateral large vestibular aqueducts. The prevalence of a narrow internal auditory canal was significantly higher in subjects with cochlear nerve canal stenosis (50.0%) than in subjects with normal cochlear nerve canals (11.1%; $p < 0.01$). There were no correlations between the type and number of malformations and hearing level.

Conclusions: The prevalence of inner-ear and/or internal auditory canal malformations detected by high-resolution temporal bone CT in children with USNHL was very high. Radiological and genetic examination provided important information to consider the pathogenesis and management of hearing loss. Temporal bone CT should be recommended to children with USNHL early in life. *SLC26A4* mutation also should be examined in cases with bilateral enlarged vestibular aqueduct.

1. Introduction

Abnormalities of the temporal bone have been associated with congenital sensorineural hearing loss (SNHL) since reported by Mondini in 1791 [1]. However, most cases of congenital SNHL were believed to be caused by abnormalities of the membranous labyrinth that could not be detected by conventional imaging techniques [2,3]. Conventional computed tomography (CT) could identify congenital cochlear malformations such as complete labyrinthine aplasia (Michel deformity), a common cavity, cochlear aplasia/hypoplasia, and incomplete partition [2–4]. Because of improvements in high-resolution CT techniques, previously unrecognized bony abnormalities—including a large vestibular aqueduct, wide and stenotic internal auditory canal (IAC), and cochlear nerve canal (CNC) stenosis—have been reported [3,5]. Currently, abnormalities found by imaging techniques not only provide diagnostic information but also aid in genetic and lifestyle counseling [1] and guide clinicians to better management of hearing loss [6].

The aim of this study was to elucidate the prevalence of inner-ear and/or IAC malformations in children with unilateral SNHL (USNHL).

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2. Patients and methods

We conducted a retrospective study of charts and temporal bone CT findings of consecutive USNHL patients 0–15 years old who were seen in the Department of Otorhinolaryngology of National Mie Hospital between January 2008 and December 2011. All procedures were approved by the Ethics Review Committee of National Mie Hospital.

2.1. Subjects

The study included 69 patients. USNHL was defined as a hearing threshold greater than 30 dB hearing level for at least one frequency (500–2000 Hz). Of the 69 patients, 32 were male and 37 were female. Their ages of diagnosis ranged from 0 to 15 years (mean ± 1 SD: 4.3 ± 6.7 years, median: 4 years). The distribution of age was shown in Fig. S1. Twenty-six (37.3% of the subjects) were infants less than 1 year old. Twenty-two children had failed newborn hearing screening (NBHL) in unilateral ear and 21 of them identified USNHL in 1 year of age. One boy who had failed NBHL first visited ENT clinic and diagnosed USNHL at the age of 3 years. There was neither subjects who passed NBHL nor ones who suspected progressive hearing loss before their diagnosis. One subject had Down's syndrome and one had tetralogy of Fallot. Patients with middle ear diseases and abnormalities, conductive and combined hearing loss revealed by pure-tone audiometry, and obvious acquired hearing loss were excluded from the study.

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2.2. Audiometric evaluations

Severity of hearing loss was defined from the pure-tone average as follows: hearing level of 21–40 dB, mild; 41–70 dB, moderate; 71–95 dB, severe; and greater than 95 dB, profound [7]. Pure-tone average was defined as the average hearing threshold at 500, 1000, and 2000 Hz. Thirty-four patients in this study were too young to be examined with pure-tone audiometry initially; for these patients, USNHL was determined on the basis of auditory brainstem response (ABR) and auditory steady state response (ASSR) using an Audera® system (Grason-Stadler). Distortion product otoacoustic emissions (DPOAE) and tympanometry were performed for all subjects.

2.3. Evaluation of temporal bone CT findings

All the patients underwent high-resolution CT of temporal bone using a single-slice helical CT (HiSpeed DX/i, GE Healthcare Japan Ltd., Tokyo Hino, Japan). Contiguous 1 mm-thick sections parallel to the infraorbitomeatal line were acquired through the temporal bone, with a field of view of 230 mm, matrix size of 512 × 512, in-plane pixel size of 0.45 mm × 0.45 mm, tube voltage of 120 kV, tube current of 150 mAs and a reconstruction kernel for bone.

CT results for each patient were examined by two otologists who did not know which ear had hearing loss. Classification of inner-ear and IAC malformations was based on Sennaroglu's classification [4] and modified as follows:

1. Cochlear malformations: Michel deformity, cochlear aplasia, common cavity deformity, cochlear hypoplasia, incomplete partition type I (IP-I), incomplete partition type II (IP-II: Mondini deformity).
2. Vestibular/semicircular canal malformations: absent vestibule, hypoplastic vestibule, dilated vestibule/absent semicircular canal, hypoplastic semicircular canal, enlarged semicircular canal.
3. IAC malformations: absent, narrow, enlarged.
4. Vestibular aqueduct finding: large.
5. CNC finding: stenosis.

We defined IAC as narrow when the diameter at the level of the porous of the IAC was less than 3 mm or 2 mm smaller than the normal side and as wide when greater than 10 mm. A large vestibular aqueduct was defined as being greater than 1.5 mm at the midpoint of the vestibular aqueduct on axial images [8]. The width of the CNC was measured at its midportion. The measurements were manually obtained using calipers [5]. CNC stenosis was defined as when the width was less than 1.5 mm [9]. An example of CNC stenosis in the right ear is shown in Fig. 1.

2.4. Genetic examinations

Patients with large vestibular aqueducts participated in genetic examination. Blood samples were obtained from the proband and his/her parents. DNA was extracted from blood samples using the Gentra Puregene DNA isolation kit (Qiagen, Hamburg, Germany), and primers specific for *SLC26A4* (GenBank NG_008489) were designed. Primer sequences for *SLC26A4* are listed in Table S1, supporting information. Screening for *SLC26A4* mutations was performed by bidirectional sequencing of amplicons generated by PCR amplification of each exon (exons 1–21) and splice sites using an Applied Biosystems 3730 DNA Analyzer (Applied Biosystems, Foster City, CA, USA) and analyzed by SeqScape v2.6 (Applied Biosystems). Examinations were conducted only after written informed consent had been obtained from each individual or parents of the patients.

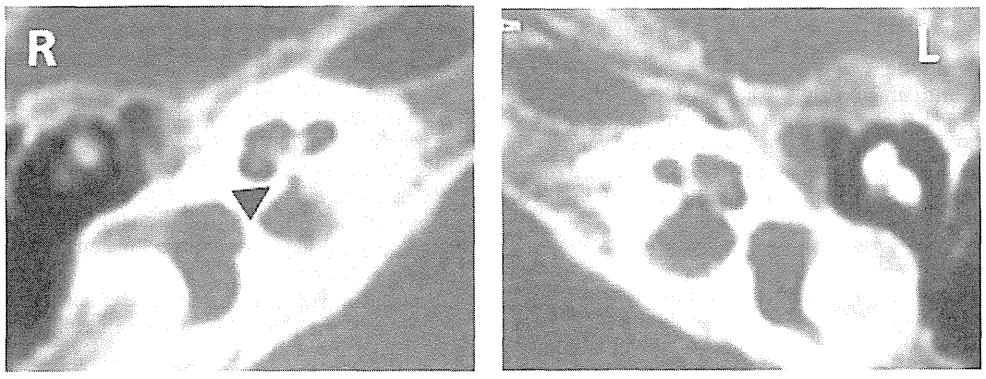

Fig. 1. Cochlear nerve canal stenosis demonstrated by transverse, thin-section CT scan of the temporal bone in three-month old boy. The left panel shows the hearing-impaired right ear (R), and the right panel shows the normal left ear (L). The arrowhead indicates the stenotic cochlear nerve canal in the right ear.

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2.5. Statistical analysis

The significance of the prevalence of the inner-ear and/or IAC malformations between infants younger than 1 year of age and children from 1 to 15 years of age, and the association between the existence of malformations and hearing level was determined by the χ^2 test.

3. Results

The prevalence of inner-ear and/or IAC malformations is shown in Fig. 2. Of the 69 subjects, 66.7% had malformations. The prevalence of malformations in infants younger than 1 year of age (84.6%) was significantly higher than that in children 1–15 years of age (55.8%; $p < 0.01$).

Table 1 shows the prevalence of each malformation. The most common anomaly was CNC stenosis of the affected ear, seen in 46.4% of the subjects. Next in frequency were IAC malformations, followed by cochlear malformations and vestibular and/or semicircular canal malformations. These anomalies were seen in the affected ear alone, except for two of five patients with vestibular and/or semicircular canal malformations. Two patients had bilateral enlarged vestibular aqueducts.

The combination of malformations we observed is summarized in Table 2. Of the 69 patients, 13 (18.8%) had CNC stenosis alone, 19 (27.5%) had CNC stenosis accompanied with other malformations, and 4 (5.8%) had narrow IAC alone. Two patients with bilateral enlarged vestibular aqueducts had cochlear or cochlear and vestibular/semicircular canal malformations. In both cases, unilateral hearing loss was found by newborn hearing screening. In one case, a 4-month-old boy, genetic examination identified a compound heterozygous mutation [p.T410M (c.1229C>T)/p.L743X (c.2228T>A)] in *SLC26A4* (Fig. S2). p.T410M was previously reported as a missense mutation [10] and p.L743X was previously reported as a nonsense mutation [11]. This nonsense substitution truncates the protein at codon 743, which is 38 amino acids from the end of the protein. This case was confirmed as Pendred syndrome. The hearing loss in his normal hearing ear developed at 1 year of age. In another case, a 2-month-old girl, pathological mutations were not found in *SLC26A4*. Her hearing level has been stable for 3 years.

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Table 3 shows the relationship between CNC malformations and IAC malformations. Of 32 cases of CNC stenosis, 16 (50.0%) were comorbid with narrow IAC. In 36 subjects with normal CNC, 4 (11.1%) had narrow IAC. The prevalence of narrow IAC was significantly higher in subjects with CNC stenosis than in subjects with normal CNC ($p < 0.01$).

Table 4 shows the combination of malformations and hearing level. There were 6 cases of mild hearing loss, 13 cases of moderate hearing loss, 7 cases of severe hearing loss, and 43 cases of profound hearing loss. DPOAE was absent in the affected ear in all subjects, except for two patients with unilateral profound hearing loss with CNC stenosis and narrow IAC without cochlear/vestibular/semicircular canal malformations. These two patients demonstrated normal responses in DPOAE in both ears. In one of these cases, ABR was performed. The threshold of wave V was 95 dBnHL (normal Hearing Level) in the affected ear and 20 dBnHL in the normal ear. This case was confirmed as unilateral auditory

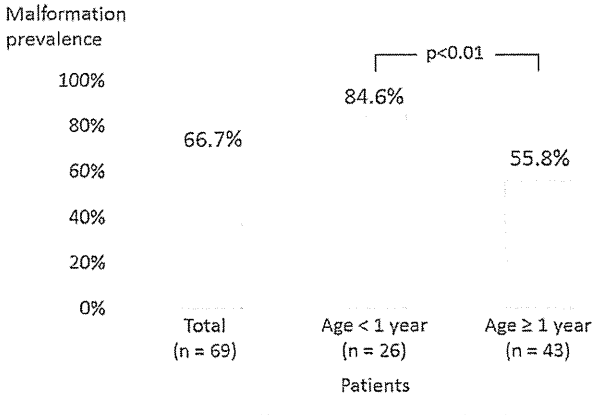

Fig. 2. Prevalence of inner-ear and/or internal auditory canal malformations found by temporal bone computed tomography.

Table 1
Prevalence of each malformation.

Malformation	Number (prevalence)
Cochlea	14 (20.3%)
Cochlear aplasia	0
Common cavity deformity	2 (2.9%)
Cochlear hypoplasia	1 (1.4%)
Incomplete partition (IP-I, IP-II)	11 (15.9%)
Vestibular/semicircular canal	5[a] (7.2%)
Internal auditory canal	22 (31.8%)
Narrow	20 (29.0%)
Enlarged	1 (1.4%)
Absent	1 (1.4%)
Vestibular aqueduct: enlarged (bilateral)	2 (2.9%)
Cochlear nerve canal: stenosis	32 (46.4%)

[a] Two cases had malformation in both ears.

Table 2
Combination of malformations.

Combination of malformations	Number (percentage)
CNC stenosis	13 (18.8)
CNC stenosis + narrow IAC	10 (14.5)
CNC stenosis + narrow IAC + C malformations	5 (7.2)
CNC stenosis + narrow IAC + V/SC malformations	1[a] (1.4)
CNC stenosis + C malformations	3 (4.3)
Narrow IAC	4 (5.8)
Large IAC	1 (1.4)
C malformations	2 (2.9)
C/V/SC malformations	2 (2.9)
V/SC malformations	2[b] (2.9)
Large VA + C malformations	1 (1.4)
Large VA + C malformations + V/SC malformations	1 (1.4)
CC with absent IAC	1 (1.4)
Normal	23 (33.3)
Total	69 (100.0)

CNC stenosis, cochlear nerve canal stenosis; IAC, internal auditory canal; C, Cochlear; V/SC, vestibular/semicircular canal; VA, vestibular aqueduct; CC: common cavity.
[a] This patient had bilateral V/SC malformations.
[b] One patient had bilateral V/SC malformations.

Table 3
Relationship between cochlear nerve canal malformations and internal auditory canal malformations.

Cochlear nerve canal		Internal auditory canal	
Stenosis	32 (46.4%)	Narrow	16 (50.0%)
		Normal	16 (50.0%)
Normal	36 (52.2%)	Narrow	4 (11.1%)
		Normal	31 (86.1%)
		Large	1 (2.8%)
Absent	1 (1.4%)	Absent	1 (100.0%)

Table 4
Combination of malformations and hearing level.

Cochlear nerve canal stenosis		+				–				Total
Narrow internal auditory canal		+		–		+		–		
Cochlear/vestibular/semicircular canal malformations		+	–	+	–	+	–	+	–	
Hearing level	Mild (21–40 dB)			1			1		4	6
	Moderate (41–70 dB)	1			3		1	4	4	13
	Severe (71–95 dB)	2			1			1	3	7
	Profound (>95 dB)	4[a]	10[b]	2	9		2	4	12	43
Total		7	10	3	13		4	9	23	69

[a] One patient had common cavity with IAC deficiency.
[b] Two patients demonstrated normal distortion product otoacoustic emissions.

neuropathy spectrum disorder. There were no correlations between the hearing level and the existence of CNC stenosis, narrow IAC, or other malformations in subjects with absence of DPOAE.

4. Discussion

The data in the present study showed a high prevalence of inner-ear and/or IAC malformations in pediatric USNHL. The prevalence was 84.6% in infants younger than 1 year of age. Most USNHL in these infants was considered as congenital, implying that more than 80% of the congenital USNHL was caused by morphological abnormality accompanied with bony anomalies.

The frequency of reported abnormal temporal bone findings in patients with USNHL varies from 7% to 44% [7]. Song et al. [8] studied CT of 322 children with USNHL and reported that 28.9% had malformations. Simons et al. [7] reported that the prevalence of CT abnormalities was 35% (29 of 83 cases), and the prevalence of magnetic resonance imaging (MRI) abnormalities was 25% (10 of 40 cases) in children with USNHL. However, they did not refer to the CNC.

The size of the CNC was first reported by Fatterpekar et al. in 2000 [5]. They demonstrated that the length and width of the CNC were significantly smaller ($p < 0.05$) in patients with congenital SNHL who had "normal" findings at thin-section temporal bone CT than in the control group. In 2008, Kono [3] investigated 118 patients without inner-ear malformations among 160 patients with USNHL, and 60% showed a significant difference in the CNC diameters between the affected and unaffected sides. Kono suggested that a diameter of less than 1.7 mm on transverse images or less than 1.8 mm on coronal images was hypoplasia. Stjernholm et al. [12] suggested that if the CNC diameter was less than 1.4 mm, then the possibility of cochlear nerve abnormality should be considered. Recent studies [9,13] demonstrated that CNC stenosis with a diameter of 1.5 mm or less as assessed with CT suggested cochlear nerve deficiency or hypoplasia as assessed with MRI. Wilkins et al. [14] showed a significant correlation between the degree of CNC stenosis and the degree of hearing loss. In the present study with the definition that the diameter was less than 1.5 mm, 46.4% of the subjects had CNC stenosis.

The exact cause of narrow CNC is unclear. Proper development of the IAC requires the presence of a normal cochlear nerve as a stimulus for attaining normal adult dimensions [5]. There is a possibility that the normal development of the CNC similarly needs the nerve for stimulus [5,15]. Fatterpekar et al. [5] speculated that, in patients with abnormality involving the membranous labyrinth, inhibition of the normal trophic effects of nerve growth factors owing to a diminutive cochlear nerve results in a small CNC. That is to say, hypoplasia of the CNC might be secondary to a hypoplastic cochlear nerve associated with some abnormality of a membranous labyrinth that could not be detected by current imaging techniques [3]. Very few of our subjects demonstrated a positive response in DPOAE, suggesting that at least the outer hair cells were affected or may not exist in most patients with USNHL.

The abnormalities found by imaging techniques provide information for diagnosis, management of hearing loss, and genetic and lifestyle counseling [1,6]. Congenital malformed inner ears may be associated with cerebrospinal fluid leakage, and thus development of meningitis is a very real possibility. Parents of children with inner-ear anomalies should be informed of the early symptoms and signs of meningitis. Consideration also should be given to immunization against common organisms implicated in meningitis [16]. Genetic examination should be recommended for patients with enlarged vestibular aqueducts. Pourova et al. [17] recommend performing SLC26A4 mutation analysis, following GJB2 analysis, in all hearing loss patients with bilateral enlarged vestibular aqueduct and/or associated thyroid impairment. Thy also mentioned that it is not reasonable to test the SLC26A4 gene in children with sporadic deafness without knowledge of their temporal bone CT/MRI images or even with its normal result. Mutations in the SLC26A4 are responsible for Pendred syndrome [18] as well as DFNB4 (non-syndromic hearing loss with inner ear abnormalities—enlarged vestibular aqueduct and/or Mondini deformity) [19]. Pendred syndrome and bilateral enlarged vestibular aqueduct correlates with the presence of two mutant alleles of *SLC26A4* [17,20,21]. Hearing loss in most patients with *SLC26A4* mutations fluctuates and is progressive [22]. Mutations in *SLC26A4* indicate the necessity for careful management of hearing and comorbidities, such as goiter.

The lack of MRI examination is one of the limitations in the present study. The results suggest the importance of temporal bone CT. Nevertheless, the risks of sedation/anesthesia for imaging in infants and young children, or indeed the radiation risk should be considered. The ideal imaging algorithm in children with unilateral or asymmetric SNHL is controversial [7]. MRI can detect soft-tissue abnormalities such as cochlear nerve deficiency with normal CNC and IAC. Simons et al. [7] suggested that virtually all children with SNHL should have an imaging study as part of their workup. They prefer high-resolution temporal bone CT as the initial study because of a high prevalence of positive findings and less cumbersome logistical issues. They also recommended that a negative CT scan should be followed by MRI to rule out SNHL caused by the central nervous system.

There are some other limitations regarding the current study. The first limitation is the diagnosis of SNHL. USNHL was determined on the basis of ABR and ASSR in 34 young patients. Middle-ear diseases and abnormalities were ruled out by CT and tympanometry; however, there is a possibility that some patients had conductive or combined hearing loss. Another limitation concerns the number of subjects. We examined 69 children, however, the evaluations should be need in the larger group.

In conclusion, a high prevalence of inner-ear and/or IAC malformations was detected by high-resolution temporal bone CT in children with USNHL. Radiological and genetic examination provided important information concerning the pathogenesis and management of hearing loss. The results of this study supported the recommendation of temporal bone CT to children with USNHL early in life. Genetic examination of *SLC26A4* also should be performed in all cases with bilateral enlarged vestibular aqueduct. The study in the larger group will likely refine the clinical protocol.

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ORIGINAL ARTICLE

Unilateral cochlear nerve hypoplasia in children with mild to moderate hearing loss

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Abstract

Conclusion: Even if hearing loss is mild to moderate, the presence of cochlear nerve (CN) hypoplasia associated with retrocochlear disorders should be considered. *Objectives:* CN hypoplasia is a term that refers to an absent cochlear nerve on high-resolution magnetic resonance imaging (MRI). Most cases of CN hypoplasia are associated with profound hearing loss. The present study reports six pediatric cases of unilateral CN hypoplasia with mild to moderate hearing loss. *Methods:* Between May 2008 and April 2011, pure-tone hearing tests were performed in 17 patients who were diagnosed with CN hypoplasia on high resolution for evaluation of unilateral sensorineural hearing loss at the National Center for Child Health and Development. Of these, six patients had average hearing levels in the affected ears of < 60 dB and were therefore included in this study. *Results:* All six ears with CN hypoplasia were associated with CN canal stenosis. DPOAEs were present in one (17%) of the six affected ears. The ABR thresholds of the ears with CN hypoplasia were significantly elevated compared with 1–4 kHz pure-tone hearing levels in one of three cases. In two of five cases, the maximum word recognition scores of the affected ears were poor compared with pure-tone hearing levels.

Keywords: *3-D constructive interference, steady-state magnetic resonance imaging, word recognition score, sensorineural hearing loss*

Introduction

Cochlear nerve (CN) hypoplasia refers to the absence of a visible CN on oblique sagittal magnetic resonance (MR) images of the lateral aspect of the inner auditory canal (IAC). CN hypoplasia is not an uncommon cause of congenital hearing loss as previously thought [1,2]. Although it is believed that most cases of CN hypoplasia are associated with profound hearing loss [1], a recent report presented a case of CN hypoplasia with moderate hearing loss limited to high frequencies [3]. The present study reports six pediatric cases of unilateral CN hypoplasia with mild to moderate hearing loss, and demonstrates that retrocochlear hearing loss is the predominant audiologic characteristic in these children.

Material and methods

Patient population

Between May 2008 and April 2011, 25 children who presented for evaluation of unilateral sensorineural hearing loss (SNHL) at the National Center for Child Health and Development were diagnosed with CN hypoplasia on high-resolution MR imaging (MRI).

Pure-tone hearing tests could be performed in 17 of these 25 children. Pure-tone audiometry was evaluated based on the three-tone average formulated by (a+b+c)/3, where a, b, and c are hearing levels at 0.5, 1, and 2 kHz, respectively. Eleven cases had profound hearing loss in the affected ears, with average hearing levels of >90 dB. The average hearing levels

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Table I. Audiologic and radiologic findings in children with unilateral cochlear nerve (CN) hypoplasia.

			Audiologic findings in affected ear				CNC diameter (mm)	
Case no.	Age (years)/sex	Side	Pure-tone hearing level (dB)	ABR thresholds (dB nHL)	Maximum speech discrimination (%)	DPOAE	Affected ear	Healthy ear
1	6/F	R	41	80	25	Absent	1.2	1.7
2	5/F	L	35	90	60	Absent	0.7	1.9
3	8/F	R	59	–	70	Absent	0.5	1.7
4	8/M	R	38	–	90	Normal	1.3	2.0
5	13/F	L	59	–	50	Absent	1.0	2.1
6	4/F	R	40	60	–	Absent	0.3	1.8

–, not evaluated; ABR, auditory brainstem response; CNC, cochlear nerve canal; DPOAE, distortion product otoacoustic emission; F, female; M, male.

of the affected ears in the remaining six (24%) cases were <60 dB (Table I). These six patients were therefore included in this study for further analysis.

The six pediatric cases consisted of one boy and five girls with a mean age of 7.3 years (range, 4–13 years). None of the children had known syndromes that could cause hearing loss or risk factors of hearing loss such as a history of prematurity, hypoxia, and hyperbilirubinemia.

Imaging

High-resolution computed tomography (HRCT) of the temporal bone in all patients was performed with

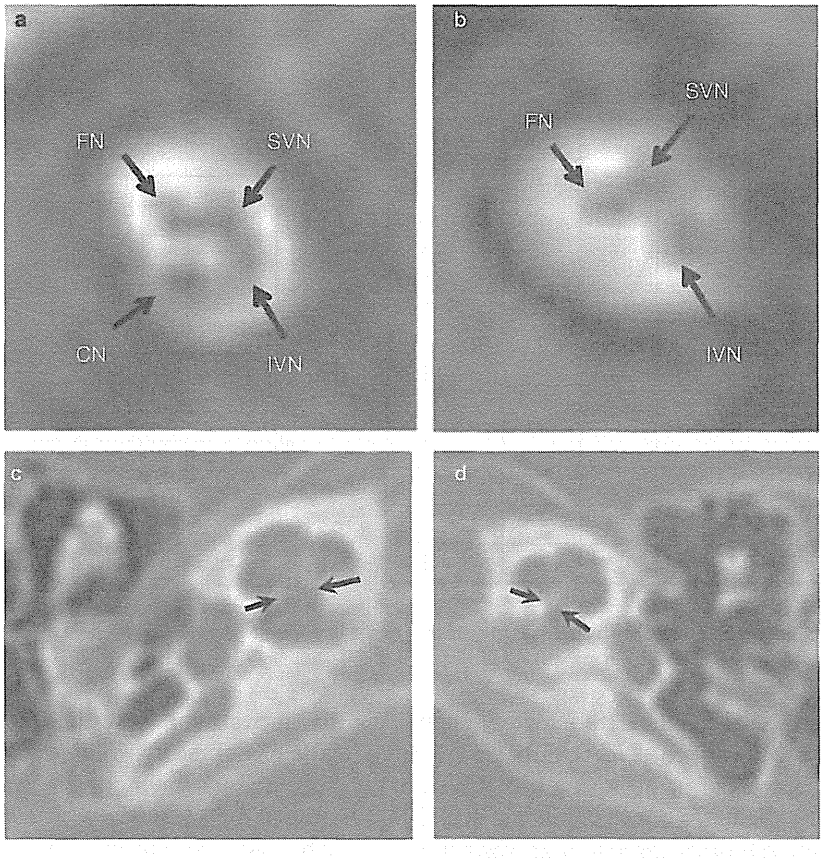

Figure 1. Cochlear nerve hypoplasia in a 5-year-old girl with left sensorineural hearing loss (SNHL) (case 2). (a, b) Oblique sagittal magnetic resonance (MR) images of the inner auditory canal (IAC). Four nerves were detected in the right (a), while the left cochlear nerve is not visible (b). CN, cochlear nerve; FN, facial nerve; IVN, inferior vestibular nerve; SVN, superior vestibular nerve. (c, d) Axial images of temporal bone high-resolution computed tomography (HRCT) show narrowing of left cochlear nerve canal (CNC) (d, 0.7 mm) compared with the right CNC (c, 1.9 mm).

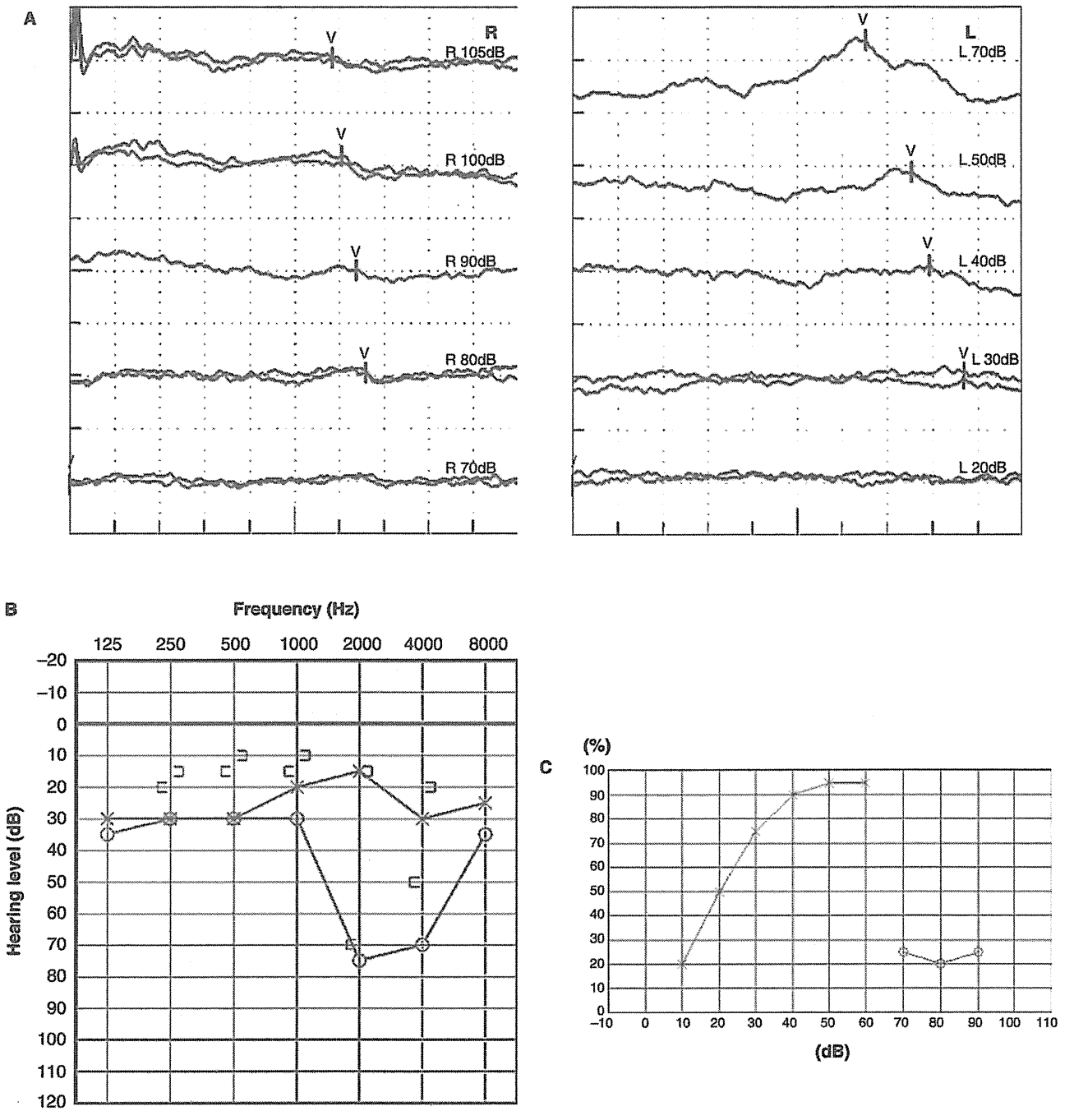

Figure 2. Audiologic findings in a child with right-sided cochlear nerve hypoplasia (case 1). (A) Auditory brainstem responses (ABRs), (B) pure-tone audiogram, (C) word recognition curve.

a multidetector-row CT scanner (8-detector, Lightspeed Ultra, GE, Milwaukee, USA). Images were acquired in the direct axial planes using a 0.652 mm slice thickness. The diameter of the CN canal (CNC) was measured along the inner margin of its bony walls at its middle portion on the axial image of the base of the modiolus.

MR images were obtained using a 1.5 Tesla system (Intera 1.5T; Philips, Belgium) according to a protocol described previously in detail [4]. The MRI scans included 3-D T2-weighted fast spin-echo sequences in axial and oblique sagittal images of the IAC with a 0.7 mm slice thickness. The 3-D constructive interference in steady-state (CISS) images was then reconstructed by traversing the IAC in a perpendicular orientation, producing images that visualized the four nerves (facial, superior vestibular, inferior vestibular, and cochlear). The

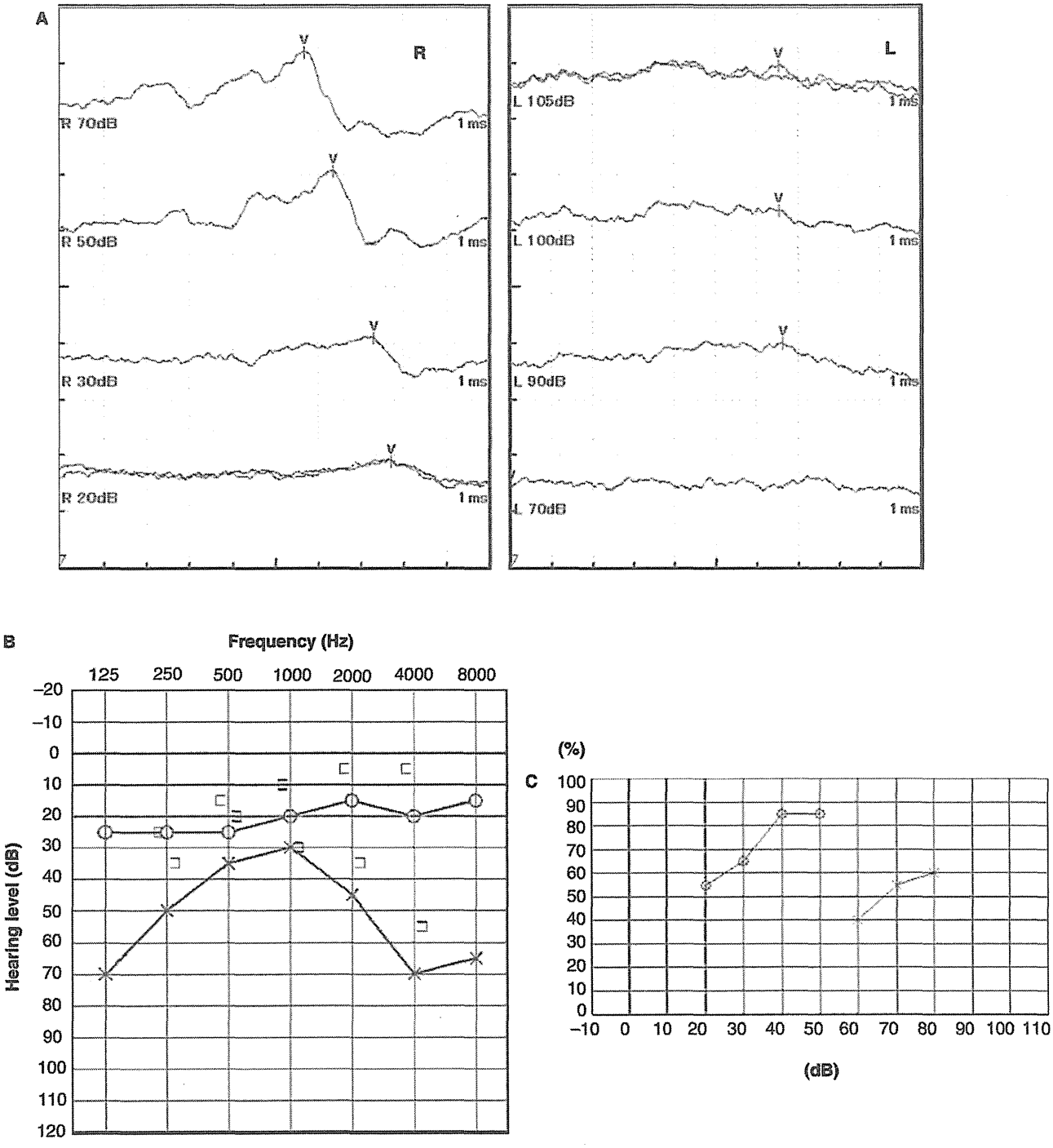

Figure 3. Audiologic findings in a child with left-sided cochlear nerve hypoplasia (case 2). (A) Auditory brainstem responses (ABRs), (B) pure-tone audiogram, (C) word recognition curve.

findings of a normal ear are shown in Figure 1a, b, (case 2, right ear).

Audiologic assessment

In addition to the pure-tone hearing test, distortion product otoacoustic emission (DPOAE) and auditory brainstem response (ABR) testing, as well as speech audiometry were performed. DPOAEs were measured in all subjects for pairs of primary tones (f1 and f2), with a fixed ratio of f2/f1 = 1.2, and fixed levels of 65 dB SPL (L1) and 55 dB SPL (L2) using the ILO292 system (Otodynamics, UK). The frequency of f2 was stepped through a range of 1–6 kHz to yield a nine-point DPGram.

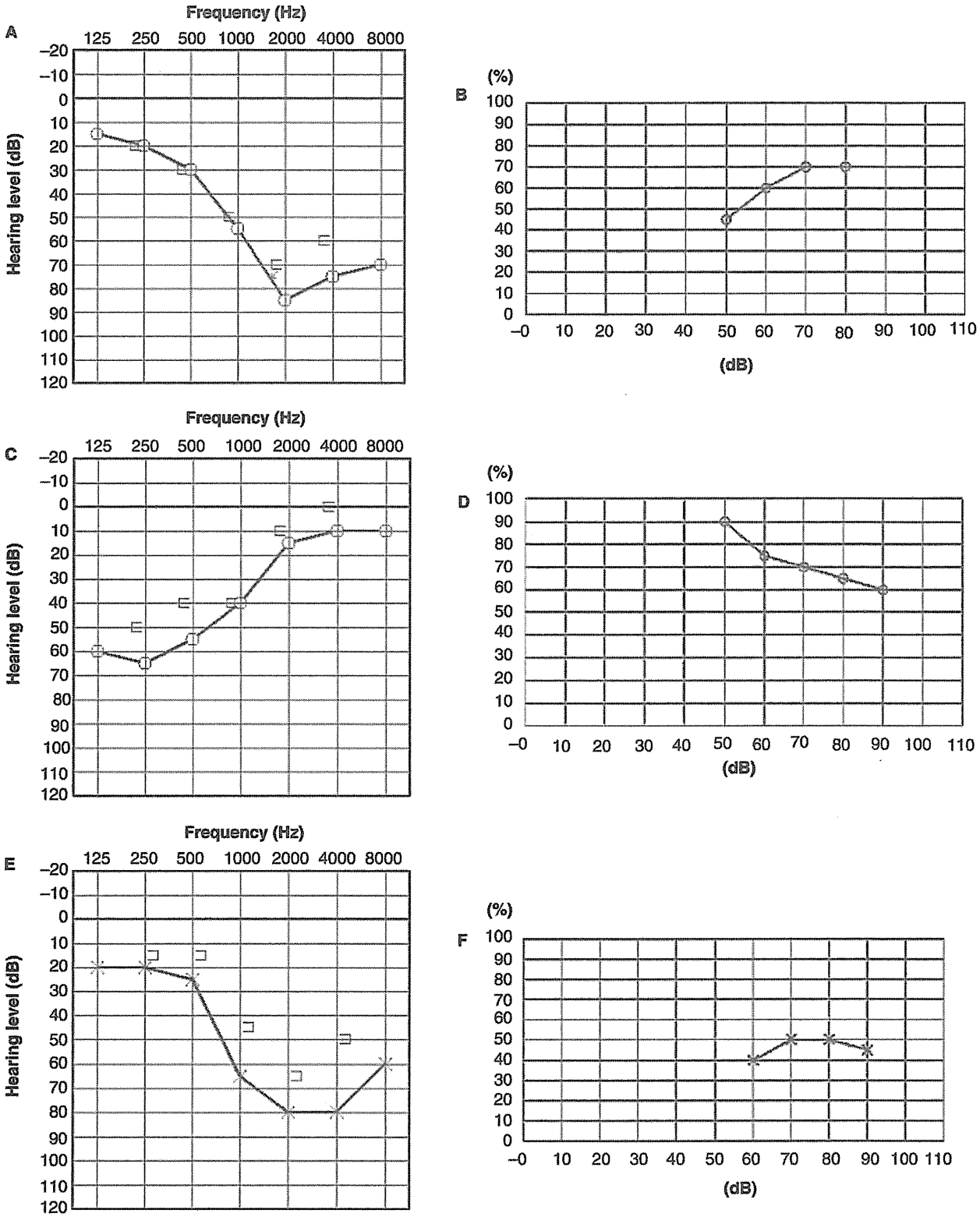

Figure 4. Audiologic findings in ears with cochlear nerve hypoplasia (cases 3–5). (a, c, e) Pure-tone audiograms: (a) case 3, (c) case 4, (e) case 5. (b, d, f) Word recognition curves: (b) case 3, (d) case 4, (f) case 5.

ABRs were recorded in three subjects using the MEB-2204 system (Nihon Koden, Japan). The 0.1 ms clicks with alternating polarity were presented monaurally at a repetition rate of 10 Hz and a maximum intensity of 105 dB nHL.

Speech audiometry was performed in five subjects, but not in case 6 where the test could not be done. The maximum word recognition scores were evaluated based on the percentage of correct answers out of 20 words, using Japanese word list 67-S. In normal-hearing subjects, the maximum word recognition scores using the word list are usually ≥90%.

Results

A summary of the findings for the six children with unilateral CN hypoplasia diagnosed by MRI is shown in Table I. On the oblique sagittal MRI image of case 2, the left CN is undetectable, while the right CN is normal (Figure 1a, b). The hearing levels of the affected ears in the six cases ranged from 35 to 59 dB (Table I). The audiogram shapes (Figures 2, 3, and 4) were high-frequency sloping (cases 3, 5, and 6), rising (case 4), 2–4 kHz notch (case 1), and inverted scoop shape (case 2). DPOAEs were present in only one (17%) of the six affected ears, and the shape of the hearing loss curve for that ear was rising.

In one of the three cases in which ABR testing was performed, the ABR threshold of the ear with CN hypoplasia was significantly elevated compared with that expected from 1000–4000 Hz pure-tone hearing levels (case 2). Speech discrimination tests were performed in five cases (Figures 2c, 3c, and 4b, d, f) but not in the 4-year-old patient (case 6). In two of these five cases (cases 1 and 2), the maximum word recognition scores of the ears with CN hypoplasia were poor compared with those of pure-tone hearing levels. In one case (case 4), the maximum word recognition score of the affected ear was 90%, but the results for 50 dB and 90 dB were 90% and 60%, respectively. Therefore, the word recognition curve in case 4 showed marked roll-over.

The click-evoked ABR tracing, pure-tone audiogram, and word recognition curves for case 1 are shown in Figure 2. The pure-tone audiogram of the right ear (affected side) showed a 2–4 kHz notch configuration. The wave V threshold of the right ear was elevated (80 dB nHL). The ABR threshold compared with the 1–4 kHz pure-tone hearing level of the right ear was slightly higher than expected. The maximum word recognition score of the right ear was 25%, which was lower than expected for pure-tone hearing.

The click-evoked ABR tracing, pure-tone audiogram, and word recognition curves for case 2 are shown in Figure 3. The pure-tone audiogram of the left ear (affected side) showed an inverted scoop shape. The ABR threshold of the left ear was 90 dB nHL and the maximum word recognition score of the left ear was 60%. These values were smaller than expected for pure-tone hearing.

All six ears with CN hypoplasia were associated with CNC stenosis (CNC diameter, <1.5 mm; mean, 0.83 mm). In contrast, the unaffected ears of the six children had CNC diameters of >1.5 mm (mean, 1.9 mm). Axial HRCT images of a representative case (case 2) of severe CNC stenosis are shown in Figure 1c and d. No cochlear malformations were seen in the six children.

Discussion

We defined CN hypoplasia as an undetectable CN on axial, coronal, or reconstructed oblique sagittal MR images. An extremely small nerve, below the limits of resolution of MRI, could appear absent and therefore should not be disregarded. Therefore, we avoid the terms deficiency, aplasia, and agenesis.

The mechanism of CN hypoplasia in children remains speculative. Both congenital deficiency and acquired degeneration of the CN might be seen in children with SNHL [5]. In pediatric cases, it is possible that a vascular insult during critical periods in development may result in isolated CN agenesis or degeneration [1]. CN hypoplasia is often associated with cochlear anomalies [4] or various coexisting syndromes such as CHARGE association [1]. Neither cochlear malformations nor known syndromes were recognized in the patients presented in this report.

CN hypoplasia is not as uncommon as previously thought [1]. Recent studies suggest that CN dysfunction accounts for up to 10% of diagnosed cases of pediatric SNHL [2]. Miyasaka et al. [4] reported CN hypoplasia in 8 of 42 (19%) ears on MRI. Of these, four ears had inner ear malformations. In the present study, no cochlear malformations were recognized in the six ears with CN hypoplasia. In addition, a relationship between CNC stenosis and CN hypoplasia was previously reported [2,6,7]. CNC diameter measurements of <1.8 mm were considered moderate stenosis, while measurements of <1.0 mm were defined as critical stenosis [8]. A CNC diameter of <1.5 mm on CT suggested CN hypoplasia [4,6,7]. All six ears with CN hypoplasia in this study were associated with CNC stenosis (CNC diameter, <1.5 mm). And also 11 ears with CN hypoplasia with profound hearing loss were associated with CNC stenosis. The mean (SD) CNC diameter was 0.83 (0.40) mm in the affected side of 6 cases with mild to moderate hearing loss, and 0.72 (0.32) mm in the affected side of 11 cases with profound hearing

loss. The CNC diameters in the group with profound hearing loss were slightly narrower than in the group with mild to moderate hearing loss, but there was no statistically significant difference ($p = 0.28$, t test). CNC may require stimulation by its contents for normal development, meaning that CNC stenosis may occur secondary to CN hypoplasia [6]. However, it was previously reported that CNC stenosis can occur without CN hypoplasia [4]. CNC stenosis on CT may therefore be indicative of the diagnosis of CN hypoplasia, but MRI should be performed to confirm the diagnosis.

In the past, it was thought that CN hypoplasia was always associated with profound SNHL [1], but a case of CN hypoplasia without profound hearing loss was reported recently [3]. In that case, an extremely small, preserved, and partially functional CN was believed to be present in the affected ear [3]. A minimal number of residual CN fibers, which were too small to be detected by MRI, may be enough to deliver sound information without threshold elevation [8].

CN hypoplasia may present as auditory neuropathy spectrum disorder (ANSD) [1,9]. In this study, DPOAEs were detected in one of six cases, which indicated normal outer hair cell function. In the 11 cases of CN hypoplasia with profound hearing loss, DPOAEs at the affected ears were detected in four cases (36%). The presence rates of DPOAEs were supposed not to relate to hearing levels. The reason for absent DPOAEs in cases of mild hearing loss is unclear, but malformations of inner ear microstructures associated with congenital CN hypoplasia are considered to be the cause of absent DPOAEs. The shape of the hearing loss curve in the case with normal DPOAE was rising (Figure 4c), and the pure-tone thresholds at 2–8 kHz were ≤15 dB. The normal DPOAE response in this case is assumed to indicate the preservation of inner ear function at high frequencies.

In case 2, who was one of three cases in which ABR testing was performed, the ABR threshold of the affected ear was significantly elevated compared with that expected from 1–4 kHz pure-tone hearing levels. The elevated ABR threshold in the case suggests disorders of CN synchrony at high frequencies.

Speech discrimination assessments showed poor maximum word recognition scores compared with that expected from pure-tone hearing levels in two affected ears. In addition, the word recognition curve of an affected ear had marked roll-over. The results of the speech discrimination tests suggested retrocochlear disorders in the affected ears. Some of the findings of the ABR and the word recognition tests in CN hypoplasia are consistent with audiologic characteristics of ANSD, which has been reported as retrocochlear hearing loss in CN hypoplasia [1]. It is believed that 6–28% of ANSD cases are due to CN hypoplasia [1,10,11]. CT is recommended for the initial screening of children with SNHL [4]. For children with ANSD, high-resolution MRI of the CN should be performed as the initial imaging study [12]. The results of the present study suggest that the imaging study for the screening of CN hypoplasia is desirable for even mild to moderate hearing loss.

Conclusion

Here, we presented six pediatric cases of CN hypoplasia with mild to moderate hearing loss. Audiologic characteristics of some ears with CN hypoplasia in this study suggested retrocochlear disorders. Even if hearing loss is mild to moderate, the presence of CN hypoplasia associated with retrocochlear disorders should be considered.

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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◆特集・補聴器に関するQ&A—診療所における対応—

2．初めて補聴器を患者さんに適合する場合について

Q3　3歳時健診にて発見された場合は？

守本倫子*

Key words：軽度難聴・中等度難聴(mild hearing loss, moderate hearing loss)，高音急墜型難聴(high frequency hearing loss)，語音明瞭度(speech perception)

Point

①3歳児健診では伝音難聴や中等度難聴が発見されることが多い．
②ある程度言葉がでていることが多いため，親が補聴器装用に同意しないことも多い．
③補聴器装用のコツは，家族が日常生活における難聴児の直面する問題点を理解すること．

はじめに

新生児スクリーニングが施行されるようになったものの，いまだに2歳半～3歳過ぎて初めて難聴がみつかる場合も少なくない．本稿では，そういった患者さんへの対応について解説する．

3歳時健診で発見される難聴とは？

新生児聴覚スクリーニングも行われているため，高度難聴は3歳前までに発見されていることが多い．しかし，軽度・中等度難聴は3歳児健診で発見されることが少なくない．

日本耳鼻咽喉科学会での報告によると3歳児健診の受診率は80%以上と高く，そのうち精密検査にて両側難聴が発見されるのは0.07%とされている．これは，新生児聴覚スクリーニングが開始される前の1997年からほとんど発見される頻度は変わっていないとされている[1]．この理由として，3歳児健診で発見される難聴児は，滲出性中耳炎による伝音難聴であることが多いこと，また感音難聴では，おそらく進行性・遅発性難聴のために新生児期には聴力が正常であった例なども含まれていることが推測される．いずれにしても，出生直後からしばらくの間は聴力が正常であった時期があるため発声がみられたりするため見落とされやすい．

森田らは，3歳以降に発見された難聴児に対してアンケート調査を行ったところ，大半は2歳過ぎには母親が聞こえが悪いことに気が付いていたものの，健診時に経過観察を指示されており，発見が遅れていると報告している[2]．診断する側も慎重に判断する必要がある．

3歳児に対する難聴の診断方法

1．難聴の診断

多くは伝音難聴であるため，鼓膜所見，ティンパノメトリーで滲出性中耳炎や慢性中耳炎を除外する．言葉を覚える大事な時期であるので，鼓膜チューブ留置術など積極的に聴力改善に努める．中耳所見に異常が認められなかった場合，遊戯聴力検査や幼児聴力検査(COR，ピープショウテスト)と耳音響放射検査(DPOAE，TEOAE)，ABR，

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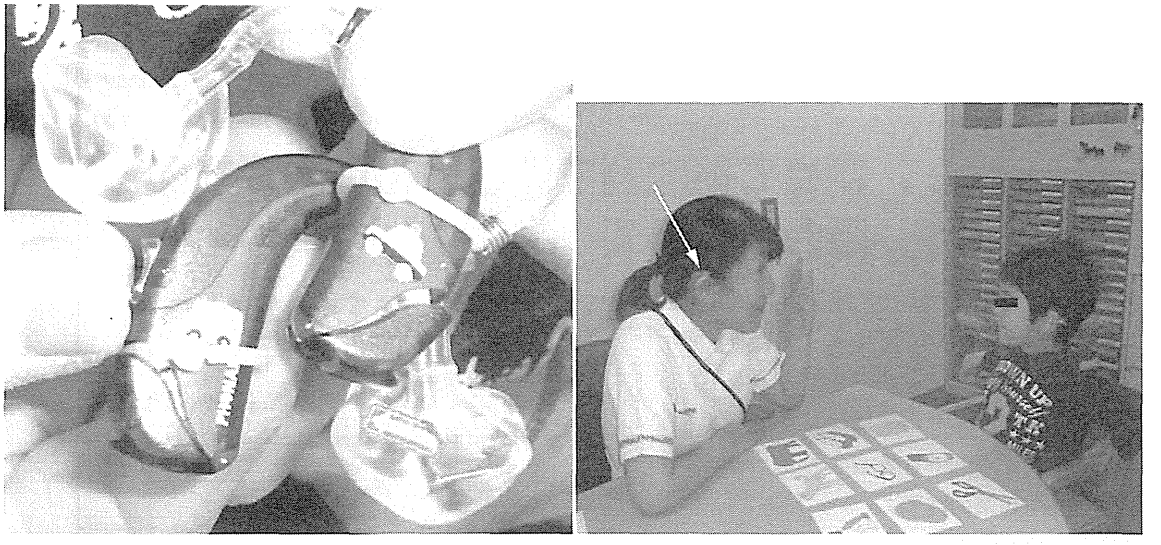

a|b

図 1.
a：補聴器に親しみを感じさせる工夫．補聴器にはそれぞれパトカーと救急車のシールが貼られているため左右が覚えやすい．イヤモールドにもシールを埋め込んで作成してある
b：3歳児の言語訓練．補聴器をつけたがらないため，言語聴覚士も同じ補聴器を耳にかけて(⇑)，仲間意識を持たせながら訓練を行っている

ASSR を組み合わせて検査する．

2．難聴の程度の評価

純音聴力検査などは3歳4〜6ヶ月以降でないとできないことが多い．しかし，高音急墜型難聴などが疑われる場合など，補聴器の調整のためにも正確な聴力検査結果が必要である．乳幼児の聴力検査では，辛抱強く何度か遊戯聴力検査を繰り返し，閾値を確定することが大切である．また，インサートイヤホンを用いての遊戯聴力検査や条件詮索反応聴力検査(visual reinforcement audiometry；VRA)は有効である．

補聴器の選択機種

1．機　種

アナログ型補聴器によるリニア増幅では，十分な利得を確保させると，最大出力が不快閾値レベルを超えてしまう．中等度難聴児では，やや高い聴覚閾値と正常やや低い不快閾値を持っているため，ボリューム調節のできない幼児では，装用に伴う不快感のみが強調されてしまう[3]．このため，ノンリニア増幅のデジタル補聴器で，COR などの聴力閾値を参考にパソコン上で出力音圧を調節することができる．また，ハウリング抑制機能を持つため，特に高音急墜型難聴に対しても利得を上げることができて有用である．小児の場合は，両耳装用が必要である．

2．見た目

子どもが装用したくなるように，カラフルなものや，補聴器にお気に入りのシールを貼るなどの工夫をする．左右を間違えないように，イヤモールドを異なる色で作成したり，シールで目印をつけるのもよい(図 1-a)．また，母も補聴器をつける真似をしたり，言語聴覚士もダミーの補聴器を装用するなどで補聴器装用に対する抵抗を感じさせなくすることも有効である(図 1-b)．

補聴器適合の際に参考とする検査

1．補聴器の必要性

軽度難聴では，親に補聴器をつけさせる必要性を理解させることから始まる．受け答えができることから，他の健聴児と比べて言葉がやや遅れている程度にしか思っていないことも少なくない．

1）発達検査

3〜4歳児では，新版 K 式言語発達検査やウェクスラー式(WPPSI)などでの言語発達の遅れの有無を評価する．明らかに言語面のみでの遅れが

ある場合は，難聴が影響している可能性から補聴器装用を検討する．

2）語彙能力検査

絵画語彙検査(PVT-R)では複数の絵を見せて説明にあった絵を選択させる．語彙数を評価することができる．日常何気なく耳に入ってくる言葉をどのくらい身に着けることができているのか，をみることで，難聴により自然には語彙数が増えていないことなどが評価できる．

2．補聴器装用効果の評価

1）音場での装用閾値検査

補聴器を装用して，乳幼児聴力検査(CORまたはピープショウテスト)を行い，補聴器により十分に聴力閾値が上昇しているかをみる．幼児の場合，言語の獲得のためには成人よりも低い閾値が求められる．

2）ことばの聞き取り検査

特に感音難聴では，「サ行」が「ハ行」に，「タ，ハ行」が「カ行」に聞き誤ることが多い．こうした音の含まれた絵カードを並べ，口元を隠して聞き取れるか検査する．

3．両親からみた満足度

補聴器を装用させた時の声の出方，語彙の増え方，音への反応の良さ，などを保護者や養育に関わる者に観察してもらう．発声や声の質などは，子ども自身が自分の声をフィードバックできているのかという指標になる．また，様々な場面(遠くから声をかけたとき，幼稚園での反応など)での違いを確認し，装用前と変化がないようであれば，調整が必要である．

親への説明(診断時，初めて装用時)

1．診断時

1）高度難聴

進行性難聴や人の表情を読み取るのがうまい子などが，この時期まで高度難聴を見過ごされてしまうことが少なくない．出生後しばらくの間は音が聞こえていた可能性や，表情のみで反応ができるようなコミュニケーション能力に長けている子

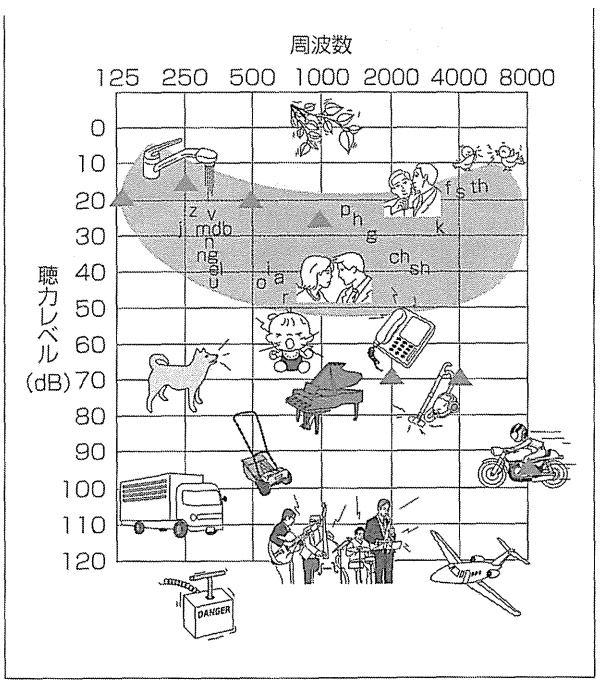

図 2. 聞こえる音と日常会話音声範囲(スピーチバナナ)の関係

などは，補聴器をつけてあげることでもっとその能力が伸びてくる可能性などを説明する．また，近い将来人工内耳留置手術を行う可能性についても言及する．

2）軽度～中等度難聴

ある程度の音が聞こえているために親も難聴であることを受容できなかったり[5]，補聴器の必要性が理解できないこともある．図2のように聴力をプロットし，いったいどのような語音が聞き取れていないのかを示す．また，構音検査などで，どのような音が正確に構音できないかを把握してもらい，単に年齢が幼少であるから「赤ちゃん言葉」で話すのではなく，音がそのようにしか聞こえていないことや，自分の構音が自分にフィードバックできていないために正確な構音に直せないことなどを認識してもらうことが大切である．

2．初めて装用時

1）装用時間

最初はうるさがったり，耳に何かをつけられるのを嫌がったりすることがある．最初は2～3時間装用から始め，徐々に装用時間を長くする．また，最初は静かなところで装用させ，装用してい

る間は親が一緒に遊ぶ，本を読むなどで補聴器が気にならないようにさせる．

2）フィッティング

最大出力音圧は成人よりやや-10 dB 程度に調整し，装用を開始する．装用時の反応を COR で確認する．この段階で調整したとしても，1回でよく聞こえるようになるわけではない．むしろ，細かいスパンで聴覚評価と再調整を繰り返し行い，徐々に適切なフィッティングに近づけていくということを理解してもらう．

限　界

静かなところではよく聞こえるようになったとしても，騒音下での聞き取りは困難であることが多い．幼小児に長時間補聴器をつけさせるコツは，家族が日常生活での難聴児の直面する問題点について理解することである[5]．

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特集 幼少児の難聴の診断・治療と聴こえと言葉のリハビリテーション

② 中耳・内耳・中枢聴覚伝導路の発達

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はじめに

「個体発生は系統発生をくり返す」ことが知られていますが，目と耳では異なった特徴を示します。脊椎動物の視器は高度に分化しています。聴器は，魚類，両生類，爬虫類は分化が乏しいですが，哺乳類では高度に分化しています。これに比べ視器の分化が進んでいる動物でも，その中枢神経系は聴覚系よりも複雑ではありません。感覚細胞は視器の網膜に約1億あり，聴器のコルチ器に約1万あります。この違いが脳の仕組みが異なっていることにつながっていると思われます。聴覚は，哺乳類ではそれぞれ脳幹レベルの中枢聴覚伝導路構造は類似していても大脳レベルでは異なり，とくに聴覚理解のための言語中枢はヒトだけにあり，右利きでは左半球に局在しているのが最たるものです。

高度に分化した感覚器の進化

Charles Darwin の『種の起源』では，感覚器の進化について次のように述べています[1]。「いくつもの事実を踏まえると，感受性の高い神経が光も感じるようになって目になり，音の空気の振動を感じるようになって耳になっても不思議はないと，私はあえていいたい」という洞察です。

Darwin は自然淘汰説に基づく『種の起源』を著しましたが，感覚器はあまりにも高度に分化しているので，うまく説明できず困ったようです。組織学的には，角膜も鼓膜も共通して上皮でできているのは，Darwin の説明の証拠かもしれません。

Darwin は多くの少年と同様に，小さい頃は昆虫の好きな少年でした。

目の進化については，『目で見る進化』という本に「光が来ると，光を感じる。神経も対応する。だんだんガラス体やレンズ，角膜ができたりする」とし，すべてが海にいる生物の中に，目の進化の初めのものからヒトの目より高度に進化したものまで存在するという解説です[2]。目は，水の惑星である地球の海の中で進化がほぼ完成したように推測されます。

聴覚は目とは異なり地球が空気の惑星になって初めて耳ができます。発声によって空気が振動し音となり，それを聴く，すなわち音声コミュニケーションのためです。耳の進化については，魚類には三半規管はありますが，音を感じるのは体表に存在する側線器という部分で，水の流れとともに音も感じると考えられ，可聴周波数は100～1,000 Hz です（図1）[3]。次の両生類では，カエルの場合は鼓膜や耳小骨，三半規管

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