MATERIALS AND METHODS

From 2010 to 2011, we prepared 3D images for 6 separate CI cases prior to surgery. These cases consisted of 5 patients (3 boys, 2 girls, ages 1-7 years), including one bilateral case. All of the patients had congenital malformations in the temporal bone structures, such as in the FN, cochlea, semicircular canal (SC), or stapes. We created 3D images using a method we developed that utilized a public personal computer.

First, we manually colored preoperative CT images, which were axial sections of the temporal bone scanned by a normal CT scanner (SOMATOM Definition; Siemens Medical, München, Germany) at a slice thickness of 0.5 mm using Photoshop CS extended. The extended version can import and edit DICOM files directly. The inner ear labyrinth, auditory ossicles, and FN were shaded in blue, red, and yellow, respectively, maintaining the shape of these structures. We had to paint every axial section of temporal bone CT images; there were about 30-40 slices. It took about two hours to accomplish the coloring process.

We then converted the colored 2D-CT images to 3D images using Delta Viewer (DV), a freeware for Macintosh available on the Internet (http://delta.math.sci.osaka-u.ac.jp/DeltaViewer/index.html). This 3D reconstruction can be done from CT images of any condition, such as the thickness of the slice of images, but the thinner the CT slices are, the more detailed and smoother the 3D images will be. This DV-3D rendering process was completed automatically within a few minutes. In this paper, we refer to the 3D images created using DV as DV-3D images.

Fig. 1 is a DV-3D image of the normal temporal bone structures. We can rotate DV-3D images freely using the DV application.

Before each CI procedure, we discussed any problems anticipated based on the DV-3D images and planned the surgery with those who would be performing the procedure. We also brought either the printed images or the notebook PC to the operating room and compared the images with the surgical findings during the CI procedure (Fig. 2).

RESULTS

Case 1. Left ear of a three-year-old boy

The patient presented with bilaterally malformed ossicles, SC hypoplasia, internal auditory canal stenosis, and an abnormal course of the FN. We had already performed CI surgery on the

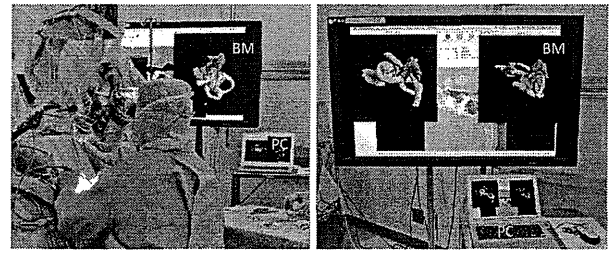

Fig. 2. The pictures of an operation room during a cochlear implantation surgery. The surgeon sees Delta Viewer 3 dimensional images displayed on the bedside monitor (BM), which is controlled by the notebook PC (Macintosh), and compares the images with surgical findings.

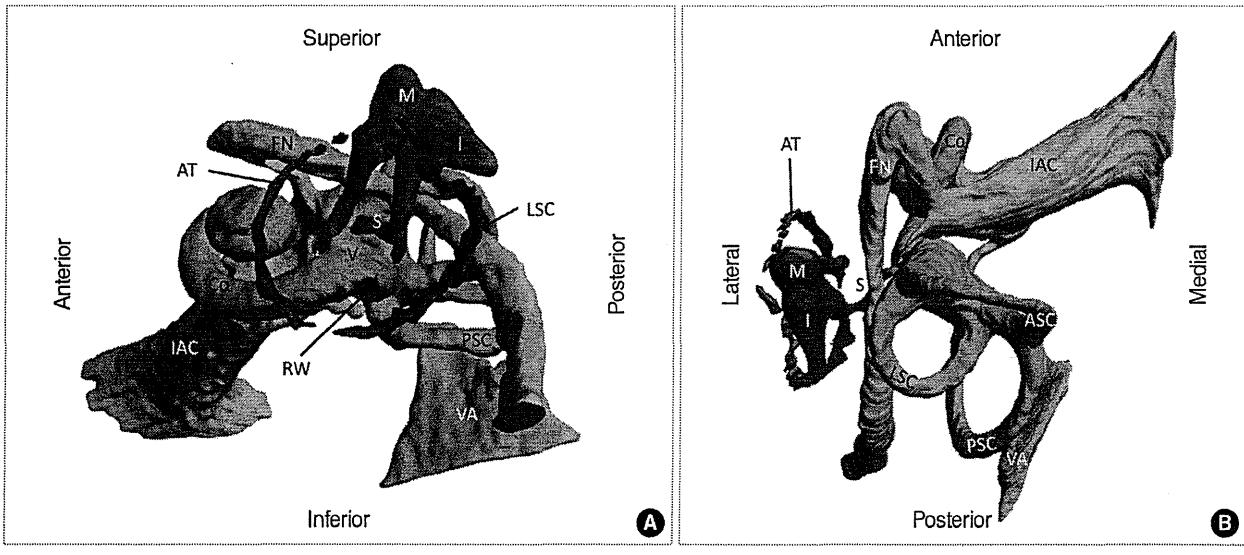

Fig. 1. Example of Delta Viewer 3 dimensional image of normal temporal bone structures of left ear based on computed tomography image. Image (A) is antero-lateral inferior view, and image (B) is superior view. The bony labyrinth was shaded in blue and includes the cochlea (Co), vestibule (V), anterior semicircular canal (ASC), lateral semicircular canal (LSC), posterior semicircular canal (PSC), and round window (RW). The internal auditory canal (IAC) and facial nerve (FN) are shaded in yellow. The ossicles are shaded in red and include the malleus (M), incus (I), and stapes (S). The annulus tympanicus (AT) and vestibular aqueduct (VA) are shaded in green and purple, respectively.

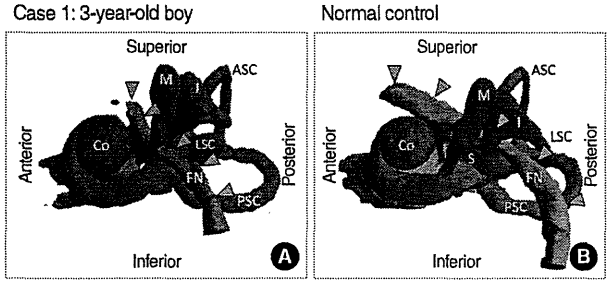

Fig. 3. (A) Lateral view of the temporal bone structures of case 1. The lateral semicircular canal (LSC) is hypoplastic, and the crus of the incus (I) and stapes (S) are absent. The labyrinthine segment of the facial nerve (FN) and the geniculate ganglion are posteriorly displaced, and the tympanic and mastoid segments of the FN are antero-inferiorly displaced, running more vertically than normal control. The cochlea (Co), malleus (M), anterior semicircular canal (ASC), and posterior semicircular canal (PSC) are intact. (B) Lateral view of normal control. The green arrowhead shows the course of the FNs.

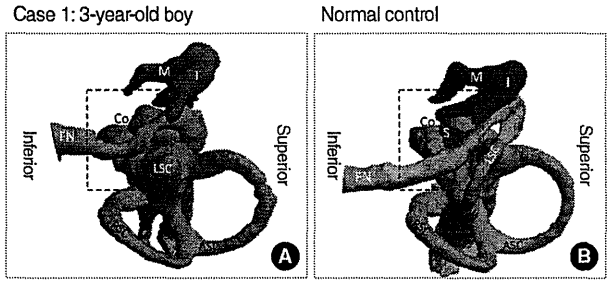

Fig. 4. (A) The Delta Viewer 3 dimensional (DV-3D) image of case 1 in the same position during cochlear implantation surgery. (B) The DV-3D image of normal control in the same position as (A). Rectangle of dashed line shows the area of Fig. 5. Co, cochlea; ASC, anterior semicircular canal; PSC, posterior semicircular canal; LSC, lateral semicircular canal; FN, facial nerve; M, malleus; I,incus; S, stapes.

patient's right ear one year before. The outcome of his speech was not as good as that of a patient with a non-malformed ear. We then planned a second CI procedure on the patient's left ear. We expected that it was going to be difficult to perform cochlear fenestration because of the facial nerve abnormality, so we prepared DV-3D images of this case before the CI surgery.

The DV-3D images (Fig. 3) indicate that the cochlear turn is intact, the lateral SC is hypoplastic, the long crus of the incus and stapes is absent, the labyrinthine segment of the FN is more posteriorly placed, and the tympanic and mastoid segments are antero-inferiorly displaced and running more vertically than normal. Fig. 4 shows the structures pictured in the same position as we found them during surgery. These images show that the stapes are absent and that the FN runs antero-inferiorly onto the oval window.

Fig. 5 shows the preoperative DV-3D image and the actual picture of the surgical findings. The space for cochlear fenestration was very narrow; however, we were able to insert an electrode based on the preoperative DV-3D image without any com-

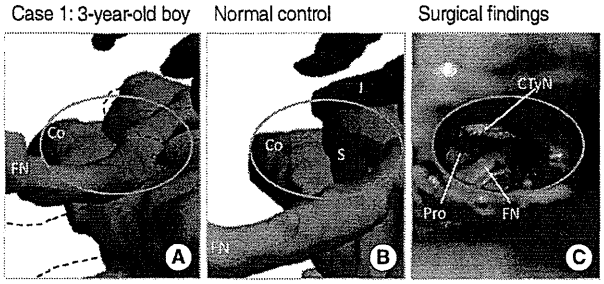

Fig. 5. The images of the surgical field of posterior tympanotomy as shown by the orange oval. (A) Case 1: close-up image of the rectangle of Fig. 4A. The stapes (S) and crus of the incus (I) are absent, and the facial nerve (FN) is antero-inferiorly displaced. Dashed line shows the position of the ossicles and facial nerve (FN) of normal control. The visible area of the basal turn of cochlea (Co) is smaller than normal. (B) Normal control: close-up image of the rectangle of Fig.4B. (C) The picture of the surgical findings of case 1. The visible area of the promontory (Pro) for cochleostomy is narrow and surrounded by the chorda tympani nerve (CTyN) and facial nerve (FN) displaced anteriorly.

Fig. 6. The X-ray of inserted electrode of case 1.

plications, such as FN palsy or stimulation. Fig. 6 shows an X-ray of the electrode: MED-EL, standard.

Case 2: Left ear of a seven-year-old boy

The patient presented with bilateral cochleo-vestibular malformations and abnormalities of the stapes and course of the FN. We had already performed CI surgery on the right ear one year before. We planned a second CI procedure on this patient's left ear for the same reason we performed surgery on the first patient.

The preoperative DV-3D images (Figs. 7, 8) indicate that the shape of the cochlea and the SC are hypoplastic, the stapes seem to be fused to the FN, and the course of the FN is abnormal, as in the previous case, with bifurcation. In addition, the round window niche is closed, so there is no landmark for cochlear fenestration. We were able to successfully perform the cochlear fenestration right next to the FN, which was detected using a FN monitor and referring to a DV-3D image of the nerve course.

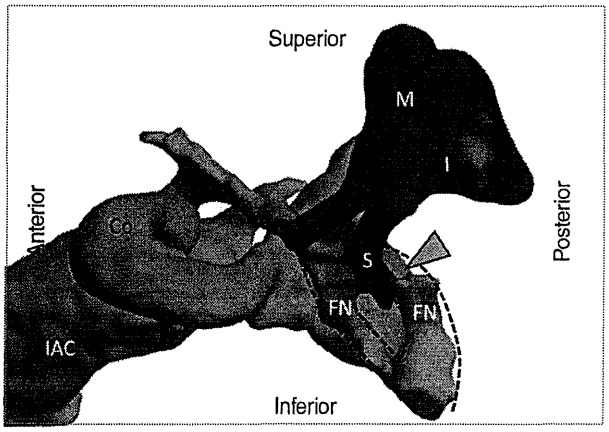

Fig. 7. Antero-lateral view of the temporal bone structures of case 2. The shape of the cochlea (Co) and semicircular canals are hypoplastic. The labyrinthine segment of the facial nerve (FN) and geniculate ganglion are posteriorly displaced, the tympanic and mastoid segments of the FN are antero-inferiorly displaced with bifurcation, and the stapes is fused to the facial nerve (arrow head). IAC, internal auditory canal; M,malleus; I, incus; S, stapes.

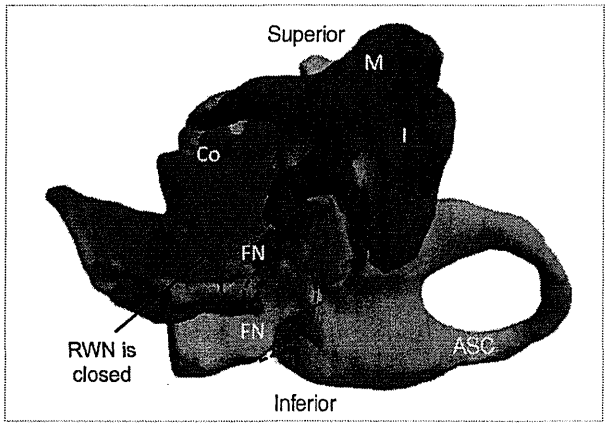

Fig. 8. The Delta Viewer 3 dimensional image of case 2 in the same position during cochlear implantation surgery. The surface of the tympanic cavity is displayed in green. The round window niche (RWN) is closed, so there was no landmark for cochlear fenestration. Co, cochlea; FN, facial nerve; ASC, anterior semicircular canal; M, malleus; I, incus.

Fig 9 shows an X-ray of the electrode: Cochlear, standard.

Table 1 consists of a list of the DV-3D images we prepared for 6 separate CI cases that involved ear malformation; in particular, an abnormal course of the FN. In all of the cases, we were able to successfully insert electrodes by referring to DV-3D images with no technical problems. Table 1 suggests that the anomaly of the stapes indicates an abnormal course of the FN.

DISCUSSION

We prepared 3D images of patients' ear malformations prior to performing CI procedures on these patients. These 3D images contributed to the successful insertion of electrodes during surgery on these patients, each of whom had an abnormal course of the FN. There is no previous report on the clinical use of 3D imaging for preoperative planning of CI surgery.

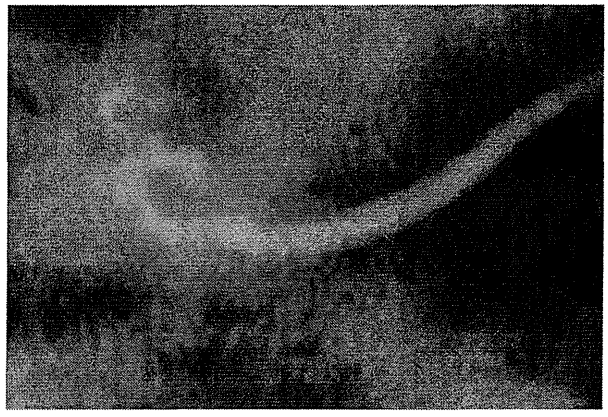

Fig. 9. The X-ray of inserted electrode of case 2.

We have been able to produce preoperative 3D images using a method we were able to develop at a low cost. Several software products are available that enable us to convert 2D medical images, such as those produced through CT or magnetic resonance imaging, to 3D. However, these imaging techniques are not suitable for creating 3D images of the temporal bone structures, because their resolution is so rough that they were developed generally for use on larger organs such as the lungs, large arteries, etc.

In very recent years, a high-performance multi-slice CT scanner and workstation have made it possible to generate detailed 3D images of the temporal bone structures for radiology diagnostics (1). Some previous articles have reported on the creation of 3D images based on a histological specimen for educational purposes (2-6).

A novelty of our method is that it enabled us to convert structures of different densities-bone structures, such as the ossicles, and soft tissue structures, such as the FN and cochlea-in the same manner on one 3D image. It is especially difficult to reconstruct a FN as a 3D image, because it is difficult to select the FN on a CT image. There is a previous report that investigated automatic FN selection through the use of special computer software; however, this method has not yet been put into use (7). There is room for further improvement in the accuracy of the DV-3D image; however, even now it is quite useful for planning a CI procedure and for avoiding surgical complications.

In this study, we suggest that an anomaly of the stapes indicates an abnormal course of the FN. It is well-established that an abnormal stapes has been associated with an anomalous course of the tympanic and mastoid segments of the facial nerve, because these are derivatives of the second branchial arch (8-10). As the labyrinthine segment of the FN is not derived from the

Table 1. Summary of cases and Delta Viewer (DV) 3D image findings

Case	Age (year) (sex)	Day of surgery (side)	DV-3D image findings					CI
			Cochlea	Semicircular canal (SC)	Stapes	Facial nerve		
						Labyrinthine segment	Tympanic and mastoid segment	
1	3 (M)	Feb 2010 Right	Intact	Hypoplasia of LSC	Absent	Posteriorly displaced	Antero-inferiorly displaced	2nd MED-EL standard
2	7 (M)	Jul 2010 Left	Hypoplasia	Aplasia of LSC and PSC	Abnormal: fusion to FN	Posteriorly displaced	Antero-inferiorly displaced with bifurcation	2nd cochlear straight
3	5 (F)	Mar 2010 Right	Intact	Aplasia of all SCs	Absent	Posteriorly displaced	Antero-inferiorly displaced	1st MED-EL standard
4	3 (M)	Feb 2010 Right	Hypoplasia	Almost intact	Absent	Posteriorly displaced	Antero-inferiorly displaced	1st cochlear straight
5	6 (F), same patient as case 3	Feb 2011 Left	Intact	Aplasia of all SCs	Absent	Posteriorly displaced	Antero-inferiorly displaced	2nd MED-EL medium
6	1 (F)	Nov 2011 Left	Almost aplasia	Almost intact	Intact	Intact	Intact	1st MED-EL medium

CI, cochlear implantation; LSC, lateral SC; PSC, posterior SC; FN, facial nerve.

second branchial arch but from the otic cupsule, the cause of the anomaly of this segment may not be discussed on the same basis as that of the other segments related to the second branchial arch.

CONFLICT OF INTEREST

No potential conflict of interest relevant to this article was reported.

REFERENCES

1. Fujii N, Inui Y, Katada K. Temporal bone anatomy: correlation of multiplanar reconstruction sections and three-dimensional computed tomography images. Jpn J Radiol. 2010 Nov;28(9):637-48.
2. Sorensen MS, Dobrzeniecki AB, Larsen P, Frisch T, Sporring J, Darvann TA. The visible ear: a digital image library of the temporal bone. ORL J Otorhinolaryngol Relat Spec. 2002 Nov-Dec;64(6):378-81.
3. Wang H, Merchant SN, Sorensen MS. A downloadable three-dimensional virtual model of the visible ear. ORL J Otorhinolaryngol Relat Spec. 2007;69(2):63-7.
4. Sorensen MS, Mosegaard J, Trier P. The visible ear simulator: a public PC application for GPU-accelerated haptic 3D simulation of ear surgery based on the visible ear data. Otol Neurotol. 2009 Jun;30(4):484-7.
5. Venail F, Deveze A, Lallemant B, Guevara N, Mondain M. Enhancement of temporal bone anatomy learning with computer 3D rendered imaging software. Med Teach. 2010;32(7):e282-8.
6. Mukherjee P, Uzun-Coruhlu H, Curthoys IS, Jones AS, Bradshaw AP, Pohl DV. Three-dimensional analysis of the vestibular end organs in relation to the stapes footplate and piston placement. Otol Neurotol. 2011 Apr;32(3):367-72.
7. Noble JH, Dawant BM, Warren FM, Labadie RF. Automatic identification and 3D rendering of temporal bone anatomy. Otol Neurotol. 2009 Jun;30(4):436-42.
8. McClerkin WW, Schuknecht HF. Facial nerve and stapes anomaly: a case report. Laryngoscope. 1974 Jul;84(7):1202-9.
9. Takahashi H, Kawanishi M, Maetani T. Abnormal branching of facial nerve with ossicular anomalies: report of two cases. Am J Otol. 1998 Nov;19(6):850-3.
10. Romo LV, Curtin HD. Anomalous facial nerve canal with cochlear malformations. AJNR Am J Neuroradiol. 2001 May;22(5):838-44.

CEO Clinical and Experimental Otorhinolaryngology Vol. 5, Suppl 1: S59-S64, April 2012 http://dx.doi.org/10.3342/ceo.2012.5.S1.S59

Original Article

What Factors Are Associated with Good Performance in Children with Cochlear Implants? From the Outcome of Various Language Development Tests, Research on Sensory and Communicative Disorders Project in Japan: Nagasaki Experience

Yukihiko Kanda, MD[1,2] · Hidetaka Kumagami, MD[2] · Minoru Hara, MD[3] · Yuzuru Sainoo, MD[3] · Chisei Sato, MD[3]
Tomomi Yamamoto-Fukuda, MD[2] · Haruo Yoshida, MD[2] · Akiko Ito[1] · Chiharu Tanaka[1] · Kyoko Baba[1]
Ayaka Nakata[1] · Hideo Tanaka[1] · Kunihiro Fukushima, MD[4] · Norio Kasai, MD[4,5] · Haruo Takahashi, MD[2]

[1]Kanda ENT Clinic, Nagasaki Bell Hearing Center, Nagasaki; [2]Department of Otolaryngology Head and Neck Surgery, Nagasaki University Graduate School of Biomedical Sciences, Nagasaki; [3]Department of Otolaryngology Head and Neck Surgery, Nagasaki University Hospital, Nagasaki; [4]Department of Otolaryngology Head and Neck Surgery, Okayama University Postgraduate School of Medicine, Dentistry, and Pharmaceutical Science, Okayama; [5]The Association for Technical Aids, Tokyo, Japan

Objectives. We conducted multi-directional language development tests as a part of the Research on Sensory and Communicative Disorders (RSVD) in Japan. This report discusses findings as well as factors that led to better results in children with severe-profound hearing loss.

Methods. We evaluated multiple language development tests in 33 Japanese children with cochlear implants (32 patients) and hearing aid (1 patient), including 1) Test for question and answer interaction development, 2) Word fluency test, 3) Japanese version of the Peabody picture vocabulary test-revised, 4) The standardized comprehension test of abstract words, 5) The screening test of reading and writing for Japanese primary school children, 6) The syntactic processing test of aphasia, 7) Criterion-referenced testing (CRT) for Japanese language and mathematics, 8) Pervasive development disorders ASJ rating scales, and 9) Raven's colored progressive matrices. Furthermore, we investigated the factors believed to account for the better performances in these tests. The first group, group A, consisted of 14 children with higher scores in all tests than the national average for children with hearing difficulty. The second group, group B, included 19 children that scored below the national average in any of the tests.

Results. Overall, the results show that 76.2% of the scores obtained by the children in these tests exceeded the national average scores of children with hearing difficulty. The children who finished above average on all tests had undergone a longer period of regular habilitation in our rehabilitation center, had their implants earlier in life, were exposed to more auditory verbal/oral communication in their education at affiliated institutions, and were more likely to have been integrated in a regular kindergarten before moving on to elementary school.

Conclusion. In this study, we suggest that taking the above four factors into consideration will have an affect on the language development of children with severe-profound hearing loss.

Key Words. *Cochlear implant, Children, Research on sensory and communicative disorders, Language development, Japan*

• Received December 1, 2011
Revision January 20, 2012
Accepted February 10, 2012

• Corresponding author: **Yukihiko Kanda, MD**
Kanda ENT Clinic, Nagasaki Bell Hearing Center, Wakakusa 4-25, Nagasaki 852-8023, Japan
Tel: +81-95-841-7038, Fax: +81-95-841-7041
E-mail: n-bell@estate.ocn.ne.jp

INTRODUCTION

Cochlear implantation (CI) is a highly specialized medical procedure for severe-to-profound hearing loss in patients all over the world. Newborn hearing screening (NHS) makes early detection and thus early intervention possible. NHS has allowed us

to test 95% of newborns in Nagasaki over the last 4 years. With the rapid increase in use of pediatric CI, there is a need to develop more intensive, longitudinal, and standardized tests for auditory, speech, and communication skills and language development. There are very few packages that include multiple language development in the world.

As a part of the Research on Sensory and Communicative Disorders (RSCD) project in Japan, we examined various language development tests for children fitted with cochlear implants. This report discusses findings as well as factors that led to better results in children with severe-profound hearing loss.

METHODS AND RESULTS

Subjects

We examined 33 Japanese children (32 cochlear-implant patients and 1 hearing-aid patient) in our hearing center for the RSCD project. Children were selected according to the following criteria: 1) aged between 48 to 155 months and 2) congenital hearing impairment with a hearing level >70 dB (average over multiple frequency bands). Children unable to complete these tests because of further disabilities were not included. A consent form was provided in 2009. The age distribution was as follows: 4 years of age (4); 5 years (5); 6 years, i.e., 1st grade in primary school (4); 7-8 years, 2nd grade (2); 8-9 years, 3rd grade (4); 9-10 years, 4th grade (6); 10-11 years, 5th grade (4); and 11-12 years, 6th grade (4). Only one patient used hearing aids in both ears, and the remaining 32 children wore cochlear implants. Ten children had gone through the NHS process, while the other 23 had not. The age at fitting of hearing aids varied from 4 months to 5 years 4 months, and the age of cochlear implant surgery varied from 1 year 6 months to 6 years 3 months.

The tests were conducted between April 2009 and March 2010.

Methods 1

We asked the children to perform the following tests:

- Test for question and answer interaction development (TQAID): This test aims to evaluate interpersonal communication skills (IPCS) with 57 questions divided into 10 categories.
- Word fluency test (WFT): This test was conducted as a productive vocabulary task. Children were asked to generate as many words as possible from a given category in 60 seconds.
- Japanese version of the Peabody picture vocabulary test-revised (PVTR).
- The standardized comprehension test of abstract words (SCTAW): This test was conducted as comprehensive vocabulary tasks, and these consist of 32 or 45 abstract words selected from Japanese textbooks.
- The screening test of reading and writing for Japanese primary school children (STRAW): This test was also conducted to examine the children's reading and writing abilities. Since preschool children have not yet learned Katakana or Kanji characters, the test for these children only included Hiragana characters.
- The syntactic processing test of aphasia (STA): The STA, a syntax test that is like the test for the reception of grammar (TROG) for Japanese language users, is a test that evaluates the comprehension and production ability of syntactic structures.
- Criterion-referenced testing (CRT) for Japanese language and mathematics.
- Pervasive development disorders ASJ rating scales (PARS) to determine autistic tendency.
- Raven's colored progressive matrices (RCPM).

Results 1

The results showed that children suffering from hearing loss exceeded the national average of all children with hearing difficulties by at least 60.6% and up to 100% (Fig. 1). A total of 76.2% of all scores exceeded the national average of children with hearing difficulties. On the CRT for Japanese language and mathematics, 70.0% of all scores exceeded the national average of scores obtained by normal-hearing children (Fig. 2). We investigated the factors believed to account for the better performances in these tests.

Methods 2

To determine the factors that allowed the children reported un-

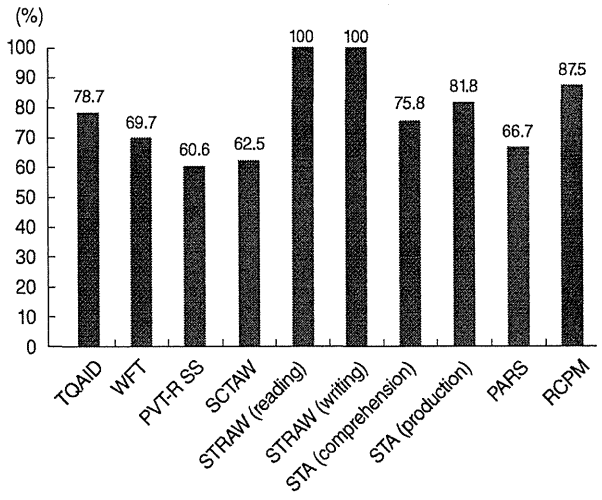

Fig. 1. The results of the various language development tests. The results show that children suffering from hearing loss exceeded the national average of all children with hearing difficulties by at least 60.6% and up to 100%. TQAID, test for question and answer interaction development; WFT, word fluency test; PVTR, Peabody picture vocabulary test-revised; SCTAW, standardized comprehension test of abstract words; STA, syntactic processing test of aphasia; PARS, pervasive development disorders ASJ rating scales; RCPM,Raven's colored progressive matrices.

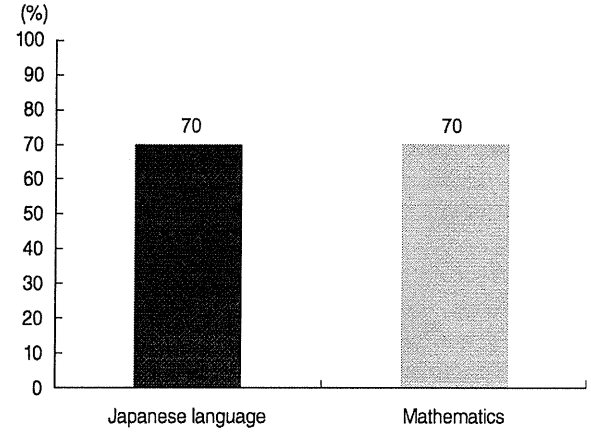

Fig. 2. On the criterion-referenced testing for Japanese language and mathematics, 70.0% of all scores exceeded the national average of scores obtained by normal-hearing children.

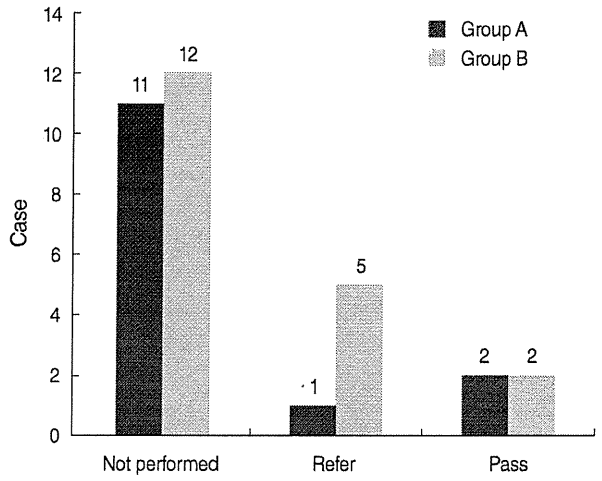

Fig. 3. Whether or not the child went through newborn hearing screening.

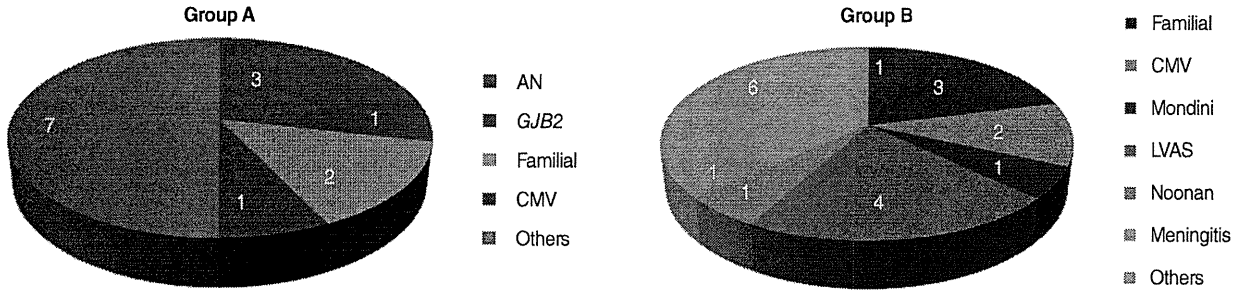

Fig. 4. Causes of deafness. AN, auditory neuropathy; *GJB2*, Gap junction protein, beta-2, 26kDa (*GJB2*) gene mutation; CMV, congenital cytomegalovirus infection; LVAS, large vestibular aqueduct syndrome.

der Results 1 to have better results, we divided the children into two groups. The first group, group A, consisted of 14 children with higher scores in all tests than the national average for children with hearing difficulty. The second group, group B, included 19 children that scored below the national average in any of the tests.

Determining criteria within each group were as follows: 1) whether the child had gone through NHS, 2) the cause for the hearing loss, 3) the age at which the child began to wear hearing aids, 4) the age at which the child received CI, 5) number of visits to our hearing center since initial examination, 6) the amount of time since CI, 7) current average hearing level, 8) current average wearing threshold, 9) whether the child has any siblings, 10) amount of time spent studying at home on a daily basis, 11) educational method (school), 12) the period of integration and the period of auditory verbal/oral education, 13) educational institution child attended before entering primary school.

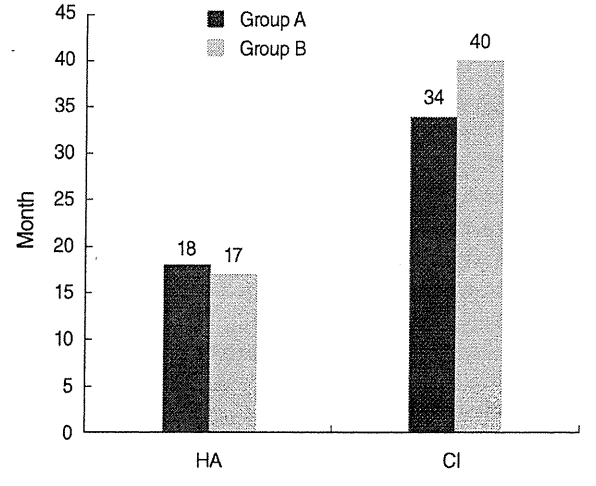

Fig. 5. The mean age for children to start wearing a hearing aid (HA) and cochlear implant (CI).

Results 2

Quite a few children underwent NHS (Fig. 3). Causes for deafness are shown in Fig. 4. There were many cases of inner ear and cochlear nerve anomaly and developmental disabilities in group B. There were no significant differences between the two groups in terms of the mean age for children to start wearing a hearing aid or the mean age for CI (Fig. 5).

The mean period of the visit at our hearing center was significantly longer in group A than in group B (P=0.049 <0.05*) (Fig. 6). The mean wearing period for the cochlear implant was significantly longer in group A than in group B (P=0.02*) (Fig. 6). The mean of the current average hearing level on their CI side was 115 dBHL for group A and 113 dBHL for group B on their CI side. On the non-operation side, it was 102.1 dBHL for group A and 97.1 dBHL for group B. The mean of the present average wearing threshold was 26.8 dBHL for group A and 28.2 dBHL for group B on their CI side. On the non-operation side, it was 59.2 dBHL for group A and 59.3 dBHL for group B. There were no significant differences in these results between the two groups (Fig. 7). Children in group A were more likely to have older siblings; however, there was no significant difference between groups

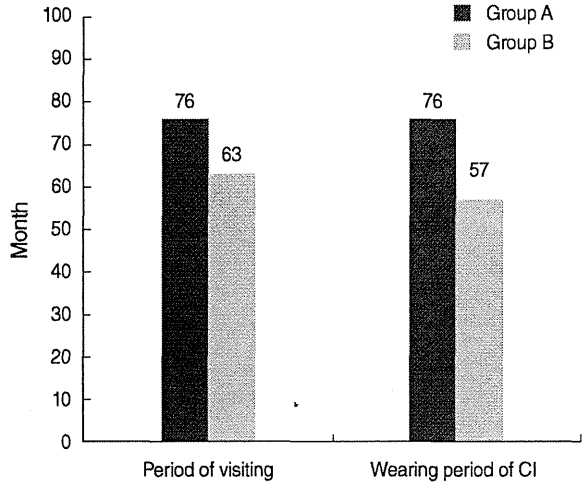

Fig. 6. The mean period of the visit at our hearing center and the mean wearing period of cochlear implant (CI).

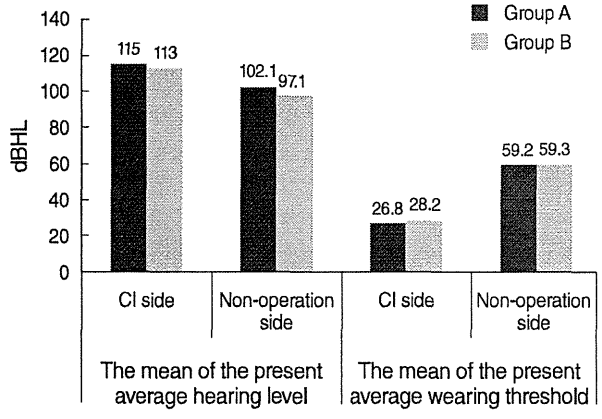

Fig. 7. The mean of the current average hearing level and the present average wearing threshold.

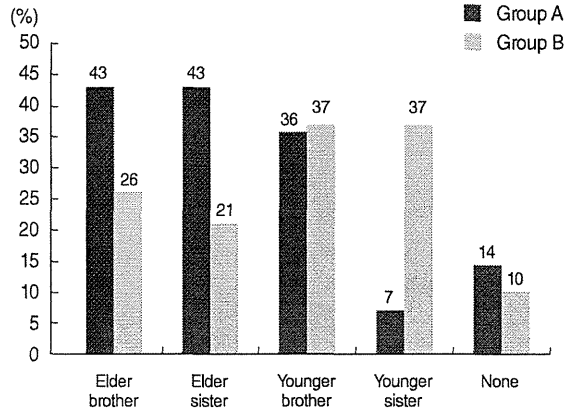

Fig. 8. Whether the child has any siblings.

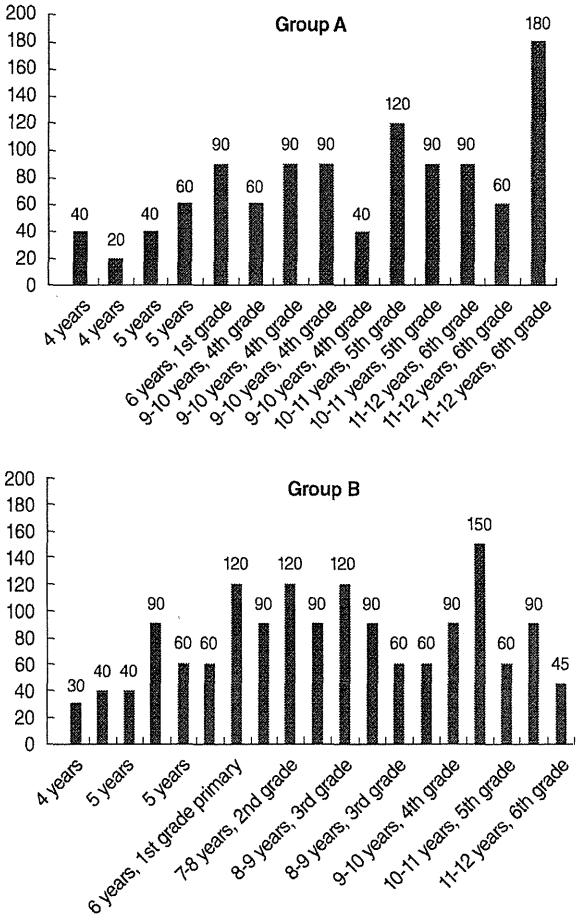

Fig. 9. The amount of time spent studying at home on a daily basis.

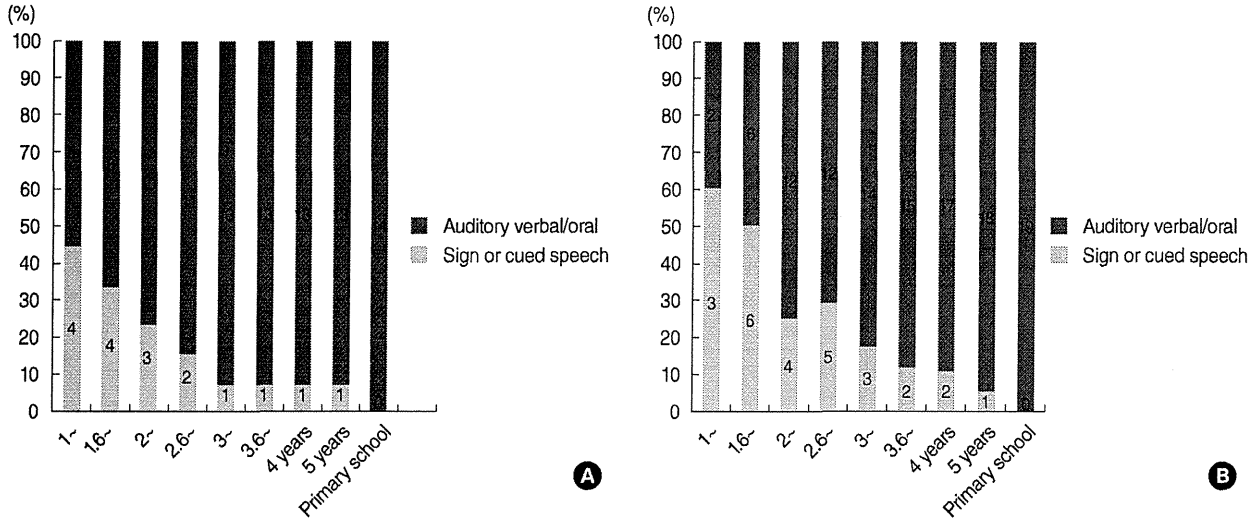

Fig. 10. Educational method (school). A, group A; B, group B.

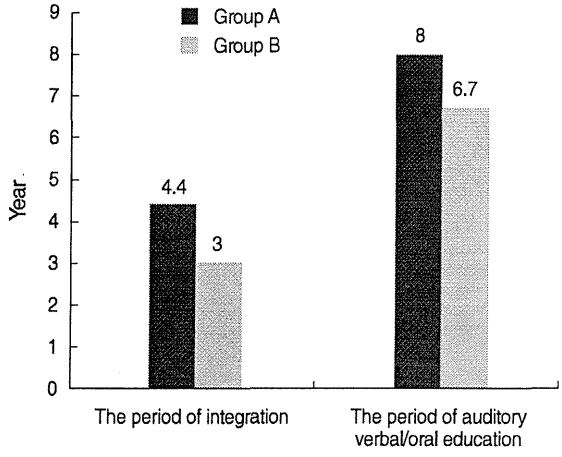

Fig. 11. The period of integration and the period of auditory verbal/oral education.

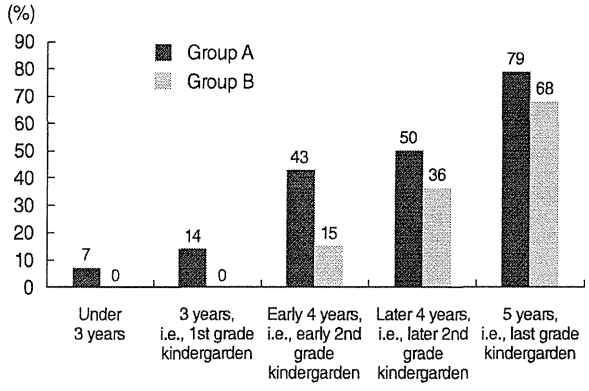

Fig. 12. Educational institution child attended before entrance to primary school.

A and B (Fig. 8). The mean amount of time spent studying at home on a daily basis was 76.4 minutes for group A and 79.2 minutes for group B; these times were not significantly different (Fig. 9). From the age of 1 year to the end of preschool, the education for group A concentrated on auditory verbal and/or oral methods, while that for group B was geared towards sign or cued speech type education; there were significant differences between groups A and B (P=0.003 <0.01**) (Fig. 10).

Children in group A attended regular school for 4.4 years, and those in group B attended for 3 years. Auditory verbal/oral education was 8 years for group A and 6.7 years for group B. While group A's education was longer than that of group B, there were no significant differences between the two groups (Fig. 11). Fig. 12 shows the percentage of children who were integrated into regular kindergarten and nursery school before attending elementary school; there were significant differences between groups A and B (P=0.01*).

DISCUSSION

What factors are associated with good performance in language development in children with cochlear implants? It is very important to gauge the effectiveness of the appropriate intervention for hard-of-hearing infants. Research and evaluation of language development for children with cochlear implants have been conducted and should continue. However, there are very few packages that include multiple language development in the world (1-8).

In 2010, Fukushima et al. planned to assess the current status of hearing impaired children in Japan using the RSCD project, and many tests were used as a part of this nationwide research

project. The study included 638 hearing-impaired children throughout Japan. To enroll hearing-impaired children, the RSCD project set up an open-invitation to various institutions, including schools for the deaf, schools for the hard-of-hearing, mainstream schools, and hospital training rooms.

We conducted multi-directional language development tests as a part of the RSVD in Japan. Overall, the results show that 76.2% of the scores obtained by the children in these tests exceeded the national average scores of children with hearing difficulty. The children that finished above average on all tests: 1). had undergone a longer period of regular habilitation in the rehabilitation center; 2) had their implants earlier in life; 3) were exposed to more auditory verbal/oral communication in their education at affiliated institutions; and 4) were more likely to have been integrated in a regular kindergarten before moving on to elementary school.

In the former report (9), age at diagnosis of hearing loss was not a significant predictor of speech-language outcomes. The children who received auditory-based rehabilitation services during the preschool years demonstrated the potential to develop spoken language communication skills (9). Our findings were similar. The lack of development of spoken language may induce restriction in learning and literacy, substantially compromising educational achievement and employment opportunities later on in life (10). There is a report that the first and second years have a lasting positive impact on language, at least until kindergarten, and the probability that a child would reach normal language levels by kindergarten increased significantly with early intervention and cochlear implant use (11). Niparko et al. (12) reported that younger age at CI was associated with significantly steeper rate increases in comprehension (1.1; 95% confidence interval, 0.5 to 1.7 points per year younger) and expression (1.0; 95% confidence interval, 0.6 to 1.5 points per year younger). Our results were similar. In this study, we suggest that taking the above four factors into consideration will have an affect on the language development of children with severe-profound hearing loss.

CONFLICT OF INTEREST

No potential conflict of interest relevant to this article was reported.

REFERENCES

1. Meinzen-Derr J, Wiley S, Choo DI. Impact of early intervention on expressive and receptive language development among young children with permanent hearing loss. Am Ann Deaf. 2011 Winter; 155(5):580-91.
2. Moon IJ, Kim EY, Chu H, Chung WH, Cho YS, Hong SH. A new measurement tool for speech development based on Ling's stages of speech acquisition in pediatric cochlear implant recipients. Int J Pediatr Otorhinolaryngol. 2011 Apr;75(4):495-9.
3. Lin HC, Yang CC, Chiang YW, Hung PW, Yang EY, Wang L, et al. Effect of identification and intervention age on language development for Mandarin-speaking deaf children with high family involvement. Int J Pediatr Otorhinolaryngol. 2011 Mar;75(3):409-14.
4. Wie OB. Language development in children after receiving bilateral cochlear implants between 5 and 18 months. Int J Pediatr Otorhinolaryngol. 2010 Nov;74(11):1258-66.
5. Wake M, Hughes EK, Poulakis Z, Collins C, Rickards FW. Outcomes of children with mild-profound congenital hearing loss at 7 to 8 years: a population study. Ear Hear. 2004 Feb;25(1):1-8.
6. Hawker K, Ramirez-Inscoe J, Bishop DV, Twomey T, O'Donoghue GM, Moore DR. Disproportionate language impairment in children using cochlear implants. Ear Hear. 2008 Jun;29(3):467-71.
7. Nikolopoulos TP, Archbold SM, Gregory S. Young deaf children with hearing aids or cochlear implants: early assessment package for monitoring progress. Int J Pediatr Otorhinolaryngol. 2005 Feb;69(2):175-86.
8. Lin FR, Ceh K, Bervinchak D, Riley A, Miech R, Niparko JK. Development of a communicative performance scale for pediatric cochlear implantation. Ear Hear. 2007 Sep;28(5):703-12.
9. Lin HW, Furman AC, Kujawa SG, Liberman MC. Primary neural degeneration in the Guinea pig cochlea after reversible noise-induced threshold shift. J Assoc Res Otolaryngol. 2011 Oct;12(5):605-16.
10. Kral A, O'Donoghue GM. Profound deafness in childhood. N Engl J Med. 2010 Oct 7;363(15):1438-50.
11. Moog JS, Geers AE. Early educational placement and later language outcomes for children with cochlear implants. Otol Neurotol. 2010 Oct;31(8):1315-9.
12. Niparko JK, Tobey EA, Thal DJ, Eisenberg LS, Wang NY, Quittner AL, et al. Spoken language development in children following cochlear implantation. JAMA. 2010 Apr 21;303(15):1498-506.

Clinical dermatology • **Concise report**

CED
Clinical and Experimental Dermatology

A case of palmoplantar lichen planus in a patient with congenital sensorineural deafness

A. Ogawa, K. Shimizu, A. Yoshizaki, S. Sato,[1] Y. Kanda,[2] H. Kumagami,[2] H. Takahashi[2] and S. Usami[3]

Department of Dermatology, Nagasaki University Graduate School of Biomedical Sciences, Nagasaki, Japan; [1]Department of Dermatology, Faculty of Medicine, University of Tokyo, Tokyo, Japan; [2]Department of Otolaryngology – Head and Neck Surgery, Nagasaki University Graduate School of Biomedical Sciences, Nagasaki, Japan; and [3]Department of Otolaryngology, Shinshu University School of Medicine, Matsumoto, Japan

doi:10.1111/j.1365-2230.2012.04374.x

Summary

We report a case of palmoplantar lichen planus in a 7-year-old Japanese girl with congenital deafness, who presented with erythematous eruptions and hyperkeratosis, with peeling and fissures on her soles, palms and digits. On histological examination of a skin biopsy from the lesion on her wrist, lichen planus was identified. Using computed tomography of the inner ears, bilateral cochlear dysplasia was found. The patient's DNA was sequenced; no sequence variants were detected in the *GJB2* gene encoding connexin-26, but she had a missense mutation in *SLC26A4* (solute carrier family 26, member 4). Mutations in *SLC26A4* are known causes of hearing loss, but this is a novel mutation, which has not been reported previously. In addition, there have been no reports of cutaneous symptoms in previously reported patients with mutations in *SLC26A4*. To our knowledge, therefore, this is the first report of palmoplantar lichen planus associated with sensorineural deafness accompanied by a mutation in the *SLC26A4* gene.

We report an unusual case of palmoplantar lichen planus (LP) presenting with sensorineural deafness, associated with a mutation in the *SLC26A4* gene.

Report

A 7-year-old girl presented with congenital bilateral sensorineural deafness and skin disorders that had been present since birth. The patient was born at term after an unremarkable pregnancy to non-consanguineous parents. There was no family history of skin disorders or auditory dysfunction.

Correspondence: Dr Asako Ogawa, Department of Dermatology, Nagasaki University Graduate School of Biomedical Sciences, 1-7-1 Sakamoto, Nagasaki, 852-8501, Japan
E-mail: asako56planetes730@yahoo.co.jp

Conflict of interest: none declared.

Accepted for publication 6 November 2011

On physical examination, erythematous eruptions and hyperkeratosis were seen on the soles, palms and digits, with peeling and fissures (Figs 1a,b). A skin biopsy was taken from the lesion on the patient's right wrist.

On histological examination, a band-like lymphocytic infiltration was seen in the upper dermis, with liquefaction degeneration (Figs 2a,b). Examination of the oral cavity and nails was unremarkable, and results of routine blood tests were normal. Specifically, thyroid hormone levels were within the normal limits and no goitre was detected by echo scintigraphy. Computed tomography (CT) scans of the patient's inner ears showed bilateral cochlear dysplasia (Figs 2c,d,e).

Based on these findings, a diagnosis of palmoplantar LP was made. Informed consent was obtained for genetic investigations.

Genomic DNA encompassing the genes *GJB2* (gap junction β-2) and *SLC26A4* (solute carrier family 26, member 4) was amplified by PCR, as described

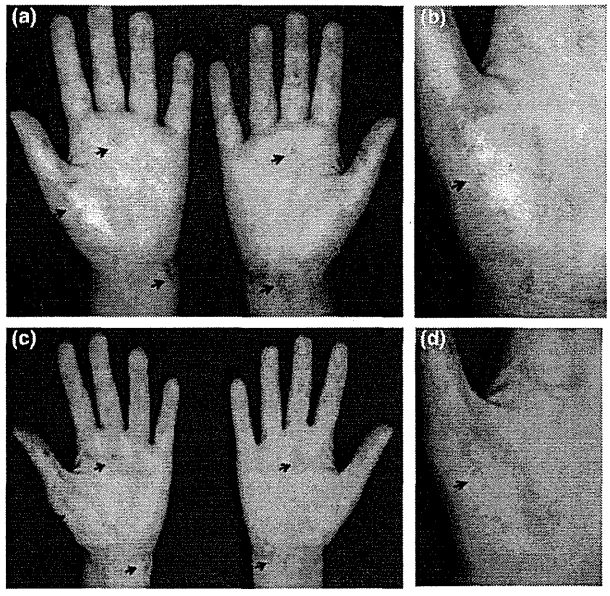

Figure 1 (a,b) Patient at the age of 7 years, showing hyperkeratotic lesions on the palms, wrists and flexor sides of the digits with underlying erythema, fissures and peeling. (c,d) At the age of 15 years, although the hyperkeratotic lesions on the palms showed some improvement, the skin lesions on the palms were unchanged.

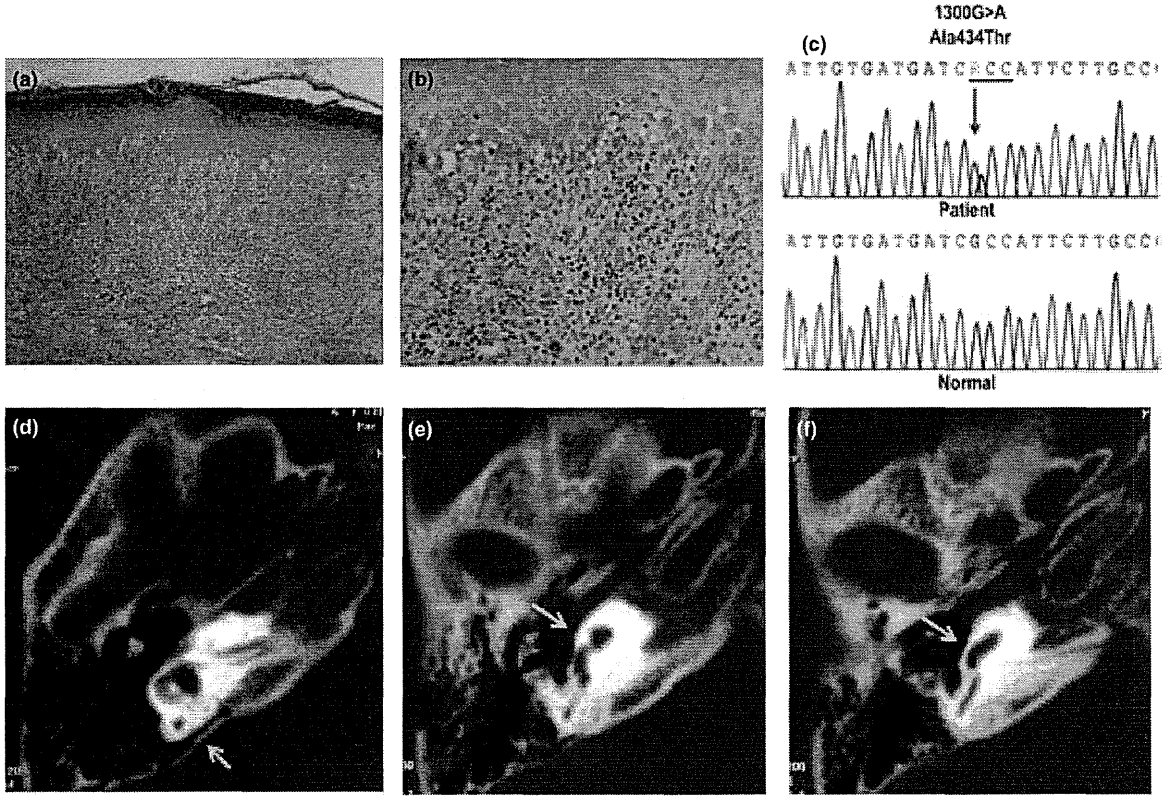

Figure 2 (a) The biopsy taken from the skin lesion on the patient's right wrist revealed hyperkeratosis, epidermal hyperplasia, and a band-like lymphocytic infiltration in the upper dermis (hematoxylin and eosin, original magnification ×40). (b) Liquefaction degeneration and Civatte bodies were detected (hematoxylin and eosin, original magnification ×100). (c) Direct sequencing analysis of the coding region of SLC26A4 revealed a G>A transition of one allele (arrow), which alters the normal alanine codon to a threonine codon. Computed tomography (CT) shows (d) no enlargement of the vestibular aqueduct (arrow); presence of (e,f) cochlear dysplasia: one and a half cochlear turns (arrow).

previously.[1,2] No sequence variants were detected in *GJB2*. The patient had a missense mutation in *SLC26A4* (p.Ala434Thr; c.1300G>A in exon 11) (Fig. 2f). The patient's mother did not have any mutations in this gene, and we did not examine the father's DNA.

The patient was prescribed several topical therapies, including steroid and urea ointment, but the skin eruptions were still visible on the palms, soles and wrists at follow-up 8 years later, at the age of 15 years (Figs 1c,d).

Cutaneous features, such as palmoplantar keratoderma, are sometimes associated with sensorineural deafness.[3] These skin disorders led us to consider conditions associated with mutations in the *GJB2* gene, which encodes connexin 26, such as keratitis–ichthyosis–deafness syndrome and Vohwinkel syndrome.[3] There have been many reports describing deafness caused by other gene mutations, including *SLC26A4*.[4] Mutations in *SLC26A4* cause autosomal recessive deafness; Pendred syndrome (hearing loss with goitre) or DFNB4 (nonsyndromic hearing loss with inner ear abnormalities).[4]

Although over 150 mutations in *SLC26A4* have been described (http://www.healthcare.uiowa.edu/labs/pendredandbor/)[5] our patient had a novel mutation, which has not been published previously, to our knowledge. In addition, there are no reports of Pendred Syndrome or NSHL, accompanied by skin lesions. It is difficult to clarify whether this heterozygous mutation is pathogenic. Yuan Y *et al.*[6] reported that some patients with inner-ear malformation carried only one missense mutation in 2009, thus it seems that such mutations can have a dominant-negative effect and/or produce different phenotypes.

SLC26A4 encodes pendrin, which is a member of the anion transporter family. Anion transporters play critical roles in the formation or preservation of gap junctions, which are related to the exchange of ions and small metabolites between adjoining cells in the epidermis of skin.[7] This intercellular communication system plays an important role in control, coordination of tissue morphogenesis, differentiation and growth.[8] Furthermore, ion channel-related drugs, such as calcium-channel blockers, are well-known causes of LP.[9] Therefore, these facts indicate that abnormality in the exchange of ions in skin tissue can cause skin diseases, including LP.

In conclusion, we report a patient with LP in whom a novel mutation of *SLC26A4* was identified. Further study is needed to confirm the links between LP and disorders of ion channels.

References

1 Kelsell DP, Dunlop J, Stevens HP *et al.* Connexin 26 mutations in hereditary non-syndromic sensorineural deafness. *Nature* 1997; **387**: 80–3.

2 Tsukamoto K, Suzuki H, Harada D *et al.* Distribution and frequencies of PDS (SLC26A4) mutations in Pendred syndrome and nonsyndromic hearing loss associated with enlarged vestibular aqueduct: a unique spectrum of mutations in Japanese. *Eur J Hum Genet* 2003; **11**: 916–22.

3 Lee JR, White TW. Connexin-26 mutations in deafness and skin disease. *Expert Rev Mol Med* 2009; **11**: e35.

4 Pourova R, Janousek P, Jurovcik M *et al.* Spectrum and frequency of SLC26A4 mutations among Czech patients with early hearing loss with and without enlarged vestibular aqueduct (EVA). *Ann Hum Genet* 2010; **74**: 299–307.

5 Cama E, Alemanno MS, Bellacchio E *et al.* Identification of a novel mutation in the SLC26A4 gene in an Italian with fluctuating sensorineural hearing loss. *Int J Pediatr Otorhinolaryngol* 2009; **73**: 1458–63.

6 Yuan Y, You Y, Huang D *et al.* Comprehensive molecular etiology analysis of nonsyndromic hearing impairment from typical areas in China. *J Transl Med* 2009; **7**: 79.

7 Willecke K, Eiberger J, Degen J *et al.* Structural and functional diversity of connexin genes in the mouse and human genome. *Biol Chem* 2002; **383**: 725–37.

8 Richard G, Brown N, Ishida-Yamamoto A, Krol A. Expanding the phenotypic spectrum of Cx26 disorders: Bart–Pumphrey syndrome is caused by a novel missense mutation in GJB2. *J Invest Dermatol* 2004; **123**: 856–63.

9 Swale VJ, McGregor JM. Amlodipine-associated lichen planus. *Br J Dermatol* 2001; **144**: 920–1.

臨　　床

長崎県における公的全県新生児聴覚スクリーニング4年半の経過

川田　晃弘[1)]・神田　幸彦[1)2)]・隈上　秀高[1)]・原　　稔[1)]
道祖尾　弦[1)]・高崎　賢治[1)]・高橋　晴雄[1)]

Universal Newborn Hearing Screening in Nagasaki Prefecture

—Experience for 4. 5 Years—

Akihiro Kawata, Hidetaka Kumagami, Minoru Hara,
Yuzuru Sainoo, Kenji Takasaki and Haruo Takahashi

(Nagasaki University Hospital)

Yukihiko Kanda

(Nagasaki University Hospital, Kanda E·N·T Clinic, Nagasaki Bell Hearing Center)

New born hearing screening was started on a trial basis in Japan in 1997.

Community-based newborn hearing screening started in Nagasaki prefecture in October 2003. We investigated 45, 924 infants who underwent universal newborn hearing screening from October 2003 to March 2008.

Ninety-four to ninety-six percent of neonates in Nagasaki prefecture were screened in from 2005 to 2008. The referral rate after the screening process was 0. 5-0. 7%. The incidence of hearing loss was 0. 15-0. 20%.

We compared the age in lunar months of the screened and non-screened infants at the time of diagnosis, intervention, fitting of the first hearing aid, and the age of cochlear implantation. The age at the time of diagnosis (p<0. 0001), intervention (p<0. 0001), fitting of the first hearing aid (p<0. 0001), and the age at which cochlear implantation was performed (p=0. 0025<0. 01) were significantly lower in the screened infants.

These results show that universal new born hearing screening is very useful for early detection of, and intervention for infants with hearing loss.

The integration of medical, educational, and social assistance is very important for infants diagnosed as having hearing loss after newborn hearing screening.

Keywords : universal newborn hearing screening, hearing aid, cochlear implant

はじめに

1960年代から米国を中心に行われるようになった新生児聴覚スクリーニングの試みは，1990年代から短時間で検査を可能にする自動聴性脳幹反応（以下自動ABR）や耳音響放射（以下OAE）の機器の出現により，欧米では飛躍的に普及した．米国では1993年にNational Institutes of Health（NIH）が，すべての新生児に生後3ヵ月までに聴覚スクリーニングを受けることを推奨して以後[1)]，

1）長崎大学大学院医歯薬学総合研究科　展開医療科学講座　耳鼻咽喉・頭頸部外科学分野
2）神田E・N・T医院，長崎ベルヒアリングセンター（長崎市）

1994年には米国耳鼻咽喉科学会，米国小児科学会および米国言語聴覚学会による Joint Committee on Infant Hearing（以下JCIH）が「生後3ヵ月までに聴覚障害を診断し，生後 6ヵ月までに介入することが望ましい」と勧告し[2]，全出生児対象の新生児聴覚スクリーニングの法制化が進んだ．10年以上経過した現在，米国では実施率は90%を越えその体制は確立されているが，一方で多数の診断所見が報告されない例や，結果の書類提示がない例などの問題点も発生しているとの報告がある[3)~5)]．

また，本邦における公的な新生児聴覚スクリーニングは聴覚障害の早期発見・診断および早期療育を目的とし，国の新生児聴覚検査モデル事業として 2001 年度に岡山県，秋田県，神奈川県，栃木県の4県で開始された[6]．長崎県においても国庫補助事業として，耳鼻咽喉科，産婦人科，小児科，療育機関，行政，県医師会が連携して，2003年10月より公的全県新生児聴覚スクリーニングが開始された．県主導での公的スクリーニングは2007年度で終了したが，2008年度からは県内の全市町が実施主体となり事業が継続されている．2008年度のスクリーニング実施率は96%となり，2005年からは4年間94%以上を維持しており，全国の報告をみてもこのように高い実施率を4年以上継続した報告は岡山県[7]以外，われわれが渉猟した範囲ではまだみられない．長崎県において施行されている公的全県新生児聴覚スクリーニングの4年半の経過について結果の検討を行い若干の考察を加え報告する．

対象と方法

2003年10月から2008年3月までの4年半に長崎県で出生した新生児59,132名のうち，長崎県新生児聴覚検査推進事業において公的スクリーニングの対象となる，長崎県に住所を有する新生児は50,307名であった．そのうちスクリーニング検査を希望し施行された新生児45,924名を対象とした．これは県内出生児数の91.3%にあたる．

分娩施設（スクリーニング実施機関）に入院中に自動ABRまたはOAE（約90%がDPOAE，10%がTEOAE）を用いて初回検査を行い，refer（要再検）であった児は退院前に再検査が行われた．再検査は初回検査と同じ検査を行い，再検査においても refer であった児については耳鼻咽喉科（精密検査実施医療機関）で確認検査および精密検査を行った．確認検査において，OAE で refer だった児については自動ABRでの検査を行い，また自動ABRで refer であった児は精密検査を行っている（図1）．ABR，聴性行動反応聴力検査（BOA），条件詮索反応聴力検査（COR）などで総合的に判断した結果，聴覚障害ありと診断された児については，必要に応じて療育機関へ紹介し，療育の介入を開始した．それ以外の児についても精密検査実施医療機関で定期的な聴力評価と経過観察を

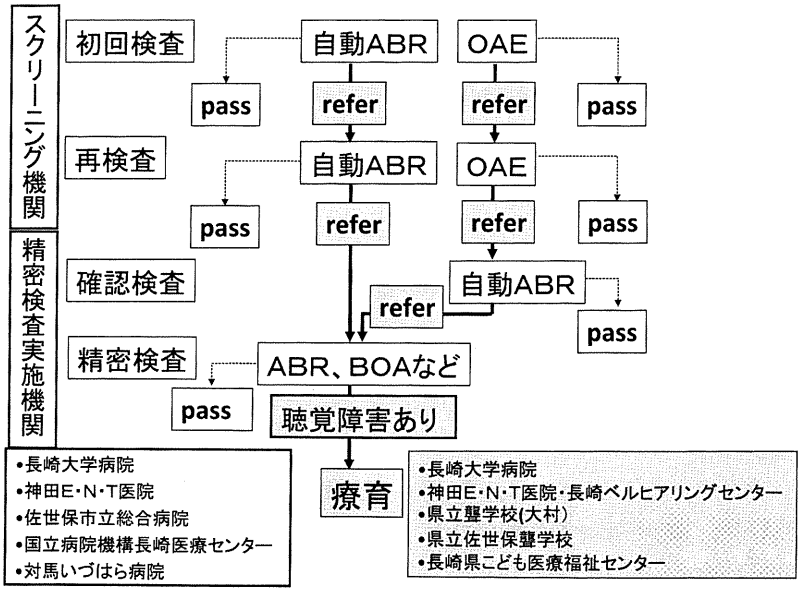

図1　スクリーニングの流れ
長崎県におけるスクリーニングの流れを示す．

行った（図1）．スクリーニング結果については長崎県においては県で集約され長崎県新生児聴覚検査推進事業検討協議会において例年報告会が行われており，そこでのデータをもとに年度ごと（各年度4月～翌年3月末，2003年度のみ10月～翌3月末までの6ヵ月）にスクリーニング受診状況および結果の検討を行った．

また，新生児聴覚スクリーニング4年半で最終的に両側聴覚障害ありと診断された児36名の経過を追い，両側平均聴力70 dBHL以上の難聴を認めた児29名について診断時期・介入時期を，また補聴器を装用した児16名について補聴器開始時期を，新生児聴覚スクリーニングが始まる前（1997年～2000年の4年間）の両側平均聴力70 dBHL以上の難聴児35名のデータと比較検討した．さらに人工内耳小児症例で長崎県在住の小児において言語習得前の症例を新生児聴覚スクリーニング施行例と未施行例に分けて検討した．

結　果

1．スクリーニング受診状況と結果

2003年度は対象児6,132名のうちスクリーニング検査受診児4,750名で受診率77%であったが，以後年々受診率は上がり，2005年度からは94%以上を維持している（表1）．2007年度には対象児10,833名のうち10,331名が検査を受けており受診率は95%に達した（表1）．スクリーニングを受けた児のうち毎年0.5～0.7%が精密検査を耳鼻咽喉科で受けており，そのほぼ半数が正常聴力であった（表2）．また，スクリーニングで聴覚障害ありと診断された児は4年半で80名（0.17%）おり，そのうち両側性は32名（40%），一側性は48名（60%）であった（表2）．検査を行った範囲では先天性サイトメガロウイルス感染症の児はいなかった．一側性難聴はそのほとんどが定期的な経過観察となっていたが，途中でドロップアウトし観察不能となった例もみられた．

2．スクリーニング後両側聴覚障害ありと診断された児の経過

スクリーニング後の経過観察で最終的に両側聴覚障害ありと診断された児は，スクリーニング時点の32名から4名増加し，36名であった．増加した4名の中にはオーディトリーニューロパシーが1名みられている．これはOAEにより診断できなかった症例である．また他の3名はNICUに長期入院した後に検査し発見された．両側聴覚障害を診断された36名のうち，補聴器を装用している児が7名，人工内耳手術を受けた児が9名であった．その他20名は，軽度から中等度難聴の児で経時的に聴力が良くなり補聴器なしで経過をみている児や，聴覚以外にも重度の障害があり発達の経過を追っている児であっ

表1　スクリーニングの受診状況

	2003	2004	2005	2006	2007	計
対象児（名）	6,132	11,453	10,819	11,070	10,833	50,307
スクリーニング受診児	4,750 (77%)	10,220 (89%)	10,231 (95%)	10,392 (94%)	10,331 (95%)	45,924 (91%)

各年度は4月～翌3月までのものを示す（ただし，2003年度のみ10月～翌3月まで）．

表2　スクリーニングの結果

	2003	2004	2005	2006	2007	計
受診児（名）	4,750	10,220	10,231	10,392	10,331	45,924
要精密検査	30（0.6%）	64（0.6%）	66（0.6%）	72（0.7%）	55（0.5%）	287（0.6%）
正常	15	31	34	39	29	148
聴覚障害	8（0.17%）	17（0.17%）	15（0.15%）	21（0.20%）	19（0.18%）	80（0.17%）
両側	4	9	7	7	5	32
一側	4	8	8	14	14	48
その他	7	16	17	12	7	59

各年度は4月～翌3月までのものを示す（ただし，2003年度のみ10月～翌3月まで）．
その他には，未受診・経過観察などが含まれる．

表3　スクリーニング後，両側聴覚障害ありと診断された児の経過

	2003	2004	2005	2006	2007	計
補聴器装用	2	1	1	0	3	7
人工内耳装用	2	1	2	3	1	9
その他	2	8	4	4	2	20
計	6	10	7	7	6	36

各年度は4月～翌3月までのものを示す（ただし，2003年度のみ10月～翌3月まで）．
その他には，軽度～中等度難聴児や経時的に聴力が良くなり補聴器なしで経過を見ている児，聴覚以外にも重度の障害を有している児，転居，死亡などが含まれる．

た．（表 3）．高度難聴と診断された児で聴力の回復が認められた児はいなかった．

補聴器および人工内耳を用いて聴覚口話で療育されている16名の原因疾患の内訳は，前庭水管拡大で進行性難聴が3名，GJB2遺伝子変異が2名，家族性が6名，不明5名であった．原因不明の5名には，双生児未熟児2名，多指症2名を認めた．

3．診断・介入時期・補聴器装用開始時期・人工内耳手術時期

新生児聴覚スクリーニング4年半で，両側中等度以上の難聴を認めた29名の診断時期は平均2.2ヵ月であり，療育の介入時期は平均3.2ヵ月であった．また，29名のうち補聴器を装用した16名（人工内耳手術を受けた児9名を含む）の補聴器装用開始時期は平均10.2ヵ月であった．これらはスクリーニングのない1997年から2000年の4年間の両側中等度以上の難聴児の診断時期の平均17.2ヵ月（$p<0.0001$），療育の介入時期の平均21.7ヵ月（$p<0.0001$），補聴器開始時期の平均24.7ヵ月（$p<0.0001$）と比べて統計学的に有意に早くなっていた（図2）．さらに，2010年8月現在，長崎大学における人工内耳小児症例で長崎県在住の小児において，言語習得前の症例38例を新生児聴覚スクリーニング施行例と未施行例に分けて検討したが，新生児聴覚スクリーニング施行例14例の人工内耳手術月齢は平均36.2ヵ月で導入前の未施行例では平均56.0ヵ月と統計学的に有意に早かった（$p=0.0025<0.01$）（図3）．

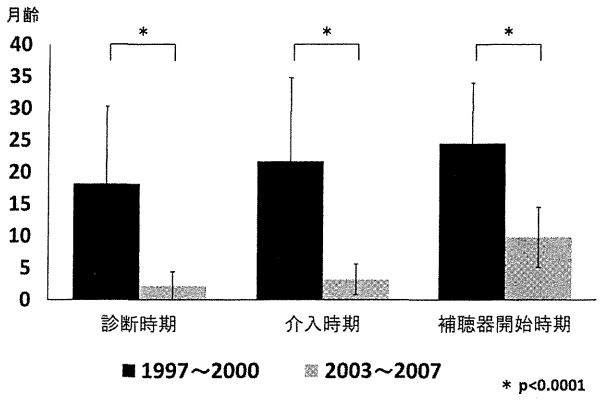

図2　診断・介入時期，補聴器装用開始時期
診断時期・介入時期・補聴器装用開始時期ともに新スク導入前と導入後では新スク導入後早くなっており，統計学的に有意差を認める．（t検定，$p<0.0001$）

考　察

米国のJCIHでは，スクリーニングの体制について1）95%以上の受診率，2）95%のフォローアップ（経過観察）率，3）4%以下のrefer率，4）3%以下の偽陽性率が望ましいと勧告している[8]．長崎県のスクリーニング体制について結果を検討すると，受診率に関しては，2007年には95%に達しており，2008年度以降も市・町により新生児聴覚検査事業が継続されており，2009年度の長崎県新生児聴覚検査推進事業検討協議会報告によると2008年度は96%であった．経過観察率に関しては，4年半で聴覚障害ありと診断された児80名のうち両側性は30名（93.8%），一側性は42名（87.5%）を経過観察しており，全体では経過観察率90%であった．refer率は0.5～0.7%であり4%以下であった．refer率が0.5～0.7%ときわめて低いのは長崎県において2段階スクリーニング体制をとっているためと思われる．また，実際は正常であったのにスクリーニングでreferとなったのは，45,924名中148名で偽陽性率は0.32%で3%以下であった．以上のことより，長崎県におけるスクリーニング体制はほぼ確立されてきている．米国ではスクリーニング率がアップし長年90%以上に達しているが，経過観察や報告の欠如が問題視されており，30%以上が書類による報告

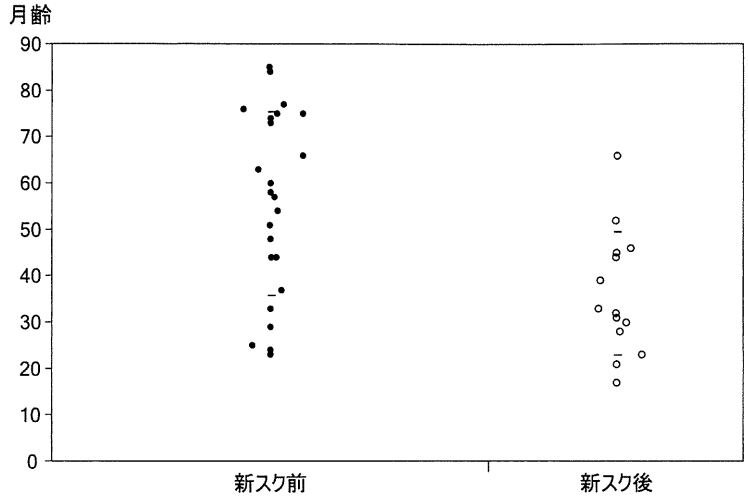

図3　人工内耳手術月齢

●は新スク導入前，○は新スク導入後である．縦軸は月齢を示す．言語習得前小児の人工内耳手術月齢は新スク導入前と導入後では新スク導入後早くなっており，統計学的に有意差を認める．(t 検定，$p<0.01$)

が行われず，介入のサービスを受けているかどうかの立証ができていない場合があると報告している[3]．米国ヘルスケアが新生児聴覚スクリーニングに関して高度に優先される項目として，1) 診断，2) 治療，3) 両親や社会の認知，4) 質的改善の継続，5) 結果の管理を推奨している[5]が，長崎県でも，今後質的な改善の継続や報告の義務と結果の管理の精度アップと継続を続けて行く事が課題である．また，スクリーニングの開始により診断時期は平均 2.2ヵ月，介入時期は平均 3.2ヵ月と早くなり，JCIH の「生後 3ヵ月までに聴覚障害を診断し，生後 6ヵ月までに介入することが望ましい」という勧告を満たしており，スクリーニングの目的である早期診断・早期療育への介入は達成できていると考えられる．補聴器装用開始時期はスクリーニング開始前の平均24.7ヵ月から開始後は平均 10.2ヵ月となっており，三科[9]，新正ら[10]の報告と同様に著明に早くなっていた．また，人工内耳の手術月齢についても新生児聴覚スクリーニング施行例と未施行例に分けて検討したが，施行例は手術月齢平均 36.2ヵ月，未施行例は平均 56.0ヵ月で施行例が有意に早かった．人工内耳の手術月齢が低いほど言語能力は，理解・表出ともに発達が良好である事は広く知られている[11)～14)]．手術月齢が早くなった事は，新生児聴覚スクリーニングにより早期診断・早期療育が可能となり，早期に人工内耳手術のみきわめができるようになったためであろう．

ポーランドでは国家をあげてスクリーニングが行われており，4 年間の報告では出生児の 96.3%にあたる 1,392,427 名の新生児がスクリーニングを受け 2485 名の難聴が診断，312 名が最重度難聴児（0.02%）で 145 名が高度難聴児（0.01%）であった[15]．われわれの 4 年半の検討でも人工内耳症例になるほどの最重度難聴児は言語習得前に限ると 45,924 名中 9 名の 0.0195%であり，ほぼ同程度であった．ポーランドのように国全体で新生児聴覚スクリーニングを行い，報告も的確にできる事は，先天性の聴覚障害児の将来および地域格差のない社会を考えると今後わが国においても重要な課題と考える．また栃木県の報告[16]では，療育体制の整備なくして新生児聴覚検査事業は継続できないとして終了した事実に関連して，難聴児に対する療育，教育を含めた事後措置を十分行える体制が整っていなければ，Universal newborn hearing screening の意義は失われるという意見[17]を挙げ，早期発見のみでは意味がなく，療育，教育など十分な事後措置が行える事が重要と述べている．

ま と め

1. 長崎県において施行されている公的全県新生児聴覚スクリーニングの 4 年半の経過について結果の検討を行った．

2. 2003 年 10 月より開始されたが 2005 年度からは実施率が 4 年連続 94%を越え，聴覚障害と診断された児は

HRCT-BASED PREDICTION FOR COCHLEAR IMPLANT OUTCOMES OF CASES WITH INNER EAR AND INTERNAL AUDITORY CANAL MALFORMATIONS

Hiroshi Yamazaki,[1,3] Sho Koyasu,[2] Saburo Moroto,[1] Rinko Yamamoto,[1] Tomoko Yamazaki,[1] Keizo Fujiwara,[1] Kyo Itoh,[2] Yasushi Naito[1,3]

[1]*Department of Otolaryngology, Kobe City Medical Center General Hospital;* [2]*Department of Radiology, Kobe City Medical Center General Hospital;* [3]*Institute of Biomedical Research and Innovation, Kobe, Japan*

Introduction

Inner ear and internal auditory canal (IAC) malformations account for approximately 20-35% of congenital sensorineural hearing loss[1,2] and an increasing number of children with inner ear and/or IAC malformations underwent cochlear implantation. According to Sennaroglu's classification of inner ear malformations, which is the most widely accepted, the inner ear malformations are divided into labyrinth aplasia, cochlear aplasia, common cavity (CC), incomplete partition type I (IP-I), type II (IP-II), and type III (IP-III), cochlear hypoplasia type I (CH-I), type II (CH-II), and type III (CH-III), and large vestibular aqueduct syndrome (LVAS).[1,3] This classification is essential to investigate the etiology of the inner ear malformations, but with respect to predicting cochlear implant (CI) outcomes, it might not be enough, because it does not include IAC malformations such as narrow IAC (NIAC) and hypoplasia of the bony cochlear nerve canal (HBCNC). These IAC malformations are highly associated with cochlear nerve deficiency (CND), which has a negative impact to CI outcomes.[4,5]

The purpose of this study was to establish a new CT-based categorization which is simple and includes both inner ear and IAC malformations for predicting CI outcomes.

Materials and methods

Between 2004 and 2010, 98 subjects who were under 20 years old underwent cochlear implantation at Kobe City Medical Center General Hospital. Among them, CT revealed that 24 subjects had inner ear and/or IAC malformations at the implanted side.

We evaluated inner ear and IAC malformations at the implanted side based on CT findings. Sennaroglu's classification was used to classify inner ear malformations and the IAC malformations were classified into NIAC and HBCNC. NIAC was diagnosed when the maximum diameter of the IAC was less than 2 mm.[2] The width of the bony cochlear nerve canal (BCNC) was evaluated at the mid-portion between the base of the modiolus of a cochlea and the fundus of the IAC on axial images. When the diameter of the BCNC is less than 1.5 mm, it is diagnosed as HBCNC.[5] CND was diagnosed when a cochlear nerve (CN) appeared smaller than the facial nerve on the parasagittal MR imaging.

We categorized inner ear and IAC malformations into four groups by two criteria: (1) the presence or absence of a bony modiolus in the cochlea; and (2) the diameters of IAC and BCNC. In this categorization, both Group 1 and Group 3 have a bony modiolus in the cochlea, while Group 2 and Group 4 lack this component. Both IAC and BCNC are normal in Group 1 and Group 2, but NIAC or HBCNC was observed in Group 3 and Group 4. Sennaroglu's classification of inner ear malformations clearly discriminates between

Address for correspondence: Yasushi Naito MD, PhD, Department of Otolaryngology Kobe City Medical Center General Hospital, 650-0047 2-1-1 Minatojima Minamimachi Chuo-ku, Kobe City, Japan. naito@kcho.jp

Cholesteatoma and Ear Surgery – An Update, pp. 371-373
Edited by Haruo Takahashi

the presence and absence of a bony modiolus in the cochlea. According to his classification, a bony modiolus is present in IP-II, CH-III, LVAS, and a normal inner ear, while CC, IP-I, IP-III, CH-I, and CH-II have a cystic cavity without a bony modiolus.[3]

We evaluated CI outcomes by category of auditory performance (CAP) scores,[6] hearing thresholds of pure-tone sounds, infant word speech discrimination scores, and monosyllabic word speech discrimination scores at one to three years after implantation. A subject with 0-4 CAP scores could not even understand common phrases without visual language and, therefore, we defined 5-7 CAP scores as a good CI outcome and 0-4 CAP scores as a poor one.

Results

We categorized our patients based on the two criteria as described above. In this study, there was no case categorized in Group 4. Group 1, Group 2, and Group 3 consisted of 11, 7, and 6 cases, respectively. MR imaging revealed CND in all cases of Group 3.

The post-operative CAP score was equal or over five in all cases of Group 1, but did not exceed four in all of Group 3. In Group 2, the post-operative CAP score was still four in two cases even after three years of CI usage, but reached to five or six in the remaining five cases. As shown in Figure 1, using our new categorization instead of the existing classifications, we can better discriminate between a good and poor outcome.

We examined speech discrimination scores of 22 cases except for two cases of Group 3 whose response to voice was poor. The correct percentage of the closed-set infant word discrimination test was ≥ 80 in all cases of Group 1, while the score ranged from 40 to 60 in tested cases of Group 3. The correct percentage of Group 2 widely varied between cases, ranging from 55 to 100. The open-set monosyllabic word discrimination test is much more difficult than the closed-set infant word discrimination test and, therefore, only 17 of 24 patients, who were over five years old and used their CI for more than two years, underwent this examination. All tested cases of Group 1 and 3 cases of Group 2 could answer correctly in equal or over 80% of accuracy. The correct percentage of the remaining cases, including all tested cases of Group 3, was ≤ 30.

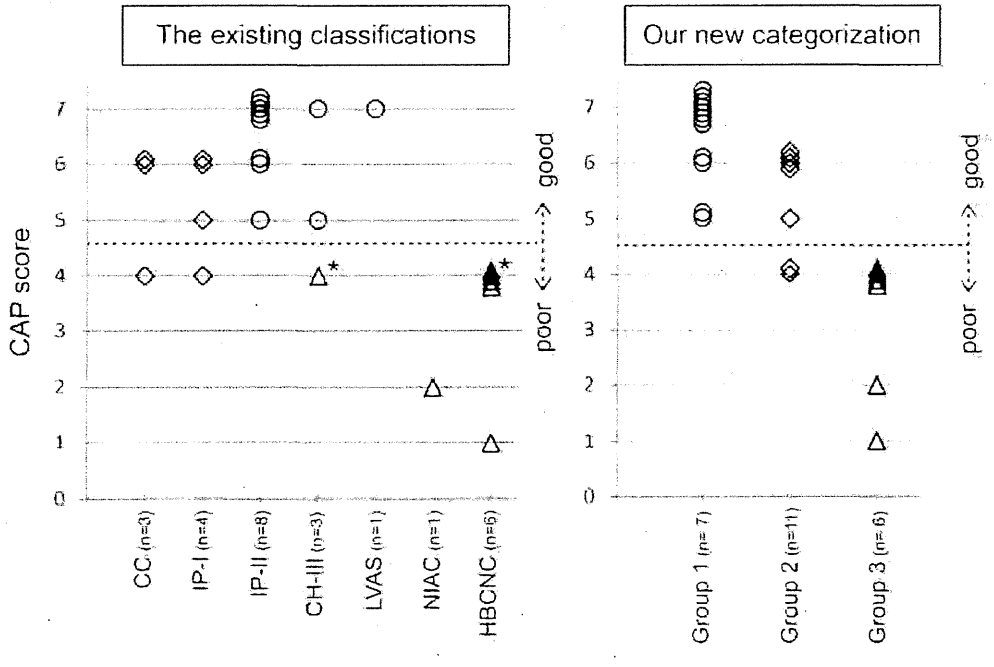

Fig. 1. A. The post-operative CAP score of each type of malformations based on the existing classifications. One case with both CH-III and HBCNC is plotted twice (*). B. The post-operative CAP score of each group of our new categorization. In both graphs, the members of Group 1, Group 2, and Group 3 are represented by a circle, diamond, and triangle, respectively.