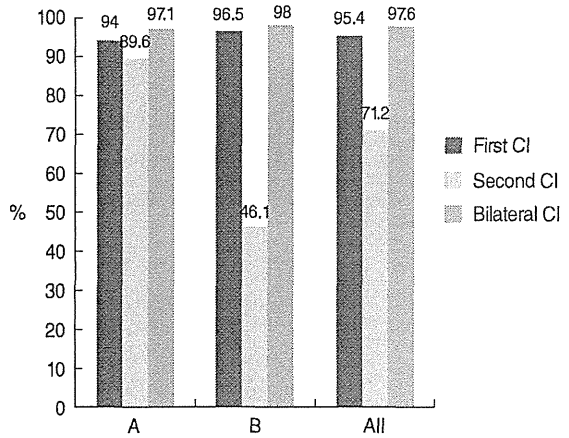

Fig. 8. Mean word recognition score at 1 m from front speaker at 70 dB SPL (0.13). CI, cochlear implantation; SPL, sound pressure level.

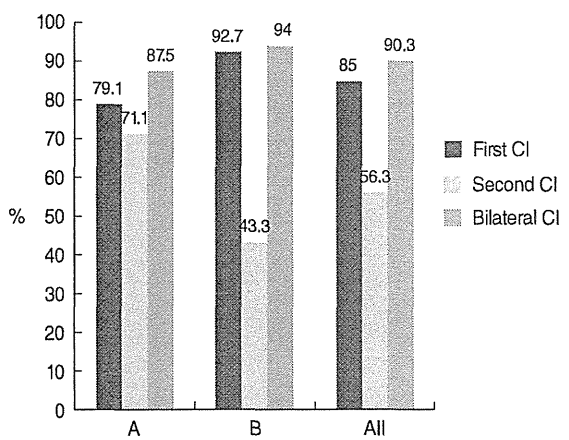

Fig. 9. Mean word recognition score at 1 m from front speaker at 60 dB SPL (P=0.05). CI, cochlear implantation; SPL, sound pressure level.

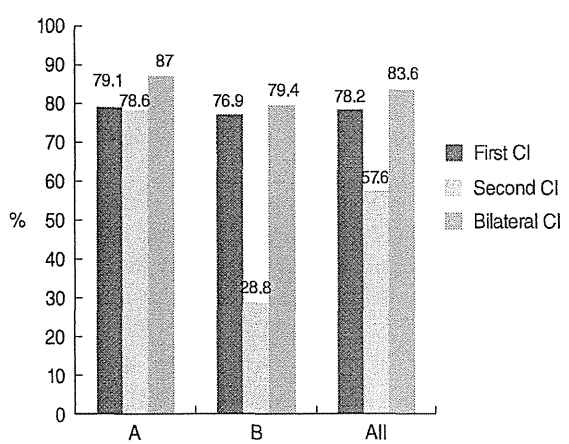

Fig. 10. Mean speech discrimination score at 70 dB SPL (P=0.02*). CI, cochlear implantation; SPL, sound pressure level.

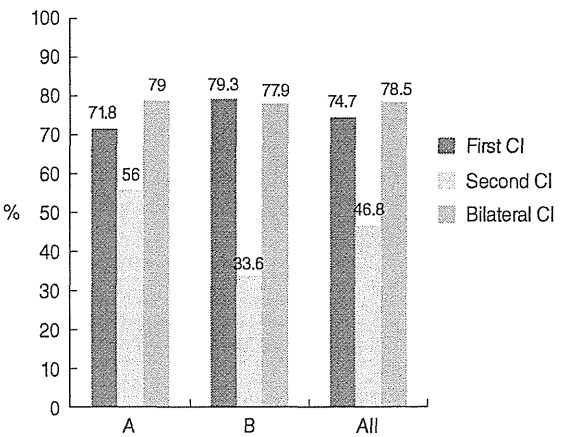

Fig. 11. Mean speech discrimination score at 60 dB SPL (P=0.24). CI, cochlear implantation; SPL, sound pressure level.

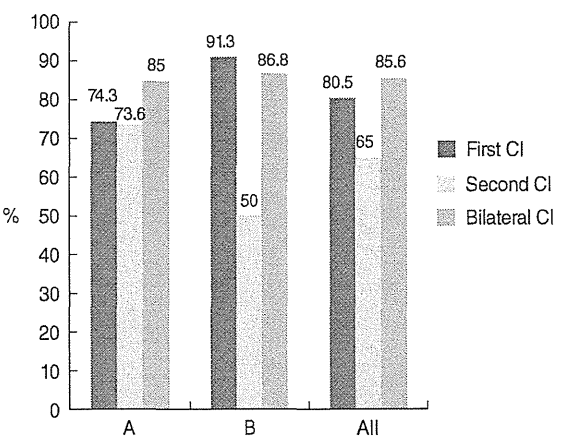

Fig. 12. Mean speech discrimination score at 1 m from second cochlear implantation (CI) side speaker at 70 dB SPL (P=0.25). SPL, sound pressure level.

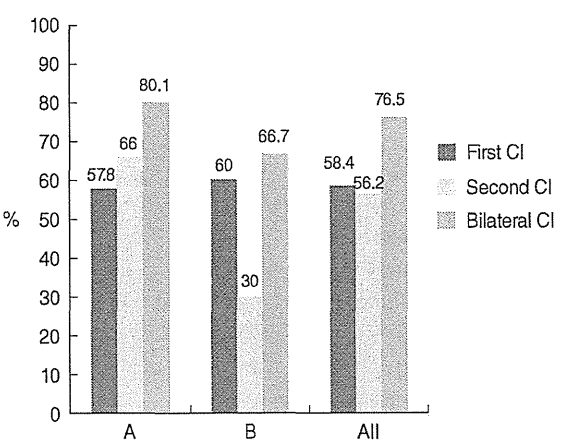

Fig. 13. Mean speech discrimination score at 1 m from second cochlear implantation (CI) side speaker at 60 dB SPL (P=0.02*). SPL, sound pressure level.

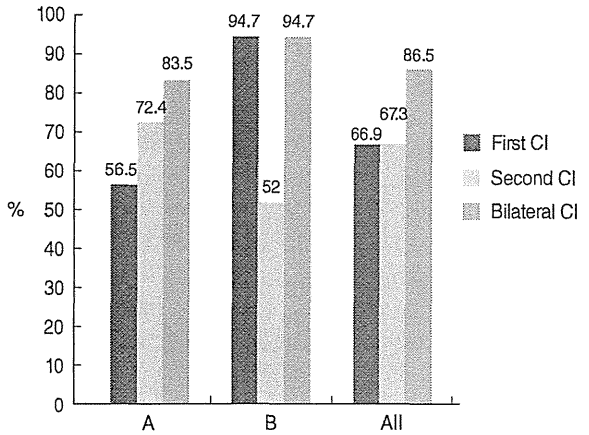

Fig. 14. Mean word recognition score at 1 m from second cochlear implantation (CI) side speaker at 60 dB SPL (P=0.02*). SPL, sound pressure level.

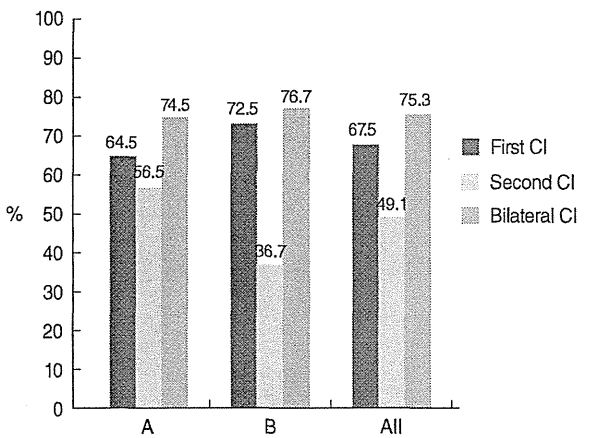

Fig. 15. Mean speech discrimination score at 1 m from front speaker under the noise (S/N=80/70, +10; P=0.01*). CI, cochlear implantation.

The mean WRS at 1 m from second CI side SP at 60 dB SPL were described in Fig. 14. The mean score for second CI in group A was superior to the mean score for first CI. There were significant differences between the results of first CI and bilateral CI at 1 m from second CI side SP on all cases at 60 dB SPL (P=0.02*).

The mean SDS at 1 m from front SP under the noise (S/N= 80/70, +10) were described in Fig. 15. The mean score for second CI in group A was similar to the one for first CI. There were significant differences between the results of first CI and bilateral CI under the noise at 1 m from front SP on all cases (P=0.01*). The mean WRS at 1 m from front SP under the noise (S/N=80/ 70, +10) were described in Fig. 16. The mean score for second CI in group A was superior to the mean score for first CI. In all cases, there were significant differences between the results of first CI and bilateral CI under the noise at 1 m from front SP (P=0.002**).

We attempted to determine until what age the second CI is effective for better language perception in various situations. We

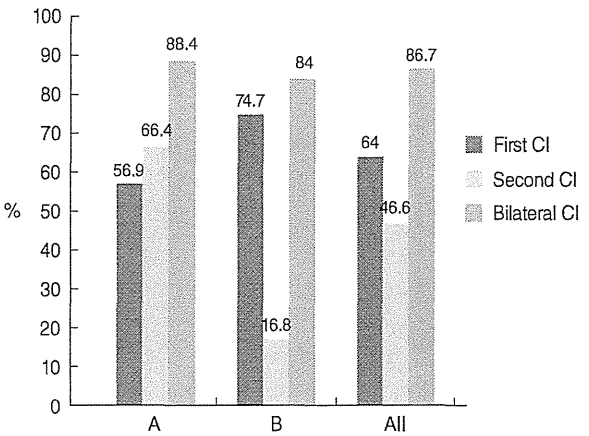

Fig. 16. Mean word recognition score at 1 m from front speaker under the noise (S/N=80/70, +10; (P=0.002**). CI, cochlear implantation.

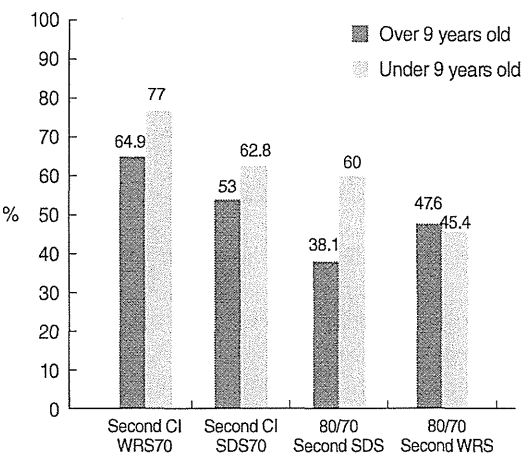

Fig. 17. Comparison of the mean word recognition score (WRS) and speech discrimination score (SDS), at 70 dB SPL at 1 m from front speaker, the mean SDS and WRS under the noise (S/N=80/70) on second cochlear implantation for over and under 9 years old. SPL, sound pressure level.

compared the results of over 9 years old with the results of under 9 years old, analyzing the mean WRS and SDS at 70 dB SPL at 1 m from the front SP, the mean SDS and WRS under the noise (S/N=80/70) on second CI (Fig. 17). The mean SDS under the noise (S/N=80/70) for the second CI (P=0.04*) shows significant differences between the over 9 years old and the under 9 years old.

We compared children that had not used their HA for over 3 years before the second CI with those that had used their HA within 10 months before the second CI using various speech understanding tests (Fig. 18). The mean WRS and SDS revealed better scores for HA usage within 10 months before the second CI than for those who stopped using their HA 3 years or more before the second CI. Especially on the WRS and SDS under the noise, there were significant differences between these two groups (P=0.01* on SDS and P=0.04* on WRS).

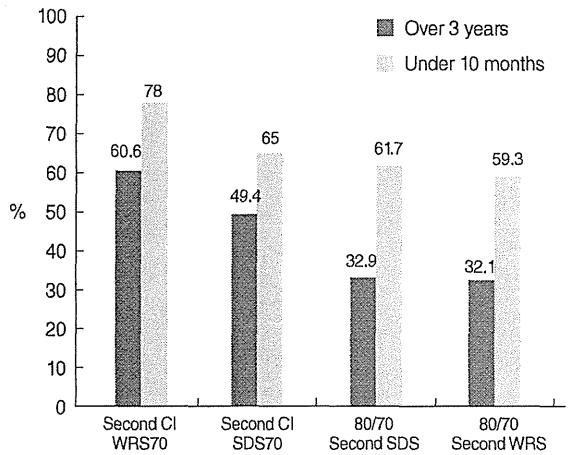

Fig. 18. Comparison of the mean word recognition score (WRS) and speech discrimination score (SDS) at 70 dB SPL at 1 m from front speaker, the mean SDS and WRS under the noise (S/N=80/70) on second cochlear implantation (CI) for those that had not used their hearing aid (HA) for over 3 years before second CI with those that had used their HA within 10 months before second CI. SPL, sound pressure level.

DISCUSSION

In all children, the WTH using the second CI was almost the same using the first CI ranging from 25 to 35 dB HL. Also, the WTH using the second CI recovered compared to the WTH using HA before their second CI (P=0.03*). A previous report (6) also describes that aided thresholds give better performance.

At every age, a second CI is very effective. However, the results of under 9 years old were better than the results of over 9 years old on the mean SDS under noise (S/N=80/70) on the second CI (P=0.04*). These results may be due to brain plasticity of the children for acquiring speech understanding under the noise (10, 14).

About use of a HA in their opposite side of first CI, on the WRS and SDS under the noise, there were significant differences between the group of over 3 years and the group of under 10 months of HA non user before second CI (P=0.01* on SDS and P=0.04* on WRS). We recommend wearing hearing aids on the opposite side after first CI. As the Japanese language uses lower frequencies, a little wearing threshold of usable frequencies remains. Also, the input from the hearing aid is very important. It is a waste to remove the HA and let the input on the opposite side of the first CI.

Most of the speech understanding scores (WRS and SDS) for children who have undergone at least 1 year habilitation after first CI and now have been fitted with a second CI show similar results to the first CI. Though the second CI eventually caught up with the first CI, it took nearly over one year.

Binaural hearing using bilateral CI is better than the first CI in all speech understanding tests. Especially, there were significant differences between the results of the first CI and bilateral CI on: 1) SDS at 70 dB SPL (P=0.02*); 2) SDS at 1 m from second CI side SP at 60 dB SPL (P=0.02*); 3) WRS at 1 m from second CI side SP at 60 dB SPL (P=0.02*); 4) SDS at 1 m from front SP under the noise (S/N=80/70, +10) (P=0.01*); 5) WRS at 1 m from front SP under the noise (S/N=80/70, +10; P=0.002**).

These results may show important binaural effectiveness such as binaural summation (1, 4, 5) and head shadow effect (2, 3).

Binaural summation (1, 3, 4, 7) and head shadow effect (2-5) are very likely to be important phenomena providing effective binaural advantages. Furthermore, binaural squelch (2, 4, 5) and sound localization (8, 9) are also well known to yield binaural advantages. In particular, in infancy there are many cases where the ability of hearing under the noise is very important for speech/language development.

The improvement of sound localization and hearing under noise that is provided in binaural hearing shows strong effectiveness in a typical infant environment and for children in a classroom setting (14). Bilateral CI is a very useful medical intervention for children with severe-to-profound hearing loss in Japan and elsewhere.

CONFLICT OF INTEREST

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

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Original Article

The Usefulness of Reconstructed 3D Images in Surgical Planning for Cochlear Implantation in a Malformed Ear with an Abnormal Course of the Facial Nerve

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Objectives. It is not unusual for a cochlear implantation (CI) candidate to have some type of ear malformation, in particular an abnormal course of the facial nerve (FN). In this study, we attempted to reconstruct a three-dimensional (3D) image of temporal bone structures with malformation using computed tomography (CT) imaging and examined its usefulness in the surgical planning of CI in a malformed ear.

Methods. We prepared 3D images for 6 separate CI cases before surgery. First, we manually colored preoperative CT images using Photoshop CS Extended. We then converted the colored CT images to 3D images using Delta Viewer, freeware for Macintosh. Before surgery, we discussed any problems anticipated based on the 3D images and plans for surgery with those who would be performing the CI.

Results. Case 1: The subject was a 3-year-old boy with malformed ossicles, semicircular canal (SC) hypoplasia, internal auditory canal stenosis, and an abnormal course of the FN. 3D image indicated that the stapes were absent, and the FN was more anteriorly displaced, so that it was difficult to perform cochleostomy. The surgical findings were similar to those depicted on the 3D image, so we could insert an electrode based on the preoperative image simulation without complications. Case 2: The subject was a 7-year-old boy with malformed stapes, atresia of the round window, cochlear and SC aplasia, and an abnormal course of the FN with bifurcation. CI was performed with no problems, in the same manner as in Case 1.

Conclusion. We were able to successfully depict the structures of the inner ear, ossicles, and FN as 3D images, which are very easy to understand visually and intuitively. These 3D images of the malformed ear are useful in preoperative image simulation and in surgical planning for those performing a CI procedure.

Key Words. *Three-dimensional image, Temporal bone, Facial nerve anomaly, Cochlear implantation*

INTRODUCTION

A number of cochlear implantation (CI) candidates have ear malformations, in particular an abnormal course of the facial nerve (FN). The course of the FN is very important for CI surgery; the more abnormal the course of the FN, the more difficult it is to perform CI surgery. However, it is extremely difficult for surgeons to understand the three-dimensional (3D) course of the FN prior to surgery through the use of two-dimensional (2D) computed tomography (CT) images.

In this study, we attempted to reconstruct a 3D image of temporal bone structures using CT imaging for a plan of CI surgery in a malformed ear and show the 3D images of the two cases.

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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

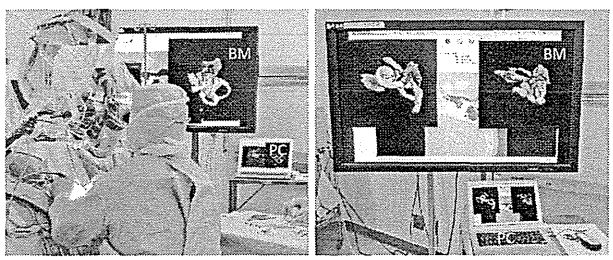

Fig. 2. The pictures of an operation room during a cochlear implantation surgery. The surgeon sees Delta Viewer 3 dimensional images displayed on the bedside monitor (BM), which is controlled by the notebook PC (Macintosh), and compares the images with surgical findings.

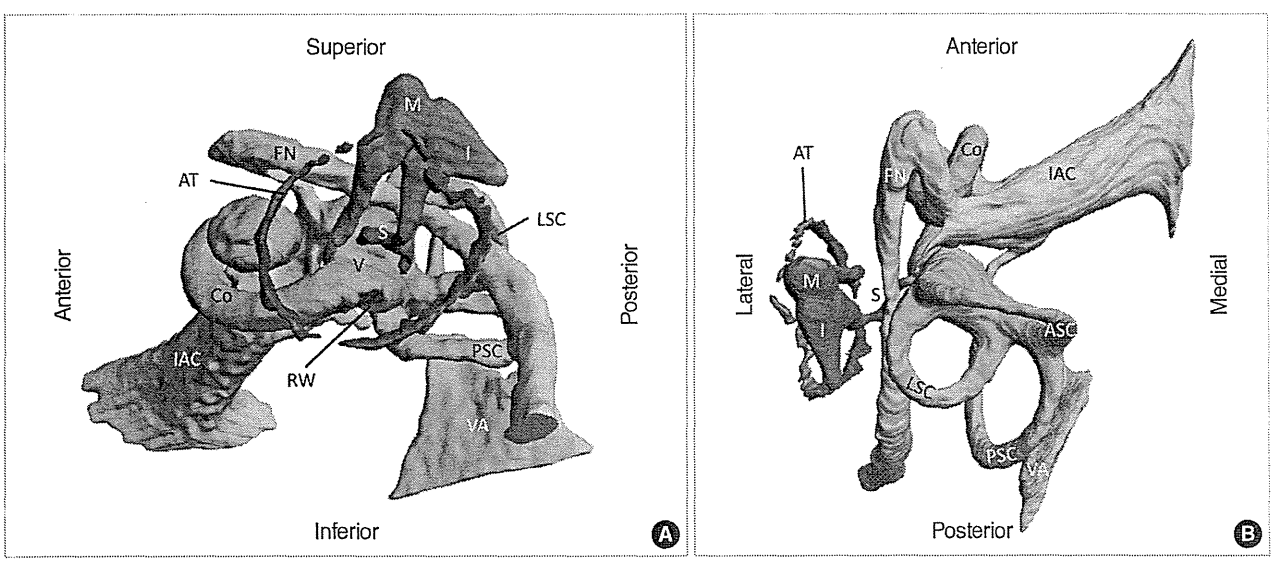

Fig. 1. Example of Delta Viewer 3 dimensional image of normal temporal bone structures of left ear based on computed tomography image. Image (A) is antero-lateral inferior view, and image (B) is superior view. The bony labyrinth was shaded in blue and includes the cochlea (Co), vestibule (V), anterior 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.

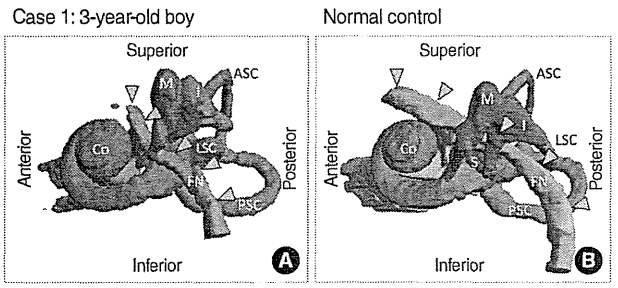

Fig. 3. (A) Lateral view of the temporal bone structures of case 1. The lateral semicircular canal (LSC) is hypoplastic, and the crus of the incus (I) and stapes (S) are absent. The labyrinthine segment of the facial nerve (FN) and the geniculate ganglion are posteriorly displaced, and the tympanic and 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.

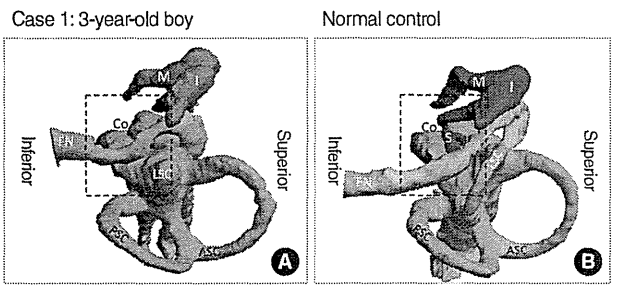

Fig. 4. (A) The Delta Viewer 3 dimensional (DV-3D) image of case 1 in the same position during cochlear implantation surgery. (B) The DV-3D image of normal control in the same position as (A). Rectangle of dashed line shows the area of Fig. 5. Co, cochlea; ASC, anterior semicircular canal; PSC, posterior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

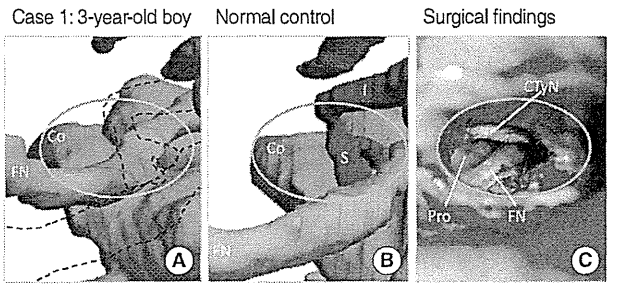

Fig. 5. The images of the surgical field of posterior tympanotomy as shown by the orange oval. (A) Case 1: close-up image of the rectangle of Fig. 4A. The stapes (S) and crus of the incus (I) are absent, and the facial nerve (FN) is antero-inferiorly displaced. Dashed line shows the position of the ossicles 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.

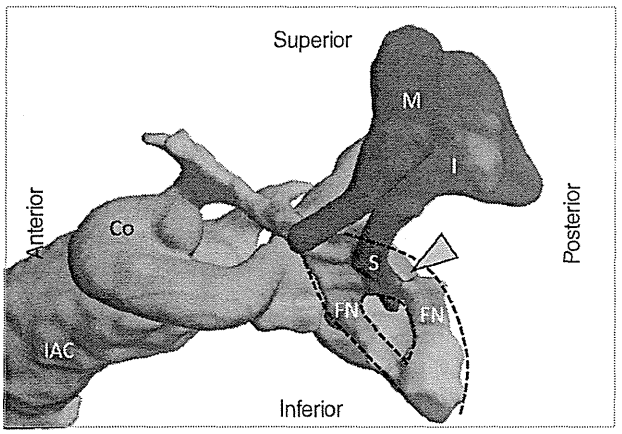

Fig. 7. Antero-lateral view of the temporal bone structures of case 2. The shape of the cochlea (Co) and semicircular canals are hypoplastic. The labyrinthine segment of the facial nerve (FN) and geniculate ganglion are posteriorly displaced, the tympanic and mastoid segments of the FN are antero-inferiorly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

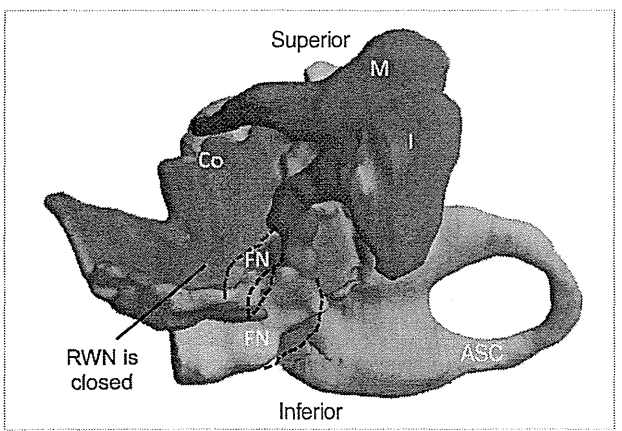

Fig. 8. The Delta Viewer 3 dimensional image of case 2 in the same position during cochlear implantation surgery. The surface of the tympanic cavity is displayed in green. The round window niche (RWN) is closed, so there was no landmark for cochlear fenestration. Co, cochlea; FN, facial nerve; ASC, anterior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

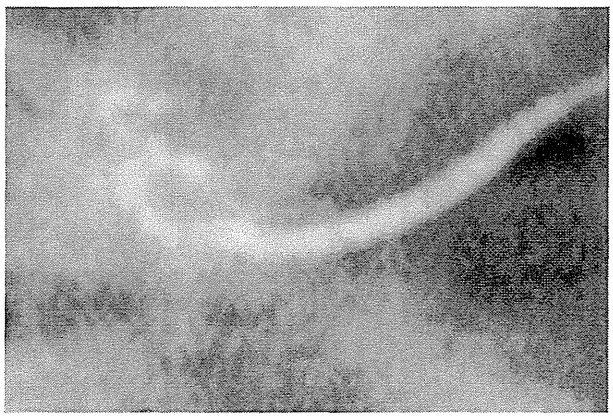

Fig. 9. The X-ray of inserted electrode of case 2.

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.

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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

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Tomomi Yamamoto-Fukuda, MD[2] · Haruo Yoshida, MD[2] · Akiko Ito[1] · Chiharu Tanaka[1] · Kyoko Baba[1]
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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*

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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-

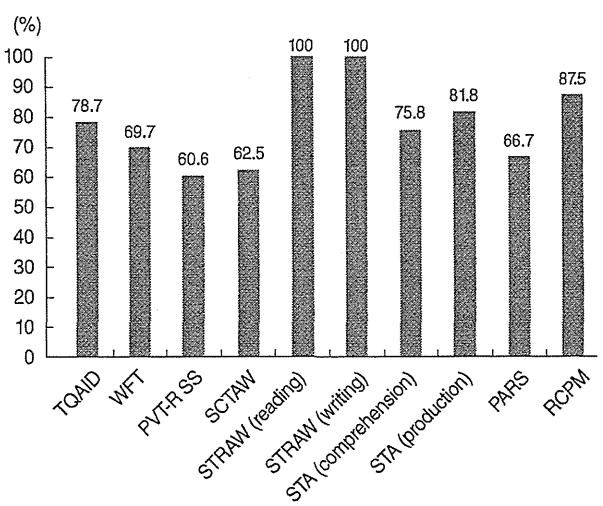

Fig. 1. The results of the various language development tests. The results show that children suffering from hearing loss exceeded the national average of all children with hearing difficulties by at least 60.6% and up to 100%. TQAID, test for question and answer interaction development; WFT, word fluency 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.

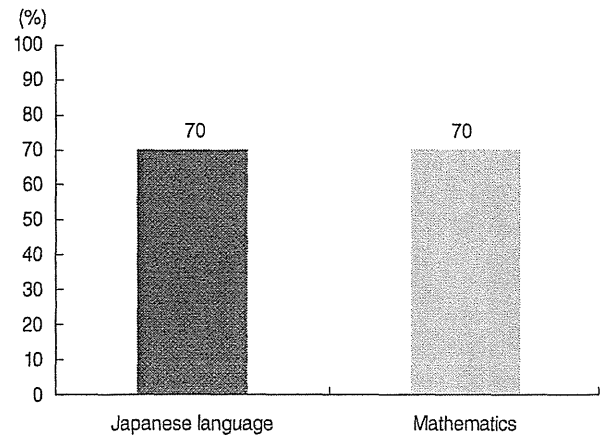

Fig. 2. On the criterion-referenced testing for Japanese language and mathematics, 70.0% of all scores exceeded the national average of scores obtained by normal-hearing children.

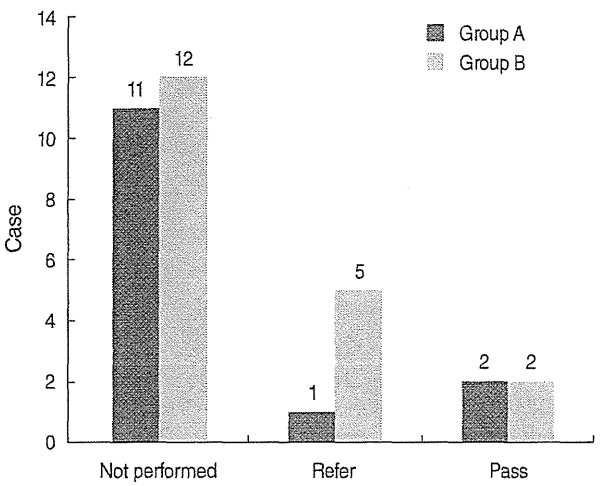

Fig. 3. Whether or not the child went through newborn hearing screening.

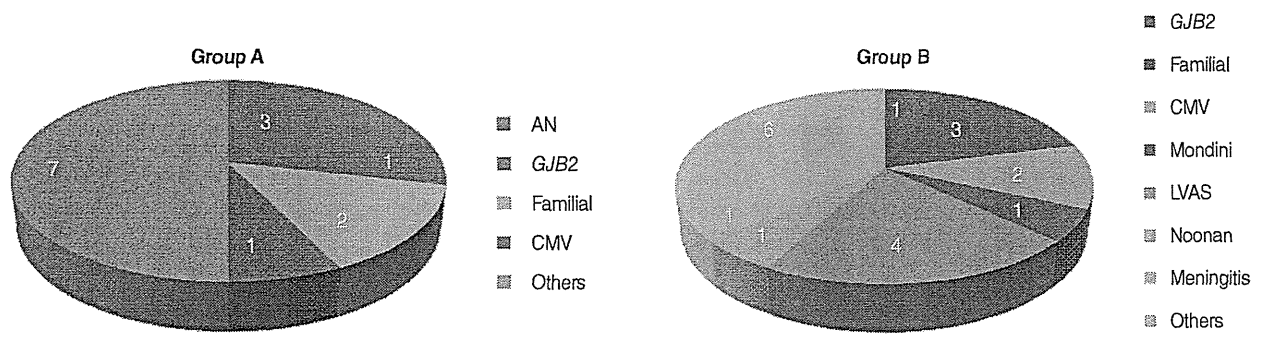

Fig. 4. Causes of deafness. AN, auditory neuropathy; *GJB2*, Gap junction protein, beta-2, 26kDa (*GJB2*) gene mutation; CMV, congenital cytomegalovirus infection; LVAS, large vestibular aqueduct syndrome.

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.

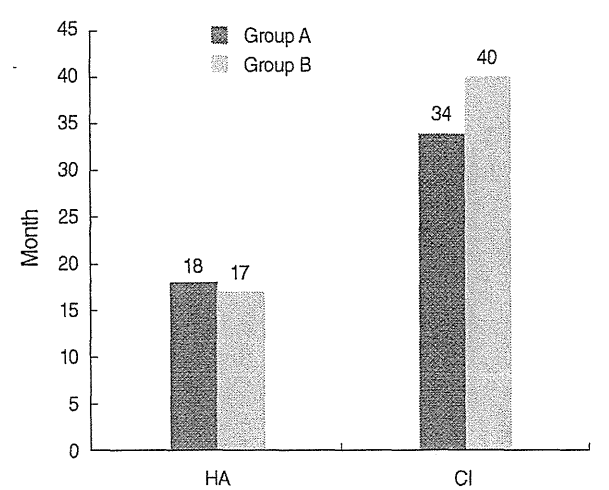

Fig. 5. The mean age for children to start wearing a hearing aid (HA) and cochlear implant (CI).

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

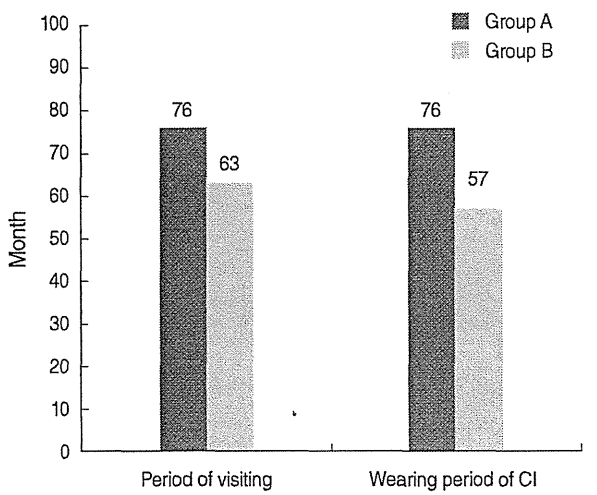

Fig. 6. The mean period of the visit at our hearing center and the mean wearing period of cochlear implant (CI).

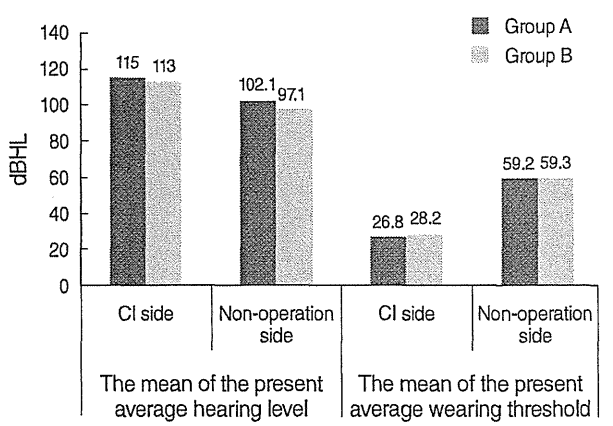

Fig. 7. The mean of the current average hearing level and the present average wearing threshold.

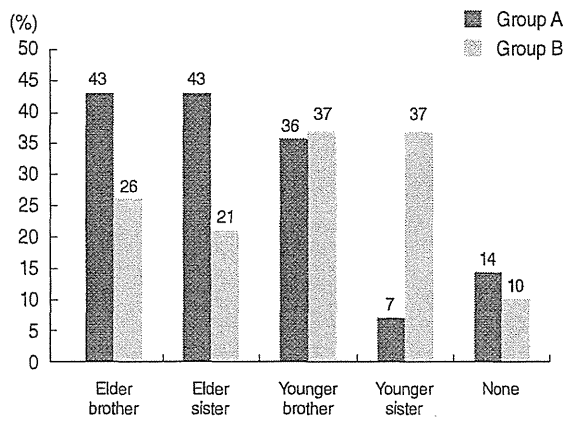

Fig. 8. Whether the child has any siblings.

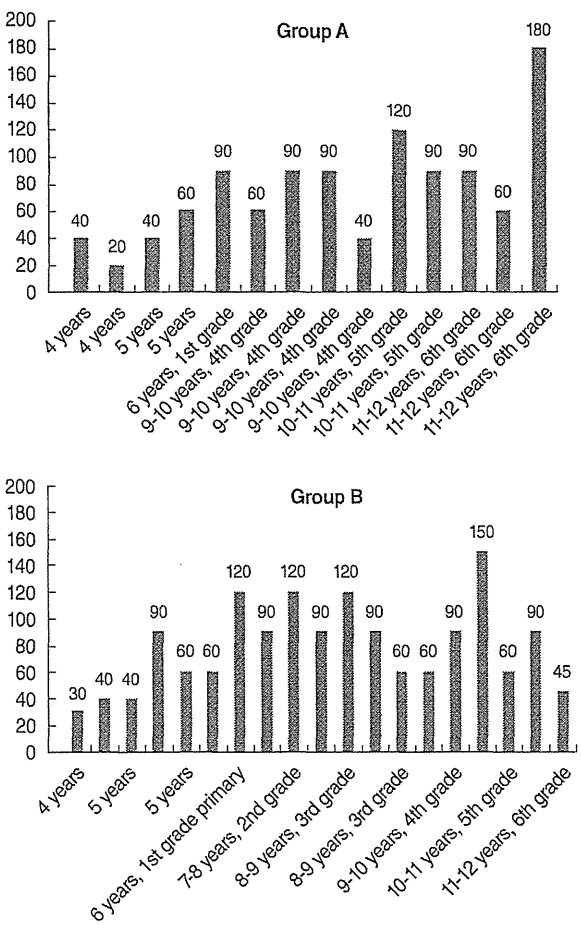

Fig. 9. The amount of time spent studying at home on a daily basis.

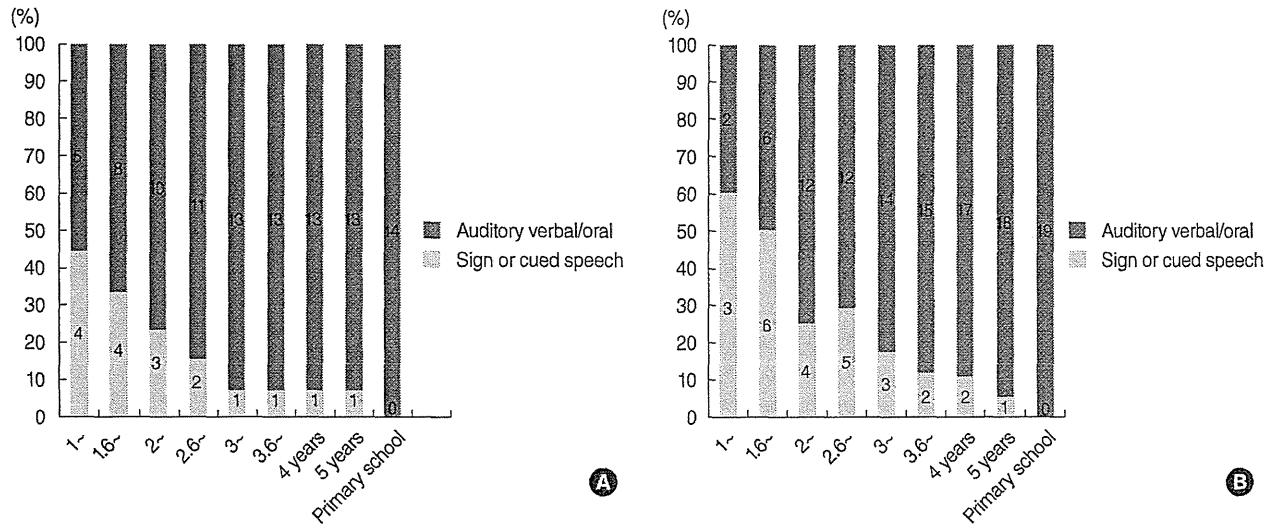

Fig. 10. Educational method (school). A, group A; B, group B.

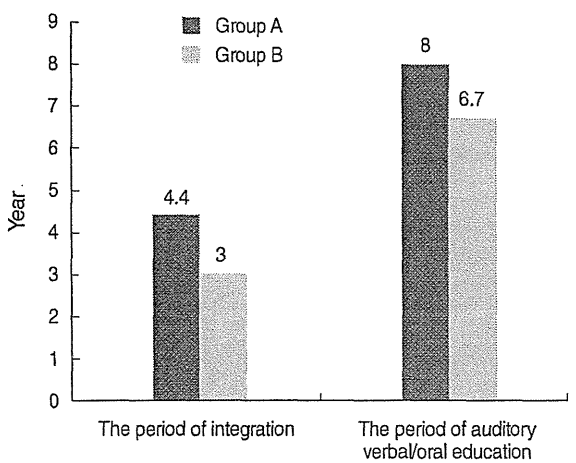

Fig. 11. The period of integration and the period of auditory verbal/oral education.

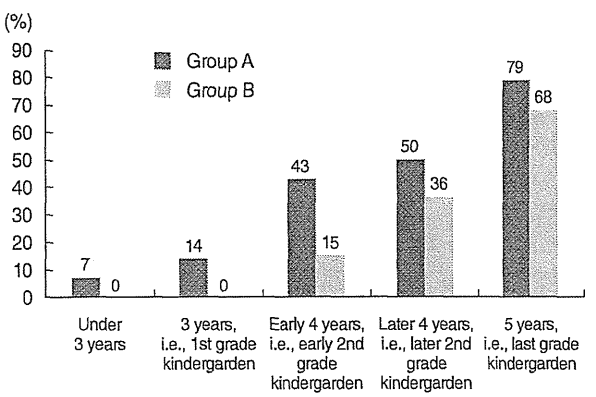

Fig. 12. Educational institution child attended before entrance to primary school.

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.

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A postmeningitic cochlear implant patient who was postoperatively diagnosed as having X-linked agammaglobulinemia

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ABSTRACT

X-linked agammaglobulinemia (XLA) is caused by a mutation in the Bruton tyrosine kinase, leading to an arrest in B cell development. Consequently, patients with XLA show significant decreases in gammaglobulin. Here, we describe a child with postmeningitic deafness and XLA who underwent a cochlear implantation. His psychomotor development had been normal and his congenital immunodeficiency was noticed only postoperatively. Immunoglobulin replacement treatment was started, but he still suffered repeated infections. Eventually, his cochlear implant was removed. A preoperative check of immunological status might be advisable in postmeningitic patients undergoing cochlear implantation to reduce the risk of postoperative infectious complications.

1. Introduction

Cochlear implantation is generally accepted as a safe and effective treatment to rehabilitate patients with bilateral severe-to-profound hearing loss. The complication rate is relatively low. The most common non-device-related complications occur around the skin flap, and the incidence of infectious complications has been reported as 12.9% [1].

Primary immunodeficiency affects as many as 10 million people worldwide. More than 150 primary immunodeficiency diseases have been identified, and they range widely in severity. Immunodeficient conditions likely increase the risk of postoperative infectious complications even in cochlear implant patients.

To the best of our knowledge, only five patients with cochlear implants who had primary immunodeficiencies have been reported to date. Hopfenspirger et al. [2] described two patients with primary immunodeficiency: one with neutropenia/chemotactic neutrophil dysfunction and one with IgA deficiency. Although they had been diagnosed with primary immunodeficiencies preoperatively, they suffered postoperative infections at the surgical sites. Eventually, the cochlear implants in both patients were removed. Yu et al. [3] reported two patients with immunodeficiency who also suffered postoperative infections after cochlear implant surgeries and were then diagnosed with primary immunodeficiencies based on results of their IgG isohemagglutinin titers decreasing postoperatively. They reported that the cochlear implant in one of the two patients was preserved with daily low-dose oral antibiotic administration, without intravenous immunoglobulin therapy [3]. More recently, Brookes et al. [4] reported on a patient who was diagnosed with deafness-dystonia-optic neuronopathy (DDON) syndrome and X-linked agammaglobulinemia (XLA). His XLA was preoperatively diagnosed and antibody replacement therapy was initiated. He did not appear to have any wound troubles after cochlear implantation.

Here, we report on the clinical course of a child with a cochlear implant who was diagnosed with XLA postoperatively. To the best of our knowledge, this is the second report of a patient with XLA who underwent cochlear implantation.

2. Case report

A 3-year-old boy with a postmeningitic profound sensorineural hearing loss (SNHL) was referred to our department in May 2006, to obtain an evaluation for cochlear implantation. A physical examination at the first visit to our department revealed that he had a partial severe retraction of tympanic membrane pars tensa in the right ear and accumulated effusions in the left ear, with purulent rhinorrhea in his nasal cavities. Play audiometry and an auditory brainstem response examination revealed bilateral profound SNHL. A CT scan and MRI examinations revealed

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ossification of the basal turn of the scala tympani of the bilateral cochleae.

In June 2006, we performed cochlear implantation surgery on the left ear. Although ossification was observed in the basal turn of the scala tympani, all electrodes (CI24RE (CA); Cochlear, Ltd., Cove, NSW, Australia) were inserted into the scala tympani. He received ceftriaxone for 10 days postoperatively. His postoperative clinical course and postoperative speech recognition ability were good.

However, in December 2007, he complained of pain where the device had been implanted, and his mother noticed swelling at the site. The swelling and his symptoms became worse (Fig. 1), although he was receiving an oral antibiotic. We then performed surgical drainage, revealing that the content of the swelling was a purulent secretion. Drain tubes were placed during the surgery. Bacteriological examination of the purulent secretion revealed *Streptococcus pneumoniae* (PISP). The local condition improved with daily postoperative lavage through the drain tubes and antibiotic administration.

In April and July 2008, the localized swelling and pain re-appeared again twice. We performed surgical drainage. The content of the swelling revealed a blood clot, and bacteriological examinations of the contents were negative. The local condition again improved with the same treatment as before. During his hospital stay, he underwent patch tests against the materials used in the cochlear implant, provided by the implant company; all the tests were negative. In June 2008, he was treated at a hospital for pneumonia. In September 2008, he underwent an immunological evaluation. His peripheral blood B cell subset was less than 1%, and his serum IgG, IgA, and IgM were 7 mg/dL, 3 mg/dL, and 30 mg/dL, respectively. Gene analysis revealed a mutation in the Bruton tyrosine kinase gene.

He was finally diagnosed with XLA. He started to undergo antibody replacement therapy every 3 weeks: his physical activity level improved significantly; and, his repeated purulent nasal discharges stopped spontaneously without usage of antibiotics,

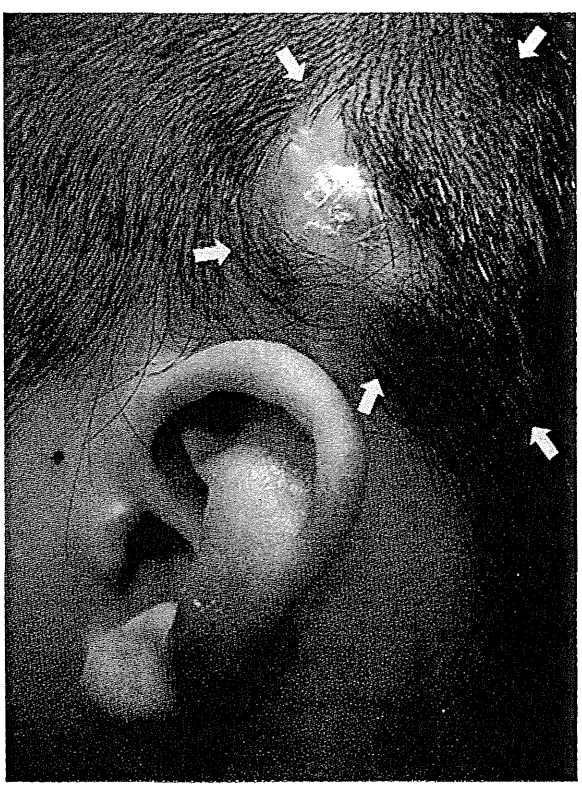

Fig. 1. Swelling at the site where the cochlear implant was implanted.

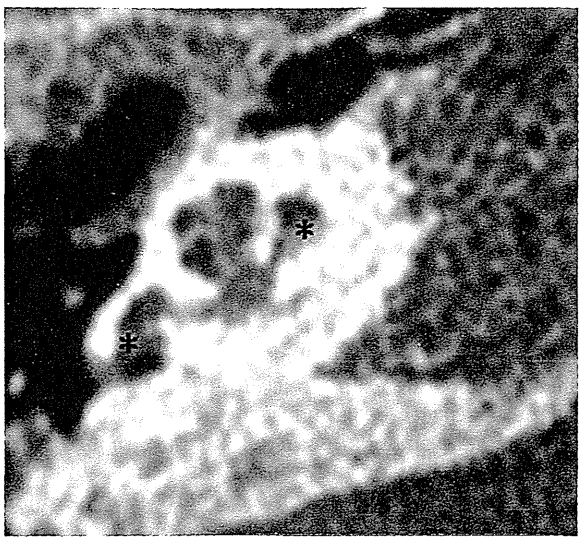

Fig. 2. A CT scan image of right ear at the second cochlear implantation. The asterisks indicate ossification of the scala tympani in the basal turn of the cochlea.

after this treatment. Despite the antibody replacement therapy, subsequently, local swelling around the implant recurred twice more. On the second occurrence, we removed granulation tissue surrounding the cochlear implant in addition to surgical drainage under general anesthesia. During the surgery, we held electrode bundle near the implant, turned it over, removed all granulation tissue surrounding the cochlear implant, to the extent possible, and washed the wound gently with saline many times, after which the cochlear implant was covered with the temporal muscle. Nevertheless, he experienced local swelling and pain again, and self-destruction of wound skin was observed. Finally, we decided to remove the cochlear implant. In February 2010, we performed cochlear implant surgery on the right ear (PULSAR; Med-El, Innsbruck, Austria) and subsequently removed the cochlear implant from the left ear. We inserted the electrode into the scala vestibule because a preoperative CT scan study revealed ossification of the scala tympani in the right ear (Fig. 2). We successfully inserted all except one electrode. Postoperatively, he received flomoxef and clindamycin for 12 days and his serum IgG level was maintained at 1000–1700 mg/dL by intravenous administration of γ-globulin perioperatively. His postoperative clinical course was good. For 22 months, no recurrence of local swelling occurred since the surgery. Postoperative speech recognition ability testing revealed 56% (words) by the Japanese speech recognition test battery, "CI–2004" under the auditory-only listening condition.

3. Discussion

XLA is caused by mutations in the Bruton tyrosine kinase, located at chromosome Xq22, leading to arrested B cell development at a pre-B cell stage. As a result, patients have no mature B cells, and IgG is less that 200 mg/dL. XLA accounted for 7.3% of primary immunodeficiency conditions in a 2007 survey in the United States. The immunodeficient condition allows bacterial meningitis, although most patients who present with bacterial meningitis do not have XLA or any other obvious deficiency in immune function [5]. Additionally, only 7.1% of child patients experience bilateral profound hearing (>90 dBSPL) after childhood bacterial meningitis [6]. Given these numbers, very rarely will a patient with postmeningitis who is a candidate for cochlear implant surgery have an immunodeficiency condition.

The most reliable strategy for the management of postoperative wound infections after cochlear implantation may be to explant

the device. Tambyraja et al. [1] summarized data from the Manufacturer User Facility and Distributor Experience (MAUDE) database, which is maintained by the Food & Drug Administration and has mandatory reporting requirements. In the pre-2002 period, 102 cases of flap problems were reported, which included flap necrosis, flap infection, flap dehiscence, and device extrusion. Approximately 70% of cochlear implants were explanted in those patients. However, current recommendations call for conservative measures [7]. Yu et al. [3] reported four patients with postoperative infections including two primary immunodeficiency patients, who showed decreased IgG isohemagglutinin titers. The postoperative cochlear implant infections in three of the four were controlled effectively with limited surgical approaches and prolonged postoperative antibiotic administration [3]. The cochlear implant was explanted due to failure of infection control in one patient with a primary immunodeficiency although antibody replacement therapy and intravenous antibiotics administration were continued [3]. Considering these reports, prolonged medical management may be effective. However, in the patient with primary immunodeficiency who did not need device removal, the report did not provide detailed levels of immunoglobulin, and her infection was successfully controlled using only a daily low-dose oral antibiotic (cephalexin) with no antibody replacement therapy; her immunodeficiency thus may not have been severe. Taken together, the conservative approach probably has limited efficacy in patients who are immunodeficient.

Controlling infectious complications after cochlear implant surgery can be difficult even in healthy patients. Recently, biofilm formation on the surface of a cochlear implant receiver–stimulator device was suggested to probably contribute to persistent cochlear implant infection. Such a bacterial biofilm on the device is highly resistant to antibiotic therapy and to removal by direct washing of the device [8]. Moreover, Pawlowski et al. [8] reported that the biofilm on the device was most substantial in the depressions along the surface of the device that were created by the manufacturer. Considering these points, the first infection on our patient was likely due to his untreated immunodeficiency condition, and biofilm on the surface of the device probably contributed to the persistent local infection.

If the patient's immunodeficiency is diagnosed preoperatively, one can add supplemental treatments perioperatively to help avoid postoperative complications. Brookes et al. [4] reported on a patient who was diagnosed with DDON syndrome and XLA. His XLA was preoperatively diagnosed and antibody replacement therapy was initiated. He did not appear to have any wound troubles after cochlear implantation. However, if the patient has not been diagnosed preoperatively, like our patient, the immunodeficient condition probably tends to increase infectious complications, ultimately leading to extraction of the implant. Also, in patients who are postmeningitic, cochlear implant surgery on the opposite side probably becomes more formidable because of postmeningitic ossification in the cochlea. Thus, considering the presence of an immunodeficient condition in patients is important, especially from symptoms suggesting such conditions such as recurrent infections. In most patients with XLA, they typically present with recurrent pyogenic infections starting at 5–6 months of age when passively placentally transferred maternal antibodies have waned [9]. Our patient had repeated purulent rhinorrhea preoperatively, which continued postoperatively. He also suffered from pneumonia once postoperatively. With hindsight, these were likely signs of his immunodeficient condition. Physicians should pay attention to the possibility of an immunodeficient condition in a patient before and after surgery. To reduce the risk of complications after a cochlear implant, we think that in addition to obtaining a careful history about any repeated infections, preoperative checks of serum immunoglobulins (IgG, IgA, IgM) and IgG subclass analyses might be needed for the diagnosis of major primary immunodeficient deficits on cochlear implant candidates who have had repeated infectious episodes. Moreover, the prevalence of acquired immune-deficiency syndrome (AIDS) continues to increase, so a preoperative check of the CD4/CD8 ratio might also be useful.

4. Conclusion

An immunodeficient condition may allow bacterial meningitis. However, most patients who present with bacterial meningitis do not have an immunodeficiency [5] and less than 10% of children with meningitis experience bilateral profound hearing after bacterial meningitis [6]. Thus, the chance of seeing a patient with XLA as a candidate for cochlear implant surgery is very low. However, we should consider an immunodeficient condition as a possible cause of meningitis in patients who are candidates for cochlear implants in helping to avoid postoperative infectious complications.

Conflicts of interest

None.

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Clinical and Experimental Dermatology

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

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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.

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Conflict of interest: none declared.

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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

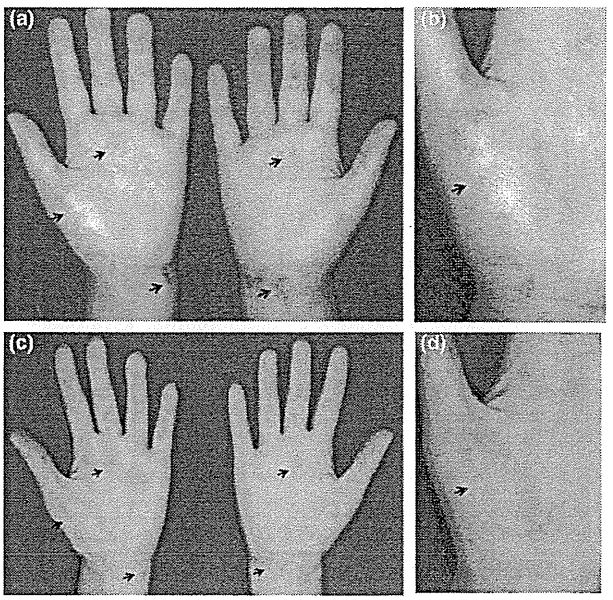

Figure 1 (a,b) Patient at the age of 7 years, showing hyperkeratotic lesions on the palms, wrists and flexor sides of the digits with underlying erythema, fissures and peeling. (c,d) At the age of 15 years, although the hyperkeratotic lesions on the palms showed some improvement, the skin lesions on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

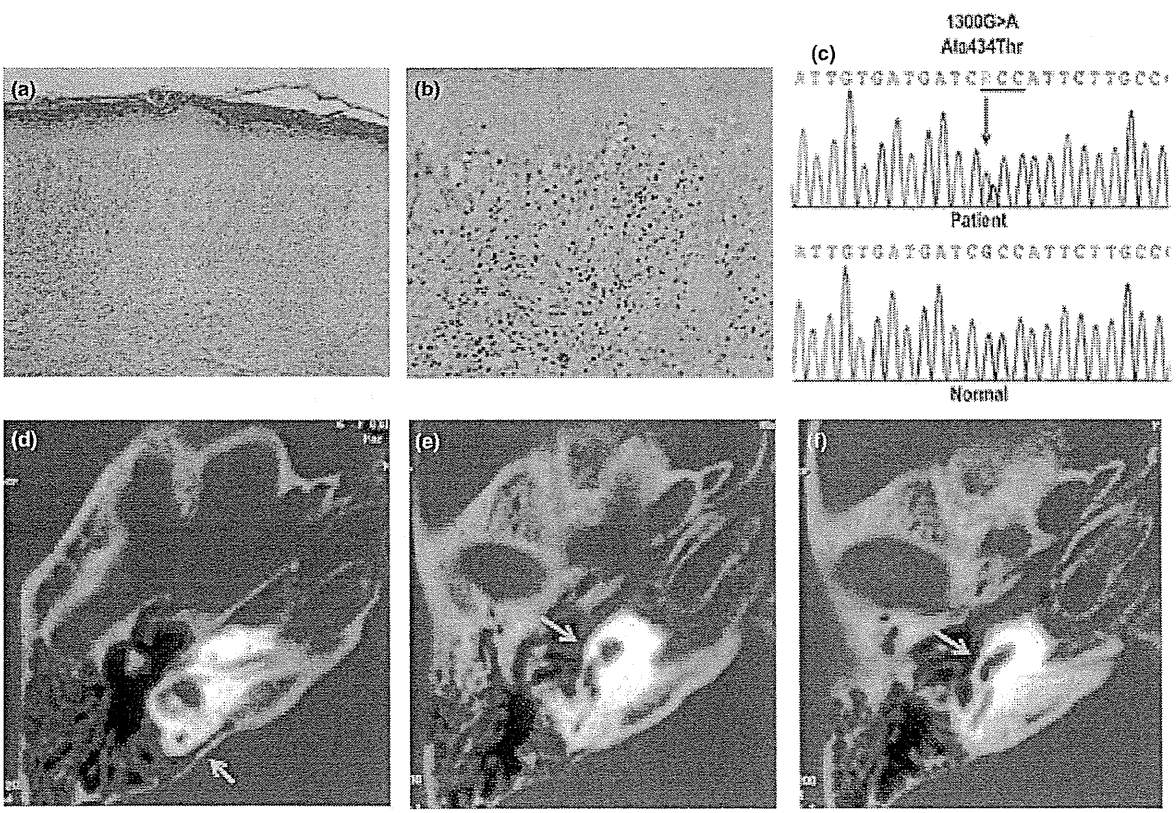

Figure 2 (a) The biopsy taken from the skin lesion on the patient's right wrist revealed hyperkeratosis, epidermal hyperplasia, and a band-like lymphocytic infiltration in the upper dermis (hematoxylin and eosin, original magnification ×40). (b) Liquefaction degeneration and Civatte bodies were detected (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).

