その他

- 当然ではあるが，爆発や突発事故的な大音響に曝露した場合には音響外傷の可能性が高く，航空機搭乗やスキューバダイビングなどの急激な気圧変化に曝露した病歴があれば圧外傷（barotrauma），さらにめまいを伴えば内耳障害の可能性が高い．
- また，アミノグリコシド系抗菌薬や白金製剤の抗癌剤投与後には薬物性難聴が考えられる．

純音聴力検査

- いうまでもなく最も基本の重要な検査である．
- 難聴の程度とともにまずみるのは気骨導差である．
- 急性の伝音難聴，感音難聴にそれぞれどのような疾患があるかはここでは割愛するが，急性の混合難聴をみることはまれで，その場合には上半規管裂隙症候群や前庭水管拡大症候群での急性内耳障害なども否定できない．
- 以下に示す聴力型（パターン）も診断に有用なことが多い．

低音型

- 伝音難聴で低音域の低下がみられれば滲出性中耳炎などの中耳貯留液が最も疑われ，感音難聴なら急性低音障害型感音難聴が疑われる．

高音漸傾型

- 伝音難聴でこのタイプは少ないが，感音難聴では突発性難聴などでよくみられる．

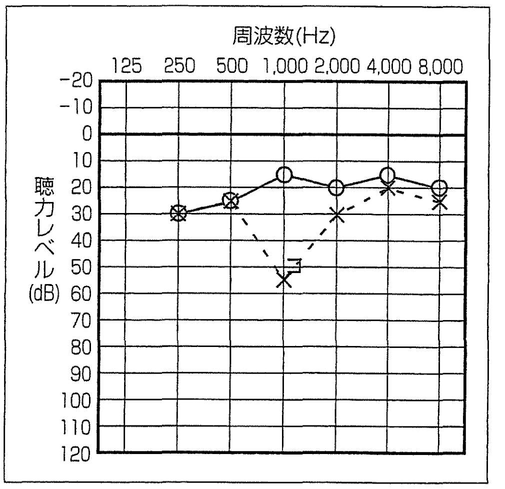
★1
As 型は外耳道容積がおよそ 0.3 mL 以下，Ad 型は外耳道容積がおよそ 1.4 mL 以上．

水平型

- 伝音難聴なら耳小骨離断でよくみられ，感音難聴ではやはり突発性難聴でみられる．

その他

- 急性難聴にはあらゆる聴力型がありうるが，時に聴神経腫瘍の初期に耳閉感のみが主訴で，周波数はさまざまであるが dip 型の難聴を示すことがあり注意を要する（❷）．

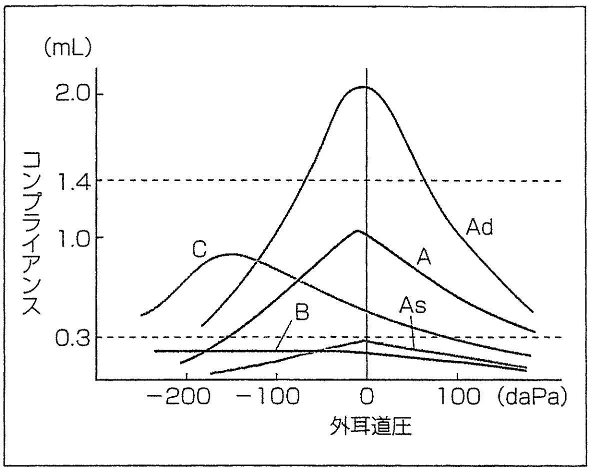


❷左聴神経腫瘍（47 歳，男性）の初診時聴力像
主訴は左耳閉感のみで症状に変動がみられたため，MRI 検査を行ったところ，左内耳道底に約 3 mm の腫瘍陰影を認めた．

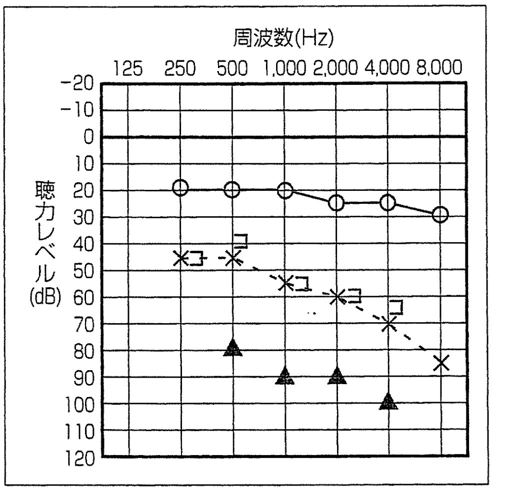
ティンパノグラム（❸）

A 型★1

- 伝音連鎖の正常パターンであるが，ピークの低い As 型が中耳伝音連鎖の固着，ピークの高い Ad 型が離断とされている[1]．
- しかし，その診断的価値（感受性）は高くなく，次のアブミ骨筋反射と組み合わせて初めて診断的価値が高まる．



❸ティンパノグラムの型分類



❹左突発性難聴（56歳，男性）の聴力像と左耳の同側音刺激によるアブミ骨筋反射閾値（▲）

左耳の最小可聴閾値は45〜85 dBで中等度難聴を示すが，アブミ骨筋反射閾値は正常耳と同等の80〜100 dBであり，補充現象が陽性である．

B型

- 急性難聴で中耳貯留液や滲出性中耳炎は少ないが皆無ではなく，あわてて受診するケースもある．
- さらに鼓膜が肥厚していると貯留液が透見できず，突発性難聴と誤診する場合がある．

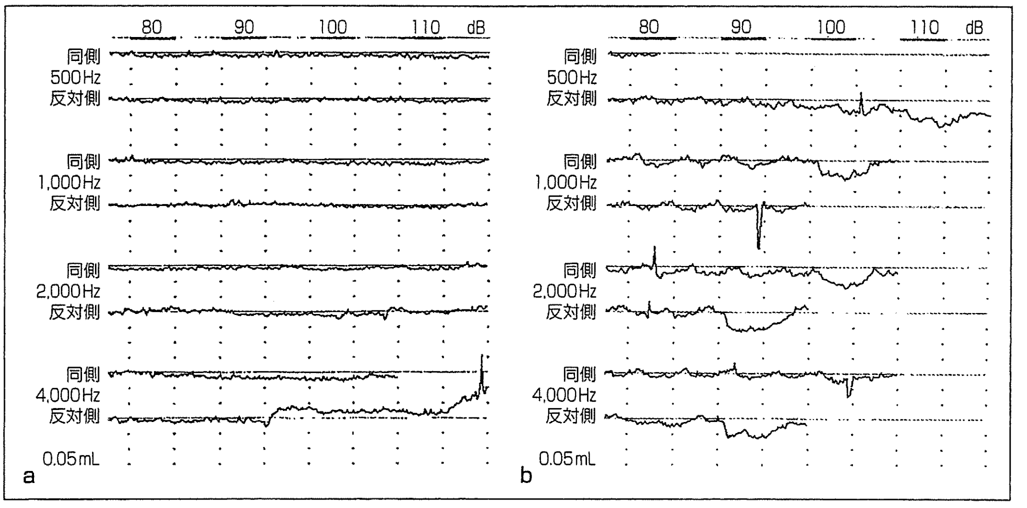
C型

- 中耳陰圧を示すが，C_2型★2では中耳貯留液がある可能性は低くはなく[2]，急性難聴の原因にもなりうる．

★2 C_2型
−200 daPaより高度の陰圧．

アブミ骨筋反射（SR）検査

- アブミ骨筋反射（stapedial reflex：SR）検査は，一般には反射の有無による中耳伝音連鎖の異常が診断できることが成書にも記されているが，感音難聴の場合に反射閾値も非常に重要であることを強調したい．
- 感音難聴には内耳性と後迷路性があるが，内耳性では補充現象（recruitment phenomenon）があるため，反射閾値は正常聴力耳と大差なく，80〜100 dBの音刺激で発生する（メッツテスト〈Metz test〉，❹）[3]．一方，後迷路性では補充現象がないので最小可聴閾値からSRが生じる閾値との差は80〜100 dBあり，軽度難聴以外では110 dBまでの音刺激では反射は生じないことが多い．
- 補充現象の検査として，SISI（short increment sensitivity index）検査，Jerger自記オージオメトリーなどがあるが，いずれも自覚的検査であり確実性を欠くが，SRは他覚的検査であるため，被検者の主観に左右されないという大きな利点がある．



❺ティンパノグラムC型（中耳陰圧）でのアブミ骨筋反射

外耳道圧を平圧にしてアブミ骨筋反射を測定すると検出されないが（a），外耳道圧を中耳圧と同じ圧（−150 daPa）にして測定すると正常に反射が検出される（b）.

ポイント

感音難聴でアブミ骨筋反射が正常耳と同等の閾値で検出されれば，ほぼ内耳性感音難聴と診断できる.

さらに鑑別が必要な場合に加える検査

歪成分耳音響放射検査（DPOAE）

- 歪成分耳音響放射検査（distortion product otoacoustic emission：DPOAE）は，内耳外有毛細胞の機能を反映する検査で，これが低下していれば内耳

Advice　SR検査のコツ

①ティンパノグラムC型のとき

⇒ 中耳圧と同じ外耳道圧にして測定

SR検査を行う際に，中耳貯留液があってティンパノグラムがB型を示すときには反応が検出されないことはよく知られているが，C型やAd型の場合も検出できないことがある.

通常SRは外耳道内が平圧の状態で測定するが，C型では中耳が陰圧なので鼓膜の可動性が悪く，SRによる微妙なコンプライアンスの低下が波形に反映されないことがある．最近は自動的に外耳道圧を中耳圧と同じ圧に設定してSR検査を行う機器も増えているが，そうでない場合は手動で外耳道圧を設定できる機器では外耳道圧を中耳圧に近づけるように設定するとSRを検出できることがある（❺）.

②ティンパノグラムAd型のとき

⇒ 中耳圧と少しずらせた外耳道圧で測定

またAd型の場合は逆で，ピーク圧付近でコンプライアンスが急激に変化することを示しており，SR測定中に脈拍や呼吸などのわずかな中耳圧変化で基線が変動するためにうまく検出できないことがある．その場合には，やはり可能なら手動で外耳道圧を中耳圧のピークから少しずらせて設定すれば，基線が安定してSRを検出できることがある.

性難聴の可能性が高い.

- 詳細は他書に委ねるが, auditory neuropathy では DPOAE が正常で, 次に述べる ABR が検出できない.
- 本検査は耳垢や中耳貯留液などの外耳, 中耳の状態に結果が左右されることが欠点である.

聴性脳幹反応 (ABR)

- 聴性脳幹反応 (auditory brainstem response : ABR) は, 4 kHz 付近の高さのクリックの音刺激に対する脳波を検出することで聴力を評価する検査で, これも客観的検査である.
- 後迷路性難聴では, 反応の欠如, 病巣より中枢側の波形が欠如, I-V 波の間隔の延長, などの所見がみられ, そのほか, 自覚的検査で難聴がみられても ABR の反応閾値が正常であれば機能性難聴(心因性難聴, 詐聴)の診断に有用である.

聴性定常反応 (ASSR)

- 聴性定常反応 (auditory steady-state response : ASSR) は, ABR と同様の聴性誘発反応による聴力検査である.
- ABR での I〜VII 波のように反応波形は出ないので, 難聴の部位診断など神経学的診断には適さない.
- しかし音の周波数ごとに反応閾値が検出できるのが最大の利点で, 幼小児などの他覚的聴力検査に応用される.

実際の検査・診断手順

- 上記の検査を用いて実際に「鼓膜に明らかな異常がない急性難聴」の患者さんの検査・診断手順は❻のようになる.

①まずは純音聴力検査を行い, 難聴の種類(伝音, 感音, 混合)を確かめる.

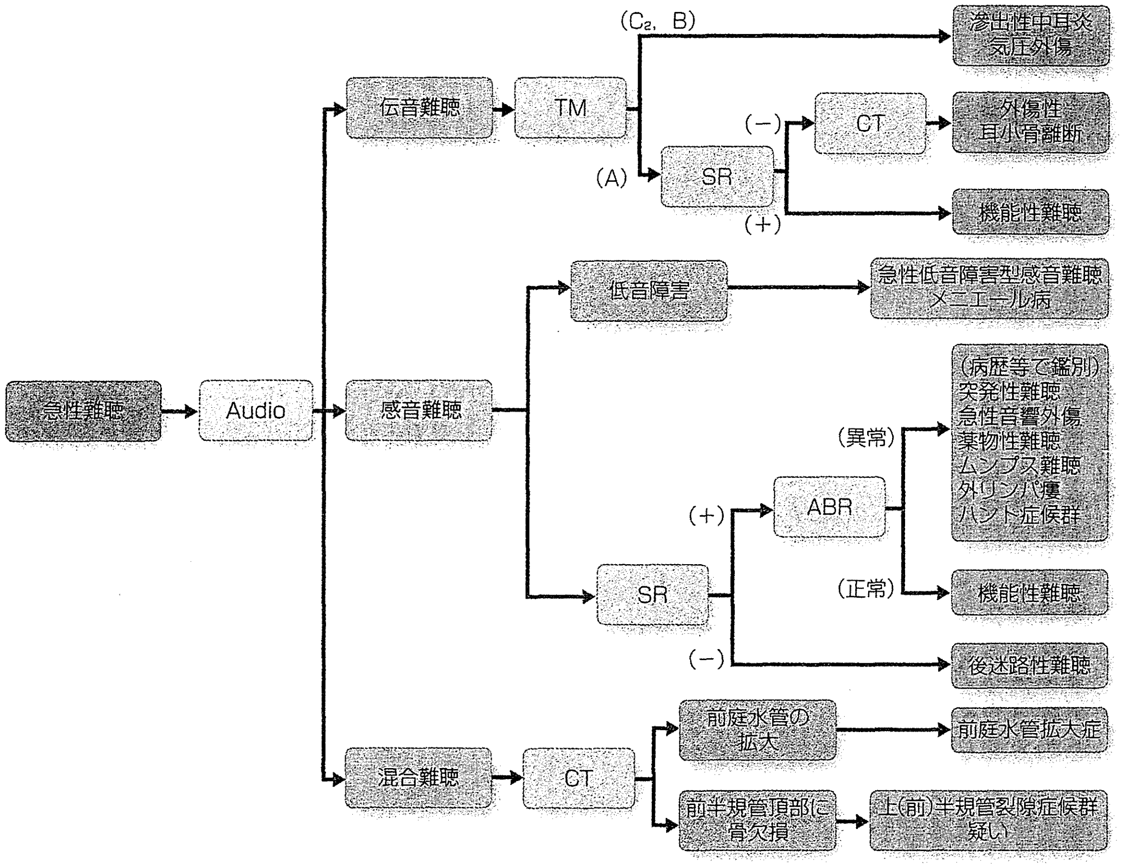
②伝音難聴の場合, 次にティンパノメトリーを行う. もし C_2 ないしは B 型であれば中耳貯留液の可能性が高く, 滲出性中耳炎や病歴によっては気圧外傷が考えられる.

③A 型の場合には SR 検査を行う. SR が検出されない場合は急性難聴としては耳小骨離断が考えやすく, CT 検査が必要となる. 正常閾値で検出される場合は機能性難聴が疑われる.

④感音難聴の場合, まず低音障害型を示せば急性低音障害型感音難聴, めまい, ふらつきを伴えばメニエール (Ménière) 病が考えられる.

Topics　低音障害型感音難聴を示す遺伝性難聴

九州などでは遺伝性難聴で低音障害型感音難聴を示すものがあり, これはめまい・ふらつきを伴わず慢性に経過する. *WFS1* 遺伝子異常といわれており, 一般に薬物治療は奏効しない.



⑯難聴診断のためのフローチャート

Audio：純音聴力検査，SR：アブミ骨筋反射検査，TM：ティンパノメトリー．

⑤低音障害型以外の感音難聴ではSR検査を行う．SRが検出できなければ後迷路性難聴の可能性があるので，随伴症状の観察やMRIなどの中枢の検査が必要となる．

⑥SRが正常閾値で検出できれば内耳性難聴か機能性難聴と考えられるので，ABRを行う必要がある．いうまでもなく，ABRが正常なら機能性難聴で，検出されないなどの異常があれば内耳性感音難聴が考えられる．

⑦内耳性感音難聴には数多くの疾患があるが，それらを鑑別する聴覚的検査はほとんどなく，特徴的病歴などで鑑別を行う．

⑧まれではあるが混合難聴の場合には，前庭水管拡大症や上（前）半規管裂隙症候群であることがあるので，CTを撮る必要がある．

（髙橋晴雄）

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Observation of Cortical Activity During Speech Stimulation in Prelingually Deafened Adults With Cochlear Implantation by Positron Emission Tomography–Computed Tomography

Haruo Yoshida, MD; Yukihiko Kanda, MD; Haruo Takahashi, MD; Izumi Miyamoto, PhD; Kenya Chiba, MD

Objectives: We evaluated the cortical activity of 2 successful prelingually deafened adult cochlear implant (CI) users who have been trained by auditory-verbal/oral communication since childhood.

Methods: Changes in regional cerebral blood flow were measured by positron emission tomography using ^{18}F-fluorodeoxyglucose while the subjects were receiving auditory language stimuli by listening to a story. Ten normal-hearing volunteers were observed as age-matched control subjects.

Results: In both cases, the auditory-related regions, when compared to same regions in the control subjects, showed hypermetabolism in the left dorsolateral prefrontal cortex and the left precentral gyrus — similar to that in successful CI users who are prelingually deafened children or postlingually deafened adults. Both subjects had the ability to activate these areas, and this ability might be one of the reasons that accounts for such exceptionally good performance in older prelingually deaf CI users. As for the visual-related regions, hypometabolism was observed in Brodmann areas 18 and 19, and this finding might be related to the intensive auditory-verbal/oral education that the subjects had received since childhood.

Conclusions: Despite the limits imposed by the small sample size and the spatial resolution of positron emission tomography, this study yielded insights into the nature of the brain plasticity in prelingually deafened adults who are successful CI users.

Key Words: auditory cortex, auditory-verbal/oral education, positron emission tomography, regional cerebral blood flow.

INTRODUCTION

Although cochlear implantation is recognized as an effective treatment for patients with severe to profound sensorineural hearing loss (SNHL),[1-3] prelingually deaf adolescents and adults have achieved only limited postimplantation improvement, and hence have not been considered good candidates for implantation.[4-7] In previous reports, age at implantation and duration of deafness were pointed out as the most important factors in influencing postimplantation performance.[2,8]

On the other hand, there has been some literature reporting good performance in language perception even in prelingually deaf adults.[9-12] We reported that cochlear implantation can be recommended to some prelingually deafened adults if they receive good habilitation with consistent auditory-verbal/oral training using well-fitted hearing aids (HAs).[13] The clinical reasons for such exceptionally good performances in these implantees are considered to be related to the recently improved quality of cochlear implants (CIs)[14-16] and/or use of an aurally based educational program.[9] However, there have been few studies on functional neuroimaging of cortical activity in successful prelingually deaf adults with CIs. The aim of this study was to evaluate the brain metabolic activity of prelingually deaf adults who are successful users of CIs and have been educated with auditory-verbal/oral communication since early childhood.

MATERIALS AND METHODS

Subjects. The subjects were 2 prelingually deafened adults who constituted 0.7% of the total of 279 patients who underwent cochlear implantation at Nagasaki University Hospital from 1997 to 2010. Details of their clinical information are shown in Table 1. Both of them showed 90 dB or worse hear-

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TABLE 1. CLINICAL INFORMATION

	Subject 1	*Subject 2*
Age at CI	34 y	29 y
Sex	Female	Male
Implanted ear	Left	Right
Time since CI	70 mo	24 mo
Age at receipt of HA	18 mo	18 mo
Education	Deaf and ordinary school	Deaf school
Cause of deafness	Unknown	Unknown
Family history of HL	Yes (son)	Yes (father and brother)

CI — cochlear implantation; HA — hearing aid; HL — hearing loss.

ing loss on auditory brain stem response testing before the age of 2 years, and thus received a diagnosis of severe to profound SNHL. Thus, both diagnoses were of prelingual deafness rather than progressive hearing loss.

Subject 1 received a diagnosis of severe hearing loss at the age of 1 year, and high-power box HAs were fitted in both ears at the age of 1½ years. She attended an ordinary elementary school in which she received habilitation by auditory-verbal/oral communication until high school. She underwent cochlear implantation at 34 years of age, and positron emission tomography (PET) superimposed with brain computed tomography images (PET-CT) was performed 70 months later, by which time her articulation was slightly distorted but her communication abilities with the CI were excellent, leading to improvements in the quality of her life. Her use of the CI is the longest and most successful among prelingually deafened adult users of CIs in our institute. The cause of her hearing loss is uncertain, as genetic testing has not been performed, but her son also has severe congenital hearing loss (Table 1).

Subject 2 received a diagnosis of severe hearing loss at the age of 1 year. He used high-power box HAs in both ears after the age of 1½ years, and attended an elementary school for deaf children in which he received habilitation by auditory-verbal/oral communication until high school. He underwent cochlear implantation at 29 years of age, and underwent our PET-CT study 24 months later, by which time his communication skills with the CI were excellent.

Ten right-handed adult volunteers (mean age, 27.1 years; age range, 22 to 34 years; 6 male and 4 female) with normal hearing (pure tone air conduction thresholds of less than 20 dB hearing level at 0.5, 1.0, and 2.0 kHz) and without any evidence of

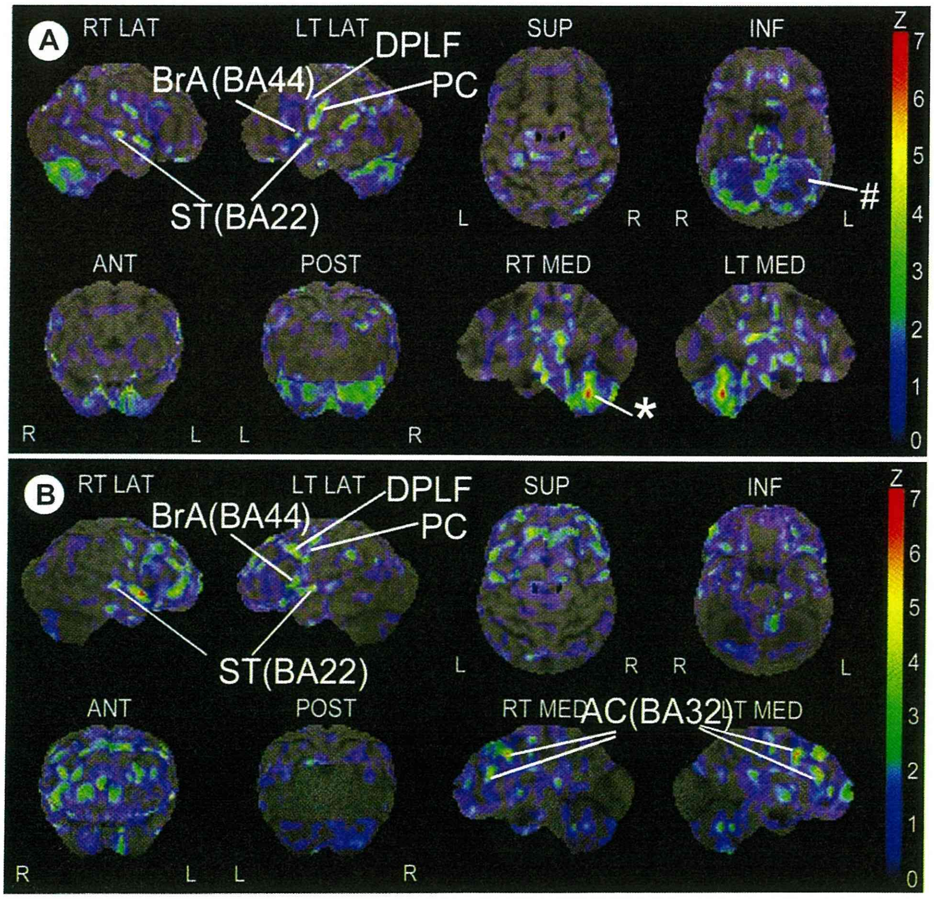

Fig 1. Z-score map computed by iSSP (Interface Stereotactic Surface Projection) application (iNEUROSTAT+) shows increased ^{18}F-fluorodeoxyglucose uptake areas examined by positron emission tomography–computed tomography. **A)** Subject 1. **B)** Subject 2. Cortical activity is shown as grading image of more-deviant regions in red (*) and less-deviant regions in purple (#) by pixel. R, RT — right; L, LT — left; LAT — lateral; SUP — superior; INF — inferior; ANT — anterior; POST — posterior; MED — medial; BA — Brodmann area; ST — superior temporal gyrus; DPLF — dorsolateral prefrontal cortex; PC — precentral gyrus; AC — anterior cingulate gyrus.

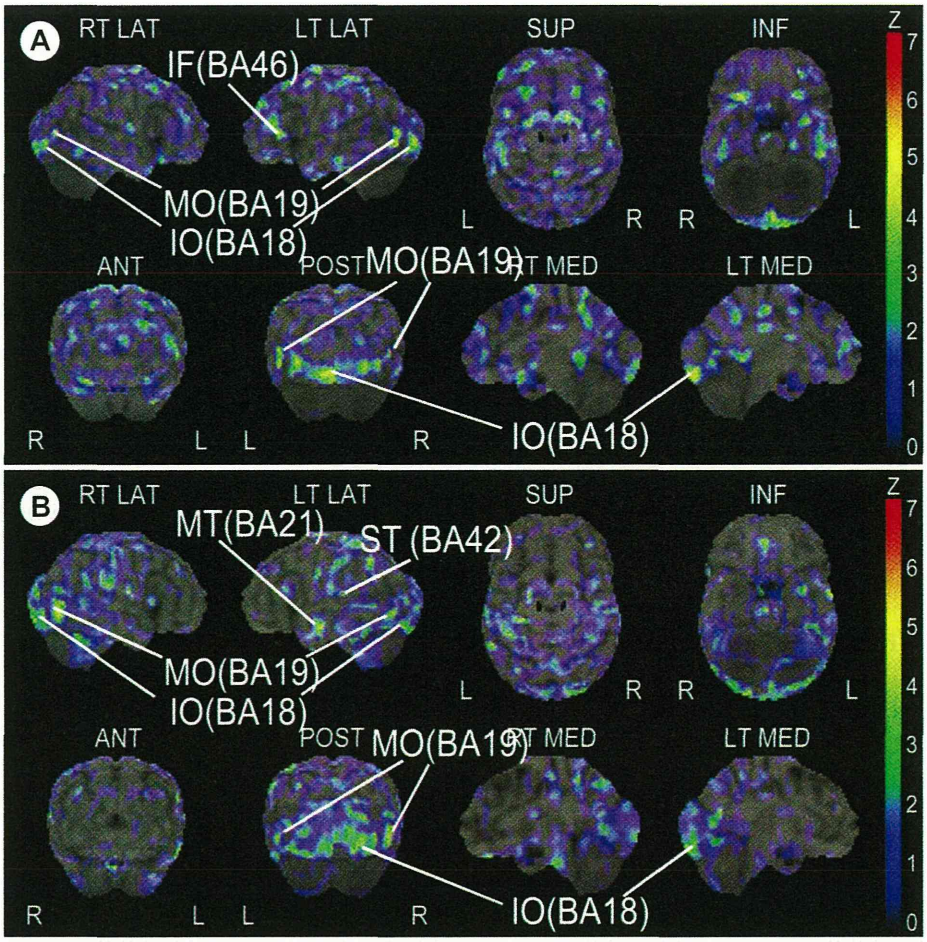

Fig 2. Decreased ^{18}F-fluorodeoxyglucose uptake areas examined by positron emission tomography–computed tomography. **A)** Subject 1. **B)** Subject 2. BA — Brodmann area; IF — inferior frontal gyrus; MO — middle occipital gyrus; IO — inferior occipital gyrus; MT — middle temporal gyrus; ST — superior temporal gyrus.

ear disease, history of noise exposure, previous ear surgery, or severe head injury were observed as control subjects who underwent the PET-CT study in the same manner as the 2 study subjects. All of the subjects gave written informed consent before participating in the study, which was approved by the ethical committee of our institute (approval number 08043067).

Scans. We injected 4 MBq/kg of ^{18}F-fluorodeoxyglucose (^{18}F-FDG) intravenously 72 seconds before the PET scan of the brain. All of the subjects were instructed to listen to a 40-minute story that was read aloud by one speaker located out of sight of the subjects. Afterward, a video recording of the session was reviewed to ensure that the participants had been listening carefully throughout, and several questions were posed on the contents of the task story to exclude those subjects from the study who were unable to correctly answer 70% of the questions. The ^{18}F-FDG–PET scans were performed with a Discovery ST PET scanner (GE Medical Systems, Milwaukee, Wisconsin).

Analysis of Scans. The original ^{18}F-FDG–PET image data were transformed into a binary format and then into a stereotactic standard Talairach space by use of the 3-dimensional Stereotactic Surface Projection (3D-SSP) application of the iNEUROSTAT+ program,[17] and the cortical radioactivity of the 2 CI users was compared with that of the group of normal-hearing adults. The usefulness of this program as compared with Statistical Parametric Mapping (SPM) has been reported.[17-19] Stereotactic coordinates and anatomic regions, including Brodmann areas (BAs), were automatically computed. The resulting images are shown as a Z-score map, which was computed by each study subject's score derived from the mean score of the standard deviation of the control subjects, and are shown as graded images from more-deviant regions in red to less-deviant regions in purple by pixel (Figs 1 and 2).

RESULTS

Table 2 shows the preoperative hearing level, the aided hearing level with HAs, the preoperative and postoperative speech discrimination scores, and the speech perception rates with CIs of the 2 study subjects. Their mean preoperative hearing levels were greater than 100.0 dB, and their aided hearing levels were 52.5 dB and 61.3 dB, respectively. After co-

TABLE 2. AUDIOLOGICAL OUTCOMES

	Subject 1	*Subject 2*
Before implantation		
Mean hearing level right ear	105.0 dB	105.0 dB
Mean hearing level left ear	105.0 dB	105.0 dB
Mean aided hearing level	52.5 dB	61.3 dB
SDS (auditory and visual)	43%	23%
SDS (auditory)	10%	0%
After implantation		
Mean hearing level with implant	28.7 dB	28.8 dB
SDS (auditory and visual)	73%	53%
SDS (auditory)	47%	37%
Speech perception rates*	90%	85%

SDS — speech discrimination score.
*Only open-set sentences without hearing aids.

chlear implantation, their hearing levels improved to 28.7 dB and 28.8 dB, respectively. Their speech discrimination scores, with auditory stimulation only, also improved, from 10% to 47% and from 0% to 37%, respectively. Their postoperative speech perception rates, which were tested by open-set sentences, were as good as 90% and 85%, respectively.

Regions showing significant increase and decrease in ^{18}F-FDG uptake compared to the control subjects are displayed in Fig 1 and Fig 2, respectively. As for the auditory-related regions, the ^{18}F-FDG uptake was increased in the left dorsolateral prefrontal cortex, the left precentral gyrus, and the superior temporal gyrus (BA22) in both subjects (Fig 1). Only in subject 2 was hypermetabolism observed bilaterally in the anterior cingulate gyrus (BA32; Fig 1B).

On the other hand, hypometabolism was observed in the left inferior frontal gyrus (BA46) of subject 1 and in the left middle temporal gyrus (BA21) of subject 2 (Fig 2). As for the visual-related regions, hypometabolism was observed bilaterally in the occipital gyrus (BA19 and BA18) in both subjects.

DISCUSSION

In many of the previous reports about prelingually deafened CI users, the results of deaf children were compared with those of adult control subjects.[20-24] However, the glucose metabolism in the brain changes with age, and this may have had an influence on the interpretation of brain activity results.[22,25] Therefore, in this study, we sought to compare prelingually deafened adults with age-matched normal-hearing adults in order to avoid age-related errors in interpreting the results and identifying neurophysiological factors that might determine the outcome of cochlear implantation. The brain metabolic activity of prelingually deaf adult CI users was evaluated by auditory stimulation only, in order to avoid visual-related effects such as lipreading.

Both subjects showed hyperactivity in the left dorsolateral prefrontal cortex. Functionally, the left dorsolateral prefrontal cortex is generally recognized as participating in higher cognitive functions such as reasoning, control of attention, and working memory.[26,27] In postlingually deafened adult subjects who showed good performance in language perception with a CI, this area was reported to be activated while the subjects were listening to voices.[28] In prelingually deaf children with a CI, postoperative speech scores were associated with enhanced metabolic activity in this area, and subjects in whom this region becomes active during spontaneous brain activity are believed to have an advantage in acquisition of auditory language.[29] The left precentral gyrus was also hypermetabolic in both of our subjects. According to the previous report, these regions are crucial for language processing and are particularly implicated in processing of speech with a CI, and deaf subjects who engage these regions tended to have better performance.[29] According to those results, the ability of our subjects to activate these areas might have been related to their exceptionally good performance with CIs.

The ^{18}F-FDG uptake was increased bilaterally, especially in the left superior temporal gyrus (BA22), in both subjects. Lee et al[29] reported that the age at implantation was positively correlated with increased activity in the right superior temporal gyrus, and their results seem compatible with our present results.

Only in subject 2, hypermetabolism was observed bilaterally in the anterior cingulate gyrus (BA32), and this area is believed to be involved in attention and arousal processes.[30,31] Although both subjects were successful CI users by the standards of prelingually deafened adults, subject 2, who is a more recent user of CIs and has less developed speech perception skills with CIs than does subject 1, often needs to pay more attention when communicating with CIs than does subject 1. In other words, subject 1 does not need to make as much of an effort when communicating with CIs in daily life.

As for the visual-related regions, the integration of audiovisual inputs in auditory speech perception was reported to be crucial for successful speech perception in subjects with CIs in a series of neuroimaging studies.[28,32] The brain activity of children with early-onset deafness was greater in the medial visual cortex and bilateral occipitoparietal junctions after cochlear implantation, and these findings suggest that speech learning resulted in a greater demand on the visual and visuospatial processing subserved by the early visual cortex and parietal cortices.[33] In our study, however, hypometabolism was observed in the visual-related regions of the posterior medial lingual cortex (BA18 and BA19). We speculate that this result may be related to the fact that both subjects had received intensive auditory-verbal/oral education without visual sensation since childhood. Because communication without visual information is a part of the auditory-verbal/oral education, it may be thought that the auditory center of a subject who was educated in this manner becomes active so as to avoid utilizing visual pathways during listening and that hypometabolism in the visual center thus develops.

In this study, we used the iNEUROSTAT+ program for anatomic standardization and 3-dimensional Stereotactic Surface Projection (3D-SSP), which can more correctly translate data from an an atrophied brain to that of a standard brain[18] and complement inappropriate anatomic standardization.[17] Moreover, Statistical Parametric Mapping (SPM) normalizes the metabolic counts to global counts, and iNEUROSTAT+ can normalize pixel values of an individual's image set to the whole brain, the thalamus, the pons, the cerebellum, or the sensorimotor region and select a suitable region as a reference region for normalization.[19]

CONCLUSIONS

Even in prelingually deafened adults, intensive training by auditory-verbal/oral communication since childhood may activate the auditory-related cortices by language stimulation through a CI as in postlingually deafened successful CI users, and might result in exceptionally good CI performance, as in the present 2 subjects. Despite the limits imposed by the small sample size and the spatial resolution of PET, this study yields insights into the nature of the brain plasticity in successful adult users of CIs with prelingual deafness.

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Original Article

Bilateral Cochlear Implantation for Children in Nagasaki, Japan

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Objectives. The number of patients with bilateral cochlear implant (CI) has gradually increased as patients and/or parents recognize its effectiveness. The purpose of this report is to evaluate the efficacy of 29 bilateral CI out of 169 pediatric CI users, who received auditory-verbal/oral habilitation at our hearing center.

Methods. We evaluated the audiological abilities 29 Japanese children with bilateral CIs including wearing threshold, word recognition score, speech discrimination score at 1 m from front speaker (SP), 1 m from second CI side SP, speech discrimination score under the noise (S/N ratio=80 dB sound pressure level [SPL]/70 dB SPL, 10 dB) at 1 m from front SP, word recognition score under the noise (S/N ratio=80 dB SPL/70 dB SPL, 10 dB) at 1 m from front SP.

Results. Binaural hearing using bilateral CI is better than first CI in all speech understanding tests. Especially, there were significant differences between the results of first CI and bilateral CI on SDS at 70 dB SPL ($P=0.02$), SDS at 1 m from second CI side SP at 60 dB SPL ($P=0.02$), word recognition score (WRS) at 1 m from second CI side SP at 60 dB SPL ($P=0.02$), speech discrimination score (SDS) at 1 m from front SP under the noise (S/N=80/70; $P=0.01$) and WRS at 1 m from front SP under the noise (S/N=80/70; $P=0.002$). At every age, a second CI is very effective. However, the results of under 9 years old were better than of over 9 years old on the mean SDS under the noise (S/N=80/70) on second CI ($P=0.04$). About use of a hearing aid (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.

Conclusion. These results may show important binaural effectiveness such as binaural summation and head shadow effect. Bilateral CI is very useful medical intervention for many children with severe-to-profound hearing loss in Japan as well as elsewhere.

Key Words: *Cochlear implant, Children, Bilateral, Binaural, Binaural summation, Head shadow effect, Japan*

INTRODUCTION

The clinical effects of both unilateral and bilateral cochlear implantation (CI) in children is well established internationally (1-15), but there are only a few reported cases of bilateral CI in Japan. The number of patients with bilateral CI has gradually increased as patients and/or parents recognize its effectiveness. The following are some of the bilateral CI cases in children that we have experienced. This is a review of bilateral CI in 169 pediatric CI users, who received auditory-verbal/oral habilitation at our clinic.

This study's aim is to obtain answers to the following questions. 1) Until what age the second CI is effective for better language perception in various situations? 2) Does the use of a hearing aid (HA) on the opposite side of first CI affect the re-

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sults after second CI? 3) Is there any critical time span between the first and second CI for their progress in language perception? 4) What is the advantage of bilateral CI over unilateral CI?

MATERIALS AND METHODS

Subjects

Since we started CI surgery in 1997, out of 169 children undergoing CI rehabilitation in our clinic, 29 children (17%) had bilateral CI for at least half a year before May 2011. The age of the children at the first CI operation ranged from 1 year 4 months to 15 years 5 months, whereas the age of children at the second CI operation ranged from 2 year 1 month to 15 years 10 months (Fig. 1). The most common age for the first CI was 1 or 2 years. The interval between first and second CI fitting ranged from 5 months to 10 years 1 month. This can be considered a relatively wide range, but the most frequent interval between the two CIs was under 1 year (Fig. 2). The period on non-use of their HA before the second CI is also valuable: it ranges from 0 month to

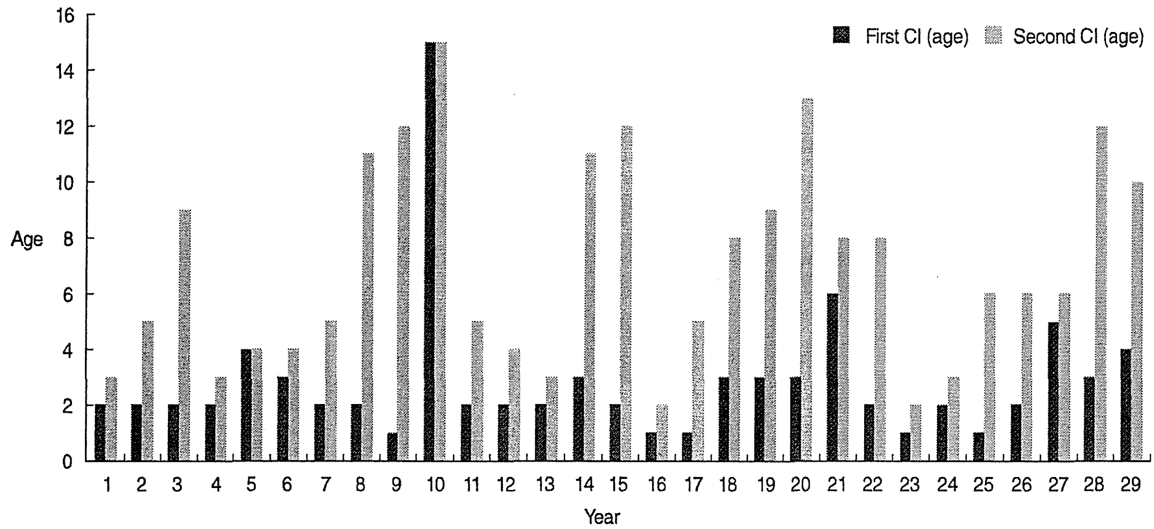


Fig. 1. Age at operation of first cochlear implantation (CI) and second CI (year).

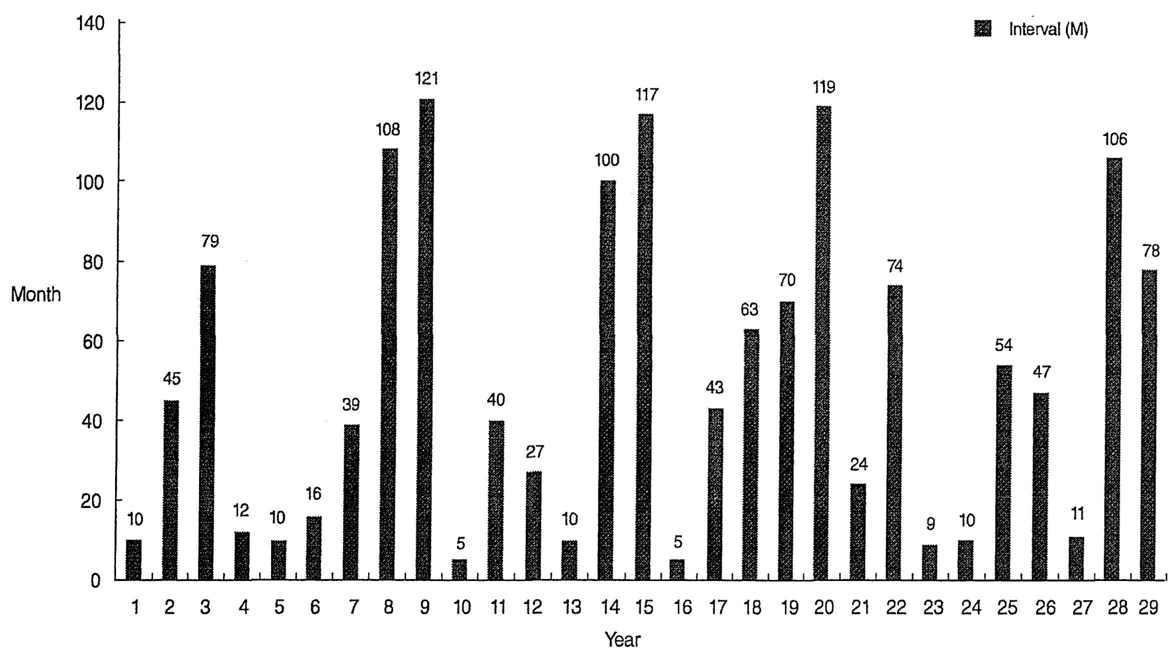


Fig. 2. Interval between first and second cochlear implantation (month).

108 months (9 years). Twelve cases did not remove their HA before the second CI (Fig. 3). Causes for deafness were described in Fig. 4. The devices used are described in Fig. 5.

We examined 19 children who acquired language with either CI or HA using various audiological tests. Children with severe anomaly or late development were not included. The children were divided into 2 groups to evaluate the amount of habilitation time after the second CI. The first group (group A) consisted of 11 children who had their second CI for at least a year. The second group (group B) included 8 children who had their second CI between 6 and 12 months.

Methods

We evaluated audiological abilities including 1) wearing threshold (WTH); 2) word recognition score (WRS, TY-89; Japanese-3 syllabic word-CD, at 60 dB sound pressure level [SPL], at 70 dB SPL); 3) speech discrimination score (SDS, 67-S; Japanese-monosyllabic word-CD, at 60 dB SPL, at 70 dB SPL) at 1 m from front speaker (SP), 1 m from second CI side SP; 4) SDS under noise (67-S; Japanese-monosyllabic word-CD, S/N ratio=80 dB SPL/70 dB SPL, 10 dB) at 1 m from front SP; 5) WRS under noise (TY-89; Japanese-3 syllabic word-CD, S/N ratio=80 dB SPL/70 dB SPL, 10 dB) at 1 m from front SP (noise: speech noise). We conducted all tests in a shielded room. Statistical analysis was done using the Student's *t*-test and paired *t*-test.

RESULTS

The mean WTH using first CI, second CI, and bilateral CI shows that all WTH is nearly the same ranging from 25 dB hearing level (HL) to 35 dB HL (Fig. 6). There were no significant differences between them. The mean WTH of their HA before he second CI was from 55 dB HL (for lower frequencies) to 65 dB HL (for higher frequencies). However, after operation the mean WTH using second CI ranges from almost 30 dB HL to 35 dB HL. There were significant difference ($P=0.03$*) between HA and

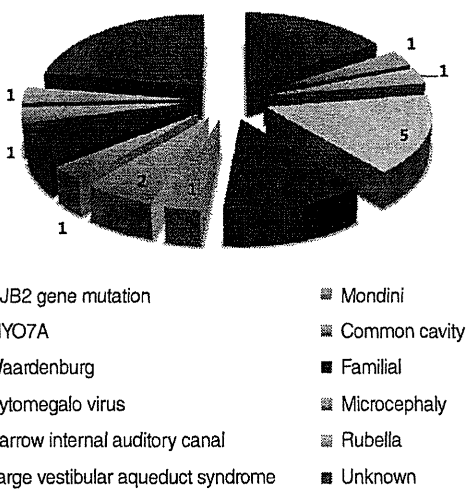


Fig. 4. Causes for deafness.

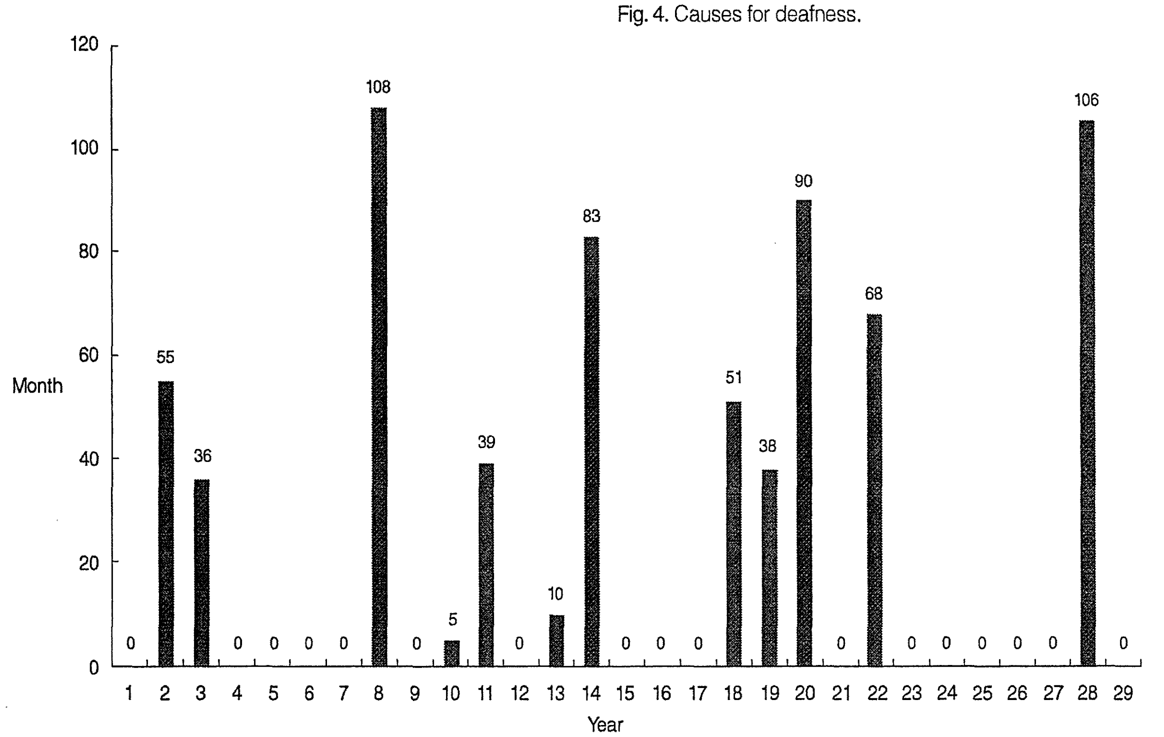


Fig. 3. Number of months child discontinued hearing aid use before second cochlear implantation.

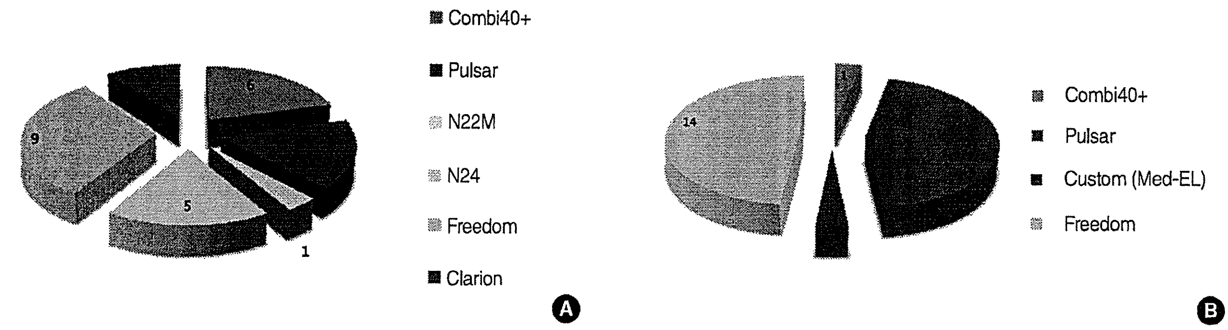
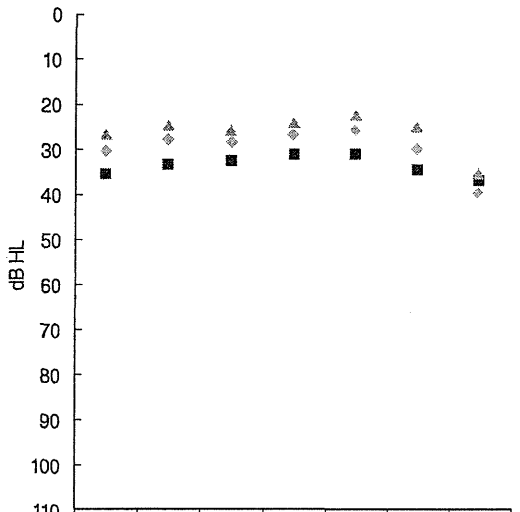
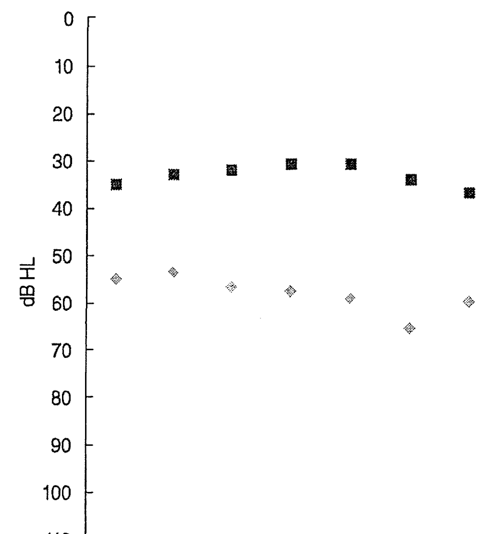


Fig. 5. Device of first cochlear implantation (CI) (A) and second CI (B).



	125 Hz	250 Hz	500 Hz	1 kHz	2 kHz	4 kHz	8 kHz
◈ First CI	30.3	27.8	28.3	26.8	25.8	29.8	39.5
■ Second CI	35.5	33.3	32.5	31	31	34.3	37
▲ Bilateral CI	26.8	24.8	25.8	24.3	22.5	25	35.4

Fig. 6. The mean wearing threshold using first cochlear implantation (CI), second CI, and bilateral CI.



	125 Hz	250 Hz	500 Hz	1 kHz	2 kHz	4 kHz	8 kHz
◈ HA	55.4	54.1	57	57.9	59.4	65.8	60
■ Second CI	35.5	33.3	32.5	31	31	34.3	37

Fig. 7. The mean wearing threshold of their hearing aid (HA) before second cochlear implantation (CI) and second CI.

second CI (Fig. 7).

The mean WRS at 1 m from the front SP at 70 dB SPL is shown in Fig. 8. The mean score for the second CI in group A was similar to the mean score for the first CI. The mean score on the WRS for all cases shows that there were no significant differences between the results of the first CI and the bilateral CI at 70 dB SPL (P= 0.13).

The mean WRS at 1 m from the front SP at 60 dB SPL is described in Fig. 9. The mean score for the second CI in group A was similar to the mean score for the first CI. For all cases, there were no significant differences between the results of the first CI and bilateral CI at 60 dB SPL (P=0.05).

The mean SDS at 1 m from the front SP at 70 dB SPL is described in Fig. 10. The mean score for the second CI in group A was similar to the mean score for the first CI. The SDS results show that there were significant differences between the results of the first CI and the bilateral CI at 70 dB SPL (P=0.02*).

The mean SDS at 1 m from the front SP at 60 dB SPL is described in Fig. 11. There were no significant differences between the results of the first CI and the bilateral CI at 60 dB SPL (P=0.24).

The mean SDS at 1 m from the second CI side SP at 70 dB SPL is described in Fig. 12. The mean score for the second CI in group A was similar to the mean score for the first CI. There were no significant differences between the results of the first CI and the bilateral CI at 1 m from the second CI side SP on all cases at 70 dB SPL (P=0.25).

The mean SDS at 1 m from the second CI side SP at 60 dB SPL is described in Fig. 13. 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*).

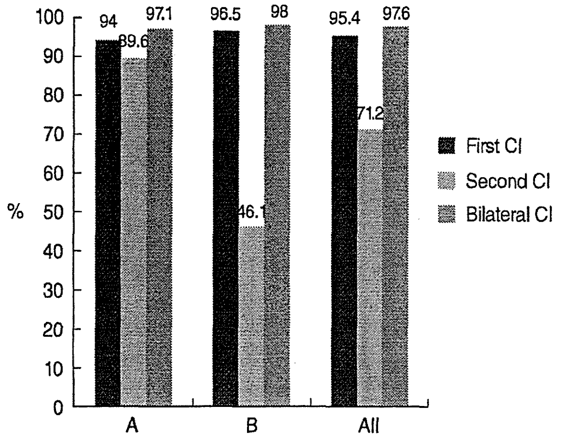


Fig. 8. Mean word recognition score at 1 m from front speaker at 70 dB SPL (0.13). CI, cochlear implantation; SPL, sound pressure level.

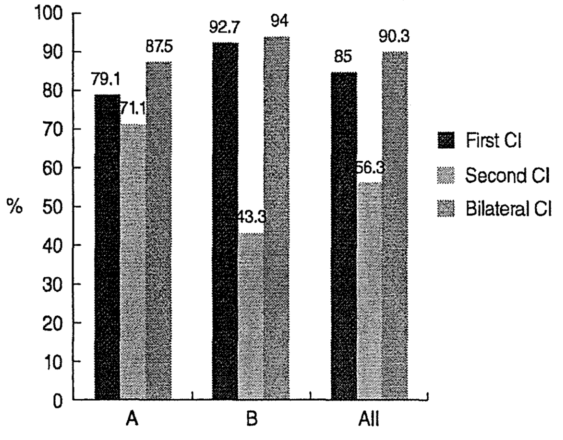


Fig. 9. Mean word recognition score at 1 m from front speaker at 60 dB SPL (P=0.05). CI, cochlear implantation; SPL, sound pressure level.

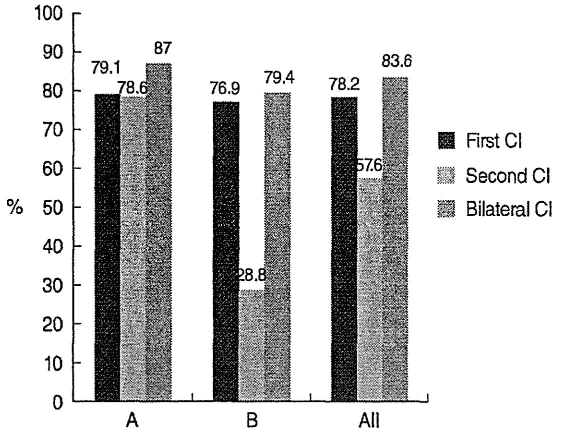


Fig. 10. Mean speech discrimination score at 70 dB SPL (P=0.02*). CI, cochlear implantation; SPL, sound pressure level.

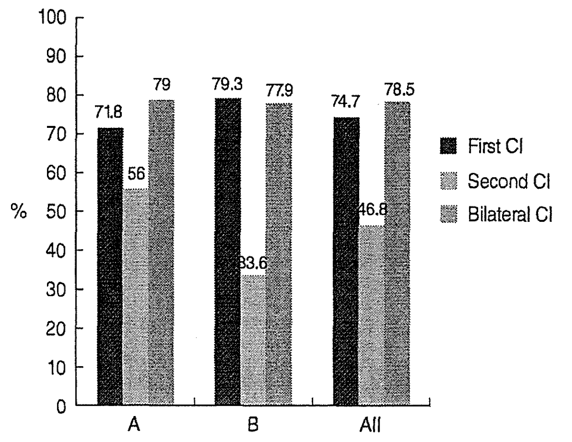


Fig. 11. Mean speech discrimination score at 60 dB SPL (P=0.24). CI, cochlear implantation; SPL, sound pressure level.

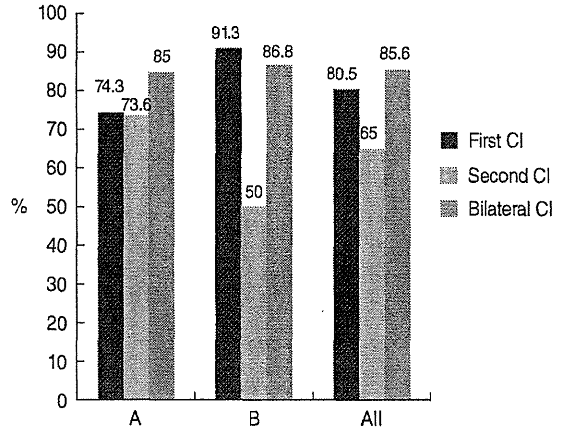


Fig. 12. Mean speech discrimination score at 1 m from second cochlear implantation (CI) side speaker at 70 dB SPL (P=0.25). SPL, sound pressure level.

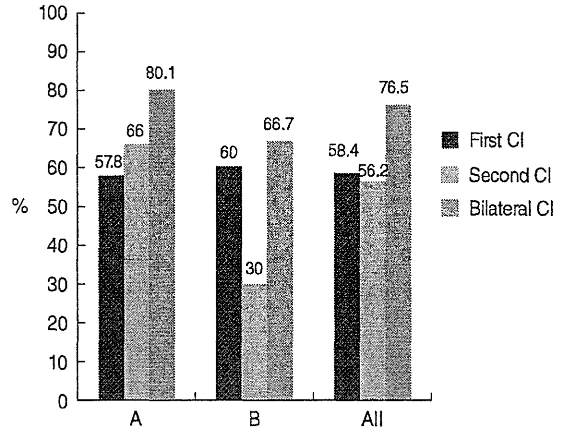


Fig. 13. Mean speech discrimination score at 1 m from second cochlear implantation (CI) side speaker at 60 dB SPL (P=0.02*). SPL, sound pressure level.

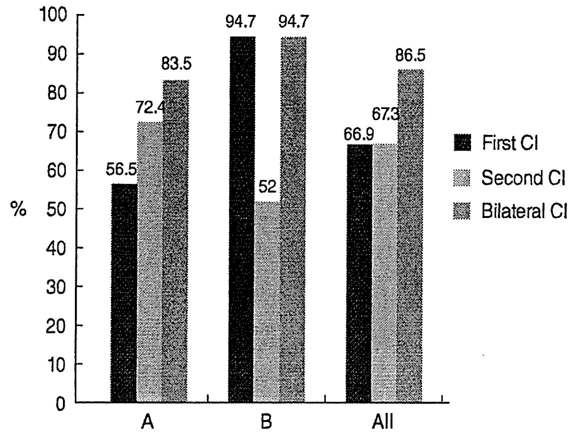


Fig. 14. Mean word recognition score at 1 m from second cochlear implantation (CI) side speaker at 60 dB SPL (P=0.02*). SPL, sound pressure level.

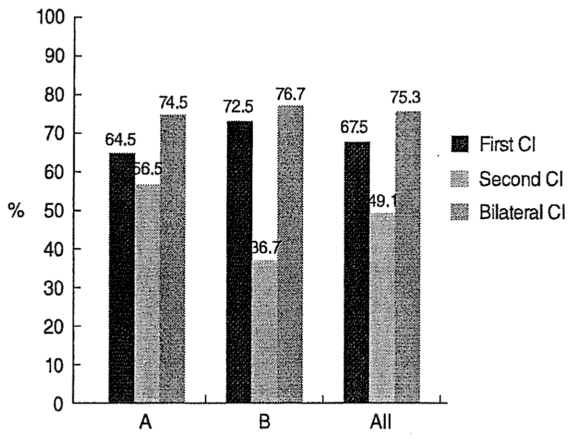


Fig. 15. Mean speech discrimination score at 1 m from front speaker under the noise (S/N=80/70, +10; P=0.01*). CI, cochlear implantation.

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

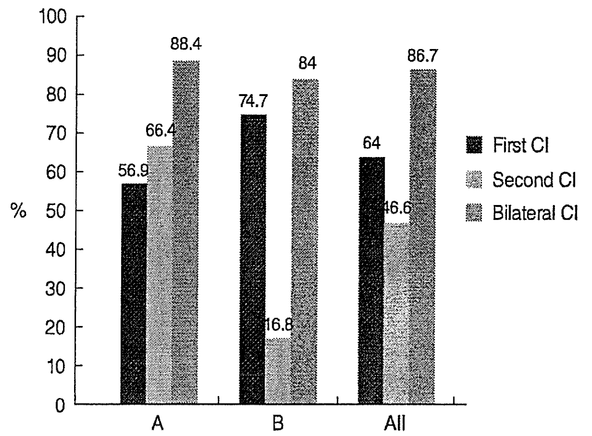


Fig. 16. Mean word recognition score at 1 m from front speaker under the noise (S/N=80/70, +10; (P=0.002**). CI, cochlear implantation.

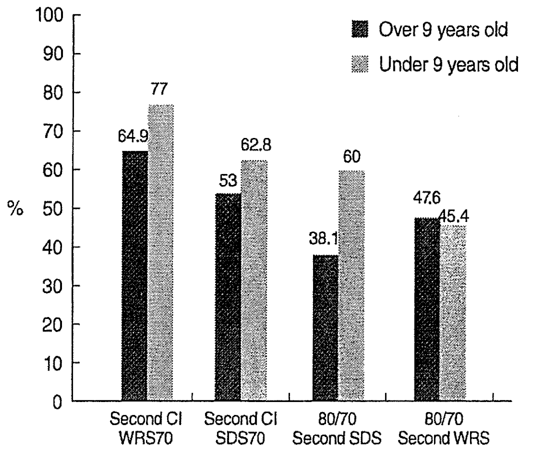


Fig. 17. Comparison of the mean word recognition score (WRS) and speech discrimination score (SDS), at 70 dB SPL at 1 m from front speaker, the mean SDS and WRS under the noise (S/N=80/70) on second cochlear implantation for over and under 9 years old. SPL, sound pressure level.

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).

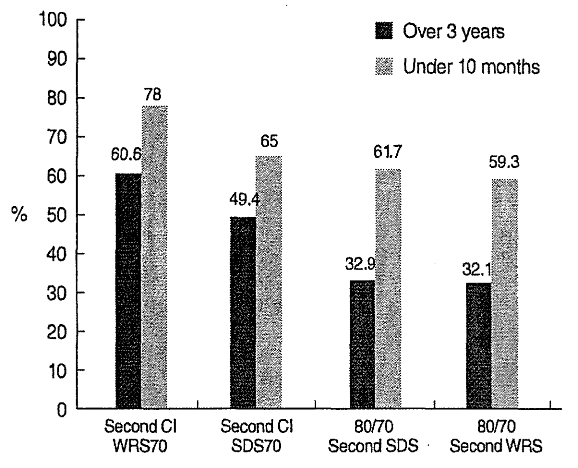


Fig. 18. Comparison of the mean word recognition score (WRS) and speech discrimination score (SDS) at 70 dB SPL at 1 m from front speaker, the mean SDS and WRS under the noise (S/N=80/70) on second cochlear implantation (CI) for those that had not used their hearing aid (HA) for over 3 years before second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