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人工内耳装用症例における静寂下・騒音下での補聴器装用効果の検討

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要旨：両耳聴取の利点は様々認められており，海外では両耳人工内耳により，聴取成績の向上が報告されている。片耳装用しか認められていない本邦では，人工内耳の対側耳に補聴器を装用することが一般的である。今回我々は，対側補聴器併用の利点について，日本人患者で検討した。静寂下条件では，片側人工内耳装用と，人工内耳と対側補聴器併用の場合との間に，聴取成績に有意な違いはなく，装用期間等ほかの要因を考慮しても違いはなかった。騒音負荷条件でも，補聴器併用による有意な正答率の向上は認められなかった。今回の研究で，対側補聴器の効果が認められない理由として，人工内耳の適応条件が厳しいこと，症例数不足により有意差が出なかった可能性，日本語と欧米言語の言語的な特徴の違いによる要因などが考えられた。今回の結果から，人工内耳装用者の聴取能力を向上には，両側人工内耳装用などにより対側耳をより積極的に活用することが必要と考える。

ーキーワードー
両耳補聴，小児人工内耳，補聴器，言語獲得，カクテルパーティー効果

はじめに

両耳聴の効果は，正常者においても，音の方向感の認知やカクテルパーティー効果[1)]による騒音下の聴取能力など，さまざまな利点があげられている。難聴者においては両側補聴器装用による聴取能力の向上が認められている[2)]。

人工内耳は1973年に3Mにより商品化されて以来，聴覚を失った患者に対して広く用いられている。人工内耳装用症例において，海外では両耳装用についての報告が増加しており，一般的になりつつある[3)]。これらの報告では，両耳装用による騒音下での聴取の向上など，さまざまな効果が認められている[4)]。本邦では，人工内耳は，1991年に輸入認可，1994年に保険適応となり，一般的に行える治療法となったが，現在まで片耳装用しか認められていない。海外では片耳装用の場合でも人工内耳の対側耳に補聴器を装用することにより，聴取能の向上が認められている[5)]。本邦でも人工内耳対側耳への補聴器装用は一般的に行われているが，その効果に関する報告は少なく，本邦でのエビデンスは確立しているとはいえない[6)]。我々の施設においても，海外の文献に基づいて聴取能の向上を期待して人工内耳対側耳に補聴器装用を行っているが，今回，我々の施設の症例に対し，人工内耳単独装用と比較して補聴器併用時に有意な聴取能向上が得られるかにつき，静寂下および騒音負荷下の条件において検討を行った。

対象と方法

1997年から2008年にかけて，東京大学医学部附属病院耳鼻咽喉科において人工内耳埋込術を施行され

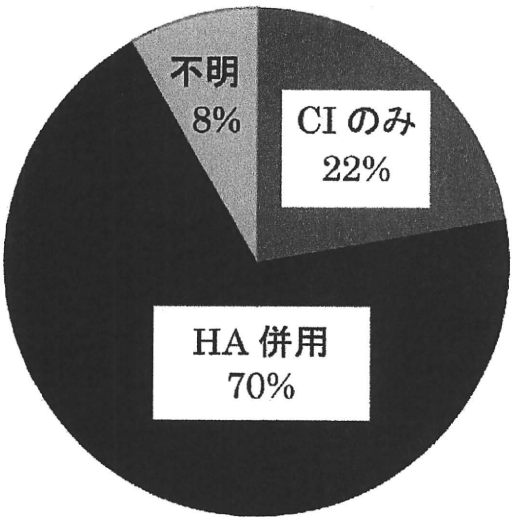

図1 当院での人工内耳装用者における補聴器の装用状況

た小児例108例のうち，補聴器を併用している症例は75例（70%）であった（図1）。人工内耳単独装用は24例（22%），不明は9例（8%）であった。

1．静寂下での聴取能評価

はじめに，静寂下での効果について検討した。対象は2008年までに当院にて人工内耳埋込術を行いかつ対側耳に補聴器を併用している小児例のうち，現在も通院中の症例の中からランダムに選んだ31例（男児15例，女児16例）であり，2006年から2009年にかけて装用効果の評価を行った。対象群の音入れの年齢は2才5ヶ月～7才4ヶ月（平均4才0ヶ月），評価時の年齢は3才5ヶ月～14才1ヶ月（平均7才5ヶ月），装用期間は0年11ヶ月～9年7ヶ月（平均3年5ヶ月）であった。検査は67-S語表を使用し，補聴器単独（HA），人工内耳単独（CI）と，対側に補聴器を併用（CI＋HA）した3条件下で検査を行った。3条件の検査順はランダムに行った。検査音は静寂下で，両側外耳道入口を結んだ正中の前方1mに設置したスピーカーより70dB SPLで呈示した。解析はpaired t 検定にて比較を行った。誤差範囲は標準誤差（SE）を呈示した。

HA単独，CI単独，CI＋HAの間で67-S語表聴取の正答率を比較した。装用効果は療育期間の長さ・対側耳の聴力レベル，補聴器の装用効果などの要因によって影響を受けている可能性もあるため，他の要素による影響があるかも併せて検討を行った。人工内耳装用開始後の療育期間は0年11ヶ月～9年7ヶ月であり，2群がほぼ同症例数になるように，36ヶ月を境にして，短期療育群（11-36ヶ月，15例）と長期間療育群（37ヶ月以上，16例）の2群にわけ検討した。人工内耳の対側耳の状況により併用効果が異なる可能性も考えられ，人工内耳適応基準の90dBHLに近い残存聴力がある症例の方が補聴器併用効果も高い可能性が存在し，その影響を検討した。対側耳の500Hz，1000Hz，2000Hz各周波数の4分法による平均聴力レベルは，80-120dBHLであり，2群がほぼ同数になるように，105dBHL以内（17例）と，105dBHL超（14例）の2群にわけ比較した。さらに対側耳で，補聴器をより良く使えている状態の方が，補聴器併用効果が高い可能性があるため，補聴器の聴取成績で2群にわけ，検討を行った。補聴器による正答率は0-75%であり，中央値で2群にわけ，0-5%の群（16例）と10%以上の群（15例）で比較を行った。人工内耳単独での正答率が良好で100%に近い場合，天井効果の為に補聴器併用効果が認められにくくなっている可能性も考えられるため，人工内耳単独での正答率によって2群に分けた検討も行った。人工内耳単独の正答率は20-100%であり，検討は2群がほぼ同数になるように80%以上の成績の群（14例）と，80%未満の群の2群（17例）に分けて，比較を行った。

2．騒音下での聴取能評価

次に騒音負荷下での効果について，静寂下の検討を行った症例の中からランダムに選んだ小児6例（男児3例，女児3例，6-9才　平均7.4才）を対象に検討を行った。これらの症例の平均装用期間は4年5ヶ月であった。検査はCI2004学童用日常生活文を使用した。刺激音は70dB SPLとし，CI2004検査実施手引書の双スピーカー法に従い，両側外耳道入口を結んだ正中の前方1mに設置した2台のスピーカーより，上方の1台からは語音を，下方の一台からはノイズを呈示した。測定は，CI，CI＋HA両条件下で行った。静寂下での評価の他に，スピーチノイズを50，60，65，70dB SPLと段階的に負荷を行った5条件で行った。条件は，静寂下から開始し，次にノイズを50dB SPLで負荷し，徐々に負荷を増大させて行った。音圧については，何れもスピーカーが呈示する音圧であり，ノイズについてはバ

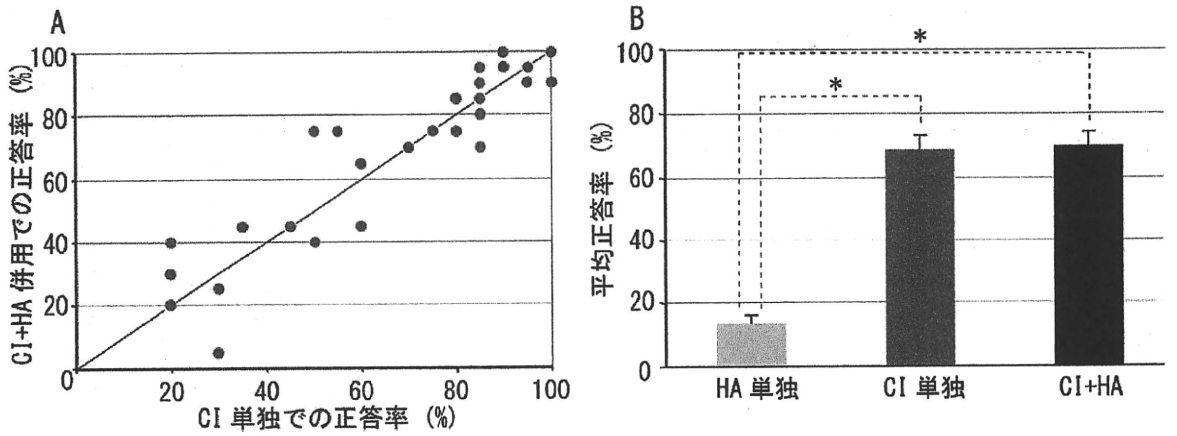

図2　補聴器併用の有無による正答率の比較
A：人工内耳単独と，補聴器併用時の正答率の分布。B：補聴器単独・人工内耳単独・補聴器併用各群の平均正答率。誤差範囲は，標準誤差。＊＜0.05

ンドノイズで校正を行った上で使用した。検査の際，被検者が疲労を訴えた場合，または段階的にノイズを負荷して，正答率が50％未満になった場合に中止とした。

結　　果

1）静寂下での効果

図2AにCI単独，CI＋HA併用の67-S語表の正答率の関係を示した。横軸はCI，縦軸はCI＋HAの正答率の分布である。CIとCI＋HAはほぼ一致した分布をしている。両条件とHA単独使用時の聴取成績のそれぞれの平均正答率を図2Bに示した。HAでは13.4±3.2%，CIでは68.7±4.7%，CI＋HAでは，69.5±13.4%であった。HAと比較して，CI，CI＋HAとも，有意に聴取成績は良好であった（p＜0.0001）。一方，CIとCI＋HAの間には有意差は認められなかった（p＝0.56）。

補聴器併用効果を得るには，長期の療育が必要な可能性も考えられるため，36ヶ月を境にして，短期療育群（11-36ヶ月）と長期間療育群（37ヶ月以上）の2群にわけ検討した。短期療育群での，CIの平均正答率は，60.3±7.0%，CI＋HAでは65.0±7.0%で，2条件間には有意差は認められなかった（p＝0.64）。長期療育群でも，CIでは76.5±5.9%，CI＋HAでは74.0±7.0%で，2条件間には有意差は認められなかった（p＝0.78）（図3）。

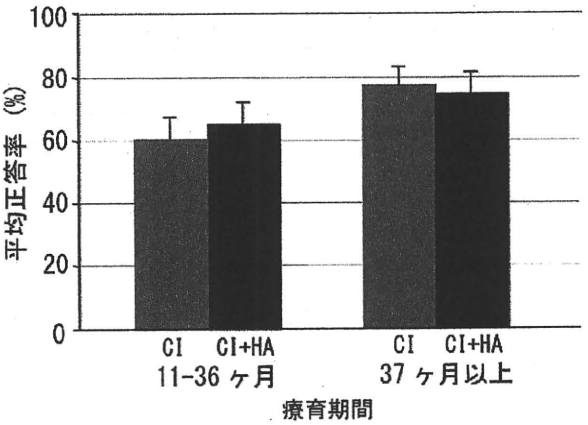

図3　療育期間の長さで分けた2群での比較

人工内耳の対側耳の残存聴力により105dBHL以内と105dBHL超で2群にわけ検討した。その結果，105dBHL以内群（CI：65.0±6.3%，CI＋HA：66.6±7.1%）でも，105dBHL超群（CI：73.2±6.9%，CI＋HA：72.9±7.3%）でも，CIと，CI＋HAでの聴取成績には有意差は認められなかった。つまり，残聴の程度による明らかな差は認められなかった（105dBHL以内群　p＝0.87，105dBHL超群　p＝0.97）（図4A）。

次に対側耳の補聴器の聴取成績の中央値で2群にわけ，0－5％の群と10%以上の群で検討を行った。0－5％群では，CIの平均正答率は，66.9±6.7%，CI＋HAは65.0±7.9%，10%以上の群ではCIは，70.7±6.8%，CI＋HAは74.0±7.0%であった。CIとCI＋HAの間には，0－5％の群と10%以上の群どちらにおいても，やはり有意差はみられなかった

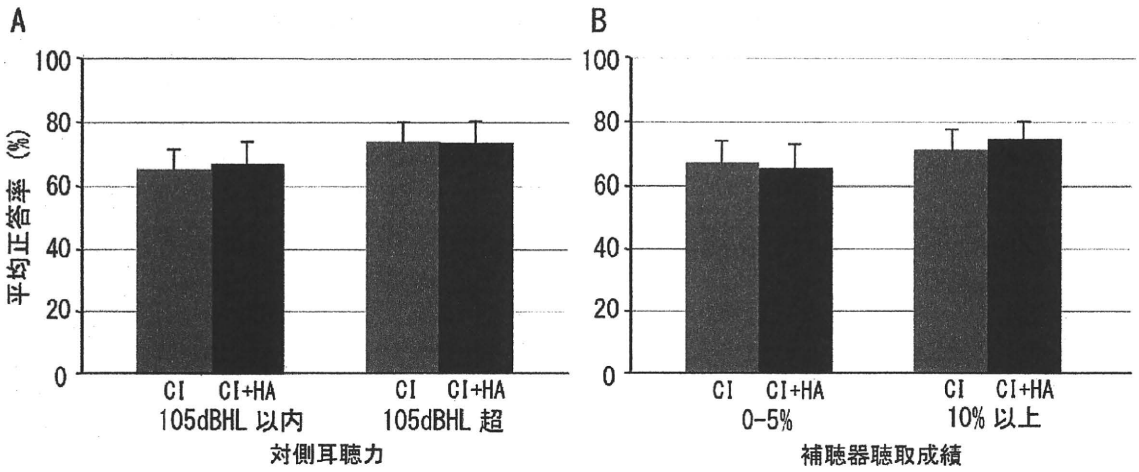

図4 対側耳の成績による比較
A：対側耳の聴力で分けた2群での比較。B：対側耳での補聴器聴取成績で分けた2群での比較。

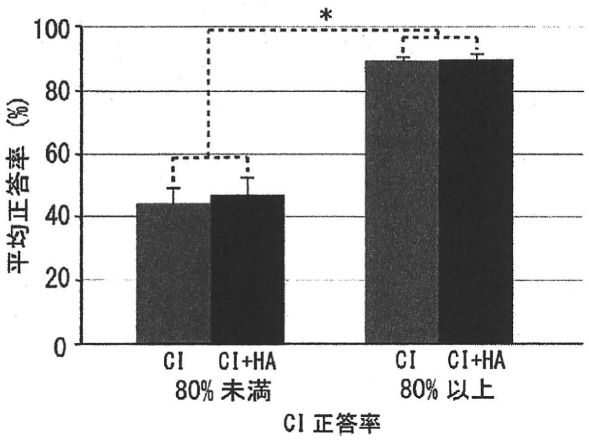

図5 人工内耳単独での聴取成績で分けた2群での比較

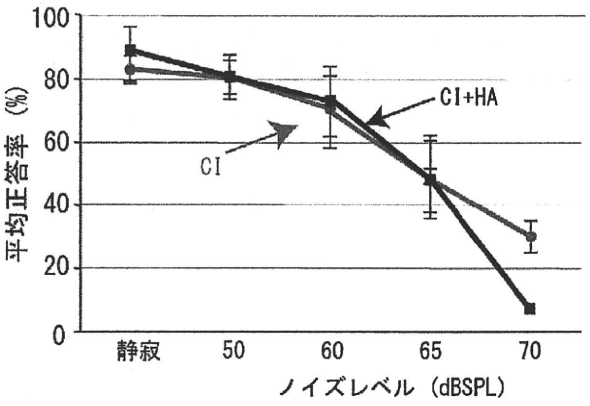

図6 騒音負荷下の人工内耳単独と補聴器併用での聴取成績
黒色は，補聴器併用条件下，灰色は，人工内耳単独条件下での聴取成績。誤差範囲は，標準誤差。

（0−5％群　$p=0.85$，10%以上群　$p=0.72$）（図4B）。

片耳人工内耳下の聴取成績における天井効果を除く為に，人工内耳単独での正答率が80%以上の成績の群と，80%未満の群の2群にわけ，検討を行った。80%以上では，CIの平均正答率は，44.3±5.0%，CI+HAは46.8±6.0%，80%未満ではCIは，88.8±1.6%，CI+HAは89.4±2.1%であった。聴取成績は，80%以上の群において，80%未満の群より良好であったが，いずれの群においても，CIとCI+HAの間では有意差は認められなかった（80%未満群　$p=0.75$，80%以上群　$p=0.84$）（図5）。

静寂下での検討結果をまとめると，CI・CI+HAとも有意差がみられず，他の要素（療育期間，対側耳の聴力，対側補聴器の聴取成績，人工内耳単独での成績）とあわせた検討でも，聴取成績に対する補聴器の併用効果は確認できなかった。

2）騒音負荷における検討

両耳聴取を行うと，カクテルパーティー効果により，騒音下での聴取に有利に働くことが考えられる。今回，片耳に人工内耳を装用した条件（CI）と，対側耳にも補聴器を併用して両耳聴できる条件（CI+HA）とにおける騒音下の聴取能比較をおこなった（図6）。ノイズ負荷が増大するにつれ，どち

らの条件下においても聴取成績が低下した。2群間の間にも統計上の有意差はみられなかった（静寂下：$p=0.32$，50dBSPL：$p=0.49$，60dBSPL：$p=0.47$，65dBSPL：$p=0.40$）。

考　察

今回，静寂下・騒音下いずれの条件の検討においても，人工内耳の対側耳に補聴器併用による聴取能の向上効果が認められなかった。この結果は海外の先行研究と異なっているが，その理由としていくつかの要因が考えられる。まず，海外に比べて，本邦では人工内耳適応基準が厳しいということが挙げられる。本邦での，人工内耳の適応においては，難聴の程度だけでなく，補聴効果が認められなかった症例を対象とするとされており，もともと補聴効果が期待できない症例が人工内耳の対象となっている。海外では，たとえば米国では，現在成人例では両側70dBHLで，補聴器で静寂下でのThe Hearing and Noise Test（HINT）の正答率が50%のレベルの難聴でも人工内耳の使用はFDAにより認可されているが，本邦では90dBHL以上の難聴者が対象であり，残聴に対する補聴効果が得られにくいと考えられる。二点目として，今回の検討では症例数が少なく，そのため統計的有意差が出にくかった可能性も考えられる。三点目としては，日本語と外国語では，発音，母音の出現頻度，周波数的特徴など，言語的な特徴に違いがあるため，日本語では補聴器併用効果が少ない可能性が考えられる。英語などの外国語では日本語に比べて子音，母音ともに種類が多く，また使われる周波数の幅も外国語の方が大きいと言われている。例えば英語の“r”と“l”のように，わずかなspectrumの違いが聞き分けに重要となってくるものも見られる[7]。チャンネル数の限られる人工内耳と異なり，補聴器ではすべての周波数を伝達することが可能であり，わずかなspectrumの違いの認知が必要な外国語の語音弁別において，より有効に働いているのかもしれない。日本語においても，松代ら[6]による成人6例の検討では，3例で補聴器併用による67-S語表の聴取成績の向上を認めている。この成人の中途失聴症例では失聴前は両耳聴が可能であったことから，両耳聴に関わる脳回路ができあがっていたと考えられる。今回のような小児例では先天的に失聴していたため，脳回路の発達が不十分で，対側耳に補聴器併用しても成人例より両耳聴が困難になっているのかもしれない。なお，今回検討を行わなかった音源定位や音楽聴取などの他の要素に補聴器併用の効果がある可能性は残されている。

人工内耳装用者において補聴器併用による静寂下および騒音下での聴取能の向上が期待されているが，今回の検討ではこれらの効果は認めなかった。現在，我々の施設では人工内耳の適応決定は日本耳鼻咽喉科学会の適応ガイドラインを遵守しており，この基準により選ばれた人工内耳装用者においては補聴器併用の効果が乏しいと言える。この基準では人工内耳の適応とならずに補聴器装用をしている患者に対して人工内耳装用を行った場合には，対側耳補聴器装用による併用効果がみられる可能性はあり，これは今後の検討課題であろう。現在の基準で適応決定を行った場合には聴取成績の向上が補聴器併用では見られなかったことから，この基準を遵守してなおかつ聴取能を更に向上させるためには，人工内耳機器の改良，新たなコード化法の開発などのデバイス機能の改善による聴取能の向上，または補聴器より以上に対側耳を積極的に活用する両側人工内耳装用などが必要と考えられる。

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Benefits of dichotic hearing under silent and noisy conditions in pediatric cochlear implant users wearing a hearing aid for the opposite ear

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Auditory localization and the cocktail party effect are examples of the advantages of dichotic hearing. Bilateral cochlear implants have already begun to be used in many countries, and been reported to enable good performances of the patients in speech recognition tasks. In Japan, cochlear implant users generally use a hearing aid for the opposite ear, because the benefits of use of a hearing aid for contralateral ears have been reported by many studies from other countries. We examined the benefits of use of a hearing aid for the contralateral ear in Japanese patients under silent and noisy conditions. Under the silent condition, there was no significant difference in the hearing ability between subjects with unilateral cochlear implants and those with unilateral cochlear implants with a hearing aid for the contralateral ear. According to further analyses taking into consideration other components, such as the period of wearing, hearing ability of the contralateral ear and that of the cochlear implant, no benefits were observed. Under the noisy condition also, no significant difference in the hearing ability was observed between the two groups. Some causes are suggested for the absence of the benefits of contralateral hearing aids in the present study: stringent criteria for the fitting of cochlear implants; the small number of cases in the study; the phonemic features of Japanese. To improve the hearing ability in cochlear implant users, we suggest aggressive use of the contralateral ears, using bilateral cochlear implants if necessary.

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ORIGINAL RESEARCH–PEDIATRIC OTOLARYNGOLOGY

Etiology and one-year follow-up results of hearing loss identified by screening of newborn hearing in Japan

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Sponsorships or competing interests that may be relevant to content are disclosed at the end of this article.

ABSTRACT

OBJECTIVE: To evaluate the incidence of newborn hearing loss in a Japanese population and to elucidate etiological factors and one-year prognosis.

STUDY DESIGN: Screening of newborn hearing.

SETTING: Children's tertiary referral center.

SUBJECTS AND METHODS: Between 1999 and 2008, 101,912 newborn infants were screened, with 693 infants (0.68%) referred. Etiology investigation included CT, detection of cytomegalovirus (CMV) DNA, and connexin 26 mutation.

RESULTS: Abnormal results (auditory brainstem response [ABR] threshold ≥ 35 normal hearing level [dB nHL] in either side) were observed in 312 infants (0.31%), and 133 subjects (0.13%) with ABR thresholds ≥ 50 dB nHL on both sides were classified into the habilitation group. In this group, inner ear/internal auditory meatus anomalies were detected in 20 of 121 subjects (17%) tested, middle/external ear anomalies in 14 of 121 subjects (12%), CMV DNA in 13 of 77 subjects (17%), and connexin 26 mutation in 28 of 89 subjects (31%). In 68 subjects undergoing all three investigations (CT, CMV, and connexin 26), 41 (60%) had positive results in at least one test. With inclusion of otitis media with effusion and perinatal problems, this rate amounted to 78% (53 subjects). Of the 97 infants in the habilitation group successfully followed up to one year, 36 (37%) showed a threshold change of 20 dB or more in either ear: 11 (11%) progression and 25 (26%) improvement, and 15 infants (15%) were reclassified into a less severe classification.

CONCLUSION: Considering that 26 percent of infants with bilateral moderate to severe hearing loss showed improvement in one year, habilitation protocols, especially very early cochlear implantation within one year of birth, should be reconsidered.

In Japan, there are about 1,060,000 newborns per year, of whom 62 percent undergo newborn hearing screening (NHS). The Saitama prefecture, situated to the north of and adjacent to the capital, Tokyo, has a population of about seven million, and has about 66,000 newborns every year. Saitama Children's Medical Center has carried out highly advanced health care for patients referred from all parts of the prefecture, and even from adjacent regions. For example, the automated audiometry brainstem response (AABR) test was first introduced to Japan in 1997 at our center. In the Saitama prefecture, the rate of NHS has been approximately 72.5 percent.

Despite the large number of newborns screened in Japan, there have been few large-scale reports on NHS,[1] in part because there are small referral institutions scattered around the country. Studies on definite etiologies and follow-up results have been even rarer; however, this has also been the case with international literature. This brief report summarizes our 10-year experience with NHS and presents the etiologies revealed and changes in hearing acuity observed in a one-year follow-up study at our center.

Methods

Subjects

Between 1999 and 2008, a total of 101,912 newborn infants were screened for hearing loss by AABRs and/or otoacoustic emission (OAE) responses (transient evoked OAE or distortion product OAE) in the 36 gynecological hospitals and clinics affiliated with Saitama Children's Medical Center. Our group has been covering almost one fifth of the population in the Saitama prefecture, in a rural part of the prefecture where few people of foreign origin, including non-Japanese Asians, reside. The 693 infants (0.68%), 396 males and 297 females, who failed the screening ("refer" results) were referred to our center for further evaluation of hearing acuity. These infants comprised the subjects of the present study. All of the referred subjects were of Japanese descent. The average age of their first visit to our center was 19 days (range 5 days to 8 months).

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Table 1
Results of hearing evaluation with ABR (of the 101,912 newborn infants initially screened, 693 [0.68%] were referred)

Group	ABR thresholds	Number	Incidence
Not abnormal	Bilaterally <35 dB	381	0.38%
Observation	≥35 dB in either ear and <50 dB in either ear	179 (unilateral: 154; bilateral: 25)	0.18%
Habilitation	Bilaterally ≥50 dB	133	0.13%

ABR, auditory brainstem response.

Examination of Hearing

On examination, the external auditory canal and ear drum were inspected and cleaned. Evaluation of hearing was done exclusively by ABRs with click stimuli. Response thresholds less than 35 dB normal hearing level (nHL) on both sides were classified as "not abnormal" and excluded from the follow-up study. Response thresholds more than or equal to 50 dB nHL on both sides were classified as the habilitation group. The others comprised the observation group.

Evaluation of Etiology

In subjects belonging to the habilitation group whose parents provided written permission, further tests were performed to elucidate the etiology. Tests included high-resolution computed tomographic scans (HRCTs, slice thickness 1.0 mm), screening for cytomegalovirus (CMV) DNA in the umbilical cord blood, and screening for connexin 26 (GJB2) mutation. HRCTs were checked by both otolaryngologists and radiologists. Both homozygous and heterozygous mutations of GJB2 were classified as positive. There was no patient selection protocol in regard to the performance of these tests, which may cause a bias in the incidence of etiology results. Family history of significant perinatal problems was checked by having parents complete questionnaires (open-ended questions). These procedures were approved by the ethics committee of our center.

Follow-up Study of Hearing Impairment

Subjects classified into the habilitation group were followed up with ABRs in our center up to one year of age. Hearing aids were fitted at three to four months if the threshold recovery was not observed. Normally, instruction to the parents was given at our center and habilitation was started at nearby institutions. Those classified into the observation group were followed up with conditioned orientation response (COR) audiometry either at nearby otolaryngologic clinics or at our center up to one year of age.

Results

Table 1 describes the results of the hearing examination. Abnormal results were observed in 312 infants (0.31% of initially screened babies). Table 2 shows positive rates in causative factors for hearing loss in the habilitation group. HRCTs of the temporal bone were obtained in 121 infants in the habilitation group to detect anomalies of the external auditory canal, the middle ear including ossicles, inner ear, and the internal auditory meatus (IAM). Inner ear/IAM anomalies were suggestive of sensorineural hearing impairment, and middle/external ear anomalies suggested conductive hearing loss. Two infants had both middle and inner ear anomalies, suggesting mixed hearing loss. Opacity of the middle ear suggested otitis media with effusion (OME), possibly augmenting the hearing impairment. As a routine procedure, apparent intratympanic fluid was cleared beforehand, but it was not unusual with a one-month-old baby for precise evaluation of the ear drums to be difficult. Therefore, we chose opacity with CT as a better indicator for middle ear fluid. As for perinatal problems (with overlapping problems permitted), 25 (19%) had apparent positive records: nine cranial anomalies, eight congenital viral infections (7 CMV, 1 rubella), 13 neonatal asphyxia, two jaundice, and seven very low birth weight (< 1500 g). Thirty-one subjects (23%) had positive family history of hearing loss. The combination of connexin 26 mutation and inner ear/IAM anomaly was observed in three infants. Of

Table 2
Positive rates in causative factors for hearing loss in habilitation group (overlapping factors are permitted)

Abnormality	No. tested	No. positive	Proportion*
Connexin 26 mutation	89	28	31%
CMV DNA	77	13	17%
HRCT findings			
Inner ear/IAM anomaly	121	20	17%
Middle/external ear anomaly	121	14	12%
Middle ear opacity	121	47	39%

CMV, cytomegalovirus, *HRCT,* high-resolution computed tomography; *IAM,* internal auditory meatus.
*Proportion = number positive/number tested.

Table 3
Causative factors for hearing loss in subjects who underwent all three tests (n = 68) in search of the etiology in the habilitation group

Abnormality	No. positive	Proportion
Connexin 26 mutation	19	28%
CMV DNA	9	13%
Anomaly identified by CT	16	24%
Overlap (connexin 26 and anomaly)	3	4%
Positive for at least one abnormality	41	60%

CMV, cytomegalovirus.

infants with positive perinatal problems, three had inner ear/IAM anomalies and one had connexin 26 mutation.

Results for all three tests (i.e., HRCT, CMV DNA, and connexin 26 mutation) were available in 68 subjects. Table 3 depicts causative factors for hearing loss in subjects (n = 68) in the habilitation group who underwent all three tests in search of the etiology. The proportion of subjects found positive in any test was 60 percent. When middle ear opacity in CT suggesting OME and positive history of perinatal problems were also accounted for, this rate amounted to 78 percent (53 of 68 subjects).

Of 133 infants classified into the habilitation group, 97 were followed with ABRs up to one year of age. Table 4 summarizes the one-year follow-up results. Overall, 37 percent of subjects demonstrated a notable change in hearing acuity, and 15 of 97 infants (15%) in the habilitation group were reclassified into the observation group at the age of one year. Of the 25 infants with improvement, nine had bilateral profound hearing loss (≥ 90 dB) initially, among whom six had shown no responses in ABRs (thresholds > 100 dB). With regard to the causative factors among these 25 subjects, two had connexin 26 mutation (heterozygotes), three had CMV DNA, eight had middle ear opacity in CT, seven had middle/inner ear anomaly, and three had perinatal problems. In ABRs, eight showed shortening of wave I latency indicative of amelioration of middle ear problems, and three showed shortening of the wave I-V interval, indicating amelioration of retrocochlear problems among these subjects. On the other hand, among 11 subjects with progressive hearing loss, two had connexin 26 mutation, two had CMV DNA, three had middle ear opacity in CT, and three had perinatal problems.

Of the 179 infants classified into the observation group, 110 were followed with CORs up to one year of age. Of those, one subject (0.9%) showed remarkable progression and was reclassified into the habilitation group and recruited for intensive habilitation programs.

Discussion

Here we present the results of a large-scale retrospective study of follow-up of newborn hearing in Asia. Considering the more detailed hearing evaluation currently performed in Western countries, as well as racial and climate differences, the authors do not know whether the data can presently be applied directly to the West. With the expectation that similar studies will be conducted in the West, we discuss the results of our present, preceding research below.

The rate of newborn hearing loss (0.31%) and bilateral moderate to severe hearing loss (0.13%) corresponded with results from previous reports.[2-6] With regard to the cause of congenital hearing loss other than anomalies, CMV infection and connexin 26 mutation have attracted attention. It has been shown that asymptomatic CMV infection causes hearing loss in six to 23 percent of cases, symptomatic infection causes hearing loss in 22 to 65 percent of cases,[7,8] and that 15 to 20 percent of bilateral moderate to profound sensorineural hearing loss could be attributable to congenital CMV infection in a Western population.[9] Our results (17%) on bilateral moderate to severe hearing loss in Japan correspond to results of these previous reports, despite the regional and racial differences. Connexin 26 mutation has been demonstrated to be responsible for 21 to 39 percent of cases of genetic nonsyndromic childhood deafness in a Western population.[10-12] Our results (31%) also fell within this reported range.

Table 4
ABR threshold changes (either ear) in habilitation group during one-year follow-up (n = 97)

Course	Threshold change	Number	Proportion
Improvement	20-30 dB, inclusively*	11	11%
	>30 dB	14	14%
	Subtotal	25	26%
No change	<20 dB	61	63%
Progression	20-30 dB, inclusively*	8	8%
	>30 dB	3	3%
	Subtotal	11	11%

*≥20 dB and ≤30 dB.

The most important finding demonstrated by the present study was the rate of change (37%) in hearing acuity within one year of birth. This was much higher than expected and should not be confused with a lack of short time reproducibility of the screening test itself.[13] Above all, the fact that not a small proportion of infants (26%) with bilateral moderate to severe hearing loss showed improvement at one year, rendering 15 percent of subjects to a less severe classification, was surprising. From the distribution of causative factors such as CMV, connexin 26, and middle ear opacity, no apparent predictor for improvement or progression could be suggested. Undeniably, the fact that a high proportion of subjects did not undergo all three tests (68 of 133; 51%) might have produced bias. This was probably because more than a few parents chose to leave the etiology unknown (especially in cases of persistent viral infection and genetic abnormality that could raise unnecessary prejudice) when no effective treatment could be provided. However, it should be noted that no intentional selection was made for entry into these tests.

The ABR study also failed to suggest known factors (e.g., resolution of OME or auditory neuropathy) as a major cause of improved hearing. Moreover, the ability to evaluate etiology from ABR results is limited because shortening of wave I may reflect only the maturity of brainstem pathways rather than auditory neuropathy, and because OME alone cannot produce profound hearing loss, leaving the possibility that it simply accompanies cochlear hearing loss. Even if the subjects with opacity in CT are excluded, 18 percent (17 of 97) showed unexplained recovery, which is not an ignorable proportion. This necessitates a follow-up study of all infants with bilateral hearing loss, regardless of the causative factors for each infant's hearing loss. Spontaneous hearing threshold recovery in sensorineural hearing loss has been demonstrated even in children around one year of age, as proven by behavioral audiometry as well as ABRs.[14] In our patients, not an ignorable number (9 infants; 9.3%) demonstrated improvement from bilateral profound hearing loss (≥ 90 dB), for which cochlear implantation could have been a choice. If the natural courses shown in the present study are taken into account, habilitation protocols may have to be reconsidered, and cochlear implantation, especially in infants less than one year of age,[15,16] remains controversial.

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Author Contributions

Nodoka Adachi, clinical follow-up and data collection; **Ken Ito,** data analysis and supervision; **Hideaki Sakata,** clinical follow-up and data collection; **Tatsuya Yamasoba,** advice on study configuration.

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Risk Factors for Hearing Loss After Pediatric Meningitis in Japan

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Objectives: We sought to identify predictors for hearing loss in Japanese children with meningitis.

Methods: We analyzed 155 cases of pediatric meningitis without other entities causing hearing loss in children admitted to Saitama Children's Medical Center between 1990 and 2005 for potential risk factors for hearing loss, using multiple logistic regression. Auditory brain stem response tests were performed to evaluate hearing loss.

Results: Of 155 children, 35 (23%) developed hearing loss (21 unilaterally and 14 bilaterally). Profound hearing loss (greater than 90 dB normal hearing level) occurred in 15 patients (9.7%; 4 unilaterally and 11 bilaterally). Of 112 patients with positive cerebrospinal fluid cultures, 27 (24%) developed hearing loss and 13 (12%) showed profound loss. Of 22 patients with *Streptococcus pneumoniae* meningitis, 11 (50%) developed hearing loss and 7 (32%) showed profound loss. Of 54 patients with *Haemophilus influenzae* meningitis, 11 (20%) developed hearing loss and 4 (7.4%) showed profound loss. High serum C-reactive protein levels and cerebrospinal fluid cultures positive for *Streptococcus pneumoniae* were identified as significant risk factors for hearing loss.

Conclusions: A high serum C-reactive protein level was first identified as a risk factor for hearing impairment after pediatric meningitis.

Key Words: Asian population, child, complication, hearing loss, meningitis, risk factor.

INTRODUCTION

Hearing impairment is one of the most important sequelae of pediatric meningitis. Although the incidence has been reported principally in white populations,[1-8] the rates vary considerably among reports, and possible differences among races have not been elucidated. Moreover, only a few articles have reported risk factors or predictors for hearing impairment in pediatric meningitis.[2,3,6,7] Thus, the issue remains controversial.

In this study, we report the first review of a large consecutive series of children with meningitis in an Asian population. We also conducted statistical analyses to elucidate risk factors for hearing impairment in our series.

PATIENTS AND METHODS

Between 1990 and 2005, a total of 192 children (up to 15 years) were admitted to Saitama Children's Medical Center with a diagnosis of meningitis. Of these patients, 155 children without other causes of hearing loss, eg, inner ear anomalies, who underwent auditory brain stem response (ABR) testing during their hospital stay comprised the study subjects. Patient ages ranged from 1 day to 13 years (median, 8 months), and there were 84 male and 71 female patients. The medical records were reviewed retrospectively. Treatment was directed by the pediatricians. The standard treatment comprised administration of antibiotics (penicillins or cephems) along with mannitol or glycerol. Antiviral drugs, gamma globulins, and steroids were sometimes used, but aminoglycosides were administered only exceptionally.

Hearing loss was defined as threshold elevation of 40 dB normal hearing level or more in either ear, determined by click-evoked ABRs. This value of 40 dB was chosen to exclude mild hearing loss, which leaves little potential handicap. Candidate predictors for hearing loss, chosen after previous reports, included patient age, gender, positive cerebrospinal fluid (CSF) culture, CSF glucose level, CSF protein level, CSF white blood cell (WBC) count, serum C-reactive protein (CRP) level, and development of seizures. In an additional analysis to evaluate the influence of bacterial strains, CSF cultures positive for *Streptococcus pneumoniae* and for *Haemophilus influenzae* replaced the parameter of an overall positive CSF culture. Table 1 shows the patient demo-

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TABLE 1. PATIENT DEMOGRAPHICS

Parameter	*Values*
Age	1 d to 13 y (median, 8 mo)
Gender	84 male, 71 female
Serum CRP (mg/dL)	0.0 to 38.2 (mean, 11.1)
CSF glucose (mg/dL)	0.0 to 151 (mean, 37.3)
CSF protein (mg/dL)	6.1 to 3,200 (mean, 277)
CSF WBCs (cells/μL)	8 to 308,000 (mean, 11,600)
Seizure	29/155 (18.7%)
Positive CSF culture	112/152 (73.7%)
Positive CSF culture for *Streptococcus pneumoniae*	22/152 (14.5%)
Positive CSF culture for *Haemophilus influenzae*	54/152 (35.5%)

CRP — C-reactive protein; CSF — cerebrospinal fluid; WBCs — white blood cells.

graphics. Possible risk factors for hearing impairment were assessed with multiple logistic regression analysis.[9]

RESULTS

Of the 155 children, 35 (23%) developed hearing loss (21 unilaterally and 14 bilaterally). Profound hearing loss (greater than 90 dB normal hearing level) occurred in 15 patients (9.7%; 4 unilaterally and 11 bilaterally). Bacteria were isolated from the CSF cultures in 112 patients. Of these, *S pneumoniae* was isolated in 22 patients and *H influenzae* in 54. Of 112 patients with positive CSF cultures, 27 (24%) developed hearing loss and 13 (12%) showed profound loss. Of 22 patients with *S pneumoniae* meningitis, 11 (50%) developed hearing loss and 7 (32%) showed profound loss. Of 54 patients with *H influenzae* meningitis, 11 (20%) developed hearing loss and 4 (7.4%) showed profound loss. Twenty-nine children developed seizures. Other apparent neurologic complications, eg, involvement of cranial nerves other than the eighth nerve and paralyses of the extremities, were not noted.

Table 2 shows the results of multiple logistic regression analyses. Of 8 candidate factors, only a high serum CRP level was identified as a significant risk factor ($p < 0.05$), for both hearing loss and profound hearing loss. To further analyze the effect of bacterial strains, we replaced the parameter of overall positive CSF culture with that of CSF cultures positive for *S pneumoniae* and for *H influenzae* (Table 3). For overall hearing loss, a CSF culture positive for *S pneumoniae* was determined as the most significant risk factor, whereas the serum CRP level failed to reach significance. However, for profound hearing loss, the serum CRP level regained significance. A CSF culture positive for *S pneumoniae* followed, but failed to reach significance for profound loss. On the basis of these results, a high serum CRP level and a CSF culture positive for *S pneumoniae* were considered important risk factors for developing hearing loss due to pediatric meningitis.

TABLE 2. LOGISTIC REGRESSION USING PARAMETER OF OVERALL POSITIVE CSF CULTURE

Parameter	*Overall Hearing Loss*	*Profound Hearing Loss*
Age (logarithmic)	0.2239	0.8357
Gender	0.1321	0.3257
Serum CRP	0.0470*	0.0216*
CSF glucose	0.2155	0.1746
CSF protein	0.3851	0.9765
CSF WBCs	0.5868	0.6318
Seizure	0.1984	0.4202
Positive CSF culture	0.6142	0.4060

Data are p values.
*Significance ($p < 0.05$).

TABLE 3. LOGISTIC REGRESSION USING PARAMETERS OF POSITIVE CSF CULTURE FOR *STREPTOCOCCUS PNEUMONIAE* AND FOR *HAEMOPHILUS INFLUENZAE* INSTEAD OF OVERALL POSITIVE CSF CULTURE

Parameter	*Overall Hearing Loss*	*Profound Hearing Loss*
Age (logarithmic)	0.2990	0.7204
Gender	0.1879	0.5142
Serum CRP	0.0755	0.0442*
CSF glucose	0.1687	0.1810
CSF protein	0.3211	0.9831
CSF WBCs	0.7218	0.7085
Seizure	0.0674	0.2044
Positive CSF culture for *Streptococcus pneumoniae*	0.0284*	0.0528
Positive CSF culture for *Haemophilus influenzae*	0.5390	0.7652

Data are p values.
*Significance ($p < 0.05$).

DISCUSSION

The incidence of hearing loss in pediatric meningitis has been reported principally in Western populations. The overall incidence of hearing impairment has ranged from 14% to 29%,[1,2,5] and that in bacterial meningitis has ranged from 7% to 31%.[1-4,6,7] The incidences reported in the present report — 23% for overall meningitis and 24% for bacterial meningitis — correspond to those in previous reports, despite racial and climate differences. The incidence of hearing loss has been greater in bacterial meningitis than in overall meningitis,[1,2] as was also confirmed by the present findings.

Streptococcus pneumoniae, *H influenzae*, and *Neisseria meningitides* are the 3 most important pathogens for pediatric meningitis. The incidence of

hearing impairment due to *S pneumoniae* meningitis ranges from 20% to 52%,[2-4,6,8] that due to *H influenzae* meningitis ranges from 22% to 41%,[1,2,4] and that due to *N meningitidis* meningitis ranges from 4% to 24%.[2-4,6] The order of preponderance in hearing involvement has remained consistent, including in the present report: *S pneumoniae* first, then *H influenzae* meningitis, and then *N meningitidis* meningitis. Other bacteria cultured in the present study included *Escherichia coli*, methicillin-resistant *Staphylococcus aureus*, other *Streptococcus* species, *Neisseria* species, *Listeria*, etc, but the small number of affected patients prevented these from being nominated as risk factors. In Japan, vaccination for *S pneumoniae* is not yet popular, and that for *H influenzae* type B has only recently been approved. Encouraging vaccination for these 2 bacteria will be important in preventing hearing loss caused by meningitis in our country.

Several risk factors or predictors for hearing impairment in pediatric meningitis have been proposed,[2,3,6-8] but the significance of risk factors has varied greatly among reports. The most common factor was a low CSF glucose level,[2,3,6,7] followed by a CSF culture positive for *S pneumoniae*.[2,6] In the present study, a CSF culture positive for *S pneumoniae* was a significant risk factor, but a low CSF glucose level failed to reach significance. Instead, a high serum CRP level was recognized as another significant risk factor — a finding not reported in previous articles. This outcome is not surprising, since the serum CRP level corresponds to the severity of overall inflammation. In our search of the English-language literature, there were no reports describing risk factors for neurologic dysfunction other than hearing impairment (eg, other cranial nerve disorders) as sequelae after meningitis.

CONCLUSIONS

Logistic regression analyses of our consecutive series of Japanese children with meningitis identified a high serum CRP level and a CSF culture positive for *S pneumoniae* as risk factors for hearing impairment after pediatric meningitis.

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Cochlear implantation in a patient with atypical Cogan's syndrome complicated with hypertrophic cranial pachymeningitis

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Abstract

A 55-year-old woman had bilateral sensorineural hearing loss (SNHL), vertigo, uveitis, and aortitis associated with Cogan's syndrome (CS). She had a history of listeria meningitis and hypertrophic cranial pachymeningitis (HCP), both of which were considered to be related to SNHL progression. She developed bilateral profound deafness within 1 year, despite medical treatment with corticosteroids and methotrexate (MTX). She underwent cochlear implantation (CI) of the left ear. Although the left and right basal turns of the cochleae were ossified, all electrodes were successfully inserted, and subsequently the inferior segment of the basal turn was drilled out. The patient did not have any postoperative complications and showed good speech perception.

Keywords: Cogan's syndrome; Hypertrophic cranial pachymeningitis; Cochlear implantation; Bilateral profound sensorineural hearing loss

1. Introduction

Cogan's syndrome (CS) is a rare disease, and an autoimmune process has been postulated to play a crucial role in its etiology. CS generally shows ear involvement (progressive bilateral sensorineural hearing loss (SNHL), tinnitus, and vertigo), ocular involvement (most commonly non-syphilitic interstitial keratitis), and variable systemic symptoms. The central nervous system is more commonly involved in CS, but hypertrophic cranial pachymeningitis (HCP), which may contribute the progression of SNHL, is rarely encountered. To date, there is only 1 case of CS complicated with HCP [1]. Fortunately, conservative steroid therapy improved SNHL in the patient.

Audiovestibular dysfunction of CS usually requires high systemic doses of corticosteroids and immunosuppressive drugs such as methotrexate (MTX) and cyclophosphamide [2]. In spite of proper treatment, 50–80% of patients develop irreversible bilateral profound SNHL [3–5]. Prognosis of cochlear implantation (CI) is generally favorable [6]. This report describes CI in a patient with a rare case of CS complicated with HCP. The patient showed good speech perception after CI.

2. Case report

A 55-year-old woman complained of sudden right SNHL in April 2005. Corticosteroids failed to improve the SNHL, and the patient developed profound deafness in her right ear (Fig. 1a). After 4 months, left SNHL and vertigo developed. Corticosteroids failed to alleviate these symptoms, and she developed profound deafness in her left ear (Fig. 1b). After 1 year, she developed conjunctival congestion and blurred vision and was diagnosed with bilateral conjunctivitis and uveitis, but she did not show headache or other cranial nerve

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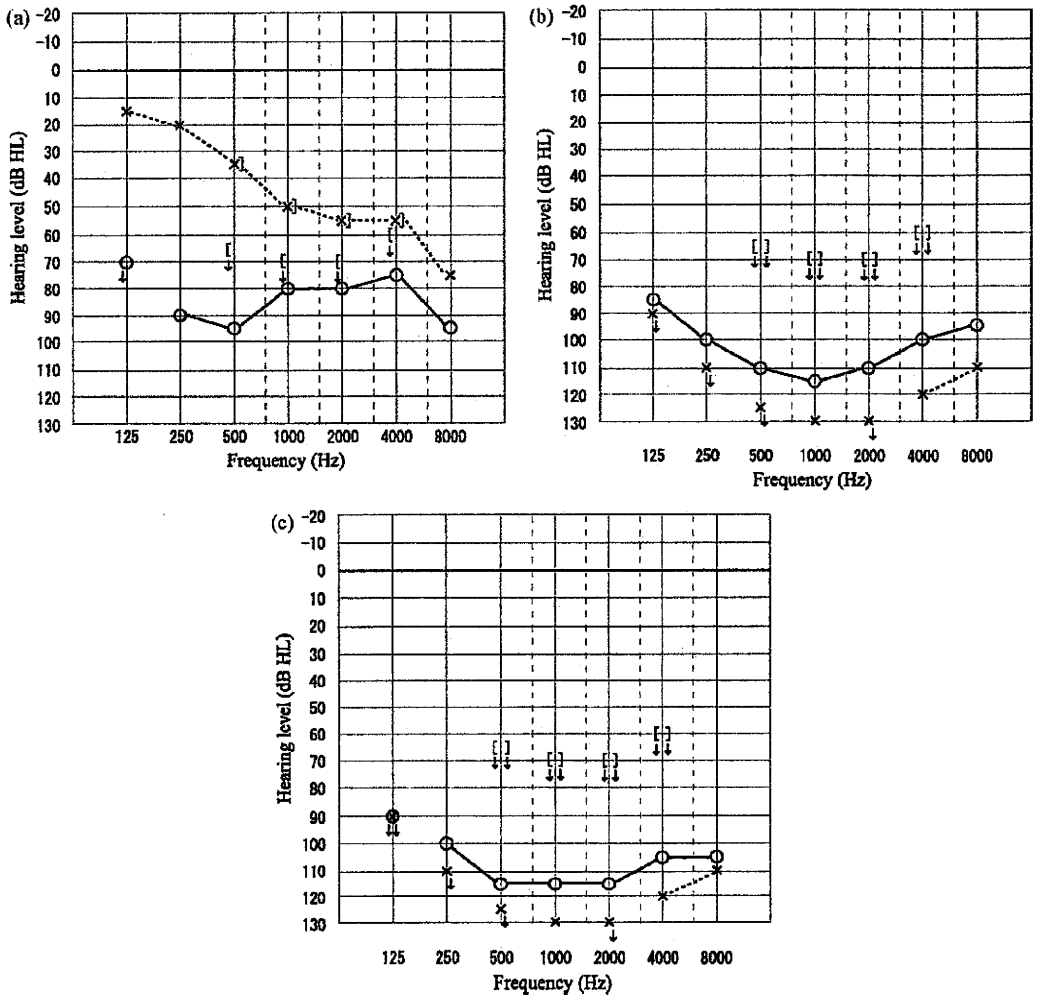

Fig. 1. (a) Audiogram at the time of developing sudden right SNHL in April 2005. (b) Audiogram at the time of developing left SNHL in April 2005. (c) Audiogram immediately after meningitis.

signs. In September 2006, she was referred to the Department of Internal Medicine at the University of Tokyo Hospital because she was suspected to have an autoimmune disease. The leukocyte count (3500/μl) was not elevated, but the erythrocyte sedimentation rate (75 mm/h) and C-reactive protein level (3.07 mg/dl) were elevated. The patient tested negative for antinuclear antibodies, myeloperoxidase-anti-eutrophil cytoplasmic antibody (MPO-ANCA), proteinase 3-ANCA, and syphilis. She was diagnosed with aortitis by F-18 fluoro-2-D-deoxyglucose (FDG) positron emission tomography (PET). The findings suggested CS. Magnetic resonance imaging (MRI) revealed meningioma at the left parietal convexity and gadolinium-enhanced T1-weighted MRI showed linear enhancement of the dura mater from the frontal lobe to the falx and along the internal auditory meatus; this finding suggests thickening of the dura mater due to HCP. The meningioma was resected, and open biopsy was performed from the dura mater at the falx. The dura mater showed intense lymphocyte infiltration, as is observed in the case of HCP. She was given a high-dose of corticosteroids and MTX, but the extent of linear enhancement of the dura mater was the same before and after as observed on the gadolinium-enhanced T1-weighted MRI (Fig. 2).

The patient was referred to our department for CI evaluation. Pure-tone audiometry showed bilateral profound SNHL similar to that depicted in Fig. 1b. High resolution computed tomography (HRCT) revealed mild ossification of the basal turns of bilateral cochleae. MRI revealed reduction of the fluid content in the basal turns of bilateral cochleae. However, we did not perform CI at this time point because she was very afraid of the disadvantage induced by implantation of an artificial device. In June 2007, she developed listeria meningitis, probably because of immunosuppression induced by long-term administration of corticosteroids and MTX. Pure-tone audiometry immediately after meningitis showed slight progression of low tone SNHL in the right ear (Fig. 1c). HRCT performed after 1 year revealed progression of ossification in the basal turns of bilateral cochleae (Fig. 3). Gadolinium-enhanced T1-weighted MRI showed dural enhancement to the same extent as was identified in the previous MRI. The

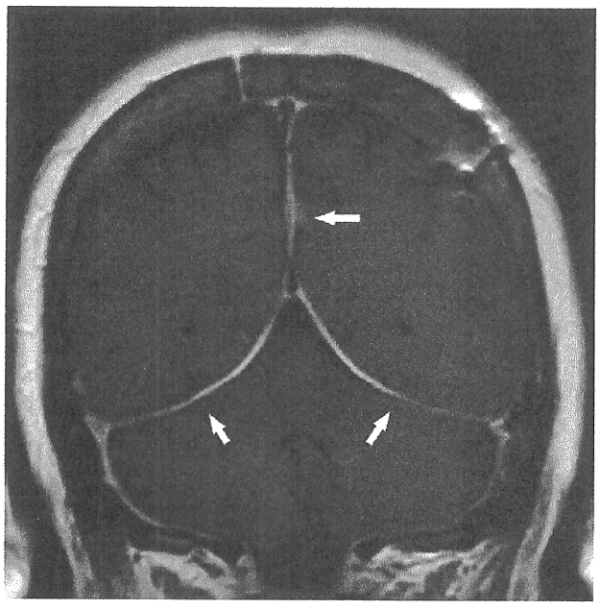

Fig. 2. Gadolinium-enhanced T1-weighted magnetic resonance imaging performed after treatment revealed pachymeningeal thickening and linear enhancement of the dura mater (arrow).

preoperative 3D fast imaging employing steady state acquisition (FIESTA) MRI revealed reduction of fluid intensity in the basal turns in both ears; this observation suggests obliteration due to ossification and the presence of fibrous tissue (Fig. 4). Therefore, we strongly recommended CI to her and decided to perform CI under her consent because further ossification would make it impossible to insert electrodes into the cochlea.

During the preoperative examination, pure-tone audiometry showed bilateral profound SNHL identical to that depicted in Fig. 1c, and results of speech audiometry showed lack of identification ability in both ears. A promontory stimulation test showed a positive response in both ears. Distortion product otoacoustic emissions (DPOAE) produced no response in both ears. The vestibular function was severely affected; vestibular evoked myogenic potential (VEMP) was absent, and the caloric test evoked no nystagmus.

We decided to perform CI to the left ear because reduction of fluid intensity in MRI was slighter in the left ear than that in the right ear. The patient underwent CI of the left ear with a Nucleus CI24R device. Although the left and right basal turns of the cochleae were ossified, all electrodes were successfully inserted following drilling out the inferior segment of the basal turn. The patient's postoperative course was uneventful. Complications including a local flap ischemia and worsening of symptoms related to CS and HCP, such as cranial nerve palsy and headache, were not observed. At the 12-month postoperative evaluation, perception score for a monosyllable, word, and sentence under the hearing-only condition was 80%, 78%, and 79%, respectively. These scores indicated that the patient had good speech perception.

3. Discussion

We showed that a patient with CS complicated with HCP successfully underwent CI and showed good speech perception after the operation.

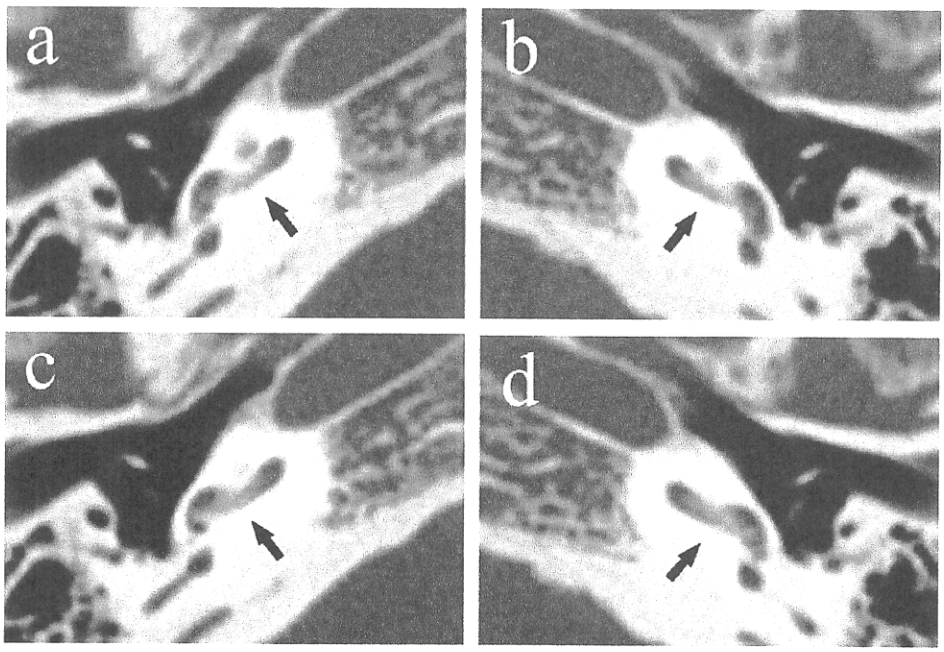

Fig. 3. Preoperative high resolution computed tomography revealed progression of ossification of the basal turn in the right (a and c) and left (b and d) ears (arrow). Figure c and d are one slice below than figure a and b.

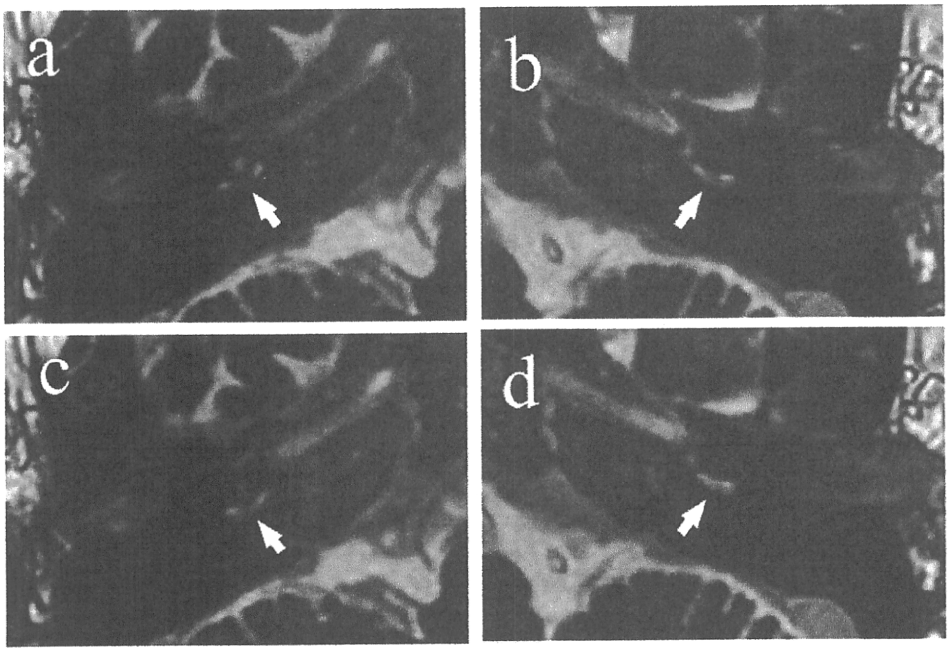

Fig. 4. Preoperative 3D fast imaging employing steady state acquisition MRI revealed reduction in fluid intensity in the basal turns in the right (a and c) and left (b and d) ears (arrow). Figure c and d are one slice below than figure a and b.

The diagnosis of typical CS was made on the basis of non-syphilitic interstitial keratitis and acute-onset SNHL [7]. CS is commonly associated with aortitis and aortic insufficiency [8]. Atypical CS is diagnosed when other inflammatory eye diseases, such as conjunctivitis, uveitis, episcleritis, iritis, and retinal vasculitis, develop, or when the interval between the onset of ophthalmologic symptoms and SNHL is $\geq$2 years [9].

The differential diagnosis of CS includes congenital syphilis, Vogt–Koyanagi–Harada syndrome, Wegener's granulomatosis, and rheumatoid arthritis. To diagnose CS, a negative result is required on serologic evaluation for syphilis. No tests can confirm the diagnosis; however, the acute-phase reactants of inflammation, including leukocyte count, erythrocyte sedimentation rate, and C-reactive protein level, were reported to be elevated in CS [10].

In our patient, acute-phase reactants, except leukocyte count, were elevated, whereas tests for antinuclear antibodies, MPO-ANCA, proteinase 3-ANCA, and syphilis gave negative results. These results excluded diseases other than CS. Although the interval between the onset of ophthalmologic symptoms and SNHL was $\leq$2 years, our patient was diagnosed with atypical CS because her ocular involvement presented with conjunctivitis and uveitis and not interstitial keratitis.

In CS, several abnormalities are detected during radiologic investigations. HRCT is useful for detecting intracochlear bony obliteration, whereas MRI is useful for detecting intracochlear soft-tissue obliteration and other central nervous lesions.

In our case, preoperative HRCT revealed bony obliteration in the basal turns of both ears. The preoperative T2-weighted MRI demonstrated reduction in fluid intensity in the basal turns in both ears; this finding suggests obliteration due to ossification and the presence of fibrous tissue. CS alone can cause intracochlear bony obliteration [11]. Listeria meningitis may also have influenced the acute progression of cochlear ossification.

Unexpectedly, in our case, gadolinium-enhanced T1-weighted MRI revealed linear enhancement of the dura mater, which suggested thickening of the dura mater and revealed complication with HCP.

HCP is a rare form of a fibrosing chronic inflammatory process, characterized by marked thickening of intracranial dura mater. Fibrous entrapment and ischemic damage of neurologic structures can cause various symptoms, including headache, decreased vision, and bilateral progressive SNHL [12]. Although HCP may cause retrocochlear hearing loss by entrapment of the eighth nerve in the internal auditory canal, the presence of sound sensation with promontory stimulation test suggested that hearing loss in our patient was due mainly to cochlear damage. It is well known that eighth nerve lesions such as acoustic neurinoma frequently cause cochlear damage as a result of the ischemia caused by the tumor and/or biochemical disturbances in the inner ear fluid. Meningitis is also known to cause cochlear damage due to the direct spread of infection from the subarachnoid space to the inner ear (suppurative labyrinthitis), but other mechanisms, such as toxic or serous labyrinthitis and secondary ischemic damage, are also proposed. Although the mechanisms are difficult to prove, secondary ischemia or biochemical disturbances in the inner ear fluid due to fibrous entrapment may account for cochlear damage in our patient. HCP generally shows a good

response to corticosteroids [13]. In our patient, however, an adequate dose of corticosteroids did not help improve linear enhancement of the dura mater; this was inferred by comparing the gadolinium-enhanced T1-weighted MRI obtained before and after treatment.

It has been reported that despite proper treatment, 50–80% of CS patients develop irreversible profound SNHL [3–5]. The prognosis of CI is favorable in CS patients with profound SNHL [5,9]. This is considered because of the following facts. (1) The patients are postlingually deaf. (2) They use hearing aids properly during the progression of SNHL. (3) The period of profound SNHL is relatively short. In accordance with previous reports, our patient showed good speech perception after the operation.

CS can have several kinds of systemic manifestations. However, the complication with HCP has been reported only in 1 case by Togashi et al. [1]. Fortunately, oral corticosteroid therapy improved bilateral SNHL in their patient, and CI was not needed. Our report was the first to describe hearing results after CI in a patient with CS complicated with HCP.

Both CS and HCP are related to an autoimmune disorder and vasculitis syndrome [14]. The major complications after CI are a local flap ischemia caused by insufficient blood supply due to vasculitis, and the worsening of CS and HCP symptoms because of stress consequent to the surgical procedure. Our patient did not show flush, infection, and necrosis at the flap site. Extrusion of the CI device was not observed. Moreover, symptoms suggesting CS and HCP recurrence were not observed after the surgery.

CI dramatically improves speech perception in patients with bilateral profound SNHL due to CS [5,9]. Hence, these patients are good candidates for CI. However, cochlear obliteration often poses a challenge for electrode insertion, requiring modifications of the surgical technique. Pasanisi et al. reported that the case of incomplete bone obliteration of basal turn, undetected on preoperative radiological examination and confined to a narrow portion, required tunneling through the ossified portion. The case of complete ossification of the inferior segment of basal turn required insertion into the scala vestibule [6]. Our patient also showed incomplete ossification of the basal turn, detected during preoperative radiological evaluations. However, we inserted all electrodes into the scala tympani by drilling out the inferior segment of the basal turn. Surgeon should be prepared to manage cochlear obliteration because cochlear obliteration is often confirmed only by intraoperative findings.

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Activation of the Auditory Cortex in a Child with a Cochlear Implant: an Optical Topography Study

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Summary

We measured neural activation of the central auditory system in a prelingually deaf cochlear implant (CI) patient on the day of first fitting. Optical topography was used to assess cortical activity non-invasively while auditory stimuli were presented through the CI. Deoxyhemoglobin (deoxy-Hb) as well as sum of oxyhemoglobin (oxy-Hb) and deoxy-Hb increased in the temporal region during the auditory stimuli, reflecting activation of the temporal cortex. Examination of the central auditory system would benefit the rehabilitation process of the CI.

Introduction

Cochlear implantation (CI) is a well-established treatment for prelingually deaf children. These days CI operation tends to be performed earlier in the patient's life, making the postoperative fitting more difficult. Although neural responses of the peripheral auditory system can be recorded by most of modern CIs, it is still difficult to evaluate the activity of the central auditory system, because functional MRI (fMRI) cannot be used for CI patients and because the parents hesitate to participate in PET studies due to irradiation. Optical topography, or Near-Infrared Optical Spectroscopy (NIRS), is a neuroimaging method which measures brain activations non-invasively. Near infrared light penetrates the tissue to a certain depth and undergoes partial absorption by blood hemoglobin (Hb) along the light path. By continuously measuring the reflected light on the skull, relative changes of the concentration of oxyhemoglobin (oxy-Hb), deoxyhemoglobin (deoxy-Hb), and total hemoglobin (total-Hb) in specific brain region can be monitored, reflecting local brain responses. In this study we used an optical topography system to study activation of the auditory cortex in an infant CI user.

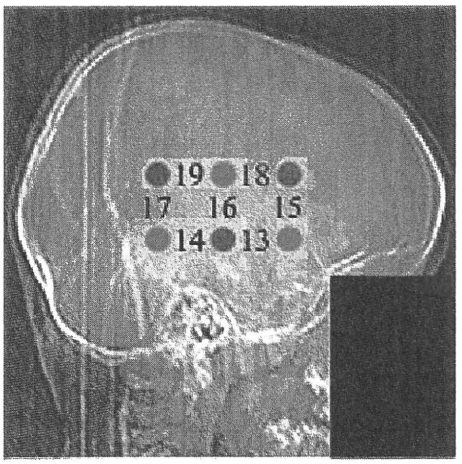

Figure 1. Schematic diagram of the three light emitting (red) and three light receiving (blue) probes on the right temporal region. Numbers indicate the layout of the seven recording sites, called channels.

Materials and Methods

A two-year-old prelingually deaf girl participated in this study. She was diagnosed as severe hearing loss at the age of 12 months, and continued to use hearing aids on both sides. Auditory brainstem response was absent at 105 dBnHL at all times. She had implanted with a Nucleus® 24 system on the left ear at the age of 24 months. This optical topography study was done 13 days after the operation, on the day of first fitting. Before the study, two speech therapists obtained neural response telemetry (NRT) and set T and C levels by evaluating the patient's behavioral responses. A portable CD player was connected to the external input of the speech processor to present auditory stimuli.

Three light emitting and three light receiving probes of an optical topography system (ETG-100, Hitachi, Japan) were attached to the temporal region of the head opposite to the CI (Figure 1). Each probe was kept 3 cm apart by a probe holder. Quantitative changes of oxy-Hb, deoxy-Hb, and total-Hb were recorded at seven different recording sites (channels), which were expected to cover the perisylvian region (Figure 1). Auditory stimuli, which were consisted of spoken words of four syllables, were presented for twenty seconds, and then turned off for twenty seconds. This was repeated for ten times, making the total measurement time for about seven minutes. The patient was tested while seated in a chair playing with toys. Written informed consent was obtained from the parents. The protocol was in compliance with the ethical committee of the institute.

Results

No obvious behavioral change was observed between the stimulated and silent periods. In channel 13, which was located in the anterior-inferior part of the measurement site, oxy-Hb and total-Hb decreased during the auditory stimuli (Figure 2). In