ける結果となった。

福田らによれば，早期発見と合わせて，人工内耳の使用が聴取能の向上に貢献し，その結果として発話明瞭度も高くなり構音獲得が容易となるため，人工内耳装用児は聴覚からの構音獲得が容易であることを報告している[10]。加藤らによれば，人工内耳装用後の言語発達には個人差があり，その要因はいくつか挙げられるが，良好な言語能力と最も相関が強かった要因を語音聴取能と報告し[11]，また，森らは人工内耳を２歳代で装用した症例は聴児と同様の構音発達が見られたと報告している[14]。一方，川崎らは人工内耳の装用が遅れた児において，語彙理解力，構音，言語性知能が２年程度遅れたと報告しており[15]，また早期の人工内耳装用が，語音弁別，発話明瞭度の改善とともに語彙・構文の産生能力の発達につながるとも述べており，今回の結果と一致する結果であると考えられる。

今回の解析により，早期からの補聴器装用により語彙・構文の理解能力は発達するものの，産生能力の発達のためには十分な聴取閾値を確保可能な補聴を早期に行う事が重要である可能性が示され，それぞれの補聴システムの持つ意義を区別して確認することができたことは意義深い結果であると考えられた。ただ，言語発達の観点から人工内耳が望ましい児を早期に判断することは困難であり，今後さらなる研究が必要と思われる。

ま と め

感覚器障害戦略研究のデータを元に，補聴器装用および人工内耳の手術年齢による言語発達の差異について検討を行った。その結果，早期の補聴開始が言語理解に，また早期の人工内耳手術が言語産生に影響するという結果が得られた。従って，早期の聴覚的介入は，良好な日本語言語発達に寄与する可能性が示された。

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Effects of early cochlear implantation and hearing aid on Japanese language development.

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This study was aimed at investigating the relationship between the age at cochlear implantation (CI) and language development using a set of Japanese language assessment test battery, consisting of communication skills testing, vocabulary testing, and syntax testing. Of the 638 hearing-impaired children enrolled, 182 children (83 males and 99 females) with CI were included in this study. They were fitted with their first hearing aids (HA) at 27 months and underwent CI at 42 months, on average. Subjects were classified into four groups according to the age at CI and at the first fitting of HA, as follows: 1) early CI and HA group, 2) early HA and delayed CI group, 3) early CI and delayed HA group, 4) delayed CI and HA group. Our study showed that early cochlear implantation was benefi-

cial for the development of productive vocabulary and syntax, and early fitting of HA was beneficial for the development comprehensive vocabulary and syntax. We concluded that early auditory input is effective for the development of language comprehension, and early development of speech discrimination is effective for the development of language production.

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難聴児における低出生時体重児の占める割合およびその言語発達に関する検討

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要旨：従来より，低出生体重が難聴のリスクファクターであることが報告されているが日本語言語発達に及ぼす影響ついて大規模に検討を行った報告は少ない状況である。本研究では難聴児の出生時体重とその言語発達に関して詳細に検討を行った結果，言語習得期前の高度難聴児627名中に占める低出生体重児は89名（14.2%）であった。通常出生体重難聴児と比較して，低出生体重難聴児では語彙レベルの検査結果には差を認めないものの，構文レベルでは有意に得点が低いことが明らかとなった。また，コミュニケーション能力を評価する質問—応答関係検査では低出生体重児群のほうが有意に低得点であった。また，レーヴン色彩マトリックス検査の得点は有意差を認めなかったが，広汎性発達障害日本自閉症協会評定尺度（現在得点）では有意差を認め，低出生体重難聴児の特徴として通常出生体重の児よりも広汎性発達障害を合併する児が多い可能性が考えられた。

ーキーワードー

難聴，低出生体重，言語発達，発達障害

はじめに

従来より，低出生体重や妊娠中感染症などが難聴のリスクファクターとして報告されている[1-4)]が，それらハイリスク児の日本語言語発達に関しては詳細な検討は行われていない。本研究では，感覚器障害戦略研究（聴覚分野）症例対象研究に参加した児を対象に，ハイリスク児の割合とその日本語言語発達に関して検討を行ったので報告する。なお，感覚器障害戦略研究（聴覚分野）症例対象研究は，平成21年から22年の間，聴覚障害児の日本語言語発達に影響を与える因子を明らかにし，発達を補償する方法を考える目的で行われた研究であり，家族・医療・教育を対象としたアンケートによる背景因子の調査と語彙・構文・コミュニケーション力を見る日本語言語発達検査が実施された。

対象と方法

1）対象児童について

厚生労働科学研究費補助金・感覚器障害戦略研究（聴覚分野）症例対照研究は，平成21年４月～平成22年３月までの１年間，聴覚障害児の日本語言語発達に影響を与える因子を明らかにし，発達を保障する方法を考える目的で行われた。対象は４歳時までに70dB以上の難聴が確認された４歳（年中児）～12歳（小学校６年生）までの児638名である。施行した検査は，日本語言語力を測定する指標として，質問応答関係検査（TQAID），教研式標準学力検査CRT-II（国語・算数），失語症構文検査（STA），改訂版絵画語い発達検査（PVT-R），標準抽象語理解

力検査（SCTAW），語流暢性検査（WFT），読み書きスクリーニング検査（STRAW），また調整変数として，広汎性発達障害日本自閉症協会評定尺度（PARS），レーヴン色彩マトリックス検査（RCPM）を実施した。なお，研究の実施に先立ち保護者に対して，研究の内容に関する十分な説明を行った後，書面で同意を取得して実施した。またデータの収集に関しては，各施設において匿名化をはじめとする個人情報保護の手続きを遵守して実施した。

2）データの補正に関して

今回実施した検査のほとんどが，検査時年齢による得点の補正を行わない形式の検査であったため，年齢の増加に併って検査結果が上昇する傾向が認められた。従って学年をまたいだ解析を行う前に，回帰により全体の成績上昇係数を求め，その係数でデータの補正を行った。

まず，対象638児の検査実施時月齢と各検査の相関解析を行い，線形回帰式を求める事で回帰係数を求め（a），今回の検査対象者のおおよその平均月齢である100ヶ月を基準とし，各児の検査月齢（x_i）との偏差（$100-x_i$）を求め，回帰係数を乗じた値を個人の成績（y_i）に加えることで次式の通り補正後の点数（y_h）を求めた。

$$y_h = y_i + a(100 - x_i)$$

補正後のデータの比較および統計解析にはIBM SPSS ver18を用いた。

結　　果

難聴児の出生時体重の分布について

本研究の対象である，生下時から難聴を有すると考えられ，～4歳までに70dB以上の難聴であることが確定診断された児638名のうち，アンケート調査の出生時体重に関する質問に回答のあった627名について解析を行った。その結果，対象児の出生時体重は444g～4760gであり，平均は2894g（標準偏差566g）であった。

対象児の出生時体重の分布を詳細に見て行くと，おおよそ3000gを中心とした大きなピークとおおよそ1000gを中心とした小さなピークの2峰性の分布を示す事が明らかとなった（図1）。厚生労働省の出生に関する統計（平成22年）によると平成21年度出生児の平均出生児体重は3020gであることより，難聴児の出生時体重の分布のうち3000gを中心とした峰は一般的な児の出生児体重の分布と同一であることが示唆される。一方，出生時体重1000gをピークとした分布は本検討の対象とした難聴児に特有のものであることが推測され，従来の報告にある低出生体重による難聴のハイリスクグループであることが示唆された。

厚生労働統計などで用いられている出生児体重の分類に倣い，出生児体重2500g未満を低出生体重児（Low Birth Weight Infant: LBWI），1500g未満を極低出生体重児（Very Low Birth Weight Infant: VLBWI），1000g未満を超低出生体重児（Extremely Low Birth Weight Infant: ELBWI）の3群に分類すると，対象児627名のうちLBWIの総数は89名（14.2%）であった。そのうち，VLBWIは18名（2.9%）であり，さらにそのうちELBWIは10名（1.6%）であった。一方，出生に関する統計（平成22年）によると平成21年度の出生児1,049,141名のうち，LBWIは87,281名（8.3%），VLBWIは6,228名（0.6%），ELBWIは2,566名（0.2%）であり[5]，高度難聴を有する児においては有意に低出生体重児

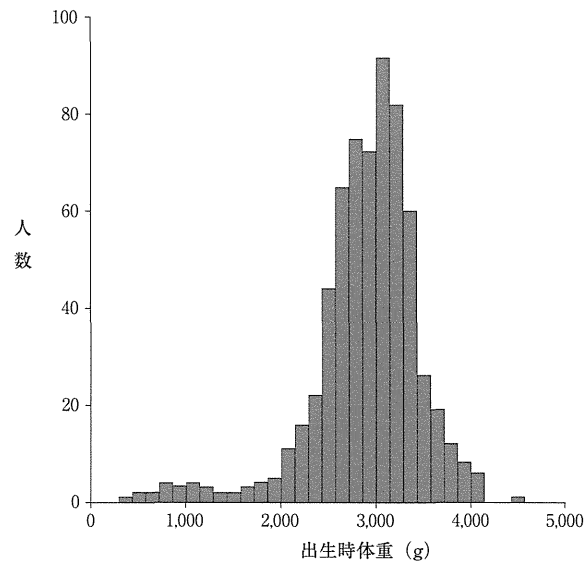

図1　対象児の出生時体重の分布
4歳までに70dB以上の高度～重度難聴と確定診断された児627名の出生時体重の分布を示す。おおよそ3000gを中心とした大きなピークと，おおよそ1000gを中心とした小さなピークの2峰性の分布を呈する。

が多いことが示された。

低出生体重難聴児の合併症の種類および頻度

難聴児における出生時体重と周産期リスクおよび出生時合併症について検討を行うため，対象児を出生時体重2500g以上の児（通常出生体重児）と，出生時体重2500g未満の低出生体重児とに分け，比較をおこなった（表1）。その結果，低出生体重の原因のひとつと考えられる妊娠中感染症の有無に関しては，低出生体重児群では約30%の児に妊娠中感染症があったのに対して，通常出生体重児では3%と大きな差が認められた。また，低出生体重児群では，人工呼吸器やNICUなどの周産期リスクファクターのあった児が約60%であるのに対して，通常出生児では約10%と大きな差が見られた。また，出生時合併症および頭頸部奇形・内耳奇形に関しては，循環器系疾患，呼吸器系疾患の合併頻度が高く，内耳奇形を伴うケースも有意に多いことが明らかとなった。

低出生体重難聴児の日本語言語発達

次に，低出生体重児の難聴発見時期や難聴の程度などの背景因子および日本語言語発達について検討を行うため，対象児を出生時体重2500g以上の通常出生体重児（538名）と，出生時体重2500g未満の低出生体重児（89名）とに分け，比較をおこなった。その結果，低出生体重児のほうが，難聴発見時期が3ヶ月程度早く，また裸耳聴力（平均）は3dBほど重度であることが明らかとなった（表2）。一方，補聴器・人工内耳の装用閾値に大きな差は認められず，最高語音明瞭度にも差は認められなかった。また，補聴器開始月齢にも差が認められなかった。非言語性知能の指標であるRCPMの結果は有意差を認めなかったが，広汎性発達障害の行動特性を評価するPARS（現在得点）では有意に低出生体重児が高得点であった。

また，日本語発達検査の結果を表3に示す。語彙レベルの日本語言語発達を評価するPVT-R,

表1 難聴児の出生時体重と周産期リスク・合併症の割合（%）

	妊娠中感染症の有無	周産期リスク（NICUなど）	頭頸部奇形	内耳奇形	循環器系疾患	内分泌系疾患	呼吸器系疾患
通常出生体重児	3.1	9.5	3.4	0.7	4.3	0.7	1.5
低出生体重児	30.6	58.3	7.4	7.4	21.9	3.8	10.7
p 値（χ^2 検定）	<0.01	<0.01	0.04	<0.01	<0.01	<0.01	<0.01

表2 難聴児の出生時体重と難聴発見時期などの背景因子の関係

	難聴発見時期	裸耳聴力	装用閾値	最高語音明瞭度	補聴器開始時月齢	レーヴン色彩マトリックス検査	広汎性発達障害日本自閉症協会評定尺度
通常出生体重児	13.3	100.7	41.8	61.4	16.7	26.8	5.3
低出生体重児	10.8	103.9	41.9	62.5	15.6	25.9	7.0
p 値（t 検定）	0.05	0.05	0.96	0.89	0.37	0.22	0.01

表3 難聴児の出生時体重と日本語発達度検査結果の比較

	質問—応答関係検査	CRT-II 国語	CRT-II 算数	絵画語い発達検査	標準抽象語理解力検査	語流暢性検査	失語症構文検査(理解)	失語症構文検査(産生)
通常出生体重児	213.5	45.0	55.6	28.2	12.9	13.1	22.5	33.3
低出生体重児	192.7	42.7	53.2	26.1	11.8	12.4	20.1	29.0
p 値（t 検定）	0.04	0.27	0.24	0.20	0.22	0.42	0.03	0.02

SCTAW，WFTのいずれも低出生体重児群のほうが通常出生体重児群より僅かに得点が低いが，２群間で有意な差は認められなかった。一方，構文レベルの日本語言語発達を評価するSTAでは，低出生体重児群のほうが通常出生体重児群より有意に得点が低かった。

またコミュニケーション能力を評価するTQAIDでは低出生体重児群のほうが通常出生体重児群より有意に得点が低かった。一方，学習習得度を評価するCRT-II国語，CRT-II算数でも，いずれも低出生体重児群のほうが通常出生体重児群より僅かに得点が低いが２群間で有意な差は認められなかった。

考　　察

本研究により，４歳までに70dB以上の難聴であると確定診断された高度～重度難聴児に占める低出生体重児の割合はおおよそ15％であり，少なくとも85％以上の児は低出生体重以外（遺伝性難聴など他の要因）による難聴である可能性が示唆された。

従来の報告では，新生児における聴覚障害の約半数は低出生体重児，重症新生児仮死，高ビリルビン血症，子宮内感染症，家族歴，先天異常症候群などのハイリスク児であることが報告されているが[1-4]，今回の調査では低出生体重児が15％程度，家族歴を有する児が10.5％，家族歴以外のハイリスクを有する児が6.7％であり，合計しても約30％であり70％の児はこれら難聴のリスクファクターを有していなかった（未発表）。

これは，本研究の対象が「４歳までに70dB以上の高度・重度難聴であることが確定診断された児」に限られているため，先天性・重度の難聴となる*GJB2*遺伝子変異などの遺伝性難聴の割合が，難聴児全体と比較して多いためと示唆される。Kimberingによると高度感音難聴児のおおよそ50％の原因は遺伝子であると推測されているが[6]，今回の結果では10％程度しか家族歴を有していなかった。これは近年の少子化により兄弟が少なくなっていることより，常染色体劣性遺伝形式をとる遺伝性難聴家系の多くが弧発例となっているためと考えられる。実際，著者らが先進医療「先天性難聴の遺伝子診断」として実施している遺伝学的検査では，弧発例のうちおおよそ40％に遺伝子変異が見出されることも，この考察を裏付ける[7]。

また，低出生体重難聴児に占める周産期リスクおよび出生時合併症について検討した結果では，低出生体重難聴児群では妊娠中感染症の頻度が通常出生体重難聴児群と比較して約10倍高頻度であり，先天性サイトメガロウイルス感染症などがその原因であることが推測された。また，低出生体重児群では，新生児仮死，人工呼吸器，NICUなどの周産期リスクファクターのあった児が約60％と過半数を占めていた。これは，低出生体重による発育不全の児が多い事を考え合わせると当然の結果であると言える。また，出生時合併症に関しては，循環器系疾患，呼吸器系疾患の合併頻度が高く，内耳奇形を伴うケースが有意に高いことから，同様に発育不全による影響が強く示唆された。近年，低出生体重がAuditory Neuropathy Spectrum Disease（ANSD）の原因の一つであることが報告されており[8-10]，今回の対象症例の中にも，低出生体重による発育不全に伴う内耳の奇形や低形成，蝸牛神経の低形成などANSDと診断される児が混在している可能性が示唆される。

また，言語発達に影響を及ぼし得る背景因子について検討を行ったところ，低出生体重児のほうが，難聴発見時期が３ヶ月程度早いことが示された。これは，低出生体重の児ほど難聴のリスクありと考えられ早期に難聴のスクリーニングがなされたことを反映しているものと思われる。一方，補聴器開始月齢にはあまり差が認められないことより，全身的な発育不全や合併症などの影響により，難聴の確定診断およびそれを受けた療育開始が遅れる傾向にあることが示唆された。また，RCPMの結果は有意差を認めなかったが，PARS（現在得点）では有意差を認めたことより，知的能力の発達に関しては大きな差は認めないものの，広汎性発達障害の行動特性を有する児が低出生体重児に多く見られることが示された。内山らによると，難聴児のうち身体的疾患・奇形を合併する例が32％，発達遅滞・自閉的発達障害・学習障害を伴う児が25％，合併症として両者を伴うものは16％であったと報告しているが[11]，今回の検討でも，低出生体重児の中に身体的疾患と広汎性発達障害の合併が示唆される症例が，通常出生体重の難聴児よりも多く含まれていることが示されたことより，低出生体重の児に対する介入および

(リ)ハビリテーション計画を立てる際には，難聴以外の身体的疾患への配慮とともに，広汎性発達障害合併の可能性を考慮する必要があると思われる。海外の報告においても，低出生体重が自閉症（Autism Spectrum Disorder）のリスクファクターであることが報告されているおり[12]，低出生体重難聴児においては，広汎性発達障害合併スクリーニング検査の実施が重要であると考えられる。さらに，低出生体重難聴児群において妊娠中感染症の有無によりPARS（現在得点）の比較を行ったところ，妊娠中感染症有群の平均が7.5点，妊娠中感染症無群の平均が5.8点であり（有意差なし），妊娠中感染症により広汎性発達障害の合併が多くなる可能性が示唆された。

また，日本語言語発達検査の結果に関しては，語彙レベルの検査項目であるPVT-R，SCTAW，WFTのいずれも低出生体重児群と通常出生体重児群に有意な差は認められなかったのに対して，構文レベルの検査項目であるSTAでは，低出生体重児群のほうが通常出生体重児群より有意に低得点であった。このことより，低出生体重難聴児では語彙レベルの発達よりも構文の理解および産生に課題を有するケースが多く含まれている可能性があり，構文発達の検査とそれに応じた指導が必要であると考えられる。

コミュニケーション能力を評価するTQAIDでは，低出生体重児群のほうが通常出生体重児群より有意に得点が低かったが，学習習得度を評価するCRT-II国語，CRT-II算数では2群間で有意な差は認められなかった。この結果と，前項の背景因子であるRCPMおよびPARSの結果をあわせて考えると，低出生体重難聴児の特徴として通常出生体重の難聴児よりも広汎性発達障害の行動特性を有する児が多く，その結果としてコミュニケーションに問題を抱える児が多い可能性が考えられた。

本研究により，難聴児における低出生体重児の占める割合およびその臨床的特徴を示すとともに，日本語言語発達の特徴が明らかとなり，また特に広汎性発達障害合併スクリーニングの重要性を明らかに出来たことは，今後の医学的介入および(リ)ハビリテーション計画立案のために極めて大きな意義を持つと考えられる。低出生体重児の中にも良好な言語発達を遂げている児も見られるが，何らかの問題を抱えている可能性を常に念頭に置き，医学的評価や日本語言語発達検査を早期から計画的に行い，その結果に基づいた個別の介入プログラムを立案・実施することが重要である。

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Prevalence of low birth weight infants among Japanese patients with hearing loss and the characteristics of their Japanese language development.

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Low birth weight has been reported as a risk factor for congenital hearing loss, however, its effect on the Japanese language development remains unclear. This study was aimed at investigating the prevalence of low birth weight infants among children with pre-lingual severe to profound hearing loss and to investigate the characteristics of their Japanese language development. Of 627 hearing-

impaired children recruited for this research, 89 children (14.2%) had a low birth weight of under 2,500g. In regard to the results of the Japanese language development tests in these children, the vocabulary development was the same as that in the normal birth weight hearing loss children, whereas the results of syntax development test and communication ability test were significantly inferior to those in the normal birth weight hearing loss children. Furthermore, the scores on the Pervasive Developmental Disorders Autism Society of Japan rating scale (PARS) in the low birth weight hearing loss children were higher than those in the normal birth weight hearing loss children. These results suggested that low birth weight may be a risk factor for hearing loss with pervasive developmental disorders.

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Hearing Handicap in Adults With Unilateral Deafness and Bilateral Hearing Loss

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Objective: To assess the perception of hearing handicap in adult patients with unilateral sudden sensorineural hearing loss (SNHL) compared with those with bilateral SNHL or unilateral congenital SNHL.
Study Design: Retrospective chart review.
Setting: Multicenter department of otolaryngology referrals.
Patients: Seventy-one subjects in the unilateral severe-profound (>70 dB) sudden SNHL group (Group 1), 17 subjects in the unilateral prelingual or congenital SNHL group (Group 2), and 121 subjects in the bilateral SNHL group (Group 3).
Interventions: Questionnaire.
Main Outcome Measures: Hearing Handicap Inventory for Adults (HHIA) and visual analogue scale (VAS) measurements of hearing handicap.
Results: Average levels of hearing loss were 92 dB in Group 1, 109 dB in Group 2, and 67 dB in Group 3. The relative percentage scores of HHIA and VAS compared with Group 3 were 72.6% and 81.0% in Group 1 and 25.4% and 50.3% in Group 2, respectively. A mild correlation between the HHIA subscale or VAS scores and degree of hearing loss could be found in Group 3. No significant correlation was found between the HHIA subscale or VAS scores and duration of hearing loss in Group 1 or Group 3. Higher scores were obtained in male subjects than in female subjects. Patients in Group 1 who were troubled by tinnitus scored significantly higher in the HHIA. In multiple logistic regression analysis, presence of tinnitus, older age, higher average hearing loss level, and group (bilateral SNHL>unilateral sudden SNHL>unilateral precongenital SNHL) revealed a significant positive association with high score (>42) of HHIA (odds ratio, 3.171, 1.021, 1.031, and 6.690, respectively).
Conclusion: The results of HHIA and VAS suggest that not only patients with bilateral SNHL but also those with unilateral sudden SNHL, particularly those who have tinnitus, experience a hearing handicap. **Key Words:** Sudden hearing loss—Hearing handicap—Questionnaire—Unilateral deafness.
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Population studies of sudden sensorineural hearing loss (SNHL) show a wide age distribution with an average of 50 to 60 years. The hearing loss is unilateral in most cases, with bilateral involvement reported in less than 5% of patients (1). The incidence of sudden SNHL has been reported to be between 5 and 30 cases per 100,000 per year (2). However, a study from Japan has shown an incidence as high as 275 cases per 100,000 per year (3).

Patients with single-side deafness (SSD) have difficulty hearing sounds coming from the deaf side, localizing a sound source, and perceiving speech against background noise, all of which have been explained by the "head shadow effect" (4,5). However, whether SSD has a noteworthy impact on the patients' well-being and social life remains under discussion.

Conventionally, the audiologic treatment of patients with SSD is a contralateral routing (CROS) hearing aid, in which a microphone, placed on the deaf side, transmits sound to the hearing ear either by wire or wireless means. Recently, the Bone-Anchored Hearing Aid (BAHA), which is a semi-implantable hearing aid and bone-conducting device, has also been applied as a treatment for patients with SSD (6,7). Cochlear implants have also been used in some patients with unilateral severe-to-profound hearing loss and ipsilateral tinnitus and were found to be beneficial in some cases (8,9). Several studies using the Hearing Handicap Inventory for Adults (HHIA) have demonstrated that unilateral hearing loss may affect the emotional and social

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well-being of adults with this condition and adults with unilateral hearing loss perceive themselves as handicapped (10–12). However, there is less information regarding the effects of unilateral sudden deafness with or without tinnitus compared with unilateral congenital deafness or bilateral hearing loss. In this study, we aimed to assess the level of hearing handicap using the HHIA and visual analog scale (VAS) for patients with unilateral sudden SNHL compared with those having unilateral congenital SNHL or bilateral SNHL in a multicenter study.

MATERIALS AND METHODS

Study Design

All subjects were enrolled in this multicenter study at 7 university schools of medicine in Japan, in institutions that belonged to the Acute Profound Deafness Research Committee (Tokyo, Japan). Questionnaire charts of 209 patients, treated between December 2009 and January 2011 at the Department of Otolaryngology of each Medical University Hospital, were reviewed retrospectively. All patients provided written informed consent for review of their records for research purposes. Each university review board approved the conduct of this study.

Subjects

Subjects were classified into 3 groups as follows: 1) unilateral severe to profound (>70 dB) sudden SNHL (Group 1), 2) unilateral severe to profound prelingual or congenital SNHL (Group 2), and 3) bilateral SNHL (Group 3). All subjects fulfilled the following criteria: a) a questionnaire with self-rated scales was completed over 6 months after the onset of hearing loss, b) patients were older than 20 years when they completed the questionnaire, c) unilateral severe-to-profound hearing loss was defined as average level of hearing loss (500, 1,000, 2,000, and 4,000 Hz) of more than 70 dB and an average level of the opposite side of below 30 dB, d) bilateral hearing loss was defined as an average level of hearing loss in the better hearing ear of greater than 30 dB, e) sudden SNHL was defined as a decrease in hearing occurring within 3 days or fewer without any identifiable cause, and f) prelingual or congenital SNHL was defined as onset of hearing loss occurring before the age of 7 years.

Questionnaire

The Japanese version of the HHIA questionnaire (Table 1) was used to evaluate the handicap. The HHIA is a self-assessment questionnaire of hearing handicap comprising 25 items, of which, 13 deal with emotional aspects (E) and 12 deal with social and situational aspects (S). For each item or situation, subjects are asked to give one of the following responses: "yes" (4 points),

TABLE 1. *The hearing handicap inventory for adults*

		tTeet p value G1-G2	t Teet p value GI-G3
S-1	Does your hearing difficulty make you use the phone less often than you would like?	0.079	0.001
E-2	Does your hearing difficulty make you feel embarrassed or out of place when you are introduced to stranger?	0.733	0.000
S-3	Does your hearing difficulty make you avoid group of people?	0.261	0.083
E-4	Does your hearing difficulty make you touchy?	0.092	0.898
E-5	Does your hearing difficulty make you feel frustrated or unhappy when talking to people of your family?	0.038	0.080
S-6	Does your hearing impairment cause any other difficulties when you go to the party or social meeting?	0.024	0.297
E-7	Does your hearing difficulties make you frustrated when talking to work mates?	0.223	0.001
S-8	Does your hearing difficulties when you go to the movies or theaters?	0.017	0.169
E-9	Does your feel harmed or down because of your hearing difficulty?	0.073	0.098
S-10	Does your hearing impairment cause difficulties when you visit friends, relatives and neighbors?	0.344	0.031
S-11	Does your hearing difficulty cause you problem to hear/understand work mates?	0.409	0.999
E-12	Does your hearing difficulty cause you nervous?	0.181	0.959
S-13	Does your hearing difficulty make you visit friends, relatives and neighbors less often than you would like to?	0.048	0.519
E-14	Does your hearing difficulty make you argue or fight with your family?	0.252	0.247
S-15	Does your hearing difficulty cause you trouble to watch TV or listen to the radio?	0.000	0.000
S-16	Does your hearing difficulty make you go out shopping less often than you would like to?	0.067	1.000
E-17	Does your hearing difficulty make you annoyed or unhappy?	0.277	0.671
E-18	Does your hearing difficulty make you prefer to be alone?	0.467	0.797
S-19	Does your hearing difficulty make you want to talk less to the people in your family?	0.140	0.137
E-20	Do you think that your hearing difficulty reduces or limit your personal or social life somehow?	0.959	0.999
S-21	Does your hearing difficulty make you trouble when you are in a restaurant with family or friend?	0.011	0.773
E-22	Does your hearing difficulty make you feel sad or depressed	0.109	0.564
S-23	Does your hearing difficulty make you watch less TV or listen to the radio less often than you would like to?	0.344	0.001
E-24	Does your hearing difficulty make you feel embarrasses or less comfortable when you talk to a friends?	0.635	0.289
E-25	Does your hearing difficulty make you feel isolated or feel aside within a group of people?	0.177	0.000

E indicates emotional subscale; G, group; S, social subscale.

"sometimes" (2 points), or "no" (0 points). Care was taken not to induce answers and to avoid interviewer bias.

In addition, subjects were asked to rate their hearing handicap in various everyday situations on a VAS, which is a psychometric measurement instrument for quantifying subjective phenomena. A VAS is presented as a horizontal line, 100 mm in length, with one end as 0 (absence of perception of hearing handicap) and the other as 100 (maximum). The subjects mark on the line the point that represents their current state; the VAS score is the distance in millimeters from the left ("absence") to the mark.

Statistical Methods

All statistical values were calculated using IBM SPSS Statistics 18 (IBM Corp. Armonk, NY, U.S.A.). We used the *t* test to compare each score of 25 items in the HHIA between groups (Group 1 to Group 2 and Group 1 to Group 3). Correlations and standard deviations within each group were examined. The significance level was set at 0.05. Pearson's correlation coefficient was used to study the relationship between the average hearing loss and subscales of HHIA or VAS score as well as the correlation between the duration of hearing loss and subscales of HHIA or VAS score. We used a multiple logistic regression analysis to assess the independent effects of age, sex, average hearing loss level, presence/absence of tinnitus, and unilateral precongenital SNHL versus unilateral sudden SNHL versus bilateral SNHL.

RESULTS

Seventy-one subjects (33 male and 38 female subjects) with a mean age of 52 years (range, 21–81 yr) were included in the unilateral sudden SNHL group (Group 1). Of these, 34 subjects (48%) were affected in the right ear. The average level of hearing loss was 92 dB (range, 70–115 dB). The average period between onset of hearing loss and completion of the questionnaires was 77 months (range, 6–237 mo). One hundred twenty-one subjects (58 male and 63 female subjects) with a mean age of 60 years (range, 20–97 yr) were included in the bilateral SNHL group (Group 3). The average levels of hearing loss in the better hearing ear, right ear, and left ear were 67 dB (range, 35–115 dB), 70.8 dB, and 71.5 dB, respectively. The average period between onset of hearing loss and completion of the questionnaires was 15 years (range, 1–56 yr). Seventeen subjects (10 male and 7 female subjects) with a mean age of 31 years (range, 20–77 yr) were included in the unilateral precongenital SNHL group (Group 2). Of these, 8 subjects (47%) were affected in the right ear. The average level of hearing loss was 109 dB (range, 75–115 dB). The causes of hearing loss were congenital deafness in 8 subjects, mumps in 7 subjects (average onset of hearing loss: 6.7 yr of age), and unknown in 2 subjects.

The mean total scores and emotional (E) and social (S) subscores together with the standard deviation values obtained from the HHIA questionnaire for the participants of Group 1, 2, and 3 were 35.8 ± 13.9 (total), 16.4 ± 13.5 (E) and 19.3 ± 14.2 (S); 12.5 ± 10.4 (total), 5.7 ± 4.4 (E) and 6.7 ± 6.5 (S); and 49.3 ± 13.6 (total), 22.4 ± 13.9 (E) and 27.0 ± 13.3 (S), respectively (Fig. 1A). Significant differences were found between all groups. Relative percentages of the HHIA scores in Group 1 and 2 compared with Group 3 were 72.6% (total), 73.2% (E) and 71.5% (S) and 25.1% (total); 25.4% (E) and 24.8% (S), respectively (Fig. 1B). The subjective handicap assessed by VAS was 51.8 ± 28.7 (Group 1), 28.5 ± 21.8 (Group 2), and 56.7 ± 29.0 (Group 3). Relative percentages of the VAS in Groups 1 and 2 compared with Group 3 were 81.0% and 50.3%, respectively (Fig. 2). Significant differences in the VAS scores ($p < 0.05$) were found in Groups 1 and 3 when compared with Group 2. Table 1 shows the comparison between the mean scores of HHIA for each item obtained for Groups 1 and 2 (G1-G2) or Group 3 (G1-G3). One item of the emotional subscale (E-5) and 5 items of the social subscale (S-6, S-8, S-13, S-15, and S-21) revealed significantly higher scores in Group 1 when compared with Group 2. Three items of the emotional subscale (E-2, E-7, and E-25) and 4 items of the social subscale (S-1, S-10, S-15, and S-23) revealed significantly higher scores in Group 3 when compared with Group 1.

Tables 2 and 3 show the Pearson's correlation between the hearing handicap (HHIA; emotional and social subscale and VAS scale) and degree and duration of hearing loss in Groups 1 and 3. A mild correlation ($0.2 < r \leq 0.4$) between

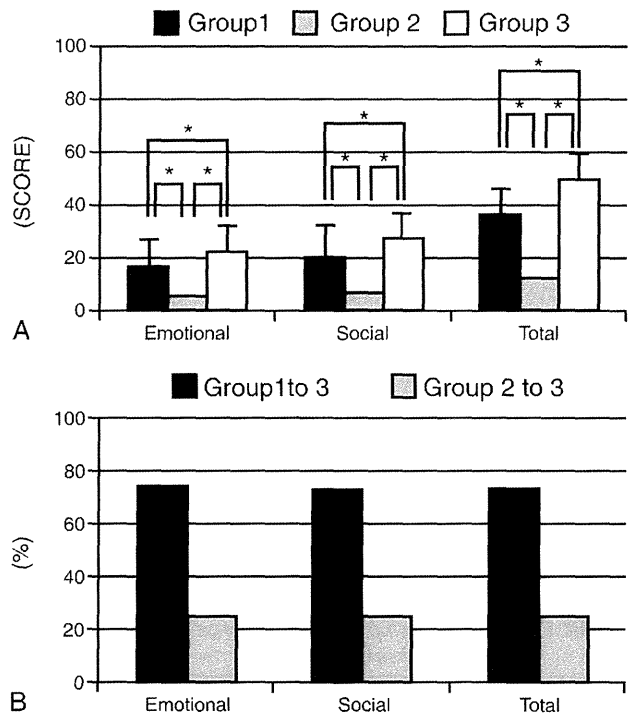

FIG. 1. Hearing Handicap Inventory for Adults (HHIA) scores for Groups 1, 2, and 3. Emotional, social, and total scores on the HHIA scale, in 3 groups of patients: Group 1, unilateral severe to profound (>70 dB) sudden sensorineural hearing loss (SNHL); Group 2, unilateral severe to profound prelingual or congenital SNHL; and Group 3, bilateral SNHL. Significant differences were found between groups (*A*). *$p < 0.05$. Relative percentages of the HHIA scores compared with Group 3 were 73.2% (E), 71.5% (S) and 72.6% (total) in Group 1 and 25.4% (E), 24.8% (S), and 25.1% (total) in Group 2 (B).

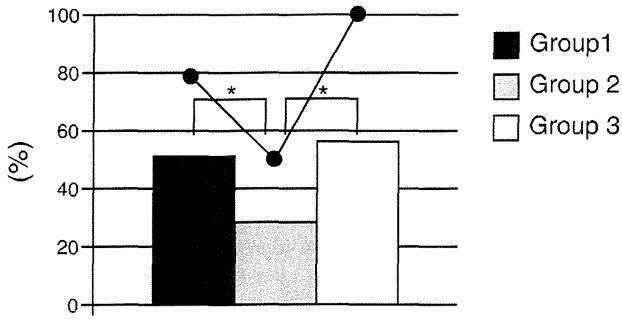

FIG. 2. Visual Analogue Scale (VAS) scores for Groups 1, 2, and 3. VAS scores in 3 groups of patients: Group 1, unilateral severe to profound (>70 dB) sudden sensorineural hearing loss (SNHL); Group 2, unilateral severe to profound prelingual or congenital SNHL; and Group 3, bilateral SNHL. Significant differences were found in Groups 1 and 3 when compared with Group 2. * $p < 0.05$. Relative percentages of the VAS were 81.0% in Group 1 to Group 3 and 50.3% in Group 2 to Group 3.

the HHIA subscale or VAS scores and degree of hearing loss could be found in Group 3. No significant correlation between the HHIA subscale or VAS scores and duration of hearing loss could be found in either Group 1 or Group 3. Figure 3 shows the mean scores of the emotional and social subscales in the HHIA and VAS related to sex in Groups 1, 2, and 3. Higher scores were found in male subjects compared with female subjects. Figure 4 shows the mean difference in the scores of HHIA (emotional and social subscale) and VAS between patients who had tinnitus and those who had no tinnitus in Groups 1 and 3. Patients with unilateral sudden SNHL (Group 1) who had tinnitus scored higher in the HHIA (E: $p < 0.05$ and S: $p < 0.05$).

We performed a multiple logistic regression analysis to determine the influence of age, sex, average hearing loss level, presence of tinnitus, and 3 groups (unilateral precongenital SNHL versus unilateral sudden SNHL versus bilateral SNHL) for the HHIA total score (Table 4). Patients who had tinnitus demonstrated a greater than 3-fold increased risk (odds ratio, 3.171) of high score (>42) in the HHIA compared with those who did not have tinnitus. High score (>42) in the HHIA indicated severe hearing handicap (10). A greater risk of high score in the HHIA

TABLE 2. *Relationship between average hearing loss and hearing handicap*

		Case	Average of HL (dB)	Average score	Pearson's correlation: r
HHIA (E)	Group 1	43	92.7	16.4	0.125
	Group 3	110	67.5	22.4	0.282
HHIA (S)	Group 1	43	92.7	19.3	0.182
	Group 3	110	67.5	27.0	0.385
VAS	Group 1	42	93.2	51.8	0.013
	Group 3	91	68.4	56.7	0.276

HHIA (E) indicates Hearing Handicap Inventory for Adults (emotional); HHIA (S), Hearing Handicap Inventory for Adults (social); HL, hearing level; VAS, visual analog scale.

TABLE 3. *Relationship between the duration of hearing loss and hearing handicap*

		Case	Average of DHL	Average score	Pearson's correlation: r
HHIA (E)	Group 1	43	78.5 Mo	16.4	0.124
	Group 3	56	189.0 Mo	21.1	0.084
HHIA (S)	Group 1	43	78.5 Mo	19.3	0.144
	Group 3	56	189.0 Mo	23.8	0.006
VAS	Group 1	42	74.7 Mo	51.8	0.106
	Group 3	51	181.2 Mo	56.5	0.135

DHL indicates duration of hearing loss.

(odds ratio, 6.690) was found in the patients with bilateral SNHL compared with those with unilateral sudden SNHL and in the patients with unilateral sudden SNHL compared with those with unilateral precongenital SNHL. The association was also significant in the patients with older age and higher average hearing loss level (Table 4).

DISCUSSION

The original HHIA (13) is in English and has high internal consistency with regard to its questions, test-retest reliability, and low standard error (14). The HHIA questionnaire has been translated into Italian (15), Brazilian Portuguese (16), and Japanese (17). The validity and reliability of the translated versions of the HHIA have also been reported in the literature. The average scores of the HHIA in adult patients with bilateral hearing loss were reported to be 52.2 ± 26.6 (total); 26.7 ± 15.3 (E) and

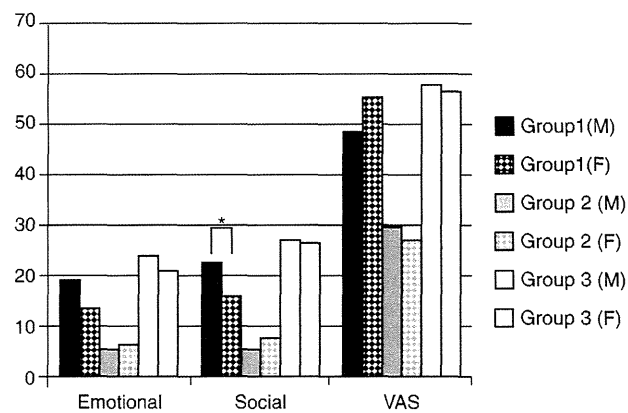

FIG. 3. Mean scores on the emotional and social subscales in the Hearing Handicap Inventory for Adults (HHIA) and Visual Analogue Scale (VAS) according to sex in Groups 1, 2, and 3. Emotional and social scores on the HHIA scale and VAS, in 3 groups of patients: Group 1, unilateral severe to profound (>70 dB) sudden sensorineural hearing loss (SNHL); Group 2, unilateral severe to profound prelingual or congenital SNHL; and Group 3, bilateral SNHL. Higher scores were found in male subjects compared with female subjects. The score of the social subscale of the HHIA in male subjects was significantly higher than that in female subjects. * $p < 0.05$.

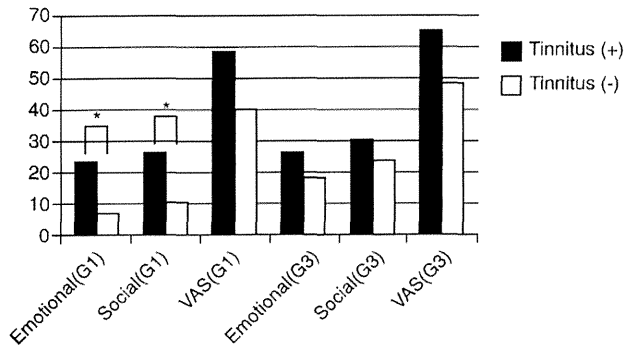

FIG. 4. Differences in the Hearing Handicap Inventory for Adults (HHIA) scores (emotional and social subscale) and Visual Analogue Scale (VAS) between patients who had tinnitus and those who did not in Groups 1 and 3. Emotional and social scores on the HHIA scale and VAS, in 2 groups of patients, some of whom also have tinnitus: Group 1, unilateral severe to profound (>70 dB) sudden sensorineural hearing loss (SNHL); and Group 3, bilateral SNHL. Those patients with unilateral sudden SNHL (Group 1) who also had tinnitus revealed significantly higher scores in the HHIA than those who were not affected. *: $p < 0.05$.

25.9 ± 12.1 (S) in Brazil (14) and 37.3 ± 16.7 (total); 21.9 ± 8.9 (E) and 15.4 ± 7.8 (S) in Italy (13). In the present study, the average score was 49.3 ± 13.6 (total); 22.4 ± 13.9 (E) and 27.0 ± 13.3 (S). Our results are therefore similar to those in the Brazilian study. The average score in the Italian study was slightly low because it seemed that the hearing threshold (hearing level from 29 to 71 dB) was also lower compared with the Brazilian subjects (hearing level from 26 to 93 dB) and the present subjects (hearing level from 35 to 115 dB). Some studies showed high correlations between the hearing handicap and degree of hearing loss in the population with bilateral hearing loss (15,17), and we confirmed weak correlations between the scores of HHIA or VAS and better ear pure-tone average in the bilateral SNHL group. Otherwise, the correlation could not be confirmed in the unilateral SNHL population. Among the population in our study, logistic regression analysis revealed that higher hearing loss level increased risk of severe hearing handicap in the HHIA score. We were also unable to confirm significant correlations between the duration of hearing loss and hearing handicap in the present study.

The HHIA and VAS scores of patients with unilateral sudden SNHL were significantly higher than in those with unilateral prelingual or congenital SNHL. This result reveals that unilateral postlingual deafness including sudden SNHL may be perceived as a hearing handicap for adults. Many patients with unilateral sudden hearing loss experience a hearing handicap in emotional and social situations. Hearing handicap, based on a score of greater than 18 in the HHIA, was previously reported in 73.1% (16) and 74.6% (17) of unilateral hearing impaired subjects. In our study, a hearing handicap was found in 69.8% of the subjects and high relative percentages of the HHIA (72.6%) and VAS (81.0%) scores were confirmed in the patients with unilateral sudden SNHL compared with those with bilateral SNHL. These scores showed that their experience of sudden SSD was almost as bad as the experience of patients with bilateral SNHL. However, subjects with unilateral prelingual or congenital SNHL revealed low relative percentages of the HHIA (25.1%) and VAS (50.3%) scores compared with subjects with bilateral SNHL. These findings thus emphasize that adults with sudden SSD experience this as a serious handicap. A greater risk of 6.69 times for severe hearing handicap in the HHIA score was found among the 3 groups. The factor of bilateral SNHL increased risk of hearing handicap in the HHIA score compared with that of unilateral sudden SNHL and the factor of unilateral sudden SNHL increased risk of the hearing handicap compared with that of unilateral precongenital SNHL.

Vicci de Araùjo et al. (10) have demonstrated a lower hearing handicap in male subjects compared with female subjects having unilateral hearing loss. However, our results show the opposite outcome, demonstrating a greater hearing handicap in male subjects compared with female subjects with either unilateral sudden or bilateral SNHL. Particularly, the outcome of the social subscale of HHIA in the unilateral sudden SNHL group was statistically significant. These findings reveal that unilateral sudden deafness may cause difficulties in life in a social environment. Disability of auditory function because of unilateral sudden deafness affects speech perception, communication in the presence of background noise, and social interaction. However, sex differences were insignificant risk factor for severe hearing handicap in the HHIA score.

The majority of people with unilateral sudden deafness experience tinnitus. Severe tinnitus can seriously impair the ability of patients to perform their activities in daily life and lower their quality of life. In the present study, the scores of HHIA and VAS were higher in patients who had tinnitus compared with those who did not feel tinnitus in Groups 1 and 3. The emotional and social subscales of HHIA were significantly higher in patients with unilateral sudden SNHL who had tinnitus. It is noteworthy that the risk of severe hearing handicap in the HHIA score among patients with tinnitus was approximately 3.71 times higher than that among those without tinnitus. The present study might indicate that unilateral sudden SNHL in adults with tinnitus causes significant hearing handicap with respect

TABLE 4. *Multiple logistic regression analysis predicting the risk of high score (>42) in the Hearing Handicap Inventory for Adults*

Variable	Odds ratio	*p*
Tinnitus	3.171	0.013
Age	1.021	0.041
Group	6.69	0.06
Average HL	1.031	0.001

HHIA indicates Hearing Handicap Inventory for Adults; Ave. HL, average hearing loss level.

Group: bilateral SNHL versus unilateral sudden SNHL versus unilateral precongenital SNHL.

to emotional and social consequences. Tinnitus adds a significant burden to those who experience this in addition to hearing loss. In recent years, cochlear implants have successfully been used to treat severe tinnitus in patients with SSD (8,9,18,19). In tinnitus cases treated with implants, 60% to 90% of patients with hearing loss revealed an improvement in perception (19). Moreover, the rehabilitation of patients with unilateral deafness with cochlear implants yields better results in speech comprehension and localization (9). We conclude that it is necessary to highlight treatment for unilateral sudden deafness in adults with tinnitus because adults who experience sudden unilateral hearing loss, particularly those who also experience tinnitus, find this a handicap in their daily lives.

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Evaluation of cortical processing of language by use of positron emission tomography in hearing loss children with congenital cytomegalovirus infection☆

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ABSTRACT

Objective: To predict cochlear implant efficacy and investigate the cortical processing of the visual component of language in profoundly deafened patients with asymptomatic congenital cytomegalovirus (CMV) infection.

Methods and cases: The cortical activity of two children with CMV-related hearing loss was evaluated with fluorodeoxyglucose-positron emission tomography (FDG-PET) with a visual language task before cochlear implantation. Total development and auditory perception ability were assessed one year after implantation.

Results: The two children with CMV-related hearing loss showed activation in the auditory association area where no activation was found in the controls, and exhibited nearly identical cortical activation patterns to those seen in patients with profound congenital hearing loss. In contrast, differences in total development in verbal ability and discrimination of sentences between the two cases were revealed one year after implantation.

Conclusion: These results might indicate that the differences of cortical activities according to hearing abilities could have been influenced by CMV infection that involves higher function of the brain directly and/or affects the cochlea peripherally. Additionally, if CMV infection might have affected only the cochlea, these cortical activation patterns were influenced secondary by the time course of hearing loss characterized by CMV infection, which had varied manifestations.

Accurate diagnosis and cochlear implantation at the appropriate time are important for successful speech development, and each patient needs a personalized habilitation program based on their etiology and brain function.

1. Introduction

Functional brain imaging provides important evidence of the plasticity of the central auditory pathway following a profound loss of hearing, and is one of the effective methods for investigating the cortical processing of language [1,2]. Previous studies have shown low levels of auditory cortical activity in subjects with profound deafness, i.e. lower levels of activity are observed with longer durations of deafness [3,4]. The importance of early hearing inputs by hearing aids or cochlear implantation (CI) has also been shown. Children with prelingual deafness can acquire spoken language by CI, but this approach is less effective in older children who have not acquired language during the critical language acquisition periods [5,6]. The development of the auditory cortex is believed to depend on the patient's auditory experience within 'critical periods' in the early lifetime. Positron emission tomography (PET) activation study by visual language task has shown that low glucose metabolism in the temporal auditory cortex predicts a good CI outcome in prelingually deafened children, which suggests that low metabolism in the

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auditory cortex may indicate its potential of plasticity for spoken language acquisition [7].

Congenital cytomegalovirus (CMV) infection is the most common environmental cause of developmental disability and sensorineural hearing loss (SNHL) in children [8]. Approximately 90% of infected infants are born with no clinical symptoms of congenital CMV infection, such as microcephaly, growth retardation, hepatomegaly, jaundice, or abnormal neurologic findings. SNHL is found in 6–23% of these asymptomatic infection cases and is often late-onset, fluctuating and progressive in nature within the first 6 years of childhood [9,10]. Hence, newborn hearing screening often does not detect problems in children with asymptomatic congenital CMV infection, and at the time of eventual SNHL diagnosis, the exact time course and manifestations cannot be determined [11]. The development of auditory skills and experiences of children with congenital CMV infection with associated hearing loss are unclear due to various clinical histories. Hearing impairment resulting from (even asymptomatic) congenital CMV infection might be not only of cochlear origin but also have central nerve involvement and entail possible risk of CMV-associated disorders later in life. Brain function and CI outcomes have not been examined in asymptomatic congenital CMV-associated hearing loss. In this study, we used ^{18}F-fluorodeoxyglucose (FDG)-PET to measure cortical glucose metabolism with a visual language task before CI in two profoundly deaf children with asymptomatic congenital CMV infection in order to assess the activities of the auditory cortex and predict the CI outcomes.

2. Methods and cases

2.1. Diagnosis of congenital CMV infection

To analyze congenital CMV infection, we used CMV DNA quantitative PCR (qPCR) analysis. Before qPCR analysis, total DNA including genomic DNA and CMV DNA was extracted from preserved dried umbilical cords. Each 10 pg total DNA was analyzed by a Step One Real-Time PCR System (Applied Biosystems, Foster City, CA, USA) using a TaqMan Universal Master Mix II (Applied Biosystems). The detailed methods of qPCR have been described previously (Furutate et al.) [12].

2.2. FDG-PET scanning and image analysis

FDG-PET scanning and image analysis were performed using the methods described by Fujiwara et al. [7]. During the time period between the intravenous injection of 370 MBq 18 F-FDG (the dose was adjusted according to the body weight of each subject) and the PET scanning of the brain, the patients were instructed to watch a video of the face of a speaking person reading a children's book. The video lasted for 30 min, and several still illustrations taken from the book were inserted (for a few seconds each) to help the subjects to follow the story. The subjects were video-recorded to confirm that they were watching the task video. PET images were acquired with a GE ADVANCE NXi system (General Electric Medical Systems, Milwaukee, WI, USA). The patients were then sedated by an anesthesiologist, and their heads were immobilized with a bandage during the scan. Spatial preprocessing and statistical analysis were performed with SPM2 (Institute of Neurology, University College of London, UK) implemented in Matlab (Mathworks, MA, USA). The cortical radioactivity of each deaf patient was compared with that of a control group by a t test in the basic model of SPM2. The control group consisted of six normal-hearing (pure tone average hearing levels within 20 dB HL) right-handed adult (27.5 ± 3.8 years) subjects. The statistical significance level was set at $p < 0.001$ (uncorrected).

2.3. Measurement of language and total development

Before CI, we evaluated the patients' mental development by the Kyoto scale of psychological development (*K*-test) in which Cognitive-Adaptive development [13] that consists of non-verbal reasoning or visuospatial perceptions is measured. This test is used commonly to assess developmental status for Japanese language users and the results are described as a developmental quotient (DQ) in comparison to normal controls. In the *K*-test, developmental delay is defined by DQ below 80.

One year after CI, auditory perception ability was assessed by word and sentence discrimination tests, which are components of the CI2004 test battery for children. Audible word discrimination tests were administered by a speech therapist with live voice stimuli presented at 70 dB in a soundproof room. We also evaluated intellectual development using the Japanese version of the WISC-III that corresponds to the Wechsler Intelligence Scale for Children (WISC) and contains non-verbal and verbal ability components. The Japanese WISC-III includes five subsets for performance IQ (PIQ) (picture completion, picture arrangement, block design, object assembly and coding) and five subsets for verbal IQ (VIQ) (information, comprehension, similarities, arithmetic and vocabulary).

This study was approved by the Ethics Committee of Shinshu University School of Medicine and prior written consent was obtained from the parents of both children after a full explanation of the study.

2.4. Details of cases

2.4.1. Case 1

This case was a 5-year-old girl. She had no particular events in the perinatal period and passed the newborn hearing screening. However at age 4 years 11 months, her mother suspected hearing loss because of poor response to sound. She only had mild expressive language impairment; her fine motor skills were unaffected. An auditory steady state response (ASSR) test showed bilateral hearing loss (approximately, right: 60 dB, left: 110 dB) (Fig. 1A). She was promptly fitted for bilateral hearing aids. After one month, a follow-up ASSR test indicated deterioration of hearing in her right ear to over 110 dB (Fig. 1C). At this point, DNA testing for hereditary hearing loss e.g. screening for mutations in the *GJB2* and *SLC26A4* genes, and checking for congenital CMV infection using preserved dried umbilical cord (above mentioned) was performed to diagnose the cause of hearing loss. These tests revealed that there were no pathological mutations causing hearing loss, but there were positive results for CMV infection. It was suspected that this late-onset, and rapidly progressive for one month, hearing loss was due to asymptomatic congenital CMV infection. Computed tomography (CT) findings of the middle and inner ear were normal. Hearing aids were not expected to be adequate to acquire spoken language, therefore CI was performed in the left ear at the age of 5 years 5 months.

2.4.2. Case 2

This 4-year-old girl had no particular events in the perinatal period and had not undergone newborn hearing screening. Her parents noticed that she did not respond to their voices when she had just turned 3 years old. She visited a hospital for a checkup where she was diagnosed by ASSR test at the age of 3 years 6 months with hearing loss that was approximately right: 60 dB, and left: 110 dB (Fig. 1B). She attended rehabilitation for hearing, using a combination of finger signing and gestures. In the following year, her hearing deteriorated further to right: 90 dB, left: 110 dB at the age of 4 years five months and her speech development was not

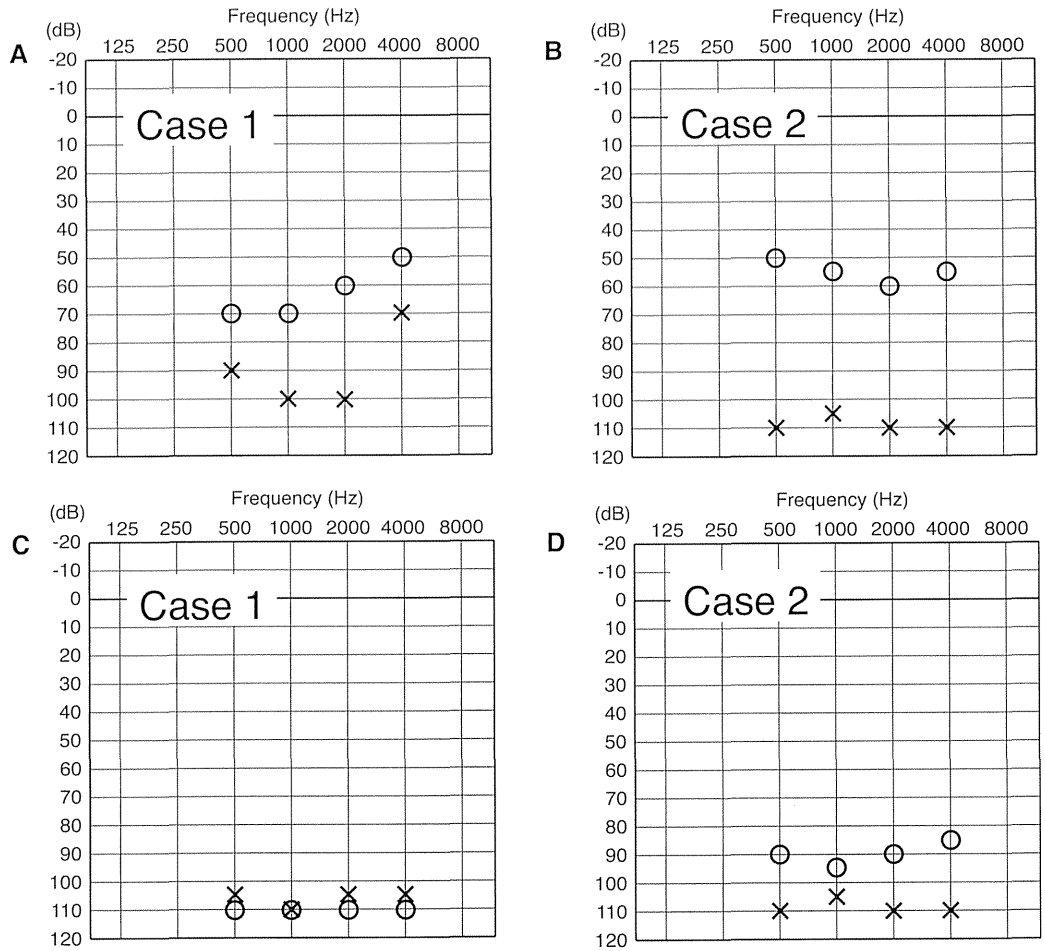

Fig. 1. (A) Case 1; a 5-year-old girl with asymptomatic congenital CMV infection (threshold using ASSR test). (B) Case 2; a 5-year-old girl with asymptomatic congenital CMV infection. These were results of first diagnosed with hearing loss. (C and D) Deterioration in hearing, for one month and for one year, respectively.

significant (Fig. 1D). She was referred to our hospital for further examinations, and her preserved umbilical cord demonstrated a positive result for congenital CMV infection. Late-onset and slowly progressive hearing loss for one year was suggested. There were no inner ear abnormalities seen in the CT findings. She underwent CI surgery in the left ear at the age of 4 years 9 months.

Each child received the same rehabilitation according to auditory oral method by the same speech therapist after implantation.

Table 1
The activated areas of the brain in profoundly deaf individuals during speech-reading.

Case	Gender/age (years)	Activated areas	
		Right hemisphere	Left hemisphere
1	Female/5	Superior temporal gyrus [BA22] Cingulate gyrus [BA31] Middle frontal gyrus [BA9]	Middle temporal gyrus [BA21] Inferior parietal lobe [BA40] Occipital gyrus [BA19] Precueus [BA7]
2	Female/5	Middle temporal gyrus [BA21] Postcentral gyrus [BA3/1/2] Middle occipital gyrus [BA20] Middle frontal gyrus [BA9]	Precentral gyrus [BA4] Precuneus [BA31] Precuneus [BA19] Cingulate gyrus [BA24]

3. Results

3.1. Brain imaging with PET

Table 1 and Fig. 2 show the areas that were activated in each child during a speech-reading task. The following cortical areas showed significantly higher metabolism during speech-reading in the children compared to normal hearing control subjects. In Case 1, the activated areas were the bilateral auditory association area [BA21], the bilateral precuneus, somatosensory cortex [BA7], the left secondary visual area [BA19], and the left inferior parietal lobule [BA40].

The activated areas in Case 2 were similar to those in Case 1, but the activation of the visual association areas in the parietal lobe were lower and smaller than in Case 1.

3.2. Assessment before cochlear implant, and outcome

Table 2 shows the children's scores in the *K*-test before CI, in the word and sentence discrimination tests, and in the Japanese WISC-III at one year after implantation. *K*-test scores that assessed Cognitive-Adaptive development of each child were almost similar. Both cases showed 30–40 dB of aided hearing thresholds at all frequencies with CI. One year after CI, the results of the Japanese WISC-III showed a clearer difference in VIQ than PIQ, in which Case 1 had a better score compared with Case 2. Case 1 did

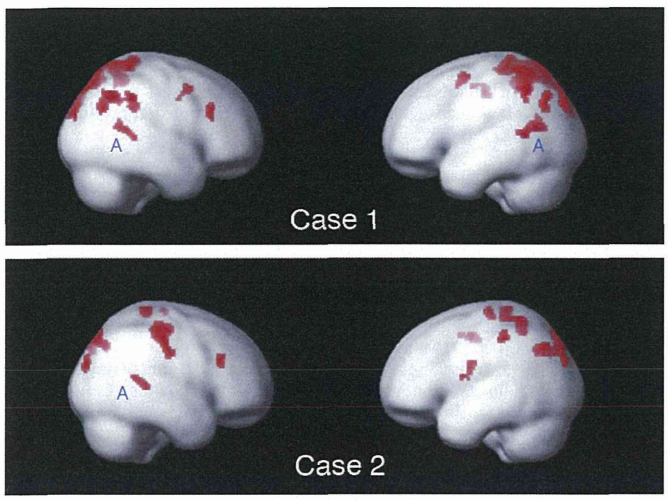

Fig. 2. Cortical activation by language-related visual stimuli in two profoundly deafened with congenital CMV infection cases. Case 1 and 2 showed significant activation in auditory association areas (A) (SPM2, $p < 0.001$, uncorrected).

Table 2
The results of total development before and after cochlear implant, and auditory assessment.

	Before CI	One year after CI	
	K-test (Cognitive-Adaptive)	WISC-III (Japanese version)	Infant word and sentence discrimination
Case 1	99	PIQ 101 VIQ 84	Word 98% Sentence 90%
Case 2	93	PIQ 93 VIQ 56	Word 100% Sentence 53%

CI, cochlear implant; *K*-test, Kyoto scale of psychological development; WISC, Wechsler Intelligence Scale for Children; PIQ, performance IQ; and VIQ, verbal IQ.

better as well in the sentence discrimination component of the auditory perception ability assessment while their results were similar regarding words in the word and sentence discrimination test for children.

4. Discussion

This is the first report on the evaluation of cortical processing of language in hearing loss children with congenital CMV infection. In infants with congenital CMV infections, as many as 20% will suffer from some degree of SNHL, either fluctuating or progressive [14]. This may present a late onset hearing loss, even if the results of newborn hearing screening were normal. The clinical courses of hearing loss in Cases 1 and 2 were typical for asymptomatic congenital CMV infection. Performance and outcome of children with CIs have a strong relation to hearing variables such as onset and course of hearing loss, age of hearing aids fitting, and social background variability, which depends on habilitation and education. According to Fukushima et al. and Kawasaki et al., children with *GJB2* mutations as the etiology for hearing loss have an advantage in their CI outcomes and speech acquisition with normal cognitive development compared with children with unknown etiologies, but this is because the hearing loss is of cochlear origin [15,16]. On the other hand, widely varying conclusions regarding CI outcomes with congenital CMV infection have been reported. Some studies reported the efficacy being not inferior to that of other CI recipients, while others reported it being much poorer [9,17–20]. Accordingly, prediction of CI outcomes for hearing loss with CMV infection is still difficult, unclear, and inconsistent because of various manifestations, progression and the possibility of involvement of higher brain function. Yamazaki et al. suggested that CI with CMV infection outcomes vary widely depending on the psycho-neurological disorders, with their differences in proportion and severity [19].

In this study, the auditory association area in the temporal lobe was activated bilaterally in Case 1 and unilaterally in Case 2. Fujiwara et al., in a FDG-PET study using the same methods and tasks as the present study, showed that subjects with better spoken language skills had less temporal lobe activation [7]. These cases exhibited nearly identical cortical activation patterns to those of congenitally deafened children, suggesting that they did not have enough hearing to develop the cortical network for audition. Previous studies have suggested that plastic changes in auditory cortices were strongly determined by the duration of auditory deprivation [21,22]. However, our two cases of children with CMV-related hearing loss were affected with severe bilateral hearing loss over a short period and were able to acquire spoken language with only a little delay for their age group. It is an interesting but unsolved question why they exhibited results that were the same as previous reports of pre-lingually deafened patients who did not receive sufficient auditory signals and therefore depended on visual cues. One possibility was that high speech-reading activation in the temporal auditory area might be linked to the condition of lacking auditory speech skills at that point, rather than reflecting a consequence of replacement by visual cross-modal plasticity due to a hearing loss of long duration. Besides, visual language activation in the auditory area may change even if affected by hearing loss of a short duration, or it might be influenced by the age-related metabolic changes during the critical period for spoken language acquisition. Another possibility was that these results might indicate that both cases had not received sufficient hearing stimulation as a foundation of language during their early years, which may be attributed to the central nervous system impairment of CMV infection.

Regarding the results of assessment after CI, there was a difference of cognitive ability with VIQ and hearing ability of sentence discrimination, with Case 1 having better CI performance than Case 2 (Table 2). In the assessment of auditory performance, Case 2 especially had difficulty in sentence discrimination despite having the same score in word discrimination as Case 1, who had better CI performance. Sentence discrimination tests require not only audible sound coded by CI, but also recognition of semantics and syntax that would be developed and established with hearing experiences during growth. Indeed, because of the differences between our two cases of the brain imaging, especially in the auditory cortex, we were uncertain whether it might be affected by CMV infection or the onset of their hearing loss itself. However, it raised the possibility that involvement of central nerve and high brain function relevant to CMV infection may lead to retardation of sentence discrimination and speech acquisition in Case 2. On the other hand, there was a difference of activation patterns in the parietal visual association areas. Case 2 showed lower and smaller than in Case 1. Fujiwara et al. predicted that the children with deafness were likely to depend more on vision than normal hearing children do. In Case 1, when someone talked to her, she might have been able to pay much more attention to their facial expression, gestures and visual cues for understanding better than Case 2. Lee et al. reported the comparison of brain metabolic activity between good and poor CI outcomes [23]. The activity patterns in the parietal regions of those with good CI outcomes in their study were similar to our result in Case 1.

We considered that these results might indicate that the differences of cortical activities according to hearing abilities could have been influenced by CMV infection that involves higher function of the brain directly and/or affects the cochlea peripherally. Additionally, if CMV infection might have affected only the

cochlea, these cortical activation patterns were influenced secondary by the time course of hearing loss characterized by CMV infection, which had varied manifestations.

Accurate diagnosis of hearing loss and early cochlear implantation are important for successful speech development. The approach using PET could help those involved in the habilitation and education of pre-lingually deafened children to decide upon the appropriate mode of communication for each individual. Brain imaging technologies to evaluate the neural basis for auditory speech skills have been developed and much evidence has been reported; however, correlation with hearing loss etiology, pathology and cross-modal plasticity of auditory cortex remains contentious. Further evaluations of the cortical metabolism before and after implantation are necessary for establishing appropriate personalized audiologic rehabilitation programs for individuals based on their etiology and brain function.

Conflicts of interest statement

We, the authors, declare that there were no conflicts of interest in conjunction with this paper.

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Hearing Handicap in Adults With Unilateral Deafness and Bilateral Hearing Loss

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Objective: To assess the perception of hearing handicap in adult patients with unilateral sudden sensorineural hearing loss (SNHL) compared with those with bilateral SNHL or unilateral congenital SNHL.
Study Design: Retrospective chart review.
Setting: Multicenter department of otolaryngology referrals.
Patients: Seventy-one subjects in the unilateral severe-profound (>70 dB) sudden SNHL group (Group 1), 17 subjects in the unilateral prelingual or congenital SNHL group (Group 2), and 121 subjects in the bilateral SNHL group (Group 3).
Interventions: Questionnaire.
Main Outcome Measures: Hearing Handicap Inventory for Adults (HHIA) and visual analogue scale (VAS) measurements of hearing handicap.
Results: Average levels of hearing loss were 92 dB in Group 1, 109 dB in Group 2, and 67 dB in Group 3. The relative percentage scores of HHIA and VAS compared with Group 3 were 72.6% and 81.0% in Group 1 and 25.4% and 50.3% in Group 2, respectively. A mild correlation between the HHIA subscale or VAS scores and degree of hearing loss could be found in Group 3. No significant correlation was found between the HHIA subscale or VAS scores and duration of hearing loss in Group 1 or Group 3. Higher scores were obtained in male subjects than in female subjects. Patients in Group 1 who were troubled by tinnitus scored significantly higher in the HHIA. In multiple logistic regression analysis, presence of tinnitus, older age, higher average hearing loss level, and group (bilateral SNHL>unilateral sudden SNHL>unilateral precongenital SNHL) revealed a significant positive association with high score (>42) of HHIA (odds ratio, 3.171, 1.021, 1.031, and 6.690, respectively).
Conclusion: The results of HHIA and VAS suggest that not only patients with bilateral SNHL but also those with unilateral sudden SNHL, particularly those who have tinnitus, experience a hearing handicap. **Key Words:** Sudden hearing loss—Hearing handicap—Questionnaire—Unilateral deafness.
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Population studies of sudden sensorineural hearing loss (SNHL) show a wide age distribution with an average of 50 to 60 years. The hearing loss is unilateral in most cases, with bilateral involvement reported in less than 5% of patients (1). The incidence of sudden SNHL has been reported to be between 5 and 30 cases per 100,000 per year (2). However, a study from Japan has shown an incidence as high as 275 cases per 100,000 per year (3).

Patients with single-side deafness (SSD) have difficulty hearing sounds coming from the deaf side, localizing a sound source, and perceiving speech against background noise, all of which have been explained by the "head shadow effect" (4,5). However, whether SSD has a noteworthy impact on the patients' well-being and social life remains under discussion.

Conventionally, the audiologic treatment of patients with SSD is a contralateral routing (CROS) hearing aid, in which a microphone, placed on the deaf side, transmits sound to the hearing ear either by wire or wireless means. Recently, the Bone-Anchored Hearing Aid (BAHA), which is a semi-implantable hearing aid and bone-conducting device, has also been applied as a treatment for patients with SSD (6,7). Cochlear implants have also been used in some patients with unilateral severe-to-profound hearing loss and ipsilateral tinnitus and were found to be beneficial in some cases (8,9). Several studies using the Hearing Handicap Inventory for Adults (HHIA) have demonstrated that unilateral hearing loss may affect the emotional and social

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