表5 言語性IQ・動作性IQと下位検査

1）言語性IQと下位検査との相関：検査実施順

	知識	単語	算数	類似	理解
全被検児	0.88**	0.85**	0.86**	0.80**	0.82**
人工内耳群	0.77**	0.86**	0.88**	0.85**	0.79**
最重度群	0.90**	0.84**	0.84**	0.76**	0.81**
重度群	0.88**	0.82**	0.80**	0.87**	0.80**
中等度群	0.89**	0.88**	0.89**	0.68**	0.77**
上位群	0.64**	0.35	0.58**	0.32	0.61**
中位群	0.37*	0.21	0.43**	0.50**	0.33
下位群	0.40*	0.27	0.72**	0.29	0.50**
最下位群	0.46	0.41	0.48	0.49	0.67**

全被検児，聴力別，言語性IQ別の各群．**：P<0.01，*：P<0.05で有意な相関

2）動作性IQと下位検査の相関：検査実施順

	動物の家	絵画完成	迷路	幾何図形	積木模様
全被検児	0.49**	0.51**	0.66**	0.65**	0.69**
人工内耳群	0.79**	0.53*	0.73*	0.75**	0.70**
最重度群	0.47*	0.54**	0.66**	0.63**	0.69**
重度群	0.42*	0.50**	0.67**	0.78**	0.62**
中等度群	0.45*	0.51**	0.58**	0.41*	0.81**
上位群	0.73**	0.58**	0.67**	0.62**	0.64**
中位群	0.39*	0.44**	0.70**	0.76**	0.59**
下位群	0.43*	0.48*	0.52**	0.45*	0.74**
最下位群	0.02	0.24	0.77**	0.71**	0.83**

全被検児，聴力別，言語性IQ別の各群．**：P<0.01，*：P<0.05で有意な相関

3）相関係数の下位検査別集計

	動物の家	絵画完成	迷路	幾何図形	積木模様
平均	0.47	0.48	0.66	0.64	0.70
標準偏差	0.21	0.09	0.07	0.12	0.08
範囲	0.02～0.79	0.24～0.58	0.52～0.77	0.41～0.78	0.59～0.83

4）下位検査間の相関係数比較での有意差一覧

迷路，積木模様>動物の家*
迷路，積木模様>絵画完成**
幾何図形>絵画完成*

Uテスト，**：P<0.01，*：P<0.05で有意な相関

ある中等度群（平均106.4）では，「知識」は有意に「単語，理解」より得点が高かった．また言語性IQ別集計では，言語性IQが70以上の上位群，中位群，下位群では「知識」は「単語，理解」より有意に得点が高かった．このことから，言語性IQが70以上の難聴児では「知識」は有意に「単語，理解」より得点が高いといえる．

言語性IQと下位検査間の相関について，全体集計は下位検査と言語性IQとの相関係数は0.80～0.88の範囲であり，高い相関が示されている．このことは，言語性IQは個々の下位検査と密接な関連があること，すなわち個々の下位検査は言語性知能の各側面を確実に測定していることを示している．

聴力別集計では言語性IQと下位検査間の相関係数は平均r＝0.83（0.68～0.90）であり，全体的には高い相関が見られる．このことは，聴力が同程度の難聴児集団であっても，動作性IQや親の教育力などにより言語力に差が生じることを示している．

言語性 IQ 別集計では，上位群，下位群では「知識，算数，理解」と有意な相関があり，中位群は「知識，算数，類似」と有意な相関があり，最下位群では「理解」のみと有意な相関がある．このことは，言語性 IQ の程度と下位検査の得点が関連していることを示唆している．すなわち，言語力の乏しい難聴児では，「理解」の課題で少しでも答えられるようになると，言語性 IQ も高くなる傾向があることを示している．同様に中位群では「算数，理解，知識」の得点が，上位群，下位群では「知識，算数，理解」の得点が向上するなら，言語性 IQ に向上が見られることが示唆されている．

下位検査の得点による順位や言語性 IQ との相関の相違は，各下位検査の内容に関連していると思われる．

得点の高い「類似」は上位概念の理解を問う課題であるが，言語訓練のなかでしばしば行う「仲間集め課題（同じ仲間はどれ？）」と類似しており，療育を受けた難聴児には「答えやすい課題」である．

次に難聴児が得点しやすい「知識」も言語訓練のなかで語彙を増やす目的で頻繁に行う課題（これは何？）に類似しており，療育を受けた難聴児には「答えやすい課題」である．

「算数」は，課題を理解できたならば，難聴児にとって困難な課題とはいえない．しかし，文で提示される課題を聴き取れない，もしくは課題自体を理解できない場合には「算数」の得点は低くなる．言語力の低い場合でも，難聴児は最初の問 7 までの計数課題（2 個，4 個，9 個の積木の数を数えるなど）の正答は可能であろう．しかし，「問 8：9 個の積木のうち 4 個を残す」課題では，言語力の乏しい難聴児の返答は，4 個を検査者に渡すことである（正答は 5 個を渡すこと）．すなわち，「算数」であっても言語力が課題解決には不可欠な要素であることを示している．

「単語」は全体集計および聴力別集計では言語性 IQ と高い相関を示しているが，言語性 IQ 別集計ではいずれの群でも有意な相関が見られない．このことは，言語力がほぼ同レベルの難聴児では，「単語」以外の課題で言語力に差が生じることを示している．

「理解」は，言語性 IQ 別中位群を除き，すべての群で言語性 IQ と有意な相関がある．特に最下位群では言語性 IQ と有意な相関は「理解」のみである．「理解」の課題では，設問である比較的長い文を聴き取り，文レベルの発話で応答することが求められる．「理解」の課題は，難聴児にとって単に知識を増やす，応答の仕方を習得するだけでは得点できない課題である．言語性 IQ 別の上位群であっても平均得点は 9.8 と低いことから，難聴児にとって明らかに「不得意な課題」であることが示された．

3．WPPSI 動作性下位検査の検討

動作性検査の全体集計では，動作性下位検査の得点が最も高い「迷路（12.6）」と最も低い「絵画完成（11.4）」について，2 つの下位検査間の差（1.2）に統計学的には有意差が見られた．しかし，他の下位検査間では得点差は見られなかった．また聴力別および言語性 IQ 別集計では，どの群でも動作性下位検査間に有意差は見られなかった．このことから，動作性下位検査間でははっきりした得点差が見られないものの，「迷路」は難聴児にとって「絵画完成」より応答しやすい課題と思われる．

動作性 IQ と下位検査得点の関係について，言語性 IQ 別最下位群での「動物の家：$r=0.02$，絵画完成：$r=0.24$」を除いた全体集計，聴力別集計，言語性 IQ 別集計で動作性 IQ と下位検査得点とに有意な相関が認められる．ただし，相関の程度は「迷路，積木模様」が「動物の家，絵画完成」よりも全般に高くなっている．このことは動作性 IQ と「動物の家，絵画完成」の相関図での得点は，動作性 IQ と「迷路，積木模様」の相関図での得点より散らばりが大きいことを示している．このことから，「動物の家，絵画完成」の課題は個々の難聴児の得意，不得意で示される能力差が「迷路，積木模様」より出現しやすい課題と思われる．

「動物の家」は作業の速さ，正確さを求める課題であり，視覚的判断，作業課題の学習，作業の速さを測定することになる．反面，「積木模様」では時間制限はあるものの，作業の速さよりは正確さが求められる．日常の療育場面で同程度の能力の幼児では，「作業の速さ」を求める課題（カルタ取り，イスとりゲームなど）では個人差（目ざとい子と反応が鈍い子）が明らかになりやすいが，正確さのみを求める課題（ぬり絵，折り紙などの製作）では個人差が目立たないことと一致している．

「迷路」は設問の背景となる知識は不必要であり，視覚的に判断するだけで解決できるため，難聴児にとって応答しやすい課題と思われる．一方「絵画完成（間違い探し）」は，設問にある対象物（例：ドア，シーソー）についての知識（どのような構造になっているか，どのように機能するか）が必要なことから，単なる視覚的な判断だけでは応答できない課題である．このような課題の特性が，全体集計で「迷路」と「絵画完成」とに有意な得点差を生じさせたと思われる．

4．文献的検討

WPPSI検査について，現実に多くの難聴幼児通園施設，児童相談所，教育機関等で難聴児に実施され，また検査結果を基に進路が検討されている[10,11]．またWPPSI検査が妥当性，信頼性のある検査であることを前提に，難聴児のWPPSI検査データを使用して研究が行われている[12-14]．しかし，文献検索上はWPPSI検査の下位検査プロフィールと難聴児の発達特徴とを関連づけた研究を見出すことができなかった．そこで，聴覚言語法による難聴児教育の専門誌である「The Volta Review」の5～6歳の難聴児を対象とした最近の論文のなかで，どのような発達検査が使用されているかを調べた[15-17]．その結果，これらの論文ではPeabody Picture Vocabulary Test (PPVT)，sequenced inventory communication development，reynell developmental language scalesなどの言語発達検査を使用しており，認知発達の評価に関しては，心理担当者(school psychologist)，親，幼稚園教員の報告の引用だけであった．さらに難聴児に関する専門書である岡田[18]，Marschark M[19]，Meadow KP[20]，住[21]，吉野[22]の著書には，いくつかのWISC検査を使用した1960～1980年代の難聴児の古典的な研究が記載されているが，WPPSI検査に関する記述はなかった．

上述の成書に記載されている研究の一つで，アメリカの6歳のろう学校在籍難聴児を対象としたSisco FH，Anderson RJの研究（1978年）によると，難聴児は健常児に比べWISC-R動作性検査（1974年版）では「組み合わせ」の得点が高く，「絵画完成，符号」の得点が低いとしている[23]．しかしながら，アメリカで公立学校での早期療育が本格的になるのは1990年代以降の新生児聴覚スクリーニングの普及が契機になっており，1990年代以前は3歳以降ろう学校で教育を開始する体制であった[24]．Siscoらの研究で対象としたろう学校在籍難聴児は，早期療育を含め，主にトータルコミュニケーションで教育されている難聴児と推定される．

これらのことから，本研究で対象とした聴覚口話法または聴覚言語法による早期療育を受けた難聴児と，先のSiscoらのWISC-R検査の研究で対象とした難聴児とは明らかに療育方法で相違している．またWISC-R検査とWPPSI検査の動作性下位検査で共通しているのは，「絵画完成，積木模様」の2課題であり，他の3課題（WISC-R検査：絵画配列，組み合わせ，符号，WPPSI検査：動物の家，迷路，幾何図形）は明らかに検査内容に相違が見られる[2,3]．このように，対象とした難聴児および検査内容に相違があることから，上記のWISC-R動作性下位検査の分析結果と本研究でのWPPSI動作性下位検査の分析結果を比較することは適切ではないと思われる．

WPPSI検査が難聴児の研究に使用されていない理由として，欧米では資格の関係上心理職（psychologist）がWPPSI検査を行っており，オージオロジスト(audiologist)や言語聴覚士（speech pathologist）は難聴児の評価に前述の言語発達検査を使用すること，さらに心理職はSchum Rが指摘するようにWPPSI検査以外の非言語性検査を使用する傾向のあることが背景にあると思われる[7]．

5．今後の課題

本研究はすべて同一施設で聴覚を最大限活用する療育を受けた難聴児の分析データを基にしている．本研究で得られた知見を一般化するためには，広く他施設で療育を受けた難聴児のデータを集め，比較検討することが必要である．このためには他施設との共同研究が必要であり，今後の課題である．

本研究では，療育開始の条件を統制するために，療育開始年齢を3歳11ヵ月以下とし，また知的発達の遅れの要因を除外するために動作性IQが90以上とした．今後は療育開始が4歳以上の難聴児，動作性IQ 90未満の難聴児，自閉症等の他障害を合併する難聴児，および特異的言語障害児（SLI児）との比較を通じて，本研究で得られた知見の検証を予定している．

結　　論

早期療育を修了した6歳の難聴児に行ったWPPSI言語性下位検査では，「類似」の評価点が最も高く，「理解」の評価点が最も低かった．このことから，「類似」課題は療育を受けた難聴児にとって得意な課題であり，「理解」は不得意な課題であることが示された．また，言語性IQが70以上の難聴児では「知識」は「類似」の次に得意な課題であることが示された．

動作性下位検査について，難聴児にとって「迷路」は応答しやすい課題であり，「動物の家」および「絵画完成」は個々の難聴児の得意・不得意で示される能力差が出現しやすい課題であることが示された．

これらの知見は，難聴児のWPPSI検査結果を解釈・判定するうえで有用であると思われる．

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付表　WPPSI 知能検査下位検査の説明

Ⅰ．言語性下位検査（番号は検査実施順を示す）
1）知識：一般的な知識を問う課題（23 問）
　　例：「耳はいくつありますか.」,「パンは何から作りますか.」
3）単語：語の意味を問う課題（22 問）
　　例：「帽子って何ですか.」「お城って何ですか.」
5）算数：計数および簡単な加算減算の課題（20 問）
　　例：「積木を 9 個数える.」
　　　「本が 2 冊と 3 冊で何冊になりますか.」
8）類似：上位概念もしくは共通事項を問う課題（16 問）
　　例：バスのほかにどういう乗り物に乗りますか.
　　　：ネコとネズミはどのように似ておりますか.
10）理解：日常生活で生じる問題の解決方法や社会的ルールを問う課題（15 問）
　　例：指をけがしたとき，どうしたらよいですか.
　　　：家に窓があるのはどうしてですか.
II．動作性下位検査の説明
2）動物の家：見本に従い 4 色ペグのうち 1 つの色ペグを穴に差し入れる課題（20 穴）
　　完成までの時間と正確さで採点する.
4）絵画完成：絵の誤り部分を指で指摘する.（23 問）
　　言語表現は不要.
6）迷路：完成までの時間と正確さで採点する.（10 問）
7）幾何図形：見本の図形（○，□，×など）を模写する.（10 問）
　　正確さで採点する.
9）積木模様：見本と同じ模様を 4 つの積木で作る.（10 問）

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ORIGINAL ARTICLE

Vestibular-evoked myogenic potentials in cochlear implant children

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Abstract

Conclusions. Our results suggest that the sacculi of most children with cochlear implants can easily be damaged, as shown by the absence of vestibular-evoked myogenic potentials (VEMPs) in response to click stimuli. Also, in most of the children, the vestibular nerve was seemingly not stimulated by the cochlear implant. These results suggest that electrical stimulation at the comfortable level can stimulate the cochlear nerve; however, this stimulation did not spread to the vestibular nerve in our children. In some children with Mondini dysplasia or vestibulocochlear nerve abnormality, the vestibular nerve was stimulated when the cochlear implant device was on, because of a VEMP response to electrical stimulation. *Objective*. To clarify the diagnostic value of VEMPs in cochlear implant patients. *Material and methods*. The click-evoked myogenic potentials of 12 children who underwent cochlear implantation surgery were investigated. The latency and amplitude of the VEMP responses were measured. *Results*. Before surgery, 6 of the 12 children showed normal VEMPs, 1 showed a decrease in the amplitude of VEMPs and five showed no VEMP response. After surgery, with the cochlear implant device off, 1 child showed a decreased VEMP and 11 showed no VEMPs. With the cochlear implant device on, four children showed VEMPs and eight did not.

Keywords: *Children, click, cochlear implant, electrical stimulation, sacculus, vestibular-evoked myogenic potential, vestibular nerve*

Introduction

Vestibular-evoked myogenic potentials (VEMPs) in response to click or short tone-burst stimuli have been used as a clinical test for the inferior vestibular nerve [1–4]. It has been suggested [5–7] that VEMPs originate from the otolith organ, particularly the saccule. In VEMPs, galvanic stimulation of the mastoid instead of the use of clicks or short tone bursts can also elicit myogenic responses from the sternocleidomastiod muscle (SCM) [8–10]. Galvanic stimulation of the mastoid is considered to stimulate the most distal portion of the vestibular nerve [8,10], but not the sacculus.

The cochlear function of both ears is impaired in cochlear implant candidates. Patients with cochlear implants can hear speech sounds, which are converted to electrical signals in the speech processor, and these signals are transmitted to the internal receiver under the scalp and conducted to the electrodes in the cochlea. Thus, cochlear nerves that are stimulated electrically convey information to the central auditory system. Cochlear implants may affect the vestibular system by means of either pathologic disruption of the sensory vestibular functions of the labyrinth or fluctuating vestibular vestibulopathy or by electrical stimulation of the vestibular system [11]. However, VEMPs have not yet been studied in cochlear implant patients. The purpose of this study was to determine the presence or absence of VEMPs in cochlear implant children when the device was off or on, and to investigate saccular function in these children.

Material and methods

Subjects

The subjects comprised 12 children (7 boys, 5 girls; mean age 3.8 ± 1.4 years; range 2–7 years) who underwent cochlear implantation (CI) surgery at the University of Tokyo Hospital. Mean pre- and

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postoperative hearing levels were 108.3±14.2 and 45.8±8.4 dB, respectively. As controls, 9 healthy volunteer children (5 boys, 4 girls; mean age 5.1±3.0 years; range 8 months to 10 years) also participated in the study.

Procedure

The children were placed in the supine position. The active electrode was placed over the upper half of the SCM, the reference electrode on the upper sternum and the ground electrode on the midline of the forehead. During recording, the children were instructed to lift their heads up to induce hypertonicity of the SCM.

Recordings

The electromyographic signal from the stimulated side was amplified and averaged using a Neuropack evoked potential recorder (Nihon Kohden Co., Ltd., Tokyo, Japan). Electromyographic activities at a constant level were recorded for each child. Rarefaction clicks (0.1 ms, 95 dB normal hearing level) were presented through headphones (type DR-531; Elega Acous Co., Ltd., Tokyo, Japan). The stimulation rate was 5 Hz, the band-pass filter intensity 20–2000 Hz and the analysis time 50 ms. VEMPs in response to 100 stimuli were averaged twice. VEMPs were recorded both before and after the operation. After the operation, VEMPs were recorded when the device was both off and on.

Measurements

We analyzed the amplitude of the first positive–negative peak, p13–n23, ipsilateral to the stimulated ear, and the latencies of p13 and n23. The average of two runs was taken for the amplitudes and latencies. An absolute value of the VEMP amplitude ratio of <0.5 was considered a decreased VEMP.

Results

Controls

Clear VEMPs were obtained from all the healthy children. The mean±SD amplitude of p13–n23 was 181.0±90.9 μv. The mean±SD latency of p13 and n23 were 10.5±0.5 and 16.1±1.3 ms, respectively.

Patients

All the children underwent CT and MRI of the ear. Four of the 12 children showed abnormalities in the CT or MRI scans, as described below.

Case 2. MRI scans demonstrated that only one branch of the vestibulocochlear nerve appeared in the internal auditory meatus.

Case 3. CT demonstrated Mondini dysplasia, with only one turn in the cochlea and the enlarged basal turn and vestibule forming a common cavity. It was also noted that communication between the internal auditory meatus and cochlea was present. The lateral semicircular canal seemed to form a sac. The superior and posterior semicircular canals were widened (Figure 1).

Case 4. CT demonstrated Mondini dysplasia, with only two turns in the cochlea. There was no distinct visualization of the apical and middle turns in the cochlea, which seemed to form a common sac. The basal turn appeared normal, but showed communication between the internal auditory meatus and cochlea. The vestibular aqueduct was enlarged (Figure 2).

Case 5. CT demonstrated large vestibular aqueduct syndrome (LVAS), but the cochlea and internal auditory meatus appeared normal.

The VEMP results of the CI children are summarized in Table I.

VEMPs and caloric tests before operation. Ten of the 12 children underwent caloric tests with irrigation of ice water. Four children showed normal caloric test results and VEMPs. Four children showed poor responses in the caloric test: one showed a decrease in the VEMP; one showed a normal VEMP; and two showed no VEMP response. Two children showed no response to either caloric or VEMP testing.

Figure 1. CT scan of Case 3. Left ear of a case of Mondini dysplasia. Note the presence of a common cavity, an enlarged basal turn of the cochlea and communication between the internal auditory meatus and cochlea. C = cochlea; V = vestibule; IAM = internal auditory meatus.

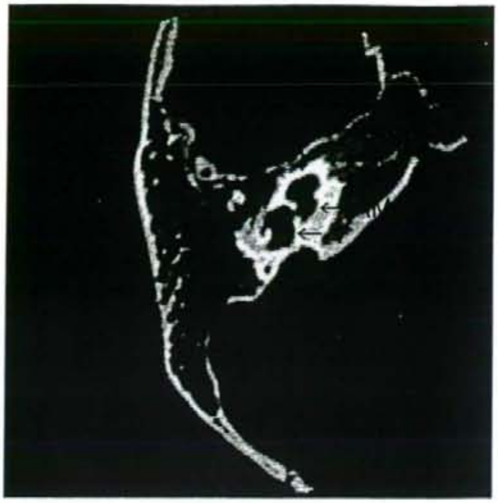

Figure 2. CT scan of Case 4. Large vestibular aqueduct of the right ear. Note the presence of communication between the internal auditory meatus and cochlea. VA = vestibular aqueduct. See Figure 1 for other abbreviations.

VEMPs before and after CI when the device was off. Six of the 12 children showed normal VEMPs before operation; 1 of these children showed a decrease in the VEMP and 5 showed no VEMP response after CI (Figure 3). Five of the 12 children showed no VEMPs both pre- and postoperatively (Figure 4). One child showed a decrease in the VEMP before operation, but this decrease was abolished after operation (Figure 5).

Comparison of VEMPs when the cochlear implant device was off and on. One child showed a decrease in the VEMP when the cochlear implant device was off and an increase when it was on. Eleven of the 12 children

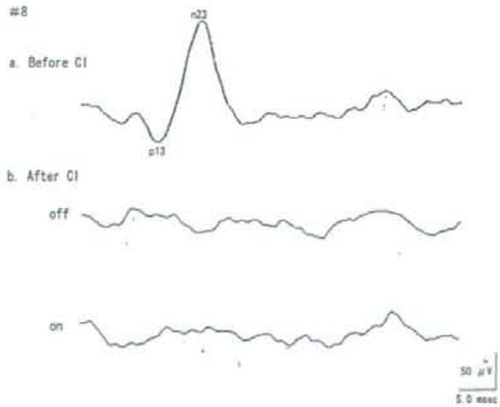

Figure 3. VEMP of Case 8. Normal VEMP before CI, absence of VEMP after cochlear implantation and absence of response on cochlear implant device activation.

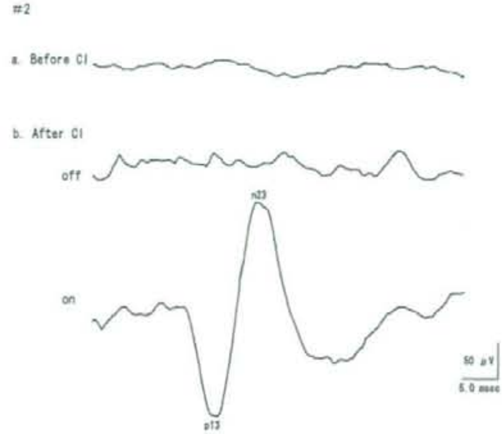

Figure 4. VEMP of Case 2. Absence of VEMP before and after CI and appearance of VEMP response on cochlear implant device activation.

did not show VEMPs when the cochlear implant device was off: 3 elicited VEMP responses (Figures 4 and 5) and 8 showed no VEMP response when it was on (Figure 3).

Discussion

Anatomically and phylogenetically, the cochlea and vestibular organs are closely related. They share a continuous membranous structure and have similar receptor cells. Therefore, inner ear diseases are likely to affect not only hearing but also the sense of balance. Regarding the ice water caloric test, our study demonstrated that 6 children (60%) had caloric hypofunction or areflexia and that 4 (40%) were normal. These results are similar to those of Ito [12], who found that, before surgery, 67% of

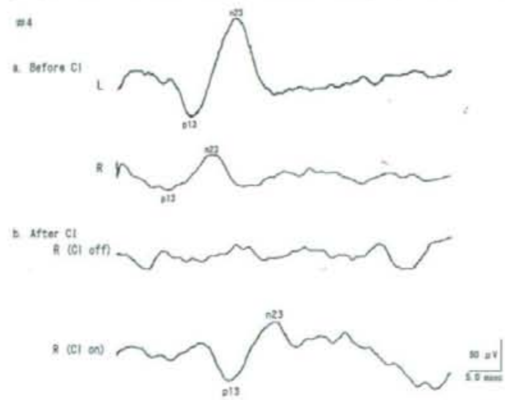

Figure 5. VEMP of Case 4. Decreased amplitude of VEMP before CI in right ear and absence of VEMP after surgery, and appearance of VEMP response on cochlear implant device activation.

Table I. Patient characteristics and data.

Patient No.	Sex	Age at CI (years)	Remarks	Before CI			After CI			
									VEMP	
				Hearing level (dB)	Caloric test	VEMP	Hearing level (dB)	Time lapse	Device off	Device on
1	M	3		111.25	+	+	43.75	2 months	Decreased	+
2	M	2	Auditory nerve abnormality	83.75		–	67.5	2 months	–	+
3	M	3	Mondini dysplasia	111.25	Poor	–	45	3 years	–	+
4	M	5	Mondini dysplasia	118.75	Poor	Decreased	45	1 month	–	+
5	M	3	LVAS	93.75	+	+	31.25	23 days	–	–
6	F	3	CMV	115	–	–	52.5	1 year	–	–
7	F	7		105	–	–	42.5	1 year	–	–
8	F	3		97.5	+	+	42.5	3 months	–	–
9	F	4		97.5	+	+	41.25	2 months	–	–
10	M	5		105	Poor	+	46.25	2 months	–	–
11	F	3		132.5	Poor	–	45	6 months	–	–
12	M	4		128.3		+	47.5	1 year	–	–

CMV = cytomegalovirus.

cochlear implant patients showed hypofunction or the absence of function in the caloric test. Buchman et al. [11] reported that nearly 70% of cochlear implant children had absent or low-intensity responses to caloric irrigation before operation.

Before operation, the VEMP test demonstrated a decrease in or the absence of VEMP responses in 6 children (50%) and normal responses in 6 (50%). The proportion of abnormal responses was therefore lower for the VEMP than for the caloric tests. These results are in agreement with the caloric test results of Tribukait et al. [13], in which the subjective visual horizontal and VEMPs of deaf children were slightly lower for the otoliths than for the semicircular canals.

Saccule effects of CI

The effects of CI on the vestibular system have been shown in numerous studies. Vestibular symptoms have been reported to occur in 0.33–70% of patients [14]. Ito [12], in a review in which he summarized the results of published electronystagmography studies, demonstrated that caloric responses changed in 71 patients (38%) and vestibulo-ocular reflex responses were reduced in as many as 40% of patients after CI. Mangham [15] reported similar findings for rotational chair testing.

Pathologic analysis of the vestibular apparatus from human temporal bones after CI has been reported [16]. Significant histopathologic damage of the vestibular end-organs was noted in 6 patients (54.5%). Fibrosis of the vestibular apparatus, saccular membrane distortion, osteoanagenesis and reactive neuromas have all been observed in patients who have undergone CI. Involvement of the scala vestibuli, as a result of damage to the osseous spiral lamina or basilar membrane in the cochlear basal turn, correlated strongly with vestibular end-organ damage.

In this study, of 12 children, 5 showed the absence of VEMPs and 2 showed a decreased VEMP amplitude after operation when the cochlear implant device was off. These results show a reduction in saccular function in 7/12 children (58.3%) after CI. Tien and Linthicum [16] suggested that the saccule is more susceptible to damage than the utricle or semicircular canals because of its proximity to the insertion pathway of the electrodes. Basilar membrane penetration was observed in 6/8 cochlear bones when a standard cochleostomy diameter (≈0.8 mm) was used. When the surgical technique was modified by making a slightly larger cochleostomy hole (≈1.8 mm) closer to the round window, the frequency of basilar membrane penetrations was restricted to two of seven bones [17].

Vestibular stimulation as a result of CI

The occurrence of vestibular stimulation as a result of CI has been shown in many studies. Black et al. [18] described two patients who experienced vestibular stimulation on initial activation of their extracochlear implants. Hoffman and Cohen [19] reported 11 patients (0.36%) who experienced device-related dizziness. Wong et al. [20] observed nystagmus in a child after the activation of a multichannel cochlear implant device using increased pulse widths (200 μs) in the monopolar stimulation mode. Bance et al. [21] described a patient who experienced significant vestibular signs during stimulation associated with a loud noise and a "shock-like" sensation. In the same study, it was found that 1/17 patients (5.9%) experienced a totally asymptomatic vestibular stimulation by their implant. Brey et al. [22] evaluated a group of 22 patients by means of pre- and postoperative posturography. In their study, postural stability worsened in one patient and improved in two after CI when the device was activated. Vestibular stimulation is possible in some patients with cochlear implant.

Recently [8,9] it has been shown that galvanic stimulations can also evoke myogenic responses in the SCM, but these myogenic potentials disappear after the activation of a selected vestibular nerve section. Goldberg et al. [23] suggested that galvanic stimulation stimulates the most distal portion of the vestibular nerve and predicted that galvanic-evoked myogenic responses would allow a better assessment of vestibular function, including the differentiation of conditions primarily involving the end-organs from those primarily involving the nerve [9]. Murofushi et al. [10] stated that the recording of galvanic-evoked myogenic responses of the SCM is useful in the differential diagnosis of labyrinthine lesions from retrolabyrinthine lesions in patients without click-evoked vestibulocollic reflexes.

The results of this study indicated that 4/12 children (33.3%) showed VEMPs when the cochlear implant device was activated. Among these four children, one showed decreased VEMPs and two showed no VEMPs before the operation; one showed decreased VEMPs and three showed no VEMPs when the device was off. Although saccular functions were damaged before surgery, VEMP responses were still elicited upon activation of the device. This suggests that VEMPs are induced by activation of the device as a result of stimulation of the vestibular nerve.

Of the 12 children, 8 showed no VEMPs when the cochlear implant device was activated, although 4 showed VEMPs before CI surgery. This finding suggests that electrical stimulation at the C level stimulated the cochlear nerve, but that the stimulation did not spread to the vestibular nerve in these patients.

Although the reasons for these differences are not totally evident, differences in patient selection, surgical procedures, cochlear implant devices, current intensity, stimulation schemes, testing paradigms and anatomical features may all be important factors.

Our study demonstrated that two children with Mondini dysplasia and one child with vestibulocochlear nerve abnormality showed VEMP responses on activation of the cochlear implant device. Sennaroglu et al. [24] reported on vestibular stimulation in the form of nystagmus in a child with a common cavity. Mondini dysplasia in our cases was characterized by the presence of communication between the internal auditory meatus and cochlea. It is possible that cochlear implant device activation may elicit VEMP responses in children with Mondini dysplasia. In one case in our study, only one branch of the vestibular nerve appeared in the internal auditory meatus and showed VEMP responses. This suggests that the vestibular nerve was intact in this child, and that the branch of the vestibulocochlear nerve that was visible on MRI scans was a vestibular nerve.

Finally, we would like to emphasize that VEMP testing is also a useful tool for diagnosing saccular function in cochlear implant patients. The sacculi of most children with cochlear implants were easily damaged, resulting in the absence of VEMP. These results suggest that electrical stimulation at the C level stimulated the cochlear nerve; however, it did not spread to the vestibular nerve in most patients. Nevertheless, it is especially noteworthy that in some children with Mondini dysplasia or vestibulocochlear nerve abnormality, the vestibular nerve can be stimulated by CI.

Acknowledgements

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ORIGINAL ARTICLE

Changes in auditory behaviors of multiply handicapped children with deafness after hearing aid fitting

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Abstract

Conclusion. The early diagnosis of deafness and the early fitting of hearing aids in multiply handicapped children are recommended for language development in these children even when their neurological or mental status is poor. *Subjects and methods.* The subjects consisted of 5 hearing-impaired infants with no other problems and 28 hearing-impaired children with multiple handicaps. Behavioral audiometry and auditory brainstem responses were used for evaluating hearing impairment. The 5 hearing-impaired infants with no other problems underwent hearing aid fitting at approximately 1 year of age and the 28 hearing-impaired children with multiple handicaps underwent hearing aid fitting at various times from 1 to 5 years of age. The effects of their hearing aids were compared on the basis of auditory behavioral changes. The developmental scale of auditory behaviors in infancy that we proposed was introduced to evaluate the development of auditory behaviors. *Results.* The auditory behaviors of the hearing-impaired children with no other problems showed constant changes with age after hearing aid fitting. However, among the 28 hearing-impaired children with multiple handicaps, 17 showed improvement in auditory behaviors, 5 showed no improvement in auditory behaviors because of the associated severe motor and mental retardation, and epilepsy, and 6 were unable to adapt to wearing hearing aids.

Keywords: *Auditory behaviors, deafness, hearing aid, multiply handicapped children*

Introduction

There are various diseases that cause congenital and acquired deafness and it has been clearly demonstrated that early detection and intervention for hearing problems in neonates, infants, and children are important issues for these children's later language development. Because of deafness, these children's hearing and communication skills will be seriously affected if they are not fitted with hearing aids and given auditory training. However, multiply handicapped children with hearing impairment have very often been ignored in terms of providing better hearing because of the absence of reliable audiometry. Since auditory brainstem response (ABR) testing was introduced as an objective audiometry method, hearing problems have become easy to detect in multiply handicapped children. Here we report a comparative study of the development of auditory behaviors between children with no other problems and children with multiple handicaps after hearing aid fitting.

Subjects and methods

Subjects

The subjects were 33 infants and children with deafness who were divided into 2 groups, i.e. type I and II. Type I comprised five hearing-impaired infants with no other problems. The age of hearing aid fitting was approximately 1 year. Type II comprised 28 hearing-impaired children with multiple handicaps aged 1–5 years (Table I).

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Table I. Profile of 28 hearing-impaired infants with multiple handicaps.

Underlying disease	Number of children affected
22 infants who were able to use hearing aids	
Cerebral palsy (athetosis type)	6
Cocaine's syndrome	3
Cerebral palsy (spastic type)	3
Mental retardation	3
Mental retardation with epilepsy	2
Spina bifida	2
Meningitis	1
Encephalitis	1
Systemic bone disease	1
6 infants who were unable to adapt to wearing hearing aids	
Cornelia de Lange syndrome	2
Mental retardation with physical handicaps	2
Cerebral palsy with epilepsy (spastic type)	1
Cerebral palsy (athetosis type)	1

Methods

Behavioral audiometry and ABRs were used for evaluating hearing problems. Our scale of developmental auditory behaviors in infancy (information was obtained from the mothers) was used for evaluation after hearing aid fitting [1,2]. Hearing aid fitting was performed on type I and II subjects by audiologists.

Results

Our scale of developmental auditory behaviors was developed on the basis of apparent responses to environmental sounds or voices and changes in the quality or amount of vocalization.

As regards type I, Figure 1 shows developmental changes in the auditory and communication behaviors of the five hearing-impaired infants with no other problems. For type II, Table I shows profiles of the 28 hearing-impaired children with multiple handicaps. Among them, 17 showed improvement in auditory behaviors, whereas 5 showed no improvement, and 6 were unable to adapt to wearing hearing aids.

Figure 2 shows developmental changes in auditory behaviors in six typical children with multiple handicaps. The effects of hearing aids were evaluated in terms of the responses to sounds or voices and changes in the quality or amount of vocalization.

Figure 3 shows a statistical comparison of developmental changes in auditory behaviors between types I and II. However, in Figure 3b five children with associated severe motor and mental retardation

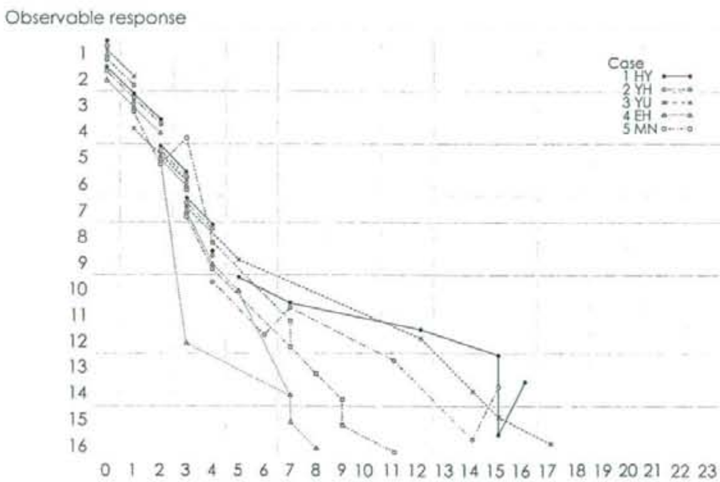

Figure 1. Type I. Developmental changes in auditory behaviors of five hearing-impaired infants with no other problems with approximately 90 dB hearing loss at the age of 1 year who were fitted with hearing aids. The changes in auditory behaviors of each subject are connected by a line. The first column indicates auditory behaviors and the first row horizontal scale indicates the time course after the first hearing aid fitting. Observable responses: 1, aware of sound; 2, increase in vocalization; 3, imitating intonation; 4, turning head for calling; 5, understanding words; 6, watching mother's mouth; 7, understanding sound; 8, reply for calling; 9, sound localization; 10, watching speaker's mouth; 11, sound localization out of sight; 12, response to verbal commands; 13, enjoy music; 14, ask object's names; 15, speak words; 16, speak two-word sentences.

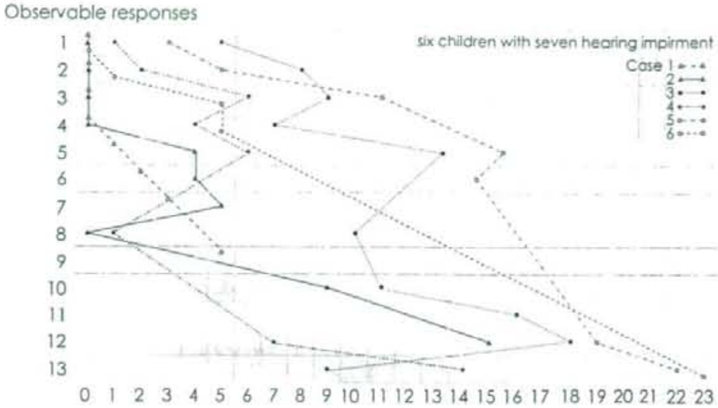

Figure 2. Type II. Developmental changes in auditory behaviors of six typical hearing-impaired children with multiple handicaps who were fitted with hearing aids. The first column indicates auditory behavior items, and the first row indicates the time course after the first hearing aid fitting. The changes in auditory behaviors of each subject are connected by a line. Observable responses: 1, more sensitive response to environmental sounds in daily life; 2, increase in vocalizations; 3, utterance of more varied kinds of vocal sounds or with inflections; 4, turning head when name is called; 5, the beginning of understanding a few words; 6, imitating speaker's mouth shape; 7, the beginning of understanding of meaning of sounds; 8, saying 'yes' or raising hands when name is called; 9, recognizing the location of different sounds; 10, watching speaker's mouth shape; 11, identifying the source of sounds that are out of sight; 12, response to simple verbal commands; 13, identifying favorite music.

and epilepsy who showed no improvement in their auditory behaviors and in Figure 3c the six children who were unable to adopt wearing hearing aid fitting were not included.

Discussion

In our study of developmental changes in auditory behaviors, the 5 hearing-impaired infants with no other problems and 28 multiply handicapped children with hearing impairment showed differences after hearing aid fitting, during the follow-up study. Our study revealed that handicapped children with severe motor and mental retardation or epilepsy due to congenital or acquired diseases were delayed in their acquisition of hearing and speech abilities despite a successful hearing aid fitting, showed no improvement in auditory behaviors, or were unable to adopt wearing hearing aids at all.

Here, it is very important to use our scale of developmental changes in auditory behaviors to observe normal babies and infants with hearing impairment. Immediately after birth, normal babies react to sudden sounds with Moro's reflex, and after a couple of months they babble and laugh when their mothers talk. Four months after birth, babies turn their head toward various sounds around them. Eight months after birth, infants perceive low-frequency sounds around them and 10 months after birth infants begin to imitate simple words such as 'Mom' [1,2].

Hearing impairment prevents developmental changes in auditory behaviors, including hearing and speech development. Hearing-impaired babies and infants with no other abnormalities can react to a calling voice or a ringing phone, and a couple of months after being fitted with a hearing aid frequently increase their amount of vocalization. Their behavioral changes are apparently observed. However, the developmental changes in auditory behaviors of hearing-impaired children with multiple handicaps is very slow or seriously delayed because of complications such as motor and mental retardation and epilepsy (Figures 2 and 3). In some children with severe mental retardation, hearing aids are useless or not used. In these hearing-impaired children, careful hearing aid fitting is encouraged and this fitting should not be precluded easily [3].

There are various etiologies of congenital deafness [4,5]. However, there is no therapeutic method for these diseases except the use of hearing aids or cochlear implants. Therefore, the goals of educational plans for hearing-impaired children with multiple handicaps are to accelerate the development of hearing and speech abilities using hearing aids for those with residual hearing and to stimulate the development of personality similarly to normal children of the same age. The achievement of language development in hearing-impaired children is the desired final goal but there are various results even if early hearing aid fitting and education are carried out as well. Then, the collaborative efforts of parents, doctors, speech pathologists, and teachers should be made as a team to improve the speech and communication skills of such children.

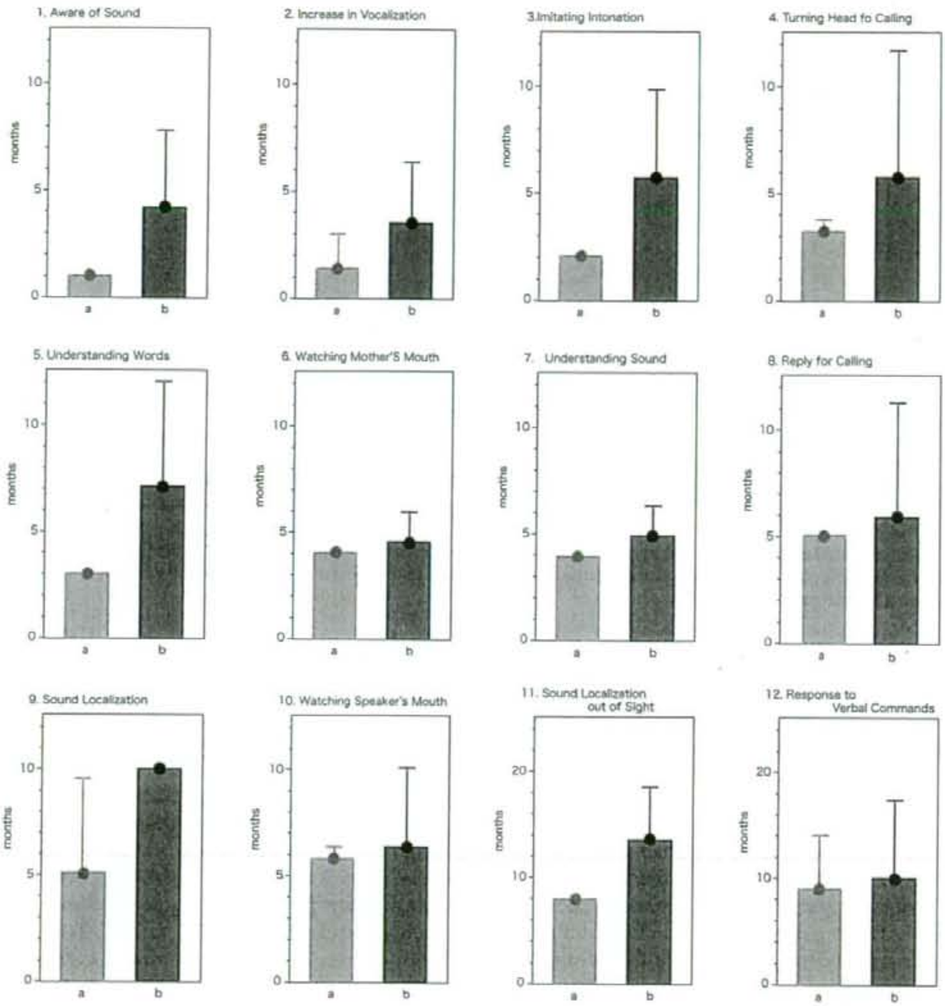

Figure 3. Statistical comparison of each auditory behavior item between 5 hearing-impaired children with no other problems and 17 hearing-impaired infants and children with multiple handicaps who showed hearing improvement. Sixteen auditory behavior items are compared using bar graphs. (a) Auditory behavior of hearing-impaired infants with no other problems ($n=5$). (b) Auditory behavior of hearing-impaired infants and children with multiple handicaps ($n=17$). With the exception of items 6, 7, and 12, all items are statistically significant.

Finally, we emphasize that the early diagnosis of hearing impairment and the early fitting of hearing aids in multiply handicapped children are recommended for the development of these children's communication skills, even if their neurological or mental status is not good.

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ORIGINAL ARTICLE

Assessment of vestibular function of infants and children with congenital and acquired deafness using the ice-water caloric test, rotational chair test and vestibular-evoked myogenic potential recording

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Abstract

Conclusions. The vestibular function can be assessed by ice-water caloric test, rotational chair test and VEMP recording in severely hearing impaired infants and young children, and 85% of these patients showed abnormal responses in these tests. *Objectives.* To evaluate the vestibular function of infants and young children with congenital and acquired deafness, we examined the semicircular canal and otolith function in their early childhood. *Materials and methods.* Our subjects were 20 children (11 boys, 9 girls; age range 31–97 months, mean age 54.2 months) with severe hearing impairment. Their vestibular functions were assessed by the ice-water caloric test, rotational chair test and vestibular-evoked myogenic potential (VEMP) recording. *Results.* Among these 20 severely hearing impaired children, only 3 (15%) showed normal responses in the caloric test, rotational chair test and VEMP recording bilaterally. Seven (35%) showed responses asymmetrically in the caloric test despite normal responses in the rotational chair test and VEMP recording bilaterally. Five (25%) showed hyporeflexia or areflexia in the caloric test bilaterally, but showed normal responses in the rotational chair test and normal reproducible or decreased VEMPs. Five (25%) showed no responses at all in the caloric test, rotational chair test and VEMP recording.

Keywords: *Children, hearing loss, vestibular function, semicircular canal, saccule, vestibular-evoked myogenic potentials*

Introduction

The cochlear and vestibular organs are closely related anatomically and phylogenetically, and the relationship of vestibular function with hearing impairment has been discussed. Since the 1950s, many studies have been performed to establish the incidence of vestibular pathology in children with congenital and acquired deafness [1–3]. Although these studies have employed the caloric test for the horizontal semicircular canal function, most of them did not evaluate the other two semicircular canals or otolithic organs, whose functions may have more influence on the development of postural control and locomotion than the horizontal semicircular canals alone.

The response in the rotational chair test had been considered to represent the bilateral horizontal semicircular canal functions. However, recent studies have shown that animals with all six semicircular canals plugged could gain vestibulo-ocular reflexes during rotations at higher frequencies [4,5]. From these results, there is a possibility that the otolith organ plays some role during rotations, and the rotational chair test may lead to the discovery of vestibular function from a new viewpoint.

Moreover, the recording of vestibular-evoked myogenic potentials (VEMPs) induced in response to clicks or short tone-burst stimuli has been carried out as a new clinical test for the inferior vestibular nerve function since 1990s, and results suggest that VEMPs originate from the otolith organ, particularly the saccule [6–8]. Investigation of vestibular function by the VEMP recording of the hearing impaired population would clarify the otolith function and the

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Table I. Results of vestibular and auditorial tests.

Patient no.	Remarks	Age at tests (months)	Hearing level (dB)		Caloric test		Rotation test		VEMP	
			Left	Right	Left	Right	Clock	Counter	Left	Right
Group A										
1	Congenital	31	106.3	106.3	+	+	+	+	+	+
2	Congenital	35	95.0	95.0	+	+	+	+	+	+
3	Congenital	40	107.5	111.3	+	+	+	+	+	+
Group B										
4	Progressive	59	87.5	92.5	L-CP	+	+	+	+	+
5	LVA	87	106.3	105.0	L-CP	+	+	+	+	+
6	LVA	46	96.3	96.3	+	R-CP	+	+	+	+
7	Progressive	97	87.5	110.0	+	R-CP	+	+	+	+
8	Congenital	40	105.0	105.0	+	R-CP	+	+	+	+
9	Congenital	80	118.8	105.0	+	R-CP	+	+	+	+
10	Congenital	54	107.5	111.3	+	R-CP	+	+	Decreased	+
Group C										
11	LVA	60	102.5	118.8	Poor	Poor	+	+	+	Decreased
12	Congenital	46	106.3	106.3	Poor	Poor	poor	+	+	Decreased
13	Congenital	39	OS	OS	–	–	+	+	+	+
14	Congenital auditory nerve disease	37	95.0	96.3	–	–	+	+	Decreased	+
15	Common cavity malformation	60	112.5	95.0	–	–	+	+	Decreased	+
Group D										
16	Rightt cochlear nerve malformation	42	OS	OS	–	–	Poor	Poor	+	–
17	Congenital	41	OS	OS	–	–	Poor	Poor	–	–
18	Progressive	64	90.0	106.3	–	–	–	–	–	–
19	Congenital CMV	62	OS	113.8	–	–	–	–	–	–
20	Meningitis	63	OS	OS	–	–	–	–	–	–

CMV, cytomegalovirus; LVA, large vestibular aqueducts.

relationship between vestibular and audiological functions further.

In children, because the vestibular function plays an important role in their gross motor development, otologists and audiologists should recognize and understand the high incidence of vestibular dysfunction in hearing impaired children and be prepared to undertake appropriate evaluations. However, there have been very few studies that investigated the entire vestibular function in deaf infants and young children. Tribukait et al. [9] assessed vestibular function in 36 deaf children by the caloric test, VEMP recording and body tilt test. In their study, 30% of the children had caloric hypo- or areflexia and 22% showed weak or no VEMPs bilaterally. However, the children in their study were over 15 years old.

Today, hearing problems can be detected mostly at the neonatal stage and early infancy, and total management including not only auditory but also vestibular function is needed. The purpose of this study was to evaluate the vestibular function of such deaf infants and young children, to establish the relationship between semicircular canal and otolith function in early childhood using the caloric test, rotational chair test and VEMP recording.

Materials and methods

Subjects

The subjects were 20 children (11 boys, 9 girls; age range 31–97 months, mean age 54.2 months) with a severe hearing impairment, fitted with hearing aids, and who planned to undergo cochlear implantation (CI) at the University of Tokyo Hospital. Before CI, their auditory and vestibular functions were evaluated. All the patients underwent audiological assessments including conditioned orientation reflex (COR) audiometry, play audiometry and standard pure tone audiometry. The audiological tests were selected for each of the patients depending on their capability to perform them. Their hearing levels ranged from 87.5 dB to off the scale, and all revealed a severe hearing loss bilaterally.

Concerning the etiology of hearing loss, 9 patients among the 20 children had congenital profound hearing loss, 3 had a progressive hearing impairment, 3 had large vestibular aqueducts (LVA) as revealed by high-resolution temporal bone computed tomography (CT) scan and inner ear magnetic resonance imaging (MRI). One patient had intrauterine cytomegalovirus infection and one had meningitis at the age of 5 years. One had congenital auditory nerve disease, one had common cavity

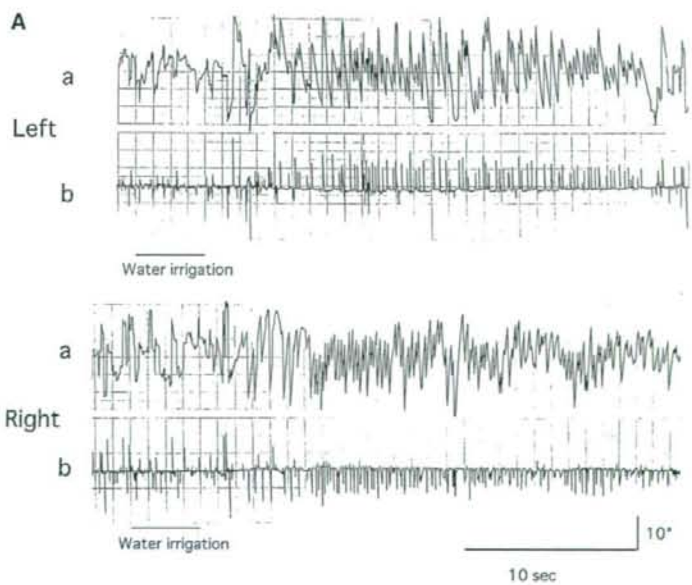

Figure 1. (A) Ice-water caloric test of patient no. 1. Bilateral normal responses are shown. Time constants: a, 3.0 s; b, 0.03 s. (B) Rotational chair test of patient no. 1. Both sides show normal responses. A, Time scale (one division per second); B, Angular displacement of eyes (time constant, 3.0 s; calibration signal, 10°); C, Rotational velocity of eyes (time constant, 0.03 s; calibration signal, 20°/s); D, Angular velocity of chair rotating. (C) VEMP recording of patient no. 1. Bilateral normal responses are shown.

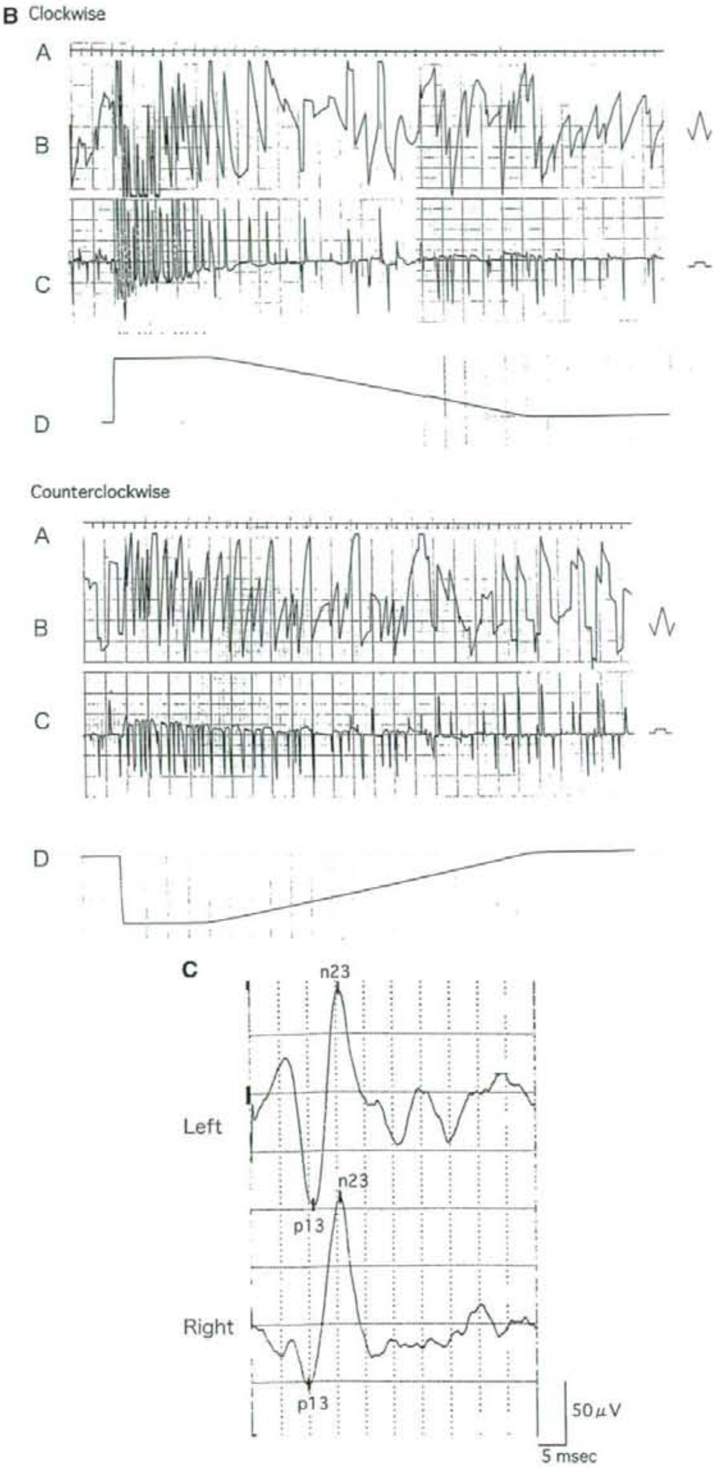

Figure 1 (*Continued*)

malformation in the inner ear and one had cochlear nerve malformation on the right side.

Methods

Vestibular assessment. The vestibular functions of the patients were evaluated by the following: ice-water caloric test, damped-rotational chair test and VEMP recording.

The ice-water (4°C and 2 ml) caloric test was performed to irrigate the external auditory meatus to induce a thermal gradient across the horizontal semicircular canal of one ear. Horizontal and vertical eye movements were recorded using standard ENG electrodes.

For the rotational chair test a rotational chair (Nagashima Co. Ltd, S-II) was accelerated to a maximum rotational velocity of 160°/s, then decayed by $4°/s^2$. The test was performed once in a clockwise direction and once in a counterclockwise direction. Eye movements were recorded by ENG and the duration and number of beats of perrotatory nystagmus were calculated for the evaluation of semicircular canals and otolith organs in both ears.

VEMPs were recorded using conventional VEMP recording procedures in our hospital [10]. Sound stimuli of clicks (0.1 ms, 95 dB nHL) were presented to each side of the ear through headphones. Electromyographic signals from the stimulated side of the sternocleidomastoid muscle were amplified. The stimulation rate was 5 Hz, the band-pass filter intensity was 20–2000 Hz, and the analysis time was 50 ms. VEMPs in response to 100 stimuli were averaged twice. VEMPs were considered significant when there was a reproducible short-latency biphasic wave (p13-n23). An absolute VEMP ratio of <0.5 was considered to indicate significant asymmetry. If the amplitude of p13-n23 was <50 μV on both sides, the subject was considered to have hypofunction bilaterally. VEMPs are responses of the sternocleidomastoid muscle via the saccules to clicks.

Results

Vestibular assessment

Table I shows the vestibular and auditorial test results of all the patients.

Caloric test

Three patients (15%) showed normal responses bilaterally. Seven patients (35%) showed asymmetrical responses, and two patients (10%) showed hypo-reactions in both ears. Eight patients (40%) showed no response bilaterally.

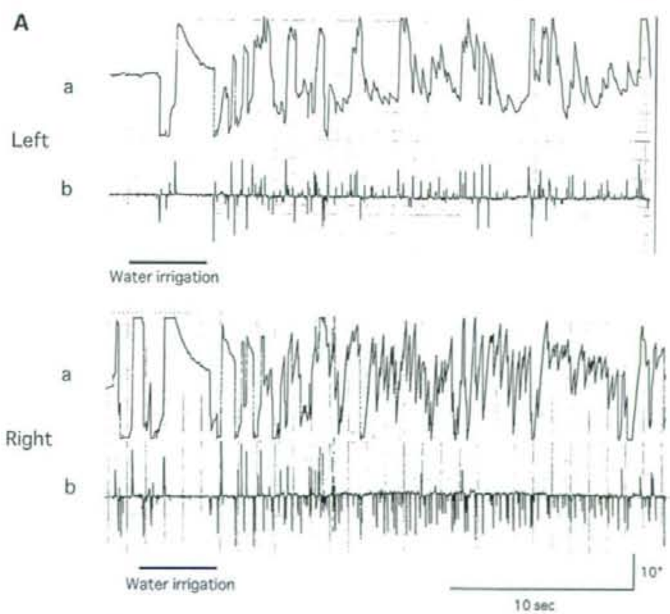

Figure 2. (A) Ice-water caloric test of patient no. 4. Canal paresis on the left side is shown. Time constants: a, 3.0 s; b, 0.03 s. (B) Rotational chair test of patient no. 4. Normal responses are shown. A, Time scale (one division per second); B, Angular displacement of eyes (time constant, 3.0 s; calibration signal, 10°); C, Rotational velocity of eyes (time constant, 0.03 s; calibration signal, 20°/s); D, Angular velocity of chair rotating. (C) VEMP of patient no. 4. Bilateral normal responses are shown.