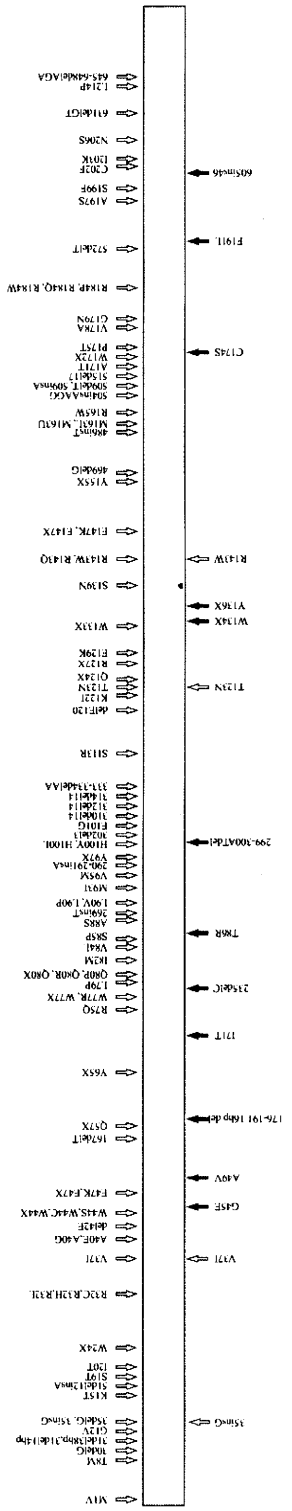

Figure 1. Differences in *GJB2* mutation spectra between Japanese populations and populations with European ancestry. Shaded arrows indicate mutations found in Caucasoid populations; filled arrows indicate mutations found in Japanese; white arrows indicate mutations reported in both Caucasoid and Japanese families.

microsatellite markers excluded a single founder mutation and therefore it is most likely a hot spot mutation [30]. In the case of the mitochondrial 1555A→G mutation, which has been reported in many ethnic populations, phylogenetic analysis of independent families with that mutation has suggested that it is sporadic and has multiplied through the evolution of the mtDNA [31]. Therefore, it is conceivable that this mutation does not have a common founder, but may be a mutational hot spot.

From the above facts, in the search for responsible genes in deafness patients, prior screening based on recurrent mutations and ethnic databases of deafness genes should be done. These facts indicate that each population may not have its own responsible genes, but may have its own mutational spectrum within the same catalog of deafness genes.

### Invader assay as a simultaneous mutation screening

Simultaneous technology, including the microarray approach as well as Invader assay, has developed rapidly and will likely facilitate great improvements in medical management and genetic counseling. The use of a microarray approach for multi-gene deafness mutation detection has been recently reported [32,33]. We have recently succeeded in screening deafness genes effectively by means of an Invader assay panel (including 41 known mutations of 9 known deafness genes) [4], enabling diagnosis of approximately 30% of the congenital hearing loss patients who had at least one mutation in *GJB2*, *SLC26A4*, and/or the mitochondrial 12S rRNA, in accordance with results in our mutation screening series. The Invader technology has excellent sensitivity and accuracy and is advantageous for the following reasons: 1) reduced contamination due to being PCR-independent, 2) extremely simple operation that can be learned quickly and does not require PCR-purification or post-hybridization washing skills, 3) detection of low-level heteroplasmy, and 4) amenability to automation. Genetic screening based on the mutation spectrum of a corresponding population may be an appropriate and effective strategy for detecting mutations in causative deafness genes.

Although mutation spectra have been demonstrated to be dependent on each population, mutations within the prevalent four genes have been extensively reported across many populations and are currently recognized to be the most common deafness genes worldwide. Therefore, the present results are consistent with a series of reports and

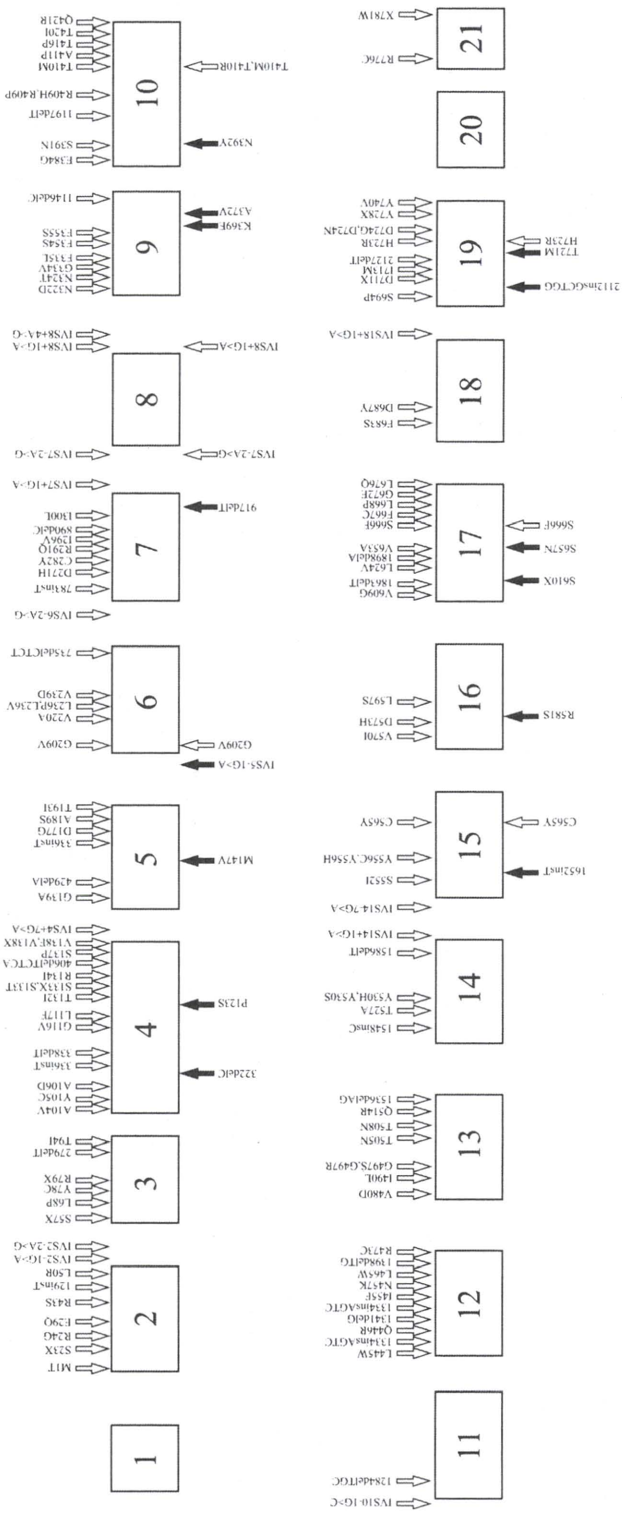


Figure 2. Differences in *SLC26A4* mutation spectra between Japanese populations and populations with European ancestry. Shaded arrows indicate mutations found in Caucasoid populations; filled arrows indicate mutations found in Japanese; white arrows indicate mutations reported in both Caucasoid and Japanese families.

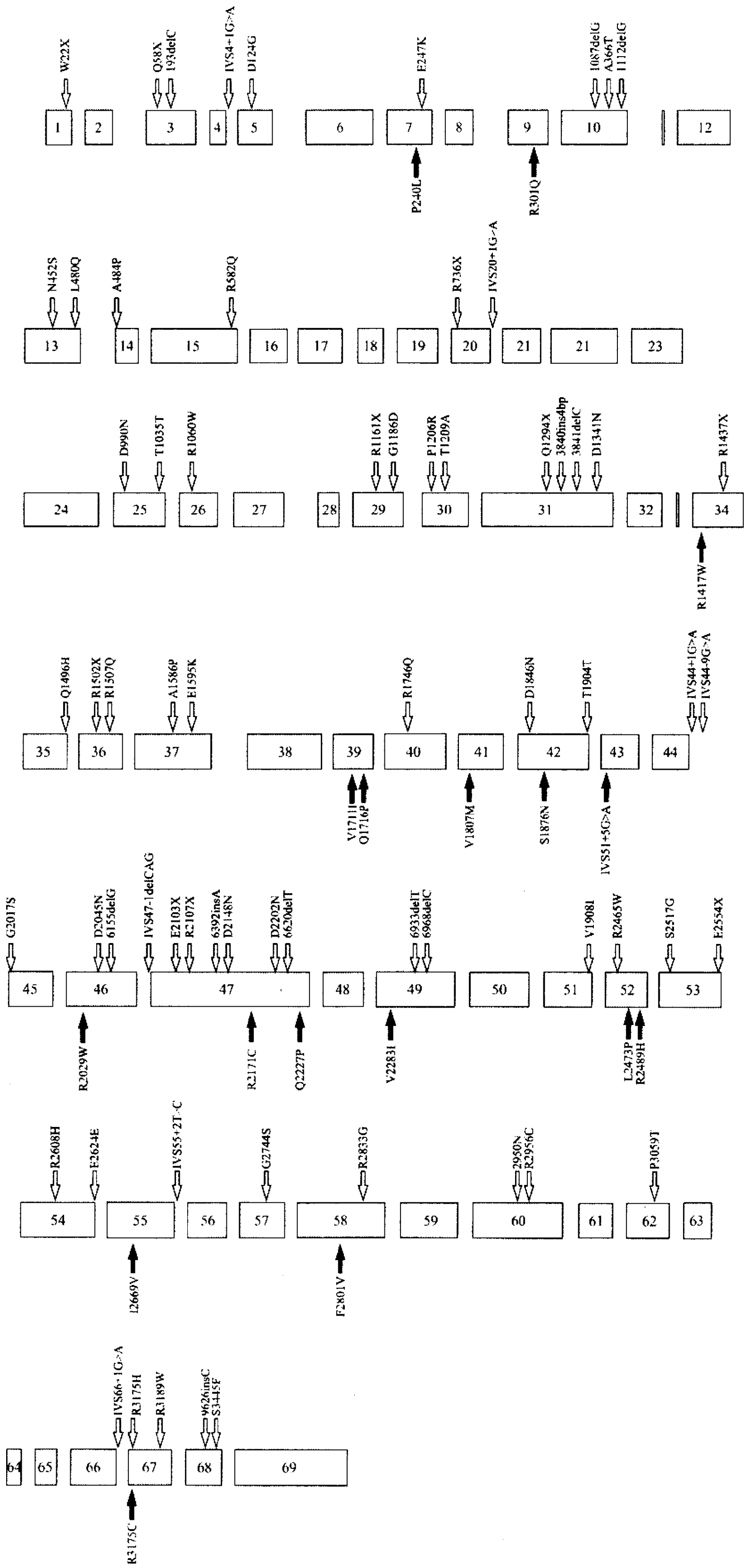

Figure 3. Differences in *CDH23* mutation spectra between Japanese populations and populations with European ancestry. Shaded arrows indicate mutations found in Caucasoid populations; filled arrows indicate mutations found in Japanese.

this strategy may also be applicable to other populations.


## Acknowledgements

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# Pilot Study of Universal Newborn Hearing Screening in Japan: District-Based Screening Program in Okayama

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**Objectives:** Newborn hearing screening was started in Okayama Prefecture in 2001 as part of a nationwide pilot study in Japan. Nearly 50,000 infants have been screened to date, and an observational study and more than 2 years of follow-up of this population are described in this report.

**Methods:** Between June 2001 and March 2005 (45 months), 47,346 neonates were screened with automated auditory brain stem response systems and followed up for at least 2 years. This total corresponds to 95% of the infants born in the 44 gynecologic institutions in this district.

**Results:** After undergoing the screening process twice, 248 infants (0.52%) received referrals; 108 of them had apparent bilaterally affected hearing, and 140 had apparent unilaterally affected hearing. Among the bilateral cases, hearing impairment was diagnosed in 40 infants, for a total prevalence of hearing impairment of 0.08%. In 3 additional infants who received a bilateral pass result and 1 infant who received a unilateral pass result, hearing impairment that was progressive or of late onset was subsequently diagnosed. The positive and negative predictive values were calculated as 40% and 99.993%, respectively.

**Conclusions:** The screening program was carefully designed to work in the Japanese society and to be well managed in Okayama Prefecture.

**Key Words:** automated auditory brain stem response, Japan, negative predictive value, newborn hearing screening, positive predictive value, prelingual deafness.

## INTRODUCTION

Bilateral, permanent hearing impairment (BPHI) is one of the most common neurologic deficits that can become obvious in the neonatal period, affecting 1 in 1,000 infants.[1-3] The presence of BPHI in early infancy can cause poor language development,[1] which can in turn hamper social participation, including school attendance and employment.[2] Appropriate intervention can reportedly prevent or reduce this problem in language development and reduce the risk of the consequences of language delay.[2,3] Given the presence of critical periods of language development, early intervention is supposed to contribute to better intervention for children with BPHI.[4] Earlier commencement of intervention requires earlier identification of BPHI. Children with BPHI can now be identified during infancy in newborn hearing screening (NHS) programs.[3] In Japan, a plan for a pilot study of a national NHS program was announced in 2000, and 3 prefectural governments (Kanagawa, Akita, and Okayama) were the first to be enrolled. Okayama is located in the western part of Japan. The population of this region was 1,959,159 in 1999, making it 21st in population of the 47 prefectures in Japan, and approximately 19,000 infants per year were born in this district.[4]

When the national NHS program was first considered, several peculiarities of Japanese society were of possible concern with regard to the application of an NHS. First, gynecologic clinics are generally more numerous, but much smaller in size, in Japan than in the United States or the United Kingdom. The annual birth numbers in most gynecologic clinics in Japan are less than 300. Organizing a large number of smaller clinics for the NHS program thus represented a challenging task. Second, in Japan, by tradition, a pregnant woman returns to her family home for the delivery and stays with her parents to get sufficient support and to rest physically and psychologically after the birth. This period of *satogaeri*

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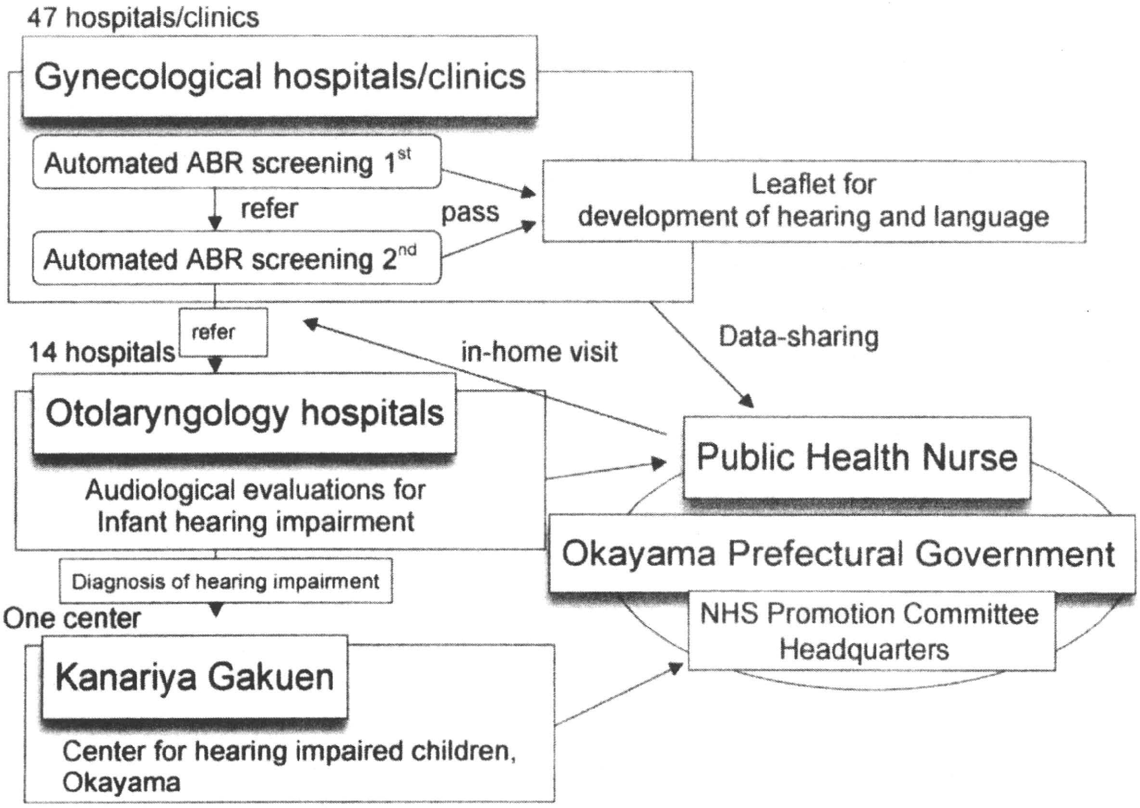


**Fig 1.** Schema of newborn hearing screening (NHS) system in Okayama. ABR — auditory brain stem response.

*bunben* (directly translated as "back-to-home childbirth")[5] usually lasts from around gestational weeks 32 to 35 to a couple of months after birth. Approximately 1,270 of the 19,000 infants born yearly in Okayama Prefecture are born in this fashion. This potentially means that many Japanese mothers and infants move from place to place around the perinatal period, and this practice can create confusion in following infants identified as having hearing impairment in the NHS program. This requires a technically more robust follow-up system for NHS. Given these specific features concerning Japanese society, skepticism has been expressed regarding the applicability of NHS to Japanese society.

To answer such doubts, we reviewed our 4 years of experience managing NHS programs in Okayama Prefecture. We examined our approach and reviewed all statistics obtained through the NHS program in Okayama to establish and remodel better NHS programs for use in Japan.

## NEWBORN HEARING SCREENING SYSTEM

*Procedure.* A pilot study of the National Screening Operation was planned by the Ministry of Health, Welfare, and Labor of the Japanese government before implementation of nationwide NHS programs. The Okayama Prefectural Government then applied to this pilot study, and the outline of the Okayama NHS program was based upon this national study program (Fig 1).

First, gynecologic hospitals and clinics in Okayama Prefecture were invited to participate in this pilot study. As a result, 44 hospitals were enrolled in the NHS program in Okayama. The annual birth numbers from these hospitals are summarized in Fig 2. All clinics and hospitals that participated in this pilot study were supposed to use an automated auditory brain stem response (ABR) system (Natus ALGO; Natus, San Carlos, California). All infants were screened only after written informed consent was obtained from a parent or legal guardian and the study design and informed consent were approved by a review board in Okayama Prefecture. The screening procedure itself was performed according to the recommendations of the system manufacturer. When the first screening results indicated "refer," a repeat screening was performed the next day while the mother and infant were still receiving inpatient care, usually within 5 days after birth. If repeat screening again indicated "refer," the infant was referred to a specialist as described below (Fig 1).

*Audiological Evaluations After Screening.* When the results indicated that an infant had passed the hearing test, the parents were given a leaflet describ-

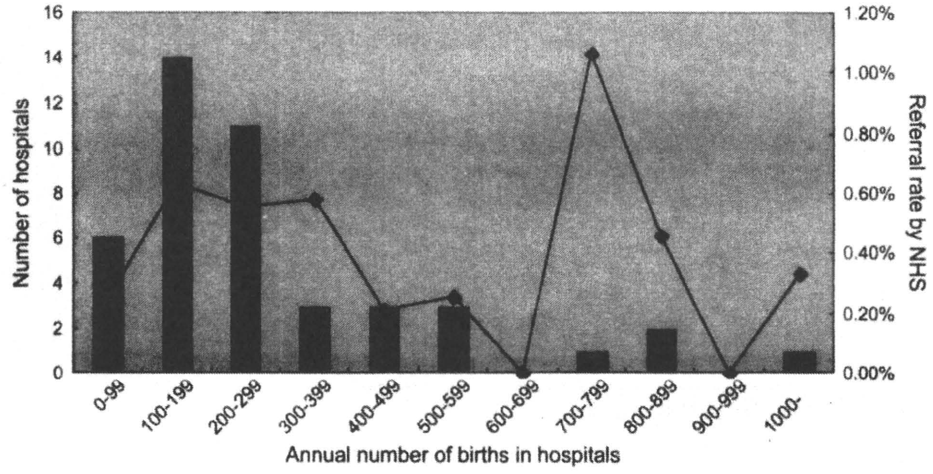


**Fig 2.** Annual number of births in gynecologic clinics and hospitals. Shaded bars demonstrate number of hospitals or clinics with given number of annual births. Lines indicate averaged referral rates for hospitals belonging to given size group. No significant tendency or correlation is apparent between hospital size and accuracy of screening.

ing the normal hearing and language development of infants, and the screening results were reported to the prefectural government under the provisions of the previously obtained informed consent.

Screened infants whose results twice indicated "refer" were then sent to one of 14 otolaryngology hospitals or clinics for further audiological evaluation. These hospitals were selected by the NHS Promotion Committee to cover the entire Okayama Prefecture. After an otolaryngologist removed earwax and checked for middle ear effusion, click-evoked ABR or auditory steady-state response testing was conducted. These children were also subjected to behavioral orientation audiometry and, later, condition orientation reaction and more complicated audiometric procedures such as the peepshow test and play audiometry.

All NHS results were documented in the *Maternal and Child Health Handbook* and used for later tracking and follow-up. This brochure is universally used in Japanese society and distributed to all pregnant women in Japan. These data were also considered in the systematic health tracking of the same children as 1.5- and 3-year-olds.

*Data Transmission and Collection.* The screening results were reported to the Okayama Prefectural Government. The cost of the screening test was paid only after the notification of screening results to the prefectural government. The results were also reported to the referral hospitals for further audiological evaluations as of 2001. Later, the program manual was modified, and the same information was also transferred to a regionally responsible public health nurse (PHN). The PHNs are Okayama government employees with special training in community health, particularly, support for the relationship between mother and infant. The PHNs can help with health concerns and parenting by using the human and social resources of the prefecture. The diagnosis and raw data of behavioral audiometry, ABR and/or ASSR, and otoacoustic emissions from each hospital were also reported to the Okayama Prefectural Government and peer-reviewed annually. Document-based information about the results of screening, finally diagnosed hearing level, commencement of training in Kanariya Gakuen (Audiology Center for Hearing Impaired Children, Okayama), and health and mental conditions of mothers and infants tracked by PHNs were also transferred and aggregated and later integrated by the Okayama Prefectural Government to monitor dropout cases during the follow-up period.

*In-Home Visit Program.* Public health nurses who received information of referral results were informed either by Okayama Prefecture or the gynecologic clinics or hospitals, and in-home visits were conducted immediately after receipt of this information. Public health nurses are obligated to take a 4-hour lecture course once a year regarding the entire NHS program, and during the in-home visits all mothers whose children had received 2 referral results were interviewed. An inquiry sheet was prepared for in-home visits to evaluate maternal concerns and anxiety, attitude toward the NHS program, and tracking for further follow-up. If any problems were encountered after the NHS, this information was supposed to be transferred to the Promotion Committee of NHS programs in Okayama.

*Identification of and Interventions for Hearing-Impaired Children.* All children in whom BPHI was diagnosed or suspected were then referred to Kanariya Gakuen.[6] Because all children in whom a hearing impairment is suspected, regardless of participation in the NHS program, are referred to Kanariya Gakuen, late-onset and/or progressive hearing loss after NHS can be identified by checking all referrals to Kanariya Gakuen from all over Okayama Prefecture. Participation in the NHS program was checked

TABLE 1. NUMBER OF NEWBORN HEARING SCREENINGS IN OKAYAMA FOR YEARS 2001 TO 2004

| | *No.* | *%* |
|---|---|---|
| Newborns | 49,839 | |
| Newborns who underwent newborn hearing screening | 47,346 | 95 |
| Infants born by *satogaeri bunben** | 4,470 | |
| Infants referred | 248 | 0.52 |
| Unilateral hearing loss | 140 | 0.32 |
| Bilateral hearing loss | 108 | 0.19 |

*Estimated from national statistics.

by both interview and documentation in the *Maternal and Child Health Handbook*. If any ambiguity remained, the staff of Kanariya Gakuen made further inquiries.

Infants with hearing impairment but no additional health problems were then introduced to the use of hearing aids. In infants who had some additional physical or mental handicaps or very mild hearing loss (around 30 dB normal hearing level), commencement of intervention was delayed in some cases. All of these infants received intervention with total communication, auditory-oral, or auditory-aural methods at Kanariya Gakuen.

Cases of unilateral hearing impairment were also followed up at otolaryngology hospitals and clinics to ensure no delays in language development. Patients who had progression of hearing loss or any degree of delay in language development were to be referred to Kanariya Gakuen again. In this article, only findings of BPHI are reported.

## RESULTS

*Screening*. A total of 44 gynecologic hospitals voluntarily applied to participate in the NHS program. In these hospitals, 49,839 infants were born, and infants whose parents did not provide written informed consent did not receive screening and were excluded from the study. As a result, 47,346 neonates were screened between June 2001 and March 2005 (45 months), corresponding to 95% of infants born in these hospitals. During the same time period, 71,198 infants were born as Okayama citizens, according to the demographic statistics. Infants in the NHS program received screening by automated ABR testing on day 3 after birth and again on day 4 or 5 after birth if the first results suggested a referral, and 248 infants (0.52%) received referrals after 2 screenings (Table 1).

*Frequency of Hearing Impairment and Positive Predictive Value*. The 248 infants included 108 with suspected bilateral hearing loss and 140 with suspected unilateral hearing loss. Of the 108 referrals for suspected bilateral loss, hearing impairment was diagnosed in 40 infants and an early intervention program was recommended. This represents 0.08% of the total number of screened infants, so the positive predictive value for bilateral hearing loss was 40% of bilateral referral cases.

*Identification of Later-Diagnosed Hearing Loss*. All infants screened by this NHS program had their screening results noted in the *Maternal and Child Health Handbook* printed by the Okayama Prefectural Government. If hearing loss was diagnosed later in the child's life, referral was still made to Kanariya Gakuen and further examination was conducted. Under this system, 3 cases of bilateral hearing loss were diagnosed 2 to 6 years after participation in the NHS program. The clinical courses of these cases are summarized in Table 2, and all 3 patients were considered to display progressive, late-onset hearing loss. In addition, 1 neonate who received a referral for suspected unilateral hearing loss finally received a diagnosis of BPHI (Table 2). This patient apparently displayed a risk factor for progressive hearing loss.

*Distribution of Gynecologic Institutes*. The annual birth numbers for each gynecologic institute are

TABLE 2. CLINICAL COURSE OF CHILDREN WITH PROGRESSIVE HEARING IMPAIRMENT

| *Screening Results* | *Age at Identification* | *Course of Identification of Deafness After Screening* |
|---|---|---|
| Unilateral pass | 6 mo | Intrapartum infection and administration of aminoglycoside after birth; first screening result was unilateral pass, but screening results before discharge changed to bilateral referral; moderate hearing loss was diagnosed, and hearing aid is now being used |
| Bilateral pass | 1 y | After meningitis, loss of response to environmental sounds was noted at 1 year of age; severe hearing impairment was diagnosed, and cochlear implant is now being used |
| Bilateral pass | 4 y | Patient started to speak after his first birthday and could call his younger brother's name; because of delayed language development, he was referred to Kanariya Gakuen at 4 years of age, and severe deafness was diagnosed; cochlear implant is now being used |
| Bilateral pass | 2 y | Response to sounds was noted by mother during infancy; because of delayed language development, she was referred to Kanariya Gakuen at 2 years of age, and severe deafness was diagnosed; cochlear implant is now being used |

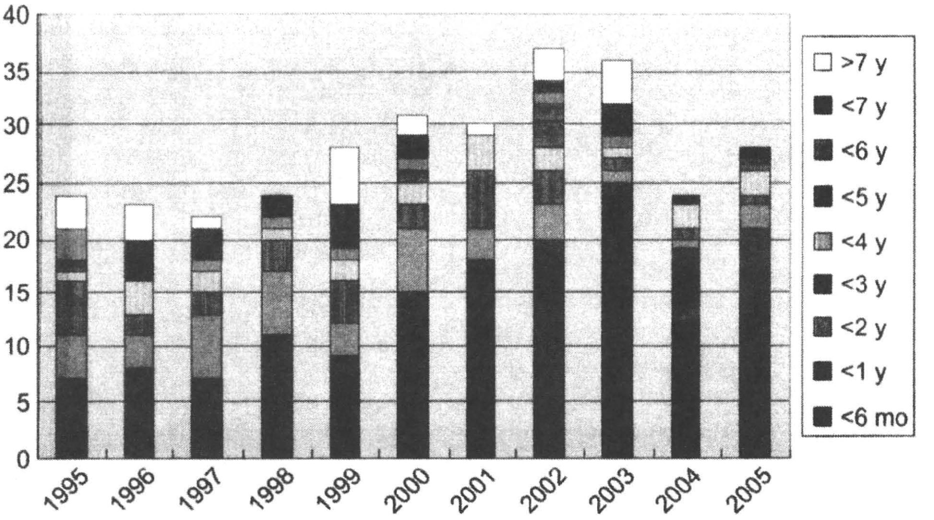


**Fig 3.** Age at referral to Kanariya Gakuen (Center for Hearing Impaired Children, Okayama). Before implementation of NHS, limited numbers of cases were referred to Kanariya Gakuen during infancy. Apparent increase in newborns (less than 6 months of age) was observed after 1997, as hearing screenings in this district were sporadic before then. Thus, nearly one third of hearing-impaired children were identified before this pilot study. Proportion of infants and total number of hearing-impaired children increased after commencement of this pilot study; however, apparent decreases have been observed in past 2 years, and total number of children referred has decreased to levels comparable to those before pilot study.

summarized in Fig 2. The annual number of births for the majority of hospitals that committed to this pilot study was less than 300 births per year, and one quarter of the hospitals had 200 to 299 births per year. The frequencies of referral results after automated ABR screening in each hospital were not associated with hospital size, and relatively low frequencies of referral (less than 0.6%) were achieved, with the exception of a few hospitals with neonatal intensive care units.

*Age at Referral.* The age distribution of hearing-impaired children on referral to Kanariya Gakuen is shown in Fig 3, and longitudinal alteration was demonstrated. Children with BPHI referred before 6 months of age represented a very small proportion in 1996, but surged to nearly 30% in 1998, when a few hospitals in Okayama started hospital-based NHS.[10] After Okayama Prefecture started the NHS program in 2001, this proportion increased gradually. The total number of children found to have BPHI also increased annually for 4 years, but then decreased in 2004. Children with BPHI referred before 6 months of age represented more than 70% of referrals as of 2005 (Fig 3).

## DISCUSSION

Newborn hearing screening programs have been effectively managed in Okayama Prefecture for 45 months. During this time, more than 40,000 infants have been screened with a very low referral rate compared with rates reported from other sources.[7-9] Most infants who received referrals were also effectively followed by PHNs in Okayama Prefecture. A careful program design that considered unique aspects of Japanese society was helpful for establishing this NHS project, and the reorganization of currently available social resources has proven highly useful for achieving NHS project goals. A very low referral rate was achieved without decreasing the negative predictive value of NHS in the present study. This was partly caused by longer in-hospital care after delivery compared to the United States and other countries, as some reports from other Japanese hospitals have likewise shown similar referral rates, and these hospitals have used similar in-patient care periods.[10]

We encountered 3 cases of bilateral hearing impairment in which "pass" results were obtained from the first screening process. One of these patients had an apparent history of postpartum hearing loss (meningitis), and the others were observed to have possible responses to sound in early infancy from the detailed history taken by audiologists. In these cases, progressive hearing loss was eventually suspected despite no apparent cause of hearing loss. Neither patient displayed enlarged vestibular aqueducts on high-resolution computed tomography (data not shown), and genetic testing including SLC26A4 or cytomegalovirus infection was unavailable, as the parents declined to undergo genetic testing. We do not know how many infants judged as passing the first screening process moved away from Okayama Prefecture in this study period. This means that we have no way of knowing the denominator to determine the true negative predictive value in this study. At least from the framework for the regional health-care network, no apparent "false-negative" cases were observed during the 2- to 6-year follow-up, and 99.993% was the predictive value for negative results from the NHS program.

Covering all of the infants born to Okayama residents represents an ongoing challenge for us. We still take nearly 20% of Okayama citizens into consideration for complete universal NHS. Because of *satogaeri bunben*, each year 1,270 infants born outside Okayama Prefecture miss their right to receive NHS, despite being born to Okayama residents. This represents approximately 7.2% of total annual births, in which the infants miss a chance to be screened. From the perspective of regional health-care system planners responsible for health care in Okayama Prefecture, this represents a group at potential risk of hearing loss who cannot be identified unless particular symptoms such as language delay appear. According to our research, the number of *satogaeri bunben* births is relatively low compared to the total birth number and coverage by the NHS service, but this still represents the largest population of infants not covered by the NHS service in Okayama Prefecture. In addition, approximately 1,500 infants per year were born in hospitals that were not participating in the NHS program, and 300 infants per year were born at a midwifery house. To capture these total 3,000 infants each year, we are now planning to implement outpatient screening protocols. These protocols were partially initiated from 2004 and will expand to cover all newborns born to Okayama residents regardless of whether the hospital of birth provides the NHS service, applying first to midwifery house deliveries and then to *satogaeri bunben* births. Expansion of the NHS programs to other prefectures all over Japan may solve this problem more simply. If all Japanese can receive NHS regardless of birthplace, *satogaeri bunben* will no longer present a problem for NHS in Japan.

Our preliminary data also showed fair language development in children after they received hearing intervention at Kanariya Gakuen. Our NHS programs require some modifications, but appear to have demonstrated preliminary success. At least, early hearing detection and intervention has been achieved with satisfactory accuracy of the screening tests and minimal psychological effects on the mothers.

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# Developmental dysgraphia with profound hearing impairment: Intervention by auditory methods enabled by cochlear implant

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**Abstract**

Learning disability combined with hearing impairment (LDHI) is a poor prognostic factor for the language development of hearing impaired children after educational intervention. A typical example of a child with LDHI and effective interventions provided by cochlear implants are presented in this report. A case of congenital cytomegaloviral infection that showed dysgraphia as well as profound deafness was reported and an underlying visual processing problem diagnosed in the present case caused the patient's dysgraphia. The dysgraphia could be circumvented by the use of auditory memory fairly established by a cochlear implant.


*Keywords:* Learning difficulty; Deafness; Congenital cytomegalovirus infection; Developmental dyslexia; Kana dysgraphia; Prelingual hearing impairment; Cochlear implant

## 1. Introduction

Hearing impairment is one of the most common causes of language developmental delay, affecting one of 1000 newborns annually. Usually, the language delay can be diminished by educational intervention [1]. The combination of learning disability (LD) and hearing impairment (HI), however, frequently makes this educational intervention highly difficult [2–6]. The combination of LD can cause additional delay in their language development. In fact, 7–11% of students in programs for the deaf or hard-of-hearing are combined with LD and this makes LD the single most frequent disorder concomitant with deafness [7,8]. Thus, the diagnosis of and intervention for the combined neurological deficits that can potentially cause LD are quite important for the educational intervention of hearing impaired children.

To the interventional procedure, we now can apply cochlear implant for profoundly deaf children to supply better phonological information of the language [9]. This implies that phonological information that can be important for LD education can be supplied though a cochlear implant. However, language output was compared between deaf child with and without LD [2,10,11] and no attention has been paid to their diagnostic, therapeutic, or interventional approach for children with LDHI. In this report, we present a set of diagnostic procedures for children with LDHI, demonstrating a case of a child with profound hearing impairment caused by congenital cytomegalovirus (CMV) infection and her learning difficulties in Kanji (Japanese ideogram) and Kata-kana (Japanese phonogram) dysgraphia that hampered her language ability. As an interventional procedure, we applied phonological education for Kanji-writing [12]. This procedure was applicable because the patient demonstrated better auditory memory with a cochlear implant in spite of her profound hearing impairment.

## 2. Case presentation

### 2.1. Present history

The patient was referred to our hospital when she was 11 years old. She was in the fifth grade of the mainstream

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school and additional training was provided in hard-of-hearing classes for mathematics and Japanese.

When the child was born, she received a diagnosis of congenital cytomegalovirus infection. Multiple cerebral infarctions in the periventricular region were noted on magnetic resonance images obtained when the child was 11 months old. Profound hearing impairment was first diagnosed by auditory brainstem response (ABR) when the child was 5 months old. She was finally diagnosed to have profoundly hearing impairment. Educational intervention with a hearing aid was started when she was 11 months old. Her communication mode when she started school was total communication. She received a cochlear implant at our hospital when she was 7 years 6 months old. During the patient's follow-up period, her teacher in the hard-of-hearing school referred her to our hospital because she demonstrated severe problems in writing Kata-kana (Japanese phonographic characters) and Kanji (Japanese ideographic characters). She was also found to have difficulties in applying a triangle ruler to square figures in mathematics class.

### 2.2. Imaging and laboratory tests

Magnetic resonance imaging was obtained prior to cochlear implant surgery and re-evaluation of these images was performed at the time of consultation (Fig. 1a). Enlargement of the lateral cerebral ventricles was observed especially in the posterior angle near the temporal, occipital, and parietal lobes. No abnormality was reported from electroencephalogram results.

### 2.3. Neuropsychological examination

The child showed hemiplegia in her right hand and she began using her left hand for writing. A part of the data has already been reported [11].

### 2.4. Visual-spatial cognitive tests

The Raven's colored progressive matrices (RCPM) test was conducted and mild (16/36, -2SD) mental retardation was suspected. The picture vocabulary test (PVT) revealed that 48/68 and results corresponding to a 9-year-old's level. The child drew six isolated faces in one plane on the cubic perspective drawing test (Fig. 2a). By Rey's complicated figure reproduction test, her scores were 6.5/36 for copying (Fig. 2b), 4/36 for immediate reproduction, and 2/36 for delayed reproduction, indicating impairment of her visual cognitive processing.

### 2.5. Auditory phonetic cognitive tests

Hearing levels with and without a cochlear implant are indicated in Fig. 3. A monosyllable speech perception test was conducted and the child recognized 50% of the combinations of a vowel and consonant and 98% of vowels of monosyllable phonems in open-set conditions without visual cues. The Rey's Auditory-Verbal Learning Test (AVLT) was conducted and her scores were 11/15 for maximum reproduction and 7/15 for delayed reproduction.

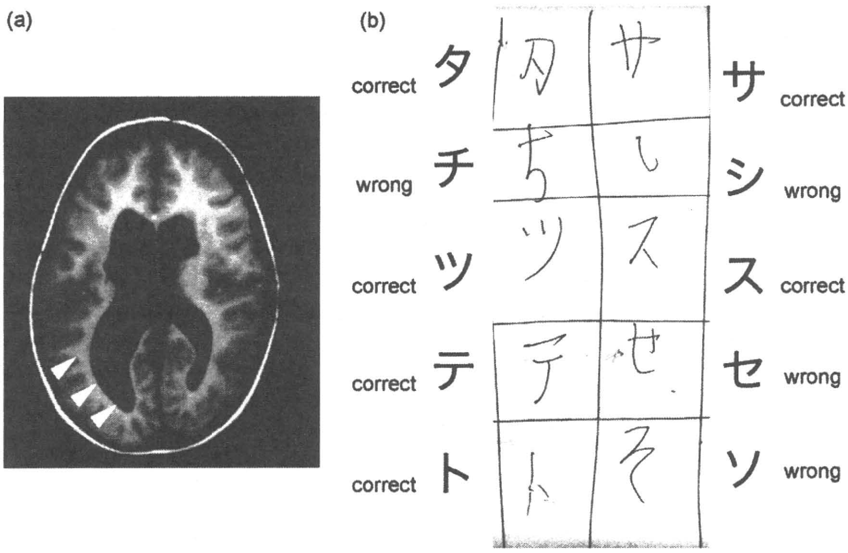


Fig. 1. (a) MRI and (b) her handwriting of Kata-kana before training. (a) Enlargement of cornu posterius (arrow head) in left hemisphere was observed. (b) Typical handwritings of /sa/, /si/, /su/, /se/, /so/, and /ta/, /ti//tu/, /te/, /to/. Correct form of Kata-kana was also indicated side of the figure. She sometimes wrote Hira-gana letters, instead of Kata-kana letters, when she could not recall proper Kata-kana letters.

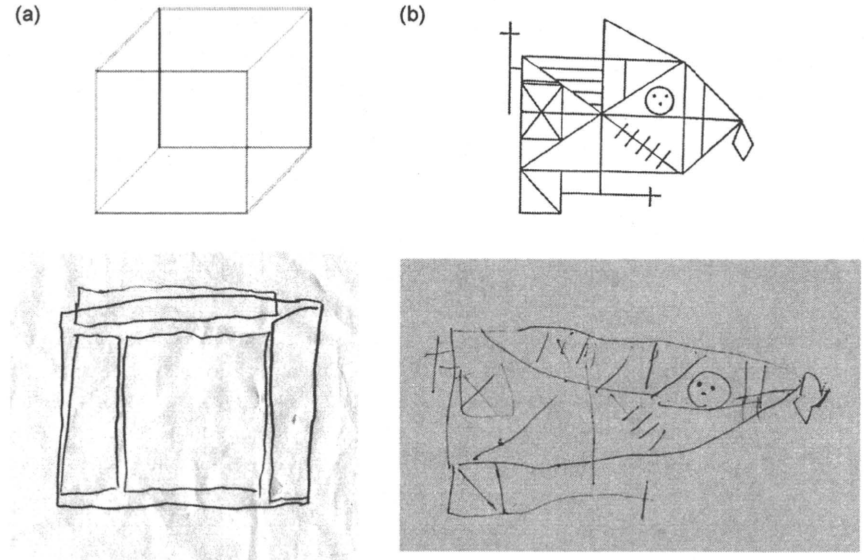


Fig. 2. Cube copy and Ray-Ostrich Complex Figure Test (RCFT). Upper: exemplar of each test, lower: copied result by the present case.

### 2.6. Language evaluation

The result of the Standard Language Test for Aphasia (SLTA) is summarized in Fig. 4. Standard Language Test for Aphasia (SLTA) [13] was used to evaluate language functions. The SLTA is the standardized battery test most commonly used in Japan, especially for aphasia patients. The correct responses in the SLTA testing are shown from 0% = most severe to 100% = normal. The different aspects of language can be evaluated separately by SLTA: auditory comprehension (to obey verbal commands), speaking (object naming, word repetition, sentence repetition), reading comprehension (follow written commands, sentence picture matching, etc.), writing (dictation of kana letters and dictation of short sentence), and calculation. Tests containing the writing process demonstrated markedly poor scores, while other auditory comprehension tests and reading comprehension tests revealed retained ability, which were comparable to the results of normal-hearing adult data.

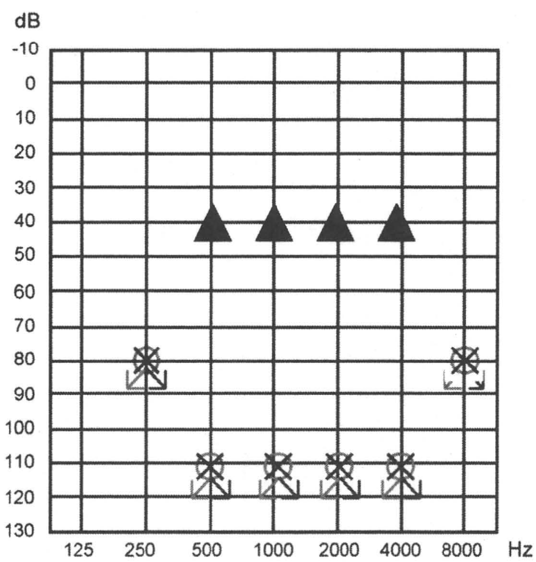


Fig. 3. Audiogram: The results of audiometry by head-set (blue and red) and sound field with cochlear implant (black triangle) was indicated.

### 2.7. Writing Kanji and Kata-kana

Screening tests for Japanese-character agraphia (14) were conducted. Hira-gana writing was within normal limits (101/

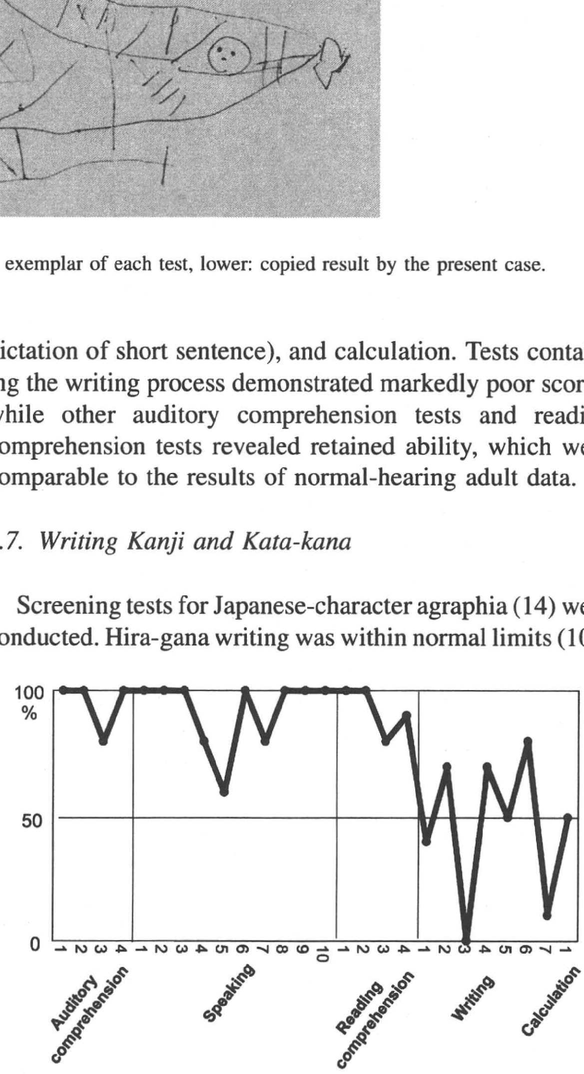


Fig. 4. The results of Standarized Language test for Aphasia (SLTA). Auditory comprehension tests include: (1) auditory word recognition, (2) sentence comprehension, (3) follow verbal commands, (4) Kana letter discrimination. Speaking tests include: (1) objective naming, (2) word repetition, (3) describe behaviors, (4) explain picture story, (5) sentence repetition, (6) listing words (words fluency), (7) read aloud Kanji words, (8) read aloud kana letters, (9) read aloud kana words, (10) read aloud short sentence. Reading comprehension tests include: (1) Kanji word-picture matching, (2) Kana word-picture matching, (3) sentence picture matching, (4) follow written commands. Writing tests include: (1) write Kanji words, (2) write kana words, (3) narrative writing, (4) dictate kana letters, (5) dictate Kanji words, (6) dictate kana words, (7) dictate short sentences calculation. Writing tasks were specifically impaired in this patient.

103) and marked deterioration (-2SD) of Kata-kana writing was observed (65/103). The child's greatest tendency was to write Hira-gana corresponding to the same mora of the questioned Kata-kana; otherwise, she responded to nothing. An Example was shown in Fig. 1b. The screening test for Kanji agraphia [14] also revealed significant (8/20 more than -1SD) delay comparing with the child's peers.

### 2.8. Intervention

Educational intervention was conducted once a week for 40 min in each session from October 2002 to March 2003. The first goal for the training was established for Kata-kana writing. The auditory training procedure reported by Uno and co-workers [12,15,16] was conducted.

Serial recitation of 46 Japanese Kata-kanas that correspond to the all resonand sound was tried, and the time to completion was recorded. During this step, serial recitation was helped by table-directed recall. Japanese resonand sound syllables consist of five vowels (a i u e o) and combinations of one of these vowels with nine consonants (k, s, t, n, h, m, y, r, w). In Japanese elementary schools, a 5 × 10 table is used to study Hiragana and Kata-kana.

Next, a serial Kata-kana writing test was conducted. When the child could not write one Kata-kana repeatedly, the Kata-kana word was decomposed into several "parts" of the lines and memorized by phonological awareness, according to the procedure previously reported by Uno et al. [16] For an example, the character corresponding to the monosyllable sound "NU" was dissolved and replaced by a phrase "line and cross" (Fig. 5) and, similarly, "TU" was replaced by "two dots and a slash". These phrases were used to help memorizing and recalling the figure of each of Kata-kana character that the child could not learn properly. This intervention was repeated until she could complete the serial Kata-kana writing within 30 s.

## 3. Results of the intervention

The first task of recitation within 30 s was achieved in the second session. Thirteen morae whose phonation could be recalled but whose phonogram could not be written properly were identified. These morae included "NE", "NU", and "O". During the fourth to seventh sessions, Uno's methods were applied and after the tenth session, the child could write all serially arranged Kata-kana characters within 1.5 min. Randomly presented morae were tested in the 12th session and the child could write all Kata-kana characters. Intervention for learning Kanji characters is now in progress, also applying the Uno method.

## 4. Discussion

Pure Kata-kana dysgraphia can be caused either by visual cognitive impairment or visual memory impairment [16,17]. However, other problems in several neurological conditions including attention, intelligence, or sequential processing can cause dysgraphia [17]. Judging from the child's mandatory communication skills and semantic understandings, neither her intelligence nor her profound hearing impairment could be the major cause of this dysgraphia, at least of the Kata-kana dysgraphia. Instead, problems in her visual cognitive processing are more likely to be the major cause of this dysgraphia, as indicated in all visual cognitive tests including the Rey's complicated figure drawing tests

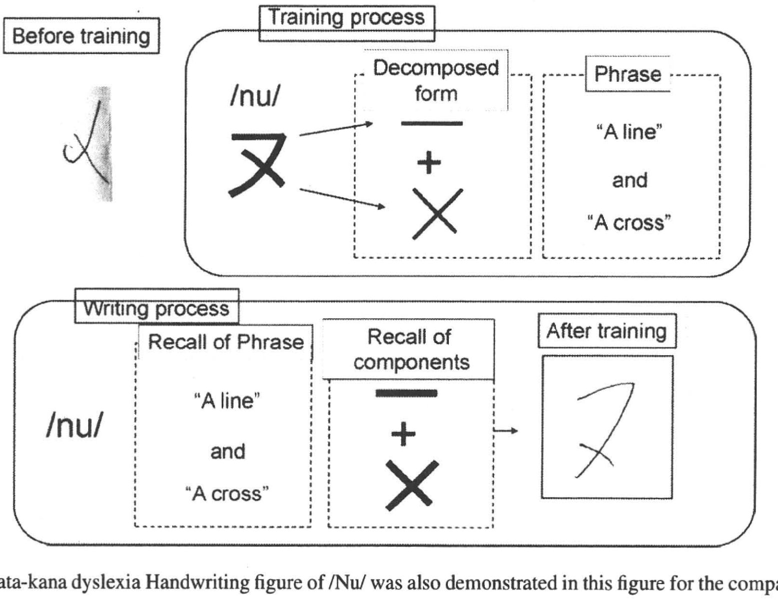


Fig. 5. Diagram of training for Kata-kana dyslexia Handwriting figure of /Nu/ was also demonstrated in this figure for the comparison between before and after intervention.

and cubic drawing test. Magnetic resonance imaging findings also support this possibility, demonstrating the same intracranial lesion as observed in cases of cerebrovascular accident with visual cognitive disorder or dyslexia/dysgraphia. Relatively low scores by RCPM might also be explained by this visual cognitive problem because RCPM itself requires rapid processing ability of visually presented materials.

Discussion of the effect of learning disabilities on individuals with cochlear implants has been focused on the negative aspects. Isaacson et al. examined 10 pediatric patients with cochlear implants postmeningitis to determine the effects of learning disability on measures of auditory perception, receptive language development, and sequential organization, and they observed slower progress, more inconsistencies, and lower test scores of the children with LD and deafness compared with the test scores of deaf children without LD [2]. It is true that in the present case, the child received a cochlear implant at an older age and had additional LD caused by a visual cognitive problem, both of which indicate poorer prognosis for language acquisition. For the educational intervention of pure dysgraphia caused by visual processing weakness, application of auditory cognitive ability for pure developmental dyslexia/dysgraphia has already been validated [15,16]. The profound hearing impairment of the child in the present case implied the possibility of poor auditory processing ability that might make this interventional approach impossible. The result of Rey's AVLT, however, indicated that the auditory processing ability and auditory memory were better than the visual cognitive processing and visual memory. In actuality, it was difficult for this child to learn how to write without auditory processed memory and this hearing ability was crucial for her education. Auditory cognitive ability and auditory-verbal learning process, which are maintained by a cochlear implant, provide an apparent benefit and the application of a cochlear implant can affirmatively be considered for cases with visual processing weakness, although this is a single case report and this result cannot always be generalized. However, the presence of LDHI cannot be the factor that persuades families to abandon the cochlear implant.

## 5. Conclusion

In this report, we demonstrated a set of approaches that contained hearing compensation by cochlear implant, neuropsychological assessment, and educational intervention with Uno methods. Neuropsychological assessment revealed an apparently "better" cognitive approach for learning process between choices of hearing and visual approaches.

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