

# A child with severe inner ear malformations with favorable hearing utilization and balance functions after wearing hearing aids

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## Abstract

Infants with congenital deafness caused by severe bilateral inner ear malformations frequently suffer from severe hearing loss and poor balance. Unfortunately, the use of hearing aids is usually ineffective in recovering hearing, necessitating cochlear implants. We report a case of a 6-year-old boy with congenital deafness and bilateral inner ear malformations (right side, incomplete partition type I [IP-I]; left side, common cavity deformity). Hearing aids had a remarkable effect in this patient, enabling sufficient and favorable hearing recovery such as to allow the patient to engage in daily conversations. Per-rotatory nystagmus was recorded on an electronystagmogram for both right and left rotations in a damped rotational chair test. It is rare for deaf children with severe bilateral inner ear malformation to demonstrate favorable development in hearing and good equilibrium function. Our findings suggest that auditory—vestibular hair cells in this patient may have been partially preserved despite IP-I in the right ear and common cavity deformity of the left ear.

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**Keywords:** Congenital hearing loss; Inner ear malformations; Rotation chair test

## 1. Introduction

Malformations of the inner ear are found in approximately 20% of children with congenital sensorineural hearing loss and are among the primary causes of congenital deafness (Jackler et al., 1987). Congenital malformations can occur due to an arrest in membranous and osseous labyrinth development during the early in the gestational period. Sennaroglu and Saatci classified severe inner ear malformations as follows: Michel deformity, cochlear aplasia, common cavity, cochlear

hypoplasia, incomplete partition type I, and incomplete partition type II (Mondini malformation) (Sennaroglu and Saatci, 2002).

Children with severe malformations of the inner ear suffer from a number of problems. First, development of language is poor even when using hearing aids; cochlear implants are therefore often indicated. Second, because of the poor hearing ability in these children, it is difficult to predict the outcome of cochlear implant surgery. Third, children with congenital deafness also tend to have balance disorders, resulting in delayed motor development in head control and independent walking (Kaga, 1999).

Here, we report the case of a child who, despite having severe bilateral inner ear malformations, achieved good hearing recovery and equilibrium function after wearing hearing aids.

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## 2. Patient and methods

### 2.1. Case history

The patient was a 6-year-old boy delivered by normal vaginal delivery after a full-term, uneventful pregnancy. The patient was brought to our hospital three months after birth for a mandatory re-examination of both ears following the newborn hearing screening. Audiological assessments included behavioral audiometry and auditory brainstem response. The auditory brainstem response showed severe hearing loss in both ears (Fig. 1). Temporal bone computed tomography demonstrated abnormal bilateral cochleae (Fig. 2). We classified the patient's cochlear malformations according to classifications by Sennaroglu and Saatci (2002). The anomaly in the right ear was identified as incomplete partition type I (IP-I); that is, the cochlea lacked the entire modiolus and cribriform area, resulting in a cystic appearance accompanied by a large cystic vestibule. The left ear was classified as a common cavity deformity with a narrow internal auditory canal, which indicated that the presence of a cystic cavity, representing the cochlea and vestibule, prevented differentiation into the cochlea and vestibule. The middle ear on both sides had no deformities. The vestibulocochlear nerve of the right ear was identifiable by magnetic resonance imaging, while that of the left ear was partly obscured (Fig. 3).

### 2.2. Auditory–verbal therapy

The patient showed severe bilateral sensorineural hearing loss, with hearing levels ranging from 100 dB HL and higher by behavioral audiometry. He was diagnosed with severe hearing loss caused by inner ear malformations without other disorders, such as mental retardation or brain malformation. The patient started wearing hearing aids provided by our

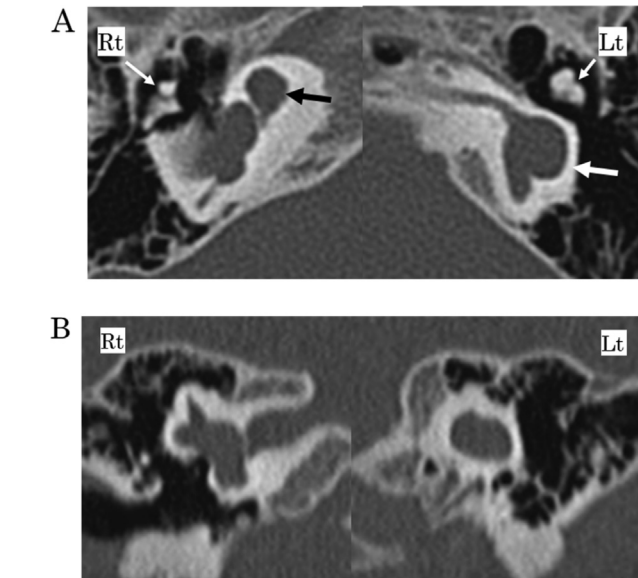


Fig. 2. (A) Axial slice of computed tomography image. (B) Coronal slice of computed tomography image demonstrating IP-I (black arrow) and common cavity deformity (white arrow). The ossicles showed no malformations (thick white arrow).

hearing clinic in both ears at five months of age and participated in auditory–verbal therapy in a kindergarten facility. By eight months of age, bilateral detailed hearing thresholds could be determined by audiometry: hearing thresholds were 95 dB HL in the right ear and 104 dB HL in the left ear (Fig. 4).

### 2.3. Development of gross motor function

Head control and independent walking were used as measures of motor development. Based on the results of the 2010 National Infant and Children Physical Development survey

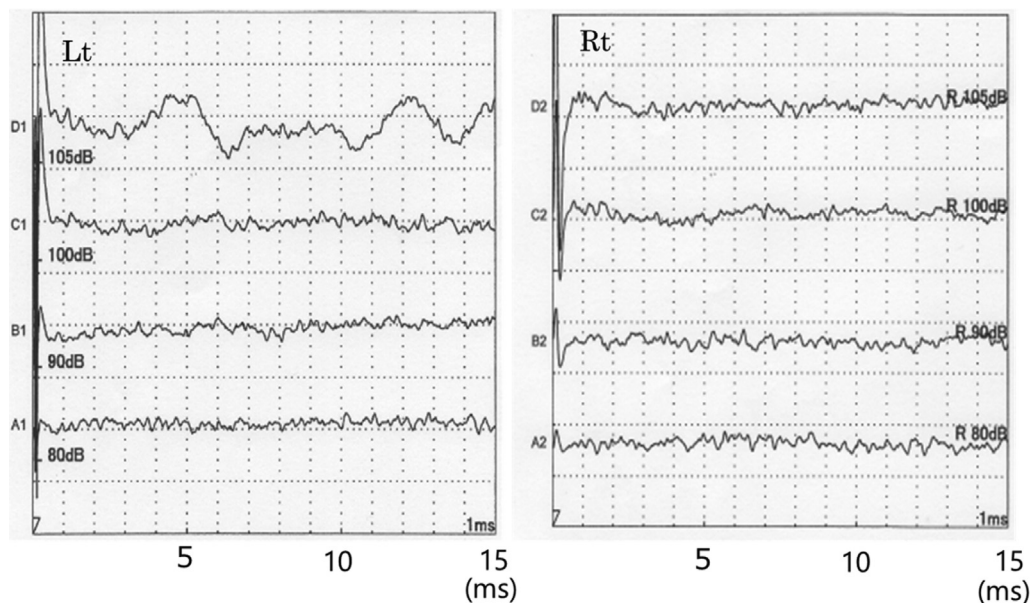


Fig. 1. Auditory brain stem response at the age of 3 months. Auditory brain stem response testing revealed no waveforms in this patient.

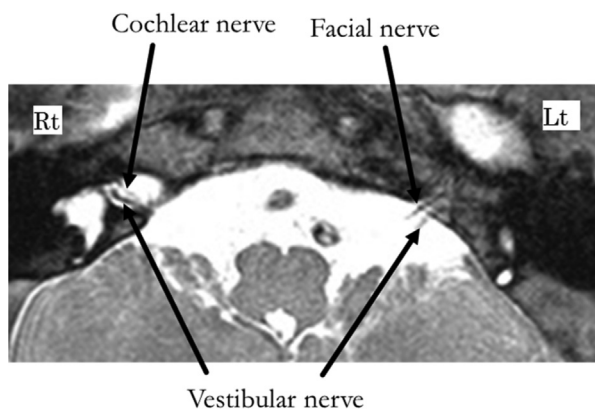


Fig. 3. Magnetic resonance imaging. The vestibulocochlear nerve was present in the right ear but indistinct in the left ear.

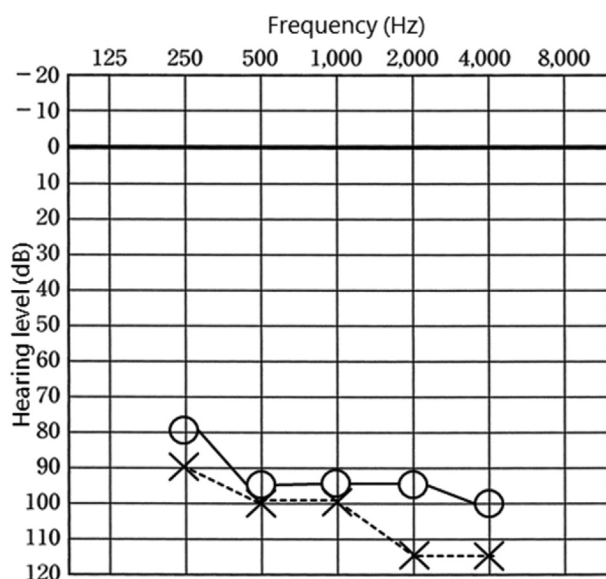


Fig. 4. Visual reinforcement audiometry using an inserted earphone at the age of 8 months. Threshold levels were 95 dB HL in the right ear and 104 dB HL in the left ear.

conducted by the Ministry of Health, Labour and Welfare of Japan, motor development is considered delayed when head control and independent walking occur later than 5 and 14 months, respectively (Masuda and Kaga, 2014). The patient first exhibited head control at 6 months and independent walking at 20 months of age. Therefore, he was considered to have delayed head control and independent walking.

A rotational chair (Nagashima Co, Ltd. S-II) was used to evaluate the patient's vestibular ocular function, by accelerating the rotation to a maximum rotational velocity of  $160^\circ/\text{s}$ , then decelerating by  $4^\circ/\text{s}^2$ . The test was performed in clockwise and counterclockwise directions. Eye movements were recorded by an electronystagmogram and duration and number of beats of per-rotatory nystagmus were calculated to evaluate the semicircular canals and otolith organs in both ears (Kaga et al., 1981) (Fig. 5). The patient showed normal duration and number of beats of per-rotatory nystagmus in the

clockwise rotation but a reduced number of beats in the counterclockwise rotation (Table 1).

#### 2.4. Assessment of hearing development

The patient spoke his first word 17 months after birth and was using two-word sentences 27 months after birth. The Japanese version of the Early Auditory Skill Development for Special Populations (EASD) questionnaire was used to assess hearing development with hearing aids (Johnson et al., 1997; Tomizawa et al., 2013). The table of developmental stages in EASD (39 items) indicates the developmental milestones of daily auditory/verbal behaviors of children between birth and 3 years of age, with a focus on five stages of qualitative changes: stages one to three correspond to the preverbal period while stages four and five correspond to the verbal period. Speech-language-hearing therapists used the EASD questionnaire to interview our patient's caregivers every 3–4 months.

- Stage one: Noticing and beginning to pay attention to sounds.
- Stage two: Making an effort to locate the source of sounds and starting to understand the meaning of sounds.
- Stage three: Following the source of sounds and making intentional vocalizations.
- Stage four: Showing improved understanding of sounds/words and using voice to communicate.
- Stage five: Starting to show auditory language comprehension and communication by conversation.

The chronological change in the patient's EASD scores showed that he had reached stage three by less than 12 months of age, approximately seven months after hearing aids were first applied. His auditory and verbal skills had developed markedly; his development was within the normal range for a 12+ month-old child, that is, at stages four to five. His developmental stage continued to correlate with age up to two years of age, when he had reached stage five and could communicate by conversations using sentences (Fig. 6). Cochlear implantation was not chosen for this patient because of the favorable development in his hearing skills.

Auditory tests showed that the patient's hearing threshold with hearing aids had reduced to 41.25 dB at the age of six (Fig. 7). He attends a mainstream primary school and a school that provides support services for the deaf. He uses an FM system in addition to his hearing aids. He is an auditory–oral communicator and does not use sign language or cued speech.

### 3. Discussion

Our patient showed favorable auditory development and balance with the use of hearing aids despite severe malformations of the inner ear. We initially considered cochlea implantation for this patient but his parents objected. Our patient's hearing threshold with hearing aids had reduced to 41.25 dB at the age of six. This favorable progress with hearing aids indicated that cochlea implants were not necessary.

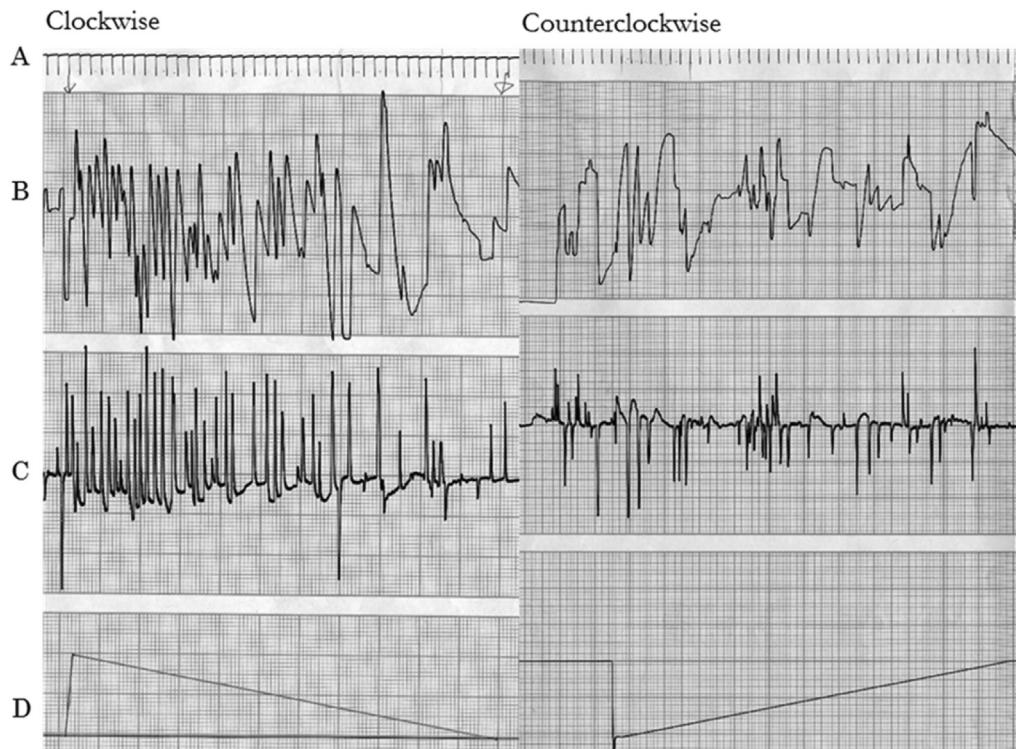


Fig. 5. Rotational chair test. (A) Time scale (one division per second). (B) Angular displacement of the eyes (time constant, 3.0 s; calibration signal,  $10^\circ$ ). (C) Rotational velocity of the eyes (time constant, 0.03 s; calibration signal,  $20^\circ/\text{s}$ ). (D) Angular velocity of the rotating chair.

Table 1

The results indicate normal responses during rotation in the clockwise direction but poor responses in the counterclockwise direction.

	Average of 6 years	Clockwise from the patient	Counterclockwise from the patient
Number of beats	30.7	33	18
Time duration (s)	19.4	37	22

Children with a hearing threshold greater than 100 dB often cannot attain a hearing threshold of 50 dB even with hearing aids; speech sounds are difficult to hear and lip reading is important for auditory understanding. For such children, cochlear implantation is often indicated. Eisenman et al. demonstrated improvements in speech perception over time for 17 children with cochlear malformations, albeit with slower progress than their age-matched peers with normal cochleae (Eisenman et al., 2001). Van Wermeskerken et al. found no significant differences in mean speech perception between 9 children with cochlear abnormalities (open set 48.8%, closed set 80%) and 22 congenitally deaf children with normal cochleae (open set 54%, closed set 81.5%) (Van Wermeskerken et al., 2007). Papsin also found no significant difference in speech perception outcomes for 103 children with anomalous cochleae compared with 195 children with normal anatomy (Papsin, 2005).

We used the EASD questionnaire to assess hearing development. In their report on EASD chronological changes in 10 children with hearing loss, Tomizawa et al. indicated that development curves generally increased with time; however,

the slope of these curves in stages four and five become shallower and individual differences increase (Tomizawa et al., 2013). Using the EASD assessment, we concluded that our patient's hearing ability was fully functional with hearing aids, such that normal hearing had been achieved and cochlear implantation was not needed. However, we think that the cochlear implant for the patients is more effective, and this is because the cochlear nerve seems to be preserved.

The mechanisms underlying the conventional fine-tuning ability of the normal cochlea has been explained by the traveling wave theory established by Bekesy (1947). The fact that hearing sensation is functional with the use of hearing aids suggests that patients with severe malformations retain some cochlear fine-tuning ability. However, it is unclear from the traveling wave theory how our patient with severe malformations had retained his cochlear-fine tuning ability. This may be attributed to the presence of residual inner hair cells.

Kaga et al. reported that children with serious congenital hearing loss showed poor vestibular function in the rotational chair test and delayed motor development in head control and independent walking (Kaga et al., 1981). One mechanism underlying delayed head control and independent walking may be the loss of the vestibulospinal reflex, which results in poor maintenance of sufficient muscle tension. As the child grows, central vestibular compensation can improve the acquisition of head control and independent walking.

A study on the vestibular function of patients with serious congenital hearing loss by the rotational chair test showed that the frequency of per-rotatory nystagmus increased across tests. This suggests that children with severe malformations can

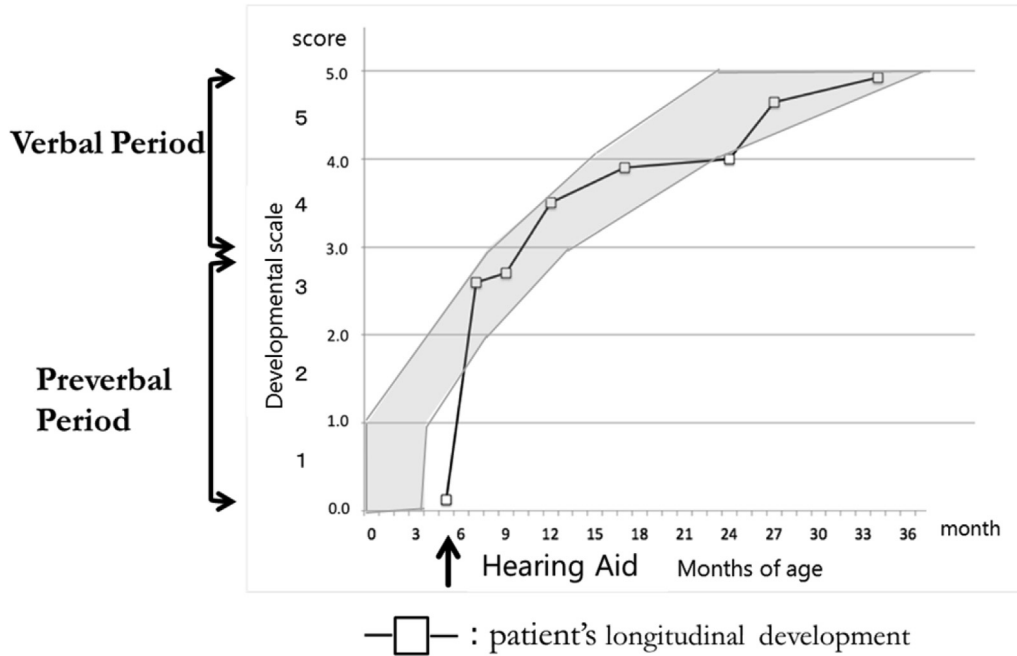
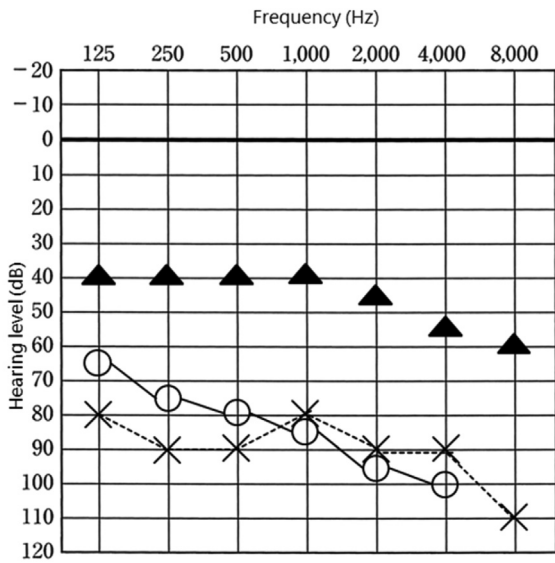


Fig. 6. Early Auditory Skill Development for Special Population scores. The patient started to wear hearing aids at the age of 5 months. The gray area shows the normal developmental range (SD:  $\pm 1$ ).



▲ : Threshold of both ears with hearing aids

Fig. 7. Visual reinforcement audiometry using an inserted earphone at the age of 6 years. Threshold level when wearing hearing aids was 41.25 dB HL.

acquire vestibular function and central vestibular sensation (Masuda and Kaga, 2014). Our patient underwent the rotational chair test at the age of six, and per-rotatory nystagmus was partially elicited, indicating that his vestibulo-ocular reflex had been preserved. This finding suggests that the patient may have possessed residual vestibular sensory cells that enabled vestibular function recovery upon maturity.

#### 4. Conclusion

Our patient had severe malformations of the inner ear, namely, common cavity deformity and IP-I. Auditory–verbal rehabilitation with hearing aids was effective, and he acquired gross motor function. These findings suggest that auditory–vestibular hair cells may have been partially preserved in this patient.

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