Available online at www.sciencedirect.com
ScienceDirect
Journal of Otology xx (2017) 1–6
www.journals.elsevier.com/journal-of-otology/

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

Yusuke Kimura [a,b,*], Takeshi Masuda [a,b], Akifumi Tomizawa [c], Hideaki Sakata [c,d], Kimitaka Kaga [a]

[a] National Institute of Sensory Organs, National Tokyo Medical Center, Tokyo, Japan
[b] Department of Otolaryngology–Head and Neck Surgery, Nihon University School of Medicine, Tokyo, Japan
[c] Department of Speech, Language and Hearing Therapy, Mejiro University, Saitama, Japan
[d] Kawagoe Otology Institute, Saitama, Japan

Received 19 January 2017; revised 3 March 2017; accepted 8 March 2017

**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.


*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.

* Corresponding author. Department of Otolaryngology–Head and Neck Surgery, Nihon University School of Medicine, 30-1 Oyaguchi Kami-cho, Itabashi-ku, Tokyo 173-8610, Japan. Fax: +81 3 3972 1321.
*E-mail address:* yusuke0617@gmail.com (Y. Kimura).
Peer review under responsibility of PLA General Hospital Department of Otolaryngology Head and Neck Surgery.

http://dx.doi.org/10.1016/j.joto.2017.03.002

## 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).

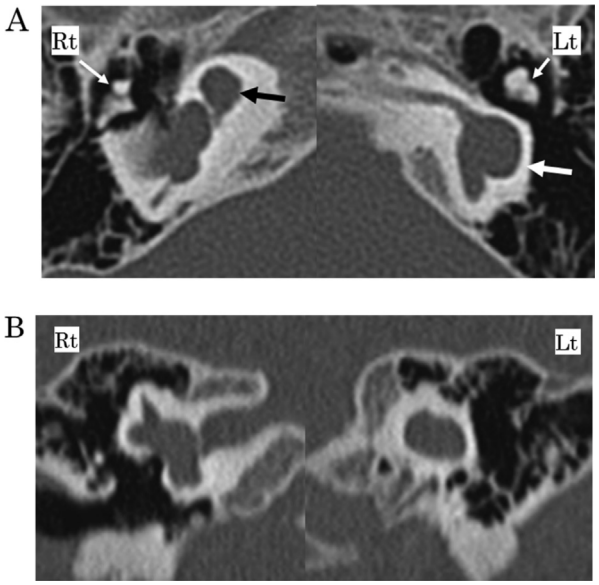


Fig. 2. (A) Axial slice of computed tomography image. (B) Coronal slice of computed tomography image demonstrating IP-I (black arrow) and common cavity deformity (white arrow). The ossicles showed no malformations (thick white arrow).

### 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 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

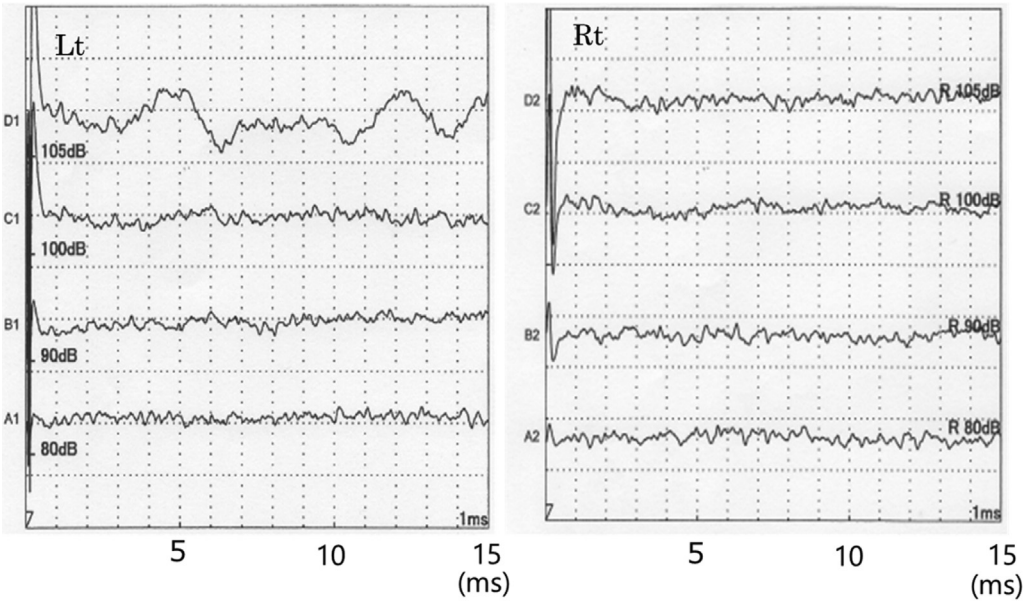


Fig. 1. Auditory brain stem response at the age of 3 months. Auditory brain stem response testing revealed no waveforms in this patient.

Please cite this article in press as: Kimura, Y., et al., A child with severe inner ear malformations with favorable hearing utilization and balance functions after wearing hearing aids, Journal of Otology (2017), http://dx.doi.org/10.1016/j.joto.2017.03.002