Case Report

A Case Report of Rapid Recovery in Speech Perception after Cochlear Implantation in a Female Child with Post-Meningitis Deafness

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Highlights

- Improved speech perception can occur rapidly after CI in post-meningitis deafness
- Deafness duration and accurate CI mapping are vital for optimal speech outcomes

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ABSTRACT

Background: It takes some time for a patient to adapt to a Cochlear Implant (CI). Usually, the improved speech perception after cochlear implantation is reported within 3–6 months. In this study, we reported a case of a female child with post-meningitis deafness who showed considerable recovery in speech perception just a few days after cochlear implantation.

The Case: The case was a 14-year-old female with complaints of severe headache, delirium, unresponsiveness to sound, and agitation, diagnosed later with meningitis. Pure tone audiometry showed total deafness in her right ear and severe to profound sensorineural hearing loss in her left ear. Seven months after deafness, she received a CI (Nucleus CI512) in the right ear. Its speech processor was activated two weeks after surgery. The first map was programmed using the advanced combination encoder strategy, along with behavioral measurements of T-levels and C-levels. Four days later, the audiometric test revealed a Pure-Tone Average (PTAve) of 35 dB HL, accompanied by a Speech Discrimination Score (SDS) of 72%. The Bamford-Kowal-Bench (BKB) sentence test yielded a score of 80% in silence. Two weeks after device activation, PTAve was 20 dB HL, the SDS was 86% and the BKB sentence test score in silence was 100%.

Conclusion: It seems that factors such as short duration of deafness, precise mapping of the CI speech processor, consistent device usage, and rich aural environment can lead to significant improvement in speech perception within two weeks after cochlear implantation in patients with post-lingual deafness.

Keywords: Cochlear implant; post-lingual deafness; children; meningitis; case report

Introduction

Meningitis is one of the most common causes of profound hearing loss in children [1]. The infection spreads to the cochlea through the cochlear aqueduct, which connects the posterior fossa to the scala tympani close to the round window membrane [2]. Inflammation of the endocochlear tissue can lead to new bone formation inside the cochlea. Ossification of the cochlear lumen may occur in a wide range of degrees. In some cases, it may not happen at all. In some cases, it can be limited to the first millimeters of the scala tympani or extend to the apical regions in varying extents [1]. Cochlear implantation is the main treatment for profound hearing loss caused by meningitis, and the ossification of the cochlear lumen is a problem for the treatment of hearing loss.

It takes some time for a patient to adapt to a cochlear implant. In post-lingually deaf patients, improvements in speech perception are typically observed within 3 to 6 months after cochlear implantation [3, 4]. Performance on speech perception tests generally reaches a plateau after 1 to 2 years [5]. In this study, we described a case with post-lingual deafness due to meningitis whose speech perception recovered extraordinarily just a few days after cochlear implantation.

Case presentation

The case was a 14-year-old female referred to the Guilan Cochlear Implant Center (GCIC) in February 2024. A few weeks before that, she had been referred to the children's hospital with complaints of severe headache, delirium, unresponsiveness to sounds, and agitation. After extensive examinations including abdominal and pelvic ultrasound scan, brain and thoracic and lumbar Magnetic Resonance Imaging (MRI), lumbar puncture, blood and urine tests, she was diagnosed as having meningitis and post-meningitis vasculitis. She was prescribed a high dose of oral corticosteroids for two weeks. Pure tone audiometry showed total deafness in the right ear and severe to profound sensorineural hearing loss with a Speech Discrimination Score (SDS) of 48% in the left ear. Auditory brainstem response and otoacoustic emissions were absent bilaterally. She was advised to receive CI in the right ear. Since the patient and her family were shocked by the report and did not accept the urgency of the situation for cochlear implantation, they refused the suggested treatment and left the clinic. Six months later, they came back to us frustrated and depressed. Meanwhile, the patient had tried hearing aids but was not satisfied. Once more, they were consulted for CI and accepted to proceed with this treatment. A High-Resolution Computerized Tomography (HRCT) scan showed no sign of ossification through the cochlear duct (Figure 1). MRI reconstruction of the inner ear also showed that the bony labyrinth was filled with fluids (Figure 2).

A Cochlear Nucleus CI512 implant was selected to be implanted in the right ear. After a normal 4-cm retroauricular incision, a mastoidectomy with facial recess approach was performed to access the cochlea. The round window niche was drilled away to improve the visualization of the round window membrane.

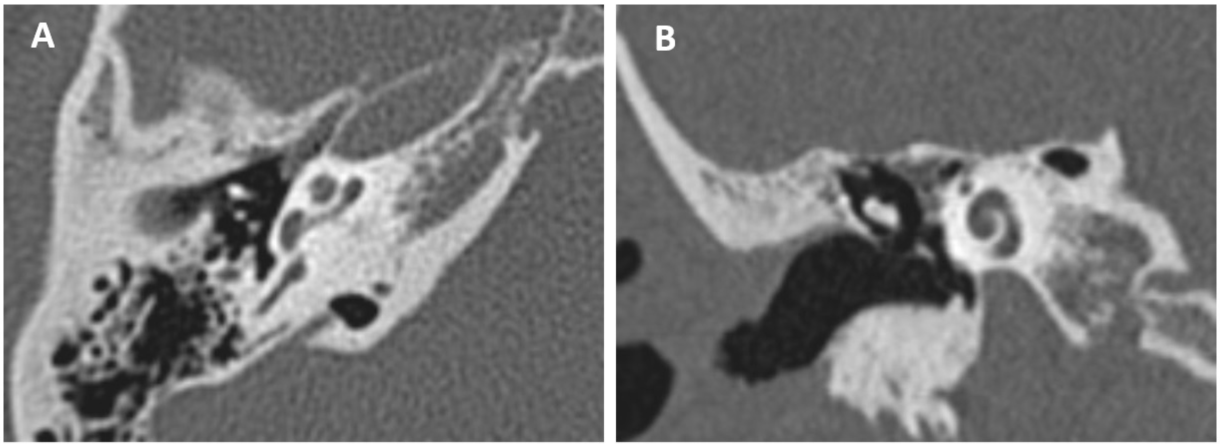

Figure 1. Pre-operative computerized tomography scan images of the right ear from axial (A) and coronal (B) views

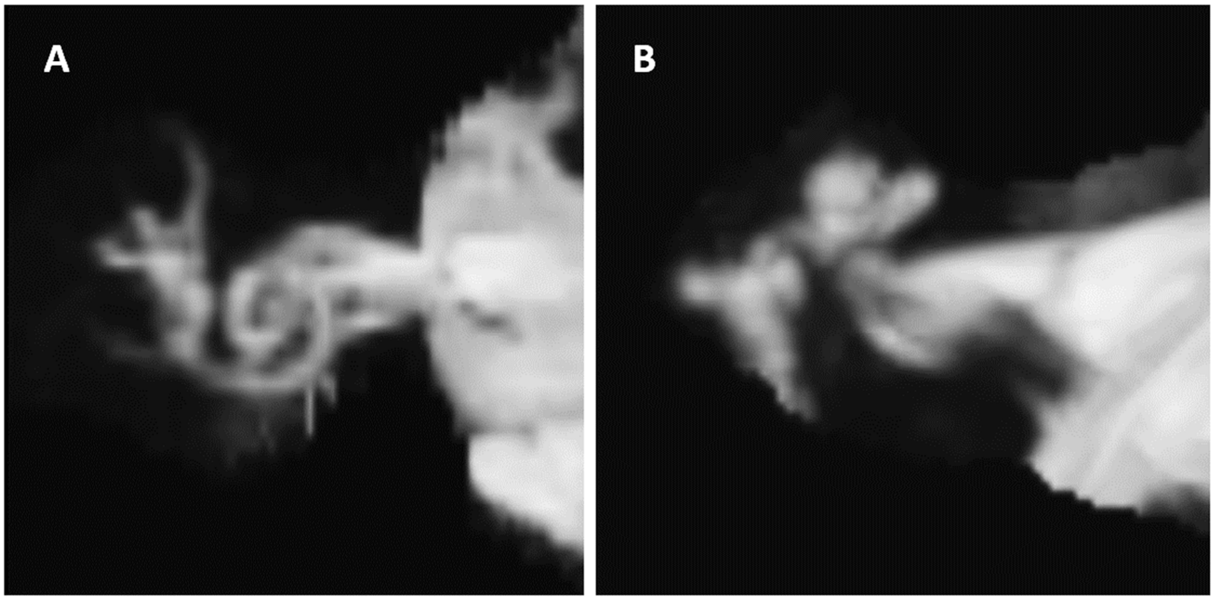

Figure 2. T2-weighted 3-dimensional magnetic resonance imaging reconstruction of right ear labyrinth from anterior (A) and inferior (B) views

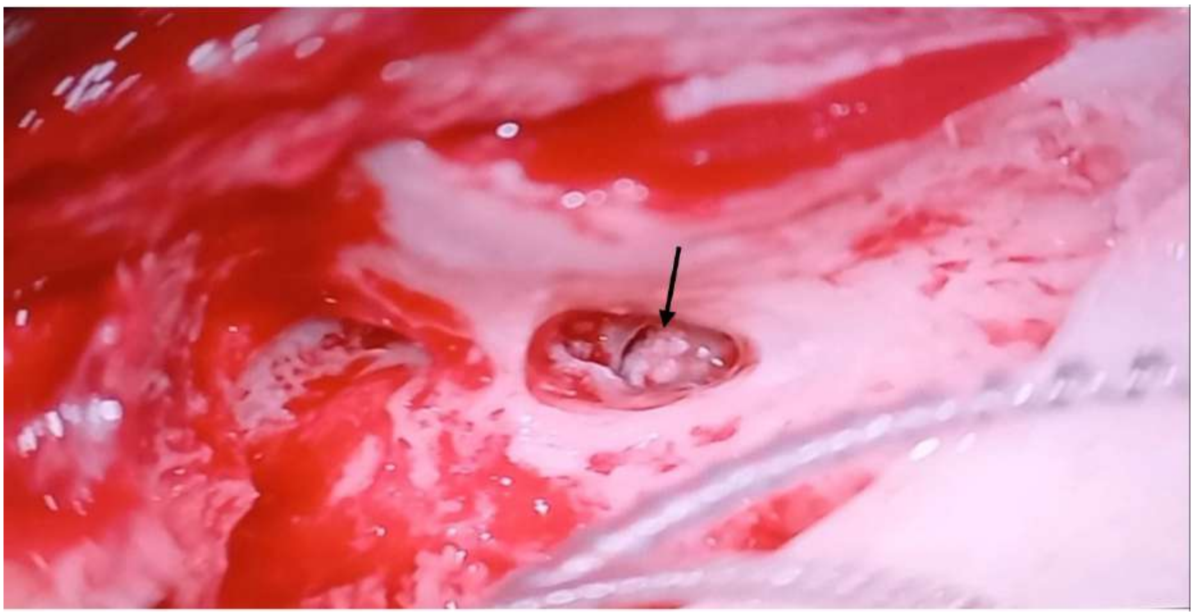

Figure 3. Surgical view of the right ear round window through the facial recess indicates bone formation

The round window niche was then obliterated, and the opening of the cochlea was found to be obstructed (Figure 3). The intraluminal line-of-sight removal of bone was done to bypass the obstruction and reach the open lumen distally. Full insertion of electrodes was done with a classic insertion method. Impedance testing and Neural Response Telemetry (NRT) were performed to ensure the integrity of the device.

Results

First mapping

The speech processor of the CI (Nucleus 7 SE) was activated two weeks after surgery. No complications occurred in relation to surgery. All electrode contacts had a normal impedance. The first map was programmed

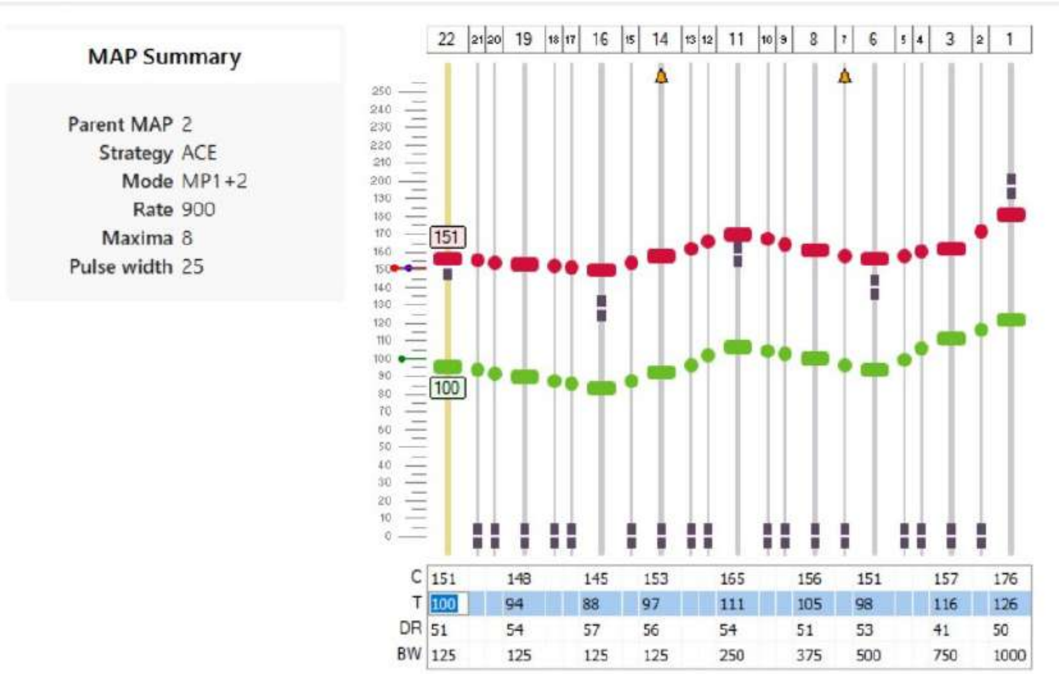

Figure 4. The last mapping parameters of the patient

using the Advanced Combination Encoder (ACE) strategy. The map parameters included: stimulation mode=MP1+2, rate=900 Hz, Maxima=8, and pulse width=25 µsec. Behavioral measurement of T-levels and C-levels was done using five electrodes (i.e., numbers 1, 6, 11, 16, and 22) and then interpolated for the remaining electrodes. The patient was instructed to use the processor during wake times and to return four days later for the next visit. To help improve her listening skills and speech comprehension, she was advised to participate in listening exercises. The exercise included: (a) listening to a short text from her school literature textbook presented several times when the speaker's lips were visible, (b) trying to identify and repeat two- to three-word phrases from the text, presented to her without lip-reading, (c) performing the exercise at least three times a day, each for 20 minutes.

Second mapping

During the second mapping session, data logging indicated an average daily device use of approximately 12 hours over the preceding four days. The second map was created by measuring the T-levels and C-levels using the same five electrodes mentioned in the first session, and interpolating for other electrodes. She was very pleased with her hearing and mentioned that she can understand speech very well. Audiometric test revealed a pure-tone average of 35 dB HL, accompanied by a Speech Discrimination Score (SDS) of 72%. Additionally, the Bamford-Kowal-Bench (BKB) sentence test yielded a score of 80% in silence. All speech assessments were carried out in a sound field at a calibrated presentation level of 65 dB SPL (A-weighted). The next appointment was set for 10 days later.

Third mapping

Two weeks after device activation, the patient came back for the third mapping session. The average daily device use was 14 hours. The average electrode impedances were decreased to 9 kΩ. The neural response telemetry was performed using the five electrodes mentioned above, and normal responses were recorded. The third map was created by measuring the T-levels and C-levels using nine electrodes (i.e., numbers 1, 3, 6, 8, 11, 14, 16, 19, and 22) through behavioral assessment. The patient mentioned that she was fully recovered and does not need any more mapping sessions. In the audiometric evaluation, the pure-tone average was 20 dB HL, the SDS was 86%, and the BKB sentence test score in silence was 100%. Figure 4 shows the details of the third map. We have obtained written informed consent to publish the details from her parents.

Discussion

Meningitis is a serious infectious disease that affects different age groups. It is among the most prevalent causes of acquired profound sensorineural hearing loss in children, and can be associated with labyrinthitis ossificans [2], which can make electrode insertion difficult or even impossible [6]. Labyrinthitis ossificans can usually be diagnosed using HRCT; however, in some cases, false-negative results may occur [7]. Moreover, fibrotic obstruction of the cochlea is difficult to identify with HRCT. It is recommended to use MRI to provide detailed images of the fluid compartments within the cochlea, enabling the identification of any obstruction caused by bacterial meningitis [8]. The sensitivity and specificity of MRI predicting intraoperative cochlear obstruction have been reported to be 94.1 and 87.5%, respectively [8]. Combining HRCT and MRI findings can increase the accuracy of cochlear obstruction diagnosis, although there may be some misdiagnoses. In our patient, both HRCT and MRI failed to identify cochlear obstruction.

Speech perception in CI patients with post-meningitis deafness has been reported to be worse than in CI patients with other etiologies. Mosnier et al. reported that in adults with post-meningitis profound hearing loss, speech performance was not improved after 6 months post-activation. They found that the patients had poorer speech scores in comparison to those in the control group. However, their performance was enhanced with new cochlear implantation technologies [9]. Helmstaedter et al. found that children with meningitis-related deafness with CI had poorer monosyllabic word perception compared to controls [10].

Learning how to process the auditory signals delivered by the CI usually requires about six months before entering a stable plateau phase [3]. There are several studies on the factors influencing speech perception in CI users. Studies have shown that precise and personalized mapping of the CI speech processor has a crucial effect on speech perception abilities [11, 12]. Durisin et al. showed that the duration of deafness after meningitis had a relationship with the auditory performance in pediatric CI recipients. Patients implanted within 6 months after meningitis had a better auditory performance and language control [13]. Similarly, in patients with post-lingual deafness, the duration of deafness is a crucial factor in predicting speech perception after cochlear implantation [5]. Oh et al. reported that adults with post-lingual deafness of less than five years had a faster rate of recovery in speech perception than those who had been deaf for more than five years [14]. Continuous usage of hearing aids before implantation provides stimulation of the cochlea and neuronal elements and supports better speech comprehension with CI [15]. In our case, high speech perception scores were achieved only two weeks after CI device activation. CI mapping was performed using meticulous behavioral measurement in nine electrode locations across the cochlear duct. Although the patient did not have proper hearing aid usage, the duration of deafness was about seven months. Short duration of deafness before CI probably has a role in speech perception outcomes.

In our study, ossification was limited to the scala tympani at the round window, and the patient had a full insertion of a pre-curved electrode via basal turn cochleostomy. A lower degree of cochlear ossification and the use of a pre-curved electrode array, as shown in our patient, are associated with better outcomes after implantation [6, 16]. The number of active electrodes within the cochlea can also affect the speech perception outcomes in CI users [17]. Consistent CI device usage and a rich aural environment are among other important factors involved in speech perception [18]. In our case, the patient had 22 well-functioning intra-cochlear electrodes. Also, she had used her speech processor for nearly 14 hours per day and had a supportive family and friends who provided a rich listening experience for her. These results suggest that a rapid recovery of speech perception may be achieved in patients with post-lingual deafness. If the duration of deafness is short, the patient is motivated, and the family can support them through the implementation process.

Conclusion

In this study, a considerable improvement in speech perception was reported within two weeks after cochlear implantation in a female child with post-meningitic deafness of seven months' duration. It seems that factors such as the short time interval between post-lingual deafness and cochlear implantation, precise mapping of the Cochlear Implant (CI) speech processor, consistent CI device usage, and a rich aural environment can lead to extraordinary improvement in speech perception within a few weeks after cochlear implantation in patients with post-lingual deafness.

Ethical Considerations

Compliance with ethical guidelines

The study approved by the Ethics Committee of Guilan University of Medical Sciences; approval ID: IR.GUMS.REC.1404.081. Written informed consent was obtained from the parents of the patient for her anonymized information to be published in this article.

Availability of data and material

Not applicable

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Authors' contributions

RP: Drafting the manuscript, interpretation of the results, preparation of the figures; SN: Study design, drafting the manuscript; MMJ: data collection, reviewing the manuscript.

Conflict of interest

The authors declare no competing interests.

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