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Case Report

Uncommon Post-Meningitis Hearing Threshold Improvement: A Case Report

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Bacterial meningitis may cause inner ear fibrosis and progressive cochlear ossification with irreversible profound hearing loss (HL). Recognition of potential ossification is essential for effective management.

We present a clinical case of a 4 year old boy who developed a progressive HL starting 3 weeks after meningitis. For the prospective risk of cochlear ossification, bilateral cochlear implantation (CI) was performed. Unexpectedly, unaided hearing threshold began to show improvement on the left ear, starting 4 months after meningitis and continuing for years post CI surgery. In order to explore the residual cochlear function, a trial of exclusively acoustic amplification was performed on the improved left side 5 years post implantation, providing good results.

A certain degree of hearing recovery may be expected after meningitis related deafness. This case encourages surgeons to always adopt atraumatic surgical techniques that can enable the preservation of cochlear structure and residual function after CI surgery.

KEYWORDS: Hearing loss, meningitis, cochlear implant, hearing threshold improvement

INTRODUCTION

Bacterial meningitis is a well-recognized cause of sensorineural hearing loss (HL), affecting up to 10% of survivors when *Neisseria Meningitidis* is the causative agent [1]. Corticosteroids started before the first antibiotic dose are useful in preventing neurological sequelae in *Hemophilus Influentiae* meningitis, but their efficacy in other cases is still debated [2]. The pattern and stability of post-meningitic HL is not well known: some studies report fluctuations, others deterioration or even improvement of the hearing threshold as the result of the infection [3-4]. Importantly, meningitis may cause inner ear fibrosis and progressive ossification leading to obliteration of the cochlear fluid spaces and secondary profound HL. Recognition of potential ossification, which may possibly evolve in few weeks, is essential for effective management [5-6]. In case of post-meningitis severe-profound HL, prompt cochlear implant (CI) surgery is often required. In presence of cochlear ossification, implantation may still be attempted, but modification of technique may be necessary, involving a drill-out of the cochlea, placement of the electrode into the scala vestibule or placement of a double electrode array. CI may gain excellent results in deaf meningitis survivors, even though the performance in ossified cochlea is poorer compared to deaf children with non-ossified cochleae [7].

We present the case of a child with post-meningitic bilateral severe-profound HL that was successfully treated with bilateral CI, and, in whom a considerable hearing threshold improvement started 4 months after CI surgery.

CASE PRESENTATION

This work has been conducted with the informed and overt parental consent, in accordance with the Code of Ethics of the World Medical Association (Declaration of Helsinki) and the standards established by the local Institutional Review Board.

This study was presented at the Mediterranean Society of Otology and Audiology (MSOA) Congress, 13-16 May 2018, Jerusalem, Israel.

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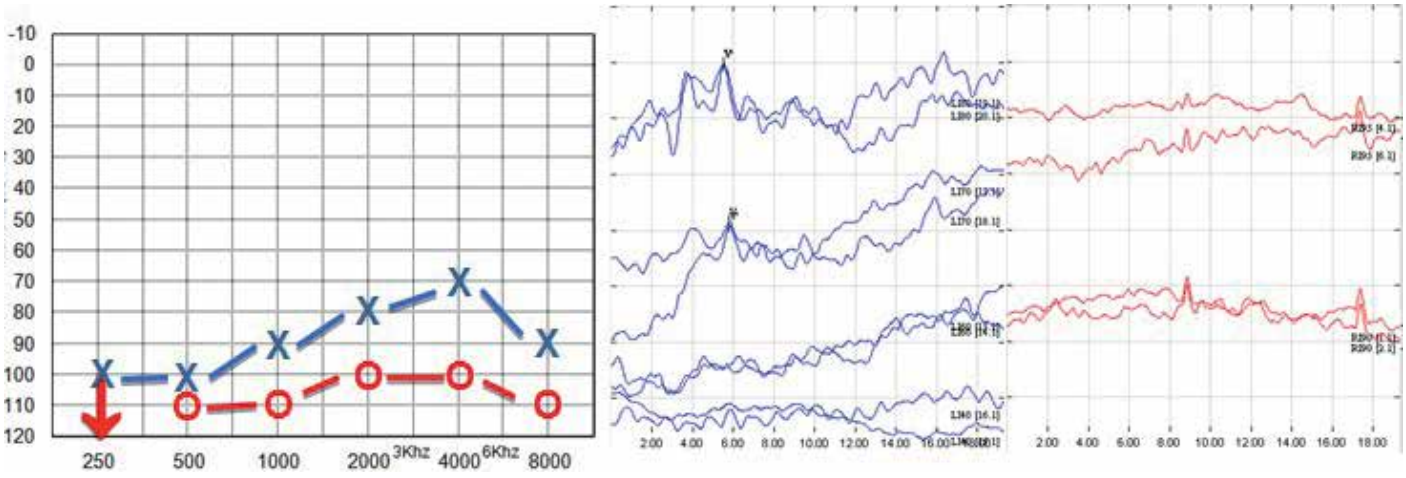


Figure 1. Hearing threshold 3 weeks after meningitis and before the CI surgery: tonal audiogram (in the left) and auditory brainsteim response (in the right).

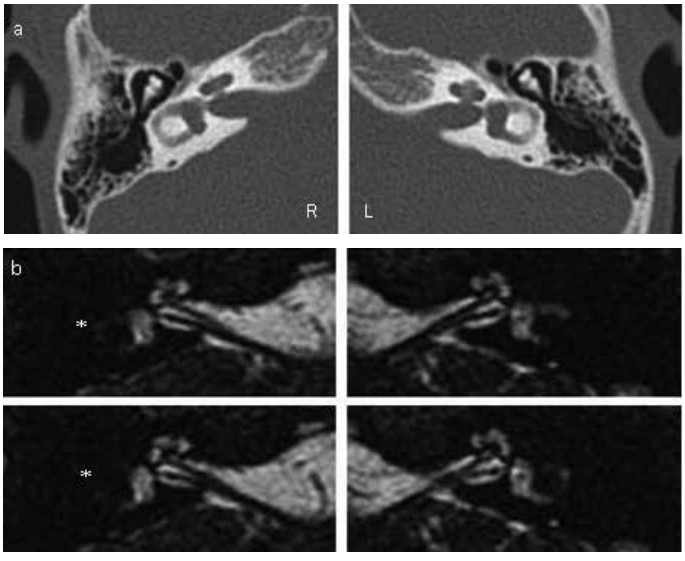


Figure 2. a, b. Inner ear imaging before CI surgery: temporal bone high-resolution computed tomography (HRCT) with apparently normal appearance on both sides (a); MRI showing decreased T2-weighted signal at the level of the semi-circular canals.
R: right side (*in b figure); L: left side

A 4 year old boy developed a *Neisseria Meningitidis type B* meningitis and was treated with intravenous Ceftriaxone. This bacterium, in association with Streptococcus pneumoniae, and H influenza type b (in areas without routine Haemophilus vaccination) are the most common meningeal pathogens in infants and small children [8].

Despite prompt infection resolution, the child reported buzzing in the ears with diminished auditory capacity. Behavioral and physiological auditory measurements confirmed a rapidly progressive bilateral cochlear deficit with mean hearing threshold reaching 105 dB HL (right ear) and 85 dB HL (left) 3 weeks after meningitis onset (Figure 1).

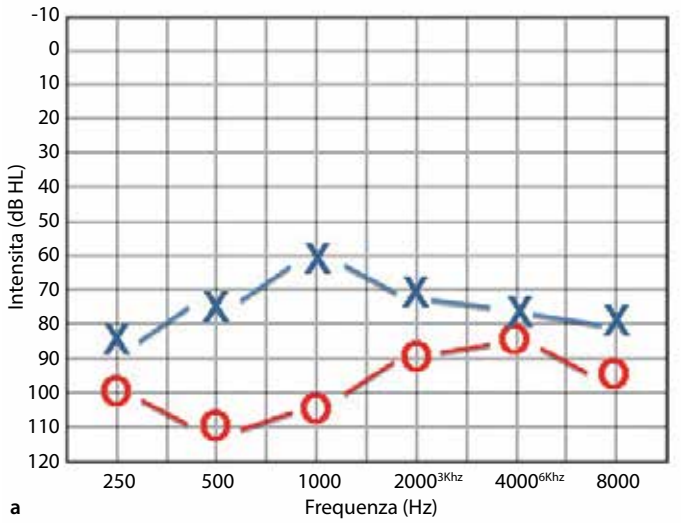
A temporal bone CT showed inner ear patency on both sides (Figure 2a), while MRI revealed a decreased T2-weighted signal in lateral semi-circular canals, interpreted as obliteration of fluid spaces due to fibrosis (Figure 2b). Due to the potential risk of cochlear ossification, simultaneous bilateral cochlear implantation was performed 45 days after infection (CI24RE receiver-stimulator with Contour Advance™ electrode array).

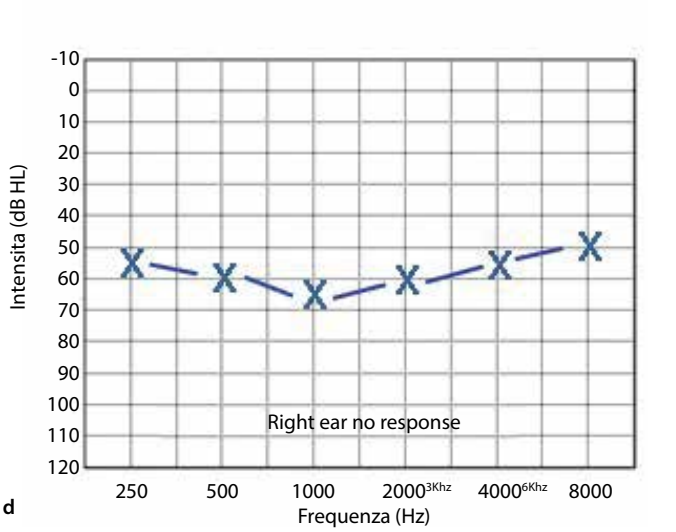
Cochlear implant surgery was performed using soft surgical technique, including slow array insertion through round window membrane incision, and both topical and systemic steroid treatment administered during and after surgery [9]. Electrode array was fully inserted as confirmed by intra-operative telemetry.

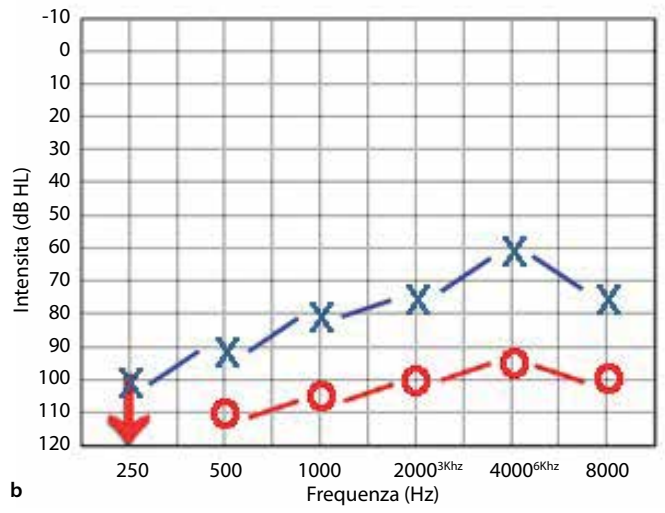
The child returned home two days after surgery. Implants were quickly accepted and the child started using the phone again about 2 months after CI activation. Neural response telemetry and impedance measurement maintained normal values without significant change over the 5 years of follow-up. Behavioral hearing threshold measurement without CI showed progression towards a total deafness on the right ear. On the contrary, left ear showed fluctuations and improvement up to a moderate HL starting 4 months after meningitis and continuing for years after CI surgery. ABR testing confirmed the subjective responses, evidencing a left wave V threshold at 60 dB nHL (Figure 3).

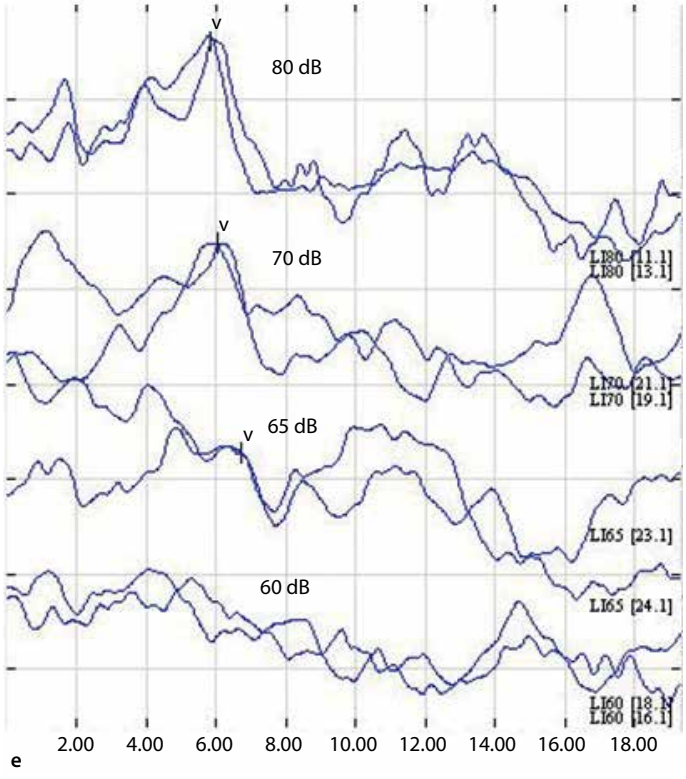
Because of the presence of auditory residuals along the whole frequency range on the left ear, we attempted to verify the efficacy of an exclusive acoustic amplification on the ear that was used to electrical hearing for many years. The acoustic amplification was not annoying to the boy and, interestingly, amplification gain preference resulted about 30% higher than the target suggested by the second-generation prescription procedures from the National Acoustic Laboratories (NAL-NL2) [10]. Free field sound detection and discrimination results without and with only the left acoustic amplification (without CI) evidenced a mean threshold (0.5-1-2-4 KHz) equal to 60 and 23.75 dB HL respectively.

Speech intelligibility was tested with monosyllabic and two-syllabic words presented at 55 dB HL in free field. Speech recognition scores improved from 22% (unaided) to 90% (aided) for the two-syllabic words, and from 15% (unaided) to 95% (aided) when monosyllabic words were presented.









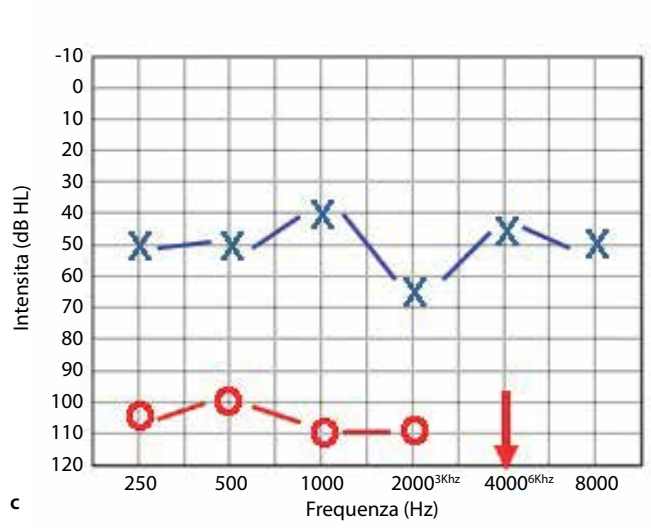


Figure 3 a-e. Hearing threshold modifications post CI: 6 months (a), 9 months (b), 2.5 years (c), 5.5 years (d) of follow-up. ABR post cochlear implant surgery (at 5.5 years of follow-up) without device (e).

DISCUSSION

This report documents the characteristics of the uncommon but possible event of a hearing threshold improvement following post-meningitis hearing impairment of severe to profound degree. In our case, meningitis led to a severe-profound HL on both sides requiring a CI surgery for the prospective risk of cochlear ossification. Starting from the fourth month after surgery the behavior of HL diversified between the 2 sides. On the right side, the residual hearing totally

deteriorated, while on the left side the auditory threshold fluctuated and improved up to a moderate level of severity. The left residual cochlear function demonstrated satisfactory discrimination skill preservation.

Bacterial meningitis is one of the most frequent cause of acquired HL [1]. Infection spread to the cochlea and intra-cochlear structures is rapid and may occur within hours of the diagnosis of meningitis, leading to a HL that is usually profound and irreversible [7]. Fibrosis and ossification of the lateral semicircular canal is a first and sensitive measure prior to evidence of cochlear involvement [5].

Early CI is critical in post meningitic severe-profound HL because the endostial inflammation can start an osteogenesis process with a resulting obliteration of the cochlear lumen [6]. Also, the fibrotic scar tissue that forms around the electrode array can severely damage the cochlear architecture, including sensory hair cells and neurons, resulting in a loss of residual hearing. Fibrotic or osseous tissue into the cochlea increases the impedance of the electrode array and consequently high levels of current are required for achieve an electrical stimulus threshold [11].

In our case, impedances and current levels did not change during CI follow-ups, even though left hearing threshold was improving and right ear threshold was declining, probably indicating the absence of intra-cochlear fibrous tissue around the CI array. The adoption of a surgical approach that is as less traumatic as possible may have partially contributed to the preservation of cochlear functionality.

Hearing threshold improvement after meningitis is rarely reported in the literature. Brookhouser et al. [3] evaluated audiological assessment in 280 children post meningitis infection. In the great majority of cases hearing threshold remained stable throughout the follow-up; 9 out of 64 children with 3 years of follow up exhibited changing in hearing threshold showing improvement, decrement or fluctuation but only 3 children showed auditory improvement in one or two ears. Roine et al. [4] reported an improvement of at least 20 dB in approximately 9% of ears during the follow up of 235 bacterial meningitis survivors with a first ABR testing indicating severe or profound HL.

The satisfactory discrimination outcomes with acoustic amplification demonstrates that a good cochlear function has been preserved in this case, and opens the likelihood of future use of new genetic or biological therapies and new hearing technologies also in patients with previous CI surgery and exclusive long term electrical hearing experience.

Moreover, this case example encourages surgeons and CI technology researchers to intensely explore techniques, therapies and technological solutions that can facilitate the preservation of inner ear structure and the still existing cochlear function during and after CI surgery in order to leave open the opportunities of new strategies adoption.

CONCLUSION

This paper presents a post-meningitic case that required an urgent CI for the prospective risk of cochlear ossification. The left hearing threshold improved gradually over a five-year period up to a HL of moderate degree and a residual cochlear function that enabled a satisfactory acoustic amplification.

A certain degree of hearing recovery may be expected after meningitis related deafness. This case encourages surgeons to always adopt atraumatic surgical techniques that can enable the preservation of cochlear structure and residual function after CI surgery.

Informed Consent: This work has been conducted with the informed and overt parental consent, in accordance with the Code of Ethics of the World Medical Association (Declaration of Helsinki) and the standards established by the local Institutional Review Board.

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Conflict of Interest: The authors have no conflicts of interest to declare.

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