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Cochlear implantation in obliterated cochlea: A case study

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Abstract
Bacterial meningitis is the most common aetiology for acquired hearing loss. One of the most common sequelae of meningitis which has high impact on general functioning is hearing loss, either unilateral or bilateral, and varying from mild to profound. This article aimed to profile a case of 14 year old cochlear implant user who presented with hearing impairment in both ears, post meningitis with bilateral labyrinthine ossification. The patient underwent scala vestibuli insertion of cochlear implant. Scala vestibuli insertion of cochlear implantation showed significant improvement in speech perception in this case with extensive fibrosis and ossification of the basal turn of left cochlea. Scala vestibuli insertion offers a valuable alternative in cases of obstructed scala tympani that can be employed for a variety of etiologies.

Keywords: scala tympani, cochlear implants, meningitis

Introduction
Bacterial meningitis is the most common aetiology for acquired hearing [1]. One of the most common sequelae of meningitis which has high impact on general functioning is hearing loss, either unilateral or bilateral, and varying from mild to profound. 17. 5% to 35% of patients with bacterial meningitis will develop permanent sensorineural hearing loss, and profound bilateral hearing loss will occur in up to 4% of patients 13-15. 90% of acquired profound hearing impairment in children are probably due to bacterial meningitis [12]. It has been observed that this occurs in the initial phase of illness [2]. The exact mechanism of hearing loss in patients with bacterial meningitis is not well understood and is likely due to multiple factors that include direct labyrinth involvement, cochlear neuroepithelial damage, and vascular insult [5]. Hearing impairment has significant consequences for a child's communication, educational achievement, and emotional and social wellbeing [3]. Early assessment of hearing in children after meningitis is recommended [4]. The early diagnosis and treatment of hearing loss has a high socio-economical value. By identifying children at risk for development of hearing loss, early rehabilitation may lessen long-term adverse outcomes [5]. Early identification is essential for successful rehabilitation, including cochlear implantation [6]. The surgical technique and the insertion technique are largely standardised, and regular CI electrodes can be inserted in the majority of cases. Special cases, however, such as the implantation of patients with anatomical malformations, obliterations of the cochlea, or re- implantations pose a challenge in cochlear implantation. The cochlear duct can be obliterated by osteoneogenesis within a few months of meningitis, which may make implantation ineffective, or impossible [7]. When the cochlea is obliterated or ossified, alternative surgical techniques, such as incomplete insertion, the implantation of double arrays [6], implantation into the scala vestibuli [7, 8], or a radical cochleostomy, must be considered. All these methods are associated with some disadvantages, such as the poor performance of the implant. However, despite a significantly higher risk for injuring the facial nerve, the internal carotid artery, or the modiolus [9], such alternative procedures are recommended in cases of partial and complete ossification.
We report a case of 14 year old cochlear implant user who presented with hearing impairment in both ears, post meningitis with bilateral labyrinthine ossification.

Case report

A 14 year old male presented with acquired sensorineural hearing loss in both ears. The patient was diagnosed with pyogenic meningitis and was treated immediately. Post treatment he noticed reduction in hearing sensitivity, and his audiological evaluation revealed bilateral profound hearing loss. The speech audiometry was carried out and could not repeat spondee and PB words. The impedance audiometry revealed bilateral A type tympanogram with ipsilateral and contralateral reflexes absent suggestive of normal middle ear status. DPOAEs were absent in both ears. BERA results revealed no clear and repeatable V peak obtained at 96 dBnHL click stimuli at 11.1 / sec repetition rate, suggestive of profound hearing loss in right ear and clear and repeatable V peak obtained till 85 dBnHL click stimuli at 11.1 / sec repetition rate, suggestive of severe hearing loss in left ear. The aided responses were out of speech spectrum.

A detailed speech and language evaluation was conducted. His speech and language skills were observed to be age adequate. The speech perception was significantly low. His revised Category of Auditory Perception (CAP) score was zero, and he was found to be unaware of environmental sounds.

The MRI brain plane with temporal bone revealed no significant abnormality detected in brain parenchyma or bilateral inner ear structures. Reduced T_2 hyperintense signal of basal turn of right cochlea and anterior aspect of basal turn of left cochlea. Completely absent lumen of bilateral semicircular canals with reduced signal of bilateral vestibule, features are suggestive of bilateral labyrinthitis ossificans. Tiny cyst in the inferior aspect of flocculus of cerebellum on right side-arachnoid cyst.

After detailed audiological and speech and language evaluations, cochlear implantation surgery was recommended.

During surgery extensive fibrosis and ossification of basal turn of cochlea was observed. Attempted dilatation of scala tympani failing which attempted scala vestibuli insertion. Electrodes were inserted till 15th electrode. The cochlear implantation was carried out in the left ear with Cochlear Nucleus CI24RE Cochlear Implant with Straight Electrode. During intraoperative measures, impedance measurements revealed open circuit in 22nd electrode and high impedance values for electrodes from 1 to 6. Neural response telemetry was carried out, no responses were obtained across all active electrodes.

The switch on was done two weeks after the surgery. The post-operative measures were similar to intraoperative measures. The switch on was carried out based on behavioural responses. The patient had better auditory perception at pulse width 37.

Following the switch on with CP802 sound processor, the patient started attending auditory training. After six months of therapy, he was observed to have a significant progression in speech perception. The revised CAP score at this point was 11, which suggested that he uses the telephone with familiar speakers.

Discussion

Scala vestibuli insertion of cochlear implantation also showed significant improvement in speech perception in this case with extensive fibrosis and ossification of the basal turn of left cochlea. Post-operative results were comparable to patients in whom conventional scala tympani insertion was performed. No adverse effects related to the site of insertion were observed. Scala vestibuli insertion offers a valuable alternative in cases of obstructed scala tympani that can be employed for a variety of etiologies [8].

The main limitation of this study is the lack of premorbid hearing thresholds of the client studied. Also, more extensive study with increased sample size will guarantee confirmation of the results.

Conclusion

Bacterial meningitis often leads to profound hearing loss, especially in children. Early detection and intervention are vital. Cochlear implantation, though challenging due to ossification, offers hope. Alternative techniques like scala vestibuli insertion show promise. Further research with larger cohorts is necessary for comprehensive management strategies.

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