



Case Report

Cochlear Implantation in Patient with Cochlear Nerve Deficiency: A Case Report

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Diagnostic imaging methods are very important for patients with bilateral sensorineural hearing loss. Magnetic resonance imaging (MRI) is able to demonstrate the vestibulocochlear nerve and facial nerve in the internal acoustic canal. Also computed tomography can be helpful to determination of the deficiency of the cochlear nerve. Cochlear nerve anomalies are classified into three group according to the magnetic resonance imagings. Patients who have cochlear nerve deficiency may not hear with cochlear amplifications. Auditory brain stem implant (ABI) is most suitable for these cases. In this case report, we presented a 3 years old girls with bilateral totally hearing loss who performed cochlear implantation despite of cochlear nerve deficiency on magnetic resonance imaging (MRI) and computed tomography (CT).

KEYWORDS: Deafness, cochlear nerve anomalies, cochlear implantation

INTRODUCTION

Cochlear implantation is an effective method for the rehabilitation of bilateral total hearing loss. House ^[1] first described this procedure in 1961. This device was developed during past decades and development of the device was resulted better hearing and recognition outcomes ^[2]. These technological developments led to wide indications for cochlear implants. However, cochlear implantation in patients with inner ear malformations is still controversial. Especially, patients with cochlear nerve deficiency are not suitable for cochlear implantation. This situation is absolute contra-indication of the cochlear implantation.

Diagnostic imaging methods are very important for the patients with bilateral sensorineural hearing loss. Magnetic resonance imaging (MRI) is able to demonstrate the vestibulocochlear nerve and facial nerve in the internal acoustic canal ^[3]. Also computed tomography can be helpful to determination of the deficiency of the cochlear nerve ^[4,5]. Cochlear nerve anomalies are classified into the three group according to the magnetic resonance imaging ^[6]. Type I is total absence of the vestibulocochlear nerve. Type IIa is absent or hypoplastic cochlear nerve or dysplasia of the vestibulocochlear labyrinth. Type IIb is absent or hypoplastic cochlear nerve and vestibulocochlear labyrinth is normal. Patients who has cochlear nerve deficiency may not benefit from cochlear implants. Auditory brain stem implant is most suitable for these cases. But some reports suggested that patients with aplasia or hypoplasia of the cochleovestibular nerve can be treated and hear with cochlear implantations ^[6].

In this case report, we presented a 3 years old girls with bilateral totally hearing loss who performed cochlear implantation despite of cochlear nerve deficiency on MRI.

CASE PRESENTATION

A girl patient was born by caesarian section with uncomplicated at secondary care center. Bilateral totally hearing loss was noticed that she was at the age of 25 months. She was referred to our hospital for advanced hearing evaluations. Tympanic membrane was seen normal and other otorhinolaryngologic examinations were seen normal. Facial nerve functions were seen normal. Otoacoustic emissions were bilaterally absent. Evoked brain stem response did not show any hearing thresholds. According to these findings, she was used bilateral hearing aids and she was evaluated for cochlear implantations. There were not seen any neurological and psychiatric contraindication for cochlear implant surgery. High resolution computed tomography (CT) of the temporal bone showed that bilateral external auditory canal and tympanic cavity were seen normal. Bilateral cochlear hypoplasia was seen and posterior and lateral semicircular canals were not seen. Bilateral internal acoustic canal were seen symmetric and normal (Figure 1). Magnetic resonance images showed that cochlear nerve was not seen bilaterally (Figure 2, 3). Left cochlear aplasia and right cochlear dysplasia were noticed. Electric stimulation of the promontorium could not elicit response at both ears. Right cochlear implantation was performed to the patient when she was 30 months old (Med-EL sonata, Med-EL; Innsbruck, Austria). Promontorium cochleostomy was performed and full electrode insertion was checked intraoperative. There was not seen any intraoperative

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Figure 1. Bilateral internal acoustic canal were seen normally on computed tomography.

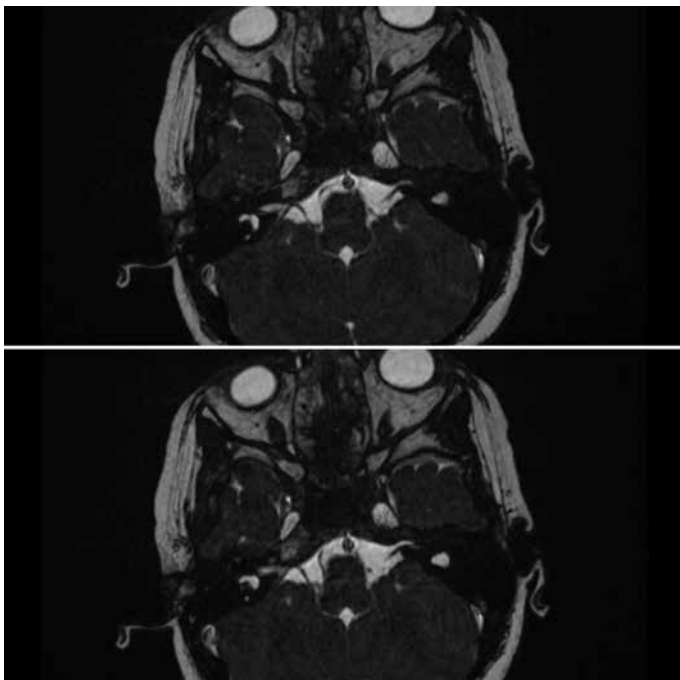


Figure 2. Axial T2 weighted MR images showed that congenital absence of the cochlear nerve.

neural response threshold (NRT). After one year, the audiological test showed thresholds of 40 dB HL and Meaningful auditory integration scale (MAIS) test result was 22/40 and listening progress profile (LIP) test result was 37/42. We received informed consent from the patient for the publication of this report.

DISCUSSION

Cochlear nerve anomalies are seen very rare in the populations. But cochlear nerve agenesis or hypoplasia is challenging situations for the patients and their physicians. McClay et al. [3] reported that cochleovestibular nerve anomalies were seen in 18% of children with sensorineural hearing loss. To be aware that situation, neuroimaging of the all patients should examined carefully and all kind of audiological tests including otoacoustic emissions, brainstem response should performed to the patients with sensorineural hearing loss. Patients who has cochlear nerve anomalies especially

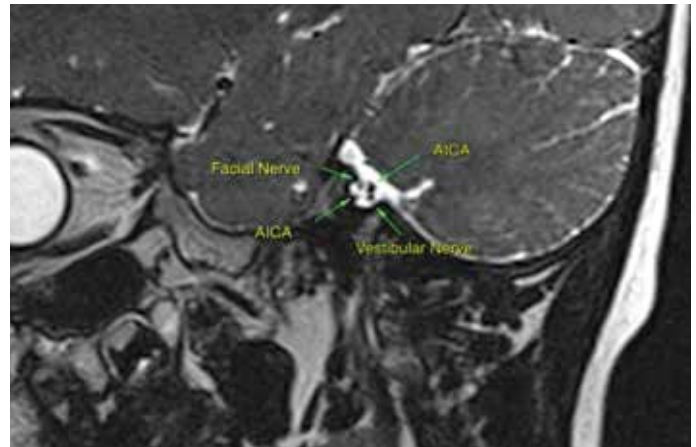


Figure 3. Oblique-sagittal T2 weighted MR image of patients with congenital totally hearing loss showed that congenital absence of the cochlear nerve parasagittal reconstruction of the MRI (AICA: anterior inferior cerebellar artery).

cochlear nerve agenesis and hypogenesis usually may not benefit from cochlear implantation. Most authors suggest that cochlear implant surgery is not suitable for these cases. Brain stem implantation may be better therapeutic option than cochlear implantation for these patients [7,8]. In a study of Govaerts and et al. [6] showed that patients with type I and type IIb aplasia did not auditive perception with their implant and they became non-users. The patients with type IIa aplasia and hypoplasia had moderate audiological results with the cochlear implant. Zhang et al. [9] reported nine cases with cochlear nerve deficiency. They found that only four children had a significant improvement in pure tone average threshold with cochlear implantation.

Electric evoked auditory brainstem response (eABR) may helpful the decision of the surgery. This test should be performed before surgery especially patients with cochlear nerve anomalies. It is critical in the evaluation of cochlear nerve anomalies. But some studies suggested that eABR is not useful because of its poor prognostic value and false positive results in general [6].

Cochlear implantation may be an option in some cases of cochlear nerve anomalies. Although it has limited effect and not cost effective, some authors have reported that patients who is seen cochlear nerve anomalies in radiological imagings may achieve good outcomes with cochlear implantation [6]. MRI may not determine the thin cochlear nerve. A very thin cochlear nerve cannot seen on MRI and it can intermingled with facial or vestibular nerve so may reported as cochlear agenesis. A few cochlear nerve branches could deliver some acoustic information to the auditory center [10].

In our case, we saw cochlear hypoplasia on temporal CT and there was not seen cochlear nerve on inner ear MRI. We performed to the patients an eABR but test result was reported as not response. But after cochlear implantation we saw good outcomes in term of audiological thresholds and speech recognitions.

In conclusion, cochlear implantation in patients with cochlear nerve anomalies is still controversial but some selected cases may be resulted with good audiological outcomes with cochlear implantation. It can

be an expensive trial for these patients but it has less complications and an easy method compared with surgery of brainstem implant.

Ethics Committee Approval: N/A.

Informed Consent: Written informed consent was obtained from the patient who participated in this study.

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REFERENCES

1. House WF. Cochlear implants. *Ann Otol Rhinol Laryngol* 1976; 85: 1-93.
2. Clark Gm. Personal reflections on the multichannel cochlear implant and a view of the future. *J Rehabil Res Dev* 2008; 45: 651-93. [\[CrossRef\]](#)
3. McClay JE, Booth TN, Parry DA, Johnson R, Roland P. Evaluation of pediatric sensorinural hearing loss with magnetic resonance imaging. *Arch Otolaryngol Head Neck Surg* 2008; 134: 945-52. [\[CrossRef\]](#)
4. Parry DA, Booth T, Roland PS. Advantages of magnetic resonance imaging over computed tomography in preoperative evaluation of pediatric cochlear implant candidates. *Otol Neurotol* 2005; 26: 976-82. [\[CrossRef\]](#)
5. Glastonbury CM, Davidson HC, Harnsberger HR, Butler J, Kertesz TR, Shelton C. Imaging findings of cochlear nerve deficiency. *AJNR Am J Neuroradiol* 2002; 23: 635-43.
6. Govaerts PJ, Casselman J, Daemers K, De Beukelaer C, Yperman M, De Ceulaer G. Cochlear implants in aplasia and hypoplasia of cochleovestibular nerve. *Otol Neurotol* 2003; 24: 887-91. [\[CrossRef\]](#)
7. Colletti V, Fiorino F, Sacchetto L, Miorelli V, Carner M. Hearing habilitation with auditory brain stem implantation in two children with cochlear nerve aplasia. *Int J Ped Otorhinolaryngol* 2001; 60: 99-111. [\[CrossRef\]](#)
8. Colletti V, Carner M, Fiorino F, Sacchetto L, Miorelli V. Hearing restoration with auditory brainstem implantation in three children with cochlear nerve aplasia. *Otol Neurotol* 2002; 23: 682-93. [\[CrossRef\]](#)
9. Zhang Z, Li Y, Hu L, Wang Z, Huang Q. Cochlear implantation in children with cochlear nerve deficiency: A report of nine cases. *Int J Ped Otorhinolaryngol* 2012; 76: 1188-95. [\[CrossRef\]](#)
10. Zanetti D, Guida M, Barezzani MG, Campovecchi C, Nassif N, Pinelli L. Favorable outcome of cochlear implant in VIIIth nerve deficiency. *Otol Neurotol* 2006; 27: 815-23. [\[CrossRef\]](#)