The Journal of International Advanced Otology

J Int Adv Otol 2020; 16(2): 271-3 • DOI: 10.5152/iao.2020.3766

Case Report

Successful Use of a Cochlear Implant in a Patient with Bony Cochlear Nerve Canal Atresia

Emel Tahir, Betül Çiçek Çınar, Hilal Burcu Özkan, Mehmet Yaralı, Bilgehan Böke, Levent Sennaroğlu

Department of Otolaryngology Head and Neck Surgery, Ondokuz Mayıs University, School of Medicine, Samsun, Turkey (ET)
Department of Audiology, Hacettepe University School of Health Sciences, Ankara, Turkey (BÇÇ, HBÖ, MY, BB)
Department of Otolaryngology Head and Neck Surgery, Hacettepe University School of Medicine, Ankara Turkey (LS)

ORCID iDs of the authors: E.T. 0000-0002-5219-0542; B.Ç.Ç. 0000-0002-6570-9876; H.B.Ö. 0000-0003-2681-3512; M.Y. 0000-0003-1870-0702; B.B. 0000-0002-5249-938X; L.S. 0000-0001-8429-2431.

Cite this article as: Tahir E, Çiçek Çınar B, Özkan HB, Yaralı M, Böke B, Sennaroğlu L. Successful Use of a Cochlear Implant in a Patient with Bony Cochlear Nerve Canal Atresia. J Int Adv Otol 2020; 16(2): 271-3.

The anatomical cause of congenital sensorineural hearing loss can be atresia of the bony cochlear nerve canal (BCNC). It has been reported that the cochlear nerve (CN) can be either hypoplastic or aplastic when the BCNC width is <1.5 mm radioanatomically. It is difficult to estimate the auditory–verbal abilities after cochlear implantation (CI) in patients with a hypoplastic CN. In such cases, it is also challenging to decide on the best treatment modality: CI or auditory brainstem implantation. In this case report, we present a 4-year-old male patient with BCNC atresia and the successful use of a cochlear implant; we also discussed the importance of audiological evaluation. A detailed radiological evaluation must be performed in every case following electrophysiological studies prior to CI. To accurately diagnose the pathology and select the surgical side, both computed tomography and magnetic resonance imaging scans should be used as complementary imaging methods in all CI candidates.

KEYWORDS: Cochlear nerve; computed tomography; cochlear implantation; audiometry; rehabilitation

INTRODUCTION

It is essential to perform medical, audiological, and radiological evaluations simultaneously for all candidates undergoing cochlear implantation (CI). Computed tomography (CT) and magnetic resonance imaging (MRI) of the temporal bone are two complementary modalities of preoperative imaging for CI [1]. CT and MRI scans provide data regarding whether CI will be functioning since a normal cochlear anatomy and functional cochlear nerve (CN) are associated with favorable CI outcomes.

The bony cochlear nerve canal (BCNC) is a bony space between the fundus of the internal auditory canal (IAC) and the base of the cochlear modiolus and it contains CN fibers [1, 2]. In patients with BCNC atresia and/or stenosis, a normal functioning CN is usually unexpected. BCNC atresia may indicate aplasia of the CN, and in such cases, CN aplasia is the cause of congenital sensorineural hearing loss (SNHL) [2, 3].

Auditory rehabilitation and language development in the post-CI period can be insufficient if CI is performed before determining the functional status of the CN. A satisfactory auditory–verbal outcome is a rare entity in patients with BCNC atresia; hence, we discuss the successful use of CI in a patient with BCNC atresia together with audiological and radiological findings.

CASE PRESENTATION

A 4-year-old male patient with bilateral profound SNHL underwent both CT and MRI. CT was evaluated in the axial plane, and images were obtained through 0.5 mm collimation and 0.5 mm thickness (Somatom Plus 4 Volume Zoom 4-channel multidetector CT scanner, Siemens, Erlangen, Germany) The BCNC was measured at the mid-modiolar level in the axial section.

This study was presented as poster at the 12th European Symposium on Pediatric Cochlear Implantation (ESPCI), June 18-21, 2015, Toulouse, France.

Corresponding Address: Emel Tahir E-mail: emeltahir@hotmail.com

Submitted: 19.03.2017 • **Revision Received:** 03.05.2019 • **Accepted:** 19.05.2019 • **Available Online Date**: 27.05.2020
Available online at www.advancedotology.org

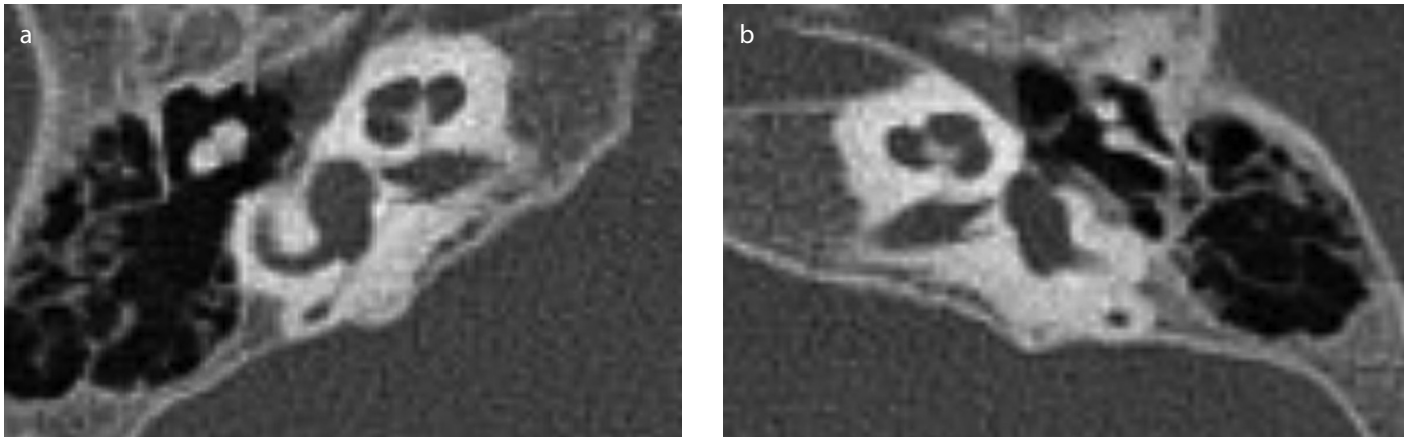

Figure 1. a, b. Axial CT images show a) right and b) left ears with normal cochlear morphology and BCNC atresia.

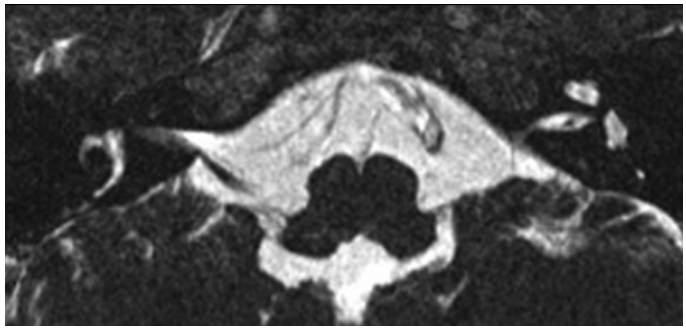

Figure 2. Axial T1-weighted MRI images show bilateral CN hypoplasia.

The number and dimension of the nerves present in the IAC were determined using the 3 Tesla MRI (Allegra, Siemens, Erlangen, Germany). The caliber of the CN was compared to the ipsilateral facial nerve, the superior and inferior vestibular nerves, and contralateral CN. The authors considered the CN to be hypoplastic if it was smaller than the other nerves mentioned above.

The patient had bilateral BCNC atresia based on CT findings (Figures 1a and 1b) and hypoplastic CN based on MRI findings (Figures 2a, 2b, and 3).

Preoperative subjective and objective audiological tests were performed. The auditory brainstem response test and otoacoustic emissions were in accordance with profound SNHL. However, in behavioral testing with inserted earphones, there were some auditory responses at 250, 500, 1000, and 2000 Hz (90, 110, 115, and 115 dB). In addition, there was some awareness to speech–noise at 85 dB bilaterally. The patient's auditory responses indicated that there might have been some functioning auditory nerve fibers. As such, the patient underwent uneventful right-sided CI. Post-CI hearing thresholds for 250-6000 Hz were 30-55 dB Hearing Level.

After using the cochlear implant for 4 years and 10 months, the auditory perception was evaluated using the Meaningful Auditory Integration Scale (MAIS), Categories of Auditory Performance (CAP)-II, Functioning After Pediatric Cochlear Implantation (FAPCI), and Parents' Evaluation of Aural Performance of Children (PEACH) when the patient was 8 years and 5 months. Before CI, the MAIS score was 3/40; this score was obtained for using the device, but the patient did not produce meaningful speech. Post-CI, his MAIS score was 32/40. Besides; his sentence recognition test (SRT) results were as follows; auditory–verbal score was 97%, and the only auditory score was 85%. His CAP-II score was 8, FAPCI score was 88.6%, and PEACH score was 78.3%. The patient has been enrolled in a mainstream primary school and has shown satisfactory academic performance. He has also developed reading comprehension skills and provided appropriate answers to various questions. Informed consent was obtained from his parents regarding this publication.

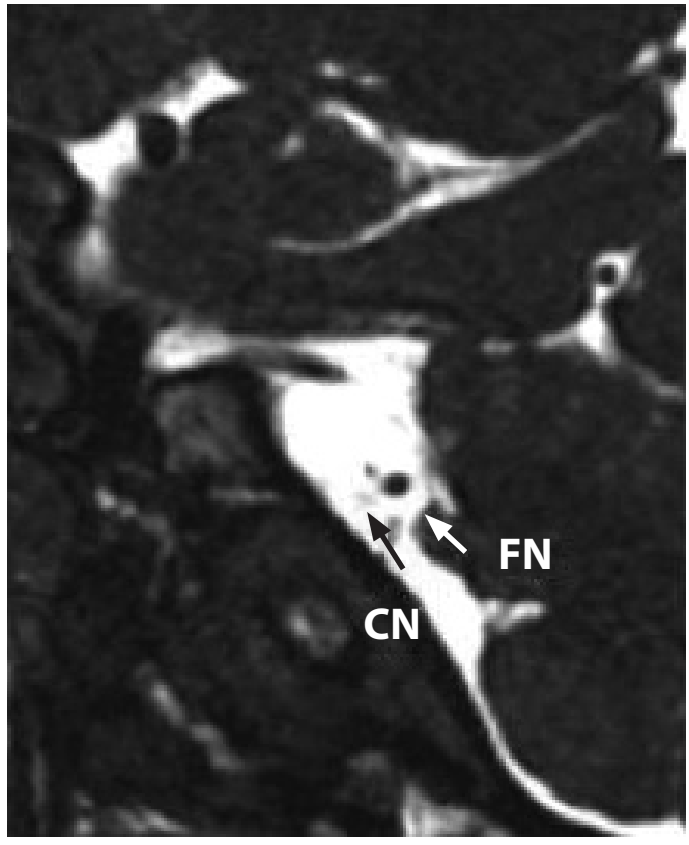

Figure 3. Oblique Sagittal T1-weighted MRI images demonstrates the poorly visualized hypoplastic CN in relation to the ipsilateral facial nerve.

DISCUSSION

Audiological and radiological examinations to determine the status of the CN should be included in the preoperative evaluation of CI candidates [4]. A CI surgeon should be able to interpret and combine

the radiological and audiological data. CT alone is not sufficient to predict the status of the CN; MRI should also be used [5]. BCNC stenosis (or atresia) is an anomaly that can lead to abnormal CN [6]. The main objective of this report was to highlight the importance of a behavioral test in a patient with BCNC atresia. Even in patients with an atretic BCNC, the CN (although hypoplastic) may function. In patients with CN hypoplasia, it is difficult to predict if there will be sufficient development of auditory–verbal skills in the post-CI period, as a detailed audiological evaluation can provide a clue whether the patient will benefit from CI or not.

It is notable that the preoperative ability to hear was evident, and outcomes were good. Therefore, the auditory fibers were adequate despite evident atretic BCNC and poorly visualized hypoplastic CN. When this patient was initially evaluated, CT and MRI findings were inconsistent with the audiological test results. The presence of a functioning nerve was confirmed through behavioral tests. Adunka et al. [7] described a similar case in which MRI and CT did not show a CN, although behavioral auditory responses were observed.

The mode of transfer of the auditory signal to the cochlear nucleus in the absence of a visible CN was unclear. One hypothesis was that the signal might have been transmitted through the nerve fibers that appear to be vestibular nerves [7]. Another hypothesis was that there were very thin but physiologically active CN fibers that were not visualized on MRI due to a narrow IAC [8]. Therefore, CI can be performed if there is a behaviorally observable auditory response and should be followed by a close observation of the auditory–verbal progress. Behavioral tests should be performed along with electrophysiological studies in such special cases. CT findings might indicate the absence of a CN because of an atretic BCNC; additional MRI does not always provide adequate information about the status of the CN. Complete audiological and radiological evaluations are both mandatory and complementary in all CI candidates.

CONCLUSION

Atretic BCNC does not merely indicate CN aplasia, as MRI does not always provide definitive information about the CN status. When CN deficiency is initially suspected based on CT findings, it must be confirmed through an MRI scan and audiological evaluation. Radiological data should always be interpreted in conjunction with electrophysiology and behavioral test results.

Informed Consent: Informed consent was obtained from the parent of the patient who participated in this study.

Peer-review: Externally peer-reviewed.

Author Contributions: Concept – L.S., E.T., B.Ç.Ç.; Design – B.Ç.Ç., E.T., H.B.Ö.; Supervision – L.S., B.B.; Resource - B.B., L.S.; Materials - L.S., B.Ç.Ç., H.B.Ö.; Data Collection and/or Processing - B.Ç.Ç., M.Y., L.S.; Analysis and/or Interpretation - L.S., M.Y., E.T.; Literature Search - E.T., Writing - M.Y., E.T., H.B.Ö., B.Ç.Ç.; Critical Reviews - L.S., B.B.

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

Financial Disclosure: The authors declared that this study has received no financial support.

REFERENCES

1. Yi JS, Lim HW, Kang BC, Park SY, Park HJ, Lee KS. Proportion of bony cochlear nerve canal anomalies in unilateral sensorineural hearing loss in children. Int J Pediatr Otorhinolaryngol 2013; 77: 530-3. [Crossref]
2. Miyasaka M, Nosaka S, Morimoto N, Taiji H, Masaki H. CT and MR imaging for pediatric cochlear implantation: emphasis on the relationship between the cochlear nerve canal and the cochlear nerve. Pediatr Radiol 2010; 40: 1509-16. [Crossref]
3. Jang JH, Kim JH, Yoo JC, Kim CH, Kim MS, Chang SO, et al. Implication of bony cochlear nerve canal on hearing in patients with congenital unilateral sensorineural hearing loss. Audiol Neurootol 2012; 17: 282-9. [Crossref]
4. Tamplen M, Schwalje A, Lustig L, Alemi AS, Miller ME. Utility of preoperative computed tomography and magnetic resonance imaging in adult and pediatric cochlear implant candidates. Laryngoscope 2016; 126: 1440-5. [Crossref]
5. Sweeney AD, Carlson ML, Rivas A, Bennet ML, Haynes DS, Wanna GB. The limitations of computed tomography in adult cochlear implant evaluation. Am J Otolaryngol 2014; 35: 396-9. [Crossref]
6. Purcell PL, Iwata AJ, Philis GS, Paladin AM, Sie KC, Horn DL. Bony cochlear nerve canal stenosis and speech discrimination in pediatric unilateral sensorineural hearing loss. Laryngoscope 2015; 125: 1961-6. [Crossref]
7. Adunka OF, Jewels V, Buchman CA. Value of computed tomography in the evaluation of children with cochlear nerve deficiency. Otol Neurotol 2007; 28: 597-604. [Crossref]
8. Warren FM, Hansberger HR, Shelton C. Apparent cochlear nerve aplasia. To implant or not to implant? Otol Neurotol 2010; 31: 1088-94. [Crossref]