



Original Article

# Radiological Imaging Findings of Patients with Congenital Totally Hearing Loss

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**OBJECTIVE:** The aim of this study was to determine and classify inner ear abnormalities in patients who had cochlear implants because of congenital sensorineural hearing loss using preoperative temporal bone computed tomography and magnetic resonance imaging.

**MATERIALS and METHODS:** Patients in the otolaryngology department who had cochlear implants because of congenital sensorineural hearing loss between January 2011 and December 2013 were included in the study. There were 167 male and 133 female patients, a total of 300. All of the patients were evaluated with 4-detector-row computed tomography and 1.5 Tesla magnetic resonance imaging.

**RESULTS:** Inner ear abnormalities were found in 136 of 600 ears (20.3%). There were six ears with incomplete partition-II (4.4%), five ears with incomplete partition-I (3.6%), two ears with Michel deformity (1.4%), two ears with cochlear hypoplasia (1.4%), two ears with cochlear otosclerosis (1.4%), and one ear with common cavity deformity (0.7%). Dilatation of the internal acoustic canal was found in 42 ears (30.9%); also, 21 ears with cochlear nerve aplasia/hypoplasia (15.4%), 5 ears with internal acoustic canal aplasia, and 1 ear with internal acoustic canal hypoplasia (0.73%) were detected. There were 10 ears with posterior semicircular canal (7.3%), 10 ears with lateral semicircular canal (7.4%), 8 ears with superior semicircular canal aplasia/hypoplasia (5.9%), and 8 ears with lateral semicircular canal-vestibular dysplasia. An enlarged vestibular aqueduct was found in 16 ears (11.7%). High jugular bulbs were found in 21 ears; however, this variation was not considered to be an inner ear abnormality.

**CONCLUSION:** Computed tomography and magnetic resonance imaging are essential for the evaluation, determination, and classification of inner ear abnormalities in patients with congenital sensorineural hearing loss who are candidates for cochlear implant operations. Also, these radiological instruments aid in determining contraindications and predicting intraoperative difficulties. Computed tomography and magnetic resonance imaging findings for these patients should be evaluated by an experienced radiologist before the operation.

**KEYWORDS:** Congenital sensorineural hearing loss, cochlear implant, inner ear abnormalities, computed tomography, magnetic resonance imaging

## INTRODUCTION

According to the degree of hearing loss, congenital sensorineural hearing loss (cSNHL) is observed in 1–3 of every 1,000 live births<sup>[1-2]</sup>. Although the inner ear structures of 80% of these patients are detected to be normal by temporal bone computed tomography (CT) and magnetic resonance, inner ear malformation can be detected in 20% of those patients<sup>[3]</sup>. Radiological examination is used to establish cases with contraindications as well as the pathologies encountered during surgery in patients with advanced hearing loss who are candidates for cochlear implants. Also, radiological examination has a role in the decision of which ear is preferable<sup>[4-6]</sup>.

Computed tomography and magnetic resonance imaging (MRI) are the two most important radiological examination techniques to evaluate patients prior to implantation. To display the bone labyrinth and internal auditory canal (IAC), the high resolution temporal bone CT technique is the first priority. Temporal bone CT provides detailed information about the condition of the otic capsule, cochlea, vestibule, oval and round window, and semicircular canals as well as the course of the facial nerve, IAC, vestibular aqueduct (VA), and cochlear aqueduct (CA)<sup>[7]</sup>.

To establish the presence of the nerves inside the IAC and to determine their diameters, a temporal bone MRI examination is performed<sup>[8]</sup>.

Our aim in this study was to use temporal bone CT and MRI to evaluate the inner ear formations and malformations of patients due to receive cochlear implants (CIs) because of cSNHL and to determine the contraindications for CI.

## MATERIALS and METHODS

The study was conducted with the authorization of Medical Ethics Committee No. 5 of the Çukurova University School of Medicine. In the Department of Ear, Nose and Throat between January 2011 and December 2013, temporal bone CT and MRI images of a total of

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**Submitted:** 23.07.2015

**Revision received:** 19.11.2015

**Accepted:** 19.11.2015

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300 patients, 167 male and 133 female, who were subsequently diagnosed with total SNHL were reevaluated retrospectively. The ages of the patients ranged from 1 to 30 years, and the average was 4.1 years.

Because only patients with cSNHL were accepted in our study, those having secondary hearing loss were not included. All of the images were evaluated preoperatively.

The parameters that we evaluated by imaging are presented in Table 1.

Computed tomography imaging was performed with a multislice device (Asteion TSX-021B Toshiba, Tokyo, Japan) with a collimation of 4 × 0.5, using 0.75 rotation time, 1 mm slice thickness, 0.750 pitch F, 100 effective mAs, and 120 kV parameters, in the supine position, angled as the orbitomeatal line to be parallel to the line between the inferior orbital edge and the upper edge of the external auditory canal (with an angle between 0 and 30 degrees). For the imaging, 400 WW/4000 WL values were generally used.

MRI was performed with a 1.5 Tesla Signa HDxt superconducting system (GE; USA). In this research, an 8HR brain coil was used. In the inner ear MRI, the axial FIESTA sequence (TE: minimum, NEX: 4.00, slice thickness: 1.0 and flip angle: 65 degrees) was used. In the brain MRI, the FLAIR sequence (TR: 9000, TE: 95.0, NEX: 1.00, slice thickness: 5.0, spacing: 1.5) was used.

**RESULTS**

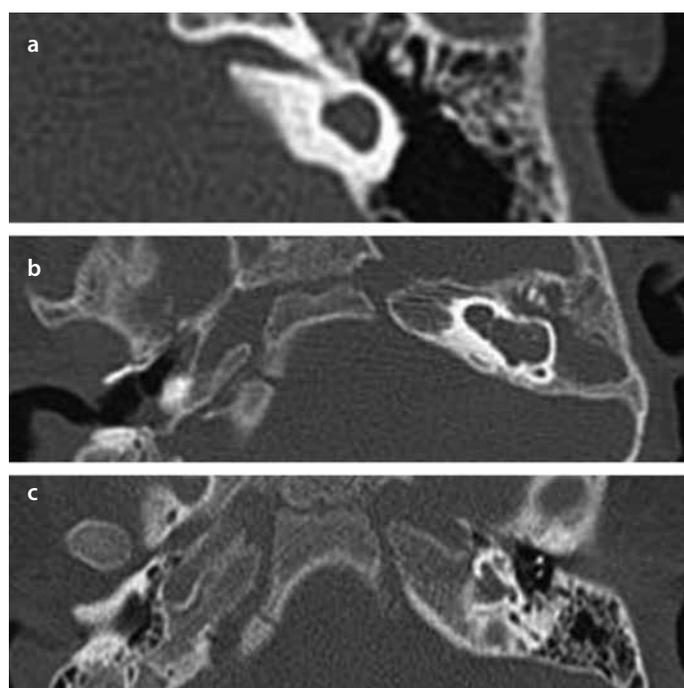
In our study, inner ear anomalies were detected in 136 of 600 ears (20.3%); 18 of these (13.2%) were cochlear anomalies. Findings compatible with Michel deformity were found in two ears (2%), and findings compatible with common cavity deformity were found in one ear (0.7%); findings compatible with IP-I were found in five (3.6%) ears, and findings compatible with IP-II (Mondini) were found in six ears (4.4%) (Figure 1). In a total of 21 ears, cochlear nerve aplasia/hypoplasia was present (Table 2). Although this anomaly is isolated in 13 of the 21 ears in which cochlear nerve aplasia/hypoplasia was found, an association with IP-II and IP-I was found in 2 cases with unilateral anomaly and in 1 case, respectively. Also, cochlear nerve aplasia/hypoplasia was found in two ears with Michel deformity, in one ear with common cavity deformity, and in two ears with IAC aplasia. Three of the isolated cases were in the right ear and two were in the left ear; in four patients, bilateral anomaly was present, and in one patient, the vestibular branch was also aplastic. In a case of IP-II, contraction was observed on the right IAC; inside the canal, a single nerve which was thought to be a facial nerve was observed. Also, in a case of IP-I, on the right eighth cranial nerve, hypoplasia and cochlear and vestibular branch aplasia were found; on the left side, vestibular branch aplasia was found (the cochlear nerve was normal).

**Table 1.** The parameters that we evaluated in temporal bone CT, inner ear MRI, and cerebral MRI in cases of congenital sensorineural hearing loss

Evaluated by CT		Evaluated by MRI	
Cochlea	Aplasia	Cochlea	Aplasia
	Hypoplasia		Hypoplasia
	Number of round		Number of round
	Cystic cavity		Cystic cavity
	Increase of sclerosis		
Vestibule	Dilate	Vestibule	Dilate
	Cystic cavity		Cystic cavity
	Aplasia		Aplasia
SSC	Aplasia/hypoplasia	SSC	Aplasia/hypoplasia
	Expand		Expand
IAC	Aplasia/hypoplasia	IAC	Aplasia/hypoplasia
	Expand		Expand
VA		Nerves within IAC	
CA		Endolymphatic bladder	
Mastoid Cells	Hypoplasia	Soft tissue and/or fluid signal in mastoid cells and in the middle ear cavity	
	Ventilation loss and/or absence	Cerebral changes	
	Soft tissue density	Cerebellar changes	
Soft tissue density in middle ear cavity			
Middle ear ossicles (malleus, stapes, and incus)			
Sigmoid sinus			
Jugular fossa			

MRI: magnetic resonance imaging; CT: computed tomography; SSC: semicircular canal; IAC: internal acoustic canal; VA: vestibular aquaduct; CA: cochlear aquaduct

A total of two ears with cochlear aplasia Michel deformity were found, and isolated cochlear hypoplasia (in addition to bilateral SSC dilatation) was present in two ears. A lack of the cochlear round window was found in six ears (Mondini cases), and cystic cavity-shaped cochlea was found in six ears (IP-1 in five ears and common cavity deformity in one ear). Sclerosis increase around the cochlea was found in two ears and evaluated as otosclerosis. When monitoring 16 ears with dilated vestibules [6 Mondini, 6 lateral semicircular canal vestibule dysplasia (LSVD)], isolated vestibular dilatation was found in two of them; also, in two cases, significant bilateral IAC dilatation accompanied vestibular dilatation on the left. Cystic cavity shaped vestibule was found in six ears (common cavity deformity in one ear and IP-I in five ears). Vestibular aplasia was found in two ears having Michel deformity. Wide VA (in 6 ears, accompanied by IP-II) was found in a total of 16 ears; VA agenesis was observed in 4 ears. In our study, aplasia/hypoplasia was observed in the LSSC in 10 ears, the PSSC in 10 ears, and the SSSC in 8 ears. Additionally, in a total of 42 ears, IAC dilatation was observed (cases with IP-I, a case with IP-II, and a case with vestibular dilatation). IAC aplasia was found in a total of five ears; unilateral IAC hypoplasia was observed in one case, and a single nerve inside the canal which was thought to be compatible with a facial nerve was present. When monitoring high positioned jugular



**Figure 1.** a-c. Inner ear malformation samples by computed tomography. Common cavity (a); incomplete partition type 1 (b); and incomplete partition type 2 (c)

**Table 2.** Inner ear malformations table

Inner ear malformation	No	(%)
Michel deformity	2	1.4
IP-I	5	3.6
IP-II	6	4.4
Common cavity deformities	1	0.7
Cochlear nerve aplasia/hypoplasia	21	15.4

IP-I: incomplete partition type I; IP-II: incomplete partition type II

bulb variation in a total of 21 ears, the condition was observed in the right ear in 13 cases, in the left ear in 2 cases, and in both the ears in three cases. When monitoring SSC aplasia/hypoplasia in 28 ears included in the study, isolated SSC aplasia/hypoplasia was present in 5 of the ears. LSSC hypoplasia was found in two of the isolated cases and PSSC hypoplasia in two others; and as with these two cases, aplasia was found in all SSCs. SSC aplasia/hypoplasia was found in one ear with IP-I, in two ears with Michel deformity, and in two ears with LSVD. Also, one ear was observed to have common cavity deformity, and the sides of the SSCs were not shaped in 12 ears; SSC dehiscence was observed in a total of 19 ears. LSSC and vestibular dilatation (LSVD) was found in a total of eight ears; in one case, isolated vestibule dilatation was present in both ears. Hypoplasia of the mastoid cells was observed in a total of nine ears, external ear canal stenosis in one ear, and anterior positioned sigmoid sinus in two ears. In two ears, dysplastic middle ear ossicles were seen. In 44 of the 300 patients in our study, cerebral and/or cerebellar pathology was monitored (7.3%) (Table 3, Figure 2-3).

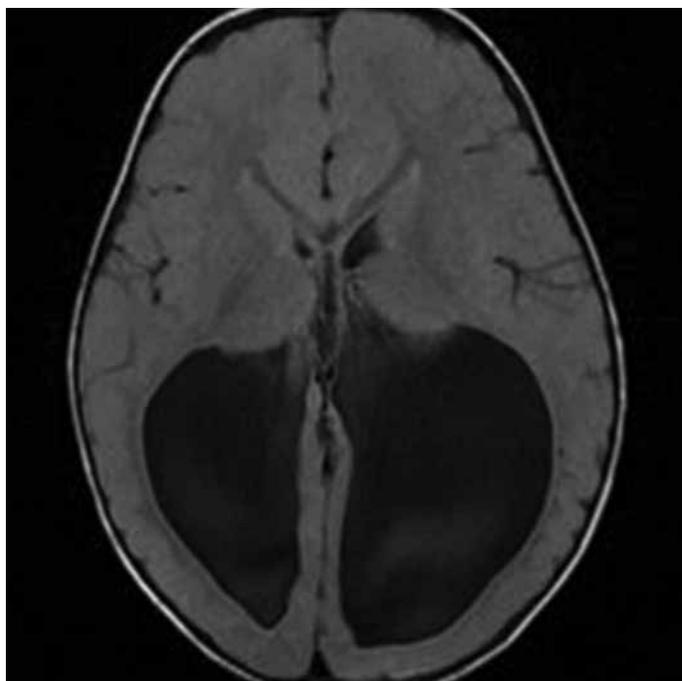
## DISCUSSION

According to the degree of hearing loss, cSNHL is observed in 1–3 of every 1,000 live births<sup>[1,2]</sup>. In infants having one or more risk factors, the incidence of evident hearing loss is more than 2–5 times greater than in those without the risk factor<sup>[3]</sup>. In many children with SNHL, their inner ear structures are detected as normal radiologically because most of the malformations are limited to the membranous labyrinth. Inner ear malformations can be detected at a rate of 20% with temporal bone CT and MRI images obtained using modern technology. The detection of these malformations is important because in some of these patients, there is a risk of recurrent meningitis and head trauma due to anomalies or sudden hearing loss due to sudden pressure changes and CSF leakage<sup>[3]</sup>.

In a CT study conducted by Antonella et al.<sup>[9]</sup> in 1999, anomalies were detected at a rate of 31% in 157 children with SNHL. In another study conducted in 2002 by McClay et al.<sup>[10]</sup>, although they detected anomalies at a rate of 17% in 185 children with SNHL, they were unable to detect any anomalies in the ears of 309 children without SNHL. In a study conducted in 2009 by Sennaroğlu et al.<sup>[23]</sup>, in which 46 inner ear malformations were monitored and in which assessments were conducted for the purpose of reclassification, they detected IP-I

**Table 3.** Cerebral and/or cerebellar changes

Cerebral and/or cerebellar changes	No	(%)
Hypoplasia of corpus callosum and colpocephaly	4	9.09
Agenesis of corpus callosum and colpocephaly	4	9.09
Band heterotopia	2	4.54
Hypoplasia of cerebellar vermis and/or hemisphere	6	13.63
Cerebral or cerebellar arachnoid cyst	6	13.63
Increase of periventricular intensity	12	27.27
Delay in myelination	2	4.54
Incomplete lissencephaly	2	4.54
Variation of cavum septum pellucidum and vergae	4	9.09
Pachygyria/polymicrogyria	1	2.27
Choroid plexus cyst	1	2.27

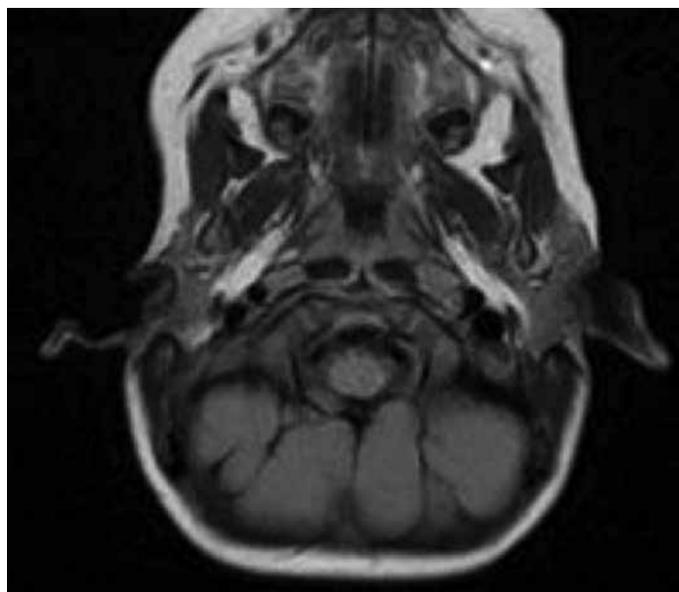


**Figure 2.** Corpus callosum agenesis is seen on FLAIR scans by magnetic resonance imaging

(17.3%) in 8 of the 46 ears in which anomalies were found and IP-II in 10 of them (21.7%). CA dilatation and any other anomalies were not monitored in any of the cases included in the study<sup>[11]</sup>. In a study conducted with 65 SNHL diagnosed patients in 2012 by Huo and Wong, they detected inner malformation in 33 ears in 20 patients. They detected cochlear malformations in 10 ears, Michel deformity in 1 ear (3%), common cavity deformity in 2 ears (6.1%), cochlear aplasia in 1 ear (3%), cochlear hypoplasia in 1 ear (3%), IP-I in 2 ears (6.1%), and IP-II in 3 ears (9.1%). In a study conducted by Kontorinski et al.<sup>[12]</sup> in 2013, in 23 patients who were found to have cochlear aplasia by inner ear CT and MRI (28 ears), they determined common cavity deformity in 6 ears (21.4%), IP-I in 4 ears (14.2%), and IAC atresia in 1 ear (3.5%).

In our study, although inner ear anomalies were detected in 136 of 600 ears (20.26%; in some cases, more than one anomaly is present), the findings were compatible with literature values. In 18 ears of the total 136 in which anomalies were detected (13.2%), cochlear anomalies were observed; 2 of these were evaluated as Michel deformity (1.4%), 1 as common cavity deformity (0.7%), 5 as IP-I (3.6%), 6 as IP-II (4.4%), and 2 as cochlear hypoplasia. In 1971, Nance et al.<sup>[13, 14]</sup> first identified X-linked deafness in association with stapes base flick and perilymphatic gusher. In a study which was conducted by Sennaroğlu et al.<sup>[15]</sup> in 2006 on 20 patients in which inner ear anomalies were detected, they determined X-linked type deafness in 1 ear (5%). In our study, this anomaly was not observed in any of the cases.

One of the most common inner ear malformations is a wide vestibule with wide, cystic, and short LSSCs. This can be easily monitored with single fluid-filled cavity-shaped CT and MRI images. This pathology is called LSVD. This anomaly may be bilateral and is generally accompanied by inner ear anomalies<sup>[16]</sup>. In a study conducted by Sennaroğlu et al.<sup>[15]</sup> in 2006, LSVD was detected in 9 of 20 ears (45%); in a study conducted by Ma et al.<sup>[17]</sup> in 2008, it was detected in 14 of 36 ears (38.8%); and Westerhof et al.<sup>[18]</sup> found LSVD in 9 of 42 ears (21.4%). In our study,



**Figure 3.** Bilateral cerebral clefts are seen on FLAIR scans by magnetic resonance imaging

out of 136 inner ear anomalies, 8 of them (5.9%) were LSVD. When monitoring two cases of bilateral VA agenesis, in one of the cases, all SSCs had a hypoplastic/dysplastic appearance in both ears. In the other patient, an additional anomaly was not observed. The number of cases with LSVD was found to be lower than other studies. The other anomalies of the semicircular canals were short, wide, or narrow canals and the partial or complete absence of the canals<sup>[16]</sup>. In a study conducted by Huo and Wang<sup>[19]</sup> in 2012, they found SSC malformation in 5 of 33 ears in which inner ear malformation was detected; in a study conducted by Pappas et al.<sup>[20]</sup> in 1990, they found a LSSC hypoplasia/dysplasia rate of 51.4%, and they emphasized that this is the most commonly detected anomaly in cases of congenital SNHL. In our study, findings compatible with SSSC were observed in 8 ears (5.88%), PSSC in 10 ears (7.35%), and LSSC aplasia/hypoplasia in 10 ears (7.35%).

Wide VA is the most common inner ear malformation that can be detected radiologically. The regular VA diameter is between 0.4 and 1 mm. To diagnose VA expansion, its diameter must exceed 2 mm. In many cases, cochlear and SSC malformations accompany VA expansion. These patients are prone to the development of sudden hearing loss, particularly after head traumas, as with other inner ear malformations<sup>[3, 21]</sup>. Even if these patients have CSF leakage risk during the operation, this does not create an absolute contraindication for CI<sup>[3, 21]</sup>. In a study conducted by Sennaroğlu et al., wide VA was observed in 12 ears out of 54 (22.2%), and in another study, they observed it in 16 ears out of 46 (34%). In a study conducted by Sennaroğlu et al.<sup>[22]</sup>, wide vestibular aqueduct was observed in 16 ears of 36 (44.4%); 6 of these ears were found to have other inner ear malformations. Also, Sennaroğlu et al.<sup>[23]</sup> found wide vestibular aqueduct in 9 ears out of 42 (21%) with SNHL. In our study, wide vestibular aqueduct was found in 16 ears out of 136 (11.7%). Mondini malformation was present in six patients with wide vestibular aqueduct.

The length of the internal auditory canal may vary from 3 to 18 mm (approximately 12 mm); its diameter may vary from 2 to 8 mm<sup>[24]</sup>. If the diameter of the canal is below 2 mm, stenosis can be mentioned. IAC stenosis accompanies congenital malformations at a rate of 12%.

When observing the facial nerves of patients with IAC stenosis, the vestibulocochlear nerve is frequently found to be largely hypoplastic or aplasic [25, 26].

The determination of IAC anomalies and the presence of vestibulocochlear nerves are fairly important in determining CI candidacy [5, 26]. The absence of the cochlear nerve is an absolute contraindication of the implantation. Because their embryological development origins are different, a normal cochlea does not indicate that the cochlear nerve is normal. An IAC narrower than 2.5 mm suggests cochlear nerve aplasia [27].

In a study conducted by Kang et al. [28] in 2010, they stated that the detection of cochlear nerve defects by MRI can be an indicator of poor results after implant.

In a study conducted by Zhang et al. [29] in 2012, which was performed in nine children patients with SNHL and cochlear nerve defects with the purpose of investigating whether CI was beneficial, they demonstrated that adequate speech intelligibility or sensing capability was not obtained in any of the children, at least in 1-year follow-ups after the surgery. Consequently, they showed that because of the limited effectiveness of cochlear implants and the uncertainty of the cost-benefit ratio, it must be considered in patients with cochlear nerve deficiencies [29].

In a study conducted by Yan et al. [30], they found that IAC anomalies can occur together with cochlear nerve anomalies. In a study conducted by Shelton et al. [31] in 1989, they reported that cochlear nerve absence is possible in patients with IAC stenosis (with a diameter of 1–2 mm). In a study conducted in 2002 by Sennaroğlu et al. [23], in 54 ears with SNHL, they did not detect the cochlear nerve in a total of 7 ears (12.9%). In a study conducted by Clemmens et al. [32] in 2013, they detected cochlear nerve anomalies in 26% of 128 patients with unilateral congenital SNHL. This rate was found to be significantly higher in patients with profound hearing loss. In our study, when monitoring cochlear nerve aplasia/hypoplasia in a total of 21 ears (15.4%) out of 136, isolated was observed in 13 of them.

The jugular foramen continues above the promontorium level in the high positioned jugular bulb variation. This variation may obstruct the access to the round window during the operation [33]. In a study conducted by Tomura et al. [34] in 1995, which was performed in 325 patients with the purpose of evaluating temporal bone variations, they found the high jugular bulb rate to be 2.4%. In our study, we did not evaluate high positioned jugular bulb variation as an ear anomaly; we detected it in 21 (3.5%) of 600 ears.

In a study conducted by Lapointe et al. [35] in 2006, they detected cerebral anomalies in 8 (20%) of 40 patients with SNHL. They also reported that some of these anomalies may negatively affect the success of the cochlear implant; hence, cerebral MRI should be added to temporal bone MRI examination in patients who are candidates for cochlear implants. In our study, cerebral and/or cerebellar pathology was observed in 44 patients (7.3%).

In conclusion, imaging methods are useful methods, both in terms of the determination of the underlying pathology and in determining

the difficulties that may be encountered in the rehabilitation phase, in cases of congenital hearing loss. This is useful to predict the success of operations and the potential complications of intraoperative and postoperative periods, particularly in patients in which cochlear implants are applied. Currently, the use of navigation devices for ears with anomalies in otologic surgeries may lead to a fair reduction in complication rates.

**Ethics Committee Approval:** Ethics committee approval was received for this study from the ethics committee of Çukurova University.

**Informed Consent:** Written informed consent was obtained from patients who participated in this study.

**Peer-review:** Externally peer-reviewed.

**Author Contributions:** Concept - N.D., M.D., E.A.; Design - Ö.S., T.B., Ü.T.; Supervision - E.A., Ü.T., F.Ç.; Resources - N.D., M.D.; Materials - N.D., M.D., Ö.S.; Data Collection and/or Processing - N.D., M.D., Ö.S.; Analysis and/or Interpretation - N.D., Ü.T., E.A.; Literature Search - N.D., Ö.S., M.D., T.B.; Writing Manuscript - M.D., Ö.S.; Critical Review - Ü.T., F.Ç., E.A., Ö.S.

**Conflict of Interest:** No conflict of interest was declared by the authors.

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

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