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De novo sensorineural hearing loss sequelae of narrow, duplicated internal auditory canal: Case series and literature review

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ABSTRACT

Introduction: A narrow duplicated internal auditory canal (IAC) is an extremely rare anomaly, likely associated with congenital sensorineural hearing loss due to aplasia/hypoplasia of the vestibulocochlear nerve or the cochlear branch alone. We aimed to review our experience with IAC duplication, describe its clinical characteristics, and present a literature review.

Case presentation: Our Otology database was searched for children who showed duplication of the IAC. Clinical characteristics of two children with bilateral duplication of the IAC are described. Data regarding clinical history, auditory assessment, magnetic resonance imaging (MRI), and computed tomography (CT) were collected and analyzed. The separated, accessory bony canals were demonstrated on high-resolution CT scans, and the nerves were demonstrated on MRI.

Discussion: To date, a few cases of narrow duplicate IAC have been reported in the literature, Approximately 20% of patients with congenital SNHL are found to show inner-ear bony abnormalities on CT, but much uncertainty still exists about the mechanism underlying IAC stenosis.[5] Imaging findings of the temporal bone in our case series demonstrated asymmetrical narrowing of both IACs, there is no clear evidence in the literature supporting the predominance of one side over the other. In our series, facial nerve function was intact bilaterally. As for our cases, both patients were enrolled in a single-sided deafness evaluation for a trial of options such as BAHA, CROS, cochlear implants, and other non-implantable hearing aids. Furthermore, addressing the important factors will optimize the outcomes including surgery at early age to optimize neural plasticity, with intense long-term therapy.

Conclusion: Congenital duplication of the IAC likely convoying sensorineural hearing loss due to aplasia/hypoplasia of the vestibulocochlear nerve. Early diagnosis and intervention are essential to optimize patient outcomes.

1. Introduction and importance

Internal auditory canal (IAC) duplication is an extremely rare congenital anomaly of the temporal bon, usually seen in association with a stenotic IAC. An estimated 0.019% rate of occurrence and only 46 cases reported in the literature. The diameter of a normal IAC is 4 mm (range, 2–8 mm). A narrow IAC is defined by a diameter of less than 2 mm, as seen on high-resolution computed tomography (CT) of the temporal bone [1–3]. It is usually unilateral and associated with cochlear nerve dysplasia and profound hearing loss [4]. Additionally, duplicated IAC is often associated with other systemic developmental anomalies such as malformations of the heart, kidneys, skeletal system, and intestinal tract [5]. The associated presence of sensorineural hearing loss (SNHL) on the affected side has been universally reported [6].

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Moreover, 20% of patients with SNHL showed inner-ear abnormalities on CT [7]. Of these, only 12% were reported to have IAC stenosis [8]. The stenotic IAC and SNHL are believed to be the result of Aplasia or hypoplasia of the vestibulocochlear nerve or its cochlear branch [9]. Most IAC stenosis cases show preserved functioning of the ipsilateral facial nerve [10,11]. Furthermore, Casselman et al. [12] and Maxwell et al. [13] showed moderate speech perception test results of IAC anomalies after cochlear implantation. Nevertheless, only a few cases of unilateral narrow duplicate IAC have been reported [14,15]. The work has been reported in line with the SCARE criteria [16]. Herein, we report two cases of a congenital sensorineural hearing loss sequelae of narrow unilateral duplicated IACs with a focused comprehensive literature review including presentation, clinical finding, status of vestibulocochlear nerve, facial nerve, associated anomalies and management options.

2. Methodology

Descriptive case series study for two children who showed duplication of the IAC. Data for this study were collected prospectively in academic university hospital. Patients were followed for an average of 12 months. Written informed consent was obtained from the patient for publication of this case report and accompanying images. Research registry Identifying number is: researchregistry7806, https://www.researchregistry.com/browse-the-registry#home/. The work has been reported in line with the SCARE criteria.

Clinical characteristics of two children with bilateral duplication of the IAC are described on the basis of the clinical evaluation and radiographic finding. Data regarding clinical history, auditory assessment, magnetic resonance imaging (MRI), and computed tomography (CT) were collected and analyzed. The separated, accessory bony canals were demonstrated on high-resolution CT scans, and the nerves were demonstrated on MRI.

3. Case presentations

3.1. Case 1

A 6-year-old boy presented to the ENT outpatient clinic with bilateral hearing loss and speech delay. The patient had undergone intensive care unit admission for 20 days after delivery due to hyperbilirubinemia, evaluation were unremarkable regarding Drug history, family history including any relevant genetic information, and psychosocial history, he was otherwise healthy. Physical examination revealed normal bilateral auricles, periauricular areas, external canals, tympanic membranes, and an intact facial nerve. A complete audiological evaluation was performed. Auditory brainstem response showed bilateral sensorineural hearing loss with absent wave 5 up to 95 dB in both ears. The tympanogram showed type A findings. High-resolution CT scan of temporal bone and MRI of the IAC show left sided narrowed, duplicated IAC (Figs. 1 and 2).

3.2. Case 2

A 10-year-old girl presented with unilateral hearing loss on the right side since childhood. The patient showed no complaints of any vestibular symptoms and no speech delay. She had no history of mumps, head trauma, or sudden exposure to loud sounds. No family history of hearing loss was observed. Physical examination revealed normal auricles, periauricular areas, and external auditory canals on both sides. Type A tympanogram was noted on both sides. The middle ear pressure was normal on both sides, with normal head and neck status. The facial nerve function was normal on both sides. A complete audiological evaluation was performed by the pediatric audiology department, and the auditory brainstem response showed sensorineural hearing loss with absent wave 5 up to 95 dB. The audiogram showing the results obtained for the dead right ear is shown in (Fig. 3). High-resolution CT scan of temporal bone and MRI of the IAC show right sided narrowed, duplicated IAC. The facial nerve canal is intact along its course (Fig. 4). High-resolution heavily weighted T2WI of the internal auditory canal showed that the right vestibular cochlear nerve cannot be appreciated (Fig. 5).

4. Clinical discussion

Congenital IAC anomalies account for 12% of all congenital temporal bone anomalies [17]. To date, a few cases of narrow duplicate IAC have been reported in the literature, which are summarized in (Table 1). Researchers agree that there might be an association between IAC anomalies such as atresia, stenosis, Aplasia, and hypoplasia, and congenital SNHL. Approximately 20% of patients with congenital SNHL are found to show inner-ear bony abnormalities on CT, but much uncertainty still exists about the mechanism underlying IAC stenosis [5]. Two widely accepted hypotheses explain the association of a narrow IAC

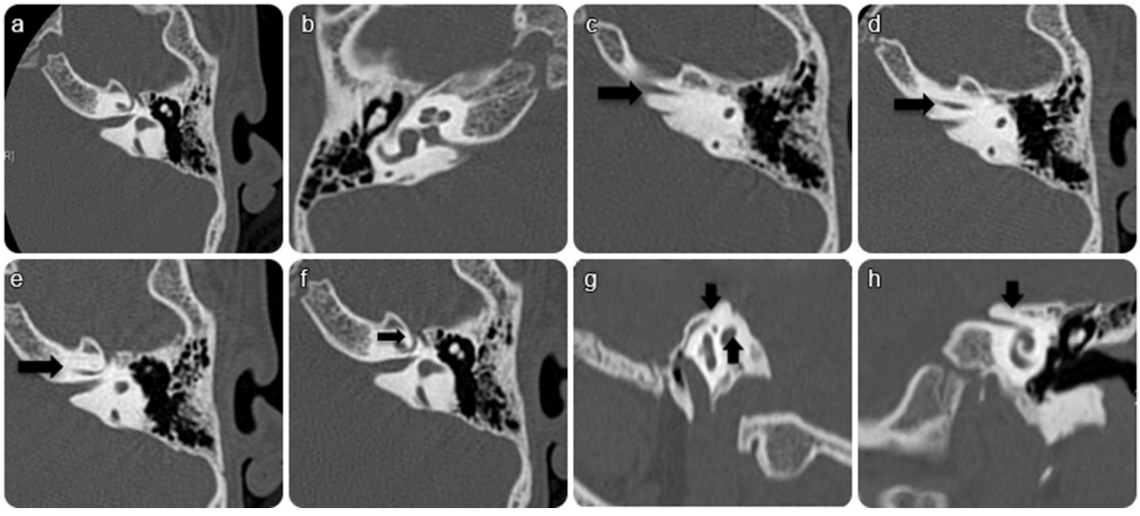

Fig. 1. A, B; High-resolution axial CT scan images of the temporal bone demonstrate asymmetrical narrowing of both internal auditory canals, the right more than the left
C, D, E, F: High-resolution axial CT images of the left temporal bone. Note the separate facial nerve canal (arrows).
G: High-resolution oblique parasagittal view of the Lt temporal bone.
H: The coronal section demonstrates the antero-superior position of the facial canal (arrow) in relation to the internal auditory canal.

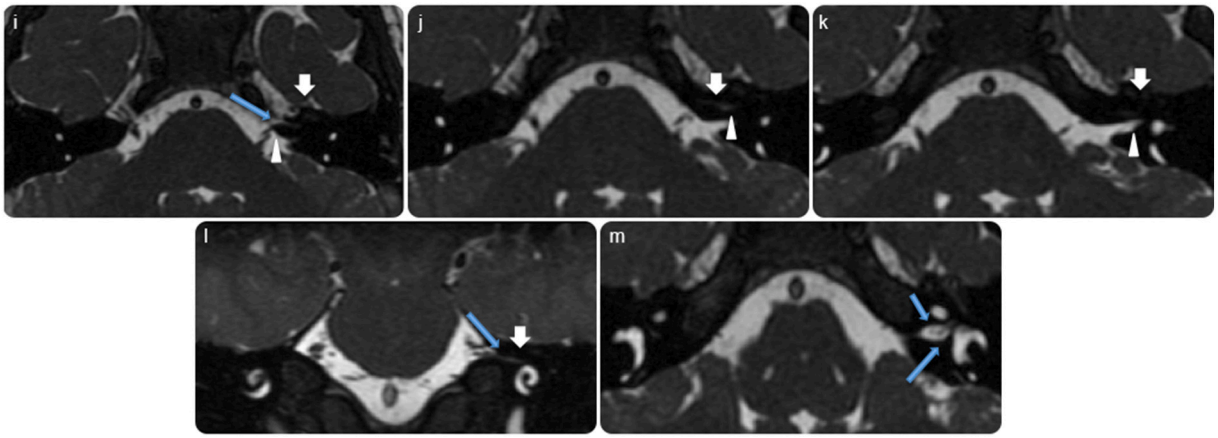

Fig. 2. High-resolution heavily weighted T2WI (3D FIESTA) of the left internal auditory meatus. The axial (I, J, K) and coronal (L) scans demonstrate the separate canal (white arrows) seen antero-superior to the internal auditory meatus (white arrowhead), with the cisternal component of the left facial nerve entering into its separate canal (small blue arrows). (M) High-resolution heavily weighted T2WI of the left internal auditory canal. Two cranial nerves are demonstrated within the fundus of the left internal auditory meatus (arrows). (For interpretation of the references to colour in this figure legend, the reader is referred to the web version of this article.)

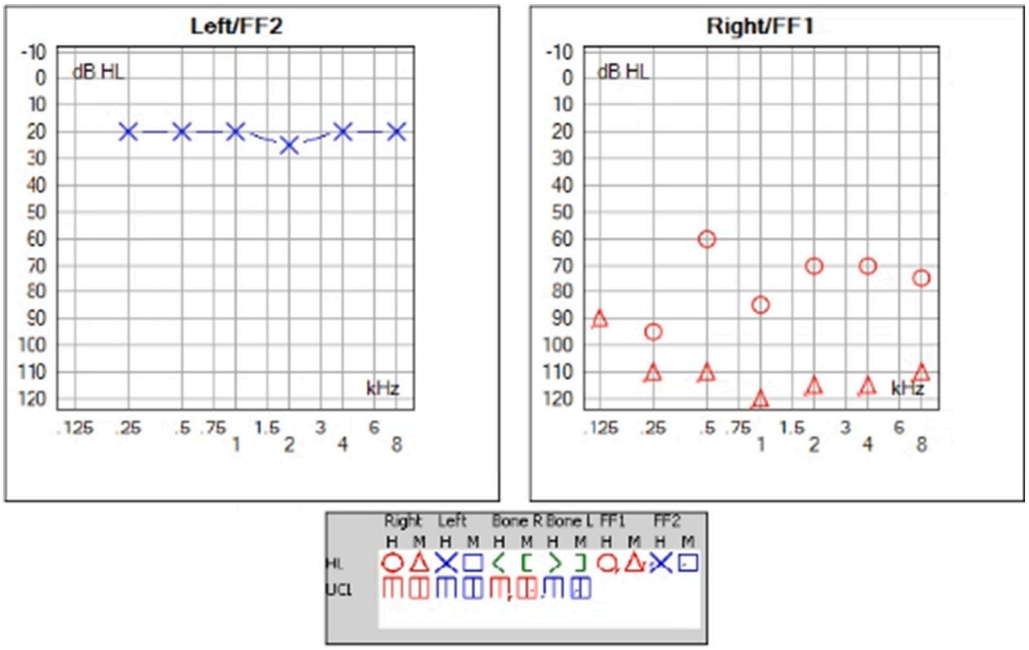

Fig. 3. Pure tone audiometry showed profound SNHL in the right ear.

with sensorineural hearing loss. The first and more widely accepted hypothesis states that the embryonic cochlea induces the growth of the vestibulocochlear nerve and that the bony canal develops around the eighth as well as the seventh cranial nerves by mesoderm chondrification and ossification in the eighth gestational week. If the vestibulocochlear nerve (VCN) is hypoplastic or aplastic, IAC does not develop properly and becomes stenotic. The other hypothesis is that the primary defect is bony stenosis that inhibits the growth of the vestibulocochlear nerve and causes impaired transmission of an induction signal from the intact cochlea and vestibule. However, this is less likely because most patients with IAC stenosis have preserved facial nerve function [6]. Imaging findings of the temporal bone in our case series demonstrated asymmetrical narrowing of both IACs, the first case on the left and the second case on the right; however, there is no clear evidence in the literature supporting the predominance of one side over the other. In our series, facial nerve function was intact bilaterally. It is well known that the facial nerve develops separately and is later surrounded as the canal forms around the vestibulocochlear nerve [18]. Most cases of IAC stenosis show preserved normal function of the ipsilateral facial nerve (Table 1). To our knowledge, there is only one report of facial palsy associated with duplicated IAC stenosis [6]. In our case, narrowing of the IAC was noticed in the CT and separate facial canal identified in the IAC. On the other hand, facial nerve function and caliber (on MRI) were normal in almost all reported cases [8]. MRI was used to identify the nerves in both canals of the duplicated IAC. Assessment and evaluation of the IAC and the cochlear nerve radiologically via high-resolution axial reformatted coronal, sagittal 3D reconstructed CT, and MRI of the vestibulocochlear nerves are crucial for developing an appropriate treatment plan [18,19]. Historically, a narrow IAC was considered a contraindication to cochlear implantation, with several reports of cochlear implant failure in the presence of such stenosis [20]. For Instance, Bakar et al. suggested that a narrow bilateral IAC in patients

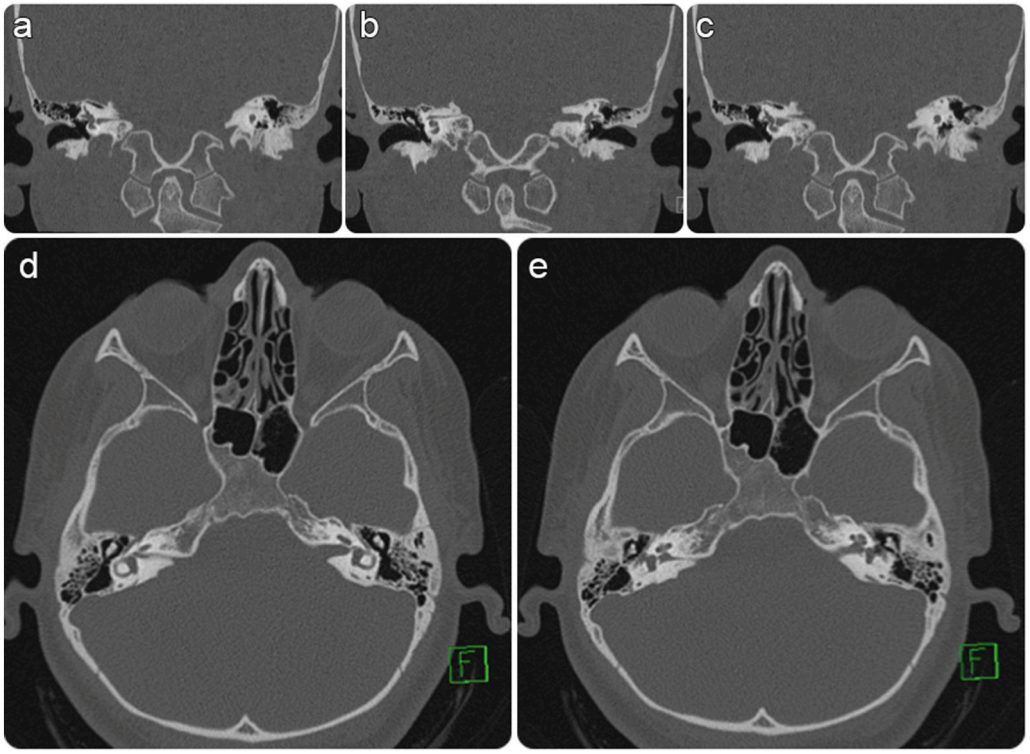

Fig. 4. Computed tomography images demonstrating a narrow duplicated IAC of the right temporal bone. The IAC is divided by a bony septum into the superior and inferior portions. The superior portion ends in a wide connection to the facial canal and a narrow connection to the vestibule. The posterior portion ends in a narrow connection to the cochlea and vestibule.

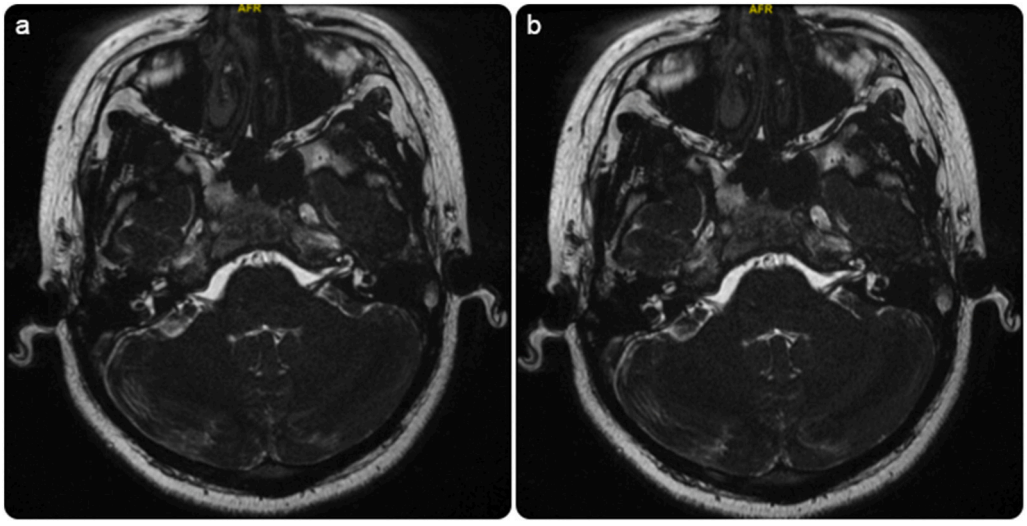

Fig. 5. High-resolution heavily weighted T2WI of the internal auditory canal showed that the right vestibular cochlear nerve cannot be appreciated.

with SNHL should be considered a contraindication for cochlear implants because of the absence or hypoplasia of the VCN [21]. However, the evidence for this relationship is inconclusive as patients who show a stenotic IAC on high-resolution HRCT do not always lack a VCN, and vice versa is also true in patients in whom IAC is normally sized on HRCT but does not demonstrate a normal VCN [7]. A few cases of improved hearing after cochlear implantation have been described in patients with hypoplasia of the cochlear branch [4,5]. Therefore, aggressive auditory testing, including cochlear promontory stimulation, along with an HRCT, MRI using 3D FT-CISS, and complete neurotological tests, are necessary to guide cochlear implant decisions [9,10]. As for our cases, both patients were enrolled in a single-sided deafness evaluation for a trial of options such as BAHA, CROS, cochlear implants, and other non-implantable hearing aids. Mary Thompson et al. highlighted that surgery at an early age to optimize neural plasticity, and intense, long-term therapy is important factors that will optimize outcomes [4]. As the two cases from a different hospital, lack of follow up periodically is present. As a result, a conflict in making multiple management plans is present.

5. Conclusions

Duplication of the IAC is a rare entity. It is usually associated with sensorineural hearing loss and other inner-ear abnormalities with various systemic developmental anomalies. The work-up for IAC duplication should include neuro-diagnostic testing and high-resolution radiography to detect abnormalities of the vestibulocochlear nerve within the IAC and decide for appropriate management. Early diagnosis and intervention options such as BAHA, CROS, cochlear implants, or other non-implantable hearing aids are essential factors that will optimize outcomes. Future studies are needed to elucidate these outcomes.

Patient consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

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CRediT authorship contribution statement

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This work was carried out in collaboration between all authors. All authors read and approved the final manuscrip.

Declaration of competing interest

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None.

Appendix A

Table 1

Reference	IAC duplication	ABR	Clinical findings		Nerve		Associated anomalies	Management
			FN	CN	FN (MRI)	VCN (MRI)		
Our case 1	L	No response	Norm	Bil SNHL	+	+	-NS	
Our case 2	R	NS	Norm	R SNHL	+	NS	NS	CI candidat HA
Manchanda et al., 2019	L	NS	Norm	Bil SNHL	NS	NS	Incomplete partition type I deformity	
Thompson et al., 2019	Bil	No response	NS	P-SNHL	+	Present in one side only	The lateral semi-circular canals on both sides were observed to be dysplastic	Bil CI
Bhattacharyya et al., 2018	Bil	No response	Norm	P-SNHL R > L	+	−	Bilateral lateral SSC aplasia	CI L
Y. Takanashi et al., 2017	L	No response	Norm	L T-SNHL	+	−	Lt vestibule, SCCs & cochlear hypoplasia.	
Y. Takanashi et al., 2017	Bil	No response	NS	P-SNHL	+	−	Rt cochlear hypoplasia	HA
Binnetoğlu et al., 2016	Bil	NS	Norm	Bil P-SNHL	+	R-hypoplastic L-absent	NS	CI R
Kishimoto et al., 2015	Bil	NS	NS	P-SNHL	+	+	Bilateral inner-ear malformation	CI
Vincenti et al., 2014	R	NS	NS	P-SNHL	+	Hypoplastic	NS	

(*continued on next page*)

Table 1 (*continued*)

Reference	IAC duplication	ABR	Clinical findings		Nerve		Associated anomalies	Management
			FN	CN	FN (MRI)	VCN (MRI)		
Kew et al., 2012	R	NS	Rt G-III Palsy	P-SNHL	–	–	NS	
Desai et al. Case 3, 2011	Bil	NS	NS	NS	+	–	NS	CI
Kessel et al., 2010	R	No response	NS	P-SNHL	+	–	NS	
L. Coelho et al., 2010	Bil	No response	NS	Bil S-SNHL	+	–	Left cochlear and vestibular dysplasia, bilateral superior SCC malformation and absence of the posterior SCC	
Kono et al., 2009	L	NS	Norm	L SNHL	+	Hypoplastic	NS	
Lee et al. (Case 1), 2009	R	NS	Norm	R S-SNHL	NS	NS	NS	
Baik HW et al., 2008	R	No response	Norm	R T-SNHL	+	–	NS	
Goktas et al., 2008	Bil	NS	NS	Bil P-SNHL	NS	NS	Mondini dysplasia, cystic dilatation of the vestibule, hypoplasia of lateral SCC, and enlarged vestibular aqueduct	
Weon et al., 2007	Bil	No response	Norm	Bil SNHL	+	_	-Right microtia, stenotic Rt EAC	
Demir et al., 2005	R	NS	Norm	Bil CHL	+	–	Klippel-Feil syndrome.	HA
Ferreira et al., 2003	R	NS	Norm	R P-SNHL	+	–	Bilateral microtia stenotic IAC	
Cho et al., 2000	R	NS	Norm	R T-SNHL	+	–	Bilateral cochlear dysplasia R > L.	
Vilain et al., 1999	R	No response	Norm	R T-SNHL	+	+	NS	
Casselman et al., 1997	L	NS	Norm	Bil SNHL	+	–	NS	
Weissman et al., 1991	L	No response	NS	NS	NS	NS	Bilateral EAC aplasia	

Abbreviations: ABR: auditory brainstem response; FN: facial nerve, CN: cochlear nerve, VCN: vestibulocochlear nerve R: right, L: left, Bil: bilateral, NS: Not specific, Norm: normal, SNHL: Sensorineural hearing loss, P-SNHL: profound, T-SNHL: total, CI: cochlear implantation, HA: hearing aids.

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