Bilateral Narrow Internal Auditory Canal With Duplication: High Resolution CT And MRI Findings

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Abstract: Inner ear malformations account for only 20 % of cases of congenital sensorineural hearing loss. A narrow internal auditory canal (IAC) with duplication is a very rare congenital anomaly that can be associated with other malformative ear abnormalities. Identification and characterization of these abnormalities will be crucial for the proper management of patients.We report two cases of bilateral duplicated internal auditory canal with other associated inner ear anomalies. [Patel N Natl J Integr Res Med, 2020; 12(1):105-111]

Key Words: Magnetic resonance imaging (MRI),Internal auditory canal (IAC),Duplicated internal auditory canal (DIAC), Vestibulo cochlear nerve (VCN),Cochlear nerve (CN), Sensorineural hearing loss (SNHL)

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Introduction: Inner ear malformations account for only 20 % of cases of congenital sensorineural hearing loss.[1] Anomalies of the internal auditory canal (IAC) are rare and account for only 12 % of all congenital temporal bone abnormalities.[2,3] Narrow internal auditory canal (IAC) has been well correlated with both unilateral and bilateral congenital sensorineural hearing loss cases. [2,3,4]

It may be associated with other ear abnormalities such as cochlear malformation (Michel deformity, common cavity deformity, cochlear aplasia, and cochlea - vestibular hypoplasia.[4] High-resolution computed tomography (HRCT) of the temporal bone with sections <2 mm thick is the imaging modality of choice to look for bony abnormalities of internal auditory canal (IAC). Magnetic resonance imaging (MRI) helps to evaluate the neural structures of IAC and three dimensional (3D) reformation of cochlea in patients with congenital sensorineural hearing loss.

Duplication of the IAC is associated with SNHL because of aplasia or hypoplasia of vestibulocochlear nerve.[5] Vestibulocochlear nerves (VCNs) or cochlear nerves (CNs) in stenotic IACs are usually dysplastic; and most of these patients can undergo electronic cochlear implantation which can be beneficial in them. [6] In comparison, in DIAC, these nerves usually appear aplastic; thus, electronic cochlear implantation may not be beneficial in them. Hence, preoperative evaluation and diagnosis of DIAC through cross sectional imaging is necessary to facilitate selection of the appropriate treatment.

Case Report: We present cases of two siblings with bilateral narrow duplicated internal auditory canal. A 7 year girl (Sibling 1, elder of two siblings) and A 4 year boy (Sibling 2, younger of two siblings) were referred to our department for the radiological evaluation for bilateral profound hearing loss since birth and for cochlear implant evaluation. Both of them had normal antenatal history (full-term pregnancy and uneventful delivery) and normal postnatal history (no NICU stay) and were otherwise develop mentally normal. There was no family history of early- onset hearing loss, no syndromic anomalies were detected, and no other risk factors for hearing loss were found.

Physical examination showed normal head and neck status. External ear features were normal. [Figure 1] [Figure 3]. The facial nerve function was normal on both sides with family history negative for hereditary SNHL. The patients had no history of mumps, head trauma, or sudden exposure to loud sound. Pure tone audiometry (PTA), Brainstem evoked response audiometry (BERA), Otoacoustic emissions (OAE) and Auditory steady state response (ASSR) all of these test shows bilateral severe to profound sensorineural hearing loss in both siblings.

Axial and coronal reformatted HRCT images of the temporal bones were obtained using 128 slice CT (Somatom Definition, Siemens, Germany) by using a collimation of 0.6 – 0.75 mm. Scans extend from the top of the petrous apex to the mastoid tip in the axial direction and from the

anterior tip of the petrous apex to the posterior margin of the mastoid in the coronal direction. For further evaluation of the seventh and eighth cranial nerves, high-resolution MR imaging was performed using the 1.5 T Achieva system (Philips Medical Systems).

MR imaging protocols included the following: A three-dimensional driven equilibrium radio frequency pulse (3D DRIVE) sequence and T2-weighted spin- echo (TSE) sequence on axial and parasagittal planes.

Case 1: On imaging, In Sibling 1 (7 year old girl), Narrowed left internal acoustic meatus seen with bony septum within dividing the IAC into anterosuperior (Facial) and inferoposterior (Vestibulocochlear) canal suggestive of Duplicated internal auditory canal on left side.[Figure2: –a,b)] [Table 1]

Narrowed right internal acoustic meatus seen with bony septum within its lateral portion(approx. 4.5 mm lateral to porus acousticus) dividing the IAC into anterosuperior (Facial) and inferoposterior (Vestibulocochlear) canal suggestive of Duplicated internal auditory canal on right side.[Figure 2 –a ,b)] [Table 1].

The anterosuperior portion ended in the facial canal and a narrow connection to the vestibule[Figure 2 –d)].

The inferoposterior portion ended in narrow connections to the cochlea and vestibule [Figure 2 –c)].
Facial nerves (FNs) in cisternal segment on either side were normal. Vestibulocochlear nerve in the cisternal segment on either side was faintly visualized. [Figure 2 –f)].

Divisions of the VCN, in the relatively narrow duplicated canals were not visualized possibility of aplastic cochlear nerve on either side. [Figure 2 –g)]

There was also non visualisation of lateral semicircular canal on either side suggest possibility of aplastic lateral semicircular canal .

Cochlea was normal on either side.[Figure 2 –h)] No external and middle ear malformations were found[Figure 1].

Figure 1: (7 Yr Old Girl) Shows Normal External Ear-No Malformation Found

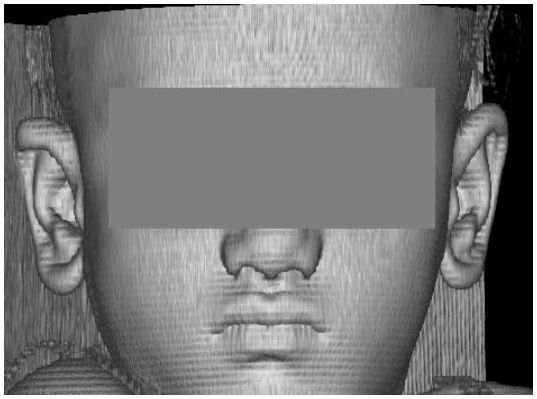

Figure 2: Narrow Internal Auditory Canal With Duplication In 7 Yr Old Girl:

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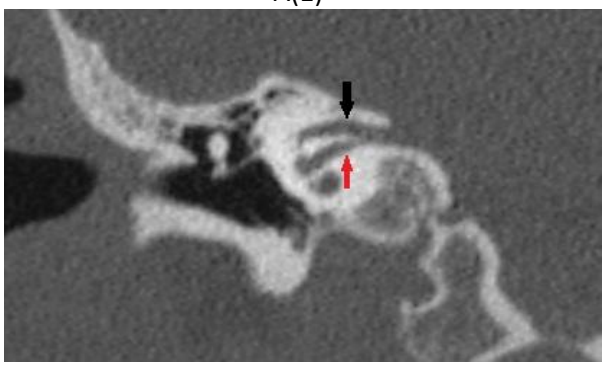

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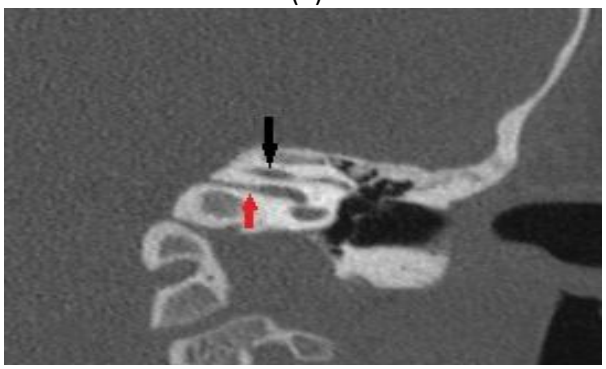

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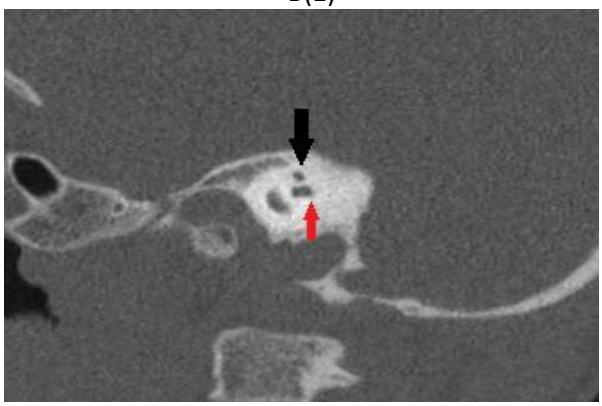

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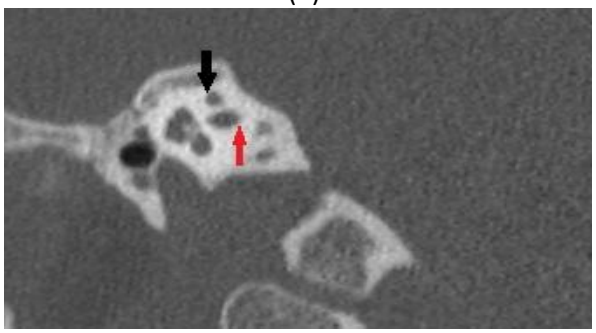

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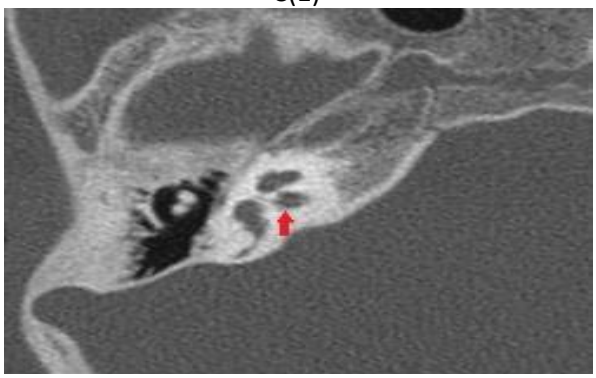

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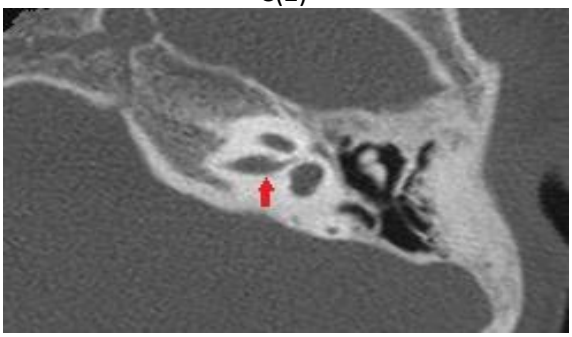

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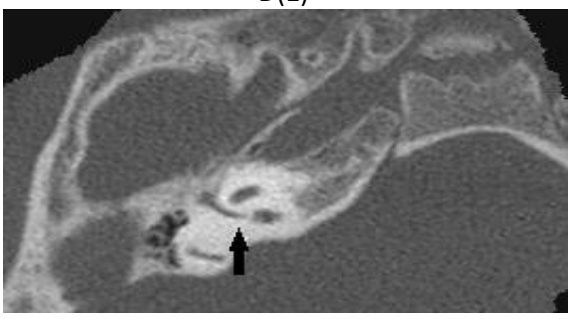

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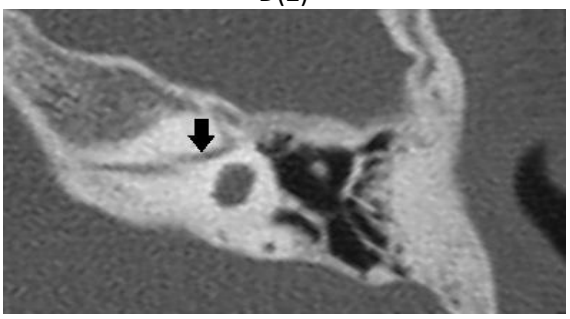

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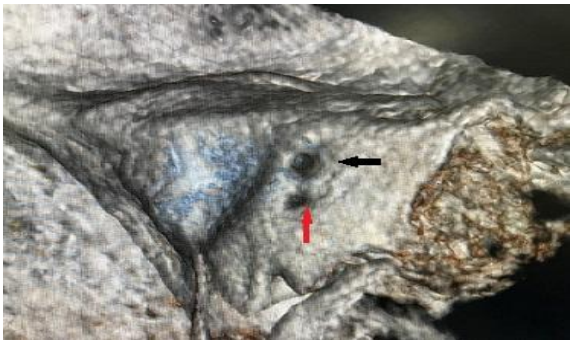

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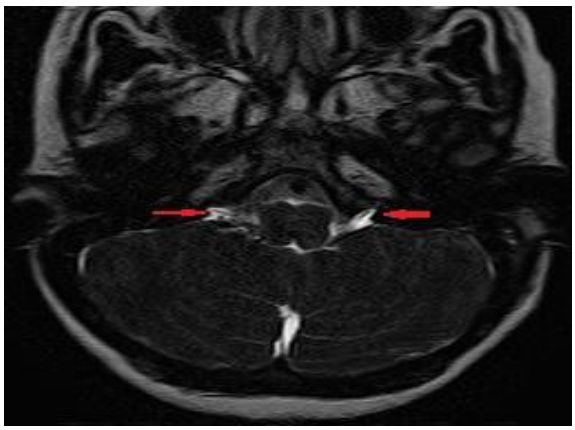

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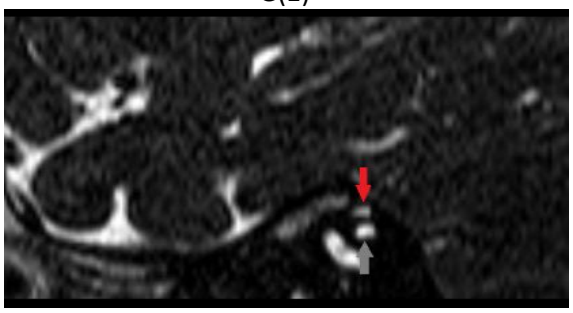

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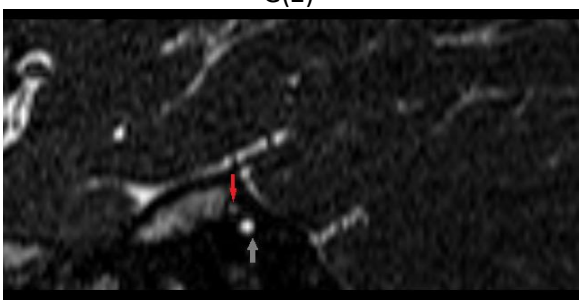

H

Figure 2: Narrow Internal Auditory Canal With Duplication In 7 Yr Old Girl:

A. (1) Right ear and (2) Left ear - Coronal CT image of temporal bone shows narrow internal auditory canal with bony septum within dividing the IAC into anterosuperior(Facial canal) (black arrow)and inferoposterior (Vestibulocochlear canal) (red arrow) .

B. (1) Right ear and (2) Left ear - Sagittal CT image of temporal bone shows same findings with anterosuperior portion ended in the facial canal (black arrow) and inferoposterior portion ended in narrow connections to the cochlea and vestibule (red arrow).

C. (1) Right ear and (2) Left ear - Axial CT image of temporal bone shows narrow internal auditory canal -inferoposterior portion vestibulocochlear canal (red arrow).

D. (1) Right ear and (2) Left ear - Axial CT image of temporal bone shows narrow internal auditory canal - anterosuperior portion facial canal (black arrow) .
E. 3D reconstruction revealed two bony internal auditory canal .

F. 3D drive T2 weighted axial MRI image, shows normal cisternal segment of facial nerve and faint visualisation of cisternal segment of vestibulocochlear nerve within internal auditory canal on either side .

G. 3D drive T2 weighted parasagittal image of internal auditory canal -(1) Right ear and (2) Left ear - Nerves were not seen in canalicular segment anterosuperior portion(Facial canal) (red arrow)and inferoposterior portion (Vestibulocochlear canal) (grey arrow).

H. 3D reconstruction of inner ear revealed normal cochlear turns on both sides with normal posterior and superior semicircular canals but absent lateral semicircular canal.

Table 1: The Measurements On HRCT

Structure Size	Sibling 1		Sibling 2	
	Right Ear	Left Ear	Right Ear	Left Ear
Anterosuperior canal(facial)(mm)	0.9	1.1	0.9	1.6
Inferoposterior canal (Vestibulo cochlear) (mm)	1.8	1.9	1.2	1.9

Case 2: In Sibling 2 (4 year old boy), On imaging, similar findings were noted. Narrowed left internal acoustic meatus seen with bony septum within dividing the IAC into anterosuperior(Facial) and inferoposterior (Vestibulocochlear) canal suggestive of Duplicated internal auditory canal on left side.[Figure 4 –a,b)] [Table 1].

Narrowed right internal acoustic meatus seen with bony septum within its lateral portion(approx. 5 mm lateral to porus acousticus) dividing the IAC into anterosuperior(Facial) and inferoposterior (Vestibulocochlear) canal suggestive of Duplicated internal auditory canal on right side[Figure 4 –a ,b)] [Table 1].

Facial nerve (FNs) in cisternal segment on either side was normal. Vestibulocochlear nerve in the cisternal segment on either side was faintly visualized[Figure 4 –f)]. Divisions of the VCN, in the relatively narrow duplicated canals were not visualized possibility of aplastic cochlear nerve on either side[Figure 4 –g)].

There was also non visualisation of lateral semicircular canal on either side except focal bulge at the site of opening of lateral semicircular canal into vestibule. Cochlea was normal on either side[Figure 4 –h)]. No external and middle ear malformations were found[Figure 3].

Figure 3: (4 Yr Old Boy) Shows Normal External Ear-No Malformation Found

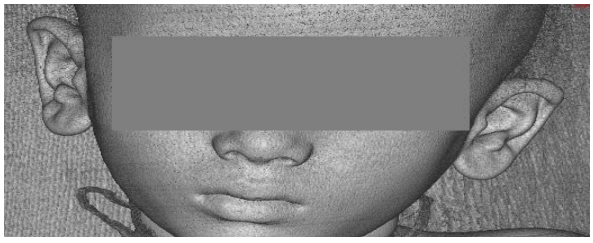

Figure 4: Narrow Internal Auditory Canal With Duplication In 4yr Old Boy:

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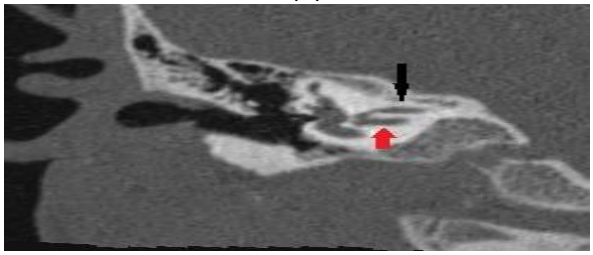

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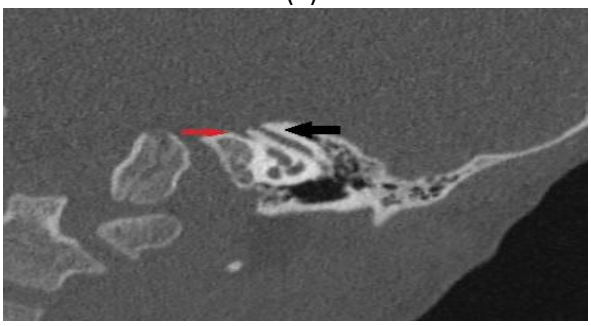

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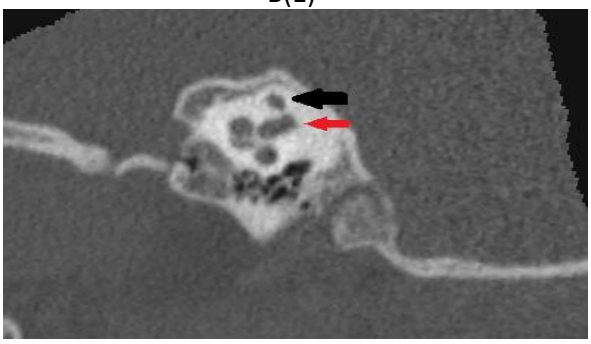

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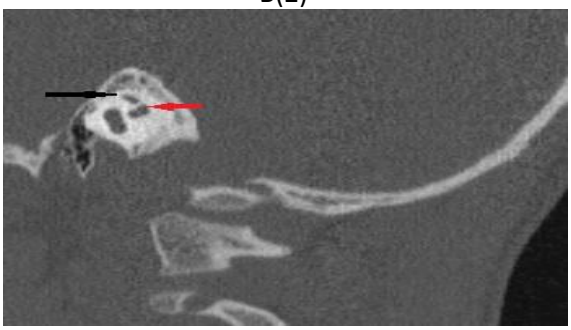

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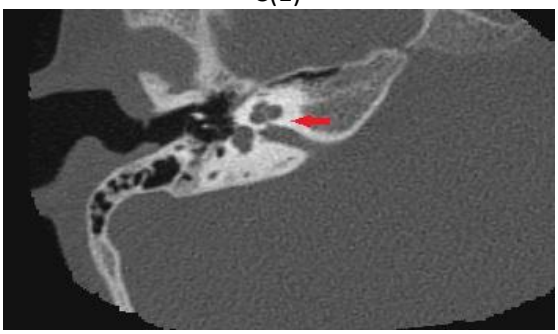

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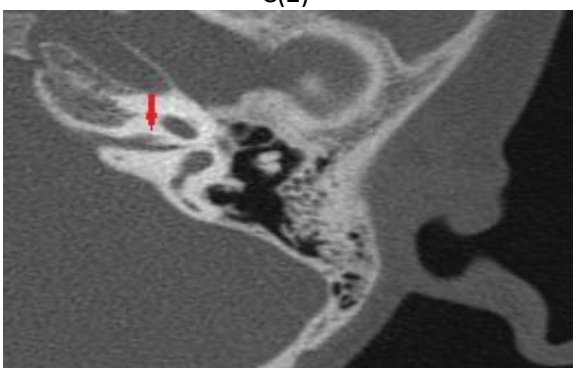

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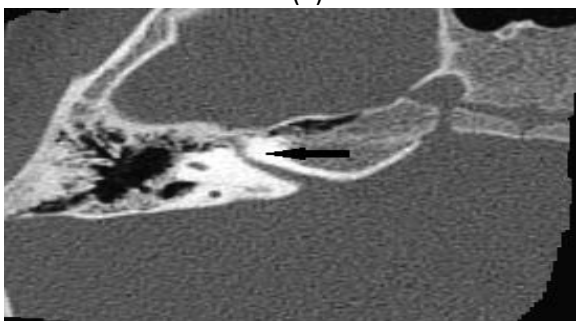

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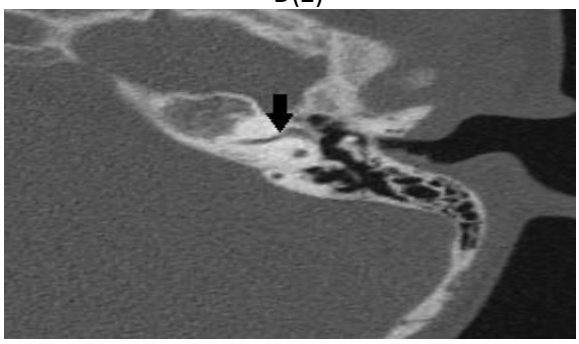

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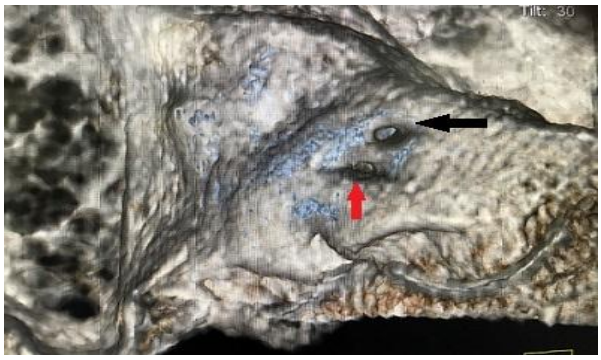

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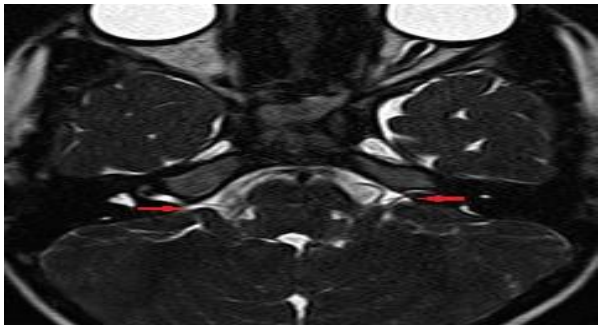

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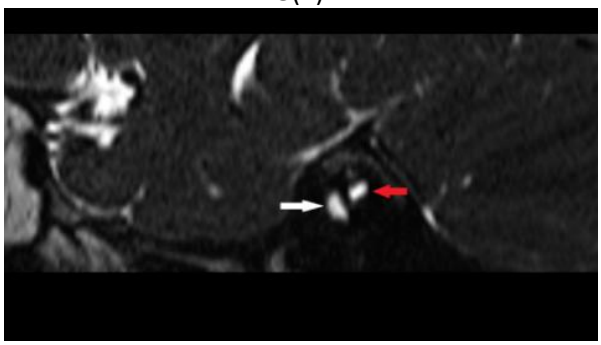

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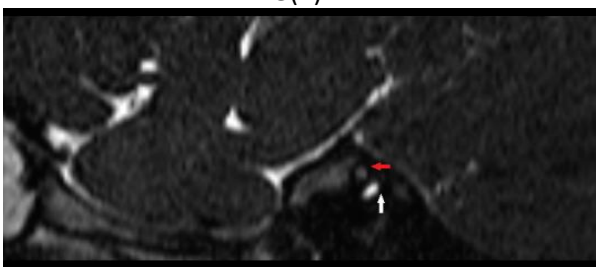

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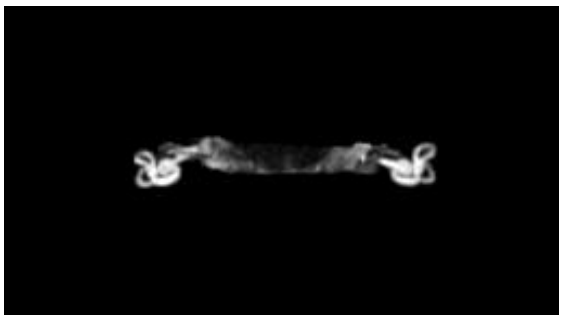

Figure 4: Narrow Internal Auditory Canal With Duplication In 4yr Old Boy:

A. (1) Right ear and (2) Left ear - Coronal CT image of temporal bone shows narrow internal auditory canal with bony septum within dividing the IAC into anterosuperior(Facial canal) (black arrow)and inferoposterior (Vestibulocochlear canal) (red arrow .

B. (1) Right ear and (2) Left ear - Sagittal CT image of temporal bone shows same findings with anterosuperior portion ended in the facial canal (black arrow) and inferoposterior portion ended in narrow connections to the cochlea and vestibule (red arrow).

C. (1) Right ear and (2) Left ear - Axial CT image of temporal bone shows narrow internal auditory canal -inferoposterior portion vestibulocochlear canal (red arrow).

D. (1) Right ear and (2) Left ear - Axial CT image of temporal bone shows narrow internal auditory canal - anterosuperior portion facial canal (black arrow).

E. 3D reconstruction revealed two bony internal auditory canals.

F. 3D drive T2 weighted axial MRI image, shows normal cisternal segment of facial nerve and faint visualisation of cisternal segment of vestibulocochlear nerve within internal auditory canal on either side.

G. 3D drive T2 weighted parasagittal image of internal auditory canal -(1) Right ear and (2) Left ear -Nerves were not seen in canalicular segment anterosuperior portion(Facial canal) (red arrow)and inferoposterior portion (Vestibulocochlear canal) (grey arrow).

H. 3D reconstruction of inner ear revealed normal cochlear turns on both sides with normal posterior and superior semicircular canals but absent lateral semicircular canal.

Discussion: Hearing impairment is the most common sensory defect in humans.[5] Hearing loss can be conductive, sensorineural or mixed type. Congenital hearing loss can be caused external, middle or inner ear abnormalities and malformations, inherited syndromes or congenital cholesteatoma.[6] The diameter of internal auditory canal varies from 2 to 8 mm in different individuals .A narrow IAC is represented by an IAC of diameter 2 mm or less.[7] This malformation comprises 12% of congenital temporal bone anomalies.[8]

Development of inner ear begins early in the 4th week of gestation, with maturation at 8–16 weeks and conversion from the cartilaginous to the bony labyrinth occurs between 16 and 23 weeks of gestation.[9] Jackler et al.[10] proposed that arrested development at various stages of embryonic development can result in distinct inner ear malformations.

The exact mechanism is unclear but there are two widely accepted hypotheses that suggest the association of a narrow IAC with SNHL. During the ninth week, the cartilaginous IAM gradually develops in synchrony with the vestibulocochlear nerve's development. Some investigators hold that when the vestibulocochlear nerve is aplastic or hypoplastic, the IAC fails to develop and becomes stenotic. 9and therefore, the absence of the vestibulocochlear nerve causes IAM aplasia or stenosis, while others believe that that the primary defect is bony stenosis that inhibits the growth of the vestibulocochlear nerve and hence transmission of induction signal from intact cochlea and vestibule gets impaired. 10 In both cases there is association of narrow internal auditory canal with sensorineural hearing loss.

Stenotic internal auditory canal is associated with vestibulocochlear nerve or cochlear nerve

dysplasia, while duplicated internal auditory canal is associated with cochlear nerve aplasia. Hence differentiation between these two anomalies is necessary for appropriate management.Duplicated internal auditory canal is a rare congenital anomaly and it is even rarer in bilateral ears. It can be associated with other temporal malformations. In our case study duplicated internal auditory canal was present in either ear in both siblings.

Facial nerve in cisternal segment was normal in both siblings with faint visualization of cisternal segment of bilateral vestibulocochlear nerve. Divisions of vestibulocochlear nerve in narrow duplicated canal were not visualized possibility of aplastic cochlear nerves .There were also aplastic lateral semicircular canal on either side in both siblings. No other malformations were present.

Conclusion: Magnetic resonance imaging and computed tomography are useful in identifying inner ear malformations and in the assessment of the anatomical structures .CT allows us to detect bone malformations, the facial canal(possible aberrant trajectory),and is useful in assessing coexisting anomalies of both the inner and middle ear. When it comes to the MRI it allows us to assess neural structures of IAC, the cranial nerves, fluid-filled spaces of the IE and any other possible intracranial anomalies. Cross sectional imaging help to provide the surgeon with the necessary pre surgical information that will allow him/her to make a proper decision, choose the appropriate implants and warn about anatomic variants and possible surgical complications that can occur.

Duplicated internal auditory canal is a rare congenital anomaly which can be associated with aplastic or hypoplastic vestibulocochlear nerve. Characteristic feature on HRCT scan is presence of bony septum in internal auditory canal with double-canal appearance, and it can be accompanied by other ear anomalies.

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