

Duplicated internal auditory canal with dysplastic ossicles and microtia: role of high-resolution CT and MRI

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DESCRIPTION

A 16-year-old girl with congenital hearing loss was referred to our department for radiological evaluation and feasibility for cochlear implantation. She had previously used various external hearing aids with no significant success. Her antenatal and post-natal history was normal with no family history of congenital or early-onset hearing loss.

Physical examination revealed shorter neck and bilateral grade 2 microtia (**figure 1A,B**) with normal external auditory canals. No syndromic stigmata-like pigmentary anomalies and craniofacial features were seen. Facial nerve functions were normal on both sides; however, her speech and language functions were markedly impaired. Complete audiological evaluation was suggestive of bilateral asymmetrical mixed hearing loss, and hence, the child underwent high-resolution CT (HRCT) temporal bone for further workup.

HRCT was performed on a 64-slice multidetector CT scanner (Philips Ingenuity, Philips Healthcare, Cleveland, Ohio, USA) which showed duplicated internal auditory canal (DIAC) on the right side, with a narrow posteroinferior and broader anterosuperior compartment. The posteroinferior canal (<1 mm diameter) was terminating into the cochlea and vestibule (**figure 1C**), while the anterosuperior compartment was continuous with the facial canal (**figure 1D**). The two canals were separated by a bony septum (**figure 1E**). Bilateral ear ossicles were dysplastic (**figure 1F**). Inner ear structures including cochlear turns, vestibule and semicircular canals were normal on both sides. Left internal auditory canal (IAC) was normal. Bilateral external ears were small and deformed; however, external auditory canals were normal. Complete atlanto-occipital assimilation was seen (**figure 1G**). For further evaluation of the VII/VIII nerve complexes, MRI was done on 3 Tesla (SIEMENS, Germany) using three-dimensional constructive interference in steady-state (CISS) images. MRI showed duplicated and narrow right IAC with non-visualisation of right cochlear nerve and normal facial nerve in the anterosuperior canal (**figure 2**). Other bilateral inner ear structures and left VII/VIII nerve complexes were normal. The absence of the cochlear nerve on the right side precluded the feasibility of cochlear implantation. Therefore, ossicular reconstruction of the left ear was advised for correcting conductive deafness, followed by plastic surgery consultation for correction of microtia.

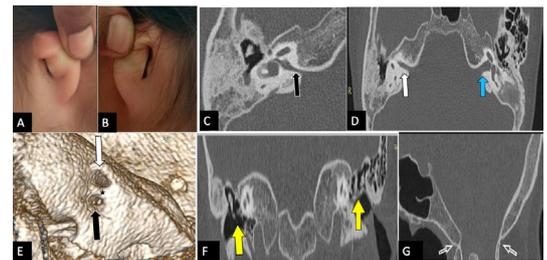


Figure 1 (A, B) Clinical picture showing bilateral grade 2 microtia (A, left ear; B, right ear). (C–G) High-resolution CT (HRCT) temporal bone. (C) HRCT axial image shows narrow posteroinferior canal (black arrow) terminating into cochlea and vestibule. (D) Axial image shows an anterosuperior canal (white arrow) terminating into the facial canal and normal left internal auditory canal (blue arrow). (E) Volume-rendered image of right petrous temporal bone shows duplicated right internal auditory canal (DIAC) separated by a horizontal bony septum (star). Anterosuperior facial nerve canal (white arrow) and narrow posteroinferior vestibulocochlear nerve canal (black arrow). (F) Coronal image shows dysplastic bilateral ear ossicles (yellow arrows). (G) HRCT in the mid-sagittal plane shows atlanto-occipital assimilation (hollow white arrows).

Duplication of IAC (DIAC) is a very rare anomaly with less than 50 cases reported in the literature.¹ Imaging features of duplicated IAC and its



Figure 2 MRI of the inner ear. (A) Axial constructive interference in steady-state (CISS) image (thicker slice) shows duplicated right internal auditory canal (DIAC) with the anterosuperior canal (red arrowhead) terminating into the facial canal and narrow posteroinferior canal (red arrow) terminating into cochlea and vestibule. Normal internal auditory canal (IAC) on the left side (blue arrow). (B, C) Axial CISS images (thinner slices) demonstrate aplastic/hypoplastic right vestibulocochlear nerve (red arrow in B) and normal facial nerve in the right anterosuperior canal (red arrowhead in C). Normal vestibulocochlear/facial nerve complex in the left IAC (blue arrow in B).



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Images in...

association with other ear malformations are not well understood. Duplication of IAC usually occurs unilaterally and is often associated with the aplastic/hypoplastic vestibulocochlear nerve.¹ It is important to differentiate IAC stenosis from DIAC

on imaging because patients with IAC stenosis have dysplastic vestibulocochlear nerve and they usually benefit from electronic cochlear implantation, while cases with DIAC have aplastic/hypoplastic nerve and are usually not benefited from electronic cochlear implantation.^{2,3} External, middle and inner ear abnormalities are occasionally seen with DIAC which are additionally responsible for the hearing loss and need consideration while planning for surgery.¹ Rarely, DIAC can be associated with syndromes like Klippel-Feil syndrome and pontine tegmental cap dysplasia.⁴ Thus, HRCT followed by MRI is necessary for the complete evaluation of cases of DIAC.

Patient's perspective

We were very worried as our daughter was not able to hear and speak properly since birth. She tried multiple hearing aids without much benefit. We were referred to the hospital for the cochlear implant. After testing her ears with Audiometry, CT & MRI, doctors told us that the cochlear implant will not be beneficial for her. However, they also explained to us that her left ear is relatively normal in which hearing could be improved by an operation. Due to my financial constraints, I am not able to get it done immediately. But I will surely get her operated as soon as finances are arranged. (Patient's father)

Learning points

- ▶ Duplication of the internal auditory canal (DIAC) is a very rare anomaly and produces a double canal appearance on cross-sectional imaging studies. The two canals are usually separated by a bony septum.
- ▶ Differentiation of IAC stenosis from DIAC is extremely important in appropriate selection of patients for electronic cochlear implantation because patients with IAC stenosis usually benefit from cochlear implantation than DIAC.
- ▶ In cases with hearing loss, DIAC is usually accompanied by other ear abnormalities. Thus, HRCT and MRI with multiplanar reconstruction and volume rendering are necessary for complete preoperative evaluation of DIAC and to rule out other associated syndromes.

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Disclaimer Case reports provide a valuable learning resource for the scientific community and can indicate areas of interest for future research. They should not be used in isolation to guide treatment choices or public health policy.

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