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Case Report

Asymmetrically severe internal auditory canal hypoplasia: A case report

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ABSTRACT

We present a case of an otherwise healthy 20-month-old with congenital sensorineural hearing loss. CT and MR imaging demonstrated bilateral asymmetrically severe hypoplasia of the internal auditory canals and vestibulocochlear nerves. Additional developmental inner ear anomalies were present in this patient, including unilateral semicircular canal hypoplasia and suspected bilateral cochlear hypoplasia. The patient retained normal facial nerve function bilaterally. We highlight the current research and understanding of congenital IAC abnormalities.

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Case report

A 20-month-old female, born 37 weeks gestation, was referred to our institution for sensorineural hearing loss. The patient had no known significant pertinent birth or perinatal medical history. The patient had no facial dysmorphism and demonstrated normal facial nerve function. The patient demonstrated abnormal auditory brainstem response testing beginning at age of 4 months. At the time of initial referral to our center, she had been using hearing aids for nearly 1 year. On clinical examination, she did not yet ambulate independently, limiting clinical assessment of gait and equilibrium. She responded only to loud noises while using hearing aids and exhibited delayed spoken language development.

CT and MRI were performed as part of her workup, both of which demonstrated bilateral asymmetrically severe inner ear abnormalities. The right side was more severely involved, exhibiting nonvisualization of both the internal auditory canal and vestibulocochlear nerve (Fig. 1), compatible with severe hypoplasia/aplasia of both structures. On the left, a hypoplastic internal auditory canal was seen, with questionable visualization of a hypoplastic vestibulocochlear nerve (Fig. 2).

A few associated findings were also noted. A normal partition of the cochlea and vestibule was seen bilaterally, though the bilateral vestibules were enlarged (Fig. 3). There was

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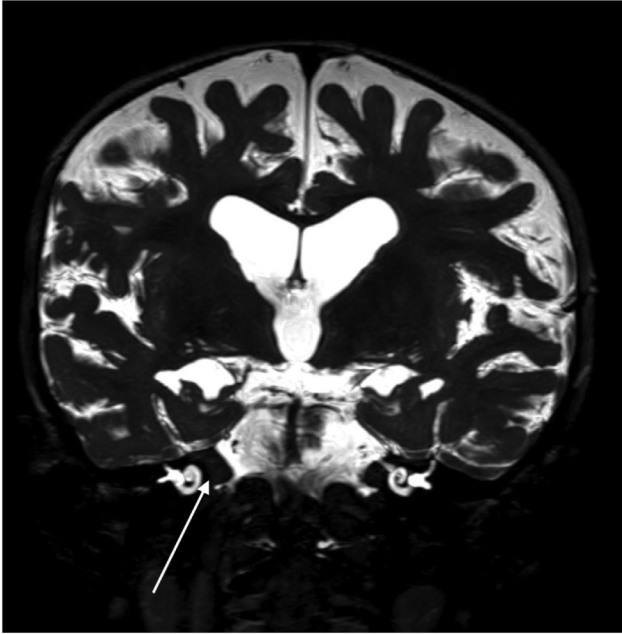


Fig. 1 – Heavily T2-weighted coronal MRI of the skull base centered at the expected region of the internal auditory canals: 4.0 mm multiplanar reconstruction of 0.5 mm slices. There is nonvisualization of the right internal auditory canal and vestibulocochlear nerve, compatible with severe hypoplasia/aplasia.

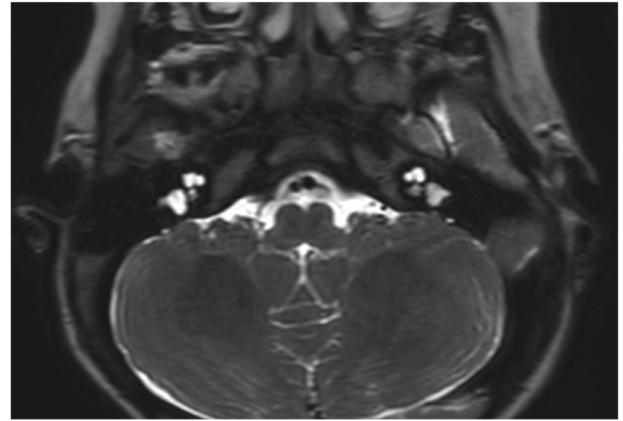


Fig. 3 – Axial T2-weighted MRI of the skull base. There is enlargement of the bilateral vestibules. There is possible bilateral cochlear hypoplasia, with the appearance of 1.5 turns.

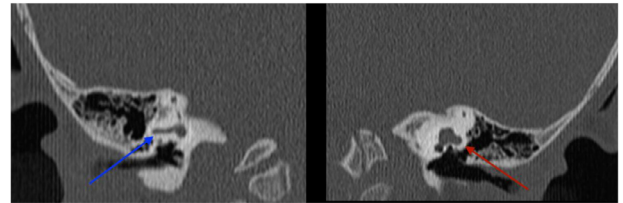


Fig. 4 – Coronal CT of the bilateral inner ear structures. The right lateral semicircular canal is intact (blue arrow), while the left lateral semicircular canal terminates as a hypoplastic short, wide trunk (red arrow). The remainder of the semicircular canals are normal in appearance (not imaged). From an embryological perspective, the lateral semicircular canal is expected to be the first malformed canal in the spectrum of inner ear developmental anomalies [11]. (Color version of figure is available online.)

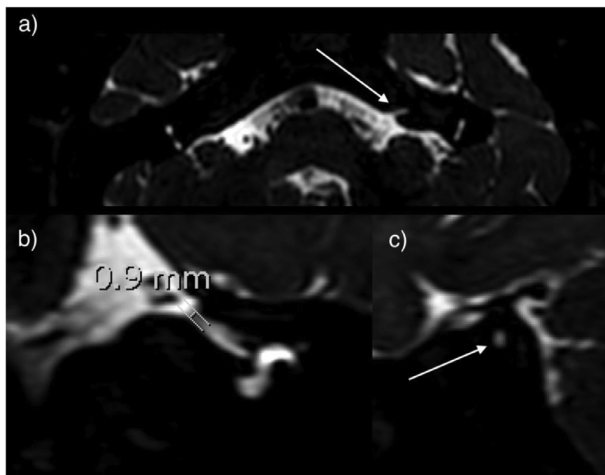


Fig. 2 – Heavily T2-weighted (A) axial, (B) coronal, and (C) sagittal MRI reconstructions of the skull base centered at the expected region of the internal auditory canals. The left internal auditory canal is hypoplastic, measuring a minimum vertical diameter of approximately 0.9 mm. Faint wispy internal intermediate signal within the narrow canal may reflect a poorly visualized hypoplastic left vestibulocochlear nerve.

suspected cochlear hypoplasia with 1.5 turns of the cochlea bilaterally (Fig. 3). The left lateral semicircular canal was noted to be hypoplastic (Fig. 4). There was no enlargement of the endolymphatic sacs bilaterally.

After obtaining the above imaging, the patient was documented in our electronic health record as pending cochlear implant consultation. No further clinical management has been since documented at the time of writing.

Discussion

Congenital sensorineural hearing loss (SNHL) affects 1 in 1000 newborns and represents a barrier to normal speech and language development in the affected child. Approximately 20%-35% patients with congenital SNHL have visualized abnormalities of the inner ear [1]. High resolution CT and MRI of the temporal bones and IAC is recommended to adequately

assess osseous anatomy, fluid spaces, and integrity of the vestibulocochlear nerve (VCN).

A vertical IAC diameter of less than 2 mm is generally considered the definition for IAC hypoplasia [2]. IAC abnormalities are frequently accompanied by hypoplasia/aplasia of the VCN, with 92% of IAC malformations exhibiting aplasia of the VCN in a case series [5]. The facial nerve, however, may retain functionality in the setting of IAC atresia (as in this case) by taking an aberrant superoventral course [6]. Such a course was not directly visualized on imaging in this patient.

The IAC develops from mesoderm enveloping the VCN, which eventually transforms into cartilage and ossifies. Current understanding suggests that development of the IAC is intricately associated with that of the VCN; atresia or hypoplasia of IAC is likely a result of altered VCN development secondary to faulty chemotactic mechanisms or lack of end organ targets [3,7]. While FGF3 mutations have been linked to IAC and other inner ear structural abnormalities, the molecular genetics governing these abnormalities remain poorly defined. It is believed that bilateral symmetric cases are usually associated with genetic factors, whereas asymmetric cases, such as seen in this patient, may be more likely due to external insults [8].

IAC hypoplasia/atresia has been associated with other inner ear abnormalities [4], such as incomplete partition variants, cochlear and semicircular canal hypoplasia, and complete labyrinthine aplasia (Michel aplasia). Numerous classification systems have been proposed to account for the myriad presentations of inner ear malformations, which will not be discussed here [9–11]. In addition to bilateral asymmetric IAC hypoplasia/atresia, this patient had left semicircular canal hypoplasia and suspected bilateral cochlear hypoplasia.

CT and MR imaging of the temporal bone should be performed prior to cochlear implantation to assess the osseous anatomy, fluid spaces, and integrity of the VCN. The absence of a VCN has generally been considered a relative contraindication to cochlear implants [12]. Auditory brainstem implantation may represent a therapeutic alternative for these patients.

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