

# Internal auditory canal hypoplasia associated with bilateral vestibulocochlear nerve aplasia and deviant facial nerve course: A case report and MRI findings

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## ABSTRACT

The evaluation of internal auditory canals and cochlea has gained significant importance due to the increasing number of cochlear implantations worldwide. This region's anatomical study is essential for cochlear implant surgery using magnetic resonance imaging as the method of choice. We report a case of a 6-year-old male patient diagnosed with a rare bilateral malformation of the internal auditory canals associated with an aberrant course of the facial nerve and vestibulocochlear nerve aplasia. This report raises the importance of identifying this rare malformation for appropriate management and reinforces awareness of possible complications.

## CASE REPORT

### CASE REPORT

A 6-year-old male patient with marked bilateral congenital sensorineural hearing loss (SNHL) presented to the outpatient clinic for evaluation of possible cochlear implantation.

The neurological and otorhinolaryngological exams showed abnormal language development and difficulties with social interaction. Absent otoacoustic emissions and brainstem auditory evoked potentials were also noted bilaterally. He tried hearing aids without improvement. No other neurodevelopmental or neurological abnormalities were observed, including spared facial nerves (FN) function.

Magnetic resonance imaging (MRI) demonstrated bilateral stenosis of the internal auditory canals (IAC) and the absence of the cisternal and intracanalicular fibers of the vestibulocochlear nerve, indicating agenesis (Figures 1 and 2). An stenotic IAC was considered if it measured less than 2 mm in the coronal plane (Figure 1). An associated anomalous intracranial course of the FN was observed bilaterally (Figures 3 and 4).

Due to patient's inner ear malformations, including cochlear nerve aplasia, cochlear implantation was contraindicated.

## DISCUSSION

### Etiology & Demographics:

The aberrant FN is not rare in patients with congenital anomaly of the middle ear. In their series of 227 subjects with middle ear anomalies, Hao and colleagues found aberration of FN in 32% of patients, with facial nerve displacement as the most commonly observed malformation [1]. The aberrant FN may cause blockage of the oval window and cross over other structures [1]. The majority of patients with FN anomaly do not present with clinical symptoms. However, when it is present, it is usually due to conductive hearing loss from an accompanying ossicular chain abnormality [1,2].

The prevalence of congenital sensorineural hearing loss (SNHL) is approximately 1-6 per 1,000 live births [3,4]. Hypoplasia of the IAC is a rare malformation, believed to be due to agenesis of the vestibulocochlear nerve (VCN) [5]. When this association is present and there is no dysfunction of the FN, it may course posteroinferiorly to the trigeminal nerve [5].

Rarely, the aberrant FN may run into a separate canal, usually preserving normal function [6]. Moreover, an aberrant anterior course of the FN often represents the smallest distance to the facial musculature [7].

### Clinical & Imaging findings:

The evaluation of IAC and cochlea has gained great importance due to the increasing number of cochlear implantations throughout the world [6]. MRI is the method of choice to investigate pathologies in this region, both congenital and acquired [8].

The diagnosis of IAC hypoplasia and VCN agenesis associated with an abnormal FN course is essential for the cochlear implant preoperative evaluation. While FN course variations commonly occur in the middle ear, these are rare in the intratemporal and intracranial compartments [9]. According to Wu C. *et al.*, it is rare to find an anomalous course of FN in patients without abnormalities in the external ear or tympanic membrane, as presented in the case [11]. The diagnosis of IAC malformations with or without variances in the FN and bilateral VCN agenesis are usually made by MRI [3,4]. Moreover, computed tomography (CT) of the temporal bone can also evaluate this anatomic region [9,12]. De Foer B. *et al.* highlights the importance of axial and parasagittal reformations of heavily 3D T2-weighted sequences for the assessment of aplasia or hypoplasia of the VCN [6].

Unfamiliarity with the FN aberrant course may lead to surgical complications, including inadvertent injuries [10, 13]. In our case, there was no dysfunction of the FN, despite its rare abnormal course. On MRI, an anomalous intracranial course of the FN was observed bilaterally. The cisternal segment was dislocated inferior to the trigeminal nerve, running towards the Gasserian ganglion and taking a sharp lateral turn into a minute canal, reaching the geniculate ganglion (Fig. 1-3).

### Treatment & Prognosis:

Cochlear nerve aplasia and other malformations of the IAC constitute contraindications for cochlear implantations. In such cases, an auditory brainstem implant (ABI) may be suggested. An ABI is a device placed directly on the cochlear nuclei in the fourth ventricle through a retrosigmoid-transmeatal approach [14-16]. This type of implant is a more recent auditory rehabilitation technique, usually performed in patients with neurofibromatosis type 2 (NF2) suffering from bilateral hearing loss due to disruption of the VCN [7]. There are only a few cases of patients with cochlear nerve aplasia undergoing this procedure [17]. Thus, further studies are needed to evaluate the efficacy of this technique in this subgroup of patients.

### Differential Diagnoses:

#### **ABERRANT FACIAL NERVE**

##### **Moebius syndrome**

Moebius syndrome is a rare congenital anomaly characterized by sixth and seventh cranial nerve palsies. Other abnormalities involving the limbs, chest wall, and spine may also be seen. Radiologically, the sixth and seventh cranial nerve nuclei are absent, as seen by flattening of the fourth ventricle and pontine hypoplasia. There may be cerebellar hypoplasia, calcification of the pons in the region of the abducens nerve, and absence of the middle cerebellar peduncles.

##### **Aberrant facial nerve (FN) canal**

The FN arises inferiorly and laterally from Meckel's cave, then courses through the petrous bone to the IAC fundus. Distally there is normal morphology of the remaining labyrinthine, tympanic, and descending mastoid facial nerve segments.

##### **Hypoplastic facial nerve canal**

The labyrinthine facial nerve is reduced in caliber. It may be isolated or associated with the other segments' hypoplasia and can be often seen in inner ear dysplasia.

##### **Hypoplastic internal auditory canal and a separate facial nerve canal**

The facial nerve may lie anterosuperior to the IAC, sometimes seen in association with inner ear dysplasia.

## TEACHING POINT

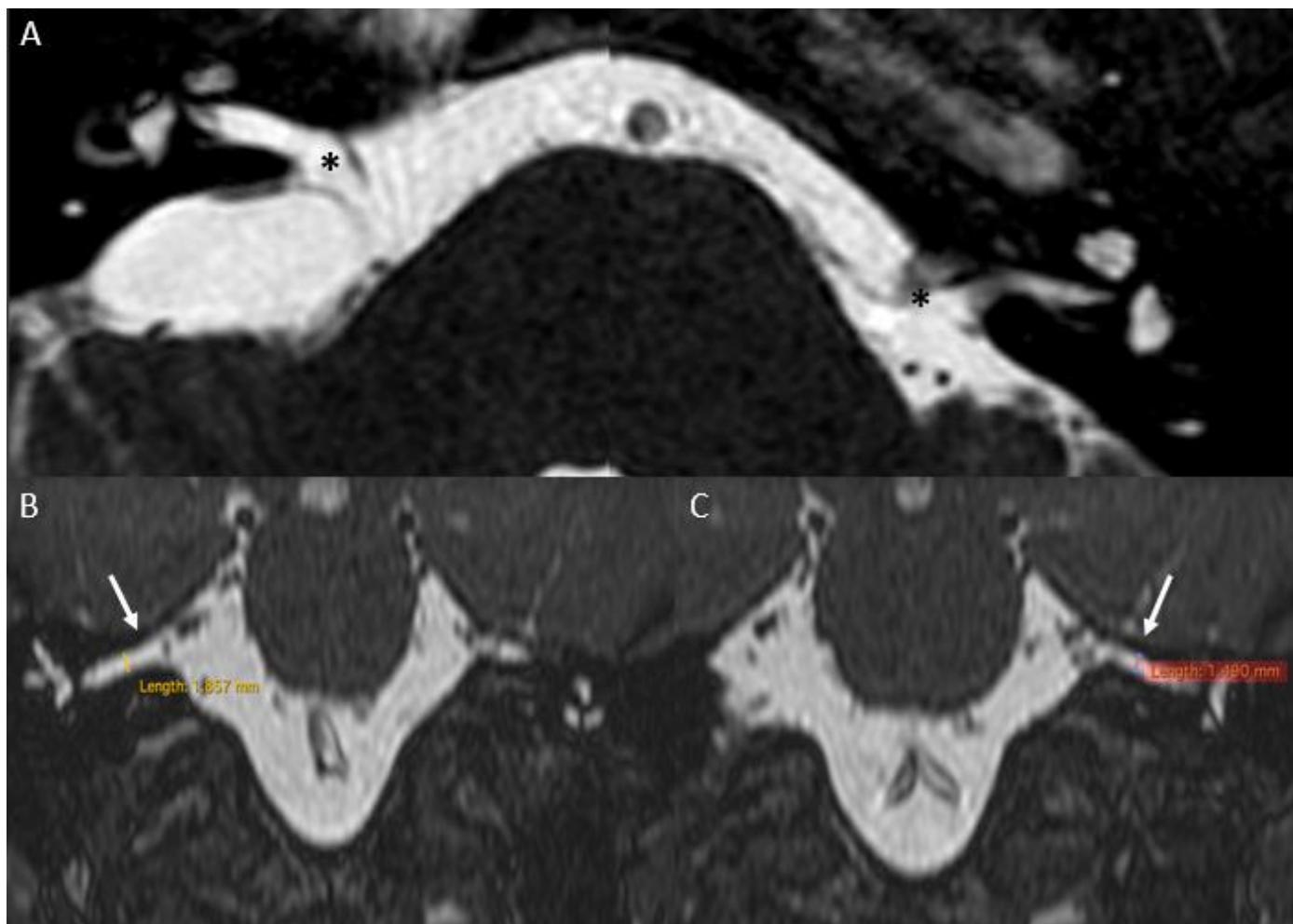
Hypoplasia of the IAC is associated with bilateral agenesis of the VCN. Anomalous FN course is a rare and important auditory system abnormality. Aberrant intracranial FN course should be investigated in patients with hypoplasia of the internal auditory canal and normal FN function.

A potential alternative treatment for children with an absent or deficient eighth cranial nerve is auditory brainstem implantation. The aberrant FN course may increase the risk of complications during this and other otorhinolaryngological procedures.

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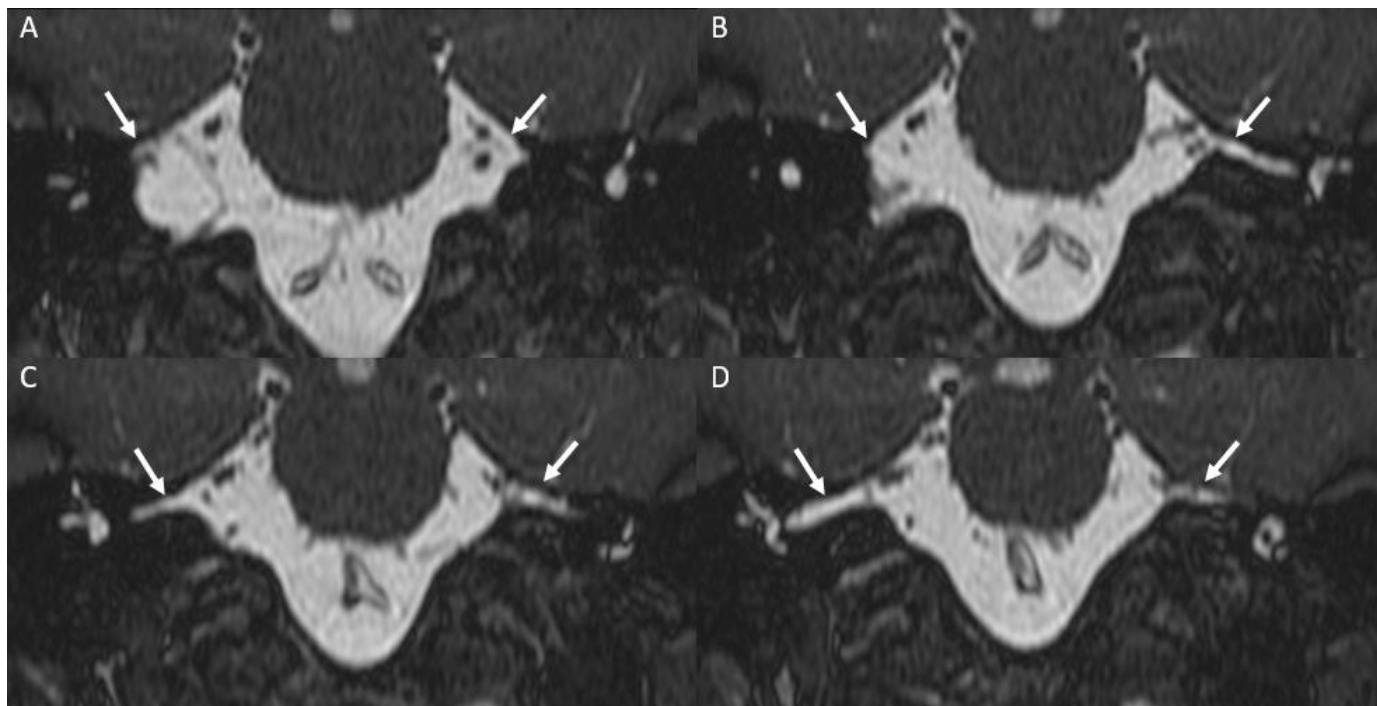
FIGURES



**Figure 1:** A 6-year-old male presented with marked bilateral congenital sensorineural hearing loss.

Findings: MRI brain performed on initial diagnosis. Non-contrast axial (A) and coronal (B and C) fast imaging employing steady-state acquisition (FIESTA) shows no vestibulocochlear nerves (black asterisks in A) and bilateral stenosis of the IAC, left (white arrow in C) greater than right (white arrow in B).

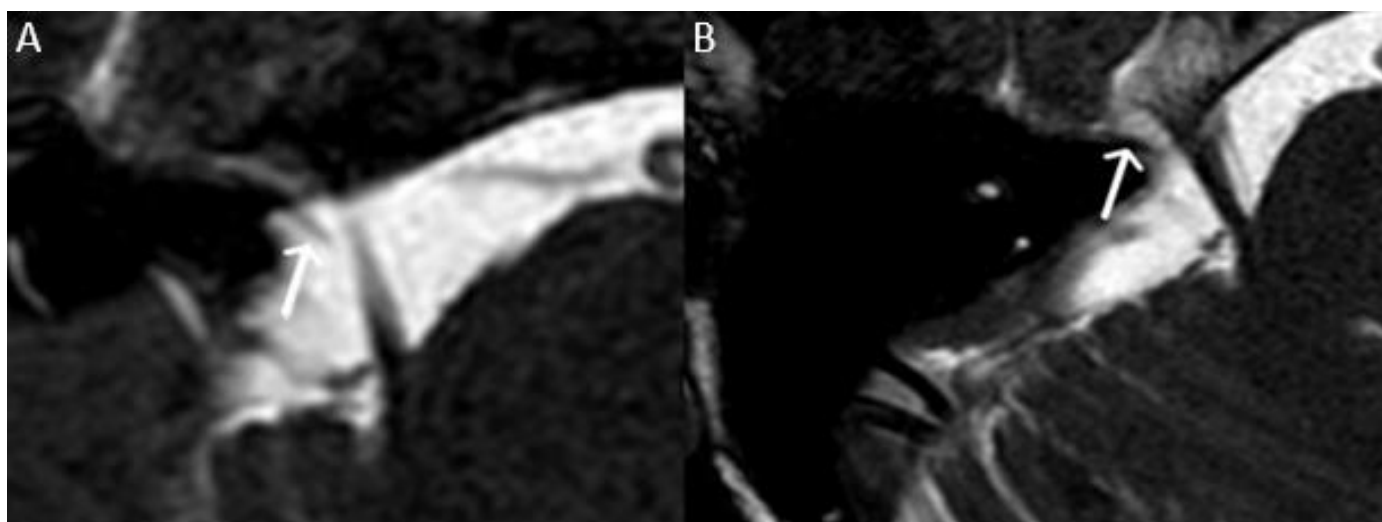
Technique: MRI, Philips 1.5-Tesla; Axial fast imaging employing steady-state acquisition (FIESTA): TR 6.9875, TE: 1.000.



**Figure 2:** A 6-year-old male presented with marked bilateral congenital sensorineural hearing loss.

Findings: MRI brain performed on initial diagnosis. Non-contrast sequential coronal images through the internal auditory canals (IAC) using fast imaging employing steady-state acquisition (FIESTA) shows vessel structures in close anatomical relationship with the IACs but no evidence of vestibulocochlear nerves (white arrows in A, B, C and D).

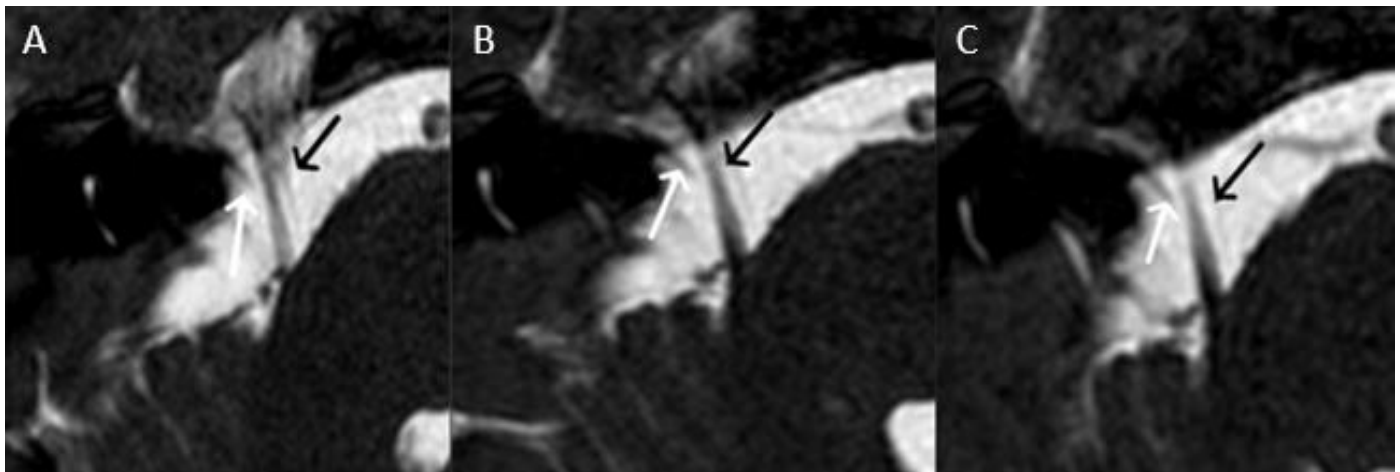
Technique: MRI, Philips 1.5-Tesla; Axial fast imaging employing steady-state acquisition (FIESTA): TR 6.9875, TE: 1.000.



**Figure 3:** A 6-year-old male presented with marked bilateral congenital sensorineural hearing loss.

Findings: MRI brain performed on initial diagnosis. Non-contrast axial fast imaging employing steady-state acquisition (FIESTA) shows the facial nerve's aberrant course in its cisternal segment (white arrow in A), and the minute facial nerve canal (white arrow in B).

Technique: MRI, Philips 1.5-Tesla; Axial fast imaging employing steady-state acquisition (FIESTA): TR 6.9875, TE: 1.000.



**Figure 4:** A 6-year-old male presented with marked bilateral congenital sensorineural hearing loss.

**Findings:** MRI brain performed on initial diagnosis. Non-contrast axial fast imaging employing steady-state acquisition (FIESTA) shows the facial nerve (white arrows in A, B and C) running below the trigeminal nerve (black arrows in A, B and C) towards the Gasserian ganglion. The facial nerve takes a sharp turn into a minute canal (white asterisks in A, B and C).

**Technique:** MRI, Philips 1.5-Tesla; Axial fast imaging employing steady-state acquisition (FIESTA): TR 6.9875, TE: 1.000.

<b>Etiology</b>	Congenital
<b>Incidence</b>	The aberrant facial nerve is a rare abnormality found in patients with congenital anomaly of the middle ear
<b>Age predilection</b>	Congenital malformation
<b>Risk factors</b>	Hypoplasia of the internal auditory canal and agenesis of the vestibulocochlear nerve
<b>Treatment</b>	Surgical treatment in selected cases
<b>Symptoms</b>	Most patients with facial nerve anomaly do not have clinical symptoms
<b>Imaging Findings</b>	The facial nerve runs beneath the trigeminal nerve towards the Gasserian ganglion and takes a sharp turn to the side into a minute canal

**Table 1:** Summary table of aberrant intracranial course of the facial nerve.

<b>Diagnoses</b>	<b>MRI</b>
<b>Moebius syndrome</b>	The sixth and seventh nerve nuclei are absent, as seen by flattening of the fourth ventricle and pontine hypoplasia. There may be cerebellar hypoplasia, calcification of pons in the region of abducens nerve, and absence of middle cerebellar peduncles.
<b>Aberrant facial nerve canal</b>	It arises inferiorly and laterally from Meckel's cave, then courses through the petrous bone to the IAC fundus. There is normal morphology of the remaining labyrinthine, tympanic, and descending mastoid facial nerve segments.
<b>Hypoplastic facial nerve canal</b>	The labyrinthine facial nerve is reduced in caliber. It may be isolated or associated with hypoplasia of the other segments, and it can be often seen in inner ear dysplasia.
<b>Aberrant course of the facial nerve</b>	The facial nerve runs beneath the trigeminal nerve towards the Gasserian ganglion and takes a sharp turn to the side into a minute canal

**Table 2:** Differential diagnosis table for aberrant intracranial course of the facial nerve.

#### ABBREVIATIONS

CT = Computer Tomography  
FN = Facial Nerve  
IAC = Internal Auditory Canal  
MRI = Magnetic Resonance Imaging  
SNHL = Sensorineural Hearing Loss  
VCN = Vestibulocochlear nerve

#### KEYWORDS

Magnetic Resonance Imaging; Vestibulocochlear Nerve Agensis; Aberrant Facial Nerve; Sensorineural Hearing Loss; Internal Auditory Canal Atresia

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