Imaging of Congenital and Acquired Sensorineural Hearing loss: Peeking through the Oval Window into the Cochlea and Beyond

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ABSTRACT

Pure sensorineural hearing loss may be congenital or acquired. It can result from a constellation of abnormalities that may involve the labyrinth, internal auditory canal, CPA, brain stem, or auditory pathways. CT and MR imaging often play a complementary role in its evaluation and postoperative assessment and in ascertaining overall prognosis. Herein we review the spectrum of pathology along the auditory pathway, extending from the labyrinth to the auditory cortex, that may present with sensorineural hearing.

Learning Objective: To recognize the imaging spectrum of sensorineural hearing loss in the pediatric and adult population.

INTRODUCTION

Sound waves reaching the ear are amplified and transmitted by the middle ear ossicles into the inner ear through the oval window. The sensory apparatus in the inner ear converts these sound waves into electrical impulses, which are then transmitted along the cochlear nerve to the brain stem and, subsequently, to the contralateral auditory cortex.[1,2]

SNHL is said to be present when the abnormality involves either the sensory (cochlea) or neural structures (cochlear nerve to the auditory cortex) and may be congenital or acquired.[1,3] Congenital SNHL is estimated to affect 1 in every 2000 neonates worldwide.[4]

The choice of initial imaging technique depends on the suspected etiology of SNHL. CT is preferred in cases of trauma or congenital dysplasias to map out osseous anatomy and the course of the facial nerve.[4,5] MR imaging is often preferred in cases of nontraumatic SNHL, otic capsule dysplasias, and retrocochlear lesions. However, combined imaging with both modalities is complementary and is being used with increasing frequency.[5-8]

The causes of SNHL may be divided, anatomically, on the basis of the location of the abnormality or, clinically, on the basis of the underlying etiology. An anatomic classification is preferred from a radiologic standpoint.[9]
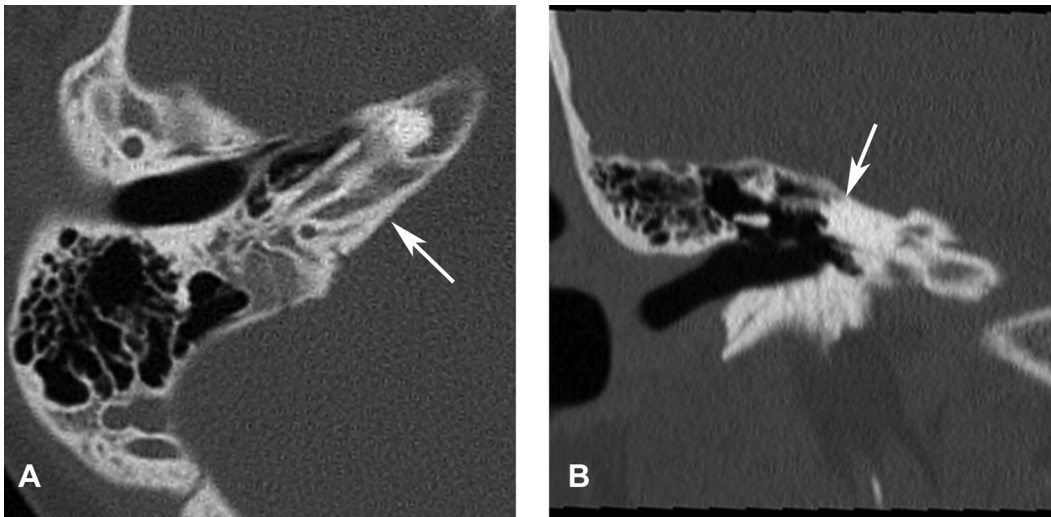
Fig 1. Complete labyrinthine aplasia. Axial CT image (*A*) with coronal reformation (*B*) shows complete absence of normal labyrinthine development (*white arrows*).
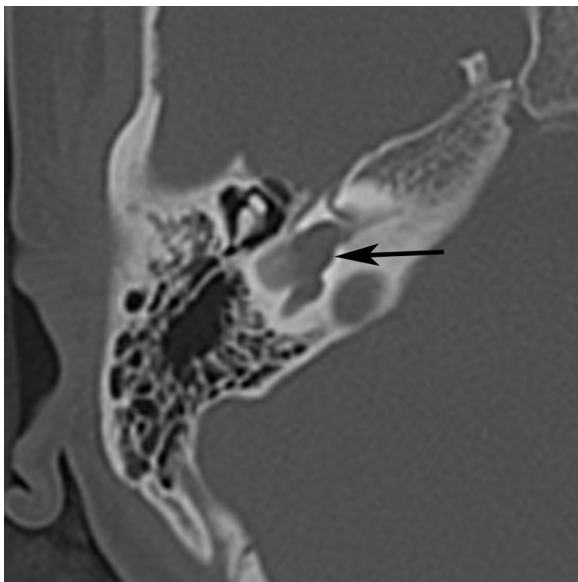

Fig 2. Common cavity anomaly. Axial CT image reveals absence of the normal cochlea and vestibule, which are replaced by a single common cavity (*black arrow*).
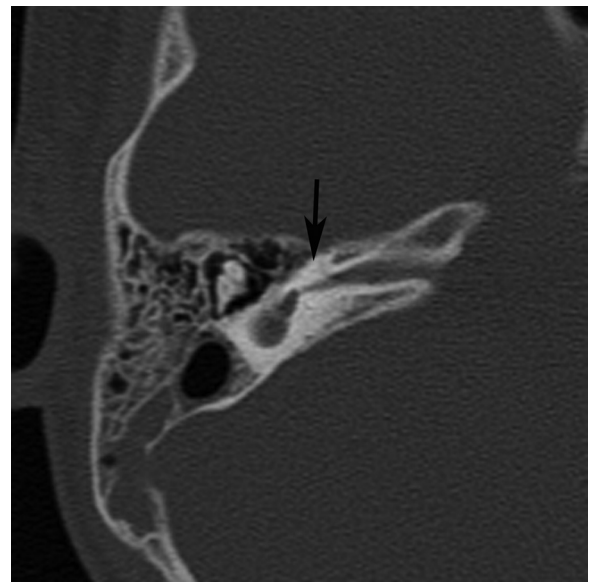

Fig 3. Cochlear aplasia. Axial CT image reveals complete absence of cochlear differentiation (*black arrow*). The vestibule is also dysplastic.

Congenital Lesions of the Labyrinth

Congenital lesions involving the labyrinth are together referred to as otic capsule dysplasias. These reflect an aberration of normal inner ear development that starts in the third week of gestation.[5,10] The cochlea and vestibule are completely formed by the eighth and 11th weeks of gestation, respectively, while the SCCs develop between weeks 19 and 22.[5] Since the embryologic development of the labyrinth takes place in a specific sequence, the timing of insults correlates well with the type of labyrinthine anomaly.[4,5,11]

Congenital anomalies vary from complete absence of inner ear structures to only mild abnormalities of cochlear partitioning.[8] On imaging, however, anomalies are seen in only approximately 20%–30% of patients with SNHL.[4,5,7,12]

Complete labyrinthine aplasia, also known as Michel's aplasia, results from arrest of otic placode development in week 3 of gestation and is extremely rare, representing <1% of all inner ear malformations.[1,4,5,11,12] Imaging shows an absence of inner ear structures. Additional findings may include IAC atresia, absence of the vestibulocochlear nerve (CN VIII), atresia of the round and oval windows, and ossicular dysplasia.[5,11,13] Hypoplasia of the temporal bone and middle ear may also occur (Fig 1).[5,13] The course of the facial nerve is often anomalous.[5,11] In addition, posterior fossa lesions, such as arachnoid cysts, may be present.[5]

The common cavity malformation results from arrested development during the fourth week of GA and accounts for approximately 25% of all cochlear malformations.[5,11] Imaging shows replacement of the cochlea and vestibule by

Fig 4. Type I incomplete partition anomaly. Axial CT images reveal a cystic appearance of the cochlea (*white arrow* in *A*) and vestibule (*black arrow* in *B*), which are separated by an incomplete bony partition (*white arrowhead* in *A*).
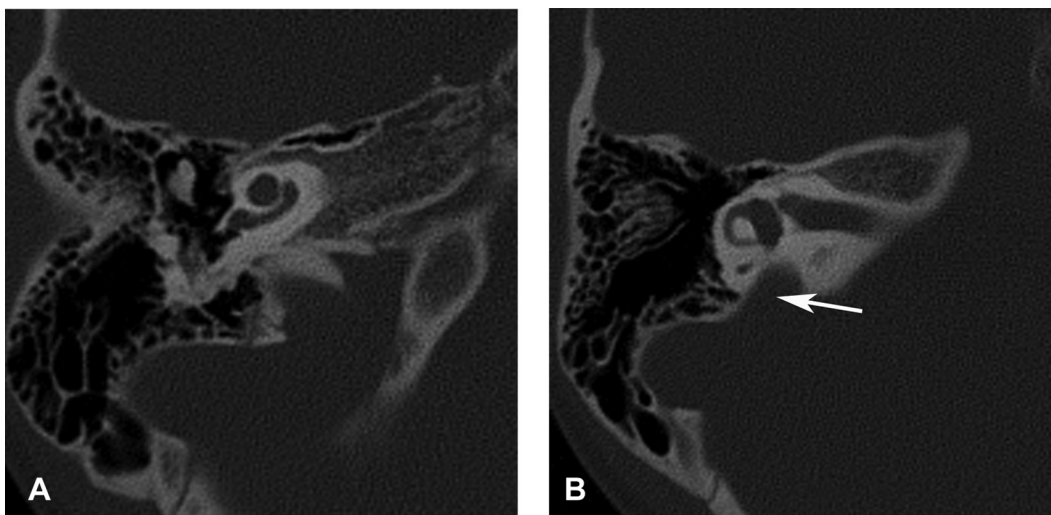

Fig 5. *A* and *B*, Mondini anomaly. Sequential CT images reveal a rounded appearance of the cochlear apex (*A*), along with EVAS (*white arrow in B*) and a mildly dysplastic vestibule.

a common cystic cavity with absent internal architecture (Fig 2).[5,11] The SSCs may be absent or malformed.[5,14]

Cochlear aplasia is also rare and represents about 3% of all cochlear anomalies.[5,11] It results from arrested development during the fifth week of GA (or the third week of GA, as described by some).[5,11] Imaging shows the absence of any cochlear framework. The vestibule and SCCs are often dysplastic (Fig 3).[5]

In the type I incomplete partition anomaly, also known as the cystic cochleovestibular malformation, the cochlear and vestibular sacs appear as a common cyst that is partly separated by a bony partition, giving rise to a "figure 8" appearance on imaging (Fig 4).[5,14] This anomaly results from developmental arrest in the fifth week of GA and accounts for approximately 6% of all cochlear malformations.[5] Modiolar deficiency often coexists, leading to transmission of intracerebral pressure to the cochlea.[11,14] Inser-

tion of a cochlear electrode in such cases leads to a sudden gush of endolymph out of the labyrinth, also referred to as a "gusher ear."[5,11]

Cochlear hypoplasia results from developmental arrest during the sixth week of GA and accounts for approximately 15% of all cochlear malformations.[5,15] The cochlea is small and may consist of a single turn or less.[5] Associated malformations of the vestibule and SCCs are again common.[5]

In the type II incomplete partition anomaly, commonly known as the Mondini deformity, developmental arrest occurs in the seventh week of GA.[5] It is, overall, the most common cochlear malformation (50%) and is associated with numerous teratogens and syndromes.[5,9] It was originally described by Mondini as a triad consisting of cochlear hypoplasia, enlarged vestibule, and a dilated vestibular aqueduct (Fig 5).[16] However, the originally described triad is
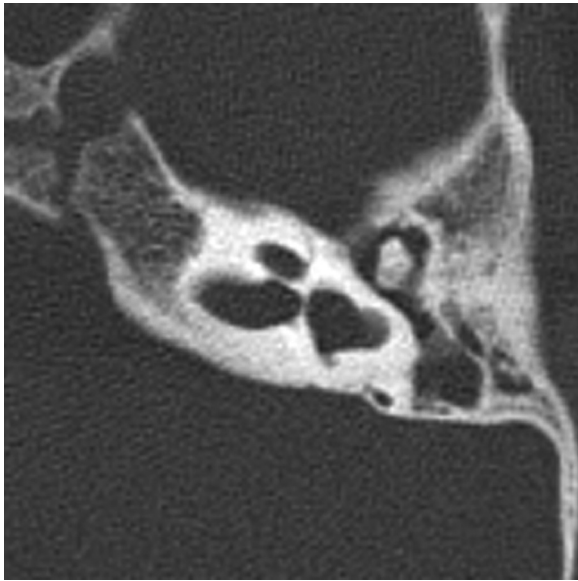
Fig 6. Lateral SCC-vestibular dysplasia. Axial CT image reveals the presence of a dysplastic vestibule with assimilation of the lateral SCC.

Fig 8. Pendred syndrome. Axial CT image reveals a modiolar deficiency (*black arrowhead*) and vestibular dysplasia (*black arrow*). Mild cochlear dysplasia is also present. Courtesy of Yutaka Sato, MD, University of Iowa hospitals and clinics.

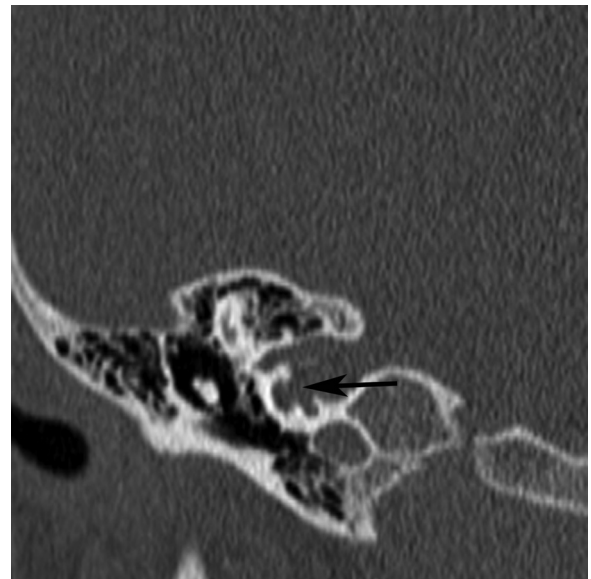**Fig 7. EVAS. Axial CT image reveals a dilated vestibular aqueduct (*white arrow*).**

Fig 9. X-linked SNHL. Coronal reformatted CT image reveals a modiolar deficiency (*black arrow*) along with a mildly dilated lateral IAC in a patient with X-linked SNHL.

seen in approximately 20% of cases.[11] On imaging, the cochlea consists of a normal basal turn with fusion of the middle and apical turns due to the absence of the interscalar septum and osseous spiral lamina.[5,11] Modiolar defects may also occur. Cochlear implantation is usually the therapy of choice.[11]

Congenital anomalies of the vestibule are frequently associated with other inner ear malformations and rarely occur in isolation. These may include dilatation, hypoplasia, or aplasia of the vestibule.[17]

Because the superior SCC develops first, followed by the posterior and lateral SCCs, isolated anomalies of the superior and posterior SCCs are rare.[4,5,11] The principal exceptions to this are Waardenburg and Alagille syndromes, both

of which may have isolated absence of the posterior SCC.[4,11] Partial or complete assimilation of the lateral SCC into the vestibule is the most common form of SCC dysplasia and is referred to as lateral SCC–vestibule dysplasia (Fig 6).[14] Unilateral dysplasias are often well-compensated, and the patient may be asymptomatic.[11]

EVAS is the most common anomaly in children with congenital SNHL.[4,11,18] It is often genetic and bilateral and is more common in females (3:2).[5,11] Patients often present with fluctuating SNHL, which worsens with barometric pressure changes and following trauma.[4,5,11] On imaging,
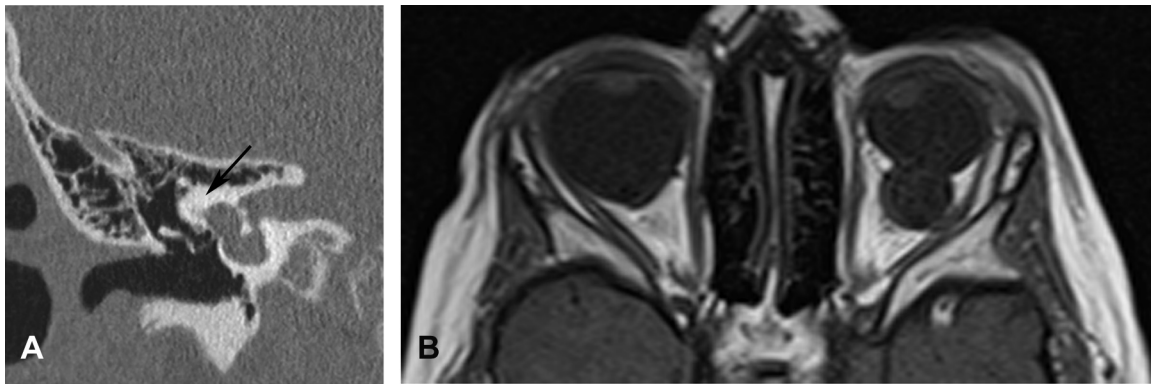
Fig 10. *A* and *B*, CHARGE syndrome. Coronal reformatted CT image (*A*) shows absence of the lateral and superior SCCs. The posterior SCC was also absent (not shown). Axial T1-weighted image (*B*) through the orbits reveals bilateral colobomas, better appreciated on left side.
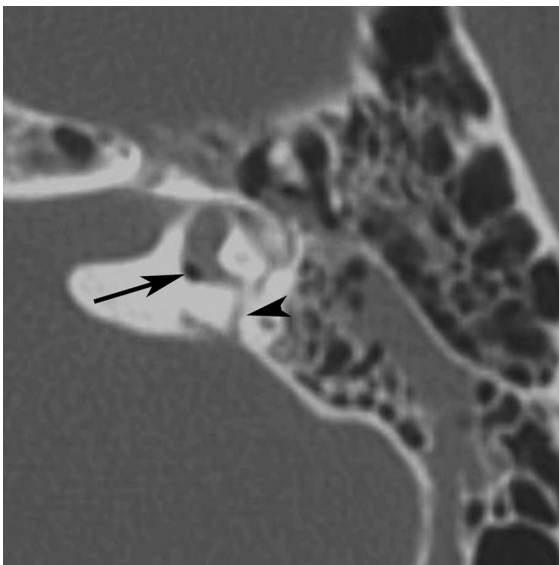

Fig 11. Labyrinthine trauma. There is a fracture line extending across the vestibule (*arrowhead*) along with a pneumolabyrinth (*arrow*).
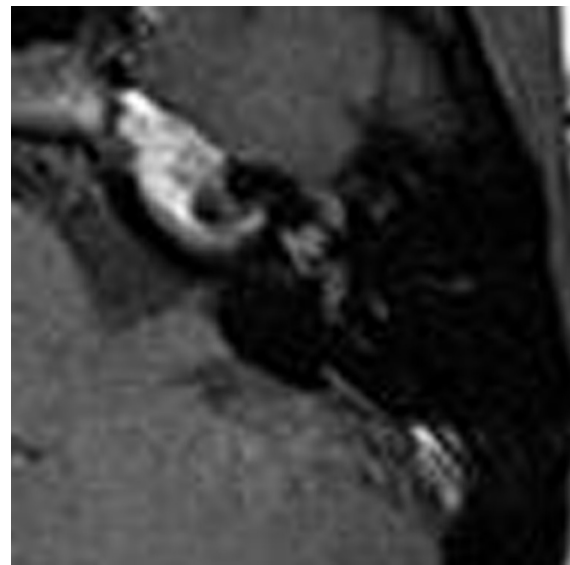

Fig 12. Labyrinthine hemorrhage. Axial T1-weighted imaging reveals diffuse labyrinthine T1 shortening, in keeping with hemorrhage.

the vestibular aqueduct is said to be enlarged if the diameter exceeds 1.5 mm at its midpoint (Fig 7).[4,5,11] Focal dilatation of the distal aqueduct, on the other hand, is a normal variant and is usually asymptomatic.[11] Patients with EVAS often show associated cochlear and vestibular anomalies of varying severity.[1,4,5] Absence of the modiolus and enlargement of the vestibule are the most common cochlear and vestibular anomalies, respectively.[18]

At times, congenital abnormalities of the inner ear may occur as part of a syndrome. It is estimated that approximately 50% of children with SNHL have a genetic cause and approximately a third of these may be associated with a distinct syndrome.[1,3,4] A complete review of imaging in these numerous syndromes is beyond the scope of this text. However, imaging in some of the more common syndromic causes of SNHL is described below.

Pendred syndrome is believed to be the most common one associated with congenital SNHL.[15,19] It shows an autosomal recessive inheritance and manifests as SNHL in association with euthyroid goiter.[15] On imaging, modiolar
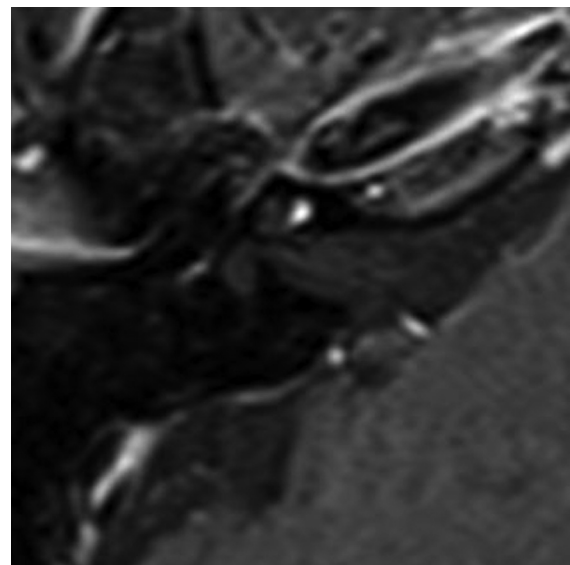

Fig 13. Cochlear schwannoma. Postcontrast axial T1-weighted, fat-suppressed image reveals focal enhancement involving the basal turn of the cochlea.
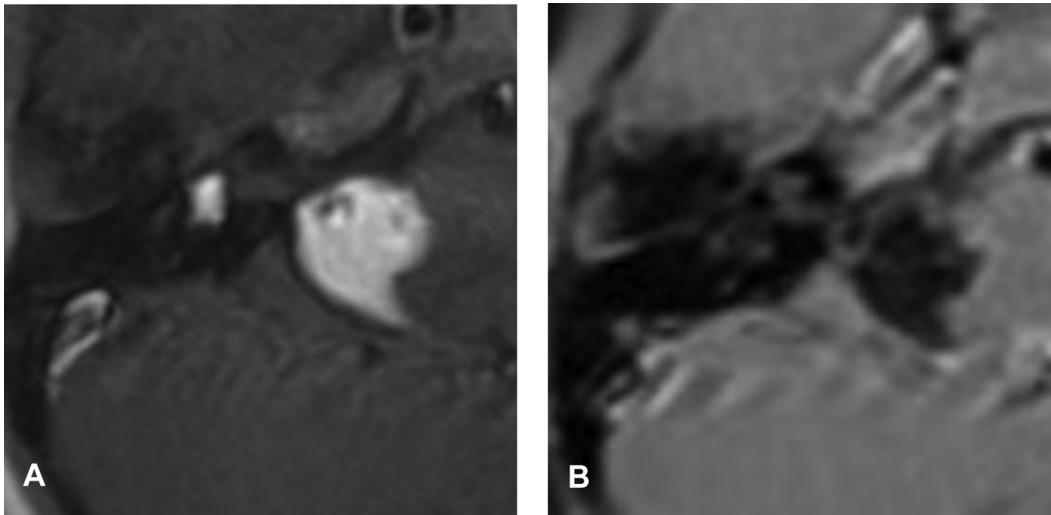
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Fig 14. Vestibular and CPA lipomas. Axial T1-weighted images, without (*A*) and with (*B*) fat suppression, reveal soft-tissue masses in the right vestibule and CPA, with signal-intensity characteristics consistent with fat.
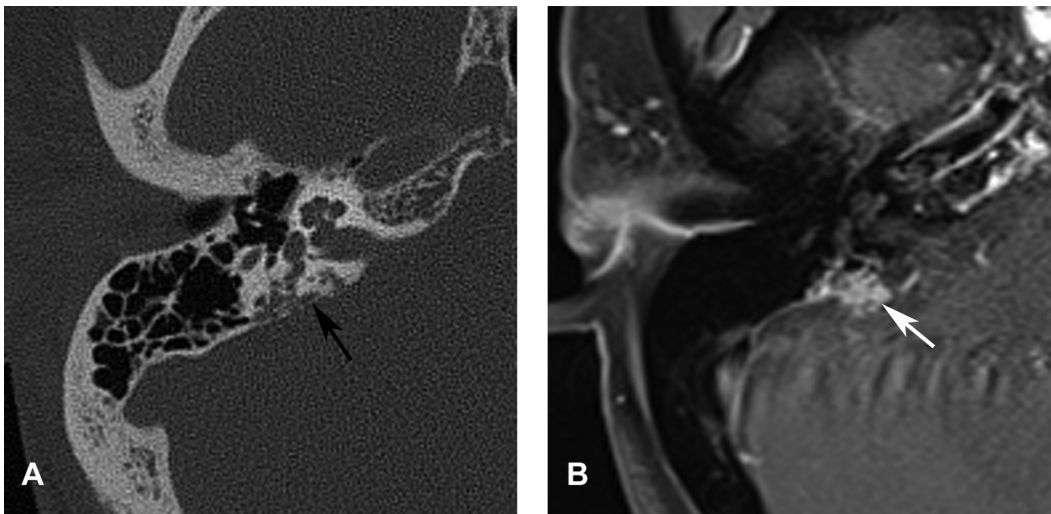

Fig 15. ELST. Axial CT (*A*) and axial T1-weighted postcontrast MR images (*B*) reveal the presence of a destructive (*black arrow* in *A*) and enhancing (*white arrow* in *B*) retrolabyrinthine mass.

deficiency and vestibular enlargement are universally seen. Other frequent findings include fusion of the middle and upper cochlear turns and a dilated vestibular aqueduct (Fig 8).[15,19]

Patients with X-linked SNHL may present with congenital or progressive, mixed or purely sensorineural hearing loss.[9,15] It predominantly affects males while female carriers may manifest only minimal symptoms.[9,15] Imaging shows dilatation of the lateral IAC, absence or severe hypoplasia of the modiolus, and hypoplasia of the cochlear base.[15] Dilatation of the labyrinthine segment of the facial nerve canal and vestibular aqueduct may also be seen (Fig 9).[9]

In CHARGE syndrome, there is SCC aplasia and vestibular dysplasia associated with colobomas, heart defects, choanal atresia, and growth retardation (Fig 10).[11,15] Cochlear partitioning defects and CND may occur.[15] Associated anomalies of the middle ear, including atresia of the

oval and round windows and ossicular chain dysplasias, can coexist.[15]

Acquired Lesions of the Labyrinth

Acquired labyrinthine causes of SNHL include trauma, labyrinthine masses, and inflammation. Fractures that violate the otic capsule are up to 7 times more likely to cause severe hearing loss compared with fractures that spare the otic capsule.[20] CT may show a fracture line, perilymphatic fistula, or pneumolabyrinth (Fig 11).[12,15] Patients with post-traumatic SNHL without fracture occasionally benefit from MR imaging, which may demonstrate cochlear hemorrhage or cochlear nerve injury.[15]

Labyrinthine hemorrhage may occur in association with coagulopathies, ELST, trauma, or labyrinthitis.[11,12] It may also occur after stapes or vestibular schwannoma surgery. MR imaging shows T1 shortening in the acute phase (Fig 12).[11,12]
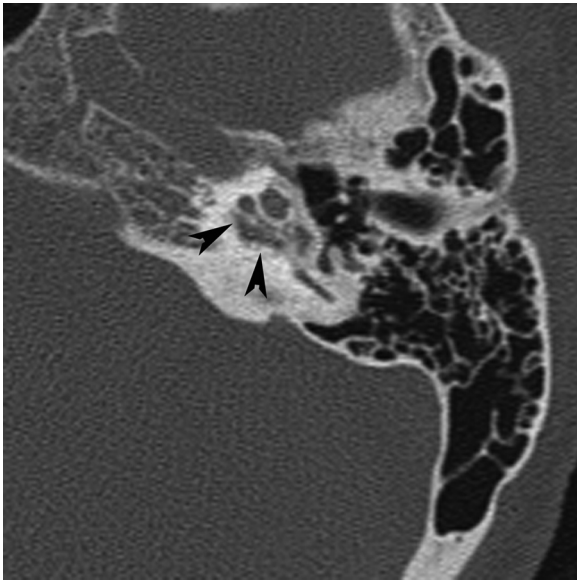
Fig 16. Retrofenestral otospongiosis. Axial CT image reveals the presence of left perilabyrinthine demineralization (*black arrowheads*). Similar findings were also present on the right (not shown).
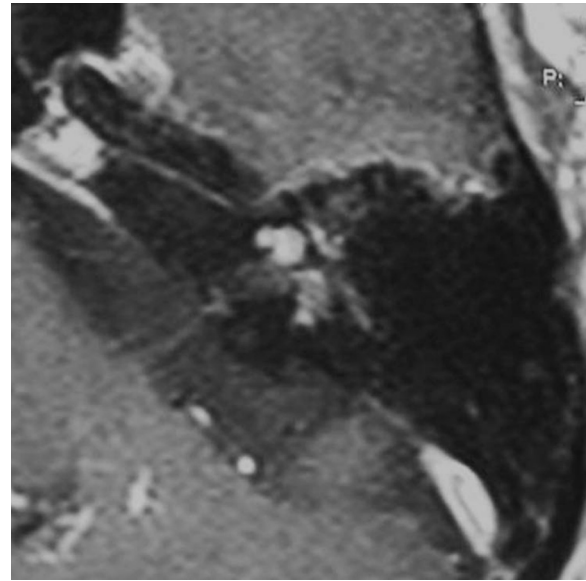

Fig 17. Acute labyrinthitis. Postcontrast axial T1-weighted image reveals the presence of diffuse labyrinthine enhancement.

Primary labyrinthine masses include schwannomas, lipomas, and ELST. Labyrinthine schwannomas most frequently involve the cochlea, though they may involve any or all segments of the labyrinth.[8] On imaging, these lesions may show slight T1 shortening and appear as a filling defect on heavily T2-weighted images.[8] They often enhance intensely (Fig 13).[11,12] Intravestibular lipomas are rare and show fat signal intensity on imaging.[11,21] They are often associated with CPA lipomas (Fig 14).[21] ELSTs are commonly sporadic but may be associated with von Hippel-Lindau disease.[11,22] Imaging shows a hemorrhagic, enhancing and destructive retrolabyrinthine mass (Fig 15).[9,11,14]

Secondary labyrinthine involvement may occur with masses of the middle ear cavity and skull base. Langerhans cell histiocytosis rarely involves the inner ear but may cause irreversible SNHL. Imaging shows an enhancing, destructive mass in a young patient.[23] Similarly, paragangliomas, metastatic deposits, and cholesteatomas may also involve the labyrinth.[11]

Otospongiosis (also known as otosclerosis) most commonly affects young females and is often bilateral (80%).[11] The fenestral subtype is more common and involves the promontory and oval window. Cochlear otospongiosis (retrofenestral subtype) affects the middle and basal cochlear turns and often occurs together with the fenestral subtype.[12,14] Cochlear otospongiosis may demonstrate a double ring sign secondary to perilabyrinthine demineralization on CT (Fig 16).[9] On MR imaging, foci of demineralization show T2 prolongation and enhance following contrast.[11,14]

Labyrinthitis is often viral in origin and self-limiting.[11,15] Bacterial labyrinthitis, on the other hand, is usually secondary to meningitis and is the most common

Fig 18. Labyrinthitis ossificans. Axial CT image in a child with SNHL reveals calcific obliteration of the left cochlea (*black arrowheads*).

cause of acquired SNHL in children.[11,15] Other causes of labyrinthitis include syphilis, autoimmune disorders, and radiation therapy.[8,11] The imaging hallmark of all types of acute labyrinthitis is membranous labyrinth enhancement (Fig 17).[11,15]

Labyrinthitis obliterans is usually secondary to meningitis. Other causes include trauma and surgery.[1] Acute labyrinthine inflammation in these cases leads to formation of granulation tissue and progressive fibrotic obliteration of the labyrinth. During this stage, imaging may only show loss of fluid signal intensity on heavily T2-weighted MR images without any obvious finding on CT. With time, metaplastic bone formation ensues and appears as labyrinthine calcification on CT (Fig 18). MR imaging in these
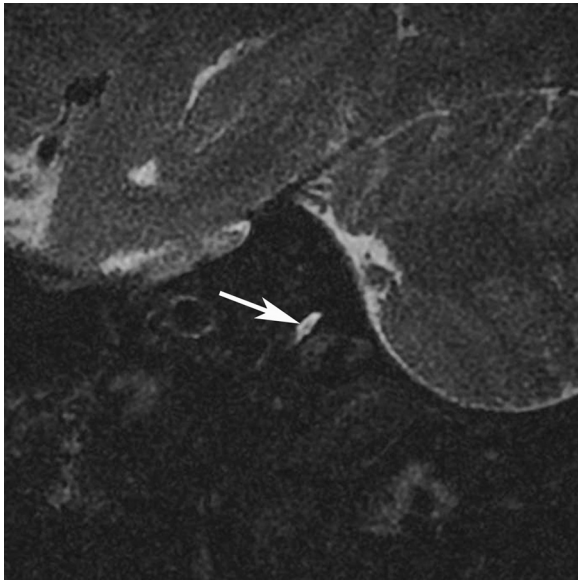
Fig 19. CN VIII aplasia. Oblique sagittal T2-weighted image in a patient with severe SNHL reveals a stenotic IAC and a single nerve (*white arrow*), which was the facial nerve in this case.
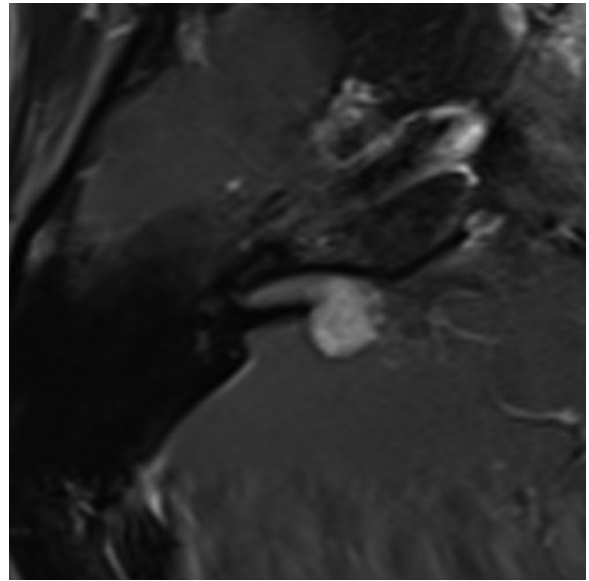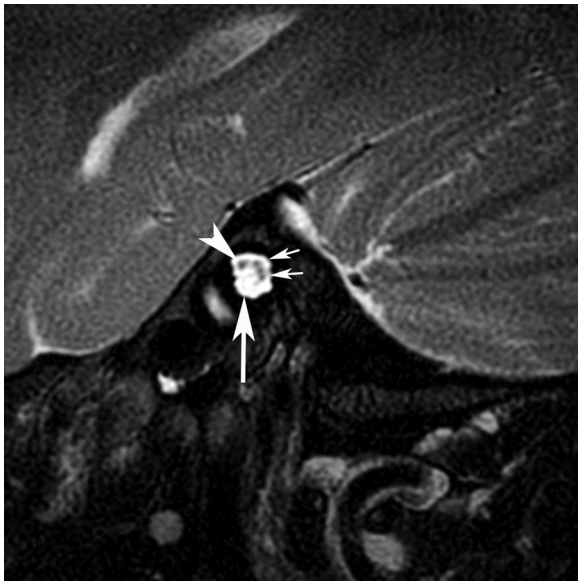

Fig 20. Oblique sagittal T2-weighted image in another patient reveals the absence of the cochlear nerve in the lower anterior quadrant (*white arrow*). The facial (*white arrowhead*), superior, and inferior vestibular nerves (*small arrows*) are visualized.

cases again reveals loss of fluid signal intensity on heavily T2-weighted images.[5,15]

Lesions Involving the IAC and CPA

CND encompasses both aplasia and hypoplasia and may be seen in up to 18% of patients with SNHL.[4] It is often congenital but may be acquired, secondary to infection or trauma.[4,24] Patients with type 1 aplasia have absence of CN VIII with stenosis of the IAC. In type 2 aplasia, there is a common CN VIII with aplasia or hypoplasia of the cochlear nerve component. Labyrinthine malformations may (type

Fig 21. Vestibular schwannoma. Axial T1-weighted postcontrast image reveals the typical "ice cream cone" appearance of the mass.

2A) or may not (type 2B) be present.[5,10] MR imaging demonstrates absence of the cochlear nerve,[4,11] and CT may show an absent or stenotic cochlear aperture.[3,4,8] Some patients may also have associated stenosis of the IAC, defined as a canal diameter <2 mm (Fig 19).[5,7,25] IAC stenosis is, however, an unreliable marker of CND because patients with a normal IAC can have cochlear nerve aplasia and vice versa (Fig 20).[5,26] Additionally, patients with bilateral CND often have associated labyrinthine and hindbrain malformations, with the latter being seen in up to 40% of patients.[25]

A normal cochlear nerve is similar in size or larger than the accompanying facial nerve in approximately 64% of cases.[24] It is considered hypoplastic when it is smaller than other nerves in the ipsilateral IAC or the normal contralateral cochlear nerve.[25]

Vestibular schwannomas are the most common CPA masses and often arise in Scarpa's ganglion.[1,12] Up to 20% of these patients may present with acute SNHL.[1] On imaging, these typically show T1/T2 prolongation and postcontrast enhancement.[1] Involvement of both the IAC and CPA may give rise to the "ice cream cone" appearance (Fig 21).[8] Widening of the porous acusticus of the IAC often occurs in larger lesions. Patients with bilateral tumors, by definition, have neurofibromatosis type II.[1,8,15] Extension of the lesion to the IAC fundus or abnormal fluid signal intensity between the mass and the cochlea portend poor hearing outcome following resection.

Meningiomas, when they involve the CPA or porus acusticus, may result in SNHL and mimic schwannomas.[1] However, they are often eccentric with respect to the porus acusticus.[8] The presence of calcifications and hyperostosis of underlying bone also favor meningioma.[1,8] IAC lipomas are rare but easily diagnosed on T1-weighted images with and without fat suppression (Fig 22).[8] Other
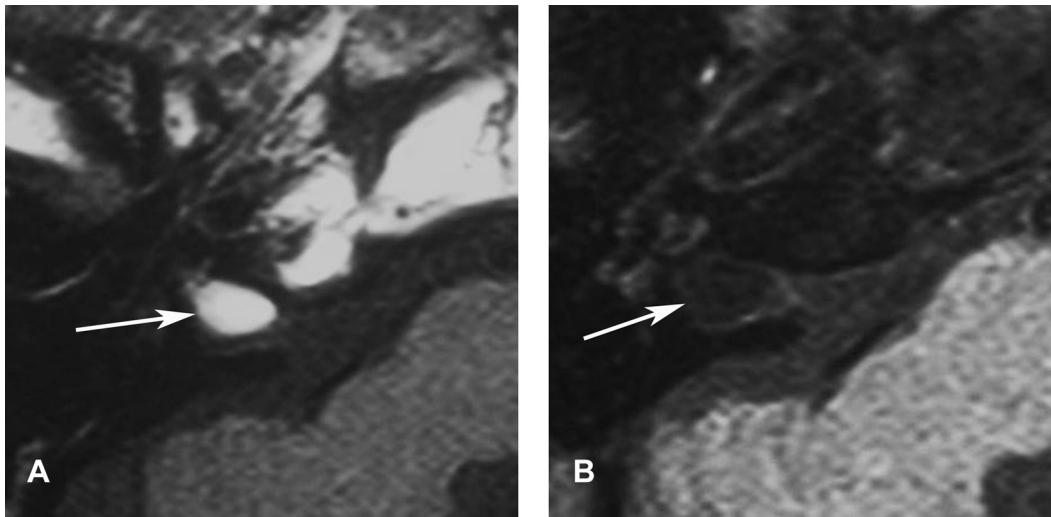
Fig 22. IAC lipoma. Axial T1-weighted images, without (*A*) and with (*B*) fat suppression, reveal a small lipoma within the IAC (*white arrows* in *A* and *B*).
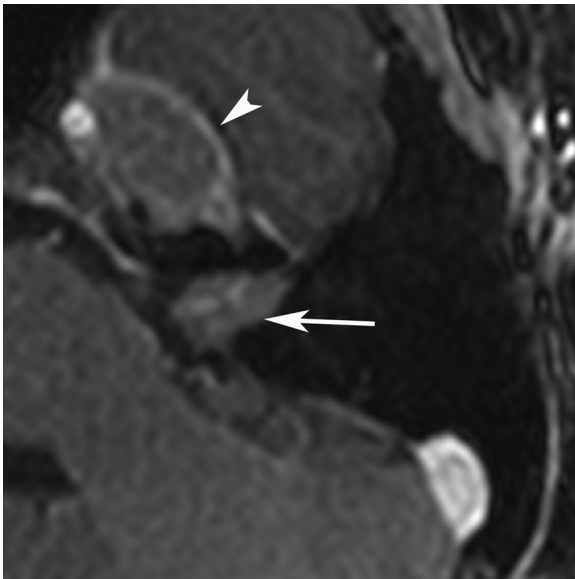

Fig 23. Metastases. Axial T1-weighted postcontrast image in a child with a pineal germ cell tumor and multiple cranial nerve palsies reveals metastatic deposits involving the left IAC (*white arrow*) and Meckel's cave (*white arrowhead*).
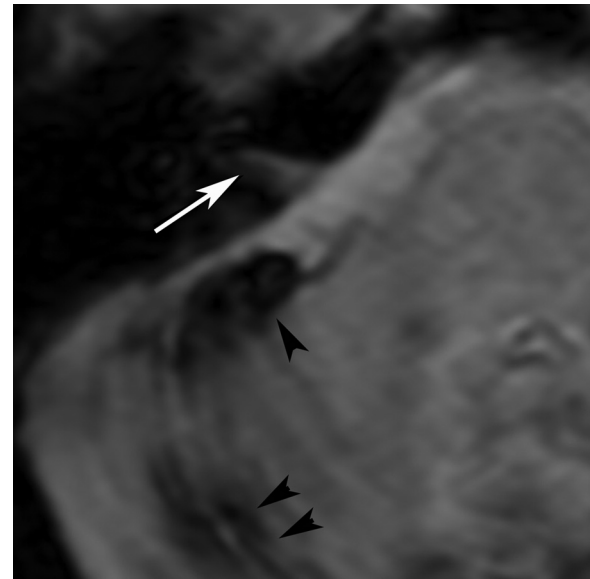

Fig 24. Superficial siderosis. Axial GRE image reveals hypointense signal intensity along the right eighth nerve (*white arrow*). Changes of superficial siderosis are also noted in the posterior fossa (*black arrowheads*).

CPA lesions associated with SNHL include metastases, dermoid and epidermoid cysts, arachnoid cysts, and congenital cholesteatomas.[1,8,15]

SNHL may also occur from cochlear nerve involvement secondary to infectious and inflammatory or neoplastic meningeal conditions (Fig 23).[1,8] SNHL in these patients is rarely the first or an isolated presentation.[8]

Siderosis of CN VIII may follow subarachnoid hemorrhage.[1,12] Patients often present with progressive gait ataxia and SNHL.[27] The nerves show peripheral low signal intensity on T2-weighted/GRE/susceptibility-weighted images (Fig 24).[12,27] Additionally, neurovascular conflicts involving CN VIII may present with fluctuating SNHL, though this is considered controversial by some authors.[1,12]

Lesions Involving the Brain Stem and Auditory Pathways

The cochlear nerve enters the brain stem at the pontomedullary junction and relays signals to the ventral and dorsal cochlear nuclei. Most of the fibers from here cross the midline through the trapezoid body and pass to the superior olivary complex. Fibers then extend cephalad through the lateral lemniscus, inferior colliculus, and medial geniculate body, finally reaching the contralateral superior temporal lobe gyrus.[1,23]

Infarctions are the most frequent lesions of the auditory pathway associated with SNHL. These show restricted diffusion in the acute stage.[23] Multiple sclerosis plaques can also affect the auditory pathway, and imaging often demonstrates additional periventricular and callosal involvement (Fig 25).[14] Brain stem tumors (primarily gliomas),
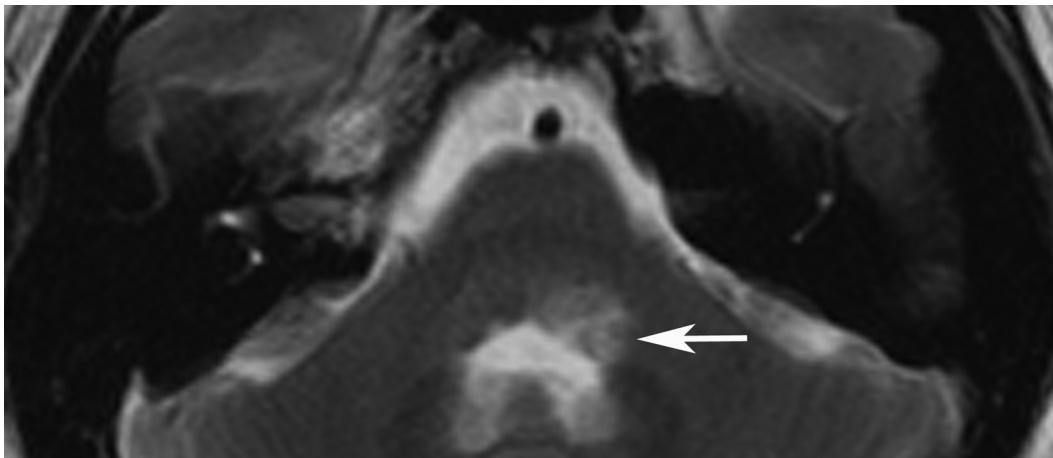
Fig 25. Demyelination. Axial T2-weighted image in a patient with multiple sclerosis reveals the presence of a demyelinating plaque in the expected location of the left eighth nerve nuclei (*white arrow*). The lesion responded to steroid therapy (not shown).
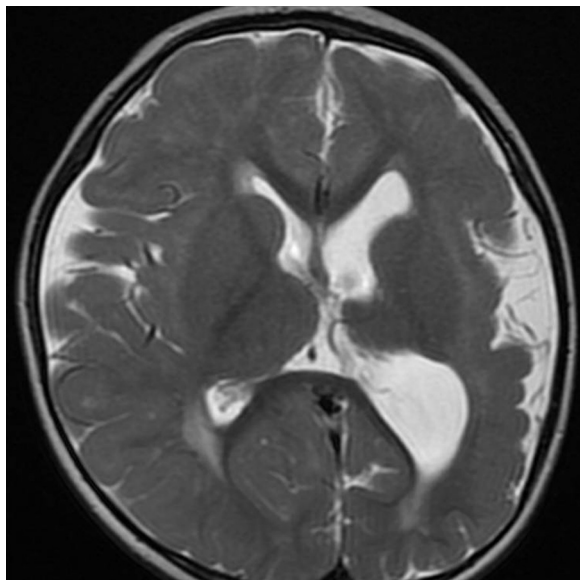

Fig 26. Congenital CMV. Axial T2-weighted image in a child with congenital CMV and bilateral SNHL. There are bilateral periventricular white matter lesions along with volume loss and pachygyria involving the left cerebral hemisphere.

metastases, and lymphomas can also cause SNHL, but involvement of adjacent brain stem structures causes additional symptoms.[23]

Sensorineural hearing loss affects 10%–15% of neonates with congenital CMV infection who are symptomatic at birth.[5] The inner ear structures in these patients are often unremarkable on imaging.[4,28] A large percentage of these children also show parenchymal volume loss, white matter lesions, and gyral abnormalities (Fig 26).[5,28]

CONCLUSIONS

A wide spectrum of pathologies, both congenital and acquired, may be associated with SNHL. Lesions may involve the cochlea or the retrocochlear apparatus. Imaging is indispensable to discern the underlying etiology and its extent and to predict overall outcome.

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