Imaging of congenital inner ear malformations

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Learning objectives

Congenital sensorineural hearing loss arises as a result of abnormalities in the inner ear, the vestibulocochlear nerve, or the processing centers of the brain. The abnormality may have a genetic cause or be a sequela of infection or injury at birth; in some cases, no cause is identified. Congenital inner ear abnormality is the major cause.

High-resolution computed tomography (CT) imaging of the temporal bones allow excellent depiction of inner ear malformations and is routinely used in the evaluation of pediatric sensorineural hearing loss.

This article illustrates the computed tomography (CT) findings associated with various congenital inner ear malformations. We propose to attend these objectives:

- Description and illustration of the congenital abnormalities of the inner ear on high resolution CT temporal bone images and correlation with developmental arrest during embryology.

- Recognize the high resolution computed tomography (HRCT) findings associated with various congenital inner ear malformations.
Background

Globally, hearing impairment occurs in about 1 to 2 per 1000 live births. As hearing loss has adverse effects on language, cognition, behavioral and socioemotional development, it is crucial to recognize this problem promptly.

The etiology of profound hearing loss in children is complex and multifactorial. It is divided into two main causes: environmental and genetic.

Environmental causes include viral infections (toxoplasma, rubella, cytomegalovirus and herpes simplex virus), bacterial meningitis, prematurity and foetal exposure to isoretinoin. Genetic errors may be either autosomal dominant or recessive and may manifest as sensorineural hearing loss alone or be associated with any of a number of syndromes (Alport, Pendred, Waardenburg, CHARGE, branchio-oto-renal and X-linked progressive hearing loss with perilymphatic gusher).

For optimal interpretation of high-resolution CT obtained in children with congenital sensorineural hearing loss, the radiologist must have a comprehensive knowledge of the normal anatomy and the embryologic development of inner ear structures as well as the spectrum of malformations that may be encountered.

1-Normal anatomy of inner ear (Figures 1 and 2):

The inner ear consists of an osseous labyrinth that encloses a membranous labyrinth. The osseous labyrinth consists of the vestibule, cochlea, semicircular canals, vestibular aqueduct, and cochlear aqueduct. The space between the osseous labyrinth and membranous labyrinth is filled with a fluid known as perilymph. A similar fluid (endolymph) fills the space within the membranous labyrinth.

The vestibule is the central, rounded portion of the osseous labyrinth. It is continuous anteroinferiorly with the cochlea and posteriorly with the semicircular canals and the vestibular aqueduct. The vestibule contains the utricle and the saccule.

There are three semicircular canals; these are designated as the superior, lateral, and posterior semicircular canals.

The cochlea consists of a canal that spirals 2½ to 2¾ times around a central column of bone (the modiolus). The cochlear aperture, also known as the cochlear nerve canal, is a small opening at the fundus of the IAC through which the cochlear nerve enters the cochlea.

The IAC extends from the labyrinth to the cerebellopontine angle and contains the seventh and eighth cranial nerves. The eighth nerve (vestibulocochlear nerve) is composed of three branches: The superior and inferior vestibular branches of the nerve
and the cochlear branch. A normal cochlear branch of the eighth nerve should have approximately the same diameter as the facial nerve.

2-Embryology (Figures 3 and 4)

At approximately the third week of gestation, otic placodes arise from the surface ectoderm on each side of the rhombencephalon. The otic placodes subsequently invaginate and form otocysts, which are the otic and auditory vesicles. At around the fifth week, diverticulum buds from the otocysts form the endolymphatic sacs and ventral pouches. By the end of the 8th week, the morphology of the labyrinth is generally identifiable. The otic capsule develops as a cartilaginous condensation of the mesenchyme from the 4th week onwards, and subsequently grows and ossifies to form the bony labyrinth. Bony labyrinthine development occurs primarily between the 4th and 8th gestational weeks, and as it continues ossification ensues between the 8th and 24th gestational weeks. In general, the saccule, endolymphatic duct and utricle are completed by the 11th week, followed by the cochleas and vestibules. The membranous cochlea achieves 1 to 1.5 turns at the end of 6 weeks, and 2.5 turns are formed at the end of the 7th week. Development of sensory epithelium including the organ of Corti occurs simultaneously with the growth and ossification of the bony labyrinth between the 8th and 24th gestational weeks.

The semicircular canals start to develop from the utricle segments of the otocysts at 7-8 gestational weeks. The superior canals form first, followed by the posterior and then the lateral canals.

That's why, when the insults occur early during the 4th to 8th gestational week, cochlear deformity results. By contrast, injury ensuing after the 8th week results in malformation of the vestibule and semicircular canals, while the cochlea is preserved.

The middle and external ear have a separate embryological origin from that of the inner ear. This explains the low frequency (about 10%) of associated middle or external ear anomalies in inner ear dysplasia.

3- Classification:

In membranous malformations, the classification is based on the histopathologic changes in the inner ear, whereas in the combined osseous/membranous deformities, radiographic appearance is used to distinguish among the various entities.

I. Malformations limited to the membranous labyrinth

A. Complete membranous labyrinthine dysplasia

B. Limited membranous labyrinthine dysplasia
1. Cochleosaccular dysplasia

2. Cochlear basal turn dysplasia

II. Malformations of the osseous and membranous labyrinth

A. Complete labyrinthine aplasia (Michel)

B. Cochlear anomalies

1. Cochlear aplasia

2. Common cavity

3. Cochlear hypoplasia

4. Incomplete partition I

5. Incomplete partition II (Mondini)

C. Labyrinthine anomalies

1. Semicircular canal dysplasia

2. Semicircular canal aplasia

D. Aqueductal anomalies

1. Enlargement of the vestibular aqueduct

2. Enlargement of the cochlear aqueduct

E. Internal auditory canal abnormalities

1. Stenotic internal auditory canal

2. Enlarged internal auditory canal.

4- CT Examinations

High-resolution CT of the temporal bone, followed by image reconstruction in both the axial and coronal planes, is required to evaluate the inner ear and its malformations. Axial scanning is performed in planes parallel to the infraorbitomeatal line. On a multidetector CT scanner, the raw axial image data set can be reconstructed with a section thickness of as little as 0.3 mm to obtain high-quality coronal reformatted images. A 512 × 512 matrix is used, and all the images are reviewed with a high-resolution bone algorithm and a small field of view (9 cm) for separate documentation of the right and left ears. Axial
images are obtained from the top of the petrous apex to the inferior tip of the mastoid bone. Coronal reformatted images are obtained from the anterior margin of the petrous apex to the posterior margin of the mastoid.
Images for this section:

**Fig. 1:** Drawing shows the normal anatomy of the inner ear: the cochlea (C); vestibule (V); superior semi–circular canal (S scc); posterior semicircular canal (P scc); lateral semicircular canal (L scc); and vestibular aque-duct, which consists of an endolymphatic duct (ED) and endolymphatic sac (ES). Note the locations of the oval window (OW) and round window (RW).

Fig. 3: drawings show the different stages of the inner ear embryologic development

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Fig. 4: Diagram shows principal congenital inner ear deformities occurring during the embryologic development

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Findings and procedure details

1. **I. Malformations limited to the membranous labyrinth**

The classification of these abnormalities is not clinically useful, as their differentiation requires histopathologic examination.

**A. Complete membranous labyrinthine dysplasia**

Complete membranous labyrinthine dysplasia is extremely rare. It has been reported in association with the cardioauditory and Usher's syndromes. Patient with this disorder has a normal CT.

**B. Limited membranous labyrinthine dysplasia**

1. **Cochleosaccular dysplasia (Scheibe)**

Incomplete development of the pars inferior is the most frequent histopathologic finding in congenital deafness, commonly known as cochleosaccular dysplasia. The organ of Corti is either partially or completely missing. The cochlear duct is usually collapsed, with Reissner's membrane adherent to the limbus. Less commonly it is distended, presumably as a result of endolymphatic hydrops. The stria vascularis is typically degenerated and may contain colloidal inclusions, describes characteristic strial changes consisting of aplasia alternating with regions of hyperplasia and gross deformity. Cochlear changes may be severe in the base turn and gradually lessen in intensity towards the apex, or they may be severe throughout. The saccule is usually collapsed and has degenerated sensory epithelium.

In cochleosaccular dysplasia, the SCCs and utricle are normal. Auditory neuronal survival is variable, but may remain normal into adulthood, at least in some cases.

2. **Cochlear basal turn dysplasia (Alexander)**

Dysplasia limited to the basal turn of the cochlea may be related to familial high frequency SNHL. No descriptions of membranous labyrinthine dysplasia limited to the pars superior was found in an extensive review of the literature. This outcome is not surprising as such individuals probably are asymptomatic. They would have normal hearing and, presumably, would have compensated for their congenital vestibular deficit.

1. **II. Malformations of the osseous and membranous labyrinth**

Congenital anomalies of the inner ear that deform the otic capsule are of special interest to the clinician, as they may be recognized during life through radiographic imaging.
1. **A. Complete labyrinthine aplasia (Michel):** figure 5

It is a rare congenital inner ear abnormality (1% of cochlear bony Malformations). This condition is the severest deformity of the membranous and osseous labyrinth, defined as the absence of inner ear structures and is caused by developmental arrest of otic placode early during the third week of gestational age. In complete labyrinthine aplasia, the otic capsule is entirely absent. Such ears are of course uniformly deaf.

A narrow, atretic IAC is seen on high-resolution CT images due to the absence of the eighth cranial nerve (not visualized on MR images). These abnormalities may be unilateral or bilateral. In patients with unilateral complete labyrinthine aplasia, the contralateral inner ear structures are often dysplastic.

Multiple associated abnormalities have been described in structures arising from the otic capsule.

**B. Cochlear anomalies**

1. **Cochlear aplasia:** (figure 6)

It is a rare anomaly, accounting for only 3% of cochlear malformations. In this deformity, the cochlea is completely absent, presumably as a result of an arrest in the development of the cochlear bud, most likely occurring in the latter part of the 3rd week of gestation or, as described by some, in the 5th week.

The vestibule and semicircular canals are often malformed (dilated or hypoplastic) but may be normal. It is necessarily to differentiate this anomaly from labyrinthine ossification in which a normal-sized bone is seen anterior to the IAC, with the bulge of the cochlear promontory produced by the basal turn of the cochlea.

2. **Common cavity** (figure 7)

The developmental arrest occurs at the fourth week of gestation and accounts for about 25% of all cochlear malformations. It is a deformity in which the cochlea and vestibule are confluent, forming an ovoid cystic space without internal architecture. The width of the cavity is typically greater than its height, with the average vertical diameter being 7 mm, and the average horizontal diameter, 10 mm. The semicircular canals are frequently malformed but occasionally normal.

3. **Incomplete partition I (cystic cochleovestibular malformation)** (figure 8)

It results from a developmental arrest in the 5th week of gestation. It entails two separated cystic cavities that represent the rudimentary cochlea (no bony modiolus) and vestibule (dilated), and assumes a 'figure of 8' or 'snowman' appearance. The vestibular aqueduct is normal. The cribriform area between the cochlea and IAC is often defective, and all
patients have a large IAC predisposing them to increased risks for meningitis and for a perilymphatic gusher in the event of surgery.

4. **Cochlear hypoplasia** (figure 9)

This deformity comprises approximately 15% of all cochlear anomalies. It is the result of an aberration in the development of the cochlear duct during the sixth week. The cochlea and vestibule can be differentiated from each other but the cochlea is consisting of a small bud of variable length (usually 1 to 3 mm) protrudes from the Vestibule and makes a single turn or less. The vestibule and semicircular canals are usually malformed but may be normal.

5. **Incomplete partition II (Mondini)** (figure 10)

This is the most common type of cochlear malformation, accounting for over 50% of all cochlear deformities. Developmental arrest occurs at the seventh week of gestation. The cochlea consists of 1½ turns, and the intercalar septum and osseous spiral lamina are absent. The basal cochlear turn is intact, but the middle and apical turns coalesce to form a cystic apex resulting in a baseball cap-like cochlear apex. The modiolus is present only at the level of the basal turn. This malformation has been described as a triad consisting of a cochlea with a normal basal turn and cystic apex, enlarged vestibular aqueduct and vestibule, and normal semicircular canals.

C. **Labyrinthine anomalies**

1. **Semicircular canal dysplasia** (figure 11)

The lateral SCC is deformed more often than the posterior or superior SCC, apparently because it forms earlier in embryogenesis (malformation of the superior and posterior semicircular canals without involvement of the lateral canal is unusual). Dysplasia of the lateral SCC is a common type of inner ear malformation. Approximately 40% of ears with a malformed cochlea will have an accompanying dysplasia of the lateral SCC. Occasionally, dysplasia of the lateral SCC exists as the sole inner ear malformation. Isolated lack of development of posterior semicircular canal is implicated in the Alagille and Waardenburg The malformed canals are usually short and wide but may be narrow. In extensive malformations, the vestibule is dilated and forms a common lumen with the lateral canal syndromes.

2. **Semicircular canal aplasia** (figure 12)

SCC aplasia is far less common than dysplasia. It refers to the absence of a semicircular canal. Presumably, it arises from a failure in the development of the vestibular anlage before the sixth week. It is usually associated with cochlear anomalies. An abnormal course of the facial nerve, atresia of the oval window, and abnormal ossicles are
frequently seen in children with aplasia of the canals. This condition is also one of the manifestations of the so-called CHARGE syndrome.

D. Aqueductal anomalies

1. Enlargement of the vestibular aqueduct *(figure 13)*

It is the most common imaging abnormality in children with congenital sensorineural hearing loss and is also commonly associated with other inner ear abnormalities. It is regarded as enlarged if the size of the mid portion is greater than 1.5 mm (measured at the midpoint of the common crus and external aperture), or its diameter is larger than that of the semicircular canal. It is usually bilateral. In many cases, vestibular aqueduct enlargement accompanies deformity of the cochlea, semicircular canals, vestibule and IAC. It also may be the sole radiographically detectable abnormality of the inner ear in a child with hearing loss. An enlarged vestibular aqueduct is frequently seen in Pendred syndrome.

2. Enlargement of the cochlear aqueduct

For enlargement of the CA to be diagnosed radiographically, the intraosseous portion coursing toward the vestibule must be enlarged beyond 1 mm, the practical resolution limit of contemporary CT scanners. If analogous criteria to enlargement of the VA are used, then an enlarged CA must have a diameter exceeding 2 mm throughout its course between the inner ear and posterior fossa. From a clinical standpoint, a CA less than 1 mm in diameter is undetectable radiographically. Despite the theoretical possibility that a widely patent CA may cause a stapedectomy gusher, abnormal connections between the internal auditory canal and vestibule appear to be etiologic in the majority of cases.

E. Internal auditory canal abnormalities *(figure 14)*

The normal diameter of the IAC ranges from 2 to 8 mm, with an average of 4 mm. Internal auditory canal abnormalities includes absence, stenosis, and enlargement. It also may have a bony septum that partitions it into two or more separate canals. The morphologic characteristics and size of the IAC are not reliable indicators of the integrity of the cochlear nerve, and normal size of the IAC and normal inner ear anatomy do not exclude a nerve deficiency (nerve may even be absent when the IAC and labyrinth are completely normal).

1. **Stenotic internal auditory canal**

A stenotic IAC may indicate a failure of eighth nerve development. When a patient has normal facial function and an IAC less than 2 mm in diameter, it is likely that the bony canal transmits only the facial nerve (absent eighth nerve). A narrow IAC may accompany inner ear malformations or may be the sole radiographically detectable anomaly in a deaf child.

1. **Enlarged internal auditory canal.**
A congenitally large canal may be an incidental finding in normal individuals and does not correlate with the level of hearing. The importance of detecting enlargement of the IAC is its association with spontaneous CSF leak and gusher during stapes surgery.
**Fig. 5:** Michel deformity - Axial HRCT showing the absence of inner ear structures(*) with normal middle ear cavity and a double internal auditory canal(+).

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**Fig. 6:** Cochlear Aplasia - Axial HRCT showed the absence of the cochlea(*) and the presence of a cystic vestibule (+) with a narrow internal auditory canal.

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**Fig. 7:** common cavity deformity axial (a, b) and coronal (c, d) CT show cochlea and vestibule forming a cystic cavity, absent modiolus and abnormal IAC (+).

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Fig. 8: Incomplete partition type I_ Axial HRCT; Two separated cystic cavities cochlea (*) and vestibule (+) dilated, and a dividing septum (arrow) between, giving the "snowman" appearance. Large IAC (1)

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Fig. 9: Cochlear hypoplasia. Axial (a, b) and coronal (c, d) HRCT: hypoplastic cochlea (bud) with no identified turn (*) and a normal vestibule in this case (+).

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**Fig. 10:** Mondini deformity_axial (a, b) and coronal (c, d) HRCT show cochlea has a turn and a half with a normal basal turn (arrows. the vestibules are intact (*)).

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**Fig. 11:** Lateral semi circular canal dysplasia_axial(a) and coronal(b) HRCT: The lateral semi circular canal is short and wide (arrow) and forms a common lumen with the vestibule(*). Dilated IAC and normal basal turn of the cochlea.

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Fig. 12: Semicircular canal aplasia_axial(a) and coronal(b) HRCTn*: The superior, posterior and lateral semicircular canals are absent with a rudimentary vestibule.

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**Fig. 13:** Axial CT: Enlarged vestibular aqueduct yellow arrow compared with width of posterior Scc(red arrow).

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**Fig. 14:** axial(a) and coronal(b) HRCT showing a enlarged internal auditory canal(*) and a narrow one (arrow).

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Conclusion

Congenital inner ear anomalies are important causes of sensorineural hearing loss in children which can be diagnosed on imaging. Being familiar with the prototype of various stages of developmental arrest and coupled with careful description of anomalies in anatomic terms allows proper communication with clinicians and facilitates multidisciplinary management. So, it is of paramount importance to understand the imaging manifestations of these conditions and to be aware of their classification and imaging appearance.
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