Imaging Diagnostics: Congenital Malformations and Acquired Lesions of the Inner Ear

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Abstract
Introduction: Congenital malformations and acquired lesions of the inner ear are characterised by small structural changes in this region. In recent decades, treatment options have improved considerably. At the same time, there has been a great advancement in diagnostic methods, obtaining high-resolution labyrinth images.

Currently, we use a 64-multislice computed tomography scanner in spiral mode (Brillance 64 Phillips, Eindhoven, the Netherlands), with an overlap of 0.66 mm and an interval of 0.33 mm, 120 kV and 300 mA. The magnetic resonance images were taken with Signa HDxt 1.5 and 3.0 T units (GE Healthcare, Waukesha, WI, USA).

We reviewed the radiological features of the lesions affecting the inner ear. They are classified as congenital (labyrinth malformation and statoacoustic nerve deficiencies) or acquired (otospongiosis, labyrinthitis, Ménière’s disease, inner ear haemorrhage, intralabyrinthine schwannoma and endolymphatic sac tumour).

Conclusion: Magnetic resonance imaging and computed tomography play an essential role in diagnosing patients with inner ear pathology. The technique selected should be chosen depending on the clinical setting. In a generic way, tomography is the method of choice for the study of traumatic pathology or otospongiosis. When tumour or inflammatory pathology is suspected, magnetic resonance is superior. In cases of congenital malformation, both techniques are complementary.

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Introduction

Congenital malformations and acquired lesions of the inner ear (IE) are characterised by small structural changes. Treatment options in this specific region have improved considerably in recent decades. At the same time, there has been great advancement in diagnostic methods, and it is currently possible to obtain high-resolution images of the labyrinth and the stacoacoustic nerves.

Computed tomography (CT) and magnetic resonance (MR) imaging of the temporal bone enable excellent anatomical definition of the inner ear and are used in daily practice to study its pathology. Computed tomography is the method of choice for studying the bony structures of the ear, whereas magnetic resonance imaging provides images of both the membranous labyrinth and the cranial nerves.

With the aid of both diagnostic techniques, the radiologist can provide referring ENT specialists with the necessary information to enable a correct treatment approach. We consider that interdisciplinary collaboration is necessary and that all parties should have extensive knowledge of disorders of the inner ear and their diagnosis through imaging.

In this article, we shall describe and illustrate the radiological characteristics of the most common congenital and acquired diseases of the IE, showing the keys to correctly interpreting them on radiological images.

Technique

In general CT images were taken with a thickness of 0.625-1mm, and reconstructed using a bone algorithm. Currently, we use a 64-multislice CT scanner (Brilliance 64 Phillips, Eindhoven, the Netherlands), with an overlap of 0.66 mm and an interval of 0.33 mm, 120kV and 300 mA. The magnetic resonance images were taken with Signa HDxt 1.5 and 3.0T (GE Healthcare, Waukesha, WI, USA).

Radiological Characteristics

The lesions found were classified according to their origin as congenital (labyrinth malformation and stacoacoustic nerve deficiencies) or acquired (otospongiosis, labyrinthitis, Ménière’s disease, intralabyrinthine schwannoma and endolymphatic sac tumour).

Congenital Malformations of the Inner Ear

Congenital malformations are defined as variations in the normal anatomical development of the inner ear, which result in functional disorders of the inner ear. The bony labyrinth develops between the fourth and eighth weeks of gestation; from the eighth week, it grows and ossifies. The structural malformations which can be diagnosed using radiological imaging of the inner ear are due to defects forming between the fourth and eighth weeks, whereas subsequent lesions affect the sensory epithelium and cannot be imaged. This is why in long-time series studies only 20% of patients with congenital hearing loss have malformations which are visible radiologically.
Cochlear Aplasia

Aplasia is a rare malformation, constituting only 3% of cochlear malformations. Embryonic development is arrested at the end of the third week of gestation and it is usually of unknown aetiology. Cochlear hypoplasia constitutes 15% of cochlear malformations and is due to an anomaly in the development of the cochlear duct during the sixth week of gestation.

Patients present with unilateral or bilateral sensorineural hearing loss from birth. Both are diagnosed using imaging tests. Cochlear absence is confirmed in cochlear aplasia. The vestibule and semicircular canals (SCC) are often malformed, globular or dilated and the cochlear nerve and its duct are missing (Fig. 1A). It is important to differentiate aplasia with cystic vestibule from cystic cochleovestibular anomaly (CCVA), since cochlear implants (CI) are contraindicated for aplasia but not for CCVA, the only possible treatment being brainstem implant. It is also important to differentiate this from labyrinthitis ossificans, in which the bulge of the promontory caused by the basal turn of the cochlea can be seen: this characteristic is not present in cochlear aplasia.

In cochlear hypoplasia, a small cochlea is observed (1–3 mm) with a single primitive turn. The vestibule and the semicircular canals can be normal or they can be malformed (Fig. 1A).

Malformation of the Vestibule and Semicircular Canals

The development of the semicircular canals starts at the sixth week of gestation and ends at the 22nd week. There are dysplasias which only affect the development of SCCs which are shorter and wider. Other malformations affect the vestibule and the SCCs, but the most common presents with an enlarged globular vestibule and dilated lateral SCC, the last to form (Fig. 1B). Aplasia of the SCCs is less common than dysplasia. It is usually associated with atresia of the oval window and an abnormal trajectory of the facial nerve. In CHARGE syndrome (Coloboma, Heart anomalies, Atresia choanae, Retarded growth and development, Genital hypoplasia, Ear anomalies) (Fig. 1B) all the SCCs can be absent.

They present clinically with vestibular alteration and can cause different degrees of hearing loss depending on their association with cochlear malformations. In both cases, the diagnostic method of choice is CT.

Deficiency of the Cochlear Nerve

In the majority of cases this is a pathology acquired in the womb or the neonatal period. In cases where a nerve has not existed (malformation at 7 weeks’ gestation) the duct will not develop either.

In cases where the nerve has not developed, CT can show atresia or stenosis of variable degree of the cochlear nerve duct, 1.4 mm being considered its normal minimum diameter. As we have already mentioned, most cases are acquired in the womb and the neonatal period; therefore, the diameter of the bony conduit and the inner auditory canal (IAC) are normal, and have low sensitivity for diagnosis. Magnetic resonance imaging is the technique of choice to show deficiencies of the cochlear nerve as it can be visualised directly along its entire length and relative...
measurements can be obtained, in particular of the facial nerve (Fig. 1C).

This pathology should be suspected in cases of profound sensorineural hearing loss, profound deafness in patients with retarded growth and development, CHARGE syndrome, auditory neuropathy spectrum disorder, sensorineural hearing loss acquired in the neonatal period, or in cases of unilateral sensorineural hearing loss.11

Incomplete Partition–Type I (IP-I)/Cystic Cochleovestibular Anomaly
IP-I is a malformation which starts at 5 weeks’ gestation and affects the development of the internal cochlear structure. There are different grades of anomaly. At its mildest, the malformation is confined to the cochlea, with normal vestibule and SCCs, at this grade there is no intercalar septum and the modiolus is absent (Fig. 2A). At its most severe, the cochlea, the vestibule and the horizontal SCC are globularly enlarged and make a single cavity which typically forms a figure-eight contour.5,12 This dysplasia is known as cystic cochleovestibular anomaly (Fig. 2A). These findings are confirmed by CT. During inner ear surgery there is a greater risk of meningitis or perilymphatic fistula due to the characteristic dilatation of the inner auditory canal.5,6

Dilated Vestibular Aqueduct Syndrome
This is the most common pathology detected by imaging in patients with progressive sensorineural hearing loss since childhood.13 The malformation is usually bilateral in 90% of cases and is more prevalent in females.

Diagnosis is established by the association of an appropriate clinical symptom context (bilateral sensorineural hearing loss) and characteristic radiological findings. With this malformation there is widening of the vestibular aqueduct and dilatation of the endolymphatic sac can be associated. Computed tomography shows a dilated bony vestibular aqueduct of a calibre exceeding 1.5 mm at the midpoint of its route between the common crusa and its external opening.14 This malformation can be associated with a dilated endolymphatic sac shown on MR, and its projection outside the bone into the posterior fossa (Fig. 2B, C).15

In 84% of cases, this malformation tends to be accompanied by another inner ear anomaly. The most common cochlear malformation is type II incomplete partition. It is also commonly associated with Prendes syndrome which presents with sensorineural hearing loss and goitre.8

Incomplete Partition–Type II (IP-II)
Also known as Mondini’s anomaly. This represents 30% of congenital malformations of the IE. There is fusion of the middle and apical turns of the cochlea, with absence of intercalar septum, whereas the basal turn is present.16 It is a result of arrested development of the ear during the 6th and 7th week of gestation; the cause is genetic or due to infectious or toxic embryopathy.

Patients characteristically present with severe hearing deficit at low frequencies due to apical malformation of the cochlea, whereas high frequencies tend to be preserved. However, other authors such as Sancho Serrano et al., confirm that there is no parallelism between histopathological lesions and hearing deficit.16

Diagnosis is made using CT (Fig. 2B). This type of malformation can be associated with anomalies in the formation of the vestibule and semicircular canals.
Incomplete Partition–Type III (IP-III)/X-Linked Stapes Gusher
Congenital autosomal recessive malformation due to a mutation of the POUF4 gene in chromosomal region Xq21: women are carriers and men are affected. It presents with progressive sensorineural hearing loss associated with conductive hearing loss, which can be masked if the sensorineural hearing loss is profound. There are very few publications to date with regard to this anomaly which describe its radiological characteristics. Computed tomography findings are absence of modiolus in the cochlea, and the interscalar septum is present (different to IP-I when both are missing) which gives it the typical “corkscrew” appearance (Fig. 2C). The external dimensions of the cochlea are normal, but its location is completely lateral to the IAC, rather than anterior which is usual. The IAC presents lateral bulbous dilatation and deficient lamina cribrosa and the facial nerve duct presents widening of the labyrinthine segments and proximal tympanic segment.

In this malformation, despite there being an absence of modiolus and free communication of cerebrospinal and perilymphatic fluid, the risk of recurrent meningitis is less common than with IP-I, due to the stapes being attached to the oval window which prevents fistula.

With regard to treatment, it is recommended that surgery is avoided using external hearing aids in cases of conductive hearing loss alone. If the patient suffers sensorineural hearing loss, a CI would be indicated taking into account the risk of perilymphatic fistula during cochleostomy caused by pressure increase in the perilymphatic space due to the absence of the lamina cribrosa.

Dehiscence of the Superior Semicircular Canal
In normal conditions, only the oval window and the round window act to compensate the movement of fluids in the IE. The SCCs are a closed system in which there is no movement of fluids when the stapes vibrate in response to sound. In patients with dehiscence of the superior semicircular canal, a “3rd window” form which creates an imbalance of this flow, and characteristically the patients will suffer vertigo and dizziness induced by intense noise (Tullio’s phenomenon), with Valsalva cough or manoeuvres.

Computed tomography is the imaging technique of choice (Fig. 3). For a safe diagnosis, it must be shown on at least two consecutive slices that there is no bony overlap. However, it has been demonstrated that there are cases in which the bony layer is so fine that it is not identified on CT scanning. Therefore, in order to make a reliable diagnosis, radiological findings should be compatible with clinical tests and the typical symptoms of vertigo on exposure to intense noise. It is also possible to evaluate dehiscence of the superior SCC using magnetic resonance imaging.

Acquired Lesions of the Inner Ear
Acquired lesions of the IE can be subdivided in turn into inflammatory lesions, traumatic lesions and tumours.

Otospongiosis
This is also called otosclerosis and is an idiopathic process characterised by presentation of reabsorption foci and pathological neoformation of spongy, immature and vascularised bone, located in the labyrinthine capsule. Vascularisation subsequently diminishes and the foci calcify. It initially affects the bony portion located anterior to the oval window (fissula ante fenestram) producing conductive hearing loss, secondary to the invasion of the footplate by abnormal bone. In advanced stages, it progresses to mixed or sensorineural hearing loss as these foci advance towards the cochlea.

We can classify otospongiosis radiologically into 4 grades, according to the extension of the lesion (Fig. 4). Grade 1 is in turn divided into 1a and 1b; 1a presents isolated lesion of the footplate, which is identified as thick (above 0.6) and hypodense. Grade 1b corresponds to an area of pre-stapedial hypodensity below or equal to 1 mm, without extension to the anterior portion of the middle cochlear turn. Grade 2 corresponds to a hypodense lesion with a diameter above 1 mm, without contact with the cochlear lumen. Grade 3 identifies a hypodense pre-stapedial focus in contact with the cochlear lumen. Grade 4 is divided into 4a and 4b as well. Grade 4a corresponds to plaques around the cochlea, and 4b corresponds to plaques around the lumen of the semicircular canals and the vestibule.

Labyrinthitis
Inflammation of the IE can occur as a result of an infection (viral or bacterial), as a result of an autoimmune process or post trauma. Inflammation can occur from direct spread from the IE or the meninges or, more rarely, from the blood. The patient usually presents with hearing loss and vertigo.

The evolution of labyrinthitis is classified into three stages: acute, fibrous and ossifying (labyrinthitis ossificans). In the acute stage, there is contrast enhancement on MR imaging, but CT is normal. In the intermediate stage, there is a loss of signal intensity of the liquid in intense T2-weighted sequences and the CT remains normal. In labyrinthitis ossificans, there is already alteration on the CT images, diffuse ossification of the entire labyrinth.
Congenital and Acquired Pathology of the Inner Ear

Figure 4  Axial CT of the ears of different patients showing the four radiological grades of otospongiosis. The arrows show the lytic foci of spongy bone. (1a) Isolated lesion of the footplate. (1b) Spongiotic focus in fissula ante fenestram less than 1 mm. (2) Otospongiotic focus above 1 mm not in contact with the middle turn of the cochlear lumen. (3) Spongiotic focus in contact with the cochlear lumen. (4) Confluent otospongiosis around the cochlea.

is characteristic (Fig. 5A). In T2 weighted MR images, there is a decrease in the intensity of the signal from the membranous labyrinth. Magnetic resonance findings in the initial non-bony forms of cochlear obliteration are very relevant with regard to cochlear implants, especially in postmeningitic cases.

Inner Ear Haemorrhage
This is usually secondary to anti-coagulant treatment or trauma (with or without fracture of the temporal bone). Other possible less frequent causes are tumours, labyrinthitis or haematological diseases such as leukaemia or coagulopathies. Clinically, it causes sudden deafness.

Figure 5  (A) Axial CT of right temporal bone showing ossification of the basal turn of the cochlea characteristic of labyrinthitis ossificans (black arrows). (B) T1 weighted coronal plane MR imaging without intravenous contrast showing an increased signal intensity of the membranous labyrinth, corresponding to intralabyrinthine haemorrhage (white arrow) (metahaemoglobin).
Figure 6  (A) MR FLAIR image after injection of intravenous contrast showing dilatation of the endolymphatic space particularly prominent in the cochleae. (B) Detail of the left ear image where white arrows indicate dilatation of the endolymphatic space in the cochlea.

Figure 7  (A₁) and (A₂) T2 and T1 weighted MR images with intravenous contrast, showing a small lesion on the inside of the basal turn of the right cochlea (white arrow) which intensely and homogeneously enhances after injection of contrast. This lesion corresponds to an intracochlear SIL. (B₁) and (B₂) T1 weighted MR images with and without intravenous contrast, showing a transmodiolar SIL with intense enhancement occupying the cochlea (white arrow) and crossing the modiolus to the bottom of the IAC (arrow tip).
Magnetic resonance imaging is the technique of choice to evaluate the membranous labyrinth. In normal conditions, in T1 weighted sequences without contrast, the membranous labyrinth is visualised as an intermediate signal structure, similar to the brain parenchyma. When there is an intralabyrinthine haemorrhage, there is a characteristic hyperintensity of signal in this sequence (Fig. 3B).33,34

Ménière’s Disease
Also known as endolymphatic hydrops, this is characterised by a triad of vertigo, tinnitus and unilateral sensorineural hearing loss. The physiopathological mechanism which triggers hydrops is an increase in endolymphatic fluid in the IE, because its absorption is reduced due to a malfunction of the endolymphatic sac.35 During acute episodes, endolymph expansion in the vestibular system causes ballooning in the perilymphatic space which ruptures the separating membrane. The mixture of the fluids of both compartments bathes the nerve receptors which trigger vertigo. The mechanical disturbance of the organ of Corti causes hearing loss.36

Visualisation of endolymphatic hydrops in Ménière’s disease has always been a major objective for clinicians, and demonstration was only possible in post-mortem histopathological studies; however, it can now be visualised using magnetic resonance imaging.37

The current method for visualising endolymphatic hydrops is based on the endolymphatic space being leak tight and isolated from perilymph, cerebrospinal fluid and blood (Fig. 6).38 After injection of intratympanic or intravenous contrast there is only contrast enhancement of perilymph and not endolymph.24 The FLAIR sequence is very sensitive to changes in T1 and other subtle alterations in the composition of the fluids, it enables perilymph to be visualised with high signal intensity (bright) surrounding the endolymphatic space which has not taken up contrast (dark).39,40

Intralabyrinthine Schwannoma
Intralabyrinthine schwannomas are benign tumours which arise de novo from the perineural Schwann cell sheath of the intralabyrinthine branches of the vestibulocochlear nerve and initially have no component in the internal auditory canal.41 According to their origin and extension, they are classified as: intracochlear (Fig. 7A), intravestibular, vestibulocochlear, transmodiolar in cases where the tumour crosses the modiolus from the cochlea to the bottom of the IAC (Fig. 7B), transmacular when it crosses from the vestibule to the bottom of the IAC, and transotic when it crosses the entire IE from the bottom of the IAC to the middle ear.

It has been published that intracochlear schwannomas originate most commonly in the scala tympani, and later growing towards the scala vestibuli.42 They present clinically with tinnitus, vertigo and unilateral progressive sensorineural hearing loss.

In T2 weighted MR sequences they are identified as a “filling” defect inside the hyperintense perilymph, and intensely enhance after contrast injection.43

Endolymphatic Sac Tumour
The endolymphatic sac is located below the dura of the posterior fossa, at the end of the vestibular aqueduct. Its function is to drain the endolymph from the membranous labyrinth. Endolymphatic sac tumour is a neoplasm which originates in the neuroectodermal epithelium of the membranous labyrinth of the IE. It is a rare tumour which presents with unilateral sensorineural hearing loss. It was initially considered an adenocarcinoma because of its high local destructive capacity and recurrence, however, no histological malignancy has been demonstrated and its capacity to metastasise is almost nil. Anatomopathologically, it is a papillary cystoadenoma originating from the pars rugosa (middle third) of the endolymphatic sac. It presents sporadically, but patients with von-Hippel-Lindau disease have a 15% predisposition for the tumour.44

Computed tomography shows localised bone destruction which is characteristically retro labyrinthine, with central intratumoral bone spicules.45 The characteristic MR image is heterogeneous, with hyperintense foci in T1 weighted sequences on haemorrhagic areas, and after injection of contrast, there are areas of decreased uptake due to calcification and haemosiderin deposit (Fig. 8).

Conclusions
Magnetic resonance imaging and computed tomography play an essential role in diagnosing patients with inner ear
pathology. These techniques are considered complementary. Computed tomography is important because it precisely delimits the bone component of the otic capsule. With the high capacity of contrast, magnetic resonance enables soft tissues to be evaluated and characterised: using this technique the membranous labyrinth and the cranial nerves can be represented in an excellent way. Furthermore, it is possible to visualise brain anomalies coexisting with malformations or other IE alterations.

We consider it necessary to understand the embryogenesis and the anatomy of the inner ear. Likewise, we must be aware of congenital and acquired disorders which might affect the inner ear. The technique of choice should be chosen in line with the clinical scenario. In general, computed tomography is the method of choice to study traumatic pathology or otospongiosis. When tumoral or inflammatory disease is suspected, magnetic resonance imaging has been shown to be superior. Both techniques are complementary in cases of congenital malformation.

Conflict of Interest
The authors have no conflict of interests to declare.

References


