RADIOLOGY THROUGH IMAGES

Inner ear malformations: A practical diagnostic approach

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Abstract Pediatric sensorineural hearing loss is a major cause of disability; although inner ear malformations account for only 20–40\% of all cases, recognition and characterization will be vital for the proper management of these patients. In this article relevant anatomy and development of inner ear are surveyed. The role of neuroimaging in pediatric sensorineural hearing loss and cochlear preimplantation study are assessed. The need for a universal system of classification of inner ear malformations with therapeutic and prognostic implications is highlighted. And finally, the radiological findings of each type of malformation are concisely described and depicted. Computed tomography and magnetic resonance imaging play a crucial role in the characterization of inner ear malformations and allow the assessment of the anatomical structures that enable the selection of appropriate treatment and surgical approach.

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PALABRAS CLAVE
Imagen diagnóstica; Oído interno; Anormalidades; Displasia coclear; Aplasia coclear; Partición incompleta

Malformaciones del oído interno: una aproximación diagnóstica práctica

Resumen La hipoacusia neurosensorial pediátrica es una causa mayor de discapacidad. Pese a que solo en el 20-40\% de los casos se identifica una malformación del oído interno, su detección es de vital importancia para el tratamiento de estos pacientes. En este artículo se repasan la anatomía y la embriogénesis del oído interno. Se valora el papel de la neuroimagen en la hipoacusia neurosensorial pediátrica y en el estudio preimplante coclear. Se destaca la necesidad de la utilización de un sistema universal de clasificación de las malformaciones del oído interno con

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implicaciones pronósticas y terapéuticas. Por último, se describen e ilustran de forma concisa los hallazgos radiológicos clave de cada tipo de malformación. La tomografía computarizada y la resonancia magnética desempeñan un papel crucial en la caracterización de las malformaciones del oído interno y permiten la valoración de las estructuras anatómicas que posibilitan la selección del tratamiento y del abordaje quirúrgico idóneos.

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Introduction

Sensorineural hearing loss in children is a major cause of disability. Its early diagnosis is very important since delayed diagnoses may affect the development of language, the academic skills and social and emotional development. It can be due to congenital or acquired anomalies; at least half of them have a genetic origin and among the acquired ones the infection due to cytomegalovirus is the most common cause.\(^1\) The prevalence of detectable cochleovestibular malformations is around 20–40\(^\circ\).\(^2\)

The cochlear implant is the standard therapeutic proceeding of mild to profound sensorineural hearing loss in children.\(^3\) Previously inner ear (IE) malformations were one counter indication for cochlear implants but advances in surgical techniques and cochlear devices have made it possible to implant cochleas with malformations.

Image modalities are crucial when it comes to doing preoperative assessments of inner ear malformations since they allow us to detect and evaluate the cochlear nerve and identify any anatomic variants and all potential surgical complications.\(^1\)

There are several categorization schemes for IE malformations and yet despite the fact that no scheme is perfect we should make the effort of trying to adopt a universal system that would allow us to share our results with the scientific community.

In this paper we will take a brief look into the anatomy and embryogenesis of the IE. We will also be evaluating the role of neuroimages in pediatric sensorineural hearing loss and in the cochlear preimplant study. We will be paying special attention to the use of a somehow universal system to categorize IE malformations with therapeutic and prognostic implications. Finally we will be describing the most widely accepted categorization while illustrating and detailing in a concise way the key radiologic findings of each type of malformation.

Anatomy and embryology of inner ear

The IE is made up of the membranous labyrinth that in turn surrounds the osseous labyrinth. The cochlea-organ responsible for hearing is a conical structure consisting of a duct that makes between 2.5 and 2.75 turns around a central core called modiolus (Fig. 1). From the modiolus a thinned-osseous spiral layer reaches out to divide the duct into vestibular duct (upper) and tympanic duct (lower).

Through the cochlear opening the cochlea meets the fundus of the internal acoustic canal (IAC) the cochlear nerve passes through. The vestibular system is made up of the vestibule and three semicircular ducts: superior, lateral, and posterior.

- Both the duct and the endolympathic sac are contained by the vestibular aqueduct reaching out from the labyrinth toward the posterior fossa epidural space.
- The IAC runs through the petrous part of the temporal bone and communicates the cerebellopontine angle cistern with the labyrinth through which the cranial nerves VII and VIII pass. In an oblique sagittal plane perpendicular to the IAC we can see the facial nerve in the anterior-superior quadrant, the cochlear nerve in the anterior-inferior quadrant and the upper and lower vestibular nerves in the posterior quadrants.
- The IE stems from the otic placode that starts developing during the 3rd week of pregnancy. The development of the cochlea is completed in the 8th week of pregnancy, the vestibule in the 11th week and semicircular ducts between the 19th and 22nd weeks of pregnancy. The first semicircular duct to develop is the superior one followed by the posterior one and the last one to develop is the lateral one.\(^4\)

The role of neuroimaging in inner ear malformations

Most candidates for a cochlear implant do not show anomalies in their temporal bone that can be identified through images,\(^1\) but if they show such anomalies finding them is very important. Both computed tomography (CT) and magnetic resonance imaging (MRI) give us an excellent representation of IE malformations and they are used in the systematic practice of the study of pediatric sensorineural hearing loss and cochlear pre-implants. Because of their complementary role the use of both image modalities is recommended since the rate of malformation detection increases dramatically.\(^5\)

CT allows us to detect bone malformations, also the facial nerve possible aberrant trajectory (more common in this population\(^1\)), vascular structure malformations and assess coexisting anomalies of both the inner and middle ears.

When it comes to the MRI it allows us to assess fluid-filled spaces of the IE and assess the cranial nerve VII and any other possible intracranial anomalies.

Combined they provide the surgeon with the necessary presurgical information that will allow him/her to make a therapeutic decision, give advise for parents and give the
Figure 1  Chart showing the EI made up of the cochlea (co), the vestibule (v) and the semicircular ducts (csc). The modiolus (m) is the base of the cochlea at the fundus of the internal acoustic canal (IAC). The cochlear coils are separated by the interscalar septum (ti); the coil is split into vestibular duct (rv) and tympanic duct (rt) by the spiral lamina (le) that stems from the modiolus. The vestibular aqueduct (av) stems from the vestibule toward the epidural space of the posterior fossa with a trajectory across the petrous part of the temporal bone and perpendicular to the IAC. (B) Representation of anatomical structures in a transverse image using computed tomography in bone window. (C) Representation of anatomical structures in a transverse image using a T2-weighted, echo-gradient, high-resolution, steady-state MRI sequence. (D) Oblique, sagittal reconstruction perpendicular to the IAC of the MRI sequence showing the facial nerve (nf) in the superior-anterior quadrant, the cochlear nerve (nc) in the inferior-anterior quadrant, the superior vestibular nerve (nvs) in the superior-posterior quadrant, ant the inferior vestibular nerve (nvi) in the inferior-posterior quadrant. In this image the anterior part is marked with an A and the posterior one with a P.

most likely prognosis for the cochlear implant, choose the surgical approach and the appropriate implants and warn about anatomic variants and any other possible surgical complications.¹

Classification

There are many classifications when it comes to IE malformations and it is precisely the absence of a common language that makes understanding hard for the scientific community. We need a universal classification system that allows us to determine the link among the different types of malformations and clinical prognosis. The most widely accepted categorization is Sennaroglu classification—this classification paper is based on it. It is based on embryogenesis. Every malformation is the consequence of an interruption in development at one time or another. Generally speaking it is useful and the more serious the malformation, the more surgical complications we will find and worse the outcome of cochlear implant will be.²,³,⁵,⁶

In our radiologic report on top of classifying malformations we also give detailed information on three anatomic structures involved in the cochlear implant: cochlear lumen—where the electrode will be placed; the modiolus—target of electrostimulation; and the cochlear nerve—the route through which the stimulus travels.²

Now let us take a look at the different types of malformations we can see (table available online).

Complete labyrinthine aplasia

Also known as Michel aplasia it is the result of the interruption of the otic placode development before the 3rd week of pregnancy. It amounts to 1% of IE malformations only.⁶ It counter indicates cochlear implantation and the best therapeutic option is the brainstem implant.⁸

Figure 2  Complete labyrinthine aplasia. (A) Transverse image using computed tomography in bone window with identifiable total absence of inner ear elements (asterisk). We can see hypoplasia in the petrous part of the temporal bone (white arrow), the atresic internal acoustic canal (IAC) with a reduced caliber (black arrow), and the flattening of the inner ear medial side (arrowhead). (B) Transverse image using T2-weighted, echo-gradient, steady-state MRI sequence where the aforementioned findings can be seen as well as the cranial pairs; in this case there is an absolute deficit of the vestibulocochlear nerve and only the facial nerve (arrowhead) can be seen in the cerebellopontine angle cistern and the IAC.
It is characterized by a total absence of IE structures (Fig. 2). The IAC is atresic, there is aplasia of the cochleovestibular nerve and the facial nerve trajectory is aberrant. It is associated with multiple anomalies of the temporal lobe among which the following ones are the most common of all: petrous apex hypoplasia, absence of round and oval windows and flattening of the middle ear medial wall due to absence of promotorium.4,9

Cochlear aplasia

It is due to developmental interruption at the end of the 3rd week of pregnancy10 and it amounts to 3% of all IE malformations.1 It is a counterindication for cochlear implantation.

The cochlea is absent and the vestibular system can be normal, dysplastic, distinguishable from the cochlea due to its posterior location with respect to the IAC (Fig. 3). The labyrinthic segment of the facial nerve has an aberrant trajectory and the inner ear medial wall is flattened.

We should remember that: in labyrinthine aplasia and cochlear aplasia there is flattening of the inner ear medial wall—useful finding to distinguish them from labyrinthitis ossificans (Fig. 4 available online). This differentiation is clinically relevant since in the labyrinthitis ossificans the cochlear implant is possible hard to accomplish.

Common cavity

It is the result of developmental interruption during the 4th week of pregnancy and amounts to 25% of all malformations.4,10 Cochlear implant is possible though hard to accomplish and the rate of complications is higher.11

It is characterized to be the confluence of the cochlea and the vestibule in one only cystic cavity without internal architecture (Fig. 5). There is a total absence of modiolus and a large communication between the cavity and the IAC.12 Semicircular ducts are usually dysplastic though there are times that they may look normal.4

Incomplete partition type I

Also known as cystic cochleovestibular malformation it is due to developmental interruption during the 5th week of pregnancy.10

The cochlea shows a total absence of the modiolus and cystic appearance without internal architecture; the vestibule is dilated. The labyrinth as a whole has the appearance of the figure eight (Fig. 6). In this malformation we can see the cochlea and the vestibule which in turn allows
Figure 5 Common cavity. (A) The chart shows common cystic cavity without internal architecture made up by rudimentary cochlea and vestibule. (B and C) Transverse image and coronal multiplanar reconstruction using computed tomography in window bone, respectively showing the common cavity (white arrows) and slightly dysplasic, widened and short semicircular ducts (black arrows). The fundus of the IAC showing cribiform plate deficits (arrowhead) enters the center of the common cavity; finding that allows us to distinguish this malformation from cochlear aplasia with dysplasic dilated vestibule in which the fundus of the IAC location is anterior to the cavity that is more posterior in the normal situation of the vestibule.

Incomplete partition type II

It is due to developmental interruption during the 7th week of pregnancy and it is the most common dysplasia of all (present in 50% of all malformations). It shows fusion of the middle and apical turns and a cystic appearance due to a defect in the apical segment of the modiolus, the interscalar septum and the osseus spiral lamina (Fig. 7). The modiolus basal coil and the basal segment look normal.

It is usually associated with an enlarged vestibular aqueduct (due to duct and endolymphatic sac dilation) and with a minimal vestibular dilation which makes up the Mondini deformity triad.  

We should remember that: malformations with cribiform plate deficits and a wide communication between the IAC and the cochlea (common cavity, incomplete partitions types I and III) predispose to a higher risk of recurrent meningitis, perilymphatic fistula and complications during surgery.

Incomplete partition type III

It is a recessive genetically inherited disease linked to chromosome X and it is not due to the developmental interruption of the otic placode. It usually affects males and mixed hearing loss of rapid progression.

There is a total absence of the modiolus though the interscalar septum is present (Fig. 8). The cochlea is located laterally to the IAC that is in turn widened and both widely communicated due to a complete deficit of the cribos plate.

We should remember that: malformations with cribiform plate deficits and a wide communication between the IAC and the cochlea (common cavity, incomplete partitions types I and III) predispose to a higher risk of recurrent meningitis, perilymphatic fistula and complications during surgery.

Figure 6  Incomplete partition type I. (A) The chart shows the cochlea with a total absence of modiolus and interscalar septum without internal architecture. The vestibule is dysplasic, slightly dilated and the labyrinth as a whole shows the shape of figure eight. (B) Transverses image using computed tomography in bone window. (C) Maximum intensity projection reconstruction of transverse image using T2-weighted, steady-state, echo gradient MRI sequence. In B and C we can see the following findings: cochlea without internal architecture (white arrow) and slightly dilated vestibule (black arrow) making up an eight shaped-labyrinth. Cribiform plate deficit (arrowhead) with wide communication between the IAC and the cochlea can be identified here. In C the cochlear nerve deficit can be seen (black asterisk in the theoretical location of the nerve). The patient shows chronic otitis media; in B we can see the occupation of the middle ear (white asterisk).
Cochlear hypoplasia

It is due to developmental interruption during the 6th week of pregnancy. The dimensions of the cochlea are smaller but there is a well-established differentiation between the cochlea and the vestibule. When in lack of experience measurements should be taken to facilitate its detection.

It is a generic term and three subtypes should be taken into consideration here (Fig. 9):

Type I, "yolk sac" of the cochlea: the cochlea is a small exsenceence stemming from the vestibule with no internal architecture and a total absence of modiolus and interscalar septum.

Type II, cystic hypoplastic cochlea: the cochlea shows reduced dimensions, no modiolus or interscalar septum but its external structure is normal. It shows wide communication with the IAC that predisposes to a higher risk of complications during surgery. The vestibular aqueduct is usually widened and the vestibule is slightly dilated.

Type III, cochlea with less than two turns: the cochlea shows less than two turns, the modiolus is smaller and the interscalar septum is shorter but both its internal and external architectures are normal. The vestibule and the semicircular ducts are usually hypoplastic.

Widened vestibular aqueduct syndrome

Its etiology is controversial; it could be due to postnatal developmental disorders. It consists of an isolated vestibular aqueduct widening due to the duct and endolymphatic sac abnormal developments being the remainder of the labyrinth normal (Fig. 10). The aqueduct is said to be...
Figure 9  Cochlear hypoplasia. (A) The chart shows the existing three types of cochlear hypoplasia: type I ‘‘yolk sac’’, like a small excrescence stemming from the vestibule; type II, cystic hypoplastic of normal external architecture but small dimensions and no internal architecture due to the absence of modiolus and interscalar septum; type III, less turns with normal external and internal architecture but shorter modiolus and interscalar septum which conditions the cochlea with less than two turns. (B) Transverse image using computed tomography (CT) in bone window showing hypoplastic ‘‘yolk sac’’-like cochlea stemming from the vestibule like a small excrescence (type I). (C) Transverse image using CT in bone window showing one hypoplastic cystic cochlea of normal external architecture but reduced dimensions and no internal architecture (type II). (D) Transverse image using CT in window bone showing one hypoplastic cochlea with less than two turns (type III).

Figure 10  Dilated vestibular aqueduct syndrome. (A) Transverse image using computed tomography in bone window showing vestibular aqueduct widening (white arrow); it is useful to use the caliber of the superior semicircular duct as a reference of the upper threshold of normalcy (arrowhead). (B) Transverse image using T2-weighted, steady-state, echo-gradient MRI sequence showing duct and endolymphatic dilation (white arrow) and the posterior semicircular duct as a reference (arrowhead). Both the cochlea and the rest of the inner ear are normal (black arrow in A and B).

widened when its caliber is larger than 0.8 mm in its central point in an oblique plane at 45° (plane of Pöschl). 19

Vestibular system malformations

It is due to developmental disorders between the 6th and 22nd weeks of pregnancy. In isolation it is not relevant for the cochlear pre-implant study; its importance has to do with a strong association with other IE malformations and certain syndromes. The vestibule and the lateral semicircular duct dysplasia is one of the strongest ones and consists of a short and wide lateral semicircular duct fused or separated through a small bony islet to the vestibule of globular appearance (Fig. 11). There is a strong correlation between the aplasia of semicircular ducts and CHARGE syndrome 20 (Fig. 12).
malformation and illustrate and give precise details on key radiological findings.

Ethical responsibilities

Protection of people and animals. The authors declare that the proceedings followed fully abide by the ethical rules and regulations from the human experimentation committee and are in full compliance with the World Health Organization (WHO) and the Declaration of Helsinki.

Confidentiality of data. The authors confirm that they have followed their centers protocols on the publication and disclosure of data from patients.

Right to privacy and informed consent. The authors confirm that they have received written informed consent from the patients and/or individuals referred to in this paper. This manuscript belongs to the corresponding author.

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Conflicts of interests

The authors declare no conflict of interests associated with this article whatsoever.

Appendix A. Supplementary data

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