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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
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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%.[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 endolymphatic 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[6]), 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

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