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ORIGINAL ARTICLE

Hearing Loss and Enlarged Internal Auditory Canal in Children☆

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KEYWORDS
Hearing loss in children;
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Abstract
Introduction: Among the temporal bone abnormalities that can be found in the etiological study of paediatric sensorineural hearing loss (SNHL) by imaging techniques, those related to the internal auditory canal (IAC) are the least frequent. The most prevalent of these abnormalities that is associated with SNHL is stenotic IAC due to its association with cochlear nerve deficiencies. Less frequent and less concomitant with SNHL is the finding of an enlarged IAC (>8 mm).
Methods: Retrospective and descriptive review of clinical associations, imaging, audiological patterns and treatment of 9 children with hearing loss and enlarged IAC in the period 1999–2012.
Results: Two groups of patients are described. The first, without association with vestibulocochlear dysplasias, consisted of: 2 patients with SNHL without other temporal bone or systemic abnormalities, one with bilateral mixed HL from chromosome 18q deletion, one with a genetic X-linked DFN3 hearing loss, one with unilateral hearing loss in neurofibromatosis type 2 with bilateral acoustic neuroma, and one with unilateral hearing loss with cochlear nerve deficiency. The second group, with association with vestibulocochlear dysplasias, was comprised of: one patient with moderate bilateral mixed hearing loss in branchio-oto-renal syndrome, one with profound unilateral SNHL with recurrent meningitis, and another with profound bilateral SNHL with congenital hypothyroidism.
Conclusions: The presence of an enlarged IAC in children can be found in different clinical and audiological settings with relevancies that can range from life-threatening situations, such as recurrent meningitis, to isolated hearing loss with no other associations.

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Hipoacusia infantil;
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Hipoacusia en niños con conducto auditivo interno agrandado

Resumen
Introducción: Entre las anomalías del hueso temporal que pueden encontrarse en el estudio etiológico de la hipoacusia neurosensorial (HANS) infantil mediante pruebas de imagen, las relacionadas con el conducto auditivo interno (CAI) se hallan entre las menos frecuentes. De ellas, la más prevalente y relacionada con HANS es el CAI estenótico por su asociación a deficiencias del nervio coclear. Menos frecuente y menos concomitante con HANS es el hallazgo de un CAI agrandado (> 8 mm).
Métodos: Estudio retrospectivo y descriptivo de las asociaciones clínicas, estudios de imagen, patrones audiológicos y opciones de tratamiento de 9 niños diagnosticados de hipoacusia en el periodo 1999–2012 con un CAI agrandado.
Resultados: Se describen 2 grupos de pacientes. El primero, sin asociación con displasias cocleovestibulares: 2 pacientes con HANS sin otras alteraciones de hueso temporal o sistémicas, una hipoacusia mixta bilateral con cromosomopatía por deleción 18q, una hipoacusia genética DFN 3 ligada a X, una hipoacusia unilateral en neurofibromatosis tipo 2 con neurinoma del acústico bilateral, y una hipoacusia unilateral con déficit de nervio coclear unilateral; y un segundo grupo con asociación a displasias cocleovestibulares: una hipoacusia mixta bilateral moderada en síndrome branquio-oto-renal, una HANS profunda unilateral con meningitis recurrentes, y una HANS bilateral profunda con hipotiroidismo congénito.
Conclusiones: La presencia de un CAI agrandado en niños puede encontrarse en diferentes contextos clínicos y audiológicos, con relevancias que pueden variar desde situaciones con riesgo vital como en meningitis recurrentes, hasta hipoacusias aisladas sin otras asociaciones.

Introduction

Progress in imaging techniques in the last several years has permitted greater precision in the aetiological and physiopathological study of paediatric sensorineural hearing loss (SNHL).[1] Such techniques are some of the most effective methods for discovering findings that make explaining the origin of hearing loss possible.[2] Middle ear malformations found in imaging tests in SNHL present great variability in both the type of structures affected and in the concomitant circumstances between the different parts of the inner ear involved.[1] Nevertheless, the enlarged vestibular aqueduct has been defined as the most frequent congenital anomaly found in radiological studies in children with SNHL.[3]

The internal auditory canal (IAC) is a part of the temporal bone whose development can be changed in the postnatal period, depending on neumatization, especially in its length, in its most medial area.[4,5] However, in the most lateral area (the fundus), the transverse or falciform crest and Bill's bar do not seem to be modified after birth.[5] Malformations related to the IAC are among the least frequent.[1] Examples such as absence, stenosis, duplication, anteversion and verticalization,[6] as well as bulbous enlargement of the IAC have been described.[7]

Among these malformations, the most prevalent and related with SNHL is stenosis in the IAC (<2 mm) because of its association with hypoplasias and aplasias of the auditory nerve; promontorial stimulation and functional magnetic resonance imaging (MRI) tests of the auditory can be required to rule out the presence of non-visualised small fibres of the auditory nerve.[7–10]

Less frequent and less concomitant with SNHL is the finding of a bulbous, dilated or enlarged IAC.[1,11] Although there are no agreed-upon criteria for a precise definition, a measurement of more than >8 mm for the outer diameter could be considered sufficient for describing an IAC as widened.[7,12,13] Its relationship with the clinical audiological associations with this finding is also not well defined. The first descriptions of a link between hearing loss and enlarged IAC in children were published in the 1970s, in some cases finding concomitances with other malformations of the temporal bones and different hearing loss patterns.[14,15]

In this context the most evident associations have been established with neurofibromatosis and DFN 3 (X-linked hearing loss with stapes gusher and widened IAC); however, the presence of an enlarged IAC has also been found either isolated or associated with other syndromic systemic pathologies, and/or with other alterations del temporal bone (cochleovestibular dysplasia, other occupying lesions, etc.).[7,16]

The most evident physiopathological mechanism attributed to IAC dilations refers to their possible link with a widening of the modiolus, cause of alterations in labyrinth pressure. It can be the origin of meningitis, fluctuating and/or progressive hearing loss, tinnitus and dizziness secondary to labyrinthine dropsy and to fistulization of the middle ear from abnormal communications between the perilymphatic and subarachnoid spaces.[17] These situations have been described as occurring especially in dysplasia of the IAC fundus with wide modiolus where the IAC opens directly into the cochlear canal, dilatation of the arachnoid sheaths around the optic nerve, cochlear dysplasia with incomplete bone separation together with dilation of the basal and vestibule turns, dilated cochlear aqueduct and DFN 3.[18–20]