ORIGINAl Article

Hearing Loss and Enlarged Internal Auditory Canal in Children

Saturnino Santos,* M. Jesús Domínguez, Javier Cervera, Alicia Suárez, Antonio Bueno, Margarita Bartolomé, Rafael López

Servicio de Otorrinolaringología, Hospital Infantil Universitario Niño Jesús, Madrid, Spain

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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).
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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* Corresponding author.
E-mail address: satursantos@gmail.com (S. Santos).
PALABRAS CLAVE
Hipoacusia infantil; Conducto auditivo interno; Malformaciones de oído interno

Hipoacusia en niños con conducto auditivo interno agrandado

Resumen
Introducción: Entre las anomalías del hueso temporal que pueden encontrarse en el estudio y 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 período 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 delección 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 defecto 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.

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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). Such techniques are some of the most effective methods for discovering findings that make explaining the origin of hearing loss possible. 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. Nevertheless, the enlarged vestibular aqueduct has been defined as the most frequent congenital anomaly found in radiological studies in children with SNHL.

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. 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. Malformations related to the IAC are among the least frequent. Examples such as absence, stenosis, duplication, anteverision and verticalization, as well as bulbous enlargement of the IAC have been described.

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.

Less frequent and less concomitant with SNHL is the finding of a bulbous, dilated or enlarged IAC. 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. 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.

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 of temporal bone (cochleovestibular dysplasia, other occupying lesions, etc.).

The most evident physiopathological mechanism attributed to IAC dilations refers to their possible link with a widening of the modiolus, cause of alterations in labyrinthine 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. 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 dilatation of the basal and vestibule turns, dilated cochlear aqueduct and DFN 3.
The objective of this study was to describe the clinical and audiological characteristics of 9 cases of children with enlarged IAC and some type of hearing loss with a concomitant sensorineural component.

Methods

This was a retrospective, described study of the case histories of 9 children with a diagnosis of hearing loss in the 1999-2012 period that presented an IAC≥8mm in its outer diameter in the imaging tests and/or a description of ‘enlarged IAC’ in the radiological report. The clinical associations, imaging studies audiological patterns found and treatment options are described.

Results

In Tables 1 and 2 the clinical, audiological and imaging technique findings, diagnoses and treatments are presented for 6 patients with enlarged IAC without any associated cochleovestibular dysplasia, and for 3 patients with enlarged IAC with associated cochleovestibular dysplasia, respectively.

Discussion

Cases not Associated With Cochleovestibular Malformations

Cases 1 and 2

The finding of a uni- or bilateral enlarged IAC in an asymptomatic patient has been considered as a variant of normality. Interpreting this finding, in the presence of SNHL, is a bit more controversial, above all in cases with slightly increased measurements and lacking other clinical or radiological associations. The majority of the studies still consider this as not significant; given that no statistical correlation with SNHL has been shown. Nevertheless, sudden hearing losses in adults related with the presence of enlarged IAC have been described. In our series, Patients 1 and 2 would fall within the group of SNHL lacking other clinical or radiological alterations as evidenced in the aetiological study, without the finding of an enlarged IAC being significant in explaining the hearing loss.

Case 3

X-linked hearing loss with stapedial gusher and enlarged IAC defined as DFN 3 (deletion of the POU3F4 gene) is characterised by a progressive, mixed loss with stapedial fixation, although it often presents as profound or rapidly progressive sensorineural hearing loss. The most important finding in imaging tests is an enlarged IAC and a separation defect of the basal wall of the cochlea, producing a cerebrospinal fluid hyper-pressure transmitted to the perilymph, justifying the mixed component and the gusher. However, this defect in relation to the basal wall of the cochlea is not present in all these patients, making the physiopathological explanation less concordant. The patient with DFN 3 in our series did indeed present this communication with the basal wall of the cochlea, as well as a probably congenital severe hearing loss in keeping with the lack of linguistic development, diagnosed with delay in the period prior to the implantation of universal neonatal auditory screening.

Case 4

Deletion 18q syndrome involves a series of abnormalities, including midfacial hypoplasia with prominent forehead, vertebral alterations, short stature from growth hormone deficit and cognitive delay. The most normal in the auditory system is stenosis of the external auditory canal, while atresia can be found. The most common hearing loss is conductive; however, as occurs with the child in our series, loss with a sensorineural component has been published in isolated cases. Although it has not been related with enlarged IAC, a greater frequency of cochleovestibular anomalies and of IAC in children with congenital syndromes and SNHL have indeed been described (Fig. 1).

Case 5

Acoustic neuroma does not generally present in children as the typical cause of IAC enlargement except in the context of a neurofibromatosis type 2 (NF2). This pathology is inherited in an autosomal dominant fashion by means of mutation 22q12 in the suppressor gene of the NF2 tumour. Two different presentations have been described: “Wishart” type, with early onset and exitus in the fourth decade, and the “Gardner” type, slower and appearing after the second decade. Auditory affectionation, the mass effect and the involvement of different cranial nerves vary significantly. Only 30% of paediatric patients begin with auditory problems.

The patient in our series commenced with mass effect symptoms caused by the tumour of ventricle III. This was partially resected, with the neurosurgery service advising radiosurgery on the acoustic neuromas.

In contrast to congenital enlargement of the IAC, whether associated to cochleovestibular malformations or not, the presence of a tumour, dural ectasia or chronic hydrocephaly can produce progressive IAC dilatation secondary to the local increase in pressure. This form of progressive enlargement has also been described in NF1 from arachnoid enlargement. In dilated IAC, considered as a variant from normality, in contrast to that produced in neuromas, the cortical borders of the bone canal and of the falciform crest are preserved.
### Table 1: Enlarged Internal Auditory Canal not Related to Cochleovestibular Dysplasia.

<table>
<thead>
<tr>
<th>Patient</th>
<th>Sex</th>
<th>Age</th>
<th>Clinical presentation</th>
<th>Audiology</th>
<th>Image</th>
<th>Treatment</th>
<th>Diagnosis</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>M</td>
<td>2 y</td>
<td>In aetiological hearing loss study, radiologist recommended MRI due to enlarged IAC on CT</td>
<td>BAEP: no bilateral response</td>
<td>IAC≈8.1 mm No other alterations</td>
<td>Cochlear implant</td>
<td>Profound bilateral SNHL</td>
</tr>
<tr>
<td>2</td>
<td>M</td>
<td>1 y and 8 m</td>
<td>Language delay. Severe bilateral SNHL</td>
<td>BAEP: RE: 80 dB LE: 70 dB</td>
<td>IAC≈8.1 mm No other alterations</td>
<td>Hearing aids</td>
<td>Severe bilateral SNHL</td>
</tr>
<tr>
<td>3</td>
<td>M</td>
<td>6 y</td>
<td>Suspicion of hearing loss, cognitive delay. 2-year-old brother with severe SNHL</td>
<td>BAEP: RE: 95 dB LE: 70 dB</td>
<td>No other alterations</td>
<td>Hearing aids</td>
<td>Severe-profound bilateral SNHL. DFN 3: deletion gene POU3F4 in X-chromosome</td>
</tr>
<tr>
<td>5</td>
<td>F</td>
<td>12 y</td>
<td>Auditory assessment for NF2</td>
<td>PTA: RE scotoma and 45 dB in 2–4 KHz</td>
<td>MRI: bilateral VIII par schwannomas, posterior fossa meningioma and III ventricle, intramedullary ependymomas</td>
<td>No</td>
<td>SNHL unilateral scotoma 2–4 KHz NF2</td>
</tr>
</tbody>
</table>

AD, autosomal dominant; BAEP, brainstem auditory evoked potentials; CT, computed tomography; dB, decibels; EAC, external auditory canal; F, female; IAC, internal auditory canal; LE, left ear; M, male; m, months; MRI, magnetic resonance imaging; NF, neurofibromatosis; PTA, pure-tone audiometry; RE, right ear; SNHL, sensorineural hearing loss; TTD, transtympanic drains; y, year(s).

### Case 6

Over the last several years, deficiencies of the cochlear nerve have been postulated as a justification for some cases of SNHL. This can be defined as an absence of the cochlear nerve or a decrease in its width with respect to the other IAC nerves in the T2-weighted MRI sequences. The deficiency is beginning to be considered as the most frequent cause of unilateral congenital sensorineural hearing loss, according to findings in imaging techniques. It varies widely, both audiologically (from slight hearing losses to cophosis, uni- or bilateral, sometimes with auditory neuropathy phenotype) and radiologically (sometimes associated with malformations of the inner ear) as in its clinical associations (prematurity, syndromes, being idiopathic, etc.).

In computed tomography scan (CT), it is characterised by stenosis of the cochlear nerve or IAC canal. The patient in our series was diagnosed with unilateral SNHL at the age of 3 years; in the aetiological study, an enlarged IAC was revealed in the CT and the posterior MRI confirmed the cochlear nerve deficit (Fig. 2). In this case, there is no IAC stenosis and the hearing loss is severe, which are findings conflicting with the radiological report of "agenesis"; the responses in that ear possibly correspond with a smaller number of cochlear nerve fibres and its complete absence is unlikely. The presence of enlarged IAC together with cochlear nerve deficit does not seem to have an embryological or physiopathological relationship. As already commented, the only epidemiological association is the greater presence of IAC alterations in syndromic children, as is the case with this patient.

### Cases Associated With Cochleovestibular Malformations

The classic classifications of internal ear malformations have not considered in depth the concomitance with IAC...
alterations and their clinical implications, especially with respect to defects in the fundus. However, Zheng et al. include hearing loss and absence of modiolus in Mondini-like dysplasia type B (1.5–2 turns of the cochlea). In the nomenclature proposed by Sennaroglu, all the patients described with a type I incomplete partition (cochleovestibular cystic anomaly) would have an enlarged IAC. The IAC can be normal in the presence of other cochleovestibular alterations, and vice versa. The different embryological origins of the two structures can make this variability concordant. Nevertheless, epidemiologically, it seems that it is more common to find enlarged IAC associated with other labyrinthine malformations; this suggests that it is necessary to study the fundus carefully and to confirm complete cochlear partition.

Case 7
This case presents with the classic clinical picture of otogenic meningitis due to an anomalous communication between the middle ear and the subarachnoid space through a malformation of the inner ear. It can happen following otitis media from bacteria penetrating through a membrane in the intact round window or by means of fistula between the middle and inner ear. The presence of a gusher and the appearance of meningitis seem more likely in incomplete partition type I deformity than in cases of type II. Our case showed the 2 situations of greatest theoretical risk: an incomplete partition type I and a fistula in the stapes footplate (Fig. 3). It is important to advise the parents about the risk of meningitis and the need for early detection of its symptoms in cases of hearing loss with incomplete partition type I defects.

Case 8
What is called branchio-oto-renal syndrome refers to the association of branchial fistulas or cysts (50%), structural kidney alterations of varying severity (67%) and hearing loss (75%–93%). Various autosomal dominant inheritance patterns (EYA1 [8q13.3], SIX5 [19q13.3] and SIX1 [14q23.1]) and a prevalence of 1/40,000 have been described, representing 2% of the profound hearing losses in children. The symptoms vary widely, depending on the temporal bone malformations and those of the kidneys. Mandibular asymmetries, facial paresis and atresia of the nasolacrimal duct

<table>
<thead>
<tr>
<th>Table 2</th>
<th>Enlarged Internal Auditory Canal Associated With Cochleovestibular Dysplasia.</th>
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</thead>
<tbody>
<tr>
<td>Patient</td>
<td>Sex</td>
</tr>
<tr>
<td>8</td>
<td>M</td>
</tr>
<tr>
<td>9</td>
<td>M</td>
</tr>
</tbody>
</table>

AD, autosomal dominant; BAEP, brainstem auditory evoked potentials; dB, decibels; IAC, internal auditory canal; LE, left ear; M, male; MRI, magnetic resonance imaging; PTA, pure-tone audiometry; RE, right ear; SNHL, sensorineural hearing loss; TTD, transtympanic drains; y, year(s).
Figure 2  Cochlear nerve deficit, right ear (RE) (from the upper left, in clockwise direction: axial MRI: the right cochlear nerve is not identified in any cut. axial MRI, dilated IACs. Parasaggital MRI, IAC in left ear (LE): cochlear nerve component is identified in anterior–inferior, in contrast to RE, where it is not identified. The arrows indicate the position of the cochlear nerve).

have also been described, relating them with the spectrum of midfacial microsomnia. The hearing loss can be conductive (30%), mixed (50%) or sensorineural (20%), stable, progressive and/or fluctuating. The alterations most frequently found on imaging tests are stenosis or atresia of the external auditory conduct, ossicular anomalies, middle ear hypoplasia and, at the level of the inner ear, cochlear hypoplasia with incomplete partition type II, dysplasia of semicircular canals, dilation of vestibular aqueduct and bulbous IAC. A greater predisposition to present congenital cholesteatoma has also been found, which would be in agreement the malformations in embryological development of the middle ear structures derived from the first and second branchial arches.

Figure 3  Recurrent meningitis: vestibular side of the footplate with spontaneous perforation, origin of perilymphatic fistula. In MRI and CT: enlarged IAC and occupation of middle ear and cavity from previous mastoidectomy.
The patient in our series presents findings on the imaging tests that are typically related with this syndrome: hypoplastic cochlea with only a single turn, hypoplastic semicircular canals and enlarged IACs. Likewise, slight-moderate SNHL is also in agreement in the clinical context (Fig. 4).

Within genetic syndromic hearing losses in children, in some entities enlarged IAC with fundus defect, associated or not to other cochleovestibular malformations (Goldenhar, Apert, Patau, CHARGE), has been described, as well as in non-genetic syndromes such as congenital infection from cytomegalovirus. There have also been sporadic references to the Usher syndrome, a diffuse dilation of the subarachnoid spaces that extends along the cranial nerves, causing bone remodelling with enlarged IAC.

Case 9
In the differential diagnosis of the association of hypothyroidism and sensorineural hearing loss, various entities have to be considered. In Pendred syndrome (SCL26A4 gene), which presents postpuberal goitre and typically dilated vestibular aqueduct (sometimes associated with cochlear dysplasia), congenital hypothyroidism is rarely found. In endemic iodine deficit, SNHL is produced in 20%–50% of the cases. In syndromes thyroid hormone resistance, SNHL is related to the functioning of the beta receptor of thyroid hormone (required for normal development of the auditory system), but no structural alterations of this system are found in imaging tests. In the child that we describe in our series, it has been impossible up to now to confirm any of the disorders mentioned above by endocrinological or genetic studies, in spite of the phenotype.

In our series, the majority of the cases have no association with other cochleovestibular malformations (n=6 vs n=3).

The clinical implications of these findings may be useful in various circumstances: terminal prognosis (ruling out NF or evaluating meningitis risk), surgery (risk of gusher in cochlear implant), epidemiological (clinical presentation seems to depend more on concomitance with other inner ear alterations than on the isolated finding of enlarged IAC, so the presence of complete partition and fundus integrity should be checked in all cases of enlarged IAC and theoretical (justify the physiopathology and clinical development–progression, fluctuation –of some SNHLs).

Conclusions
The presence of enlarged IAC in children can be found in different clinical and audiological contexts, with a relevance that can vary from situations with life-threatening risk such as in recurrent meningitis, up to isolate hearing losses without any other associations.

Carrying out an appropriate audiological and aetiological differential diagnosis when faced with this finding can provide greater perisurgical safety and better knowledge of the physiopathology, clinical evolution and prognosis of some SNHLs.

Conflict of Interests
The authors have no conflict of interest to declare.

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