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ORIGINAL ARTICLE

Malformation of the Eighth Cranial Nerve in Children☆

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KEYWORDS
Cochlear nerve;
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Abstract
Introduction and objectives: Prevalence of congenital sensorineural hearing loss (SNHL) is approximately 1.5–6 in every 1000 newborns. Dysfunction of the auditory nerve (auditory neuropathy) may be involved in up to 1%–10% of cases; hearing losses because of vestibulocochlear nerve (VCN) aplasia are less frequent.

The objectives of this study were to describe clinical manifestations, hearing thresholds and aetiology of children with SNHL and VCN aplasia.

Methodology: We present 34 children (mean age 20 months) with auditory nerve malformation and profound HL taken from a sample of 385 children implanted in a 10-year period. We studied demographic characteristics, hearing, genetics, risk factors and associated malformations (Casselman's and Sennaroglu's classifications). Data were processed using a bivariate descriptive statistical analysis (P<.05).

Results: Of all the cases, 58.8% were bilateral (IIa/IIa and I/I were the most common). Of the unilateral cases, IIb was the most frequent. Auditory screening showed a sensitivity of 77.4%.

A relationship among bilateral cases and systemic pathology was observed. We found a statistically significant difference when comparing hearing loss impairment and patients with different types of aplasia as defined by Casselman's classification. Computed tomography (CT) scan yielded a sensitivity of 46.3% and a specificity of 85.7%. However, magnetic resonance imaging (MRI) was the most sensitive imaging test.

Conclusions: Ten percent of the children in a cochlear implant study had aplasia or hypoplasia of the auditory nerve. The degree of auditory loss was directly related to the different types of aplasia (Casselman's classification) Although CT scan and MRI are complementary, the MRI is the test of choice for detecting auditory nerve malformation.

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Oído interno;
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Malformaciones del octavo par en niños

Resumen
Introducción y objetivos: La prevalencia de la hipoacusia neurosensorial (HNS) congénita es aproximadamente de 1,5-6 de cada 1.000 nacimientos. La disfunción del nervio auditivo (neuropatía auditiva) puede estar presente hasta en el 1-10% de los casos de HNS en niños, siendo menos frecuentes aquellas pérdidas debidas a una aplasia o hipoplasia del nervio auditivo.

Los objetivos del estudio son describir la clínica, umbrales auditivos y etiología en una muestra de niños con HNS y aplasia o hipoplasia del octavo par.
Metodología: Presentamos 34 niños (edad media 20 meses) con malformación del nervio auditivo e HNS profunda de una muestra de 385 niños implantados en los últimos 10 años. Estudiamos las características demográficas, clínicas y malformaciones asociadas (clasificaciones de Casselman y Sennaroglu). Los datos fueron procesados usando un análisis estadístico descriptivo bivariante ($p < 0,05$).
Resultados: Un 58,8% fueron bilaterales (IIa/IIa y I/I fueron los más frecuentes). De los unilaterales el IIb fue el más frecuente. La sensibilidad del cribado auditivo fue de un 77,4%. Encontramos diferencias estadísticamente significativas entre el grado de hipoacusia y los distintos tipos de aplasia (clasificación de Casselman).

La sensibilidad de la TC fue del 46,3% y la especificidad del 85,7%. La RNM fue la prueba de imagen más sensible.
Conclusiones: Un 10% de los niños en estudio para un implante coclear tienen una aplasia o hipoplasia del nervio auditivo. El grado de pérdida auditiva está directamente relacionado con los distintos tipos de aplasia (clasificación de Casselman). Aunque la TC y la RNM son complementarias, la RNM es el test de elección para la detección de malformaciones del nervio auditivo.

Introduction

Congenital sensoneural hearing loss (SNHL) is one of the most common problems in newborns, affecting between 1.5 and 6 children out of every 1000 living newborns.[1] Although the cause of the problem may be located anywhere in the auditory pathway, it is well known that the most common site is the cochlear, whether this be due to congenital or acquired causes, and in particular at sensory hair cell level. Recent studies suggest that the dysfunction of the auditory nerve (auditory neuropathy) may be involved in 1%–10% of cases of SNHL in children, with auditory losses resulting from anatomical changes to the nerve (aplasias and hypoplasias) being less frequent.[2,3]

In our experience 10% of children in a cochlear preimplant study will present with an aplasia or hypoplasia of the auditory nerve and this finding has increased over time due to improvements in magnetic resonance imaging. However, and as we will describe below, radiologic findings regarding aplasia of the auditory nerve are not 100% proof of a total absence of auditory function.

Material and Methods

The sample included 34 children patients (mean age: 20.5±27.0 months) who had been diagnosed with SNHL due to aplasia or hypoplasia of the auditory nerve with or without associated vestibulocochlear nerve malformations. We based our diagnosis and study primarily on the MRI imaging findings in accordance with the techniques described by Casselman[4]: (T2) 3 Teslas MRI: slices of 0.4–0.7 mm perpendicular to the pathway of the facial nerve and of the eighth cranial nerve in the internal auditory canal and at the cerebellopontine angle level (parasagittal reconstruction) and volume reconstruction in the axial plane (three-dimensional Fourier transformation-constructive interference in steady state [3DFT-CISS]), and at the same time a routine axial T2 weighted brain imaging study to rule out central nervous system pathologies. The imaging study was completed with high-resolution CT of the temporal bone (axial plane performed in slices parallel to the infraorbitary line up to 0.3 mm thick. A 512×512 matrix was used and the images of each ear were separated with a field of vision of approximately 9 cm).

Hearing tests included: tone and/or speech audiometry (Affinity 2.0/Equinox 2.0), tympanometry (Impedance AZ 26 Audiometer), otoacoustic emissions (Madsen Accuscreen) and brainstem auditory and steady state evoked potentials (Navigator Pro Biologic: PEA and MASTER II).

Malformations of the auditory and vestibulocochlear nerves were grouped according to the Casselman[4] (Figs. 1–5) and Sennaroglu[5] classifications respectively. All data were processsed using bivariate statistical analysis (P<.05).