Auris Nasus Larynx xxx (2016) xxx–xxx

Contents lists available at ScienceDirect

Auris Nasus Larynx

journal homepage: www.elsevier.com/locate/anl

Audiologic and radiologic findings in cochlear hypoplasia

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ARTICLE INFO

Article history:
Received 12 July 2016
Accepted 19 December 2016
Available online xxx

Keywords:
Audiological evaluation
Inner ear malformation
Cochlear hypoplasia
Cochlear aperture
Cochlear nerve
Cochlear implantation

ABSTRACT

Objective: The aim of the current study is to evaluate audiologic and radiologic findings of cochlear hypoplasia which is a subgroup of inner ear malformations.
Methods: This study was a prospective clinical study and based on voluntary participation from cases with cochlear hypoplasia diagnosis. The study was conducted at Hacettepe University, Department of Otolaryngology, Head and Neck Surgery and Department of Audiology. Subjects were selected from an inner ear malformations database. Inclusion criteria were having cochlear hypoplasia for at least one ear. There were 66 subjects with an age range of 12 months and 60 years 5 months. For each subject, pure tone audiometry and tympanometry were applied according to chronological and cognitive age. And also, auditory brainstem response test was applied to when it is need. Subjects' radiologic results were reevaluated to confirm cochlear hypoplasia, cochlear nerve and cochlear aperture.
Results: Cochlear hypoplasia types were statistically significantly different in terms of HL degree. This difference was caused by cochlear hypoplasia type IV group being was statistically different from the other three groups. Like with degree of HL, cochlear hypoplasia groups were statistically different from other three groups in terms of type of hearing loss. Cochlear aperture and cochlear nerve status showed variation according to cochlear hypoplasia type but these differences were not statistically approved.
Conclusions: In the current study, incidence of cochlear hypoplasia was 23.5% in all inner ear malformation. With this study, it was seen that subtypes of cochlear hypoplasia showed variability in terms of degree and type of hearing loss and also cochlear aperture and cochlear nerve status. Especially cochlear hypoplasia type IV differs from other three cochlear hypoplasia types.

1. Introduction

Inner ear malformations (IEMs) are the cause of congenital sensorineural hearing loss in approximately 20% of hearing impaired children [1]. IEM is used as a general term for all cochlea types that differ from normal structures. Sennaroglu et al. classified IEM by using computerized tomography (CT) and magnetic resonance imaging (MRI) [1,2]. This latest classification includes eight groups; labyrinthine aplasia (Michel deformity), rudimentary otocyst, cochlear aplasia, common cavity, incomplete partition (IP) of the cochlea (IP), cochlear hypoplasia (CH), large vestibular aqueduct syndrome (LVAS) and cochlear aperture abnormalities (CA) [2]. All these groups show variability in terms of radiological findings [1,3,4]. The latest classification is summarized in Table 1.

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http://dx.doi.org/10.1016/j.anl.2016.12.002
0385-8146/

Table 1
Classification of inner ear malformations.

Name of the IEM	Inner ear malformations (IEM)							
	Labyrinthine aplasia (Michel deformity)	Rudimentary otocyst	Cochlear aplasia	Common cavity	Incomplete partition (IP) of the cochlea	Cochlear hypoplasia (CH)	Large vestibular aqueduct syndrome (LVAS)	Cochlear aperture abnormalities (CA)
Short explanations	The cochlea, vestibule, semicircular canals, vestibular and cochlear aqueducts are absent	Incomplete millimetric representations of otic capsule (round or ovoid in shape), without an internal auditory canal	The cochlea is absent	Cochlea and vestibule are represented by an ovoid or round structure. Cochlea and cochlear vestibular neural structures are present	External dimensions are similar to normal cochleae but the internal architecture is deficient. Three different types: (IP-I), incomplete (IP-II) and (IP-III)	External dimensions are reduced relative to the normal cochlea. According to internal architecture, four different types of CH: CH-I, CH-II, CH-III and CH-IV	An enlarged vestibular aqueduct in the presence of a normal cochlea, vestibule and semicircular canals	Cochlear aperture or cochlear nerve canal transmits the cochlear nerve from the Internal acoustic canal to the cochlea

Cochlear hypoplasia (CH), a subgroup of IEM, also has four different types identified according to CT results. Radiological views of CH types are shown in Fig. 1.

Cochlear hypoplasia type I (CH-I) is defined as a bud like cochlea (round or ovoid) arising from the internal acoustic canal (IAC). Internal architecture is severely deformed; no modiolus or interscalarsepta can be identified. There may or may not be a septum separating IAC from the cochlea. In the provided Fig. 1 (ISS), there is a thin septum separating the two structures.

Cochlear hypoplasia type II (CH-II) is defined as cochlea with smaller dimensions and without modiolus and ISS. But external architecture is similar to that of a normal cochlea and makes a wide connection with the existing IAC. The vestibular aqueduct is enlarged, and the vestibule is minimally dilated.

Cochlear hypoplasia type III (CH-III) has a shorter modiolus than normal and the cochlea has a reduced number of turns (<2 turns). Internal and external architecture are similar to normal cochlea, with smaller dimensions. The vestibule and semicircular canals are hypoplastic.

Finally, cochlear hypoplasia type IV (CH-IV) has a cochlea with approximately a normal sized basal turn and severely hypoplastic middle and apical turns. The labyrinthine segment of the facial nerve may be located anterior to the cochlea.

Fig. 2 shows a schematic view of CH types. CT and MR images and schematic view of CH types show difference. When the features of CH are evaluated histopathologically, it can be seen that there are very thin intersepta coming from the modiolus in schematic view. Unfortunately the resolution of HRCT and that of MRI are not precise enough for the present time, to detect these very rudimentary developments. Therefore, it is possible to evaluate these as completely cystic with the present day imaging modalities. But in future, we will most probably be able to detect these thin septa, with much developed imaging resolution.

It was known that cochlear aperture (CA) and cochlear nerve (CN) status also show variability across IEM types [1,3]. On MRI; the diameters, area, and signal intensity of the CN were measured and compared to the ipsilateral facial nerve. Also the width of the CA was measured on axial images and it was defined as 'stenotic' when the width was less than 1.5 mm [5,6].

Some studies have investigated audiological findings of IEM but not according to types of IEM.

The aim of the current study is to evaluate audiological and radiological findings of cochlear hypoplasia a subgroup of IEM. As described above, cochlear hypoplasia has four different types identified by Sennaroglu [1,2]. These CH groups are compared according to degree and type of hearing loss, status of CA and CN.

2. Materials and methods

The current study was conducted at Hacettepe University, Department of Ear-Nose-Throat and Department of Audiology. Hacettepe University's Non-invasive Ethical Committee approved this study (No: GO 14/195-30).

In our IEM database, there were 481 subjects when this study conducted and 113 of them have CH diagnosis. Inclusion criteria