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Original Article

Egyptian children with congenital sensorineural hearing loss candidates for cochlear implant: Is MRI evaluation necessary?

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ABSTRACT

Objectives: Evaluating the value of MRI for assessing congenital anomalies of the inner ear in children with congenital sensorineural hearing loss (SNHL) before cochlear implantation.
Patients and methods: This study included 56 children, suffering from severe to profound bilateral congenital SNHL. All included patients were potential candidates for cochlear implants. All patients were evaluated by MRI of the temporal bone.
Results: This study detected abnormalities in 27 (24.11%) ears out of 112 ears. We detected dilated vestibular aqueduct in 12/27 (44.44%) of abnormal ears, incomplete partition type-II in 8/27 (29.63%) of abnormal ears, cochlear hypoplasia in 8/27 (29.63%) of abnormal ears, semicircular canals hypoplasia in 2/27 (7.41%) of abnormal ears, Michel deformity in 2/27 (7.41%) of abnormal ears, common cavity deformity in 2/27 (7.41%) of abnormal ears and cochlear aplasia in 1/27 (3.7%) of abnormal ears.
Conclusion: MRI is a valuable tool in the evaluation of inner ear abnormalities in children with congenital SNHL. It allows for the detection of aplasia of the cochlea or the cochlear nerve which represent absolute contra-indications to a cochlear implant surgery.

1. Introduction

Children born with bilateral congenital sensorineural hearing loss (SNHL) can suffer long term consequences; since in addition to being deaf, they will fail to develop language skills. SNHL is caused by abnormalities of the inner ear, eighth cranial nerve or higher center in the brain. These may be inherited from the parents or may result as a sequel of intrauterine infection or a birth injury, yet, in many cases the underlying cause remains obscure [1].

Cochlear implants represent a source of hope for children with SNHL, allowing them to develop hearing abilities and acquire language communication potentials. Therefore, it is highly advisable to undergo the surgery as early as can be in candidate children [2].

That being said, it is of utmost importance to identify contra-indications to this surgery, which would result in poor post-operative outcomes. Conditions, such as absent cochlea or cochlear nerve, should absolutely contraindicate the procedure [3]. Many of such conditions are readily identifiable on imaging studies, which are an integral part for assessing those patients before undergoing the operation [4]. Therefore, radiologists should be acquainted with normal and abnormal imaging findings of the inner ear and temporal bone [5].

Recent developments in computed tomography (CT) and magnetic resonance (MR) imaging have greatly improved diagnosis and management of patient with petrous bone abnormalities by allowing more accurate preoperative diagnosis [6]. Discussions debating which imaging tool, either MRI or CT, is better for pre-procedural assessment of candidate children with SNHL, have been going on, each advocating pros and cons for either modality. Those in favor of CT debate the higher resolution and better visualization of the bony ossicles, canal for facial nerve, ossicular labyrinth and jugular bulb, which can be valuable for planning the surgical pathway [7–9]. Those debating for MRI discuss the better visualization of soft tissues, especially the cochlear nerve and the fluid filled inner ear structures. Lack of cochlear nerve visualization on MRI is related to unfavorable outcome after cochlear implantation [3]. Yet, MRI has longer exam durations, something to be considered when dealing with young children, and increased cost [10].

In this study, we aimed at evaluating the value of MRI for discovering congenital conditions that may affect the conduction or the outcome of cochlear implantation in Egyptian children, by describing our experience in a tertiary care centre.

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2. Patients and methods

This is a prospective study that was carried out at the Department of Radiology, in collaboration with the Department of Audiology, of our tertiary care hospital, during the duration from October 2015 to August 2016. We included 56 children, suffering bilateral congenital SNHL, which was graded in a range of severe to profound. Their age ranged from 1 to 14 years. All included patients were potential candidates for cochlear implants and an audiological battery of investigations to assess cochlear implant candidacy including tympanometry and hearing assessment according to age, including sound field audiometry, play audiometry, in addition to auditory brainstem response audiometry and otoacoustic emission testing. Radiological investigations were also done including MRI of the inner ear and temporal bone. Sedation was used to overcome artifacts resulting from movement of children on the scanner. We obtained approval of both the scientific and ethics committees of our institution. Informed written consents were signed from the patients' parents or legal guardians.

The work up of our candidate patients began with detailed history taking including perinatal, natal and post-natal history. The perinatal details included history of any maternal intake of drugs or exposure to radiation during pregnancy. In utero infection such as syphilis, rubella, toxoplasmosis, cytomegalovirus or herpes (sTORCH). Syndromes associated with SNHL, neurodegenerative disorders. Developmental history, including history of diminution of hearing and trials at correction using hearing aids, were included. Also, history of associated medical conditions and family history of consanguinity among parents, and of permanent childhood SNHL were put into consideration.

All children included in our study suffered bilateral congenital SNHL. Those who developed diminution of hearing after a certain acquired factor, and not congenital deafness, were excluded.

2.1. MRI imaging protocol

All MRI exams were performed on a 1 T Philips panorama MRI machine (Philips Medical Systems, Netherlands) using an 8-channel head coil. We included a high resolution 3D T2-weighted fast spin-echo sequence (DRIVE) of the temporal bone in the axial plane with a slice thickness of 0.7 mm and a 130 mm FOV. Para-sagittal oblique cuts were acquired in a plane perpendicular to the course of the nerves along the internal auditory canal (IAC). The fluid filled inner ear structures were MIP reconstructed. Both axial and coronal T1 temporal bone images and axial T2 brain images were acquired. The scan duration was approximately 30 min without intravenous contrast agents being used.

2.2. Analysis of the MRI images

MRI images were evaluated for malformations. Different parts of inner ear were studied including the cochlea, vestibule, semicircular canals, vestibular aqueduct, IAC and cochlear nerve. Images were also evaluated for any abnormalities present and any inner ear malformations were classified according to the classification described by Sennaroglu and Saatci [11] in the following manner:

Cochlear Malformations

(1) Michel deformity: completely absent cochlea and vestibule.
(2) Cochlear aplasia: completely absent cochlea, present vestibule.
(3) Common cavity anomaly: non-differentiated single cystic cavity that represents both the cochlea and vestibule.
(4) Cochlear hypoplasia: both the cochlea and vestibule are present and separate, yet are smaller in size than normal. A hypoplastic cochlea can look like a small bud that arises from the IAC.
(5) IP-I (cystic cochleovestibular anomaly): cystic-appearing cochlea lacking entire modiolus and cribriform area, large cystic vestibule.
(6) IP-II (Mondini deformity): the cochlea consists of 1.5 turns in which the middle and apical turns fuse to form a cystic apex, this is accompanied by a dilated vestibule and vestibular aqueduct.

Vestibular Malformations

They include Michel deformity, common cavity deformity, absent vestibule and dilated vestibule.

Semicircular canal malformations

They include absent, hypoplastic and enlarged.

IAC malformations

They are absent, narrow or enlarged.

Vestibular and Cochlear Aqueduct malformations

They are described as enlarged or normal.

We considered the cochlear nerve as hypoplastic when its diameter was less than the diameter of the adjacent facial nerve in the IAC, at its middle to lateral thirds [12]. If the cochlear nerve was not visualized on any plane of the MRI scan, we considered it as aplastic. If the least dimension of the IAC in the axial and parasagittal oblique planes was smaller than 3 mm, it was considered as stenotic [13]. Vestibular aqueduct diameter $\geq$2 mm at the operculum and/or $\geq$1 mm at the midpoint is considered enlarged [14].

All data was entered and analyzed using Microsoft office excel 2013. Descriptive statistical analyses including minimum, maximum, mean, standard deviation and percentages were done.

3. Results

A total of 56 children (112 ears) with bilateral congenital SNHL were radiologically evaluated with MRI of the temporal bone in this study. Their age ranged from 1 to 14 years, with a mean of 3.48 and standard deviation of 2.08. There were 29 male and 27 female patients in this study. Out of the 56 children, 42 (75%) were normal and 14 (25%) were abnormal. Thirteen children had bilaterally abnormal inner ear while 1 child had unilateral abnormal ear. Thus, this study detected abnormalities in 27 (24.11%) ears. The anatomic locations and types of the malformations is summarized in Table 1. Table 2 provides a classification of the malformations.

From the 13 patients with bilateral abnormalities, 12 had bilateral symmetrical abnormalities. This study showed only one patient with bilateral asymmetrical abnormalities, with cochlear aplasia and

Table 1
Summary of MRI findings.

Anatomic description	Number
Cochlea	
Normal	91
Incomplete partition type-II	8
Aplasia	3
Hypoplasia	8
Cystic cavity of cochlea and vestibule	2
Vestibule	
Normal	96
Aplasia	2
Enlarged	12
Cystic cavity of cochlea and vestibule	2
Semicircular canals	
Normal	99
Aplasia	11
Hypoplasia	2
Vestibular aqueduct	
Normal	98
Dilated	12
Aplasia	2
Internal auditory canal	
Normal	102
Stenosis	10
Cochlear nerve	
Normal	99
Aplasia	5
Hypoplasia	8