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Diagnostic value of magnetic resonance imaging in children with congenital sensorineural hearing loss

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ABSTRACT

This study aimed to investigate the value of Constructive Interference In Steady State (CISS) magnetic resonance imaging (MRI) assessment in evaluating the cranial nerve VIII, and the branches of this nerve for detection of the inner ear abnormalities. Ninety pediatric patients diagnosed with congenital sensorineural hearing loss (SNHL) between November 2006 and September 2008 were assessed by 3D-CISS MRI to evaluate the cranial nerve VIII with respect to congenital abnormalities of the inner ear. Of the 90 patients included in the study group, 44 were male (48.9%) and 46 were female (51.1%) at an age varying from 1 to 16 years old (5.97±4.04 years). The structures of 180 inner ears of 90 pediatric patients were assessed. Pathology was detected in 48 inner ears (26.6%) with any of the cochlear, vestibular, semi-circular canal and vestibular aqueduct abnormalities. Among 24 patients (26.6%), Michel deformity (1.66%), cochlear aplasia (4.98%), common cavity deformity (2.22%), cochlear hypoplasia (3.33%) and Mondini malformation (2.77%) was detected in 3, 9, 4, 6 and 5 inner ears, respectively. Internal auditory canal (IAC) assessment revealed dilatation in 11 inner ears, hypoplasia in 6, aplasia in 1 and vestibular aqueduct (VAD) dilatation in 9 inner ears. CISS MRI is an effective imaging technique for assessing the inner ear abnormalities, the presence of nerve VIII in patients with congenital sensorineural hearing loss. Thus, we believe CISS MRI imaging should be performed before any operation in all patients, who may be candidates for cochlear implantation.

1. Introduction

While the incidence of congenital sensorineural hearing loss (SNHL) varies among different societies, the rate is approximately 0.5-2.1 per 1000 deliveries (Pabla et al., 1991; Vartiqinen et al., 1997). Cochlear implant is an effective treatment method for cases with SNHL (Van den Broek et al., 1995; O'Donoghue, 1996). Radiologic assessments are guiding in detecting inner ear abnormalities, if any, in potential implant recipients, and determining the feasibility of operation and the type of implant to be used. The anatomy of major inner ear structures such as the facial nerve, and some conditions where implantation is contraindicated are evaluated by pre-operative radiologic assessments. Relevant radiologic assessments include computed tomography (CT), magnetic resonance imaging (MRI) and three-dimensional applications of these (Casselman et al., 1993; Dahm et al., 1993; Woolley et al., 1997; Lo, 1998; Batman et al., 2001; Incesulu et al., 2003; Miyasaka et al., 2010).

While CT provides a detailed assessment on bony labyrinth, it remains insufficient in imaging the membranous labyrinth. However, 80% of the patients with SNHL have membranous labyrinth abnormalities (Jackler et al., 1987; Sennaroğlu and Saatçi, 2002). Therefore, CT assessments fail to provide an adequate evaluation in a noticeable portion of the patients. This study investigated the value of 3D MRI investigation in detecting the inner ear abnormalities and the

Table 1. The examined sequences and assessment parameters of MRI

	TR/TE	FOV (cm)	NEX	Slice number	Slice thickening (mm)	Matrix	Examination time (second)
T1 weighted axial spin-echo	400/7.8	23X23	2	20	3	173X320	142
T2 weighted axial spin-echo	4480/108	23X23	2	21	5	199X384	114
T2 weighted coronal spin-echo	4270/108	23X23	2	20	3	199X384	108
CISS	13.48/6.74	23X23	1	88	0.6	192X256	359

FOV: Field of view; **NEX:** Number of excitations; **MRI:** Magnetic resonance imaging

value of Constructive Interference In Steady State (CISS) MRI in assessing the presence of cranial nerve VIII, and the vestibular and cochlear nerve branches.

2. Methods

90 pediatric patients <18 years old, who were diagnosed with sensorineural hearing loss between November 2006 and September 2008 through physical examination and audiological tests, were assessed by 3D-CISS MRI to screen the congenital abnormalities of the inner ear. Of the 90 patients included in the study, 44 were male (48.9%) and 46 were female (51.1%) at an age varying from 1 to 16 years old (5.97±4.04 years). Patients with a clinical history of a known traumatic, infectious, or toxic cause of hearing loss were excluded.

All assessments were performed using a head coil at the MRI device with a 1.5 T magnetic field strength (Magnetom, Symphony-Quantum, Siemens, Erlangen, Germany). While patients who were able to adjust did not receive pre-assessment sedation, patients who could not adjust due to age or mental insufficiency were administered IV Pethidin HCl of 5 mg/kg under monitorization before the assessment. No contrast material was administered during the assessment. The examined sequences and assessment parameters are given in Table 1. On T2-weighted coronal sections involving the whole brain, pathologies that may cause intracranial signal increase or additional congenital abnormalities were evaluated.

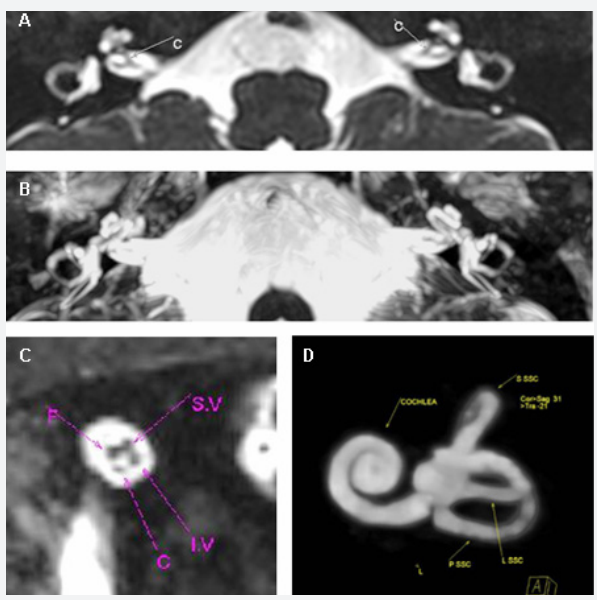

Fig. 1. Axial (A) and sagital (C) Constructive Interference In Steady State (CISS) magnetic resonance imaging (MRI) sequence show 8th cranial nerve in bilateral internal acoustic canal. 3-D volume rendering image of the inner ear (B and D).

All the images were transferred to the work station (Leonardo, Siemens Medical Solutions, Forcheim, Germany) for analysis after the procedure, and at the CISS MRI series, cranial nerve VIII in the bilateral internal acoustic canals (IAC) and the presence of cochlear branch were evaluated using minimum-intensity projection (MIP) images at axial and sagittal planes. On CISS MRI series, using the thin and thick MIP images from the sections at the inner ear level, and the 3D images obtained using the volume rendering technique, the general morphology and localization of the inner ear structures were examined with respect to abnormalities (Fig. 1).

Table 2. Sennaroglu Classification

Cochlear malformations

Cochlear malformations include the following:

1. Michel deformity. There is complete absence of all cochlear and vestibular structures.

2. Cochlear aplasia. The cochlea is completely absent.

3. Common cavity deformity. There is a cystic cavity representing the cochlea and vestibule, but without showing any differentiation into cochlea and vestibule

4. Cochlear hypoplasia. Malformation is further differentiated so that the cochlea and vestibule are separate from each other but their dimensions are smaller than normal. Hypoplastic cochlea resembles a small bud off the internal auditory canal (IAC).

5. Incomplete partition type I (IP-I). The cochlea is lacking the entire modiolus and cribriform area, resulting in a cystic appearance. This is accompanied by a large cystic vestibule.

6. Incomplete partition type II (IP-II) (Mondini deformity). The cochlea consists of 1.5 turns, in which the middle and apical turns coalesce to form a cystic apex, accompanied by a dilated vestibule and enlarged VAD.

Vestibular malformations

Vestibular malformations include Michel deformity, common cavity, absent vestibule, hypoplastic vestibule, and dilated vestibule.

Semicircular canal malformations

Semicircular canal malformations are described as absent, hypoplastic, or enlarged.

Internal auditory canal malformations

Internal auditory canal malformations are described as absent, narrow, or enlarged.

Vestibular and cochlear aqueduct findings

Vestibular and cochlear aqueduct abnormalities are described as enlarged or normal.

The inner ear abnormalities found in the patients enrolled in the trial were classified using the Sennaroğlu Classification (Sennaroğlu and Saatçi, 2002) (Table 2).

Definitions

IAC dilatation was defined 2.0 mm or more widening of any portion of the IAC (Fraser and Carter, 1975).

Vestibular aqueduct dilatation was defined diameter of >1.5 mm in the mid-portion of the descending hind limb (Reardon et al, 2000).

3. Results

The structure of 180 inner ears in 90 pediatric patients was analyzed. Pathology was detected in 48 inner ears (26.6%) with any of the cochlear, vestibular, semi-circular canal and vestibular aqueduct abnormalities. Cochlear abnormality was detected in 27 inner ears (15%) of 15 patients (16.6%) (12 of these 15 patients were bilaterally detected). Among 24 patients (26.6%), Michel deformity (1.66%), cochlear aplasia (5.0%), common cavity deformity (2.22%), cochlear hypoplasia (3.33%) and Mondini malformation (2.77%) was detected in 3, 9, 4, 6 and 5 inner ears, respectively (Fig. 2, 3). In 21 inner ears with any of the vestibular, semi-circular canal, IAC and vestibular aqueduct abnormalities (11.6%), normal cochlear development was observed to be completed. None of the patients experienced an incomplete partition type-I abnormality (Table 3).

Given the distribution of the detected vestibular abnormalities, no abnormality was detected in 166 inner ears whereas vestibular agenesis was observed in three inner ear structures with Michel deformity. A dilated vestibule was seen in one inner ear with cochlear hypoplasia, one inner ear with cochlear aplasia, five inner ears with Mondini deformity and one inner ear with common cavity deformity. While 11 patients had agenetic semi-circular canals, 20 patients were found to have dysgenesis (Table 3).

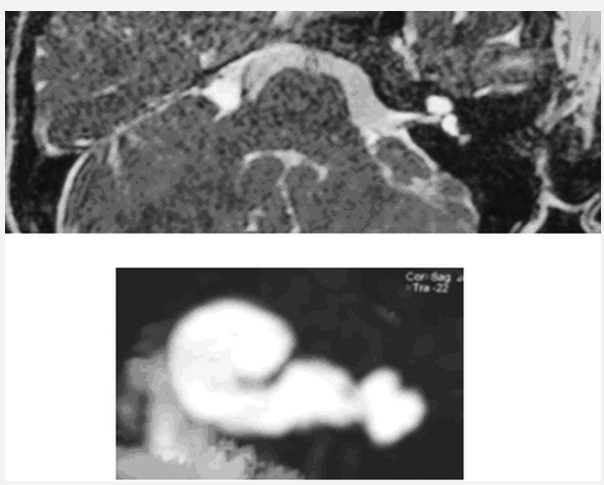

Fig. 2. Axial (A) Constructive Interference In Steady State (CISS) magnetic resonance imaging (MRI) sequence and (B) 3-D volume rendering images show Michel deformity on right side and Mondini malformation on the left

IAC assessment revealed dilatation in eleven inner ears, hypoplasia in six, and aplasia in one inner ear while 162 inner ears were considered normal (Table 3). In one inner ear with Michel deformity, the IAC was aplasic while six inner ears had a hypoplasic IAC, including one with mild hypoplasia. Among six inner ears with IAC hypoplasia, two had cochlear aplasia, two had cochlear hypoplasia and one had common cavity deformity. While one of the ears had a completely normal inner ear structure, the IAC was found to be hypoplasic. Among eleven inner ears, there were three cases of cochlear hypoplasia, two cases of Mondini deformity, two cases of Michel deformity and two cases of cochlear aplasia accompanied by IAC dilatation and two inner ear structures were normal.

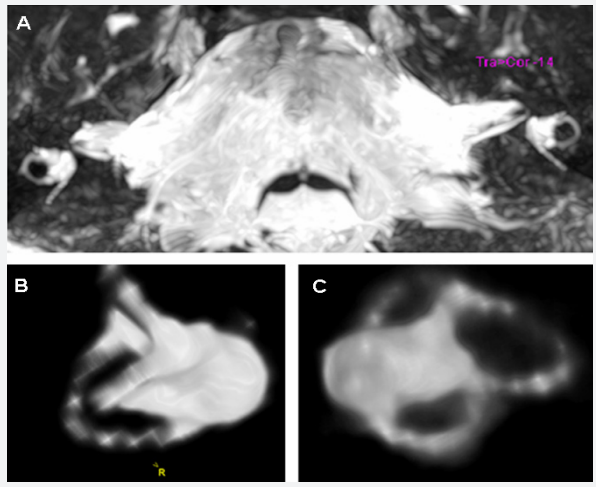

Fig. 3. Axial (A) Constructive Interference In Steady State (CISS) magnetic resonance imaging (MRI) sequence and (B, C) 3-D volume rendering images of the bilateral cochlear aplasia.

Assessment of the vestibular aqueduct (VAD) structures revealed dilatation in nine inner ears, while 171 patients were considered normal (Table 3). One of these patients had cochlear aplasia, one had cochlear hypoplasia, three had Mondini deformity while four didn't have any pathology in the inner ear.

In 132 inner ears (73.3) evaluated in the study, the membranous labyrinth formation and the IAC were considered normal. Table 3 summarizes the CISS MRI imaging results from 24 patients with several abnormalities.

4. Discussion

Cochlear implant is an organ prosthesis that is placed inside the inner ear and stimulates the auditory nerve electrically. The development of new treatment methods such as cochlear implant and Auditory brainstem implant (ABI) has shown promise for pediatric patients with severe sensorineural hearing loss in the recent years (Witte et al., 2003). In addition, radiologic imaging techniques have gained a wide area of usage in the assessment of the pre-operative treatment choices and post-operative follow-up of the cochlear implant candidates (Witte et al., 2003).

The inner ear is made up of two separate nested structures called the bony labyrinth and the membranous labyrinth. Approximately 20% of the cases with sensorineural hearing loss experience bony labyrinth abnormalities (Jackler et al., 1987). The remaining 80% have a normal labyrinth structure while exhibiting pathologies in the membranous labyrinth or at the cellular level (Sennaroğlu and Saatçi, 2002). CT is considered as the first-choice imaging method in the clinical practice for these patients due to the inexpensive and fast scanning as well as its advantages in evaluating the bony labyrinth. However, the cranial nerve VIII and the inner ear membranous labyrinth structures can only be assessed by MRI. MRI yields perfect anatomic details in structures composing the membranous labyrinth filled with liquid via high soft tissue resolution. The ability to establish axial, sagittal, coronal images and 3D images may offer advantages in the diagnosis and follow-up of children with sensorineural hearing loss. In a study by Zhu et al. (2002) 3D-CT and

Table 3. The evaluation of inner ear in patients with SNHL

	Inner ear in Patients with SNHL (n=180)	
	n	**%**
Cochlear abnormalities		
Cochlear aplasia	9	4.98
Cochlear hypoplasia	6	3.33
Mondini malformation	5	2.77
Common cavity deformity	4	2.22
Michel deformity	3	1.66
Incomplete partition type-I abnormality	0	2.77
Normal	153	85
Vestibular abnormalities		
Normal	166	92.2
Dilatation	11	6.1
Agenesis	3	1.6
Semi-circular canal abnormalities		
Normal	149	82.7
Dysgenesis	20	11.1
Agenesis	11	6.1
Internal acoustic canal abnormalities		
Normal	162	90
Aplasia	1	0.5
Dilatation	11	6.1
Hypoplasia	6	3.3
Vestibular aqueduct abnormalities		
Normal	171	95
Dilatation	9	5

Internal acoustic canal dilatation was defined 2.0 mm or more widening of any portion of the internal acoustic canal (Fraser and Carter, 1975).
Vestibular aqueduct dilatation was defined diameter of >1.5 mm in the mid-portion of the descending hind limb (Reardon et al., 2000).
SNHL: Sensorineural hearing loss

3D-MRI reconstruction techniques were compared in the pre-operative assessment of the patients, who were to receive cochlear implant, and 3D-MRI reconstruction techniques were detected to show the inner ear structures better. Another study by Miyashita et al. (2000) demonstrated that cochlear axis and the turns could be imaged thoroughly on the 3D reformatted images obtained from the MIP images. In our trial, thin MIP, thick MIP, and 3D images using VR technique have been obtained in all patients with cochlear abnormality. A 3D virtual reality was established for the membranous labyrinth structures by obtaining 3D reformatted images, using a 0.6-mm section thickness, thereby facilitating the definition and classification of the abnormalities and helping the surgeon to plan the operation appropriately by reviewing the 3D inner ear structures before the operation.

The absence of vestibulocochlear nerve is a contraindication for cochlear implantation. While CT can not demonstrate the vestibulocochlear nerve and cochlear branch, MRI may prove the presence of the nerve inside the IAC. An acoustic canal narrower than 2 mm as detected on CT is a key clue for vestibulocochlear nerve hypoplasia or aplasia, and such patients definitely need to be evaluated by MRI. Bamiou et al. (2001) investigated the narrow IAC diameter accompanying the absence of cochlear nerve. A patient was detected to have cochlear nerve hypoplasia with a normal acoustic canal diameter. In all the patients with cochlear nerve hypoplasia detected in the study, IAC narrowness accompanied the hypoplasia. In a trial by Morris et al. (2000) a patient with a normal IAC diameter on CT was found to have agenetic cochlear nerve as shown by MRI. All this data suggest that implant candidates should be evaluated by MRI in the pre-operative period so that isolated cochlear nerve agenesis that is not detected on CT is not omitted. In our study, two patients with a hypoplastic IAC and undetectable cochlear nerves were referred to brain stem implantation. A patient with Michel deformity was found to have acoustic canal aplasia. In one patient with mildly hypoplasic unilateral IAC, bilateral cochlear nerves were demonstrated; however, brain stem implantation was recommended since the patient had bilateral cochlear aplasia.

Pre-operative assessment via MRI is a must in potential cochlear implant recipients to assess the suitability of the implant and determine the type of the implant. Considering that the patient group is a pediatric group, MRI, as a non-radiation containing, non-invasive imaging method, should be the first-choice radiologic modality. Since temporal CT still maintains its superiority in assessing the bony labyrinth, ossicle chain and secondary bone changes, we believe that the combination of thin-slice CT and CISS MRI that evaluates the presence of the cochlear nerves and the structures originating from the membranous labyrinth would provide the optimum imaging.

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