The Journal of Laryngology & Otology
October 2002, Vol. **116,** pp. 804–810

Magnetic resonance imaging versus computed tomography in pre-operative evaluation of cochlear implant candidates with congenital hearing loss

Levent Sennaroglu, Isil Saatci*, Ayse Aralasmak*, Bulent Gursel, Ergin Turan

Abstract

Recent reports indicate that the cochlear nerve may be absent in some cases of congenital sensorineural hearing loss. The aim of this prospective study was to determine the incidence of cochlear nerve anomaly in cochlear implant candidates with congenital hearing loss using magnetic resonance imaging (MRI). Twenty-seven patients with congenital profound bilateral sensorineural hearing loss who were being evaluated for the cochlear implant procedure were studied.

These patients had high-resolution computerized tomography (CT), through the petrous bone in axial sections. MRI examinations consisted of T1 and turbo spin echo (TSE) T2-weighted 3 mm axial images, and additional 3D Fourier Transform T2-weighted TSE sequences obtained on three different planes (axial, perpendicular and parallel to the internal auditory canal (IAC) i.e. oblique sagittal and coronal, respectively) for the purpose of cochlear nerve demonstration. Results showed that all of the 14 patients with normal CT of the temporal bone, had four distinct nerves in the distal part of the IAC on TSE-MRI. Thirteen patients demonstrated various bony malformations of the cochleovestibular system on CT. MRI revealed the absence of the cochleovestibular nerve in four patients where the IAC was very narrow or completely absent on CT. One patient with severe Mondini malformation who had an enlarged IAC demonstrated an isolated absent cochlear nerve.

Key words: Cochlear Implants; Hearing Loss; Sensorineural; Cochlear Nerve; Magnetic Resonance Imaging

Introduction

Pre-operative radiographic evaluation of cochlear implant candidates is clinically important in deciding whether the patient is suitable for implantation, and choosing the side of implantation. High resolution computerized tomography (CT) and magnetic resonance imaging (MRI) are the two principal radiological investigations for cochlear implantation. There is still a continuing debate on which method should be used in the pre-operative evaluation of these patients. While some authors prefer CT,[1] some favour MRI[2] and some use both CT and MRI[3] before cochlear implantation.

CT scan gives excellent bone detail of the temporal bone.[1] Bony malformations of the otic capsule, and modiolus deformity can be demonstrated clearly on CT. The diameter of the internal acoustic canal can be measured accurately. Also ossification and fracture lines can be identified easily on CT. In addition, pneumatization of the mastoid and facial nerve position provide valuable information to the surgeon. However, CT lacks detail regarding neural structures, inner ear fluid and fibrosis. MRI, on the other hand, is superior to CT in the demonstration of nerves in the internal auditory canal, retrocochlear pathologies and membranous inner ear pathologies which result in characteristic fluid changes. However, MRI lacks information about the bony structures, particularly the course of the facial canal.

Recently, Casselman *et al.*[4] demonstrated the congenital absence of the cochlear nerve on MRI. Cochlear implantation in a patient with an absent cochlear nerve would be a catastrophe for the patient and the cochlear implant team. Therefore, it is imperative to diagnose this pre-operatively and inform the family. Maxwell *et al.*[2] and Gray *et al.*[5] reported two cases of implant failure due to an absent cochlear nerve in patients with a narrow IAC. Both groups had to explant the device. This topic formed the aim of our study where we used TSE-MRI in order to determine the incidence of cochlear nerve anomaly in cochlear implant candidates with congenital hearing loss.

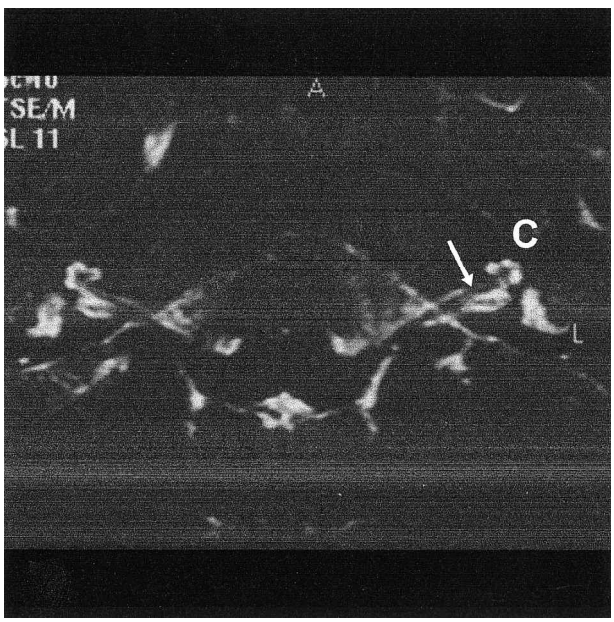
From the Departments of Otolaryngology – Head and Neck Surgery and Radiology*, Hacettepe University, Turkey.
Accepted for publication: 21 May 2002.

Materials and methods

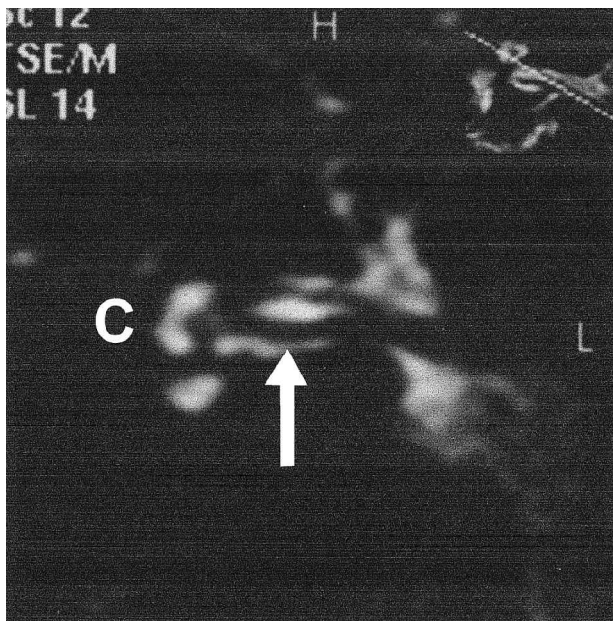
This study was carried out in the departments of Otolaryngology – Head and Neck Surgery and Radiology in Hacettepe University. Patients with profound bilateral sensorineural hearing loss who were being evaluated for cochlear implant procedure have been the subjects of this investigation. There were 27 patients, 14 male and 13 female. Their ages ranged from three to 26 (mean 11 years).

They had thorough ENT examination. Only patients with congenital hearing loss were included in this study. Patients with acquired hearing loss from such causes as viral diseases, meningitis, progressive hearing loss, ototoxicity and temporal bone fracture were excluded. All underwent complete audiological evaluation. All patients had bilateral profound sensorineural hearing loss.

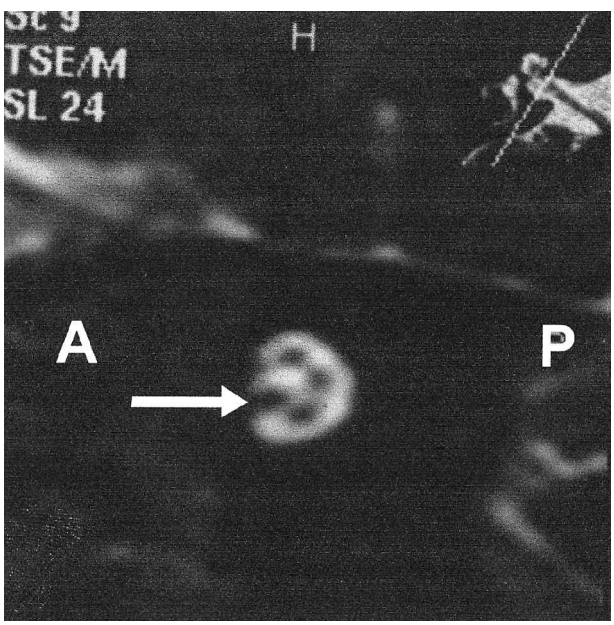
The patients had high-resolution CT, with contiguous 1 mm-thick images obtained through the petrous bone in axial sections. MRI examinations were performed on 0.5 T equipment (Philips 0.5 T Gyroscan NT System). Following scout views, T1 and TSE T2 weighted 3 mm axial images, and additional 3D Fourier Transform T2-weighted TSE sequences (TR/TE = 4000/250 msec) were obtained on three different planes (axial, perpendicular and parallel to IAC i.e. oblique sagittal and coronal, respectively) (Figures 1(a), 1(b), 1(c)) were obtained



(a)



(c)



(b)

FIG. 1

Normal MRI findings from 3D TSE T2 weighted sequence (C = cochlea, white arrow = cochlear nerve) (a) Transverse section parallel to internal acoustic canal; (b) Oblique sagittal section perpendicular to cochlear nerve (A = anterior, P = posterior); (c) Oblique coronal section parallel to the cochlear nerve.

https://doi.org/10.1258/0022215026029361 9 Published online by Cambridge University Press

TABLE I
COMPUTERIZED TOMOGRAPHY FINDINGS IN PATIENTS WITH MALFORMATIONS

1	Bilateral common cavity deformity with slightly enlarged IAC
2	Common cavity deformity with very narrow IAC on the left (Figure 2a) common cavity deformity with narrow IAC on the right
3	Common cavity deformity on the left side, normal IAC Mondini malformation on the right, normal IAC
4	Common cavity on the left side, wide IAC cochlear aplasia with dilated vestibule on the right, wide IAC
5	Mondini deformity on the left, normal IAC hypoplastic cochlea on the right, normal IAC
6	Bilateral Mondini deformity with enlarged IAC
7	Hypoplastic cochlea and vestibule with short IAC on the left side hypoplastic cochlea, absent vestibule with short and narrow IAC on the right (Figure 3a)
8	Hypoplastic cochlea, absent vestibule, narrow IAC on the left side severe Mondini malformation, enlarged IAC on the right
9	Mondini malformation, enlarged IAC, on the left side Mondini malformation, enlarged IAC, on the right side (Figure 4a)
10	Common cavity, enlarged IAC on the left severe Mondini malformation, enlarged IAC on the right
11	Mondini malformation, enlarged IAC on the left Mondini malformation, enlarged IAC on the right
12	Mondini malformation, enlarged IAC on the left Mondini malformation, enlarged IAC on the right
13	Michel deformity, absent IAC on the left Michel deformity, absent IAC on the right

since reconstruction images may not be satisfactory for the purpose of cochlear nerve demonstration. 3D T2-weighted TSE sequences included 1.2 mm thick slices were 0.6 mm overlapping, being in two separate stacks for oblique coronal and sagittal images. Field of view (FOV) was 130 mm with rectangular FOV of 75 per cent for axial and 160 mm with rectangular FOV of 60 per cent for the other two sequences.

Patients were divided into two groups:

(1) Membranous malformations: patients with normal CT findings (normal cochlea, vestibule, internal auditory canal (IAC), vestibular aqueduct and cochlear aqueduct).

(2) Bony malformations: patients with bony inner ear malformations demonstrated by CT. In this group of patients the classification of Jackler *et al.*[6] was used. These malformations are Michel deformity, cochlear aplasia, cochlear hypoplasia, common cavity and Mondini deformity. Deformities of the IAC were classified as normal, enlarged, narrow or absent.

CT was reviewed for malformations of the bony otic capsule and internal auditory canal. On TSE-MRI, neural structures (cochlear, facial, superior vestibular and inferior vestibular nerves) in the internal auditory canal were examined.

Results

There were 14 patients with normal CT of the temporal bone, i.e. with no bony malformation involving the cochlea, modiolus, vestibule, IAC, vestibular or cochlear aqueducts. In a TSE-MRI of these patients four distinct nerves in the distal part of the IAC were identified, and no abnormality of inner ear structures was present (Figure 1).

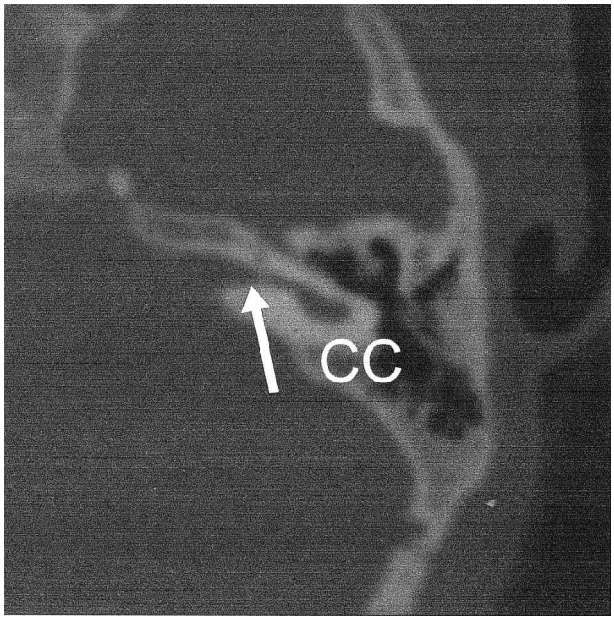
Thirteen patients demonstrated various bony malformations of the cochleovestibular system on CT. TSE-MRI gave additional information in these patients. CT findings are summarized in Table I. Altogether seven ears (27 per cent) demonstrated absence of the cochlear nerve. In six ears the cochleovestibular nerve was absent with a narrow or absent IAC. In one patient with severe Mondini malformation the cochlear nerve branch was absent.

In the group with inner ear malformations TSE-MRI revealed the absence of the cochlear nerve in patients 2, 7, 8 and 13 where the IAC was very narrow or completely absent on CT. In patient 2, IAC was very narrow on both sides (Figure 2(a)). There was no cochleovestibular nerve on the right side. On the left two nerves were visible in CPA but there was no nerve entering the common cavity (Figures 2(b), 2(c), 2(d)). Patient 7 had a short and narrow IAC on the right where no cochleovestibular nerve was present (Figures 3(a), 3(b), 3(c)). The facial nerve had a separate channel. On the left side a rudimentary cochleovestibular nerve was identified. In patient 8 there was narrow IAC and hypoplastic cochlea on the right side. Again the cochleovestibular nerve was absent on this side. Patient 13 had Michel deformity and absent IAC on both sides. Therefore, the cochleovestibular nerve was absent bilaterally in this patient.

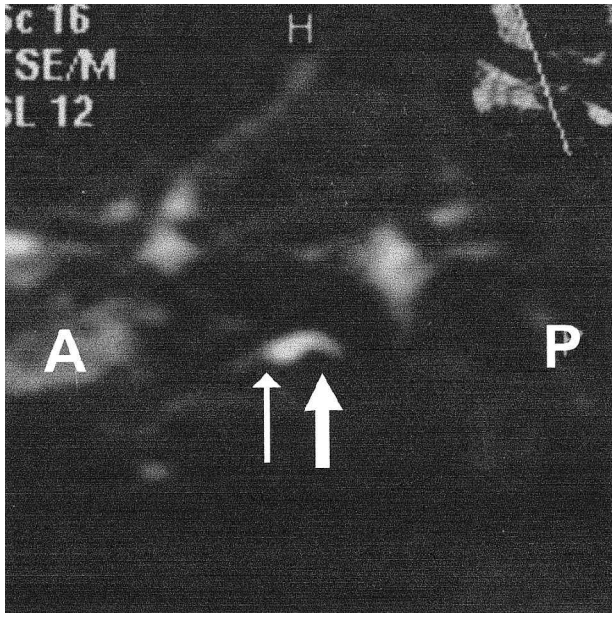
There was only one patient who had an isolated cochlear nerve absence. This patient (9) had severe Mondini malformation (Figure 4(a)) with an enlarged IAC. A TSE-MRI section perpendicular to the IAC just before the beginning of malformed cochlea showed only three nerves (facial, superior and inferior vestibular nerves) in the IAC (Figure 4(b)). The cochlear nerve, which should occupy the space anterior and inferior to the IAC, was absent.

In patients 1, 3 and 4 only two nerves (probably the facial and common cochleovestibular nerves) could be demonstrated in MRI. In all the remaining patients all four nerves in the IAC could be demonstrated.

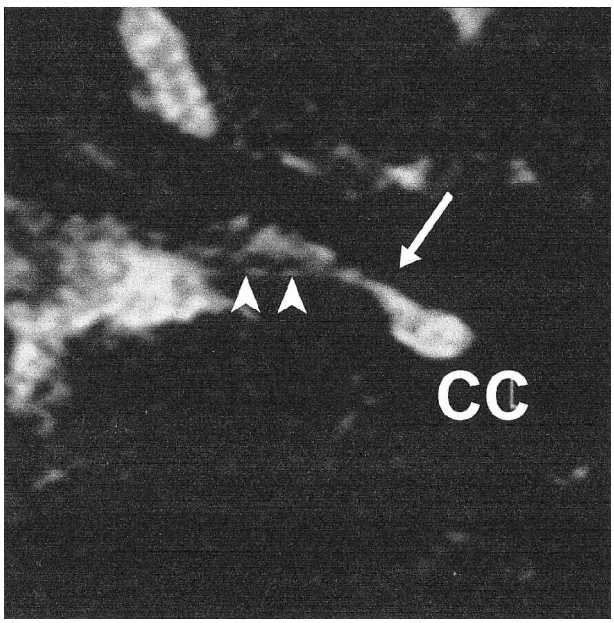
https://doi.org/10.1258/00222150260293619 Published online by Cambridge University Press



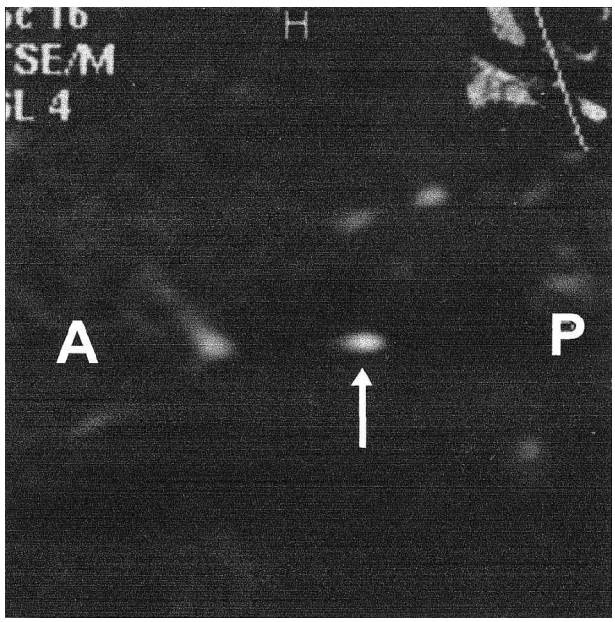
(a)



(c)



(b)



(d)

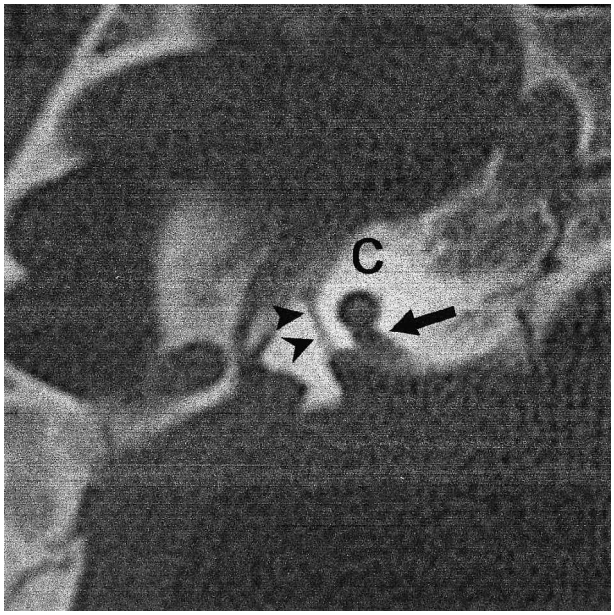
FIG. 2

Patient 2 with narrow IAC and common cavity. (a) Axial CT image demonstrating narrow IAC (white arrow) and common cavity deformity (CC); (b) Transverse MR image showing cochleovestibular nerve in the proximal IAC (white arrowheads), but no nerve in the distal IAC (white arrow) entering the cavity (CC); (c) Oblique sagittal MR image perpendicular to left IAC in the proximal part of the canal showing facial nerve (thin white arrow) and cochleovestibular nerve (thick white arrow); (d) Oblique sagittal MR image perpendicular to left IAC in the distal part of the canal showing absence of any neural structure within the IAC (white arrow).

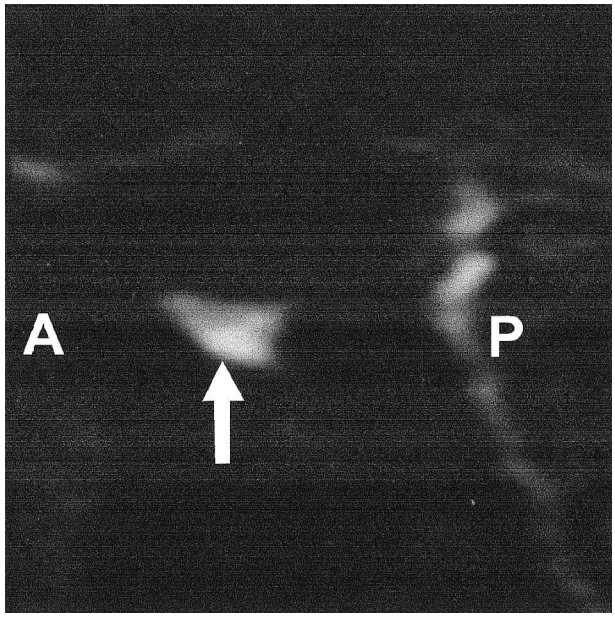
Discussion

The aim of this study was to investigate the isolated absence of the cochlear nerve in the presence of normal CT. Shelton *et al.*[7] pointed to the possible association between a narrow IAC on CT and the possibility of absent vestibular or cochlear nerves. Occasional case reports by Gray *et al.*[4] and Maxwell *et al.*[2] which reported an absent cochlear nerve, had a narrow internal auditory canal. On the contrary, Casselman *et al.*[4] in their report of seven cases with absence or hypoplasia of the vestibulocochlear nerve, had one patient with normal CT and an absent cochlear nerve on MRI. Morris *et al.*[8] also reported a case with an absent cochlear nerve where the IAC had normal dimensions. It is very rare to encounter an absent cochlear nerve in the presence

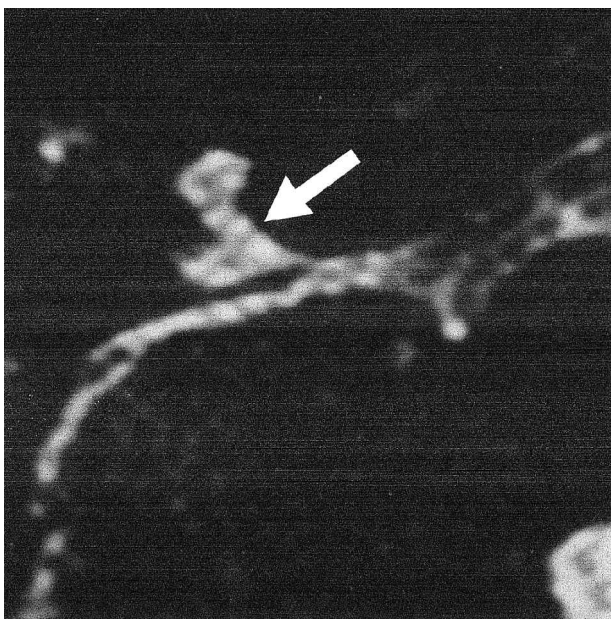
https://doi.org/10.1258/0022215026029361 9 Published online by Cambridge University Press



(a)



(c)



(b)

FIG. 3

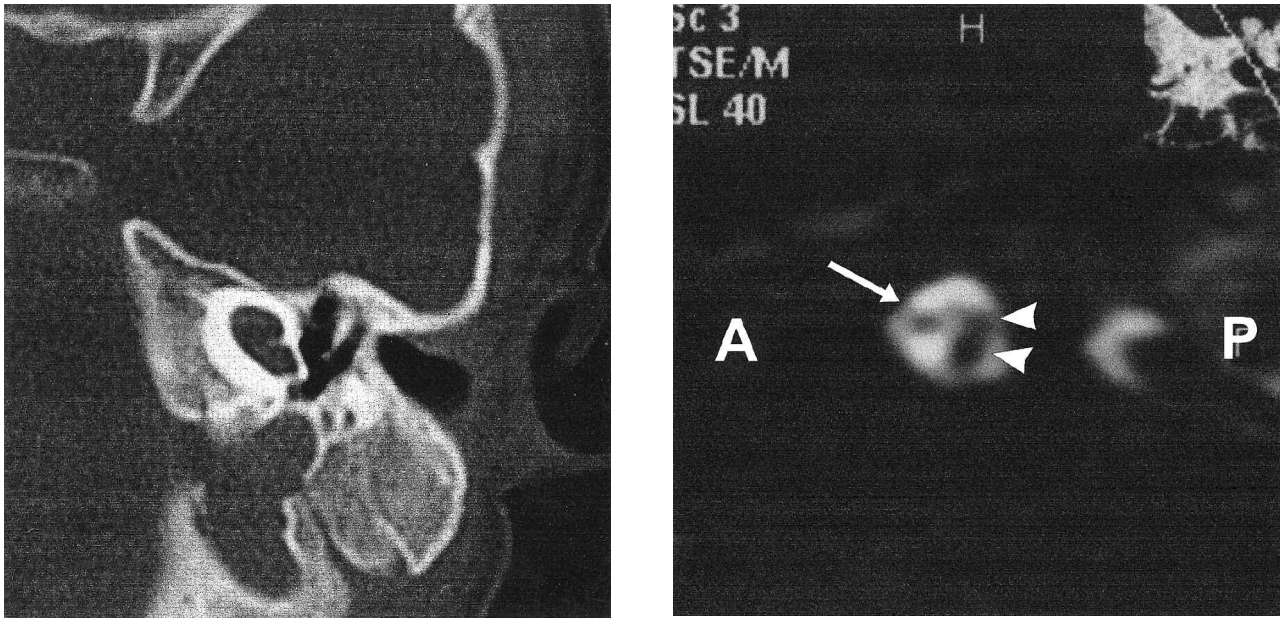
Patient 7 with short and narrow IAC. (a) Axial CT image demonstrating short and narrow IAC (black arrow) and hypoplastic cochlea (C). Note the separate canal for the facial nerve (black arrowheads) MR images parallel (b), and perpendicular (c) IAC, disclosing no neural structure within the IAC (A = anterior, P = posterior).

of normal CT findings. Our findings in the first 14 patients who had normal inner ear and internal auditory canal findings on CT demonstrated normal cochlear, vestibular (superior and inferior) and facial nerves in the IAC. Therefore, in our series the isolated absence of the cochlear nerve was not detected in patients with congenital hearing loss who had normal temporal CT findings. Recently, Ellul *et al.*[9] compared CT and MRI in cochlear implants candidates and it can be concluded from their cases that all patients with congenital hearing loss had normal cochlear nerves as identified with FSE-MRI.

What should be the radiological investigation of cochlear implant candidates? The controversy still continues. The main reason for radiological work-up is to detect cochlear aplasia and a very narrow IAC, that are the two radiologic contraindications to cochlear implantation. According to Lo[1] CT should be the primary investigation in cochlear implant preoperative radiological assessment and MRI should be reserved for special occasions. He stated that a narrow IAC less than 2.5 mm may be associated with absent cochleovestibular nerve which is a contraindication to implant surgery. He argued that few surgeons would be willing to replace CT with MRI, which adds cost to an expensive procedure, and requires sedation and monitoring in children.

On the contrary, Maxwell *et al.*[2] reported that they used MRI as the primary investigation method and reserved CT for special situations. They presented a

https://doi.org/10.1258/0022215026029361 9 Published online by Cambridge University Press



(a) (b)

FIG. 4

Patient 9 with bilateral severe Mondini malformation. (a) axial CT image demonstrating incomplete partition in the cochlea; (b) oblique sagittal section perpendicular to internal auditory canal showing only three nerves (white arrow = facial nerve; white arrowheads = superior and inferior vestibular nerves) with no cochlear nerve branch (anterior and inferior part of the IAC).

case with aplasia of a cochlear nerve. The patient had a narrow IAC on CT and cochlear nerve aplasia at the same side on MRI. Because of the failure of the implant in this patient they came to the conclusion that MRI should be the primary radiological investigation. We believe that, as the patient had a narrow IAC, this was already an indication for MRI. Ellul *et al.*[9] also suggested FSE-MRI should be the initial imaging study before implantation and came to the conclusion that FSE-MRI should replace CT. They stated that FSE-MRI is very good at the demonstration of the cochlear nerve and determining the patency of the cochlea. This technique, however, cannot show the course of the facial canal. Therefore, in the presence of a complex ear malformation they perform CT to trace the course of the facial nerve. Additionally they are of the opinion that in this way the patient is prevented from an unnecessary radiation dose. As can be seen from their report, they did not demonstrate an absent cochlear nerve in congenital cases. It is interesting to note that they had absent cochlear nerve in a patient with profound hearing loss following a viral infection. They pointed out that this was due to an acquired degeneration of the cochlear nerve following a viral illness.

Arriaga *et al.*[3] in their 13 case-review diagnosed one case with inner ear fibrosis, one vestibular neuroma and disproved a suspected fibrosis with MRI. They pointed out that as cochlear implantation is a contraindication to future MRI, the preoperative evaluation is the last opportunity for MRI in a patient with vestibular schwannoma. Also by reducing the examination to a single fast spin-echo T2-weighted sequence the cost of this examination is decreased. Therefore, this group preferred to use both CT and MRI together. Frau *et al.*[11] stated that they preferred CT as the method of investigation and argued that fibrosis undetected by CT usually did not appear to compromise the surgical access.

Recently Thai Van *et al.*[12] reported a case where the patient had bilateral narrow IACs with normal labyrinth. 3DFT-CISS showed only a small nerve at the level of IAC bilaterally. This was considered as the facial nerve. When the patient showed improvement in vocalization during the follow up, functional MRI (FMRI) was obtained. This showed activation in an area corresponding to Brodmann's area. They concluded that spatial resolution limits of MRI may have resulted in the failure of visualization of very thin acoustic fibres. According to the authors another possible explanation for this might be a common nerve carrying facial, acoustic and vestibular fibres. FMRI appears to be technique of choice in investigating the auditory cortex. In patients such as these promontory ABR is also very helpful for demonstrating the neural pathway.

In our cases absence of cochlear nerve is correlated with a narrow or absent internal auditory canal rather than the pathology of the inner ear (common cavity, hypoplasia, or Mondini). Therefore, our findings support the results of Shelton *et al.*[7] Recently Bamiou *et al.*[10] also reviewed the anomalies of the cochlear nerve in congenital patients and all of their cases with absent cochlear nerve were associated with a narrow IAC. There was one patient with a hypoplastic cochlear nerve where the dimensions of IAC were normal. In our series there was only one patient with an enlarged IAC who had an

https://doi.org/10.1258/00222150260293619 Published online by Cambridge University Press

absent cochlear nerve branch. This was a patient with severe Mondini malformation. In three patients with malformations of the inner ear (patients 1, 3, and 4) only two nerves were present in IAC. These most probably represented the facial and cochleovestibular nerve, where the latter did not branch into cochlear, superior and inferior vestibular nerves. The branching anomaly of the cochleovestibular nerve appears to be related to the end organ pathology rather than the IAC anomaly.

In our department we use CT as the primary investigation method. We share the same view as Gray *et al.*[5] that in the presence of total hearing loss, narrow IAC and cochleovestibular malformation we use TSE-MRI for the demonstration of the cochlear nerve. Also in acquired pathologies, which may have cause within the cochlea (meningitis, labyrinthitis, transverse temporal fracture through the cochlea, and autoimmune disorders), we think that TSE-MRI should be used.

Conclusion

Although this study failed to show aplasia of the cochlear nerve on MRI in the presence of normal CT findings in patients who had congenital hearing loss, we think that the findings of Casselman *et al.*[4] and Morris *et al.*[7] should always be kept in mind. If, however, there is a bony malformation on CT (particularly a narrow internal auditory canal), MRI should be done in order to demonstrate the presence of any neural pathology in the IAC. In addition if the patient has total hearing loss in spite of normal CT, we think this is also an indication for FSE-MRI as there is a possibility of an absent cochlear nerve. If, however, any abnormality is detected on CT or in the case of an acquired cause capable of producing fibrosis inside the cochlea, MRI is definitely indicated.

References

1 Lo WW. Imaging of cochlear and auditory brain stem implantation. *Am J Neuroradiol* 1998;**19**:1147–54
2 Maxwell AP, Mason SM, O'Donoghue GM. Cochlear nerve aplasia: Its importance in cochlear implantation. *Am J Otol* 1999;**20**:335–7
3 Arriaga MA, Carrier D. MRI and clinical decision in cochlear implantation. *Am J Otol* 1996;**17**:547–53
4 Casselman JW, Offeciers FE, Govaerts PJ, Kuhweide R, Geldof H, Somers T, *et al.* Aplasia and hypoplasia of the vestibulocochlear nerve: diagnosis with MR imaging. *Radiology* 1997;**202**:773–81
5 Gray RF, Baguley DM, Vanat Z, Begg J, Phelps PD. Cochlear implant failure due to unexpected absence of the eighth nerve – a cautionary tale. *J Laryngol Otol* 1998;**112**:646–9
6 Jackler RK, Luxford WM, House WF. Congenital malformations of the inner ear: a classification based on embryogenesis. *Laryngoscope* 1987;**97**:2–14
7 Shelton C, Luxford WM, Tonokawa LL, Lo WW, House WF. The narrow internal auditory canal in children: a contraindication to cochlear implants. *Otolaryngol Head Neck Surg* 1989;**100**:227–31
8 Morris DP, Walsh R, Ramsden RT, Gillespie JE. Nerve agenesis and hypoplasia in the internal auditory meatus: the importance of early radiological diagnosis in cochlear implantation. Presented at the 5th European Symposium on Pediatric cochlear implantation. June 2000, Antwerp, Abstract Book #054
9 Ellul S, Shelton C, Davidson HC, Harnsberger HR. Preoperative cochlear implant imaging: Is magnetic resonance imaging enough? *Am J Otol* 2000;**21**:528–33
10 Bamiou DE, Worth S, Phelps P, Sirimanna T, Rajput K. Eighth nerve aplasia and hypoplasia in cochlear implant candidates: The clinical perspective. *Otol Neurotol* 2001;**22**:492–6
11 Frau GN, Luxford WM, Lo WW, Berliner KI, Telishi FF. High resolution computed tomography in evaluation of cochlear patency in implant candidates: a comparison with surgical findings. *J Laryngol Otol* 1994;**108**:743–8
12 Thai Van H, Fraysse B, Berry I, Berges C, Deguine O, Honegger A, *et al.* Functional magnetic resonance imaging may avoid misdiagnosis of cochleovascular nerve aplasia in congenital deafness. *Am J Otol* 2000;**21**:663–70

Address for correspondence:
Levent Sennaroglu, M.D.,
28 Cadde, Hardem Apt., 3/A Blok No: 8,
Cigdem Mahallesi,
Karakusunlar, Balgat,
Ankara,
Turkey.

Fax: 90-312-3113500
E-mail: Isennar@tr.net

L Sennaroglu, M.D. takes responsibility for the integrity of the content of the paper.
Competing interests: None declared