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The bony cochlear nerve canal in children with absent or hypoplastic cochlear nerves

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ABSTRACT

Objective: To correlate presence and size of the bony cochlear nerve canal [BCNC] with size of the internal auditory meatus [IAM] on CT in children with absent or hypoplastic cochlear nerves [CNs] as compared to age matched controls.

Methods: This retrospective case-notes review was based in the departments of Cochlear Implantation and Neuroradiology at a tertiary paediatric hospital. Twenty-five ears of fifteen children (subjects) with profound sensorineural deafness (SND) and absent or hypoplastic CN on MRI scan were compared to age matched controls. Two groups of controls were included; a control group of nineteen ears of twelve children with normal hearing or conductive hearing loss [control group 1] and a second control group of twenty one ears of eleven children with severe to profound hearing loss related to GJB2 mutations [control group 2]. Both control groups had evidence of the presence of the CN. Two neuroradiologists independently assessed presence and size of BCNC and IAM on CT and presence of CN on MRI in subjects and controls. The BCNC and IAM size was compared between subjects and both control groups. The presence of BCNC was correlated with the IAM size on CT, presence/absence of CN on MRI and audiological evaluation in subjects.

Results: The mean IAM width was significantly smaller in subjects as compared to controls. The BCNC was absent in 17/25 subject ears and present in all control ears. Absent BCNC correlated with a narrow IAM in 13/17 subject ears. Presence of the BCNC supported presence of a CN although this was not seen on MRI. However, BCNC absence may be associated with presence of a CN as was seen in two subject ears. Five subject ears out of 22 [22%] with absent CN on MRI had other evidence of a present cochlear nerve.

Conclusions: BCNC is an additional parameter to assess presence of the cochlear branch of the CN. Presence of the BCNC may indicate cochlear nerve presence. Caution should be used in assessing candidacy of cochlear implants based on MRI alone and a combination of imaging and audiological tests should be used to assess presence of the CN.

1. Introduction

Imaging of the temporal bone is routinely performed as a part of the initial assessment of cochlear implant candidates to determine the presence of the cochlear nerve [CN] as successful implantation depends on the integrity of the cochlear nerve fibres. High resolution CT and/or MRI are used to visualize the size of the internal auditory meatus [IAM] and the anatomy of the CN within the IAM. A narrow IAM is often an indication of a hypoplastic or absent CN [1,2]. However, aplasia of the cochlear nerve has been reported in the presence of a normal sized IAM [3–6] and cochlear nerve fibres may be present despite narrow IAMs [7–9]. Furthermore, the vestibular and cochlear branches of the CN may not be clearly visualized on MRI in the presence of a narrow IAM [6,10]. Hence it may not be always possible to conclusively infer the presence of a CN, based on either assessment of the size of the IAM on CT or visualizing the nerves on MRI despite targeted high resolution multiplanar imaging of the petrous bones and IAMs. The choice of imaging modality in cochlear implant candidates differs between performing either CT scan, MRI scan or a combination of the two, based on clinical features [11].

The bony cochlear nerve canal [BCNC] is the canal between the fundus of the internal auditory canal and the base of the cochlea [Fig. 1] and carries the cochlear nerve fibres from the spiral ganglion to reach the CN [12]. Normative data on the measurements of the BCNC were reported in earlier studies [13,14], and an

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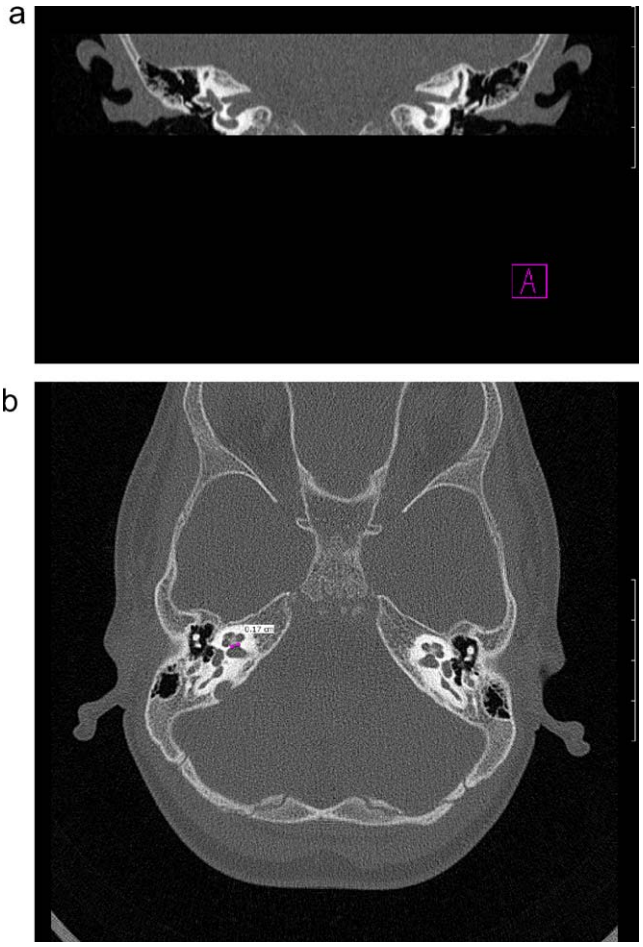

Fig. 1. (a) Coronal and (b) Axial CT showing BCNC and its measurement.

earlier study suggested that the BCNC is small in subjects with profound bilateral sensorineural deafness (SND) [15]. There are few studies on the presence or absence of BCNC in children with absent CNs [6,9,16]. This study aims to compare the presence and size of the BCNC, in children with absent or hypoplastic CNs and age matched controls. The study also aims to correlate the presence and size of the BCNC with the dimensions of the IAMs on CT and presence of CN on MRI.

2. Subjects and methods

This retrospective study was based jointly at the Department of Cochlear Implantation and Department of Radiology in a tertiary paediatric hospital receiving referrals from throughout the UK. Children referred for cochlear implants at this centre undergo a detailed assessment including a medical, audiology, speech & language and psychology review and both CT and MRI scans to visualize the inner ear anatomy before deciding on suitability for implantation. Children with a bilateral pre-lingual severe to profound SND, referred for cochlear implantation were considered for this study. All these children had CT and MRI according to the protocol for cochlear implant assessment at this centre. Fifteen such children who were reported to have absent or hypoplastic cochlear nerves on MRI scans done at this centre were included in the study [referred to as 'subjects']. Two sets of controls were age matched to these subjects. Twelve age matched controls were selected from children with audiologically proven normal hearing or conductive hearing loss referred for high resolution CT of the petrous bones [referred to as 'control group 1'] for reasons other than bilateral SND. This group did not have an MRI scan, but presence of the CN was inferred from results of hearing tests indicating normal hearing or conductive hearing loss. The second set of controls ['control group 2'] comprised of eleven children with bilateral severe to profound sensorineural hearing loss due to GJB2 mutations. This group had evidence of presence of the CN on MRI. Medical case-notes of subjects and the two control groups were reviewed for clinical details and hearing test results.

CT of the petrous bones was performed by direct scanning in both axial and coronal planes to produce a slice thickness of 1 mm. Axial images were acquired as a spiral with a pitch of 1 and coronal images were acquired sequentially. MRI imaging performed on a 1.5 T scanner included Three-Dimensional Constructive Interference in Steady State [3D FT-CISS] MRI reformatted in axial, coronal and parasagittal planes, the latter oriented in the transverse plane of the IAM.

The CT and MRI of subjects and the CT of both control groups were reviewed by two independent neuroradiologists blinded to the clinical information. For all the subjects, the morphology of the membranous labyrinth and the presence of the CNs were assessed on MRI. The CN was considered absent when it could not be visualized on the axial and parasagittal perpendicular sections on the MRI through the IAM. When the BCNC was not visualized on axial sections, coronal views were used to confirm its absence. The CN was considered hypoplastic by subjective visual assessment and by comparison with the size of the adjacent neural structures of the facial and vestibular nerves within the IAM. The size of the BCNC was recorded and the maximum width was measured on axial CT in all subjects and controls. The width of the IAM, configuration of the modiolus and the presence of any cochleovestibular anomalies were recorded. The BCNC and IAM width was measured to the nearest 0.1 mm. A small IAM was defined as a width of less than 3 mm [17–20]. Statistical analysis was done using SPSS [Statistical Package for Social Sciences]. All measurements between the two observers were compared with correlation coefficients [Pearson]. The mean of the two observers' measurements, of the IAM and BCNC of subjects and controls, were compared using paired nonparametric tests [Wilcoxon paired test]. The project was approved by the Research and Development directorate at the authors' institution.

3. Results

3.1. Clinical features [Table 1]

Twenty-five ears from fifteen subjects [age 9 months to 11 years] with prelingual bilateral severe-profound SND and absent or hypoplastic CN on MRI were included in the study. Ten subjects had bilaterally absent or hypoplastic nerves. Five subjects showed unilateral absent or hypoplastic nerves, and the ears showing presence of a normal CN on MRI were excluded from the study. Clinical features and syndromic diagnosis are detailed in Table 1. The median age of diagnosis of SND in the subjects was 3.5 months [range: birth to 22 months, mean age: 6 months]. Of the two subjects with associated systemic malformations one had anal atresia, congenital heart disease and ear tags and another had choanal atresia, but a unifying diagnosis could not be identified. Thirteen subjects had delayed motor milestones and 11 had dysmorphic facial features. Two subjects [one each with Moebius and CHARGE syndrome] had associated facial palsy and the others had normal function of the facial nerve clinically. The 25 ears of the subjects were age matched to 19 ears of 12 children in control group 1 [age 11 months to 12 years] and to 21 ears of 11 children in control group 2 [age 8 months to 12 years]. Control group 1 ears affected by microtia or SND were excluded from the analysis.