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Cochlear implants for congenital deformities

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Abstract
There have been few accounts of multi-channel cochlear implants in patients with congenital structural deformities of the inner ear which are associated with severe and sometimes progressive deafness. These malformations can now be recognized easily on 2 plane thin section high resolution CT studies which are mandatory for the pre-implantation assessment. However, no attempt seems to have been made to describe which of these malformations would be suitable for an implant or for which would this procedure be contra-indicated. True Mondini deformity of both the cochlea and dilated vestibular aqueduct type would appear suitable for a multi channel implant, but this type of implant should not be used for a primitive otocyst, severe labyrinthine dysplasia or the characteristic X-linked deformity.

Introduction

The insertion of a multichannel electrode into the coils of the cochlea, the so-called cochlear implant, has become a practical and successful operation for improving hearing in some very deaf patients. Despite their high cost cochlear implant programmes in Great Britain have long waiting lists and there has been some financial support from the government. Careful selection of suitable patients is therefore essential and rigorous assessment programmes necessary. It soon became apparent that imaging of the petrous temporal bones was most important for the screening of candidates for cochlear implants. The best results from cochlear implants are achieved in patients who have become severely deaf after speech has been acquired; the so-called post lingual deafness. Many such patients have been the victims of suppurative labyrinthitis of either tympanogenic or meningitic aetiology or severe otospongiosis/otosclerosis. A previously undiagnosed cause of deafness is often revealed only by the imaging done as part of the pre-implant assessment programme (Phelps *et al.* 1990). The obliteration or partial obliteration of the cochlear coils by labyrinthitis ossificans is shown reliably by thin section high resolution computed tomography in the axial plane and this has been described in several publications (Harnsberger *et al.*, 1987; Jackler *et al.*, 1987; Gantz *et al.*, 1988; Müeller *et al.*, 1989).

There have as yet been few attempts to use cochlear implantation for severe congenital deafness. Successful implantation in a patient with bilateral Mondini deformities was described by Silverstein *et al.* (1988) and Shelton *et al.* (1989) warn of the importance of recognizing a very narrow internal auditory meatus (IAM). They reported that three patients with this anomaly failed to respond to the sensation of sound after receiving an implant.

Material and methods

The following is an account of imaging studies of congenital structural abnormalities in severely deaf children who may be assessed for possible cochlear implantation. High resolution computed tomography with contiguous 1 mm section in the axial and coronal planes forms the basis of the examination. Magnetic resonance has a less clearly defined role but may be useful for confirming the presence of the audiovestibular nerve and confirming that the cochlea contains fluid.

1. *Narrow IAM*

An IAM with a calibre less than 2 mm diameter is outside the normal range and is therefore unlikely to contain an audiovestibular nerve. Moreover, if a narrow IAM continues laterally into the labyrinthine portion of the facial nerve canal with no change of calibre obviously no other neural components can be present (Phelps and Lloyd, 1990). It is most important, however, to confirm the narrow calibre by imaging in two planes (Fig. 1).

2. *Primitive Otocyst*

The Michel deformity, complete absence of any vestige of inner ear, is in fact exceedingly rare. A commoner lesion is the primitive otocyst with endolymphatic appendage as shown in Fig. 2. The IAM is usually absent but a tract for the facial nerve runs anterior to the otocyst. These cases with total anacusis are however unsuitable for implantation.

3. *Severe labyrinthine dysplasia*

A dilated vestibule communicating widely with an amorphous sac representing the cochlea is also associated with total anacusis. Moreover this lesion is associated with a very real risk of the development of a spontaneous cerebrospinal fluid fistula through a defect in the footplate of the stapes or of recurrent attacks of meningitis. The wide direct communication between the subarachnoid

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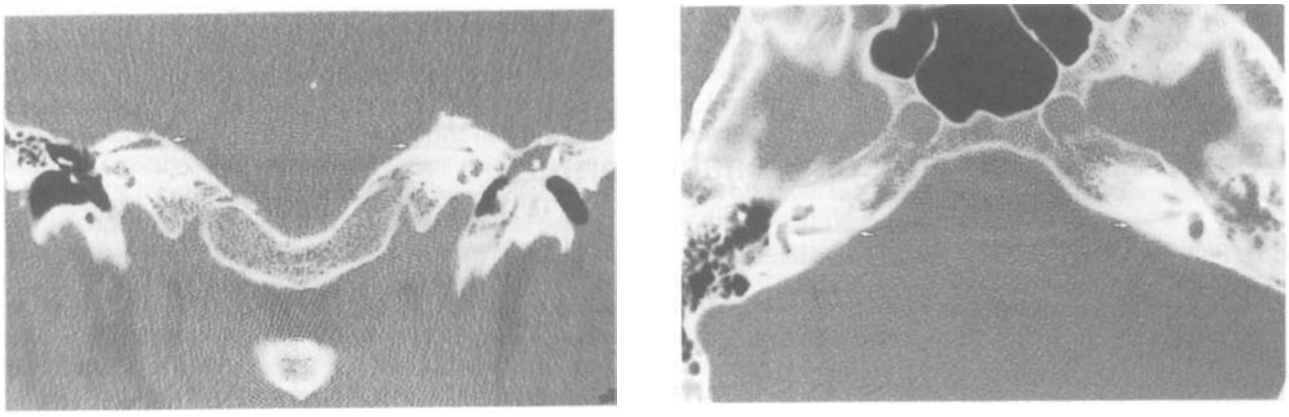

FIG. 1

Coronal (a) and axial (b) CT sections through the petrous temporal bones showing a very narrow internal auditory meatus on both sides (arrows).

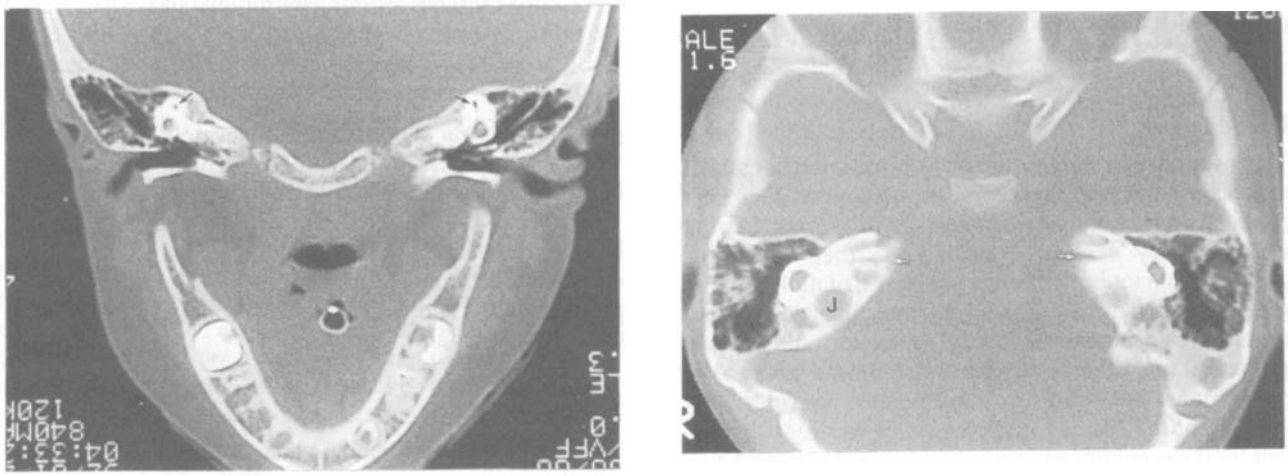

FIG. 2

Coronal (a) and axial (b) sections showing primitive otocysts with endolymphatic appendage in both sides (black arrow). The facial nerve canal (white arrow) is anterior to the otocysts. There is a high but normal jugular bulb (J) on one side.

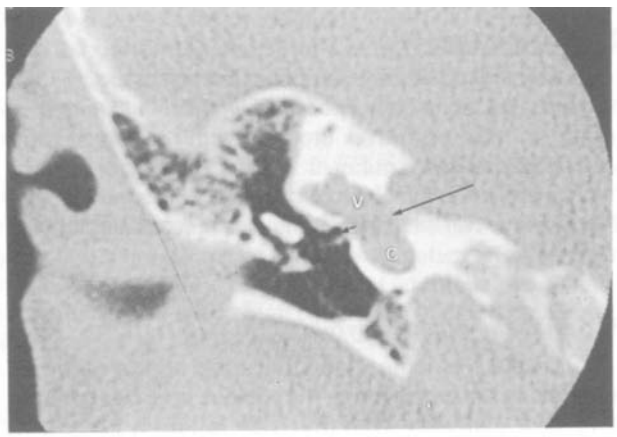

FIG. 3

Coronal CT section showing severe dysplasia of the labyrinth. The dilated vestibule (V) forms a single cavity with the amorphous cochlea (C). The patient had recurrent meningitis and when the stapes with a dehiscence in the footplate was disturbed there was a gush of CSF. The route from the tapering IAM is depicted by the arrows.

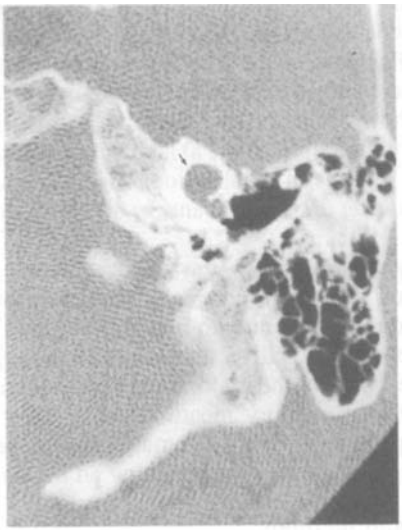

FIG. 4

Axial CT section showing a true Mondini dysplasia which would probably be suitable for an intracochlear implant. Note the normal calibre basal turn and the distal sac (arrow).

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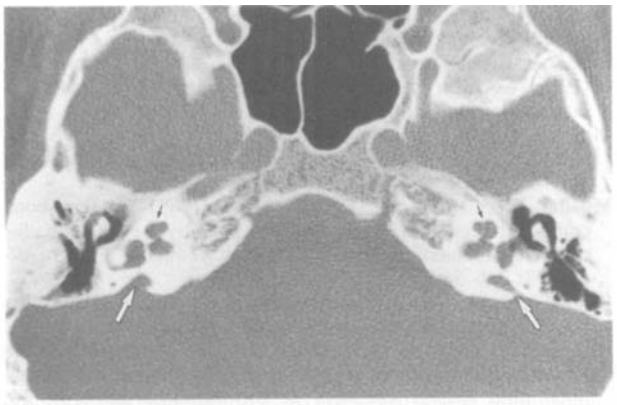

FIG. 5

True Mondini deformity of cochlear coils and vestibular aqueduct. This axial CT section shows normal vestibule and lateral semi circular canals but deficient interscalar septum in the cochlear coils resulting in a distal sac (black arrow). There are also dilated vestibular aqueducts bilaterally (white arrow).

space in the IAM and the middle ear (Fig. 3) make this type of deformity unsuitable for a cochlear implant. Surprisingly a recent case of ours which died as the result of meningitis was found to have an audiovestibular nerve present despite a negligible endorgan in the single tube cochlea.

4. *Deficient distal coils (Mondini deformity)*

The true Mondini deformity consists of a normal basal turn and distal sac (Fig. 4). Consequently some hearing is possible and we have seen several such cases. Moreover the deafness may be progressive, as in the case recorded by Silverstein *et al.* (1988), and therefore these patients become suitable for an implant if sufficiently deaf.

5. *Dilated vestibular aqueduct*

A wide vestibular aqueduct was a feature of the temporal bones dissected by Mondini in 1791 and therefore the so called 'vestibular aqueduct syndrome' is part of the Mondini dysplasia. Moreover we have seen several such cases identified by imaging (Fig. 5). Progressive and fluctuant hearing loss often associated with trivial head trauma is a characteristic feature of 'dilated vestibular aqueduct syndrome'.

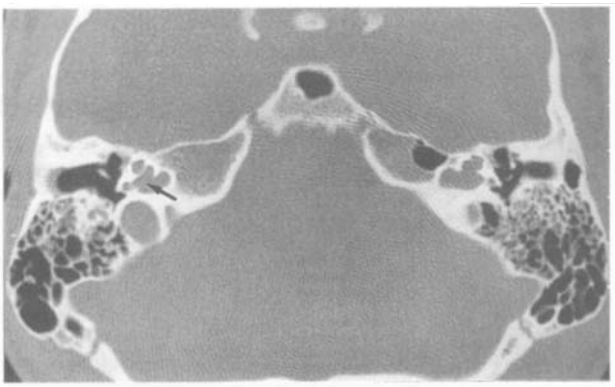

FIG. 6

The characteristic feature of the inner deformity in one variety of X-linked deafness is not so much the bulbous IAM but the deficient bone between the fundus of the IAM and the basal turn of the cochlea (arrow) shown on this axial CT section.

6. *X-linked deformity*

An association of X-linked mixed deafness with a stapes gusher (a profuse flow of perilymph or cerebrospinal fluid if the stapes is disturbed) has been recognized for twenty years. However, it is only recently that a distinct type of deformity has been recognized by imaging in some severely deaf males (Phelps *et al.*, 1991). It would seem that the most important aspect of the deformity is deficient bone between the lateral end of the bulbous IAM and basal turn of the cochlea (Fig. 6). This would preclude the insertion of a multi channel electrode as the deficient bone would almost certainly mean that the electrode would enter the IAM rather than staying in the cochlear coils. However, genetic studies have shown that there are at least two types of X-linked deafness (Reardon *et al.*, 1991) some of which have normal inner ears as shown by CT and are therefore suitable candidates for implantation.

Discussion

Fifteen years ago and again more recently we described inner ear deformity associated with a risk of a cerebrospinal fluid fistula (Phelps and Lloyd, 1978; Phelps, 1986). We pointed out that this was not the abnormality described originally by Mondini but a more severe dysplasia probably representing arrest of development at an earlier stage. Such cases would not seem suitable for an intracochlear implant because of the communication or potential communication with the subarachnoid space in the IAM which could lead to problems with a CSF leak. An extracochlear implant would also be unsuitable because of the absence or extreme deficiency of any cochlear endorgan. Jackler *et al.* (1987) also consider that describing several types of inner ear abnormality under the single umbrella term of 'Mondini Dysplasia' is unwarranted. The term should be kept only for the most prevalent form of minor cochlear deformity with incomplete partition: a normal basal turn and a distal sac. These patients often have some hearing present and in my opinion are not at risk of developing a spontaneous CSF leak into the middle ear. Moreover if the deafness in a true Mondini deformity becomes severe enough then these patients are suitable for an intracochlear implant. Only slight improvement in hearing was recorded in the case described by Silverstein *et al.* (1988) but it is not clear why these authors should have performed a pre-operative lum-

FIG. 7

Thin section (1.5 mm) 3DFT of petrous temporal bone in the axial plane showing the cochlear coils (large arrow) and the audiovestibular nerve (small arrows) in a normal subject.

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bar puncture. They apparently thought there was a risk of CSF fistula through a dilated cochlear aqueduct but the relevant CT scan seems to show dilated vestibular aqueducts, not cochlear aqueducts.

The dilated vestibular aqueduct with or without a true Mondini cochlear deformity is associated with progressive deafness and these patients become suitable for an implant although to my knowledge no implants in such cases have been described.

Progression of severe deafness is also a feature of the X-linked inner ear deformity, but our studies appear to have confirmed that there are at least two types with entirely different radiological features (Phelps *et al.*, 1991). The patients on whom the axial CT scan show normal appearances of the petrous temporal bones would be suitable for an implant. Those with the characteristic deformity of deficient bone between the lateral end of the internal auditory meatus and the basal turn of the cochlea would be unsuitable for a multi-channel implant for the same reason as the patients with the severe cochlear dysplasia: the electrode would be likely to penetrate into the IAM. A single channel extracochlear electrode would however be feasible. It is these patients with the characteristic X-linked deformity who develop a stapes gusher if the stapes is disturbed.

Careful assessment by CT imaging is mandatory for any patient who is being considered for an intracochlear implant. Rapid developments in MRI however mean that this type of imaging may assist or even supplant CT for showing the audio-vestibular nerves and also the soft tissue densities in the cochlear coils (Fig. 7).

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