The Journal of Laryngology and Otology
July 1998, Vol. **112,** pp. 646–649

Clinical Records

Cochlear implant failure due to unexpected absence of the eighth nerve – a cautionary tale

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Abstract
We present a case of bilateral absence of the eighth cranial nerve in the internal auditory meatus (IAM). This caused total failure of responses after cochlear implantation in a six-year-old patient with congenital deafness. Pre-operative magnetic resonance (MR) imaging is important to show not only the anatomy of the middle and inner ears but also the structures in the IAM.

Key words: Cochlear implant; Hearing loss, sensorineural, congenital; Ear, anatomy; Magnetic resonance imaging

Introduction

Cochlear implantation has now become an accepted method of managing profound sensorineural deafness both congenital and acquired (Summerfield and Marshall, 1995) in cases where there is no medical, surgical or developmental contra-indication. The surgical technique is safe and reasonably straightforward with few complications in the hands of an experienced otologist (Cohen and Hoffman, 1991). The East of England Cochlear Implant Programme at Addenbrooke's Hospital has been in existence for 11 years, with 132 patients implanted with intra-cochlear devices: of these 42 are children.

The selection process for paediatric cochlear implantation has an implicit assumption that a profound congenital sensorineural hearing loss is cochlear in origin. A case is reported here where this assumption was invalid. Profound hearing loss was the result of bilateral congenital abnormality of the cochleo-vestibular nerve. This anatomical abnormality was not initially detected and the hearing loss was managed with a multichannel cochlear implant. Stimulation of the implant did not lead to any auditory perception.

The case is presented as a cautionary tale with a suggested strategy for identifying such cases in future so that this situation may not be repeated.

Case report

Child S (female) was born at term as breech presentation with outlet forceps delivery following an uneventful pregnancy in 1991 and did not need neonatal intensive care. She was a first child with normally hearing parents and there was no family history of hearing impairment or any congenital anomalies. Neonatal screening for hearing impairment was not performed. The child was suspected as having hearing loss by her parents around the age of ten months. This was confirmed at the age of 15 months by auditory brainstem responses performed under general anaesthetic with an indication of the loss being profound. Grommets were inserted at this time for associated glue ears. Hearing aids were fitted and were worn regularly thereafter. There was no appreciable improvement in her hearing or speech and language development as had been expected. Sign supported English was started to supplement her communication skills.

She was suspected to be suffering from a visual problem due to the absence of the blink reflex but this was ruled out by ophthalmic investigations. Developmental milestones were noted to be slightly delayed with regard to her motor skills and she learned to walk at the age of 22 months. Suspicion of a microcephaly prompted careful search for other somatic anomalies (Feingold, 1978). After careful consideration the full picture was difficult to fit into the named syndromes. Chromosomal studies were normal. Facial nerve function was normal.

Referral for implantation

This child with congenital profound deafness was referred by her local hospital to our centre for consideration of cochlear implantation during the latter part of 1995. Assessment involved a detailed history and clinical examination followed by speech and language assessment and trial of hearing aids with new moulds. Thereafter the aided thresholds with visual reinforced audiometry were at 70 dB (at 250 Hz) and 90 dB (at 500 Hz) with no responses at the higher frequencies. Functional listening was assessed: the child would respond to a loud drum but not to any loud voiced sounds unless she could see the lip pattern. Lip-reading skills were limited although she was able to attempt to copy lip patterns. She did not use her voice other than to attract attention. High definition computed tomography (CT) scans of the temporal bones revealed patent cochlear ducts on both sides. Cochlear

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Accepted for publication: 19 April 1998.

morphology was reported as normal. This child met the selection criteria for paediatric cochlear implantation.

Operative procedure

As the left middle ear cleft was still open with a grommet and had active discharge around that, it was decided to implant the other ear which had a healthy tympanic membrane. A Nucleus 22 intra-cochlear device was implanted on the right side in January 1996 using a vertical postaural (Gibson) incision, cortical mastoidectomy, posterior tympanotomy and cochleostomy. Dacron ties and bone cement were used to anchor the electrode array. The surgical procedure was uneventful. A good insertion was obtained with 22 functional plus five supporting electrode rings inside the cochlea. Electrical integrity testing of the device was performed and deemed satisfactory. At this time electrical auditory brainstem response (EABR) was being introduced in this centre: the results were equivocal, this being ascribed to the lack of experience in this technique. Electrical stapedial reflex testing was not undertaken due to lack of time. Post-operative recovery and wound healing were satisfactory.

Tuning

The first tuning session took place in March 1996. The child's reaction to electrical stimulation was clear but seemed to be in response to a sensation produced at the right eyelid. This type of 'non-auditory stimulation' occurs when other nerves in the vicinity of the cochlea (in this case the facial nerve) are affected by spread of electrical energy. It was difficult to collect repeatable, reliable responses from child S but the team were eventually able to gather information on six electrodes. Stimulation levels were set very low to avoid triggering the eye twitch but it reappeared when the map was activated. Adjustments were made to the map but after a few days the twitch was apparent again. This became the pattern over the next few sessions: electrodes were set conservatively, the eye twitch was not apparent, then after a day or so the eye twitch would return. Suspect electrodes were inactivated from the map and the parameters of the map adjusted. The most reliable data from sound field testing was as seen in Table I.

It was not possible to determine whether these implant-aided responses were auditory or non-auditory. In the first few months of implant use the family reported that the child was responding to environmental sounds such as her baby sister crying, a wrapper being scrunched, running water and pages turning in a book. But, again, we could not be sure whether auditory or non-auditory stimulation was responsible for her awareness.

Progress

The child communicated through sign language and was beginning to attempt some sounds and lip patterns. Her language development in sign gave no cause for concern but her development of spoken language was progressing very slowly. She could say 'bye-bye', 'please', 'dada' but could no longer say 'mummy'. She knew all her colours in sign and played imaginative games. It was difficult to maintain her concentration as she liked to take control of an activity or offer detailed observations which made it difficult to keep her on task.

TABLE I
COCHLEAR IMPLANT-AIDED AUDIOGRAM (MAP 3) (SENSITIVITY CONTROL ON 2.0) [DATE 18.4.96]

Frequency (Hz)	250	500	1K	2K	4K
DBA	65	65	NR	NR	NR

TABLE II
MANCHESTER PICTURE TEST

Mode	% Correct (chance 25%)
Sign plus speech	100
Lip reading only (3 lists)	30, 30, 20
Lip reading plus voice (4 lists)	60, 60, 50, 40

Further assessment examined lip-reading alone and lip-reading with speech using the Manchester Picture test. All eight lists of the test were used to investigate the relationship between visual input from the processor (Table II).

These results suggested that the speech processor was delivering a small but significant amount of information which could be used to decode single words in a closed set activity. However, it is more likely that this was not sound but non-auditory stimulation to the facial nerve. During this session eye twitches were noted when the child was presented with warble tones at 2K and 6K at about 70 dB.

Integrity tests

Electrical integrity testing of the internal implanted parts of the device was carried out by a representative from the manufacturers on July 1996. This confirmed that all elements of the implanted equipment were working appropriately.

Second opinion and explanation

The child was referred to another cochlear implant centre (Nottingham) for an independent second assessment Integrity testing again showed an intact and normally working implant. However electrical auditory brainstem response testing showed no evidence of stimulation of any part of the auditory pathway. Kinking of the electrode array was suggested. Therefore, an ultra high resolution CT scan was performed: general anaesthesia was not required. The receiver and the electrode array were seen to be in satisfactory position. New information obtained with this scan was the narrow appearance of the internal auditory meatus (Figure 4) especially on the implanted side with absence of Bill's Bar. However in retrospect this was also apparent in the original CT scans but had not been reported. This seemed a clue to abnormality of

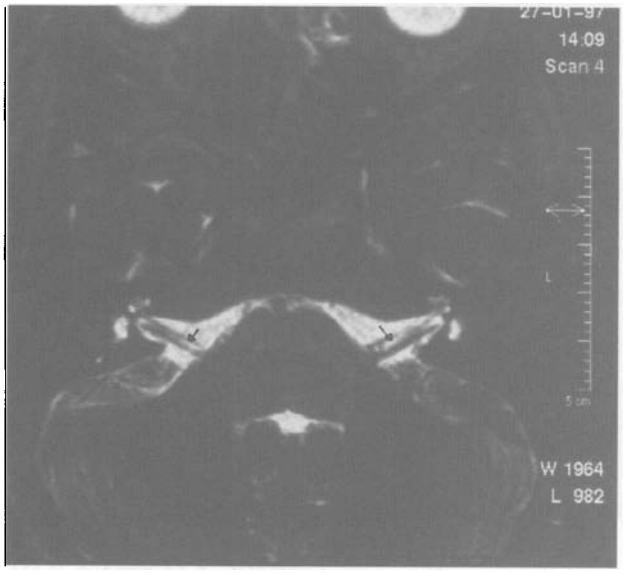

FIG. 1
Axial 0.7 mm T_2 weighted MR image of IAM showing normal structures (arrow) (Normal subject)

https://doi.org/10.1017/S0022215100141349 Published online by Cambridge University Press

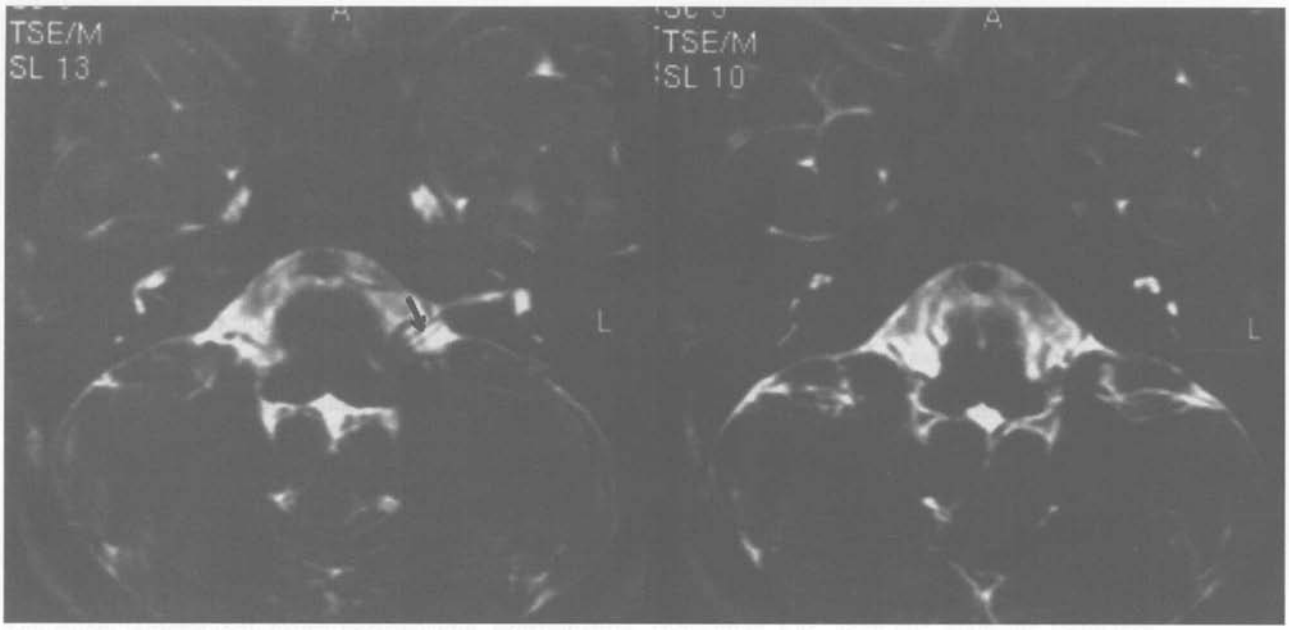

FIG. 2

Axial 0.7 mm T_2 weighted MR image of IAM showing absence of nerve bundle on the right side and a hypoplastic nerve on the left side (arrow). (Patient).

structures in the IAM (Phelps, 1992). Detailed views of the IAM and the cerebello-pontine angle (CPA) were required and this was performed by magnetic resonance imaging (MRI). The Nucleus 22 device lacked a removable magnet. After much discussion with the family the device was explanted under general anaesthesia, 15 months after implantation. The surgery was uneventful and nothing was found to explain the failure of responses.

MRI scan

A detailed three-plane MRI scan of the petrous temporal bones was requested under a general anaesthetic.

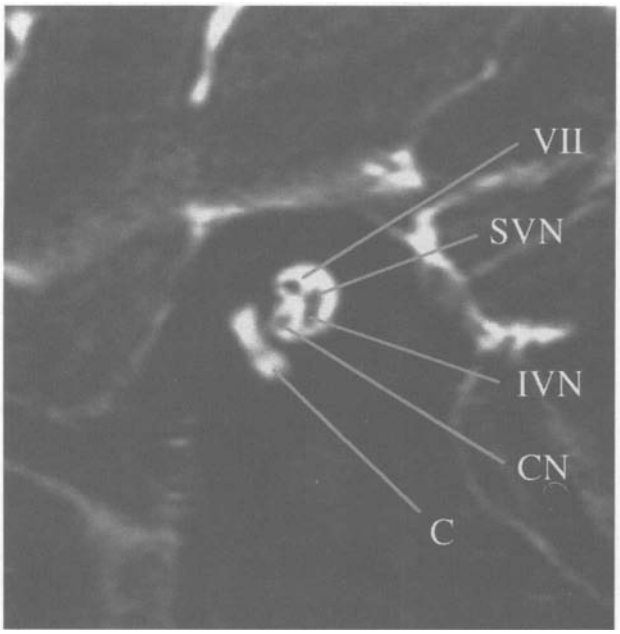

FIG. 3

Saggital 0.7 mm T_2 weighted MR image of IAM showing normal structures. VII = Facial nerve; CN = Cochlear nerve; C = Cochlea; SVN = Superior vestibular nerve; IVN = Inferior vestibular nerve. (Normal subject).

On both sides a single prominent nerve (presumably the facial nerve) was seen to cross the CP Angle and enter the IAM. There was, however, another much thinner single structure alongside this and this was reported to be a hypoplastic cochleo-vestibular nerve (Figure 2).

In view of these new findings it was decided not to re-implant this child on either the previously implanted nor the contralateral side. Given the child's perseverance and enthusiasm and also the family support available she was fitted with a vibrotactile aid to help her speech perception.

Discussion

The assumption that a congenital hearing loss is caused by a cochlear lesion has been demonstrated to be invalid. The aetiology of the SNHL in this case was a vestigial

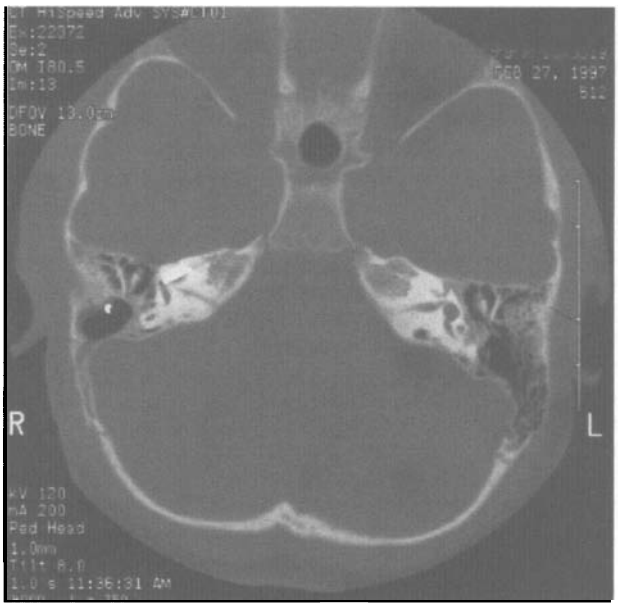

FIG. 4

Ultra high definition CT scan (axial view) showing narrowing of IAMs on both sides. (Patient).

https://doi.org/10.1017/S0022215100141349 Published online by Cambridge University Press

cochleo-vestibular nerve in the IAM only identified by MRI. If an implant candidate demonstrates auditory function (aided or unaided) (not vibrotactile responses) then there should be an eighth nerve. In the absence of such responses care must be taken to demonstrate a cochleo-vestibular nerve in the IAM before implantation.

CT or MRI for implant candidates?

The radiological investigation of choice in a cochlear implant assessment has been CT scanning (Bath *et al.*, 1993). This technique images bone accurately, particularly the all important cochlear duct and gives an indication of the patency of the basal turn of the cochlea. CT scanning has also been shown to identify a narrow IAM (Shelton *et al.*, 1989) and this morphological anomaly has been associated with an absent eighth nerve. The cochleo-vestibular nerve cannot be seen by CT however, and MRI is better for this purpose. The use of MRI for cochlear implant assessment has been proposed by Arriaga and Carrier (1996).

If a child demonstrates auditory thresholds then assessment of the patency of the cochlear duct by CT is sufficient. Where there are no auditory thresholds some thought must be given to the investigative strategy. A CT scan would provide information as to the width of the IAM, but it is not known what extent of narrowing indicates an absent eighth nerve. Casselman *et al.* (1997) described MRI of the IAM in seven cases with 'congenital or unexplained hearing loss' and abnormalities of the cochleo-vestibular nerve. Aplasia of the cochleo-vestibular nerve was demonstrated in two cases, with associated stenosis of the IAM. In three cases the IAM was of normal morphology yet the cochlear branch of the cochleo-vestibular nerve was absent or hypoplastic. CT would thus miss three out of five eighth nerve hypoplasias. The best investigation in such cases is MRI, in the hands of a radiologist experienced in imaging the contents of the IAM (Figures 1 and 3). Either submillimetric gradient echo images such as 3DFT-CISS described by Casselman should be used or alternatively equally thin high resolution T2-weighted sections by a two-dimension technique can be obtained. The latter is faster with better spatial resolution; the former allows for three-dimensional resolutions.

Child S met the audiological criteria for implantation. It is apparent however that peri- and post-operative audiological investigation could have identified the existence of the anatomical anomaly earlier than was the case had our experience been greater. Pre-operative promontory stimulation testing by subjective response would have been helpful although perhaps impractical in a child. Peri-operative stapedial reflex testing would have been helpful in assessing the functional integrity of the auditory pathway to the level of the superior olivary nucleus. Concerns about the progress of child S with the implant led to investigation in the form of integrity testing, which was normal. This did not identify the cause of the problem but EABR was definitive. It is now our strategy to demonstrate the functional integrity of the auditory pathway in such cases with no auditory thresholds by peri-operative EABR and stapedial reflex measurements. In such cases where progress is slower than expected the functional integrity of the auditory pathway should be demonstrated with EABR: integrity testing is insufficient. However post-operative tests are all too late if there is an absent eighth nerve which should have been identified pre-operatively.

Developmental delays and failure to progress should also prompt the implant team to look for syndromal features (Brunner and Winter, 1991) especially if there are associated somatic stigmata.

The twitches in the upper eyelid with auditory input were probably due to stimulation of the facial nerve or its branches by the spread of electrical energy around the region. This is also hardly surprising, the seventh cranial nerve being the only normal structure passing through the IAM in this case. This non-auditory response was a false indication of progress in tuning, and the cochlear implant scientist should be mindful of this possibility.

Our experience in this case is presented as a cautionary tale. The assumption that a total congenital SNHL can be ascribed to a cochlear lesion is invalid, and the anatomical integrity of the auditory pathway should be demonstrated by MRI.

Conclusions

Although not common, IAM pathologies do occur in congenitally deaf patients. Some are associated with abnormal cochlear morphology and these should be carefully looked for in the initial films. Narrow IAM on CT scan is an indirect pointer to abnormality in the neural structures passing through it and should prompt further imaging of the region.

Responses on tuning cannot be trusted if they could be non-auditory, particularly in children who cannot describe the sensations. We recommend that non-auditory responses are regarded with extreme suspicion and EABR responses are the most valuable evidence of a true hearing response.

We also recommend that in all cases of total congenital hearing loss anatomical integrity of the auditory pathway be demonstrated using MRI prior to cochlear implantation.

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