Cochlear Implants – Surgical, Audiological and Electrophysiological Issues

Jaydip Ray

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Promotores:	Prof.dr. C.W.R.J. Cremers
	Prof. W.P.R. Gibson (Sydney, Australia)
Copromotores:	Mr. D.W. Proops (Birmingham, United Kingdom)
	Mr. R.F. Gray (Cambridge, United Kingdom)
Manuscriptcommissie:	Prof.dr. H.P.H. Kremer (voorzitter)
	Prof.dr. F.E. Offeciers (Medisch Instituut St. Augustus, Wilrijk, België)
	Dr.Ir. A.F.M. Snik

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CHAPTER 1

GENERAL INTRODUCTION

1.1

INTRODUCTION

COCHLEAR IMPLANTS – HISTORICAL PERSPECTIVE

Cochlear implants are now widely accepted as one of the best ways to restore useful hearing in profound hearing loss and data supporting is now overwhelming.

The idea of using electrical energy to produce hearing sensation goes back to Alessandro Volta, who in 1790, placed metal rods in his ears and connected them to an electrical source, losing consciousness in the process. He fortunately woke up to remember bubbling noises inside his ears as a result of the electrical stimulus[1].

Around 1957, Djourno and Eyries in France applied a single copper wire to the auditory nerve of a totally deaf man who had a mastoid cavity from cholesteatomatous disease. An induction coil and an indifferent electrode were placed in the temporalis muscle and an active electrode was placed on a segment of the auditory nerve visible through the vestibule. The coil was later stimulated by induction currents from a second coil placed against the overlying skin. The subject reported sounds like “crickets” [2].

However the real origins of modern day Cochlear Implants lies in the work of William House in Los Angeles who in 1961 implanted a single channel implant in the scala tympani of the cochleas of two patients. Both had a hard wire gold electrode placed in the scala tympani through the round window[3]. Both patients reported sensation of hearing with electrical stimulation.

With this initial success other institutes like the San Francisco group[4] and the Stanford group[5] focussed their attention on cochlear implantation. Simultaneous with these groups another investigator working in Australia, Graham Clark, was making great progress in designing a multichannel cochlear implant. In 1978 and 1979 three profoundly deaf adult patients received multichannel cochlear implants with 20 channel electrode arrays[6,7,8].

In Europe Drs Ingeborg J Hochmair-Desoyer and Erwin S. Hochmair along with Dr. Kurt Burial implanted five patients with multichannel devices in 1970. The number of electrodes varied between six and eight[9].

COCHLEAR IMPLANT BASICS

In the profoundly deaf ear most of the hair cells are absent or non functioning severing the connection between the peripheral and control auditory systems. The cochlear implant bypasses the hair cell by delivering electrical signals directly to the surviving elements of the auditory nerve. This it does by a combination of *transduction, amplification, compression, filtering, feature recognition, feature extraction* and *encoding*

The basic design of the cochlear implant was derived from the cardiac pacemaker industry. The internal components are usually encased in biocompatible titanium silastic or ceramic casing. The electrodes are made of platinum iridium wires encased in biocompatible silastic carrier. The device is comprised of external and internal parts. The external components include a microphone, speech processor and transmitter. The internal component is the receiver / stimulator.

It is interesting to note that over thirty different device designs have been developed over the first fifteen years[10].Cochlear implants differ in their basic appearance and also in the following aspects:

- Type of electronic microphone
- Type of speech processor
- Speech processing strategy
- Transfer of energy across scalp
- Number of electrodes
- Placement of electrodes
- Signal delivery, current flow and charge density

Microphone: This is mostly housed in the “behind the ear” unit. But a separate “clip on” microphone can be placed remotely and attached with a cable. Direction microphones are useful in competing noise.
Speech processor: This converts the microphone input into patterns of electrical stimulations and can either be *ear level* or *body worn*.
Speech processing strategies have evolved over the years in an attempt to match electrical stimulation with acoustic input. The commonly used strategies are:
Compressed Analogue (CA) strategy was amongst the first to be used with multichannel cochlear implants and was widely used with the now discontinued Ineraid implants. The CA stimuli represent a large proportion of the information in unprocessed speech and provided significant open set speech recognition[11]. A

more recent form of analogue stimulation is *Simultaneous Analogue Stimulation (SAS)* which is supported by the Clarion S-series implant processor[12]. The SAS strategy is fully simultaneous and reaches stimulation rated of 90,000 samples per second.
However there seems to be a trade off between attempts to improve temporal resolution by increasing simultaneous channel stimulation and introducing distorsion from electrical field interaction. Therefore other strategies like *Hybrid Analogue / Pulsatile (HAP), Quadruple Pulsatile Sampler (QPS)* and *Pulsed Pulsatile Sampler (PPS)* with lower stimulation rates have also been tried.

Continuous Interleaved Sampling (CIS) on the other hand is a completely sequential strategy and attempts to represent voicing information in a more natural way. The envelope signal in CIS is claimed to convey slow variations in the vocal tract [13].

Multipeak (MPEAK) and later *Spectral peak (SPEAK)* strategies used in the Nucleus devices were developed by Cochlear Ltd and the University of Melbourne. The input is filtered into twenty bands. A postprocessor scans the output envelopes and gives good speech recognition[14].

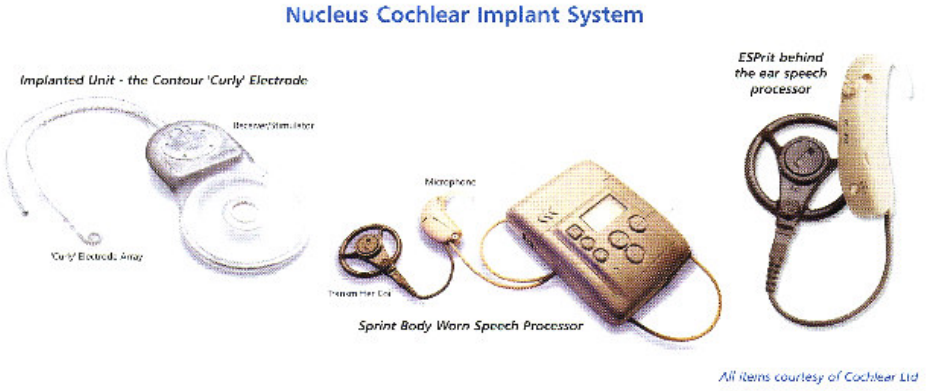
Advanced Combination Encoder (ACE) uses faster rates of stimulation than SPEAK and the additional information has been shown to have improved word recognition scores[15,16].

Transmitter: The transmitter transfers power information from the speech processor to the receiver / stimulator either *percutaneously* through a hard wired connection or *transcutaneously* through the skin (as radiofrequency signal or magnetic inductionn). Percutaneous connection provides easy transmission of stimuli and also enables direct recording of intracochlear evoked potentials[17,18]. Transcutaneous connection on the other hand limits the amount of stimuli that can be transferred at one time. However it has the advantage of reducing the risk of infection because of an intact scalp cover.

Electrodes: The electrodes and the electrode carrier are together called the *electrode array*. This can either be *single channel* or *multi channel*. The electrodes can be ball shaped, concentric rings or plates.

The electrode array is commonly inserted into the scala tympani. Electrode arrays can be different in number, placement, stiffness and cross sectional area. Perimodiolar placement of the electrodes is achieved either with a contoured electrode array (Nucleus devices) or with a *positioner* (Clarion devices)[19,20]. These *modiolus hugging* electrode arrays tend to take up a position close to the spiral lamina. The proximity of the electrodes to the neural elements reduce stimulation levels and thus power requirements[21].

The stimulus itself can be bipolar or monopolar. In bipolar mode each intra-cochlear electrode is stimulated with respect to another intracochlear electrode. Different pairs of electrodes can be used to stimulate different segments of the cochlea. In monopolar mode the intracochlear electrode can be stimulated with respect to another remote electrode.



SAFETY ISSUES

Overall cochlear implantation is a relatively safe procedure. Head growth is not affected by cochlear implants and otitis media with effusion and meningitis are no more or less common in these patients. A history of recurrent otitis media should not inordinately delay cochlear implantation[22]. In a study of fifty children with cochlear implants 74% had otitis media before and 16% after implantation. 28% required ventilating tubes for recurrent otitis media before implantation. All episodes of postimplant otitis media were successfully treated with routine oral antibiotics without any sequelae[23]. The commonest complications are wound infection and flap breakdown. This has been addressed in the section on surgical issues[24,25,26,27].

Recently however some concerns were raised both in the US[28] and in Europe[29] about the link between cochlear implantation and the risk of meningitis. The Food and Drug Administration noted that 91 cases of meningitis were reported worldwide in patients who had received cochlear implants and a total of 17 deaths had resulted from this infection. Majority of the patients were younger than 7 years of age although some adults were also affected. The pathogenesis of the meningitis was uncertain but implant design, surgical technique and associated congenital inner ear malformations were presumed to be precipitating factors. One particular implant design using a special positioner was thought to be at a higher risk of causing meningitis and was withdrawn from the market. Some calculated the risk of meningitis in implanted patients to be 30 times higher than an age matched US population[30]. European[31] and Australian[32] implant safety data was also scrutinised and recommendations for prevention[33] published. Prompt antibiotic treatment of acute otitis media is needed in implanted patients. Vaccination against the most prevalent types of bacterial meningitis (Streptococcus pneumoniae and Haemophilus influenzae) should be considered in patients of all ages who have implants or are candidates for implantation.

PSYCHOSOCIAL ISSUES

Cochlear implants have the potential to impact on every aspect of the recipients' life. Other types of surgical procedures like sight restoration or cosmetic surgery which alter self perception may be associated with psychological distress as it can change social relationship and alter their worlds. In the early stages of implantation

there was tremendous scepticism about cochlear implantation in children[34] but many of the worries have been dispelled by the better than expected results obtained in younger children. Adolescents find the device most difficult. Similar to the deaf adults whose social contacts are tied closely to deafness, they may view cochlear implants as something that may stigmatise them amongst their peers. There is a high risk of rejection of a normally working implant[35,36] and this issue is addressed in the chapter on non-users.

In working adults there is evidence of under-employment in deafened people. Implantation brings about a marked improvement in working life with greater confidence and job security[37].

COCHLEAR IMPLANT CANDIDACY

Decisions on cochlear implant candidacy can sometimes be associated with a lot of emotional turmoil and this is especially true in case of children and the prelingual deaf adolescents. The issue of neural plasticity of the auditory cortex is now well recognised[38,39,40] and there is also constant pressure to reduce the age of cochlear implantation in profoundly deaf children in order to improve outcome. There is evidence to show that children implanted under 2 years of age achieve better open set speech recognition than children implanted after 2 years of age[41]. Implantation has now been undertaken in children between 5 and 11 months[42]. The assessment time in these children needs to be appropriately shortened while maintaining the best standards[43].

COCHLEAR IMPLANT TEAMS

A cochlear implant service is best provided by a multidisciplinary team[44] which in most places comprises of at least:

- Otologist
- Chief audiologist
- Audiological scientist
- Hearing therapist
- Psychologist
- Medical physicist
- Speech and language therapist

- Teachers of the deaf
- Co-ordinator

The prospective patient is seen jointly by team members at the assessment clinic, work up clinic, decision clinic and after implantation in the follow up clinics.
After initial referral a detailed history is taken enumerating birth details, family history, developmental milestones including motor skills and speech and language. Clinical examination includes a detailed otoneurological examination. A systemic examination is also important to rule out any associated anomalies.
Frank discussion about the patient's awareness about cochlear implantations and the expectations of the patient and the immediate family are very important in order to avoid future disappointments. Relevant patient information leaflets are supplied.
Speech and language are assessed and aided puretone thresholds are obtained followed by a trial of well fitting and powerful hearing aids. Some centres additionally perform electric response audiometry. A thorough vestibular assessment is also carried out.
CT / MRI scanning are crucial to visualising cochlear morphology and anatomical integrity of the auditory pathway.

COCHLEAR IMPLANT SURGERY

The surgery is performed under general anaesthesia. Facial nerve monitoring is routine. A post aural approach is common. The skin incision and the scalp flap are crucial considerations in order to avoid flap related problems. From large incisions of the early years the current trend is for smaller incisions and lesser scalp mobilisation. The chapters on surgical issues deal with this problem.
A cortical mastoidectomy is performed creating a cavity with overhanging edges. A bed for the receiver/stimulator package is drilled. The size, shape and site of this depends on the device and on the age of the patient and the shape of the skull. The facial recess is opened through a posterior tympanotomy and a cochleostomy is fashioned just anterior-inferior to the round window niche. The implant is secured in the bed and the electrode array is inserted gently into the scala tympani through the cochleostomy. The cochleostomy is then sealed with soft tissue plug and the wound closed in layers.

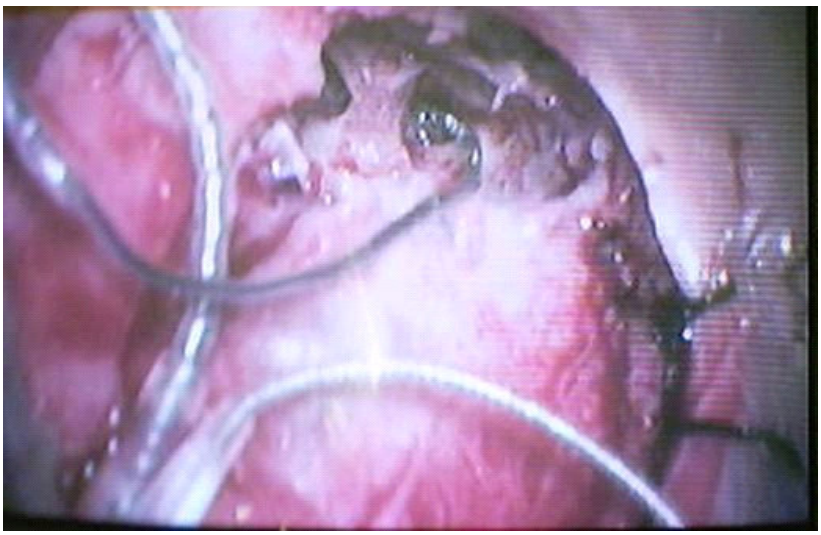


Figure 1. Mastoidectomy cavity with posterior tympanotomy. Electrode array visible in position through cochleostomy.

The concept of “soft surgery” is widely practiced for preserving residual hearing during implantation[45,46]. The technique involves deferring cochleostomy until immediately before electrode insertion, flattening the promontory with a large burr, cochleostomy with a small burr just anterior-inferior to the round window, exposing endosteum, smoothing bony edges, limited opening of scala tympani with preservation of the endosteum, no suctioning of perilymph, use of lubricant like hyaluronidase and gentle electrode insertion. Clinical and experimental data suggest this atraumatic procedure limits damage to the cochlea and preserves some residual hearing[47].

Most units perform some form of electrical testing on the implant perioperatively using the manufacturer provided software. This usually includes some form of neural response telemetry and integrity testing. Apart from reassuring the surgical team regarding the electrode placement and function the test provides a rough estimate of the threshold and comfort levels and can be used as a guide at the time of switch on.

An x ray is taken in some units to confirm correct insertion and alignment of the electrode array. The device is usually switched on in three to four weeks time.

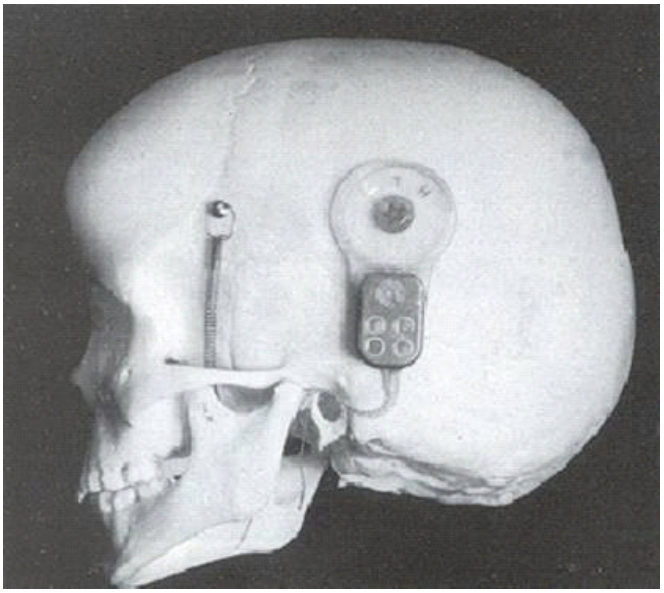


Figure 2. Surgical considerations in a paediatric skull

Cochlear implant surgery in the paediatric population requires special considerations particularly in relation to the delicate tissues and small dimensions. Allowances must be made for skull growth and subsequent electrode migration. There is no evidence that head growth is affected by the implant or that it would result in complete extrusion of electrode or implant[48,49,50]. A more vertical placement of the device may sometimes be needed in a paediatric skull.

Inner ear dysplasias were considered absolute contraindications in the past. Techniques for implantation of dysplastic cochleas have been described[47] and results vary according to the degree of abnormality. Severe forms of cochlear abnormality or auditory nerve aplasia do poorly. Two such cases are illustrated in the section on unusual cases[52,53] . Less severe forms of deformity like Mondini and large vestibular aqueduct have excellent results[54]. Children with congenital inner ear anomalies have a higher possibility of facial nerve anomaly. The incidence of abnormal course of the facial nerve can be as high as 16%[55] .There is also a potential for developing a cerebrospinal fluid gusher at the time of cochleostomy. Labyrinthitis ossificans is as much a problem in children as in adults. Advanced degrees of ossification can be overcome by short electrode insertion, scala vestibule insertion, bifid electrodes or cochlear drillout[56,57,58]. Cochlear implantation in chronically discharging ears have been discussed[59].

COCHLEAR IMPLANT OUTCOME

In 1985 John Ballantyne[60] wrote “every group working with implants – notably in the United States, Europe and Australia – has been able to claim some success, especially in terms of awareness of environmental sounds and of facilitation of lip reading skills; and a number of subjects by whatever mode of stimulation have been able in the short term to detect changes in rhythm and some of the prosodic features of speech. But the understanding of normal consecutive speech is still a long way off and the long term results are far from certain”

From this limited benefit, cochlear implantation has improved steadily over the last two decades and continues to improve further with better software and speech processing strategies. Most patients get very good closed set speech perception. Post lingually deafened adults and prelingually deafened children implanted promptly get very good open set speech perception. Factors closely related to speech perception abilities in implanted children are communication mode and educational setting[61]. Many of the children are able to attend mainstream education. Other factors include cause of deafness, age at onset of deafness and length of deafness[62]. Patients implanted for otosclerosis generally do well but non auditory stimulation especially that of the facial nerve can be a problem[63,64]. Transverse fractures of the temporal bone can cause severe disruption to the otic capsule and the results may be variable[65].

Recent surveys show that age at implantation is decreasing, children in oral education programs obtain more benefit from a cochlear implant than children in total communication programs, children who undergo implantation before 2 years of age show greater benefit than children who undergo implantation after 2 years of age and more children with good auditory skills before implantation and more residual hearing are undergoing implantation[66].

Different patients using identical devices may have different speech perception scores. The reasons for this are likely to be

- survival of neural elements in the cochlea
- proximity of the electrodes to the target neurons
- depth of insertion of electrode array
- integrity of central auditory pathway
- cognitive and language skills

As mentioned perilingually deafened adolescents may perform poorly and the outcome may not match with their expectations. This can result in the unfortunate outcome of elective non use.

COCHLEAR IMPLANTS AS A TOOL TO STUDY AUDITORY PHYSIOLOGY

Cochlear implants provide an unsurpassed access for studying the electro-physiology of the inner ear and the auditory pathway. The electrode array being adjacent to the spiral lamina allows both the best possible stimulation mode and recording arrangement in vivo. Various stimulation modes in combination with different current levels can be used to test specific segments of the cochlea. The implant evoked electrical auditory brainstem responses provide valuable information on the state of the inner ear and the auditory pathway. A specially adapted Nucleus 24 device was encased in a perspex case (Figure 3). This has a multipin socket for connection to the experimental recording set up (Figure 4). This has allowed direct recording of auditory electrical activity and has provided an insight into the patterns of maturation of the auditory pathway as a function of age[67]. Auditory neuropathy is another newly recognised contentious issue where the role of cochlear implantation has been debated[68]. Both these issues have been addressed in the section on electrophysiology.

Figure 3. External Cochlear Implant (Nucleus®) with connections

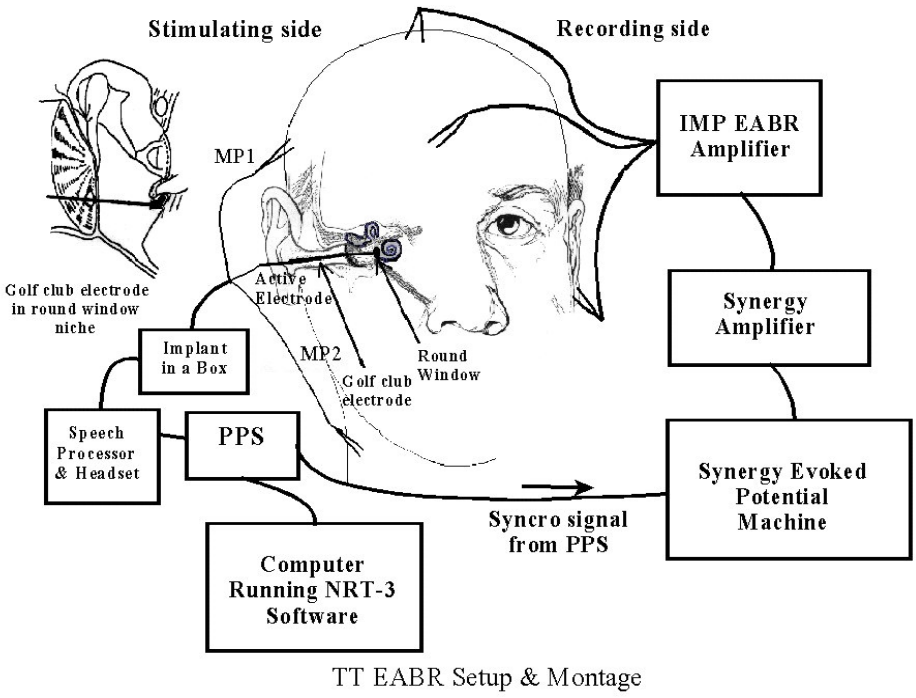


Figure 4. Set up for electrophysiological measurements.

THE FUTURE

Research into cochlear implantation continues to push the boundaries of expectation. As cochlear implant technology and outcome tends to improve the promising future possibilities look like becoming realities. These include:

- Binaural use of cochlear implants.
- Modified electrode design to utilise residual hearing.
- Hybrid acoustic / electric stimulation
- Totally implantable devices
- Auditory brainstem implantation

BACKGROUND TO THE WORK

The Midlands Adult Cochlear Implant Programme, Birmingham.

The midlands cochlear implant programme is based in Birmingham, UK. This was set up in 1990 and has grown over the years under the leadership of Mr. David Proops. Till date over 500 patients have been implanted in this programme and majority of the patients are funded by the National Health Service. The implants

used are the Nucleus device (Cochlear Corporation, Australia) Clarion device (Advanced Bionics, USA) and the MedEl device. The adult and paediatric services are administered separately. There is a smooth transition from one to the other at the age of sixteen. In addition to cochlear implantation the unit also undertakes middle ear implantation (Symphonix device) and insertion of Bone Anchored Hearing Aids.

The East of England Cochlear Implant Programme, Cambridge.
The East of England Cochlear Implant Programme is based in Cambridge at Addenbrooke's Hospital. The programme was established in 1986 with the help of the Cambridge Hearing Trust. All implants are funded by the National Health Service but the trust supports staff and research activities. In the early stages the Smith Nephew Richards Ineraid device had been used. Subsequently the programme has used the Nucleus device. The lead surgeon is Mr Roger F Gray. The unit also undertakes insertion of Bone Anchored Hearing Aids and runs courses for cochlear implant surgeons.

The Sydney Cochlear Implant Programme, Sydney, Australia.
The Sydney Cochlear Implant Programme was established in 1984 under the leadership of Professor William P R Gibson. Till date 950 patients have been implanted. The programme not only provides cochlear implant services to the residents of New South Wales but also has an overseas programme which caters to many south East Asian countries. There is a strong commitment to research which is funded through University of Sydney and charitable funds. In addition the Graham Fraser Memorial Foundation sponsors one UK otolaryngology trainee each year to go over and undertake research work with the Sydney group.

DATA COLLECTION

Most of the results are based on data gained from using the Nucleus® multichannel cochlear implant (Cochlear corporation, Australia). Any research work is dependant on data collection and analysis. The results and conclusion can only be as good as the quality of the data. For this purpose specifically designed database systems were used[69]. The process involved in designing and running such a system is described in the next chapter.

PURPOSE OF THE STUDY

The current study explores several issues relating to cochlear implantation. The first question is *whether smaller incisions and flaps reduce complication rates*. Sections 2.1 and 2.2 address this. Next we enquire *why implants need removing* and discuss *the results of reimplantation* in Sections 2.3 and 2.4. In Section 2.5 the results of a *technique to render a discharging mastoid cavity safe for implantation* are described along with analysis of intermediate term results. In Section 3.1 we investigate *why some implant recipients become limited users or non users* of their devices. The cochlear implant has proved an excellent tool to study the auditory pathway in vivo and chapter 7 uses implant derived electrophysiological data to investigate the *extent to which the maturation of the auditory pathway depends on auditory stimulation*. Section 4.2 explores the hypothesis that the term *auditory neuropathy actually comprises of separate subgroups with different aetiopathogenesis* and *different outcomes* following implantation. Sections 5.1 and 5.2 look at two challenging cases and explore the dilemmas facing implant teams in such situations.

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1.2

A COMPUTERISED COCHLEAR IMPLANT DATABASE SYSTEM

J. Ray

R.F. Gray

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ABSTRACT

In an environment of Clinical Governance with increased demands for accountability it is very important that accurate, reliable and secure data records be maintained for easy retrieval, analysis and presentation when required. A database is a very versatile tool for this purpose. We describe here our experience in designing a database for cochlear implant patients in Cambridge together with guidance for prospective designers in their chosen sub-speciality.

INTRODUCTION

The ability to collect, store, retrieve and analyse data is critical in providing a health care system that is timely, efficient and cost-effective[1]. Computer support is essential for such labour intensive works[2]. One useful computer tool is the database management system (DBMS). This provides a conceptual framework to assist in organising data and can physically store, maintain, retrieve and analyse this meaningfully.

Healthcare information needs to be shared for: (1) Audit (2) Clinical Governance (3) Research (4) Finance and (5) Data comparison between centres. Quite often various sub-units collect and store the data as is required for their field of work. This produces unnecessary data duplication and fails to recognise the potential for increased efficiency of integrating pertinent information to produce timely and useful reports[3].

Faced with the problems of data storage and retrieval and recognising the advantages of a DBMS we set out to design a database of all the patients who had received Cochlear Implantation at the East of England Cochlear Implant Programme.
This has proved to be very useful. The steps for setting up a database are discussed and can easily be applied to other areas of the speciality.

MATERIAL AND METHOD OF DATABASE DESIGNING

A database is a computer based information system where the stored data can be used by a wide variety of applications[4].

Figure 1. Numeric Coding System used for entering data. Summary table

Surname:	
First Name:	
Medical Records Number (CRN):	
Date of Birth:	
Sex:	1 = male 2 = female
Occupation:	
Operation date:	
Age at onset of deafness:	
Age at operation:	
Type of operation:	1 = normal cochlea 2 = obliterated cochlea 3 = congenitally deformed cochlea 4 = post CSOM fat obliteration of middle ear
Revision operation:	1 = yes 2 = no 3 = explant
Implant type:	1 = Ineraid 2 = Nucleus 22 3 = Nucleus 20 + 2 4 = Nucleus 24 5 = Single channel (RNID) 6 = Single Channel (Medel) 7 = Other
Aetiology of deafness in implated ear	0 = not recorded 1 = congenital idiopathic 2 = meningitis 3 = congenital progressive 4 = otosclerosis 5 = head injury 6 = CSOM 7 = Ototoxicity 8 = syndromal
Time Course of Deafness	
Left ear	0 = not known 1 = congenital 2 = sudden 3 = progressive
Right ear	0 = not known 1 = congenital 2 = sudden 3 = progressive

Planning and design

The most difficult stage was the planning stage. Careful thought was required when designing the database. As much as possible was planned on paper[5]. It is most important to consider what data will need to be extracted in future. This in

turn determines the data to be collected and stored and how they relate to each other. Needs of all potential users was investigated and draft paper copies were circulated for approval by individual members. This exercise in itself streamlined the data acquisition process and spotted several areas of duplication of data. Free text was avoided to minimise ambiguity and confusion. All data was coded (Figure 1) using existing nomenclature used regularly in the department.

Choice of software package

Choosing a DBMS which suits the purpose was not difficult. An available existing system (preferably millennium proof) usually proves to be cheaper and user friendly because of familiarity and compatibility amongst users. We chose Microsoft Access as this was freely available on all the computers in the department. However any available software package would do as long as it fulfils the purpose.

Tables

This is the basic framework to store information and is the equivalent of a file (Figure 2). Each *entity* (e.g. patient) is a *record* and is displayed in horizontal rows. Each *attribute* that describes the entity (e.g. surname, address, symptom etc) is a *field* and is displayed in columns. Although entities may share attributes, an unique attribute (called the *Primary key*) is used to identify an entity. The primary key serves a very important purpose as we shall see later. We have used the patient's hospital number (CRN) as the primary key.

The nature of information needed and collected by different sub units of the department are different. Therefore instead of storing information in one large table it is much more convenient for inputting and storing if this is broken up into smaller tables. A master table holds the summary of patient details and forms the minimum core dataset (Figure 1) by which an individual patient or groups of patients can be identified. The smaller tables contain only a certain aspects of the patients' dealings with the department. We divided our tables (Figure 4) broadly according to the sub-units which deals with the patients separately e.g. patient details, audiology, radiology, surgery, complications, switch-on and tuning.

Sections on follow-up were dealt separately and repeated at each follow-up visit. Adults and children were also dealt separately due to the difference in the test battery.

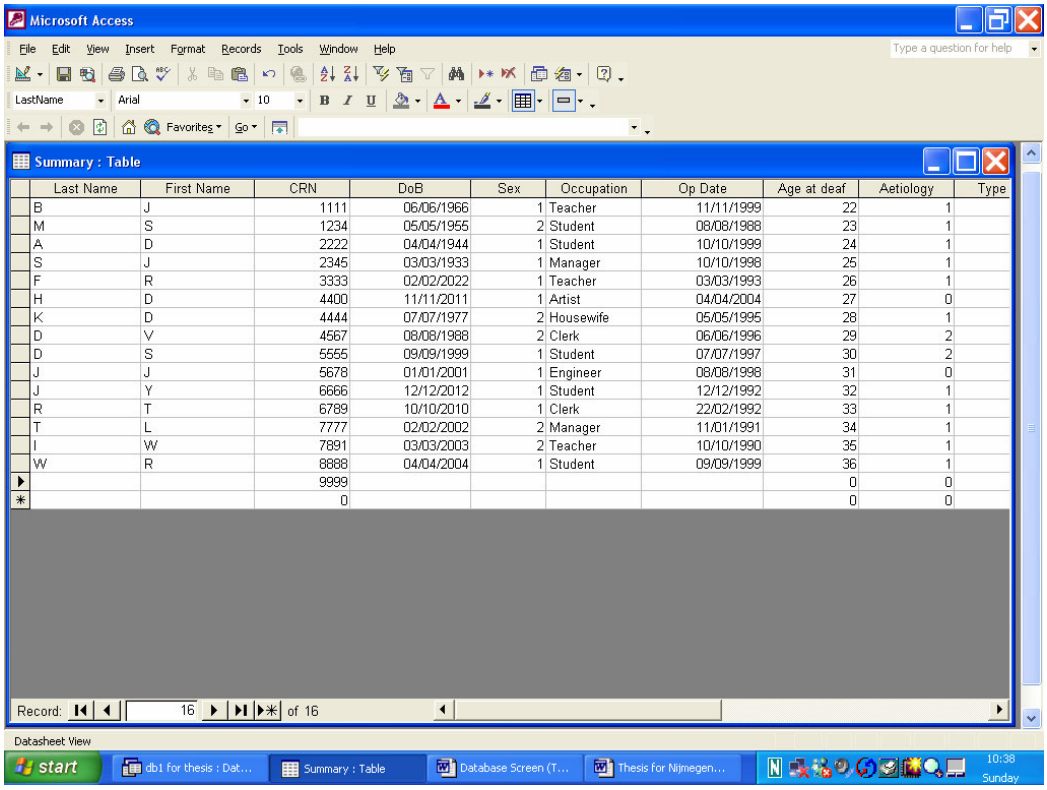

Figure 2. Table structure

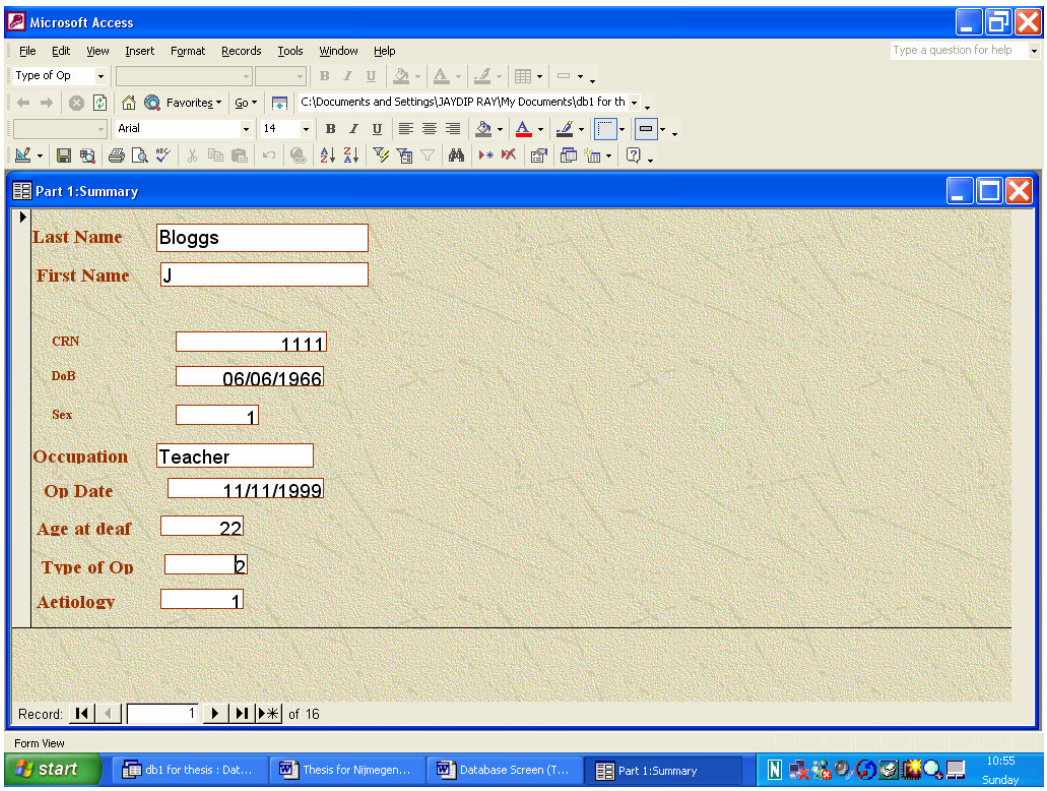

Figure 3. Form view for entering, viewing and editing data.

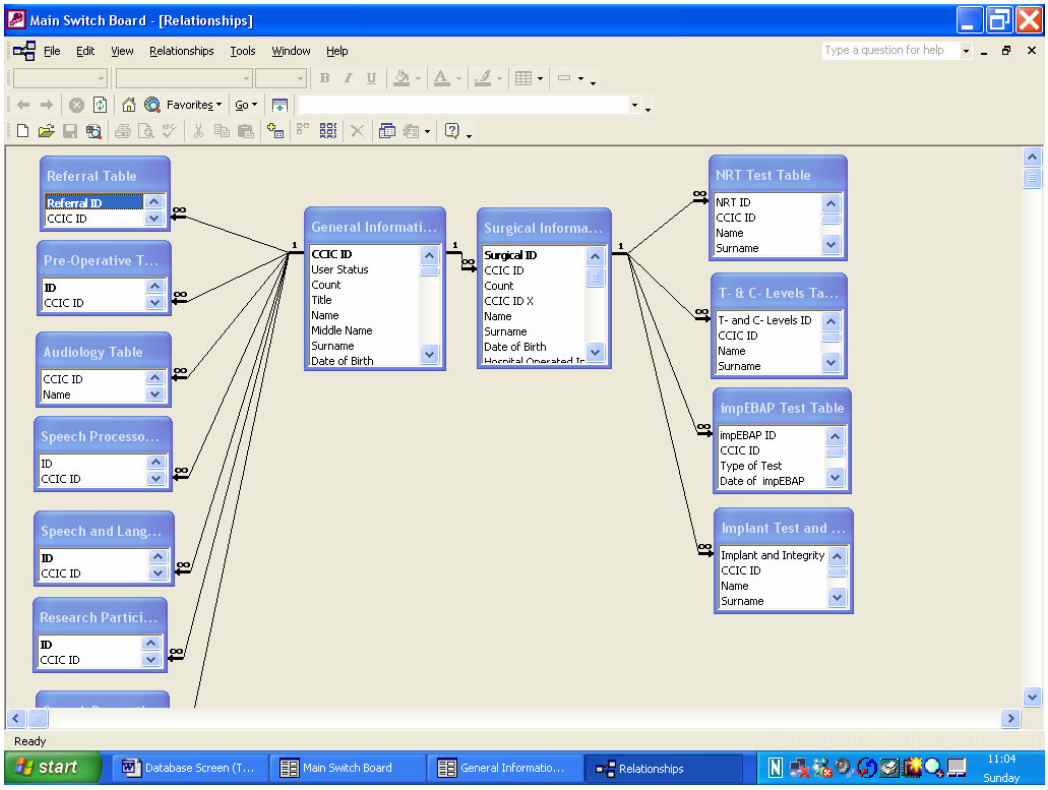

Figure 4. Various tables and their relationships.

Forms

Consideration must be given to the needs of each individual user who will collect and input the data. Forms (Figure 3) were designed to view, input, edit, control and present data easily. The ease of inputting data is important and simple forms to facilitate this ensures compliance amongst users and keeps the database up to date.

Relationships, Primary key and Referential integrity

A database with multiple tables needs to be cohesive and therefore a *relationship* has to be established between the tables. Relationships can be one-to-one, one-to-many or many-to-one. We used the commonest relationship i.e. “one-to-many”. Thus all the smaller tables were linked to the master table (Figure 4). This implies that a record from the master table can have more than one matching record in a second table but the reverse cannot happen e.g. one patient can have many test results but each test result matches only one patient. This has been ensured by establishing what is called *referential integrity.* Referential integrity helps to synchronise data in related tables and prevents the data from getting out of step. This is crucial for accuracy and reliability. It also prevents deletion of data from the

master table by any editing in the peripheral tables. In our database the backbone of relationships and referential integrity has been the *primary key* (patient hospital number). This set up also helps trap duplicates at the point of entry thus maintaining accuracy.

Confidentiality

The final aspect that is of paramount importance is the issue of confidentiality and security. This has been resolved by utilising a unique user password to determine who has access to the data. It can be taken one step further by organising users into groups and setting security levels. However ours being a small cohesive unit this was unnecessary. At present the database exists on password secure laptop computers which are easily transportable to assessment, operative and rehabilitation sites. The data thus collected is subsequently transferred to a main computer which is owned by the Implant Programme under overall supervision of the programme manager and maintained by the IT department of Addenbrooke's Hospital.

Current Position

After the above steps have been done the database was ready for entering and storing information. As the Cochlear Implant Programme at Cambridge had been in existence for more than twelve years, enormous amount of data had to be transferred from the files to the database. But once the backlog had been cleared it became easy to update records of every consecutive patient that joins the programme. At present this is done on the coded paper hard copy of the forms. The data is then transferred manually onto the computer by the database manager at the earliest opportunity. With increased familiarity amongst users data can be directly entered into the computer using the form view.

With the system up and running information can now be extracted, analysed and presented in a meaningful manner using the built in facilities of Report and Query wizard.

DISCUSSION

The usefulness of databases in clinical practice is already well recognised and the "Impeval Data-ease" database used for the Evaluation of the National Cochlear Implant Programme in 1990-94 is a prime example[6]. The Head and Neck

Database developed recently by the British Association of Otolaryngologists and Head & Neck Surgeons is another example.

A good database must provide *(1) Data Integrity* i.e. ensure that the data is accurate, consistent and reliable *(2) Data Security* i.e. data should not be lost *(3) Data Accessibility* i.e.data should be available in a meaningful way to all users who need it and *(4) Data Confidentiality* i.e. protect it from access and alteration by unauthorised users[5]. Microsoft Access fitted these requirements very well. Access is a very powerful DBMS and can store almost limitless amount of information yet it is easy to use with tremendous flexibility and control over data. It allows Dynamic Data Exchange (DDE) and compatibility with other applications like spreadsheets with basic statistical applications (e.g. Excel) and slide presentations (e.g. Powerpoint). The feature of Object Linking an Embedding (OLE) can be used to include scanned images (e.g. x-rays, scans, operative photographs). These can be projected directly from the database during presentations and can also prove very useful in medicolegal issues.

Well structured forms prevent things from being forgotten[7] and ensures staff acceptance and compliance thus achieving a comprehensive data entry[8]. This is important because completeness and accuracy of the data entered determines the quality of the database[9]. In designing our tables and forms we have used drop down boxes to provide prompts on the coding system used. This allows direct entry of data without the need to refer to paper copies. A numeric coding system also facilitates easy data entry and retrieval[10]. The initial apathy of transferring data on to the computer has now passed and user friendly forms with prompts have ensured compliance even amongst the most reluctant users.

The use of the patients hospital number as the primary key has ensured data accuracy.
To avoid data duplication only the hospital number recorded in the notes of the cochlear implant programme is used and any other number from other hospital files are ignored.

At present the database is not linked to the hospital information system for reasons of hospital data protection and held on a main computer under overall responsibility of the manager of the implant programme. In future it might be possible to have a central database with multiple user terminals with high level of integration with hospital information systems[11] and with varied user data access

security levels. The eventual aim would be to integrate comparable databases at different Cochlear Implant Programmes.

As newer equipments, tests and surgical procedures continue to emerge the designs of the forms and tables can easily be edited to encompass changing data collection needs without affecting the existing data.

CONCLUSION

The data that is used by an organisation is one of its valuable resources and is expensive and time consuming to gather. It is therefore essential that the data be organised and arranged so that best use can be made of it.

A database system is accurate, robust, timeless, time saving and convenient to use with ease in generating reports. Now that computers in the workplace are a fact of life it will not be long before databases to store and maintain data become a necessity rather than a luxury.

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CHAPTER 2

SURGICAL ISSUES

2.1

SURGICAL COMPLICATIONS OF 844 CONSECUTIVE COCHLEAR IMPLANTATIONS – LARGE VERSUS SMALL INCISIONS

J. Ray
W. Gibson
H.Sanli

Cochlear Implants International (In press)

ABSTRACT

Objectives: To evaluate the long term difference in wound and flap problems between large and small incisions and the problems encountered with primary and revision cochlear implant surgery

Patients: 844 consecutive patients underwent cochlear implantation at the Sydney Cochlear Implant Centre. 212 cases were operated on prior to October 1994 using the retroauricular 'C' shaped incision or a post auricular incision with a horizontal posterior limb. After October 1994 a new small vertical post aural incision was used in all patients. Post operative problems were analysed.

Study design: Prospective longitudinal study of cochlear implantees from 1984 to 2003.

Setting: Tertiary care referral centre.

Intervention: Change in incision for cochlear implantation.

Main outcome measure: Wound and flap problems.

Results: 5 patients out of 212 (2.3%) in the first group encountered wound and flap problems. In comparison 7 out of 632 patients (1.10%) from the later group experienced wound and flap problems using the new incision. 80 out of the total 844 patients underwent revision procedures for various reasons

Conclusion: There is a reduced incidence of wound and flap problems with small skin incisions and minimal scalp mobilisation. Device failure, wound and flap problems are still the commonest causes of explantation. Performance of the replacement device usually was similar to the original device and was not related to the aetiology of deafness or to the cause of explantation. This data will be useful in counselling patients for reimplantation / revision surgery.

INTRODUCTION

Cochlear implantation has established itself as a reliable way of providing an auditory input to profoundly deaf patients[1]. No major perioperative life threatening complications have been reported although recently concerns have been raised regarding late onset meningitis in infant recipients[2,3]. However complications still occur and such events have serious financial and psychological implications for both the patient and the implant team. The devices like any other electrical devices are liable to occasional malfunction or even breakdown. The surgical process itself carries its own share of complications that may compromise the functioning of the

device requiring it to be removed. One such event is wound infection and flap breakdown.

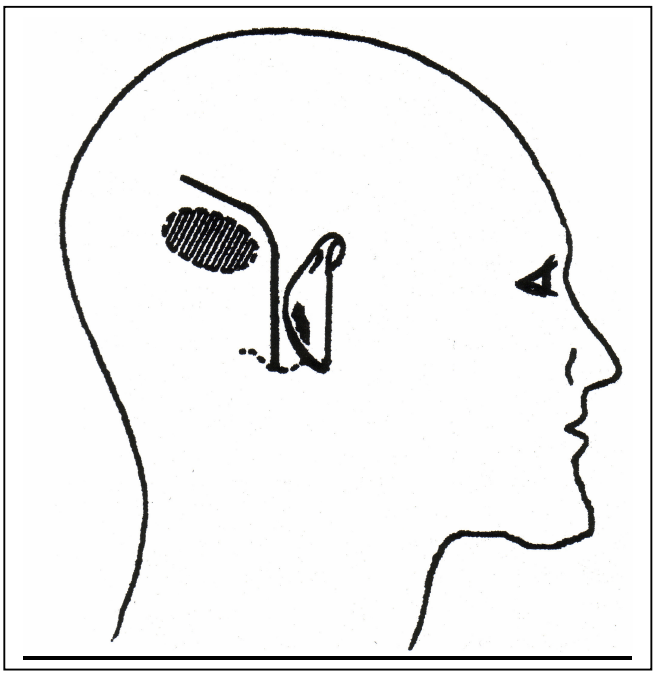

Figure 1. Vertical incision with horizontal limb

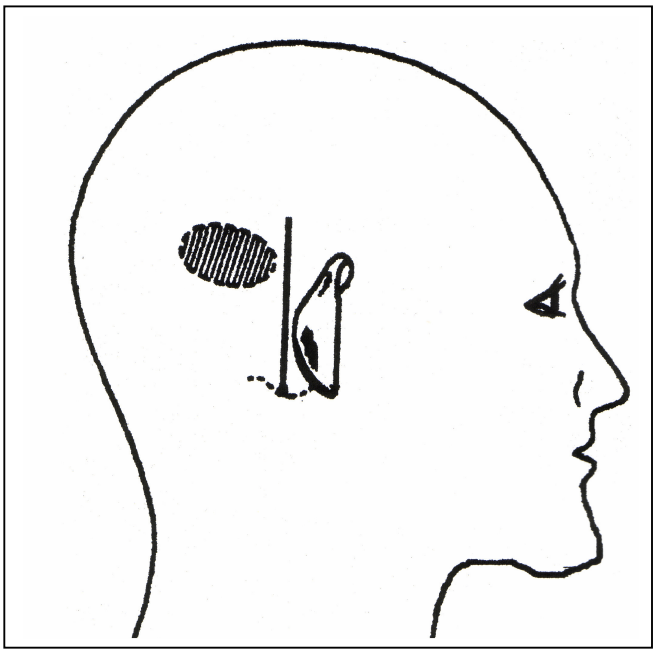

Figure 2. Small vertical post aural incision

We have looked at our data on 844 consecutive cochlear implant patients to analyse the post operative problems encountered between 1984 and 2003. The wound and flap problems have been specifically analysed relative to the patients' chronological position in the surgical series.

PATIENTS AND METHODS

The study is a retrospective review of all cochlear implant patients who had undergone initial or subsequent cochlear implant surgery between 1984 and 2003. All patients were under the care of the senior author (WPRG). All patients were categorised according to the aetiology of deafness, and the time and type of problems encountered.

All patients underwent detailed preoperative audiological, electrophysiological and radiological investigations. Adult patients underwent vestibular assessment and promontory stimulation.

The surgical procedure has evolved through the last nineteen years. The initial incision was a large retroauricular 'C' shaped incision which was followed by a vertical postaural incision with a horizontal posterior limb (Figure 1). After 1994 all patients underwent implantation using a new incision (Figure 2). This involved a small (3-7cm) vertical post aural incision (Figures 2 and 3) with limited mobilisation of the scalp[4].The implant was housed in a tight periosteal pocket above the temporo-parietal suture line. The wound was closed with absorbable sutures. The details of the procedure are to be found in an article published from this programme[4]. Intra-operative integrity testing was conducted on all implants. In addition implant evoked auditory brainstem responses and neural response telemetry was conducted to check the array is correctly positioned within the cochlea and that the spiral ganglion was viable in different areas. Post operatively speech perception was assessed using the Melbourne Speech Perception Categories[5]. This is a validated speech perception measure after cochlear implantation where patients are scored between 1 and 7 in ascending order of performance (Table1).

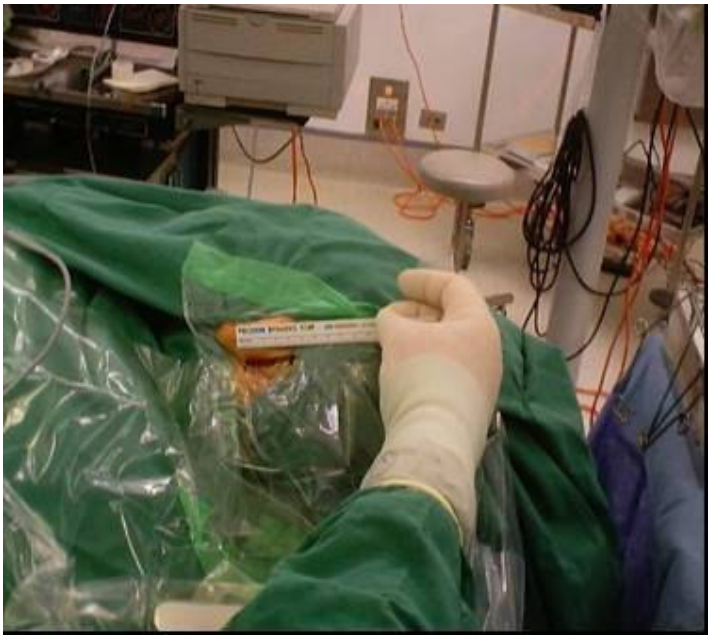

Figure 3. Small Post aural incision

Table 1. Melbourne Speech Perception Categories

Category	Description
1	Detection of speech sounds only
2	Discrimination of suprasegmental aspects of speech in addition to 1
3	Discrimination and recognition of vowels in addition to 1 and 2
4	Discrimination and recognition of consonants in addition to 1-3
5	Minimal open set speech perception in addition to 1-4
6	Open set speech perception (>20% phoneme scores for PBK words)
7	Good open set speech perception (>50% phoneme score for PBK words)

RESULTS

Patient demographics

A total of 844 patients underwent cochlear implantation in the Sydney Cochlear Implant Centre between 1984 and 2003. 212 were done prior to 1994 and 632 were done after 1994.The age range was 5 months to 92 years (Mean 27.1 years). Fifty-six patients belonged to the overseas programme and had been implanted in Sydney after being referred from Southeast Asian countries. 80 patients underwent revision procedures during this period. The range of follow up was 19 years to three months (mean 7.8 years)

Of the total 80 revision surgery candidates there were 42 (52.5%) adults and 38 (47.5%) children in the series. 47 (58.75%) were male and 33 (41.25%) female. The Nucleus® device (Cochlear Corporation, Australia) was used in all 844 patients at the Sydney centre. Ten patients needed more than three revision episodes.

Types of revision procedures (Table 2)

Of the 80 patients 52 (65%) underwent explantation and ipsilateral reimplantations while 20 (25%) underwent explantation and contralateral reimplantation. Five (6.25%) had repositioning the device and thinning the flap and two (2.5%) had reinsertion of the electrode array. One (1.25%) procedure was to replace the magnet in the housing and reinforce the area. Three devices were upgraded during the revision procedure.

Table 2. Type of procedure

Procedure	Proportion
Explant and ipsilateral re-implant	52 (65%)
Explant and contralateral re-implant	20 (25%)
Repositioning	5 (6.25%)
Reinsertion	2 (2.50%)
Reinforcing the magnet	1 (1.25%)

Intra-operative problems (Table 3)

Eleven patients (1.3%) with labyrinthitis ossificans needed drillout procedures to position the electrode array. In one child there was unanticipated fibrous occlusion of the scala tympani. The procedure had to be abandoned, however the contralateral ear was operated on during the same session and the cochlea was found to be patent so implantation proceeded uneventfully. Post-operatively good speech scores were obtained (7 on Melbourne Categories).

Full insertion was not possible in 24 (2.84%) patients for various reasons. The range of partial insertion was 5 to 21 electrodes (mean 17 electrodes). 14 were due to labyrinthine ossification in post meningitis patients and two had common cavity deformity of inner ears. In 3, (0.35%) there was unexpected difficulty in electrode insertion possibly due to fibrous occlusion. In the remaining 7, no obvious cochlear abnormality was evident. The range of speech scores in these patients was 1 to 7 (Melbourne Categories) and the mean score was 4.7.

Table 3. Intra-operative Problems

Problem	% of total
Abandoned implantation	1 (0.12%)
Partial insertion	24 (2.84%)
Meningitis	
Drill out	11 (1.3%) (meningitis) basal turn drilled on promontory)
Double array	3 (0.36) (meningitis)
Common cavity	2
Fibrous obliteration	3
No obvious cochlear abnormality	5

Post operative problems (wound and flap related) (Tables 4a and4b)

Wound and flap related problems were experienced in 12 (1.42%) patients (16 episodes). There were 5 (2.35%) patients in the initial group of 212 where the larger incisions had been used. There were 7 (1.10%) instances of wound and flap problems in the later group of 632 where the smaller vertical incision was used[2].

Table 4a. Post operative problems (wound related)

Problem	% of total
Infection	11 (wound) 1 (middle ear – fat & blind sac) Total 12 (1.42%)
Non-auditory stimulation (VII)	5 (0.59%)
VII temporary weakness	1 (0.12%)
Repositioned	5 (0.59%)
Erosion of EAC/TM	2 (0.24%)

Table 4b. Effect of change in incision

Period	Total Numbers	Wound complications (%)
Pre 1994 (Large incision)	212	5 (2.35%)
Post 1994 (Small incision)	632	7 (1.10%)

In one patient the middle ear had been previously obliterated with autologous abdominal fat with blind sac closure of the external canal. The external canal had broken down due to infection and the fat had diminished in size. The device was removed and the procedure was revised. Explantation had to be carried out in all twelve patients. Staged ipsilateral reimplantation was undertaken in 2 (0.24%) while contralateral reimplantation was undertaken in 10 (1.18%). Full insertion of all active electrodes was achieved in all.

Five patients experienced problems due to excessive scalp thickness over the receiver stimulator package. All were boys who had been implanted at an early age and had grown to massive proportions as teenagers. These cases had to be revised and in 4 of these the receiver stimulator had to be repositioned to a more superiorly place site.

Reconstruction of the posterior canal wall had to be undertaken in one patient. In another patient the ear drum had to be repaired. In both cases the electrode array had extruded and had to be reinserted and anchored with soft tissue. There was no change in the speech perception scores after revision in both cases.

Post operative problems (device related) (Table 5)

Total device failure occurred in 17 (2.01%) cases. Majority 13(1.54%) were unexplained while in 2 (0.24%) the precipitating cause was direct trauma to the implant. Sudden electrostatic discharge was responsible for device failure in 2 (0.24%), one being due to a lightening strike.

Table 5. Post operative problems (device related)

Problems	% of total
Device malfunction ('perceptional')	4 (0.47%)
Device malfunction (confirmed)	16 (1.89%)
Device breakdown (cause unknown)	13 (1.54%)
Device breakdown (user related)	4 (0.47%)
Total device related problems	37 (4.38%)

In 20 patients (2.37%) there was suboptimum performance from the device. Four cases (0.47%) were categorised as 'perceptional' as it was reported by the patients without any demonstrable loss of implant integrity (2 patients reported uncomfortable cracking noises heard through the implant and 2 experienced loud bangs through the implant on several occasions). In 16 (1.89%) the problems with the device could be demonstrated by electrical testing. All patients underwent explantation and ipsilateral reimplantation. In five of these patients speech perception scores improved on an average by 2 grades (Melbourne Categories) after revision surgery. In the remainder speech scores remained the same and there was no deterioration in any patient.

Post operative problems (others) (Table 6)

Non auditory responses in the form of facial nerve stimulation was experienced in 5 (0.59%) patients. The aetiologies in these patients were common cavity and otosclerosis.

Table 6. Post operative problems (others)

Problems	% of total
Dislodged magnet	1 (0.12%)
Extrusion of electrode array	2 (0.24%)
Extrusion of device	2 (0.24%)

Two extrusions of electrodes occurred (0.24%). These were reinserted in due course and the cochleostomy and the posterior tympanotomy sealed with soft tissue.
A partial and temporary facial weakness was experienced in one patient despite continuous facial nerve monitoring. The facial nerve canal was intact and the complication is thought to have arisen from heating from the drill. Fortunately this patient made a complete recovery over the next three months.

An unusual and hitherto unreported problem occurred in a six year old patient about two years after implantation. The magnet seemed to pop out of its housing several times without any precipitating factors. Initially it was possible put it back in place by gentle manipulation. Eventually it became necessary to explore the area through a separate incision and the magnet was repositioned in the casing using non absorbable sutures in the surrounding tissue to retain it in place. The device itself remained uncompromised.

One patient had left sided congenital profound deafness and used conventional amplification hearing aids on the right side. He subsequently lost hearing in the right ear and the ear was implanted. Six years later there was progressive loss of speech perception despite a functional implant and a large acoustic neuroma was discovered. The implant was explanted at surgery for removal of the acoustic neuroma and subsequently reimplanted in the contralateral ear.

DISCUSSION

Over the last two decades cochlear implantation surgery has evolved through various techniques and devices. Unfortunately there are occasions when the implant needs to be removed and replaced. Despite the increasing experience that has come with the growing number of implant surgeries over the last decade, data on failures and explantations has been scanty.

The initial evaluation of the safety of cochlear implantation in the UK was undertaken by the Medical Research Council in 1994[1]. Overall complication rates were very low although patient numbers were small. Wound and flap problems have been the commonest complication experienced in many series[6,7]. It is slowly becoming clear that a smaller incision with minimal scalp mobilisation is associated with lesser wound and flap problems[8,9].
This series confirms that smaller incisions are associated with less complications. The wound and flap problems were halved in this study with use of a small (3-7cm) vertical post aural incision with minimal mobilisation of the scalp tissues[4]. The wound healed unobtrusively and without any compromise of the vascularity or viability. Minor infections were easily controlled by intensive antibiotic treatment[10,11]. Nevertheless 12 infected wounds (1.42%) failed to settle down with a conservative approach and an explantation / reimplantation procedure was necessary. These figures were clinically significant but failed to reach statistical significance as the number of wound infections were small.

In cases of middle ear infections, the electrode array was cut close to the cochleostomy and the main body of the device was removed. The electrode was retained as a “lumen keeper” and this maintained the patency of the cochlea until reimplantation was possible[6,12].

Reimplantation was first reported by Hochmair-Desoyer and Burian Hochmair-Desoyer and Burian 1985[13]. Although the internal device design has improved, device failure continues to account for majority of the explantations[14]. Failure rates of 3% for adults have been quoted for the Nucleus devices[15]. In our series the overall device failure problems rate was 2.01%. Histological studies[16] have shown that the trauma from explantation and reimplantation is no greater than the implantation itself.

Overall explantation / reimplantation rates between 8.5%[17] and 10%[18] have been reported. Miyamoto et al[18] have stated that in the majority of reimplantations the depth of insertion and the number of active channels remained unaltered. In our series, explantation / reimplantation rates were around 8.53% which is comparable to similar series reported from other centres.
There was no relationship between the performance of the replacement device and the duration of original device use, surgical complications, insertion depths and preoperative variables. Similar findings have been reported in a retrospective

multicentre study of reimplantation surgery[19] for twenty-eight failed Nucleus 22 cochlear implants in eighteen US implant programmes.

CONCLUSIONS

Several key features about cochlear reimplantation surgery have been evaluated in this study and this data has been useful in counselling patients.

Device failure, wound and flap problems are the commonest causes for explantation, reimplantation or revision. Larger skin incisions and wider scalp mobilisation are associated with increased wound and flap problems.

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2.2

MEDICAL AND SURGICAL COMPLICATIONS OF SECOND HUNDRED CONSECUTIVE ADULT COCHLEAR IMPLANT PATIENTS IN BIRMINGHAM

S. Dutt
J. Ray
E. Hadjihannas
H.R. Cooper
I. Donaldson
D.W. Proops

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ABSTRACT

One hundred consecutive adult cochlear implant patients numbered 101 to 200 in the series were prospectively monitored and data collected along the same protocol as for the first 100 patients. The study period was 1999 to 2001. The total number of procedures was 122. Change in practice after first hundred implants included a standard surgical technique with smaller incision and lesser flap mobilisation and routine facial nerve monitoring. As far as practicable the same commercial type of implant was used. There were 111 (91%) implantation episodes, 5 (4%) explantation episodes, 4 (3.3%) reimplantation episodes and 2 (1.7%) revision procedures. 89 patients underwent unilateral implantation and 11 underwent bilateral implantation. Major complications included flap breakdown (1.6%), extrusion of electrode array (0.8%) and device failure (0.8%). Overall major complication rate was 3.2% (4/122). The overall minor complication rate was 18%. In addition 7.3% experienced transient vertigo and 5.7% local discomfort lasted 3 days on an average with complete resolution. The overall complication rate in the second hundred implant patients is lower than the first hundred. Smaller incision and lesser flap mobilisation has reduced the minor complication rate.

INTRODUCTION

At the Midland Adult Cochlear Implant Programme in Birmingham 252 adult patients have been implanted till the end of 2001. The outcome and complications in the first hundred patients in the series have been analysed in a previous paper from this programme[1]. This article follows on from the first one and forms part of the ongoing process self monitoring of performance.

One of the earliest reports on complications of cochlear implants came from a survey of surgeons in the US[2] and a major complication rate of 4.8% was reported. Subsequent survey on a larger cohort showed an overall major complication rate around 10%[3].The safety of cochlear implantation in the UK was initially evaluated from pooled data by Summerfield and Marshall[4]. They have followed the same cohort of patients through till 1998 and reported an overall major complication rate of 8%[5]. Other authors have since reported on their complications and lessons learnt from them[6,7,8,9] .

This paper looks at the experience gained from the second cohort of hundred patients and compares it with the first cohort and also compares it with the available literature. This data is useful for preoperative counselling of patients.

PATIENTS AND METHODS

The study is a prospective longitudinal survey of one hundred consecutive adult cochlear implant patients with serial numbers 101 to 200 in the series in the Midland Adult Cochlear Implant Programme.

Since the analysis of our first one hundred patients, medical, surgical and audiological data was prospectively collected in the same format as for the first cohort. As before the patients filled in a self response questionnaire while the medical personnel filled in their relevant forms.

All patients were categorised according to their age at implantation, aetiology of deafness, position in the series, time and type of problems encountered and remedial actions taken.

Complications were divided into major and minor and also into perioperative and post operative problems. The latter was further divided into flap related, patient related and device related problems.

All patients underwent detailed preoperative audiological and radiological investigations. The surgical procedure had been consistent all through the series. All patients were operated by one of the senior authors (DWP or ID). A “lazy S” post aural incision (4-5cm in length) (Figure 1B) with limited mobilisation of the scalp was used in all cases. A separate superiorly based periosteal flap was used and the wound was closed in two layers. Facial nerve monitoring was employed in all case throughout the procedure and neural response telemetry was conducted perioperatively to test integrity of the device and of the auditory pathway. Perioperatively three doses of prophylactic antibiotics were used. A post operative radiograph was taken on the next day and the device was switched on after one month.

RESULTS

Out of the second hundred adult cochlear implant patients 41 were male and 59 were female. The mean age at implantation was 51.3 years (range 17-76 years). The main aetiology was idiopathic (47%) followed by otosclerosis (8%), hydrops

(7%) and meningitis (7%). Table 1 shows a detailed break up of the various aetiologies.

Table 1. Etiology

Etiology	Total patients (%)
• Idiopathic	47
• Otosclerosis	8
• Meningitis	7
• Hydrops	7
• Measles	5
• Head injury	5
• Ototoxicity	4
• Genetic (Non syndromic)	3
• LVAS	3
• CSOM	3
• Usher Syndrome	3
• Rubella	2
• Mumps	1
• Blast injury	1
• Wegener's Granuloma	1

The total number of procedures undertaken in these patients was 122. Of these 111 were for implantation (including 89 unilateral en 11 bilateral implantations), 5 were for explantation, 4 for reimplantation and 2 for revision surgery. The average hospital stay was 4.3 days (Range 3 to 10 days). (Table 2) During the early part of the series hospital stay was longer but became shorter subsequently. The longest stay of ten days was due to a stubborn wound infection which initially failed to respond to conservative treatment.

Table 2. Surgery

• Unilateral Implantations	89
• Bilateral Implantations	11 9 straightforward, 1 problems on contralateral side, 1 no response on contralateral side.
• Implantation • Explantation • Reimplantation • Revisions	111 (91%) 5 (4%) 4 (3.3%) 2 (1.7%)
• Total Procedures	122 (Patients:100)

Overall major complication rate was 3.2% (4/122). The overall minor complication rate was 18.85%. All the complications are listed in Table 3. These have been described as peri-operative and post-operative problems. The latter has been further divided into flap-related, patien- related and device-related problems.
Major complications included wound breakdown in two (1.6%), extrusion of electrode array in one (0.8%) and device failure in one (0.8%).(Table 3)

Table 3. Complications

Intraoperative Problem		Number
Damage to Chorda tympani		10 (8.1%)
Damage to tympanic annulus and/or posterior meatal wall		6 (4.9%)
Exposure of facial sheath		1 (0.8%)
Soft tissue occlusion of basal turn (after cochleostomy)		2 (1.6%)
Partial ossification		3 (2.4%)
Drillout		1 (0.8%)
Simultaneous repair of tympanic membrane		1 (0.8%)
Postoperative problems		
Flap problems:	Number(%)	Comments
Wound infection	11 (9.0%)	i.v. antibiotics
Flap breakdown	2 (1.6%)	Explant / Reimplant (contralateral)
Thick flap	1 (0.8%)	Revised electively
Intermittent flap oedema	1 (0.8%)	Treated conservatively
Haematoma	2 (1.6%)	One needed aspiration
Patient related problem		
CSF Leak	1 (0.8%)	Detected 3ds later. Spont. resolution
Otitis externa	1 (0.8%)	i.v. and topical antibiotics
Middle ear infection	1 (0.8%)	i.v. antibiotics
Severe vertigo	1 (0.8%)	Lasted 4 days. Resolution in 10 days
Transient vertigo	9 (7.3%)	Avg 3 days. Complete resolution
Local pain / discomfort	7 (5.7%)	Avg 3 days. Complete resolution
Tinnitus worsened	2 (1.6%)	Counselling and remapping
Non auditory stimulation	2 (1.6%)	Remapping
Implant related problems		
Electrode protrusion	1 (0.8%)	Reinserted and wound repaired
Device Failure	1 (0.8%)	Reimplanted

Intra operative problems

The chorda tympani was inadvertently lost during surgery in 10 (8.1%) cases. There was concomitant injury to the posterior meatal wall and the tympanic annulus in 6 (4.9%) cases. In one patient there was a pre-existing drum perforation which was repaired at the time of implantation. The facial nerve sheath was exposed in one during drilling of the posterior tympanotomy. However there

was no facial weakness. There were no instances of post operative facial nerve weakness in the whole series.

Soft tissue occlusion of the cochlear lumen at the site of cochleostomy was encountered in two patients. This was confined to the basal turn and was successfully overcome. Complete insertion of electrode array was possible in one while the other one had partial insertion. The aetiologies in these patients were idiopathic and congenital hearing loss.

Severe ossification of the basal turn necessitated a “drillout” to identify the cochlear lumen in one patient with otosclerosis. Partial insertion of the electrode up to 8 rings was possible here. Partial ossification was encountered in three other otosclerotic cochleas but implantation proceeded uneventfully with full insertion in one case. In the other two cases difficulty in insertion of the electrode array was encountered and both had partial insertion with less than 15 electrode rings in the cochlea.

Post operative problems

Wound infection was encountered in eleven cases (9%). All occurred in the immediate post operative period and were managed effectively by prolonged course of antibiotics.

In one patient dehiscence of the overlying flap occurred several months after the initial implantation procedure. After conservative treatment failed to heal the area the implant was removed and staged contralateral reimplantation carried out. The other case of flap breakdown occurred within a month of surgery and again explantation and staged contralateral reimplantation was carried out. Both patients were male and over 60 years of age.

Two episodes of post operative wound haematoma were encountered one of which had to be aspirated. The implant performance or flap integrity were not compromised. Otitis externa and otitis media were reported in one each and was managed effectively by oral and topical antibiotics. Unexplained intermittent flap oedema not requiring surgical intervention was observed in one patient.

Delayed cerebrospinal fluid leakage occurred in one patient 3 days after the implantation. This patient had a tear in the middle fossa dura at the time of initial surgery that was repaired with temporalis fascia, surgicel and ‘fibrin glue’. The leak was treated conservatively and settled spontaneously in a day without recourse to re-exploration.

Two patients (2.6%) reported worsening of their tinnitus after implantation and this was managed by tinnitus counselling and remapping. 7.3% experienced transient vertigo and 5.7% local discomfort lasted 3 days on an average with complete

resolution before discharge from the hospital. Severe vertigo occurred in the immediate post operative period in one elderly female. This took about 14 days to abate completely.

Two other patients experienced non auditory stimulation in the form of facial nerve twitching when their implants were working. One of suffered from cochlear otosclerosis and had a partial insertion. The other patient had deafness of idiopathic origin and had a complete insertion of the electrode array. Both the cases were treated by altering the mapping and the speech processing strategy.

Device related problems

There was one case of unexpected device failure involving a Nucleus 22 implant. This was removed and replaced with a new Nucleus 24 implant.

The other device related major complication was late electrode extrusion from the cochleostomy and through the tympanic membrane (anecdotally referred to as the 'Nessie sign' after the Loch Ness monster by some surgeons) two years after implantation. At the time of initial implantation this patient had a tympanic membrane perforation (secondary to middle ear mucosal disease) which was closed with temporalis fascia graft. He underwent revision surgery and reimplantation without any detriment to speech recognition scores. The subsequent drum perforation was repaired with temporalis fascia.

Implants types and performance

At the time of conclusion of the study both the Clarion devices were fully functional. Of the 113 Nucleus devices, 104 had more than 15 electrodes switched on, 8 had between 10 and 15 electrodes switched on and one had less than 10 electrodes working. Of the 115 cochlear implants used during this period (111 first implants and 4 reimplantations), 113 were Nucleus 24 (Cochlear Corporation) and two were Clarion (Advanced Bionics, USA). (Table 4). Full insertion (i.e., all active rings and the supporting or stiffening rings inside the cochlea) was possible in 107 Nucleus devices and on both the Clarion devices used (94.8%). Amongst the partial insertions 3 (2.6%) had more than fifteen electrode rings within the cochlea while 3 (2.6%) had less than fifteen rings within the cochlea. Great difficulty in insertion was encountered in 3 (2.6%), slight difficulty in 8 (6.9%) and no difficulty in 104 (91.2%). Hylaruronidase (Healon®) was used in all implantation episodes to ease insertion and minimise trauma to basilar membrane.

Table 4. Electrode insertion:

Ease of insertion:			
• No difficulty:	104 (90.5%)		
• Slight difficulty:	8 (6.9%)		
• Great difficulty:	3 (2.6%)		
Depth of insertion:			
Implant type	Depth of insertion	Number	Ease of insertion
CLARION (n=2)	Full	2	No difficulty
Nucleus 24 (n=113)	Full	107 (93%)	• No difficulty (101) • Slight difficulty (5) • Great difficulty (1)
	20	1(0.8%)	No difficulty
	18	1(0.8%)	Slight difficulty
	16	1(0.8%)	Slight difficulty
	13	1(0.8%)	Great difficulty
	12	1(0.8%)	Slight difficulty
	8	1(0.8%)	Great difficulty
Electrodes switched off:			
>15 electrodes working		104 (92.1%)	
10 – 15 electrodes working		8 (7.1%)	
<10 electrodes working		1 (0.8%)	

Table 5. Imaging

Pre-operative Scans (CT and MRI)::	
• Possible fibrous occlusion:	2
• Partial ossification:	3
• Severe ossification:	1 (this needed a drillout)
• Patent cochlea:	94
	Total 100 patients
Post-operative scans x rays:	
• Normal appearance:	85
• Partial insertion:	4
• Abnormal position:	1
• Not known:	27
	Total 117 implantation episodes

Preoperative imaging

Pre operative imaging (computed tomography and magnetic resonance imaging) was very useful in predicting patency of cochlear lumen and this was corroborated by the surgical findings. (Table 5).

DISCUSSION

The initial evaluation of the safety of cochlear implantation in the UK was undertaken by the Medical Research Council in 1994[4]. Overall complication rates were low and around 34%.

The rates of major complications reported following cochlear implantation range between 8%[5] and 10% [4,3.] The major complication rate in our first 100 consecutive patients was 3%. This has remained steady with the current at 3.2% for the second hundred patients reported here.

The complication rates have an inverse relationship with the position of the patient in the case series[5] i.e. complication rates drop with increasing experience of the surgical practice. This is reflected by the halving of minor complications rate from 39% in the first[1] cohort to18% in the second.

Wound and flap problems have been one of the commonest complication experienced in many series[10,3]. Flap problems were commoner with the flap designs which utilise large post aural incisions and wide scalp mobilisation. Series with smaller incisions report lesser wound and flap problems[11,12]. After starting with a grossly extended endaural incision (Figure 1A) we have modified our incision to a smaller “lazy S” post aural incision (Figure 1B) with minimal scalp mobilisation and the wound is closed in two layers with a periosteal flap to reinforce this. The wound heals better and without any compromise of the flap vascularity or viability. The wound infection rate now stands at 9% compared to 11% in the first hundred patients.

In 2 patients (1.6%) there was late flap breakdown. This failed to settle down with a conservative approach and an explantation / reimplantation procedure had become necessary. Both patients were elderly males.

Although the device design has improved internal device failure still continues to account for many explantations[3]. Failure rates of 3% for adults have been quoted for the Nucleus device[4]. In this series the device failure rate was 0.8%.

In our series explantation rates were around 4% and is comparable to that reported from other centres. Overall explantation reimplantation rates of between 8 and 10% have been reported[5,6,7] and the commonest causes of explantation were device failure.and flap necrosis. Histological studies[14] have shown that the trauma from explantation and reimplantation is no greater than the implantation itself.

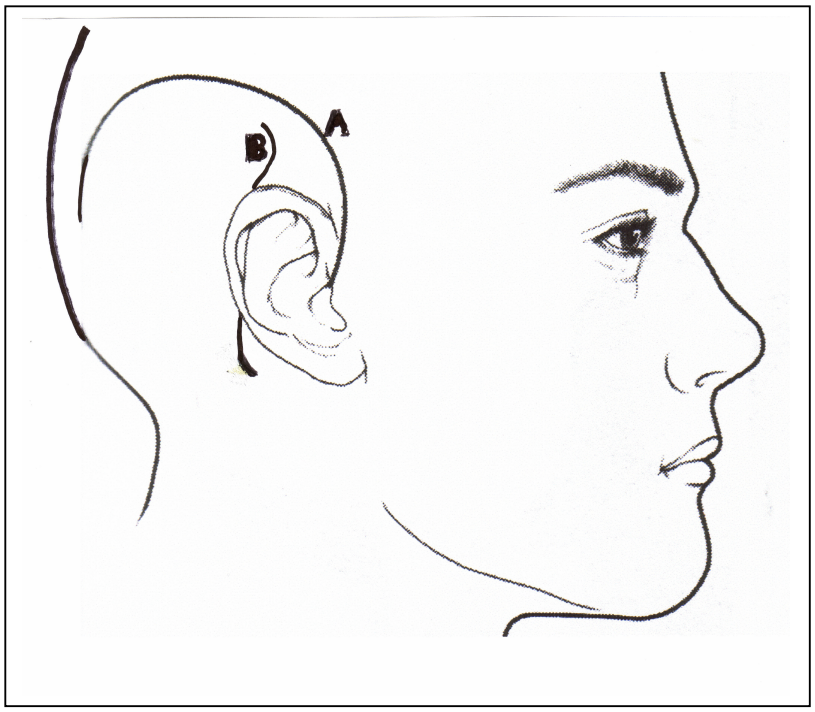

Figure 1. Incisions used (A) Extended endaural incisions used in the initial stages. (B) Small “lazy S” postaural incision used in later stages.

There is therefore no surgical contraindication for ipsilateral reimplantation for device failure. In most series the majority of insertion depths and the number of active channels remained unaltered after reimplantation. Over the years we have tried to use only one commercial variety of cochlear implants so the results can be monitored consistently. As in our series retrospective multicentre studies of reimplantation surgery[10,13] for failed Nucleus 22 cochlear implants in US implant programmes found no relationship between the performance of the replacement device and the duration of original device use, surgical complications, insertion depths and preoperative variables.
Minor infections were easily controlled by intensive antibiotic treatment as has been reported by some[9, 11]

In the first hundred cochlear implantations, three patients had transient facial nerve weakness with full recovery. Experience and the routine use of facial nerve monitoring have prevented any facial nerve complications. Like many other centres[10] we have used per-operative neural response telemetry. This includes device integrity testing and has helped to detect faulty devices and provides predictive information about the threshold and comfort levels and the afferent neural pathways leading to the brainstem.

CONCLUSIONS

Device failure, wound and flap problems are still the commonest complications. Smaller incisions and minimal scalp mobilisation certainly helps to reduce wound and flap problems. Many minor complications can be dealt with conservatively without recourse to explantation. Many without complications have gone on to bilateral implantation.

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2.3

SURGICAL REMOVAL OF 11 COCHLEAR IMPLANTS- LESSONS FROM THE 11 YEAR OLD CAMBRIDGE PROGRAMME

J. Ray
R.F. Gray
I. Court

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ABSTRACT

This is a retrospective study of 10 patients (11 ears) out of 132 cochlear implant patients of the Cambridge Cochlear Implant Programme. These patients have all been explanted. Individual problems have been studied, relevant literature reviewed and the pitfalls of implant surgery re-examined in the light of our experience.

INTRODUCTION

Cochlear implantation (CI) has now become an established means to gain access to sound in profound deafness far beyond the reach of conventional hearing aids[1]. Although the surgical procedure involved has remained essentially the same, experience and expertise has matured with time .The equipment has become more complicated as improvements have taken place. Some of the earlier implants had failed and needed replacing and some needed upgrading to take advantage of the ever improving results with newer devices.

The Cambridge Cochlear Implant Programme started in 1986 and in over 11 years, 90 adults and 42 children have been implanted (Table 1). In this review article we have looked at our data on implanted patients and report on 11 cases (in 10 patients) where explantation was necessary (Table 2). Six patients with multichannel implants had significant complications requiring explantation and a further four with single channel implants were upgraded to newer devices where performance could be improved.

Table 1. Explant Details

Cambridge Cochlear Implant Programme (1986-1997)
• Total - 132
• Adults -90 (Multi Channel - 84; Single Channel - 6)
• Child -42 (Multi Channel - 41; Single Channel - 1)
Total Implants Explanted - 11(8.3%)
• *Group A)* Explantation for problems -7 (5.3%)
• *Group B)* Explantation for upgrading to better devices -4 (3%)
Adult-11(8.3%) M=7 (5.3%)
Child - 2 (1.5%) F =6 (4.5%)
Type of implants explanted
RNID/UCH 4(3%)
Ineraid 5(3.7%)
Nucleus 2(1.5%)
Awaiting explantation for upgrading - 2 [RNID/UCH] (1.5%)

Table 2. Reasons for Explantation

• Trauma	4 (3.0%)
• Flap Problems	1 (0.7%)
• Upgrades	4 (3.0%)
• IAM Pathology	1 (0.7%)
• Psychiatric Problems	1 (0.7%)

PATIENTS AND METHODS

Case details of patients implanted in the Cambridge Cochlear Implant Programme between 1986 and 1997 were examined to find those who have needed explantation surgery following CI. In all 11 explantations in 10 patients (one twice) are reported (Table 3).

Table 3. Details of 10 patients (one twice)

Pt.	Sex	Group	Aetiology	Implant	Problem	Cause	Interim action	Current Position
Group A: Explantation for problems								
LF	M	Child	Meningitis	N22	Electrical failure	Direct Trauma	Explanted	Implant N22 (ipsi)
SE	F	Child	Unknown	N22	No response/ Nil EABR	IAM pathol	Explanted/ MRI	Vibrotactile aid
CH	M	Adult	Unknown	Ineraid	Psychiatric problems	CSOM/ Tinnitus (contra)	Explanted	_____ _
CT	M	Adult	CSOM	Ineraid	#Pedestal	Direct traums	Explanted	Implant N24 (contra)
CL	M	Adult	Meningitis	Ineraid	#Pedestal	Direct trauma	Explanted	Implant Ineraid (ipsi)
RS	M	Adult	Head Injury	Ineraid(1)	#Pedestal	Direct trauma	Explanted	Awaiting N24 (contra)
				Ineraid(2)	Discharge at pedestal	Flap infection		
Group B: Explantation to upgrade from single to multichannel								
JH	M	Adult	Progressive SN	RNID/ UCH	Limited perception	Upgrade	Explanted	Implant Ineraid (contra)
JW	F	Adult	Meningitis	RNID/ UCH	Limited perception	Upgrade	Explanted	Implant N22 (ipsi)
WW	M	Adult	Progressive SN	RNID/ UCH	Limited perception	Upgrade	Explanted	Implant N22 (ipsi)
IB	F	Adult	Progressive SN	RNID/ UCH	Limited perception	Upgrade	Explanted	Implant N22 (contra)

SN= sensorineural deafness; ipsi=ipsilateral; contra=contralateral; N22/24=Nucleus

Pre-operative assessment (Adults):

Pre-operative assessment included a detailed history and clinical examination after initial referral. The patients were also given a questionnaire prior to this. Pure tone audiometry (AC and BC) and aided free field audiometry were performed followed by speech audiometry using live voice BKB sentences and Boothroyd word lists. A trial of high powered hearing aids with good fitting ear moulds was given to rule out any useful benefit from conventional aiding. High resolution CT Scans of the cochleas and internal auditory meati (IAM) were used to assess the patency of the cochlear duct and to rule out any congenital anomaly. Magnetic Resonance Imaging (MRI) has been substituted for CT in suspected congenital anomalies in the IAMs and the cochleas. All single channel implantees with the RNID/UCH device were fitted with a temporary round window electrode through a tympanotomy a few weeks prior to implantation as part of the assessment.

Pre-operative assessment (Paediatric):

Paediatric implantees had Visual Reinforcement Audiometry and speech and language assessment plus trial of hearing aids. CT Scanning or MRI (under general anaesthetic if necessary) was undertaken in all cases. Stapedial reflex integrity tests were carried out in the operating theatres immediately post insertion while the child was still under the anaesthetic.

Post-operative Tests (Adults) :

Post implantation electrode position was confirmed by perorbital skull x-rays next day. After switch on the audiological test battery includes Aided Free Field Audiograms. Speech perception was tested using both live voice (BKB sentences, Boothroyd word lists and connected discourse tracking) and taped lists (BKB sentences, vowel-consonant-vowel setup, gap detection). Awareness of familiar environmental sounds was also tested from taped lists.

Post-operative Tests (Paediatrics):

Post implantation electrode position was confirmed as in adults by perorbital skull x rays. After switch on audiological tests for awareness of sound and speech were carried out using sound detection, speech discrimination, pattern recognition, Ling 5 sound tests, Manchester picture test and modified open set breakfast picture scores.

Study Group:

Our study group of 10 patients comprised essentially of two sub groups: Group (A) consisting of patients who had needed explantation for some complication related to implantation and Group (B) consisting of patients who have needed revision surgery because of unsatisfactory performance of a single channel device or for upgrading to multichannel devices (Table 1).

RESULTS

Group A: Explantations for problems

In this group a male: female ratio of 5:1 was noted. Of these 4 were adults and 2 were children. Two cases had been implanted for meningitis, one following head injury, one following recurrent ear infections and one following congenital deafness, while in the last one the aetiology was unknown.

Adults(n=4)

Hardware failure due to trauma (n=3); Flap problems (n=1) More complications occurred with the Smith & Nephew Richards Ineraid devices (formerly Symbion) and the commonest reason for reimplantation was trauma leading to breakage of the pedestal base (three out of the five devices involved). One of them (R.S.) was successfully reimplanted into the same ear after withdrawing the original electrode through an enlarged cochleostomy. However on close scrutiny four of the six ball electrodes were missing from the lead cable on the old device, presumably detached and left inside the cochlea. Audiological results were poor. Later flap problems due to overwhelming skin growth despite flap debridement under anaesthesia and recurrent infection and discharge from pedestal prompted its subsequent explantation. This patient is currently being assessed for reimplantation into the other ear.

From this experience electrode withdrawal has not been attempted in any further Ineraid patients. In the remaining three patients the wires were cut and left in situ while the pedestal was removed and reimplantation carried out in the contralateral cochlea in two cases.

Another patient (C.T.) with the traumatic breakage unfortunately had a contra-lateral discharging mastoid cavity. This was successfully prepared using the technique of cavity obliteration with autologous abdominal fat and blind pit closure of the external auditory canal[2]. He has now been implanted with a Nucleus 24 device in the prepared ear.

Psychiatric problems (n=1) One patient (C.H.) who had a history of psychiatric problems found it difficult to cope with the implant, because of discharge from the mastoid cavity and felt that the tinnitus in the ear had been worsened by implantation. Surprisingly both tinnitus and discharge were in the contralateral ear. This implant, although functioning well and with satisfactory speech discrimination and audiological results (38 per cent BKB with implant alone and 60 percent with implant and lip reading (Table 4), had to removed in response to the patient's wishes.

Table 4. Audiological performance pre explantation and post reimplantation

Patient	Pre explantation BKB sentences (implant + lip reading)	Post reimplantation BKB sentences (implant + lip reading)
LF	Limited language	Limited language
SE	Limited language	Limited language
CH	60%	Not reimplanted
CT	100%	100%
CL	100%	100%
RS	74%	70%
JH	90%	100%
JW	80%	96%
WW	8%	52%
IB	94%	6% (electrode movement)

Children (n=2)

The two cases in this group were children who had received the multichannel Nucleus 22 device.

Hardware failure due to trauma (n=1). The first complication was an electrical failure in the circuitry following a direct trauma to the implant as a result of a fall from his bed (L.F.). The Nucleus device was very easily removed intact and reimplantation through the same cochleostomy into the still patent cochlear duct was straightforward. The results have been as good with the replacement device as with the original device.

Unanticipated 8th. Nerve pathology (n=1). One of the most intriguing problems encountered in our experience was that of nonstimulation in a 5 year old congenitally deaf child (S.E.) born at term through normal delivery and without any

family history of deafness. After assessment and CT Scanning the right ear was chosen to implant the Nucleus 22 device because of the risk of otorrhoea from a troublesome grommet in the other ear. The surgical procedure and post operative period were unremarkable and a good insertion of the electrode array was seen on perorbital plain films. However the perioperative EABR showed poor waveforms and subsequently there was a complete failure to stimulate the auditory pathway at multiple failed tuning sessions. Integrity testing showed all parts of the Nucleus 22 device to be working normally. A few inconsistent responses were obtained but most were suspected to be due to non auditory stimulation of the ipsilateral facial nerve which was producing ipsilateral facial twitches on exposure to noise. Several electrodes were switched off and remapping was tried several times (including twice by Nucleus UK). EABR again showed failure to stimulate any part of the auditory pathway. The only clue was now obtained from a repeat of ultra high definition CT scan. Although this showed perfect positioning of the implant and the electrode array, a closer look revealed a narrowing of the Internal Auditory Meatus (IAM) on the implanted side with the absence of the bony protrusion in the IAM known as Bill's bar. This was taken as an indirect pointer to IAM pathology[3] and a detailed MRI scan of the cerebellopontine angle (CPA) and IAMs in 3 planes was planned. As the device was not MRI compatible (due to the absence of a removable magnet in the receiver/stimulator) it had to be explanted 15 months after implantation. The MRI revealed abnormalities of structures in both IAMs and CPAs but more pronounced on the implanted side. These constituted an absent or a rudimentary eighth nerve along with a single facial nerve trunk occupying the IAM and accounted for the consistent failure to respond and also for the non auditory responses. Contralateral reimplantation was considered but thought to offer no certainty of hearing responses and therefore not carried out. A vibrotactile aid has now been provided to augment auditory and speech training.

Group B: Explantations for improving performance with multichannel device

This group comprised of adult patients who had received the original RNID/UCH Single Channel implant with round window ball electrode. The predominant aetiology was idiopathic progressive sensorineural hearing loss while one was deaf due to meningitis. The male: female ratio was equal (Table 3).

Device failure (n=1)

One of our initial patients with a RNID/UCH device (I.B.) had problems due to device failure and this had to be explanted and replaced with another RNID/UCH device on the ipsilateral side. Subsequently this has been upgraded with a

Nucleus 22 multichannel device in the contralateral ear. Unfortunately after 3 years of trouble free performance electrode movement has now been suspected and electrodes have required extensive remapping. Should further changes occur then reinsertion would seem to be an appropriate course of action.

Poor performance (n=3)

Four of our patients with RNID/UCH devices have now been upgraded to Multichannel devices to improve performance. One of these patients has received the Ineraid multichannel device in the contralateral ear. The other two patients have been reimplanted with Nucleus 22 multichannel device into the same ear after removal of the original RNID/UCH device under the same anaesthetic. Surgical procedures have been uneventful and straightforward and results have been satisfactory. Gain in scores for BKB sentences were as follows:

Patient	BKB(%) single channel (Lip reading + Implant)	BKB(%) multichannel (Lip reading + Implant)
J.H.	90	100
J.W.	80	96
W.W.	8	52

N.B.:Awaiting explantation (n=2)

Two patients are awaiting explanation of single channel devices which have remained unused after receiving the upgraded devices in the contralateral ear.

DISCUSSION

Cochlear implantation, like all surgical procedures carries with it a range of complications both major (which necessitates return to the operating theatre and revision surgery under general anaesthesia) and minor (those which can be sorted out in clinic)[4].

Complications

Most common major problem:

In our series the commonest problem was related to the breakage of the Ineraid percutaneous pedestal due to direct trauma (2.2 per cent). This has been unavoidable because the biocompatible pyrolised carbon pedestal is brittle and it is not always possible to repair it in situ using adhesives.

Most serious major problem:
Flap problems with overwhelming skin growth and discharge from the pedestal site due to recurrent infection (0.7 per cent) has forced us to explant an Ineraid device which was working satisfactorily. This experience has taught us to modify our flap design and to thin it down by removing subcutaneous tissue. This has averted any further flap overgrowth. Infection and discharge continue to be problems with the percutaneous pedestals; meticulous care, hygiene and follow up seems to make no difference.

Most avoidable major problem:
The problem we have encountered with non stimulation of auditory pathway (0.7 per cent) due to absence of the eighth nerve is now avoidable with high resolution, sub-millimetric fast spin-echo T2-weighted MRI scans with three-dimensional reconstruction.

Most unavoidable major problem:
By far the most unavoidable problem has been the need to replace older devices with newer ones (three per cent) in order to improve performance and improve quality of life. At the current pace of research and development in integrated circuits it is not difficult to foresee the fact that the present devices might need replacing in future.

What can be done in future?
Manufacturers
Osseointegration of a Percutaneous sytem presently offers the best answer to the brittleness of the original Ineraid pedestals which led to fractures and total loss of device[5]. The initial results of the trial with the University College London Implantable Device [UCLID][6] have been very promising. Percutaneous systems may be upgraded to new speech processing strategies without operative surgery (for example Med El CIS link attached to Ineraid pedestals).

Design modifications in the Nucleus system have also helped to overcome several problems highlighted in our series. Stress relief loops and extended silastic covering leading on to the lead cable promises to solve disruption of circuitry due to direct trauma as was seen in one of our paediatric patients[7].
The excellent detail being shown by MRI scans by experienced radiologists means this will be used more often[8] for potential CI candidates. MRI for patients who have already been implanted has been looked into carefully and the alternative to

complete explantation purely to do an MRI lies in the use of newer devices with a removable magnet or in the use of osseointegrated titanium percutaneous pedestals which are non magnetic and therefore MRI compatible and have a detachable circuitry.

Cochlear Implant Teams

The need for strict patient selection criteria and also use of high resolution imaging techniques especially in cases of congenital hearing loss cannot be overemphasised. Lack of experience in patient selection has resulted in two explantations in this series (psychosis and eighth nerve agenesis). The latter has prompted us to request sub-millimetric high-resolution fast spin-echo T2-weighted MR images with three-dimensional reconstruction in such patients and possibly for all patients in future.

Flap design plays a very important role in both the percutaneous and transcutaneous systems, more so in the former. In these the flap thickness should be kept down to a minimum in order to avoid overgrowth.

Peri-operative Averaged Electrode Voltages (AEVs) and Electrically Evoked Auditory Brainstem Responses (EABRs) provides a good assessment of the internal receiver at the time of initial surgery. However having already embarked on the procedure this will not prevent inappropriate implantation.

As this is such a rapidly advancing field where computer science, electronics and otology go hand in hand it is quite likely that what seems like state of the art technology today might soon be antiquated. Therefore the possibility of needing to upgrade present devices to take fullest advantage of any improvements should always be kept in mind when designing any modification in the surgical procedure, in particular, taking care that at least the device is removable when needed without irrevocable damage to the middle ear or cochlea. This has happily been our experience with four single channel implants who have opted for upgrading to multichannel devices.

Patients

The attitude of the patient is critical if explants are to be avoided. Motivation must be good and expectations must be realistic. Patients with psychosis or history of psychiatric must be regarded with circumspection.

Contact sports and physical activities should be tailored to avoid direct trauma and device breakages.

Possible future explantations

Electrode migration and extrusion is still a vexing problem. Use of cement to anchor the electrode array in the cavity in addition to dacron ties has kept our number of migrating electrodes down. However in the unfortunate eventuality of that happening a wait and watch policy is adopted along with careful remapping when required.

CONCLUSION

Explantations for various reasons still remain an unpleasant reality in a small proportion of any prosthetic surgery. Cochlear implants are no exception in this regard. In our series the leading causes of explantation were: upgrading to improve performance (3.0 per cent); hardware failure due to direct trauma (3.0 per cent); psychiatric problems (0.7 per cent); flap problems (0.7 per cent); IAM pathology (0.7 per cent).

Although cochlear implantation has now come of age it still continues to be a rather difficult and exacting procedure needing considerable skill and experience in elements of otology and neurotology[9]. Past problems and failures have highlighted the pitfalls associated with the procedure. To avoid these problems attention should be given to: patient assessment strategy; flap design and handling; choice and siting of the receiver/stimulator package and care in electrode placement and anchorage. Together, they will determine the outcome of future procedures.

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2.4

EXPLANTATION AND REIMPLANTATION OF COCHLEAR IMPLANTS

J. Ray
D. Proops
I. Donaldson
C. Fielden
H. Cooper

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ABSTRACT

This study looks at the rates of explantation and reimplantation surgery in adult cochlear implant patients between 1990 and 2002 and also evaluates the surgical and audiological implications.

15 (5.5%) out of 272 adult cochlear implantees (288 cochlear implants: 282 Nucleus, 4 MedEl Combi 40, 2 Clarion) needed their devices removed (explanted). 14 out of the 15 patients selected received explantation, reimplantation or revision surgery. The main reasons included device failure (2.2%), wound and flap problems (1.8%) and electrode extrusion (0.73%). Wound and flap problems were more common with larger skin incisions. In staged reimplantations reinsertion was made easier if the electrode was retained in situ till reimplantation. Performance of the replacement device was not related to the aetiology of deafness or to the cause of explantation. This data will be useful in counselling patients.

INTRODUCTION

Cochlear implantation has established itself as a reliable way to rehabilitate selected profoundly deaf patients. Like any device they are prone to malfunction and may breakdown. Similarly complications of the surgical process itself may compromise the functioning of the device requiring it to be removed. Such an event has serious financial and psychological implications for both the patient and the implant team and is reported far less commonly than the successes of implantation. We have looked back at our data on 272 consecutive adult cochlear implant patients' notes to analyse the rates of explantation and reimplantation between 1990-2002. This data is useful for preoperative counselling and avoiding future complications.

MATERIALS AND METHODS

This is a retrospective study looking at all the implantations, explantations and reimplantations undertaken at the Midlands Adult Cochlear Implant Programme between 1990 and 2002. Case notes were analysed and the relevant audiological data reviewed in the selected patients.

272 adult patients received cochlear implantation (286 cochlear implants: 282 Nucleus, 4 MedEl Combi 40, 2 Clarion) in our programme between 1990 and 2002. Of these 272 adult patients fifteen (5.5%) needed explantation, reimplantation and/or revision surgery. These patients were further analysed regarding their aetiology of deafness, performance, reason for explantation and subsequent reimplantation.

Performance was assessed by measuring speech perception with Bench, Kowal, Bramford (BKB) sentences. The tests were administered in the auditory alone setting with pre-recorded sentence lists using a male voice in quiet environment. Patients were tested at 3 months, 6 months and 9 months after implantation and annually thereafter.

During the initial stages of the programme the surgical approach involved a grossly extended endaural incision (Figure 1A) with wide mobilisation of scalp tissue to have access to the site of the receiver stimulator package. Subsequent surgical steps involved cortical mastoidectomy, posterior tympanotomy, cochleo-stomy and electrode insertion. The endaural incision was later replaced with a much smaller “lazy S” post auricular incision (Figure 1B) in 1998.

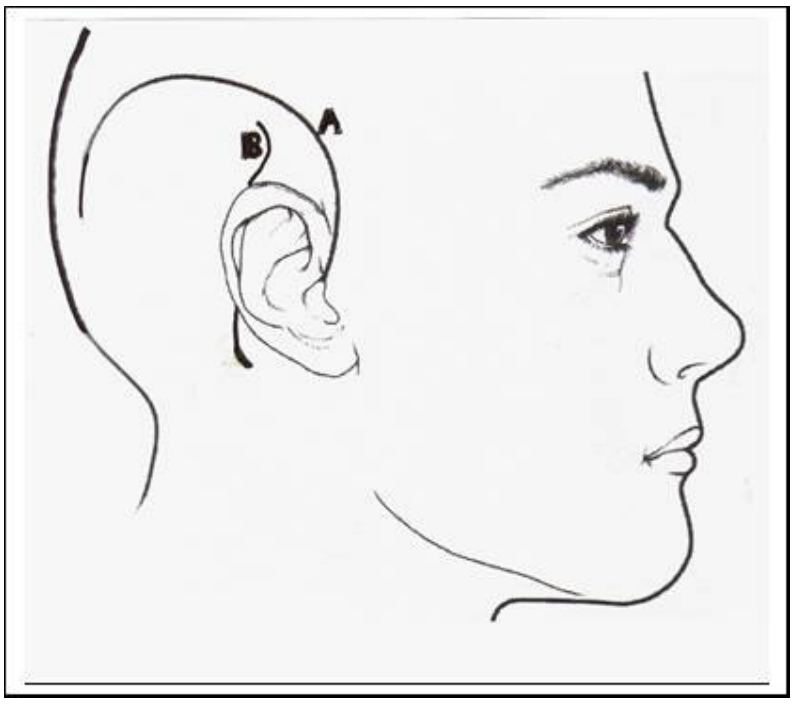

Figure 1. Incisions used. (A) Extended endaural incision used in the earlier stages. (B) Small “lasy S” postaural incision used in the later stages.

The wound was closed in two layers and per operatively antibiotics were administered. Neural response telemetry was performed routinely over the last three years. A post operative radiograph was taken.

RESULTS

Fifteen patients (5.5%) out of a total of 272 adults implanted between 1990 and 2002 underwent explantation, reimplantation and/or revision procedures. Details of aetiology, duration of deafness, implant types and speech perception scores (BKB sentences) with each implant are shown in Table 1. Eighteen implants (twice in three patients each; P9, 10, 11) were removed (explanted) for various reasons. One patient could not be reimplanted because of progressively worsening multiple sclerosis and depression (P15). The electrode was repositioned and reinserted without explantation in one patient. The remaining thirteen patients were reimplanted either ipsilaterally or contralaterally. The mean age at initial surgery was 55.2years (range 38 – 77 years). The mean duration of bilateral profound hearing loss at the time of first implantation was 12.86 years (range 1 – 30 years). Time to explantation surgery ranged from 12 months to 9 years (mean 2.7 years) and follow up after reimplantation ranged from 14 months to 6 years (mean 3.9 years).

The aetiology of deafness included chronic suppurative otitis media in one, measles in one, otosclerosis in two and meningitis in one. The cause was unknown in eight. In this group the deafness was of sudden onset in two and progressive in six. In one patient the cause was a combination of measles in infancy and subsequent chronic suppurative otitis media and in another it was a combination of head injury and subsequent chronic suppurative otitis media (Table 2).

Table 2: Aetiology of deafness

CSOM	1 (0.4%)
Measles	1 (0.4%)
Otosclerosis	2 (0.73%)
Meningitis	1 (0.4%)
Measles + CSOM	1 (0.4%)
Head injury + CSOM	1 (0.4%)
Idiopathic	8 (2.94%) (sudden 2; progressive 6)

Table 1. Details of patients (Pts), age at first implantation, aetiology, duration of profound deafness in years, implant types, performance (BKB scores with 1st, 2nd, 3rd implant) and relevant problem. R=right side, L=left side. N22/N24=Nucleus device. NAS=non auditory stimulation. MS=multiple sclerosis. Explant interval refers to the gap between explantation and reimplantation

Pts	Age (Yr)	Aetiology	Durn Deaf (Yr)	1st Imp	2nd Imp	3rd Imp	Explant interval (Yr)	1st BKB Score	2nd BKB Score	3rd BKB Score	Problems
P1	40	Sudden	1	MedEl L	N24 L		3	84%	93%		*Device failure*
P2	59	CSOM + Measles	28	MedEl R	N22 R		1	50%	82%		*Device failure*
P3	70	Otosclerosis	10	N22 L	N24 L		2	48%	12%		*Device failure*
P4	65	Otosclerosis	30	N24 L	N24 L		4	48%	58%		*Device failure*, Partial insertion, *NAS*,
P5	55	Head Injury + CSOM	2	N22 L	*Reinserted*		2	30%	27%		Electrode extruded;
P6	60	Meningitis	1	N24 R	N24 L		1	0%	73%		No response, device working
P7	40	Progressive	5	N24 L	N24R		2	0%	86%		No response, electrode curled before insertion,
P8	56	Progressive	20	N22 R	N24 L		9	48%	78%		Electrode extruded. RNID(L)single channel explanted, Reimplanted(L)
P9	40	Progressive	4	N22 L	N24 L	N24R	4 + 3	0%	0%	86%	*Device failure*, Contour tip flip back, *2nd Device failure*
P10	36	Sudden	2	N22 R	N24 L	N24R	2 + 6	60%	20%	12%	Cholesteatoma (R); Revised R then explanted R Electrode cut; Implant L (poor result), Reimplant R (poor result)
P11	50	Progressive	10	N24 R	N24R	N24L	3 + 3	86%	8%	38%	*Wound inf. Flap problem*, *revised, reinserted*, explanted, Reimplant R,
P12	70	Progressive	10	N24 R	N24 L		2	55%	65%		*Flap problem*
P13	70	Progressive	30	N24 L	N24 R		2	0%	0%		*Wound breakdown*,
P14	77	CSOM	30	N22 R	N24 R		2	0%	0%		*Wound breakdown* , infected cavity, Flap revised, electrode cut,
P15	38	Measles	10	N24 R	N/A		1	20%	N/A		*Wound breakdown* , MS, Depression, Not reimplanted, HA

Reimplantation into the ipsilateral ear was carried on eight occasions. The reasons were device failure (5), wound and flap problems (2) and cholesteatoma formation (1) requiring staged reimplantation. The cholesteatoma was dealt with at the time of the explantation and the device was removed after dividing the electrode array close to the cochleostomy. Reimplantation was staged over six months. At this time the old electrode array was removed and the new one inserted in the same sitting. In all these patients the depth of insertion remained unchanged. Post reimplantation speech perception scores (BKB sentences) improved in three, worsened in two (otosclerosis and cholesteatoma) and remained poor in two (wound infection and flap problems) (Table 1 and Table 3).

Table 3. Details of procedures

Ipsilateral	8 (2.94%)
Contralateral	8 (2.94%)
Revised	2 (0.73%)
Not reimplanted	1 (0.4%)

Contralateral reimplantation was carried out on eight occasions. All of these patients had suffered bilateral profound hearing loss ranging between 1 and 30 years. The reasons for contralateral reimplantation included infection (1), flap problems (1), second consecutive device failure (1) and device upgrade (1), poor response with working implant (3) and wound breakdown (1). Post reimplantation speech perception scores (BKB sentences) improved remarkably in five, worsened in two and remained poor in one (Table 1 and Table 3).

The two main reason for explantation were device failure (2.2%) and wound and flap related problems (1.8%). The other reasons included electrode extrusion (0.73%), cholesteatoma formation (0.4%) and device upgrade (0.4%) (Table 4).

Table 4. Reasons for explantation

Patient factors	
Wound breakdown	3 (1.1%)
Flap problems	2 (0.73%)
Electrode extrusion	2 (0.73%)
Cholesteatoma	1 (0.4%)
Device related	
Upgrade	1 (0.4%)
Device failure	6 (2.2%)

Of the 288 cochlear implant devices used in 272 patients 282 were Nucleus 22 , 24 and contour (Cochlear Ltd, Australia), four were Combi40 (MedEl Corp, Austria) and two were Clarion (Advanced Bionics, USA) devices. Two of the four MedEl devices (50%) suffered unexplained and unexpected hard failure after working well between 3 and 5 years. Four out of the 282 Nucleus devices (1.42%) failed necessitating explantation and reimplantation (Table 5).

Table 5. Failed Devices

Devices	Total used	Failed	%
MedEl Combi 40	4	2	50
Nucleus 22 & 24	282	4	1.42
Clarion	2	0	0

Wound breakdown was encountered in three cases. All were from the earlier operations where an extended endaural incision (Figure 1A) with a large flap was used. Intensive antibiotic treatment, wound debridement and flap rotation failed to save the implants from being compromised making explantation inevitable. Another problem that was difficult to manage was persistent flap swelling necessitating contralateral reimplantation.

Electrode extrusion occurred in two patients. Revision surgery was carried out without explantation in one with noticeable worsening of speech perception (P5). In the other patient (P8) contralateral reimplantation was carried out after removing the old RNID single channel device and there was significant improvement in speech perception (BKB sentences).

At the initial surgery full electrode insertion had been achieved in 14 patients including those with otosclerosis and meningitis. Partial insertion was achieved in one patient with otosclerosis (P4). He also experienced significant non auditory stimulation in the form of throat sensation. Device failure necessitated ipsilateral explantation and reimplantation with improved speech perception scores (BKB sentences). The most notable problem was involving a Nucleus 24 contour device (P9). The tip of this device seemed to have folded back on itself at the time of withdrawal of the stylus (as evidenced by subsequent x rays) leading to difficulty in obtaining a suitable map. This necessitated explantation and reimplantation. Unfortunately the second device also failed and subsequently reimplantation was carried out successfully in the contralateral ear. Interestingly this problem of

excessive curving of the contour electrode was experienced in yet another device which curled up excessively even before insertion into the cochlea.

Three patients had an overall deterioration of their speech perception scores as a result of explantation and reimplantation surgery (P3, 10, 11). Explantations were due to wound infection, cholesteatoma and device failure in an otosclerotic cochlea. The patients with persistent poor performance were above seventy years of age (P13, 14). However there was no definite relationship between the performance of the replacement device and the aetiology of deafness or that of the explantation or the duration of prior implant use.

DISCUSSION

Over the last two decades cochlear implantation surgery has evolved through various techniques and devices. However there are occasions where the implant needs to be removed and replaced. Despite the increasing experience that has come with the growing number of implantees over the last decade, data on failures and explantations have been few.

The initial evaluation of the safety of cochlear implantation in the UK was undertaken by the Medical Research Council in 1994[1]. Overall complication rates were very low although patient numbers were small. Reimplantation was first reported by Hochmair-Desoyer and Burian[2]. Device failure continues to account for majority of the explantations[3]. Failure rates of 3% for adults have been quoted for the Nucleus device[4]. In our series the overall device failure rate was 2.2% (1.4% for Nucleus device).

Myamoto et al have reported an overall reimplantation rate of 10% and stated that for the majority the insertion depth and the number of active channels remained unaltered[5]. Ray et al reported explantation rates of around 8.3% where the commonest cause of explantation was device failure[6]. In our series explantation rates were around 3.7%. This is comparable to similar series reported from other centres[7]. Over the last three years we have used only one commercial variety of cochlear implant so that results can be monitored consistently. As in our series a retrospective multicentre study of reimplantation surgery[8] for twenty-eight failed Nucleus 22 cochlear implants in eighteen US implant programmes found no relationship between the performance of the replacement device and the duration

of original device use, surgical complications, insertion depths and preoperative variables.

The second important problem encountered was wound infection, flap swelling and flap breakdown. Flap problems were commoner with the previous flap design which utilised the grossly extended endaural incision. Minor infections were easily controlled by intensive antibiotic treatment as has been reported by some[9] but 1.83% failed to settle down with a conservative approach and an explantation / reimplantation procedure had become necessary. No flap breakdowns have been recorded after change to a smaller “lazy S” incision with lesser mobilisation of the scalp tissues. This experience is also echoed in other series[7,10].
In case of cholesteatoma, granulations and middle ear adhesions the best option is to cut the electrode array close to the cochleostomy and remove the main body of the device. This facilitates exploration of the middle ear and also maintains a pathway within the cochlea till reimplantation is undertaken[7].
Like many other centres[11] we use of per operative neural response telemetry. This includes device integrity testing and has helped to detect faulty devices. A back up device is always available for such eventuality.

CONCLUSIONS

Several key features about cochlear reimplantation surgery have been evaluated in this study and this data has been useful in counselling patients.
Device failure, wound and flap problems are the commonest causes for explantation, reimplantation or revision. Larger skin incisions and wider scalp mobilisation are associated with wound and flap problems. In staged procedures retaining the electrode array in the cochlea facilitates subsequent reinsertion. Performance of the replacement device is not related to the aetiology of deafness or to the cause of explantation.

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2.5

FURTHER EXPERIENCE WITH FAT GRAFT OBLITERATION OF MASTOID CAVITIES FOR COCHLEAR IMPLANTATION

R.F. Gray

J. Ray

D.J. McFerran

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ABSTRACT

Obliteration of old mastoids and wet middle ears with autologous abdominal fat seems to be a reliable technique to render chronically discharging mastoid cavities or open middle ears dry and closed. There have been two other papers on this topic from the same department[1,2]. This paper is the third in the series and looks at the intermediate results of this procedure at five years. Of the 16 patients (one bilateral) 94.1 per cent of the ears are dry and uninfected with closed external meati. Recurrent cholesteatoma was found in two patients at implantation and removed.

INTRODUCTION

Chronically discharging middle ears pose a challenge to otologists. There is little wonder why so many attempts have been made to overcome the problem with obliterative techniques. The abundance of techniques point to the fact that no single procedure is perfect. Moreover not all techniques lend themselves to subsequent otological procedures. We adopted the use of autologous fat grafts to obliterate the middle ear and mastoid cavities along with blind pit closure of the external canal either as an isolated procedure or as a preliminary to cochlear implantation. We use the same technique whether the problem is a discharging perforation (safe CSOM) with mucosal disease or an old radical cavity with recurrent cholesteatoma. The intermediate term results of 17 cases of mastoid obliteration (in 16 patients) performed in our centre between 1992 and 1997 are presented here.

PATIENTS AND METHODS

All 16 patients (nine male and seven female) suffered from long-term chronic middle ear disease with chronic sepsis leading in hearing loss (Table 1). While 12 (75 per cent) had safe variety of CSOM with perforations, four (25 percent) had cholesteatomatous disease. Pre-existing mastoid cavities (with discharge due to middle-ear mucosal disease) were present in 10 (62.5 per cent) cases at referral. The disease was bilateral in 14 (87.5 per cent).

Table 1. Clinical details of patients

16 patients (9 male; 7 female) [17 patients, one patient had bilateral obliterations] Age: 44-68 years	
Type of disease	*Number*
Bilateral CSOM	14
Unilateral CSOM	2
Middle ear mucosal disease	12
Recurrent mastoid cholesteatoma	4

Indications

The majority (16 procedures) were undertaken prior to cochlear implantation. However one operation had to be done *after* cochlear implantation due to extrusion of the cochlear implant (Nucleus 22) electrode into the ear canal through the old post-surgical deep meatal stenosis (Table 2). This patient had previously undergone bilateral mastoid explorations 40 years ago and the cavities had remained disease free.

Table 2. Surgery

(17 ears; 16 patients; one bilateral)	
Single obliteration + Implant	11
Obliteration + Implant + 3rd Look	2
Implant + Single obliteration (post implant)	1
Obliteration + Simultaneous implantation	1
Obliteration + Revision + Implant	2 (Total=17 ears)
Implants	Ineraid = 7
	Nucleus 22 = 7
	Nucleus 24 = 3

Procedure

This is usually a two stage operation at three months interval. In brief it involves:

Stage I: Post aural incision
Radical / Revision mastoidectomy
Permanent obliteration of the Eustachian tube opening.
Positioning of silicone strip over the promontory and round window
Obliteration of mastoid bowl with autologous free abdominal fat graft
Blind sac closure of the external auditory canal.

Stage II: Revision incision through old scar
Preparation of implant bed
Elevation of fat graft that now becomes encapsulated with fibrous tissue.
Removal of silicone strip
Cochleostomy and implantation
Closure

A detailed description of surgical technique will be found in the initial paper[2].

RESULTS

Obliteration of mastoid cavities with autologous abdominal fat was performed in 17 ears (16 patients; one bilateral) in association with cochlear implantation (Table 3).

Table 3. Details of fat obliteration

No	Pt	Previous Procedure	Our procedure and implant type (R)	(L)	Complication	Current position
1	TT	Middle ear MD (bilateral)	Oblit. Ineraid	-	Nil	Dry
2	GD	Mastoid cavity MD (L)	-	Oblit N22	Nil	Dry
3	AH	Middle ear MD (bilateral)	-	Oblit N22	Nil	Dry
4	JT	Middle ear MD (bilateral)	-	Oblit N22	Nil	Dry
5	AW	Mastoid cavity MD (bilateral)	Oblit N22	-	Nil	Dry
6	RT	Middle ear MD (bilateral)	-	Oblit Ineraid	Chole at 2nd look Clear at third look	Dry
7	MA	Mastoid cavity MD (bilateral)	-	Rev. Oblit. N24	Hole in blind pit	Dry
8	DM	Mastoid cavity (bilateral) CHOLE	-	Oblit. Ineraid	Temporary 7th palsy. Breakdown of blind pit. Exposed electrode	Discharging ear
9	EO	Mastoid cavity MD (bilateral)	-	Oblit N24	Nil	Dry
10	GH	Mastoid cavity (bilateral) CHOLE	-	Oblit N22	Nil	Dry
11	CT	Mastoid cavity (bilateral) CHOLE	Oblit. Ineraid	Oblit. Rev. Oblit. N24	Graft abscess (L). Chole. Hole in blind pit. Clear at third look	Dry # Ineraid
12	GC	Mastoid cavity MD (bilateral)	-	Ineraid	Nil	Dry
13	BR	Mastoid cavity MD (R)	Oblit N22	-	Nil	Dry
14	PK	Mastoid cavity MD (bilateral)	-	N22 Oblit (post implant)	Nil	Dry
15	PC	(L) Vestibular Neurectomy + (R) Labyrinthectomy CHOLE	Oblit Ineraid (Single stage)	-	Wound infection. Settled	Dry # Ineraid
16	ON	Middle ear MD (bilateral)	Oblit Ineraid	-	Nil	Dry

Abbreviations:
MD = mucosal disease
CHOLE = Cholesteatoma
N22/N24 = Nucleus cochlear implants
# = Ineraid pedestal broken due to trauma
Oblit = Fat obliteration of middle ear or mastoid cavity

Initial obliteration of the mastoid cavity and subsequent implantation three months later was carried out in 13 cases (76.4 per cent). In two (11.7 per cent) of these a third look was required to rule out residual / recurrent cholesteatoma (Table 2).

Obliteration and implantation were done simultaneously in a single operation in one (5.9 per cent). In another case (5.9 per cent) cochlear implantation had to be followed by obliteration because breakdown of the previously closed ear canal had resulted in protrusion of a loop of electrode out of the ear canal. This patient had undergone bilateral mastoid explorations for cholesteatomatous disease 40 years ago. The ears were stable and trouble free.

The procedure had to be revised in two cases (11.7 per cent) due to breakdown of external canal skin before cochlear implantation.

Complications

(1) *Temporary facial palsy.* In one patient (5.9 per cent) a temporary lower motor facial palsy occurred when the tip of a 18F gauge sucker strayed into the facial nerve canal. The nerve had been exposed by previous surgery. The palsy recovered completely at three months (Table 4).

(2) *Breakdown of ear canal skin closure (blind pit) after implantation.* In one patient the blind pit closure broke down three months after implantation and the ear started to discharge again (5.9 per cent). This was the patient who had the temporary facial palsy. Further surgery has been postponed indefinitely.

(3) *Recurrent / Residual Cholesteatoma.* Residual cholesteatoma pearls were noted and removed at the second operation in two cases (11.7 per cent) and a third look was needed to make sure that adequate clearance was done (Table 2).

(4) *Breakdown of ear canal skin closure (blind pit) before implantation.* In two instances (11.7per cent) the blind pit closure of the external canal broke down and the procedure had to be revised. The second attempt to obliterate the septic cavity has been successful in both cases.

(5) *Wound infection.* Post-operative wound infection occurred in one case (5.9 per cent) two weeks after surgery but settled with conservative measures.

Current Position

Out of 17 ears (16 patients) treated by this procedure 16 (94.1 per cent) are dry at the end of five years (Table 4). Fifteen patients have working implants while one is

not using his Ineraid implant which broke following trauma to the pedestal. In one patient (same as I) the exposed electrode is visible through the breakdown in the blind pit. However the implant (Ineraid device) still continues to function and he is not keen to undergo revision surgery.

Table 4. Results of surgery

Complication	
Cholesteatoma	2 (pearl found at implantation)
Recurrent sepsis	2 (abscess in fat graft and holes in blind pit)
Facial palsy for 3 months	1 (obliteration break down; implant still working)
Wound infection	1
Current situation	
Dry ears	16
Discharge , exposed electrode	1
Working implants	15
Broken implants	2

DISCUSSION

A variety of techniques of obliteration of the mastoid cavity using different flaps and materials have been described. Our technique has been successful in eliminating middle ear sepsis in 94.1 per cent of cases and has lent itself very well to in preparing the septic ear for a sterile cochlear implant device.

Obliterating materials

Bone paté or hydroxyapatite obliteration are popular in CSOM cases where the loss is conductive. But the obliterating material may mature to solid bone. This would have to be partly drilled away for implantation of an electrode and is therefore considered undesirable in these special circumstances.

Pedicled temporalis muscle grafts may shrink and sometimes cause morbidity when the patient finds chewing painful and restricted post-operatively.

The intermediate results with fat are encouraging and prove the material to be quite stable. The choice of autologous abdominal fat is logical if one considers its: (1) abundant supply (2) easy accessibility (3) low metabolic rate and (4) resistance to necrosis when used as a free graft in a bony cavity. Being a single block of tissue which develops a fibrous capsule around itself, it is easy to elevate or reduce at the second stage for implantation[2].

Pre-existing cholesteatoma

The risk of occult and residual cholesteatoma after obliteration is always present. By using the operation for implantation as the "second-look" procedure such as is recommended for combined approach technique, may help minimise the risk. Where pearls of cholesteatoma are found and removed at implantation a "third look" will be needed as it is well known that cholesteatoma pearls are picked up much earlier clinically than by imaging. This has been necessary in two of our patients (11.7 per cent).

MRI scanning

Imaging to monitor the recurrence of cholesteatoma is also desireable but difficult in these situations because cochlear implant patients cannot readily have MR imaging. However with the newer MR compatible implants fat suppression sequence with gadolinium will be helpful in making this distinction.

CT scanning

High resolution CT scanning can be done if needed. However, the interpretation of the scan is difficult and the predictive value is low. The confidence with which recurrent cholesteatoma can be detected depends on several factors:

(1) The slice angle and the window level determines the amount of a particular tissue that is included in that window.
(2) The amount of tissue influences the "volume averaging" which the computer utilizes to measure tissue density.
(3) The greater the difference in Hounsfield units (i.e.the greater the difference in densities) between the two tissues the easier it is to differentiate between them.

For example hydroxyapatite (Hounsfield Unit +1900) is easily distinguished from cholesteatoma (-40)[3] but hydroxyapatite is not useful in cochlear implant surgery for reasons mentioned above. Fat (-100) can also be differentiated from cholesteatoma (-40) but with difficulty. The distinction between fibrous tissue (+40) and recurrent cholesteatoma (-40) is also not easy. The difference between muscle (+40) and cholesteatoma (-40) is similarly blurred and is open to observer interpretation error. A Hounsfield difference of at least 100 units and an adequate tissue volume is necessary for confident distinction between adjacent tissues. Therefore the sensitivity of CT scanning to detect recurrent cholesteatoma is poor.

Serial CT scans can be useful and may at best postpone the need for a third look. Delayed cases may show evidence of bone erosion. However at present the use of high resolution CT cannot replace the need for surgical exploration of the obliterated cavity. The role of spiral CT scanning also remains to be evaluated.

Clinical monitoring

The patients undergo regular close clinical monitoring and open surgical exploration is the preferred option at the earliest suspicion of cholesteatoma. The advantage with cochlear implant patients is that they need lifetime care in the same hospital department and seldom default on appointments. Schuknecht's series of neglected huge cholesteatoma with facial palsy[4] is therefore unlikely in our patients who are under regular otologic supervision. Obliteration of the middle ear and mastoid cavity not only makes patients suitable for cochlear implantation but improves their quality of life further by allowing them at least one dry ear.

CONCLUSION

The technique which was started five years ago has proved satisfactory and the results are encouraging. The proof of the operation will be if others find it equally satisfactory for cochlear implant candidates who present with intractable bilateral CSOM.

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CHAPTER 3

NON/LIMITED USE OF COCHLEAR IMPLANTS

3.1

NON-USERS AND LIMITED USERS OF COCHLEAR IMPLANTS

J. Ray
T. Wright
C. Fielden
H. Cooper
I. Donaldson
D.W. Proops

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ABSTRACT

This study evaluates the incidence of non use and limited use of cochlear implants and attempts to identify predictors of such outcome. This involved a retrospective analysis of questionnaires, clinical and audiological data of 423 cochlear implants recipients from the Midland Adult Cochlear Implant Programme and Birmingham Paediatric Cochlear Implant between 1990 and 2000. Of the 172 children in the paediatric programme 5 (2.9%) were non / limited users and of the 251 adults in the adult programme 7 (2.78%) were non / limited users. Thus in total 12 (2.82%) recipients were deemed to be either limited users (0.94%) or non-users (1.89%) of their implants. The mean age at implantation of the non user group was 22.2 years (range 9-56 years) and 42.5 years (range 21-64 years) for the limited user group. The mean duration of deafness prior to implantation was around 10 years in both the paediatric and adult groups.

In the paediatric group peer pressure played a prominent role in the non-use of implant. On the other hand depression, tinnitus, concomitant neurological problems and non auditory stimulation seemed to be the predominant reasons in the older age group. The reasons for limited use were cognitive slowing and background noise. Non users tend to be younger than the limited users.

INTRODUCTION

Non use and limited use of implants is a recognised phenomenon but there is little published in the literature. The majority of reports of non-use are anecdotal and isolated. But as the cohort of cochlear implant recipients grows this may well be an unwelcome evolving phenomenon. It therefore becomes imperative to monitor instances of non use and to identify their antecedents.

Cochlear implantation is a reasonably safe procedure with few complications[1]. On the whole, recipients accept implants well and rejection rate is very low. One large study placed implant non use at 3%[2]. However the implications of non-use are significant for both the patient and the implant team. Most importantly the cost of an elective non-use of implant has to be accounted for in the health care budgeting[3]. Most implant teams are keen to avoid such occurrences and hence the continued search for predictors of good performance.

This study attempts to analyse predisposing factors for elective non use or limited use of cochlear implants.

PATIENTS AND METHODS

This is a retrospective study of all individuals implanted in the Midland Adult Cochlear Implant Programme and Birmingham Paediatric Cochlear Implant Programme between 1990 and 2000. The transfer from the paediatric to the adult section takes place at sixteen years of age.

All cochlear implant recipients were required to complete a range of self report questionnaires at the time of assessment for implantation and also subsequently. These along with the clinicians notes provided the background information on the individual cochlear implant users. Details of interviews with recipients conducted by the audiologist, hearing therapist and speech therapists were reviewed. The following factors were specifically assessed: (i) aetiology of deafness (ii) age at onset of deafness and at implantation (iii) duration of deafness (iv) surgery and minor or major complications (v) reasons for limited use or non use. Limited use was defined as use of implant for less than two hours per day. Non use was complete rejection of implant use.

The duration of deafness was defined as the length of time that the ear subsequently implanted was deaf. In case of progressive and congenital hearing loss values for the overall duration of hearing loss and that for the profound deafness were recorded separately. The age referred to in the paper is the age of the patient at implantation.

All potential recipients underwent preliminary audiological assessment including pure tone audiometry and auditory brainstem responses and computed tomography and all children and selected adults underwent magnetic resonance imaging. All potential recipients and / or their parents were consulted and counselled prior to surgery. The decision to proceed to cochlear implantation was a multidisciplinary one.

The surgery was standardised involving a cortical mastoidectomy, posterior tympanotomy and cochleostomy. Peri operative tests included stapedial reflex testing and electrical auditory brainstem responses and more recently neural

response telemetry. Post operatively all recipients underwent a check x-ray to confirm correct position of the electrode array within the cochlea. All recipients were planned to be switched on at one month post implantation. Audiological results were collected at one month, three months and nine months post implantation. Subsequently these cochlear implant users were reviewed at varying intervals by the audiological scientist and hearing therapist on the team.
Recipients who missed appointments were contacted directly by the hearing therapist (TW) and the audiologist (CF). In the event of non use of partial use both telephone and postal enquires were made and where possible visits were undertaken. Problems were discussed freely and frankly with the patient and the family and attempts were made to address the issues that arose from the discussions.

RESULTS

A total of 423 individuals had been implanted in the Birmingham programmes (adult and paediatric) between 1990 and 2000. Of these 172 were children and 251 adults. The age range at implantation for the paediatric programmes was 18 months to 16 years and for the adult programme was 20 years to 86 years.
Over the ten year period 12/423 (2.82%) recipients were deemed to be either limited users (0.94%) or non-users (1.89%) of their implants. (Table 1) In the paediatric group 5/172 (2.9%) were non/limited users and in the adult group 7/252 (2.78%) were non/limited users (Table 1).

Age (non/limited user group)
The age range at implantation of this group was 9-64 years (Mean 29.2 years, median 21 years). The nonuser group had a mean age of 22.2 years (range 9 -55 years) at implantation and the commonest aetiology was either unknown or meningitis. The limited user group on the other hand was of varied aetiology and the mean age at implantation was 42.5 years (range 21 -64 years).
The mean age at implantation was 11.2 years in the paediatric group and 42.1 years in the adult group (Table 1).
The recipients with limited use or non use of implants were divided into four groups i.e. children less than ten years age, teenagers less than twenty years age, adults under forty years and adults over forty years. The number of individuals in each group were 3 (0.7%), 2 (0.5%), 4 (0.9%) and 3 (0.7%) respectively (Table 1).

Table 1. Patient demographics

Programme	*Recipients*	*Non/Limited user*	*Mean age at implantation (Yrs)*	*Mean duration of deafness (Yrs)*
Paediatric	172	5 (2.90%)	11.2	10
Adult	251	7 (2.78%)	42.1	11.15
Combined	423	12 (2.8%)	29.2 [9 - 64]	6.5 [2 – 20]
Non-users	8 (1.89%)	Mean age: 22.2 years		
Limited users	4 (0.94%)	Mean age: 42.5 years		
Male:Female :: 8 : 4				
Age grouping of non / limited users				
Child (<10)	3 (0.7%)			
Teens (<20)	2 (0.5%)			
Adult (<40)	3 (0.7%)			
Adult (>40)	4 (0.9%)			

Duration of hearing loss (non/limited user group)

The average duration of deafness in the non user group was 9.2 years while that for the limited user group was 14.2 years.

In the paediatric group the mean duration of deafness was 10 years while in the adult group this was 11.15 years.

Aetiology and type of deafness (non/limited user group)

The aetiology was unknown in five, meningitis in three, head injury and subsequent meningitis in one, otosclerosis, measles and road traffic accident in one each (Figure 1).

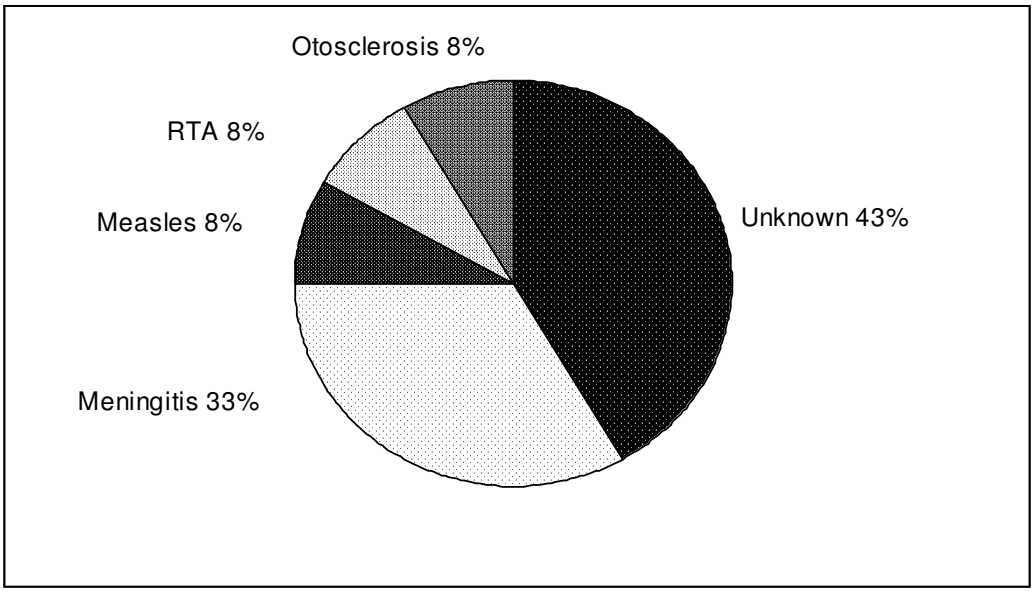

Figure 1. Aetiology of Deafness

The deafness was progressive in 5 and sudden in 7. One individual had other serious co morbidities from meningitis in the form of blindness, bilateral labyrinthine failures, severe tinnitus and multiple neurodeficits. (Table 2)

Table 2. Clinical details

No	Sex	Age Implanted	Aetiology	Years Deaf	Insertion	Implant	Status	Other Problems
1	M	9	Unknown	9	Complete	N22	Non user	Embarrassed with peers Family pressure
2	F	9	Unknown	9	Complete	N22	Non user	Hydrocephalus, VP shunts(bilateral),
3	M	10	Meningitis	6	Complete	N22	Non user	Disliked CI, Embarrassed with peers,
4	M	12	Unknown	12	Complete	N22	Non user	Hasty Decision, Embarrassed with peers
5	M	16	Measles	14	Complete	N22	Non user	NAS, unpleasant sensation
6	F	41	Meningitis	1	Partial	N22	Non user	Neurodeficits, Disorientation, Tinnitus, Labyrinthine failure, Blindness
7	F	28	Meningitis	20	Complete	N22	Non user	Tinnitus, NAS, Unpleasant Noise
8	F	56	Unknown	3	Single Channel	Med El	Non user	Tinnitus, Depression, Arthritis, Labyrithitis ossificans
9	M	30	RTA	2	Complete	N22	Partial	Cognitive slowing
10	M	21	Unknown	12	Complete	N22	Partial	Embarrassed with peers,
11	M	55	Meningitis	1	Complete	N24	Partial	Cognitive slowing
12	M	64	Otosclerosis	42	Complete	N24	Partial	Factory worker, Noise

Abbreviations: N: Nucleus, NAS: non auditory stimulus, RTA: road traffic accident.

Reasons for non/limited use

The reasons for non use of the implant were lack of facility in the older implanted children and pressure from family and peers (Table 3). The age range for these recipients was 9 – 12 years at the time of implantation. The range of implant use in these individuals was between 9 months and 14 months most of which was intermittent. Two implant users had worsening tinnitus and blamed it on the implant. One had received the single channel device for labyrinthitis ossificans and suffered from depression and disabling arthritis. The other also experienced unpleasant loud noises from his implant. Cochlear implant recipients with meningitis who had other neurological sequelae in addition to their deafness readily became non users. One recipient had deafness due to measles at the age of two years and was implanted at sixteen years of age. He complained of unpleasant non auditory stimulation and became a non user within a year of implantation.

Table 3. Reasons for reduced usage

Reasons in the younger patients:
Peer pressure and family pressure
Poor performance in older children
Meningitis with other sequelae
Unrealistic expectations
Reasons in the mature patients:
Neurological problems
Cognitive slowing
Tinnitus, Depression, Noise
Non-auditory stimulation

Amongst the limited users the youngest person (21 years at implantation) felt embarrassed to use the implant initially and after three years has started wearing this at home for a few hours occasionally. The oldest person in the group (progressive deafness due to otosclerosis from age of 22 years and implanted at 64 years) found the implant difficult to use in his factory environment and therefore became a limited user using it only at home. This man remains a limited user even in his retirement from factory work. The other two individuals suffered cognitive slowing due to coexisting medical conditions.

DISCUSSION

A cochlear implant user may elect to be a non user of their device because of perceived lack of benefit they derive from the device. However they form a small minority of the implantees but nevertheless a very important group. They fall into either non users or limited user (less than two hours of implant use per day). Although partial or non use of implants is known, published reports of these are few.

The implant user may have a deficit in the central auditory pathways or may be experiencing non auditory stimulation. There may have been a medical or a surgical complication or the individual may feel they do not subjectively gain any benefit from the device. Inappropriate expectations, early onset deafness, co morbidity from medical conditions, tinnitus and depression may all contribute towards non use. While some may perform poorly at formal testing and yet be pleased with their implants, others may choose not to use their implant for no predictable reason. Summerfield and Marshall[1] quoted an elective non use rate of

around 3.5% and predicted a cumulative elective non use rate of around 11%. Our non use rate was 1.89% and limited use rate was 0.94%.

It has been thought that medical or surgical complications of cochlear implantation may make the recipient a non user[4]. Complications are classified as major when there is extension of the original inpatient episode or requiring readmission and removal of the implant or an event like facial nerve damage. Minor complications are those requiring non-routine outpatient care. In a multicentre study it was found that 19.8% non users experienced minor complications and 8% experienced at least one major complication[4]. This is similar to that reported in North American series[5]. However the later series did not report on non-use arising out of complications. It is interesting to note that none of the recipients in this study experienced any major or minor complications that could have contributed to them becoming non-users. Several reports of revision surgery and reimplantation of cochlear implants have been published[6-13]. Revision surgery also does not seem to render a patient a non user of the implant.

Poorer outcome has been shown in older recipients and in those who have been profoundly deaf for longer periods prior to implantation[14]. The mean duration of deafness prior to implantation in both paediatric and adult groups in our study was around 10 years. Age at implantation also has a bearing on whether an implant user becomes a non user or a limited user. In our series the age of the individuals at implantation did affect whether they became a non user or a limited user. The non user group was much younger (mean age 22.2 years) than the limited user group (mean age 42.5years).

Those with meningitis, especially if they have other neurological sequel apart from their deafness tend to reject the implant more commonly. The other aetiologies of deafness do not seem to determine the degree of future implant use.

CONCLUSION

In children the commonest reason for non-use of implant was lack of facility probably because they were older children at implantation. Amongst this group teenage peer pressure played a prominent role because they tend to have their educational and social support from the deaf community. Meningitis, especially with other neurological sequel in addition to deafness increases possibility of

rejection of the implant On the other hand depression, tinnitus, concomitant neurological problems, and non auditory stimulation seemed to be the predominant reasons in the older age group. The reasons for limited use were cognitive slowing and background noise. Non users tend to be younger than the limited users. The mean duration of deafness prior to implantation in both the paediatric and adult groups was about ten years.

It is worth distinguishing between limited use and non use as the former outcome is a lesser disappointment for the providers of health care.

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CHAPTER 4

ELECTROPHYSIOLOGY

4.1

THE ROLE OF AUDITORY STIMULATION IN THE MATURATION OF THE AUDITORY PATHWAY

J. Ray
W.P.R. Gibson
H. Sanli

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ABSTRACT

Objective: To compare the maturation of the auditory pathway, as shown by electrical brainstem auditory potentials (EABR), in ears in which there had been prior auditory stimulation and ears in which no prior auditory stimulation had occurred.
Study design: Prospective, longitudinal, analysis.
Main outcome measure: Change in waveform of eV and absolute latency of wave eII, eIII and eV on Implant Evoked Auditory Brainstem Response (ImpEABR).
Setting: Tertiary referral centre
Materials and methods: Electrophysiological data collected prospectively from ears which received cochlear implants. ImpEABR were recorded. 70 children, implanted after January 2000, were selected according to a strict inclusion exclusion protocol. All the children had received a 22 channel Nucleus cochlear implant (CI24 series). Intraoperatively, ImpEABR were recorded using the Medelec Synergy® system in conjunction with the Nucleus NRT® software. The ImpEABR latencies of wave eII, eIII, eV and morphology of wave eV were assessed.
Results: The ImpEABR alter during the first 12 months of life. The latency becomes shorter during this period and the morphology of eV alters from a broad shape to a more distinct waveform. This appears to occur independently even in the absence of auditory stimulation.
Conclusions: The development of the electrical brainstem auditory potentials is not dependant on auditory stimulation.

INTRODUCTION

Commonly, it has been supposed that prior exposure to acoustic stimulus is a prerequisite for maturation of the auditory neural pathway. However recent works[1,2] have indicated that this precondition may not be an absolute necessity and the peripheral auditory pathways may develop despite auditory acoustic deprivation. This paper is an attempt to assess the role of auditory stimulation in maturation of the auditory pathway.

Children with a congenital hearing loss in which it was determined that there was no useful hearing prior to cochlear implantation were compared with children who

had an acquired hearing loss (meningitis) and had had normal hearing prior to the onset of the deafness.

PATIENTS AND METHODS

Only patients who had full insertion of 22 electrodes of the Nucleus 24 device were included in the study.

Meningitis group

This group comprised of 19 patients who suffered sudden profound deafness due to meningitis. None had any previous history of hearing loss. There was no cochlear abnormality on CT, and no brain abnormality was seen on MRI. Apart from the hearing loss, there were no other neurological problems. Pre-operatively, these ears had no potentials on round window electrocochleography (RW ECochG) and no auditory brainstem responses (ABR) using stimuli between 500Hz and 8kHz at 100dBHL. The mean duration of deafness (from meningitis to implantation) was 6.69 months. All 19 patients were implanted unilaterally with Nucleus CI24 multi channel cochlear implant (Cochlear Corporation, Australia). Post-operatively all children had made satisfactory progress using their implants.

Congenital group

This group comprised of 70 children who were profoundly deaf from birth. This was confirmed preoperatively by RW ECochG and only patients with thresholds worse than 100dB between 500Hz and 8kHz were included. None of the children included in the study had any useful hearing aid experience prior to the testing. There was no cochlear abnormality on CT, and no brain abnormality was seen on MRI. Apart from the hearing loss, there were no other neurological problems. All children received unilateral implantation with Nucleus (Cochlear Corporation, Australia) multi channel cochlear implant. Excluded from the study were children with syndromic deafness. Premature infants were included if born after 34 weeks gestational age.

Test criteria

Implant evoked auditory brainstem potentials (ImpEABR) recorded immediately after cochlear implantation in monopolar (MP) 1+2 mode of stimulation. Data was collected prospectively using the same Medelec Synergy® Evoked Response system to maintain consistency. The Nucleus® Neural Response Telemetry

software (Cochlear Corporation, Australia) was used as the stimulator to evoke the ImpEABR via the Nucleus 24 device. For channel 22 recordings a current level of 228 implant units with a pulse width of 25 microseconds was used while a current level of 112 implant units with 25 microsecond pulse width was used for channel 11 recordings using the Nucleus 24 device. These recordings were thought to provide representative traces from two different segments of the cochlea.

Absolute latencies and the morphology of the waves were used for analysing the data. SPSS® for Windows statistical software package was used for the statistical evaluations.

Three different absolute latencies were measured along each trace, eII, eIII and eV. A classification for defining the waveform morphology was devised for this study. This classification divides the waveform into three categories according to the shape of eV. Type I is where there is a definite peak visible, Type II refers to a smoother and rounder trace while Type III has a much flatter trace.

RESULTS

Meningitis Group

The nineteen patients in the control group suffered sudden deafness due to bacterial meningitis after variable periods of normal hearing. Sixteen were pneumococcal, two streptococcal and one meningococcal in origin. The mean age of the group was 49.9 months (range 11-203months). The waveform morphology of eV was well formed (Type I) after 12 months (Figure 1 traces B and C). The waveform (Figure 1 traces B and C).and latencies (Table 1) of the waves 1.45ms for eII, 2.10ms for eIII and 4.10ms for eV are similar to that obtained in adults after implantation (Figure 5).

Congenital group

This group of seventy patients was used to evaluate the effects of age, development and maturation on the electrophysiological parameters. (Table 2). The mean age of this group was 29 months (range 5-60 months, SD 13.8 months). The latencies of all waves show a steady decrease in value (probably signifying maturation) until the age of one year (traces A-F in Figure 2 and traces A-D in Figure 3). After this age, the latencies of the waves remain quite stable and values conformed with those obtained in adults (Figure 5). The trend for change in wave latencies with age is shown in Figure 4.

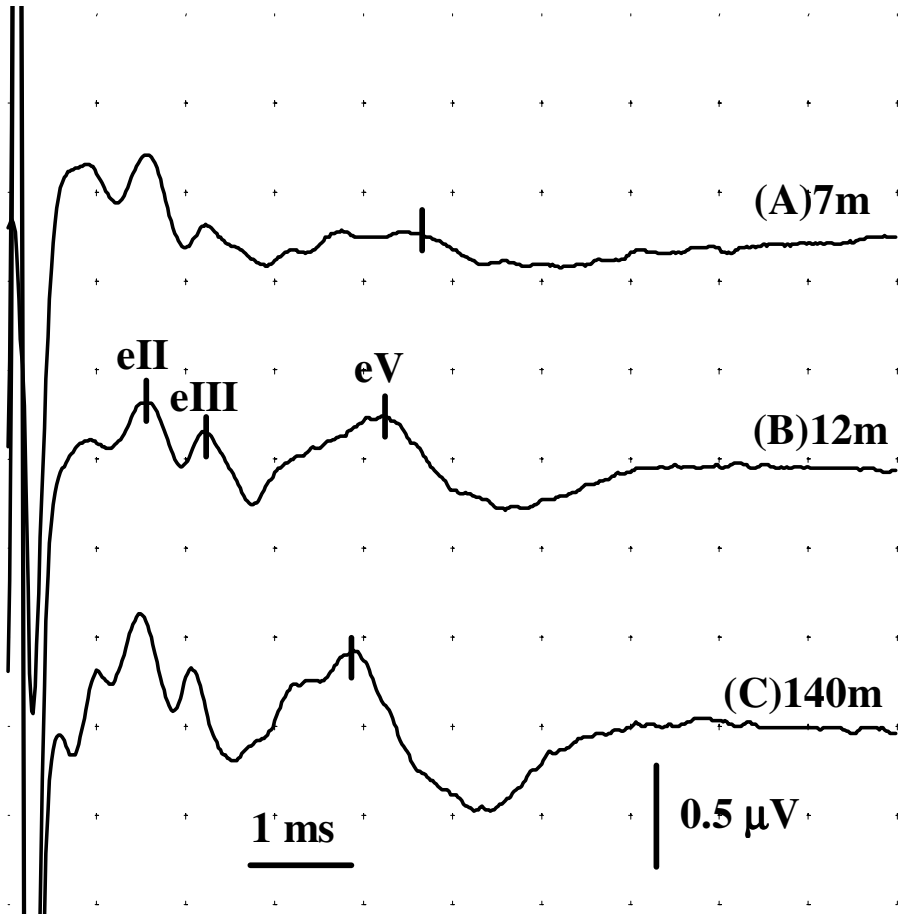

Figure 1**.** Changes in IMP EABR latencies and waveforms at various ages (in months) in the meningitis group. A and B are early traces while C represents typical mature trace. Peaks of waves are marked eII, eIII and eV.

Table 1. Absolute latencies of ImpEABR waves in the meningitis group

Age Range at onset of deafness	Number of patients	Mean age (months) (SD)	Absolute Latencies eII (ms) (SD)	Absolute Latencies eIII (ms) (SD)	Absolute Latencies eV (ms) (SD)
3 -12 m	2	11m	1.6	2.25	4.31
13-24 m	6	20.28m (3.49)	1.53 (0.24)	2.22 (0.16)	4.26 (0.24)
>24 m	11	72.36 (57.3)	1.45 (0.19)	2.10 (0.21)	4.10 (0.46)
N=19 Age range 11-203m					

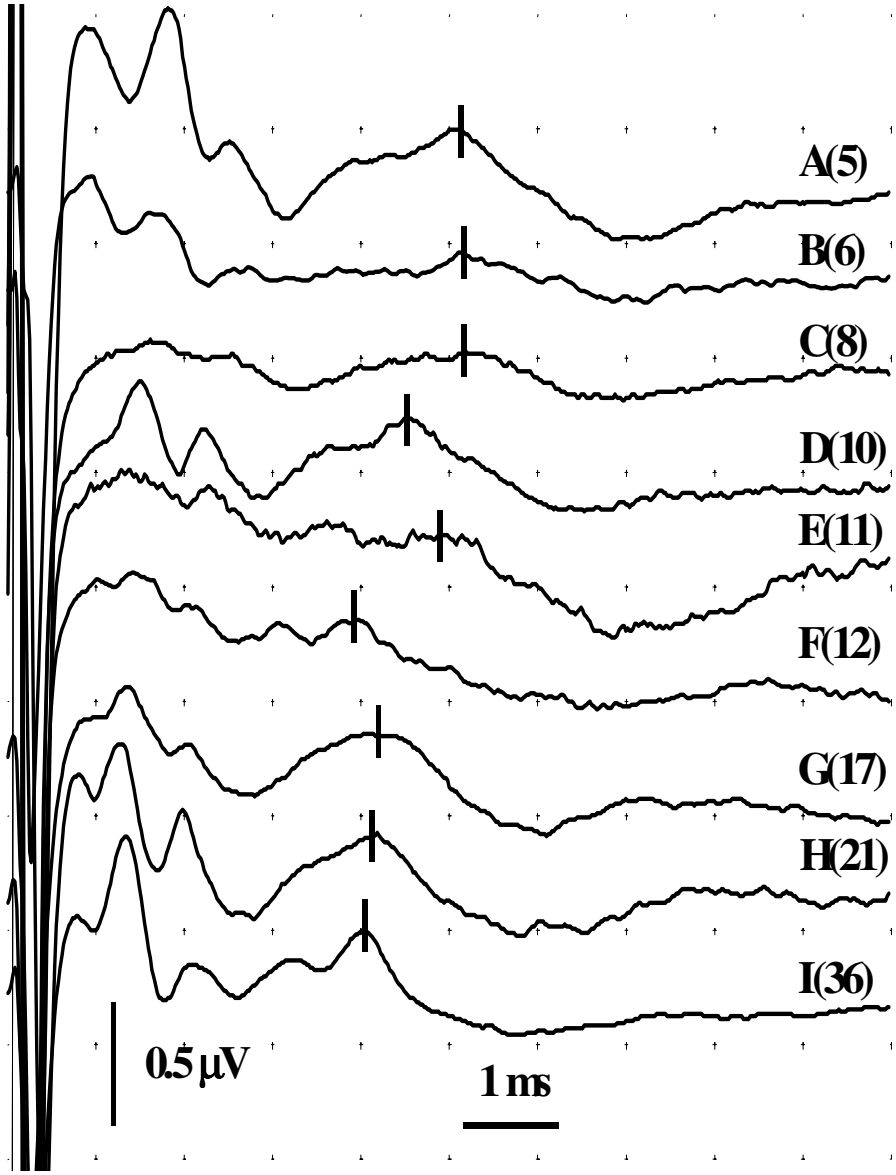

Figure 2. Changes in IMP EABR latencies and waveforms (Channel 11 recordings) at various ages in the congenital deafness group. A-I are patients with age in months. Peaks of waves are marked eII, eIII and eV.

Similar changes in the waveform of eV of ImpEABR occurred and the mature waveform of eV was reached by 12 months age (traces G – I in Figure 2 and traces E- H in Figure 3). The mean latencies of eII and eV after 12 months are 1.4msec and 4msec respectively.

The rates of development of the latencies in the two groups as a function of age are very similar.

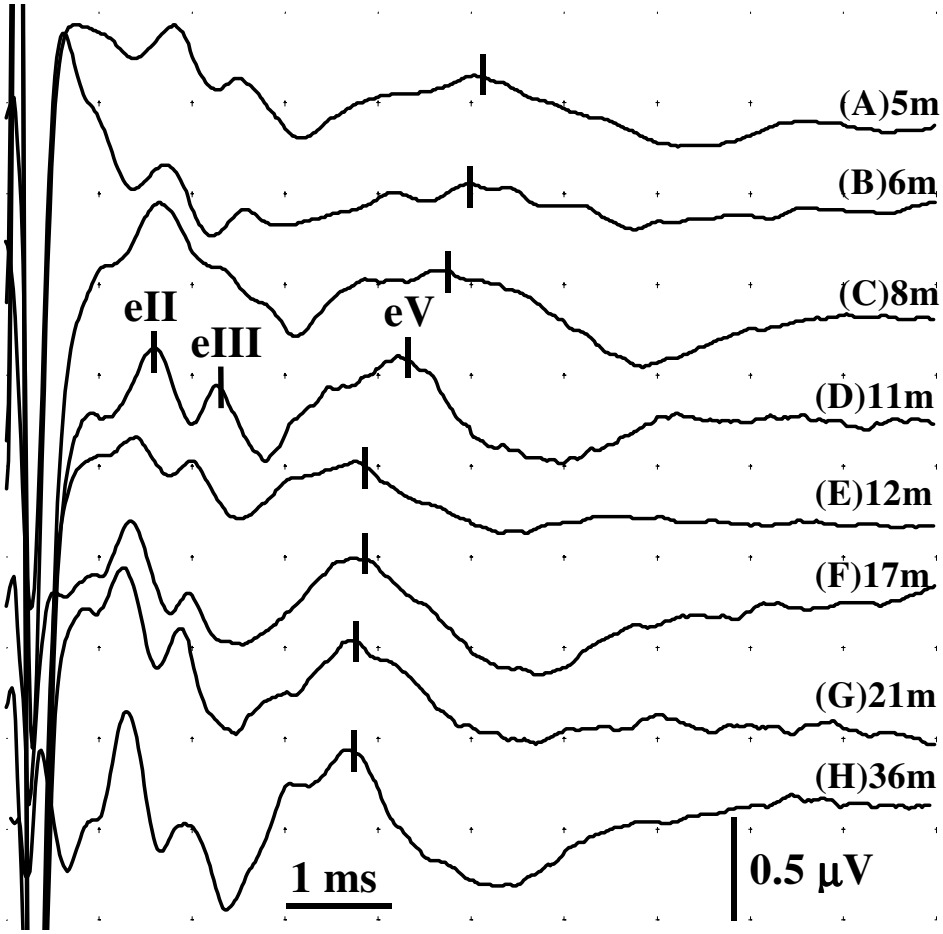

Figure 3. Changes in IMP EABR latencies and waveforms (Channel 22 recordings) at various ages in the congenital deafness group. A-H are patients with age in months. Peaks of waves are marked eII, eIII and eV.

Table 2. Absolute latencies of ImpEABR in the congenital deafness group

Age Range at implant (testing) months	Number of patients	Mean age of group in months (SD)	Absolute Latencies eII (ms)	Absolute Latencies eIII (ms)	Absolute Latencies eV (ms)
3 – 6 m	2	5.5 m	1.83 (0.03)	2.37 (0.10)	5.02 (0.03)
7 – 9 m	2	8 m	1.63 (0.07)	2.23 (0.05)	4.63 (0.11)
10 – 12 m	4	11m	1.45 (0.13)	2.11 (0.12)	4.05 (0.25)
13 – 24 m	22	18.95 (2.9)	1.41 (0.14)	2.02 (0.28)	4.15 (0.47)
25 – 36 m	20	28.68 (3.7)	1.49 (0.18)	2.08 (0.16)	4.09 (0.38)
37 – 60 m	20	47.21 (6.7)	1.39 (0.48)	2.06 (0.21)	4.02 (0.31)

N = 70; Mean age of group 29 months;
Range: 5-60 months; SD : 13.86months

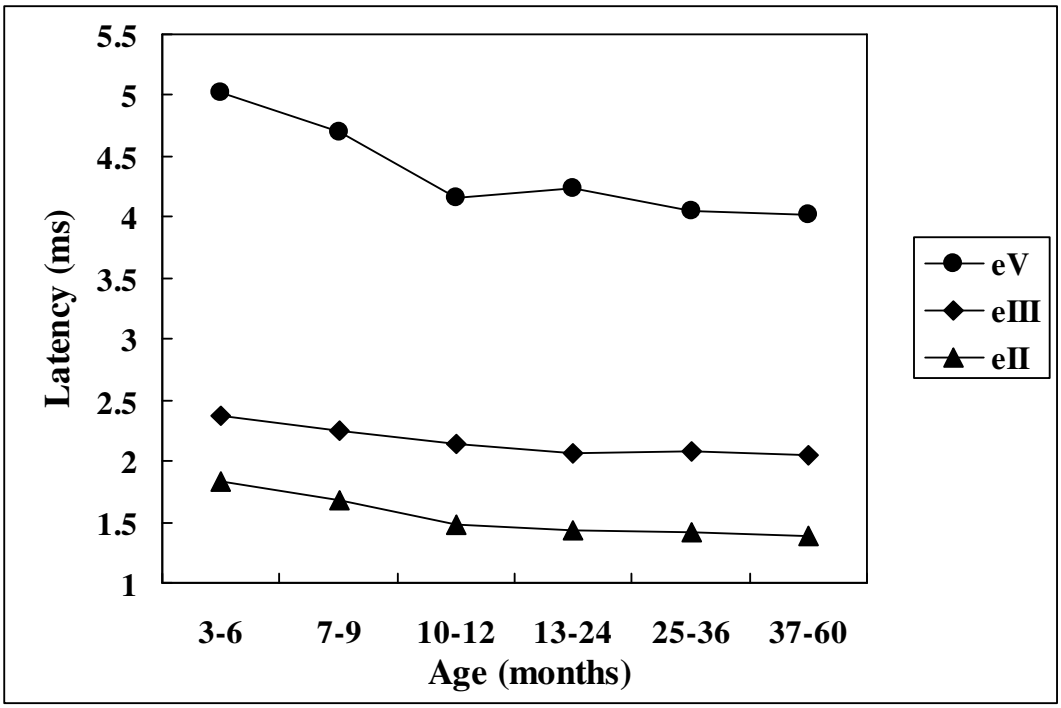

Figure 4. Trend of maturation latencies of IMP EABR waves over time

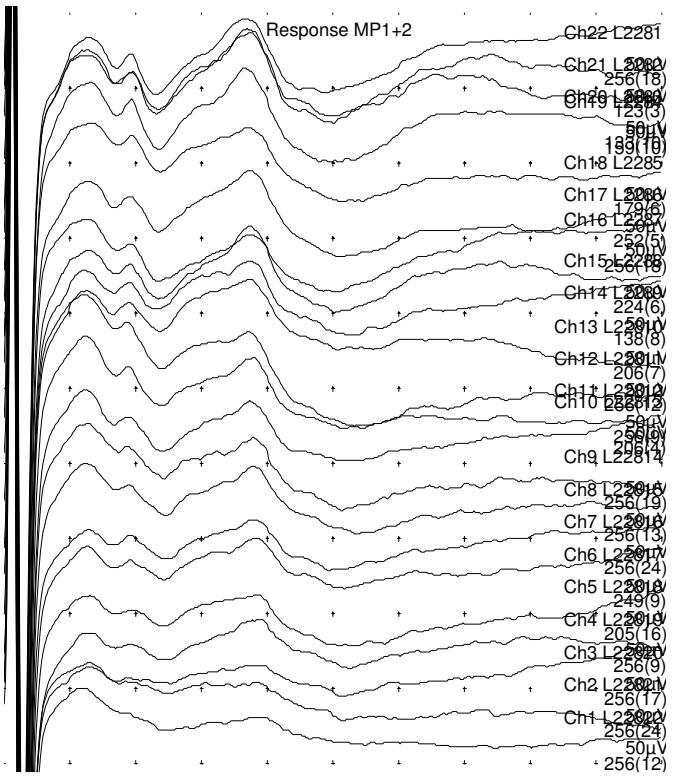

Figure 5. Typical Imp EABR traces in adult

Analysis

The null hypothesis in this study was that there is a difference in waveform latencies between those deafened at birth and those deafened later. The results from our studies failed to show any statistically significant difference between the two groups. This implies that auditory acoustic stimulation probably plays no role in the maturation of the auditory pathway.

DISCUSSION

Traditional view

It has been presumed that auditory stimulation is necessary for the morphological maturation of the auditory pathways and various studies have been quoted to support this view. The Shaker-2 mouse mutant is an animal model, where a cochleo-saccular type of genetically induced inner ear degeneration occurs. Morphological signs of degeneration are evident in the 3rd postnatal week and a severe and almost total degeneration occurs by the age of 6-9 weeks. In this mouse variety, the brainstem auditory nuclei stop developing around age day 14 while the brainstem itself continues to grow until day 150. It was postulated that early cochlear degeneration prevented the maturation of the brainstem auditory pathway[3].

In an examination of 39 human brains (ranging in age from the 29th week of gestational age to the 70th year of life), it was demonstrated that myelination takes place during the1st year of life and this was thought to be a prerequisite for functional maturation[4]. It was presumed that normal auditory development depended on adequate stimulation during this sensitive period of life.

The correlation between the functional and morphological maturation of the auditory pathway was studied in preterm and term infants by Inagaki et al[5]. They found that the peak latencies and I-V interpeak latencies auditory brainstem response (ABR) gradually decreased during the third trimester and the first 2 years after birth. They also calculated the pontine auditory conduction velocity (PACV) which showed dramatic changes during the first 2 years of life. The PACV value at the ages between 2-4 years was similar to that of adults. From magnetic resonance imaging studies they commented that myelination in the lateral lemniscus proceeded from the late foetal to the infantile period, and the myelin sheaths of large diameter nerve fibres increased mainly in the infantile period.

Based on this histomorphometrical investigation, the development of PACV was determined to reflect the maturation of nerve cells in the upper nuclei as well as the myelination of small and large fibres in the auditory pathway.

Many other authors[6-9] have demonstrated a decrease of average latency values of auditory brainstem responses during the first 2 years after birth. It has also been shown that peripheral structures of the hearing pathway are significantly more mature at birth than are central auditory structures.

Some authors[10] have felt that the auditory pathway may be functionally more intact in congenitally deaf children than in postmeningitic deaf children. Our experience however shows that the rate of maturation in the two groups is comparable. This was also the finding at a previous work published from this department[11].

ABR and EABR

Most of these studies have used auditory brainstem responses to quantify the state of maturation of the auditory nervous system both in humans and animals. Eggermont[12] has pointed out that although single unit recording using evoked potentials can be made in animals to compare the rate of maturation, the reliability and applications of such findings to humans is uncertain.

One study compared ABR traces in neonates in a neonatal intensive care unit (NICU). This study concluded that although ABR contributes information on the brainstem maturation, ABR alone is a poor predictor of the neurological outcome because of the large inter-individual variability of the responses in neonates[13].

ABR and EABR traces have been compared in human and animal recordings[14] and it has been proposed that latency, morphology, and magnitude be used for identification and analysis of EABR components.

Evolving Concept

Eggermont[15] has observed that the development and maturation of the human auditory system appears to occur in parallel at all levels from middle ear to cortex. Premature birth does not seem to affect the maturation rate or the maturity of the auditory brainstem potentials. He deduced that previous sound exposure did not seem to affect physiological maturation of the auditory pathway.

It is however known that the development the central auditory system is affected by prolonged periods of sound deprivation extending into early childhood[16]. The onset and duration of the period of deafness prior to cochlear implantation is an

important determinant of outcome. This is the reason why early implantation is considered crucial in congenital deafness for a satisfactory outcome.

Another study[1] showed that characteristic forms of cochlear nucleus neurons develop normally despite the absence of cochlear nerve input (produced by early destruction of the otocyst, embryonic precursor of the inner ear, in chick embryos). Otocyst removal however induces formation of permanent functional aberrant axonal projections to the ipsilateral cochlear nucleus from the contralateral cochlear nucleus.

In a recent study one group looked at EABR latencies in acoustically deprived cats and compared these with ABR from normal hearing cats[2]. In addition, morphological analyses of the cochlear nuclei and the auditory cortex and their subdivisions were examined. The EABR latencies demonstrated that the peripheral auditory pathway is more independent of auditory or external electrical stimulation than the central regions.

CONCLUSION

Our study suggests that EABR latencies will decrease and will assume the adult waveform independent of any acoustic or electrical input.

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4.2

BRAINSTEM AUDITORY NEUROPATHY, HAIR CELL DESYNCHRONY AND COCHLEAR IMPLANTATION

J. Ray
W.P.R. Gibson
H. Sanli
A. Haddon

Otology Neurotology (submitted)

ABSTRACT

Objective: The term Auditory neuropathy (AN) has been coined to describe a spectrum of hearing disorders characterised by absent or abnormal brainstem responses in the presence of normal otoacoustic emission. Typically ears affected by AN perform poorly using conventional hearing aids. A new classification based on the site of lesion revealed by electrophysiological tests is evaluated. The outcome of cochlear implantation in different types of AN is discussed.
Study Design: A longitudinal, prospective, cohort study of all cochlear implant patients between 1984 and 2003.
Setting: Tertiary care centre.
Patients: All subjects were tested using round window electrocochleography (RWEcochG), trans-tympanic electric auditory brainstem responses (TTEABR), neural response telemetry (NRT) and implant-evoked electric auditory brainstem responses (ImpEABR).
Intervention: Cochlear implantation
Main outcome measure: Presence of abnormal positive potentials (APP) on RWEcochG**,** Implant evoked auditory brainstem responses (ImpEABR) and speech discrimination scores after implantation.
Results: AN was classified into the following categories: hair cell desynchrony (HCD) when the pathology was localised to the outer hair cells or the hair cell-auditory nerve synapses, and brainstem auditory neuropathy (BAN) when the pathological lesion affected the brainstem afferent auditory pathway. A further group Central Auditory Neuropathy (CAN) exists but this is not discussed in this review.
A study cohort of 65 (16.17%) patients with diagnosed auditory neuropathy were discovered among 402 paediatric patients who underwent preoperative and intraoperative electrophysiological testing for cochlear implantation between 1984 and 2003. 42 (65.6%) were confirmed as HCD with the presence of APP while 9 (13.9%) were BAN. 14 (21.5%) had evidence of both HCD and BAN. An age matched group of 70 patients without HCD or AN was used as controls for the study. The mean post implant speech perception scores (Melbourne categories) were 6.26 for the HCD group and 2.57 for the BAN group ($p<0.005$). The mean speech perception scores in the control group was 4.66 ($p<0.005$).
Conclusion: After cochlear implantation**,** children with HCD usually have far better speech perception outcomes than those with BAN. Surprisingly, HCD children had better speech perception outcomes than the control group.

INTRODUCTION

Auditory neuropathy (AN) is a recently recognised phenomenon characterised by varying degrees of sensorineural hearing loss which performs poorly using conventional hearing aids. AN has been associated with an absent or severely abnormal ABR and normal otoacoustic emission (OAE)[1]. This led to the belief that the cochlear function was relatively normal and that the dysfunction primarily affected the neural auditory pathway. Unfortunately this model failed to account for the unexpectedly good performance of most of these ears using a cochlear implant[2-7]. This report looks at the differing outcomes of cochlear implantation in two subgroups of AN patients differentiated by preoperative electrical testing and proposes a new classification for AN.

PATIENTS AND METHODS

Audiological and electrophysiological data have been collected prospectively on all cochlear implant patients between 1984 and 2003. The medical and obstetric records provided details of intrauterine and perinatal complications.

All patients underwent detailed preoperative audiological, electrophysiological and radiological investigations. Majority of the patients were referred with otoacoustic emission (OAE) results. Preoperatively all patients underwent round window electrocochleography (RWEcochG), auditory brainstem responses (ABR), and some underwent transtympanic electric auditory brainstem responses (TTEABR). The results in a patient with normal hearing are shown in Figure 1. The surgical procedure has been consistent all through the series and all patients were under the care of the senior author (WPRG). During surgery, all implants were tested for integrity and using implant evoked electric auditory brainstem potentials (ImpEABR) and neural response telemetry (NRT) (Figure 4,6,8).

Only patients who had full insertion of 22 electrodes of the Nucleus 24 device were included in the study. Data was collected prospectively till March 2003 using the Medelec Sensor until September 1998 and afterwards using the Medelec Synergy® Evoked Response system. The Nucleus® N R T software (Cochlear Corporation, Australia) was used to generate the ImpEABR via the Nucleus 24 device. A current level of 228 implant units with a pulse width of 25 microseconds was used in all recordings. The traces from channel 22 of the Nucleus 24 device

were compared. Absolute latencies and the morphology of the waves were used for analysing the data. Three different absolute latencies were measured along each trace i.e. eII, eIII and eV. The morphology of eV was used for the study.

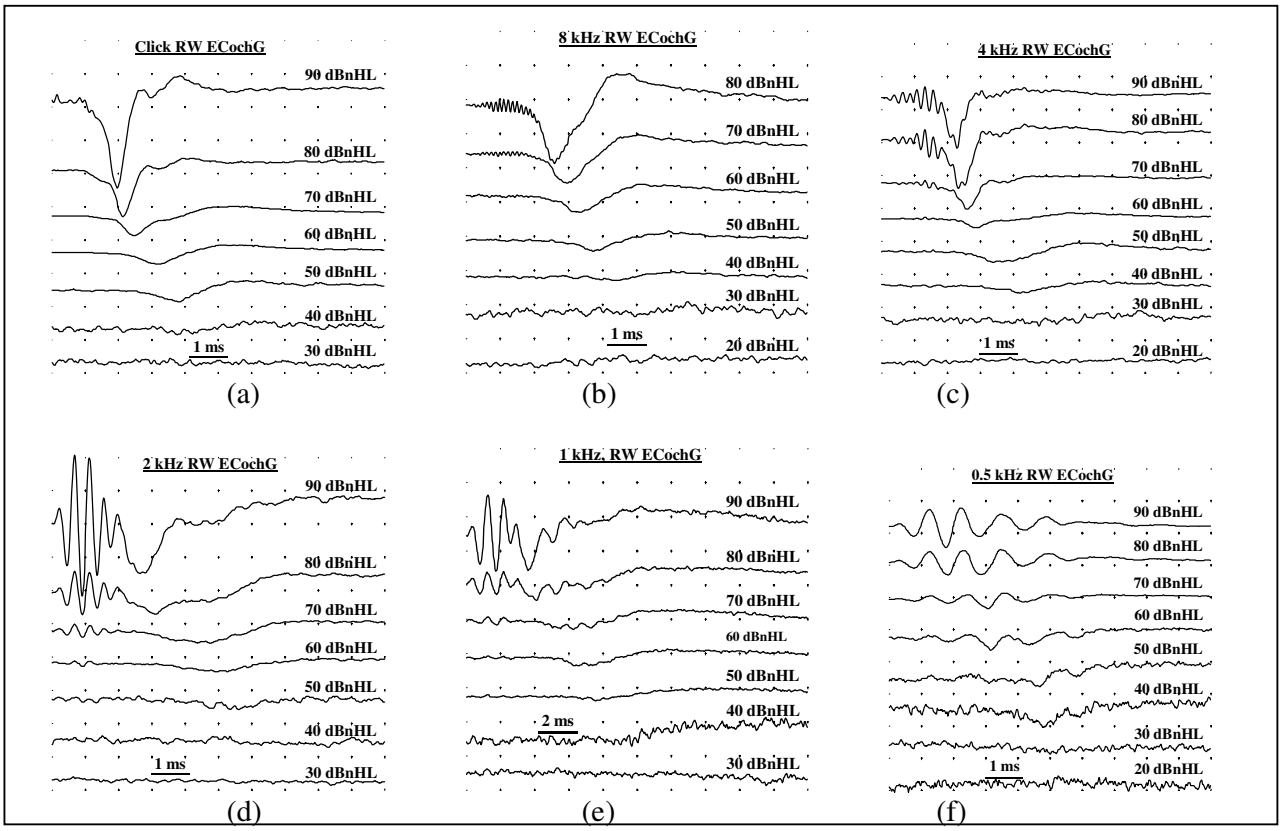

Figure 1. Electrophysiological test results in a hearing patient (a-f) Electrocochleography.

The study group

A working classification based on the electrophysiological tests was used for the study (Table 1). This divided the patients into three categories according to the possible site of lesion:

(i) Hair cell desynchrony (HCD)

(ii) Brainstem auditory neuropathy (BAN)

(iii) Cortical auditory neuropathy (CAN)

The HCD category is characterised by the presence APP on RWEcochG[8] often associated with OAE, absent or abnormal ABR, and normal TTEABR and Imp EABR. The BAN category had absent or delayed waveforms on TTEABR and ImpEABR.

Table 1. Proposed Classification of AN

Subgroup	Site of lesion	Diagnostic findings	Speech Scores	Prognosis for CI
Hair Cell Desynchrony *(HCD)*	Hair Cell Hair Cell – Auditory Nerve Synapse	APP in RWEcochG ±OAE	6.25 (SD: 1.3) *(n=42)*	Good
Brainstem Auditory Neuropathy *(BAN ±HCD)*	Auditory Nerve Afferent – Efferent Connections	Abnormal TTEABR and ImpEABR	2.5 (SD: 2.02) *(n=23)*	Poor

Notes: APP abnormal positive potentials; RWEcochG round window elctrocochleography; OAE otoacoustic emission; TTEABR transtympanic electric auditory brainstem response; ImpEABR implant evoked auditory brainstem response; SD standard deviation

The control (non HCD or BAN) group

The control group comprised of 70 deaf ears in which HCD and BAN was excluded by electrical testing. The children in this group were profoundly deaf from birth without any history of intrauterine or perinatal complications. There was no cochlear abnormality on CT, and no brain abnormality was seen on MRI. Apart from the hearing loss, there were no other neurological problems. Pre-operatively, these ears had no APP on RW EcochG and appropriate ABR recordings for their degree of deafness. All children received a Nucleus (Cochlear Corporation, Australia) multi channel cochlear implant. Intraoperatively all these ears were judged to have well-formed ImpEABR.

One year after surgery, the speech perception scores using the Melbourne Categories[10] were obtained (Table 2).

Table 2. Melbourne Speech Perception Categories

Category	Description
1	Detection of speech sounds only
2	Discrimination of suprasegmental aspects of speech in addition to 1
3	Discrimination and recognition of vowels in addition to 1 and 2
4	Discrimination and recognition of consonants in addition to 1-3
5	Minimal open set speech perception in addition to 1-4
6	Open set speech perception (>20% phoneme scores for PBK words)
7	Good open set speech perception (>50% phoneme score for PBK words)

All data was statistically evaluated using standard statistical package (SPSS for Windows® 1998). Mann-Whitney U Test for non-parametric data was used to test for significant difference. Significance level $P<0.005$ (two tailed).

RESULTS

The control (non HCD, BAN) group

A control group was used to evaluate the effects of age, development and maturation on the electrophysiological parameters. The mean age of this group was 29 months (range 5-60 months, SD 13.8 months). The ImpEABR waves shorten in latency with maturation and stabilise to adult values around twelve months of age. With maturation the eV waveform develops a definite discernible peak (Figure 2). The mean latencies of eII and eV after 12 months are 1.4msec and 4msec respectively. (Table 3)

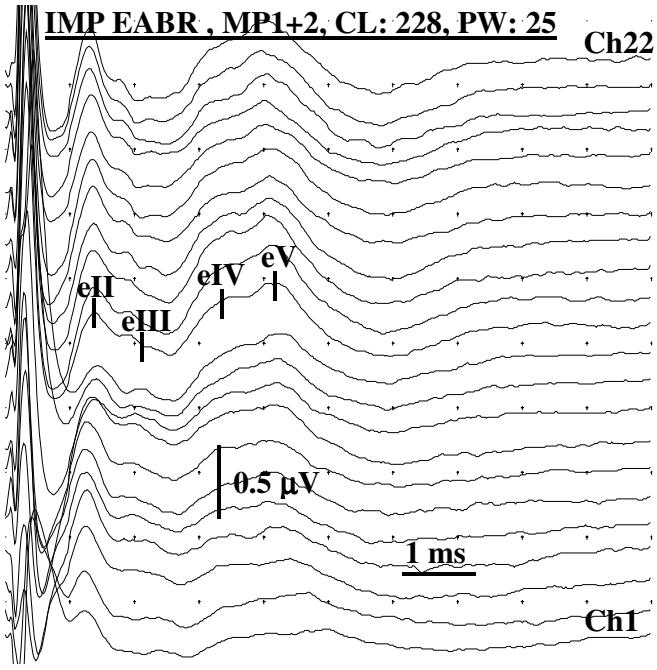

Figure 2. Implant evoked electrical auditory brainstem response in the control (non HCD, non BAN) group.

Table 3 Control (Non HCD/BAN) Group ImpEABP Values

Age Range at implant (testing) months	Number of patients	Mean age of group in months (SD)	Absolute Latencies IIe (ms)	Absolute Latencies IIIe (ms)	Absolute Latencies Ve (ms)
3 – 6 m	2	5.5 m	1.83	2.37	5.02
7 – 9 m	2	8 m	1.63	2.23	4.63
10 – 12 m	4	11m	1.45	2.11	4.05
13 – 24 m	22	18.95m (2.9)	1.41	2.02	4.15
25 – 36 m	20	28.68m (3.7)	1.49	2.08	4.09
37 – 60 m	20	47.21m (6.7)	1.39	2.06	4.02
N = 70; Mean age of group 29 months Range: 5-60 months; SD : 13.86months					

The study group

65 (8.46%) children were diagnosed with auditory neuropathy out of the 402 children who underwent cochlear implantation between 1984 and 2003. 42 (64.6%) of the 65 had hair cell desynchrony (HCD), 9 (13.9%) had brainstem auditory neuropathy (BAN). 14 (21.5%) had both HCD and BAN.

Predisposing factors

24 (36.9%) of the 65 children had been born prematurely. The mean gestational age at birth of the group was 36.17months (Range 23 weeks – 42 weeks; SD 5.73 weeks). Of these 16 (24.6%) had a history of neonatal jaundice and 16 (24.6%) had experienced episodes hypoxia at birth of varying degree and duration. 5 (7.6%) had both neonatal jaundice and hypoxia. (Table 4).

Table 4. Summary of Patients' Outcomes (Study Group)

Total tested	402	
Total diagnosed AN	65 (16.2%)	
Subgroups		
HCD	42 (64.6%)	
BAN	9 (13.9%)	
HCD + BAN	14 (21.5%)	
		n=65
Associated problems		
Prematurity	24 (36.9%)	
Mean gestational age	36.17weeks	
Hyperbilirubinaemia	16 (24.6%)	
Neonatal hypoxia	16 (24.6%)	
Jaundice + Hypoxia	5 (7.6%)	

Notes: HCD hair cell desynchrony, BAN brainstem auditory neuropathy

Many of the children had passed OAE screening tests and sometimes their parents had been falsely reassured about their hearing status. The average age at the time of detection of deafness was 11.67 months (Range 1-36 months; SD 10.60 months).

Test results and intervention

The mean age at cochlear implantation for this group was 43.6 months (range 12–120 months, SD 25.17months).

Intra-operative Imp EABR showed well formed and easily identifiable waveforms in HCD (Figure 3). NRT responses tended to be good in this group (Figure 4). The Imp EABR waveform in the BAN (n=9) were poorly formed and often no clear waveforms were identifiable (Figure 5) as was the case with NRT responses (Figure 6). The group with both HCD and BAN (n=14) also had poorly formed or absent ImpEABR.(Figure 7). NRT responses were highly variable and unpredictable (Figure 8).

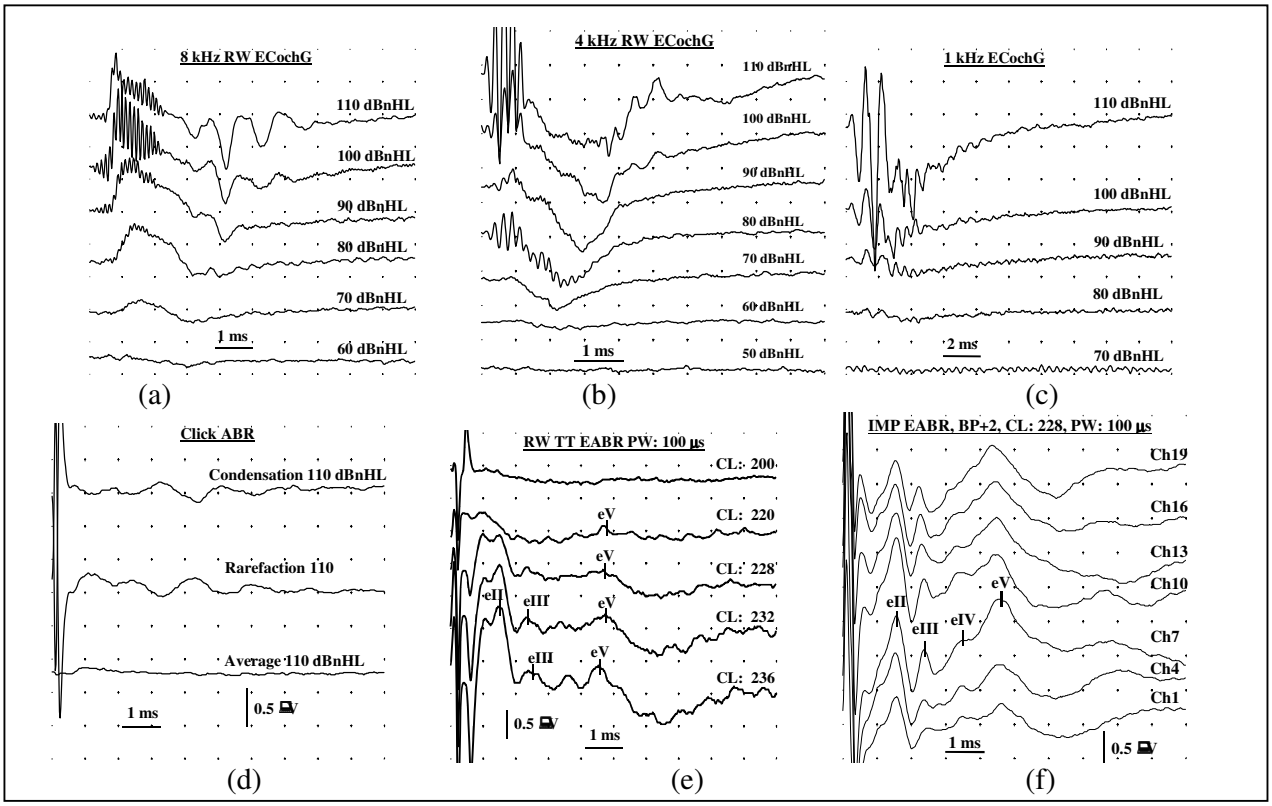

Figure 3. Electrophysiological test results in hair cell desynchrony. (a-c) Electro-cochleography, (d) Auditory brainstem response, (e) Trans-tympanic electrical auditory brainstem response, (f) Implant evoked electrical auditory brainstem response.

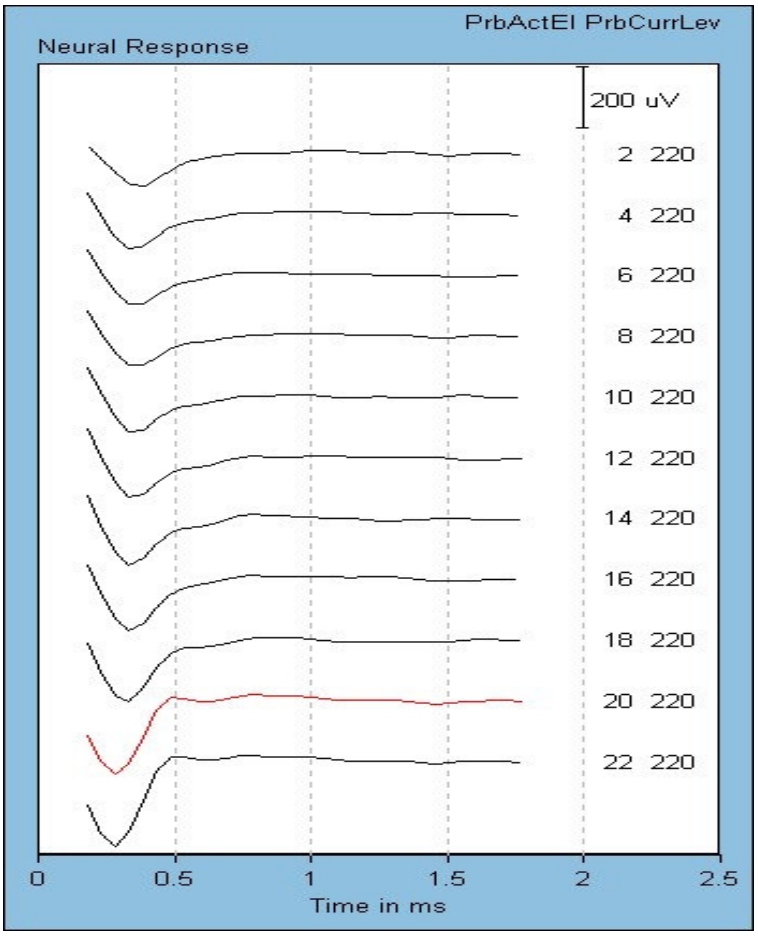

Figure 4. Neural Response Telemetry results in controls and hair cell desynchrony.

Outcome of cochlear implantation

All 65 patients in the study group underwent cochlear implantation. Speech perception scores at 12 months were obtained. The mean speech perception scores in the HCD group was 6.26 (SD 1.32) while that in the BAN group it was 2.57 (SD 2.03) (Melbourne categories). The mean speech perception score in the control (non HCD and BAN) group was 4.66 (SD 2.48). It is interesting to note that the speech perception outcome in the HCD group was even better than the control group (two tailed p value <0.005, Mann Whitney U test).

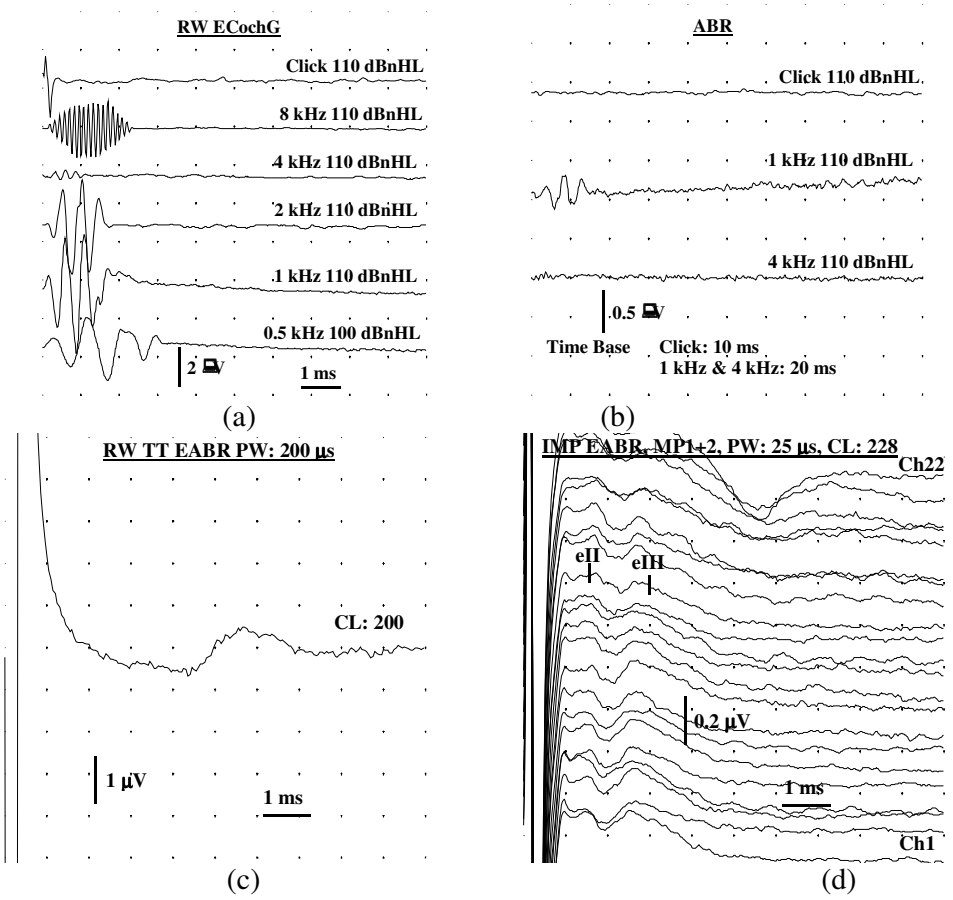

Figure 5. Electrophysiological test results in brainstem auditory neuropathy.
(a) Electrocochleography, (b) Auditory brainstem response, (c) Trans-tympanic electrical auditory brainstem response, (d) Implant evoked electrical auditory brainstem response.

DISCUSSION

Classification and Tests

The term “auditory neuropathy” has been coined to describe those ears with a hearing loss in which OAE are present but the ABR are absent[11]. We consider that this concept is too broad diagnostically and provides little information for prognostic decision making. We propose the term Hair Cell Desynchrony (HCD) when the problem only involves the hair cells and Brainstem Auditory Neuropathy (BAN) when the neural pathway is affected. A third group termed Central Auditory Neuropathy (CAN) is proposed to describe central cortical deafness (Table 1).

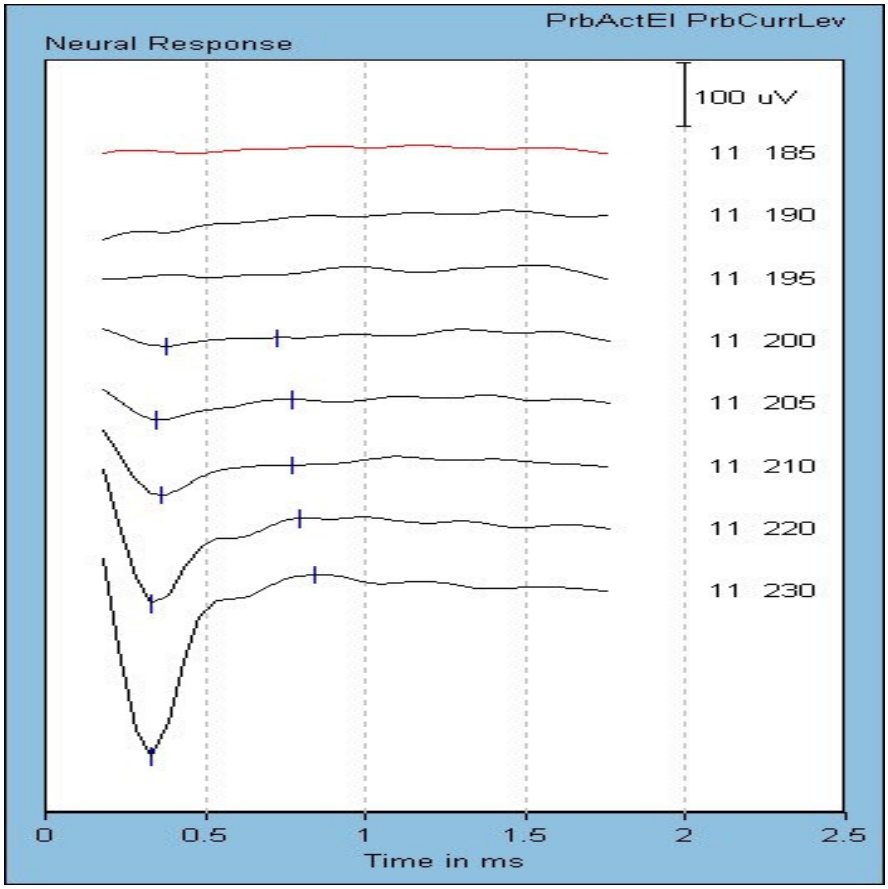

Figure 6. Neural Response Telemetry results in brainstem auditory neuropathy

Hair cell desynchrony (HCD)

The presence of OAE and the absence of ABR can be explained by the presence of outer hair cells (OHC) when there is a significant loss of inner hair cells (IHC). OAE are primarily generated by OHC, while the IHC are responsible for the afferent neural signal.

Studies performed in chinchillas[12] with carboplatin (an anticancer drug from the same group as cisplatin) have shown an extensive loss of IHC while the OHC remain intact. In response to an acoustic stimulus the preserved outer hair cells produce both an electrical signal recorded as the cochlear microphonic (CM), and an acoustic signal recorded as the OAE. Despite the CM and OAE being present, these animals have absent ABR and a profound hearing loss. The Bronx waltzer mouse is a mutant mouse in which IHC are absent and OHC are present. These mice will also provide a large CM response and OAE in the absence of hearing and ABR[13]. The Beethoven mouse is a similar model for this study[14]. Chronic cochlear hypoxia was achieved in these animals and produced swelling of IHC but

the OHC appeared normal[15]. This is of particular note as many premature infants are reported to suffer periods of hypoxia[9].

In the above situations both CM and OAE may be recorded from the cochlea, yet the animal has a hearing loss and no ABR. This can explain why a few infants pass OAE screening tests when they have a profound hearing loss on ABR testing.

Round window electrocochleography (RW EcochG) measures cochlear function and abnormal positive potentials (APP) have been described in ears affected by HCD[16]. When initially described nearly 30 years ago APP were thought to be due to desynchrony of the auditory nerve[17]. It is now believed that APP result from the activity of persistent OHC, in the presence of inactive or absent IHC. These APP appear to be large cochlear microphonics (CM) with an asymmetric output displayed as a positive summating potential (SP). When the APP are large, OAE are usually present[2].

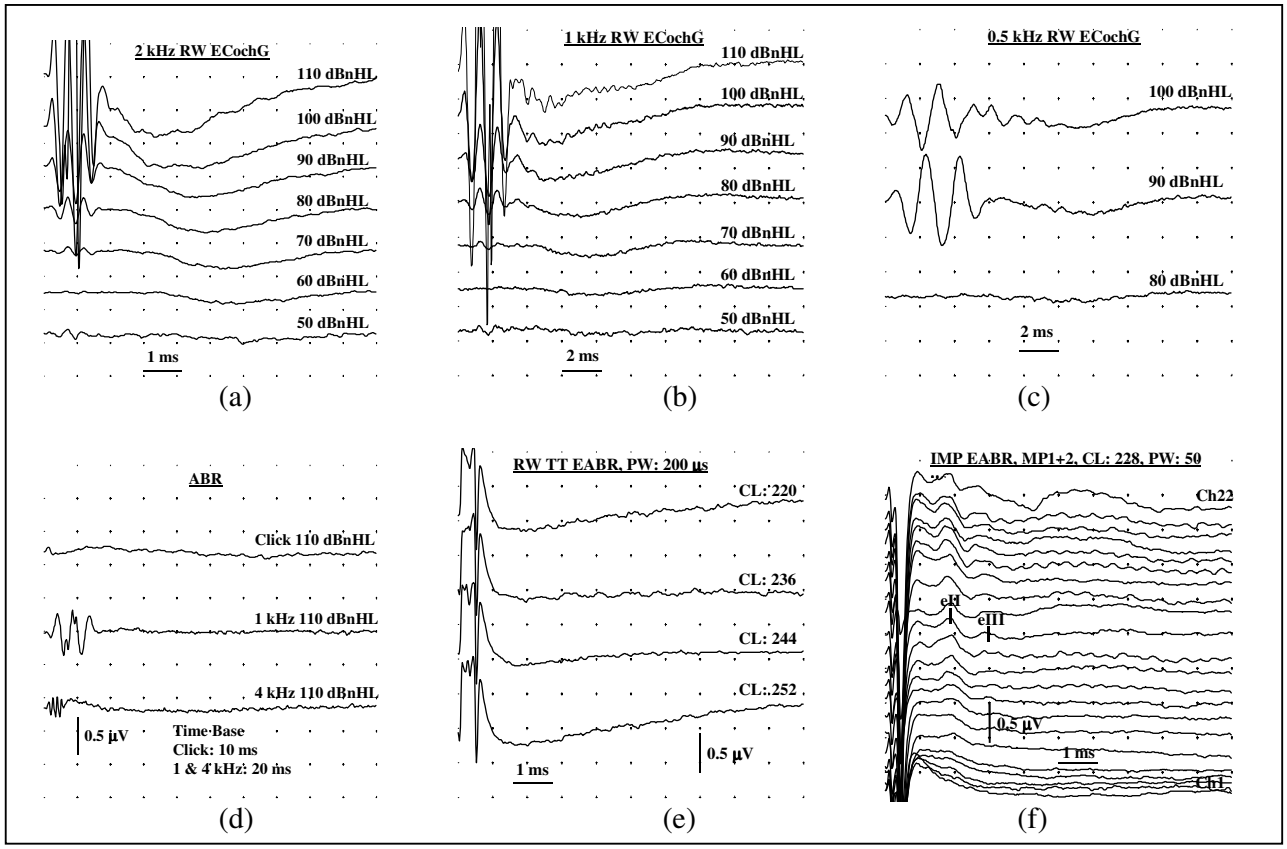

Figure 7. Electrophysiological test results in hair cell desynchrony and brainstem auditory neuropasthy (a-c) Electrocochleography, (d) Auditory brainstem response, (e) Trans-tympanic electrical auditory brainstem response, (f) Implant evoked electrical auditory brainstem response.

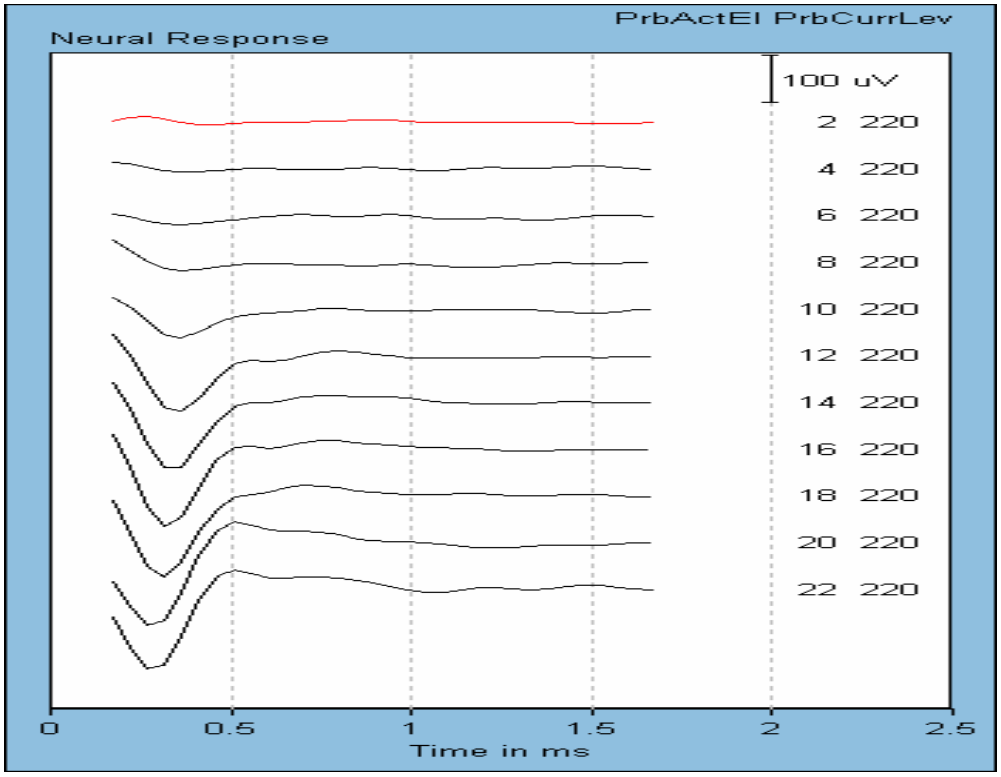

Figure 8. Neural Response Telemetry results in hair cell desynchrony and brainstem auditory neuropasthy

Brainstem auditory neuropathy (BAN)

Acoustically evoked auditory brainstem potentials (ABR) can be used to distinguish the site of a lesion within the auditory nerve or brainstem[18]. Similarly, we propose that electrically evoked auditory brainstem responses (EABR) can provides information on the site of brainstem auditory lesions. A control group of age matched deaf ears which were believed not to have HCD or BAN were evaluated to provide a diagnostic template.

The control (non HCD and BAN) group

This group has been age matched and selected as close to the study cohort as practicable. All ears that have suffered a profound hearing loss will have some loss of spiral ganglion cells and other neural structures, but it was believed that the amount of auditory neuropathy was insufficient to adversely affect the use of a cochlear implant.

Habilitation of children affected by AN and the value of the HCD/BAN classification

The proposed classification has important prognostic value when auditory habilitation is considered. The use of conventional hearing aids in AN is debatable. In one study[19] only 17% of hearing aid users derived marginal benefit and even this was insufficient for development of speech and language.

Cochlear implants have a better outcome in some of these patients, but not all children with auditory neuropathy do well with cochlear implants. A cochlear implant restores afferent neural function in the HCD group much better than in the BAN group. A possible explanation becomes obvious on examining ImpEABR traces, in the HCD group there are clearly formed ImpEABR while in the BAN group there is a complete lack of meaningful waveforms.

Interestingly the HCD group had a better outcome on speech perception testing than the control group. This may suggest that there is better brainstem auditory neural function in HCD than in a cohort of deaf ears which are not suspected as suffering from HCD or BAN. In all deaf ears some loss of spiral ganglion cells and neural structures occurs. Perhaps this is less marked in many HCD ears.

It is hoped that pre-operative testing involving RW ECochG and TT EABR will help to distinguish these children before implant surgery is undertaken so that appropriate counselling of the family is available.

CONCLUSION

A new classification of auditory neuropathy has been proposed based on possible site of lesion as shown by electrophysiological tests. Children with HCD do better with cochlear implants than those with BAN. Implant teams and parents of prospective implant candidates should be aware of this difference in outcome at the time of pre-surgery evaluation.

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CHAPTER 5

UNUSUAL CASES

5.1

THE SCHEIBE COCHLEA DEFORMITY WITH MACROCEPHALY: A CASE FOR SINGLE CHANNEL IMPLANTATION

J. Ray
R.F. Gray
Z.H. Vanat
J.H. Begg

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ABSTRACT

An 11 year old congenitally deaf child with bilateral primitive common cavity (Scheibe type) cochleosaccular dysplasia and benign familial macrocephaly was implanted with an extracochlear single channel device with an ear level speech processor. This paper describes the assessment, findings, dilemmas in decision making, surgical procedure and the favourable outcome after implanting. The relevant literature has been reviewed and our case is presented for the unusual combination of features.

INTRODUCTION

Congenital anomalies of cochlear morphology are well known causes of profound sensorineural deafness. An 11 year old boy from a farming family with Scheibe type cochleo-saccular dysplasia was assessed and found to be profoundly deaf in both ears beyond the reach of hearing aids but not suitable for a multichannel cochlear implant because of 1. Lack of tonotopic representation and 2. Middle ear sepsis with drum perforation and discharge. He was tried with a vibrotactile device and later implanted with an extracochlear single channel device to augment his lip reading skills and provide awareness of environmental sounds.

The unusual association of sensorineural deafness due to cochlear dysmorphology with hypotonia and autosomal dominant benign familial macrocephaly aroused our interest.

CASE REPORT

The child JH was first suspected to be deaf by the childminder at one year age when there was no response to a door slamming shut. After failing three health visitor distraction tests he was referred to the local ENT department where bilateral post aural hearing aids were fitted at the age of 18 months.

Reassessment at 2 years showed very little language development (only single indistinct words). Tympanograms were flat and responses on electrocochleography were equivocal at 95dBA. At this stage grommets were fitted. These were later replaced by T-tubes. Repeated ear infections added to the problems and they

were removed after 4 years leaving small perforations which were still discharging and troublesome when assessment for cochlear implant was in progress. These eventually closed over.

Associated features

JH was the elder child of parents with normal hearing and without any family history of deafness. He was a normal term birth but was thought to have infantile hypotonia. It was also noted that the head circumference was 1 standard deviation (SD) greater than the mean for his age and this continued to increase steadily until it was greater than 4SD of the mean at 4 years age. A CT scan showed enlarged lateral ventricles. A review of the family tree and CT scanning of the parents (when similar findings were seen in the father) by the Medical Genetics Department revealed the existence of a rare disorder – benign familial macrocephaly. This was inherited as an autosomal dominant disorder from the father's side. The head growth stopped at 4SD from the mean head circumference (findings in his father were the same).

Interestingly, from the mother's side the child had inherited an idiopathic ptosis on the right eye which had run through at least 3 generations. Other somatic abnormalities seen in the child were increased distance between the medial canthus with downslanting palpebral fissures, an upturned nose, broadened forehead and a flat midface. His motor skills were rather delayed and he learned to walk only at the age of 3 years. Thereafter he made steady progress and by 7 years he was riding a bicycle without stabilisers.

Initial referral

JH was referred to our assessment clinic at the age of 9 years for consideration of cochlear implantation. Ultra high resolution CT scanning showed " widened and dysplastic basal turn of the cochlea without any round window or promontory which in turn was separated from the internal auditory meatus (IAM) by a thin bony septum (Schiebe type cochleosaccular dyslasia). Semicircular canals also appeared to be hypoplastic. The carotid canals were prominent bilaterally and only just separated from the middle ear cavity by a thin bony septum" (Figure 1). Audiological results were as follows:

Unaided Pure Tone Audiogram

Frequency(Hz)	250	500	1K	2K	4K
dBHL Left Ear	110	95	120	115	NR
dBHL RightEar	90	95	115	115	NR

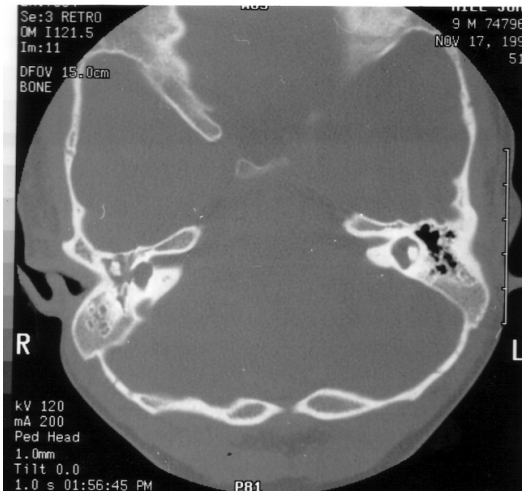

Figure 1. CT scan (1mm axial sections) of the patient showing bilateral cochleosaccular dysplasia and internal auditory meatus separated from the middle ear by a thin bony partition.

An intracochlear implant was not thought to be advisable because of the risks of a CSF leak[1] compounded by chronic middle ear sepsis. Furthermore it was also felt that he would derive very limited benefit from a multichannel cochlear implant as it was not expected that those cochlear structures that had developed would be organised in the usual tonotopic way. The child, JH was fitted with a TRILL vibro-tactile aid.

Reassessment

At one year JH was reassessed and found not to be using the TRILL at all as he found the waist worn box attached to wristbands very cumbersome. Ear infections had settled by then and the perforations had closed over with granular myringitis which could be improved by topical antibiotic cream. Promontory Stimulation and Auditory Brainstem Response tests showed a few inconsistent responses. Detailed 0.7mm section T2 weighted fast spin echo MR scans were done to demonstrate the anatomical integrity of the auditory nerve pathway from the internal auditory meatus (Figure 2). This showed “bilateral primitive common cavity lesions on both sides but no definable cochlea. However there were several nerves going through the IAMs”.

Further testing was carried out to confirm the functional value of his communication. He was performing well on lip reading screens but was unable to detect any meaning from voice alone.

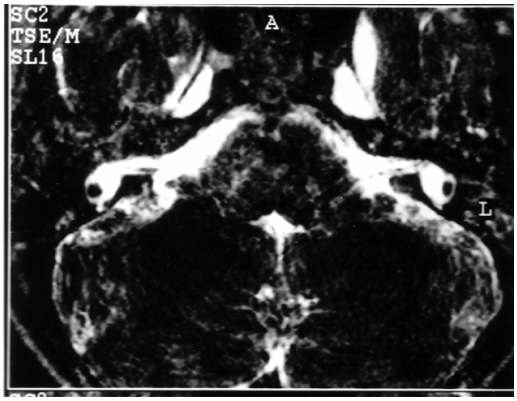

Figure 2. Axial 0.7mm T2 weighted MR image of IAMs showing existence of neural structures in both IAMs and leading upto a primitive common cavity.

Device Selection

Experience with similar pathologies at other centres in Europe and North America was reviewed and it was decided to recommend a single channel extracochlear device. The parents were counselled. They were sure that a body worn device would be rejected and that JH would only take to an ear level device. The Med El Extracochlear implant with an ear level single channel speech processor (being the only ear level device available then) was chosen. The right ear was selected for implantation.

Surgery

Usual approach for a cochlear implant was used with no surprises. A high promontory was visualised but no identifiable round window or niche could be found. The stapes and the stapedius tendon were present and normal. The ball electrode of the single channel implant was sited in a shallow depression drilled out where the normal anatomical round window should have been. This was held in position with a small amount of ionomeric bone cement. The reference electrode was buried beneath the temporalis muscle and the receiver held in place with prolene ties. Electrode position was confirmed the following morning by post operative perorbital skull x-ray (Figure 3).

Switch-on

The device was switched on a month later and with further tuning comfort levels ranged from –10 to –7.1(dBV) with sensitivity at 2.5 and volume at 3. There were occasional non-auditory sensations during tuning sessions in the form of a discomfort in the throat, neck and ear. This was probably due to the leakage of

electrical current into the Glossopharyngeal nerve via the tympanic plexus on the promontory where the electrode was sited. This did not diminish his enthusiasm. At subsequent tuning sessions the low frequency levels were reduced to minimise non-auditory sensations.

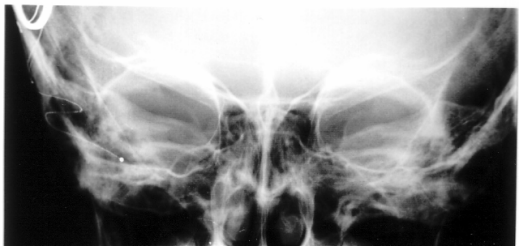

Figure 3. Post operative perorbital skull x-ray of the patient showing position of the single channel extracochlear ball electrode.

Post operative results

There have been several problems with reliability of the equipment which is not robust enough to cope with the lifestyle of a 10 year old boy. However the support from the manufacturers with multiple spare parts has been good. The relevant changes in audiological scores were as follows:

Functional Listening	Pre-implant	Post-implant
Syllable pattern (closed set of 12 words)		
Lip pattern and voice:	100%	80%
	0%	42%
Voice alone:		
Manchester Picture Test (voice alone)	0%	30%

He is a skilled lip reader and is beginning to gain additional information from the implant as is borne out by the scores.

DISCUSSION

Congenital sensorineural deafness

Jackler et al have proposed a classification of congenital malformations of the middle ear based on embryogenesis[2] in order to differentiate “true Mondini” deformities from the other varieties. The Mondini type inner ear dysplasia is

probably the commonest variety of cochlear dysmorphology causing sensorineural deafness seen in the clinical setting. Scheibe type malformations are less common. What makes this case of interest is its association with the rare condition of benign familial macrocephaly and infantile hypotonia. It is not clear if the deafness is a separate problem or an unusual manifestation of benign familial macrocephaly. The criteria for diagnosis of autosomal dominant hereditary hearing loss are (1) male to male inheritance pattern; (2) characteristic hearing loss demonstrated audiometrically over three successive generations; and (3) exclusion of other causes of deafness[3]. None of these features were seen in JH and so it can be assumed that this was a chance association and the risk of recurrence of deafness is low but indeterminate.

Benign familial macrocephaly

Benign familial macrocephaly is a rare autosomal dominant disorder with incomplete penetrance[4]. The features are same as seen in JH and CT scans show ventricular dilation in most cases[5]. The risk of recurrence is 1 in 2.

Related syndromes

Further literature search shows coexistence of macrocephaly and hypotonia has been noted in benign familial macrocephaly, Ruvaclaba-Myhre-Smith syndrome and Bannayan-Zonana syndrome[6]. It has been postulated that the above three disorders may be represented in the same gene locus.The ptosis appears to have been idiopathic and probably an incidental association.

Vibrotactile device

Varying degrees of inner ear aplasias have been described. There are several instances of leakage of cerebrospinal fluid and meningitis either spontaneously[1,7] or after cochlear implantation[8]. In our patient this risk was compounded by the fact that the child was suffering from recurrent middle ear infections. So at the initial assessment a vibrotactile device (TRILL) was tried out. Although this gives prosodic information about the rhythm and length of sounds, the equipment is cumbersome (especially for children) and was soon discarded.

Multichannel cochlear implant and risks

Multichannel cochlear implants have been inserted in cases with mild bilateral Mondini deformities[9]. But in severe cochleosaccular aplasia there is a definite risk of development of spontaneous cerebrospinal leak or recurrent attacks of meningitis because of wide direct communication between the middle ear and the

subarachnoid space in the IAM. Therefore an extracochlear single channel device with an ear level speech processor was considered the only feasible option.

MRI scan

It was also necessary to assess the anatomical integrity of the auditory pathway medial to the aplastic cochlea. It has been seen at autopsy that an intact eighth nerve may be present despite a negligible end organ in a single tube cochlea[10]. With the newer MRI techniques it was possible to demonstrate this in our patient.

Middle ear abnormalities

Anomalies of the round window are also common in subjects with dysplastic inner ears as this develops from the otic capsule and not the branchial arches[11]. This observation was also corroborated by our findings at surgery.

Single channel cochlear implant

Unlike a vibrotactile device a single channel cochlear implant provides information on pitch changes upto 300Hz in addition to prosodic information and this would allow improved speech discrimination and environmental awareness[12]. One of the key factors considered before implanting this child was the need for sound awareness as a safety issue in a farm environment. Not only can he now hear a tractor engine but can reliably turn his head when his name is called.

Current situation

His spontaneous speech contains some vowel sounds and he is beginning to use t,d,k and g. He now has some awareness of his own voice and is beginning to monitor final consonants. Speech intelligibility has improved since the implant was activated. He is able to make himself partly understood to sympathetic strangers. He has won the annual award at his residential school for the deaf for the most improved speech.

CONCLUSION

Severe sensorineural hearing loss due to severe cochlear dysmorphology can be ameliorated by single channel extracochlear devices without the risks of cerebro-spinal fluid leak or iatrogenic meningitis.

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5.2

COCHLEAR IMPLANT FAILURE DUE TO UNEXPECTED ABSENT EIGHTH NERVE – A CAUTIONARY TALE

R.F. Gray
J. Ray
D. Baguley
Z. Vanat
J. Begg
P.D. Phelps

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ABSTRACT

We present a case of bilateral absence of the eighth cranial nerve in the internal auditory meatus. This caused total failure of responses after cochlear implantation in a 6 year old patient with congenital deafness. Pre operative MR imaging is important to show not only the anatomy of the middle and inner ears but also the structures in the IAM.

INTRODUCTION

Cochlear implantation has now become an accepted method of managing profound sensorineural deafness both congenital and acquired[1] in cases where there is no medical, surgical or developmental contraindication. The surgical technique is safe and reasonably straightforward with few complications in the hands of an experienced otologist[2]. The East of England Cochlear Implant Programme at Addenbrooke's Hospital has been in existence for eleven 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 ten months. This was confirmed at the age of fifteen 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 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[3]. 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 70dB (at 250Hz) and 90dB (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 CT scans of temporal bones revealed patent cochlear ducts on both sides. Cochlear 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 5 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. Postoperative 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 6 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 1.

Table 1. 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

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 would say “bye-bye”, “please”, “dada” but was no longer saying “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.

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

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

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 70dB.

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 EXPLANTATION

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 1) 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 structures in the IAM[4]. Detailed views of the IAM and the cerebello-pontine angle (CPA) were required and this was to be 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.

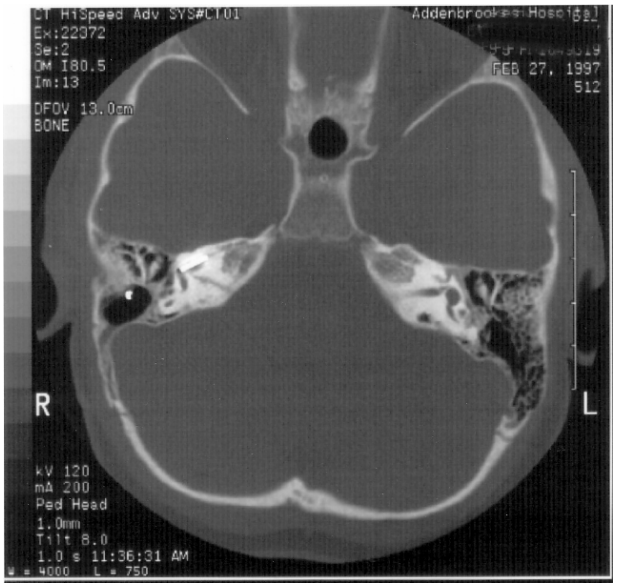

Figure 1. Ultra high definition CT scan (axial view) showing narrowing of IAMs on both sides (Patient).

MRI SCAN

A detailed 3 plane MRI scan of the petrous temporal bones was requested under a general anaesthetic. 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 (Figures 2 and 3).

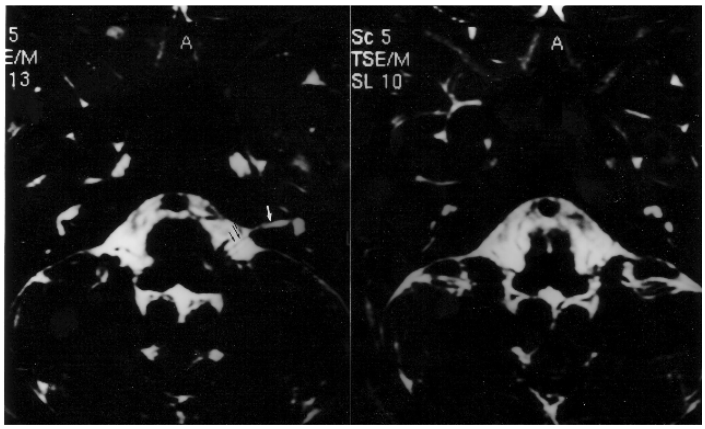

Figure 2. Axial 0.7mm T2 weighted MR image of IAM showing absence of nerve bundle on the right side and a hypoplastic nerve on the left side (arrow). (Patient).

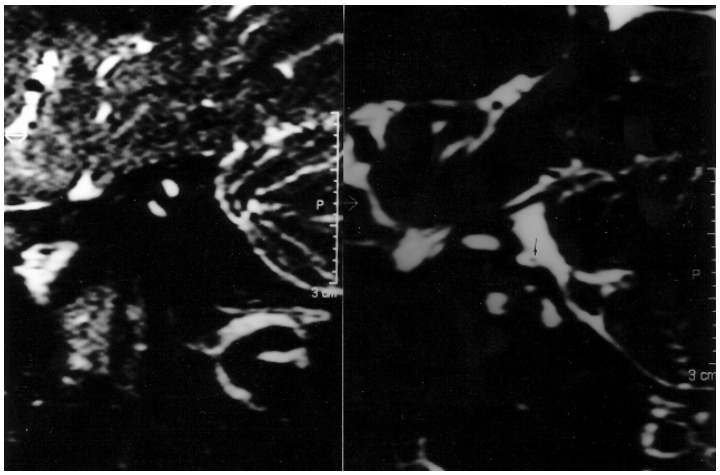

Figure 3. Saggital 0.7mm T2 weighted image of IAM showing abnormality of structures. Arrow shows the hypoplastic seventh nerve. (Patient).

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 childs 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 vestigeal 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 Computerised Tomography (CT) scanning[5]. 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[6] and this morphological anomaly has been associated with 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[7].

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[8] 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 3 cases the IAM was of normal morphology yet the cochlear branch of the Cochleo-vestibular nerve was absent or hypoplastic. CT would thus miss 3 out of 5 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 4 and 5). 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 3 dimensional resolutions.

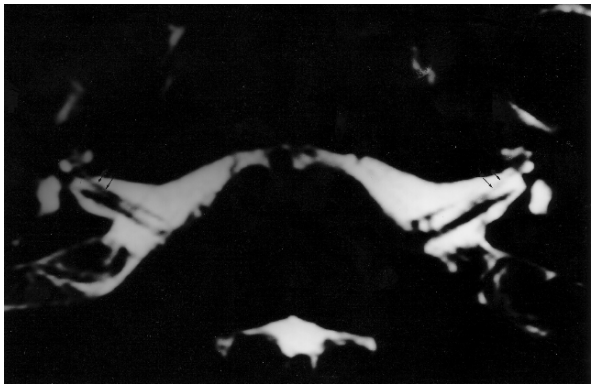

Figure 4. Axial 0.7mm T2 weighted MR image of IAM showing normal structures (arrow) (Normal subject)

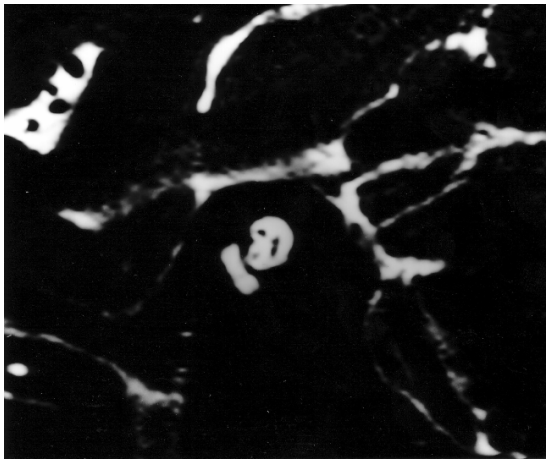

Figure 5. Saggital 0.7mm T2 weighted MR image of IAM showing normal structures.VII=Facialnerve;CN=Cochlear nerve; C=Cochlea; SVN=Superior vestibular nerve; IVN=Inferior vestibular nerve. (Normal subject).

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 though 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[9] 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

1. 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 of neural structures passing through it and should prompt further imaging of the region.

2. 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.

3. 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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CHAPTER 6

SUMMARY AND CONCLUSIONS

Over the last half a century cochlear implantation has evolved from being a mere quirky experiment through being a controversial medical device to a widely accepted otological intervention. The journey has not always been smooth but the doubters and the sceptics have only helped to refine all aspects of this technology.

Initially only post lingually deafened adults were selected. The indications have been steadily widened to include a broader candidature. This means that much younger patients can be implanted to take advantage of the neural plasticity. This has resulted in the satisfactory outcomes that are witnessed in these children many of whom achieve very high open set speech perception and attend mainstream schools. Many congenital inner ear anomalies are also considered suitable for implantation thus challenging traditionally held misgivings about surgical intervention in these patients.

The introductory section takes us through the background of cochlear implantation, the current standard of practice and the future. This work was carried out at three large cochlear implant programmes (Cambridge, Birmingham and Sydney) and their backgrounds have been described. Data collection, management and analysis have been addressed in the chapter on databases. This provides a guideline on how to construct a simple database and its basic structure.

The basics of cochlear implant (CI) technology and the surgical steps involved have been described. Candidacy and psychosocial issues are closely related to outcome and cost benefit analysis. Recent concerns about the possible risk of meningitis in CI patients have also been discussed.

CHAPTER 2

Although cochlear implantation is a relatively safe procedure it carries with it its share of surgical complications.

Section 2.1 looks at a very large series of 844 patients in the Sydney CI programme. Several key features about cochlear reimplantation surgery have been addressed. Device failure, wound and flap problems are the commonest causes of explantation and reimplantation. This also shows how a smaller incision and flap reduces the incidence of wound and flap problems.

Section 2.2 adopts a clinical audit approach and compares the medical and surgical complications in the second hundred CI patients to the first hundred in Birmingham. Device failure, wound and flap problems are again the commonest complications. This again reiterates how the use of a modified and smaller incision reduces wound and flap problems.

There are many instances where an implanted device may need removal and / or replacement.

Section 2.3 looks at the series from the Cambridge group where the commonest causes for explantation were for upgrading (3%) and device failure (3%). The importance of flap design and handling and the importance of patient assessment have also been stressed.

Section 2.4 looks at the series from the Birmingham group. The main reasons for explantation, reimplantation include device failure (2.2%), wound and flap problems. The latter were associated with larger incisions and wider scalp mobilisation. The strategies needed in each situation for a satisfactory outcome are discussed.

Section 2.5. In the initial stages of cochlear implantation only healthy and clean mastoid cavities were implanted and the history of chronic suppurative otitis media was considered a contraindication for cochlear implantation. Several techniques have been described to render chronically discharging ears dry. However not all techniques are suitable for subsequent cochlear implantation. The Cambridge programme adopted the technique of obliterating discharging mastoid cavities with autologous abdominal fat prior to cochlear implantation. This chapter looks at a technique and interim results at five years when 94.1% of these obliterated and implanted ears were dry.

CHAPTER 3

Very little has been published about non or limited use of CIs, although this is widely known anecdotally. This chapter explores the issues of unrealistic expectation, adverse effect of co morbidities and the influence of deaf culture and peer pressure resulting in non or limited use of implants. Different patterns of

patient characteristics were identified. The commonest cause for non use in the younger patients was poor performance and peer pressure. Depression, tinnitus and concomitant neurological problems played an important part in the older patients.

CHAPTER 4

Cochlear implant provides an unsurpassed tool to study electrophysiology of the auditory pathway in vivo. The Sydney programme has collected a large volume of electrophysiological data from its cochlear implant patients and this proves a useful starting point for researching the auditory system.

Traditionally it has been believed that prior exposure to auditory acoustic stimulus is a necessary prerequisite for the maturation of the auditory neural pathway. However recent work suggests that the auditory pathway continues to mature irrespective of auditory stimulation. In *Section 4.1* the latencies of implant evoked electrical brainstem responses (ImpEABR) have been used to compare auditory maturation in congenitally deafened (without any useful hearing prior to implantation) and later deafened patients (with normal hearing prior to sudden deafness). There seems to be no difference in the two groups and the latencies continue to decrease and the traces acquire adult waveforms independent of any acoustic or electrical input.

Section 4.2 deals with the recently recognised concept of Auditory Neuropathy. Typically ears affected by auditory neuropathy have been predicted to perform poorly with cochlear implantation. However electrophysiological tests show that the term has actually been used to describe different sets of patients with diverse audiological conditions. Using data from otoacoustic emission (OAE), round window electrocochleography (RWEcochG), auditory brainstem responses (ABR), transtympanic electrical auditory brainstem responses (TTEABR) and implant evoked electrical auditory brainstem responses (ImpEABR) a prognostic classification (based on the possible site of lesion) has been proposed. This divides patients into Hair Cell Desynchrony (HCD) and Brainstem Auditory Neuropathy (BAN) groups. Patients with HCD do much better with CI than those with BAN.

CHAPTER 5

This chapter looks at the challenges facing the cochlear implant teams with unusual cases.

Section 5.1 describes the dilemma faced in implanting a child with bilateral primitive common cavity (Schiebe type cochleosaccular dysplasia) and benign familial macrocephaly with infantile hypotonia (Ruvaclaba-Myhre-Smith syndrome).

Section 5.2 is a cautionary tale illustrating how one might be caught unawares in cases of unsuspected auditory tract abnormalities and emphasises the role of high quality imaging.

CONCLUSION

This thesis examines several important issues relating to CI and draws from the experience of three large and reputed CI programmes. The surgical section discusses the issue of complications, explantation and reimplantation. Device failure, wound and flap problems continue to be the main problems. Smaller incisions and minimal scalp mobilisation are crucial in reducing flap problems. A surgical technique using autologous abdominal fat to render discharging mastoid cavities safe for implantation is described. Reduced use or rejection of CIs remains an unpleasant truth. Poor performance, unrealistic expectation, adverse effect of co morbidities, influence of deaf culture and peer pressure result in non or partial use of implants. The section on electrophysiology illustrates the use of CIs as a powerful tool to study the auditory pathway in vivo. A prognostic classification for auditory neuropathy into Hair Cell Desynchrony and Brainstem Auditory Neuropathy has been proposed with a better CI outcome predicted in the former. Electrophysiolgical data in CI patients also suggests that the auditory pathway continues to mature in the early years of life independent of prior acoustic stimulation. The last section describes two unusual cases which can pose a challenge to a cochlear implant team.

CHAPTER 7

CURRICULUM VITAE

JAYDIP RAY was born in India on 2nd September 1965 and gained his basic medical qualification from University of Calcutta in 1989. He completed his Diploma in Otolaryngology in 1991. In 1994 he attained the first position in the Master of Surgery from the same university and was awarded the gold medal. His thesis was on the “Epidemiology of glue ear”.

In 1994 he was offered an attachment to the Guy’s and St. Thomas’ Hospital, London and moved to the UK. His higher surgical training in otorhinolaryngology has been in the West Midlands Rotation in the UK.

His sub speciality interest in Implantation Otology started as a research fellow in the cochlear implant unit in Cambridge between1997-98 with Mr. R.F. Gray. The main project was entitled “Imprinting of speech in sudden childhood deafness; role of neural plasticity and auditory memory” and was supported by the Cambridge Hearing Trust.

Another year (2001-2002) at the Otology Neurotology unit at the University Hospital Birmingham provided tremendous exposure to all varieties skull base surgery and implantation otology including middle ear implants and bone anchored hearing aids under the tutelage of Mr. D.W. Proops. The support of the Midland Adult Cochlear Implant Programme proved invaluable in continuing the research work into cochlear implants.

Mr. Ray was awarded the Graham Fraser Memorial Fellowship 2003 to undertake further clinical research into profound deafness and cochlear implantation with Professor WPR Gibson at Sydney University. This also provided him with a unique opportunity to visit the research facilities at Cochlear Corporation, Bionic Ear Institute and Melbourne University. He also spent some time with the Royal Australian Flying Doctor Service to experience the health problems in the Australian outbacks. He was also awarded the Journal of Laryngology and Otology Travelling Fellowship in 2003 to visit the House Ear Institute, Los Angeles and Stanford University, California. This visit also enabled him to see the surgery for the first penetrating auditory brainstem implant.

Mr. Ray has published extensively and presented regularly at both national and international meetings and has been the recipient of several awards. Notable amongst these are Certificate of Merit at the National Science Talent Search

Competition 1984 and the Politzer Prize for the *Best Clinical Paper* at the 23rd Meeting of the Politzer Society 2002.

The association with Nijmegen University started when he was awarded the Rotha Abraham Trust Scholarship 2001 to attend the 36th Post Graduate Ear Surgery Course in Nijmegen. This has culminated in the production of this thesis with the compilation of major clinical work from three large cochlear implant programmes. He is scheduled to complete training in October 2004 and is currently attached to the otolaryngology department of the University Hospital Birmingham.

CHAPTER 8

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