Paediatric cochlear implantation

In 1986 William House, one of the pioneers of cochlear implantation wrote, 'Cochlear implants have now reached a robust childhood'.1 This quotation is particularly apposite today with cochlear implantation being more widely offered to profoundly deaf children. More than 800 children worldwide have now received the Nucleus multichannel device, one of the most widely used of the implants currently available.

The concept of electrical stimulation of the auditory system dates back to Volta; the recent history of cochlear implantation has been summarised by Luxford and Brackman.2 It is really only in the last decade that cochlear implantation has moved from the experimental field into wider clinical application.

What is a cochlear implant?

In essence a cochlear implant is a very sophisticated (and somewhat expensive) hearing aid, bypassing the inner ear hair cell system and stimulating the ganglion cells of the VIIIth nerve directly.4 The basic components of any cochlear implant system comprise a microphone which is connected to an externally worn speech processor which filters speech and extracts formant information. These data are transmitted to the receiver/electrode package which is the implanted part.

An implant may be extracochlear, stimulating the nerve via an electrode positioned adjacent to the round window, or it may be intracochlear, the electrode(s) being inserted into the lumen of the cochlea. Implants may be single channel, such as the UCH/RNID device5 or they may be multichannel such as the Ineraid6 or Nucleus7 devices. The Ineraid uses a transcutaneous electrical connection, the Nucleus depending on electromagnetic induction for transmission of the signal to the implanted package. Intracochlear multichannel devices provide significantly greater benefit than single channel ones and are, therefore, the treatment of choice.6 8 9

Indications for paediatric cochlear implantation

Cochlear implantation is indicated for profound sensory (cochlear) hearing loss; it depends on the integrity of the cochlear nerve. In the UK the prevalence of severe or profound sensorineural deafness in children is between one and two per 1000.10 11 It is only a small proportion of such children that is likely to need cochlear implantation as the majority can be helped by suitable conventional hearing aid systems, coupled with appropriate habilitation or rehabilitation. While the detailed audiological criteria for implantation in children may vary in different centres, there appears a broad consensus that the child needs to have had an adequate trial of suitable conventional amplification. In the Federal Food and Drug Administration (FDA) programme administered in 24 centres in the USA, and in the Melbourne programme in Australia, the criteria include auditory thresholds of greater than 90 dB averaged over 500 Hz, 1 kHz and 2 kHz, aided auditory thresholds of 55 dB or greater, or speech perception scores of less than 20% using standard sentences prepared by the Central Institute for the Deaf, USA.

In the Nottingham programme our criteria include total loss of hearing with aided responses at thresholds greater than 80 dB across the frequency range from 500 Hz to 4 kHz. In the light of others' experience this latter criterion has been relaxed to aided thresholds of 70 dB, although to date no child has been implanted using that new criterion.

Much debate has centred around the question of whether to implant prelinguistically deaf children or only those who have had some experience of language. In the FDA trials 64% of those implanted were deafened at or before 24 months of age and are considered prelinguistic, with 43% being congenitally deaf.12 In Nottingham we have moved more cautiously: of a total of 20 children who have undergone this surgery, only two have had a congenital hearing loss although a further eight belong in the prelinguistic category. Encouraged by our early assessment of the progress of these two children, and in the light of results from other centres, we have now elected to include within our criteria congenitally deaf children under the age of 5 years.

Special needs of children

ASSESSMENT

Considerable skill is needed in evaluating not only the hearing of young children but also their spoken language and other communication skills.13 Conventional tests of spoken language may be used for children with functional language, while in the preverbal stage video analysis of a child with a known adult may be used,14 both before and after implantation. Behavioural audiometry results should be supported by those from evoked response audiometry.

As well as an auditory assessment, in our programme the child also undergoes general medical and otological evaluation, including radiological assessment using computed tomography. The child should be generally medically fit and there should be no active middle ear infection. A computed tomogram is an important element in the assessment process; in cases of deafness due to meningitis,
labyrinthitis ossificans (new bone growth within the lumen of the cochlea) may occur and prove a contraindication to implantation of an intracochlear device. Congenital inner ear malformation may be demonstrated, such as Mondini dysplasia or abnormalities of the internal auditory meatus.

Finally, in assessing a child for possible implantation, the expectations of the parents need to be taken into consideration; these should be realistic. Time must be allocated in the evaluation phase for counselling the parents; they need to be aware of the long road ahead if their child undergoes implantation. The rehabilitation period is a very long one. It is essential that appropriate rehabilitative support be available locally for the child and that the potential as well as the limitations of the implant should be known to the local teacher(s) of the deaf.

Surgery

Implantation in children poses special challenges. Preparation of the child is all important, and close involvement of the parents is essential. It is helpful to have shared photographs and books about children previously implanted. If the deafness has resulted from meningitis then the operative period may prove particularly distressing for the family. Sensitive preoperative counselling may do much to help reduce the stress.

The surgical incision should be planned so as to be well away from the actual implant. The squamous temporal bone is much thinner in a child and special caution needs to be exercised when fashioning the bed for the implant package. The remainder of the surgery is little different from adult implantation.

In the postoperative period seemingly simple matters such as suture removal may prove very traumatic and the Nottingham team has now adopted the use of absorbable subcutaneous sutures.

Children are in hospital for only two or three days postoperatively, which is no different from any other major middle ear surgery. Postoperative vertigo has not been encountered in any of the children in the Nottingham programme.

Tuning and Rehabilitation

Each electrode of the multichannel array has to be tuned to obtain threshold levels and upper, comfort, levels. Again, as with preoperative evaluation, this requires the specialised skills of those used to assessing young deaf children. The initial reaction of many children to the initial switch-on is to burst into tears; careful preliminary counselling will avoid distress to the parents from this. Tuning can be a very time consuming process, and it is hoped that current work involving peroperative measurements of electrically evoked stapedius reflex levels and brain stem auditory responses will help reduce this.

After the initial tuning, the process of auditory rehabilitation starts. The skills of the teacher of the deaf become paramount, not only in direct contact with the child but also in involving many others in this process, including parents and other members of the family, other teachers, and many more who come into daily contact with the child.

During the rehabilitation period, there is regular assessment of communication skills and also language development, the latter involving the skills of the speech therapist.

It will have become obvious from the foregoing that paediatric cochlear implantation demands a team approach at all stages, involving audiologist, otologist, teacher(s) of the deaf, speech therapist, medical physicist, radiologist, and not least the child and his or her parents and other family members.

Special benefits to children of cochlear implantation

The benefits of cochlear implantation in children may be greater than those for adults. Loss of hearing in young children is likely to result in failure to develop, or loss of already developed speech and language. Hearing is essential for this development. Mecklenburg et al have listed the benefits as primary and secondary (table 1).15

There may also be tertiary effects, with benefit to education and intellectual development and longer term employment prospects. The costs of cochlear implantation may be offset against reduced educational costs, reducing the additional costs of placement of a child in a special school for the deaf. The concept of the developmental critical period has become important in the past decade,16 with anatomical and metabolic changes in some brain stem auditory nuclei being demonstrated.17 Loeb notes that language acquisition, whether verbal or visual, is greatly compromised if training is not begun early.16 He comments that if critical periods exist in the development of auditory perceptive capabilities, this would introduce an urgency into the decision of whether to implant devices (that are now primitive but rapidly evolving).

While there is a clear trend towards continued improvement in performance with longer term use of implants, it remains to be seen whether prelinguistically deaf children will continue to progress, although preliminary evidence is encouraging. In Nottingham we have seen a steady improvement in performance with time. Preoperatively none of the children receiving implantation were even aware of environmental sounds; of the seven children assessed at 12 months after operation, three could discriminate some speech without lip reading and four could understand speech without lip reading.

Paediatric cochlear implantation in the United Kingdom

In 1989 the Department of Health and Social Security announced special funding for six centres to conduct assessment of cochlear implantation. The Nottingham paediatric cochlear implant programme received funding from this initiative, having carried out an initial five implants supported by charitable funds. The Nottingham programme is the only dedicated paediatric implant centre within the United Kingdom although a limited number of implants has been carried out in other parts of the country. Using the Nucleus 22 channel intracochlear device 19 children aged between 2 years and 10 years have received

**Table 1 Benefits of cochlear implantation**

<table>
<thead>
<tr>
<th></th>
<th>Primary</th>
<th>Secondary</th>
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<tbody>
<tr>
<td>Detection of sound at comfortable loudness levels</td>
<td>Improved voice quality</td>
<td>Improved speech production (accuracy)</td>
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<tr>
<td>Improved identification of the possible elements of speech</td>
<td>Increased phonological repertoire</td>
<td></td>
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<tr>
<td>Improved speech perception for closed, and open, set speech stimuli</td>
<td>Improved single word receptive vocabulary</td>
<td></td>
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<tr>
<td>Enhanced lip reading skills</td>
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**Table 2 Hearing before and after implantation in 20 children**

<table>
<thead>
<tr>
<th>Performance</th>
<th>Before implant (n=20)</th>
<th>After implant</th>
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<tbody>
<tr>
<td></td>
<td>Immediate (n=19)</td>
<td>6 months (n=11)</td>
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<tr>
<td>Can freely use telephone</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Understands speech without lipreading</td>
<td>1</td>
<td>4</td>
</tr>
<tr>
<td>Discrimination of some speech sounds without lipreading</td>
<td>6</td>
<td>3</td>
</tr>
<tr>
<td>Understands speech with lipreading</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Recognises environmental sounds</td>
<td>1</td>
<td>4</td>
</tr>
<tr>
<td>Responds to spoken language</td>
<td>11</td>
<td></td>
</tr>
<tr>
<td>Aware of environmental sounds</td>
<td>7</td>
<td></td>
</tr>
<tr>
<td>No awareness of sounds</td>
<td>20</td>
<td></td>
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Detection of minimal residual disease in leukaemia

The advent of modern intensive treatment has ensured that the majority of children with acute lymphoblastic leukaemia (ALL) will attain complete haematological remission. The length of this remission is, however, dependent on effective elimination of malignant cells as relapses are almost certainly due to residual leukaemic cells unresponsive to initial treatment. Such cells are referred to as minimal residual leukaemia or minimal residual disease (MRD) which is effectively the lowest number of abnormal cells identifiable with the available methods.

Detection of minimal residual disease
Remission in leukaemia is conventionally defined by a level of 5% leukaemic blasts or less in the bone marrow. However, identification of low numbers of leukaemic cells in a background of normal regenerating bone marrow precursors presents difficulties for even experienced haematologists. More sensitive and specific methods have been developed to complement morphology,¹ ² and these will be discussed.

Immunological analysis
Immunological analysis has shown that aberrant or asynchronous surface markers are often present on leukaemic cells but absent from their normal counterparts.³ ⁴ Two colour immunofluorescent techniques were initially developed to detect residual blasts in the bone marrow of patients with T cell acute lymphoblastic leukaemia as the phenotype CD7+/cytoplasmic CD3+/terminal transferase (TdT)+ is normally found only on thymic precursors. ³ The coexpression of the CD10 antigen on blasts from patients with ‘common’ ALL together with the myeloid associated antigens CD13 or CD33, or of CD10 with the T cell markers CD2 or CD7, is a useful indicator of MRD which can be exploited using two colour immunofluorescence and fluorescence activated cell sorter analysis. The technique can detect one leukaemic cell in 10⁴ normal cells.⁵ This technique is limited by the small numbers of leukaemias expressing such aberrant surface markers. In addition, the frequency of normal regenerating precursor cells expressing unusual combinations of surface markers has not been well studied and may be more common than realised, giving rise to false positive results.

Cytogenetic abnormalities
Consistent cytogenetic abnormalities in the malignant cells are useful markers to monitor MRD, but chromosome analysis is dependent on dividing cells, and in ‘remission’ narrows the 1–5% abnormal cells present may not enter mitosis, thus limiting its use. The method is labour intensive, requires fresh samples, and at least 100 cells need to be screened for each sample.

Molecular analysis
Molecular analysis provides a sensitive and specific technique to monitor MRD, particularly in cases of ALL.¹ The DNA rearrangements in the genes coding for the immunoglobulin heavy chain (IgH) in B cells, or the T cell receptor complex in T cells, can be detected in the majority of cases of ALL of T or B cell origin.¹ Detection of clonally derived cells has been shown to be useful to evaluate the remission status of patients undergoing treatment and to predict early relapse.⁸ ⁹

B LINEAGE LEUKAEMIAS
In B lineage ALL, the IgH gene rearrangements are unique...