Chapter 1
Current trends in paediatric cochlear implantation

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Cochlear implantation now ranks as one of the most successful forms of management enabling profoundly deaf individuals to gain a sense of hearing and to develop and maintain spoken language skills. The introduction of a new healthcare technology such as cochlear implantation demands a close scrutiny of benefits and eventual outcomes to justify both the investment of resources and the risks associated with any medical and surgical intervention. In the case of children the responsibility for making decisions rests with their parents and it is vital that, prior to considering implantation for their children, they should be informed of a full range of risks, benefits and outcomes so that they can make an informed choice based on evidence.

There are complex issues surrounding the assessment of benefit in young deaf children. Benefit may be defined differently by parents, purchasers of healthcare, representatives of the deaf community and eventually by the children themselves as they become teenagers and adults. Progress for deaf children could, of course, be influenced by a wide range of variables including duration of deafness, age at implantation or hearing-aid fitting, degree of residual hearing, etiology, efficiency of hearing aid or cochlear implant use, mode of communication, educational management, degree of home support and commitment, to name just a few. These variables and their complex interactions pose challenges to investigators in their search for definitive predictions of benefit for individual cases. It is necessary, therefore, to consider observable trends in order to

enlighten our knowledge about cochlear implantation and its success or failure. It is instructive to consider these trends under various headings.

Audiological trends

The availability of long-term outcomes from cochlear implantation has produced an interesting dilemma. Early candidates, for whom the long-term outcomes are now available, often outperform their profoundly deaf peers who continue to use hearing aids and many now function on a par with severely deaf hearing-aid users (a full discussion is given in Chapter 4). Performance and progress can be assessed using a wide variety of measures that will be outlined in later chapters of this book but the most obvious changes are seen in spoken language perception and speech production skills, described in more detail in Chapter 10. The limitations of hearing aids for different groups of children now need to be considered in the light of the strong evidence of the benefits of cochlear implantation. The early candidates for cochlear implantation tended to be deafer, with longer durations of deafness, and tended to be older at the time of implantation than children who have received cochlear implants in more recent years. Each of these factors could militate against optimum performance with the devices and yet many of these children have still often outperformed their hearing-aided peers. What sort of long-term performance might we expect from children for whom all factors are working in their favour? Present cochlear implant systems are more sophisticated than the older devices and the application of the latest technology to children who have greater levels of neural survival (associated with significant amounts of residual hearing), and who receive their implants at more optimum times, should produce far better results.

Cochlear implant selection criteria have gradually relaxed over the years as a direct consequence of the documented high level of performance of the users, giving rise to the trend to implant children with greater amounts of residual hearing. The improvement of speech intelligibility and of access to telephone use are now very real possibilities. These were not envisaged as outcomes in the very early days of cochlear implantation when the expectation was to impart some basic and rather rudimentary stimulation of the auditory system. The significant changes in the audiological selection criteria that have taken place will be detailed in Chapter 4.

Trends in implantation age

At the beginning of the new millennium early implantation has become a factor of great importance in paediatric cochlear implantation and serious debate has taken place about the most appropriate age to implant a young deaf child. Auditory discrimination abilities have been demonstrated in human subjects very early in life, as normally hearing neonates have a preference for the voice to which they were mostly exposed *in utero* (DeCasper and Prescott, 1984). Infants with hearing are becoming linguistically sophisticated by one year of age, although this may not be clearly visible to naïve observers. During this period of time, the neurones in the auditory pathways are maturing and billions of major neural connections are being formed. The auditory system appears to adapt in response to psychophysical and electrophysiological stimuli over time (Ruben, 1997). When sensory input to the auditory nervous system is interrupted, especially during early development, the morphology and functional properties of neurones in the central auditory system can break down (Sininger, Doyle and Moore, 1999). It seems that the ability of neuroplasticity in the developing auditory system declines as children grow older and therefore early implantation could result in shorter auditory deprivation and better spoken language acquisition.

When reviewing the literature with regard to age at implantation there have been conflicting conclusions in the past. This may arise from studies which contain small numbers of children, and widely heterogeneous groups, and which have only comparatively short-term results. (Nikolopoulos et al, 1999). Shea, Domico and Lupfer, (1994) for example, found that age at implantation was positively correlated with outcomes in prelingually and peri-lingually deafened children. This earlier study may be thought to support later, rather than earlier, implantation. However, only 26 children were studied with ages at implantation ranging from two to 16 years old, and more than half of them had been implanted before less than 1.5 years. Children older at implantation may initially do better than younger children, because of their better language levels and ability to carry out testing, but when one follows the younger implanted children over a longer time-scale, the better outcomes in these younger children become apparent, as the following study shows.

Outcomes in terms of speech perception and production were measured on open and closed-set measures of speech perception and on two hierarchical scales of auditory perception and speech intelligibility (Nikolopoulos et al, 1999). The hierarchical scale of auditory perception was Categories of Auditory Performance (see Chapter 3; Archbold et al, 1998) and the scale for speech intelligibility was the Speech Intelligibility Rating (see Chapter 10; Allen et al, 1998). Results were reported on 126 congenitally or prelingually deaf children. Up to two years after implantation, a positive correlation between these outcomes and age at implantation was found. However, at three and four years after implantation, a negative correlation was found (all statistically significant, $p< 0.5$). This study demonstrated the importance of long-term follow-up in those children implanted early. Although the results support early implantation, older children should not be excluded from assessment on the basis of age alone, and may do well.

The effect of age at implantation on the long-term functional outcome is illustrated in Figures 1.1 and 1.2. Figure 1.1 illustrates the comparative percentages of children achieving either of the two top categories of the rating scale, Categories of Auditory Performance (Archbold, Lutman and Nikolopoulos, 1998). Significantly more children implanted below the age of five achieve these categories four years after implantation than those implanted over

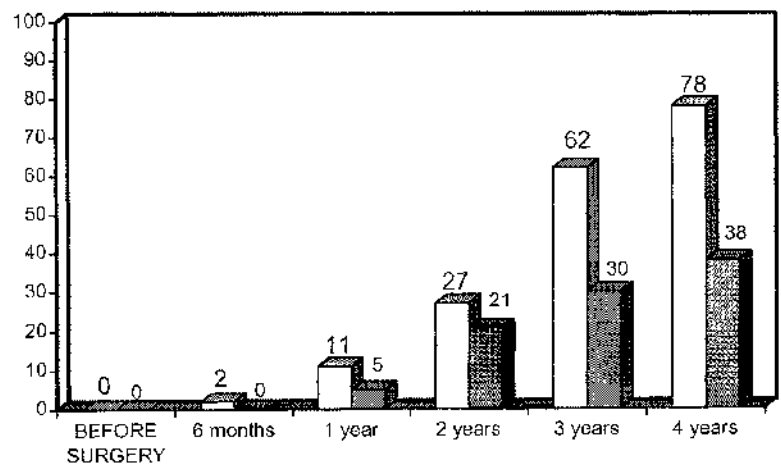

Figure 1.1. Percentages and actual numbers (on top of columns) of congenitally deaf children achieving the category of 'understanding conversation without lipreading' or 'use of telephone with a known speaker' using Categories of Auditory Performance (CAP), comparing those implanted under five years of age with those implanted at five years of age and over.

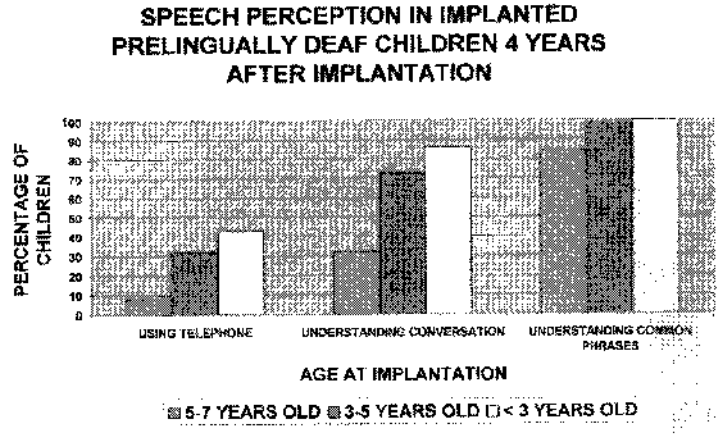

Figure 1.2. Pre-lingually deaf children achieving understanding of common phrases without lipreading, understanding conversation without lipreading and using the telephone, four years after implantation, comparing implantation at different ages.

five years of age. Figure 1.2 breaks this down further: it shows children born deaf or pre-lingually deafened, four years after implantation, and their attainments as rated by Categories of Auditory Perfomance on using the telephone, understanding conversation without lipreading and understanding common phrases without lipreading, comparing those implanted below the age of three, with those implanted between three and five, and those implanted over five. While there is little difference in outcomes on closed-set measures, such as understanding common phrases, there is a significant difference due to age at implantation in those achieving the understanding of conversation and the use of the telephone. A similar result is achieved using the Speech Intelligibility Rating, and shown in Chapter 10.

A consensus is now being established on the importance of early implantation, summarized by Miyamoto (2002) who considered that six months of age is now the youngest age to consider implantation. Other studies have supported the idea of implanting deaf children younger than two years of age (Lenarz in Honjo and Takahashi, 1997; Waltzman and Cohen, 1998), but there is, as yet, no strong evidence that a child implanted at one year of age will outperform a similar child implanted at two years of age. On the other hand, certain concerns have been raised with regard to implantation of very young children including:

- the relatively small size of the skull and its growth in young children with potential surgical difficulties of access and

possible increase in surgical risk (later complications of electrode displacement);
- the high prevalence of otitis media in this age group and subsequent related complications; and
- difficulties in tuning, rehabilitation and identification of other difficulties.

These issues are discussed more fully in Chapter 3, but the data so far have not supported these concerns and implantation in very young age is feasible and relatively safe (Waltzman and Cohen, 1998; Miyamoto, 2002).

Other issues, such as post-meningitic ossification, may expedite the decision to proceed, irrespective of the child's very young age. On the other hand, adequate time is needed for parents to realize the existence of, and adjust to, their child's deafness and for audiologists to establish true hearing thresholds and to assess the potential benefits of conventional hearing aids.

In conclusion, the current trend in paediatric cochlear implantation is to implant younger children, and this is likely to continue with the introduction of Newborn Hearing Screening.

Trends in educational management

The educational management of children following implantation has become a major issue.

Educational placement is an important outcome measure from paediatric cochlear implantation as it is likely to have educational implications (Archbold et al., 1998). Comparisons of the educational settings of profoundly deaf children implanted in the Nottingham programme with those of profoundly deaf children in the UK revealed that profoundly deaf children implanted early (when they are still at the pre-school stage) are three times more likely to attend mainstream school three years following implantation than profoundly deaf children of the same age using hearing aids (Archbold et al., 2002). The educational settings of implanted children with three years experience with their devices were: 41% in mainstream schools, 52% in units in mainstream schools and only 7% in schools for the deaf. The respective percentages for age-matched profoundly deaf children with hearing aids were: 13% in mainstream schools, 62% in units of mainstream schools, and 25% are placed in schools for the deaf (Figure 1.3).

Data from the Nottingham programme also revealed that children implanted at an age younger than five years old are three

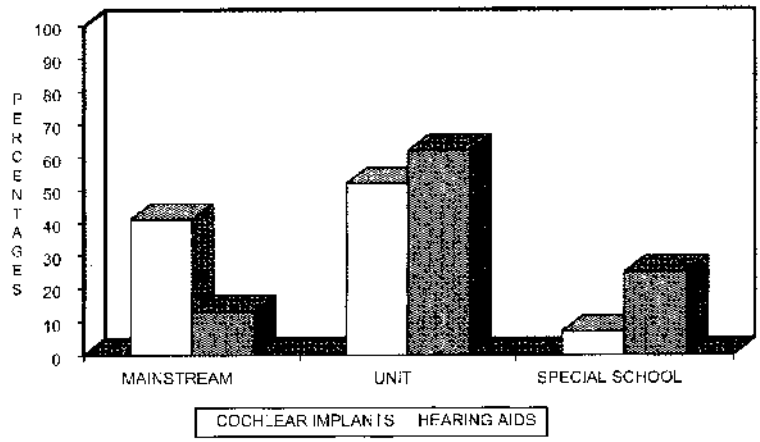

Figure 1.3. Educational placement of profoundly deaf children of the same age, comparing those with hearing aids and those with cochlear implants. Cochear implant children are at the three-year interval following implantation. All implanted children were implanted before five years of age.

times more likely to be placed in mainstream schools and five times less likely to be placed in schools for the deaf in comparison with children implanted above the age of five (Figure 1.4). However, educational placement continues to be monitored; with the challenges of secondary education, will this trend continue in the long term? Chapter 3 gives a fuller discussion of the educational issues, with its complexities.

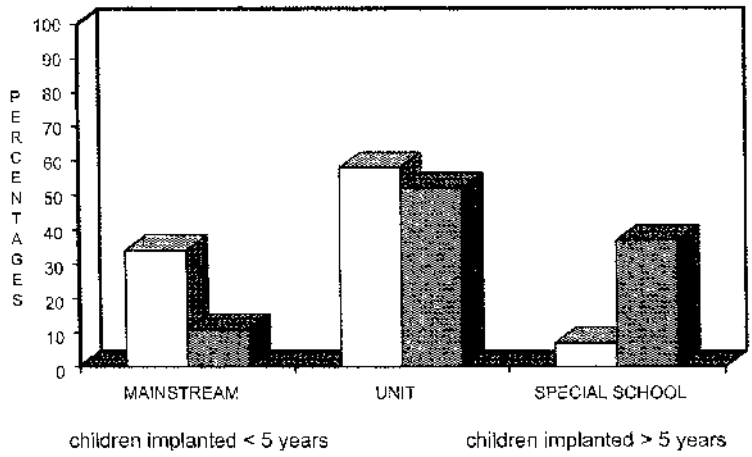

Figure 1.4. Educational placement of implanted children three years after implantation. Comparisons of those children implanted under five years of age with those implanted over five years of age.

A debate also exists with regard to the most appropriate mode of communication for deaf children. We explored the relationship between the approach to communication and measures of speech perception and production three, four, and five years following cochlear implantation in young pre-lingually deaf children (Archbold et al., 2000). The communication approach used by each child was classified by his/her teacher of the deaf at each interval into one of two categories: oral communication and signed communication. The results revealed that, at all intervals, those children classified as using oral communication significantly exceeded those using signed communication on measures of speech perception and intelligibility ($p < 0.05$). However, when those children who had changed from using signed to oral communication during the three years after implantation were compared at the three-year interval with those who used oral communication throughout, there was no significant difference in their results. It remains to be explored whether children use oral communication after implantation because they are doing well, or whether they do well because they are using oral communication.

Trends in patient-oriented healthcare

Parents' perspectives are very important and therefore there is a compelling need to involve them fully in assessment of outcomes. It is now becoming more broadly accepted that any new method of treatment should engage the wider public in discussion about what a national health service should provide and who should decide it (Chisholm, 1999). Moreover, it has also been suggested that repeated evaluation of patients' (or parental) views should become an integral part of routine healthcare (Richards, 1999).

However, the methods of assessment of quality of life issues with regard to both children and parents following paediatric cochlear implantation are extremely limited; as described in Chapter 11, parents at the Nottingham programme are asked to complete a questionnaire prior to implantation, and annually following implantation. The results show that their expectations were met, or surpassed, following implantation (Nikolopoulos et al., 2001; and see Chapter 11 for fuller discussion). A further questionnaire has been developed, based on open parental responses three years after implantation, which has been found to be robust and repeatable (Archbold et al., 2002). Content analysis of the open responses revealed the three most common constructs mentioned by parents to be the positive changes in confidence (linked to communication) seen in their children, the need for

continuing links with the implant centre, particularly for long-term technical support, and the importance of liaison between the cochlear implant centre and the local educational service. Clearly, paediatric cochlear implantation is a service that necessarily closely interacts with parents and the local professionals providing long-term support to the child, and ways in which this can be organized are discussed in Chapter 3.

Trends in measures of effectiveness

It is no longer sufficient to provide information on safety or efficacy of healthcare services; we are now required to demonstrate cost effectiveness (Niparko in Niparko et al., 2000). This obliges services to produce information both on accurate costings and on outcomes, requiring careful data collection over time. Several measures, including Categories of Auditory Performance, and Speech Intelligibility Rating (Allen, Nikolopoulos and O'Donoghue, 1998) have been developed by the Nottingham programme to be accessible to the non-specialist, including purchasers of services, and to demonstrate outcomes over a long time-frame from different patient groups. Chapters 3 and 10 give further details. In addition, a major indication of effectiveness is the usage rate of the device. Unlike other implantable devices, the child can choose not to wear the external equipment, and thus continued high usage rates are an indication of patient satisfaction and an effective use of the financial investment. The Nottingham programme constantly monitors the usage rates via parents and teacher reports. Five years after implantation, 99% of our children are wearing their devices all or most of the time, according to parents, and only 1% are non-users at this stage of the programme. We have a few children who have later chosen to wear their devices only for some of the time, and 1% who are now choosing not to wear their devices at all. Maintaining this high usage rate relies on careful assessment of candidatures, appropriate tuning of devices, careful monitoring of functioning, and liaison with parents and local services.

Over the 10 years since the establishment of the programme, the necessary infrastructure to support this long-term effective use has become more evident, and the various chapters in this book describe the developing service provision, as the numbers grow. Dramatic changes have been seen in candidature, expectations and outcomes. Inevitably, practice has changed too, and those of us working in this challenging and ever-changing field have a responsibility to ensure that our children have the best possible lifelong care.

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