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Cochlear Implants in Children: A Review

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1. Introduction

In 1980, the first child in the world was implanted with the single-channel House cochlear implant device (Eisenberg & House, 1982). Children who initially received cochlear implants during this first paediatric clinical trial were quite old compared to current ages (the average age in the first House clinical trial was 8 years, whereas children are now being implanted as young as 6 months of age), and the majority communicated using sign language (Eisenberg & Johnson, 2008). It is now known that implanting older children who do not communicate orally gives little chance of speech perception or spoken language development. In 1985, the first children received a multichannel cochlear implant in Australia (Clark et al., 1987). This clinical trial selected children who had a higher potential for success, including shorter duration of deafness and a commitment to oral communication both at home and in their educational programs. At this time, it was unknown whether the speech processing schemes used with adults who had lost their hearing after developing language (ie. post-lingually deafened) would be appropriate for facilitating the speech perception and language development of young children with immature auditory systems. It is important also to note that the desired outcomes for adults and children differed; while the goal for adults was to improve auditory skills and communication using previously acquired cognitive, spoken language, and social skills, the goal for children was to develop these skills using the auditory information provided by the cochlear implant, having had no useful auditory experience (and therefore presumably no neural development of their auditory system) until they received their cochlear implant. The implantation of children was also highly controversial. For many years, cochlear implantation in children was opposed by the Deaf Community, on the grounds that deafness in children should be considered as a cultural and linguistic difference rather than as a disability that could be remediated by a cochlear implant. Over time, this view has changed such that in 2000, a position paper of the National Association of the Deaf in the U.S. stated that "cochlear implantation is a technology that represents a tool to be used in some forms of communication, and not a cure for deafness" (National Association of the Deaf, 2000).

It is now well documented that children with severe-profound hearing loss receive significant benefits from cochlear implants in terms of speech perception and language development (Blamey et al., 2006; Geers et al., 2008; Moog, 2002; Nicholas & Geers 2007). Cochlear implants are becoming the standard of care for children with severe-profound hearing loss, with increasing uptake of simultaneous bilateral implants over recent years. There is a large variation in implementation of cochlear implant technology around the

world, and also within regions in some countries. Bilateral implantation is becoming the standard of care for children in developed countries, such as Germany, England and the United States, while in developing countries it is very infrequent. In less developed countries, many children are still receiving unilateral single-channel cochlear implants, which are cheaper to manufacture, and many are not able to access the technology at all due to high cost. For example, of the estimated 1 million children with profound hearing loss in India, only approximately 5000 are reported to have cochlear implants. It is difficult to estimate how many children worldwide have received cochlear implants to date, as reports vary widely. However, in December 2010, the U.S. Food and Drug Administration (FDA) reported that approximately 219,000 people worldwide had received implants (National Institute on Deafness and other Communication Disorders, 2011). Despite variations in estimates, it is generally accepted that approximately half of the number of cochlear implant recipients are children.

2. Suitability for a cochlear implant

2.1 Criteria for candidature

In the early days of cochlear implantation in children, children were only considered as suitable recipients for a cochlear implant when they had no useable aided hearing, and therefore had nothing to lose if the outcome were not good, as cochlear implantation damages the inner ear such that acoustic hearing is not usually possible post-operatively. As technological improvements in electrode design, speech processing strategies, receiver/stimulator design and programming have gradually facilitated improving outcomes with cochlear implants, the clinical perspective has changed.

Determining suitability in children is a more complicated process than it is for post-lingually deafened adults, whose speech production and language skills are fully developed. Whereas for adults it can be assumed that the ability to perceive speech is limited by hearing ability alone, for children, speech perception is limited by language knowledge and speech production skills as well as by residual hearing quality and quantity (DesJardin et al., 2009; Sarant et al., 1997). Unsurprisingly, speech perception scores (obtained from measuring the number of sounds, words, or sentences perceived correctly on a test) in children are more highly correlated with spoken language abilities than with any other factor (Blamey et al., 2001a), and are also influenced by speech production skills (Paatsch et al., 2004). Therefore, basing decisions about cochlear implant candidature for children on speech perception scores alone could risk implanting some children who have sufficient aided hearing to develop spoken language through hearing aids, but who are limited in their speech perception ability by their undeveloped spoken language skills. This risk has increased over time as the age at which children receive cochlear implants has decreased. Further, as speech perception results with cochlear implants have improved, the amount of hearing being risked in order to achieve the potential benefits of a cochlear implant has increased. Given this increasing risk, accurate prediction of a particular child's potential to benefit from a cochlear implant has become even more important.

Blamey and Sarant (2002) proposed a method of combining speech perception and language assessment scores to calculate an objective criterion for cochlear implant suitability, so that a child's pre-operative aided speech perception performance is compared to a distribution of

speech perception scores for children with cochlear implants who are matched according to language ability (Blamey & Sarant, 2002). While this approach is helpful for older children with some language ability, it is not suitable for use with very young children whose speech perception, production and language skills are undeveloped, independent of their degree of hearing loss, and for whom, due to behavioural and cognitive developmental issues, it is very difficult to assess speech perception ability.

Since the 1990's, several researchers have proposed alternate methods of determining suitability for a cochlear implant in children. Osberger et al. (1991) classified children using hearing aids into 'gold', 'silver' and 'bronze' categories, based on their unaided pure tone thresholds (PTA) averaged across 0.5, 1, and 2 kHz. Initially, it was predicted that only children in the 'bronze' category (mean >110dbHL and >110dbHL at two of the three frequencies) were suitable candidates for a cochlear implant. These categories were revised when it became apparent that children with cochlear implants were outperforming not only hearing aid users in the bronze, but also in the silver (mean = 104dbHL and 101-110 dbHL at two of the three frequencies) and gold (mean = 94dbHL and 90-100 dbHL in two of the three frequencies) categories. A further methodology that compared speech perception results for children using hearing aids and cochlear implants in order to determine criteria for suitability used the concept of 'equivalent hearing loss' (EHL). Boothroyd and Eran (1994) compared the abilities of children using hearing aids with those using a cochlear implant on an imitative test of phonetic (speech sound) contrasts, and derived EHL by plotting speech perception results against the three-frequency unaided PTA for each ear. Linear regression statistical analysis was used to transform the speech perception scores of the children into EHL values. Although the EHL for the children using cochlear implants suggested that their potential for speech perception was similar to that of children with a severe hearing loss using hearing aids, there were still children using cochlear implants whose speech perception skills were no better than those of children with a profound hearing loss using hearing aids. In 1997, Boothroyd reported that children with cochlear implants who were educated mostly in oral communicative environments achieved speech perception scores equivalent to those of children using hearing aids with a hearing loss in the 70-89 dbHL (severe) range (Boothroyd, 1997). Similar results have been reported more recently (Davidson, 2006; Eisenberg et al., 2004).

Throughout the current decade, several studies of large numbers of children with cochlear implants have reported speech perception results that are comparable to those achieved by post-lingually deafened adults using cochlear implants, and even to those achieved by children with a moderate hearing loss using hearing aids (Geers et al., 2003; Svirsky et al., 2004; Tajudeen et al. 2010; Wie et al. 2007). In response to these achievements, the criteria for suitability have again changed such that even very young children with a severe to severe-profound hearing loss are now deemed suitable recipients for cochlear implants, and children with significant, or useable, residual hearing are currently being implanted in centers not under the jurisdiction of the United States FDA (Geers & Moog, 1994; Leigh et al., 2011; Svirsky & Meyer, 1999; Zwolan et al., 1997). Currently, the more conservative FDA guidelines approve cochlear implantation in children aged 12-23 months with bilateral profound sensorineural hearing loss (>90dbHL) and in children aged 2 years and older with severe-profound hearing loss (greater than or equal to 90dBHL in the better hearing ear).

The introduction of neonatal hearing screening programs in developed countries over the past decade has meant that hearing loss is now identified in babies as young as a few days or months old, and there is earlier referral and diagnosis than ever before (Dalzell, 2000; White & Maxon, 1995; Yoshinaga-Itano, 2003a). Very young infants and toddlers now represent the majority of paediatric cochlear implant candidates in these countries, and for these children, decisions about candidacy must currently be based solely or primarily on audiometric information if cochlear implants are to be given early, as there are limited tools available to measure speech perception or language abilities in this age group. The audiometric information is usually objective data obtained from the transient evoked auditory brainstem response (ABR) used in hearing screening, otoacoustic emissions, or auditory steady state responses (ASSR). These results may be combined with behavioural data derived from testing conducted by audiologists, depending on the protocol of individual cochlear implant programs. Most recently, an "equivalent PTA" model was derived to be applied to audiometric data for very young children from a comparison of the open-set speech perception scores of preschool and elementary school-aged children using cochlear implants and hearing aids. The model gives equivalent PTA for a 75% through to 95% chance of improvement in speech perception outcomes in 5% steps. Using a less conservative 75% chance of improvement criterion (as opposed to the 95% criterion that has until now been applied), the model recommends that children with bilateral profound hearing loss through to children with unaided pure tone average thresholds of 75 to 90 dBHL are suitable recipients for cochlear implants, while children with lesser hearing loss than 75dbHL are encouraged to continue with hearing aid use (Leigh et al., 2011).

2.2 Children with additional disabilities: Implications for candidacy

It is well established that 30-40% of children with severe-profound hearing loss also have an additional physical and/or cognitive disability, such as visual impairment, cognitive impairment, learning disabilities, autistic spectrum disorders (ASD) or developmental delay (Archbold & O'Donoghue, 2009; Edwards, 2007; Holt et al., 2005). Often, the additional disability is related to the cause of deafness, and is part of a syndrome or other grouping of disabilities. Children with additional disabilities present a further challenge with regard to determining suitability for cochlear implants, because the degree of benefit derived by the 'average' child with hearing loss is unlikely to be experienced by these children due to the effects of their additional disabilities. For this reason, children with additional disabilities were not considered suitable cochlear implant candidates for many years. Although excluded from FDA clinical trials in the past (Holt et al., 2005), small numbers of children with additional disabilities have received cochlear implants. Little is known about the degree of benefit children with hearing loss and additional disabilities derive from cochlear implants with regard to speech perception and spoken language development, for several reasons. Firstly, much of the research effort around cochlear implants has been directed at identifying outcomes and predictive factors for the majority of children with cochlear implants who do not have additional disabilities. Secondly, due to the fact that there are smaller numbers of children with additional disabilities, and many are unable to complete standardised assessment procedures, quantitative analysis of outcomes has been difficult. A further challenge is that there are a large number of additional disabilities spread across a relatively small population of children, therefore obtaining sufficient numbers to define the

aspects of each disability and its impact on communication development after implantation has been difficult.

The few studies of children with cochlear implants and additional disabilities have generally reported poorer performance on speech perception, production and language assessments, particularly when higher level speech processing abilities are required. For example, in one of the first studies of these children, (Pyman et al., 2000) found that although 90% of 75 children with motor and/or cognitive delays could discriminate consonants and vowels after four years of cochlear implant use, only around 60% of the children were able to use this information to perceive open-set sentences (those presented with no context), compared to over 80% of children without additional disabilities. Similarly, a further study of children with a variety of disabilities, such as attention-deficit disorder, cerebral palsy, central auditory processing disorder, dyspraxia and autism, showed some speech perception skill development at a slower rate than for the general population (Waltzman et al., 2000). Children whose additional disability is mild can derive significant benefit from cochlear implants, whereas children with more severe disabilities have much less favourable outcomes, with some showing almost no progress (Edwards, Frost & Witham, 2006; Filipo et al., 2004; Hamzavi et al., 2000; Holt & Kirk, 2005; Meinzen-Derr et al., 2011; Vlahovic & Sindija, 2004). Most studies have highlighted that children with additional disabilities require longer periods of implant use before demonstrating any benefit, and as for children in the general cochlear implant population, variation in outcomes is wide for children with additional disabilities (Hamzavi et al., 2000; Waltzman et al., 2000). It was reported for some children that the assessment tasks were too difficult to complete (Donaldson et al., 2004; Waltzman et al., 2000), which is a factor that has added to the difficulty of determining outcomes for this population.

Children with autistic spectrum disorders (ASD) have historically been considered poor cochlear implant candidates, but as the age at which children are being implanted has decreased, there are now a number of children who have been implanted before their diagnosis of ASD. The single published study of progress in a group of children with ASD reported that smaller gains on tests of speech perception and language had been made in comparison to those reported for the cochlear implant general population, but that parent reports suggested positive improvements in their children's functioning and responsiveness (Donaldson et al., 2004).

In summary, although the degree of benefit obtained from cochlear implants is often lower for children with additional disabilities, many children still receive measurable benefit from their devices, and this benefit adds to their quality of life. Some of the observed benefits cannot be quantified on standardised tests, and have been instead reported anecdotally, with observations of improvements in social interaction and responsiveness to the environment, behaviour, vocalization, self-help skills, motor skills and the ability to follow instructions (Donaldson et al., 2004; Filipo et al., 2004; Fukuda et al., 2003; Waltzman et al., 2000; Wiley et al., 2005). There is still a need to determine the impact of additional disabilities on post-operative benefit with cochlear implants, and to define more clearly what benefits might reasonably be expected for children with different additional disabilities. The point at which a cochlear implant will not be beneficial also needs to be determined with regard to the degree of severity of additional disabilities, and the definition of benefit should be carefully explored, with improved psychological well-being, children's

maximum potential, and quality of life being taken into consideration in addition to quantitative outcomes on tests.

2.3 Candidacy and selected aetiologies/pathologies of deafness

A further group of children for whom candidacy issues are more complex are those with selected pathologies that not only cause severe-profound hearing loss but may also impact on outcomes with cochlear implants. Although there are many such pathologies, the most common of these will be discussed as examples of the impact aetiology, or cause of hearing loss, may have on post-implantation outcomes.

In the 1990's, auditory neuropathy (AN) was defined as a distinct type of hearing disorder that disrupts neural activity in the central and peripheral auditory pathways (Starr et al., 1996). Auditory neuropathy is characterised by normal outer hair cell function in the cochlea (which enables many babies to pass newborn hearing screening if otoacoustic emission testing is used), and a retro cochlear lesion (dysfunction in the inner hair cells or auditory [eighth] nerve), which manifests as an absent or abnormal response to auditory brainstem response (ABR) testing. Features of this pathology include poorer than expected speech perception abilities in relation to degree of hearing loss in the majority of children, with some children who have only a mild hearing loss demonstrating a severely impaired ability to use their hearing for speech understanding (Rance et al., 2007). This pathology affects approximately 0.23% of at-risk children (Rance et al., 1999). Given the unusual pattern of perceptual deficits that characterises AN, much of the research in this area has focused on whether or not a cochlear implant can assist these children to understand speech through their hearing. The few published investigations on speech perception have varied widely, reporting no benefit (Miyamoto et al., 1999; Teagle et al., 2010) through to benefit comparable to that received by the general population of children with cochlear implants (Buss et al., 2002; Peterson et al., 2003; Rance & Barker, 2008; Trautwein et al., 2000). For the children who demonstrated significant benefit, it was noted that electrical stimulation via the cochlear implant elicited ABR responses, which suggests that the implant was able to enable greater neural synchrony and therefore to overcome the desynchronization thought to underlie AN. Studies of language and speech production outcomes for these children are again limited, and results are similar to those for speech perception, with wide variation in outcomes, but also with some children demonstrating the same level of development as the general population of children with cochlear implants (Jeong et al., 2007; Madden, 2002; Rance et al., 2007). For parents of children with this pathology, there is reasonable evidence to suggest that children may benefit from a cochlear implant, although expectations may need to be lower than for the general population of children with sensorineural hearing loss.

Usher syndrome is the most common condition that affects both hearing and vision, and its major symptoms are congenital or progressive deafness resulting in severe-profound hearing loss, and progressive loss of vision due to retinitis pigmentosa, an eye disorder which causes night blindness and a loss of peripheral vision. Many children with Usher syndrome also have significant balance problems, which can delay walking in very young children. Approximately 6-12% of children with hearing loss, or 4 in every 100,000 births in the United States (Boughman et al., 1983) and 6 per 100,000 births in England (Hope et al., 1997) have Usher Syndrome, which is a genetic condition. Once children have lost their vision, the auditory information provided by a cochlear implant is their only means of

connecting and communicating with the world, so it is very important that these children are diagnosed and receive their cochlear implants early in order to establish communication through audition prior to the loss of vision. Usher syndrome is one of the 20% of causes of deafness that involve abnormalities in cochlea-vestibular anatomy. These abnormalities increase the potential for surgical difficulties and complications, such as damage to the facial nerve and incomplete insertion of the implant electrode array in the cochlea (Bauer et al., 2002; Chadha et al., 2009).

Some other children with congenital deafness also have cochlear abnormalities, often due to a range of genetic causes, another of which is CHARGE syndrome. Children with this rare genetic syndrome have deafness, visual problems, and a variety of other physical abnormalities, including serious heart defects, colobomas (or holes) in one or both eyes, growth retardation, genital abnormalities and external and internal ear malformations. Anatomical abnormalities in the structure of the cochlea can also create difficulties for programming, with reduced dynamic ranges for children with more severe cochlear abnormalities (Papsin, 2005). For these reasons, malformation of the cochlea was considered a contra-indication to cochlear implant surgery in the early years of cochlear implantation in children, and it is still not possible to implant some of these children (Bamiou et al., 2001). Despite these difficulties, initial results for small numbers of children with cochlear anomalies have shown that implantation is possible, with some children achieving speech perception and language results similar to those without anatomical abnormalities (Chadha et al., 2009; Dettman et al., 2011). Children with a common cavity anomaly (a single cavity in the cochlea) and other more severe syndromic anomalies have achieved much poorer results (Bauer et al., 2002; Chadha et al., 2009; Lanson et al., 2007; Loundon et al., 2003; Young et al., 1995).

Children with viral causes of deafness such as rubella, cytomegalovirus (CMV), toxoplasmosis and meningitis also require special consideration, as these viruses can cause developmental neurological deficits, including learning and cognitive difficulties (Edwards, 2007; Grimwood et al., 2000; Isaacson et al., 1996). A significant difference between children with deafness caused by meningitis and that caused by the other viruses is that while CMV, toxoplasmosis and rubella are contracted perinatally, children who have had meningitis will have experienced sound prior to infection and may have developed some spoken language skills. A further complication of meningitis is ossification (bone growth) within the cochlea, which is usually bilateral and can commence within four weeks of the illness (Durisin et al., 2010). This makes it imperative that children who have had meningitis are diagnosed with hearing loss and receive cochlear implants as soon as possible, before ossification limits both the potential for a full insertion and for benefit. Again, limited reports of post-operative benefit for children with these causes of deafness show a wide range of speech perception skills, intelligibility and language outcomes, with some children doing well (Francis et al., 2004; Lee et al., 2005) and others doing poorly (Isaacson et al., 1996; Ramirez Inscoe & Nikolopoulos, 2004; Wie et al., 2007).

3. Benefits of unilateral cochlear implants

3.1 Environmental awareness

At the most basic level, cochlear implants provide children with an auditory awareness of their environment. Through their cochlear implant, children can hear many environmental

sounds that would not be audible to them through hearing aids. These include high frequency sounds such as water running, birds singing, the kettle whistling, the car indicators ticking and the phone ringing. Being able to hear what is going on in their environment gives children a feeling of connectedness with the world, and also provides them with a greater degree of safety, although localization of sound sources is usually not achieved by most children with only one cochlear implant. Children are more easily able to hear their name being called, to determine when someone is speaking to them, and even to enjoy music. In the early days of cochlear implantation, when only older children received implants, and before it was realised that children could use cochlear implants to develop spoken language, environmental awareness was a prime motivating factor in the decision to implant some children (Sarant et al., 1994).

3.2 Speech perception

The cochlear implant assists children to process spoken auditory information in their environment both as an aid to lip reading, which is particularly useful in noisy educational environments, and also as a source of auditory information that can be relied upon without lip reading in appropriate listening conditions. As briefly mentioned earlier, speech perception results for children have steadily improved over time with advances in device hardware and software, surgical techniques, and experience with programming speech processors and habilitation. Initially, it was not expected that children with congenital hearing loss would be able to achieve the speech perception abilities shown by post-lingually deafened adults, but many children have exceeded these levels of perceptual ability. By the mid 1990's, 60 to 80% of children with unilateral implants achieved open-set word and sentence speech perception abilities comparable to those achieved by adults using audition only (Dowell et al., 1995; Dowell et al., 1997; Geers et al., 2003; Sarant et al., 2001).

More recent studies of children implanted at younger ages and using more recent technologies report even better speech perception abilities. While it has been suggested for some time that children with cochlear implants perform at a level equivalent to that of a child with a severe hearing loss using hearing aids (Blamey et al., 2001a; Boothroyd, 1997; Svirsky & Meyer, 1999), it has recently been reported that very young children can perform on tests of speech recognition at a level equivalent to that of children with a moderate hearing loss using hearing aids (Leigh et al., 2008b). Recent long-term studies have also shown that high proportions of children (79% and 60%) can use the telephone (Beadle et al., 2005; Uziel et al., 2007). These are considerable achievements for children who have been profoundly deaf since birth, and who have developed their auditory processing abilities through the reduced and fragmented sound provided by cochlear implants. It is also worth noting that a meta-analysis of 1916 reports on speech perception performance in children with cochlear implants suggested that, rather than levelling out, speech perception benefits continue to increase as children grow older (Cheng et al., 1999).

The assessment of speech perception abilities in adults and older children is relatively straightforward. It may involve an individual listening to a sound or word and pointing to a picture that best represents that sound or word (closed-set testing) or could involve the individual listening to and repeating a sound, word or sentence spoken by the assessor with no context (open-set testing). Children with age-appropriate cognitive abilities are able to complete these sorts of tasks from the age of around 3 to 4 years, when they have developed

the ability to label sounds, letters or words (Spencer et al., 2011). With the introduction of earlier diagnosis of hearing loss through newborn hearing screening, a need to assess very young children has developed in order to determine their suitability for cochlear implants. There are several methods of doing this, but these are less objective, and are much more reliant on the expertise and judgement of professionals in observing behavioural responses in very young children, and also on parent reports of responses to speech sounds and specific familiar words. Reports using these modified forms of speech perception testing in very young children have suggested that speech perception skills can develop rapidly within the first two years of cochlear implantation for children implanted before 4 years of age (Robbins et al., 2004a; Svirsky et al., 2004; Tajudeen et al., 2010; Wie et al., 2007).

As previously mentioned, much of the improvement in speech perception scores is attributable to advancements in technology, and particularly to the development of more effective speech processing strategies for the three commercially available cochlear implant devices (the Nucleus/Cochlear device, the Clarion device, and the Med-El device). The development of speech processing technology in the Cochlear device, which retains a dominant international market share of around 70% (Patrick et al., 2006) will be discussed as an example of this progress. In the early Nucleus 22-channel cochlear implant device, speech feature extraction schemes that presented only the fundamental frequency of speech and the first two formants (or bursts of energy) of speech (F0F2 and F0F1F2) were used (Clark et al., 1983). These strategies provided an aid to lip reading and very limited speech perception ability (Dowell et al., 1985). They had several disadvantages, such as not discriminating between speech and non-speech sounds, causing some environmental noises to sound quite unnatural, and providing no information above 3kHz, which made it impossible for users to perceive unvoiced information about consonants (such as 's', 'sh', 'f',' th' etc.).

In the early 1990's, a new strategy, known as Multipeak (MPEAK), was introduced with the goal of improving consonant recognition scores. MPEAK still used feature extraction algorithms, but also provided information about high frequency sounds on three fixed bands of the implant electrode array. The MPEAK strategy represented an improvement in that it distinguished between voiced and unvoiced sounds, and some electrodes were allocated to the representation of high frequency consonant information, which is extremely important for speech perception. The additional information provided through this speech processing strategy led to improved speech perception scores, particularly for fricatives (eg. 's', 'sh'), in both quiet and noise conditions (Clark, 1989; Dowell et al., 1991). Despite the improvements in benefit with MPEAK, an ongoing disadvantage of formant extraction strategies was that in background noise the speech processor made errors.

By 1995, improvements in electronics technology had allowed a new approach to speech processing to be adopted, using bandpass filtering principles in order to provide more information about the speech spectrum. The Spectral (SPEAK) speech processing strategy used bandpass filters to select 6 to 10 of the largest spectral components in each analysis time period and assigned these to particular electrodes in the cochlea. In this strategy, groups of electrodes, rather than single electrodes, were stimulated to represent particular speech features such as vowels, and stimulation occurred at a much higher rate than for previous strategies, which meant that more information could be presented more quickly. The selection of the highest amplitude information increased the chance of presenting only

the most salient speech information, and of suppressing lower amplitude background noise. The SPEAK strategy resulted in large increases in speech perception benefit for children and adults, particularly in background noise (Cowan et al., 1995; McKay et al., 1991), and probably contributed to the largest increase in speech perception benefit of all the technological advancements made before or since that time. Other significant technological improvements in cochlear implants over the last decade have included new receiver-stimulators, smaller, body-worn and behind-the-ear digital speech processors, and further high-rate speech processing strategies. These have facilitated further improvements in speech perception benefit, particularly in very young children who have had access to all of the recent technology.

One of the most challenging findings of research on speech perception ability in children with cochlear implants is the enormous variation in performance between individuals (Cowan et al., 1997; Pyman et al., 2000; Sarant et al., 2001; Staller et al., 1991). While many reports describe 'average' performance, this concept minimises and perhaps even disguises the fact that while many children do reasonably well, and some do as well as their peers with normal hearing, there are still children who derive very little benefit from their cochlear implant. This variation in outcomes makes it difficult to predict how a particular child will perform after implantation, and therefore to determine which children are suitable for a cochlear implant, particularly when they risk losing useable residual hearing in order to have one. Several factors that have been identified as predictive of post-operative performance will be discussed in section 4 of this chapter.

3.3 Speech production

The development of speech production has always been a significant problem for children with severe-profound hearing loss, as they do not have the auditory capacity to monitor their own speech or to hear the speech of normal-hearing individuals. For many years, most children using hearing aids with this degree of hearing loss have been rated as unintelligible, or as having very low intelligibility, to adult listeners unfamiliar with the speech of children with hearing loss (Bamford & Saunders, 1992; Gold, 1980; Spencer et al., 2011). Cochlear implants can provide children with auditory information that makes their own speech and that of others audible, so that they can learn from speakers with normal hearing, and self-monitor their own speech production. As with speech perception, children with cochlear implants show a wide range of speech production abilities, with many children performing at a very high level, and others showing low levels of performance (Connor et al., 2006; Spencer & Oleson, 2008; Tobey et al., 2003), but even children implanted at relatively late ages and with only a few years of implant use are generally rated as much more intelligible than their peers with a similar degree of hearing loss using hearing aids (Connor et al., 2006; Flipsen, 2008; Tobey & Hasenstab, 1991; Tye-Murray et al., 1995). Speech production outcomes have improved over time, as a result of longer periods of implant experience and improved hardware and speech processing strategies, although for many children they are still not equivalent to those of children with normal hearing (Chin et al., 2003; Peng et al., 2004).

Speech production skills and speech intelligibility ratings equivalent to those of 'gold' hearing aid users have been reported after less than 3 years of implant use (Blamey et al., 2001b; Svirsky et al., 2000). Children who are implanted at younger ages and use more

recent technology demonstrate the greatest achievements, with intelligibility ratings of 60-75% and much higher rates of speech production accuracy reported for children implanted as preschoolers (Ertmer et al., 2007; Flipsen, 2008; Peng et al., 2004; Tobey et al., 2003). More recent reports of children followed for longer post-operative periods of up to ten years have reported speech intelligibility rates of 77%, 90%, and 67% respectively, and suggest that the development of intelligibility does not plateau after a few years, but increases over time with chronological age and increased length of cochlear implant use (Beadle et al., 2005; Blamey et al., 2001c; Chin et al., 2003; Uziel et al., 2007). Beadle and colleagues showed that although 48% of the children in their study had developed connected speech that was intelligible to the average listener after 5 years of cochlear implant use, after 10 years this figure had increased to 77% (Beadle et al. 2005).

It was initially unknown whether children with cochlear implants would follow the same pattern of sound acquisition as their peers with normal hearing, or what their rate of progress would be compared to the former. In children with normal hearing, speech acquisition generally takes between 4 to 7 years (Chin et al., 2003). Studies of consonant and vowel acquisition in children with cochlear implants suggest that, on average, these children demonstrate a pattern of phoneme (or speech sound) acquisition similar to that of children with normal hearing (Ertmer et al., 2007; Serry et al., 1997), although their rate of development is often slower (Blamey et al., 2001b). This has meant that the speech acquisition process has still been incomplete at the age at which children with normal hearing have mastered speech production, but with little or no evidence that a plateau in development has been reached for children implanted between 2 and 5 years of age (Blamey et al., 2001c). Initial investigations of a small number of children implanted before the age of 12 months have yielded conflicting results, with one study reporting that the rate of speech production development for children implanted under the age of 12 months matched that of children with normal hearing (Leigh et al., 2008c), and another finding that children implanted before age 12 months and those implanted between 12-24 months showed no difference in their speech production development (Holt & Svirsky, 2008). Future research will hopefully clarify the critical period during which children should receive cochlear implants in order to facilitate speech production outcomes that are similar to those of children with normal hearing.

3.4 Language development

Language development is generally measured using standardised assessments of vocabulary and grammatical knowledge. In the early 1990s, most reports on language were case studies demonstrating changes thought to be associated with cochlear implantation, but knowledge in this area has grown over time, and there is now solid evidence for large numbers of children regarding language outcomes. Initial research concentrated on whether children with cochlear implants developed language more quickly than their peers with hearing aids, or compared development to predictions based on pre-operative language development with hearing aids. One of the earlier studies compared language development in three groups of children with cochlear implants, hearing aids and tactile aids (body-worn aids that provide vibratory or electrical stimulation) over 3 years (Geers & Moog, 1994). On average, the language growth of children with cochlear implants in this study was equal to or exceeded that for the other groups of children, and even approached that of children with hearing aids who had 20dB better hearing. Earlier this decade, children with cochlear

implants were reported to be developing at a rate similar to that of children with a severe hearing loss of around 78dbHL (Blamey et al., 2001a), and it is now well established that on average, children with cochlear implants demonstrate significantly faster spoken language development than their peers with similar levels of hearing loss who use hearing aids (Connor et al., 2000; Miyamoto et al., 1999; Svirsky et al., 2000; Tomblin et al., 1999). Given these promising results, the focus changed to comparing the progress of children with cochlear implants to that of their normally-hearing peers.

By the late 1990's, although language outcomes for children with cochlear implants had improved compared to those for children with similar degrees of loss using hearing aids, on average, children with cochlear implants were still demonstrating language growth rates of only 50-60% of the rate of children with normal hearing (Blamey et al., 2001a; Davis & Hind, 1999; Geers, 2002; Ramkalawan & Davis, 1992; Wake et al., 2004). Given the fact that these children were already delayed in their language development by the amount of time it had taken for diagnosis and implantation to occur, this slower rate of growth meant that by the time they were of school age, many children were delayed by at least 1 year, and approximately half had a severe language delay (ie. greater than 2 standard deviations below the mean for children with normal hearing). This rate of progress clearly has severe implications for academic achievement and functional literacy outcomes.

Over the past decade, with a decreasing average age at implantation and improved cochlear implant speech processing technology and hardware, language outcomes have further improved for children with cochlear implants, such that some children now acquire spoken language as do children with mild to moderate hearing loss (Spencer et al., 2011). More recently, several studies have shown that children who have received their cochlear implants at very young ages (and have had several years of experience) can achieve spoken language development at similar rates to children with normal hearing (Connor et al., 2006; Duchesne et al., 2009; Geers, 2006b; Schorr et al., 2008; Svirsky et al., 2004; Tomblin et al., 2005). For example, Dettman et al. 2008 reported that children implanted before the age of 2.5 years showed an average vocabulary development rate of 85% of that of children with normal hearing. This means that the gap between chronological age and language age for these children remains more constant, and for some diminishes instead of growing, as has commonly been reported in the past.

Greater proportions of children are showing age-appropriate development in receptive and expressive vocabulary (50% & 58%; Geers et al., 2009) and receptive and expressive language (47% & 39%; Nicholas & Geers, 2008) than previously. It has also been observed that some children with cochlear implants are even able to learn language more quickly than the average child with normal hearing and therefore 'catch up' some of the delay in language acquisition incurred before they received a cochlear implant, with reports of language development at age-appropriate levels between the ages of 4 and 7 years (Yoshinaga-Itano et al., 2010). As with speech perception and speech production, there is still enormous variation in language skills between individuals and between different populations of children (Spencer et al., 2003), with recent reports still documenting many children with significant language delays (Ching, 2010; Connor et al., 2000; Nikolopoulos et al., 2004; Sarant et al., 2009; Young & Killen, 2002).

The capacity for learning language in children with normal hearing is so great that they are not only able to develop fluency in their native language, but can also become fluent

speakers of more than three other languages without specific instruction. However, there have long been concerns that language delay in bilingual children is due to simultaneously learning two languages, due to the fact that learning a second language delays the learning process with the first language. It seemed logical that, for children with impaired auditory systems who are facing even greater challenges in language acquisition, learning two languages simultaneously would further delay the acquisition of the first language. More recently, it has been found that delays in vocabulary and slower progress in learning the second language dissipate in the early primary school years, and are likely to be due to the amount of exposure children have to each language (ie. the language that is used the most develops more quickly). It has also been demonstrated that language impairments found in bilingual children are due to individual children's innate capacity to learn language, and are not caused by simultaneous language learning (Genesee, 2001).

Despite the significant challenge inherent in mastering one spoken language with a cochlear implant, there is emerging evidence that it is also possible for children with cochlear implants to develop competence in a second spoken language. Robbins et al (2004b) reported on 12 children implanted before age 3 years, who not only demonstrated exceptional proficiency in their first language (almost all children had age-appropriate first language) but also solid progress in their second language over the 2 years during which they were followed. The children who were most proficient in their second language development had parents who spoke the second language at home, had opportunities to use the second language outside home, and had extensive cochlear implant experience. It was noted that, as a group, many of these children were 'ideal' cochlear implant recipients; half had hereditary deafness without additional disabilities, none had less than a full electrode array insertion, all had received intensive auditory-oral therapy prior to and after implantation, and none had meningitis-caused deafness. Two other studies have documented the ability of children with cochlear implants to develop competency in a second language. Of 18 children who received their cochlear implants by the age of 5 years and had a mean usage time of 4.5 years, the majority had achieved age-appropriate receptive and/or expressive language skills in their primary language, although their second language skills were still in the early stages of development (Waltzman et al., 2003). Uziel and colleagues (2007) also documented that some of the children in their study showed some ability to develop competency in a second language.

3.5 Social and emotional development

Children with profound hearing loss, including children with cochlear implants, are at increased risk for adverse life outcomes such as loneliness, poorer quality personal relationships, behaviour problems, drug and alcohol problems, and generally poorer quality of life than their normally hearing peers (Meadow, 1980; Watson et al., 1990). These problems can be attributed to a reduced ability to acquire many of the skills that underpin social functioning due to hearing loss (Marschark, 1993), despite their improved auditory capabilities. It is also important to note, however, that not all children with profound hearing loss and/or cochlear implants develop these problems. The impact of hearing loss on children's social and emotional development is also affected by several factors external to the children themselves, such as parental acceptance of and adaptation to their child's hearing loss, quality of family life, the ability of the family to cope, school and community

support, and resources (Calderon, 2000; Montanini-Manfredi, 1993). Of course, a child's personality and method of interacting with their social environment also contributes significantly. The few studies that examine the psychosocial development of children with cochlear implants show mixed results (Martin et al., 2011).

It has been reported that children with cochlear implants often have limited pragmatic skills, which can lead to poor social integration (Bat-Chava et al., 2005). Pragmatic skills include using language for different purposes (eg. greeting people, requesting information, demanding information), being able to change language according to the situation or listener (eg. speaking to an adult versus a toddler), and following conversational rules (eg. turn-taking in conversations, using facial expressions and eye contact, rephrasing when misunderstood). Children with poor pragmatic skills may say inappropriate things during conversations, may show little variety in the language they use, or may relate stories in a disorganised, illogical way. These behaviours often lead to a higher incidence of communication breakdown, and can lower social acceptance, as many children may choose to avoid having uncomfortable interactions with others who have pragmatic difficulties. Pragmatic problems are often related to delayed language development, which may include a limited vocabulary, and deficits in knowledge of grammar and age-appropriate slang.

It is not uncommon for children with severe-profound deafness to demonstrate significantly reduced emotional development and social maturity (Bat-Chava et al., 2005; Hintermair, 2006). These children also report loneliness, a lack of close friendships and other psychosocial difficulties more frequently than do their normally-hearing peers (Most, 2007; Stinson & Whitimire, 2000), and some studies show that this is the same for some children with cochlear implants (Boyd et al.,2000; Dammeyer, 2010; Leigh et al., 2009). Older children with cochlear implants (aged 9-14 years) are generally more affected by loneliness than younger children (aged 5-9 years), with children who receive their implants when older being most affected (Schorr, 2006). This may reflect the fact that social interaction becomes increasingly complex in adolescence, and peer group size tends to increase at this time, making communication more difficult due to increased acoustic and social challenges (Bat-Chava & Deignan, 2001; Martin et al., 2011).

Unsurprisingly, loneliness and psychosocial difficulties are greatest for children with additional disabilities, particularly those with low speech intelligibility and poor communication skills, as this increases communication breakdown and results in poorer peer attitudes towards children with these difficulties, who may be rejected or ignored by their peers (Dammeyer, 2010; Hintermair, 2007; Most, 2007; van Gent et al., 2007). Conversely, other studies have found no increased incidence of loneliness and psychosocial difficulties in children with cochlear implants compared to children with normal hearing (Percy-Smith et al., 2008a; Schorr, 2006), and children have been observed by parents to have improved communication skills and social relationships as a result of cochlear implantation (Archbold et al., 2008b; Bat-Chava & Deignan, 2001; Bat-Chava et al., 2005; Huber, 2005; Huttunen & Valimaa, 2010). Children with cochlear implants have been reported to be more likely to be acculturated to hearing society than those with a severe-profound hearing loss using hearing aids (Leigh et al., 2008a).

A statistically significant association has also been found between the level of social well-being in children with cochlear implants and their speech perception, production and

language skills (Dammeyer, 2010; Percy-Smith et al., 2008b). Social development usually follows language skill development, and improvements in both have been observed to occur more quickly for children with cochlear implants than for children with severe-profound hearing loss using hearing aids (Bat-Chava et al., 2005). It has been suggested that improved spoken language and communication skills facilitate psychosocial development through an ability to communicate and a subsequent increase in confidence (Bat-Chava & Deignan, 2001). Children with severe-profound deafness have historically been found to have lower levels of self-esteem than their peers with normal hearing (Nicholas & Geers, 2003), with the self-esteem of adolescents being lower than that of younger children (Schorr, 2006). It has been suggested that unless their language skills match those of their hearing peers, children with cochlear implants cannot fully integrate into the hearing community and develop positive self-esteem (Crouch, 1997; Lane & Grodin, 1997). However, as with many recent outcomes for children with cochlear implants, more recent research has shown equivalent levels of self esteem in children with cochlear implants and children with normal hearing (Loy et al., 2010; Martin et al., 2011; Sahli & Belgin, 2006).

Recent studies have also used measures of health-related quality of life (QOL) to investigate psychosocial development in children with cochlear implants, using both parental and child reports. QOL is considered to be an assessment of well-being across various areas of life such as social interaction, school adjustment, friendships, communication, and listening ability. Although a potential limitation of QOL measures can be that although parents are well-informed of their children's level of physical functioning, they have a tendency to underestimate their psychosocial functioning (Zaidman-Zait, 2011), QOL assessments are still regarded as a useful method of obtaining a more holistic measure of benefit. Loy and colleagues (2010) found no significant differences between overall reported QOL for children with cochlear implants compared to their peers with normal hearing, in either younger (8-11 years) or older (12-16 years) groups, although the younger group rated QOL more highly than did the adolescent group. Others have reported similar findings for children of various ages (Huber, 2005; Warner-Czyz et al., 2009).

Several factors have been found to influence psychosocial development in children with cochlear implants. Children who are implanted earlier and therefore have a longer duration of implant use are reported to be more socially competent (Leigh et al., 2008a; Martin et al., 2011), with girls outperforming boys (Martin et al., 2011; Nicholas & Geers, 2003; Percy-Smith et al., 2008b). As mentioned earlier, children implanted at older ages appear to be at greater risk of loneliness (Schorr, 2006), and it has been suggested that this may be due to the fact that they do not develop feelings of belonging and inclusion at a young age, as do children with normal hearing, due to their delayed language prior to implantation. It is also reported that children with cochlear implants in mainstream educational settings who are exposed to spoken, rather than signed, language at home have a higher level of social well-being (Percy-Smith et al., 2008b; van Gent et al., 2007). This may be because children in these settings are more likely to have hearing parents, and therefore are continuing to speak their first language in these settings, rather than using sign language at home and spoken language at school, as would children of many deaf parents. There is also no evidence that children with cochlear implants in mainstream educational settings, where speech is used exclusively for communication, have an increased incidence of social or emotional difficulties compared to children in special educational settings (Filipo et al., 1999; Nicholas & Geers, 2003; Percy-Smith et al., 2008b).

Once again, there is enormous variability between individuals in their communication and social development after cochlear implantation, with some children progressing at, or even above, the average rate of children with normal hearing, and others who lag behind their peers. Although there appear to be no negative reports on social/emotional development of children as a result of cochlear implantation, a cochlear implant will not guarantee that the social difficulties experienced by many children with severe-profound hearing loss are avoided (Punch & Hyde, 2011). The research does offer hope, however, that an early cochlear implant may not only facilitate the development of speech and language skills, but can also give children the potential to develop a healthy and positive social identity and competent interactional skills.

3.6 Literacy and academic outcomes

With documented improvements in speech perception, production and language outcomes clearly attributable to the improved auditory access provided by cochlear implants, there has been an expectation that academic outcomes for children with cochlear implants would also improve, with implanted children showing significantly better performance than their peers with hearing aids. However, although the proportion of children with cochlear implants who are enrolled in mainstreamed education settings is increasing steadily (Geers & Brenner, 2003), the degree to which cochlear implants have impacted on academic outcomes in children with severe-profound hearing loss is not yet clear. Much of the research on children with hearing loss is limited mainly to studies of reading ability, and few children who have received cochlear implants at a young age are currently old enough for longer-term outcomes to be measured.

Many children with severe-profound hearing loss, including those with cochlear implants, have 4 to 5 year delays in spoken language development by the time they enter secondary school (Blamey et al., 2001a; Dahl et al., 2003; Davis & Hind, 1999; Ramkalawan & Davis, 1992; Sarant et al., 2009). Generally, the greater the degree of hearing loss, the larger the language delay (Boothroyd et al., 1991). It is well known that poor spoken language ability is a primary cause of difficulty in learning to read for children with normal hearing, and it is therefore unsurprising that literacy achievement for children with hearing loss has historically been low, with many children failing to progress in reading beyond the identification of a limited number of words, or the fourth grade level of primary school (Geers et al., 2008; Moeller et al., 2007). Reported rates of progress have varied from 1 to 6 months for every year of education, with the delay in reading widening in adolescence (Geers et al., 2008; Thoutenhoofd, 2006). A significant proportion of graduating students with hearing loss are functionally illiterate (Helfand, 2001; Moeller et al., 2007; Traxler, 2000; Walker et al., 1998), having not even acquired mastery of spoken language, which is necessary not only for the development of literacy but also for the development of literate thought (Paul 1998). Low literacy achievement and low academic outcomes have seriously impacted on the ability of many children with hearing loss to obtain employment as adults, with resulting low skill employment and reduced income for some, and others simply not having sufficient literacy skills to succeed in the workplace at all.

One of the key language skills required for learning to read is vocabulary, which is often limited in children with hearing loss due to phonetic and phonological delays (Connor & Zwolan, 2004; James et al., 2008; Johnson & Goswami, 2010; Moeller et al., 2007; Moores &

Sweet, 1990). Phonological processing occurs when a child analyses words into their constituent parts, repeats strings of syllables that form new words, or quickly names common words. These processing abilities enable word decoding to occur, which in turn facilitates word recognition and comprehension of word meaning. Delayed phonological awareness, and a subsequently delayed vocabulary, makes it difficult to learn to read. Further compounding this difficulty is the fact that reading is a skill that must be learned through explicit instruction, some of which may be 'missed' due to compromised perceptual abilities caused by hearing loss, and also through an inability to understand some of the instruction due to poorer language skills (Moeller et al., 2007). It has been shown, however, that vocabulary development accelerates after cochlear implantation (Connor et al., 2006; Dawson, 1995; Geers et al., 2007; Johnson & Goswami, 2010; Nicholas & Geers, 2008), although there are conflicting reports regarding whether vocabulary growth rates slow over time, particularly for children who received their cochlear implants at older ages (El-Hakim et al., 2001) or remain constant (James et al., 2007). There is wide variability in vocabulary development between children (Connor et al., 2000), and long-term follow up of some children in their teen or early adult years still documents many children not having attained age-appropriate vocabulary (Uziel et al., 2007).

Reading outcomes to date for children with cochlear implants are promising, with evidence that children with cochlear implants are often achieving better reading outcomes at a faster rate than their peers with hearing loss who use hearing aids (Marschark et al., 2007), although many children are still significantly delayed. The number of children with cochlear implants who achieve age-appropriate reading skills is increasing (Geers, 2002; 2003). It has also been documented that almost 4 times as many children who have used a cochlear implant for at least 2 years have achieved a reading level beyond that of fourth grade compared to children with severe-profound hearing loss of similar ages using hearing aids (Spencer et al., 2003; Vermeulen et al., 2007). Higher levels of reading performance have been documented for girls than for boys (Moog & Geers, 2003), as has been observed in children with normal hearing. As with normally-hearing children, the factor that most affects reading outcome is language ability (Connor & Zwolan, 2004; Geers, 2003; Johnson & Goswami, 2010; Spencer et al., 2003), with children who are more competent in producing an oral narrative attaining better reading comprehension skills (Crosson & Geers, 2001). Cognitive ability (Geers & Hayes, 2011), speech intelligibility and speech perception ability have also been shown to be strong predictive factors of reading outcomes (Geers, 2003; Johnson & Goswami, 2010; Spencer & Oleson, 2008).

There is increasing evidence that some children with cochlear implants can not only acquire better reading outcomes than their peers with hearing aids, but can even achieve similar outcomes to their peers with normal hearing (Archbold et al., 2008a; Spencer et al., 2003; Spencer & Oleson, 2008). James and colleagues (2008) reported that children implanted between the ages of 2 to 3.6 years achieved reading scores that were within one standard deviation of the hearing normative mean, scoring higher than children implanted between ages 5 and 7 years. Geers and Hayes (2011) also documented 47-66% of adolescents who received their implants as pre-schoolers achieving reading abilities within the average range for their hearing peers. Other studies have reported similar results, with 70%, 61%, and 51% of children reading within age-appropriate levels (Moog, 2002; Geers, 2003, Johnson & Goswami, 2010 respectively).

Other studies have shown that although early cochlear implantation facilitates improved reading outcomes in terms of both decoding and reading comprehension, a significant number of children are still not reading at the same level as their normally-hearing peers, and are falling behind over time (Archbold et al., 2008a; Connor & Zwolan, 2004). Geers and colleagues showed that only 44% of secondary school students showed age-appropriate reading performance, compared to 56% of the same group when in primary (elementary) school (Geers et al., 2008). Although the group of children was reading, on average, at an age-appropriate level when aged 8-9 years, the same children were delayed on average by almost 2 years in their reading by age 15-16 years. More recently, Geers and Hayes (2011) also reported that although 72% of the adolescents in the same sample had retained their reading standing in comparison with hearing peers since primary school, (demonstrating age-appropriate growth in reading skills over that time), 60% were still delayed overall. For many children, the reading gap between children with cochlear implants and their peers with normal hearing still widens as they grow older. Some studies still report that some children still do not make any progress at all (James et al., 2008).

Studies of writing in children with hearing loss have evaluated syntax (or grammar), looking specifically at complexity, productivity and grammaticality. The writing of children with hearing loss has generally been found to be composed of shorter sentences than those used by their hearing peers (Kretchmer & Kretchmer, 1986), repetitive phrasing, and many subject-object-verb constructions (Lichtenstein, 1998; Wilbur, 1977). There are also many errors of omission, substitution and word addition (Myklebust, 1964), including the omission of articles, prepositions, copulas, pronouns and conjunctions (Crosson & Geers, 2001). Lichtenstein also noted many errors of morphology such as plurality, verb agreement and tense in the writing of children with hearing loss. It has been concluded that children with hearing loss have even greater difficulties with writing than with learning to read (Paul, 1998).

During the primary (or elementary) school years, early writing patterns appear to follow those of spoken language development (ie. children write as they would speak). As their writing skills develop, they use more sophisticated forms of language so that their writing becomes more "detached" from their spoken language (Spencer et al., 2003). Children with cochlear implants are reported to persist in the documented pattern of immature writing skills, with shorter, less complex sentences containing more errors reported for a group of 9-year-old children using cochlear implants (Spencer et al., 2003). In this study, correlations between language abilities and writing productivity suggested that the children had not yet 'detached' their written from their spoken language. Geers and Hayes (2011) also documented the poor spelling and writing skills of children with cochlear implants compared to their peers with normal hearing. Children in this study continued to struggle with phonological processing tasks, and performed at delayed levels on measures of phonological awareness, expository writing, and spelling.

Academic success relies on reading and writing abilities, and there is now a body of work focused on literacy in children with cochlear implants. However, information on overall academic performance of these children is scarce. Spencer and colleagues (2004) examined academic achievement in science, social studies and humanities in young adults with cochlear implants, finding that consistent users of cochlear implants performed comparably to their hearing peers, achieving an overall mean standard score of 103.88 on the relevant

subtests of the Woodcock-Johnson Tests of Achievement (the expected average score for children with normal hearing would be 100). This study is novel because it is the only report of fully comparable academic performance for children with cochlear implants. A more recent report on educational and employment achievements in France showed that although 42-61% of the children had failed one grade (or year level) at school (a higher rate of failure than for children with normal hearing), over 60% of those aged 18 years and over either held a university degree and/or were employed at levels similar to those of their peers with normal hearing. These figures were reported as being very similar to those for the general population of France, where 53% of individuals have at least a high school diploma (Venail et al., 2010). A third study of Malaysian children reported that for children implanted relatively late (aged 3-4 years), 56% performed below the average level academically, with greatest achievement in mathematics rather than language (Mukari et al., 2007).

As with other areas of development, wide variability in literacy and academic outcomes has been reported. As children are implanted at younger ages and enter school with better language skills, it is likely that future research will show a further narrowing of the gap in literacy and academic performance between children with cochlear implants and children with normal hearing. However, although many younger children are reported to be performing at age appropriate levels, some studies suggest that this level of performance is not sustained long-term by all children. Currently, the effect of cochlear implants on the long-term academic outcomes of children appears promising, but unclear.

4. Factors affecting speech perception, production and language outcomes

Despite the significant improvements made in cochlear implant technology, and the large body of clinical knowledge gained over time regarding likely benefits for children with cochlear implants, one of the remaining significant challenges is to identify predictors of post-implant outcomes, as there is great variation in benefits between individuals. Several factors have currently been identified as influential in children's speech perception, speech production, language and academic development after implantation, and the most important of these are discussed below.

4.1 Age at diagnosis

With the establishment of newborn hearing screening in many developed countries around the world, the average age of diagnosis of hearing loss in these countries has dropped to 12-25 months, with many babies identified as young as 3 months of age (Dalzell et al., 2000; Harrison et al., 2003; Watkin et al., 2007). As mentioned previously, the earlier identification of hearing loss has resulted in a rapid rise in the numbers of children receiving cochlear implants at younger ages (ASHA, 2004). It was estimated that the number of children receiving cochlear implants before the age of 2 years between 1991 and 2002 increased forty fold (Drinkwater, 2004), and it is likely that this growth rate has not declined. However, there are still many children in developed countries who are not receiving cochlear implants early in life. It is disappointing to note that despite earlier identification of hearing loss through newborn hearing screening programs, many families (and almost half of the families in the U.S. who are referred for further hearing assessment of their newborn babies) still do not receive early intervention services by the age of 6 months, as is recommended by the 2007 Position Statement of the Joint Committee on Infant Hearing (JCIH, 2007). The

reasons for this are varied, and include a lack of understanding of the importance of early identification and intervention, problems with follow-up systems, lack of access to appropriate services and other issues related to babies' health (Sass-Lehrer, 2011). It is also reported that around one third of pediatric implant recipients who passed the newborn hearing screening assessment subsequently become implant candidates through progressive hearing loss in the first years of life due to genetic causes such as the Connexin 26 mutation, Usher Syndrome, or to other causes such as auditory neuropathy or congenital Cytomegalovirus (CMV) (Young et al., 2011), and these children also receive cochlear implants when older.

Although age at diagnosis has been reported by many studies to be an influential factor in outcomes for children with cochlear implants, some studies have not found this link (Geers et al., 2009; Geers, 2004; Harris & Terlektsi 2011; Sarant et al., 2009; Wake et al., 2005). Two of these studies included a greater proportion of children who were diagnosed late and were therefore implanted at older ages, reporting poorer performance than other studies of children whose hearing loss was identified earlier. Nicholas and Geers (2006) reported that age at diagnosis was not a significant predictive factor in language outcomes unless it led to children receiving a cochlear implant before 24 months of age. Evidence that age at diagnosis is an important factor has become stronger as children receive cochlear implants at younger ages. Several studies have reported excellent speech perception abilities and age-appropriate language outcomes for many young children who were diagnosed with hearing loss in the first six months of life (Apuzzo, 1995; Yoshinaga-Itano, 2003b; Yoshinaga-Itano et al., 1998), and there is mounting evidence that early-diagnosed children are developing language at a faster rate than their later-diagnosed peers (Connor & Zwolan, 2004; Kennedy et al., 2006).

4.2 Age at implant/duration of profound deafness

Age at implantation is often quite close to time of diagnosis early in life due to newborn screening. For children with congenital hearing loss, 'age at implant' is equal to 'duration of deafness'. Many human and animal studies of the development of the neurosensory pathways of the primary auditory cortex in the brain have suggested that the plasticity, or potential for development, of neural pathways is greatest during early development, and that there is therefore a 'critical period', during which auditory stimulation must occur in order for neural maturation to occur (Kral et al., 2001; Sharma et al., 2002). If stimulation does not occur within this timeframe, the auditory system degenerates (Kral et al., 2001; Shepherd, 1997). In humans with normal hearing, maturation of the central auditory system continues throughout childhood through to adolescence. Research with humans has shown that the central auditory system can retain its plasticity for some years without auditory input, and when stimulated by a cochlear implant will commence maturation at the same rate as for children with normal hearing, with the maturational sequence delayed by the period of sensory deprivation (Ponton et al., 1996).

It has been found, however, that after long periods of deprivation, such as in children who have used a unilateral implant for several years and have then received a second, bilateral implant, that there were abnormalities in spatial patterns of cortical activity in the brain not observed in children who received a second cochlear implant after a shorter time (Gordon et al., 2010). Further physiological studies suggest that in the absence of normal auditory

stimulation there is a period of about 3.5 years during which the central auditory system retains its maximum plasticity. This can extend in some children up to the age of approximately 7 years, after which it is significantly reduced (Sharma et al., 2005; Sharma et al., 2002). Harrison and colleagues (2005), who examined the speech perception performance of children implanted at different ages, argue that the situation is not quite as simple as this. They hypothesize that although central auditory plasticity is limited for children implanted at older ages, there is no age at which there is a clear cut-off, but instead there is an age-related plasticity effect that depends to some extent on the tests used to assess performance.

Early research on cochlear implantation in children supported the biological plasticity theory, showing a strong negative relationship between duration of deafness (or age at implant) and speech perception outcomes (Apuzzo, 1995; Nikolopoulos et al., 1999; Osberger, 1991; Staller et al., 1991). Initially, speech perception results for children who were not congenitally deaf, received their cochlear implant relatively quickly, and therefore had a shorter period of deafness, were superior to those for children with congenital deafness and later implantation (Pisoni et al., 1999; Staller et al., 1991). As Marshark (2007) noted, children who have later onset hearing loss have usually developed better language skills prior to implantation, and therefore show better achievement afterwards (for example, Moog & Geers, 2003). For children with congenital deafness, a significant correlation between age at implantation and outcomes has also been documented in many recent studies. Children implanted earlier show faster growth of speech perception (Tajudeen et al., 2010; Uziel et al., 2007), language (Connor et al., 2000; Nikolopoulos et al., 2004; Schorr et al., 2008; Tomblin et al., 2005) and reading abilities (Archbold et al., 2008a; Geers et al., 2008; James et al., 2008; Johnson & Goswami, 2010), and also have improved psychosocial outcomes (Schorr, 2006). Development of speech production is also associated with age at implantation, with slower rates of development shown by children who received their implants later (Flipsen, 2008; Peng et al., 2004; Tye-Murray et al., 1995). Interestingly, for children implanted very early, early age at implantation and speech production have been observed to have the opposite association, with one study documenting slower vocal development for children implanted when younger. Greater physical, cognitive, and social maturity were thought to provide children implanted at older ages with an advantage for early speech development (Ertmer et al., 2007).

More recently, there have been reports of even better outcomes in children implanted around the age of 2 years or younger, with higher proportions of children achieving speech perception, language and reading skills commensurate with those of their hearing peers (Duchesne et al., 2009; Geers, 2004; Niparko et al,. 2010; Svirsky et al., 2004). These results have been observed to be "consistent with the existence of a 'sensitive period' for language development, and a gradual decline in language acquisition skills as a function of age" (Svirsky et al., 2004). Svirsky and colleagues qualify this observation by suggesting that the auditory information provided by a cochlear implant is significantly inferior to that received by children with normal hearing, and that it is possible that sensitive periods for speech and language development may exist for cochlear implant users and not for children with normal hearing because of the diminished auditory signal the former receive.

Nicholas and Geers (2007) studied the language development of 76 children who had received a cochlear implant by their third birthday. They concluded that children who received an implant by 12-16 months, before substantial spoken language delay had

developed, were more likely to achieve age-appropriate spoken language. These children 'caught up' with their hearing peers by 4.5 years of age, whereas children implanted after 24 months of age did not. Both Nicholas and Geers (2007) and Tomblin and colleagues (2005) observed an early burst of language growth in children implanted before the age of 18 months which was not seen in children implanted after this age. More recent studies suggest implanting children as early as before 12 months of age, with strong development of speech perception and language skills reported at age-appropriate rates for many or all of the children (Svirsky et al., 2004; Tajudeen et al., 2010; Waltzman & Roland, 2005; Wie, 2010).

A review of recent studies concluded that the evidence suggests that cochlear implantation before the age of 2 years is more effective than after this time, but that it is not yet clear whether implantation of children under 12 months of age provides greater benefit (Ali & O'Connell, 2007). As implantation of children under the age of 2 years is a relatively recent practice, limited evidence has been obtained for short-term outcomes (only up to approximately 5-8 years post-implantation) and the effect of implantation at a very young age on longer-term outcomes is still unknown (Ali & O'Connell, 2007). It is also not yet known whether children implanted at older ages, who have been shown to develop more slowly, will eventually reach equivalent long-term milestones to those implanted earlier. Some more recent longer term studies support this view, showing that although age at implantation strongly influences outcomes in younger children, the effect of this factor appears to wane with increasing age and implant experience (Geers, 2004; Hay-McCutcheon et al., 2008; Moog & Geers, 2003). Finally, when considering these reports, it is also important to remember that children implanted at younger ages are more likely to use oral communication, a factor that has also been shown to improve speech perception and spoken language outcomes.

4.3 Degree of hearing loss

There is conflicting evidence regarding the influence of degree of hearing loss on outcomes for children with cochlear implants. This factor has been reported as highly predictive of outcomes for children with cochlear implants in many studies. Speech perception abilities, language development and reading in children with hearing loss and those with cochlear implants have been found to decrease with increasing severity of hearing loss (Boothroyd et al., 1991; El-Hakim et al., 2001; Holt & Svirsky, 2008; Wake et al., 2005; Zwolan et al., 1997). Nicholas and Geers (2007) observed that children with better hearing prior to implantation showed faster language growth with increasing implant experience than did children with less pre-implant hearing. Conversely, some other studies that included more children who were older when implanted and at testing have not found a significant correlation between degree of hearing loss and speech perception, vocabulary or speech production outcomes (Blamey et al. 2001a; Harris & Terlektsi, 2011). The majority of published evidence supports a significant influence of degree of hearing loss on outcomes.

4.4 Cognitive ability

Non-verbal cognitive ability has been identified as one of the most influential factors on language outcomes in preschool children with hearing loss. The influence of cognitive skills is no less important for outcomes in children with cochlear implants, and several studies

have reported it to be one of the most significant factors of all those examined, having much greater influence than other variables (Geers et al., 2009; Geers, 2003). Non-verbal IQ has been shown to have a significant effect on the development of vocabulary (Mayne, 2000), language (Geers et al., 2009; Geers et al., 2008; Sarant et al., 2009; Sarant, Hughes, & Blamey, 2010), reading (Moog & Geers, 2003), and speech production (Tobey et al., 2003). Although, after adjusting for the effect of language, cognitive ability usually has no direct effect on speech perception performance, it does have an indirect effect on this outcome. This is because language is strongly influenced by cognitive ability, and is the medium through which speech perception assessments are conducted; children have to comprehend the language used in speech perception tests and respond using spoken language (Sarant et al., 2010). Many studies have demonstrated a strong association between language and speech perception ability (for example, Blamey et al., 2001; Niparko et al., 2010).

Cognitive delay has been associated with reduced development of speech perception and production skills in populations of children with diagnosed additional disabilities (Holt & Kirk, 2005; Pyman et al., 2000; Waltzman et al., 2000), but is also a predictive factor for children who are in the average range for non-verbal cognitive abilities (Moog & Geers, 2003). Pisoni and colleagues emphasized the importance of cognitive factors such as memory, attention, and verbal rehearsal speed in determining outcomes after implantation (Pisoni & Cleary, 2003; Pisoni et al., 1999), and postulated that 'central' cognitive factors might explain some of the previously unexplained variance in outcomes for children with cochlear implants (Pisoni & Cleary, 2003; Pisoni et al., 1999). Geers and Sedey (2011) added credence to this theory with their recent observation that faster verbal rehearsal speed contributed to better language outcomes in children implanted between 2 and 5 years of age with more than 10 years of cochlear implant experience. In further support of Pisoni and colleagues' theory, it has recently been reported that when compared to children of the same age and cognitive ability, children with cochlear implants still demonstrate language delays that are disproportionate to their cognitive potential (Meinzen-Derr et al., 2011). The cognitive processes underlying this performance-functional gap need to be investigated and understood in order to implement appropriate intervention strategies to close the gap and improve outcomes for a greater proportion of children with cochlear implants.

4.5 Communication mode

Communication mode, often dichotomized into oral communication and total communication (signing plus speaking), has long been investigated as a source of variance in outcomes for children with cochlear implants, with mixed results. Proponents of oral communication maintain that maximal auditory benefit from cochlear implants can only be gained if hearing and speech are the only media for communication. There are several reports of children attending oral communication programs achieving higher speech perception and language scores than children in total communication programs (Archbold et al., 2000; El-Hakim et al., 2001; Geers et al., 2003; Meyer et al., 1998; Moog & Geers, 2003). Similarly, speech production outcomes are reported to be better for children in oral education settings. Tobey et al (2003) found oral-aural communication and teaching methods that emphasized speaking and listening to be the most influential factors in determining speech production development in children implanted by age 5 years. These

environments were found to enhance speech production development, regardless of whether the environment was a mainstream school or a special school, although children in mainstream environments outperformed those in special education environments.

Proponents of the total communication approach maintain that children will obtain maximal information through the use of both speech and some form of manually coded English, as the latter will provide information that may be missed due to insufficient auditory abilities. Improved vocabulary development has been documented for children implanted early and enrolled in total communication educational programs over those in oral programs (Connor et al., 2000). There are also reports that mode of communication does not significantly influence some outcomes. Yoshinaga-Itano and Snyder (1996) found that mode of communication and learning did not significantly affect students' performance in the lexical/semantic characteristics of their written language. They hypothesized that written language is acquired in such a way that students need only one well-established language in order to acquire the written form of their language, and that both oral and signed communication methods may provide students with sufficient bases from which to learn written English. Similarly, several studies of speech perception, production, language, reading and later academic outcomes of children with cochlear implants have not found oral or total communication modes to be predictive of better results (Geers, 2003; Miyamoto et al., 1993; Niparko et al., 2010; Robbins et al., 1999; Uziel et al., 2007).

The absence of overwhelming evidence of the superiority of one communication method over the other may be due to differences in the characteristics of the children studied. Children who are implanted at younger ages are more likely to use an oral communication method, and particular educational programs may also have selection biases towards children with characteristics such as greater preoperative residual hearing or higher cognitive ability (Geers, 2006a). Some non-government funded educational programs are not accessible to families of lower socioeconomic status, and in this way only children from families with greater financial means and likely higher educational achievements will be enrolled in particular programs. When considering the effect of mode of communication, it is unclear in many cases whether children use oral communication after cochlear implantation because they are progressing well, or whether their rate of progress is due to their use of oral communication.

4.6 Family characteristics

Several family characteristics have been found to contribute to various outcomes for children with hearing loss, including those with cochlear implants. Family size has been observed to impact on speech production outcomes, with children from smaller families making faster progress (Moog & Geers, 2003; Tobey et al., 2003). This is presumably due to the fact that parents of smaller families may have more time and/or resources to devote to assisting their children's communication development. Similarly, children from families of higher socioeconomic status have achieved better speech production, language and literacy outcomes (Connor & Zwolan, 2004; Dollaghan et al., 1999; Holt & Svirsky, 2008; Niparko et al., 2010; Tobey et al., 2003). Greater parental involvement in children's intervention programs has also been associated with improved language development (Moeller, 2000; Sarant et al., 2009; Watkin et al., 2007). This is presumably due to increased follow-up and improved communication at home, as parents who become involved in intervention have

been shown to demonstrate better communication skills and make higher contributions to children's progress than non-participating parents (Fallon & Harris, 1991).

Unsurprisingly, maternal communication skills are also a significant indicator for language development, early reading skills, and psychosocial development, with children of mothers who are better communicators developing better reading and language skills and having fewer behaviour problems (Calderon, 2000; Niparko et al., 2010). Children with a more highly educated parent caregiver have been reported to have better language, even in studies where the average educational level was relatively high (Geers et al., 2009; Sarant et al., 2009). It has been suggested that the relationship between socioeconomic status and language outcomes is actually mediated solely by properties of maternal speech that differ as a function of socioeconomic status (Hoff, 2003; Hoff & Tian, 2005). Gender also contributes to the variation in outcomes between children, with females consistently achieving better results with regard to speech production (Tobey et al., 2003), reading (Moog & Geers, 2003) and language development (Geers et al., 2009).

4.7 Other factors

Cochlear implant and speech characteristics such as the number of active electrodes in the implant array, larger dynamic ranges in speech processor maps, greater growth of loudness and length of time using the latest speech processing strategies have been found to significantly influence speech production and language outcomes in children implanted by age 5 years (Connor et al., 2000; Moog & Geers, 2003; Peng et al., 2004; Tobey et al., 2003). The number of surviving nerves has also been postulated to contribute to outcomes (Pyman et al., 2000).

5. Limitations in outcomes with unilateral cochlear implants

Historically, the consequences of unilateral hearing loss (UHL) have been underestimated, both for children with normal hearing and those with a unilateral cochlear implant, as spoken language can still be developed with one hearing ear. Prior to the introduction of neonatal hearing screening, many children with UHL were undiagnosed until they attended school, where communication difficulties in noisy educational environments or failure to progress academically at the expected rate raised suspicions of hearing loss. Although there has been limited research on the effect of UHL on the development of spoken language, mild through to significant delays have been reported in several studies of children with UHL and normal hearing in the unimplanted ear, although there has been insufficient follow-up to determine whether the reported delays persisted through childhood (Cho Lieu, 2004). A review of the literature in this area also found that school-aged children with UHL have increased rates of academic failure (22-35% rate of repeating at least one grade), additional needs for educational assistance (12-41%), and behavioural problems in the classroom (Cho Lieu, 2004).

Despite the fact that many children with a unilateral implant demonstrate excellent speech perception abilities in the controlled testing environment of a sound proof booth (Cheng et al., 1999; Leigh et al. 2008c; Sarant et al., 2001), this performance does not represent their speech perception abilities in the real world. The difficulties experienced by children with one normal hearing ear and one ear with UHL are similar (but worse) for children with a

single cochlear implant and a severe-profound or profound hearing loss in the non-implanted ear. These include difficulty understanding speech that is soft, or speech in noisy environments, such as the playground or classroom, and difficulty locating sound sources, such as their peers in a group conversation, or their teachers in the classroom. These auditory challenges can limit their ability to follow or take part in a group conversation, or to focus in the correct direction when the teacher begins to speak. The amount and quality of speech heard by children with one cochlear implant and a significant hearing loss in the other ear is greatly reduced and fragmented compared to what is heard by children with normal hearing. Further, understanding what they do hear is made difficult by their often delayed language skills. With poor language knowledge, many of these children are unable to piece together poorly heard or overheard information, and therefore to learn incidentally (without direct teaching), as do children with normal hearing. The inability to 'overhear' spoken conversations limits the access of these children to many avenues of incidental learning, and therefore restricts their acquisition of knowledge of language, social interaction, and how the world works, stifling their development in many areas.

A unilateral cochlear implant does not guarantee the development of language, speech production, academic or social skills comparable to those of children with normal hearing. Although there are many children with a unilateral cochlear implant who are able to develop these skills at an age-appropriate rate, there also remain many who show delayed development in these areas, some of whom maintain or increase their delay through to adulthood. Given the difficulties of unilateral hearing loss, giving children bilateral cochlear implants could potentially improve outcomes.

6. Bilateral cochlear implants

A recent report on worldwide trends in bilateral cochlear implantation estimated that 59%of bilateral cochlear implant recipients in the U.S., and 78% of recipients in other countries are currently children (Peters et al., 2010). It was observed that by the end of 2007, 70% of all bilateral cochlear implants had been received by children, with children aged 3-10 years being most highly represented in this group (33% of all bilateral surgeries; Peters et al., 2010). 70% of children received 2 cochlear implants in sequential operations (2 separate operations). Of the remaining 30%, children aged less than 3 years were the only group for whom the majority (58%) received bilateral cochlear implants simultaneously (during the same operation). Bilateral cochlear implantation in children is a growing trend worldwide; in 2010, implant manufacturers' databases indicated that there were 4986 children with bilateral implants (Peters et al., 2010).

6.1

6.1.1 Decision making

The decision to give a child one or two cochlear implants is a difficult one for parents, despite the growing trend toward implanting children at a young age with simultaneous bilateral cochlear implants. Until recently, there has been a lack of strong evidence to support bilateral implantation, particularly with regard to longer term outcomes (Hyde et al., 2010). For parents of children with no useable residual hearing, the decision is more straightforward, as binaural hearing offers significant benefits over monaural hearing. However, parents of children with useable aided residual hearing face a more difficult

decision, as loss of functional and useful hearing is being risked for a probable, but not guaranteed, benefit. Parents usually take into account their child's degree of hearing loss in both ears (if the child has no cochlear implants) or in the non-implanted ear, professional recommendations, costs (typically between $US40,000 -$US60,000 (Papsin & Gordon, 2007), their own attitudes and desires for their child, and surgical/medical and other risks (see section 6.1.4). Parents of children who are deemed eligible by an implant team for bilateral cochlear implants may still choose to give their child a unilateral implant. Reasons for this decision have included a desire to see what the benefits of one implant are before proceeding with another, concerns about the appearance of children wearing two speech processors, saving an ear for future technological developments (see 6.1.3), and difficulty accepting children's hearing loss.

6.1.2 Physiological and functional arguments for bilateral cochlear implantation

The arguments for bilateral cochlear implantation include stimulation of both auditory nerves to ensure that the better ear is stimulated, as the benefits of cochlear implantation are not necessarily symmetrical for each ear. As previously discussed, many factors influence outcomes, and although some factors will be the same for both ears in a particular individual (for example, communication mode, cognitive ability etc.), others may not. These could include the anatomical structure and physiology of the ears, effects of the pathology that caused the hearing loss, and in the case of children who receive two cochlear implants separated in time (sequential implantation), the duration of deafness will differ between the ears. A further reason for bilateral cochlear implantation is to prevent the neural degeneration that has been documented in humans and animal studies as a result of auditory deprivation (Hardie, 1998; Sharma et al., 2002; Shepherd, 1997). Bilateral implantation also ensures that children still have hearing in the case of speech processor or device failure in one ear, which can significantly reduce stress for children and their families if these events occur. Finally, having bilateral cochlear implants may facilitate binaural hearing, which requires the perception of auditory information in both ears. As discussed earlier, children with unilateral cochlear implants experience the difficulties associated with unilateral deafness, such as an inability to localize sounds, and difficulty perceiving speech in background noise. For the relatively small number of children who have sufficient hearing to use a hearing aid in their non-implanted (or contralateral) ear, the literature shows that binaural benefit is gained through use of the cochlear implant and hearing aid together (Frush Holt et al., 2005; Mok et al., 2007). However, for many children with a bilateral profound or severe-profound hearing loss, the use of a contralateral hearing aid in the non-implanted ear is not a viable option, due to a lack of residual hearing. For these children, bilateral cochlear implantation is the only means of providing binaural hearing.

6.1.3 Access to future technology

Arguments against bilateral implantation include 'saving' an ear for future technology while using a hearing aid with residual hearing (if there is sufficient residual hearing). Although it is known that changes in the cochlea occur after implantation, and that these are permanent, it is not known whether repeated re-implantation with cochlear implants or with other future technology is possible after many years of cochlear implant use (although re-implantation is usually successful in the case of device failure). It is also unknown if or

when future technologies such as gene therapy or neural regeneration will become available for clinical use, and it is accepted that there is a critical time period for central auditory brain and language development, beyond which future technology may not be beneficial. Without knowing what form future technologies may take, it is not possible to predict how useful they may be for individuals who have 'waited' and not proceeded with the current cochlear implant technology.

6.1.4 Risks

Many parents have concerns about the risks of cochlear implant surgery, and some of these risks are increased with two separate implant operations, as is the case with sequential implant procedures. Simultaneous implant operations require less than double the surgery time and eliminate the need for, and risks of, two anaesthetics and recovery periods. Complications as a result of cochlear implant surgery can be categorised as major and minor, and most occur very close to the time of surgery, although some have been reported up to 14 years post-surgery, and can recur. Major complications include infections or skin flap breakdown in the area around the implant, extrusion of the end of the electrode array outside the cochlea, device failure (requiring explantation of the device), cholesteatoma, permanent facial nerve damage, persistent eardrum perforation, cerebrospinal fluid leak with subsequent meningitis, and magnet displacement. For children with anatomical deformities of the cochlea (such as Mondini deformity, in which there are less than the normal two and a half turns in the cochlea), the risk of facial nerve damage is greater. However, reported major complication rates are very low, ranging from 2 - 5% (Bhatia et al., 2004; Cohen et al., 1989; Loundon et al., 2010).

Minor complications are those which can be resolved without surgery, and include vertigo with or without nausea, persistent otitis media (middle ear infection), facial palsy, tinnitus, mild skin flap infection, flap swelling, hematoma (bruising), taste disturbance, and pain around the operation site. The incidence of minor complications is higher and more subject to variation between cochlear implant centers; studies of large numbers of patients ranging from 4% - 20% (Bhatia et al., 2004; Dutt et al., 2005; Loundon et al., 2010). Other risks include those of any surgical procedure, including the risks associated with an anaesthetic and blood loss. Some risks are increased for younger children, including an increased risk of anaesthetic complications. A further risk is due to the relatively small size of their skulls. Although their cochleae are adult-sized at birth, their small skull size increases the risk of displacement of the electrode array with subsequent significant skull growth. There is also a high prevalence of otitis media in this age group, which raises the risk of significant infection in the implant area as a result of infection spread from the middle ear. Due to these concerns, the FDA currently approves cochlear implantation in children only from the age of 2 years and older (ASHA, 2004). In summary, although there are several possible complications of cochlear implant surgery, the incidence of life-threatening complications is extremely low, and the rates of major and minor post-operative complications are also low, making cochlear implant surgery in children a reliable and safe procedure.

Longer term risks of cochlear implantation include device failure. Although cochlear implants are designed to last for a lifetime, about 2% of devices do fail (ASHA, 2004). Device failure can result in a changed auditory percept or a total lack of function, and re-implantation is the only solution. Fortunately, most re-implants function as well as, or better

than, the original implants, but the risks, costs, and inconvenience of surgery must be undertaken. Another longer-term risk is the increased risk of bacterial meningitis, due to the fact that the cochlear implant is a foreign body, and can act as a nidus for infection when there is a bacterial illness (ASHA, 2004). This risk is highest for children with malformed cochleae, those who contract meningitis prior to cochlear implantation, children aged less than five years, and children with otitis media or immunodeficiency. A further longer-term complication is facial nerve stimulation, which can occur at any time after cochlear implantation, but is rare. Children most at risk of this are those with malformed cochleae. Fortunately, it is a simple procedure for an audiologist to switch off the electrode/s causing the unwanted sensation.

A final and important risk that is unique to sequential bilateral cochlear implantation is that some (usually older) children may not like the sound of their second cochlear implant, and will eventually become non-users. While many children, particularly those who have had one cochlear implant and have another after a significant period of time, may not initially like the sound of their second implant, most adapt to it over time with encouragement and support. However, some children never adapt, and show a pattern of inconsistent use over several years that culminates in rejection when they are older. There have been no reports in the literature to date about adaptation and non-user rates for either large groups of children with bilateral implants or for simultaneously implanted children. Factors thought to contribute to this outcome in children with a unilateral cochlear implant include older age at implantation, dislike of the auditory percept, facial nerve pain or twitching, peer pressure in secondary school, family issues, non-mainstream school settings, use of signed communication, lack of involvement in the decision-making process (older children), and poor speech intelligibility after several years of cochlear implant use (Archbold et al., 2009; Ray et al., 2006; Watson & Gregory, 2005).

Published information on the current non-user rate for children with unilateral cochlear implants suggests the risk of rejection is low; the reported non-user rate is currently around 3% (Archbold et al. 2009; Uziel et al. 2007). However, for children receiving a second, sequential cochlear implant, the situation is entirely different, as they must adapt to a second, different sound percept; one that may not compare favourably with that provided by their first cochlear implant. In the first study to be published on adaptation in children with bilateral implants, Galvin and Hughes (in press) noted that a higher proportion of children who were implanted simultaneously adapted to full-time use of their devices (95%) than those implanted sequentially (70%), and that adaptation to bilateral implant use was not easy for almost 20% of the 46 children studied. Both Galvin and colleagues, and Archbold (2009; in a study of long term use of unilateral cochlear implants in children) noted that children who eventually become non-users often first demonstrate a pattern of inconsistent use. Archbold also noted that children who became non-users usually had disabilities additional to their hearing loss. The possibility of this eventuality should be taken into account by parents, and also by children old enough to participate in the decision-making process.

6.2 Benefits of bilateral cochlear implants

When a person with normal hearing listens with two ears (rather than just one) sound quality is improved, it is easier to locate the source of a sound, and it is easier to understand

speech, particularly in background noise. The improved sound quality with two ears is commonly described as fuller, more spacious, and more natural. To locate sound sources, the listener primarily uses the differences in timing and level of sound arriving at each ear, with sound arriving later and being softer at the ear furthest from the sound source. This localization ability allows the listener to locate sounds in the environment, to find the speaker in a group conversation, and to be more aware of changes in their auditory environment. Speech perception is improved with two ears because the brain has two opportunities to process the same signal (binaural redundancy), and because the combined signal is slightly louder (binaural summation). The benefits of two ears are particularly significant when speech and noise are coming from different directions. Firstly, due to the physical barrier of the head (the head-shadow effect), the noise level will be lower at the ear that is furthest from the noise source. Given that speech will usually be arriving from in front of the head, the level of the speech signal is equal at both ears. The listener is therefore better able to perceive speech by attending primarily to the ear at which the noise level is lower. Secondly, with speech and noise coming from different directions, each ear receives a different balance of speech and noise. The brain is able to compare these two different signals and reduce the impact of the noise to increase the salience of the speech signal (binaural unmasking).

6.2.1 Speech perception

The speech perception abilities of children with bilateral cochlear implants have been explored using both standardized measures and a variety of study-specific measures in quiet conditions and in various noise conditions (for example, Galvin et al., 2007a, b; Scherf et al., 2007; van Deun et al., 2010). A review of the research found that 11/13 of the studies reported significant improvement in children's speech perception in noise abilities (Johnston et al., 2009). Some of these improvements were due simply to the head shadow effect, or to the ability to concentrate on the sound from one ear over another (Galvin et al., 2008a; Galvin et al., 2007a; Litovsky et al., 2006a). A recent study found that although they did not perform as well as children with normal hearing, bilaterally implanted children performed significantly better than unilaterally implanted children on tests of speech perception performance in noise (Lovett et al., 2010). As with outcomes for children with unilateral cochlear implants, the degree of improvement varies widely between individuals. Improved speech perception in noise has been associated with shorter periods of hearing loss in the second ear in some studies (Litovsky et al., 2004; Peters et al., 2007; Steffens et al., 2008), but not all have found this link (Kuhn-Inacker et al., 2004; Litovsky et al., 2006a; Wolfe et al., 2007). Two studies that did not find improvements in speech perception in noise included children who had a long time period between their first and second cochlear implants. There have also been reports of improved speech perception performance in quiet conditions with bilateral implants (Scherf et al., 2007; Zeitler et al., 2008).

6.2.2 Localization of sound

Bilateral cochlear implantation has not yet shown a clear benefit for sound localization. In assessments of localization performance for long-term users to date, some children can localize sounds well (Litovsky et al., 2006b; Lovett et al., 2010). Bilateral implantation has been associated with increases of 18.5% in the accuracy of sound localization (Lovett et al.,

2010). Other children are more limited in their localization ability; able to lateralize sounds from the left or right side of their heads confidently and with high accuracy, but unable to determine the direction of the sound source (as occurs with true binaural processing) as the stimulus is presented closer to the front and centre of their heads (Galvin et al., 2008b; Grieco-Calub & Litovsky, 2010). Many other bilaterally implanted children (particularly older children) have shown no ability at all to locate sound sources (Galvin et al., 2007a). Of the children who show some spatial awareness, many do not differ significantly in their ability to children with bimodal stimulation (a cochlear implant plus hearing aid), and none have the abilities of children with normal hearing (Sparreboom et al., 2010). Although overall the best performers are younger, not all young children demonstrate an ability to locate sound sources (Galvin et al., 2010; Galvin et al., 2007a).

6.2.3 Broader outcomes of bilateral implantation

Most of the research on outcomes for children with bilateral cochlear implants has focused on speech perception in noise and sound localization abilities. There is little research to date comparing broader outcomes of children with unilateral versus bilateral cochlear implants, and at the time of writing, there were no reports of speech production or academic outcomes. An initial theoretical analysis of the cost effectiveness of bilateral implantation suggested that it "is possibly a cost-effective use of resources", but that further data on the costs and benefits of bilateral implantation compared with unilateral implantation are required to reach a definitive conclusion (Summerfield et al., 2010). To date, two studies using standardized quality-of-life measures have attempted to determine whether bilateral implants facilitate improved quality of life in children, however neither reported a significant improvement for children with bilateral implants (Beijen, 2007; Lovett et al., 2010).

Information on the impact of bilateral cochlear implantation on language is currently limited. A recent study comparing the preverbal communication of children implanted before age 3 years (27 bilaterally; 42 unilaterally) reported that children with bilateral cochlear implants were significantly more likely to use vocalisation to communicate and to use hearing when interacting with an adult than were children with unilateral implants (Tait et al., 2010). After statistically controlling for the influence of age at implantation and length of deafness, it was found that bilateral implantation contributed to 51% of the variance in outcomes. A multi-center study of 91 children with unilateral (n=60) and bilateral (n=31) implants reported that bilateral implantation was not associated with improved expressive or receptive language development (Niparko et al., 2010). Similarly, Nittrouer & Chappman (2009) examined the vocabulary, receptive and expressive language abilities of 58 children tested at age 3.5 years and also found no differences in outcomes between 15 children with unilateral and 26 with bilateral cochlear implants. Both of these studies provide no support, in terms of language development, for providing young children with bilateral cochlear implants.

However, recent initial results of another prospective, multicentre study comparing outcomes for children with unilateral and bilateral cochlear implants showed a significant advantage for bilaterally implanted children with regard to language development (Sarant et al., in press). The groups of unilaterally (n= 11) and bilaterally (n=17) implanted 5-year-old children in this study did not differ with regard to average non-verbal cognitive ability,

parent involvement in intervention or parent stress levels, and children with bilateral cochlear implants achieved significantly higher expressive and total language scores than did children with unilateral cochlear implants. Initial results of a Belgian study of 25 bilaterally implanted children matched for 10 factors with 25 unilaterally implanted children also reported significantly better receptive and expressive language outcomes for the bilaterally implanted children (Boons et al., in press).

Considering other benefits of bilateral implants, Galvin and colleagues' research and clinical experience with older children and young adults indicates that there are more general benefits, such as ease of listening, awareness of the auditory environment, and increased confidence in social situations, that are of great functional value to children with bilateral implants (Galvin & Hughes, in press). For this group, self-motivation and external support and encouragement were particularly important, as adapting to a second implant at a later age is a more difficult process. Parent questionnaire data from this study for 38 children and young adults showed that 79% of children were using two cochlear implants more than 60% of the time, and 68% reported using bilateral implants more than 90% of the time. Reports of perceived benefit in everyday life also indicated that there was no upper age limit beyond which additional benefit could not be gained from bilateral implants. When considering the risks, time and effort required to obtain bilateral implants versus any additional benefit gained, 79% of the families reported that the second cochlear implant was worthwhile, 16% were unsure, and only 5% felt that obtaining bilateral implants had not been worthwhile (Karyn Galvin, personal communication, August 16th, 2011).

6.3 Timing of first and second cochlear implants; sequential and simultaneous implantation

It is reasonable to assume that children who receive a second cochlear implant early in life will have greater neural plasticity of the central auditory system, and that the first implant will have dominated the auditory neural pathways for a shorter period of time also. Electrophysiological studies of auditory brainstem responses in children with early onset of deafness support this view, showing prolonged wave latencies in the second implanted ear for children implanted sequentially compared to those implanted simultaneously (Gordon, 2008; Gordon et al., 2010). Follow up of children has shown that wave latencies improve over time, particularly for children implanted under 3 years of age (Gordon et al., 2007), and that cortical evoked responses are fundamentally different for children implanted before and after age 3.5 years in terms of wave morphology and latency (Bauer et al., 2006; Sharma et al., 2005). These studies suggest that the shortest delay possible (ie. simultaneous bilateral implantation) will maximise the chance of developing true binaural auditory processing. The clinical evidence reported to date supports the electrophysiological findings. Children who receive bilateral implants sequentially when younger adapt more quickly (Dowell et al., 2011; Galvin & Hughes, in press; Scherf et al., 2009) and generally have better speech perception and sound localization outcomes than those implanted when older (Galvin et al., 2007a).

There appears to be a consensus that children receiving a second implant over the age of 4 years perform much more poorly on speech recognition and sound localization tasks, and do not show evidence of true binaural processing (for example, Galvin et al., 2007a; Johnston et al., 2009; Wolfe et al., 2007). Current evidence suggests that simultaneous bilateral

implantation is a safe surgical procedure, and may also offer advantages to ease of adaptation, although there may be greater challenges associated with programming and managing two devices in younger children (Ramsden et al., 2009).

6.4 Factors affecting outcomes

Outcomes with bilateral implants are influenced by many of the same interacting factors as with unilateral implants (see section 4). As with unilateral implants, factors such as age at time of first implant and amount of pre-operative auditory stimulation in the ear implanted second contribute to outcomes, with younger children and those with pre-implant hearing aid use achieving better results (Galvin et al., 2007a; Peters et al., 2007; Wolfe et al., 2007; Zeitler et al., 2008) . Consistency of device use also influences outcomes, with most children implanted at younger ages adapting more quickly and with greater ease to using bilateral implants, whether they are simultaneously or sequentially implanted (Galvin et al., 2008a; Scherf et al., 2009). Older sequentially implanted children and young adults (who are responsible for their own consistency of device use) must be highly self-motivated in order to persist with learning to use their second cochlear implant; this can be particularly difficult for children aged 7-12 years, especially if they have not been involved in the decision-making process (Galvin et al., 2009). Children implanted at younger ages are also more likely to achieve similar listening abilities with either device, and appear to have greater potential for the development of localization abilities.

For children implanted sequentially, greater improvements in speech perception and localization abilities are demonstrated when there is a shorter time period between the first and second implants (Galvin et al., 2008a; Schafer & Thibodeau, 2006). Factors associated with poor outcomes include poorer than expected outcomes with the first implant, a long time delay between the first and second implants, and limited experience and/or habilitation using the second implant on its own (Dowell et al., 2011). Given the limited information about outcomes for bilaterally implanted children to date, it is not currently possible to accurately predict outcomes for individuals.

6.5 Limitations in current knowledge of outcomes with bilateral implants

The early literature is limited in showing what is possible for bilaterally implanted children. Many studies have included children with very little experience at the time of assessment (as low as 6-12 months for many studies; Sparreboom et al., 2010). We know from the experience of both adults and children with unilateral implants that speech perception and other skills can improve over a period of years, and from bilateral studies that localization skills also require time to develop, therefore it is reasonable to expect that results could improve over time. The evidence is also limited in terms of the number of children who have been followed. A review of paediatric bilateral implant research noted that over half of the published studies reviewed had only 10 or fewer participants (Johnston et al., 2009). Although there are no reports to date on outcomes for children who have received bilateral cochlear implants aged under one year, it would not be unreasonable to expect that very early bilateral implantation would also further optimize outcomes, given the electrophysiological and other evidence collected to date (Peters et al., 2010). There is also currently a lack of evidence regarding quality of life, language, literacy and academic outcomes for children with bilateral compared to unilateral implants. As more children

receive bilateral implants, studies with larger numbers of participants observed over longer periods of time will be conducted, as has occurred with unilateral implants. These studies will no doubt provide further information on which the magnitude of the effect of bilateral implants on outcomes can be measured.

7. Conclusion

Enormous progress has been made over the past three decades in the development of cochlear implants. We have progressed from uncertainty and controversy around whether children could use the incomplete auditory information provided by a unilateral cochlear implant to develop spoken language, to documenting outstanding and life-transforming success for many children with unilateral or bilateral cochlear implants. Cochlear implants are now accepted as the standard of care for children with severe-profound hearing loss. They have allowed many children to attend regular schools, and to develop their language, social and academic skills to levels that exceed those for their peers with severe-profound hearing loss using hearing aids. For some children, cochlear implants have facilitated outcomes such as those their hearing peers achieve, including post-secondary school study, fulfilling employment, and rich social relationships in the hearing world. However, there are still a significant number of children with cochlear implants whose speech intelligibility, speech perception, spoken language, academic and social development are far below that of children with normal hearing. There remains enormous variation in outcomes between individuals with both unilateral and bilateral cochlear implants. Other influences related to neural maturation and development, and also to complex interactions between demographic variables, environmental factors, intervention and learning processes, are not yet understood. A challenge for the future will be to make progress in our understanding of these factors and processes in order to improve outcomes for a greater proportion of children with cochlear implants. Further follow-up of children with unilateral and bilateral cochlear implants is required in the future to determine what the best outcomes will be.

8. References

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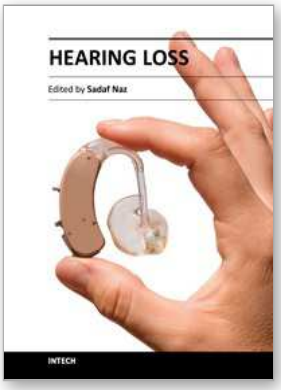

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Authored by 17 international researchers and research teams, the book provides up-to-date insights on topics in five different research areas related to normal hearing and deafness. Techniques for assessment of hearing and the appropriateness of the Mongolian gerbil as a model for age-dependent hearing loss in humans are presented. Parental attitudes to childhood deafness and role of early intervention for better treatment of hearing loss are also discussed. Comprehensive details are provided on the role of different environmental insults including injuries in causing deafness. Additionally, many genes involved in hearing loss are reviewed and the genetics of recessively inherited moderate to severe and progressive deafness is covered for the first time. The book also details established and evolving therapies for treatment of deafness.