Outcomes and Predictive Factors with Cochlear Implants for Adults with a Significant, Early-Onset Hearing Loss

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ABSTRACT

Background and Aims

Adults with an early-onset hearing loss are being referred to cochlear implant clinics more frequently in recent years than was observed during the early years of cochlear implantation. Open set speech perception outcomes for adults with early onset hearing loss and late age at cochlear implant are typically below those measured for adults with an acquired hearing loss, and show wider variability. This variability in outcomes makes it difficult to provide appropriate and realistic counselling regarding potential to benefit in pre-operative sessions. To address this issue, this study investigated cochlear implant outcomes and predictive factors for adults with a significant early-onset hearing loss.

Method

The study design involved a prospective, longitudinal evaluation of outcomes and predictive factors for a study group of 29 adults with a significant, early-onset hearing loss who received cochlear implants at the Royal Victorian Eye and Ear Hospital, Melbourne Australia.

In addition to standard pre-operative clinical assessments, receptive language was assessed using the PPVT, speech intelligibility was rated using the NTID rating scale, and communication modes (reliance on sign or oral communication) were investigated. Temporal processing was assessed via gap detection testing, non-verbal IQ was assessed using the Raven’s Progressive Matrices test (RPM), and cortical auditory evoked potentials (CAEP) were recorded. Post-operatively, speech perception was measured at 3, 12 and 24 months after surgery. At the same assessment points, electrically evoked CAEP were recorded, and correlated with speech perception results.

A study questionnaire was administered post-operatively to determine if a recipient’s satisfaction with their cochlear implant was correlated with performance. Speech
perception, gap detection and questionnaire results were compared to a control group of adults with an acquired hearing loss.

Results

The study cohort of 29 adults with significant early-onset hearing loss gained significant benefit from their cochlear implant, as measured by increased speech perception results post-operatively. Results, however, were significantly lower than those obtained from 576 adults with an acquired hearing loss from the Royal Victorian Eye and Ear Hospital.

Receptive language was not correlated with post-operative speech perception, but was correlated with speech intelligibility; participants with greater speech intelligibility achieved better post-operative speech perception results. Those participants who used purely oral language achieved better speech perception and had better speech intelligibility than those who used some signing for communication.

Gap detection was correlated with performance, with those participants with lower gap detection scores achieving better speech perception results. Both cortical evoked responses and non-verbal IQ were not correlated with outcomes.

Adults with an early-onset hearing loss scored significantly lower than adults with an acquired hearing loss in all domains of the questionnaire, and speech perception performance was correlated with perceived communication abilities, confidence, and device satisfaction.

Conclusion

Adults with a significant early-onset hearing loss have the potential to gain significant improvements in speech perception scores after receiving a cochlear implant. Results from the current study suggested that the most important factor in predicting cochlear implant outcomes for adults with a significant early-onset hearing loss, may be the level of useful hearing an individual had during the critical period for maturation of the auditory system and associated spoken communication (as reflected by the
participants’ speech production intelligibility), and most importantly, how they used this hearing. Due to the difficulties in obtaining an accurate hearing history, rating speech production is a clinically useful tool to provide insight into adults’ prior hearing levels and use of audition. Questionnaire results suggested that recipient satisfaction with their device was dependent on their speech perception performance. This highlighted the importance of the clinician’s role in guiding a patient through the decision-making process to enable adults with early-onset hearing loss to make fully informed decisions based upon realistic expectations of outcomes.
DECLARATION

THIS IS TO CERTIFY THAT:

1. THE THESIS COMPRISSES ONLY MY ORIGINAL WORK TOWARDS THE PHD EXCEPT WHERE INDICATED IN THE PREFACE

2. DUE ACKNOWLEDGEMENT HAS BEEN MADE IN THE TEXT TO ALL OTHER MATERIAL USED

3. THE THESIS IS FEWER THAN 100,000 WORDS IN LENGTH EXCLUSIVE OF TABLES, MAPS, BIBLIOGRAPHIES, AND APPENDICES

ALEXANDRA MAY ROUSSET

6 SEPTEMBER 2017
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### ABBREVIATIONS

<table>
<thead>
<tr>
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<th>Description</th>
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<tbody>
<tr>
<td>ABR</td>
<td>Auditory Brainstem Response</td>
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<tr>
<td>AEO-HL</td>
<td>Adults with Early-Onset Hearing Loss</td>
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<tr>
<td>AUSLAN</td>
<td>Australian Sign Language</td>
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<td>BB</td>
<td>Broad Band</td>
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<tr>
<td>CAEP</td>
<td>Cortical Auditory Evoked Potentials</td>
</tr>
<tr>
<td>CI</td>
<td>Cochlear Implant</td>
</tr>
<tr>
<td>CNC</td>
<td>Consonant-Nucleus-Consonant</td>
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<tr>
<td>dB</td>
<td>Decibels</td>
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<tr>
<td>EEG</td>
<td>Electroencephalography</td>
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<tr>
<td>fMRI</td>
<td>Functional Magnetic Resonance Imaging</td>
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<tr>
<td>HA</td>
<td>Hearing Aid</td>
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<td>Hz</td>
<td>Hertz</td>
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<tr>
<td>IQ</td>
<td>Intelligence Quotient</td>
</tr>
<tr>
<td>LAPL</td>
<td>Loudest Acceptable Presentation Level</td>
</tr>
<tr>
<td>M</td>
<td>Mean</td>
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<tr>
<td>MCIC</td>
<td>Melbourne Cochlear Implant Clinic</td>
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<tr>
<td>MLR</td>
<td>Middle Latency Response</td>
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<tr>
<td>MMN</td>
<td>Mismatch Negativity</td>
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<tr>
<td>MRI</td>
<td>Magnetic Resonance Imaging</td>
</tr>
<tr>
<td>ms</td>
<td>Millisecond</td>
</tr>
<tr>
<td>NF2</td>
<td>Neurofibromatosis Type 2</td>
</tr>
<tr>
<td>NTID</td>
<td>Northern Technical Institute of the Deaf</td>
</tr>
<tr>
<td>PET</td>
<td>Positron Emission Tomography</td>
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<tr>
<td>PPVT</td>
<td>Peabody Picture Vocabulary Test</td>
</tr>
<tr>
<td>Abbreviation</td>
<td>Description</td>
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<td>--------------</td>
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</tr>
<tr>
<td>PTA</td>
<td>Pure Tone Average</td>
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<tr>
<td>QoL</td>
<td>Quality of Life</td>
</tr>
<tr>
<td>rCBF</td>
<td>Resting Cerebral Blood Flow</td>
</tr>
<tr>
<td>RPM</td>
<td>Ravens Progressive Matrices</td>
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<tr>
<td>RVEEH</td>
<td>Royal Victorian Eye and Ear Hospital</td>
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<tr>
<td>SD</td>
<td>Standard Deviation</td>
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<tr>
<td>SL</td>
<td>Sensation Level</td>
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<tr>
<td>SS</td>
<td>Standard Score</td>
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<tr>
<td>TC</td>
<td>Total Communication</td>
</tr>
<tr>
<td>WVA</td>
<td>Wide Vestibular Aqueduct</td>
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<td>Yrs</td>
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1 INTRODUCTION

Since the first adult received a cochlear implant at the Melbourne Cochlear Implant Clinic (MCIC) at the Royal Victorian Eye and Ear Hospital (RVEEH) in 1978 (Clark, 2006), cochlear implantation has become the intervention of choice for the majority of adults with an acquired hearing loss, and young children with a hearing loss which is severe or profound. Outcomes with cochlear implants (CI) for these patient populations are generally very successful, with the majority of adults regaining open-set speech perception, and young children developing oral speech and language (Blamey et al., 1996; Dettman et al., 2016; Dowell et al., 2004; Geers, 2004; Leigh et al., 2013). As the successful outcomes with cochlear implants became more widespread, interest began to arise amongst the Deaf community as to the feasibility of cochlear implantation for adults with an early onset hearing loss. Although there was initially some resistance amongst the Deaf community toward implantation, acceptance of the device as an appropriate intervention strategy is increasing, and cochlear implant centres around the globe are receiving growing numbers of referrals for adults with an early-onset hearing loss (Hladek, 2002; Shpak et al., 2009; Zwolan et al., 1996). Currently, 403 adults with an early-onset hearing loss have received cochlear implants at the RVEEH, Melbourne.

The term ‘prelingual’ is often used to describe patients who present with an early-onset hearing loss, as their hearing loss is generally believed to have occurred prior to the acquisition of oral language. When working with adult cochlear implant recipients, however, determining the exact age of onset of hearing loss is often problematic. The categorisation of ‘prelingual’ may be based on the case history provided by the patient, rather than an exact diagnosis. Due to the ambiguity surrounding the use of the term ‘prelingual’, for the purpose of this study, adults who are believed to have experienced a hearing loss of a significant degree during infancy or early childhood, shall be referred to as Adults with Early Onset Hearing Loss (AEO-HL) in this thesis.

AEO-HL have traditionally been considered ‘poor’ cochlear implant candidates as they generally achieve lower scores on standardised speech perception testing than adults.
with a hearing loss that was acquired after speech and language had developed normally (Caposecco et al., 2012; Eisenberg, 1982; Kaplan et al., 2003). It is difficult to predict outcomes for any recipient with an early-onset hearing loss with any certainty, as they tend to show much wider variability in results compared to adults with an acquired hearing loss. While some AEO-HL may gain significant speech perception benefit with their cochlear implants, some only achieve an aid to lipreading or environmental sound awareness, and a proportion go on to become non-users (Peasgood et al., 2003; Shpak et al., 2009; Teoh et al., 2004). This variability makes it difficult for clinicians to determine what patient factors can be used to help predict outcomes. Self-report measures and questionnaires have been used extensively with this patient group, with results suggesting that regardless of the generally observed low speech perception scores, the majority of AEO-HL CI recipients wear their device full time and report functional benefit (Zwolan 2006, Kaplan 2003, Chee 2004).

Previous research suggests that prolonged periods of auditory deprivation causes degeneration of the central auditory system, and prevents appropriate maturation of language processing areas within the auditory cortex (Kral & Eggermont, 2007; Sharma & Dorman, 2006). As the auditory system can only develop appropriately with auditory experience (Kral & Eggermont, 2007; Kral et al., 2016), it is now accepted as best practice to provide a CI to a congenitally deaf child as early as practicable, in order to minimise the detrimental effects of auditory deprivation (Sharma et al., 2002b). Studies investigating the development of Cortical Auditory Evoked Potentials in children have suggested a sensitive period for central auditory development of 3.5 years, and that after age seven, auditory plasticity is significantly reduced (Sharma et al., 2015; Sharma et al., 2002b; Sharma et al., 2007). The development of language skills requires the presence of a robust auditory input guiding the organisation of the secondary auditory cortex and association areas during this sensitive period (Teoh et al., 2004). For these reasons, implanting children at a young age maximises their chances of achieving age appropriate speech and language skills (Sharma et al., 2002a).
AEO-HL who receive cochlear implants, may have experienced a disruption to the development of their auditory system by the presence of a significant hearing loss during the critical period of cortical plasticity, and therefore do not attain the same level of auditory skills with their cochlear implant (Lammers, Versnel, et al., 2015). Age at onset of hearing loss is a critical factor to identify as it has been correlated with postoperative outcomes in many studies involving children (Dettman et al., 2007; Geers, 2004; Leigh et al., 2013). Obtaining an accurate hearing history from an AEO-HL is often difficult, however, making it challenging to accurately determine the age of onset of deafness, the age of intervention or hearing aid use, and most importantly how the patient utilised any remaining residual hearing during infancy and childhood.

The literature has demonstrated that outcomes with cochlear implants for AEO-HL are variable, and any one individual prospective recipient may attend a clinic with unrealistic expectations. It is often the case that a recipient has friends or acquaintances with cochlear implants who may have achieved significant speech understanding with their implant. The prospective recipient may hope to achieve the same outcomes, but due to the heterogeneity of this patient cohort, and the differences in early hearing history, expectations of speech understanding may not be realistic. There are two issues to discuss. Firstly, if the recipient does not gain the same benefit as their friend/acquaintance, this has the potential to cause significant stress and disappointment. Secondly, the degree of mismatch between pre-implant expectations and post-implant benefit is likely to also cause stress and disappointment and may lead to non-use of the device.

The clinician guiding an AEO-HL through the decision-making process is faced with a difficult counselling situation. Unrealistic expectations around the benefits of cochlear implantation need to be addressed, and the patient needs to understand the limitations of the technology and uncertainty around outcomes. Conveying the complex interaction of variables known to affect potential benefit from the implant (age at onset, degree of hearing loss, and effective utilisation of hearing during childhood) can be difficult, particularly as the adults may have reduced receptive and
expressive language skills that are due to their early-onset hearing loss. Sometimes young adults with early-onset hearing loss are accompanied to the clinic by their parents who may have made the decision many years ago not to implant. These appointments can be quite emotional as the participant and their family deal with their feelings; realising that decisions, or a lack of awareness of the option of implantation during childhood, are impacting decisions to be made in the present.

There exists a need for clinicians to be able to better counsel AEO-HL cochlear implant candidates pre-operatively. Clinicians require more in depth understanding of how to accurately predict outcomes to ensure that the recipient is aware of the likelihood of success, how to define and frame what success is for each individual case, and how to counsel for an outcome that may not meet the recipient’s expectations. There are four key limitations when reviewing the literature regarding cochlear implant outcomes in this population: study numbers are typically small; many papers report on case studies which is a poorer level of evidence (Rychetnik et al., 2002); studies use a retrospective study design which limits the range of factors that can be investigated; and finally, studies have tended to focus on either objective or subjective results, rather than exploring numerous facets of auditory processing. This study aimed to address these research limitations by exploring multiple domains for 29 AEO-HL prospective CI candidates at the MCIC, Australia.

This study aimed to investigate the auditory processing skills of AEO-HL recipients via psychophysical, electrophysiological, linguistic, and non-verbal IQ testing. It was anticipated that results would provide valuable insight into the likelihood that the recipient had utilised any existing residual hearing at a young age. Objective results were compared with a self-report measure. It was hypothesised that these results would provide valuable information about individual patient variables which may be used to more accurately predict objective outcomes for AEO-HL. The information provided by this study will allow clinicians to counsel prospective patients appropriately, enabling the patient to make informed decisions about cochlear implantation based upon realistic expectations of outcomes.
2 SUMMARY AND ANALYSIS OF THE RELEVANT LITERATURE

Due to the many different topics and tests covered in this thesis, each specific area of investigation was allocated its own chapter. The literature review has been organised in a similar fashion. After an introduction into cochlear implantation, adults with a significant early-onset hearing loss, and the implications of auditory deprivation there is a section titled ‘Assessment of the auditory pathway’. This section is further subdivided into the various subsections which are allocated their own chapter within the thesis body; namely language, gap detection, cortical auditory evoked potentials, and non-verbal IQ. This is then followed by a section investigating self-report questionnaires.

2.1 Cochlear Implantation

Cochlear implantation has now become a widely-accepted intervention for adults with an acquired hearing loss and young children with hearing loss of a severe degree or worse. It has been well documented that a cochlear implant can restore the perception of speech, enabling adults to gain significant improvement as measured by open-set speech perception, and in most cases, use the telephone (Blamey et al., 2013; Skinner et al., 2002). Young children provided with CIs have the potential to develop normal speech and language, and many are attending main stream schools (Dettman et al., 2016; Dettman et al., 2007; Leigh et al., 2013).

The first documented use of electrical stimulation of the auditory nerve occurred in 1957 by Djourno and Eyries (Djourno & Eyries, 1957). Subsequently, House and colleagues began investigating a clinical application for the concept. In 1972, they developed a single channel cochlear implant which was successfully able to elicit sound sensations in many deaf adults. Concurrently, a separate group of researchers in Australia, Europe and the United States began researching multi-channel cochlear implants (Clark, 2006). The first adult was implanted with the Nucleus multichannel device developed by Clark and colleagues (Clark et al., 1979) at the Melbourne Cochlear Implant Clinic in 1978. By 1985, the United States Food and Drug
Administration approved the commercial distribution of the multi-channel cochlear implant developed by Clark and his colleagues, in collaboration with the University of Melbourne and Cochlear Pty Ltd (Clark, 2006). Since the late 1970s there have been many technological advances in the internally implanted device, the externally worn speech processor, and the speech coding strategies. Each generation of speech processor has seen a reduction in size and an increase in the number of features available for recipients. Improvement in technology and advances in speech coding strategies have facilitated an improvement in speech perception performance, from around 15% open-set sentence perception with the earliest feature extraction strategies, to an average of over 80% with the newest strategies (Leigh, Moran, et al., 2016; Skinner et al., 2002). Even though the majority of adult cochlear implant recipients are able to understand enough speech with audition alone to use the telephone with some degree of success, a wide variability in outcomes is still observed (Blamey et al., 1996; Leigh, Moran, et al., 2016).

Once improvements in early technology have been accounted for, individual patient factors have been shown to significantly influence cochlear implant outcomes. It has been suggested that these factors can be divided up into primary factors, which are difficult to measure, and secondary factors, which are more easily measured (Blamey et al., 1996). The authors suggested that these primary factors are unique to the individual and may include cognitive abilities and the central nervous system and auditory system of the patient. Secondary factors may include aspects such as age at onset of hearing loss, duration of hearing loss, prior use of residual hearing and duration of implant use (Blamey et al., 1996). Ultimately, it is likely that the primary factors of an individual play a large role in determining cochlear implant speech perception results. It is not possible to accurately measure the primary factors of an individual, however, but it is possible that the secondary factors are related to these primary factors. For example the duration of deafness of an individual, which is easy to measure, may indicate the degree of degeneration within the cochlea, which is difficult to measure (Blamey et al., 1996). Quantifying these secondary factors may therefore provide an avenue for estimating the primary factors of an individual.
The age at onset of hearing loss has been shown to be one of the most important factors in determining cochlear implant outcomes, with those patients who first experienced hearing loss prior to the acquisition of language performing significantly poorer on speech perception testing than those patients who have an acquired hearing loss (Boisvert et al., 2015; Dowell et al., 2004; Kraaijenga et al., 2015). For those patients with an acquired hearing loss, duration of severe to profound hearing loss has been shown to affect outcomes, with longer durations associated with poorer outcomes (Blamey et al., 1996; Dowell et al., 2004). Longer durations of implant use has been shown to be positively correlated with results (Blamey et al., 1996; Blamey et al., 2013) and age at implantation has also been demonstrated to be influential, with those patients implanted at a more elderly age achieving lower scores (Dowell et al., 2004). Those recipients who had demonstrated better pre-operative auditory skills have been shown to achieve greater performance with their cochlear implant (Blamey et al., 2013; Dowell et al., 2004). More recent studies have indicated that effective use of residual hearing is not restricted to the implanted ear, but that the successful use of the residual hearing in the contralateral ear pre-operatively can also positively influence post-operative results. This suggests that it is the total auditory stimulation the individual has been exposed to which is the important factor (Boisvert et al., 2015; Lazard et al., 2012). The above mentioned studies have all indicated that the unexplained variance in performance remains large (Blamey et al., 1996). It is therefore crucial that a prospective cochlear implant recipient is aware of the potential benefits and limitations of the cochlear implant prior to surgery, and has realistic expectations.

Despite the observed variability in cochlear implant outcomes in adult recipients, overall results were very promising, and investigations began into the the option of cochlear implantation for children. In 1985, the first two young persons (aged 15 and 9 years) received multichannel cochlear implants in Melbourne. These two cases were selected as they both experienced profound to total hearing loss following meningitis, that is, they had developed auditory pathways prior to auditory deprivation. They were also able to complete an extensive test protocol, and could provide reliable feedback
regarding their experiences with the cochlear implant. The internal implant at this stage was too large for a young child’s skull, and required a wire bent to hold the transmitter coil in place (Patrick & Clark, 1991). Both of these external speech processor features precluded their use with small children or infants. One year later, clinical trials began in Australia and the USA for children aged 2-17 (Mecklenburg et al., 1991). These initial studies regarding implantation of older children and adolescents yielded mixed results. For some of these children, signed communication had been their only option during the period of auditory deprivation, and use of hearing aids was reported to be sporadic. Speech perception outcomes were poor, and a proportion of these older children/adolescents went on to become non-users. It was evident that adolescents with early onset hearing loss were not the ideal candidates for cochlear implantation due to the duration of auditory deprivation between birth/meningitis and implant surgery. Investigations continued into the feasibility of implanting younger children, in order to take advantage of neural plasticity. In late 1985, the Mini Nucleus 22 became available, allowing for the implantation of younger children. This device contained an internal magnet therefore eliminating the use of the cumbersome wire/head band previously used to hold the external coil in place. Clark and his colleagues in Melbourne implanted the first young child, aged 5.4 years, in March 1986. The current 1990 FDA guidelines approve the application of cochlear implants for all children/persons older than 12 months of age, who meet audiological and medical criteria.

Since this time, cochlear implantation has become widely accepted as a suitable management option for children with severe hearing loss or worse. The majority of children using multichannel cochlear implants develop the ability to understand some speech in the open-set context (Dowell et al., 2002). The observed variability in outcomes, however, prompted widespread research into factors that may contribute to this variability. The literature suggests that the individual factor which accounts for the largest proportion of variance in post-operative outcomes is the age of implantation (Dettman et al., 2007; Geers & Brenner, 2003; Leigh et al., 2013). It has now become best-practice in many cochlear implant clinics, to provide a cochlear
implant to suitable children prior to 12 months of age, in order to maximise the child’s opportunities to develop age-appropriate language skills (V. Colletti et al., 2005; Leigh, Dettman, et al., 2016). It has been shown that cochlear implantation can be a safe procedure for infants and very young children when there are no other medical issues. Recommendations suggest proceeding with cochlear implantation in infants younger than 12 months only when experienced surgical teams include specialised paediatric anaesthetists (Kempf et al., 1999; Young, 2002). In addition to age at implantation, other factors which have been shown to have a positive effect on outcomes include higher relative socio-economic status (Geers & Brenner, 2003), higher maternal education (Ching et al., 2010) and greater family involvement in the habilitation process (Moeller, 2000; Niparko et al., 2010). The presence of any additional disabilities has been shown to have a negative effect on outcomes (Dettman et al., 2004; Pyman et al., 2000). The provision of an early cochlear implant to a child who meets the audiological, radiological, and medical selection criteria, coupled with optimum family support and involvement, and the provision of robust and consistent auditory input, provides the greatest opportunity for acquisition of age appropriate communication skills.

2.2 Cochlear Implant Results for Adults with a Significant Early-Onset Hearing Loss

As access to cochlear implants for young children increased over the 1980s, anxiety and concern were experienced by many members of the Deaf community (or Deaf culture). A culture is typically thought of as a group of people who share the same attributes and ideas. In the case of Deaf culture, the most important attribute is its signed language (Paul & Quigley, 1990). Members of this group are proud of their culture and language (Tyler, 1993), and some deaf individuals within this community prefer to be referred to as deaf with a capital D (Lane, 1990) as it denotes their identity as a linguistic and cultural minority group.

In the 1980s, members of the Deaf community rejected the concept that the medical profession was trying to find a ‘cure’ for deafness. They argued that deaf people
should not be thought of as disabled, but as members of a minority cultural group (Sparrow, 2005). It was believed that cochlear implants threatened the very future of this culture by ensuring that deaf children grew up speaking an oral language, rather than the signed language of the deaf. The community was so exclusive that even marrying outside of the Deaf community was considered a form of ethnocide by some Deaf activists (Hladek, 2002). The American National Association of the Deaf (NAD) position paper on cochlear implants in children in 1991 stated that cochlear implants in children were ‘unsound scientifically, procedurally and ethically’ and that ‘far more serious are the ethical issues raised through decisions to undertake invasive surgery upon defenceless children’ (NAD 1991,1).

Whilst recognising that the language, education, social and cultural aspects of Deaf culture have been impacted significantly by the introduction of the cochlear implant (Hladek, 2002), the majority of deaf children are born to hearing parents. Hearing parents tend to express their desire for their children to grow up in their own culture, speaking their own language. That is, hearing parents tend to stick with the communication forms that they know and are poor adopters of sign language. Deaf children of hearing parents have admitted that they do not intimately know their own biological families due to difficulties with communication (Hladek, 2002).

As the numbers of parents choosing cochlear implants for their children increased in the 1990s, acceptance of the device within the Deaf community as an ‘assistive listening device’ also increased. It was no longer seen as a threat to the existence of the Deaf community. In their 2000 position paper on cochlear implants, the NAD stated that ‘The NAD recognizes the rights of parents to make informed choices for their deaf and hard of hearing children, respects their choice to use cochlear implants and all other listening devices, and strongly supports the development of the whole child and of language and literacy’ (NAD 2000). The position paper instilled the limitations of the technology, in particular commenting that the device ‘represents a tool to be used in some form of communication’. They advocated visual language during infancy to ensure the child had access to some language prior to implantation.
Support was cautiously expressed and the position paper suggested that ‘parents presently have a reasonable basis upon which to decline implantation for their child’. The paper also advocated counselling with a member of the Deaf community prior to surgery, to ensure the family was aware of all options available to them.

An important focus of the NAD 2000 position paper was on the necessity of appropriate counselling prior to cochlear implantation; both for parents and also for AEO-HL. The paper identified limitations in outcomes for adult recipients, and stressed the importance of the patient understanding these issues prior to surgery (NAD 2000). Cochlear implant clinicians learned that they needed to show sensitivity towards the beliefs of the Deaf community when working with these individuals.

Although the NAD and the literature discussed above was American based, it was evident that the increase in acceptance of cochlear implant technology within the Deaf community was international. Increases in referrals for AEO-HL were seen in Cochlear Implant Centres around the globe. In the Melbourne Cochlear Implant Clinic alone, one AEO-HL was implanted in 1990, 10 in 2000, 33 in 2009, and 34 in 2015. There are currently 403 AEO-HL who have been implanted at the MCIC, out of a total of 2111 adults, as of April 2017.

There is a great deal of evidence in the literature to demonstrate that AEO-HL, as a group, attain poorer levels of speech perception with their cochlear implants than adults with a postlingual hearing loss (Caposecco et al., 2012; Klop et al., 2007; Svirsky et al., 2004). There are a number of factors that have been shown to influence speech perception outcomes for AEO-HL such as degree of residual hearing, prior educational setting, use of hearing aids, and communication mode. Despite their poorer average open set speech perception, there is a large degree of variability (Santarelli et al., 2008). Some recipients become eventual non-users, and some obtain only environmental sound awareness (Peasgood et al., 2003), and yet others gain significant open-set speech perception (Dowell et al., 2002; Shpak et al., 2009). This range in outcomes makes it difficult to counsel patients prior to cochlear implantation.
regarding their individual potential to benefit from a CI in terms of speech understanding.

Previous research suggests that prolonged periods of auditory deprivation causes degeneration of the central auditory system, and prevents appropriate maturation of language processing areas within the auditory cortex (Kral & Eggermont, 2007). The development of language skills requires the presence of a robust auditory input guiding the organisation of the secondary auditory cortex and association areas during the sensitive period (Svirsky et al., 2004). Many studies show that for these reasons, implanting children at a young age maximises their chances of achieving greater speech and language skills (Geers, 2004; Leigh et al., 2013). Prior to the introduction of universal new born screening, and in the absence of any known risk factors, identification of hearing loss before 12 months of age was rarely possible. Parents generally did not suspect a hearing loss until their child failed to develop speech at around 1-2 years of age (Yoshinaga-Itano et al., 2000). Many of the case histories provided by AEO-HL are consistent with this. That is, their hearing loss was not detected or aided until after the critical period for cortical plasticity. The lack of auditory input during this period is a likely factor in the more limited auditory skills in this group after receiving a cochlear implant.

Coupled with sensitive periods for cortical development, each AEO-HL’s use of any residual pre-operative hearing may also contribute to post-operative outcomes. Santarelli and colleagues investigated outcomes for a group of recipients with severe to profound hearing loss diagnosed prior to age three, who received their implant after 12 years of age. All participants had used hearing aids with some success and used an oral/aural means of communication. Although post-operative speech perception scores were significantly lower than those typically seen with adults with an acquired hearing loss, scores continued to increase up to three years after surgery. The authors suggested that the use of hearing aids helped facilitate some auditory input, and that together with an auditory-oral communication mode, this positively influenced the cochlear implant outcomes for this group of patients (Santarelli et al., 2008). Yoshida
also found that post-operative speech perception performance was correlated with early fitting of hearing aids and some use of hearing pre-operatively through the use of oral communication (Yoshida et al., 2008).

The relative benefit or functional use of residual hearing is likely to be further influenced by each AEO-HLs preferred communication mode. A study by Kos and colleagues (2009) compared cochlear implant outcomes for a group of AEO-HL who had all worn hearing aids but who had different modes of communication. One group who had exclusively used sign language for communication was compared with a group who had used cued speech. Results showed significantly poorer post-operative open-set phoneme perception for the group who had only used sign language (p=0.03). The authors suggested that despite similar pre-implant speech perception performance, and similar post-implant detection of sound, the exposure to the structure of oral language, through cued speech and lipreading, gave an advantage when using a cochlear implant. The authors suggested that the reason for the poor results in sign language users was not the lack of auditory stimulation but a lack of understanding and familiarity with the acoustic and temporal structure of oral language (Kos et al., 2009). The previous knowledge of the structure of oral language positively influenced the post-operative outcome for this group of adults who received cochlear implants relatively late in life. These results were supported by Kaplan and colleagues who investigated outcomes among a group of AEO-HL and adolescents with different communication modes. All recipients were satisfied with their implant and reported a significant improvement in their quality of Life (QoL), but significantly greater speech perception performance was measured for the patients who used oral communication as opposed to those using Total Communication (signing support) (Kaplan et al., 2003).

Dowell and colleagues (2002) demonstrated that pre-operative hearing (pure-tone average) was not predictive of outcomes in 25 children/adolescents implanted between eight and 18 years. The pre-operative speech perception and receptive language scores, however, were correlated with post-implant speech perception. They
suggested that the ability of the recipients to use their minimal pre-operative hearing to gain some speech discrimination, carried over to effective use of the signal received via the cochlear implant (Dowell et al., 2002).

In addition to the above pre-operative variables (age at implant, residual hearing, and communication mode) reported in the literature to influence cochlear implant outcomes, variables more specific to AEO-HL such as attendance and motivation have been investigated. A case-study of eight recipients by Zhang and colleagues (2014) suggested that attendance at medical, programming and rehabilitation appointments was correlated with the implant recipient’s motivation. They found that those recipients who were motivated were more likely to wear their speech processors in a variety of situations, to adapt to the auditory stimulation and use it as an aid to communication (Zhang et al., 2014). A pilot study investigating the experiences of eight AEO-HL cochlear implant recipients demonstrated that all recipients had predominantly positive experiences. All recipients were highly motivated and reported that they were not pressured by family members to receive the implant. Two of the patients reported non-auditory sensations when their implants were first activated (experienced as a sensation in the arm or chest), which resolved over the first few weeks of use (Jeffs et al., 2015).

In summary, although AEO-HL CI recipients often did not achieve the post-operative speech perception performance achieved by recipients with a late-onset and shorter duration of hearing loss, they did demonstrate appropriate device usage, satisfaction and perceived benefit (Caposecco et al., 2012). AEO-HL recipients generally reported subjective benefit from their implants such as improved environmental sound awareness, music enjoyment, self-confidence and self-esteem (Most et al., 2010; Straatman et al., 2014; Zwolan et al., 1996).

In addition to speech perception tests, and recipient’s completion of subjective questionnaires, another measure of the relative success or benefit from cochlear implantation can be measured via non-user rates. That is, while cochlear implantation is generally regarded a success for most recipients, there are a small number of
recipients who elect not to continue wearing their speech processor. Factors such as age at implantation, duration of hearing loss, and degree of benefit are likely to influence use and non-use. Generally, rates of non-use of cochlear implants amongst adults with an acquired hearing loss are low. A review of non-use in 313 adult patients across 9 different hospitals in the UK reported rates of non-use from 1-10% (Summerfield & Marshall, 2000). A survey of 423 CI recipients from Birmingham, comprising both postlingually deaf adults and children, revealed that 2.8% of recipients were either limited (0.9%) or non-users (1.9%) of their implant. For the paediatric group, peer pressure was a common reason for rejecting the implant, and for the adults, depression, tinnitus, other neurological issues, and non-auditory stimulation appeared to be the most prominent reasons. Finally, recipients who experience little benefit are most at risk for elective non-use of their cochlear implant (Ray et al., 2006).

The rates of elective non-use for AEO-HL are low, with reported instances of 5-12% (Chee et al., 2004; Kaplan et al., 2003; Zwolan et al., 1996), although do appear to be slightly greater than for children and adults with an acquired hearing loss (Summerfield & Marshall, 2000). Instances of non-use following failure to reach expectations highlights the need for appropriate counselling pre-operatively. Setting appropriate expectations, especially for AEO-HL recipients, is more likely to result in more satisfied implantees and reduce the prevalence of non-use (Shpak et al., 2009).

Due to the wide variability in outcomes and a large number of factors affecting outcomes, it can be difficult for cochlear implant clinicians to counsel prospective recipients appropriately. Pre-implant expectation counselling must be predicated on the individual’s case history. Three problems exist however for the clinician who is trying to obtain a complete and accurate hearing history. Firstly, the recipient rarely recalls the details of their age at onset, degree of hearing loss, and course of hearing loss (stable or progressive). Secondly, the recipient seldom recalls how they made use of any residual hearing. Often the name of a pre-school and school is the only detail provided. Thirdly, due to the very nature of the long-standing hearing loss, the recipient may have relatively poor speech and language, and communication regarding
the case history may occur via a sign language interpreter. These three aspects create
difficulty for the clinician who is attempting to provide counselling regarding
expectations that are grounded and evidence based.

Many authors have stressed the importance of appropriate and targeted counselling. It
may be argued that, for the AEO-HL group, this counselling is even more important for
two reasons. Firstly, the literature suggests a higher likelihood of poor outcomes for
AEO-HL. Secondly, any mismatch between pre-implant expectations and post-implant
satisfaction may lead to anxiety, dejection, anger and perhaps non-use (Fitzpatrick et
al., 2004; Kaplan et al., 2003; Schramm et al., 2002). The issues with this research are
that much of the available data relevant to AEO-HL implantees involves small subject
numbers, and case-study type reports. Further investigation of additional factors, with
a larger subject group, may provide clinicians with better evidence and resources for
predicting outcomes and thus lead to better pre-implant counselling for this AEO-HL
group.

2.3 Implications of Auditory Deprivation

The development of the human auditory system is sensitive to both intrinsic and
extrinsic factors, with early anatomical development occurring in utero without the
need for external stimuli, and the final stages of development relying on external input
extending into childhood (Moore & Linthicum Jr, 2007). Auditory experience begins in
utero, when the cochlear approaches maturation at 24-26 weeks of age (Kral et al.,
2016). With extensive myelination occurring throughout the brainstem around 27 – 29
weeks of age (Moore & Linthicum Jr, 2007), it is at this point that consistent
behavioural and physiological responses to sound are seen (Kuhlman et al., 1988).
Development of the basic anatomy of the cochlea and brainstem proceeds even in the
presence of congenital deafness.

The meaningful development of the auditory cortex, however, is prolonged, involving
progressive myelination in a stimulus-driven manner, with full maturation not achieved
until approximately 12 years of age (Moore & Linthicum Jr, 2007; Ponton et al., 1999).
The beginning of cortical processing of auditory stimuli occurs between 4 months and 5 years of age as axons of thalamocortical afferents begin projecting into the cortical layers. Between 5 and 12 years of age, interhemispheric and intrahemispheric connections begin developing. These are the axons which connect various speech and language areas, allowing more complex processing of speech signals (J. K. Moore, 2002; Ponton et al., 1999). An excellent review into the development of auditory-evoked potentials in the cortex by Eggermont and Ponton (2003) shows that before 4 months of age, only layer 1 axons of the cortex are mature, corresponding to the ABR. Between 4.5 months and 5 years of age, layer IV, V and VI axons start to mature, corresponding to the MLR, T-complex and MMN waveforms. Between 5 and 12 years of age Layer II and III axons start to mature, along with a corresponding decrease in synaptic density. This corresponds with the maturation of the P1 waveform, and the emergence of the N1. Beyond 12 years of age all cortical axons are mature, corresponding with a mature N1 response. (Eggermont & Ponton, 2003; Huttenlocher & Dabholkar, 1997; Moore & Guan, 2001; Ponton et al., 2000; Ponton et al., 1999).

Consistent and robust auditory input is critical during the early stages of development to ensure that appropriate maturation of the auditory pathway takes place. The central auditory system either cannot mature properly in the absence of auditory stimulation, or degenerates due to a lack of stimulation. Plasticity is a term used to describe the ability of a sensory system to be guided and shaped depending on the stimulus, or lack of stimulus, that it receives. A critical period is generally used to describe a window early in development, during which a system is maximally plastic and therefore open to structuring or restructuring on the basis of input from the environment (Werker & Hensch, 2015). There are three main stages that define a critical period. The first is the onset, which is triggered by sensory input, or sensory deprivation, which prompts activation of biological markers responsible for cell excitatory-inhibitory balance. Once the critical period is open, there is an open window during which synaptic pruning and homeostasis of existing synapses occurs. The final stage involves closure of the critical period, which involves applying molecular brakes to consolidate the neural circuit from a plastic to a stable state, which has the potential
to limit any further structural changes in the future (Werker & Hensch, 2015). An infant is born with infinite possibilities, and it is the experiences the infant endures which guide development.

Early onset hearing loss has the potential to not only affect the primary auditory cortex, but can have widespread effects on brain development in general, therefore it can be thought of as a connectome disease. ‘The connectome model is a network map of effective synaptic connections and neural projections that comprise a nervous system and shape its global communication and integrative functions’ (Kral et al., 2016). Kral and colleagues suggested that the presence of an early onset hearing loss may affect the various connections within the brain and may result in stronger couplings with the visual system, for example, and poorer couplings with other higher-order cognitive functions. The auditory component of the connectome includes ‘bottom-up’ connections from the auditory cortex to the higher order auditory areas, such as declarative memory, language, executive function, attention, working memory, motor planning, attention, and object identity. The ‘top-down’ channel of processing is the connections between these higher order areas back down to the primary auditory cortex (Edeline, 2012). Kral proposed that variability in development of these neural feedback loops in response to hearing loss may affect higher order neurocognitive and psychosocial outcomes after cochlear implantation (Kral et al., 2016).

Differences in brain structure due to the presence of early onset deafness have been investigated by Shibata and colleagues (2007) using voxel-based morphometry (VBM). Results showed that the deaf recipients had less white matter in the left posterior temporal gyrus, adjacent to the language cortex, but had the same grey matter asymmetries as normal hearing adults. The authors suggest that the white matter deficit in the left temporal lobe, but not the right, may represent decreased volume for auditory processing, specifically related to speech but not to general auditory function (Shibata, 2007). Altered brain structure has also been investigated by Emmorey and colleagues (2003) using volumetric analysis of MRI data for 25 congenitally deaf adults and 25 normal hearing adults. Results from this study also showed a reduction in white
matter in the left and right Heschl’s gyrus portion of the cortex, suggesting that auditory deprivation from birth results in less myelination and/or fewer fibres projecting to and from the auditory cortices. The amount of grey matter volume in Heschl’s gyrus was the same for the deaf and normally hearing adults, suggesting that hearing loss did not cause cell loss within the primary auditory cortex. The deaf adults therefore exhibited a larger grey-white matter ratio than the normally hearing adults (Emmorey et al., 2003).

The neuronal synapses within the human auditory cortex are believed to increase rapidly (synaptogenesis) within the first year and then decrease gradually as they revise their functions as a result of auditory experience, in a developmental process known as ‘synaptic revision’ or pruning (Huttenlocher & De Courten, 1986). Synaptic density has been measured in normal hearing humans posthumously, with results indicating that new synapse formation begins in the prenatal period and continues for the first four years of life. After age four, synapses begin to be eliminated as a result of the auditory experience of the child (Huttenlocher & Dabholkar, 1997), which makes the system far more efficient, and ensures that the child is responsive to the stimuli which matter. These stimuli will be a result of the environment in which the child is immersed. They are dependent on the hearing status of the child, but also on parental interaction, acoustic input and on the availability of a rich language environment.

During development, new synapses are generated by these early auditory experiences without any competition from existing synapses, but the number of synapses in the adult primary cortex remain constant (Kral et al., 2005). In the case of congenital hearing loss, synaptogenesis and synaptic elimination will not occur, which results in a naïve auditory cortex that is not capable of processing incoming auditory signals appropriately (Kral et al., 2006). Regional cerebral blood flow, (rCBF) at rest represents the density of the neurons and decreases as synaptic revision occurs. A study by Hirano and colleagues (2000) found that the rCBF in the auditory association areas was significantly higher in AEO-HL than in adults with either normal hearing or an acquired hearing loss. This suggested that synaptic revision had not occurred. In addition, listening to a speech stimulus through a cochlear implant did not result in an increase
in CBF in the association areas of the AEO-HL. The authors suggested that the functional differentiation of the auditory association areas in the AEO-HL was not mature enough for appropriate processing of the auditory stimulus (Hirano et al., 2000).

The impact of early hearing loss on the cortical activation patterns within the auditory system in response to speech stimuli has also been investigated via the use of Positron Emission Tomography (PET) in cochlear implant users (Naito et al., 1997). Naito and colleagues investigated the patterns induced by white noise, sentences, and no sound for adults with normal hearing, adults with an acquired hearing loss, and AEO-HL. For adults with normal hearing or an acquired hearing loss, the noise stimulated the primary auditory area, whereas the sentences stimulated the primary auditory area and the auditory association areas surrounding them. In AEO-HL, activation was only observed in the primary auditory cortex, even with the speech stimuli. Naito and colleagues showed that even though the primary auditory cortex of AEO-HL may remain responsive to intracochlear electrical stimulation, there was much less activation of the secondary auditory cortex in response to speech than that found in adults with either normal hearing or an acquired hearing loss (Naito et al., 1997).

In the presence of congenital hearing loss, sensory input is diminished. The unstimulated pathways and connections between the auditory cortex and association areas subsequently remain unpruned, and therefore may provide a conduit for other modalities to access auditory areas (Sharma et al., 2007). Cross-modal plasticity is the term used to describe this recruitment of underutilised cortical processing resources of a sensory system by another system (Buckley & Tobey, 2011). Cross-modal reorganisation does not affect all auditory areas equally. Studies on congenitally deaf cats have shown localised cross-modal plasticity of the interconnections between auditory association areas, which may disrupt top-down processing (Kral & Sharma, 2012). These results supported those of Naito and colleagues (1997) who showed that poor performing CI users showed less activation in the auditory association areas than good CI users and normal hearing adults (Naito et al., 1997), and those of Gilley who
showed that auditory stimulation did not activate auditory association areas in a normal fashion in late-implanted children (P. Gilley et al., 2006). Lee and colleagues showed that in AEO-HL, the amount of activation of the higher order auditory cortices by a cochlear implant decreases with increasing age at implantation (Lee et al., 2005). Nishimura and colleagues also investigated the activation of the primary auditory cortex for AEO-HL. They found that a spoken word was able to stimulate the primary auditory cortex, but not the adjacent language areas, and consequently the subject was able to hear the word, but could not understand it (Nishimura et al., 1999).

Many language areas are multimodal and receive input from both the visual and auditory systems. Multimodal areas are capable of cross-modal plasticity. Therefore if the auditory cortex is not utilised, it has the potential to be recolonised by the visual system (Kral et al., 2001). Functional magnetic resonance imaging (fMRI) has been used to demonstrate that early deafness results in the processing of visual stimuli in the auditory cortex (Finney et al., 2001). The impact of the use of sign language on cross modal plasticity has also been investigated by Fine and colleagues using fMRI to analyse activation patterns. Visual stimuli were presented to a group of normally hearing adults who were fluent in sign language, normally hearing adults who did not sign, and deaf adults who were fluent in sign language. Activation of the auditory cortex in response to the visual stimuli was only observed for the deaf adults, therefore the authors suggested that it was the presence of hearing loss, rather than the use of sign language, that resulted in cross-modal reorganization of the auditory association areas.

In summary, current literature regarding auditory pathway development has important repercussions for the current study. Results of the above-mentioned studies have demonstrated that the presence of a congenital or early-onset profound hearing loss can have far reaching effects on the development of the auditory system. Appropriate maturation of the auditory system can only occur in the presence of auditory input, therefore a significant hearing loss during the critical period results in the altered development of the auditory system (Kral & Eggermont, 2007; Kral et al., 2016). A
fundamental consequence of early hearing loss involves a lack of synaptogenesis and synaptic pruning, resulting in a naïve auditory cortex that is incapable of responding to an auditory stimulus in an appropriate manner (Huttenlocher & Dabholkar, 1997; Huttenlocher & De Courten, 1986; Kral et al., 2006). The literature has demonstrated that early onset hearing loss can result in the decoupling of auditory association areas from the primary auditory cortex via a disruption to the auditory connectome (Kral et al., 2016; Kral et al., 2006), thereby preventing the formation of important auditory feedback loops, and providing a possible conduit for other sensory modalities to colonise the underutilised auditory processing areas (Sharma et al., 2007; Sharma et al., 2009). The functional effects of a lack of synaptic pruning, decoupling of the auditory association areas and cross modal recolonization are evident in studies investigating cortical activation patterns in AEO-HL. Studies have shown that visual stimuli can be processed in the auditory cortex (Finney et al., 2001), and that a speech signal may activate the primary auditory cortex, but not the associated language areas resulting in a cochlear implant recipient who was able to hear the word, but could not understand it (Nishimura et al., 1999). Post-operative speech perception correlates with the activation of higher-order auditory cortices, therefore cross-modal remapping may have a negative effect on outcomes for these recipients. These findings support the notion that early implantation for congenitally deaf children maximises the chances of a good outcome, and that late implanted AEO-HL may struggle to interpret speech signals due to the immature development of pathways and the possible re-organisation of their auditory cortex.

2.4 Assessment of the Auditory Pathway

There are several assessments available designed to evaluate the performance of the auditory system. These tests may provide insight into the development of the auditory system and the availability and appropriate utilisation of auditory stimuli.
2.4.1 Language

The basic anatomical structure of the auditory cortex is immature at birth (Kral et al., 2001). Studies have demonstrated the need for appropriate auditory stimulation at an early age, while the auditory system is most plastic (Sharma et al., 2002a), enabling the brain to develop the necessary association areas and connections for speech and language development (Svirsky et al., 2004). Auditory deprivation during this period prevents appropriate connections between the primary auditory cortex and auditory association areas, impairing the function of top-down and bottom-up feedback loops (Kral, 2016). This disruption to the connectome prevents appropriate maturation of language processing areas within the cortex and results in long lasting effects on the eventual speech perception and spoken language ability of the cochlear implant recipient (Kral et al., 2001). Although the different aspects of language may be restricted by their own critical periods of development, they are all interdependent and a disruption to one will affect the maturation of the others. A hearing loss obtained in the first year will disrupt the acquisition of phonology, and therefore impact the future semantic and syntactic capabilities of the child (Ruben, 1999).

The ability to learn spoken language is innate, but its acquisition is complex and must be learned under appropriate acoustic stimulation. During the first 12 months, children with normal hearing develop advancing skills in speech segmentation, word learning, syntax acquisition, and communication (Levine et al., 2016). Ruben has suggested that there are three fundamental aspects of language which mature at different times in a child’s development; phonology (sounds), semantics (words), and syntax (grammar) (Ruben, 1997). Phonological perception matures during the first 8-10 months of life, basic semantic abilities during the first 2-4 years, and syntactic abilities mature over a longer period and are fully developed by the age of 15 years (Kral et al., 2001; Ruben, 1999).

Studies have demonstrated that new-born infants show a preference for their mother’s voices, suggesting that the ability to discriminate sounds begins in utero (Levine et al., 2016). A study by Werker and colleagues investigated the ability of
infants to discriminate phonemes within their native English language, and two foreign
dialects. At 6-8 months of age the infants could discriminate phonemes in all three
languages, but by 10 – 12 months they displayed a relative difference in discrimination
between English and foreign phonemes. This suggested that by the end of the first
year, a child has developed the ability to categorise and identify phonemes of their
native language (Werker et al., 1981). From birth to 12 months, infants also begin to
experiment with vocalisations; establishing pitch and intonation. Between 12 and 24
months, first words begin to appear and speech gradually begins to replace gesture
and non-word vocalisations. By age three, typically developing children have achieved
extensive vocabularies and syntax skills, and their speech production is generally 50%
intelligible to unfamiliar speakers, and 100% intelligible by age four (Bleile, 2004).

Prelingual hearing impairment generally refers to the onset of hearing loss in the first
few years of life, before the most important aspects of speech are developed.
Substantial delays in language are observed for children with severe to profound
deafness, even for those who are well aided and involved in early intervention
(Nicholas & Geers, 2006). Many studies have highlighted the importance of early
identification of hearing loss (Yoshinaga-Itano et al., 1998) and subsequent early
intervention to minimise the disruption to language development and to maximise the
potential benefit provided by such intervention (Colletti et al., 2011; Connor et al.,
2006; Niparko et al., 2010). Leigh and colleagues have suggested that the critical
period for speech perception may extend beyond the first year of life, but that the
critical period for language development lies within the first 12 months (Dettman et
al., 2016; Leigh et al., 2013). AEO-HL have missed this window of opportunity for
language development via auditory channels. Depending on the duration of hearing
loss, the impact of interventions such as cochlear implants are restricted by disruptions
to language development which occurred in infancy.

2.4.1.1 Communication Mode

It is generally acknowledged that two main philosophies exist in the education of deaf
children; Oral/Aural and Total Communication, but the actual approaches vary widely.
An Oral/Aural approach refers to the concept of dependence on hearing and auditory information, with the intention of developing and utilising spoken language. Total Communication refers to a combination of listening, lipreading, gesture and sign language. Geers and colleagues (2003) described a communication mode on a continuum from purely oral to purely sign language, with differing degrees of reliance on vision and audition for any particular communication mode. The relative benefits of communication mode on speech and language outcomes for hearing aid and cochlear implant users have been debated for decades in the literature. One methodological problem encountered in this research is that any correlations or relationships between communication mode and outcomes may not be definitively the result of a causal effect. The use of sign language may be necessary due to intrinsic or extrinsic patient factors, such as limited access to sound during infancy, late cochlear implantation or cognitive or motor deficits, or may reflect parental choice. The poorer speech perception scores observed for children who use sign language may be a result of confounding factors and may not actually be caused by the use of sign language per se (Dowell et al., 2002).

Parents of deaf children are faced with a range of choices regarding the communication mode and educational setting they wish to embrace. Professionals need to support parents through this stage with sensitivity, but none the less need to ensure that parents are aware of the consequences of such decisions. A document has recently been generated in Germany in order to guide parents. The handout presents an excellent review of the literature with a strong bias towards starting sign language as soon as a diagnosis of hearing loss has been made (Humphries et al., 2015). The authors suggest that the starting point of learning to talk is language, and that early access to sign language can ensure language acquisition and avoid cognitive deficits associated with linguistic deprivation. They believe that deaf children need to be raised bilingual, and that sign language is the best way to give deaf children an early and solid foundation in language. Unfortunately, few studies have examined the acquisition and proficiency of hearing parent’s sign language. Learning sign language can be difficult,
but whilst it is important that the entire family become fluent in sign to provide the
deaf child with a rich linguistic environment, this may not always be the case.

Studies have demonstrated different outcomes for children using cochlear implants
whilst being raised in a bilingual (speech and sign) environment. Vieu and colleagues
found that children learning sign language had poorer syntax and language levels than
those children using oral or cued speech. The authors felt that cued speech was a
better way of coding syntax as it reproduces the native language sentence pattern
(Vieu et al., 1998). Kirk and colleagues (2000) found no difference in language
measures between a group of children utilising oral communication and a group using
Total Communication (Kirk et al., 2000) while another study found a greater vocabulary
improvement in children enrolled in total communication programs when compared to
those in oral programs (Connor et al., 2000). A recent study by Yanbay and colleagues
investigated language outcomes for a group of 42 children enrolled in three different
educational approaches Auditory/Verbal, Auditory/Oral and Total Communication. It
was found that family involvement and socio-economic status had a significant effect
on post-operative language measures. Once these factors had been accounted for, no
significant differences in language scores were observed across the three approaches
(Yanbay et al., 2014). These results suggested that age appropriate language can be
achieved regardless of the communication approach the child received.

The use of a visual mode of communication has been shown to be associated with
poorer cochlear implant speech perception results, than for similar implanted patients
who had utilised audition for communication (Dowell et al., 2002; Kaplan et al., 2003;
Kos et al., 2009). Kaplan and colleagues investigated cochlear implant outcomes
amongst a group of AEO-HL and adolescents with different communication modes. All
recipients were satisfied with their implant and reported a significant improvement in
their quality of Life (QoL), but significantly greater speech perception performance (p =
0.05) was measured for the patients who used oral communication as opposed to
those using Total Communication (signing support). The effect of communication
mode in children was investigated by Kirk, who found significantly poorer speech
perception performance in children utilising total communication than those children using oral communication (Kirk et al., 2000) again supporting the importance of the utilisation of audition during the critical period for the development of auditory processing skills.

In summary, the literature demonstrates irrefutably that crucial aspects of language development are compromised by the presence of a significant hearing loss in infancy and early childhood (Geers, 2004; Leigh et al., 2013). The introduction of sign language in early childhood has not been shown to disadvantage children in terms of their language development (Kirk et al., 2000; Yanbay et al., 2014), and in fact some authors encourage sign language to give deaf children an early foundation in language (Humphries et al., 2015). Sign language has, however, been shown to be negatively associated with speech perception (Dowell et al., 2002; Kirk et al., 2000). The greater majority of these studies, however, involve children who have received their cochlear implants after the first crucial stage of language development; phonological development. Cochlear Implant Clinics are increasingly implanting children prior to 12 months of age, and demonstrating significant benefits in terms of language development and speech perception abilities (Colletti et al., 2011; Dettman et al., 2016; Leigh et al., 2013). The most recent studies that have examined outcomes for adolescents who received their first cochlear implant younger than 12 months, suggest that early device fitting facilitates speech and language ability that is comparable to hearing peers. Age at implant and other parental factors (socio-economic and maternal education) account for significantly more variation in scores than communication mode (Yanbay et al., 2014).

### 2.4.1.2 Speech Intelligibility

The production of speech is complex, involving rapid, and fine tuned motor skills (Bleile, 2004). Profound deafness in early childhood impairs the complex processes involved in the development of speech perception and speech production, by
impairing the acoustic-motor feedback loops. That is, the infant with normal hearing and typical development spends many hours vocalising and babbling and thus developing and strengthening auditory and motor pathways for speech.

Clarity of speech production may be measured with naturalistic samples of speech, more formalised articulation tests, and by subjective measures of speech intelligibility. Formally, a speech intelligibility measure provides a ‘quantitative measure or estimate of the probability that listeners will correctly identify items within classes of linguistic units’ (Boothroyd, 1985) There are two methods of measuring speech intelligibility which have been used most often in the literature. The first involves generating a word-identification percentage score, and the second involves use of a rating scale. Both methods are subjective and susceptible to speaker and listener variables. It is generally recognised that intelligibility of the speaker can be influenced by grammatical context, vocabulary, word familiarity, and the probability of a word occurring (Monsen, 1983), and that how the sample is judged can be influenced by the experience of the listener to speech of the hearing impaired.

Word-identification techniques are generally considered a more accurate measure of intelligibility as they generate an objective measurement of how much of the speech sample the listener actually understood (Porter & Bradley, 1985). Some studies, however, have employed the use of both write-down procedures and rating scales, and have found the two methods to be highly associated (Peng et al., 2004).

Rating scales, where the listener records their perception of the speaker’s overall intelligibility, are a clinically useful tool for those who seek a general indication of how well an individual’s speech will be understood by others (Doyle, 1987). When comparing rating scales, Doyle and colleagues found that the National Technical Institute for the Deaf, NTID scale (Subtelny, 1980) can be used effectively by audiologists with high inter- and intra-rater reliability, and that specific training in the tool was not necessary. The reliability of the NTID was also demonstrated by Porter and Bradley (1985). The NTID is a 5-point scale, allowing listeners to rate speech from a score of 1 representing completely unintelligible speech, to 5 which represents speech
which is completely intelligible. These descriptions serve to remind the listener that the speech of the speaker is to be compared with completely intelligible speech. Judgements made in this comparative way tend to be more accurate than when a listener is required to give an absolute judgement (Doyle, 1987). The SIR (Speech Intelligibility Rating) is another five point hierarchical scale which was designed to classify children’s global speech production, and which has been used extensively in relevant research (Allen et al., 2001; O’Donoghue et al., 1999; Wilkinson & Brinton, 2003).

Speech intelligibility can be measured with a speech-sample that has been elicited in a variety of ways. The subject may read a passage from a book, read a list of sentences, repeat sentences modelled by another person, or engage in natural conversation. Subtelny (1977) found that intelligibility ratings generated by spontaneous speech or the reading of a passage from a book, had the potential to differ significantly. For some subjects, greater intelligibility may be achieved by the reading of a passage, as the context, syntax and semantics are correct. For others, if the content of the book is of a higher literacy level than the subject has attained, their intelligibility may be negatively affected and may in fact be greater during spontaneous speech (Subtelny, 1977).

The measured speech intelligibility of a patient is often correlated with speech perception, and many studies have shown a significant and positive correlation between these two measures, for both children and AEO-HL (Habib et al., 2010; Kaplan et al., 2003; Kos et al., 2009; Svirsky et al., 2007). Habib and colleagues (2010) investigated the effects of age at implantation on a group of 37 paediatric cochlear implant recipients, all of whom used oral communication. The children were split into three groups; those implanted prior to 12 months of age, those implanted between 12 and 24 months, and those implanted between 24 and 40 months. The speech sample was elicited by a speech pathologist modelling a sentence for the child to repeat. Responses were recorded and judged by a panel of three inexperienced adults. They firstly wrote down what they heard, and gave a judgement rating from a scale of 1 (not intelligible at all) to 5 (completely intelligible). The written responses were scored by
the number of key words correctly heard, and averaged across the three listeners. An average rating for each child was also obtained: both measures were correlated. A significantly greater speech intelligibility score was found for those children implanted prior to two years of age compared to those implanted after two years of age (Habib et al., 2010). Svirsky’s data also supported the hypothesis that earlier implantation, preferably prior to two years of age, resulted in better speech intelligibility (Svirsky et al., 2007). Their study suggested the existence of a sensitive period for the development of intelligible speech post-implant; rates of development were faster for children who received their cochlear implants prior to two years of age. A gradual loss of the ability to acquire certain communicative skills with increasing age was observed.

The correlation between speech intelligibility and speech perception was also investigated by O’Donoghue and colleagues. The SIR scale was used to quantify speech intelligibility of paediatric cochlear implant recipients, as it was felt that it was more appropriate to use a rating scale in a clinical setting. A significant positive correlation was found, whereby those children with greater intelligibility achieved better speech perception scores, even after controlling for age at implantation, duration of deafness, age at onset of deafness, and etiology of deafness (O’Donoghue et al., 1999). A key finding of this study was that as early as two years after receiving a cochlear implant, speech perception appeared to predict later speech intelligibility. These results highlighted the need to provide a robust auditory signal, and appropriate rehabilitation, whilst the auditory system was at its most plastic, in order to form optimum speech production clarity.

Speech intelligibility can also be successfully quantified in adult cochlear implant recipients. Van Dijkhuizen and colleagues investigated the predictive value of speech intelligibility for a group of 25 AEO-HL deaf adult cochlear implant candidates (van Dijkhuizen et al., 2011). Participants underwent a battery of speech intelligibility tests, and it was found that scores obtained were largely independent of the listener. Nine of the 25 subjects were implanted, as the study criteria stipulated that only those subjects with above average speech intelligibility were to receive a cochlear implant.
Participants all scored above chance-level on speech perception testing, and the most intelligible in the group had all used exclusively oral communication modes in childhood. The subjects with the best speech intelligibility achieved the best post-operative speech perception scores. Post-operative speech perception scores were also significantly correlated with pre-operative speech perception. The authors suggested that speech intelligibility may be a more reliable predictor of post-operative performance than pre-operative speech perception, as a drop in pre-operative hearing may cause a decrease in speech perception. The speech intelligibility result, however, may better reflect the amount of residual hearing the recipient had when younger, and thus provide a window into the individual’s hearing history. A potential flaw in the study design however, meant that the 16 participants with the poorest intelligibility were not implanted, potentially influencing the study results, and leaving a small study cohort of only nine recipients.

The literature has also demonstrated a significant correlation between speech intelligibility and communication mode. Cochlear implant recipients who use oral language tend to have better speech intelligibility than those who use total communication. There is a significant relationship between the occurrence of functional speech in congenitally deaf adults and prolonged hearing aid use coupled with an education environment with oral-aural emphasis (Sims et al., 1980). Osberger compared the speech intelligibility in cochlear implantees who used oral communication and total communication via a write down procedure (Osberger, 1994). They found that the intelligibility of the oral group was significantly greater than that of the total communication group (48% vs 21%). Similarly, for children, Tobey and colleagues examined the impact of communication mode and classroom placement on speech intelligibility for a group of 131 cochlear implant recipients pre-operatively, and at various points post-operatively up until 8-9 years of age. Significantly greater speech intelligibility was obtained by those children who used oral communication, rather than total communication. Classroom placement before and after implantation was significantly related to speech intelligibility scores obtained at 8 to 9 years of age. Lower speech intelligibility scores at 8-9 years were observed for those children placed
in self-contained special educational programs (Tobey et al., 2004). Some caution in interpretation is required; it cannot be concluded that the placement of a child in a mainstream setting, or that the use of oral communication generates greater speech intelligibility. It may be that those children with a natural tendency toward oral communication may be directed towards a mainstream program, or that those children with additional learning difficulties may be more comfortable in a supported unit (Tobey et al., 2004).

2.4.1.3 Summary

Prior to the introduction of universal new born hearing screening, and in the absence of any known risk factors, early identification of hearing loss was rarely possible. Parents generally did not suspect a hearing loss until their child failed to develop speech at around 1-2 years of age (Yoshinaga-Itano et al., 1998). This is frequently the early hearing history retold by adults with early onset hearing loss in cochlear implant clinics in the present day. These adults may have received limited auditory input during sensitive periods of cortical plasticity, and therefore have reduced potential to develop auditory skills with their cochlear implant. The literature has demonstrated the impact of early onset hearing loss on the development of speech and language (Nicholas & Geers, 2006). Lack of appropriate auditory stimulation during the critical period has been shown to result in changes to the auditory cortex and language association areas (Kral et al., 2001; Sharma et al., 2002a). Mode of communication has consistently been shown to be correlated with speech perception, but not necessarily language skills (Dowell et al., 2002; Kirk et al., 2000; Yanbay et al., 2014). Those recipients who use total communication generally obtain lower speech perception scores than those who use oral language, and generally have poorer speech intelligibility (Osberger, 1994; Tobey et al., 2004). Greater speech intelligibility has been shown to be positively correlated with speech perception results (Tobey et al., 2004; van Dijkhuizen et al., 2011).
2.4.2 Gap Detection

The auditory system of a normal hearing listener is able to make use of its psychoacoustic abilities to resolve fine details in a signal, including spectral shape and timing information. These abilities are important for the accurate perception of speech cues. Temporal processing disorders are related to phonologic processing deficits. An inability to detect small differences in timing in ongoing speech can create speech discrimination errors (Keith, 2000; Yalçinkaya et al., 2009). Temporal processing is most often estimated using the psychophysical gap detection task, which can be viewed as a test of temporal integrity at the level of the cortex (Keith, 2000). It is generally acknowledged that auditory temporal processing improves over the first few years of life, and studies have shown that auditory deprivation during infancy has the potential to result in impaired temporal processing skills (Trehub et al., 1995), implying that the gap detection test can be used as one possible measure of cortical maturation.

Typically, gap detection tasks require a listener to attend to three signals, one of which contains a gap in the sound. The listener is required to select which of the three signals contains the gap. This gap is initially large enough to be easily heard, but after being correctly identified by the listener, the gap is progressively shortened to the point where it can no longer be identified, and the three signals are perceived as identical. The threshold at which the gap is sufficiently wide to be heard as two sounds is the ‘gap detection threshold’.

2.4.2.1 Gap Detection in normal hearing listeners

For normal hearing listeners, the performance for gap detection thresholds quoted in the literature is generally in the order of 2 – 6ms. Moore and colleagues suggest that the gap detection threshold for adults with normal hearing using broad band stimuli is 2-3ms, whereas it is 4ms for a sinusoid above 400Hz (Moore, 1993). Trehub et al looked at gap detection in infants, children and adults, using a 500Hz tone pip, and found normal hearing adults had gap detection thresholds on average of 5.2ms
(Trehub et al., 1995). Samelli and Schochat found a mean gap detection threshold for normal hearing young adults, of 4.19ms using a white noise stimulus (Giannela Samelli & Schochat, 2008). This supports the 4ms found by Musiek et al (2005) and Florentine and Buus (1984), although is slightly longer than the 2 – 3ms suggested by Moore, 1996 (Florentine & Buus, 1982; Moore, 1993; Musiek et al., 2005).

Gap detection thresholds are highly dependent on both the stimulus level and the stimuli used. Variations in these two parameters may account for some of the variation seen in the literature. For normal hearing listeners, gap detection thresholds have been shown to improve up to a 30dB sensation level (SL), above which there is no further improvement (Nelson & Thomas, 1997). Many studies have demonstrated that gap detection abilities are better when a high frequency stimulus is used (Fitzgibbons & Gordon-Salant, 1987; Nelson & Thomas, 1997), although Moore and Glasberg suggest that they do not change much between 400 and 2000Hz when a sinusoid signal is the stimulus (Moore & Glasberg, 1988). Moore and colleagues found that gap detection thresholds measured with sinusoid stimuli are smaller than those measured with bandpass noise for subjects with normal hearing, hearing loss, and cochlear implant recipients. This may be due to fluctuations in the noise which may be confused with the gap, and therefore sinusoid stimuli may give a better measure of gap detection (Moore, 1993).

### 2.4.2.2  Gap detection in listeners with a cochlear hearing loss

The presence of sensorineural hearing loss frequently results in a measured gap detection threshold that is larger than normal, although temporal resolution ability in listeners with hearing loss is strongly affected by the stimulus intensity and the stimulus type (Moore, 1993; Moore & Glasberg, 1988). Whilst gap detection thresholds might be larger than normal at an equivalent presentation level, it has been shown that at a comparable sensation level and with a sinusoid stimulus, the gap detection thresholds of adults with an acquired hearing loss are generally in the same range as adults with normal hearing (Fitzgibbons & Gordon-Salant, 1987; Moore et al., 1989; Nelson & Thomas, 1997). Studies have demonstrated that listeners with mild-
moderate hearing losses have gap detection thresholds that are similar to normal hearing listeners (Fitzgibbons & Gordon-Salant, 1987; Nelson & Thomas, 1997), while listeners with a moderate to severe hearing loss have been shown to be able to resolve gaps within normal limits at equivalent sensation levels, independent of the degree of hearing loss (Fitzgibbons & Gordon-Salant, 1987). When a broadband, or narrow band noise signal is used as the stimulus, however, larger gap detection thresholds are observed with subjects with a hearing loss. This may be a result of the configurations of the hearing loss and a corresponding decrease in sensitivity in high frequency regions, rather than impaired processing skills (Fitzgibbons & Gordon-Salant, 1987; Moore & Glasberg, 1988). Another possibility for the increased gap detection thresholds observed with hearing impaired listeners with a noise stimulus may be due to the presence of recruitment. It is possible that recruitment may result in the inherent fluctuations in a noise stimulus sounding louder than normal for the listener. The inherent dips in the noise might therefore be confused with the gap to be detected (Moore, 1993).

### 2.4.2.3 Gap detection in cochlear implant recipients

Cochlear implant recipients have demonstrated varying abilities to perform gap detection tasks via electrical stimulation. Shannon (1989) investigated gap detection abilities in a group of 17 adults with acquired hearing loss. Ten of these adults had received a Nucleus implant, and completed gap detection testing using a biphasic pulse train. Seven adults had received a Symbion cochlear implant, and performed gap detection using a sinusoid stimulus via the percutaneous connection. For both groups of patients, gap detection thresholds of 20-50ms were obtained near threshold, but that decreased to 1-5ms at a perceived ‘loud’ stimulation level. There was no difference in gap detection threshold between the biphasic or sinusoidal stimulus, or between the apical and basal end of the cochlea. The range and magnitude of gap detection thresholds observed were similar to those observed in normal hearing listeners (Shannon, 1989). It is also possible to measure gap detection thresholds by presenting the signal in the free field. Using this method, Wei and colleagues looked at
Gap detection thresholds for normal hearing subjects, and hearing-impaired subjects with an early-onset or postlingual hearing loss using a multichannel cochlear implant. He used a white noise stimulus, and found average gap detection thresholds of 2ms for adults with normal hearing, 11ms for implanted adults with a postlingual hearing loss, and 41ms for implanted adults with an early-onset hearing loss (Wei et al., 2007). Presenting the stimulus in the free field, however, exposes the signal to limitations posed by both the electrical stimulation and the processing strategy of the individual recipients preferred processor settings.

The relationship between gap detection thresholds and speech perception results shows wide variation in the literature, whether measured acoustically, or via the cochlear implant. Kishon-Rabin and colleagues observed that the relationship between speech perception and psychoacoustic performance was such that subjects with very impaired gap detection abilities, classified as thresholds over 40ms, had poor speech perception (Kishon-Rabin et al., 2009). A study by Shannon and colleagues however, investigated gap detection via direct electrical stimulation through cochlear implants, and found a wide range of speech perception performance which was not correlated with gap detection (Shannon, 1989). A significant correlation between speech perception and gap detection was found by Tyler and colleagues using a 500Hz and 4000Hz narrow band noise in a group of hearing impaired listeners (Tyler et al., 1982). Many studies have demonstrated that subjects with larger gap detection thresholds have lower speech perception scores, (Bosman & Smoorenburg, 1997; Muchnik et al., 1994; Tyler et al., 1982; Tyler et al., 1989), and have nominated 40ms as the upper limit of gap detection thresholds sufficient to perceive temporal information in speech (Bosman & Smoorenburg, 1997; Tyler et al., 1989).

2.4.2.4 Gap detection in AEO-HL

The investigation of gap detection abilities in AEO-HL appears to be mainly restricted to cochlear implant users. A study by Busby and colleagues showed that gap detection measured with an electric stimulus for cochlear implant users who were implanted at a young age, were typically within the normal ranges. They found that those subjects
with larger gap detection thresholds had been implanted at a later age, suggesting that gap detection thresholds may be influenced by auditory deprivation prior to cochlear implantation (Busby et al., 1992). A follow up study looked at gap detection abilities of a group of deaf adolescents with an early onset hearing loss and an average age of 9.4 at implantation. While there was wide variation in outcomes (1.8 - 32.1ms), subjects who became deaf at a later age had lower gap detection thresholds (Busby & Clark, 1999). They also found that subjects with smaller gap detection thresholds showed greater improvement in their perception of sentences presented in the auditory-visual condition compared to pre-operative measures. Their findings suggest that auditory deprivation in early childhood increases the likelihood of poorer gap detection thresholds.

A study by Tong et al (Tong et al., 1988) investigated a range of psychophysical tasks, speech perception and language for three cochlear implant patients with early-onset hearing loss, and found that the patient who used manual communication performed significantly poorer on all tests than the two subjects who used oral communication, and that all three were significantly poorer than a group of adults with acquired hearing loss. These results supported the idea of early cochlear implantation to increase the chance of a sufficient maturation of the auditory pathway, and that auditory experience such as that demonstrated by the oral subjects, provided the opportunity for better performance on psychophysical tasks and performance on speech perception tests. The concept of early cochlear implantation in order to minimise the amount of auditory deprivation can also be supported by psychophysical testing, as shown by Trehub and colleagues (Trehub et al., 1995) who investigated Gap Detection in infants, children and adults. Their results suggest that temporal resolution in infants is nearly adult like by 6-12 months of age. Auditory deprivation for prolonged periods after this time may result in impaired temporal processing skills.

Temporal processing abilities have also been shown to be disrupted in children with auditory neuropathy. In their study of gap detection in children, Yalcinkaya and colleagues found that these children with auditory neuropathy were unable to perform
A gap detection task within normal time limits, with results suggesting that the processing disorder may imply issues at a cortical level. The authors suggested that children with gap detection thresholds greater than 20ms may be unable to perceive rapid changes in formant frequencies of ongoing speech (Yalçınkaya et al., 2009). Abnormal temporal resolution findings were also reported by Rance and colleagues for a group of 14 children with auditory neuropathy, but not age matched peers with normal hearing or sensorineural hearing loss. In this study, temporal processing (as measured by temporal resolution rather than gap detection tests) was found to be correlated with speech perception abilities (Rance et al., 2004). It would appear that disruption to auditory processing by the presence of auditory neuropathy may influence gap detection abilities in a similar way to the presence of an early onset hearing loss.

2.4.2.5 Summary

Acoustic gap detection abilities of AEO-HL, prior to receiving a cochlear implant, have not been widely investigated, most likely due to the difficulty in obtaining a sufficient sensation level. Studies have shown that at an equivalent sensation level, adults with an acquired hearing loss or using a cochlear implant, demonstrate gap detection thresholds in the same range as adults with normal hearing. This would suggest that temporal resolution as measured by the ability to detect gaps, is not impaired by cochlear (hair cell) damage (Moore, 1993; Shannon, 1989). Gap detection thresholds have been correlated with speech perception performance in adults with normal hearing and an acquired hearing loss, and children with auditory neuropathy. It is therefore possible that larger acoustic gap detection thresholds obtained from AEO-HL may indicate deficits in temporal processing abilities which may correlate with poorer speech perception outcomes post operatively.

2.4.3 Cortical Auditory Evoked Potentials

Cortical Auditory Evoked Potentials, or CAEP, refers to the recording of electrical potentials generated by the auditory cortex in response to sound stimuli, from the
scalp of a listener. The presence and characteristics of the response allow conclusions to be drawn about the hearing level of the subject, or the performance of their auditory pathway. CAEPs are endogenous responses, as opposed to the Auditory Brainstem Response (ABR) which is an exogenous response. Exogenous responses are a product of the stimulus, whereas endogenous responses are generated by a higher region of the central nervous system, and are highly dependent on the stimulus context, such as the state of awareness of the subject and any change in the stimulus (Hall III et al., 2007).

In a normal hearing adult, the typical cortical evoked response may have P1, N1, P2 and N2 waves present. P1 is the first positive wave, occurring at 40-60ms following the onset of the tone. N1 is the first negative wave occurring between 80-120ms. The N1 is a robust wave, and along with P1 is generally the most prominent feature of the response. P2 occurs at 150 – 200ms and N2 occurs at 200-300ms (Beagley, 1971). Both the amplitude of the waveforms seen, and the latency of the wave peaks are influenced by the intensity of the stimulus. As the stimulus decreases in level, the latency increases and the amplitude decreases. By manipulating the level of the stimulus, it is possible to estimate the audiometric threshold of the subject, and to assess the auditory pathway. Eddins and Peterson (1999) found that the N1 can be detected for normal hearing listeners at 8dB on average above the audiometric threshold (Eddins & Peterson, 1999). CAEPs are instrumental in the diagnosis of non-organic hearing loss, as they do not require the co-operation of the subject, apart from a passively alert state of concentration.

The generation of a CAEP involves dynamic interactions between various structures found in the auditory cortex within the temporal lobe, the frontal lobe, the limbic system and subcortical regions. The P1 response is generated by auditory thalamic and cortical sources (Sharma et al., 2002a). A study by Moore and Guang (2001) on the structure of the human auditory cortex showed that a mature number of neurons are present at birth in the cortex, but that the axons that carry information to these cells mature at differing rates during the first ten years of life (Moore & Guan, 2001). The P2
component matures at the same time as the brain stem response, at approximately 4.5 months of age, and originates from the primary auditory cortex. The maturity of the P1 waveform represents the start of the maturation process of the axons in layer II and III of the auditory cortex between the ages of 5 and 12. It is only at this point that the N1 wave begins to emerge, with maturation not occurring until after age 12 when the cortical axons are all fully mature (Moore & Guan, 2001; Ponton et al., 2000; Ponton et al., 1999). Contrary to the P1 and P2 wave of the CAEP response, the N1 wave is therefore not fully mature until well into adolescence (Eggermont & Ponton, 2003).

The P1 latency systematically decreases with increasing age, and can therefore be used as a biomarker for investigating the maturation of central auditory pathways in infants and children (Sharma et al., 2002a). The latency of the P1 waveform is a reflection of the delay in propagation through the peripheral and central auditory pathways (Eggermont et al., 1997). The P1 response is a robust wave, occurring at 100-300ms in young children, and decreasing in adulthood. Wunderlich and Cone-Wesson studied the maturation of CAEPs in infants and children, and found that the response from young infants was dominated by a broad positive peak at 200 – 250ms, and then a broad negative trough at around 400 – 500ms (Wunderlich et al., 2006). Latencies are best used to judge maturity, as they are much less variable than amplitude (Eggermont, 1988). The general waveform morphology reaches maturity around 12 years of age, but latencies and amplitude continue to develop beyond this age (Wunderlich et al., 2006).

CAEPs can be also be evoked via an electrical stimulus, generally via a cochlear implant. The stimulus can be presented via a loudspeaker, in which case it would be processed by the speech processor, or a stimulus can be directly presented to a nominated electrode of the cochlear implant. While latencies are shorter than for auditory evoked potentials due to the electrical stimulus directly activating neural pathways and therefore being unaffected by time delays associated with acoustic travel time, the effect is much smaller for CAEP than it is for the ABR (Hall III et al.,
Consistent with an acoustic CAEP, latency decreases and amplitude increases as stimulus level increases.

The use of CAEP responses to monitor development of the auditory system in children after cochlear implantation is now widely accepted. Sharma and colleagues have shown that congenitally deaf children who receive their cochlear implants prior to age 3.5 develop normal P1 latencies within six months of listening with their implant (Sharma et al., 2002b). This study investigated P1 latencies for 104 congenitally deaf children who received cochlear implants between the ages of 1.3 to 17.5 years. The participants in this study were either congenitally deaf, or acquired a severe to profound hearing loss prior to age one. Recordings were made at least six months after the device was switched-on using a synthetic speech syllable presented in the free field. Participants were divided into the ‘early’ implanted group, who received their cochlear implants prior to age 3.5, the ‘middle’ implanted group, who received their implants between 3.5 and seven years of age, and the ‘late’ implanted group, who received their implants after age seven. In the ‘early’ implanted group, 55/57 children showed P1 latencies within the normal range after six months of cochlear implant use, whereas children who received their implant after age seven often did not develop normal latencies, and those implanted between age 3.5 and seven showed a wide variation in latencies. The authors have suggested that these results show that the auditory system of congenitally deaf children remains maximally plastic up to the age of 3.5. Beyond this age plasticity appears to decrease, and beyond the age of seven the substantial alteration in the P1 latency in response to sound is likely due to a significant reduction of neural plasticity (Sharma et al., 2002a).

A longitudinal study looking at two children who received cochlear implants at age six showed that while the P1 matured at a normal rate with a latency delay approximately equal to the duration of deafness, the N1 waveform never emerged (Ponton et al., 1999). The authors have proposed that these results support the hypothesis that while the deeper cortical layers (responsible for P1) may mature in the absence of sound stimulation, the maturation of the superficial layers may require sound stimulation.
during a critical period. If a child receives a cochlear implant prior to age 3.5 they generally develop an N1 that is similar in morphology and latency to normally hearing children, but for those implanted after age seven an N1 generally does not develop (Ponton & Eggermont, 2002). These results support the much cited findings that implanting a child at a young age when the auditory system is at its most plastic will facilitate the best speech and language outcomes by enabling appropriate maturation of the auditory pathways, as indicated by a P1 at an appropriate latency (Sharma et al., 2002b) and the presence of the N1 (Eggermont & Ponton, 2003).

Burdo and colleagues (2006) looked at CERA results for five groups of cochlear implant recipients, including children and AEO-HL, pre- and post-operatively. They used an acoustic tone burst at 500Hz and 2000Hz presented via an audiometer and coupled directly to the participant’s hearing aid for the pre-operative assessment and to the speech processor microphone for the three and 12-month post-operative assessment. Results showed that the N1 and P2 latency decreased from three to 12 months post-operatively for all participants, but these changes were significantly different between cochlear implant recipients with acquired deafness, those implanted as children, and adult recipients with early onset deafness (p<0.00001). The largest decrease was seen for AEO-HL, confirming the immaturity of auditory cortical function, while the smallest decrease was seen for the children who had been good hearing aid users (Burdo et al., 2006).

Cortical responses have been used to investigate cases where an adult cochlear implant recipient has had a poor outcome. Results show that some recipients who had a less than optimal result with their cochlear implant, had an abnormal cortical result, categorised as a response that could not be repeated, or did not occur within an appropriate time window (Psarros et al., 2009). A case study of one AEO-HL cochlear implant recipient demonstrated evoked CAEPs with an immature morphology, similar to that recorded in young infants, consisting of a broad, large amplitude positive peak (Mcneill et al., 2009). Atypical cortical responses were also found in children with cochlear implants who achieved only limited speech perception with their device.
Results showed that children implanted after age four showed a range of waveform morphologies, whilst those implanted at a younger age showed more typical morphologies. Atypical cortical responses were associated with those children with poorer speech perception results, and the authors suggest this may reflect a persistent immaturity in the cortex, or aberrant cortical organisation (Gordon et al., 2005).

Results from the above-mentioned studies suggest that variations in the ‘typical’ P1-N1-P2 evoked potential response can be observed in both adult and paediatric cochlear implant recipients. These variations include delays in the expected latencies of the P1 and N1 waveform, absence of the N1 waveform, and atypical morphologies. These variations have been shown to be moderately correlated with age at implant for paediatric cases, and with speech perception performance for both adult and paediatric recipients (Gordon et al., 2005; Mcneill et al., 2009; Psarros et al., 2009). The variability observed in both latencies and morphology is likely to be a result of cortical reorganization stimulated by auditory deprivation (Dorman et al., 2007).

In contrast to the above studies demonstrating aberrant CAEP responses obtained by AEO-HL cochlear implant recipients, there exists a selection of studies which have shown more ‘typical’ responses from this patient subgroup. A case study of two AEO-HL who underwent acoustic CERA testing via high-powered hearing aids, showed that at sensation levels of 30dB and 10dB, typical P1-N1-P2 waveforms were successfully recorded (Abraham et al., 2015). Both of these recipients were believed to have a congenital profound hearing loss and communicated via gesture only, with no spoken language. Lammers and colleagues (2014) investigated CAEPs in adult cochlear implant recipients, 12 of whom became deaf prior to age two (as far as could be determined) but who received their implant as an adult. Results showed that a typical N1-P2 waveform could be recorded irrespective of speech perception score and onset of deafness. The AEO-HL group however, did show shorter N1 latencies with larger amplitudes than the group of adults with an acquired hearing loss (Lammers, Versnel, et al., 2015). Similar results were obtained by Gordon and colleagues (2008) when
investigating CAEP results from a group of 16 children. The children in the study who scored less than 50% on speech perception testing showed a significantly shorter N1 latency than those who scored over 50% (80ms vs 100ms) (Gordon et al., 2008). It is generally acknowledged that the deprivation of sound prevents the appropriate maturation and development of the auditory system, which can be observed through atypical CERA results (Dorman et al., 2007; Eggermont & Ponton, 2003; Sharma et al., 2002b; Sharma et al., 2007). It is therefore interesting that ‘typical’ P1-N1-P2 waveforms have been successfully recorded in AEO-HL both pre-operatively via an acoustic stimulus (Abraham et al., 2015), post-operatively through the cochlear implant (Lammers, Versnel, et al., 2015), and in implanted children with poor functional outcomes (Gordon et al., 2008). The post-operative studies of Lammers and Gordon, however, have demonstrated that the latency of the N1 is earlier for AEO-HL, and with a bigger amplitude, when compared to adults with an acquired hearing loss (Lammers, Versnel, et al., 2015), and earlier for poor performing children compared to children with good speech perception performance (Gordon et al., 2008). Gordon has suggested that the poor functional outcomes associated with ‘normal-like’ responses suggest that these peaks reflect different activity than that occurring in normal hearing children (Gordon et al., 2008). The possibility of a loss of synaptic pruning as a result of early onset hearing loss in the adult patients in Lammer’s study (Huttenlocher & Dabholkar, 1997), combined with the possibility of ‘corticocortical de-coupling’ (Kral & Eggermont, 2007), may have resulted in a larger cortical area responding to a stimulus, leading to the larger and earlier N1 peak observed (Lammers, Versnel, et al., 2015).

One of the main limitations present in the literature regarding the use of CAEPs in evaluating cochlear implant performance, is the small subject numbers in each study. With the exception of the large studies by Sharma and colleagues (2002) which analysed results in 104 children, and which was then replicated by Dorman (2011) with 245 children (Dorman et al., 2007; Sharma et al., 2002b), the majority of papers have limited numbers or report on case studies of one or two cochlear implant patients. Whilst the large body of work by Sharma, Dorman and indeed also by Wunderlich and Cone-Wesson (2006), allows confident conclusions to be drawn about the maturation
of the P1 latency with development in children, the incidental reports in much of the other literature makes it difficult to draw definitive conclusions (Sharma et al., 2002b; Wunderlich et al., 2006).

In summary, as CAEP results have successfully been used both to assess the maturity of the auditory cortex, and to investigate cochlear implant recipients with less than optimal results, their use pre-operatively has the potential to contribute some valuable information as to the development of a recipient’s auditory system. This may be of particular interest when working with AEO-HL, who are often unaware of their very early hearing history. The presence of a mature cortical waveform may indicate that the auditory system had been sufficiently stimulated at an early age to enable appropriate development to occur (Pantev et al., 2002). The ability to successfully record acoustic CAEP responses from pre-operative cochlear implant recipients is hindered by the severity of the hearing loss. This issue may be overcome by following up with post-operative CAEPs using direct stimulation to the cochlear implant. Without the limitations of a minimal sensation level, it may be possible to monitor the development of the response over time, and to investigate whether those recipients who display responses within a defined morphology guideline or latency period, demonstrate greater speech perception with their cochlear implant.

2.4.4 Non-Verbal IQ

The investigation of intelligence in AEO-HL has not been widely reported. While intelligence is widely discussed among psychologists, there is no standard definition of what exactly constitutes ‘intelligence’. Some researchers have suggested that intelligence can be defined as a general ability while others believe that it encompasses a range of skills, although most agree that genetics and environment both play a role in determining intelligence. In 1904, Spearman described a concept he referred to as general intelligence, or the ‘g’ factor (Spearman, 1904). He argued that because people who scored well on one test tended to also perform well on another, intelligence could be thought of as a general cognitive ability that could be measured
and numerically represented. Often intelligence is referred to as IQ, or Intelligence Quotient, but there are other test materials available which can give a variety of measures. Intelligence testing is regularly performed to obtain funding for education or community support, and is widely used in schools.

Nonverbal intelligence is the ability to analyse information and solve problems using visual or hands-on reasoning, as opposed to verbal intelligence, which is the ability to comprehend and solve language-based problems. Nonverbal intelligence is critical as it allows a person to analyse and solve complex problems without being limited by language abilities.

Nonverbal tasks involve skills such as:

- the ability to recognise visual sequences and remember them
- understanding the meaning of visual information and recognising relationships between visual concepts
- performing visual analogies
- recognition of causal relationships in pictured situations.

Raven’s progressive matrices (RPM) were developed as a non-verbal intelligence test in order to remove language barriers in the estimation of intellectual aptitude, making them a good choice in assessing people with reading difficulties or hearing impairment (J. C. Raven, 1936). The test measures the two main components of general intelligence (Spearman, 1904): the ability to think clearly and make sense of complexity, which is known as educative ability, and the ability to store and reproduce information, known as reproductive ability. A study by Wu and colleagues (2008) investigated the intellectual ability of Mandarin speaking children using CIs, and utilised the Wechsler Intelligence scale for children. The study found that results on performance IQ, or non-verbal IQ, were not significantly different to the published norms for children, but that results on the verbal IQ were significantly poorer. The authors suggested that the use of verbal IQ tests for deaf children may not be a true
representation of intelligence, and that it may in fact be another form of measuring spoken language development (Wu et al., 2008).

Non-verbal IQ has been studied extensively in children with hearing impairment. Non-verbal IQ has been found to be linked to academic achievement in the hearing impaired student (Watson et al., 1986) although it accounted for only a third of the variability in academic achievement scores. The reading level attained by deaf children with cochlear implants has also been shown to be related to nonverbal intelligence (Archbold et al., 2008). It was found that children with low scores on the Ravens Coloured Matrices test (developed for use with children) were unlikely to have age appropriate reading scores. A ten-year study by Coletti and colleagues (2010) found that children who received their implants as infants, performed better on cognitive non-verbal tests than those who received their implants at a later age. The authors suggested that early implantation had a positive effect on complex non-verbal cognitive functions and played a fundamental role in the development of higher cognitive functions utilising multisensory integration (Colletti et al., 2011). Results from these studies suggested that hearing impaired children with lower non-verbal IQ scores tended to achieve lower reading skills and academic achievement levels than children with higher non-verbal IQ scores, and that children implanted earlier tended to attain better scores on cognitive non-verbal tests than those children implanted later (Archbold et al., 2008; Colletti et al., 2011; Watson et al., 1986). The prior literature considers children and adults who are within the normal range of development. There are also studies which have examined cochlear implant outcomes for children with recognised learning difficulties and developmental delays. Performance on speech perception tasks has been shown to be negatively associated with the presence, and severity of additional disabilities (Dettman et al., 2004; Pyman et al., 2000). In a study of non-verbal IQ in 13 children with cochlear implants, Park and colleagues (2015) found a positive correlation with post-operative auditory performance. The Wechsler Intelligence scale for children was used in the study, and a strong positive correlation was observed between the performance (or non-verbal) IQ and post-operative CAP (Categories of Auditory Performance) scores. In particular, the
subtests of picture completion and picture arrangement had the highest correlation. These subtests reflect social cognition, which was found to be generally poorer in CI users than the normal hearing population (Park, Song, et al., 2015).

The literature suggests that cognition, and in particular performance on non-verbal tests of IQ, is linked to post-operative cochlear implant results, reading ability and academic achievement in children (Archbold et al., 2008; Watson et al., 1986; Wu et al., 2008). Early implantation has also been shown to influence the development of cognition, and the use of phonological memory in speech perception and production (Colletti et al., 2011; Pisoni, 2000). It is feasible to assume that the same conclusions can be drawn for adults with a hearing loss who may also have language delays. Non-verbal IQ may be a more accurate measurement of intelligence for these patients.

Little research seems to exist, however, regarding the use of non-verbal tests of intelligence with adults with an early onset hearing loss.

### 2.4.5 Self-Report Outcome Questionnaire

Despite relatively poor objective results on traditional speech perception measures, the majority of AEO-HL who receive cochlear implants wear their devices full time and report high levels of overall satisfaction (Zwolan et al., 1996). It is apparent that traditional outcome measures may not always be sensitive enough to capture the advantages provided to this recipient group by cochlear implant technology. Studies have suggested that qualitative measures, such as Quality of Life questionnaires, may be more sensitive at capturing functional benefit (Caposecco et al., 2012; Chee et al., 2004; Most et al., 2010).

According to the World Health Organisation (WHO) QoL working group, ‘Quality of Life’ (QoL) is defined as an ‘individual’s perception of their position in life in the context of the culture and value systems in which they live and in relation to their goals, expectations, standards and concerns’. (Osberger et al., 1993). The QoL is a multi-dimensional construct, with contributions from several different aspects, or ‘domains’
of life. The primary aim of medical intervention is to improve a patient’s quality of life. In the case of cochlear implantation, this means not only improving a person’s ability to hear, but impacting their self-esteem, social interactions and daily functioning. The generic quality of life instruments available have proven to be insensitive to some specific health-related aspects of hearing loss, therefore it is important to use a disease-specific quality of life instrument when investigating the effects of hearing loss (Hinderink et al., 2000).

In order to more effectively study the improvements in QoL gained by cochlear implantation, Hinderink and colleagues at the Nijmegen Cochlear Implant Clinic developed a quantifiable, self-assessment HRQoL tool for use specifically with cochlear implant users (Hinderink et al., 2000). The Nijmegen Cochlear Implant Questionnaire (NCIQ) measures important improvement in 3 main domains; Physical, Psychological, and Social. Many studies have successfully used the NCIQ to measure QoL improvements facilitated by cochlear implantation (Damen et al., 2007; Hinderink et al., 2000; Hirschfelder et al., 2008). Other studies have utilised the Glasgow Benefit Inventory, which is designed for measuring outcomes after otolaryngological procedures (Lassaletta et al., 2006), while others have designed their own study questionnaires (Caposecco et al., 2012; Chee et al., 2004). The important feature of these tools is that they are designed to measure functional benefit provided by the cochlear implant, as perceived by the recipients themselves.

Improvements in QoL have been well documented for adult cochlear implant recipients with a postlingual hearing loss. Krabbe investigated the effect of cochlear implant use on a group of postlingually deaf adults. Three questionnaires; the NCIQ, a generic HRQoL, and a health-state classification system HUI-2, were sent to 45 adult cochlear implant recipients with a postlingual hearing loss, and 46 adult patients waiting for a cochlear implant (Hinderink et al., 2000). Results showed that the cochlear implant led to a significant improvement in QoL. Lassaletta and colleagues demonstrated that in addition to improvement in hearing and speech perception, daily
activities, social functioning, and self-esteem were also affected by cochlear implantation (Lasaletta et al., 2006).

More recently, research has focused on cochlear implant outcomes for AEO-HL. Many studies have focussed on speech perception outcomes, which are generally significantly poorer than those of adults with a postlingual hearing loss (Klop et al., 2007; Most et al., 2010; Zwolan et al., 1996). More studies are now investigating the impact that cochlear implantation has on QoL for these patients. It is observed that even when there is no measurable improvement in speech perception, recipients reported high levels of satisfaction with their cochlear implant (Zwolan et al., 1996), and improvement in communication, social skills, academic and work performance, and self-esteem after implantation (Most et al., 2010).

It has been suggested that subjective benefit obtained from the CI is not solely related to a recipients’ speech perception performance (Hinderink et al., 2000). This finding is supported by those of Zwolan et al (1996); despite the 12 adults with early onset hearing loss demonstrating no open-set speech recognition and very little closed set speech recognition post-operatively, all recipients reported that they were satisfied with their cochlear implant, and that it improved both their expressive and receptive communication skills (Zwolan et al., 1996). Straatman and colleagues (2014) also demonstrated a lack of correlation between speech perception performance and quality of life measures in 28 adults with early onset hearing loss. They suggested that quality of life is not only influenced by speech recognition, but by the ability to hear environmental sounds and the availability of additional auditory cues (Straatman et al., 2014).

In addition to evaluating speech perception performance for a group of 30 and 38 adult cochlear implant recipients with early-onset hearing loss respectively, Chee (2004) and Caposecco (2012) developed their own study questionnaires to capture perceived benefit provided by the implant. As well as multiple choice questions, both studies asked recipients to list advantages and disadvantages of the cochlear implant. Results from both studies showed improved awareness of surroundings, improved
communication, and improved ability to understand speech. The listed disadvantages were minor practical issues surrounding use of a speech processor, such as the high battery consumption (Caposecco et al., 2012) and the physical discomfort associated with wearing the processor (Chee et al., 2004). It must be noted that some of this data was derived from speech processors that were body worn, before behind-the-ear speech processors became available in 1998.

It was apparent from the literature that irrespective of speech perception performance, most AEO-HL cochlear implant recipients chose to wear their devices for the greater part of the day. Zwolan found that all recipients reported wearing their devices more than 10 hours per day, and that this was unrelated to speech recognition scores (Zwolan et al., 1996). Of the 38 recipients, 81% wore their implants more than 8 hours per day and 13% wore it 4-8 hours per day (Caposecco et al., 2012), and out of 30 participants, 67% wore their implant ‘always’ and 30% ‘almost always’ (Chee et al., 2004). This high rate of device use, which was unrelated to speech perception performance, suggested that the recipients themselves were noticing and appreciating subtle benefits in their day to day lives that were not measured by traditional speech perception testing.

When evaluating post-operative performance of AEO-HL cochlear implant recipients, the measurement of improvement via a health related QoL instrument may be more sensitive than traditional speech perception testing. The impact of greater environmental sound awareness, for example, must not be overlooked. It has been suggested that for this cochlear implant population, benefit and performance should be viewed as two separate outcomes (Most et al., 2010), and that due to the lack of correlation between speech perception performance and quality of life, both speech recognition and quality of life measures should be used to evaluate performance (Straatman et al., 2014).
As cochlear implantation has become more widely accepted within the Deaf community as an appropriate intervention strategy, cochlear implant clinics worldwide are receiving increasing numbers of referrals for AEO-HL.

The literature suggests that speech perception outcomes for AEO-HL who receive cochlear implants are poorer than typically measured for adults with an acquired hearing loss. Results show great variability, however, from significant open-set speech perception through to sound awareness only, and a proportion of these recipients may reject their device. The large variation in observed outcomes makes it difficult for the clinicians working with this patient cohort to appropriately counsel and guide a prospective candidate through the decision-making process. Clinicians are often faced with unrealistic expectations from the patient, which must be addressed to ensure that the patient fully understands the benefits and limitations of cochlear implantation.

Typically, past studies investigating cochlear implants for AEO-HL have focussed on one specific aspect of auditory processing, or subjective benefit, and few have recruited sufficient participant numbers to draw definitive conclusions. The current state of knowledge regarding cochlear implantation for this group suggests that speech perception results are variable, temporal processing skills may be poor, language skills are often delayed, and abnormal electrically evoked cortical responses may be associated with poor performance. Despite these objective results, many studies report that AEO-HL achieve significant subjective benefit from cochlear implantation and wear their device all day.

It remains uncertain as to whether poor pre-operative gap detection abilities and the inability to measure a cortical response, despite sufficient sensation levels, reflect deficiencies of the auditory system, leading to a poor post-operative outcome. Alternatively, the significant degree of hearing loss often observed in this population may infact be the cause for the poor gap detection thresholds and lack of a
measurable cortical response. Language and speech production tests may provide further indication of the availability of auditory input at a young age and are of particular interest as many recipients may have insufficient hearing thresholds to obtain reliable psychoacoustic or electrophysiological data pre-operatively. It is anticipated that this project may provide some insight into these questions for a group of AEO-HL by investigating the relationships between post-operative speech perception performance and the pre-operative non-verbal IQ, language, psychophysical and electrophysiological measures for each recipient. Post-operative objective performance will also be compared with post-operative evoked CAEP and subjective results.
4 STATEMENT OF THE RESEARCH QUESTIONS

The first aim of this study was to investigate the speech perception outcomes with cochlear implants for a group of 29 AEO-HL from the Melbourne Cochlear Implant Clinic (MCIC), RVEEH, Australia. Speech perception was measured pre-operatively, and at 3, 12 and 24 months post-operatively to determine if performance improved after implantation and with device experience. Results from this patient cohort were compared to a larger group of adults from the MCIC with an acquired hearing loss.

The second aim of this study was to investigate aspects of auditory processing through linguistic, psychophysical, and electrophysiological testing, in order to determine if results offered insight into the early auditory experience of an individual. Whilst it remains difficult to accurately document the early hearing history for AEO-HL, the literature suggests that investigation in to the pre-operative measures used in this study may help to predict outcomes as they may be associated with the early auditory experience of an individual.

The third aim of this study was to investigate results from a self-report measure specifically designed for this project. Results were correlated with objective performance, and compared to a group of adults with acquired hearing loss from the MCIC.

The project aimed to establish whether any of the above-mentioned tests held any significant predictive value for post-operative objective outcomes, and whether these outcomes correlated to the recipient’s self-report of subjective benefit.

The specific hypotheses tested were:

1. That CI recipients with better language scores on standardised tests will attain better scores on speech perception testing with their cochlear implant than those with poorer scores.
2. That CI recipients with better speech intelligibility will achieve better scores on speech perception testing with their cochlear implant than those with poor intelligibility.

3. That CI recipients who use sign language will achieve lower scores on speech perception testing with their cochlear implant than those who use oral communication.

4. That temporal processing, as measured with acoustic stimuli pre-operatively using a gap-detection test, will be poorer in AEO-HL than in adults with an acquired hearing loss.

5. That the relationship between gap detection and speech perception is such that shorter gap detection thresholds are associated with better speech perception scores.

6. That those participants showing a measurable cortical waveform pre-operatively will achieve better speech perception scores post-operatively than those patients who did not have a measurable cortical response.

7. AEO-HL will demonstrate waveform latencies for auditory cortical responses which are delayed when compared to published norms for adults with normal hearing.

8. That those participants with lower scores on the Raven’s Matrices, non-verbal test of IQ, will achieve lower post-operative speech perception scores than those who achieved higher scores.

9. That the use of a self-report outcome measure will accurately reflect post-operative results on speech perception testing.

10. That results of the self-report outcome questionnaire will be lower for AEO-HL than those with an acquired hearing loss.
5 STUDY DESIGN

Approval for this project was granted by the Human Ethics Committee of the Royal Victorian Eye and Ear Hospital, project number 09/922H. All AEO-HL who attended the Melbourne Cochlear Implant Clinic between June 2010 and October 2014 who met the inclusion criteria, were invited to take part in this study. A total of 29 participants were recruited.

In addition to the clinical evaluations that were routinely administered in the standard care and management at the MCIC, this study completed further psychophysical, electrophysiological, language, non-verbal IQ and self-report questionnaire measures pre-operatively, and at 3, 12 and 24 months post-operatively. All electrophysiology and psychophysical data required testing at The University of Melbourne. A study questionnaire was administered at 12 and 24 months post-operatively for the AEO-HL group, and at 12 months post-operatively for the group of adults with an acquired hearing loss.

Compensation for the extra sessions was provided to the participants to cover travel expenses at the rate decided by the Melbourne University/CRC Research Committee in 2008. This involved re-imbursement of $10 per day, plus $20 per 100km travelled and the cost of all parking and tolls.

For each of the four data collection points, the extra sessions were restricted to one 2-hour session where possible. Extra sessions were required if the subject felt fatigued, or if any unforeseen technical difficulties were encountered.

Test Battery included:

- Standard Assessments at MCIC (2 hours)
- Audiological Evaluation (as per standard protocol – see Appendix A)
- Open Set Speech Perception Testing (as per standard protocol)
- Closed Set Speech Perception Testing (as per standard protocol)
Extra Assessments at Melbourne University: (2 hours)

- Cortical Auditory Evoked Potential Testing
- Gap Detection Test
- PPVT Receptive Language Test
- RPM Non-Verbal IQ Test
- Questionnaires

**Audiological Evaluation**

- Detailed History
- Standard Audiological/ENT investigations
- Pure tone Audiometry, Tympanometry, Contralateral Acoustic Reflexes
- Hearing Aid Evaluation, including insertion gain, to ensure that the hearing aid used met NAL prescriptive targets.
- If a participant was not currently wearing a hearing aid, but had aidable hearing, then a clinic loan aid was fitted for the duration of the assessments, and a three-month trial period allowed prior to testing.

**Summary of Data Collection Points for AEO-HL Group:**

**Table 1: Summary of data collection points**

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<thead>
<tr>
<th>Measure</th>
<th>Pre-Op</th>
<th>3mths Post-Op</th>
<th>12mths Post-Op</th>
<th>24mths Post-Op</th>
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<tr>
<td>Audiogram</td>
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<td>Hearing Aid Evaluation</td>
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<td>Questionnaire</td>
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5.1 Thesis Structure

Due to the different topics and tests covered in this project, each specific area of investigation was allocated its own chapter. It was anticipated that this structure would allow each topic to be discussed in detail; including a separate methods, results, and discussion section within each chapter. The thesis concludes with an overall discussion conclusion to bring all the findings together.

- Chapter 6: Study participants
- Chapter 7: Speech perception; methods, results, and discussion
- Chapter 8: Language; methods, results, and discussion
- Chapter 9: Gap Detection; methods, results, and discussion
- Chapter 10: CAEP; methods, results, and discussion
- Chapter 11: Non-Verbal IQ; methods, results, and discussion
- Chapter 12: Self-Report Outcome Questionnaire; methods, results, and discussion
- Chapter 13: An overall discussion chapter, tying in all facets of the project
- Chapter 14: Conclusions
6 PARTICIPANTS

Twenty-nine AEO-HL from the Melbourne Cochlear Implant Clinic Royal Victorian Eye and Ear Hospital, were recruited for this study. Participants were approached by their case manager during a routine appointment, and asked if they would like to participate in the project. All participants were medically, radiologically and audiologically suitable for a cochlear implant, as per the standard clinic guidelines (Leigh, Moran, et al., 2016). All participants used hearing aids which provided limited access to speech sounds, used English, AUSLAN (Australian Sign Language) or a combination of both, as their primary language, and had no anatomical deformity of the inner ears. All participants were between 18 and 65 years of age at the time of recruitment. Audiological guidelines at the MCIC suggest that a cochlear implant can be offered to an AEO-HL if there is evidence that auditory cues assist communication, and the prospective recipient has realistic expectations.

Four adults with a postlingually acquired hearing loss were recruited as a comparison group. Two of these volunteers received bilateral cochlear implants. In order to make a more meaningful comparison between results from the AEO-HL group and recipients with an acquired hearing loss, an additional 15 recipients were recruited for the temporal processing evaluation and labelled PostL1, and an additional 16 for the administration of the questionnaire, labelled PostL2. Additional data was required for these portions of the project as normative data was not available.

The additional 15 participants in the PostL1 group completed the gap detection testing as part of a separate study approved by the Royal Victorian Eye and Ear Hospital Human Research Ethics Committee (project number 14-1182H. The equipment and procedures used in the two studies were identical, and it was felt the additional recipients were required for adequate statistical power. The selection criteria for this CRC study were less stringent than the current study, but the average PTA for the group was 70dB in the ear to be implanted, with an average pre-operative CVC phoneme score of 26%. PTA was taken as the average hearing threshold at 500Hz,
1KHz and 2KHz. These 19 cochlear implant recipients (15 from the CRC study and four recruited for the current study) were referred to as PostL1.

In order to compare questionnaire results from AEO-HL with cochlear implant recipients with an acquired hearing loss, a further 16 were administered to recipients with acquired hearing loss at the MCIC. A total of 31 questionnaires were posted to recipients who were below 65 years of age, had a severe to profound hearing loss pre-operatively, with a PTA greater than 70dB, and received their cochlear implant between 2011 and 2014, ensuring that they had 12 months or more of device use at the time the questionnaire was administered. These selection criteria were used to ensure the participants matched the AEO-HL study group as closely as possible. Sixteen cochlear implant recipients returned the questionnaire. Questionnaire data for this group, referred to as PostL2 was only collected at the 12-months post-operative point.

A certain degree of participant selection bias existed within this project, both within the AEO-HL group, and PostL1 and PostL2 groups. While an attempt was made to discuss the project with all AEO-HL who fit the selection criteria and presented to the Melbourne Cochlear Implant Clinic between January 2011 and June 2014, many patients declined to take part. Ninety-one AEO-HL received a cochlear implant at the Melbourne CIC between this time, and only 29 were recruited for the project, which may represent a more motivated patient cohort than the general population of AEO-HL. The inclusion of participants with an acquired hearing loss in PostL1 and PostL2 was also likely to introduce a degree of bias, as it is possible that only the motivated CI recipients agreed to take part in the research project, or returned the questionnaire. It is important to note that these are not completely random samples, and therefore inferences made from the results obtained should be viewed cautiously.

All participants were counselled pre-operatively as to the possible outcomes with a cochlear implant, and had realistic expectations as to what benefits the cochlear implant may be able to provide. All participants received a Nucleus Freedom, CI422 or CI512 Cochlear Implant and used a CP810 or CP910 speech processor which utilised the ACE speech processing strategy.
Table 2: Participant demographics for the three study groups

<table>
<thead>
<tr>
<th>DEMOGRAPHICS</th>
<th>AEO-HL</th>
<th>POSTL 1</th>
<th>POSTL 2</th>
<th>SIGNIFICANCE</th>
</tr>
</thead>
<tbody>
<tr>
<td>AGE AT IMPLANTATION (YRS)</td>
<td>43</td>
<td>58</td>
<td>47</td>
<td>p&lt;0.001</td>
</tr>
<tr>
<td>RANGE</td>
<td>26-63</td>
<td>25-77</td>
<td>27-65</td>
<td></td>
</tr>
<tr>
<td>UNAIDED PTA: CI EAR RANGE</td>
<td>111 dB</td>
<td>81 dB</td>
<td>102 dB</td>
<td>p&lt;0.001</td>
</tr>
<tr>
<td>UNAIDED PTA: NON-CI EAR RANGE</td>
<td>65-125 dB</td>
<td>75-108 dB</td>
<td>72-125 dB</td>
<td></td>
</tr>
<tr>
<td>UNAIDED PTA: NON-CI EAR RANGE</td>
<td>107 dB</td>
<td>70 dB</td>
<td>85 dB</td>
<td>p&lt;0.001</td>
</tr>
<tr>
<td>BEST PRE-OP CVC PH RANGE</td>
<td>13%</td>
<td>62%</td>
<td>38%</td>
<td>p&lt;0.001</td>
</tr>
<tr>
<td>BEST PRE-OP CVC PH RANGE</td>
<td>0-72%</td>
<td>19-97%</td>
<td>0-83%</td>
<td></td>
</tr>
<tr>
<td>BEST PRE-OP CVC PH RANGE</td>
<td>8%</td>
<td>26%</td>
<td>13%</td>
<td>p&lt;0.001</td>
</tr>
<tr>
<td>BEST PRE-OP CVC PH RANGE</td>
<td>0-44%</td>
<td>0-62%</td>
<td>0-47%</td>
<td></td>
</tr>
<tr>
<td>GENDER</td>
<td>Female 16</td>
<td>Female 10</td>
<td>Female 8</td>
<td></td>
</tr>
<tr>
<td>GENDER</td>
<td>Male 13</td>
<td>Male 9</td>
<td>Male 12</td>
<td></td>
</tr>
</tbody>
</table>

A one-way ANOVA using the Tukey’s post hoc pairwise comparison was used to compare age, unaided PTA, and pre-operative CVC phoneme scores between the three groups. There was no significant difference between the ages of the AEO-HL and PostL2 group, but the PostL1 group was significantly older. The PTA in the ear to be implanted of the AEO-HL group was significantly higher than both the PostL1 and PostL2 group. The PTA in the contralateral ear was significantly different between all three groups. The pre-operative CVC phoneme score in the ear to be implanted was significantly higher for the PostL2 group than both the PostL1 and AEO-HL group. The pre-operative CVC phoneme score in the contralateral ear was significantly different for all three groups, with the AEO-HL being the poorest. It is possible that the differences in pre-operative PTA and CVC scores of the study groups contributed to the differences in post-operative speech perception and questionnaires results measured. These differences will be discussed more thoroughly in the corresponding thesis chapters.

Best pure tone average (PTA) was taken as the average hearing threshold at 500Hz, 1KHz and 2KHz in the better hearing ear. When the unaided hearing level exceeded the maximum output of the audiometer, the PTA was taken as 5dB above that maximum
output, ie 125dB (Ching et al., 2001). The best pre-operative phoneme score may have been obtained from either ear and in many cases, was from the contralateral ear, which generally had better hearing thresholds.

**Aetiology**

The aetiology of hearing loss was unknown for 33% of the AEO-HL Group, 44% of PostL1 Group and 55% of PostL 2 Group. A large percentage (27%) of the AEO-HL group had a hearing loss attributed to maternal Rubella. This group would have been born prior to the widespread use of the rubella vaccination, which was introduced in Australia in 1971 (Menser et al., 1985). A search of the paediatric cochlear implant patients at the Melbourne Cochlear Implant Clinic revealed only seven with hearing loss believed to be caused by maternal Rubella. Five of these children were born prior to 1989, one in 2007 and one was born overseas in 2011. It appears that the incidence of profound hearing loss attributed to maternal Rubella has significantly decreased.

Figure 1: Aetiologies – AEO- HL

- Unknown n = 10
- Genetic n=5
- Rubella n=8
- Prematurity n=3
- Ushers n = 1
- Waardenburg n=2
- Meningitis n = 1

Figure 2: Aetiologies – PostL 1 Group

- Unknown n=8
- Genetic n=2
- ME Disease n=3
- Meniere’s n=2
- Trauma n=2
- WVA n=1

Figure 3: Aetiologies - PostL 2 Group

- Unknown n-11
- Genetic n=3
- Rubella n=1
- Meniere’s n=2
- Otosclerosis n=2
- NF2 n=1
7 SPEECH PERCEPTION

7.1 Speech Perception Methods

Speech perception evaluations were completed with each of the 29 participants in the AEO-HL group pre-operatively, and at 3, 12 and 24 months post-operatively, and with PostL1 and PostL2 pre-operatively and at 3 and 12 months post-operatively. Participants used their preferred hearing aid volume and program settings pre-operatively, and their preferred speech processor settings post-operatively.

Open set testing was completed as per the standard MCIC protocol in a sound treated booth, see Appendix A (Leigh, Moran, et al., 2016). Pre-operatively, testing was completed in the right, left and binaural condition. When testing individual ears, participants were required to turn off the contralateral hearing aid. Post-operatively, participants were tested with their cochlear implant alone, and binaurally. Participants were required to listen to a list of CVC (Consonant-Vowel-Consonant) words and 2 lists of CUNY (City University New York) sentences, presented in quiet audition alone, via a loudspeaker, at a level of 65dB SPL. Participants were required to repeat the word or sentence that they heard, which was then scored by the audiologist. Any confusion in correctly interpreting a repeated word or sentence was clarified via finger spelling or a written word.

CUNY sentences (Boothroyd, 1985) were recorded by a female Australian speaker, and included 72 lists of 12 sentences. The length of the sentences varied from three to 14 words. There were 102 words per list, and the test was scored as the percentage of words correctly identified. CVC words were recorded by a male Australian talker, according to the rules stipulated to ensure a balanced list of phonemes (Peterson & Lehiste, 1962). CVC words are a list of 50 monosyllabic words scored according to the number of whole words repeated correctly, and the number of phonemes repeated correctly. Each list contained 150 phonemes; 50 vowels, and 100 consonants.
If participants scored greater than 30% correct on CUNY sentences in quiet, then CUNY sentences were presented in the presence of background noise in that particular condition, which comprised of multi-talker babble at a signal to noise ratio of +10dB. This was completed for each ear that obtained the score of >30%, and in the binaural condition. If the participant scored <30% for only one ear, this ear was not tested in noise to avoid distress for the participant. If the participant scored <30% with each ear individually, then testing in noise was not conducted in any condition. Two lists of CUNY sentences were presented in each condition.

The CVC phoneme score was chosen as the value of interest for analysis due to the expected language limitations of the AEO-HL group of participants. It has been suggested that individuals with a language score below age 7;5 may struggle with the language complexity of sentence perception tasks (Dowell et al., 2002).

The CVC phoneme score was compared to the phoneme score collected from 576 adults with an acquired hearing loss from the MCIC. All adults with a known acquired hearing loss who received either a CI24RE, CI422, CI512 or CI522 cochlear implant were selected from the database for analysis. A two sample t test was performed to analyse differences in scores between the two groups.

### 7.2 Speech Perception Results

#### 7.2.1 AEO-HL Post-Operative Speech Perception Results

The majority of AEO-HL participants gained significant benefit from their cochlear implant. Figure 4 compares the pre- and post-operative CVC phoneme score in the implanted ear. The ‘best’ post-operative score was chosen for analysis. Twenty-six of the 30 participants, or 87% of participants, showed an improvement in their post-operative phoneme score, when compared to their pre-operative score in the implanted ear. Two participants obtained the same score pre- and post-operatively (10% and 0%) and two demonstrated a post-operative decrease in scores, although for both of these participants the scores were close to chance level both pre- and post-
operatively (7% to 4%, and 10% to 7%). A paired t test was used to determine whether the difference between the pre- and post-operative scores were significant. The group average demonstrated significant improvement (p<0.001) in their best post-operative score (34.55%) when compared to the pre-operative score in the ear to be implanted (7.55%). To assess the strength of this finding, Cohen’s d was used to indicate the amount of difference between the phoneme scores. This difference corresponded to a large effect size of 1.23.

Figure 4: Pre-operative and post-operative CVC Phoneme scores in the implanted ear (%) for 29 AEO-HL.

Figure 5 shows the best post-operative CVC phoneme score compared to the best pre-operative CVC phoneme score, which was generally obtained from the contralateral, or non-implanted ear. Again, the majority of participants demonstrated significant improvement in speech perception performance, even when compared with their better hearing ear. Twenty-four of the 30 subjects, or 80% of subjects, showed an improvement in their post-operative phoneme score, when compared to their best pre-operative score. One participant obtained the same score pre- and post-operatively (10%) and five did not achieve a post-operative score greater than their
best pre-operative score. A paired t test was used to determine whether the difference between the pre- and post-operative scores was significant. The group average demonstrated significant improvement (p<0.001) in their best post-operative score (35.61%) when compared to the pre-operative score in the ear to be implanted (13.43%). This difference corresponded to a large effect size (Cohen’s d= 0.86).

![Chart showing pre-operative and post-operative CVC Phoneme scores for 29 AEO-HL subjects.](chart.png)

**Figure 5: Best pre-operative and best post-operative CVC Phoneme score (%) for 29 AEO-HL**

No significant correlation was observed between the best pre-operative phoneme score and the best post-operative score (r = 0.914, p = 0.313) however, a significant correlation was observed between the pre-operative CVC phoneme score obtained in the implanted ear, and the best post-operative phoneme score (r = 0.382, p = 0.041). Higher pre-operative phoneme scores in the ear to be implanted were associated with higher post-operative phoneme scores.
Figure 6: Relationship between pre-operative and post-operative CVC phoneme Score (%)

Figure 7 shows the improvement in speech perception, CI alone, over time for the AEO-HL group of participants. A General Linear Model was used, with participant as the random factor and time (post-operative assessment point) as a covariate. The ANOVA (GLM) was significant for time. Fisher post hoc pairwise comparisons showed that the improvement in phoneme score in the implanted ear alone from pre-operative to 3-month post-operative and the improvement from 3 to 12-months post-operative were significant. No significant improvement was observed between 12 months (mean 34.31%) and 24 months (mean 35.27%) post-operatively.

A one-way ANOVA was also performed to compare pre-operative scores in the implanted ear, and the ‘best’ pre-operative score, with the post-operative performance at 3, 12 and 24 months. A significant improvement from pre- to post-operative was found using Dunnett’s multiple comparisons when comparing both the implanted ear, and the best score. There was no significant difference found between the best pre-operative score and the pre-operative score in the implanted ear. The
asterix denote outliers. A significant improvement was seen between all data points, except 12 to 24-months post implantation.

![Graph showing improvements in speech perception over time](image)

**Figure 7**: Improvements in speech perception over time, for the 29 AEO-HL.

### 7.2.2 Comparison with results for adults with acquired hearing loss.

The 12 months post-operative CVC phoneme score for 576 adult cochlear implant recipients with acquired hearing loss from the Melbourne Cochlear Implant Clinic were analysed. All patients with a CI24RE, CI422, CI512 or CI522 cochlear implant were selected. The average phoneme score obtained CI alone was 64.44%, with a standard deviation of 24.27% and ranging from 0 – 98%.

CI alone, CVC phoneme scores from the 29 AEO-HL were compared to this group of 576 adults with an acquired hearing loss using a 2-sample t test. There was a significant difference in speech perception scores between the groups, with the adults with an acquired loss achieving a greater score (t=-7.02, p<0.001)
Figure 8: Comparison of speech perception results between the study cohort of 29 AEO-HL (mean 34%) and the 576 adults with an acquired hearing loss (mean 64.4%).

7.3 Speech Perception Discussion

Post-operative speech perception performance for this group of AEO-HL is significantly lower than the scores observed for the wider group of adults with postlingual hearing loss, although the group demonstrated a significant improvement in scores post-operatively. A large effect size was observed for the AEO-HL when post-operative performance was compared to pre-operative scores from the implanted ear and the contralateral ear, indicating that AEO-HL can gain significant overall improvement in their speech perception performance when they receive a cochlear implant.

Improvements in speech perception post-operatively have been widely reported in the literature for AEO-HL, although it is generally accepted that objective performance remains below that recorded for adults with a postlingual hearing loss (Kaplan et al., 2003; Kos et al., 2009; Santarelli et al., 2008; Schramm et al., 2002; Yoshida et al., 2008). The post-operative mean for this group of 29 AEO-HL of 34.55% is consistent with the figures reported in the literature of 31% for 28 participants in Straatman’s
study, 45% for 9 participants in Van Dijk’s study and 23% for 48 participants in Lammer’s study (Lammers, van Zanten, et al., 2015; Straatman et al., 2014; van Dijkhuizen et al., 2011).

The pre-operative phoneme score in the implanted ear was found to be correlated with post-operative performance for the AEO-HL while the PTA in the implanted or contralateral ear was not, suggesting that it is not necessarily the presence of any residual hearing pre-operatively that is important for outcomes, but rather the individual’s ability to use their remaining hearing to assist in communication. For those participants who have demonstrated that they can use auditory input to process a speech signal, it can be inferred that their central auditory system remains relatively intact and functional. For those participants who did have some residual hearing pre-operatively, but who were unable to utilise this hearing to gain any speech understanding, it is less certain whether their central auditory system has the necessary skills for speech perception. These results are supported by those of Dowell and colleagues (Dowell et al., 2002) who found that those subjects who were able to gain some benefit from their minimal pre-operative hearing, performed better with their cochlear implant, and by Yoshida and colleagues (Yoshida et al., 2008) who found that post-operative speech perception performance was correlated with early fitting of hearing aids and some use of hearing pre-operatively for oral communication.

Variability in outcomes for AEO-HL who receive cochlear implants has been well documented in the literature (Klop et al., 2007; Schramm et al., 2002). Results from the current study, however, show that whilst the group of recipients with acquired hearing loss have a significantly higher mean speech perception result than the AEO-HL, both groups show a similar spread of scores, from 0-90% for the AEO-HL and 0-100% for the group with acquired loss. A study by Santarelli and colleagues (2008) also found less variability than that which is generally documented. They felt this may be due to their homogenous patient group (Santarelli et al., 2008). The subjects in Santarelli’s study all used hearing aids pre-operatively, all used the same mode of communication, and all had auditory-oral speech therapy when younger. It was
suggested that the presence of auditory input throughout childhood via hearing aids, and the lack of any visual mode of communication, may have enabled the auditory cortex to remain sensitive to auditory information (The influence of communication mode will be discussed in more detail in Chapter 8). In support of this theory, Santarelli found that speech perception for AEO-HL continued to improve for three years post-operatively, whereas many studies, including this one, found that performance did not significantly increase beyond the 12 months post-operative point (Lammers, van Zanten, et al., 2015; Zeitler et al., 2012). The lack of further significant increases in speech perception beyond one year after surgery has also been documented for cochlear implant recipients with an acquired hearing loss (Blamey et al., 2013; Hamzavi et al., 2003).

Many studies have focussed on cochlear implant outcomes and predictive factors for cochlear implant recipients with an acquired hearing loss. It is generally acknowledged that restoration of a degraded auditory signal with a cochlear implant enables the recipient to gain significant auditory benefit. A retrospective study by Dowell and colleagues revealed that early vs late onset of significant hearing loss, duration of severe to profound hearing loss, age at implantation and pre-operative auditory skills were found to have a significant association with post-operative speech perception (Dowell et al., 2004). A large retrospective study by Blamey and colleagues investigated factors affecting performance with 2251 adult cochlear implant recipients with an acquired hearing loss across 15 international cochlear implant centres as a follow up to a previous study performed 15 years earlier (Blamey et al., 1996; Blamey et al., 2013). Performance increased as a function of duration of implant experience up to 3.5 years after implantation, with the most dramatic improvements occurring during the first 12 months of device use. In the original 1996 study, duration of deafness had a strong negative effect on performance, age at implantation had a slight negative effect, which increased after age 60, and duration of experience had a significant positive effect. In 2011, length of cochlear implant experience became the most significant factor, and age at implantation and duration of deafness became less significant. Blamey suggested that the changes in these factors may be related to the
widening of patient selection criteria, and improvements in technology and surgical technique. With both studies, the amount of variance unaccounted for remained large.

With regards to outcomes for cochlear implants for AEO-HL, the literature suggests that prior use of residual hearing, the use of hearing aids, and communication mode may influence outcomes (Kos et al., 2009; Santarelli et al., 2008; Yoshida et al., 2008). Although both participant groups in the current study demonstrated significant benefit from their cochlear implants, the differences observed between the two groups is assumed to be due to the differences in age at onset of significant hearing loss. The large degree of variability observed in post-operative outcomes in cochlear implant recipients with an early onset hearing loss may be due to differing degrees of impact of hearing loss on the developing auditory system and language processing areas of the cortex. The effects of auditory deprivation and cross modal plasticity have been well documented, and will be discussed further in Chapter 13. The consequences of early onset hearing loss are difficult to measure accurately for an individual, making it challenging for clinicians to counsel prospective implant recipients appropriately. There exists a need for further research to identify potential clinical tools which may be able to provide some predictive value for this patient group.
8 LANGUAGE

As the results from the previous chapter have shown, the post-operative CVC phoneme scores obtained from the current study cohort of AEO-HL are significantly lower than those seen in adults with an acquired hearing loss. The literature would suggest that part of the reason lower results are seen for this patient cohort may be due to decoupling of the primary auditory cortex from higher-order association areas (Kral et al., 2006). This decoupling may interrupt the necessary feedback loops and top-down, bottom-up processing of speech perception (Boothroyd, 1985), interrupt the connectome (Kral et al., 2016), or lead to the cross-modal remapping of previously under-utilised secondary association areas (Buckley & Tobey, 2011; Nishimura et al., 1999). For the purpose of investigating aspects of language and communication, which may provide insight into the appropriate development of the secondary association areas, the receptive language, speech intelligibility and communication mode of the current study participants were investigated.

Specifically, the following hypotheses were tested:

Hypothesis 1: That CI recipients with better language scores on standardised tests will attain better scores on speech perception testing with their cochlear implant than those with poorer scores.

Hypothesis 2: That CI recipients with better speech intelligibility will achieve better scores on speech perception testing with their cochlear implant than those with poor intelligibility.

Hypothesis 3: That CI recipients who use sign language will achieve lower scores on speech perception testing with their cochlear implant than those who use oral communication.
8.1 Language Methods

In order to investigate the impact of early hearing loss on the communication abilities of this study cohort, three aspects of language and communication were measured; the receptive language as estimated by the Peabody Picture Vocabulary Test PPVT, speech intelligibility as measured by the Northern Technical Institute of the Deaf (NTID) rating scale, and communication mode.

Receptive Vocabulary Testing (PPVT)

The Peabody Picture Vocabulary Test (PPVT), 4th edition (L Dunn, 1997) was used to estimate a receptive vocabulary standard score of each participant enrolled in the study, prior to receiving their cochlear implant. The PPVT-4 is a measure of single word receptive vocabulary, and had two parallel sets of questions, Form A and Form B. Form A was used in this study. The test consisted of 204 items, with each item containing four different pictures. The examiner administered the test in accordance with the manual, and presented the test item either live voice, or in a written format. The participant was then required to indicate which of the four pictures best represented the test item. The item was not able to be presented via sign language due to the potential for iconicity, or the sign itself offering some clue as to the test word (Geers & Brenner, 2003). Lipreading, however, was allowed. The task required an understanding of concrete words, abstract words and concepts, and was administered according to the test manual. The manual for the PPVT-4 provides normative data from the ages of 2½ to 90+ years. The participant’s raw score was then converted to a standard score, percentile rank, normal curve equivalent, and an age-equivalent score in years.

Speech Production (NTID)

The NTID (Northern Technical Institute of the Deaf) is a five-point scale used to rate speech intelligibility:
• 1 = Speech is completely unintelligible

• 2 = Speech is very difficult to understand - only isolated words or phrases are intelligible

• 3 = With difficulty, the listener can understand about half the content of the message (intelligibility may improve after a listening period),

• 4 = Speech is intelligible with the exception of a few words or phrases

• 5 = Speech is completely intelligible.

Speech Intelligibility was rated pre-operatively using the NTID rating scale for judging speech intelligibility (Subtelny, 1980). The use of Rating Scales for measuring intelligibility enables the listener to record their perception of overall intelligibility on a continuum from completely unintelligible (where the listener is unable to understand the speech at all) to completely intelligible (where the speech can be clearly understood). Rating scales enable clinicians to gain a general indication of how well an individual’s speech will be understood by others (Doyle, 1987).

The use of the NTID has appeal as a quick means of assessing overall intelligibility. The NTID has been shown to be used effectively by audiologists with high inter- and intra-rater reliability to rate hearing-impaired children’s speech, and that training in the use of such scales does not appear necessary for reliable judgements (Doyle, 1987). The use of a rating scale was chosen over the word identification method due to the ease with which a rating scale can be used within a clinical setting. The spontaneous speech of the participant was used to generate the intelligibility rating. It was felt this method would be more suitable for this participant group, as there exists the possibility that the literacy level required for the reading of a book passage may be too high for some participants, hence negatively affecting their intelligibility (Subtelny, 1977).
Each participant in this study was given a rating by two audiologists working in the Melbourne Cochlear Implant Clinic, or in some cases the second rating was given by a Master of Clinical Audiology student from the University of Melbourne. In the case of a disagreement between the two ratings, an average was taken, and if necessary was rounded up to the highest whole number to enable correlations with speech perception and other study measures. Percentage agreement and Cohen’s Kappa was used to demonstrate the level of agreement between the two audiologists. As the NTID has only five rating scales, it is possible that an overly inflated percentage agreement could be observed, therefore Cohen’s Kappa was also performed. The Kappa statistic estimates the degree of consensus between the two audiologists, after correcting for the effects of chance, where 0.0 indicates no agreement beyond that which could be expected due to chance alone, and 1.0 indicates perfect agreement (Kohen, 1960). A score of greater than 0.7 is generally acknowledged to be a good agreement between two audiologists (Landis & Koch, 1977).

**Communication Mode**

Participants were divided into two separate communication categories based on their use of sign language. The ‘Oral’ category comprised those participants who did not use sign language in any form. The ‘Signing Support’ category comprised the remaining participants, who exhibited varying dependence on sign language. There was no participant in this study who relied entirely on manual communication, that is, all participants could either lipread to a certain extent or produce some spoken language.

### 8.2 Language Results

The following section demonstrates the range of receptive vocabulary seen for the AEO-HL study group, the correlations between vocabulary and post-operative speech perception scores and speech intelligibility. Correlations are shown between intelligibility and speech perception, and comparisons made between the vocabulary, speech perception and intelligibility of the oral group and the signing support group.
8.2.1 Receptive Language

For the PPVT, a standard score of 100 is the mean in the normal population with a standard deviation of 15. A score of less than 70 represents a score more than two SD below the mean, and indicates a significant receptive language delay. The mean PPVT standard score for the group of 29 AEO-HL was 79.90, with a standard deviation of 19.09 and a range of 32-107. 24% of participants scored more than two SD below the mean, 21% scored between one and two SD below the mean, and 55% of participants scored in the normal range (+/- 1SD of the mean). No participant scored more than one SD above the mean. No significant difference in speech perception scores was observed between those who scored more than one SD below the mean and those who scored in the normal range (+/- one SD of the mean, t=-0.21, p=0.834) or between those who scored more than two SD below the mean and those who scored in the normal range (t = -0.42, p=0.680).

It is evident from the above data that approximately one quarter of the participants in this study demonstrated well below average receptive language skills.

![Relationship between PPVT Standard Score and Post-Operative CVC Phoneme Score for 29 AEO-HL participants](image)

**Figure 9:** Relationship between PPVT Standard Score and Post-Operative CVC Phoneme Score for 29 AEO-HL participants
Figure 9 shows the comparison between each participant’s receptive language score and post-operative CVC phoneme score. The solid line represents the population mean, and the dashed lines represent one standard deviation above, and below, the mean. Post-operative speech perception was not correlated with the PPVT receptive language standard score ($r = 0.074$, $p = 0.7$). While there is no significant direct correlation, all those participants with a standard score less than 60 did not achieve a CVC phoneme score greater than 50%. It is possible that a reasonable level of language is a requirement for a good speech perception score.

### 8.2.2 Speech Intelligibility

The average speech intelligibility rating for the study group was 3.82, with a standard deviation of 1.00 and a range of 2-5. Error! Reference source not found. shows the correlation between speech intelligibility, as measured by the NTID rating scale, and post-operative CVC phoneme scores. A significant positive correlation was observed, whereby those participants with a higher speech intelligibility rating achieved better post-operative CVC phoneme scores ($t=0.602$, $p = 0.001$). While the speech production ability of the participants is a possible confounding factor, every precaution was taken to avoid this. As observed in Figure 10, no subject in this study achieved an NTID rating of one, that is, there was no participant with completely unintelligible spoken language.

Regression analysis showed that a significant proportion of variance (36%) in post-operative CVC phoneme scores was accounted for by the speech intelligibility rating in this study cohort.

When investigating the reliability of the two ratings of speech intelligibility per participant, a percentage agreement of 55% was obtained, indicating that in 55% of cases, or for 16 participants, both audiologists gave the exact same rating. A Kappa statistic of 0.480 was also observed, which indicates a moderate agreement between the two audiologists when adjusted for chance (Landis & Koch, 1977; Viera & Garrett, 2005). The Kappa statistic and percentage agreement achieved using this rating scale
with two audiologists did not give the degree of agreement that was anticipated. Whilst a moderate agreement was deemed to be valid for analysis, further work is warranted to determine a more appropriate method of assessing speech intelligibility when using two different raters.

No significant correlation was observed between speech intelligibility and receptive language score (r=0.105, p=0.586) in this cohort.

![Diagram](image)

**Figure 10**: Relationship between speech intelligibility and post-operative CVC phoneme scores for 29 AEO-HL.

### 8.2.3 Communication Mode

Figure 11 demonstrates the observed difference in post-operative speech perception between those participants who used oral communication, and those who used Signing Support. The mean post-operative CVC phoneme score for the oral group was 43.13%, with a standard deviation of 23.41 and a range of 7 – 88%. The mean phoneme score for the Signing Support group was 24.29% with a standard deviation of 17.86, and a range of 0 – 68%. Mode of communication was found to have a significant effect on speech perception outcomes. Those participants who used an exclusively oral
mode of communication scored significantly higher on their post-operative speech perception testing, than those who used sign language, either exclusively or in addition to oral language (t=2.45 p=0.011).

Figure 11: Post-operative CVC phoneme score as a function of communication mode. Error bars indicate +/- 2 SE of the mean.

There was a significant difference in speech intelligibility, as measured by the NTID, between the oral and signing support group, with the oral group mean speech intelligibility of 4.27 significantly higher than that of the signing support group (3.36) (t=2.72 p=0.006).
Figure 12: Speech Intelligibility as a function of communication mode. Error bars indicate +/- 2 standard errors of the mean.

There was no significant difference in receptive vocabulary, as measured by the PPVT, between the oral and signing support groups (t=1.33 p=0.097).

8.2.4 Language Discussion

Of the three aspects of language investigated in this study, speech intelligibility and communication mode were observed to have significant relationships with post-operative speech perception scores. Receptive language, as measured by the PPVT standard score, was not found to be significantly correlated with speech perception or speech intelligibility. There was no significant difference in receptive vocabulary between participants who used an exclusively oral communication mode, and those who utilised signing support.

The PPVT receptive language results of this study compare the participant’s raw score with the scores of people of the same age. For this test, a standard score of 100 is normal. A score of less than 70 indicates a significant receptive language delay. It is evident from the results obtained that 24% of subjects in this study demonstrated
below average receptive language skills, with a standard score more than two SD below the mean. The average language score for the 29 subjects was 79.8, which falls below one standard deviation (SD) of the general population mean (85 – 115). The lack of observed correlation between the receptive language score and post-operative speech perception (r=0.074, p=0.703) is in contrast to a previously published study, which found a moderate correlation (r=0.395, p<0.05) (Rousset et al., 2016). Although there was some overlap in participants between these two papers, the additional subjects unique to each study may have generated the observed difference in results. The published study used clinical results, with more participants. There were no participants in the current study who scored greater than 1SD above the mean, whereas 9 (21%) scored more than 1SD above the mean in the previous study. The average standard score for the previous study was 85.95, compared to 79.8 in this study, indicating that there may have been a subgroup of participants in the previous study with higher receptive language scores. It is unlikely that there is a complete lack of relationship between language and speech perception, but it may be more complex than a linear correlation. While a linear correlation was not observed in the current study, it is worth noting that all those participants with a PPVT standard score less than 60 scored less than 50% on speech perception, suggesting that a reasonable language ability is required to attain a reasonable speech perception score.

When evaluating open set speech perception skills for children, or AEO-HL, it is possible that the results may be influenced by speech production limitations and lack of lexical knowledge, as well as hearing abilities (Paatsch et al., 2004). Blamey and colleagues showed that there was a strong relationship between speech perception, language and speech production of deaf children using both hearing aids and cochlear implants (Blamey et al., 2001). Although the current study did not demonstrate any correlation between speech intelligibility and receptive vocabulary, or speech perception and receptive vocabulary, nearly 25% of the group did show delayed language skills. It is reasonable to assume that for the participants in the current study, limited receptive language would also make it difficult to ‘fill in the gaps’ when performing speech perception tasks with a degraded auditory signal. It has been
shown that if white noise substitutes a single phoneme in a recorded word, normally hearing subjects not only fail to notice the substitution, they report to have clearly heard it. The brain is able to fill in the missing sound using top down processing (Warren, 1970). It is likely that most cochlear implant recipients with an acquired hearing loss can draw on their language skills and world knowledge to overcome loss of information from a degraded auditory input. This top-down processing ability may be less developed in AEO-HL participants, contributing to their generally poorer speech perception outcome. CVC words were chosen as the speech perception test measure in this project, as opposed to sentence material, to try and minimise the potential influence of top-down processing.

Adult cochlear implant candidates with a significant early onset hearing loss often struggle to accurately report on their hearing history. This group often tend to show large variability in post-implant outcomes, from little speech perception benefit to great benefit, therefore it is important to try to identify factors which may assist with counselling. The results from this study, and those of van Dijkhuizen and colleagues, suggest that a candidate’s speech intelligibility may possess some predictive value for cochlear implant speech perception outcomes. It has been proposed by van Dijkhuizen, that a review of the literature suggests that the presence of intelligible speech in AEO-HL may be a necessary condition for success with a cochlear implant (van Dijkhuizen et al., 2011). It is possible that intelligibility may reflect the amount of hearing an individual retained during infancy and the amount of auditory input and stimulation they received (Santarelli et al., 2008; Teoh et al., 2004). Van Dijkhuizen’s study investigated cochlear implant results from nine adults with reasonable speech intelligibility. They found a significant correlation between intelligibility and post-operative speech perception. They also found that the most intelligible subjects in their study all used oral communication as opposed to total communication. There are potential limitations involved in the interpretation of this study, however, as only participants with reasonable speech intelligibility were included. No inferences can be made about potential recipients with poor intelligibility (van Dijkhuizen et al., 2011). The finding of better post-operative outcomes correlating with better speech
intelligibility was also observed in the current study. The recipients using completely oral communication demonstrated significantly more intelligible speech than those who relied on some signing support. Other studies in the literature have also shown differences in intelligibility between oral and manual modes of communication for both children and adult cochlear implant recipients (Osberger, 1994; Sims et al., 1980; Tobey et al., 2004).

Of the 29 participants in this study, 14 (48%) used some form of sign language to supplement their communication. Those participants who used a purely oral mode of communication scored significantly better on post-operative speech perception with their cochlear implant than those who used signing support (p=0.011). There was no difference in receptive language observed between the two groups, but significantly greater speech intelligibility ratings were recorded for the oral group. These results reflect those generally observed in the literature (Habib et al., 2010; van Dijkhuizen et al., 2011).

Many studies support the finding that better speech perception results are obtained by those recipients who focus entirely on their hearing and use a purely oral mode of communication (Kaplan et al., 2003; Kos et al., 2009; Santarelli et al., 2008; Yoshida et al., 2008). Two separate studies by Kos and Kaplan (2009 & 2003) both found significantly better speech perception outcomes for AEO-HL who used oral language, as opposed to those using sign language. Kos proposed that knowledge of the structure of oral language significantly influenced the post-operative performance for AEO-HL with cochlear implants (Kos et al., 2009). Kaplan et al found that even though all participants in both the oral and signing group were satisfied with their implant and reported a significant improvement in their quality of Life (QoL), significantly greater speech perception performance was measured for the patients who used oral communication as compared to those using Total Communication (p = 0.05) (Kaplan et al., 2003).

The current study showed no difference in receptive vocabulary between those participants who used oral language, and those who used signing support. Similar
results have been observed for children with cochlear implants (Dawson et al., 1995; Kirk et al., 2000; Yanbay et al., 2014). A recent study by Yanbay and colleagues investigated language outcomes for a group of 42 children enrolled in different communication programs at school; auditory/oral, auditory verbal therapy, or signing support. Once the results had been adjusted for socio-economic status and family involvement, which were found to have a significant effect on results, no significant difference in receptive vocabulary (as measured with the PPVT) was found across the three modalities (Yanbay et al., 2014). Results are also supported by those of Dawson, who found that communication mode was not a significant predictor of receptive vocabulary as measured with the PPVT (Dawson et al., 1995), and by Kirk, who investigated language outcomes in 56 children using oral communication and 50 children using total communication. Although significant differences were observed in speech perception between the groups, receptive and expressive language scores were not statistically different (Kirk et al., 2000). It was suggested that the discrepancy may be due to the fact that language testing was performed in the child’s preferred communication mode, whereas speech perception testing was performed audition alone.

There are always difficulties interpreting results for children, or adults, who use some degree of sign language due to the very nature of the communication mode. There is a great deal of variability in the amount of reliance on sign between individuals, which introduces some degree of uncertainty when analysing results. This was observed by Yanbay in her large study of 42 children (Yanbay et al., 2014), and was certainly seen in this study. Some participants in the current study relied on Auslan interpreters for discussions with clinicians and researchers, whereas others only used Auslan when speaking with deaf friends but were confident in their ability to listen and lipread when participating in this study. There were no participants in this study who relied entirely on Auslan with no ability to use some degree of oral language. It has been mentioned in the literature that most children born profoundly deaf are the children of hearing parents, many of whom may never learn to communicate with their deaf child effectively using sign language, generating a potential for language delay (Svirsky et al.,
Humphries and colleagues (2015) encourage parents of signing children to attempt to learn sign language, in order to create a stronger family unit and provide appropriate support for their child (Humphries et al., 2015). All participants in this study were deaf children of hearing parents, so it is possible that a strong signing language model was not provided at home. This may be a contributing factor to the vocabulary delay seen in many participants in this current study.

An additional inherent limitation in interpreting results regarding communication mode, is the uncertainty regarding the nature of any relationship. It cannot be assumed that any observed correlation is the result of a causal effect (Dowell et al., 2002). It is unclear if the lower speech perception abilities observed are due to the fact that the cochlear implant recipient uses sign language, or if they use sign language due to the fact that they have additional needs, or have received an implant after the critical period for oral language development. The participants in the current study all had an early onset significant hearing loss, and received a very ‘late’ cochlear implant as an adult. These recipients did not have access to a robust auditory signal when younger, and the introduction of sign language would have been a logical step for many families to take.

Results from the current study, as well as many in the literature, would suggest that the development of language, in particular receptive vocabulary, may not be dependent on a purely oral approach to language. It appears that the foundations for language and vocabulary acquisition can be generated through sign language as effectively as through oral language. When we investigate aspects of oral language however, in particular speech production and speech perception, an oral background appears to be crucial. In this study, and other published studies, the use of sign language was associated with significantly poorer levels of speech perception through the cochlear implant, and significantly poorer speech intelligibility. For AEO-HL cochlear implant recipients, the disruption of natural language development due to a lack of appropriate audition during infancy appears to significantly affect their eventual speech perception, and speech production, abilities. It has been suggested that an
appropriate and stimulating auditory environment in the early years is crucial for the development of auditory skills (Dowell et al., 2002). It would appear that for an AEO-HL cochlear implant recipient to achieve a reasonable level of post-operative speech perception ability with their cochlear implant, they may need more exposure to audition when younger. As it is very difficult to obtain an accurate hearing history from these recipients, the evaluation of their speech production abilities and communication mode may be able to provide some insight as to their hearing levels, use of amplification and early intervention as an infant.

### 8.3 Language Conclusion

Three language measures were investigated for possible associations with cochlear implant outcomes for a group of AEO-HL recipients. Specifically, the following hypotheses were tested in this chapter:

**Hypothesis 1:** That CI recipients with better language scores on standardised tests will attain better scores on speech perception testing with their cochlear implant than those with poorer scores.

**Hypothesis 2:** That CI recipients with better speech intelligibility will achieve better scores on speech perception testing with their cochlear implant than those with poor intelligibility.

**Hypothesis 3:** That CI recipients who use sign language will achieve lower scores on speech perception testing with their cochlear implant than those who use oral communication.

Results of the current study showed that receptive language, as measured with the PPVT, did not appear to be significantly correlated with the speech perception, speech intelligibility, or communication mode of the study group. Results did not support Hypothesis 1. The study participants who had better speech intelligibility, and utilised an exclusively oral communication mode, performed better on speech perception testing with their cochlear implant than those with poorer intelligibility. Results did
support Hypothesis 2. These results may be due to a better ability to utilise the auditory signal provided by the cochlear implant, possibly due to more hearing and exposure to oral language when young. Those recipients who used some form of sign language for communication had poorer intelligibility, and lower post-operative speech perception scores. Results support Hypothesis 3.

The findings of the present study suggest that individual cochlear implant benefit can be predicted to some degree from the hearing history and speech intelligibility of the participant. This supports the inclusion of a speech intelligibility measure in the assessment protocol for AEO-HL considering cochlear implantation.
9 GAP DETECTION

As the results from the previous chapter have shown, post-operative speech perception was able to be predicted by pre-cochlear implant ratings of speech intelligibility and adult’s descriptions of their communication mode. Due to the difficulties in obtaining an accurate hearing history for AEO-HL, rating speech production is a clinically useful tool to provide insight into adults’ prior hearing levels and use of audition. Additional information about the development of the auditory system as a result of early auditory experience may be provided by the investigation of gap detection thresholds. It has been shown that this aspect of temporal processing is adult like by 6-12 months of age (Trehub et al., 1995), therefore it is possible that intact gap detection abilities may imply that early access to audition had been available and appropriately utilised.

Specifically, the following hypotheses were tested:

Hypothesis 4: That temporal processing, as measured with acoustic stimuli pre-operatively using a gap detection test, will be poorer in AEO-HL than in adults with an acquired hearing loss

Hypothesis 5: That the relationship between gap detection and speech perception is such that shorter gap detection thresholds are associated with better speech perception scores

9.1 Gap Detection Methods

Temporal processing was evaluated pre-operatively using a gap detection task, performed using Macarena software developed at the University of Zurich by Wai Kong Lai. It is hypothesised that those subjects with better temporal processing skills pre-operatively, as measured by smaller acoustic gap detection thresholds, will achieve better speech perception results with their cochlear implant, and that the AEO-HL study group would have poorer gap detection thresholds than the adults with an acquired hearing loss in group PostL1.
The gap detection procedure involved the participant listening to three sounds, one of which contained a gap in the middle. The participant was required to identify which of the three signals contained the gap. For the first trial, the gap was 60ms in duration, ensuring it could easily be heard, but then decreased in duration to the point where it could no longer be identified. The software used a 3IFC (three interval forced choice) paradigm to converge on 70.7% correct (Levitt, 1971). An adaptive two down one up procedure was used; after two correct responses the gap decreased and after each incorrect response it increased. Initially the step size was 4ms, reducing to 2ms. The testing proceeded until eight reversals occurred. The gap duration of the last four reversals was averaged and taken as the gap detection threshold, or the smallest gap that the subject could detect. Two trials were completed, following a training trial to familiarise the subject with the task, and an average was taken for analysis. While many studies have focussed on gap detection thresholds post cochlear implant surgery, this methodology is subject to the limitations of electrical stimulation and speech processor characteristics (Park, Won, et al., 2015), and it has been suggested that the cochlear implant itself does not provide sufficient temporal information to promote adequate temporal resolution (Duarte et al., 2015). For these reasons, and the fact that the use of gap detection as a possible predictor of outcome was the focus in the current study, gap detection was measured pre-operatively with an acoustic signal.

Two different stimuli were used, a broadband (BB) stimulus, and a 500Hz sinusoid. Both stimuli had a total duration of 1000ms, including the gap. The duration of stimulus either side of the gap was identical, and adjusted for each stimulus of differing gap duration, to ensure that the total stimulus duration remained at 1000ms. For the largest gap duration of 60ms, the duration of stimulus either side of the gap was 470ms, and for the smallest gap duration of 2ms, the duration of stimulus each side was 499ms.

The BB stimulus was generated using a white noise, with a Butterworth lowpass filter (Robertson & Dowling, 2003) with a cut off of 6000Hz and an attenuation rate of 24dB.
per octave applied. The second stimulus used was a 500Hz sinusoid. The preserved phase condition (where the phase of the signal continues after the gap as though the gap did not exist) was used to minimise spectral splatter, which is the term given to the higher and lower frequency energy generated by the onset and offset of the tone at the edges of the gap. Remaining splatter was masked using white noise with a notch at 500Hz. The spectral shape of the noise was identical to that generated by Moore and presented at 30dB spectrum level below the average level of the tone (Moore & Glasberg, 1988). This masking noise was sufficient to mask any spectral splatter below 3KHz. It was assumed that the small amount of splatter still present above 3KHz was not audible to the participants involved in this study.

Each stimulus used in the gap detection task was presented at a sensation level of 30dB (Fitzgibbons & Gordon-Salant, 1987) for those participants who had low enough thresholds to allow for this. The signal was presented via 3A tubephones, through an audiometer connected to an external Edirol Sound Card which enabled the generation of additional volume. Both ears were tested. The tubephones used had a maximum output of 135dB SPL. Audiometric threshold was determined immediately prior to testing with a 500Hz pure tone threshold using the same audiometer and tubephones. Gap detection was then performed through the external sound card, and the dial reading converted to dB HL via the use of a previously determined calibration level generated with a Bruel and Kjaer Sound Level Meter and 2CC coupler. As the maximum output for the gap detection task was 130dB SPL for the 500Hz signal, and 135 dB SPL for the Broadband signal, for those participants with a pure tone threshold greater than 100dB the full sensation level was not available. Audiometric threshold was not determined for the BB signal, therefore presentation level was based on the threshold determined at 500Hz. Each stimulation level was checked for comfort with the participant using a loudness scale, validated by Hawkins for use in setting the SSPL90 in hearing aids (Hawkins et al., 1987). Refer to Appendix B.

Gap Detection thresholds obtained from the AEO-HL recipients were correlated with post-operative speech perception results, communication mode and speech
intelligibility. They were also compared to a group of cochlear implant recipients with an acquired hearing loss. Four of these recipients were those enrolled in the current study, and the remaining 15 were enrolled in a large HEARing CRC sponsored study approved by the Royal Victorian Eye and Ear Hospital Human Research Ethics Committee (project number 14-1182H). Further discussion on these additional participants can be found in Chapter 6: ‘Participants’. It must be acknowledged that these additional 19 participants do not represent a true control group. They were not matched in any demographic to the study cohort of AEO-HL, and due to the nature of their involvement, it is likely that they represent a smaller subset of motivated cochlear implant recipients as opposed to cochlear implant recipients in general. The equipment and procedures used in the two studies were identical, and it was felt the additional recipients were required in order to make a meaningful comparison. The acquired hearing loss study group only performed the gap detection task using the 500Hz sinusoid signal in the ear to be implanted.

9.2 Gap Detection Results

9.2.1 AEO-HL Group: Gap Detection with 500Hz Sinusoid

Gap detection thresholds using the 500Hz sinusoid signal, were able to be measured in the ear to be implanted in 18 AEO-HL. Fourteen of these participants had a 500Hz threshold at or below 100dB, enabling the desired 30 dB sensation level to be used. One participant had a 25 dB SL, two had a 20dB SL and one a 15 dB SL. Statistical analysis was done including and excluding these four recipients. Four participants were unable to detect the gap at the maximum duration, two of these did not have the full 30dB SL available to them. A gap detection threshold of 60ms was assigned to these participants. Gap detection thresholds were measured in 17 participants in the contralateral ear. As hearing thresholds in the contralateral ear tend to be better than the ear to be implanted, the full 30 dB SL was able to be achieved for all participants. One participant was unable to complete the task with the contralateral ear and was therefore assigned a threshold of 60ms.
Figure 13 shows a scatterplot of 500Hz gap detection thresholds in the ear to be implanted vs post-operative phoneme score, showing a general trend, with scores decreasing with larger gap detection thresholds. The squares represent the four recipients who did not have the full 30dB SL available to them. While there is not a significant linear correlation, inspection of the data shows that all speech perception scores over 50% were for participants with gap detection thresholds less than 10ms. Those participants who achieved a gap detection threshold less than 10 ms (n=7) scored significantly better on post-operative speech perception testing than those who had a gap detection threshold greater than 10 ms (n=11) using the Mann-Whitney test (W=95.5, p= 0.0049). A logarithmic transformation was used to fit the data to a negatively exponential function. Results show a significant correlation between the 500Hz gap detection threshold in the ear to be implanted, and the postoperative CVC phoneme score (p=0.01). In this instance, the gap detection threshold accounted for 34.8% of the variance in the post-operative performance.

Figure 13: Regression analysis of the 500Hz gap detection threshold in the ear to be implanted and post-operative performance for the 18 AEO-HL participants.
Correlation analysis was repeated with the four recipients unable to achieve the suggested 30dB SL removed, but no difference in results was observed (\(\rho = -0.464, p = 0.095\) vs \(\rho = -0.425, p = 0.078\) for all 18 participants). All subsequent analysis was therefore performed with all 18 participants included.

No significant difference in 500Hz gap detection threshold was observed between the ear to be implanted, and the contralateral ear, for the group as a whole, and for individual recipients. A Wilcoxon signed rank test was used for the 14 recipients who were able to perform the task for both ears (Wilcoxin statistic = 32.0, \(p = 0.364\)), and the Mann Whitney was run with all recipients included (\(W = 344, p = 0.519\)). There was no observed correlation between the gap detection threshold in the contralateral ear and post-operative performance.

### 9.2.2 AEO-HL Group: Gap Detection with Broad Band Signal

The gap detection task with the BB stimulus proved difficult for many recipients. Almost all participants reported that the BB stimulus was much softer than the 500Hz stimulus, even though the measured SPL was identical, potentially due to the configuration of their hearing loss. A significant difference was observed between the Median 500Hz gap detection threshold (18.25ms), and the Median BB gap detection threshold (59ms) in the ear to be implanted for the AEO-HL group (\(W = 239.5, p = 0.028\)), and the contralateral ear (\(W = 227.5, p = 0.046\)). There was no significant difference between the BB gap detection threshold in the ear to be implanted and the contralateral ear (\(W = 30.0, p = 0.107\)).

Regression analysis with transformed data showed no significant correlation between the BB gap detection threshold in the ear to be implanted and speech perception (\(p = 0.828\)) or between the contralateral ear and speech perception (\(p = 0.939\)).
9.2.3  AEO-HL Group: Relationship between Gap Detection and Language Measures

The relationship between temporal processing and language measures was investigated by analysing gap detection thresholds, speech intelligibility, and communication mode for the 18 participants who had results available for all tests.

Regression analysis showed that speech intelligibility was highly significant (p=0.002) and accounted for 46.5% of the variance in post-operative phoneme scores in this subset of 18 participants. Although gap detection and speech intelligibility were not significantly correlated with each other (r=-0.382, p=0.118) there was some degree of correlation between the two factors, and it is possible that with more participants, a significant correlation may have been observed. It is reasonable to assume that a correlation between the two factors does exist, as both speech intelligibility and temporal processing appear to be related to access to auditory input during infancy.

Multiple regression analysis was performed with the factors that demonstrated a significant relationship (p=0.05) with the post-operative speech perception results: the log of the 500Hz gap detection threshold in the ear to be implanted (F=4.68, p=0.001) and speech intelligibility (F=8.98, p=0.047). The 500Hz gap detection threshold in the CI ear and speech intelligibility accounted for 59.22% of the variance in post-operative outcomes in this subset of 18 participants, and gave a regression equation of:

Post-op phonemes = 8.9 − 24.9 (log GD) + 14.19 (NTID)

Within this analysis, gap detection accounted for one third of the variance and speech intelligibility accounted two thirds of the variance. These figures are less than those demonstrated in the individual regression analyses (34.8% for GD and 46.5% for NTID) as some of the variance is shared between gap detection and speech intelligibility due to the observed small degree of correlation between the two factors (r=-0.382, p=0.118).

Figure 14 shows the relationship between the actual CVC phoneme score and the predicted scores with a 90% prediction interval, when using the above regression
equation for this cohort of 18 participants. When a CVC phoneme score less than 30% is predicted, this cohort of recipients suggests that a phoneme score of 30% or less will be measured. More variability is observed with higher speech perception results, with a higher predicted score (greater than 30%) showing an actual phoneme score between 30 and 100%. It is evident that a significant, and strong correlation exists (r=0.769, p<0.001) suggesting that the above regression equation represents a reasonable assumption of performance based on the 500 Hz gap detection threshold and speech intelligibility of an individual with significant early onset hearing loss.

Figure 14: Relationship between the ‘predicted’ CVC phoneme score given the above regression equation, and the ‘measured’ CVC phoneme score.

An interval plot was used to investigate the relationship between speech intelligibility and the 500Hz gap detection threshold in the ear to be implanted. Participants with a speech intelligibility of two or three were combined in order to make a valid comparison. The figure shows the mean gap detection threshold (and 95% confidence
interval) for each level of speech intelligibility. Better speech intelligibility was associated with a smaller gap detection threshold (rho = -0.493, p=0.044).

![Graph showing relationship between gap detection and speech intelligibility](image)

*Individual standard deviations are used to calculate the intervals.*

**Figure 15: Relationship between gap detection and speech intelligibility for the 18 AEO-HL study cohort.**

No correlation between the BB gap detection threshold and speech intelligibility was observed for either the ear to be implanted (rho = 0.134, p=0.648) or the contralateral ear (rho = 0.418, p=0.155).

There was no significant difference seen between the 500Hz gap detection threshold of the AEO-HL cochlear implant recipients who used oral language, and those who used signing support in either the ear to be implanted (W=65, p=0.135) or the contralateral ear (W=64, p=0.469). No significant difference was seen between the BB gap detection threshold in the ear to be implanted for the oral versus signing support group (W=45, p= 0.339) or between the two groups for the BB gap detection threshold in the contralateral ear (W=39, p= 0.052).
9.2.4 Adults with an Acquired Hearing Loss: Gap Detection Results

No correlation was observed between the 500Hz gap detection threshold in the ear to be implanted and post-operative speech perception for the study group with an acquired hearing loss (rho=0.113, p=0.645). No gap detection thresholds were obtained from the contralateral ear in this group. Fourteen of the 19 participants, or 74%, achieved a gap detection threshold less than 10ms.

Figure 16: Relationship between post-operative CVC phoneme score and 500Hz gap detection threshold for the PostL1 study group.

A significant difference was observed between the AEO-HL study group, and the group with acquired hearing loss for the 500Hz gap detection threshold in the ear to be implanted (W=437.5, p= 0.0038). Far greater variability in results was observed for the AEO-HL group, as can be seen in figure 17. All 19 participants with an acquired hearing loss had the full 30dB SL available to them, while four of the AEO-HL group had a reduced SL available. Removal of these four participants from the analysis did not influence the results. A corresponding difference in speech perception was observed.
between the AEO-HL group (mean 33.8%) and the postlingual group (mean 74.7%), \( t = -6.10, p = 0.000 \).

**Figure 17**: 500Hz gap detection threshold in the ear to be implanted for the 18 AEO-HL participants and the 19 participants with an acquired hearing loss.

### 9.3 Gap Detection Discussion

This section of the project investigated the temporal processing skills of a study cohort of 18 AEO-HL as measured with a gap detection task. A significant correlation between the 500Hz gap detection threshold in the ear to be implanted and speech perception was observed, with those recipients with smaller gap detection thresholds achieving better speech perception results. Gap detection and speech intelligibility accounted for 59% of the variance in post-operative performance for AEO-HL. Gap detection abilities for a group of 19 adults with an acquired hearing loss were significantly better and demonstrated less variability than the results obtained from the study cohort of AEO-HL.
Temporal processing skills have been widely assessed in the literature utilising gap detection procedures. There exists a large range of reported thresholds which reflects differences in procedures use; for example gap detection thresholds measured acoustically have proven to be sensitive to stimulus type, stimulus intensity and stimulus frequency, with differing threshold ranges reported for each. It is generally accepted that typical gap detection thresholds for normal hearing listeners are in the range of 2-10ms, for listeners with acquired SNHL or a CI around 10ms, and around 40ms for CI users with an early onset hearing loss (Giannella Samelli & Schochat, 2008; Moore & Glasberg, 1988; Shannon, 1989; Wei et al., 2007). In the current study, the average 500Hz gap detection thresholds obtained for the group with acquired hearing loss of 8.76ms, sits well within the reported literature averages. Little research appears to have been done with AEO-HL participants pre-operatively, but the average results in the current study of 27.5ms was smaller than the average published figure of 40ms for cochlear implant recipients with an early-onset hearing loss. Figure 17 demonstrates the greater amount of variability in thresholds observed in the AEO-HL group when compared to the PostL1 group. Despite equivalent sensation levels, the ability of the AEO-HL participants to accurately detect temporal gaps in a stimulus showed wide variation. It has been suggested that variability in gap detection results may be the result of differences in the degree of survival of auditory neurons at the cochlea level (Moore & Glasberg, 1988). It is possible that those participants with larger gap detection thresholds may have had a more profound loss from early infancy, and consequently had less auditory neurons surviving and greater disruption to the development of their auditory pathway, leading to reduced temporal resolution.

Larger gap detection thresholds were measured when a broad band stimulus was used, with an average of 44.4ms recorded for the AEO-HL group. These findings are consistent with those of Moore and colleagues, who found that gap detection thresholds measured with sinusoid stimuli were smaller than those measured with bandpass noise for subjects with normal hearing, hearing loss, and cochlear implant recipients. They suggested that this may be due to fluctuations in the noise which may be confused with the gap, and therefore felt sinusoid stimuli may give a better
measure of gap detection (Moore, 1993). All participants in the current study reported that the BB signal sounded softer than the 500Hz sinusoid. This may have been due to the configuration of their hearing loss. Many participants only had a corner of low frequency acoustic hearing prior to cochlear implantation, which meant they could only detect a portion of the BB signal.

It can be difficult to predict speech perception based purely on psychophysical tests, such as gap detection. Gap detection measures the response of the auditory system to an incoming signal, whereas speech perception also reflects linguistic knowledge and top-down processing which can allow distorted incoming signals to be processed at higher levels (Kishon-Rabin et al., 2009). The influence of linguistic knowledge, and the sensitivity of gap detection thresholds to stimulus intensity and frequency, may account for some of the variation in observed correlations between speech perception and gap detection reported in the literature. The choice of speech perception material may also prove influential in the study of gap detection with adults with an early onset hearing loss, who present with language delays. Kishon-Rabin and colleagues showed that subjects with acoustic gap detection thresholds greater than 40ms performed poorly on speech perception testing (Kishon-Rabin et al., 2009), and were also able to demonstrate that those listeners with poor temporal resolution also had degraded frequency resolution. In support of this study, Tyler found significant correlations between gap thresholds obtained with noise bursts and speech perception in noise (Tyler et al., 1982).

Gap detection results for the AEO-HL group in the current study, showed significant correlation with regression analysis between the 500Hz gap detection threshold in the ear to be implanted, and the post-operative phoneme score, accounting for 34.8% of the variance in speech perception results. A significant difference in phoneme scores was observed between those participants with a gap detection threshold less than 10ms, and those with a gap detection threshold greater than 10ms. Results suggest that for an AEO-HL, a gap detection threshold less than 10ms is required to achieve a CVC phoneme score greater than 50%. This is not unexpected as the ability to perceive
the rapid temporal changes in a speech envelope is a requirement for speech understanding. These results support the majority of published data which suggest a mean gap detection threshold of 8-10 ms for listeners with an acquired hearing loss, who on average, achieve phoneme scores greater than 50%. The observed trend for differences in speech perception for those AEO-HL participants who scored below or above 10 ms does not hold for the group of adults with an acquired hearing loss, however, some of whom achieved significant speech perception results with larger gap detection thresholds. The current study did not observe any significant correlation between gap detection thresholds and speech perception results for the PostL1 study group, but this may be due to the fact that 74% of the participants with an acquired hearing loss all achieved a gap detection threshold less than 10 ms. Little variability in gap detection thresholds was observed within this group, suggesting that the majority were able to complete the task with little difficulty. This is consistent with the results of Shannon, who also found no correlation between gap detection and speech perception for 24 implant recipients with an acquired hearing loss (Shannon, 1989). Although it is difficult to reach definitive conclusions with small study numbers, results imply that in order for an AEO-HL cochlear implant recipient to achieve similar speech perception results to recipients with an acquired hearing loss, they must also be able to achieve temporal processing thresholds in the same vicinity.

The literature has shown that at an equivalent sensation level, adults with an acquired hearing loss can demonstrate gap detection thresholds in the same range as adults with normal hearing. This would suggest that temporal resolution as measured by the ability to detect gaps, is not impaired by cochlear damage (Moore, 1993; Shannon, 1989). These results may imply that it is not the presence of a cochlear hearing loss, but rather the implications of an early-onset hearing loss, which result in poorer gap detection abilities for AEO-HL. It has been shown that gap detection abilities in infants is nearly adult-like by 6-12 months of age, and that auditory deprivation up to this time may result in impaired processing skills (Trehub et al., 1995). On average, the AEO-HL group required the duration of the gap to be over three times greater than that of the PostL1 group, to be able to reliably detect it, despite equivalent sensation levels. These
differences may reflect differences in the development of the auditory pathways of the
two groups of participants as a consequence of the significant early-onset hearing loss.
These results are supported by two studies by Busby and colleagues. In a study
conducted in 1992, psychophysical abilities were investigated in a group of 10 cochlear
implant recipients who all had an onset of hearing loss prior to age four and received a
cochlear implant between the ages of five and 23. It was found that those recipients
who received their cochlear implants at a later age had longer gap detection
thresholds (Busby et al., 1992). A follow up study investigated gap detection abilities in
a group of 15 early deafened cochlear implant recipients. Results showed that
recipients who became deaf at a later age, in comparison to those with congenital
deafness, had lower gap detection thresholds. A significant correlation was also
observed between gap detection thresholds and the perception of sentences in the
auditory-visual condition. The authors commented that ‘findings suggest that the lack
of both normal hearing at birth and auditory stimulation in early childhood, as was the
case for the congenitally deaf subjects, increases the likelihood of poorer gap-
detection thresholds’ (Busby & Clark, 1999).

The development of intelligible speech requires a robust and meaningful auditory
signal at a young age, and has been shown to correlate with post-operative
performance with a cochlear implant. The lack of significant correlation in the present
study between speech intelligibility and gap detection thresholds may suggest that the
two tests reflect different aspects of the participant’s auditory development. Speech
intelligibility reflects higher level language development, and more specifically cortical
links between auditory, language and speech centres (Kral et al., 2016; Sharma et al.,
2009), and gap detection reflects auditory psychophysical abilities (Keith, 2000; Moore,
1993). Multiple regression analysis showed that gap detection and speech
intelligibility accounted for 59.22% of the variance in post-operative phoneme score
for this subset of 18 participants. Speech intelligibility accounted for approximately 2/3
of this variance, and gap detection the remaining 1/3. The correlation equation
performed well, with a significant correlation between the actual and predicted
phoneme scores (r=0.769, p<0.001). Results would suggest that the use of gap
detection thresholds, in combination with a rating of an individual's speech intelligibility, has significant predictive value for post-operative speech perception performance.

An early study by Tong, Busby and Clark demonstrated that the gap detection performance of three subjects with early onset hearing loss was significantly poorer than for subjects with an acquired hearing loss. Although subject numbers in this study were extremely small, it was also found that of the subjects with early onset hearing loss, the two who utilised total communication performed significantly worse than the subject who used oral communication. The authors suggested that early intervention would increase the chances of a more normal development of the auditory pathway during maturation (Tong et al., 1988), and that the importance of auditory experience and training was also demonstrated by the better psychophysical and speech perception performance of the oral participant. In the current study, gap detection abilities were also found to be better in oral participants than those who used signing support. The differences in gap detection thresholds between communication modes almost reached significance for both the BB stimuli results in the contralateral ear and for the 500Hz signal in the ear to be implanted. Once again it is difficult to reach definitive conclusions when study numbers are small. In this case seven participants used oral communication and eight used signing support, but the differences observed suggest that the processing of temporal information may be better in individuals who rely on audition for communication.

Although significant correlations were observed within this chapter, it is important to bear in mind the existing limitations in the methodology. It is difficult to draw definitive conclusions when working with small subject numbers, such as the 18 AEO-HL and 19 adults with an acquired hearing loss, although the current study did have more participants than many of the published studies. Four of the 18 AEO-HL did not have the full 30dB SL available to them, which may have affected results. There were also a proportion of participants who were unable to meaningfully complete the task, despite sufficient SL, which resulted in a grouping of participants with a result of 60ms
gap detection threshold. The group of 19 adults with an acquired hearing loss in group PostL1 were unlikely to represent the typical cochlear implant recipient as they were motivated recipients who had volunteered for a time-intensive study.

9.4 Gap Detection Conclusions

In the current study, acoustic 500Hz gap detection thresholds were significantly correlated with speech perception performance for AEO-HL, and were shown to be significantly poorer and display more variation than those measured in adults with an acquired hearing loss. It was observed that those participants with lower gap detection thresholds had better speech intelligibility, used oral language and achieved better speech perception results. Gap detection thresholds and speech intelligibility accounted for 59% of the variance in speech perception results within a multiple regression analysis for this study group.

Results from the current study support the following hypotheses:

Hypothesis 4: That temporal processing, as measured with acoustic stimuli pre-operatively using a gap detection test, will be poorer in AEO-HL than in adults with an acquired hearing loss

And Hypothesis 5: That the relationship between gap detection and speech perception is such that shorter gap detection thresholds are associated with better speech perception scores

It is possible that those AEO-HL who are able to achieve gap detection thresholds in the same range as adults with an acquired hearing loss may have an auditory system which is better developed, perhaps as a consequence of greater access to audition during infancy. Temporal processing skills, in combination with speech intelligibility, appears to have some predictive value for post-operative performance for AEO-HL.
10 CORTICAL AUDITORY EVOKED POTENTIALS

As results from the previous chapters have shown, AEO-HL tend to achieve lower scores on speech perception testing with cochlear implants, and display greater variability in performance, than the results typically measured from adults with an acquired hearing loss. For the current study cohort of 29 AEO-HL, speech intelligibility, communication mode and gap detection abilities have been shown to hold some predictive value for post-operative speech perception performance. The production of intelligible speech and the development of gap detection skills have the best chance of occurring in the presence of auditory input during early childhood (Santarelli et al., 2008; Teoh et al., 2004; Trehub et al., 1995). These results would suggest that it is the presence and utilisation of early audition which leads to greater speech intelligibility, better gap detection abilities and higher speech perception scores for this patient cohort.

The literature has shown that early auditory deprivation leads to the delayed maturation of CAEP in children, and that adult cochlear implant users with poor performance may display abnormal cortical responses (Mcneill et al., 2009; Psarros et al., 2009; Sharma & Dorman, 2006; Sharma et al., 2002a). For these reasons, the presence of acoustically evoked CAEP responses were investigated, and the latencies of waveform peaks in post-operative evoked CAEP were analysed over time and correlated with speech perception performance. It was considered possible that the CAEP responses of this patient cohort may provide more insight into the availability and utilisation of residual hearing during early childhood.

Specifically, the following hypotheses were tested:

Hypothesis 6: That those participants showing a measurable cortical waveform pre-operatively will achieve better speech perception scores than those patients who did not have a measurable cortical response.
Hypothesis 7: AEO-HL will demonstrate waveform latencies for auditory cortical responses which are delayed when compared to published norms for adults with normal hearing.

10.1 CAEP Methods

Cortical auditory evoked potentials were obtained for each subject pre-operatively, and three, 12 and 24 months post-operatively, using a Medelec Synergy evoked potential system.

Pre-operatively, an acoustic 500Hz tone burst signal of 50ms duration was used, presented via Synergy tubephones to each ear. The rise/fall time of the stimulus was 10ms, with a 30ms plateau as these stimulus parameters have been shown to produce the greatest N1-P2 amplitude (Onishi & Davis, 1968). The maximum stimulation level available was 115dB SPL. Due to the severity of hearing loss in the participants involved in this study, it was not always possible to attain a sufficient sensation level to record a response. The minimum sensation level required to record a cortical response in normal hearing listeners is 10dB (Eddins & Peterson, 1999). Testing of each ear was still attempted if the 500Hz threshold in that ear was less than 115dB. The stimulation level was generally increased to the maximum of 115dB, and for the majority of subjects in the study, 115dB was classified as ‘comfortable’ using the same Loudness Scale as used in the gap detection task pre-operatively, see Appendix B (Hawkins et al., 1987). A run was also completed at a level of 10dB, to get a baseline of any noise that may have been present.

The tone burst stimuli were presented at a rate of 0.5Hz, with an alternating polarity. A recording window of 500ms was used and the data was low-pass filtered at 30Hz. Artefact rejection was set at 100μv. Thirty accepted sweeps were averaged, and repeated to confirm the response for each stimulus. Although many researchers average a few hundred sweeps, Crowley and Colrain (2004) as cited by Hall (Hall III et al., 2007) reported decreased N1 amplitudes for signals within a train of stimuli, indicating short term habituation, and decreased amplitude from train to train,
indicating long term habituation. Gordon has successfully used just 50 sweeps for research purposes (Gordon et al., 2005; Gordon et al., 2008) and therefore the use of 30 sweeps has been deemed sufficient and will minimise the time spent on the task enabling it to be of use in a busy clinical setting.

Post-operatively, cortical responses were measured by stimulating individual electrodes directly. Three different electrodes were measured, typically electrode 20, 11 and three, representing the apical, mid, and basal portions of the electrode array.

Cochlear’s Custom Sound EP software was used to generate the stimuli. A monopolar stimulus mode was used. The stimuli contained 30 pulses per burst, at a rate of 250Hz with a duration of 116ms. The repetition rate, or the interval between the onset of the first stimulus burst and the onset of the next burst, was 0.5Hz and the total number of sweeps recorded was 30. Responses were recorded using gold cup electrodes positioned in a three-electrode montage. The non-inverting electrode was placed on the Vertex (Cz), the inverting electrode was placed on the contralateral mastoid, and the reference electrode on the ipsilateral mastoid. The vertex to contralateral ear montage minimises electrical artefact generated by the cochlear implant, and maximises the amplitude of the recorded response (Sharma et al., 2002a). The RF (Radio Frequency) free period was set at 500ms to ensure that there was no RF sent from the processor coil to the implant during the time window of the recording. RF has the potential to introduce interference in the recording. Despite the above protocols, artefact was observed randomly within some test sessions. The appearance of artefact did not appear to be patient dependent, as for certain participants it was observed at some test sessions and not others. For one bilateral recipient, it was observed on one ear, but not the other. At the suggestion of Cochlear Europe Limited’s Clinical Technical Support Manager, if this artefact was observed, the RF period was actually turned off. It is believed that it may be the offset and onset of the RF burst that causes the baseline deviation, or artefact, for some CI recipients and not the actual RF. This method of dealing with artefact proved successful for five participants, but there were
still seven test sessions, involving five different participants, which generated artefact prior to the implementation of Cochlear’s suggestion to turn off the RF free period.

Stimuli were presented at the Loudest Acceptable Presentation Level (LAPL) for that electrode. This level was determined by increasing the stimulation level on the electrode to be tested to the point that was the loudest level the subject could tolerate. Typically, this level exceeded the subject’s map C level. LAPL was chosen rather than the C level as a greater current level is more likely to elicit a response. Two measurements were obtained at each subject’s LAPL for each electrode to look for repeatability of morphology and latency of the N1-P2 complex, and at 10CL lower to observe a decrease in response amplitude. A measurement was also obtained at 10CL to observe any background EEG noise levels.

Waveforms were analysed by two audiologists experienced in interpreting electrophysiological responses. Both audiologists made a decision as to whether a waveform was present or not based on repeatability, morphology and latencies. If a discrepancy existed between the two judges as to whether a waveform was present or absent, a conservative approach was taken and the response was deemed to be absent. Latencies for the P1, N1 and P2 peaks were marked independently by both audiologists. An average was calculated for the latency of each peak. The maximum discrepancy in latencies between the two audiologists was 37ms, which was deemed to be acceptable.

Given that the primary question was whether or not a cortical response pre-operatively is a predictor of post-operative performance, results were classified by the presence, or absence, of a repeatable waveform. Poor waveform morphology, due to insufficient sensation levels and long-standing early-onset hearing losses, meant that analysis of latency was not always possible. The Mann Whitney test was used to determine if there were differences in post-operative speech perception between those participants who had a measurable CAEP and those who did not. Comparisons were also made between speech intelligibility and temporal processing abilities of the two groups.
Post-operatively, the latencies of P1, N1 and P2 were investigated over time for the whole study group, and the six participants who attended and had measurable responses at all three test sessions. A General Linear Model was used to determine if the latencies of the above-mentioned waveforms changed over time. Latencies were then correlated with post-operative speech perception. Latencies were chosen for analysis rather than amplitude as they have been shown to generally display less variability (Eggermont, 1988).

10.2 CAEP Results

10.2.1 Pre-Operative CAEP Results

Acoustically evoked CAEP measurements showed wide variability in responses. An attempt was made to record a CAEP for 17 participants, seven of whom had a measurable response. For the remaining nine participants, repeatable responses were unable to be elicited. The range of sensation level available to the group who had measurable CERAs was 15-65 dB, and for the group without measurable responses it was 5-35dB.

Figure 18 shows an example of an acoustic CAEP, recorded from one of the participants in the current study. Latencies and morphology follow the typical patterns, with P1 = 60ms, N1=95ms, P2 = 190ms.

![Figure 18: Example of a pre-operative acoustic CAEP recorded in the current study.](image-url)
Figure 19 and 20 show the pre-operative acoustic CAEP and the three-month post-operative CAEP for another participant. Although this participant had a 500Hz pure tone threshold of 85dB, a response was unable to be recorded acoustically at the maximum stimulation level of 115dB. After three months of cochlear implant use, however, a repeatable electrically evoked CAEP was able to be obtained with P1, N1 and P2 occurring at expected latencies.

![Figure 19: Pre-Operative acoustic CAEP for Participant 8.](image)

![Figure 20: Three-month Post-Operative CAEP for Participant 8.](image)

The Mann-Whitney test was used to determine if those participants with measurable pre-operative CAEP responses obtained greater post-operative speech perception than those without measurable responses. There was no significant difference in speech perception between the two groups (W=51.1, p= 0.4269).
There was no significant difference in speech intelligibility between those participants with, and without a measurable CAEP response ($W=55$, $p=0.6575$). There was no significant difference in gap detection abilities between those with, and without a measurable CAEP ($W=44$, $p=0.68$).

There was no significant difference in the pre-operative 500Hz threshold between those participants with, and without a measurable CAEP ($W=45$, $p=0.1302$) or the 1KHz threshold between the two groups ($W=43.5$, $p=0.1588$).

The above calculations were repeated with the one participant with only a 5dB SL removed, but there was no change to the results. There remained no significant difference between the two groups for any of the factors investigated.

The presence of a pre-operative CAEP response did not appear to be predictive of post-operative outcome, or to be correlated with speech intelligibility or gap detection thresholds.

### 10.2.2 Post-Operative CAEP Results

Post-operatively, 17 participants had a measurable electrically evoked CAEP (eCAEP) on at least one electrode at the three-month assessment point. One participant had persistent artefact and one had no response on any electrode. At the 12-month assessment point, 11 participants had a measurable CAEP on at least one electrode. Three participants had persistent artefact, and three had no repeatable responses. Two of these three participants had a LAPL of just 4-6CL above their map C (comfort) level, therefore it is possible that this was not a sufficient sensation level to generate a response. The remaining participant with no repeatable response had a SL of 15-30CL.

At the 24-month assessment point, 11 patients had a measurable CAEP on at least one electrode and three had pure artefact.

Figure 21 represents an example of the type of artefact that was seen during this project. The size of the artefact was not dependent on the stimulus level, it was
equally as large with an inaudible stimulus level of just 10 CL, making it easy to identify, and supporting the suggestion that this artefact was a result of the RF onset and offset pulse. Persistent artefact was observed for seven test sessions prior to discussions with Cochlear Ltd, which resulted in the suggestion of turning off the RF free period in the Custom Sound software. As can be seen in figure 21, the artefact appears to begin at around 120ms after the stimulus, which corresponds to the offset of the stimulus burst used in this study (116ms). It is likely that this artefact may be due to an RF offset baseline deviation. Removing the RF free period from the recording set-up resulted in noisier traces, but it did eliminate the artefact for all five participants who showed artefact in their standard measurements.

Figure 21: Example of artefact encountered during post-operative recordings of eCAEPs.
10.2.3 Waveform Latencies

Changes in the latency of the P1, N1 and P2 waveforms were analysed using a GLM, with subject, time and electrode as factors, and subject allocated as a random factor. The analysis was first done with all participants pooled, and secondly with the five participants who had measurable responses at all three assessment points.

For the pooled data, there was no significant change in latencies across electrode, or time, for any of the waveforms, but there was a significant difference due to participant. There was no significant difference due to time for the P1 or the N1 waveform.

Table 3: GLM for pooled data from 20 participants

<table>
<thead>
<tr>
<th>Waveform Peak</th>
<th>Factor</th>
<th>F Value</th>
<th>Significance Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>P1</td>
<td>Subject</td>
<td>7.91</td>
<td>p&lt;0.001</td>
</tr>
<tr>
<td></td>
<td>Time</td>
<td>0.65</td>
<td>p=0.528</td>
</tr>
<tr>
<td></td>
<td>Electrode</td>
<td>0.82</td>
<td>p=0.447</td>
</tr>
<tr>
<td>N1</td>
<td>Subject</td>
<td>7.27</td>
<td>p&lt;0.000</td>
</tr>
<tr>
<td></td>
<td>Time</td>
<td>0.30</td>
<td>p=0.739</td>
</tr>
<tr>
<td></td>
<td>Electrode</td>
<td>0.32</td>
<td>p=0.726</td>
</tr>
<tr>
<td>P2</td>
<td>Subject</td>
<td>6.96</td>
<td>p&lt;0.001</td>
</tr>
<tr>
<td></td>
<td>Time</td>
<td>0.19</td>
<td>p=0.825</td>
</tr>
<tr>
<td></td>
<td>Electrode</td>
<td>2.46</td>
<td>p=0.096</td>
</tr>
</tbody>
</table>

Table 4: Average peak latencies for 20 participants

<table>
<thead>
<tr>
<th>Waveform Peak</th>
<th>Time Point</th>
<th>Mean Latency</th>
<th>Standard Deviation</th>
</tr>
</thead>
<tbody>
<tr>
<td>P1</td>
<td>3 months</td>
<td>58.16ms</td>
<td>13.06</td>
</tr>
<tr>
<td></td>
<td>12 months</td>
<td>58.62ms</td>
<td>9.22</td>
</tr>
<tr>
<td></td>
<td>24 months</td>
<td>57.12ms</td>
<td>5.36</td>
</tr>
<tr>
<td>N1</td>
<td>3 months</td>
<td>109.45ms</td>
<td>27.24</td>
</tr>
<tr>
<td></td>
<td>12 months</td>
<td>111.76ms</td>
<td>23.47</td>
</tr>
<tr>
<td></td>
<td>24 months</td>
<td>109.21ms</td>
<td>13.82</td>
</tr>
<tr>
<td>P2</td>
<td>3 months</td>
<td>183.06ms</td>
<td>22.46</td>
</tr>
<tr>
<td></td>
<td>12 months</td>
<td>184.62ms</td>
<td>22.77</td>
</tr>
<tr>
<td></td>
<td>24 months</td>
<td>191.42ms</td>
<td>21.70</td>
</tr>
</tbody>
</table>
As there was no significant difference in peak latencies across time or across electrode, all latencies were combined and averaged. The average P1 peak latency for 77 recordings, was 58.05ms, the N1 from 78 recordings was 110.01ms, and for 75 recordings of P2 the latency was 185.61ms. These latencies sit well within the published norms for normal hearing adults, of 40-60ms for P1, 80-120ms for N1 and 150-200ms for P2 (Beagley, 1971). GLM analysis for the five participants with measurements at all three time points did not show a significant effect of subject, time or electrode.

Table 5: GLM analysis for the five participants with results at all three time points.

<table>
<thead>
<tr>
<th>Waveform Peak</th>
<th>Factor</th>
<th>F Value</th>
<th>Significance Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>P1</td>
<td>Subject</td>
<td>2.03</td>
<td>p=0.118</td>
</tr>
<tr>
<td></td>
<td>Time</td>
<td>0.88</td>
<td>p=0.429</td>
</tr>
<tr>
<td></td>
<td>Electrode</td>
<td>0.44</td>
<td>p=0.652</td>
</tr>
<tr>
<td>N1</td>
<td>Subject</td>
<td>0.81</td>
<td>P=0.553</td>
</tr>
<tr>
<td></td>
<td>Time</td>
<td>0.90</td>
<td>p=0.422</td>
</tr>
<tr>
<td></td>
<td>Electrode</td>
<td>0.13</td>
<td>p=0.880</td>
</tr>
<tr>
<td>P2</td>
<td>Subject</td>
<td>0.72</td>
<td>P=0.618</td>
</tr>
<tr>
<td></td>
<td>Time</td>
<td>1.27</td>
<td>p=0.301</td>
</tr>
<tr>
<td></td>
<td>Electrode</td>
<td>0.51</td>
<td>p=0.610</td>
</tr>
</tbody>
</table>

Table 6: Average latencies across time points for the different waveform peaks, for the five participants with results at all three time points.

<table>
<thead>
<tr>
<th>Waveform Peak</th>
<th>Time Point</th>
<th>Mean Latency</th>
<th>Standard Deviation</th>
</tr>
</thead>
<tbody>
<tr>
<td>P1</td>
<td>3 months</td>
<td>55.08ms</td>
<td>7.89</td>
</tr>
<tr>
<td></td>
<td>12 months</td>
<td>55.58ms</td>
<td>3.58</td>
</tr>
<tr>
<td></td>
<td>24 months</td>
<td>57.44ms</td>
<td>4.10</td>
</tr>
<tr>
<td>N1</td>
<td>3 months</td>
<td>103.86ms</td>
<td>5.01</td>
</tr>
<tr>
<td></td>
<td>12 months</td>
<td>104.75ms</td>
<td>8.96</td>
</tr>
<tr>
<td></td>
<td>24 months</td>
<td>107.78ms</td>
<td>5.47</td>
</tr>
<tr>
<td>P2</td>
<td>3 months</td>
<td>177.29ms</td>
<td>13.65</td>
</tr>
<tr>
<td></td>
<td>12 months</td>
<td>173.58ms</td>
<td>12.28</td>
</tr>
<tr>
<td></td>
<td>24 months</td>
<td>184.00ms</td>
<td>11.31</td>
</tr>
</tbody>
</table>

As there was no significant difference in peak latencies across time or across electrode for these 5 participants, all latencies were combined and averaged. The average P1
peak latency for 34 recordings, was 55.88ms, the N1 from 35 recordings was 105.17ms, and for 35 recordings of P2 the latency was 177.74ms. Again, these latencies sit well within published norms (Beagley, 1971).

10.2.4 Correlations with Speech Perception

As there were no significant differences in latencies seen across electrodes or assessment points for any participant, an average latency for each waveform was calculated for each participant. This overall latency was then correlated with post-operative CVC phoneme score. No significant correlations were observed between latencies and speech perception results for any of the peaks (P1: r=0.305 p=0.219, N1: r=0.312, p=0.207, P2: r=0.233, p=0.352). An example of the lack of significant relationship between the N1 latency and CVC phoneme score for the 17 participants can be seen in figure 22.

![Figure 22: Relationship between N1 latency and post-operative phoneme score for 17 participants.](image-url)
Peak latencies were not correlated with speech intelligibility (P1: r=-0.386 p=0.126, N1: r=0.031 p=0.906, P2: r=-0.040 p=0.880) or with the 500Hz gap detection threshold in the implanted ear (P1: r=0.087 p=0.798, N1: r=-0.475 p=0.139, P2: r=-0.334, p=0.316).

10.3 CAEP Discussion

Results from the current study show that there was no difference in post-operative speech perception scores between the group of seven participants who had a measurable CAEP pre-operatively, and the nine participants who did not. There was also no difference in speech intelligibility, gap detection thresholds or the acoustic 500Hz and 1KHz threshold. The presence of a pre-operative CAEP response did not appear to be predictive of post-operative outcome, or to be correlated with speech intelligibility or gap detection thresholds. Acoustic auditory cortical responses are not widely used as a clinical test with AEO-HL due to the difficulty in recording (Burdo et al., 2006). This was certainly evident in the current study, where a difference in pre-operative 500Hz and 1KHz threshold was not observed between participants with a measurable response, and those without. A recent case study of two AEO-HL by Abraham and colleagues (2015) showed that acoustically evoked CAEP responses were successfully recorded via hearing aid amplification at sensation levels of 10 and 30dB. The two adults in this case study had very early-onset hearing loss, believed to be congenital, derived no perceived benefit from amplification, and had developed no oral language. With such a small sample size it is not possible to make any conclusions, but the authors have suggested that the successful recording of responses may reflect two very different scenarios. The first possibility may be that the recorded CAEP responses reflect cortical decoupling resulting in the innervation of the primary auditory cortex (as well as the secondary cortex) by neighbouring mature structures, allowing the N1 to be recorded, or in the opposite vein, the responses may reflect the maturation of cortical neurons even when there is a lack of auditory stimulation (Abraham et al., 2015). While the results of the current study seem to support the claims made by Abraham and colleagues, that it is indeed possible to measure a typical CAEP result from AEO-HL (Abraham et al., 2015), the lack of a response does not
appear to correlate with any pre-operative patient demographic, or post-operative speech perception performance. As noted by Burdo and colleagues (2006), the successful measurement of acoustic CAEP waveforms from this patient population remains problematic, and may not hold much clinical significance (Burdo et al., 2006).

Post-operatively, electrically evoked CAEP were able to be recorded in 17/19 participants at the 3-month point, 11/17 at the 12-month point, and 11/14 at the 24-month point. Electrical artefact, assumed to be due to the offset of the stimulus burst causing a baseline deviation, prevented a result being measured in eight participants. Only four recipients demonstrated no measurable response. Utilising the recording montage suggested by Sharma and colleagues (2002) ensured that responses were successfully recorded for the greater majority of participants (Sharma et al., 2002b).

Unfortunately, the data for this section of the project was incomplete. Technical issues involving the presence of artefact caused some recordings to be unusable. It was also difficult to ensure that all participants attended all post-operative sessions. Generally, participants were very willing at the 3-month assessment point, as they were all still very much involved in the early programming sessions at the RVEEH and enthusiastic about their progress. By 12 and 24-months post-surgery, however, enthusiasm seemed to wane, and it became more difficult to encourage participants to attend extra research appointments.

Analysis of the latencies of the P1, N1 and P2 waveform peaks in the current study showed a significant difference between participants, but not across electrodes, or across time. A recent study by Lammers and colleagues (2015) which also recorded evoked CAEPs via direct stimulation, also showed no difference in latencies between basal, mid and apical electrodes (Lammers, Versnel, et al., 2015). The lack of significant difference in latencies across time and electrode in the current study meant that averages for the different waveform peaks could confidently be calculated. The group average of 58ms for P1, 110ms for N1 and 186ms for P2 sits well within the published norms of 40-60ms for P1, 80-120ms for N1, and 150-120ms for P2 (Beagley, 1971). In contrast to the results from this study, changes in latencies post-operatively have been
observed both in late-implanted children and AEO-HL who receive an implant as an adult (Burdo et al., 2006; Sharma et al., 2002a). For both studies, bigger changes in latencies were observed for those recipients with longer durations of deafness. For children implanted after the age of seven, a rapid decrease in latencies was observed in the first month, but then the latencies stabilised, or only changed very slightly after that point and never reached the normal limits (Sharma et al., 2002a). Burdo has suggested that these larger decreases seen with AEO-HL with no auditory memory, confirm the immaturity of auditory cortical function in congenital deaf subjects without any hearing experience (Burdo et al., 2006).

In the current study, no correlation was observed between waveform latencies and speech perception performance. There is a contradiction within the literature regarding the correlation of CAEP responses with speech perception performances. Some studies have shown a distinct correlation with latencies or morphology (Gordon et al., 2005; Kelly et al., 2005; Mcneill et al., 2009; Psarros et al., 2009), whereas others have shown typical waveforms irrespective of speech perception performance (Gordon et al., 2008; Lammers, Versnel, et al., 2015). A study by Kelly and colleagues (2005) showed a significant correlation between speech perception and the latency of P2 for a group of 12 adult CI recipients with an acquired hearing loss, with earlier latencies associated with greater speech perception scores (Kelly et al., 2005). A second study has shown that some adult recipients who had a less than optimal result with their cochlear implant, also had an abnormal cortical result, categorised as a response that could not be repeated, or did not occur within an appropriate time window (Psarros et al., 2009). A case study of one AEO-HL cochlear implant recipient demonstrated evoked CAEPs with an immature morphology, similar to that recorded in young infants, consisting of a broad, large amplitude positive peak (Mcneill et al., 2009). Atypical cortical responses were also found in children with cochlear implants who achieved only limited speech perception with their device (Gordon et al., 2005). Results showed that children implanted after age four showed a range of waveform morphologies, whilst those implanted at a younger age showed more typical morphologies. It has been suggested that the association between poor speech perception results and
atypical CAEP responses, for both adults and children, may reflect central processing issues, a persistent immaturity in the cortex, or aberrant cortical organisation (Gordon et al., 2005; Psarros et al., 2009). Results from the above-mentioned studies suggest that variations in the ‘typical’ P1-N1-P2 evoked potential response can be observed in both adult and paediatric cochlear implant recipients. These variations include delays in the expected latencies of the P1 and N1 waveform, absence of the N1 waveform, and atypical morphologies. These variations have been shown to be moderately correlated with age at implant for paediatric cases, and with speech perception performance for both adult and paediatric recipients (Gordon et al., 2005; Mcneill et al., 2009; Psarros et al., 2009). The variability observed in both latencies and morphology is likely to be a result of cortical reorganization stimulated by auditory deprivation (Dorman et al., 2007).

In contrast to the above studies demonstrating aberrant CAEP responses obtained by AEO-HL cochlear implant recipients, there exists a selection of studies which have shown somewhat ‘typical’ responses from this patient subgroup. A case study of two AEO-HL who underwent acoustic CAEP testing via high-powered hearing aids, showed that at sensation levels of 30dB and 10dB, typical P1-N1-P2 waveforms were successfully recorded (Abraham et al., 2015). Lammers and colleagues (2015) investigated CAEPs in adult cochlear implant recipients, 12 of whom became deaf prior to age two (as far as could be determined) but who received their implant as an adult. Results showed that a typical N1-P2 waveform could be recorded irrespective of speech perception score and onset of deafness. The AEO-HL group however, did show shorter N1 latencies with larger amplitudes than the group of adults with an acquired hearing loss (Lammers, Versnel, et al., 2015). Similar results were obtained by Gordon and colleagues (2008) when investigating CAEP results from a group of 16 children. The children in the study who scored less than 50% on speech perception testing showed a significantly shorter N1 latency than those who scored over 50% (80ms vs 100ms) (Gordon et al., 2008). It is generally acknowledged that the deprivation of sound prevents the appropriate maturation and development of the auditory system, which can be observed through atypical CAEP results (Dorman et al., 2007; Eggermont &
Ponton, 2003; Sharma et al., 2002b; Sharma et al., 2007). It is therefore interesting that ‘typical’ P1-N1-P2 waveforms have been successfully recorded in AEO-HL both pre-operatively via an acoustic stimuli (Abraham et al., 2015), post-operatively through the cochlear implant in the current study and the recent study by Lammers and colleagues (Lammers, Versnel, et al., 2015), and in implanted children with poor functional outcomes (Gordon et al., 2008). The post-operative studies of Lammers and Gordon, however, have demonstrated that the latency of the N1 is earlier for AEO-HL, and with a bigger amplitude, when compared to adults with an acquired hearing loss (Lammers, Versnel, et al., 2015), and earlier for poor performing children compared to children with good speech perception performance (Gordon et al., 2008). Gordon has suggested that the poor functional outcomes associated with ‘normal-like’ responses suggest that these peaks reflect different activity than that occurring in normal hearing children (Gordon et al., 2008). The possibility of a loss of synaptic pruning as a result of early onset hearing loss in the adult patients in Lammer’s study (Huttenlocher & Dabholkar, 1997), combined with the possibility of corticocortical de-coupling (Kral & Eggermont, 2007), may have resulted in a larger cortical area responding to a stimulus, leading to the larger and earlier N1 peak observed (Lammers, Versnel, et al., 2015).

The duration of deafness before implantation in children has been shown to have significant effects on the changes seen in post-operative CAEP recordings. Cortical development depends on extrinsic stimulation, therefore congenital deafness can disrupt the functional connectivity and development of the auditory system (Sharma et al., 2015). Sharma and colleagues (2002) investigated CAEP results in 245 congenitally deaf children implanted at different ages. The children who received their cochlear implant prior to age 3.5 all developed P1 waveforms within the normal limits within six months after receiving their cochlear implant. Those implanted between 3.5 and seven years showed variability in their latencies, and the majority of those implanted after age seven did not achieve normal latencies even after several years of experience with their cochlear implant. (Sharma et al., 2002b). The emergence of the N1 waveform appears to be dependent on auditory input during a critical period. Eggermont and Ponton (2003) found that the N1 component of the CAEP was absent for two subjects.
who were deaf for more than three years prior to receiving their implants at age six (Eggermont & Ponton, 2003). Sharma has also shown that children implanted after age seven generally do not develop an N1 response, whereas those implanted prior to age 3.5 show an N1 peak with similar morphology and latency to that found in normal hearing children (Sharma & Dorman, 2006). There were two late-implanted patients in Sharma’s study who did develop an N1 response. One of these had a progressive hearing loss and the other showed mild-moderate aided thresholds. It is likely that they both had sufficient residual hearing to provide adequate auditory stimulation to the cortex during childhood (Sharma & Dorman, 2006). Results from the above mentioned studies seem to suggest a sensitive period for central auditory development of about 3.5 years (Sharma et al., 2007) and that beyond age seven auditory plasticity is significantly reduced (Sharma et al., 2015).

It has been suggested, at least in children, that the absence of the N1 component indicates abnormal higher-level auditory cortical development (Sharma & Dorman, 2006), and that the P1 and N1 peaks will not reach typical latencies if there is an absence of auditory input during the critical period (Eggermont & Ponton, 2003; Sharma et al., 2015; Sharma et al., 2002b). The current study, however, demonstrated typical waveforms with appropriate P1 and N1 latencies for the small cohort of AEO-HL who did not receive a cochlear implant until adulthood. There are two possible explanations for these results. The first is that the patients had sufficient residual hearing during the critical period for the primary auditory cortex, and the secondary auditory cortical regions (including the supragranular layers and intra-hemispheric cortico-cortical connections) to develop along relatively normal lines (Eggermont & Ponton, 2003; Godey et al., 2001). This suggestion is supported by the fact that every participant in the current study had some oral language, suggesting they had access to at least some audition during infancy. No participant had a confirmed congenital total hearing loss.

The second explanation could be that provided by Lammers and colleagues from their study in 2015. Results showed that although the waveforms observed in AEO-HL
implant recipients demonstrated typical morphology, the N1 latency was earlier and the amplitude bigger, than those observed in adults with an acquired hearing loss. As suggested by Lammers, it is possible that the recordable CAEP may have been due to the combination of a lack of synaptic pruning (Huttenlocher & Dabholkar, 1997) and decoupling of the auditory cortex (Kral & Sharma, 2012), leading to the activation of a wider area of axons representing more innate and less complex auditory cortical networks (Lammers, Versnel, et al., 2015). In the current study, a proportion of participants with poor speech intelligibility and poor speech perception showed typical waveform morphologies with latencies within the normal range, with no correlation observed between latencies and speech intelligibility or gap detection thresholds. It is therefore possible that the waveforms observed in the current study reflect the activation of a wider area within the cortex and a lack of synaptic pruning, rather than a specific region of cortical activation.

While evoked potentials have successfully been used to assess maturity of the auditory pathway in children (Sharma et al., 2002b; Wunderlich et al., 2006), such an application may not be applicable, or appropriate, when assessing AEO-HL. Recent results, including those from the current study, evoked responses from Lammers and acoustic responses from Abraham, suggest that typical waveforms can be obtained by adults with minimal prior exposure to audition (Abraham et al., 2015; Lammers, Versnel, et al., 2015). These results may reflect a different origin of the waveform generators (Gordon et al., 2008; Lammers, Versnel, et al., 2015) or may in fact indicate that an individual may have had access to sound during infancy, allowing the auditory pathway to develop appropriately.

10.4 CAEP Conclusions

In the current study, seven of seventeen participants had a measurable acoustic CAEP response. The presence or absence of a response was not correlated with speech perception, speech intelligibility or gap detection abilities. Post-operatively, the majority of participants had a measurable eCAEP response. Waveform peak latencies
for this cohort of AEO-HL were well within published norms, did not change with implant experience, and were not correlated with speech perception performance.

Results from the current study do not support the following hypotheses:

Hypothesis 6: That those participants showing a measurable cortical waveform pre-operatively will achieve better speech perception scores post-operatively than those patients who did not have a measurable cortical response.

Hypothesis 7: That AEO-HL will demonstrate waveform peak latencies for auditory cortical responses which are delayed when compared with published norms for adults with normal hearing.

While CAEPS have been used successfully to assess the maturity of the auditory system in children, this does not appear to be appropriate for AEO-HL. Results from the current study have shown that it is possible to record CAEP with typical morphologies and latencies from AEO-HL giving limited clinical value to the measurement of CAEP in this patient population.
11 NON-VERBAL IQ

Previous chapters have demonstrated that whilst a portion of the variance in post-operative results observed in the study cohort of 29 AEO-HL can be accounted for via speech intelligibility and gap detection abilities, there remains a degree of unexplained variance. In cochlear implant recipients in general, once individual demographics have been accounted for, it has been suggested that additional observed variance may be due to differences in cognitive function (Pisoni & Cleary, 2003). For this reason, the non-verbal IQ of the study group of AEO-HL was investigated via the Raven’s Progressive Matrices test.

Specifically, the following hypothesis was tested:

Hypothesis 8: That those participants with lower scores on the Raven’s Matrices non-verbal test of IQ, will achieve lower post-operative speech perception scores than those who achieved higher scores.

11.1 Non-Verbal IQ Methods

Non Verbal IQ was measured using Raven’s Progressive Matrices (J. C. Raven, 1936) which is a widely used intelligence test designed to measure a person’s ability to reason and solve abstract problems independent of language. The test measures two components of general intelligence (Spearman, 1904); “the ability to think clearly and make sense of complex data (educative ability) and the capacity to store and reproduce information (reproductive ability) (J. C. Raven, 1936). The test attempts to remove language barriers in the estimate of intelligence, and so is particularly well suited to measuring the intelligence of people with hearing loss or reading problems.

The Standard Progressive Matrices test – Classic Version was used in this study. It was designed to be used for ages 6 – 80 years, taking approximately 20 – 45 minutes to complete. The test can be administered by an allied health professional, such as an audiologist, negating the need to employ a psychologist for the purpose of this study.
The standard set consisted of 60 problems divided into sections A, B, C, D and E, each made up of 12 problems. The participant was required to find the missing item to complete the pattern in each problem. In each set the first problem was, as nearly as possible, self-evident. The problems which follow built on the first problem and became progressively more difficult, requiring greater cognitive capacity to encode and analyse. Participants were required to complete the test independently, by marking on a test form which item would successfully complete each pattern. Results were scored by adding up the number of correct responses in each set. To ensure that the pattern of correct responses was consistent, results were compared to the normal distribution of scores across the five sets for each possible total score.

The participant’s total score was compared against norms collected from military and civilian subjects between the ages of 20 and 65, which were collected in 1992. A participant’s score was classified based on the percentage of people in the same age range who obtained higher, or lower, scores, and given a corresponding grade.

- Grade I: ‘Intellectually superior’ If the score lay at or above the 95th percentile for people of the same age group
- Grade II: ‘Definitely above the average in intellectual capacity’. If the score lay at or above the 75th percentile (designated II+ if it lay at or above the 90th percentile)
- Grade III: ‘Intellectually Average” if the score lay between the 25th and 75th percentile (designated III+ if it was above the 50th percentile, and III- if it was below)
- Grade IV: ‘Definitely below average in intellectual capacity’ if the score lay at or below the 25th percentile (designated IV- if it lay below the 10th percentile)
- Grade V: ‘Intellectually impaired’ If the score lay at or below the 5th percentile for that group.

Comparisons between the Raven’s raw scores obtained from the AEO-HL and acquired hearing loss study groups were made using two sample t tests. IQ measures are
designed to be normally distributed within the general population, therefore parametric statistics were used for this section of data.

For the AEO-HL study group, Pearson correlations were used to analyse relationships between the Raven’s raw score and speech perception, receptive language and speech intelligibility. A two-sample t test was used to analyse differences in raw score between the oral, and signing support group, and a one-way ANOVA using Tukey’s pairwise comparisons was used to investigate differences in the speech perception mean between Raven’s grades.

11.2 Results

Speech perception and Raven’s results were measured for 24 participants within the AEO-HL study group, and all four study participants with an acquired hearing loss. A two-sample t test showed there was no significant difference between the average Raven’s raw score of the AEO-HL study group (mean 45.96) and the acquired hearing loss study group (mean 49), (t=1.43, p=0.179).

11.2.1 AEO-HL Study Group

Figure 23 shows the distribution of grades observed within the AEO-HL study group. Thirty-three percent of participants achieved a Grade III score, representing ‘Intellectually Average’, and 42% received a Grade IV, representing ‘Definitely below average in intellectual capacity’. Two participants achieved a Grade of V, suggesting ‘Intellectually impaired’, and two achieved a Grade I, suggesting ‘Intellectually superior’.
Figure 23: Distribution of 24 AEO-HL participants amongst RMP Grades.

Figure 24: Average post-operative phoneme score for each RPM Grade for the 24 AEO-HL.

The differences in measured speech perception obtained by participants falling within the five Raven’s grades were investigated using a one-way ANOVA using Tukey’s
pairwise comparisons. No statistically significant difference between the means of each Grade was observed (p=0.176).

No correlation was observed between the Raven’s raw score and post-operative CVC phoneme score (r=-0.232, p=0.276) or between the Raven’s Grade and post-operative CVC phoneme score (r=0.340, p=0.104).

**Figure 25: Relationship between the post-operative phoneme score and Raven's raw score for the 24 AEO-HL.**

A significant correlation was observed between the Raven’s score and the receptive language score, as measured by the PPVT standard score, for both the Raven’s raw score (0.720, p<0.001) and Grade score (r=-0.588, p=0.003).
Figure 26: Relationship between receptive language and Raven’s raw score for the 24 AEO-HL.

No significant correlation was observed between the Raven’s raw score and speech intelligibility ($r=-0.024$, $p=0.911$).

No difference in Raven’s raw score was observed between those participants using oral communication ($n=12$, mean = 44.83), and those utilising some signing support ($n=12$, mean = 47.08) ($t=-0.70$, $p=0.494$).

### 11.2.2 Acquired Hearing Loss Study Group

Three of the participants with an acquired hearing loss achieved a Grade III and one achieved Grade IV on the Raven’s Progressive Matrices.

No correlation was observed between the Raven’s Raw Score and post-operative CVC phonemes for the acquired hearing loss participants ($r=0.162$, $p=0.838$), or between the Raven’s raw score and PPVT standard score ($r=0.134$, $p=0.866$), although it must be noted that with a sample size of just four, it is not possible to make any conclusions.
11.3 Non-Verbal IQ Discussion

Seventy-five percent of the AEO-HL study group fell within Grade III or IV on the Raven’s progressive matrices, putting them in either the ‘Intellectually average’ or ‘Definitely below average in intellectual capacity’ group. Two participants scored ‘Intellectually superior’, and two ‘intellectually impaired’. Although there was no significant difference between speech perception results between the different grades, the two participants who scored ‘Intellectually superior’ had speech perception scores of 0% and 4%, and one of these participants no longer uses their device. While it is difficult to make inferences from small participant numbers, the lack of correlation between the RPM raw score and CVC phoneme score suggested that the use of the RPM, as a measure of non-verbal intelligence, does not appear to predict post-operative performance within this cohort.

Cochlear implant outcomes in children have been shown to be influenced by the presence of any additional disability, in particular cognitive delays. Studies have shown that the presence of a cognitive delay can result in poorer speech perception performance (Dettman et al., 2004; Dowell et al., 2002; Pyman et al., 2000). A study by Pyman and colleagues (2000) demonstrated that slower progress was made with a cochlear implant if the child also had either a motor and/or cognitive delay. The severity of any existing delay was not quantified, rather labelled as present, or absent, but results suggested that the presence of such a delay may even result in the absence of any measurable speech perception (Pyman et al., 2000). The influence of the degree of severity of delay was investigated by Dettman and colleagues (2004), who found that children with a moderate and severe developmental delay had significantly poorer speech perception than children without such delay (Dettman et al., 2004). Although no such correlation was observed in the current study, using the RPM, only two participants fell within the ‘intellectually impaired’ grade. It is possible that with larger study numbers, a difference in post-operative performance may have been observed.

Non-verbal intelligence, as measured with the RPM in the current study, was not significantly different between the AEO-HL and acquired hearing loss study groups. The
RPM test was designed to remove language barriers, making it a useable test for populations with delayed language skills. No difference in score was observed between the AEO-HL participants who used oral communication and those who used signing support. The language mode engaged by this cohort of participants, whether purely oral or with the addition of some signing support, did not influence results on this test. These results support the use of the RPM as an appropriate test to assess intelligence in this cohort of participants.

It is generally acknowledged that vocabulary is linked to cognition. Various hypotheses have been suggested as to the reason for this link, such as greater vocabulary being the result of wider reading habits, leading to a greater ability to store and utilise verbal concepts (LM Dunn & Dunn, 2007). According to Spearman, general intelligence, or ‘g’ has two components, the educative component which is measured by Raven’s Progressive Matrices, and reproductive ability (Spearman, 1904). Reproductive ability is the ability to recall, and use explicit, verbalised concepts, and is measured by the Mill Hill Vocabulary Scale. The Raven’s Progressive Matrices and Mill Hill Vocabulary Scale are often used together in order to make a statement about the two main components of ‘g’, and generally the scores on both correspond. It is suggested that if a person’s general vocabulary score is below their score on the Matrices test, then it may be that the person has not been exposed to, or been able to achieve, the use of language that their intellectual capacity warrants (J. Raven, 2008).

Although results are inconclusive, some studies have proposed a ‘verbalisation hypothesis’ with regards to the approach taken by participants on a non-verbal test of intelligence (Burke & Bingham, 1969; Jensen et al., 1968). It has been suggested that participants may ‘talk’ their way through the problems on the page, and therefore an appropriate score on the Raven’s progressive matrices may in fact be somewhat reliant on adequate language skills (Burke & Bingham, 1969). Goetzinger et al (1967) however, showed no correlation between the language skills and non-verbal reasoning of deaf children (Goetzinger et al., 1967). The results of these studies seem to suggest that while it is possible that verbalisation may play a role in problem solving skills, it is
not always the case. It is conceivable that the verbalisation hypothesis may help to explain the positive correlation seen between the PPVT standard score and the Raven’s progressive matrices score in the current study. Those participants with higher vocabulary scores also tended to score better on the Raven’s Progressive Matrices test. The Mill Hill Vocabulary test of intelligence was not chosen for this study as the main objective was to measure non-verbal intelligence, but the correlation observed by Raven and colleagues appears to also apply to the PPVT test in the present study (J. Raven, 2008).

Intelligence test results are designed to be normally distributed within the general population. Although the current study has small participant numbers, the distribution approaches normality, with 75% of AEO-HL participants and all 4 postlingual participants falling within Grade III or IV. With very few participants falling within the Grades at each extreme, it was difficult to observe any significant differences in performance. It is possible that with greater participant numbers, a difference in performance may have been observed, at least between the two ends of the scale.

Research suggests that tests of working memory may be a better predictor of post-operative cochlear implant performance than non-verbal IQ. Zekveld and colleagues (2007) investigated the relationship between hearing loss and performance on non-verbal, cognitive tests of attention and memory. They found no association between hearing loss and performance on these tests, and in fact suggested that working memory was used by people with hearing loss to compensate for the degraded auditory signal. It is likely that auditory working memory plays a crucial role in speech understanding, and that the more severe the hearing loss, the more a listener may rely on working memory (Zekveld et al., 2007). In support of these results, a review by Pisoni (2000) suggested that the phonological representations of words and the mapping of sound patterns onto meanings in memory, plays a crucial role both in the perception, and the production of speech. Pisoni noted that children who performed well on speech perception tasks, often also performed well on other language measures and had better speech production skills. Pisoni suggested that this may have
been due to a better ability to recall and use words stored in the lexicon and to access sensory-motor patterns from memory to imitate spoken words (Pisoni, 2000). The specific speech perception material chosen for measuring outcomes has been shown to have an effect on the correlation with working memory in Mandarin-speaking cochlear implant users (Tao et al., 2014). Tao and colleagues found that working memory and efficiency was associated with sentence recognition.

Significant correlations between speech perception results and cognitive functioning skills have been reported by Pisoni and colleagues (2003 and 2000) for large groups of implanted children. In a study of 176 implanted children, strong correlations in performance were observed between speaking rate and both forward and backward digit span, a test which is often used to assess working memory. Results from this study suggested that up to 20% of the unexplained variance in outcome may be due to cognitive factors related to the speed and efficiency that phonological and lexical information is retrieved (Pisoni & Cleary, 2003). The assessment of the rate of speech correlates with a concept referred to as ‘verbal rehearsal’ (Landauer, 1962; Standing & Curtis, 1989). Verbal rehearsal typically refers to the concept of an individual silently repeating to themselves the verbal information to be remembered, as a method of maintaining information in their working memory (Atkinson & Shiffrin, 1971). Pisoni and Cleary (2003) have proposed that working memory capacity and verbal rehearsal speed may contribute an extra source of variance to post-operative outcome measures. They found that means for the digit span tests were shorter in the implanted children than age-matched normally hearing children, suggesting that deaf children have atypical development of short-term working memory capacity. Better digit span results were obtained from the children using oral communication compared to those who used some signing support, and the authors suggested this may be due to children from signing backgrounds being exposed to less oral language, leading to problems in processing and rehearsing auditory information in short-term memory. Pisoni and Cleary propose that variability in performance on traditional outcome measures in children may reflect fundamental differences in the speed of
information processing such as verbal rehearsal and the rate of encoding phonological and lexical information in working memory (Pisoni & Cleary, 2003).

In the current study, no significant correlations between non-verbal IQ as measured with the Raven’s Progressive Matrices test were observed with post-operative speech perception results. Future research may find that analysis of working memory, speed of speech analysis and comparisons with sentence recognition may provide more significant associations. It is also possible that when investigating possible correlations with outcome measures, greater subject numbers may be required to reach significance.

11.4 Non-Verbal IQ Conclusions

Results from the current study show no correlation between the RPM raw score or standard score and post-operative phoneme score or speech intelligibility score. There was no difference in RPM raw score between those AEO-HL participants who used oral language and those who used signing support. There was however, a significant correlation with receptive language as measured with the PPVT.

Results do not support Hypothesis 8: That those participants with lower scores on the Raven’s Matrices non-verbal test of IQ will achieve lower post-operative speech perception scores than those who achieved higher scores

Measurement of non-verbal IQ, as measured with the RPM, does not appear to predict post-operative performance on speech perception tests for this cohort of 24 AEO-HL.
12 SELF-REPORT OUTCOME QUESTIONNAIRE

Results from previous chapters have demonstrated that post-implant speech perception performance for AEO-HL is variable, but has been shown to correlate with the speech intelligibility, reported communication mode, and gap detection abilities of an individual recipient. It is possible that the development of intelligible speech and the ability to accurately detect temporal gaps may reflect the presence and utilisation of audition during infancy. The measurement of acoustic or evoked CAEP, whilst useful in children, has not provided extra insight into the development of the auditory pathway for these study participants. Non-Verbal IQ was not shown to influence post-operative performance. This study aimed to investigate outcomes and predictive factors for AEO-HL. While the previous chapters have explored such predictive factors, the relationship between objective and subjective benefit for recipients will be now be explored.

Specifically, the following hypotheses were tested:

Hypothesis 9: That the use of a self-report outcome measure will accurately reflect post-operative results on speech perception testing.

Hypothesis 10: That results of the self-report outcome questionnaire will be lower for AEO-HL than for those adults with an acquired hearing loss.

12.1 Self-Report Outcome Questionnaire Methods

The present study developed a questionnaire to explore recipients’ use of the cochlear implant, their own view on their communication abilities, and their confidence and satisfaction with the device. The questionnaire consisted of 15 questions with a 5-point scale. In all questions ‘5’ was the most positive response. Refer to Appendix C for a copy of the questionnaire.

The questionnaire was divided up into four sections. Section one consisted of two questions regarding device use. Section two consisted of nine questions regarding
communication. Section three consisted of two questions regarding confidence, and section four consisted of two questions regarding satisfaction with the cochlear implant. The questionnaire was administered to all AEO-HL participants at 12 and 24 months following their cochlear implant operation. Twenty-four questionnaires were completed at the 12-month point, and 22 at the 24-month point. Eighteen subjects completed the questionnaire at both the 12 and 24-month post-operative point.

This novel questionnaire was developed for the purposes of this study, but there were two questions which were unclear to the participants, for example questions three and four appeared to generate some confusion amongst participants. Question three asked how participants mainly communicated prior to receiving their cochlear implant, and question four asked how they mainly communicated after receiving their cochlear implant (5 = Listening, 4 = Listening and Lipreading, 3 = Lipreading, 4 = Sign Language, 1 = Written Notes). Some participants seemed to select every mode of communication they may have used, rather than selecting the one mode that they used most of all. If a participant selected two adjacent categories, then an intermediate value was assigned, essentially creating a ten-point scale. If three categories were selected, or two non-adjacent categories were selected then that data point was removed from analysis. This confusion regarding interpretation of Q3 and Q4 needs to be taken into consideration before interpreting the results.

In order to compare results from the AEO-HL group with persons who had an acquired hearing loss, additional questionnaires were administered to adults with acquired hearing loss from the general Melbourne Cochlear Implant Clinic recipient group at 12-months post-implant. Questionnaires were sent to 31 recipients who were under 65 years of age, had a severe to profound hearing loss pre-operatively, and received their cochlear implant between 2011 and 2014, ensuring that they had 12 months of device use at the time the questionnaire was administered. A total of 15 questionnaires were returned, and combined with the four collected from the four participants with acquired hearing loss who completed the full battery of tests for this project, a total of
19 questionnaires were completed. This group of 19 adults with acquired hearing loss will be referred to as the PostL2 group.

12.2 Self-Report Outcome Questionnaire Statistical Analysis

As questionnaire data is ordinal data, when comparing results between the AEO-HL and PostL2 groups and between the 12 and 24-month AEO-HL questionnaires, the non-parametric Mann-Whitney test was used to analyse differences between the two sample medians.

Question 1, 5, 14, 15, and by default the Device Satisfaction section, were given a result of ‘5’ by every subject in the PostL2 group. Therefore, a 1 sample t test was necessary to compare the AEO-HL group mean with a perfect score for these questions.

The relationship between questionnaire data and speech perception results was analysed using the Spearman correlation analysis, to evaluate the relationship between two variables which may not change at a constant rate.

Analysis was performed for each question individually, and for each section. When analysing results for each section, there were some questions that had been missed by individual participants. This question was removed from the analysis.

12.3 Self-Report Outcome Questionnaire Results

The questionnaires collected from the AEO-HL group at 12 and 24 months were compared. Results were then compared between the AEO-HL and PostL2 groups. The relationship between scores for the four domains of the questionnaire and speech perception outcomes were evaluated using correlation analysis.
12.3.1 AEO-HL Questionnaire results – 12-months post-operative vs 24-months post-operative.

Questionnaires from both the 12 and 24-month post-operative point were collected from 18 participants.

To investigate any differences between the two collection points, results were compared for each question using the Mann-Whitney non-parametric test.

Q 1 ‘How many hours per day do you wear the device?’

Q 2 ‘How many hours do you wear a device on the contralateral ear?’

Table 7: Statistical analysis for Q1 and Q2 for the AEO-HL group

<table>
<thead>
<tr>
<th></th>
<th>Q1</th>
<th></th>
<th>Q2</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Mann Whitney</td>
<td></td>
<td>Mann Whitney</td>
<td></td>
</tr>
<tr>
<td></td>
<td>W = 375</td>
<td></td>
<td>W = 324</td>
<td></td>
</tr>
<tr>
<td></td>
<td>p= 0.8793</td>
<td></td>
<td>p= 1.0</td>
<td></td>
</tr>
</tbody>
</table>

There was no significant difference in how many hours per day the group wore their cochlear implant, or a device on the contralateral ear from 12 to 24-months post-operative.

Q 4 ‘With your cochlear implant, how do you mainly communicate?’

Table 8: Statistical analysis for Q3 and Q4 for the AEO-HL group

<table>
<thead>
<tr>
<th></th>
<th>Q4 Comparison between 12 &amp; 24 mths</th>
<th></th>
<th>Q3 Comparison between pre- and 12m post-op</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Mann Whitney</td>
<td></td>
<td>Mann Whitney</td>
</tr>
<tr>
<td></td>
<td>W = 303.5</td>
<td></td>
<td>W = 216</td>
</tr>
<tr>
<td></td>
<td>p= 0.1787</td>
<td></td>
<td>p= 0.0051</td>
</tr>
</tbody>
</table>

There was no significant difference between how the AEO-HL group felt they mainly communicated between 12 and 24 months post-operatively.
There were no other observed significant differences between 12 and 24-months post-operative for any of the remaining questions in the study questionnaire. The only significant difference observed was between the main mode of communication pre- and post-operatively. Please refer to Appendix D for a full table of statistical analyses.

12.3.2 Comparison between the AEO-HL and PostL2 Group.

Questionnaires received from the AEO-HL group and PostL2 group were compared by section; CI use, Communication, Confidence and Device Satisfaction, and also by individual question. As there were no differences observed between the 12 and 24-month point for the AEO-HL group, any responses provided at 24 months that were not provided at 12 months, were included in the analysis.

Table 9: Comparison between questionnaire domain results for AEO-HL and PostL2 groups

<table>
<thead>
<tr>
<th>CI Use (Q1)</th>
<th>Communication (Q4-11)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1 Sample T Test</td>
<td>t = -3.19</td>
</tr>
<tr>
<td>p = 0.003</td>
<td></td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Confidence (Q12-13)</th>
<th>Device Satisfaction (Q14-15)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mann Whitney</td>
<td>W = 549</td>
</tr>
<tr>
<td>p &lt; 0.001</td>
<td></td>
</tr>
</tbody>
</table>

There was more variability seen between hours of cochlear implant use in the AEO-HL group compared to the PostL2 group. All 19 participants in the PostL2 group wore their device for more than eight hours a day, compared to 21 (or 75%) of AEO-HL. Five AEO-HL participants wore their device between four and eight hours a day and two participants wore it between 1 and 4 hours per day.

There was a significant difference in the way the recipients perceived their communication abilities with a cochlear implant, with the PostL2 group rating their abilities higher than the AEO-HL group. The PostL2 group also rated themselves
significantly more confident and significantly more satisfied with their CI, than the AEO-HL group.

12.3.3 Comparison per question:

The Mann Whitney test was used to analyse any differences between the AEO-HL and PostL2 groups for each individual question, except for question one, 14 and 15 which required a single sample t test due to the PostL2 group scoring 100%. Please refer to Appendix D for a full table of statistical results. To display the results for each question in a graphical form, any intermediate categories were rounded up.

Q1 How many hours per day do you wear the device?

![Image of pie charts showing hours of device use]

Figure 27: Comparison between AEO-HL and PostL2 groups for Q1.

There was a significant difference between the AEO-HL and PostL2 group for the number of hours of device use ($t = -3.19$, $p = 0.003$). Every participant in the PostL2 group wore their cochlear implant for more than eight hours per day. There was some variation in the AEO-HL group, but 75% of participants still wore their implant more than eight hours a day. Eighteen percent wore their device between four and eight hours, and 7% wore it between one and four hours per day. One AEO-HL participant wore their device between four and eight hours a day at the 12-month point, but became a non-user before the 24-month point. There was no difference in PPVT standard score between the AEO-HL who wore their processor less than eight hours a
day compared to those who wore it over 8 hours per day \( (t=0.13, p=0.896) \). Those who wore it more than eight hours a day scored significantly better on CVC phonemes than those who wore it less \( (t=-2.2, p=0.045) \).

**Q2 Do you wear a hearing aid on your opposite ear?**

![Figure 28: Comparison between AEO-HL and PostL2 groups for Q2.](image)

There was no significant difference between the two groups regarding use of a hearing aid on the contralateral ear, although there was more variability observed in the AEO-HL group. It appears that the PostL2 group either wore a hearing aid full time, or not at all, whereas the AEO-HL group varied in the time they wore a contralateral hearing aid.

**Q3 Before you had your cochlear implant, how did you mainly communicate?**

![Figure 29: Comparison between AEO-HL and PostL2 groups for Q3.](image)

There was a significant difference observed between the main communication mode selected for the AEO-HL and PostL2 group \( (W=424, p=0.023) \) with 84% of PostL2
subjects choosing ‘listening and lipreading’ as their main mode of communication compared to 56% of the AEO-HL group.

Q4. With your cochlear implant, how do you mainly communicate?

Figure 30: Comparison between AEO-HL and PostL2 groups for Q4.

After receiving their cochlear implant, there was again a significant difference seen between the two groups in terms of their main mode of communication. The majority (74%) of the PostL2 group reported that they relied only on listening for communication, with 26% requiring some lipreading support. For the AEO-HL group on the other hand, 20% reported that they could communicate by listening alone, with 76% reporting that listening and lipreading together was necessary, and one participant who relied on lipreading alone for communication.

Questions three and four were the most poorly answered by the AEO-HL group, due to some degree of confusion. There were 29 participants who responded to Q1, yet only 23 who provided a valid answer to Q3 and 25 to Q4. These questions specifically asked for the main mode of communication used prior to cochlear implantation and that used post-operatively. Six AEO-HL participants appeared to choose every communication mode they had used, making their responses invalid. All six of these subjects were those who had selected sign language. All 19 participants in the PostL2 group provided valid answers to these questions.
Q5. Do you feel that the cochlear implant has made it easier for you to communicate?

Figure 31: Comparison between AEO-HL and PostL2 groups for Q5.

Results from Question five indicated that the cochlear implant made communication ‘much easier’ in 100% of participants in the PostL2 group, and 55% of the AEO-HL group. Of the 29 AEO-HL participants, 34% reported that the implant made communication a little bit easier, 3% reported it made no difference, and 7% reported that it actually made communication a little more difficult. This equated to a significant difference between the groups (t = -4.07, p =0.000)

Q6. How do you rate your ability to understand speech when combined with lipreading?

Figure 32: Comparison between AEO-HL and PostL2 groups for Q6.
There was a significant difference between the groups in their perceived ability to understand speech when combined with lipreading, with the PostL2 group reporting that they understood more speech ($W=603.5$ $p=0.0072$) than the AEO-HL group. For both groups however, the most popular category was ‘I understand most things’ which was selected by 72% of the AEO-HL group, and 68% of the PostL2 group. In the PostL2 group, 32% of recipients reported that they could understand everything, compared to 10% of AEO-HL. In the AEO-HL group, 14% reported that they could understand about half, and one participant reported that they could understand some things. No participant in either group reported being unable to understand any speech when combined with lipreading.

**Q7. How do you rate your ability to understand speech with no lipreading?**

![Figure 33: Comparison between AEO-HL and PostL2 groups for Q7.](image)

A significant difference was observed between the rated ability to understand speech with no lipreading, with the majority of the PostL2 group (63%) reporting the ability to understand most of the conversation without lipreading, compared to 17% of the AEO-HL group ($W=518$, $p=0.000$). Twenty-one percent of the PostL2 group reported that they could understand about half of speech, compared to 28% of the AEO-HL participants. The most popular rating for the AEO-HL group, at 41%, was the category of being able to understand ‘some’ things, whereas only one PostL2 participant
selected this category. None of the PostL2 group reported being unable to understand any speech without lipreading, whereas 14% of the AEO-HL group did.

Q8. How do you rate your ability to follow conversation in a group?

![Pie charts comparing AEO-HL and PostL2 groups for Q8](image)

**Figure 34: Comparison between AEO-HL and PostL2 groups for Q8.**

A significant difference was observed between the two groups in the amount of group conversation which could be understood (W=605.5, p=0.0188). The majority of PostL2 participants (59%) reported being able to understand ‘most things’ when listening in a group, whereas an equal number of AEO-HL participants reported being able to understand ‘most things’ (38%) and ‘some things’ (34%). Only one PostL2 participant reported that they could only understand some things in a group.
Q9. How do you rate your ability to hear and recognise environmental sounds?

Figure 35: Comparison between AEO-HL and PostL2 groups for Q9.

There was no significant difference in perceived ability to hear and recognise environmental sounds between the two groups (W = 670.5, p = 0.3556). Both groups reported being able to hear and recognise most things (59% for the AEO-HL group and 58% for the PostL2 group) and no participant from either group reported that they were unable to recognise any environmental sounds.

Q10. Do you enjoy listening to music with your cochlear implant?

Figure 36: Comparison between AEO-HL and PostL2 groups for Q10.
There was no significant difference between the two groups in terms of music enjoyment (W=667.5 p= 0.4129). The majority of both groups (63% of AEO-HL and 53% of PostL2 group) reported that listening to music was very enjoyable with their cochlear implant. One participant in each group did not enjoy listening to music at all.

Q11. Are you able to hear on the telephone with your cochlear implant?

![Figure 37: Comparison between AEO-HL and PostL2 groups for Q11.](image)

There was a significant difference regarding telephone use between the two groups, with the PostL2 group significantly more comfortable hearing on the telephone (W=493.5, p= 0.0013). Only one participant in the PostL2 group reported being unable to hear on the telephone at all, whereas 14 AEO-HL participants, or 52%, could not hear on the telephone.
Q12. Do you feel more confident in a group when wearing your cochlear implant?

![Comparison between AEO-HL and PostL2 groups for Q12.](image)

There was a significant difference in reported confidence levels between the two study groups when participants wore their speech processor in a group situation ($W=572$, $p=0.0015$). Of the PostL2 group, 79% felt much more confident, with the remaining 21% feeling slightly more confident. There was a much wider range of confidence levels reported in the AEO-HL group, however 38% still reported feeling ‘much more confident’ and 31% feeling ‘slightly more confident’.
Q13. Do you feel more confident with unfamiliar people when wearing your speech processor?

![Comparison between AEO-HL and PostL2 groups for Q13.](image)

A significant difference was observed between the two groups in terms of how confident they felt with unfamiliar people when wearing their speech processor, with the PostL2 group reporting higher levels of confidence ($W=511.5$, $p=0.0001$). Again, a much wider range of confidence levels was reported for the AEO-HL group, however 36% still reported feeling ‘much more confident’, and 32% felt ‘slightly more confident’. All participants in the PostL2 group reported feeling either ‘much more’, or ‘slightly more’, confident.
Q14 How satisfied are you with your cochlear implant?

Figure 40: Comparison between AEO-HL and PostL2 groups for Q14.

All participants in the PostL2 group reported feeling ‘very satisfied’ with their cochlear implant, which was significantly different to the AEO-HL group (t= -3.33, p= 0.002). More variability was observed in the AEO-HL group, however the majority 66% reported feeling ‘very satisfied’. One participant reported feeling very dissatisfied, but it is suspected that this was a response made in error, as they reported feeling ‘very pleased’ that they received the cochlear implant in the following question.
Q15 Are you pleased that you had the cochlear implant?

![Pie charts showing responses to Q15](Image)

**Figure 41: Comparison between AEO-HL and PostL2 groups for Q15.**

There was no significant difference between the two groups when recipients were asked if they were pleased they had received the cochlear implant. All participants in the PostL2 group reported being very pleased that they had received a cochlear implant, as did the greater majority of AEO-HL participants (86%). No participant, from either group, reported being either somewhat, or very, displeased that they had received the cochlear implant ($t=-1.98$, $p=0.057$)

### 12.3.4 Correlations with speech perception:

Speech perception was positively correlated with perceived communication rating for both the AEO-HL and PostL2 groups, with participants who scored higher on CVC phonemes rating their communication abilities higher than those who achieved lower scores. There was no correlation between device use and speech perception for either group.
Figure 42: Relationship between perceived communication ability rating and post-operative speech perception performance for the 29 AEO-HL.

Figure 43: Relationship between perceived communication ability rating and post-operative speech perception performance for the 19 participants in the PostL2 group.
Confidence was not correlated with speech perception for the PostL2 group \((\rho = 0.216, p=0.374)\) with all participants rating their confidence in group situations and with unfamiliar people higher with their cochlear implant. Fourteen of the 19 participants rated their confidence at 100%, four at 90% and one at 80%. There was greater variability seen in the confidence ratings of the AEO-HL group, with a range of 40 – 100%. The mean was 70% and the median 80%. A positive significant correlation with speech perception was observed with the Spearman coefficient \((\rho=0.411 \ p=0.037)\).

![Figure 44: Relationship between the Confidence Rating and speech perception performance for the 29 participants in the AEO-HL group.](image)

The PostL2 group all rated their overall satisfaction with their cochlear implant at 100%. Speech perception and device satisfaction was positively correlated for the AEO-HL group \((\rho=0.404, p=0.03)\). There was less variability seen in the satisfaction ratings of the AEO-HL group than observed in their confidence ratings, with 18 of the 29 participants rating their overall satisfaction at 100%. The group mean was 92% with a median of 100%, indicating that irrespective of speech perception results, the AEO-HL
group was very satisfied with their cochlear implant and very pleased that they had chosen to be implanted.

Figure 45: Relationship between the overall Satisfaction Rating and speech perception results for the 29 participants in the AEO-HL group.

12.4 Self-Report Outcome Questionnaire Discussion

Analysis of questionnaire data demonstrated that both AEO-HL and PostL2 participants reported that the cochlear implant had made a positive impact on their life. Analysis of the data from the AEO-HL study group showed no significant difference between results obtained 12 months post-operatively, and 24 months post-operatively. This finding contrasts with anecdotal reports from clinicians which suggest that AEO-HL recipients tend to take a greater amount of time to adjust to their cochlear implant and reach their full potential than adults with an acquired hearing loss. These results do, however, support the finding in the current study of no significant difference in speech perception between the 12 and 24 month post implant point; consistent with Zeitler and colleagues (Zeitler et al., 2012). It is possible that one reason the participants in the present study adapted fairly quickly to their cochlear implant may be that they all used some form of spoken language for communication; either
auditory/oral alone, or auditory/oral in combination with some signing support. No AEO-HL participant in the present study communicated through sign language alone.

For all four domains of the questionnaire; Device Use, Communication, Confidence and Satisfaction, significant differences were observed between the AEO-HL and PostL2 groups, with higher scores obtained from the PostL2 group. Further investigation into the individual questions has highlighted areas of similarity and differences between the two population groups.

There was more variability in the AEO-HL group with regards to the amount of time per day the device was worn, when compared to the PostL2 group who all wore their device full time by the 12-month post-operative point. A significant difference in speech perception was observed between those AEO-HL who wore their CI more than eight hours per day, and those who wore it less than eight hours. The AEO-HL who wore it more than eight hours demonstrated higher speech perception scores, and therefore may have elected to wear the processor more as a reflection of the benefit provided by the CI. When asked about the use of a contralateral hearing aid, results for the PostL2 group showed that a hearing aid was either worn full time, or not at all, suggesting that if benefit was gained through use of a hearing aid then it was worn, and if there was no perceived benefit then it was not used. Results for the AEO-HL group, however, were more variable. Hearing aid use varied in the number of hours worn per day, perhaps reflecting the variability in the number of hours the cochlear implant was also worn. It may be that the AEO-HL group, as a whole, was less reliant on their hearing devices. This may be a consequence of their history of limited audition; these participants may be more comfortable spending some time per day in quiet, confident in their ability to lipread if necessary. The participants in the PostL2 group, on the other hand, have had access to a good auditory signal for the majority of their life, and may find times of silence disconcerting. It is also possible that the PostL2 group received more benefit from their contralateral hearing aid, as the group showed a significantly lower PTA of 84dB compared to 107dB in the AEO-HL group.
Pre-operatively, a combination of listening and lipreading was the main form of communication for both groups, and both demonstrated a significant shift towards a more oral mode of communication post-operatively, which was reflected in their improvement in speech perception scores post-operatively. The PostL2 group did rate themselves significantly more ‘oral’ than the AEO-HL group, however, both pre- and post-operatively. These questions appeared to generate some confusion amongst the AEO-HL group, with some participants selecting multiple communication modes, thus rendering their score invalid. These participants were those who also relied on sign language to some extent. Therefore, the participants who utilised sign support may not be accurately represented in the results for these questions. It has been suggested that the use of written questionnaires may not be valid for the AEO-HL population, as they rely on a certain level of literacy (Connolly et al., 2006). Whilst every attempt was made to ensure that the language in this questionnaire was kept simple, it is possible that some of the questions were still too complex for some participants, or that the wording was sometimes ambiguous.

A significant difference was observed between the two groups for all questions relating to speech understanding, listening in noise and use of the telephone, with the PostL2 group rating themselves significantly higher than the AEO-HL group. No difference was observed between the two groups in terms of hearing and recognising environmental sounds, enjoyment of music, and when asked if they were pleased they had received a cochlear implant. The majority of both groups felt they could recognise most environmental sounds. The benefits of improved ability to hear environmental sounds should not be underestimated. In a traditional clinical setting, outcomes are generally quantified by performance on speech perception tests, but for AEO-HL recipients the ability to hear and connect to their environment can be just as important. Two participants in this study made interesting anecdotal comments about their environment. Participant 25 was a busy parent of two children. Participant 25’s phoneme score was 32%, but one of the biggest benefits reported was the ability to hear the washing machine beep from another room. Participant 27, who scored 7% for phonemes, was new to parenthood. Participant 27 reported that the implant actually
detracted from lipreading ability, but was very grateful that the implant enabled
detection of his baby crying. These two recipients may be considered ‘poor’
performers in terms of speech perception scores, but they both reported their
implants had been a success, and both participants reported they were ‘very pleased’
they had received their implants.

The degree to which a recipient may or may not report benefit from implantation may
be based on their own expectations of the cochlear implant. This is an area where
cochlear implant clinicians can influence and guide a recipient’s expectations
throughout the candidacy evaluation. A study by Chee and colleagues reported that
the majority of their 30 prelingually deaf cochlear implant recipients reported
increased levels of safety due to increased sound awareness (Chee et al., 2004),
highlighting the fact that investigating areas other than objective speech perception
performance is important for this patient group.

In the present study, there was no significant difference in the enjoyment of music
between the two groups, with 63% of AEO-HL participants and 53% of the PostL2
participants rating music as ‘very enjoyable’. Anecdotal reports from cochlear implant
clinicians suggested that implant recipients with acquired deafness often comment
that music does not sound the same as they remembered it. These recipients have a
clear memory of how a piece of music should sound, and the imperfect processing of
the music by the implant may not match this memory. AEO-HL cochlear implant
recipients, on the other hand, often comment that they really enjoy listening to music.
They may not be able to understand the lyrics, but can follow the beat and melody of
the song. These recipients do not have a memory of how the piece of music should
sound, in its entirety. They may only have had access to an incomplete version of the
music through their hearing aids, so the addition of the high frequency information
provided by the implant may allow them an enhanced signal with which to follow a
piece of music. The findings of the present study are supported by those of Moran and
colleagues (2016) who also found an equivalent enjoyment of music between a group
of 15 adult cochlear implant recipients with early onset hearing loss and 15 recipients
with an acquired hearing loss (Moran et al., 2016). This study analysed music appreciation through the use of the University of Canterbury Music Listening Questionnaire (UCMLQ) (Looi & She, 2010) and found that while the accurate ‘perception’ of musical styles was poorer for the AEO-HL group, the enjoyment and appreciation between the two groups was equivalent, and suggested that perception and enjoyment should be considered separately. Both the current study, and the study by Moran et al., found a trend for higher levels of enjoyment for the AEO-HL group, but differences between the AEO-HL and PostL2 group were not significant. The enjoyment of music is likely due to the provision of extra information by the cochlear implant that was not available through a hearing aid (Eisenberg, 1982).

The overall differences observed between the AEO-HL and PostL2 participant groups across all questions of the questionnaire, reflect the fundamental differences in auditory development between these two groups. The cochlear implant is merely replacing lost information for the PostL2 group, and once the signal had been restored the majority of this participant group were able to utilise the information to assist in communication and perception. Certain situations, such as noisy groups, are still difficult due to the limitations of microphones and speech processor capabilities, but for many recipients the implant is able to restore them to a purely oral mode of communication (Blamey et al., 2013; Skinner et al., 2002). For the AEO-HL group, however, the cochlear implant may be providing entirely new information, rather than replacing lost information. The ability of an individual to utilise this new information is variable, and may depend on the development of the auditory system (Caposecco et al., 2012). The present study showed that the additional information delivered by the implant provided the AEO-HL participants with the ability to hear and recognise environmental sounds, and enjoy music to the same degree as the PostL2 participants.

A number of studies have investigated the use of questionnaires in addition to speech perception testing, to evaluate benefit for cochlear implant recipients in a more functional capacity. Health related QoL tools have been used to demonstrate significant improvements in QoL, self-esteem and confidence for cochlear implant
recipients with acquired hearing (Damen et al., 2007; Hinderink et al., 2000; Lassaletta et al., 2006). Comparable questionnaire results have been observed between adult cochlear implant recipients with an acquired or early-onset hearing loss, regardless of the difference in speech perception performance, (Straatman et al., 2014). A lack of correlation between speech perception results and quality of life measures for AEO-HL has been reported in a number of studies, indicating that factors other than objective performance are important to this patient group (Straatman et al., 2014; Zwolan et al., 1996). Zwolan and colleagues looked at questionnaires administered pre- and post-operatively for a group of AEO-HL. The study group demonstrated little or no improvement in open set speech perception, but the majority wore their device more than 10 hours per day and reported that they were satisfied with their cochlear implants (Zwolan et al., 1996).

Results from this present study differed from the majority of published literature as the study questionnaire demonstrated a significant correlation between objective performance of the AEO-HL group and three of the four domains investigated. Speech perception was positively correlated with ‘perceived’ communication for both groups, and a significant positive correlation was observed for device satisfaction and confidence in different listening situations for the AEO-HL group. There was no correlation between device use and post-operative speech perception for either group, indicating that irrespective of objective benefit, most recipients wore their device more than eight hours a day. Results showed that that even though the AEO-HL group demonstrated equivalent scores to the PostL2 group for the questions relating to music appreciation and environmental sound awareness, they show significant differences in all other domains. Although there was no difference between the two groups when asked if they were ‘pleased’ they had received the implant, it is important to consider that this may be viewed as a leading question. The process of cochlear implantation involves a considerable investment of time, energy, and emotion for a recipient. For the recipient to then regret their decision, and not report being ‘pleased’ that they had received the implant, may take a degree of strength. Results from this question may therefore not be a true reflection, and it may be more
prudent to consider that the study did demonstrate a correlation between the ‘Device Satisfaction’ domain (of which this question forms 50%) and objective performance.

The specifically designed questionnaire used in this study was developed to address particular areas of communication that experienced clinicians felt would present difficulties for AEO-HL implant recipients, as well as questions relating to areas commonly reported as being improved by a cochlear implant. It is possible that the specific nature of this questionnaire was responsible for the observed correlations with performance that may not be observed in other studies.

### 12.4 Self-Report Outcome Questionnaire Conclusions

Although post-operative speech perception performance is poorer for AEO-HL compared to adults with an acquired hearing loss, both groups demonstrated significant benefit from their cochlear implants. Recipients in both groups rated their communication abilities higher when they obtained greater results on speech perception testing. Confidence and Device Satisfaction was correlated with objective performance for the AEO-HL group, however overall levels of satisfaction were high, suggesting that the way a recipient evaluates their decision to receive a cochlear implant is not only dependent on their performance on speech perception testing.

Results from the current study support the following hypotheses:

Hypothesis 9: That the use of a self-report outcome measure will accurately reflect post-operative results on speech perception testing.

Hypothesis 10: That results of the self-report outcome questionnaire will be lower for AEO-HL than for those adults with a postlingual hearing loss.

Results from the study questionnaire do reflect objective speech perception results in most domains, suggesting that questionnaire data may be a valid means of assessing post-operative outcomes with this patient group.
13 DISCUSSION

13.1 Introduction

Cochlear implantation has become the intervention of choice for the majority of adults with an acquired severe to profound hearing loss, and young children with hearing loss of a severe degree or worse. Cochlear implant outcomes for these patient groups are generally excellent, with the majority of adults regaining the ability to follow speech through audition, and many children developing age appropriate speech and language skills (Blamey et al., 1996; Chee et al., 2004; Dowell et al., 2004; Geers et al., 2003; Kral et al., 2016; Leigh et al., 2013). Increasing numbers of adults with a significant early-onset hearing loss are now exploring the option of cochlear implantation, as acceptance of the device as an appropriate intervention aid is growing within the Deaf Community (Hladek, 2002). The Melbourne Cochlear Implant Clinic alone now has 403 AEO-HL who have received cochlear implants, out of a total of 2111 as of March 2017.

It has been well documented that cochlear implant results on objective speech measures for AEO-HL are on average below those typically recorded for adults with an acquired hearing loss, and tend to show greater variability; from good open set speech perception, through to an aid to lipreading, environmental sound awareness, or no auditory percept at all (Caposecco et al., 2012; Eisenberg, 1982; Kaplan et al., 2003). This wide variability in outcomes makes it difficult for the clinicians working with these implant candidates to provide appropriate counselling, thereby ensuring that the candidate has realistic expectations as to the limitations and possible benefits of cochlear implantation.

This study was designed to attempt to address some of the difficulties encountered by cochlear implant clinicians by investigating aspects of auditory development. A series of studies were performed to address the language, communication, psychophysical, electrophysiological, cognitive and self-report measures of individual study participants. This study aimed to provide insights into the variables that may contribute to cochlear implant outcomes. The relationship between objective
outcomes and self-report measures were also investigated, as it has been shown that the two are not necessarily well correlated (Chee et al., 2004; Eisenberg, 1982; Zwolan et al., 1996).

13.2 Objective Outcome Measures

*Neural Plasticity*

The literature has shown that appropriate development of the auditory system can only occur with auditory experience (Kral & Eggermont, 2007; Kral et al., 2016). For children with congenital profound hearing loss, it is best practice to provide a cochlear implant as soon as practicable in order to minimise the effects of auditory deprivation and optimise communication outcomes (Sharma & Dorman, 2006; Sharma et al., 2002b). Adults with a significant early-onset hearing loss can be considered to be the children who were not given the opportunity for early implantation. It has been suggested that ‘inter-individual variability in the brain’s adaptation to sensory loss underpins much of the observed variation in outcome of cochlear implantation’ (Kral et al., 2016). Whilst variability in communication outcomes is observed amongst all cochlear implant recipients, the degree of variability in speech understanding is greater in adults with an early onset hearing loss. Results from the current study suggested that auditory deprivation and lack of auditory language input in early childhood may be the greatest contributing factor to speech perception outcomes. It is possible that some of the variability in adaptation to sensory loss, as discussed by Kral and colleagues, may in fact be linked to the severity and age of onset of hearing loss, and the individual’s functional use of residual hearing to develop communication.

Kral and Eggermont [2007] have suggested that auditory experience is crucial for the proper activation of higher-order auditory areas within the brain. In the absence of hearing, the primary auditory cortex remains capable of responding to auditory stimuli, but the functionality of the auditory cortex is significantly affected by the deprivation (Kral et al., 2006). In studies on the primary auditory cortex of congenitally deaf cats, it was shown that there was a decrease in activation in the infragranular
layers (Kral et al., 2001). These layers, at least in humans, are the points of origin for the top-down processing pathways from the higher-order areas. The absence of activity in these infragranular layers can be interpreted as a functional decoupling of the primary cortex from higher-order auditory cortices. The presence of a congenital hearing loss in humans not only affects the primary auditory cortex, but also the synaptic connections of the auditory association areas. The auditory component of the connectome model includes ‘bottom up’ connections from the auditory cortex to the higher order auditory areas, and the top down channel of processing, consisting of the connections between these higher order areas back down to the primary auditory cortex (Kral et al., 2016). In the absence of auditory input, these important feedback loops are weakened (Sharma et al., 2009). In the congenitally deaf model, without this top-down processing and developmental reduction in synapses, plasticity will become non-adaptive and the auditory cortex will lose the ability to learn and adapt to the environment (Kral & Eggermont, 2007).

At the end of the sensitive period, and in the absence of auditory stimulation, primary and secondary auditory areas become decoupled; these areas are no longer able to develop interconnections, even when a cochlear implant is provided, and feedback loops are unable to develop (Sharma et al., 2009). Gilley, Sharma and Dorman (2008) used high density EEG measures to analyse the source of the CAEP in response to a speech sound in children implanted prior to 3 ½ years and after 7 years of age. In the early implanted children, activation was observed in the auditory cortical areas contralateral to their cochlear implant, which is the same as the activation pattern observed in normally hearing children. In the late implanted children, however, activation was observed outside of the auditory cortical areas (often in the visual or parietotemporal area). The authors suggested that the absence of activation within the auditory cortical area of the late implanted children indicated absent or weak connections between primary and association areas (Gilley et al., 2008). This functional disconnection between the primary and higher-order cortices may also underly the end of the sensitive period in children with congenital profound hearing loss (Sharma et al., 2009).
Large studies investigating cortical maturation in children with congenital deafness after cochlear implantation have consistently shown a period of developmental plasticity which is maximal up to age 3.5, and which decreases significantly after age seven (Sharma & Dorman, 2006; Sharma et al., 2002b). Lee and colleagues used PET scans to investigate resting cortical metabolic rate and found significant differences for resting glucose metabolic rate between those children implanted prior to age four and those implanted after age seven. The late implanted children showed normal metabolism in the higher order auditory cortices in the absence of speech sounds, suggesting that these cortical areas had been recolonised (Lee et al., 2005). Post-mortem studies have shown that synaptic density peaks at age 2-4, and that after this point, in the case of significant hearing loss and a corresponding lack of auditory experience, synaptic pruning does not take place (Huttenlocher, 1984; Huttenlocher & Dabholkar, 1997). This results in a naïve auditory cortex, which is unable to appropriately process the incoming signal provided by the cochlear implant (Kral et al., 2006). It has been suggested that stimulation via cochlear implantation during the sensitive period is crucial for synaptogenesis and subsequent pruning and refinement of synaptic networks to become established, allowing for a functional and specialised auditory system (Kral & Sharma, 2012). These multitudes of studies highlight aspects of neurodevelopment which point to a significant decrease in cortical plasticity in the auditory system after age three or four.

Functionally, studies of speech perception and language outcomes in children support this age cut off, with children achieving far greater speech perception when they receive an implant prior to the end of the critical period. When we investigate the more subtle aspects of language development, as opposed to pure speech perception, it would seem that implantation prior to 12 months of age provides the child with the best possible opportunities (Dettman et al., 2016; Leigh et al., 2013; Levine et al., 2016). There is a pronounced reduction in synaptic plasticity in the auditory cortex during early deafness, but some residual plasticity does remain into adulthood (Kral & Sharma, 2012; Most et al., 2010). AEO-HL who receive cochlear implants, however, often struggle to interpret the signal provided by the cochlear implant effectively, even
after long periods of device use. It has been suggested that the decoupling of the cortical layers which can occur with early onset hearing loss, disrupts the necessary bottom-up input, intrinsic processing and cortical top-down feedback, which is necessary for the correct interpretation of a speech signal (Kral & Sharma, 2012).

A functional impact of ‘top-down’ and ‘bottom-up’ processing has been proposed by Boothroyd’s model of speech perception (Boothroyd, 1985). The implications of disruptions to the connectome model between the primary auditory cortex and associated higher-order areas (Kral et al., 2016), can be related to this functional understanding of speech perception. Top-down language processing refers to the understanding of speech perception as meaningful, and bottom-up refers to the analysis of the incoming sound by the cochlear, the auditory nerve and the auditory areas of the cerebral cortex. Boothroyd suggested that the comprehension of speech is comprised of evidence available to the listener, their knowledge of language, and their ability to quickly analyse the incoming signal while extracting the meaning (Boothroyd, 1985). When a cochlear implant recipient with an acquired hearing loss receives a degraded auditory signal through their cochlear implant, they are often able to make use of top-down processing, or their knowledge of language, to ‘fill-in’ any missing gaps. This ability is not always available to AEO-HL CI recipients due to the likely disruption to the connectome, poorly developed auditory association areas, or auditory association areas which have been colonized by the visual system. These recipients often have a language delay and do not have the ability to utilise top-down processing to ‘fill-in’ the gaps, and therefore if sentence tests were used for analysis of outcomes, the interpretation of scores would need to consider any language issues as well as processing of auditory input. To address this factor, the present study chose to use the monosyllabic word test phoneme score as the speech perception measure for analysis. Although some recipients may still try to replace an unknown word with a known word, the test is more of an imitative task than a sentence perception test which may be more susceptible to language delays. For these reasons it has been suggested that phoneme scores are a more sensitive measure of speech perception when working with patients who may have language delays (Dowell et al., 2002).
Whilst the speech perception results obtained on CVC phonemes in the current project were significantly lower for the study cohort of 29 AEO-HL than those observed from the MCIC averages for adults with an acquired hearing loss, the group still showed significant improvement over pre-operative speech perception scores. The average post-operative CVC phoneme score for the group of 576 adults with an acquired hearing loss was 64.44%, compared to 35.61% for the AEO-HL group. The lower scores observed for the AEO-HL recipient group are likely the result of several factors such as a disruption to their auditory connectome, and potential language delays possibly leading to decreased use of top-down processing. These factors may conceivably be a consequence of the difference in early hearing history between the two groups of recipients, leading to a disruption in the maturation of the auditory processing and oral communication abilities of the AEO-HL.

*Language*

Even without the use of hearing, a typical trajectory of language development can be achieved if a visual language is provided early in life (Mayberry, 2010). Thus it appears that the critical periods of language development are independent of modality (Werker & Hensch, 2015). Mayberry and colleagues (2011) used fMRI to investigate linguistic tasks in sign language. They found that for those individuals born deaf who learned sign language at an early age, neural activation was observed in the classic language areas of the brain. For those deaf individuals who were not given the opportunity to learn sign language during infancy, and who were unable to learn spoken language due to the severity of their hearing loss, observed activation patterns deviated from the expected norms (Mayberry et al., 2011) A crucial finding of the study, was that the functional anatomy for spoken and signed language processing was similar, but only observed when language acquisition began early in life. The authors suggested that first language acquisition, either spoken or signed, needed to occur during the period of brain growth, in order for the classic neural network of language processing to reach its maximum potential in the adult brain.
Results from the current study did not show significant correlation between receptive language, as measured by the PPVT and post-operative speech perception scores. A language delay, specifically receptive vocabulary knowledge that was two or more standard deviations below the mean was demonstrated by 25% of the AEO-HL study group. The average score for the group was greater than one standard deviation below the population mean. It has been shown that identification of hearing loss, and appropriate intervention needs to be implemented prior to six months of age (Yoshinaga-Itano et al., 1998) in order to minimise language delays. As universal hearing screening has only been in practice since 2005 in Victoria, Australia and all patients in the study cohort were deaf children of hearing parents, it is highly unlikely that they would have received the appropriate diagnosis and intervention prior to six months of age. This delay in intervention may have contributed to the language delay observed. There was no difference in receptive language observed between the oral group and the signing support group, which reflected past published results (Habib, Waltzman 2010, van Dijkhuizen 2011). The development of language may not be dependent on a purely oral mode of communication, and as Mayberry’s study has demonstrated, fMRI shows that sign language is able to activate the appropriate language areas in the brain for children who were immersed in a fluent sign language environment at the appropriate ages, thereby meeting the critical periods for language acquisition (Mayberry et al., 2011). It would appear that the provision of an appropriate sign language model during infancy has the potential to lay the foundations for language acquisition as effectively as oral communication.

Whereas ‘language’ can be effectively generated through audition or signing, providing the critical periods are met, the aspects of ‘oral’ language such as speech production and speech perception, are more dependent on access to the relevant acoustic cues via audition. In the present study, significantly higher post-operative speech perception results and significantly higher speech intelligibility ratings, were obtained from the group using oral communication. Typically, the development of intelligible speech production requires robust and consistent auditory input at an early age. The current study showed a significant positive correlation between speech intelligibility,
and post-operative speech perception performance. This relationship has been demonstrated in other studies (Santarelli et al., 2008; Teoh et al., 2004; van Dijkhuizen et al., 2011). Speech intelligibility as a predictive factor of post-operative objective performance has been proposed by van Kijkhuizen and colleagues, who have suggested that the presence of intelligible speech pre-operatively is necessary for a successful outcome with a cochlear implant (van Dijkhuizen et al., 2011).

The impact of the use of sign language on speech perception in cochlear implant users is a contentious topic. Results from the current cohort of recipients showed that significantly poorer speech perception results were obtained from the group which used signing support as opposed to those who used purely oral language. There was no statistically significant difference in receptive vocabulary knowledge between the two groups, but significantly greater speech intelligibility was measured for the group using oral communication. These results do not necessarily suggest a causal relationship between the use of sign language and poor speech perception. It is possible that those participants who had less hearing during infancy used sign language out of necessity, and that concurrently the poorer hearing lead to the development of less intelligible speech. The poorer speech perception results observed for the signing support group may not therefore be caused by the fact that the group used sign language, but the fact that they used sign may be a result of the minimal hearing available during infancy.

**Gap Detection**

The perception of speech requires the discrimination of spectral and temporal cues. The spectral discrimination of the AEO-HL participants in the current study was unable to be determined preoperatively due to an insufficient sensation level. Temporal discrimination was measured via gap detection acoustically at 500Hz pre-operatively, with results showing that those participants with smaller gap detection thresholds achieved better speech perception results. Gap detection thresholds obtained from AEO-HL were significantly larger, and showed greater variability, than the gap detection thresholds obtained from a group of adults with an acquired hearing loss. The
literature has shown that at an equivalent sensation level, adults with an acquired hearing loss, or using a cochlear implant, demonstrate gap detection thresholds in the same range as adults with normal hearing. This would suggest that temporal resolution as measured by the ability to detect gaps, is not impaired by cochlear damage (Moore, 1993; Shannon, 1989). It may therefore not be the presence of a cochlear hearing loss, but rather the implications of an early-onset hearing loss, which result in poorer gap detection abilities for an individual. As gap detection abilities are adult-like by 6-12 months of age (Trehub et al., 1995), and studies have shown that poorer gap detection is measured in patients with a congenital compared to progressive hearing loss, and in patients who use sign language (Busby & Clark, 1999; Busby et al., 1992), the AEO-HL participants in the current study who demonstrated lower gap detection thresholds may have had more access to audition during infancy and may have more developed auditory pathways. It is possible that the measurement of temporal resolution via gap detection may be related to the amount of auditory input an individual with an early-onset hearing loss received as an infant.

Results for the AEO-HL in the current study suggest that a gap detection threshold less than 10ms is required in order to achieve a post-operative phoneme score greater than 50%, and that gap detection and speech intelligibility accounted for 59% of the post-operative variance in speech perception scores. Those recipients with lower gap detection thresholds and better speech intelligibility achieved greater speech perception scores. The regression equation enabled a calculated ‘predicted’ phoneme score to be plotted against the ‘actual’ phoneme score for this cohort of AEO-HL, demonstrating a significant correlation (r=0.769, p<0.001). This gives these two tests some predictive value for post-operative outcomes, as both measures appear related to the access and use of auditory input during infancy. Gap detection may indicate how well the auditory system has developed through early auditory input, while speech intelligibility is likely a measure of higher level cortical interconnections. These two measures may be thought of as the secondary factors, as suggested by Blamey and colleagues (1996), which are easily measured and likely to reflect the primary factors
of an individual which are not easily measured, such as the development of the auditory system (Blamey et al., 1996).

*Cortical Auditory Evoked Potentials*

Cortical auditory evoked potentials have been used extensively to measure the development of the auditory system in children (Sharma & Dorman, 2006; Sharma et al., 2002b; Wunderlich et al., 2006). Due to the P1 being generated by the primary auditory cortex, and the N1 by the secondary auditory cortical regions, both with individual time courses of development, the P1 and N1 provide reliable biomarkers of maturation in congenitally deaf children (Gilley et al., 2008; Godey et al., 2001; Sharma et al., 2015). Sharma and Dorman examined the P1 latency in 245 congenitally deaf children who received cochlear implants at different ages. They found that children who were implanted prior to 3½ years of age achieved normal P1 latencies, those implanted between 3½ and seven years had variable latencies, and those implanted after age seven had abnormal cortical latencies. A follow-up study compared 80 children with CIs against 41 typically hearing children. They found that the N1 began to emerge in normal hearing children at around 3-6 years of age, whereas detectability of the N1 in implanted children was variable. Very few children who received an implant after seven years of age developed an N1 response. Results from these studies suggest that the sensitive period for central auditory development diminishes significantly after seven years of age (Sharma et al., 2015; Sharma & Dorman, 2006; Sharma et al., 2002b). Sharma has suggested that once the sensitive period ends, there is high likelihood of de-coupling of the primary auditory cortical areas from surrounding higher-order cortices and cross-modal reorganization of secondary auditory cortical areas (Sharma et al., 2009).

The use of CAEPs as a measure of the maturity of the auditory system does not appear to be appropriate with adult cochlear implant recipients. The current study, and other recent studies, have shown that it is possible to measure CAEPs with typical morphologies and latencies from AEO-HL (Abraham et al., 2015; Lammers, Versnel, et al., 2015). Results from the current study showed that acoustic and electrically evoked
CAEPs with typical morphologies and latencies were able to be recorded from the majority of AEO-HL study participants. Although the measurement of an acoustic CAEP pre-operatively was severely limited by the available sensation level, the presence or absence of a response was not correlated with post-operative speech perception scores. Post-operatively, peak latencies did not change with implant experience and were not correlated with performance. These results are in contrast to much of the published literature in children, showing decreases in latencies with implant experience (Sharma et al., 2002a).

There are two possible explanations for the typical CAEP results seen in the current study. The first could be due to these particular participants having had more access to sound during early childhood, allowing sufficient maturation of the auditory pathway for a CAEP response to be recorded. This idea is supported by the fact that every participant in the current study had been able to develop some oral language; no participant had a congenital profound/total hearing loss. This concept is supported by a study by Sharma and Dorman who were able to record an N1 response in two children who were implanted after seven years of age. One of the children in their study had an acquired hearing loss, and one had a progressive hearing loss, suggesting that they both had sufficient hearing during early infancy for the waveform to mature (Sharma & Dorman, 2006).

The second explanation may relate to atypical waveform generators (Gordon et al., 2008; Lammers, Versnel, et al., 2015). Lammers and colleagues (2015) suggested that the ability to record a CAEP from an adult with minimal prior exposure to audition may be a reflection of the activation of a wider area of the auditory cortex due to a lack of synaptic pruning (Huttenlocher & Dabholkar, 1997) and decoupling of the auditory cortex (Kral & Sharma, 2012), rather than a specific region of cortical activation (Lammers, Versnel, et al., 2015).

The measurement of CAEP and eCAEP in this cohort of AEO-HL did not provide any predictive value for post-operative speech perception outcomes. While the duration of deafness before implantation has been shown to cause significant changes in post-
operative CAEP recordings in children (Sharma et al., 2015), we did not observe these changes in adults. Results from this study suggested that the measurement of CAEP may not be clinically useful for this patient population, and that a CAEP recording may not be a clinically useful tool for assessing the maturation of the auditory pathway in adults.

Non-Verbal IQ

Whilst a ten-year longitudinal study by Coletti and colleagues (2011) found that children who received their implants during infancy performed better on non-verbal IQ tests than children who received their implants at a later age, the same result was not observed with the adult participants in the current study. The participants in the current study did not receive their implants during infancy, yet showed no difference in non-verbal IQ scores when compared to adults with an acquired hearing loss. Coletti suggested that early auditory input is necessary for the development of higher cognitive functions utilising multisensory integration (Colletti et al., 2011). Pisoni suggested that children utilising oral communication develop better verbal rehearsal speeds than children using sign language, thereby giving them more efficient scanning and retrieval of information from short-term memory, and potentially better outcome measures with cochlear implants (Pisoni & Cleary, 2003). Results from both Coletti and Pisoni suggested that early auditory input was necessary for the development of non-verbal cognition, and verbal rehearsal skills. Pisoni suggested that those children who used sign language did not develop verbal rehearsal speeds to the same extent as oral children, as they did not have the same access to audition.

The AEO-HL in the current study who utilised some signing support for communication demonstrated significantly lower speech perception and speech intelligibility than those who relied on purely oral communication. Receptive vocabulary for the two groups, however, was equivalent. Results from the current study partly support the results obtained in children by Pisoni and colleagues, but not entirely (Pisoni & Cleary, 2003). As verbal rehearsal is linked to both speech perception and speech intelligibility, this may help to explain why participants in the current study demonstrated poorer
speech perception and intelligibility for the adults who rely on signing support as opposed to those who use oral language only. Verbal rehearsal skills have also been shown to be important for vocabulary, but there was no difference in receptive vocabulary for the AEO-HL who use oral language and those who use signing support. It is possible that the verbal rehearsal speed theory does not apply to the adult population in the same way as children. The measurement of verbal rehearsal speed and working memory for the current cohort of study participants may have provided additional information and explanations for the results obtained.

Verbal rehearsal speed has been shown to be a critical component in outcome measures typically used for cochlear implant recipients, such as speech perception, vocabulary, comprehension and speech intelligibility (Pisoni & Cleary, 2003). Pisoni and Cleary suggested that differences in verbal rehearsal strategies may be a key feature of the differences in post-operative cochlear implant outcomes, at least for children (Pisoni & Cleary, 2003). Whilst it certainly appeared to be an important investigation for children with a congenital hearing loss, the measurement of verbal rehearsal speed in a large number of adults with a significant early-onset hearing loss may provide more insight into the significance of such a measure in this population. It is possible that verbal rehearsal abilities may help to explain the poorer speech perception and speech intelligibility results observed in the adults who utilised some signing support for communication. The equivalent language scores obtained between both the oral and signing support group, however, may suggest that adults are able to use additional strategies in this domain, or may reflect the fact that the receptive language abilities of the group in general demonstrate a delay due to a common late diagnosis of hearing loss and the lack of a fluent signing language model during infancy. Verbal rehearsal may therefore be more sensitive at explaining vocabulary differences in children who receive an early implant and have the potential of achieving age appropriate language abilities.

It is possible that the participants in the current study, who all had some oral language, did have access to some audition during infancy. This may explain why the differences
in non-verbal IQ as suggested by Coletti were not observed, and that it was possible to measure a typical CAEP with latencies within normal limits in the current study (Colletti et al., 2011). Those participants with greater auditory access, however, and who utilised a purely oral language mode may have developed better temporal processing abilities and better speech intelligibility and speech perception, perhaps as a result of better verbal rehearsal speeds as a consequence of better development of the auditory system and secondary cortical links.

**Summary of Objective Measures:**

It has been well established in the literature that for an individual with an early-onset, significant hearing loss, the timing of cochlear implantation and the linguistic environment the individual is exposed to are critical for outcomes.

For children with significant hearing loss it has been shown that:

- Implantation prior to 12 months of age leads to optimal language outcomes
- Implantation prior to 3.5 years is necessary for the appropriate development of the auditory system as measured by typical latencies of the P1 and N1 waveform
- Children implanted early achieve better non-verbal cognitive abilities
- Children immersed in oral language develop better verbal rehearsal strategies, leading to better speech perception, speech intelligibility and language skills
- Children exposed to a fluent and rich signing culture can develop age appropriate vocabulary

(Colletti et al., 2011; Leigh et al., 2013; Mayberry et al., 2011; Pisoni & Cleary, 2003; Sharma & Dorman, 2006; Sharma et al., 2002a)
For the 29 adults in the current study with significant early-onset hearing loss who received late cochlear implants:

- All participants were the deaf children of hearing parents, and were likely not given the opportunity of early exposure to fluent sign language, therefore the group shows delays in receptive vocabulary
- All participants had some oral speech production, therefore it is probable that they had some access to audition during infancy
- This minimal access may have been enough for the maturation of a typical CAEP waveform with N1 and P2 peaks occurring at typical latencies
- No correlation was observed between non-verbal IQ and any demographic or outcome measure, apart from receptive vocabulary
- The group utilising some signing support for communication demonstrated significantly poorer speech perception and speech intelligibility; their lack of reliance on audition during early childhood may have impacted the development of cortical interconnections.
- Those participants with better gap detection abilities and better speech intelligibility achieved significantly better results on speech perception, which may have been due to more exposure to, and better utilisation of, audition during early childhood.

There exists a dynamic interplay of sensory stimuli, language and neurocognitive maturation during development which are all connected and support each other to provide the functional outcomes observed in cochlear implant recipients (Kral et al., 2016). Kral and colleagues (2016) suggested that the sensory input provided by a CI is related to stronger and more robust language skills, stronger language skills predict better neurocognitive outcomes, and neurocognitive functioning acts to support language. This positive feedback loop, whereby the use of spoken language consolidates the auditory skills that are required for the development of language in
the first place, plays a central role in cochlear implant outcomes in young children. When the development of the auditory system and this positive feedback loop has been disrupted by the presence of an early onset hearing loss, and a cochlear implant has not been provided until adulthood, the ability of the brain to make use of this incoming, degraded auditory signal is limited. Results of this current study suggested that the most important factor in predicting cochlear implant outcomes may be how much hearing an individual had during the critical period for maturation of the auditory system and language development, and more importantly, how they used this hearing. This proposal is supported by Kral and colleagues, who have suggested that late cochlear implantation only provides meaningful input if the auditory system has been able to mature due to previous acoustic hearing (Kral et al., 2016).

13.3 Subjective Outcome Measures

QoL instruments may be a more appropriate cochlear implant outcome measure for AEO-HL due to the low speech perception scores generally recorded for this patient group (Caposecco et al., 2012; Chee et al., 2004; Most et al., 2010). It has been suggested that such QoL measures may be more sensitive to the subtle benefits that can make a functional difference to the daily lives of AEO-HL, such as improved confidence, environmental sound awareness and improved personal safety due to the additional auditory cues provided by the cochlear implant (Caposecco et al., 2012; Chee et al., 2004). Studies typically report a lack of correlation between objective and subjective performance, with some patients obtaining minimal levels of open set speech perception, but reporting great device satisfaction and full time use (Hinderink et al., 2000; Most et al., 2010; Straatman et al., 2014; Zwolan et al., 1996). Results from the current study differed from previous published literature due to the significant correlation observed between speech perception performance and results from three of the four domains of the study questionnaire.

The study questionnaire developed for the purposes of this project was divided into four separate domains. The first explored the participant’s use of the device, the
second their perceived communication abilities, the third was confidence in different
listening situations, and the fourth was satisfaction with the device. Results for all four
domains were significantly lower than those obtained from a group of adults with an
acquired hearing loss (PostL2). Results for perceived communication ability were
correlated with post-operative speech perception performance for both the AEO-HL
and PostL2 group. Confidence and Device Satisfaction was also correlated with
performance for the AEO-HL group only. It is possible that it was due to the specific
nature of this questionnaire that significant correlations with performance were
measured for this AEO-HL patient cohort.

Whilst results from the current study showed that this group of 29 AEO-HL gained
significant benefit from their cochlear implants, eight participants achieved post-
operative CVC scores that were less than 20%. Device use was not correlated with
speech perception performance with many recipients scoring poorly for open set
speech perception still wearing their device more than eight hours a day. These results
are similar to those of Caposecco and colleagues, who found that 81% of their 38
participants wore their device more than eight hours per day (Caposecco et al., 2012),
and Craddock and colleagues who reported that 12 out of 20 AEO-HL wore their device
more than 10 hours per day despite no improvement in speech perception (Craddock
et al., 2016). It has been well documented that cochlear implant recipients with early
onset hearing loss tend to wear their cochlear implant full time, irrespective of
objective performance (Chee et al., 2004; Kaplan et al., 2003; Zwolan et al., 1996). It is
likely that the high rate of full-time device use amongst this population relates to the
quality of pre-operative counselling provided by the cochlear implant clinician.
Typically, much time is spent during appointments ensuring that each recipient
understands the limitations of cochlear implant technology, and has set their own
goals appropriately. It is often found that whilst post-operative expectations may
initially be somewhat unrealistic, by the end of the pre-operative period potential
recipients have tailored their goals to more achievable levels. If these goals, such as
being able to detect (but not comprehend) a call them from another room, or being
able to detect a car approaching or the front doorbell ringing are attained, then the
implant may be considered a success by the individual ensuring that it is worn consistently.

It is important to remember that each prospective cochlear implant candidate needs to be treated as an individual. For the three participants in the current study with a speech intelligibility rating of two (speech is very difficult to understand—only isolated words or phrases are intelligible), speech perception outcomes were low; 0%, 9%, and 19% respectively, but perceived benefit varied. The participant with zero open set speech perception became a non-user of their device, as they perceive no functional benefit and communicates via sign language. In contrast, the participant with a phoneme score of 9% uses a combination of lipreading and listening and has found the CIs input so positive that she is scheduled to receive a second side implant. Whilst the detection and discrimination of sound through the implant appeared to be similar for these two participants, their different reliance on audition for communication may have influenced their perception of the functional benefit.

Predicting the rate of device rejection has been investigated by Lammers and colleagues (2015) in a group of 48 adults with an onset of profound hearing loss prior to age two. The average post-operative phoneme score for the group was 25%, with 52% of participants obtaining open set speech perception, 27% obtaining sound detection, and 21% obtaining no benefit. The ten participants (21%) with no benefit from their cochlear implant went on to reject their devices and become non-users. (Lammers, van Zanten, et al., 2015). Four of these ten participants used sign language as their sole communication mode. The authors concluded that those participants who rejected their device appeared to have minimal residual hearing prior to cochlear implantation and were more likely to use sign language as their primary mode of communication. Rates of device non-use in the current study were much lower than the study by Lammers (Lammers, van Zanten, et al., 2015), with only one of the 29 rejecting their device by the 24-month post-operative mark. Supporting the ideas suggested by Lammers, this participant relied on sign language for communication and was rarely heard to use his voice to communicate. The rate of non-use in the current
study is similar to those reported by Craddock and colleagues, who reported that one participant became a non-user in their study of 20 adults with early-onset hearing loss (Craddock et al., 2016). It is possible that device rejection relates to the benefit perceived by an individual, which may have some basis in the preferred communication mode and reliance on audition by an individual.

Clinician expectations regarding future cochlear implant benefit has been used successfully to account for some degree of post-operative variance by Lammers and colleagues (2015). They found that the expectations of the CI team, pre-operative CVC scores and the presence or absence of residual hearing explained 69% of the variance in outcomes. A study by Craddock (2016), however, found no such relationship. The authors used the Adult Pre-Lingually Profoundly Deaf Implant Profile (APDIP) form to assess the levels of concern clinicians had for individual candidates regarding aspects of onset and duration of profound hearing loss, hearing aid use and mode of communication. Results for 20 adults with early-onset hearing loss showed no apparent relationship between the level of concern measured pre-operatively on the APDIP and hours of daily device use or speech perception outcomes (Craddock et al., 2016). Cochlear Implant teams often express reservations regarding possible outcomes for AEO-HL, but results from the two studies above, and the current study, would suggest that while these reservations should be discussed with the prospective patient to help guide post-operative expectations, these concerns may not actually correlate with post-operative performance, or perceived benefit.

The literature has shown that AEO-HL can demonstrate similar rates of improvement in QoL measures after cochlear implantation when compared to adults with an acquired hearing loss, even though the same improvements in speech perception are not observed (Peasgood et al., 2015; Straatman et al., 2014). No significant correlations between improvement in QoL and speech perception have been observed in the published literature, whether generic or health-specific QoL measures are used, or if a study specific questionnaire has been used (Chee et al., 2004; Kumar et al., 2016; Most et al., 2010). The questionnaire in the current study, however, was
designed by experienced clinicians and targeted specific domains regarding device use, perceived communication abilities, confidence, and device satisfaction. Significant correlations were observed between speech perception and perceived communication, confidence, and device satisfaction for the AEO-HL, which contrasts with previous publications. These results would suggest that questionnaire data can be used as an appropriate outcome measure for this patient population. In agreement with the published literature, no correlation was observed between outcomes on speech perception testing and device use (Craddock et al., 2016; Zwolan et al., 1996), or whether the recipient reported that they were ‘pleased they had received the CI’. Results from the current study, and the study by Lammers and colleagues (2015) suggest that the probability of a participant rejecting their cochlear implant is greater for those individuals with minimal residual hearing and who rely purely on sign language for communication (Lammers, van Zanten, et al., 2015).

It is apparent that traditional outcome measures may not always be sensitive enough to capture the advantages provided to this recipient group by cochlear implant technology. Many studies have suggested that qualitative measures, such as Quality of Life questionnaires, may be more sensitive at capturing functional benefit (Caposecco et al., 2012; Chee et al., 2004; Most et al., 2010). The degree to which a recipient may or may not report benefit from implantation may be based on their own expectations of the cochlear implant. This may be a direct result of the counselling provided to the individual in the pre-operative period. Appropriate and realistic counselling is required for this patient cohort to ensure that they understand the limitations of the technology and to minimise the rate of non-use (Kumar et al., 2016; Summerfield & Marshall, 2000).

13.4 Clinical Implications

Cochlear implantation has now become an accepted and widely utilised option for adults with a significant early-onset hearing loss. Results from the current study, and the published literature, have shown that the benefits of cochlear implantation for this
patient group are highly variable, and in general lower in terms of traditional speech perception measures, than those typically observed for adult recipients with an acquired hearing loss (Eisenberg, 1982; Kos et al., 2009). This variability creates challenges for cochlear implant clinicians, as it becomes difficult to predict the chance of improvement post-operatively, and therefore to counsel prospective patients appropriately.

Clinicians are often faced with unrealistic expectations from prospective recipients in the pre-operative period, such as the patient wanting to advance their career, meet a partner, or even learn to talk. Many potential recipients have friends with cochlear implants, some of whom may have had a successful outcome, or who may have developed oral language as a child. Young adults who have an early-onset hearing loss may also attend a cochlear implant clinic with their parents. In some cases, these families had attended the clinic 20 years earlier, and the decision may have been made by the family not to proceed with cochlear implantation at that time. These families may express feelings of regret, guilt or pressure as they come to terms with the fact that the expected outcomes for a two-year-old are very different to a 22-year-old. The prospective candidate needs to understand that a goal of learning to talk as an adult is unrealistic. It is vital that such expectations of communication change are explored in detail as it is important that the potential recipient understands the limitations of the cochlear implant technology, and has realistic goals to strive for post-operatively.

The literature suggests that early auditory deprivation may result in a compromised auditory system, leading to a poor post-operative outcome (Kral et al., 2006; Sharma & Dorman, 2006). In a recent paper, Kral has suggested that for an AEO-HL to achieve a successful CI outcome, they must have had access to enough sound during infancy to allow for at least rudimentary development of the auditory system (Kral et al., 2016). Results from the current study are consistent with this proposition that the most important factor in predicting post-operative speech perception results for AEO-HL may be the amount of residual hearing an individual had during the critical period for maturation of the auditory system and language development.
It can be difficult to establish an accurate early hearing history from adult candidates, however, making it problematic to determine the amount of residual hearing available during the critical period for communication development (Lammers, Versnel, et al., 2015). The patient themselves often do not know what their hearing thresholds were when they were diagnosed with a hearing loss. They may not even know how old they were when a diagnosis was first made, or when they received their first hearing aids. They may be unaware if they had received any auditory or speech therapy during childhood, or what early educational approach was taken. Due to the difficulties in obtaining an accurate hearing history, the quality of speech production and psychoacoustical measures of temporal processing may provide insight into an adults' prior hearing levels and use of audition. The combination of gap detection thresholds and speech intelligibility accounted for 59% of the variance in outcomes for the 18 AEO-HL who were able to complete the gap detection task. Gap detection measurements, however, requires the patient to have some residual acoustic hearing at 500Hz. From a practical point of view, the test also requires additional equipment and an extra pre-operative measurement session, placing additional burden on cochlear implant clinics. In a busy clinical setting, the rating of speech intelligibility requires minimal training and effort. The NTID rating scale used in the current study was significantly correlation with objective performance, accounting for 36% of the variance in post-operative outcome for the 29 AEO-HL study participants. Incorporating such a straightforward tool into clinical protocols may help clinicians ascertain how much exposure to audition an individual may have received during early childhood, thereby allowing them to facilitate the setting of realistic post-operative listening goals.

Objective performance on speech perception is not the only measurement of benefit for this patient cohort. While three of the four domains of the self-report questionnaire used in the current study did correlate with speech perception results, the majority of AEO-HL wore their device more than eight hours per day, enjoyed listening to music and reported the same levels of environmental sound awareness as cochlear implant recipients with an acquired hearing loss. Overall levels of device
satisfaction were high, suggesting that the way a recipient evaluates their decision to receive a cochlear implant is not only dependent on their performance on speech perception testing.

The incorporation of a speech intelligibility rating scale (as a proxy for the utilisation of residual hearing in childhood) into clinical protocols may provide insight into the prospective cochlear implant candidate’s early hearing history, allowing clinicians to predict post-operative performance with greater accuracy. It is important to consider that whether a recipient feels 'pleased' they received a cochlear implant may be based upon their own expectations, and may be irrespective of speech perception results. This highlights the importance of the clinician’s role in guiding a patient through the decision-making process, enabling adults with a significant early-onset hearing loss to make fully informed decisions based upon realistic expectations of outcomes.

13.5 Study Limitations

There were certain limitations in the current study, some inherent to clinical research, and some specific to this patient population. Small study numbers are a common issue in clinical research, especially when investigating specific patient populations. It is difficult to draw definitive conclusions from a study cohort of just 29 participants. This study may have had more power, and may have found additional significant relationships, if greater numbers had been available.

Another limitation of the current study, specific to this population, is the ambiguity surrounding the age at onset of significant hearing loss. While these participants all fit the criteria of having a ‘prelingual’ hearing loss in the clinical sense (classified at the Melbourne Cochlear Implant Clinic as the onset of severe hearing loss prior to 3 years of age) it was very difficult to quantify this statement based on patient report only. Adult cochlear implant recipients often had only a limited knowledge of their early hearing history. The specific age at diagnosis and severity of hearing loss at diagnosis was often not known to them. For this reason, the study participants were identified as
having a significant early-onset hearing loss, as opposed to categorising them as having a ‘prelingual’ hearing loss.

It is generally accepted within cochlear implant and hearing aid research, that two lists of monosyllabic words are used in each test condition, giving a total of 100 words. For the current study, however, only one list was used. The rationale for the reduced list was to minimise distress for the study participants. AEO-HL generally achieve only minimal open-set speech perception. While the majority of CI recipients with an acquired hearing loss also achieve only minimal speech perception pre-operatively, as a clinician it is possible to alleviate any pre-operative distress by explaining to the candidate that it is necessary to establish a baseline performance prior to surgery, in order to then measure improvements post-operatively. With AEO-HL, however, post-operative performance may still remain relatively low, and forcing a research participant to listen to 100 words that they may not be able to hear, becomes very distressing for the participant, and also for the clinician. The decision was made, therefore, to administer just one list of CVC words at each assessment point. The phoneme score was taken as the measure of choice as it removes the influence of language (that is, vocabulary) effects and provides a score out of 150 which is more statistically reliable than the word score out of 50.

The NTID speech intelligibility scale used in this study has benefits in that it requires minimal training, and is reported to have high inter- and intra-rater reliability. Two speech intelligibility ratings were obtained for each participant in this study. A percentage agreement and Kappa statistic were calculated to enable a comment to be made on the reliability of the rating. The percentage agreement of 55% and the Kappa Statistic of 0.48 obtained in the study were lower than anticipated. It is possible that training or greater clinician experience with this patient cohort may help to improve the reliability of the rating scale.

The current data set, specifically within the eCAEP section of the study, contained a proportion of missing data points. Whilst the majority of participants were happy and willing to attend pre-operative research test sessions, and the 3-month post-operative
test session, enthusiasm for extra appointments waned somewhat after this point. It became increasingly difficult to encourage participants to attend all appointments once the acute programming and rehabilitation stage at the MCIC had passed. Seven sessions within the eCAEP section were also unable to be used for analysis due to the presence of artefact. These issues prevented a full data set of results being available for analysis.

The gap detection and self-report analyses required a comparison to be made between the study cohort of AEO-HL and a group of adults with an acquired hearing loss. For the gap detection comparison, results were obtained from Cochlear Ltd, as part of a larger cohort study. It is unlikely that this subset of participants was completely representative of the entire population of CI recipients with an acquired hearing loss. The study at Cochlear Ltd was intensive and required long assessment sessions, therefore it is probable that those patients who agreed to be part of this study were highly motivated and enthusiastic. For the self-report chapter, recent cochlear implant recipients who best fit the inclusion criteria were chosen from the MCIC database. Questionnaires were then posted to these participants at home. It is possible that only motivated recipients, who were generally pleased with their cochlear implant, took the time to fill in the questionnaire and post it back to the clinic. There exists the possibility that the adults with acquired hearing loss who provided the comparison data for these studies do not, therefore, represent a true randomised control group, but rather a subset of motivated and enthusiastic cochlear implant recipients.

13.6 Future Directions

The use of a speech intelligibility rating scale, administered by audiologists, proved highly significant in terms of predicting post-operative speech perception for AEO-HL. The use of such a scale would be further validated if the results were compared with a full articulation and speech production assessment performed by a speech pathologist. A favourable correlation would enable the rating scale to be incorporated into a clinical setting with increased confidence.
Typical eCAEP waveform morphologies and latencies were observed for this cohort of AEO-HL CI recipients. Latencies of the peaks did not change with experience. These results are in contrast to those typically observed with young children, where peak latencies can be confidently used as a biomarker of maturation of the auditory system (Sharma et al., 2002b; Wunderlich et al., 2006). Typically, these studies involved congenitally profoundly deaf children, whose auditory systems are naïve to auditory input. The provision of a cochlear implant provides the stimulus required for the auditory system to mature, allowing peaks to emerge and latencies to change. The adults in the current study all had some oral language, indicating that they probably had some access to audition at an early age. The fact that typical eCAEPs were able to be recorded may suggest that their auditory systems had been provided with sufficient input for them to develop to a certain degree. A theoretical question exists as to what the eCAEP may look like for an adult CI recipient with a congenital profound/total hearing loss. It is possible that we may see the same type of maturation and latency shifts that are observed in congenitally deaf young children who receive cochlear implants. This question remains theoretical, however, as an adult candidate with a congenital total hearing loss is unlikely to be recommended to proceed with cochlear implantation.

Studies by Pisoni and Cleary (2003) have suggested that both working memory capacity and verbal rehearsal speed may help to explain some of the observed variance in post-operative cochlear implant measures in children (Pisoni & Cleary, 2003). The measurement of verbal rehearsal speed in adults with a significant early-onset hearing loss may help to provide support for the observation that many individuals who utilise sign language have poorer speech intelligibility and lower speech perception scores after cochlear implantation.

13.7 Summary

Cochlear Implant outcomes for adults with an acquired hearing loss and young children with a congenital hearing loss are typically very positive, with the majority of adult
patients re-establishing functional speech understanding and children developing the ability to listen and speak (Blamey et al., 1996; Geers et al., 2003). Communication outcomes for adults with an early-onset hearing loss, however, are generally poorer than those observed for adults with an acquired hearing loss, and display greater variability (Most et al., 2010; Zwolan et al., 1996).

The literature has demonstrated that the auditory system can only mature in the presence of a robust auditory signal, and it is for this reason that early cochlear implantation is generally recommended for children (Kral & Eggermont, 2007; Kral et al., 2016). Early cochlear implantation minimises the period of auditory deprivation and maximises post-operative outcomes (Sharma et al., 2005; Sharma et al., 2002b). Establishing an accurate early hearing history from AEO-HL in order to determine their access to audition during infancy, is typically problematic. The clinician involved must therefore attempt to establish whether any useable hearing was available to the patient during infancy, and more importantly, make some assumptions about whether the patient made use of this hearing, to be able to counsel these patients appropriately.

The current study attempted to design a series of tests to investigate aspects of auditory processing and spoken language skills in a group of 29 adults with a significant early-onset hearing loss who received cochlear implants at the MCIC. In addition to standard clinical protocols a series of language, gap detection, electrophysiological and subjective tests were conducted.

Results from the current study demonstrated that although speech perception results for the group of 29 AEO-HL were significantly lower than those obtained from a group of 576 adults with an acquired hearing loss, the group did show significant improvement in speech perception results post-operatively. Receptive language was not shown to be correlated with speech perception performance. Those participants who used a purely oral communication mode achieved better speech perception and demonstrated greater speech intelligibility than those who used some sign support. Speech intelligibility was strongly correlated with post-operative results, with those
participants with greater speech intelligibility achieving higher speech perception scores. Speech intelligibility accounted for 36% of the variance in performance.

Pre-operative gap detection thresholds using a 500Hz acoustic stimulus were successfully recorded in 18 AEO-HL. Gap detection thresholds were significantly larger, and displayed greater variability, than those measured in a group of 19 adults with an acquired hearing loss in study group PostL1. A significant correlation was observed between the 500Hz gap detection threshold in the ear to be implanted, and the post-operative CVC phoneme score for the AEO-HL group. Gap detection and speech intelligibility accounted for 59% of the variance in post-operative outcomes for this group of 18 participants. The regression equation provided a reasonable estimation of performance, as the correlation between the actual and predicted CVC phoneme score was highly significant.

The presence of an acoustic CAEP was not correlated with speech perception, speech intelligibility or gap detection abilities. Post-operatively, typical waveform morphologies and latencies were measured. Latencies did not change over time and were not correlated with speech perception results. Non-Verbal IQ was not correlated with speech perception performance or speech intelligibility, and there was no difference in IQ between those participants who used oral communication and those who used signing support. A significant correlation was observed between non-verbal IQ and receptive language.

AEO-HL scored significantly below the group of adults with an acquired hearing loss in the study group PostL2 on three out of four domains of the study questionnaire. For the specific questions regarding music enjoyment, environmental sound awareness or whether they were 'pleased' they had received the cochlear implant, however, there were no differences between the groups. There was no significant correlation between device use and speech perception for either group. Speech perception was positively correlated with perceived communication abilities for both groups. Confidence and Device Satisfaction was positively correlated with speech perception for the AEO-HL group.
Results from the current study do support:

- Hypothesis 2: That AEO-HL CI recipients with better speech intelligibility will achieve better scores on speech perception testing with their cochlear implant than those with poor intelligibility.
- Hypothesis 3: That CI recipients who use sign language will achieve lower scores on speech perception testing with their cochlear implant than those who use oral communication.
- Hypothesis 4: That temporal processing, as measured with acoustic stimuli pre-op using a gap detection test, will be poorer in AEO-HL than in adults with an acquired hearing loss.
- Hypothesis 5: That the relationship between gap detection and speech perception is such that shorter gap detection thresholds correlate with better speech perception scores.
- Hypothesis 9: That the use of a self-report outcome measure will accurately reflect post-operative results on speech perception testing.
- Hypothesis 10: That the results of the self-report outcome questionnaire will be lower for AEO-HL than those with a postlingual hearing loss.

Results from the current study do not support:

- Hypothesis 1: That CI recipients with better language scores on standardised tests will attain better scores on speech perception testing with their cochlear implant than those with poorer scores.
- Hypothesis 6: That those participants showing a measurable cortical waveform pre-operatively will achieve better speech perception scores post-operatively than those patients who did not have a measurable cortical response.
- Hypothesis 7: AEO-HL will demonstrate waveform latencies which are delayed when compared to published norms.
- Hypothesis 8: That those participants with lower scores on Raven’s Matrices, non-verbal test of IQ, will achieve lower post-operative speech perception scores than those who achieve higher scores.

Results from the current study suggest that the use of a speech intelligibility rating scale and the measurement of gap detection thresholds, may offer some insight into the availability, and appropriate use of acoustic cues during early childhood. Clinically, the incorporation of a speech intelligibility rating scale into clinic protocols requires no training, and minimal time. The highly significant correlation with post-operative performance may help guide patient expectations during the pre-operative counselling process. It is important to remember that while objective speech perception results are typically the outcome measure utilised in cochlear implant programmes worldwide, the self-report questionnaire measure used in the current study showed that regardless of speech perception scores, most AEO-HL CI recipients still chose to wear their speech processor all day, enjoyed listening to music, were able to identify environmental sounds and were very pleased they had received a cochlear implant. While these are not the traditional outcome measures used, they are no less important to a hearing-impaired person who may not have had the opportunity to connect with their environment and enjoy music prior to implantation, and these factors may be appropriate and achievable expectations to discuss in the pre-operative counselling process.
14 CONCLUSIONS

In summary, while the speech perception results obtained from adult cochlear implant recipients with a significant early-onset hearing loss are poorer, and more variable than those seen for recipients with an acquired hearing loss, the cohort in the current study still demonstrated significant benefit from their implants. Improvements in speech perception were observed for the majority of the group, with results significantly correlated with speech intelligibility and gap detection thresholds. These factors may indicate the availability and use of acoustic hearing during infancy, reflecting the appropriate maturation of the auditory system during development. Most importantly, the majority of recipients in the study cohort wear their speech processor all day, and are very pleased that they received a cochlear implant.
15 LIST OF ASSOCIATED PUBLICATIONS AND PRESENTATIONS

Departmental Seminar July 2010

Departmental Seminar July 2011

Departmental Seminar August 2012

RHD Colloquium October 2014

Receptive Language as a Predictor of Cochlear Implant Outcome for Prelingually deaf Adults. Alexandra Rouset, Richard Dowell, Jaime Leigh – Published IJA 2016

Music Appreciation and Music Listening in pre- and postlingually deaf cochlear implant recipients. Michelle Moran, Alexandra Rouset & Valerie Looi– Published IJA 2016

The Use of a Study Questionnaire to Evaluate Performance of Prelingually Deaf Adult Cochlear Implant Recipients. Alexandra Rouset & Richard Dowell – Submitted IJA June 2016


Oral Presentation, Audiological Society of Australia Conference Adelaide, ‘Receptive Language Skills for Prelingually Deaf Adults who Proceed with the Cochlear Implant‘ July 2012, presented by my colleague, Jaime Leigh

Poster Presentation, Audiology Australia National Conference May 2016 ‘The Use of a Study Questionnaire to Evaluate Performance of Prelingually Deaf Adult Cochlear Implant Recipients’

Oral Presentation, Audiology Australia National Conference May 2016’ Speech Intelligibility as a Predictor of Cochlear Implant Outcome for Adults with Prelingual Hearing Loss’

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REFERENCES


## APPENDIX A – COCHLEAR IMPLANT AUDIOLOGICAL EVALUATION PROTOCOL FOR ADULTS

### Cochlear Implant Audiological Evaluation

<table>
<thead>
<tr>
<th>Patient</th>
<th>Audiology</th>
<th>Documentation</th>
</tr>
</thead>
<tbody>
<tr>
<td>Follow up questions from previous discussion appointment</td>
<td>Dorsal examination</td>
<td>Audogram scanned into patient’s DHR</td>
</tr>
<tr>
<td>Explain need for assessments to assess functionality of each hearing pathway</td>
<td>Measure air and bone conduction hearing thresholds</td>
<td>Note speech perception scores in patient’s DHR</td>
</tr>
<tr>
<td>Instruct patient for audiometric measures</td>
<td>Custom HA function with HIT and PDM</td>
<td></td>
</tr>
<tr>
<td>Instruct patient for speech perception testing</td>
<td>Perform aided speech perception measures</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Administer CNC Words in each ear</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Binaurally</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Administer SRT if phoneme scores &gt;30%</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Administer MAP Words A &amp; V</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Phoneme accuracy &gt;30%</td>
<td></td>
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<td></td>
<td></td>
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<tr>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Explain audiogram and speech perception results</td>
<td>Discuss relative merits of HA and CI given results at hand</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Explain further course of action and seek patient’s choice</td>
<td>Note further course of action in patient’s DHR</td>
</tr>
</tbody>
</table>
APPENDIX B: SOUND LEVELS

LEVELS OF LOUDNESS

Painfully Loud

Extremely Uncomfortable

Uncomfortably Loud

Loud, But OK

Comfortable, But Slightly Loud

Comfortable

Comfortable, but Slightly Soft

Soft

Very Soft
APPENDIX C - STUDY QUESTIONNAIRE

Your Name: ________________________________
Date: ________________________________
Date of cochlear implant: ________________________________

Use of Cochlear Implant

1. How many hours per day do you wear the device?
   - More than 8 hours a day
   - 4 - 8 hours per day
   - 1 – 4 hours per day
   - Less than 1 hour a day
   - Never

2. Do you wear a hearing aid on your opposite ear?
   - More than 8 hours a day
   - 4 - 8 hours per day
   - 1 – 4 hours per day
   - Less than 1 hour a day
   - Never

Communication

3. Before you had your cochlear implant, how did you mainly communicate?
   - Listening
   - Listening & lipreading
   - Lipreading
   - Sign Language
   - Written Notes

4. With your cochlear implant, how do you mainly communicate?
   - Listening
   - Listening & lipreading
   - Lipreading
   - Sign Language
   - Written Notes
5. Do you feel that the cochlear implant has made it easier for you to communicate?
- Much easier
- A little more difficult
- A little bit easier
- Much more difficult
- No difference

6. How do you rate your ability to understand speech when combined with lipreading?
- I understand everything
- I understand some things
- I understand most things
- I can’t understand anything
- I understand about half

7. How do you rate your ability to understand speech with no lipreading?
- I understand everything
- I understand some things
- I understand most things
- I can’t understand anything
- I understand about half

8. How do you rate your ability to follow conversation in a group?
- I understand everything
- I understand some things
- I understand most things
- I can’t understand anything
- I understand about half

9. How do you rate your ability to hear and recognise environmental sounds?
- I recognise everything
- I recognise some things
- I recognise most things
- I can’t recognise anything
- I recognise about half

10. Do you enjoy listening to music with your cochlear implant?
- Very enjoyable
- Slightly unpleasant
- Slightly enjoyable
- Not at all
- Neutral
11. Are you able to hear on the telephone with your cochlear implant?
Yes, with no difficulty  [ ]  Not really; very difficult  [ ]
Yes, with slight difficulty  [ ]  Not at all  [ ]
Only with familiar speakers  [ ]

Confidence

12. Do you feel more confident in a group when wearing your speech processor?
Much more confident  [ ]  Slightly less confident  [ ]
Slightly more confident  [ ]  Much less confident  [ ]
Neutral  [ ]

13. Do you feel more confident with unfamiliar people when wearing your speech processor?
Much more confident  [ ]  Slightly less confident  [ ]
Slightly more confident  [ ]  Much less confident  [ ]
Neutral  [ ]

Satisfaction with Device

14. How satisfied are you with your cochlear implant?
Very Satisfied  [ ]  Somewhat dissatisfied  [ ]
Somewhat Satisfied  [ ]  Very dissatisfied  [ ]
Neutral  [ ]

15. Are you pleased that you had the cochlear implant?
Very Pleased  [ ]  Somewhat displeased  [ ]
Somewhat Pleased  [ ]  Very displeased  [ ]
Neutral  [ ]
APPENDIX D: COMPARISON OF STUDY QUESTIONNAIRE RESULTS

Comparison of study questionnaire results at 12 and 24 months post-operatively for the AEO-HL Study group – Statistical Analysis

<table>
<thead>
<tr>
<th>Q1</th>
<th>Q2</th>
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<tbody>
<tr>
<td>Mann Whitney</td>
<td>Mann Whitney</td>
</tr>
<tr>
<td>W = 375</td>
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<th>Q4</th>
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<td>p = 0.1787</td>
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<td>W = 312</td>
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<td>W = 307.5</td>
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<td>W = 363</td>
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<th>Q12</th>
</tr>
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<tbody>
<tr>
<td>Mann Whitney</td>
<td>Mann Whitney</td>
</tr>
<tr>
<td>W = 299</td>
<td>W = 320.5</td>
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<tr>
<td>p = 0.3834</td>
<td>p = 0.9390</td>
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<table>
<thead>
<tr>
<th>Q13</th>
<th>Q14</th>
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<tbody>
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<td>Mann Whitney</td>
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</tr>
<tr>
<td>W = 328</td>
<td>W = 340</td>
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<tr>
<td>p = 0.6539</td>
<td>p = 0.4691</td>
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<td>Mann Whitney</td>
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<tr>
<td>W = 362</td>
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<td>p = 0.9775</td>
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Comparison of study questionnaires by individual question, between AEO-HL study group and PostL2 group.

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<tr>
<th>Q1</th>
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<td>1 sample T Test</td>
<td>t = -3.19</td>
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<th>Q4</th>
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<td>p &lt; 0.001</td>
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<td>Mann Whitney</td>
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<td>p = 0.3556</td>
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<tr>
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<tr>
<td></td>
<td>t = -3.33</td>
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<tr>
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<td>p = 0.0001</td>
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<table>
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<tbody>
<tr>
<td>1 sample T Test</td>
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Author/s:  
Rousset, Alexandra May

Title:  
Outcomes and predictive factors with cochlear implants for adults with a significant, early-onset hearing loss

Date:  
2017

Persistent Link:  
http://hdl.handle.net/11343/194904

File Description:  
Outcomes and predictive factors with cochlear implants for adults with a significant, early-onset hearing loss

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