Mild and moderate congenital hearing loss in childhood: trends and associations with language outcomes

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Abstract

**Background:** Children born with mild and moderate bilateral hearing loss experience poorer language outcomes than peers without hearing loss. Following implementation of universal newborn hearing screening (UNHS) in the early 2000s, most children are now fitted with hearing aids very early, even without firm evidence of efficacy. Filling this knowledge gap would support clinicians and families to make appropriate decisions for affected children.

**Aims:** In population-based samples of children born with mild and moderate bilateral hearing loss in two age groups, I aimed to describe the: 1) language and child wellbeing outcomes at 5-8 years of age examined both by system of hearing loss detection and age at detection; 2) relationship between measures of hearing ability (aided and unaided) and speech/language outcomes at 5-7 years of age; and 3) expressive vocabulary of children at age 1-3 years, with comparison to same-aged children without hearing loss.

**Methods:** All participants were drawn from population-based studies or databanks with comparable measures and demographic composition in Victoria and New South Wales. In Aim 1, 21 directly assessed children (5-7 years) were analysed with 125 children (5-8 years) from historical hearing loss cohorts and 1217 children (7 years) without known hearing loss. For Aim 2, hearing aid data were obtained for 19 directly assessed children. In Aim 3, questionnaire data were compared between 20 children (1-3 years) with and 1711 children (2 years) without known hearing loss. Language was assessed or parent-reported, with additional data collected by parent questionnaire. Aim 1 was analysed using unadjusted and adjusted regression models, with potential confounders identified *a priori*. For Aims 2 and 3, analyses were descriptive and exploratory.

**Results:** Outcomes for 5-7 year olds with moderate loss improved across detection systems spanning opportunistic through to UNHS (mean expressive language p for trend .05, receptive vocabulary p for trend .06) and were better when diagnosed before age 6 months. Children with mild loss did not show
similar benefit, despite trends toward earlier diagnosis and hearing aid fitting. Children with mild-moderate loss had poorer expressive language than children without loss (adjusted mean difference -8.9 points, 95% CI -14.7 to -3.1). Audibility measures correlated with unaided hearing (r=-.60 to -.79, p=.02 to <.001) and speech recognition ability (r=-.63 to -.73, p=.04 to .01). Unaided and aided hearing measures did not show associations with speech and language. On average 1-3 year olds with, versus without, hearing loss had lower expressive vocabulary raw scores even though older at assessment. Language of children with mild loss was on average 9 months behind expectations (SD 5.6 months), moderate loss on average 6.6 months behind (SD 6.7 months).

**Conclusions:** Children with mild and moderate hearing loss show early language delays that persist into primary school. Children with moderate loss showed some benefit from earlier detection and fitting with hearing aids. Children with mild loss did not, which is concerning given the current practice of earlier hearing aid fitting. A population-based trial of hearing aids for mild losses would clarify the benefits versus costs of this approach.
Declaration

This is to certify that:

i. the thesis comprises only my original work towards the PhD except where indicated in the Preface;

ii. due acknowledgement has been made in the text to all other material used;

iii. the thesis is fewer than 100 000 words in length, exclusive of tables, maps, bibliographies and appendices.

Peter Carew
Preface
This thesis comprises my original work and contains no material previously published or written by another person, except where due reference has been made.

Activities undertaken to obtain the described data were nested within the broader activities of the Victorian Childhood Hearing Impairment Longitudinal Databank (VicCHILD). This ongoing study is housed at the Centre for Community Child Health at the Murdoch Children’s Research Institute, Melbourne, Australia. The VicCHILD team, including the PhD candidate, was responsible for all aspects of the databank. This included study design, participant recruitment, collecting data and pursuing linkage with other data sources.

Of particular relevance to the contained body of work, I was responsible for the development of data collection protocols covering two age ranges: 1-3 years and 5 to 7 years. I collected over 70% of the included data and performed all statistical analyses under the guidance of my supervisory team and a member of my PhD advisory committee, biostatistician Dr Fiona Mensah.

To examine secular trends in language outcomes of children with hearing loss at age 5 to 7 years, I had access to datasets outside of VicCHILD. Namely, this comprised full access to both the Statewide Comparisons of Outcomes (SCOUT) study and the Children with Hearing Impairment in Victoria Study (CHIVOS), each of which I was a named study investigator. For comparison to language outcomes of children without hearing loss, I received access to data from the longitudinal Early Language in Victoria Study (ELVS).
Publications

During my candidature, I contributed to the following publications. Publication 1 arose directly from my PhD data. It addresses the first aim of my thesis and appears in Chapter 5. Publications 2-4 represent work conducted within the Murdoch Children’s Research Institute during my candidature but did not directly relate to my thesis; thus, they appear in Appendix I-K.


Acknowledgements

During any PhD study there would surely be a multitude of people who have contributed and deserve acknowledgement. The truly unique aspect of each PhD is the way in which these people have helped, supported, facilitated and nurtured. I would like to specifically acknowledge the following individuals and groups. Without you, I would not have been able to achieve all that this thesis represents.

Firstly, I thank all the children and families who have participated in the three arms of this project for making my PhD possible. These families were often involved with Australian Hearing for hearing rehabilitation and I thank the many clinical and support staff that readily provided information without delay, despite it adding to their daily demands. Likewise, staff from various hearing loss research projects housed at the Murdoch Children’s Research Institute over a 20-year period have, through their work with recruitment, assessment and on-going management, made this PhD possible.

Secondly, thank you to my supervisors. You have each provided me with such unique support informed by your diverse backgrounds and experiences. Melissa, thank you on many fronts. Thanks for the opportunity to work on this exciting project, for generously sharing your incredible knowledge across many fields, for persisting with me on my journey towards clearer academic writing and for always referencing me to the bigger picture. Traci, your supervision at a geographic distance has not compromised your support of me through many foreign concepts and challenges. Thank you for being an ever-available sounding board for my ideas and tangents. Zeffie, thank you for your encouragement, your ability to consistently steady me along the way and for being the wealth of information you modestly deny to be. Gary, thank you for your ever-present support and pragmatic approach that has helped me keep everything in context.

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from a diverse group of researchers. The Centre for Community Child Health at the Murdoch Children’s Research Institute and the Department of Audiology and Speech Pathology at the University of Melbourne have physically housed me and provided wonderful environments full of passionate and intelligent individuals who inspire my work. I wish to acknowledge several colleagues and friends in particular for their support. Fiona, for sharing your wealth of statistical knowledge and your thoughtful input on my analyses. Laura, Shannon, Alanna and Nardia, for experiencing alongside me the peaks and troughs of PhD candidature. Donella, Kelley and Dani, for being friends and colleagues who just ‘get it’ and who provided unwavering support. Carina, for being chief cheerleader from afar, and Billy, for your ability to save my tables and margins from myself.

Finally, I must thank my family. To my parents, Ginny and Paul, thank you for accepting and supporting my decision to return to study. You have never discouraged me from challenging myself and I deeply appreciate both the stated and implicit encouragement you give me. To Susan and Guy, thank you also for your support and understanding.
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**Abbreviations**

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<th>Abbreviation</th>
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<tbody>
<tr>
<td>3FAHL</td>
<td>Three-frequency average hearing loss</td>
</tr>
<tr>
<td>4FAHL</td>
<td>Four-frequency average hearing loss</td>
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<tr>
<td>AABR</td>
<td>Automated Auditory Brainstem Response</td>
</tr>
<tr>
<td>ABR</td>
<td>Auditory Brainstem Response</td>
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<tr>
<td>CELF-4</td>
<td>Clinical Evaluation of Language Fundamentals – Fourth Edition</td>
</tr>
<tr>
<td>CHIVOS</td>
<td>Children with Hearing Impairment in Victoria Outcome Study</td>
</tr>
<tr>
<td>dB HL</td>
<td>Decibels Hearing Level</td>
</tr>
<tr>
<td>dB SPL</td>
<td>Decibels Sound Pressure Level</td>
</tr>
<tr>
<td>dBA</td>
<td>A-weighted decibels</td>
</tr>
<tr>
<td>dBnHL</td>
<td>Decibels normalised hearing level</td>
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<tr>
<td>ELVS</td>
<td>Early Language in Victoria Study</td>
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<tr>
<td>LiSN-S</td>
<td>Listening in Spatialised Noise – Sentences test</td>
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<tr>
<td>LOCHI</td>
<td>Longitudinal Outcomes of Children with Hearing Impairment</td>
</tr>
<tr>
<td>MCDI</td>
<td>MacArthur Bates Communicative Development Inventory</td>
</tr>
<tr>
<td>NIH</td>
<td>National Institutes of Health</td>
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<tr>
<td>NPVT</td>
<td>NIH-adaptive Picture Vocabulary Test</td>
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<tr>
<td>OAE</td>
<td>Otoacoustic Emissions</td>
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<tr>
<td>OCHL</td>
<td>Outcomes of Children with Hearing Loss</td>
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<tr>
<td>PEACH</td>
<td>Parents’ Evaluation of Aural/Oral Performance of Children</td>
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<td>PTA</td>
<td>Pure tone average</td>
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<td>SCOUT</td>
<td>Statewide Comparison of Outcomes</td>
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<td>SEIFA</td>
<td>Socio-Economic Indexes for Areas</td>
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<tr>
<td>SII</td>
<td>Speech Intelligibility Index</td>
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<tr>
<td>SRT</td>
<td>Speech Reception Threshold</td>
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<td>UNHS</td>
<td>Universal newborn hearing screening</td>
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<tr>
<td>USPSTF</td>
<td>United States Preventative Services Task Force</td>
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<tr>
<td>VicCHILD</td>
<td>Victorian Childhood Hearing Impairment Longitudinal Databank</td>
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<td>VIHSP</td>
<td>Victorian Infant Hearing Screening Program</td>
</tr>
<tr>
<td>WHO</td>
<td>World Health Organisation</td>
</tr>
<tr>
<td>WNV</td>
<td>Wechsler Nonverbal Scale of Ability</td>
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1. Introduction

Children born with mild and moderate bilateral hearing loss on average experience poorer language outcomes compared to their peers without hearing loss. Due to universal newborn hearing screening, these less severe losses are now routinely detected earlier than was historically possible. As a result, there has been a rapid change in practice such that most children detected with mild loss are now fitted with hearing aids very early – despite lack of firm evidence of benefit. Compared to more marked losses, less is known about how this earlier detection has impacted outcomes for children with mild and moderate loss. Furthermore, there are limited population-level data on how measures of hearing associate with language outcomes and at what age these poorer than expected outcomes become apparent. Knowledge both about language outcomes and impact of early aiding could help support clinicians and families to make appropriate decisions for impacted children.

Therefore, in population-based samples of children born with mild and moderate bilateral hearing loss in two age groups, I aimed to describe the: 1) language and child wellbeing outcomes at 5-8 years of age examined both by system of hearing loss detection and age at detection; 2) relationship between measures of hearing ability (aided and unaided) and speech/language outcomes at 5-7 years of age; and 3) expressive vocabulary of children at age 1-3 years, with comparison to same-aged children without hearing loss.

1.1 Research plan

This study examined speech and language outcomes in children born with mild or moderate bilateral congenital hearing loss. Data were collected for two age groups: children aged 5-7 years and 1-3 years. The older children completed a direct assessment of language ability (approx. 120 minutes) and their primary caregivers completed surveys on their child and family (approx. 20 minutes). At 1-3 years of age parents completed surveys which included a report of their
child's expressive language skills (approx. 20 minutes). These data collected for this thesis were integrated and compared with existing data sourced from multiple harmonised population-based cohorts of children both with and without hearing loss.

1.2 Chapter summary

Chapter 2 is a literature review. It starts by explaining hearing and hearing loss, the different methods used to quantify losses and how hearing loss detection has changed over time. The role of Universal Newborn Hearing Screening is highlighted in the context of how mild and moderate losses are now detected early. Language development in the presence of hearing loss is then considered. Theories of language development are outlined, leading into a comprehensive overview of language outcomes of children with mild and moderate hearing loss. Management of these hearing losses is also considered. Throughout, knowledge gaps are highlighted, setting the scene for this PhD study.

Chapter 3 outlines the rationale for this study, including the individual study aims and hypotheses.

Chapter 4 reports the study methodology. A brief overview of the contributing historical cohorts provides context for the reader on studies that provided data for comparative purposes. An in-depth description of the cohort unique to this PhD study leads into a detailed description of the novel study design, procedures and measures used in this thesis. Specific procedural details relevant to Aim 2’s hearing aid focus are clearly delineated from the common methods used for both Aims 1 and 2. The statistical methods and analysis plan for each aim rounds out this procedural chapter.

Chapter 5 is the first of three results chapters. It reports the results of Aim 1, describing the outcomes of children born with mild and moderate hearing loss exposed to 4 different systems of hearing loss detection over a period of two decades during which the average age of detection and fitting of amplification fell markedly. The results have been published in Child: care, health and
development; therefore, this published paper forms the chapter. The results section outlines the sample characteristics, followed by adjusted regression models to examine the association between language outcomes and system of diagnosis and age at diagnosis in children aged 5-7 years. These outcomes are compared to those achieved by children without hearing loss in a large cohort drawn from the same population.

Chapter 6 reports the results of Aim 2, examining the relationships among measures of hearing (aided audibility, unaided hearing and speech recognition ability), and then their associations with speech/language outcomes. This aim used a restricted sample of children aged 5-7 years from Aim 1, comprising those detected under the current hearing screening system and wearing hearing aids. As these results have not been published, this is written as a traditional chapter. The chapter begins by describing the sample characteristics and their speech and language ability. Correlations between aided audibility and unaided hearing and speech recognition ability are presented, followed by correlation and linear regression analyses of these measures of hearing with speech/language outcomes. Observed variation in the participants hearing and language assessment outcomes is explored.

Chapter 7 is the final results chapter. It reports the results of Aim 3, which focuses on parent-reported expressive vocabulary outcomes in children aged 1-3 years. As these results have not been published, this is written as a traditional chapter. It begins by describing the sample characteristics of these new cohorts of children with and without hearing loss, followed by analysis of the effect of age at assessment on parent-reported expressive vocabulary outcomes. Next the patterns in expressive vocabulary development are examined, followed by exploration of associations between parent-reported audibility and expressive vocabulary outcomes.

Chapter 8 is the final chapter of this thesis. It critically discusses the results of the study. It begins by outlining the principal findings of each aim, before discussing the overall strengths and limitations of the study. Interpretation of
the study results with regards to the posed hypotheses is followed by a discussion of how they fit with previous literature. The findings from Aim 1 are further elaborated upon following a brief discussion in the published paper. The unpublished Aim 2 and 3 findings are interpreted in depth for the first time. Implications of the findings at a population level and for clinical practice are outlined, followed by conclusions and an outline of future research.
2. Review of the literature

2.1 Overview

This chapter begins by exploring hearing and congenital hearing loss, to orient the reader to the quantification, changes in detection, and impact of hearing loss. The chapter then broaches language development in the presence of hearing loss, to highlight what impact this sensory deficit has on developmental outcomes. This leads into an exploration of the literature behind management decisions for hearing loss, highlighting the evidence gaps for the current strategy of early hearing aid fitting for milder losses. The chapter ends with a summary to lead the reader into the specific aims of the PhD thesis. More detail of the distinct sections of this literature review are below.

The first section of this review comprises an overview of hearing and hearing loss includes how hearing loss is quantified and how congenital losses have been detected over time. An exploration of the population health principles that influence current hearing loss detection methods illustrates why we now have early detection of hearing loss of all degrees. The term “milder” hearing loss is introduced, which is the focus of this thesis and is used to cover both mild and moderate bilateral losses. Specific literature on the detection of milder losses is accompanied by an overview of its prevalence in the population and the broad impacts this degree of loss can have on affected individuals.

The second section of this review focuses on language outcomes in children with milder hearing loss. The aim here is to highlight where deficits in development are thought to occur. A summary of typical language development and the role of hearing leads into a comprehensive review of the literature on language outcomes at two specific periods of life: the early primary school years and the preschool years, since this is when differential outcomes of treatment are likely to emerge. Following on, management of milder losses covers current trends towards earlier amplification. This section considers the strength of the evidence that this management is effective.
2.2 Hearing and hearing loss

This section reviews how we hear, what the different types of hearing loss are, and how hearing loss can occur. Differences across the literature in quantifying hearing loss, including the various methods of measuring hearing loss impact, illustrate the diversity that exists in describing both hearing loss and its impact.

2.2.1 Process of hearing

The ability to hear is dependent on a precisely organized sequence of processes and events. The process begins at the most peripheral point of the auditory pathway, with sound received at the pinna funneled down the external auditory canal towards the eardrum (see Figure 2.1, modified from Cole & Flexer (2007)1). The eardrum and ossicular chain of the middle ear are responsible for both translating and magnifying this vibration of air particles into mechanical energy. Mechanical vibrations transmitted by the stapes footplate create traveling waves within the fluid-filled cochlea.2 The cochlea is innervated by sensory hair cells arranged in a systematic fashion, with cells at the basal end coding for high frequencies and cells at the apical end coding low frequencies. These hair cells are stimulated as a result of the travelling wave moving from base to apex and synapse with terminal dendrites connecting to the spiral ganglion cells in the cochlea, transforming mechanical energy into electrical impulses.3 Neural conduction of the impulses reach the cochlear nucleus of the brainstem, travelling onwards along synapses formed with the nuclei of the inferior colliculus, medial geniculate body of the thalamus and reach the auditory cortex of the temporal lobe.3 The auditory centre of the brain is responsible for translating these electrical signals into a recognizable format, generating a perception of sound that can then be transformed and acted upon.
The process illustrated above must occur without disruption in order to deliver normal hearing ability. An impairment to hearing ability, or hearing loss, is the product of dysfunction along at least one point of this auditory pathway. There are different types of hearing loss. These losses can result from different aetiological causes, both temporary and permanent, with varying functional impact for the individual.

2.2.2 Types of hearing loss
Two main types of hearing loss exist: a conductive loss which has pathological roots in the outer or middle ear, and a sensorineural or sensory loss which has a
site of lesion in the cochlea. A mixed hearing loss occurs when both a sensorineural and conductive component are present. Auditory neuropathy, a rarer form of hearing loss, exists and is estimated to impact 7-10% of those born with a hearing loss.\textsuperscript{4, 5} It is characterized by normal cochlear function, yet disordered afferent neural conduction of electrical signals along the auditory pathway.\textsuperscript{6} When considering the two main types, conductive hearing losses are often temporary, dependent on the pathological cause. Sensorineural losses are most often permanent.

### 2.2.3 Causes of hearing loss

Hearing loss can result from numerous causes that may be broadly grouped into two categories: congenital and acquired. The cause of congenital hearing loss may be genetic (such as single-gene mutations) and/or non-genetic (such as infections, a result of birth issues, or a consequence of treatment received in the newborn period with or without genetic predisposition).\textsuperscript{7} Acquired hearing loss can occur at any stage during life, from infectious, accidental or ageing processes. Hearing loss can also have a late onset, with the incidental cause being an event from earlier in life (e.g. infection) or a genetic process that manifests with such a phenotype. Hearing losses can progress, in that hearing loss may worsen over time. Unlike conductive losses, very few sensorineural losses improve over time.

Approximately 50 to 60% of permanent hearing losses identified in children are thought to have a genetic cause. Inheritance is commonly autosomal recessive, with both parents carrying the gene for hearing loss and often with no extended family history of gene expression.\textsuperscript{8} Syndromic causes account for approximately 30% of children with genetically derived loss, with the hearing loss being one of a constellation of defining clinical features. Such hearing losses may affect one or both ears (i.e. a unilateral or bilateral loss), may progress in severity and may be present at birth or have a later onset. The remaining 70% of genetic causes of hearing loss are non-syndromic. The most common example is a mutation to the gene GJB2, responsible for producing a protein labeled Connexin 26. Hearing loss resulting from Connexin 26 is usually measurable from birth, occurs bilaterally
and is audiometrically stable across many years; it ranges in severity from mild to profound.\(^7\)

Often termed “environmental”, congenital causes of hearing loss to which some children may be particularly genetically susceptible include maternal infections during pregnancy (e.g. Cytomegalovirus infection, CMV) and perinatal causes, including birth hypoxia, severe jaundice and ototoxic medications.\(^7\) to \(^10\) Such identifiable factors are termed risk factors for hearing loss.

For a large number of children diagnosed with hearing loss a specific aetiology is not determined, often due to a lack of investigations. Many of these cases are likely caused by unidentified or unknown non-syndromic genetic anomalies and/or congenital infections for which diagnostic testing was not performed during a narrow diagnostic window (especially relevant to CMV infection).

### 2.2.4 Quantifying hearing loss

Hearing loss is usually measured by obtaining reliable behavioural responses to calibrated stimuli of known frequency and intensity. The subject indicates the softest sound they can detect, which is defined as their threshold. Those too young or unable to comply with this procedure can alternately be assessed using non-volitional methods. Measuring elicited physiological processes known to occur within the inner ear and along the auditory pathway allows estimation of expected behavioural hearing thresholds. Irrespective of the method used, both approaches have the same goal: to define both the type and degree of any hearing loss present.

When sound detection thresholds fall outside what is considered the normal range, they are typically categorised into “degrees” of hearing loss. A specified range of sound sensitivity, measured in units of decibels hearing level (dB HL), defines each degree of loss. As all categorisation systems are arbitrary, many exist. Figure 2.2 displays degrees of hearing loss adopted by various audiology and health organisations around the world. All include mild, moderate, severe and profound categories. Some add a “slight” category between normal and mild
hearing loss\textsuperscript{11,12} and some a moderately severe category.\textsuperscript{11} Variation in the range of sound intensities that are defined by these degrees of loss also exist. For example, the upper threshold covered by a moderate loss label ranges across 55, 60, 65 to 70 dB HL between systems of categorisation. A hearing loss is classified as mild once a threshold is either 20 or 26 dB HL. It is plausible that such differences between categorisation systems are why some recent studies of hearing loss prevalence in adults have defined mild hearing loss as 26 dB HL or greater,\textsuperscript{13-16} as 26 dB is common to all categorisation systems’ definitions of mild hearing loss. The horizontal lines in Figure 2.2 denote the categorisations used for this PhD, as discussed in Section 2.4.1.

\begin{figure}[h]
\centering
\includegraphics[width=\textwidth]{figure2.2.png}
\caption{Variation in degrees of hearing loss internationally}
\end{figure}

An additional complication in the literature is the varied use of the term “minimal” hearing loss. It either indicates a hearing loss of slight degree, or is used as an umbrella term for both bilateral mild losses and unilateral losses of any degree. It is evident that terminology has not remained uniform over time or across disciplines.\textsuperscript{17} This has resulted in a literature that is difficult to navigate and presents challenges for correctly interpreting participant outcomes often
described in relation to the category of loss (e.g. slight, mild, minimal) rather than the audiometrically measured hearing levels.

Some variation in categorising degrees of loss may also result from the differing needs of research- versus clinically-based audiometry. For clinical purposes, degrees of loss can be broken into the broad categories listed in Table 2.1, which assign roughly evenly-sized audiometric ranges to each category and closely aligns to the divisions of hearing loss used in an Australian rehabilitative setting.18

<table>
<thead>
<tr>
<th>Degree of hearing loss</th>
<th>Audiometric range (dB HL)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Normal hearing</td>
<td>-10 to 20</td>
</tr>
<tr>
<td>Mild</td>
<td>21 to 40</td>
</tr>
<tr>
<td>Moderate</td>
<td>41 to 65</td>
</tr>
<tr>
<td>Severe</td>
<td>66 to 90</td>
</tr>
<tr>
<td>Profound</td>
<td>&gt;90</td>
</tr>
</tbody>
</table>

2.2.4.1 The Pure Tone Average (PTA)

An audiogram (the entire plot of an individual’s hearing thresholds) may be assigned a degree of hearing loss by visual observation of the general decibel region in which the thresholds lie. Alternatively, a specific frequency average is calculated (also known as a pure tone average or PTA) which averages measured thresholds across a frequency range considered to be especially relevant for understanding speech. This averaging method is used routinely in a clinical setting of hearing aid prescriptions; therefore, it features prominently in research that is conducted with amplified populations. Two common frequency average calculations are the three frequency average hearing loss (3FAHL) which averages thresholds at 500 Hz, 1000 Hz and 2000 Hz, and the four frequency average hearing loss (4FAHL) which also includes the 4000 Hz threshold. The 3FAHL or 4FAHL is expressed as a single number, which is then assigned a degree of loss based on where within the classification system it falls.
As with the multitude of categorisation systems outlined in Section 2.2.4 above, there also exist alternative PTA calculations in the literature. These alternate calculations are commonly used in hearing loss prevalence research, typically where subtler degrees of loss are detected in a general population sample rather than a clinically-defined cohort. Further detail of other PTA calculations in the literature is in the section on milder hearing loss prevalence (Section 2.4.3).

### 2.2.4.2 Aided audibility and the Speech Intelligibility Index (SII)

The above methods of grouping hearing loss into specific degrees conventionally apply to true or unaided hearing thresholds. Several potential limitations of the PTA itself as a measure of hearing ability warrant discussion. Firstly, the PTA calculation cannot fully reflect the range of audiometric patterns that could produce the same average value. For example, a steeply sloping high frequency loss, a rising low frequency loss and a flat loss could share the same PTA value. This is despite the very different access to speech sounds that these three separate audiograms would connote. Secondly, the PTA no longer reflects the hearing levels of an aided child. Hearing aids enable children with hearing loss to hear sounds they otherwise could not. A child who has been fitted with amplification is encouraged to wear their device during waking hours and is, therefore, in this aided state as they actively and incidentally learn language across their typical daily activities. It follows that the PTA may not best quantify hearing ability in a child’s everyday life as their hearing thresholds are improved while wearing hearing aids. Therefore, the PTA may not be best able to account for language outcomes in early-amplified children.

Consequently, researchers have recently focused on the concept of ‘aided audibility’. Aided audibility refers to a method of quantifying hearing that incorporates the amplified acoustic signal. The Speech Intelligibility Index (SII) is one example of an aided audibility measure. The SII\textsuperscript{19} is calculated by dividing the speech frequency range into discrete frequency bands, to each of which two functions are applied. The audibility function represents the amount of speech that is supra-threshold (i.e. audible as it is louder than the individual’s threshold), contributing to speech recognition. An importance function weights
particular frequency bands according to their relative significance for overall speech recognition, taking account of the phonemes that occur within a specific language.\textsuperscript{20} Within each band the audibility function is multiplied by the importance function, the product of which is summed across all frequency bands to obtain one SII value. A higher SII score represents greater speech intelligibility.

Originally designed for understanding the relationship between acoustic speech signals and speech recognition in adults, there is evidence that the SII can quantify speech audibility\textsuperscript{21} and predict speech recognition scores in children.\textsuperscript{22} The use of the SII as a tool to explore the relationship between audibility and language outcomes is covered in more detail in Section 2.5.3.1.

\textbf{2.2.4.3 Functional assessment tools}

Whilst not a measure or method of quantifying hearing loss itself, the impact of hearing and aided audibility on an individual may be observed through the use of functional assessment tools. Such indirect tools aim to examine the effectiveness of amplification and to understand how well hearing is used in daily life.\textsuperscript{23} Use of such tools could provide insight into whether audibility, when measured observationally via a functional assessment, influences development of age appropriate speech and language. One validated functional hearing measure recommended for use in audiological protocols\textsuperscript{24} is the Parents’ Evaluation of Aural/Oral performance of Children (PEACH) rating scale.\textsuperscript{23} In this tool, caregivers rate the frequency of the child’s reactions to auditory input in different listening conditions. Higher PEACH scores are reported as positively correlating with language measures for children with bilateral loss at 3 years of age.\textsuperscript{25} Functional assessment questionnaires and their contribution to understanding speech and language outcomes of children with hearing loss are considered in Section 2.6 of this literature review, as part of the management of hearing losses.
2.3 Detection of hearing loss

Earlier detection of congenital hearing loss has facilitated both earlier diagnosis and, subsequently, earlier management. The purpose of this section of the literature review is to contextualise the environment in which congenital hearing loss is now detected. It starts by covering the original drivers for changes in detection systems, which were the attempts to improve the overwhelmingly poor outcomes experienced by children whose severe and profound congenital losses were detected late. This is achieved by contrasting historical to contemporary approaches of hearing loss detection. Throughout, the progression towards earlier detection is outlined for the state of Victoria, where this PhD study was conducted. The impact of these changes on the detection of mild and moderate hearing losses is also highlighted by exploring the population health principles that guide screening activities. This explains the inevitable earlier detection of mild hearing loss that has occurred alongside moderate and greater losses.

2.3.1 Historical detection

Early systematic screening originated from a desire to detect more children with hearing impairment than was possible by incidental or passive observation of apparent hearing loss alone.\textsuperscript{26} As early as the 1940s, professionals were forming the opinion that identifying children with hearing losses earlier en masse could lead to improved language, education and whole of life outcomes, compared to the typically disastrous outcomes seen in those with significant hearing losses identified late. Delays attributed to congenital losses were measurable at under 3 years of age\textsuperscript{27,28} and on average, deaf high school graduates were equipped with language and academic skills comparable to fourth-grade children without hearing loss.\textsuperscript{29} Influential drivers of improvements to the age of detection include improvements in hearing assessment methods,\textsuperscript{30} advances in technology\textsuperscript{31} and government policy changes/expert body position statements (e.g. National Institutes of Health,\textsuperscript{32} Joint Committee on Infant Hearing\textsuperscript{33}).

A growing understanding of behavioural responses to sound influenced initial screening activities targeted at younger children. For example, by the 1960s in
the United Kingdom (UK), children experienced two separate hearing screenings during early childhood. A distraction test was performed at 7-8 months of age, and a hearing screen was performed in the first year of formal education. The distraction test was a structured behavioural observation of sound awareness, modified from work that showed hearing testing before 12 months of age was feasible. The hearing screen consisted of a supra-threshold presentation of swept pure tones under headphones. This activity was viewed as valuable from an educational standpoint, in addition to acting as confirmation of earlier screening results. The hearing screening pre-dated the distraction test, having been employed in various iterations since the 1930s.

Early screening in the state of Victoria, Australia, resembled some of the activities that were occurring in the UK. Both localities operated comprehensive community nursing systems, which were employed to perform early behavioural hearing screenings across the population. These screenings in Victoria, again based on the work by Ewing, were performed without standardisation across the state, and screen outcomes were vulnerable to inconsistencies in referrals for further hearing assessment. Without clear pathways towards diagnosis, an individual child's path after not passing a behavioural hearing screen was not always commenced, completed or optimised.

The first iteration of the Victorian Infant Hearing Screening Program (VIHSP) commenced operation in 1992, aiming to systematically reduce the age at diagnosis and fitting of hearing aids for children with a moderate or greater (>40dB HL) bilateral hearing loss to under 12 months of age. To achieve this, two distinct, statewide screening methods were employed in tandem. The first was a screen of newborns for the presence of a risk factor for hearing loss (see Section 2.2.3 for risk factors), which if identified would trigger a referral to an audiologist for a prompt electrophysiological diagnostic hearing assessment. The second screening method was a behavioural screen of all children without any risk factors for hearing loss, after they turned 8 months of age but within their first year of life. This two-tier screening system was devised because screening children for hearing loss based on risk factors alone could only theoretically
achieve a maximum diagnosis yield of 60% because so many children had no overt risk factors.\textsuperscript{35}

Both prior to commencing and throughout this two-tier screening program, thorough training was conducted to ensure that community nurses and audiologists were operating within the same testing parameters. Referral pathways for children who did not pass the screen were formalised and tightened, in attempts to reduce the number of children lost to follow up, and aid prompt initiation of intervention.\textsuperscript{35} The results of this exercise were an increased number of babies diagnosed with hearing loss by six months of age, and more infants being fitted with hearing aids than pre-1992. The mean age at diagnosis of a moderate or greater loss (i.e. $>$40 dB HL) fell from 20.3 months to 14.2 months. The program achieved a significant lowering of the median age of diagnosis for those children with severe hearing loss (from 22.4 to 11.2 months). However, the results did not indicate significant earlier median age of diagnosis for children with moderate and profound hearing loss.\textsuperscript{35} Median age at diagnosis of mild hearing loss paradoxically appeared to increase across this study period, from 39.6 months to 52.1 months of age. This was likely artefactual - a result of the systematic screening approaches and more widely available diagnostic facilities resulting from VIHSP, leading to increased ongoing surveillance of child hearing using behavioural techniques across the early years of life. Such surveillance could result in a higher proportion of older children being diagnosed with mild hearing loss. Thus the two-tier screening approach used in the first iteration of VIHSP would have increased the chances of not only children born with but also those developing mild losses being detected more readily than in the past.

This trial\textsuperscript{35} enabled more infants to be diagnosed early with hearing loss and shifted diagnosis to a lower median age for a sub-set of those targeted. However, the overall results were considered disappointing and highlighted a need for more direct methods of early detection. Importantly, the trial affirmed the success of technological advances allowing the diagnostic assessment of infants for hearing loss. It was during this period that these diagnostic tests were
recognised as having potential use as a screening tool, to objectively screen hearing across the newborn population.

2.3.2 Technological advances

Prior to the technological era of the last 50 years, the only method for evaluating hearing was to observe a behavioural response to an auditory stimulus. By varying the properties of the stimulus, be it the intensity or the frequency, an audiogram could be documented. An audiogram is a graphical representation of the softest sounds that an individual is able to reliably detect, typically plotted across a frequency range that is most important for communication purposes. Today, behavioural test techniques are still considered the gold standard technique for participants from approximately 12 months of age onwards. However, this is not possible for infants – the group for whom diagnosis is now seen as most urgent. Electrophysiological measures now exist that harness the involuntary responses of the auditory system when stimulated. These evoked measures provide an alternative to behavioural assessment for those in the population where information on hearing status is crucial for appropriate early intervention and ongoing management. These measurements are used to provide information that is interpreted to estimate hearing levels in those unable to provide reliable behavioural responses to sound, including instances of physical and intellectual disability, non-organic hearing impairment, and infancy.

Two commonly used measures have been successfully modified to function as hearing screening tools: the electro-acoustic Otoacoustic Emissions (OAEs) and the electrophysiologic Auditory Brainstem Response (ABR) test. As noted in Section 2.2, all steps in the auditory pathway must be operational for normal sound detection to occur. Therefore, testing towards the end of this pathway (i.e. at the cochlear level) will in effect test the entire peripheral pathway.

OAEs were identified in the 1940s but were not used for diagnostic purposes until the 1980s. OAEs are an elicited sound of biological origin recorded in the ear canal by a microphone. These sounds originate from healthy, correctly functioning outer hair cells within the cochlea, functioning in response to an
auditory stimulus delivered in the ear canal. Therefore, present OAEs measure successful transmission of a stimulus through the outer, middle and inner ear only.\textsuperscript{36} OAEs are primarily used in a diagnostic sense as supportive evidence for hearing status rather than the indicator of an exact threshold level; the measured OAE intensity does not directly relate to cochlear ‘strength’.\textsuperscript{37}

By comparison, the ABR can estimate a hearing threshold and has been in use clinically since the 1970s.\textsuperscript{38} The test involves recording neural activity at the auditory nerve and brainstem level, elicited by an auditory stimulus presented to the outer ear. Responses from electrodes on the scalp measure the sound transduction through the outer, middle and inner ear and into the auditory brainstem. Hearing thresholds using the ABR are determined by altering stimulus properties such as frequency and intensity.\textsuperscript{36}

These two measures required some modification to perform the newborn hearing screening tasks within required parameters. Of particular relevance for the ABR, practical issues such as length of testing time, portability of equipment and staffing by experts who could interpret the results were limiting factors.\textsuperscript{38} To function as an efficient screening tool, these diagnostic tools interpretable solely by experts needed to be transformed into devices that could provide a binary (pass/fail) outcome. With technological advances, these two measures were successfully adapted into screening form, the automated auditory brainstem response (AABR) and the OAE screener. Both tools could be applied to population-wide screening of newborns.\textsuperscript{39}

2.3.3 Universal Newborn Hearing Screening trials and evaluations

The primary desired outcome of Universal Newborn Hearing Screening (UNHS) is to lower the age at diagnosis for children born with hearing loss, allowing earlier amplification and improving the chances of better language, education and overall outcomes for affected individuals.\textsuperscript{40} In turn, this is expected to improve their ability to actively interact within the mainstream hearing society at an acceptable cost-benefit ratio.\textsuperscript{41} Well before evidence of efficacy of UNHS was measured, peak bodies were advocating for earlier diagnosis of hearing
impairment\textsuperscript{42} under the educated assumption that earlier identification would translate into more effective intervention and subsequently improved outcomes across the lifespan. The United States National Institutes of Health (NIH) released in 1993 a Consensus Development Conference Statement that recommended every newborn have a hearing screen in the first few months of life.\textsuperscript{43, 44} The Joint Committee on Infant Hearing (JCIH), representing various American Academies and invested national professional bodies, released a series of Position Statements dating from 1982 that document shifting recommendations mirroring improved technology over time. Early statements advocated for targeted risk factor screening.\textsuperscript{45, 46} The 1994 Position Statement\textsuperscript{47} endorsed the 1993 NIH statement for early screening for all newborns, prompted by evidence that approximately half of infants diagnosed with hearing loss did not possess any risk factors for hearing impairment. By the year 2000, UNHS was being advocated, following the “1,3,6 principle” (screen before 1 month, diagnose before 3 months, enroll in intervention by 6 months).\textsuperscript{33}

Published evidence of the partial efficacy of UNHS programs mostly occurred within a 20-year period from the early 1990s onwards. Reviews of these trials occurred for the purpose of position updates of various authorities and expert panels. One of the more rigorous studies was carried out between 1993 and 1996 in four hospitals in Wessex, UK\textsuperscript{48}, as a controlled trial on the efficacy of neonatal hearing screening in increasing the rate of early diagnosis of hearing loss.

The trial, using a two-stage OAE/AABR screening approach, was designed to test the hypothesis that UNHS in hospital in addition to the standard behavioural screen performed at 8 months (i.e. the distraction test described in Section 2.3.1) would increase rates of early referral, diagnosis and management of congenital hearing loss, over and above the use of the behavioural screen alone. Of the 53,781 babies in the trial, approximately half were born during the period of UNHS, with hearing outcomes by 10 months of age then compared to those of infants born prior to the trial. Key findings included an 80% yield of diagnoses (90 cases) during UNHS, based on the expected prevalence for bilateral hearing
loss of greater than 40 dB HL. Under UNHS conditions, 62 more babies born with moderate or greater losses were referred for audiology before six months of age compared to the referrals under behavioural screening alone. Therefore, early confirmation and management of infants with hearing loss was significantly increased for UNHS newborns and the type II error rate (i.e. the false-negative result of screening activities) dropped significantly from 27% in the distraction test alone setting to 4% in the UNHS setting.48

The results of the Wessex trial also showed that UNHS was a worthwhile exercise for all infants, not just the “at-risk” population of those infants with specific risk factors. Consistent with previous findings,49 UNHS did not contribute other harms such as increased parent anxiety. This universal value to all infants from UNHS has been confirmed in the years since the Wessex trial, notably in the Victorian context by Barker and colleagues (2013).50 Barker et al (2013) found via a population audit that a UNHS program that targeted every baby (whether born healthy or admitted to the neonatal intensive care unit (NICU)) outperformed NICU-only screening activities on measures including mean capture and follow-up rates and mean age at diagnostic audiology attendance.50 Reassuringly, recent examination of the diagnostic yield of target condition hearing losses pre- and post-UNHS implementation are comparable, with 1.12 diagnoses per 1000 live births reported in 199751 and approximately 1 in every 1000 infants reported in 2015.52

2.3.4 Universal Newborn Hearing Screening adoption
Adoption of UNHS occurred within a short period of time in advantaged countries around the world. In Europe, the Netherlands had started using UNHS rather than distraction techniques from 20029 and the UK had commenced UNHS roll-out in 2000.53 In Canada there was province-wide UNHS in Ontario in 2002.54 In the USA federal funding was provided in 2000 for UNHS, and by April 2001 there were 38 states that had either legislated or had legislation pending for UNHS to be a requirement.43 Such changes contributed to the significant rises observed in screening rates: from fewer than 5% of infants in developed countries screened in 1993, to greater than 90% in 2004.55 In Australia, in 2002
Western Australia, closely followed by New South Wales, was the first to commence UNHS, with Victoria commencing UNHS activities in 2005.

The majority of UNHS programs target congenital losses of bilateral moderate or greater severity (i.e. typically >40 dB HL). In order to achieve this, milder degrees of loss than those targeted are commonly detected. A few jurisdictions have specifically set the target of UNHS to include unilateral and mild bilateral losses, operating under the rationale that knowledge of any degree of hearing loss is valuable for ongoing management. Therefore, UNHS activities have led to earlier diagnosis of milder degrees of hearing loss. This earlier diagnosis of mild losses is reviewed in more detail in Sections 2.3.5 and 2.4.

The direct outcome of UNHS programs, a lowering of the age at which congenital hearing loss is detected, has been established by the above studies and numerous others. However, further evidence was required to confirm that this success at early detection translated into improved longer-term language outcomes. Documenting the improved longer-term outcomes of those earlier-diagnosed infants when compared to children later-diagnosed would provide important evidence in the UNHS cost-effectiveness debate, as governments initially funded programs without strong empirical evidence of population-level efficacy.

The United States Preventive Services Task Force (USPSTF), a panel of experts in evidence-based medicine, commissioned two systematic reviews of the evidence surrounding UNHS and subsequently released Recommendation Statements of their main findings. In their 2001 Statement, based on the review by Thompson et al., the USPSTF concluded that at that time there was not enough evidence to advocate for or against UNHS. It noted clear evidence that UNHS decreased the age of diagnosis for moderate or greater bilateral hearing loss, but this was not accompanied by evidence of better long-term language outcomes of these early-identified children; more studies were called for. The next USPSTF Statement in 2008 concluded that sufficient evidence existed to indicate that children diagnosed at an earlier age had improved scores on measures of expressive and
receptive language ability. The systematic review of the literature, which formed the evidence base for the 2008 Statement, relied primarily on a single retrospective cohort study assessing language outcomes of 120 children at a mean age of approximately 8 years. Higher adjusted mean z scores for language (receptive language mean difference 0.82, 95% CI 0.31 to 1.33, expressive mean difference 0.70, 95% CI 0.13 to 1.26) were observed when hearing loss confirmation occurred prior to 9 months of age. Overall, the 2008 USPSTF Statement concluded with moderate certainty that there was benefit to performing infant hearing screening universally.

However, the USPSTF position was not a viewpoint accepted by all. A further systematic review of literature published up to October 2007 highlighted serious study design issues in the majority of identified studies. This tempered the favorable differences observed in language development for early- versus late-identified children that the USPSTF based their recommendation of UNHS upon. Consequently Wolff et al (2010) concluded that, based on available evidence, there was only a slight indication of benefit from earlier diagnosis.

In summary, the impact of UNHS on reducing the age at which hearing loss is detected at a population level is not in doubt. If this were the only goal of UNHS programs, they would be comfortably labeled a success. However, UNHS programs were envisaged to be the fix for the longer-term impact of hearing loss on affected individuals’ quality of life, most immediately measured by improvements in childhood language development. In this regard, it is not yet conclusively established that earlier detection of hearing loss leads to clear benefits for language outcomes of affected children. Further review of the literature on the language outcomes of children with hearing loss occurs in Section 2.5.

2.3.5 Mild hearing loss detection via Universal Newborn Hearing Screening

For any screening program, population health principles influence the outcomes achieved. As described above, UNHS has successfully reduced the age at
detection of moderate and greater losses. At the same time, this activity has reduced the age of mild loss detection, which is not the target of most UNHS programs. The principles behind why this occurs are important to consider. They explain why it is inevitable that mild losses are now detected earlier than ever before, and what this earlier detection means for those individuals diagnosed.

Screening programs must be carefully designed to ensure they can provide a valid, reliable and accurate measure of what is aimed to be measured.\textsuperscript{66} Awareness of these requirements has grown in the decades since the publication of Wilson and Jungner’s widely referenced 10 principles of screening.\textsuperscript{67} These principles were proposed to act as an initial checklist, whereby research into the feasibility of screening for a particular condition would only be warranted if the broad conditions in the checklist were met. However, in practice screening programs can only effectively be appraised retrospectively, when the product of the activity can be observed.

In screening terminology, sensitivity represents how well the test can detect the condition of interest when the condition is truly present. Specificity is how well the test can confirm the absence of the condition being screened for, when it is truly absent. Table 2.2 (modified from Raffle & Muir Gray 2007\textsuperscript{66}) illustrates how these two measures of screening efficacy are calculated, with sensitivity $= A/(A+C)$ and specificity $= D/(B+D)$. To be clinically useful, a screening test needs to have both good sensitivity and specificity.\textsuperscript{68}

<table>
<thead>
<tr>
<th>Screen result</th>
<th>Target condition</th>
<th>Present</th>
<th>Absent</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>Positive</td>
<td>A: true positive</td>
<td>B: false positive</td>
<td>A + B</td>
<td></td>
</tr>
<tr>
<td>Negative</td>
<td>C: false negative</td>
<td>D: true negative</td>
<td>C + D</td>
<td></td>
</tr>
<tr>
<td>Total</td>
<td>A + C</td>
<td>B + D</td>
<td>A + B + C + D</td>
<td></td>
</tr>
</tbody>
</table>
sensitivity 100%, specificity >98%, see Kwon & Farrell (2010)\textsuperscript{69}. Such screen performance, when illustrated on a Receiver Operator Characteristic (ROC) curve with sensitivity on the y-axis and 1 - specificity on the x-axis, would see the plot hugging the left side and top of the graph and result in an area under the curve approaching 1.\textsuperscript{66} Such a plot illustrates that for the chosen cut-point of the screen, the test is able to successfully identify essentially everyone with the condition in the screened population. That is, there has been no significant trade-off in specificity in the process of obtaining maximum sensitivity. Both high sensitivity and specificity is more readily achievable when the screen cut-point is closer to what is achieved in a diagnostic test. This is the case in PKU screening for example, where the screen looks for presence or absence of the enzyme phenylalanine hydroxylase.\textsuperscript{70}

Other screening tests, such as screening for congenital hearing loss, experience a trade-off between sensitivity and specificity performance. Congenital hearing loss is a condition where there is less clear delineation between condition presence versus absence, as hearing loss presents along a continuum of severity. As mentioned earlier, most UNHS programs target moderate or greater hearing loss. This is a somewhat arbitrary target in terms of condition presence versus absence, as a loss is still measurable at decibel levels lower than the screening target set. Therefore, in order to maintain optimal sensitivity (i.e. not miss any children born with moderate or greater hearing loss), the specificity of the test will necessarily be compromised by the inadvertent detection of mild hearing loss. This means some children with mild losses will be detected earlier than was historically possible, a direct result of the efforts of population-level newborn screening to identify all children with moderate and greater hearing loss.

2.3.6 The balance between benefit and harm in screening
A related yet slightly different issue is the balance between benefit and harm of screening and how this factors in decision-making on the degree of hearing loss to target. Within the UNHS literature, the challenge of choosing a cut point at which to screen has been articulated via the statement, “...at some point on the
severity continuum, evidence in a number of domains becomes equivocal, open to multiple interpretation, poorly-controlled or just non-existent, largely because the sub-population of mild or unilateral permanent childhood hearing impairment may be difficult to identify with the requisite certainty.”". This scenario reflects the commonly encountered reduction in measurable benefit with a lowering of the “threshold” for detection of the target condition. This is visualised in Figure 2.3, showing the effect of lowering the cut-point of hearing loss targeted by screening. As the cut-point shifts further to the left of the graph (representing a lesser degree hearing loss targeted by screening), a larger number of children will be flagged by the screen in the ever-present attempt to not “miss” any child with the condition of interest. This will increase the false positive rate, exposing more families to potential harm from a scenario of a diminishing return on the expected benefit of earlier diagnosis when the condition is truly present. A false positive in this scenario of a condition along a continuum is also somewhat arbitrary, being linked to the decision about a screening cut-point. Does treatment exist that warrants the detection and justifies the inevitable costs?

![Figure 2.3 Distribution of hearing loss severity across the population](image)

It has been said, “all screening programs do harm; some do good as well, and, of these, some do more good than harm at a reasonable cost”. At the most basic level, the money invested in screening represents a resource that could otherwise have been spent on something else, which in an environment of finite resources represents harm. Applying this concept of harm from screening activities to the question above of the appropriate level of hearing loss to screen
for presents the following conundrum. The balance between benefit (e.g. the amount of improvement in child language outcomes resulting from optimised hearing and earlier intervention, improved ease of communication, positive impact on social integration) and harm (e.g. cost to the healthcare system of excess investigations, diagnosis stigma, burden to the family in time, stress and inconvenience)\textsuperscript{72} is susceptible to shifting towards harm as attempts are made to detect less severe conditions.

In this UNHS scenario, for those ultimately diagnosed with a “non-target” mild hearing loss, the costs of this earlier diagnosis may not be fully actualised at the time of detection and instead continue to accrue over time. This may occur via “treatment creep”, where the well-meaning desire to intervene for a diagnosed condition happens without evidence of efficacy. As described earlier, the literature on outcomes of children with more severe hearing loss is clear in that providing improved hearing earlier can improve childhood outcomes. As with all conditions, there is a larger number of children in the population with milder degrees of hearing loss than those with greater degrees of loss (see Section 2.4.3). Providing the same management options (e.g. earlier hearing aid fitting) to those with milder degrees of loss, based upon the extrapolation of good evidence of benefit in more severely impacted individuals may not be effective, as the benefit/harm ratio may be quite different.

Parallels can be drawn with scenarios where treatment proven as effective in one setting actually leads to harm when used in alternate settings. One recently published example highlighted that the success observed in reducing pre-term infant mortality via the use of corticosteroids in at-risk women in high-income countries actually resulted in increased overall infant mortality when the same treatment was administered in low- and middle-income countries.\textsuperscript{73} This treatment approach was not controversial and supported by prominent bodies such as the United Nations\textsuperscript{74} and World Health Organization,\textsuperscript{75} yet is an example of how treatment shown to be effective in one population cannot be assumed to be equally successful in other settings.
This “new” population of children diagnosed earlier with milder degrees of hearing loss, identified as a result of UNHS, may be experiencing management that is not grounded in relevant evidence. This current section of the literature review has provided context as to why some mild congenital losses are now necessarily detected earlier than historically possible and has alluded to how management practices are developing. Greater details of management for milder losses are considered as part of Section 2.6 of this literature review.

To set the scene, the next section of the literature review defines the level of milder hearing loss of interest to this PhD. It covers in more detail how these losses are typically detected, prevalence estimates and what is known regarding impact on affected individuals.

### 2.4 Milder hearing losses

The purpose of this section of the literature review is to focus attention on the degrees of hearing loss that are the subject of this PhD study, i.e. mild and moderate bilateral loss. These degrees of hearing loss are the topic of this PhD due to their status as an under-researched group in the literature and for whom there is some management uncertainty. From this point onwards the literature review will be concentrating on children with these degrees of hearing loss. Initially the term *milder* hearing loss is defined, as it is a study-devised term for use in this thesis. Then the remainder of this section will focus on trends in detection, prevalence and the general impact of milder hearing loss. The impact of milder hearing loss on language outcomes specifically is covered in Section 2.5.3.

#### 2.4.1 Milder hearing loss definition

Throughout this thesis, the term *milder* hearing loss refers to bilateral, sensorineural hearing loss comprising both mild and moderate degrees. The decibel range that the term milder hearing loss covers is defined as a PTA of 21 to 65 dB HL.
The lower end of mild hearing losses is variably defined according to an interpretation of where normal hearing ends, and whether “slight” hearing losses are defined. For example mild hearing loss is noted by the American Speech-Language-Hearing Association (ASHA), the Centers for Disease Control and Prevention (CDC) and the World Health Organization as spanning 26-40 dB HL. Both the British Society of Audiology and Australian Hearing indicate that mild hearing loss covers the average threshold range of 20 or 21-40 dB HL. Both ASHA and CDC systems include a “slight” degree of loss, for average thresholds spanning 16-25 dB HL. These differences between systems are illustrated in the earlier Figure 2.2 in Section 2.2.4.

As illustrated, the upper limit of the mild hearing loss range is widely accepted to be around 40 dB HL, with thresholds higher (poorer) than this level yet lower (better) than approximately 65 dB HL classified as a moderate hearing loss. As explained earlier, the averaging of thresholds across either three or four frequencies is commonly used to determine where an individual falls within a classification system. From the horizontal lines drawn in Figure 2.2 denoting this PhD’s chosen definition of mild and moderate hearing loss, it is clear that these two degrees combined under the label “milder” hearing loss approximates the broad divisions used internationally to label hearing loss.

2.4.2 Milder hearing loss detection
UNHS programs have facilitated the routine diagnosis of moderate hearing loss at ages considerably earlier than either behavioural or risk factor screenings could systematically achieve. From a historical age of detection at around 2 years of age for moderate and greater losses and a mean age of almost 3 years for moderate losses specifically, modern UNHS programs now report a median age of 7 weeks for the confirmation of moderate or greater loss. Whilst this result groups children with moderate, severe and profound losses together, it is safe to assume that the inclusion of moderate loss as a target condition of UNHS (see Section 2.3.4 above) results in earlier diagnosis.
A minority of UNHS programs, typically found across North America, aim to detect mild hearing loss.\textsuperscript{52} Technological parameters are set to enable this (i.e. a lower intensity eliciting stimulus), with an acceptance of a higher program false-positive rate in order to detect the less severely impacted.\textsuperscript{79} However, the majority of screening programs internationally do not target mild loss and instead focus on detecting losses where there is greater consensus of benefit from earlier detection, i.e. moderate and greater.\textsuperscript{52, 58, 60} As discussed in Section 2.3.5, when screening targets a particular (and somewhat arbitrary) degree condition that exists on a continuum of severity, it will inevitably detect some cases of lesser severity than the stated target in an attempt to avoid any false negative screening outcomes. In these UNHS systems programmed to detect moderate and greater hearing loss, the detection of mild hearing loss could be considered a “by-product” of UNHS.

Certain characteristics of the screen stimulus, typically an acoustic click, could potentially explain the detection of untargeted mild losses.\textsuperscript{80} Primarily, the aim of detecting all congenital losses of moderate and greater degree necessitates an AABR click stimulus intensity of less than moderate levels (e.g. in the Victorian scenario, VIHSP uses a 35 dBnHL click). This intensity setting would yield some false positives in the form of mild hearing losses in the 35-40 dB range. Additionally, the multiple frequencies contained within the acoustic spectrum of a click stimulus leave screening technology vulnerable to individual audiometric configuration differences. An infant whose frequency-average hearing loss falls within the mild range could fail a newborn hearing screen due to the sloping “shape” of hearing thresholds in the frequency range analysed. Equally, the infant may pass the screen if their shape is less sensitive to the properties of the stimulus. The former scenario contrasts to an audiometric flat loss in the mild range (e.g. at 25 dB HL) that would not be detected by UNHS, as the infant would produce an electrophysiological response to the supra-threshold eliciting stimulus.

It is also possible that for some infants born with mild hearing losses, attributes of their auditory response to sound, versus the background noise during
newborn hearing screening, leave them vulnerable to not passing an AABR screen. The signal-to-noise ratio, known to be crucial for ABR identification in a diagnostic setting, may be below the criteria required by the AABR technology to register a pass result in some infants with mild losses.

Irrespective of the mechanism by which mild hearing loss is detected by a screen targeting moderate and greater loss, additional evidence for earlier detection for mild losses in the UNHS era comes from rising hearing aid fitting rates in very young children. National figures from Australia display recent growth in the number of fittings occurring prior to 12 months of age when the better hearing ear is audiometrically within the mild hearing loss range (i.e. 21 to 40 dB HL). Such fittings are occurring at an age before which mild hearing losses were historically detected, indicating that UNHS has had a major impact not only on age of detection but on actual treatment of mild hearing loss. With age at identification often inversely related to the degree of loss when relying on behavioural observation of deficits, the impact of UNHS on driving down the age of detection of mild hearing loss is clear.

2.4.3 Milder hearing loss prevalence

Prevalence estimates for milder degree hearing losses display considerable variation, likely resulting from methodological differences between studies. However, an understanding of approximate prevalence of milder hearing loss provides context to the value of the question being explored in this PhD.

In the late 1990s/early 2000s there existed no national data on the expected prevalence of congenital hearing loss of any degree, either in the United States or the United Kingdom. Subsequently, attempts at national data collection have been made using a multitude of data collection methodologies and platforms across different age ranges. Variation in subject age, sampling procedures and participation rates, as well as differences in the definition of hearing loss have been noted. This has led to a literature base requiring careful interpretation to understand the prevalence estimates for mild and moderate hearing loss.
As outlined in Section 2.4.2, one of the secondary products of UNHS has been the early identification of mild hearing losses. This has seen reported age of detection drop from historical levels of four to six years of age\textsuperscript{54,87,88} to much younger, with some studies reporting a median detection age of 0.8 years\textsuperscript{54} and less. Despite age at detection decreasing, there is not a wide literature on the prevalence of mild losses in the newborn period. A longitudinal study of 35668 children who were exposed to newborn hearing screening in the United Kingdom recently reported a prevalence of mild bilateral hearing loss of 1.32/1000 children when assessed by the first year of formal schooling\textsuperscript{89} but it is impossible to know what proportion of these children developed hearing loss post-neonatally.

Prior to the availability of such UNHS cohorts to facilitate longitudinal population-level prevalence estimates from infancy onwards, prevalence was typically established via cross-sectional studies at an age where wide sampling was feasible. Therefore, most prevalence estimates for mild loss have focused on school-aged children. Table 2.3 presents some prevalence estimates in the literature for slight/mild and moderate losses. The prevalence estimates for slight/mild losses are particularly broad, ranging from 1.32 to 36 in every 1000 children. Characteristics of two widely-cited studies included in Table 2.3, that of Wake et al (2006)\textsuperscript{90} and Bess et al (1998)\textsuperscript{84}, are explored below to help explain the substantial differences in prevalence estimates and inform interpretation of the broader literature.
<table>
<thead>
<tr>
<th>Study (year), design</th>
<th>n</th>
<th>Sample details (degree of loss, age/ schooling level)</th>
<th>Hearing loss calculation</th>
<th>Prevalence rate per 1000 children</th>
<th>95% confidence interval</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Slight / mild loss</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Bess et al (1998)</td>
<td>1218</td>
<td>Mild bilateral loss defined as 20-40 dB HL bilaterally in grades 3, 6 and 9.</td>
<td>3FAHL (0.5, 1, 2kHz)</td>
<td>16</td>
<td>-</td>
</tr>
<tr>
<td>Wake et al (2006)</td>
<td>6581</td>
<td>Slight/mild bilateral loss defined as 16-40 dB HL better ear in second and sixth year of formal schooling.</td>
<td>3FAHL 0.5, 1, 2kHz (LPTA) and 3, 4, 6kHz (HPTA)</td>
<td>8.8 (slight/mild)</td>
<td>0.66 to 1.15</td>
</tr>
<tr>
<td>Watkin and Baldwin (2011)</td>
<td>35668</td>
<td>Mild bilateral loss defined as 20-39 dB HL better ear by first year of formal schooling.</td>
<td>4FAHL (0.5, 1, 2, 4kHz)</td>
<td>1.32</td>
<td>0.94 to 1.69</td>
</tr>
<tr>
<td>Stevens et al (2013)</td>
<td>111951</td>
<td>Mild bilateral loss defined as 20-34 dB HL aged 5-14 years.</td>
<td>4FAHL (0.5, 1, 2, 4kHz)</td>
<td>17</td>
<td>12.4 to 25</td>
</tr>
<tr>
<td>Feder et al (2017)</td>
<td>178000</td>
<td>Mild bilateral loss in poorer ear defined as 21-40 dB HL ages 6-18 years. 26-40 dB HL at age 19 years.</td>
<td>4FAHL (0.5, 1, 2, 4kHz)</td>
<td>36</td>
<td>28 to 44</td>
</tr>
<tr>
<td>Study (year), design</td>
<td>n</td>
<td>Sample details (degree of loss, age/schooling level)</td>
<td>Hearing loss calculation</td>
<td>Prevalence rate per 1000 children</td>
<td>95% confidence interval</td>
</tr>
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<tr>
<td><strong>Moderate loss</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Fortnum et al (2001)92</td>
<td>17160</td>
<td>Better ear moderate bilateral loss (41-70 dB HL) aged 3 years and 9-16 years.</td>
<td>4FAHL (0.5, 1, 2, 4kHz)</td>
<td>0.45 to 0.60 (3 years)</td>
<td>0.40 to 0.50, 0.54 to 0.66</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>0.89 to 1.21 (9-16 years)</td>
<td>0.86 to 0.91, 1.18 to 1.24</td>
</tr>
<tr>
<td>Stevens et al (2013)94</td>
<td>111951</td>
<td>Moderate bilateral loss (35-64 dB HL) aged 5-14 years.</td>
<td>4FAHL (0.5, 1, 2, 4kHz)</td>
<td>0.5 to 2.7</td>
<td>0.4 to 0.8, 2.0 to 4.0</td>
</tr>
</tbody>
</table>
In a study of 6581 Australian children (3367 aged 6-7 and 3214 aged 10-11 years) Wake et al (2006)\(^9\) reported a prevalence of mild loss of 2.4/1000 children. Bess et al (1998),\(^8\) when studying 1218 children in the USA (comprising 565, 350 and 303 in Grades 3, 6 and 9 respectively), determined a prevalence of mild loss of 16/1000 children. Both studies were cross-sectional, meaning that late onset or progressive losses that were not present at birth may have been measured by school age sampling.

Briefly, the study by Bess and colleagues in Nashville, Tennessee, aimed to determine the prevalence of minimal hearing loss (mild bilateral and unilateral of any degree) and its impact on the educational performance of school children. It was the first large study to document the greater than previously identified difficulty experienced by children with this degree of hearing loss in the educational setting. It also documented a significantly higher level of grade retention (i.e. not progressing with age-matched peers into the next year of schooling) for the children identified with minimal hearing loss overall (37%), compared to school district norm retention rates (less than 10% averaged across the three school grades). The study by Wake and colleagues in Melbourne, Victoria established prevalence of slight/mild degree of bilateral loss in a population of school children that overlapped in age with that of Bess et al (1998). However, they found very little impact of the hearing loss when outcomes were assessed using strong objective standardised measures of educational progress and achievement.

The studies differed in participation uptake rates: the Victorian study achieving an 85% response rate in their true random sample, whereas the Tennessee study had an overall rate of 44%, falling from 65% for children in third grade to 30% of approached students in 9th grade. This drop in uptake rates for older children may suggest bias towards participation by students who were experiencing difficulties. The Tennessee study also had poor return rates for some teacher-completed outcome measures. These teachers, despite being blinded to the hearing status of the child, may have introduced reporting bias by preferentially returning measures related to students regarding whom they harboured
particular concerns. Whilst there was overlap in the age of participating children between the studies, Wake et al’s (2006) sample was overall younger and was seeking losses greater than 15 dB HL, rather than greater than 20 dB HL in Bess et al (1998). Such factors may contribute to the almost eightfold difference in prevalence estimates between the two studies.

Wake et al (2006)\textsuperscript{90} reported a lower prevalence than expected,\textsuperscript{84, 93} following air and bone conduction audiometric testing and tympanometry assessment. Upon attempting to explain this difference in prevalence, the authors contrasted their 0.88% prevalence rate of combined slight/mild losses with that obtained in Niskar and colleagues’ US National Health and Nutrition Examination Survey (NHANES) III.\textsuperscript{12} In their sample of over 6000 children aged 6 to 19 years, the NHANES study reported an overall prevalence rate of slight or worse bilateral hearing loss (i.e. $\geq 16$ dB HL) of approximately 3% determined via air conduction hearing screens only (no bone conduction or tympanometry). The clear majority of losses detected by NHANES III were slight/mild in severity, providing a comparable severity comparison with the similarly sized Wake et al (2006)\textsuperscript{90} study. Wake and colleagues suggested a plausible underlying 2% prevalence of conductive hearing loss in the US population sampled by NHANES could in fact indicate the Victorian study determined a similar prevalence of slight/mild loss, with respect to those with likely sensorineural type losses. It should be noted Bess and colleagues also delineated conductive from sensorineural losses. This suggests other factors regarding sampling and study design (as highlighted above) may have influenced the high prevalence estimates for mild losses noted.

A meta-analysis documenting global hearing loss prevalence provides estimates using data from studies conducted from 1973 to 2010.\textsuperscript{91} When restricting results to 18 high-income country studies with a combined sample of over 100000, estimates of mild hearing loss (20-34 dB HL) in children aged 5-14 years reach 17/1000 (95% CI 12.4 to 25.0). These estimates were all obtained from population-based studies with large sample sizes and excluded studies which were school-based due to concerns regarding environmental noise when detecting mild losses. Despite marked differences in inclusion criteria, the
similarity between the estimate of Stevens et al (2013)\(^91\) and Bess et al (1998),\(^84\) who both defined mild hearing loss across a similar decibel range, is of interest and provides support for a larger number of mild losses in school-aged children.

For moderate hearing losses, prevalence data sets are also scarce. This is partly due to the grouping of moderate with severe and profound losses in prevalence estimates under an umbrella covering the full range of typical degrees of loss targeted by UNHS activities. Table 2.3 highlights two studies (one retrospective cross-sectional,\(^92\) one meta-analysis\(^91\)) which provide prevalence estimates specifically for moderate losses, ranging from 0.45 to 2.7 in every 1000 children. The retrospective study focused on a 15-year birth period in the UK via a survey of health and education services for children with hearing loss.\(^92\) They reported a prevalence of moderate bilateral loss (41-70 dB HL) in children aged 3 years of 0.45/1000 children (95% CI 0.40 to 0.50) to 0.60/1000 children (95% CI 0.54 to 0.66). As expected, prevalence increased to between 0.89/1000 children (95% CI 0.86 to 0.91) and 1.21/1000 children (95% CI 1.18 to 1.24) at age 9-16 years.\(^92\) This finding was in line with other prevalence studies that show a similar trend in increasing rates of hearing loss when assessing older subjects.\(^94,\)\(^95\) From the meta-analysis of global hearing loss prevalence described earlier,\(^91\) moderate hearing loss in children (35-64 dB HL) is estimated to range 0.5-2.7/1000 (95% CIs 0.4 to 0.8 – 2.0 to 4.0).

Other large population-level studies provide estimates of a combined moderate, severe and profound prevalence. A birth cohort study of 64116 Victorian children born in 1993 found, for children aged at least 6 years who had been fitted with amplification, a prevalence of moderate or greater hearing loss of 1.12/1000 children (95% CI 0.88 to 1.41).\(^96\) In the same longitudinal study of 35668 UK children exposed to UNHS described earlier,\(^89\) moderate or greater hearing loss was detected in the first year of schooling for 1.51/1000 children (95% CI 1.11 to 1.92). A Canadian cross-sectional sample of 56000 children found a prevalence of moderate or greater loss (i.e. >40 dB HL) in 6 to 19 year old children of 11/1000 (95% CI 5.0 to 23.0).\(^86\)
In summary, much variation in the prevalence estimates of congenital milder degree hearing loss relates to unavoidable limitations resulting from differing study design. Most estimates have been made at school-age, with inherent limitations on concluding whether these losses were present from birth. However, despite methodological variations and study limitations outlined above, overall the prevalence of mild loss in school-aged children is likely higher than moderate loss.

2.4.4 Milder hearing loss impact

The broader impact of milder hearing loss on affected children is unclear. There may exist a perception that children with milder losses will experience relatively minor difficulties when compared to those with severe/profound losses. This perception has likely influenced the amount of research conducted on children with hearing losses in the milder range.\textsuperscript{97,98} Whilst this PhD focuses on language outcomes, the challenges in multiple domains experienced by impacted children are highlighted below.

Broadly, areas of known challenges include poorer performance in adverse listening conditions,\textsuperscript{99-101} lower speech recognition scores,\textsuperscript{102} challenges with academic performance,\textsuperscript{84,103} reported levels of fatigue,\textsuperscript{104} and measured levels of stress,\textsuperscript{105} compromised psychosocial outcomes\textsuperscript{106} and language development.\textsuperscript{107,108} However, we have a poor understanding of the specific challenges these children are at risk of experiencing, or at what thresholds they begin to surface. The recognised heterogeneity in outcomes achieved by children with mild losses can limit conclusions (e.g. Winiger et al (2016)\textsuperscript{109}). For example, some children with mild losses will develop spoken language without significant difficulties and may not be identified as experiencing a significant negative impact of their hearing loss,\textsuperscript{110} whereas some will experience significant language difficulties. Sections 2.5.3 and 2.6.3 will explore the literature on language development in children with milder losses in more depth, including the difficulty with isolating whether lower than expected scores in language domains are a product of the hearing loss itself or another contributing factor.
Of particular relevance for children with mild hearing loss, uncertainty in outcomes likely influences both parent and clinician decision-making on appropriate management and intervention approaches. Without a clear understanding of the possible impact of milder losses, clinicians cannot appropriately advise and parents do not have a firm basis from which to make the best decisions for their child. Section 2.6 of this chapter will examine the literature on management of milder hearing loss.

Section 2.4 of this literature review has defined milder hearing loss as the focus of this PhD study. By explaining how it is inevitable with UNHS that milder degree losses are detected earlier than historically possible, an understanding of milder loss prevalence was gained from the limited existing research. The broader impacts milder loss can have on multiple domains in childhood highlight the potential challenges these children may face as they grow up. The next section of this review switches to the development of language, to initially provide a firm understanding of what should occur during development and then what challenges children with milder hearing loss face in this domain.

### 2.5 Language

Language has been defined as the knowledge of a code used for representing ideas about the world through a conventional system of arbitrary signals for the purpose of communication. This code can be in a spoken, written, or other symbolic (e.g. sign language) form. Throughout this thesis, the use of the term “language” will refer to language in a spoken form. In mainstream society, mastery of spoken language skills confers a lifetime of benefits, maximising communicative opportunities and ultimately leading to a greater likelihood of leading a fulfilling existence. In other words, the importance of developing adequate oral language ability in a society where the primary communicative method is oral cannot be overstated.

Broadly, spoken language can be described across five domains: phonology (rules governing how language is used at the speech sound level), morphology and syntax (or morpho-syntax, how morphemes/minimal meaningful units of
language are used and the rules governing how words are combined to form sentences), semantics (the meaning of words and word combinations in language) and pragmatics (rules pertaining to the use of language in conversation and society). How these pertain to both expressive and receptive spoken contexts are outlined in Table 2.4 (adapted from American Speech-Language-Hearing Association114).

**Table 2.4. Overview of the domains of language**

<table>
<thead>
<tr>
<th>Domain</th>
<th>Receptive (understanding)</th>
<th>Expressive (producing)</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Phonology</strong></td>
<td>Ability to identify and distinguish sounds while listening (phonological awareness)</td>
<td>Correct use of sound patterns while speaking</td>
</tr>
<tr>
<td><strong>Morphology</strong></td>
<td>Understanding morphemes/minimal meaningful units of language</td>
<td>Using morphemes/minimal meaningful units of language correctly while speaking</td>
</tr>
<tr>
<td><strong>Syntax</strong></td>
<td>Understanding sentence structure elements when listening</td>
<td>Using correct sentence structure when speaking</td>
</tr>
<tr>
<td><strong>Semantics</strong></td>
<td>Understanding vocabulary</td>
<td>Speaking vocabulary</td>
</tr>
<tr>
<td><strong>Pragmatics</strong></td>
<td>Understanding conversational exchanges, the social aspects of spoken language</td>
<td>Social use of language, including production of relevant and cohesive messages during conversation</td>
</tr>
</tbody>
</table>

The purpose of this section of the literature review is firstly to outline typical childhood language development and to highlight the role auditory access plays in this development. Next, the evidence for language outcomes in children with hearing loss is reviewed, first generally and then for children with milder losses specifically. Throughout, elements of research design and methodology will be highlighted to illustrate the differences between clinical and population-level results. If the literature shows that degraded auditory input has a negative impact on language outcomes, this would warrant further investigations into management methods used in this population. If on the other hand the literature indicates no negative impact, this would suggest that current management practices are sufficiently addressing their needs.
2.5.1 Language development

It is recognised that elements of language learning, such as vocabulary growth\textsuperscript{115} and semantic and pragmatic development,\textsuperscript{116} continue across the lifespan. However, the first few years of life are when foundations of phonologic and morpho-syntactic language development are laid. It is no coincidence this is believed to also be a time when the developing brain is primed for rapid acquisition of fundamental communication skills for language development. Section 2.5.1.1 chronologically outlines some of the expected milestones of language development, which are often used to monitor rates of language attainment in children across the first years of life. The milestones outlined culminate in a level of development that equips the child for continued language learning.

2.5.1.1 Expected development of language milestones

Foundations of language are acquired during the early years of life. This typically occurs in a predictable manner. In Table 2.5 this development is outlined with an intentional focus on the building of receptive and expressive skills relevant to spoken language.
### Table 2.5. Expected language development across 0-48 months of age

<table>
<thead>
<tr>
<th>Age</th>
<th>Receptive (understanding)</th>
<th>Expressive (producing)</th>
</tr>
</thead>
<tbody>
<tr>
<td>0-12 months</td>
<td>Receptive language skills are expected to precede expressive language, with children showing evidence of word comprehension at 8-10 months.(^{117}) Children respond to their name at around 9-10 months.(^{118})</td>
<td>Typically developing children commencing babbling around 6 months of age, progressing from the canonical form (e.g. “dadada”) to variegated babbling with differing consonant/vowel combinations (e.g. “da ba by doy”) by 8-10 months of age. By one year of age, there is growth in the number of vowel and consonant sounds produced, usually coupled with the first word being produced.(^{117}) Population studies suggest there is a wide range of normal for the first utterance: 33% of 8 month-olds are reported to be using a recognizable word with intent, with this proportion increasing to 77% by 12 months of age.(^{119})</td>
</tr>
<tr>
<td>12-24 months</td>
<td>Comprehension of single words progresses to understanding simple one-step commands. Paired initially with a gesture to increase understanding, this becomes redundant by around 15 months of age.(^{120}) By 24 months of age, children typically understand two-step commands.</td>
<td>Expressive vocabulary grows at approximately 5-10 words per month.(^{121}) A vocabulary spurt occurs following the first 50 words, with an average of 40 new words being spoken per month.(^{122}) Two-word combinations are usually produced as the typically developing child approaches two years of age.</td>
</tr>
<tr>
<td>24-36 months</td>
<td>Children typically understand questions about familiar objects, people and events.(^{123}) By their third birthday, questions seeking meaning are typically understood.(^{124})</td>
<td>In their third year of life, children correctly use questions about familiar objects, people and events.(^{123}) By their third birthday, questions seeking meaning (i.e. “why”) are typically produced.(^{124}) Expressively, three word utterances are produced expanding to full sentences.(^{125}) Speech production is 75% intelligible by 3 years,(^{126}) as children can approximate two-thirds of adult speech sounds.(^{125})</td>
</tr>
<tr>
<td>36-48 months</td>
<td>Expanded capacity for understanding three-step commands and complex concepts including past and future, same versus different.(^{121})</td>
<td>Sentences are produced with grammatically correct multiple clauses(^{125}) including use of conjunctions and tense.(^{123})</td>
</tr>
</tbody>
</table>
Between 4 and 5 years of age, with school entry approaching or commenced, a typically developing child is expected to have sufficiently developed the language fundamentals outlined above in Table 2.5 to facilitate effective communication. Speech production is now expected to be essentially fully intelligible. This development does not stop at 5 years, with continued expansion of expressive and receptive vocabulary and increased complexity of language use throughout childhood and into adulthood. The above outlined milestones will vary between children in the timing of skill acquisition but will generally follow the expected progression.

The next section considers theories behind how this language development occurs. The roles of audition and social influences in language development theories are highlighted. The explanation of the physiological development of hearing provides evidence for how one particular language development theory occurs in parallel (see Section 2.5.1.3).

2.5.1.2 Theories of how language develops

For the majority of children, language develops without the need for explicit teaching and without great effort (see preceding Section 2.5.1.1 for details). It is generally accepted to be the product of both biology and environment, which interact to produce language that is proficient for purpose and yet varies in competency between individuals.\textsuperscript{127} The methods by which children achieve language proficiency are long debated: from the concept of nativism whereby language acquisition is an innate or hard-wired function of the brain,\textsuperscript{128} to the active learning of language through cumulative experience and operant conditioning.\textsuperscript{129} More recent theories move away from these two poles of thought operating in isolation, instead drawing upon combinations of social and neurological constructs to form hypotheses for language learning. One relatively recent hypothesis is the interaction of general human computational ability with social skills to aid early language acquisition: the neural commitment theory.\textsuperscript{130, 131}
The neural commitment theory describes the inherent ability of infants to improve their discrimination of native language speech sounds as they develop. In this theory, auditory input lays the groundwork for neural commitment, with exposure to spoken language the catalyst for the neural mapping of inputs (i.e. auditory stimuli). Early detection of phonetic and prosodic aspects of speech then allows development of sensory patterns (e.g. recognition of auditory cues unique to a language), facilitating further language specialisation. This neural circuitry is believed to be bi-directional, where neural connections are both primed for speech sounds relevant for native language development and perceptually desensitised to non-native speech sounds that do not match the forming language template. Evidence for this neural development is seen in magnetoencephalography studies, where the processing of non-native speech sounds was observed to recruit greater brain resources of increased duration in both hemispheres than processing native language auditory signals.

As a language-learning model, the neural commitment theory suggests that an infant uses statistical learning in combination with social interaction to facilitate their growing proficiency in a primary language. Statistical learning is an example of implicit rather than explicit learning, where individuals identify and learn from statistical regularities in a stream of sensory information (in this case, speech). From a total of approximately 600 consonants and 200 vowels across the world’s languages, any infant is potentially faced with a challenging task to identify the approximately 40 phonemes that comprise their primary language. It is not surprising then that a purely statistical-based model is believed insufficient for language learning and that social interaction appears to be important. This is illustrated in a study where nine-month-old infants exposed to Mandarin via either audio or audiovisual DVD presentations did not display evidence of phonetic learning. This contrasted with the successful phonetic learning of other same-age infants learning Mandarin from a live person. Such results highlight the importance of social interaction in providing positive feedback for aspects of language learning such as phonemic development. The age at which this learning of the foundations of language occurs is also believed important, as discussed below.
Neural commitment occurs early in life and exploits periods of neural sensitivity and plasticity across early development, within the bounds of what was previously referred to as critical periods for the acquisition of skills. These periods may be better referred to as “sensitive” or “optimal” periods. The precise timing in development for optimal periods to learn different levels of language (e.g. phonetic and syntactic) varies and in some cases overlaps. In typically developing children learning only one language, an important period for phonetic learning is before the end of the first year of life, with syntactic development accelerating at between 1.5 and 3 years of age. Further evidence of neural coding occurring early is found in other domains of language development. An example is seen in the work of Travis and colleagues (2011), who employed brain imaging techniques to show that neural mechanisms for encoding lexico-semantic information are laid early, and are followed by refinements into adulthood. Neural pathways observed to be present in 12-18 month-old infants were found to be fundamentally the same brain pathways operating in adulthood. Evidence these neural attributes used to understand words are present in infants who have both a very small lexicon and limited expressive ability is supportive of the theory that foundations for language learning are built early. Further evidence that there are optimal periods for attributes of language development is highlighted in Section 2.5.1.3. This is where, of particular relevance to this thesis, the role of audition in the neural commitment theory is explained. Prior to this, the general role of audition in language is considered.

To capitalise on this innate ability to develop language during the period of time when the brain is primed to acquire language, access to a clear, undistorted auditory signal is required. An auditory feedback loop is essential to assist with correcting the errors made when producing novel speech sounds and learning the rules and intricacies of language. There is evidence that permanent childhood hearing loss can be irreversibly damaging, because it impacts not only on the acquisition of skills that are required for normal communication in an oral environment, but on underlying brain and auditory
development. For example sub-optimal stimulation of auditory cortical pathways occurs in children with hearing loss of less than profound degree, compared to children with normal hearing.\textsuperscript{142} This manifests as extended latencies of evoked recordings, believed to represent synaptic delays across both the peripheral and central auditory pathways resulting from under-stimulation.\textsuperscript{143, 144}

**2.5.1.3 The role of audition in the neural commitment theory**

Kuhl's neural commitment theory includes audition as the primary input for development of spoken language. Audition begins early in life. The human infant's ability to hear is developed as early as approximately 25 weeks gestational age. By this stage of development not only does gross cochlear morphology appear complete and functional (20 weeks gestation\textsuperscript{145}), but success at recording responses from the auditory brainstem suggests adequate completion and connection of associated auditory neural pathways by ages ranging from 25 to 28 weeks gestational age.\textsuperscript{140, 146, 147} At this point in development, a sufficiently strong stimulus can reliably elicit a measurable response from the auditory nerve in a normally hearing foetus.\textsuperscript{146} A hearing infant born at term, therefore, has already been exposed to over 10 weeks of auditory experience.

It is doubtful this intra-uterine hearing experience has been limited to simple sound detection. There has likely been exposure to auditory cues that are important to language development, such as prosody (characteristics of speech at a syllabic rather than individual phonemic level), intonation (the variation in the pitch of spoken words) and rhythm.\textsuperscript{148} Evidence for this early listening rather than early sound detection is supported by the measurement of infant preference, at less than three days of age, for its mother's voice over those voices of other female speakers.\textsuperscript{149} The cries of newborns have been documented to mimic some prosodic properties of the language they were exposed to in utero.\textsuperscript{150}
As highlighted above, audition is crucial to multiple domains of language learning in early life. We know that infants are born as a language blank canvas, possessing the ability to auditorily differentiate and discriminate any speech sound contrast.\textsuperscript{151, 152} This blank canvas aids the task of learning the spoken language relevant to their environment, but does not last a lifetime. Between 8-10 months of age, as monolingual infants’ competency in their native language grows, their ability to discriminate speech sounds unique to nonnative languages becomes redundant.\textsuperscript{136} An example of this was observed in infants aged 6 to 12 months, utilising the /l-r/ contrast in English and the absence of such differentiation in the Japanese language. As these infants developed, regional differences were observed. By 12 months of age, while American infants improved their ability to discriminate between native /l-r/ speech sounds, Japanese infants decreased their nonnative /l-r/ discrimination ability.\textsuperscript{153} This highlights a narrowing of attention by infants towards sounds with functional relevance to their auditory environment. The neural commitment theory suggests statistical learning from auditory stimuli is one component in this narrowing process.

Additionally, it is feasible that audition is important for mediating the role of social factors in the neural commitment theory. It is hypothesised that social inputs are required to effectively harness or “gate” the brain’s computational power, to then be constructive in the task of language acquisition.\textsuperscript{154} In the aforementioned example of Mandarin phoneme learning (see Section 2.5.1.2), it is likely that auditory input alongside social interaction provided by a live instructor rather than a recording-only presentation is advantageous in the progressive building of phonemic categories that are the foundation for learning language. The live instructor provides positive social reinforcement in the use of auditory input.

This hypothesis is of particular interest in the context of this PhD, due to both the supportive speech perception evidence for this language-learning hypothesis and the potential implications for language learning that a degraded auditory input may have.
2.5.2 Language outcomes in children with hearing loss

The preceding sections on language development, including theories of how language develops and the role of hearing, have led to this section reviewing the language outcomes achieved by children with hearing loss. In this section, the research highlighted is that of large-scale population-based Australian studies. These outline the general trends in outcomes of children with hearing loss in an environment where age of detection and subsequent intervention is trending downwards. Of note, the two studies considered do not emphasise the outcomes of children within specific degrees of loss, nor do they focus on milder losses. Instead, they consider hearing loss represented as a continuous variable. Those studies that do consider language outcomes in children specifically stratified by degree of loss will be considered in Section 2.5.3 of this thesis.

The Longitudinal Outcomes of Children with Hearing Impairment (LOCHI)\textsuperscript{155} and Statewide Comparisons of Outcomes (SCOUT)\textsuperscript{156} studies were conducted in parallel across multiple Australian states when congenital hearing loss detection across the population was routinely occurring earlier than ever before. Descriptive details of the two studies are found in Table 2.6. This was a period when research interest was focused on the potential benefits in outcomes of those detected earlier versus later.

Notably, these two large-scale and population-based studies have provided greater insight into the impact of earlier hearing loss detection. LOCHI is a longitudinal study of 451 children amplified for mild to profound hearing loss, of whom approximately half were fitted with hearing aids before age 6 months. It examined outcomes following birth in one of three Australian states across which there was a lack of uniformity in hearing loss detection systems. With assessment occurring at 6 and 12 months post amplification fitting and again at age 3 and 5 years, results indicated those fitted earlier in life showed strong evidence of better language outcomes at age 5 years upon comparison to those fitted later.\textsuperscript{157} LOCHI assessed these children on a range of language measures to
determine a composite language score. This score indicated the outcomes of children with hearing loss were up to 1.5 standard deviations below population normative values at age 3 years, and at both the 3 and 5 year old assessments a child’s increasing degree of hearing loss was increasingly negatively associated with their outcomes.\textsuperscript{158} Common to both preschool age assessments was the significant influence of poorer maternal education and presence of comorbid conditions in depressing the child’s language outcomes.

The cross-sectional SCOUT population-based study, which exploited the secular trends in systems of congenital hearing loss detection across two Australian states, found results of a similar nature to LOCHI, yet in older children. At around school entry, Wake and colleagues (2016)\textsuperscript{156} showed the decrease in diagnosis age afforded by UNHS led to clear benefits in expressive language and receptive vocabulary skills exhibited in a sample with hearing loss ranging mild to profound. Similar to LOCHI, these children were also all fitted with amplification and results excluded those with intellectual disability. No impact of earlier detection was observed on child behaviour or quality of life, and consistent with the LOCHI study a general trend toward lower language and vocabulary scores was observed as degree of loss increased. Notably, overall language performance of children with hearing loss was still poor compared to population norms and the nonverbal cognitive potential of these children.\textsuperscript{156}

These two studies\textsuperscript{156, 158} provide contemporary evidence of the impact of systematic earlier detection of congenital hearing loss on the language outcomes of affected children. By looking across the spectrum of hearing loss from mild to profound, trends in outcomes are observed, such as the correlations between lower age at detection and better outcomes, and conversely the correlation between increasing degree of loss and poorer outcomes. Whilst Ching and colleagues (2013)\textsuperscript{159} predominantly split their sample at age 3 years into those fitted with hearing aids versus cochlear implants to examine the impact of alternate amplification strategies, Wake and colleagues (2016)\textsuperscript{156} used different statistical methods to examine outcomes by degree of loss. Wake et al (2016)\textsuperscript{156} found earlier diagnosis led to greatest improvement in receptive vocabulary for
children with severe loss, and receptive language for profound losses. These results make intuitive sense, whereas the lack of a severity relationship on expressive language skills is more difficult to explain.

What neither the LOCHI nor SCOUT study do is look comprehensively at the outcomes of children detected with milder hearing losses. With children being diagnosed earlier than ever before with these degrees of loss, the literature examining the impact of earlier detection on outcomes requires exploration to determine whether the general outcome trends seen in large population-based studies are seen in studies that specifically focus on a defined degree of loss sample. This literature should be able to distinguish the effect of greater degrees of loss on any of these general trends noted thus far.
<table>
<thead>
<tr>
<th>Study</th>
<th>Subject groups</th>
<th>Inclusion Criteria</th>
<th>Outcome Measures</th>
<th>Findings</th>
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<tr>
<td>Ching et al (2010, 2013, 2017)</td>
<td>At age 3 years &lt;br&gt;n=451 children, comprising: &lt;br&gt;n=239 first fitted with hearing aids &lt;6 months of age. &lt;br&gt;n=356 assessed</td>
<td>Permanent bilateral loss born between 2002-2007 and accessing services from Australian Hearing in NSW, Vic or southern QLD under age 3 years. Loss defined by 4FAHL (0.5,1,2,4kHz), 98.6% of sample fitted with amplification at age 3 years.</td>
<td>Receptive and expressive language: Preschool Language Scale – 4&lt;br&gt;Receptive vocabulary: The Peabody Picture Vocabulary Test&lt;br&gt;Speech production: Diagnostic Evaluation of Articulation and Phonology&lt;br&gt;Child language and psychosocial skills: Child Development Inventory, Parents’ Evaluation of Aural/oral performance of Children (PEACH).</td>
<td>At age 3 years&lt;br&gt;Global language development 1.5 standard deviations below population norms. Significant predictors of poorer outcomes included:&lt;br&gt;• greater severity of hearing loss,&lt;br&gt;• male gender,&lt;br&gt;• presence of additional disabilities,&lt;br&gt;• lower maternal education,&lt;br&gt;• older age of cochlear implantation.&lt;br&gt;Age at amplification showed a weak non-significant impact on outcomes.</td>
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<td>At age 5 years &lt;br&gt;n=472, comprising: &lt;br&gt;n=384 with non-progressive loss. &lt;br&gt;n= 350 assessed</td>
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<td>At age 5 years&lt;br&gt;Increasing benefit from earlier intervention with increasing loss. Children fitted with amplification at 24 months, versus 3 months, had language outcomes:&lt;br&gt;• 11.8 score points poorer for 70dB HL losses (95% CI: -18.7 to -4.8)&lt;br&gt;• 6.8 score points poorer for 50 dB HL losses (95% CI: -10.8 to -2.8).&lt;br&gt;Early performance measures taken at under 2 years (parent-reported PEACH, child PLS assessment) significantly predicted language development at 3 and 5 years.</td>
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<tr>
<td>Study</td>
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| Wake et al (2016)\textsuperscript{156} | Population-based quasi-experimental study (SCOUT) | n=220, comprising:  
Aged 5-6 years  
n=69 born during period of UNHS (NSW)  
n=65 born during risk factor screening (Vic)  
Aged 7-8 years (existing population cohort)  
n=86 born when detection was opportunistic (Vic) | Permanent bilateral loss believed congenital >25 dB HL in better ear calculated by 3FAHL (0.5,1,2kHz), fitted with hearing aids or cochlear implants by age 4 years. NB: 37% of sample was already participating in the LOCHI study. | Receptive and expressive language: Preschool Language Scale – 4  
Receptive vocabulary: The Peabody Picture Vocabulary Test  
Letter knowledge: Phonological Abilities Test  
Nonverbal cognition: Wechsler Nonverbal Scale of Ability  
Health-related quality of life: Health Utility Index 3 parent self-report, parent-proxy child report. Pediatric Quality of Life Inventory 4.0  
Behaviour and emotion: Strengths and difficulties questionnaire: Aust version for 4-10 year olds | Children were diagnosed younger if born during UNHS versus risk factor screening (adjusted mean difference -8 months, 95% CI -12.3 to -3.7).  
Excluding children with intellectual disability, statistically significant improvements seen between children born during opportunistic detection, to risk factor to universal screening in the following areas:  
• Age at diagnosis (22.5 vs 16.2 vs 8.1 months, p<.001)  
• Receptive language (81.8 vs 83.0 vs 88.9, p=.05)  
• Expressive language (74.9 vs 80.7 vs 89.3, p<.001)  
• Receptive vocabulary (79.4 vs 83.8 vs 91.5, p<.001)  
No significant difference in parent reported child behaviour and health-related quality of life.  
Performance of children with hearing loss remains lower than cognitive potential (mean 10.3.4, SD 15.2). |
2.5.3 Language outcomes in children with milder hearing loss

This section of the literature review specifically examines what is understood of language development outcomes for children with milder (mild and moderate) hearing loss. Outcomes for two separate age groups of children are considered; those in the early primary school years and those of early preschool age. These age groups encapsulate two periods where language development is of particular interest; to maximise the child’s learning potential with entry to formal education and to monitor the early consolidation of language skills, respectively. In other words, the difference between these two age groups is the shift in focus from learning language, to using language to learn.160 These two age groups also match the two populations examined in this PhD study. Considered are the different ways these studies have quantified hearing loss and the impact on the ability to generalise their conclusions. Studies that do not delineate results by degree of loss are excluded, as the outcomes achieved by children are highly likely to vary by hearing loss severity (as demonstrated by the studies covered in Section 2.5.2). The trends observed in outcomes of population-based versus clinical/convenience samples and early versus late identified losses are examined to illustrate the potential effect of research methodology on published findings.

2.5.3.1 Early primary-school language outcomes in children with milder loss

The ideal study design at this age group would be a large population-based study of children early detected with milder loss. This would likely be achieved through the activities of UNHS. These children would be assessed on standardised language tasks that allow comparison to comprehensively normed expected scores, in addition to comparison with normally-hearing peers drawn from the same population. Measures of hearing would cover both unaided and aided hearing ability, to incorporate the impact of amplification on outcomes. These study requirements would, if able to be met, allow for the greatest generalisation of contemporary outcomes for the children of interest that is representative of the broader population.
Table 2.7 outlines papers focusing on the language outcomes achieved by primary-school aged children with mild-moderate bilateral hearing loss. These studies either specifically sampled mild and/or moderate losses, or delineated the results of children with these degrees of loss from other loss types and severities. All studies defined their participant hearing losses using a variant of the PTA calculation. Other studies exist but are not included as they do not specifically report the outcomes of children with hearing losses of interest in isolation. Instead, they group children with mild and unilateral losses\(^8^4,1^6^1\) or report mild and moderate outcomes as trends rather than discrete interpretable results.\(^1^5^6,1^5^9,1^6^2\)

The pattern of language outcomes seen in samples of children with slight/mild losses was of isolated difficulties in aspects of language processing, perhaps an indication of an impairment but not a broader language disability. At the mildest end of the severity spectrum in the literature considered, Wake et al (2006)\(^9^0\) reported children with slight/mild losses had significantly weaker skills in phonological processes in comparison to peers without hearing loss. Specifically, these tasks involved the storing and recalling word sound structures and discriminating between different sounds in words. These difficulties were measured despite overall expressive and receptive language skills being comparable to those with normal hearing.\(^9^0\) However, this study recorded a mean PTA of 22.4 dB HL in their population-based study of participants with hearing loss, indicating a very mild hearing impairment. Therefore, it is not entirely surprising that such an isolated impact to specific language skills highly reliant on auditory cues was the only domain to indicate deficits in this sample of children who did not wear hearing aids.

Those studies that reported mild or moderate outcomes from a larger cohort covering the broader spectrum of hearing loss observed a pattern of poorer language outcomes with increasing degree of loss.\(^1^0^7,1^6^3,1^6^4\) For example, Wake et al\(^1^0^7,1^6^3\) observed the language outcomes achieved by children at 7-8 years of age with mild loss ranged from approximately one third to close to one standard deviation below the expected performance levels on standardised assessment of
language and receptive vocabulary. Tomblin et al (2015)164 noted significantly poorer language outcomes of children with severe loss compared to mild loss, but interestingly no significant difference between children with mild and moderate losses. These two groups of children were scoring across the ages 2-6 years at either close to the expected standardised language score (mild loss) or at one third of a standard deviation below expectations (moderate loss). A similar result was seen in a Swedish study165 of 4-6 year old children with mild to severe loss. These researchers noted language problems were measured as participant PTA reached the early moderate level (from 50 dB HL onwards).165 It is important to note that such performance was measured against standardised scores. When scores are compared to those achieved by peers without hearing loss from the same population, the children with hearing loss are performing more poorly than reliance on the test normative score would suggest.164 This highlights the importance of an understanding of expected performance on language tasks drawn from a similar population to the actual participants of interest.

It is possible that the trend toward poorer language performance with increasing severity of PTA described above is influenced by the performance of children with greater degrees of loss in the sample. These children with severe losses may skew the overall performance trajectory. When considering studies comprised solely of children with mild-moderate loss, there are examples of language outcomes not correlating with increasing severity of PTA.166,167 Alternatively, the broad heterogeneity in language skills is observed by studies that focus solely on the language outcomes of children with mild-moderate loss166,168,169 may simply reflect the impact of a range of inherent language skills in children that are independent from influence of their degree of hearing loss. Factors such as cognitive ability, which may be influencing the language skills displayed in these children, are explored in more detail in Section 2.6.3.

Regarding the design of the included studies, the majority were clinical/convenience samples and the majority of participants were not early identified via systematic UNHS activities.84, 90, 107, 163, 166-169 The proportion of
participants who were early detected within individual studies also varied. For example, approximately 75% of participants from the Outcomes of Children with Hearing Loss (OCHL) study were detected (via UNHS activity) with a mean age at diagnosis of 7.34 months.\textsuperscript{111,164} This is contrasted with another study where 3% of their population was early-diagnosed at under 1 year of age despite being a non-UNHS sample.\textsuperscript{165} The lack of studies including children with very early-diagnosed milder losses, plus the potential bias from convenience sampling, limits the ability to generalise these studies’ findings to the contemporary early-detected general population. As stated above, a study would ideally be conducted using a sample of UNHS-detected children derived from the broader population. This design would both reflect the current trend in earlier population-wide hearing loss detection and reduce the risk of a highly selected sample through a population-based approach to recruitment.

Two studies in Table 2.7, both drawn from the OCHL study, used the Speech Intelligibility Index (SII) in addition to the conventional PTA to interpret their participant language skills. The SII (see Section 2.2.4.2 for details) incorporated the impact of aided audibility via hearing aids on language outcome trends. Using the SII, a link has been established between improved speech/language development and children experiencing improved audibility via well-fitted hearing aids. This was a result observed even in children with the lowest degree of impairment, i.e. mild hearing losses.\textsuperscript{170}

The OCHL study concluded that optimally fitted hearing aids reduced the risk of language delay in children aged 2-6 years with hearing losses spanning mild to moderately-severe. Children who had greater aided audibility also experienced a faster rate of language growth as measured by a composite language score.\textsuperscript{160} In school-aged children with PTA ranging normal to profound, Stiles and colleagues (2012)\textsuperscript{171} showed those with higher aided audibility, quantified via the SII, achieved better language outcomes than those with a poorer SII. The authors also showed that aided SII was more strongly associated than the PTA with performance on receptive vocabulary and word/non-word repetition tasks. These results were reported despite limited ability to account for the influence of
confounding factors due to sample size restrictions. Also, Stiles and colleagues (2012) did not report outcomes specifically at the mild or moderate loss level, which limited the conclusions that can be made about the value of the SII measurement at interpreting the language outcomes within this milder range of losses.

In summary, from the included studies of children with milder losses in the early primary school years, there are mixed reports of their language outcomes. There is some evidence of very limited impact on outcomes when the degree of loss is slight/mild, and mixed evidence for the impact as degree of loss increases to the mild-moderate level. This is likely a reflection of differences in study design, covering aspects of sampling, analyses and the inherent variability of language abilities in children. There is a growing body of work that looks at using alternatives to the PTA as methods of quantifying hearing ability and relationships between hearing ability and language performance.
### Table 2.7. Primary school language outcomes of children with milder loss

<table>
<thead>
<tr>
<th>Author (country)</th>
<th>Population characteristics</th>
<th>Study design</th>
<th>Audiological measures and language outcome domains</th>
<th>Outcomes/conclusions</th>
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<tr>
<td>Gilbertson &amp; Kamhi (1995) (USA)</td>
<td>n=20 mild-moderate loss aged 7:9 to 10:7 years. All aided. Mean 3FAHL 42 dB (SD 21 dB). n=20 normal hearing comparison group aged 5:1 to 9:7 years. Non-UNHS sample</td>
<td>Cross-sectional. School sourced convenience sample – Memphis and Arkansas teaching body.</td>
<td>Aud: 3-5 frequency PTA Lang: Novel word acquisition, recognition and retention, phonological processing.</td>
<td>50% of hearing loss sample achieved comparable levels on all tasks to children with normal hearing. Remaining 50% performed at lower levels on measures of word acquisition, general language and multisyllabic phonological processing. Labelled low-functioning children as language impaired, as cognitive functioning fell within normal age limits. Degree of hearing loss not significantly related to language performance or ability to learn words. Potential influence of socio-cultural factors, 7 of 9 African-American children in lower performing group.</td>
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<tr>
<td>Author (country)</td>
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50% of hearing aid users rated use as poor, very poor/not at all users, or missing data  
Lang: rec/exp vocab, auditory memory, grammatical skills. | Some delay in the age of development of two word utterances observed for all mild losses (conductive and sensorineural, unilateral and bilateral).  
Receptive and expressive vocabulary size in children with hearing loss without significant comorbidities did not differ from those seen in normally hearing children.  
Receptive vocabulary T score (n=14 mild bilateral sensorineural) 53 (SD 14.8), range 23-67.  
Expressive vocabulary T score 56 (SD 8.6), range 50-71.  
Nonverbal IQ 53.9 (SD 8.1), range 44-64. |
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| Wake et al (2004/05)   | n=86 mild to profound aged 7-8 years (n=19 mild, n=27 moderate). All aided by 4.5 years. No major physical or intellectual disability. 67% response rate. Referenced to normative scores, mean 100, SD 15. Non-UNHS sample. | Cross-sectional. Population-based from entire geographic state. | Aud: 3-frequency PTA  
Lang: rec/exp lang, rec vocab  
Sp: articulation  
Other: nonverbal intelligence, health related QoL, behaviour, school functioning, development | Children with hearing loss show major deficits in language and schooling areas assessed. Significant association between poorer PTA and poorer performance on measures of total language (p <0.001) and receptive vocabulary (p<0.001). Children with mild loss had total language scores one third SD lower than population norm (mean score 94.8, standard error 3.9), receptive vocabulary 0.8 SD lower (mean score 87.5, standard error 3.5). Children with moderate loss were over 1 SD poorer than population norm on both total language and receptive vocabulary measures. Severity of loss at diagnosis, at assessment and nonverbal IQ were correlated with most speech/language measures. Age of diagnosis did not influence language scores, after adjustment for severity of loss and nonverbal IQ. |
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<tr>
<th>Author (country)</th>
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<tr>
<td>Wake et al (2006) (Aus)</td>
<td>n=6240 children in 1st (age 7.2 years) and 5th (age 11.1 years) grade of schooling.</td>
<td>Cross-sectional cluster sample survey.</td>
<td>PTA: 3-frequency PTA at either low (0.5,1,2 kHz) or high (3,4,6 kHz) frequency range.</td>
<td>Significantly poorer performance by children with slight/mild loss on measure of phonologic short-term memory (mean difference -11.4 points; 95% CI -16.8 to -6.1; p&lt;0.001).</td>
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<td>Prevalence survey for slight/mild loss (better ear 16-40 dB HL).</td>
<td>Population-based.</td>
<td>Lang: rec/exp lang, phonologic short term memory, reading ability, phonological awareness and discrimination.</td>
<td>Significantly poorer phonologic discrimination (scores &lt;90%) by children with hearing loss compared to controls (18.8% versus 5.2%; odds ratio of 4.0 after adjustment for non-verbal IQ and matching; 95% CI 1.6 to 10.3, p=0.004).</td>
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<td>85% response rate.</td>
<td>89 schools participated from one geographic state.</td>
<td>Other: nonverbal intelligence, health-related QoL, behaviour, school learning measures.</td>
<td>No significant difference in performance between children with slight/mild and normally hearing peers on remaining language and other measures.</td>
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<td>n=55 (0.88% of sample) identified with slight/mild loss (mean PTA 22.4 dB HL). Slight n=39, mild n=16.</td>
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<td>Referenced to normal hearing controls (peer matched).</td>
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<td>Unaided sample.</td>
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<td>Non-UNHS sample.</td>
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<td>Author (country)</td>
<td>Population characteristics</td>
<td>Study design</td>
<td>Audiological measures and language outcome domains</td>
<td>Outcomes/conclusions</td>
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<tr>
<td>Borg et al (2007) (^{165}) (Sweden)</td>
<td>n=156 Swedish speaking children with better ear hearing loss up to 80 dB HL, aged 4-6 years. No significant diagnosed comorbidities. 59% response rate. Bilateral and unilateral losses included. Bilateral losses divided into four 20 dB severity groups. Approx. 50% hearing aid users, remainder unaided. n=97 controls</td>
<td>Cross-sectional. Population-based clinical sample, all children in national hearing care system aged 4-6 years.</td>
<td>Aud: 3-frequency PTA Lang: 9 tests covering sensory processes, language processes and speech motor functions. Includes phoneme discrimination, rec vocabulary, measures of grammar, word finding expressive vocabulary test.</td>
<td>Range of abilities observed amongst children with milder losses; normal language scores observed for some children. Word finding expressive vocabulary test performance most impacted by hearing loss. Expressive delays estimated at 1.5-2 years for children with losses 41-80 dB HL. Prosodic phrase repetition showed decreasing initial stress accuracy with increasing degree of loss: 91% correct for children with normal hearing, 84% for children with losses 21-60 dB HL, 61% for children 61-80 dB HL. Language problems became apparent with PTA reaching 50-60 dB HL. Children with greater degrees of loss showed greater use of hearing aids (rho=0.35, p&lt;0.001).</td>
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<tr>
<td>Author (country)</td>
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<tr>
<td>Dokovic et al (2014)(^{169}) (Serbia)</td>
<td>n=144 children with mild bilateral loss (20-40 dB HL) mean age 8.85 years (range 7.5-11 years).</td>
<td>Cross-sectional. School health check based in city of Belgrade.</td>
<td>Aud: 4-frequency PTA Standardised tests of perceptual, verbal and nonverbal abilities.</td>
<td>Children with mild loss had poorer phonological short-term memory (mild loss mean score 10.22 SD 3.24, normal hearing 11.25 SD 3.24, p&lt;0.001) and phonological discrimination ability (mild loss mean score 14.61 SD 5.43, normal hearing 16.31 SD 4.75, p&lt;0.001) than children with normal hearing.</td>
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<td></td>
<td>Unaided, no access to hearing aids.</td>
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<td>Adapted for use in Serbia, designed to assess adequacy of existing skills for academic learning.</td>
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<td></td>
<td>n=160 age matched controls with normal hearing.</td>
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<td>Non-UNHS sample.</td>
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<tr>
<td>Tomblin et al (2014)(^{170}) (USA)</td>
<td>n=180 children aged 3 (n=74) and 5 years (n=106) with mild (26-45 dB HL) to severe bilateral loss.</td>
<td>Observational cross-sectional. Multi-centre clinical sample.</td>
<td>Aud: 3 or 4-frequency PTA, SII Lang: rec/exp lang, Speech: Articulation ability</td>
<td>Children with mild loss had significantly better speech articulation ability than those with moderate-severe loss (2 age groups combined: mild loss mean 95.36 SD 15.01; moderate-severe mean 86.60 SD 18.04; p&lt;0.001). Overall composite language performance of both categories of children was similar to that expected for children with normal hearing (mild loss Z score mean 0.26 SD 0.87, moderate-severe loss -0.08 SD 0.97). Benefits of hearing aids were similar between mild and greater losses.</td>
</tr>
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<td></td>
<td>Divided into two categories: mild (n=76) and moderate to severe (n=104).</td>
<td></td>
<td>Composite language score derived from all assessment results.</td>
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<td></td>
<td>All but 4 children fitted with hearing aids.</td>
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<td>Referenced to population norms, mean 100 SD 15.</td>
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<td></td>
<td>Partial UNHS sample.</td>
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<td>Author (country)</td>
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<tr>
<td>Tomblin et al (2015) (USA)</td>
<td>n=290 children with mild to severe bilateral loss. Aged 2-6 years. n=112 normal hearing controls. Partial UNHS sample.</td>
<td>Accelerated longitudinal design. Multi-centre clinical sample.</td>
<td>Aud: 4-frequency PTA, SII Composite language score derived from all assessment results. Mean 100, SD 15.</td>
<td>At all ages overall language ability of children with hearing loss was significantly poorer than children with normal hearing. Mean scores of children with hearing loss were often less than 0.5 SD lower than expected score. Trend observed of increasing PTA significantly associated with decreasing language scores. However, no significant difference in performance was observed between children with mild and moderate loss.</td>
</tr>
</tbody>
</table>
2.5.3.2  **Preschool language outcomes of children with milder losses**

The ideal study design for this younger population would be a population-based longitudinal study. Early identified infants would be recruited via a system that is independent of their pathway post-diagnosis (e.g. amplification, early intervention enrolment, watchful waiting). This recruitment system would be accessible to the broad community, with participation by families with diverse levels of economic means and parental education. If this system achieved substantial geographic coverage, an adequate sample size to allow for potential confounding variables would be a possibility, as would a comparison of outcomes between those who have chosen different interventions (e.g. hearing aid use). The age group of interest mandates contribution of data from parent-report measures, ideally using well-validated and comprehensively normed tools. As the children develop, a greater use of directly assessable outcomes could be measured to document changes in performance across consistent domains over time. Access to children without hearing loss from the same general population would provide a point of reference for performance that is experienced by peers with comparable demographics.

More studies of preschool aged children than primary school aged children were derived from UNHS samples. Such studies could potentially be more representative of outcomes achieved under current trends in hearing loss detection and intervention practices in developed countries. Other factors influencing the ability to generalise from any samples, including UNHS-derived samples, are considered below. For those studies where the language outcomes of children with mild and moderate loss could not be clearly distinguished from other degrees of loss, trends toward better outcomes with lesser degree of loss/earlier amplification were seen in either specific domains such as speech sound production skills\(^{173}\) or in general composite language scores.\(^{157}\) However, this was not always observed, with Stika et al (2015)\(^{98}\) reporting that degree of loss was not strongly associated with language skills and Koehlerling and colleagues (2013) only observing a small contribution of degree of loss to grammatical development in children aged 3 and 6 years.\(^{162}\) As noted in Section 2.5.3.1, the OCHL cohort shows across the age range 2-6 years (which overlaps
both of this PhD’s age groups) that children with mild loss do not have significantly different language outcomes on a composite language score than children with moderate loss who are performing at lower than expected levels.164

Amongst the small number of studies where preschool language outcomes could be discerned for mild and moderate losses (see Table 2.8), important study design limitations place caveats on the outcomes. Recruiting children from Early Intervention services, whilst convenient especially at preschool age, likely increases the risk of bias toward a skewed sample of children either doing well or poorly and thereby reduces generalisability. Those children performing well in early intervention may be more likely to remain enrolled rather than seek an alternative intervention strategy. At the other end of the ability range, children experiencing poorer than expected development may be more likely to remain engaged in an intervention service. Fulcher et al’s (2012)174 study of language outcomes at 3, 4 and 5 years of age is one potential example, with speech outcomes better in children with severe and profound loss rather than mild and moderate. In the same study, mean language scores for children with mild and moderate loss were consistently higher than expected normative values. This may be a reflection of another recognised limiter on generalisability, with all highlighted researchers acknowledging the advantaged and highly educated families in their samples.174-176

Fitzpatrick et al (2015)176 noted mean language development in a sample of children at 3 and 4 years with minimal (mild bilateral and unilateral) loss as being no different from the development seen in normal hearing peers. This study was reliant on parent-reported outcome measures, completed by parents recruited from Early Intervention centres. Whilst parent reported outcomes are an accepted and common method of obtaining information on young children’s development, participating families from an early intervention program may not represent the general experience of children with hearing loss in the broader community. As noted previously (see Section 2.2.4), the lack of a consistent separation of outcomes for children with mild bilateral and unilateral losses
contributes to difficulties in drawing conclusions specifically for children with bilateral hearing loss.

Vohr et al.'s study of the effect of very early intervention on language outcomes for children with all degrees of loss recruited participants via the local UNHS program records. Whilst this population approach is independent of engagement with Early Intervention and would have a better chance at providing a more representative sample than a sample of an Early Intervention service only, a response rate at just over 50% may indicate some bias in the parents who chose to participate. A small sample size and combining mild bilateral losses (n=2) with unilateral losses (n=10) into a minimal group for analysis purposes places restrictions on generalisability for the less represented bilateral mild losses.

In summary, variation in experimental design is a common factor influencing the limited literature available on language outcomes of preschool children with milder hearing losses. This presents a challenge in drawing general conclusions on how these children are performing. For preschool children with mild loss whose parents are well-educated and have access to early intervention programs and resources, outcomes are reported as comparable to expected performance levels and scores of peers without hearing loss. The outcomes of children with moderate losses have been reported as not significantly different to those with mild loss. It is unclear whether these children would have performed as well as they have if they were not early detected, rather as a result of factors independent of their hearing loss.

This section of the literature review has examined the language outcomes of children with milder hearing loss at two age groups. The following section of this chapter continues the focus on milder hearing losses, shifting towards the current management of children born with these losses.
Table 2.8. Preschool language outcomes of children with milder loss

<table>
<thead>
<tr>
<th>Author (country)</th>
<th>Population characteristics</th>
<th>Study design</th>
<th>Audiological measures and language outcomes</th>
<th>Outcomes/conclusions</th>
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</thead>
<tbody>
<tr>
<td>Vohr et al (2008/2011)</td>
<td>n=30 children with hearing loss (n=18 moderate to profound, n=12 minimal loss comprising 10 unilateral, 1 mild bilateral auditory neuropathy, 1 mild bilateral). 76% of sample amplified by 18-24 month assessment. No implant recipients. Child with mild loss aided at 6 months. n=96 normal hearing controls UNHS cohort, 52% response rate, face-to-face interviews conducted.</td>
<td>Prospective longitudinal age matched cohort study</td>
<td>AUD: calculation method undefined, loss status obtained from state UNHS program. Lang: expressive vocabulary (parent report)</td>
<td>At 12-16 months, children with minimal loss had expressive vocabulary skill similar to normal hearing controls (words produced raw score, SD: minimal loss 20, SD 32; control 17, SD 25). At 18-24 months, children with hearing loss had fewer words than children with normal hearing. Those children with minimal loss enrolled in early intervention at less than 3 months of age showed greatest growth in vocabulary between 12-16 and 18-24 months (142 ±161 words). Children with moderate to profound loss enrolled after 3 months of age showed the slowest growth (22 ±29 words).</td>
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<td>Fulcher et al (2012)</td>
<td>n=94 children with hearing loss mild to profound comprising: n=45 early identified (&lt;12 months), with n=24 mild/moderate loss n=49 late identified (&gt;12 months and &lt;5 years of age), with n=34 mild/moderate loss. Amplified sample. Partial UNHS cohort.</td>
<td>Early Intervention service sample</td>
<td>AUD: calculation method undefined. Lang: rec vocab, rec/exp lang Sp: articulation test</td>
<td>At 3 years of age, speech outcomes of severe/profound group (M=107.43, SD=9.15) significantly better than both mild (M=93.15, SD=10.29) and moderate group (M=97.2, SD=10.12, p&lt;0.001). At 4 years of age, severe/profound group speech outcomes (M=109.81, SD=4.17) significantly better than mild group (M=97.0, SD=9.81, p=0.001) and moderate group (M=97.83, SD=11.74, p=0.05). No difference observed in mean scores between mild and moderate groups. At age 3 years, total language ability of severe/profound group (M=102.17, SD=11.33) was significantly poorer than mild group (M=110.58, SD=9.71, p=0.026). By 4 and 5 years of age, language outcomes were not significantly different between the different degrees of loss. Mean language scores for mild and moderate losses were consistently above expected normative values.</td>
</tr>
<tr>
<td>Author (country)</td>
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<td>Fitzpatrick et al (2015) (Can)</td>
<td>n=55 children with minimal hearing loss (n=24 bilateral mild, n=31 unilateral). Mean mild bilateral hearing loss 32.4 dB HL. Amplification recommended for 80% of sample with minimal loss n=45 normal hearing controls. UNHS cohort, mean identification age 4.2 months.</td>
<td>Cross-sectional, recruitment via Early Intervention centres.</td>
<td>AUD: mild defined as PTA 20-40 dB HL at ≥2 frequencies above 2000 Hz. Lang: mean length of utterance, expressive language, language comprehension. Aud: Auditory development measures (all parent report)</td>
<td>Mean parent report language development at 2 and 3 years of age of children with minimal loss did not differ from normal hearing peers. No results available for solely mild bilateral losses.</td>
</tr>
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</table>
2.6 Management of milder losses

As discussed in Section 2.4, current management of milder hearing loss is influenced by earlier detection facilitating hearing aid fittings at younger ages than ever before. Contemporary audiological practice is designed around the tenet that well-fitted amplification can mitigate the impact of hearing loss on auditory skills crucial for spoken language development.²⁴, ¹⁷⁸ When hearing losses are detected and amplified early, this practice has resulted in children born with severe and profound loss displaying consistent gains in language outcomes compared to those detected later.¹⁵⁶, ¹⁵⁷ This apparent benefit of early amplification is less consistently reported for children with mild and moderate loss.

The estimated impact of milder losses, whilst not as significant as severe and profound, is nonetheless of interest. A mild bilateral hearing loss may result in a child missing anywhere from 25% to 50% of speech sounds, dependent upon listening conditions (i.e. background noise levels), distance from the speaker and audiometric threshold configurations.¹⁰¹, ¹⁷⁹-¹⁸² Further limitations on access to the speech signal would be experienced by children with untreated moderate hearing loss, for whom spoken communication may be mostly or entirely inaudible unless presented at a raised level or within a close proximity.¹⁸¹ Thus despite a lack of explicit research evidence, early amplification for moderate hearing loss has long been advocated for and forms part of routine service provision. Less consensus exists for the early amplification of mild losses. There is historically a lack of understanding of factors that contribute to outcomes in children with mild loss,¹⁸³ meaning the decision to fit amplification is less clear.¹⁸⁴, ¹⁸⁵

This section of the literature review firstly outlines current practice in amplification fitting both in Australia and internationally. Theories as to why this management practice may occur despite a lack of outcomes-based evidence of efficacy follows. This then leads into an exploration of what evidence exists for efficacy of amplification specifically for milder losses. Finally, other factors that may play a role in management of mild loss are considered.
2.6.1 Current management practice for milder losses

Management of moderate hearing loss is considered relatively straightforward, with clear recommendations for early amplification. Moderate losses, alongside greater degrees of loss, are specifically included as the target for UNHS programs (see Section 2.3.4). Amplification guidelines in Australia and internationally recommend early hearing aid fitting.\textsuperscript{24, 186, 187} Enacting these recommendations is grounded in the educated belief that children with moderate losses benefit from early amplification. There is little conjecture on this, therefore, the focus in the remainder of this section of the literature review is specifically on mild hearing loss.

Patterns of amplification provision for mild hearing loss are more varied than for moderate loss. Options range from watchful waiting\textsuperscript{54} until there is clear evidence of difficulties that may be assisted via amplification, through to the routine provision of amplification of children with mild loss.\textsuperscript{188} Providing frequency modulation systems (which can assist in hearing over distance or in background noise) is a recognised option for children with mild loss, but typically occurs when children are older, are in an educational setting, or are observed to be having specific situation-based difficulties.\textsuperscript{186} There exist guidelines which advocate for amplification being considered for children with mild loss\textsuperscript{187, 189} without specific mention of age at which this should occur. A Canadian amplification protocol suggests due to risks of academic difficulty, infants with a loss of at least 30 dB HL are potential amplification candidates.\textsuperscript{24} A frequently noted delay between a mild loss diagnosis and when amplification is subsequently fitted, in the absence of research stating this is the correct management method, may suggest uncertainty around the benefit of early intervention using hearing aids.\textsuperscript{54, 188}

The Australian paediatric amplification protocol highlights the variable impact of mild and unilateral hearing loss on individuals as one reason for parents deciding, alongside the advice from their audiologist, whether to amplify a newly diagnosed infant.\textsuperscript{186} In Australia, fitting of hearing aids appears to be occurring more commonly than previously observed, with pronounced spikes in the
number of first fittings for children with a better-ear mild loss both at under 1 year of age and in the early primary school ages of 4-7 years. This may be influenced by the restricted access to other management options for children with mild losses in Australia, as outlined below.

In Australia, management options at the mild end of the hearing loss spectrum are limited. Unlike those with moderate hearing loss, children with mild loss are ineligible for government-funded early intervention education services as their audiogram does not exceed an average hearing loss of 40 dB HL across the frequency range 0.5-4 kHz. The effect of this restriction is that families of a child with mild hearing loss would need to privately pay for any educational services, unless they qualified for funding via another avenue (e.g. their child has a fundable co-morbidity). This limits the options for no direct cost management options for children with mild hearing loss to the fitting of amplification. The current arrangement in Australia is for government funding of hearing devices for those aged 0-26 years of age. Therefore, it is possible that children born with mild hearing loss are only able to access no-cost hearing devices, without access to early intervention education services. Ineligibility for support services has been hypothesised as a potential limiter to outcomes of children with less severe degrees of loss.

The reason for this current management practice for children born with mild loss may be a reflection of a well-meaning desire to intervene. Put simply, in the absence of clear guidelines, protocols or evidence of how best to assist children detected early with mild hearing loss, those families presenting for hearing services may be offered a treatment that has been recognised as effective for children with greater than mild hearing loss. As outlined earlier in Section 2.3.5, this may be an example of treatment creep, where a particular treatment such as hearing aid fitting is implemented in a less severely impacted population without compelling evidence of the positive benefit/harm ratio. The focus of the next section of this literature review will be on what evidence does exist, gathered from situations where hearing aids have been fitted to children with mild losses. Specifically, it will focus on the literature examining aspects of hearing aid use.
2.6.2 Evidence of hearing aid efficacy for mild losses

Ideally, to measure the efficacy of hearing aid fitting for children with mild hearing loss, a comparison of outcomes in children with and without hearing aids would occur. If the goal were to measure improved outcomes resulting from earlier hearing aid fitting, a sufficiently sized sample of children with hearing aids would be needed for a comparison of those children fitted earlier versus later. To account for potentially confounding factors in participants, this study would be of a prospective experimental study design such as a randomised controlled trial, rather than an observational design. A population-based recruitment method would also assist in enhancing the generalisability of the study results to the broader population of children born with mild hearing loss.

It is apparent that such an ideal study as described above has not been reported in the literature. Indeed, studies that focus on outcomes of children with mild loss from the specific angle of the impact of wearing hearing aids are not common. What does exist is an exploration of patterns of hearing aid use in this population of children, both with and without analysis of the association between aided status and language outcomes.

Findings of observational studies suggest that aided children with mild loss are achieving better language outcomes than aided children with greater degrees of loss. 170 This is much the same as what has been documented in most literature that has only considered degree of loss and not aided status. 107,156,160 However, there is also the consistent finding that children with milder degrees of hearing loss also use their hearing aids less. 165,193,194 As highlighted by McCreery et al (2015),195 this creates challenges for understanding the impact of hearing aids on outcomes. At first glance, an erroneous conclusion that could be reached is that children who wear hearing aids more frequently have on average poorer outcomes than those that wear hearing aids less. Thus, another detail of the ideal study to determine efficacy of hearing aids in improving the language outcomes in children with mild loss is for the results obtained to either be solely for this degree of loss, or clearly delineate results by degree of loss.
Examples of research on hearing aid use specifically in children with mild hearing loss are particularly useful as they allow sound understanding of clinical practices within a defined hearing loss range. There are few such studies and the majority are of an observational study design, often combining mild bilateral losses and unilateral losses of any degree under the minimal label (see Section 2.2.4). In a Canadian retrospective chart review of all children identified with minimal loss over a 16 year period at one paediatric hospital, the decision to aid children with mild bilateral loss was significantly related to the age at which the loss was detected (older age, odds ratio 1.24; 95% CI 1.13 to 1.36) and what PTA the child had (poorer better ear hearing loss, odds ratio 1.08; 95% CI 1.04 to 1.12). That is to say that the odds of amplification being prescribed at the time of hearing loss confirmation were 24% higher for each additional year in diagnosis age.

This same Canadian research group found that less than two thirds of children with minimal hearing loss consistently used their hearing aids, with no significant difference in amplification uptake between variants of mild bilateral loss (mild bilateral or mild high frequency bilateral) and unilateral loss. Whilst details are absent of what age children were when this hearing aid use was calculated, it is a result that parallels with that from work from the OCHL cohort, where younger age, better unaided hearing and lower maternal education have been noted as a barrier to greater hearing aid use. In a study also of OCHL participants that divided children with mild loss into categories of hearing aid use, those children who were in the full time use category (>8 hours per day) had significantly higher scores in receptive vocabulary and expressive grammar than non-users. Whilst hearing aid use was calculated from two data sources, parent report and hearing aid data-logging, it is possible this result may illustrate reverse causality. Children in the full time use category may have had more intelligent parents with higher health literacy, who both understood the potential benefits of increased hearing aid use and may have been facilitating greater language development via other methods in addition to consistent aiding. This would fit with the finding that maternal education level influenced trends in hearing aid use when examined longitudinally, yet degree of loss was
only an influence on amount of use (poorer hearing leading to greater use) once children were attending school. Other factors that may be influencing the results seen are considered in Section 2.6.3.

If hearing aids are influential in improving the language outcomes of children with mild hearing loss, measures of aided audibility may provide additional evidence of hearing aid efficacy. As outlined in Section 2.2.4, a SII score can be calculated to represent the aided audibility a child experiences through wearing a device, which can then be used to measure a relationship between amplified hearing and language outcomes. Aided audibility may also be observed through the use of functional assessment tools. These tools aim to examine the effectiveness of amplification and to understand how well hearing is used in daily life. In a group of children with losses from mild to profound, the PEACH functional assessment tool administered shortly after amplification fitting predicted language outcomes at age 3 years.

In examining the relationship between hearing aid use, the SII, functional assessment tools including the PEACH and language outcomes, McCreery and colleagues (2015) reported a generally consistent trend in the OCHL study population (aged 2-6 years spanning mild to severe losses). They found that higher parent ratings on the PEACH correlated with better SII, higher amounts of hearing aid use and greater language ability. This is of interest, as functional measures step beyond an objective prediction of speech recognition scores (the SII) to provide a glimpse of how a child is using aided audibility in their commonly encountered environments. The relationship may again be influenced by reverse causality, with children who are inherently brighter and, therefore, possessing better language skills perhaps able to respond to both unamplified and amplified sounds better. Such relationships are yet to be explored at a population-level for broader applicability to the general population.

2.6.3 Other factors influencing management of mild hearing loss
Parents of children with hearing loss are key to the successful use of hearing aids, as they try to act in the best interests of their child across their
development. Part of this is their responsibility for hearing aid use particularly at younger ages. For parents of children with hearing loss of any degree, adequate support is crucial for optimising the chances of successful use. From a large US-based population survey of over 350 parents, 48% of respondents perceived the information provided to them about hearing aid orientation and management was insufficient.\textsuperscript{199}

Aside from device management, the literature notes parent uncertainty regarding the appropriate recommendations for helping their child with a mild hearing loss. This relates to factors such as whether a device is required and when is the optimal time to fit this device. This uncertainty experienced by parents is likely influenced by the quality and clarity of advice provided by clinicians.\textsuperscript{112, 196} This may be due to clinician perceptions that children with milder losses are only going to experience relatively minor difficulties across broad language domains when compared to those with severe/profound losses. This may be interpreted by parents as the professional minimising the milder hearing loss that they are struggling to understand.\textsuperscript{112} Their confidence in choosing a management strategy may be further hampered by differing advice from various professionals, such as between audiologists and otologists.\textsuperscript{112}

The mutable factors outlined above are sensitive to improvement in management practices which could positively influence outcomes of children with mild hearing loss. Relatively fixed factors also must be considered for their impact on management. One is the extent to which families have financial means to access services and support overall child development. There is some evidence that children wear hearing aids for a lower number of hours per day as family socioeconomic status decreases.\textsuperscript{193} In a system like that of Australia, where there is no financial cost to families for hearing aid purchase, other socioeconomic factors (e.g. family capacity to attend appointments rather than attend work) could still compound developmental delays to which children with hearing loss as a group are sensitive.
Child cognition could also be a factor in determining the efficacy of management decisions for children with hearing loss. There are known strong relationships between cognition and language outcomes, and between language ability and speech perception skills.\textsuperscript{200} If the outcomes of interest in children with hearing loss are language or auditory performance based, then cognition must be considered or controlled for in evaluating efficacy of management. In a group of 62 children fitted with either hearing aids or cochlear implants, IQ was found to impact speech perception skills indirectly via influence on language outcomes.\textsuperscript{200} Similarly, a significant correlation between non-verbal IQ and speech recognition ability has been observed in both children with normal hearing and those with a diagnosis of auditory processing disorder.\textsuperscript{201} Careful experimental design to account for cognition may allow for the impact of this factor in interpreting study results. This may be of particular relevance for children with mild hearing loss, as the underlying cause of hearing loss may systematically co-vary with severity and cognition. For example, a hearing loss resulting from a Connexin 26 mutation will often be severe or profound in degree yet occur in isolation, with no known comorbidity and generally intact cognitive functioning.\textsuperscript{202} With a large proportion of congenital losses believed to be of unknown genetic cause, it is plausible that a proportion of children born with mild losses display lags in aspects of their development (such as language) that reflect their cognition.

\subsection*{2.7 Summary}

Earlier diagnosis of congenital hearing loss facilitates intervention that contributes to better language outcomes for impacted children. However, evidence of how these children are typically impacted and how to best assist them reach their potential becomes less clear as hearing loss severity decreases. With management decisions for milder losses now occurring earlier than before, the strength of evidence for improved language outcomes resulting from this recent major change in clinical practice is equivocal. Therefore, there is a clear need to address these evidence gaps, to improve our understanding of how earlier detected milder losses impact children at the population level and what influence current management approaches play in improving their outcomes.
3. Rationale, aims and hypotheses

3.1 Rationale
Children born with mild and moderate bilateral hearing loss experience poorer language outcomes than peers without hearing loss. With universal newborn hearing screening now widespread, many of these children are now fitted with hearing aids very early. This is occurring without firm evidence of efficacy. Filling this knowledge gap would support clinicians and families to make appropriate decisions for affected children.

3.2 Aims
Three broad aims were the focus of this PhD study, specifically:

1. For children with mild or moderate congenital hearing loss aged 5-8 years of age drawn from four population-derived samples:
   a. compare language, behaviour and quality of life outcomes between samples,
   b. determine whether age of detection predicts outcomes within the four samples pooled, and
   c. compare mean outcomes of the contemporary sample to a typically developing sample of similarly-aged children.

2. For a sub-set of early-identified children with mild or moderate congenital hearing loss at 5-7 years of age:
   a. establish the relationship between a measure of aided audibility and both (i) unaided hearing acuity and (ii) speech recognition ability, and
   b. quantify the extent to which (i) unaided hearing acuity and (ii) measures of aided audibility are associated with speech/language outcomes.

3. For early-identified children with mild or moderate congenital hearing loss at 1-3 years of age:
   a. describe parent reports of expressive vocabulary, and
   b. compare mean outcomes to a representative sample of similarly-aged children without hearing loss.
3.3 Hypotheses

The hypotheses that map to the above stated aims are:

1. Both children with mild and moderate hearing loss would display:
   a. secular trends toward improved language, behaviour and quality of life,
   b. better outcomes if detected with hearing loss earlier, and
   c. comparable mean outcome performance to typically developing children without hearing loss.

2. 
   a. Aided audibility values would substantively correlate to both unaided hearing acuity and speech recognition ability.
   b. Measures of aided audibility would be more highly associated with speech/language outcomes than unaided hearing acuity.

3. 
   a. Children with mild hearing loss would display mean expressive vocabulary development similar to children without hearing loss.
   b. Children with moderate hearing loss would display significantly poorer mean expressive vocabulary development than children without hearing loss.
4. Methodology

4.1 Overview

This chapter provides a detailed description of the methods for this PhD study. After an initial overview of all contributing cohorts, it presents an in-depth description of the cohort that comprised the unique data creation for this PhD. The features common to all cohorts are then detailed, followed by descriptions of the design, setting, participants and ethical approval. Finally, it delineates the procedural details for the two age groups (5-7 and 1-3 years) relevant to the three sub-studies and their respective outcome measures. Statistical methods are presented for each overall PhD aim.

4.2 Overview of cohorts

Five cohorts of children from four population-based source studies contributed data for this PhD (see Figure 4.1 for overview); four cohorts were of children with hearing loss and the fifth comprised children without hearing loss. This section provides a very brief overview of the cohorts to orient the reader, with in-depth cohort descriptions from Section 4.3 onwards. Central to this PhD’s epidemiological underpinnings, all cohorts were population-derived as described later in the chapter.

The Children with Hearing Impairment in Victoria Outcome Study (CHIVOS) reported language outcomes at age 7-8 years of children born between January 1991 and July 1993, derived from the statewide register of children with aided hearing loss. These children were born during a period when there was effectively no systematic approach to detection of congenital hearing loss. A subset of children from the longitudinal CHIVOS project was incorporated into Study 1 (addressing Aim 1) of this PhD and were defined as the opportunistic detection group. Further details of the CHIVOS cohort is in Section 4.4.1.
Figure 4.1. Overview of source cohorts comprising studies 1-3 of this PhD
The Statewide Comparison of Outcomes (SCOUT) quasi-randomised trial comprised children born March 2003 to February 2005, drawn across two Australian states from the national register of children with aided hearing loss. At the time these children were born, New South Wales had just (December 2002) commenced universal newborn hearing screening (UNHS) with automated auditory brainstem response (AABR) technology. Victoria had just commenced AABR screening for admissions to its four neonatal intensive care and attached special care nurseries. This was complemented by a well-established universal population-based risk factor screening process operating throughout the state. This comprised risk factor screening at 2 weeks and 8 months of age, with behavioural hearing screening at 8-10 months of age. These children with hearing loss were all assessed at 5-6 years of age and were split into two subsets for the purposes of Study 1 in this PhD. These were defined as the risk factor screening group from Victoria and the newly established UNHS group from New South Wales. Further details of the overall SCOUT study is in Section 4.4.2.

The Victorian Childhood Hearing Impairment Longitudinal Databank (VicCHILD) yielded the fourth hearing-impaired cohort. From this ongoing databank of children of all ages, those born in two date ranges (April 2007 to April 2010 and August 2012 to February 2014) were eligible. At the time these children were born, UNHS had been in operation in Victoria for up to nine years. Streamlined systems were in place to minimise delay between identification, diagnosis, and access to early intervention services (hearing technology and education). Children born in 2007-10 were assessed at 5-7 years of age and were defined as the mature UNHS group in Study 1 of this PhD. A sub-group of the mature UNHS group, those who wore hearing aids at the time of assessment, formed the participants for Study 2 (addressing Aim 2). Caregivers of the children born in 2012-2014 were surveyed at child age 1-3 years of age for Study 3 (addressing Aim 3) of this PhD. VicCHILD is detailed in Section 4.3.

The fifth group of children used in this PhD was the large population-based birth cohort known as the Early Language in Victoria Study (ELVS). These children,
born in 2003, were selected from Victorian metropolitan local government areas spanning the spectrum of disadvantage-advantage to maximise population representation. This thesis draws on data from waves 3 and 8 of ELVS when participants were aged 2 and 7 years respectively. This cohort of children is referred to in this PhD as the children without hearing loss group for Study 1 and the representative sample for Study 3. Further details of the ELVS is in Section 4.4.3.

Figure 4.2 summarises the periods of birth and mean ages of detection for the four cohorts contributing to this PhD, in the context of changing population-wide hearing detection systems in place since the early 1970s in Victoria and New South Wales. This figure shows the impact on mean age of congenital hearing loss detection by birth year.
Figure 4.2. Timeline of population-wide systemic hearing detection measures used in two Australian states

Abbreviations: VIC, Victoria; NSW, New South Wales; CHIVOS, Children with Hearing Impairment in Victoria Outcome Study; SCOUT, Statewide Comparison of Outcomes study; VicCHILD, Victorian Childhood Hearing Impairment Longitudinal Databank; ELVS, Early Language in Victoria Study.
4.3 In-depth cohort description: VicCHILD

The bulk of active data design and acquisition in this PhD occurred within the Victorian Childhood Hearing Impairment Longitudinal Databank (VicCHILD) cohort described briefly in Section 4.2. This section provides a detailed description of the VicCHILD cohort, which contributed participants in two age groups and within which all of this PhD’s new data were collected.

VicCHILD is a population-level databank for all children born with congenital hearing loss of any degree or type in the state of Victoria. Data collection for this PhD was harmonised with scheduled VicCHILD data activities, at preschool and early school age. Information about the databank relevant to this PhD is summarised below.

The VicCHILD research project is coordinated by a team at the Murdoch Children’s Research Institute at Melbourne’s Royal Children’s Hospital, serving the state of Victoria (population 5.8 million people), Australia. VicCHILD aspires to advance research about childhood hearing loss and, as a databank, aims to continue recruiting children and storing information continuously and confidentially. As such, it is a growing resource. Different types of information are collected from various sources with collection burden minimised for participating families. Data linkage, biological samples, exchange of information and data sharing are all elements of the VicCHILD project.

VicCHILD commenced recruitment of Victorian-born participants in 2010 using two approaches: a once-off retrospective approach to families with a hearing impaired child born since universal newborn hearing screening (UNHS) commenced in 2005, and via ongoing prospective recruitment for children born after VicCHILD became operational in 2010. Both recruitment processes are relevant to this PhD and occur in cooperation with the Victorian Infant Hearing Screening Program (VIHSP), the statewide UNHS detection system.

With a consistently high uptake rate (over 99%), VIHSP effectively screens all Victorian newborns for congenital hearing loss and facilitates referral to
diagnostic assessment for infants who do not pass. Audiologists return results from diagnostic assessment to VIHSP. Following a diagnosis of permanent hearing loss of any degree or type, VIHSP contacts families via letter with a brief explanation of the VicCHILD project. Families are provided with a two-week opt-out period to decline contact from VicCHILD researchers. Following this period, VicCHILD project staff contact families by phone to seek verbal permission to post a baseline pack, which interested families then complete. This pack contains a participant information statement, consent form, questionnaires and saliva collection materials.

Data for VicCHILD are collected longitudinally with repeated measures from participants and families. Parent questionnaires and direct assessment (when children are old enough) of speech and language development occur at enrolment, in the preschool years, at school entry, at late primary and late secondary school. This VicCHILD-specific longitudinal whole-of-childhood data is intended to supplement data obtained from other sources (e.g. state and national databases) that detail service use, diagnoses, educational outcomes and intervention status and use.

At baseline enrolment all parents/caregivers complete basic demographic questions concerning their family broadly and their child with hearing loss specifically. Importantly, they choose whether to provide their informed consent for different aspects of the VicCHILD project (see Appendix A for information statement and consent form). These additional consents cover: the accessing of existing biological samples and the provision of new samples, permitting the exchange of information about the child with hearing loss with other professionals where doing so would reduce child burden of repeated assessments, and agreeing to their data being shared in a de-identified manner with other research projects. Two other consents are sought at baseline which are crucial to this PhD study: families choose whether to be re-contacted by VicCHILD for future waves of direct assessment data collection and whether they agree that VicCHILD could seek to link their child’s data to existing datasets held in other organisations (e.g. hospital records and health-related databases).
Families have consistently responded very positively to these additional consents, with over 90% of families agreeing to all options, barring a slightly lower uptake of consent for biological samples.

All data collected by VicCHILD, either from other sources or via direct contact with participating families, are stored within the online application Research Electronic Data Capture (REDCap). REDCap is a secure web-based tool that can be used for building and managing databases. Each VicCHILD participant is assigned a unique identifier used to record all participant contact and information obtained.

4.4 Historical cohorts

The purpose of this section is to provide further detail on the historical cohorts outlined in Section 4.2. The similarities of the population-level studies to the VicCHILD cohort will be highlighted.

4.4.1 CHIVOS

CHIVOS was conceived in response to methodological flaws in then-influential research into outcomes for children with hearing loss that limited generalisability to the broader population. Important studies by Yoshinaga-Itano and Moeller had suggested the combination of very early identification and intervention services would improve language outcomes to normal levels, irrespective of severity of loss and intelligence and socioeconomic status. However, at the time of CHIVOS' development, there was an absence of population-level studies that could incorporate epidemiological practices to minimise selection bias and improve generalisability. Such a study would include those children with hearing loss for whom continued intervention attendance within specific programs was not achievable, would include information on innate cognitive abilities, and would look beyond preschool language outcomes to longer term outcomes achieved after childhood language trajectories had stabilised. These attributes were largely absent from previous studies, thus CHIVOS was established in order to substantiate the longer-term benefit of earlier diagnosis.
Children were eligible for inclusion if they were born January 1991 to July 1993 in the state of Victoria, Australia, were fitted with hearing aids for bilateral congenital hearing loss by age 4.5 years, and did not have intellectual or major physical disability. Australian children diagnosed with a hearing loss that may require amplification are referred to Australian Hearing, a federally funded organisation responsible for providing no-cost amplification following nationally standardised processes to the paediatric population. At the time of CHIVOS, Australian Hearing routinely provided Victoria’s hearing screening program with data on all children classified with congenital hearing loss, which formed the sampling frame for the study. This enabled researchers to assess all eligible and consented families from across the state.

A flowchart detailing participant recruitment is shown in Figure 4.3 (sourced from Wake et al (2005)\textsuperscript{163}). Language outcomes were measured using standardised assessment tools at age 7-8 years. The overall response rate was 67%, with a participant hearing loss at diagnosis breakdown being: 21% mild, 34% moderate, 21% severe and 24% profound.
Figure 4.3. CHIVOS participant recruitment flowchart
4.4.2 SCOUT

The quasi-randomised SCOUT trial compared outcomes of children with congenital hearing loss born in two Australian states. At the time of birth, New South Wales (NSW) was undertaking AABR newborn hearing screening. Victoria (VIC) was conducting risk factor screening of the majority well-baby population, with only babies admitted to the neonatal intensive care unit receiving AABR screening. During the period of participant birth (2003-2005) the states were similar when examining measures of socioeconomic advantage, ethnic diversity, education system characteristics and access to health services (both general and audiology related). Australian Hearing, as introduced in Section 4.4.1, used national protocols to deliver the same rehabilitative services for all children in the trial. Thus these two states significantly differed only in their approach to congenital hearing loss detection.

Children were eligible for inclusion in the SCOUT study if they were born in either NSW or VIC between March 2003 and February 2005 with a bilateral hearing loss believed to be congenital of greater than 25 decibels hearing level in the better hearing ear. Children needed to have had amplification fitted by 4 years of age. Exclusions comprised children of parents with insufficient English to participate, hearing loss that was unilateral, temporary, acquired or in the normal range at age of assessment (5-6 years), and logistic (e.g. living outside Australia) or social reasons that precluded participation. Unlike the CHIVOS study, presence of an intellectual disability was not an exclusionary criterion. National Acoustics Laboratory accessed AH’s national database of children fitted with amplification, resulting in state-wide coverage of children with congenital hearing loss.

Recruitment into the SCOUT study followed a two-stage opt-in process. Researchers initially audited AH records to determine who met eligibility criteria, obtaining permission from the treating audiologist to approach the family on their behalf. Researchers then posted an information letter from the audiologist outlining the study, inviting families interested in participating to contact the SCOUT research team directly. Figure 4.4 (sourced from Wake et al
(2016) displays a flowchart detailing participant recruitment across the two states. Non-participants (57%) did not significantly differ from participants (43%) on measures of initial or current hearing level. Standardised assessment tools were used to measure outcomes, as per the CHIVOS study detailed in Section 4.4.1. These covered measures of language, receptive vocabulary and non-verbal IQ. Parent-completed questionnaires measured general child behaviour and health-related quality of life.

Figure 4.4. SCOUT participant recruitment
Abbreviations: RFS, risk factor screening; UNHS, universal newborn hearing screening; dB HL, decibels hearing level; PTA, pure tone average; Qre, questionnaire; HUI, health utilities index.
4.4.3 ELVS

ELVS is a community-based longitudinal cohort study of speech and language development in a general population. Participants in this 2003 birth cohort were recruited from six of 31 local government areas in metropolitan Melbourne. A measure of social disadvantage derived from 2001 Australian census data was used to determine from which six local government areas ELVS should recruit participants. This social disadvantage index (SEIFA) represented population attributes such as low income, low education completion rates and high unemployment. By stratifying the local government areas into three tiers using this SEIFA index, two non-contiguous local government areas from each tier were selected to ensure that ELVS recruited from geographic areas that covered the spectrum of advantage/disadvantage.

Recruitment targeted all children aged 7.5 to 10 months of age living in the six local government areas who did not have development delay, cerebral palsy or other known serious intellectual or physical disability, and whose families had sufficient English for study purposes. Methods used to recruit within the local government areas were aimed at maximising the number of participants. In cooperation with maternal and child health programs which provide services to approximately 82% of families at around 8 months of age, maternal and child health nurses were asked to consecutively approach parents of infants attending their 8 month health check. Parents were also approached at their children’s 8 month behavioural hearing screen in operation at the time, and parents could also directly approach ELVS researchers if interested. A participant sample of over 1900 was achieved at the inception of ELVS, which represented approximately 35% of all eligible participants in the six local government areas. ELVS is an ongoing longitudinal study with scheduled data collection waves. Participation at each wave was sought from all families originally recruited, unless they had formally withdrawn from the study prior. As such, the size of the ELVS cohort fluctuates across the data collection waves, illustrated in Figure 4.5 (sourced from Reilly et al (2017)).
Figure 4.5. Participant flowchart for the ELVS cohort across waves of data collection
Standardised measures administered repeatedly across data collection waves form the backbone of ELVS data, along with parent-completed questionnaires. This PhD utilised ELVS data from wave 3 and wave 8, when participants were 2 and 7 years of age respectively. Total cohort number inevitably decreased over time, with the cohort sample size being 1741 at wave 3, and 1189 at wave 8.

4.5 Design

This PhD has a significant cross-sectional component, which comprised its active data design and data acquisition. This was complemented by the cohorts drawn from longitudinal studies, which helped to shape the method of the overall study.

The cross-sectional component of this study involved active data collection from children with mild or moderate congenital bilateral hearing loss in two age ranges: 5-7 years and 1-3 years. For Study 1, data on children aged 5-7 years were used. Firstly, cross-sectional data were used that allowed quasi-experimental analyses. Secondly, data were pooled to enable longitudinal analyses in a sub-aim. For Study 2, a subgroup of the 5-7yo children was supplemented by additional cross-sectional data. For Study 3, data on children aged 1-3 years were analysed.

The next section outlines potential sources of error, bias and confounding relevant to this PhD study. It details strategies used to deal with these issues in the cross-sectional component of this PhD to which they apply.

4.5.1 Internal and external validity

Validity is an essential attribute of research. Research quality is dependent on two aspects of validity: internal and external. Internal validity represents an indication of how much the results of the study reflect the true situation for the studied sample, in the absence of alternative explanations. Low internal validity may be the result of chance, confounding, or bias. External validity is how generalisable the results of the current study are to the wider population. Higher levels of both internal and external validity increase the confidence in the conclusions drawn and provide weight towards the broader relevance of the
findings. The following section outlines the strategies undertaken to enhance validity throughout this study.

4.5.1.1 Minimising random error

Random error can occur if any factor is introduced into a study that impacts the measurement of a variable. Random error decreases the precision of estimates in a cumulative manner (i.e. as more bias is introduced, the precision will decrease, often seen via a widening of confidence intervals). Random error, as the name implies, is difficult to predict or control.

A possible source of random error in this study was a parent misunderstanding the meaning of a question in the questionnaire, which may lead to a response that is not an accurate answer to the true question. Random error was a potential risk to the internal validity of this study due to the limited sample size of the contemporaneous group of participants. Small sample sizes are a risk to precision as there is reduced ability to collect more data upon which to average out the variation in measurement. If analyses were to be based solely upon the participants sourced from the VicCHILD cohort, a lack of precision around results could then impact the conclusions drawn. Therefore, efforts were made to restrict the likelihood of random error influencing study results using a combination of methodological decisions and tools.

In this study, random error was minimised primarily through adherence to standardised research manuals. As my PhD project was nested within VicCHILD, the VicCHILD Technical Protocol (relevant pages in Appendix B) and VicCHILD Project Manual (relevant pages in Appendix C) were used, which detailed methods for child recruitment and contact for the project broadly. Adhering to the processes outlined in the protocol and manual ensured all participant contact was carried out in a consistent manner. Additionally, the manual provided detailed methods for measure administration and general data collection that enhanced consistency across the data collection period. Standardised and detailed multi-item measures were used wherever possible to enhance construct validity. Such standardised measures were chosen as they demonstrated high
reliability and validity, and multi-item measures assist in minimising random error by collecting multiple responses that all contribute to the precision of the underlying construct. During analyses, reliance on samples with limited sample size were restricted wherever practicable, to limit the influence of random error further on the ability to draw conclusions.

4.5.2 Minimising bias

4.5.2.1 Recall bias

When seeking information that requires participants to recall information from the past it is inevitable that there will be some error. If such error were random, and occurred across all participant groups, it is likely to influence effect estimates towards the null. However, recall bias occurs when the study systematically creates a tendency for some participants to recall past experiences or events differently to others. This type of bias can be particularly prominent in cross-sectional case-control studies, where two distinct participant groups are compared and when information is sought about past rather than recent events.\(^{206}\)

Recall bias was a potential risk to internal validity of this study due to seeking information from caregivers relating to past events. Details of past hearing status and device usage (e.g. age of hearing loss diagnosis and hearing device fitting, usage estimates) and participant behaviour were sought. The risk with such questions is the respondent could recall requested information incorrectly, due to elapsed time or an internally held bias (e.g. social desirability might lead to reporting hearing aid use higher than actual, due to the belief greater use benefits the child). Such bias could lead to altered (weakened/strengthened) associations between examined factors and outcomes compared to the true association.

Recall bias was minimised in this study by collecting information as close as possible to the time frame of interest. Objective information was obtained from other sources (e.g. a database that recorded hearing aid fitting dates) where practicable. Verification of information from respondents was checked against
these other sources where possible, with further investigation conducted if sources were not in agreement. Information requested from caregivers on past events was restricted to the recent past (e.g. child’s behaviour over the preceding six months). Further minimisation of the impacts of recall bias occurred via the use of multiple measurements for the same information (e.g. hearing aid use measured as a single estimate for total number of hours of use for the day, as well as via an hourly scale across waking hours and within a separate questionnaire).

4.5.2.2 Selection bias

Selection bias occurs when there is a lack of representation or commonality between the selected, studied sample and the population of interest. Selection bias in cross-sectional studies can occur at the point of recruitment and at any follow-up.

The generalisability of findings can be impacted by the manner in which participants are selected. Bias can be introduced via the study setting, location, inclusion and exclusion criteria. There needs to be a balance between these aspects and still being able to address the research question of interest. A further manner in which selection bias can occur is via systematic differences between individuals who were approached and became participants and non-participants. This is a very challenging issue to address.

Attempts to decrease selection bias occurred through the method of recruitment from the VicCHILD project. VicCHILD is open to all children in the Australian state of Victoria with a congenital hearing loss, with defined methods to maximise population representation (see Section 4.6 for further details). Inclusionary criteria were as broad as possible in order to maximise the representativeness and diversity of the overall population of children with congenital hearing loss.
4.5.2.3 Minimising confounding

Confounding occurs if an association between an exposure and an outcome is distorted by another variable. There are three conditions that must be met for a variable to be a confounder: (i) it must be associated with the outcome variable; (ii) it must be associated with the exposure variable; and (iii) it cannot lie on the causal pathway between the exposure and the outcome. Confounders can impact the association observed between exposure and outcome in numerous ways, via over- or under-estimation, by masking an effect to appear as if there is none, or by creating the appearance of an effect when this is not true.

Confounding can be controlled via decisions made at the time of study design, through randomisation, restriction or matching. This is contingent on the study design chosen being amenable to such attempts to control confounding. In general, the best approach to confounding is to first set out to avoid all that is avoidable, bearing in mind that the only design able to remove unknown confounders is randomisation. Secondly, if it is unavoidable, attempts should be made to try and adjust for the effect of known potential confounders during the analysis phase of the study. Statistical methods exist which permit adjustment for potential confounding variables. We can identify potential confounders _a priori_, such as what has been observed in prior literature, or through statistical techniques. Commonly both approaches are used.

In this study, confounding was handled during the analytic phase to maintain generalisability within our already restricted participant base. Known potential confounders were identified _a priori_ from the literature, acknowledging that a small sample size limited the number of factors that could be adjusted for. Multivariable models were used where possible to examine the effect of potentially confounding variables and the balance between selection bias and confounders.
4.6 Setting and location
Coordination of this study occurred from the Murdoch Children’s Research Institute, located at The Royal Children’s Hospital Melbourne, in the state of Victoria, Australia. Participating children were sourced from either an existing databank of children born with hearing loss (VicCHILD) or from past studies of language outcomes for children from Victoria and New South Wales. The true population nature of this project is a key strength; participation was equally open to children from both metropolitan and rural areas within the two Australian states of birth. To increase participation, direct language assessments typically occurred in the family home, or at the Royal Children’s Hospital if preferred. When families lived in locations impractical for direct language assessments (e.g. overseas or other Australian states), their participation was maintained where possible via the completion of written questionnaire components of the study.

4.7 Participants
This section on participants is restricted to those on whom active data collection was conducted: the participants from the VicCHILD databank. Details of participants sourced as sub-groups of historical studies (CHIVOS, SCOUT and ELVS) are located in Section 4.4.

As outlined above, participants in this PhD were a subset drawn from the VicCHILD databank. Therefore, eligibility criteria for this study were primarily those of the databank, with further refinement to restrict the sample to the degree of hearing loss and age groups of interest.

4.7.1 Inclusion criteria
Any child enrolled in the VicCHILD project and aged 5-7 years or 1-3 years was eligible for inclusion if they met the following criteria:

1. Their degree of hearing loss at the time of diagnosis was bilateral mild or moderate sensorineural in the better hearing ear. This information was provided to the VicCHILD database by VIHSP and was derived from
newborn hearing assessments following a UNHS referral. If a VicCHILD participant was of the correct age but this diagnosis information was missing, the caregiver was contacted to determine whether their child had either a mild or moderate loss in the better ear when diagnosed, as defined immediately below in Section 4.7.1.1.

2. The child’s age at the time of data collection would fall within the age range 5;0 to 7;11 or 1;4 to 3;0.

4.7.1.1 Hearing measures

One component of the VicCHILD project is to obtain participant information from other sources, such as national databases. Audiometric information for children with permanent hearing loss is predominantly held within Australian Hearing, with which, at the time of this study, VicCHILD was yet to complete data linkage. Thus the only hearing sensitivity information routinely available within VicCHILD was the diagnostic hearing assessment results from the VIHSP, featuring results that are electrophysiological in nature. From these results, hearing levels can be predicted with reasonable confidence, but are not considered the gold standard in audiological measurement (see Section 2.3.2 for details). So whilst electrophysiological information was sufficient for initial identification of children with mild or moderate hearing loss, behavioural hearing thresholds were required to calculate specific pure tone averages.

Behavioural audiometric data were obtained from Australian Hearing. When families were contacted to confirm their participation in the 5-7 year old VicCHILD assessment, requests were made to Australian Hearing for not only the audiogram most recent to the VicCHILD assessment date, but also the first reliable behavioural audiogram obtained for the child (see Appendix D for cover letter for request). The request covered any available threshold information across the octave frequency range 250 Hz to 8000 Hz. These audiometric thresholds were entered into the VicCHILD database and were used to calculate either a three frequency (500 Hz, 1000 Hz and 2000 Hz) or four frequency (addition of 4000 Hz) average hearing level. Two methods of calculating the pure
tone average (PTA) were used for harmonisation reasons; the three-frequency PTA had been used in historical studies included in Study 1, whereas more audiometric frequencies could be designed into Study 2. Using the first audiogram allowed verification of mild or moderate hearing loss in the better hearing ear as close as possible to the time of diagnosis. Using the audiogram closest to VicCHILD assessment allowed for the current hearing levels to be articulated and used in analyses for potential confounding.

For respondents to the 1-3 year old ELQ, audiometric information about the respondent’s child was requested from Australian Hearing following receipt of their completed questionnaire. This provided both initial and current PTAs from behavioural audiograms for use in Study 3.

4.7.2 Exclusion criteria
Children were excluded if they met the following criterion:

1. The child and family had insufficient English to complete study requirements, i.e. a direct child language assessment (age-dependent) and parent completion of questionnaires written at a Grade 6 English reading level.

4.8 Procedure
As mentioned earlier in this chapter, participants were in two age groups (5-7 and 1-3 years). The activities conducted with these two groups are distinct. Therefore, for ease of reading the details of procedures are stratified by age group. Details of measures clearly denote to which age group of children the measure was used. It will be noted where there were common procedures to both age groups.

4.8.1 Early primary school aged children
All children within this age group (5-7 years) were already recruited into the established overarching research project. Thus the procedures centered on
confirming participation in this wave of data collection under the auspices of VicCHILD and associated activities to prepare for assessment.

For the given data collection period of January 2015 to August 2015, VicCHILD provided the researcher with contact details and diagnosis hearing levels defined by degree and type for all children who would be within the age range 5;0 to 7;11 and who had consented to being contacted for the purpose of direct assessment (see Section 4.3 for details). The research team began telephoning primary caregivers to ascertain their willingness and availability for a direct assessment to occur. Primarily these visits were to occur in the family home, with an alternate option of the child attending the Royal Children's Hospital for assessment if the family chose. For those who did not agree to direct assessment, researchers requested that parents complete questionnaires only. These could be mailed to caregivers and returned via reply paid post. Procedures and call scripts were documented in the VicCHILD Project Manual (see Appendix C).

Once agreement to direct assessment had been obtained and date and time for the assessment were arranged, follow up questions were asked regarding any recent speech and language assessment, possible difficulties with completing required tasks, and details of hearing status and any devices used. These supplementary questions were necessary as this point of contact with families was quite often the first since recruitment into VicCHILD, and these details may not have been known at the time of their enrolment.

Specifically, this information facilitated several activities:

1. Obtain any speech, language or psychosocial measures recently administered, to ascertain whether to administer again or obtain results from external sources if it had occurred recently (within the last 6 months)

2. Understand whether any educational funding application or other research project participation was ongoing or planned, for which
VicCHILD results could be provided to reduce assessment burden on the child

3. Adjust delivery of assessment (if permissible within published test parameters) due to any physical or intellectual need

4. To determine aided status, and to obtain the most recent hearing assessment data, which was required for one component of the test battery (the LiSN-S, see Section 4.10.1.3 for details).

Details of associated organisations and permission to contact these services were obtained verbally, and subsequent efforts were made to obtain the required information. This is detailed in the VicCHILD Project Manual in Appendix C.

Of note is the acquisition of hearing assessment data. All families were asked if their child attended Australian Hearing for their audiological monitoring. Details of the branch they attend were obtained, and under data linkage consent from VicCHILD, the researcher contacted these centres to obtain current audiograms. The cover letter for this request is in Appendix D.

Following agreement to participate, all families were posted a pack containing a confirmation letter with assessment date, time and location specified, a parent questionnaire and several other caregiver-completed measures (Assessment of quality of life – four dimensions, Mill Hill vocabulary scale and others listed in Figure 4.6). These were posted to allow for completion prior to the assessment date if possible. All documentation is in Appendix E. Reminder processes occurred either by phone call or SMS message to confirm the meeting the day prior, with families contacting the researcher if they needed to reschedule.

Assessment took place in the agreed location, where a schedule of assessments was followed. Figure 4.6 shows a schema outlining the sequence and time allowance for each of the measures at 5-7 years; a subset of these measures
contributed to this PhD, and are further detailed in Section 4.10. Figure 4.6 also shows the full range of tasks that caregivers completed during the child’s assessment. The researcher encouraged the parent to complete all questionnaires before the end of the assessment, and hand to the researcher; if this was not possible, the parent retained any unanswered measures to complete and return as soon as possible in a reply-paid envelope. As per the VicCHILD project manual schedule for follow up of missing data, VicCHILD researchers made contact with families via SMS message and phone calls over three weeks at weekly intervals in an effort to have questionnaires returned. If no materials were returned after the third reminder, no further reminders occurred.

VicCHILD aims to be a low-burden project for participating families. To this end, the project allowed one face-to-face visit to conduct and collect all assessment data. Consistent monitoring of progress through the VicCHILD test battery during assessment ensured that the maximum possible number of PhD-related measures were completed. Of note, the clear majority of participants were able to complete all prescribed assessments within the allotted assessment time.
Figure 4.6. Timed schema of participant progression through 5-7 year old assessment tasks

4.8.1.1 Early primary school aged children who wore hearing aids

The overall group of children aged 5-7 years were selected on the basis of their hearing loss at the time of diagnosis being a bilateral mild or moderate congenital loss. By the assessment at 5-7 years of age, some of these children's hearing losses had progressed or improved. The second PhD aim was to explore the impact of wearing hearing aids on language outcomes, and some children were either not users of amplification or had progressed to using cochlear implants. Thus, the pool of participants at 5-7 years for Study 2 (addressing Aim 2) narrowed to include only those who were using hearing aids at the time of assessment. This sub-group formed the group of children with hearing loss within Study 2 of this PhD. For this sub-group, further procedural detail is required to explain how the hearing aid information was obtained.

In order to simulate real-ear measures to determine hearing aid output and derive a measure of aided audibility (Study 2), details of the hearing aid fitting were required alongside access to the appropriate devices in order to replicate their programming. Parents reported details of hearing device use (e.g. whether hearing aids, cochlear implants or no amplification were used) at the time of contact with families to confirm participation in the 5-7 year old VicCHILD assessments. At the conclusion of the direct assessment period, the list of participants who were current hearing aid users was provided to Australian Hearing. Australian Hearing accessed their database to obtain the hearing aid manufacturer and model information fitted to the participating children. As these children were of a similar age and had similar requirements for hearing aid power, there were a small number of different hearing aids prescribed.

Australian Hearing provided hearing aids that matched the make and model of those prescribed to the participants. All the aids were digital and non-linear. Australian Hearing also provided the hearing aid programming files for each child. The Noah System version 3 was used for all hearing device activities. Noah is software commonly used in hearing aid fittings, acting as an integrated system to measure and fit hearing aids from multiple suppliers and device manufacturers.207 Programming a hearing aid consists of applying a specific
prescription to the unaided measure of an individual’s hearing, in attempts to optimise hearing levels. Programming also includes the activation or deactivation of device-specific features and functions (e.g. automatic directional microphones, feedback suppression algorithms). Each time changes are made to the programming of a hearing aid (for example, due to parent feedback, a change in hearing threshold or child report), a programming file is created within the Noah software. All children were fitted with the hearing aid prescription method predominantly used in Australia, the National Acoustics Laboratory – Non-Linear Second edition (NAL-NL2) prescription.

Within the Noah file for each participant is the record of each programming session that had occurred on that child’s current and former devices. For each child, the Noah file reflecting the child’s hearing aid characteristics at the time of their VicCHILD assessment was identified. This was a critical step, as the participant may have undergone language assessment early in the assessment period (January to August 2015) and had a hearing aid adjustment between that time and when the Noah files were obtained from Australian Hearing. If an adjustment had occurred, the most recent programming record in the Noah file may not have necessarily reflected the child’s aided hearing at the point of assessment. Therefore, the appropriate programming file was carefully identified and the correct model hearing aid was programmed accordingly. Only one hearing aid per child was programmed; this was the aid fitted to the better hearing ear based off audiometric data from around the time of VicCHILD assessment.

After the hearing aid was correctly programmed, the output from the hearing aid was measured using coupler measures in a test box. Coupler measures were performed as real ear measures, which would have taken into account the influence of the child’s ear canal resonance on the output of the hearing aid, could not feasibly be performed as part of the VicCHILD assessment. Following all hearing aid measurements, the researcher compiled this data with audiometric data for hand entry into the Situational Hearing Aid Response Profile (SHARP) program to determine a speech intelligibility index (SII) score.
(an aided audibility measure) to varying intensity stimuli. The procedure followed to achieve these steps is documented in Appendix F.

4.8.1.1 Hearing aid use data logging
It was originally planned that data logging activity would be collected from the children’s hearing aids, as supplementary information to the parent reported hearing device use in the questionnaires obtained at direct assessment. Data logging provides objective data on hours of device switch-on and often a breakdown of different programs used/listening environments exposed to in the time between hearing aid adjustments. However, activation of data logging functionality did not form part of the standard Australian Hearing protocol at the time of assessment and as such few children had this data recorded. Also by this age (5-7 years), the timing of Australian Hearing review assessment for most children becomes less fixed than the regular appointments occurring in the preschool years and more responsive to issues of hearing difficulty or fluctuation. This meant that, even if data logging were activated, the period over which information was collected varied between children. Whilst approximate daily hours of use were estimated in the software, this was calculated over a time period that varied between children (observed to range from a couple of weeks to over 12 months). Consequently, direct data logging information were not used and the only information on device use was the parent-reported estimates of daily use completed in the written questionnaires (see Section 4.10.2).

4.8.2 Preschool aged children
As per the early primary school aged children, all preschool aged children (1-3 years) were already recruited into the overarching VicCHILD databank. The procedures again centered around confirming families were able to participate in this wave of data collection, which was instigated solely for this PhD study.

For the data collection period of July to December 2015, VicCHILD provided the researcher with contact details for all children consented into the databank with a mild or moderate bilateral hearing loss at diagnosis and who were within the
age range of 16 to 30 months. This age range was selected as it matched the age range of the main outcome measure to be used in this arm of the study.

Attempts were made to obtain parent reports of emerging language development in children with hearing loss at as close to 2 years of age as possible. This would allow for direct comparisons with the representative sample of children without hearing loss from ELVS who were assessed at the same age. However, due to the limited number of children turning 2 years during the data collection window, a larger spread of ages at questionnaire completion was foreseen for the cohort with hearing loss. This had implications for analysis, as detailed in results Chapter 7. The researcher developed the questionnaire for this arm of the study (known as the Early Language Questionnaire (ELQ)), posting this to families with a cover letter highlighting the reasons for collection and instructions on how to complete (see Appendix G). Follow up of these posted questionnaires occurred as per the VicCHILD reminder process for retrieving parent-completed data, explained in Section 4.8.1 above and in the VicCHILD Project Manual.

Early in the data collection period, a bulk mail out occurred to families whose eligible child had passed 24 months of age but was within the 30 month maximum age limit. This was conducted in an attempt to boost participant numbers. Similarly, towards the end of the data collection period, those children aged 16 months to 24 months were approached en masse via mail out to boost numbers. Splitting the approach to families in this manner served to both increase the number of responses received overall, and to maximise the responses received around 24 months of age.

Despite our use of proven reminder processes for the return of the ELQ, some families did not complete the required materials within the desired 16-30 month age bracket of development. When this occurred, the researcher retained the data collected for these older children and accounted for these in the statistical approach taken towards analysis.
Upon return of the completed ELQ and associated materials, the researcher hand-entered data into the VicCHILD databank data repository. Data accuracy checks were performed as detailed in Section 4.11.3.

4.9 Ethics
As a PhD project with activities fully embedded within the VicCHILD project, ethics approvals were by way of VicCHILD ethics approval from the Royal Children’s Hospital Human Research Ethics Committee. Any alterations to documents sent to participants, or to procedures or communications with participants, were ratified by the ethics committee. Ethics approval certificates are in Appendix H.

4.10 Outcome measures
This section on outcome measures combines the data collection activities from the two age groups studied.

As outlined above, this study was nested within broader VicCHILD activities. Therefore, PhD-related measures were intertwined with the assessment protocol for VicCHILD. The full list of measures is available in the VicCHILD Project Manual; Figure 4.6 (see Section 4.8.1 above) shows the timed flow of assessment administration and data collection at the direct assessment for the older group of children (5-7 years). The following section of this chapter provides more details about the PhD-related measures, including the derivation of subscales. Table 4.1 summarises the key outcome measures across the three studies comprising this PhD. Table 4.2 specifically relates to Study 1 of this PhD, outlining the different outcome measures used in the source cohorts over time. Outlined throughout Section 4.10, under the individual measure descriptions, are any differences between measures used in the active data collection component of this PhD to those obtained from historical cohorts.
<table>
<thead>
<tr>
<th>Construct</th>
<th>Measure</th>
<th>Completed by</th>
<th>PhD study/aim</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td>Child</td>
<td>Parent</td>
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<tr>
<td>Receptive and expressive language</td>
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<td>Receptive vocabulary</td>
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<td>Non-word repetition</td>
<td>Children's test of Non-word Repetition (CNRep)</td>
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<tr>
<td>Speech articulation</td>
<td>Goldman Fristoe Test of Articulation 2 (GFTA-2)</td>
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<tr>
<td>Speech recognition</td>
<td>Listening in Spatialized Noise – Sentences (LiSN-S)</td>
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<td>Pediatric Quality of Life Inventory 4.0(PedsQL 4.0)</td>
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<tr>
<td>Behaviour and emotion</td>
<td>Strengths and Difficulties Questionnaire: Australian version for 4-10 year olds (SDQ)</td>
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<tr>
<td>Non-verbal cognition (IQ)</td>
<td>Wechsler Nonverbal Scale of Ability (WNV)</td>
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Table 4.2. Outcome measures used across historical and contemporary cohorts included in PhD Study 1

<table>
<thead>
<tr>
<th>Construct</th>
<th>Source and measure</th>
<th>Cohort</th>
<th>VicCHILD</th>
<th>SCOUT</th>
<th>CHIVOS</th>
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<td>Clinical Evaluation of Language Fundamentals – Third ed. (CELF-3)</td>
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<td>Preschool Language Scale-4. Aust. language adaptation (PLS-4)</td>
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<td>Receptive vocabulary</td>
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<td>Behaviour and emotion</td>
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<td>Non-verbal cognition (IQ)</td>
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<td>Wechsler Nonverbal Scale of Ability (WNV)</td>
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<td>Wechsler Abbreviated Scale of Intelligence (WASI)</td>
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<td>Wechsler Intelligence Scale for Children (WISC)-Third ed.</td>
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4.10.1 Child-completed measures

Child-completed measures focused on assessing language skills which, as outlined in Chapter 2, is a developmental process known to be vulnerable to interruptions to auditory input.\textsuperscript{209}

Child-completed measures are described by domain. Measures were selected to both collect data on domains of interest and to harmonise with previous studies for comparison purposes. All basal requirements and ceiling rules for the following measures were adhered to as per standardised, published requirements. A basal level, when met, typically indicates an appropriate entry level in a standardised test and indicates that all items prior to this level are considered to be scored correct irrespective of whether they were delivered or not. A ceiling level represents the point at which test administration ceases due to the meeting of a criterion for the number of incorrect responses. All items after this ceiling level are considered incorrect. Often a measure requires both a basal and ceiling be established to calculate a raw score.

All child-completed measures were restricted to participants aged 5-7 years who underwent direct assessment.

4.10.1.1 Speech and language measures

Receptive and expressive language

Receptive and expressive language skills were assessed using the Clinical Evaluation of Language Fundamentals (CELF-4) Australian Standardised Edition for 5-8 year olds.\textsuperscript{210} The CELF-4 is a standardised and normed measure that is widely used in educational and research settings, for both children with and without hearing loss.\textsuperscript{93,158,163,178} The measure has high reliability, with stability coefficients for subtests ranging from .71 to .86, and .88 to .92 for composite scores when based on the standardised population.\textsuperscript{210} The CELF-4 examiner’s manual gives extensive validation information, including a study of 32 students diagnosed with mild to moderate hearing loss who performed more poorly on almost all subtests than controls matched for age, gender and parent education
level. This study suggested that performance on the subtests of the CELF-4 was not due to auditory acuity issues, as the children with hearing loss performed well on a number repetition task; rather, it was hypothesised that underlying difficulties with processing language may have appeared secondary to the hearing problems of the participants.210

The six selected subtests yielded a receptive and expressive language standard score for each participant. Receptive language was assessed using three subtests: Concepts & Following Directions, Word Classes-Receptive, and Sentence Structure. Concepts & Following Directions involves asking the child to point to objects in a stimulus book in response to spoken instructions (e.g. “Point to the big fish and the white apple. Go.” The child responds by pointing to a page showing multiple items.). These commands the child needs to follow increase in length and complexity. The Word Classes-Receptive task requires the child to select two of three words that are related. Instructions are given verbally and accompanied by visual prompts in the stimulus book (e.g. “Tell me the two words that go together best: Cat, whiskers, nest.” The child responds with two of the words?). In the Sentence Structure task, the administrator reads a sentence and the child points to one of four pictures on a single page in the stimulus book that illustrates the sentence (e.g. “Point to ‘The girl has a big, spotted, black-and-white dog.’”). This subtest evaluates the child’s understanding of grammatical rules.

Expressive language was determined by combining scores on the subtests: Word Structure, Recalling Sentences, and Formulated Sentences. Briefly, Word Structure involves the child completing a sentence with a word. (e.g. when faced with a picture in the stimulus book of a boy reading a book, the examiner states the sentence “The boy likes to read. Every day he...” and the child is required to respond with the word “reads”). This subtest focuses on expressive grammar. The Recalling Sentences task requires the student to imitate the sentence that the tester has just presented (e.g. “The tractor was followed by the bus”), which increase in complexity, and is scored according to the number of errors made in their re-telling. In Formulated Sentences, the child is asked to create a sentence
using a target word that is accompanied by a visual prompt in the stimulus book (e.g. when presented with a picture of children leaving a school, the child is asked to make a sentence using the word “car”).

As described above, most material was presented using a stimulus book alongside oral instructions. The Recalling Sentences subtest was performed using recorded sentence stimuli, delivered to the participant via external speakers positioned at a consistent distance of 60 centimetres directly in front of the child. This resulted in an average at-ear presentation level of 60 dBA, consistent with everyday conversation levels. Participant responses were recorded on a USB voice-recorder and/or scored live and transcribed onto CELF-4 record forms.

The receptive and expressive language measures used in the historical cohorts (Study 1) ELVS (CELF-4) and CHIVOS (CELF-3) were either the same or the earlier version of the measure used in active data collection in this PhD. The PLS-4 used in the SCOUT study, as with the various CELF versions, yields a normative mean of 100 with a SD of 15. High stability in language scores over time would suggest that these two standardised measures of expressive and receptive language are measuring the same constructs and are comparable.156

Receptive vocabulary

Receptive vocabulary represents the lexical aspect of language, which involves the understanding and meanings of words. Lexical knowledge is also related to general measures of “crystallized intelligence” (i.e. the ability to use an individual’s skills, knowledge and experience), overall cognitive functioning and success in educational achievement.211 Receptive vocabulary ability could be compromised in the presence of unamplified or poorly-managed hearing loss.

Receptive vocabulary was measured using an automated, adaptive test based on the Picture Vocabulary Test from the National Institutes of Health Toolbox Cognition Battery (NIH-PVT).211 The NIH-PVT presents an audio file of a target word simultaneous to the appearance of four pictures on the computer screen.
The participant would choose the image by tapping the picture that best represents the word heard.

This test has been comprehensively normed, accounting for demographic and cultural factors known to impact test performance. High convergent validity of the NIH-PVT was measured against the Peabody Picture Vocabulary Test-4th edition (PPVT) \( (r=0.78, \ p<0.001) \). Reassuringly, weak discriminant validity measured against the Brief Visuospatial Memory Test-revised and the Rey Auditory Verbal Learning Test \( (r=0.08, \ p<0.001) \) suggests a lack of a relationship with measures evaluating different constructs. A strong intraclass correlation coefficient of 0.84 (95% CI 0.75 to 0.90) for children aged 3-15 years indicates good test-retest reliability. Across the same age range a strong correlation was observed between higher vocabulary scores and increasing age \( (r=0.82, \ p<0.001) \). In summary, the NIH-PVT was shown to be a valid measure of receptive vocabulary ability, obtained in an efficient and computerised adaptive manner.

In order for a vocabulary test to fit within the small available time window of the overall assessment test battery, the test chosen needed to be shorter than standard measures of receptive vocabulary (e.g. the PPVT). Our version of the NIH Picture Vocabulary Test (NPVT) maintained the key attributes of the NIH-PVT as outlined above. Changes involved delivery of the task via an iPad through a custom designed app that the test subject could run independently of the researcher. Features such as a repeat button were available for the stimulus word to be replayed if required, and unambiguous images formed the forced-choice options. The use of external speakers again ensured consistent delivery of the stimulus at a known audibility level. Minor changes to the NIH-PVT included the use of an Australian female speaker with substitutions for any words deemed North American-centric. The custom-designed NPVT was used across multiple studies within the Murdoch Children’s Research Institute that had a need for a rapid, adaptive vocabulary test. Raw scores were converted to scaled scores, and then to age-corrected standard scores using normative data and formulae.
The receptive vocabulary measures used in the historical cohorts with and without hearing loss (Study 1) were, as detailed above, measures that displayed high convergent validity to the NIH-PVT (e.g. the PPVT-4 used in SCOUT) or were the earlier iteration of this measure (i.e. the PPVT-3 in CHIVOS and ELVS). This provides reassurance that the necessary use of different measures across time provide outcomes that can be readily compared.

**Phonological awareness**

Phonological awareness refers to an individual’s awareness of the structure of sounds in their oral language.\(^{213}\) It is suggested that children with awareness that spoken words are constructed from strings of phonemes have an advantage in learning to read,\(^{214}\) with flow-on educational and vocational impact. Phonological awareness in young children (5-10 years) with mild-moderate sensorineural hearing loss has been reported as significantly compromised when compared to controls (mean=97.5, SD=6.4; hearing loss mean=90.6, SD=13.7, \(p=.021\)), yet did not significantly differ from scores achieved by children with specific language impairment (mean 80.6, SD=22.1).\(^{168}\) Yet it has been asserted on the basis of language ability measured in multiple domains, certainly for slight/mild losses, that ultimately normal language could be achieved despite a language system ‘weakened’ by deficits in some domains such as phonology.\(^{88}\)

Phonological awareness was assessed using the Comprehensive Test of Phonological Processing– Second edition (CTOPP-2).\(^{214}\) This measure was normed on a large (n=1900 aged 4-24 years) sample selected to be demographically representative of the US population. Reliability analyses strongly suggest that the measure has little test error for content, time and scorer differences, when considering both subtest and composite score levels.\(^{214}\) The validity of the CTOPP-2 has been documented extensively within the examiners manual across three domains: content-description, criterion-prediction and construct-identification validity.

Three subtest scaled scores of the CTOPP-2 were summed to produce a phonological awareness composite score. Within the age range of participants
(5-7 years), a slightly different combination of subtests was required for those younger (5-6 years) versus older (7 years). Thus all children undertook the Elision and Blending Words subtests. The younger children also completed the Sound Matching subtest, whereas the older children instead completed the Phoneme Isolation subtest.

The Elision subtest assesses the ability to remove phonological segments from spoken words to form a different word. For example, the child is asked to say the word “toothbrush”, then is instructed to say “toothbrush” without saying “tooth”. Or say the word “cup” without saying /k/. The Blending Words subtest assesses the ability to form words from speech sounds. For example, the child is asked what do these sounds make? “t-oi” and the child is required to answer “toy”. The Sound Matching subtest (5-6 years) is a forced choice scenario that measures the child’s ability to select words with the same initial or final sound as an example word. For example, which word starts with the same sound as rain? Tape, line or rope, with the child required to say “rope”. The Phoneme Isolation subtest (7 years) measured the ability to isolate an individual sound within a word. For example, the participant is told that the word man has three sounds, /m/-/a/-/n/, man. What is the first sound – the one at the beginning – in the word man? The child is required to respond with the phoneme /m/. The Blending Words subtest was delivered by a recording of a female North American speaker. Other materials used were a stimulus picture book and an examiner record booklet.

Non-word repetition

Short term memory is how a finite amount of information is temporarily stored in the mind in a very accessible manner. Problems with short-term memory can manifest as an inability to attend to or complete even simple verbal instructions. Such problems can hamper acquisition of skills essential for basic language and literacy competence. Children with poor short-term memories have difficulties learning new words, resulting in slow vocabulary growth which can then impact upstream spoken and written communication. Related to reading ability, short-term memory deficits may reduce a child’s ability to successfully blend and build sounds from letters presented in novel words. For
children with mild to moderate hearing loss, there is evidence of compromised phonological short-term memory.\textsuperscript{168}

Short-term memory was measured using the Children's Test of Non-word Repetition (CNRep).\textsuperscript{216} The test was standardised on children aged 4-9 years who had no known hearing problems. Reliability of the CNRep was good, with a test-retest coefficient of .77 after a four week interval, and a correlation coefficient of .72 following re-administration 14 months after initial testing. Cultural and socio-economic influences are reduced through the use of unfamiliar spoken “non-words”. Validity studies on the CNRep suggest significant correlations with a child's ability to read single words at age 5 and 8 years, in addition to their receptive vocabulary knowledge at 4, 5 and 8 years.\textsuperscript{217}

To complete the CNRep task, the child needed to repeat a non-word out loud immediately after they had heard it. After two practice words of two-syllable length, a further 40 words ranging in length from two to five syllables were presented for the child to repeat. An identical test stimulus (an audio recording of an adult Australian female) was used for all participants, delivered using the same computer speaker configuration detailed above to maximise consistent levels of audibility across participants. A total raw score was the number of non-words pronounced without error. Standard scores were derived from external norms supplied with the CNRep manual.

\textit{Articulation}

Hearing loss can compromise the development of not only language (a system of words and symbols used to communicate) but also speech (the production of spoken language). The ability to successfully produce speech sounds impacts a person’s speech intelligibility. This can be a major issue for children with hearing loss, because it impedes their ability to be understood and also their social interactions. Access to speech sounds in an aided or unaided state can influence articulation skill development in children born with hearing loss.\textsuperscript{170}
Via spontaneous and imitative sound production, the Goldman Fristoe Test of Articulation 2 (GFTA-2)\textsuperscript{218} Sounds-in-Words subtest was used to assess the speech articulation of children. This measure is a widely used tool for assessing production of English consonants. It is a standardised test, normed on 2350 subjects across the United States aged between 2 to 21 years. Psychometrically, the GFTA-2 Sounds-in-words subtest displays high reliability across both sexes, and 98% agreement across error judgement for initial, medial and final sounds during test-retest analyses.\textsuperscript{218} Both content and construct validity were deemed acceptable.

The researcher used an easel of pictures to prompt the child to say the name of particular items illustrated. Across the duration of the test the child named 53 single words, eliciting 77 consonant and consonant cluster sounds in different positions of the spoken word (i.e. initial, medial or final position). The child’s responses were audio recorded for scoring and checking purposes. Target phonemes were recorded as correct, incorrect or not produced during administration. Raw scores ranged from 0 to 77, representing the number of articulation errors across the test. Standard scores were derived from these raw scores, with possible scores ranging from <40 to 116, dependent on child sex and corrected age at assessment.

\subsection*{4.10.1.2 Nonverbal IQ}

\textit{Non-verbal intelligence}

In both children with normal hearing and hearing loss, intelligence is a known determinant of language and learning outcomes (see Section 2.6.3). However, hearing loss is 40 times more prevalent in those with intellectual disability versus the general population,\textsuperscript{219} with intellectual disability believed to increase the impact of hearing loss on childhood cognitive and language development.\textsuperscript{220} Therefore, non-verbal intelligence was measured to be able to consider this construct as a potential confounder via statistical analysis. Varying intelligence between participants, if uncontrolled, has the potential to confound results as within any defined population such as children with hearing loss. Those with greater cognitive skills may have a greater likelihood of scoring better than those
with lower cognition. In order to conclude how all participants within a defined population are performing, the potentially advantageous factors need to be accounted for.

The Wechsler Non-Verbal Scale of Ability (WNV) measured general cognitive ability using nonverbal subtests through the use of pictorial directions. It is a widely-used assessment designed specifically for linguistically diverse populations, including for those with hearing loss. There is high concurrent validity between the WNV and other standardised measures of ability, with the full scale (four-subtest) measure correlating with the Wechsler Intelligence Scale for Children-IV (WISC) (.76) and Wechsler Preschool and Primary Scale of Intelligence (.71), amongst others. No significant difference in either subtest or full scale scores was seen when performance on the WNV by a cohort of participants with hearing impairment was compared to a matched control group. In this PhD the two-subtest WNV battery was used in order to obtain a broad examination of general ability within a tight assessment time period. The overall reliability of the two-subtest full scale score is 0.91, with reliability in a hearing impaired population for the two subtests ranging 0.93-0.97. Adequate stability in WNV scores are reported across time.

The two subtests used for the age range 4 to 7 years are Matrices and Recognition. For the Matrices subtest, a stimulus book placed in front of the child was used to assess how the child understood a pattern of increasingly complex geometric figures. The child is required to understand and interpret a certain geometric design and select, by pointing, the appropriate answer from four or five similar options. The subtest was stopped once the child scored four incorrect answers on five consecutive items. For the Recognition subtest, the child was again presented with a stimulus book. The child is shown a geometric design for three seconds on one page, then on the next page is required to point to the identical design when presented along with three or four other options. As per the Matrices subtest, the Recognition task increased in complexity and followed the same discontinue rule. T scores were calculated from subtest raw
scores using age appropriate values, with a sum of T scores then converted to a full-scale standardised score.

The non-verbal intelligence measures used in the historical cohorts with hearing loss (Study 1) were either identical (SCOUT) or had high concurrent validity (CHIVOS) to the WNV employed in the active data collection of this PhD. The Wechsler Abbreviated Scale of Intelligence used in ELVS (assessing children without hearing loss) was designed to allow sub-test score substitution with more comprehensive intelligence measures such as the Wechsler Intelligence Scale for Children,222 providing confidence that scores across these standardised measures are generally comparable.

4.10.1.3 Speech recognition

**Speech recognition**
Knowledge of not just how an individual detects sound but actually perceives auditory information is valuable for understanding the functional impact of hearing loss. In a population with hearing loss, this can inform attempts to either improve the quality of the signal received at the ear, or focus attention on auditory training to improve use of the signal.223 A task of identifying speech in the presence of competing background noise was used in this PhD. Assessment of auditory recognition under conditions as close to real-life situations as possible can be highly informative, as it more accurately reflects the challenges experienced in regular communication scenarios.

The speech recognition test used in this study was the Listening in Spatialized Noise – Sentences test (LiSN-S).224 An adaptive test, the LiSN-S rapidly determines a signal to noise ratio at which the subject can correctly identify 50% of the stimuli. The child wears headphones, which are connected to a computer to deliver the test as outlined below.

Algorithms are employed to create a virtual three-dimensional acoustic space under the headphones in which different listening conditions are assessed. Of the
four test configurations available, the Different Voices ± 90° (DV90) condition was used, with target sentences sounding as if spoken directly in front of the child and the competing speech from different voiced talkers presented binaurally. This stimuli configuration was selected as it best replicated the challenging listening environment that these school-aged children often encounter. The Prescribed Gain Amplifier (PGA) within LiSN-S delivered both the competing and target speech at levels calculated within the software as consistent with the child's aided hearing levels. If the child was unaided, the PGA was not used and delivery occurred without amplification. In the unamplified scenario the LiSN-S task starts with the target sentence presented at 62 dB SPL, adjusted against a constant level in the competing speech of 55 dB SPL via the process outlined below.

Up to 30 open set sentences of a maximum seven words were presented. The number of words correctly repeated was scored per sentence. Based on this score, the next target sentence was automatically adapted relative to the competing speech. If the child repeated less than half the words in the target sentence correctly, the subsequent sentence’s intensity was raised, resulting in an easier signal-to-noise ratio. If more than half the words in a sentence were repeated correctly, the sentence intensity (loudness) was lowered relative to the competing speech, creating a more challenging signal-to-noise ratio. If exactly half the number of words in the sentence were repeated correctly, no change was made to the signal-to-noise ratio for the next test item. An adaptive procedure such as this is valuable to researchers and clinicians alike, allowing for rapid assessment of speech recognition. A Speech Reception Threshold (SRT) was calculated automatically either (a) once the 30 sentences were all complete, or (b) the minimum number of sentences had been delivered (n=22) and the standard error in their scores is less than 1dB. Australian norms exist for people aged 6 to 60 years. As some of the participants in this study were younger than the norms range, raw SRT scores were used in analyses.
4.10.2 Parent completed measures

Parent‐completed measures relating to their child are described by domain. As with child‐completed measures, harmonisation with data collected in comparison studies influenced the decision to use specific measures. Parents or caregivers were asked to complete these measures at the child’s direct assessment (or by mail when direct assessment was not agreed to) for children aged 5‐7 years. For children in age group 2 who were 1‐3 years of age, parents or caregivers completed the measures when the ELQ was received in the mail. Only the measures relevant for this PhD study are described; in practice they were presented to parents within the same questionnaire as the broader set of VicCHILD measures.

**Behavioural difficulties (Appendix E, section 2)**

Behavioural difficulties have been reported to occur in children with hearing loss, including milder degrees of loss.84, 226 Parents or caregivers of a child with a hearing loss at 5‐7 years completed the Strengths and Difficulties Questionnaire: Australian version for 4‐10 year olds (SDQ).227 This is a widely used and validated measure of psychological characteristics, screening for child emotional and behavioural problems. Satisfactory reliability, good internal consistency (.73) and good test‐retest reliability across a 4‐6 month interval (.62) is reported.227 Normative data exists for both older and younger aged children.228, 229 There are a total of twenty‐five statements divided under five subscales: conduct, emotional symptoms, hyperactivity, peer relationships, and pro‐social behaviour. Higher scores within each subscale (excluding the pro-social scale) represented more problems within that domain. For each item a score from 0‐2 was assigned if a parent considered the statement ‘not true (0)’, ‘somewhat true (1)’ or ‘certainly true (2)’ although, several items were reverse coded. A total difficulties score was calculated by summing the four difficulties scales, excluding the pro-social questions. Again, higher scores represented greater behavioural difficulties.

Of the historical cohorts included in Study 1, two also measured behavioural difficulties using the SDQ (SCOUT and ELVS). The Revised Rutter Parent Scale for
School-age Children and teacher version was used in the CHIVOS study to measure the same construct. Scores on the SDQ and Revised Rutter are shown to be highly correlated \((r=0.88-0.92)\) and have comparable predictive validity.\textsuperscript{230} Therefore, the use of different tools to measure behavioural difficulties across the included cohorts is unlikely to have significantly impacted result interpretation.

\textit{Health-related quality of life (Appendix E, section 1)}

Children with hearing loss are frequently noted to have poorer scores on quality of life measures than children without hearing loss.\textsuperscript{84,107} The Pediatric Quality of Life Inventory 4.0 (PedsQL)\textsuperscript{231} was used to assess health-related quality of life for the child with hearing loss. The measure has been validated for children aged 2-18 years, and the versions relevant to 1-3 year olds and 5-7 year olds were collected in this PhD. Reliability for the total scales score approach or exceed the recommended reliability criterion of .90.\textsuperscript{232} The scale consisted of 23 items split across four sections for 5-7 year olds: physical, social, emotional and school functioning. The scale contained 16 items split across the same four sections for children at age 1-3 years. Caregivers rated how much of a problem the individual items had been for their child over the past one month, using a five point scale: ‘never (0)’, ‘almost never (1)’, ‘sometimes (2)’, ‘often (3)’, and ‘almost always (4)’. Scores were re-coded to range from 0-100 \((0=100, 1=75, 2=50, 3=25, 4=0)\). The total scale score was the sum across the four sections outlined, with a higher score indicating better health-related quality of life.

The PedsQL was used widely across the historical cohorts included in Study 1 of this PhD. However, CHIVOS used the 28-item Child Heath Questionnaire. Both the PedsQL and the Child Health Questionnaire are consistent in being parent/caregiver-proxy report measures. In two separate studies in children with sickle cell disease, both the Child Health Questionnaire and the PedsQL were found to be valid and reliable tools to measure health-related quality of life in children.\textsuperscript{233, 234} This is some evidence towards the convergence of two measures of quality of life in children with a chronic health condition.
Macarthur Bates Communicative Development Inventory: Words and Sentences (Appendix G, section 4)

In comparison to those children in the 5-7 year old age group, there is increased difficulty in obtaining language measures directly from children at younger ages. Therefore, parents of children in the 1-3 year old age group completed the Macarthur-Bates Communicative Development Inventory (MCDI): Words and Sentences version\textsuperscript{235} to obtain a measure of language ability in this younger age bracket. The version of the MCDI included in the ELQ harmonised with that used for the comparison cohort of children without hearing loss; both had 24 vocabulary items substituted to accommodate differences between American and Australian English. Whilst the entire MCDI was included in the ELQ, only the expressive vocabulary production scores were used in this PhD. Raw scores were calculated and percentile scores were obtained.

Norming for this measure has been updated since the first sample was collected in 1988-89 at 3 sites in the United States. The updated sample of 1461 children (728 girls) ranging in age from 16 to 30 months were more diverse than the original sample in ethnicity and educational attainment of participants. The updated sample was drawn from 8 geographic states in the US\textsuperscript{235} Internal consistency of the MCDI is reported as high, with Cronbach’s coefficient alpha of .96. This indicates high agreement that different items within the same category are measuring the same content domain\textsuperscript{235} Reliability of the MCDI Words and Sentences vocabulary production was .95 (p<.01), indicating high correlation between test administrations to the same participants separated on average by 1.38 months. Concurrent validity is also reported to be high. Numerous studies provide correlational validity for the parent-report vocabulary measure with actual child performance on standardised measures of language development. This includes studies of children with hearing loss\textsuperscript{235,236}

Functional assessments (Appendix E)

Functional assessments or functional auditory outcome measures are used to assess how an individual is using their hearing (see Section 2.2.4.3 for further details). Such assessments also make it possible to gauge whether there are any
concerns that could be addressed by providing or adjusting amplification. Completed by caregivers or teachers, these tools provide insight into how well hearing is used in daily life.

The Parents’ Evaluation of Aural/oral performance of Children (PEACH) rating scale\textsuperscript{23} was used to obtain a measure of functional performance from parents or caregivers of a child with a hearing loss in both study age ranges. Three yes/no pre-rating questions aim to determine if the responses to the assessment questions will be reflective of the ideal scenario of (1) a healthy child, (2) who has been wearing their amplification and (3) the amplification has been working correctly. The PEACH rating scale was designed such that if “no” was answered to any of the three questions, the measure should not be used. However, there were some children in this PhD sample who were not fitted with amplification. As such, only a response of “no” to question 1 regarding child health was used to exclude subsequent parent/caregiver responses.

The PEACH then poses 2 questions assessing how often the amplification is worn and whether loud sounds cause distress or complaints, followed by 11 questions about a range of scenarios relating to listening performance. The PEACH asks parents to rate their child’s listening behaviour in respect to their observations over the previous week. The areas these questions cover include responsiveness to environmental sounds, communication in both noise and quiet, and telephone use. A five-point scale is used to rate these questions, with an accompanying percentage of time guide: ‘never (0) 0%’, ‘seldom (1) 1-25%’, ‘sometimes (2) 26-50%’, ‘often (3) 51-75%’, ‘always (4) 75-100%’.

Raw scores were transformed if reverse scoring applied to the item, and then summed to calculate two subscale performance scores, for quiet and noise conditions. These two scores were summed to calculate an overall PEACH score. Published norms for the earlier PEACH Diary version have been shown to be valid for use with the PEACH rating scale.\textsuperscript{23} Low PEACH scores are reported at younger ages: low scores reported up to 40 months of age for the original diary version\textsuperscript{23} and scores reported as significantly lower for children under 20
months of age compared to children over 20 months of age. These scores reportedly rise to approximately 85% by around 30 months of age in a normal hearing sample from a Canadian validation study. These same researchers also report that a score of 90% by a child’s third birthday is a reasonable goal, a statement attributed to the authors of the PEACH.

Hearing device use supplement (Appendices E and G)

As indicated in Section 4.5.1.2.1, to minimise the risk of recall bias the participant hearing device use was to be ascertained from two sources: data logging downloaded from hearing device programming records, and via parent questionnaire. As explained in Section 4.8.1.1.1, device data logging could not be reliably obtained. To this end, the sole method of determining hearing device use was via parent questionnaires (i.e. the PEACH and this described supplement). The hearing device use supplement obtained parent reports of how many hours in total and over what waking hours their child used their hearing aids. Parents initially were asked to report the total hours of hearing aid use for a typical weekday. They were then asked to indicate, via a visual scale, over which hours of a typical weekday their child was wearing their hearing aid. The scale of the day was broken into hourly-blocks from 7am to 10pm (or 6am to 8pm for children in the 1-3 year old age group). It was believed that this dual method of obtaining parent-reported hearing aid usage data could potentially provide a more accurate estimation than asking a simple hours of use per day question alone.

The supplement then asked whether there was a difference in hearing aid usage between weekdays and weekend days. If there was, the dual method of determining hours of weekend hearing aid use was then completed. Questions were also asked about how much of the time (rated from 0 = never, 1 = seldom, 2 = sometimes, 3 = often, 4 = always, 5 = don’t know) the child used their devices in particular age-appropriate situations (e.g. playing with parents, in the car, etc.). These situational-based questions were not included in the data analysis of this PhD. See Appendices E and G for the layout of these questions.
4.10.2.1 Demographic measures
The majority of participants in VicCHILD, the study in which this PhD is nested, provided demographic information at the time of enrolment and subsequent updates at scheduled points of contact throughout the child's development. Relevant demographic details were extracted from these broader VicCHILD parent questionnaires administered to all participants at enrolment, 1-3 years or 5-7 years of age.

Child hearing and communication (Appendix E – question 4.1 to 4.3 (5-7 years) and Appendix G – question 2.3 to 2.5 (1-3 years))
These questions were asked as hearing device fitting may not have occurred by enrolment date. Parents completed the question whether the child had ever been fitted with hearing aids, and if so at what age (in years and months). If hearing aids were no longer used, the age at which use ceased was also collected, in years and months. The responses to these questions were used to check whether there was a discrepancy with data obtained from Australian Hearing, with caregivers contacted for clarification if any questions arose about current aided status and age at first hearing aid fitting.

Child sex (not included in the Appendices)
Child sex was asked in the VicCHILD participant enrolment questionnaire and confirmed against VIHSP-supplied sex details. Therefore, for this study child sex was obtained from the VicCHILD database.

Language used in the home (Appendix E – question 7.5 and 7.6 (5-7 years))
The language mainly used in the home was asked, with the choice being between ‘English’, ‘Auslan’, or ‘Other’ with space provided to specify the language. The second question then asked whether any other languages are regularly used in the home. The responses to these two questions were used to determine whether a child lives in a home where a language other than English is spoken.

If there was any ambiguity about this classification based on the responses to the two questions, the caregiver was contacted by telephone for clarification.
Language used in the home for participants in the younger age group (1-3 years) was obtained from the VicCHILD participant enrolment questionnaire.

**Caregiver education level (not included in appendices)**

Caregiver education level had been collected from all VicCHILD participating families in the participant enrolment questionnaire and entered into the VicCHILD databank. Questions around caregiver education level were not repeated in the follow-up questionnaire for the 1-3 year old or 5-7 year old assessment. In the enrolment questionnaire, caregivers were asked to report their last year of completed schooling (i.e. year 9 or less, year 10, 11, or 12) and their highest level of post-school education (i.e. trade apprenticeship, tertiary degree, postgraduate degree, does not apply). From these measures, one binary variable was used: completed tertiary study (yes/no).

**Neighbourhood disadvantage for the family (not included in Appendices)**

The Socio-Economic Indexes for Areas (SEIFA) was used to attribute a measure of disadvantage for participating families. SEIFA is a national Australian measure devised by the Australian Bureau of Statistics from Census data. SEIFA is comprised of four indexes which each focus on different aspects of socio-economic advantage and disadvantage, representing a summary of relevant census variables. The Australian Bureau of Statistics defines relative socio-economic advantage and disadvantage in terms of “people's access to material and social resources, and their ability to participate in society”. For all households within a given geographic area (represented by postcode), SEIFA averages the responses for relevant variables reported in the Census. The SEIFA score derived at this postcode or neighbourhood level can then be used to represent the degree of disadvantage experienced at an individual family level. The SEIFA is standardised to have a mean national score of 1000 (SD 100), with higher scores reflecting less disadvantage.

Postcodes were obtained from the VicCHILD databank for all families in both age ranges. Via statistical software accessing SEIFA standardised scores, the researcher calculated a SEIFA ranking for each participating family.
4.11 Data management

4.11.1 Overview
This section describes in general terms the data management methods employed in this PhD study. It covers strategies for minimising missing data and methods of ensuring data accuracy. The analysis approach is then addressed in Section 4.12.

4.11.2 Missing data
As questionnaires were completed and returned to the researcher at the time of the child's direct assessment, visual scanning for missing data occurred in the presence of the caregiver. If it was apparent that one section or questionnaire had not been completed, the caregiver was asked to complete it on the spot. More comprehensive inspection of questionnaires occurred once back at the Murdoch Children’s Research Institute on all questionnaires, both those returned at the assessment and those posted back independently. If further omissions were observed, the respondent was contacted by phone and asked to complete the question/s verbally. These actions resulted in very little missing data for parent-completed questionnaires.

For direct measures administered to children at assessment, missing data on different measures occurred primarily as a result of non-compliance and/or insufficient time. The test battery was long, and fatigue was a factor in the decision to not administer some measures. Where possible, missing data were avoided by taking short breaks and returning to tasks where time allowed, along with the use of motivational rewards (e.g. a sticker chart documenting progress). Where possible, basal scores were imputed for those children who were unable to perform the measure due to a lack of comprehension of the task/intellectual disability.

4.11.3 Accuracy
Data accuracy was maintained using several techniques. Firstly, close attention was paid to the labeling of data with participant ID, to ensure no confusion about the origins of the data on multiple record sheets. Researchers verified that the
participant ID matched the record number of the VicCHILD REDCap database (see Section 4.3 for more detail) when data were converted from physical record to electronic record.

A VicCHILD data cleaning manual assisted in initial material screening. For example, this manual was used when deciding which response to enter in the database if multiple responses were indicated for a single‐response item. Scoring of measures requiring calculation of subscales and total scores occurred according to instructions in the VicCHILD Visit Protocol Manual. For any scoring questions not covered by the manual, a research team meeting was regularly convened to determine the correct method of scoring and to document the decision for future reference.

Further cleaning occurred once data were in electronic format, by randomly selecting 10% of physical copies to visually check against the entered data. If errors were identified, these were checked and corrected. Overall an error rate of 1% across the 10% of physical questionnaires checked was deemed acceptable. If a higher error rate was detected, a further 10% of physical data were to be checked against electronic data. However, an error rate on the initial randomly selected physical copies was below 1% and no further physical checks were conducted.

The PhD researcher exported all the required data from REDCap in a Microsoft Excel .csv format and then imported into Stata version 14. Each variable was labeled and scored in Stata. All data cleaning and derivation files generated new data files, to ensure the integrity of the raw data was not compromised. Further checks to those described above occurred in Stata, particularly for any measures (direct assessment or parent completed) where standard scores were calculated, to ensure the results fell within the expected range. Outliers were identified via histograms and box and whisker plots. Checking of outliers against hard copy documents occurred to ensure the data had been accurately captured. Using this method, summary measures were verified as accurate and not influenced in their direction by inaccurate outlier values.
4.12 Analysis plan

The purpose of this analysis plan is to describe the types of analyses undertaken, outcomes and exposure variables, and a priori confounders. This section is structured by the three broad PhD aims, with each aim restated for reference.

4.12.1 Aim 1 (Chapter 5)

For children with mild or moderate congenital hearing loss aged 5-8 years of age drawn from four population-derived samples: a) compare language, behaviour and quality of life outcomes between the samples, b) determine whether age of detection predicts outcomes within the four samples pooled, and c) compare mean outcomes of the contemporary sample to a typically developing sample of similar-aged children.

Aim 1 used data from all contributing studies (n=146 children with hearing loss, n=1217 children without hearing loss). It involved both between-groups and longitudinal analyses. Outcomes were receptive and expressive language standard scores, receptive vocabulary standard scores, measures of behaviour problems and health-related quality of life (all continuous variables). Degree of hearing loss, categorised as mild or moderate, was an exposure variable common to all parts of this aim.

For the first part of this aim (i.e. to compare language, behaviour and quality of life outcomes between systems of hearing loss detection), the second exposure variable was system of detection, categorised as: mature UNHS (reference group), newly established UNHS, risk factor screening and opportunistic detection. A linear regression model was constructed with initial control for demographic a priori confounders, including: child sex; parent education level; language other than English household; and family disadvantage (SEIFA scores). A fully adjusted linear regression model was then conducted, incorporating additional a priori confounders of child non-verbal IQ and hearing loss at the time of assessment with the demographic confounders listed above. Analyses were initially conducted on all children from the mature UNHS, newly established UNHS and risk factor screening groups. Subsequently, all children
with intellectual disability were excluded to allow the inclusion of the fourth group (opportunistic detection) in analyses. Trends in outcomes were plotted across system of detection with tests of significance performed. To enable accurate harmonisation with historical cohorts, parent education level was treated as a binary (tertiary/no tertiary education completed) potential confounder. Additional potential confounders to those already listed, such as the presence of other disabilities, could not be included due to sample size limitations and a lack of harmonised data across the component studies (see Section 8.4 for further discussion).

For the second part of this aim (i.e. to determine whether age of detection predicts outcomes within the four samples pooled) the second exposure variable became age of detection in months, categorised as: 0-6 months (reference group), 7-18 months, 19-30 months, 31-42 months and 43-54 months of age. Children with intellectual disability were again excluded to allow pooling of children from four time-points. Fractional polynomials were plotted to graphically observe the influence of age at detection on outcomes for children with mild and moderate hearing loss combined. Demographic and fully adjusted linear regression models (potential confounders as above) were used to estimate mean differences in outcomes by age of detection, striated by degree of loss at diagnosis.

The third part of this aim restricted the hearing loss sample to those detected under the mature UNHS system and introduced the children without hearing loss group (reference group). These two groups became the second exposure variable. The receptive vocabulary outcome was removed as no score for this domain was available for the children without hearing loss group. Demographic and fully adjusted linear regression models (potential confounders as above, other than hearing loss at time of assessment) were used to estimate mean differences in outcomes between the groups.
4.12.2 Aim 2 (Chapter 6)

For a sub-set of early-identified children with mild or moderate congenital hearing loss at 5-7 years of age: a) establish the relationship between a measure of aided audibility and both (i) unaided hearing acuity and (ii) speech recognition ability, and b) quantify the extent to which (i) unaided hearing acuity and (ii) measures of aided audibility are associated with speech/language outcomes.

Aim 2 used data from a sub-set of the mature UNHS cohort (n=14) in cross-sectional analyses. Outcomes were speech reception thresholds and standard scores measuring receptive and expressive language, receptive vocabulary, phonological awareness, non-word repetition and speech articulation (all continuous variables). Exposure variables were measures of hearing: unaided pure tone averages, aided measures of speech intelligibility index and a functional assessment measure (all continuous).

For the first part of this aim (i.e. establish the relationship between a measure of aided audibility and both hearing acuity and speech recognition ability) correlation analyses were performed with accompanying statistical tests of significance.

For the second part of this aim (i.e. quantify the extent to which hearing acuity and measures of aided audibility associate with language and speech outcomes) correlation analyses were first performed. Linear regression modeling was then conducted, with control for *a priori* confounders, including: child sex; parent education level; language other than English household; family disadvantage (SEIFA scores); and child non-verbal IQ. Post-hoc step-wise regression was used to investigate the loss of a significant correlation between measures of hearing and speech articulation. Further descriptive exploration of the variance seen between participants in outcome and calculated exposure variables also occurred post-hoc. Due to the small sample size, these additional analyses must be considered exploratory and are interpreted with due caution.
4.12.3 Aim 3 (Chapter 7)

For early-identified children with mild or moderate congenital hearing loss at 1-3 years of age: a) describe parent reports of expressive vocabulary, and b) compare mean outcomes to a representative sample of similarly-aged children without hearing loss.

Aim 3 focused on the outcomes of children in a separate age group to those in aims 1 and 2. The participants for this aim were sourced from the mature UNHS group (1-3yo, n=20) and the children without hearing loss group (2yo wave 3, n=1711). The outcome of interest was expressive vocabulary raw score (continuous). The exposure variable common across all parts of this aim was degree of hearing loss, categorised as none, mild or moderate.

For the first part of this aim (i.e. report a caregiver-completed measure of expressive vocabulary) descriptive statistics were used to quantify the range of performance seen for children with mild or moderate hearing loss at diagnosis.

For the second part of this aim (i.e. compare mean outcomes to a representative sample of similarly-aged children) linear regression modeling was to occur with adjustment for potential a priori confounders occur as per aim 1 and 2. However, different trends in vocabulary production between the cohorts as participant age increased necessitated a change in analysis plans. Descriptive exploration of the data occurred between the cohorts and in reference to published norms.

Further detail of all analyses can be found in the respective Results chapters.
5. Language outcomes in children aged 5-7 years with milder hearing loss (Aim 1)

5.1 Overview

The results that make up this chapter have been published in the journal *Child: care, health and development*. The paper summarises the relevant literature, followed by an overview of methodology and results presentation. The results section summarises sample characteristics presented by system of hearing loss detection, with graphical representations of adjusted linear regression results showing changes in outcomes across system of detection and age at detection. Adjusted linear regression models are used to estimate mean differences in outcomes between children with hearing loss and those from a community cohort without hearing loss. The results are briefly discussed and are elaborated upon in Chapter 8.
Mild–moderate congenital hearing loss: secular trends in outcomes across four systems of detection


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Abstract

Background Universal newborn hearing screening (UNHS) targets moderate or greater hearing loss. However, UNHS also frequently detects children with mild loss that results in many receiving early treatment. The benefits of this approach are not yet established. We aimed to (i) compare language and psychosocial outcomes between four hearing loss detection systems for children aged 5–8 years with congenital mild–moderate hearing loss; (ii) determine whether age of detection predicts outcomes; and (iii) compare outcomes between children identified via well-established UNHS and the general population.

Methods Linear regression adjusted for potential confounding factors was used throughout. Via a quasi-experimental design, language and psychosocial outcomes were compared across four population-based Australian systems of hearing loss detection: opportunistic detection, born 1991–1993, n = 50; universal risk factor referral born 2003–2005, n = 34; newly established UNHS, born 2003–2005, n = 41; and well-established UNHS, born 2007–2010, n = 21. In pooled analyses, we examined whether age of detection predicted outcomes. Outcomes were similarly compared between the current well-established UNHS system and typically developing children in the Early Language in Victoria Study, born 2003, n = 1217.

Results Age at diagnosis and hearing aid fitting fell steadily across the four systems. For moderate losses, mean expressive language (P for trend .05) and receptive vocabulary (P for trend .06) improved across the four systems, but benefit was not obvious for mild losses. In pooled analyses, diagnosis before age six months predicted better language outcomes for moderate losses. Children with mild–moderate losses exposed to well-established UNHS continue to experience expressive language scores well below children in the general population (adjusted mean difference −8.9 points, 95% CI −14.7 to −3.1).

Conclusions Treatment arising from UNHS appears to be clearly benefitting children with moderate hearing losses. However, rigorous trials are needed to quantify benefits, versus costs and potential harms, of early aiding of children with mild losses.
Introduction

Early diagnosis and amplification offer the potential to prevent, rather than remediate, language disability resulting from bilateral congenital hearing loss (Wake & Carew 2016). Universal newborn hearing screening (UNHS) has now revolutionized the management landscape across the entire spectrum of hearing loss. This includes children born with mild and moderate loss, whose language outcomes show some evidence of better performance when identified early, but on average remain poorer than population norms. For example, in the contemporaneous Outcomes of Children with Hearing Loss (OCHL) study, children with mild and moderate loss had outcomes on average between two-thirds and one standard deviation below their normally hearing peers at age 5 years (Tomblin et al. 2015). Therefore, it is not surprising that audiologic practice has rapidly encompassed amplification for milder losses that were not initially the target condition for UNHS (Bagatto et al. 2016; King 2010).

Any treatment, including early fitting of amplification, should only become routine practice when there is clear evidence that benefits are cost-effective and exceed any treatment burden, stress or negative impact on quality of life (Grimes & Schulz 2002). There is evidence from quasi-randomized and observational studies of better language outcomes when intervention occurs early (Nelson et al. 2008), but also that these benefits are greater with more severe losses (Ching 2015). This suggests the maximal benefit of very early detection and amplification may not be as sizeable for mild losses, when compared with the benefit expected for moderate losses. Quantifying this potential benefit requires targeted research at the milder end of the hearing loss spectrum. Unfortunately, these children comprise a historically less-researched group than those with severe and profound loss (Moeller et al. 2015; Stika et al. 2015).

Ideally, this evidence would be attained by conducting randomized experimental trials of amplification versus no amplification, stratified by degree of loss. However, the surging number of hearing aids fitted for very young children with a mild loss in the better hearing ear has occurred in the absence of such trials (Australian Hearing 2015). At this point, therefore, the best evidence may come from natural experiments, i.e. quasi-experimental studies that compare outcomes of different detection systems within populations that are otherwise similar. Observational longitudinal studies can also provide valuable evidence regarding age of amplification, although there is substantial potential for residual confounding which is likely to promote over-estimates of efficacy (Craig et al. 2012).

Four Australian population-based state-wide detection systems provide the opportunity to compare both quasi-experimental and longitudinal approaches to the question, ‘Does earlier diagnosis and amplification benefit children with mild and moderate hearing loss?’. It is also important to know the extent of deficits these children continue to experience. Over two decades, these detection systems advanced from opportunistic detection, to risk factor screening, through to ‘newly established’ UNHS and to the current ‘mature’ (well-established) UNHS system. Therefore, for population-derived samples of 5 to 8-year-old children with mild or moderate bilateral hearing loss believed congenital, we aimed to:

1. Compare mean language, behaviour and quality of life outcomes between detection systems,
2. Determine whether age of detection predicts these outcomes in the pooled sample, and
3. Compare mean outcomes between the sample exposed to the mature UNHS program and a contemporaneous, typically developing cohort.

Methods

This was primarily a quasi-experimental study utilizing state-wide population cohorts of children with suspected congenital hearing loss. Cohorts were separated by year and/or geographic state of birth. Victoria’s Royal Children’s Hospital Ethics Committee approved all studies, and parents provided written informed consent.

Population samples with hearing loss

Over nearly 20 years, Wake et al. have recruited four distinct cohorts capturing the evolution in detection systems in two Australian states that mirror international changes. The states share similarities in birth cohort size (approximately 75 000 and 88 000 annually in Victoria and New South Wales (NSW) respectively), socio-demographic characteristics and access to diagnostic and intervention services (Wake et al. 2016).

To assemble the cohorts, researchers approached parents of all children registered in state-wide databases as having mild-profound congenital hearing loss that was believed to be present from birth or soon after. The first three cohorts are historical, while the fourth reflects the current UNHS program in Victoria.


4. Mature UNHS – children born April 2007–April 2010 and participating in the state-wide Victorian Childhood Hearing Impairment Longitudinal Databank (VicCHILD), open to all children born with hearing loss in Victoria. The UNHS program was similar to that of Group 3, but with greatly streamlined follow-up processes that minimise delays between identification and accessing services.

These four cohorts were subsequently pooled to conduct longitudinal analyses for our second aim.

Population sample without hearing loss

The Typically Developing group comprised participants from the Early Language in Victoria Study (ELVS) (Reilly et al. 2006), born and recruited in 2003 from six metropolitan local government areas. Australian census data indicated that these spanned the disadvantage–advantage spectrum, thus maximizing population representation. The inclusion of this cohort allowed analyses addressing our third aim.

All source cohorts excluded families with insufficient English to participate (e.g. questionnaires written at Grade 6 level), CHIVOS additionally excluded children with known intellectual disability. Of the hearing loss cohorts, only VicCHILD is open to children who have never used a hearing device. SCOUT and CHIVOS required fitting by 4 and 4.5 years of age respectively.

Sampling and procedures

Participants were aged between 5 and 8 years at assessment, and all (except the typically developing group) were classified as having either mild (20 to 40 decibels pure tone average (dB PTA)) or moderate (41 to 65 dB PTA) hearing loss in the better ear at birth or from soon after. Researchers audited individual study databases for children who met inclusion and exclusion criteria, extracting assessment data for the Opportunistic, Risk Factor, Newly Established UNHS and Typically Developing groups. Procedures to obtain these data were consistent with those outlined for the Mature UNHS group below.

For the Mature UNHS group (VicCHILD sourced), eligible families were approached for participation at which point the expected time commitment was outlined (i.e. a 2-h home visit). Families either agreed to fully participate or opted to complete parent questionnaires only. Children used their usual amplification during assessment if applicable.

Measures

Outcome measures are shown in Table 1. At age 5 to 7 years, Mature UNHS participants were directly assessed on standardized measures of language, receptive vocabulary and non-verbal IQ. Parents reported their child’s behaviour and health-related quality of life. Newly Established UNHS/Risk Factor and Opportunistic Detection participants were similarly assessed in language, receptive vocabulary and IQ at age 5 to 6 years and 7 to 8 years respectively. Data from Typically Developing participants at 7 years of age were also assessed for similar domains. Children from the risk factor, newly established and mature UNHS groups whose physical or intellectual disabilities precluded direct assessment were assigned the following basal scores: receptive/expressive language (basal score = 50; n = 13, 9%), receptive vocabulary (basal score = 20; n = 9, 6%).

Potential a priori confounders covered demographic factors of child sex, parent education level, English as the child’s second language and the Australian census-based Disadvantage Index (SEIFA, national mean 1000, SD 100; higher scores reflect less disadvantage) (Australian Bureau of Statistics 2008). Others were non-verbal IQ (intellectual disability score < 70 or unable to be assessed) and current hearing loss (most recent better ear three-frequency PTA).

Statistical analysis

For Aim 1, we estimated mean group differences between all outcomes using linear regression, adjusting for demographic and then the non-demographic potential confounders listed above. We conducted analyses with all children from the Mature UNHS, Newly Established UNHS and Risk Factor groups, and then repeated analyses only for children without intellectual disability. This facilitated inclusion of the
Table 1. Key measures

<table>
<thead>
<tr>
<th>Construct</th>
<th>Source and measure</th>
<th>Cohort</th>
<th>Additional information</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age at diagnosis</td>
<td>DB: Australian Hearing</td>
<td></td>
<td>Age at confirmed hearing loss</td>
</tr>
<tr>
<td>Receptive and expressive</td>
<td>DA: Clinical Evaluation of Language Fundamentals -</td>
<td></td>
<td>Recursive language (auditory comprehension) and expressive language (expressive</td>
</tr>
<tr>
<td>language</td>
<td>Fourth ed. (KEF-4) (Semel et al. 2006)</td>
<td></td>
<td>communication) scales. All measures are</td>
</tr>
<tr>
<td></td>
<td>DA: Clinical Evaluation of Language Fundamentals -</td>
<td></td>
<td>standardized; normative mean 100, SD 15.</td>
</tr>
<tr>
<td></td>
<td>Third ed. (KEF-3) (Semel et al. 1995)</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>DA: Preschool Language Scale-4. Austral. language</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>adaptation (PLS-4) (Zimmerman et al. 2002)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Receptive vocabulary</td>
<td>DA: NIH-adaptive Picture Vocabulary Test (NPVT)</td>
<td></td>
<td>NPVT: assessment via automated, adaptive iPad delivery with age-corrected standard</td>
</tr>
<tr>
<td></td>
<td>(Weintraub et al. 2013)</td>
<td></td>
<td>scores with mean 100, SD 15.</td>
</tr>
<tr>
<td></td>
<td>DA: The Peabody Picture Vocabulary Test-4 (PPVT-4)</td>
<td></td>
<td>PPVT: 228 items, standardized; mean 100, SD 15.</td>
</tr>
<tr>
<td></td>
<td>(Dunn &amp; Dunn 2007)</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>DA: The Peabody Picture Vocabulary Test-3 (PPVT-3)</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>(Dunn &amp; Dunn 1997)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Health-related quality of</td>
<td>PI: Pediatric Quality of Life Inventory 4.0 (PedQL 4.0)</td>
<td></td>
<td>PedQL 4.0: 23 items. Total score is sum of</td>
</tr>
<tr>
<td>life</td>
<td>(Manni et al. 2003)</td>
<td></td>
<td>physical, social, emotional and school functioning sections. Range: 0 = worst health</td>
</tr>
<tr>
<td></td>
<td>PI: Child Health Questionnaire (Landgraf et al. 1998)</td>
<td></td>
<td>to 100 = best possible health. CHQ: 28 items. Physical and psychosocial summary</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>scores used. Higher score indicates better health.</td>
</tr>
<tr>
<td>Behaviour and emotion</td>
<td>PI: Strengths and Difficulties Questionnaire (Australian</td>
<td></td>
<td>SOQ: 25 items. Scales: conduct, emotional</td>
</tr>
<tr>
<td></td>
<td>version for 4–10 year olds (Goodman 2001)</td>
<td></td>
<td>symptoms, hyperactivity, peer relationships, prosocial behaviour. Total</td>
</tr>
<tr>
<td></td>
<td>PI: Revised Rutter Parent Scale for School-Age</td>
<td></td>
<td>Difficulties score is sum of all difficulties scales excluding prosocial. Range: 0–40</td>
</tr>
<tr>
<td></td>
<td>Children (Hogg et al. 1997)</td>
<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>Revised Rutter: Emotional, conduct and total</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>difficulties scales. Score ≥ 13 indicates behaviour</td>
</tr>
<tr>
<td>Non-verbal cognition (IQ)</td>
<td>DA: Wechsler Nonverbal Scale of Ability (WNV)</td>
<td></td>
<td>WNV: Two-subtest version (Matrices and</td>
</tr>
<tr>
<td></td>
<td>DA: Wechsler Abbreviated Scale of Intelligence (WASI)</td>
<td></td>
<td>Range: 30 to 170.</td>
</tr>
<tr>
<td></td>
<td>(Wechsler 1999)</td>
<td></td>
<td>WASI: Two-subtest version (Block design and</td>
</tr>
<tr>
<td></td>
<td>Third ed. (Wechsler 1991)</td>
<td></td>
<td>scores indicate greater cognitive ability.</td>
</tr>
</tbody>
</table>

Abbreviations: TD: Typically Developing; New Est UNHS; Newly Established UNHS; Oppor, Opportunistic Detection; DA, Direct Assessment; PI, Parent Response; DB, database; VHSP, Victorian Infant Hearing Screening Program.
Opportunistic group, as this cohort originally excluded children with intellectual disability. Plotting fully adjusted means with 95% confidence intervals enabled visualization of trends across the systems of detection against reference means, significance tests for linear trend are also presented.

Aim 2 drew on pooled data from the four hearing loss groups. Fractional polynomials graphically and empirically examined whether age at detection may influence outcomes, independent of detection system, among children without intellectual disability. Linear regression with identical adjustment to Aim 1 quantified the extent to which continuous outcomes were predicted by age of detection, categorised ordinarily into a 0 to 6-month period and then in 12-month divisions to achieve even participant distributions.

For Aim 3, adjusted linear regression estimated mean differences in language outcomes between the Mature UNHS and Typically Developing groups.

Results

Figure 1 shows the participant flow for the four groups. Detailed recruitment figures for the three historical groups are reported elsewhere (Wake et al. 2004; Wake et al. 2016) from the 84 children aged 5–7 years registered with VicCHILD, 31 had mild/moderate hearing loss of whom 67% participated in the Mature UNHS group.

Initial and current hearing losses and non-verbal IQ were similar across the groups (Table 2). A total of 15 children (10.3%) were categorized with intellectual disability: 2 (9.5%) in the Mature UNHS and 13 (17.3%) in the Newly Established UNHS and Risk Factor groups. All children in the historical groups and 19 of the 20 in the Mature UNHS group used amplification. Maternal educational attainment increased over time, in keeping with Australian educational trends.

Age of detection and hearing aid fitting fell significantly over time (Table 3). Thus, children with mild loss detected under Mature UNHS had an estimated diagnosis on average 9 months (95% CI – 22.8 to 4.8) and fitting 19 months (95% CI – 33.2 to –4.8) earlier than under the Newly Established UNHS system.

Secular trends across the four detection systems (Aim 1)

For moderate losses, Fig. 2 appears to indicate benefits associated with the shift from Opportunistic through to UNHS systems for expressive language (P for trend .05) and receptive vocabulary (P for trend .06). Receptive language improvement was modest and not statistically significant. The very large stepwise reduction in age of aiding for mild losses across the four periods (Table 3) was not paralleled by similarly clear upward trends for mild losses, whose expressive language and receptive

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**Figure 1.** Study recruitment from hearing loss cohorts.

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Table 2. Characteristics of the sample

<table>
<thead>
<tr>
<th></th>
<th>Typically Developing (n = 1217)</th>
<th>Mature UNHS (n = 20)</th>
<th>Newly Established UNHS (n = 41)</th>
<th>Risk Factor Screening (n = 34)</th>
<th>Opportunistic Detection (n = 50)</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Child</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Age at assessment in months, mean (SD)</td>
<td>88.2 (2.2)</td>
<td>75.4 (12.3)</td>
<td>65.6 (7.1)</td>
<td>63.0 (3.8)</td>
<td>95.0 (5.7)</td>
</tr>
<tr>
<td>Male sex, %</td>
<td>50.5</td>
<td>47.6</td>
<td>56.1</td>
<td>55.0</td>
<td>62.0</td>
</tr>
<tr>
<td>Severity – 3 freq dB PTA better ear, mean (SD)</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Initial</td>
<td>—</td>
<td>43.6 (11.3)</td>
<td>43.6 (9.4)</td>
<td>41.0 (11.7)</td>
<td>47.0 (10.0)</td>
</tr>
<tr>
<td>Current</td>
<td>—</td>
<td>44.0 (13.6)</td>
<td>50.8 (22.9)</td>
<td>48.0 (26.0)</td>
<td>46.1 (12.0)</td>
</tr>
<tr>
<td>Non-verbal IQ, mean (SD)</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Whole sample</td>
<td>104.7 (14.7)</td>
<td>99.1 (19.2)</td>
<td>94.5 (29.2)</td>
<td>94.1 (25.2)</td>
<td>—</td>
</tr>
<tr>
<td>Children without intellectual disability</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Family</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Language other than English household, %</td>
<td>5.8</td>
<td>14.3</td>
<td>7.3</td>
<td>11.8</td>
<td>6.0</td>
</tr>
<tr>
<td>Disadvantage Index, mean (SD)</td>
<td>1036.0 (60.7)</td>
<td>1007.9 (55.2)</td>
<td>1010.3 (71.1)</td>
<td>1036.8 (50.9)</td>
<td>1016.9 (74.6)</td>
</tr>
<tr>
<td>Parent with at least undergraduate level education, %</td>
<td>38.6</td>
<td>57.1</td>
<td>26.8</td>
<td>41.2</td>
<td>32.0</td>
</tr>
</tbody>
</table>

Abbreviations: PTA, Pure Tone Average; questionnaire, ax. assessment.
1 Percent missing data (mature UNHS sample): non-verbal IQ 9.1%, all other measures complete.
2 Percent missing data (newly established UNHS sample): non-verbal IQ 12.2%, LOTE 2.4%, parent education 9.8%, all other measures complete.
3 Percent missing data (risk factors: LOTE 2.9%, parent education 5.9%, all other measures complete.
4 Percent missing data (opportunist): LOTE 0%, disadvantage index 2%, parent education 0%, all other measures complete.
11 Non-verbal IQ including un-accessible children for whom the basal standard score (30) was imputed.

Table 3. Ages of detection and hearing aid fitting by severity across detection systems (intellectual disability excluded)

<table>
<thead>
<tr>
<th>Outcome in months</th>
<th>Fully adjusted mean 1</th>
<th>Fully adjusted mean difference (95% CI)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Mature Established UNHS (n = 10)</td>
<td>Newly Established UNHS (n = 31)</td>
</tr>
<tr>
<td>Mild</td>
<td>Age detected</td>
<td>4.5</td>
</tr>
<tr>
<td></td>
<td>Age hearing aid fitted</td>
<td>8.3</td>
</tr>
<tr>
<td>Moderate</td>
<td>Age detected</td>
<td>5.5</td>
</tr>
<tr>
<td></td>
<td>Age hearing aid fitted</td>
<td>12.4</td>
</tr>
<tr>
<td>Mild moderate combined</td>
<td>Age detected</td>
<td>5.4</td>
</tr>
<tr>
<td></td>
<td>Age hearing aid fitted</td>
<td>11.2</td>
</tr>
</tbody>
</table>

Percent missing data: Age hearing aid fitted 9.3%; Age detected complete.
1 Adjusted for parent education, English as a second language, disadvantage index, sex, non-verbal IQ and current hearing loss.

Vocabulary remained around two thirds to one standard deviation below normative means. In this context, an apparent late upwelling in receptive language with mature UNHS only (Fig. 2) is difficult to interpret. Health-related quality of life and behaviour appeared largely unaffected by system of detection for either group.
Figure 2. Relationships of mean receptive and expressive language and receptive vocabulary outcomes with evolving systems of hearing loss detection. Horizontal solid and dashed lines represent normative scores and standard deviations. Whiskers represent 95% confidence intervals. Statistical test of significance, P, for trend. Participants with intellectual disability excluded. Abbreviations: Opport., opportunistic; NE UNHS, newly established UNHS; M UNHS, mature UNHS. [Colour figure can be viewed at wileyonlinelibrary.com]
Age of detection and language outcomes in the pooled sample (Aim 2)

Figure 3a shows a single fractional polynomial line of best fit reflecting the trend in each language outcome by age of detection for the pooled mild–moderate sample, while Fig. 3b shows these same trends for the four detection systems overlaid. Language and vocabulary outcomes were higher with detection age before 6 and, unexpectedly, after 30 months, and lower with detection between 6 and 30 months. However, there was little separation of the lines by era of detection.

The regression analyses (Table 4) show that the benefits of earlier detection were almost wholly experienced by children with moderate, not mild, losses. Compared with a diagnosis under 6 months, children with moderate loss diagnosed between 18 and 30 months had poorer receptive and expressive language (fully adjusted mean difference −16.7 points, 95% CI −36.3 to −3.0; −27.0, 95% CI −49.2 to −15.7, respectively) and receptive vocabulary (−16.4, 95% CI −28.1 to −4.7). Outcomes for mild losses showed little association with age at detection, other than significantly poorer receptive language when detected between 6 and 18 months.

Comparison of mature UNHS and typically developing groups (Aim 3)

Expressive language was significantly poorer in the mild–moderate combined Mature UNHS than the Typically Developing group (fully adjusted mean difference −8.9 points, 95% CI −14.7 to −3.1). Adjusted receptive language, behaviour and health-related quality of life scores were similar to Typically Developing children (Table 5).

Discussion

This population-level study showed that, over a nearly 20-year period of births, expressive language and receptive vocabulary in children diagnosed with moderate congenital hearing loss improved steadily as hearing detection systems shifted from opportunistic to mature UNHS. While our small sample size
Table 4. Outcomes by age of detection (in months) for children without intellectual disability.

<table>
<thead>
<tr>
<th>Diagnosis age</th>
<th>0-6 months</th>
<th>Mean diff (95% CI)</th>
<th>P</th>
<th>6-18 months</th>
<th>Mean diff (95% CI)</th>
<th>P</th>
<th>18-30 months</th>
<th>Mean diff (95% CI)</th>
<th>P</th>
<th>30-42 months</th>
<th>Mean diff (95% CI)</th>
<th>P</th>
<th>42-54 months</th>
<th>Mean diff (95% CI)</th>
<th>P</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mild intellectual disability</td>
<td>Receptive language</td>
<td>95.7</td>
<td>80.8</td>
<td>-1.4 (-2.7 to -0.1)</td>
<td>0.02</td>
<td>84.2</td>
<td>-1.5 (-2.7 to 0.7)</td>
<td>0.16</td>
<td>93.5</td>
<td>-2.3 (-3.5 to 1.3)</td>
<td>0.04</td>
<td>98.1</td>
<td>2.4 (-1.9 to 6.7)</td>
<td>0.24</td>
<td></td>
</tr>
<tr>
<td>Moderate intellectual disability</td>
<td>Receptive language</td>
<td>93.1</td>
<td>87.6</td>
<td>-8.5 (-16.1 to 0.1)</td>
<td>0.05</td>
<td>80.1</td>
<td>-8.0 (-15.1 to 0.4)</td>
<td>0.06</td>
<td>71.8</td>
<td>-3.5 (-11.6 to 4.6)</td>
<td>0.03</td>
<td>93.0</td>
<td>-0.9 (-16.0 to 0.4)</td>
<td>0.06</td>
<td></td>
</tr>
<tr>
<td>Mild-moderate combined</td>
<td>Receptive language</td>
<td>93.1</td>
<td>87.6</td>
<td>-8.5 (-16.1 to 0.1)</td>
<td>0.05</td>
<td>80.1</td>
<td>-8.0 (-15.1 to 0.4)</td>
<td>0.06</td>
<td>71.8</td>
<td>-3.5 (-11.6 to 4.6)</td>
<td>0.03</td>
<td>93.0</td>
<td>-0.9 (-16.0 to 0.4)</td>
<td>0.06</td>
<td></td>
</tr>
</tbody>
</table>

*Adjusted for parent education, English as a second language, disadvantage index, sex, non-verbal IQ, and current IQ loss.

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### Table 5. Outcomes by severity for Mature UNHS versus Typically Developing group

<table>
<thead>
<tr>
<th>Outcome</th>
<th>Nomative mean (SD)</th>
<th>Typically developing</th>
<th>Mature UNHS</th>
<th>Mean diff (95% CI)</th>
<th>P</th>
<th>Fully adjusted mean *</th>
<th>Typically developing</th>
<th>Mature UNHS</th>
<th>Mean diff (95% CI)</th>
<th>P</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Mild</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Receptive language</td>
<td>100 (15)</td>
<td>94.3</td>
<td>95.9</td>
<td>1.6 (−7.8 to 11.0)</td>
<td>.74</td>
<td>94.3</td>
<td>101.2</td>
<td>6.9 (−1.9 to 15.7)</td>
<td>.12</td>
<td></td>
</tr>
<tr>
<td>Expressive language</td>
<td>100 (15)</td>
<td>98.9</td>
<td>86.1</td>
<td>−12.8 (−21.9 to −3.7)</td>
<td>.006</td>
<td>99.0</td>
<td>91.5</td>
<td>−7.4 (−16.3 to 1.4)</td>
<td>.10</td>
<td></td>
</tr>
<tr>
<td>Behaviour problems</td>
<td>6.9 (5.1)</td>
<td>7.0</td>
<td>8.8</td>
<td>1.8 (−16 to 5.1)</td>
<td>.30</td>
<td>7.0</td>
<td>9.5</td>
<td>2.6 (−1.0 to 6.1)</td>
<td>.16</td>
<td></td>
</tr>
<tr>
<td>Health-related QOL</td>
<td>81.9 (12.6)</td>
<td>83.4</td>
<td>80.0</td>
<td>−3.5 (−10.6 to 3.6)</td>
<td>.34</td>
<td>83.6</td>
<td>77.7</td>
<td>−5.9 (−13.2 to 1.4)</td>
<td>.11</td>
<td></td>
</tr>
<tr>
<td><strong>Moderate</strong></td>
<td></td>
<td></td>
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<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Receptive language</td>
<td>100 (15)</td>
<td>94.3</td>
<td>95.1</td>
<td>0.6 (−7.7 to 9.3)</td>
<td>.85</td>
<td>94.3</td>
<td>94.9</td>
<td>0.6 (−0.9 to 0.9)</td>
<td>.88</td>
<td></td>
</tr>
<tr>
<td>Expressive language</td>
<td>100 (15)</td>
<td>98.9</td>
<td>89.1</td>
<td>−9.7 (−18.0 to −1.5)</td>
<td>.02</td>
<td>99.0</td>
<td>89.0</td>
<td>−10.0 (−17.5 to −2.4)</td>
<td>.01</td>
<td></td>
</tr>
<tr>
<td>Behaviour problems</td>
<td>6.9 (5.1)</td>
<td>7.0</td>
<td>8.9</td>
<td>1.8 (−1.1 to 4.8)</td>
<td>.22</td>
<td>7.0</td>
<td>8.4</td>
<td>1.4 (−1.8 to 4.6)</td>
<td>.39</td>
<td></td>
</tr>
<tr>
<td>Health-related QOL</td>
<td>81.9 (12.6)</td>
<td>83.4</td>
<td>85.0</td>
<td>2.5 (−3.6 to 8.7)</td>
<td>.42</td>
<td>83.6</td>
<td>85.3</td>
<td>1.9 (−4.6 to 8.3)</td>
<td>.56</td>
<td></td>
</tr>
<tr>
<td><strong>Mild moderate combined</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Receptive language</td>
<td>100 (15)</td>
<td>94.3</td>
<td>95.5</td>
<td>1.2 (−5.1 to 7.6)</td>
<td>.70</td>
<td>94.3</td>
<td>97.5</td>
<td>3.2 (−2.6 to 8.9)</td>
<td>.28</td>
<td></td>
</tr>
<tr>
<td>Expressive language</td>
<td>100 (15)</td>
<td>98.9</td>
<td>87.9</td>
<td>−10.0 (−17.2 to −4.8)</td>
<td>.001</td>
<td>99.0</td>
<td>90.1</td>
<td>−8.9 (−14.7 to −3.1)</td>
<td>.003</td>
<td></td>
</tr>
<tr>
<td>Behaviour problems</td>
<td>6.9 (5.1)</td>
<td>7.0</td>
<td>8.8</td>
<td>1.8 (−0.4 to 6.0)</td>
<td>.11</td>
<td>7.0</td>
<td>8.9</td>
<td>1.9 (−0.5 to 4.3)</td>
<td>.11</td>
<td></td>
</tr>
<tr>
<td>Health-related QOL</td>
<td>81.9 (12.6)</td>
<td>83.4</td>
<td>83.3</td>
<td>−0.1 (−4.8 to 4.6)</td>
<td>.97</td>
<td>83.6</td>
<td>82.0</td>
<td>−1.5 (−6.5 to 3.4)</td>
<td>.54</td>
<td></td>
</tr>
</tbody>
</table>

*Abbreviation: QOL, quality of life.
* Adjusted for parent education, English as a second language, disadvantage index, sex.
* With additional adjustment for non-verbal IQ.

which are plausible and backed up by our larger pooled analyses. We acknowledge that a larger sample size would have firmly conclusions for the mild group, but, unfortunately, to our knowledge no other harmonized cohorts spanning such systems with prospective, same-age standardized outcomes exist internationally nor could now be generated. Our historical cohorts did not record detail on participant recruitment at the hearing loss subgroup level, unlike our thoroughly documented youngest cohort (mature UNHS). How this may have affected generalizability is unknown. Our findings may not fully generalize to children with intellectual disability who were not assessed in the oldest cohort (opportunistic detection). Nonetheless, three-group comparisons including these children did not suggest different conclusions, and their exclusion from the four-group comparisons reduced concerns regarding skewness due to basal scores (Wake et al., 2016). As with all population studies from pre-UNHS era, we cannot be certain that all hearing losses were congenital in two of our four groups. The better outcomes of children detected after age 30 months could be explained by any of chance, late-onset hearing loss being misclassified as congenital and/or better outcomes leading to later diagnosis (i.e. reverse causation). With all children bar one being hearing aid users, results may not generalize to children with unaided losses. As no information on intervention program enrollment or approach was available, we could not quantify any impact on outcomes. Anecdotally, there has been an ongoing trend towards intervention being offered more frequently and inclusively to milder losses. The fact that this was not mirrored in steadily improving performance is concerning.

Unlike children with moderate losses, it is difficult to conclude that children with mild losses are experiencing clear benefit from the profound shift in practice to earlier detection and aiding. This is even more surprising given that both maternal education levels in the general population and amplification technology improved markedly across the two decades separating the oldest from the youngest children. We consider two possible reasons. Children with mild losses may simply not wear adequately fitted aids for enough time each day (Fitzpatrick et al., 2010; Walker et al., 2015), when amplification is subtle and may require prolonged delivery to accrue significant benefit. Alternatively, mild hearing loss could represent ‘overdiagnosis’, defined as identification of a real condition for which treatment does not actually benefit an individual’s outcomes (Coon et al., 2014). This would imply that these children’s developmental deficits might not be attributable solely to their hearing acuity. If so, the decision to amplify mild losses early could represent not only overtreatment (i.e. treatment that cannot deliver benefit) but also active harm (costs, burden, stigmatization).

Under current detection practices, large numbers of children born with mild bilateral hearing loss will continue to be identified early. There is a need for carefully constructed and controlled trials that compare systematic hearing aid provision to no provision, and that accurately measure usage. Outcomes should include both specific and broad language outcomes as well as costs and health-related quality of life.
Such trials would need to be of adequate size to explore both the hearing thresholds and the minimum ‘dose’ (percent of time hearing aids must be worn and quality of fitting) beyond which benefits appear to accrue. Otherwise, it will remain impossible to move to an evidence-based and cost-effective system that optimizes management for all children with congenital hearing loss based on need and benefit.

In conclusion, for children born with mild hearing loss, we observed limited evidence of benefit to language outcomes as a result of earlier diagnosis and amplification. However, we do observe clear benefits in children born with moderate hearing loss. These results suggest that, despite the best intentions made possible by universal newborn hearing screening, our current approach to treating children with mild loss is not producing consistent improvements across time.

**Key Messages**

- Universal newborn hearing screening (UNHS) often targets moderate or greater hearing losses, but it also frequently detects mild losses.
- Despite the benefits versus costs/harms of earlier detection of mild losses yet to be established, early amplification for these children has rapidly become standard practice.
- For children with moderate losses, language outcomes have improved as detection has moved from opportunistic to risk factor screening to newly established UNHS and mature UNHSs.
- Clear benefits of earlier detection and advanced amplification were not obvious in the measured outcomes for children with mild losses.
- An evidence base for effective clinical care of children with mild losses needs to be established, e.g. using randomised trials of early hearing aid provision.

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**Conflict of interests**

The authors have no conflict of interest to disclose. The authors have no relevant financial relationships to disclose.

**Contributors’ statement**

Mr Carew conceptualized and designed the study, was responsible for the acquisition and interpretation of data, drafted and revised the manuscript.

Dr Mensah provided guidance with analysis and interpretation of the data, and reviewed and revised the manuscript.

Associate Professor Rance assisted with the design of the study, assisted with interpretation of the data and reviewed and revised the manuscript.

Dr Flynn and Poulakis provided guidance regarding data collection instruments and reviewed and revised the manuscript.

Professor Wake led the CHIVOS, SCOUT and VicCHILD cohorts, conceptualized the study, provided guidance regarding data collection instruments, analysis and interpretation of the data and reviewed and revised the manuscript.

**Acknowledgements**

We would like to thank all children, parents and researchers involved in the CHIVOS, SCOUT, VicCHILD and ELVS studies that formed this body of work.

**References**


Australian Hearing (2015) Demographic details of young Australians aged less than 26 years with a hearing impairment, who have been fitted with a hearing aid or cochlear implant at 31 December 2014 [Internet]. Australian Hearing.


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6. Aided audibility in children aged 5-7 years with milder hearing loss (Aim 2)

6.1 Overview
Aim 2 (a) was to establish the relationships of aided audibility with hearing acuity and speech recognition ability in early-identified children with mild and moderate bilateral congenital hearing loss. Aim 2 (b) was to quantify the association between measures of hearing (both unaided and aided) and speech/language outcomes. The findings for this aim are presented in traditional chapter format. The chapter begins by defining the characteristics of the analysed sample, first demographically and then in terms of their hearing characteristics and participant speech and language ability. Results for parts (a) and (b) are then reported, followed by a case study for a participant of particular interest. Findings and their limitations are critically discussed in Chapter 8.

6.2 Sample characteristics
As outlined in methods chapter Section 4.5, the participants in this chapter were a sub-group of the broader sample of children aged 5-7 years born with either a mild or moderate bilateral sensorineural hearing loss. This sub-group comprised the 19 participants who were hearing aid users. Five children were subsequently excluded, one for non-compliance with assessments, and the others for varying audiometric reasons (e.g. auditory neuropathy diagnosis, a bilateral loss that was functionally unilateral). Hence the sample comprised the remaining 14 children (six boys, eight girls, mean 76 months of age, range 60–95 months).

6.2.1 Demographic factors
Table 6.1 summarises participant characteristics. The sample mean non-verbal IQ of 104.8 was slightly higher than the population norm of 100, with a range skewed towards higher cognitive ability. Fatigue accounted for one participant not completing IQ tasks. Half of parents reported university level education, and household disadvantage broadly fell within one standard deviation above and below the population norm. With 21% of households not speaking English as the
primary language at home, this supported the decision to include home language as a potential confounder.

The majority of participants were early-diagnosed and early-amplified. Participants had on average been wearing hearing aids since their first birthday, following a mean diagnosis at age 3.6 months. A sole participant diagnosed at 28 months and fitted a month later influenced the range in age of diagnosis. Two-thirds of participants were fitted with hearing aids by 7 months of age. At the time of language assessment, parent-reported hearing device use suggested participants wore their hearing aids on average 10 hours during weekdays and 8 hours on weekend days. All hearing aids used were those prescribed and maintained by Australian Hearing. No adjustments were made to the hearing aids prior to assessment as part of the research protocol.
<table>
<thead>
<tr>
<th>Characteristic</th>
<th>Mean (SD) n=14</th>
<th>Median (IQR) n=14</th>
<th>Range</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Child</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Male sex, %</td>
<td>43</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>Age at assessment in months</td>
<td>75.8 (12.9)</td>
<td>72.5 (65 to 89)</td>
<td>60 to 95</td>
</tr>
<tr>
<td>Age at diagnosis in months</td>
<td>3.6 (7.2)</td>
<td>1.5 (1.1 to 2.4)</td>
<td>0.5 to 28.6</td>
</tr>
<tr>
<td>Age hearing aid first fit in months</td>
<td>12.8 (15.5)</td>
<td>4.2 (2.9 to 16.2)</td>
<td>2.2 to 47.0</td>
</tr>
<tr>
<td>Hearing aid use in hours, weekdays</td>
<td>10.3 (2.3)</td>
<td>10.5 (8.0 to 12.0)</td>
<td>6.0 to 13.0</td>
</tr>
<tr>
<td>Hearing aid use in hours, weekend days</td>
<td>8.4 (4.7)</td>
<td>9 (6.0 to 12.0)</td>
<td>0 to 14.0</td>
</tr>
<tr>
<td>Non-verbal IQ*</td>
<td>104.8 (6.8)</td>
<td>103 (101 to 109)</td>
<td>95 to 117</td>
</tr>
<tr>
<td><strong>Family</strong></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Language other than English household, %</td>
<td>21</td>
<td>-</td>
<td>-</td>
</tr>
<tr>
<td>Disadvantage Index</td>
<td>1006 (60.9)</td>
<td>1024 (996 to 1049)</td>
<td>878 to 1068</td>
</tr>
<tr>
<td>Parent with university level education, %</td>
<td>50</td>
<td>-</td>
<td>-</td>
</tr>
</tbody>
</table>

*n=13*
6.2.2 Hearing characteristics

A better ear pure tone average (PTA) of 43.4 dB HL was calculated for the group (Table 6.2). No significant asymmetry in hearing levels between the ears was observed. Figure 6.1 graphically displays mean audiometric thresholds and standard deviations for the group. Table 6.2 shows both calculated and parent-reported measures of aural function. A speech reception threshold (SRT) was calculated for 11 participants who completed Listening in Spatialised Noise – Sentences (LiSN-S) testing, with a mean threshold of speech stimuli 3.5 dB lower than the background noise level. This is poorer than the performance of children with normal hearing, with norms indicating a mean SRT score ranging from -9.1 to -12.7 dB across the ages 5-7 years. A wide range of speech intelligibility index (SII) scores were obtained for each speech input level, reflecting what appeared to be considerable variation in aided audibility. Mean SII scores indicated that for each 10 dB increase in the loudness of speech, from soft to medium and medium to loud, audibility of speech improved by approximately 25%.

The mean PEACH total score, combining results of the quiet and noise subscales for how frequently children with hearing loss react to auditory input, was 79.1% (standard deviation 11.3, range 65.9% to 97.7%). Whilst a wide range of scores were obtained on the PEACH, this group mean was lower than the approximately 85% overall PEACH score expected in normally hearing children for the mean age of our study cohort.
Table 6.2. Summary of hearing related, language and speech outcome scores for Aim 2 participants

<table>
<thead>
<tr>
<th>Measure</th>
<th>n</th>
<th>Mean (SD)</th>
<th>Median (IQR)</th>
<th>Range</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Hearing</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Better ear Pure Tone Average, dB HL</td>
<td>14</td>
<td>43.4 (10.9)</td>
<td>42.0 (35.0 to 51.0)</td>
<td>31.0 to 65.0</td>
</tr>
<tr>
<td>Speech Reception Threshold, dB SNR</td>
<td>11</td>
<td>-3.5 (2.6)</td>
<td>-3.7 (-5.9 to -2.4)</td>
<td>-7.0 to 1.6</td>
</tr>
<tr>
<td><strong>Aided Audibility - SII</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Soft speech (55 dB SPL)</td>
<td>14</td>
<td>28.2 (14.5)</td>
<td>30 (14 to 37)</td>
<td>6 to 50</td>
</tr>
<tr>
<td>Medium speech (65 dB SPL)</td>
<td>14</td>
<td>50.0 (16.3)</td>
<td>49 (41 to 63)</td>
<td>15 to 70</td>
</tr>
<tr>
<td>Loud speech (75 dB SPL)</td>
<td>14</td>
<td>76.5 (18.9)</td>
<td>80 (75 to 90)</td>
<td>26 to 91</td>
</tr>
<tr>
<td><strong>Functional Assessment</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>PEACH total score, %</td>
<td>14</td>
<td>79.1 (11.3)</td>
<td>78.4 (68.1 to 90.9)</td>
<td>65.9 to 97.7</td>
</tr>
<tr>
<td><strong>Language</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Receptive language</td>
<td>13</td>
<td>101.8 (17.9)</td>
<td>98 (90 to 118)</td>
<td>75 to 125</td>
</tr>
<tr>
<td>Expressive language</td>
<td>13</td>
<td>95.2 (20.1)</td>
<td>99 (80 to 102)</td>
<td>61 to 138</td>
</tr>
<tr>
<td>Receptive vocabulary</td>
<td>14</td>
<td>91.2 (22.8)</td>
<td>98.5 (78.4 to 103.1)</td>
<td>48 to 119</td>
</tr>
<tr>
<td>Phonological awareness</td>
<td>12</td>
<td>89 (12.1)</td>
<td>91 (80 to 96)</td>
<td>67 to 112</td>
</tr>
<tr>
<td>Non-word repetition</td>
<td>13</td>
<td>93.8 (21.1)</td>
<td>96 (89 to 104)</td>
<td>55 to 135</td>
</tr>
<tr>
<td><strong>Speech</strong></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Articulation</td>
<td>13</td>
<td>100.2 (10.0)</td>
<td>105 (89 to 107)</td>
<td>81 to 110</td>
</tr>
</tbody>
</table>

Abbreviations: dB HL, decibels hearing level; dB SNR, decibels signal to noise ratio; dB SPL, decibels sound pressure level.
Figure 6.1. Mean participant audiometric thresholds, plotted with standard deviations per threshold

6.2.3 Speech and language ability of the sample

Participants included in the analyses for this aim were a sub-group of the broader early identified group of 5-7 year old children reported in Chapter 5.

Contrary to the expected equivalence between language and cognitive outcomes, all mean scores on speech/language measures were lower than the mean non-verbal IQ for the sample. They ranged from meeting the population normative mean (receptive language, articulation) to falling around two thirds of a standard deviation below (expressive language, receptive vocabulary, phonological awareness and articulation) (Table 6.2). Similar to the SII and PEACH results, much wider variability was seen in individual speech/language outcomes than in the sample non-verbal IQ.
6.3 Association between SII, PTA and speech recognition (Aim 2 (a))

This section reports the relationship of aided audibility (SII) with hearing acuity and speech recognition ability in early-identified children with mild and moderate bilateral congenital hearing loss.

Similar and large significant negative correlations were observed between better ear PTA and same-ear SII calculated at the three different input levels (SII soft speech \( r = -0.60, p = 0.02 \); SII medium speech \( r = -0.78, p = 0.001 \); SII loud speech \( r = -0.79, p < 0.001 \)) (Figure 6.2). In other words, children with worse hearing acuity (higher thresholds) also had poorer speech intelligibility indices (lower SII). Better ear PTA and speech recognition as defined by the speech reception threshold (SRT) also showed a large correlation \( r = 0.65, p = 0.02 \), with poorer speech recognition observed for children with poorer PTAs. Tests of correlation between SRT and SII scores are illustrated in Figure 6.3, with similarly large and significant negative correlations documented for soft \( r = -0.63, p = 0.04 \), medium \( r = -0.73, p = 0.01 \) and loud \( r = -0.70, p = 0.02 \) inputs. These results provide support for the Situational Hearing Aid Response Profile-calculated SII being consistent with both the unaided measure of hearing ability (PTA) and actual amplified speech recognition scores in competing speech for this group of children with mild and moderate hearing loss.
Figure 6.2. Correlation between SII and better ear PTA
Soft speech correlation coefficient $p < .02$, medium speech $p = .001$ and loud speech $p < .001$, $n=14$.

Figure 6.3. Correlation between SII and Speech Reception Threshold
Soft speech correlation coefficient $p < .04$, medium speech $p = .01$, loud speech $p < .02$, $n=11$. 
6.4 Association between measures of hearing and speech/language outcomes (Aim 2 (b))

This section outlines the results that address the second aim of this chapter, i.e. to establish associations between measures of hearing (both unaided and aided) and speech/language outcomes.

Correlation analyses of unaided hearing (PTA) and aided audibility (SII, speech recognition and functional assessments) exposures with speech/language outcomes were performed. Correlations were small in size and did not reach statistical significance for receptive language, expressive language, receptive vocabulary, phonological awareness or non-word repetition scores (see Table 6.3). However, better articulation was correlated with both better (i.e. lower) PTA ($r = -.63, p=.02$) and better (i.e. higher) SII for loud speech ($r=.57, p=.04$).
Table 6.3. Correlation coefficients for language outcomes and measures of aided and unaided hearing

<table>
<thead>
<tr>
<th>Language outcome</th>
<th>BEPTA (4FA dB HL)</th>
<th>SII Soft (55 dB SPL)</th>
<th>SII Medium (65 dB SPL)</th>
<th>SII Loud (75 dB SPL)</th>
<th>SRT</th>
<th>PEACH</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>r</td>
<td>p</td>
<td>r</td>
<td>p</td>
<td>r</td>
<td>p</td>
</tr>
<tr>
<td>Receptive language (n=13)</td>
<td>-.02</td>
<td>.94</td>
<td>.02</td>
<td>.95</td>
<td>.09</td>
<td>.77</td>
</tr>
<tr>
<td>Expressive language (n=13)</td>
<td>-.11</td>
<td>.70</td>
<td>-.11</td>
<td>.71</td>
<td>.13</td>
<td>.67</td>
</tr>
<tr>
<td>Receptive vocabulary (n=14)</td>
<td>-.07</td>
<td>.82</td>
<td>.40</td>
<td>.16</td>
<td>.40</td>
<td>.16</td>
</tr>
<tr>
<td>Phonological awareness (n=12)</td>
<td>-.19</td>
<td>.56</td>
<td>.17</td>
<td>.59</td>
<td>.31</td>
<td>.32</td>
</tr>
<tr>
<td>Non-word repetition (n=13)</td>
<td>.06</td>
<td>.84</td>
<td>-.14</td>
<td>.65</td>
<td>-.01</td>
<td>.96</td>
</tr>
<tr>
<td>Articulation (n=13)</td>
<td>-.63</td>
<td>.02</td>
<td>.23</td>
<td>.46</td>
<td>.46</td>
<td>.11</td>
</tr>
</tbody>
</table>

Abbreviations: BEPTA, better ear pure tone average; SII, speech intelligibility index; SRT, speech reception threshold; PEACH, Parents’ Evaluation of Aural/Oral performance of Children.
Regression analyses were then conducted, adjusting for *a priori* potential confounders (child’s sex, languages spoken in the home, maternal education level, social disadvantage based on residential location, non-verbal IQ). This occurred with the acknowledgement that such analyses must be considered exploratory given the very small sample size.

Table 6.4 shows the attenuation of effect estimates resulting from these analyses. The association between speech articulation and unaided and aided measures of hearing was no longer observed. To explore this, potential confounders were included into the linear regression models in a step-wise manner. When social disadvantage level, child’s sex and non-verbal IQ were added to the PTA model as the third, fourth and fifth potential confounders, this resulted in the reduction in size of the articulation outcome regression coefficient and an increasing p-value. In the SII for loud speech model, the addition of any potential confounders immediately resulted in the same pattern as previously described. To examine whether any other variable impacted the association between measures of hearing (PTA and SII) and the articulation outcome, modelling using only two potential confounders was conducted (i.e. non-verbal IQ and one other from the list of *a priori* confounders above). From this, the only combination of potential confounders observed to further attenuate the association between hearing and articulation was non-verbal IQ and child’s sex.

Subsequent analyses were conducted examining any association between age at diagnosis and first fitting of hearing aids (both measured in months) and speech/language outcomes. Table 6.5 shows older age at diagnosis was significantly negatively correlated with phonological awareness and articulation scores. Once *a priori* potential confounders were included in linear regression analyses, the association between poorer articulation and later age at diagnosis attenuated fully. However, the association between phonological awareness and age at diagnosis remained significant in the model incorporating the 5 potential confounders (Table 6.6). Hearing aid use was also examined. No association between the hours of daily hearing aid use and speech/language outcomes was observed (Table 6.5 and Table 6.6)
Across all these results the effect sizes observed were generally small. Coupled with wide confidence intervals, this highlights a degree of uncertainty in the outcomes of these analyses and likely reflects the very small sample size. This echoes the caution expressed earlier regarding the exploratory nature of the above analyses.
Table 6.4. Results of linear regression analyses of speech/language outcomes by measures of unaided and aided hearing

<table>
<thead>
<tr>
<th>Language outcome</th>
<th>BEPTA (4FA dB HL)</th>
<th>SII Soft (55 dB SPL)</th>
<th>SII Medium (65 dB SPL)</th>
<th>SII Loud (75 dB SPL)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>β</td>
<td>95% CI</td>
<td>P</td>
<td>β</td>
</tr>
<tr>
<td>Receptive language (n=12)</td>
<td>.39 (-.39, 1.16)</td>
<td>.26 (-.06, -.67, .54)</td>
<td>.80 (.16, -.72, .40)</td>
<td>.50 (.17, -.67, .34)</td>
</tr>
<tr>
<td>Expressive language (n=12)</td>
<td>.15 (-.87, 1.17)</td>
<td>.72 (.27, -.91, .36)</td>
<td>.32 (-.08, -.76, .60)</td>
<td>.78 (.01, -.62, .64)</td>
</tr>
<tr>
<td>Receptive vocabulary (n=13)</td>
<td>.73 (-.51, 1.96)</td>
<td>.20 (.16, -.74, 1.05)</td>
<td>.68 (.05, -.86, .95)</td>
<td>.91 (-.08, -.95, .79)</td>
</tr>
<tr>
<td>Phonological awareness (n=12)</td>
<td>-.05 (-.86, .77)</td>
<td>.89 (.04, -.59, .52)</td>
<td>.87 (.05, -.48, .59)</td>
<td>.81 (.03, -.47, .52)</td>
</tr>
<tr>
<td>Non-word repetition (n=12)</td>
<td>-.52 (-2.57, 1.53)</td>
<td>.55 (.13, -.12, 1.04)</td>
<td>.79 (.07, -.11, 1.24)</td>
<td>.88 (.03, -.11, 1.10)</td>
</tr>
<tr>
<td>Articulation (n=13)</td>
<td>-.28 (-.85, .29)</td>
<td>.27 (.03, -.43, .37)</td>
<td>.86 (.03, -.37, .43)</td>
<td>.87 (.08, -.30, .46)</td>
</tr>
</tbody>
</table>

All regression coefficients reflect the mean difference in speech/language ability score according to each point increase in the measure of unaided or aided hearing (adjusted for maternal education, languages spoken in the home, social disadvantage, child’s sex, non-verbal IQ).
Table 6.5. Correlation coefficients for language outcomes by age at diagnosis, age of first hearing aid fitting and hearing aid use

<table>
<thead>
<tr>
<th>Language outcome</th>
<th>Age at diagnosis (months)</th>
<th>Age at first aid fitting (months)</th>
<th>Hearing aid use, weekday (hours)</th>
<th>Hearing aid use, weekend day (hours)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>r</td>
<td>p</td>
<td>r</td>
<td>p</td>
</tr>
<tr>
<td>Receptive language (n=13)</td>
<td>-.08</td>
<td>.80</td>
<td>-.36</td>
<td>.23</td>
</tr>
<tr>
<td>Expressive language (n=13)</td>
<td>-.26</td>
<td>.39</td>
<td>-.28</td>
<td>.35</td>
</tr>
<tr>
<td>Receptive vocabulary (n=14)</td>
<td>-.43</td>
<td>.13</td>
<td>-.18</td>
<td>.53</td>
</tr>
<tr>
<td>Phonological awareness (n=12)</td>
<td>-.59</td>
<td>.04</td>
<td>-.47</td>
<td>.12</td>
</tr>
<tr>
<td>Non-word repetition (n=13)</td>
<td>-.39</td>
<td>.18</td>
<td>-.04</td>
<td>.89</td>
</tr>
<tr>
<td>Articulation (n=13)</td>
<td>-.55</td>
<td>.05</td>
<td>-.06</td>
<td>.85</td>
</tr>
</tbody>
</table>
Table 6.6. Results of regression analyses of speech/language outcomes by age at diagnosis, age at first hearing aid fitting and hearing aid use

<table>
<thead>
<tr>
<th>Language outcome</th>
<th>Age at diagnosis (months)</th>
<th>Age at first aid fitting (months)</th>
<th>Hearing aid use, weekday (hours)</th>
<th>Hearing aid use, weekend day (hours)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>β  95% CI  P</td>
<td>β  95% CI  P</td>
<td>β  95% CI  P</td>
<td>β  95% CI  P</td>
</tr>
<tr>
<td>Receptive language (n=12)</td>
<td>.42 (-.58, 1.43) .33</td>
<td>-.17 (-.65, .32) .42</td>
<td>1.16 (-3.82, 6.14) .58</td>
<td>.11 (-2.18,2.40) .91</td>
</tr>
<tr>
<td>Expressive language (n=12)</td>
<td>-.08 (-1.38, 1.21) .88</td>
<td>-.19 (-.75, .37) .43</td>
<td>-.09 (-6.08, 5.89) .97</td>
<td>.78 (-1.73,3.29) .46</td>
</tr>
<tr>
<td>Receptive vocabulary (n=13)</td>
<td>-.50 (-2.22, 1.23) .51</td>
<td>-.18 (-.96, .60) .59</td>
<td>-1.33 (-8.19,5.52) .65</td>
<td>-.78 (-4.09,2.53) .59</td>
</tr>
<tr>
<td>Phonological awareness (n=12)</td>
<td>-.75 (-1.30, -.19) .02</td>
<td>-.13 (-.58, .32) .48</td>
<td>-1.82 (-6.03,2.39) .32</td>
<td>-.57 (-2.55,1.42) .50</td>
</tr>
<tr>
<td>Non-word repetition (n=12)</td>
<td>-1.13 (-2.99,.73) .18</td>
<td>-.05 (-1.06, .97) .91</td>
<td>-3.91 (-11.50,3.68) .24</td>
<td>-1.11 (-5.29,3.07) .52</td>
</tr>
<tr>
<td>Articulation (n=13)</td>
<td>-.43 (-1.10,.24) .17</td>
<td>-.11 (-.44,.23) .47</td>
<td>-1.05 (-3.96,1.85) .41</td>
<td>-.13 (-1.63,1.37) .84</td>
</tr>
</tbody>
</table>

All regression coefficients reflect the mean difference in speech/language ability score according to each month increase in the age at diagnosis or first hearing aid fitting (adjusted for maternal education, languages spoken in the home, social disadvantage, child’s sex, non-verbal IQ).
6.5 Variation between participants

Throughout this results chapter, there was considerable variation observed across the sample in individually measured audibility values and speech/language outcome scores for many participants. Table 6.7 displays, at a participant level, the better ear PTA, SII and speech/language outcome scores measured. Also displayed are parent-reported hours of daily hearing aid use and measured non-verbal IQ scores. Several patterns are seen in this participant-level data that highlight the challenges likely faced by clinicians working with this population.

Around one third of participants (participant 7, 9, 10, 11, 14) followed the group trends observed, with individual speech and language outcome scores generally lower than measured non-verbal IQ. Whilst this may reflect what is generally observed in Australian children, showing lower language and higher IQ (e.g. Levickis et al. 2017\textsuperscript{241}), other contributing factors are worth highlighting. Four out of these five participants had better ear PTA scores that indicated a moderate hearing loss and all were reported to wear amplification for at least 8 hours per day on weekdays. Two participants lived in households where English is not the main language used (participant 7 and 9). Reviewing SII scores showed that participant 9, who had the highest better ear PTA (worst hearing), also had the lowest measured SII scores. These scores reflect poor aided audibility even for loud speech (75 dB SPL) at the time of assessment. This participant appears to have had the largest consistent discrepancy between non-verbal IQ and individual speech and language outcomes of all the participants in the group.

Closer examination of hearing and demographic factors for participant 9 indicates that this child was identified and fitted early (diagnosed at 1.6 months, fitted at 3 months). The child’s parents had completed tertiary education and the family had a measure of social disadvantage similar to the population norm (disadvantage index 1020 versus a population mean of 1000, where higher scores indicate less neighbourhood disadvantage). The juxtaposition between factors that could contribute to the large discrepancy between participant 9’s poor language vs high IQ scores (i.e. moderate hearing loss, poor aided audibility,
English as a second language household) and those factors that are intuitively held to be beneficial to child outcomes (i.e. early diagnosis and aiding, well-educated parents, high reported daily hearing aid use) illustrate the challenges faced by clinicians at an individual level. Variability in outcomes and inconsistencies in measures pose not unsubstantial difficulties for treating clinicians.

6.6 Summary

A small sample size dictates that all regression analyses undertaken are essentially exploratory and must be interpreted as such. The variable and unpredictable effect estimates and large confidence intervals are likely an illustration of this fact. However, a robust result in this chapter was the significant association between measures of hearing (i.e. unaided thresholds/PTA and aided audibility/SII) and speech recognition. Such results fit with the pattern of better speech articulation ability in those participants with better unaided or aided measures of hearing. The influence of a priori potential confounders could not reliably be estimated. Individual variation between participant scores highlights the challenges encountered by treating clinicians in providing appropriate management and assisting families with obtaining the best outcomes for the child with mild or moderate hearing loss.
<table>
<thead>
<tr>
<th>Participant</th>
<th>BEPTA (dB HL)</th>
<th>Hearing device use</th>
<th>SII</th>
<th>Language outcomes</th>
<th>Non-verbal IQ</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Weekday</td>
<td>Weekend</td>
</tr>
<tr>
<td>1</td>
<td>31</td>
<td>8</td>
<td>0</td>
<td>50</td>
<td>69</td>
</tr>
<tr>
<td>2</td>
<td>55</td>
<td>12</td>
<td>12</td>
<td>11</td>
<td>41</td>
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<tr>
<td>3</td>
<td>58</td>
<td>13</td>
<td>13</td>
<td>14</td>
<td>26</td>
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<tr>
<td>4</td>
<td>43</td>
<td>10</td>
<td>10</td>
<td>23</td>
<td>49</td>
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<tr>
<td>5</td>
<td>31</td>
<td>8</td>
<td>7</td>
<td>37</td>
<td>63</td>
</tr>
<tr>
<td>6</td>
<td>35</td>
<td>6</td>
<td>0</td>
<td>50</td>
<td>70</td>
</tr>
<tr>
<td>7</td>
<td>48</td>
<td>10</td>
<td>7</td>
<td>37</td>
<td>49</td>
</tr>
<tr>
<td>8</td>
<td>51</td>
<td>13</td>
<td>13</td>
<td>42</td>
<td>63</td>
</tr>
<tr>
<td>9</td>
<td>65</td>
<td>8</td>
<td>6</td>
<td>6</td>
<td>15</td>
</tr>
<tr>
<td>10</td>
<td>33</td>
<td>9</td>
<td>4</td>
<td>35</td>
<td>63</td>
</tr>
<tr>
<td>11</td>
<td>41</td>
<td>12</td>
<td>12</td>
<td>16</td>
<td>43</td>
</tr>
<tr>
<td>12</td>
<td>35</td>
<td>11</td>
<td>8</td>
<td>31</td>
<td>62</td>
</tr>
<tr>
<td>13</td>
<td>35</td>
<td>12</td>
<td>12</td>
<td>14</td>
<td>39</td>
</tr>
<tr>
<td>14</td>
<td>46</td>
<td>13</td>
<td>14</td>
<td>29</td>
<td>48</td>
</tr>
</tbody>
</table>

Abbreviations: BEPTA, better ear PTA; artic, articulation.
7. Expressive vocabulary of children aged 1-3 years with milder hearing loss (Aim 3)

7.1 Overview
To address Aim 3, the researcher quantified expressive vocabulary at 1-3 years of age via parent report for children born with mild or moderate bilateral hearing loss. These children were compared to a representative sample of similarly-aged children without hearing loss from the same geographic state of Australia. The findings for this aim are presented in a traditional chapter format. It begins by defining the characteristics of the analysed sample, and then explains the statistical approach (which was influenced by age differences between the samples). It reports and describes expressive vocabulary for children both with and without hearing loss and compares the abilities of the two groups. Finally, evidence for the impact of audibility on expressive vocabulary of the children with hearing loss is outlined. Findings are critically discussed in Chapter 8.

7.2 Sample characteristics
As outlined above, two distinct samples of children were included in analyses. The scope of this PhD included approaching and actively collecting data from all eligible children with hearing loss within the overarching VicCHILD study (see Section 4.8.2 for procedures). Data for the sample of children without hearing loss were obtained from a completed wave of assessment from the longitudinal Early Language in Victoria Study (ELVS). Sections 4.3 and 4.4.3 detail the methods for the two source studies and waves.

7.2.1 Children with hearing loss
Approaches were made to all families recorded in the VicCHILD databank as having a child aged between 16 and 36 months of age born between August 2012 and March 2014 with a bilateral mild or moderate sensorineural hearing loss. Twenty-two of the 34 families approached responded (64% uptake rate). Two
families were excluded prior to analysis due to corrected diagnoses of profound bilateral hearing loss at birth. Therefore, 20 children were included in analyses.

7.2.2 Children without hearing loss
Of the 1917 children originally consented into ELVS, at the time of 2 year old data collection there were outcome data available for 1711 children, all of which were included in analyses. From baseline at 8 months of age to the 2 year old data collection wave, the ELVS study had achieved a 91.1% retention rate.

7.2.3 Demographic factors
The samples were comparable on participant sex breakdown, levels of family disadvantage and parent tertiary education levels (Table 7.1). However, the samples differed on two demographic characteristics: child age at assessment and the primary language other than English being spoken at home. Children with hearing loss were on average older (mean age 29.5 versus 24.1 months, p=.0002) but assessed across a wider age range than children without hearing loss (20 to 38 months versus 23 to 28 months). This wider assessment age range was necessary to maximise the number of included children with hearing loss available from the VicCHILD databank. Thirty percent of families with a child with hearing loss reported using a primary language other than English at home, compared to less than 4% in the sample of children without hearing loss (p<.0001). In part, this difference may reflect the growth in home language diversity over the approximate 10-year birth gap between the two groups of children. Data from the 2016 Australian Census indicates 26% of families in Victoria now speak a language other than English at home. The sample of children without hearing loss may have also been drawn from local government areas (see Section 4.4.3 for recruitment details) that had lower diversity in home languages, compared to the VicCHILD sample which has participants residing across the entire geographic state of Victoria.
Table 7.1. Aim 3 participant demographic characteristics

<table>
<thead>
<tr>
<th>Characteristic</th>
<th>VicCHILD (born VIC 2007-10) (n=20)</th>
<th>ELVS (born VIC 2003) (n=1711)</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Child</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Age at assessment in months, mean (SD); range</td>
<td>29.3 (5.0); 20 to 38</td>
<td>24.1 (0.4); 23 to 28</td>
</tr>
<tr>
<td>Male gender, n (%)</td>
<td>12 (60)</td>
<td>868 (50)</td>
</tr>
<tr>
<td><strong>Family</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Primary language other than English household, n (%)</td>
<td>6 (30.0)</td>
<td>61 (3.7)</td>
</tr>
<tr>
<td>Disadvantage Index, mean (SD)</td>
<td>1021.0 (45.5)</td>
<td>1038.1 (59.4)</td>
</tr>
<tr>
<td>Parent with tertiary level education, n (%)</td>
<td>8 (40.0)</td>
<td>641 (37.5)</td>
</tr>
</tbody>
</table>

7.2.3.1 Characteristics unique to the sample with hearing loss

Of the 20 children with hearing loss, 17 had their hearing loss degree at diagnosis (i.e. mild or moderate, supplied to VicCHILD from VIHSP) confirmed by their initial behavioural audiograms conducted at Australian Hearing. Additional behavioural hearing data from hearing testing around the time of language assessment were available for 16 children, providing a current severity of hearing loss (see Table 7.2); without exception, hearing losses for all children remained within the mild or moderate range. The researcher sighted all available behavioural audiograms. For those children (n=4) whose behavioural hearing data were not available, parents were called and verbally confirmed that both their child’s initial and current hearing loss met inclusion criteria. Based on reported hearing at diagnosis, the sample was evenly divided into 10 children with mild and 10 with moderate loss.

The majority of participating children were diagnosed early in life. Nineteen of the 20 children were diagnosed at less than 6 months of age, 18 at less than 3 months. One child was identified at age 15 months, skewing the diagnosis age range. With the exclusion of this child, the mean age at diagnosis reduced from 2.3 months (SD 3.3 months) to 1.6 months (SD 1.0 month). Despite the later diagnosis, this one child was retained in the sample for analysis purposes.
The majority of children were fitted bilaterally with amplification, albeit with a wide range of reported hearing aid use. Eighteen children were current hearing aid users, with a mean age of first fitting of 7.9 months (SD 6.6 months). Parents completed an hourly breakdown of hearing aid use across typical daytime waking hours (6am through to 8pm) for both weekday and weekend days at around the time of survey completion. Table 7.2 shows children on average wore their hearing aids for between 6 and 7 hours per day, but this ranged from as little as 0-2 hours to as much as 12 hours. Parents reported that 16 of the 20 children (80%) were receiving some form of early education intervention at the time of expressive vocabulary assessment.
Table 7.2. Hearing-related characteristics of participants aged 1-3 years

<table>
<thead>
<tr>
<th>Characteristic</th>
<th>n</th>
<th>Mean (SD); range</th>
</tr>
</thead>
<tbody>
<tr>
<td>Initial severity of hearing loss, 3FAHL (dB HL)</td>
<td>17</td>
<td>41.3 (9.0); 20 to 56</td>
</tr>
<tr>
<td>Current severity of hearing loss, 3FAHL (dB HL)</td>
<td>16</td>
<td>37.9 (9.2); 20 to 55</td>
</tr>
<tr>
<td>Age at diagnosis (months)</td>
<td>20</td>
<td>2.3 (3.3); 0.3 to 15.6</td>
</tr>
<tr>
<td>Age at fitting (months)</td>
<td>18</td>
<td>7.9 (6.6); 1 to 26</td>
</tr>
<tr>
<td>Weekday hours of hearing aid use</td>
<td>15</td>
<td>7.4 (3.1); 2 to 12</td>
</tr>
<tr>
<td>Weekend day hours of hearing aid use</td>
<td>16</td>
<td>6.5 (3.7); 0 to 12</td>
</tr>
</tbody>
</table>

Abbreviations: 3FAHL, three frequency average hearing loss; dB HL, decibels hearing level

7.3 Impact of age at assessment on analyses

As mentioned in Section 7.2.3, there were differences in age structure at assessment between the two samples (with children without hearing loss on average 5 months younger and with a much narrower spread of ages). To determine whether this difference would impact analyses, preliminary investigations were conducted on the effect of child age at the time of expressive vocabulary assessment.

Differences in the pattern of vocabulary production were observed between the two samples. Figure 7.1 shows scatterplots of raw expressive vocabulary scores by age at assessment for children with and without hearing loss, with the accompanying linear line of best fit trend lines. These show that, for children with mild hearing loss, there was a general tendency toward higher expressive vocabulary in older children. This tendency was less pronounced in children with moderate loss, with the trend line appearing close to horizontal. With mild and moderate losses combined, a slight visual tendency is observed toward greater expressive vocabulary scores with increasing age.

A different pattern was observed in the sample with no hearing loss. The scatterplot shows a concentration of scores around the 24-month age point.
where data collection was targeted. The trend line suggests that children in this sample who were older at the time of assessment had lower expressive vocabularies than their younger peers. This does not reflect what we expect to see in a natural population trend in language by age. Instead, this pattern may reflect the data collection process in the ELVS study and what is known about parental agency. Motivated families with more resources, who may have children with greater inherent language ability, may have completed the assessment more promptly than families who completed the assessment at a later child age.

These differences in patterns of expressive vocabulary production between the children with and without hearing loss imposed restrictions on analysis options. The approach originally outlined to conduct linear regression and adjust for age at assessment and other potentially confounding factors was no longer appropriate. To adjust for age would essentially mean the adjustment is being driven by a non-representative pattern, observed in the small age range of children without hearing loss, being extrapolated linearly across the entire range of assessment ages. For this reason, analysis and comparison of results between children with and without hearing loss was conducted descriptively, in reference to published sex-specific fitted percentile scores for children aged 16-30 months and by “vocabulary age” (see Section 7.4). Each set of analyses is outlined below.
Figure 7.1. Scatterplot of expressive vocabulary raw score by age at assessment
7.4 Expressive vocabulary of children with hearing loss

Expressive vocabulary was assessed using parent responses from the 680-word vocabulary checklist from the MacArthur Bates Communicative Development Inventory (MCDI) Words and Sentences questionnaire.\textsuperscript{235} For the 20 children with hearing loss, a mean of 223 words (SD 166 words) was produced. Raw scores ranged from 0 to 462 words. By degree of hearing loss, the following raw scores were obtained: mild, 171 words (SD 167 words); moderate, 275 words (SD 157 words).

To understand how such scores placed these children with hearing loss within the broader population, percentile ranks from published norms were assigned for individual participants according to their age at assessment.\textsuperscript{235} Details of the United States norming sample are found in Section 4.10.2 of Chapter 4. If a child’s raw score lay between two percentile ranks, the conservative approach described by Fenson et al (2006)\textsuperscript{235} was used, where the lower of the two percentiles was assigned for the child’s score. However, when children scored below the 5\textsuperscript{th} percentile, they were arbitrarily assigned the 5\textsuperscript{th} percentile rank. For children aged over 30 months, percentile ranks were assigned from the highest available (i.e. 30 month) normed age group available. This occurred for 8 of the 20 children (40\% of the sample), with a maximum age of 38 months. Four of these 8 children had expressive vocabulary falling within the 5\textsuperscript{th} percentile for children aged 30 months, despite their chronological age ranging from 33 to 35 months. The other half of children aged over 30 months at assessment were ranked in the 10\textsuperscript{th}, 20\textsuperscript{th} (two children) and 25\textsuperscript{th} percentile for expected ability at 30 months, despite chronological age at assessment ranging from 33 to 38 months.

As 40\% of the children with hearing loss were older than the standardised percentile ranks for the outcome measure used, age equivalents based on language performance were estimated for each child. These equivalents were determined by relating individual raw scores to the 50\textsuperscript{th} percentile scores within the published tables of expected performance across age bands 16 to 30 months. The band within which the child’s raw score crossed the 50\textsuperscript{th} percentile was
assigned as the “vocabulary age”, distinct from the child’s “chronological age”. Using these two ages, a expressive vocabulary differential was calculated for each child by subtracting the chronological age from vocabulary age. These multiple measures of expressive vocabulary are listed in Table 7.3 per child with hearing loss. Sixteen children’s vocabulary age was below their chronological age (range 5 to 17 months) while only 3 showed a vocabulary age ahead of their chronological age and this was across a much narrower range (1 to 3 months).
<table>
<thead>
<tr>
<th>Participant number</th>
<th>Age at assessment (months)</th>
<th>Expressive vocabulary raw score</th>
<th>Percentile rank</th>
<th>Vocabulary age (months)</th>
<th>Vocabulary differential (months)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mild at diagnosis</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Participant 1</td>
<td>33</td>
<td>113</td>
<td>5*</td>
<td>20</td>
<td>-13</td>
</tr>
<tr>
<td>Participant 2</td>
<td>35</td>
<td>85</td>
<td>5*</td>
<td>19</td>
<td>-16</td>
</tr>
<tr>
<td>Participant 3</td>
<td>35</td>
<td>235</td>
<td>15*</td>
<td>23</td>
<td>-12</td>
</tr>
<tr>
<td>Participant 4</td>
<td>30</td>
<td>434</td>
<td>20</td>
<td>25</td>
<td>-5</td>
</tr>
<tr>
<td>Participant 5</td>
<td>25</td>
<td>120</td>
<td>15</td>
<td>20</td>
<td>-5</td>
</tr>
<tr>
<td>Participant 6</td>
<td>26</td>
<td>0</td>
<td>5</td>
<td>&lt;16</td>
<td>-10</td>
</tr>
<tr>
<td>Participant 7</td>
<td>22</td>
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<td>5</td>
<td>&lt;16</td>
<td>-6</td>
</tr>
<tr>
<td>Participant 8</td>
<td>30</td>
<td>236</td>
<td>10</td>
<td>23</td>
<td>-7</td>
</tr>
<tr>
<td>Participant 9</td>
<td>33</td>
<td>33</td>
<td>5*</td>
<td>&lt;16</td>
<td>-17</td>
</tr>
<tr>
<td>Participant 10</td>
<td>27</td>
<td>462</td>
<td>60</td>
<td>28</td>
<td>+1</td>
</tr>
<tr>
<td>Moderate at diagnosis</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Participant 11</td>
<td>38</td>
<td>398</td>
<td>30*</td>
<td>27</td>
<td>-11</td>
</tr>
<tr>
<td>Participant 12</td>
<td>33</td>
<td>81</td>
<td>5*</td>
<td>18</td>
<td>-15</td>
</tr>
<tr>
<td>Participant 13</td>
<td>30</td>
<td>44</td>
<td>5</td>
<td>16</td>
<td>-14</td>
</tr>
<tr>
<td>Participant 14</td>
<td>35</td>
<td>328</td>
<td>25*</td>
<td>25</td>
<td>-10</td>
</tr>
<tr>
<td>Participant 15</td>
<td>26</td>
<td>66</td>
<td>5</td>
<td>16</td>
<td>-10</td>
</tr>
<tr>
<td>Participant 16</td>
<td>24</td>
<td>410</td>
<td>75</td>
<td>27</td>
<td>+3</td>
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<tr>
<td>Participant 17</td>
<td>35</td>
<td>238</td>
<td>25</td>
<td>21</td>
<td>-14</td>
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<tr>
<td>Participant 18</td>
<td>26</td>
<td>444</td>
<td>50</td>
<td>26</td>
<td>0</td>
</tr>
<tr>
<td>Participant 19</td>
<td>33</td>
<td>409</td>
<td>25*</td>
<td>25</td>
<td>-8</td>
</tr>
<tr>
<td>Participant 20</td>
<td>20</td>
<td>335</td>
<td>80</td>
<td>23</td>
<td>+3</td>
</tr>
</tbody>
</table>

*percentile rank assigned based on sex specific norms for children at age 30 months.
7.4.1 Comparison of expressive vocabulary between children with and without hearing loss

Raw expressive vocabulary scores of children without hearing loss were on average higher than children with hearing loss. Of the 1711 children without hearing loss, a mean of 260 words (SD 162 words) was produced. Raw scores ranged from 0 to 679 words. Expressive vocabulary percentile ranks are plotted in Figure 7.2. Common to both the VicCHILD (filled bars) and ELVS (open bars) plots is the high proportion of children falling in the 5th percentile for expected vocabulary production. Forty percent of children with hearing loss, and 12% of children without hearing loss achieved at or below the 5th percentile published for the US norming sample.235 This highlights that there was a tendency within both samples for expressive vocabulary to be lower than expected according to age. When defined as achieving a score at or below the 10th percentile, close to 20% of children without hearing loss would be classified as having delayed expressive vocabulary at two years. Children with hearing loss were around 2.5 times more likely to have delayed expressive vocabulary than the representative sample of children without hearing loss. This figure could be even higher given the limitations of assigning percentile ranks to children chronologically over 30 months of age. Visual inspection of Figure 7.2 highlights the clustering of scores within the hearing loss sample below the expected mean performance. Fewer than 20% of children with hearing loss scored higher than the 50th percentile.
Vocabulary age and vocabulary differential scores were also calculated for the sample of children without hearing loss. When the mean chronological age of 24.1 months was subtracted from the mean vocabulary age of 22.3 months, children from the representative sample without hearing loss (ELVS) had a vocabulary differential of -1.8 months (SD 4.0 months). Table 7.4 displays the differential for the sample of children with hearing loss, against the differential for children without hearing loss as the reference. To highlight any impact on vocabulary differential calculations, the hearing loss sample was considered both with and without the 2 children with no reported words produced. The hearing loss sample had a greater delay in expressive vocabulary for every method of comparison conducted: the full sample (n=20), the sample excluding the 2 children reported to have no expressive vocabulary, and by degree of hearing loss at diagnosis. Children with mild loss had scores that were on average 9
months lower than expected (SD 5.6 months), whereas children with moderate loss had scores that were 6.6 months lower than expected (SD 6.7 months).

Table 7.4. Vocabulary differential in months, stratified by sample

<table>
<thead>
<tr>
<th>Characteristic</th>
<th>ELVS (n=1711)</th>
<th>VicCHILD (n=20)</th>
<th>VicCHILD mild (n=8)*</th>
<th>VicCHILD mild (n=10)</th>
<th>VicCHILD mod (n=10)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Vocabulary age at ax, mean (SD)</td>
<td>22.3 (4.0)</td>
<td>21.5 (4.2)</td>
<td>22.1 (4.0)</td>
<td>20.6 (4.1)</td>
<td>22.4 (4.4)</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Chronological age at ax, mean (SD)</td>
<td>24.1 (0.4)</td>
<td>29.3 (5.0)</td>
<td>29.9 (4.9)</td>
<td>29.6 (4.5)</td>
<td>29 (5.7)</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Differential</td>
<td>-1.8 (4.0)</td>
<td>-7.8 (6.1)</td>
<td>-7.8 (6.5)</td>
<td>-9 (5.6)</td>
<td>-6.6 (6.7)</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Abbreviations: ax, assessment.</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
*excluding those children with 0 MCDI scores

In the scatterplot of vocabulary differential by expressive vocabulary raw score with linear trend lines (Figure 7.3), children with hearing loss exhibit greater expressive vocabulary delay (i.e. poorer vocabulary differentials) than children without hearing loss. This performance pattern is observed despite the use of a vocabulary differential. This differential allows comparability across the hearing and hearing loss samples by accounting for differences in age at assessment.
A lower proportion of children with hearing loss recorded positive vocabulary differential scores than those children without hearing loss. This is visualised via the horizontal line in Figure 7.3 plotted at vocabulary differential point 0, representing the point where the child’s vocabulary age equals their chronological age. Therefore, all points above this line reflect children who have a positive differential score or a vocabulary age that is higher than their chronological age. Within the sample of children without hearing loss, 31.3% (n=535) had a positive vocabulary differential. This contrasted with the sample of children with hearing loss, of whom 20% (n=4) recorded positive vocabulary differentials. Three of these 4 children were diagnosed with a moderate bilateral loss, and one with mild bilateral hearing loss.

Figure 7.3. Scatterplot of vocabulary differential by expressive vocabulary raw score
7.4.2 The association between parent-reported child audibility and child expressive vocabulary

Parents of children with hearing loss completed the Parents’ Evaluation of Aural/oral performance of Children (PEACH) rating scale at the same time they reported their child’s expressive vocabulary. The PEACH is a functional auditory outcome measure, designed for use by parents or caregivers of a child with hearing loss, which probes how well the child appears to function in a variety of listening conditions (see details in Section 4.10.2). Table 7.5 summarises the PEACH responses obtained from 19 of the 20 families with a child with hearing loss (95% completion rate).

No significant differences were observed in mean percentage performance scores for questions categorised into “quiet”, “noisy” or “overall” (quiet and noisy combined) conditions, whether looking at the children with hearing loss as one group, or divided by degree. Thus, the PEACH overall scores were used to describe child listening performance, with a mean score for children with mild and moderate loss combined of 66%. A higher percentage score indicates less listening difficulty as perceived by the caregiver. As outlined in Section 4.10.2, a score of 90% by 3 years of age is considered a reasonable goal for expected performance. Given the mean age at assessment of children with hearing loss was 29.3 months, on average it would appear this group of children are perceived by parents as experiencing poorer listening performance than expected. However, a considerable range in PEACH scores was noted, spanning approximately 35% to 80-95% when considering both degrees of loss. This wide range of scores is consistent with the broad heterogeneity in outcomes achieved by children with mild and moderate loss across all studies in this PhD.
Table 7.5. PEACH performance values across participants with hearing loss

<table>
<thead>
<tr>
<th>Characteristic</th>
<th>n</th>
<th>Mean (SD); range</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Mild moderate combined</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Subscale performance</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Quiet condition percentage</td>
<td>19</td>
<td>68.2 (17.7); 33.3 to 95.8</td>
</tr>
<tr>
<td>Noisy condition percentage</td>
<td>19</td>
<td>63.4 (14.3); 40 to 95</td>
</tr>
<tr>
<td><strong>Overall performance</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Combined conditions percentage</td>
<td>19</td>
<td>66.0 (15.2); 36.3 to 95.5</td>
</tr>
<tr>
<td><strong>Mild</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Subscale performance</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Quiet condition percentage</td>
<td>9</td>
<td>68.5 (20.0); 33.3 to 91.7</td>
</tr>
<tr>
<td>Noisy condition percentage</td>
<td>9</td>
<td>62.2 (12.5); 40 to 80</td>
</tr>
<tr>
<td><strong>Overall performance</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Combined conditions percentage</td>
<td>9</td>
<td>65.7 (15.6); 36.4 to 81.8</td>
</tr>
<tr>
<td><strong>Moderate</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td><strong>Subscale performance</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Quiet condition percentage</td>
<td>10</td>
<td>67.9 (16.6); 41.6 to 95.8</td>
</tr>
<tr>
<td>Noisy condition percentage</td>
<td>10</td>
<td>64.5 (16.4); 40 to 95</td>
</tr>
<tr>
<td><strong>Overall performance</strong></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Combined conditions percentage</td>
<td>109</td>
<td>66.4 (15.7); 40.9 to 95.5</td>
</tr>
</tbody>
</table>

Parents of children with hearing loss who reported their child as having a higher expressive vocabulary also reported fewer observations of their child having listening difficulties in commonly encountered environments and scenarios. PEACH scores showed a moderate positive correlation with expressive vocabulary raw scores (Figure 7.4; r = .5, p<.05). Children with mild and moderate losses at diagnosis are combined in this scatterplot. Dichotomisation by degree of loss did not exhibit any tendency towards performance differences between mild and moderate losses.
Figure 7.4. Correlation between PEACH score and expressive vocabulary raw score for children with hearing loss

Whilst not reaching statistical significance, parents of children who were further behind in their expressive vocabulary (represented by a more negative language differential) tended to report more hours of hearing aid use each day. Against expectations and perhaps reflecting reverse causation, this weak association was observed for both children with mild ($r = -.4$, $p = .2$) and moderate ($r = -.4$, $p = .2$) hearing loss at diagnosis, as displayed in Figure 7.5. A similar association was observed for mild and moderate losses combined ($r = -.3$, $p = .2$).
7.5 Summary

Despite differences in the age distribution of the two samples of children, analysis of expressive vocabulary performance would suggest that children with mild and moderate hearing loss are performing more poorly than children without hearing loss at around the age of 2 years. In reference to published norms, this poorer performance by children with hearing loss was observed when considering both raw scores and derived scores that represented a vocabulary age and a differential score for expected language performance. In this small sample, children with hearing loss were 2.5 times more likely to have delayed expressive language than children without hearing loss, with a delay defined as scoring at or below the 10th percentile for expected performance. Only one fifth of children with hearing loss were meeting or exceeding expectations of expressive vocabulary performance, compared to around one third of children without hearing loss. Whilst acknowledging a wide distribution of scores, parent-reports of listening performance may suggest these children with hearing loss
are functioning at a lower than expected level in commonly encountered listening conditions. This is reported alongside wide estimates of daily hearing device use, with the suggestion of greater use by children who have poorer expressive vocabulary.
8. Discussion

8.1 Overview
This final chapter critically discusses the principal findings of this PhD. The findings for Aim 1 are expanded from the brief discussion in the publication comprising Chapter 5. Aim 2 and 3 findings are discussed for the first time. A summary of overarching strengths and weaknesses of the study follows. Next, each aim is interpreted in context with prior literature. To conclude, clinical implications of this study and future research directions are highlighted.

8.2 Principal findings

8.2.1 Aim 1
For children with mild or moderate congenital hearing loss aged 5-8 years of age drawn from four population-derived samples, similar except for the early detection system in place when they were born (opportunistic, risk factor detection, newly-implemented universal newborn hearing screening (UNHS), well-established UNHS), to:
(a) compare language, behaviour and quality of life outcomes between the samples,
(b) determine whether age of detection predicts outcomes within the four samples pooled, and
(c) compare mean outcomes of those exposed to well-established UNHS to a population-based sample of similarly-aged children without hearing loss.

Age of detection and hearing aid fitting decreased significantly with evolution over time to the systems and processes for the detection of congenital hearing loss. This trend was seen both for children born with mild and moderate bilateral hearing loss (p <.01); notably children with mild loss detected under the contemporary mature UNHS system were diagnosed on average 9 months (95% CI -22.8 to 4.8) and fitted 19 months (95% CI -33.2 to -4.8) earlier than under the immediately preceding newly established UNHS system.
For moderate losses, clear benefits were associated with the shift from opportunistic to UNHS systems of detection for expressive language (p for trend .05) and receptive vocabulary (p for trend .06). Receptive language improvement was modest and not statistically significant. Little evidence was observed of similar trends for mild losses, with mean expressive language and receptive vocabulary remaining about one standard deviation below normative means across all four periods. Health-related quality of life and behaviour problems appeared mostly unaffected by system of hearing loss detection regardless of severity.

Visual trends for the pooled mild-moderate sample suggested better language and vocabulary outcomes resulted when age of detection was prior to 6 or after 30 months. Trends showing poorer outcomes with detection between 6-30 months of age were common to each detection system analysed. Regression analyses illustrated these benefits of earlier detection were almost entirely experienced by children with moderate, not mild, losses. Compared to a diagnosis under 6 months, children with moderate loss diagnosed between 18-30 months had poorer receptive and expressive language by between one and two standard deviations (fully adjusted mean differences -16.7 points, 95% CI -30.3 to -3.0 and -27.0, 95% CI -40.2 to -13.7, respectively) and over a standard deviation poorer receptive vocabulary skills (-16.4, 95% CI -28.1 to -4.7). Outcomes for children with mild losses showed little association with age at detection.

Expressive language was around 0.6 standard deviations lower in the mild-moderate combined mature UNHS group when compared to the children without hearing loss group (fully adjusted mean difference -8.9 points, 95% CI -14.7 to -3.1). Adjusted receptive language, behaviour and health-related quality of life scores were similar to those of children without hearing loss.

8.2.2 Aim 2

For children aged 5-7 years fitted with hearing aids for mild or moderate congenital hearing loss, who were identified under the well-established UNHS
system of detection, to quantify relationships of (i) unaided hearing acuity and (ii) aided audibility with: (a) measured speech recognition ability, and (b) speech/language outcomes.

Aided audibility, represented by the speech intelligibility index (SII), significantly correlated with unaided hearing ability (pure tone average, PTA) at soft \( r = -0.60, p = 0.02 \), medium \( r = -0.78, p = 0.001 \) and loud \( r = -0.79, p < 0.001 \) speech inputs. Unaided hearing and speech recognition ability were significantly correlated; poorer speech recognition was observed for children with poorer sound detection thresholds \( r = -0.65, p = 0.02 \). SII scores and speech recognition scores significantly correlated at all three inputs (soft speech \( r = -0.63, p = 0.04 \); medium speech \( r = -0.73, p = 0.01 \); loud speech \( r = -0.70, p = 0.02 \)). These findings support the SII calculation used as being consistent with both unaided measures of hearing ability and amplified sentence recognition scores in competing speech for a sample of children fitted with hearing aids after early identification of mild or moderate bilateral hearing loss.

It appeared that better (i.e. lower) PTA and better (i.e. higher) SII scores for loud speech inputs translated into better speech articulation, a skill that generally requires good auditory perception of phonemes (PTA \( r = -0.63, p = 0.02 \); SII \( r = 0.57, p = 0.04 \)). However, and contrary to what was hypothesised, hearing measures (unaided acuity and aided audibility) were not significantly associated with receptive language, expressive language, receptive vocabulary, phonological awareness or non-word repetition scores. Furthermore, exploratory linear regression modelling adjusted for \textit{a priori} potential confounders saw the loss of the significant association between speech articulation and both unaided and aided measures of hearing.

There was considerable variation observed across the sample for measured audibility values and speech/language outcome scores. Coupled with a low sample size, these factors impacted the ability to interpret the results obtained.
8.2.3 Aim 3

For 1-3 year old children with mild or moderate congenital hearing loss detected by a system of well-established UNHS: (a) describe parent-reported expressive vocabulary, and (b) compare mean outcomes to a population-based sample of similarly-aged children without hearing loss.

In a sample of early diagnosed and amplified children with mild or moderate bilateral hearing loss of mean age 29 months, the mean expressive vocabulary raw score was 223 words (SD 166 words). Children with a mild bilateral loss had a lower average raw score (171 words, SD 167 words) than children with moderate loss (275 words, SD 157 words). This contrasts to a mean raw score of 260 words (SD 162 words) in a representative sample of children without hearing loss who were on average 5 months younger (mean age 24 months).

Age differences between the samples challenged meaningful direct comparison of raw vocabulary scores. However, proportionally more children with hearing loss were distributed in the lower percentile ranks for expected performance than were children without hearing loss.

By using the normed ages of where a child’s raw score crosses the 50th percentile for expected achievement (i.e. defining their “vocabulary” age), a vocabulary differential score highlighted variance between vocabulary and chronological ages. Against the published norms, the representative sample without hearing loss scored 1.8 months lower than expected for the size of their expressive vocabulary. Children with hearing loss displayed younger age equivalents; those with mild loss were on average 9 months lower than expected (SD 5.6 months), moderate loss on average 6.6 months lower (SD 6.7 months).

Slightly over 30% of the sample of children with no hearing loss had a positive vocabulary differential, i.e. they were performing at a level that met or exceeded the expressive vocabulary expectations for their age. Twenty percent of the children with hearing loss were in the same positive range: 3 children with moderate loss and one with mild loss.
For the sample of children with hearing loss, parents who reported their child showed better listening ability also reported larger expressive vocabularies \((r = .5, \ p < .05)\). Whilst not reaching significance, parents of children with lower expressive vocabulary scores tended to report more hearing aid use in their children than did parents of children with higher scores for both mild and moderate losses.

8.2.4 Synopsis of principal findings
For children born with mild hearing loss, there was limited evidence of benefit to language outcomes in the early primary school years as a result of earlier diagnosis and amplification. However, there were clear benefits seen for children born with moderate hearing loss. These results suggest that, despite the best intentions made possible by UNHS, our change in approach to treating children with mild loss has not produced consistent improvements across time.

Both unaided and aided measures of hearing closely agreed with speech recognition performance in a group of children with aided mild or moderate loss. Despite this association, these same measures of hearing did not show evidence of a relationship to children’s speech/language outcomes. Whilst limitations with the data exist, the possibility of non-auditory factors contributing to outcomes achieved by these children must be considered.

Delays in language development appear evident at around two years of age in children with mild and moderate hearing loss. Expressive vocabulary production lags the production reported in children without hearing loss. Of interest, this delayed vocabulary appears to be greatest in children with mild, rather than moderate loss. These observed delays were despite early detection and amplification use.

8.3 Strengths
The three studies that form this PhD have a number of common factors that support the validity of the findings. These strengths centre upon the quasi-
experimental design. Population-based studies were the source of all participants across both age groups. Nesting the PhD within an existing research project, with protocols designed to optimise participation by all eligible children in the geographic state of Victoria, strengthened the ecological validity of the results presented. Children were recruited from the statewide VicCHILD database that enrolls based on detection of permanent hearing loss, rather than any specific treatment. Population-based comparison groups, both in the children without hearing loss\textsuperscript{119,245} and with hearing loss,\textsuperscript{107,156,163} provide a robust platform from which to measure secular trends (Aim 1) and performance compared to children representing those without hearing loss (Aims 1 and 3).

Broad participant inclusion criteria across all aims of this PhD study should further enhance generalisability of the findings. Children with identified intellectual disability were only excluded for analyses that required comparison with historical cohorts (aim 1). Unlike much existing research into the outcomes of children with hearing loss, no other exclusions for additional disabilities were applied. This likely strengthened the representative nature of the sample, given that reports of additional disabilities in children with congenital hearing loss range from 20-40\%.\textsuperscript{61,246} The good study uptake rate (67\% for Aim 1, 64\% for Aim 3) suggests the samples obtained had sufficient reach into the population of interest, enhancing generalisability and reducing the possible impact of bias.

Drawing participants from the broader population, rather than a clinical sample, is a comparative strength when viewed alongside limited existing research focused on children with milder hearing loss. This PhD’s outcomes reflect the plausible scenario of broad heterogeneity in language outcomes for children with this degree of loss, as seen in the results for all three aims. This contrasts to more selected samples, in this field commonly sourced from early intervention agencies, whereby outcomes may be measured from within a narrower and potentially skewed pool of ability (e.g. participation by enrolled early intervention participants, who may have remained in the program due to successful outcomes).\textsuperscript{155} Additionally, the participants with hearing loss in Aim 3 all had bilateral losses. This contrasts with other research on the same age group
reporting outcomes for children with “minimal” hearing loss, which combines unilateral hearing losses with mild bilateral losses. This PhD provides data that is specifically applicable to children with bilateral losses - a group whom audiologists manage differently to children with unilateral losses.

Aspects of diagnosis and management of hearing loss were reflective of current practice in Australia. The contemporaneous samples (i.e. well-established UNHS participants from Aim 1, the participants in Aim 2, and children with hearing loss in Aim 3), all drawn from a population-level databank, were in the majority early-diagnosed and early-amplified. This is consistent with expectations resulting from earlier identification under UNHS systems and likely replicates the pattern of intervention seen in the general community. Australian Hearing, independent of the PhD candidate, conducted all aspects of hearing aid programming and management. This means that the assessed children likely used hearing aids as they are typically prescribed and worn. This contrasts to clinically-based samples, where amplification may be verified and adjusted prior to measuring outcomes. In a large multi-state study in the United States, up to 55% of children wore amplification with insufficiently prescribed gain in both ears. It could be argued that under the Australian system, where audiologists from one national organisation (Australian Hearing) are responsible for prescribing amplification following a defined protocol, the likelihood of insufficiently prescribed amplification is lower than reported in other jurisdictions. Nonetheless, if under-amplification does occur in Australian practice, the population-base of this PhD likely documents outcomes achieved wearing typical rather than optimal amplification. Therefore, such results are likely to be highly representative of everyday challenges faced by children with amplification.

8.4 Limitations
The component studies of this PhD also have a number of important limitations. One is the size of the samples achieved. There is a generally recognised relationship where precision is inversely proportional to sample size: a smaller sample would lead to less precision around effect estimates. For Aim 1,
the number of participants approached for active data collection, combined with those from historical cohorts, led to an adequate sample size. The analyses conducted support the conclusions that suggest improving secular trends for moderate but not mild losses. Unlike Aim 1, for Aims 2 and 3 the final samples were considerably smaller. However, the number of children with hearing loss in these two aims represented all available and eligible children in the population registered with the over-arching databank that this PhD was nested within. This meant that, despite the lowering of precision around the measured effect estimates and acknowledged restriction of analysis options available, these results still represent the best estimates obtainable for this population.

The conclusions drawn from Aim 1 examining the secular trends in outcomes across different systems of hearing loss detection may not be fully generalisable to the small proportion of children with both hearing loss and intellectual disability. This is because such children were originally excluded from the oldest historical cohort used in comparative analyses, the Children with Hearing Impairment in Victoria Outcome Study (CHIVOS107). It was important to include CHIVOS children, as they allowed for an extension of comparisons of outcomes to before systematized approaches to hearing loss detection were in place in Victoria. This resulted in conclusions about trends over time that are relevant for most the population without intellectual disability.

Conclusions across all aims may also not apply to those children with hearing loss who are unaided. This is likely to be of minimal significance as outlined below. The aided status of the PhD participants is due to two reasons. The first is that historical cohorts used for comparisons of outcomes were originally designed to draw participants only from amplified populations. The second is that children who were part of active data collection in both age groups of this PhD reflect what is current common practice and were majority fitted with hearing aids for mild and moderate degree losses. It could be hypothesised that children with these degrees of loss who are unaided are not the children who are of specific interest to this PhD. Presumably, children with known hearing loss without hearing aids are achieving acceptable oral language outcomes, else no-
cost assistance in the form of device fitting would likely have been pursued. The population-based recruitment platform used in the active data collection activities of this PhD does not exclude unamplified children. Simply, it appears that the databank may reflect both the frequent decision to aid these children and the population trend towards earlier amplification. Therefore, the results reported are highly applicable to the majority of children identified with these hearing losses.

As mentioned in Section 7.3, children with hearing loss are reported to have rates of comorbidities approximating 20-40% (see Cupples et al (2014)246 for a summary). In this PhD, there was very limited information about known comorbidities of the participants. This precluded any analyses incorporating disabilities into the relationship between hearing loss and language outcomes. However, children with comorbidities can be included in studies of outcomes for children with hearing loss, even those designed specifically to exclude them. How frequently this would occur is dependent on the severity of the additional disability, which would influence the chance of it being pre-diagnosed. Also influential is the age of the participants, with studies of younger children likely to have a greater proportion of undiagnosed conditions compared to samples of older children from the same population. As a population-based study, children with comorbidities were almost certainly included in the sample and analyses, even despite Aim 1’s intentional exclusion of children with intellectual disability. Additionally, given the previously noted sample size restrictions, it is unlikely that information on comorbidities could have been used in other than a descriptive manner. What this PhD study provides is an overall population-level analysis of the outcomes of children with mild-moderate hearing loss, considering all children in the one population.

No direct assessment of language skills occurred for the children in Aim 3 of this PhD. Parent-report of expressive vocabulary was the only measure of language ability, in contrast to the directly-assessed language skills in the older age-group of children. Given the age of the younger children (around 2 years at assessment), parent-report was the most efficient and appropriate method for
obtaining language data. Comprehension of and compliance with performance-based tests would have likely reduced an already small sample size, and limited availability of appropriate age-based normative scores are reasons why a direct measure may not be appropriate.²³⁶ Had a direct measure been used, this may have also biased results towards better performance. This is because those children with better language skills would have had greater likelihood of successfully completing the required tasks. Use of a direct measure would have also prevented a direct comparison to the children without hearing loss from the population-based ELVS cohort. This large sample from the same geographic state was valuable for comparison of parent-reported early language performance, providing additional contextualisation of language skills other than reliance solely upon the published test norms. Concerns around the validity of parent-report measures have decreased over recent years, with the McArthur Bates Communicative Development Inventory used in this PhD specifically designed to optimise validity²⁴⁹ and has been used in multiple studies of language ability in children with hearing loss.⁹⁸, ¹⁷³, ¹⁷⁵, ¹⁷⁷, ²⁵⁰

Parent-reported data were also relied on for understanding child hearing aid use (specific to Aims 2 and 3). Unfortunately, this study revealed that activating electronic data logging of hearing aid use was not routine practice for all clinicians at Australian Hearing. When logging was activated, the needs-based review schedule for children led to daily estimates of hearing aid use being calculated from variable periods of usage data (e.g. a couple of weeks to close to 12 months of use). This reduced confidence in objective measures of device use within the limited number of children where data logging was activated. Whilst the literature suggests that parent-reported hearing aid use typically overestimates true use by around 2.5 hours per day,¹⁹³ these parent estimates of device use significantly correlate with actual data logging figures (r=.76). This suggests most parents overestimate their child’s hearing aid use by around the same amount, providing some reassurance for use of subjective parent-reported data when considering trends of hearing aid use rather than direct associations. Given this, it is possible that the weak correlation seen between higher hearing aid use and lower expressive vocabulary (Aim 3) is either a true weak
association that requires further investigation with the use of objective device use data, or it reflects concern with vocabulary development and higher use as a result (i.e. reverse causation).

The hearing aids used by participants were their regular aids provided by the government funded Australian Hearing. As the researcher was not affiliated with Australian Hearing, direct output from the hearing aids via real ear measures of hearing aid performance to verify how suitably the devices were fit to a child’s hearing loss could not be obtained (see Chapter 4 for methodological details). Had verification occurred, this may have meant that children “under fitted” with amplification would have been identified and had their hearing aids adjusted prior to their language assessment (particularly relevant for Aim 2 of this PhD). The alternative approach of using test box rather than real ear measures, despite being similar to extant published research,\textsuperscript{196} could have introduced some unmeasured variance between simulated and actual hearing aid performance. Whilst the rapid ear canal growth of the first few years of life has slowed by the age of the assessed group (5-7 years)\textsuperscript{195} meaning a reduced risk of significant real ear to coupler measure differences, the absence of the child's hearing aid earmoulds for the coupler measures may still have influenced the results obtained. It is possible the language abilities measured were somewhat poorer than the child’s potential when optimally aided.

\section*{8.5 Interpretation in light of prior literature}
The following sub-sections explores the key PhD findings and how they relate to previous studies.

\subsection*{8.5.1 Aim 1}
The three hypotheses relating to Aim 1 were discussed briefly in the paper forming Chapter 5 and are elaborated upon in this chapter, within discrete sub-headings and then at the overall theme level.
8.5.1.1 Aim 1a

Hypothesis: Children with both mild and moderate congenital hearing loss would display:

a. Secular trends toward improving language, behaviour and quality of life with evolving systems of early detection

There was partial support for this hypothesis. Children with moderate hearing loss displayed secular trends toward improving language as hearing loss detection systems evolved. No consistent trend was observed for children with mild loss. Behaviour and quality of life outcomes remained largely unchanged for both degrees of loss across a nearly 20-year period of births. Scores for children in the contemporary well-established UNHS cohort were not significantly different when compared to those of children with normal hearing or the test normative values.

As there is not a body of literature that examines secular trends specifically for milder hearing losses, interpreting the Aim 1a findings in light of prior research is challenging. Similar studies reporting outcomes in children identified under differing systems of detection have either defined the hearing loss of interest as moderate or greater,61,62 or if including mild losses did not specifically present outcomes at the degree of loss level. 156 This PhD’s reported trend towards an improvement in language outcomes for children with moderate loss of approximately 0.6-0.7 SD as detection systems evolved from risk factor to UNHS is slightly higher than those observed in a similar population-based study by Wake et al (2016)156 (approximately 0.3-0.5 SD improvement with detection shifting from risk factor to UNHS). The likely reason for this difference is the overall poorer language skills of children with greater degrees of loss (i.e. severe and profound) included in the Wake et al (2016) sample but excluded from this PhD sample. These greater degrees of loss may have depressed the group outcomes, compared to the results of this PhD study. Wake and colleagues also reported a pattern of poorer language-related outcomes with increasing hearing
loss severity. This was a pattern not clearly observed in the results presented for Aim 1a. By restricting the degree of hearing loss to mild and moderate for a small and heterogeneous sample, this reduced the ability to observe performance patterns across the narrower severity range.

Of note, an interesting trend was observed in the Outcomes of Children with Hearing Loss (OCHL) study in the United States that is similar to this PhD’s results. For children aged 2-6 years across the severity range mild to severe, poorer language outcomes with increasing severity were noted, which is consistent with other large studies of children with hearing loss.\textsuperscript{156,159} However, Tomblin and colleagues noted that whilst the outcomes of their children with mild loss were significantly different from children with severe loss, they were not significantly different to those achieved by children with moderate loss.\textsuperscript{164} It is plausible that the agreement seen between the current PhD study and Tomblin et al (2015) may also be observable in other large studies if analyses between specific degrees of loss were conducted. Unfortunately this does not appear to be a commonly conducted analysis, which in a study of broad range of hearing loss raises challenges with drawing conclusions for smaller subsets of losses.\textsuperscript{97}

8.5.1.2 Aim 1b

Hypothesis: Children with both mild and moderate congenital hearing loss would display:

\begin{itemize}
\item[b.] \textit{Better outcomes if detected with hearing loss earlier, regardless of detection system to which exposed}
\end{itemize}

This hypothesis was also supported for children with moderate hearing loss, but not for children with mild loss. Children with moderate loss experiencing better language outcomes if diagnosed prior to 6 months of age is consistent with findings that provided early evidence for the continued UNHS expansion internationally.\textsuperscript{93,204} Such a result is also consistent with more recent studies with robust samples and methodologies that attempted to quantify the benefit to
language outcomes of earlier detection. All of these recent studies included children with moderate degree of hearing loss. Of the three that included children with mild loss, unfortunately none quantified outcomes within discrete degrees of loss by their age at diagnosis. The absence of a literature base on the effect that earlier detection of mild hearing loss has on outcomes is not surprising; historically the bulk of outcomes evidence has been gathered from children diagnosed late, as such studies pre-dated newborn hearing screening.

Several factors may contribute to why children with moderate, but not mild, loss show benefit in measured outcomes with earlier diagnosis. For illustrative purposes, these factors are outlined in a hypothesised sequential manner from the point of an early diagnosis of both a mild and a moderate hearing loss. Firstly, the moderate hearing loss may have been diagnosed more quickly than the mild loss: mild diagnoses are vulnerable to delays as a result of lower confidence in hearing loss threshold estimation compared to greater degrees of loss and challenges differentiating mild permanent losses from temporary conductive losses. At confirmation of diagnosis, the parents of the infant with moderate loss may receive clearer recommendations on how to assist their child. This would facilitate confident decisions on the importance of early aiding and intervention: several studies have highlighted the impact on families of clinician uncertainty regarding advice for mild loss management. As the children develop, the parents of the child with moderate hearing loss may receive more consistent behavioural evidence of the hearing loss compared to the parents of the child with mild loss. These indicators of difficulty hearing may reinforce the importance of the chosen intervention method for the child with moderate loss, just as subtler or inconsistent difficulties displayed by the child with mild loss may reduce the resolve of families to adhere to any intervention provided. Access to intervention other than device fitting may be less of an option for the family of the child with mild loss. At the time this study was conducted, unlike the child with moderate loss, the child with mild loss was ineligible for state-funded early intervention (personal communication) and
those out of pocket options may not provide the same degree of intense therapy that is historically provided to children with moderate to profound loss.87

The above series of events may contribute to a divergence in the language skills of children early diagnosed with mild versus moderate hearing loss by the time they reach the early school years when researchers often assess abilities. The cumulative auditory experience that the child experienced up to this point,196 if device provision and use is actually beneficial for all losses, could be an important factor. Cumulative auditory experience refers to the combined effects over time of optimised audibility of speech, consistent hearing aid use and intervention occurring early to take advantage of optimal learning periods.111 Also, the sensitivity of the measures used to assess language skill is important, with construct validity to document benefit to language required for both degrees of loss at the appropriate child age.250 Somewhat related to the previous point, the gains from earlier diagnosis measured for the child with moderate loss may be greater than that of the child with mild loss. This is because of greater scope for improved outcomes in the more severely impacted early diagnosed child.

With regards to amplification, there is evidence internationally of considerable delays between diagnosis and fitting for children with a mild hearing loss, whereas earlier amplification is typically observed with greater severity losses.87,188 If earlier diagnosis252 and amplification170 is truly influential in improving the outcomes of children with mild loss, then prompt provision of devices would be critical. The results from this PhD study are not in alignment with delays in amplification trends seen internationally; on average children with mild loss in the well-established UNHS cohort were fitted 3.8 months after diagnosis (at age 8.3 months), compared to 6.9 months after diagnosis for children with moderate loss (at age 12.4 months). Thus in this sample of children, those with mild loss were not “disadvantaged” by a later fitting of amplification by comparison to their peers with moderate loss.
A crucial element for hearing aids is also their actual use after fitting. Children with mild losses typically wear their amplification for less time each day when compared to children with greater degrees of loss, with use of amplification in younger children lower and more variable than in older children.\textsuperscript{193, 194} Whilst hearing aid use was not specifically measured as part of Aim 1, a discussion of the impact of amplification fitting must consider use. Further discussion on this topic can be found in Sections 8.5.1.4.1 and 8.5.2.3.

The conclusion from screening literature is that detection in and of itself is not the answer to improved outcomes, it is the combination of detection and treatment that likely leads to measurable differences.\textsuperscript{61, 64} It has been proposed throughout this section that optimisation of device use is a factor that should lead to improved outcomes for children with mild loss. An alternative should be considered, which is that earlier device use by children with mild loss may not result in a functional gain sufficient to improve their language outcomes. Section 8.5.1.4.2 specifically discusses non-auditory factors and is where this alternative theory is outlined.

**8.5.1.3 Aim 1c**

*Hypothesis: Children with both mild and moderate congenital hearing loss would display:*

\textit{c. Comparable mean outcome performance for the group exposed to the well-established UNHS system and typically-developing children without hearing loss.}

The evidence for this hypothesis varied by outcome. Mean outcome performance between children with hearing loss exposed to well-established UNHS was comparable to the representative sample of children without hearing loss in the domains of health-related quality of life, behaviour, and receptive language. However, in expressive language ability, children with both mild and moderate hearing loss performed around two thirds of a standard deviation below the representative sample without hearing loss. A comparison of receptive...
vocabulary outcomes was not possible as the historical sample without hearing loss was not assessed in this language domain.

Receptive language skills reaching those of children with normal hearing is not the typical finding for studies of children with hearing loss, unless the degree of loss is very mild or the sample had limited generalisability to the broader population (e.g. measuring outcomes of children enrolled in intensive early intervention programs). More commonly, a pattern is observed of better receptive versus expressive language skill, yet with both receptive and expressive language ability still found to be below age-appropriate scores. The pattern of receptive language results in this PhD study is challenging to explain. Examining the scores of children in both the hearing loss and non-hearing loss cohorts separately may help. Firstly, the ELVS cohort were scoring below the expected normative score by approximately one third of a standard deviation (94.3 versus expected score of 100). Secondly, the children with hearing loss scored quite differently depending on their degree of loss. Children with mild loss were as a group scoring slightly above the expected score (101.2) and those with moderate loss at the same level at children without hearing loss (94.9). Thus when combined, the receptive language score of children with mild-moderate loss was slightly better than that of children without hearing loss (97.5 versus 94.3, fully adjusted means). These scores were all derived from performance on the same Australian standardised measure, the CELF-4.

It is possible that the Australian norms for the CELF are not representative of a population-based sample. At age 4 years the ELVS cohort without hearing loss were as a group already scoring lower than standardised norms (mean score 96.7, SD 14.9, measured using the preschool version of the CELF). This approximate 0.2 SD lower than expected performance is similar to the performance seen at age 7 years in this study and may suggest a consistent under-performance against normative scores in receptive language ability. Alternatively, the sampling framework of the ELVS study (see Section 4.4.3) may not have achieved a truly representative cohort of children, with some skew towards poorer receptive language ability. This is perhaps the least likely
scenario, as expressive language skill in this population of children without hearing loss has reached expected norms at both 4 and 7 years of age (mean scores 99.6 and 99.0 at 4 and 7 years respectively).

If there is concern regarding the normative scores for measurement of receptive language in children without hearing loss, this should also apply to children with hearing loss from the same population. Instead, mean performance exceeding the normative score was observed in the children with mild hearing loss as described above. The sample size of children with hearing loss, alongside heterogeneity in ability, should also be considered. Figure 2 in Chapter 5 displays wide 95% confidence intervals around mean receptive language performance of children detected with mild and moderate hearing loss under the mature or well-established UNHS system (approximately 25 points for mild loss). This limited precision from a sample of 9 children with mild hearing loss may have left the mean receptive language estimate for this degree of loss vulnerable to imprecision. This is of particular interest as the pattern of very good performance in receptive language is not mirrored in other closely associated language domains.

In summary, the receptive language pattern of results remains challenging to explain. Possible factors at play include the norming of this language domain for a population-based sample, alongside variation in scores resulting from a small sample size. However, the overall patterns observed for expressive language are unambiguous and are expanded upon below.

This sample of children with mild and moderate hearing loss detected under a mature UNHS system display evidence of an expressive language delay at age 5-7 years compared against peers without hearing loss. A deficit in a broad domain such as expressive language performance is consistent with reports of children with slight/mild and mild to moderate losses having poorer language skills within specific areas such as phonological short-term memory, phonological discrimination and morphosyntactic ability. These studies often
reported deficits in specific language domains without relating them to a broader global language measure such as that used in this PhD study.

This study’s results also align with those from recent larger samples, indicating children with mild and moderate loss continue to have lower than expected language skills, even when identified early.\textsuperscript{157,164} Such studies derived a global language measure from results in multiple speech/language domains, prior to controlling for predictor variables in analyses. In language assessments up to the age of 6 years, the OCHL group showed that children with a later diagnosis were able to “catch up” with their earlier detected peers. However, as a group, these children were still performing more poorly compared to their peers with normal hearing (e.g. at age 6 years, children with mild loss were on average 10 composite language score points below peers with normal hearing, children with moderate loss 15 points).\textsuperscript{164} This represents a lower performance by children with hearing loss ranging two thirds to one standard deviation dependent on hearing loss severity. The consistency of these results to the current PhD study support a hypothesis of continuing delay in language outcomes over time, despite general trends towards earlier identification.

\textbf{8.5.1.4 Aim 1 overall theme of results} 

Overall, it appears children with mild loss may not experience clear benefit from earlier detection and aiding. This contrasts with trends observed for children with moderate loss. Both the quasi-experimental and longitudinal results pointed to this same conclusion. This agreement between experimental approaches provides confidence in the conclusions\textsuperscript{256} on possible benefits of earlier diagnosis and fitting with amplification varying by degree of loss.

This pattern of results is all the more surprising given that, over the two decades that separate the oldest and youngest children, there are many reasons other than early hearing loss detection for childhood language outcomes to have improved. Maternal education levels have risen markedly, which should correlate with increased consistency of hearing aid use\textsuperscript{193} and stronger child language outcomes have resulted from increased and more responsive parental
language skills.\textsuperscript{257} Hearing technology itself has improved, as has the ability to verify fitting to prescribed amplification targets despite the known challenges with maintaining an optimal fit during childhood.\textsuperscript{247} Thus, even had the age of detection and fitting remained constant and not actually reduced at all, we might have expected better outcomes over time. Reasons for the difference in the trends in outcomes over time between the two degrees of loss are considered below.

8.5.1.4.1 Amplification-related factors

One possible reason for no clear benefit from earlier identification and amplification is that children with mild loss may not wear adequately-fitted hearing aids for enough time each day in order to accrue significant benefit. Many factors could contribute to this scenario. These cover aspects of obtaining amplification and maintaining use, and the terminology commonly used in this space to understand the degree of loss and the impacts it may have on the child and family.

Commencing hearing aid use in children with mild loss is contingent upon caregiver decision-making. These decisions are made ‘within’ a hearing loss detection system, an aside to the broader secular trends theme that was the focus of this aim. From the OCHL sample in the US, a study of the influence of hearing aid use on outcomes of children with mild bilateral loss determined an initial delay between detection and diagnostic confirmation of hearing loss for the 61\% of the sample identified through UNHS.\textsuperscript{196} The average age at confirmation of hearing loss was reported to be 19.5 months. Holte et al (2012),\textsuperscript{78} for the same OCHL cohort, reported that one reason for such delay was the subjective parental observation of their child’s responses to sound. Such hesitation by parents is understandable; the environments that young children are often in are unlikely to elicit the known challenges of mild hearing loss (see earlier Section 2.4.4). Activities with the child are likely to occur within close proximity, often speech will be directed to the child and occur within their ‘listening bubble’, the optimal zone around the child for hearing success.\textsuperscript{258}
Reassurance from such visual displays of hearing may be reinforced by a perception of ‘mild’ hearing loss not being significant.\textsuperscript{112} These viewpoints may be held not only by families but also by professionals.\textsuperscript{79, 112} Potential misinterpretation of the language used to describe hearing loss within the range of approximately 15-40 dB HL was highlighted in 2004 by Fred Bess who, when discussing minimal hearing loss (i.e. the umbrella term for both unilateral and mild bilateral hearing loss) said “Minimal is not inconsequential”.\textsuperscript{251}

Haggard and Primus (1999)\textsuperscript{34} exposed 36 normally hearing parents with normal hearing children aged 7 years and younger to filtered auditory recordings designed to simulate hearing losses. These recordings were assigned labels using common terms such as slight, mild and moderate and were engineered to represent increasing difficulty hearing. After listening to each recording presented in random order, parents completed questionnaires that probed their subjective impression of the simulated hearing losses as if their own child had that hearing loss. These questionnaires asked parents to choose terms that best represent the hearing loss they had just heard. They also required parents to numerically rate their perception of how much difficulty their child would have in various hearing-related tasks if their child had that hearing loss. Parents consistently described the simulated losses using terminology that would suggest substantially greater impact of the hearing loss than the conventional labels for classifying losses (slight, mild, moderate) would indicate. This suggests that the caregiver, when exposed to a simulation of what their child hears, may appreciate the severity of listening challenges more than what they would assume their child experiences based off the commonly used terms for degree of loss.

This PhD’s most recent sample (the well-established UNHS sample) was identified and fitted with amplification early. This has effectively minimised concerns that delays in fitting amplification is why children with mild losses may not experience the clear benefits from early detection. However, the challenges with common interpretation of language such as ‘mild’ used to describe this
degree of loss that were outlined in the preceding paragraphs could impact the amount of hearing aid use once fitted.

It is noted in the literature that consistent amplification use by children with mild loss occurs less than 50% of the time, but improves with increasing age. Maintaining hearing aid use, particularly at younger ages, has been linked to improved language outcomes of children with hearing loss, including those with mild losses. Hearing aids fitted from an early age, during the period of maximal neural plasticity, consistently worn and optimised to hear speech, should lead to improved outcomes. Walker et al (2015) refer to this process as defining a child’s ‘cumulative auditory experience’. Parents having difficulty observing a benefit of hearing aid use in either language development or behaviour are suggested as limiters to maintaining consistent use of amplification. Challenges with maintaining hearing aid use at younger ages likely act alongside the impact of hearing loss severity (i.e. less consistent use with milder degree of loss) to compromise attempts at maintaining use in the long term.

8.5.1.4.2 Non-auditory factors
Discussed above is the possibility that non-optimal use of hearing aids has precluded the true language benefits that would accrue from optimal early aid fitting and use. This would be the most likely contributor to the poorer than desired outcomes observed in this study, if ideal amplification-related practices are the enabler for improving outcomes of children with mild loss. However, based on the reported results in this thesis, an alternative scenario to consider is one where children with mild loss may not be able to benefit from even optimised amplification practices. Whilst this hypothesised scenario is at odds with the limited existing literature on the benefits of amplification for mild losses in clinical or advantaged samples, it warrants consideration because there is not yet conclusive evidence of benefit from optimised amplification in children with mild loss at a population level.
The concept of overdiagnosis could be considered as part of this scenario. This concept was briefly discussed in the paper forming Chapter 5 of this thesis specifically in relation to mild congenital hearing loss. “Overdiagnosis” is defined as the identification of a real condition (i.e. not a misdiagnosis), for which the treatment provided does not benefit an individual’s outcome. Overdiagnosis is more prevalent in literature focused on adult subjects (e.g. Bleyer & Welch (2012)), but is emerging as a recognised factor in paediatric study populations. Neuroblastoma screening is highlighted as one example of overdiagnosis in children, as a proportion of neuroblastomas spontaneously regress without any treatment during infancy. Screening for this condition leads to the increased identification of lower-stage cancers. However, in population-level studies such screening has not reduced mortality rates from end-stage neuroblastoma.

Unfortunately, lack of benefit does not equate to lack of harm. Earlier diagnosis may not always be worth the costs incurred. There are reports of overdiagnosis leading to unnecessary risks for the child and potential harm to the family. In a review of articles published in one calendar year on paediatric overuse, Coon et al (2017) highlighted the risk of developing malignancy associated with phototherapy exposure as the result of hyperbilirubinaemia treatment thresholds set too low. Another identified example was head circumference screening for neurocognitive disorders. This screening was neither sensitive nor specific for the target condition, as at the population-level neurocognitive disorders are more common in children with normal head circumference. Routine measurement can lead to unnecessary and costly follow-up, as well as increased family anxiety.

In the case of hearing loss, the balance between benefit versus harm from early diagnosis and resulting earlier intervention should also be considered (see Section 2.3.6 for benefit versus harm discussion). Children identified early with mild losses and aided early are still exposed via treatment (i.e. hearing aids) to the same potential harms (e.g. burden, stigma) as those children with more significant losses. These harms may be harder to justify in a scenario of
diminishing gains, such as the reduction in benefit to measurable outcomes you might expect with decreasing hearing loss severity. For example, children with mild losses are closer to the expected language scores than children with severe losses, which means the magnitude of any improvements in outcomes following treatment would be smaller in those with milder losses assuming both groups of children have the same language and cognitive potentials. This may mean that evidence of overall no harm to parents and families as a result of UNHS49,267-269 may no longer be balanced on the side of no harm when considering children with mild loss, given the projected benefits from the earlier detection and aiding that function to off-set any treatment harm are demonstrably lower.

The concept of mild hearing loss being an example of “overdiagnosis” would serve to delineate mild from moderate and indeed greater hearing loss; children with the greater degrees exhibit clear benefits to their language outcomes as a result of their “treatment” being the earlier fitting of hearing aids (moderates - this PhD study) or cochlear implants for severe and profound losses (e.g. Ching (2015)157). If mild hearing loss is an example of an overdiagnosis and early hearing aid fitting considered an overtreatment (due to a lack of documented improvement in these PhD findings resulting from earlier reported hearing aid fitting), a reasonable question remains as to why these children have poorer language outcomes than their normal hearing peers. One possible reason is that these poorer language outcomes are not a direct product of their hearing status. Instead, they may result from some other developmental difficulty.

Other developmental difficulties, possibly tied to the cause of permanent mild loss (which in the absence of identifiable causes could be presumed genetic, see Section 2.2.3), may contribute to the documented language delay. There is a reported comorbidity rate in children with hearing loss that ranges from 20-40% and is hypothesised to have a role influencing, amongst others, language outcomes.61, 246 This may mean that these children cannot demonstrate a functional gain via intervention with hearing aids that would otherwise be sufficient to change language outcomes in the absence of comorbidities. If this were the case, then optimising and maintaining hearing aid use alone may be
ineffectual to improving speech and language outcomes. Excluding the amplification factors discussed in Section 8.5.1.4.1 (which were not accounted for in this Aim), the presented results do lend some support to this theory of developmental difficulties rather than hearing ability impacting measured language outcomes. It also remains possible that amplification and non-amplification related factors could interplay in the management of these children. Vigilance regarding additional disabilities, diagnosed or otherwise, should play a role in monitoring speech and language progress rather than focussing solely on amplification factors.

8.5.2 Aim 2

8.5.2.1 Aim 2a

Hypothesis: For children with mild or moderate bilateral hearing loss who wear hearing aids:

a. Aided audibility values would correlate to both unaided hearing acuity and speech recognition ability

This hypothesis was supported. The calculated speech intelligibility index (SII) values correlated strongly to both unaided hearing acuity (i.e. PTA thresholds, correlations ranging from -.60 to -.79) and children's ability to recognise open-set sentences in the presence of competing speech (i.e. LiSN-S speech reception thresholds, correlations ranging - .63 to -.73). These findings support the SII calculation used as being an appropriate measure for estimating aided audibility in this sample of early-identified children with mild or moderate bilateral hearing loss.

The association between the SII and PTA thresholds was reassuring, as essentially both values quantify a form of hearing ability. Therefore, some degree of relationship was expected between the scales from which the values were derived. Calculating the SII requires the multiplication of audibility functions (speech that is supra-threshold within specified frequency bands) by band
importance functions for speech (in this instance the ANSI 2007 standard band importance function). Had there been lack of an association, this would have suggested an error in the SII calculation. The Situational Hearing Aid Response Profile program used to calculate the SII has a reported prediction accuracy of within 5 decibels of the actual measured hearing aid output in a small verification sample.\textsuperscript{270} Similarly, the convention in audiometric threshold seeking (which determines the thresholds from which the PTA is calculated) also acknowledges a ±5 decibel test-retest variance.\textsuperscript{271}

The above correlation between SII values and speech recognition scores is consistent with previous reported findings.\textsuperscript{21, 22, 171} In a sample of four adults and 29 children (14 children with mild to severe hearing loss), Scollie (2008)\textsuperscript{22} determined that SII scores could be used to predict consonant recognition scores with differing accuracy for adults versus children. For children, greater accuracy in predictions was observed when both age and hearing loss were incorporated into transfer functions fitted to the data. In a larger cohort of 116 children and 19 adults, all with normal hearing, McCreery and Stelmachowicz (2011)\textsuperscript{21} reported similar findings to Scollie (2008). The use of age-specific data within the SII was necessary to avoid overestimation of speech recognition performance for children, tested in their study using consonant-vowel-consonant non-word stimuli. In a study of 40 children (16 with hearing loss, ranging mild to profound), SII scores were shown to significantly correlate with word recognition performance measured using the Lexical Neighborhood Test and the Multisyllabic Lexical Neighborhood Test.\textsuperscript{272}

Of note, the results reported in the present PhD study extend the published SII findings for children with hearing loss. In a novel finding, a correlation between SII and speech recognition scores was observed for sentence-level material, delivered in the presence of competing speech. Such stimuli are highly representative of common listening environments where children commonly encounter difficulty. These results provide evidence of an accurate and convenient method of predicting speech recognition ability under realistic
listening conditions for children with aided mild or moderate bilateral hearing loss.

8.5.2.2 Aim 2b

Hypothesis: For children with mild or moderate bilateral hearing loss who wear hearing aids:

b. Measures of aided audibility would be more highly associated with speech/language outcomes than unaided hearing acuity

No evidence was observed to support the hypothesis that aided audibility is more highly associated with speech/language outcomes than unaided hearing acuity. The results obtained suggest no association between the outcomes examined and any hearing measures, including the novel SII measures. This pattern of results is consistent with findings by some but not others. This PhD study’s results should be considered in context of the limitations described in Section 8.4, including the challenge of a small sample size, plus heterogeneity in outcomes. The literature is discussed below in Sections 8.5.2.2.1 and 8.5.2.2.2, according to method of quantifying hearing (i.e. unaided and aided).

8.5.2.2.1 Unaided hearing acuity and speech/language outcomes

The existing literature on the relationship between speech/language outcomes and unaided hearing ability in children with mild-moderate hearing loss presents somewhat mixed results. Where some purport no association, consistent with this Aim’s results, there are reports that support an association. Differences in experimental design and approach to analyses could factor into this lack of consistency. All literature showing no association between unaided hearing ability and speech/language outcomes was either conducted on children with hearing losses specifically in the mild to moderate range, or explicitly tested the relationship between the two degrees of loss as part of analysis of a greater range of hearing loss.
The one report of poorer performance in expressive grammar and word repetition with increasing severity of unaided hearing loss was conducted in French-speaking adolescents with mild-moderate loss aged 11-15 years.\textsuperscript{254} The hearing in schools study\textsuperscript{90} also highlighted poorer phonological discrimination ability in children aged 7-11 years with slight/mild loss, yet no relationship was observed between degree of loss and performance across what was a very narrow decibel range of participant hearing loss. The differences in language and age of French-speaking participants in the study by Delage and Tuller (2007)\textsuperscript{254} when compared to other reported results (majority in younger English-speaking children) may have influenced the differing pattern of results in studies of children with mild-moderate loss.

Studies of broad ranges of hearing loss approach analysis of language ability between participants with different degrees of hearing loss in different manners. Some do not report on inter-degree trends (e.g. between mild and moderate losses, see Ching et al (2013)\textsuperscript{159}) and instead focus on hearing loss as a continuous variable. Others provide an overview of both, which is of interest in this discussion. For example, Wake and colleagues (2005) studied 7 to 8 year old children born with mild to profound hearing loss and reported overall that worsening PTA was associated with decreases in language scores.\textsuperscript{163} However, the authors also conducted analyses using severity of loss as a categorical variable, estimating language scores adjusted for age at diagnosis and non-verbal IQ. These analyses revealed total language scores (receptive and expressive subsets combined) that on average did not significantly differ between children with mild and moderate loss (mild mean score 88.8, standard error 4.7; moderate mean score 82.7, SE 3.5).

A similar result was noted by Tomblin et al (2015)\textsuperscript{164} for children with hearing loss ranging mild to severe. At the cohort level they reported that worsening PTA was associated with decreases in global predicted language scores for participants aged 2 to 6 years. Through supplementary analyses they reported that whilst children with mild loss performed significantly better on language tasks than children with moderately severe loss, there was no significant
difference observed in language ability between participants with mild and those with moderate loss. This statement is not accompanied by estimates of language scores, instead language ability by chronological age is displayed graphically.\textsuperscript{164} A similar pattern was observed in the OCHL group’s spontaneous language outcomes of 3-year-old children, which aimed to observe language skills in children with hearing loss in conversation rather than via standardised measures. Quality (a label derived from a number of measures, including utterance length and number and number of different words), rather than quantity (total number of words and utterances, words per minute), of the elicited language sample was found to be associated with PTA. Children with moderate and severe loss had significantly poorer quality language than children with normal hearing. Children with mild loss had significantly better language than those with severe loss. However, no significant difference in language quality was noted between children aged 3 years with mild and moderate loss.

To summarise, the lack of an association between unaided hearing ability and speech/language outcomes in this PhD study is generally in agreement with the existing literature that considers the outcomes of children with mild and moderate hearing loss in a categorical rather than continuous manner.

8.5.2.2.2 Aided audibility and speech/language outcomes

Contrary to previously reported relationships,\textsuperscript{170,171} this study observed a lack of evidence for an association between the SII and speech/language outcomes. This may reflect a novel result within a defined group of children with milder degree hearing losses. However, there are a number of factors that may contribute to a discrepancy in findings between this PhD study and the published literature. These are mainly encompassed by differences in experimental design and are detailed below, following an outline of the findings in the literature that present with contrary findings to this PhD study.

Stiles et al (2012)\textsuperscript{171} compared the lexical ability of 16 children aged 6-9 years with hearing loss spanning mild to profound to children with normal hearing. To determine whether the PTA or the aided SII was more predictive of outcomes,
the authors ran regression models incorporating either one or two predictor variables found to be significant. They found that the aided SII was a stronger predictor than the PTA for non-word repetition ability and receptive vocabulary after accounting for degree of hearing loss. It is possible that both the limited use of covariates plus a robust contrast in performance between children with normal hearing and those with a broad range of hearing losses were contributing factors to the significant effects reported.

Tomblin and colleagues (2014) examined a group of 180 children aged 3-5 years with hearing loss ranging mild to moderately severe to observe the impact of hearing aids on speech and language outcomes. They firstly reported that unaided hearing based off a PTA was associated with speech and language development using results of standardised speech and language tests combined into a single composite language score. Then, using an aided SII score, they showed a correlation with their composite language score. This significant association between an aided audibility measure (SII) and speech and language development occurred for all degrees of loss studied (mild to moderately severe). The strength of these associations were reported as approaching moderate levels and, similar to Stiles et al (2012), were measured after adjusting for unaided hearing levels.

Considering the participants in these studies, it is unlikely that the age difference between children in the two studies described above (3-5 years and 6-9 years) and the age of participants in this PhD study (5-7 years) is a key contributor to the lack of an observed association between SII and speech/language outcomes. Considering the trajectory of language development across childhood, no reports could be found of increased variation in language outcomes specifically in the middle age band (5-7 years) that could have altered any association seen at both the earlier and later ages. The present study recruited participants from a population-level databank, versus a more clinically/convenience-derived sample. This distinction is conceptually important. There is a reported common challenge with maintaining optimal hearing aid fitting in children; 55% of a sample of 195 children with hearing aids were listening using
amplification that did not meet prescribed targets. In a population-based sample there could conceivably be less monitoring and checking of appropriate amplification than in a clinical experimental sample. In turn, this un-optimised audibility may impact language performance.

Still considering aspects of hearing aids and aided audibility, different methods used to calculate the SII should be examined. In this PhD study, after selecting the better hearing ear based off the participant’s unaided PTA, a SII figure was then calculated for that ear and used in analyses. Other studies used real ear measures and determined which ear to use based off the better ear SII\(^\text{171}\) or used a residual SII that represented the difference between the aided and predicted SII.\(^\text{170}\) Thus the two published studies had confidence in their choice of the best SII score based off calculations on both ears, whereas this PhD study was reliant upon the better-ear PTA and better ear SII being the same ear. Whilst it is unlikely that there would be large discrepancies between these two hearing-related measures, it cannot conclusively be stated that the best ear audibility value was always used in analyses. This could have resulted in weaker associations between the aided audibility and speech/language outcomes given the ear with the better SII may not have always been factored into calculations.

Another influencing factor could be the use in this study of average values rather than real ear measures. The use of average values is similar to that used elsewhere,\(^\text{196}\) yet could have introduced some unmeasured variance between simulated and actual hearing aid performance. Also, earmould acoustics were not accounted for. Due to each child’s unique ear canal anatomy and properties, it is difficult to predict the direction and magnitude of any variation to measured SII/language associations resulting from the measurement of hearing aid performance in a test box without customised earmoulds attached. All of the factors discussed above may have impacted upon the measurement of hearing aid output, with direct flow-on to the association analyses performed.

Functional auditory outcome measures, often caregiver-completed, allow for an understanding of the impact aided audibility has in commonly encountered
listening environments. The functional assessment tool used in this PhD study (the PEACH, see Section 4.10.2) has also been shown to be associated with language outcomes. PEACH scores negatively correlate with worsening PTA levels\textsuperscript{23} and when administered prior to 2 years of age, were shown to be associated with language ability at age 3 and 5 years.\textsuperscript{157, 197} In a study with 356 children with losses ranging mild to profound, the PEACH score was combined with 8 other language measures to create a global outcomes score. With 15 predictor variables in the analyses, the inclusion of the PEACH score for 102 children administered at less than 2 years of age saw the amount of variation in 3 year old language outcomes that could be explained by the regression model increase from 40% to 45%. Due to the scale and sample size of the current PhD study, there were restricted options to explore any similar associations between the PEACH and speech/language outcomes.

8.5.2.3 Aim 2 overall theme of results

In summary, the aided audibility measure (the SII) calculated in this study strongly correlated with both the unaided hearing (PTA) and speech recognition ability (the LiSN-S) of participants aged 5-7 years with mild and moderate hearing loss. Neither unaided (PTA) nor aided (SII) measures of hearing were associated with speech and language outcomes: the unaided finding is generally consistent with trends in the literature, the aided finding is novel and at odds with the limited published studies.

If these results are correct, it would appear that no measure of hearing ability (i.e. unaided or aided) is consistently able to explain speech and language outcomes in children with mild and moderate loss at the population-level. Contrary to what has been suggested by others,\textsuperscript{171} this would restrict the ability to use aided audibility measures as a tool to guide predictions of speech and language outcomes for children with hearing losses specifically in the mild to moderate range. Children with these losses continue to perform poorly in their speech and language outcomes as a group despite earlier aiding.\textsuperscript{156, 157, 164} Thus if correct, a potentially useful tool linked to aided audibility may not be helpful to
guide improvements in the speech and language outcomes for a subset of children with hearing loss for whom there is still a significant number of unknowns. This may be due to early hearing aid fitting not benefitting this group, despite children fitted with hearing aids earlier than historically achieved.

It is important to note some overall limitations with the results for Aim 2. The sample size achieved did restrict options for analyses and limit the conclusions drawn. The correlation between language outcomes and measures of aided and unaided hearing resulted in medium effect sizes (in the range of .3 to .4) that, despite their size, were non-significant. Such results could suggest that sample size was insufficient for bivariate correlations. Therefore, it was not surprising that the exploratory linear regression analyses with multiple predictor variables produced effect estimates with wide confidence intervals. Also, for this sample it remains unknown what an individual child’s hearing aid use was (e.g. number of hours hearing aids are routinely used on a daily basis) over time since the age of early initial fitting through to language assessment at 5-7 years. Patterns of hearing aid use right across these critical early development years could impact an individual’s cumulative auditory experience, discussed in Section 8.5.1.4.1. Such variation in device use across these years could be influential on language outcomes measured at 5-7 years. Both the sample size and hearing aid use factors could influence the overall theme of results presented here.

A novel finding in Aim 2 yet to be discussed is the strong relationship demonstrated between the calculated measure of aided audibility (the SII) and speech recognition ability using ecologically valid stimuli (the LiSN-S). It could be reasonably expected that those who score well on speech recognition tasks should also perform well with language tasks, particularly when the evidence of speech recognition is, as in this study, from open-set stimuli where language knowledge is particularly influential. Therefore, it was unexpected that speech recognition ability was not consistently associated with speech and language outcomes as documented elsewhere.178
It is prudent to note that factors other than hearing acuity (unaided or aided) could be contributing to the lack of an observed relationship between speech recognition and language. One example is the relationship between cognition, language outcomes and speech perception. It is known that there are strong relationships between cognition and language outcomes and between language ability and speech perception skill. In a group of 62 children fitted with either hearing aids or cochlear implants, Sarant et al (2010) found non-verbal IQ had an impact on speech perception performance not directly, but indirectly via influence on language outcomes. Tomlin et al (2015), using the same measure of speech recognition as this PhD study, showed a significant correlation between non-verbal IQ and speech recognition ability for both children with normal hearing and those diagnosed with auditory processing disorder. The relationship between non-verbal IQ and speech recognition was not an aim of this PhD study and as such the influence of non-verbal IQ on this population remains unanswered.

As highlighted so far and considering the discussion in the next section of this chapter, there are multiple themes in the results for Aim 2 that may influence the association between aided audibility and speech and language outcomes. Clearer conclusions are necessary for this group of children who are being early aided yet are still performing below expectations for their language outcomes. The best way to provide generalisable and robust evidence would be through rigorous studies of larger numbers of children in a population-based setting.

8.5.2.4 Discussion of an interesting finding in an individual child

When addressing Aim 2, a consistent theme of variability in speech/language outcomes between participants was evident. Isolating one participant (participant 9) who had the most notable difference between cognitive ability and speech/language outcomes provided an opportunity to highlight factors that may contribute to this discrepancy in measured ability. Participant 9 had a non-verbal IQ score of 117, representing a cognitive potential over 1 standard deviation above the expected mean score. However, language and speech
standardised scores ranged from 70 to 94 which represented skills approximately 0.5 to 2 standard deviations below expected performance levels. This participant lives in a household where the primary language spoken is not English, and where parents have tertiary level education and less social disadvantage than the population norm.

Cognitive ability, as measured here by non-verbal ability, is reported to be a strong predictor of language outcomes in children under 5 years with hearing loss. Cognitive ability also continues to impact language development and academic outcomes throughout childhood. With IQ reported to be relatively stable across childhood, the discrepancy between high cognitive potential and underperformance on standardised assessments warrants exploration from several angles as detailed below.

It is possible that the child being distracted, tired or uncooperative at the time of assessment, influenced this discrepancy between cognitive ability and speech/language performance. However, the experienced researcher who conducted the assessment did not observe this had occurred. Demographic factors mentioned above (i.e. home language, parent education, family disadvantage) could have opposing impact on language ability. From a longitudinal study of language development in typically developing children, at the age of 2 years a non-English speaking background was associated with lower vocabulary production and strongly associated with low expressive and receptive language at age 4 years. However, these children were likely to catch up to their peers by age 7 years, which matches the age at assessment for participant 9. The influence of parent education and family disadvantage is somewhat clearer. Both higher maternal education and less social disadvantage are likely to be protective factors, which should support better language outcomes.

A measurable language difficulty could produce the pattern of results observed. For example, the term specific language impairment (SLI) describes a language difficulty occurring in the absence of other developmental deficits. Whilst the
use of this label has recently attracted controversy for its arbitrary and somewhat indiscriminate use, several reports exist of comparisons between the language skills of children with normal hearing (both with and without a SLI diagnosis) and children with mild-moderate hearing loss. These studies highlight similarities in language performance patterns between the hearing loss and SLI groups, suggesting the deficits observed in children with hearing loss (phonological short term memory, phonological discrimination and awareness) are similar to those seen in children with language impairment.\textsuperscript{166, 168} One challenge for considering a language impairment occurring for participant 9 is, despite the discrepancy between cognition and language ability that in isolation could lend itself to a label of SLI, the presence of hearing loss could be considered a "developmental deficit".

It is likely that auditory factors for participant 9 are valuable to understanding the poorer than expected language performance when considering cognitive potential. This participant had a PTA of 65 dB HL across four frequencies, the poorest of all participants and bordering a severe loss in the better hearing ear. Research is quite consistent in showing that increasing severity of hearing loss is associated with poorer speech and language outcomes.\textsuperscript{156, 157, 164} It has also been suggested that provision of amplification early, and consistent usage leads to improved outcomes compared to late fitting and less use.\textsuperscript{164} This participant was an early identified and fitted child, with up to 8 hours of hearing aid use reported per weekday in the period immediately prior to language assessment. Therefore, questions should be asked about the benefit provided by these hearing aids at the time of assessment. Participant 9’s calculated SII for loud speech only registered 26 points out of a possible 100, where 100 points represents full audibility. Given the evidence that over 50% of children fitted with hearing aids do not meet prescribed amplification targets,\textsuperscript{247} there is a strong likelihood that this participant was under-fitted with amplification at the time of assessment and vulnerable to lack of audibility during assessment. Unfortunately no further clues regarding audibility were available, as due to the severity of hearing loss, participant 9 did not complete speech discrimination in noise testing (the LiSN-S) that could have illustrated audibility deficits.
8.5.3 Aim 3

Hypothesis: 1-3 year old children with mild, but not moderate, hearing loss exposed to a well-established UNHS system would display mean expressive vocabulary development similar to children without hearing loss.

No evidence from this PhD supported this hypothesis. Both children born with mild and moderate loss displayed poorer mean expressive vocabulary and greater expressive vocabulary delay (defined by a negative vocabulary differential) than their peers without hearing loss at around age 2 years. At a sub-group level, children with mild loss had poorer vocabulary than those with moderate loss.

This hypothesis originated in part from literature suggesting children with hearing loss can achieve language outcomes similar to their normally hearing peers. This was reported early in life for both children with mild/minimal losses specifically\textsuperscript{175, 177} and in the range mild to severe,\textsuperscript{98} as well as into preschool aged children with mild and unilateral loss\textsuperscript{176} and even those ranging mild to profound.\textsuperscript{174} At best, such results could suggest that earlier identification and intervention allows children with all degrees of hearing loss to “keep up with” rather than “catch up to” children without hearing loss.\textsuperscript{174} Alternatively, children with losses of milder degrees may only start to lag in their language outcomes at later ages when environmental challenges and language learning demands increase.\textsuperscript{175} A third scenario, not evidenced in the published studies discussed so far, is that children with hearing loss already show delays in aspects of language development, even from an early age.

The results of the present study suggest that this third scenario of early measurable delays may be occurring. An early impact of hearing loss on expressive vocabulary is seen when data are collected at around 2 years of age. These observed results are in agreement with those reported from larger samples. From the OCHL study, at the earliest data collection age of 2 years those participants with mild loss were close to two thirds of a standard deviation lower in language scores than same-aged children with normal hearing.\textsuperscript{164}
Children with moderate loss were more than one standard deviation below their peers with normal hearing. The same trend occurred in an Australian cohort of children with hearing loss, who at age three years showed delays in language acquisition at all severity levels, but systematically increasing across the hearing loss spectrum from mild to profound.\textsuperscript{25} Whilst the present study used a single measure of vocabulary at age 2, the reported findings were similar to those from these larger studies\textsuperscript{25, 164} with more robust summary outcomes comprising a range of speech and language outcomes.

Study design limitations highlighted in previous sections of this discussion are also of consideration with this younger population of children with hearing loss. For example, sample size is again a limitation when interpreting the results. The small number of children with hearing loss who had expressive vocabulary assessed within the same window of age as the representative sample of children meant that direct comparisons between the two groups were more challenging. This was an unavoidable limitation of different experimental designs of the two source cohorts. The children with hearing loss were recruited from a newly established longitudinal databank open to children of any age. The children without hearing loss were from a large cohort of children recruited if born within a specified period and residing in one of several geographic localities. These differences resulted in notable disparity between groups in both the number of children and the distribution of ages, ultimately eliminating the use of standard scores. However, given that the children with hearing loss were in fact older than those without hearing loss, it is striking that their raw vocabulary scores were worse. This is especially notable, as we would expect rapid growth in vocabulary between the ages of 2 and 3 years.\textsuperscript{117, 273} This finding does provide some evidence of a real difference between children with and without hearing loss at around 2 years of age, given the comparison of outcomes was made with children from the same population tested on the same measure.

Of note, this PhD’s population-sourced cohort of younger children with hearing loss comprised only those with bilateral loss. This is a distinguishing feature when compared to the literature noted prior, whereby the grouping of children
with two distinct phenotypes of hearing loss, mild bilateral and unilateral of any degree, into a minimal hearing loss group\textsuperscript{175-177} may be problematic. Some children will be listening and learning language with the benefit of one uncompromised ear, versus two mildly impaired ears. Grouping would imply that the distinct challenges these two groups of children face\textsuperscript{251} are comparable in the net effect on language; this is not necessarily so. Universal intervention protocols are not considered possible for children with minimal loss because of large individual variability in outcomes and performance in the existing literature.\textsuperscript{109} Therefore, the benefit of solely including bilateral losses in this study means a clearer picture of language development can be obtained for children with two affected ears.

A study recruitment process linked to early intervention program attendance or wearing of devices/clinical samples\textsuperscript{98, 174, 175, 177} could impact the heterogeneity of the outcomes seen. Such samples could display skewed performance, likely positive for successful attendees of early intervention programs, when compared to what might be expected from population-sourced studies. The children in this sample were sourced from a population-based study, without inclusion criteria requiring early intervention attendance or hearing device fitting and use. All had early identified bilateral losses of mild or moderate degree at both diagnosis and age of assessment and the majority of families had chosen to fit amplification. Results showed children with any hearing loss had poorer expressive vocabulary than children without loss, but the degree of delay was - surprisingly - greater for children with mild loss than those with moderate losses. Whilst a chance occurrence in a small sample size cannot be discounted, this result may suggest children with mild loss are not benefiting from early aiding in the manner observed for children with moderate loss. This result dovetails with those seen in this PhD’s 5-7 year old sample (Aim 1), where in the well-established UNHS cohort the benefit of earlier identification and amplification was only consistently measurable for children with moderate loss. This novel pattern of language performance across a narrow range of hearing loss has been discussed prior (see Section 8.5.1), but there are aspects unique to the 1-3 year old group that warrant mention.
Parents of children with hearing loss who reported their child as having a higher expressive vocabulary also reported fewer observations of their child having listening difficulties in commonly encountered environments and scenarios. Those parents observing greater listening difficulties reported a lower expressive vocabulary in their child. Having concerns about a child’s listening performance may influence patterns of hearing aid use. It is acknowledged that children with mild loss as a group tend to wear hearing aids less often than those children with greater degrees of loss. An explanation put forward for this is that parents may experience uncertainty around what benefit hearing aids may provide their child with mild loss. Parents may be requiring evidence they acknowledge (e.g. observing their child struggling to hear, or having fewer words than similar-aged children) before committing to greater hearing aid use. It is possible that between their diagnosis and the age at which expectations of spoken language start to increase, the parents of children with mild loss were unaware of the impact of their child’s diagnosis on language development. In turn, this may have contributed to less consistent amplification use for their child. The observed slight trend toward greater hearing aid use in children with lower expressive vocabularies, whilst not statistically significant, may be in response to parent concerns regarding poorer expressive performance within the age bracket where language development is expected.

The influence of others outside the family unit may also be a factor. The decision to ensure more consistent and longer hearing aid use may be instigated by professionals involved with the family, via monitoring of global child development. Input from such professionals may occur too late to be of maximum assistance to the child’s development. Ideally the family would be educated and supported on methods of assisting their child’s development from a very early age. However, there are recognised discrepancies in the access to early intervention for families with children with lesser degrees of loss versus severe and profound losses. These differences cover both the access to and frequency of intervention sessions, which are opportunities to educate families on the potential impact of hearing loss and evidence of benefits for amplification.
use.\textsuperscript{174} To a certain extent, this difference in service delivery across the hearing loss spectrum is a product of our limited knowledge of the impact milder losses have on children’s development and what benefits outcomes.

### 8.6 Population and clinical implications

There has been little debate for the early identification and aiding of moderate bilateral hearing loss. However, there has also been less research evidence showing benefits resulting from this intervention when compared to evidence for greater degrees of loss. This PhD provides support for early amplification of moderate loss, showing secular trends toward improving language outcomes over an approximate 20 year period during which mean detection age and age at hearing aid fitting has decreased. This in the face of a growing literature base that shows children with moderate loss continue to display poorer than expected language development.\textsuperscript{156, 157, 164}

The implications for mild bilateral hearing loss differ from those for moderate losses. Davis et al (1997)\textsuperscript{26} stated, in the debate around the role of UNHS in detecting congenital hearing loss, that “…at some point on the severity continuum, evidence in a number of domains becomes equivocal, open to multiple interpretation, poorly controlled, or just non-existent…”. Tellingly, this was written in response to the debate in the mid-1990s regarding what hearing level should be set as the cut-point for newborn hearing screening. The findings of this PhD uphold the concerns expressed by Davis et al (1997).\textsuperscript{26} Despite the rapid changes in clinical practice where mild losses detected by UNHS are virtually always aided in this population, there has been no consistent change in outcomes. When considering implications at a population level, this PhD provides no support for lowering newborn hearing screening cut-points.

The outcomes of this PhD specific to mild losses do not clearly flag specific changes to clinical practice around the management of mild bilateral losses. There remains a lack of certainty about the best approach to take to optimise outcomes in a manner that is acceptable by the child, parents and health professionals. From a clinician’s sense, the question of what is the right care is
often a challenge due to uncertainty of benefit or harm between individuals. 277 The inclination to attempt improving children’s outcomes by intervening is balanced by a desire to not cause undue burden, cost and even harm. The fear of doing too little rather than too much is recognised as a motivator but also a reflection of inner biases.278 Frank and frequent self-knowledge checking is a key component to improving the translation of the available evidence, rather than educated hunches, into clinical decisions.

With the lack of clear evidence about the best way forward, current clinical practice of providing amplification for mild losses on a case by case basis is likely appropriate. Such careful consideration of individual factors such as hearing loss configuration and magnitude of impairment within the mild decibel range, projected benefits of the amplified signal for the child, comorbidities and family readiness for amplification is already recommended in existing clinical guidelines.24 186 Where amplification is chosen, greater family support in evidence-based decision-making and monitoring of family motivation for ongoing amplification use will likely optimise any benefits that result from aiding mild losses. Alternatives to hearing aids, such as remote microphone systems, could also become an option for early diagnosed infants with mild losses, outside of scenarios where use is typically recommended (e.g. a classroom setting).186

Family-centered counseling or training on the listening challenges that children with mild hearing loss experience in routine interactions may provide additional information that is able to maximise hearing aid use. Influencing parent decision-making to persevere with amplification (i.e. maintaining daily use across all waking hours), despite no obvious outward signs of benefit may be crucial to maximising the child with hearing loss’ cumulative auditory experience. The early intervention space and family counseling is poorly researched for children with mild loss.188 Early education and intervention may play an important role for these families, particularly if all amplification options have been exhausted or if amplification in general is unable to improve outcomes for this population (see Section 8.5.1.4.2).
8.7 Conclusions and future directions

The population-level findings of this PhD suggest that, both across the two age groups and across time, children born with mild bilateral hearing loss are not experiencing the clear benefits to language development that children with moderate loss experience. This is despite both groups of children now being detected and fitted with hearing aids earlier than ever before. Seeking conclusive evidence in the arena of how best to support the outcomes of children with mild bilateral hearing loss presents many challenges. Yet it is an important research area to address. Under current detection practices, large numbers of children with mild bilateral hearing loss will continue to be identified early. Children with mild loss account for approximately 30% of all impacted individuals.\textsuperscript{54,87} This indicates that the present gaps in the evidence relate to a clinically significant proportion of children.

Carefully constructed and controlled trials are needed to compare the systematic provision of hearing aids versus no hearing aids for children with mild bilateral hearing loss. The most important aspect of this trial would be providing support to maximise hearing aid use within the intervention group. Integral to this would be an accurate method to measure usage, quality of the fit, and examination of outcomes from both an unaided and aided hearing acuity perspective. These outcomes would by necessity cover multiple domains, including both specific skills and broad language outcomes, as well as measures of costs and health-related quality of life. An adequate sample size would facilitate exploring both the effect of hearing threshold within the mild range and the amount of time well-fitted devices are used that lead to observable benefits, quantified via the use of appropriately sensitive measures. Monitoring outcomes from an early age would inform decision-making on cost-effectiveness of systems designed to optimise management for children with congenital hearing loss based on need and benefit, without the creation of undue costs and harm.
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10. Appendices

Appendix A: VicCHILD parent information statement and consent form

The VicCHILD project aims to advance research about childhood hearing loss. It started in 2011 and is based at The Royal Children’s Hospital, Melbourne. We hope it will help us understand what leads to the best outcomes for these children. In turn, this may lead to better counseling and treatment.

VicCHILD's full name is the ‘Victorian Childhood Hearing Impairment Longitudinal Database’. It is a databank for children born with permanent hearing loss. A databank is a special kind of research database. It keeps collecting children indefinitely, and stores their information confidentially for many years.

Databanks have special advantages. They can:
- Answer questions that need very large numbers of participants.
- Answer new research questions quickly using the stored information.
- Show how children’s outcomes change over time, as new treatments and approaches develop.
- Find all children who are eligible for new research projects, so they have the chance to take part.

VicCHILD pulls together different kinds of information into a single databank. Most of this information is already collected – we just need your permission to access it. Its value comes from joining it up in new ways. This means that VicCHILD doesn’t need to make big demands on your or your child.

You may choose to provide some or all of the information listed in Section 4 and 5 to VicCHILD. VicCHILD is open to children with a permanent hearing loss from birth or soon after. Around 60 to 100 babies are joining the databank each year. Later, it may expand to other Australian states and territories. Some other countries are also interested in joining.
2. Who is funding this research project?
The Murdoch Childrens Research Institute (MCRI) and the Centre for Community Child Health at the Royal Children's Hospital.

3. Why are we being asked to be in this research project?
The Victorian Infant Hearing Screening Program (VIHSP) has passed on your information as your child was identified has having a hearing impairment.

4. What do we need to do to be in this research project?
VicCHILD would like to contact you every 5-7 years. We hope to re-contact families around the start and end of primary school, and around the end of high school.

The VicCHILD project is made up of parts – you may want your child to be in all parts, or only some parts. It collects some information directly from families. It also gathers information from other sources, with your permission. This means we collect accurate information without a great deal of your time.

To take part in the VicCHILD project, your child will need to complete:
- A questionnaire every 5-7 years, once he/she is old enough. It is short, taking around 15-30 minutes. It asks about your child's own well-being and quality of life. For younger children a VicCHILD researcher may help. Older children can fill out the questionnaires themselves.
- An assessment of language and learning, starting from age 5-7 years. We aim for a VicCHILD researcher to assess your child at ages 5-7 years, 10-12 years and 15-17 years. This depends on VicCHILD's resources being available to do the assessment. It takes about 1-1.5 hours, depending on your child's abilities. We use standard tasks suited to the child's age. For example, we show pictures and ask questions, then listen to their answers and their speech. The tasks are short and varied, and most children enjoy them greatly. If you agree, we may audio- or video-record parts of the assessment to help with scoring and the recordings also become part of the database.

To take part in VicCHILD, you will need to complete:
- A questionnaire mailed to you every 5-7 years. This takes about 15-30 minutes to fill out. You can post it to us, or we might collect it when we visit. It asks about your child, and about you and your family.
  - About your child: We ask about your child's health, quality of life, language and learning. We also ask about the services your child uses and your experiences with these services.
  - About you and your family: We ask about your family history and demographics (where you live, your current work etc.). We also ask about your own health and feelings, and how your child's hearing loss has affected your family.
- A 1-1.5 hour home visit soon after each questionnaire: We make a time for a researcher to visit you at home. If you prefer, the visit could be somewhere convenient to you (like childcare or school), or you can come to the Royal Children's Hospital. At the visit:
  - The researcher goes through any consent forms needed at that time, to make sure you understand about each part of VicCHILD. You can ask questions that you may not have thought of at the first phone call. Then you can consent to the aspects of VicCHILD you want to take part in.
  - We may ask you to complete a second short questionnaire. This covers your child's development, and more detailed questions about the services your child uses and their costs to you. The researcher can help you, or you can complete it by yourself. The researcher sees your child (see section above).
  - If a visit isn't possible, we can interview you by phone instead. However, it's not possible to collect all of the information over the phone.

5. What are other ways I can take part in VicCHILD?
You can give permission for your child to take part in some or all parts of VicCHILD. The following parts of VicCHILD are optional. They don't require you to do anything. With your permission VicCHILD will carry them out on your behalf.
We would like to get your permission for VicCHILD to:

A. Link your child’s information with other existing datasets (“data linkage/data retrieval”)

As Australian children grow up, services and agencies collect and store information. It’s used for purposes like budgets and reports about Australians’ health and education. It is carefully protected and confidential.

You can give permission for VicCHILD to access some of the information stored about your child. It may relate to your child’s hearing loss, its diagnosis and cause; devices to assist hearing; the services children use; their progress; and demographic information.

Below, we list the main organisations that store these data. Some of these are only available as your child gets older. Also, most children don’t have information with every organisation.

- **Organisations:**
  - Victorian Perinatal Data Collection, Victorian Birth Defects Register, Victorian Infant Hearing Screening Program (VIHSP), Medicare/PBS (Pharmaceutical Benefits Scheme), Australian Hearing, Hospital Admissions data, Royal Victorian Eye and Ear Hospital records & Cochlear Implant Clinic records

- **Sources of data:**
  - School Entry Health Questionnaire, Australian Early Development Index, National Assessment Program – Language and Literacy (NAPLAN), Victorian Certificate of Education, Victorian Certificate of Applied Learning (VCAL)

Over time there may be other databases that could help hearing research. We are also asking your permission to access such information. This would also need approval from the Ethics Committee.

We retrieve information in one of two ways. This depends on the organisation that holds the data.

- **Confidential data retrieval:** VicCHILD sends a request to the organisation. The request includes your child’s name, birth date, or other details needed to identify your child’s records. The organisation then gives the information directly and confidentially to the VicCHILD researchers.

- **Re-identifiable linkage:** Linkage is done confidentially by VicCHILD researchers, or by ethically-approved data linkage organisations like Bioqip or the Victorian Data Linkage Unit. Your child’s personal information is stored separately and given a unique linkage code. This code allows linkage and updates of your child’s data, as more information is collected. Only the research team has access to this code.

B. Access and collect biological samples from you and/or your child

We would like to access two samples from your child.

- Your child’s newborn screening (“ Guthrie”) card. This dried blood sample was collected from your child’s heel soon after birth. It is stored by the Victorian Clinical Genetics Service at the Murdoch Childrens Research Institute. It can be studied for factors that might influence hearing, like viral infections during pregnancy. It can also be used for genetic analysis.

- A genetic sample via a buccal (cheek) swab or saliva sample. This lets us study genetic causes of hearing loss. These samples are securely stored in locked freezers at the Biochemistry Faculty of the Murdoch Childrens Research Institute for the duration of VicCHILD. Collecting the sample is very simple—it doesn’t hurt at all and takes about a minute to do.

We would also like to access Mother’s prenatal test, if available.

- Most mothers have a blood test when they are 10-12 weeks pregnant, to screen for conditions like Down Syndrome. The serum is currently stored by the Victorian Clinical Genetics Service at the Murdoch Childrens Research Institute. The baby’s mother can give permission to access it. It could address factors very early in pregnancy that might influence how hearing develops.

C. Exchange of information

Sometimes, two agencies may ask to see your child around the same time. For instance, we might contact you soon after a school entry assessment. Or a secondary school might request results from the VicCHILD 10-12 year old assessment. To avoid multiple assessments, you can let us exchange results with other professionals (like speech pathologists, audiologists, and teachers). This might be by letter or phone.

D. Re-contact you for new research projects

Over time, knowledge grows. This can lead to new research questions, and perhaps new research
projects of interest to you. This might involve new technologies and/or new treatments, beyond the information you already have from your child. You could find out more and then decide whether you wanted to take part. You would then decide whether or not to consent to any new projects.

The project might be with VicCHILD researchers or with other research teams. If the new research was with a different research team, we would send you an invitation letter on their behalf. We only pass on your contact information if you agree to this.

E. Use your child’s VicCHILD data for future/research projects (data sharing)

VicCHILD is a resource for researchers around Australia and internationally. They can request access to data to answer different research questions. This ensures that your valuable information is used to the full. Any such requests need to be approved by the VicCHILD team, and the research must be ethically approved. Researchers can access only the data items they need from VicCHILD. When they use your child’s VicCHILD data, they would not be able to identify you or your child.

We would also like to be able to join your VicCHILD information with larger data repositories to answer questions about language or hearing. For example, Professor Melissa Wake, VicCHILD’s Chief Investigator, is also a lead researcher in the MRCI’s “Centre of Research Excellence in Children’s Language” (CRE-CL). Families from several of the MRCI’s large language studies have agreed that their data be added to a single large database for the CRE-CL. This makes a much bigger dataset containing children’s language assessments than any one project could manage by itself. We hope this will lead to major new discoveries about how children learn language, with or without hearing loss. To do this, we need to share some identifying information with the CRE-CL administrators. However, this does not go in the dataset, so researchers using your data are not able to identify you or your child.

6. What are our alternatives to taking part in this project?

You don’t have to take part if you don’t want to. If you consent and then change your mind, you can withdraw at any stage. This won’t affect any care or treatment your child receives from The Royal Children’s Hospital.

If you decide to withdraw, we would like to keep the information collected up to that point. However, if you don’t want us to keep the information, you can tell us when you withdraw. Then we would remove your child’s information and destroy it.

7. What are the possible benefits for my child?

Your child may not directly benefit from taking part in VicCHILD. However, in the longer term, the project may lead to better treatment for children with hearing impairments. This might benefit your child.

If we assess your child’s language, you can ask us for a short report of the results.

We may offer you or your child the chance to be in other research projects. These may or may not benefit your child. You would choose at the time whether to join any extra projects we tell you about.

8. What are the benefits for other people in the future?

Knowledge from VicCHILD may help other children with hearing loss in the future. This might include better diagnosis, counselling, treatment or interventions. These could improve children’s speech and language, hearing, and well-being.

9. What are the possible risks, side effects and/or discomforts?

We don’t expect any harm to you or your child. Answering questionnaires may cause you to think of new questions about your child’s hearing condition. You might not have thought of these before. If so, we encourage you to contact your GP, paediatrician or ENT specialist. You can also contact VicCHILD.

There is always a risk to privacy when keeping a database, particularly with ongoing data storage. However, we have strict privacy and confidentiality policies to minimize this risk. If you have any questions or concerns about the VicCHILD project, please contact us at any time. Our phone number and email are at the end of this statement.

10. What are the possible inconveniences?

The main inconvenience is to your time. Each parent questionnaire takes about 15-30 minutes. A first visit takes about 30 minutes. Visits to assess your child take around 1.5 hours. We hope these visits and questionnaires will happen every 5-7 years (this does depend on ongoing funding).
11. How do I know my child's information is confidential?

All the information you give us is private. We can only disclose it according to the consent that you give us, except as required by law. In accordance with Australian and/or Victorian privacy and other relevant laws, you have the right to access and correct the information we collect and store about you. Please contact us if you would like to access your information.

Information collected by VicCHILD is re-identifiable. This means we remove your names and use a special code number instead whenever your child’s information is analysed. That way, your child’s identity is always protected. However, VicCHILD can still go back and match your names to your VicCHILD code number for administrative purposes, if needed.

Questionnaires are stored in locked filing cabinets at the Murdoch Childrens Research Institute or a secure storage facility. Genetic samples are stored in locked freezers at the Biorepository Facility of the Murdoch Childrens Research Institute. All information is stored on a password-protected computer database.

We plan to present results at conferences and in professional journals. We only report information for groups of children—no one will be able to identify you, your child or your family.

12. Will we be informed of the results?

VicCHILD is building up slowly, as children are born and join the project. We can begin to study the information once the database has been running for 2-3 years. We mail a newsletter each year to families. We also have a VicCHILD webpage. It contains information about VicCHILD and in time will also hold newsletters and links to research publications.

We won’t routinely send you results of your child’s language and learning assessments. However, you can ask us for a short written report if you would like one.

13. Who are the researchers?

Professor Melissa Wake, a paediatrician, is Director of Research at the Centre for Community Child Health. Dr Zofia Poulakia, a psychologist, is the Director of the Victorian Infant Screening Program (VISP).

Dr Kathryn Mueller, a research officer in the Hearing, Language and Literacy Group at the MRCI.

Dr Rachel Burt, a senior research fellow at the MRCI, is group leader of the Molecular Hearing Group.

Professor Jane Halliday, an epidemiologist, is head of the Public Health Genetics Group at the MRCI.

Mr Luke Stevens is the Data Management Coordinator at the MRCI.

Ms Sherriyn Tobin, a provisional psychologist, VicCHILD’s senior research coordinator.

(Over time, the research team may change, as VicCHILD and its participants grow.)

If you would like more information about the project or if you need to speak to a member of the research team in an emergency please contact:

Names: Sherriyn Tobin
Contact telephone: (03) 9345 4215
Email: vic-child@rch.org.au
Website: http://www.rch.org.au/ccch/for_researchers/VicCHILD/

If you have any concerns about the project or the way it is being conducted, and would like to speak to someone independent of the project, please contact:

Director, Ethics & Research, The Royal Children’s Hospital on telephone: (03) 9345 5044.
CONSENT FORM

HREC Project Number: 31081
Research Project Title: Victorian Childhood Hearing Impairment Longitudinal Databank (VicCHILD)
Version Number: 4  
Version Date: 30 August 2013

- I voluntarily consent for my child and I to take part in this research project.
- I believe I understand the purpose, extent and possible effects of my child's and my involvement in this project.
- I have had an opportunity to ask questions and I am satisfied with the answers I have received.
- I understand that this project has been approved by The Royal Children’s Hospital Human Research Ethics Committee and is carried out in line with the National Statement on Ethical Conduct in Human Research (2007).
- I understand I will keep this Parent/Guardian Information Statement and one copy of the Consent Form.

Child's Name

Parent/Guardian Name  Parent/Guardian Signature  Date

Note: All parties signing a Consent Form must date their own signature

### ADDITIONAL CONSENT

<table>
<thead>
<tr>
<th></th>
<th>YES</th>
<th>NO</th>
</tr>
</thead>
<tbody>
<tr>
<td>A. Linking your child’s information with other existing datasets (data linkage)</td>
<td>YES, I do consent</td>
<td>NO, I do not consent</td>
</tr>
<tr>
<td>B. Biological samples</td>
<td>Access to newborn screening card</td>
<td>YES, I do consent</td>
</tr>
<tr>
<td></td>
<td>Cheek swab or saliva sample</td>
<td>YES, I do consent</td>
</tr>
<tr>
<td></td>
<td>Access to prenatal maternal serum</td>
<td>YES, I do consent</td>
</tr>
<tr>
<td>C. Exchange of information</td>
<td>YES, I do consent</td>
<td>NO, I do not consent</td>
</tr>
<tr>
<td>D. Re-contact for research purposes</td>
<td>YES, I do consent</td>
<td>NO, I do not consent</td>
</tr>
<tr>
<td>E. Using your child’s VicCHILD data for future/other research projects (data sharing)</td>
<td>YES, I do consent</td>
<td>NO, I do not consent</td>
</tr>
</tbody>
</table>
Appendix B: VicCHILD Technical Protocol

(Royal Children’s Hospital Human Research Ethics Committee reference number 31031)

VICTORIAN CHILDHOOD HEARING IMPAIRMENT LONGITUDINAL DATABANK

VicCHILD

RESEARCH PROTOCOL

VERSION 14. 5th March 2015

CONFIDENTIAL

This document is confidential and the property of the VicCHILD Team at the Centre for Community Child Health.

No part of it may be transmitted, reproduced, published, or used without prior written authorisation from the institution.

STATEMENT OF COMPLIANCE

This document is a protocol for a population-based, condition-specific longitudinal database. The study will be conducted in compliance with all stipulations of this protocol, the conditions of ethics committee approval, the NHMRC National Statement on Ethical Conduct in Human Research (2007) and the Note for Guidance on Good Clinical Practice (CPMP/ICH-135/95).
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# Protocol Synopsis

<table>
<thead>
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<th>Title</th>
<th>The Victorian Childhood Hearing Impairment Longitudinal Databank (VicCHILD)</th>
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| **Objectives** | 1. To establish VicCHILD as a world-first population-based databank for children with congenital hearing impairment.  
2. To facilitate population-based research that will:  
   • Describe secular trends in outcomes;  
   • Support population-based quality improvement activities;  
   • Identify and quantify factors that predict outcomes; and  
   • Facilitate randomised controlled trials of interventions. |
| **Design** | Longitudinal databank. |
| **Project Duration** | Collection of data on individual participants will be ongoing.  
Although this application covers only the first 5 years of the project, we envisage that the VicCHILD databank itself will:  
   • Recruit newly-diagnosed babies into the future, assuming sufficient funding for minimum activities.  
   • Remain in existence and useable indefinitely. |
| **Population** | 1. All children will be invited to participate at 3-6 months of age who:  
   • Are born in the state of Victoria, Australia, from January 2011 onwards  
   • Have bilateral or unilateral congenital hearing impairment  
   • Are recorded by the Victorian Infant Hearing Screening Program.  
Assuming a 70% uptake, we expect approximately 80 Victorian babies to join VicCHILD annually (see Section 4.2 of this Protocol).  
2. In one-off retrospective recruitments, we will also invite the following to contribute their data retrospectively, then participate in ongoing data collection:  
   • 2011: recruitment of 157 participants (based on 70% uptake) from two recently-completed population-based studies (CHIVOS and SCOUT).  
   • 2012-3: all eligible children born Mar 2005- Dec 2010, i.e., during the statewide roll-out of VIHSP’s universal newborn screening activities. |
| **Outcomes** | 1. An established population-based databank for hearing-impaired children.  
2. Greater understanding of developmental, health and well-being outcomes for hearing-impaired children and how to improve them.  
3. Potential for new partnerships/capacity for population-based research into:  
   • Congenital hearing impairment in Victoria and other states/countries  
   • Other conditions detected by screening before or soon after birth. |
GLOSSARY OF ABBREVIATIONS

<table>
<thead>
<tr>
<th>ABBREVIATION</th>
<th>TERM</th>
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<tbody>
<tr>
<td>AEDI</td>
<td>Australian Early Development Index</td>
</tr>
<tr>
<td>CCCH</td>
<td>Centre for Community Child Health</td>
</tr>
<tr>
<td>CEBU</td>
<td>Clinical Epidemiology &amp; Biostatistics Unit</td>
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<tr>
<td>CHIVOS</td>
<td>Children with Hearing Impairment in Victoria Outcomes Study</td>
</tr>
<tr>
<td>CMV</td>
<td>Cytomegalovirus</td>
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<tr>
<td>dBHL</td>
<td>Decibel Hearing Level</td>
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<tr>
<td>FAHL</td>
<td>Frequency Average Hearing Loss</td>
</tr>
<tr>
<td>MCRRI</td>
<td>Murdoch Children's Research Institute</td>
</tr>
<tr>
<td>NAPLAN</td>
<td>National Assessment Program - Literacy and Numeracy</td>
</tr>
<tr>
<td>PTA</td>
<td>Pure Tone Audiogram</td>
</tr>
<tr>
<td>RCH</td>
<td>Royal Children's Hospital</td>
</tr>
<tr>
<td>SCOUT</td>
<td>Statewide Comparison of Outcomes of Hearing Loss</td>
</tr>
<tr>
<td>VDL</td>
<td>Victorian Data Linkages (Victoria Department of Health)</td>
</tr>
<tr>
<td>VIHSP</td>
<td>Victorian Infant Hearing Screening Program</td>
</tr>
</tbody>
</table>

1. INVESTIGATORS AND FACILITIES

1.1 Study Location/s

Centre for Community Child Health
Level 2 East, Royal Children's Hospital
Flemington Road,
PARKVILLE VIC 3052

Murdoch Children's Research Institute
Level 5, Royal Children's Hospital
Flemington Road,
PARKVILLE VIC 3052

1.2 Study Management

1.2.1 Principal Investigator
Prof. Melissa Wake
Associate Director and Director of Research
Centre for Community Child Health
(Ph) (03) 9345 5937
(Fax) (03) 9345 5900

Prof. Wake is well-placed to lead the Operations Team of VicCHILD at the Royal Children's Hospital, given her extensive experience in hearing-related public health research and in population-based longitudinal and intervention child health research more generally. Her role will be to set out the strategy for the establishment of the databank, ensure adequate resources and personnel are allocated, offer guidance on the various issues that potentially
will arise, publicise the databank and its potential to other research groups, and lead and/or contribute to dissemination activities (e.g., publications, presentations).

1.2.2 Associate Researchers

Dr Zefi Poulakis
Director - Victorian Infant Hearing Screening Program (VIHSP)
Research Officer - Hearing, Language and Literacy Stream, Healthy Development Theme, Murdoch Childrens Research Institute
Centre for Community Child Health

As Director of VIHSP, Dr Poulakis has an in-depth understanding of the statewide screening program from which Victoria's will spring, and is well-connected to the many professional groups that may contribute to and/or use the data from the databank. She has extensive knowledge of the epidemiology and outcomes of congenital hearing impairment and nearly 20 years of experience in population-based research into this condition. Her Doctorate in Clinical Psychology and experience as a clinical psychology group leader will specifically enrich Victoria's research into psychological predictors and outcomes for children who are deaf and hard of hearing.

Associate Professor Jane Halliday
Group leader
Public Health Genetics
Murdoch Childrens Research Institute

The Public Health Genetics group at the MCRI uses epidemiological methods to investigate the prevalence of and aetiology of birth defects, evaluate genetic health services utilisation, assess consumer interest in, and understanding of, genetic testing, and examine the health and well-being of infants, children, and young adults following technological interventions during pregnancy and conception. As leader of this group, Professor Halliday will be well-placed to provide a wealth of epidemiological research advice to the Victoria team.

Mr Luke Stevens
Data Management Coordinator,
Clinical Epidemiology & Biostatistics Unit
Murdoch Childrens Research Institute

Mr Stevens links the MCRI's statistical and IT arms and is the custodian for REDCap, the secure web-based application designed to support data capture that will underpin Victoria's data management. He will provide technical advice and support for the setup and management of the databank.

Dr Rachel Burt
Postdoctoral Research Fellow,
Hearing, Language and Literacy Stream, Population health genes and environment
Murdoch Childrens Research Institute

Dr Burt is a group leader and senior research fellow in the Molecular Hearing group at MCRI. In this project, both Dr Burt and Dr Mueller will:
• Advise on coordination of sample collection, processing and storage using already-validated processing protocols and state-of-the-art 2D sample tracking system and associated database

• Provide expert advice on research questions related to the examination of genetic and/or epigenetic variation in future analyses

• Oversee the design and validation of genetic and epigenetic assays for specific candidate genes of interest

• Oversee the statistical analysis of generated data using biostatistical expertise recently developed within MCRI for such analyses

Ms Sherryn Tobin  
VicCHILD Project Manager  
Community Health Services Research, Population Health Theme  
Murdoch Childrens Research Institute

Ms Tobin is a practicing Psychologist and has worked as a Senior Research Assistant for over 13 years, with specific experience on projects relating to early hearing (HISS, CHIVOS, SCOUT and VIHSP) and language (Let’s Learn Language and Language for Learning). Ms Tobin will manage the day to day running of the project under supervision of the research team.

Mr Peter Carew  
PhD student  
Community Health Services Research, Population Health Theme  
Murdoch Childrens Research Institute

Mr Carew is a paediatric audiologist with 10 years’ experience in clinical, public health and research settings. He is currently completing his PhD on a subgroup of VicCHILD participants and brings his audiological skills to the project.

All: Royal Children’s Hospital & Murdoch Childrens Research Institute  
Flemington Road  
Parkville VIC 3052

1.3 Funding and resources

A variety of sources have supported VicCHILD. An MCRI Healthy Development Theme internal seed grant of $35,000 allows for establishment of the VicCHILD databank structure and operational processes, supplemented by internal Centre for Community Child Health funding throughout 2011. $30,000 was obtained from the Phyllis Connor Philanthropic Trust in 2012. Since 2013 the MCRI’s CRE in Child Language has part-supported project staff. In 2014 the Cooperative Research Centres (CRC) program awarded a five year extension to the HEARing CRC, with some funding flowing to the Murdoch Childrens Research Institute, from which a portion was allotted to the VicCHILD databank.
Additional funding applications will be made in 2014/15.

We anticipate funding applications to:

- The National Health and Medical Research Council
- The Australian Research Council Linkage Grants scheme
- Philanthropic bodies such as the Deafness Foundation
- The US National Institutes of Health (e.g., National Institute of Deafness & Communication Disorders)
- International collaborative funds (e.g., NHMRC-EU Collaborative Research Grant scheme).

2. BACKGROUND AND RATIONALE

One in 800 children in Australia is born with a permanent bilateral childhood hearing impairment of moderate or greater degree (>40dB HL in the better ear), and many more have milder and/or unilateral losses. Moderate or greater hearing losses have significant impacts on a child’s speech, language and general development, incurring lifelong social, educational and economic costs, with a cost to society of more than $1.5 million per child. The proposed data repository will be a resource for systematic, coordinated, population-based prospective research to improve these outcomes.

The impact of hearing screening

Recent advances in hearing screening make this new repository both necessary and feasible.

Universal Newborn Hearing Screening (UNHS) programs now identify children affected by hearing impairments much sooner after birth compared to earlier methods such as distraction hearing tests which are generally offered and conducted when infants are around nine months of age. Research published throughout the 1990s strongly suggested that very early detection (<6 months of age) of hearing loss led to normal language and developmental outcomes that were similar to those of hearing children. This formed the platform from which national UNHS programs have been implemented throughout the world.

However, this optimism has been tempered by subsequent evidence arising from epidemiological approaches and stronger research designs. When compared with risk factor screening and/or distraction screening techniques, universal newborn hearing screening is followed by much earlier detection, amplification, and entry to early intervention. Mean language outcomes also improve, but the effect size is much less than desired and, as of 2010, it is clear that they remain below the mean values for hearing populations throughout the preschool and early primary school years (e.g., Kuever et al., 2010, Kennedy et al., 2006 NEJM).

Therefore, UNHS programs are both a stimulus (because so much remains to be achieved to normalise outcomes) and a resource (because they detect and track all children with congenital hearing loss in whole populations) for research capitalising on detection that is occurring earlier than ever before.

The Victorian Infant Hearing Screening Program (VIHSP) is the statewide newborn hearing screening program responsible for testing the hearing of all newborn babies born in Victoria. The testing occurs during their time in hospital after their birth, or at an outpatient appointment within the first weeks of life. The VIHSP NHS service is expected to be fully implemented in
all maternal hospitals by early 2011, providing the rationale and recruitment mechanism for this proposal.

Understanding hearing impairment outcomes

Previous research initiatives were established at CCCH to better understand the outcomes of children living with hearing impairments. Two studies, the Statewide Comparison of Outcomes and Hearing Loss (SCOUT) and the Children with Hearing Impairment in Victoria Outcomes Study (CHIVOS), examined language and developmental outcomes in children with hearing impairments. These studies offer both a secondary means of recruitment for a statewide databank for children with hearing impairments, as well as a source of data to link to.

Statewide Comparison of Outcomes and Hearing Loss (SCOUT): The SCOUT study aimed to investigate the effectiveness and cost-effectiveness of universal newborn hearing screening. SCOUT compares population outcomes at age five years in children with congenital bilateral permanent hearing loss born between March 2003 and February 2005 in New South Wales or Victoria. At that time, NSW offered universal newborn hearing screening to all babies (UNHS program) while Victoria screened only babies at risk (risk-factor program). Children included in SCOUT have bilateral congenital hearing loss, and were fitted with hearing aids or cochlear implants before their fourth birthday. All eligible children in both states were approached to undergo assessments of their language, non-verbal cognition, vocabulary and speech intelligibility at age 5 years, supplemented by parent and service data.

Children with Hearing Impairment in Victoria Outcomes Study (CHIVOS): CHIVOS is an ongoing longitudinal, population-based study of 88 Victorian children with mild to profound congenital hearing loss. Starting in 1999, CHIVOS children have been seen at age 1-5, 12-14 and 17-19 years of age (anticipated 90% retention). At each wave, participants have undergone direct assessments of language, non-verbal cognition and academic achievement, supplemented by parent- and child-reported data.

Advancing hearing impairment outcomes

While NHS and early intervention programs improve speech and language outcomes, it has become evident that other factors may play a part in the poorer (although variable) outcomes in hearing-impaired children. Despite the research conducted in the CCCH, our knowledge of these factors and their relative effects on outcomes is inadequate, as information regarding childhood hearing loss is hampered by a lack of population-based prospective research, as articulated by the US National Institute of Deafness & Communication Disorders (NIDCD 2006). With the introduction of VICHSP, and the existence of other comprehensive population databases (including healthcare, other congenital problems and academic outcomes data), Victoria has the opportunity to create an important repository to advance outcomes research at a population level.

Unraveling the genetics through identification of biomarkers

Population data repositories (termed ‘biobanks’) when they include biological samples can foster innovative research and treatment breakthroughs for conditions in which complex interactions of genetic, molecular, disease and social/environmental susceptibility may impact upon disease risk or treatment. Condition-specific data repositories are typically smaller than whole-of-population biobanks (such as the Western Australia Genome Project and the UK Biobank) because effect sizes are larger. One of the key strengths of VicCHILD will be the availability of biospecimens for the identification of biomarkers associated with early hearing loss. Further, VicCHILD will support longitudinal and genetic discovery research, while also monitoring secular trends in outcomes. It responds to the 2005 National Institute of Deafness & Communication Disorders’ call for the development of data repositories for the research
community... [and] funding mechanisms... for secondary analyses of existing data' (NICDC, 2006).

Better understanding of the environmental and/or genetic causes of congenital hearing loss will result in better and targeted treatment, as well as improved prognosis and counselling options. Environmental risk factors include premature birth, ototoxic drugs and infections (in particular, congenital cytomegalovirus (CMV) infection) (Joint Committee on Infant Hearing, 2007). As part of VicCHILD, we hope to collect maternal serum in the first trimester of pregnancy, allowing measurement of many different metabolites and infectious agents, including CMV viral load. Limited previous studies have suggested that children with congenital CMV infection following first trimester maternal infection are more likely to have CNS sequelae, especially sensorineural hearing loss (Pass, 2008). We will be perfectly placed to test this finding in a large cohort.

Genetic factors contribute to approximately 50% of all hearing loss (Trenholme, 2000). Mutations in more than 50 identified genes can cause non-syndromic deafness - many more cause syndromic deafness. Unravelling the genetics of hearing loss, however, remains challenging due to large heterogeneity and classification difficulties. Within the next 1-5 years, gene enrichment techniques and next-generation DNA sequencing will allow a detailed and cost-effective sequence analysis of all known deafness genes, if not all genes in a human DNA sample. The proposed VicCHILD database will be poised to support such comprehensive genetic studies, both retrospectively and prospectively, on whole populations of children with hearing impairment.

In recent years there has been intense interest in the likely role of epigenetic disruption as a major mediator of cumulative risk associated with gene-environment interactions in complex human diseases. Epigenetic mechanisms, known to be sensitive to environmental perturbation, encompass a range of regulatory modifications and factors that modify the activity state of underlying DNA in the absence of changes to primary DNA sequence (Bird, 2007). This includes the regulation of all gene expression, with differences in overall epigenetic and associated gene expression, profiles accounting for the wide range of different cell and tissue types in our bodies, each of which contain essentially the same DNA sequence. A role for epigenetic disruption in hearing development and loss is supported by several lines of evidence (Provenzano, 2007) including studies examining phenotypic differences of monozygotic twins (containing identical genetic information). In one study involving twins with Usher syndrome, one sibling presented with a much more severe phenotype than the other sibling. The authors posited a possible epigenetic mechanism for the differences in gene expression (Ellestad, 1997). Epigenetic analysis of biospecimens collected in VicCHILD therefore offers vast opportunities to increase our understanding of hearing and hearing loss.

Setting population research precedents

We believe that this would be the first in the world; neither the US nor the UK has the supporting intervention or population outcomes databases essential to such an initiative. This longitudinal database would allow us to monitor cross-sectional outcomes over time, undertake longitudinal research and ultimately lead to better intervention strategies for hearing-impaired children. Databases can provide the opportunity for collaboration of research across varying disciplines and organisations statewide, as well as facilitate relationships both state- and worldwide.

Longitudinal research will provide information about how the children's outcomes (such as language development) change with age (in comparison to normative data), as well as how trends in outcomes change over years — this will allow examination of how different services and interventions might improve outcomes for hearing-impaired children at a population level. MCRI researchers have the expertise from successful outcomes studies, for which we have...
achieved high participation rates, as well as expertise in biobanking and biomarker discovery. Victoria is placed in a unique situation from which it is possible to engage in longitudinal studies aimed at identifying factors that influence outcomes in hearing-impaired children and young adults. Victoria has standardised tests of hearing, early development and academic performance that enable effective comparisons to be made. If we can access and coordinate information from 1) existing resources, 2) new genetic studies and 3) a standardised protocol for assessment of speech and language in deaf children, we will have unrivalled possibilities for data linkage, with which to investigate factors that affect outcomes.

A databank that incorporates (1) data from existing databases, (2) biological markers and (3) tailored standardised preciotor and outcomes information directly from the participants themselves will be able to address many different research questions, including questions that are as yet not articulated. Therefore, we do not specify any one hypothesis or research question here. Examples of research questions that could be addressed by the databank include:

- What factors (genetic and environmental) best explain the great variability in language outcomes for children with any given severity or type of hearing impairment?
- Has the incidence of prenatal CMV changed over time and, if so, is this reflected in changes in the epidemiology of congenital hearing impairment?
- Does amplification for mild congenital hearing loss improve outcomes?
- Does amplification for unilateral hearing loss improve outcomes?

3. STUDY AIMS AND OBJECTIVES

3.1 Aim

Our primary aim in the 5-year period 2011-2015 is to successfully establish VicCHILD as a world-first population-based databank for hearing-impaired children.

3.2 Objectives

Our overall objective is to provide a new resource for efficient and flexible population-based research into congenital hearing impairment, available to researchers across disciplines. Ultimately, we expect VicCHILD to lead to better intervention and outcomes for hearing-impaired children, flowing on to greater societal and economic participation in adulthood.

Our specific objectives are to facilitate population-based research that will:
- Describe secular trends in outcomes over time for repeated birth cohorts;
- Support population-based quality improvement activities over time;
- Identify and quantify factors that predict outcomes; and
- Facilitate randomised controlled trials of new management and treatment approaches.

3.3 Secondary Objectives

Harmonising state, national and international longitudinal datasets could greatly enhance power to address important new questions about hearing loss. If the model is successful, it could also be applied to other conditions detected by population screening before or soon after birth. Therefore, we also aim to:
• Facilitate new partnerships, training opportunities and capacity amongst Victorian researchers for population-based research in congenital hearing impairment;

• Set up VicCHILD so that the same methods and database could readily be adopted for other population data collections, separately or pooled with VicCHILD data, for:
  o congenital hearing impairment in other Australian states/territories, and/or
  o other conditions detected via prenatal or newborn population screening in Victoria;

• Develop collaborations with other countries who may wish to join VicCHILD (so far, Singapore, New Zealand, Austria and the Netherlands have expressed interest); and

• Present the findings to the wider community through publications and media.

4. METHODOLOGY

4.1 Design

VicCHILD will be a condition-specific, prospective, longitudinal databank. Briefly, the databank will amalgamate 3 main types of data, described more fully in later sections:

• Limited questionnaire information collected directly from parents, supplemented by child-reported information and direct assessment data when available;

• Stored biosamples; and

• Participating children’s relevant administrative and other existing data held by various government, health and education sector agencies.

From mid-2011, VicCHILD aims to undertake continuous, ongoing recruitment targeting all children with congenital permanent hearing loss born from Jan 1 2011 and recorded in the Victorian Infant Hearing Screening Program (VIHSP). This prospective recruitment will be augmented by two additional, discrete, retrospective recruitment waves to bring older children and their existing data into VicCHILD:

• 2011: a one-off retrospective recruitment of 157 participants (based on 70% uptake) from two recently-completed population-based studies (CHIVOS and SCOUT).

• 2012: all eligible children born between March 2005 (when VIHSP first began systematically offering UNHS to around 30% of the state’s births) and December 2010 (when the statewide roll-out became virtually complete) and recorded in the VIHSP database.

These participants will then contribute to the same ongoing prospective data collection as the other participants.

4.2 Participants

4.2.1 Eligibility

Victorian children will be invited to participate in VicCHILD if they have a mild or greater permanent unilateral hearing loss in the affected ear or bilateral sensorineural hearing loss in the better ear, where the hearing loss is believed to be congenital (i.e., present at or developing soon after birth).^1

^1 A mild or greater hearing loss is defined as averaging at least 25dBHL or more. Either or both of the 3 and 4 frequency PTAs (Pure Tone Audiogram) must meet these criteria. A 3 frequency average hearing loss (3FAHL) is averaged across 500, 1k and 2k Hz; a 4FAHL is averaged across 500, 1k, 2k and 4k Hz.
Children with temporary hearing impairment will be excluded from VicCHILD, as will children with conductive hearing loss. Families of non-English speaking backgrounds (NESB) will be eligible but until funding is available to support translation services, families will require sufficient English to provide informed consent (i.e., read and understand the participant information statement and consent form) and complete the baseline questionnaires with researcher assistance. We hope that future funding will enable fuller NESB participation.

4.2.2 Population

VicCHILD will be populated with data obtained through both prospective and retrospective recruitment of participants as detailed below.

Prospective component: From June 2011 ongoing

Victorian infant Hearing Screening Program (VIHSP)

Since January 2011 Victoria has fully implemented the VIHSP, meaning that every Victorian newborn is eligible for a hearing screen. This provides the population-based sampling frame, and thus the impetus, for VicCHILD.

The relevant key figures pertaining to VicCHILD’s accrual are as follows:

- 72,000 babies are born in Victoria each year (2009-10 year), of which VIHSP screens around 80%.
- Permanent bilateral moderate or greater hearing loss affects 0.96/1000 babies screened (approx 67 children/year).
- Permanent mild bilateral or unilateral losses affect a further 0.61/1000 babies screened (approx 43 children/year).

Thus, we expect around 110 babies to be eligible annually.

- Assuming a 70% uptake, we expect approx 80 infants annually to join VicCHILD.
- VIHSP infants recruited prospectively will be those born or diagnosed after Jan. 2011.

Retrospective components

In addition to prospective recruitment from VIHSP, we aim to have three one-off recruitment processes for children diagnosed through VIHSP born in 2005-10 and for participants in the SCOUT and CHIVGS projects, both of which are conducted through the Centre for Community Child Health. These aim to boost early numbers in the VicCHILD databank and capitalise on rich data already collected in previous studies.

(i) Statewide Comparison of Outcomes of Hearing Loss (SCOUT) (populate end 2011-beginning 2012)

SCOUT is briefly described in Section 2, page 3, above. Once data linkage occurs, the SCOUT study provides a means of populating the database with participants who have already contributed rich outcomes data.

- 137 children from SCOUT will be approached to take part in VicCHILD.
- Assuming a 70% uptake, we expect approximately 95 children
- In 2011, these individuals will be 6-8 years of age.

(ii) Children with Hearing Impairment in Victoria Outcomes Study (CHIVGS) (populated end 2011-beginning 2012)
CHIVOS is also briefly described in Section 2, page 8, above. The sample’s children are now young adults with mild to profound congenital hearing impairment. Once data linkage occurs, CHIVOS provides a means of populating the database with young adult participants who have already contributed rich outcomes data.

- 88 children who took part in CHIVOS will be approached to take part in the VicCHILD project.
- Assuming a 70% uptake, we expect approximately 62 young adults to join the databank.
- In 2011, these individuals will be 18–19 years of age.

(iii) Victorian Infant Hearing Screening Program (VIHSP) (2012–13)
We aim to retrospectively approach all children meeting criteria born between March 2005 (when VIHSP began offering UNHS to around 30% of Victoria’s births) and December 2010 (when statewide roll-out was virtually complete) through the VIHSP database. This retrospective recruitment commenced in 2012, once VicCHILD had been successfully established by the end of 2011.

- As of October 2010, 256 infants had been detected by VIHSP as having a permanent bilateral or unilateral hearing loss.
- Assuming a 70% uptake, ~180 children could be recruited retrospectively from VIHSP.
- In 2011-2, these children will be 1-8 years of age.

Table 1 shows the anticipated accrual to VicCHILD based on the above assumptions. Appendices 1.1 and 1.2 show the lower accrual if uptake rates were 50% and 30%, respectively.
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4.2.3 Recruitment and consent

Figure 1 summarises graphically how participants will be recruited to VICCHILD through VIHSP, SCCUT and CHIVOS. Families may also hear about VICCHILD from some other source outside of these studies and will be free to make contact with the VICCHILD team. More specific details related to mechanisms for recruitment through each of the three recruitment sources are shown in flowcharts presented in Appendices 2.1 – 2.3.
Figure 1. VicCHILD recruitment flowchart
As each child becomes eligible for ViCHILD, we will ask parents to consent to join the longitudinal databank. As the databank will accrue indefinitely, we are not specifying an encide or sample size for this process.

This initial consent will cover many, but not all, of the databank’s activities over time as the child matures. It will cover as much future data linkage and collection as we can foresee and is reasonable. Additional consent will be sought as necessary. Specifically:

- Medicare consent covers only the 5 years of Medicare/PBS data preceding the consent. In order to develop a lifetime health services utilisation repository, it is likely that parents will need to be asked to re-consent every 5 years to access that quinquennium’s data.
- We may request further consent if new data sources, relevant to outcomes of hearing loss but unforeseen at the time of this Ethics application, become available.
- The Victorian Department of Health is currently piloting achieving explicit written parental consent for research use of the newborn screening card (Guthrie card) at the time of sample collection. This consent applies only to use of non-identifiable/anonymous samples for research use. As we require samples to be identifiable, we will be seeking explicit consent for access to the newborn screening cards.

Consent mechanisms will vary somewhat according to the recruitment portal (i.e., through VHSP, SCOUT or CHIVOS) as this determines both the commencement age and the data that are already available for the child. Separate information and consent forms have been prepared for each of the three recruitment mechanisms (coded version names are shown in the flowcharts presented in Appendices 2.1 – 2.3, as discussed below). The following sections outline recruitment and consent processes specific to each recruitment type.

The broad stages of consent are summarised in the following Box:

| A | VHSP/SCOUT/CHIVOS contacts families by letter, briefly explaining ViCHILD and offering ‘opt-out’ by reply-paid slip from being contacted by ViCHILD. If families do not opt out within 2 weeks, ViCHILD will phone to seek verbal assent to send the Participant Information Statement and Consent Form to the participant/parent. Parents will be aware that accepting this information does not constitute consent. Parents of children recruited through VHSP will also receive 2 questionnaires to complete. SCOUT parents will also receive a buccal sample collection kit, and questionnaire to complete. CHIVOS young adults will only receive the PIS/consent forms and a buccal (check) sample collection kit (no questionnaire). CHIVOS parents will receive only the PIS/consent forms (no questionnaire).

| B | Where funding permits, a ViCHILD researcher schedules a single home visit to explain the information and consent form face-to-face and to take a saliva sample or buccal swab from the child (ViCHILD recruits only). If a home visit is not made (i.e., SCOUT and CHIVOS recruits and VHSP recruits who either live too far from Melbourne or decline the home visit), we will offer the same process over the phone. At this point the parent’s written consent is sought for:
  - At that visit:
    - The initial questionnaire (VHSP and SCOUT recruits only)
    - Collecting a saliva sample or buccal swab (by mail if no home visit)
    - Access to newborn screening card
    - Access to prenatal maternal serum
    - Use of data
      - Linking their child’s information with other existing datasets (i.e., “data linkage/retrieval”)
      - Exchange of information with service providers, if relevant (see Section 8.1)
      - Re-contact to participate for research projects
      - Using their child’s ViCHILD data for future/other research projects (i.e., “data sharing”).
  - At 6-7, 10-12 and 15-17 years:
Recruitment through VIHSP

The cornerstone of recruitment to VicCHILD will be via the Victorian Infant Hearing Screening Program. Recruitment through VIHSP will be both prospective (those diagnosed after January 2011) and retrospective (those diagnosed prior to January 2011). The flowchart presented in Appendix 2.1 shows details of this recruitment process, including coded version names for associated letters and the participant information statement and consent form.

Parents of newborn babies who agree to a VIHSP screen (nearly all whom VIHSP approaches) consent to VIHSP keeping their babies’ basic demographic, risk factor and screening data for quality purposes. The ~1% who do not pass the VIHSP screening test are referred to an audiologist for further tests to confirm a diagnosis, and parents sign a further consent to allow retention of data for quality, evaluation and audit purposes, re-contact and communication between relevant professionals.

Our proposed recruitment mechanism is modelled on that used by the Sleeping Sound with ADHD trial (EHRC 30033). This was developed in consultation with a representative from the Victorian Health Services Commissioner’s Office who advised that it is in agreement with the Privacy Act of Victoria. For recruitment to VicCHILD of children diagnosed with a hearing impairment through VIHSP, we propose the following:

- VIHSP sends an approach letter to parents telling them about VicCHILD, and a pending invitation to take part. This will usually occur at around 4-6 months of age. This timing for approach to potential participants has been developed in consultation with workers from the VIHSP Early Support Service program (funded by the Victorian Department of Education and Early Childhood Development). This time point is approximately 3-4 months post-diagnosis, and therefore aims to avoid the period of maximum distress, attendance at appointments and decision-making that typically follows the diagnosis.

Using an ‘opt-out’ approach, parents who do not wish to learn more about the study will be asked to notify the VIHSP team within 2 weeks by returning the ‘opt-out’ slip at the bottom of the letter by mail.

In the Sleeping Sound with ADHD trial (EHRC 30033), paediatricians send the ‘opt out’ letter to their eligible patients. Of the 436 patients contacted thus far, 35 (8.2%) have opted out (personal communication, Dr Emma Sciberras, 13 Dec 2010).

If VIHSP receives the envelope back marked ‘Return to Sender’, a member of the VIHSP team will attempt to contact these parents by phone to ascertain willingness to hear about VicCHILD. If no contact can be made, details will not be passed to VicCHILD.

- Two weeks after sending the letter, VIHSP will pass contact details of those parents who did not decline from being contacted about the study to the VicCHILD team.
• The VicCHILD team will conduct an initial phone call to the parent to confirm interest in the study and gain verbal consent to send a study information pack which contains a cover letter, the participant information statement and consent form, parent questionnaires (study generated and the Ages and Stages) and a buccal swab or spit pot. Multiple attempts will be made to reach families on different days and at different times of the day. A maximum of 2 answering machine messages will be left. Where telephone contact is not made a brief introductory letter will be posted to the family, requesting that they contact the VicCHILD team either to decline participation or confirm interest in VicCHILD. Where telephone contact is successful, a time will be arranged to conduct a home visit to go through the various aspects of the consent form with the potential participant and to gain written consent for participation in VicCHILD. Where home visits will not occur, a “follow-up” phone call will be made 1 week after sending the initial pack. Where telephone numbers are found to be incorrect (or unavailable) and an email address is available, an attempt to contact families by email will be made.

• Where a home visit is to occur, the “SKYPE” online, password secure program (http://www.skype.com) will be used to send SMS text message one day prior to the home visit as a reminder to families. The SMS requests confirmation of availability for the visit by the family, in the form of a return SMS. Those without a mobile phone will be called as a reminder for their visit:

   “Hi [parent], [Researcher] from VicCHILD will be visiting you on [Day] the [date] at [time]. Please reply “yes” to confirm OR call 93454215 to reschedule. Thanks!”

• Where a home visit will not occur, and where the VicCHILD study pack including consent forms are not received by mail one week after “follow-up” call is made to the participant, an SMS reminder will be sent:

   “Hi [parent name]. Just a quick reminder to return your VicCHILD pack. Questions or need a replacement? Call 93454215. Thanks!”

   along with an email reminder (where an email address is available).

   Those without a mobile phone will be called. Following the SMS (or call) and email, the VicCHILD team members will wait one week for contact to be made by the family, in terms of either a returned recruitment pack or a statement of intent (by either phone, SMS or email) by the family that they will do so. Contact will then be made by phone to confirm interest and act as a further reminder to return forms, and a maximum of 2 messages will be left on answering machines. A reminder letter and replacement forms will be posted where necessary.

**Retrospective recruitment through SCOUT**

The same approach will be used to invite parents of SCOUT children into VicCHILD, except that SCOUT researchers will send the initial approach letter. The flowchart presented in Appendix 2.2 details this process for SCOUT recruits, including coded version names for associated letters and the participant information statement and consent form. Home visits may be offered (funding dependent) to those families who have expressed interest in participating in VicCHILD but have not returned materials, despite completion of the full reminder process (i.e., as for VLHSP recruits who do not have a home visit, see above).

**Retrospective recruitment through CHIVOS**
The main difference for CHIVOS participants is that, being 17-18 years old by the third wave of the CHIVOS study in 2010, all will have reached the legal age of consent by late 2011. Thus, CHIVOS participants can consent autonomously to join VicCHILD.

CHIVOS will therefore address the VicCHILD approach letter both directly to the young CHIVOS adult and to their parents, using the same 'opt-out' approach.

CHIVOS parents will be asked only to provide their written consent to include the CHIVOS data contributed by them about themselves and their households in the three waves since 1999 into VicCHILD.

The process for CHIVOS recruits is detailed in the flowchart presented in Appendix 2.3. Coded version names for associated letters and participant information statements and consent forms are included. Again, as for SCOUT recruits, home visits may be offered (funding dependent) to those CHIVOS families who have expressed interest in participating in VicCHILD but have not returned materials, despite completion of the full reminder process.

Sibling recruitment

CHIVOS and SCOUT families may have other hearing-impaired children whom they would like to be included in VicCHILD. These siblings will be eligible to join VicCHILD if they have been born since the 1st of January 2006.

Document version codes:

There are a number of documents associated with the VicCHILD recruitment process, with different versions of each of these documents associated with each recruitment source. This is required due to the difference in age for children in the recruitment sources, and due to the difference in data being collected for each recruitment source. These document version codes are shown throughout the flowcharts presented in Appendices 2.1 – 2.3. The coding system has been designed to be as intuitive as possible. The table in Appendix 2.4 lists each code and its associated document, thereby demonstrating the coding system.

5. COLLECTION AND STORAGE FROM EXISTING POPULATION AND HEARING-SPECIFIC DATABASES

5.1.1 Database sources

The power of VicCHILD lies in the wealth of population-based general and specialist predictor and outcome data available from existing data sources. This complements the relatively small amount of information that will be collected directly from parents and children themselves.

Data custodians from a number of the below resources have agreed in principle to providing data to VicCHILD. For further information about the data anticipated for collection from these agencies, please see Appendix 4. Potential data sources for data collection are shown below; R indicates that data will most likely be acquired by retrieval and L by linkage (see next section for further discussion).

- Retrospective:
  - Victorian prenatal maternal serum screening program (R)
  - Victorian newborn bloodspot screening program (Guthrie cards) (R)
  - Victorian Perinatal Database (R or L)
  - Victorian Birth Defects Register (R or L)
  - Victorian Infant Hearing Screening Program (VIHSP) (R)
• SCOUT and CHIVOS data for the children in these studies (R)

• One-off prospective:
  • School Entrance Health Questionnaire (SEHQ) (school entry) (R or L)
  • Australian Early Development Index (AEDI) (school entry, if available) (R or L)
  • VCE/VCAL (school leaving) marks

• Repeated prospective:
  • Australian Hearing (0-18 years) (R)
  • Medicare and Pharmaceutical Benefit Scheme databases (every 5 years) (R)
  • National Assessment Program - Literacy and Numeracy (NAPLAN) – (school Years 3, 5, 7 and 9) (R)
  • Hospitals admissions data (linkage directly to hospitals via BioGrid) (L)
    • VAED (Victorian Admitted Episodes Data)
    • VEMD (Victorian Emergency Minimum Dataset)
    • The Royal Victorian Eye and Ear Hospital & Cochlear Implant Clinic

• Additional potential sources include:
  • Victorian Deaf Education Institute
  • Program for Students with Disabilities (PSD)
  • Longitudinal Disability Databank (under development)
  • Early intervention services (private and publicly funded)

5.1.2 Data retrieval and linkage

Confidential data retrieval

Direct data retrieval will occur for data sources where retrieval is more straightforward than linkage, or where organisations do not have the infrastructure for data linkage. At specified times, VicCHILD will confidentially provide participant details and copies of consent forms to the organisation’s data custodian. The custodian will then retrieve relevant data for the specified children to send to VicCHILD.

Re-identifiable Data linkage

Data can potentially be linked using linkage platforms such as BioGrid and the Victorian Data Linkages (VDL, Victorian Department of Health). Data linkage via these means would be anonymous, although data would be in re-identifiable form (via linkage keys) for purposes of matching to VicCHILD participants. These data linkage processes are described in more detail below.

BioGrid (http://www.biogrid.org.au/wps/portal) is an Australian data-linkage resource which has been developed, using stringent ethical processes, to facilitate access to and sharing of clinical and genetic research data in a privacy-protected and controlled manner. Before data are linked, patient-identifying data are replaced by a unique linkage code and kept on a separate database to clinical information. No actual data are held by BioGrid. Instead, individual organisations make a local data store visible to BioGrid using encrypted channels, over the internet. No patient identifiers are attached to the clinical data records other than the unique linkage code. Only authorised clinicians and researchers are allowed to access BioGrid data (no access to identifiable data). The access is strictly controlled by a process whereby BioGrid’s Management and Scientific Advisory Committees’ and the data custodians’ approval must be met, before data become accessible or are released. Both BioGrid and the organisations which participate in data linkage abide by rigorous governance and behavioural standards, which include a strict approval process. BioGrid data owners have total control over the data they make available.
The Victorian Data Linkages (VDL) Unit is in the early stages of establishing a statewide linkage capacity to progress Victorian population research. Currently, VDL allows for linkage to the Victorian Admitted Episodes Data Set (VAED) and the Victorian Emergency Minimum Dataset (VEMD), and is in preliminary consultation with other medical and educational data sources. VDL has strict policies which follow the best practice protocol for health data linkage and federal privacy legislations in order to protect the confidentiality of individuals who have data linked within the database. All data are returned post-linkage with no patient identifiers are attached to the clinical data records, with each individual assigned a data linkage key. All data custodians must have approved the data linkage prior to data being released for linkage by VDL.

Data retrieval and linkage schedule

Timing of data retrieval and linkage will be pre-specified for each data source to maximise data completeness and value while minimising cost for VicCHILD and burden for the data source. Table 2 below shows the current projected timing of data collection for retrospective, one-off prospective and repeated prospective database sources listed above.
Table 2. Projected timing of data linkage/extraction for VicCHILD participants by age and year from 2011-2021

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</table>

*P Pre-P perinatal data: Maternal prenatal serum, Guthrie card access, Victorian Perinatal Data Collation, VSEP
*V Victorian Birth Defects Register
*H Australian Hearing
*A Australian Early Development Index (if available)
*E European Language and Literacy
*VCE/VGAL results
*R Retrospective linkage to OHVOD data which collects AIMS (AIMS was replaced by NAPLAN in 2007), NAPLAN & VCE results, information from the ophthalmic implant centre and audiology tests from Australian hearing.
6. MEASURES COLLECTED BY VICCHILD

6.1 Data collection processes

VICCHILD collects measures throughout each participant’s childhood in three ways: through parent questionnaires, child assessments and child completed questionnaires (section 6.2.1 and section 6.2.2). Questionnaires are completed by parents firstly at the time of enrolment in VICCHILD, then periodically at every scheduled 5-7 year follow up. To introduce the project (see Page 18) or to set-up follow-up visits (usually at 5-7 years, 9-12 years, and 15-17 years), contact is typically made to parents by phone (or mail, text or email where judged to be more appropriate, such as instances where parents are hearing impaired). Where parents agree to an assessment home visit, a selection of measures is sent to parents in advance of the visit, giving them the opportunity to commence completing them. For example, these include a self-report vocabulary measure for each parent to complete (the Mill Hill) and parent reported quality of life measures. At the child assessment, which usually occurs in a family’s home or alternatively at the Murdoch Childrens Research Institute, any remaining parent and child completed questionnaires are administered in addition to direct assessment measures. Where possible, all questionnaires are collected by the researcher on the day of the visit; alternatively, parents can mail them back.

Commencing early 2015 VICCHILD will be trialling an additional point of contact with parents via a mailed questionnaire, with scope to incorporate this into the permanent data collection processes if the exercise proves acceptable to families. As this point of contact solely involves questionnaire activity, further details can be found in section 6.2.1 Parent completed questionnaires.

6.2 Questionnaires and language assessments

We plan to collect three types of information from participants and parents at the key 5-yearly time points:

- Parent-reported questionnaires covering:
  - child development, health and well-being, education and resource use
  - parent and family health and demographics

- Child-reported questionnaires at 9-12 and 15-17 years covering:
  - Health, well-being and quality of life

- A home visit Assessment visit module (should funding permit) includes:
  - Direct assessment of child’s language, cognition and academic achievement
  - Parent completed questionnaire relating to child’s development, health status and their own vocabulary levels
  - (Existing SCGUT and CHIVOS assessment data provided retrospectively to VICCHILD.)

As noted in 6.1 above, this protocol necessarily covers all of VICCHILD from its conception through its lifespan, including different ages of children at different time periods. Therefore,
this section covers all measures, not all of which are being asked at any given time. Table 3 shows the VicCHILD schedule of measures from 2011 until mid-2014; Table 4 shows the schedule of measures from mid-2014 until further notice.

6.2.1 Parent completed questionnaires

At baseline and approximately 5-yearly, parents complete a (mailed) questionnaire which reports on the development of their child, and their own and their child's health and wellbeing. CHIVOS participants and their parents are not asked to complete questionnaires, as many of the measures used in VicCHILD have already been completed by the parents during their participation in the CHIVOS study. SccUT parents do not complete the 5-7 year questionnaire for the same reason. Families recruited to VicCHILD through VIHSP and SccUT also complete short enrolment questionnaires which provide basic demographic data and details of child's birth, family history (hearing loss-related) and child's hearing loss (also impact on family of child's hearing loss for VIHSP recruits).

Child and adult health and well-being outcomes are assessed using valid and reliable measures currently being used in other large-scale Australian research projects studying the language, development and/or well-being of children with and without hearing loss. The parent questionnaires also contain items relating to family demographics, child education and resource and service use.

When funding permits, participants undergo language and cognitive assessments at (approximately) the 5-7, 10-12 and 15-17 year-old time points to track their development and progress using age-appropriate measures. These assessments are usually conducted during home visits or alternatively at the Murdoch Children's Research Institute, for approximately 60 minutes duration. The ages at which these measures are collected are summarised in Table 3 (2011-mid-2014) and Table 4 (mid-2014 onwards).

The trial of an additional point of contact with families via a mailed questionnaire will occur for children aged 1-3 years and will include measures that are summarised in this document, being the MDQI, PEACh, PedsQL, and study generated questions relating to education and child's hearing. Reminder processes for the follow up of the questionnaire will mirror those previously approved for the recruitment of participants into VicCHILD (see Section 4.1 Recruitment and Consent, page 19 of this document)

Parent completed questionnaires comprise the following measures of health and well-being:

- **Kessler-6 (K-6) (about the parent)**
  - Psychological distress scale, used to provide a global indicator of mental health. The 6 item measure takes approximately 2 minutes to complete.

- **PedsQL Generic Core Scale (0-25 years)**
  - Child quality of life measure relating to physical, emotional, social & school functioning, with a completion time of less than 4 minutes.
  - Infant scales for 1-12 months (36 items) and 13-24 months (45 items)

- **PedsQL General Wellbeing Scale (8-18 years)**
  - 7 item measure relating to feelings about self, support from family and friends, general health, health hopes for the future. Completion time of less than 1 minute.
• **PedSQL Family Impact Module (about the family, no age specifications)**
  - Items relating to the primary parent/guardian’s communication with others and worry about their child and relevant condition, the family activities and relationships.
  
  Note: The author of the PedSQL measure has approved small item modifications for the CHILD. For items 7 & 8 the words “illness” and “condition” have been replaced with “hearing loss.”

• **PedSQL Multidimensional Fatigue Scale**
  - A generic symptom-specific instrument designed to measure fatigue in patients with acute and chronic health conditions, as well as healthy populations.
  - Versions exist for age groups spanning toddlers from 2 years to adults over 26 years, with parent and self-reports.
  - The completion time for the measure (parent report for young children ages 5-7 years) is less than 3 minutes (15 items).

• **Strengths and Difficulties Questionnaire (3-17 years)**
  - 25 item behaviour and mental health screening questionnaire, with a completion time of approximately 5 minutes.

• **The Ages and Stages Questionnaire (4-66 months)**
  - Developmental milestones, including communication, gross motor, fine motor, problem solving and personal-social milestones, are completed by parents of all VHSP recruits.

• **The Children’s Communication Checklist 2 (CCC-2; 4-16 years)**
  - Pragmatic language impairment subscale, which has a completion time of approximately 5 minutes.

• **The Communication and Symbolic Behaviour Scales (GSBS; 5-24mo)**
  - Communication milestones including expressive and receptive vocabulary.

• **MacArthur Communicative Developmental Inventory (MCDI; 8-37mo)**
  - Communication milestones including expressive and receptive vocabulary.

Parent-completed questionnaires also contain items related to the following:

• **Demographic information**
  - Reporting parents gender, relationship to child, level of completed education, country of birth, and date of birth, employment status.
  - Household income, benefits, Health Care Card.

• **Health items**
  - “In general how do you rate (your/your partner’s/your child’s) health?”
  - “Do (you/your partner) have any illness or condition that requires you to see a health professional frequently?”

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• **Education**
  - Name, suburb and type of school attended by the child (if school-aged)
  - Date (year) child began attending school & number of years child has attended school

• **Resources and service use**
  - Use of assistive devices such as hearing aid and cochlear implant
  - Service use (e.g. early intervention services)
  - Financial support (Centrelink, health care cards)
  - Health professional use (e.g. speech pathologist)
  - Satisfaction with services and resources, and any associated costs to families

• **Child’s hearing**
  - Name cause of hearing impairment if known (The cause of the child’s hearing impairment is discussed in more detail in the assessment module. This discussion is aided by the causes list, provided in the assessment module booklet).
  - Use of hearing aid, cochlear implant, and/or FM system (including willingness of child to use, the usefulness of the device & frequency of use)

• **Child’s communication**
  - How the child communicates (spoken language, sign, etc.) and how well the child is understood

• **Family history of hearing, speech or language conditions or disorders**

• **For children recruited via VIHSP or via self-referral at the baseline visit only**
  - Child pre- and post-natal information

Table 3 summarises the content of enrolment and 5-7 year questionnaires for VIHSP recruits, and enrolment questionnaire for SCOUT recruits, during VicCHILD’s establishment period 2011-mid 2014. Specific measures and items included in each questionnaire are detailed. A number of the VicCHILD questionnaire measures and items have been used in the SCOUT and CHIVOS studies and within VIHSP. Table 3 maps these measures and items across VicCHILD, SCOUT, CHIVOS and VIHSP questionnaires in order to demonstrate the complementary (rather than overlapping) nature of the VicCHILD data collection with these studies. The VCHWS (Victorian Child Heath and Wellbeing Survey; Department of Education and Early Childhood Development, 2011) is also included in this table to demonstrate the potential to compare VicCHILD findings with population-level data for 0-12 year-old Victorian children. Assessment visit measures are also included (see section 5.2.2).
Table 3. VicCHILD measures and items 2011- mid 2014

<table>
<thead>
<tr>
<th>Measures</th>
<th>VICCHILD data collection</th>
<th>Data collected by other projects linking to VicCHILD</th>
<th>norm data</th>
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<tr>
<td></td>
<td>Enrolment Q data: VHSP recruits¹</td>
<td>5-7yr Q data: Visit; VHSP recruits¹</td>
<td>Enrolment Q data: SCOUT recruits</td>
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<tr>
<td>Ages and Stages Questionnaire</td>
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| Clinical Evaluation of Language 
Function (P2 or Version 5) | | | | | | | |
| Hearing screening Q data² | | | | | | | |
| Bus Story² | | | | | | | |
| AQoL-4D³ | | | | | | | |
| CHUQ² | | | | | | | |
| Heel prick | | | | | | | |
| Nil Hill Vocabulary Test (for parents)³ | | | | | | | |
| Peabody Picture Vocabulary Test | | | | | | | |
| Peds-QL | | | | | | | |
| Peds-QL Family impact module | | | | | | | |
| Causes of hearing loss | | | | | | | |
| SDQ | | | | | | | |
| Velonon Non-verbal ³ | | | | | | | |
| Items | | | | | | | |
| Birth weight | | | | | | | |
| Gestational age | | | | | | | |
| Type of birth | | | | | | | |
| Cochlear implant/ hearing aid use | | | | | | | |
| Global health item (parent/partner) | | | | | | | |
| degree of hearing impairment on child | | | | | | | |
| Hearing impairment cause³ | | | | | | | |
| Has child started primary school | | | | | | | |
| Name and suburb of school | | | | | | | |
| Type of school attended | | | | | | | |
| Primary parent gender, DOB, relationship to child | | | | | | | |
| Primary parent education, employment status | | | | | | | |
| Partner gender, DOB, relationship to child | | | | | | | |
| Partner parent education, employment status | | | | | | | |
| Language spoken at home | | | | | | | |
| Numeracy income | | | | | | | |
| Who does child live with | | | | | | | |
| Total number of children | | | | | | | |
| Family history | | | | | | | |

¹ Data collected by other projects which will populate the VicCHILD database via linkage or retrieval.

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Table 4 summarises the measures collected through questionnaires and assessment visits (see section 5.2.2) for all recruited children aged birth to 12 years old from mid-2014 onwards (the consolidation phase of VicCHILD). Slight differences from the 2011-2014 questionnaires incorporate new measures and ensure that an appropriate questionnaire is available for every child in VicCHILD no matter what age they join (such as older SCOUT recruits and retrospective recruits), or what recruitment source they came from (including via siblings or word of mouth). This means that we can provide both (1) an age-independent enrolment questionnaire and (2) an age-specific questionnaire every 5-7 years, no matter what age the child.
### Table 4. VicCHILD measures and items from mid 2014, via questionnaires and Assessment visits.

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**Note:** Items in bold indicate new measures or changes from previous years.
6.2.2 Home Assessment visit module

When funding is available, outcomes in terms of child language, communication and development are assessed (usually in the child’s home or alternatively at the Murdoch Childrens Research Institute,) using the measures listed below. These assessments are conducted at the ages of 5-7 years (for VIHSP children only), 9-12 years (VIHSP and SCOUT children only), and 15-17 years (VIHSP and SCOUT children only). Assessments are usually arranged with parents via the phone, and a reminder is sent before the visit in the same manner as before the recruitment home visits (see section 4.2.3). SMS and email contact may be used instead of phone contact where more appropriate for families. At this time researchers enquire about whether the participating child has any additional diagnoses or developmental difficulties that would be helpful for VicCHILD researchers to be aware of prior to conducting the assessment. This information may be necessary in order to accommodate the different needs of children during the assessment process.

For inter-rater reliability and language sample purposes, and to assist with scoring, these direct child assessments may be audio recorded. With parents’ consent, the recording is stored as part of the VicCHILD database to provide samples for language comparison data over time. If parents request assessment feedback, VicCHILD sends a short report visually summarizing their child’s language outcomes. This report follows the previously-approved format for Language for Learning (HREC 30311) and Let’s Learn Language (HREC 260323), used for over 1500 families without incident. The feedback letter is sent within six weeks from the date of assessment. In instances where a feedback letter is requested and the child scores below the tenth percentile on the expressive and/or receptive indices of the CELF, the VicCHILD Project Manager (ST) will phone the families to discuss the results prior to mailing out the feedback letter.

Child assessment: measures (see Table 4 for timing of VicCHILD administration; all are validated with population norms for the age range specified below, unless otherwise stated)

Depending on the age of the child:

1. (a) Clinical Evaluation of Language Function (CELF-P2) (3-6.11 years)
   • Assesses receptive and expressive language skills, completion time of 30-60 minutes.
   This is used only for 5-6 year olds with language skills that are lower than needed to complete the CELF-4 or -5.

2. (b) Clinical Evaluation of Language Function (CELF-4 or -5) (5-21 years)
   • Assesses receptive and expressive language skills, completion time of 30-60 minutes.
   The transition from CELF-4 to CELF-5 will be made in line with clinical practice, once Australian norms are available.

3. Wechsler Nonverbal Scale of Ability (WNV) (4-21 years)
   • Non-verbal measure of cognitive ability, completion time of 15-20 minutes. This is the only assessment that also has norms for children with hearing loss, as well as for the hearing population.

4. (a) Peabody Picture Vocabulary Test (PPVT-4) (2.5-90+ years)
• Instrument for measuring the receptive (hearing) vocabulary of children and adults, completion time of 10-15 minutes.

OR

3. (b) National Institutes of Health Picture Vocabulary Test (NPVT) (3-85 years)

• Measure of receptive vocabulary of children and adults, completion time of 4 minutes. We are trialling this test for ViCHILD in late 2014 and will make the permanent change from the PPVT to the NPVT if it proves to be shorter, less burdensome, and more fun for the participants.

4. (a) The Bus Story Test – Revised Edition (3-8 years)

• A test of narrative language for children that correlates highly with subsequent language scores. The child is told a story and then asked to retell it using picture prompts, from which information content, sentence length and grammatical usage can be scored in the context of naturalistic speech; has age norms.

OR

4. (b) Expression: Reception and Recall of Narrative Instrument (ERRNI) (4-15+ years)

• A structured test of narrative language that assesses ability to relate, comprehend, and remember a story after a delay. This provides information on the child’s ability to remember content, their use of grammatical structures, and their comprehension. Has Australian age norms and is suitable for children too old for the Bus Story Test.

5. Child Health Utility 9D (CHU9D) (7-17 years)

• A brief 9 item questionnaire that is a measure of health related quality of life in children and allows the calculation of quality adjusted life years (QALYs). The questionnaire asks children about how their health affects their lives.

6. Goldman-Fristoe Test of Articulation (GFTA) (7-21 years)

• Measures spontaneous and imitative sounds production; approximately 5-10 minutes.

7. Comprehensive Test of Phonological Processing (CTOPP) (5-24 years)

• Has several subtests measuring the ability of the child to perceive and manipulate sounds. Takes approximately 15-20 minutes.

8. The Children’s Test of Nonword Repetition (CNRep) (4-8 years)

• Tests children’s ability to repeat nonwords; measures are closely related to language skills more generally. Takes approximately 4 minutes.

9. Listening in Simulated Noise – Sentences Test (LSN-S) (5-80 years)

• An adaptive listening task that measures speech perception ability in noisy environments. This test is normed on those with normal hearing and hearing
impairment up to moderate degree, and takes approximately 5 minutes per subtest to complete (four subtests).

10. **Wide Range Achievement Test (WRAT-4) (5-94 years)**
   - This test assesses the academic achievement of individuals. It takes approximately 20 minutes.

**Parent-completed measures at the assessment**
The following measures are completed by the parent. For parents who decline an assessment, **VocCHILD** mails the questionnaire booklets, and completes the cause of hearing impairment questions by phone:

1. **Mill Hill Vocabulary Scales (parent's own self-reported vocabulary)**
   - Assesses parent vocabulary and verbal reasoning ability. This involves each parent completing a short written questionnaire, with parallel forms (A and B) being used for the two parents.

2. **Health Utilities Index Self-administered questionnaire, Proxy assessment (HUI-2 & 3) (5 years +)**
   - Child health related quality of life (utility measure), centered around sensation, mobility, emotion, cognitive, self-care, pain. Completion time of 10-15 minutes.

3. **Assessment of Quality of Life (AQoL)-4D**
   - Parent quality of life (utility measure) including 4 dimensions (independent living, relationships, mental health, senses) and a global utility score. 12 items, 1-2 minutes.

4. **Speech Intelligibility Rating Scale (SIRS)**
   - Parents complete this scale of speech intelligibility where a score is given from 1-5. A higher score indicates that for an inexperienced listener, the child is easy to understand. A lower score indicates that even listeners who know the child find their speech difficult to understand.

5. **PedoQL Multidimensional Fatigue Scale (PedoQL MFS)**
   - Parents complete 15 items, split across three areas, General fatigue, Sleep/rest fatigue and Cognitive fatigue. A score is given which rates how much of a problem has the child had with each item over the past one month, ranging from Never (0) to Almost Always (4). A higher score indicates a greater level of perceived fatigue.

6. **Use of hearing assistance devices.**
   - A study-designed set of questions regarding the child’s hearing aid and/or cochlear implant and/or FM system use during different types of activities during the day.

6. **Parental Evaluation of Oral/Aural performance of Children (PEACH) (early childhood to high school ages)**
   - Parents describe children’s aural and oral functioning in noisy and quiet conditions. This takes approximately 5-10 minutes and is suitable for use with children with any degree of aided hearing impairment.

7. **Children’s Home Inventory for Listening Difficulties Questionnaire (CHILD) (3-12 years)**
The measure is administered to parents of children with hearing impairment that is either fitted or not fitted with any assistive listening device. This measure takes approximately 5-10 minutes to complete.

8. Cause of hearing impairment

- A study-designed list of hearing impairment causes, by which parents can identify the specific cause associated with their child's hearing impairment. This will be administered using a laminated flash card with responses recorded on a researcher data collection form.

6.3 Biological samples

VicCHILD requests written consent to collect buccal swabs or saliva samples, and to access prenatal maternal serum screening samples and Guthrie blood spots, in order to use these samples in research arising from the VicCHILD project. While specific research questions regarding these biosamples have not currently been determined, participant consent for the collection and storage of these samples and/or access to them will “future-proof” VicCHILD, thereby maximising the research potential of this valuable cohort. At this stage, any genetic testing will be limited to genes already identified as playing a role in hearing and hearing loss. However, given the ongoing identification of such genes and the potential for whole-of-genome analyses, it is not possible to define a comprehensive list at this stage. Any additional analyses will be the subject of future RCH HREC modifications, e.g., should other genes beyond hearing- and hearing loss-related genes become a target of arising research questions.

6.3.1 Buccal swab or saliva sample

Buccal swabs are easily collected and processed, yielding good quality DNA. DNA will be obtained using commercially available kits, e.g. Epicentre’s Catch-All™ (Buccal swabs) and Qiagen’s QIA-500 saliva sample collection kits (DNA Genotek). The samples can be taken either by a parent, by the older child themselves, or a researcher during the home visit, or when it is most convenient. If a home visit is not conducted, kits can be posted to the participant and returned by reply-paid return envelope after the sample has been taken by the parent or child. The buccal swab is a gentle and relatively non-invasive procedure in which samples are taken by gently rubbing a swab up and down six times against the inside of each cheek, and then along the grooves next to the top and bottom gums on the same side. Providing a saliva sample involves participants delivering approximately 4ml of saliva into a purpose-designed collection kit. The procedure is not associated with any possible bodily harm except for the inconvenience of producing a saliva sample.

DNA is extracted from saliva sample and buccal swab kits using standard protocols. Where the DNA yield is insufficient, i.e., less than 1000ng (according to anticipated requirements), participants may be requested to provide a second sample at the next point of contact (e.g., at 5-7 years for VHISSP recruits). However, we note that this may not prove necessary as DNA capture methods are rapidly improving.) DNA samples are de-identified and stored in the Murdoch Children’s Research Institute (MCR) biobanking facility. The storage location of individual samples is maintained in a secure database. The biobanking facilities are secured by swipe card and only researchers involved with projects in the facility have access.

6.3.2 Prenatal Maternal Serum

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PLEASE NOTE, unlike buccal swabs, saliva samples are fine to stay at room temp for up to 2 years so don’t stress about them not being in a freezer — it’s that easy!

8.2.5 Storage of saliva DNA (to be updated by RB)

Rachel Burt’s Molecular Hearing group will be responsible for storing the VicCHILD saliva samples. Saliva DNA will be extracted and stored as below.

8.2.6 Processing saliva DNA

[Rachel Burt to insert]

9. Early Language Questionnaire

This is a 12 month trial (July 2015-July 2016) of an additional point of contact with families to complete a mailed out questionnaire looking at early language development, in line with international trends towards collecting standardised measures in the early years. Children eligible are those with any degree of hearing loss who are within the age limits of the main outcome measure used in the Early Language Questionnaire (ELQ), the MacArthur Bates Words and Sentences test (16 to 30 months). A PhD project looking specifically at mild to moderate bilateral losses at the time of diagnosis is nested within this mail-out, and has an upper eligibility cut-off of 36 months.

9.1 Eligibility and priorities of mail out activities

The priorities for who to approach and when are dependent on several factors:

1. Eligible age for main measure used in ELQ, the MRCDI Words and Sentences (16-30 months)

2. The need to maximise responses from those diagnosed with mild to moderate bilateral losses (PhD focus)

3. The idea that this questionnaire, if it proceeds past the 12 month feasibility trial, is best administered around 24 months of age to be able to compare to international data collection trends

In order to maximise the number of participants that can be approached during the trial duration (July 2015- July 2016, with ethics approval expiring August 2016), oldest children will be approached first.

An initial mail out will occur in late July 2015 that covers the age range 23 months to 29.5 months for all participants regardless of type and severity of hearing loss.

Monthly afterwards: new children turning 23 months should be sent the ELQ pack.

PhD specific activities:

Also in July 2015, the children in VicCHILD between 29.5-36 months will be audited to determine their hearing loss at diagnosis, and those who were bilateral mild or moderate will also be sent the
ELQ pack. This is in an attempt to increase numbers of responses from mild/moderate families and on the assumption that responses can be incorporated into the analysis.

In October 2015, all children from approximately 15 months to 23 months who had an initial diagnosis of bilateral mild or moderate should be sent the ELQ, to be able to complete data collection by December 2015.

9.2 ELQ pack and mail out activities

Follow the steps listed below to create the mail-merged cover letters for families with children who are eligible for the Early Language Questionnaire (ELQ).

Open: REDCap database

Under ‘reports’ Choose “ELQ Pack Due” then click on [Microsoft excel] [CSV]

This will generate a excel list of all families who have children within the eligible age range.

Copy: Columns C to I (first name, last name, address, suburb, state & postcode) of the list (don’t worry about seeing this document)

Open: [Excel to mail merge excel template] Paste the selected data into the excel mail merge template (over the top of old data). Click Save.

Close both excel documents.

Cover letters

Open: Open the cover letter for families who are to receive the Early Language Questionnaire

Z:\3. RESEARCH UNIT\Hearing\VicCHILD\Mailout materials\Cover letters\VHSP\ HREC3181 Cover Letter_ELQ_V2_20June_2015

To update the mail merge for the letters choose the ‘mailings tab’ and then ‘finish and merge’ and ‘edit individual documents’. This will automatically update the letter however do look over them to double check they match the list in the excel file.

Print the letters (in colour and NOT double sided)

DO NOT SAVE and close.
Materials in pack (assembled in the order below please)

- Cover letter
- Early Language Questionnaire with completed ID boxes to front page and page 1 (found at Z:\3. RESEARCH UNIT\Hearing\VicCHILD\Assessments Questionnaires & Measures\Questionnaires HREC_31081_Oct_1-3yr_Early_Language_V1_23_June_2015)
- PEACH Questionnaire with VicCHILD ID written on top right of front cover and page 1
- CS Reply paid envelope
- Using a paper clip, clip the cover letter, together with the early language questionnaire and PEACH questionnaire, and reply paid envelope behind (i.e., not clipped). Fold paper materials in half and seal envelope.
- On the front of the pack:
  - Please hand write the family’s address
  - Put a VicCHILD sticker on the pack
  - Stamp with the VicCHILD project stamp

Once the recruitment materials pack has been sent

Recruitment and Tracking Early Language Questionnaire Tab

ELQ Pack Sent - Pack sent variable

  Change to: ‘Yes’

  Date Pack sent: put in the date the pack was sent

Date reminders should begin: Date should be entered as 1 week after pack was sent. Found in ELQ Reminder Process sub-section

9.3 ELQ pack reminder process

Reminder call script

Reminder process for Early Language Questionnaire is as per recruitment reminder process.

<table>
<thead>
<tr>
<th>Workflow</th>
<th>Action</th>
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<tbody>
<tr>
<td>✔</td>
<td>Send pack to all eligible families</td>
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<tr>
<td>✔</td>
<td>1 week after pack is sent, make the 1st reminder phone call or email</td>
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<td>(if phone numbers found to be incorrect, email will be used)</td>
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<tr>
<td>✔</td>
<td>1 week after follow-up/reminder call made, a 2nd SMS/Email reminder can be made</td>
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</table>
1 week after SMS / email, make a 3rd follow-up reminder call
(Maximum of two messages to be left on the answering machine)
If still unable to contact a family, change status to 'Lost during follow-up, reminder process complete.'

If after approximately one week past the mail out the ELO the materials have not been returned, a phone call reminder can be completed.

Script for 1st reminder call

Hello, my name is [your name] and I am calling as part of the VicCHILD research project about hearing impairment from the Royal Children’s Hospital. Am I speaking with [parent’s name]? Great. This is a friendly reminder call to check that you have received a recent VicCHILD questionnaire pack about language in the mail?
Yes?
- That’s great, we just like to make sure that the pack has been sent to the correct address.
- Have you had a chance yet to read over the materials yet? If so, do you have any questions?
- Thank you for taking the time to read over the materials and we would be very appreciative if you would be able to complete this valuable information about [child name]’s language. If you have any questions about the study please feel free to call us on 9345 4215.
Yes?
- Oh I’m terribly sorry about that. Would you mind if I checked the address details that we have for you to make sure we have sent the pack to the correct address?
Correct address, offer to send a new pack in the mail to family
Incorrect address, write the new address on excel spread-sheet, apologise to family and let them know that we will send another pack out as soon as possible.
- Thank you for taking the time to read over the materials and we would be very appreciative if you would be able to complete this valuable information about [child name]’s language. If you have any questions about the study please feel free to call us on 9345 4215.
### Script for 1st reminder email

**Hi Parent**

We hope that you have received in the mail a VicCHILD questionnaire pack about [child name]'s language. This is just a quick reminder to return the materials if you have completed them. If you have any questions or need any replacement materials, please reply to this email and we will organise to send them out.

Greatly Appreciated,

---

### Redcap recordings

- **Open:** The Early Language Questionnaire form for the family, by selecting their ID.
- **Change:** Change the variable "Reminder 1 Phone Call Completed" to "Yes" and enter the date.

### Reminder 2: SMS or Email Reminders

If, after two weeks past the pack sent date, a family's pack has not been returned, an SMS reminder can be sent or an email (if an email address is available).

This is to be monitored manually at this stage, ideally through the mail out list.

### Skype instructions

- **Log on:** Open Skype on your computer. (You will need MCR IT to approve and to install it on your computer)
  - Log in using the following details:
    - User name: 
    - Password: 
- To send an SMS follow these steps
  - Click on Menu: Call
  - Click on :Call phones
  - Use the pin pad to enter the number
> Click on the black sms symbol
> Type message in the box and click send

Script for 2nd reminder text

Hello. Just a quick reminder to return your VicCHILD pack. Questions or need a replacement? Call 9345 4215. Thanks!

Script for 2nd reminder email - Please email the family from the VicCHILD email address

Hi Parent:

Just a quick reminder to return the VicCHILD materials about [child name]'s language. If you have any questions or need any replacement materials, please reply to this email and we will organise to send them out.

Greatly Appreciated,

Redcap recordings

Open: The Early Language Questionnaire form for the family, by selecting their ID.
Change: Change the variable "Reminder 2 SMS or email Completed" to "Yes" and enter the date

Reminder 3: Final Reminder Phone Call

If after approximately three weeks following pack sent date, the families still haven't been returned, a phone call reminder can be completed.

Script for 3rd and final reminder call
Hello, my name is [your name] and I’m calling as part of the VicCHILD research project about hearing impairment from the Royal Children’s Hospital. Am I speaking with [parent’s name]?

Great. This is a friendly reminder call to let you know that it is not too late for you to return the VicCHILD questionnaire about [child name]’s language.

- Have you had a chance yet to read over the materials as yet? If so, do you have any questions? Do you need another copy of any of the materials?
- Thank you taking the time to read over the materials and we would be very appreciative if you would be able to complete this valuable information about [child name]’s language. If you have any questions about the study please feel free to call us on 9345 4215.

Redcap recordings

Open: The Early Language Questionnaire form for the family, by selecting their ID.

Change: Change the variable “Reminder 3 Completed” to “Yes” and enter the date.

If the reminder process has been exhausted and no contact with the family has been established, a reminder pack (or a duplicate pack) can be sent to the family. Refer to the instructions in Section 9.2 for this information.

Redcap recordings

Open: The Early Language Questionnaire form for the family, by selecting their ID.

Change: Change the variable “Reminder pack sent” to “Yes” and enter the date.

Note at this point of time the reminder process starts from reminder 1 again.

9.4 ELQ completed pack processing

When packs are resumed, the Early Language Questionnaire section of the participant’s Redcap recruitment and tracking record should be completed.

Specifically the ELQ pack returned question should be changed to yes, with the date indicated.

Tick off the two components, the Early Language Parent Questionnaire (blue cover) and the PEACH (white cover) if both have been returned. For this trial, it is not unusual for parents to not return the PEACH if their child is unaided, as the PEACH instructions specifically indicate use for children fitted with hearing aids or cochlear implants. If this point of contact becomes standard for VicCHILD ongoing, the format of the PEACH will need to be addressed. The decision of whether to pursue return of the PEACH is dependent on answers contained in the blue questionnaire.
Change the ELQ reminder needed prompt to No, leaving all other fields completed in this section of the form.

Complete the record of the two consent questions (consent to provide MCDI to EI services and consent to get updated contact details from EI services) and indicate which is the current EI service attended. All information is contained in the blue questionnaire. Any information relevant but not having a specific section to be entered can be written in the notes field. This is usually the scenario where parents have consented yes to both questions, but they are not enrolled with EI services.

Hand clean both questionnaires for missing answers, and follow up with family if there is any need. Ensure VicCHILD ID numbers are on all pages of both questionnaires, update the contact details on the back of the blue questionnaire and place the two questionnaires in the data cupboard.

9.5 ELQ entry into data repository

Both the Early Language Parent Questionnaire (blue cover) and the PEACH (white cover) are entered into one form in the Data Repository, labelled ELQ T23. Entry for the most part is self-explanatory.

The largest section of the ELQ is section 4, the Vocabulary checklist which is entirely sourced from the MacArthur-Bates Communicative Development Inventories Words and Sentences (MCDI W&S). The vocabulary checklist is identical to that used in wave 3 of the Early Language in Victoria Study (ELVS) with adaptations for Australian English. A tick against the vocab items indicates the child produces that word and is given a Yes in REDCap.

Section 4.29 has the question “Has your child begun to combine words yet, such as “mother cracker” or “doggie bite”?” and three possible answers: Not yet, sometimes and often.

If “Not yet” is chosen, do NOT complete the next section (4.30 Complexity) – regardless of what answers the parent may have completed for these items. Branching logic exists so that the option to enter section 4.30 is removed if not yet is chosen.

If “Sometimes” or “Often” is chosen, complete section 4.30. If parents indicated both options for the one question (eg indicated yes to both two shoe and two shoes), select the lower/more complex version of the sentence (it is always the bottom choice). If there is no answer selected for any pair, leave blank. This is a very important step as the complexity score is derived from the second of the two examples in each instance.

10. Direct Assessment Bookings

At present (as of 27/02/2015), direct assessments are being offered to families when children are aged between 5 and 7 years, and between 9 and 12 years of age.

10.1 General protocols booking direct assessments

Ideally we seek to explain the assessment visits to the caregiver verbally over the phone. As such we may not seek to leave a voicemail message too early, as our overarching conventions on the number
of times we may contact families and the number of messages we can leave still apply for this purpose.

Therefore the following is a suggested approach, with obvious exceptions acceptable on a case by case basis:

- Attempt to speak to caregiver when they answer the phone at least twice initially, is no message to be left.

- From the third attempt onwards, a message may be left for the caregiver outlining the reason for the call in case this promotes a call back to us or facilitates the caregiver answering our next attempts.

10.2 Protocols for booking assessments when not contactable via phone

For situations where the caregiver telephone number is not of use (disconnected, not belonging to user anymore) the preference for contact next is email, then alternative contacts. The suggested email template to use is as follows:

“Dear [parent name],

I am emailing you to discuss the next exciting stage of VicCHILD as we have unfortunately not been able to contact you by telephone.

For some time now, you and [child name] have been taking part in the VicCHILD project. Now that [child name] is between [five and seven years of age] OR [nine and twelve years of age], we are hoping to complete a language and learning assessment with him/her. Collecting this type of information on the development of children who have a hearing loss is really valuable.

If you would like to hear more information about these assessments with VicCHILD, or if you have any questions, please email us back or phone us on 9345 4215.

With kind regards,

RA Name
(VicCHILD Email Signature)

Following this email being sent, allow a further two weeks for contact from the family before then proceeding to approaching alternate contacts for new telephone contact details.

10.3 Phone Script: 5-7 Year Assessments

APRIL 2015 edit: Jump from question 1 to question 7 if answer is anything other than both ears either mild or moderate (any combination).
Hi. This is [your name] calling from the VicCHILD hearing research project at the Royal Children’s Hospital. Am I speaking with [parent name]? The reason for my call is to give you a little information about our study assessments which are starting. Is now a good time to speak with you?

Yes, happy to hear more

For some time now, you and [child name] have been taking part in the VicCHILD project. Now that [child name] is between five and eight years of age, we are hoping to complete a language and learning assessment with her/him. [If required, can add the following: Collecting this type of information on the development of children who have a hearing loss is really valuable.]

What we would like to do is to come and see both [child name] and yourself at home, or alternatively you have the option of coming in to see us at the Royal Children’s Hospital. We expect to be with you for a couple of hours, during which you will complete some questionnaires about [child], and [child name] will do a few activities on language and learning which most children enjoy.

Do you have any questions for me so far?

Are you happy for us to make a time now to complete the visit with you and your child?

No – refusal to participate in Assessments (boo)

Okay, that’s fine we totally understand. Instead of an assessment, would you be happy to fill out a questionnaire if we mailed it to you? You could just post it back to us in the reply-paid envelope.

We are trying to improve our process for the study. Can I ask the reason you’re not keen on doing an assessment?

Yes, happy to hear more

That’s great! Now before we arrange a day and time to meet, we do need to collect a little information so that we are prepared for the visit. I have a few questions which shouldn’t take too long to answer.

1. First up could I please confirm what degree of hearing loss [child] has?
   Confirm which ear, if they give a range, e.g. moderate to severe on the left ear, enter as the lesser degree of loss (i.e. moderate)
   If answer is both ears mild or moderate (any combination) then proceed with questions 2-6, otherwise jump to question 7.

2. And does [child] wear hearing aids?
   Confirm if bilateral or unilateral fitted, if has cochlear implant (CI) then we don’t need any further hearing information, proceed to booking on the spot (Question 7 onwards).
   If child is unfitted (i.e. no hearing aid or cochlear implant), we need to finish all questions with appropriate modifications and omissions, to determine where hearing is measured (may be Australian Hearing or could be a regular centre like a hospital).

3. Where does [child] go to have their hearing and hearing aids checked?
   Answer should be Australian Hearing, need to narrow down the specific centre.
4. You wouldn’t by any chance know the name of the audiologist who [child] sees? 
   *No fuss if they don’t know. (Not for use when obtaining audiogram, but for use by PC later)*

5. Do you remember when your last appointment at Australian Hearing was? 
   *Seeking a date, for use by PC later*

6. Do you know when the next appointment is scheduled for? 
   *Seeking a date, for use by PC later*

7. Thanks very much for that information, now for our assessment, would you prefer us to 
   come out to you at home, or would you like to come into the Royal Children’s Hospital? 

8. And in general, is there a particular day of the week or time of day that would suit you best? 
   *Have usual booking date and time discussion*

9. And will [child name] need an Auslan interpreter to complete the activities?

10. Does [child name] currently attend any Early Intervention services such as speech/language 
    therapy? 
    *If yes, have they completed any assessments/tests lately and who with? Seek appropriate 
     details to enable follow up.*

11. Is [child name] completing assessments with LOCHI in the near future? If yes, then we need 
    to call the LOCHI team to follow up re: e.g., CELF **

12. And is [child name] undergoing a funding application currently? 
    *If yes: we need to know contact details of the person at the school who is dealing with the 
     application, to check and see what assessments have been done/may be done in the future.*

13. And just checking, would there be a reasonably quiet place to do the assessment in that is 
    not too distracting for [child]? 
    *No fuss if they can’t promise that, but appreciate anything they can do to keep environment 
     ideal.*

14. In order for us to bring the correct materials, we would just like to know whether [child] has 
    any difficulties with: 
    - Sitting up at a table 
    - Any difficulties with vision 
    Are there any modifications that happen in the classroom to make it easier for 
    [child] to learn? We ask because we might be able to include these when we are 
    working with him/her.

15. *If a home visit:** Okay the last thing that I need to do is check a couple of details with you 
    before we come out to your home. These are questions we ask everyone just to ensure our 
    researchers are safe at work, is that ok?
a. Are there generally any problems with finding the house & parking safely? Is access to the house ok?

b. Is there any difficulty with mobile phone coverage? (If they say YES) Is there a landline available just in case of emergency?

c. Are there any pets or animals (that we need to know about?)
   (Just in case you needed us to call ahead so you could put less-friendly pets away!)

15. If hearing aid user: And just a reminder that [child] will need to have their hearing aids for the assessment, just letting you know in case they are usually kept at school.

Thank you very much for all that information [parent name], and for your time today. We’ll send a pack out today which confirms the visit time and day, as well as some questionnaires that you can fill out either before or during the visit. It will also have our contact details in case you need them.

Okay! We’ll see you and [child name] on [date of assessment], and we’ll send you a reminder a day or so before we visit. Goodbye.

10.4 Phone Script: 9-12 Year Assessments

Hi. This is [your name] calling from the VicCHILD hearing research project at the Royal Children’s Hospital. Am I speaking with [parent name]? Is now a good time?

Yes

For some time now, you and [child’s name] have been taking part in the VicCHILD project. Now that [child’s name] is between nine and twelve years of age, we are hoping to complete a language and learning assessment with her/him. Collecting this type of information on the development of children who have a hearing loss is really valuable.

What we would like to do, is to come and see both [child name] and yourself at home, or alternatively you have the option of coming in to see us at the Royal Children’s Hospital. We expect to be with you for a few hours, during which you will complete some questionnaires about [child], and [child name] will do a few activities on language and learning which most children enjoy.

Do you have any questions for me so far? Are you happy for us to make a time now for us to complete the visit with you and your child?

Yes, happy to hear more

Great, let’s work out when we could come for the [assessment] visit. [QUESTIONS]

1. For our assessment, would you prefer us to come out to you at home, or would you like to come into the Royal Children’s Hospital?

2. And is there a particular day of the week or time of day that would suit you best? What time would suit? usual recr visit seating time discussion

3. And just checking, would there be a reasonably quiet place to do the assessment in that is not too distracting for [child]?
We are trying to improve our process for the study. Can I ask the reason you’re not keen on doing an assessment?

- Check if the child is in LOCHI. If so please state this clearly in the notes and let me know so that I can follow-up with the Victorian representative of the LOCHI study.

- Check if the child is engaged with a speech therapist. If so, obtain verbal permission from the parent to contact the speech therapist. If granted, get their contact details, and call the speech therapist to double check what assessments have been done and when, and what assessments are planned. By applying the rules that we used in L4L that Sheena decided on, we can repeat the CELF or a similar assessment if it has not been completed in the last 6 months. If the speech therapist was planning to complete a CELF with the child and we are booked into see the child AND if Mum has consented to exchange of information with health professionals, we can send a copy of the CELF record form with a template letter DIRECTLY to the speech therapist (see below).

- Check if the child is undergoing a funding application. If so get the contact details for the relevant person at the school, and call them to double check what assessments have been done and when, and what assessments are planned. By applying the rules that we used in L4L that Sheena decided on, we can repeat the CELF or a similar assessment if it has not been completed in the last 6 months. If the school professional was planning to complete a CELF with the child and we are booked into see the child, AND if Mum has consented to exchange of information with health professionals, we can send a copy of the CELF record form with a template letter DIRECTLY to the school professional (see below).

10.5 Children in other research/seeing health pros/completing funding apps

When booking in direct assessments, it is very important that we enquire with families about the following:

1. Are you and your child currently taking part in other research studies, for example the LOCHI study?

2. Is your child currently seeing a speech pathologist and/or psychologist, and have they had speech pathology and/or psychology assessments within the last 6 months?

3. Is your child currently participating in assessments as part of a funding application?

We need this because if the CELF-4 or the WNV have been administered in the last SIX months we do not need/want to re-administer these tests within the VicCHILD assessment battery. Additionally if the child is seeing a speech pathologist and/or psychologist, it may be helpful for such professionals to have copies of the VicCHILD assessment battery (NB: This requires parents to have consented to exchange of information).
In regards to the LOCHI study, as of 27/02/15 LOCHI are also visiting their participants who are in the age range of 9-12 years. Duplicated assessments between the VicCHILD and LOCHI assessment batteries include the CELF-4 and the WNV (identical subtests across both measures). Thus to avoid duplication and invalidating a measure that VicCHILD or LOCHI has administered, we routinely check in with Laura Button who is a Melbourne researcher working on the LOCHI study. This ensures that we are communicating prior the VicCHILD assessment, and thus will hopefully prevent double ups whilst also permitting exchange of information between the two studies and thus reducing parental burden. If a child is in LOCHI, please see procedures below.

***It is important that you only ever provide a copy of a child’s record forms and/or summary/standard scores to a qualified health professional such as a speech pathologist and/or psychologist. This ensures that only professionals who are trained in interpreting standardised tests see the child’s results, and second it protects the copyright of the content of the respective measures. See section 9.5.3 for specific details of what materials can be sent to whom.***

9.5.1 When a parent indicates that their child has had a ‘recent’ assessment

- Update the appropriate REDCap fields to reflect this on the direct assessment booking page
- Ask the parent what assessment has been completed and when it was completed (a lot of parents are not able to provide accurate information about this). In most instances we will then need to contact the child’s Speech Pathologist and/or Psychologist to check if the CELF or the WNV were administered within a period of SIX months.
- Visually check the REDCap database to ensure that the parent/guardian has provided consent to “Exchange of Information”. On the parent information statement (Fig 3), exchange of information is defined as:

**Exchange of Information**

Sometimes, two agencies may ask to see your child around the same time. For instance, we might contact you soon after a school entry assessment. Or a secondary school might request results from the VicCHILD 10-12 year old assessment. To avoid multiple assessments, you can let us exchange results with other professionals (like speech pathologists, audiologists, and teachers). This might be by letter or phone.

- If a parent has consented to exchange of information, please ask the parent to provide the speech pathologist’s and/or psychologist’s name and contact details, and record these in the relevant REDCap notes section.
- You are then allowed to contact the speech pathologist/psychologist that the parent has nominated. Before discussing the child with you, the speech pathologist may want to see a copy of the consent form so feel free to email or fax a copy to them.

- During the phone call to the speech pathologist please check that it was the CELF-4 that was administered and it was definitely within the last six months. Please also check what subtests were administered to ensure that we are collecting all of the relevant information that VicCHILD requires.

  - If a CELF-4 WAS administered in the last six months, please request a copy of the entire record form (not a summary- it must be a copy of the actual record form). This can be emailed or faxed to the VicCHILD team. Once received, please give a copy of the form to the RA who will be completing the assessment with this child. Please make sure that the RA is aware that they do not need to re-administer the CELF-4. Please make all progress notes in the database, including current status i.e. I'm waiting to receive copy of CELF-4 Record Form from Speech Pathologist (Date and Initials); or CELF-4 Record Form received and passed to RA name who is completing the assessment (Date and Initials)

  - If the CELF-4 WAS NOT administered in the last six months, please let the speech pathologist know that we intend to complete one and that we can send them a copy of the record form or a summary of the standard scores and percentiles- whatever the clinician would prefer (refer to section 9.5.3 for protocols re sending results)

If the parent indicates that the child has had assessments but is unsure what and when

  - If the parent/guardian has provided consent to exchange information, you should then contact the speech pathologist/psychologist to find out what assessments have been completed and when.

  - Then follow the protocols as described above.

If the parent indicates that the CELF-4 hasn't been completed but would like a copy sent to the child's speech pathologist

  - If the parent/guardian has provided consent to exchange information, you should then contact the speech pathologist to discuss the child's role in VicCHILD and sending them a copy of the child's CELF results.

  - Refer to section 9.5.3 for protocols re sending results.
If the parent indicates that she would like the results of her child’s language assessment shared with a speech pathologist/psychologist or another professional at their child’s school

- If the parent/guardian has provided consent to exchange information, then you are able to share the child’s results with another health professional. HOWEVER, record forms should only ever be sent to speech pathologists or psychologists

- If a parent feels strongly that a school professional should have a copy of the results, then we are able to send a summary of the scores which details the percentile results for the measures requested.

- Refer to section 9.5.3 for protocols re sending results

9.5.2 When a child is taking part in the LOCHI study

If the parent/guardian has provided consent to exchange information, please

- Complete the following template and email to ST as soon as possible. ST routinely emails Laura (laura.button@hearing.com.au) on a weekly basis and Laura then confirms if the child is in LOCHI and when they had their last assessment

<table>
<thead>
<tr>
<th>Name</th>
<th>VicCHILD ID</th>
<th>Notes</th>
<th>State</th>
<th>VicCHILD RA scheduled for Visit &amp; Date of Visit</th>
<th>In LOCHI or NOT</th>
<th>Next Step</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

9.5.3 Sending VicCHILD results to external professionals/research studies

General notes

- Copies of record forms and/or summaries of percentiles **should NOT** be emailed. They must all be mailed by registered post only.

- For any record forms that are shared, you must black out the child’s VicCHILD ID number

- In terms of what data we share with other professionals, the default is the CELF measure only, however if parent’s have provided consent for data exchange and other measures are requested then this is fine too.

1. **For Speech Pathologists and/or Psychologists**

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2. For Other Health Professionals (this includes School Principals and Teacher of the Deaf)
   - Do not send a copy of the record form, please use template below and only provide percentiles
   - Cover Letter & Results Template (delete tests that are not required).
   - Use a registered post envelope to send the results (please don’t email)

3. For Other Health Professionals (this includes School Principals and Teacher of the Deaf)
   - Cover Letter:
   - Use a registered post envelope to send the results (please don’t email)

9.6 Children who require signing interpreters

Please update the relevant RedCAP variables accordingly.
The following template then needs to be complete and emailed from the VicCHILD email address to: rch.interpreters@rch.org.au

<table>
<thead>
<tr>
<th>Date</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>Time</td>
<td></td>
</tr>
<tr>
<td>Name</td>
<td></td>
</tr>
<tr>
<td>U/R Number OR Date of Birth (if U/R is not known)</td>
<td></td>
</tr>
<tr>
<td>Language</td>
<td></td>
</tr>
<tr>
<td>Clinic &amp; Location</td>
<td></td>
</tr>
<tr>
<td>Doctor or Professional</td>
<td></td>
</tr>
<tr>
<td>Estimated length of appointment</td>
<td></td>
</tr>
<tr>
<td>Please confirm booking has been made to: Name</td>
<td></td>
</tr>
<tr>
<td>Cost Centre</td>
<td>0136</td>
</tr>
</tbody>
</table>

9.7 Information sent to families when a direct assessment is scheduled

9.7.1 Cover letters: 5-7 and 9-12 assessments

- The cover letter can be merged utilizing the standard mail merge template. Please use the mail merge function as the letters need to be identical for all families.

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• The cover letter contains two blue highlighted fields: a) him/her please change child’s
gender as appropriate (i.e. delete the incorrect gender); and b) Home Visit/The MCRI/Royal
Children’s Hospital please change as appropriate to the booking (i.e. just delete the incorrect
location)
• When merged, the date and time are not very user-friendly. Thus please change the date to
the following format “Tuesday 2nd December, 2014”; and the time to “2:00pm”.

9.7.2 Measures to be included in the 5-7 year direct assessment pack

- VicCHILD 5-7 Questionnaire
- AQQL-4D (Parent/Guardian)
- Mill Hill A (Parent/Guardian)
- Mill Hill B (Partner - if Applicable)

9.7.3 Measures to be included in the 9-12 year direct assessment pack

- VicCHILD 8-10 OR 11-12 Questionnaire
- AQQL-4D (Parent/Guardian)
- Mill Hill A (Parent/Guardian)
- Mill Hill B (Partner - if Applicable)

9.8 MCRI Room Booking Procedures

• Using your Outlook account, enter your clinic or individual appointment on your calendar
• Click on the ‘Invite Attendees’ button
• Click on the ‘Rooms’ button and the list of 4 West rooms will appear (see screen shot below)
• Select your room, invite to the appointment and then click OK

You can then send your appointment which will go to Asha and Louise (Room Delegates) to confirm
the room booking. Please note: Your room is not ‘booked’ until you receive a confirmation email

To help you manage your clinic, you can enter as much or as little information to the appointment or
clinic as you wish, though remember the Room Delegates will be able to view that information.
10 Direct Assessment Procedures

10.1 Items Required for an assessment (5-7 & 9-12)

Before leaving for a visit, make sure your suitcase contains the following:

**KEY:**
- **green=5-7, red=9-12**

- Staff ID badge and “Working With Children Check”
- CELF 4 Stimulus Book
- CELF 4 Concepts and Following Directions laminated script
- WRAT Word Reading List [3-12]
- WRAT testing kit and Blue response forms (Maths, Spelling, Word Reading)
- Bus Story picture book [5-7]/ ERRNI Picture book [9-12]
- Bus story script [5-7]/ ERRNI script [5-12]
- Portable speakers
- CNRep [5-7, 9-12] & CTOPP [5-7] on the USN-S laptop
- NPVT iPAD
- GFTA stimulus book [5-7, 9-11]
- USN-S kit (laptop with soundcard and software; headphones.) [5-7 Mild/Moderate]
- Digital Notetaker (MP3 player USB)
- Stopwatch
- Causes of Hearing Loss laminated question card

- Additional questionnaires for parent and/or child completion (VicCHILD questionnaire 5-7, 8-10 or 11-12; Mill Hill A & B, CTRSD 9-12, PedsQL MFS [5-7], ACQOL-4D; CHPD [5-7 M/M]) or PEACH (all others). PedsQL Wellbeing [5-12], PedsQL Generic Core Scale Child-report [5-12]. NB: items in *bold* have not been sent out in advance

- Stickers and stamps
- Coloured pencils for drawing and colouring pages
- Stationery

**Spare Forms Folder**

Every suitcase should also have a spare forms folder containing:

- Spare copies of all visit parent questionnaires and assessment record forms
- Spare buccal and spit pots
• Reply paid envelopes with VicCHILD stamp (for returning any Parent Questionnaires)

**Things To Consider**

• Make sure that all of the record forms and questionnaires in the family visit pack contain the child’s ID number

• All families will receive a reminder of their visit (a phone call or SMS) the day before their assessment

• Check if the family has returned their parent questionnaire before you leave for the visit.

---

**10.2 Direct Assessment Flow**

Please wear MCRI ID tag and WWCC ID

↓

Greet parents & complete consent procedures for assessment (if not completed and obtained prior), and other consents necessary (e.g., dataashing only), and hearing loss causes (see laminated card)

↓

Ask parents which room would be the most appropriate to complete the assessment

↓

Ask parent to complete QUESTIONNAIRES

(For) We have a couple of brief questionnaires for you to complete while I am doing the activities with ______. (VicCHILD parent questionnaire; Mill Hill A (and B for other parent if there), AQOL-4D; CHILD AND PEACH [all others], PedsQL MFS proxy [5-7])

↓

(For) Now it’s time for me to start the activities with your child.

While I’m doing the activities, please try your best not to repeat or re-word (or translate) any of the instructions that I give your child, as we need to make sure that each child is given the activities in exactly the same way. The questions that I’m going to ask your child will increase in difficulty as we go on, so we don’t expect kids to get everything right.

If necessary engage in warm up play (either with the child’s toys or drawing together) or chat (For) Now we are going to do some listening and talking activities together. We will also be looking at lots of pictures. Every time we finish an activity we will put a sticker/stamp on this sheet.

When the page is full you will know you are all finished!
Complete the CELF-4 (CFD, RS, FS, WC subtests [all], also WS and SS [5-7]) ** not in Auslan

(Give the child a 2-5 minute break as needed)

Then complete 5-7 visit in the following order: Mo/Mo: NPVT ▶ ILSN ▶ CNReg ▶ WNV ▶ Bus Story ▶ CTOPP ▶ GFTA and Se/Prot: NPVT ▶ CNReg ▶ Bus Story ▶ WNV ▶ CTOPP ▶ GFTA

OR

Then complete 9.12 visit in the following order: ERRNI pt1 ▶ WNV ▶ GFTA ▶ WRAT ▶ CHU90 ▶ NPVT ▶ PedsQL Wellbeing ▶ PedsQL Core Child report

Thank the parent and child for their involvement in the study and for allowing us to visit them.

(Check Q's are completed and go through each with parent to ensure that there are no missing questions.) Final task is to take the spit pot sample.

If parents ask for a feedback letter, tell them we can send something about their child’s results within 4 wks.

10.2 Feedback Letters

For direct assessments completed as part of VICCHILD, we don’t routinely offer feedback/results letters to parents.

In instances where feedback is requested AND the complete CELF (i.e., all core receptive and expressive subtests) have been administered, then a brief feedback letter can be provided.

The template for this letter can be found:

\RCHFS1\emtse\3. RESEARCH UNIT\Hearing\VICCHILD\Restricted Access\Assessments\Feedback.doc

PLEASE NOTE: The for children who score below the 10th percentile on their receptive and/or expressive scores, please call the family first to inform them of this prior to sending out the feedback letter.

In instances where the full CELF is not administered, it is not possible to provide meaningful feedback in the context of the reduced measures. Thus please inform parents something along the following lines. Please also send them a list of speech pathology services in their area.

“As you will have noticed these are shortened/abbreviated tests that are meaningful for groups of children but not for individual children. Therefore we are not able to provide results for your individual child”.

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10.3 Parent Support

When parents specifically request information about parent support services in the context of having a child with a HI:

1. If young baby, Early Support Services:
   Victorian Infant Hearing Screening Program Early Support Service
   Ph: (03) 9345 4941
   Fax: (03) 9345 5049
   Email: eis.vihsp@rch.org.au

2. IF attending EI, seek assistance there

3. M&CH nurse (info at:

4. Information and resources:
   infanthearing.vihsp.org.au

5. Parent only resource - Aussie Deaf Kids (online support community, for parents by parents, no expert info/feedback etc):

For information on child health and stages:

6. RCH Kids Health Info (Family information materials) web site:
   http://www.rch.org.au/kidsinfoc/#tab-All

7. Educational Resource Centre have made the above site into an app:
   http://www.rch.org.au/khapp/

10.4 Tips for Standardised Assessments

Testing Environment

- Test administration should ideally be in a quiet, well-lit room that is free from interruptions and distractions.
- Advise the parent that it is important to refrain from talking, repeating or rewording questions or helping their child during test administration.
- Make sure any stimulus book being used is visible to both you and the child.
- During testing do not allow the child to see the record form or the examiner’s side of the stimulus book.
Rest periods/Breaks

- For 5-7yr olds and 9-12yr olds, as testing will be quite a long time, a break is recommended.
- If a break is needed earlier than the usual time point, schedule the break at the end of a test (or sub test), so as not to interrupt administration of the test.
- Discourage eating during the actual testing and encourage snacks to be saved until the break/ end of the appointment, where possible.
- While the child is taking a break, this is a good opportunity to answer any questions the parent has about the questionnaires.

Encouragement/Reinforcement

- It is important to maintain a rapport with the child, especially young children who are not familiar with testing situations.
- Establishing and maintaining a good rapport facilitates the child’s interest and cooperation during test administration.
- If the child is not comfortable, take time to engage him/her in play or discussion before beginning test administration.
- If the child is comfortable, you can begin test administration straight away.
- While administering a test, do not tell the child if their responses are right or wrong, or how many items they have answered correctly. (Unless otherwise stated on the scoring sheet as for some items of the CTOPP)
- Make general encouraging comments or reinforcing statements such as:
  
  “We’re almost done”

  “I like the way you’re working”

  “Good listening”

  “One more question and you get another sticker!”

  “Good try”

  “Good pointing”

10.5 Calculating Chronological Age

Before beginning any tests, complete the information about the child and the examiner on the front page of all record forms (i.e. child’s ID, child’s DOB, date of assessment and examiners initials). To
compute the child’s chronological age, subtract the child’s birth date from the test date. In doing so, you need to remember 2 points:

1. When borrowing days of the month, always borrow 30 days, regardless of the month
2. When borrowing months, always borrow 12 months.

<table>
<thead>
<tr>
<th>Year</th>
<th>Month</th>
<th>Day</th>
</tr>
</thead>
<tbody>
<tr>
<td>2004</td>
<td>1</td>
<td>16</td>
</tr>
<tr>
<td>1999</td>
<td>9</td>
<td>30</td>
</tr>
<tr>
<td>9</td>
<td>9</td>
<td>16</td>
</tr>
</tbody>
</table>

A child’s age is never rounded up to the next month or year. Chronological age is always rounded down to the previous month. Days are not considered. For example 7 yrs. 2 months, 29 days is considered 7 yrs. 2 months.

10.6 NESB children and families

- Please note: the measures that directly assess the children’s abilities (e.g., CELF, CTOPP) CANNOT be interpreted. All items must be delivered in English only.

- The two exceptions to this rule are as follows:

  1. Assistance to understand and complete forms: If the parent wants/needs assistance when completing either the questionnaires about the child (i.e., AQoL-4D) this is fine. You can give as much assistance as necessary to help the parent to understand this information.

  2. Copy: The concept of ‘copy’ can be explained/interpreted during the CELF Recalling Sentences subtest. For example, if a NESB child does not appear to understand the concept of ‘repeat’/‘copy’ after the RA has attempted the demo items with the child, then it is OK to ask the parent to explain the concept of ‘copy’ in their own language to their child, so that the child understands what they’re being asked to do. However, we can then only score responses in English that come out of the child’s mouth (i.e. we cannot score any responses interpreted or repeated by the parent, in either language).
It is important to outline to the parent at the beginning of the assessment that a parent cannot provide or translate any information in another language. If a parent does interpret an item then it cannot be scored.

10.7 Auslan speaking families and children

Families and children who require Auslan interpreters will complete almost all of the same assessments and questionnaires.

- Parents will complete the VicCHILD questionnaire, the Mill Hill, the AQOL-4D, and (for 5-7 only) the PedsQL-MP3 (proxy). Parents who are hearing impaired do not complete the CHILD or PEACH.
- Children who will use Auslan during the assessment can complete the Bus Story or ERRNI (and video it), the WNV, the WRAT [not Word Reading unless they are able to read the words in spoken English], and the CHU-9D and PedsQL Wellbeing and PedsQL Self-report Core questionnaires.

Please make sure that parents do not translate or interpret instructions or assessment content for their children, as these need to come from the interpreter (i.e. we cannot score any responses interpreted or repeated by the parent, in either language).

10.8 Using the Digital Note Taker – MP3

Charging the battery

The player’s battery is recharged while the player is connected to a running computer. Remove the USB cap and connect the USB connector to a USB port on your computer. When the remaining battery indication of the display shows “Full”, charging is complete.

Turning the player on

Make sure that the MP3 player is not “on hold” side of player. The player resumes the operation from the point at which you last turned it off.

Press the ‘Play’ button to turn on the player.

When recording a subtest

Please state the child’s ID number and the name of the subtest you are recording prior to the administering the subtest Items.
10.9 Parent Questionnaires

10.9.1 Mill Hill
This questionnaire assesses verbal ability in adults. Make sure the parent knows they need to circle the word in each group of 6 that is closest in meaning to the word in heavy type. Make sure they only circle one answer for each question and that they complete both sides of the page.

10.9.2 Assessment of Quality of Life (AQOL-4D)
This questionnaire asks the parent about their own health related quality of life.

10.9.3 CHILD (5-7, Mild to Severe)
This questionnaire asks the parent about their child’s hearing functioning, in 15 scenarios commonly encountered in their home environment. Ensure that the parent knows they do not have to try each situation with their child, but can recall how their child has behaved in these situations over the last week. Parents need to consider their child in each situation wearing their hearing aid if applicable. Parents need to rate each scenario described according to the levels 1-6 on the “Understand-O-Meter”, which describes how well they perceive their child’s hearing to be functioning. The parent must choose the level that describes their child’s ability most closely.

10.9.4 PEACH
This questionnaire asks the parent about their child’s hearing functioning. Parents need to rate their child’s listening behaviour over the past week on a Likert-type scale. The questions are designed for children wearing their hearing aids or cochlear implants. If the child has never been fitted with a hearing device, the parent can answer question 1 with never and proceed with the 11 remaining questions.

10.9.5 PedsQL MFS (5-7)
This questionnaire asks the parent about their child’s quality of life with respect to fatigue. Parents respond to 18 symptoms of fatigue when considering their child’s behaviour over the last one month, using a Likert-type scale. Parents must complete all three sub-sections.

10.10 Child Assessments

10.10.1 Clinical Evaluation of Language Fundamentals (CELF-4)
The CELF 4 measures a broad range of expressive and receptive language skills.

CELF administration for 5-7 year olds involves 6 subtests:
○ Concepts and following directions
○ Word structure
○ Recalling sentences  **iPad version  **record
○ Formulated Sentences  **record
○ Word Classes 1 (receptive component only)
○ Sentence structure  **record

CELF administration for 9-12 year olds involves 4 subtests:

○ Concepts and following directions
○ Recalling sentences  **iPad version  **record
○ Formulated Sentences  **record
○ Word Classes 2 (receptive and expressive)  **record

Scores on these subtests provides an overall score of Language Ability (Core Language) as well as scores on language facets; Receptive Language (language comprehension) and Expressive Language (language use).

Please Note: CELF-4 Administration

- For signing children subtests will be presented in sign language as well as aurally
- Refer to the stimulus manual and record form for all directions and stimulus questions.

<table>
<thead>
<tr>
<th>Subtest</th>
<th>Notes</th>
</tr>
</thead>
<tbody>
<tr>
<td>Recalling Sentences</td>
<td>Cannot be administered in Auslan (but signed English is OK, if child uses this)</td>
</tr>
<tr>
<td>Formulated Sentences</td>
<td></td>
</tr>
<tr>
<td>Word Classes- receptive &amp; expressive</td>
<td>Cannot be administered in Auslan (but signed English is OK, if child uses this)</td>
</tr>
<tr>
<td>Concepts and following directions</td>
<td>Cannot be administered in Auslan (but signed English is OK, if child uses this)</td>
</tr>
<tr>
<td>Word Structure</td>
<td>Cannot be administered in Auslan</td>
</tr>
<tr>
<td>Sentence Structure</td>
<td>Cannot be administered in Auslan</td>
</tr>
</tbody>
</table>
Important notes for CELF administration

- Administer the subtests in the order they appear on the record form.
- Before administering any subtest item, present the respective demonstration and trial items so the child can practice the task and become familiar with the nature of the stimuli.
- Trial items allow encouragement, demonstration, repetition and prompting required to elicit the targeted response.
- If a child revises a response or self-corrections before the next item is presented, score the revised response EVEN IF IT IS INCORRECT.
- If a subtest allows repetitions, repeat the stimulus question if the child requests repetition or does not respond within 10 seconds. Repeat stimulus ONLY ONCE.
- Each subtest has a discontinuance rule, where the subtest is ended after the child reaches a specified number of consecutive zero scores.
- For repetition and discontinuance rules, see specific subtest considerations below. Also see the stimulus manual and record form.

Specific Subtest Considerations

Concepts and following directions [NOT IN AUSLAN]

- To evaluate the child’s ability to interpret spoken directions of increasing length and complexity that contain concepts that require logical operations, and to remember the names, characteristics and order of mention of pictures.
- Try to get into the habit of using the stimulus sheet. It enables you to sit alongside the child and watch exactly what they are pointing to. You will need to read the script from the manual for the familiarisation items and trials as this information is not on the stimulus sheet.
- There are two familiarisation items, which ensure the child knows the names of the animals presented in the test items.
- If the child is unable to identify an animal correctly during the first presentation of the familiarisation item, repeat the animal name until the child is familiar with it and can readily point to it when requested.
- During the trial items it is important to train the children to understand the ‘go’ instruction and emphasise that the order of pointing is also important.
NO ADDITIONAL PROMPTING FOR THE CHILD TO “WAIT FOR GO” CAN BE GIVEN DURING THE TEST ITEMS

5-7yo take Set 1 Demo and Trials and start at Item #1 and must take Items s #1-#23; 9-12yo take Set 2 Demo and Trials and start at Item #24.

If a child responds to an instruction accurately but does not wait for ‘go’ then the item is recorded as incorrect.

Wait until you are certain that the child has completed their response to each item before presenting the next item.

No repetitions allowed. Before administering test items you can say to child “Listen carefully. I can only say this once”.

Discontinue after 7 consecutive zero scores. For 5-7yo, start counting zeros at Item #24.

2 familiarisation items, 2 trial items.

Word Structure [5-7]

- Evaluates the child’s ability to apply word structure rules (morphology) to mark inflections, derivations, and comparison, and to select and use appropriate pronouns to refer to people, objects and possessive relationships.

- Some questions have multiple correct responses allowed, listed in the stimulus book.

- If the child gives a response that is different from the target response but demonstrates the targeted structure and is meaningful to the context of the item, write the response on the record form and score as correct.

  **Example:** Targeted structure: Progressive (go)

  Correct Response: sleeping

  Child’s response: lying on the bed

- The targeted structure of each question is listed for each question in the stimulus book in black bold font.

- If the child gives a response that is related, but not identical to the target (e.g. ‘bad blown’ for irregular past ‘blew’) prompt one time with “Can you say it another way?” and repeat the item. Give the whole prompt again.

- If a child gives a response that is not listed as a correct response, and you are uncertain if it meets the above criteria, write the child’s response verbatim for later scoring considerations.
Start at item #1
Several items go over two pages (demonstration, then trial) — make sure you maintain continuity when delivering these items.
Repetition allowed.
Discontinue rules: none. Administer all items.
One demo item, 2 initial trial items, and 3 additional trial items for harder questions.

Recalling sentences (NOT IN AUSLAN, ***Record)
- Evaluates the child’s ability to (a) listen to spoken sentences of increasing length and complexity, and (b) repeat the sentences without changing word meanings, inflections, derivations or comparisons (morphology), or sentence structure (syntax).
- No repetitions allowed.
- 2 trial items.
- Use LSAC iPad version

I am going to say a sentence. I want you to listen carefully and repeat exactly what I say. Let’s try. [pause] My sister is in grade six.

Let’s try again. Listen carefully and say exactly what I say. [incorrect answer] You need to say “My sister is in grade six.”

Listen to another sentence and say exactly what I say. [pause] Does Mr Lopez teach reading?

Now let’s try some more. Remember to listen carefully and say exactly what I say.

- 5-7yo start at item #1; 9-12yo start at item #6
- The child’s response must be a repetition of the sentence VERBATIM.
- Recording and Scoring at the same time — please score according to the following guidelines:
  - Self correction by the child is ok.
  - Using “Um” is not an error.
  - Press the tick (✓) if the sentence is definitely right (it is said word for word)
  - Press the question mark (?) if unsure or the sentence is partially right (has a few [1-3] errors)
  - Press the cross (✗) if the sentence definitely has more than 4 errors

-1 error for each word that is omitted, repeated, added, substituted
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-1 error for each word or segment transposed (if the transposition doesn’t change the meaning of the sentence)
-2 errors for each word or segment transposed (if the transposition changes the meaning of the sentence)

c. This can be tricky as self-corrections, which may contain many word repetitions, are not errors

e. **Item 3.** Did you remember to bring your lunch?

c. “Did you remember to bring your food, I mean lunch?” = no errors

c. “Did you remember to bring your food, lunch?” = no errors

e. **Item 8.** The big brown dog ate all of the cat’s food.

c. “The big brown dog ate all of the cats food.” = no errors

e. **Item 13.** The book was not returned to the library by the teacher

c. “The book was not returned by the teacher” = no errors (disregard the initial part before the self-correction)

c. Therefore, only press cross (x) if the sentence **doesn’t sound at all like the original version**, or the child has said ‘I’m not sure’

c. The test will discontinue when the cross has been selected 3 times

c. When in doubt, press the question mark (?)

c. The main aim is to finish administering the test NOT to mark it. If considering what to press take too long please press **d.e.t**.

Formulated sentences (NOT IN AUSLAN, ***Record***)

c. Evaluates the examinee’s ability to formulate complete, semantically and grammatically correct spoken sentences of increasing length and complexity using given words and contextual constraints imposed by illustrations.

c. 5-7yo start Set 1 at Item #1-23; 9-12yo start Set 2 at Item #8-28

c. Recording and Scoring at the same time: please always record recalling sentences using your USB and also attempt to score at the same time just in case the recording doesn’t work.

c. The child’s response must contain the word or phrase stimulus, with no change to tense/plurality (excluding colloquialisms), no change to semantics, and must be about something in the picture.

c. Child’s response can be first person, animal as speaker, can be in question or statement form, can be a dialogue between people in the picture.
Sentence Structure

- Evaluates child's ability to interpret spoken sentences of increasing length and complexity.
- Start at Item #1
- Repetitions allowed.
- Discordant rules: none. Administer all items
- One demo item and 2 trial items.

10.10.2 Wechsler Non-Verbal Scale of Ability (WNV)

The WNV measures general cognitive ability using nonverbal subtests in children 4y to 31y5sm.

Verbal requirements are minimized by using pictorial directions to communicate the demands of the subtests, with additional brief verbal prompts permitted.

VicCHILD is using the 1-subtest battery of.
- Matrices and Recognition subtests for the 5-7yo visits
- Matrices and Spatial Span subtests for the 9-12yo.

Administration

- Administer both subtests in 1 session. If not feasible, schedule 2nd session ASAP e.g. within 1 week
- Place the closed stimulus book on the table with the coil-bound edge toward the examinee and the Spatial Span board with numbers facing you.
- You must be sitting opposite the child for each subtest
- Ensure the stimulus book is flat and at a distance that enables the child to point to response options
- To ensure standard administration do not allow child to turn pages in stimulus book
- Keep stopwatch out of child's view on the table out of their reach
- Maintain a clear view of the stimulus book in order to gesture to the pictorial directions and observe child's responses.

Pictorial directions and gestures

- Pictorial directions provide a non-verbal, engaging method of communicating task requirements. They illustrate what the examiner and examinee will do during subtest administration
Administration of pictorial directions involves several gestures. Become familiar with the following standard phrases and descriptions of administration gestures:

<table>
<thead>
<tr>
<th>Phrase</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sweep your hand</td>
<td>Guide your hand, with your palm upward, in a line just above the position(s)</td>
</tr>
<tr>
<td>Drag your finger</td>
<td>Move your finger along the page</td>
</tr>
<tr>
<td>Point</td>
<td>Briefly touch or hold your finger above the indicated position</td>
</tr>
</tbody>
</table>

- If the pictorial directions and supplemental verbal directions are insufficient, provide additional help to ensure that they understand the task requirements. Do not proceed to administer the test items until you are sure the examinee understands.
- It is not appropriate to instruct the child on different methods that could be used to determine the correct answer to Matrices items but is ok to help them learn that their task is to select one option that goes into the space labelled with question mark.
- If child refuses to respond to the pictorial directions or sample items on one of the subtests you can temporarily suspend the administration and proceed to the next. Return to the previous subtest when they are more engaged and have obtained some degree of success in the test situation.

**Encouraging Responses**

- Time prompt: if no response within 30 secs gesture to them to encourage them to respond.
- Do not use 30 sec guide rigidly. Adjust time prompts to examinee performance see p. 27 manual.

**Encouraging responses/responding to child test behaviours** with the following standard phrases:

<table>
<thead>
<tr>
<th>Child test behaviour/response</th>
<th>Phrase to Use</th>
</tr>
</thead>
<tbody>
<tr>
<td>To encourage child</td>
<td>“You’re working hard”</td>
</tr>
<tr>
<td>If they try and answer before you have completed directions</td>
<td>“wait until I’m finished”</td>
</tr>
<tr>
<td>If they try to turn the stimulus book when solving</td>
<td>hold in place and say “don’t turn the book”</td>
</tr>
<tr>
<td>--------------------------------------------------</td>
<td>---------------------------------------------</td>
</tr>
<tr>
<td>If they say they can’t perform a task</td>
<td>“Just try your best”</td>
</tr>
<tr>
<td>If the child asks for your help</td>
<td>“You do it”</td>
</tr>
<tr>
<td></td>
<td>“You need to do it yourself”</td>
</tr>
<tr>
<td>To restore confidence (if they perform poorly on an entire subtest and are aware of it)</td>
<td>“That was a hard one! ” “Let’s do something else”</td>
</tr>
</tbody>
</table>

**Recording Responses**

- Make an entry on the record form for all administered items, to distinguish them from items that were not administered (score, checkmark or examinee’s response). Note verbal and nonverbal responses.
- Recommended abbreviations for recording a child’s response are:

<table>
<thead>
<tr>
<th>Abbreviation</th>
<th>Use</th>
</tr>
</thead>
<tbody>
<tr>
<td>R</td>
<td>Examiner repeated item or Instructions or examinee requested repetition</td>
</tr>
<tr>
<td>SC</td>
<td>Self-corrected</td>
</tr>
<tr>
<td>DK</td>
<td>Stated that they did not know the answer</td>
</tr>
<tr>
<td>NR</td>
<td>No response</td>
</tr>
</tbody>
</table>

**Start Points**

Begin at age-specific start point (inclusive i.e. 4-5 includes those up to 5/11m)

**Introduction**

“You will be doing several different things today. I will show you some pictures that will help you understand what to do. Look carefully to see what the children in the pictures are doing. That will show you what to do. You can also ask me questions.” (see p. 39)
Establishing a Basal Set (perfect scores on 2 consecutive items)

- If perfect scores achieved on first two items administered then basal has been established. Award full credit for non-administered items prior to the start point.

- If they obtain a perfect score on the first item administered but not the second, count the first toward the reversal criterion of two consecutive perfect scores and administer reversal items in reverse sequence until achieve perfect scores on two consecutive items.

- If no perfect score obtained on either the first or second item, administer reversal items in reverse sequence until achieve perfect scores on two consecutive items.

- Then resume testing forwards.

Giving Credit to Non-administered Items

- To distinguish between non-administered items for which credit was given and items actually administered, draw a slash over the non-administered items and write the score next to the item prior to the start point.

- If start on item prior to a child’s age-specific start point (e.g. suspected of cognitive deficiency), regardless of their performance on items preceding age-specific start point, full credit is awarded for preceding items if perfect scores are obtained on the age-specific start point and subsequent item.

Matrices

- Child looks at an incomplete matrix and selects the missing portion from 4 to 5 options.

- Do all Demo and Sample items. Start at item #1 for 5-7, start at item #7 for 9-12yo.

- The discontinue rule for Matrices and Recognition subtests is 4 scores of 0 on 5 consecutive items (i.e. discontinue after 4 consecutive scores of 0, or after 4 scores of 0 on 5 consecutive items).

- Scores on items administered in reverse sequence count towards the discontinue rules.

Recognition

- Child looks at geometric designs for 3 seconds and identifies option out of 4 or 5.

- Use stopwatch to ensure precise timing of stimulus exposures (3 secs).

- No strict time limits for responses (10-30s should be sufficient). Scoring based on response accuracy.
• After Demo and Sample items, 5yo start at Item #1; 6-7yo start at Item #4

• The discontinue rule for Matrices and Recognition subtests is 4 scores of 0 on 5 consecutive items (i.e. discontinue after 4 consecutive scores of 0, or after 4 scores of 0 on 5 consecutive items).

• Scores on items administered in reverse sequence count towards the discontinue rules.

Spatial Span (Forwards and Backwards)

• Child watches examiner tap a series of blocks and then taps them her/himself in either the same order or backwards order to the examiner.

• Remove pictorial instructions before starting the Spatial Span Sample item.

• No strict time limits for responses (10-30s should be sufficient). Scoring based on response accuracy.

• The discontinue rule for Spatial Span is after scores of zero on both trials of an item (each item = 2 trials).

• Scores on items administered in reverse sequence count towards the discontinue rules.

• Begin with Demonstration Item (2 trials), then Sample Item (2 trials), then Item #1.

• Administer Spatial Span backwards regardless of child’s performance on Spatial Span Forwards.

• In Auslan, use the instructions “touch the same ones I touched, in the reverse order”. This seems to translate more clearly than “only backwards”

10.10.3 The NIH Picture Vocabulary Test (NPVT)

The NPVT is used to measure receptive vocabulary.

• The child is asked to touch the picture on an iPad (out of 4 options) which represents the word they hear e.g., “peeking”

• Automated scoring and discontinue procedure; this does not require the RA to administer the items or to keep score.

• Once you have securely backed up the Recalling Sentences audio and the NPVT results, you can delete those participants from the ALT app.

• From the results page of the ALT app, touch the participant’s ID and swipe left. The option to Delete will appear in red. Press Delete.
Set-up

- Unlock the iPad — Kit 2 [pad pass code is 8229]
- Select the ALT app
- Press New Test +
- Select ______ (top left of the screen)
- Press on the text Participant Identifier and type in the participant’s ID
- Press Return on the keyboard
- Select the participant’s Date of Birth from the scroll
- Press Validate (bottom left of the screen) and ensure that it says participant found
- Get back to the main screen by pressing the iPad’s central button
- Open up ALT-NIH
- Press LAST SCORE
  (If there are previous scores, press CLEAR SCORE)
  (Press LAST SCORE again to ensure that it is blank or has null on the screen)

PLEASE NOTE** It is imperative that you clear the score before beginning the test:
- Press START (top right)
- Select the child’s age
- Press BEGIN
- Select the participant’s highest grade at school (3rd Grade)
- Press OK under NIH Toolbox Picture Vocabulary Test

Administration

To begin the test, the child must respond correctly and without help to at least 2 of the training items.

“You are going to see some pictures and hear some words. Press on the picture that you think best represents the word that you hear. Make sure you have a good look at each picture before you decide, and press the picture you have selected only once. If you’re not sure, have a guess. To hear the word again, press the play button. Let’s try two practice questions.”

- [Examiner presses OK]
- If the participant gives an incorrect response on either practice question, an ‘Incorrect Response’ message comes up. Press ‘OK’ and say “This is the banana [point], can you press on the picture of the banana”
“Now we’re going to do some more. Some of the words will be easier and some will be more difficult. Just try your best on each question. If you press a picture but nothing happens, wait a few seconds before trying again [demonstrate pressing finger on table]. If you keep pressing [demonstrate pressing finger repeatedly on table] it will skip through questions.”

- [Examiner presses OK]
- No response after 10 seconds: Press the play button to replay the word. Say “If you’re not sure, just take a guess”
- Child can press repeat (the play button) as many times as they like to hear the word stimulus

**Scoring**

- Select the entire scoring text by pressing down on the white space to the right of the text until it is all highlighted in blue
- Press COPY
- Get back to the main screen by pressing the iPad’s central button
- Press ALT (the participant’s ID and DOB should already be entered, double check that it is correct)
- Press down on the large white square (under the line below the words: paste the results) until the word Paste shows up
- Press Paste so that the participant’s score appears

**PLEASE NOTE** It is imperative that these scores are the exact scores from the ALT-NIH app. It is possible to have pasted the results from the previous child if the results were not adequately copied

- Press Save Results (bottom right of the screen)
- Press the back button (top left of the screen)
- Press Participants
- Select the participant’s ID on the left of the screen
- Write their score down on the Data Collection Summary Sheet. Their score is found in the circle above the word Score which is on the top right of the screen. The score ranges from -10 to 10. It can therefore be a negative number.

**PLEASE NOTE** Remember to include the negative sign if the score has a negative sign in front of the number.

- Once you have copied the results into ALT and have noted down the right score, go back to the ALT-NIH app and press CLEAR SCORE so that the test is ready for the next participant
- Remember to back up the score by emailing it to yourself (instructions below)
NOTE: the participant’s overall test score is the last Theta score in column 3. If there is ever a problem with the ALT app you can write the participant’s score down from the output you get from the NIH-ALT app directly following the test. However, this may be tricky because if the participant answered the last question incorrectly their overall Theta score shifts into the 4th column and does not appear to be in the Theta column. If you encounter any problems copying the participant’s results into the ALT app, follow the directions below.

- In the ALT app results screen, find and select the relevant participant ID from the left of the screen.
- The NPVT score will be displayed under the NPVT section on the right side of the screen.
- Take note of whether it is a positive or negative number.

Alternative scoring method (use if the method above fails):

If the ALT app ever fails, please copy and paste the score into Notes, following the directions below.

- Press the app NOTES.
- Write the date, participant number and their DOB.
- Double press on the yellow note and press PASTE under the participant’s number.
- Write the last Theta score on the Data Collection Summary Sheet, under NPVT. This is usually in column 3 but may be in column 4 if the last answer is incorrect.
- Email the note to yourself when you get back to the MCRI in order to back up the data.

Backing up the score:

This must be done as soon as you have internet access. You have access to the internet at the MCRI. The iPads automatically connect to the internet when you are in the office.

10.10.3 Expression, Reception and Recall of Narrative Instrument (ERRNI; 9-12)

- The ERRNI measures a person’s expressive language and story comprehension through their ability to relate a story, comprehend it, and remember it after a delay.
- In the research context it provides an index of syntactic maturity and narrative skill.
- Two parallel versions of the ERRNI: Fish Story and Beach Story (allows for retesting while minimising practice effects). We will do the Beach Story in this round of assessments.
- Both stories have a false belief and comprehension questions to check whether the examinee has understood this.
10.10.4 **LSN-S (5-7)**

The LSN-S is a test of hearing functioning in specified noise, conducted under headphones. The task involves verbal repetition of a target sentence, when listening in the presence of non-target, distracting speakers. The child will complete two of the four sub-tests, namely the:

- DV-30 or high cue condition, and
- SV-0 or low cue condition

This test uses Australian English, and requires three demographic fields to be completed before the test can begin. These demographic details should be pre-loaded in the software prior to the visit.

- Child first name
- Child family name
- Child date of birth

If the child is a hearing aid user, they must complete the task with their audiogram entered into the Prescribed Gain Amplifier (PGA), which will replicate their aided hearing when wearing headphones. Due to this requirement, it is essential that their audiogram be obtained prior to the assessment visit. This should also be entered into the system prior to the visit.

***If the child has cochlear implants, they are not able to complete the LSN-S***

**Set-up**

- Headphones with soundcard attached must be connected to computer via USB in order to launch LSN-S software.

- Mandatory fields to commence testing are “Last name”, “First name”, and “Date of Birth”.

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• For VicCHILD, enter VicCHILD ID number into both name fields, using the child's correct date of birth.

• Press the “Add” button and the participant's details should appear in the bottom half of the screen under the “all” tab.

• Use the blue arrow on the left side of the screen labelled “Next” to proceed.

Client Session

The next screen encountered is a list of all LCN-5 sessions that have been performed under this participant ID number. This is the screen that you would open if you wanted to review past results. For any new test, this screen requires you to either choose or enter your name in the “Tester” box.

• Press the “New” button on the right hand side of the screen, confirm that English (Australian) is the selected language, then either write or select the tester name and validate by pressing the “Ok” button.

At this stage, a pop-up window will appear asking whether you wish to use the Prescribed Gain Amplifier (PGA).

• Answer “No” if the child does not wear hearing aids.

• Answer “Yes” if the child does wear hearing aids.
The process for administering the LiSN-S deviates at this point, into what will be referred to as the "unaided stream" and the "aided stream". Please note: for BOTH streams, if the child's date of birth is under 6 years 0 months, a pop-up window will warn that norms are not available for this age. Always press "Ok" and proceed through this screen. You would then press the blue "Next" arrow to continue through to the Diagnostic screen.

Unaided stream

By selecting "No" to the PGA pop-up question, your next screen will be the Diagnostic screen where testing occurs, once you've pressed the "Next" arrow. For further instructions, refer to section "Diagnostic Screen".

Aided stream

By selecting "Yes" to the PGA pop-up question, you will be prompted to enter audiogram data for the PGA on the next screen. By selecting "Use Last PGA" to the PGA pop-up question, you will be confirming that any previous audiogram entered for the participant is the one to be used. This is what you would see if the audiogram had been pre-loaded before your visit and it is ok to choose "Yes".

In order to enter the audiogram, this data needs to have been obtained from Australian Hearing prior to the assessment (if consent to date linkage exists, otherwise it will be entered from a parent-supplied audiogram at the time of assessment). The audiogram should be pre-loaded into the software before the visit.
To enter the audiogram data, click directly onto the chart, separated by ear. Ensure the “AC” button is active for input of air conduction values (red circle for Right Ear; blue cross for Left Ear).

Rules for entering audiometric data:

- **Thresholds are recorded** in 5dB increments, and need to be entered for all frequencies listed from 250Hz to 8000Hz. The audiograms obtained from Australian Hearing may not have all frequencies required by the software completed. In these instances, the following two rules are applied:

  1. If the missing threshold is between two completed frequencies, the missing threshold is the average of the neighbours (e.g., 2000Hz is 25 dBHL, 4000Hz is 45 dBHL, the missing 3000Hz is entered as 35dBHL).

  2. If the missing threshold does not have a neighbouring frequency entered on both sides (e.g., audiogram complete from 250Hz to 4000Hz, 6000Hz and 8000Hz missing), the thresholds are entered as the same as the last completed threshold (e.g., in the example before, 6kHz and 8kHz are entered as the same value as 4kHz).

For unilateral losses, leave the unaffected ear at the default setting of 0 dBHL, for all frequencies.

- When you are finished press the “Save” button, then press the “Next” button which will take you through to the Diagnostic Screen, where you will see a progress bar while the sound file for the PGA is adjusted.
**Diagnostic Screen:**

![Diagnostic Screen](image)

**Unaided stream**

Ensure that the “Different Voices ≤90°” is selected in the menu on the left side of the screen. Give participant instructions for task and then put headphones on.

**Instructions for the “Different Voices ≤90°”**

Accompany these instructions with visual cues to aid the participant’s understanding (e.g., indicating sounds coming from in front, not worrying about the sounds from the side):

- This next task is a listening one where you wear these comfy headphones (and you do it with your hearing aids off – if applicable)
- You are going to hear a lady’s voice saying some sentences. It will sound like she is speaking in front of you. There is a beep before each sentence. Your job is to listen to the sentence and then tell me what you heard.
At the same time, there are going to be some other people talking at the side, but don't worry about them, just listen to the lady who speaks after the beep.
It is really important that you tell me everything that you heard the lady say, even if you didn't hear it all. It will get tricky, but that is ok, just do your best.

- By pressing “start” on the top right of the screen, the competing speakers commence.
- Press the blue “next” button at the bottom right of the screen to present the first target sentence. The target sentence is delivered while the following symbol is on the screen. Once this symbol disappears, you can ask the child for what they heard the lady say if they haven't told you already.

For each subtest, there is a minimum delivery of 22 sentences, with a maximum of 30 sentences. Scoring all occurs online, with the level of the next sentence dictated by the score achieved on the previous. It is important to keep giving motivational praise to ensure continued compliance with the test. Phrases such as “good work” “well done” “ok next one!” “that was tricky, here is an easier one” work well.

**Scoring**

Scoring is a count of the number of words correct per sentence, with the following guidelines:

- A word is considered correct if it is repeated exactly as it appears on the playback screen
- If an extra word is included, there is no penalty
- If a substitution, omission or plurality occurs, the number of correct words is adjusted accordingly
- Word order within the sentences does not matter, score is not influenced by word placement.
• Select the number of words correct, then press "next" and proceed until the test informs you that you have finished the Different Voice ±90° testing.

• Then choose the subtest condition "Same Voice ±0°" from the menu on the left side of the screen. You will need to re-instruct with instructions specific to this subtest.

Instructions for the "Same Voice ±0°"

For the next part, things are going to change a little!
Those other people talking at the side are now going to sound like they are in front of you, as well as the lady you are listening to.
All you need to do is keep listening for the beep, and tell me what the lady says after that.
Ok? Let's give it a try...
• Score as per the previous subtest.
• Once the testing has completed, proceed to the detailed results page.

Aided stream

• Use the same instructions as for the unaided stream for both subtests to be performed, but deliver the instructions with the child wearing their hearing aids. This necessitates the child putting their hearing aids back on in between the different voice and same voice subtests.
Please be aware that you will be trying to encourage a child with hearing impairment wearing headphones, so loud voice may be required and more visual cues.

- In the screen for the aided stream, only the “Different Voices ±50” subtest appears. That is because you must complete the “Quick LBM S PTA test” section before you can access the “Same Voice ±0” in the “Full LBM S PTA test” section.

Throughout the testing, in both the aided and unaided stream, the goal for each subtest is to obtain a speech reception threshold (SRT). If the test needs to be aborted before stopping automatically, it is necessary to record the SRT and the number of sentences that were successfully delivered up to the point where the test was abandoned. For example, in the example below you would record 30 sentences completed (labelled on screen as “Seq”) and a SRT of -3.9 dB on the assessment data collection sheet. If this is not recorded manually, it cannot be retrieved at a later date (in contrast to when the test is run until completion).

![Image](image)

If there is no problem with compliance with the entire test, then you can obtain the required results from the Detailed results screen.

**Detailed results screen**

![Image](image)

From this detailed results screen you can obtain the following information for both the “Different Voice ±50” (labelled on this screen as High-Cue SRT) and “Same Voice ±0” (Low-Cue SRT):...
• Client’s Score (dB) – this is the SRT that is displayed at the end of each test condition in the LSNN-5.

• Variance from Average in StDev – this indicates how far, in standard deviations, this child’s score deviates from the average score for age.

Note you must leave the Phonak Headphones in until after you have exited the program or it will not quit.

Frequently Asked Questions

Q. How did my child score on the LSNN-5? What is his/her hearing like?

A. This isn’t a routine hearing test, and the results take a little while to calculate and interpret. What we are hoping to measure is how child performs in different listening conditions, and these can’t be easily reported for an individual child, they need to be looked at once we have finished assessments on all children.

10.10.5 The Wide Range Achievement Test (WRAT; 9-12)

The WRAT is an achievement test which tests an individual’s ability to read words, comprehend sentences, spell and compute mathematical problems. We will be administering 3 subtests of the WRAT to the 9-12s: Word reading, Spelling, and Maths.

“5 rule” for Word Reading and Math Computation

Basal of 5: Min 5 items must be answered correctly in order to waive administration of 15 preliminary in each subtest (i.e., don’t get 5 right in Part 2 for WR or MC, have to go back and do Part 1)

“10 rule” for Word Reading

Discontinue rule for the WR subtest

<table>
<thead>
<tr>
<th>Examiner behaviour</th>
<th>Examiner response</th>
</tr>
</thead>
<tbody>
<tr>
<td>If the examinee asks if a response was correct</td>
<td>Say something similar to: “That’s fine”</td>
</tr>
<tr>
<td>If it’s obvious they can’t answer a difficult item</td>
<td>Say something similar to: “That was a hard one. Let’s try another!”</td>
</tr>
</tbody>
</table>

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• Materials: The Bus Story picture book, voice recorder, and laminated instruction card

• Record this subtest with the voice recorder. Video with the iPad if the test will be in Auslan, keep recorder on for audio of interpreter.

  o Place the picture book in front of the child so that it is visible to both of you
  o Open at the first page of pictures and let the child look at them for a few moments
  o Say “I’m going to tell you a story about this bus” (Point deliberately to each of the three pictures in order on Page 1 so that the child realises that the pictures are all referring to the same bus.) “Then, when I’m finished, I want you to tell me the story about this bus.” (Point to each of the three pictures again if you think it may be necessary.)
  o Tell the story, keeping strictly to the text, but adjusting the speed of your speech to match the child’s ability to concentrate
  o When you have finished the story telling, switch on the recorder and give the ID no. Turn back to page 1.

“How you tell me the story. Once upon a time there was a...” Turn the pages as the child tells the story. As soon as the child blocks, any prompting given should be minimal and indirect e.g. “And then...?” “So...?”

• If the child says no more, use a dash (·) after the last word spoken to indicate a block. Discontinue prompting, but carry on showing the pictures.
• Should the child spontaneously start again to tell the story, record the response for scoring; but if the child cannot or will not, say anymore, accept the situation without comment.

10.10.7 Comprehensive Test of Phonological Processing (CTOPP: 5-7, not in Auslan)

The CTOPP assesses phonological awareness, phonological memory and rapid naming. A deficit in one or more of these kinds of phonological processing abilities is viewed as the most common cause of learning disabilities in general, and reading disabilities in particular. In addition to their role in learning to read, phonological processing abilities also support effective mathematical calculation, listening comprehension, and reading comprehension.

General Administration:
• Record this test

• Feedback: All of the subtests require the examiner to give feedback to the examinee during the administration of initial items. This is done to help the examinee learn to master the task while responding to the items.

• Discontinuation of Testing: Each subtest is discontinued when the child scores zero for three items in a row. The test is also discontinued with no score entered for the subtest if the child doesn’t get at least one of the first three items correct.

• Entry Points & Ceilings: For each examinee, regardless of age, the examiner begins administering every subtest with the first item. All of the subtests for the CTOPP-2 are terminated when the individual misses three items in succession. If for some reason items were given above the ceiling and a few were ‘passed’, these ‘passed’ items would be scored as incorrect.

Eision (5-7 year olds, ***Record this subtest)  

Measures the ability to remove phonological segments from spoken words to form other words

• Record correct items as 1 and incorrect items as 0. The correct responses are noted in the Profile/Examiner Record Booklet. The total raw score is the number of correct test items up to the ceiling.

• Administer the subtest until discontinuation rule is reached unless child does not answer at least one of the practise items correctly. Do not score if the child did not complete at least one of the first three items correctly.

• Discontinue subtest when child scores 3 consecutive zeros.

• Feedback may be given for the first 14 items only Feedback examples:

  That’s right, let’s try the next one.

  That’s not quite right, toothbrush without saying tooth is brush (see manual or record booklet for more examples of how to give feedback).

Directions

Let’s play a word game.

Say ‘toothbrush’. Now say ‘toothbrush’ without saying ‘tooth’.

Correct? -> That’s right. Let’s try to next one.

Incorrect? -> That’s not quite right. ‘Toothbrush’ without saying ‘tooth’ is ‘brush’.

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Let’s try some more.

...

Okay, now let’s try some where we take away smaller parts of the words.

Blending Words [5-7 year olds]

Measures the ability to synthesise sounds to form words

• Please have audio tracks 1-33 ready on the laptop

• The examiner receives 1 point for the correct response and 0 for incorrect. The total score for this subtest is the number of correct test items up to the ceiling.

• Administer the subtest until discontinuation rule is reached unless child does not answer at least one of the first three items correctly. Do not score if the child could not complete the first item correctly.

• Discontinue subtest when child scores 3 consecutive zeros.

• Feedback may be given for the first 12 items only. Feedback examples:
  o When you put cow-boy together it makes cowboy. You try it: cow-boy makes _____.
  o Try to say the sounds all together as a real word. (see manual or record booklet for more examples of how to give feedback).

Directions:

On this one, you will hear some words in small parts, one part at a time. I want you to listen carefully and then put these parts together to make a whole word. Ready? Let’s try one.

Correct? -> That’s right, let’s try the next one.

Incorrect? -> That’s not quite right. When you put “cow-boy” together, it makes “cowboy”. You try it: cow-boy makes ________?

Let’s try the next one.

....

Let’s try some more words. Each time, you will hear the word one part at a time. Listen carefully and put the parts together to make a whole word.

Correct? -> That’s right.
Incorrect? -> When you put pen-sel together, it makes ‘pencil’

...

Okay, now let's try some more where we say even smaller parts of words, one part at a time. I want you to listen carefully and then put these parts together to make a whole word.

Ready? Let's try one.

Sound Matching (5-6 year olds ONLY, don't administer to 7 year olds)

Measures the ability to select words with the same initial and final sounds

- Materials: Picture book
- The examinee receives 1 point for a correct response and 0 for incorrect.
- Discontinue when the examinee three items in a row OR if child does not answer at least one of the first three items correctly. Do not score if the child did not complete at least one of the first items correctly.
- Feedback may be given for subtest items 1-6 in Pt 1 and 14-19 in Pt 2 only (see manual or record booklet for examples of how to give feedback).
- Pause 1 sec after pronouncing the target word and before pronouncing the three alternative answers; point to the pictures as their names are pronounced.
- The total score is the number of correct test items up to the ceiling.

Directions:
We're going to play a game with words. I will show you pictures to help you remember the words.

Look at the first picture. This is a sock. Now look at these two pictures. This is a sun, and this is a bear. The word sock starts with the /s/ sound.

Which of these picture words starts with the /s/ sound like ‘sock’? sun or bear?

Correct? -> That’s right. ‘Sock’ and ‘sun’ start with the same sound, /s/. Let’s try the next one.

Incorrect? -> That’s not quite right. The answer is ‘sun, because ‘sock’ and ‘sun’ [emphasize the first sound] start with the same sound, /s/. Let’s try another one.

[etc for next 9 items]

...
We're going to change our word game. Look at the first picture. This is a can. Look at these two pictures: pot and sun. The word “can” ends with the /n/ sound.

[etc. for next 12 items]

Phoneme Isolation (7 year olds ONLY)

Measures the ability to isolate individual sounds within words

- The examinee receives 1 point for a correct response and 0 for incorrect.
- Follow instructions on Test Record form.
- Discontinue when the examinee three items in a row OR if child does not answer at least one of the first three items correctly. Do not score if the child did not complete at least one of the first items correctly.
- Feedback may be given for the subtest items 1-7 (Pt 1) and 17-23 (Pt 2) only (see manual or record booklet for examples of how to give feedback).
- The total score is the number of correct test items up to the ceiling.

Directions

Now we are going to do a word game where I will ask you to say parts of words.
The word “man” has three sounds, /m/ - /a/- /n/, “man”. What is the first sound – the one in the beginning – in the word “man”?

Correct? -> That’s right, the first sound in “man” is /m/.

Incorrect? -> That’s not quite right. The first sound in “man” is /m/.

No response? -> The first sound in “man” is /m/.

Let’s try some more.

[continue items 2-16]

Now let’s try a few words with more than three sounds.

[etc.]

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10.10.8 Children’s Test of Non-word Repetition (CNRep) Not in Auslan

The CNRep is a test of short-term phonological memory. It assesses basic abilities to process and learn language.

- Materials: CNRep audio cued up on the laptop, USB audio recorder & CNRep Score Sheet
- The child’s responses are audio recorded for scoring purposes. You should score online.
- If the child becomes restless, stop half way through the CNRep (after item 20) to reinforce to the child they are doing a good job and to keep trying. If a child shows no signs of fatigue or boredom it will not be necessary to take this small break. Use your judgement to decide.
- There are no repetitions allowed in this test. The only time the item/s can be repeated is if there is a major disruption that means the child was not able to hear the item.

Scoring

- You need to familiarize yourself with the items in order to decide whether the child’s repetition attempt is correct. In most cases it will be obvious if the repetition is correct or not. Incorrect attempts may be inaccurate in a number of ways. For example:
  - The child might omit one or more phonemes from the original item (e.g. when the non-word “blatow” is repeated as “alow”)
  - The child might replace one phoneme with others (e.g. when a child repeats “barazon” with “barazon”)

Child might make compounds of these error types

- There are a minority of repetition attempts which are difficult to score. You may find that you are uncertain whether the child’s ambiguous sound corresponds to the correct phoneme in the test item or another similar sounding one.
- The rule of thumb is to judge whether a listener, who had not heard the item on the recording, would be able to correctly reproduce the original non-word from the child’s repetition attempt. If so, score the attempt as correct. If not, score as an error. Always check the auditory recording after the appointment if you are unsure.
- Allowance should be made to children who make highly systematic speech errors in their spontaneous speech. For example, you may see a child who you notice consistently replaces the sound “k” in a word with the sound “t”. If this child produced “contramponist” in response to “contramponist”, the repetition attempt should be scored correct.
- Some common speech errors you might come across in six year olds, for example, are:
“$w$” for “$w$”
“$f$” for “$f$”
“$n$” for “$n$”

- If the child does not repeat the item in the silent interval press the pause button and ask
  “Can you say it?” If the child asks to hear the item again, the tester should say it aloud and
  the child attempt to repeat it. Irrespective of whether the repetition attempt is successful
  or not, an error should be scored in this situation.
- You need to make sure that you cover your mouth from view when speaking the nonwords if
  you need to say the words aloud rather than relying on the audio. This is to avoid the child
  using lip movements to supplement their memory for the spoken nonword.
- Encourage the child to sit quietly and listen carefully.
- Encourage the child to talk a bit louder if their responses are too soft.
- There is no discontinuing rule. All items are to be administered even if children do not
  respond. Try (as best you can) to administer the complete test to all children.

Administration

Script which will play on recording once you being to explain the task to the child

“... you will hear a funny made-up word. I would like you to say the funny word back to me
as soon as you have heard it.”

“So, if the funny word was ‘nooo’, you should say ‘nooo’ back to me. Let’s try that now, shall
we?”

Trial 1

“nooo”

Child attempts repetition. If repetition attempt is incorrect:

“That was a good try, but it wasn’t quite right. Let’s try again.”

Repeat procedure until a satisfactory repetition attempt is made by child. Don’t continue test if child
cannot successfully repeat:

“Very good. Now let’s switch the recording on and hear some more funny words.”

Make sure child is paying attention by saying

“Are you ready? Listen carefully.”
10.10.9 Goldman Fristoe Test of Articulation  

The GFTA is a test of articulation. It assesses the level to which children can correctly pronounce the sounds of English.

- **Materials**: The GFTA stimulus booklet
- **Administration**: Point at the stimulus and ask "what is this" as per instructions – participants often start naming before this is asked, however. Note that some stimuli have different probe questions.
- **Record this subtest**.
- **Start with Item #1 and use scoring sheet for Sounds in Words**.
- **Score for error**.
- **Write down the sounds the child used if you can**.

Once completed and scored (and recorded in database), pass on recording to Ange Morgan (via Georgie) to do the deep scoring/management.

10.10 Export of Recalling Sentences and NPVT Data

With the expertise of CURVE Tomorrow, we can now export the RS and NPVT data directly from each of the two VicCHILD iPads into RedCAP.

**Protocol**

- Each Wednesday, ST will export data from both iPads.
- In the rare occurrence where the NPVT and the RS subtest of the CELF are not administered on the same day, then the RA needs to email Sheri requesting a delay in export of data from the specific iPad.
- Select the ALT app on each iPad.
- Select Results.
- Select Upload to RedCAP.
- The files should automatically start uploading. Don’t be concerned if you see a red circle around an ID number with the note “Upload Failed”, this just means that the file has already been exported. A green circle around an ID number indicates a successful upload.
Notes: on future use of the Data/RedCAP Integration

- The current set up does not account for how we record multiple administrations per participant (e.g, we see them at 5yrs and 3yrs old). In the future if the NPVT and RS subtests are repeated then we will need to adjust the VicCHILD ID (record ID in project) so that it delineates what data they're at. e.g, V0260 at 5-7yr direct assessment becomes "V0260_d57" V0260 at 9-11yr direct assessment becomes "V0260_d911"

- At present (April 2015), the need to use an ID number with a further code at the end to indicate time-point is not required and future follow-up has not been confirmed (ST).

11 Scoring Rules for Direct Assessments

11.1 Parent Questionnaires

11.1.1 Mill Hill
Calculate the total number of correct responses.

There is a list of correct answers to Forms A and B in the VicCHILD Direct Assessment Manual to help you score the items.

11.1.2 AQOL
This measure will be scored using STATA syntax according to weights and scores.

11.1.3 CHILD
Average all the responses for a representative mean of listening function

11.1.4 PEACH
- Quiet subscale score: \[
\text{score} = \frac{\text{Total correct items}}{24} 
\]
- Noise subscale score: \[
\text{score} = \frac{\text{Total correct items}}{20} 
\]
- Overall PEACH score: Add all scores/24

<table>
<thead>
<tr>
<th>Scoring: To be completed by professional</th>
</tr>
</thead>
<tbody>
<tr>
<td>RAW Score</td>
</tr>
<tr>
<td>QUIET</td>
</tr>
<tr>
<td>NOISE</td>
</tr>
<tr>
<td>OVERALL</td>
</tr>
</tbody>
</table>

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11.1.5 PedsQL Measures

All PedsQL measures will be scored using STATA syntax.

11.2 Child Direct Assessments

11.2.1 CELF-4

- For queries about scoring individual items see the separate scoring document with item-specific questions and the scoring tables.
- For each of the 4 or 5 subtests calculate the Raw Score (the sum of the item scores). Transfer the raw scores for each subtest to the inside cover (pg.2) of the record form.
- To convert the subtest raw scores to norm referenced scaled scores, use the age appropriate table (Table B; see separate scoring tables document). Raw scores for each subtest are listed below their respective headings and associated scaled scores are listed in the outer columns on the left and right sides.
- Enter the subtest scaled scores on the front page and the inside cover of the record form.
  
  Add the subtest scaled scores below each composite score listed. For example, to compute the Core Language Score, for 5-7yo add the Concepts and Following Directions, Word Structure, Recalling Sentences and Formulated Sentences subtest scales scores; for 9-11yo, add the Concepts and Following Directions, Recalling Sentences, Formulated Sentences, and Word Classes 2 Total subtest scores. Record the total in the box labelled Sum of Subtest Scaled Scores in the Core Language score column.

- Refer to Table C for relevant age group (see separate document) for converting the sum of subtest scaled scores into Core Language and Index Scores.
- Percentile ranks for the Core Indices are in Table C and percentile ranks for individual subtests are listed in Table E (see separate document).

Recording the Raw Score

- The total raw score for a subtest is the sum of the item scores.
- Record the total raw score for each subtest administered in the appropriate box on Page 2 of the record form.

Recording the Scaled Score
• Using the age-appropriate tables in the scoring document convert each subtest raw score to a norm-referenced scaled score.

• Subtest norms in the scoring document have 3 parts. Part A contains the subtest scaled scores. (Except for the Word Classes-Total scaled score [part B].) Part C lists the plus/minus (+/-) score points that are used to build confidence intervals.

• Raw scores for each subtest are listed below their respective headings and the associated scaled scores are listed in the outer columns on the left and right sides.

• To use the table, locate the examinee’s total raw score in the appropriate subtest column, then read across to the left or right to the number in the scaled score column. (This is the scaled score equivalent of the total raw score for the subtest.)

• Enter each subtest scaled score on Page 2 of the record form.

Confidence Intervals

• You can use section C of Table C to obtain a confidence interval for each subtest scaled score.

• Select the 95% level of confidence then record the number of scaled score points (+/- column on page 2 of record form).

• Compute the upper limit by adding points to the subtest scaled score.

• Compute the lower limit by subtracting points from the subtest scaled score.

• Record both numbers in the CI interval box on page 2 of the record form.

• The lower and upper limits have to span the obtainable score range for a given scale e.g. 1 to 19.

Subtest Raw Scores of Zero

• If an examinee obtains a raw score of 0 on a CELF-4 subtest that score does not indicate that they entirely lack the ability measured by the subtests. It indicates, rather, that their ability cannot be determined by the particular set of subtest items.

• If an examinee obtains a raw score of 0 on only one of the subtests that form a composite score, you can still derive the composite score by using the appropriate norm tables.

• If two of the subtests that form a composite score have total raw scores of 0, you cannot derive the composite score.

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• In order to convert a total subtest raw score of 0 to the subtests scaled score, the raw score must be an earned score of 0 (i.e. meaning items that were administered scored 0 until the discontinue rule was met).

**Percentile Ranks**

• Percentile indicates the percentage of people who have scored equal to or lower than a specific score
• Convert each subtest scaled score and each composite standard score to a percentile using the Table in scoring document
• Locate subtest scaled score in subtest column and read across to the right to Percentile Rank column
• Record percentile rank for each score in appropriate box on page 2
• Repeat for Composites

**Norm-referenced Standard Scores**

• The Core Language and Index Scores are norm-referenced standard scores which enable you to compare the examinee’s score to that of others the same age
• They are on a normalised standard score scale with a mean of 100 and SD of 15
• The table below shows the relationship of standard scores and percentile ranks to distances from the mean in SD units

<table>
<thead>
<tr>
<th>Standard Score</th>
<th>Distance from Mean</th>
<th>Percentile Rank</th>
</tr>
</thead>
<tbody>
<tr>
<td>145</td>
<td>+3SD</td>
<td>99.9</td>
</tr>
<tr>
<td>130</td>
<td>+2SD</td>
<td>98</td>
</tr>
<tr>
<td>115</td>
<td>+1SD</td>
<td>84</td>
</tr>
<tr>
<td>100</td>
<td>Mean</td>
<td>50</td>
</tr>
<tr>
<td>85</td>
<td>-1SD</td>
<td>16</td>
</tr>
<tr>
<td>70</td>
<td>-2SD</td>
<td>2</td>
</tr>
</tbody>
</table>
Concepts and Following Directions

- Give 1 point per correct item. Add scores for each item administered and 1 point for each item not administered before the child’s start point (if basal was reached) for raw score.

Recalling Sentences

Transfer audio files to computer

- Ensure iTunes is installed on the computer
- Connect the iPad to the computer using the USB cable
- iTunes will automatically launch (if not, open iTunes)
- In the top left corner of the iTunes screen, click on the iPad icon to view contents of iPad.

From the menu at the left of the screen, click on Apps:
- Scroll to the bottom of the screen. Under File Sharing, your Apps will be listed. Select the ALT app. All of the audio files stored in the app will be displayed to the right of the screen:

<table>
<thead>
<tr>
<th>File Sharing</th>
<th>ALT Documents</th>
</tr>
</thead>
</table>
| The apps listed below can transfer documents between your iPad and this computer. | ALT tagged
|                                          | ALT tagged-ahm
|                                          | ALT tagged-ahm-1
|                                          | Participant 100 - Response 5.m4a
|                                          | Participant 100 - Response 6.m4a
|                                          | Participant 100 - Response 7.m4a
|                                          | Participant 100 - Response 8.m4a
|                                          | Participant 100 - Response 9.m4a
|                                          | Participant 100 - Response 10.m4a
|                                          | Participant 100 - Response 11.m4a
|                                          | Participant 100 - Response 12.m4a
|                                          | Participant 100 - Response 13.m4a
|                                          | Participant 100 - Response 14.m4a
|                                          | Participant 100 - Response 15.m4a
|                                          | Participant 100 - Response 16.m4a
|                                          | Participant 100 - Response 17.m4a
|                                          | Participant 100 - Response 18.m4a
|                                          | Participant 100 - Response 19.m4a
|                                          | Participant 100 - Response 20.m4a

- Highlight all of the audio files you wish to transfer and click the ‘Save to’ button to locate your chosen folder (alternatively, you can drag the files to your chosen folder)

- As each participant will have up to 32 audio files (one audio file per sentence), you may wish to create a folder for each participant in which to store their audio files.

2. Scoring
- Press the back arrow ← (top left) to go to the ALT app main screen.
- Press on the Results icon to enter the Results screen.
- Find and select the participant ID from the left side of the screen.
- The participants audio recordings and markings will be displayed to the right of the screen.
- Check for sentences marked as completely correct or completely incorrect (i.e. green tick or red cross).
- For those sentences marked with a green tick circle 3 on the paper record form.
- For those sentences marked with a red cross circle 0 on the paper form.
- For those sentences marked with a question mark, you will need to listen to these sentences to determine the score.
- Note: for RAs new to the test, it is recommended that you listen back to all of the sentences to double-check your scoring.
- You may listen to the sentences from the Results screen of the ALT app (tap each sentence to listen) or from the transferred audio files on the computer.
- As you listen, use the record form the mark the participant’s errors (you can use the editing symbols suggested at the top of the record form).
- Score the item by comparing the participant’s response to the stimulus sentence. Count the number of errors in the response and classify it according to the following rules:
  - 0 0K No errors made. Circle 3
  - 1 Error One error made. Circle 2
  - 2-3 Errors Two or three errors made. Circle 1
  - 4+ Errors Four or more errors made. Circle 0
- Use extra scoring information from page 35 of manual.
- Decide on number of errors per item using table below.
- This is not automated, and so needs to be calculated as you go.

**Guidance from Melissa Wake in regards to scoring RS items for the VicCHILD Study**

Final Scoring Decision for Recalling Sentences by NW (guided by manual & other projects). NW understands that this may systematically lower some scores but she doubts by much (see highlighted print out of reasoning from authors AND Pearson).

1. If the error is in morphology (i.e., grammatical errors - for example instead of “followed” the child says “follow”), even if the reason could be phonological, this is scored as an error.
2. If the error is not morphological (e.g., missing a terminal consonant of a noun i.e., “rabbit” instead of “rabbit”) then you don’t score it as an error. You do not count misarticulations as errors because these do not tap morphology or memory.

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These rules apply in addition to the standard rules of omissions, repetitions, additions, transpositions and substitutions being scored as errors.

Additional pointers for scoring RS from other projects (i.e., ELVS and LAL)

<table>
<thead>
<tr>
<th>Number of errors</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>1. Look at the whole sentence first.</strong></td>
</tr>
</tbody>
</table>
| **2. Each transposition that changes the meaning of the sentence**  
  e.g. The tractor was followed by the bus/The bus was followed by the tractor | 2 errors |
| **3. Each transposition that does NOT change the meaning of the sentence**  
  e.g. The computers and printers were donated by the school board/The printers and computers were donated by the school board | 1 error |
| **4. A substituted word in a contraction**  
  e.g. Didn’t the boys eat the apples/Won’t the boys eat the apples  
  *(Won’t for didn’t (will not for did not))* | 1 error |
| **5. One substitution + one addition**  
  e.g. Did the girl catch the baseball/Couldn’t the girl catch the baseball  
  *(Couldn’t for did equals two errors (could not for did))* | 2 errors |
| **6. Substituting an appropriate contracted form**  
  e.g. The rabbit was not put in the cage by the girl/The rabbit wasn’t put in the cage by the bird | 0 errors |
| **7. Substituting a non-contracted form for a contracted form**  
  e.g. Didn’t the boys eat the apples/Did not the boys eat the apples? | 0 errors |
| **8. Regional and cultural patterns or** | 0 errors if part of the examiner’s language |
 Variations that reflect dialectical differences. Refer to Appendix B of manual.

E.g., Substituting that for who in sentences 17 and 20.

<table>
<thead>
<tr>
<th>Number</th>
<th>Rule</th>
</tr>
</thead>
<tbody>
<tr>
<td>3</td>
<td>If they repeat the sentence with 0 errors</td>
</tr>
<tr>
<td>2</td>
<td>If they make 1 error (i.e. single word changed, added, substituted or omitted)</td>
</tr>
<tr>
<td>1</td>
<td>If they make 2-3 errors (i.e. 2-3 words changed, added substituted or omitted)</td>
</tr>
<tr>
<td>0</td>
<td>If they make 4+ errors (i.e. 4+ words changed, added, substituted, omitted or reversed; resequencing of phrases containing four or more words)</td>
</tr>
</tbody>
</table>

Score the subtest:
1. Add scores that examinees obtained on each item administered.
2. Add in 3 points per each non-administered item (i.e. those preceding the start point if basal was retained).
3. Add the scores in each column. Enter this in column subtotal boxes.
4. Add column subtotals to get subtest raw score.

Examples of Scoring

<table>
<thead>
<tr>
<th>Item</th>
<th>Errors</th>
<th>Stimulus Sentence</th>
<th>Examinee Response</th>
<th>Scoring</th>
</tr>
</thead>
<tbody>
<tr>
<td>11</td>
<td>1</td>
<td>The sand castle was built by the girls and boys.</td>
<td>The sand castle was made by the girls and</td>
<td>One word substitution</td>
</tr>
<tr>
<td></td>
<td></td>
<td>boys</td>
<td></td>
<td></td>
</tr>
<tr>
<td>---</td>
<td>---</td>
<td>------</td>
<td>---</td>
<td></td>
</tr>
<tr>
<td>20</td>
<td>2</td>
<td>The student who won the award at the art show was very excited.</td>
<td>The student who won the prize at the art show was very surprised.</td>
<td>• 2 substitutions  • Score=1</td>
</tr>
<tr>
<td>18</td>
<td>4</td>
<td>If the rain doesn’t stop before noon, the excursion will have to be cancelled.</td>
<td>If the rain doesn’t stop by noon, the trip will be cancelled.</td>
<td>• 2 substitutions  • 2 omissions  • Score=0</td>
</tr>
<tr>
<td>27</td>
<td>5</td>
<td>The librarian has twelve new year-eight science books reserved for us.</td>
<td>The science teacher has eight new year-12 books for us.</td>
<td>• 1 substitution  • 1 addition  • 1 transposition (diff meaning)  • 2 omissions</td>
</tr>
</tbody>
</table>

**Word Structure**

- Give 1 point per correct item. If child’s response is different from target but demonstrates the target structure and is meaningful in the context, mark as correct.
- Add scores for each item to calculate a raw score.

**Word Classes 1 — Receptive**

- Sum the number of correct responses for the raw score. Refer to Table for Scaled score.

**Word Classes 1 — Receptive & Expressive**

Three scores obtained from this subtest:

1. Word Classes-2-the receptive score (WC-R)
2. Expressive Score (WC-E)
3. Total Score (WC-T)

- Part 1 (Receptive Score) Circle the words that the examinee gives in response to the first part of each item (correct words are printed in colour on record form).
Part 2 (Expressive Score) Correct responses to the 2nd part of each item are listed below the line on record form. If the examinee’s response is similar to that listed, circle the listed response and score 1 point. If the response is not listed, write it verbatim for later scoring consideration.

Score 0 if incorrect or no response.

**Deriving the Word Classes 2-Receptive scaled score**

- Sum the receptive scores (first part) that the examinee obtained on each item administered
- Add in 1 point per each non-administered item (i.e. those preceding the start point if basal was reached)
- Using the scoring tables, convert this raw score into the Word Classes 2-Receptive scaled score

**Deriving the Word Classes 2-Expressive scaled score**

- Sum the expressive scores (2nd part) that the examinee obtained on each item administered
- Add in 1 point per each non-administered item (i.e. those preceding the start point if basal was reached)
- Using the scoring tables, convert this raw score into the Word Classes 2-Expressive scaled score

**Deriving the Word-Classes 2-Total scaled score**

- Add the Word Classes 2-Receptive scaled score and Word Classes 2-Expressive scaled score
- Using section B of age-appropriate tables in the scoring tables, convert this sum to a total scaled score

**Word Classes Scaled Score**

- The shaded columns are used to covert Word Classes scores
- Follow the steps below to convert the Word Classes raw scores to scaled scores and then to convert those scaled scores to the WC-Total Scaled Score
Enter the Word Classes-Receptive and Expressive raw scores in the Raw Score column of the subtest scores summary on page 2 of the record form.

Looking at the shaded column in section a of the Table C convert the raw scores to scaled scores.

Enter the scaled scores in the Scaled Score column on page 2 of the record form.

Add the Receptive and Expressive scaled scores and record the sum in the shaded box.

Refer to section b of Table C and find the sum of the Receptive and Expressive scaled scores in the shaded column to find the Total Scaled score that corresponds to the sum. Record the WCT Scaled Score box on page 2 of the record form.

Sentence Structure

- Give 1 point per correct item and 0 for incorrect or no response.
- Add scores for each item to calculate a raw score.

Formulated Sentences

- Use the credit and no-credit rules below to decide if a sentence should receive credit at all.

### No-credit Rules (Score=0)

- The stimulus word(s) must be used.
  (To assist with scoring underline the stimulus word(s) in each sentence.)

- Tense and plurality of stimulus word must not be changed, e.g. give instead of gave.

- Exceptions:
  1. Dialectical or colloquial substitutions for the stimulus word e.g. till for until; 'cause for because
  2. Stimulus word may be used in a possessive form (children's for children)

- Semantic meaning of stimulus word must be used unchanged, e.g. 'Gave is eating dinner' is 0 points.

- Sentence must be about something in the picture (responses that are even remotely related to the stimulus picture should be accepted).

### Credit Rules

- Sentence response may be part of a dialogue between or among people pictured.
• An animal may be considered the speaker.

• The sentence may be stated in the first person (i.e. student puts herself or himself in the stimulus picture as one of the speakers).

• Credit regional and cultural patterns or variations that reflect dialectal differences if they are appropriate for the examinee’s language background. Score these variations as intact sentences according to the structural rules of the student’s dialect. Refer to Appendix B.

• Responses may be in the form of questions or statements.

• If 2 or more sentences are given in response to an item, score each sentence as if it was the student’s only response.

• Give credit for the one sentence earning the highest score, but do NOT add the scores for the sentences.

• Some examinees respond with the target word as the first word of their sentence response, somewhat like a starter. Give credit for such sentences if all the rules for scoring are met.

Score each response

• Use scoring guidelines in Table 2.4 of Manual (page 43 onwards)

• Use examples of scored responses in scoring document

• Read sentence and underline the stimulus word(s).

• Assign a score according to the following Scoring Key (decide if the sentence has correct structure and count the number of syntactic and/or semantic errors)

1. Add scores that examinee obtained on each item administered
2. Add in 2 points per each non-administered item (i.e. those preceding the start point if basal was reached)
3. Add the scores in each column. Enter this in column subtotal boxes.
4. Add column subtotals to get subtotal raw score.

<table>
<thead>
<tr>
<th>Score</th>
<th>Rule</th>
<th>Example</th>
</tr>
</thead>
</table>
| 2     | • Complete sentence.  
        • Semantically and grammatically correct.  
        • Uses correct structure.  
        (i.e. logical, meaningful, complete and | Item 16: The police officer was directing  
the cars because the traffic lights were out. |
<table>
<thead>
<tr>
<th>Type</th>
<th>Description</th>
<th>Example</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Complete sentence</td>
<td>Item 21: We have to pay, otherwise we will have to do the dishes.</td>
</tr>
<tr>
<td>Correct structure</td>
<td>Item 25: As soon as Julie had got her lunch, Carlos was finished. As soon as he ate his sandwich, lunch will be over.</td>
<td></td>
</tr>
<tr>
<td>Only 1 or 2 deviations in syntax or semantics</td>
<td></td>
<td></td>
</tr>
<tr>
<td>0</td>
<td>Type (a)</td>
<td>Item 21: Until I'll be done.</td>
</tr>
<tr>
<td>Incomplete sentence</td>
<td>Item 19: The boy could have rode his bike if he broke his arm although he could skateboard.</td>
<td></td>
</tr>
<tr>
<td>Does not demonstrate correct structure</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Type (b)</td>
<td>Complete sentence</td>
<td>Item 26: Mark slept late in order to catch the bus.</td>
</tr>
<tr>
<td>Correct structure</td>
<td></td>
<td></td>
</tr>
<tr>
<td>More than 2 deviations in syntax or semantics</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Type (c)</td>
<td>Complete sentence</td>
<td>Item 14: He is buying cabbage or lettuce.</td>
</tr>
<tr>
<td>Not logical or meaningful</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Type (d)</td>
<td>Required stimulus word is NOT used</td>
<td>Item 16: Because I go to school, I learn.</td>
</tr>
<tr>
<td>Type (e)</td>
<td>Response is totally unrelated to the picture</td>
<td></td>
</tr>
</tbody>
</table>

**Core Language Score**

The core language score is a measure of general language ability that quantifies the examinee's overall language performance. It sums the scores from the following subtests: [5-7] Concepts and
Following Directions, Recalling sentences (RS), Formulated sentences (FS), Word Structure (WS); [9-12] Recalling sentences (RS), Formulated sentences (FS), Word Structure (WS), Total (WC2)

- Transfer the subtest scaled scores from page 2 to the appropriate spaces on page 1 (making sure you record them in the correct age-appropriate column).
- To assist, the subtests required for the Core Language score are in bold type in the Subtests Score Summary table on page 2 of the record form.
- Sum the subtest scaled scores below each composite score listed.
- Record the sum in the box labeled Sum of Subtest Scaled Scores and covert the sum to the Core Language Score.
- Record the Core Language score in the Standard Score box in the Core Lang score column.
- Repeat this for the following: RLI, ELI (see below for subtests in each composite).
- Establish CIs for the Core Lang Score and RLI and ELI in the same way that you did for the subtest scaled scores (remembering once again that CIs span the obtainable score range of a score scale e.g. for Core Lang 40-160).

**Index Scores**

To obtain an index score sum the scaled scores of subtests that comprise each index:

- The RLI is a measure of listening and auditory comprehension: 5-7 is CFD + WC2/2 + SS; 9-12 is CFD + WC2R.
- The ELI is an overall measure of expressive language skills: 5-7 is WS + RS + FS; 9-12 is RS + FS + WC2-E.

### 11.2.2 WNV

- Standard Scores on the WNV are provided for subtests and for the full scale score and are as follows:
  - T scores: mean=50, SD=10
  - Full Scale Standard Scores: mean=100, SD=15

- If an examinee responds to a previously administered item after beginning the next one, award the appropriate credit for the response or self-correct, if they should be awarded appropriate credit. If it was intended to replace a previous response, score only the intended response, regardless of accuracy.
• Calculate the child’s age on the summary page (front page of record form) using the first date if tested during two sessions. Assume months have 30 days and do not round up.
• Calculate total raw score for a subtest by summing the item scores: add scores including reversal items and non-administered items (items where credit was given) prior to the start point.
• Write the total raw score in the box at bottom of each subtest.
• Transfer these scores to the raw score column of the ‘total raw score to T score conversion’ table.
• To derive T scores for Matrices and Recognition subtest raw scores use table A1 for 5-7yo.
• To derive T scores for Matrices and Spatial Span subtest raw scores use table A1 for 9-12yo.
• If a child obtains a raw score of zero on either of the subtests comprising the 2-subtest battery the full scale score should not be derived.
• To calculate the full scale score sum all subtest T scores and transfer this sum into ‘sum of T scores box’ in the ‘Sum of T scores to full scale score conversion’ table. Use table A3 (p. 141-142) to derive the Full Scale score equivalents and corresponding percentiles.
• Determining test age equivalents: use table A5. For each age group the total raw score corresponding to a T score of 50 represents the median test performance for that particular age. The table shows raw scores for indicated test ages.
• Subtest level analysis: Table B3 provides required diffs b/w subtest T scores to attain statistical significance (critical values) at .15 (6 points for 4-7 year olds) and .05 levels (11 points for 4-7 year olds).
  If the absolute value of the difference scores equals or exceeds the critical value, circle the Y in the box labelled significant difference. If the absolute value does not equal or exceed the critical value circle N in the subtest comparison table.
• If the difference between subtest T scores is significant, use table B4 (p.156 onwards) to determine the cumulative percentage of the normative sample that obtained a same or greater diff between the subtest T scores. These base rates are separated into minus and plus columns based on the direction of the difference.

11.2.3 NPVT
• Item Response Theory (IRT) is used to score the NPVT. A score known as a theta score is calculated for each participant; it represents the relative overall ability or performance of the participant. A theta score is very similar to a z-score, which is a statistic with a mean of zero and a standard deviation of one.
• Age-Adjusted, Fully Adjusted and Unadjusted Scale Scores, as well as a national percentile
rank that corresponds to the age-adjusted scale score, are provided for the NPVT.

- In addition, the theta score is converted to a “Computed Score” that appears in the Assessment Scores output file available after each test.
- Download this to VicCHILD scoring folder to get the theta and t-score, and match results to appropriate age norm in the NIH Toolbox Manual (see p.19.) The Computed Score for NPVT ranges from roughly 200 to 2000 and can be used for simple vocabulary ability comparisons over time.

**Interpretation**

The NPVT is a measure of general vocabulary knowledge and is considered to be a strong measure of crystallized abilities (those abilities that are more dependent upon past learning experiences and are consistent across the life span). To interpret individual performance, one can evaluate all three types of scale scores:

- A participant’s age-adjusted scale score at or near 100 indicates vocabulary ability that is average for the age level. Scores around 115 suggest above-average vocabulary ability, while scores around 150 suggest superior ability — in the top 2 percent nationally for age, based on Toolbox normative data.
- Conversely, a score of 85 suggests below-average vocabulary ability, while a score in the range of 70 or below suggests significant impairment in language ability, which may also be indicative of difficulties in school (for children) or trouble functioning in work environments with a language demand.

An unadjusted scale score allows us to view the participant’s performance in comparison to the entire Toolbox national sample, allowing for a more absolute view of the participant’s ability. The fully adjusted scale scores have been statistically adjusted to level the playing field interpretively, such that an individual’s score can be compared to a narrower group, more similar demographically. The NPVT Computed score provides a way of gauging raw improvement or decline from Time 1 to Time 2 (or subsequent assessments). Such a score is useful because a raw score does not provide relevant information on a computer-adaptive test. (Raw scores are useful for monitoring absolute improvement/decline over time when statistical transformations are not used in the scoring process, as is the case in IRT-based scoring or in the Flanker or DCCS measures, described below.) Thus, a computed score of 660 at Time 1 and 640 at Time 2 represents real improvement by the participant in vocabulary knowledge; however, this individual’s Age-Adjusted Scale Score may or may not have increased, depending on how his/her performance at Time 1 and Time 2 compared to the age cohorts used in the national norms.
11.2.4 ERNRI
Accuracy of scoring is dependent upon quality of transcription

- Transcribe each narrative, starting each utterance on a new line (this is essential for MLU calculation)
- The scoring will be done by an RA dedicated to this and manual gives guidelines.
- the MLU and Type/token ratios will be computed using text analysis software

Converting ERNRI raw scores to percentiles and standard scores

- The scoring document contains tables for converting raw scores for story ideas, comprehension and MLU to percentiles
- The scoring document (Table 1) is used to convert percentiles into standard scores (required if you want to compute the Forgetting score which is the difference between the standard scores for ideas on initial story-telling and recall). Percentile equivalent of this difference score is in Table 2 of the scoring document.

11.2.5 LSN-S

- For each subtest, there is a minimum delivery of 22 sentences, maximum of 30 sentences
- Scoring all occurs online, with the level of the next sentence dictated by the score achieved on the previous.
- Scoring is a count of the number of words correct per sentence, with the following guidelines:
  - A word is considered correct if it is repeated exactly as it appears on the playback screen
  - If an extra word is included, there is no penalty
  - If a substitution, omission or change in tense/plurality etc occurs, the number of correct words is adjusted accordingly (i.e., the word form must be identical)
  - Word order does not matter, score is not influenced by word placement.
- The Speech Reception Threshold (SRT) which is progressively calculated as the test proceeds is the measure of interest. Once the subtest is complete, the SRT is stored. If the subtest is aborted or incomplete, then the number of sentences delivered and the progressive SRT needs to be recorded.
The results are automated from the tester’s online scoring; look at the results tab and note down the scores shown for each subtest, and for the combined two tests into the data collection form in the spaces provided.

Save the results screen as a screenshot? iPad photo? so that we have back up of the results.

11.2.6 WRAT

Word Reading

- Part 1: Letter Reading & Part 2: Word Reading, which consists of 55 words.
- Pay careful attention to the pronunciation guide (provided in the Test form and by the Australian English way files in the VICCHILD scoring folder) during subtest administration in order to score the participant’s responses correctly.
- One point is given for each letter and/or word read correctly.
- You may use any notation system that you prefer to record Participant responses. The recommended procedure is to circle the letter items or the item number of the word items if the response is correct. Cross out the letter or the item number of the word if the response is incorrect.
- If the participant changes his/her response, score the last attempt.
- Add up the number of words the child read correctly up to the ceiling and write in the box on the scoring form.

- The total for the Word Reading subtest is the Letter Reading raw score (out of 15) + Word Reading raw score (out of 55). Thus the Word Reading Total Raw Score is out of 70.
- Transfer the Word Reading Total Raw Score to the Assessment Summary Sheet.

Spelling

- Add up the number of words the child spelt correctly up to the ceiling and write in the box on the scoring form, on the front of the form and on the summary sheet.
- On Part 1: Letter Writing, give 1 point for any correctly written letter from the child’s own name, with a maximum of 2 points, and 1 point for each correctly written letter from the list.
worth 1 point. Record a number “1” or “2” next to sentences which contain the words/phrases listed. If a synonym of a word listed or word in a local dialect is used, please record for later scoring discussions at the next RA meeting.

- Do NOT give points for information that is given completely out of order
  - If the referent (bus, policeman, etc.) is not specified at each change of “actor”, the information score for the activity referred to should have one point deleted. For example:
    - “The bus ran away
      - Went in a tunnel”
  - Instead of giving 2 points for “in tunnel”, the child only gets 1, because at first the child was referring to the bus, and did not state that now he/she is talking about the train when he/she said “went in a tunnel”
- Tally up all the points and write the total in the “total” box on the bottom left hand side.
- Transfer the Information, Subordinate Clause, and Sentence Length scores on to the front side of the page.
- Using the pages below, find the age ranges for the child’s Information, Subordinate Clause and Sentence Length Scores, and record these ranges on the front side of the scoring sheet, and on the summary sheet.
  - For example, if a child scored “9” for Sentence Length, look at which ages “9” is an appropriate score for. You will see that it is from 5:1 through to 7:11, so write 5:1-7:11 in the “age range for score” box on the front page so Sentence Length.

11.2.8 CTOPP-2

- The total raw score for each of the three subtests is the total number of correct test items up to the ceiling item.
- The raw composite score for Phoneme Awareness is the sum of the: (5-6yo) Elision, Blending, and Sound Matching; and (7yo) Elision, Blending, and Phoneme Isolation
- Use the tables in separate scoring document to covert the child’s raw score into a standard score, percentile rank, Age equivalent & Grade Equivalent score & enter on the record form & summary sheet.
  - Table A1 pp.100-101 for age equivalencies from raw scores

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11.2.9 CNRep

- The total raw score is the number of items pronounced correctly
- Use Table 1 below to determine the Standard Score
- Add up the totals in the columns to get Raw Scores by syllable
- Enter the total raw score and standard score on the front page of the record form.
- If you were unsure of any responses, listen back to the audio recording to determine the score
- To calculate the raw score, count the number of items correct
- To calculate the raw score by syllables:
  - Transfer the item scores to the Syllable Template page
  - Count the number of items correct in each column

Transfer audio files

- Connect your recording device to the computer to save the audio files in your chosen folder (process will vary depending on recording device)
- If a child has a raw score less than the minimal for a standard score, their standard score needs to be marked as less than the minimal. So for a raw score of 1-4, mark this on the record form and the summary sheet as less than the lowest possible standard score (i.e. SS = <64). A raw score of 0 or 0 cannot be given a standard score so use a dash (-).
- Use Table 2 below to work out the child’s % range (range of centiles). Using the column appropriate for the child’s age find the raw score and look across to the left hand column to find the centile point. If the exact raw score is not found, calculate the range of centiles in which the raw score falls (e.g. a raw score of 26 for a 6yo child would give a centile range of 50-75)
• Note that there are no norms for 9;0-11;11, so for the 8-12 testing group we have to use the 8;11 scores as basals for now. We may in the future compare these to results from the comparably-aged ELVS groups.

11.2.10 Goldman-Fristoe Test of Articulation 2

- Make sure that all errors have been noted in the appropriate cells of the recording grid
- Count the total number of articulation errors, including on consonant clusters (blends). The total raw score is the number of items pronounced incorrectly, i.e., the total score is the number of errors (/77). Write the raw score in the total box on the front right cover of the response forms.
- Use Table B1 in the scoring document to determine the Standard Score, Confidence Interval, & Percentile
- Make sure the above are gender and age appropriate
- For age equivalence locate the raw score and read across to determine the age (see Table B1 in scoring document), and transfer the standard, percentile and 95% confidence interval to the appropriate box on the score summary box of the response form.
- Once you’ve entered these scores into the database, hand over the recording and response form to Angela Morgan for type of error scoring.

NB: if you have extreme scores (raw score of 0, or standard score less than 40), refer to manual in scoring document (p.30 ff. in GFTA-2 manual)

12 Office Procedures after Direct Assessments (to be updated still)

12.1 Database updates

- When you return to the MCRI and have completed the assessment scoring, update the child’s visit information in REDCap.
- Any Developmental issues to be noted. We will usually be aware of issues prior to this visit, but please update any information that you learn about a child from the visit that has not been previously noted.
- Any issue(s) raised: information that comes up from the visit that might be important for future contact with the family.
Appendix D: Audiogram request cover letter

[DATE]

Dear [Australian Hearing CENTRE NAME],

Child Name: 
Child Date of Birth: 

The above named child and their family are participants in VicCHILD, the Victorian Childhood Hearing Impairment Longitudinal Databank. Periodically, VicCHILD assesses the language and hearing of participating children. It needs some audiologic information prior to assessment.

What is this request for?
This child’s most recent audiometric thresholds. Please provide as much information as was obtained at the time.

Am I allowed to give you this information?
Yes, you are. Attached is signed consent from the child’s parent or guardian. Australian Hearing has reviewed and approved this consent form, as has the Royal Children’s Hospital Ethics in Human Research Committee.

How do I get this information to you?
There are several ways you can provide this information:
1. Verbally, over the phone (VicCHILD will call you)
2. By completing, scanning and emailing this letter to vic-child@rch.org.au
3. By completing this letter and faxing to (03) 9345 5900

Alternatively, you can provide VicCHILD with a copy of the child’s audiogram.

What is the deadline?
We’d be very grateful if you could provide this information by 3 business days. VicCHILD cannot complete assessments until we have this information.

If you have any questions please contact VicCHILD on (03) 9345 4215 or vic-child@rch.org.au. On behalf of the VicCHILD team, thank you for your cooperation with this request.

[Signature]
Professor Melissa Wake
Associate Director of Research
The Royal Children’s Hospital
Centre for Community Child Health

[Signature]
Dr. Zelie Pouliakos
Clinical Psychologist
The Royal Children’s Hospital
Centre for Community Child Health

[Signature]
Ms Sherryn Tobin
Project Coordinator
The Royal Children’s Hospital
Centre for Community Child Health

Please turn over
CONFIDENTIAL: Please return to VicCHILD
Email: vic-child@rhh.org.au
Fax: 03 9345 5500

Child Name: ____________________________
Child Date of Birth: _______________________
Date of hearing assessment: ___________________

<table>
<thead>
<tr>
<th>Ear</th>
<th>250Hz</th>
<th>500Hz</th>
<th>1000Hz</th>
<th>2000Hz</th>
<th>3000Hz</th>
<th>4000Hz</th>
<th>8000Hz</th>
<th>3FAHL</th>
<th>4FAHL</th>
</tr>
</thead>
<tbody>
<tr>
<td>AC</td>
<td>Right</td>
<td></td>
<td></td>
<td></td>
<td></td>
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<td></td>
<td></td>
</tr>
<tr>
<td></td>
<td>Left</td>
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<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>BC</td>
<td>Right</td>
<td></td>
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<tr>
<td></td>
<td>Left</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

Transducer used (please tick):

- [ ] Headphones
- [ ] Inset earphones
Appendix E: confirmation letter and parent questionnaires

Address

Date

Dear [parent],

We appreciate your continued support of the Victorian Childhood Hearing Impairment Longitudinal Databank (VicCHILD).

Thank you for making a time for us to visit at your home. We're looking forward to meeting [child name], and our language and learning assessments with [him/her].

We will be seeing you and [child name] at [location] on [date] at [time].
The visit takes approximately 30 minutes.
If you need to change the day or time, please contact us on (03) 9345 4215.

This package contains:

- 1 parent questionnaire

We'll collect it when we visit. You can either fill it out at the visit, or make a start beforehand.

We look forward to meeting you both. Any questions? Contact VicCHILD on (03) 9345 4215 or vicchild@uch.org.au. Thank you again for your time!

Professor Melissa Wake
Associate Director of Research
The Royal Children's Hospital Centre for Community Child Health

Dr. Zefire Poulaka
Clinical Psychologist
The Royal Children's Hospital Centre for Community Child Health

Ms Libby Smith
Project Coordinator
The Royal Children's Hospital Centre for Community Child Health

HREC 31031 CL-V-4 Assess V1 19 Aug 2014
### 1. About your child’s WELLBEING

We know that the physical, social, and emotional wellbeing of children can differ. Below is a list of things that might be a problem for your child. Please tell us how much of a problem each one has been for your child in the past ONE month by filling the circle for:

0 if it is never a problem  
1 if it is almost never a problem  
2 if it is sometimes a problem  
3 if it is often a problem  
4 if it is almost always a problem

There are no right or wrong answers. Some may not fully apply to your child yet – just do the best you can.

*In the past ONE month, how much of a problem has this been for your child?*

<table>
<thead>
<tr>
<th>Physical Functioning (problems with...)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1.1 Walking more than one block</td>
</tr>
<tr>
<td>1.2 Running</td>
</tr>
<tr>
<td>1.3 Participating in sports activity or exercise</td>
</tr>
<tr>
<td>1.4 Lifting something heavy</td>
</tr>
<tr>
<td>1.5 Taking a bath or shower by him or herself</td>
</tr>
<tr>
<td>1.6 Doing chores, like picking up his or her toys</td>
</tr>
<tr>
<td>1.7 Having hurts or aches</td>
</tr>
<tr>
<td>1.8 Low energy level</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Emotional Functioning (problems with...)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1.9 Feeling afraid or scared</td>
</tr>
<tr>
<td>1.10 Feeling sad or blue</td>
</tr>
<tr>
<td>1.11 Feeling angry</td>
</tr>
<tr>
<td>1.12 Trouble sleeping</td>
</tr>
<tr>
<td>1.13 Worrying about what will happen to him or her</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Social Functioning (problems with...)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1.14 Getting along with other children</td>
</tr>
<tr>
<td>1.15 Other kids not wanting to be his or her friend</td>
</tr>
<tr>
<td>1.16 Getting teased by other children</td>
</tr>
<tr>
<td>1.17 Not being able to do things that other children his or her age can do</td>
</tr>
<tr>
<td>1.18 Keeping up when playing with other children</td>
</tr>
</tbody>
</table>

---

VicCHILD 5-7 year questionnaire  
Page 2 of 15  
HREC 31081  
V5 19/08/14
1.24 In general, how would you say your child’s current health is? (Fill one circle only)

O Excellent  O Very good  O Good  O Fair  O Poor

2. About your child’s BEHAVIOUR

We know that behaviour varies between children, and would like to know about your own child.

For each item, please mark the circle for Not True, Somewhat True or Certainly True. It would help us if you answered all items as best you can, even if you are not absolutely certain. Please give your answers on the basis of your child’s behaviour over the last six months.

Remember to fill the circles like this ●

<table>
<thead>
<tr>
<th>Item</th>
<th>Not True</th>
<th>Somewhat True</th>
<th>Certainly True</th>
</tr>
</thead>
<tbody>
<tr>
<td>2.1 Considerate of other people’s feelings</td>
<td>O</td>
<td>O</td>
<td>O</td>
</tr>
<tr>
<td>2.2 Restless, overactive, cannot stay still for long</td>
<td>O</td>
<td>O</td>
<td>O</td>
</tr>
<tr>
<td>2.3 Often complains of headaches, stomach-aches or sickness</td>
<td>O</td>
<td>O</td>
<td>O</td>
</tr>
<tr>
<td>2.4 Shares readily with other children, for example toys, treats, pencils</td>
<td>O</td>
<td>O</td>
<td>O</td>
</tr>
<tr>
<td>2.5 Often loses temper</td>
<td>O</td>
<td>O</td>
<td>O</td>
</tr>
<tr>
<td>2.6 Rather solitary, prefers to play alone</td>
<td>O</td>
<td>O</td>
<td>O</td>
</tr>
<tr>
<td>2.7 Generally well behaved, usually does what adults request</td>
<td>O</td>
<td>O</td>
<td>O</td>
</tr>
<tr>
<td>2.8 Many worries or often seems worried</td>
<td>O</td>
<td>O</td>
<td>O</td>
</tr>
<tr>
<td>2.9 Helpful if someone is hurt, upset or feeling ill</td>
<td>O</td>
<td>O</td>
<td>O</td>
</tr>
<tr>
<td>2.10 Constantly fidgeting or squirming</td>
<td>O</td>
<td>O</td>
<td>O</td>
</tr>
<tr>
<td>2.11 Has at least one good friend</td>
<td>O</td>
<td>O</td>
<td>O</td>
</tr>
<tr>
<td>2.12 Often fights with other children or bullies them</td>
<td>O</td>
<td>O</td>
<td>O</td>
</tr>
<tr>
<td>2.13 Often unhappy, depressed or tearful</td>
<td>O</td>
<td>O</td>
<td>O</td>
</tr>
</tbody>
</table>
### 3. About your child’s SCHOOLING

We would like to ask you for some information about your child’s schooling. This will help us understand how educational needs differ for children with hearing impairments.

*Remember to fill the circles like this •*

#### 3.1 Has your child started attending primary school? (Fill one circle only)
- Yes: Please continue to item 3.2
- No: Please skip to item 3.6

#### 3.2 What is the name of your child’s school? (please specify)

#### 3.3 What is the postcode of your child’s school?

#### 3.4 Including prep year, is this your child’s: (Fill one circle only)
- 1st year of school
- 2nd year of school
- 3rd year of school or more

#### 3.5 What type of school does your child attend? (Fill as many circles as apply)
- Mainstream school
- Mainstream school with a special unit or program for children with hearing loss
- School for children with hearing loss
- Special school for children with other disabilities
- Other (please specify)

#### 3.6 (Only answer this question if your child hasn’t started school yet) Does he/she attend any of the following? (Fill one circle only)
- Day care
- Mainstream program (e.g. Childcare or crèche, with or without kinder program)
- Mainstream setting with special program (e.g. Integration for children with hearing impairment)
- Setting for children with hearing impairment
- Doesn’t attend any setting on a regular basis
- Other (please specify)
4. About your child's HEARING and COMMUNICATION

We would now like to know about your child's hearing impairment, and any devices your child uses in everyday life.

4.1 Has your child ever had hearing aids fitted? (Fill one circle only)
   □ Yes  □ No (go to Question 4.7)

4.2 How old was your child when hearing aids were first fitted? (answer to the nearest month if possible)
   □ years □ months

4.3 Does your child use a hearing aid now?
   □ Yes (Now go to Question 4.4)  □ No
   If No, how old was your child when he/she stopped using hearing aids altogether? (answer to the nearest month if possible)
   □ years □ months (Now go to Question 4.7)

4.4 How often does your child wear hearing aids now? (Fill one circle on each line)

<table>
<thead>
<tr>
<th></th>
<th>Always</th>
<th>Most of the time</th>
<th>Sometimes</th>
<th>Hardly ever/never</th>
</tr>
</thead>
<tbody>
<tr>
<td>a. at home</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>O</td>
</tr>
<tr>
<td>b. at school, kinder or day care</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>O</td>
</tr>
<tr>
<td>c. during social activities</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>O</td>
</tr>
</tbody>
</table>

4.5 How willing is your child to wear hearing aids now? (Fill one circle on each line)

<table>
<thead>
<tr>
<th></th>
<th>Very</th>
<th>Quite</th>
<th>A little</th>
<th>Hardly at all</th>
</tr>
</thead>
<tbody>
<tr>
<td>a. at home</td>
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<td>O</td>
<td>O</td>
</tr>
<tr>
<td>c. during social activities</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>O</td>
</tr>
</tbody>
</table>

4.6 How helpful are the hearing aids for your child? (Fill one circle on each line)

<table>
<thead>
<tr>
<th></th>
<th>Very</th>
<th>Quite</th>
<th>A little</th>
<th>Hardly at all</th>
</tr>
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<td>O</td>
</tr>
<tr>
<td>c. during social activities</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>O</td>
</tr>
</tbody>
</table>
4.7 Has your child ever been evaluated for a cochlear implant? (Fill one circle only)
   O No (go to Question 4.13)
   O Yes, but cochlear implant never fitted (go to Question 4.13)
   O Yes, and cochlear implant fitted

4.8 How old was your child when a cochlear implant was first fitted? (answer to the nearest month if possible)
   [ ] years [ ] months

4.9 Does your child use a cochlear implant now?
   O Yes (Now go to Question 4.10)  O No

If no, how old was your child when he/she stopped using a cochlear implant altogether? (answer to the nearest month if possible)
   [ ] years [ ] months (Now go to Question 4.13)

4.10 How often does your child use the cochlear implant now? (Fill one circle on each line)

<table>
<thead>
<tr>
<th></th>
<th>Always</th>
<th>Most of the time</th>
<th>Sometimes</th>
<th>Hardly ever /never</th>
</tr>
</thead>
<tbody>
<tr>
<td>a. at home</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>O</td>
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<tr>
<td>b. at school, kinder or day care</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>O</td>
</tr>
<tr>
<td>c. during social activities</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>O</td>
</tr>
</tbody>
</table>

4.11 How willing is your child to use the cochlear implant now? (Fill one circle on each line)

<table>
<thead>
<tr>
<th></th>
<th>Very</th>
<th>Quite</th>
<th>A little</th>
<th>Hardly at all</th>
</tr>
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<tbody>
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<td>O</td>
</tr>
<tr>
<td>b. at school, kinder or day care</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>O</td>
</tr>
<tr>
<td>c. during social activities</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>O</td>
</tr>
</tbody>
</table>

4.12 How helpful is the cochlear implant for your child? (Fill one circle on each line)

<table>
<thead>
<tr>
<th></th>
<th>Very</th>
<th>Quite</th>
<th>A little</th>
<th>Hardly at all</th>
</tr>
</thead>
<tbody>
<tr>
<td>a. at home</td>
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<td>O</td>
</tr>
<tr>
<td>b. at school, kinder or day care</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>O</td>
</tr>
<tr>
<td>c. during social activities</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>O</td>
</tr>
</tbody>
</table>

4.13 Has your child ever had a Remote Microphone system (e.g., FM or Roger device) fitted? (Fill one circle only)
   O Yes  O No (go to Question 4.10)
4.14 How old was your child when a Remote Microphone system was first fitted? (answer to the nearest month if possible)

☐ years ☐ months

4.15 Does your child use a Remote Microphone system now?

☐ Yes (Now go to Question 4.16) ☐ No

If no, how old was your child when he/she stopped using a Remote Microphone system altogether? (answer to the nearest month if possible)

☐ years ☐ months (Now go to Question 4.19)

4.16 How often does your child wear the Remote Microphone system now? (Fill one circle on each line)

<table>
<thead>
<tr>
<th></th>
<th>Always</th>
<th>Most of the time</th>
<th>Sometimes</th>
<th>Hardly ever/never</th>
</tr>
</thead>
<tbody>
<tr>
<td>a. at home</td>
<td>☐</td>
<td>☐</td>
<td>☐</td>
<td>☐</td>
</tr>
<tr>
<td>b. at school, kinder or day care</td>
<td>☐</td>
<td>☐</td>
<td>☐</td>
<td>☐</td>
</tr>
<tr>
<td>c. during social activities</td>
<td>☐</td>
<td>☐</td>
<td>☐</td>
<td>☐</td>
</tr>
</tbody>
</table>

4.17 How willing is your child to wear the Remote Microphone system now? (Fill one circle on each line)

<table>
<thead>
<tr>
<th></th>
<th>Very</th>
<th>Quite</th>
<th>A little</th>
<th>Hardly at all</th>
</tr>
</thead>
<tbody>
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<td>☐</td>
<td>☐</td>
</tr>
<tr>
<td>b. at school, kinder or day care</td>
<td>☐</td>
<td>☐</td>
<td>☐</td>
<td>☐</td>
</tr>
<tr>
<td>c. during social activities</td>
<td>☐</td>
<td>☐</td>
<td>☐</td>
<td>☐</td>
</tr>
</tbody>
</table>

4.18 How helpful is the Remote Microphone system for your child? (Fill one circle on each line)

<table>
<thead>
<tr>
<th></th>
<th>Very</th>
<th>Quite</th>
<th>A little</th>
<th>Hardly at all</th>
</tr>
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<td>☐</td>
<td>☐</td>
</tr>
<tr>
<td>b. at school, kinder or day care</td>
<td>☐</td>
<td>☐</td>
<td>☐</td>
<td>☐</td>
</tr>
<tr>
<td>c. during social activities</td>
<td>☐</td>
<td>☐</td>
<td>☐</td>
<td>☐</td>
</tr>
</tbody>
</table>

4.19 Overall, how much do you think your child’s hearing impairment affects his/her everyday life? (Fill one circle on each line)

<table>
<thead>
<tr>
<th></th>
<th>A great deal</th>
<th>Quite a bit</th>
<th>A little</th>
<th>Hardly at all</th>
</tr>
</thead>
<tbody>
<tr>
<td>a. at home</td>
<td>☐</td>
<td>☐</td>
<td>☐</td>
<td>☐</td>
</tr>
<tr>
<td>b. at school, kinder or day care</td>
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<td>☐</td>
<td>☐</td>
<td>☐</td>
</tr>
<tr>
<td>c. during social activities</td>
<td>☐</td>
<td>☐</td>
<td>☐</td>
<td>☐</td>
</tr>
</tbody>
</table>
4.20 Do you know the cause of your child's hearing loss? (Fill one circle only)
   ○ Yes  ○ No
   If Yes, please describe the cause for your child's hearing loss. Causes include syndromes, genetic conditions, pre-natal causes, etc.

4.21 How easy is it to understand your child? (Fill one circle only)
   ○ When speaking in sentences, the child is easy for all listeners to understand. Child is understood easily in everyday situations.
   ○ When speaking in sentences, the child is easy to understand for listeners who have had little experience with a deaf person’s speech.
   ○ When speaking in sentences, the child is possible to understand if the listener is concentrating and lipreading.
   ○ When speaking in sentences, the child is difficult to understand. Single words can be okay to understand when there are context and lipreading cues.
   ○ When speaking in sentences, the child is not able to be understood.

4.22 The next checklist contains a series of statements describing how children communicate. For each statement, you are asked to give information about your child. You are asked to judge whether you have observed that behaviour.

   Please enter a number in the box in the right hand column, as follows:
   0 = less than once a week (or never)
   1 = at least once, but not every day
   2 = once or twice a day
   3 = several times (more than twice) a day (or always)

   a. ☐ Talks repetitively about things that no-one is interested in
   b. ☐ Looks blank in a situation where most children would show a clear facial expression – e.g. when angry, fearful or happy
   c. ☐ Says things that s/he does not seem to fully understand (may appear to be repeating something s/he’s heard an adult say). So, for instance, a 5-year-old girl may be heard to say of a teacher “she’s got a very good reputation”
   d. ☐ Does not look at the person s/he is talking to
   e. ☐ Misses the point of jokes and puns (though may be amused by non-verbal humour such as slapstick)
   f. ☐ Uses favourite phrases, sentences or longer sequences in rather inappropriate contexts. E.g., might say “all of a sudden” rather than “then”, as in “we went to the park and all of a sudden we had a picnic”. Or might habitually start utterances with “by the way”
<p>| | |</p>
<table>
<thead>
<tr>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>g.</strong></td>
<td>Gets confused when a word is used with a different meaning from usual: e.g. might fail to understand if an unfriendly person was described as “cold” (and would assume they were shivering)</td>
</tr>
<tr>
<td><strong>h.</strong></td>
<td>Stands too close to other people when talking to them</td>
</tr>
<tr>
<td><strong>i.</strong></td>
<td>Talks to people too readily: e.g. without any encouragement, starts up a conversation with a stranger</td>
</tr>
<tr>
<td><strong>j.</strong></td>
<td>Pronounces words in an over-precise manner: accent may sound affected or “put-on”, as if child is mimicking a TV personality rather than talking like those around him/her</td>
</tr>
<tr>
<td><strong>k.</strong></td>
<td>Ability to communicate varies from situation to situation - e.g. may cope well when talking one-to-one with a familiar adult, but have difficulty expressing himself in a group of children</td>
</tr>
<tr>
<td><strong>l.</strong></td>
<td>Repeats back what others have just said. For instance, if you ask, “what did you eat?” might say, “what did I eat?”</td>
</tr>
<tr>
<td><strong>m.</strong></td>
<td>Ignores conversational overtures from others (e.g. if asked, “what are you making?” does not look up and just continues working)</td>
</tr>
<tr>
<td><strong>n.</strong></td>
<td>Takes in just 1-2 words in a sentence, and so misinterprets what has been said. E.g. if someone says “I want to go skating next week”, s/he may think they’ve been skating, or want to go now</td>
</tr>
<tr>
<td><strong>o.</strong></td>
<td>It’s difficult to stop him/her from talking</td>
</tr>
<tr>
<td><strong>p.</strong></td>
<td>Tells people things they know already</td>
</tr>
<tr>
<td><strong>q.</strong></td>
<td>Fails to recognise when other people are upset or angry</td>
</tr>
<tr>
<td><strong>r.</strong></td>
<td>Is over-literal, sometimes with (unintentionally) humorous results. E.g., a child who was asked “Do you find it hard to get up in the morning” replied “No. You just put one leg out of the bed and then the other and stand up.” Another child who was told “watch your hands” when using scissors, proceeded to stare at his fingers.</td>
</tr>
<tr>
<td><strong>s.</strong></td>
<td>Includes over-precise information (e.g. exact date or time) in his/her talk, e.g. when asked “when did you go on holiday” may say “13th July 1995” rather than “in the summer”</td>
</tr>
<tr>
<td><strong>t.</strong></td>
<td>Asks a question, even though s/he has been given the answer</td>
</tr>
<tr>
<td><strong>u.</strong></td>
<td>Appreciates the humour expressed by irony. Would be amused rather than confused if someone said “isn’t it a lovely day!” when it is pouring with rain</td>
</tr>
<tr>
<td><strong>v.</strong></td>
<td>Makes good use of gestures to get his/her meaning across</td>
</tr>
</tbody>
</table>
5. About your FAMILY LIFE

Families sometimes have special concerns or difficulties because of their children's health.

Below is a list of things that might be a problem for you. Please tell us how much of a problem each one has been for you during the past ONE month by filling the circle for:

0 if it is never a problem
1 if it is almost never a problem
2 if it is sometimes a problem
3 if it is often a problem
4 if it is always a problem

There are no right or wrong answers.

In the past ONE month, as a result of your child's health, how much of a problem have you had with...

<table>
<thead>
<tr>
<th>Communication (problems with...)</th>
<th>Never (0)</th>
<th>Almost Never (1)</th>
<th>Sometimes (2)</th>
<th>Often (3)</th>
<th>Almost Always (4)</th>
</tr>
</thead>
<tbody>
<tr>
<td>5.1 I feel that others do not understand my family's situation</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>5.2 It is hard for me to talk about my child's health with others</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>5.3 It is hard for me to tell doctors and nurses how I feel</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Worry (problems with...)</th>
<th>Never (0)</th>
<th>Almost Never (1)</th>
<th>Sometimes (2)</th>
<th>Often (3)</th>
<th>Almost Always (4)</th>
</tr>
</thead>
<tbody>
<tr>
<td>5.4 I worry about whether or not my child's medical treatments are working</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>5.5 I worry about the side effects of my child's medications/medical treatments</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
</tbody>
</table>
Below is a list of things that might be a problem for your family. In the past ONE month, as a result of your child’s health, how much of a problem has your family had with ...

<table>
<thead>
<tr>
<th>Daily Activities (problems with...)</th>
<th>Never (0)</th>
<th>Almost Never (1)</th>
<th>Sometimes (2)</th>
<th>Often (3)</th>
<th>Almost Always (4)</th>
</tr>
</thead>
<tbody>
<tr>
<td>5.9 Family activities taking more time and effort</td>
<td>o</td>
<td>o</td>
<td>0</td>
<td>0</td>
<td>o</td>
</tr>
<tr>
<td>5.10 Difficulty finding time to finish household tasks</td>
<td>o</td>
<td>o</td>
<td>0</td>
<td>0</td>
<td>o</td>
</tr>
<tr>
<td>5.11 Feeling too tired to finish household tasks</td>
<td>o</td>
<td>o</td>
<td>0</td>
<td>0</td>
<td>o</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Family Relationships (problems with...)</th>
<th>Never (0)</th>
<th>Almost Never (1)</th>
<th>Sometimes (2)</th>
<th>Often (3)</th>
<th>Almost Always (4)</th>
</tr>
</thead>
<tbody>
<tr>
<td>5.12 Lack of communication between family members</td>
<td>o</td>
<td>o</td>
<td>0</td>
<td>0</td>
<td>o</td>
</tr>
<tr>
<td>5.13 Conflicts between family members</td>
<td>o</td>
<td>o</td>
<td>0</td>
<td>0</td>
<td>o</td>
</tr>
<tr>
<td>5.14 Difficulty making decisions together as a family</td>
<td>o</td>
<td>o</td>
<td>0</td>
<td>0</td>
<td>o</td>
</tr>
<tr>
<td>5.15 Difficulty solving problems together</td>
<td>o</td>
<td>o</td>
<td>0</td>
<td>0</td>
<td>o</td>
</tr>
<tr>
<td>5.16 Stress or tension between family members</td>
<td>o</td>
<td>o</td>
<td>0</td>
<td>0</td>
<td>o</td>
</tr>
</tbody>
</table>

6. About YOU and YOUR PARTNER

We know that a child’s environment and family plays a big part in their development. We would now like to ask some questions about you, as the parent or primary carer of this child, and about your partner.

Please answer within the boxes, or by filling the circles like this ∗

6.1 What is today's date? (Answer within boxes)

```
[ ] [ ] / [ ] / [ ]
```

day / month / year

6.2 Which of the following best describes YOUR relationship to this child? (Fill one circle only)

0 Biological parent 0 Step parent 0 Grandparent
0 Other (please specify) __________
6.3 In general, how would you say your current health is? (Fill one circle only)

- Excellent
- Very good
- Good
- Fair
- Poor

We know that raising young children can at times affect parent's general health and wellbeing. The next question asks about how you have been feeling in the past 2 weeks.

6.4 In the last two weeks, about how often did you feel:

<table>
<thead>
<tr>
<th>Feeling</th>
<th>None of the time</th>
<th>A little of the time</th>
<th>Some of the time</th>
<th>Most of the time</th>
<th>All of the time</th>
</tr>
</thead>
<tbody>
<tr>
<td>a. so sad nothing could cheer you up</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>O</td>
</tr>
<tr>
<td>b. worried or frightened</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>O</td>
</tr>
<tr>
<td>c. restless or stressed</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>O</td>
</tr>
<tr>
<td>d. hopeless</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>O</td>
</tr>
<tr>
<td>e. that everything was an effort</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>O</td>
</tr>
<tr>
<td>f. worthless</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>O</td>
</tr>
</tbody>
</table>

Kessler et al., KB (2002).

6.5 Do you currently have a partner who lives with you (Fill one circle only)

- Yes Please go to question 6.6
- No Please go to question 7.1

We would like to learn more about your child's family, so would now like to know more about your partner.

Please answer within the boxes, or by filling the circles like this •

6.6 Which of the following best describes your PARTNER'S relationship to this child? (Fill one circle only)

- Biological parent
- Step parent
- Grandparent
- Other (please specify) ___________________________

6.7 In general, how would you say your partner's current health is? (Fill one circle only)

- Excellent
- Very good
- Good
- Fair
- Poor

7. About your HOUSEHOLD

As mentioned before, family life and surrounding environment is an important factor for children's development. The next questions ask for updates about your family and household.

Please answer by filling the circles like this •

7.1 What is the main source of income for this household?

<table>
<thead>
<tr>
<th>Source of Income</th>
<th>Two parents working full time</th>
<th>One parent working full time</th>
<th>Two parents working part time</th>
<th>One parent working part time</th>
<th>Pension</th>
<th>Student</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>O</td>
</tr>
</tbody>
</table>

- Other (please describe) ___________________________

16/CHIL 5-7 year questionnaire  Page 12 of 16  HREC 31061  V5 10/09/14
7.2 What is your FAMILY’S annual income? This is the total income of everyone in your household, BEFORE tax, per year. (Fill one circle only)

- Less than $31,199 per year
- $31,200-$51,999 per year
- $52,000-$77,999 per year
- $78,000-$103,999 per year
- $104,000-$155,999 per year
- $156,000 or more per year

7.3 Do you or your partner have a Health Care Card (from Centrelink)? (Fill one circle only)
- Yes
- No

7.4 Do you receive any kind of disability allowance for this child? (Fill one circle only) (E.g., Carer’s Allowance, Child Disability Assistance Payment, Youth Disability Supplement, Mobility Allowance, etc.)
- Yes
- No

7.5 What language is mainly used in your home? (Fill all circles that apply).
- English
- Auslan
- Other, please specify ________________________________

7.6 Are any other languages regularly used in your home?
- No
- Yes, English
- Yes, Auslan
- Yes (please specify) ________________________________

8. About SUPPORT and SERVICES

Families use a range of services for assistance with their children’s hearing loss. The next questions ask about the services you have used over the last 12 months to help with your child. Please indicate if you have used the following services, and how frequent this use is.

Over the last 12 months:

8.1 Did you use Australian Hearing services?
- Yes
- No (go to Question 8.2)

If Yes, how many visits have you made in the past 12 months? ______ visits

8.2 Did you use other audiology services?
- Yes
- No (go to Question 8.3)

If Yes, how many visits have you made in the past 12 months? ______ visits

8.3 Did you use childcare services (including after school care)?
- Yes
- No (go to Question 8.4)

If Yes, how many visits have you made in the past 12 months? ______ visits per week

8.4 Did you use cochlear implant clinic services?
- Yes
- No (go to Question 8.5)

If Yes, how many visits have you made in the past 12 months? ______ visits
8.5 Did you use any **early intervention** services?
   - Yes
   - No (go to Question 8.6)
   If Yes, how many visits have you made in the past 12 months? ______ visits per month

8.6 Did you use **Ear, nose and throat specialist** services?
   - Yes
   - No (go to Question 8.7)
   If Yes, how many visits have you made in the past 12 months? ______ visits

8.7 Did you use **general practitioner (GP)** services?
   - Yes
   - No (go to Question 8.8)
   If Yes, how many visits have you made in the past 12 months? ______ visits

8.8 Did you use **Genetic Services**?
   - Yes
   - No (go to Question 8.9)
   If Yes, how many visits have you made in the past 12 months? ______ visits

8.9 Did you use **maternal child health** services?
   - Yes
   - No (go to Question 8.10)
   If Yes, how many visits have you made in the past 12 months? ______ visits

8.10 Did you use **speech pathologist** services?
   - Yes
   - No
   If Yes, how many visits have you made in the past 12 months? ______ visit

8.11 Did you use any other services?
   - Yes
   - No
   If yes, please specify:

________________________________________________________________________
9. Contact details for you and your nearest relative or friend

We would like to be able to keep in touch in the future, please help us by filling in your current contact details below.

<table>
<thead>
<tr>
<th>Address</th>
<th>Suburb</th>
</tr>
</thead>
<tbody>
<tr>
<td>Postcode</td>
<td>State</td>
</tr>
<tr>
<td>Your Email</td>
<td>Your Mobile</td>
</tr>
</tbody>
</table>

Partner's contact details:

| Partner's Email | Partner's Mobile |

In case you move house, it can be helpful to contact a relative or friend who knows where you've moved. If you can, please write your relative's/friend's details below.

<table>
<thead>
<tr>
<th>Name of relative or friend</th>
</tr>
</thead>
<tbody>
<tr>
<td>Relationship to you (e.g., parent, friend, sister etc.)</td>
</tr>
<tr>
<td>Address</td>
</tr>
<tr>
<td>Postcode</td>
</tr>
<tr>
<td>Email</td>
</tr>
</tbody>
</table>

This is the end of the questionnaire.

Please check that you have answered every question.

If a VicCHILD team member is visiting you, please give them your completed questionnaire.

If not, please check that you have answered all of the questions and return your questionnaires (as well as saliva samples and consent forms, if you have them) in the reply paid envelope.

Questions?

Contact VicCHILD on (03) 9345 4215 / vic-child@rch.org.au

OR visit the VicCHILD website at:

www.rch.org.au/ccch/vic-child

Thank you for your time!
Parents’ Evaluation of Aural/Oral
Performance of Children
(P.E.A.C.H.)

Developed by Teresa Ching & Mandy Hill

<table>
<thead>
<tr>
<th>Child’s Name:</th>
<th>Your Name:</th>
</tr>
</thead>
<tbody>
<tr>
<td>D.O.B.:</td>
<td>Interviewer:</td>
</tr>
<tr>
<td>Number &amp; Interval:</td>
<td>Date:</td>
</tr>
</tbody>
</table>
Parents’ Evaluation of Aural/Oral Performance of Children
(P.E.A.C.H.)

Developed by Teresa Ching & Mandy Hill

What is the PEACH?
- The PEACH (Parents’ Evaluation of Aural/oral performance of Children) is a questionnaire designed to record how your child is hearing and communicating with others when using his/her hearing aids and/or cochlear implant. We ask you to observe your child’s listening behaviour in everyday life and give a rating in relation to a range of hearing and communication scenarios.

The PEACH is not a test. Remember even normal hearing people have some difficulty hearing in some situations. Children’s listening skills improve as they grow and develop and as they get more listening practice.

Why use the PEACH?
- The PEACH is used to evaluate the effectiveness of your child’s hearing aids and/or cochlear implant. Your PEACH ratings will be used to build a picture of your child’s functional performance in everyday life situations. The results can be used by your child’s audiologists to tailor audiological intervention to address the specific difficulties experienced by your child. The PEACH scores collected at several intervals over time can also be used to monitor your child’s progress with intervention.

How do I do it?
- Think about your child’s behaviour over the past week in relation to each question.
- Give a rating, based on the estimated percentage of time that your child displays the described behaviour.

What happens next?
- After you return a completed PEACH, a researcher may contact you to talk through your ratings. The researcher may ask you further questions to make sure they have a thorough understanding of the abilities and needs of your child.

Results from the PEACH will be used to monitor your child’s progress. The information will also be passed onto your child’s audiologist to guide intervention.

Pre-Rating Checklist

<table>
<thead>
<tr>
<th>Question</th>
<th>Yes</th>
<th>No</th>
</tr>
</thead>
<tbody>
<tr>
<td>Has the child been wearing his/her hearing aids and/or cochlear implant?</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Has the child been well/healthy?</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Have the child’s hearing aids and/or cochlear implant been working properly?</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

*If the PEACH is used to assess performance when aided, it should only be completed when the answer to all of the above items is YES.*
Please reflect on your child’s listening behaviour over the past week and circle the appropriate number.

<table>
<thead>
<tr>
<th>Question</th>
<th>Never 0%</th>
<th>Seldom 1 - 25%</th>
<th>Sometimes 26 - 50%</th>
<th>Often 51 - 75%</th>
<th>Always 75-100%</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. How often has your child worn his/her hearing aids and/or cochlear implant?</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
<td>4</td>
</tr>
<tr>
<td>2. How often has your child complained or been upset by loud sounds?</td>
<td>4</td>
<td>3</td>
<td>2</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>3. When you call, does your child respond to his/her name in a quiet situation?</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
<td>4</td>
</tr>
<tr>
<td>4. When asked, does your child follow simple instructions or do a simple task in a quiet situation?</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
<td>4</td>
</tr>
<tr>
<td>5. When you call does your child respond to his/her name in a noisy situation when he/she can’t see your face? (examples of responses include looks up, turns, answers verbally)</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
<td>4</td>
</tr>
<tr>
<td>6. When asked, does your child follow simple instructions or do a simple task in a noisy situation?</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
<td>4</td>
</tr>
<tr>
<td>7. When you are in a quiet place reading with your child, how often does he/she pay close attention to what you are saying? OR if your child is listening to stories/songs on the TV or CD when there is no other background noise how often can he/she follow what is being said?</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
<td>4</td>
</tr>
<tr>
<td>8. How often does your child initiate/participate in conversation in a quiet situation?</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
<td>4</td>
</tr>
<tr>
<td>9. How often does your child initiate/participate in conversation in a noisy situation?</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
<td>4</td>
</tr>
<tr>
<td>10. How often does your child understand what you say in the car/bus/train?</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
<td>4</td>
</tr>
<tr>
<td>11. How often does your child recognise people’s voices without seeing who was talking?</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
<td>4</td>
</tr>
<tr>
<td>12. How often does your child successfully use a phone?</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
<td>4</td>
</tr>
<tr>
<td>13. How often does your child respond to sounds other than voices?</td>
<td>0</td>
<td>1</td>
<td>2</td>
<td>3</td>
<td>4</td>
</tr>
</tbody>
</table>
Please provide comments regarding any of the above items:

__________________________________________________________________________
__________________________________________________________________________
__________________________________________________________________________
__________________________________________________________________________
__________________________________________________________________________
__________________________________________________________________________

Scoring: To be completed by professional

<table>
<thead>
<tr>
<th></th>
<th>RAW Score</th>
<th>% Score</th>
</tr>
</thead>
<tbody>
<tr>
<td>QUIET</td>
<td>(Q ≥ 5+6+7+8+11+12)</td>
<td>(A/24) × 100</td>
</tr>
<tr>
<td>NOISE</td>
<td>(Q ≥ 5+6+8+10+13)</td>
<td>(B/20) × 100</td>
</tr>
<tr>
<td>OVERALL</td>
<td>(A + B)</td>
<td>(C/44) × 100</td>
</tr>
</tbody>
</table>

Copyright 2005 Australian Hearing
To be completed by parent about your child

It is important to know when and in what situations children wear their hearing aids/amplification in order for us to examine the benefits.

1a. On a weekday (Monday-Friday), how many hours a day does your child currently wear their hearing device?

Weekday (Monday-Friday): ___________ hours

1b. On a typical weekday, my child wears his/her hearing device: (Fill all circles that apply in the row below)

<table>
<thead>
<tr>
<th>7-8am</th>
<th>8-9</th>
<th>9-10</th>
<th>10-11</th>
<th>11-12pm</th>
<th>12-1</th>
<th>1-2</th>
<th>2-3</th>
<th>3-4</th>
<th>4-5</th>
<th>5-6</th>
<th>6-7</th>
<th>7-8</th>
<th>8-9</th>
<th>9-10</th>
</tr>
</thead>
<tbody>
<tr>
<td>O</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>O</td>
</tr>
</tbody>
</table>

2a. Are there differences in wearing time on a weekend day (Saturday-Sunday)?

○ Yes  ○ No (go to Question 3)

Weekend (Saturday-Sunday): ___________ hours

2b. On a typical weekend, my child wears his/her hearing device: (Fill all circles that apply in the row below)

<table>
<thead>
<tr>
<th>7-8am</th>
<th>8-9</th>
<th>9-10</th>
<th>10-11</th>
<th>11-12pm</th>
<th>12-1</th>
<th>1-2</th>
<th>2-3</th>
<th>3-4</th>
<th>4-5</th>
<th>5-6</th>
<th>6-7</th>
<th>7-8</th>
<th>8-9</th>
<th>9-10</th>
</tr>
</thead>
<tbody>
<tr>
<td>O</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>O</td>
</tr>
</tbody>
</table>

3. Overall, how much of the time does your child use their hearing aid and/or cochlear implant and/or remote microphone in the following situations? Fill one circle only for each row.

<table>
<thead>
<tr>
<th>Situation</th>
<th>Never (6)</th>
<th>Seldom (1)</th>
<th>Sometimes (2)</th>
<th>Often (3)</th>
<th>Always (4)</th>
<th>Don't Know (5)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Play with parents</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>O</td>
</tr>
<tr>
<td>Play alone</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>O</td>
</tr>
<tr>
<td>Play with other children</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>O</td>
</tr>
<tr>
<td>Mealtime</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>O</td>
</tr>
<tr>
<td>Public places</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>O</td>
</tr>
<tr>
<td>In the car</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>O</td>
</tr>
<tr>
<td>At preschool or school</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>O</td>
</tr>
<tr>
<td>TV/computer/iPad</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>O</td>
<td>O</td>
</tr>
</tbody>
</table>
4. Think about your child in each of the activities listed below. When they are doing this activity, what hearing device are they using? Fill all circles that apply for each row.

<table>
<thead>
<tr>
<th>Situation</th>
<th>Hearing Aid Used</th>
<th>Cochlear Implant Used</th>
<th>Remote Microphone Used</th>
<th>Don’t Know</th>
<th>Not Applicable</th>
</tr>
</thead>
<tbody>
<tr>
<td>Play with parents</td>
<td>○</td>
<td>○</td>
<td>○</td>
<td>○</td>
<td>○</td>
</tr>
<tr>
<td>Play alone</td>
<td>○</td>
<td>○</td>
<td>○</td>
<td>○</td>
<td>○</td>
</tr>
<tr>
<td>Play with other children</td>
<td>○</td>
<td>○</td>
<td>○</td>
<td>○</td>
<td>○</td>
</tr>
<tr>
<td>Meal times</td>
<td>○</td>
<td>○</td>
<td>○</td>
<td>○</td>
<td>○</td>
</tr>
<tr>
<td>Public places</td>
<td>○</td>
<td>○</td>
<td>○</td>
<td>○</td>
<td>○</td>
</tr>
<tr>
<td>In the car</td>
<td>○</td>
<td>○</td>
<td>○</td>
<td>○</td>
<td>○</td>
</tr>
<tr>
<td>At preschool or school</td>
<td>○</td>
<td>○</td>
<td>○</td>
<td>○</td>
<td>○</td>
</tr>
<tr>
<td>TV/computer/iPad</td>
<td>○</td>
<td>○</td>
<td>○</td>
<td>○</td>
<td>○</td>
</tr>
</tbody>
</table>
To be completed by Parent/Guardian

Version A

1) Tomato
   0 fly 0 croak
   0 wood 0 dunce
   0 fruit 0 step
   0 corn 0 arm

2) Toss
   0 throw 0 catch
   0 hide 0 roll
   0 dive 0 pull

3) Damp
   0 light 0 bag
   0 sweet 0 letter
   0 wet 0 flag

4) Rest
   0 dry 0 sing
   0 go away 0 taste
   0 run up 0 lie down

5) Cruel
   0 clean 0 green
   0 pretty 0 found
   0 water 0 unkind

6) Receive
   0 walk 0 accept
   0 believe 0 empty
   0 money 0 drive

7) Battle
   0 stroll 0 light
   0 snow 0 fight
   0 bowl 0 last

8) Patch
   0 mend 0 watch
   0 hand 0 bang
   0 switch 0 cook

9) Disturb
   0 transfer 0 skip
   0 link 0 upset
   0 doubt 0 fire

10) Blaze
    0 kitchen 0 coat
    0 grass 0 root
    0 flare 0 side

11) Malaria
    0 basement 0 fever
    0 theatre 0 fruit
    0 ocean 0 tune

12) Fascinated
    0 illustrated 0 modelled
    0 poisoned 0 charmed
    0 frightened 0 copied

13) Liberty
    0 freedom 0 worry
    0 rich 0 serviette
    0 forest 0 cheerful

14) Stubborn
    0 steady 0 hopeful
    0 obstinate 0 hollow
    0 orderly 0 slack

15) Precise
    0 natural 0 exact
    0 faulty 0 grand
    0 stupid 0 small

16) Resemblance
    0 memory 0 fondness
    0 assemble 0 repose
    0 attendance 0 likeness

17) Anonymous
    0 applicable 0 magnificent
    0 insulting 0 fictitious
    0 nameless 0 untrue

18) Elevate
    0 raise 0 move
    0 resolve 0 work
    0 waver 0 disperse

Please turn over the page....
<table>
<thead>
<tr>
<th>19) task</th>
<th>29) bombastic</th>
<th>39) vagary</th>
</tr>
</thead>
<tbody>
<tr>
<td>horn</td>
<td>democratic</td>
<td>vagabond</td>
</tr>
<tr>
<td>trap</td>
<td>bombastic</td>
<td>obscure</td>
</tr>
<tr>
<td>problem</td>
<td>anxious</td>
<td>vulgarity</td>
</tr>
<tr>
<td></td>
<td></td>
<td>evasion</td>
</tr>
<tr>
<td></td>
<td></td>
<td>fallacy</td>
</tr>
</tbody>
</table>

| 20) courteous | 30) levity  | 40) spacious |
|               |            |              |
| dreadful      | parsonry   | fallacious   |
| polite        | frivolity  | coeval      |
| curtesy       | salutary   | palatial    |
|               | villainy   | typical     |
|               | taniff     | nutritious   |
|               |            | flexible     |

| 21) prosper | 31) whim  | 41) sedulous |
|            |          |              |
| imagine    | complain  | rebellious   |
| succeed    | noise     | dilatory     |
|           |          |              |
|           |          |              |

| 22) lavish | 32) ruse  | 42) nugatory |
|           |          |              |
| unaccountable | limb   | inimitable   |
| romantic   | noise    |              |
| extravagant | trick   |              |
|            |          |              |
|            |          |              |

| 23) immerse | 33) recumbant | 43) adumbrate |
|             |              |              |
| frequent    | fugitive     | forshadow    |
| reverse     | uncomfortable | protect      |
| rise        | unwieldy     |              |
|            | repelling    |              |
|            |              |              |

| 24) consolidate | 34) quenulous | 44) minatory |
|                |              |              |
| congregate    | assertive    | implacable   |
| party         | fearful      |              |
| compress      | petulant     |              |
|              | curious      |              |
|              | inquiring    |              |
|              | spurious     |              |

| 25) envisage | 35) temerity | 45) minatory |
|             |              |              |
| enfeebled   | impatience   | diminutive   |
| surround    | rashness     |              |
| activate    | nervousness  |              |
|            | stability    |              |
|            | punctuality  |              |
|            |              |              |

| 26) amulet | 36) fecund | 46) minatory |
|           |            |              |
| cameo      | ascendent   |              |
| flirtation |族将    |              |
| charm      | querulous   |              |
|            |              |              |

| 27) garrulous | 37) abrogate | 47) minatory |
|              |            |              |
| talkative    | contradict  |              |
| massive      | doery      |              |
| ridiculous   | renounce    |              |
|            | execute    |              |
|            | belle      |              |
|            | assemble   |              |

| 28) libertine | 38) traduce | 48) minatory |
|              |            |              |
| profligate   | challenge   |              |
| fast         | suspend    |              |
|             | establish  |              |
|             |             |              |

| 29) bombastic | 39) vagary  | 40) spacious |
|              |            |              |
|              |            |              |

| 30) levity  | 41) sedulous | 42) nugatory |
|            |              |              |
|            |              |              |

| 43) adumbrate | 44) minatory | 45) minatory |
|              |              |              |
|              |              |              |

| 46) minatory | 47) minatory | 48) minatory |
|              |              |              |
|              |              |              |

| 49) minatory | 50) minatory | 51) minatory |
|              |              |              |
|              |              |              |

| 52) minatory | 53) minatory | 54) minatory |
|              |              |              |
|              |              |              |

| 55) minatory | 56) minatory | 57) minatory |
|              |              |              |
|              |              |              |

| 58) minatory | 59) minatory | 60) minatory |
|              |              |              |
|              |              |              |

| 61) minatory | 62) minatory | 63) minatory |
|              |              |              |
|              |              |              |
AQL-4D Basic Data Collection Copy

Tick the box next to the response that best fits your situation

1. Do you need any help looking after yourself? (For example: dressing, bathing, eating)
   - I need no help at all
   - Occasionally I need some help with personal care tasks
   - I need help with the more difficult personal care tasks
   - I need daily help with most or all personal care tasks.

2. When doing household tasks: (For example: cooking, cleaning the house, washing)
   - I need no help at all
   - Occasionally I need some help with household tasks
   - I need help with the more difficult household tasks
   - I need daily help with most or all household tasks.

3. Thinking about how easily you can get around your home and community:
   - I get around my home and community by myself without any difficulty
   - I find it difficult to get around my home and community by myself
   - I cannot get around the community by myself, but I can get around my home with some difficulty
   - I cannot get around either the community or my home by myself.

4. Because of your health, your relationships (for example: with your friends, partner or parents) generally:
   - Are very close and warm
   - Are sometimes close and warm
   - Are seldom close and warm
   - I have no close and warm relationships

5. Thinking about your relationship with other people:
   - I have plenty of friends, and am never lonely
   - Although I have friends, I am occasionally lonely
   - I have some friends, but am often lonely for company
   - I am socially isolated and feel lonely.

6. Thinking about your health and my relationship with my family:
   - My role in the family is unaffected by my health
   - There are some parts of my family role I cannot carry out
   - There are many parts of my family role I cannot carry out
   - I cannot carry out any part of my family role.

7. Thinking about your vision, including when using your glasses or contact lenses if needed:

Centre for Health Economics, Monash University
Appendix F: Hearing aid processes documents

Post assessment hearing device activities

- AK provided Noah files coded according to AHCIS number on USB, and hearing devices
- PC imported Noah files and identified child, and most recent Noah file in situation of multiples
- PC renamed Noah files using VicCHILD ID number, stored within Noah 3.0 held at University of Melbourne Audiology Clinic
- From each Noah file, PC identified aid fitted, appropriate date of fitting to use to simulate child’s hearing aid and data logging details
- PC programmed hearing aid via HIPRO to replicate aid settings when child conducted VicCHILD ax for the child’s better hearing ear based off audiograms supplied to VicCHILD by AH
- PC then switched off all adaptive features – directionality, noise reduction, whistle block, etc for all programs (basically any adaptive features that may influence the hearing aid performance in the test box). Also frequency compression was checked – if it were in action then the FCC and FCR needed to be noted down (hasn’t happened yet – see supervision notes from TF 16/07 for more details)
- Hearing device was then disconnected and Affinity launched through the Noah interface
- HIT tab selected from RHS of screen, correct ear selected, protocol to use was Clinical HIT Peter, with speech stimuli ISTS, 2cc coupler, plotting hearing aid output/response to 4 input levels (50, 65, 80, MPO – likely 90)
- Screenshot of response curves taken when all four fully visible on graph, added to document participant test box screen shots.doc
- Using cross hair tool from LHS of HIT screen, selecting each curve, output was recorded for all required frequencies and recorded in excel document 5-7 post ax hearing devices and logging info
- Process was then repeated for each participant
- At end of process, all participant data is to be deleted from the University of Melbourne Audiology Clinic system

Next steps relate to using test box measures in SHARP to obtain SII

- After launching SHARP program, new file allows entry of data by hand, specifying ear, transducer, age and participant ID. Stimuli is left on Carrots, RECD values are apparently redundant but age specific values entered from Dillon’s Hearing Aids book. 80 curve is redundant but still recorded.
  After saving, the main SHARP window displays the calculation of SII – ear needs to be specific, Speaker switched to female, stimulus remains as carrots and mode needs to stay as real ear

- SII needs to be recorded for five situations; soft, medium and loud speech, average conversation at 1m and average conversation at 4m
- Screenshots can be taken, but as raw data table is saved within the program then there is no need to really do this
- Note from 30/11/15: when choosing which was the better ear, the 3FA was considered first. If both ears were equal, 4FA was considered, and the better hearing ear was identified. 3FA was always recorded as the PTA. Except for V0139 for whom LE was better 3FA, but RE was better 4FA. For this participant, for consistency the 3FA was chosen.
1. Obtain Noah file and hearing devices
   a. 06/07/2015 – collecting Noah files and 4 models of the 7 required devices
      i. When opening files will be able to determine which device is being used
      ii. Remaining devices are being sourced, additional Noah file is being sourced
2. Create location for Noah files on Unimeb computer
   a. AH requirement to re-name the Noah files to remove identifying information as much as possible
3. Interested in Noah files from fitting prior to the language assessment case for fine tuning details purposes
   a. Need dates listed for participants to use as reference - DONE
4. Obtain data logging information from either date after language assessment date OR a period of time in 2015
   a. Important to work out the breakdown between these two possible time points – re worthwhile calling clinician for more up to date data logging use values if possible.
5. Upload Noah file to appropriate hearing aid one by one, stick with one hearing aid type first to minimise changes
6. Obtain required information for SHARP
   a. Exact requirements outlined in figure below:
      i. Thresh HL audiograms exist at MCRY
      ii. Need to obtain simulated real-ear dB SPL for a series of input levels
         1. Minimum of two levels required, better to get three: 55/55/75
      iii. MPO values
7. Put this information into excel spreadsheet while on site at Unimeb
8. Once have all information required from hearing aids, delete Noah files from Unimeb computer
Appendix G: ELQ cover letter and questionnaire

Dear «Parent_1_First_Name»

Thank you for being part of VicCHILD (the Victorian Childhood Hearing Impairment Longitudinal Databank). Nearly 450 families across Victoria have now joined VicCHILD. With your help, we are learning more about hearing impairment and how children reach the best outcomes.

We were last in contact when you and «Child_First_Name» joined VicCHILD. Now that «Child_First_Name» is between 1 and 3 years of age, we are keen to find out how his/her language is developing. We hope you’ll tell us by filling out two questionnaires. The white ‘PEACH’ questionnaire asks about «Child_First_Name»’s hearing and communication. The blue questionnaire asks about «Child_First_Name»’s hearing, language and health, plus a few questions about you and your family. Altogether, they take around 30 minutes.

Your answers will help understand why and how some children develop language more easily than others. Many countries have started to collect similar information at this age. Your answers will also tell us how Victorian toddlers with hearing impairment are doing compared to other parts of the world.

To take part:
- Fill out the blue and white questionnaires
- Return them in the reply-paid envelope - if possible, within the next 14 days.

Questions? Call VicCHILD on (03) 9345 4215 or vic-child@rch.org.au. Thank you again for your time!

<table>
<thead>
<tr>
<th>Professor Melissa Wolfe</th>
<th>Dr. Zafia Poulos</th>
<th>Dr. Sherryn Tobin</th>
</tr>
</thead>
<tbody>
<tr>
<td>Associate Director of Research</td>
<td>Clinical Psychologist</td>
<td>Project Coordinator</td>
</tr>
<tr>
<td>The Royal Children’s Hospital Centre for Community Child Health</td>
<td>The Royal Children’s Hospital Centre for Community Child Health</td>
<td>The Royal Children’s Hospital Centre for Community Child Health</td>
</tr>
</tbody>
</table>

HREC 31081 Cover Letter_ELQ_V2 Page 1 of 1 Dated 20 May 2015
Early Language Questionnaire

Thank you for your continued support of VicCHILD! This questionnaire asks you about your child with a hearing loss, and about you and your family. It takes about 30 minutes to complete. It is private, and your answers are confidential.

Instructions:
- Please fill in the circles like this ☐ (do not tick or cross).
- If you make a mistake, put a cross through it ☒. Then fill in and put a circle around the correct one ☐.
- Some questions ask you to write extra information (e.g. please specify ________).
- Please use a blue or black pen.
- Try to answer all questions. If you’re not sure just give your best guess!

Please send this back to us in the reply-paid envelope.

Questions? Call VicCHILD on (03) 9345 4215 or email vic-child@rch.org.au
### 1. About your CHILD

1.1 In general, how would you say your child's current health is? *(Fill one circle only)*

- ○ Excellent
- ○ Very good
- ○ Good
- ○ Fair
- ○ Poor

Below is a list of things that might be a problem for your child. Please tell us how much of a problem each one has been for your child during the past ONE month by filling the circle for:

0 if it is never a problem

1 if it is almost never a problem

2 if it is sometimes a problem

3 if it is often a problem

4 if it is always a problem

There are no right or wrong answers. Some may not fully apply to your child yet – just do the best you can.

In the past ONE month, how much of a problem has your child had with...

<table>
<thead>
<tr>
<th>Physical Functioning (problems with...)</th>
<th>Never (0)</th>
<th>Almost Never (1)</th>
<th>Sometimes (2)</th>
<th>Often (3)</th>
<th>Almost Always (4)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1.2 Walking</td>
<td>○</td>
<td>○</td>
<td>○</td>
<td>○</td>
<td>○</td>
</tr>
<tr>
<td>1.3 Running</td>
<td>○</td>
<td>○</td>
<td>○</td>
<td>○</td>
<td>○</td>
</tr>
<tr>
<td>1.4 Participating in active play or exercise</td>
<td>○</td>
<td>○</td>
<td>○</td>
<td>○</td>
<td>○</td>
</tr>
<tr>
<td>1.5 Lifting something heavy</td>
<td>○</td>
<td>○</td>
<td>○</td>
<td>○</td>
<td>○</td>
</tr>
<tr>
<td>1.6 Helping to pick up his or her toys</td>
<td>○</td>
<td>○</td>
<td>○</td>
<td>○</td>
<td>○</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Emotional Functioning (problems with...)</th>
<th>Never (0)</th>
<th>Almost Never (1)</th>
<th>Sometimes (2)</th>
<th>Often (3)</th>
<th>Almost Always (4)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1.7 Feeling afraid or scared</td>
<td>○</td>
<td>○</td>
<td>○</td>
<td>○</td>
<td>○</td>
</tr>
<tr>
<td>1.8 Feeling sad or blue</td>
<td>○</td>
<td>○</td>
<td>○</td>
<td>○</td>
<td>○</td>
</tr>
<tr>
<td>1.9 Feeling angry</td>
<td>○</td>
<td>○</td>
<td>○</td>
<td>○</td>
<td>○</td>
</tr>
<tr>
<td>1.10 Worrying</td>
<td>○</td>
<td>○</td>
<td>○</td>
<td>○</td>
<td>○</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Social Functioning (problems with...)</th>
<th>Never (0)</th>
<th>Almost Never (1)</th>
<th>Sometimes (2)</th>
<th>Often (3)</th>
<th>Almost Always (4)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1.11 Playing with other children</td>
<td>○</td>
<td>○</td>
<td>○</td>
<td>○</td>
<td>○</td>
</tr>
<tr>
<td>1.12 Other kids not wanting to play with him or her</td>
<td>○</td>
<td>○</td>
<td>○</td>
<td>○</td>
<td>○</td>
</tr>
<tr>
<td>1.13 Getting teased by other children</td>
<td>○</td>
<td>○</td>
<td>○</td>
<td>○</td>
<td>○</td>
</tr>
</tbody>
</table>

*Please complete this section if your child attends childcare or daycare*

<table>
<thead>
<tr>
<th>Childcare/Daycare Functioning (problems with...)</th>
<th>Never (0)</th>
<th>Almost Never (1)</th>
<th>Sometimes (2)</th>
<th>Often (3)</th>
<th>Almost Always (4)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1.14 Doing the same childcare/daycare activities as peers</td>
<td>○</td>
<td>○</td>
<td>○</td>
<td>○</td>
<td>○</td>
</tr>
<tr>
<td>1.15 Missing childcare/daycare because of not feeling well</td>
<td>○</td>
<td>○</td>
<td>○</td>
<td>○</td>
<td>○</td>
</tr>
<tr>
<td>1.16 Missing childcare/daycare to go to the doctor or hospital</td>
<td>○</td>
<td>○</td>
<td>○</td>
<td>○</td>
<td>○</td>
</tr>
</tbody>
</table>
2. About your CHILD’S hearing loss

2.1 What is today’s date?

Day / Month / Year

2.2 Do you know the cause of your child’s hearing loss? (Fill one circle only)

O Yes       O No

If Yes, please describe the cause for your child’s hearing loss. Causes include syndromes, genetic conditions, prenatal causes, etc.

2.3 Has your child ever had hearing aids fitted? (Fill one circle only)

O Yes       O No (go to Question 2.4)

2.4 How old was your child when hearing aids were first fitted?

(Answer to the nearest month if possible)

[ ] years  [ ] months

2.5 Does your child use a hearing aid now? (Fill one circle only)

O Yes       O No

If Yes, which Australian Hearing Centre do you attend? (Please specify below)

[ ]

(Now go to Question 2.6)

If No, how old was your child when he/she stopped using hearing aids altogether?

(Answer to the nearest month if possible)

[ ] years  [ ] months (Now go to Question 2.6)

2.6 Has your child ever been evaluated for a cochlear implant? (Fill one circle only)

O No (go to Question 2.5)

O Yes, but cochlear implant never fitted (go to Question 2.7)

O Yes, and cochlear implant fitted

2.7 How old was your child when a cochlear implant was first fitted?

(Answer to the nearest month if possible)

[ ] years  [ ] months
2.3  Does your child use a cochlear implant now?  (Fill one circle only)

- Yes  (Now go to Question 2.3)
- No

If no, how old was your child when he/she stopped using a cochlear implant altogether?
(Answer to the nearest month if possible)

[ ] 3 [ ] 4 [ ] 5 [ ] 6 [ ] 7 [ ] 8 [ ] 9 [ ] 10 [ ] 11 [ ] 12 [ ]

2.9  It is important to know when children wear their hearing aids/amplification in order for us to understand the benefits.

If your child does not wear any hearing device, go to Question 2.12
(a) About how many hours per day does your child currently wear their hearing device:

On a weekday (Monday-Friday) [ ] [ ] hours

On a weekend day (Saturday/Sunday) [ ] [ ] hours

(b) On a typical day, which periods does your child wear his/her hearing device?
(Fill all circles that apply in the rows below)

<table>
<thead>
<tr>
<th>Typical day</th>
<th>Morning</th>
<th>Lunch/Afternoon</th>
<th>Evening</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>8-7am</td>
<td>7-8</td>
<td>8-9</td>
</tr>
<tr>
<td>Weekday</td>
<td>[ ]</td>
<td>[ ]</td>
<td>[ ]</td>
</tr>
<tr>
<td></td>
<td>9-10</td>
<td>10-11</td>
<td>11-12</td>
</tr>
<tr>
<td></td>
<td>[ ]</td>
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</tr>
<tr>
<td></td>
<td>12-1pm</td>
<td>1-2</td>
<td>2-3</td>
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<td>[ ]</td>
<td>[ ]</td>
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<tr>
<td></td>
<td>3-4</td>
<td>4-5</td>
<td>5-6</td>
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<td>[ ]</td>
<td>[ ]</td>
</tr>
<tr>
<td></td>
<td>6-7</td>
<td>7-8</td>
<td>7-9pm</td>
</tr>
<tr>
<td></td>
<td>[ ]</td>
<td>[ ]</td>
<td>[ ]</td>
</tr>
</tbody>
</table>

2.10  In general, how much of the time does your child use ANY of their hearing devices in the following situations? Please think about all of this child’s hearing devices (e.g., hearing aids, cochlear implants). (Fill one circle on each row)

<table>
<thead>
<tr>
<th>Situation</th>
<th>Never</th>
<th>Seldom</th>
<th>Sometimes</th>
<th>Often</th>
<th>Always</th>
<th>Don’t know</th>
</tr>
</thead>
<tbody>
<tr>
<td>Play with parents</td>
<td>[ ]</td>
<td>[ ]</td>
<td>[ ]</td>
<td>[ ]</td>
<td>[ ]</td>
<td>[ ]</td>
</tr>
<tr>
<td>Play alone</td>
<td>[ ]</td>
<td>[ ]</td>
<td>[ ]</td>
<td>[ ]</td>
<td>[ ]</td>
<td>[ ]</td>
</tr>
<tr>
<td>Play with other children</td>
<td>[ ]</td>
<td>[ ]</td>
<td>[ ]</td>
<td>[ ]</td>
<td>[ ]</td>
<td>[ ]</td>
</tr>
<tr>
<td>Meal times</td>
<td>[ ]</td>
<td>[ ]</td>
<td>[ ]</td>
<td>[ ]</td>
<td>[ ]</td>
<td>[ ]</td>
</tr>
<tr>
<td>Public places</td>
<td>[ ]</td>
<td>[ ]</td>
<td>[ ]</td>
<td>[ ]</td>
<td>[ ]</td>
<td>[ ]</td>
</tr>
<tr>
<td>In the car</td>
<td>[ ]</td>
<td>[ ]</td>
<td>[ ]</td>
<td>[ ]</td>
<td>[ ]</td>
<td>[ ]</td>
</tr>
<tr>
<td>At day care or kinder</td>
<td>[ ]</td>
<td>[ ]</td>
<td>[ ]</td>
<td>[ ]</td>
<td>[ ]</td>
<td>[ ]</td>
</tr>
<tr>
<td>TV/computer/Pad</td>
<td>[ ]</td>
<td>[ ]</td>
<td>[ ]</td>
<td>[ ]</td>
<td>[ ]</td>
<td>[ ]</td>
</tr>
</tbody>
</table>
2.11 Think about your child in each of the activities listed below. When they are doing this activity, what hearing device do they usually use? (Fill all circles that apply for each row.)

<table>
<thead>
<tr>
<th>Situation</th>
<th>Hearing aid used</th>
<th>Cochlear implant used</th>
<th>Don’t know</th>
<th>Doesn’t apply</th>
</tr>
</thead>
<tbody>
<tr>
<td>Play with parents</td>
<td>○</td>
<td>○</td>
<td>○</td>
<td>○</td>
</tr>
<tr>
<td>Play alone</td>
<td>○</td>
<td>○</td>
<td>○</td>
<td>○</td>
</tr>
<tr>
<td>Play with other children</td>
<td>○</td>
<td>○</td>
<td>○</td>
<td>○</td>
</tr>
<tr>
<td>Meals times</td>
<td>○</td>
<td>○</td>
<td>○</td>
<td>○</td>
</tr>
<tr>
<td>Public places</td>
<td>○</td>
<td>○</td>
<td>○</td>
<td>○</td>
</tr>
<tr>
<td>In the car</td>
<td>○</td>
<td>○</td>
<td>○</td>
<td>○</td>
</tr>
<tr>
<td>At day care or kinder</td>
<td>○</td>
<td>○</td>
<td>○</td>
<td>○</td>
</tr>
<tr>
<td>TV/computer/Pad</td>
<td>○</td>
<td>○</td>
<td>○</td>
<td>○</td>
</tr>
</tbody>
</table>

2.12 Overall, how much do you think your child’s hearing impairment affects his/her everyday life? (Fill one circle on each row)

<table>
<thead>
<tr>
<th>Situation</th>
<th>A great deal</th>
<th>Quite a bit</th>
<th>A little</th>
<th>Hardly at all</th>
</tr>
</thead>
<tbody>
<tr>
<td>a. At home</td>
<td>○</td>
<td>○</td>
<td>○</td>
<td>○</td>
</tr>
<tr>
<td>b. At school, kinder or day care</td>
<td>○</td>
<td>○</td>
<td>○</td>
<td>○</td>
</tr>
<tr>
<td>c. During social activities</td>
<td>○</td>
<td>○</td>
<td>○</td>
<td>○</td>
</tr>
</tbody>
</table>

3. About your CHILD and EARLY INTERVENTION

We would now like to ask some questions about any Early Intervention that your child has received because of their hearing loss:

3.1 Has your child ever been enrolled with or had visits from an Early Intervention service for children with hearing loss?
   ○ Yes ○ No (Now go to Section 4)

3.2 Is your child currently enrolled with or having visits from an Early Intervention service for children with hearing loss?
   ○ Yes ○ No (Now go to Question 3, 4)

3.3 Does your child currently attend or have visits from any of these services? (Fill one circle only)
   ○ Aurora School Early Intervention
   ○ Early Education Program for Hearing Impaired Children (EEP)
   ○ Taralga Oral Language Centre Hearing Deaf Children
   ○ Parent Advisor (Dianella, Bencigo)
   ○ RIDE (Tasla-Health)
   ○ None of the above

3.4 How old was your child when they started with this Early Intervention service?
   (Round to the nearest month if possible)

   [ ] years   [ ] months
3.5 Has your child ever attended another Early Intervention service for hearing loss?  
C Yes C No (Now go to Section 4)  

<table>
<thead>
<tr>
<th>Service</th>
<th>Yes</th>
<th>No</th>
<th>Start</th>
<th>Finish</th>
</tr>
</thead>
<tbody>
<tr>
<td>Aurora School Early Intervention</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Early Education Program for Hearing Impaired</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Children (EEP)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Jalada oral language centre for deaf children</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Parent Advisor (Dianella, Bendigo)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>RIDDC (Tele-health)</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Other: Please specify</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Other: Please specify</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Other: Please specify</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

4. About your CHILD’S language and vocabulary

Vocabulary Checklist
Children understand many more words than they say. We are particularly interested in the words your child SAYS. Please go through the list and mark the words you have heard your child use. If your child uses a different pronunciation of a word (for example, “ottle” instead of “giraffe” or “spagh” for “spaghetti”), mark the word anyway. Remember that this is a “catalogue” of all the words that are used by many different children. Don’t worry if your child knows only a few of these right now.

Please note, this checklist looks long, but it is actually very quick to complete!

<table>
<thead>
<tr>
<th>4.1 SOUND EFFECTS AND ANIMAL SOUNDS</th>
<th></th>
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<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>bee, buzz</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>cock-a-doodle</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>croak</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>oink</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>quack</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>quick quack</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>ugh, oh</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>vroom</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>woof woof</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>yum, yum</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
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</table>

<table>
<thead>
<tr>
<th>4.2 ANIMAL NAMES (Real or Toy)</th>
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<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>alligator</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>bear</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>bee</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>bird</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>bug</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>bunny</td>
<td></td>
<td></td>
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<td></td>
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<tr>
<td>caterpillar</td>
<td></td>
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</tr>
<tr>
<td>duck</td>
<td></td>
<td></td>
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<tr>
<td>elephant</td>
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</tr>
<tr>
<td>falcon</td>
<td></td>
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<td></td>
<td></td>
</tr>
<tr>
<td>fox</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>giraffe</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>goose</td>
<td></td>
<td></td>
<td></td>
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</tr>
<tr>
<td>horse</td>
<td></td>
<td></td>
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</tr>
<tr>
<td>penguin</td>
<td></td>
<td></td>
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</tr>
<tr>
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</tr>
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<tr>
<td>rooster</td>
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</tr>
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</tr>
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</tr>
<tr>
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</tr>
<tr>
<td>snail</td>
<td></td>
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</tr>
<tr>
<td>snake</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>turtle</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>tiger</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Item</td>
<td>Item</td>
<td>Item</td>
<td></td>
<td></td>
</tr>
<tr>
<td>----------------------</td>
<td>----------------------</td>
<td>----------------------</td>
<td></td>
<td></td>
</tr>
<tr>
<td>cat</td>
<td>lamb</td>
<td>turkey</td>
<td></td>
<td></td>
</tr>
<tr>
<td>chicken</td>
<td>lion</td>
<td>turtle</td>
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<td></td>
</tr>
<tr>
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<td>monkey</td>
<td>wolf</td>
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<tr>
<td>deer</td>
<td>wombat</td>
<td>zebra</td>
<td></td>
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</tr>
<tr>
<td>dog</td>
<td>mouse</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>donkey</td>
<td>owl</td>
<td></td>
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<td></td>
</tr>
</tbody>
</table>

### 4.3 VEHICLES (Real or Toy)
- airplane
- firetruck
- tractor
- bicycle
- helicopter
- train
- boat
- motorcycle
- bicycle
- bus
- sled
- truck
- car
- pusher

### 4.4 TOYS
- ball
- chalk
- pencil
- balloon
- crayon
- play dough
- bat
- doll
- present
- block
- game
- puzzle
- book
- glue
- story
- bubbles
- pen
- toy

### 4.5 FOOD AND DRINK
- apple
- French fries
- popcorn
- banana
- grapes
- strawberries
- beans
- green beans
- potato
- bread
- gum
- potato chip
- butter
- hamburger
- pretzel
- cake
- ice
- pudding
- olives
- ice cream
- pumpkin
- carrots
- jam
- raisin
- cereal
- jelly
- salt
- cheese
- juice
- sandwich
- chicken
- meat
- soda pop
- chocolate
- melon
- soup
- coffee
- milk
- spaghetti
- coke
- muffin
- strawberry
- biscuit
- noodles
- toast
- corn
- nuts
- tuna
- croaker
- orange
- vanilla
- donut
- pancake
- vitamins
- drink
- peanut butter
- water
- egg
- peas
- yoghurt
- fish
- piyama
- jumper

### 4.6 CLOTHING
- beads
- jacket
- slipper
- belt
- jeans
- runners
- bib
- mittens
- tethers
- boots
- necklace
- sook
- button
- pyjama
- jumper
<table>
<thead>
<tr>
<th>coat</th>
<th>pants</th>
<th>sights</th>
</tr>
</thead>
<tbody>
<tr>
<td>nappy</td>
<td>scarf</td>
<td>underpants</td>
</tr>
<tr>
<td>dress</td>
<td>skirt</td>
<td>zipper</td>
</tr>
<tr>
<td>gloves</td>
<td>shoe</td>
<td></td>
</tr>
<tr>
<td>hat</td>
<td>shorts</td>
<td></td>
</tr>
</tbody>
</table>

4.7 **BODY PARTS** *or words used in your family*

<table>
<thead>
<tr>
<th>ankle</th>
<th>feet</th>
<th>nose</th>
</tr>
</thead>
<tbody>
<tr>
<td>arm</td>
<td>finger</td>
<td>owe/boo boo</td>
</tr>
<tr>
<td>belly button</td>
<td>hair</td>
<td>penis*</td>
</tr>
<tr>
<td>buttocks/bottom*</td>
<td>hand</td>
<td>shoulder</td>
</tr>
<tr>
<td>cheek</td>
<td>head</td>
<td>tooth</td>
</tr>
<tr>
<td>chin</td>
<td>knee</td>
<td>toe</td>
</tr>
<tr>
<td>ear</td>
<td>leg</td>
<td>tongue</td>
</tr>
<tr>
<td>eye</td>
<td>lips</td>
<td>tummy</td>
</tr>
<tr>
<td>face</td>
<td>mouth</td>
<td>vagina*</td>
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</tbody>
</table>

4.8 **SMALL HOUSEHOLD ITEMS**

<table>
<thead>
<tr>
<th>basket</th>
<th>glasses</th>
<th>plate</th>
</tr>
</thead>
<tbody>
<tr>
<td>blanket</td>
<td>hammer</td>
<td>purse</td>
</tr>
<tr>
<td>bottle</td>
<td>jar</td>
<td>radio</td>
</tr>
<tr>
<td>box</td>
<td>keys</td>
<td>sissors</td>
</tr>
<tr>
<td>bowl</td>
<td>knife</td>
<td>soap</td>
</tr>
<tr>
<td>broom</td>
<td>lamp</td>
<td>spoon</td>
</tr>
<tr>
<td>brush</td>
<td>light</td>
<td>tape</td>
</tr>
<tr>
<td>bucket</td>
<td>medicine</td>
<td>telephone</td>
</tr>
<tr>
<td>camera</td>
<td>money</td>
<td>tissue/tissues</td>
</tr>
<tr>
<td>can</td>
<td>mop</td>
<td>toothbrush</td>
</tr>
<tr>
<td>clock</td>
<td>nail</td>
<td>towel</td>
</tr>
<tr>
<td>comb</td>
<td>serviettes</td>
<td>rubbish</td>
</tr>
<tr>
<td>cup</td>
<td>paper</td>
<td>tray</td>
</tr>
<tr>
<td>dish</td>
<td>penny</td>
<td>vacuum</td>
</tr>
<tr>
<td>fork</td>
<td>picture</td>
<td>walker</td>
</tr>
<tr>
<td>garbage</td>
<td>pillow</td>
<td>watch</td>
</tr>
<tr>
<td>glass</td>
<td></td>
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</tbody>
</table>

4.9 **FURNITURE AND ROOMS**

<table>
<thead>
<tr>
<th>basement</th>
<th>drawer</th>
<th>rocking chair</th>
</tr>
</thead>
<tbody>
<tr>
<td>bathroom</td>
<td>dryer</td>
<td>room</td>
</tr>
<tr>
<td>bathtub</td>
<td>garage</td>
<td>shower</td>
</tr>
<tr>
<td>bed</td>
<td>high chair</td>
<td>sink</td>
</tr>
<tr>
<td>bedroom</td>
<td>kitchen</td>
<td>sofa</td>
</tr>
<tr>
<td>chest</td>
<td>living room</td>
<td>stairs</td>
</tr>
<tr>
<td>chair</td>
<td>oven</td>
<td>stove</td>
</tr>
<tr>
<td>cupboard</td>
<td>play pen</td>
<td>table</td>
</tr>
<tr>
<td>couch</td>
<td>porch</td>
<td>TV</td>
</tr>
<tr>
<td>cot</td>
<td>potty</td>
<td>washing machine</td>
</tr>
<tr>
<td>door</td>
<td>refrigerator</td>
<td>window</td>
</tr>
</tbody>
</table>

*VOC量表问卷*  
*(Early Language Questionnaire)*  
*HREC 31531_V6 23/08/2015*  
*Page 8 of 15*
### 4.10 OUTSIDE THINGS

- backyard
- cloud
- flag
- flower
- garden
- grass
- hose
- ladder
- lawn mower
- moon
- pool
- rain
- rock
- roof
- sandbox
- shovel
- footprint
- sky
- slide
- snow
- sprinkler
- star
- stick
- stone
- street
- sun
- swing
- tree
- water
- wind

### 4.11 PLACES TO GO * or words used in your family

- beach
- camping
- church*
- circus
- country
- city
- farm
- petrol station
- home
- house
- movie
- outside
- party
- picnic
- playground
- school
- shop
- woods
- work
- yard
- zoo

### 4.12 PEOPLE * or words used in your family

- aunt
- baby
- babysitter
- babysitter's name
- boy
- brother
- child
- clown
- cowboy
- daddy *
- docaort
- fireman
- friend
- girl
- grandpa*
- lady
- postman
- man
- mummy*
- nurse
- own name
- people
- person
- pet's name
- sister
- teacher
- uncle

### 4.13 GAMEs AND ROUTINES

- bath
- breakfast
- bye
- ring (on phone)
- dinner
- give me five!
- got you!
- go potty
- hi
- hello
- lunch
- nap
- night
- no
- please
- thanks
- shopping
- snack
- so big!
- thank you
- this little piggy
- turn around
- yes
- za z za z shush/shush

### 4.14 ACTION WORDS

- bite
- blow
- break
- bring
- build
- drive
- drop
- dry
- dump
- eat
- hug
- hurry
- jump
- kick
- kiss
- read
- ride
- rip
- run
- say
- swim
- swing
- take
- talk

---

McChild questionnaire
(Early Language Questionnaire)

Page 9 of 15  HREC 3185_V1  23/06/2015
<table>
<thead>
<tr>
<th>bump</th>
<th>fall</th>
<th>knock</th>
<th>see</th>
<th>fear</th>
</tr>
</thead>
<tbody>
<tr>
<td>buy</td>
<td>feed</td>
<td>lick</td>
<td>shake</td>
<td>think</td>
</tr>
<tr>
<td>carry</td>
<td>find</td>
<td>like</td>
<td>share</td>
<td>throw</td>
</tr>
<tr>
<td>catch</td>
<td>finish</td>
<td>listen</td>
<td>show</td>
<td>tickle</td>
</tr>
<tr>
<td>chase</td>
<td>fit</td>
<td>look</td>
<td>sing</td>
<td>touch</td>
</tr>
<tr>
<td>clap</td>
<td>fox</td>
<td>love</td>
<td>sit</td>
<td>wait</td>
</tr>
<tr>
<td>clean</td>
<td>get</td>
<td>make</td>
<td>skate</td>
<td>walk</td>
</tr>
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<td>climb</td>
<td>fire</td>
<td>open</td>
<td>sleep</td>
<td>walk</td>
</tr>
<tr>
<td>close</td>
<td>go</td>
<td>paint</td>
<td>slide</td>
<td>wash</td>
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<td>hate</td>
<td>pick</td>
<td>smile</td>
<td>watch</td>
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<tr>
<td>cover</td>
<td>have</td>
<td>play</td>
<td>spill</td>
<td>wipes</td>
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<td>cry</td>
<td>hear</td>
<td>pour</td>
<td>splash</td>
<td>wish</td>
</tr>
<tr>
<td>cut</td>
<td>help</td>
<td>pretend</td>
<td>stand</td>
<td>work</td>
</tr>
<tr>
<td>dance</td>
<td>hide</td>
<td>pull</td>
<td>stop</td>
<td>write</td>
</tr>
<tr>
<td>draw</td>
<td>hit</td>
<td>push</td>
<td>sweep</td>
<td></td>
</tr>
<tr>
<td>drink</td>
<td>hold</td>
<td>put</td>
<td></td>
<td></td>
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### 4.15 DESCRIPTIVE WORDS

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<th>brown</th>
<th>careful</th>
<th>clean</th>
<th>cold</th>
<th>cute</th>
<th>dark</th>
<th>dirty</th>
<th>dry</th>
<th>empty</th>
<th>fast</th>
<th>fine</th>
<th>first</th>
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<tr>
<td>full</td>
<td>green</td>
<td>hungry</td>
<td>hurt</td>
<td>last</td>
<td>little</td>
<td>long</td>
<td>loud</td>
<td>mad</td>
<td>naughty</td>
<td>new</td>
<td>nice</td>
<td>noisy</td>
<td>old</td>
</tr>
<tr>
<td>orange</td>
<td>heavy</td>
<td>sleepy</td>
<td>soft</td>
<td>story</td>
<td>stuck</td>
<td>thirsty</td>
<td>tiny</td>
<td>tired</td>
<td>wet</td>
<td>white</td>
<td>windy</td>
<td>yellow</td>
<td>lucky</td>
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### 4.16 WORDS ABOUT TIME

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<th>before</th>
<th>day</th>
<th>later</th>
</tr>
</thead>
<tbody>
<tr>
<td>morning</td>
<td>right</td>
<td>now</td>
<td>time</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>today</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>tomorrow</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>tonight</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>yesterday</td>
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### 4.17: Pronouns

<table>
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<tr>
<th>He</th>
<th>Me</th>
<th>Their</th>
<th>We</th>
</tr>
</thead>
<tbody>
<tr>
<td>Her</td>
<td>Mine</td>
<td>Them</td>
<td>You</td>
</tr>
<tr>
<td>Hers</td>
<td>My</td>
<td>These</td>
<td>Your</td>
</tr>
<tr>
<td>Him</td>
<td>Myself</td>
<td>They</td>
<td>Yourself</td>
</tr>
<tr>
<td>His</td>
<td>Our</td>
<td>This</td>
<td></td>
</tr>
<tr>
<td>I</td>
<td>She</td>
<td>Those</td>
<td></td>
</tr>
<tr>
<td>It</td>
<td>That</td>
<td>Us</td>
<td></td>
</tr>
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### 4.18: Question Words

<table>
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<th>How</th>
<th>When</th>
<th>Which</th>
<th>Why</th>
</tr>
</thead>
<tbody>
<tr>
<td>What</td>
<td>Where</td>
<td>Who</td>
<td></td>
</tr>
</tbody>
</table>

### 4.19: Prepositions and Location

<table>
<thead>
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<th>About</th>
<th>Down</th>
<th>On top of</th>
</tr>
</thead>
<tbody>
<tr>
<td>Above</td>
<td>For</td>
<td>Out</td>
</tr>
<tr>
<td>Around</td>
<td>Here</td>
<td>Over</td>
</tr>
<tr>
<td>At</td>
<td>Inside</td>
<td>In there</td>
</tr>
<tr>
<td>Away</td>
<td>Into</td>
<td>To</td>
</tr>
<tr>
<td>Back</td>
<td>Next to</td>
<td>Under</td>
</tr>
<tr>
<td>Behind</td>
<td>Of</td>
<td>Up</td>
</tr>
<tr>
<td>Beside</td>
<td>Off</td>
<td>With</td>
</tr>
<tr>
<td>By</td>
<td>On</td>
<td></td>
</tr>
</tbody>
</table>

### 4.20: Quantifiers and Articles

<table>
<thead>
<tr>
<th>A</th>
<th>Each</th>
<th>Other</th>
</tr>
</thead>
<tbody>
<tr>
<td>All</td>
<td>Every</td>
<td>Some</td>
</tr>
<tr>
<td>A lot</td>
<td>More</td>
<td>Some</td>
</tr>
<tr>
<td>An</td>
<td>Much</td>
<td>The</td>
</tr>
<tr>
<td>Another</td>
<td>Not</td>
<td>Too</td>
</tr>
<tr>
<td>Any</td>
<td>None</td>
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</tr>
</tbody>
</table>

### 4.21: Helping Verbs

<table>
<thead>
<tr>
<th>Am</th>
<th>Does</th>
<th>Need/need to</th>
</tr>
</thead>
<tbody>
<tr>
<td>Are</td>
<td>Don't</td>
<td>Try/try to</td>
</tr>
<tr>
<td>Be</td>
<td>Gonna/going to</td>
<td>Wanna/want to</td>
</tr>
<tr>
<td>Can</td>
<td>Could</td>
<td>Got to</td>
</tr>
<tr>
<td>Could</td>
<td>Have to</td>
<td>Were</td>
</tr>
<tr>
<td>Did/didn't</td>
<td>Is</td>
<td>Will</td>
</tr>
<tr>
<td>Do</td>
<td>Don't want</td>
<td>Would</td>
</tr>
</tbody>
</table>

### 4.22: Connecting Words (6)

<table>
<thead>
<tr>
<th>And</th>
<th>But</th>
<th>So</th>
</tr>
</thead>
<tbody>
<tr>
<td>Because</td>
<td>If</td>
<td>Then</td>
</tr>
</tbody>
</table>
### 4.25 HOW CHILDREN USE WORDS

<table>
<thead>
<tr>
<th></th>
<th>Not Yet</th>
<th>Sometimes</th>
<th>Often</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. Does your child ever talk about past events or people who are not present? For example, a child who saw a parade last week might later say parade, clown, or band.</td>
<td>☐</td>
<td>☐</td>
<td>☑</td>
</tr>
<tr>
<td>2. Does your child ever talk about something that’s going to happen in the future, for example, saying “choc choc” or “airplane” before you leave the house for a trip, or saying “swing” when you are going to the park?</td>
<td>☐</td>
<td>☐</td>
<td>☑</td>
</tr>
<tr>
<td>3. Does your child talk about objects that are not present such as asking about a missing or absent toy, referring to a pet out of view, or asking about someone not present?</td>
<td>☐</td>
<td>☐</td>
<td>☑</td>
</tr>
<tr>
<td>4. Does your child understand if you ask for something that is not in the room, for example, by going to the bedroom to get a teddy bear when you say “where’s the bear?”</td>
<td>☐</td>
<td>☐</td>
<td>☑</td>
</tr>
<tr>
<td>5. Does your child ever pick up or point to an object and name an absent person to whom the object belongs? For example, a child might point to mummy’s shoe and say “mummy”.</td>
<td>☐</td>
<td>☐</td>
<td>☑</td>
</tr>
</tbody>
</table>

### 4.24 WORD ENDINGS / PART 1

<table>
<thead>
<tr>
<th></th>
<th>Not Yet</th>
<th>Sometimes</th>
<th>Often</th>
</tr>
</thead>
<tbody>
<tr>
<td>1. To talk about more than one thing, we add an “s” to many words. Examples include cards (for more than one card), shoes, dogs, and keys. Has your child begun to do this?</td>
<td>☐</td>
<td>☐</td>
<td>☑</td>
</tr>
<tr>
<td>2. To talk about ownership, we add an “s”, for example, Daddy’s key, kitty’s dish, and baby’s bottle. Has your child begun to do this?</td>
<td>☐</td>
<td>☐</td>
<td>☑</td>
</tr>
<tr>
<td>3. To talk about activities, we sometimes add “ed” to verbs. Examples include running and crying. Has your child begun to do this?</td>
<td>☐</td>
<td>☐</td>
<td>☑</td>
</tr>
<tr>
<td>4. To talk about things that happened in the past, we often add “ed” to the verb. Examples include kissed, opened, and pushed. Has your child begun to do this?</td>
<td>☐</td>
<td>☐</td>
<td>☑</td>
</tr>
</tbody>
</table>

### WORD FORMS
Following are some other words children learn. Please mark any of these words that your child uses.

#### 4.25 NOUNS

<p>| | | | |</p>
<table>
<thead>
<tr>
<th></th>
<th></th>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>children</td>
<td>men</td>
<td>teeth</td>
<td>mice</td>
</tr>
</tbody>
</table>

#### 4.26 VERBS

<p>| | | | |</p>
<table>
<thead>
<tr>
<th></th>
<th></th>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td>ate</td>
<td>fell</td>
<td>made</td>
<td>saw</td>
</tr>
<tr>
<td>blew</td>
<td>flew</td>
<td>ren</td>
<td>sat</td>
</tr>
<tr>
<td>bought</td>
<td>got</td>
<td>sat</td>
<td>took</td>
</tr>
<tr>
<td>broke</td>
<td>had</td>
<td>sang</td>
<td>took</td>
</tr>
<tr>
<td>came</td>
<td>heard</td>
<td>took</td>
<td>took</td>
</tr>
<tr>
<td>drank</td>
<td>held</td>
<td>went</td>
<td>took</td>
</tr>
<tr>
<td>drove</td>
<td>lost</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
WORD ENDINGS / PART 2

Young children often place the wrong endings on words. For example, a child might say "Aunte goed home." Mistakes like this are often a sign of progress in language. In the following lists, please mark all the mistakes of this kind that you have heard your child say recently.

### 4.27: NOUNS

<table>
<thead>
<tr>
<th>Blanket</th>
<th>Pants</th>
<th>Sock</th>
<th>Toes</th>
</tr>
</thead>
<tbody>
<tr>
<td>children</td>
<td>men</td>
<td>men</td>
<td>tees</td>
</tr>
<tr>
<td>child</td>
<td>mice</td>
<td>mice</td>
<td>toses</td>
</tr>
<tr>
<td>fooots</td>
<td>mouse</td>
<td>mouse</td>
<td>toouths</td>
</tr>
<tr>
<td>fooots</td>
<td>shoes</td>
<td>shoes</td>
<td></td>
</tr>
</tbody>
</table>

### 4.28: VERBS

<table>
<thead>
<tr>
<th>Stole</th>
<th>Some</th>
<th>good</th>
<th>ruined</th>
</tr>
</thead>
<tbody>
<tr>
<td>blewed</td>
<td>deed</td>
<td>goped</td>
<td>runned</td>
</tr>
<tr>
<td>blowed</td>
<td>dranked</td>
<td>haveed</td>
<td>sweeted</td>
</tr>
<tr>
<td>bought</td>
<td>eated</td>
<td>holded</td>
<td>sidde</td>
</tr>
<tr>
<td>brokeed</td>
<td>failed</td>
<td>losted</td>
<td>takeed</td>
</tr>
<tr>
<td>brokeed</td>
<td>flaced</td>
<td>losted</td>
<td>wanted</td>
</tr>
<tr>
<td>creamed</td>
<td>gapped</td>
<td>moosed</td>
<td></td>
</tr>
</tbody>
</table>

### 4.29: Has your child begun to combine words yet, such as “nother cracker” or “doggie bite?”

<table>
<thead>
<tr>
<th>Not Yet</th>
<th>Sometimes</th>
<th>Often</th>
</tr>
</thead>
</table>

### 4.30: COMPLEXITY

In each of the following pairs, please mark the one that sounds MOST like the way your child talks right now.

If your child is saying sentences even longer or more complicated than the two provided, just pick the second one.

1. Two shoe, Two shoes.
2. Two foot, Two feat.
3. Daddy car, Daddy’s car.
4. (Talking about something happening right now), Kitty sleep, Kitty sleeping.
5. (Talking about something happening right now), I make tower, I making tower.
6. (Talking about something that already happened), I fall down, I fall down.
7. More biscuit, More biscuits.
8. These my tooth, These my teeth.
10. (Talking about something that already happened), Doggie kiss me, Doggie kissed me.
11. (Talking about something that already happened), Daddy pick me up, Daddy picked me up.
12. (Talking about something that already happened), Kitty go away, Kitty went away.
13. Doggie table, Doggie on table.
14. That my truck, That’s my truck.
15. Baby crying, Baby is crying.
16. You fix it, Can you fix it?
17. Read me story, Mummy, Read me a story, Mummy.
18. No wash dolly, Don’t wash dolly.
4.31 Thanks for telling us about your child’s language in Section 4 (above). Early Intervention services tell us that they too would value this information on your child’s current language.

Do you give permission for VicCHILD to share Section 4 with your Early Intervention service?

☐ Yes  ☐ No, I don’t want VicCHILD to share section 4  ☐ No, doesn’t attend

4.32 If we lose touch with you, could we ask your current Early Intervention service if they know which service/school your child moved on to? We would ask them to contact you on our behalf, or provide your current contact details so we can get in touch directly.

☐ Yes  ☐ No, I don’t want VicCHILD to ask this service  ☐ No, doesn’t attend

5. About YOU and your FAMILY LIFE

We know that a child’s environment and family plays a big part in their development. We would now like to ask some questions about you, as the parent or carer of this child.

5.1 Are you male or female? (Fill one circle only)

☐ Female  ☐ Male

5.2 Which of the following best describes YOUR relationship to this child? (Fill one circle only)

☐ Biological parent  ☐ Step parent  ☐ Grandparent  ☐ Other (please specify)

5.3 In general, how would you say YOUR current health is? (Fill one circle only)

☐ Excellent  ☐ Very good  ☐ Good  ☐ Fair  ☐ Poor
5.4 Do you currently have a partner who lives with you?

- Yes Please go to question 5.5
- No Please go to Section 6

5.5 Is your PARTNER male or female? (Fill one circle only)

- Female
- Male

5.6 Which of the following best describes your PARTNER’S relationship to this child? (Fill one circle only)

- Biological parent
- Step parent
- Grandparent
- Other (please specify) __________________________

5.7 In general, how would you say your PARTNER’S current health is? (Fill one circle only)

- Excellent
- Very good
- Good
- Fair
- Poor

6. Contact details for you and your nearest relative or friend

We’d like to stay in touch as your child grows up - your current contact details will help.

Address ____________________________ Suburb ____________________________
Postcode __________ State ____________ Home Phone ____________________________
Your Email __________________________ Your Mobile ____________________________
Partner’s contact details:
Partner’s name __________________________
Partner’s Email __________________________ Partner’s Mobile ____________________________

In case you move house, it can be helpful to contact a relative or friend who knows where you’ve moved. If you can, please write your relative’s friend’s details below.

Name of relative or friend __________________________
Relationship to you (e.g., parent, friend, sister etc.) __________________________
Address ____________________________ Suburb ____________________________
Postcode __________ State ____________ Home Phone ____________________________
Email __________________________ Mobile ____________________________

Thank you for your time!
Please check that you’ve answered all of the questions, and return your questionnaires in the reply paid envelope.

Questions? Contact VicCHILD on (03) 9345 4215 / vic-child@rch.org.au
OR visit http://www.rch.org.au/ccch/fbr_researchers/VicCHILD/
## Appendix H: Ethics certificates

**RCH HUMAN RESEARCH ETHICS COMMITTEE APPROVAL**

<table>
<thead>
<tr>
<th>HREC REF. No.</th>
<th>31081 O</th>
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<tbody>
<tr>
<td>PROJECT TITLE:</td>
<td>Victorian Childhood Hearing Impairment Longitudinal Databank (VioCHILD)</td>
</tr>
<tr>
<td>DOCUMENTS APPROVED:</td>
<td></td>
</tr>
<tr>
<td>Assessment Cover Letter CL-V-4 v1 dated 30 Jul 2014</td>
<td></td>
</tr>
<tr>
<td>Assessment Cover Letter CL-V-5 Follow up v1 dated 30 Jul 2014</td>
<td></td>
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<tr>
<td>Feedback Letter (not in range) v1 dated 7 Aug 2014</td>
<td></td>
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<tr>
<td>Feedback Letter (within range) v1 dated 7 Aug 2014</td>
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<td>Questionnaire (5-7 year) v5 dated 19 Aug 2014</td>
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<td>Questionnaire (13-24 months) v4 dated 19 Aug 2014</td>
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<td>APPROVED PROTOCOL:</td>
<td>Protocol v12 dated 19 August 2014</td>
</tr>
<tr>
<td>PRINCIPAL INVESTIGATOR:</td>
<td>Melissa Wake</td>
</tr>
<tr>
<td>DATE OF MODIFICATION APPROVAL:</td>
<td>30 September 2014</td>
</tr>
<tr>
<td>DURATION:</td>
<td>23 months</td>
</tr>
<tr>
<td>DATE OF APPROVAL EXPIRY:</td>
<td>17 August 2016</td>
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</tbody>
</table>

CONDITIONS: Approved for access to participants in ethically approved research.

**SIGNED:**

---

**COMMITTEE REPRESENTATIVE**

---

**APPROVED SUBJECT TO THE FOLLOWING CONDITIONS:**

1. The study must not commence until all Research Agreements have been executed (if applicable).
3. Any proposed change in the protocol or approved documents or the addition of documents must be submitted to the Human Research Ethics Committee (HREC) for approval prior to implementation, including:
   - Flyer, brochure, advertising material
   - Increase in recruitment target
4. The Principal investigator must notify Research Development & Ethics of:
   - Any serious adverse events of the study on participants and steps taken to deal with them.
   - Any unforeseen events (e.g. protocol violations or complaints).
   - Investigators withdrawing from or joining the project.
5. A progress report must be submitted annually and at the conclusion of the project.
6. RCH HREC approval must remain current for the entire duration of the project. If the project is not completed in the allocated time a renewal request must be submitted to the Research Development & Ethics. Investigators
**RCH HUMAN RESEARCH ETHICS COMMITTEE APPROVAL**

<table>
<thead>
<tr>
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**SIGNED:** [Signature]

**COMMENTS:** This approval also reflects a change to the research team

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**APPROVED SUBJECT TO THE FOLLOWING CONDITIONS:**

1. The study must not commence until all Research Agreements have been executed (if applicable).
3. Any proposed change to the protocol or approved documents or the addition of documents must be submitted to the Human Research Ethics Committee (HREC) for approval prior to implementation, including:
   - Flyers, brochures, advertising material
   - Increase in recruitment target
4. The Principal Investigator must notify Research Ethics & Governance of:
   - Any serious adverse effects of the study on participants and steps taken to deal with them.
   - Any unforeseen events (e.g. protocol violations or complaints).
   - Investigators withdrawing from or joining the project.
5. A progress report must be submitted annually and at the conclusion of the project.
6. RCH HREC approval must remain current for the entire duration of the project. If the project is not completed in the allocated time, a renewal request must be submitted to the Research Ethics & Governance. Investigators undertaking projects without current HREC approval risk their indemnity, funding and publication rights.

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**CLINICAL TRIALS**
Science, Not Philosophy, Will Help Deaf and Hard-of-Hearing Children Reach Their Potential

Melissa Wake, MD;^1,^2 Peter Darow, MAd)^3

Survival, developmental outcomes, and quality of life have been transformed over the past 30 years for infants suffering a wide range of previously devastating conditions. Children affected by prematurity, cystic fibrosis, and congenital heart disease now survive and indeed thrive into adulthood. Technical advances have proceeded alongside coordinated programs of multicenter registries and trials, driven by clinicians and researchers motivated to not only continually push the boundaries of postdiagnosis treatment but to show that children reap the benefits.

The past 30 years have also heralded astonishing advances for children born deaf and hard of hearing. The first pediatric cochlear implantation in 1985 was followed by the discovery of the first of many genes for nonsyndromic hearing impairment^1^ in the 1990s. Widespread implementation of universal newborn hearing screening programs in the early 2000s means that hearing impairment is now routinely diagnosed in the first 3 postnatal months. For the first time in history, we have the opportunity to learn how to prevent language delay, so that development never falls behind that of hearing children.

The systematic review by Fitzpatrick et al^3^ uncomfortably illustrates that we may not yet have seized this opportunity. Noting that "no consensus exists on optimal interventions for spoken language development," they examined the effectiveness of early sign and oral language intervention compared with oral language intervention only for children with permanent hearing loss. Only 11 observational studies could be included. Not one randomized trial was located; indeed, not a single article reported any kind of experimental design. Eight of the 11 articles focused solely on cochlear implant users, where the large majority of children use standard hearing aids. The total end result is 320 children with implants and a further 207 in samples heterogeneous for hearing aids and implants. For every language, speech, and speech reception outcome, the quality of evidence was rated as "very low."

The need for better intervention is real and present. So many questions remain unanswered. Is there a hearing threshold below which amplification has few benefits? Do children with unilateral losses benefit from hearing aids? How is language best learned in the presence of hearing that is not normal, even with the best modern technology? How is best taught? What are the most effective ways for parents to promote communication? Does sign language help, hinder, or make no difference to oral language outcomes? Even the simplest question of "Are we making progress, year on year?" remains out of reach.

Given these questions, what underlies the dearth of high-quality randomized trials? Perhaps the advocacy for universal newborn hearing screening^4^ and cochlear implantation were just

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too persuasive, our job seemed done! Unfortunately, it is becoming very clear from large-scale epidemiologic studies that, on average, language and learning outcomes still fall well below both population norms and the children’s own cognitive potential. The Australian Statewide Comparison of Outcomes of Hearing Loss quasi-experimental study reports incremental gains of ~0.3 to 0.5 SDs in language scores with each step from statewide opportunities to risk factor to universal screening. Nonetheless, even in the latter group, mean language scores for young schoolchildren born in 2003 to 2005 without intellectual disability still hovered just below 90 (~1 SD below their cognitive ability). These figures nearly mirror those of the observational Longitudinal Outcomes of Children with Hearing Impairments (following ~400 Australian children across the full range of hearing loss) and Outcomes of Children with Hearing Loss (following ~300 hard-of-hearing children from multiple US states) studies.

Whatever the reason, the lack of intervention trials is deeply troubling. Contrast this with childhood cancer, whose cumulative childhood incidence of ~1.0 to 2.5 per thousand is similar to that of congenital hearing loss. Acute lymphoblastic leukemia was invariably fatal in the 1950s; now 90% of children survive. Today, one might argue that hearing impairment is the more disabling condition. Like cancer, gains for childhood hearing impairment will happen only with painstaking comparative trials that are powered for incremental benefit and rigorous in avoiding confounding.

As noted by Fitzpatrick et al., the debate about optimal outcomes has resulted in a plethora of philosophy-driven interventions. Winston Churchill said that “However beautiful the strategy, you should occasionally look at the results.” A coordinated international investment is needed to support and incentivize randomized trials, ideally springing from harmonized, web-based registries that can both monitor secular year-on-year improvements and provide large numbers of parents eager to make tomorrow better than today. Science, not philosophy, will optimize the future for countless unborn children with permanent hearing impairment.

REFERENCES
Appendix J: Epidemiology of hearing loss paper

Cross-sectional epidemiology of hearing loss in Australian children aged 11–12 years old and 25-year secular trends

Jing Wang,1,2 Carlijn M P le Clercq,1,3 Valerie Sung,1,2 Peter Carey,1,4 Richard S Liu,1,2 Fiona K Mensah,1,2 Rachael A Burn,1,2 Lisa Gold,1,2 Melissa Woke1,2

ABSTRACT

Objective: In a national study of Australian children aged 11–17 years old, we examined the (1) prevalence and characteristics of hearing loss, (2) its demographic risk factors, and (3) evidence for secular increases since 1990.

Methods: This is a cross-sectional CheckPoint wave within the Longitudinal Study of Australian Children (LSAC) cohort. 1465 children (49.8% retention; 49.7% boys) underwent air conduction audiometry, a new technology that allow for rapid and accurate hearing assessment. Results were recorded for 10 hearing thresholds at 500, 1000, 2000, and 4000 Hz.

Results: The prevalence of bilateral and unilateral hearing loss ≥60 dB HL was 9.3% and 13.3%, respectively. Bilateral hearing loss (BH) was more prevalent than unilateral hearing loss (UHL) (6.8% vs 2.5%). Bilateral UHL was more prevalent than unilateral BH (12.5% vs 9.6%). The prevalence trend was similar across age groups, with the highest prevalence in the 11–12-year-old age group (18.0%).

Conclusions: The prevalence of hearing loss in children is a major public health concern. Early identification and intervention are critical to prevent the development of hearing loss and its associated complications. The results of this study provide valuable information for the development of evidence-based hearing loss prevention strategies.
Original article

earphones in 2005–2006 compared with 19.8% in 1998–1994, and reported personal stereo usage has been associated with a 70% increased risk of slight to mild sensorineural hearing loss. Second, factors related to the more general rise in non-communicable diseases could also contribute, including obesity, cardiovascular disease, diabetes, hypertension and dyslipidaemia. Consistently increasing levels of inflammation are a putative risk factor for presbycusis. Childhood obesity, which has risen dramatically over the last three decades, has been cross-sectionally associated with higher hearing thresholds and an almost doubling risk in low-frequency hearing loss. Third, social disparities—increasing in many societies—have been associated with noise-induced hearing threshold shift in children of lower socioeconomic status. While gender balance is stable, sex should be considered because of its possible interactions with other risk factors.

The Longitudinal Study of Australian Children’s (LSAC) recent cross-sectional CheckPoint biophysical module provides an opportunity to study current epidemiology in hearing loss at age 11–12 years. Here, we (1) describe the current prevalence and characteristics of hearing loss; (2) quantify its demographic risk factors, and (3) examine secular trends, drawing on international studies published in 1996–2015.

METHODS

Study design and participants

The Child Health CheckPoint is a cross-sectional population-derived study nested within the national LSAC. LSAC recruited two nationally representative cohorts in 2004, of which the B cohort (5107 infants) is relevant to this paper. In a two-stage sampling design, 10% of all Australian postcodes were randomly selected, stratified by state and urban/rural domicile; in-children were then randomly selected from the Australian Medicare database and followed biennially. The B cohort response rate was 57.2% (2004), of whom 75% were retained to wave 6 (2014). LSAC interviewers obtained consent at the wave 6 interview (10–11 years) to pass contact details to CheckPoint, which was open to all retained B cohort children aged 11–12 years between LSAC’s wave 6 (2014) and wave 7 (2016). Parents provided written consent and children verbal assent.

Procedures

From December 2014, CheckPoint contacted each family to ascertain interest and book a single appointment for the child, as the CheckPoint assessment centre visited each Australian state sequentially between February 2015 and February 2016. Each child underwent assessments of multiple body systems; we report on the ‘Listen Up’ station, which was offered in the first 30–45 min of either a 3.5-hour appointment at the main assessment centre (17 stations) or a 2.5-hour appointment at the ‘mini-centre’ (13 stations) in smaller regional cities.

Measures

Audiometry

Trained examiners conducted air-conduction pure-tone audiometry using an Oscilla USB-330 (V3.3.4) computer-based audiometer with Oscilla headphones, and a standardised modified Hughson-Westlake audiometric technique. Testing of the first frequency began at 30 dB HL if within normal limits, testing for other frequencies began at 20 dB HL. Participants were asked to remove hearing aids and/or cochlear implant speech processors if worn; testing of the first frequency began at 60 dB HL and of successive frequencies at 50 dB HL. shown that adjacent frequencies threshold. If participants’ hearing thresholds at two or more frequencies in at least one ear were >20 dB HL, parents’ written feedback stated that hearing was outside the usual range and they should consider a clinical audiologic assessment. For the first 143 participants, only three frequencies were tested (1, 2, and 4 kHz) for each ear across an intensity range of 10 to 120 dB HL. As CheckPoint systems became faster and additional funding was sourced, testing at 8 kHz (n=1342), tympanometry (n=1090) and soundproof booths (n=930) were successively added.

Tympanometry (middle ear function)
The Oscilla TSM500 automatically calculated ear canal volume, middle ear pressure and compliance during a pressure sweep.

Table 1: Summary of derived variables

<table>
<thead>
<tr>
<th>Construct/measure</th>
<th>Definition</th>
<th>Rationale</th>
</tr>
</thead>
<tbody>
<tr>
<td>Continuous</td>
<td></td>
<td></td>
</tr>
<tr>
<td>High Fletcher Index</td>
<td>The average of thresholds at 1, 2 and 4 kHz</td>
<td>Best reflects audibility at speech frequencies</td>
</tr>
<tr>
<td>Four frequency average</td>
<td>The average of thresholds at 1, 2, 4 and 8 kHz</td>
<td>Reflects audibility at usual speech-speech frequencies</td>
</tr>
<tr>
<td>Lower frequency average</td>
<td>The average of thresholds at 1 and 2 kHz</td>
<td>Reflects audibility of lower frequency speech sounds, for example, most vowels and letter combinations (th, ed, etc); characteristic of condutive or sensorineural hearing loss</td>
</tr>
<tr>
<td>Higher frequency average</td>
<td>The average of thresholds at 4 and 8 kHz</td>
<td>Reflects audibility of higher frequency speech sounds, for example, most consonant and letter combinations (th, etc); particularly disabling with background noise; suggests noise-induced and/or sensorineural hearing loss</td>
</tr>
<tr>
<td>Categorical</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Unilateral hearing loss</td>
<td>Hearing thresholds ≥15 dB HL in worse hearing ear and hearing thresholds ≤15 dB HL in better hearing ear</td>
<td>Hearing loss in one ear</td>
</tr>
<tr>
<td>Bilateral hearing loss</td>
<td>Hearing thresholds ≥15 dB HL in better hearing ear and hearing thresholds ≤15 dB HL in better hearing ear</td>
<td>Hearing loss in both ears</td>
</tr>
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<td>Type A tympanogram</td>
<td>Ear canal volume: 0.6–1.5 cm3; middle ear pressure: −100 to +50 daPa; compliance: 0.25–1.8 mmHg</td>
<td>Suggests normal middle ear function</td>
</tr>
<tr>
<td>Type B tympanogram</td>
<td>Ear canal volume: 0.6–1.5 cm3; middle ear pressure: −100 to +50 daPa; compliance: &lt;0.25 mmHg</td>
<td>Suggests middle ear effusion, tympanic membrane perforation, common effusion or a probe sealed against the canal wall</td>
</tr>
<tr>
<td>Type C tympanogram</td>
<td>Ear canal volume: 0.6–1.5 cm3; middle ear pressure: &lt;−100 daPa; compliance: 0.25–1.8 mmHg</td>
<td>Suggests negative middle ear pressure</td>
</tr>
</tbody>
</table>

db HL, decibel hearing level.


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Figure 1  Audiometric recruitment and participation flow in Child Health CheckPoint.

Tympograms were classified as types A (normal compliance), B (no or negligible compliance) and C (normal compliance, negative middle ear pressure), with criteria and interpretation detailed in table 1.11

Sociodemographic exposures
CheckPoint recorded children's age, sex and Socio-Economic Indexes for Areas disadvantage index. This composite neighbourhood index ranks postcodes nationally according to data from the 2011 five-yearly Australian Census. Contributing items include average household education levels, income levels, employment status and disability for that postcode. Higher scores reflect less disadvantage, with a national mean of 1000 and standard deviation (SD) of 100. Distribution of index scores is divided into five national quintiles.

Data management
Table 1 details all derived variables with their constructs and rationale. Our primary pure-tone average outcome was the high Fletcher Index (mean hearing threshold across 1, 2 and 4 kHz). As its range of frequencies maps most closely to the range of speech sound frequencies, it is likely to have the greatest functional relevance for oral communication.25 We also calculated the following indices to maximise cross-study comparability: four-frequency average (1, 2, 4 and 8 kHz), lower frequency average (1 and 2 kHz) and higher frequency average (4 and 8 kHz, believed to be most affected by noise exposure). We defined the following severity groupings: normal (−10 to 15 dB HL), slight (16–25 dB HL), mild (26–40 dB HL) and moderate or worse (≥41–60 dB HL) in line with the American Speech-Language-Hearing Association guidelines26 and other prevalence studies.14 We reported hearing abilities both by better and worse ear on high Fletcher Index. For all indices, we defined bilateral hearing loss as thresholds ≥16 dB HL in the better ear, and unilateral hearing loss as thresholds ≥16 dB HL in the worse ear but normal hearing (≤15 dB HL) in the better ear.14
Table 2: Prevalence of bilateral and unilateral hearing loss

<table>
<thead>
<tr>
<th>Variable</th>
<th>−10 to 15 dB normal</th>
<th>16−25 dB, slight</th>
<th>26−40 dB, mild</th>
<th>≥41 dB, moderate or worse</th>
<th>≥61 dB, slight or worse</th>
<th>≥26 dB, mild or worse</th>
</tr>
</thead>
<tbody>
<tr>
<td>Males</td>
<td>90.7 (88.1 to 92.1)</td>
<td>8.5 (2.0 to 10.1)</td>
<td>0.6 (0.3 to 1.2)</td>
<td>0.1 (0.3 to 0.6)</td>
<td>0.1 (0.3 to 0.6)</td>
<td>0.1 (0.3 to 0.6)</td>
</tr>
<tr>
<td>FEMA</td>
<td>92.7 (91.2 to 94.0)</td>
<td>6.6 (2.5 to 8.0)</td>
<td>0.7 (0.3 to 1.3)</td>
<td>0.1 (0.3 to 0.6)</td>
<td>0.0 (0.3 to 0.6)</td>
<td>0.0 (0.3 to 0.6)</td>
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<tr>
<td>FEMA*</td>
<td>89.0 (87.3 to 90.5)</td>
<td>10.0 (6.0 to 11.7)</td>
<td>0.9 (0.5 to 1.3)</td>
<td>0.1 (0.3 to 0.6)</td>
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<tr>
<td>FEMA†</td>
<td>91.1 (90.1 to 92.4)</td>
<td>6.0 (4.8 to 7.4)</td>
<td>0.6 (0.4 to 1.0)</td>
<td>0.1 (0.3 to 0.6)</td>
<td>0.0 (0.3 to 0.6)</td>
<td>0.0 (0.3 to 0.6)</td>
</tr>
</tbody>
</table>

Unilateral hearing loss

<table>
<thead>
<tr>
<th>Variable</th>
<th>−10 to 15 dB normal</th>
<th>16−25 dB, slight</th>
<th>26−40 dB, mild</th>
<th>≥41 dB, moderate or worse</th>
<th>≥61 dB, slight or worse</th>
<th>≥26 dB, mild or worse</th>
</tr>
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<tr>
<td>Males</td>
<td>77.4 (75.0 to 79.6)</td>
<td>12.3 (8.9 to 14.9)</td>
<td>0.7 (0.3 to 1.3)</td>
<td>0.2 (0.5 to 1.2)</td>
<td>0.1 (0.3 to 0.6)</td>
<td>0.1 (0.3 to 0.6)</td>
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<tr>
<td>FEMA</td>
<td>78.4 (76.1 to 80.5)</td>
<td>13.4 (10.7 to 16.1)</td>
<td>0.7 (0.3 to 1.3)</td>
<td>0.2 (0.5 to 1.2)</td>
<td>0.1 (0.3 to 0.6)</td>
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<tr>
<td>FEMA*</td>
<td>75.5 (73.9 to 76.9)</td>
<td>14.5 (12.8 to 16.4)</td>
<td>0.7 (0.3 to 1.3)</td>
<td>0.2 (0.5 to 1.2)</td>
<td>0.1 (0.3 to 0.6)</td>
<td>0.1 (0.3 to 0.6)</td>
</tr>
<tr>
<td>FEMA†</td>
<td>81.6 (80.4 to 82.6)</td>
<td>10.2 (8.7 to 11.7)</td>
<td>1.0 (0.6 to 1.6)</td>
<td>0.3 (0.5 to 1.8)</td>
<td>0.1 (0.3 to 0.6)</td>
<td>0.1 (0.3 to 0.6)</td>
</tr>
</tbody>
</table>

*Subsample only: children with audiometry at 8 kHz (n=1342).
†FEMA, four-frequency average; *FEMA, higher frequency average; †FEMA, high Fletcher index; FEMA, lower frequency average.

Statistical analyses

Statistical analyses were performed in Stata version 14.0. For aim 1, we calculated the prevalence estimates of hearing loss with 95% CIs. To determine whether it was justifiable to combine those with and without likely middle ear pathology for analyses, we used analysis of variance (ANOVA) to compare the mean hearing thresholds between those with type A, B, and C tympanograms. ANOVA was conducted for these comparisons despite a slight skew in the distribution of hearing thresholds, supported by application of the central limit theorem for a study of this size (n=1090 participants with tympanometry). For aim 2, we used logistic regression to estimate ORs with 95% CIs for hearing loss according to sociodemographic characteristics. For aim 3, we selected population studies from online supplementary table e1 that performed air-conduction audiology in children of comparable ages using similar hearing loss definitions. We then plotted four secular trend lines (representing four definitions) for studies reporting data collected since 1990, each summarizing published prevalence by midyear of data collection, with CheckPoint providing the final point. We report P value for trend using logistic regression.

RESULTS

Baseline characteristics

Figure 1 presents the study flow from wave 6 of LSAC onwards. The analyses included 1485 children. The mean age was 11.4 years with approximately equal numbers of boys and girls. The mean disadvantage index was 1026.2, indicating a slight skew towards less disadvantaged children compared with the general Australian population (mean 1000) (online supplementary table e2 and figure 1).

Audiometry and tympanometry

Online supplementary table e3 shows that the mean hearing thresholds for individual frequencies and all four indices were similar when including all children (n=1485), and the subsamples with audiometry at 8 kHz (n=1342) and with tympanometry (n=1090), with the lowest (best) for the highest frequency average and highest (worst) for the lower frequency average. The mean hearing thresholds for children with type A, B, and C tympanogram in better and worse ears differed significantly (all P<0.001), but these differences were small with high degrees of overlap of the distributions (online supplementary figure e1 and online supplementary table e3).

Prevalence of hearing loss (aim 1)

Table 2 shows the prevalence of bilateral and unilateral hearing loss using four indices. Regarding our primary outcome (high Fletcher Index), the prevalence of bilateral and unilateral hearing loss (≥16 dB HL) was 9.3% and 13.3%, respectively. Most hearing loss was slight (16−25 dB HL) (bilateral 8.5%, unilateral 12.3%). Using four frequency averages, the prevalence of bilateral and unilateral hearing loss was 7.3% and 14.3%, respectively. Lower frequency losses were more common than higher frequency losses (bilateral 11.0% vs 6.9%; unilateral 12.4% vs 11.5%) (table 2).

In sensitivity analyses, the prevalence of bilateral and unilateral hearing loss in children with type A tympanograms (n=956) (online supplementary figure e2) and children tested in soundproof booths (n=739) (online supplementary table e4) was similar to our overall prevalence estimates.

Sociodemographic risk factors (aim 2)

Girls appeared slightly more likely to have bilateral (OR=1.55, 95% CI 1.09 to 2.22, P=0.02) or unilateral (OR=1.38, 95% CI 1.02 to 1.86, P=0.04) losses using high Fletcher index. Bilateral/unilateral hearing losses were not significantly associated with age or disadvantage index (table 3). In sensitivity analyses, prevalence was similar by age, gender and disadvantage index in children with type A tympanograms (n=956) (online supplementary table e5).

Secular trends since 1990 (aim 3)

The four secular trend lines representing four definitions (bilateral lower frequency, bilateral higher frequency, unilateral lower frequency, and unilateral higher frequency) are plotted in figure 2, with CheckPoint providing the final point for each line. For all four definitions, there was evidence of rising prevalence since 1990 for hearing loss ≥16 dB HL (all P for trend <0.001). To investigate non-linear secular trends, we conducted post-hoc tests by running logistic regression models with categorical time points, time-squared and time-cubed, respectively. The results show no evidence of non-linearity (data available from authors on request).
DISCUSSION

Principal findings

Nearly 10% of Australian children aged 11–12 years old had bilateral hearing loss ≥ 16 dB HL across the main speech frequencies, with the majority slight (16–25 dB HL) in degree. Unilateral losses were more prevalent, and lower frequency losses were more common than higher frequency losses. While demographic characteristics did not convincingly predict hearing loss, prevalence estimates have substantively increased since 1990.

Strengths and limitations

Strengths include our population-based sample, standardised measurement using air-conduction audiology and tympanometry, and classification of hearing loss via four indices allowing a thorough exploration of hearing thresholds. The children were old enough for good compliance with testing protocols and accurate ascertainment of even slight losses. Sensitivity analyses indicated that, in a best-case scenario (normal middle ear function, minimal external noise), the prevalence was robust. To our knowledge, we are the first to examine secular trends using different definitions, enabling us to make comparisons with ‘like’ studies. This revealed a consistent and concerning upswing in prevalence and will support future comparisons.

There were also limitations. First, the time and cost constraints of a “whole child” examination, where hearing was just one of multiple health domains measured, precluded additional frequencies (e.g., 0.5, 1, 2 and 4 kHz) included in some studies. While this could have provided a finer-grained understanding of hearing profile, it is unlikely to have greatly altered prevalence. The 3 and 4 kHz do not add greatly to conclusions drawn from the adjacent frequencies, and we excluded the 0.5 kHz because it is less relevant to spoken speech and is most affected by the residual background noise (this could partly explain the larger upswing in lower frequency losses). Second, bone-conduction audiology would have more accurately classified sensorineural, conductive and mixed losses, but would have reduced comparability with other population studies, which mostly did not use bone-conduction audiometry. Our tympanometry indicated little influence of middle ear status on mean hearing thresholds at this age. Third, under-representation of disadvantaged families may limit generalisability to the wider socioeconomic context. Finally, we compared secular trends across studies reporting similar but not identical definitions, since this proved impossible.

Interpretation in light of other studies

Compared with the selected comparable studies, we note an upward secular trend in prevalence since 1990. Design differences would not explain away these trends. Exclusion of children with abnormal tympanograms (type B or C) by the Canadian study would have reduced prevalence, but would not alter the overall trends. Additionally, our prevalence differed slightly regardless of including or excluding children with abnormal tympanograms. Second, as the first NPH children were not assessed in soundproof booths, background noise could have contributed to some false-positive results. However, prevalence when restricted to the 930 children tested in soundproof booths was slightly higher with more frequent identification of slight lower frequency losses (online supplementary table e4). Third, although racial/ethnic make-up differs somewhat between USA, Canada, and Australia, this would not explain the changes over time seen within those countries, for example in the NHANES studies.
Figure 2: Secular trend in hearing loss ≥16 dB HL by types in school-age children: Child Health Checkpoint and historical studies. dB HL, decibels hearing level; NHANES, National Health and Nutrition Examination Survey.

Regarding risk factors, it is not surprising that age was not significantly associated with hearing loss due to our narrow range of 11-12 years. Other studies have variably noted differences in hearing loss prevalence by gender\textsuperscript{10,11} and socioeconomic status.\textsuperscript{6} We think that our few and inconsistent significant associations most likely reflect chance, but recommend further exploration with a wider range of indicators and preferably replication across studies.

Implications

It is widely accepted that non-communicable diseases are progressive, with first perturbations often starting early in life.\textsuperscript{24,25} Presbycusis >25 dB HL in either ear affects around 15%, 30% and 50% of individuals aged 40-49, 50-59 and 60-69 years, respectively.\textsuperscript{43} If slight childhood hearing losses are a harbinger of presbycusis, then this documented rise is concerning, particularly while the underlying pathology of hair cell death remains irreversible.

Implications flow: First, it is important to confirm and monitor secular trends in children, young adults and older adults. Second, to cater for multiple definitions across studies and to provide evidence to move towards a single “best” outcomes-driven definition, we recommend setting up an international repository of deidentified person-level population hearing data, similar to the accepted standard for genomic studies. Third, longitudinal studies should document hearing trajectories to establish whether these children with slight losses are indeed the adults with disabling presbycusis of the future. We urge national studies including children approaching adolescence to consider implementing audiometric assessment in future waves; ideally, this would collect a wider range of aural parameters such as bone-conduction audiometry and information on tinnitus. Fourth, causal research, especially in slight losses, is needed to identify prevention and treatment strategies. Noise exposure from sustained headphone use shows inconsistent associations especially at younger ages with likely individual differences in susceptibility to noise-induced hearing loss.\textsuperscript{33,34} Other avenues of exploration include risk factors for non-communicable diseases (eg, inflammation and adiposity as outlined in the Introduction),\textsuperscript{12,14} ear diseases (eg, otitis media), infections (eg, congenital cytomegalovirus)\textsuperscript{35} and genetics (eg, late-onset genetic losses, polygenic influences and mitochondrial DNA mutations).\textsuperscript{33} Trials are needed to determine whether reducing the effects of slight and mild hearing losses (eg, school building design, sound field systems, teaching styles) improve functional and learning outcomes.\textsuperscript{36} All of these factors could be collected in population-based studies. Finally, age-specific current and future prevalence, burden and costs should be modelled to provide policymakers with the information they need to act.

CONCLUSIONS

Childhood hearing loss is prevalent and has risen since 1990. Future research should investigate the causes, course and impact of these changes.

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Contributors

MW conceived the Checkpoint study with the Checkpoint team. MW was the primary student supervisor, along with RAB and VS, and oversaw all aspects of the study and the manuscript preparation. RSJ contributed to hearing data collection and, under the guidance of PC, designed the hearing protocols. JW and CMPC conducted data extraction, cleaning and handling. JW performed data analysis and wrote the main paper. MW, PC, RM and UG advised on statistical issues and interpretation. All authors critically reviewed the manuscript and had final approval of the submitted and published version of this paper. MW and JW had full access to all the data in the study and take responsibility for the integrity of the data and the accuracy of the data analysis.

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Original article

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Cross-sectional epidemiology of hearing loss in Australian children aged 11–12 years old and 25-year secular trends

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Appendix K: 16p11.2 deletion paper

Deep phenotyping of speech and language skills in individuals with 16p11.2 deletion

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Running timing: Speech and language in 16p11.2 deletion cases

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Abstract

Recurrent deletions of a ~600-kb region of 16p11.2 have been associated with a highly penetrant form of childhood apraxia of speech (CAS). Yet prior findings have been based on a small, potentially biased sample using retrospectively collected data. We examine the prevalence of CAS in a larger cohort of individuals with 16p11.2 deletion using a prospectively-designed assessment battery. The broader speech and language phenotype associated with carrying this deletion was also examined. Fifty-five participants with 16p11.2 deletion (47 children, 8 adults) underwent deep phenotyping to test for the presence of CAS and other speech and language diagnoses. Standardized tests of oral motor functioning, speech production, language, and non-verbal IQ were conducted. The majority of children (77%) and half of adults (50%) met criteria for CAS. Other speech outcomes were observed including articulation or phonological errors (i.e., phonetic and cognitive-linguistic errors, respectively), dysarthria (i.e., neuromuscular speech disorder), minimal verbal output, and even typical speech in some. Receptive and expressive language impairment was present in 73% and 70% of children, respectively. Co-occurring neurodevelopmental conditions (e.g., autism) and non-verbal IQ did not correlate with the presence of CAS. Findings indicate that CAS is highly prevalent in children with 16p11.2 deletion with symptoms persisting into adulthood for many. Yet CAS occurs in the context of a broader speech and language profile and other neurobehavioral deficits. Further research will elucidate specific genetic and neural pathways leading to speech and language deficits in individuals with 16p11.2 deletions, resulting in more targeted speech therapies addressing aetiological pathways.

Key words: 16p11.2 deletion, speech, language, childhood apraxia of speech
1 INTRODUCTION

A recurrent ~600-kb heterozygous deletion of 16p11.2, at approximate position
~29.5-30.2 Mb in the reference genome (GRCh37/hg19), is widely acknowledged to
affect speech and language development in individuals who carry it.\textsuperscript{14} Up to 70% of
affected individuals present with some form of communication disorder.\textsuperscript{2} Yet there is
a paucity of evidence delineating the specific types of speech and/or language
disorders associated with the deletion, limiting precise definition of the
communication phenotype.

Specificity of phenotyping is important given that childhood apraxia of speech
(CAS), a complex diagnosis, has been identified as a core communication deficit in
children with the canonical 16p11.2 deletion.\textsuperscript{2,4,8} CAS is a severe motor speech
disorder affecting the precision and consistency of speech production.\textsuperscript{5} The only study
to systematically assess speech in children with 16p11.2 deletion found all nine verbal
participants had CAS.\textsuperscript{2} These findings are of considerable interest given that the
prevalence of CAS in the general population is estimated at only 0.01–0.02%.\textsuperscript{10}

However, the study was limited by a small and potentially biased sample and the use
of retrospectively collected data.\textsuperscript{2} Further work is thus required across a larger sample
and using prospective assessments carefully designed to conduct differential diagnosis
of communication disorder.

At a broader level of communication phenotyping, beyond CAS, even the
simple differentiation between speech and language impairments in individuals with
16p11.2 deletion has received little focus,\textsuperscript{11} with some studies conflating the two. Yet
speech production\textsuperscript{12-15} is supported by cognitive and neural mechanisms distinct from
those that support higher-level language processing.\textsuperscript{2} Similarly, there has been a lack
of differentiation of expressive versus receptive language abilities, although both are
seemingly affected. Moreover, no study has examined performance in specific language subdomains (e.g., semantics, syntax, phonology). Limited evidence is also available for the related outcome of literacy, with reading and writing often examined in case reports rather than larger cohorts. Finally, given that language outcomes have often been described in cases selected for the presence of autism spectrum disorder (ASD), deep speech and language phenotyping in a sample unselected for neurodevelopmental conditions is needed to better understand the specificity and prevalence of communication deficits across the more general population of children with 16p11.2 deletion. The use of specific diagnoses reached by valid methods is essential for clinical management, as these distinct categories necessitate disorder-specific interventions.

Our aims were threefold: (1) Examine the prevalence of CAS in a large unselected cohort of individuals with 16p11.2 deletion, using a prospectively determined assessment battery; (2) Characterize specific co-occurring speech production and language diagnoses to provide a comprehensive study of the communication phenotype; and (3) Examine the relationship between CAS and other behaviors of language, oral motor function, non-verbal cognition, as well as the presence of neurodevelopmental conditions (e.g., epilepsy and/or seizures, autism) in an attempt to better understand the underlying cognitive architecture associated with 16p11.2 deletions.

SUBJECTS AND METHODS

Participants

Eligibility criteria for probands included a diagnosis of 16p11.2 recurrent microdeletion (~600-kb including DOC2A and TBX5) and an age between 2;11 and 18;0 years. Affected siblings and parents of the proband were also eligible.
Participants were excluded if they demonstrated an additional copy-number variant, a neurological condition impacting speech, or had taken part in our previously published work.  

Participants were recruited from two sources: Victorian Clinical Genetics Services (VCGS, Australia) and the 2015 Simons Variation in Individuals Project (VIP) Connect Family Meeting (Washington, US). For the US sample, the study was advertised to families attending the Simons VIP Meeting. Families within Australia were identified via the VCGS database. Probands on the database who met study eligibility (as described above) were contacted. The Human Research Ethics Committees at The Royal Children’s Hospital, Melbourne (#27053), and the Massachusetts Institute of Technology (#1306005763) approved the study.

Measures

Participants completed a range of face-to-face clinical measures, as described below. The battery took approximately 2-2.5 hours to administer. The number of participants contributing to each measure is available as supplemental material. Demographic data (e.g., hearing, vision, gross and fine motor) were obtained from parents and the Simons VIP.

Aim 1: Prevalence of CAS, and Aim 2: Broader speech and language phenotype

Speech Production (including CAS)

Differential diagnosis of speech occurred across: articulation disorder (deficit at the phonetic, motoric level of speech e.g., phonetic distortions such as a lisp or being unable to produce a particular sound); phonological disorder (deficit at the phonemic level reflecting cognitive-linguistic errors e.g., a sound can be produced but
Articulation deficits were evaluated using the Goldman-Fristoe Test of Articulation-2 (GFTA-2). GFTA-2 responses were analyzed for phonological processes and compared to normative data to identify disordered, delayed or age-appropriate processes (see Supplemental Material Table S1 for definitions). This distinction is essential given that disordered phonology (compared to delayed) is associated with persistent speech difficulties and impaired phonological awareness and literacy. Dysarthria was diagnosed based on 1) the presence of oromotor dysfunction (i.e., impaired Focal Oromotor Control based on speech and non-speech tasks, as described below) signaling impaired neuromuscular tone or co-ordination of movements; and 2) concomitant deviant perceptual speech features rated during a 10-minute conversational speech sample, affecting articulation, resonance, prosody, respiration and/or phonation.

For CAS, diagnostic criteria were adapted from Murray et al. based on three American Speech and Hearing Association consensus-based criteria, as in our previous work: 1) inconsistent production of the same word; 2) lengthened and disrupted coarticulatory transitions; and 3) prosodic errors (see Table 2). The same criteria were used for child and adult participants given the absence of diagnostic criteria for adults with CAS. Inconsistent production was examined using the standardized Inconsistency subtest of the Diagnostic Evaluation of Articulation and
Phonology (DEAP). Prosodic, syllable and sound sequencing errors typifying disrupted co-articulatory transitions were analysed using the Single-Word Test of Polysyllables. Overall speech accuracy ratings of Percent Consonants Correct (PCC) and Percent Vowels Correct (PVC) were also derived from this tool. Where this test could not be completed (n=8), speech accuracy ratings were calculated using the GFTA-2. PCC scores were categorized by severity: mild (>85), mild-moderate (65-85), moderate-severe (50-64), and severe (<50). The 10-minute conversational speech sample was also analyzed to identify further features of CAS (see Table 2).  

**Oral Motor Function**

Oral motor function was assessed in children aged ≥3 years using the Verbal Motor Production Assessment for Children (VMPAC), the most psychometrically robust oral motor assessment available for children. Three scales were administered: Global Motor Control, a measure of neuromotor innervation to peripheral muscles in the torso, neck, head and face; Focal Oromotor Control assessing control of the jaw, lips, face and tongue during speech and non-speech tasks; and Sequencing, examining performance of oral and speech movements in sequential order. Children <3 years were evaluated using the Oral and Speech Motor Control Protocol (OSMCP). The DEAP Oromotor subtest, which includes tasks examining isolated and sequenced non-speech movements, was used for two children unable to complete the VMPAC due to reduced attention. Oromotor abilities of adults (> 18 years) were measured using the Frenchay Dysarthria Assessment-2 (FDA-2). These tools assess both speech and non-speech oral motor tasks.
Language & Literacy

Australian participants <21 years were evaluated with the Clinical Evaluation of Language Fundamentals (CELF-Preschool 2 or CELF-4, depending on age). The CELF Core Language Score, reflecting overall receptive and expressive language competence, was used to identify impaired language. Children ≤3 years were assessed using the Preschool Language Scale-5 (PLS-5), a measure of receptive and expressive language. Due to time constraints, language functioning of the US child cohort (n=5) was measured using the CELF-5 Recalling Sentences subtest. Deficits in receptive and expressive language were recorded based on the CELF Core Language subtests, with impairments in specific subdomains (i.e., semantics, syntax, morphology) further identified. For those who completed the PLS-5, deficits were based on participants’ overall scores (Auditory Comprehension and Expressive Communication) and the Profile form. Auditory short-term memory was also examined in the Australian cohort aged >5 years (n=26) using the Forwards Number Repetition subtest of the CELF-4.

Seven of 8 adults were examined using the Peabody Picture Vocabulary Test-4 (PPVT-4), a measure of receptive vocabulary, and the Test for Reception of Grammar-2 (TROG-2), a measure of grammatical comprehension. The Nonword Repetition subtest of the Comprehensive Test of Phonological Processing-2 (CTOPP-2) was completed by children (n=29) and adults (n=7) as an indicator of phonological processing and awareness, early precursors of literacy. Literacy outcomes of the Australian cohort (n=26) were measured using the Word Reading and Spelling subtests of the Wide Range Achievement Test-4 (WRAT-4).
Aim 3: Associations between CAS and other neurologic cognitive-behavioral domains

Information regarding additional diagnoses (e.g., epilepsy, autism, attention deficit hyperactivity disorder (ADHD), gross and fine motor impairment) was obtained from parents and the Simons VIP. Non-verbal cognition was measured with the Wechsler Abbreviated Scale of Intelligence-2 (WASI-2)\textsuperscript{39} for participants >6 years (n=38) or Kaufman Brief Intelligence Test-2 (K-BIT-2)\textsuperscript{10} for those <6 years (n=2).

Analysis

Results for the Australian and US cohorts were combined given that demographic characteristics were comparable (Table 1). Where different measures were used across children, scores from measures examining similar constructs were grouped where possible (e.g., CELF-4, PLS-5). For Aim 1, CAS ratings were completed by CM. A second speech pathologist (AM), who was blinded to CAS status, also completed ratings for n=15, ~30% of the sample, to confirm CAS diagnoses. For Aim 2, data were transcribed and analyzed, and standard scores were computed for all relevant tests by CM to provide a differential diagnosis of speech, language, and literacy.

For Aim 3, the relationship between CAS and children’s broader phenotype (e.g., autism, ADHD, epilepsy and/or seizures, non-speech oromotor) was examined using the Fisher’s exact test. The Mann-Whitney U-test was used to compare the language, auditory short-term memory and non-verbal IQ scores of children with and without CAS. The association between CAS and speech-related oromotor impairment was examined using the Spearman's rank correlation. This test was also used to investigate the correlation between language and non-verbal IQ.
RESULTS

Altogether, 55 participants were recruited: 47 Australian (40 children, 7 adults) and 8 American (7 children, 1 adult). Sample characteristics are detailed in Table 1. Age of participants ranged from 2;11 to 17;9 years for probands (n=44) and child siblings (n=3) and 20;4 to 48;2 years for the remaining affected adult siblings (n=1) and parents (n=7). Of the child sample, 51% (24/47) were currently receiving speech and/or language therapy while 38% (18/47) had received this type of therapy in the past.

As not all participants completed each assessment (due to factors such as age, time constraints, compliance), the denominators presented below vary. Denominators used reflect the number of participants who completed each assessment, as detailed in the supplemental material (Table S2).

-Table 1-

Aim 1: Prevalence of CAS, and Aim 2: Broader speech and language phenotype

Speech production

Three child participants were excluded from speech analyses due to minimal verbal output (n=2) or missing data (n=1). Of remaining participants, 89% (39/44) had impaired speech (Figure 1). Children demonstrated a range of PCC severity ratings: mild (51%, 22/43), mild-moderate (33%, 14/43), moderate-severe (9%, 4/43), and severe (2%, 1/43). PCC data were not available for one child.

- Figure 1 -

Diagnostic criteria for CAS were met by 77% (34/44) of children (Table 2). Only 29% (10/34) of this sample had previously received a clinical diagnosis of CAS.
Whilst CAS was prevalent, articulation and phonological speech errors were also common, present in 43% (19/44) and 82% (36/44) of children, respectively. Of the 19 children with articulation errors (e.g., interdental or lateral fricatives, labiodental production of bilabials), 11 had structural deficits likely to be causing the phonetic distortion (e.g., malocclusion, submucous cleft palate, misaligned teeth). Phonological processes were classified as delayed in 36% (16/44) and disordered in 45% of children (20/44) (supplemental material Table 83).

For children, CAS occurred in conjunction with both articulation and phonological errors (23%, 10/44), phonological errors alone (34%, 13/44), articulation errors alone (5%, 2/44) or dysarthria with co-occurring articulation and/or phonological errors (16%, 7/44). Five of the ten children without a CAS diagnosis had other forms of speech impairment; isolated phonological (n=2) or articulation errors (n=1), and co-occurrence of these errors (n=2).

For the adult cases, 50% (4/8) met the CAS diagnostic criteria (Table 2). Three adults presented with articulation errors (i.e., interdental fricatives) and/or phonological processes involving ‘th’ (i.e., stopping, fronting), respectively used by 25% (2/8) of adults (one demonstrated co-occurring articulation and phonological errors). The remaining five cases showed no errors during less demanding speech tasks (e.g., conversation, GFTA-2), but produced one or more errors (e.g., metathesis, epentheses, assimilation) during more complex tasks such as producing polysyllables.

PCC speech severity ratings were mild in all impaired adult cases (71%, 5/7).

- Table 2 -

Oromotor

In the Australian child cohort, oromotor functioning was impaired in 85% (28/33).

Oromotor functioning was not assessed in 14 children. Thirty children completed the
Global Motor Control scale of the VMPAC, with performance classified as within
normal limits for 27% (8/30), moderately impaired for 23% (7/30), and severely
impaired for 50% (15/30). Within this scale, impairments were often characterized by
reductions in tongue strength (53%, 16/30), soft palate contraction (47%, 14/30) and
smoothness or range of movement (33%, 10/30).

The Focal Oromotor Control scale was completed by 29 children with
impaired functioning seen in 76% (22/29). Severity of impairment was classified as
mild (7%, 2/29), moderate (10%, 3/29), or severe (59%, 17/29). Poor performance
during single non-speech oromotor movements reflected impaired lingual movements
(48%, 14/29; e.g., reduced tongue elevation and laterализation), labial-facial
movements (31%, 9/29; e.g., poor lip rounding and, in two cases, asymmetry), and
mandibular control (10%, 3/29; e.g., reduced jaw excursion and stability). Impaired
double non-speech oromotor movements (e.g., “smile and kiss”) were common (43%,
13/29), typified by impaired transition and precision of movements. With regard to
speech-related oromotor movements, deficits were frequently noted during the
production of triple oromotor sequences (e.g., /a-m-u/; 72%, 21/29) and
words/sentences (e.g., “bow, toe, go”; 74%, 20/27). In comparison, impaired single
(e.g., /a/) and double speech movements (e.g., /a-u/) were seen in 55% (16/29) and
45% (13/29), respectively.

Twenty-nine children completed the Sequencing scale. Performance on this
scale revealed the greatest difficulty, with severe impairment noted in 79% (23/29) of
children. Mild and moderate impairments were infrequent (14%, 4/29, and 7%, 2/29)
as were intact sequencing skills (14%, 4/29). Sequencing deficits were seen across all
tasks (i.e., sequencing of non-speech oromotor movements, phonemes, and words).
Oromotor functioning was assessed in 6 of the 8 adults, with all showing some degree of impaired laryngeal and tongue control (during speech and/or non-speech tasks). Common features included reductions in phonation time, volume control, and non-speech and speech tongue movements (elevation and alternating sequences). Palatal and lip deficits were also common, both occurring in 67% (4/6) of adults. These deficits reflected poor palatal movement during phonation and reduced alternating lip movements during speech. Reflexes (e.g., cough, swallow) and intelligibility were relatively spared (both areas impaired in 1/6 cases). Reduced respiratory support for speech was observed in two adults although this was associated with a chest infection.

Language, Cognition & Literacy

For the child cases, language was not evaluated in seven due to poor child compliance (n=4), severity of verbal impairment (n=1), or missing data (n=2). Of those assessed, 83% (33/40) had a language impairment, with Core Language Scores (i.e., overall receptive-expressive abilities) greater than two standard deviations below the normative mean (Table 3). Language impairment in children commonly affected both expressive and receptive abilities (60%, 24/40) rather than only expressive (5%, 2/40) or only receptive (3%, 1/40) abilities. Differences in expressive and receptive language were not examined in 6 children as only one or the other domain was evaluated due to severe expressive language difficulties (n=2), selective mutism (n=1), or time constraints (n=3). Impairments were present across the subdomains of semantics, syntax, and morphology (Table 3). Non-verbal IQ and literacy scores for children were greater than one standard deviation below the normative mean (Table 3). Auditory short-term memory (measured using the CELF-4) and phonological
memory (based on the CTOPP-2) were respectively impaired in 80% (20/25) and 94% (32/34) of children (Table 3).

For the adults, 86% (6/7) had impaired language, with performance on the PPVT-4 and TROG-2 greater than one standard deviation below the mean (Table 3). Language was not assessed with one adult. Non-verbal IQ and literacy scores for this group were two standard deviations below the normative mean (Table 3).

**Aim 3: Associations between CAS and other neurologic cognitive-behavioral domains**

Results from the Fisher’s exact test did not identify an association between CAS status in the children and autism \( (p = 0.15) \), ADHD \( (p = >0.999) \), gross or fine motor function \( (p = 0.27, \text{ and } p = 0.70, \text{ respectively}) \), epilepsy and/or seizures \( (p = 0.47) \), Similarly, the Mann-Whitney U test did not reveal an association between CAS and language \( (U = 64, p = 0.12) \) and non-verbal IQ \( (U = 69, p = 0.15) \), although CAS was associated with poor short-term auditory memory \( (U = 21, p = 0.01) \).

The association between CAS and non-speech oromotor function in children, measured by the Fisher’s exact test, was not statistically significant \( (p = 0.55 \text{ for mandibular control, } p = 0.37 \text{ for labial-facial control, and } p = 0.39 \text{ for tongue function). Similarly, results from the Spearman’s rank test revealed that Focal Oromotor Control percentage scores (i.e., speech and non-speech oromotor performance) were not correlated with GFTA-2 standard scores for children with CAS \( (r = 0.13, p = 0.58) \). This pattern suggests that articulation was not associated with oromotor performance for this group. Performance on the Sequencing scale was related to DEAP Inconsistency scores \( (r = -0.93, p = <0.001) \) but not GFTA-2 scores.
for children with CAS, indicating that children with poorer sequencing abilities (during speech and non-speech tasks) produced a higher number of inconsistent speech errors. Children’s language scores (reflecting overall receptive and expressive language competence) were not correlated with non-verbal IQ ($r = -0.08, p = 0.73$), even when children with hearing impairment were excluded ($r = -0.04, p = 0.86$). Notably, the language abilities of 6 children were $\geq 2$ standard deviations below their non-verbal IQ.

**DISCUSSION**

In line with our previous work,\textsuperscript{2} we found that CAS is prevalent (77%) in individuals with canonical 16p11.2 deletions. Further, our findings suggest that speech and/or language disorder is more common than previously estimated in this population, with 98% of children demonstrating some form of speech or language impairment and a significant proportion (57%) having co-occurrence of these deficits. Other neurobehavioural conditions (e.g., epilepsy and/or seizures, autism) were not correlated with a diagnosis of CAS.

Although most children (89%) demonstrated impaired speech, severity ratings were mild or mild-moderate for the majority of the cohort (i.e., PCC ratings $>65$), and only two cases were minimally verbal. Interestingly, only 29% of children with CAS had previously received a clinical diagnosis of CAS (despite all previously having seen a SLP), suggesting potential misdiagnosis in the clinical community and, consequently, lack of targeted intervention of motor planning and programming deficits, as needed for improving these children’s outcomes. This result is not surprising considering the variable criteria used by clinicians for the diagnosis of
CAS. Our findings highlight the need for clinicians to systematically test for the presence of CAS based on the ASHA consensus criteria in children with 16p11.2 deletion to improve management of speech in this population. Further, our data show that CAS commonly occurs with other speech disorders (dysarthria, articulation, and phonological), hence a careful differential diagnosis is required and clinical problem solving needed to therapeutically target the specific level(s) of deficit. In addition to managing surface behaviors, health professionals may consider referring children with CAS for clinical genetic testing if indicated to determine potential underlying causes such as 16p11.2 deletion.

In relation to language, a fairly consistent profile was demonstrated in that deficits involved both receptive and expressive language. This is in line with previous reports where a mixed receptive-expressive language impairment profile has been associated with 16p11.2 deletion. Here, however, we highlight that the language deficit is not restricted to a particular subdomain, rather it is generalized affecting morphology, syntax and semantics. This finding is shared by other syndromes associated with intellectual disability (e.g., Floating Harbour and Kabuki). Our findings further refine the phenotype associated with 16p11.2 deletion by revealing that it is not linked to a syndrome-specific language profile where only specific subdomains are affected.

Although it is probable that children’s broader cognitive profile is related to their language attainment given their shared genetic bases mean language scores were nearly one standard deviation below non-verbal IQ. In contrast, children’s literacy outcomes, as measured by the WRAT-4, were a relative strength and more commensurate with average non-verbal IQ performance. This differs from previous research where strengths in literacy were not as apparent in comparison to language.
For instance, in Hanson et al. word reading abilities were, on average, just below verbal IQ (76 and 79, respectively) whereas here, a greater split between word reading and language abilities was apparent (83 and 69, respectively). This difference could potentially be due to differences in measures or sample characteristics (e.g., in Hanson et al. 46% had language impairment whereas here it occurred in 83% of children).

With regard to adults, all had mild speech severity ratings and typically only issues during more demanding speech motor tasks (e.g., polysyllabic words). Notably, the diagnostic criteria for CAS were met by 50% of adults with the remaining cases showing some symptoms despite not meeting the pre-defined criteria for a formal diagnosis. Importantly, the phenotype of CAS differed between adults and children, consistent with the broader CAS literature. Specifically, children demonstrated features such as syllable segregation and voicing errors that were not seen in adults. Although CAS is recognized as a persistent speech disorder, how it evolves during adulthood remains poorly understood, including for individuals with 16p11.2 deletions. The cross-sectional design of the current study makes it difficult to infer the progression of speech disorders for this population, necessitating the need for future large scale prospectively designed longitudinal studies.

In terms of associated factors, CAS occurred independent of other neurobehavioural conditions, such as autism, ADHD, and epilepsy and/or seizures in children. Non-verbal IQ was also not predictive of CAS. These results should be interpreted with some caution however, given the limitations of our study (e.g., small sample size and the wide age range of participants, which may have impacted on analyses that included neurodevelopmental conditions that are not typically diagnosed in early infancy e.g., ADHD). Our finding of an association between CAS and
memory deficits is consistent with prior reports. Speech (articulation and phonology) and language outcomes could have been impacted by hearing impairment (conductive), which occurred in 19% of the child sample. This is particularly plausible for speech outcomes as all children with impaired hearing had a speech deficit, typically articulation and/or phonological errors that occurred in the presence of CAS. A limitation of our data is that hearing status was obtained via parent-report, potentially under-representing the occurrence of hearing impairment in this cohort.

It is most probable that CAS observed in people with 16p11.2 deletions is associated with specific neural and genetic markers yet to be explored in this population. While the speech phenotype associated with 16p11.2 deletion is largely expressed as CAS, the following profiles may be seen: i) typical speech-sound development, ii) isolated developmental articulation disorder or phonological delay, iii) CAS with or without dysarthria, articulation or phonological errors, or even iv) non-verbal presentation. Of interest would be to determine the combination of genetic and neural markers that can predict children’s speech outcome. Although language impairment has been linked to the number of genomic copies of the 16p11.2 region and abnormal development of neural pathways supporting language function (i.e., left arcuate fasciculus) in cases with 16p11.2 deletion, the mechanisms underlying speech dysfunction for this group remain unknown.

Future research may also examine whether more severe phenotypes observed in individuals with 16p11.2 deletion (e.g., non-verbal presentations) are associated with the ‘two-hit’ model suggested by Girirajan et al. This model proposes that severe phenotypes linked to 16p12.2 microdeletions result from a secondary insult (e.g., an additional copy number variant, gene mutation or environmental factor). The
model has been applied to children with 16p11.2 deletion but it has yet to be determined how relevant it is for explaining speech outcomes.

Whilst we aimed to recruit a sample less biased than previous reports and unselected for neurodevelopmental conditions (e.g., ASD), our cohort may have been biased towards children with developmental concerns due to our method of recruitment (i.e., a genetics service where individuals are often referred due to specific clinical indications). Additionally, it is possible that being advertised as a speech and language study, our sample could be biased towards individuals with speech and language conditions. We, however, aimed to mitigate this by indicating that all individuals, even those without speech and language disorders, were eligible.

This study is the largest to date to systematically assess speech and language skills in children with 16p11.2 deletion. We confirm that CAS is a core component of the speech phenotype seen in children with 16p11.2 deletion. A potential extension of this work is to compare the phenotype against intra-familial controls and 16p11.2 duplication cases. Future research should be directed at examining causal pathways in order to identify the specific genetic and neural mechanisms underpinning CAS. This will provide much needed data informing the etiology of CAS given that only a small proportion of cases can currently be explained by known genetic mutations (e.g., FOXP2 and GRN2A). Supplementary information is available at European Journal of Human Genetics’ website.
REFERENCES


1. Titles and legends to figures
2. Figure 1. Prevalence of speech production disorders.
Table 1: Participant characteristics

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<thead>
<tr>
<th></th>
<th>Children</th>
<th>Adults</th>
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*a Submucous cleft palate.
*b Differences between child cases (Australian and US cohorts) significant at 0.05 level (Fisher’s exact test).
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*Not completed by 12 children and all of the adults. *Not completed by 12/34. *Not completed by 2/34. *Unknown for one child (who did not meet the criteria for CAS) since their data was obtained from clinical reports.

C/V: consonant/vowel. DDK: diadochokinesis.
### Table 3 Language, literacy and cognitive outcomes

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<th>Child cases (n=47)</th>
<th>Adult cases (n=8)</th>
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<tr>
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<td>Impaired n (%)</td>
<td>Mean, SD (Range)</td>
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<td>Core language</td>
<td>26/31 (84%)</td>
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<td>Receptive semantics</td>
<td>22/27 (81%)</td>
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<tr>
<td>Receptive syntax</td>
<td>3/3 (100%)</td>
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<tr>
<td>Expressive semantics</td>
<td>18/22 (82%)</td>
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</tr>
<tr>
<td>Expressive morphology</td>
<td>11/11 (100%)</td>
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<td>Expressive semantics and morphosyntax</td>
<td>25/30 (83%)</td>
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<td>PPVT-4*</td>
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<td>TROG-2*</td>
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<tr>
<td>Average</td>
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<td>Borderline</td>
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<td>Extremely low</td>
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<td>Auditory short-term memory</td>
<td>20/25 (60)</td>
<td>5.2, 1.9 (1-9)</td>
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<tr>
<td>CTOPP-2*</td>
<td>32/34 (94%)</td>
<td>4.4, 2.4 (1-9)</td>
</tr>
</tbody>
</table>

---

*Normative mean = 100 (SD 15). *2Normative mean = 10 (SD 3). Scores greater than 1SD below the normative mean were classified as impaired except for non-verbal IQ, which was classified as high average (110-119), average (90-109), low average (80-89), borderline (70-79) and extremely low (<70).
*4US cohort only included as they only completed the Recalling Sentences subtest. Language was unable to be assessed in 7 of the Australian children due to reasons such as the cooperation of the child, profound language difficulties, or the absence of a recent language assessment for children whose data were obtained from clinical reports.
*5Cognitive testing not possible for 14 children (due to the child’s age or capabilities) and 1 adult.
*6*1 child completed the PPVT-4, a measure of receptive vocabulary, as they were unable to complete the CELF-4 due to severe speech and language impairment.
*7*Measured using the CELF-4 Number Repetition Forwards subtest (Australian cohort only).
*8*Not completed by 5 children who were unable to complete the sample mean.
Author/s: Carew, Peter

Title: Mild and moderate congenital hearing loss in childhood: trends and associations with language outcomes

Date: 2018

Persistent Link: http://hdl.handle.net/11343/214534

File Description: Complete thesis

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