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Early detection and intervention for unilateral hearing loss and mild bilateral hearing loss in children: Clinical practices and outcomes

Edited by
Teresa Y. C. Ching, Elizabeth Fitzpatrick, Kerttu Huttunen, Carmen Kung, Ulrika Löfkvist and Valerie Sung

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Early detection and intervention for unilateral hearing loss and mild bilateral hearing loss in children: Clinical practices and outcomes

Topic editors

Teresa Y. C. Ching — Macquarie University, Australia
Elizabeth Fitzpatrick — University of Ottawa, Canada
Kerttu Huttunen — University of Oulu, Finland
Carmen Kung — Macquarie University, Australia
Ulrika Löfkvist — Uppsala University, Sweden
Valerie Sung — Murdoch Childrens Research Institute, Royal Children's Hospital, Australia

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Alessandro Martini,
University of Padua, Italy

*CORRESPONDENCE
Teresa Y. C. Ching
✉ teresaychingagmail.com

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Editorial: Early detection and intervention for unilateral hearing loss and mild bilateral hearing loss in children: clinical practices and outcomes

Teresa Y. C. Ching[1,2,3]*, Elizabeth M. Fitzpatrick[4,5], Kerttu Huttunen[6,7,8], Ulrika Löfkvist[9,10], Carmen Kung[11] and Valerie Sung[12,13,14]

[1]NextSense Institute, NextSense, Sydney, NSW, Australia, [2]Macquarie School of Education, Macquarie University, Sydney, NSW, Australia, [3]School of Health and Rehabilitation Science, University of Queensland, Brisbane, QLD, Australia, [4]Faculty of Health Sciences, University of Ottawa, Ottawa, ON, Canada, [5]Child Hearing Laboratory, CHEO Research Institute, Ottawa, ON, Canada, [6]Research Unit of Logopedics, Faculty of Humanities, University of Oulu, Oulu, Finland, [7]Department of Otorhinolaryngology, Head and Neck Surgery, Oulu University Hospital, Oulu, Finland, [8]Medical Research Center Oulu, University of Oulu, Oulu, Finland, [9]Department of Public Health and Caring Sciences, Uppsala University, Uppsala, Sweden, [10]Department of Clinical Service, Intervention and Technology (CLINTEC), Karolinska Institute, Stockholm, Sweden, [11]Department of Linguistics, Macquarie University, Sydney, NSW, Australia, [12]Prevention Innovation, Population Health, Murdoch Children's Research Institute, Parkville, VIC, Australia, [13]Centre for Community Child Health, Royal Children's Hospital, Parkville, VIC, Australia, [14]Department of Pediatrics, University of Melbourne, Parkville, VIC, Australia

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Editorial on the Research Topic
Early detection and intervention for unilateral hearing loss and mild bilateral hearing loss in children: clinical practices and outcomes

Most newborn hearing screening (NHS) programs aim to identify children with permanent moderate to profound bilateral hearing loss. In addition to these children, NHS programs may identify children with unilateral hearing loss (UHL) and those with mild bilateral hearing loss (MBHL). Previously, these children have typically been identified at 4–5 years of age. The very early identification of children with UHL raises questions about early management, particularly about early provision of amplification for which there is no clear evidence. A similar lack of evidence for the efficacy of intervention exists for children with MBHL. The contributions to this special issue reflect state-of-the-art evidence on clinical practices and outcomes of children who are impacted by a hearing loss during an important developmental period, especially for speech, language, listening effort, emotions and behaviour, and quality of life.

Early detection has provided an opportunity to better map the audiologic characteristics and trajectory of hearing loss in children with UHL. This is the focus of two papers in this issue, which reported findings from studies in Australia and Canada. Zhang et al. provided detailed audiologic and clinical profiles of 91 children with congenital UHL. The authors drew attention to the challenge of obtaining reliable behavioral audiologic profiles because despite early diagnosis at an average of 2.1

months, almost half of the children were over age 3 years before the first reliable audiogram was obtained. This study also showed that 78% of children showed deterioration in hearing by the time of their first behavioral audiogram (average age 3 years). Notably, 73% of the children with deterioration progressed to severe to profound hearing loss. In the second paper, Fitzpatrick et al. also undertook an in-depth examination of the trajectory of unilateral hearing loss. Almost half of 177 children followed for an average of 58.9 months experienced further deterioration in hearing including 12% who developed bilateral hearing loss. These reports underscore the importance of careful monitoring of even very mild degrees of UHL, given that 2–3 of every 4 children appear to be at risk for further hearing deterioration in one or both ears.

Other challenges in clinical management of UHL are reported in two papers. Horrocks et al. study showed that children admitted to neonatal intensive care units with UHL, when compared to matched controls without hearing loss, were more likely to have congenital anomalies, developmental impairments and requirements for speech and language therapy. The authors highlighted the need for screening for this group of children because many of the congenital anomalies were not detected at birth, including genetic and clinical follow-up. Patel et al. showed that the lack of evidence in guiding early management of UHL has resulted in much of the decisions on trialing hearing devices being parent- or child-led rather than clinician-led. Of the children who were fitted with hearing devices in the reported cohort, most occurred late (mean age 4.7 years). The authors also highlighted the lack of funding for support services and cochlear implants, despite 28.5% of the families reporting concerns around their care.

The presence of hearing loss reduces auditory input, potentially affecting outcomes. Three studies reported on psychosocial, language and quality-of-life outcomes. Ong et al. showed that school-aged children with UHL and MBHL were just as likely as those with moderate to profound hearing loss to experience more emotional/behavioural difficulties, poorer health-related quality of life, and higher distress reported in their parents compared to population norms. Carew et al. showed that, on average, children with MBHL had poorer language outcomes than those with UHL, and both groups had lower scores compared to population norms. The total health-related quality of life scores were, on average, similar between UHL and MBHL groups. On the other hand, Cupples et al. found that children with congenital UHL had language, functional performance, speech intelligibility and quality-of-life outcomes similar to population norms, but passage comprehension and speech perception in noise were significantly below the typical range. They identified a relationship between better nonverbal cognitive ability and language results, which underscored the importance of examining cognitive ability in future studies.

Binaural processing can help listeners locate sound sources and improve their ability to hear and understand target speech in noisy environments. As these benefits relied on combining auditory inputs from both ears, they are lessened when hearing is reduced in one or both ears. Two studies explored the consequences of decreased audibility. Lewis et al. reported that children with untreated UHL or MBHL located talkers more easily and achieved better speech perception in noise when assessed in the audio-visual condition than in the auditory-only condition. On average, children with UHL exhibited better speech perception than children with MBHL, but both groups performed more poorly than peers with typical hearing. Dahlgren et al. showed that children with unilateral aural atresia experienced difficulties in localisation, with performance inversely related to degree of hearing thresholds in the atretic ear. However, early aiding with bone conduction hearing aids had mixed effects. A further consequence of decreased audibility is listening-related fatigue, which may underpin problems experienced by listeners with hearing loss. Adams et al. described fatigue experienced by 6- to 16-year-old children with and without hearing loss. Compared with children with typical hearing, listening can cause more fatigue not only for children with bilateral hearing loss but also for children with UHL. This calls for improving the acoustics of learning environments for children.

The articles in this research topic exemplify the diverse outcomes of contemporary cohorts of children with UHL and MBHL, part of which may be attributed to the lack of evidence-based guidelines for management. Despite an unsuccessful attempt to conduct a randomized control trial of amplification for children with MBHL by Sung et al. they shared learnings on engaging families in trials that might generate high-quality evidence. Cupples et al. called for research to evaluate the fitting of hearing devices using random assignment to avoid any confounding influence of degree of hearing loss or past/current level of progress. More research is needed to understand factors influencing the somewhat atypical speech, language and psychosocial outcomes. Given the heterogeneity of children with UHL or MBHL, and the clear risk of progression of hearing loss, it is crucial to develop tailored intervention options and evidence-based guidelines for management.

Author contributions

TC: Writing – original draft, Writing – review & editing. EF: Writing – original draft, Writing – review & editing. KH: Writing – original draft, Writing – review & editing. UL: Writing – original draft, Writing – review & editing. CK: Writing – original draft, Writing – review & editing. VS: Writing – original draft, Writing – review & editing.

Conflict of interest

The authors declare that the research was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

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EDITED BY
Teresa Y. C. Ching,
NextSense Institute, Macquarie University, Australia

REVIEWED BY
Barbara Cone,
University of Arizona, United States
Karen Liddle,
Children's Health Queensland, Australia

*CORRESPONDENCE
Sally K. Thornton
✉ sally.thornton@nottingham.ac.uk

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Multiple congenital anomalies and adverse developmental outcomes are associated with neonatal intensive care admission and unilateral hearing loss

Lucy M. Horrocks[1], Pádraig T. Kitterick[1,2], Dulip S. Jayasinghe[3], Karen R. Willis[4], Katherine R. M. Martin[5], Abhijit Dixit[6] and Sally K. Thornton[1,2]*

[1]Hearing Sciences, School of Medicine, The University of Nottingham, Nottingham, United Kingdom, [2]NIHR Nottingham Biomedical Research Centre, Nottingham University Hospitals NHS Trust, Nottingham, United Kingdom, [3]Neonatal Intensive Care Unit, City Hospital Campus, Nottingham University Hospitals NHS Trust, Nottingham, United Kingdom, [4]Children's Audiology, Nottingham University Hospitals NHS Trust, Nottingham, United Kingdom, [5]Children's Development Centre, City Hospital Campus, Nottingham University Hospitals NHS Trust, Nottingham, United Kingdom, [6]Clinical Genetics, City Hospital Campus, Nottingham University Hospitals NHS Trust, Nottingham, United Kingdom

Aim: To determine congenital and developmental outcomes of children with Unilateral Hearing Loss (UHL) who were admitted to the Neonatal Intensive Care Unit (NICU).

Method: Retrospective, single-site study that followed 25 children with permanent congenital UHL *and* a NICU admission to a NICU of Nottingham University Hospital. Birth and two-year developmental follow-up data were collected. They were compared to matched control group who had a NICU admission but no hearing loss (matched on gestational age, weight and sex).

Results: The median birthweights, gestational ages and number of days spent on the NICU for the UHL population were 2510 g, 36 weeks, and 12 days respectively. Most children (20/25; 80%) with UHL *and* a NICU admission were diagnosed with a congenital anomaly within the first two years of life. Only half (13/25) of these children were diagnosed with a congenital anomaly at discharge. Children with UHL *and* a NICU admission were more likely than the matched group (NICU admission only; $p < .001$) to have multiple congenital anomalies. We found a positive association between multiple congenital anomalies and developmental impairment for the NICU graduates with UHL ($p = .019$). This UHL-NICU group were also more likely than the matched NICU children to have developmental impairment (7/25 vs. 0/25; $p = .01$), speech and language therapy (13/25 vs. 1/25; $p < .001$), inner ear malformations (14/25 vs. 0/25, $p < .001$) or craniofacial anomalies (12/25 vs. 2/25; $p = .004$).

Interpretation: Children with UHL *and* a NICU admission were at high risk of congenital anomalies and certain adverse developmental outcomes. Improved congenital anomaly screening is needed at birth for this population. Having multiple congenital anomalies suggests closer developmental monitoring is needed. This study contributes towards producing clinical screening and management guidelines to ensure consistent high-quality care for this unique population.

KEYWORDS

unilateral hearing loss, neonatal intensive care, congenital anomalies, developmental outcomes, congenital hearing loss, paediatrics

Introduction

Babies born with unilateral hearing loss (UHL) who were admitted to the neonatal intensive care unit (NICU) are a very understudied population, with *no* current literature on their specific developmental outcomes. Currently there are *no* national hearing or NICU guidelines surrounding the management of this population or of the birth population of children with UHL. UHL is hearing loss in only one ear, it's prevalence in the birth population is estimated at 0.3–1 per 1,000 births (1–3) and whilst there is little literature documenting it's prevalence in the NICU population, some studies estimate 1.2%–4.6% (4, 5).

There has been no research investigating two-year outcomes of children with UHL *and* a NICU admission, although there has been research into the birth population of children with UHL and for children with normal hearing with a NICU admission.

Approximately 40% of children with UHL require speech and language therapy (SLT) (6). A need for further academic support has also been identified in this population, with studies finding that 45% of children with UHL need an Individualised Education Program (IEP) (7) and almost a third of children with UHL fail a grade (8). It is established in the literature that congenital anomalies such as craniofacial abnormalities and inner ear malformations are common in this population (3, 9–11) and the prevalence of bony malformations in the inner ear and/or internal auditory canal was markedly higher in infants with congenital UHL than in infants with bilateral hearing loss (9), with about two thirds (66.7%) of children with UHL having inner-ear and/or internal auditory canal malformations (12). However, little research has been undertaken into the number and further "non ear" related variety of congenital anomalies present in children with UHL.

For children admitted to the NICU, Schiariti et al. found that 12.6% of term babies had a congenital anomaly; 3.1% of them were cardiac and/or circulatory anomalies, 2.7% were gastrointestinal anomalies and 0.4% were face and neck anomalies (13). Those that are preterm and/or have a low birth weight are more likely to have visual and auditory impairments (14), with 3%–5% of babies <1500 g having a hearing impairment, compared to less than 1% of babies born at term (15). Babies born pre-term and/or of low birth weight are more likely to have learning difficulties (14). One study found that a third of preterm babies (32–36 weeks) had motor, speech and educational difficulties in childhood (14).

The absence of published literature surrounding the developmental outcomes of children with UHL *and* a NICU admission has highlighted the need for this exploratory study. The primary aim is to describe the congenital, anatomical and behavioural outcomes of this population, with the aim to determine a constellation of features specifically associated with this group of children. Genetic screening is not currently funded for children with UHL in the UK.

This information from a larger cohort could inform the development of clinical guidelines on the follow up and management of this understudied population, ensuring effective and consistent care.

Materials and methods

Participants

This retrospective, longitudinal case-controlled cohort study documents routinely collected NICU discharge data, two-year developmental follow-up data and hearing aid treatment data which was collected and analysed for patients with UHL and without UHL *and* a NICU admission to Nottingham University Hospitals. For the UHL infant group the inclusion criteria included a NICU admission and a referral from the UK National Newborn Hearing Screening Programme (NHSP) with a subsequent diagnosis of permanent UHL confirmed *via* auditory brainstem response (ABR) for birth dates between February 28, 2008, and 12 July 2019. This gave the final cohort of 25 patients (13 females, 12 males; median gestational age 36 weeks; median birthweight 2510 g).

Data was also collected for matched and peer controls. The matched control patients had passed the NHSP screening and were matched on sex, birthweight (±10 g), gestational age (±1 week) and whether they spent ≥48 h NICU, as it is already well established that these variables impact a baby's

development. Few exceptions were made when no matched babies fit these criteria; the weight range was increased by 10 g, and one patient was not matched on sex. If more than one baby fit the matched control criteria, then the baby born most recently was chosen. Peer control patients had passed the NHSP screening and were matched on date of birth (±4 days) and sex. Their weights, gestational ages and days spent on the NICU were recorded. No significant difference was detected regarding the birthweights, gestational ages and number of days spent on the NICU between the children with UHL and their matched controls or peer controls ($p > 0.05$ for all comparisons).

The NEAT database has been approved by Research Ethics Committee (REC 22/SC/0337; IRAS 292263).

Procedure

Data recorded between 0 and 2 years included data from (pre-term) birth to 2 years. Data recorded at birth was taken from NICU discharge notes. We analysed all data from birth to the time of the study and utilised paper notes or NHS databases. Demographic data, discharge summaries and the patient's first postcode were recorded from the neonatal database, Badger. If a patient had multiple anomalies within one organ or organ system, this was still recorded as one anomaly.

Developmental impairment is a diagnosis of their development which is not in the normal range for their corrected gestational age. Specifically, a child was diagnosed with developmental impairment if their developmental skills fell two SD or more below the population mean in two or more developmental domains. Developmental impairment was included in this study if it was stated in the patient notes that they had a global developmental delay, fine motor delay, delay in communication, developmental impairment or had been referred for developmental needs.

In terms of eligibility for SLT in the study area, the local SLT specialist hearing impaired team only accept referrals for children with severe/profound bilateral hearing loss and auditory spectrum neuropathy disorder. Referral to local community SLT is possible for children with other degrees of hearing loss, however this is not done as routine at point of diagnosis by the current service.

Home postcode on discharge from NICU was used to determine deprivation index (16, 17). The lower the index of multiple deprivation decile, the more deprived the area was that the patient lived [1 = most deprived, 10 = least deprived (16, 17)]. These patients were then split into two groups – (i) index of multiple deprivation decile of <5 (more deprived) and (ii) index of multiple deprivation decile of ≥5 (less deprived).

Follow-up data for the UHL cases and their matched counterparts were collected and used to determine abnormal anatomical features between 0 and 2 years. There were four exceptions in the UHL cohort; three eye problems and one inner ear malformation were detected *after* 2 years. A consultant neonatologist determined which anomalies were congenital.

The date of first fitting of a hearing device and the types of devices trialled were recorded. It was then calculated how many patients had their device fitted before 1 year of age. Patients not recorded on the database were assumed to have never trialled a hearing device.

Hearing thresholds for the patients with UHL were recorded and their most recent audiology report was used. The patients with UHL were then split into two groups, those that had mild/moderate hearing loss and those that had severe/profound hearing loss according to the British Society of Audiology guidelines (18).

See **Supplementary Appendix S2** for the full list of diagnoses included and definition of major and minor craniofacial abnormalities.

Missing data: The number of children with UHL may be higher than we recorded in this population as cases may be missed for a number of reasons: Firstly, while many US screening programmes include babies with permanent mild BHL and UHL in their target group, NHSP in the UK does not. It aims to identify all children with a moderate-profound permanent HL in the better hearing ear. As a by-product, the screen will identify babies who have UHL and, in some cases, mild permanent hearing loss, as well as temporary hearing loss. Babies with UHL may also be missed if they had moved out of area after NICU admission or if their data was not available on Badgernet.

Statistical analysis

A power calculation was not performed as the sample size was limited by the number of available UHL cases with NICU admissions registered at the Nottingham University Hospitals, 2008–2019. Mann–Whitney *U* test was performed for non-categorical data and chi-squared for categorical data (or Fischer's Exact if f $n \leq 5$ in either group being compared).

Shapiro-Wilk test indicated that the non-categorical data were not normally distributed.

Results

In this study, 14,538 babies were admitted to the NICU, 25 of whom had UHL, making the prevalence of UHL in this NICU cohort approximately 0.17%. This is lower than previous reports (1.23%–4.6%) (4, 5).

Congenital anomalies

Congenital anomalies that were detected at birth, and between discharge and two years, were documented in Table 1. Over half (13/52) of cases with UHL were diagnosed with a congenital anomaly at birth, with the majority of cases being diagnosed with multiple congenital anomalies (8/13). Surprisingly, analyses reveal that more than a quarter ($n = 7$) of patients with UHL were diagnosed with an anomaly that was thought to be congenital, post-discharge, but within the first 2 years of life. Therefore, when the total number of children with UHL with congenital anomalies detected at birth and post-discharge are combined, the majority (20/25) of babies with UHL had a least one congenital anomaly. Most (16/20) of these cases had multiple congenital anomalies. Children with UHL were significantly more likely to have a congenital anomaly that was diagnosed either at birth ($p < 0.001$) or post NICU discharge between 0 and 2 years ($p < 0.001$) compared to their matched counterparts. They were also significantly more likely to have multiple congenital anomalies (>1 anomaly) at birth ($p = 0.004$) and between 0 and 2 years ($p < 0.001$) compared with the matched control group.

Abnormal anatomical features

Table 2 presents the abnormal anatomical features in the UHL cohort between 0 and 2 years. A statistically significant difference was found regarding total number of anomalies between children with UHL (71) and their matched controls (21) ($p < 0.001$). Children with UHL were 6 times more likely to have vision and eye issues, 5 times as likely to have other malformations, 4 times as likely to have neurological and spinal issues, 3.5 times as likely to have gastrointestinal problems, 3 times as likely to have renal anomalies and 2.5 times as likely to have cardiac abnormalities than their matched counterparts. However, these differences were not found to be statistically significant ($p > 0.05$). Only the differences in inner ear malformations (IEM) and craniofacial anomalies were statistically significant, yielding a p-value of <0.001 and 0.004 respectively. Division of craniofacial anomalies into subgroups revealed a significant difference from matched controls only detected for ear anomalies ($p < .05$). No statistical differences were found between the patients with UHL and their matched controls for respiratory, metabolic or neuromotor anomalies.

TABLE 1 Congenital anomalies detected at birth.

	At birth		Between birth and 2 Years	
Number of congenital anomalies	UHL ($N = 25$)	Matched controls	UHL ($N = 25$)	Matched controls ($N = 25$)
0[a]	12 (48%)	24 (96%)	5 (20%)	21 (84%)
≥1[a]	13 (52%)	1 (4%)	20 (80%)	4 (16%)
≥2[a]	8 (32%)	0 (0%)	16 (64%)	2 (8%)
≥3	4 (16%)	0 (0%)	13 (52%)	2 (8%)
≥4	2 (8%)	0 (0%)	8 (32%)	0 (0%)

[a]Indicates there was a statistically significant difference between the cases (UHL) and matched controls using a chi-squared test ($n > 5$), or Fischer's Exact where $n \leq 5$ ($p < .05$).

Behavioural outcomes

Table 3 describes the behavioural outcomes of the UHL patients that were recorded to date. 52% of patients with UHL had SLT, 28% had a developmental impairment and 16% had a learning disability. There was a significant difference between the UHL group and their matched controls for SLT ($p < 0.001$) and developmental impairment ($p = .01$). It was observed that *all* of the UHL patients with a developmental impairment also had multiple congenital anomalies, with 4/7 (57%) having ≥4 congenital anomalies. Analyses revealed that patients with UHL *and* a developmental impairment were statistically more likely to have multiple congenital anomalies than those without a developmental impairment ($p = .019$).

Syndromic and genetic data

Genetic testing was carried out on twelve patients with UHL, and eleven results were obtained. Four of these patients had a genetic variant, two of which were pathogenic and two of uncertain significance. It is not known if these variants were causative of hearing loss. Three out of the four patients that had a genetic variant also had a phenotypic syndrome diagnosed by a paediatrician.

Six patients with UHL were diagnosed with a syndrome, four of which are known to be associated with hearing loss. These were Oculo-auriculo-vertebral Syndrome (19), Large Vestibular Aqueduct Syndrome (20), Klippel Feil Syndrome (21) and Beckwith Wiedemann Syndrome (22). Of those with a syndromic diagnosis, all 6 had multiple congenital anomalies, 4 had a developmental impairment and 4 were enrolled in SLT.

No syndromic or genetic data were recorded for the matched controls.

Hearing data

Fourteen of the 25 patients with UHL had mild or moderate hearing loss and 11 had severe or profound hearing loss in their affected ear. More than half of the patients (14/25) had an index

TABLE 2 Describing abnormal anatomical features detected between birth and 2 years.

	Detected between birth and 2 years		Detected between birth and current age	
Anatomical anomaly	UHL ($N = 25$)	Matched controls ($N = 25$)	UHL ($N = 25$)	Matched controls ($N = 25$)
Inner ear malformations	14 (56%)	0 (0%)	14 (56%)[a]	0 (0%)
Craniofacial anomalies	12 (48%)	2 (8%)	12 (48%)[a]	2 (8%)
Major anomaly only	4 (16%)	0 (0%)	4 (16%)	0 (0%)
Minor anomaly only	6 (24%)	2 (8%)	6 (24%)	2 (8%)
Major and minor anomaly	2 (8%)	0 (0%)	2 (8%)	0 (0%)
Ear anomaly only	5 (20%)	0 (0%)	5 (20%)[a]	0 (0%)
Face or head anomaly only	4 (16%)	2 (8%)	4 (16%)	2 (8%)
Ear and face or head anomaly	3 (12%)	0 (0%)	3 (12%)	0 (0%)
Neurological and spinal	5 (20%)	1 (4%)	8 (32%)	2 (8%)
Gastrointestinal	5 (20%)	1 (4%)	7 (28%)	2 (8%)
Vision and eye	1 (4%)	0 (0%)	6 (24%)	1 (4%)
Other malformations	5 (20%)	1 (4%)	5 (20%)	1 (4%)
Cardiac	5 (20%)	0 (0%)	5 (20%)	2 (8%)
Respiratory	3 (12%)	0 (0%)	4 (16%)	3 (12%)
Metabolic	2 (8%)	0 (0%)	4 (16%)	5 (20%)
Renal	1 (4%)	1 (4%)	3 (12%)	1 (4%)
Neuromotor	1 (4%)	0 (0%)	3 (12%)	2 (8%)
Total number of anomalies	41	6	71[b]	21

[a]Indicates there was a statistically significant difference between the cases (UHL) and matched controls using Fischer's Exact (as $n \leq 5$).
[b]Indicates there was a statistically significant difference between the cases (UHL) and matched controls using Mann–Whitney *U* test.

TABLE 3 Behavioural outcomes recorded to date.

	UHL ($N = 25$)	Matched controls ($N = 25$)
Speech and language therapy[a]	13 (52%)	1 (4%)
Developmental impairment[a]	7 (28%)	0 (0%)
Learning disability	4 (16%)	0 (0%)
Autism	1 (4%)	0 (0%)

[a]Indicates there was a statistically significant difference between the cases (UHL) and matched controls using Fischer's Exact (as $n \leq 5$).

of multiple deprivation decile of <5 (associated with lower socio-economic status according to their postcode) on discharge from the NICU. Nearly half (12/25) had trialled a hearing device, and most of these patients (10/25) still used a hearing device at the time of the study. The patient's degree of hearing loss or deprivation decile did not significantly affect whether they had a hearing device, their age of fitting of the device or what device they used. Most patients with UHL that were fitted with a hearing device were fitted after their first birthday (8/12;67%) irrespective of their degree of hearing loss or deprivation, with an average age of first fitting of 3 years 1 month. Patients with UHL who had SLT or developmental impairment were significantly more likely to have trialled a hearing device than those that did not ($p = .047$ and $p = 0.019$ respectively). Five of the seven children with UHL and a developmental impairment also received SLT.

Discussion

Children with UHL *and* a NICU admission were at high risk of multiple congenital anomalies and certain adverse developmental outcomes. Targeted clinical screening—genetic and clinical follow-up is needed at birth for this discrete population.

These data indicate that children with UHL *and* a NICU admission were more likely than their matched counterparts to have congenital anomalies, developmental impairment and SLT. Approximately two thirds (64%) of patients with UHL had multiple congenital anomalies but not all congenital anomalies were detected at discharge (7/25, detected post-discharge). This information suggests better screening for

congenital anomalies would be advantageous in this population. This would be particularly beneficial as a congenital anomaly was found to be positively associated with a developmental impairment and could be used as an indicator for closer developmental follow up in early life.

The prevalence of UHL and NICU admission was 0.17%—lower than previous reports (1.23%–4.6%) (4, 5). Varied methodology could explain this difference, for example babies on the NICU for ≤5 days were excluded in one of these studies. Any baby with suspected sepsis is routinely admitted to the NICU in Nottingham University Hospitals which differs from other regions. Another possibility is despite universal NHSP, UHL is under-reported or undetected in this cohort.

Congenital anomalies

In this study, half (14) of UHL cases were diagnosed with a congenital anomaly at birth. A further quarter (7) patients had congenital anomalies that weren't detected at birth but were detected within the 2 years following. This prevalence is higher than the 29% of congenital anomalies recorded in the literature for the general population of babies with UHL (3); this suggests that NICU-UHL is a red flag for anomalies that don't come to light until post-discharge. This population of babies with UHL and NICU admission were also more likely than their matched counterparts (normal hearing and NICU admission) to have multiple congenital anomalies (64% vs. 8%), further highlighting the importance of screening and detection for NICU-UHL babies.

Abnormal anatomical features

This study supports the already published literature that IEM and craniofacial anomalies are positively associated with UHL and also suggests that NICU admission doesn't increase the likelihood of having these conditions within the UHL cohort, as the prevalence is similar to the UHL well-baby population (3, 9, 10).

Yelverton et al. (2013) showed that 2.4% of babies with UHL (combined well and NICU cohort patients) also had a gastrointestinal problem detected at birth (3), which is 10× lower than the 28% of patients (UHL-NICU) who had a gastrointestinal problem recorded in this study. A NICU admission and diagnosis of UHL could be strongly associated with gastrointestinal problems, or it may be that gastrointestinal issues develop over time. It is possible that we had a lower detection threshold for documenting gastrointestinal problems than Yelverton. Patients with UHL were more likely to have a gastrointestinal problem than their matched counterparts (7/25 vs. 2/25), and most of the associated gastro-intestinal problems (5/7) were detected at birth. Further research needs to be conducted into this association.

In **Table 2** it is apparent that one third, (32%) of the UHL cases were identified as having neurological and spinal issues and a quarter (24%) had vision and eye problems by the age of two. Again we detected a much larger prevalence than reported by Yelverton et al.'s birth population UHL study (4.3%) (3). These data indicate that admission to the NICU and having UHL presents a higher cumulative risk of neurological and/or eye issues. Furthermore, many of these problems may develop after birth; 38% (3/8) of those with spinal and neurological issues and 83% (5/6) of those with vision and eye problems in this cohort developed them in early childhood. This particularly high prevalence of vision and eye problems, 6 times greater than their matched counterparts, suggests a need for closer ophthalmic follow up in this population with UHL.

20% of patients with UHL *and* a NICU admission were found to have cardiac anomalies between 0 and 2 years, which lies between the two values for the general UHL population (41%) and 12% for babies who also have a co-existing JCIH – US risk factor (3) these are all higher than the 8% of matched controls. Many cardiac anomalies that were documented (for example, heart murmur, atrioventricular septal defect), were discounted as they are extremely common in the general NICU population as they are usually not significant, result from prematurity and often resolve with age. Methodological considerations (what is counted as a cardiac anomaly) may account for some of the differences between our data and the current literature.

Behavioural outcomes

This study identified a prevalence of 52% of UHL cases needing SLT, 13 times greater than their matched controls, but in keeping with the majority of the current literature for well babies with UHL and no NICU admission (6, 7). This suggests it is the UHL and not the other underlying health conditions associated with admission to the NICU which increases the likelihood of needing SLT for patients with UHL. This again highlights the need to target NICU graduates with UHL for referral and follow-up.

It was interesting that over a quarter of the UHL cases had a developmental impairment, which was significantly more than their matched controls (28% UHL vs. 0% controls). There is little literature surrounding the association between UHL and developmental impairments. One study found one fifth of children with UHL (both well and NICU populations) were diagnosed with developmental delay (23), which is similar to the findings of this study (28%). Specific developmental follow-up for patients with UHL could help to identify developmental impairment earlier and provide earlier interventions, which could lead to better outcomes and quality of life. Furthermore, all UHL cases in this study with

developmental impairment had multiple congenital anomalies, suggesting that having multiple congenital anomalies could be an indicator for closer developmental monitoring in this population throughout early childhood.

Syndromic and genetic outcomes

In this study, nearly one quarter (6/25) of the UHL cases were diagnosed with a recognised syndrome, 4 of which were found to be associated with hearing loss. It is interesting that these were not the syndromes most associated with UHL (for example, Waardenburg Syndrome), suggesting that perhaps there is a different subset of syndromes yet to be identified, that are more likely to be associated with UHL *and* NICU admission. One study found that 1 in 110 patients (0.9%) with UHL had a syndrome that was associated with hearing loss, which is much lower than the 16% found in this study (3). This suggests that infants with a syndrome and UHL are more likely to also have a NICU admission.

Studies have revealed the percentage of UHL associate with a family history is approximately 3.7%–13% (9, 24, 25), which is similar to the number of UHL cases with genetic variants in this study (16%). There is little to no research on the specific genetic variants associated with UHL. Furthermore, three of the four patients with a genetic variant also had a diagnosed syndrome, suggesting that the syndromes may be linked to specific genetic variants. A national study investigating genetics and UHL cases would need to be conducted to confirm this. Currently genetic screening is not recommended or funded for infants diagnosed with UHL in the UK.

Hearing outcomes

In this study, 40% of patients with UHL were currently using a hearing device, which is similar to pre-existing literature (6). Patients with UHL that received SLT were significantly more likely to have trialled a hearing device than those that had not received SLT. A study into children with bilateral hearing loss (BHL) by Tomblin et al. (2015) identified that hearing aids can improve language outcomes over time in these children (26). There is sparse literature available to indicate that children with UHL that are struggling with their speech and language development may benefit more from a hearing device or a trial of a device in their early years. There is some evidence to suggest that wearing a hearing device can improve quality of life, especially in those suffering with speech and language or academic and behavioural issues, whereas other studies have found that hearing devices may not be beneficial for younger children with UHL and do not improve speech recognition (27–30). Cochlear implantation is not currently funded for children with UHL in the UK, recent preliminary studies have shown improvement in some areas for children with UHL following implantation (31). More research is needed in this area. Furthermore, the majority (6/7) of patients with UHL that had a developmental impairment had trialled a hearing device, which may be due to the positive association between developmental delay and SLT. Furthermore, as there are no current UK NHS guidelines for the management of UHL in children, audiologists may use SLT or diagnosis of a developmental impairment to guide them on management with a device, as well as use this information to suggest to parents that their child trial a device. However, by the time the child needs SLT this may be too late; earlier device trials could be important during the critical period for language acquisition. Parents may be more willing to trial a device if they see the developmental effects UHL has on their child. Out of the home, eg in nursery and playgroups it could be vital for children with UHL (particularly NICU graduates) to use a hearing device and employ a remote microphone system as it is known that deciphering speech in noise is particularly difficult for people with UHL. Currently there is not consistent funding for remote microphone systems and SLT for children with UHL.

Future work

Not all patients with UHL *and* a NICU admission go onto develop congenital anomalies or abnormal anatomical or behavioural outcomes; further research is required into why this is the case and are their neuroprotective factors which help pre or postnatally (eg maternal magnesium or prenatal steroids). It is possible that there is a subset of patients in this cohort that have certain risk factors that increase their chance of having adverse developmental outcomes. For example, further research into the association between multiple congenital anomalies and developmental outcomes could be conducted.

Conclusion

Research into the developmental outcomes of patients with UHL has mainly focused on the general population, not the cohort that has also been admitted to the NICU. This longitudinal study identified many adverse outcomes in this unique population, which is a step towards identifying a constellation of features associated with UHL in babies who have been admitted to the NICU. This study can contribute towards developing guidelines surrounding the screening, follow up and management of these patients, which would benefit both clinicians and patients.

Data availability statement

The data analyzed in this study is subject to the following licenses/restrictions: Currently datasets are not available to researchers outside of NUH Nottingham. Datasets available on application to NEAT database team. Requests to access these datasets should be directed to dulip.jayasinghe@nuh.nhs.uk.

Ethics statement

The NEAT database has been approved by South Central - Berkshire Research Ethics Committee (REC 22/SC/0337; IRAS 292263). Written informed consent from the participants' legal guardian/next of kin was not required to participate in this study in accordance with the national legislation and the institutional requirements.

Author contributions

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Conflict of interest

The authors declare that the research was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

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Supplementary material

The Supplementary Material for this article can be found online at: https://www.frontiersin.org/articles/10.3389/fped.2022.1068884/full#supplementary-material.

References

1. Berninger E, Westling B. Outcome of a universal newborn hearing-screening programme based on multiple transient-evoked otoacoustic emissions and clinical brainstem response audiometry. *Acta Otolaryngol.* (2011) 131(7):728–39. doi: 10.3109/00016489.2011.554440

2. Prieve B, Dalzall L, Berg A, Bradley M, Cacace A, Campbell D, et al. The New York State universal newborn hearing screening demonstration project: outpatient outcome measures. *Ear Hear.* (2000) 21:104–17. doi: 10.1097/00003446-200004000-00005

3. Yelverton JC, Dominguez LM, Chapman DA, Wang S, Pandya A, Dodson KM. Risk factors associated with unilateral hearing loss. *JAMA Otolaryngol Head Neck Surg.* (2013) 139(1):59–63. doi: 10.1001/jamaoto.2013.1097

4. Chang J, Oh SH, Park SK. Comparison of newborn hearing screening results between well babies and neonates admitted to the neonatal intensive care unit for more than 5 days: analysis based on the national database in Korea for 9 years. *PLoS One.* (2020) 15(6):e0235019. doi: 10.1371/journal.pone.0235019

5. Stadio AD, Molini E, Gambacorta V, Giommetti G, Volpe AD, Ralli M, et al. Sensorineural hearing loss in newborns hospitalized in neonatal intensive care unit: an observational study. *Int Tinnitus J.* (2019) 23(1):31–6. doi: 10.5935/0946-5448.20190006

6. Lieu JE, Tye-Murray N, Fu Q. Longitudinal study of children with unilateral hearing loss. *Laryngoscope.* (2012) 122(9):2088–95. doi: 10.1002/lary.23454

7. Rachakonda T, Shimony JS, Coalson RS, Lieu JEC. Diffusion tensor imaging in children with unilateral hearing loss: a pilot study. *Front Syst Neurosci.* (2014) 8:87. doi: 10.3389/fnsys.2014.00087

8. Klee TM, Davis-Dansky E. A comparison of unilaterally hearing-impaired children and normal-hearing children on a battery of standardized language tests. *Ear Hear.* (1986) 7(1):27–37. doi: 10.1097/00003446-198602000-00006

9. Masuda S, Usui S. Comparison of the prevalence and features of inner ear malformations in congenital unilateral and bilateral hearing loss. *Int J Pediatr Otorhinolaryngol.* (2019) 125:92–7. doi: 10.1016/j.ijporl.2019.06.028

10. Paul A, Marlin S, Parodi M, Rouillon I, Guerlain J, Pingault V, et al. Unilateral sensorineural hearing loss: medical context and etiology. *Audiol Neurootol.* (2017) 22(2):83–8. doi: 10.1159/000474928

11. Cone-Wesson B, Vohr BR, Sininger YS, Widen JE, Folsom RC, Gorga MP, et al. Identification of neonatal hearing impairment: infants with hearing loss. *Ear Hear*. (2000) 21(5):488–507. doi: 10.1097/00003446-200010000-00012

12. Masuda S, Usui S, Matsunaga T. High prevalence of inner-ear and/or internal auditory canal malformations in children with unilateral sensorineural hearing loss. *Int J Pediatr Otorhinolaryngol*. (2013) 77(2):228–32. doi: 10.1016/j.ijporl.2012.11.001

13. Schiariti V, Klassen AF, Houbé JS, Synnes A, Lisonkova S, Lee SK. Perinatal characteristics and parents' perspective of health status of NICU graduates born at term. *J Perinatol*. (2008) 28(5):368–76. doi: 10.1038/jp.2008.9

14. Saigal S, Doyle LW. An overview of mortality and sequelae of preterm birth from infancy to adulthood. *Lancet*. (2008) 371(9608):261–9. doi: 10.1016/S0140-6736(08)60136-1

15. Newnam K, Parrott J. The NICU graduate: implications for pediatric primary care. *Newborn Infant Nurs Rev*. (2013) 13(2):94–100. doi: 10.1053/j.nainr.2013.03.005

16. Ministry of Housing CaLG. English indices of deprivation 2019 Postcode Lookup 2019. Available from: http://imd-by-postcode.opendatacommunities.org/imd/2019.

17. Ministry of Housing CaLG. The English Indices of Deprivation 2019 Frequently Asked Questions (FAQs) 2019. Available from: https://assets.publishing.service.gov.uk/government/uploads/system/uploads/attachment_data/file/853811/IoD2019_FAQ_v4.pdf.

18. Audiology BSo. Pure-tone air-conduction and boneconduction threshold audiometry with and without masking. (2018) updated August 2018. Available from: https://www.thebsa.org.uk/wp-content/uploads/2018/11/OD104-32-Recommended-Procedure-Pure-Tone-Audiometry-August-2018-FINAL.pdf

19. Strömland K, Miller M, Sjögreen L, Johansson M, Joelsson B-ME, Billstedt E, et al. Oculo-auriculo-vertebral spectrum: associated anomalies, functional deficits and possible developmental risk factors. *Am J Med Genet A*. (2007) 143A (12):1317–25. doi: 10.1002/ajmg.a.31769

20. Arcand P, Desrosiers M, Dubé J, Abela A. The large vestibular aqueduct syndrome and sensorineural hearing loss in the pediatric population. *J Otolaryngol*. (1991) 20(4):247–50. PMID: 1920576

21. McGaughran JM, Kuna P, Das V. Audiological abnormalities in the Klippel-Feil syndrome. *Arch Dis Child*. (1998) 79(4):352. doi: 10.1136/adc.79.4.352

22. Schick B, Brors D, Prescher A, Draf W. Conductive hearing loss in Beckwith–Wiedemann syndrome. *Int J Pediatr Otorhinolaryngol*. (1999) 48 (2):175–9. doi: 10.1016/S0165-5876(99)00015-4

23. Haffey T, Fowler N, Anne S. Evaluation of unilateral sensorineural hearing loss in the pediatric patient. *Int J Pediatr Otorhinolaryngol*. (2013) 77(6):955–8. doi: 10.1016/j.ijporl.2013.03.015

24. Friedman AB, Guillory R, Ramakrishnaiah RH, Frank R, Gluth MB, Richter GT, et al. Risk analysis of unilateral severe-to-profound sensorineural hearing loss in children. *Int J Pediatr Otorhinolaryngol*. (2013) 77(7):1128–31. doi: 10.1016/j.ijporl.2013.04.016

25. Niu K, Brandström A, Skenbäck S, Duan M, Uhlén I. Risk factors and etiology of childhood hearing loss: a cohort review of 296 subjects. *Acta Otolaryngol*. (2020) 140(8):660–6. doi: 10.1080/00016489.2020.1757753

26. Tomblin JB, Harrison M, Ambrose SE, Walker EA, Oleson JJ, Moeller MP. Language outcomes in young children with mild to severe hearing loss. *Ear Hear*. (2015) 36(Suppl 1(0 1)):76S–91S. doi: 10.1097/AUD.0000000000000219

27. Krishnan LA, Van Hyfte S. Management of unilateral hearing loss. *Int J Pediatr Otorhinolaryngol*. (2016) 88:63–73. doi: 10.1016/j.ijporl.2016.06.048

28. McKay S. To aid or not to aid: children with unilateral hearing loss Audiology Online [updated 07/22/2002]. Available from: https://www.audiologyonline.com/articles/to-aid-or-not-children-1167

29. Briggs L, Davidson L, Lieu J. Outcomes of conventional amplification for pediatric unilateral hearing loss. *Ann Otol Rhinol Laryngol*. (2011) 120:448–54. doi: 10.1177/000348941112000705

30. Banga R, Doshi J, Child A, Pendleton E, Reid A, McDermott AL. Bone-anchored hearing devices in children with unilateral conductive hearing loss: a patient-carer perspective. *Ann Otol Rhinol Laryngol*. (2013) 122(9):582–7. doi: 10.1177/000348941312200908

31. Brown KD, Dillon MT, Park LR. Benefits of cochlear implantation in childhood unilateral hearing loss (CUHL trial). *Laryngoscope*. (2022) 132(Suppl 6(Suppl 6)):S1–S18. doi: 10.1002/lary.29853

32. England PH. Patient Journey from screen to referral GOV UK2020 [updated 14/12/2020]. Available from: https://www.gov.uk/government/publications/newborn-hearing-screening-programme-nhsp-operational-guidance/6-patient-journey-from-screen-to-referral

33. England PH. Newborn hearing screening programme (NHSP): care pathways for babies in neonatal intensive care units (NICU) GOV UK2020 [updated 29/10/2020]. Available from: https://www.gov.uk/government/publications/newborn-hearing-screening-care-pathways/newborn-hearing-screening-programme-nhsp-care-pathways-for-babies-in-neonatal-intensive-care-units-nicu

34. England PH. Newborn hearing screening programme (NHSP): care pathways for well babies GOV UK2020 [updated 29/10/2020]. Available from: https://www.gov.uk/government/publications/newborn-hearing-screening-care-pathways/newborn-hearing-screening-programme-nhsp-care-pathways-for-well-babies

35. England PH. Guidelines for surveillance and audiological referral for infants and children following newborn hearing screen GOV UK2019 [updated 19/07/2019]. Available from: https://www.gov.uk/government/publications/surveillance-and-audiological-referral-guidelines/guidelines-for-surveillance-and-audiological-referral-for-infants-and-children-following-newborn-hearing-screen

36. Approach to the Child with Birth Defects. (2001). Available from: http://www.med.umich.edu/lrc/coursepages/m1/humangenetics/HG501-Petty-Lec16.html

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Kerttu Huttunen,
University of Oulu, Finland

REVIEWED BY
Kevin Munro,
The University of Manchester, United Kingdom
Lynn K. Perry,
University of Miami, United States

*CORRESPONDENCE
Bethany Adams
bethany.adams@nottingham.ac.uk

†These authors have contributed equally to this work and share last authorship

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Exploring listening-related fatigue in children with and without hearing loss using self-report and parent-proxy measures

Bethany Adams[1,2]*, Sally K. Thornton[1,2], Graham Naylor[1,2], Ruth V. Spriggs[1,2], Ian M. Wiggins[1,2†] and Padraig T. Kitterick[3†]

[1]National Institute for Health and Care Research (NIHR) Nottingham Biomedical Research Centre, Nottingham, United Kingdom, [2]Hearing Sciences, Mental Health and Clinical Neurosciences, School of Medicine, University of Nottingham, Nottingham, United Kingdom, [3]National Acoustic Laboratories, Australian Hearing Hub, Macquarie University, Sydney, NSW, Australia

Children with hearing loss appear to experience greater fatigue than children with normal hearing (CNH). Listening-related fatigue is often associated with an increase in effortful listening or difficulty in listening situations. This has been observed in children with bilateral hearing loss (CBHL) and, more recently, in children with unilateral hearing loss (CUHL). Available tools for measuring fatigue in children include general fatigue questionnaires such as the child self-report and parent-proxy versions of the PedsQL™-Multidimensional Fatigue Scale (MFS) and the PROMIS Fatigue Scale. Recently, the Vanderbilt Fatigue Scale (VFS-C: child self-report; VFS-*P*: parent-proxy report) was introduced with a specific focus on listening-related fatigue. The aims of this study were to compare fatigue levels experienced by CNH, CUHL and CBHL using both generic and listening-specific fatigue measures and compare outcomes from the child self-report and parent-proxy reports. Eighty children aged 6–16 years (32 CNH, 19 CUHL, 29 CBHL), and ninety-nine parents/guardians (39 parents to CNH, 23 parents to CUHL, 37 parents to CBHL), completed the above fatigue questionnaires online. Kruskal-Wallis H tests were performed to compare fatigue levels between the CNH, CUHL and CBHL. To determine the agreement between parent-proxy and child self-report measures, Bland-Altman 95% limits of agreement were performed. All child self-report fatigue measures indicated that CBHL experience greater fatigue than CNH. Only the listening-specific tool (VFS-C) was sufficiently able to show greater fatigue in CUHL than in CNH. Similarly, all parent-proxy measures of fatigue indicated that CBHL experience significantly greater fatigue than CNH. The VFS-*P* and the PROMIS Fatigue Parent-Proxy also showed greater fatigue in CUHL than in CNH. Agreement between the parent-proxy and child self-report measures were found within the PedsQL-MFS and the PROMIS Fatigue Scale. Our results suggest that CBHL experience greater levels of daily-life fatigue compared to CNH. CUHL also appear to experience more fatigue than CNH, and listening-specific measures of fatigue may be better able to detect this effect. Further research is needed to understand the bases of fatigue in these populations and to clarify whether fatigue experienced by CBHL and CUHL is comparable in nature and degree.

KEYWORDS

fatigue, children, unilateral hearing loss, self-report, hearing loss, quality of life

1. Introduction

There is no universally accepted definition of fatigue, though it is generally described as an overall feeling of tiredness, lack of energy or vigour, or decreased motivation (1). Fatigue often presents after insufficient sleep or increased mental or physical exertion, but usually resolves after rest or mental stillness.

It has been widely reported that both adults (2, 3) and children (4–9) with hearing loss experience an increase in daily life fatigue, compared to their normal hearing peers. Individuals with a hearing impairment may have to allocate more cognitive resources to effortful listening than those with normal hearing (10), which can lead to fatigue. This fatigue, being a result of listening, is referred to as 'listening-related fatigue'. Children with hearing loss (CHL) have to exert greater listening effort than children with normal hearing (CNH), potentially leading to poorer sentence recognition (11) and lower processing speed (12) in listening tasks.

Moreover, children are more affected by unfavourable noise conditions than adults (13) as classroom teaching often takes place in a reverberant and noisy environment (14, 15), requiring considerable listening effort (16). The available literature suggests that for CHL it can be mentally exhausting spending an entire day listening to their teacher's speech, against excessive classroom noise levels. It is also more difficult for children with unilateral hearing loss (CUHL) to localise sounds, compared to CNH (17).

Children with hearing loss have reported fatigue in qualitative studies. Davis et al. (18) found that fatigue in CHL is expressed in many ways, such as difficulty concentrating, lack of motivation, and physical tiredness. Parents of CHL and clinicians who manage CHL have also noted that children with varying degrees of hearing loss experience fatigue, especially following sustained listening demands at school Bess et al. (4). Although the consequences of fatigue have not yet been measured in children with hearing loss, it has been reported that children with fatigue due to a chronic health disorder have poor academic performance, decreased motivation, increased distractibility, poorer social functioning, and more depressive symptoms (19–21). If such negative consequences also are present in CHL, then there is a pressing need for further elucidation.

Currently, fatigue is most often measured *via* self-report. This is often in the form of questionnaires but can also be *via* qualitative interviews. Often, subjective measures of fatigue are multidimensional, which capture health or activities commonly associated with fatigue, such as sleep, cognition, and social functioning (22).

The well-known and widely used Pediatric Quality of Life Multidimensional Fatigue Scale (PedsQL-MFS 23, 24); has been used in many studies to quantify fatigue in CHL (7, 9, 25, 26). This questionnaire assesses three domains of fatigue, namely *General Fatigue*, *Sleep/Rest Fatigue* and *Cognitive Fatigue*. A *Total Fatigue* score is calculated by summing the above three domains.

Hornsby et al. (2014) were the first to measure fatigue in CHL using self-report questionnaires. The PedsQL-MFS was used to measure fatigue in ten school aged children with bilateral hearing loss (CBHL), and ten aged-matched CNH. They found higher levels of fatigue in CHL compared to their CNH peers in all domains. These findings were confirmed in a larger study by the same group (9) in which parent-proxy reports of fatigue were also collected. Here, they compared fatigue ratings between 60 CBHL and 43 CNH. In this study, CBHL rated higher levels of fatigue across all domains, which was significant in the *Cognitive* and *Total* domains. They showed that parent ratings of fatigue were significantly different to both ratings by CHL and CNH in *Cognitive*, *Sleep/Rest* and *Total* domains of fatigue (9). In the parent-proxy report, parents often report higher than their children's self-report (higher scores represent lower fatigue), especially in the *Sleep/Rest* domain, suggesting that parents often underestimate their child's fatigue. However, it is important to note that this standardised tool was designed to measure fatigue in children with chronic health disorders, such as rheumatoid arthritis. It was not designed to measure fatigue in children with hearing loss, and so may not be considered the best tool to measure listening-related fatigue. A new, recently validated, measure for fatigue in CHL is the pediatric version of the Vanderbilt Fatigue Scale (VFS-Peds 27);. This stands out from the PedsQL-MFS, as it is the only tool which has been specifically designed to measure listening-related fatigue in children.

Until recently, researchers measuring fatigue in CHL did not include CUHL. Unilateral hearing loss (UHL) has historically been regarded as being a minor inconvenience, and CUHL are generally offered less support when compared to CBHL (28). However, increasing evidence has shown that having UHL can affect many aspects of a child's development in ways that can be considerably impactful; socially, educationally, and behaviourally (29–32).

A recent qualitative study provided supporting narrative evidence for the incidence of fatigue in CUHL (4). Focus groups with parents of children with UHL included the observation that "My daughter is exhausted most days after school or when she has to listen for a long time". One audiologist observed that "Our kids with UHL are similar to children with mild to moderate hearing losses – they require auditory breaks throughout the day and struggle more academically than one would expect given their hearing loss." Hornsby et al. (27) utilised the Vanderbilt Fatigue Scale (VFS-C: child self-report; VFS-*P*: parent-proxy report) to measure fatigue in CNH, CUHL, and CBHL, with self-report from the children and proxy reports from their parents. As expected, CBHL rated significantly more fatigue than CNH. Though CUHL rated a greater level of fatigue than CNH, this was not significant. Parents of CNH, CUHL and CBHL (PNH, PUHL, PBHL, respectively) completed the parent-proxy report (VFS-*P*). Interestingly, both PUHL and PBHL had very similar fatigue ratings, which were both significantly higher than PNH. PBHL or PUHL were approximately four times more likely to report that their child experiences moderate-to-severe fatigue than PNH (4). This exploratory study was the first to include CUHL and quantitatively show that they experience similar levels of fatigue to CBHL.

Two more recent studies have measured fatigue in CUHL using the highly validated PedsQL-MFS. Sindhar et al. (26) compared fatigue levels between CUHL and CBHL aged 5–18 years (mean age 10.7 years), children with normal hearing (CNH) children (obtained from 23) using both child self-report and parent-proxy versions of the PedsQL-MFS. In the child self-report version, CBHL reported significantly greater fatigue than CNH in the *Total*, *General* and *Cognitive* fatigue domains, but not the *Sleep/Rest* domain. Children with UHL also reported greater levels of fatigue than CNH, though there were no significant differences throughout domains. Conversely, in the parent-proxy reported fatigue scores,

both PUHL and PBHL rated significantly greater levels of fatigue than CNH across all domains and there were no significant differences in parent-proxy reports between CUHL and CBHL. Similarly to reports from the VFS in Bess et al. (2020), parents of children with hearing loss who completed the parent-proxy PedsQL-MFS (26) reported much higher levels of fatigue than their children's self-report ratings, with a much greater distinction from the CNH. This contrasts to Hornsby et al. (9), where parents underestimated their children's fatigue levels. Carpenter et al. (25) compared fatigue levels in children aged 5–18 years (mean age 10.44 years) with unilateral sensorineural hearing loss (USNHL) and unilateral conductive hearing loss (UCHL) to CNH (obtained from 23), again using child self-report and parent-proxy versions of the PedsQL-MFS. They found that children with USNHL reported significantly greater levels of fatigue than CNH children across all fatigue domains other than the *Sleep/Rest* domain. Children with UCHL reported similar levels of fatigue to CNH children. Conversely, in the parent-proxy reported outcomes, parents of children with both USNHL and UCHL rated their children's fatigue at similar levels (though parents of children with USNHL rated the highest fatigue levels), both of which were significantly greater than the CNH scores, across all four fatigue domains. Carpenter et al. (25) suggested that the lack of perceived fatigue reported by children with UCHL, compared to children with USNHL, was because the former have better habilitation options, such as bone conducting hearing aids. Carpenter et al. also noted that in this study, children with USNHL had a greater degree of hearing loss than those in the UCHL group.

In most current literature, both the non-hearing specific PedsQL-MFS and the hearing specific VFS have been used to measure fatigue in CUHL and CBHL. These questionnaires have not yet been directly compared to each other to determine if they are in fact measuring the same type of fatigue. In this study, we compared fatigue levels between CNH, CUHL and CBHL using the PedsQL-MFS and the newly validated VFS. We also introduced the PROMIS Fatigue Short Form (33), a highly standardised measure of fatigue in children, that has not previously been used to quantify fatigue in children with hearing loss.

This study aims to explore the fatigue levels in CNH, CUHL and CBHL using generic and listening-specific fatigue questionnaires. We also aim to explore whether parent-proxy reporting is a reliable measure of fatigue in children with hearing loss.

2. Methods

Children aged 6–16 years with unilateral hearing loss (UHL), bilateral hearing loss (BHL) or normal hearing (NH), and their parents/guardians were invited to take part in an online questionnaire. The online questionnaire was advertised *via* NHS audiology clinics, schools, and online *via* social media. Parents/guardians who were interested in taking part were first invited to a webpage, which contained the participant information sheet and parent consent form. Parents/guardians were encouraged to read through this page with their children before signing up to take part. Once signed up, parents/guardians were sent two password protected study links *via* email. One for the parent/guardian to complete and one for the child to complete. For children aged 6–7, parents were advised to assist in the completion of questionnaires by reading out each individual statement and recording responses if necessary. For children aged 16 years, the study link was sent directly to them.

The study was created and hosted using Online Surveys (https://www.onlinesurveys.ac.uk/) and distributed electronically with personalised links *via* email. Before launch, the study was piloted by pediatric audiologists, clinical hearing scientists and their children (one CUHL and three CNH). For children aged 6–15 years, the study link first opened up to the child participant information sheet and consent and assent page. Parents were asked to electronically sign the consent form, and children were asked to sign the assent form before they could move onto the series of questionnaires. Children aged 16 were also presented with a participant information sheet and asked to sign the consent form before accessing the questionnaires.

2.1. Inclusion criteria

Children were eligible if they were aged 6–16 years with a permanent unilateral or bilateral hearing loss or normal hearing. Hearing loss was confirmed *via* parental self-report of diagnosis from a clinical audiologist. The study was entirely virtual, so formal hearing assessments were not possible. The following eligibility criteria were applied:

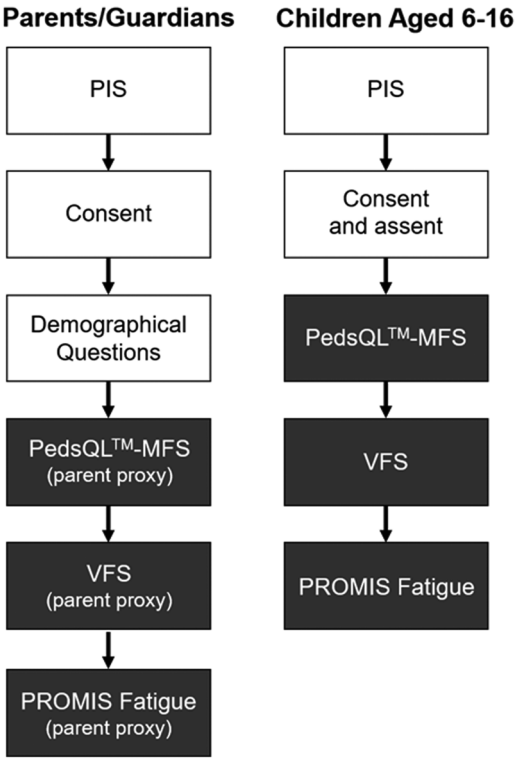

FIGURE 1
Flowchart of study format for parent/guardian and child participants. For child self-report section, parent consented for children aged 6-15 years old, and children assented. Children who were aged 16 provided their own consent. PIS = Patient Information Sheet. Shaded boxes are fatigue questionnaires.

2.1.1. Children

- Aged 6–16
- Has received a diagnosis of permanent unilateral hearing loss or permanent bilateral hearing loss
- Has normal or corrected-to-normal vision confirmed *via* parental self-report
- Uses English in home environment or at school
- Able and willing to give informed consent (16 years or older), or to give assent together with consent from a parent/guardian
- Has access to a computer/smart phone or tablet with internet.

2.1.2. Parents/guardians

- A parent/guardian to a child who meets the above criteria
- Able to speak fluent English
- Has access to a computer with internet
- Able and willing to give informed consent

2.2. Study format

The study format is outlined in **Figure 1**.

2.3. Recruitment

Participants were recruited through advertisement on online forums, in schools, and in audiology clinics.

2.4. Ethics

This study was approved by the Health Research Authority (HRA), Wales Research Ethics Committee (20/WA/0233) and the Nottingham Research Committee (52–0720).

2.5. Questionnaires

2.5.1. PedsQL-MFS

The Pediatric Quality of Life Inventory Multidimensional Fatigue Scale (PedsQL-MFS; 23, 24) is a standardised self-report instrument designed to measure fatigue in paediatric patients. It consists of three separate fatigue domains: *General Fatigue* (6 items), *Sleep/Rest Fatigue* (6 items), and *Cognitive Fatigue* (6 items), and also gives a score of *Total Fatigue*. It includes both child and parent-proxy reports, allowing both perspectives. The PedsQL-MFS has been used by children with a wide variety of health conditions, including cancer (24) and rheumatoid arthritis, and has been shown to have good internal consistency, reliability and validity (23). For ease of use in younger children (aged 5–7), the PedsQL-MFS has simplified terminology, and answers are anchored to a happy/sad faces scale (23).

2.5.2. Vanderbilt fatigue scale

The Vanderbilt Fatigue Scale (VFS; 4, 27) is a self-report tool designed to measure listening-related fatigue in children. All items in the VFS are directly related to hearing or listening, for example "My brain gets tired after listening all day" (27). The VFS includes three versions: child self-report (VFS-C), parent-proxy (VFS-*P*) and teacher-proxy (VFS-T).

2.5.3. PROMIS paediatric fatigue short form

The Patient-Reported Outcomes Measurement Information System (PROMIS) paediatric self-report (33) and parent-proxy (34) fatigue short-forms (v10a) are validated questionnaires used to assess fatigue in multiple populations. An example item from this short form includes "I was too tired to enjoy the things I like to do". Although these tools have not yet been used to measure fatigue in children with hearing loss, they have been shown to have a high content validity (35).

2.6. Scoring and statistics

The PedsQL-MFS responses were summed to produce a score for each domain (*General Fatigue*, *Sleep/Rest Fatigue* and *Cognitive Fatigue*). These scores were then combined to produce the *Total Fatigue* score. A lower score indicates a greater level of fatigue. The VFS was scored using Item Response Theory (IRT) using R Studio, as outlined by Hornsby et al. (27). Higher IRT scores indicated a greater level of fatigue. The PROMIS Fatigue Short Form raw scores were converted to T-Scores [https://www.assessmentcenter.net/]. T Scores are standard scores with a mean of 50 and a standard deviation of 10 in a reference population (36). Higher T Scores indicate a greater level of fatigue, and a score above 50 indicates greater fatigue than the population average.

Descriptive statistical analysis and normality tests were conducted using IBM SPSS Statistics for Windows, version 28.0 (IBM Corp., Armonk, NY). Data were not normally distributed. Normality of response measures was assessed visually with histograms and with the Shapiro-Wilk test. Kruskal-Wallis H tests, with Mann-Whitney post-hoc tests, were performed to compare fatigue levels between the CNH, CUHL and CBHL.

To determine the agreement between parent-proxy and child self-report measures of fatigue, Bland-Altman limits of agreement (37) were performed with exact 95% CI for the limits of agreement, and supplemented by Pearson correlation coefficients. Agreement calculations between the VFS-*P* and VFS-C were not possible due to different outcomes between parents and children (VFS-C gives one *Total Fatigue* score, whereas VFS-*P* gives separate scores of Mental Fatigue and Physical Fatigue).

3. Results

The study was open between August 2020 and September 2021. Ninety-nine parents/guardians completed the study, out of which 37 were parents to CBHL, 23 were parents to CUHL and 39 were parents to children with NH. Eighty children between the ages of 6 to 16 completed the questionnaires (mean age = 10.25; SD = 3.02), of which 29 children had BHL, 19 had UHL and 32 had NH. In 77 cases, both the parent and child completed the questionnaire

(26 BHL, 19 UHL and 32 NH). Participant demographics can be found in Table 1.

3.1. PedsQL-MFS

In the PedsQL-MFS, lower scores indicate a greater level of fatigue. Median scores, interquartile ranges (IQR), and minimum and maximum values for the child self-reported and parent-proxy PedsQL-MFS are reported in Table 2. Figure 2 displays the child self-reported medians and IQRs for CNH, CUHL and CBHL and parent-proxy reported means for CNH, CUHL and CBHL. Lower scores on the PedsQL-MFS indicate a higher level of fatigue.

CUHL and CBHL reported greater levels of fatigue than CNH across all domains. CUHL reported similar levels of fatigue to CBHL. A Kruskal-Wallis test was conducted to determine if there were differences in fatigue scores between groups (NH, UHL and BHL). No significant differences in fatigue scores were found in the *Total* ($H(2) = 5.235$, $p = 0.07$) or *Sleep/Rest* ($H(2) = 0.688$, $p = 0.709$) domains. Significant differences in fatigue scores between groups were found within the *General* ($H(2) = 7.591$, $p = 0.022$) and *Cognitive* ($H(2) = 8.141$, $p = 0.017$) domains. Subsequently, Mann-Whitney pairwise comparisons were performed with Bonferroni correction for multiple comparisons in the *General* and *Cognitive* fatigue domains. This *post hoc* analysis revealed statistically significant differences between CBHL and CNH in the *General* and *Cognitive* domains. CBHL reported statistically significantly greater levels of fatigue than CNH in the *General* ($p = 0.031$), and borderline significance for *Cognitive* ($p = 0.05$), domains of fatigue.

TABLE 1 Demographics include 99 cases where the parent filled out the questionnaires. HA; hearing aids, CI; cochlear implants.

	Normal Hearing $N = 39$	Unilateral Hearing Loss $N = 23$	Bilateral Hearing Loss $N = 37$
Gender of child			
Male	**25**	**11**	**21**
Female	**14**	**12**	**16**
Age of child			
6-7	5	6	11
8-12	19	13	18
13-15	13	4	5
16	2	0	3
Hearing Status		**23**	**37**
Permanent Hearing Loss		**21**	**33**
Fluctuating Hearing Loss		**2**	**4**
Hearing Devices			
HA	0	**14**	**23**
CI	0	**1**	**11**
FM System	0	**3**	**22**

CUHL reported significantly greater levels of fatigue than CNH in the *Cognitive* domain only, and this difference was marginally significant ($p = 0.047$).

Both groups of parents (CUHL and CBHL) reported greater fatigue for their children (indicated by lower PedsQL-MFS scores) than parents of CNH across all domains of fatigue (Table 2; Figure 2).

In the parent-proxy PedsQL-MFS, significant differences between child groups were found in all domains (*General*: $H(2) = 23.926$, $p < 0.001$; Sleep/Rest: $H(2) = 10.508$, $p = 0.005$; *Cognitive*: $H(2) = 23.008$, $p < 0.001$; *Total* $H(2) = 23.828$, $p < 0.001$). Parents of CBHL reported significantly greater levels of fatigue than CNH in the *Total* ($p < 0.001$), *General* ($p < 0.001$), *Sleep/Rest* ($p = 0.004$) and *Cognitive* ($p < 0.001$) domains. Additionally, parents of CUHL also rated significantly greater levels of fatigue than CNH across the *Total* ($p = 0.017$), *General* ($p = 0.012$) and *Cognitive* ($p = 0.025$) domains. There were no significant differences in parent-proxy ratings of fatigue between CUHL and CBHL.

3.1.1. Parent-Proxy vs. Child self-report

For parent-proxy vs. child self-report comparisons, $N = 77$ for both groups. There were no significant differences in scores between child self-report and parent-proxy reports in the *Total* and *General* domains of the PedsQL-MFS. However, score differences between child self-report and parent-proxy reports of the *Sleep/Rest* and *Cognitive* domains were significant.

3.1.1.1. Total

Agreement by Bland-Altman plots are shown in Supplementary Figure S1A with a mean difference of PedsQL-MFS *Total* score of −5.63 to 2.13; the limits of agreement were −35.21 to 31.71. The interclass correlation coefficient was 0.847 (95% CI: 0.759–0.903,

TABLE 2 Medians (interquartile range) and [min, max] of child self-report and parent-proxy report scores from the pedsQL-MFS.

	NH	UHL	BHL
Child Self-Report	*N = 32*	*N = 19*	*N = 29*
Total	76.4 (36.5) [20.8, 100]	61.1 (33.3) [5.6, 98.6]	63.9 (23.6) [8.3, 91.7]
General	83.3 (36.5) [8.3, 100]	66.7 (41.7) [0.0, 100]	70.8 (35.4) [8.3, 95.8]
Sleep/Rest	66.7 (40.6) [25.0, 100]	62.5 (33.3) [0.0, 100]	66.7 (37.5) [16.7, 91.7]
Cognitive	81.3 (41.7) [0.0, 100]	54.2 (41.7) [0.0, 95.8]	50.00 (43.8) [0.0, 95.8]
Parent-Proxy Report	*N = 39*	*N = 23*	*N = 37*
Total	80.56 (2.2) [43.0, 100]	62.5 (34.7) [9.7, 97.2]	56.9 (38.9) [8.3, 98.6]
General	83.3 (25.0) [54.2, 100]	58.3 (50.0) [12.5, 100]	58.3 (41.7) [0.0, 95.3]
Sleep/Rest	83.3 (16.7) [37.5, 100]	75.0 (33.3) [8.3, 100]	62.5 (39.6) [8.3, 100]
Cognitive	79.2 (45.8) [0.0, 100]	54.2 (45.8) [0.0, 100]	37.5 (50.0) [0.0, 100]

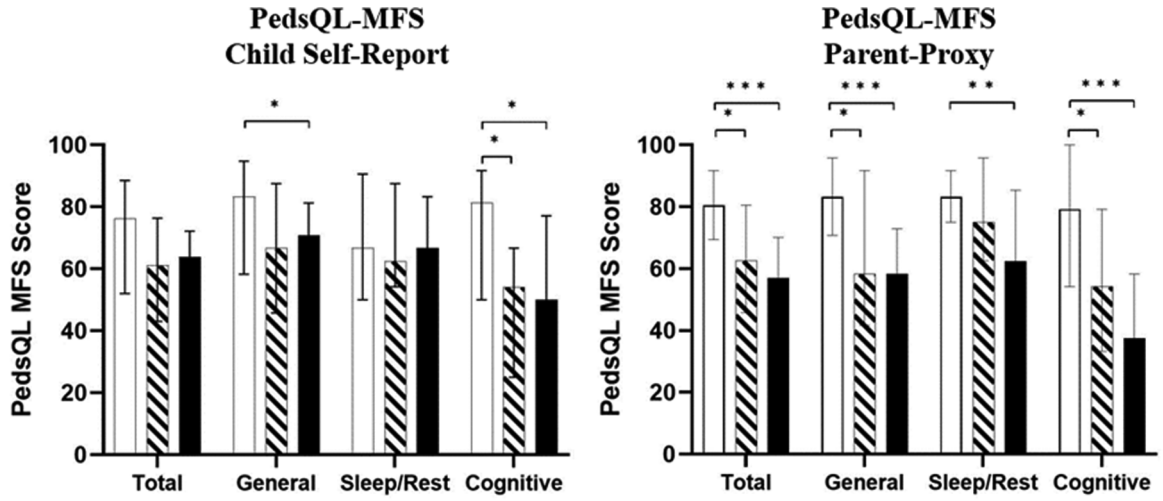

FIGURE 2
Median pediatric quality of life inventory multidimensional fatigue scale (pedsQL-MFS) scores for child self-report (left) and parent-proxy (right) respondents. Error bars shown represent medians ± IQRs. Lower values indicate greater fatigue. CNH; white bars, CUHL; oblique stripes, CBHL; black bars respectively. Asterisks show significant differences between groups (*$p < 0.05$, **$p < 0.01$, ***$p < 0.001$).

$p < 0.01$). Regression analysis between the score means and the difference between scores were not significant (-0.52, $p = 0.596$), meaning there is no proportional bias.

3.1.1.2. General

Agreement by Bland-Altman plots are shown in **Supplementary Figure S1B** with a mean difference of PedsQL-MFS *General* score of −4.61 to 5.26; the limits of agreement were −42.21 to 42.96. The interclass correlation coefficient was 0.785 (95% CI: 0.662–0.863, $p < 0.001$). Regression analysis between the score means and the difference between scores were not significant (-0.007, $p = 0.949$), meaning there is no proportional bias.

3.1.1.3. Sleep/rest

Parent-proxy and child self-report PedsQL-MFS *Sleep/Rest* scores were significantly different ($t(76) = -4.394$, $p < 0.001$), so no Bland-Altman plot could be created, and proportional bias cannot be assessed. The interclass correlation coefficient was 0.845 (95% CI: 0.756–0.902, $p < 0.001$).

3.1.1.4. Cognitive

Parent-proxy and child self-report PedsQL-MFS *Cognitive* scores were significantly different ($t(76) = 6.858$, $p < 0.001$), so therefore no Bland-Altman plot could be created, and proportional bias cannot be assessed. The interclass correlation coefficient was −0.389 (95% CI: −1.186 - −0.117, $p = 0.923$).

3.2. The VFS

Median item response theory (IRT) scale scores, IQRs and minimum-maximum scores are shown in **Table 3**. **Figure 3** displays the median IRT scores with their respective IQRs for CNH, CUHL and CBHL for both child self-report and parent-proxy report from the VFS. A higher score indicates a greater level of fatigue.

TABLE 3 Medians (interquartile range) and [min, max] of child self-report and parent-proxy report scores from the VFS.

	NH	UHL	BHL
VFS-C	$N = 32$	$N = 19$	$N = 29$
Total Fatigue	−1.2 (2.4) [−2.5, 2.7]	0.6 (2.3) [−2.5, 2.7]	0.3 (2.0) [−1.7, 2.7]
VFS-*P*	$N = 39$	$N = 23$	$N = 37$
Mental Fatigue	−1.4 (1.6) [−2.0, 0. 7]	0.3 (1.8) [−2.0, 2.3]	0.9 (1.8) [−1.7, 2.4]
Physical Fatigue	−0.9 (1.2) [−2.3, 0.8]	−0.2 (1.6) [−2.3, 2.0]	0.3 (1.8) [−2.3, 2.0]

In the VFS-C (child self-report), both hearing loss groups (CUHL and CBHL) reported greater levels of fatigue than CNH. Fatigue scores were statistically significantly different between CNH, CUHL and CBHL ($H(2) = 13.892$, $p < 0.001$). Pairwise comparisons with Bonferroni corrections found CBHL experience significantly greater fatigue than CNH ($p = 0.001$). CUHL also experienced significantly greater fatigue than CNH ($p = 0.029$). There were no significant differences in fatigue levels between CUHL and CBHL in the VFS-C, though CUHL reported a higher level of fatigue than CBHL.

The VFS-*P* (parent-proxy) splits fatigue into two subdomains, mental fatigue and physical fatigue. Fatigue scores were statistically significantly different between CNH, CUHL and CBHL in both the Mental fatigue ($H(2) = 48.644$, $p < 0.001$) and Physical fatigue ($H(2) = 30.364$, $p < 0.001$) subdomains. In the VFS-*P*, again both CUHL and CBHL reported significantly greater levels of fatigue than CNH in the Mental fatigue domain (CBHL-CNH, $p < 0.001$; CUHL-CNH, $p < 0.001$) and the Physical fatigue domain (CBHL-CNH, $p < 0.001$; CUHL-CNH, $p = 0.011$). Parents of CBHL reported the greatest level of fatigue (for their children) compared to parent ratings for CUHL and CNH in both the Mental and Physical fatigue domains, whilst fatigue levels rated by parents of

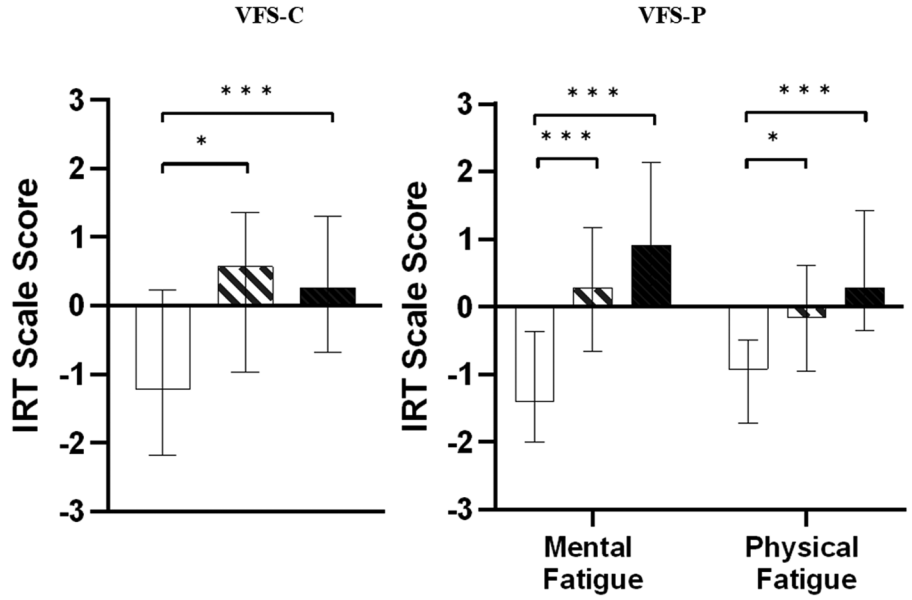

FIGURE 3
Median item response theory (IRT) scale scores for the VFS-children (left) and VFS-parent (right). Greater values indicate a greater level of fatigue. Bars and error bars shown represent medians ± IQRs. CNH; white bars, CUHL; oblique stripes, CBHL; black bars respectively. Asterisks show significant differences between groups (*$p < 0.05$, **$p < 0.01$, ***$p < 0.001$).

TABLE 4 Medians (interquartile range) and [min, max] of child self-report and parent-proxy report from the PROMIS fatigue short form.

	NH	UHL	BHL
PROMIS Fatigue Child Self-Report	$N = 32$	$N = 19$	$N = 29$
T Score	39.8 (24.1) [30.3, 76.3]	51.1 (28.2) [30.3, 81.5]	53.0 (18.7) [30.3, 80.0]
PROMIS Fatigue Parent-proxy	$N = 39$	$N = 23$	$N = 37$
T Score	42.1 (14.6) [34.1, 61.0]	50.7 (21.9) [34.1, 75.2]	57.4 (18.2) [34.1, 80.8]

CUHL fall between ratings by parents of CBHL and parents of CNH. There were, however, no statistically significant differences between fatigue levels for CUHL and CBHL as rated by parents.

3.3. PROMIS fatigue short form

T-scores for the child self-report and parent-proxy PROMIS Fatigue Short Form are shown in Table 4. Figure 4 displays the median T-scores with their respective IQRs for CNH, CUHL and CBHL for both child self-report and parent-proxy report from the PROMIS Fatigue Short Form. In the child self-report, there was a statistically significant difference between fatigue score and hearing loss groups (CNH, CUHL and CBHL; $H(2) = 8.33$, $p = 0.016$). Both CUHL and CBHL scored higher than CNH, though only CBHL scored a significantly greater level of fatigue than CNH ($p = 0.017$).

In the parent-proxy report, fatigue scores were statistically significantly different between groups ($H(2) = 28.325$, $p < 0.001$). Parents of CBHL and parents of CUHL scored significantly greater fatigue scores than parents of CNH (CBHL-CNH, $p < 0.001$; CUHL-CNH, $p = 0.015$).

3.3.1. Parent-Proxy vs. Child self-report

Agreement by Bland-Altman plots are shown in Supplementary Figure S1C with a mean difference of PROMIS Fatigue score of −1.72 (95% CI: −4.16 – 0.72); the limits of agreement were −22.78 (95% CI: −18.60 – −26.96) to 19.34 (95% CI: 15.15–23.53). The interclass correlation coefficient was 0.813 (95% CI: 0.705–0.881, $p < 0.001$). Regression analysis between the score means and the difference between scores was not significant (0.70, $p = 0.482$), meaning there is no proportional bias.

4. Discussion

In this study, we compared fatigue levels experienced by children with NH, UHL and BHL using three separate measures of fatigue to: a) understand which questionnaires were sufficiently able to discriminate levels of fatigue between children with different types of hearing loss; and b) help us to understand whether parent-proxy reports of fatigue using these measures were good alternative measures to child self-reports. We found that both the general (PedQL-MFS and PROMIS Fatigue) and listening-specific (VFS) fatigue measures were able to detect that CBHL experienced significantly greater levels of fatigue than CNH, in both child self-report and parent-proxy measures. Out of the three child self-report fatigue measures used, only the listening-specific VFS had the ability to discriminate between fatigue levels in CUHL and CNH. Further, parents of CUHL reported significantly greater fatigue compared to parents of CNH in both the VFS-*P* and the

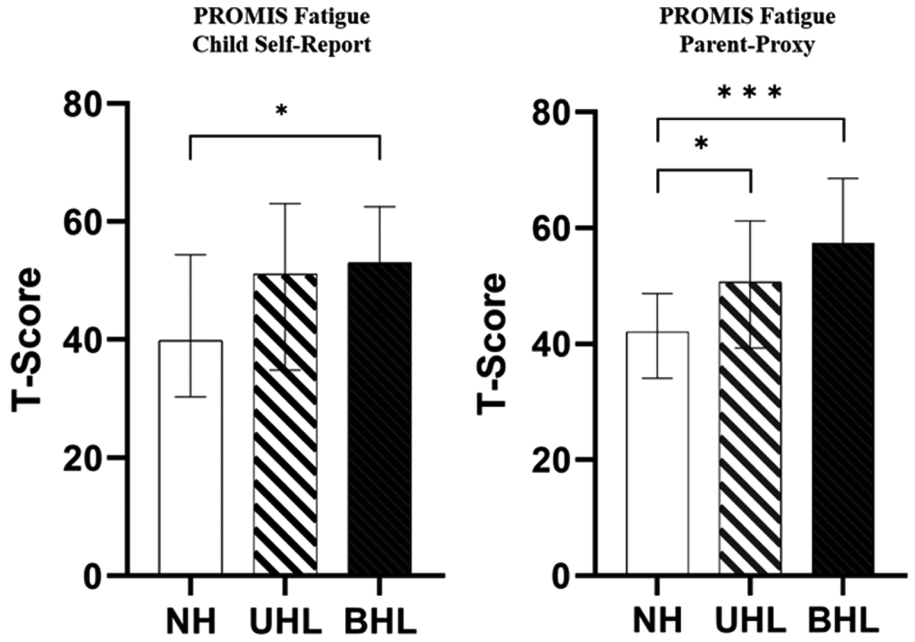

FIGURE 4
Median PROMIS fatigue short form 10a T-scores for the child self-report (left) and parent-proxy (right) respondents. Higher values indicate greater fatigue. Bars and error bars shown represent medians ± IQRs. CNH; white bars, CUHL; oblique stripes, CBHL; black bars respectively. Asterisks show significant differences between groups (*$p < 0.05$, **$p < 0.01$, ***$p < 0.001$).

PROMIS Fatigue Parent-Proxy measures, as well as the *Total*, *General* and *Cognitive* domains of the PedsQL-MFS.

The PedsQL-MFS has been used the most extensively to measure fatigue in CHL (7, 9, 25, 26). In our data, both CUHL and CBHL and their parents rated significantly greater *Cognitive* fatigue than CNH and their parents. These data most closely relate to the data presented by the listening-related VFS, and are suggestive of an increase in cognitive load due to listening effort. Marsella et al. (38) demonstrated an increase in cognitive load in children with asymmetric hearing loss by measuring alpha power in the parietal cortex using EEG. They found significantly higher parietal alpha power levels in noisy conditions compared to quiet listening conditions. These increased cognitive demands in children with hearing loss led to an increase in cognitive fatigue. In a study by Brännström et al. (39), native and non-native language speaking children underwent listening comprehension tests whilst pupil dilation was measured using pupillometry. They found pupil dilation to be greater in poorer listening conditions, indicating an increased listening effort. They also found that baseline pupil size decreased over the listening comprehension trials, indicating listening-related fatigue. This study demonstrates the dynamics between listening effort and cognitive load during difficult listening situations. However, Alhanbali et al. (3) assessed fatigue and effort in hearing-impaired participants using the Fatigue Assessment Scale and Effort Assessment Scale. There were no significant differences between effort and fatigue scores, and the measures were only weakly correlated. These findings could suggest that listening effort is not the only predictor of fatigue and that there are potentially many other factors that could give rise to fatigue in hearing-impaired individuals. The development of fatigue may be produced by the lack of motivation to sustain effort within a particular task (40, 41). Throughout most domains of the PedsQL-MFS, parents of CUHL and CBHL rated lower levels of fatigue than their children. This is similar to the effect seen in Hornsby et al. (9), where parents rated their children as having significantly less fatigue than child self-report in the *Sleep/Rest*, *Cognitive* and *Total* domains. In our findings, though there was agreement between parent and child reports within the *Total* and *General Fatigue* domains, no agreement was found within the *Cognitive* and *Sleep/Rest* domains. This differs from Sindhar et al. (26) and Carpenter et al. (25) where no significant differences between parent proxy and child reports were found in any fatigue domains. There is little consistency in literature concerning the agreement between parent-proxy and child self-report outcomes. Ultimately, this is most likely due to individual relationships between parents and their children. It could also be due to different circumstances under which participants completed the questionnaires. The lack of consistency between parent-proxy and child self-report comparisons suggests that, if able to, children should complete their own fatigue self-report measures if required clinically.

This study is the first to compare fatigue levels in children with differing levels of hearing loss within the United Kingdom, the former studies were conducted within the United States. Parent child relationships across different cultures vary greatly (42) and understanding of fatigue may vary between nations. Furthermore, this study took place online as it took place during the Covid-19 pandemic, whilst Sindhar et al. (26), Carpenter et al. (25) and Hornsby et al. (9) administered the questionnaire in person. Online and in-person questionnaires both have their benefits and risks. Participants completing a survey online may feel more comfortable and free, as they are safe behind an extra layer of anonymity. However, completing a questionnaire online also gives

the freedom to complete it at any time of the day, rather than a booked session that would usually be within working hours.

Although the PROMIS Fatigue Short form has not previously been used to measure fatigue in CHL, and was initially designed to measure fatigue in children with or without chronic pain (43), it was able to detect more fatigue in children with hearing loss compared to children with normal hearing in our sample. Though this questionnaire is unidimensional, results show similarity to scores within the PedsQL-MFS. The PROMIS showed more consistency between child self-report ratings and parent-proxy ratings than the multidimensional PedsQL-MFS.

Out of the three fatigue questionnaires, unsurprisingly the results show that the VFS was the most sensitive measure, as it was able to detect significantly greater fatigue in both CUHL and CBHL, compared to CNH, in both the child self-report and parent-proxy tools. This tool is an important step in helping us to understand fatigue in this population. This is a newly developed and newly validated tool, and is currently the only measure available that was designed to specifically assess listening-related fatigue in CHL (4, 27).

Until recently, the effects of UHL in children have been understudied, and assumed to be minimal (44). For example, children with UHL in the United Kingdom are not routinely funded to have a remote microphone system (RMS), a wireless microphone system that transmits sound from a talker to the receiver's ear (28), even though use of RMS has been shown to improve performance in sustained auditory attention ability (45). Unilateral hearing loss does, in fact, have many consequences (46); CUHL perform worse in localisation tasks compared to CNH (17, 47), and have poorer speech and language comprehension, reduced word recognition (17) and lower IQ scores (48, 49), to name a few. In this study, we found no significant differences in fatigue scores between CUHL and CBHL in child self-report questionnaires. Our data illustrate the need for continued research into the effects and impacts of unilateral hearing loss on children, to support the development of interventions to reduce this impact and the recognition of the needs of CUHL in policy and service provision.

4.1. Limitations

This study was open during national lockdowns due to the COVID-19 pandemic. Most children who took part in the study took part in school lessons from home, separated from their usual routines, whilst some children carried on going into school. Changes in their usual routines could have greatly affected responses in questionnaires. For example, children who still attended school may have been subjected to louder environments than usual due to mixing of age groups in classrooms, therefore exerting more listening effort. In classrooms, a predictor of fatigue, measured using the PedsQL-MFS, was found to be perceived listening difficulty (50). Children from single-child households may have had a quieter, more peaceful, work environment compared to children living in multiple-child households. As the study took place online, participants were not able to ask for clarifications from the researcher, as they would be able to do if they were completing questionnaires in person. It is also possible that children and parents completed the questionnaires together, causing the results to lack independence. Furthermore, the study was open for a year, and so fatigue levels may have differed depending on seasonal effects such as time spent outdoors.

Using a generic rather than a disease-specific outcome measure may be common in the clinical environment, but not always advisable for children with hearing loss. However, for CBHL the generic fatigue questionnaire measures were found to be acceptable and overall, produced quite similar findings to the outcomes from hearing specific questionnaires. Unfortunately, where CUHL measures are concerned then only the VFS-C was able to discriminate between the fatigue levels for CUHL and CNH.

Fatigue is complex and definitions and descriptions of fatigue can vary greatly, often depending on their source. For example, physical fatigue, defined as "the reduced desire to take part in physically demanding tasks, or reduced ability to maintain optimal performance" (51), is different from mental fatigue, described as a "reduced ability or desire to perform tasks that require concentration, attention, clear thinking and memory" (52). The broadness of fatigue as a construct renders measurement difficult, so it is therefore prudent to ascertain which aspects of fatigue demand most focus. Multidimensional fatigue scales, such as the 18-item PedsQL-MFS, can be advantageous as they capture a plurality of distinct domains in which fatigue may manifest, therefore providing a more nuanced description of impact than unidimensional scales. A negative consequence of this is that they are time-consuming and individual questions are not always relevant to the respondent. This is particularly prominent in questionnaires that are designed for the general population, rather than for specific cohorts. By contrast, the VFS-Peds is a 10-item questionnaire, which focuses on one source of fatigue (listening), but integrates several outcome domains in one score. For children with hearing loss, the VFS-Peds is more relevant and less time consuming than the PedsQL-MFS.

5. Conclusion

This study has measured fatigue in children with normal hearing (CNH), children with unilateral hearing loss (CUHL) and children with bilateral hearing loss (CBHL) using three separate tools for measuring fatigue. Our results suggest that both CUHL and CBHL experience more fatigue than their normal hearing peers, and that the listening-specific VFS may be better able to detect fatigue in CUHL. Research is needed to determine the potential differences or similarities in the experiences of CUHL, compared to CBHL, in order to understand the nature of fatigue in these populations so that we can ultimately find ways to improve their quality of life and reduce their fatigue.

Data availability statement

The raw data supporting the conclusions of this article will be made available by the authors, without undue reservation.

Ethics statement

The studies involving human participants were reviewed and approved by the Health Research Authority (HRA), Wales Research Ethics Committee (20/WA/0233) and the Nottingham Research Committee (52-0720). Informed consent to participate in this study was provided by the participants, or participants' legal guardian/next of kin if under the age of 16 years old.

Author contributions

Conceptualization, BA, ST, GN, IW, PK; Data curation, BA, ST, PK; Formal analysis, BA; Investigation, BA; Methodology, BA, PK; Project administration, BA; Supervision, ST, IW, GN, PK; Validation, BA; Writing—original draft, BA; Writing—review and editing, BA, ST, GN, RS, PK. All authors have read and agreed to the published version of the manuscript. All authors contributed to the article and approved the submitted version.

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Conflict of interest

The authors declare that the research was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

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Supplementary material

The Supplementary Material for this article can be found online at: https://www.frontiersin.org/articles/10.3389/fped.2023.1127578/full#supplementary-material.

References

1. O'Connor PJ. Evaluation of four highly cited energy and fatigue mood measures. *J Psychosom Res.* (2004) 57(5):435–41. doi: 10.1016/j.jpsychores.2003.12.006

2. Holman JA, Hornsby BWY, Bess FH, Naylor G. Can listening-related fatigue influence well-being? Examining associations between hearing loss, fatigue, activity levels and well-being. *Int J Audiol.* (2021) 60(sup2):47–59. doi: 10.1080/14992027.2020.1853261

3. Alhanbali S, Dawes P, Lloyd S, Munro KJ. Self-Reported Listening-Related Effort and Fatigue in Hearing-Impaired Adults. *Ear Hear.* (2017) 38(1):e39–48. doi: 10.1097/AUD.0000000000000361

4. Bess FH, Davis H, Camarata S, Hornsby BWY. Listening-Related Fatigue in Children With Unilateral Hearing Loss. *Lang Speech Hear Serv Sch.* (2020) 51(1):84–97. doi: 10.1044/2019_LSHSS-OCHL-19-0017

5. Bess FH, Hornsby BWY. The Complexities of Fatigue in Children with Hearing Loss. *Perspectives on Hearing and Hearing Disorders in Childhood.* (2014) 24(2):25–39. doi: 10.1044/hhdc24.2.25

6. Gustafson SJ, Key AP, Hornsby BWY, Bess FH. Fatigue Related to Speech Processing in Children With Hearing Loss: behavioral, Subjective, and Electrophysiological Measures. *J Speech Lang Hear Res.* (2018) 61(4):1000–11. doi: 10.1044/2018_JSLHR-H-17-0314

7. Hornsby BW, Werfel K, Camarata S, Bess FH. Subjective fatigue in children with hearing loss: some preliminary findings. *Am J Audiol.* (2014) 23(1):129–34. doi: 10.1044/1059-0889(2013/13-0017)

8. Lindburg M, Ead B, Jeffe DB, Lieu JEC. Hearing Loss–Related Issues Affecting Quality of Life in Preschool Children. *Otolaryngol–Head and Neck Surg.* (2020) 164(6):1322–9. doi: 10.1177/0194599820962475

9. Hornsby BWY, Gustafson SJ, Lancaster H, Cho SJ, Camarata S, Bess FH. Subjective Fatigue in Children With Hearing Loss Assessed Using Self- and Parent-Proxy Report. *Am J Audiol.* (2017) 26(3s):393–407. doi: 10.1044/2017_AJA-17-0007

10. Pichora-Fuller MK, Kramer SE, Eckert MA, Edwards B, Hornsby BW, Humes LE, et al. Hearing Impairment and Cognitive Energy: the Framework for Understanding Effortful Listening (FUEL). *Ear Hear.* (2016) 37(Suppl 1):5s–27s. doi: 10.1097/AUD.0000000000000312

11. Lewis D, Schmid K, O'Leary S, Spalding J, Heinrichs-Graham E, High R. Effects of Noise on Speech Recognition and Listening Effort in Children With Normal Hearing and Children With Mild Bilateral or Unilateral Hearing Loss. *J Speech Lang Hear Res.* (2016) 59(5):1218–32. doi: 10.1044/2016_JSLHR-H-15-0207

12. McGarrigle R, Gustafson SJ, Hornsby BWY, Bess FH. Behavioral Measures of Listening Effort in School-Age Children: examining the Effects of Signal-to-Noise Ratio, Hearing Loss, and Amplification. *Ear Hear.* (2019) 40(2):381–92. doi: 10.1097/AUD.0000000000000623

13. Klatte M, Hellbrück J, Seidel J, Leistner P. Effects of Classroom Acoustics on Performance and Well-Being in Elementary School Children: a Field Study. *Environ Behav.* (2010) 42(5):659–92. doi: 10.1177/0013916509336813

14. Crandell CC, Smaldino JJ. Classroom Acoustics for Children With Normal Hearing and With Hearing Impairment. *Lang Speech Hear Serv Sch.* (2000) 31(4):362–70. doi: 10.1044/0161-1461.3104.362

15. Klatte M, Lachmann T, Meis M. Effects of noise and reverberation on speech perception and listening comprehension of children and adults in a classroom-like setting. *Noise Health.* (2010) 12(49):270–82. doi: 10.4103/1463-1741.70506

16. Howard CS, Munro KJ, Plack CJ. Listening effort at signal-to-noise ratios that are typical of the school classroom. *Int J Audiol.* (2010) 49(12):928–32. doi: 10.3109/14992027.2010.520036

17. Reeder RM, Cadieux J, Firszt JB. Quantification of speech-in-noise and sound localisation abilities in children with unilateral hearing loss and comparison to normal hearing peers. *Audiol Neurootol.* (2015) 20(Suppl (1):31–7. doi: 10.1159/000380745

18. Davis H, Schlundt D, Bonnet K, Camarata S, Hornsby B, Bess FH. Listening-Related Fatigue in Children With Hearing Loss: perspectives of Children, Parents, and School Professionals. *Am J Audiol.* (2021) 30(4):929–40. doi: 10.1044/2021_AJA-20-00216

19. Nap-van der Vlist MM, Dalmeijer GW, Grootenhuis MA, van der Ent K, van den Heuvel-Eibrink MM, Swart JF, et al. Fatigue among children with a chronic disease: a cross-sectional study. *BMJ Paediatr Open*. (2021) 5(1):e000958. doi: 10.1136/bmjpo-2020-000958

20. Hockenberry-Eaton M, Hinds P, Howard V, Gattuso J, O'Neill JB, Alcoser P, et al. Developing a conceptual model for fatigue in children. *Eur J Oncol Nurs*. (1999) 3 (1):5–11. doi: 10.1016/1462-3889(91)80005-7

21. Garralda ME, Rangel L. Impairment and coping in children and adolescents with chronic fatigue syndrome: a comparative study with other paediatric disorders. *J Child Psychol and Psychiatr*. (2004) 45:543–52. doi: 10.1111/j.1469-7610.2004.00244.x

22. Whitehead L. The measurement of fatigue in chronic illness: a systematic review of unidimensional and multidimensional fatigue measures. *J Pain Symptom Manage*. (2009) 37(1):107–28. doi: 10.1016/j.jpainsymman.2007.08.019

23. Varni JW, Burwinkle TM, Szer IS. The PedsQL Multidimensional Fatigue Scale in pediatric rheumatology: reliability and validity. *J Rheumatol*. (2004) 31(12): 2494–500.

24. Varni JW, Burwinkle TM, Katz ER, Meeske K, Dickinson P. The PedsQL in pediatric cancer: reliability and validity of the Pediatric Quality of Life Inventory Generic Core Scales, Multidimensional Fatigue Scale, and Cancer Module. *Cancer*. (2002) 94(7):2090–106. doi: 10.1002/cncr.10428

25. Carpenter D, Dougherty W, Sindhar S, Friesen T-N, Lieu J, Kesser BW. Are children with unilateral hearing loss more tired? *Int J Pediatr Otorhinolaryngol*. (2022) 155:111075. doi: 10.1016/j.ijporl.2022.111075

26. Sindhar S, Friesen TL, Carpenter D, Kesser B, Lieu JEC. Fatigue in Children With Unilateral and Bilateral Hearing Loss. *Otol Neurotol*. (2021) 42(9):e1301–7. doi: 10.1097/MAO.0000000000003225

27. Hornsby BW, Camarata S, Cho S-J, Davis H, McGarrigle R, Bess Fred H. Development and Evaluation of Pediatric Versions of the Vanderbilt Fatigue Scale for Children With Hearing Loss. Journal of speech. *Language and Hearing Res*. (2022) 65 (6):2343–63. doi: 10.1044/2022_JSLHR-22-00051

28. Bagatto M, DesGeorges J, King A, Kitterick P, Laurnagaray D, Lewis D, et al. Consensus practice parameter: audiological assessment and management of unilateral hearing loss in children. *Int J Audiol*. (2019) 58(12):805–15. doi: 10.1080/14992027.2019.1654620

29. Brookhouser PE, Worthington DW, Kelly WJ. Unilateral hearing loss in children. *Laryngoscope*. (1991) 101(12):1264–72. doi: 10.1002/lary.5541011202

30. Lieu JE. Unilateral hearing loss in children: speech-language and school performance. *B-ENT*. (2013) 21(Suppl):107–15. PMID: 24383229; PMCID: PMC4382076

31. Lieu JE, Tye-Murray N, Karzon RK, Piccirillo JF. Unilateral hearing loss is associated with worse speech-language scores in children. *Pediatrics*. (2010) 125(6): e1348–55. doi: 10.1542/peds.2009-2448

32. Fitzpatrick EM, Gaboury I, Durieux-Smith A, Coyle D, Whittingham J, Nassrallah F. Auditory and language outcomes in children with unilateral hearing loss. *Hear Res*. (2019) 372:42–51. doi: 10.1016/j.heares.2018.03.015

33. Irwin DE, Stucky BD, Thissen D, Dewitt EM, Lai JS, Yeatts K, et al. Sampling plan and patient characteristics of the PROMIS pediatrics large-scale survey. *Qual Life Res*. (2010) 19(4):585–94. doi: 10.1007/s11136-010-9618-4

34. Irwin DE, Gross HE, Stucky BD, Thissen D, DeWitt EM, Lai JS, et al. Development of six PROMIS pediatrics proxy-report item banks. *Health Qual Life Outcomes*. (2012) 10:22. doi: 10.1186/1477-7525-10-22

35. Crichton A, Knight S, Oakley E, Babl FE, Anderson V. Fatigue in child chronic health conditions: a systematic review of assessment instruments. *Pediatrics*. (2015) 135(4):e1015–31. doi: 10.1542/peds.2014-2440

36. Rothrock NE, Amtmann D, Cook KF. Development and validation of an interpretive guide for PROMIS scores. *J Patient-Reported Outcomes*. (2020) 4(1):16. doi: 10.1186/s41687-020-0181-7

37. Bland JM, Altman DG. Statistical methods for assessing agreement between two methods of clinical measurement. *Lancet*. (1986) 1(8476):307–10. doi: 10.1016/S0140-6736(86)90837-8

38. Marsella P, Scorpecci A, Cartocci G, Giannantonio S, Maglione AG, Venuti I, et al. EEG activity as an objective measure of cognitive load during effortful listening: a study on pediatric subjects with bilateral, asymmetric sensorineural hearing loss. *Int J Pediatr Otorhinolaryngol*. (2017) 99:1–7. doi: 10.1016/j.ijporl.2017.05.006

39. Brännström KJ, Rudner M, Carlie J, Sahlén B, Gulz A, Andersson K, et al. Listening effort and fatigue in native and non-native primary school children. *J Exp Child Psychol*. (2021) 210:105203. doi: 10.1016/j.jecp.2021.105203

40. Alhanbali S, Dawes P, Lloyd S, Munro KJ. Hearing Handicap and Speech Recognition Correlate With Self-Reported Listening Effort and Fatigue. *Ear Hear*. (2018) 39(3):470–4. doi: 10.1097/AUD.0000000000000515

41. Hockey R. *The psychology of fatigue: Work, effort and control*. Cambridge: Cambridge University Press (2013).

42. Trommsdorff G. Parent-child relations over the lifespan: A cross-cultural perspective. In: Rubin KH, Chung OB, editors. *Parenting beliefs, behaviors, and parent-child relations: a cross-cultural perspective*. New York: Psychology Press (2006). p. 143–83.

43. Sommer A, Grothus S, Grochowska K, Claus BB, Stahlschmidt L, Wager J. Assessing fatigue in children and adolescents: psychometric validation of the German version of the PROMIS® Pediatric Short Form v2.0 - Fatigue 10a in school children and pediatric chronic pain patients. *Qual Life Res*. (2022) 31(4):1257–66. doi: 10.1007/s11136-021-03032-8

44. Oyler RF, Oyler AL, Matkin ND. Unilateral Hearing Loss. *Lang Speech Hear Serv Sch*. (1988) 19(2):201–10. doi: 10.1044/0161-1461.1902.201

45. Mondelli MFCG, Jacob RTdS, Honório HM. The use of remote microphone systems in unilateral hearing loss: a preliminary study among Brazilian children and teenagers. *J Appl Oral Sci: Revista FOB*. (2019) 27:e20180744–e20180744. doi: 10.1590/1678-7757-2018-0744

46. van Wieringen A, Boudewyns A, Sangen A, Wouters J, Desloovere C. Unilateral congenital hearing loss in children: challenges and potentials. *Hear Res*. (2019) 372:29–41. doi: 10.1016/j.heares.2018.01.010

47. Van Deun L, van Wieringen A, Van den Bogaert T, Scherf F, Offeciers FE, Van de Heyning PH, et al. Sound localization, sound lateralization, and binaural masking level differences in young children with normal hearing. *Ear Hear*. (2009) 30(2):178–90. doi: 10.1097/AUD.0b013e318194256b

48. Purcell PL, Shinn JR, Davis GE, Sie KCY. Children with unilateral hearing loss may have lower intelligence quotient scores: a meta-analysis. *Laryngoscope*. (2016) 126 (3):746–54. doi: 10.1002/lary.25524

49. Niedzielski A, Humeniuk E, Błaziak P, Gwizda G. Intellectual efficiency of children with unilateral hearing loss. *Int J Pediatr Otorhinolaryngol*. (2006) 70(9):1529–32. doi: 10.1016/j.ijporl.2006.02.011

50. Gustafson SJ, Camarata S, Hornsby BWY, Bess FH. Perceived Listening Difficulty in the Classroom, Not Measured Noise Levels, Is Associated With Fatigue in Children With and Without Hearing Loss. *Am J Audiol*. (2021) 30(4):956–67. doi: 10.1044/2021_AJA-21-00065

51. Gandevia SC. Some Central and Peripheral Factors Affecting Human Motoneuronal Output in Neuromuscular Fatigue. *Sports Med*. (1992) 13(2):93–8. doi: 10.2165/00007256-199213020-00004

52. Chalder T, Berelowitz G, Pawlikowska T, Watts L, Wessely S, Wright D, et al. Development of a fatigue scale. *J Psychosom Res*. (1993) 37(2):147–53. doi: 10.1016/0022-3999(93)90081-P

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EDITED BY
Valerie Sung,
Royal Children's Hospital, Australia

REVIEWED BY
Sally Kay Thornton,
University of Nottingham, United Kingdom
Jing Wang,
Royal Children's Hospital, Australia

*CORRESPONDENCE
Elizabeth M. Fitzpatrick
✉ elizabeth.fitzpatrick@uottawa.ca

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Trajectory of hearing loss in children with unilateral hearing loss

Elizabeth M. Fitzpatrick[1,2]*, Flora Nassrallah[2], Isabelle Gaboury[3], JoAnne Whittingham[2,4], Bénédicte Vos[5], Doug Coyle[6], Andrée Durieux-Smith[1,2], Marie Pigeon[4] and Janet Olds[2,4]

[1]Faculty of Health Sciences, University of Ottawa, Ottawa, ON, Canada, [2]Child Hearing Laboratory, CHEO Research Institute, Ottawa, ON, Canada, [3]Faculty of Medicine and Health Sciences, Université de Sherbrooke, Longueuil, QC, Canada, [4]Audiology Clinic, CHEO, Ottawa, ON, Canada, [5]School of Public Health, Université libre de Bruxelles (ULB), Brussells, Belgium, [6]School of Epidemiology, Public Health and Preventive Medicine, University of Ottawa, Ottawa, ON, Canada

Introduction: The aim of this study was to quantify the amount of deterioration in hearing and to document the trajectory of hearing loss in early identified children with unilateral hearing loss (UHL). We also examined whether clinical characteristics were associated with the likelihood of having progressive hearing loss.

Methods: As part of the Mild and Unilateral Hearing Loss Study, we followed a population-based cohort of 177 children diagnosed with UHL from 2003 to 2018. We applied linear mixed models to examine hearing trends over time including the average amount of change in hearing. Logistic regression models were used to examine the relationship between age and severity at diagnosis, etiology, and the likelihood of progressive loss and amount of deterioration in hearing.

Results: The median age of the children at diagnosis was 4.1 months (IQR 2.1, 53.9) and follow-up time was 58.9 months (35.6, 92.0). Average hearing loss in the impaired ear was 58.8 dB HL (SD 28.5). Over the 16-year period, 47.5% (84/177) of children showed deterioration in hearing in one or both ears from their initial diagnostic assessment to most recent assessment including 21 (11.9%) who developed bilateral hearing loss. Average deterioration in the impaired ear ranged from 27 to 31 dB with little variation across frequencies. Deterioration resulted in a change in category of severity for 67.5% (52/77) of the children. Analysis for children who were followed for at least 8 years showed that most lost a significant amount of hearing rapidly in the first 4 years, with the decrease stabilizing and showing a plateau in the last 4 years. Age and severity at diagnosis were not significantly associated with progressive/stable loss after adjusting for time since diagnosis. Etiologic factors (ENT external/middle ear anomalies, inner ear anomalies, syndromic hearing loss, hereditary/genetic) were found to be positively associated with stable hearing loss.

Conclusion: Almost half of children with UHL are at risk for deterioration in hearing in one or both ears. Most deterioration occurs within the first 4 years following diagnosis. Most children did not experience sudden "large" drops in hearing but more gradual decrease over time. These results suggest that careful monitoring of UHL especially in the early years is important to ensure optimal benefit from early hearing loss detection.

KEYWORDS

unilateral hearing loss, children, progressive loss, trajectory, etiology

Introduction

Unilateral hearing loss (UHL) in children has gained increasing attention as a clinically important hearing disorder. Permanent childhood hearing loss is relatively common affecting 3–4 per 1,000 children when all degrees of bilateral and unilateral loss are considered during childhood (1). An estimated 20%–30% of these children have unilateral hearing loss (1, 2). In contrast to historical practices, a substantial proportion of children with UHL are now diagnosed in infancy or early childhood due to widespread population level newborn hearing screening (NHS) (3, 4). Permanent UHL affects about 1 per 1,000 infants based on newborn screening cohorts (5, 6).

Historically, the clinical implications of UHL were not well understood. Unlike children with bilateral hearing loss, these children have access to speech and develop spoken language without intervention. However, there is a growing consensus that UHL affects typical development of auditory pathways and auditory function with implications for communication and academic development for at least some children (7–10). Difficulties in language and academic performance can persist at school age (11–13). However, some uncertainty remains about the consequences of UHL and who is most at risk for difficulties and the need for intervention and overall best practices continue to receive attention (14–17). Parental uncertainty about the effects of UHL has been reflected in parent-focused literature and studies suggest considerable indecision about intervention recommendations (18–20).

There is some variation in NHS programs worldwide in defining hearing disorders including whether mild bilateral and UHL, historically considered to be minimal losses, are specifically targeted (17, 21, 22). Arguably, one reason for including UHL is that as a public health intervention, screening aims not only to improve developmental outcomes but also to prevent delay through early audiologic management and intervention. Programs such as the Infant Hearing Program in Ontario, Canada (23), target the early detection of UHL on the basis that there may be negative consequences associated with any hearing loss and that children are at risk for deterioration in hearing in the other ear. Several studies have reported that children with hearing loss are at risk for further deterioration in hearing with wide variation in rates of progressive loss documented (24–28). Purcell et al. (29) reported that 32.8% of 128 children with sensorineural UHL who had their first audiologic assessment at age 7.7 years showed progressive hearing loss. Paul et al. (30) reported that 19% of 80 children showed progressive loss but 68% of children were initially identified with severe-profound hearing loss and further deterioration in hearing thresholds may not have been captured. Importantly, there has been little focus on the trajectory of hearing loss in children with UHL (31), particularly in early identified children. Datasets available prior to NHS included few children with early-detected UHL, limiting the possibility to document changes in hearing (3). Therefore, little is known about when and how much change in hearing occurs.

Relatively little is known about the relationship between the clinical characteristics (e.g., etiology, age at diagnosis, severity of hearing loss) of children with UHL and the risk of progression in hearing loss. Like bilateral hearing loss, etiology is related to both genetic and environmental factors. While genetics are the most common cause of bilateral hearing loss (1, 32, 33), structural and environmental causes make up a large part of the etiological distribution of UHL (33, 34). While several environmental factors including prematurity and ototoxicity have been associated with non-genetic hearing loss, congenital cytomegalovirus (cCMV) has emerged as the most common cause (32, 35). Congenital CMV accounts for 15%–20% of childhood hearing loss, including UHL, and has been associated with both late onset and progressive hearing loss (36–39). However, in an etiologic study, Dahl et al. (40) found no relationship between CMV or common genetic etiologic factors and progression of hearing loss. Structural anomalies such as enlarged vestibular aqueduct (EVA) and cochlear nerve deficiency (29, 33), common causes of UHL, have also been associated with progressive loss (29, 41). In an investigation of children with UHL, Purcell et al. (29) reported that children with bony cochlear nerve stenosis were at greater risk of progression in hearing. In the same study, risk of progression was not significantly different for children with and without EVA or for those with temporal bone anomalies versus normal imaging results. Overall, the research suggests that the relationship between etiologic factors and the risk of progressive hearing loss is rather inconclusive.

The Joint Committee on Infant Hearing (JCIH) has historically identified risk indicators for late onset and progressive permanent hearing loss, which have guided screening surveillance programs (22, 42). Our previous research on a cohort of children with bilateral/unilateral loss found no significant association between risk indicators and progressive loss except that children with craniofacial anomalies were more likely to have stable hearing loss (24). Permanent conductive (structural) loss, generally associated with craniofacial anomalies such as aural atresia has been reported in 25%–33% of children with UHL (2, 43). Onset of hearing loss, which can be related to etiology, and type and severity of hearing loss at diagnosis have also not been well-investigated in relation to progressive hearing loss.

Understanding the trajectory of hearing loss has implications for management practices including the need for surveillance and potential adjustments in intervention. Screening aims to improve developmental outcomes by detecting and managing hearing loss early and provides new opportunities to better understand the evolution of childhood hearing loss. Consistent with these goals, we have followed a population-based cohort of children with permanent hearing loss in one Canadian audiology center. The purpose of this study was to examine the clinical characteristics and the evolution of hearing loss in children with UHL. Specifically, the objectives were to: (1) determine the proportion of children with UHL, the amount of deterioration in hearing thresholds, and the trajectory of hearing loss; and (2) examine whether there was an association between clinical characteristics at diagnosis including etiology, age at diagnosis (related to onset), and severity of hearing loss and the likelihood of progressive hearing loss.

Methods

Design and setting

This longitudinal study was conducted as part of a research program examining development outcomes in children with mild bilateral or UHL. As part of this project, population-level data related to diagnosis and intervention were collected prospectively on all children with permanent hearing loss followed in the Eastern Ontario region of Canada and diagnosed from 2003 to 2018. For this study on progressive hearing loss, we also extracted all post-diagnostic audiometric data from the medical records.

The study was conducted at CHEO, a pediatric hospital which is the sole audiologic diagnostic center for infants in the area screened through a province-wide early hearing detection and intervention (EHDI) program. Screening targets include mild bilateral and UHL. The clinic also provides services for children who relocate to the area. Well-established clinical protocols for identification and follow-up of hearing loss are in place (44). Services are publicly funded through the provincial health system. The program was fully implemented in 2003 and data for this study are population-based, covering a birth cohort of approximately 240,000 infants during the 16-year study period. Services for all children confirmed with permanent hearing loss include audiologic follow-up at 3- and 6-month intervals respectively in the first and second year after identification and then annually up to age 6 years. Intervention services for communication development are also provided within the audiology service.

Participants

The study population included all children followed at CHEO who were identified with permanent UHL (2003–2018). UHL was determined based on the National Workshop on Mild and Unilateral Hearing Loss (45) definition as hearing loss in one ear only with a pure-tone average (PTA at 0.5, 1, 2 kHz) of 20 dB HL or >25 dB at two or more frequencies above 2 kHz. Research Ethics Committees at the CHEO Research Institute (file #09-64X), and the University of Ottawa (file #H10-09-11) approved the study protocol.

Procedures

Data collection for this study took place in two phases. In phase 1, as part of a longitudinal study on all children with hearing loss, clinical characteristics have been collected prospectively from medical records since full NHS implementation in 2003. Data were entered in a study-specific database and included child (e.g., sex, screening status) and hearing loss details (e.g., onset, age of diagnosis, type of loss, severity of hearing loss, middle ear status, etiology, risk indicators). In addition, medical records were re-examined for this study to update the child's profile with any new etiologic information from clinical areas such as genetics (e.g., family history and/or genetic testing), infectious diseases (e.g., cCMV infections) and ENT services (e.g., imaging results). A researcher with experience in medical chart data also entered risk indicators for hearing loss based on JCIH (42) descriptions and coding was verified with an audiologist or physician if needed.

In addition to the audiologic data entered at diagnosis, all follow-up audiologic and hearing-related medical assessment results were retrospectively extracted from paper or electronic (after 2013) medical charts and entered into an SPSS database including audiometric thresholds and middle ear status (e.g., immittance results and ENT clinical notes). Category of hearing loss (mild, moderate, moderately severe, severe, profound) was assigned based on 4-frequency 0.5, 1, 2, 4 kHz) pure-tone average (PTA) applying standard audiologic definitions (shown in **Table 1**). All clinical assessment data were available to the researchers.

Determination of progressive hearing loss

A definition used in our previous research (24), adopted from Dahl et al. (40), was applied: (1) a decrease of 10 dB or greater at two or more adjacent frequencies between 0.5 and 4 kHz or a decrease in 15 dB at one octave frequency in the same frequency range. Children were categorized as having progressive hearing loss (vs. stable hearing) if there was worse hearing in the impaired ear or if the ear with normal hearing developed a loss. The presence of progressive hearing loss was determined based on a comparison of initial and most recent audiologic profiles. The initial confirmation of permanent hearing loss was based on the audiologic assessment conducted, either diagnostic auditory brainstem response (ABR) testing (using tone pip stimuli) or behavioral audiometry results. The relationship between behavioral and ABR thresholds has been well-documented and correction factors have been established to predict behavioral thresholds from ABR results (47–49). For the, ABR results, clinical audiologists had recorded the estimated behavioural thresholds (eHL) in the medical chart, applying correction factors used by the Ontario Infant Hearing Program (47) and these eHL thresholds were entered for all ABR tests. Most children, due to their age, were assessed using behavioral audiometry at their most recent assessment. Therefore, determination of progressive hearing loss for children who were initially diagnosed using ABR assessments, required a comparison of ABR (eHL thresholds) and behavioral thresholds.

Decision rules consistent with our previous research on progressive hearing loss (24) were applied. Inconclusive or incomplete results were not included for the analysis. If middle ear function was abnormal (based on tympanometry and/or ENT medical chart notes) at any assessment, audiograms with >10 dB changes in thresholds compared to previous/subsequent assessments were excluded. Assessments which included sound field results only were also excluded. Any unclear results were discussed between two researchers and reviewed with a clinical audiologist on the research team, as needed. For each audiological assessment entered, time from the confirmation of the hearing loss was calculated in months. For longitudinal analysis, the audiometric thresholds closest to and within 6 months of the year of follow-up (e.g., year 1, 2, 3, etc.) were selected (e.g., Year 2 encompassed thresholds obtained between 18 and 30 months).

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TABLE 1 Baseline clinical characteristics of the study sample ($n = 177$).

Characteristic	Study sample	Stable HL	Progressive HL
	$n = 177$	$n = 93$	$n = 84$[a]
Sex, *n* (%)			
Female	87 (49.2)	42 (45.2)	45 (53.6)
Male	90 (50.8)	51 (54.8)	39 (46.4)
Screening status			
Exposed to screening	134 (75.7)	69 (74.2)	65 (77.4)
Not exposed to screening	43 (24.3)	24 (25.8)	19 (22.6)
Onset hearing loss, *n* (%)			
Congenital/Early[b]	95 (53.7)	44 (47.3)	51 (60.7)
Late onset[c]	46 (26.0)	30 (32.3)	16 (19.0)
Acquired	7 (4.0)	6 (6.5)	1 (1.2)
Unknown	29 (16.4)	13 (14.0)	16 (19.0
Age diagnosis (months), median (IQR)	4.1 (2.1, 53.9)	24.3 (2.0, 58.9)	3.7 (2.2, 44.8)
Type of hearing loss, *n* (%)			
Sensorineural	119 (67.2)	57 (61.3)	62 (73.8)
Mixed	23 (13.0)	10 (10.8)	13 (15.5)
Conductive[d]	35 (19.8)	26 (28.0)	9 (10.7)
PTA (4 frequency) at diagnosis (impaired/worse ear), mean (SD)	58.8 (28.5)	63.3 (30.3)	53.8 (30.3)
Degree of hearing loss at diagnosis (impaired/worse ear), *n* (%)			
High frequency[e]	17 (9.6)	9 (9.7)	8 (9.5)
Mild (20–40 dB HL)	39 (22.0)	15 (16.1)	24 (28.6)
Moderate (41–55 dB HL)	33 (18.6)	18 (19.4)	15 (17.9)
Moderately severe (56–70 dB HL)	42 (23.7)	25 (26.9)	17 (20.2)
Severe (71–90 dB HL)	23 (13.0)	9 (9.7)	14 (16.7)
Profound (>90 dB HL)	23 (13.0)	17 (18.3)	6 (7.1)
Risk factors at diagnosis, *n* (%)			
Craniofacial anomalies	32 (18.1)	22 (23.7)	10 (11.9)
Syndromes (associated with HL)	9 (5.1)	3 (3.2)	6 (7.1)
Family history	9 (5.1)	4 (4.3)	5 (6.0)
NICU	7 (4.0)	1 (1.1)	6 (7.1)
CMV	4 (2.3)	0	4 (4.8)
Meningitis	4 (2.3)	3 (3.2)	1 (1.2)
Oncology treatment	3 (1.7)	3 (3.2)	0
No risk factors	109 (61.6)	57 (61.3)	52 (61.9)
Etiology, *n* (%)			
ENT anomaly-external/middle ear	28 (15.8)	22 (23.7)	6 (7.1)
ENT anomaly-inner ear	14 (7.9)	8 (8.6)	6 (7.1)
Syndrome (associated with HL)	21 (11.9)	8 (8.6)	13 (15.5)
Hereditary/genetic	15 (8.5)	8 (8.6)	7 (8.3)
CMV	8 (4.5)	1 (1.1)	7 (8.3)
NICU admission[f]	5 (2.8)	1 (1.1)	4 (4.8)
Meningitis	4 (2.3)	3 (3.2)	1 (1.2)
Oncology	3 (1.7)	3 (3.2)	0
Unknown	79 (44.6)	39 (41.9)	40 (47.6)
Total assessments, median (IQR)	7.0 (5.0, 11.0)	6.0 (5.0, 9.0)	9.0 (6.0, 14.0)
Time to most recent audiogram (months), median (IQR)	58.9 (35.6, 92.0)	50.6 (32.6, 88.5)	64.3 (39.3, 92.2)
Age at most recent audiogram (months), median (IQR)	87.5 (55.1, 139.0)	82.2 (52.1, 139.0)	88.8 (55.9, 140.6)

CMV, cytomegalovirus; ENT, ear, nose, and throat; HL, hearing loss; NICU, neonatal intensive care unit.
[a]Includes 7 children who developed hearing loss in the normal hearing ear; impaired ear remained stable.
[b]Early onset ≤6 months of age.
[c]Late onset: >6 months of age.
[d]Includes only permanent conductive hearing loss.
[e]Defined as >25 dB HL at >2 frequencies above 2 kHz.
[f]The children with NICU admission had no other determined etiologies (e.g., syndrome) and had one of the JCIH treatments or conditions (ECMO, assisted ventilation, ototoxic medication, and hyperbilirubinemia requiring exchange transfusion). A total of 16 children were admitted to the NICU but other children were classified in specific etiologic categories, e.g., 5 children with syndromes and 6 children with other etiologies such as CMV and ENT anomaly/inner ear.

Data analysis

Statistical analyses were conducted using SPSS (version 26). Participant characteristics were summarized using descriptive statistics including means and standard deviations, medians and interquartile ranges, and frequency counts as appropriate. One outcome of interest was the proportion of children with progressive hearing loss. Differences in clinical characteristics (e.g., onset, type, severity of hearing loss, etiology, risk indicators) were compared for children with progressive and stable hearing levels using *t*-tests or Mann-Whitney *U* tests (as appropriate) for continuous variables and chi-square tests for categorical variables.

The amount of change in hearing loss across frequencies was calculated from first to most recent audiometric assessment. For the longitudinal analysis, the trajectory of hearing loss (for the impaired ear at initial diagnosis) was analyzed using mixed linear models (with the identity correlation matrix) that were fit with maximum-likelihood estimation techniques to evaluate the trajectory across individual frequencies (0.5–4 kHz). To control for intra-subject variability of trajectories, a random effect was added on the linear term of the model. Another random effect on the intercept was added to control for the variability between individual baseline thresholds. The time effect was modeled as linear, quadratic, and cubic factors to be able to detect a loss in hearing (linear effect) and a change in the rate of decrease over time (quadratic and cubic effects). Analyses were conducted with all available data without imputation, as estimation of parameters using the maximum-likelihood method is considered adequate to address missing data (50, 51).

Using logistic regression, we also evaluated the relationship between clinical characteristics (age at diagnosis, severity of hearing loss at diagnosis, etiology) and status of hearing loss (stable versus progressive). Four multivariable models were also fit to evaluate the relationship between these covariates and the amount of deterioration in hearing at individual audiometric frequencies from 0.5–4 kHz. All models were adjusted for time since diagnosis. Two-tailed tests were applied for all analyses with statistical significance set at $p < 0.05$.

Results

Study population and characteristics

Figure 1 shows the selection of participants for the analysis. From 2003 to 2018, a total of 730 children were identified with permanent hearing loss in the clinic, of whom 197 (27.0%) had UHL at diagnosis. After removing children with limited follow-up and those with auditory neuropathy spectrum disorder due to the fluctuating nature of hearing loss, 177 children were available for detailed analysis. A total of 1,565 audiologic assessments were examined (median of 7.0 assessments per child; IQR 5.0, 11.0; range 3–31) to determine whether hearing loss was progressive or stable.

Description of participants

Table 1 shows the characteristics of the 177 children included in the analysis. Most children (134/177, 75.7%) were known to be exposed to newborn screening and UHL was diagnosed in infancy at a median age of 4.1 months (IQR 2.1, 53.9). Of the 43 (24.3%) children without screening, 17 (9.6%) were confirmed as not screened and 26 (14.7%) were born outside the province/country and were not screened or information was not available. Children had been followed for a median of 58.9 months (IQR 35.6, 92.0) and had a median age of 87.5 months (IQR 55.1, 139.0) at most recent assessment.

Hearing loss was determined to be congenital or early onset (<6 months) for 53.7% (95/177), late onset for 26.0% and acquired (e.g., meningitis or other known causes) for 4.0%. Onset was unknown for the remaining 16.4% of children due to unknown screening status and no early diagnostic assessment. Most (142/177, 80.2%) children presented with sensorineural (67.2%) or mixed (13.0%) hearing loss at diagnosis and the remaining 19.8% with permanent conductive (structural) loss. The mean hearing loss at diagnosis (4-frequency PTA in the impaired ear) was 58.8 dB (SD 28.5) with 74.0% (131/177) of children having <70 dB HL (mild to severe). One or more known risk indicators for hearing loss was documented for 38.4% (68/177) of the children. Etiology was known for 55.4% (98/177) of children with the most common etiologies being external/middle ear anomalies (15.8% of total), inner ear anomalies (7.9%), syndromes associated with hearing loss (11.9%) and hereditary/genetic causes (8.5%), together accounting for 88.7% of causes (details in Table 1).

Proportion and severity of progressive loss

Overall, 84 of 177 (47.5%) children showed deterioration in hearing in one or both ears from initial diagnosis to most recent assessment. For 63 (35.6%) children, hearing loss remained unilateral with further deterioration in the impaired ear only, and 21 (11.9%) children developed bilateral hearing loss including 14 (7.9%) who showed deterioration in both ears since initial diagnosis and another 7 (4.0%) who developed a loss in the normal hearing ear only. For these 21 (11.9%) children, the loss in the normal hearing ear was identified at a median of 22.1 months (IQR 10.4, 43.3). In summary, 27.7% (98/354) of all ears showed a drop in hearing since initial diagnosis (77 impaired ears plus 21 previously unaffected contralateral ears).

Since this was an early identified cohort, we verified whether there was a difference in progressive hearing loss in children who were identified using objective ABR versus behavioral audiometry at baseline. At the final assessment there were 198 ears with hearing loss (177 impaired ears and 21 ears originally within normal limits), 51.5% (102 of 198) were identified through ABR testing at baseline and 48.5% with behavioral audiometry. Chi-square analysis showed no significant difference in the percentage of ears with progressive hearing loss when the initial diagnostic

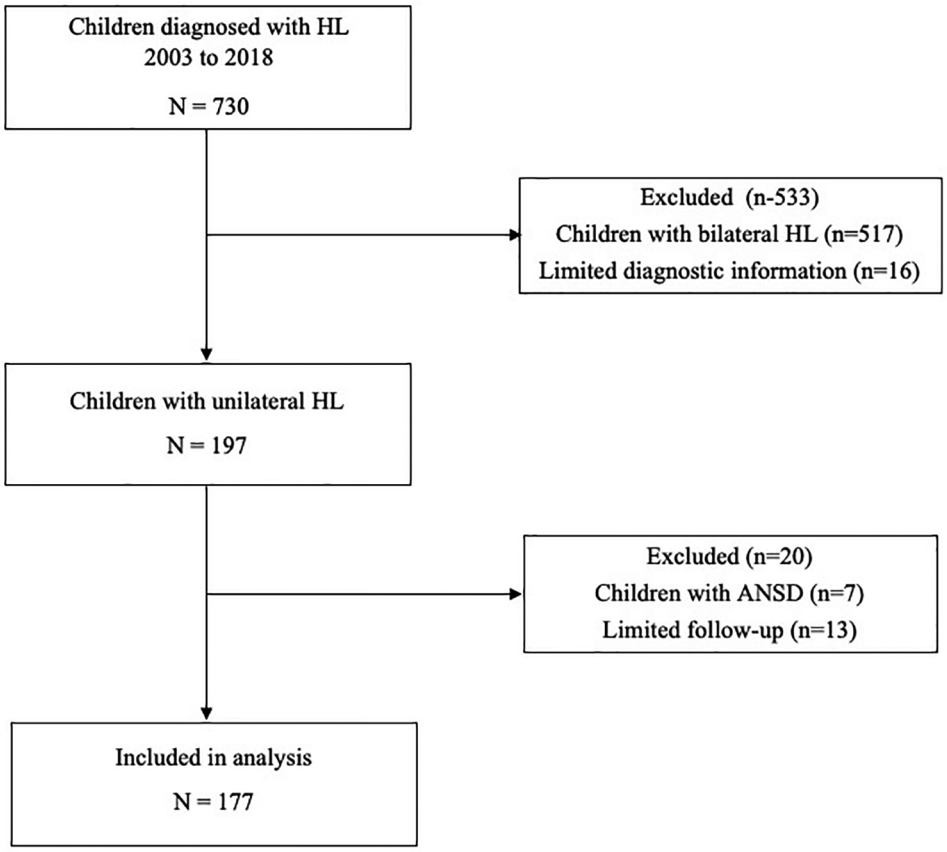

FIGURE 1
Selection of study participants. ANSD, auditory neuropathy spectrum disorder; HL, hearing loss.

assessment was conducted using ABR versus behavioral audiometry [X^2 (1) = 0.866, $p = 0.352$].

The characteristics of the 84 children with progressive hearing loss and the 93 with stable hearing are shown in Table 1. While there was no significant difference in the length of follow-up time for children with progressive loss compared to those with stable loss ($p = 0.072$), the children with progressive loss had more audiologic assessments ($p < 0.001$). As shown, more children with progressive hearing loss had congenital/early onset hearing loss ($p = 0.042$). Children with progressive hearing loss were diagnosed at a median age of 3.7 months (2.2, 44.8) compared to 24.3 months (2.0, 58.9) for those with stable hearing thresholds. It is important to note that children with late onset hearing loss are not necessarily identified at the initial onset of the loss but rather when it becomes severe enough to be noticed, therefore changes in hearing prior to diagnosis are unknown. There was no significant difference in age of diagnosis for children diagnosed with late onset hearing loss in the later 5-year period (2014–2018) compared to those diagnosed in the previous 11 years (2003–2013) ($p = 0.074$). Compared to children with stable hearing, children with progression had more sensorineural/mixed loss ($p = 0.015$) and had less severe hearing loss at diagnosis ($p = 0.013$). The latter finding may reflect that there were more children in the stable group with profound hearing loss at diagnosis in the impaired ear (17 vs. 6), and further deterioration may not have been captured if hearing loss had reached the limits of measured hearing thresholds. When considering only the 154 children with better than profound hearing loss, 50.6% (78/154) showed deterioration in at least one ear.

Severity of hearing loss

Figure 2 shows the average drop in hearing by frequency (0.5–4 kHz) in the impaired ear for the 77 children with deterioration from first to last audiometric assessment. As shown, there was substantial deterioration in hearing across all frequencies. Average deterioration ranged from 27 to 31 dB with little variation across frequencies. For example, at diagnosis, average thresholds ranged from 53 dB HL at 1 kHz to 58 dB HL at 4 kHz and at last assessment from 80 to 86 dB HL. For the 21 children who developed bilateral loss (not shown in Figure 2), 16 (76.2%) children initially presented with high frequency only or mild hearing loss in the previously normal hearing ear; 14 showed further progression in that ear over time.

Figure 3 shows the changes in category of severity of hearing loss for individual ears, classified according to PTA across the four frequencies (0.5, 1, 2, 4 kHz). Changes are shown separately for

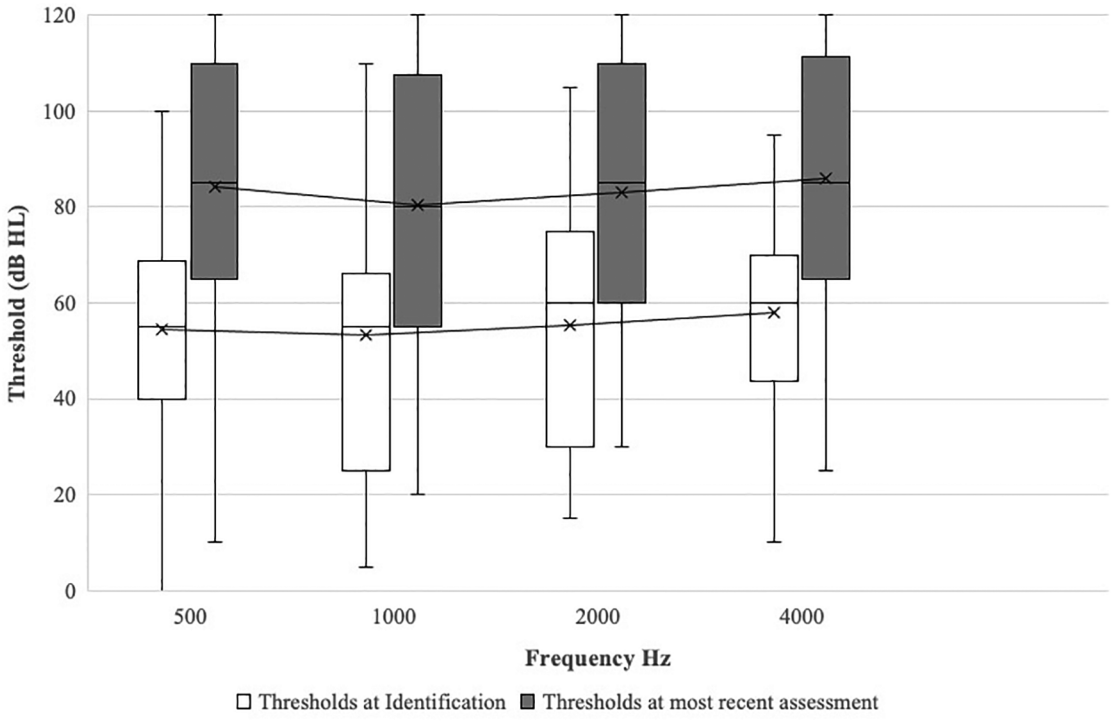

FIGURE 2
Average change in hearing thresholds across frequencies from initial diagnosis to most recent assessment ($n = 77$). The boxes indicate the 25th, 50th, and 75th percentiles. The whiskers above and below the box boundaries show the largest and smallest observed values. x on graph refers to mean thresholds.

the 77 impaired ears and for the 21 ears that started with normal hearing. For the 77 impaired ears, deterioration was sufficient to result in a change in category of hearing loss severity for 67.5% (52/77). For example, 22 ears with mild hearing loss in the impaired ear at diagnosis showed a moderate or worse loss at last assessment and 14 moved from a severe to a profound loss category. For the 21 normal hearing ears, 8 showed a moderate hearing loss or greater at most recent assessment.

By definition, children with progressive loss did not show improvement in hearing levels. Of the 100 impaired ears that were coded as not progressive (stable), 3 showed >10 dB improvement (in 4-frequency PTA) from baseline to most recent assessment (range: 11.4–20.0 dB change). Two of these children had structural conductive hearing loss and all three children continued to present with hearing loss. One normal hearing ear that later developed hearing loss also showed an improvement of 13.8 dB from diagnosis to final audiogram but continued to show a mild loss.

Trajectory of hearing loss

Using the series of audiometric assessments recorded over the first 8 years of follow-up for this cohort (n at baseline of 48, 34, 60, and 50, for 0.5 1, 2 and 4 kHz respectively), we examined the trajectory of hearing loss in the impaired ear to document patterns of changes in hearing over time. Figure 4 shows that most children lost a significant amount of hearing rapidly in the first 4 years of follow-up ($p < 0.001$ for all four frequencies). On average, the loss was estimated at 27.1 dB, 23.1 dB, 24.1 dB, and 22.5 dB at 0.5 1, 2, and 4 kHz respectively. Subsequently, the decrease in thresholds showed a statistically significant stabilization in deterioration for all frequencies ($p < 0.001$) followed by a plateau in the last 4 years of observation.

Relationship between child characteristics and progressive hearing loss

Logistic regression was carried out to assess the association between known clinical characteristics and progressive hearing loss in the 77 impaired ears that showed deterioration. Age at diagnosis and severity of hearing loss (4-frequency PTA) were not significantly associated with progressive/stable hearing loss (Table 2) after adjusting for time since diagnosis. However, the etiologic factors (ENT anomaly-external/middle ear, ENT anomaly-inner ear, syndrome, hereditary/genetic) entered in the model were found to be positively associated with stable hearing loss (i.e., protected against progressive hearing loss). For example, children with hereditary/genetic etiology have a 95% chance of stable hearing loss. However, given the relatively small number of children in some etiologic categories, the results should be interpreted with caution. After applying a Bonferroni correction [adjusted alpha level of 0.001—(.05/35)], only the factor ENT anomaly-external/middle ear remained statistically significant.

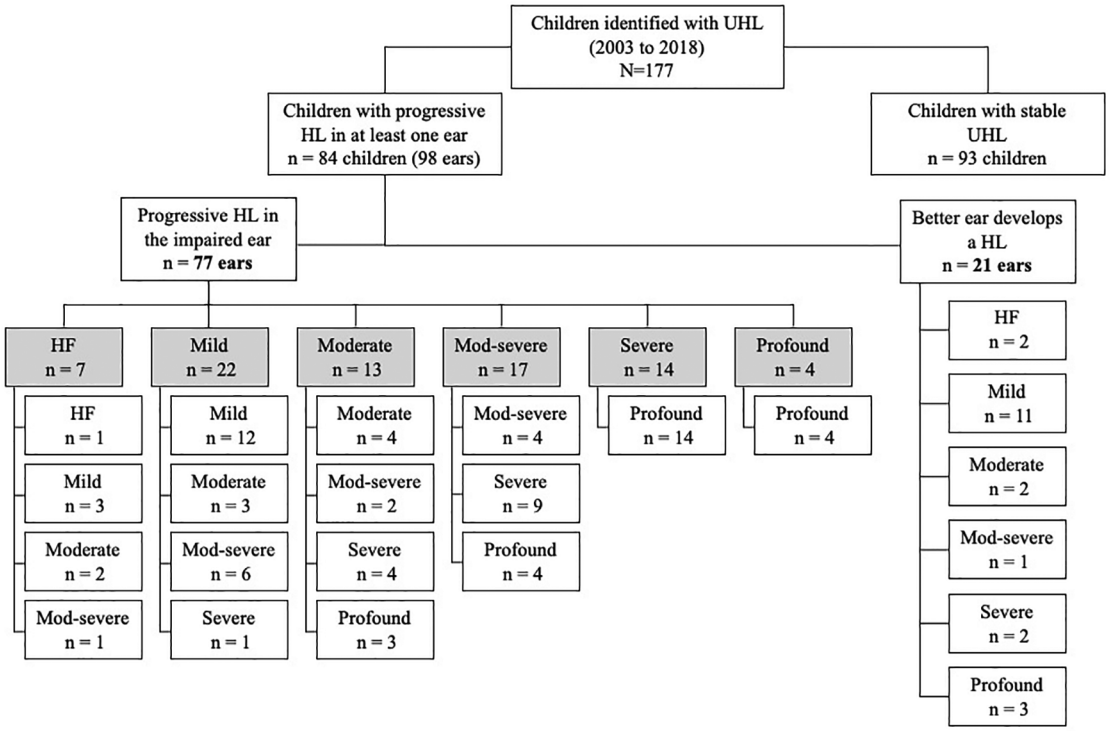

FIGURE 3
Category of hearing loss severity at diagnosis and at most recent assessment. The shaded boxes represent the degree of hearing loss in the impaired ear at diagnosis and the unshaded boxes the degree at final assessment. For the 21 ears that developed hearing loss (previously normal hearing), degree of hearing loss at final assessment is shown. UHL, unilateral hearing loss; HL, hearing loss.

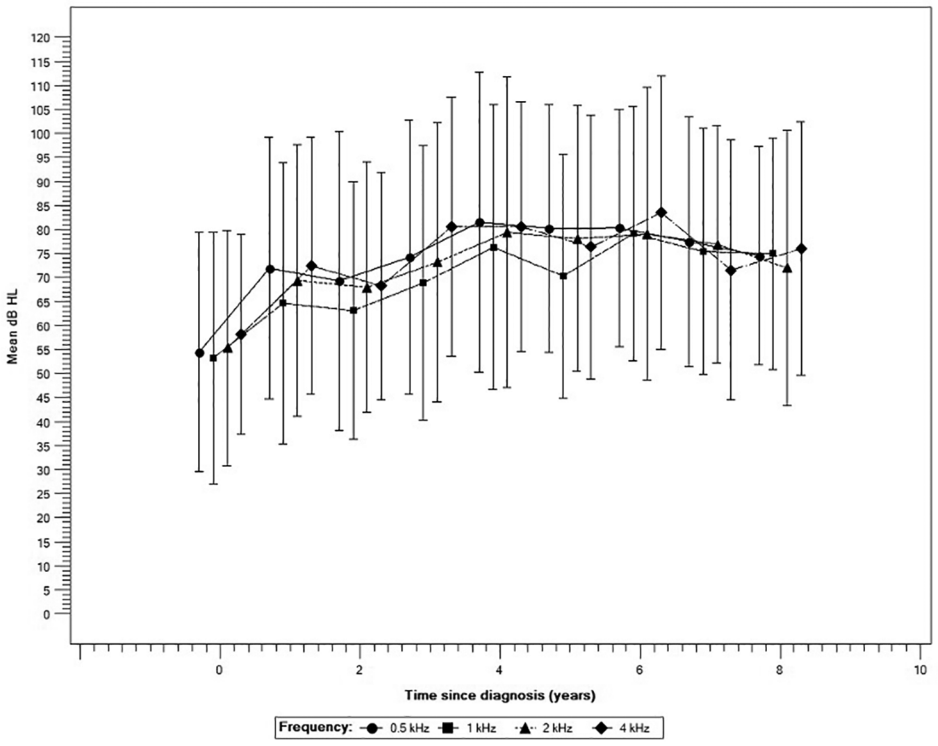

FIGURE 4
Trajectory of hearing loss across four frequencies (n = 77). Error bars represent one standard deviation.

TABLE 2 Factors associated with progressive hearing loss ($n = 77$).

Factor	Adjusted odds ratio (95% CI)	*p*-value
Age at diagnosis	1.00 (0.98, 1.02)	0.733
Severity (PTA) at baseline	0.98 (0.96, 1.00)	0.103
Etiologic factors		
ENT anomaly-external/middle ear	0.01 (0.00, 0.13)	<0.001
ENT anomaly-inner ear	0.08 (0.01, 0.98)	0.049
Syndrome (associated with HL)	0.08 (0.01, 0.93)	0.044
Hereditary/genetic	0.05 (0.00, 0.71)	0.026
CMV[a]	n/a	n/a

CMV, cytomegalovirus; ENT, ear, nose, throat; HL, hearing loss; PTA, pure-tone average.
[a]Insufficent number of participants for the regression model; 1 of 8 with CMV had stable hearing loss.

Using the same variables and adjusting for time since diagnosis, linear regression models were fit to examine the association between clinical characteristics and the total amount of deterioration in hearing at individual frequencies 0.5–4 kHz (**Table 3**). Age at diagnosis was not a significant predictor of change in hearing except at 0.5 kHz, where younger age at diagnosis was associated with more deterioration in hearing. This difference was small, translating to 2.2 dB more deterioration in the threshold at 0.5 kHz threshold when a child was diagnosed at age 12 months compared to 24 months. Applying a Bonferroni correction, (adjusted alpha level of 0.001) the result would no longer be statistically significant. There was no significant association between any etiology and amount of deterioration in hearing at any frequency.

Discussion

This population-based study showed that about 1 in 4 children with permanent hearing loss present with UHL at initial diagnosis. Based on a large dataset of longitudinal audiometric data, we found that almost half (47.5%, $n = 84$ of 177) of children first diagnosed with UHL experienced deterioration in hearing in the impaired ear or developed a hearing loss in the normal hearing ear. While deterioration for most children was limited to the impaired ear (43.5% of all children) 11.9% also developed bilateral hearing loss. We observed a trend towards a greater drop in hearing in the first 4 years after diagnosis with the decrease slowing over time and a plateau effect noted in the next 4 years.

Our overall findings related to the proportion of children who experience changes in hearing are consistent with our previous report on 330 children across the spectrum of hearing loss, both unilateral and bilateral loss (24). In that study 48% showed some amount of deterioration in hearing over time, including 37% of the 73 children with UHL. Almost half of all children had more than 20 dB drop in average hearing levels. In a subsequent study, we found that 42% of children with UHL showed deterioration, including 17% who developed bilateral loss (2). However, in both these studies we limited our analyses to a comparison of initial diagnostic and most recent audiologic results to determine progression. A study from another Canadian center reported that about one-third of 128 children with UHL showed progression (29). The current study adds another contribution to our understanding in detailing the trajectory of hearing loss. Through our analysis of multiple audiograms, we mapped out trends for children with UHL across a span of 8 years.

Severity

The amount of change in hearing loss is important in planning optimal audiologic management of these children. During the study period, children lost an average of about 30 decibels across the individual speech frequencies (impaired ear) and more than two thirds of the deterioration happened over the first 4 years post-diagnosis (average of over 20 dB decrease in thresholds). These are clinically important changes. For example, for the 77 children with impaired ears that showed progressive hearing loss, the drop in average hearing levels was sufficient to result in two-thirds (67.4%) of them being reclassified to a more severe category of hearing loss at last audiometric assessment. This resulted in an almost doubling of the number of children with severe or profound hearing loss, in at least one ear (39 ears vs. 18 at diagnosis). Furthermore, 11.9% of children developed bilateral hearing loss placing them at greater risk for delays in auditory and communication development. Of the

TABLE 3 Factors associated with amount of deterioration in hearing across frequencies ($n = 77$).

	500 Hz		1,000 Hz		2,000 Hz		4,000 Hz	
	Coefficient[a]	*p*-value	Coefficient	*p*-value	Coefficient	*p*-value	Coefficient	*p*-value
Age at diagnosis (mos)	−0.18 (−0.35, −0.02)	0.028	−0.09 (−0.23, 0.06)	0.218	−0.01 (−0.21, 0.01)	0.095	−0.01 (−0.13, 0.11)	0.905
Severity (PTA) at baseline	0.01 (−0.26, 0.28)	0.935	0.10 (−0.13, 0.33)	0.383	−0.01 (−0.19, 0.16)	0.879	0.14 (−0.08, 0.35)	0.215
Etiology factors								
ENT anomaly-external/middle ear	−18.96 (−41.70, 3.78)	0.100	N/A		−14.18 (−46.90, 18.53)	0.388	−4.44 (−27.18, 18.31)	0.696
ENT anomaly-inner ear	0.47 (−24.80, 25.73)	0.971	−7.41 (−32.72, 17.90)	0.556	6.93 (−7.36, 21.22)	0.335	6.17 (−12.95, 25.29)	0.518
Syndrome (associated with HL)	−11.71 (−28.53, 5.11)	0.167	3.00 (−14.16, 20.16)	0.725	1.54 (−11.25, 14.32)	0.810	−2.25 (−16.71, 12.21)	0.755
Hereditary/genetic	8.25 (−33.12, 49.62)	0.689	4.30 (−14.12, 22.72)	0.638	0.62 (−13.66, 14.91)	0.930	−4.04 (−17.53, 9.45)	0.549
CMV	8.48 (−12.19, 29.15)	0.412	7.63 (−11.68, 26.93)	0.428	8.90 (−6.91, 24.71)	0.264	5.18 (−13.41, 23.77)	0.577

CMV, cytomegalovirus; ENT, ear, nose, throat; HL, hearing loss; PTA, pure-tone average.
[a]In the table, coefficient refers to the difference in hearing level (at each frequency) for every 1 unit change in the factor examined (independent variable, e.g., age, severity). In the model, only age at diagnosis at 500 Hz was significant, i.e., for each month later age of diagnosis, there was a very slight improvement in hearing. For the etiologic (categorical) factors, the coefficient represents the amount of change in hearing level when the etiologic factor is not present vs. present. In the model, no coefficients were statistically significant, i.e., no factors were associated with a difference in hearing level at any frequency.

21 normal hearing ears that developed a loss, 5 showed severe or profound hearing loss by study end.

The increase in severity of hearing loss in one or both ears is an important finding for two reasons. First, severity of hearing loss has important implications for the type of technology that these children are likely to require. Current guidelines generally support the use of conventional hearing aids for children with UHL who present with less than severe hearing loss (14, 16). For children with hearing aids, recommendations and counseling related to use may change with greater hearing loss severity. In addition, management options for children with severe to profound hearing loss, commonly referred to as single-sided deafness, have evolved in recent years with more children now considered for cochlear implants (15, 52–54). Changes in hearing might lead to different hearing technology (e.g., cochlear implant) and management options being considered for about one-quarter of the children (24.9%, $n = 44$) in our study compared to 10.2% at initial diagnosis. A recent review primarily based on adult UHL studies suggests that early cochlear implantation can prevent or reduce auditory deprivation in individuals with UHL (55). Secondly, there is some evidence from a systematic review to indicate that children with severe and profound UHL have more difficulty than those with less severe loss in at least some aspects of speech and language development (11). Earlier awareness of the presence of more severe loss may result in the fitting of optimal technology and provision of speech-language intervention in a timely manner, therefore, careful monitoring of these children would seem to be warranted.

Trajectory

Knowing about the trajectory of hearing loss and any change in audiometric profiles over time can provide useful information for parents and can underscore the importance of monitoring their child's hearing. It can also be useful for clinicians and decision-makers in establishing appropriate clinical follow-up protocols. Our longitudinal analysis showed that the most important changes in hearing levels were observed in the first 4 years. Hearing continued to decrease over time but at a slower rate and the drop was much less pronounced 5–8 years after diagnosis. It is of clinical importance that most children did not experience sudden "large" drops in hearing but a more progressive, gradual decrease over time. These small changes in hearing thresholds are likely not noticeable by parents or therapists who see the children in everyday environments, especially since most continue to have one ear with normal hearing. When hearing loss drops suddenly, services may be initiated quickly. In contrast, our findings of more gradual progression indicate the need for close surveillance of hearing in these children in the first few years after diagnosis. Greater awareness about the possibility of worse hearing in one ear or the development of hearing loss in the contralateral ear can be valuable in guiding the families of these young children. For example, our previous research has shown that hearing aid use in the preschool years tends to be lower in these children, even when compared with mild bilateral loss (56). Timely information about a change in hearing may help parents decide to move forward with recommendations for amplification or motivate them to increase their child's hearing aid use. It is possible that concrete information about the evolution of UHL may influence parents' decisions early in their child's life and potentially prevent or reduce delays in later childhood.

Factors

Predicting who is most likely to lose further hearing would also be helpful in guiding families and in establishing clinical protocols. However, our examination of factors showed no clear relationship with age or severity of hearing loss at diagnosis. Our analysis of the available etiologic factors showed only that all were associated with stable hearing loss. In a previous study investigating risk factors (24), we also found a positive association between structural conductive conditions (e.g., atresia) and stable hearing loss. In that research, there was no relationship between any other risk factors and progressive/stable hearing loss. Dahl et al. (40) also did not find a relationship between severity of hearing loss or etiology and progressive loss over the first 3 years of life. It is important to note that our study was conducted prior to the implementation of cCMV or systematic genetic screening in the hearing program, resulting in almost half of the sample having unknown etiology. With further expansion of molecular screening in population-based NHS to detect infants at risk for late onset or progressive loss, more comprehensive analyses in sub-populations of children may eventually shed further light on progressive hearing loss (32, 57). In our study, we also could not show any relationship between age at diagnosis and the magnitude of deterioration in hearing levels across the speech frequencies except a small difference at 0.5 Hz, where a diagnosis 12 months earlier resulted in a loss of 2.2 dB more hearing per year. The etiologies examined also had no significant impact on the amount of deterioration in hearing.

Limitations

A strength of this study is access to a population-based cohort in a publicly funded, health care system with comprehensive medical chart data available. Well-established diagnostic and follow-up protocols were in place in the clinic. Clinical characteristics and initial audiometric information were collected prospectively as children were diagnosed. However, our study has some limitations. A comparison of early audiologic (ABR and behavioral thresholds) and later behavioral assessments can introduce some error. Although we used estimated behavioral thresholds (eHL) to document ABR results, the agreement with behavioral thresholds is not perfect and there is some evidence that predicted behavioral thresholds may be underestimated in children with moderate and greater degrees of hearing loss (49, 58). In addition, behavioral threshold responses obtained for infants and young children, with normal hearing, particularly for VRA, are likely to be in the 20 dB HL range and become lower

as their age increases (59, 60). This could lead to some underestimation of the number of children who experienced deterioration in hearing.

Our study depended on clinical audiologic data and despite clear follow-up protocols, the nature of clinical management is that assessments do not always follow the planned schedule. For young children, assessment results may be incomplete or require several test sessions. Furthermore, compared to children with bilateral hearing loss, audiologic follow-up for these children may be less consistent due to less concern on the part of parents about communication development, less frequent intervention sessions, and higher levels of amplification non-use. It is possible that children are less likely to present for follow-up visits if there is no concern. In addition, children in this study were diagnosed over a 16-year period and had variable lengths of follow-up. While we controlled for time since diagnosis in the regression analyses, this resulted in a smaller sample size for the longitudinal analysis of trajectory of hearing loss and requires that these results be interpreted with caution. Finally, the lack of specific etiologic data (e.g., based on radiologic findings or cCMV screening) and the relatively small number of children in some etiology groups precluded more extensive analyses of conditions (e.g., cCMV, enlarged vestibular aqueduct) previously reported to be associated with progressive hearing loss (37, 39, 41).

Conclusion

Early identified children with UHL represent a new clinical population in the last 20–30 years since the widespread implementation of NHS. An important goal of screening is to improve developmental outcomes for children with hearing loss of any degree. Using population-level data to track the evolution of hearing loss, this study provides evidence that almost half of the children with UHL are at risk for further deterioration in hearing in the impaired ear or for bilateral loss especially in the first 4 years after diagnosis. The extent of the problem and the magnitude of the hearing deterioration, coupled with the potential impact on intervention decisions seem to justify efforts to regularly monitor these children to identify additional needs as early as possible.

Data availability statement

The datasets presented in this article are not readily available because Data are not available outside of the research team as per Ethics approvals. Requests to access the datasets should be directed towards the corresponding author.

Ethics statement

The studies involving human participants were reviewed and approved by Research Ethics Committees at the CHEO Research Institute (file #09-64X), and the University of Ottawa (file #H10-09-11). Written informed consent from the participants' legal guardian/next of kin was not required to participate in this study in accordance with the national legislation and the institutional requirements.

Author contributions

EF conceived the overall project. EF, IG, AD-S, DC, JO, MP, and JW developed the methods and procedures and provided input throughout the study. JW, FN, and BV managed data collection and data entry. IG oversaw and conducted statistical analysis and interpretation of the analyses. JW assisted with the statistical analysis. EF drafted the first version of the manuscript. All authors contributed to the article and approved the submitted version.

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Conflict of interest

The authors declare that the research was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

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Publisher's note

References

1. Morton CC, Nance WE. Newborn hearing screening-a silent revolution. *New Eng J Med*. (2006) 354(20):2151–64. doi: 10.1056/NEJMra050700

2. Fitzpatrick EM, Al-Essa RS, Whittingham J, Fitzpatrick J. Characteristics of children with unilateral hearing loss. *Int J Audiol*. (2017) 56(11):819–28. doi: 10.1080/14992027.2017.1337938

3. Fitzpatrick EM, Whittingham J, Durieux-Smith A. Mild bilateral and unilateral hearing loss in children: a 20 year view of characteristics and practices. *Ear Hear*. (2014) 35(1):10–8. doi: 10.1097/AUD.0b013e31829e1ed9

4. Ghogomu N, Umansky A, Lieu JE. Epidemiology of unilateral sensorineural hearing loss with universal newborn hearing screening. *Laryngoscope*. (2014) 124 (1):295–300. doi: 10.1002/lary.24059

5. Prieve BA, Stevens F. The New York state universal newborn hearing screening demonstration project: introduction and overview. *Ear Hear*. (2000) 21(2):85–91. doi: 10.1097/00003446-200004000-00006

6. Johnson JL, White KR, Widen JE, Gravel JS, James M, Kennalley T. A multicenter evaluation of how many infants with permanent hearing loss pass a two-stage otoacoustic emissions/automated auditory brainstem response newborn hearing screening protocol. *Pediatrics*. (2005) 116(3):663–72. doi: 10.1542/peds.2004-1688

7. Kishon-Rabin L, Kuint J, Hildesheimer M, Ari-Even Roth D. Delay in auditory behaviour and preverbal vocalization in infants with unilateral hearing loss. *Dev Med Child Neurol*. (2015) 57(12):1129–36. doi: 10.1111/dmcn.12812

8. Fitzpatrick EM, Gaboury I, Durieux-Smith A, Coyle D, Whittingham J, Nassrallah F. Auditory and language outcomes in children with unilateral hearing loss. *Hear Res*. (2019) 372:42–51. doi: 10.1016/j.heares.2018.03.015

9. Purcell PL, Cushing SL, Papsin BC, Gordon KA. Unilateral hearing loss and single-sided deafness in children: an update on diagnosis and management. *Cur Otorhinolaryngol Rep*. (2020) 8(3):259–66. doi: 10.1007/s40136-020-00293-8

10. Lieu JEC. Permanent unilateral hearing loss (UHL) and childhood development. *Cur Otorhinolaryngol Rep*. (2018) 6(1):74–81. doi: 10.1007/s40136-018-0185-5

11. Anne S, Lieu JEC, Cohen MS. Speech and language consequences of unilateral hearing loss: a systematic review. *Otolaryngol Head Neck Surg*. (2017) 157(4):572–9. doi: 10.1177/0194599817726326

12. Nassrallah F, Fitzpatrick EM, Whittingham J, Sun H, Na E, Grandpierre V. A descriptive study of language and literacy skills of early school-aged children with unilateral and mild to moderate bilateral hearing loss. *Deafness Educ Int*. (2020) 22 (1):74–92. doi: 10.1080/14643154.2018.1555119

13. Lieu JE, Tye-Murray N, Karzon RK, Piccirillo JF. Unilateral hearing loss is associated with worse speech-language scores in children. *Pediatrics*. (2010) 125(6): e1348–55. doi: 10.1542/peds.2009-2448

14. Bagatto M, DesGeorges J, King A, Kitterick P, Laurnagaray D, Lewis D, et al. Consensus practice parameter: audiological assessment and management of unilateral hearing loss in children. *Int J Audiol*. (2019) 58(12):805–15. doi: 10.1080/14992027.2019.1654620

15. Appachi S, Specht JL, Raol N, Lieu JEC, Cohen MS, Dedhia K, et al. Auditory outcomes with hearing rehabilitation in children with unilateral hearing loss: a systematic review. *Otolaryngol Head Neck Surg*. (2017) 157(4):565–71. doi: 10.1177/0194599817726757

16. Fitzpatrick EM, Cologrosso E, Sikora L. Candidacy for amplification in children with hearing loss: a review of guidelines and recommendations. *Am J Audiol*. (2019) 28(4):1025–45. doi: 10.1044/2019_aja-19-0061

17. Laugen NJ, Erixon E, Huttunen K, Mäki-Torkko E, Löfkvist U. Newborn hearing screening and intervention in children with unilateral hearing impairment: clinical practices in three nordic countries. *J Clin Med*. (2021) 10(21):5152. doi: 10.3390/jcm10215152

18. Fitzpatrick EM, Grandpierre V, Durieux-Smith A, Gaboury I, Coyle D, Na E, et al. Children with mild bilateral and unilateral hearing loss: arents' reflections on experiences and outcomes. *J Deaf Stud Deaf Educ*. (2016) 21(16):34–43. doi: 10.1093/deafed/env047

19. Grandpierre V, Fitzpatrick EM, Na E, Mendonca O. School-aged children with mild bilateral and unilateral hearing loss: parents' reflections on services, experiences, and outcomes. *J Deaf Studies Deaf Edu*. (2018) 23(2):140–7. doi: 10.1093/deafed/enx049

20. Reeve K. *Amplification and family factors for children with mild and unilateral hearing impairment. National workshop on mild and unilateral hearing loss: workshop proceedings*; Brekenridge, CO: Centers for Disease Control and Prevention (2005).

21. Wood SA, Sutton GJ, Davis AC. Performance and characteristics of the newborn hearing screening programme in England: the first seven years. *Int J Audiol*. (2015) 54:353–8. doi: 10.3109/14992027.2014.989548

22. Joint Committee on Infant Hearing. Year 2019 position statement: principles and guidelines for early hearing detection and intervention programs. *J Early Hearing Det Int*. (2019) 4(2):1–44. doi: 10.15142/fptk-b748

23. Hyde ML. Newborn hearing screening programs: overview. *J Otolaryngol*. (2005) 34(Suppl 2):S70–8. PMID: 16076420.

24. Barreira-Nielsen C, Fitzpatrick EM, Hashem S, Whitingham J, Barrowman N, Aglipay M. Progressive hearing loss in early childhood. *Ear Hear*. (2016) 37(5): e311–21. doi: 10.1097/AUD.0000000000000325

25. Kenna MA, Feldman HA, Neault MW, Frangulov A, Wu B-L, Fligor B, et al. Audiologic phenotype and progression in GJB2 (connexin 26) hearing loss. *Arch Otolaryngol Head Neck Surg*. (2010) 136(1):81–7. doi: 10.1001/archoto.2009.202

26. Watkin P, Baldwin M. The longitudinal follow up of a universal neonatal hearing screen: the implications for confirming deafness in childhood. *Int J Audiol*. (2012) 51 (7):519–28. doi: 10.3109/14992027.2012.673237

27. Martinez-Cruz CF, Poblano A, Garcia-Alonso Themann P. Changes in tonal audiometry in children with progressive sensorineural hearing loss and history of neonatal intensive care unit discharge. A 20 year long-term follow-up. *Int J Pediatr Otorhinolaryngol*. (2017) 101:235–40. doi: 10.1016/j.ijporl.2017.08.022

28. Acke FRE, De Vriese C, Van Hoecke H, De Leenheer EMR. Twelve years of neonatal hearing screening: audiological and etiological results. *Eur Arch Otorhinolaryngol*. (2022) 279(7):3371–8. doi: 10.1007/s00405-021-07060-5

29. Purcell PL, Shinn JR, Coggeshall SS, Phillips G, Paladin A, Sie KCY, et al. Progression of unilateral hearing loss in children with and without ipsilateral cochlear nerve canal stenosis: a hazard analysis. *Otolo Neurotol*. (2017) 38(6): e138–e44. doi: 10.1097/mao.0000000000001452

30. Paul A, Marlin S, Parodi M, Rouillon I, Guerlain J, Pingault V, et al. Unilateral sensorineural hearing loss: medical cntext and etiology. *Audiol Neurootol*. (2017) 22 (2):83–8. doi: 10.1159/000474928

31. Wang J, Sung V, Carew P, Burt RA, Liu M, Wang Y, et al. Prevalence of childhood hearing loss and secular trends: a systematic review and meta-analysis. *Acad Pediatr*. (2019) 19(5):504–14. doi: 10.1016/j.acap.2019.01.010

32. Mitchell CO, Morton CC. Genetics of childhood hearing loss. *Otolaryngol Clin North Am*. (2021) 54(6):1081–92. doi: 10.1016/j.otc.2021.08.008

33. Mehta D, Noon SE, Schwartz E, Wilkens A, Bedoukian EC, Scarano I, et al. Outcomes of evaluation and testing of 660 individuals with hearing loss in a pediatric genetics of hearing loss clinic. *Am J Med Genet A*. (2016) 170 (10):2523–30. doi: 10.1002/ajmg.a.37855

34. van Beeck Calkoen EA, Engel MSD, van de Kamp JM, Yntema HG, Goverts ST, Mulder MF, et al. The etiological evaluation of sensorineural hearing loss in children. *Eur J Pediatr Audiol*. (2019) 178(8):1195–205. doi: 10.1007/s00431-019-03379-8

35. Nance WE, Lim BG, Dodson KM. Importance of congenital cytomegalovirus infections as a cause for pre-lingual hearing loss. *J Clin Virol*. (2006) 35(2):221–5. doi: 10.1016/j.jcv.2005.09.017

36. Fowler KB. Congenital cytomegalovirus infection: audiologic outcome. *Clin Infect Dis*. (2013) 57(Suppl 4):S182–4. doi: 10.1093/cid/cit609

37. Goderis J, De Leenheer E, Smets K, Van Hoecke H, Keymeulen A, Dhooge I. Hearing loss and congenital CMV infection: a systematic review. *Pediatrics*. (2014) 134(5):972–82. doi: 10.1542/peds.2014-1173

38. Fletcher KT, Horrell EMW, Ayugi J, Irungu C, Muthoka M, Creel LM, et al. The natural history and rehabilitative outcomes of hearing loss in congenital cytomegalovirus: a systematic review. *Otol Neurotol*. (2018) 39(7):854–64. doi: 10.1097/mao.0000000000001861

39. Lanzieri TM, Chung W, Flores M, Blum P, Caviness AC, Bialek SR, et al. Hearing loss in children with asymptomatic congenital cytomegalovirus infection. *Pediatrics*. (2017) 139(3):e20162610. doi: 10.1542/peds.2016-2610

40. Dahl HH, Ching TY, Hutchison W, Hou S, Seeto M, Sjahalam-King J. Etiology and audiological outcomes at 3 years for 364 children in Australia. *PLoS One*. (2013) 8 (3):e59624. doi: 10.1371/journal.pone.0059624

41. Alemi AS, Chan DK. Progressive hearing loss and head trauma in enlarged vestibular aqueduct: a systematic review and meta-analysis. *Otolaryngol Head Neck Surg*. (2015) 153(4):512–7. doi: 10.1177/0194599815596343

42. Joint Committee on Infant Hearing. Year 2007 position statement: principles and guidelines for early hearing detection and intervention. *Pediatrics*. (2007) 120:898–921. doi: 10.1542/peds.2007-2333

43. Acke FRE, Van Hoecke H, De Leenheer EMR. Congenital unilateral hearing loss: characteristics and etiological analysis in 121 patients. *Otol Neurotol*. (2021) 42 (9):1375–81. doi: 10.1097/mao.0000000000003248

44. Ontario ministry of children and youth services ontario infant hearing program. Ontario infant hearing program: a guidance document, version 2017.01. 2017.

45. National Workshop on Mild and Unilateral Hearing Loss. Workshop Proceedings. Breckenridge, CO: Centers for Disease Control and Prevention (2005).

46. Stapells D. Threshold estimation by the tone-evoked auditory brainstem response: a literature meta-analysis. *J Speech Lang Pathol Audiol*. (2000) 24:74–83.

47. Ministry of Children, Community and Social Services: Ontario Infant Hearing Program. Protocol for auditory brainstem response-based audiological assessment (ABRA), guidance version 2018.01. (2018).

48. Vander Werff KR, Prieve BA, Georgantas LM. Infant air and bone conduction tone burst auditory brain stem responses for classification of hearing loss and the relationship to behavioral thresholds. *Ear Hear*. (2009) 30:350–68. doi: 10.1097/AUD.0b013e31819f3145

49. McCreery RW, Kaminski J, Beauchaine K, Lenzen N, Simms K, Gorga MP. The impact of degree of hearing loss on auditory brainstem response predictions of behavioral thresholds. *Ear Hear*. (2015) 36(3):309–19. doi: 10.1097/AUD.0000000000000120

50. Little RJ, Rubin DB. *Statistical analysis with missing data*. 3rd ed. Hoboken, NJ: John Wiley & Sons (2019). 462.

51. van Buuren S. *Flexible imputation of missing data*. Boca Raton, FL: CRC Press, Taylor & Francis Group (2012). 342.

52. Arndt S, Prosse S, Laszig R, Wesarg T, Aschendorff A, Hassepass F. Cochlear implantation in children with single-sided deafness: does aetiology and duration of deafness matter? *Audiol Neurootol*. (2015) 20(Suppl 1):21–30. doi: 10.1159/000380744

53. Peters JP, Ramakers GG, Smit AL, Grolman W. Cochlear implantation in children with unilateral hearing loss: a systematic review. *Laryngoscope*. (2016) 126 (3):713–21. doi: 10.1002/lary.25568

54. Benchetrit L, Ronner EA, Anne S, Cohen MS. Cochlear implantation in children with single-sided deafness: a systematic review and meta-analysis. *JAMA Otolaryngol Head ANeck Surg*. (2021) 147(1):58–69. doi: 10.1001/jamaoto.2020.3852

55. Vanderauwera J, Hellemans E, Verhaert N. Research insights on neural effects of auditory deprivation and restoration in unilateral hearing loss: a systematic review. *J Clin Med*. (2020) 9(3):812. doi: 10.3390/jcm9030812

56. Fitzpatrick EM, Gaboury I, Durieux-Smith A, Coyle D, Whittingham J, Salamatmanesh M, et al. Parent report of amplification use in children with mild bilateral or unilateral hearing loss. *J Am Acad Audiol*. (2019) 30(2):93–102. doi: 10.3766/jaaa.17020

57. Shearer AE, Shen J, Amr S, Morton CC, Smith RJ. A proposal for comprehensive newborn hearing screening to improve identification of deaf and hard-of-hearing children. *Genet Med*. (2019) 21(11):2614–30. doi: 10.1038/s41436-019-0563-5

58. Lee CY, Jaw FS, Pan SL, Hsieh TH, Hsu CJ. Effects of age and degree of hearing loss on the agreement and correlation between sound field audiometric thresholds and tone burst auditory brainstem response thresholds in infants and young children. *J Formos Med Assoc*. (2008) 107:869–75. doi: 10.1016/S0929-6646(08)60203-X

59. Widen JE, Folsom RC, Cone-Wesson B, Carty L, Dunnell JJ, Koebsell K, et al. Identification of neonatal hearing impairment: hearing status at 8 to 12 months corrected age using a visual reinforcement audiometry protocol. *Ear Hear*. (2000) 21(5):471–87. doi: 10.1097/00003446-200010000-00011

60. Hulecki L, Small S. Behavioral bone-conduction thresholds for infants with normal hearing. *J Am Acad Audiol*. (2011) 22:81–92. doi: 10.3766/jaaa.22.2.3

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EDITED BY
Elizabeth Fitzpatrick,
University of Ottawa, Canada

REVIEWED BY
Ryan William McCreery,
Boys Town National Research Hospital, United States
Martijn Agterberg,
Radboud University, Netherlands

*CORRESPONDENCE
Sally K. Thornton
✉ sally.thornton@nottingham.ac.uk

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Characterisation of the treatment provided for children with unilateral hearing loss

Roshni Patel[1], Derek J. Hoare[1,2], Karen R. Willis[3], Shammas Tabraiz[1], Paul K. Bateman[1,2] and Sally K. Thornton[1,2]*

[1]Hearing Sciences, Mental Health and Clinical Neurosciences, School of Medicine, The University of Nottingham, Nottingham, United Kingdom, [2]NIHR Nottingham Biomedical Research Centre, Nottingham University Hospitals NHS Trust, Nottingham, United Kingdom, [3]Children's Audiology, Ropewalk House, Nottingham, United Kingdom

Background: Children with permanent unilateral hearing loss (UHL) are an understudied population, with limited data to inform the guidelines on clinical management. There is a funding gap in healthcare provision for the children with UHL in the United Kingdom, where genetic screening, support services, and devices are not consistently provided or fully funded in all areas. They are a disparate population with regard to aetiology and their degree of hearing loss, and hence their device choice and use. Despite having one "good ear", some children with UHL can have similar outcomes, socially, behaviourally, and academically, to children with bilateral hearing loss, highlighting the importance of understanding this population. In this longitudinal cohort study, we aimed to characterise the management of the children with UHL and the gaps in the support services that are provided for the children in Nottingham, United Kingdom.
Methods: A cohort study was conducted collecting longitudinal data over 17 years (2002–2019) for 63 children with permanent congenital confirmed UHL in a large tertiary regional referral centre for hearing loss in Nottingham, United Kingdom. The cases of UHL include permanent congenital, conductive, mixed, or sensorineural hearing loss, and the degree of hearing loss ranges from mild to profound. The data were taken from their diagnostic auditory brainstem responses and their two most recent hearing assessments. Descriptors were recorded of the devices trialled and used and the diagnoses including aetiology of UHL, age of first fit, degree of hearing loss, when and which type of device was used, why a device was not used, the support services provided, concerns raised, and who raised them.
Results: Most children (45/63; 71%) trialled a device, and the remaining 18 children had no device trial on record. Most children (20/45; 44%) trialled a bone-conduction device, followed by contralateral routing of signal aid (15/45; 33%) and conventional hearing aids (9/45; 20%). Most children (36/45; 80%) who had a device indicated that they wore their device "all day" or every day in school. Few children (8/45; 18%) reported that they wore their device rarely, and the reasons for this included bullying (3/8), feedback from the device (2/8), and discomfort from the device (2/8). Only one child reported that the device was not helping with their hearing. The age that the children were first fitted with their hearing device varied a median of 2.5 years for hearing aids and bone-conduction devices and 7 years for a contralateral routing of signal aid. The length of time that the children had the device also varied widely (median of 26 months, range 3–135 months); the children had their bone-conduction hearing aid for the longest period of time (median of 32.5 months). There was a significant trend where more recent device fittings were happening for children at a younger age. Fifty-one children were referred by the paediatric audiologist to a support service, 72.5% (37/51) were subsequently followed up by the

referred service with no issue, whilst the remaining 27.5% (14/51) encountered an issue leading to an unsuccessful provision of support. Overall, most children (65%, 41/63) had no reported concerns, and 28.5% (18/63) of the children went on to have a documented concern at some point during their audiological care: five with hearing aid difficulties, five with speech issues, four with no improvement in hearing, three facing self-image or bullying issues, and one case of a child struggling to interact socially with friends. Three of these children had not trialled a device. We documented every concern reported from the parents, clinicians, teachers of the deaf, and from the children themselves. Where concerns were raised, more than half (58.6%, 10/18) were by schools and teachers, the remaining four concerns were raised by the family, and further four concerns were raised by the children themselves.

Conclusion: To discover what management will most benefit which children with permanent UHL, we first must characterise their treatment, their concerns, and the support services available for them. Despite the children with UHL being a highly disparate population—in terms of their aetiology, their device use, the degree of hearing loss, and the age at which they trial a device—the majority report they use their device mostly in school. In lieu of available data and in consideration of the devices that are available to them, it could be useful to support families and clinicians in understanding the devices which are most used and where they are used. Considering the reasons for cessation of regular device use counselling and support services would be vital to support the children with UHL.

KEYWORDS

unilateral hearing loss (UHL), children, hearing aids, contralateral routing of signal (CROS), bone-conduction device (BCD)

Introduction

Within the United Kingdom (England, Northern Ireland, Wales, and Scotland), there is a current lack of national management guidelines for clinicians looking after children with unilateral hearing loss (UHL), and treatment is widely debated (1–3). Numerous organisations have established candidacy guidelines for paediatric amplification for the children with UHL (see **Supplementary Material**) and there is considerable variation in the guidelines with candidacy criteria ranging from 15 to 30 dB HL (4). Air or bone-conduction devices (BCDs) or cochlear implants (CIs) may be recommended at the upper limit of candidacy, for example, in single-sided deafness (SSD) where amplification is not possible, and for the children with UHL, many recommend a case-by-case decision approach (1, 46–48).

Uptake of devices for UHL can be variable with as few as 31% of children (9–18 years) with UHL using a hearing device (6). Low device uptake and current lack of guidelines most likely reflect the disparate nature of the degree of UHL, its aetiology, and the variety of treatment options available. The lack of guidelines poses a major problem for clinicians to advise the families of the children affected by UHL, since the impact of UHL can be highly variable across individual children as are their needs, desired outcomes, and personal preferences (7–9).

In 50% of the cases, the aetiology of UHL is unknown (10). There are several risk factors that have been associated with UHL (11) including premature birth, trauma, craniofacial anomalies, genetic causes, and bacterial or viral infections (12, 13). Children who have a family history of hearing loss are more likely to have bilateral hearing loss (BHL) whilst UHL is more likely to be present in those with craniofacial anomalies (8).

The listening difficulties that the children with UHL experience can mainly be explained by their loss of bilateral input. The range of loss of input can range from a child having no available hearing in their affected ear, often called SSD (weak or absent nerve for example), to very mild losses where they have some, albeit asymmetrical bilateral input. Their degree of hearing loss and aetiology will impact their device choice. The choice for a child to use a device and their device choice are also likely to be indicative of having an ear that works well and the ability of the child to use that monaural input to good effect. It can also reflect their listening environment, for example, if their device helps them in background noise or whether they are predominately in a quiet home and are able to position themselves for optimal listening. Counselling by a multidisciplinary team is advocated to avoid treatment bias for children with conductive UHL (14), and this can benefit children with either sensorineural UHL or aural atresia. However, it is critical to monitor speech and language development since they are at risk when listening with one ear and device trial should be implemented early (15). Furthermore, it is important to note that hearing is still limited with a BCD especially for children with SSD and/or in a noisy listening setting, possibly because the BCD does not restore the binaural hearing for children with conductive UHL (15, 16).

In a small study of children with Trisomy 13 and conductive UHL (mostly moderate hearing loss), the conservative approach of watchful waiting was often adopted, and when hearing aids were implemented, they were on the whole successful (14).

Where there is a lack of auditory input, this will impede binaural summation of loudness, the head shadow effect, and binaural release from masking which affects their ability to decode speech in noise and localise sound in space (15–19). In a recent study with children who have no access to sound in one ear (SSD) and who had a cochlear implant (CI), the implanted group exhibited an improved speech perception in noise and better sound localisation skills, compared with their non-implanted peers (20).

The morbidity of UHL can be similar to bilateral hearing loss (BHL), and there is a wealth of evidence that UHL can affect preverbal vocalisation, speech and language development, and cognition (8, 9, 21–25). The children with UHL can also struggle with listening fatigue particularly in noisy environments, and their degree of fatigue is very variable but can be very similar to the level of fatigue experience by children with BHL (26, 27).

There are a number of hearing devices that will not restore bilateral input but can be used to help the children with UHL—contralateral routing of signal (CROS) aids and bone-conduction devices help alleviate the head shadow effect as they re-route the sound via air or bone-conduction, respectively. BCDs and CROS-aids are commonly used for severe losses, fluctuating hearing loss, profound conductive losses or where the auditory nerve is absent or weak, whereas mild to moderate losses are often treated with conventional hearing aids.

The data from adult studies often imply that BCDs are better than CROS-aids but both devices impair speech perception when the noise emanates from the impaired ear side. Also, it has been shown that a BCD does not improve sound localisation abilities (30), but it is believed that they do improve speech perception when noise is on the side of the better ear (30–33).

A study assessing orienting head-movement responses in people with SSD investigated whether a BCD would jeopardise their directional hearing based on monaural spectral and/or level cues. They found that 5 out of 19 participants could localise certain sounds (broad-band and high-pass but not low-pass filtered noises) in the horizontal plane in the unaided condition and that a BCD did not deteriorate their localisation abilities (28).

The outcomes for the children with UHL can be as disparate as the aetiology and their type of hearing loss. For example, evidence of the impact of unilateral conductive hearing loss in children can be varied and sometimes mild, and there is limited evidence to indicate that hearing devices benefit every child in relation to their speech and language development. Aetiology is important since listeners with conductive UHL can activate the auditory pathways of their impaired ear through their own voice, and through bone and tissue conduction (29). If the UHL is conductive, BCDs can provide a form of binaural input where the cochlea is stimulated on the side of the loss as well as re-routing the sound to the better-hearing ear (when an inner ear hearing apparatus is functional). With the BCD, the cochleae can receive stimulation with negligible interaural attenuation so that a cross-hearing is experienced. The concomitant stimulation of the contralateral cochlea could impair the ability to process interaural differences in level and timing, which could limit improvements in the binaural hearing (30, 31). Prior studies indicate that BCDs provide speech recognition-related benefits; however, it is not agreed whether more accurate sound localisation occurs with BCDs (32, 33). With congenital conductive UHL, the data are contentious. There has been reported improvement in horizontal spatial hearing with a BCD for some listeners with conductive UHL despite the inherent problems of time delay and cross-hearing (32, 34), and one study shows that this can improve predominantly on the aided side with a BCD (35).

It was postulated that some listeners with congenital conductive UHL have adapted to their UHL and learned to rely on the spectral shape cues and monaural head shadow effect cues (36). In summary, how BCD stimulation affects spatial hearing abilities and the predictive factors that may affect the degree of the benefit provided by BCDs remains unknown.

Collation and comparison of data sets for paediatric UHL studies in the current literature can be difficult; a systematic review and consensus paper noted no firm evidence for the efficacy of current available devices nor evidence to inform decisions as to which devices are most suitable (5, 37). The limited data available suggest a trend towards improvement in speech perception with hearing devices, particularly with listening attention (35). Frequency modulation (FM) systems were shown to have the most benefit for speech recognition in noise, and studies evaluating CROS hearing aids demonstrated variable outcomes (38).

Several adult studies have demonstrated the long-term implications of uncorrected hearing loss; with an increased likelihood of experiencing social isolation and emotional distress as well as an increased risk of developing dementia in later life (39–41). Whilst the above implications are yet to be investigated in cases of early-onset UHL, other factors, for example, maternal education and earlier aiding, have been shown to ameliorate poorer outcomes (42). Watchful waiting should also be a possible option, particularly for milder hearing losses (14) as we have yet to show that treatment of UHL is associated with improved academic performance.

Future research may highlight the importance of early detection and appropriate treatment for some children with UHL.

To promote consistent and effective care for the children with UHL, it is important to both identify the current standard of care and support provided, as well as characterise the affected cohort of patients. It is also important to identify gaps in the funding and support for the children with UHL.

Moreover, an understanding of the respective thoughts of patients, parents, and clinicians on the care provided is essential to ensure a unified approach to UHL management.

In this current longitudinal cohort study, we describe all aspects, we can reliably ascertain from their data over 17 years in a large tertiary centre, on the management of children with confirmed permanent UHL. Consequently, there is a potential for further research into the future development of evidence-based management guidelines and promoting informed treatment decisions for clinicians, parents, and children.

The objectives of this study are the following:

1. Establish a database from the advent of NHSP (2002) to document the **demography and aetiology** of UHL in children in a tertiary referral centre.

2. Characterise **management of** UHL—e.g., type of device trialled, the age of the child when they first trial a device, when and how long they use their current device, and why they do not use their device.
3. Additional **support services** for the children with UHL and the documentation of any **concern** related to the impact of their hearing loss.

Methods

The routinely recorded and collected data in this study formed part of a service evaluation 2002–2019 at Nottingham University Hospitals. The data was also used from the Nottingham research database (NEAT) under ethical approval to analyse routinely collected data (REC project ID: 292263), South Central Berkshire Research Ethics Committee.

Inclusion and exclusion criteria

The criteria for inclusion included the patient referred on the NHSP and having a diagnosis of permanent UHL (hearing loss in one ear lasting for greater than 6 months). Children who did not speak English as a first language were also included.

The criteria for exclusion included children with acquired, fluctuating, or bilateral hearing losses as well as any child who did not have their new-born hearing screening (NHS) conducted in Nottingham.

Of the 89 cases identified as having UHL from the new-born hearing screening or referred from the UHL clinic, 26 (29.2%) cases were excluded as they were acquired or progressive, leaving a total of 63 cases of permanent congenital confirmed UHL. Of the 26 excluded, 13 cases were excluded for the reason that the hearing loss was progressive and developed to a bilateral hearing loss, 11 were acquired UHL, 1 case was both acquired and bilateral, and 1 case was excluded as the child did not have their new-born hearing screening in Nottingham.

Classification of UHL

The cases of UHL include permanent congenital, conductive, mixed, or sensorineural hearing loss, and the degree of hearing loss range from mild to profound. The aetiology and severity of hearing loss could be identified in many cases, but we could not reliably determine the number of cases with conductive UHL since the tympanometry data were viewed to be not of high enough quality. The guidelines of the British Society of Audiology (BSA) were used to classify the severity of UHL in the poorer hearing ear (British Society of Audiology, 2018). Hearing loss in each ear was recorded and averaged over four frequencies (0.5, 1, 2, and 4 kHz). Occasionally, only the hearing level at two frequencies was recorded (most often 1 and 4 kHz) usually due to the attention or ability of the child to remain engaged in the testing (depending on their age or degree of development). Hearing loss in the affected ear was classified as Mild: between 20 and 40 dBHL; Moderate: 41–70 dBHL; Severe: 71–95 dBHL; or Profound: over 95 dBHL.

Data extraction and analyses

The data were entered into an Excel spreadsheet from 89 patient paper notes (2002–2019) and electronic hospital databases (Practice Navigator, MedWay and NoTis) in a regional referral centre, Children's Audiology at Ropewalk House in Nottingham. The patient demographics included date of birth, sex, age at diagnosis of hearing loss, formal diagnosis of hearing loss, aetiology of UHL, and birth history. The results from the NHSP hearing test of the child and their diagnostic auditory brainstem response (ABR) as well as the two most recent hearing test results were recorded for frequencies 0.25, 0.5, 1, 2, 4, and 8 kHz, if present. When the data permitted, this was recorded for both ears. The age of the child at the hearing tests was recorded as was the method of testing—pure tone audiometry (PTA), visual reinforcement audiometry (VRA), and play audiometry. If the child had a hearing device, the device type, ear fitted, make and model, and date of first fit were recorded. For the patients who stopped using a device, the reasons for this were documented, and for those who had no device, the reasons why no trial was undertaken were noted. For device use, we catalogued when the device was worn and for how long, for example, none of the time, some of the time, at school, at home, or all the time, alongside any relevant quantification if present. The data-logging information from the hearing devices was not available. The support service data was also documented and included the teachers of the deaf (ToD), speech and language therapy (SLT), missed appointments, and comments made by the relevant parties relating to the management of the child and their respective outcomes.

We documented every concern reported from parents, clinicians, ToD, or the children themselves. Where data were of poor quality, verbatim quotes were lifted from patient notes.

Missing data

The cases of UHL may have been missed if the NHSP of the child took place out of the area, or if the child was not referred for diagnostic follow-up following a UHL found on the new-born hearing screening. The data were not available if the parents did not engage with audiology services after the initial appointment and diagnosis of UHL for their child.

The onset of hearing loss was categorised as congenital or acquired. Those with an acquired hearing loss were defined as developing hearing loss after having passed their new-born hearing screening. Congenital hearing loss was defined as children who were referred on the NHS as a neonate and then went on to have a confirmed permanent hearing loss by ABR and then later VRA or play audiometry (depending on the age or developmental ability of the child at the time of testing). The average of the hearing loss of the child (across frequencies,

TABLE 1 Aetiology of UHL: craniofacial abnormalities including atresia, microtia, maldevelopment of labyrinthine structures.

Aetiology	Frequency ($n = 32$)
Craniofacial abnormality	18
Malformation or absence of auditory nerve	7
Post-infection	4
Genetic	4
Birth complications	22
Caesarean section	8
NICU admission	7
Jaundice	3
Prematurity	3
Breech	2
Meningitis	1

NICU, neonatal intensive care unit.
Birth complications were present in 22 cases of the children with UHL. More than one birth complication may be documented for each child.

recorded in dB SPL) was recorded for both ears, and the data were used from the most recent hearing test (most often PTA). Where this information was not available, then the data from the second latest hearing test were used. In a number of cases, the most recent hearing tests only documented the data from the better-hearing ear (usually when the poorer ear was described as congenitally "dead"), in which cases the *current* severity of hearing loss remained unknown and these cases are documented as severe-profound*. These are listed separately in **Tables 2**, **3**.

The software used included Microsoft Excel (Internet) version 2302 to record the data. The Kruskal–Wallis (K–W) non-parametric ANOVA for multiple comparisons and Pearson correlation (r) were performed using SPSS version 28.0. The GraphPad Prism version 8.0 was used to graph the data.

Results

Of the total 89 cases of children with hearing loss identified between February 2002 and March 2019 analysed in this study, 77 children had permanent congenital hearing loss, 12 had an acquired hearing loss, and 63 children were identified with permanent congenital UHL.

1. Demography and aetiology: Of the 63 children with permanent congenital UHL, 50.7% (32/63) were male and 49.2% (31/63) female. Birth complications were recorded in 34.9% (22/63) of the cases where the most common were C-sections (36.3%; 8/22) and NICU involvement 31.8% (7/22). The aetiology of the hearing loss and the number of birth complications reported are shown in **Table 1**.

TABLE 3 The severity of hearing loss is for the poorer ear only and is categorised according to BSA guidelines. *Severe-Profound group: the poorer ears for children in this group have not currently been tested. The most recent hearing tests only documented data from the better hearing ear (usually when the poorer ear was described as congenitally 'dead'), in which cases the current severity of hearing loss remained unknown and these cases are documented as severe-profound* and listed separately in the table.

Frequency of use	Not at all/rarely, $n = 8$ (%)	All day, $n = 36$ (%)
Hearing loss		
Mild	0 (0)	0 (0)
Moderate	2 (25)	13 (36)
Severe	1 (12.5)	1 (3)
Profound	1 (12.5)	7 (19)
Severe-profound*	4 (50)	15 (42)
Device used		
BCD	3 (37.5)	16 (44)
HA	3 (37.5)	6 (8)
CROS	2 (25)	14 (39)

Degree of hearing loss is the hearing loss in their poor ear and is categorised according to the BSA guidelines (see Methods). "All day" includes when the device was reported to be worn all day in school.

The known aetiologies for UHL in the children were documented in 50.7% (32/63) of the cases. The aetiologies were broadly classed into categories and described in the notes as follows: craniofacial—structural malformation 56.3% (18/32) including atresia, microtia, and poor development of labyrinthine structures; malformation or absence of auditory nerve 21.9% (7/32); post-infection 12.5% (4/32); and genetic 9.38% (4/32).

The genetic testing revealed four pathogenic variants, and two had variants of unknown significance. The genetics data have previously been cited in a recent study (44).

With regard to the hearing severity of the poorer ear of the child, 1.6% (1/63) was mild, 31.7% (20/63) moderate, 15.9% (10/63) severe, and 15.9% (10/63) profound. In the remaining 34.9% (22/63), the degree of severity of their hearing loss was not documented in their most recent two hearing tests. In these 22 cases, the poorer ear remained untested for a prolonged period, sometimes since the diagnostic ABR as in the majority of the cases, it was recognised that the ear was

TABLE 2 A hearing loss in relation to device trial.

	Mild	Moderate	Severe	Profound	Severe-profound*
(A) Trial/severity					
Device trial, $n = 45$	0	16	2	8	19
No trial, $n = 18$	1	4	8	2	3
(B) Hearing loss in relation to device type, $n = 45$					
HA, $n = 9$	0	9	0	0	0
BCD, $n = 20$	0	5	1	3	11
CROS, $n = 16$	0	2	1	5	8

The severity of hearing loss is for the poorer ear only and is categorised according to the BSA guidelines.
*Severe-profound group: the poorer ears for children in this group have not currently been tested (see Methods).

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effectively "dead" due to a number of reasons. We do not have the current degree of hearing loss recorded for this group, so we have labelled this group separately in Tables 2, 3 as severe-profound*.

Most children with UHL who trialled a device had either a moderate hearing loss 36% (16/45) or were in the severe-profound category of hearing loss (19/45; 42%). Most of those not trialling a device had severe hearing losses 44.4% (8/18) (Table 2A). There were five children who would have had an aidable mild ($n = 1$) or moderate ($n = 4$) loss in their poorer ear but who never trialled a device. Only the children who owned and reported that they wore a hearing aid (HA) had a moderate degree of hearing loss ($n = 9$); however, there were children with moderate losses who also used a BCD ($n = 5$) or a CROS-aid ($n = 2$) (Table 2B). Of the two children with severe losses, one used a CROS-aid and the other used a BCD. Three children with profound hearing losses used a BCD, whereas five children used a CROS-aid. More than half of the children who had severe-profound losses* were currently using a BCD ($n = 11$), and eight children used a CROS-aid (Table 2B).

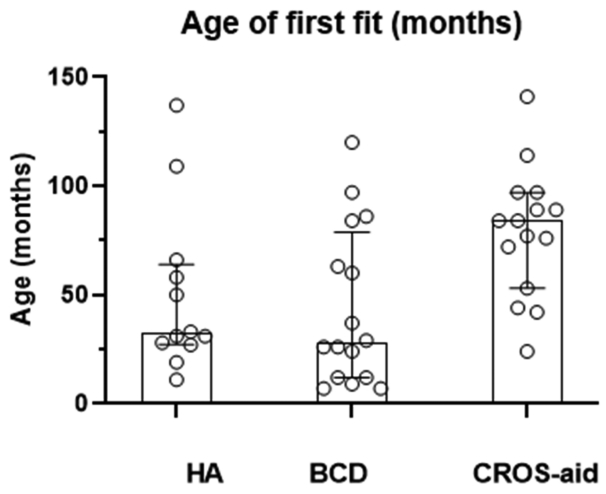

FIGURE 2
When a child is first fitted with their hearing device. The age of first fitting of the device (in months) for each child is represented by an open symbol for children who have a HA, hearing aid; a BCD, bone-conduction device; or a CROS-aid, contralateral routing of signal aid.

Management of UHL

Forty-five children (45/63; 71%) with UHL trialled a device with the remaining 18 (18/63; 28.5%) children having had no device trial (Figure 1). Twenty (20/45; 44%) children trialled a BCD, 16 (16/45, 36%) a CROS-aid, and 9 (9/45, 20%) conventional hearing aids.

The range of ages at first device fit was wide with a mean age of 56 months or 4.7 years ($n = 41$). The ages of the child when they are first fit with their first device are plotted individually in Figure 2, and the overlain boxplots illustrate the median and interquartile ranges (IQR) for age at first fit by device type. A significant difference between the groups was found (K–W ANOVA, $K = 8.4$; $P = 0.015$). The medians and minimum–maximum ranges (in months) for age of first fit within device types were the following: HA (median, 32 months; 11–137 months), BCD (median, 28 months; 7–120 months), and CROS-aid (median, 84 months; 24–141 months).

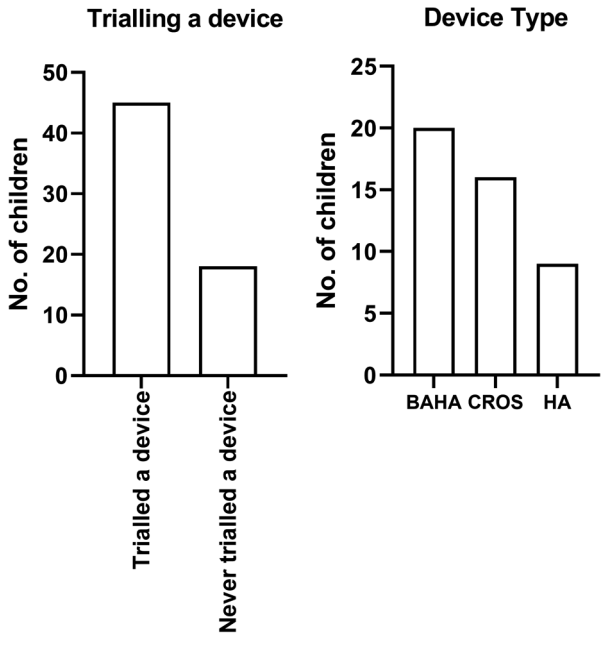

FIGURE 1
The number of children who trial a device and the device type trialled.

When a device is worn and who leads the device choice

A device was reported to be predominately worn "at school" or "all day" (Figure 3A). A parent-led decision for the device trial was documented in half, 51% (23/45) of all the cases, followed by child-led decision 13.3% (6/45) and clinician-led decision 8.9% (4/45). In six cases (13.3%, 6/45), "other" represents cases where the overall lead for the decision in these cases was unclear and likely to be a joint decision between the clinician, parent, and child (Figure 3B). In six cases (13.3%, 6/45), no reason was documented for device trial.

In the 36 children with UHL who wore their device all day, 13 had moderate hearing loss, 1 had severe, 7 had profound, and 15 had an unknown degree of hearing loss, likely to be severe-profound* (Table 3). BCDs were most used by children who wore their device all day, including at school (16/36, 44%), followed by CROS-aids (14/36, 39%). Six (6/36, 8%) children wore their hearing aids all day, including at school.

The children kept their BCDs for longer periods of time (median 32.5 months), but this was not statistically significantly longer than for HAs (median 24 months) or length of time they kept their CROS-aids (median 22 months) ($n = 44$; Kruskal–Wallis ANOVA

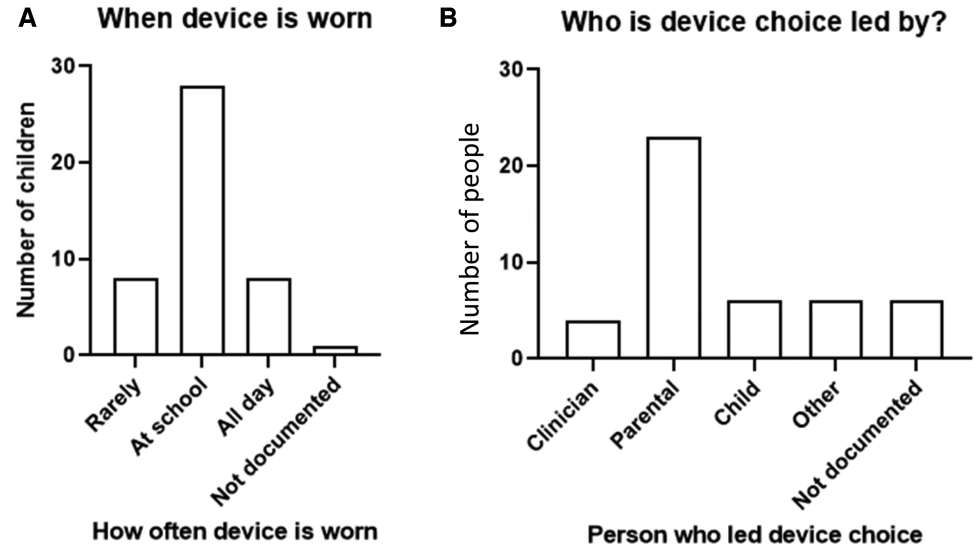

FIGURE 3
(A) When a child with UHL wears their device; (B) device trial lead, $n = 63$.

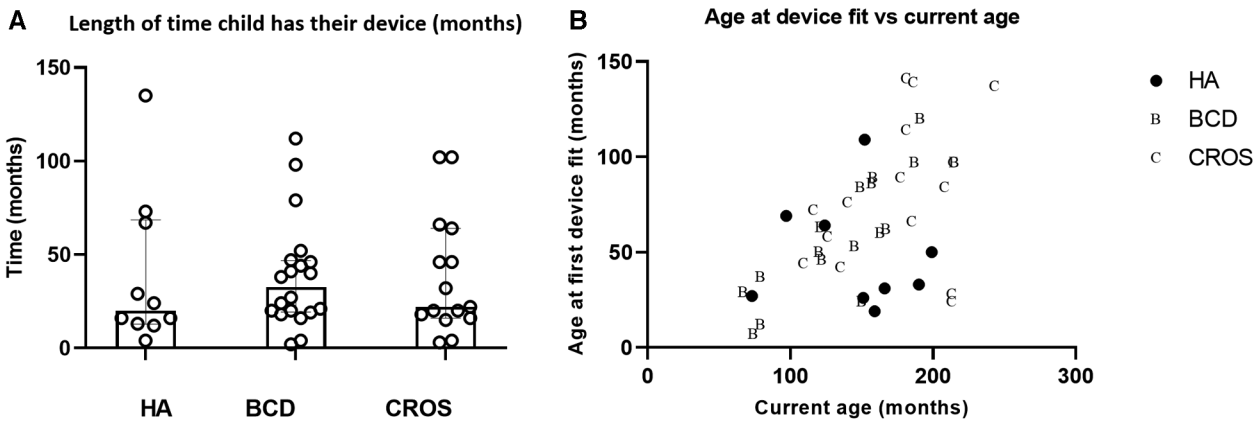

FIGURE 4
(A) Length of time was approximated to the child's last hearing test minus the date of their first trial of their device. Medians and interquartile ranges plotted in (B). In (B) age when the child was first fitted with their device (in months) is plotted against their current age in months. Filled symbols represent children with a hearing aid, letter B for children with a BCD and C for children fitted with a CROS aid. HA, hearing aid; BCD, bone-conduction device; CROS-aid, contralateral routing of signal aid.

$P = 0.91$). Across device types, the length of time a device is used is widely varying (Figure 4A); however, these data give no indication of whether the device was worn or not. Nevertheless, the child would return to Paediatric Audiology to have their hearing device serviced and updated, and if it was stated that the device was not being worn, it would be returned to the service. Unsurprisingly, there is a positive correlation between the age of first device fit and the current age of the child ($r = 0.52$; $P = 0.0004$) (Figure 4B) —useful to note as it shows that there is a trend where children who are born more recently are more likely to be fitted at a younger age. Also, younger children were more likely to be fitted with a BCD or a HA than a CROS-aid.

Children who have a device but rarely wear their device

There were eight children who have had device trials and currently wear their device but only infrequently. For the three children, it was documented that infrequent wearing was because they experienced bullying wearing their hearing devices (and/or appearance of their ear), two children experienced feedback from their device, two found them uncomfortable to wear (one also found the sound robotic and stopped wearing because of appearance), and one found their HA not helpful with their hearing.

Not trialling a device

In terms of their aetiology, the majority who have not trialled a device in this group have external or inner ear abnormalities (n = 5), two have auditory neuropathy spectrum disorder (ANSD), three have sensorineural hearing loss, one had CMV, and one had a skull fracture as an infant. In the six cases, the aetiology was unknown or not documented.

Of the 18 children who did not trial a device, it was discussed between the child and the parent in 12 cases. In 50% (9/18) of the cases, the parent decided for the child not to trial a device, and it was an ENT clinician-led decision (mild UHL) in one case (5.6%, 1/18). A further two cases were joint clinician and parent-led decisions, and six cases had no documented reason. However, despite there being no documented reason for "no trial of a device," it was reported in three cases of the child doing well, having a good understanding of speech and no developmental concerns. One child was lost to follow-up.

Follow-up during audiological care

Support services

The percentage of children who had a documented referral to a support service was 81.6% (51/63); the remainder had no documented support referral.

Of the 51 children who were referred, 72.5% (37/51) received their intended support with no issue, whilst the remaining 27.5% (14/51) encountered an issue leading to an unsuccessful provision of support. The documented obstructions to this support were 86% (12/14) due to the child not meeting referral criteria and 14% (2/14) due to parental refusal of support. Overall, 93 support referrals were made: 74% (69/93) for a teacher of the deaf and 20.4% (19/93) for speech and language therapy. A further 5.38% (5/93) to "other" services. Within these 51 children with referrals, most, 37/51 (72.5%), had combined referrals to several support services and 14/51 (27.5%) had a single referral to a specific service.

Documented concerns surrounding UHL

A total of 65% (41/63) of the children with UHL had no reported concerns, and 28.5% (18/63) of the children had a documented concern: 5 children reported difficulties with their hearing devices, 5 had problems with their speech, 4 had no improvement in their hearing, 3 faced self-image or bullying issues, and 1 case of a child struggling to interact socially with friends. In three cases, the reason for the concern was not documented.

Where a concern was raised during audiological care, the hearing level severity included 0% (0/18) mild, 17% (3/18) moderate, 17% (3/18) severe, 17% (3/18) profound, and 50% (9/18) of the cases had an unknown degree of hearing loss—likely to be severe-profound*. Furthermore, in the 18 children where a concern was reported most trialled a device, 8 children had trialled a BCD, 4 an HA, 3 a CROS-aid, and 3 children had no device trial on record. Within these 3 cases with no documented device trial, 2 cases received documented support service input. Only in one case did a child with raised concerns has no device trial and no support.

Half, 56% (10/18), of the concerns were raised by schools and teachers, 22% (4/18) of the concerns were raised by the family, and a further 22% (4/18) of the concerns were raised by the children themselves.

Discussion

A number of organisations have established candidacy guidelines for paediatric amplification, but the guidelines for the children with UHL remain more ambiguous, and recommendations vary (4). Currently, there are no national clinical practice hearing guidelines surrounding the treatment and management of the children with UHL in the United Kingdom. Most guidelines specify criteria for amplification as audiometric threshold levels and considering the disparate nature of the condition, the treatments, and its aetiology, and it has been postulated that individually treating each child is optimal (45).

Most children with permanent congenital UHL in our cohort wore a device, most often a BCD or a CROS-aid; they wore it at school or all day indicating that, at least for school age children, a trial of a device should be prioritised. In our cohort, it was the parents who usually made the decision for their child to trial a device. Many children with UHL are at high risk of certain adverse developmental outcomes (9, 23), thus funding should be made available for all the children with UHL who need a referral to the support services that they require and for their preferred device.

The prevalence of hearing loss in one ear is estimated at 0.3–1 per 1,000 births (46); our estimation for this cohort is 0.4 per 1,000 births. The prevalence may be higher as UHL cases (particularly mild UHL) go undetected or under-reported; NHSP guidance in the United Kingdom does not aim to identify milder hearing losses, but as in the United States and Canada, milder losses may be identified as a by-product of the screening procedures. Furthermore, the parents who did not engage with the audiological service following identification of UHL on the NHSP are not included in this study.

To improve the current standard of care, it is first important to identify the cohort of patients affected and to understand the treatment provided. This study provides much needed information about current practice and the reasons behind the interventions and choices made by the care team of the children and their families in this central tertiary regional referral centre over a 17-year period.

Demography and aetiology

Similar to previous studies, we found that the aetiology of UHL was documented in half of the cases and over half of these were structure abnormalities related to the ear, 22% had absent or malformed auditory nerve, 12% post-infection, and 9% genetic causes. The aetiology and degree of hearing loss are very

important to ascertain as they can impact device choice for the child with UHL. It is established in the literature that congenital anomalies, for example, craniofacial abnormalities and ear malformations, are common in this population (12, 47), and the prevalence of malformations in the inner ear and/or internal auditory canal are higher in infants with congenital UHL than in infants with bilateral hearing loss (48), with two-thirds of the children with UHL having inner ear and/or internal auditory canal malformations (49). The structural abnormalities of the inner ear structures can mean that the conductive losses are sufficient to warrant the fitting of CROS-aids or BCDs. Aetiology is an important consideration since more than half of the children in this study who had never trialled a device had ANSD or abnormalities of the external or inner ear. Furthermore, where a concern was raised during audiological care, 50% of the cases had an unknown degree of hearing loss—likely to be severe-profound*—and these are the cases where the hearing was non-functioning from birth.

Previous studies have revealed that the percentage of UHL associated with a family history is approximately 3.7%–13% (48, 50, 51), which is similar to the number of UHL cases with genetic variants in this study (9%). Currently, genetic screening is not recommended or funded for all infants diagnosed with UHL in the United Kingdom, only for those where a syndrome is suspected. There is little to no research on the specific genetic variants associated with UHL. Three of the four patients with a genetic variant also had a diagnosed syndrome, suggesting that the syndromes may be linked to specific genetic variants (52). UHL and a NICU admission can be a red flag for additional congenital anomalies and developmental delay (44).

Characterisation of the management of treatment for UHL

In this study, most (71%) children with UHL trialled a device, which in some cases is higher than pre-existing literature. In Purcell's study of 50 children, aged 5–19 years, a similar number to our study ($n = 34$ children; 68%) had trialled a hearing device. Fewer, however, ($n = 20$, 40%) continued to use their device. In a study of 31 children with congenital, acquired, or unknown onset UHL (age range: 1–10 years), it was reported that 81% of the children with moderately severe or better UHL accepted the use of a hearing aid. However, when the UHL was severe or profound, the parents reported very poor or no use of the hearing aid (53). One reason could be that the data we report are more recent compared with the previous studies. Also, we include all types of devices and a wider age range (0–18 years), and we did not include the children whose parents did not engage with audiological services after the initial diagnosis of UHL.

Appachi et al. evaluated auditory outcomes from various modalities of hearing rehabilitation, including FM systems, hearing aids, CROS-aids, and BCDs, where the use of FM system was beneficial for speech recognition in noise, and hearing aid use showed a trend of improvement in speech perception. CROS-aid use was associated with mixed auditory outcomes. BCD use was associated with consistent gain in speech reception thresholds (SRTs) and speech discrimination, and an improved hearing in noise, but findings for sound localisation were inconsistent. Similarly, Liu et al. systematically reviewed the role of BCDs and found consistent gain in SRT and speech discrimination, but inconsistent results with sound localisation. The measurement of the quality of life showed a high rate of usage and benefit in the learning domain.

The age of the first device trial was disparate and depended on the device type. It also depended on the age of the child where the children who have been seen more recently (younger) were fitted earlier, most likely due to change in management over time. The average age of fitting of a device was around school age (4.7 years); BCD and HA can be fitted earlier and were fitted around preschool age. This age of fit was later in comparison with other studies for children with BHL, for example, Walker et al. reported an average age of fitting at 10.99 months (range: 5 months to 7 years, 3 months) within a cohort of 211 children identified with BHL (54). However, it should be noted that the ages of their participants were skewed to the younger age group than that of the current study. There is a disagreement in the literature concerning the best age to fit a child who has UHL, and a case-by-case approach is optimal (1, 46–48).

One interpretation of the wide range of the ages of first fit in this study indicates that the children with UHL are followed up, and when they have trouble, they have no problem returning to audiology services and requesting a trial even into their teenage years.

Current NICE guidelines indicate those babies who are confirmed deaf by the NHSP should receive a hearing aid within 2 months. Meanwhile, the British Academy of Audiology provide no fixed time over which amplifications should be provided, stating "amplification be provided in accordance with family centred care guidelines" (45). Whilst there is little evidence to suggest early aiding is beneficial to *all* the children with UHL, there is some evidence to suggest that wearing a hearing device can improve the quality of life, especially in those suffering with speech and language or academic and behavioural issues. A few studies have shown that early intervention may be crucial particularly for speech reception thresholds and speech discrimination, especially in noisy environments (33, 55, 56); however, the data measuring improvement in sound localisation with aids are inconsistent. A study that examined the quality of life measures reported a high usage rate of BCDs among children (33) whereas another study found low usage (1.3 h/day) but with an improvement in CHILD scores and speech in noise testing following amplification with a BCD (52).

Other studies have found that hearing devices may not be beneficial for younger children with UHL and do not improve speech recognition (33). Such contrasting evidence is also mirrored in our anecdotal data, with one parent stating that their child is "happier and much more responsive" with hearing aid use (case 59) and another reporting they are "unsure of the hearing aid and are unable to notice a difference" (case 89). As such, more research is needed for the best age to trial a device for the individual child with UHL.

The uncertainty about age at which to fit a hearing device is complex and is dependent not only on the importance of stimulating the binaural neural pathway during development but also the degree and aetiology of hearing loss. For some children, bilateral stimulation can only be achieved with a CI (not yet commissioned in the United Kingdom for UHL/SSD, and no CI use is reported in this study). CROS-aids have a very different role, and evidence for the benefit of the early fitting of CROS-aids for children is not available (in the United Kingdom, they are not fit before 6–7 years of age). CROS-aids do not stimulate the unaided ear and only provide access to the acoustically blind area, so there is no binaural access to sound. It is thought that CIs for severe-profound losses and hearing aids for mild–moderate losses are vital to stimulate the auditory pathway during the crucial periods of neural development for the acquisition of speech and language (4, 56, 57).

With regard to the devices used by the children in this study, nearly half wore BCDs, a third wore CROS-aids, and a fifth wore conventional hearing aids often reflecting the age of the child, aetiology, and type of hearing loss experienced. No children in this cohort had a cochlear implant despite their associated benefits seen in children and adults with severe-profound UHL (60, 61). This is likely due to current lack of funding for cochlear implantation for UHL in the United Kingdom and may also reflect limited evidence. For the children who trialled devices, very few did not wear their device, and of these cases, the degree of severity was spread across moderate, severe, and profound groups with no clear majority, similar to a prior study (24). Most children with moderate losses used a hearing aid. There were no children with mild, severe, or profound losses currently using a hearing aid. Unsurprisingly, all children with severe, profound, or severe-profound* losses currently used a BCD or a CROS-aid. The children who have profound losses were more likely to have a CROS-aid (63% CROS; 37% BCD), and the children with severe-profound* losses were more likely to have a BCD (58% BCD; 42% CROS). We do not know why most children who did not trial a device in our cohort had a severe level of hearing loss. One possibility is that mild and moderate losses are easily treated with hearing aids and profound losses with BCD or CROS-aid whereas severe losses fall between the two and thus there could be indecision about the best device to trial.

Most children who wore a device reported that they wore it all day or at school. A parent-led decision for device trial was most often documented (51%), followed by child-led decision (13%) and clinician-led decision (9%). For the children who did not trial a device, half were documented as a parent-led decision, very few were clinician- or child-led decisions. With regard to "decision to trial a device," specific decision making and counselling were often poorly documented and unclear. It is likely that the decision to trial a device is complex and child-specific; further investigation would be useful into how we could better understand both parental and child concerns. This may be particularly important in cases where device was not trialled or parents were not engaged with audiological services. Funding is also a point of concern as with limited resources, a CROS-aid or hearing aid is much more economical than a BCD. There are no official recommendations in the United Kingdom for CROS-aids, but there is a practical consideration when fitting them to young children. CROS-aids are best fitted on open fit slim tubes so that the sound to the better ear is not attenuated. Young children do not have the appropriate size of ear canal to enable fitting of open slim tubes, and hearing aid manufacturers do not make domes/slim tubes small enough for the ear canals of children. Furthermore, due to the small removable components (e.g., domes), there are choking hazards for small children. If CROS-aids were fit to a small child, an occluding ear mould would be required to aid retention (rather than the ideal open fit slim tube), and the occlusion effect would need to be overcome in the hearing aid programming. For these reasons, CROS-aids are not fit for children under 6 years old in Nottingham, United Kingdom. The children with severe and profound losses are not usually fitted with conventional hearing aids because of interaural attenuation due to the high levels of gain required, resulting in a cross-hearing into the better ear and therefore likely distortion of the sound perceived in the normal hearing ear.

Our data illustrate the importance of BCD and CROS-aids and are in contrast to an early study where 27 children with UHL who were fitted with an HA but usually did not wear it, 26% reported wearing it all of the time, 4% reported wearing it only in school, and 50% reported never wearing it (62).

The data that predate commissioning of the BCD use for UHL found that hearing aid use was high ($n = 31$; 81%) *but* only for users with moderately severe or better levels of hearing loss, whereas children with severe or profound UHL had poor or no use of hearing aids (53).

This is likely because those for whom a BCD or CROS-aid would be particularly beneficial did not have access to those devices at that time.

Whilst it can be beneficial for children to be wearing their devices throughout the day, self-reported use of hearing aid frequency can be inaccurate. It has been shown that although the estimates and data-logging of the parents were significantly correlated, the results indicated that the parents overestimate the amount of time their children wear their hearing aids by about 2.5 h (47). In a recent study of babies and carers, no correlation was found between hours of daily hearing aid use and self-reported hearing aid management skills or factors having a negative impact on hearing aid use (63).

It is possible in this current study that both child and parental reports of device use could be misleading. However, the children with UHL in this study were overall older than the children reported in prior studies, and it has been shown that longer hearing aid use relates to older age, poorer hearing, and higher maternal education (42, 54). Future recording of data-logging and environmental assessments from technology and hearing devices would enable greater insight into the device use for the children with UHL.

Where it was reported that the children only rarely wore their devices, in this current study, the predominant reasons were because of bullying at school, feedback from the device, and discomfort. A prior study of 15 children with UHL found that the most common reason for the cessation of device use was

discomfort, followed by lack of benefit (64). The children with UHL may also be particularly vulnerable if they have had surgery for craniofacial anomalies. In a recent study, hearing impairment among adolescents was associated with increased reported rates of bullying victimisation: 34% of children with hearing loss were bullied, and children who do not use hearing aids had even greater odds of being bullied (OR = 2.40, 95% confidence interval: 1.18–4.86, $P = 0.015$) (65). It is difficult to know if the non-hearing aid users were not using their devices because of the bullying or if their speech and other developmental problems were the cause of them being targeted. Further research is needed to investigate how anti-bully interventions can support this vulnerable group. A 2019 meta-analysis and systematic review found that school-based anti-bullying programmes significantly reduced bullying perpetration and victimisation; this could be vital for all children with hearing loss (66).

Follow-up during audiological care, support services

More than half of all children in our cohort had a referral to a support service; this is in keeping with most of the current literature. A previous study indicated that 39% of the children with permanent UHL received speech therapy, 54% had received an individualised education programme (IEP), and 36% had received additional educational assistance ($n = 46$, ages 6–12 years) (67). In a later study, the need for further academic support was identified in this population—more children with UHL received IEPs (45%) and speech therapy (41%) than the children with normal hearing (5% for both IEPs and speech therapy) (68). Three quarters of these children received their intended support with no issue, whilst the remaining quarter of the children with UHL ($n = 14$) encountered an issue leading to an unsuccessful provision of support. The majority (86%) of documented obstructions to this support were due to the child not meeting referral criteria, defined by local support teams, only two cases were due to parental refusal of support. The criteria for referral are defined by the support service rather than audiologists—this is an area of inconsistency that needs further evaluation.

Documented concerns surrounding the UHL of the child

A fifth of the children with UHL went on to have documented concerns; these included speech and language problems, self-image or bullying issues, hearing aid difficulties, and/or poor social interactions with friends. The majority (40/51) of the children with UHL that had a referral to a support service had trialled a hearing device. Whilst the struggles faced by each child differ, thematic analysis (69), using a focus group comprising of a mix of children with permanent hearing losses, their parents, and audiologists, suggests that there are six main domains in which hearing loss can affect children. These include behaviour, feelings, environments, social/activities, family, and hearing equipment. The anecdotal data from our own study are in line with these findings highlighting the universal impact of hearing loss on children as well as the consequent importance of ensuring their adequate support. Within adults with UHL, core rehabilitation outcomes include the following: (1) spatial orientation, (2) group conversations in noisy social situations, and (3) impact on social situations (70). Consequently, it would be beneficial to identify the core outcomes for the children with UHL; in doing so, management plans can be better tailored and the outcomes of the child more easily monitored and assessed.

Guidelines and funding for the children with UHL

Given the lack of UK National Health Service and NICE guidelines for the management of paediatric UHL, audiologists often use SLT or a diagnosis of developmental impairment to guide them on their treatment plan, and to suggest to the parents that their child should trial a device. Unfortunately, by the time the child requires SLT, this may be too late, especially when early device trials may impact speech and language acquisition, as has been noted by the LOCHI study (42). Most device trials in our cohort were parent-led, and without counselling, it may be the case that the parents are more willing to trial a device if they see the adverse developmental effects of UHL. Deciphering speech in noise is particularly tricky for the children with UHL, and therefore in a noisy home or at preschool, it would be strategic for the children with UHL to use a hearing device. Also, certainly in school/preschool, a sound-field system would benefit all children regardless of device use. Currently, there is consistent funding neither in the National Health Service nor in education authorities for FM/remote microphone systems, sound-field systems, and support, including SLT for the children with UHL.

More than half of the concerns about a child with UHL were raised by teachers, and a fifth were raised by the family, and a further fifth were raised by the children themselves. This is likely due to teachers spending most of the day with the child in an environment wherein auditory cues are paramount, and they may also be highlighting the developmental delay of the child and paucity of support in school. The parents/families/clinicians may also seek their advice from specialist interest groups, social media, and charitable web sources such as the National Deaf Children's Society.

The recognition of hearing impairments within schools and the consequent supporting facilities they can provide is also likely to affect the response of the child. Prior to the NHSP, the school hearing screening programme (SHSP) was used throughout the United Kingdom to recognise the children with undiagnosed hearing impairments (42). School age screening continues in many, but not all, parts of the country. Within the United Kingdom, Fortnum et al. have found the diagnostic accuracy of school hearing tests not to be cost effective (71) and that the distinct lack of quality data numbers is one of the reasons that

funding for the SHSP has since ceased. Unfortunately, the late identification of hearing loss is likely to be of detriment to the children with acquired forms of hearing loss (72–74). However, screening and early diagnosis is only the start. The availability of high-quality early years support for hearing loss remains a major barrier to the progress of the children. Failure to provide this high-quality support means that the potential benefits of new-born hearing screening are not being realised consistently across the United Kingdom. Almost a third of families did not feel they got the support they needed to make sure their child made good progress and developed well after diagnosis through new-born hearing screening (75). This is perhaps why, in comparison, the siblings of the children with UHL have been shown to perform better in a number of domains (behaviourally, socially, and academically). One-fifth of the children with UHL were diagnosed with developmental delay (76).

Strength and limitations

This study has a number of strengths and limitations.

It is important to note that for the children and families in this study, although there are no national guidelines for treatment of UHL, there are some guidelines about device candidacy (see the **Supplementary Material**). Thus, device choice for a child may not only depend on parental/child choice. In the United Kingdom, the National Health Service provides free hearing devices for all children at the point of service. Device type is not impacted by a plan under the health insurer of the family, and audiologists working for the National Health Service endeavour to provide an equal service for all children with hearing loss. There are no care guidelines for individuals with UHL; however, under current guidelines, the recommended care pathway for individuals with SSD in the National Health Service involves initially trialling a conventional hearing aid, followed by a CROS-aid, and then a BCD (see **Supplementary Material** for details). Bone-conduction implantations are funded by the National Health Service, but CIs for UHL/SSD are not. The current guidance for eligibility of individuals for consideration of a CI within the NHS includes a requirement for bilateral severe-profound hearing loss; children and adults with UHL/SSD in the United Kingdom following National Health Service care pathways are currently ineligible for this intervention.

A further limitation to this study is potential sampling bias since many children with mild losses can be missed; they may either not be picked up on UNHS or the parent may not follow up with audiological services following diagnosis.

Another limitation of the study is that we are not able to make inferences about device benefits since one cannot extrapolate from the reporting of wearing a particular device to the device being beneficial for the child in all environments.

A strength of this study is that there are advantages to following the same children over time in a single large centre, and the continuity of care for these children are reflected in consistently documented follow-up notes over a prolonged period. There was a consistency to record keeping, which contrasts to the data that are amalgamated from multiple sites and have different local policies and different record keeping regimens.

Future work

We do not know why some children with UHL go on to require speech and language services and struggle to develop academically and behaviourally whilst others do not. The fitting of the hearing device, degree of hearing loss, and maternal education are key (42), but research is required investigating the contributing genetic and environmental factors.

Future research into how the brains processing of monaural cues are impacted by late identification of hearing loss or later aiding is needed, as these could impact the outcomes of the children (77). This is particularly important since some studies have shown that asymmetric hearing loss causes a reweighting of cues that are used and postulate that adapted monaural cues may be utilised for sound localisation (78, 79). A recent study in children with congenital conductive UHL showed that they may rely on monaural spectral cues for horizontal sound localisation (35). Context of listening cues can also be important for sound localisation and would be interesting to investigate (80).

Long-term follow-up into adulthood would be beneficial for this cohort to examine which and when are the best devices to trial for a child and the most favourable support services. It will also be important to understand the reasons the families do not engage with hearing services and to quantify the outcomes of their children. Setting up anti-bullying campaigns within schools could be vital for children with hearing loss as they are particularly vulnerable, and their device use is likely most useful in school. A core-outcome set of what is important for the children with UHL and their families is important to define.

It will be important to determine why particular children with UHL struggle in school, and whether instructional training for the parents and teachers improves the likelihood of a positive outcome for the child. This may be particularly important for the parents of the children with UHL who in a recent study underestimated the fatigue of their child (27).

Future research should concern the items that enable the children with UHL to succeed and discover biomarkers that can accurately quantify stress and the quality of life. These factors are likely to be complex, multi-faceted, and relate to their frustration, attention, anxiety, fatigue, peer relations, social confidence, independence in the classroom, and emotional maturity, which are the important variables in educational success for children. Researching these aspects of a child's education could be key to understanding their struggles and thus providing specific support they need to help them succeed.

Conclusion

In our study, most children with UHL wore a BCD or a CROS-aid and reported they wore it for the duration of their school day. There was a very wide-ranging age of first device fit, but on average,

it happened at school age (4.7 years). The children who trialled a BCD or hearing aid were fitted earlier at around 2.5 years. Additional support with speech and language via support services were available for three quarters of this cohort, but for those who were unable to access this support, it was primarily because the child did not meet the referral criteria. Several areas of provision of support services provided for the children with UHL are currently under resourced. Individualised treatment plans are essential for this distinct cohort but where devices are not trialled or worn then sound-field amplification systems in the nursery and school would improve all the outcomes of the children regardless of their hearing status.

The funding for genetic testing and consistent provision of support services, counselling, and anti-bullying campaigns within schools for this understudied group is vital.

Data availability statement

The raw data supporting the conclusions of this article will be made available by the authors, without undue reservation.

Ethics statement

Ethical review and approval was not required for the study on human participants in accordance with the local legislation and institutional requirements. Written informed consent from the participants' legal guardian/next of kin was not required to participate in this study in accordance with the national legislation and the institutional requirements.

Author contributions

SKT and KRW: conceptualization, methodology. RP, SKT, ST, and KRW: investigation. ST, RP: validation and formal analysis. RP, PKB, SKT, and DJH: data curation, writing – original draft preparation. RP, SKT, KRW, and DJH: writing – reviewing and editing. All authors contributed to the article and approved the submitted version.

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Conflict of interest

The authors declare that the research was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

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Supplementary material

The Supplementary Material for this article can be found online at: https://www.frontiersin.org/articles/10.3389/fped.2023.1197713/full#supplementary-material

References

1. Doshi J, Banga R, Child A, Lawrence R, Reid A, Proops D, et al. Quality-of-life outcomes after bone-anchored hearing device surgery in children with single-sided sensorineural deafness. *Otol Neurotol.* (2013) 34:100–3. doi: 10.1097/MAO.0B013E318277A3DD

2. Briggs L, Davidson L, Lieu JEC. Outcomes of conventional amplification for pediatric unilateral hearing loss. *Ann Otol Rhinol Laryngol.* (2011) 120:448–54. doi: 10.1177/000348941112000705

3. Krishnan LA, Van Hyfte S. Management of unilateral hearing loss. *Int J Pediatr Otorhinolaryngol.* (2016) 88:63–73. doi: 10.1016/J.IJPORL.2016.06.048

4. Fitzpatrick EM, Cologrosso E, Sikora L. Candidacy for amplification in children with hearing loss: a review of guidelines and recommendations. *Am J Audiol.* (2019) 28:1025–45. doi: 10.1044/2019_AJA-19-0061

5. Bagatto M, DesGeorges J, King A, Kitterick P, Laurnagaray D, Lewis D, et al. Consensus practice parameter: audiological assessment and management of unilateral hearing loss in children. *Int J Audiol.* (2019) 58:805–15. doi: 10.1080/14992027.2019.1654620

6. Purcell PL, Edwards TC, Wisneski M, Chan DK, Ou H, Horn DL, et al. Unilateral hearing loss in youth: development of candidate items for a condition-specific validated instrument. *Otolaryngol Head Neck Surg.* (2018) 159:1043–50. doi: 10.1177/0194599818797092

7. Dwyer NY, Firszt JB, Reeder RM. Effects of unilateral input and mode of hearing in the better ear: self-reported performance using the speech, spatial and qualities of hearing scale. *Ear Hear.* (2014) 35:126–36. doi: 10.1097/AUD.0b013e3182a3648b

8. Rohlfs AK, Friedhoff J, Bohnert A, Breitfuss A, Hess M, Müller F, et al. Unilateral hearing loss in children: a retrospective study and a review of the current literature. *Eur J Pediatr*. (2017) 176:475–86. doi: 10.1007/s00431-016-2827-2

9. Fitzpatrick EM, Gaboury I, Durieux-Smith A, Coyle D, Whittingham JA, Nassrallah F. Auditory and language outcomes in children with unilateral hearing loss. *Hear Res*. (2019) 372:42–51. doi: 10.1016/j.heares.2018.03.015

10. Fitzpatrick EM, Durieux-Smith A, Gaboury I, Coyle D, Whittingham J. Communication development in early-identified children with mild bilateral and unilateral hearing loss. *Am J Audiol*. (2015) 24:349–53. doi: 10.1044/2015_AJA-15-0003

11. Tharpe AM, Sladen DP. Causation of permanent unilateral and mild bilateral hearing loss in children. *Trends Amplif*. (2008) 12:17–25. doi: 10.1177/1084713807313085

12. Yelverton JC, Dominguez LM, Chapman DA, Wang S, Pandya A, Dodson KM. Risk factors associated with unilateral hearing loss. *JAMA Otolaryngol Head Neck Surg*. (2013) 139:59–63. doi: 10.1001/jamaoto.2013.1097

13. Howell JB, Appelbaum EN, Armstrong MF, Chapman D, Dodson KM. An analysis of risk factors in unilateral versus bilateral hearing loss. *Ear Nose Throat J*. (2019) 98:330–3. doi: 10.1177/0145561319840578

14. Stewart R, Gallagher D, Leyden P. Diagnosis and management of conductive hearing loss in children with trisomy 21. *J Paediatr Child Health*. (2018) 54:1242–5. doi: 10.1111/JPC.14065

15. Corbin NE, Buss E, Leibold LJ. Spatial release from masking in children: effects of simulated unilateral hearing loss. *Ear Hear*. (2017) 38:223–35. doi: 10.1097/AUD.0000000000000376

16. Griffin AM, Poissant SF, Freyman RL. Speech-in-noise and quality-of-life measures in school-aged children with normal hearing and with unilateral hearing loss. *Ear Hear*. (2019) 40:887–904. doi: 10.1097/AUD.0000000000000667

17. Corbin NE, Buss E, Leibold LJ. Spatial hearing and functional auditory skills in children with unilateral hearing loss. *J Speech Lang Hear Res*. (2021) 64:4495–512. doi: 10.1044/2021_JSLHR-20-00081

18. Morrongiello BA. Infants' monaural localization of sounds: effects of unilateral ear infection. *J Acoust Soc Am*. (1989) 86:597–602. doi: 10.1121/1.398749

19. Auslander MC, Lewis DE, Schulte L, Stelmachowicz PG. Localization ability in infants with simulated unilateral hearing loss. *Ear Hear*. (1991) 12:371–6. doi: 10.1097/00003446-199112000-00001

20. Arras T, Boudewyns A, Swinnen F, Zarowski A, Philips B, Desloovere C, et al. Longitudinal auditory data of children with prelingual single-sided deafness managed with early cochlear implantation. *Sci Rep*. (2022) 12. doi: 10.1038/S41598-022-13247-5

21. Lieu JEC, Tye-Murray N, Karzon RK, Piccirillo JF. Unilateral hearing loss is associated with worse speech-language scores in children. *Pediatrics*. (2010) 125:e1348–e55. doi: 10.1542/peds.2009-2448

22. Lieu JEC. Speech-language and educational consequences of unilateral hearing loss in children. *Arch Otolaryngol Head Neck Surg*. (2004) 130:524–30. doi: 10.1001/archotol.130.5.524

23. Lieu JEC. Permanent unilateral hearing loss (UHL) and childhood development. *Curr Otorhinolaryngol Rep*. (2018) 6:74–81. doi: 10.1007/S40136-018-0185-5

24. Purcell PL, Shinn JR, Davis GE, Sie KCY. Children with unilateral hearing loss may have lower intelligence quotient scores: a meta-analysis. *Laryngoscope*. (2016) 126:746–54. doi: 10.1002/lary.25524

25. Ead B, Hale S, DeAlwis D, Lieu JEC. Pilot study of cognition in children with unilateral hearing loss. *Int J Pediatr Otorhinolaryngol*. (2013) 77:1856–60. doi: 10.1016/J.IJPORL.2013.08.028

26. Bess FH, Davis H, Camarata S, Hornsby BWY. Listening-related fatigue in children with unilateral hearing loss. *Lang Speech Hear Serv Sch*. (2020) 51:84–97. doi: 10.1044/2019_LSHSS-OCHL-19-0017

27. Adams B, Thornton SK, Naylor G, Spriggs RV, Wiggins IM, Kitterick PT. Exploring listening-related fatigue in children with and without hearing loss using self-report and parent-proxy measures. *Front Pediatr*. (2023) 11:1127578. doi: 10.3389/FPED.2023.1127578

28. Agterberg MJH, Snik AFM, Van de Goor RMG, Hol MKS, Van Opstal AJ. Sound-localization performance of patients with single-sided deafness is not improved when listening with a bone-conduction device. *Hear Res*. (2019) 372:62. doi: 10.1016/J.HEARES.2018.04.007

29. Snik FM, Teunissen B, Cremers WRJ. Speech recognition in patients after successful surgery for unilateral congenital ear anomalies. *Laryngoscope*. (1994) 104:1029–34. doi: 10.1288/00005537-199408000-00020

30. Stenfelt S, Goode RL. Bone-conducted sound: physiological and clinical aspects. *Otol Neurotol*. (2005) 26:1245–61. doi: 10.1097/01.MAO.0000187236.10842.D5

31. Koci V, Seebacher J, Weichbold V, Zorowka P, Wolf-Magele A, Sprinzl G, et al. Improvement of sound source localization abilities in patients bilaterally supplied with active middle ear implants. *Acta Otolaryngol*. (2016) 136:692–8. doi: 10.3109/00016489.2016.1155232

32. Vyskocil E, Liepins R, Kaider A, Blineder M, Hamzavi S. Sound localization in patients with congenital unilateral conductive hearing loss with a transcutaneous bone conduction implant. *Otol Neurotol*. (2017) 38:318–24. doi: 10.1097/MAO.0000000000001328

33. Liu CC, Livingstone D, Yunker WK. The role of bone conduction hearing aids in congenital unilateral hearing loss: a systematic review. *Int J Pediatr Otorhinolaryngol*. (2017) 94:45–51. doi: 10.1016/J.IJPORL.2017.01.003

34. Liu Y, Zhao C, Yang L, Chen P, Yang J, Wang D, et al. Characteristics of sound localization in children with unilateral microtia and atresia and predictors of localization improvement when using a bone conduction device. *Front Neurosci*. (2022) 16:973735. doi: 10.3389/FNINS.2022.973735/BIBTEX

35. Vogt K, Frenzel H, Ausili SA, Hollfelder D, Wollenberg B, Snik AFM, et al. Improved directional hearing of children with congenital unilateral conductive hearing loss implanted with an active bone-conduction implant or an active middle ear implant. *Hear Res*. (2018) 370:238. doi: 10.1016/J.HEARES.2018.08.006

36. Vogt K, Wasmann JW, Van Opstal AJ, Snik AFM, Agterberg MJH. Contribution of spectral pinna cues for sound localization in children with congenital unilateral conductive hearing loss after hearing rehabilitation. *Hear Res*. (2020) 385:107847. doi: 10.1016/J.HEARES.2019.107847

37. Huttunen K, Erixon E, Löfkvist U, Mäki-Torkko E. The impact of permanent early-onset unilateral hearing impairment in children: a systematic review. *Int J Pediatr Otorhinolaryngol*. (2019) 120:173–83. doi: 10.1016/J.IJPORL.2019.02.029

38. Appachi S, Specht JL, Raol N, Lieu JEC, Cohen MS, Dedhia K, et al. Auditory outcomes with hearing rehabilitation in children with unilateral hearing loss: a systematic review. *Otolaryngol Head Neck Surg*. (2017) 157:565–71. doi: 10.1177/0194599817726757

39. Dawes P. Hearing interventions to prevent dementia. *HNO*. (2019) 67:165–71. doi: 10.1007/s00106-019-0617-7

40. Naylor G, Dillard L, Orrell M, Stephan BCM, Zobay O, Saunders GH. Dementia and hearing-aid use: a two-way street. *Age Ageing*. (2022) 51:afac266. doi: 10.1093/AGEING/AFAC266

41. Jayakody DMP, Wishart J, Stegeman I, Eikelboom R, Moyle TC, Yiannos JM, et al. Is there an association between untreated hearing loss and psychosocial outcomes? *Front Aging Neurosci*. (2022) 14:868673. doi: 10.3389/FNAGI.2022.868673

42. Ching TY, Dillon H, Leigh G, Cupples L. Learning from the longitudinal outcomes of children with hearing impairment (LOCHI) study: summary of 5-year findings and implications HHS public access. *Int J Audiol*. (2018) 57:105–11. doi: 10.1080/14992027.2017.1385865

43. Pure tone air and bone conduction threshold audiometry with and without masking—British Society of Audiology. Available at: https://www.thebsa.org.uk/resources/pure-tone-air-bone-conduction-threshold-audiometry-without-masking/ (Accessed January 20, 2023).

44. Horrocks LM, Kitterick PT, Jayasinghe DS, Willis KR, Martin KRM, Dixit A, et al. Multiple congenital anomalies and adverse developmental outcomes are associated with neonatal intensive care admission and unilateral hearing loss. *Front Pediatr*. (2023) 10:1068884. doi: 10.3389/FPED.2022.1068884

45. Quality standards in paediatric audiology. BAA (British Academy 1028 of Audiology. (2022) p. 1–22.

46. Berninger E, Westling B. Outcome of a universal newborn hearing-screening programme based on multiple transient-evoked otoacoustic emissions and clinical brainstem response audiometry. *Acta Otolaryngol*. (2011) 131:728–39. doi: 10.3109/00016489.2011.554440

47. Paul A, Marlin S, Parodi M, Rouillon I, Guerlain J, Pingault V, et al. Unilateral sensorineural hearing loss: medical context and etiology. *Audiol Neurootol*. (2017) 22:83–8. doi: 10.1159/000474928

48. Masuda S, Usui S. Comparison of the prevalence and features of inner ear malformations in congenital unilateral and bilateral hearing loss. *Int J Pediatr Otorhinolaryngol*. (2019) 125:92–7. doi: 10.1016/j.ijporl.2019.06.028

49. Masuda S, Usui S, Matsunaga T. High prevalence of inner-ear and/or internal auditory canal malformations in children with unilateral sensorineural hearing loss. *Int J Pediatr Otorhinolaryngol*. (2013) 77:228–32. doi: 10.1016/j.ijporl.2012.11.001

50. Friedman AB, Guillory R, Ramakrishnaiah RH, Frank R, Gluth MB, Richter GT, et al. Risk analysis of unilateral severe-to-profound sensorineural hearing loss in children. *Int J Pediatr Otorhinolaryngol*. (2013) 77:1128–31. doi: 10.1016/j.ijporl.2013.04.016

51. Niu K, Brandström A, Skenbäck S, Duan M, Uhlén I. Risk factors and etiology of childhood hearing loss: a cohort review of 296 subjects. *Acta Otolaryngol*. (2020) 140:660–6. doi: 10.1080/00016489.2020.1757753

52. Glob J. Genetic hearing loss-syndromes locus name-DFNA. *Glob J Otolaryngol*. (2017) 5. doi: 10.19080/GJO.2017.05.555665

53. Kiese-Himmel C. Unilateral sensorineural hearing impairment in childhood: analysis of 31 consecutive cases. *Int J Audiol*. (2002) 41:57–63. doi: 10.3109/14992020209101313

54. Walker EA, Spratford M, Moeller MP, Oleson J, Ou H, Roush P, et al. Predictors of hearing aid use time in children with mild-to-severe hearing loss. *Lang Speech Hear Serv Sch*. (2013) 44:73–88. doi: 10.1044/0161-1461(2012/12-0005)

55. Johnstone PM, Nábělek AK, Robertson VS. Sound localization acuity in children with unilateral hearing loss who wear a hearing aid in the impaired ear. *J Am Acad Audiol*. (2010) 21:522–34. doi: 10.3766/JAAA.21.8.4

56. Russo FY, De Seta D, Orlando MP, Ralli M, Cammeresi MG, Greco A, et al. Hearing attention and quality of listening in children with unilateral hearing loss with and without hearing aid. *Acta Otorhinolaryngol Ital*. (2022) 42:169–75. doi: 10.14639/0392-100X-N1746

57. Chandrasekar B, Hogg ES, Patefield A, Strachan L, Sharma SD. Hearing outcomes in children with single sided deafness: our experience at a tertiary paediatric otorhinolaryngology unit. *Int J Pediatr Otorhinolaryngol*. (2023) 167:111296. doi: 10.1016/J.IJPORL.2022.111296

58. Entwisle LK, Warren SE, Messersmith JJ. Cochlear implantation for children and adults with severe-to-profound hearing loss. *Semin Hear*. (2018) 39:390–404. doi: 10.1055/S-0038-1670705

59. Calcus A, Tuomainen O, Campos A, Rosen S, Halliday LF. Functional brain alterations following mild-to-moderate sensorineural hearing loss in children. *eLife*. (2019) 8:e46965. doi: 10.7554/ELIFE.46965

60. Sharma SD, Cushing SL, Papsin BC, Gordon KA. Hearing and speech benefits of cochlear implantation in children: a review of the literature. *Int J Pediatr Otorhinolaryngol*. (2020) 133:109984. doi: 10.1016/j.ijporl.2020.109984

61. Oghalai JS, Bortfeld H, Feldman HM, Chimalakonda N, Emery C, Choi JS, et al. Cochlear implants for deaf children with early developmental impairment. *Pediatrics*. (2022) 149(6):e2021055459. doi: 10.1542/PEDS.2021-055459/188094

62. Davis A, Reeve K, Hind S, Bamford J. Children with mild and unilateral hearing loss. In: *A sound foundation through early amplification*. Proceedings of the Second International Conference (2001). p. 179–86.

63. Visram AS, Roughley AJ, Hudson CL, Purdy SC, Munro KJ. Longitudinal changes in hearing aid use and hearing aid management challenges in infants. *Ear Hear*. (2021) 42:961–72. doi: 10.1097/AUD.0000000000000986

64. Purcell PL, Jones-Goodrich R, Wisneski M, Edwards TC, Sie KCY. Hearing devices for children with unilateral hearing loss: patient- and parent-reported perspectives. *Int J Pediatr Otorhinolaryngol*. (2016) 90:43–8. doi: 10.1016/J.IJPORL.2016.08.029

65. Joo H, Huang EY, Schoo D, Agrawal Y, Chen JX. Bullying victimization in adolescents with hearing impairment. *Int J Pediatr Otorhinolaryngol*. (2023) 167. doi: 10.1016/J.IJPORL.2023.111489

66. Gaffney H, Ttofi MM, Farrington DP. Evaluating the effectiveness of school-bullying prevention programs: an updated meta-analytical review. *Aggress Violent Behav*. (2019) 45:111–33. doi: 10.1016/J.AVB.2018.07.001

67. Lieu JEC, Tye-Murray N, Fu Q. Longitudinal study of children with unilateral hearing loss. *Laryngoscope*. (2012) 122:2088–95. doi: 10.1002/LARY.23454

68. Rachakonda T, Shimony JS, Coalson RS, Lieu JEC. Diffusion tensor imaging in children with unilateral hearing loss: a pilot study. *Front Syst Neurosci*. (2014) 8: e2021055459. doi: 10.3389/FNSYS.2014.00087

69. Lindburg M, Ead B, Jeffe DB, Lieu JEC. Hearing loss–related issues affecting quality of life in preschool children. *Otolaryngol Head Neck Surg*. (2021) 164:1322–9. doi: 10.1177/0194599820962475

70. Katiri R, Hall DA, Hoare DJ, Fackrell K, Horobin A, Hogan N, et al. The Core Rehabilitation Outcome Set for Single-Sided Deafness (CROSSSD) study: international consensus on outcome measures for trials of interventions for adults with single-sided deafness and for the Core Rehabilitation Outcome Set for Single-Sided Deafness (CROSSSD) initiative. *Trials*. (2022) 23:764. doi: 10.1186/s13063-022-06702-1

71. Fortnum H, Ukoumunne OC, Hyde C, Taylor RS, Ozolins M, Errington S, et al. A programme of studies including assessment of diagnostic accuracy of school hearing screening tests and a cost-effectiveness model of school entry hearing screening programmes. *Health Technol Assess*. (2016) 20:1–210. doi: 10.3310/HTA20360

72. Ching TYC, Crowe K, Martin V, Day J, Mahler N, Youn S, et al. Language development and everyday functioning of children with hearing loss assessed at 3 years of age. *Int J Speech Lang Pathol*. (2010) 12:124–31. doi: 10.3109/17549500903577022

73. Moeller MP, McCleary E, Putman C, Tyler-Krings A, Hoover B, Stelmachowicz P. Longitudinal development of phonology and morphology in children with late-identified mild-moderate sensorineural hearing loss. *Ear Hear*. (2010) 31:625–35. doi: 10.1097/AUD.0B013E3181DF5CC2

74. Shojaei E, Jafari Z, Gholami M. Effect of early intervention on language development in hearing-impaired children. *Iran J Otorhinolaryngol*. (2016) 28:13–21. doi: 10.3310/hta20360

75. NDCS. Right from the start a campaign to improve early years support for deaf children. Published by the National Deaf Children's Society (2016) p. 1–34. Available at: https://www.ndcs.org.uk/media/1283/right_from_the_start_campaign_report_final.pdf (Accessed March 21, 2023).

76. Haffey T, Fowler N, Anne S. Evaluation of unilateral sensorineural hearing loss in the pediatric patient. *Int J Pediatr Otorhinolaryngol*. (2013) 77:955–8. doi: 10.1016/j.ijporl.2013.03.015

77. Kumpik DP, King AJ. A review of the effects of unilateral hearing loss on spatial hearing. *Hear Res*. (2019) 372:17–28. doi: 10.1016/J.HEARES.2018.08.003

78. Keating P, Dahmen JC, King AJ. Context-specific reweighting of auditory spatial cues following altered experience during development. *Curr Biol*. (2013) 23:1291–9. doi: 10.1016/J.CUB.2013.05.045

79. Kim JH, Shim L, Bahng J, Lee HJ. Proficiency in using level cue for sound localization is related to the auditory cortical structure in patients with single-sided deafness. *Front Neurosci*. (2021) 15:1345. doi: 10.3389/FNINS.2021.749824/BIBTEX

80. Arras T, Snapp H, Sangen A, Snels C, Kuntz I, Theunen T, et al. Instant improvement in monaural spatial hearing abilities through cognitive feedback. *Exp Brain Res*. (2022) 240:1357. doi: 10.1007/S00221-022-06333-7

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EDITED BY
Andrea Ciorba,
University of Ferrara, Italy

REVIEWED BY
Ryan William McCreery,
Boys Town National Research Hospital, United States
Meredith Spratford,
Boys Town National Research Hospital, United States

*CORRESPONDENCE
Valerie Sung
valerie.sung@rch.org.au

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Mild matters: trial learnings and importance of community engagement in research for early identified bilateral mild hearing loss

Valerie Sung[1,2,3]*, Teresa Y. C. Ching[4,5,6], Libby Smith[1,2], Vivienne Marnane[7], Michelle Saetre-Turner[7], Alison King[8], Rachael Beswick[9], Claire E. Iseli[10,11] and Peter Carew[1,12]

[1]Prevention Innovation, Population Health, Murdoch Children's Research Institute, Parkville, VIC, Australia, [2]Centre for Community Child Health, Royal Children's Hospital, Parkville, VIC, Australia, [3]Department of Paediatrics, University of Melbourne, Parkville, VIC, Australia, [4]NextSense, NextSense Institute, Sydney, NSW, Australia, [5]Macquarie School of Education, Macquarie University, Sydney, NSW, Australia, [6]School of Health & Rehabilitation Sciences, The University of Queensland, St Lucia, QLD, Australia, [7]National Acoustic Laboratories, Sydney, NSW, Australia, [8]Hearing Australia, Doncaster, VIC, Australia, [9]Children's Health Queensland Hospital and Health Service, South Brisbane, QLD, Australia, [10]Otolaryngology Department, Royal Children's Hospital, Parkville, VIC, Australia, [11]Department of Surgery, University of Melbourne, Parkville, Victoria, Australia, [12]Department of Audiology and Speech Pathology, University of Melbourne, Parkville, VIC, Australia

Introduction: Early identification of mild hearing loss has resulted in early hearing amplification without adequate evidence of effectiveness. This paper describes learnings from a pilot trial, combined with a qualitative study, to highlight the importance of community engagement in designing research studies to determine whether early amplification benefits young children with bilateral mild hearing loss.

Methods: PART 1 of the study is a proof-of-concept non-blinded multi-centre randomised controlled trial (RCT) of hearing device fitting vs. no fitting aimed to gather preliminary data and determine its acceptability/feasibility in children <2 years old with bilateral mild hearing loss.

Results: PART 2 is a qualitative study to understand the barriers/enablers to RCT participation. Of 40 potentially eligible families, nine (23%) declined, three were uncontactable (7%), 26 (65%) ineligible: of these, nine (35%) did not meet hearing threshold inclusion criteria, 11 (42%) were already fitted or had made decisions on fitting hearing device, two (7%) had conductive loss and four (16%) were ineligible for other reasons. Two of 11 (18%) eligible families were randomised. With the limited sample size, outcome measures were not compared between groups. Both participants completed the trial, reported the RCT to be acceptable, and neither changed group post-enrolment.

Discussion: Whilst recruitment uptake could potentially be increased by altering the eligibility criteria, better communication with and reimbursement of clinicians as recruiters, and improving awareness of the study amongst external stakeholders, the RCT methodology does not conform to family-centred practice, and potentially raises ethical concerns regarding potential adverse consequences of not offering early amplification. Parental perception of losing

control over choice of management due to randomisation is not an easily modifiable factor. Alternative methodological approaches without randomisation are required to determine whether hearing amplification benefits infants with mild hearing loss.
Clinical Trial Registration: identifier [ACTRN12618001608257].

KEYWORDS

pediatric mild bilateral hearing loss, hearing amplification, newborn hearing screening, randomized controlled clinical trial (RCT), acceptability and feasibility

1. Introduction

Congenital hearing loss affects 1–3 in every 1,000 children, and can have adverse impacts on communication, social and emotional development, and academic outcomes in children with flow-on effects on employment and quality of life in adulthood (1). In Australia, universal newborn hearing screening (UNHS) has led to detection of any degree of hearing loss, from mild to profound, very soon after birth so that infants can receive intervention (hearing devices, cochlear implants, speech/sign intervention) early in the pre-lingual years (2). While UNHS in Australia does not aim to detect hearing loss of less than moderate degree, mild losses are being detected as a by-product. The National Workshop on Mild and Unilateral Hearing Loss defined permanent mild bilateral hearing loss as "when the diagnosis indicates there is, in both ears, a calculated or predicted average pure tone air conduction threshold at 0.5, 1, 2 kHz between 20 and 40 decibels hearing level (dB HL) or pure tone air conduction thresholds greater than 25 dB HL at two or more frequencies above 2 kHz (i.e., 3, 4, 6, 8 kHz)"(3).

Earlier detection has led to improvements in outcomes for children with moderate or greater degrees of hearing loss (2, 4–7); however, this may not be the case for children with mild hearing loss (5, 8). The studies that have examined outcomes of mild hearing loss mainly reported on school-aged children and have mixed results. Some studies reported school-aged children with mild hearing loss to have higher grade-retention rates and more dysfunction in the domains of stress, social support and self-esteem than children with normal hearing (9, 10), while other studies have not demonstrated the same (8, 11, 12). Differences in study methodology may account for some of this outcome variability. Some report outcomes only for children with slight/mild bilateral loss (11), vs. minimal (mild bilateral and unilateral combined) losses. Others recruited from populations of children known to have hearing loss (12), vs. those who reported outcomes of children with hearing loss detected via large population screenings.

A recent study of 5–7-year-olds with mild hearing loss found that full time hearing device users performed significantly better on grammar and vocabulary measures than non-users, but found no difference in articulation or speech perception (13). Other reports of school aged children, including those with mild hearing loss, have shown aided hearing can support listening comprehension (14) and oral language outcomes (15). A few studies of younger children with mild hearing loss, who received newborn hearing screening and were majority engaged with early intervention services, suggested they did as well as their normally hearing peers (16, 17). Fitzpatrick et al. assessed the outcomes at four years of a group of infants identified with unilateral and bilateral mild hearing loss; the majority (80%) of infants were recommended for amplification (17). Many parents of young children with mild hearing loss do not perceive clear benefits of early hearing amplification while others feel more positive (18, 19). There have been no randomised controlled trials (RCTs) examining the effectiveness of early hearing devices on pre-lingual children with mild hearing loss.

In the past, clinical management of mild hearing loss has relied on auditory considerations about deprivation and assessment of developmental progress in post-lingual children (20). More recently however, increasing numbers of pre-lingual infants/children with mild hearing loss are being fitted with hearing devices due to increasing detection of mild losses within weeks of birth. In 2020, Hearing Australia [the national government-subsidised hearing service provider for all children and youth aged 26 years and under in Australia (21)] recorded that 56.7% of hearing device fittings in Australian children less than 2 years old had mild hearing loss in the better ear of ≤40 dB HL (22). Two recent studies examined the parental and audiologist perceptions of early management of mild bilateral hearing loss. Parents reported significant stress around the diagnostic processes, guilt about the potential future negative effects of not fitting hearing devices for their infants, and a multitude of challenges around hearing device compliance and maintenance (18). Many parents felt the decision for hearing device fitting was often left up to them to make (18). This was reflected in audiologists reporting that they considered multiple child and family-related factors and the perspectives of parents and families in making decisions about fitting in this population (23). Indeed, audiologists perceived the clinical management of these children to be challenging, mainly due to the lack of evidence to guide management (23).

Evidence on the effectiveness of early hearing device fitting in infants and pre-lingual children with mild hearing loss is therefore needed to guide management of these children, especially in the face of healthcare costs and potentially significant burdens for these families and society. We attempted to answer this research question through a proof-of-concept RCT aimed to gather preliminary data, to be used towards planning for a possible future more definitive RCT, to compare, in children less than 2 years old with bilateral mild hearing loss, language outcomes of those fitted with hearing devices vs. those without hearing device fitting, 6 months post-randomisation. The secondary aims were

to collect 6 months post-randomisation data on child social abilities, functional performance and listening effort, parental morale, parent-child relationship and quality of life, as well as determine the acceptability and feasibility of the RCT. However, as our trial failed to recruit sufficient participants, we engaged clinical audiologists and families of young children with mild bilateral hearing loss to conduct a qualitative study to understand the barriers/enablers to RCT participation (PART 2). This paper overall aims to describe our learnings from both the RCT and the qualitative study to highlight the importance of community engagement to help develop the impetus, design and implementation of future research studies to determine whether early amplification benefit young children with bilateral mild hearing loss.

2. Materials and methods

2.1. PART 1: randomised controlled trial

Here, we describe the essential details of our trial's recruitment methodology in the context of an unsuccessful trial from which key lessons were learnt. The RCT is registered with the Australian and New Zealand Clinical Trials Registry (ACTRN12618001608257); the full protocol is available on the ANZCTR website. The study has ethics approval from the Royal Children's Hospital Human Research and Ethics Committee HREC 38112 (HREC/45275/RCHM-2018-151266).

We set out to conduct a proof-of-concept non-blinded multi-centre RCT comparing hearing devices (intervention) with no hearing devices (control) in children less than 2 years old with bilateral mild hearing loss (21 to 40 dB HL) across at least 3 octave frequencies between 250 and 4000 Hz by objective or behavioural testing. Recruitment occurred in three states in Australia: Victoria (VIC), New South Wales (NSW) and Queensland (QLD). Children must have met all of the following criteria to be enrolled in the study: (a) born in VIC, NSW, or QLD and eligible for services of Hearing Australia (Australian resident/citizenship status), (b) less than 2 years old, (c) had parents/carers who spoke English adequately to give consent, (d) had, within the last 3 months, been confirmed to have bilateral mild hearing loss (21 to 40 dB HL) across at least 3 octave frequencies between 250 and 4,000 Hz by objective or behavioural testing, and (d) had pure sensorineural hearing loss. Children with any of the following criteria were excluded from the study: (a) families who had already made a decision of fitting/not fitting hearing devices for their children, or children who were already fitted with hearing devices, (b) complex medical problems/major disabilities (e.g., recurrent seizures, major cardiac problems requiring multiple operations), (c) any conductive hearing loss, (d) hearing threshold of <21 dB or >40 dB HL at any frequency, (e) medical contraindication to hearing device fitting, and (f) families who definitively planned to move, during the following 6 months, to a location where follow-up assessment was not possible or practical.

Six months after the study started, in response to the poor recruitment rate, inclusion criteria for hearing thresholds were broadened to the following: had, within the last 3 months, been confirmed to have bilateral mild hearing loss [at least three frequency average ≤40 dB eHL (estimated hearing level) between 250 and 4,000 Hz] by objective or behavioural testing. That is, children with three- or four-frequency average threshold of <21 dB or >40 dB HL were excluded.

The study was conducted at Hearing Australia clinics in the three states from 1st February 2019 to 31st January 2020. At enrolment, the participant child was randomised to either intervention or control. Children in the intervention group received hearing devices as per standard Hearing Australia protocol, and were followed up for hearing device compliance (monthly parent-report via a five-question survey sent to the parent's mobile phone or email, and hearing device data logging) over 6 months. They also received normal audiological care and parental support as clinically required following Hearing Australia protocol over 6 months, including assessments and clinical counselling by a Hearing Australia audiologist regarding hearing devices fitting, fitting adjustment and follow up appointments at the Hearing Australia centre (21). Timing of fitting, model of hearing devices and clinical care were determined by the Hearing Australia audiologist. Children in the control group were not fitted with hearing devices and received normal audiological care and parental support as clinically required following Hearing Australia protocol over 6 months, including assessments and clinical counselling by a Hearing Australia audiologist.

The randomization methodology, primary and secondary outcome measures are available from the ANZCTR website and are not reported here as they are not the focus of our learnings in this paper. The feasibility of the RCT was measured by the: (a) number of children enrolled as a proportion of eligible children; (b) number of children who dropped out as a proportion of enrolled children; (c) number of children who changed treatment group from original treatment allocation, as a proportion of enrolled children; and (d) device use—measured by automated data logging in hearing devices over 3 months, and monthly parent report on proportion of device use during waking hours over the last week, during the 6 months after fitting. The acceptability of the RCT was measured by parent-report at the 6 month follow-up through survey with the following questions: (a) "How do you feel about your child being allocated to the hearing aids group vs. the no hearing aids group?", (b) "Overall, do you feel your child has been advantaged or disadvantaged by being assigned to a fitting or no fitting group?", and by free text responses.

We had estimated our expected sample size according to the known incidence of mild bilateral hearing loss [0.4/1,000 newborns (24)]; approximately 100 infants would be eligible from all 3 states over one year. Anticipating a consent rate of 60% and a drop-out rate of 20%, approximately 48 children with mild bilateral hearing loss would be enrolled in the RCT over the study period of one year, with approximately 24 in each intervention arm. The expected recruitment numbers per site

were ~15 from VIC, ~18 from NSW and ~15 from QLD. Data collection was via REDCap.

2.2. PART 2: qualitative study

Subsequent to determining the feasibility of the RCT, a qualitative study was undertaken to explore the factors that influenced parental uptake of the RCT. This qualitative study received ethics approval (as above). Over a four-month period from June 2020 to September 2020, we invited caregivers who met the same eligibility criteria for the RCT, but without excluding those who had already made a decision of fitting/not fitting hearing devices for their child, to participate in a semi-structured phone interview and complete a basic demographic child and parent questionnaire via REDCap (see **Supplementary Material Interview Guide**). Due to COVID-19 restrictions in place at the time of data collection, face-to-face interviews could not be offered. During the 4-month period, we also invited by email diagnostic and rehabilitation audiologists to participate in semi-structured phone or videoconference interviews to explore their perceptions of factors that influenced parental uptake of an RCT. Audiologists also filled in a brief demographic questionnaire via REDCap about their audiological experience and frequency of managing children with mild hearing loss. For both caregiver and audiologist groups, data collection continued until no new themes emerged (saturation of themes). This was verified during data collection through reflective discussion after each interview between the researcher conducting the interview and project team members. Two researchers conducted the interviews, one completing the caregiver group and the other completing the audiologist group. Interviews were transcribed using a third party transcription service, and transcriptions were reviewed by the researchers who conducted each interview for accuracy and to allow reflexive thought to identify any assumptions the researcher may bring to the research purpose (25).

We theorised that factors influencing parental uptake in a RCT worked together to influence the decision-making process, therefore Grounded Theory methodology was applied for analysis of all interviews together. Grounded Theory is an iterative, inductive methodology that results in the generation of a theoretical explanatory process relating to a phenomenon, in this case the parental decision making process (26). In a similar approach to other qualitative analysis methods such as thematic analysis, interview transcripts were coded to categorise and assign meaning to data to allow the identification of similarities, differences, and patterns. Through iterative processes the coding was organised within a framework that denoted interactions between concepts (27).

The same researchers who completed the interviews independently completed initial coding of each interview transcript, and the two researchers discussed areas of discrepancy until consensus was reached on how to complete initial coding of all transcripts. A further six transcripts underwent initial coding and discussion held with the wider research team (VS, TC, VM, LM, MS, RB) to ensure consensus on initial codes before all transcripts were coded. After initial coding was completed, a small group of researchers met (VM, MS, LM) to undergo intermediate coding. Intermediate coding is the process of identifying categories and concepts from the initial codes and beginning to identify relationships between the concepts for a theory to merge from the data. The initial theory on parental decision making to participate in a RCT was discussed with a wider research team (VS, TC, LS, VM, LM, MS, RB) and then refined in the final stage of advanced coding, where the final framework was derived and interrelated concepts were established.

3. Results

3.1. PART 1: RCT

Here we report only on the outcomes of recruitment for the RCT in view of the aims of this paper. Forty infants were referred to the study. Fifteen referrals were from VIC, 16 from NSW, and 9 from Queensland. All referrals were from audiologists or the Victorian Infant Hearing Screening Program. No referrals were from ENT specialists or paediatricians. Of the 40 potentially eligible families, 3 could not be contacted, 26 were ineligible and 11 were eligible. Of those eligible, 2 participated and 9 declined (**Figure 1**).

Table 1 shows the reasons for ineligibility. The main reason for ineligibility was not meeting the audiometric threshold criteria (35%). The study team identified this as an issue halfway through the recruitment period and relaxed the inclusion criteria. Six families (23%) had already made the decision to fit hearing aids or had decided that they did not want hearing aids. Five infants were already fitted with hearing aids (19%). Other infants were excluded because the family had insufficient English to consent, were moving away, or the infant had conductive hearing loss, or had complex medical issues contraindicating hearing aid fitting.

Two infants participated. One was randomised to the intervention "hearing devices" group and the other to the control "no hearing devices" group. Both participants were followed up at 6 months. Follow-up of outcomes concluded by 16th April 2020. The participant characteristics, and the primary and secondary outcomes for each participant, are reported in the **Supplementary Tables**. We did not compare primary and secondary outcomes between the two groups due to the limited sample size.

The following describes the outcomes of the feasibility measures:

- 2 out of 11 (18%) eligible participants consented to take part.
- Neither of the two participants dropped out of the study.
- Neither of the participants changed treatment group from the original treatment allocation.
- Device use: The participant parent from the intervention group completed 5 out of 6 monthly surveys on device use. The child was ill during two of the reporting weeks and the parent indicated they were not typical weeks for device use, so these

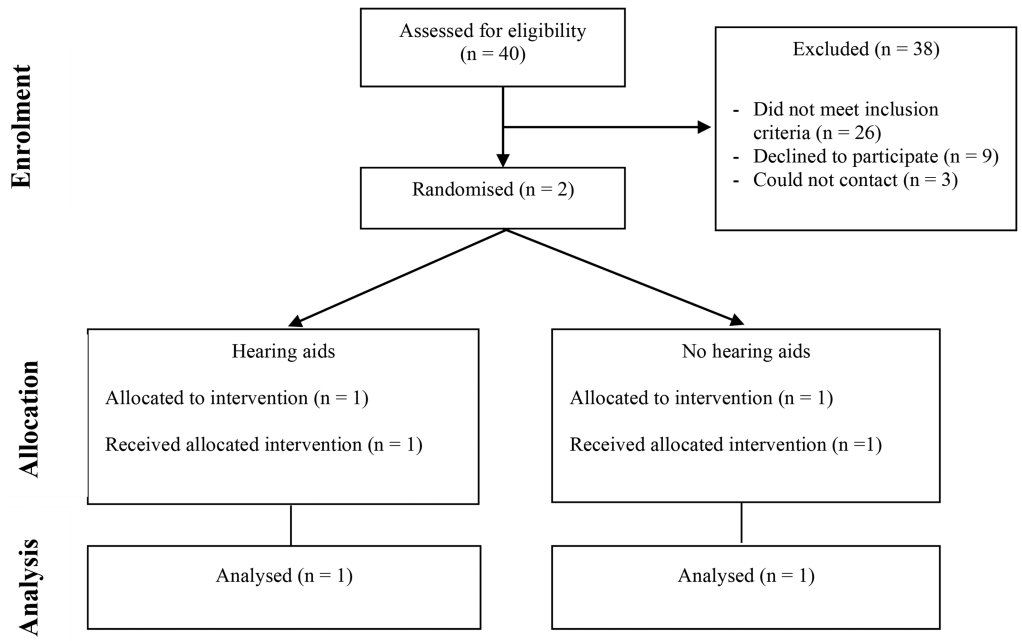

FIGURE 1
Participant flowchart.

two weeks were excluded from analysis. By parent report, devices were worn on average by the participant for 47% of waking hours on weekdays (3.5 h), and 21% of waking hours on weekend days (1.7 h). This is compared to data logging indicating the participant wore the devices on average 4.6 h (right) and 4.5 h (left) per day over 3 months of the study period.

Both participant families filled in surveys about the acceptability of the study. They reported positive or neutral feelings about participating in the study. The intervention family reported feeling positive about their child being allocated to that group and felt their child was "highly advantaged", commenting that being assigned to the fitting group was "good for his learning development". The control family felt "neutral" about being allocated to that group and did not feel advantaged or disadvantaged. They commented that they were glad their child "didn't have the trouble of a hearing aid" but were also "worried that it might have been good for her". They also indicated they understood the purpose of the study well with the comment "I suppose that's the whole point of the study—we just don't know what's best!".

TABLE 1 Reasons for ineligibility (total *n* = 26).

Reason ineligible	Frequency (%)
Did not meet hearing threshold inclusion criteria	9 (35%)
Already decided to fit/not fit hearing aids	6 (23%)
Already fitted	5 (19%)
Conductive or other hearing loss	2 (7%)
Insufficient English	1 (4%)
Complex medical	1 (4%)
Moving away	1 (4%)
More than 3 months since diagnosis	1 (4%)

3.2. PART 2: qualitative study

3.2.1. Recruitment

Ten caregivers and 11 audiologists completed interviews. Tables 2, 3 outline the demographic characteristics of both groups.

3.2.2. Interview outcomes

The decision that a parent would ultimately make was the result of a complex interplay of: (1) individual circumstances and beliefs, (2) study design factors, (3) perceived benefits of participation, and (4) perceived costs of participation. These four themes and their corresponding subthemes generated a decision-making framework that was underpinned by a major theme of "parent altruism" that was common across interviews and represented in Figure 2.

3.2.2.1. Overarching theme: parent altruism

Overwhelmingly, parents spoke of the challenges they faced when making decisions about hearing amplification for their child. Perhaps unlike parents of children with more severe hearing loss, parents of children with mild hearing loss experienced "overwhelming indecision around whether to get hearing aids" [P401]. Parents felt the need to do the right thing by their child:

TABLE 2 Demographic characteristics of child and caregiver for interviews.

ID	Gender	Age of child (months)	Fitted with hearing aids	Parent with hearing loss	Primary language spoken at home	Caregiver completing interview
P201	Male	2	Yes	Father	Cantonese	Father
P203	Female	9	No	Mother	English	Mother
P205	Male	3	No	Father	English	Mother
P206	Male	3	No	No	Maltese	Mother
P306	Female	3	Yes	No	English	Mother
P311	Female	2	No	No	Greek	Mother
P312	Female	1	No	No	English	Mother
P401	Male	3	No	No	English	Mother
P402	Female	4	No	No	English	Mother
P404	Male	4	Yes	No	English	Mother
P405	Male	5	Yes	No	English	Mother

> *Family say, "Well just do what"s best for her", and I was like, yeah, well I'm gunna, but I didn't know what that is* [P312].

Many parents had experienced high levels of stress making decisions around hearing aid fitting due to the lack of evidence surrounding the effectiveness of fitting hearing aids for this population. As such, a common theme was feeling inclined to participate in the trial for altruistic reasons, or "the general good of everyone" [P201]:

> *When we were trying to make the decision about whether to get aids or not, basically it felt really hard to make the decision because there was no information and there was no studies that were really conclusive. I'd want to participate so that future people could make the decision easier.* [P401]

Similarly, audiologists with experience attempting to recruit parents into the trial described how the uncertainty surrounding whether to fit hearing aids would lead parents to act altruistically. Parents would "*jump on anything they can do to help*" [A405]:

> *I found that sometimes they're a little bit easier to recruit only because there's that 'Do we fit? Do we not fit?' Some families are like 'Oh, I'd really love to know. I'd love to be part of this, if it makes that decision easier for somebody else down the track.'* [A201]

The desire to help future parents was often the first factor mentioned when discussing trial participation. When probed further on their decision-making process, parents would relate their reasoning to this anchoring argument, either reinforcing or weakening the initial desire to participate. This is illustrated in the decision-making overview in **Figure 2** and explained further through the remaining themes and subthemes.

3.2.2.2. *Individual circumstances and preferences*

During the interviews, parents and audiologists described how a family's circumstances and preferences would influence their willingness or capacity to participate in the trial. Two subthemes were identified: timing of study presentation, and hearing aid preferences.

3.2.2.2.1. Study timing: during coping with the diagnosis. Recruitment into the hypothetical trial and subsequent randomisation (fitted or not fitted with hearing aids) would have occurred when the child was very young, soon after the diagnosis of hearing loss was made. Consideration of the context surrounding families and their emotional state at this point in time demonstrates how the time at which recruitment is conducted can affect uptake. Parents described how receiving their child's diagnosis was an overwhelming experience. In the early stages post-diagnosis, parents felt bombarded by information, needed to attend multiple appointments, and some struggled to come to terms with the diagnosis:

TABLE 3 Demographic characteristics of audiologist completing interviews.

ID	Gender	Age Group (years)	Area of practice	State	Number of years peadiatric audiology experience	Number of children with bilateral mild hearing loss seen in last 12 months
A201	Female	31–40	Rehabilitation	NSW	5–9	≤5
A202	Female	41–50	Rehabilitation	NSW	15–19	≥15
A300	Female	≤30	Mixed	VIC	15–19	6–10
A307	Female	31–40	Rehabilitation	VIC	5–9	≤5
A309	Female	51–60	Diagnostic	VIC	20+	≤5
A310	Female	51–60	Diagnostic	VIC	20+	≤5
A312	Female	31–40	Rehabilitation	VIC	15–19	≤5
A400	Male	≤30	Rehabilitation	QLD	5–9	6–10
A403	Female	41–50	Rehabilitation	VIC	2±	11–15
A405	Female	≤30	Rehabilitation	VIC	<5	≤5

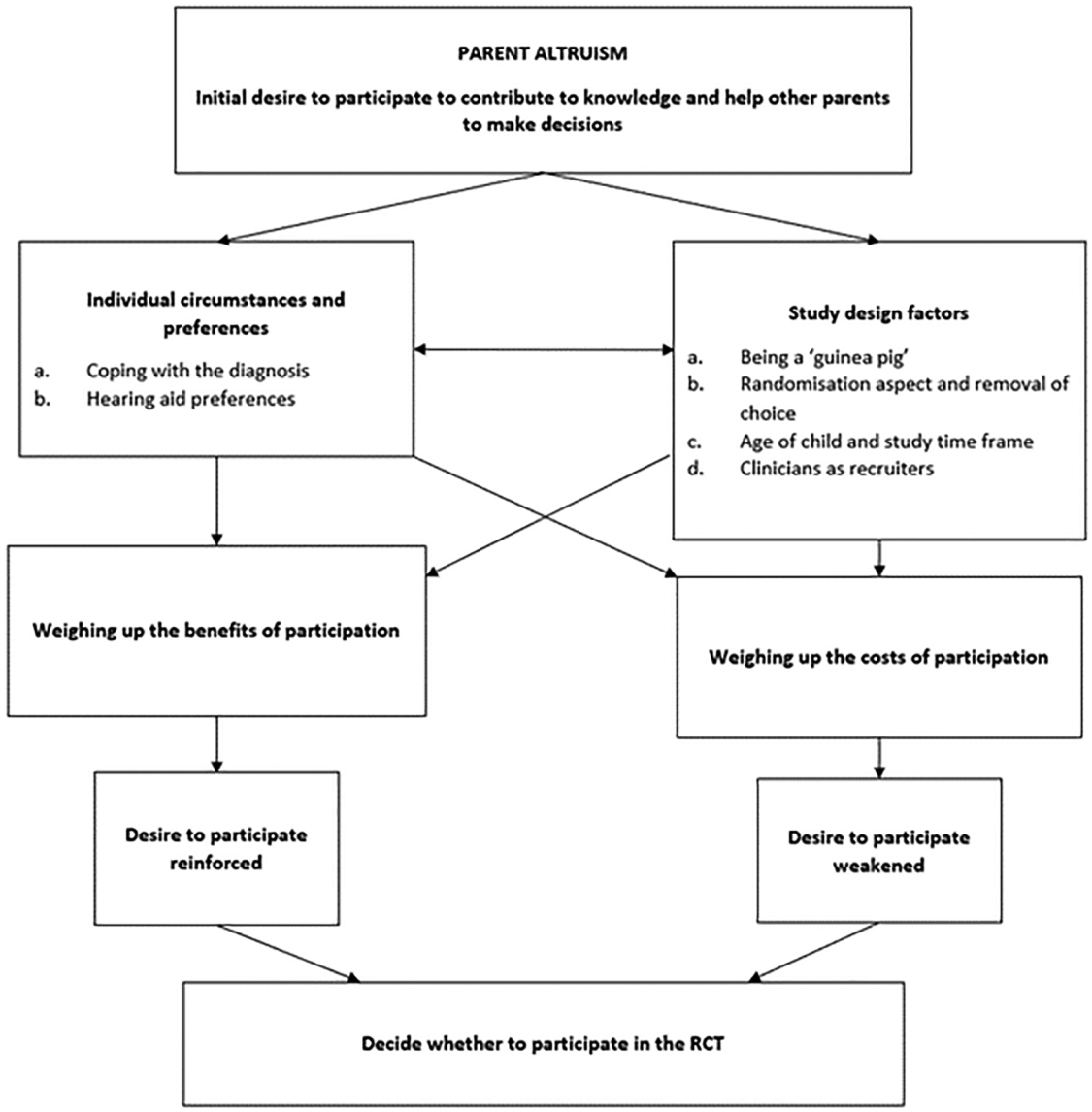

FIGURE 2
Overview of decision making (figure adapted from McCann et al. (2010) (28)).

> *As soon as the diagnostic came through, I received numerous emails with attachments that I can read through the hearing loss. But then like those things to me are—I know it's factual but, still, it doesn't really feel very real to me.* [P201]

Some parents found the invitation to participate in a study brought the emotions surrounding diagnosis and the decision about providing amplification to the forefront. The timing of study presentation is a factor that likely influenced uptake of the trial:

> *So, I guess there's so many decisions, like so many different appointments and things we go through just to get to this stage and you just had a baby. There's just so much going on. It [participating in research] feels like an extra thing without even starting to think about making that decision, and I guess like we're right now in the decision process of whether to get hearing aids or not and we're trying to get as much information as we can to make that decision. I feel like we're half agonising over the decision… Maybe in like another month I would maybe think about it [participating].* [P401]

3.2.2.2.2. Preferences for fitting or not fitting. Another factor that influenced parent willingness to partake in the trial was attitudes towards fitting of hearing aids. Preferences for fitting or not fitting varied among the interviewees; some had strong preferences either way, and some had no preference or had not yet made up their mind. One parent described how participating in the trial would make her feel anxious about her child's development:

> *I think my major thought process would be I'm someone who's very for hearing aids. So, my thought process would be—if he*

> *was chosen not to have hearing aids, would that impact his language or development in the future and things like that? I've always put it as if there was a gap or a little stone missing in your steppingstones, if I can replace that for my son I will. So, yeah, it would probably be quite anxious for me. I don't think I would actually join something like that, just in case we were picked to not have them, because I wouldn't want any negative effects on his development or hearing or language.* [P405]

Somewhat unexpected was that even when parents had strong preferences for fitting or not fitting, this did not necessarily mean they would decline participation in the trial. One parent (who had decided against hearing aids for her child) described how her personal experience of hearing loss had influenced her preferences for her daughter:

> *Well for me being hearing impaired, like it's a little bit confronting to see your daughter with hearing aids and knowing that the stigma around wearing hearing aids at times with other kids and other people that don't really understand.* [P203]

However, this parent suggested that she would not decline to participate and would not be disappointed being randomly selected for the fitted group as she understood "*that it's for the purpose of the study*" [P203]. This highlights the importance of ensuring that parents have a good understanding of the purpose and benefits of a study and its design.

3.2.2.3. Study design factors

This theme speaks to the challenges faced by researchers when attempting to strike a balance between ensuring that a study design can answer a research question using the highest possible level of evidence, whilst simultaneously maximising participation rates. The responses from parents and audiologists demonstrate that researchers should have clear reasons for the choice of study design, and this reasoning should be made available to recruiters as well as to participants. The researcher conducting the interviews noted that for parents without a background in research or experience participating in trials, the nature of a randomised control trial was understandably foreign. This theme has four subthemes, (1) being a "guinea pig", (2) randomisation aspect and removal of choice, (3) age of child and study time frame, and (4) clinicians as recruiters.

3.2.2.3.1. Being a "guinea pig". A common concern voiced by parents was that their child would be participating in something where the outcome or long-term effect was unknown:

> *I do think people usually have a tendency of being scared of trying to do things like this. Like, they feel a bit like they're guinea pigs, I would say. Like, why should I partake in an experimental study, or why should I kind of put my information, all that, out there?* [P311]

> *Thinking about it from a child's perspective, realistically they're like a guinea pig, using them—will this work, will that not work, how is this going to affect them, will it not affect them at all?* [P405]

Parents stated that the complex nature of the study design and the uncertainty could have an impact on parental uptake:

> *Clearly a lack of knowing… the lack of knowledge, like, of knowing what the study is and what the whole process is, would probably make other people around parents suggest not to, not to do it.* [P311]

When parents had a good understanding of the importance of the research, and indeed the benefits that a randomised control trial design can give, they were more supportive:

> *My husband and I, we're both scientists and we kind of know that research and studies like this are needed to find out things that are going to help people and the greater good.* [P203]

3.2.2.3.2. Randomisation aspect and removal of choice. Perhaps the most pertinent study design factor that parents and audiologists commented on was the randomisation into the fitted and non-fitted group. Hearing aid preferences varied across interviewees, and it seemed as though some parents were not comfortable having the decision regarding devices taken away. In addition, it raises a potential ethical concern with not providing amplification for these children when failure to do so could potentially negatively impact communication development:

> *You're basically signing up to get the decision taken off you. It feels a bit scary. Like you might be doing the wrong thing by your kid if you got allocated not and your child might be six months behind if they don't have them, but they actually needed them… I guess if I had to commit to a decision for six months, I wouldn't want it to be random, so I probably wouldn't take part in it* [P401].

Audiologists also noted that some parents were not comfortable with randomisation:

> *When you're doing these random assignments I know that some parents don't like that because it's taking the control away from them* [A202].

One audiologist suggested that for parents who were undecided or struggling with the decision surrounding hearing aids, the removal of choice can be positive:

> *I found it especially helpful offering a research project or trial like that, offering it to the families that were very undecided, those that were really in two minds. They had absolutely no idea which way they were going to go. This [participating in*

> *trial] meant that they didn't have to decide. It was kind of decided for them* [A400].

3.2.2.3.3. Age of child and study time frame. Another factor influencing participation in an RCT was the relatively short time frame of the study, and the age requirement. Parents generally felt comforted that their participation would be "*only six months*" and they could go back to their original decision about hearing aids or change their mind after this period [P203]. Similarly, a number of parents thought the impact of participation would be minimal due to their child's young age:

> *Because I still think at his early months—he's only not even three months—to have without the hearing aids for a period of up to six months, it probably won't hurt him that much as compared to when he is two years old, three years old or at school age.* [P201]

One parent suggested six months was a long period of time, and that the commitment could "become a bit of a burden" (401). However, the opportunity for closer monitoring of the child's development was a strong benefit to participation for this parent, and it would factor in to whether she would agree to participate:

> *I guess if you're involved your child's got more touch points and getting more monitoring that could help identify if there was an issue because I'm assuming if it was identified that he really was not hitting milestones, it'd be easier… I guess people checking in on him and seeing how he's progressing.* [P401]

As will be discussed in the following theme, the support of audiologists for the study design would be important in this context since clinicians were recruiting families. Some audiologists voiced their concern over the study design:

> *It's good in that it's not a huge length of time to commit to. I don't know how much difference six months would show given they are quite young and they are still so close to the parents and all that kind of stuff. So I don't actually know if it would be long enough to show any significant differences or not* [A201].

3.2.2.3.4. Clinicians as recruiters. Due to the relationship with the hearing service provider, audiologists working clinically were tasked with recruiting participants into the study. This could be more effective or beneficial than researchers as researchers are unfamiliar to families, and would not attain as high level of trust as clinicians for facilitating the informed consent process. However, having someone outside the research team responsible for recruitment adds further complexity to the factors that can influence parental uptake of a trial. Like parents, audiologists were generally supportive of the research because they had experienced the challenges of providing recommendations without definitive evidence:

> *I think it would give clinicians more confidence in knowing which way to advise parents about the benefits of amplification* vs. *non-amplification, or any kind of intervention, I guess because right now it's really hard. I find it very difficult to sort of know what to say to parents* [A300].

Nevertheless, audiologists have a relatively short time with families and were justifiably focused on their main responsibility of providing family-centred clinical care. As one audiologist put it: "*there's a lot to fit into that appointment time*" [A403]. Some would simply forget to recruit, "*because in the middle of a diagnosis that's the last thing on your mind*" [A300], and others spoke of recruitment potentially undermining the recommendations they were making:

> *So if I'm having difficulty convincing them to go through with various recommendations, I would probably not add 'And would you like to participate in a study?' into the mix either, and because I think it undermines the recommendation. It's like 'I'm recommending that you go and get some hearing aids, and by the way, we don't really know yet whether it's going to make a difference.' It's like, yeah, maybe not.* [A310].

Audiologists had the best interests of the family in mind, and wanted to make sure that participating in the trial would benefit the family:

> *I think either way just being involved with the study, I'd be happy with that because I know that even the children that were randomly allocated into the unaided group, they still received ongoing reviews and speech assessment at certain periods. So, they weren't necessarily just left with nothing.* [A400]

It was clear that utilising clinicians as recruiters meant that audiologists had to balance their clinical role with their recruitment responsibility. Clinicians have the benefit of knowing how the family is coping with the diagnosis and other priorities in their life, and some stated they would select which families they would attempt to recruit based on how they were "managing the news and the diagnosis" [A310]. Most, however, were of the view that they generally "wouldn't deny anyone the knowledge of the research" [A309]. Unlike a member of the research team, clinicians may not always prioritise recruitment, particularly if they had less understanding of, or experience with, research:

> *I think that because I have worked in research before I have an appreciation of the benefits of research as well as the challenges of recruitment. So I feel like most other audiologists wouldn't probably give as much energy to this sort of thing as I would. I would probably be more pro supporting research than the general audiologist, and I would find the time, but I don't think that a lot of other audiologists would* [A403].

Discussion

Our paper highlights the importance of community engagement in designing and conducting research to determine whether early amplification benefits infants and young children

with bilateral mild hearing loss. Important lessons have been learnt from the failure of recruitment for our proof-of-concept RCT. Our subsequent qualitative study explored the barriers and enablers of participation in a RCT, and identified useful concepts that could be applied to future research studies that attempt to address the research question.

Over one year from 1st February 2019 to 31st January 2020, 40 infants were referred to the RCT, which was much fewer than expected. During this period, according to data from hearing screening programs and diagnostic audiology services, approximately 146 children were diagnosed with bilateral mild hearing loss in the three states; therefore, only approximately 27% (40/146) were referred to the study team. There were a few possible reasons for why 106 infants were not referred to the study. First, and anecdotally the most common reason, diagnostic and rehabilitation audiologists indicated that they did not refer families to the study if they did not meet the audiology threshold criteria (e.g., infants originally diagnosed with mild bilateral hearing loss may subsequently have normal hearing or moderate hearing loss). Second, a database error in Victoria accounted for 9 potentially eligible families missed from being referred. Third, audiologists indicated that they did not refer families if families had insufficient English to give consent.

Of the potentially eligible participants referred, the majority were ineligible (26/40, 65%). The main reason was infants not meeting the hearing threshold criteria. Our inclusion criteria depended upon the infant meeting particular thresholds of hearing loss over three or four frequencies in two ears. Infants may be diagnosed with bilateral mild loss, but even if a single hearing threshold in one (or both) ears changed on subsequent testing, they may have been assigned a different degree of loss. These infants would then have become ineligible for the study. This reflects the fact that diagnosis of mild hearing loss can be uncertain and challenging, and often requires multiple diagnostic audiology appointments to confirm the hearing status (16). Conversely, infants initially diagnosed with a different degree of loss may subsequently become eligible, but we may have missed the window to recruit them.

The other main reason for ineligibility was the infant having already been fitted with hearing aids (4/26, 19%), or the family having already made the decision to fit or not fit hearing aids (6/26, 23%), at the time when they were approached by the researcher. Nine out of 11 eligible families declined to take part (9/11, 82%). Parents did not want their child's treatment decision to be randomised, or did not want to be involved in research. The high ratio of families who declined compared to those who participated indicates that there may have been considerable barriers to participation, and could possibly reflect parents' preference to make their own choice in hearing device fitting. There are many possible reasons for this: parents may feel empowered to take action for their child's hearing loss by fitting hearing aids; parents may perceive benefits to hearing device fitting; and parents may feel potential guilt of denying the child the opportunity to access a full range of sounds, especially if the child has subsequent language delays (16). These factors were evident from our subsequent qualitative study (see below).

There may also be other reasons why we received a lower than expected number of referrals for the study. Some of the families may not have been referred because they did not speak sufficient English to provide consent. In Victoria, around 24% of families of children with congenital hearing loss are culturally and linguistically diverse; the exact proportion of families who do not speak sufficient English is not known (Z. Poulakis, Victorian Infant Hearing Screening Program, personal communication, 29th Nov 2021). It is also possible some children with complex medical needs were not referred. In an audit of a Victorian clinical service for children with hearing loss, nine out of 129 (7%) of children with mild hearing loss had complex medical needs (29). As this was a clinical service for children with medical needs, we would expect this proportion to be lower at a population level.

Our use of threshold averages for determining hearing loss degree was consistent with other studies that included children with mild losses (30). The design of our trial where we excluded children who were already fitted or had already decided about fitting hearing devices meant that most referrals for recruitment were for young infants, where it was necessary to rely on objective evoked potential threshold estimates. Our strict exclusion of children who had hearing thresholds outside the desired range at any frequencies aimed to maximise the rigor of the RCT. However, a more pragmatic approach to accommodate potential uncertainties around diagnostic thresholds, particularly for evoked potential threshold estimates, may have allowed for more referrals for consideration for recruitment in a real-life setting. Since the completion of our RCT, others have demonstrated the utility of unaided audibility to identify those children who, without amplification, may be at risk of language delays (31).

Two families participated in the RCT. Both families completed baseline and follow up data collection. The family randomised to the intervention group completed five out of six of the compliance questionnaires. Even though there was only one participant in the intervention group, monitoring device use by a short monthly parent report in REDCap may be an acceptable compliance monitoring method. The two participant families indicated that the experience of taking part was either positive or neutral, and completed the study protocols without issue. Although it was not possible to draw conclusions from two participants, there was no indication that the families found participating difficult or onerous. The low number of participants meant we could not address our primary aims.

To further understand the barriers and enablers to participation in an RCT, we subsequently conducted a qualitative study to understand perceptions of participation, by interviewing parents of children <2 years old with newly diagnosed bilateral mild hearing loss, and audiologists. We demonstrated the overarching facilitator to participation of parental and audiologists' desire to contribute to research to help determine whether hearing devices should be offered to newborns with mild hearing loss. This was in the setting of most parents and audiologists experiencing the stresses and challenges of uncertainty in the early management of mild hearing loss,

congruent to previous research (18, 23). Individual circumstances, including how the family was coping with the new diagnosis, and their hearing aid preferences, could strongly influence their preferences for participation. Past studies on paediatric cancer trials have indicated that when consent for a child's participation in a trial is sought from parents soon after diagnosis, parents are likely to make decisions when they are distressed and vulnerable (32).

The strongest barrier, and perhaps the least modifiable factor, to participating in an RCT of hearing device fitting, was parents' reluctance for their child to be randomised to a treatment group, due to parental perception of losing control over choice of hearing device fitting. This was also against audiologists' values of family-centred practice. The perception of guilt of potentially causing harm to their child by not fitting hearing devices early was a notable barrier to participation. This has been shown in previous research, where anticipation of possible regret often accompanies a parent's sense of responsibility to protect their child in their decision-making while considering participating in trials (33). These may be challenges that cannot be easily overcome. In addition, there may be potential ethical concerns with not providing amplification for these children when failure to do so could potentially negatively impact communication development.

Nevertheless, we have learnt there are some potentially modifiable factors to improve uptake in research studies involving families of infants and young children with mild hearing loss. Parental perceptions of their child being a "guinea pig" in research could be addressed by increasing general awareness of research and better or clearer information about the study methodology and what was involved. Involving parents as study recruiters could also be a way to breakdown misconceptions and improve uptake rate. Potentially modifiable factors to study design to improve study uptake could include relaxation of eligibility criteria and increasing awareness amongst other child hearing health stakeholders (e.g., early intervention services, maternal child health nurses) of the study. The benefits of utilising clinicians as recruiters are many; however, it is important that researchers consider the burden they may be placing on busy professionals. In particular, we note that no ENT specialist or paediatrician made a referral for study recruitment. Many RCTs rely on clinicians as recruiters, with up to 50% failing to recruit target numbers (34). A 2013 systematic review identified 11 qualitative studies that centre around 8 themes relating to clinician's involvement and recruiting to RCTs; these would be of salience for any future trials in this population (34). A strong relationship and open communication between clinicians and the research team, and remunerating clinicians as recruiters (including protected time added to appointments to discuss the study) are paramount. Identifying clinicians with a passion for the study (such as participant A403), and further supporting their role in recruitment may also be a strategy that could improve parental uptake of the trial. It is possible that stronger engagement with diagnostic audiologists may have improved referral rates. In the state of VIC, we bypassed the need for diagnostic audiologists to refer to the study team by identifying potentially eligible participants through the state's newborn hearing screening program; extending this method to the other states may have helped. Nevertheless, even if referrals to the study were increased, the 82% parental decline rate means that unless we could address the reasons for parental decline, we would unlikely have been successful with recruiting enough participants.

Our study's greatest limitation was the inability to recruit sufficient participants for the RCT. Its strength was to use qualitative methodology to identify factors influencing participation in a RCT on hearing device fitting in infants with mild bilateral hearing loss, and in so, engaging the community in future study design. This research question is not answerable through a RCT design as the removal of parental choice through randomisation may not align with family centred practice. Therefore, alternative methodologies must be considered. These may involve novel methods of measuring infants' ability to hear [e.g., objective assessments which measure speech discrimination in infants (35, 36)], and analysing outcomes data of aided and unaided infants with mild hearing loss from large observational studies, such as the proposed National Health and Medical Research Council (NHMRC) funded Australian National Child Hearing Health Outcomes Registry (ANCHOR), which aims to link data from child hearing health services in Australia to track child hearing outcomes.[1]

In conclusion, our attempted trial highlighted many barriers and challenges around trial recruitment involving randomisation for families at a very vulnerable and stressful time of their children's lives, shortly after their hearing loss diagnosis. Community engagement is paramount in designing and conducting research to determine whether early amplification benefits infants and young children with bilateral mild hearing loss. Important lessons have been learnt from the failure of recruitment for our proof-of-concept RCT. Better engagement of audiologists as recruiters, and additional supports for parents, may be necessary to improve recruitment rate in designing future studies. However, the RCT methodology takes away caregiver choice and control, may not align with family centred practice and may present a potential ethical concern for future adverse consequences if early amplification is not offered. Alternative research methodological approaches without randomisation are ultimately required to answer the important question of whether early hearing amplification benefits infants with mild bilateral hearing loss.

Data availability statement

The datasets presented in this article are not readily available because Data collected in this study is subject to the National Acousitc Laboratory's Privacy Policy, which can be found here: http://www.hearing.com.au/privacy-policy/. Participant information collected remains strictly confidential. Only the researchers directly

[1]https://www.mcri.edu.au/research/projects/anchor

involved with the study can access the information. We have undertaken to disclose the information only with expressed written permission from the participants, except as required by law. Requests to access the datasets should be directed to valerie.sung@rch.org.au.

Ethics statement

The studies involving human participants were reviewed and approved by Royal Children's Hospital Human Research and Ethics Committee HREC (38112 (HREC/45275/RCHM-2018-151266)). Written informed consent to participate in this study was provided by the participants' legal guardian/next of kin.

Author contributions

VS had oversight of the whole study. VS, TC and RB acquired funding. VS, TC, PC, AK and CI conceptualised and designed the study. VM and LS organised the database. VM, LS, AK, PC and MS collected the data. VS, TC, RB, VM, LS and MS performed the data analyses. VS, TC, RB, PC, CI provided supervision. VS wrote the original draft. VM and MS wrote sections of the manuscript. All authors contributed to the article and approved the submitted version.

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Conflict of interest

AK was Principal Audiologist for Paediatric Services at Hearing Australia at the time of the study. Hearing Australia, funded by the Australian Government, is the National provider of hearing services and devices to children and young people up to 26 years of age in Australia. Hearing Australia did not receive any funding for this study. TC and VM were employed by the National Acoustic Laboratories, the research division of Hearing Australia. RB was employed by Children's Health Queensland Hospital and Health Service. All other authors declare no conflict of interest. The funders had no role in the design of the study; in the collection, analyses, or interpretation of data; in the writing of the manuscript, or in the decision to publish the results.

Publisher's note

Supplementary material

The Supplementary Material for this article can be found online at: https://www.frontiersin.org/articles/10.3389/fped.2023.1197739/full#supplementary-material

References

1. Smith RJ, Bale JF Jr., White KR. Sensorineural hearing loss in children. *Lancet*. (2005) 365(9462):879–90. doi: 10.1016/S0140-6736(05)71047-3

2. Wake M, Ching TY, Wirth K, Poulakis Z, Mensah FK, Gold L, et al. Population outcomes of three approaches to detection of congenital hearing loss. *Pediatrics*. (2016) 137(1):1–10. doi: 10.1542/peds.2015-1722

3. National Workshop on Mild and Unilateral Hearing Loss: Workshop Proceedings. Breckenridge (CO): Centers for Disease Control and Prevention. (2005).

4. Moeller MP. Early intervention and language development in children who are deaf and hard of hearing. *Pediatrics*. (2000) 106(3):E43. doi: 10.1542/peds.106.3.e43

5. Ching TYC, Dillon H, Button L, Seeto M, Van Buynder P, Marnane V, et al. Age at intervention for permanent hearing loss and 5-year language outcomes. *Pediatrics*. (2017) 140(3):e20164274. doi: 10.1542/peds.2016-4274

6. Yoshinaga-Itano C, Sedey AL, Coulter DK, Mehl AL. Language of early- and later-identified children with hearing loss. *Pediatrics*. (1998) 102(5):1161–71. doi: 10.1542/peds.102.5.1161

7. Ching TY, Dillon H, Marnane V, Hou S, Day J, Seeto M, et al. Outcomes of early- and late-identified children at 3 years of age: findings from a prospective population-based study. *Ear Hear*. (2013) 34(5):535–52. doi: 10.1097/AUD.0b013e3182857718

8. Winiger AM, Alexander JM, Diefendorf AO. Minimal hearing loss: from a failure-based approach to evidence-based practice. *Am J Audiol*. (2016) 25(3):232–45. doi: 10.1044/2016_AJA-15-0060

9. Tharpe AM. Unilateral and mild bilateral hearing loss in children: past and current perspectives. *Trends Amplif*. (2008) 12(1):7–15. doi: 10.1177/1084713807304668

10. Bess FH, Dodd-Murphy J, Parker RA. Children with minimal sensorineural hearing loss: prevalence, educational performance, and functional status. *Ear Hear*. (1998) 19(5):339–54. doi: 10.1097/00003446-199810000-00001

11. Wake M, Tobin S, Cone-Wesson B, Dahl HH, Gillam L, McCormick L, et al. Slight/mild sensorineural hearing loss in children. *Pediatrics*. (2006) 118(5):1842–51. doi: 10.1542/peds.2005-3168

12. Porter H, Sladen DP, Ampah SB, Rothpletz A, Bess FH. Developmental outcomes in early school-age children with minimal hearing loss. *Am J Audiol*. (2013) 22(2):263–70. doi: 10.1044/1059-0889(2013/13-0013)

13. Walker EA, Holte L, McCreery RW, Spratford M, Page T, Moeller MP. The influence of hearing aid use on outcomes of children with mild hearing loss. *J Speech Lang Hear Res*. (2015) 58(5):1611–25. doi: 10.1044/2015_JSLHR-H-15-0043

14. Walker EA, Sapp C, Dallapiazza M, Spratford M, McCreery RW, Oleson JJ. Language and reading outcomes in fourth-grade children with mild hearing loss compared to age-matched hearing peers. *Lang Speech Hear Serv Sch*. (2020) 51 (1):17–28. doi: 10.1044/2019_lshss-ochl-19-0015

15. Tomblin JB, Oleson J, Ambrose SE, Walker EA, McCreery RW, Moeller MP. Aided hearing moderates the academic outcomes of children with mild to severe hearing loss. *Ear Hear*. (2020) 41(4):775–89. doi: 10.1097/aud.0000000000000823

16. Nassrallah F, Tang K, Whittingham J, Sun H, Fitzpatrick EM. Auditory, social, and behavioral skills of children with unilateral/mild hearing loss. *J Deaf Stud Deaf Educ*. (2020) 25(2):167–77. doi: 10.1093/deafed/enz041

17. Fitzpatrick EM, Durieux-Smith A, Gaboury I, Coyle D, Whittingham J. Communication development in early-identified children with mild bilateral and unilateral hearing loss. *Am J Audiol*. (2015) 24(3):349–53. doi: 10.1044/2015_AJA-15-0003

18. Lin JJ, Gillam L, Smith L, Carew P, King A, Ching TYC, et al. Mild matters: parental insights into the conundrums of managing mild congenital hearing loss. *Int J Audiol*. (2022) 61(6):500–6. doi: 10.1080/14992027.2021.1954248

19. Fitzpatrick E, Grandpierre V, Durieux-Smith A, Gaboury I, Coyle D, Na E, et al. Children with mild bilateral and unilateral hearing loss: parents' reflections on experiences and outcomes. *J Deaf Stud Deaf Educ*. (2016) 21(1):34–43. doi: 10.1093/deafed/env047

20. Gelfand SA, Silman S. Apparent auditory deprivation in children: implications of monaural versus binaural amplification. *J Am Acad Audiol*. (1993) 4(5):313–8. PMID: 8219298

21. King AM. The national protocol for paediatric amplification in Australia. *Int J Audiol*. (2010) 49(Suppl 1):S64–9. doi: 10.3109/14992020903329422

22. Aided Child Demographics. Hearing Australia. (2020).

23. Ching TYC, Saetre-Turner M, Marnane V, Scarinci N, Choik C, Tulloch K, et al. Audiologists' perspectives on management of mild bilateral hearing loss in infants and young children. *Int J Audiol*. (2022) 61(9):752–60. doi: 10.1080/14992027.2021.1961170

24. Watkin PM, Baldwin M. Confirmation of deafness in infancy. *Arch Dis Child*. (1999) 81(5):380–9. doi: 10.1136/adc.81.5.380

25. Bolam B, Gleeson K, Murphy S. "Lay person" or "health expert"? exploring theoretical and practical aspects of reflexivity in qualitative health research. *Forum Qual Sozialforschung*. (2003) 4(2):26. doi: 10.17169/fqs-4.2.699

26. Tie Y C, Birks M, Francis K. Grounded theory research: a design framework for novice researchers. *SAGE Open Med*. (2019) 7:2050312118822927. doi: 10.1177/2050312118822927

27. Suddaby R. From the editors: what grounded theory is not. *Acad Manage J*. (2006) 49(4):633–42. doi: 10.5465/amj.2006.22083020

28. McCann SK, Campbell MK, Entwistle VA. Reasons for participating in randomised controlled trials: conditional altruism and considerations for self. *Trials*. (2010) 11(1):31. doi: 10.1186/1745-6215-11-31

29. Olivier N, Shepherd D, Smith L, Carew P, Paxton G, Downie L, et al. Etiology, comorbidities, and health service use in a clinical cohort of children with hearing loss. *Ear Hear*. (2022) 43(6):1836–44. doi: 10.1097/AUD.0000000000001253

30. Tomblin JB, Walker EA, McCreery RW, Arenas RM, Harrison M, Moeller MP. Outcomes of children with hearing loss: data collection and methods. *Ear Hear*. (2015) 36 Suppl 1(0 1):14S–23S. doi: 10.1097/AUD.0000000000000212

31. McCreery RW, Walker EA, Stiles DJ, Spratford M, Oleson JJ, Lewis DE. Audibility-Based hearing aid fitting criteria for children with mild bilateral hearing loss. *Lang Speech Hear Serv Sch*. (2020) 51(1):55–67. doi: 10.1044/2019_LSHSS-OCHL-19-0021

32. Kodish ED, Pentz RD, Noll RB, Ruccione K, Buckley J, Lange BJ. Informed consent in the childrens cancer group: results of preliminary research. *Cancer*. (1998) 82(12):2467–81. doi: 10.1002/(sici)1097-0142(19980615)82:12<2467::aid-cncr22>3.0.co;2-m

33. Shilling V, Young B. How do parents experience being asked to enter a child in a randomised controlled trial? *BMC Med Ethics*. (2009) 10(1):1. doi: 10.1186/1472-6939-10-1

34. Fletcher B, Gheorghe A, Moore D, Wilson S, Damery S. Improving the recruitment activity of clinicians in randomised controlled trials: a systematic review. *BMJ open*. (2012) 2(1):e000496. doi: 10.1136/bmjopen-2011-000496

35. Ching TYC, Zhang VW, Ibrahim R, Bardy F, Rance G, Van Dun B, et al. Acoustic change complex for assessing speech discrimination in normal-hearing and hearing-impaired infants. *Clin Neurophysiol*. (2023) 149:121–32. doi: 10.1016/j.clinph.2023.02.172

36. Mao D, Wunderlich J, Savkovic B, Jeffreys E, Nicholls N, Lee OW, et al. Speech token detection and discrimination in individual infants using functional near-infrared spectroscopy. *Sci Rep*. (2021) 11(1):24006. doi: 10.1038/s41598-021-03595-z

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EDITED BY
Carmen Kung,
Macquarie University, Australia

REVIEWED BY
Dawna Lewis,
Boys Town National Research Hospital, United States
Nicole Corbin,
University of Pittsburgh, United States

*CORRESPONDENCE
Hanna Josefsson Dahlgren
✉ hanna.josefsson@ki.se

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A pilot study on spatial hearing in children with congenital unilateral aural atresia

Hanna Josefsson Dahlgren[1,2]*, Cecilia Engmér Berglin[1,2], Malou Hultcrantz[2] and Filip Asp[1,2]

[1]Department of Clinical Science, Intervention and Technology, Karolinska Institutet, Stockholm, Sweden, [2]Department of Otorhinolaryngology, Karolinska University Hospital, Stockholm, Sweden

Despite normal hearing in one ear, individuals with congenital unilateral aural atresia may perceive difficulties in everyday listening conditions typically containing multiple sound sources. While previous work shows that intervention with bone conduction devices may aid spatial hearing for some children, testing conditions are often arranged to maximize any benefit and are not very similar to daily life. The benefit from amplification on spatial tasks has been found to vary between individuals, for reasons not entirely clear. This study has sought to expand on the limited knowledge on how children with unilateral aural atresia recognize speech masked by competing speech, and how horizontal sound localization accuracy is affected by the degree of unilateral hearing loss and by amplification using unilateral bone conduction devices when fitted before 3 years of age. In a within-subject, repeated measures design, including 11 children (mean age = 7.9 years), bone conduction hearing device (BCD) amplification did not negatively affect horizontal sound localization accuracy. The effect on speech recognition scores showed greater inter-individual variability. No benefit from amplification on a group level was found. There was no association between age at fitting and the benefit of the BCD. For children with poor unaided sound localization accuracy, there was a greater BCD benefit. Unaided localization accuracy increased as a function of decreasing hearing thresholds in the atretic ear. While it is possible that low sound levels in the atretic ear provided access to interaural localization cues for the children with the lowest hearing thresholds, the association has to be further investigated in a larger sample of children.

KEYWORDS

unilateral aural atresia, unilateral conductive hearing loss, UCHL, sound localization, speech recognition, BCD, bone conduction device, early fitting

1. Introduction

Individuals with unilateral conductive hearing loss due to unilateral aural atresia (UAA) report a high degree of difficulties in tasks related to binaural hearing, such as sound localization and recognition of speech in noise (1). Treatment using bone conduction hearing devices (BCDs) aim to restore hearing in the atretic ear and aid in binaural hearing. From infancy, children might be offered a passive transcutaneous BCD fitted on a softband. The standard percutaneous skin-penetrating BCD attached to a titanium screw osseointegrated in the cortical bone superior and posterior to the pinna provide the user with higher amplification compared to the softband (2). It has been the first hand choice for treating hearing loss in UAA at our clinic for several years as surgery is minimally invasive and serious adverse events are rare. However, in a study from 2015,

authors found that 47% of children implanted with a percutaneous BCD had discontinued using the implant 5 years after surgery (3). Insufficient benefit from amplification was one of the most commonly stated reasons for non-usage (3). Pure-tone thresholds improve from amplification (4, 5) but the effect on binaural hearing needs to be investigated further. There seem to be a large inter-individual variability on the effect from amplification on speech recognition thresholds (SRTs) (6, 7) as well as on sound localization accuracy (SLA) (6, 8). It is known that some individuals localize fairly good in the monaural unaided setting and thus will not benefit as much from BCD (6, 8, 9). A possible explanation for the high inter-individual variability in benefit from a BCD is that some individuals with congenital UAA learn to use monaural spectral cues for localization in the horizontal plane (10). The age at which the child is fitted with the BCD has also been suggested as a factor influencing the benefit from amplification (11, 12). In children with unilateral sensorineural hearing loss, sound localization accuracy seems to improve for children fitted with a hearing aid by 5 years of age, whereas not for children fitted by 9 years of age (13). In most studies regarding children implanted with a BCD, the study participants have started using their BCD at 4–6 years of age at the earliest (9). Studies presenting results from surgically implanted percutaneous or transcutaneous devices rarely disclose whether the participants in the studies have previously been using a different system for bone conduction and for how long. As binaural hearing and the central auditory pathways develop during the first 5–6 years of life (14–16), early treatment might be beneficial on binaural tasks such as horizontal sound localization and speech recognition in acoustically challenging conditions (11, 12).

2. Aim

The aim of the present study was to quantify the effect of early access to unilateral bone conduction amplification on sound localization accuracy and recognition of speech in symmetrically separate competing speech in a cohort of children with UAA fitted with a BCD before 3 years of age.

3. Materials and methods

3.1. Study participants

Children with congenital UAA were recruited from a list of patients that had attended the atresia clinic at the Hearing Habilitation Unit at Rosenlund's Hospital from 2015 to 2017. Forty-one individuals were eligible based on the following inclusion criteria: 5–10 years of age, unilateral congenital atresia, fitted with a BCD, and fluent in the Swedish language. Ten subjects were excluded based on the following exclusion criteria: syndrome-associated atresia ($n = 4$); sensorineural hearing loss ($n = 1$); contralateral air conduction pure-tone average across 500, 1,000, 2,000, and 4,000 Hz (PTA_4) >20 dB hearing level (HL) ($n = 4$); and surgical ear canal repair ($n = 1$). Investigations took place during January 2018, November 2020, and spring 2021. Four individuals that were not able to come in for testing in 2018 had grown too old for inclusion in 2020. One subject had moved abroad and could not be contacted. Thirteen individuals declined or could not attend any of the visits for different reasons. One subject repeatedly did not show up for measurements. One subject did not fulfill any aided measurements due to lack of time and was excluded from analysis (**Figure 1**).

3.2. Study design

In a 3 h visit, recognition of speech in spatially separate competing speech and horizontal sound localization accuracy were tested in a within-subject repeated measures design (aided and unaided, test order was pseudo-randomized based on the last digit of the subject's national ID number). Aided and unaided pure-tone hearing thresholds were also measured. The children used their own BCD, either a Cochlear or an Oticon processor (**Table 1**), for all aided measurements. The devices had been previously programmed using the fitting software provided by the manufacturer and had been fine-tuned according to the preferences of the child. Background data were retrieved from the caregiver of the study participants and from patient charts. Data on mean usage per day were retrieved from the device using the fitting software. The study participants had been provided with a copy of Parents Evaluation of Aural/Oral Performance of Children (PEACH) to fill out before the visit. Ethical approval was obtained from the regional ethics committee in Stockholm, 2012/1661-31/3. Written consent was acquired from all study participants.

3.3. Unaided and aided hearing thresholds

Unaided air- and bone conduction hearing thresholds were measured according to ISO 8253-1 (2010) using TDH39 supra-aural headphones and the Radioear B71 bone transducer. Masking of the non-test ear was applied as appropriate. To estimate the degree of amplification provided by the BCD, aided hearing thresholds were quantified by measuring frequency-modulated tone thresholds in sound field using a fixed-frequency Békèsy technique. While the reliability of this technique is not quantified in children, it is characterized by high reliability and reproducibility in adults (17, 18). During the measurements, the contralateral normal ear was plugged by an earplug (EAR Classic foam earplug; 3M, Minneapolis, MN, USA) and a circum-aural hearing protector (Bilsom 847 NST II, Honeywell Safety Products, RI, USA) was placed over the ear plug. The combination of the plug and the circum-aural hearing protector was previously estimated to provide an average of 39 dB attenuation of the PTA_4, based on recordings in adults ($n = 8$) with normal hearing according to ISO 4869-1 (1990) (19, 20). Mean (SD) attenuation values as recorded in those adults ($n = 8$)

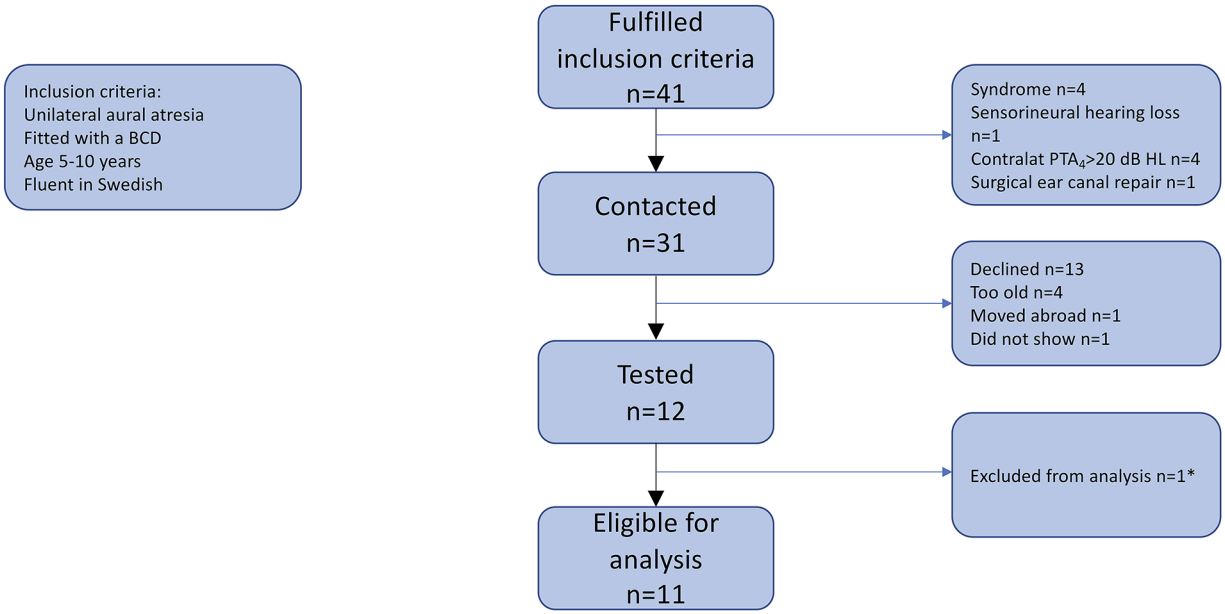

FIGURE 1
Flow chart for inclusion in the study. Thirteen eligible participants declined participation due to different reasons. Four patients were between 5 and 10 years old in 2018 but could not make it to any of the appointments available at that time and had grown too old to include in the study when more opportunities opened in 2021. *One participant did not perform any aided measurements due to lack of time and was therefore excluded from analysis.

were 34.7 dB (6.3 dB) at 0.5 kHz, 35.1 dB (6.0 dB) at 1 kHz, 40.5 dB (2.9 dB) at 2 kHz, 47.2 dB (5.6 dB) at 3 kHz, 49.4 dB (4.4 dB) at 4 kHz, and 46.2 dB (6.4 dB) at 6 kHz.

3.4. Horizontal sound localization ability

An eye-tracking technique was used to determine the perceived sound location. The setup, stimuli, and quantification of sound localization responses have been previously described in detail (21). The rationale for using this test was that it allows for rapid determination of horizontal localization accuracy (approximately 3 min recording time) and has previously been used for measuring sound localization accuracy in children with unilateral hearing loss (22) as well as in measuring the difference in performance between bilateral and unilateral sound stimulation (23).

3.4.1. Setup

Measurements were conducted in a double-walled sound booth [ambient sound level = 25 dBA, reverberation time T_{30} = 0.11 s at 500 Hz, as recorded with a B & K 2238 Mediator and a B & K

TABLE 1 Background data including type of device, age, gender, and degree of usage from the computer log of the BCD.

Subject ID	Device	Processor	Age at fitting (year)	Age at testing (year)	Duration of device use (year)	Usage (h/day)	Gender	Atretic side
1	PC	Baha 4	1.75	7.1	5.7	7.5	M	R
2	PC	Baha 5	2.33	10.1	7.8	5.2	M	R
3	PC	Ponto Pro	2.83	8.1	5.3	—[a]	M	R
4	PC	Baha 5	2.83	8.5	4.8	7.5	M	R
5	Softband	Ponto Pro	0.5	5.3	4.9	6.6	F	L
6	PC	Baha 5	0.42	5.5	5.2	5.7	M	R
7	Softband	Baha 5	1.25	8.5	7.2	5.3	M	R
8	BAHA Attract	Baha 5	0.17	9.9	7.2	0.3	M	R
9	Softband	Baha 5 SP	0.25	5.11	5.6	1.1	F	R
10	PC	Baha 5	2.0	7.8	5.5	4.2	M	R
11	PC	Baha 5	2.33	10.10	8.45	—[a]	M	R
Mean ± SD			1.5 ± 1.0	7.9 ± 1.9	6.3 ± 1.3	4.8 ± 2.6		

PC, percutaneous fixture.
All study participants were initially fitted with a BCD on softband. The time of surgery for a percutaneous device or BAHA attract was unavailable to the authors.
[a]System for reading the computer log of the device not functioning.

2260 Investigator (Brüel & Kjær), respectively]. Twelve active loudspeakers each coupled to a 7-inch video display (LD pairs) were placed equidistantly in a 110° arc in the frontal horizontal plane, resulting in loudspeaker positions at ±55°, ±45°, ±35°, ±25°, ±15°, and ±5° relative to the subject who was seated facing the loudspeaker array. The distance from the LD pairs to the head of the study participant was approximately 1.2 (loudspeaker) and 1.1 m (screen). The LD pairs were vertically adjusted to the height of the study participant using a motorized stand, situating the loudspeakers at ear level (**Figure 2**).

To record the gaze of the study participants in relation to the LD pairs, an eye-tracking system was used (Smart Eye Pro, Smart Eye AB, Gothenburg, Sweden). The coordinates of the LD pairs were defined in three dimensions in the eye-tracking system, resulting in areas of interest [AOIs (21, 24)]. Twelve AOIs (width = 0.17 m; height 0.55 m) constituted a continuous array of AOIs in a 3D model, corresponding to the physical LD pairs.

3.4.2. Stimulus

The visual stimulus was a colorful children's cartoon. The auditory part of the stimulus consisted of a broadband musical melody with a long-term frequency spectrum similar to that of a female voice and naturally occurring amplitude modulations. The stimulus was presented at 63 dB sound pressure level (SPL) (A). The rationale for using this stimulus was that it allows comparison with previous findings in children and adults with normal hearing (21), with children with congenital unilateral sensorineural hearing losses (22), and with adults with congenital unilateral atresia (25).

3.4.3. Test procedure and quantification of localization responses

Study participants were familiarized with the auditory–visual stimulus during a gaze-calibration procedure in which the stimulus was presented from different azimuths. The test started by presenting the stimulus from the LD pair at −5°. After approximately 7 s, the visual stimulus was stopped, and the sound immediately shifted to a randomized loudspeaker. After 1.6 s, the visual stimulus was reintroduced at the azimuth of the sounding loudspeaker. Azimuthal shifts were repeated 24 times following a beforehand generated order of randomized shifts.

Children were allowed to move their head freely. They were instructed to look where they perceived the sound was coming from and informed that they would be guided by audition only during sound-only presentation and that the visual part of the stimulus would reappear at the same azimuth as the sound.

The position of the study participant's pupil relative to the LD pairs was sampled at 20 Hz during the 1.6 s sound-only periods. The median pupil position from the last 500 ms of the sound-only period was defined as the perceived sound source location. SLA was quantified as an error index (EI) [for calculations, see Asp et al. (21)] ranging from 0 to 1, where 0 represents a perfect performance and 1 a random performance. Based on test–retest analyses in infants and young children, also from Asp et al. (21), a within-subject difference in the EI of ±0.12 was considered statistically significant at the 95% confidence level.

3.5. Speech recognition thresholds in competing speech

Measurements of SRT were performed in a setup resembling a challenging everyday listening situation (19, 22, 26) using a matrix test (22, 26). Participants were seated in the middle of a double-walled sound booth facing a loudspeaker presenting target speech

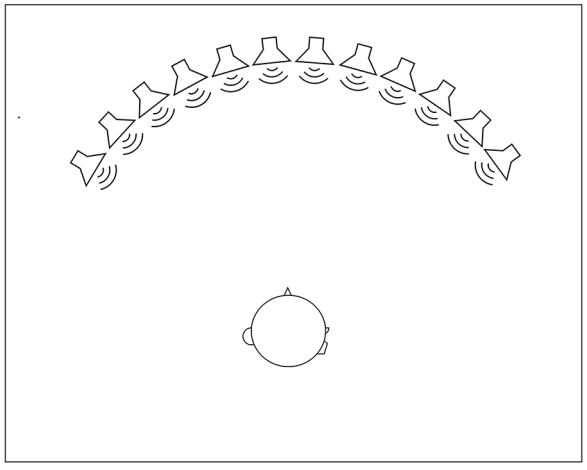

FIGURE 2
Setup for testing of SLA. Twelve loudspeakers each coupled to a 7-inch video display (LD pairs) were placed in a 110° arc in the frontal horizontal plane relative to the subject who was seated facing the loudspeaker array. The distance from the LD pairs to the head of the study participant was approximately 1.2 (loudspeaker) and 1.1 m (screen). The LD pairs were adjusted to be at ear level of the study participant.

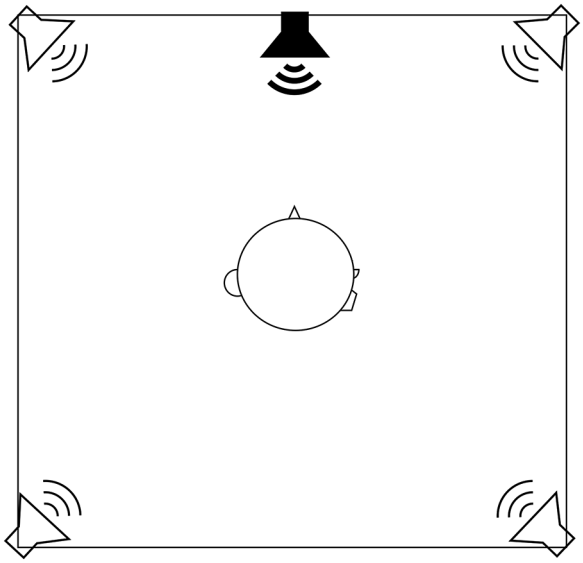

FIGURE 3
Setup for testing of SRT. Participants were seated facing a loudspeaker presenting target speech at 0° azimuth. Interfering speech was presented from four spatially symmetrically separated loudspeakers at ±30° and ±150° azimuth at a fixed overall level of 63 dB SPL measured at the position of the subject's head.

at 0° azimuth. Interfering speech was presented from four spatially and symmetrically separated loudspeakers at ±30° and ±150° azimuth at a fixed overall level of 63 dB SPL measured at the position of the subject's head (**Figure 3**). The interferers comprised four non-correlated recordings of a single male talker reading a novel. The target speech (the Hagerman sentences) was a female voice (27). Each sentence consisted of five words that formed a grammatically correct sentence with low semantic predictability in a fixed syntax (e.g., "Peter höll nio nya lådor," in translation: "Peter held nine new boxes").

Study participants were instructed to face the loudspeaker presenting target speech and asked to repeat the sentences from three lists (one training list), each containing 10 sentences. This resulted in the presentation of 30 sentences per listening condition (aided and unaided). No sentence was repeated. Oral responses were recorded and scored by an audiologist outside the test room. Both the aided and unaided assessments started by presenting the first sentence of the training list at a signal-to-noise ratio (SNR) of +10 dB. For the following training sentences, the target speech level decreased up to three times in 5 dB steps, then up to three times in 3 dB steps, and then in 2 dB steps until the number of correct words in a sentence was ≤2. When the number of correct words in a sentence was ≤2, or the training list ended, the training was terminated. Subsequently, two lists (i.e., 20 sentences) were presented. The level adjustment of the target speech aimed at a threshold of 40% words correctly repeated according to the following scheme: the target speech level was changed +2 dB for zero correctly identified words, +1 dB for one correctly identified word, 0 dB for two correctly identified words, −1 dB for three correctly identified words, −2 dB for four correctly identified words, and −3 dB for five correctly identified words. The 40% threshold and the adaptive scheme for level adjustment were based on computer simulations and analysis of the maximum steepness of the psychometric function (27–29). The SRT was defined as the mean of the SNRs for the last 10 of the totally 20 presented sentences (29, 30). The mean (SD) test–retest difference for this task was previously estimated in adults to 1.1 dB (1.4 dB) (26).

TABLE 2 Individual hearing thresholds of the impaired ear.

Subject ID	AC PTA_4 imp (dB HL)	BC PTA_4 imp (dB HL)	AC PTA_4 aided imp (dB HL)	AC PTA_4 better ear (dB HL)
1	65	5	18.5	6
2	65	11	16.0	0
3	65	11	29.1	8
4	65	0	17.3	5
5	66	11	42.7	3
6	65	10	17.5	9
7	61	4	32.2	6
8	63	10	31.2	5
9	74	9	30.1	5
10	53	0	18.3	3
11	64	4	19.6	4
Max	74	11	42.7	9
Min	53	0	16.0	0
Mean ± SD	64.2 ± 4.9	6.8 ± 4.4	24.8 ± 8.7	4.9 ± 2.5

Imp, impaired ear; PTA_4, pure-tone average; AC, air conduction threshold; BC, bone conduction threshold.
Air conduction thresholds of the better ear are also presented.

3.6. Subjective assessment

The PEACH score is a questionnaire consisting of 13 questions assessing auditory behaviors of the child in different situations and is to be filled in by the caregiver. It is divided into two domains, quiet and noise. The questionnaire was developed from the more extensive PEACH diary (31) and is validated in Swedish (32). It is also used in the national pediatric hearing register in Sweden. The questionnaire was sent home to the study participants in advance along with written instructions on how to fill out the form. If the caregiver had not filled out the form at home, the caregiver was allowed to fill out the questionnaire on the day of testing.

3.7. Statistical calculations

PTA_4 were calculated as the mean of hearing thresholds at 500, 1,000, 2,000, and 4,000 Hz. Statistical calculations were conducted using GraphPad Prism 9.3.1 (350). Correlations with age, age at first fitting of the BCD, mean time of usage per day, duration of device use, and unaided PTA_4 as predictor variables and SLA and SRT as dependent variables were performed using non-parametric tests (Spearman correlation) due to skewed distribution of the data. Paired comparisons (unaided vs. aided listening) were also performed using non-parametric tests (Wilcoxon signed rank).

4. Results

The final sample comprised 11 children aged 5.3–10.8 years (mean 7.9 years, SD 1.9). All study participants had a normal eardrum on the non-atretic side. Background data are presented in **Table 1**. For all participants, age of first fitting was decided as when they first started using a BCD on softband mean (SD) of 1.5 (1.8) years. At the time of testing, three individuals used a conventional BCD on softband, one participant used a passive transcutaneous BAHA Attract, and seven individuals used active percutaneous devices. Historical data on device use were not available to the authors. Data on mean time of usage per day since their last control were retrieved from the fitting software. The study participants used their BCD for a mean (SD) of 4.8 (2.6) h per day (**Table 1**).

4.1. Hearing thresholds

The mean (SD) PTA_4 of the atretic ear was 64.3 (4.9) dB HL (**Table 2**). All study participants improved their PTA of the atretic ear when using their BCD, resulting in a mean (SD) PTA_4

of 24.9 (8.8) dB HL. Six of the study participants (subjects 1, 2, 4, 6, 10, and 11) reached aided hearing thresholds <25 dB HL.

Individual results from the sound localization test and speech recognition are summarized in Table 3 and Figure 4.

4.2. Sound localization accuracy

SLA data from two study participants could not be interpreted and were excluded from further analysis (one participant did not cooperate to testing, possibly due to tiredness; one participant had a congenital eye anomaly that made eye tracking not possible). The mean (SD) of unaided EI was 0.48 ± 0.17, whereas the mean (SD) of aided EI was 0.37 ± 0.05; individual results are plotted in Figure 4A and presented in Table 3. There was no statistically significant difference between the unaided and aided results ($p = 0.078$, $n = 9$, Wilcoxon matched pairs).

Based on previous calculations on test–retest reliability for the localization task, and an estimate of the 95% confidence interval for a single error index value based on this reliability (95% CI = ±0.054 for adults; 95% CI = ±0.12 for infants), we analyzed intra-individual performance differences (unaided vs. aided) in localization accuracy. The three study participants who showed the poorest unaided SLA (participant 2, 6, and 9) showed intra-individual statistically significant improvements when tested with the BCD (±0.12) ($p < 0.05$).

Age at testing did not have a statistically significant effect on SLA performance on a group level (unaided $\rho = -0.44$, $p = 0.239$; aided $\rho = 0.29$, $p = 0.45$, Spearman correlation) (Figure 5A), or on the benefit from amplification on the task (i.e., the difference between unaided and aided SLA, $\rho = 0.62$, $p = 0.08$, Spearman correlation). The two youngest participants had the worst unaided SLA performance, as well as the most benefit from amplification. Age at first fitting did not correlate with SLA performance in listening condition (aided $\rho = -0.28$, $p > 0.05$; unaided $\rho = 0.02$, $p > 0.05$, Spearman correlation), time of usage per day (unaided $\rho = 0.05$, $p = 0.92$; aided $\rho = -0.41$, $p > 0.05$, Spearman correlation), or duration of device use (aided $\rho = 0.18$, $p > 0.05$; unaided $\rho = -0.34$, $p > 0.05$, Spearman correlation). The participant with the lowest time of usage performed worse in the aided compared to the unaided setting. Unaided SLA was found to be correlated to unaided PTA_4 of the atretic ear ($\rho = 0.93$, $p = 0.007$, Spearman correlation) (Figure 6A), indicating increased localization accuracy with increasing unaided hearing sensitivity. There was no such correlation for aided hearing thresholds and aided SLA (Figure 6B) ($\rho = 0.24$, $p = 0.525$, Spearman correlation).

4.3. SRT in competing speech

One subject did not finish the speech recognition test due to tiredness and was not included in the analysis, i.e., 10 children provided data for this test. Data are presented in Table 3. The mean (SD) SRT was comparable for aided [−7.9 (3.5) dB] and unaided [−7.7 (3.4) dB] listening conditions ($p > 0.05$, Wilcoxon matched pairs) (Table 3 and Figure 4B). SRTs improved with increasing age (unaided $\rho = -0.88$, $p = 0.002$; aided $\rho = -0.69$, $p = 0.033$, Spearman correlation) (Figure 5B) but was not found to be affected by age at first fitting of the BCD (unaided $\rho = -0.22$, $p = 0.505$; aided $\rho = 0.02$, $p = 0.755$, Spearman correlation) or time of usage per day (unaided $\rho = 0.17$, $p = 0.703$; aided $\rho = 0.12$, $p = 0.793$, Spearman correlation). Aided SRTs were found to improve with increased duration of use in years ($\rho = -0.67$, $p = 0.04$, Spearman correlation); however, duration of use in years also correlate with the age of the study participants. Correlation of duration of use was not found in the unaided performance ($\rho = -0.40$, $p = 0.25$). There was a trend toward a correlation between unaided PTA_4 of the atretic ear to unaided performance (Figure 6C); this was however not significant ($p = 0.061$, Spearman correlation). No correlation was found between aided hearing thresholds and aided SRT (Figure 6D) ($\rho = -0.10$, $p = 0.785$, Spearman correlation).

4.4. PEACH questionnaire

Caregivers of all participating subjects filled out the PEACH questionnaire ($n = 11$). The caregiver of one child could not fill in the unaided part of the questionnaire as the study participant used its BCD "during all waking hours." One subject had not been well during the last week, and one had not been using the BCD during the last week. These questionnaires were excluded from analysis. The total score and the scores for the quiet and noise domains are presented in Table 4. Parents reported significantly higher scores in quiet than in noise for both unaided ($p < 0.05$, Wilcoxon matched pairs) and aided ($p < 0.05$, Wilcoxon matched pairs) conditions, indicating that the study participants might have greater difficulties listening in noisy than in quiet environments. There was no significant difference between unaided and aided scores in any of the domains [Total score $p = 0.945$, quiet domain $p = 0.375$, noise domain $p = 0.125$ (Wilcoxon matched pairs)].

5. Discussion

The aim for this study was to investigate the impact of unilateral bone conduction amplification before 3 years of age on horizontal sound localization and recognition of speech in spatially separate competing speech in children with UAA. Although all study participants improved their hearing thresholds in the aided condition, the intra-individual variability in the benefit for SLA and SRTs was large. Results from this pilot study indicate that fitting with a BCD before the age of 3 does not seem to negatively affect horizontal sound localization accuracy in children with UAA and might be beneficial to some individuals. Results on speech recognition were more diverse, where four individuals showed a worse performance in the aided setting. In a review by Vogt et al. (33), they found that aided hearing thresholds did not approach normal levels in six out of nine included studies and also suggested that an insufficient degree of amplification might be a part explanation for poorer aided speech recognition scores. Several of the study participants

TABLE 3 Individual results on SLA and SRT, aided and unaided.

Subject ID	AC PTA_4 (dB HL)[a]	SLA unaided (EI)	SLA aided (EI)	SLA benefit (EI)	SRT unaided (dB)	SRT aided (dB)	SRT benefit (dB)
1	65	0.410	0.330	−0.080	−5.1	−9.1	−4.0
2	65	0.520	0.340	−0.180[b]	−8.6	−7.3	1.3
3	65	—	—	—	−7.4	−8.5	−0.9
4	65	0.470	0.360	−0.110	−9.7	−6.7	3.0
5	66	—	—	—	−5.4	−0.3	4.1
6	65	0.630	0.360	−0.270[b]	—	—	—
7	61	0.330	0.330	0.000	−9.5	−10.2	−2.3
8	63	0.360	0.450	0.090	−10.8	−12.7	−1.9
9	74	0.870	0.420	−0.450[b]	−0.3	−6.4	−5.9
10	53	0.360	0.280	−0.080	−7.9	−5.7	2.2
11	64	0.377	0.423	0.046	−12.1	−11.8	0.3
Max	74	0.87	0.45	−0.450	−12.1	−12.7	−5.9
Min	53	0.33	0.28	0.090	−0.3	−0.3	4.1
Mean ± SD	64.2 ± 4.9	0.48 ± 0.17	0.37 ± 0.05	−0.11 ± 0.17	−7.7 ± 3.4	−7.9 ± 3.5	−0.41 ± −3.18

A more negative SRT value indicates a better performance. There was no significant benefit from amplification on SLA or SRT on a group level ($p > 0.05$, Wilcoxon matched pairs). Three individuals showed significant intra-individual change in SLA when comparing unaided to aided scores (a change of ± 0.12 being statistically significant in infants ($p < 0.05$).

[a]Unaided air conduction thresholds of the atretic ear.

[b]Significant intra-individual change $p < 0.05$.

in the present study did not reach normal hearing levels of the atretic ear in the aided setting. However, aided hearing thresholds did not significantly affect SRT on a group level.

Age at fitting was not related to SLA or SRT, suggesting that early treatment with a BCD for congenital UAA will not negatively affect these abilities on a group level. Amplification benefits for SLA and SRT were more evident in the younger individuals, who were also fitted at an earlier age (before 1 year of age, subjects 5, 6, and 9). Two of these individuals (6 and 9; no SLA data were collected for subject 5) improved their SLA performance and all three improved their SRTs in the aided condition. Subjects 6 and 9 also had the worst unaided PTA_4 as well as the worst unaided SLA performance. Agterberg et al. (34) suggested that individuals with worse unaided SLA might benefit more from amplification, consistent with the above stated findings.

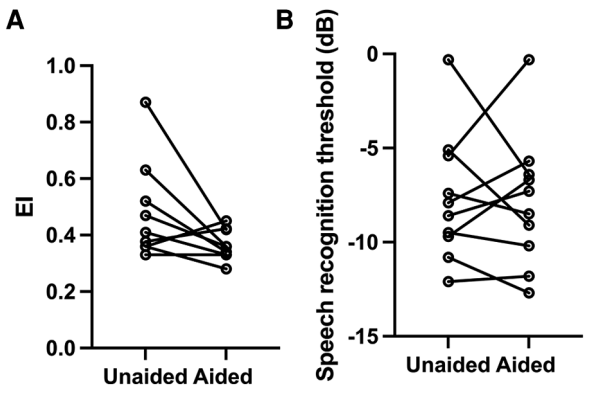

FIGURE 4
(A) Individual values of unaided and aided SLA performance. A lower EI indicates a better performance. There was no significant benefit from amplification on a group level (Spearman correlation, $p > 0.05$). In the aided setting, the three individuals with the highest unaided EI had intra-individual statistically significant benefit compared to the unaided measurements ($p < 0.05$). (B) There was a high degree of inter-individual variability on SRT both aided and unaided. A more negative SRT (dB) value indicates a better performance.

Both aided and unaided SRTs increased with increasing age, which might be expected since development of speech recognition is known to continue into adolescence (35). For individuals with congenital UAA, an age effect on SRT is further confirmed by a comparison with data from the current study and adults with UAA [$n = 12$, mean (SD) −10.9 dB (1.4 dB), $p = 0.008$, unpaired t-test] (25).

The time of usage of the BCD in this study was quite low and varied between 0.3 to 7.5 h per day but did not seem to influence the results on SLA or SRT. The duration of device use was found to affect aided SRTs but correlates also with the age of the study participants making it hard to draw conclusions from this finding. All study participants had been initially fitted with a BCD on softband before receiving a percutaneous or transcutaneous device, but eight out of 11 subjects had changed to a different system when the study took place. Information on at what age these individuals had received their surgically implanted solutions and audiological data from the previously used BCD on softband with correlated data on time of usage per day was unavailable to the authors. It is possible that children who use their BCD more frequently as toddlers might have greater benefit from the device. Compared to percutaneous devices, passive transcutaneous devices and conventional devices have approximately 10–15 dB lower amplification due to the attenuation of the skin and soft tissues of the scull (36, 37). Even though we found no correlation between aided hearing thresholds and the effect on SLA or SRT from amplification, an effect on the results from the study participants using different BCD systems cannot be ruled out. Due to the small sample size, we were unable to analyze whether the use of different systems might have influenced the results.

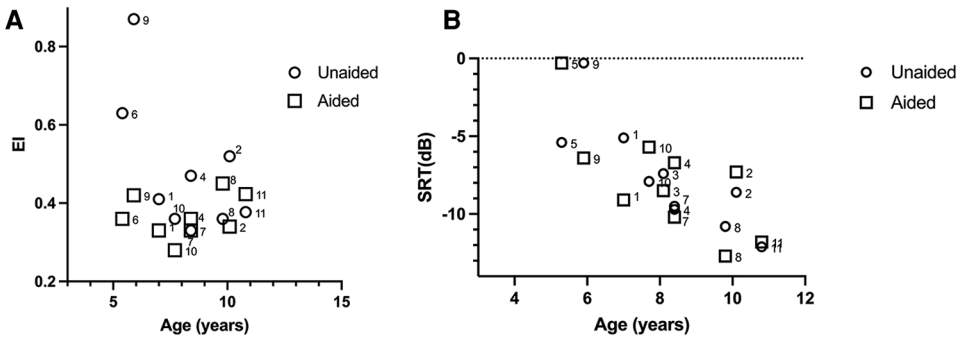

FIGURE 5
(A) SLA performance unaided and aided correlated to age. An EI value of 0 indicates a perfect performance and a value of 1 a random performance. Two of the youngest participants (6 and 9) had the worst unaided SLA performance, but there was no significant correlation between SLA and age. The youngest individuals had the most benefit from amplification. (B) SRT correlated with age. A more negative value indicates better speech discrimination. Both aided and unaided SRTs were correlated to age ($p < 0.05$), where the older study participants had lower (better) SRTs.

Binaural cues, such as interaural time differences (ITDs) and interaural level differences (ILDs) are known to be important for localizing sound in the horizontal plane (38). For individuals with UAA, detection and processing of ITDs and ILDs may be compromised because of reduced audibility in one ear. For horizontal SLA in normal binaural hearing, ITDs have been shown to be dominant, overthrowing ILD and spectral cues for low frequency sounds (39). Monaural spectral cues, resulting from acoustic reflections in the pinna, shoulders, and body are used for localization in the vertical plane in normal hearing

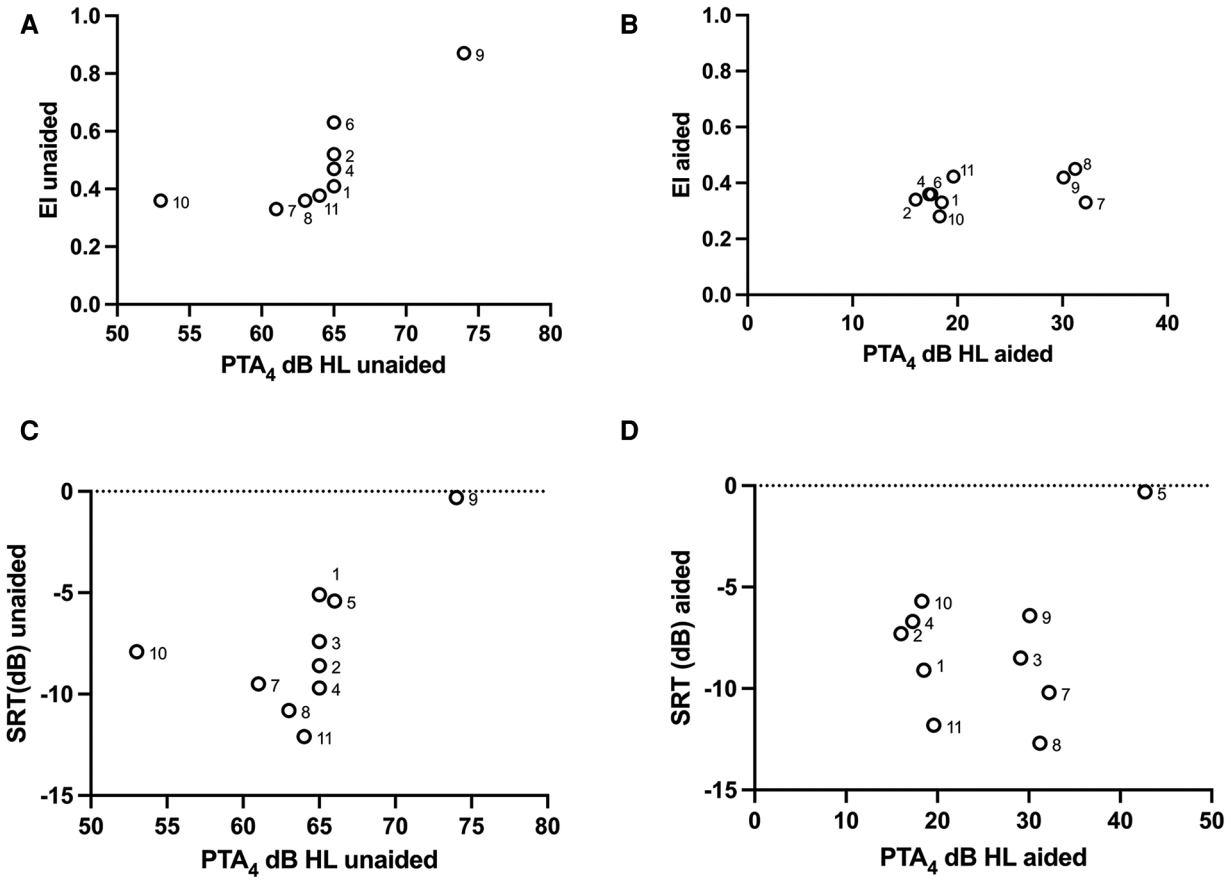

FIGURE 6
(A) Unaided SLA was correlated to unaided PTA_4 ($p < 0.05$). (B) There was no correlation between aided PTA_4 and aided SLA performance. (C) Unaided SRT related to unaided PTA_4. There was no significant correlation between unaided audibility in the atretic ear and unaided speech recognition thresholds ($p > 0.05$, Spearman correlation). (D) No significant correlation was found between aided SRTs and aided PTA_4 ($p > 0.05$, Spearman correlation).

TABLE 4 PEACH scores aided and unaided separated by domain.

Total PEACH score %, mean (SD)		Quiet domain %, mean (SD)		Noise domain %, mean (SD)	
Uniaded	Aided	Unaided	Aided	Unaided	Aided
82.5 (11.3)	81.6 (9.6)	88.5 (10.6)	91.3 (8.0)	70.0 (14.1)	73.5 (11.8)

Results were significantly higher in the quiet domain than in the noise domain in the unaided and in the aided condition ($p < 0.05$, Wilcoxon matched pairs). There was however no significant effect from amplification in any of the domains ($p > 0.05$, Wilcoxon matched pairs).

listeners (40) and might be of importance for unaided horizontal sound localization in individuals with UAA (8, 11, 33, 34). Here, monaural cues were likely available to the children, but not very prominent given the naturally occurring amplitude modulations of the sound. As such, processing of interaural cues should be important to reach good performance in the task used in the present study. The statistically significant correlation between unaided PTA_4 and unaided SLA observed needs to be evaluated in a larger sample due to co-variating factors such as the age at first fitting and the duration of time during which amplification had been available. Also, PTA_4 values of the children tested here were clustered around 65 dB HL (**Figure 6A**) and the correlation observed depended heavily on two outliers. Notwithstanding that this correlation may not be present in a larger study group, a discussion on the possible influence of the audibility of the atretic ear on localization accuracy is warranted. First, it might be that individuals with less severe hearing loss secondary to UAA might be able to utilize interaural differences for localizing sound in the horizontal plane, since even very low sound levels in the ear with poorer thresholds provide access to interaural localization cues (39). Second, also giving some support to our finding that the PTA_4 of the atretic ear may be an important predictor of localization accuracy, a similar relationship has been observed in adults with UAA (25) using the same localization technique as in the present study. Third, previous studies in individuals with UAA have demonstrated an increase in localization accuracy at high presentation levels (41, 42). An interpretation of this localization improvement is that both cochleae are stimulated (because of the increased presentation level) and interaural differences may be utilized for sound source localization despite the unilateral hearing loss. This may also be what occurred in the present study in the children with the lowest hearing thresholds.

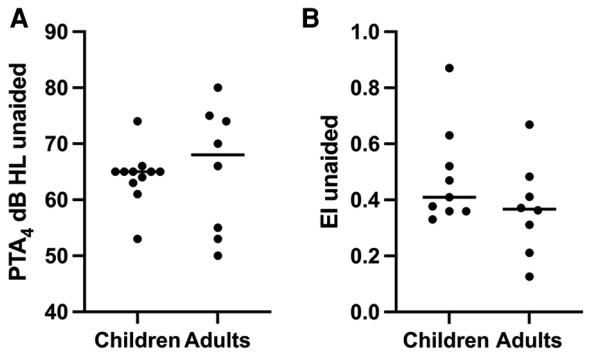

FIGURE 7
Comparison of unaided PTA_4 (A) and SLA (B) between children from the present study and adults from the study by Siegbahn et al. (25). The adults had a tendency toward a higher PTA_4 and lower EI compared to the children in the present study where the spread tends to approach lower PTA_4 and a higher EI.

When comparing the localization results of the present study to those of adults with UAA (25), the adults had a tendency toward a lower EI (i.e., better localization) compared to the children in the present study (**Figure 7**). This is noteworthy, since localization accuracy for normal binaural hearing seems mature at approximately 5–6 years of age (15, 16). The younger participants in the present study showed the worst unaided SLA; however, the effect on amplification was more evident in these individuals as they approached the EI of the older study participants in the aided setting. This could indicate that maturation of SLA is not delayed in these children compared to normal hearing individuals.

Similar to scores from children with moderate unilateral sensorineural hearing loss, results from the overall PEACH scores were lower in the aided and unaided condition than those for normal hearing children in the same age group (22, 31). Parental ratings of aural/oral performance were comparable for unaided and aided listening, suggesting that parents are not able to discriminate whether the BCD is beneficial to the child in the situations described in the questionnaire.

6. Study limitations

The statistical power of this study is limited. The authors were only able to present a small sample as only 11 out of 31 individuals that met inclusion criteria decided to take part in the study. Several of the predictor variables co-varied making it difficult to draw conclusions from the results. Aided hearing thresholds also varied in the studied cohort, where five study participants had aided hearing thresholds within the range of mild-to-moderate hearing loss. However, the studied cohort was homogenous regarding hearing thresholds of the non-atretic ear and all study participants had normal BC thresholds in the atretic ear. There was no formal procedure to ensure that the device was fully functioning prior to testing, but since all participants of the study improved their hearing thresholds in the aided setting, we assumed that the BCD was functional. Information on longitudinal device use was not available to the authors.

7. Conclusion

Collectively, the results from the current pilot study indicate that the introduction of BCD amplification before 3 years of age in children with UAA does not seem to affect horizontal sound localization accuracy and might result in benefit for horizontal sound localization for some individuals. The effect of early access to amplification on recognition of speech in spatially and symmetrically separated competing speech is more diverse. While

there is no significant effect on a group level, some individuals might perform worse in the aided setting. In the future, it would desirable to be able to predict which individuals might benefit more from amplification. The effects of early fitting of a BCD in UAA on spatial hearing needs to be evaluated in a larger sample.

Data availability statement

The raw data supporting the conclusions of this article will be made available by the authors, without undue reservation.

Ethics statement

The studies involving humans were approved by the regional Ethical Committee in Stockholm 2012/1661-31/3. The studies were conducted in accordance with the local legislation and institutional requirements. Written informed consent for participation in this study was provided by the participants' legal guardians/next of kin. Written informed consent was obtained from the minor(s)' legal guardian/next of kin for the publication of any potentially identifiable images or data included in this article.

Author contributions

HJ, CB, MH and FA significantly contributed to the design of the study. HD and CB recruited study participants and did the clinical examinations. FA conducted measurements of spatial hearing. HD collected the data and performed statistical calculations along with FA. HD, FA and CB interpreted the data. All authors contributed to the article and approved the submitted version.

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Conflict of interest

The authors declare that the research was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

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References

1. Priwin C, Jönsson R, Magnusson L, Hultcrantz M, Granström G. Audiological evaluation and self-assessed hearing problems in subjects with single-sided congenital external ear malformations and associated conductive hearing loss. *Int J Audiol.* (2007) 46(4):162–71. doi: 10.1080/14992020601077984

2. Reinfeldt S, Håkansson B, Taghavi H, Eeg-Olofsson M. New developments in bone-conduction hearing implants: a review. *Med Devices (Auckl).* (2015) 8:79–93. doi: 10.2147/mder.S39691

3. Nelissen RC, Mylanus EA, Cremers CW, Hol MK, Snik AF. Long-term compliance and satisfaction with percutaneous bone conduction devices in patients with congenital unilateral conductive hearing loss. *Otol Neurotol.* (2015) 36 (5):826–33. doi: 10.1097/MAO.0000000000000765

4. Pfiffner F, Kompis M, Stieger C. Bone-anchored hearing aids: correlation between pure-tone thresholds and outcome in three user groups. *Otol Neurotol.* (2009) 30 (7):884–90. doi: 10.1097/MAO.0b013e3181b4e8eb

5. Nadaraja GS, Gurgel RK, Kim J, Chang KW. Hearing outcomes of atresia surgery versus osseointegrated bone conduction device in patients with congenital aural atresia: a systematic review. *Otol Neurotol.* (2013) 34(8):1394–9. doi: 10.1097/MAO.0b013e3182a36065

6. Kunst SJ, Leijendeckers JM, Mylanus EA, Hol MK, Snik AF, Cremers CW. Bone-anchored hearing aid system application for unilateral congenital conductive hearing impairment: audiometric results. *Otol Neurotol.* (2008) 29(1):2–7. doi: 10.1097/mao.0b013e31815ee29a

7. Hol MK, Snik AF, Mylanus EA, Cremers CW. Does the bone-anchored hearing aid have a complementary effect on audiological and subjective outcomes in patients with unilateral conductive hearing loss? *Audiol Neurootol.* (2005) 10(3):159–68. doi: 10.1159/000084026

8. Vyskocil E, Liepins R, Kaider A, Blineder M, Hamzavi S. Sound localization in patients with congenital unilateral conductive hearing loss with a transcutaneous bone conduction implant. *Otol Neurotol.* (2017) 38(3):318–24. doi: 10.1097/MAO.0000000000001328

9. Vogt K, Frenzel H, Ausili SA, Hollfelder D, Wollenberg B, Snik AFM, et al. Improved directional hearing of children with congenital unilateral conductive hearing loss implanted with an active bone-conduction implant or an active middle ear implant. *Hear Res.* (2018) 370:238–47. doi: 10.1016/j.heares.2018.08.006

10. Vogt K, Wasmann JW, Van Opstal AJ, Snik AFM, Agterberg MJH. Contribution of spectral pinna cues for sound localization in children with congenital unilateral conductive hearing loss after hearing rehabilitation. *Hear Res.* (2020) 385:107847. doi: 10.1016/j.heares.2019.107847

11. Agterberg MJH, Hol MKS, Cremers C, Mylanus EAM, van Opstal AJ, Snik AFM. Conductive hearing loss and bone conduction devices: restored binaural hearing? *Adv Otorhinolaryngol.* (2011) 71:84–91. doi: 10.1159/000323587

12. Priwin C, Jönsson R, Hultcrantz M, Granström G. BAHA in children and adolescents with unilateral or bilateral conductive hearing loss: a study of outcome. *Int J Pediatr Otorhinolaryngol.* (2007) 71(1):135–45. doi: 10.1016/j.ijporl.2006.09.014

13. Johnstone PM, Nabelek AK, Robertson VS. Sound localization acuity in children with unilateral hearing loss who wear a hearing aid in the impaired ear. *J Am Acad Audiol.* (2010) 21(8):522–34. doi: 10.3766/jaaa.21.8.4

14. Van Deun L, van Wieringen A, Van den Bogaert T, Scherf F, Offeciers FE, Van de Heyning PH, et al. Sound localization, sound lateralization, and binaural masking level differences in young children with normal hearing. *Ear Hear.* (2009) 30 (2):178–90. doi: 10.1097/AUD.0b013e318194256b

15. Eklöf M, Asp F, Berninger E. The development of sound localization latency in infants and young children with normal hearing. *Trends Hear*. (2022) 26:23312165221088398. doi: 10.1177/23312165221088398

16. Litovsky RY. Review of recent work on spatial hearing skills in children with bilateral cochlear implants. *Cochlear Implants Int*. (2011) 12(Suppl 1):S30–4. doi: 10.1179/146701011X13001035752372

17. Berninger E, Olofsson Å, Leijon A. Analysis of click-evoked auditory brainstem responses using time domain cross-correlations between interleaved responses. *Ear Hear*. (2014) 35(3):318–29. doi: 10.1097/01.aud.0000441035.40169.f2

18. Berninger E, Gustafsson LL. Changes in 2f1−f2 acoustic distortion products in humans during quinine-induced cochlear dysfunction. *Acta Otolaryngol*. (2000) 120 (5):600–6. doi: 10.1080/000164800750000405

19. Asp F, Jakobsson AM, Berninger E. The effect of simulated unilateral hearing loss on horizontal sound localization accuracy and recognition of speech in spatially separate competing speech. *Hear Res*. (2018) 357:54–63. doi: 10.1016/j.heares.2017.11.008

20. ISO. *ISO 4869-1: Acoustics-hearing protectors-part 1: subjective method for the measurement of sound attenuation*. Geneva: International Organization for Standardization (1990).

21. Asp F, Olofsson Å, Berninger E. Corneal-reflection eye-tracking technique for the assessment of horizontal sound localization accuracy from 6 months of age. *Ear Hear*. (2016) 37(2):e104–18. doi: 10.1097/AUD.0000000000000235

22. Johansson M, Asp F, Berninger E. Children with congenital unilateral sensorineural hearing loss: effects of late hearing aid amplification—a pilot study. *Ear Hear*. (2020) 41(1):55–66. doi: 10.1097/AUD.0000000000000730

23. Denanto FM, Wales J, Tideholm B, Asp F. Differing bilateral benefits for spatial release from masking and sound localization accuracy using bone conduction devices. *Ear Hear*. (2022) 43(6):1708–20. doi: 10.1097/aud.0000000000001234

24. Gredebäck G, Johnson S, von Hofsten C. Eye tracking in infancy research. *Dev Neuropsychol*. (2010) 35(1):1–19. doi: 10.1080/87565640903325758

25. Siegbahn M, Engmér Berglin C, Hultcrantz M, Asp F. Adults with unilateral congenital ear canal atresia - sound localization ability and recognition of speech in competing speech in unaided condition. *Acta Otolaryngol*. (2021) 141(7):689–94. doi: 10.1080/00016489.2021.1921843

26. Berninger E, Karlsson KK. Clinical study of Widex Senso on first-time hearing aid users. *Scand Audiol*. (1999) 28(2):117–25. doi: 10.1080/010503999424842

27. Hagerman B. Sentences for testing speech intelligibility in noise. *Scand Audiol*. (1982) 11(2):79–87. doi: 10.3109/01050398209076203

28. Hagerman B. Reliability in the determination of speech reception threshold (SRT). *Scand Audiol*. (1979) 8(4):195–202. doi: 10.3109/01050397909076321

29. Hagerman B, Kinnefors C. Efficient adaptive methods for measuring speech reception threshold in quiet and in noise. *Scand Audiol*. (1995) 24(1):71–7. doi: 10.3109/01050399509042213

30. Plomp R, Mimpen AM. Improving the reliability of testing the speech reception threshold for sentences. *Audiology*. (1979) 18(1):43–52. doi: 10.3109/00206097909072618

31. Bagatto MP, Scollie SD. Validation of the parents' evaluation of aural/oral performance of children (PEACH) rating scale. *J Am Acad Audiol*. (2013) 24 (2):121–5. doi: 10.3766/jaaa.24.2.5

32. Brännström KJ, Ludvigsson J, Morris D, Ibertsson T. Clinical note: validation of the Swedish version of the parents' evaluation of aural/oral performance of children (PEACH) rating scale for normal hearing infants and children. *Hearing Balance Commun*. (2014) 12(2):88–93. doi: 10.3109/21695717.2014.903030

33. Vogt K, Desmet J, Janssen AM, Agterberg MJH, Snik AFM. Unexplained variation in benefit of treatment of congenital unilateral aural atresia: a review of the literature. *Audiol Neurootol*. (2021) 26(5):295–302. doi: 10.1159/000512245

34. Agterberg MJ, Snik AF, Hol MK, van Esch TE, Cremers CW, Van Wanrooij MM, et al. Improved horizontal directional hearing in bone conduction device users with acquired unilateral conductive hearing loss. *J Assoc Res Otolaryngol*. (2011) 12 (1):1–11. doi: 10.1007/s10162-010-0235-2

35. Koopmans WJA, Goverts ST, Smits C. Speech recognition abilities in normal-hearing children 4 to 12 years of age in stationary and interrupted noise. *Ear Hear*. (2018) 39(6):1091–103. doi: 10.1097/AUD.0000000000000569

36. Verstraeten N, Zarowski AJ, Somers T, Riff D, Offeciers EF. Comparison of the audiologic results obtained with the bone-anchored hearing aid attached to the headband, the testband, and to the "snap" abutment. *Otol Neurotol*. (2009) 30 (1):70–5. doi: 10.1097/MAO.0b013e31818be97a

37. Håkansson B, Tjellström A, Rosenhall U, Carlsson P. The bone-anchored hearing aid. Principal design and a psychoacoustical evaluation. *Acta Otolaryngol*. (1985) 100(3–4):229–39. doi: 10.3109/00016488509104785

38. Portfors CV, von Gersdorff H. Macrocircuits for sound localization use leaky coincidence detectors and specialized synapses. *Neuron*. (2013) 78(5):755–7. doi: 10.1016/j.neuron.2013.05.034

39. Wightman FL, Kistler DJ. The dominant role of low-frequency interaural time differences in sound localization. *J Acoust Soc Am*. (1992) 91(3):1648–61. doi: 10.1121/1.402445

40. Grothe B, Pecka M, McAlpine D. Mechanisms of sound localization in mammals. *Physiol Rev*. (2010) 90(3):983–1012. doi: 10.1152/physrev.00026.2009

41. Thompson NJ, Kane SLG, Corbin NE, Canfarotta MW, Buss E. Spatial hearing as a function of presentation level in moderate-to-severe unilateral conductive hearing loss. *Otol Neurotol*. (2020) 41(2):167–72. doi: 10.1097/MAO.0000000000002475

42. Canfarotta MW, Kane SLG, Buss E. Effects of presentation level on spatial hearing with and without bone-conduction amplification in congenital unilateral aural atresia. *Otol Neurotol*. (2021) 42(4):e388–92. doi: 10.1097/MAO.0000000000002991

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EDITED BY
Amanda Jane Leach,
Charles Darwin University, Australia

REVIEWED BY
Todd Wine,
University of Colorado Anschutz Medical Campus, United States
Ryan William McCreery,
Boys Town National Research Hospital, United States

*CORRESPONDENCE
Peter Carew
pcarew@unimelb.edu.au

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Language and health-related quality of life outcomes of children early-detected with unilateral and mild bilateral hearing loss

Peter Carew[1,2]*, Daisy A. Shepherd[1,3], Libby Smith[1], Qi Rui Soh[1] and Valerie Sung[1,3,4]

[1]Murdoch Children's Research Institute, Royal Children's Hospital, Parkville, VIC, Australia, [2]Department of Audiology and Speech Pathology, The University of Melbourne, Parkville, VIC, Australia, [3]Department of Paediatrics, The University of Melbourne, Parkville, VIC, Australia, [4]Centre for Community Child Health, Royal Children's Hospital, Parkville, VIC, Australia

Introduction: We aimed to describe the language and health-related quality of life (HRQoL) outcomes of children early-identified with unilateral or mild bilateral permanent hearing loss. This was a cross-sectional community-based study of children with mild bilateral or unilateral permanent hearing loss (including unilateral auditory neuropathy spectrum disorder (ANSD)), drawn from a population-based databank in Victoria, Australia.
Methods: Enrolment in this databank is independent of early intervention and amplification approaches. Language and caregiver-reported HRQoL outcomes are described by type and degree of loss at three timepoints across child development: at age 2 years (n = 255), 5–7 years (n = 173) and 9–12 years (n = 45).
Results: Across all age groups, average language outcomes were poorer than population normative scores by between a half to two thirds of a standard deviation. Children with mild bilateral hearing loss demonstrated poorer average language outcomes than children with unilateral hearing loss, particularly at younger ages. Children with unilateral ANSD showed language outcomes comparable to their peers with unilateral profound hearing loss. Children had poorer HRQoL psychosocial scores compared to physical scores, without obvious patterns of outcomes linked to degree or type of hearing loss.
Discussion: This study demonstrates children with early-identified unilateral or mild bilateral hearing loss have average language and HRQoL outcomes poorer than population normative expectations from an early age. These outcomes are observed at later ages across childhood. These findings provide a contemporary description of language and quality of life outcomes for children identified but not targeted by universal newborn hearing screening and raise questions of how to provide better support for these populations of children and their families.

KEYWORDS

unilateral hearing loss, mild bilateral hearing loss, unilateral auditory neuropathy spectrum disorder, early-identified, language outcomes, health-related quality of life

1. Introduction

Universal newborn hearing screening (UNHS) has had a transformational effect on the development pathways and early life outcomes for children born with congenital hearing loss. It is now common for identification of hearing loss to occur in the first weeks of life (1), facilitating interventions such as amplification and enrolment into early intervention

programs earlier than previously routinely possible (2). Earlier identification of hearing loss has led to improved language outcomes, although many children still have language development below expected for their age and cognitive potential (3, 4). The impact of early hearing loss identification on health-related quality of life (HRQoL) is less clear, with some studies documenting improved HRQoL in children whose hearing loss was identified through UNHS compared to without UNHS (5), whilst other studies showed no difference (4).

Many UNHS programs (e.g., in Australia and the United Kingdom) target screening for bilateral hearing losses of moderate or greater degree (6), a cut-point chosen because of evidence that earlier detection of these degrees of losses led to improved language outcomes (7). However, UNHS can and does also identify children with mild degrees of hearing loss and unilateral hearing losses—whether planned (8) or as a "by-product" of targeting bilateral moderate or greater degrees (9). Whether early or later detected, there is growing evidence of harmful effects of mild and unilateral hearing loss on several developmental domains including speech and language (10, 11). Recent amplification data from population-wide government hearing services indicate that children with mild and unilateral hearing loss represent a substantial proportion of the paediatric population presenting for amplification services. Hearing Australia, the national provider of hearing amplification for children in Australia, reports the highest proportion of children first fitted with amplification under 12 months of age have an average hearing loss in the better hearing ear in the range of 0–40 decibels (i.e., a mild bilateral or unilateral loss) (12). Historical age at detection for these children was commonly reported to occur (prior to UNHS) around 4–5 years of age (13), or up to 8 years of age for children with unilateral loss (14). Therefore, UNHS could be viewed to have unintentionally supported the creation of a new group of children with hearing loss—those early detected with mild bilateral or unilateral hearing loss.

This "new" population (15) comes with new challenges from the time that they do not pass their newborn screen. It is recognized that diagnosis—both the duration of time to reach a diagnosis and the certainty of diagnosis—is a different process from significant bilateral losses. The number of appointments required to reach a diagnosis can be much more than for children with larger degrees of loss (16). It is likely that this leads to some stress for those involved, particularly families but also professionals (16, 17). With limited evidence for the outcomes of early-detected children with these types of loss, clinical management of these children is challenging (16, 17). Clinical practice guidelines reflect the uncertainty in outcomes for children with mild and unilateral hearing loss, with references to individual observations, watchful waiting, behavioral verification of hearing levels and needs-based approaches to the decision of if and when to provide amplification (e.g., King (18), Fitzpatrick et al. (19)).

Uncertainty, both in outcomes and management approaches, also exists for children with unilateral auditory neuropathy spectrum disorder (ANSD). This is a rare hearing profile, with estimates suggesting individuals with unilateral ANSD comprise 1%–7% of all ANSD cases (20). However, these children are also detected early and the parental uncertainty regarding appropriate approaches to supporting development of language and communication reported for children with bilateral ANSD (21) may also be a factor for their unilateral ANSD peers. Outside of case reports, published studies including individuals with unilateral ANSD have focused on describing the clinical characteristics of impacted individuals (20, 22) or detail electrophysiological traits and characteristics (23) rather than their developmental outcomes.

This study addresses the gap in literature on the outcomes of children with early-detected mild and unilateral hearing loss. We describe the language and HRQoL outcomes of a contemporary population of children with different degrees of non-target hearing loss (i.e., hearing loss that was not the target for UNHS in Australia) including unilateral ANSD at different ages across child development.

2. Methods

2.1. Study design and participants

This was a cross-sectional study of Australian children whose degree of permanent hearing loss at diagnosis was either of mild degree in at least the better ear (grouped as mild bilateral), or unilateral of any degree (mild, moderate, severe or profound). This group represents the group of children whose hearing was not the target for UNHS (i.e., not bilateral moderate to profound) in Australia. Children with a diagnosis of unilateral ANSD were also included. Outcomes of participants, collected between 2014 and 2023, were drawn from set data-collection points of a databank built to track the developmental outcomes of children born with permanent hearing loss, the Victorian Childhood Hearing Longitudinal Databank (VicCHILD).

VicCHILD is a population-level data repository, open to all children born or living in the state of Victoria, Australia, with any degree and type of permanent hearing loss. Recruitment into VicCHILD is currently still active, and since its inception in 2012 has over 1,200 participant families. Most VicCHILD participants are under one year of age at enrolment. The majority of participants also have access to government-supported hearing amplification and early intervention programs. Data are collected longitudinally via repeated measures across childhood, at enrolment and at key developmental stages: preschool (~2 years), primary school entry (5–7 years), and primary school exit (9–12 years). Data are collected either via caregiver-report or direct assessment, across domains covering health, physical development, quality of life, language and listening. More details on the VicCHILD methodology are available elsewhere (24). VicCHILD has ethics approval from the Royal Children's Hospital Human Research and Ethics Committee (approval number 31081), with parent/caregivers providing written informed consent.

2.1.1. Recruitment

The primary recruitment mechanism for VicCHILD is via Victoria's UNHS program, the Victorian Infant Hearing Screening Program (VIHSP), which routinely screens 99.5% of babies in the days and weeks after birth and supports families through to the point of definitive diagnosis of hearing loss (25). VIHSP sends a letter about VicCHILD to eligible families whose child has a confirmed hearing loss diagnosis from diagnostic audiology. This letter provides a two-week window for families to opt-out of learning about VicCHILD, after which time VIHSP passes contact details to the VicCHILD research team who contacts eligible families. The VicCHILD research team describes the databank and obtains consent to provide further details—after which time families decide whether to join the databank and provide consent to participate.

2.2. Outcome measures

This study reports VicCHILD's language and HRQoL outcomes in 3 different age groups, using normed and standardized measures, as described below. They were collected as part of multiple other outcome measures collected at the 3 different developmental age brackets (further details described elsewhere) (24).

2.2.1. Language measures

2.2.1.1. 2 years: expressive vocabulary

At around age 2 years, VicCHILD families received and completed either a paper-based or online survey. The primary language outcome collected for this age-group is caregiver-reported expressive vocabulary. This was measured by the 100-word checklist from the Sure Start Language Measure (SSLM) (26), designed for expressive vocabulary assessment across ages 16–30 months. To complete this measure, caregivers indicate which words from the provided list their child says. This measure, based upon the MacArthur Bates Communicative Development Inventory: UK Short Form (27), demonstrates high reliability and concurrent validity (26) and is standardized (based on the child's sex and age in months) with a mean expected score of 100, standard deviation of 15.

2.2.1.2. 5–7 years and 9–12 years: expressive and receptive language and receptive vocabulary

At both 5–7 years and 9–12 years timepoints, language outcomes were collected by direct-assessment measures, completed at a location convenient to the family (at home, at the Royal Children's Hospital, or online during the COVID−19 pandemic). For children who underwent the same assessments within the specified age brackets as part of their usual clinical care, families provided permission for these assessment results to be shared with the research team.

2.2.1.2.1. Clinical evaluation of language fundamentals recalling sentences test. The Clinical Evaluation of Language Fundamentals fourth edition (CELF-4, Australian Version) is a normed measure used both clinically and in educational settings to assess receptive and expressive language (28). The Recalling Sentences test is one subscale from the CELF-4, which along with three other subscales is used to calculate a Core Language Score. However, the Recalling Sentences test administered in isolation has been demonstrated in a large Australian population-based study to be a strong predictor of the total CELF Core Language scores (29). Consequently, we used the Recalling Sentences test as a marker of both expressive and receptive language ability. The Recalling Sentences test is standardized for the ages 5–21 years.

The Recalling Sentences test was administered via an iPad, with children repeating an audio-recorded sentence they have heard, verbatim. This method allows assessment without visual cues. Sentence length and difficulty would progress across the test. Responses are scored live by trained research assistants, rated as either "correct" (no errors), "intermediate/uncertain" (two or three errors) or "incorrect" (four or more errors). The Recalling Sentences test ends after 32 sentences, or after three consecutive "incorrect" scores. A raw score is obtained for each child ranging from 0 to 96. From this, conversion to an age-related scaled score occurs (possible values spanning 1 to 18), with a normative data mean of 10 and standard deviation of 3.

2.2.1.2.2. National institute of health toolbox picture vocabulary test. Receptive vocabulary was assessed using an adaptive test, the National Institutes of Health Toolbox Picture Vocabulary Test (NPVT) (30). The NPVT is a validated measure of general vocabulary knowledge for children aged between 3 and 17 years. On an iPad, children see four images and are required to select the image that best/most closely represents the audio recording of a word played to them. Following two practice items, up to 25 test items with a wide range of difficulty are delivered, with adjustment to difficulty made automatically according to the child's performance on the preceding word.

A theta score (similar to a *z*-score) is reported by the application at the conclusion of the test; representing the relative overall performance of the child. The NPVT provides age-adjusted, fully adjusted and unadjusted scale scores (standard scores), as well as a national percentile rank that corresponds to the age-adjusted scale score. VicCHILD calculates the standard score, which is the receptive vocabulary outcome used in this study. Based on Toolbox normative data, all scaled scores can be interpreted to understand individual performance. An age-adjusted scale score around 100 suggests vocabulary ability is at the expected level for the child's age, with scores of 115 suggesting above-average ability. A score of 85 represents below-average vocabulary ability.

2.2.2. Health-related quality of life measures: all age groups

To measure HRQoL, the Pediatric Quality of Life (PedsQL) (31) was used. A generic instrument validated for use in populations with hearing loss, the PedsQL is a standardized measure with 23 items; we used the Generic Core Scale, V4.0 in this study. The tool comprises 23 items across four domains: Physical, Emotional, Social, and School Functioning (31). With a

five-point response scale for each item reverse scored and transformed to a 0–100 scale, a score of 100 represents the best possible HRQoL in relation to questions about how much certain tasks or activities were a problem for the child.

In addition to the total score, two summary metrics are also produced from the PedsQL questionnaire: the physical health summary score, and the psychosocial health summary score. The caregiver proxy-report version was used at all ages in this study, a format demonstrated to have reliability and validity in these age groups of interest (32). Caregivers were asked to consider the child over the past one month when answering each item. Caregivers completed the PedsQL at 2 years or around 5–7 years and 9–12 years at the time of the language assessment.

2.3. Hearing loss characteristics

The definition of hearing loss for this study reflects that used by VicCHILD (24). The primary source of information on hearing loss at enrolment were UNHS records. At scheduled contact points with participating families, hearing loss records were updated using caregiver-supplied audiology reports.

Degree of hearing loss was classified using decibel ranges used by the national provider of hearing amplification, Hearing Australia (33): mild (21–40 dB), moderate (41–60 dB), severe (61–90 dB) and profound (>90 dB). Participants were recorded as having either a unilateral or bilateral hearing loss based on the presence/absence of hearing loss in the second ear. A diagnostic report stating the presence of unilateral ANSD, with normal hearing in the second ear, was used to identify our unilateral ANSD sample for this study.

Type of hearing loss for VicCHILD is not restricted to sensorineural losses. Due to this, our study sample included a small number of children identified with permanent conductive and mixed hearing losses. Children identified with unilateral aural atresia were excluded from this study as their outcomes are reported elsewhere.

2.4. Other participant characteristics

Participant characteristics were collected at enrolment and updated at each data collection point. The participant characteristics included in this study's analyses were demographic characteristics (sex, age at assessment, socioeconomic disadvantage, household income, household primary language, maternal education level) and health-related characteristics (number of comorbidities, gestational age, non-verbal IQ and whether an individual was admitted to NICU). From 2020 onwards, caregivers were asked to report on their child's additional health needs or medical diagnoses.

2.5. Study sample selection

This study included all VicCHILD participants identified to have a hearing loss diagnosis satisfying the criteria of mild bilateral hearing loss in the better ear, or a unilateral hearing loss of any degree, identified by VIHSP, born between 2005 and 2020, with data collected between December 2014 and March 2023. For each age group, children were included in the study sample if they had at least one outcome (language or HRQoL) measured at that data collection point. Hearing and demographic data were collated from data recorded at the first two collection points (enrolment and age 2 years). Updated service and device use data were also obtained at each subsequent collection point (age 5–7 years and 9–12 years).

Three study samples were formed corresponding to the three timepoints across child development, at age 2 years (early life), 5–7 years (entry to primary school) and 9–12 years (transition to secondary school), respectively. Due to the longitudinal nature of the VicCHILD databank, data from some participants were included across multiple age groups and therefore the three samples were not completely independent.

2.6. Statistical analysis

For each of the three age groups, key hearing-related, demographic and health-related characteristics were summarized. Continuous measures were reported as means and standard deviations (SD) or medians and interquartile range limits (IQR) depending on their distribution, with categorical characteristics reported as frequencies and proportions. The number of participants common to multiple age groups were quantified and reported.

Outcome measures were reported for all individuals, and then further stratified by degree of hearing loss. For each age group, the mean language measures (i.e., SSLM score, CELF recalling sentences, NPVT) were reported, alongside the SD and associated 95% confidence interval (CI). Due to the skewed nature of the PedsQL measure, the median PedsQL score and IQR were reported, alongside an estimated 95% CI using the Binomial distribution. When stratified by degree of hearing loss, the older age group (9–12 years) had small sample sizes and therefore the CI was not estimated due to low precision. We considered mean scores to represent below average performance if scores were greater than 1 standard deviation below the normative mean, with above average performance represented by scores greater than 1 standard deviation above the normative mean.

All analyses were conducted in R version 4.1.2 (34) using complete case analysis.

3. Results

3.1. Participant characteristics

Data in this study represent 473 individual records of child outcomes, spread across three timepoints: 2 years ($n = 255$), 5–7 years ($n = 173$) and 9–12 years ($n = 45$). Data from 8 participants were included in all age groups; 79 participants' data were included in both the 2 year and 5–7 years age

groups, and 34 participants' data were included in the two older age groups.

Table 1 describes the participant characteristics. Sex proportions across the three timepoints varied somewhat, with 40%–44% of participants at 2 years and 5–7 years reported female, and 53% female at 9–12 years. On average, across all ages, participants lived in areas of slightly less socio-economic disadvantage compared to the Australian population norm (mean Socio-Economic Indexes for Areas (SEIFA) scores of 1,004, 1,007 and 1,023 in increasing age group order, where a higher number represents less disadvantage, compared to normative score of 1,000). Over 80% of participants at all timepoints had reported maternal education completion being at least year 12 (completed high school), and most participants lived in households with high levels of reported income. Participants whose data were collected at the youngest timepoint (2 years) reported the highest proportion of languages used in the home being other than/additional to English. Participants were predominantly well babies, with mean gestational ages reflective of full term pregnancies and more than 80% of births not requiring admission to a neonatal intensive care unit.

Consistent with expectations of UNHS, children were diagnosed with hearing loss early in life with median age at detection ranging from 1.2 to 1.5 months across all three timepoints (Table 1). A quarter to a third of participants at all timepoints were diagnosed with a mild bilateral hearing loss; most children with unilateral hearing loss had a profound degree of loss. Participants with unilateral ANSD represented 15% of our sample at 2 years. The majority of participants had sensorineural hearing loss, with smaller proportions with mixed and permanent conductive losses, reflecting the source of the sample—from a population-based databank inclusive of all children with permanent hearing loss of any degree or type. Around 60%–75% of participants were reported to have one or more additional health need or medical diagnosis in addition to hearing loss.

The majority of participants had no hearing device fitted at the time of assessment (2 years, 55%; 9–12 years, 58%) or had hearing aid only (5–7 years, 51%). For those fitted with hearing device(s), the median age of first fitting was lowest in the younger data collection timepoints, with a median age of 6 months for participants at age 2 years (IQR: 3.0, 12.8 months) (Table 1). We observed greater proportions of hearing device use at timepoints when participants were older. Half (50%) of participants had never engaged with an early intervention program at the time data were collected at 2 years and 5–7 years. At the two timepoints where non-verbal IQ testing was possible, mean IQ scores reflected population normative scores (5–7 years, mean IQ 102 (SD 18); 9–12 years, mean IQ 100 (SD 18)).

3.2. Language

3.2.1. Early life (2yo)

When considered as a single group, children at age 2 years with unilateral or mild bilateral loss in our sample demonstrated, on average, caregiver-reported expressive vocabulary approximately two thirds of a standard deviation below population normative scores ($n = 197$, mean 90.5, 95% CI: 88.22, 92.74) (Table 2).

Children with mild bilateral and moderate unilateral losses demonstrated the poorest expressive vocabulary, with mean scores approaching a full standard deviation below population normative scores at this young age (mean 88.4, 95% CI: 84.3–92.5, and 86.3, 95% CI: 80.3, 92.3, respectively).

When considering unilateral sensorineural losses, we observed little difference in expressive language across children with mild, severe and profound losses on average, with mean vocabulary scores ranging from one third to two thirds of a standard deviation poorer than population normative scores (Figure 1), although not substantially lower comparatively to the population scores (e.g., 95% CIs presented in Table 2).

Children with unilateral ANSD demonstrated expressive vocabulary scores around two thirds of a standard deviation below population normative scores (mean 91.6, 95% CI: 86.7–96.4), a comparable mean outcome to those with profound unilateral loss (mean 91.9, 95% CI: 85.5–98.2) (Figure 1; Table 2).

3.2.2. Entry to primary school (5–7yo)

Language outcomes at this age group were, in general, poorer than population normative scores. Used as a marker of expressive and receptive language, scores on the CELF Recalling Sentences subscale suggest that when considered as a single group, children in the early primary school years with unilateral or mild bilateral hearing loss in our sample were scoring approximately two thirds of a standard deviation, on average, poorer than population normative scores ($n = 146$, mean 8.1, 95% CI: 7.4–8.7) (Table 2).

At this age point, children with mild bilateral hearing loss were, on average, one standard deviation below population normative scores (mean 6.9, 95% CI: 5.8–8.0), the poorest average performance of any hearing loss group (Figure 2; Table 2). Across unilateral sensorineural losses, we observed mean language performance within one standard deviation of population normative scores, and those with moderate, severe or profound losses having some scores approaching and exceeding the expected standardized score of 10 (Figure 2; Table 2).

Children with unilateral ANSD demonstrated a range of language performances roughly similar to children with profound unilateral losses (mean 8.6, 95% CI: 6.6–10.7, and mean 8.2, 95% CI: 7.2–9.3, respectively) (Table 2).

Of the 144 children on whom receptive vocabulary assessment had been conducted, we observed the greatest variability in performance for children with mild bilateral hearing loss in our sample (SD 25.5 points, Figure 2). On average, these children had receptive vocabulary scores in the below average range (mean 82.3, 95% CI: 75.2–89.4) (Table 2).

For children with unilateral sensorineural loss, their mean receptive vocabulary scores were closer to the expected score of 100, but still slightly poorer than population normative scores (Figure 2; Table 2) with a smaller spread of scores than observed for mild bilateral losses. Of the 15 children with

TABLE 1 Characteristics of the three study samples.

		Age 2 years		Age 5–7 years		Age 9–12 years	
		N = 255		*N* = 173		*N* = 45	
		Missing[a], *n* (%)		Missing[a], *n* (%)		Missing[a] *n* (%)	
Hearing-related characteristics							
Age at detection/diagnosis (months)—median [IQR]		21 (8.24)	1.20 [1.20, 1.20]	9 (5.20)	1.20 (1.20, 2.40)	3 (6.67)	1.50 (1.11, 2.40)
Hearing loss severity—*n* (%)		0 (0)		0 (0)		0 (0)	
Bilateral:	Mild		93 (36.47)		63 (36.42)		12 (26.67)
Unilateral:	Mild		18 (7.06)		14 (8.09)		5 (11.11)
	Moderate		34 (13.33)		25 (14.45)		6 (13.33)
	Severe		35 (13.73)		21 (12.14)		8 (17.78)
	Profound		35 (13.73)		34 (19.65)		12 (26.67)
ANSD (unilateral)			40 (15.69)		16 (9.25)		2 (4.44)
Type of hearing loss—*n* (%)		0 (0)		1 (0.62)		0 (0)	
SNHL			199 (78.04)		144 (83.72)		28 (84.44)
Auditory neuropathy			40 (16.59)		16 (9.30)		2 (4.44)
Mixed HL			8 (3.14)		5 (2.91)		1 (2.22)
Conductive HL			5 (1.96)		7 (4.07)		4 (8.89)
Not available/applicable			3 (1.18)		0 (0)		0 (0)
Amplification status at time of survey—*n* (%)		44 (17.25)		25 (14.45)		4 (8.89)	
No device			117 (55.45)		67 (45.27)		24 (58.54)
Hearing aid(s) only			89 (42.18)		76 (51.35)		17 (41.46)
CI (unilateral or bilateral) only			4 (1.90)		0 (0)		0 (0)
Hearing aid and CI			1 (0.47)		5 (3.38)		0 (0)
Frequency of device use at time of survey: *n* = 94/81/17		3 (3.19)		31 (38.27)		4 (23.53)	
<4 h			24 (26.37)		1 (2.00)		0 (5.26)
4–8 h			39 (42.86)		17 (34.00)		7 (53.85)
>8 h			28 (30.77)		32 (64.00)		6 (46.15)
Age first device fitted (months)[b]—median (IQR)		129 (50.59)	6.00 (3.00, 12.75)	92 (53.18)	18.00 (6.00, 46.00)	22 (48.89)	24.00 (9.00, 54.50)
Enrolled in early intervention services							
Ever—*n* (%)		36 (14.88)	107 (48.86)	72 (41.62)	51 (50.50)	DNC	DNC
Age at enrolment—median [IQR]		160 (62.75)	8.00 [5.00, 13.50]	DNC	DNC	DNC	DNC
Demographic characteristics							
Age at language assessment (years)—mean (SD)		58 (22.75)	2.14 (0.16)	19 (10.98)	6.90 (0.78)	8 (17.78)	11.27 (0.94)
Age at PedsQL completion (years)—mean (SD)		2 (0.08)	2.31 (0.26)	36 (20.81)	6.72 (0.79)	3 (6.67)	11.21 (0.99)
Sex of child: Female—*n* (%)		0 (0)	103 (40.39)	0 (0)	76 (43.93)	0 (0)	24 (53.33)
Socioeconomic disadvantage (SEIFA)—mean (SD)		0 (0)	1,004.72 (63.35)	0 (0)	1,007.17 (69.05)	0 (0)	1,023.93 (68.34)
Household income—*n* (%)		28 (9.80)		23 (13.29)		4 (8.89)	
<$31,199			19 (8.37)		10 (6.67)		3 (7.32)
$31,199—$51,999			21 (9.25)		18 (12.00)		3 (7.32)
$52,000—$103,999			94 (41.41)		75 (50.00)		20 (48.78)
>$104,000			93 (40.97)		47 (31.33)		15 (36.59)
Household primary language—*n* (%)		33 (12.94)		42 (24.28)		29 (64.44)	
English only			113 (50.90)		75 (57.25)		11 (68.75)
Bilingual/multilingual (English + other)			75 (33.78)		43 (32.82)		2 (12.50)
Other language(s) only			34 (15.32)		13 (9.92)		3 (18.75)
Maternal education—*n* (%)		19 (7.45)		28 (16.18)		21 (53.33)	
Year 10 or less			24 (10.17)		16 (11.03)		3 (12.50)
Year 11			12 (5.08)		6 (4.14)		1 (4.17)

(*Continued*)

TABLE 1 Continued

	Age 2 years		Age 5–7 years		Age 9–12 years	
	N = 255		*N* = 173		*N* = 45	
	Missing[a], *n* (%)		Missing[a], *n* (%)		Missing[a] *n* (%)	
Year 12		61 (25.85)		51 (35.17)		11 (45.83)
Tertiary or postgraduate		139 (58.90)		72 (49.66)		9 (37.50)
Health-related characteristics						
Number of comorbidities—*n* (%)	122 (47.84)		67 (38.73)		15 (33.33)	
None		54 (40.60)		35 (33.02)		8 (26.67)
1		45 (33.83)		29 (27.36)		7 (23.33)
2		22 (16.54)		22 (20.75)		8 (26.67)
3 or more		12 (9.02)		20 (18.87)		7 (23.33)
NICU admissions: yes—*n* (%)	8 (3.14)	48 (19.43)	9 (5.20)	26 (15.85)	1 (2.22)	7 (15.91)
Gestational age—mean (SD)	4 (1.57)	38.46 (2.38)	0 (0)	38.82 (2.43)	1 (2.22)	39.02 (2.44)
Non-verbal IQ—mean (SD)	*DNC*	*DNC*	17 (9.83)	102.51 (18.39)	2 (4.44)	100.00 (18.22)

[a]Relative to sample size for each age point unless specified in the left-hand column.
[b]Note the high level of missing information due to a large proportion of individuals most likely not ever having a device. However, this information was not collected via our data collection tool so we are unable to quantify this.
DNC corresponds to a cell in which that data/information was not collected at that time point.

TABLE 2 Summary of language and vocabulary scores for each age group.

Age 2 years		Expressive vocabulary (SSLM)							
		n	Mean	SD	95% CI				
Overall		197	90.48	16.09	(88.22, 92.74)				
By hearing loss									
Bilateral:	Mild	69	88.39	16.88	(84.34, 92.45)				
Unilateral:	Mild	11	95.73	18.75	(83.13, 108.32)				
	Moderate	28	86.32	15.47	(80.32, 92.32)				
	Severe	30	94.90	16.05	(88.91, 100.89)				
	Profound	26	91.85	15.61	(85.54, 98.15)				
ANSD (unilateral)		33	91.55	13.76	(86.67, 96.42)				
Age 5–7 years		**CELF Recalling sentences**				**NPVT**			
		n	Mean	SD	95% CI	*n*	Mean	SD	95% CI
Overall		146	8.05	3.76	(7.44, 8.67)	144	90.85	21.28	(87.34, 94.36)
By hearing loss									
Bilateral:	Mild	53	6.92	3.97	(5.83, 8.02)	52	82.32	25.54	(75.21, 89.44)
Unilateral:	Mild	10	7.60	4.58	(4.33, 10.87)	10	88.73	22.91	(72.35, 105.12)
	Moderate	22	9.18	3.59	(7.59, 10.78)	20	94.84	12.84	(88.83, 100.85)
	Severe	17	9.59	3.74	(7.66, 11.51)	16	96.15	17.85	(86.64, 105.67)
	Profound	30	8.23	2.88	(7.16, 9.31)	31	95.67	17.52	(89.24, 102.1)
ANSD (unilateral)		14	8.64	3.48	(6.63, 10.65)	15	100.87	14.20	(93.01, 108.74)
Age 9–12 years		**CELF Recalling sentences**				**NPVT**			
		n	Mean	SD	95% CI	*n*	Mean	SD	95% CI
Overall		37	8.49	3.49	(7.32, 9.65)	36	98.42	17.88	(92.37, 104.46)
By hearing loss									
Bilateral:	Mild	9	9.22	3.19	–	8	104.93	12.52	–
Unilateral:	Mild	5	7.60	5.41	–	5	89.49	34.46	–
	Moderate	5	8.60	1.82	–	5	96.54	9.12	–
	Severe	7	8.29	3.68	–	7	100.92	14.29	–
	Profound	9	7.89	3.72	–	9	99.95	17.52	–
ANSD (unilateral)		2	10.5	3.54	–	2	83.69	8.30	–

unilateral ANSD, receptive vocabulary performance was, in general, within the expected performance range (85 to 115) (Figure 2).

3.2.3. Transition to secondary school (9–12yo)

Similar to the pattern of performance seen at the entry to primary school age group, more individual performance variation was

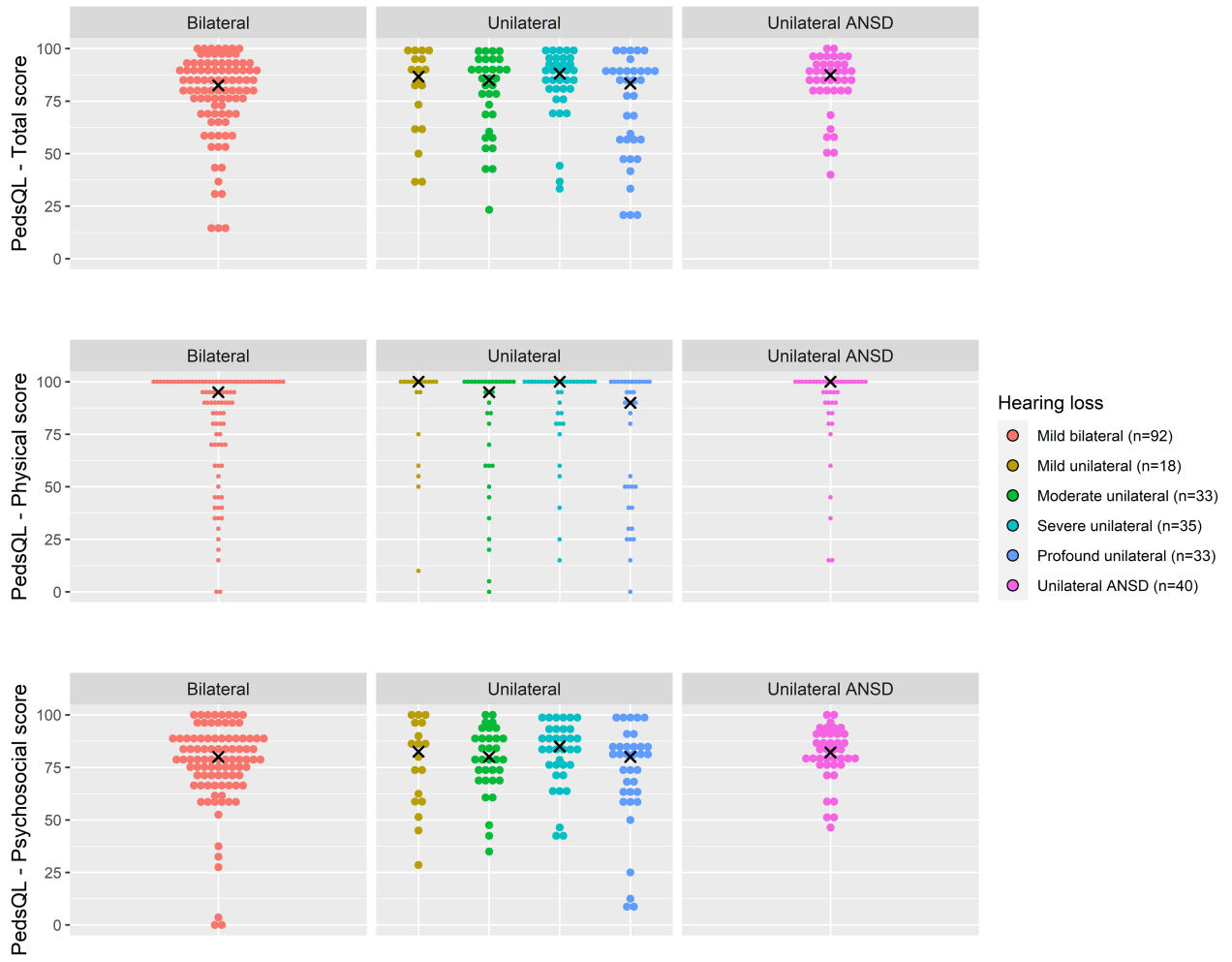

FIGURE 1
Expressive vocabulary (standardised SSLM scores) for age 2 years across hearing loss groups.

observed for expressive and receptive language outcomes in the transition to secondary school group as opposed to patterns of performance for receptive vocabulary outcomes—where mean scores approximated population normative scores (Figure 3; Table 2).

Due to small sample sizes within discrete degrees of hearing loss in this age group, aggregate results were described. Overall, mean expressive and receptive language outcomes at this age in our sample were around half a standard deviation poorer than population normative scores ($n = 37$, mean 8.5, SD 3.5) (Table 2). Mean receptive vocabulary scores were close to population normative scores ($n = 36$, mean 98.4, SD 17.9).

3.3. Health-related quality of life

Due to skewed distribution of HRQoL scores, median scores were presented. Overall, physical PedsQL scores in all age groups and for all degrees of loss were higher than psychosocial PedsQL scores (Table 3). Psychosocial PedsQL scores had a wider distribution in individual performance than physical PedsQL scores; this was particularly noticeable at our early life (2 years) and entry to primary school (5–7 years) timepoints, and was observed for all degrees of loss.

The cluster of high HRQoL scores seen at age 2 years was not so pronounced at 5–7 years and this was reflected in the shift in median psychosocial PedsQL scores (2 years psychosocial PedsQL median 80.0, IQR 70 to 90, 5–7 years psychosocial PedsQL median 70.0, IQR 60 to 82.5) (Table 3). Of note, the highest median psychosocial PedsQL score at age 5–7 years was seen in children with unilateral ANSD (median 82.5, IQR 65 to 85).

Total PedsQL scores, comprising physical and psychosocial scales, were generally similar across degrees of loss and at all age groups (Figures 4–6).

4. Discussion

4.1. Principal findings

This study describes language and HRQoL outcomes at multiple age timepoints in a large sample of children across childhood, all of whom had early identified hearing losses not

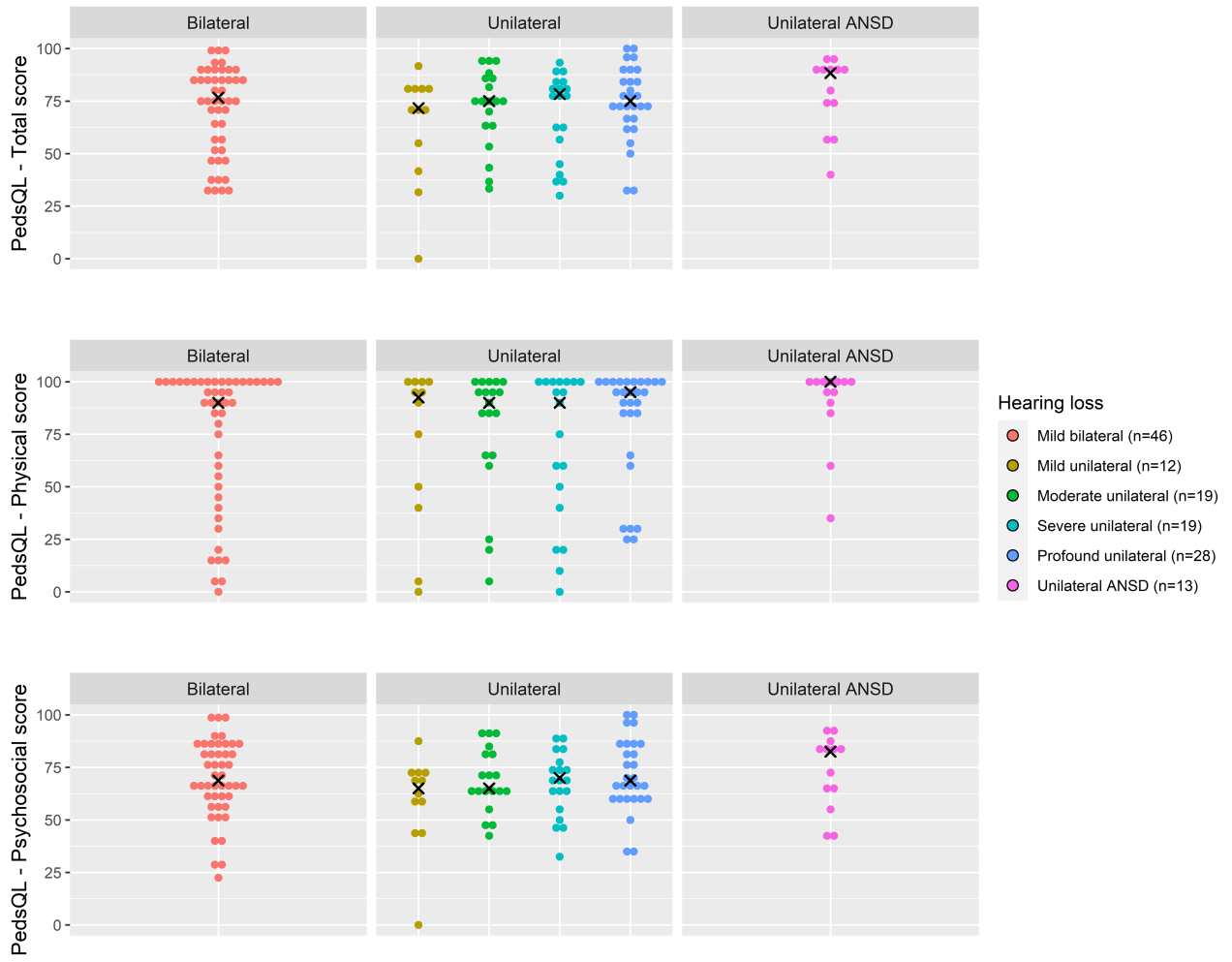

FIGURE 2
Language (CELF recalling sentences) and vocabulary (NPVT) scores for age 5–7 years across hearing loss groups.

targeted by UNHS in Australia—mild bilateral and unilateral losses.

4.1.1. Language

Across all age groups, overall language outcomes were on average a half to two thirds of a standard deviation poorer than population normative scores.

Children with mild bilateral hearing loss tended to demonstrate poorer language outcomes than those with unilateral loss or unilateral ANSD. This pattern of outcomes was particularly evident at the early life (2 years) and entry to primary school (5–7 years) timepoints.

For children with unilateral hearing loss, receptive vocabulary performance at entry to primary school appeared to be approximating population normative levels. However, receptive and expressive language outcome results tended to be poorer than population normative scores.

Children with unilateral ANSD, across early life and entry to primary school timepoints, demonstrated language performance comparable to children with unilateral profound sensorineural hearing loss. Average language outcome scores were around two thirds of a standard deviation poorer than population normative scores, with similar distributions of performance observed.

Interpreted cautiously due to low participant numbers, children at the transition to secondary school (9–12 year) timepoint were either in general at or within two thirds of a standard deviation below the population normative levels irrespective of degree or type of hearing loss.

4.1.2. Health related quality of life

Across all age groups, children had poorer psychosocial HRQoL scores compared to physical HRQoL scores. Distribution of individual scores appeared to follow the same pattern across all ages and degrees of loss, with most HRQoL scores within the upper quartile scores suggesting many of these children experience high quality of life.

4.2. Strengths of the study

A strength of this study is the population-level databank that was the source of participating children. Through this

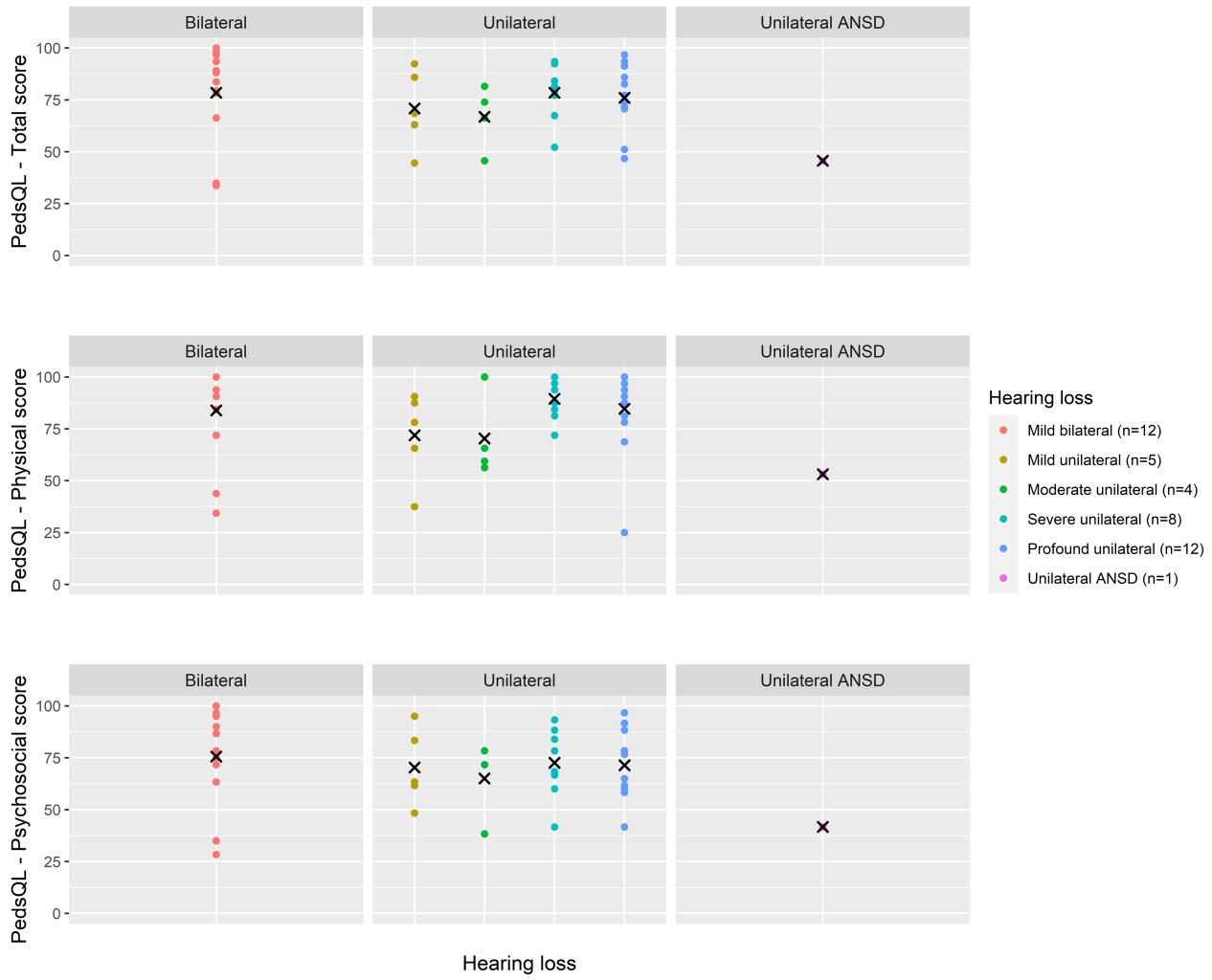

FIGURE 3
Language (CELF recalling sentences) and vocabulary (NPVT) scores for age 9–12 years across hearing loss groups.

databank we were able to confirm method of hearing loss identification (all detected via UNHS activities) and access outcomes on standardized measures. By using all available timepoints we have been able to maximize the number of results to report outcomes from a large group of children with non-target losses (including unilateral ANSD which has very sparse reporting of language outcomes) detected as by-products of UNHS activities. Our study also provides a description of outcomes at multiple timepoints across child development. This has resulted in a study of the contemporary population that reflects current detection trends (early) and availability of intervention—something that is to our knowledge not available in the extant literature.

Through our recruitment source, we have optimized the reported levels of diversity in participant characteristics that are comparable to the general population—such as levels of socio-economic disadvantage that reflect the expected levels in the Australian population. When compared to clinical samples of children with the same hearing diagnoses, we believe our results are representative of the wider population by documenting varied decisions taken by families around intervention and use of amplification.

4.3. Limitations

In reporting descriptive outcomes of children detected with unilateral or mild bilateral hearing loss under contemporary conditions, whilst we have achieved a large sample at 473 data points, we have not explored any causal relationships between degree and type of hearing loss and outcomes for these children. Our study design—drawing on available outcomes from the first decade of an established and growing databank that serves as a repository of outcomes—meant that we cannot yet comment on trajectories of performance across child development, but rather describe age-groups independently. We were also limited to using responses from those families who actively participate in the databank activities but note there were no significant differences in the characteristics of participant responders (Table 1) vs. non-responders (Supplementary Table S1).

TABLE 3 Summary of health-related quality of life outcomes for each age group.

		n	PedsQL Total score			PedsQL Physical score			PedsQL Psychosocial score		
			Median	IQR	95% CI	Median	IQR	95% CI	Median	IQR	95% CI
Age 2 years											
Overall		253	85.00	[69.52, 92.86]	(83.33, 86.67)	95.00	[70.00, 100.00]	(95.00, 100.00)	80.00	[70.00, 90.00]	(78.57, 82.50)
By hearing loss											
Bilateral:	Mild	92	82.50	[70.00, 91.41]	(79.27, 86.67)	95.00	[70.00, 100.00]	(90.00, 100.00)	80.00	[69.46, 89.29]	(75.00, 82.50)
Unilateral:	Mild	18	86.67	[65.83, 95.83]	(63.33, 96.67)	100.00	[80.00, 100.00]	(75.00, 100.0)	82.50	[60.63, 93.75]	(60.00, 95.00)
	Moderate	33	85.00	[68.33, 91.67]	(73.33, 90.48)	95.00	[60.00, 100.00]	(70.00, 100.00)	80.00	[70.00, 90.00]	(72.50, 87.50)
	Severe	35	88.1	[80.36, 94.17]	(81.67, 92.86)	100.00	[82.50, 100.00]	(90.00, 100.00)	85.00	[75.00, 92.50]	(77.50, 89.29)
	Profound	35	83.33	[55.00, 89.40]	(58.33, 88.33)	90.00	[45.00, 100.00]	(50.00, 100.00)	80.00	[63.39, 85.36]	(67.50, 85.00)
ANSD (unilateral)		40	87.38	[80.00, 93.33]	(83.33, 90.48)	100.00	[88.75, 100.00]	(90.00, 100.00)	82.14	[76.88, 90.00]	(78.57, 87.50)
Age 5–7 years											
Overall		137	76.67	[56.67, 86.67]	(73.33, 80.00)	95.00	[60.00, 100.00]	(90.00, 95.00)	70.00	[60.00, 82.50]	(65.00, 72.50)
By hearing loss											
Bilateral:	Mild	46	76.67	[54.17, 86.67]	(65.00, 85.00)	90.00	[51.25, 100.00]	(75.00, 100.00)	68.75	[60.00, 84.38]	(65.00, 80.00)
Unilateral:	Mild	12	71.67	[51.67, 80.42]	(41.67, 81.67)	92.50	[47.50, 100.00]	(40.00, 100.00)	65.00	[54.38, 72.50]	(45.00, 72.50)
	Moderate	19	75.00	[63.33, 85.83]	(61.67, 86.67)	90.00	[65.00, 97.50]	(65.00, 100.00)	65.00	[62.50, 81.25]	(62.50, 82.50)
	Severe	19	78.33	[50.83, 82.50]	(45.00, 83.33)	90.00	[45.00, 100.00]	(40.00, 100.00)	70.00	[58.75, 76.25]	(55.00, 72.50)
	Profound	28	75.00	[66.67, 85.83]	(71.67, 85.00)	95.00	[80.00, 100.00]	(85.00, 100.00)	68.75	[60.00, 85.00]	(60.00, 82.50)
ANSD (unilateral)		13	88.33	[73.33, 90.00]	(56.67, 91.67)	100.00	[90.00, 100.00]	(85.00, 100.00)	82.50	[65.00, 85.00]	(55.00, 87.50)
Age 9–12 years											
Overall		42	78.80	[66.58, 88.86]	[70.65, 84.09)	89.06	[71.88, 96.88]	(81.25, 93.75)	73.33	[61.67, 86.67]	(63.33, 78.33)
By hearing loss											
Bilateral:	Mild	12	85.57	[75.27, 94.29]	–	93.75	[81.25, 100.00]	–	82.50	[69.58, 91.25]	–
Unilateral:	Mild	5	68.48	[63.04, 85.87]	–	78.13	[65.63, 87.50]	–	63.33	[61.67, 83.33]	–
	Moderate	4	70.11	[61.14, 75.82]	–	62.50	[58.59, 74.22]	–	71.67	[63.33, 73.33]	–
	Severe	8	80.43	[74.73, 86.17]	–	90.63	[83.59, 97.66]	–	73.33	[65.00, 85.03]	–
	Profound	12	75.54	[70.65, 87.23]	–	92.19	[80.47, 96.88]	–	70.83	[61.25, 80.83]	–
ANSD (unilateral)		1	45.65	–	–	53.13	–	–	41.67	–	–

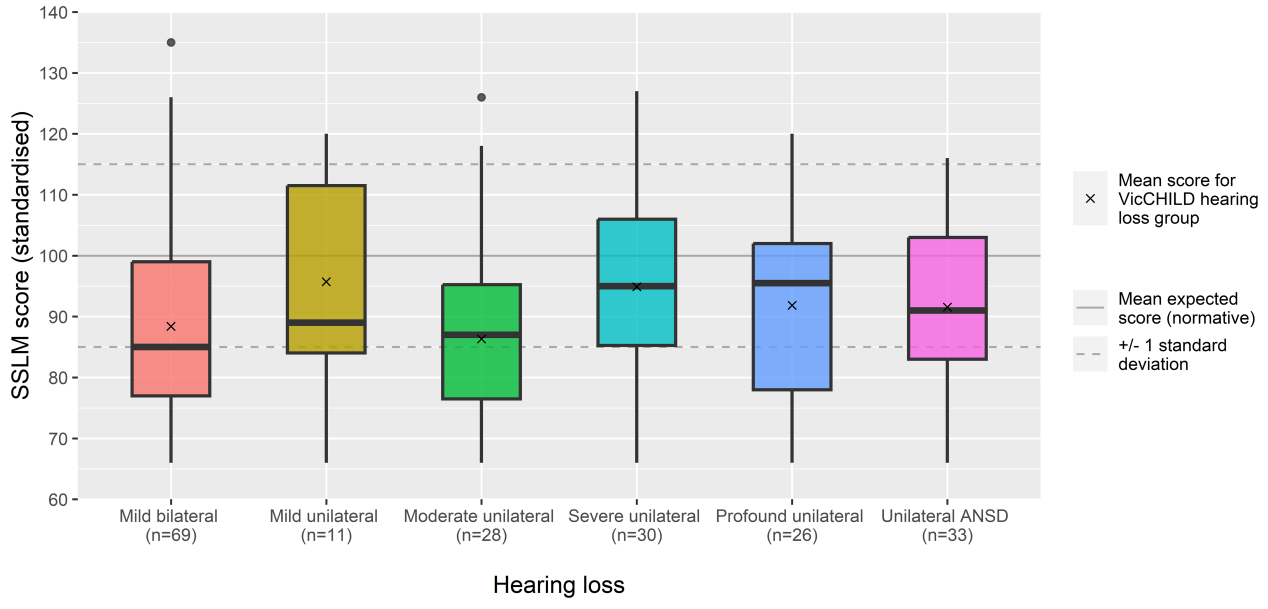

FIGURE 4
Health-related quality of life (PedsQL 4.0) scores for age 2 years across hearing loss groups.

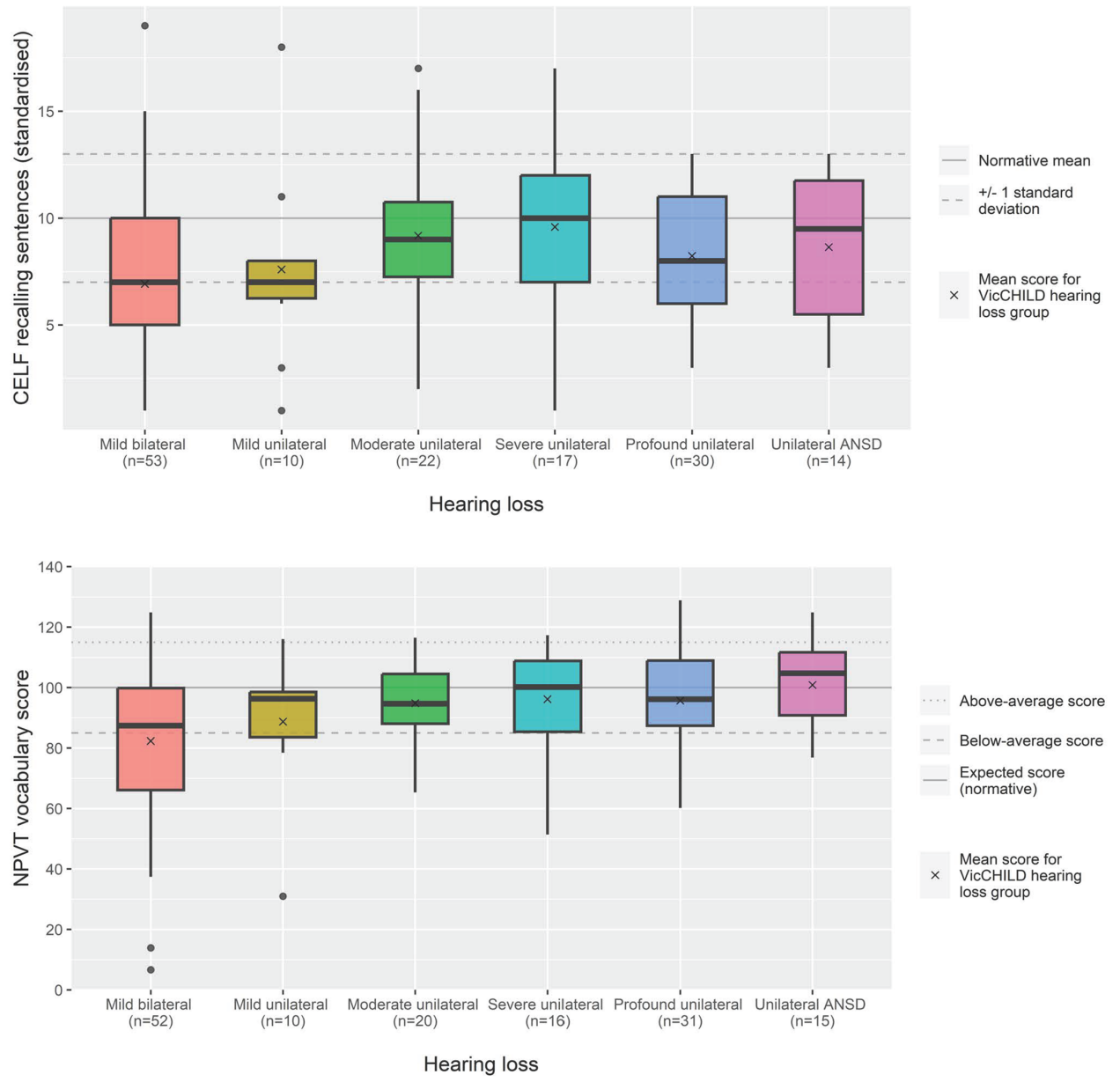

FIGURE 5
Health-related quality of life (PedsQL 4.0) scores for age 5–7 years across hearing loss groups.

The nature of the population databank—where different individual measures need to be as short as possible to reduce participant burden and encourage participant retention over time—precludes the ability to include outcome measures that may have been more sensitive to discrete groups of children with hearing loss. For example, whilst the PedsQL is validated for use in populations with chronic health conditions (32) and has been used in prior studies involving children with hearing loss (35, 36), it may not be as sensitive an instrument as alternate instruments such as the HEAR-QL, in demonstrating potentially more nuanced challenges faced by children with unilateral and mild bilateral hearing loss (37). Moreover, it is not unusual for large databanks that span many years to be challenged by missing data. For example, data about additional health needs or medical diagnoses were collected only after 2020 with a high proportion of missing data for this variable. The reported rate of additional health needs in our sample is higher than that reported in the existing literature (38). This may be because participant families reported against a more comprehensive list of medical diagnoses as compared with previous studies. The higher than expected proportion of children with additional health needs or medical diagnoses could theoretically affect the outcomes measured; however, we do not have complete data for this variable, and we suspect many of our families may have reported on medical diagnoses unrelated to the child's hearing or vocabulary outcomes.

Whilst the number of children we have included in this study is large in relation to many other studies of unilateral

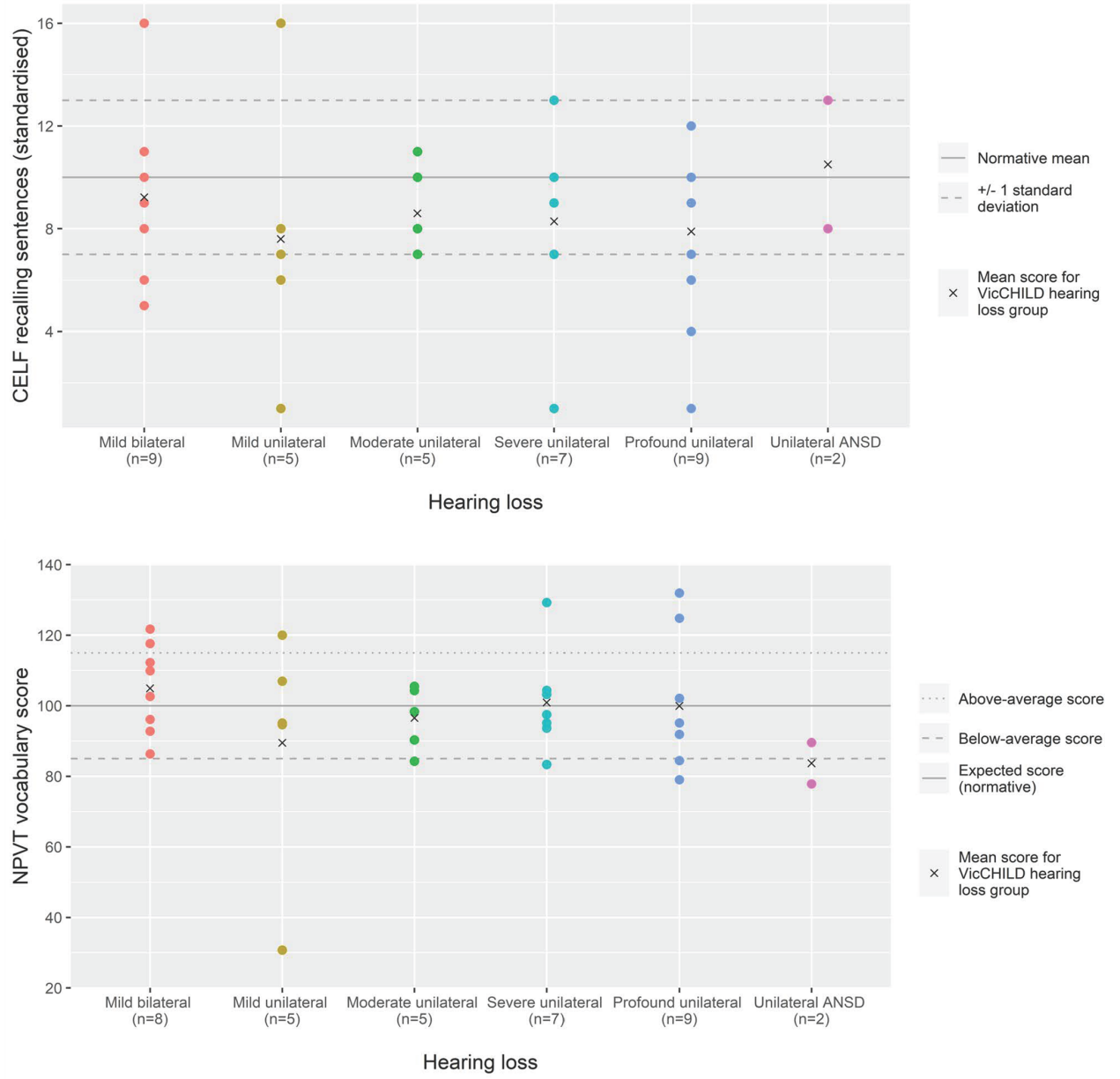

FIGURE 6
Health-related quality of life (PedsQL 4.0) scores for age 9–12 years across hearing loss groups.

and mild bilateral hearing loss, we occasionally interpreted all children's results in one combined group of "minimal" non-target hearing losses. This raises the critique of analyzing outcomes for two different types of hearing loss as one group. It is important to note that children with these hearing loss types are actually heterogenous groups that instead share some common challenges of hearing loss, such as uncertainties in early clinical management and possibly inconsistent early hearing device use (17), and low access to/engagement in early intervention services as demonstrated by our data (less than 50% ever accessed early intervention services). It is possible that the reasons for these challenges may differ between mild bilateral and unilateral losses (39), and it would be preferable to uniformly report their outcomes as discrete groups.

4.4. Interpretation in light of other studies

Our study is, in effect, an audit reporting language and HRQoL outcomes in a large group of children born with mild bilateral or unilateral hearing loss. Participants represent the diversity seen in the community with regards to decisions on amplification and intervention that is harder to achieve in clinical samples. Due to the duration of UNHS in Victoria and the size of the databank where participants were drawn from, our study is able to

describe outcomes across a larger sample of universally early-identified children than we are aware has been performed prior. Therefore, we believe this study represents a valuable addition to the literature on language and HRQoL outcomes that are seen in the contemporary hearing detection landscape where early detection is common and management decisions vary.

4.4.1. Mild bilateral and unilateral hearing loss

Our early life timepoint results demonstrated poorer expressive vocabulary performance than population norms, aligning with other reports of early life impact of unilateral hearing loss. In a UNHS detected sample with a median age 9.4 months, children with unilateral hearing loss were shown to demonstrate delays in auditory behaviour and preverbal vocalizations when compared to age-matched peers from the same population with normal hearing (40). However, not all reports agree, with another report of early detected children with unilateral and mild bilateral hearing losses showing language development meeting expectations through to four years of age (41). Of note, less than half of our sample of children engaged with early intervention services, possibly a reflection of the availability of these services to this non-target group of children, or low engagement due to perceptions these children may not require such services. With early detection of mild and unilateral losses now routine, it is important to reflect on whether this group of children have access to and are adequately supported to enroll in early intervention services.

Our entry to primary school timepoint demonstrated differences in performance across language and vocabulary outcome measures. This may have to do with task complexity, with our measure of receptive and expressive language (CELF Recalling Sentences) appearing more robust at highlighting performance differences compared to our receptive vocabulary (NPVT) assessment task. With receptive and expressive language requiring skills in morphological and phonological awareness, semantics, syntax and working memory, it may not be surprising that our children with mild bilateral or unilateral loss showed more variation in performance on this task—and lower achievement levels—when compared to the receptive vocabulary task that relies on semantics alone. Challenges in discrete areas of language may be supported by other findings, such as Nassrallah et al. (42) who reported findings of a descriptive study of children aged 5–9 years of age. They reported poorer than expected phonological processing skills, with 46% of children with mild bilateral or unilateral loss more than one standard deviation poorer than the expected level on a phonological memory task.

The results of this study support the conclusion that children with mild bilateral or unilateral hearing loss are at greater development risk (43) than their peers without hearing loss. The lower scores and large variation in scores on the caregiver-reported psychosocial HRQoL domain, as compared to the other HRQoL domains, may be a demonstration of this developmental risk. Such a result aligns with other reports of poorer quality of life in school and social domains for children with unilateral hearing loss (44). Uncertainty on appropriate management of hearing loss may also lead to this perception of development risk, with caregivers and audiologists recently reporting challenges in decision-making around best ways to support children born with mild bilateral hearing loss (16, 17).

4.4.2. Unilateral auditory neuropathy spectrum disorder

Our finding that, on average, children with unilateral ANSD demonstrated language performance similar to those with unilateral sensorineural profound losses is novel but not unexpected when considering what is known of outcomes for children with bilateral ANSD. Children with bilateral ANSD, typically in an early-identified setting and users of amplification (hearing aids or cochlear implants), have been shown to demonstrate early language abilities (up to 7 years of age) not significantly different to their peers with bilateral sensorineural loss (45, 46). In comparison, very little is documented on the outcomes of children with unilateral ANSD, likely a consequence of the rarity of this type of unilateral hearing loss (47). By including children with unilateral ANSD in our descriptive study, we are able to report on language outcomes in this under-described group.

5. Conclusions

Routine early identification of mild bilateral and unilateral hearing loss has driven recent focus on understanding outcomes for impacted children and their families. Whilst population-based studies, such as this one, describe the unadjusted development outcomes under contemporary detection methods, understanding the factors that mediate these outcomes is required to guide what steps will optimize appropriate support for these children. In particular, more attention needs to be paid in evaluating whether these children have access to or are adequately supported to enroll in early intervention programs, and whether early intervention programs for these children are effective. Similarly, future research needs to focus on the impact of early amplification as well as consistency in amplification use in these children on their language and quality of life outcomes. Efforts to harmonize outcome measures across databanks and projects focused on mild bilateral and unilateral hearing loss, such as the upcoming Australian National Child Hearing health Outcomes Registry (ANCHOR, NHMRC grant 2015735) should enable this transition from descriptive reports to more predictive analyses, particularly as the number of early detected children with these degrees of loss continues to grow.

Data availability statement

The raw data supporting the conclusions of this article will be made available by the authors, without undue reservation.

Ethics statement

The studies involving humans were approved by Royal Children's Hospital Ethics Review Board. The studies were conducted in accordance with the local legislation and institutional requirements. Written informed consent for participation in this study was provided by the participants' legal guardians/next of kin.

Author contributions

PC conceptualized and designed the study, contributed to data interpretation, drafted and revised the manuscript; he takes overall responsibility for all aspects of the study; DS conceptualized and designed the study, was responsible for the analysis and interpretation of the data, drafted and revised the manuscript; LS was responsible for acquisition, cleaning and preparation of the data and reviewed and revised the manuscript; QS assisted with designing the study and reviewed and revised the manuscript; VS conceptualized and designed the study, provided guidance regarding the data analysis and reviewed and revised the manuscript. All authors contributed to the article and approved the submitted version.

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Conflict of interest

The authors declare that the research was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

Publisher's note

Supplementary material

The Supplementary Material for this article can be found online at: https://www.frontiersin.org/articles/10.3389/fped.2023.1210282/full#supplementary-material

References

1. Ching TY, Oong R, van WE. The ages of intervention in regions with and without universal newborn hearing screening and prevalence of childhood hearing impairment in Australia. *Aust N Z J Audiol.* (2006) 28(2):137–50. doi: 10.1375/audi.28.2.137

2. Mauk GW, White KR, Mortensen LB, Behrens TR. The effectiveness of screening programs based on high-risk characteristics in early identification of hearing impairment. *Ear Hear.* (1991) 12(5):312. doi: 10.1097/00003446-199110000-00003

3. Ching TYC, Dillon H, Leigh G, Cupples L. Learning from the longitudinal outcomes of children with hearing impairment (LOCHI) study: summary of 5-year findings and implications. *Int J Audiol.* (2018) 57(sup2):S105–11. doi: 10.1080/14992027.2017.1385865

4. Wake M, Ching TY, Wirth K, Poulakis Z, Mensah FK, Gold L, et al. Population outcomes of three approaches to detection of congenital hearing loss. *Pediatrics.* (2016) 137(1):1–10. doi: 10.1542/peds.2015-1722

5. Korver AM, Konings S, Dekker FW, Beers M, Wever CC, Frijns JH, et al. Newborn hearing screening vs later hearing screening and developmental outcomes in children with permanent childhood hearing impairment. *JAMA.* (2010) 304(15):1701–8. doi: 10.1001/jama.2010.1501

6. Holstrum WJ, Biernath K, McKay S, Ross DS. Mild and unilateral hearing loss: implications for early intervention. *Infants Young Child.* (2009) 22(3):177–87. doi: 10.1097/01.IYC.0000357455.38816.b8

7. Pimperton H, Kennedy CR. The impact of early identification of permanent childhood hearing impairment on speech and language outcomes. *Arch Child*. (2012) 97(7):648–53. doi: 10.1136/archdischild-2011-301501

8. Hyde ML. Newborn hearing screening programs: overview. *J Otolaryngol*. (2005) 34(Suppl 2):S70–8.

9. Wood SA, Sutton GJ, Davis AC. Performance and characteristics of the newborn hearing screening programme in England: the first seven years. *Int J Audiol*. (2015) 54 (6):353–8. doi: 10.3109/14992027.2014.989548

10. Lieu JEC, Kenna M, Anne S, Davidson L. Hearing loss in children: a review. *JAMA*. (2020) 324(21):2195. doi: 10.1001/jama.2020.17647

11. Walker EA. Evidence-based practices and outcomes for children with mild and unilateral hearing loss. *Lang Speech Hear Serv Sch*. (2020) 51(1):1–4. doi: 10.1044/2019_LSHSS-19-00073

12. Aided Child Demographics. Hear Aust. (2020).

13. Grandpierre V, Fitzpatrick EM, Na E, Mendonca O. School-aged children with mild bilateral and unilateral hearing loss: parents' reflections on services, experiences, and outcomes. *J Deaf Stud Deaf Educ*. (2018) 23(2):140–7. doi: 10.1093/deafed/enx049

14. Brookhouser PE, Worthington DW, Kelly WJ. Unilateral hearing loss in children. *Laryngoscope*. (1991) 101(12):1264–72. doi: 10.1002/lary.5541011202

15. Fitzpatrick E, Grandpierre V, Durieux-Smith A, Gaboury I, Coyle D, Na E, et al. Children with mild bilateral and unilateral hearing loss: parents' reflections on experiences and outcomes. *J Deaf Stud Deaf Educ*. (2016) 21(1):34–43. doi: 10.1093/deafed/env047

16. Lin JJ, Gillam L, Smith L, Carew P, King A, Ching TYC, et al. Mild matters: parental insights into the conundrums of managing mild congenital hearing loss. *Int J Audiol*. (2021) 0(0):1–7. doi: 10.1080/14992027.2021.1954248

17. Ching TYC, Saetre-Turner M, Marnane V, Scarinci N, Choik C, Tulloch K, et al. Audiologists' perspectives on management of mild bilateral hearing loss in infants and young children. *Int J Audiol*. (2022) 61(9):752–60. doi: 10.1080/14992027.2021.1961170

18. King AM. The national protocol for paediatric amplification in Australia. *Int J Audiol*. (2010) 49(Suppl 1):S64–9. doi: 10.3109/14992020903329422

19. Fitzpatrick EM, Cologrosso E, Sikora L. Candidacy for amplification in children with hearing loss: a review of guidelines and recommendations. *Am J Audiol*. (2019) 28(4):1025–45. doi: 10.1044/2019_AJA-19-0061

20. Song M, Li J, Lan L, Xie L, Xiong F, Yu L, et al. Clinical characteristics of patients with unilateral auditory neuropathy. *Am J Otolaryngol*. (2021) 42(5):103143. doi: 10.1016/j.amjoto.2021.103143

21. Uus K, Young A, Day M. Parents' perspectives on the dilemmas with intervention for infants with auditory neuropathy spectrum disorder: a qualitative study. *Int J Audiol*. (2015) 54(8):552–8. doi: 10.3109/14992027.2015.1020970

22. Barreira-Nielsen C, Fitzpatrick EM, Whittingham J. Characteristics and outcomes of children with auditory neuropathy Spectrum disorder. *Can J Speech-Lang Pathol Audiol*. (2016) 40(1):67–79.

23. Buchman CA, Roush PA, Teagle HFB, Brown CJ, Zdanski CJ, Grose JH. Auditory neuropathy characteristics in children with cochlear nerve deficiency. *Ear Hear*. (2006) 27(4):399–408. doi: 10.1097/01.aud.0000224100.30525.ab

24. Sung V, Smith L, Poulakis Z, Burt RA, Carew P, Tobin S, et al. Data resource profile: the victorian childhood hearing impairment longitudinal databank (VicCHILD). *Int J Epidemiol*. (2019) 48(5):1409–1409. doi: 10.1093/ije/dyz168

25. Carew P, Mensah FK, Rance G, Flynn T, Poulakis Z, Wake M. Mild-moderate congenital hearing loss: secular trends in outcomes across four systems of detection. *Child Care Health Dev*. (2018) 44(1):71–82. doi: 10.1111/cch.12477

26. Roy P, Kersley H, Law J. *The sure start language measure standardisation study*. London: City University (2005).

27. Dale PS, Dionne G, Eley TC, Plomin R. Lexical and grammatical development: a behavioural genetic perspective. *J Child Lang*. (2000) 27(3):619–42. doi: 10.1017/S0305000900004281

28. Semel E, Wiig EH, Secord WA. *Clinical evaluation of language fundamentals-fourth edition, Australian standardised edition*. Sydney: NSW: Psychological Corporation (2006).

29. Smith J, Wang J, Grobler AC, Lange K, Clifford SA, Wake M. Hearing, speech reception, vocabulary and language: population epidemiology and concordance in Australian children aged 11 to 12 years and their parents. *BMJ Open*. (2019) 9 (Suppl 3):88. doi: 10.1136/bmjopen-2018-023196

30. Clifford S, Davies S, Gillespie A, Lange K, Liu M, Wang Y, et al. Longitudinal Study of Australian Children's Child Health CheckPoint Data User Guide—June 2021. (2021). Available at: https://mcri.figshare.com/articles/journal_contribution/Longitudinal_Study_of_Australian_Children_s_Child_Health_CheckPoint_Data_User_Guide_December_2018/5687590/3 (Cited April 22, 2023).

31. Varni JW, Seid M, Kurtin PS. PedsQL 4.0: reliability and validity of the pediatric quality of life inventory version 4.0 generic core scales in healthy and patient populations. *Med Care*. (2001) 39(8):800–12. doi: 10.1097/00005650-200108000-00006

32. Varni JW, Limbers CA, Burwinkle TM. Parent proxy-report of their children's health-related quality of life: an analysis of 13,878 parents' reliability and validity across age subgroups using the PedsQL 4.0 generic core scales. *Health Qual Life Outcomes*. (2007) 5:2. doi: 10.1186/1477-7525-5-2

33. Rose E, Sung V. Otolaryngology. In: Harding K, Mason DS, Efron D, editors. *Paediatric handbook*. 10th ed. Newark, United Kingdom: John Wiley & Sons, Incorporated (2021):461. Available at: http://ebookcentral.proquest.com/lib/unimelb/detail.action?docID=6375231 (Cited August 3, 2022).

34. R Core Team. R: A language and environment for statistical computing. (2021). Available at: https://www.r-project.org/ (Cited April 21, 2023).

35. Wake M, Tobin S, Cone-Wesson B, Dahl HH, Gillam L, McCormick L, et al. Slight/mild sensorineural hearing loss in children. *Pediatrics*. (2006) 118(5):1842–51. doi: 10.1542/peds.2005-3168

36. Ching TYC, Dillon H, Button L, Seeto M, Van Buynder P, Marnane V, et al. Age at intervention for permanent hearing loss and 5-year language outcomes. *Pediatrics*. (2017) 140(3):4. doi: 10.1542/peds.2016-4274

37. Lieu JEC. Permanent unilateral hearing loss (UHL) and childhood development. *Curr Otorhinolaryngol Rep*. (2018) 6(1):74–81. doi: 10.1007/s40136-018-0185-5

38. Cupples L, Ching TY, Crowe K, Seeto M, Leigh G, Street L, et al. Outcomes of 3-year-old children with hearing loss and different types of additional disabilities. *J Deaf Stud Deaf Educ*. (2014) 19(1):20–39. doi: 10.1093/deafed/ent039

39. Lewis DE. Speech understanding in complex environments by school-age children with mild bilateral or unilateral hearing loss. *Semin Hear*. (2023) 44(Suppl 1):S36–48. doi: 10.1055/s-0043-1764134

40. Kishon-Rabin L, Kuint J, Hildesheimer M, Ari-Even Roth D. Delay in auditory behaviour and preverbal vocalization in infants with unilateral hearing loss. *Dev Med Child Neurol*. (2015) 57(12):1129–36. doi: 10.1111/dmcn.12812

41. Fitzpatrick EM, Durieux-Smith A, Gaboury I, Coyle D, Whittingham J. Communication development in early- identified children with mild bilateral and unilateral hearing loss. *Am J Audiol*. (2015) 24(3):349–53. doi: 10.1044/2015_AJA-15-0003

42. Nassrallah F, Fitzpatrick EM, Whittingham J, Sun H, Na E, Grandpierre V. A descriptive study of language and literacy skills of early school-aged children with unilateral and mild to moderate bilateral hearing loss. *Deaf Educ Int*. (2020) 22 (1):74–92. doi: 10.1080/14643154.2018.1555119

43. Walker EA, Spratford M, Ambrose SE, Holte L, Oleson J. Service delivery to children with mild hearing loss: current practice patterns and parent perceptions. *Am J Audiol*. (2017) 26(1):38–52. doi: 10.1044/2016_AJA-16-0063

44. Roland L, Fischer C, Tran K, Rachakonda T, Kallogjeri D, Lieu JEC. Quality of life in children with hearing impairment: systematic review and meta-analysis. *Otolaryngol Head Neck Surg*. (2016) 155(2):208–19. doi: 10.1177/0194599816640485

45. Ching TYC, Day J, Dillon H, Gardner-Berry K, Hou S, Seeto M, et al. Impact of the presence of auditory neuropathy spectrum disorder (ANSD) on outcomes of children at three years of age. *Int J Audiol*. (2013) 52:S55–64. doi: 10.3109/14992027.2013.796532

46. Walker E, McCreery R, Spratford M, Roush P. Children with auditory neuropathy Spectrum disorder fitted with hearing aids applying the American academy of audiology pediatric amplification guideline: current practice and outcomes. *J Am Acad Audiol*. (2016) 27(3):204–18. doi: 10.3766/jaaa.15050

47. van Wieringen A, Boudewyns A, Sangen A, Wouters J, Desloovere C. Unilateral congenital hearing loss in children: challenges and potentials. *Hear Res*. (2019) 372:29–41. doi: 10.1016/j.heares.2018.01.010

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EDITED BY
Zhengqing Hu,
Wayne State University, United States

REVIEWED BY
Johannes Fellinger,
Hospitaller Brothers of Saint John of God Linz, Austria
Sally Kay Thornton,
University of Nottingham, United Kingdom

*CORRESPONDENCE
Valerie Sung
✉ valerie.sung@rch.org.au

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Emotional behavioral outcomes of children with unilateral and mild hearing loss

Jun Jean Ong[1,2,3], Libby Smith[1,2], Daisy A. Shepherd[2,4], Jessica Xu[1,2], Gehan Roberts[1,2,4] and Valerie Sung[1,2,4]*

[1]Centre for Community Child Health, The Royal Children's Hospital, Melbourne, VIC, Australia, [2]Murdoch Children's Research Institute, Melbourne, VIC, Australia, [3]Department of Paediatrics, School of Medicine, International Medical University, Kuala Lumpur, Malaysia, [4]Department of Paediatrics, The University of Melbourne, Melbourne, VIC, Australia

Background: Deaf and hard-of hearing (DHH) children often experience emotional/behavioral difficulties. The impact of unilateral/mild hearing loss (HL) on children's emotion and behavior are unclear. We aimed to describe emotional/behavioral, health related quality-of-life (HRQoL) and parent psychological distress outcomes of school-age children with unilateral/mild HL, compared to children with moderate to profound HL, and in relation to population norms; and identify predictive factors of emotional/behavioral difficulties.

Methods: Data of 339 DHH children, 5–12 years, enrolled in the Victorian Childhood Hearing Longitudinal Databank (VicCHILD), which include demographics, early development, medical/audiological characteristics and parent rated questionnaires of emotion/behavior, HRQoL and parental psychological distress collected at various stages of child's life were analyzed. We used Cohen's d to investigate the outcomes by measuring the mean score differences of both groups with published norms and logistic regression to analyze the factors predictive of emotional/behavioral difficulties.

Results: The proportion of children with unilateral/mild HL and moderate to profound HL who experienced emotional/behavioral difficulties was similar (18.3% vs. 20.6%), with hyperactivity and poor prosocial behavior reported as the predominant symptoms in both groups. Mean emotional/behavioral scores of both groups were comparable and substantially higher than normative population scores. This was also the case for HRQoL and levels of parent distress. Among children with unilateral/mild HL, additional health needs were the strongest predictive factor, demonstrating an approximately 1.7-fold increase in odds of emotional/behavioral difficulties (OR = 1.67; 95% CI 1.29–2.17, $p < 0.001$) with every additional health need. Early developmental concerns, other than communication milestone and attending mainstream schoolshowed weaker evidence of association.

Conclusion: Children with unilateral/mild HL were just as likely as those with moderate to profound HL to experience more emotional/behavioral difficulties, poorer HRQoL and higher parental distress scores compared to population norms. Our findings justify the provision of early intervention, support and medical services for all DHH children to identify those at risk of poorer outcomes.

KEYWORDS

unilateral, mild, hearing loss, emotional behavioral difficulties, deaf or hard of hearing, children

Introduction

Deaf and hard-of hearing (DHH) children often experience emotional and behavioral difficulties (1, 2). Previous studies, which mainly include children with moderate to profound hearing loss (HL) have reported high prevalence of externalizing and internalizing behavioral symptoms compared to normal hearing population (2). In a review by Stevenson et al. (2015), peer problems were rated by both parents and teachers as the predominant emotional/behavioral symptoms, whereas a recent longitudinal study showed that hyperactivity/inattention symptoms were most reported by parents and low prosocial behavior by teachers (2, 3). Deficits in socio-emotional regulation due to delayed executive function and social cognitive development are hypothesized to be reasons DHH children are more vulnerable to emotional/behavioral problems (4). Poor social skills and low prosocial behavior are equally reported despite improved language development due to persistent pragmatic developmental challenges (5). However, more recent studies have described minimal differences in emotional/behavioral mean scores among DHH children compared to normative data (2, 6). The narrowing of the gap in emotional/behavioral outcomes are likely contributed by the implementation of universal newborn hearing screening (UNHS) and the advancement of audiological intervention, speech-language therapy and counselling services in the past decade. Nevertheless, emotional/behavioral outcomes remain highly variable with a large proportion of DHH children continuing to experience difficulties despite earlier access to intervention and improvement of language and academic performances (7). Ongoing research to investigate possible contributing factors is much needed.

Language and communication development are among the most well-studied predictors of emotional/behavioral outcomes in DHH children (6, 8, 9). The severity of emotional/behavioral problems are influenced by the level of language abilities, with good receptive language and communication skills associated with lower risk of emotional/behavioral difficulties (8, 9). Aside from language abilities, additional health needs and nonverbal cognitive ability were significant factors identified among DHH children at 3 and 5 years of age (6, 10). Given that approximately two thirds of DHH children are reported to have an additional disability that could impact their education or development, Wiley et al. (2011) proposed the need for interdisciplinary medical evaluation for all DHH children (11). Studies have also shown that early detection of hearing loss and early access to intervention were associated with favorable academic and language performance (2, 8). However, degree/laterality of HL have not been shown to influence outcomes in several studies, with Wake et al. (2004) reporting that DHH children have poorer psychosocial, quality of life and language outcomes, irrespective of the severity of HL (1, 7, 12). In addition, Carew et al. (2018) reported poorer expressive language skills in children with mild HL compared to population means, despite early detection through the well-established UNHS (13).

Increasing detection of children with milder HL through UNHS and the awareness of their challenges have led to a rise in research interest exploring outcomes of unilateral/mild HL (1, 14, 15). Studies suggest children with unilateral/mild HL score lower in academic tests, are more likely to fail at least one grade and are delayed in various developmental abilities compared to hearing peers (16–18). They experience more emotional/behavioral difficulties than their peers with one fifth of children with unilateral HL reported by their teachers to have behavioral problems and requiring classroom accommodations (16, 17). Studies of children with unilateral/mild HL published after the implementation of UNHS continue to report high socio-emotional problems despite early diagnosis (13). A 3-year follow-up longitudinal study of children with unilateral HL described high prevalence of behavioral problems that improved with intervention; however, 10% or more continue to have problems with inattention, externalizing and internalizing symptoms (19). Porter et al. (2013) showed that differences in academic performance between unilateral/mild HL and hearing children were not apparent, but greater attention difficulties in the classroom were identified among children with unilateral/mild HL (20). Le Clercq et al. (2020) further emphasized the association between emotional/behavioral outcomes and hearing threshold, with higher inattention and social problems among children with slight to mild hearing loss (21). However, the effects of unilateral/mild HL on emotional/behavioral outcomes remain inconclusive as available studies also showed contradicting results, reporting no additional behavior problems compared to typical hearing children (22, 23).

As not all DHH children experience emotional/behavioral difficulties, recognition of predictive factors is imperative to identify those at high risk of poorer outcome. Factors associated with emotional/behavioral outcomes among children with moderate-profound bilateral HL are well explored, but likely differ from children with unilateral/mild HL due to differences in their experiences and access to sound. Possible factors such as lower maternal education, later age of amplification and intervention were suggested to be associated with poorer outcomes among children with unilateral/mild HL (19, 20, 24). However, evidence is scarce and the additional benefits of audiological intervention on emotional/behavioral outcomes among children with unilateral/mild HL remain uncertain.

The study analyzed data of families enrolled during the first 10 years of the Victorian Childhood Hearing Longitudinal Databank (VicCHILD) which recruited children with permanent HL of any degree and laterality (25). Information from a considerably large sample of children with unilateral/mild HL were able to be included in the study to address the following research questions:

1. What are the emotional/behavioral, health related quality-of-life (HRQOL) and parent psychological distress outcomes of children with unilateral/mild HL compared to children with moderate to profound HL and the normal hearing population?
2. Which factors are predictive of emotional/behavioral difficulties among children with unilateral/mild HL?

Material and methods

Participants

Our cross-sectional study included DHH children enrolled during the first 10 years of VicCHILD (between 2012 and 2022), and whose families completed a survey including emotional/behavioral outcomes at age 5–12 years. VicCHILD is a statewide population-based longitudinal databank open to every child with permanent hearing loss in Victoria, Australia (25). The majority of families are recruited through the statewide UNHS program, the Victorian Infant Hearing Screening Program (VIHSP) which screens more than 99% of newborns in Victoria. Families of DDH children attending the Royal Children's Hospital Caring for Hearing in Children Clinic are also invited to participate. Information and assessments are collected at enrolment and various stages of the child's life course, with the details of methodology described elsewhere (25). Recruitment and assessments at different age timepoints are still ongoing. This study described data on emotional/behavioral outcomes, assessed during primary school-age (5–12 years) and related information collected at different stages. The study has ethics approval from the Royal Children's Hospital Human Research Ethics Committee (approval number 31081).

Procedure

Parents provided sociodemographic, audiological and medical information about their child during enrolment and subsequent stages of assessment. Audiological data from the time of diagnosis was provided by the VIHSP. Where possible, updated audiological data was obtained at the time emotional/behavioral outcomes were collected, either from Hearing Australia, a service provider throughout Australia tasked with providing monitoring and rehabilitation services for deaf/hard of hearing children, or from the Caring for Hearing in Children Clinic, a pediatric service based at the Royal Children's Hospital. Audiological data included information about the child's type of HL, degree/laterality of HL, use of hearing devices at the time of assessment (or unaided) and age of first fitting, where available. Degree of hearing loss was classified using decibel ranges used by the national provider of hearing amplification, Hearing Australia (26): mild (21–40 dB), moderate (41–60 dB), severe (61–90 dB) and profound (>90 dB). We grouped the children according to the degree/laterality of HL: unilateral/mild HL vs. moderate-profound HL. Children with unilateral HL have mild to profound HL in one ear (≥20 dB) and normal hearing in the contralateral ear (<20 dB). Children with mild HL have mild HL (21–40 dB) in the better hearing ear. Children with moderate-profound HL have at least moderate HL (≥ 40 dB) in the better ear.

We collected data of children's developmental profile and additional health needs during enrolment. We measured early developmental profile using the Ages and Stages Questionnaire (ASQ),a brief parental questionnaire of a child's current skills and development from 1 to 66 months of age (27). Parents answered 6 questions "yes", "sometimes" or "not yet" in each of 5 domains of development: communication, gross motor, fine motor, problem-solving skills, and personal social skills based on what their child is able to do. Each answer was scored and the sum scores for each domain were calculated. Sum scores below cut-off, defined as 2 standard deviations (SD) below the mean was considered a positive early developmental concern. We only included early developmental profile assessed during the first 36 months of age for analysis. Information of additional health needs were based on parents' selection from a comprehensive list of health conditions, comprising of conditions related to neurodevelopmental, genetic and neurological disorders, malignancy, allergy, visual impairment and other chronic disorders.

We later collected outcome measures on child's emotion/behavior, quality of life and parental well-being during early school years, assessed using standardized parent rated questionnaires. The types of information and the timepoints at which data were collected from each participant child and family are described in detail elsewhere (25).

Outcome measures

Emotional behavioral outcome

The Strengths and Difficulties Questionnaire (SDQ) is a 25-item parent-rated screening measure designed to identify emotional/behavioral difficulties in children (28). The instrument comprises of five subscales: conduct problems, hyperactivity, emotional symptoms, peer problems and prosocial behavior with each subscale containing 5 items. Each item is rated on a 3-point response scale from 0= "not true" to 1= "somewhat true" and 2= "certainly true". Higher total scores for the first four subscales and "total difficulties" score which is the sum of the first four subscales (excluding prosocial behavior) reflect difficulties, while higher scores for the prosocial subscale reflect strength. The cut-off scores for "abnormal" category corresponds to the 90th (10th for prosocial subscale) percentiles, therefore total difficulties scores falling in the top 10% of the normative distribution is indicative of clinically significant emotional/behavioral difficulties (see **Supplementary Table S2**). Different cut-off scores and mean scores from Australian normative data are available for children 4–6 years and 7–12 years (29, 30).

Health related quality of life outcome (HRQoL)

The Pediatric Quality of Life Inventory version 4.0 (Peds-QL), parent completed version was used to measure HRQoL of the child (31). The inventory comprises 23 items from four domains: physical health, emotional functioning, social functioning and school functioning and each item is rated on a 5-point Likert scale. Items are scored from 0 to 100, with higher scores indicating better HRQoL. The total scores are the mean score of the sum of all domains and the psychosocial mean score represents sum of emotional, social, and school functioning domains.

Parent psychological distress outcome

The Kessler Psychological Distress Scale (K6) is a 6-item self-report measure of psychological distress for adults (32). Parents

indicate how often they experienced feeling sad, nervous, hopeless, restless, that everything was an effort and worthless during the past 30 days, using a 5-point Likert scale. Scores above clinical cut-off point indicate significant psychological distress.

Statistical analysis

The data was analyzed using Microsoft Excel and SPSS Statistic Package 26. We summarized participant characteristics for each DHH groups using means [with standard deviations (SD)] for continuous variables, medians [with interquartile ranges (IQR)] if not normally distributed and counts (with proportions) for categorical variables. Normality analysis showed that data of outcomes measures were slightly skewed and not normally distributed. For the first research question, we calculated the standardized mean difference using Cohen's d effect size to compare emotional/behavioral outcomes of both groups with published Australian norms according to age; 4–6 years and 7–17 years. Although outcomes were not normally distributed, mean scores were used for analysis as available normative population data for comparison were described in mean (SD). Same method of statistical analysis was used to compare HRQoL and parent distress outcomes in both groups compared to population norms. The outcomes (means and SD) of both DHH groups were also described and compared. Spearman's rank correlation was used to estimate correlations between continuous outcomes to describe the general observed patterns in our sample. For the second research question, we used univariable logistic regression to estimate associations between key predictors (separately) with emotional/behavioral difficulties among children with unilateral/mild HL. These predictors include categorical factors (gender, age groups (5–6 and 7–12years), hearing laterality, type of HL, unaided or aided hearing, using speech or other communication mode, attending mainstream or special/other schools and presence of early developmental concerns) and continuous factors (additional health needs, hearing devices first fitting age). Variables with multiple categories (communication mode, type of HL and school) were dichotomized for analysis due to the small numbers in several subgroups.

Results

Between 2012 and 2022, a total of 1202 DHH children were enrolled in VicCHILD. Of these, 834 DHH children had turned age 5–12 years old at the time of data analysis in late 2022, of which 339 families had completed the survey that included the SDQ as a measure of emotion/behavior. Of those who completed the SDQ (339 families), 186 families had completed the ASQ at 36 months or younger; of these 100 had unilateral/mild HLand 86 had moderate-profound HL, 337 completed the Peds-QL and 246 completed the K6. **Table 1** shows the participant demographic, audiological and medical characteristics. The study sample included a total of 339 children aged 5–12 years old, consisting of 169 children with unilateral/mild HL (49.9%) and 170 children with moderate-profound HL (50.1%). The characteristics of non-participants are summarized in **Supplementary Table S1**. Non-participating families were from slightly more disadvantaged areas and less likely to report use of English as a primary language at home compared to study participants. Otherwise, the groups were similar in demographic and audiological details, including maternal education and degree of HL.

Compared to children with moderate to profound HL, children with unilateral/mild HL were first fitted with hearing aid at an older age (median (IQR) 21.0 (43.0) months vs. 4.1 (9.0) months) with fewer children fitted before 36 months of age (68.6% vs. 94.0%). A higher proportion of children with unilateral/mild HL were also unaided with hearing devices (39.1% vs. 6.5%), using speech as main communication mode (84.6% vs. 67.1%) and attending mainstream school (82.2% vs. 69.4%). However, around half of parents of children with moderate to profound HL reported early communication developmental concerns, double in proportion compared to children with unilateral/mild HL. Approximately one fourth of parents with moderate-profound HL children also reported early developmental concerns in all domains aside from communication milestone, higher than children with unilateral/mild HL (**Table 1**).

Aim 1: outcomes of children with unilateral/mild HL compared to children with moderate-profound HLand population norms

Figure 1 shows the prevalence of emotional/behavioral difficulties of children with unilateral/mild HL and moderate to profound HL in our sample. A similar proportion of children from both groups experienced emotional/behavioral difficulties (18.3% vs. 20.6%), with hyperactivity and poor prosocial behavior, the most frequent symptoms reported. More children with unilateral/mild HL reported emotional symptoms (19.5% vs. 14.7%) while children with moderate to profound HL more often reported peer problems (14.8% vs. 21.8%).

In **Figure 2**, we demonstrate emotional/behavioral difficulties, total and the subscales mean score differences of both DHH groups, according to age groups. Emotional/behavioral difficulties total scores of children with unilateral/mild HL and moderate to profound HL were comparable (mean 9.8, SD 6.4 and mean 10.5, SD 6.5). However, younger children 5–6 years experienced more emotional/behavioral difficulties, with total scores within the top 20% (borderline) range (mean 10.1, SD 6.2 for unilateral/mild HL and mean 11.4, SD 6.9 for moderate-profound HL), while scores for children 7–12 years in both groups were within normal range (mean 9.6, SD 6.5 and mean 10.2, SD 6.3), for both DHH groups.

Compared to population normative scores, children with unilateral/mild HL and moderate to profound HL were reported to have higher emotional/behavioral difficulties scores for both age groups. Among all the subscales, hyperactivity symptoms had

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TABLE 1 Participant characteristics broken down by hearing loss group.

	Unilateral/Mild HL	Moderate-profound HL
	N = 169	*N* = 170
Child Characteristics		
Age at diagnosis of HL (months) - mean (SD)	2.1 (4.6)	2.6 (8.6)
Age at SDQ completion (years) - mean (SD)	8.0 (2.2)	8.4 (2.3)
Age group at SDQ completion (years)—*n* (%)		
5–6 years	52 (30.8)	42 (24.7)
7–12 years	117 (69.2)	128 (75.3)
Gender, male—*n* (%)	90 (53.3)	93 (54.7)
Family characteristics; *n* (%)		
Maternal education		
Year 11 or less	10 (5.9)	20 (11.8)
Year 12	51 (30.2)	41 (24.1)
Tertiary or postgraduate	71 (42.0)	58 (34.1)
Unreported	37 (21.9)	51 (30.0)
SEIFA disadvantage index[a]- mean (SD)	1,013.9 (71.1)	1,012.8 (65.3)
Family history of HL	17 (10.1)	10 (5.9)
English as primary language at home	138 (81.7)	126 (74.1)
Audiological/Medical characteristics, *n* (%)		
Degree of HL		
Unilateral-mild[b]	14 (8.3)	
Unilateral-moderate[b]	19 (11.2)	
Unilateral-severe[b]	16 (9.5)	
Unilateral-profound[b]	37 (21.9)	
Unavailable	10 (5.9)	
Bilateral mild[c]	73 (43.2)	
Bilateral-moderate[c]		63 (37.1)
Bilateral-severe[c]		32 (18.8)
Bilateral-profound[c]		49 (28.8)
Unavailable		26 (15.3)
Hearing device		
Unaided	66 (39.1)	11 (6.5)
Hearing aid/s only	66 (39.1)	57 (33.5)
Cochlear implant	6 (3.5)	80 (47.1)
Unreported	31 (18.3)	22 (12.9)
Age hearing aid first fitting		
Median (IQR), months	21.0 (43.0)	4.1 (9.0)
≤36 months, *n* (%)[d]	70 (68.6)	141 (94.0)
Age of cochlear implantation		
Median (IQR), months	35.5 (32.5)	18.0 (21.0)
≤36 months, *n* (%)[d]	3 (50.0)	62 (77.5)
Hearing loss types		
Sensorineural	123 (72.8)	130 (76.5)
Auditory neuropathy	7 (4.2)	17 (10.0)
Mixed	21 (12.4)	20 (11.7)
Permanent conductive	10 (5.9)	1 (0.6)
Atresia	8 (4.7)	2 (1.2)
Communication mode		
Speech only	143 (84.6)	114 (67.1)
Sign language (Auslan) only	0	6 (3.5)
Simultaneous sign and speech	3 (1.8)	13 (7.6)
Non-verbal/Key word signing/ gestures.	1 (0.6)	10 (5.9)
Unreported	22 (13.0)	27 (15.9)
School		
Mainstream ± special unit	139 (82.2)	118 (69.4)
School for DHH	2 (1.2)	15 (8.8)
Special school for children with disabilities	5 (3.0)	20 (11.8)
Others	5 (3.0)	3 (1.8)
Unreported	18 (10.6)	14 (8.2)
Additional health needs	131 (77.5)	124 (72.9)
Early developmental concerns ≤36 months[d]		
Communication	27 (27.0)	47 (54.7)
Gross motor	16 (16.0)	23 (26.7)
Fine motor	13 (13.0)	24 (27.9)
Problem solving	14 (14.0)	25 (29.1)
Social/Adaptive skill	13 (13.0)	21 (24.4)

[a]Socio-Economic Indexes for Areas (SEIFA) Index of Relative Socio-Economic Disadvantage (national mean 1,000, SD 100, with higher values representing less disadvantage).
[b]Degree of HL in the worse ear.
[c]Degree of HL in the better hearing ear.
[d]Total number may vary from total participants.

the highest mean scores differences compared to population normative scores, for both age and DHH groups. Table 2 estimates the standardized mean difference in HRQoL and parent distress outcome scores of children with unilateral/mild HL and moderate to profound HL compared to norms. Parents of DHH children from both groups reported poorer HRQoL for their child and higher parent distress scores compared to normative population scores (Table 2). HRQoL total and parent distress mean scores between both DHH groups were similar (74.1 (SD 18.4) vs. 72.8 (SD 17.9) and 15.5 (SD 3.6) vs. 15.0 (SD 3.7)). Poorer HRQoL in all domains and increased parent distress were also correlated with greater emotional/behavioral difficulties, of moderate effect size (r) ranging from 0.22–0.71 ($p < 0.001$), indicating that children with emotional/behavioral difficulties tended to have poorer HRQoL and lived with a parent with high levels of psychological distress.

Aim 2: factors associated with emotional/behavioral outcomes in children with unilateral/mild HL

Table 3 illustrates the association between each potential factor and emotional/behavioral difficulties (as measured by the SDQ scores; dichotomized) in children with unilateral/mild HL. The strongest evidence of association were additional health needs, demonstrating an approximately 1.7-fold increase in odds of emotional/behavioral difficulties

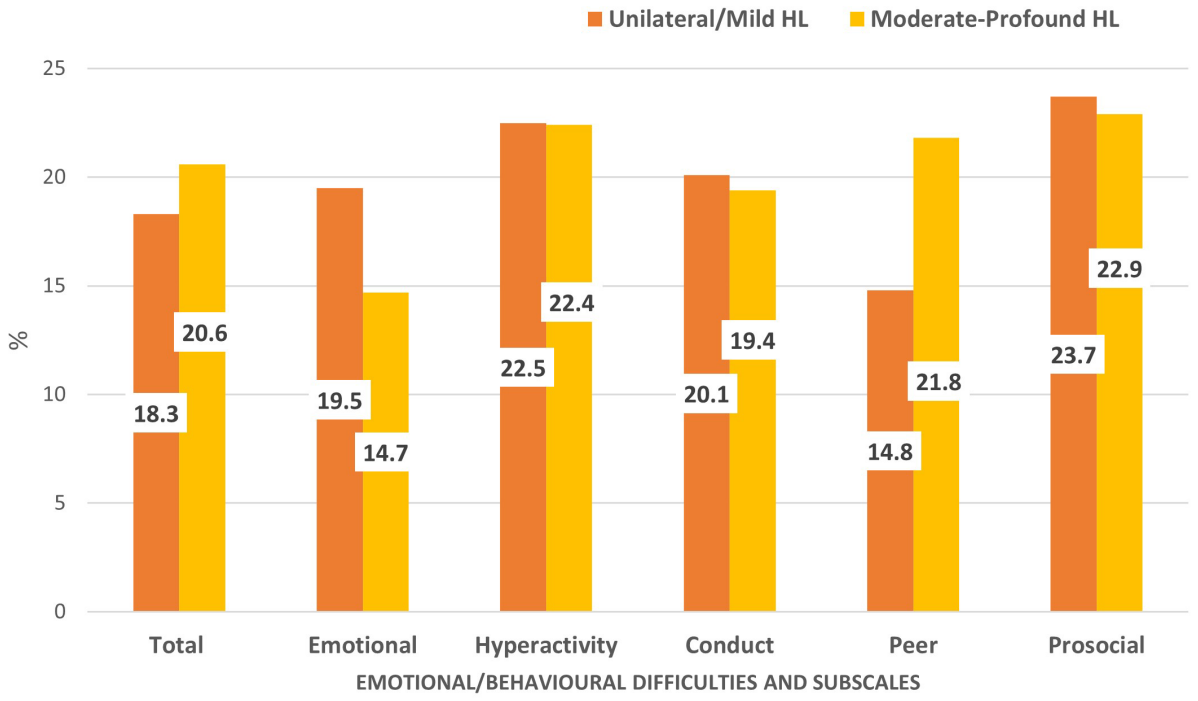

FIGURE 1
Proportion of children with unilateral/mild HL ($n = 169$) and moderate-profound HL ($n = 170$) with emotional/ behavioral difficulties and subscales scores above cut-off. * Cut-off scores are based on Australia normative data, retrieved from Kremer et al, 2015 and Mellow D., 2005.

(OR = 1.67; 95% CI 1.29–2.17, $p < 0.001$) with every additional health need. There was weaker evidence of association with demographic characteristics and early developmental concerns, nevertheless, the association between early developmental concerns, particularly gross motor milestone and attending mainstream school with emotional/behavioral difficulties were noteworthy. To investigate the association between each developmental domain and additional health needs, correlation analysis showed that children with unilateral/mild HL and additional health needs were more likely to have early problem-solving developmental concern reported at 36 months or younger, [r (98): 0.30, $p = 0.002$]. No association between additional health needs and other early developmental domains were demonstrated (Communication: r(98):0.09, $p = 0.36$; gross motor: r(98): 0.09, $p = 0.35$; fine motor: r (98): 0.08, $p = 0.41$; social: r(98): 0.04, $p = 0.70$).

More than one third (39.5%) of children with unilateral/ mild HL were unaided. The use of hearing devices and other audiological factors showed weak evidence of association with emotional/behavioral difficulties (Table 3). Table 4 further compared audiological factors and outcomes between children with unilateral HL and mild HL. More children with unilateral HL were unaided (65.5% vs. 17.6%) and had their first fitting with hearing aids at an older age (median age 24.0 months vs. 16.7 months) compared to children with mild HL. However, emotional/behavioral, HRQoL and parental psychological distress outcomes of unilateral and mild HL were comparable.

Discussion

Key findings

In a cross-sectional study of a large prospective cohort of DHH children, we showed that approximately one fifth of children 5–12 years old with unilateral/mild HL experienced emotional/ behavioral difficulties, measured as higher mean SDQ scores compared to Australian normative population data. Compared to peers with moderate-profound HL, children with unilateral/mild HL experienced comparable rates of emotional/behavioral difficulties and similar child health related quality-of- life and levels of parental psychological distress. Children with unilateral/ mild HL with additional health needs were at risk of emotional/ behavioral difficulties. Early developmental concerns, other than communication milestone and attending mainstream school showed weaker evidence of association.

Outcomes of unilateral/mild hearing loss

To our knowledge, this is one of the largest studies of DHH children demonstrating children with unilateral/mild HL experiencing comparable emotional/behavioral difficulties in comparison to peers with moderate-profound HL and greater than the normative population. Even though emotional/ behavioral mean scores were within normal range, the wide standard deviations and high proportion of scores indicative of

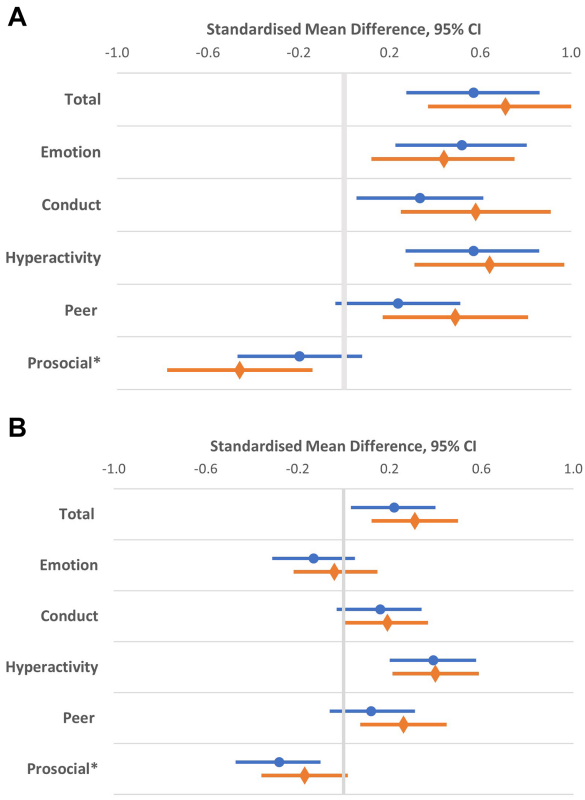

FIGURE 2
Compared to Australian population normative scores, children with unilateral/mild HL (•) and moderate to profound HL (◊) were reported to have higher mean differences for total and emotional/behavioral subscales according to age (A) 5-6 and (B) 7-12 years. *Lower prosocial behavior scores indicate difficulties.

TABLE 3 Estimated associations (quantified as odds ratios) between child characteristics and emotional/behavioral outcomes for children with unilateral/mild HL.

	Emotional/ behavioral difficulties	Odds ratio	*P* value
	(*N* = 31)	(95% CI)	
Gender, male, *n* (%)	15 (48.4)	1.08 (0.50, 2.36)	0.839
Age, 5– 6 years, *n* (%)	10 (32.3)	0.92 (0.40, 2.12)	0.842
Hearing laterality, bilateral, *n* (%)	16 (51.6)	0.91 (0.41, 2.00)	0.807
Hearing aid first fitting (*n* = 102), median, (IQR) months	24.0 (49.8)	1.01 (0.99, 1.03)	0.55
Cochlear implant first fitting (*n* = 6), median (IQR), months	87.0[a]	-	-
Type of hearing loss, sensorineural, *n* (%)	19 (61.3)	1.93 (0..85,4.39)	0.115
Hearing device, unaided, *n* (%)	13 (41.9)	0.91 (0.39, 2.13)	0.836
Communication mode, speech only, *n* (%)	25 (80.7)	4.72 (0.63, 35.12)	0.13
School, mainstream ± special unit, *n* (%)	22 (70.9)	3.30 (0.99, 11.02)	0.053
Additional health needs, *n* (%)	31 (100.0)	1.67 (1.29, 2.17)	<0.001
Early developmental concerns, *n* (%), (*n* = 100)			
Communication	4 (26.7)	0.98 (0.28,3.39)	0.975
Gross motor	5 (33.3)	3.36 (0.97, 11.70)	0.056
Fine motor	4 (26.7)	3.07 (0.81, 11.69)	0.1
Problem solving	4 (26.7)	2.73 (0.73, 10.22)	0.137
Social/Adaptive skill	4 (26.7)	3.07 (0.81, 11.69)	0.1

[a]Numbers too small for analysis.

emotional/behavioral difficulties suggests high variability in emotional/behavioral outcomes among children with unilateral/mild HL. Prior studies of DHH children with more severe degrees of HL have reported similar results; however, our participants with unilateral/mild HL demonstrated emotional/behavioral difficulties of large effect sizes compared to norms (2, 7).

Our study demonstrated a high prevalence of hyperactivity symptoms among both groups of DHH children. Although a review study reported contradicting results (2), many earlier studies have demonstrated ADHD-like symptoms among DHH children (6, 33). Hyperactivity symptoms are not unexpected, as children with HL and ADHD share similar difficulties in executive function and self-regulation (34, 35). The reduction in emotional/behavioral difficulties scores observed among the older children in both DHH groups suggest that developmental gap narrows with age in response to intervention, adaptation to challenges and with maturity. These age-related changes were also observed in a 3-year follow-up longitudinal study that showed improvement in behavioral problems among a proportion of school age children with unilateral HL, highlighting the positive gains of intervention in a selected group of DHH children (19).

TABLE 2 Child related HRQOL and parent distress standardized mean differences (SMD) of children with unilateral/mild bilateral HL and moderate-profound HL compared with norms.

	Norm(ref)	Unilateral/ mild HL	Effect size	*p*-value	Moderate- profound HL	Effect size	*p*-value
		Mean (SD)	(SMD) (95% CI)		Mean (SD)	(SMD) (95% CI)	
Parent distress	5.9 (4.3)	15.5 (3.6)	2.64 (2.27, 3.0)	<0.001	15.0 (3.7)	2.44 (2.08, 2.80)	<0.001
HRQOL							
Total	81.3 (15.9)	74.1 (18.4)	−0.39 (−0.54, −0.23)	<0.001	72.8 (17.9)	−0.47(−0.63, −0.31)	<0.001
Physical health	83.3 (20.0)	78.8 (23.9)	−0.18 (−0.22, −0.03)	0.022	78.4 (23.9)	−0.21(−0.36, −0.05)	0.008
Psychosocial	80.2 (15.8)	72.1 (16.6)	−0.49 (−0.65, −0.33)	<0.001	69.9 (17.6)	−0.78(−0.93, −0.59)	<0.001
Emotion	80.3 (17.0)	68.9 (18.6)	−0.61(−0.77, −0.44)	<0.001	69.7 (19.3)	−0.55(−0.71, −0.38)	<0.001
Social	82.2 (20.1)	76.3 (21.4)	−0.28(−0.43, −0.12)	<0.001	71.7 (23.1)	−0.46(−0.61, −0.30)	<0.001
School	76.9 (20.2)	71.1 (18.2)	−0.32(−0.47, −0.16)	<0.001	68.5 (18.8)	−0.45(−0.61, −0.29)	<0.001

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TABLE 4 Comparison of audiological factors and outcome scores of children with unilateral and mild hearing loss.

	Unilateral HL, $n = 96$	Mild HL, $n = 73$
Audiological characteristics		
Hearing device, *n* (%)		
Unaided	57 (65.5)	9 (17.6)
Hearing aid/s only	27 (31.0)	39 (76.5)
Cochlear implant	3 (3.4)	3 (5.9)
Age at first fitting of hearing aid		
Median (IQR), months	24.0 (47.0)	16.7 (26.3)
≤36 months, *n* (%)	20 (51.3)	53 (84.1)
Age of cochlear implantation, ($n = 6$)		
Median (IQR), months	30.0 (38.0)	41.0 (9.0)
≤36 months, *n* (%)	2 (66.7)	1 (33.3)
Outcomes, mean (SD); median (IQR)		
Emotional/behavioral Difficulties scores	9.8 (6.4); 8.0 (8.0)	9.6 (6.4); 9.0 (7.5)
n (%)	17 (17.7)	14 (19.2)
HRQOL scores,		
Total	73.8 (18.5); 77.2 (23.9)	74.6 (18.5); 78.8 (26.5)
Physical health	79.3 (25.3); 89.5 (31.0)	78.1 (24.9); 88.0 (34.0)
Psychosocial	71.5 (16.1); 71.7 (25.0)	72.9 (17.3); 75.0 (24.2)
Emotion	68.2 (18.4); 70.0 (25.0)	70.1 (18.9); 75.0 (28.8)
Social	76.3 (21.2); 80.0 (35.0)	76.2 (21.7); 80.0 (35.0)
School	70.1 (17.8); 75.0 (30.0)	72.5 (18.8); 75.0 (25.0)
Parent psychological distress (K6) scores	15.9 (3.6); 16.0 (4.0)	15.1 (3.7); 14.0 (5.0)

Emotional/behavioral outcomes were strongly correlated with child health related quality-of-life and parental distress levels. This association is consistent with prior reports, where parents of DHH children and adolescents with high externalizing and internalizing behaviors were more likely to report mental health problems and be burdened by the challenges faced (12, 36). Dammeyer et al. (2019) additionally reported that the degree of HL was not an influencing factor of the family's well-being (37). Families of children with unilateral/mild HL described different but consequential challenges compared to families of children with moderate to profound HL (37, 38). Parents reported feeling less support and empathy from the DHH community as their children with milder HL were perceived to be "Not deaf enough" and the significance of mild HL was minimized by healthcare providers (37). With the majority of children with unilateral/mild HL attending mainstream school, they would be required to fully rely on listening and speaking to communicate and experienced high expectations regarding their performances in academic, language and social skills. Furthermore, the perceived benefits of using hearing aids may be less obvious to both child and parents, hence many families may struggle with compliance and be frustrated or guilty when not able to follow through with intervention (37, 38). To address these unique challenges, further research capturing the various experiences of families with or without hearing devices may guide future recommendations and support required to optimize management of children with unilateral/mild HL.

Factors associated with emotional behavioral outcomes

Among children with unilateral/mild HL, our study identified additional health needs as the only predictive factor of emotional/behavioral outcomes. Although a few studies have suggested maternal education level and age of diagnosis/intervention as possible factors (20, 24), we did not identify other demographic or audiological factors that were significantly associated with emotional/behavioral outcomes. Numerous studies of DHH children have shown that additional disabilities and lower cognitive skills are associated with poorer outcomes and our study has identified a similar risk factor among children with unilateral/mild HL (10, 39, 40). Our finding is further supported by Wake et al. (2006)'s study that showed excellent outcomes among selected children of slight/mild hearing loss with no additional medical illness or intellectual disabilities (23). Children with HL and additional cognitive or physical comorbidities have more challenges that would impact their early milestones and response to audiological interventions. Furthermore, families of children having additional health needs besides HL are more likely to have marital and psychological distress that affects parent-child relationship (36). With over two thirds of children with unilateral/mild HL in this study having additional health needs, the need for early medical and developmental screening is warranted regardless of the degree/laterality of HL.

Our study is the first to examine early developmental profiles and the association with later emotional/behavioral outcomes among children with unilateral/mild HL. Early developmental screening of young DHH children may identify children at risk of later cognitive and educational difficulties (41). Among the five developmental domains screened, early problem-solving development is the domain most representative of early cognition and adaptive skills, hence it was not surprising that it was the only milestone found to be associated with the presence of additional health needs. Therefore, although no association was observed with emotional/behavioral outcomes, identifying young children with problem solving developmental concerns may help recognize children at risk of additional health problems. With close to one fifth of children with unilateral/mild HL reporting developmental concerns in all domains other than communication by 36 months of age, the possibility of association between early developmental concerns, particularly gross motor milestone with emotional/behavioral difficulties should not be disregard. Several studies have described the close relationship between hearing loss and motor development, suggesting that DHH children were at higher risk of deficits in balance and fine motor skills, with 12% of young DHH children detected with early gross motor developmental delay and one fifth of school age DHH children to be less competent in gross and fine motor skills compared to typical hearing peers (9, 11, 42). Children with slight/mild HL have also been reported to have significantly poorer physical HRQoL compared to typical hearing children (33) hypothesized to be related to inner ear

abnormalities or the lack of environmental exposure due to sensory deprivation. Concurrent gross motor delay among DHH children is likely to intensify emotional/behavioral difficulties as the presence of motor delay in typical hearing children have been reported to have more emotional/behavior difficulties (43). However, the relationship between motor development, hearing loss and outcomes is still poorly understood and requires more substantial evidence to warrant any recommendations.

Available evidence of the benefits of hearing assistive devices among children with unilateral/mild HL in preventing emotional/behavioral difficulties are limited and debatable. Our study showed no association between the use of hearing devices and age of amplification with emotional/behavioral outcomes. An outcome study of DHH children detected across four hearing screening systems similarly found that behavior and HRQoL to be largely unaffected by the advancement in hearing screening and expressive language continued to be lower than expected among children with mild and moderate hearing loss (13). However, other studies proposed that early age of diagnosis and amplification were associated with better social skills and psychoeducation outcomes (20, 24). Fitzpatrick et al. (2022) also found no additional behavioral problems and parenting stress among 4-year-old children with unilateral/mild HL who were identified early at median age of 4.5 months (22). Regardless, substantial evidence demonstrating clear benefits of using hearing aids to promote socio-emotional growth in children with unilateral/mild HL is unavailable. Studies of DHH children with various hearing loss severity have also shown that the use of hearing devices may not be protective of emotional/behavioral difficulties despite improvement in language abilities (6, 7, 10). Despite the possible lack of association between hearing aids and emotional/behavioral outcomes, our result should be interpreted with care due to the high probability of additional unmeasured confounding factors such as hours of aided time, quality of fitting and parental psychosocial barriers (44, 45).

Our study further described the differences in characteristics and outcomes among children with unilateral HL and mild HL. Children with unilateral HL had similar scores as children with mild HL in emotional/behavioral, HRQOL and parental distress outcomes. However, more children with unilateral HL were unaided and were first fitted with a hearing aid at a later age indicating that children with unilateral HL had less access to early intervention and support compared to children with mild HL. The differences observed highlight the possible additional challenges faced by children with unilateral HL which are yet to be explored.

Strengths and limitations

Among the many strengths, our study has included data from a large number of children with unilateral/mild HL with and without hearing devices. These children are often under-represented in research as they are less likely to have regular healthcare appointments or appear in clinical databases. We used the SDQ, a validated measure of emotional/behavioral outcomes for the range of ages of our participants, enabling referencing of our results to population norms. Several limitations have also been identified. While VicCHILD is a population-based cohort, responders lived in areas of relatively less socioeconomic disadvantage compared to non-responders, and therefore results may not be generalizable to those living in more disadvantaged areas The duration of daily usage of hearing aids and level of audibility among users of hearing devices were not explored and would have provided information about the compliance of hearing aid usage particularly among unilateral/mild HL children. Likewise, information of prior usage of hearing devices and duration of use among unaided children during time of assessment may provide better understanding of its influence on socioemotional development. When analyzing the association between emotional/behavioral difficulties with different variables, we noted several odds ratio with wide confidence interval. The lack of precision may be due to the small sample size of children with unilateral/mild HL and emotional/behavioral difficulties. Larger sample sized studies may be able to explore, for example, the relationship between the accumulative effects of early developmental concerns and emotional/behavioral difficulties. Similarly, for the type of school and early developmental concerns, the result should be interpreted judiciously due to the small sample of unilateral/mild HL children not attending mainstream school and with developmental concerns; however the possibility of association with other factors should not be disregard. Single parent rated assessments used in this study to evaluate emotional/behavioral outcomes may not provide a complete perspective due to differences in child behavior and assessor priorities across different settings. However, despite differences in emotional/behavioral symptoms reported among parents and teachers, recognition of emotional/behavioral difficulties by both assessors were shown to be significantly correlated (3). Hence incorporating teacher- rated measures may not influence the result but will provide a better clinical understanding of the child's behavior throughout the day. The study strength of including DHH children of variable audiological and intervention background may also be a limitation, due to the high heterogeneity of the participants. However, the variation in the participants' characteristics reflects the real world and the different challenges faced by families of children with unilateral/mild HL. We were not able to further analyze the specifics effects of additional health needs on outcomes due to incomplete data, as additional health needs were collected only after 2020. The high reported rate of additional health needs in our sample observed, may also likely be due to the comprehensiveness of the list of medical diagnosis provided to parents. Characterizing additional health needs in future studies will provide important knowledge to accurately predict high risk DHH children and understand how they influence DHH children's emotion and behavior.

Conclusion

This study demonstrated that children with unilateral/mild HL were just as likely as children with moderate- profound HL to

experience more emotional/behavioral difficulties, poorer HRQoL and higher parental distress compared to the general population. Our study results justify the provision of early access to services and support among children with unilateral/mild HL. Early developmental screening of additional health needs is crucial to identify children with unilateral/mild hearing loss who are at risk of emotional/behavioral difficulties, as early individualized intervention may improve quality of life and parental well-being.

Data availability statement

The raw data supporting the conclusions of this article will be made available by the authors, without undue reservation.

Ethics statement

The studies involving humans were approved by Royal Children's Hospital Human Research Ethics Committee (approval number 31081). The studies were conducted in accordance with the local legislation and institutional requirements. Written informed consent for participation in this study was provided by the participants' legal guardians/next of kin.

Author contributions

JJO, LS, GR and VS were primarily responsible for the conception and design of the study. LS and JX were responsible for acquisition of the data. JJO analyzed the data. DS provided statistical advice. All authors contributed to the interpretation of data, critical revision of manuscript, read and approved the final version of the manuscript. All authors contributed to the article and approved the submitted version.

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Conflict of interest

The authors declare that the research was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

Publisher's note

Supplementary material

The Supplementary Material for this article can be found online at: https://www.frontiersin.org/articles/10.3389/fped.2023.1209736/full#supplementary-material

References

1. Wake M, Hughes EK, Poulakis Z, Collins C, Rickards FW. Outcomes of children with mild-profound congenital hearing loss at 7 to 8 years: a population study. *Ear Hear.* (2004) 25(1):1–8. doi: 10.1097/01.AUD.0000111262.12219.2F

2. Stevenson J, Kreppner J, Pimperton H, Worsfold S, Kennedy C. Emotional and behavioural difficulties in children and adolescents with hearing impairment: a systematic review and meta-analysis. *Eur Child Adolesc Psychiatry.* (2015) 24:477–96. doi: 10.1007/s00787-015-0697-1

3. Wong CL, Ching TY, Cupples L, Leigh G, Marnane V, Button L, et al. Comparing parent and teacher ratings of emotional and behavioural difficulties in 5-year old children who are deaf or hard-of-hearing. *Deafness Educ Int.* (2020) 22(1):3–26. doi: 10.1080/14643154.2018.1475956

4. Vissers CT, Hermans D. Social-emotional problems in deaf and hard-of-hearing children from an executive and theory of mind perspective. *Evid Based Pract Deaf Educ*. (2018) 28:455–76.

5. Szarkowski A, Young A, Matthews D, Meinzen-Derr J. Pragmatics development in deaf and hard of hearing children: a call to action. *Pediatrics*. (2020) 146 (Supplement_3):S310–5. doi: 10.1542/peds.2020-0242l

6. Wong CL, Ching TY, Leigh G, Cupples L, Button L, Marnane V, et al. Psychosocial development of 5-year-old children with hearing loss: risks and protective factors. *Int J Audiol*. (2018) 57(sup2):S81–92. doi: 10.1080/14992027.2016.1211764

7. Wong CL, Ching TYC, Cupples L, Button L, Leigh G, Marnane V, et al. Psychosocial development in 5-year-old children with hearing loss using hearing aids or cochlear implants. *Trends Hear*. (2017) 21:2331216517710373. doi: 10.1177/2331216517710373

8. Theunissen SC, Rieffe C, Netten AP, Briaire JJ, Soede W, Schoones JW, et al. Psychopathology and its risk and protective factors in hearing-impaired children and11 adolescents: a systematic review. *JAMA Pediatr*. (2014) 168(2):170–7. doi: 10.1001/jamapediatrics.2013.3974

9. Hogan A, Shipley M, Strazdins L, Purcell A, Baker E. Communication and behavioural disorders among children with hearing loss increases risk of mental health disorders. *Aust N Z J Public Health*. (2011) 35(4):377–83. doi: 10.1111/j.1753-6405.2011.00744.x

10. Leigh G, Ching TY, Crowe K, Cupples L, Marnane V, Seeto M. Factors affecting psychosocial and motor development in 3-year-old children who are deaf or hard of hearing. *J Deaf Stud Deaf Educ*. (2015) 20(4):331–42. doi: 10.1093/deafed/env028

11. Wiley S, Arjmand E, Dixon M. Findings from multidisciplinary evaluation of children with permanent hearing loss. *Int J Pediatr Otorhinolaryngol*. (2011) 75 (8):1040–4. doi: 10.1016/j.ijporl.2011.05.019

12. Overgaard KR, Oerbeck B, Wagner K, Friis S, Øhre B, Zeiner P. Youth with hearing loss: emotional and behavioral problems and quality of life. *Int J Pediatr Otorhinolaryngol*. (2021) 145:110718. doi: 10.1016/j.ijporl.2021.110718

13. Carew P, Mensah FK, Rance G, Flynn T, Poulakis Z, Wake M. Mild–moderate congenital hearing loss: secular trends in outcomes across four systems of detection. *Child Care Health Dev*. (2018) 44(1):71–82. doi: 10.1111/cch.12477

14. Walker EA. Evidence-based practices and outcomes for children with mild and unilateral hearing loss. *Lang Speech Hear Serv Sch*. (2020) 51(1):1–4. doi: 10.1044/2019_LSHSS-19-00073

15. Tomblin JB, Harrison M, Ambrose SE, Walker EA, Oleson JJ, Moeller MP. Language outcomes in young children with mild to severe hearing loss. *Ear Hear*. (2015) 36(0 1):76S. doi: 10.1097/AUD.0000000000000219

16. Tharpe AM. Unilateral and mild bilateral hearing loss in children: past and current perspectives. *Trends Amplif*. (2008) 12(1):7–15. doi: 10.1177/1084713807304668

17. Bess FH, Dodd-Murphy J, Parker RA. Children with minimal sensorineural hearing loss: prevalence, educational performance, and functional status. *Ear Hear*. (1998) 19(5):339–54. doi: 10.1097/00003446-199810000-00001

18. Đoković S, Gligorović M, Ostojić S, Dimić N, Radić-Šestić M, Slavnić S. Can mild bilateral sensorineural hearing loss affect developmental abilities in younger school-age children? *J Deaf Stud Deaf Educ*. (2014) 19(4):484–95. doi: 10.1093/deafed/enu018

19. Lieu JE, Tye-Murray N, Fu Q. Longitudinal study of children with unilateral hearing loss. *Laryngoscope*. (2012) 122(9):2088–95. doi: 10.1002/lary.23454

20. Porter H, Sladen DP, Ampah SB, Rothpletz A, Bess FH. Developmental outcomes in early school-age children with minimal hearing loss. *Am J Audiol*. (2013) 22(2):263–70. doi: 10.1044/1059-0889(2013/13-0013)

21. le Clercq CM, Labuschagne LJ, Franken MC, de Jong RJ, Luijk MP, Jansen PW, et al. Association of slight to mild hearing loss with behavioral problems and school performance in children. *JAMA Otolaryngology–Head & Neck Surgery*. (2020) 146 (2):113–20. doi: 10.1001/jamaoto.2019.3585

22. Fitzpatrick EM, Jiawen W, JoAnne W, Flora N, Isabelle G, Andrée DS, et al. Parent-Reported stress and child behavior for 4-year-old children with unilateral or mild bilateral hearing loss. *J Deaf Stud Deaf Educ*. (2022) 27(2):137–50. doi: 10.1093/deafed/enab042

23. Wake M, Tobin S, Cone-Wesson B, Dahl HH, Gillam L, McCormick L, et al. Slight/mild sensorineural hearing loss in children. *Pediatrics*. (2006) 118(5):1842–51. doi: 10.1542/peds.2005-3168

24. Laugen NJ, Jacobsen KH, Rieffe C, Wichstrøm L. Social skills in preschool children with unilateral and mild bilateral hearing loss. *Deafness Educ Int*. (2017) 19(2):54–62. doi: 10.1080/14643154.2017.1344366

25. Sung V, Smith L, Poulakis Z, Burt RA, Carew P, Tobin S, et al. Data resource profile: the victorian childhood hearing impairment longitudinal databank (VicCHILD). *Int J Epidemiol*. (2019) 48(5):1409–10h. doi: 10.1093/ije/dyz168

26. Rose E, Sung V. Otolaryngology. In: *Paediatric Handbook*. 10th edition. Hoboken, NJ: Wiley-Blackwell (2021). p. 451–64.

27. Bricker D, Squires J, Mounts L, Potter L, Nickel R, Twombly E, et al. *Ages and stages questionnaire*. Baltimore, MD: Paul H. Brookes (1999).

28. Goodman R. The strengths and difficulties questionnaire: a research note. *J Child Psychol Psychiatry*. (1997) 38(5):581–6. doi: 10.1111/j.1469-7610.1997.tb01545.x

29. Kremer P, de Silva A, Cleary J, Santoro G, Weston K, Steele E, et al. Normative data for the strengths and difficulties questionnaire for young children in Australia. *J Paediatr Child Health*. (2015) 51(10):970–5. doi: 10.1111/jpc.12897

30. Mellor D. Normative data for the strengths and difficulties questionnaire in Australia. *Aust Psychol*. (2005) 40(3):215–22. doi: 10.1080/00050060500243475

31. Varni JW, Burwinkle TM, Seid M, Skarr D. The PedsQL^{TM*} 4.0 as a pediatric population health measure: feasibility, reliability, and validity. *Ambul Pediatr*. (2003) 3(6):329–41. doi: 10.1367/1539-4409(2003)003<0329:TPAAPP>2.0.CO;2

32. Kessler RC, Barker PR, Colpe LJ, Epstein JF, Gfroerer JC, Hiripi E, et al. Screening for serious mental illness in the general population. *Arch Gen Psychiatry*. (2003) 60(2):184–9. doi: 10.1001/archpsyc.60.2.184

33. Hancock KJ, Brennan-Jones CG, Vithiatharan R, Payne D, Runions K, Lin A, et al. Mental health problems among 4–17-year-olds with hearing problems: results from a nationally representative study. *Hearing Balance Commun*. (2017) 15 (3):145–55. doi: 10.1080/21695717.2017.1325094

34. Soleimani R, Jalali MM, Faghih HA. Comparing the prevalence of attention deficit hyperactivity disorder in hearing-impaired children with normal-hearing peers. *Archives de Pédiatrie*. (2020) 27(8):432–5. doi: 10.1016/j.arcped.2020.08.014

35. Hintermair M. Executive functions and behavioral problems in deaf and hard-of-hearing students at general and special schools. *J Deaf Stud Deaf Educ*. (2013) 18 (3):344–59. doi: 10.1093/deafed/ent003

36. Dammeyer J, Hansen AT, Crowe K, Marschark M. Childhood hearing loss: impact on parents and family life. *Int J Pediatr Otorhinolaryngol*. (2019) 120:140–5. doi: 10.1016/j.ijporl.2019.02.027

37. Lin JJ, Gillam L, Smith L, Carew P, King A, Ching TY, et al. Mild matters: parental insights into the conundrums of managing mild congenital hearing loss. *Int J Audiol*. (2022) 61(6):500–6. doi: 10.1080/14992027.2021.1954248

38. Fitzpatrick E, Grandpierre V, Durieux-Smith A, Gaboury I, Coyle D, Na E, et al. Children with mild bilateral and unilateral hearing loss: parents' reflections on experiences and outcomes. *J Deaf Stud Deaf Educ*. (2016) 21(1):34–43. doi: 10.1093/deafed/env047

39. Theunissen SC, Rieffe C, Kouwenberg M, De Raeve LJ, Soede W, Briaire JJ, et al. Behavioral problems in school-aged hearing-impaired children: the influence of sociodemographic, linguistic, and medical factors. *Eur Child Adolesc Psychiatry*. (2014) 23(4):187–96. doi: 10.1007/s00787-013-0444-4

40. Dammeyer J. Psychosocial development in a danish population of children with cochlear implants and deaf and hard-of-hearing children. *J Deaf Stud Deaf Educ*. (2010) 15(1):50–8. doi: 10.1093/deafed/enp024

41. Charkaluk ML, Rousseau J, Calderon J, Bernard JY, Forhan A, Heude B, et al. Ages and stages questionnaire at 3 years for predicting IQ at 5–6 years. *Pediatrics*. (2017) 139(4):e20162798. doi: 10.1542/peds.2016-2798

42. Rajendran V, Roy FG, Jeevanantham D. Postural control, motor skills, and health-related quality of life in children with hearing impairment: a systematic review. *Eur Arch Oto-Rhino-Laryngol*. (2012) 269:1063–71. doi: 10.1007/s00405-011-1815-4

43. Rodriguez MC, Wade TJ, Veldhuizen S, Missiuna C, Timmons B, Cairney J. Emotional and behavioral problems in 4-and 5-year old children with and without motor delays. *Front Pediatr*. (2019) 7:474. doi: 10.3389/fped.2019.00474

44. Walker EA, Spratford M, Moeller MP, Oleson J, Ou H, Roush P, et al. Predictors of hearing aid use time in children with mild-to-severe hearing loss (2013).

45. Munoz KF, Nichols N, Hill S. Psychosocial experiences of parents of young children who use hearing devices: a scoping review. *Journal of Early Hearing Detection and Intervention*. (2021) 6(1):90–5. doi: 10.26077/2656-d109

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EDITED BY
Zhengqing Hu,
Wayne State University, United States

REVIEWED BY
Ryan William McCreery,
Boys Town National Research Hospital, United States
Holly Teagle,
The University of Auckland, New Zealand

*CORRESPONDENCE
Vicky W. Zhang
✉ Vicky.Zhang@nal.gov.au

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Audiological characteristics of children with congenital unilateral hearing loss: insights into Age of reliable behavioural audiogram acquisition and change of hearing loss

Vicky W. Zhang[1,2]*, Sanna Hou[1], Angela Wong[1], Christopher Flynn[1,3], Jane Oliver[1,4], Michelle Weiss[1,5], Stacey Milner[1,6] and Teresa Y. C. Ching[1,7,8,9]

[1]Audiological Science Department, National Acoustic Laboratories, Sydney, NSW, Australia, [2]Department of Linguistics, Macquarie University, Sydney, NSW, Australia, [3]Lutwyche centre, Hearing Australia, Brisbane, QLD, Australia, [4]Upper Mt Gravatt centre, Hearing Australia, Brisbane, QLD, Australia, [5]Dandenong centre, Hearing Australia, Melbourne, VIC, Australia, [6]Cheltenham centre, Hearing Australia, Melbourne, VIC, Australia, [7]NextSense Institute, Macquarie Park, Sydney, NSW, Australia, [8]Macquarie School of Education, Macquarie University, Sydney, NSW, Australia, [9]School of Health and Rehabilitation Sciences, University of Queensland, St Lucia, QLD, Australia

Objectives: The aims of this study were to report the audiological characteristics of children with congenital unilateral hearing loss (UHL), examine the age at which the first reliable behavioural audiograms can be obtained, and investigate hearing changes from diagnosis at birth to the first reliable behavioural audiogram.
Method: This study included a sample of 91 children who were diagnosed with UHL via newborn hearing screening and had reliable behavioural audiograms before 7 years of age. Information about diagnosis, audiological characteristics and etiology were extracted from clinical reports. Regression analysis was used to explore the potential reasons influencing the age at which first reliable behavioural audiograms were obtained. Correlation and ANOVA analyses were conducted to examine changes in hearing at octave frequencies between 0.5 and 4 kHz. The proportions of hearing loss change, as well as the clinical characteristics of children with and without progressive hearing loss, were described according to two adopted definitions: Definition 1: criterion (1): a decrease in 10 dB or greater at two or more adjacent frequencies between 0.5 and 4 kHz, or criterion (2): a decrease in 15 dB or greater at one octave frequency in the same frequency range. Definition 2: a change of ≥20 dB in the average of pure-tone thresholds at 0.5, 1, and 2 kHz.
Results: The study revealed that 48 children (52.7% of the sample of 91 children) had their first reliable behavioural audiogram by 3 years of age. The mean age at the first reliable behavioural audiogram was 3.0 years (SD 1.4; IQR: 1.8, 4.1). We found a significant association between children's behaviour and the presence or absence of ongoing middle ear issues in relation to the delay in obtaining a reliable behavioural audiogram. When comparing the hearing thresholds at diagnosis with the first reliable behavioural audiogram across different frequencies, it was observed that the majority of children experienced deterioration rather than improvement in the initial impaired ear at each frequency. Notably, there were more instances of hearing changes (either deterioration or improvement), in the 500 Hz and 1,000 Hz frequency ranges compared to the 2,000 Hz and 4,000 Hz ranges. Seventy-eight percent ($n = 71$) of children had hearing deterioration between the diagnosis and the first behavioural audiogram at one or more frequencies between 0.5 and

4 kHz, with a high proportion of them (52 out of the 71, 73.2%) developing severe to profound hearing loss. When using the averaged three frequency thresholds (i.e., definition 2), only 26.4% of children ($n = 24$) in the sample were identified as having hearing deterioration. Applying definition 2 therefore underestimates the proportion of children that experienced hearing changes. The study also reported diverse characteristics of children with or without hearing deterioration.

Conclusion: The finding that 78% of children diagnosed with UHL at birth had a decrease in hearing loss between the hearing levels at first diagnosis and their first behavioural audiogram highlights the importance of monitoring hearing threshold levels after diagnosis, so that appropriate intervention can be implemented in a timely manner. For clinical management, deterioration of 15 dB at one or more frequencies that does not recover warrants action.

KEYWORDS

unilateral hearing loss, children, audiological characteristics, behavioural audiogram, progressive hearing loss, etiology

1. Introduction

Universal Newborn Hearing Screening (UNHS) programs have played a crucial role in identifying unilateral hearing loss (UHL) in infancy. Since its implementation, there has been a significant increase in the prevalence of UHL in newborns, rising from approximately 0.3–0.6 per 1,000 (1, 2) to 1–2.4 per 1,000 children (3–6). This increase has drawn increased attention to the impact of UHL during early childhood. Previous studies have shed some light on the significant impacts of UHL on various aspects of development in certain children. These impacts include difficulties in sound localization, speech recognition in noise (7–13), and higher-level language skills such as cognition, comprehension, reading, and communication (12, 14). Long-term impacts have also been reported, indicating that children with hearing loss may experience poorer outcomes such as overall quality of life (15), academic achievements, and psychosocial challenges, compared to children with normal hearing in both ears (16). The early identification of UHL is essential to ensure timely and effective intervention for optimal developmental outcomes. However, clinical management of UHL still presents several challenges due to limited knowledge regarding the audiological characteristics, underlying causes, and a lack of evidence-based information regarding the long-term consequences in this target population.

The accurate diagnosis and timely intervention as early as possible are widely recognized as crucial for optimal audiological management of hearing loss configuration, progression, and long-term outcomes in children with hearing loss (17, 18). Currently, electrophysiological tests such as auditory brainstem response (ABR) and auditory steady-state response (ASSRs) measures are commonly used to estimate children's behavioural audiograms. It should note that these tests only provide approximations. For instance, tone-burst evoked auditory brainstem response (TBABRs) has an error range of ±5–20 dB when determining behavioural results at different frequencies (19–21). Additionally, there is no reliable method to estimate hearing threshold for the impaired ear of a child with unilateral auditory neuropathy spectrum disorder until they can provide reliable individual ear behavioural testing. These highlight the need for a reliable behavioural audiogram for each ear. However, acquiring ear-specific reliable behavioural audiograms can be challenging, especially for children with UHL, as they must be able to accept the use of insert-phones or headphones during the testing. Furthermore, due to the asymmetrical hearing levels, masking is often required, which also adds to the complexity during behavioural testing. The challenge is further exacerbated by some other non-technical factors such as the child's developmental stage, general cooperation, and limited time allocated for clinical appointments. There is limited information available regarding the average age at which reliable behavioural hearing levels can be obtained in children with congenital UHL diagnosed via UNHS.

Another challenge lies in understanding the prevalence rate of progressive hearing loss in children with UHL, and its potential risk factors. Despite the fact that children with UHL can still perceive sound through their unaffected ear, the importance of obtaining reliable behavioural audiograms and regularly monitoring hearing thresholds for each ear may have been insufficiently emphasized. Studies have indicated that a considerable proportion of children with UHL are at risk of progressive hearing loss, either in one or both ears (22, 23). However, the percentage of children with UHL who exhibit hearing deterioration varies across reports. This may be due to differences in definitions of progressive hearing loss used, age ranges of the children, measurement methods, follow-up durations and specific sub-groups of hearing loss under investigation (22, 24–30). For example, Dahl and colleagues (25) defined progressive hearing loss as a decrease in 10 dB or greater at two or more adjacent frequencies between 0.5 and 4 kHz or a decrease in 15 dB at one octave frequency in the same frequency range over the period of investigation. Studies on UHL that adopted this definition have reported progressive hearing loss in 37%–47.5% of children with UHL, with 11.9%–19% eventually developing bilateral hearing loss (22, 23, 26). The figures are not directly comparable, which may be due to the different sample

sizes and variations in baseline and the most recent audiometric assessment points used in the studies. Several other studies have also used averaged hearing thresholds from three or four frequencies, considering hearing changes of ≥10 (31), ≥15 (32), or ≥20 dB (16) as indicative of progressive hearing loss in children with UHL. For instance, according to the Fitzpatrick study (16), 12.9% of children with UHL were reported to have a change of ≥20 dB in the pure-tone average of three frequency thresholds from 0.5 to 2 kHz. In comparison, Purcell et al. (31) found that 32.8% of children with UHL, specifically associated with ipsilateral bony cochlear nerve canal stenosis, experienced progressive hearing loss, defined as a change of ≥10 dB in pure-tone thresholds averaged across 0.5, 1 and 2 kHz. The study started hearing assessments from a mean age of 7.7 years and followed-up participants for approximately 3 years (1,126 days). Additionally, Paul et al. (33) reported a rate of 19% for progressive hearing loss when using a similar criterion of >10 dB change in the pure-tone average of four frequency thresholds from 0.5 to 4 kHz. In terms of measurement points and lengths of time between measures, different studies have focused on various approaches. For example, Fitzpatrick et al. (23) compared the diagnostic audiogram (median age 3.3 months) to the most recent audiometric assessment (median age 88.8 months) with a median length of 64.3 months between measurements. They found that 47.5% of the children showed progressive loss, and 11.9% progressed to bilateral loss. Another study by Fitzpatrick et al. (34) reported that 8 out of 62 children with UHL showed progressive hearing loss (12.9%). The figure, based on data collected from 1990 to 2010, is lower than the 2023 report, which could be due to some children in the 2014 study not being identified with hearing loss until after 5 years of age (i.e., after the implementation of a UNHS program). Moreover, the potential factors contributing to progressive hearing loss in UHL children still remain inconclusive (35). Some studies suggested that cytomegalovirus (CMV) (36–38) and mutations in the gap junction β 2 gene (GJB2) (39) are risk factors. However, other studies have not found significant associations between progressive hearing loss and indicators such as genetic mutations and, NICU admission, family history, craniofacial anomalies, syndromes, postnatal infections (22, 25), or factors like age at diagnosis, severity of hearing loss, or etiologic (23, 26).

Due to the challenges associated with obtaining a reliable behavioural audiogram during early childhood, and the variations in reported proportions and potential risks of progressive hearing loss documented in different studies on UHL, there is a need for more evidence to determine the age at which a reliable audiogram can be obtained, as well as to gather more information on the extent of hearing changes in children with UHL. This knowledge will contribute to the development of optimal management and intervention strategies. Considering these challenges and the importance of early intervention, this study aims (1) to describe the audiological characteristics and etiology of a group of children diagnosed with congenital unilateral hearing loss (CUHL); (2) to examine the age at which reliable behavioural audiograms can be obtained and explore potential factors that could delay obtaining these audiograms; and (3) to investigate changes in hearing levels from birth to the first reliable behavioural audiogram, as well as the potential risks of progressive hearing loss.

2. Materials and methods

2.1. Participants

Participants included 91 children who were enrolled in the Children with Unilateral Hearing Loss study in New South Wales ($n = 28$), Victoria ($n = 24$), and Queensland ($n = 39$). All participants were identified with UHL via newborn hearing screening with a subsequent diagnosis of permanent UHL confirmed via electrophysiological hearing tests at diagnostic centres and hospitals. These include tympanometry, distortion product otoacoustic emission (DPOAEs), and TBABRs or ASSRs testing on both ears at a diagnostic centre or hospital for birth dates between March 19, 2014, and February 8, 2018. Following diagnosis of the hearing loss, children were referred to Hearing Australia (the national government funded organisation that provides hearing services to all children with hearing loss under the age of 26 years in Australia) to receive further audiological services, which includes ongoing hearing assessments, hearing device fitting and verification. Inclusion criteria for this report included children enrolled in the study on unilateral hearing loss (reported separately) who had (1) a diagnosis of UHL; (2) frequency-specific audiometric thresholds estimated from electrophysiological testing at diagnosis; (3) reliable behavioural audiograms in early childhood at least at one low frequency (0.5 or 1 kHz) and one high frequency (2 or 4 kHz) at Hearing Australia (see details in Section 2.2, "Audiological data collection for the first reliable behavioural audiogram"). Additionally, children diagnosed with Auditory Neuropathy Spectrum Disorder (ANSD) were excluded from the sample due to the inaccuracies in estimated hearing thresholds using electrophysiological hearing tests at the time of diagnosis. After a comprehensive review of records at Hearing Australia, data of children who met the inclusion criteria were included in this report. This study has been approved by Hearing Australia Human Research Ethics Committee (No. AHHREC2014-28 and No. AHHREC2019-9).

2.2. Data collection procedures

2.2.1. Audiological data collection at diagnosis

Diagnostic data for this study were collected at the time when children were diagnosed with UHL. The hearing thresholds at diagnosis in each ear were measured by audiologists at diagnostic centres or hospitals using objective electrophysiological tests of TBABRs or ASSRs. The correction factors applied to convert the electrophysiological hearing results into estimated behavioural hearing threshold (dB eHL) were as follows: for TBABRs, 500 Hz was adjusted by 10 dB, 1,000 Hz by 10 dB, 2,000 Hz by 5 dB, and 4,000 Hz by 0 dB; for ASSRs, 500 Hz was adjusted by 15 dB, 1,000 Hz by 10 dB, 2,000 Hz by 10 dB, and 4,000 Hz by 10 dB

(19, 40, 41). The age at diagnosis was defined as the age of hearing loss confirmed at the diagnostic centre or hospital using relevant electrophysiological methods.

2.2.2. Audiological data collection for the first reliable behavioural audiogram

The behavioural audiological assessments after diagnosis for all children were performed by clinical paediatric audiologists at Hearing Australia according to the national audiological protocols. The behavioural hearing thresholds were obtained using visual reinforcement audiometry (VROA), conditioned play audiometry (PA), or a combination behavioural method of VROA and PA if needed, depending on the child's age and ability. Serial behavioural audiological results after the time of diagnosis for each child were retrospectively reviewed by experienced research audiologists from clinical records held at Hearing Australia, to identify reliable behavioural audiograms. The definition of a reliable behavioural audiogram for the impaired ear includes:

- clinical note shows reliable behavioural results on the tested frequencies.
- must have hearing thresholds at least at one low frequency (0.5 or 1 kHz) and one high frequency (2 or 4 kHz).
- must have masked hearing thresholds when required, for air and bone conduction thresholds.

Specific decision rules were developed for this study, such as if an audiogram associated with temporary abnormal middle ear function, it was not recorded as a reliable result, and subsequent audiological results were examined accordingly. The date and the detailed audiological results (e.g., hearing thresholds, transducer, assessment method, clinical comments) of the first reliable behavioural audiogram were then recorded for further analysis.

2.2.3. Factors affecting the age of obtaining reliable behavioural audiograms

To identify potential factors affecting the age at which reliable behavioural audiograms were obtained, children's demographic and basic audiological information, as well as the etiological details were extracted from available clinical reports of Hearing Australia and provided reports by other diagnostic organizations to Hearing Australia. To gain further insights of clinicians' experiences and challenges on the potential reasons that may affect how early a behavioural audiogram is obtained, we invited clinical audiologists at Hearing Australia to complete an informal online survey[1]. According to the survey results and content analysis of additional comments provided by audiologists, a few other reasons were identified, apart from the collected children's demographic data, audiological characteristics and etiological details. These additional factors included children's behaviour/compliance, staff and equipment resources, scheduled appointments time, as well as caregivers' attitudes toward UHL and family availability.

Considering the suggested reasons from the survey, we reviewed the clinical case notes of earlier testing appointments before obtaining the first reliable behavioural audiogram. We then grouped the comments reported during the actual appointments into three categorised reasons: (1) children's behavioural issues (such as loss of interest in tasks, attention issues, or intolerance of earphones), (2) challenges unrelated to children's behavioural issues (such as insufficient masking information for children requiring masking, or insufficient appointment time for masking), and (3) family availability (such as lost contact or failure to attend scheduled appointments). Cases exhibiting any of these issues were labelled as "Yes", while those without such issues were labelled as "No". These three potential reasons and the other four clinical variables (presence or absence of reported etiology, degree of hearing loss at diagnosis, ongoing middle ear problems in the impaired ear, and hearing device fitting) were included in subsequent statistical analysis and discussion on the reasons of the delay in obtaining behavioural audiogram in this report.

2.2.4. Definition of degree of hearing loss

The degree of hearing loss in the impaired ear at diagnosis and behavioural audiological assessments was further determined by three-frequency averaged thresholds at 0.5, 1, and 2 kHz (22, 26). All children included in this report had all three frequency-specific audiometric thresholds (0.5, 1, and 2 kHz) estimated from electrophysiological testing at diagnosis in each ear. For the behavioural audiograms in the impaired ear, all children had measured thresholds at 1 kHz, whilst 15 children had missing data at 0.5 kHz, 19 children had missing data at 2 kHz. To address this, a general rule using an estimation method by extrapolating from the available measured frequency-specific results (as described in (42) was used to make the best estimate of behavioural hearing thresholds for the missing values:

[1]The survey consisted of two main questions: (A) what is the clinician's perspective regarding the time of getting reliable behavioural results for child's individual ear. Audiologists were instructed to select a response from a list of 5 responses including: (1) <1-year-old; (2) 1–1.5 years old; (3) 1.5–2 years old; (4) 2–3 years old; and (5) >3-year-old. (B) what are the most relevant reasons with regards to the potential delays in obtaining the behavioural audiogram. Audiologists were asked to select the top 3 most relevant reasons including (1) Appointment time; (2) Equipment resources (e.g., equipment setup, booth availability, testing materials); (3) Staff resources; (4) Child's behaviours/compliance; (5) Degree of hearing loss; (6) Type of hearing loss; (7) Caregivers' attitude on UHL; (8) etiology; (9) Child's device fitting status; (10) Others. In addition to the closed response questions, audiologists also had the opportunity to provide comments and specifications on each selected option and additional comments or insights on these questions. The results subsequently used for the following statistical analyses and discussion on the reasons of delaying in getting behavioural audiogram of this report.

- If there is no measured threshold at 0.5 kHz, extrapolate the results by decreasing 10 dB from 1 kHz;
- The missing threshold at 2 kHz is calculated as the mid value between the 1 and 4 kHz;

The three frequencies averaged hearing level (3FAHL) in the impaired ear was then calculated using the thresholds at 0.5, 1, and 2 kHz.

2.2.5. Definition of changes in hearing level

For the purposes of this study, the changes in hearing were determined by comparing the hearing thresholds measured from the first reliable behavioural audiogram to the baseline hearing thresholds at diagnosis. A positive difference between two thresholds indicated a deterioration in hearing, while a negative difference indicated an improvement. To be consistent with recent literature, the following two definitions were adopted for the analysis of significant deterioration:

- Definition 1: a decrease of ≥10 dB at two or more adjacent frequencies between 500 and 4,000 Hz, or a decrease of >15 dB at one octave frequency in the same frequency range (22, 23, 25, 26).
- Definition 2: a change of ≥20 dB in the three frequencies (500, 1,000, and 2,000 Hz) pure-tone average (16, 22, 28).

Additionally, this study employed a similar version of definition 2 to determine a significant improvement in hearing thresholds. Specifically, an increase of greater than or equal to 20 dB (i.e., a change of ≤−20 dB) in the three-frequency average hearing level (3FA HL) was defined as an improvement. Any change that did not meet the criteria for deterioration or improvement (i.e., fell within the range of −20–20 dB in 3FA HL) was categorized as stable hearing loss between the estimated baseline hearing threshold and the first reliable behavioural audiogram.

2.3. Statistical analysis

The main interests of analysis in this study were the age at the first reliable behavioural audiograms and the proportion of children with change of hearing threshold levels. Descriptive statistics, such as mean and standard deviation, median and percentiles, interquartile range were used to report quantitative outcomes. Regression analyses were used to examine relationships between potential factors and the ages at which reliable behavioural audiograms were obtained. The potential reasons for delayed behavioural audiograms in our sample were extracted from clinical notes and classified based on a survey's results (see the footnote in Section 3.2). Pearson's correlation analysis was used to assess the amount of change in hearing thresholds at individual frequencies (0.5–4 kHz) between the baseline hearing thresholds at diagnosis and the hearing thresholds measured from the first reliable behavioural audiogram. A two-way ANOVA test was further conducted to examine the frequency effects on changes in hearing levels. Comparisons were also made between the initial diagnosis results and the first reliable behavioural audiometric results to determine the proportion and extent of hearing changes. Further explorations on the differences in clinical characteristics were performed by comparing children with and without hearing deterioration using Fisher's exact test or Chi-Square analysis, as appropriate. All analyses used two-tailed tests, with statistical significance set at $p < 0.05$. The statistical analysis was performed using IBM SPSS Statistics for Windows v.29 (43).

3. Results

3.1. Demographic characteristics

Table 1 provides descriptive statistics of the demographic characteristics at diagnosis of the sample. The etiology records were extracted from clinical reports or provided diagnosis reports available at Hearing Australia and were carefully reviewed by experienced research audiologist. Among a total of the 91 children, etiology was known for 35 children (38%), and the remaining 56 (62%) children had no reported etiology. Of the 35 children with known etiology, 10 (11%) of them had absent or abnormal auditory nerves, 9 (10%) were born with atresia and/or microtia, 8 (9%) had Cytomegalovirus (CMV), 4 (4%) had inner ear anomalies (enlarged vestibular aqueduct syndrome), 4 (4%) had syndromic hearing loss (1 Down's syndrome, 1 Goldenhar syndrome, 1 Noonan's syndrome, 1 Global developmental delay). Among the 56 children without a reported etiology for their hearing loss, 4 individuals were suspected to have a genetic basis, as one of their parents had a history of hearing loss, suggesting a potential hereditary component to their hearing loss. The distribution of degree of hearing loss at diagnosis in the sample showed that out of the 91 children, 1.1% had a high-frequency mild hearing loss, 16.5% had a mild hearing loss, 13.2% had a moderate hearing loss, 25.3% had a moderate to severe hearing loss, 15.4% had a severe hearing loss, and the remaining 28.6% had a profound hearing loss. The use of hearing aids was also recorded. By the time the children obtained their first reliable behavioural audiogram, 62 out of 91 children were fitted with hearing aids. Another 2 children were fitted with hearing aids within one month after obtaining the first behavioural audiogram. The remaining 27 children were not fitted with hearing aids at the time of the first behavioural audiogram.

3.2. Age when the first reliable behavioural audiogram is obtained

Figure 1 shows a histogram of the age at which the first reliable audiogram was obtained, with a mean age of 3.0 years (SD: 1.5; IQR: 1.8, 4.1). Although the mean age at diagnosis of the sample was 2.1 months old, it has been noted that about half of the children (47.3%) did not obtain their first reliable behavioural audiogram until after 3 years of age. A correlation analysis showed no significant relationship between the degree of hearing

TABLE 1 Demographic characteristics of children with congenital hearing loss ($N = 91$).

Characteristics	Children
Gender	
Male, *n* (%)	46 (50.5%)
Female, *n* (%)	45 (49.5%)
Birthweight, kilograms	
Mean (SD)	3.2 (0.65)
Median	3.3
Interquartile range (IQR)	2.9–3.6
Missing data, *n* (%)	18 (14%)
Gestation, weeks	
Mean (SD)	38.1 (2.8)
Median	39.0
IQR	37–40
Missing data, *n* (%)	16 (17.6%)
Age at diagnosis, months	
Mean (SD)	2.1 (1.2)
Median	1.7
IQR	1.2–2.6
Affected ear at diagnosis	
Left ear, *n* (%)	48 (52.7%)
Right ear, *n* (%)	43 (47.3%)
Etiology, *n* (%)	
Absent/abnormal auditory nerve	10 (11%)
Atresia/Microtia	9 (10%)
Cytomegalovirus (CMV)	8 (9%)
Large vestibular aqueduct syndrome (LVAS)	4 (4%)
Other syndromes	4 (4%)
No reported etiology	56 (62%)
Degree of hearing loss at diagnosis[a], *n* (%)	
High frequency	1 (1.1%)
Mild [20–40 dB]	15 (16.5%)
Moderate [41–55 dB]	12 (13.2%)
Moderate to Severe [56–70 dB]	23 (25.3%)
Severe [71–90 dB]	14 (15.4%)
Profound (>90 dB)	26 (28.6%)
Hearing device fitting, *n* (%)	
Before the first reliable behavioural audiogram	62 (68.1%)
One month within the first reliable behavioural	2 (2.2%)
5 months after the first behavioural audiogram	5 (5.5%)
Not fitted by time of the study	22 (24.2%)

[a]Degree of hearing loss was categorized as: high frequency loss only: ≥25 dB eHL at ≥2 frequencies above 2 kHz (23); mild hearing loss: 20–40 dB; moderate hearing loss: 41–55 dB; moderate severe hearing loss: 56–70 dB; severe hearing loss: 71–90 dB; profound hearing loss: >90 dB (23, 25).

loss (3FA HL) and the age at which the behavioural audiometry was obtained ($p > 0.05$).

To explore the potential factors contributing to the delay in obtaining reliable behavioural audiograms, a multiple regression analysis was conducted. The dependent variable was the age at which the first reliable behavioural audiogram was obtained. The independent variables consisted of 4 clinical variables (presence or absence of reported etiology, degree of hearing loss at diagnosis, ongoing middle ear problems in the impaired ear, and hearing device fitting), as well as 3 additional factors (whether children comply or not with the testing, presence or absence of other faced challenges unrelated to children's behavioural issues, and family availability) (see details Section 2.2. "Factors affecting the age of obtaining reliable behavioural audiograms"). Regarding the hearing device fitting factor, it was categorized into two groups. One group included cases where a hearing aid was fitted before or around the time of obtaining the first reliable audiogram. The other group included cases where a hearing aid was fitted long after the first reliable audiogram or where no hearing devices were fitted at all.

The results in Table 2 revealed a significant regression model [$F(7, 83) = 2.63$, $p = 0.017$] with the full set of predictors accounted for 11.3% of total variance in the age of getting the first reliable audiogram. Among these predictors, children's behavioural issues ($\beta = 12.9$, $p = 0.003$) and the presence or absence of reported ongoing middle ear issues ($\beta = -10.84$, $p = 0.04$) demonstrated significant associations with the dependent variable. Specifically, children with reported behavioural issues were likely to be older when getting their first reliable audiogram. Conversely, those with reported ongoing middle ear issues tended to be younger when obtaining their first reliable audiogram. However, for the remaining predictors, there was insufficient evidence to demonstrate significant associations with the dependent variable.

3.3. Change of hearing threshold levels

3.3.1. Relationship between the hearing thresholds at diagnosis and the first reliable behavioural assessment across tested frequencies

Figure 2 illustrates the relationships between the baseline hearing thresholds of the impaired ear at diagnosis and the values measured at the first reliable behavioural audiogram across tested frequencies. The results depict significant correlations between the estimated hearing threshold (dB eHL) and the measured hearing thresholds (dB HL) from behavioural assessment at each tested frequency ($r > 0.75$, $p < 0.001$). The strongest positive correlation between the two measures was observed at 1 kHz [$r(89) = 0.8$, $p < 0.001$], while the lowest correlation was found at 500 Hz [$r(74) = 0.75$, $p < 0.001$] with greatest variability in hearing changes [mean difference of 14.3; SD: 25.0, IQR: (0, 21.3)]. In Figure 2, the data points above the solid line represent participants whose hearing thresholds were higher in the behavioural testing, indicating a deterioration, while points below the solid line represent an improvement in hearing threshold at the behavioural test.

A two-way ANOVA test was conducted to examine the effects of four tested frequencies (0.5, 1, 2, and 4 kHz) and five age ranges on the dependent variable of hearing level changes. The age ranges considered were under 2 years, 2–3 years, 3–4 years, 4–5 years, and over 5 years, which correspond to the age at which the first reliable behavioural audiogram was obtained. The results revealed a statistically significant difference in the overall changes in hearing thresholds among the four frequencies [$F(3, 307) = 2.83$, $p = 0.039$]. Post-hoc analysis indicated that the changes in hearing levels were significantly higher at 500 Hz compared to

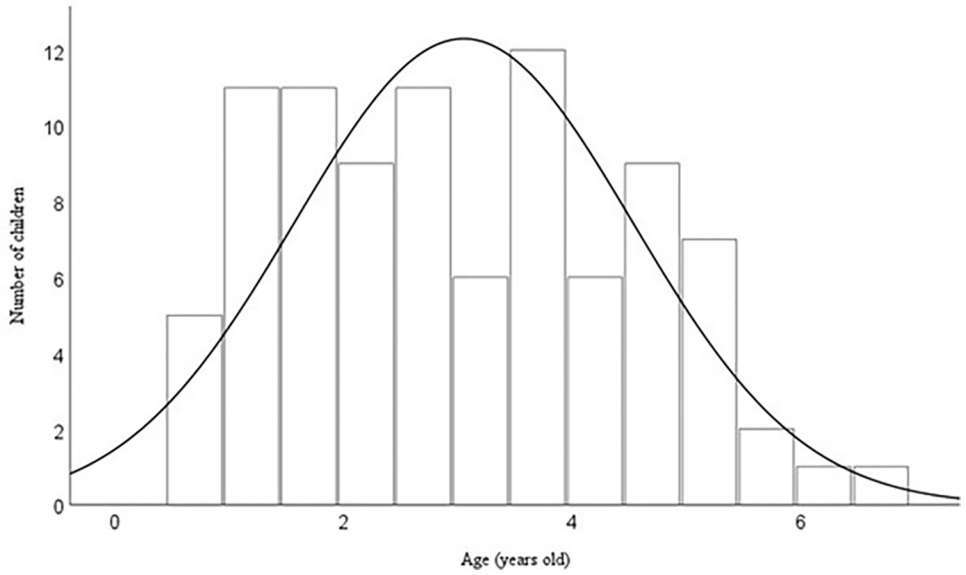

FIGURE 1
Histogram of age (years) at which the first reliable behavioural audiogram was obtained.

TABLE 2 Effect size (unstandardized coefficient estimates Beta-values), 95% confidence intervals (95% CI), and significance levels (*p*-values) of predictor variables for age at first reliable behavioural audiogram (*N* = 91).

	Beta	95% CI	*p*-value
Presence or absence of reported etiology	1.70	(−5.69, 9.09)	0.65
Degree of hearing loss at diagnosis	−0.01	(−0.14, 0.13)	0.94
Ongoing middle ear problems	−10.72	(−20.76, −0.67)	0.04*
Hearing device fitting	−1.44	(−9.54, 6.67)	0.73
Presence or absence of children's behavioural issues	13.12	(4.94, 21.30)	0.002*
Presence or absence of other faced challenges	−4.37	(−19.69, 10.95)	0.58
Family availability	9.23	(−3.02, 21.48)	0.14
Adjusted R^2	0.112		

*Depict significance at 0.05 probability level.

the other frequencies, followed by 1 kHz, 2 kHz, and the lowest changes were observed at 4 kHz. The main effect of age of getting the behavioural audiogram on the changes in hearing levels [$F(4, 307) = 0.83$, $p = 0.51$], and the interaction between frequency range and age [$F(12, 307) = 0.45$, $p = 0.94$] were not statistically significant.

Although there were generally high correlations between the hearing thresholds at two measured points, a considerable number of children exhibited either deterioration or improvement at each frequency. To explore this further, a detailed analysis of hearing level changes at tested frequencies was conducted (as shown in Table 3). The results indicated that majority of children experienced deterioration rather than improvement in the initial impaired ear at each frequency. For instance, at 1,000 Hz, 58 children (63.8%) had hearing deterioration of 10 dB or more, while 12 (13.2%) demonstrated hearing improvement of 10 dB or more. Overall, there were more instances of hearing changes (either deterioration or improvement) in the 500 and 1,000 Hz frequency ranges. Approximately 79% and 77% of ears showed a hearing change of 10 dB or more at 500 and 1,000 Hz, respectively. In contrast, less than 62.5% of ears experienced a hearing change at 2,000 Hz and 4,000 Hz.

By analysing the averaged hearing level changes in children with either deteriorated or improved hearing thresholds, it was observed that although the mean values of deterioration or improvement of thresholds were similar across all tested frequencies, the degree of thresholds difference was in general higher for deterioration than for improvement in these children. For example, at 500 Hz, the mean deterioration in hearing thresholds was 28.9 dB, compared to the mean improvement of 18.9 dB. The statistical analysis with independent *t*-test indicated that these differences were statistically significant at 500 and 2,000 Hz [at 500 Hz, $t(38) = 2.4$, $p = 0.01$; at 2,000 Hz, $t(35.2) = 2.0$, $p = 0.024$].

3.3.2. Examination of the proportion of children who had hearing loss changes

Table 4 shows the proportions of hearing loss changes in the impaired ear. Using definition 1, approximately 78% of the 91 children ($n = 71$) experienced deterioration at the time of the first reliable behavioural audiogram. Among the 71 children with hearing deterioration, 34 of them demonstrated deterioration by 3 years of age. As shown in Figure 3, two children progressed from unilateral to bilateral hearing loss by 3 years of age. Out of the 69 children who had hearing deterioration in the impaired ear only, 53 of them had a decrease in 10 dB or greater at two or more adjacent frequencies between 0.5 and 4 kHz, and the other 16 children had a hearing decrease in 15 dB or greater at one octave frequency in the same frequency range. For the 20 children

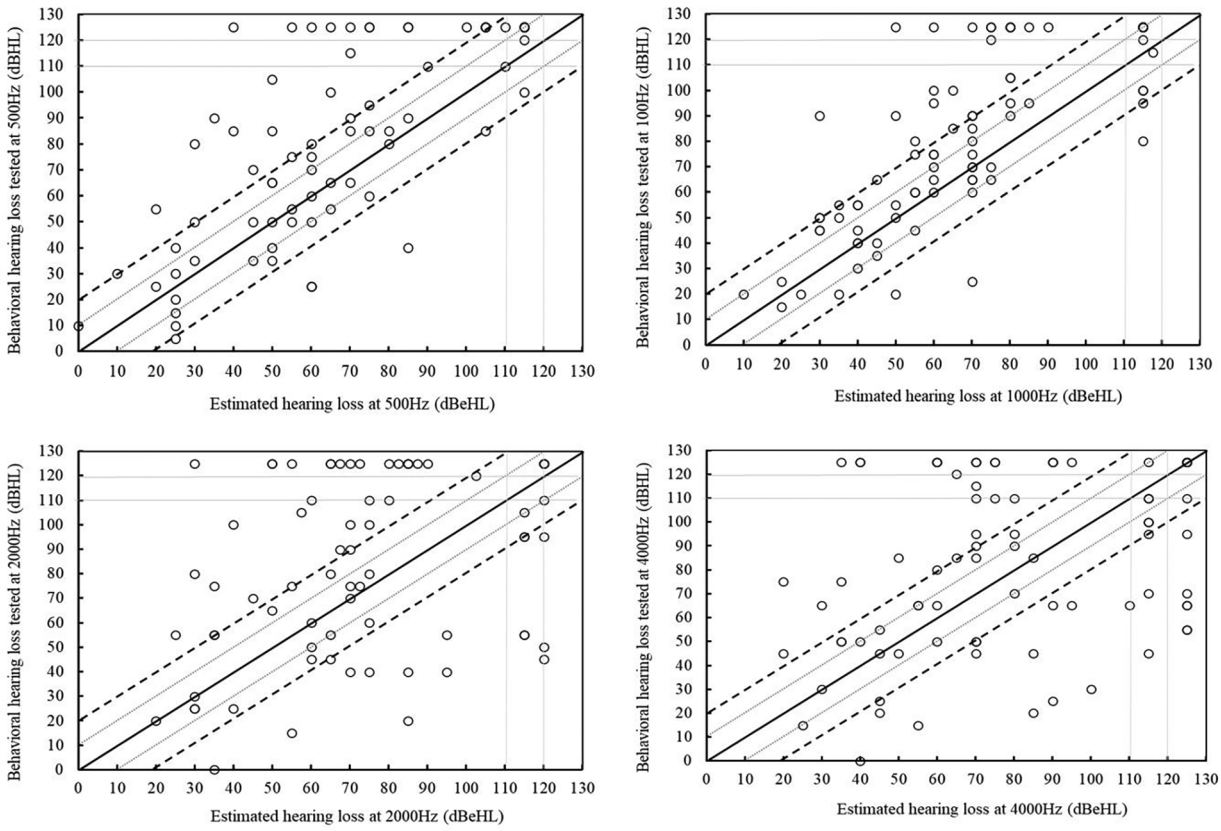

FIGURE 2
Scatter plots showing the estimated threshold at diagnosis (horizontal axes) against the hearing thresholds at the first reliable behavioural assessment (vertical axes) at 500, 1,000, 2,000, and 4,000 Hz. The solid diagonal line shows agreement. Points above the solid line indicate deterioration. The range between the two grey lines depicts +/− 10 dB, and that between the two dotted depicts +/− 20 dB.

TABLE 3 Number (%) of children demonstrating hearing changes and average hearing level change (mean ± SD) at tested frequencies.

		Frequency (Hz)			
		500	1,000	2,000	4,000
Available data points (*n*)		76	91	72	88
Number (%) of children demonstrating hearing change					
Deterioration	≥20 dB	30 (39.5%)	28 (30.8%)	18 (25.0%)	19 (21.6%)
	≥10 and <20 dB	16 (21.1%)	30 (33.0%)	13 (18.1%)	15 (17.0%)
	subtotal	46 (60.6%)	58 (63.8%)	31 (43.1%)	34 (38.6%)
Improvement	≥20 dB	5 (6.6%)	4 (4.4%)	5 (6.9%)	9 (10.2%)
	≥10 and <20 dB	9 (11.8%)	8 (8.8%)	9 (12.5%)	11 (12.5%)
	subtotal	14 (18.4%)	12 (13.2%)	14 (19.4%)	20 (22.7%)
Any hearing change (include deterioration or improvement)	≥20 dB	35 (46.1%)	32 (35.2%)	23 (31.9%)	28 (31.8)
	≥10 and <20 dB	25 (32.9%)	38 (41.8%)	22 (30.6%)	26 (29.5%)
	Total	60 (79%)	70 (77%)	45 (62.5%)	54 (61.4%)
Average change of hearing level in dB					
Deterioration	Mean (SD)	28.9 (19.4)	23.9 (16.9)	28.7 (18.4)	26.1 (16.6)
Improvement	Mean (SD)	18.9 (11.3)	18.8 (11.7)	18.9 (12.9)	22 (12.0)
	Difference *p*-value*	0.011	0.11	0.024	0.15

*Independent Samples *t*-tests were used to compare the average change of hearing level between the two deterioration and improvement groups at each frequency.

who had no hearing deterioration in both ears, two of them had behavioural thresholds within the normal hearing levels in both ears.

As shown in Table 4, when applying definition 2 (i.e., 3FAHL ≥ 20 dB), 26.4% of the children in the sample experienced hearing deteriorated by the time of the first reliable

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TABLE 4 Proportion of hearing loss changes in the impaired ear ($N = 91$).

Definition of progressive hearing loss		Number (%) of children with hearing loss changes in the present study		
		Deterioration (*n*, %)	No deterioration (*n*, %)	
Definition 1	criterion (1): a decrease in 10 dB or greater at two or more adjacent frequencies between 0.5 and 4 kHz, or criterion (2): a decrease in 15 dB or greater at one octave frequency in the same frequency range	71 (78.0%),[Including 55 (60.4%) children who met criterion 1; and an additional 16 (17.6%) children who met criterion 2]	20 (22.0%)	
		Proportion of children progressed to severe to profound loss 52/71 (73.2%)		
		Deterioration (*n*, %)	Stable[a] (*n*, %)	Improvement[b] (*n*, %)
Definition 2	a change of ≥20 dB in the three frequencies (500, 1,000, and 2,000 Hz) pure-tone average	24 (26.4%)	65 (71.4%)	2 (2.2%)
		Proportion of children progressed to severe to profound loss 22/24 (91.7%)		

[a]Definition of stable: the three-frequency (500, 1,000, and 2,000 Hz) average hearing level (3FA HL) of the behavioral results is less than 20 dB difference (either increase or decrease) from the diagnostic results.
[b]Definition of improvement: an increase of greater than or equal to 20 dB (i.e., ≥20 dB improvement) in the 3FA HL.

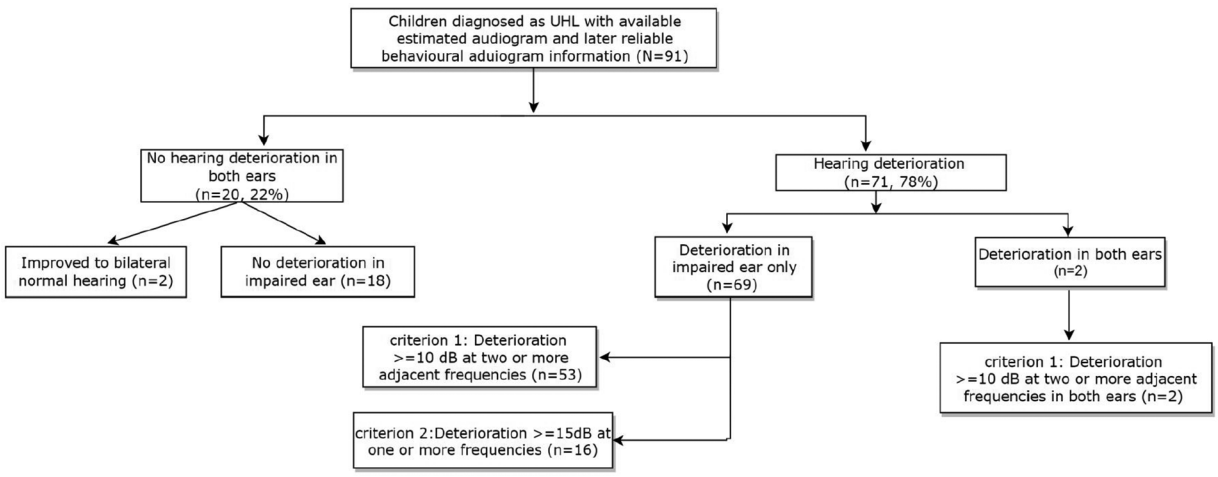

FIGURE 3
Flowchart of the number of children with and without hearing deterioration, including details on the extent of hearing deterioration based on definition 1 for progressive hearing loss.

behavioural audiogram, while 2.2% showed improvement in the hearing levels. The majority of children (71.4%) had stable hearing between the two measurement points. Among those with hearing deterioration ($n = 24$), 23 demonstrated deterioration in the initially impaired ear, and one child was identified as having hearing deteriorated in both ears.

We also observed that a high proportion of children in our study developed severe to profound hearing loss from a milder degree of loss. Specifically, when applying definition 1, out of the total of 71 children who showed progressive hearing loss, 52 of them (73.2%) deteriorated to severe to profound hearing loss, compared to their initial hearing thresholds at the time of diagnosis. When using definition 2, 22 out of the 24 children, an overwhelming majority of them (92%) deteriorated to severe to profound hearing loss during behavioural assessments.

3.3.3. Clinical characteristics of children who had changes in hearing loss

Table 5 showed the breakdown of clinical characteristics and etiology information for children with or without hearing deterioration based on the two different definitions. According to the frequency-specific definition of progressive hearing loss (Definition 1), children with varying degrees of hearing loss at diagnosis may exhibit hearing deterioration in at least one frequency. On the other hand, based on definition 2 (i.e., 3FAHL ≥ 20 dBHL), apart from the moderate ($n = 12$) and severe ($n = 14$) two groups, more than 70% of the children in any other degrees of hearing loss at diagnosis are likely to remain in the stable group. For example, among the 15 children diagnosed with mild hearing loss, 12 of them (80%) showed no hearing deterioration based on definition 2. This is because definition 2

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TABLE 5 Number (proportion) of children with or without hearing deterioration for clinical characteristics and etiology information based on the two definitions.

Characteristics	*N* = 91	Definition 1[a]		Definition 2[b]	
		Deterioration (*n* = 71)	No deterioration (*n* = 20)	Deterioration (*n* = 24)	No deterioration (*n* = 67)
Gender					
Female	45	37	8	14	31
Male	46	34	12	10	36
Hearing loss ear					
Left	48	36	12	15	33
Right	43	35	8	9	34
Degree of HL at diagnosis					
High frequency	1	1	0	0	1
Mild [20-40 dB]	15	11	4	3	12
Moderate [41–55 dB]	12	10	2	10	2
Moderate to Severe [56–70 dB]	23	18	5	7	16
Severe [71–90 dB]	14	11	3	6	8
Profound (>90 dB)	26	20	6	6	20
Hearing device[c]					
Fitted	62	49	13	18	44
Not fitted	29	22	7	6	23
Etiology					
Absent or abnormal auditory nerves	10	8	2	5	5
Atresia/Microtia	9	2	7	0	9
CMV	8	6	2	4	4
LVAS	4	4	0	2	2
Syndromic	4	2	2	0	4
No reported etiology	56	49	7	13	43

[a]Definition 1: criterion (1): a decrease in 10 dB or greater at two or more adjacent frequencies between 0.5 and 4 kHz, or criterion (2): a decrease in 15 dB or greater at one octave frequency in the same frequency range.
[b]Definition 2: a change of ≥20 dB in the three frequencies (500, 1,000, and 2,000 Hz) pure-tone average.
[c]Hearing device fitted or not fitted before the first behavioural audiogram.

may not capture further deterioration in certain frequencies. By excluding all children diagnosed with profound hearing loss, whose hearing loss may have already reached the limits of measured hearing thresholds, 60% (51 out of 85) would demonstrate deterioration according to definition 1, while 27.7% (18 out of 85) would show deterioration based on definition 2.

Furthermore, the deterioration rates were similar between the group of children fitted with hearing devices and the group without hearing devices, based on both definitions. According to Definition 1, 79% (49 out of 62) of the fitted group showed hearing deterioration, while 21% (13 out of 62) remained stable. In the not fitted group, 76% (22 out of 29) demonstrated deterioration, and 24% (7 out 29) remained stable. Similarly, based on Definition 2, the proportions were similar, with 79% of the fitted group experiencing deterioration, and 21% remaining stable. Among the not fitted group, 76% showed deterioration, and 24% remained stable.

Regarding the documented constellation of etiology, using Definition 1, 8 out of 10 children with absent or abnormal auditory nerves, 6 out of the 8 children with CMV, and all the children with LVAS demonstrated further hearing deterioration in at least one frequency. Among the 4 children with syndromic hearing loss, 2 experienced hearing deterioration, while the other 2 did not. The results by using definition 2 showed that the number of children whose documented constellation of etiology (including absent or abnormal auditory nerves, CMV, LVAS) were the same between the two groups of children with and without hearing deterioration. None of the 4 children with syndromic hearing loss showed hearing deterioration. Among the total of 9 children who were born with atresia and/or microtia, both definitions suggested a higher likelihood of stable hearing.

4. Discussion

The primary objective of this study was to report the clinical characteristics in a group of children with congenital unilateral hearing loss. We aimed to gather demographic information, determine the age at which the first reliable behavioural audiogram was obtained, and identify possible factors for any delays in acquiring the audiogram. Another important aspect of the present report was to directly compare the estimated hearing thresholds at the time of diagnosis with the first reliable behavioural audiogram, to investigate the hearing changes between these two measured points. This knowledge will contribute to the development of optimal audiological

management, which will enable clinicians to promptly identify any hearing changes and make necessary adjustments to intervention strategies.

4.1. Demographic characteristics and etiology

The demographic characteristics of our sample demonstrate balanced representations of male and female children, as well as left and right of UHL. The equitable distributions suggest that UHL affects both genders and ear laterality without exhibiting any significant preference or potential biases. Among the 91 children included in our study, the etiology was known for 35 of them (38%). Absent/abnormal auditory nerve ($n = 10$) and ENT malformations (atresia or microtia) ($n = 9$) accounted for approximately half of the cases with known etiology. Among the 8 children with CMV, 2 were diagnosed with mild or moderate hearing loss, whilst the remaining 6 had severe to profound hearing loss. This finding aligns with previous reports indicating that CMV infection is associated with such hearing loss severity (36–38). Considering this significant impact, it underscores the importance of implementing CMV screening and genetic testing for children diagnosed with UHL. In addition, another 8 out of the 91 children (8%) in our sample had UHL associated with LVAS ($n = 4$) or other syndromes ($n = 4$). Among these children, the degree of hearing loss at diagnosis ranged from mild to profound, with a significantly higher percentage of moderate hearing loss observed in children with LVAS (3 out of 4 children with LVAS). Notably, 62% of the children in our sample had no reported etiology. Although this finding aligns with previous research reporting a high proportion of UHL cases with unknown etiology or no reported risk factors (23, 26, 34), the fact that a significant number of cases remain of unknown etiology presents a challenge for clinicians in understanding the underlying causes of UHL and developing targeted intervention strategies for these children.

4.2. Age at first reliable behavioural audiograms

Numerous published guidelines by various international organisations have outlined recommendations of early identification, assessment, and management of children with all forms of hearing loss, including that are UHL [e.g., (44–46)]. The results of this study revealed that 52.7% of the children ($n = 48$) identified with UHL through UNHS had their first reliable behavioural audiograms in the impaired ears by 3 years of age, despite a national pediatric clinical protocol recommending that ear-specific behavioural thresholds at all frequencies from 0.5 to 4 kHz should be obtained by 18 months of age for infants with hearing loss (47). This finding also contradicts the perspectives of clinical paediatric audiologists based on our online survey (see Footnote 1), where 64% of clinicians believed that a behavioural audiogram for individual ears could be reliably measured before 2 years of age, and 83% believed it could be obtained before 3 years of age for children with UHL. The potential reasons for this discrepancy could be related to clinicians' opinions on the clinical management of individual children and prioritizing of other clinical needs or activities for children with certain characteristics. For instance, in the case of children with severe to profound loss or absence of auditory nerve in one ear, clinicians tend to focus on the hearing in the normal ear and middle ear status. Similarly, this also applies to children with unilateral microtia or atresia as the hearing thresholds in the affected ear would not impact device settings.

The regression analysis from this report indicated the effects of degree of hearing loss, etiology, or hearing device fitting were not significant factors influencing the age at which behavioural audiograms were obtained. This differs from the aspects being identified as potential influencing factors by clinicians in an online survey, as detailed in Section **2.2**, "Factors affecting the age of obtaining reliable behavioural audiograms". Instead, younger ages of obtaining the first reliable behavioural audiogram were significantly associated with better child's behaviour and the presence of ongoing middle ear issues in the impaired ear.

Children with middle ear issues tended to have more appointments with clinicians, providing them with more opportunities to have hearing tests at early age. However, ongoing middle ear pathology might also delay the behavioural testing due to the potential for inconsistent test results caused by fluctuations in hearing levels or the need for a recovery period after medical treatment for middle ear conditions. It should be noted that only audiograms taken outside periods of temporary middle ear dysfunction, and meeting other criteria as outlined in Section **2.2**, were considered as reliable for further analysis in this study. This exclusion may underestimate the age at which reliable audiograms could be obtained, as temporary middle ear dysfunction may not always impact hearing thresholds or diagnostic classifications. In addition, the factors identified in this report only accounted for 11% of the total variance in age at behavioural audiometry. Hence, additional research is needed to identify and gain a deeper understanding of the various factors contributing to delays in obtaining reliable behavioural audiograms. In particular, the attitudes of clinicians and parents or caregivers towards management of UHL in young children should be investigated (48–52). Understanding these perspectives will provide insights into how decision is made regarding audiological follow-up appointments, the prioritization of clinical activities, and the perceived importance of complete ear-specific audiograms. This information can help develop targeted strategies to address challenges and ensure children with UHL receive timely and comprehensive hearing rehabilitation.

4.3. Change in hearing threshold levels

Children's hearing thresholds may experience changes over time, which could be partially due to changes in ear canal acoustics (53, 54). However, the magnitude of such changes

because of coupling changes is much smaller than what has been observed in the current findings (42). Past studies underscore the risk of hearing deterioration in children with any degree or type of hearing loss, and the deterioration can occur in one or both ears and its severity can range from mild to severe (16, 22, 29). Moreover, these changes can impact a child's perception of sounds across frequencies, thus affecting their language and learning development and overall functional performance (55–57). Hence, it is crucial to obtain reliable behavioural audiograms at an early age to identify and monitor any hearing changes.

This study indicates that changes in hearing levels were observed in each testing frequency. These changes could be classified as deterioration to improvement and categorised into a significant change (≥20 dB) or minor change (≥10 dB but <20 dB) at each frequency. The mean hearing deterioration for frequencies at 0.5, 1, 2, and 4 kHz ranged from 23.9 to 28.9 dB, while the mean hearing improvement ranged from 18.8 to 22 dB across the same frequencies (see **Table 3**). The analysis of the percentage of children experiencing hearing deterioration and improvement shows a greater proportion of deterioration rather than improvement at each frequency. Moreover, there are more instances of hearing changes in the 500 Hz and 1,000 Hz frequency ranges compared to the 2,000 Hz and 4,000 Hz ranges. These results are not consistent with a recent report by Fitzpatrick et al. (23) that showed similar hearing changes across frequencies. Allocating sufficient time and resources to obtain behavioural hearing thresholds at all frequencies is crucial. The present results suggest the importance of prioritizing the acquisition of low-frequency thresholds, considering the significance of low-frequency hearing for speech understanding, especially in noisy environments (58).

The current study found that 71 (78%) children diagnosed with UHL at birth experienced hearing deterioration between the diagnosis and first behavioural audiogram at one or more frequencies between 0.5 and 4 kHz (definition 1, see **Table 4**). Remarkably, among these 71 children, a high proportion of them (73.2%) developed severe to profound hearing loss (see **Table 4**). Previous studies using the same definition reported a deterioration rate of about 37%–47% of children with UHL when comparing the diagnostic audiogram with the most recent audiometric assessment (22, 23, 26). One possible explanation for the difference in proportions between the current study and previous reports may be attributed to the characteristics of the samples across studies. In Fitzpatrick et al.'s studies (23, 26), only about half of their samples were congenital UHL (i.e., 53.7% were congenital UHL in the 2023 study; 47.2% had congenital UHL in the 2017 study). The recent study (2023) reported that among the congenital UHL group ($n = 95$), 51 (54%) showed progressive HL. Another possible explanation may be that both Fitzpatrick studies compared the diagnostic audiogram with the most recent audiometric assessment. The mean length of time between assessments for children with progressive hearing loss was 64.3 and 50.3 months in the 2023 and 2017 studies, respectively. In contrast, our study only included children with congenital UHL, and we compared the results by examining the initial estimated hearing thresholds with the first behavioural audiogram. This approach allowed us to provide timely support after UNHS, for early identification and intervention in children with hearing deterioration. Using averaged three frequency thresholds (i.e., definition 2) to define deterioration, we only identified 26.4% of children in the sample as having hearing deterioration. This suggests that adopting averaged thresholds to define deterioration may underestimate the proportion of children experiencing hearing changes, which could have direct impact on management decisions.

This study also revealed that among the 48 children who had their first reliable behavioural audiogram before 3 years of age, 34 of them (71%) experienced progression of hearing loss in at least one frequency. This finding also emphasises the importance of monitoring hearing thresholds at all audiometric frequencies after diagnosis through newborn hearing screening. Early identification of deterioration has direct implications for considerations such as hearing device fitting and adjustments, evaluations of the impact of hearing loss on a child's development and determining appropriate intervention.

4.4. Strengths, limitations and future directions

One strength of this study is that it only includes children diagnosed with congenital UHL via UNHS, and it conducts a direct comparison between the initial estimated hearing thresholds from electrophysiological measures with the first reliable behavioural audiogram. Both of these methods have been recognised as the gold standard tests for threshold estimation for young infants (59). In this way, clinicians can promptly identify any hearing changes and adjust intervention plans as soon as needed. Another strength of this study is that the behavioural assessment procedures adhere to a nationally standardised clinical protocol implemented by Hearing Australia, a government-funded hearing service organization. By applying the same clinical protocol and using standard training and equipment, this ensures a relatively consistent and reliable approach in conducting the assessments. The diagnostic protocols are also well-established in audiological diagnostic hospitals/centres across Australia. This ensures the methodological rigour and quality of the data collected. To strive for an early intervention goal of 1-2-3 (i.e., hearing screening by one month of age, audiologic diagnosis by two months of age, and enrolment in early intervention by three months of age), our findings highlight the challenges in obtaining timely behavioural audiograms for children with UHL. The observed delays suggest that additional strategies and resources may be needed to meet the desired early intervention goal.

The current study has some limitations that should be considered. Firstly, the current data were drawn from participants in a research study, which may restrict the generalizability of the results to the general population. Secondly, the behavioural audiogram is obtained retrospectively via the Hearing Australia database, which means that certain factors that could influence the test results, such as children's behavioural

issues, family's engagement, resources or other challenges during the appointment, might not be recorded in the case notes. This could affect the analysis of the potential factors contributing to the delay in obtaining reliable behavioural audiograms. Additionally, the low variance explained by our regression model indicates that there may be other factors affecting the age at which children obtain their first reliable behavioural audiogram that were not considered in our analysis. Future research could explore potential factors, such as parental and clinician attitudes towards UHL, as well as the frequency of attended appointments. These investigations may inform clinical practice and guide early intervention strategies. Thirdly, identifying potential etiology causes of progressive hearing loss in children with UHL is crucial for clinicians to develop targeted interventions. However, a significant challenge arises from the unknown etiology of many cases in our sample, making the clinical characteristics and risks associated with hearing progression in children with UHL still inconclusive due to the lack of specific information. Close collaboration with ENT specialists may be beneficial in addressing this challenge, as it could facilitate a more efficient acquisition of etiological information. Further research is necessary to investigate risk factors associated with hearing progression in this population. Lastly, the age of the first reliable behavioural audiogram depends on the child and family's availability to attend audiological appointments. As a result, the actual age of hearing deterioration may be even younger than what was reported, but the identification of hearing changes would rely on when the behavioural test was conducted. Additionally, this report does not include follow-up behavioural audiograms after the first reliable audiogram. Future investigations will examine the comparison between the first and subsequent reliable behavioural thresholds to capture the hearing changes in children with UHL during early life.

5. Conclusion

The findings from this study contribute to the understanding of the demographic, audiological, and etiological characteristics of children with UHL, highlighting the importance of early monitoring of hearing changes and factors that influence the age at which reliable behavioural audiograms are obtained. By gaining a better understanding of this information and its implications for children's developmental outcomes, clinicians and researchers can strive to optimize early clinical management strategies for children with UHL. Overall, the results indicate that closely monitoring hearing loss after initial diagnosis is essential to ensure optimal interventions are implemented at the earliest age for this target group of children.

Data availability statement

The datasets presented in this article are not readily available because the datasets are not available outside of the research team as per Hearing Australia Ethics approvals. Requests to access the datasets should be directed to the corresponding author (Vicky W. Zhang, Vicky.Zhang@nal.gov.au) and Hearing Australia Human Research Ethics Committee Secretary (Phillip Nakad, Phillip.Nakad@nal.gov.au).

Ethics statement

The studies involving humans were approved by Hearing Australia Human Research Ethics Committee (No. AHHREC2014-28 and No. AHHREC2019-9). The studies were conducted in accordance with the local legislation and institutional requirements. Written informed consent for participation in this study was provided by the participants' legal guardians/next of kin.

Author contributions

VZ: Conceptualization, Investigation, Methodology, Supervision, Project administration, Formal Analysis, Writing – original draft, Writing – review & editing. SH: Data curation, Writing – review & editing. AW: Data curation, Writing – review & editing. CF: Data curation, Writing – review & editing. JO: Data curation, Writing – review & editing. MW: Data curation, Methodology, Writing – review & editing. SM: Data curation, Writing – review & editing. TC: Conceptualization, Investigation, Methodology, Supervision, Writing – review & editing, Funding acquisition.

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Conflict of interest

The authors declare that the research was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

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References

1. Watkin PM, Baldwin M. Confirmation of deafness in infancy. *Arch Dis Child*. (1999) 81(5):380–9. doi: 10.1136/adc.81.5.380

2. Vohr B, Simon P, McDermott C, Kurtzer-White E, Johnson MJ, Topol D. Early hearing screening, detection and intervention (EHDI) in Rhode Island. *Med Health R I*. (2002) 85(12):369–72.

3. Berninger E, Westling B. Outcome of a universal newborn hearing-screening programme based on multiple transient-evoked otoacoustic emissions and clinical brainstem response audiometry. *Acta Otolaryngol*. (2011) 131(7):728–39. doi: 10.3109/00016489.2011.554440

4. Dalzell L, Orlando M, MacDonald M, Berg A, Bradley M, Cacace A, et al. The New York state universal newborn hearing screening demonstration project: ages of hearing loss identification, hearing aid fitting, and enrollment in early intervention. *Ear Hear*. (2000) 21(2):118–30. doi: 10.1097/00003446-200004000-00006

5. Johnson JL, White KR, Widen JE, Gravel JS, James M, Kennalley T, et al. A multicenter evaluation of how many infants with permanent hearing loss pass a two-stage otoacoustic emissions/automated auditory brainstem response newborn hearing screening protocol. *Pediatrics*. (2005) 116:663–72. doi: 10.1542/peds.2004-1688

6. Morton CC, Nance WE. Newborn hearing screening-A silent revolution. *N Engl J Med*. (2006) 354(20):2151–64. doi: 10.1056/NEJMra050700

7. Bess FH, Tharpe AM, Gibler AM. Auditory performance of children with unilateral sensorineural hearing loss. *Ear Hear*. (1986) 7(1):20–6. doi: 10.1097/00003446-198602000-00005

8. Crandell CC. Speech recognition in noise by children with minimal degrees of sensorineural hearing loss. *Ear Hear*. (1993) 14(3):210–6. doi: 10.1097/00003446-199306000-00008

9. Johnson CE, Stein R, Broadway A, Markwalter T. "Minimal" high-frequency hearing loss and schoolage children: speech recognition in a classroom. *Lang Speech Hear Serv Sch*. (1997) 28(1):77–85. doi: 10.1044/0161-1461.2801.77

10. Kenworthy OT, Klee T, Tharpe AM. Speech recognition ability of children with unilateral sensorineural hearing loss as a function of amplification, speech stimuli and listening condition. *Ear Hear*. (1990) 11(4):264–70. doi: 10.1097/00003446-199008000-00003

11. McCreery RW, Walker EA, Spratford M, Oleson J, Bentler R, Holte L, et al. Speech recognition and parent ratings from auditory development questionnaires in children who are hard of hearing. *Ear Hear*. (2015) 36(Suppl 1):60S–75S. doi: 10.1097/AUD.0000000000000213

12. Moore DR, Zobay O, Ferguson MA. Minimal and mild hearing coss in children: association with auditory perception, cognition, and communication problems. *Ear Hear*. (2020) 41(4):720–32. doi: 10.1097/AUD.0000000000000802

13. Ruscetta MN, Arjmand EM, Pratt SR. Speech recognition abilities in noise for children with severe-to-profound unilateral hearing impairment. *Int J Pediatr Otorhinolaryngol*. (2005) 69(6):771–9. doi: 10.1016/j.ijporl.2005.01.010

14. Walker EA, Sapp C, Dallapiazza M, Spratford M, McCreery RW, Oleson JJ. Language and reading outcomes in fourth-grade children with mild hearing loss compared to age-matched hearing peers. *Lang Speech Hear Serv Sch*. (2020) 51(1):17–28. doi: 10.1044/2019_LSHSS-OCHL-19-0015

15. Lieu JE. Speech-language and educational consequences of unilateral hearing loss in children. *Arch Otolaryngol Head Neck Surg*. (2004) 130(5):524–30. doi: 10.1001/archotol.130.5.524

16. Fitzpatrick EM, Whittingham J, Durieux-Smith A. Mild bilateral and unilateral hearing loss in childhood: a 20-year view of hearing characteristics, and audiologic practices before and after newborn hearing screening. *Ear Hear*. (2014) 35(1):10–8. doi: 10.1097/AUD.0b013e31829e1ed9

17. Bagatto MP, Moodie ST, Seewald RC, Bartlett DJ, Scollie SD. A critical review of audiological outcome measures for infants and children. *Trends Amplif*. (2011) 15(1):23–33. doi: 10.1177/1084713811412056

18. Ching TYC, Dillon H, Leigh G, Cupples L. Learning from the longitudinal outcomes of children with hearing impairment (LOCHI) study: summary of 5-year findings and implications. *Int J Audiol*. (2018) 57(sup2):S105–11. doi: 10.1080/14992027.2017.1385865

19. Stapells DR. Frequency-specific threshold assessment in young infants using the transient ABR and the brainstem ASSR. In: Seewald RC, Tharpe AM, editors. *Comprehensive handbook of pediatric audiology*. San Diego: Plural Publishing, Inc. (2011). p. 409–48.

20. Rance G, Roper R, Symons L, Moody LJ, Poulis C, Dourlay M, et al. Hearing threshold estimation in infants using auditory steady-state responses. *J Am Acad Audiol*. (2005) 16(5):291–300. doi: 10.3766/jaaa.16.5.4

21. Stapells DR, Gravel JS, Martin BA. Thresholds for auditory brain stem responses to tones in notched noise from infants and young children with normal hearing or sensorineural hearing loss. *Ear Hear*. (1995) 16(4):361–71. doi: 10.1097/00003446-199508000-00003

22. Barreira-Nielsen C, Fitzpatrick E, Hashem S, Whittingham J, Barrowman N, Aglipay M. Progressive hearing loss in early childhood. *Ear Hear*. (2016) 37(5):e311–21. doi: 10.1097/AUD.0000000000000325

23. Fitzpatrick EM, Nassrallah F, Gaboury I, Whittingham J, Vos B, Coyle D, et al. Trajectory of hearing loss in children with unilateral hearing loss. *Front Pediatr*. (2023) 11:1149477. doi: 10.3389/fped.2023.1149477

24. Baganha S, Marques T, Migueis A. Asymmetric sensorineural hearing loss in children: progression and involvement of the contralateral ear. *Acta Otolaryngol*. (2022) 142(3–4):285–90. doi: 10.1080/00016489.2022.2051602

25. Dahl HH, Ching TY, Hutchison W, Hou S, Seeto M, Sjahalam-King J. Etiology and audiological outcomes at 3 years for 364 children in Australia. *PLoS One*. (2013) 8(3):e59624. doi: 10.1371/journal.pone.0059624

26. Fitzpatrick EM, Al-Essa RS, Whittingham J, Fitzpatrick J. Characteristics of children with unilateral hearing loss. *Int J Audiol*. (2017) 56(11):819–28. doi: 10.1080/14992027.2017.1337938

27. Hura N, Stewart M, Walsh J. Progression of hearing loss and cochlear implantation in large vestibular aqueduct syndrome. *Int J Pediatr Otorhinolaryngol*. (2020) 135:110133. doi: 10.1016/j.ijporl.2020.110133

28. Johansen IR, Hauch AM, Christensen B, Parving A. Longitudinal study of hearing impairment in children. *Int J Pediatr Otorhinolaryngol*. (2004) 68(9):1157–65. doi: 10.1016/j.ijporl.2004.04.010

29. Pittman AL, Stelmachowicz PG. Hearing loss in children and adults: audiometric configuration, asymmetry, and progression. *Ear Hear*. (2003) 24(3):198–205. doi: 10.1097/01.AUD.0000069226.22983.80

30. Berrettini S, Ravecca F, Sellari-Franceschini S, Matteucci F, Siciliano G, Ursino F. Progressive sensorineural hearing loss in childhood. *Pediatr Neurol*. (1999) 20(2):130–6. doi: 10.1016/S0887-8994(98)00123-4

31. Purcell PL, Shinn JR, Coggeshall SS, Phillips G, Paladin A, Sie KCY, et al. Progression of unilateral hearing loss in children with and without ipsilateral cochlear nerve canal stenosis: a hazard analysis. *Otol Neurotol*. (2017) 38(6):e138–e44. doi: 10.1097/MAO.0000000000001452

32. Acke FRE, Van Hoecke H, De Leenheer EMR. Congenital unilateral hearing loss: characteristics and etiological analysis in 121 patients. *Otol Neurotol*. (2021) 42(9):1375–81. doi: 10.1097/MAO.0000000000003248

33. Paul A, Marlin S, Parodi M, Rouillon I, Guerlain J, Pingault V, et al. Unilateral sensorineural hearing loss: medical context and etiology. *Audiol Neurootol*. (2017) 22(2):83–8. doi: 10.1159/000474928

34. Fitzpatrick EM, Lambert L, Whittingham J, Leblanc E. Examination of characteristics and management of children with hearing loss and autism spectrum disorders. *Int J Audiol*. (2014) 53(9):577–86. doi: 10.3109/14992027.2014.903338

35. Levi H, Tell L, Feinmesser M. Progressive hearing loss in hard-of-hearing children. *Audiology*. (1993) 32(2):132–6. doi: 10.3109/00206099309071862

36. Dahle AJ, Fowler KB, Wright JD, Boppana SB, Britt WJ, Pass RF. Longitudinal investigation of hearing disorders in children with congenital cytomegalovirus. *J Am Acad Audiol*. (2000) 11(5):283–90. doi: 10.1055/s-0042-1748054

37. Fowler KB. Congenital cytomegalovirus infection: audiologic outcome. *Clin Infect Dis*. (2013) 57 Suppl 4 (Suppl 4):S182–4. doi: 10.1093/cid/cit609

38. Vos B, Noll D, Whittingham J, Pigeon M, Bagatto M, Fitzpatrick EM. Cytomegalovirus-a risk factor for childhood hearing loss: a systematic review. *Ear Hear*. (2021) 42(6):1447–61. doi: 10.1097/AUD.0000000000001055

39. Kenna MA, Feldman HA, Neault MW, Frangulov A, Wu BL, Fligor B, et al. Audiologic phenotype and progression in GJB2 (connexin 26) hearing loss. *Arch Otolaryngol Head Neck Surg*. (2010) 136(1):81–7. doi: 10.1001/archoto.2009.202

40. Small SA, Stapells DR. Multiple auditory steady-state response thresholds to bone-conduction stimuli in young infants with normal hearing. *Ear Hear*. (2006) 27 (3):219–28. doi: 10.1097/01.aud.0000215974.74293.b9

41. Werff KR V, Prieve BA, Georgantas LM. Infant air and bone conduction tone burst auditory brain stem responses for classification of hearing loss and the relationship to behavioral thresholds. *Ear Hear*. (2009) 30(3):350–68. doi: 10.1097/AUD.0b013e31819f3145

42. Dillon H. *Hearing aids*. 2nd ed. New York: Thieme Medical Publishers, Inc. (2012).

43. IBM Corp. *IBM SPSS statistics for windows (version 29.0)*. Armonk, New York, United States: IBM Corp. (2022).

44. Bagatto M, DesGeorges J, King A, Kitterick P, Laurnagaray D, Lewis D, et al. Consensus practice parameter: audiological assessment and management of unilateral hearing loss in children. *Int J Audiol*. (2019) 58(12):805–15. doi: 10.1080/14992027.2019.1654620

45. American Academy of Pediatrics. Joint committee on infant hearing year 2007 position statement: principles and guidelines for early hearing detection and intervention programs. *Pediatrics*. (2007) 120(4):898–921. doi: 10.1542/peds.2007-2333

46. American Academy of Audiology. Audiologic guidelines for the assessment of hearing in infants and young children. (2012).

47. Hearing Australia. *Infant amplification guidelines (circular 073)*. Available at: https://hearingcomau.sharepoint.com/:w:/r/sites/PoliciesandProcedures/_layouts/15/Doc.aspx?sourcedoc=%7B58F8F421-73CA-4ADC-9668-949332706EA4%7D&file=CLINICAL_073_INFANT%20AMPLIFICATION%20GUIDELINES%20AND%20CI%20REFERRAL_AUDCIRC.doc&action=default&mobileredirect=true&DefaultItemOpen=1 (Accessed 2023 July 22, 2022).

48. Chen PH, Lim TZ. Newborn hearing screening and early auditory-based treatment in Taiwan: action trends of families with children who are hearing impaired. *Int J Audiol*. (2021) 60(7):514–20. doi: 10.1080/14992027.2020.1837970

49. Fitzpatrick EM, Gaboury I, Durieux-Smith A, Coyle D, Whittingham J, Salamatmanesh M, et al. Parent report of amplification use in children with mild bilateral or unilateral hearing loss. *J Am Acad Audiol*. (2019) 30(2):93–102. doi: 10.3766/jaaa.17020

50. Hung YC, Chen PH, Lin TH, Lim TZ. Children with unilateral hearing loss after newborn hearing screening in Taiwan. *Am J Audiol*. (2022) 31(3):646–55. doi: 10.1044/2022_AJA-22-00019

51. Hussain S, Pryce H, Neary A, Hall A. Exploring how parents of children with unilateral hearing loss make habilitation decisions: a qualitative study. *Int J Audiol*. (2021) 60(3):183–90. doi: 10.1080/14992027.2020.1804080

52. Porter A, Sheeran N, Hood M, Creed P. Decision-making following identification of an infant's unilateral hearing loss: parent and professional perspectives. *Int J Pediatr Otorhinolaryngol*. (2021) 148(110822):110822. doi: 10.1016/j.ijporl.2021.110822

53. Bagatto MP, Scollie SD, Seewald RC, Moodie KS, Hoover BM. Real-ear-to-coupler difference predictions as a function of age for two coupling procedures. *J Am Acad Audiol*. (2002) 13(8):407–15. doi: 10.1055/s-0040-1716004

54. Seewald RC, Moodie KS, Sinclair ST, Scollie SD. Predictive validity of a procedure for pediatric hearing instrument fitting. *Am J Audiol*. (1999) 8(2):143–52. doi: 10.1044/1059-0889(1999/017)

55. Ching TY, Hill M, Dillon H. Effect of variations in hearing-aid frequency response on real-life functional performance of children with severe or profound hearing loss. *Int J Audiol*. (2008) 47(8):461–75. doi: 10.1080/14992020802116128

56. Moore BC, Stone MA, Fullgrabe C, Glasberg BR, Puria S. Spectro-temporal characteristics of speech at high frequencies, and the potential for restoration of audibility to people with mild-to-moderate hearing loss. *Ear Hear*. (2008) 29 (6):907–22. doi: 10.1097/AUD.0b013e31818246f6

57. Walker EA. The importance of high-frequency bandwidth on speech and language development in children: a review of patricia stelmachowicz's contributions to pediatric audiology. *Semin Hear*. (2023) 44(Suppl 1):S3–16. doi: 10.1055/s-0043-1764138

58. Moore BC, Stainsby TH, Alcantara JI, Kuhnel V. The effect on speech intelligibility of varying compression time constants in a digital hearing aid. *Int J Audiol*. (2004) 43(7):399–409. doi: 10.1080/14992020400050051

59. The Joint Committee on Infant Hearing. Year 2019 position statement: principles and guidelines for early hearing detection and intervention programs. *J Early Hear Detect Interv*. (2019) 4(2):1–44.

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Ulrika Löfkvist,
Uppsala University, Sweden

REVIEWED BY
Catherine Killan,
University of Nottingham, United Kingdom
Allison Mackey,
Karolinska Institutet (KI), Sweden

*CORRESPONDENCE
Dawna Lewis
✉ dawna.lewis@boystown.org

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Impact of room acoustics and visual cues on speech perception and talker localization by children with mild bilateral or unilateral hearing loss

Dawna Lewis[1,2]*, Sarah Al-Salim[3], Tessa McDermott[1], Andrew Dergan[1] and Ryan W. McCreery[2]

[1]Listening and Learning Laboratory, Boys Town National Research Hospital, Omaha, NE, United States, [2]Auditory Perception and Cognition Laboratory, Boys Town National Research Hospital, Omaha, NE, United States, [3]Clinical Measurement Program, Boys Town National Research Hospital, Omaha, NE, United States

Introduction: This study evaluated the ability of children (8–12 years) with mild bilateral or unilateral hearing loss (MBHL/UHL) listening unaided, or normal hearing (NH) to locate and understand talkers in varying auditory/visual acoustic environments. Potential differences across hearing status were examined.
Methods: Participants heard sentences presented by female talkers from five surrounding locations in varying acoustic environments. A localization-only task included two conditions (auditory only, visually guided auditory) in three acoustic environments (favorable, typical, poor). Participants were asked to locate each talker. A speech perception task included four conditions [auditory-only, visually guided auditory, audiovisual, auditory-only from 0° azimuth (baseline)] in a single acoustic environment. Participants were asked to locate talkers, then repeat what was said.
Results: In the localization-only task, participants were better able to locate talkers and looking times were shorter with visual guidance to talker location. Correct looking was poorest and looking times longest in the poor acoustic environment. There were no significant effects of hearing status/age. In the speech perception task, performance was highest in the audiovisual condition and was better in the visually guided and auditory-only conditions than in the baseline condition. Although audiovisual performance was best overall, children with MBHL or UHL performed more poorly than peers with NH. Better-ear pure-tone averages for children with MBHL had a greater effect on keyword understanding than did poorer-ear pure-tone averages for children with UHL.
Conclusion: Although children could locate talkers more easily and quickly with visual information, finding locations alone did not improve speech perception. Best speech perception occurred in the audiovisual condition; however, poorer performance by children with MBHL or UHL suggested that being able to see talkers did not overcome reduced auditory access. Children with UHL exhibited better speech perception than children with MBHL, supporting benefits of NH in at least one ear.

KEYWORDS

unilateral hearing loss, mild bilateral hearing loss, speech perception, audiovisual, room acoustics

Introduction

Children with mild bilateral hearing loss (MBHL) or unilateral hearing loss (UHL) make up at least 5% of school-age children in the United States (1, 2), representing approximately 2.5 million children from pre-kindergarten through 12th grade (3). Children with MBHL or UHL are typically educated in mainstream classrooms alongside peers with normal hearing (NH), in acoustic environments that often do not meet recommended standards for children with hearing loss (4–8). Spratford et al. (8) tested noise and reverberation in 164 general education classrooms. They reported that 87.3% of the classrooms had unoccupied noise levels above the recommended level of 35 dBA. Reverberation times were above the 0.3 s recommended for classrooms educating children who are deaf/hard of hearing or have other communication issues in 62.2% of classrooms.

In classrooms where children are learning, acoustic environments change often, with a variety of talkers and noise sources around the classroom that fluctuate in level. To hear and understand talkers in these environments, children will need to identify and separate the ones they want to listen to from other voices and sounds in the environment. They also may need to quickly shift their attention among multiple sound sources. Depending on the task, children with MBHL or UHL may perform more poorly than children with NH when attempting to understand speech in noise and reverberation (1, 9–15). However, much of the research examining speech understanding in children with MBHL or UHL has not taken real-world listening conditions into account. Not doing so could result in overestimations of how these children will perform in real listening conditions, which could, in turn, impact the provision of educational services that would support listening and learning for children with MBHL and UHL in general education classrooms (16, 17).

When acoustics make it difficult to hear speech, seeing a talker's face can improve children's speech understanding—a skill that improves with age (18–21). As a result of reduced auditory access, children with MBHL or UHL may depend on these visual cues more than children with NH. Recent work by Lalonde and McCreery (18) revealed that school-age children who were hard of hearing exhibited greater audiovisual benefit for sentence recognition in noise than children with NH. Being able to quickly locate talkers to see their faces may strengthen speech understanding in classrooms with poor acoustics and multiple talkers. However, a challenge exists for children with MBHL or UHL since the effort to locate talkers in the presence of reduced auditory access may use cognitive effort that might otherwise be used for speech understanding and learning.

Although children with MBHL or UHL may demonstrate similar difficulties in speech understanding in noise and reverberation, speech and language development, and academic performance (1, 12, 13, 15, 17, 22–31), the underlying mechanisms for these difficulties are likely to be different. For children with UHL, access to binaural cues can be reduced or absent depending on the degree of hearing loss in the poorer ear. Binaural cues are used for locating and separating auditory signals, benefiting speech understanding in background noise (32). Reduced access to these cues may negatively impact speech understanding in children with UHL, particularly for talkers from the direction of their poorer hearing ear. Children with MBHL have access to binaural cues, particularly when hearing levels are symmetrical across ears. However, they experience reduced access to signals from both ears when compared to listeners with NH or UHL. Poor access to speech signals may hinder how well children with MBHL or UHL understand talkers.

To address the conditions children will experience in the real world, numerous studies since the early 2000s have assessed speech understanding in children with NH and children with MBHL or UHL using complex listening tasks and acoustic conditions (12, 33–37). For example, Griffin et al. (33) reported that even when presentation levels were individualized based on sentence-recognition performance, children with UHL performed more poorly than children with NH on a comprehension task. Lewis et al. (12) used audiovisual tasks of speech understanding designed to simulate plausible listening conditions in a classroom to examine the impact of MBHL and UHL on sentence recognition and comprehension. Children with NH and children with MBHL or UHL were tested using a traditional single talker auditory-only sentence recognition task and an audiovisual comprehension task presented by multiple talkers, both presented from multiple locations. Overall, sentence recognition scores were high across all groups, suggesting little impact of hearing status. For the comprehension task, children with MBHL or UHL performed more poorly than those with NH but there were no differences in performance for the two hearing-loss groups. These findings suggested that complex listening tasks in realistic acoustic environments can negatively affect speech understanding in children with MBHL or UHL to a greater extent than children with NH.

The current study was designed to further examine the ability of children with MBHL or UHL to locate and understand talkers under a range of conditions, with a goal of differentiating performance across hearing status groups using tasks that were less complex than our previous comprehension tasks but more complex than simple sentence recognition tasks.

Visual cues directing a listener to the location of a sound can improve identification of that sound for adults with NH or hearing loss (38, 39). Visually guiding children with MBHL or UHL to the talker's location has the potential to reduce effort required to locate that talker as the acoustic environment varies but has not been examined to date. However, locating a talker, even in adverse acoustics, may not require as much effort as locating that talker and understanding what they are saying. Two tasks were used address children's ability to locate talkers and understand them in complex listening conditions, In a localization-only task, children with NH and children with MBHL or UHL were asked to locate talkers under auditory-only and visually guided auditory conditions in three different acoustic environments that children might experience in classrooms. In a speech perception task, children with NH and children with MBHL or UHL were asked to locate multiple talkers and repeat back what each talker said under varying

auditory and auditory-visual conditions in a single acoustic environment.

This experiment addressed the following research questions.

1. *Does acoustic environment impact the ability of children with MBHL, UHL, or NH to locate talkers in auditory-only* vs. *visually guided conditions and how does performance compare across groups?*
2. *Does acoustic environment impact looking time of children with MBHL, UHL, or NH who correctly locate talkers in auditory-only and visually guided conditions and how does performance compare across groups?*
3. *Do auditory and visual accessibility impact speech perception for children with MBHL, UHL, or NH and how does performance compare across groups?*
4. *For children with MBHL or UHL, do audiological (audibility in better (MBHL) or poorer (UHL) ear) and cognitive (vocabulary, working memory) factors help to explain individual differences in speech perception?*

Methods

Test environment and stimuli

A simulated acoustic environment was created following the procedures described in Valente, et al. (37). The simulated room was acoustically treated with acoustic wall and ceiling tiles, carpeting, and a velour curtain. The unaltered acoustic environment in the test space had a 37.4 dBA LEQ background noise level and a 0.18 s reverberation time (T30 mid). As previously described in Salanger et al. (40), participants were seated in the center of the test space surrounded by stands with five 32-inch high-definition televisions (HDTVs; Samsung Syncmaster 2,433) and loudspeakers [M-Audio Studiophile AV (40)] that were arranged around the participant's location at 0°, 90°, 121°, −121°, and −90° (**Figure 1**). Virtual microphone control [ViMiC (41)], generated the simulated environment. Speech-shaped noise was radiated incoherently through the five loudspeakers. The direct sound and first-order reflections were processed through ViMiC and combined with late reverberation and speech shaped noise to create the simulated acoustic space. The audio signals were positioned in a virtual room model to simulate appropriate source distance, reflections, and reverberation.

A custom-built wireless attitude and heading reference system (AHRS) tracked participants' head movements. Head movements were processed in real time using a microcontroller, to provide attitude and heading solutions as Euler angles over Bluetooth.

Stimuli consisted of 96 five-to-six-word low-predictability sentences, video-recorded by four adult female talkers of American English. The sentences were syntactically correct but semantically incorrect with four keywords each (e.g., "The collar charged the silly cement", "The magic ceilings guess far"; keywords underlined) that were chosen to be within the lexicon of children in the first grade (42).

Clinical assessments

Clinical assessments were administered by audiologists and speech-language pathologists who had experience working with

FIGURE 1
Experimental set-up.

children with hearing loss. Audiometric thresholds were measured by an audiologist in a sound-treated, double-walled booth. The Automated Working Memory Assessment [AWMA (43)] Odd One Out subtest was used to measure visuo-spatial working memory. In this task, the child must first indicate the "odd one out" or different shape from a set of three shapes and then recall the position of the different shape on an empty grid. The number of grids in each sequence increases when the child attains four correct answers in a set of six sequences. The Peabody Picture Vocabulary Test–4 [PPVT-4 (44)] was used to assess receptive vocabulary. In the PPVT-4, the child selects a picture that matches a target word from a set of four choices. Visual acuity of all participants was screened using a Sloan letters chart (45). Participants with prescription glasses or contacts were required to wear them during the screening. To pass the screening, the participant must have had a visual acuity screening threshold of 20/32 or better in both eyes.

Participants

Forty children with NH [21 male (52.5%)], 21 children with MBHL [10 male (47.6%)], and 17 children with UHL [12 male (70.6%)] participated. The number of participants was motivated by a power analysis for main effects by group. Children were included if their age was within three months of the target age range of 8 to 12 years. The mean age for the children with NH was 10.5 years (range: 8.1–13.0). For the children with MBHL, the mean age was 10.3 years (range: 8.1–12.8) and for the children with UHL it was 10.0 years (range: (7.9–13.3). Twenty of the children with NH participated in the localization-only task and 20 participated in the speech recognition task. Although not required, all except two children with MBHL or UHL participated in both tasks; one child with MBHL participated only in the localization-only task and one child with UHL participated only in the speech recognition task.

For the current study, children were considered to have NH if their air-conducted thresholds were 15 dB HL or better at all octave frequencies 250–8,000 Hz in both ears. MBHL was defined as a 4-frequency better-ear pure-tone average (BEPTA;.5, 1, 2, 4 kHz) threshold >20 and ≤45 dB HL or thresholds >25 dB HL at one or more frequencies above 2 kHz in both ears.

For 20 of the children with MBHL, the mean BEPTA was 33.7 dB HL (SD = 7.14). One participant with MBHL had a high-frequency hearing loss, with a BEPTA for the frequencies with hearing loss (6–8 kHz) of 67.5 dB HL. Children with MBHL presented with sensorineural hearing loss in both ears ($n = 16$), conductive hearing loss in both ears ($n = 2$), mixed hearing loss in both ears ($n = 2$), sensorineural hearing loss in one ear and mixed hearing loss in the other ($n = 1$), or undetermined ($n = 1$).

Unilateral hearing loss was defined as a 4-frequency pure-tone average threshold >20 dB HL in the poorer ear (PEPTA) and <20 dB HL in the better ear, or thresholds >25 dB HL at one or more frequencies above 2 kHz and ≤15 dB HL at frequencies below 2 kHz in the poorer ear. Eight children had UHL in the right ear and nine had UHL in the left ear. For 15 of the children with UHL, the PEPTA was 58.7 dB HL (SD = 29.9). One of those participants did not have thresholds in the poorer ear that were within the limits of the audiometer. For analysis purposes, that participant's PEPTA was included as 125 dB HL. Two participants with UHL presented with high-frequency hearing loss. For one of those participants, the PEPTA for the frequencies with hearing loss was 40 dB HL (4 kHz, left), and for the other it was 40 dB HL (3, 6, 8 kHz, right). In the poorer ear, children with UHL presented with sensorineural hearing loss ($n = 9$), conductive hearing loss ($n = 4$), mixed hearing loss ($n = 2$), or undetermined ($n = 2$).

TABLE 1 Audiological characteristics, vocabulary, and working memory for participants with MBHL and UHL.

	MBHL	UHL
Age of identification (months)	M = 36.3	M = 35.8
	Mdn = 36.0	Mdn = 24.0
	Range = 0–108	Range = 1–108
Better ear PTA (MBHL; dB HL)	M = 32.5 Mdn = 32.5	
	Range = 8.8–42.5	
Poorer ear PTA (UHL; dB HL)		M = 58.2
		Mdn = 46.3
		Range = 12.5–125
Fitted with at least 1 HA	17 [81%]	11 [64.7%]
Type of hearing aid	Bilateral BTEs (15)	BTE (10)
	Unilateral BTE (1)	CROS (1)
	Unilateral bone-anchored device (1)	
Age of initial HA fitting (months)	M = 48.3	M = 60.7
	Mdn = 51.5	Mdn = 60.0
	Range = 2–96	Range = 22–108
Language (PPVT)	M = 108.52	M = 112.13
	SD = 14.37	SD = 9.63
Working memory (Odd One Out; AWMA)	M = 110.40	M = 113.31
	SD = 19.68	SD = 13.94

NH, normal hearing; MBHL, mild bilateral hearing loss; UHL, unilateral hearing loss; M, mean; Mdn, median; SD, standard deviation; PTA, pure-tone average; HA, hearing aid; BTE, behind-the-ear; CROS, contralateral routing of signals; PPVT, peabody picture vocabulary test; AWMA, automated working memory test.

Audiological, vocabulary, and working memory characteristics of participants with MBHL and UHL are summarized in Table 1. Age of onset of hearing loss and possible progression of hearing loss for children with MBHL or UHL were not available. Testing was completed without personal hearing aids.

Procedures

For the localization-only and speech perception tasks, sentences were presented randomly by the four talkers from each of the five locations around the listener, at 60 dBA. Conditions were randomized for each task and sentence order and talker within conditions were randomized within tasks.

Looking behavior was monitored using the AHRS to assess both speed and accuracy of localization. Pilot testing determined the minimum angle (in degrees relative to 0° azimuth) at which head turn plus eye turn toward a loudspeaker and screen would allow participants to visualize each of the five screens. Minimum

angles for the four non-zero-degree locations were determined to be ±30° (for loudspeakers at ±90° turning right/left), ±85° (for loudspeakers at ±121° turning right/left).

Participants could move their upper body to allow for more natural looking behaviors. Localization was recorded as angular data in the horizontal plane. Looking accuracy was coded as correct when the participant looked into the region for the loudspeaker/screen of the target talker but did not look past that region. If the participant did not look in the correct region or he/she moved beyond that region, accuracy was coded as incorrect. Looking time was analyzed only for those trials coded as correct for looking accuracy.

Localization-only task

Participants heard sentences presented in two conditions (auditory only, visually guided auditory) and three acoustic environments (favorable, typical, poor). In the auditory-only condition, no visual cues were available. In the visually guided condition, the TV screen located above a loudspeaker illuminated blue if the sentence was presented from that loudspeaker. Acoustic environments were chosen to represent a range of listening environments for classroom listening: *Favorable* (noise = 22 dB signal-to-noise ratio [SNR], reverberation time [T30 mid] = 0.5 s); *Typical* (noise = 6 dB SNR, T30 mid = 0.7 s); and *Poor* (noise = 0 dB SNR, T30 mid = 1.3 s). Listeners were instructed to look at the talker's location as quickly as possible after she began speaking. After locating each talker, participants were required to return to the 0° azimuth position before the next sentence was presented.

Speech perception task

Participants listened to sentences presented under four randomized conditions: (1) auditory-only, (2) visually guided auditory (3) audiovisual, (4) baseline (single location auditory-only at 0° azimuth). In the two auditory-only conditions, no visual cues were available. The visually guided condition was the same as in the localization-only task. For the audiovisual condition, recordings of the talkers were presented on the HDTVs using custom software developed in Max 6.

Reverberation for the speech perception task was T30 mid = 0.6 s and SNRs were 0 dB or 3 dB for children with NH or MBHL/UHL, respectively. The different SNRs for NH vs. MBHL/UHL participants were chosen to allow a range of speech-perception performance levels for all groups without ceiling or floor effects.

For the auditory only, visually guided auditory, and audiovisual conditions, sentences were presented randomly by one of four talkers from each of the five locations around the listener. For the baseline condition, each of the four talkers was presented randomly from the speaker at 0° azimuth. For all conditions except baseline, listeners were asked to locate the talker as quickly as possible as each sentence was presented and then repeat the sentence. After locating each talker, participants were to return to face the 0° azimuth position. For the baseline condition, they were asked to look forward throughout the condition. Responses were scored by number of keywords correct (keyword scoring) and by whether all keywords were correct (sentence scoring). A researcher scored the sentences as they were administered. Responses also were video recorded to allow the researcher to recheck scores.

Statistical methods

Linear mixed effects models were conducted using R Statistical Software [R Core Team, v. 4.1.3 (46)] and the lme4 (47) and lmerTest (48) packages. Figures were created using the ggplot2 package [v.3.3.5 (49)]. Descriptive statistics for each group were calculated. Pearson correlations were calculated for children with MBHL or UHL for variables that were only collected for children with hearing loss including: age of hearing loss identification (in months), audiological (better-ear and poorer-ear PTA for mild bilateral and unilateral participants, respectively), language [PPVT-4 (44)], and working memory [Odd One Out subtest of the AWMA (43)]. All linear mixed effects models included a random intercept for each participant to account for correlations between repeated measures within the same participants. Effects are reported as raw coefficients to support interpretation of effects.

Separate models were used in each experiment to examine percent correct looking, looking time, and speech perception. In the localization-only task, the fixed effects were age (in years), acoustic condition (favorable, typical, and poor), audiovisual cues (auditory-only or visually guided), and hearing group (NH, UHL, and MBHL). The auditory-only, favorable condition was coded as the reference in the localization-only task model. In the speech perception task, the fixed effects were age (in years), condition (auditory-only, visually guided, audiovisual, and single location baseline), and hearing group (NH, UHL, and MBHL) with the single-location baseline coded as the reference condition. For both models, hearing group was coded in contrast to the children with MBHL. Model assumptions were confirmed by examining the normality of the distribution of model residuals. *Post-hoc* tests for significant main effects with multiple comparisons were interpreted with *p*-values adjusted using the False Discovery Rate procedure to control for Type I error rate with multiple comparisons (50).

Results

Does acoustic environment impact the ability of children with MBHL, UHL, or NH to locate talkers in auditory-only vs. *visually guided conditions and how does performance compare across groups?*

The initial analyses addressed percent correct looking by acoustic environment and auditory/visual cues for children with NH, UHL, or MBHL (**Figure 2** and **Table 2**). **Table 3** shows the statistics for the linear mixed effects model for percent correct looking. The main effects of acoustic environment and auditory/visual cues on percent correct looking were significant, but none of the differences between hearing groups or higher-order interactions were statistically significant. *Post-hoc t*-tests showed

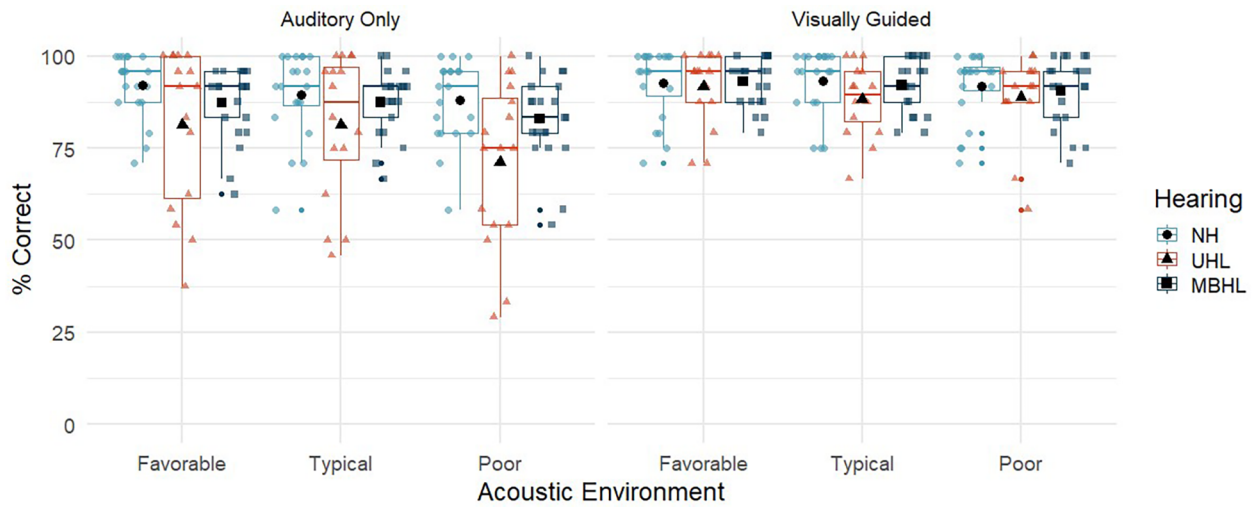

FIGURE 2
Percent correct looking for children with normal hearing (NH; light blue circles), children with unilateral hearing loss (UHL; red triangles) and children with mild bilateral hearing loss (MBHL, dark blue squares) for the three acoustic environments (favorable, typical, poor). Results are shown for the Auditory-Only (left panel) and Visually Guided (right panel) conditions. Boxes represent the interquartile range, and whiskers represent the 5th and 95th percentiles. For each box, lines represent the median. Colored symbols represent individual data points. Black filled symbols represent means.

that percent correct looking in the poor acoustic environment was poorer than in the typical (Coefficient = −5.9, $p < 0.001$) and favorable (Coefficient = −6.8, $p < 0.001$) environments, but the difference between typical and favorable environments was not significant (Coefficient = −0.88, $p = 0.58$). Visually guided conditions had higher percent correct looking than auditory-only conditions (Coefficient = 5.3, $p < 0.001$). The condition by hearing group interaction was not significant (Coefficient = 0.72, $p = 0.58$).

Does acoustic environment impact looking time of children with MBHL, UHL, or NH who correctly locate talkers in auditory-only and visually guided conditions and how does performance compare across groups?

Figure 3 and Table 4 show looking time in seconds for each group across acoustic environments and auditory/visual cues. The pattern of looking time across group and conditions (Table 5) was the same as the percent correct looking results, with *post-hoc t*-tests showing that typical and favorable acoustic conditions were not different (Coefficient = .007, $p = 0.66$) but both had significantly shorter looking time than the poor condition (Coefficient = .08, $p < 0.001$). Visually guided conditions had shorter looking times than auditory-only conditions (Coefficient = −0.17, $p < 0.001$). There were no significant effects of hearing group, age, or higher-order interactions related to looking time in the localization-only task.

Do auditory and visual accessibility impact speech perception for children with MBHL, UHL, or NH and how does performance compare across groups?

Figure 4 and Table 6 show speech perception in percent correct by scoring method (keyword vs. sentence) and conditions (auditory-only, visually guided, audiovisual, and single location

TABLE 2 Mean (standard deviation) for percent correct looking by group and listening condition.

Condition	NH	UHL	MBHL
Favorable (AO)	92.1 (8.7)	81.3 (21.5)	87.3 (9.8)
Favorable (VG)	92.7 (9.6)	91.7 (10.0)	93.1 (6.5)
Typical (AO)	89.4 (11.7)	81.3 (19.7)	87.5 (8.9)
Typical (VG)	93.3 (9.0)	88.3 (9.6)	92.1 (7.2)
Poor (AO)	87.9 (11.1)	71.1 (22.2)	82.9 (11.6)
Poor (VG)	91.9 (9.3)	88.9 (11.6)	90.5 (9.2)

AO, auditory only; VG, visually guided; NH, normal hearing, UHL, unilateral hearing loss; MBHL, mild bilateral hearing loss.

TABLE 3 Linear mixed effects model for group by condition.

Predictors	Percent correct looking		
	Estimates	CI	*p*
(Intercept)	79.41	62.55–96.27	**<0.001**
Poor vs. favorable	−5.92	−9.01 to −2.84	**<0.001**
Typical vs. favorable	−0.88	−3.96 to 2.21	0.576
AO vs. VG	5.26	2.17–8.34	**0.001**
NH vs. MBHL	2.22	−3.50 to 7.94	0.446
UHL vs. MBHL	−4.79	−10.92 to 1.34	0.125
Age (years)	0.83	−0.75 to 2.40	0.303
Condition/AV interaction	3.77	−0.60 to 8.15	0.091
Condition/hearing group interaction	0.72	−2.58 to 3.61	0.58
Random effects			
σ^2	70.10		
$\tau_{00\ ID}$	74.82		
ICC	0.52		
N_{ID}	57		
Observations	341		
Marginal R^2/conditional R^2	0.147/0.588		

AO, auditory only; VG, visually guided; NH, normal hearing, UHL, unilateral hearing loss; MBHL, mild bilateral hearing loss.
Estimates represent the coefficients for each variable in the model. For categorical predictors, the estimate represents the mean difference. For continuous predictors, the estimate represents the change in looking time for a one unit change in the predictor.
All *p*-values for significant effects are bolded.

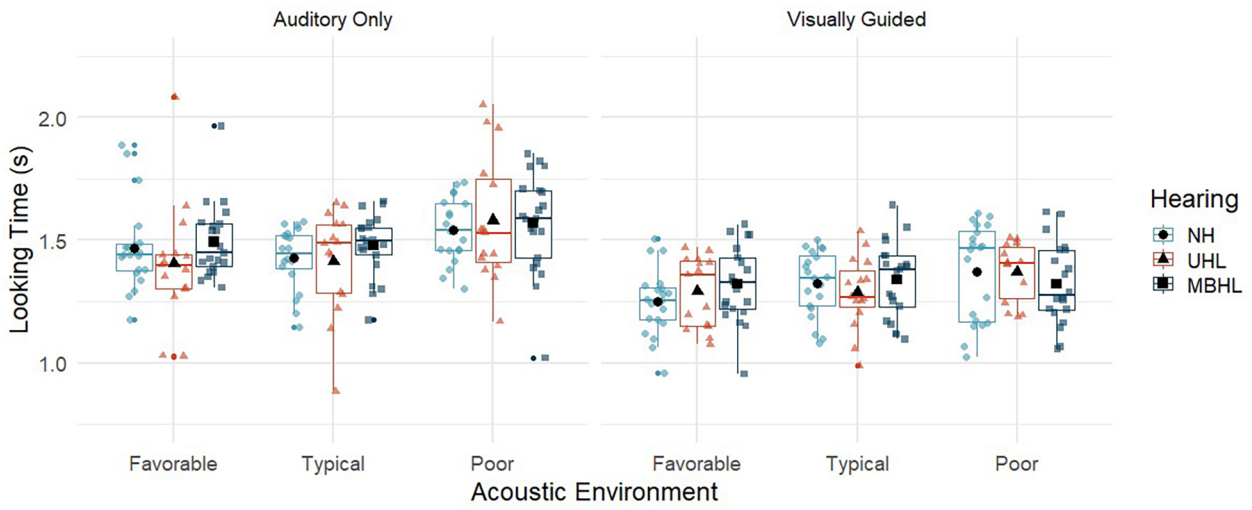

FIGURE 3
Looking time for children with normal hearing (NH; light blue circles), children with unilateral hearing loss (UHL; red triangles) and children with mild bilateral hearing loss (MBHL, dark blue squares) for the three acoustic environments (favorable, typical, poor). Results are shown for the Auditory-Only (left panel) and Visually Guided (right panel) conditions. Boxes represent the interquartile range, and whiskers represent the 5th and 95th percentiles. For each box, lines represent the median. Colored symbols represent individual data points. Black filled symbols represent means.

baseline) for children with NH, UHL, or MBHL. The linear mixed effects models allow for a comparison of two different scoring methods (and their correlation within participants) on the outcome of the models. There could be differences in the model depending on whether the scoring was based on keywords correct or whether the entire sentence was correct. We included a term in the model to account for this potential effect. The main effect indicated that keyword scoring was approximately 30% better than the whole sentence scoring, but that none of the interactions depended on the scoring method. When accounting for this effect, we use the term speech recognition because the main effects of other variables reflect an overall composite of keyword and whole sentence scores for each participant. This can be interpreted that the main effects of group and condition were the same regardless of how the sentences were scored.

Table 7 shows the statistics for the linear mixed effects model for speech recognition. The main effects of condition, hearing group, age, and scoring, and the condition by hearing group interaction were statistically significant. Percent correct looking was not associated with speech recognition. For every one-year increase in age, there was a 3.4% increase in speech recognition. *Post-hoc* *t*-tests were used to assess the effects of condition, hearing group and their interaction. Children with NH had speech recognition that was 27.1% higher ($p < 0.001$) than children with MBHL and 13.9% higher ($p = 0.005$) than children with UHL across conditions. Children with UHL had speech recognition that was 13.2% higher ($p = 0.004$) than children with MBHL across conditions. For each listening condition, the *post-hoc* tests were conducted in reference to the single location baseline condition, which was the condition with the poorest

TABLE 4 Mean (standard deviation) for looking time (seconds) by group and listening condition.

Condition	NH	UHL	MBHL
Favorable (AO)	1.5 (0.2)	1.4 (0.3)	1.5 (0.2)
Favorable (VG)	1.3 (0.1)	1.3 (0.1)	1.3 (0.2)
Typical (AO)	1.4 (0.1)	1.4 (0.2)	1.5 (0.1)
Typical (VG)	1.3 (0.1)	1.3 (0.1)	1.3 (0.2)
Poor (AO)	1.5 (0.1)	1.6 (0.3)	1.6 (0.2)
Poor (VG)	1.4 (0.2)	1.4 (0.1)	1.3 (0.2)

AO, auditory only; VG, visually guided; NH, normal hearing, UHL, unilateral hearing loss; MBHL, mild bilateral hearing loss.

TABLE 5 Linear mixed effects model for group by condition.

	Looking time in seconds		
Predictors	Estimates	CI	*p*
(Intercept)	1.55	1.33–1.77	**<0.001**
Poor vs. favorable	0.08	0.05–0.12	**<0.001**
Typical vs. favorable	0.01	−0.03 to 0.04	0.657
AO vs. AV	−0.17	−0.20 to −0.14	**<0.001**
NH vs. MBHL	−0.02	−0.10 to 0.05	0.529
UHL vs. MBHL	−0.04	−0.12 to 0.04	0.377
Age (years)	−0.01	−0.03 to 0.01	0.479
Random effects			
σ^2	0.02		
$\tau_{00\ ID}$	0.01		
ICC	0.39		
$N_{\ ID}$	56		
Observations	334		
Marginal R^2/conditional R^2	0.237/0.536		

AO, auditory only; VG, visually guided; NH, normal hearing, UHL, unilateral hearing loss; MBHL, mild bilateral hearing loss.
Estimates represent the coefficients for each variable in the model. For categorical predictors, the estimate represents the mean difference. For continuous predictors, the estimate represents the change in speech recognition for a one unit change in the predictor.
All *p*-values for significant effects are bolded.

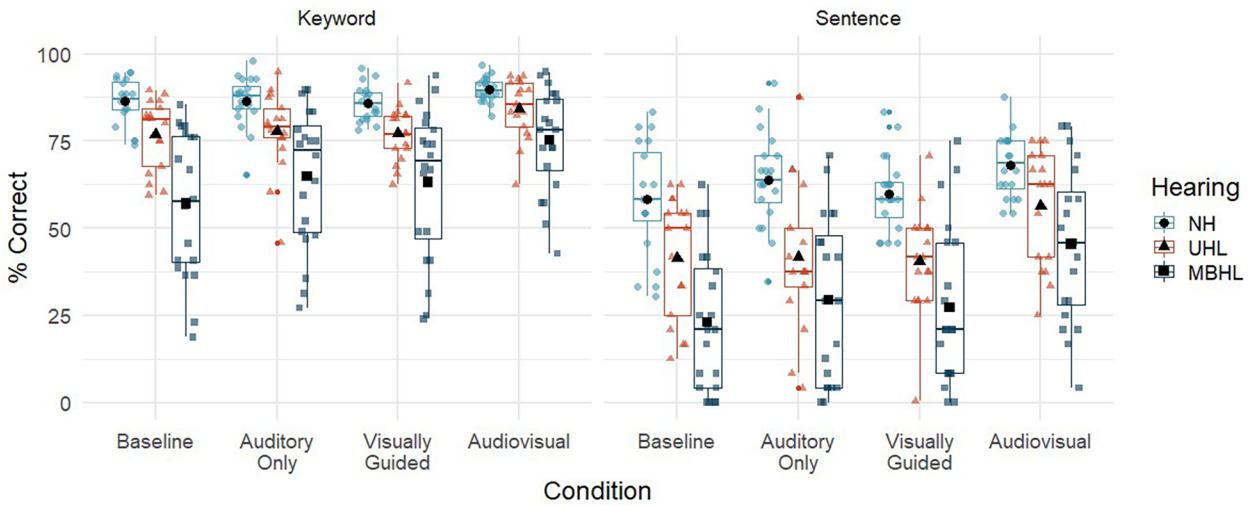

FIGURE 4
Speech perception (% correct) for children with normal hearing (NH; light blue circles), children with unilateral hearing loss (UHL; red triangles) and children with mild bilateral hearing loss (MBHL, dark blue squares) for the four listening conditions (auditory-only, visually guided, audiovisual, baseline). Results are shown for scoring by keyword (left panel) and sentence (right panel). Boxes represent the interquartile range, and whiskers represent the 5th and 95th percentiles. For each box, lines represent the median. Colored symbols represent individual data points. Black filled symbols represent means.

speech recognition across groups. Speech recognition in the auditory-only (+7.1%, $p < 0.001$) and visually guided (+5.3%, $p < 0.001$) conditions was significantly higher than the baseline condition. Speech recognition in the audiovisual condition was higher than the baseline (+20.3%, $p < 0.001$), auditory-only (+13.3, $p < 0.001$) and visually guided (+15%, $p < 0.001$) conditions. The significant interaction between hearing group and condition was driven by a larger difference in speech recognition between children with NH and children with MBHL in the audiovisual and baseline conditions than between children with NH and children with UHL in those conditions.

For children with MBHL or UHL, do audiological (audibility in better (MBHL) or poorer (UHL) ear) and cognitive (vocabulary, working memory) factors help to explain individual differences in speech perception?

TABLE 6 Mean (standard deviation) for speech perception (%) by scoring method (keyword, sentence), group (NH, UHL, MBHL) and listening condition (baseline, AO, VG, AV).

Condition	NH	UHL	MBHL
Keywords			
Baseline	86.4 (6.5)	77.0 (10.1)	57.1 (20.8)
AO	86.5 (7.1)	77.8 (11.6)	64.8 (20.0)
VG	85.9 (4.9)	77.0 (7.8)	63.2 (21.7)
AV	89.8 (3.5)	84.3 (8.7)	75.3 (14.9)
Sentences			
Baseline	58.2 (16.1)	41.4 (17.6)	22.9 (20.3)
AO	63.9 (13.3)	41.7 (21.1)	29.4 (24.0)
VG	59.9 (10.6)	40.5 (15.4)	27.3 (23.0)
AV	68.1 (9.1)	56.4 (16.7)	45.4 (22.3)

AO, auditory only; VG, visually guided; AV, audiovisual; NH, normal hearing, UHL, unilateral hearing loss; MBHL, mild bilateral hearing loss.

To examine the factors that led to individual differences in keyword recognition for children with UHL or MBHL, a separate linear mixed effects model was constructed with the same

TABLE 7 Linear mixed effects models for group by condition.

Predictors	Sentences and keywords correct		
	Estimates	CI	*p*
(Intercept)	27.14	5.06–49.22	**0.016**
NH vs. MBHL	27.00	18.52–35.48	**<0.001**
UHL vs. MBHL	13.26	4.40–22.11	**0.003**
AV vs. AO	13.00	8.55–17.45	**<0.001**
Baseline vs. AO	−7.39	−11.98 to −2.80	**0.002**
VG vs. AO	−1.96	−6.09 to 2.17	0.351
Age (years)	3.40	1.36–5.43	**0.001**
Scoring	−30.65	−32.35 to −28.95	**<0.001**
Correct Looking	0.01	−0.07 to 0.09	0.784
NH AV vs. MBHL AV	−9.45	−15.19 to −3.71	**0.001**
UHL AV vs. MBHL AV	−2.72	−8.70 to 3.27	0.373
NH baseline vs. MBHL baseline	4.22	−1.52 to 9.97	0.149
UHL baseline vs. MBHL baseline	6.44	0.40–12.48	**0.037**
NH VG vs. MBHL VG	−0.45	−6.18 to 5.28	0.877
UHL VG vs. MBHL VG	0.81	−5.19 to 6.81	0.791
Random effects			
σ^2	84.95		
$\tau_{00\ subid}$	142.05		
ICC	0.63		
N_{subid}	57		
Observations	456		
Marginal R^2/conditional R^2	0.649/0.868		

AO, auditory only; VG, visually guided; AV, audiovisual; NH, normal hearing, UHL, unilateral hearing loss; MBHL, mild bilateral hearing loss.
Estimates represent the coefficients for each variable in the model. For categorical predictors, the estimate represents the mean difference. For continuous predictors, the estimate represents the change in speech recognition for a one unit change in the predictor.
All *p*-values for significant effects are bolded.

TABLE 8 Linear mixed effects model for children with MBHL or UHL.

Predictors	Keyword recognition for MBHL and UHL		
	Estimates	CI	*p*
(Intercept)	78.60	29.84–127.36	**0.002**
degree	−1.87	−2.57 to −1.17	**<0.001**
UHL vs. MBHL	−49.87	−76.02 to −23.72	**<0.001**
PPVT	0.18	−0.16 to 0.51	0.298
AWMAOdd	0.12	−0.13 to 0.37	0.331
Degree * hearing status [UHL]	1.86	1.14–2.58	**<0.001**
Random effects			
σ^2	468.07		
$\tau_{00\ subid}$	73.46		
ICC	0.14		
N_{subid}	37		
Observations	296		
Marginal R^2/conditional R^2	0.244/0.346		

UHL, unilateral hearing loss; MBHL, mild bilateral hearing loss; PPVT, Peabody Picture Vocabulary Test; AWMAOdd, Odd One Out subtest for the Automated Working Memory Assessment. Estimates represent the coefficients for each variable in the model. For categorical predictors, the estimate represents the mean difference. For continuous predictors, the estimate represents the change in keyword recognition for a one unit change in the predictor.
All *p*-values for significant effects are bolded.

structure as the full model that included children with NH, but also included audiological variables, vocabulary, and working memory (**Table 8**). Degree of hearing loss was represented as the better-ear pure tone average for the children with MBHL and the poorer-ear pure tone average for the children with UHL. The main effects of this model mirrored the full model including children with NH. The degree of hearing loss was significantly related to keyword recognition, but there was a significant interaction with hearing group that suggested the pattern of degree of hearing loss and keyword recognition was different between children with MBHL and children with UHL. Specifically, the effect of degree of hearing loss on keyword recognition was stronger for children with MBHL than children with UHL. None of the other audiological factors, vocabulary, or working memory had a significant relationship with keyword recognition after controlling for other factors.

Discussion

The current study examined the impact of MBHL or UHL on children's ability to locate and understand talkers under a range of acoustic and auditory/visual conditions. Identifying potential differences in performance across hearing status groups may help to guide intervention for these children.

The localization-only task addressed the ability of children with NH and children with MBHL or UHL to locate talkers who were presented auditory only or with a visual guide to the talker's location in three acoustic environments that children might experience in classrooms. Overall, children were better able to correctly locate talkers in the visually guided condition than in the auditory-only condition. This finding is consistent with findings from adults (38, 39), suggesting that visual information about a talker's location can improve localization of that talker for children with NH, MBHL, or UHL. There were no effects of age on looking behavior, suggesting that the children in the age range studied here were similarly adept at locating the talkers. The impact of acoustics on looking behavior were mixed. Overall, children correctly located talkers least in the poorest acoustic environment. The absence of a difference between typical vs. favorable acoustics suggests that children may be able to tolerate a range of acoustic environments without impacting their ability to find talkers in environments similar to the ones simulated in the current study. There also was no effect of hearing status. The absence of this effect was somewhat surprising, particularly for the poor acoustic condition where auditory access would be expected to have a greater impact on the two hearing-loss groups than on children with NH. However, the results suggest that even with reduced audibility, children with MBHL or UHL exhibited similar abilities to their peers with NH when attempting to locate talkers, suggesting that the task was not more difficult for them even with poorer auditory access. Although average percent correct looking scores were not significantly different across the three groups, the pattern of scores for the children with UHL in the auditory-only condition (see **Figure 2**), suggests a greater negative effect for some of these children when visual cues were unavailable. Studies using a greater variety of acoustic conditions and talker locations could be helpful in further differentiating potential hearing status effects on looking behaviors.

When children correctly located talkers, their looking times followed the same patterns as the correct looking scores. Looking times were shorter for visually guided than for auditory-only conditions and were longer in the poor auditory environment than in the typical and favorable environments. There were no effects of age or hearing group. Even with reduced auditory access, children with MBHL or UHL may not take longer to locate talkers than children with NH during some listening tasks.

Localization-only results suggest that children can benefit from the addition of visual information that guides them to talker locations across varying acoustic environments often found in educational settings, particularly in poor acoustics. Modifications as simple as having the teacher point to students who are raising their hands can give other children the opportunity to locate a particular talker before they speak. It also could be helpful to arrange desks in such a way that talkers are easily located (e.g., positioning in an arc rather than rows).

The speech perception task examined the ability of children with NH and children with MBHL or UHL to both locate multiple talkers and repeat back what those talkers said under varying auditory/visual conditions in a single acoustic environment. This task was not expected to be as difficult as the comprehension task used in our earlier study (12). However, it had the potential to address differences between children with MBHL and UHL that may have been masked by the difficulty of a complex comprehension task. It was anticipated that the syntactically correct/semantically incorrect sentences used in the current study would provide an additional level of difficulty over

previous findings that used sentences that were both syntactically and semantically correct, and that differences in SNRs for children with NH vs. children with HL would avoid floor and ceiling effects for speech recognition.

Overall, the findings support other studies that have shown that speech understanding in noise for children with NH and children with hearing loss improves with age (51–54). Children's speech recognition was highest in the audiovisual condition and lowest in the baseline (single location at 0° azimuth) condition. Speech recognition was better in the visually guided and auditory-only conditions than in the baseline condition but providing visual guidance to talker location did not improve speech recognition over auditory-only presentations. There was no significant effect of correct looking on speech recognition. These findings suggest that being able to find talkers more quickly does not necessarily result in better speech understanding if individuals do not see the talkers speaking once they have been located. It is possible that benefits of visual guidance for locating talkers will vary with the task. In tasks with high cognitive load, for example, visual guidance and audiovisual input could work together to improve speech understanding. Additional research would be needed to address this issue.

Poorer speech recognition in the baseline relative to the auditory-only condition was unexpected. In the baseline condition there was no need to locate talkers before repeating the sentences, potentially resulting in less listening effort than when talkers were in multiple locations. It is possible that children were less attentive in this condition, which they may have expected to be easier, than in conditions where they were required to find talkers. However, this could not be verified in the current study. Further research with this specific set of conditions and methodology is needed to address the issue.

As previously noted, the number of participants was motivated by a power analysis for main effects by group; however, we did not conduct a power analysis to determine how many participants would be required for group by condition interactions. Thus, it is possible that we may be underpowered for those comparisons. Many of the statistically significant effects observed in this study were small to medium effect sizes, suggesting there was sufficient power to address the research questions of interest.

Despite listening to speech at a poorer SNR, children with NH demonstrated better speech recognition than either children with MBHL or UHL. Seeing the talkers improved speech recognition for all groups, but children with MBHL or UHL continued to perform more poorly than their peers with NH, even in the audiovisual condition. These findings suggest that being able to see talkers as they are speaking is beneficial, but not sufficient to overcome reduced auditory access for children with MBHL or UHL. Children with UHL performed better than children with MBHL. In the current study, NH in one ear provided benefit for speech recognition in complex conditions that was not available for children with mild hearing loss in both ears.

Factors that may impact individual differences in speech understanding were examined for the children with MBHL or UHL. Only degree of hearing loss was shown to have a significant effect. Degree of hearing loss in the better ear of children with MBHL had a greater impact on keyword understanding than did degree of hearing loss in the poorer ear for children with UHL. This occurred despite a better mean and smaller range of BEPTAs for the children with MBHL than for PEPTAs in the children with UHL (see **Table 1**). These findings support the benefit of NH on speech perception, even when that NH occurs in only one ear.

Previous research has suggested that degree of hearing loss in the poorer hearing ear may impact speech perception and localization abilities in children with UHL (23). Although population-based studies show poorer ear thresholds in children with UHL are equally represented across a wide range of severity levels (55, 56), the hearing loss levels of participants in individual studies, including the current study, may not include similar numbers of children representing this wide range of severity levels (57). Further research that includes a larger number of children across a representative range of severity for the poorer ear is needed to further address how degree of hearing loss in the poorer ear may differentially impact outcomes in children with UHL.

Hearing aids may improve auditory access for children with MBHL or UHL (58–60); however, there is currently no clear consensus regarding personal amplification recommendations for these populations and both hearing aid recommendations and hearing aid use may be delayed and/or inconsistent (55, 61–64). In the current study, all children were tested without amplification to represent potential worst-case outcomes based on hearing status. Future studies in complex conditions reflecting real-world listening should include measures with amplification to address how improving audibility, in both ears for children with MBHL or one ear for children with UHL who are able to use a hearing aid in the poorer hearing ear, can impact outcomes. Such studies should also examine consistency of hearing aid use in children who are fitted with personal amplification to determine potential effects on outcomes.

Author's note

The content of this manuscript is the responsibility and opinions of the authors and does not necessarily represent the views of the National Institutes of Health.

Data availability statement

The data presented in this article are not readily available as per ethics approvals. Requests to access the data should be directed toward the corresponding author.

Ethics statement

This study was approved by the Institutional Review Board for Boys Town National Research Hospital (08-06 XP). Informed Consent to participate in this was provided by the participants' legal guardians. In addition, participants provided informed assent. Participants were paid $15 per hour for their participation and received a book at the end of the study.

Author contributions

DL conceived the overall project, developed the methods and procedures with Timothy Vallier (see Acknowledgements), and provided input throughout the study. TM, AD, and SA-S managed data collection and data entry. RM oversaw and conducted statistical analysis and interpretation of the analyses. DL drafted the first version of the manuscript. DL, RM, SA-S, TM, and AD contributed to review and editing. All authors contributed to the article and approved the submitted version.

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Conflict of interest

The authors declare that the research was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

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References

1. Bess F, Dodd-Murphy J, Parker R. Children with minimal sensorineural hearing loss: prevalence, educational performance, and functional status. *Ear Hear*. (1998) 19:339–54. doi: 10.1097/00003446-199810000-00001

2. Niskar A, Kieszak S, Holmes A, Esteban E, Rubin C, Brody D. Prevalence of hearing loss among children 6 to 19 years of age. *J Amer Med Assoc*. (1998) 279:1071–5. doi: 10.1001/jama.279.14.1071

3. National Center for Education Statistics. *Fast Facts: Back-to-school statistics*. National Center for Educational Statistics (2023). Available at: https://nces.ed.gov/fastfacts/display.asp?id=372#K12-enrollment (Accessed June 5, 2023).

4. American National Standards Institute. *12.60–2010/Part 1, acoustical performance criteria, design requirements and guidelines for schools, part 1: Permanent schools*. New York: Acoustical Society of America (2010).

5. Crukley J, Scollie S, Parsa V. An exploration of non-quiet listening at school. *J Educ Audiol*. (2011) 17:23–35.

6. Gremp MA, Easterbrooks SR. A descriptive analysis of noise in classrooms across the US and Canada for children who are deaf and hard of hearing. *Volta Rev*. (2018) 117(1–2):5–31. doi: 10.17955/tvr.117.1.2.781

7. Ronsse L, Wang L. Relationships between unoccupied classroom acoustical conditions and elementary student achievement measured in Eastern Nebraska. *J Acoust Soc Am* (2013), 133(3):1480–95. doi: 10.1121/1.4789356

8. Spratford M, Walker E, McCreery R. Use of an application to verify classroom acoustic recommendations for children who are hard of hearing in a general education setting. *Am Jl Audiol*. (2019) 28:927–34. doi: 10.1044/2019_AJA-19-0041

9. Bess F, Tharpe AM. Case history data on unilaterally hearing-impaired children. *Ear Hear*. (1986) 7:14–9. doi: 10.1097/00003446-198602000-00004

10. Crandell C. Speech recognition in noise by children with minimal degrees of sensorineural hearing loss. *Ear Hear*. (1993) 14:210–6. doi: 10.1097/00003446-199306000-00008

11. Griffin AM, Poissant S, Freyman RL. Speech-in-noise and quality-of-life measures in school-aged children with normal hearing and with unilateral hearing loss. *Ear Hear*. (2019) 40:887–904. doi: 10.1097/AUD.0000000000000667

12. Lewis D, Valente DL, Spalding J. Effect of minimal/mild hearing loss on children's speech understanding in a simulated classroom. *Ear Hear*. (2015) 36:136–44. doi: 10.1097/AUD.0000000000000092

13. Lewis D, Smith N, Spalding J, Valente DL. Looking behavior and audiovisual speech understanding in children with normal hearing and children with mild bilateral or unilateral hearing loss. *Ear & Hear*. (2018) 39:783–94. doi: 10.1097/AUD.0000000000000534

14. McCreery RW, Walker EA, Spratford M, Oleson J, Bentler R, Holte L, Roush P. Speech recognition and parent ratings from auditory development questionnaires in children who are hard of hearing. *Ear Hear* (2015) 26(suppl. 1):60S–75S. doi: 10.1097/AUD.0000000000000213

15. Moore DR, Zobay O, Ferguson MA. Minimal and mild hearing loss in children: association with auditory perception, cognition, and communication problems. *Ear Hear*. (2020) 41:720–32. doi: 10.1097/AUD.0000000000000802

16. Yoshinaga-Itano C, DeConde Johnson C, Carpenter K, Stredler Brown A. Outcomes of children with mild bilateral hearing loss and unilateral hearing loss. *Sem Hear*. (2008) 29:196–211. doi: 10.1055/s-2008-1075826

17. Lieu JEC, Tye-Murray N, Fu Q. Longitudinal study of children with unilateral hearing loss. *Laryngoscope*. (2012) 211:2088–95. doi: 10.1002/lary.23454

18. Lalonde K, McCreery RW. Audiovisual enhancement of speech perception in noise by school-age children who are hard of hearing. *Ear Hear*. (2020) 41:705–19. doi: 10.1097/AUD.0000000000000830

19. Massaro DM, Thompson LA, Barron B, Laren E. Developmental changes in visual and auditory contributions to speech perception. *J Exper Child Psychol*. (1986) 41:93–116. doi: 10.1016/0022-0965(86)90053-6

20. Ross LA, Molholm S, Blanco D, Gomez-Ramirez M, Saint-Amour D, Foxe JJ. The development of multisensory speech perception continues into the late childhood years. *Eur J Neurosci*. (2011) 33:2329–37. doi: 10.1111/j.1460-9568.2011.07685.x

21. Wightman F, Kistler D, Brungart D. Informational masking of speech in children: auditory-visual integration. *J Acoust Soc Am*. (2006) 119(6):3940–9. doi: 10.1121/1.2195121

22. Bess FH, Klee T, Culbertson JL. Identification, assessment, and management of children with unilateral sensorineural hearing loss. *Ear Hear*. (1986) 7:43–51. doi: 10.1097/00003446-198602000-00008

23. Bess F, Tharpe AM, Gibler AM. Auditory performance of children with unilateral sensorineural hearing loss. *Ear Hear*. (1986) 7:20–6. doi: 10.1097/00003446-198602000-00005

24. Blair J, Peterson M, Viehweg S. The effects of mild sensorineural hearing loss on academic performance of young school-age children. *Volta Rev*. (1985) 87:87–93.

25. Dancer J, Burl N, Waters S. Effects of unilateral hearing loss on teacher responses to the SIFTER. *Am Ann Deaf*. (1995) 140:291–4. doi: 10.1353/aad.2012.0592

26. Fischer C, Lieu J. Unilateral hearing loss is associated with a negative effect on language scores in adolescents. *Intl J Ped Otorhinolaryngol*. (2014) 78:1611–7. doi: 10.1016/j.ijporl.2014.07.005

27. Lewis D, Schmid K, O'Leary S, Heinrichs-Graham E, Spalding J, High R. Effects of noise on speech recognition and listening effort in children with normal-hearing and children with mild bilateral or unilateral hearing loss. *J Speech Lang Hear Res*. (2016) 59(5):1218–32. doi: 10.1044/2016_JSLHR-H-15-0207

28. Lieu J, Tye-Murray N, Karxon R, Piccirillo J. Unilateral hearing loss is associated with worse speech-language scores in children. *Pediatrics*. (2010) 125:2009–448. doi: 10.1542/peds.2009-2448

29. Oyler R, Oyler A, Matkin N. Unilateral hearing loss: demographics and educational impact. *Lang Speech Hear Serv Schools*. (1988) 19:201–10. doi: 10.1044/0161-1461.1902.201

30. Ruscetta MN, Arjmand EM, Pratt SR. Speech recognition abilities in noise for children with severe-to-profound unilateral hearing impairment. *International J Ped Otorhinolaryngol*. (2005) 69:771–9. doi: 10.1016/j.ijporl.2005.01.010

31. Wang J, Quach J, Sung V, Carew P, Edwards B, Grobler A, et al. Academic, behavioural, and quality of life outcomes of slight to mild hearing loss in late childhood: a population-based study. *Arch Dis Child*. (2019) 104(11):1056–63. doi: 10.1136/archdischild-2019-316917

32. Stecker GC, Gallun G. Binaural hearing, sound localization, and spatial hearing. In Tremblay K & Burkhardt R, editors. *Translational perspectives in auditory neuroscience: normal aspects of hearing*. San Diego: Plural Publishing, Inc. (2012) p. 83–433.

33. Griffin AM, Poissant SF, Freyman RL. Auditory comprehension in school-aged children with normal hearing and with unilateral hearing loss. *Lang Speech Hear Schools*. (2020) 51:29–41. doi: 10.1044/2019_LSHSS-OCHL-19-0020

34. Klatte M, Lachmann T, Meis M. Effects of noise and reverberation on speech perception and listening comprehension of children and adults in a classroom-like setting. *Noise Health*. (2010) 12:270–82. doi: 10.4103/1463-1741.70506

35. McFadden B, Pittman A. Effect of minimal hearing loss on children's ability to multitask in quiet and in noise. *Lang Speech Hear Serv Schools*. (2008) 39:342–51. doi: 10.1044/0161-1461(2008/032)

36. Picou E, Davis H, Lewis D, Tharpe AM. Contralateral routing of signals systems can improve speech recognition and comprehension in dynamic classrooms. *J Speech Lang Hear Res*. (2020) 63(7):2468–82. doi: 10.1044/2020_JSLHR-19-00411

37. Valente DL, Plevinsky H, Franco J, Heinrichs E, Lewis D. Experimental investigation of the effects of the acoustical conditions in a simulated classroom on speech recognition and learning in children. *J Acoust Soc Am*. (2012) 131 (1):232–46. doi: 10.1121/1.3662059

38. Best V, Ozmeral EJ, Shinn-Cunningham BG. Visually-guided attention enhances target identification in a complex auditory scene. *J Assoc Res Otolaryngol*. (2007) 8:294–304. doi: 10.1007/s10162-007-0073-z

39. Best V, Marrone N, Mason C, Kidd G Jr, Shinn-Cunningham B. Effects of sensorineural hearing loss on visually guided attention in a multitalker environment. *J Assoc Res Otolaryngol*. (2009) 10:142–8. doi: 10.1007/s10162-008-0146-7

40. Salanger M, Lewis D, Vallier T, McDermott T, Dergan A. Applying virtual reality to audiovisual speech perception tasks in children. *Amer J Audiol*. (2020) 29 (2):244–58. doi: 10.1044/2020_AJA-19-00004

41. Braasch J, Peters N, Valente DL. A loudspeaker-based project technique for spatical music applications using virtual microphone control. *Comput Music J*. (2008) 32(3):55–71. doi: 10.1162/comj.2008.32.3.55

42. McCreery RW, Spratford M, Kirby B, Brennan M. Individual differences in language and working memory affect children's speech recognition in noise. *Int J Audiol*. (2017) 56(5):306–15. doi: 10.1080/14992027.2016.1266703

43. Alloway TP. *Automated working memory assessment*. London, England: Pearson (2007).

44. Dunn LM, Dunn DM. *Peabody picture vocabulary test (4th ed.)*. Circle Pines, MN: American Guidance Service (2007).

45. Ferris FL, Kassoff A, Bresnick GH, Bailey I. New visual acuity charts for clinical research. *Amer J Ophthalmol*. (1982) 94(1):91–6. doi: 10.1016/0002-9394(82)90197-0

46. R Core Team R: *A language and environment for statistical computing*. R Foundation for Statistical Computing, Vienna, Austria. (2023) (Version 4.1.3)

47. Bates D, Maechler M, Bolker B, Walker S. *lme4: Linear mixed effects models using Eigen and S4*. (2022). R Package Version 1.1-33.

48. Kuznetsova A, Brockhoff PB, Christensen RHB, *lmerTest Package: Tests in Linear Mixed Effects v. 3.3* (2020). Available at: https://github.com/runehaubo/lmerTestR

49. Wickham H. *ggplot2l Elegant Graphics for Data Analysis*. New York: Springer-Verlag (2016) *(Version 3.3.5)*

50. Benjamini Y, Hochberg Y. Controlling the false discover rate: a practical and powerful approach to multiple testing. *J Royal Stat Soc: Series B (Methodological)*. (1995) 57(1):289–300. doi: 10.1111/j.2517-6161.1995.tb02031.x

51. Corbin NE, Yarnell Bonino A, Buss E, Leibold LJ. Development of open-set word recognition in children: speech-shaped noise and two-talker speech maskers. *Ear Hear*. (2016) 37:55–63. doi: 10.1097/AUD.0000000000000201

52. Elliott LL, Connors S, Kille E, Levin S, Ball K, Katz D. Children's understanding of monosyllabic nouns in quiet and in noise. *J Acoust Soc Am*. (1979) 66:12–21. doi: 10.1121/1.383065

53. Hieber F, Tillman T, W T. Room acoustics effects on monosyllabic word discrimination ability for normal and hearing impaired children. *J Speech Hear Res*. (1978) 21:440–58. doi: 10.1044/jshr.2103.440

54. Walker EA, Sapp C, Oleson JJ and McCreery RW. Longitudinal speech recognition in noise in children: effects of hearing status and vocabulary. *Front. Psychol*(2019) 10:1–12. doi: 10.3389/fpsyg.2019.02421

55. Fitzpatrick EM A-E, Whittingham J R, Fitzpatrick J. Characteristics of children with unilateral hearing loss. *Int J Audiol*. (2017) 56(11):819–28. doi: 10.1080/14992027.2017.1337938

56. Fitzpatrick EM, Durieux-Smith A, Whittingham J. Clinical practice for children with mild bilateral and unilateral hearing loss. *Ear Hear*. (2010) 31(3):392–400. doi: 10.1097/AUD.0b013e3181cdb2b9

57. Fitzpatrick EM, Nassrallah F, Gaboury I, Whittingham J, Vos B, Coyle D, et al. Trajectory of hearing loss in children with unilateral hearing loss. *Front Ped*. (2023) 11:1–13. doi: 10.3389/fped.2023.1149477

58. Lieu JEC, Karzon RK, Ead B, Tye-Murray N. Do audiologic characteristics predict outcomes in children with unilateral hearing loss? *Otol Neurotol*. (2013) 34 (9):1703–10. doi: 10.1097/MAO.0000000000000190

59. Briggs L, Davidson L, Lieu JEC. Outcomes of conventional amplification for pediatric unilateral hearing loss. *Ann Otol Rhinol Laryngol*. (2011) 120(7):448–54. doi: 10.1177/000348941112000705

60. Johnstone PM, Nabelek AK, Robertson VS. Sound localization acuity in children with unilateral hearing loss who wear a hearing aid in the impaired ear. *J Am Acad Audiol*. (2010) 21:522–34. doi: 10.3766/jaaa.21.8.4

61. Walker EA, Holte L, McCreery RW, Spratford M, Page T, Moeller MP. The influence of hearing aid use on outcomes of children with mild hearing loss. *J Speech Lang Hear Res* (2015) 58:1611-25. doi: 10.1044/2015_JSLHR-H-15-0043

62. American Academy of Audiology (2013). *American academy of audiology clinical practice guidelines: Pediatric amplification*. Clinical Practice Guidelines: Pediatric Amplification—The American Academy of Audiology (Accessed June 14, 2023).

63. Bagatto M, DesGeorges J, King A, Kitterick P, Laurnagaray D, Lewis D, et al. Consensus practice parameter: audiological assessment and management of unilateral hearing loss in children. *Int J Audiol*. (2019) 58(12):805–15. doi: 10.1080/14992027.2019.1654620

64. Walker EA, Spratford M, Ambrose SE, Holte L, Oleson J. Service delivery to children with mild hearing loss: current practice patterns and parent perceptions. *Am J Audiol*. (2017) 26(1):38–52. doi: 10.1044/2016_AJA-16-0063

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EDITED BY
Erin Plyler,
University of Tennessee Health Science Center (UTHSC), United States

REVIEWED BY
Ashley Flagge,
University of South Alabama, United States
Saravanan Elangovan,
University of Tennessee Health Science Center (UTHSC), United States

*CORRESPONDENCE
Linda Cupples
linda.cupples@mq.edu.au

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Speech, language, functional communication, psychosocial outcomes and QOL in school-age children with congenital unilateral hearing loss

Linda Cupples[1]*, Teresa Y. C. Ching[2,3,4] and Sanna Hou[5]

[1]Department of Linguistics, Centre for Language Sciences, Macquarie University, Sydney, NSW, Australia, [2]NextSense Institute, NextSense, Sydney, NSW, Australia, [3]Macquarie School of Education, Macquaarie University, Sydney, NSW, Australia, [4]School of Health and Rehabilitation Sciences, The University of Queensland, Brisbane, QLD, Australia, [5]National Acoustic Laboratories, Hearing Australia, Sydney, NSW, Australia

Introduction: Children with early-identified unilateral hearing loss (UHL) might be at risk for delays in early speech and language, functional communication, psychosocial skills, and quality of life (QOL). However, a paucity of relevant research prohibits strong conclusions. This study aimed to provide new evidence relevant to this issue.
Methods: Participants were 34 children, ages 9;0 to 12;7 (years;months), who were identified with UHL via newborn hearing screening. Nineteen children had been fitted with hearing devices, whereas 15 had not. Assessments included measures of speech perception and intelligibility; language and cognition; functional communication; psychosocial abilities; and QOL.
Results and discussion: As a group, the children scored significantly below the normative mean and more than one standard deviation below the typical range on speech perception in spatially separated noise, and significantly below the normative mean on written passage comprehension. Outcomes in other aspects appear typical. There was however considerable *within participant* variation in the children's degree of hearing loss over time, raising the possibility that this pattern of results might change as children get older. The current study also revealed that participants with higher levels of nonverbal ability demonstrated better general language skills and better ability to comprehend written passages. By contrast, neither perception of speech in collocated noise nor fitting with a hearing device accounted for unique variance in outcome measures. Future research should, however, evaluate the fitting of hearing devices using random assignment of participants to groups in order to avoid any confounding influence of degree of hearing loss or children's past/current level of progress.

KEYWORDS

unilateral hearing loss, congenital hearing loss, children, speech perception, language ability, school-age children

Introduction

There is general agreement in the literature that the introduction of Universal Newborn Hearing Screening (UNHS) has resulted in the identification of an increased number of children with unilateral hearing loss (UHL) at an earlier age (1–3). This increase has brought with it a strengthened research focus on the impact of early

identified UHL on children's language and other outcomes [e.g., (4)], and a related interest in evaluating the benefits of audiological rehabilitation with hearing aids (HAs) or cochlear implants (CIs) for this population [e.g., (5, 6)]. The aim of this research was to build on current literature; first, by examining a range of outcomes for a sample of 9-year-old children with congenital UHL; and second, by examining the association between children's outcomes and a set of predictor variables, including whether or not they had been fitted with hearing devices.

The outcome variables of direct interest in the current study were speech perception and production, language and cognition, functional auditory performance, psychosocial skills, and quality of life (QOL). Our particular focus was on children who presented with *congenital* unilateral hearing loss. A similar set of outcomes was the focus of a systematic review by Huttunen et al. (4), whose search of the literature up to February 2018 produced "no high-quality studies reporting on consequences of pre- or perilingual UHI [unilateral hearing impairment]" (p. 181). Consistent with this finding, Huttunen et al. stated that the literature they reviewed enabled them to draw "no definitive conclusions … on the impact of early-onset UHI on children's development" (p. 181). Nevertheless, individual research reports, especially those published since 2019, provide some support for the view that children with early-onset UHL achieve poorer outcomes than their age-matched peers with normal hearing (NH).

Fitzpatrick et al. (7) reported on 38 children with early-identified UHL. Thirty-five of the children presented with a congenital hearing loss, and no child was diagnosed with a severe developmental delay. The children's speech production, language, and functional auditory performance were assessed at 48 months of age, on average. When compared to a control group of age-matched children with NH, the children with UHL performed similarly on tests of receptive vocabulary and speech production, but significantly more poorly on assessments of receptive and expressive language and functional auditory performance.

Other researchers have also reported evidence of a selective impact of UHL on children's language and functional auditory outcomes. For example, Nasrallah et al. (8) reported that a group of children, ages 5–9 years, with UHL or mild bilateral HL, achieved outcomes within the average range of test normative means for receptive vocabulary, language, and speech production, but below expectations for functional auditory performance. Moreover, this pattern was true for both children with UHL and children with bilateral HL, whose scores did not differ significantly from one another. Griffin et al. (9) reported that a sample of 25 unaided children with UHL (15 congenital, ages 7;0 to 12;0 years;months) performed more poorly than a group of 14 NH children on an auditory story comprehension task when presented under challenging (noisy) conditions but not in quiet. Canête et al. (10) compared outcomes for a group of 12 participants, ages 7–16 years, with UHL due to congenital aural atresia, with results for 15 NH controls. Children with UHL generally performed more poorly on speech recognition in noise tasks, and especially for recognition of sentences.

Smit et al. (11) also reported on a participant sample with congenital conductive UHL due to aural atresia. Twenty-nine children and young adults, ages 6–21 years, took part in the research. Twelve of the 29 participants had an additional syndrome or medical condition, and 13 had used hearing amplification. All outcome measures were assessed using questionnaires. They included hearing QOL (in domains of spatial, speech, and quality of hearing), general QOL, language, and social-emotional-behavioural domains. The results show that study participants achieved lower scores in hearing QOL than children without hearing loss reported in the literature, and there was no effect of amplification. On the other hand, measures of general quality of life, language, and social-emotional-behavioural domains all fell within the normal range. Smit et al. (11) concluded that their study results provide evidence for a normal pattern of development in children and young adults who have conductive UHL due to aural atresia, while acknowledging that the "limited size and selection of the study population" might have contributed to their failure to detect real group differences (p. 6).

Irrespective of Smit et al.'s (11) concerns regarding possible methodological weaknesses in their study, the findings receive some support from related research. Nasrallah et al. (12) reported that a group of children, ages 5–9 years, with UHL or mild bilateral HL, achieved outcomes within the average range of test normative means for social and behavioural skills, as rated by parents and teachers. Moreover, this pattern of results was true for both children with UHL and children with bilateral HL, whose scores did not differ significantly from one another. On the other hand, findings reported by Griffin et al. (13) confirmed a significant difference in hearing-related QOL between children with UHL and those with NH.

In sum, recent studies examining the impact of UHL on children's development provide evidence of poorer outcomes relative to children with NH in functional auditory performance [e.g., (7, 8)], hearing-related quality of life [e.g., (11, 13)], and speech perception [e.g., (10, 13)]. On the other hand, non-significant differences have been observed in general QOL and psychosocial skills [e.g., (11, 12)], receptive vocabulary [e.g., (7, 8)], and speech production [e.g., (7, 8)]. With respect to language outcomes, results are inconsistent: Fitzpatrick et al. (7) found that language outcomes were worse for children with UHL compared to children with NH, whereas Nasrallah et al. (8) and Smit et al. (11) found evidence of outcomes within the typical range.

While these previously reported findings are suggestive, they do not enable strong conclusions to be drawn in regard to the impact of *congenital* UHL on children's outcomes, because most participant samples were diverse with respect to onset of hearing loss. Furthermore, children's cognitive development appears to have been overlooked in many recent published studies, despite evidence from a 2016 meta-analysis which showed that children with UHL scored significantly lower than expected on both full-scale IQ results and performance IQ (14). Hence, the first aim of the current research was to examine the impact of congenital UHL across a representative set of outcome variables including measures of speech perception and production, language and nonverbal cognition, functional auditory performance, psychosocial skills, and QOL.

The second aim of the current research was to examine the association between children's outcomes and a small set of concurrent predictor variables, which included fitting status (being fitted with a hearing device or not), nonverbal cognitive ability, and speech perception. Nonverbal ability and speech perception were included because of their demonstrated role in previous studies of speech and language outcomes achieved by children with congenital bilateral HL [e.g., (15–17)]. On the other hand, degree of hearing loss was not included as a concurrent predictor because it was not shown to play a consistent role in previous research involving children with UHL [e.g., (7, 9, 13)]. Failure to find a consistent association between degree of hearing loss and outcomes in this population might reflect, at least in part, changes in children's degree of hearing loss over time, as documented in several recent studies [e.g., (18–20)]. However, the current study was not designed to address this issue.

The current study

The current aims were addressed in a cross-sectional study of a group of school-aged children with congenital UHL. Three primary research questions were addressed.

1. Do children with congenital UHL exhibit speech and language deficits compared to norms at school age?
2. Do children with congenital UHL exhibit functional communication deficits compared to norms at school age?
3. Do children with congenital UHL exhibit deficits in psychosocial outcomes and QOL compared to norms at school age?

In accordance with findings reported in the literature, we predicted that children with congenital UHL would achieve poorer functional auditory outcomes than expected relative to norms, but similar psychosocial outcomes and QOL. Predictions regarding speech and language outcomes were less clear, with the possibility that different outcome measures might reveal different patterns of results; for example, children with UHL might achieve similar outcomes in speech production and receptive vocabulary but poorer outcomes on speech perception and other language measures.

Two additional questions were more exploratory.

4. What might account for variation in the outcomes achieved by children with UHL (e.g., nonverbal cognitive ability, speech perception, use or not of a hearing device)?
5. Why might children who are *not fitted* with a hearing device achieve better outcomes than fitted children?

Method

General procedure

The protocol for this study was approved by the Australian Hearing Human Research Ethics Committee. After enrolment in the study, parents completed a questionnaire to provide demographic information, including their own level of education and any additional disabilities affecting their children. Parents also completed questionnaires soliciting information on their children's use of language and hearing in real-world environments, behavior and emotions, and QOL. Child participants completed a battery of tasks comprising audiological assessments, which were performed at the children's local hearing centres, questionnaires regarding their use of language and hearing in real-world environments, and QOL. Research speech pathologists completed direct assessments of children's spoken and written language skills and nonverbal cognitive ability. They also rated the intelligibility of children's speech. These assessments were performed at either the children's homes or hearing centres. They were conducted between age 9;0 (9 years; 0 months) and 12;7.

The definition of UHL used in this study was based on the National Workshop on Mild Bilateral and Unilateral Hearing Loss (21). In particular, UHL was defined as the average pure tone air conduction threshold at 0.5, 1, 2 kHz of any level greater than or equal to 20 dB HL or pure tone air conduction thresholds greater than 25 dB HL at two or more frequencies above 2 kHz in the affected ear with an average pure tone air conduction threshold in the good ear less than or equal to 15 dB HL.

Participants

The current participant sample was drawn from a larger group of 153 children who were diagnosed with UHL at birth between 2002 and 2007 in New South Wales, Australia. The children were identified through Australia's nationwide newborn hearing screening program. Of the 153 children, 128 aged 9 years or older were invited to take part in the study after removing 6 children who lived remotely and a further 19 whose contact details were incomplete. Thirty-nine children and their families accepted the invitation to participate. After omitting children who subsequently withdrew from the study or did not have results available for an assessment of nonverbal cognitive ability, a final sample of 34 children remained (20F, 14M), 19 of whom were fitted with a hearing device and 15 of whom were not. Tables 1, 2 contain demographic and audiological characteristics of the final participant sample, including details of hearing devices. Just under half of the children were identified as having a disability in addition to their hearing loss (see Table 1). Disability types included: learning disability, cranio-facial

TABLE 1 Demographic characteristics of participants ($N = 34$).

Characteristics	Participants ($N = 34$)	
Gender		
Male	14	41.2%
Female	20	58.8%
Age at Diagnosis in months, mean (range)	2.3	(0.4, 9.7)
Age at Assessment in years;months, mean (range)	10;4	(9;0, 12;7)
Additional Disability (excl. ANSD)		
Yes	15	44.1%
No	15	44.1%
Not specified	4	11.8%

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TABLE 2 Participants' audiological characteristics ($N = 34$).

Audiological characteristics	Participants ($N = 34$)			
Hearing Loss affected ear				
Right	17		50.0%	
Left	17		50.0%	
Type of Hearing Loss	@ diagnosis			
SNHL	19	55.9		
Conductive	9	26.5		
Mixed	2	5.9		
ANSD	2	5.9		
Not specified	2	5.9		
Degree of Hearing Loss (affected ear)[a]	@ diagnosis		@ assessment	
Typical range	0	0	5	14.7
Mild	9	26.5	6	17.6
Moderate	13	38.2	4	11.8
Severe	8	23.5	9	26.5
Profound	3	8.8	10	29.4
Not specified	1	2.9	0	0.0
Device Configuration @ assessment[b]			@ assessment	
No device			15	44.1%
15×Unilateral fitting, 4×bilateral fitting			19	55.9%

[a]Degree of hearing loss based on a four-frequency average (0.5, 1, 2, and 4 kHz) of hearing thresholds, such that Mild <= 40 dB HL, moderate = 41–60 dB HL, severe = 61–90 dB HL, profound >= 91 dB HL.
[b]Of 4 bilateral devices: 2 bilateral HAs + remote microphones (FMs), 1 bilateral CIs + remote microphones, 1 bilateral HAs; Of 15 unilateral devices: 5 remote microphones only, 8 unilateral HAs + remote microphones, 2 unilateral HAs only.

abnormality, developmental delay, Golden Har syndrome, Autism Spectrum Disorder, Attention Deficit Hyperactivity Disorder, and vision problems.

All children were diagnosed with hearing loss via UNHS in their first year of life [*Mean* = 2.3 months; *standard deviation (SD)* = 2.2 months]. Following diagnosis of hearing loss, all children were referred to Hearing Australia (the national government-funded hearing service provider) for audiological management, which includes ongoing hearing assessments, hearing device fitting and verification using real-ear measures according to national pediatric protocols (22).

For purposes of the current study, degree of hearing loss is expressed as mild (averaged hearing loss <=40 dB HL), moderate (41–60 dB HL), severe (61–90 dB HL) or profound (>=91 dB HL) based on a four-frequency average (0.5, 1, 2 and 4 kHz) of hearing thresholds. At diagnosis, the majority of children (61.8%) had a sensorineural or mixed hearing loss, and most (88.2%) had hearing losses in the mild to severe range. Three children (8.8%) had a profound loss at diagnosis (see Table 2). When assessments were conducted, however, the number of children with hearing losses in the profound range had increased to 10 (29.4%), and the number with mild to severe losses had dropped to 19 (55.9%). The remaining 5 children had hearing within the typical range at assessment (see Table 2). Consistent with these findings, degree of hearing loss changed from diagnosis to assessment for most individual children ($n = 26$), but most (21) of these changes involved adjacent categories (e.g., from mild to moderate or from moderate to severe). Fourteen children had a higher degree of loss at assessment than diagnosis, whereas 12 children had a higher degree of loss at diagnosis.

Assessments

Audiology

Standard pure tone audiometry and tympanometry were conducted only if a child's current records were not within six months of assessment.

Speech and language

Speech perception

Speech perception was assessed using nonsense syllables (Vowel-Consonant-Vowel or VCV syllables) and sentences [Beautifully Efficient Speech Test (BEST), (23)]. Nonsense syllables were presented in collocated noise (VCV-N) at a signal-to-noise ratio (SNR) of 5 dB from a loudspeaker positioned at 0° azimuth at a distance of 0.75 metre, and performance was measured as percent correct. Sentences were presented in collocated noise at 0° azimuth (BEST-S0N0) or speech from the front at 0° and uncorrelated noise from +90° and −90° azimuth from both sides (BEST-S0N90). Performance was measured as speech reception thresholds, which were expressed in decibels (dB) SNR. Normative means and SDs for the BEST were taken from Ching et al. (24). There were no normative data available for the VCV.

Speech intelligibility

The Speech Intelligibility Rating scale [SIR, (25, 26)] was used to rate how easy or difficult it was to understand the children's speech. Ratings were assigned by parents and research speech pathologists (referred to as "other") using a 6-point scale, from 1 (always understand the child with little or no effort) to 6 (almost never understand the child's speech). Normative means and SDs were obtained from a related study in our laboratory.

Language

The following language assessments were administered to participants by research speech pathologists.

The Peabody Picture Vocabulary Test 4th Edition [PPVT-4; (27)] is a standardized test of receptive vocabulary, using a four-alternative forced-choice, picture-pointing format in administration. It gives an overall standard score for receptive vocabulary (*Mean* = 100, *SD* = 15).

The Clinical Evaluation of Language Fundamentals—4th Edition [CELF-4; (28)] is a standardized test of spoken English (*Mean* = 100; *SD* = 15). The test includes verbal tasks which enable children to demonstrate understanding of and ability to produce English language structures. In this study an overall core language score was computed along with three subtest scores—receptive language, expressive language, and language memory.

The Woodcock Johnson III® Diagnostic Reading Battery [WJ III® DRB; (29)] comprises a set of individually administered tests, three of which were used here. Letter-word identification and word attack assessed children's ability to read aloud single words and non-words respectively; and passage comprehension assessed children's understanding of words, phrases, and/or short passages using word-picture matching and cloze procedures. The test gives an individual standard score for each

test (*Mean* = 100, *SD* = 15), and a separate "Basic Reading" score, which combines results for letter-word identification and word attack.

Cognitive ability

Nonverbal cognitive ability was assessed using the Wechsler Nonverbal Scale of Ability [WNV; (30)], which was designed specifically for linguistically diverse populations, including people with hearing loss. This test provides a nonverbal IQ score (*Mean* = 100; *SD* = 15).

Functional auditory performance

The Parents' Evaluation of Aural/Oral Performance of Children [PEACH; (31)] and the Self-Evaluation of Listening Function [SELF; (32)] were used to measure children's functional auditory performance in real life. The PEACH was designed to assess children's listening and communicative behaviour in 10 real-world environments, based on observations by parents. The SELF was based on items in the PEACH with appropriate adaptations, and relied on subjective reports from children. Each item is rated on a five-point scale: never (0%), seldom (1%–25%), sometimes (26%–50%), often (51%–75%), and always (>75% of the time) by the respondent. Each assessment gives an overall score and two subscale scores, quiet and noise. Normal values for both tests were taken from a related study in our laboratory.

Psychosocial skills: behavior and emotions

The Strengths and Difficulties Questionnaire [SDQ; (33)] was used to assess children's behaviour and emotional difficulties. Parents completed the questionnaire, which comprises 25 items, making up five subscales: conduct problems, emotional symptoms, hyperactivity, peer relationships, and pro-social behavior. Each subscale consists of five items. The first four subscale scores (excluding prosocial behavior) were summed to make a "total difficulties score". Australian normative data by age group (7–10 years) and gender (34) were used to calculate *z*-scores. All "difficulties" scores were reversed so that higher *z*-scores reflect less problems.

Quality of life (QoL)

The Pediatric Quality of Life Inventory version 4.0 Generic Core Scales (PedsQL) were used to measure children's health-related quality of life. The inventory was completed by children (PedsQL-C) and their parents (PedsQL-P). It comprises 23 items from four domains: physical functioning, emotional functioning, social functioning, and school functioning. A psychosocial health summary score was calculated as the mean score over the items answered across the emotional, social and school functioning scales. Each item is rated on a 5-point Likert scale, from 0 (never a problem) to 4 (almost always a problem). Items were reversed-scored and rescaled to a 0–100 scale, where higher scores indicate better QoL. For scale and total scores, the mean was computed as the sum across all items divided by the number of items answered. *Z*-scores were computed using normative means and SDs from Varni et al. (35).

Data analysis

Data analysis was conducted in three stages. Stage 1 addressed the question of whether the current sample of children with congenital UHL achieved outcomes that differed from those achieved by a normative sample of children the same age. This question was addressed, first, by noting whether the mean scores achieved by the current sample were within one SD of their respective normative means; and second, using a series of 39 single-sample *t*-tests to compare the current sample's mean scores to the relevant normative means using an adjusted α-level of.001 (.05 ÷ 39). Stage 2 addressed the question of whether children using a hearing device would achieve different outcomes than those who did not use a hearing device. This question was addressed using a series of 41 independent samples *t*-tests with an adjusted α-level of .001 (.05 ÷ 41). Stage 3 addressed the question of what additional variables might account for variation in the outcomes achieved by the current sample of children. Correlational and regression techniques were used to address this question, using an α-level of .001 for correlations and .005 (.05 ÷ 10) for regressions.

Results

The first three research questions asked whether school-aged children with congenital UHL would exhibit deficits compared to age-matched norms in speech and language, functional communication, psychosocial abilities, and QOL. To address these questions, mean scores were computed for all individual outcome measures across all participants. These mean scores and standard deviations are shown in Tables 3–7, along with normative values where available.

For the most part, these data support the view that children with UHL in the current study performed at a level similar to typically developing children of the same age. The mean scores achieved by the current sample were within one SD of their respective normative means for outcomes in language and nonverbal cognition, functional auditory performance, behavior and emotion, and QOL. On the other hand, children performed outside the typical range on speech perception in noise, when speech and noise were spatially separated. The mean SNR for the BEST S0N90 was −1.11 dB for the current group, above the expected range of −6.8 to −2.2 dB; and the observed spatial release from masking (SRM) was 0.26 dB for the current group, below the expected range of 1.1–4.5 dB (Table 3).

This pattern of results was confirmed for the most part using a series of single-sample *t*-tests to compare the current sample's mean scores with the corresponding normative means. Using a corrected α-level of.001 (.05 ÷ 39 individual comparisons), three differences reached statistical significance. They were the results for BEST S0N90 and BEST SRM, which confirmed our previous analysis; and the result for WDRB Passage Comprehension ($t[33] = 7.15$, $p < .001$) on which children with UHL underperformed relative to norms (see Table 4).

TABLE 3 Speech outcomes for the current sample: comparison with norms and fitting status.

Measure	Scale	Mean (SD)		Prob[a]	Mean (SD)		Prob[b]
		Norms	Current		Fitted	Not Fitted	
Speech perception							
VCV[c]	Quiet %	NA	96.1 (5.4)	NA	95.6 (5.9)	96.7 (5.1)	.588
	Noise %	NA	88.3 (9.9)	NA	85.9 (11.5)	91.4 (6.6)	.138
BEST[d,e]	S0N0 SNR	−1.7 (1.7)	−.85 (1.8)	.014	.02 (1.7)	−1.99 (1.1)	.001*
	S0N90 SNR	−4.5 (2.3)	**−1.11 (4.3)**	<.001*	.78 (3.7)	−3.58 (3.9)	.004
	SRM	2.8 (1.7)	**.26 (3.6)**	<.001*	−.76 (2.8)	1.59 (4.1)	.072
Speech production							
SIR[f,g]	Parent	1.1 (0.3)	1.3 (0.5)	.068	1.4 (0.5)	1.2 (0.4)	.199
	Other	1.2 (0.5)	1.5 (0.6)	.050	1.6 (0.7)	1.3 (0.6)	.233

NA, not available; VCV, nonsense syllables; BEST, beautifully efficient speech test; SIR, speech intelligibility rating scale. $\alpha = .001$. Bold font is used to indicate the current group's scores that fall outside the typical range.
[a]Probability is computed using a one-sample *t*-test comparing the current sample to norms.
[b]Probability is computed using an independent samples *t*-test to compare children from the current sample who differ in fitting status.
[c]$n = 29$.
[d]$n = 30$.
[e]BEST normative means and SDs from Ching et al. (24).
[f]$n = 26$.
[g]SIR norms come from a related study in our laboratory.
*$p \leq .001$.

TABLE 4 Language and cognitive outcomes for the current sample: comparison with norms and fitting status.

Measure	Scale	Mean (SD)		Prob[a]	Mean (SD)		Prob[b]
		Norms	Current		Fitted	Not Fitted	
Receptive vocabulary							
PPVT SS		100 (15)	99.3 (13.5)	.772	96.6 (13.7)	102.7 (12.9)	.196
Language							
CELF4 SS	Core	100 (15)	93.9 (16.4)	.038	90.2 (16.0)	98.6 (16.2)	.141
	Rec Lang	100 (15)	91.4 (16.2)	.004	86.8 (17.0)	97.3 (13.5)	.060
	Exp Lang	100 (15)	95.9 (15.8)	.136	92.7 (15.2)	99.8 (16.2)	.200
	Lang Mem	100 (15)	94.1 (16.5)	.044	90.5 (15.7)	98.5 (17.0)	.164
Reading							
WDRB SS	Word ID	100 (15)	100.2 (14.5)	.944	97.1 (13.0)	104.1 (15.7)	.168
	Word Att	100 (15)	101.7 (11.3)	.377	98.3 (9.2)	106.1 (12.4)	.042
	Pass Comp	100 (15)	90.1 (8.1)	<.001*	88.1 (7.9)	92.6 (7.8)	.108
	Basic Read[c]	100 (15)	101.1 (13.3)	.636	97.6 (11.2)	105.5 (14.8)	.088
Nonverbal cognitive ability							
WNV SS	Full Scale	100 (15)	99.0 (13.2)	.653	93.7 (12.2)	105.7 (11.5)	.007

N = 34. PPVT, Peabody Picture Vocabulary Test 4th edition; CELF, Clinical Evaluation of Language Fundamentals 4th edition; WDRB, Woodcock-Johnson III Diagnostic Reading Battery; WNV, Wechsler Nonverbal Scale of ability. $\alpha = .001$.
[a]Probability is computed using a one-sample *t*-test comparing the current sample to norms.
[b]Probability is computed using an independent samples *t*-test to compare children from the current sample who differ in fitting status.
[c]WDRB Basic Reading scale combines Word ID and Word Attack.
*$p \leq .001$.

Research question 4 addressed what might account for variation in the outcomes achieved by children with UHL, in particular, aspects such as cognitive ability, speech perception, and whether or not a hearing device was fitted. In a related vein, question 5 addressed why children who were *not fitted* with a hearing device might achieve better outcomes than fitted children. To shed light on these issues, children were first divided into groups according to whether they were fitted with a hearing device or not. As might be expected, these groups differed in degree of hearing loss. To illustrate, Figure 1 shows the percentage of fitted vs. non-fitted participants with different degrees of hearing loss at the time of their assessment. On the SELF questionnaire, of the 19 children with hearing devices, 2 (10.5%) had missing data, 2 (10.5%) reported using their devices 50% of the time, and 15 (78.9%) reported using their devices ≥75% of the time.

The next step was to compare the assessment outcomes achieved by fitted and non-fitted children. Mean scores were computed for all individual outcome measures for fitted vs. non-fitted participants separately. These mean scores and standard deviations are shown in Tables 3–7. Using a corrected α-level of.001 (.05 ÷ 41 individual comparisons), outcomes for fitted vs. non-fitted participants differ

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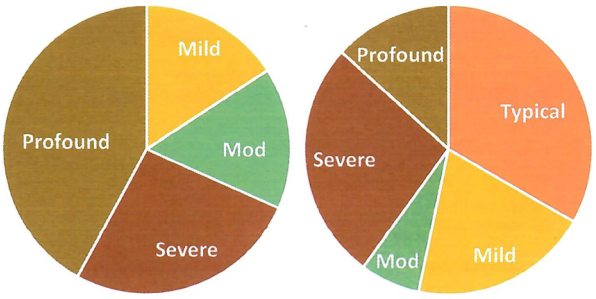

FIGURE 1
Percentage of fitted vs. non-fitted participants with different degrees of hearing loss at assessment.

significantly on only one measure, the BEST S0N0 ($t[28] = 3.66$, $p = .001$) assessment of speech perception in collocated noise (see Table 3). This result reflected better performance (lower speech reception thresholds) in participants who had *not* been fitted with a hearing device. Although no other individual comparisons were significant, children who had *not* been fitted with a hearing device generally performed better across the range of measures than children who had been fitted.

Correlation and regression techniques provide another approach to investigate within-group variability in outcomes. For these analyses, a limited set of 10 outcome measures was used. These measures were chosen because they provided an overall reflection of performance on the various assessments. They were: SIR (other) to measure speech intelligibility; PPVT-4, CELF-4 core language, WDRB basic reading, and WDRB passage comprehension to measure language skills; PEACH total, and SELF total to measure functional auditory performance; SDQP Total to measure psychosocial skills; and PedsQL-C Total and PedsQL-P Total to measure QoL. Table 8 shows the Pearson product-moment correlations between these variables and three potential predictors: device-fitting status, WNV, and speech perception in collocated noise (BEST S0N0). Tables 9, 10 show the summary results from 10 regression analyses using these three potential predictors, one for each outcome measure.

To summarise the correlation results: There were significant positive correlations ranging from.58 to.78 between the four language measures, indicating that children who performed well on one measure tended to score well on the other measures, as would be expected. In addition, WNV scores were positively associated with three of the four language measures (excluding PPVT), indicating that children with higher levels of nonverbal cognitive ability achieved better language outcomes. With regard to associations involving other variables, significant correlations

TABLE 5 Functional auditory performance for the current sample: comparison to norms and fitting status.

Measure	Scale	Mean (SD)		Prob[a]	Mean (SD)		Prob[b]
		Norms	Current		Fitted	Not Fitted	
PEACH[c]	Quiet %	87.7 (12.8)	84.6 (14.6)	.292	82.7 (18.1)	86.5 (10.5)	.514
	Noise %	82.8 (15.1)	71.5 (20.0)	.008	68.2 (20.8)	74.9 (19.4)	.402
	Total %	85.5 (13.2)	78.1 (16.0)	.027	75.4 (17.8)	80.8 (14.3)	.404
SELF[d]	Quiet %	87.4 (10.4)	85.0 (15.2)	.385	83.1 (18.8)	87.7 (7.8)	.410
	Noise %	83.9 (13.2)	84.7 (17.5)	.806	81.7 (21.6)	88.9 (8.5)	.267
	Total %	85.7 (10.8)	84.8 (15.9)	.765	82.4 (20.0)	88.3 (6.5)	.315

PEACH, Parents' Evaluation of Children's Aural/Oral Performance; SELF, Self Evaluation of Listening Function. Norms for PEACH and SELF are from a related study in our laboratory. $\alpha = .001$.
[a]Probability is computed using a one-sample *t*-test comparing the current sample to norms.
[b]Probability is computed using an independent samples *t*-test to compare children from the current sample who differ in fitting status.
[c]$n = 26$.
[d]$n = 31$.
*$p \leq .001$.

TABLE 6 Psychosocial outcomes—behavior and emotion—for the current sample: comparison to norms and fitting status.

Measure	Scale	Mean (SD)		Prob[a]	Mean (SD)		Prob[b]
		Norms	Current		Fitted	Not Fitted	
SDQP—*Z*	Emotional	0 (1.0)	−.12 (1.2)	.624	−.27 (1.38)	.02 (1.04)	.558
	Conduct	0 (1.0)	−.28 (1.1)	.215	−.20 (1.13)	−.35 (1.07)	.740
	Hyperactivity	0 (1.0)	−.15 (.88)	.401	−.12 (1.02)	−.18 (0.78)	.873
	Peer relations	0 (1.0)	−.01 (1.0)	.945	−.04 (1.21)	.01 (0.86)	.922
	Prosocial	0 (1.0)	.05 (.98)	.802	.19 (0.92)	−.08 (1.06)	.493
	Total[c]	0 (1.0)	−.18 (.95)	.360	−.20 (1.19)	−.16 (0.72)	.920

$N = 25$. SDQP—*Z*, Strengths and Difficulties Questionnaire *Z*-scores. $\alpha = .001$.
[a]Probability is computed using a one-sample *t*-test comparing the current sample to norms.
[b]Probability is computed using an independent samples *t*-test to compare children from the current sample who differ in fitting status.
[c]The first four subscales (emotional, conduct, hyperactivity, and peer relationships) were summed to make the Total Difficulties score.
*$p \leq .001$.

TABLE 7 Quality of life (QOL) outcomes for the current sample: comparison to norms and fitting status.

Measure	Scale	Mean (SD)		Prob[a]	Mean (SD)		Prob[b]
		Norms	Current		Fitted	Not Fitted	
PEDSQLC—Z[c]	Physical	0 (1.0)	−.16 (0.93)	.328	−.38 (1.08)	.11 (0.62)	.141
	Emotional	0 (1.0)	−.51 (0.91)	.004	−.56 (1.08)	−.44 (0.68)	.720
	Social	0 (1.0)	−.11 (1.10)	.561	−.20 (1.10)	−.01 (1.12)	.628
	School	0 (1.0)	−.32 (0.88)	.049	−.47 (0.90)	−.12 (0.83)	.268
	Psychosoc	0 (1.0)	−.38 (1.01)	.039	−.50 (1.08)	−.24 (0.93)	.480
	Total	0 (1.0)	−.34 (1.03)	.071	−.50 (1.15)	−.13 (0.86)	.325
PEDSQLP—Z[d]	Physical	0 (1.0)	.18 (0.95)	.335	.33 (0.52)	.03 (1.25)	.434
	Emotional	0 (1.0)	−.57 (1.31)	.035	−.61 (1.58)	−.54 (1.02)	.897
	Social	0 (1.0)	−.15 (1.01)	.472	−.20 (1.15)	−.09 (0.91)	.778
	School	0 (1.0)	−.22 (0.90)	.231	−.28 (1.00)	−.15 (0.83)	.715
	Psychosoc	0 (1.0)	−.39 (1.11)	.087	−.46 (1.36)	−.32 (0.84)	.766
	Total	0 (1.0)	−.17 (0.93)	.349	−.15 (0.95)	−.20 (0.96)	.903

PEDSQLC—Z, Pediatric Quality of Life Inventory Child Self Report Z-scores; PEDSQLP—Z, Pediatric Quality of Life Inventory Parent Proxy Report Z-scores. Normative means and SDs from Varni et al. (35). $\alpha = .001$.

[a]Probability is computed using a one-sample *t*-test comparing the current sample to norms.

[b]Probability is computed using an independent samples *t*-test to compare children from the current sample who differ in fitting status.

[c]$n = 32$.

[d]$n = 26$.

*$p \leq .001$.

TABLE 8 Correlations between outcome measures for the current sample.

	BEST SONO	WNV	SIR	Language				Functional auditory performance		SDQP	Quality of life	
	BEST S0N0	WNV	SIR	PPVT	CELF	WDRB Basic	WDRB Comp	PEACH	SELF	SDQP	Peds QL-C	Peds QL-P
Fitting	−.57* (30)	.46 (34)	−.24 (26)	.23 (34)	.26 (34)	.30 (34)	.28 (34)	.17 (26)	.19 (31)	.02 (25)	.18 (32)	−.03 (26)
BEST	1.00	−.24 (30)	.38 (22)	−.39 (30)	−.38 (30)	−.31 (30)	−.28 (30)	−.45 (22)	−.56 (27)	−.17 (21)	−.26 (28)	−.10 (22)
WNV		1.00	−.25 (26)	.46 (34)	.60* (34)	.54* (34)	.58* (34)	−.04 (26)	.12 (31)	.10 (25)	.51 (32)	.04 (26)
SIR			1.00	−.46 (26)	−.44 (26)	−.19 (26)	−.28 (26)	−.50 (26)	.26 (23)	−.57 (25)	−.46 (24)	−.42 (26)
PPVT				1.00	.71* (34)	.58* (34)	.78* (34)	.20 (26)	.24 (31)	.06 (25)	.31 (32)	−.05 (26)
CELF					1.00	.67* (34)	.69* (34)	.16 (26)	.41 (31)	.04 (25)	.55* (32)	.01 (26)
WDRB B						1.00	.67* (34)	.13 (26)	.37 (31)	−.02 (25)	.51 (32)	−.02 (26)
WDRB C							1.00	−.13 (26)	.23 (31)	.04 (25)	.47 (32)	−.13 (26)
PEACH								1.00	.37 (23)	.44 (25)	.17 (24)	.44 (26)
SELF									1.00	−.17 (22)	.42 (31)	−.03 (23)
SDQP										1.00	.40 (23)	.62* (25)
PedsQL-C											1.00	.44 (24)
PedsQL-P												1.00

SIR, Speech Intelligibility Ratings by research speech pathologists; CELF, CELF Core Language Score; WDRB B, WDRB Basic Reading score; WDRB C, WDRB Passage Comprehension score; PEACH, PEACH Total; SELF, SELF total; SDQP, SDQP total Z-score; PedsQL-C, PedsQL child report z-score; PedsQL-P, PedsQL parent proxy report z-score. $\alpha = .001$.

*$p <= .001$.

revealed that: device fitting was associated with higher speech reception thresholds indicating poorer performance ($r = -.57$, $p < .001$); children who achieved better outcomes in CELF-4 core language also scored higher on the PedsQL-C ($r = .55$, $p < .001$); and parents' ratings of their children on the SDQP were positively associated with their ratings on the PedsQL-P ($r = .62$, $p < .001$).

Consistent with the results for the correlational analyses, multiple regressions revealed that only nonverbal cognitive ability as reflected in WNV scores accounted for significant unique variance in outcomes; in particular for CELF-4 core language and WDRB passage comprehension (see Tables 9, 10). In accordance with these results, the total variance explained in the regression analyses was generally small and non-significant. Only one regression analysis accounted for significant total variance, that of CELF-4 core language scores, with 52.2% of variance explained. The other nine analyses accounted for nonsignificant variance ranging from 2.3% to 37.1%.

Discussion

This study was designed to investigate the outcomes achieved by a group of school-aged children with congenital UHL. Children's

TABLE 9 Regression summary table for speech and language measures (speech intelligibility, PPVT receptive vocabulary, CELF4 core language score, WDRB basic Reading, WDRB passage comprehension).

Independent variable	Outcome measure									
	SIR		Language							
	SIR		PPVT-4		CELF-4 core		WDRB basic reading		WDRB passage comprehension	
	R^2									
Fit status Y/N	.019		.057		.057		.132		.113	
BEST S0N0	.137		.097		.085		.016		.012	
WNV	.199		.180		.379 ($p < .001$)		.162		.246 ($p = .004$)	
Total R^2	.355		.334		.522 ($p < .001$)		.310		.371	
n	22		30		30		30		30	
	Regression coefficients									
	Beta	Sig	Beta	Sig	Beta	Sig	Beta	Sig	Beta	Sig
Fit status Y/N	.526	.085	−.305	.197	−.441	.033	−.035	.882	−.123	.587
BEST S0N0	.641	.020	−.443	.033	−.449	.012	−.213	.295	−.208	.285
WNV	−.540	.030	.519	.013	.752	<.001	.492	.020	.605	.004

SIR, Speech Intelligibility ratings by research speech pathologists. Standard scores used for PPVT4, CELF-4 core language score, WDRB Basic reading, and WDRB Passage Comprehension. $\alpha = .005$ (.05 ÷ 10).

TABLE 10 Regression summary table for functional auditory performance (PEACH, SELF), behaviour and emotion (SDQP), and quality of life (pedsQL-C, pedsQL-P).

Independent variable	Outcome measure									
	Functional auditory performance				SDQP		Quality of life			
	PEACH		SELF		SDQP		PedsQL-C		PedsQL-P	
	R^2									
Fit status Y/N	.011		.067		.002		.045		.000	
BEST S0N0	.239		.245		.031		.029		.013	
WNV	.001		.002		.010		.201		.010	
Total R^2	.251		.314		.043		.275		.023	
n	22		27		21		28		22	
	Regression coefficients									
	Beta	Sig	Beta	Sig	Beta	Sig	Beta	Sig	Beta	Sig
Fit status Y/N	−.305	.340	−.014	.957	−.164	.649	−.229	.355	−.162	.653
BEST S0N0	−.632	.031	−.580	.010	−.255	.418	−.246	.244	−.185	.556
WNV	.043	.864	−.062	.791	.119	.684	.546	.016	.121	.673

PEACH, PEACH total; SELF, SELF total; SDQP, SDQP total z-score; PedsQL-C, PedsQL child report z-score; PedsQL-P, PedsQL parent proxy report z-score. $\alpha = .005$ (.05 ÷ 10).

performance was evaluated on a comprehensive set of assessments targeting speech perception and production, language and cognition, functional auditory performance, behaviour and emotions, and QOL. The current participants achieved similar outcomes to the normative groups on all but three of the outcome measures: they required higher SNRs for speech perception in noise under conditions when speech and noise were spatially separated; they showed less spatial release from masking; and they underperformed on a test of written passage comprehension.

This pattern of results is similar in some respects to findings reported in the literature. Participants achieved typical outcomes in general QOL and psychosocial skills (behavior and emotions), consistent with previous reports by Smit et al. (11) for children and young adults ages 6–21 years, and Nasrallah et al. (12) for children ages 5–9 years. Participants also showed no marked weakness in receptive vocabulary, in accord with Fitzpatrick et al. (7) and Nasrallah et al. (8). There was limited evidence that children might exhibit a weakness in some aspects of language but not others, which might help to explain inconsistencies in findings between studies [e.g., (7, 8, 11)]. Finally, our finding that children performed below the typical range on a task assessing perception of sentences presented in noise is consistent with results described by Canête et al. (10), whose participant sample was similar to the current group in age (at 7–16 years of age) and congenital onset.

Setting aside these similarities, the results stand in contrast with reports in the literature that children with UHL achieve outcomes below expectations for functional auditory performance at 48 months of age (7) and 5–9 years of age (8). A possible explanation for this inconsistency across studies lies in the current study's focus on older children, of 9–12 years of age. As

the data in Table 2 show, for the participants in this study, there were marked differences in degree of hearing loss across the period from diagnosis to assessment, and these changes raise the possibility that assessment results might be influenced considerably by the timepoint at which they are administered.

Another point of investigation in the current study was the identification of factors that might underlie variability in the outcomes achieved by children with UHL. As a first step, participants were allocated to groups according to whether they were fitted with a hearing device or not. Comparison of the groups' performance across the full range of outcomes revealed one significant difference: Device fitting was associated with higher speech reception thresholds indicating poorer performance. No other individual comparisons were significant, although there was a general trend in the data for fitted children to achieve worse outcomes than children who were not fitted. While this overall pattern might seem counterintuitive, it presumably reflects the fact that the decision to fit or not was influenced by children's severity of hearing loss (with aids more likely for children with more severe losses), and by how well they were progressing, that is, the decision to fit was not independent of performance, but rather, prompted partly by poor progress. The only way to ensure that results are not confounded in this way is to *randomly* assign children to participant groups according to fitting status in future studies.

Correlations and regression analyses were used to provide further evidence regarding factors that might account for variability in children's outcomes. Two factors were targeted in addition to fitting status: They were nonverbal cognitive ability and speech perception in noise. Only cognitive ability accounted for significant variance in any outcome measure, and in particular for two language measures: CELF-4 core language and WDRB passage comprehension. This finding is consistent with our previous research examining concurrent predictors of language in 5-year-old children with congenital, bilateral hearing loss (17); and more importantly, it underscores the importance of including cognitive ability in future studies of outcomes in similar participant groups.

Strengths, limitations, and future directions

This report focuses on the outcomes achieved by a cohort of 34 children with UHL at 9 years of age. A major strength of the study lies in its inclusion of a group of children who were diagnosed with UHL at birth through Australia's universal newborn hearing screening program. By contrast, many previous studies of UHL have included more diverse groups of children with UHL and/or mild bilateral HL of varying onset. A second strength of the study lies in its use of data that were collected across a limited age range (from 9;0 to 12;7 years of age) using questionnaires and directly administered tests to assess a comprehensive set of representative outcome variables including speech production and perception, language and cognitive ability, functional auditory performance, psychosocial skills (behavior and emotions), and QOL.

Despite these strengths, the study is not without its limitations. With no longitudinal component, the results can provide only a snapshot in time with respect to children's outcomes. A longitudinal component could be particularly informative given the demonstrated variability in degree of hearing loss across time *within participants*, which is evident in the current sample and other recent investigations of children with UHL (18–20). A second limitation is the small sample size of 34, which restricted the number of independent variables that could be included in multiple regression analyses and therefore contributed to the small percentage of variance explained. Finally, as noted earlier, the effect of fitting status on outcomes was confounded in the current study because the decision to fit a hearing device was not independent of children's degree of hearing loss or their current progress.

Conclusion

The current study investigated outcomes in 9-year speech perception and production, language and cognition, functional auditory performance, psychosocial skills (behavior and emotions), and QOL in a cohort of 34 children who were identified with UHL through Australia's universal newborn hearing screening program. As a group, the children scored significantly below the normative mean and more than one SD below the typical range on a measure of speech perception in spatially separated noise. They also scored significantly below the normative mean on written passage comprehension. Outcomes in other aspects, such as spoken language ability, psychosocial skills, QOL, and nonverbal ability appear typical. It will be important, however, to discover whether this pattern of results changes as children get older, especially in light of the within participant variation evident in the current children's degree of hearing loss over time. On a practical level, these findings enhance our understanding of the difficulties experienced by children with congenital unilateral hearing loss at school age. In particular, observed difficulties with speech perception in noise are likely to have a negative impact on children's ability to learn effectively in classrooms, which are generally noisy places. The findings in this regard underscore the importance of reducing the impact of noise in classrooms and closely monitoring children's learning on a regular basis, especially for children with unilateral hearing loss.

The current study also revealed that participants with higher levels of nonverbal cognitive ability demonstrated better general language skills and better ability to comprehend written passages. On the other hand, neither perception of speech in collocated noise nor fitting with a hearing device accounted for unique variance in outcome measures. However, further research in this area is required before strong conclusions can be drawn. For example, the effect of fitting hearing devices should include random assignment of participants to groups according to whether they are fitted or not. If random allocation is not possible, there is a strong likelihood that the decision to fit will be influenced by confounding variables, such as degree of hearing loss (children with more severe losses are more likely to be fitted with a hearing device) and past/current progress (parents of a child who is experiencing difficulties may be more likely to try something that might help than parents of a child who is doing well).

Data availability statement

The datasets presented in this article are not readily available. Individual subjects will be referred to by a subject number, which is arbitrarily selected for each subject. All data will be stored according to the National acoustic Laboratories (NAL) protocol for confidential storage. All data collected as part of the research study must be kept for at least 7 years after the publication of the findings of the study, in accordance with the requirements of the Health Privacy Principles. Paper consent forms and assessment records will be put into secure document storage at the conclusion of the study. The document storage will be managed by NAL. A scanned copy of all forms will be saved by NAL, in accordance with the requirements on data storage at NAL. Requests to access the datasets should be directed to Viji Easwar, Viji.Easwar@nal.gov.au.

Ethics statement

The studies involving humans were approved by Hearing Australia Human Research Ethics Committee, which is registered with National Health and Medical Research Council. The studies were conducted in accordance with the local legislation and institutional requirements. Written informed consent for participation in this study was provided by the participants' legal guardians/next of kin.

Author contributions

LC: Conceptualization, Data curation, Formal Analysis, Funding acquisition, Investigation, Methodology, Writing – original draft, Writing – review & editing. TC: Conceptualization, Data curation, Formal Analysis, Funding acquisition, Investigation, Methodology, Project administration, Resources, Supervision, Writing – review & editing. SH: Data curation, Investigation, Methodology, Project administration, Writing – review & editing, Conceptualization.

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Conflict of interest

The authors declare that the research was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

Publisher's note

References

1. Anne S, Lieu JEC, Cohen MS. Speech and language consequences of unilateral hearing loss: a systematic review. *Otolaryngol Head Neck Surg.* (2017) 157(4):572–9. doi: 10.1177/0194599817726326

2. Fitzpatrick EM, Durieux-Smith A, Whittingham J. Clinical practice for children with mild bilateral and unilateral hearing loss. *Ear Hear.* (2010) 31(3):392–400. doi: 10.1097/AUD.0b013e3181cdb2b9

3. Hung Y-C, Chen P-H, Lin T-H, Lim TZ. Children with unilateral loss after newborn hearing screening in Taiwan. *Am J Audiol.* (2022) 31:646–55. doi: 10.1044/2022_AJA-22-00019

4. Huttunen K, Erixon E, Löfkvist U, Mäki-Torkko E. The impact of permanent early-onset unilateral hearing impairment in children—a systematic review. *Int J Pediatr Otorhinolaryngol.* (2019) 120:173–83. doi: 10.1016/j.ijporl.2019.02.029

5. della Volpe A, Ippolito V, Roccamatisi D, Garofalo S, De Lucia A, Gambacorta V, et al. Does unilateral hearing loss impair working memory? An Italian clinical study comparing patients with and without hearing aids. *Front Neurosci.* (2020) 14:905. doi: 10.3389/fnins.2020.00905

6. Nicolas S, Gallois Y, Calmels M-N, Deguine O, Fraysse B, Marx M. Quality of life of children treated for unilateral hearing loss: a systematic review and meta-analysis. *Arch Dis Child.* (2021) 106:1102–10. doi: 10.1136/archdischild-2020-320389

7. Fitzpatrick EM, Gaboury I, Durieux-Smith A, Coyle D, Whittingham J, Nassrallah F. Auditory and language outcomes in children with unilateral hearing loss. *Hear Res.* (2019) 372:42–51. doi: 10.1016/j.heares.2018.03.015

8. Nasrallah F, Fitzpatrick EM, Whittingham J, Sun H, Na E, Grandpierre V. A descriptive study of language and literacy skills of early school-aged children with

unilateral and mild to moderate bilateral hearing loss. *Deafness Educ Int*. (2018) 22 (2):74–92. doi: 10.1080/14643154.2018.1555119

9. Griffin AM, Poissant SF, Freyman RF. Auditory comprehernsion in school-aged children with normal hearing and with unilateral hearing loss. *Lang Speech Hear Serv Sch*. (2020) 51:29–41. doi: 10.1044/2019_LSHSS-OCHL-19-0020

10. Canête OM, Purdy SC, Brown CRS, Neeff M, Thorne PR. Behavioural performance and self-report measures in children with unilateral hearing loss due to congenital aural atresia. *Auris Nasus Larynx*. (2021) 48:65–74. doi: 10.1016/j.anl.2020.07.008

11. Smit AL, Burgers YRW, Swanenburg de Veye HFN, Stegeman I, Breugem CC. Hearing related quality of life, developmental outcomes and performance in children and young adults with unilateral conductive hearing loss due to aural atresia. *Int J Pediatr Otorhinolaryngol*. (2021) 142:110590. doi: 10.1016/j.ijporl.2020.110590

12. Nasrallah F, Tang K, Whittingham J, Sun H, Fitzpatrick EM. Auditory, social, and behavioral skills of children with unilateral/mild hearing loss. *J Deaf Stud Deaf Educ*. (2020) 25(2):167–77. doi: 10.1093/deafed/enz041

13. Griffin AM, Poissant SF, Freyman RF. Speech-in-noise and quality-of-life measures in school-aged children with normal hearing and with unilateral hearing loss. *Ear Hear*. (2018) 40(4):887–904. doi: 10.1097/AUD.0000000000000667

14. Purcell PL, Shinn JR, Davis GE, Sie KCY. Children with unilateral hearing loss may have lower intelligence quotient scores: a meta-analysis. *Laryngoscope*. (2016) 126:746–54. doi: 10.1002/lary.25524

15. Ching TYC, Cupples L, Zhang V. Predicting 9-year language ability from preschool speech recognition in noise in children using cochlear implants. *Trends Hear*. (2022) 26:1–13. doi: 10.1177/23312165221090395

16. Ching TYC, Dillon H, Leigh G, Cupples L. Learning from the longitudinal outcomes of children with hearing impairment (LOCHI) study: summary of 5-year findings and implications. *Int J Audiol*. (2018) 57(Suppl 2):S105–11. doi: 10.1080/14992027.2017.1385865

17. Cupples L, Ching TYC, Button L, Seeto M, Zhang V, Whitfield J, et al. Spoken language and everyday functioning in 5-year-old children using hearing aids or cochlear implants. *Int J Audiol*. (2018) 57(Suppl 2):S55–69. doi: 10.1080/14992027.2017.1370140

18. Fitzpatrick EM, Al-Essa RS, Whittingham J, Fitzpatrick J. Characteristics of children with unilateral hearing loss. *Int J Audiool*. (2017) 56:819–28. doi: 10.1080/14992027.2017.1337938

19. Fitzpatrick EM, Nasrallah F, Gaboury I, Whittingham J, Vos B, Coyle D, et al. Trajectory of hearing loss in children with unilateral hearing loss. *Front Pediatr*. (2023) 11:1149477. doi: 10.3389/fped.2023.1149477

20. Zhang VW, Hou S, Wong A, Flynn C, Oliver J, Weiss M, et al. Audiological characteristics of children with congenital unilateral hearing loss: insight into age of reliable behavioural audiogram acquisition and change of hearing loss. *Front Pediatr*. (2023) 11:1279673. doi: 10.3389/fped.2023.1279673

21. Centers for Disease Control and Prevention. *National workshop on mild and unilateral hearing loss. Workshop Proceedings*; Breckenridge, CO (2005).

22. King AM. The national protocol for paediatric amplification in Australia. *Int J Audiol*. (2010) 49(Suppl 1):S64–9. doi: 10.3109/14992020903329422

23. Best V, McLelland M, Dillon H. *The BEST (beautifully efficient speech test) for evaluating speech intelligibility in noise. Poster Presented at the World Congress of Audiology*; Brisbane, Australia (2014).

24. Ching TYC, van Wanrooy E, Dillon H, Carter L. Spatial release from masking in normal-hearing children and children who use hearing aids. *J Acoust Soc Am*. (2011) 129(1):368–75. doi: 10.1121/1.3523295

25. Nikolopoulos TP, Archbold SM, Gregory S. Young deaf children with hearing aids or cochlear implants: early assessment package for monitoring progress. *Int J Pediatr Otorhinolaryngol*. (2005) 69(2):175–86. doi: 10.1016/j.ijporl.2004.08.016

26. Allen C, Nikolopoulos T, Dyar D, O'Donoghue GM. Reliability of a rating scale for measuring speech intelligibility after pediatric cochlear implantation. *Otol Neurotol*. (2001) 22(5):631–3. doi: 10.1097/00129492-200109000-00012

27. Dunn LM, Dunn DM. *Peabody Picture Vocabulary Test*. 4th ed. Minneapolis, MN: Pearson Assessments (2007). doi: 10.1037/t15144-000

28. Semel E, Wiig EH, Secord WA. *Clinical Evaluation of Language Fundamentals, Fourth Edition (CELF-4)*. San Antonio, TX: The Psychological Corporation (2003).

29. Woodcock RW, Mather N, Schrank FA. *Woodcock Johnson III® Diagnostic Reading Battery (WJ III® DRB) 3rd ed. Rolling Meadows*. IL, USA: The Riverside Publishing Company (2004).

30. Wechsler D, Naglieri JA. *Wechsler Nonverbal Scale of Ability*. San Antonio, TX: PsychCorp (2006).

31. Ching TYC, Hill M. The parents' evaluation of aural/oral performance of children (PEACH) scale: normative data. *J Am Acad Audiol*. (2007) 18(3):220–35. doi: 10.3766/jaaa.18.3.4

32. Ching TYC, Hill M, Dillon H. Effect of variations in hearing-aid frequency response on real-life functional performance of children with severe or profound hearing loss. *Int J Audiol*. (2008) 47(8):461–75. doi: 10.1080/14992020802116128

33. Goodman R. The strengths and difficulties questionnaire: a research note. *J Child Psychol Psychiatry*. (1997) 38:581–6. doi: 10.1111/j.1469-7610.1997.tb01545.x

34. Mellor D. Normative data for the strengths and difficulties questionnaire in Australia. *Aust Psychol*. (2005) 40(3):215–22. doi: 10.1080/00050060500243475

35. Varni JW, Burwinkle TM, Seid M, Skarr D. The PedsQL 4.0 as a pediatric population health measure: feasibility, reliability and validity. *Ambul Pediatr*. (2003) 3:329–41. doi: 10.1367/1539-4409(2003)003<0329:TPAAPP>2.0.CO;2

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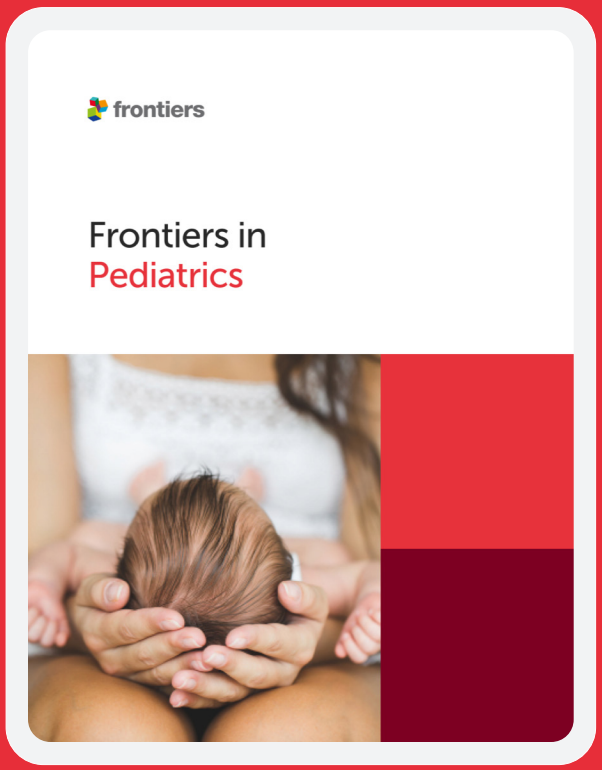

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