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PSYCHOSOCIAL SKILLS AND QUALITY OF LIFE IN DEAF AND HARD OF HEARING CHILDREN: LEARNING FROM THE FIRST TEN YEARS OF THE LOCHI STUDY

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This article discusses findings from the first 10 years of the Longitudinal Outcomes of Children with Hearing Impairment (LOCHI) Study, which includes more than 450 deaf and hard of hearing Australian children whose hearing losses were identified variously through newborn hearing screening or later paths to confirmation and intervention.

The Longitudinal Outcomes of Children with Hearing Impairment (LOCHI) Study is a population-based prospective study in Australia that, since 2005, has periodically assessed a wide range of outcomes for a cohort of children who are deaf or hard of hearing (DHH) to investigate the effectiveness of early identification and early intervention and the factors that account for variability in outcomes.

Prior to the LOCHI study, research into the effect of earlier identification comprised mainly either (a) retrospective comparisons of convenience samples of children that were often drawn from single early intervention, educational, or audiological services that tended to show positive effects of earlier identification (e.g., Calderon & Naidu, 1998; Moeller, 2000; Yoshinaga-Itano et al., 1998); or (b) population based studies that tended to show no, or limited, benefits of earlier identification of hearing loss (e.g., Kennedy et al., 2006; Korver et al., 2010).

Given the widespread acceptance of Universal Newborn Hearing Screening (UNHS) and the extent to which governments around the world made commitments to its implementation, it was an anomaly that the evidence for its efficacy at the initial stages of its development was

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not strong. Indeed, as late as 2008, a systematic review (Nelson et al., 2008) noted that there was only “moderate certainty that the net benefit of screening all newborn infants for hearing loss is moderate”, noting the need for evidence from prospective examination of whether earlier identification of hearing loss resulted in improved developmental and educational outcomes for children born with hearing loss. It was in that context that the LOCHI study was conceived.

LOCHI STUDY DESIGN

The need for a prospective study of the efficacy of earlier identification of hearing loss presented a significant design dilemma for researchers in contexts where newborn hearing screening was already operating at population level. In such jurisdictions, random assignment of children to be either screened or not screened would not be ethical and the idea of random assignment to treatment groups (early intervention versus later intervention) would be even more ethically egregious. To overcome the need for random assignment, the LOCHI study took advantage of a window of time in Australia between 2002 and 2007 when several Australian states were implementing UNHS programs at different rates. The three most populous states—New South Wales, Victoria, and Queensland—each commenced programs within a three-year period from 2002 to 2005. New South Wales implemented UNHS across all regions of the state simultaneously, whereas in Queensland and Victoria the roll-out of the program was staged across regions over 2 years and 6 years, respectively (Leigh, 2006).

As a prospective study, all children identified with hearing loss who were born in those three states across a 5-year period from 2002 to 2007 were invited to participate, resulting in a cohort of approximately 470 children that was naturally divided into those who were identified through UNHS and those who were identified later, allowing age of identification to be a primary predictor variable in a prospective research design (Ching et al., 2013). In addition, data on numerous other predictor variables were also collected, including (a) child-related variables (i.e., birth weight, gender, age of identification/diagnosis, degree of hearing loss, age at hearing aid fitting, age at cochlear implantation, non-verbal cognitive ability presence of additional disabilities, and presence of auditory neuropathy); (b) family-related variables (i.e., parental level of education, socio-economic status, and language/communication mode used at home); and (c) educational/early intervention variables (i.e., communication mode, changes in communication mode over time). Notably, all participating children, regardless of whether they were identified via UNHS or standard care, received the same hearing intervention service from a single national provider (Hearing Australia), at no cost to families.

A range of outcomes have been measured at multiple points in the lives of the LOCHI study participants (i.e., at 6 and 12 months after first fitting with hearing aids or cochlear implants (CIs), and then at each of 3, 5, and 9 years of age) including: receptive and expressive language, functional communication ability, speech perception and production, literacy, educational attainment, and, importantly in the context of this review, psychosocial development and quality of life (QOL).

SOCIAL AND EMOTIONAL DIFFICULTIES AND QUALITY OF LIFE

For the last three decades, researchers have increasingly considered the social and emotional development of children who are DHH. Studies have variously reported on general concepts of psychosocial functioning, often associated with the overall construct of QOL, and sometimes more specifically on mental health and psychopathology. While far from being unanimous in their findings, researchers have frequently shown an increased risk of some social and emotional difficulties associated with decreased QOL and, to a lesser extent, mental health problems (Castellanos et al., 2018; Fellinger et al., 2009; Hindley et al., 1994; Overgaard et al., 2021; Stika et al., 2021; Theunissen et al., 2014a, b).

As broadly consistent as these findings have been, it is notable that there has been considerable heterogeneity in the samples drawn from the diverse population of children and adolescents who are DHH. Multiple dimensions of difference within this population make generalized conclusions problematic. Increasingly, studies have focused on the psychosocial outcomes in subsections of this population, particularly children with CIs. A further limitation of previous research in this area is that most studies were based on cross-sectional designs, often with convenience samples and a limited range of predictor variables, making it difficult to draw conclusions regarding both generalizability and causality for any reported difficulties.

As a longitudinal population-based study, the LOCHI study provides the opportunity to consider the social and emotional development of a population of children at 3, 5, and 9 years of age, and their health-related QOL at 9 years of age. Examining these outcomes and range of independent variables longitudinally provides the opportunity to consider both the prevalence of social and emotional difficulties and possible predictors of those difficulties.

PSYCHOSOCIAL DEVELOPMENT AT 3 YEARS OF AGE

Detailed description of the findings regarding social and emotional development at the 3 year age interval of the LOCHI study were reported by Leigh et al. (2015). At that point, in addition to the usual demographic survey and the range of language and communication assessments, caregivers completed two questionnaires: the Child Development Inventory (CDI; Ireton, 2005) and the Parents' Evaluation of Aural/Oral Performance of Children (PEACH; Ching & Hill, 2007).

The CDI (Ireton, 2005) is a questionnaire-based measure that contains 300 statements that describe observable aspects of child behavior from 15 months to 6 years of age. Although there are eight subscales (Social, Self Help, Gross Motor, Fine Motor, Expressive Language, Language Comprehension, Letters, and Numbers), the two of interest here were those that address aspects of psychosocial development: (a) the Social subscale that probes social behaviors, initiative, independence, and social interaction; and (b) the Self-Help subscale that addresses independence, self-care skills, and personal responsibility. Each subscale has 40 items requiring caregivers to answer Yes or No to indicate whether they have observed a behavior in their child. The PEACH (Ching & Hill, 2007) contains 11 questions that seek caregivers' ratings of children's ability to communicate in quiet and in noise and to respond to sounds in everyday situations, and two questions that address rating their use of their sensory devices. For each question parents are asked to rate the child's ability on a five-point scale and the frequency of occurrence of behaviors observed over a week. It yields an overall functional performance score calculated from the response to the 11 questions.

The most notable finding regarding the children's performance at 3 years of age was that average scores on the two measures of psychosocial development were within the typical range for children of that age. This was a notable contrast with much previous research that concluded DHH children were at higher risk of poorer psychosocial outcomes than their typical hearing same age peers. However, in a trend that would be observed at subsequent age intervals in the LOCHI study, standard deviations were high, with some children achieving scores well below the mean. In contrast with psychosocial scores, mean quotients for the children's language and communication ability and their standard scores on the PEACH were well below the range expected for their typically developing same age peers (Leigh et al., 2015). Further, the children's scores for receptive and expressive language ability measured by the CDI and functional communication ability as measured by the PEACH were significantly correlated with both measures of psychosocial development. Regarding the PEACH, children who were rated by caregivers as being better functional communicators tended to score higher on both social development and self-help skills—raising awareness of functional communication abilities as a predictor of psychosocial strengths.

PSYCHOSOCIAL DEVELOPMENT AT 5 YEARS OF AGE

At 5 years of age, data were available for 356 children whose parents completed at least one of the questionnaire-based assessments of their social and emotional development (Wong et al., 2017, Wong et al., 2018). Once again, assessments included the CDI (Social subscale), the Strengths and Difficulties Questionnaire (SDQ; Goodman, 1997), and the PEACH. The SDQ is a 25-item screening measure designed to identify behavioral and emotional problems in children. It comprises five subscales: conduct problems, hyperactivity, emotional symptoms, peer problems, and prosocial behavior. Each subscale consists of 5 items rated by parents on a three-point response scale from 0 = 'not true', to 1 = 'somewhat true', to 2 = 'certainly true'. Scores from the first four subscales are used to create a "total difficulties score," with higher scores indicating greater social or emotional difficulties and potential psychopathology. Higher scores on the prosocial scale indicate psychosocial strengths. Receptive and expressive language abilities at five years were assessed using the Preschool Language Scale—fourth edition (PLS-4; Zimmerman et al., 2002) and non-verbal cognitive ability was measured using the Wechsler Nonverbal Scale of Ability (WNV; Wechsler & Naglieri, 2006).

Scores for each of the measures were converted to Z-scores using the most recently published normative data. To reduce measurement error and random variation across the various subscales, a “global psychosocial score” was created by averaging the Z-scores for the SDQ total difficulties scale, SDQ prosocial scale, and the CDI social skills scale.

In a pattern of results like those at 3 years of age, the analysis of the children’s global psychosocial scores at 5 years of age showed that, on average, they were within the range of typically developing children (Z-score = -0.67). Notably, however, once again there was high variability among scores (SD=1.05) and a much higher than expected proportion of children falling more than 2 SDs below the normative mean (i.e., 11.7% versus the 2.5% expected in a normal distribution). Further, in contrast with the results at 3 years of age, scores on the Social Skills scale of the CDI were on average 1.4 SDs below age expectation for typically hearing children at that age (Wong et al., 2017).

To determine the possible predictors of these highly variable outcomes, hierarchical multiple regression analyses were conducted with the global psychosocial score as the dependent variable. The factors that predicted better psychosocial functioning were higher non-verbal cognitive ability, the absence of additional disabilities, language ability, and functional communication skills (i.e., as measured by the PEACH). Regarding the latter two predictors, consistent with the findings at 3 years of age, scores on both the PLS-4 and the PEACH were highly significant predictors of global psychosocial outcomes at 5 years old. Most notably, average PEACH scores showed the strongest effect size on global psychosocial outcomes, suggesting that, even if children develop good structural (i.e., morphosyntactic) language abilities, they may experience social or emotional difficulties if they do not communicate effectively at a functional level. The influence of functional communication abilities on psychosocial functioning was an issue deemed particularly important to address at the 9 years old level of the LOCHI study.

PSYCHOSOCIAL DEVELOPMENT AT 9 YEARS OF AGE

Given the impetus to further investigate the predictive value of functional communication skills for children’s psychosocial functioning, the 9 years old stage of the LOCHI study sought to expand the range of functional communication abilities under consideration, and the range of outcomes measured. Regarding the first issue, it was apparent that functional communication ability includes the ability to be understood by others (i.e., speech intelligibility) and the ability to use language socially to manage relationships with others (i.e., pragmatic language abilities). Not only are these important components of any definition of functional language and communication skills, but they have also been shown by others to be associated with psychosocial functioning (Freeman et al., 2017; Zadman-Zait & Most, 2020). Studies that address the potential for relationship between functional communication ability and QOL, however, are less common. Therefore, the 9 years of age interval of the LOCHI study took the opportunity to consider the relationship of functional language use (including both speech intelligibility and pragmatic abilities) to both psychosocial functioning and QOL while controlling for the influence of structural language abilities (Ching et al., 2021).

At the 9 years of age interval there were 144 children for whom data points were available for each of the following predictor and dependent variables:

Predictors

- Structural language ability: Clinical Evaluation of Language Fundamentals—4th Edition (CELF-4; Semmel et al., 2003)
- Nonverbal cognitive ability: Wechsler Nonverbal Scale of Ability (WNV; Wechsler & Naglieri, 2006)
- Pragmatic use of spoken language: Children's Communication Checklist—Second Edition (CCC-2; Bishop, 2003).
- Auditory functional performance: Parents' Evaluation of Aural/Oral Performance of Children (PEACH; Ching & Hill, 2007).
- Speech intelligibility: Speech Intelligibility Rating scale (SIR; Allen et al., 2001).
- Degree of hearing loss.
- Age at intervention (age at first hearing aid fitting).

Dependent Variables

- Behaviour and emotion (psychosocial functioning): Strengths and Difficulties Questionnaire (SDQ, Goodman, 1997)
- Health-related quality of life (HRQOL): Pediatric Quality of Life

> Inventory version 4.0 (PedsQL 4.0; Varni et al., 2001). This inventory comprises 23 items from four domains: physical functioning, emotional functioning, social functioning, and school functioning. Each item is rated on a 5-point Likert scale, from 0 (never a problem) to 4 (almost always a problem). Z-scores were computed for the mean scores for each scale and the total score using published norms (Varni et al., 2003).

As occurred at both the 3- and 5-year age points, the children's mean scores for the SDQ—both the total scale and all subscales—were within the normal range. However, as before, a much larger than expected proportion of children had scores that were clinically significant for psychosocial difficulties. Indeed, 4.9% had total difficulties scores more than two SDs below the mean, in contrast with the normative population where just 2.3% would be expected to fall in that range. Similarly, for the PedsQL, 6.9% of children had total scores that were more than two standard deviations below the normative expectation.

Relationships Among Measures

As expected from previous research findings, better structural language skills were correlated with better psychosocial skills, less behavioral difficulties, and better QOL. There were also significant positive correlations between pragmatic language skills and all subscale and total scores on the PedsQL and SDQ (Ching et al., 2021). Hierarchical multiple regression analyses—accounting for the effects of other variables—provided a much clearer understanding of the contributory significance of the various predictors. Table 1 shows the results of multiple regression analyses with the total difficulties, prosocial, and individual subscale scores of the SDQ and the total, psychosocial health, physical health, and individual scale scores on the PedsQL. Functional auditory abilities (PEACH) and pragmatic language abilities (GCC), together with degree of hearing loss and hearing device (i.e., CI vs hearing aid) were significant predictors of total scores for both the SDQ and the PedsQL, accounting for 33.2% of the total variance. Notably, after accounting for the effects of all other variables, neither receptive nor expressive language ability scores were significant predictors of any SDQ scores (i.e., neither total nor any of the subscale scores). For the PedsQL, functional auditory abilities and pragmatic language abilities

Table 1. Multiple regression model using the total scale scores and individual subscale scores for the SDQ and PedsQL at 9 years of age as dependent variables

PREDICTOR	SDQ												PedsQL											
	SDQ Total		Emotion		Conduct		Hyper-activity		Peer prob.		Prosocial		QOL Total		Psychosocial		School		Social		Emotion		Physical	
	Est	*p*-Value	Est	*p*-Value	Est	*p*-Value	Est	*p*-Value	Est	*p*-Value	Est	*p*-Value	Est	*p*-Value	Est	*p*-Value	Est	*p*-Value	Est	*p*-Value	Est	*p*-Value	Est	*p*-Value
Age at intervention	0.05	0.45	0.02	0.84	0.05	0.51	0.08	0.27	0.001	0.99	- 0.13	0.09	0.08	0.36	0.05	0.52	0.05	0.51	0.03	0.7	0.04	0.67	0.12	0.16
Hearing loss (BE4FA)	**0.47**	**0.003**	**0.45**	**0.01**	**0.42**	**0.018**	**0.33**	**0.04**	0.20	0.26	0.22	0.20	0.29	0.08	**0.34**	**0.04**	0.28	0.08	0.11	0.52	**0.46**	**0.01**	0.22	0.22
Device (HA or CI)	**0.40**	**0.10**	**0.52**	**0.005**	0.33	0.07	0.24	0.16	0.11	0.55	0.09	0.61	0.25	0.14	0.29	0.09	0.1	0.53	0.11	0.55	**0.52**	**0.006**	0.2	0.28
IQ (WNV)	0.13	0.11	- 0.01	0.95	0.14	0.14	**0.23**	**0.01**	0.001	0.99	0.15	0.09	0.06	0.53	0.05	0.56	0.02	0.82	0.03	0.74	0.08	0.42	0.05	0.63
Receptive lang. (CELF)	0.13	0.36	0.17	0.31	0.11	0.48	0.05	0.75	0.07	0.65	- 0.32	0.05	0.16	0.31	0.21	0.18	0.2	0.18	0.12	0.47	0.20	0.23	0.03	0.84
Expressive lang. (CELF)	- 0.13	0.38	- 0.25	0.13	- 0.01	0.94	- 0.06	0.68	- 0.03	0.84	0.22	0.16	- 0.14	0.36	- 0.19	0.21	- 0.07	0.63	- 0.17	0.28	- 0.22	0.18	- 0.01	0.93
Pragmatic lang.: (GCC)	**0.29**	**0.004**	0.19	0.09	0.06	0.6	**0.35**	**<0.001**	0.20	0.08	0.14	0.19	0.27	0.01	**0.25**	**0.02**	**0.30**	**0.003**	**0.28**	**0.01**	0.03	0.78	0.21	0.07
Auditory performance (PEACH)	**0.33**	**<0.001**	**0.31**	**0.002**	**0.32**	**<0.001**	0.17	0.06	**0.21**	**0.08**	**0.23**	**0.01**	0.33	<0.001	**0.38**	**<0.001**	**0.29**	**<0.001**	**0.28**	**0.004**	**0.37**	**0.004**	0.18	0.08
Speech intelligibility (SIR)	-0.10	0.87	-0.04	0.69	0.04	0.65	0.05	0.55	0.11	0.55	- 0.08	0.36	- 0.01	0.87	0.02	0.78	0.07	0.42	- 0.02	0.82	0.01	0.88	- 0.07	0.45
Adjusted R^2	**0.332**		**0.129**		**0.166**		**0.262**		**0.135**		**0.194**		**0.249**		**0.268**		**0.308**		**0.160**		**0.114**		**0.094**	

Bold values indicate significance at $p < 0.05$. Est = parameter estimate (beta).

For SDQ columns: SDQ Total = Total difficulties score of Strengths and Difficulties Questionnaire; Emotion = emotional difficulties score; Conduct = conduct score; Hyperactivity = hyperactivity score; Peer Prob. = peer problems score; Prosocial = prosocial behavior score

For PedsQL columns: QOLT = total score of Pediatric Quality of Life Inventory or PedsQL; Psychosocial = psychosocial health score; School = school functioning score; Social = social functioning score; Emotion = emotional functioning; Physical = physical health score.

For predictors: BE4FA = Better ear four frequency average hearing level (average of hearing threshold level at 0.5, 1, 2, and 4 kHz); HA or CI = hearing aids or cochlear implants; WNV =Wechsler Nonverbal Scale of Ability score; Receptive lang. (CELF) = receptive language score of CELF-4; Expressive lang. (CELF) = expressive language score of CELF-4; GCC = General Communication Composite, pragmatic language score; PEACH = functional hearing score; SIR = Speech Intelligibility Rating.

were the only significant predictors of total scores, accounting for 24.9% of the total variance.

CONCLUSIONS AND IMPLICATIONS FOR INTERVENTION

The investigations of children's psychosocial functioning at each of the three age intervals in the LOCHI study reviewed here have consistently suggested a relationship between language and communication abilities and the children's abilities in the psychosocial domain (Ching et al., 2021; Leigh, et al., 2015; Wong et al., 2017; Wong et al., 2018). Recalling that the genesis of the LOCHI study lay in the question of whether developmental outcomes for DHH children are improved by earlier identification and intervention, it warrants attention that no such relationship was evident for psychosocial development in the analyses that have been reviewed in this paper. What is clear, however, is that language and communication development is affected by the age at which intervention is commenced. Ching et al. (2017) described how, at the 5 year age interval, children whose intervention commenced at 3 months had significantly better language skills than those whose intervention commenced at 12 or 24 months with the effect size increasing with the severity of hearing loss—particularly for children receiving CIs. Indeed, on average, children who received their first CI at age 24 months had average scores for language ability that were 21.4 standard score points (i.e., 1.4 standard deviations) below the average score for children who received their first CI at 6 months. Given the relationships between language abilities and psychosocial abilities, it is apparent therefore that, indirectly through improved language skills, earlier intervention does affect psychosocial development and quality of life.

What has been made clearer by the findings reported across the age intervals of the LOCHI study is the importance of functional language and communication skills in predicting psychosocial abilities and difficulties. At each of the 3-, 5- and 9 year age intervals, functional communication skills, as evidenced in children's scores on the PEACH, showed a significant positive association with psychosocial skills, and ultimately quality of life. Further, pragmatic language skills—an important indicator of functional language ability—were also shown to account for significant positive variance in children's psychosocial skills.

However, even though structural language abilities (i.e., morphosyntactic skills, vocabulary) are foundational to the development of good functional language abilities, the relationship between the latter and children's psychosocial development has proved to be particularly robust. Indeed, at the 9 year age interval, structural language abilities were not significant predictors of SDQ or QOL scores after allowing for the effects of functional auditory performance and pragmatic language abilities.

The findings summarized in this review bear close attention in the determination of strategies for intervention into children's language and communication development. They suggest several critical considerations for early intervention and ongoing educational endeavors with children who are DHH, including: (a) ensuring the assessment of children's language and communication skills (especially their functional communication abilities) from the earliest possible age; (b) ensuring the use of evidence-based strategies to develop and/or to remediate delays or difficulties in both structural and functional language abilities; and (c) ensuring that appropriate assessment strategies are in place to identify children at risk of significant psychosocial or psychological difficulties so that timely referral for specialist intervention can be made to ameliorate that risk.

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LET'S PLAY AND TALK: LINKING CHILDREN'S LANGUAGE AND PRETEND PLAY ABILITIES

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Play, while complex, is essential for children's learning and development. It is well established in the literature that there is a strong link between children's pretend play abilities and their language skills, particularly in relation to the use of language in social contexts. For many children who are deaf and hard of hearing (DHH), pretend play and language skills are often delayed, highlighting the important role of adults in supporting these skills. For many educators working with young DHH children, the focus is often centred on supporting communication and language, and less on play. We argue that play provides a rich and authentic context for supporting both language and pretend play abilities, with many educators requiring professional development to build knowledge and to implement a play-based approach to support children's language and pretend play. This paper outlines the key characteristics of pretend play and highlights the relationship between pretend play and language. Findings from several recent research studies are presented including the outcomes from a qualitative study that implemented a professional learning program that supported educators to implement a play-based approach to foster young DHH children's play and language abilities.

SPOKEN LANGUAGE ABILITIES IN CHILDREN WHO ARE DEAF AND HARD OF HEARING

Language is a complex system of symbols that are socially and culturally shared to help us convey our ideas, thoughts, needs, and desires with others in our world (Paatsch & Nolan, 2020). Language is developed through a multifaceted, dynamic process involving the mastery of the rules for combining these symbols in both the expressive and receptive forms (Creaghe, 2020; Paatsch et al., 2023, Pruden et al., 2006). Children learn language when they are exposed to frequent, rich, meaningful, and socially interactive environments, supported by more sophisticated language users such as caregivers and teachers (Beiting et

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al., 2022). Strong language abilities, particularly the use of language in social contexts (known as pragmatics), are key to successful social interactions. Pragmatics play a critical role in a child's ability to take part in social settings such as home, preschool, school, and the wider community (Paul et al., 2020), and includes skills such as turn-taking, use of eye-gaze, taking the perspective of others, and the ability to be contingent on the contributions made by others in the interaction. Together, these pragmatic skills involve the complex interplay of cognitive abilities, language skills, and social awareness (Toe et al., 2019). Therefore, it is not surprising that language outcomes are strongly linked to academic success and social and emotional abilities, and have been reported to be the best predictor of school readiness and later school success (Hirsh-Pasek et al., 2015; Levine et al., 2016).

Research has shown that many children who are deaf and hard of hearing (DHH) are at risk of language delays despite early identification of hearing loss, early intervention, and greater access to spoken language through more advanced hearing technologies such as cochlear implants and hearing aids (Paatsch & Toe, 2020; Paul et al., 2020). In particular, many DHH children who use Listening and Spoken Language as their main mode of communication have delayed pragmatic skills compared with their peers who have typical hearing, and experience fewer opportunities for interaction with a wide variety of conversational partners. For example, parental ratings of the conversational skills of typical hearing and DHH children aged 12-36 months showed that on average, DHH children were greater than two standard deviations below the ratings for typical hearing children, with fewer than one-third who were within the normal range (Nicholas, 2000). Research exploring primary school-aged DHH children has also shown that their pragmatic skills often differ from those of age-matched typical hearing peers (Church et al., 2017; Paatsch et al., 2017; Paul et al., 2020). Findings from these studies showed that some DHH children were able to take-turns, request for clarification, initiate topics, and answer questions, but did not extend topics, repair conversations when there was a breakdown, or provide responses that were contingent on their conversational partner's contributions. Such findings suggest that many of these pragmatic language delays can create social challenges for many DHH children in forming friendships with peers and participating in classroom activities.

Researchers advocate for the need to provide young DHH children with abundant, rich and authentic opportunities to engage in meaningful interactions with a variety of conversational partners throughout everyday activities (Paatsch et al., 2017; Paatsch & Toe, 2020). Such opportunities provide rich environments for DHH children to collaborate with others, take turns, understand and use appropriate verbal and non-verbal cues necessary for language development, take the perspective of another person, acknowledge the contributions of others during interactions, and provide feedback to their conversational partners. We argue that a critical and naturally occurring context for supporting DHH children's language abilities is through play, particularly pretend play.

PRETEND PLAY

Defining play, while difficult and complex, has resulted in much debate and interest amongst researchers, philosophers, teachers, policymakers, and parents for centuries. However, despite the challenge of accepting one universal definition, there is growing consensus that play is an activity that is intrinsically motivated, fun, social, involves joyful discovery, and is critical for children's learning and development (Yogman et al., 2018; Zosh et al., 2018). Vygotsky (1967) theorizes that it is through play that children learn to use their imagination, to create, to experiment, and to discover, particularly when they play with others more competent at those skills. It is through play that children engage in social interactions where a zone of proximal development (ZPD) is created—the difference between children's current level of independent performance and the level where they can learn when supported by more competent peers and adults.

Yogman and colleagues (2018) reported that "play is not frivolous: it enhances brain structure and function and promotes executive function (i.e., the process of learning), which allows us to pursue goals and ignore distractions" (p. 1). There are many different types of play that children can engage in, including construction play (e.g., building a spaceship with blocks), physical or gross motor play (e.g., climbing, running), visual-perceptual play, games with rules (e.g., chasey), and pretend play (e.g., use of symbols in the play). Pretend play, also known as symbolic, imaginative, and social play, refers to "the non-literal use of objects, action or attributes…that appears to develop on a relatively fixed schedule and is present in every culture" (Creaghe et al., 2021, p. 981). Pretend play involves many distinctive behaviours including: (1) object substitution, where children use an object for something else [e.g., a stick for a person; a block for a computer]; (2) role play, where children may enact the role of a character in play [e.g., person or animal]; (3) play sequences, where children plan a series of actions in their play that become more complex over time; (4) play scripts, where children develop stories with single or multiple problems and resolutions that may commence with reflecting what children do (e.g., pretending to have a drink) then move to more complex stories; and (5) characters in play, where children use dolls and figurines to impose emotions and show an understanding of mental states of others (Elkonin, 2005; Robertson et al., 2020; Stagnitti & Paatsch, 2018). Together these pretend play skills involve higher-order thinking, creativity, imagination, theory of mind, self-regulation, self-awareness, and problem-solving—all skills critical for learning 21st century skills (Yogman et al., 2018). Furthermore, pretend play creates a sense of safety, which reduces stress and anxiety and prepares children for learning (Hirsch-Pasek, 2024; Paatsch et al., 2023).

LINK BETWEEN PRETEND PLAY AND LANGUAGE

There is a substantial body of research that has shown a strong link between pretend play and language acquisition, leading to later literacy development, including understanding and use of narrative language (Creaghe et al., 2021; Lillard et al., 2013). Both language and play, particularly pretend play, share many conceptual similarities including the understanding and use of complex symbols and the reliance on communication within social contexts (Creaghe & Kidd, 2022; Quinn et al., 2018). In addition, both activities depend on the support and scaffolding by competent language users and players (Creaghe & Kidd, 2022; Creaghe et al., 2021; Paatsch et al., 2023; Quinn et al., 2018).

In a recent study by Stagnitti and colleagues (2020) the relationship between children's pretend play abilities and their spoken language skills were investigated in 30 typically hearing children at the commencement of their first year of formal schooling. Results showed that children who were able to substitute objects for something else, particularly those who could use many objects to create a new object or use any object for anything in their play, predicted these children's overall receptive and expressive language abilities. Similar links between pretend play and broad language skills were also reported in a study by

Kizildere et al. (2020) who found that pretend play skills of 119 preschool children at 4 years of age predicted their receptive language skills at 5 years of age. Further research with children with typical hearing has also shown the strong link between pretend play and specific language skills such as semantic development, including vocabulary, semantic problem solving, use of mental state words (Creaghe et al., 2021 Melzer & Palermo, 2016); syntax, including grammatical knowledge (Creaghe et al., 2021; Stagnitti et al., 2016); and pragmatic development, including narrative development, turn-taking, and social interaction (Creaghe et al., 2021; Fekonja et al., 2005; Holmes et al., 2022).

Similar relationships between pretend play and language abilities in DHH children have also been reported. Spencer (1996) investigated the association between pretend (symbolic) play and expressive language in three groups of 2 year olds: (1) deaf children with typical hearing parents; (2) deaf children with deaf parents; and (3) typical hearing children with typical hearing parents. Results showed a consistent pattern of association between language levels and symbolic play but not between play and hearing status, confirming the strong link between expressive language and play. Brown and colleagues (2001) investigated the relationship between pretend play and word production in 10 typical hearing and 10 DHH children at 28, 29, and 30 months of age. Results showed an association between word production and levels of pretend play for both groups, with lower levels of language and play skills evident in the group of DHH children. Similar reports of delayed pretend play and word learning were reported in a later study by Quittner et al. (2016). Specifically, children with cochlear implants who received implants after the age of 2 years were delayed in their achievement of pretend play and novel noun learning when compared to their age-matched typical hearing peers. In a recent study by Da Silva et al. (2022) investigating 12 DHH preschool children's social interactions during recess time, results showed that these children spend less time in social interactions compared to their typical hearing peers. Specifically, these children avoided pretend play and showed a high prevalence of onlooking during recess time, suggesting they may experience social isolation and challenges with forming friendships. This link between social-cognitive development, the social use of language, and pretend play abilities was also compared in a group of 82 DHH children and 91 typical hearing peers, aged between 3 and 8 years (Sidera et al., 2019).

Results showed that the group of DHH children had difficulties in understanding pretend emotions, which was strongly related to their pragmatics and expressive vocabulary skills.

Together, these findings highlight the importance of pretend play as a fertile context for supporting young children's language and communicative development, as they engage in rich meaningful interactions with others. However, the implications of these findings emphasize the need for teachers, teachers of the deaf, and allied health professionals to understand the importance of pretend play and the ways in which they can support children to develop play abilities to foster language learning. The following sections present an example of how a tailored professional learning program supported staff at a school for young DHH children to build knowledge, implement a play-based approach, and change practice.

EMBEDDING A PLAY-BASED APPROACH TO SUPPORT YOUNG DHH CHILDREN'S LANGUAGE DEVELOPMENT

Background

This three-year research project, *Teachers Deliberating Practice*, was undertaken by researchers at Deakin University, Australia (Nolan, Paatsch, and Robertson) in collaboration with staff at a school for DHH children aged birth to 7 years old. Specifically, the researchers were approached by the school leadership to support them to embed a play-based approach to teaching and learning in their school as a context for supporting DHH children's communication and language development.

Research Project Aims

This project aimed to establish a research culture within the school that supported knowledge building and led to practice change relating to a play-based approach to teaching and learning. Specifically, the project focussed on supporting teachers, teachers of the deaf, educators, support staff and allied health professionals to build knowledge about pretend play and language, as they embedded a play-based approach across the school. In addition, the project implemented the Participatory Action Research (PAR; Kemmis & McTaggart, 2005) approach in supporting teachers to research their own practice and document any practice changes as a result of implementing a play-based approach.

Context of the Study

The school offers programs for DHH and deafblind children from diagnosis to 7 years old across the state of Victoria, Australia. Programs include early intervention, early education that includes kindergarten and first year of schooling, and an inclusive 3–4-year-old community kindergarten for DHH and typical hearing children. Staff at the school include teachers, educators, teachers of the deaf, support staff, and allied health professionals including speech-language pathologists, social workers, and physiotherapists.

The Program

The research project consisted of three phases: (1) understanding and building knowledge; (2) researching practice; and (3) building capacity for a sustainable program.

The first phase focused on building staff knowledge in the areas of play and language. Specifically, the first workshop with 60 staff included content knowledge building about the types and principles of play, with a focus on pretend play, the link between play and learning, the components of language, and the interaction between language and play. The second workshop involved building staff knowledge on their role in supporting children's play and language, reflecting on their own talk patterns that facilitate language learning, and the principles of implementing a play-based approach.

Phase two of the research project involved 30 staff participating in three online workshops and two full-day, face-to-face workshops. The aims of this phase were to introduce the staff to assessment of play and the four-step process of PAR (Observe, Reflect, Plan, and Act), as well as to work with them to design a research project around a case study child in their class. This included developing a research question to explore their own practice in supporting children's language with a play-based approach. Exploration included undertaking assessments, videoing their practices, collecting evidence, reflecting, and planning for implementation. Staff completed three cycles of the four-step process of PAR as part of their own research project. At the conclusion of the third cycle, staff participated in a 'Showcase of Practice' where each staff member presented their findings from their PAR to the rest of the staff.

Phase three of the project included four full-day, face-to-face workshops at the school to build capacity for sustainability of the program for the future. The staff and researchers developed a working document that embedded the content and resources that were introduced and developed during the first two phases. Specifically, this involved co-developing a 'Play and Practice: Connecting play and language' document for the school that included research-evidence and teacher planning and assessment documents specific to play and language, as well as ways to support a play-based approach in the school. Further sections of the document included key considerations for implementing PAR, and ways of working with colleagues within a transdisciplinary practice approach.

The Outcomes

Throughout the three phases of the research project, staff constantly discussed their deepening understandings of play and language. At the end of Phase two, staff were invited to complete an online survey to share their learnings as a result of the project. Overall results showed that while "there's a lot to learn from all these workshops", staff had gained a better understanding of the ways in which young DHH children's play and language abilities can be supported through a play-based approach. In particular, staff noted that "play is an integral part of developing communication skills" and that "play opportunities are everywhere". Staff also reported on the many features of Phase two that supported their learning including the "new knowledge and putting it into practice", "the analysis process of videos and refining the research question", "having time to view videos, discuss and plan with colleagues", and "that the action research cycle allows me to reflect critically on my practice and then implement a plan that is specific to my students".

Staff also commented on the value of the "Showcase of Practice". One staff member commented that they "enjoyed hearing the perspective of my peers as to what play looks like in their individual spaces and how ongoing reflection is key to improving our programs and supporting our students." Other staff commented on how the presentations "helped to articulate our understandings", "to consolidate the learning", and to "see the students' growth in play and language", while others reported that listening to colleagues was "amazing" as "we learnt a lot" from "different perspectives".

Staff were also invited to reflect on the differences they made to their practice during participation in Phase 2. Staff saw the importance of setting up an authentic play environment to encourage children to use language with their peers and teachers, and the need to "add play as part of their goals, not just language". Some also changed their practices to be more involved with the children. As one staff member reported, "I was more reflective about what was going on, what role I was playing, and how I could shift this to challenge the child." Another teacher commented:

> "My level of involvement in my student's play needed to increase as I discovered from watching the videos of her play, that she needed more modelling on how to extend her play from the few repetitive actions she knew for a given scenario. So instead of just setting up experiences for her to play with her peers, I would join her in play and model."

CONCLUSION

This paper has presented research relating to the key characteristics of pretend play and the relationship between pretend play and language. The findings from research have also shown that many DHH children are often delayed in their pretend play and language skills, with many researchers highlighting the need to provide professional development for those who work with these children in supporting these skills. The current research project outlined in this paper has highlighted one example of how researchers and staff in one school in Victoria, Australia, worked together to build knowledge in play and language and to implement this new knowledge into practice. Educators, teachers of the deaf, education support staff, and allied health professionals valued working across transdisciplinary teams to build their understandings, to change current practice, and to develop a sustainable program for all staff working in the school. These staff can make a difference to children's learning by taking time to build knowledge of children's pretend play abilities, the ways in which a play-based approach provides a rich context for language learning, and to reflect on their own practices from an evidence-informed perspective. However, further research is warranted to investigate the specific pretend play and language outcomes for DHH children and the relationship between these outcomes and teachers' practice when implementing a play-based approach.

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The Volta Review, Volume 124(1)

FUELING SPOKEN LANGUAGE DEVELOPMENT THROUGH AUDITORY ACCESS

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Children can only develop spoken language through consistent exposure to the acoustic cues that comprise speech and language. Until recently, hearing levels from the clinical audiogram were the primary measure used to define typical hearing and the presence or degree of a child's hearing loss. While the clinical audiogram remains an important diagnostic measure for audiologists, the degree of hearing loss from the audiogram is an inconsistent indicator of a child's functional hearing capacity, particularly once the child is fitted with hearing aids or cochlear implants. Speech audibility is an estimate of a child's access to the acoustic cues that comprise spoken language and have recently been used to accurately quantify a child's functional hearing levels with and without devices. This article will review the limitations of quantifying auditory access based on the clinical audiogram, as well as approaches for estimating speech audibility for children with hearing loss.

The audiogram is a clinical measure of hearing sensitivity based on a child's behavioral responses to narrowband or pure-tone stimuli across a frequency range that is most important for understanding speech. The audiogram can also be estimated based on neural responses to tone burst or chirp stimuli from the auditory brainstem response or auditory steady-state response in infants or children who cannot complete a behavioral hearing test. The audiogram can be divided into ranges that quantify the amount or degree of hearing loss. Figure 1 shows an audiogram of a child with mild hearing loss in both ears. The audiogram is often used to make a medical diagnosis of hearing loss. However, the audiogram also has some functional limitations, particularly for measuring the impact of hearing loss on communication.

Although the audiogram measures hearing levels related to the discrete sound frequencies that are most important for understanding speech, research has suggested that the degree of hearing loss on the audiogram is not a strong predictor of how well a child with hearing loss

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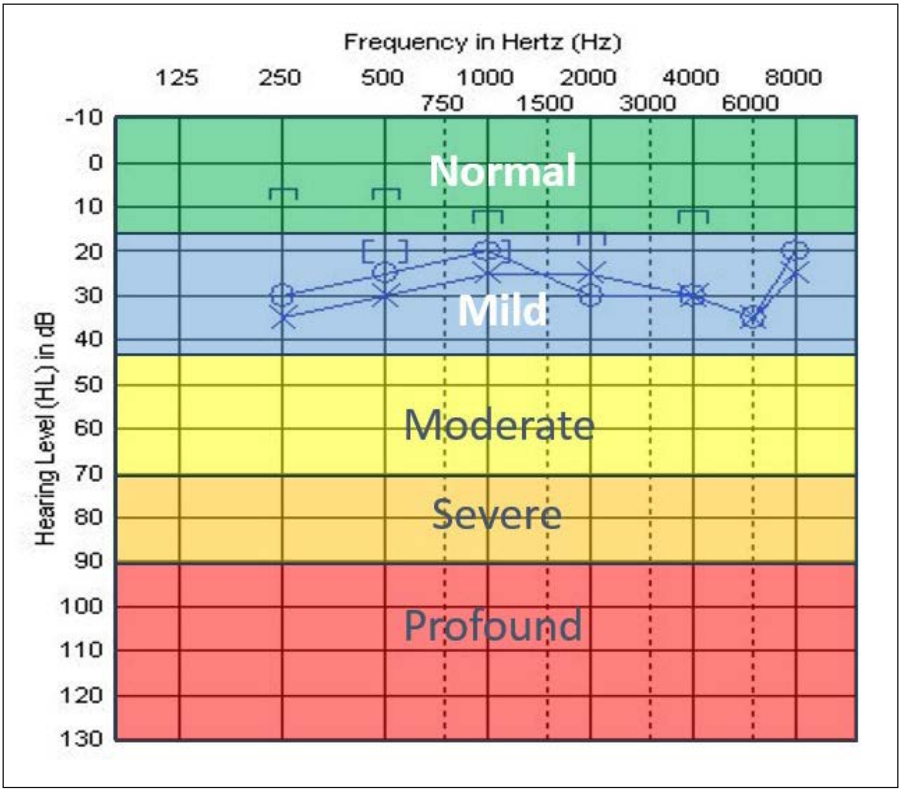


Figure 1. An audiogram for a child with mild hearing loss in both ears. Circles are hearing thresholds for the right ear, and triangles are hearing thresholds for the left ear. Brackets are bone conduction thresholds. Shaded areas represent different degrees of hearing loss: Normal = Green, Mild hearing loss = Blue, Moderate hearing loss = Yellow, Severe hearing loss =Orange, and Profound hearing loss = Red.

will develop spoken language or perform academically in school. Several studies have suggested that children with mild degrees of hearing loss may be at greater risk for delays in spoken language (Walker et al., 2020) and academic challenges (Tomblin et al., 2020) than children with moderate or greater degrees of hearing loss. The intervention that a child with hearing loss receives is much more likely to predict a child's developmental progress than the amount of hearing loss from the audiogram. For example, Walker and colleagues (2015) found that children with mild bilateral hearing loss who received and consistently used hearing aids had stronger language outcomes than children with mild bilateral hearing loss who did not use hearing aids. Likewise, children with hearing aids that were fitted to optimize their audibility for speech had greater language growth (Tomblin et al., 2015), executive function skills (McCreery & Walker, 2022), and math and reading outcomes (Tomblin et al., 2020) than children with less audibility through their hearing aids. The degree of hearing loss from the audiogram does not reflect these differences in intervention for children with hearing loss.

Based on this research and other studies, our research team has been developing new methods to quantify auditory access that may be more useful metrics for parents and clinicians who support children with hearing loss and are more effective at predicting risk and resilience among children who use hearing aids.

UNAIDED SPEECH AUDIBILITY

Unaided speech audibility is a measure that represents the proportion of speech energy that a child can hear without a hearing aid in a specific listening situation. The most common listening situation that we use to estimate audibility is speech at an average conversational level when the talker is one meter away facing the listener, which is approximately 60 dB SPL. We can use the hearing threshold levels from the audiogram to estimate the proportion of speech at an average conversational level that a child can hear with their hearing thresholds. Figure 2 shows the unaided speech spectrum plotted with the right ear audiogram from the child with mild to moderate hearing loss from Figure 1.

We can measure the amount of the speech spectrum that falls above a child's thresholds by converting the audiogram thresholds to dB SPL to allow direct comparison to speech using a graph called the SPL-o-gram (Figure 2). The SPL-o-gram allows us to visualize how much of the speech signal is audible in the right ear for an average speech signal without a hearing aid (unaided). We can also apply a calculation known as the speech intelligibility index (SII; ANSI S3.5-2007) that numerically quantifies the weighted proportion of the speech spectrum that is audible or above the child's thresholds. In the example in Figure 2, the unaided SII for the right ear is 0.47. This means that for an average conversational level in a quiet listening situation, the child hears approximately 47% of the cues for spoken language. Visually, most of the speech cues that are audible in this example are at frequencies below 2000 Hz. Parents of children with hearing loss are often surprised to learn that mild hearing loss means that less than half of the speech spectrum is audible in quiet.

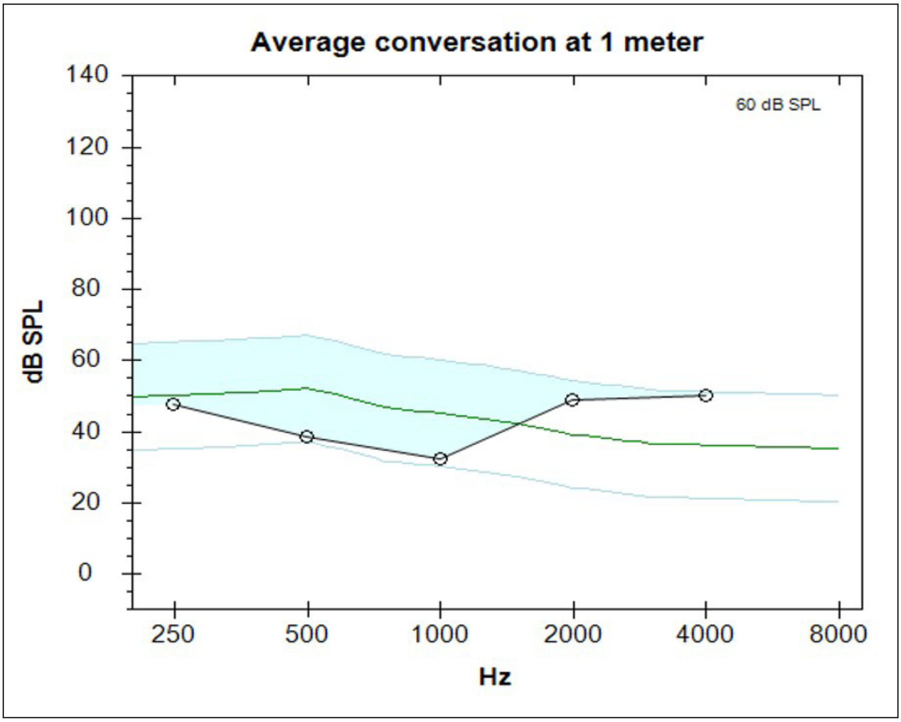


Figure 2. An SPL-o-gram for the right ear of the child with mild hearing loss from Figure 1. Note that the y-axis has typical hearing at the bottom and increasing sound levels as the y-axis increases. The frequencies from the audiogram remain on the x-axis. The circles connect with the solid black line are the hearing thresholds for the right ear converted to dB SPL. The green line represents the average level of speech at 60 dB SPL across frequency, and the blue lines represent the range of speech levels. The blue shaded area represents the portion of the speech spectrum that is audible in the right ear without hearing aids.

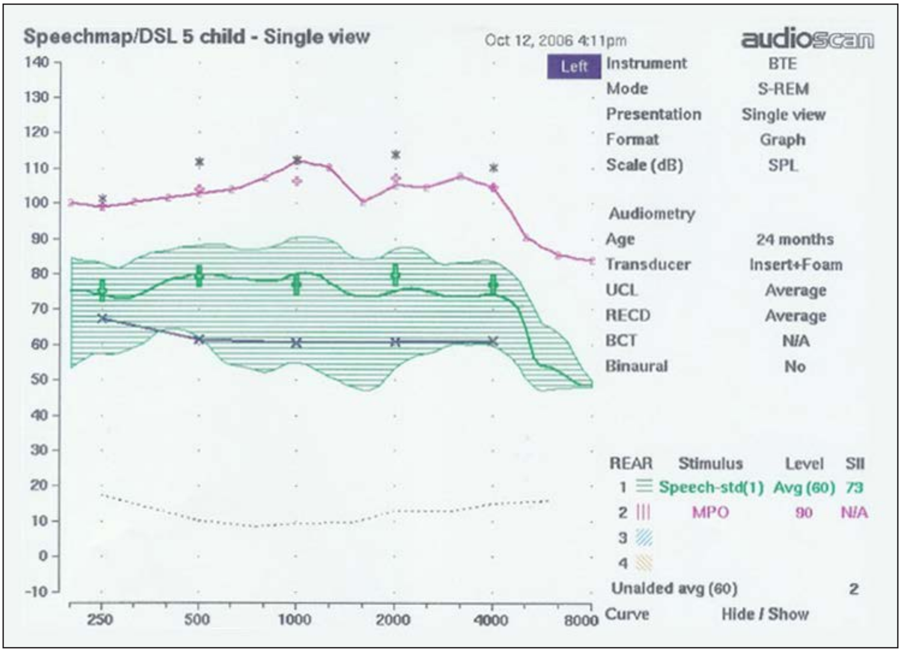
A study by Sapp and colleagues (2023) compared the level of parental concern based on traditional descriptive categories of hearing loss (e.g. mild, moderate, severe, and profound) compared to audibility-based descriptions of hearing loss and hearing loss simulations. Parents felt that hearing loss simulations and audibility-based descriptions of hearing loss more accurately conveyed the functional impact of hearing loss than using terms like “mild” to describe the problem. Measures of unaided audibility can be useful counseling tools for parents of children with hearing loss or other professionals who may be involved in the child’s medical or educational support.

AIDED SPEECH AUDIBILITY

Audibility can also be calculated with hearing aids based on measurements of the hearing aid output in the child's ear canal or in a coupler with appropriate correction factors, known as aided audibility. Aided audibility is based on the same SII calculation as unaided audibility, but the measures of hearing aid output are compared to the child's hearing thresholds instead of a standard unaided speech spectrum. Figure 3 shows an example of an SPL-o-gram for aided audibilty. In this example, the amplified speech spectrum is compared to the child's thresholds from the left ear on the audiogram. Without amplification, the child can hear 2% of the speech spectrum (unaided SII) and with the hearing aid, the child can hear 73% of the average speech spectrum (aided) based on the SII. As noted above, the aided SII is a useful tool for estimating how much access a child has to speech through their hearing aids. Currently, there is no standardized method for calculating the speech audibility for children who use cochlear implants, as there is no way to directly measure how much of the speech spectrum is accessible through the cochlear implant.

One of the limitations of the aided SII is that the amount of speech audibility that can be restored through a hearing aid decreases as the child's degree of hearing loss increases (McCreery et al., 2013). This means that there is no optimal SII value that can be achieved across children who have varying degrees of hearing loss. The Western Ontario Pediatric Amplification Protocol has published normative values for the aided SII as a function of the degree of hearing loss that are available for clinicians and built into many hearing aid verification systems (Bagatto et al., 2016). A recent study by Wiseman and colleagues (2023) showed that children with hearing loss who use hearing aids who had aided SII less than 50% were at risk of language delays compared to children with typical hearing, even if hearing aids were consistently used. This work suggests that an aided SII in quiet where less than half of the aided speech spectrum is audible may not provide adequate audibility to support spoken language development and may require modifications to the child's intervention, including a more powerful hearing aid or referral for cochlear implant evaluation if greater audibility cannot be achieved through acoustic hearing.

The audiogram is an important measure for diagnosis and quantification of hearing loss. However, the audiogram is known not to reflect



Instrument BTE
Mode S-REM
Presentation Single view
Format Graph
Scale (dB) SPL

Audiometry
Age 24 months
Transducer Insert+Foam
UCL Average
RECD Average
BCT N/A
Binaural No

REAR	Stimulus	Level	SII
1 ☰	Speech-std(1)	Avg (60)	73
2 ▥	MPO	90	N/A
3 ▨			
4 ▨			
Unaided avg (60)			2
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Figure 3. An SPL-o-gram showing aided audibility. The X symbols connected with the blue line represent the child's hearing thresholds from the audiogram in the left ear. The green shaded area is the speech spectrum measured through the child's hearing aid. The aided (73) and unaided (2) SII values for average speech (60 dB SPL) are shown on the bottom right side of the graph.

the functional listening challenges that children with hearing loss experience in their daily lives. Speech audibility, as measured by the SII, is a way to use the audiogram along with either unamplified or amplified speech spectra to estimate the proportion of the speech that will be audible to children in different listening situations. Clinicians can use measurements of audibility to counsel parents and caregivers of children with hearing loss who use hearing aids to help increase understanding of how hearing loss may impact communication. Parents of children with hearing loss can ask their child's audiologist or other care providers about speech audibility to help understand how much hearing aids can support a child's communication access.

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CONGENITAL CYTOMEGALOVIRUS (CMV) INFECTION: A COMMON YET OVERLOOKED CAUSE OF HEARING LOSS AND NEURODEVELOPMENTAL DISORDERS

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Congenital cytomegalovirus (cCMV) infection is a prevalent yet under-recognized cause of sensorineural hearing loss and neurodevelopmental disorders in children. While CMV infection is typically asymptomatic in healthy individuals, its transmission during pregnancy can result in significant and lasting health consequences for the fetus. Despite its global impact, awareness of cCMV remains low, particularly among pregnant women and healthcare professionals, contributing to the under-diagnosis and inadequate management of affected infants. This manuscript reviews the epidemiology, clinical manifestations, and long-term outcomes of cCMV infection, emphasizing the critical need for preventive measures, including increased awareness and education. The paper also discusses the role of screening programs, highlighting the potential benefits of universal or targeted approaches in early detection and intervention. Despite advances in understanding cCMV, significant gaps remain in knowledge about its long-term cognitive effects and the effectiveness of various interventions. Further research is needed to explore the lived experiences of individuals with cCMV and their caregivers, identify gaps in healthcare, and develop comprehensive, family-centered intervention strategies.

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WHAT IS cCMV INFECTION?

CMV is a herpesvirus transmitted through body fluids that establishes lifelong latency after the initial infection. This means the virus can reactivate later in life, and individuals can also be reinfected with different strains of CMV. For most people, the infection is either asymptomatic or causes only mild "flu-like" symptoms and fever, without serious health issues.

However, in individuals with weakened immune systems—such as those with HIV or recent transplants—CMV can lead to severe illness (Taylor, 2003). Congenital CMV (cCMV) infection occurs when the virus is transmitted from a pregnant woman to her unborn fetus through the placenta, a process known as vertical transmission. Most pregnant women acquire the infection through contact with saliva from young children who shed CMV more readily than adults (Dollard et al., 2014). For example, sharing utensils or kissing a toddler on the mouth can lead to CMV transmission. To prevent CMV infection, pregnant women are advised to practice good hygiene routines, such as always washing hands after changing diapers or wiping a young child's nose, and avoiding the sharing of utensils, food, and beverages with others (Fowler & Boppana, 2018).

The risk of transmission is significantly higher if the mother contracts a primary CMV infection during pregnancy (32%) compared to a recurrent infection (1.4%) (Kenneson & Cannon, 2007). With an average prevalence of 0.5%, cCMV is the most common congenital virus infection in developed countries (Orb et al., 2024; Ssentongo et al., 2021) In developing regions across Africa, Asia and Latin America, prevalence rates range from 0.6% to 6.1% (Kenneson & Cannon, 2007).

Research by Fowler and colleagues (2018) has highlighted the impact of race and ethnicity on the prevalence of cCMV. They found that black infants and infants of multiracial origin might have a significantly higher risk of contracting cCMV. These findings underscore the need for preventive strategies to raise awareness of cCMV among all women of childbearing age and for healthcare professionals to recognize group differences related to race and origin (Fowler et al., 2018). Notably, no significant differences were found between races or ethnicities in the likelihood of children having symptomatic cCMV infection or associated hearing loss.

Seroprevalence refers to the proportion of adults or older children in a population who have detectable antibodies against a specific pathogen, such as CMV, in their blood. This measure increases with age, an indication of how widespread CMV is within a community or population at a given time (Fowler et al., 2022). In regions with higher seroprevalence, there are typically higher rates of cCMV infection, even though the risk of infecting unborn children is greater in cases of primary infection (De Vries et al., 2013).

Approximately 1 in 5 infants born with cCMV infection may have birth defects or long-term health issues. Congenital CMV infection has also been associated with fetal death and stillbirths in some studies (Griffiths et al., 1980; Iwasenko et al., 2011). Children born with cCMV infection are typically categorized into two groups; those with symptomatic cCMV (10%) and asymptomatic cCMV (90%) at birth (Kenneson & Cannon, 2007).

Symptomatic cCMV infection refers to cases where the individual exhibits clinical manifestations of the infection at birth. In congenital cases, these symptoms might include an enlarged liver, jaundice, rash, sensorineural hearing loss, vision problems, and neurological issues such as microcephaly, seizures, or developmental delays (Naing et al., 2016). These symptoms are often detected at birth or shortly thereafter and can lead to serious long-term health issues (Lanzieri et al., 2017).

Asymptomatic cCMV infection refers to cases where the infected individual does not show observable symptoms or signs of illness in early life (Stoyell et al., 2024) or later childhood (Lopez et al., 2017; Novelli et al., 2022). Around 90% of children with cCMV are asymptomatic during the newborn period (Gantt et al., 2017). Although these individuals may not display clinical manifestations at the time of diagnosis, they can still carry the virus and might develop late-onset hearing loss. Around 10–15% of asymptomatic infants with cCMV will develop permanent sensorineural hearing loss (Fowler & Boppana, 2018). Additionally, while visual and cognitive impairments can occur, they are less compared to those observed in symptomatic cases of cCMV (Dollard et al., 2007).

IMPACT OF cCMV INFECTION

Congenital CMV infection is a leading cause of acquired unilateral or bilateral sensorineural hearing loss in children, which can range from mild to profound (Goderis et al., 2014). This hearing loss may be present at birth, progress over time, or develop as a late-onset condition during childhood. Due to the potential for progressive or delayed hearing loss, ongoing auditory monitoring is crucial. Timely interventions are essential to support language and communication development, ensuring that the affected children receive the necessary resources and care.

Cochlear implantation is used for children with cCMV and bilateral deafness, but it is also beneficial for those with single-sided deafness (SSD) (Aldè et al., 2023). Unilateral cochlear implantation in children with cCMV and SSD can significantly improve their ability to localize sounds and understand speech in noisy environments. Additionally, there is a considerable risk to losing hearing in the unaffected ear over time for individuals with cCMV. By promoting early cochlear implantation in children with SSD, the child can preserve as much hearing capability as possible, supporting their overall auditory and speech development (Aldè et al., 2023).

Hearing loss is a prominent neurodevelopmental disorder resulting from cCMV infection (Goderis et al., 2014), but it does not fully account for the range of cognitive difficulties observed in children with cCMV infection (Karltorp et al., 2014). Children with hearing loss and cCMV infection are at risk for various cognitive impairments, including poorer executive functioning (Löfkvist et al., 2020). In a study by Löfkvist and colleagues (2020), pragmatic skills were assessed through parent questionnaires, revealing that children with cCMV had significantly worse pragmatic skills and phonological working memory compared to well-matched controls with genetic cause of deafness. These cognitive challenges can, in turn, negatively impact academic achievement and social interactions (Löfkvist et al., 2020).

Furthermore, cCMV is also associated with vestibular problems (Dhondt et al., 2021; Karltorp et al., 2014; Kokkola et al., 2023; Shears et al., 2022). In a systematic review, Shears et al. (2022) demonstrated that even children with asymptomatic cCMV infection, including those with typical hearing, may experience vestibular dysfunction, which can be progressive. Karltorp et al. (2014) investigated comorbidities in children with hearing loss comparing those to controls with a genetic cause of deafness (connexin 26), which typically does not include cognitive issues. They found that children with cCMV had poorer balance, vision loss, unexpected eating disorders, and neuropsychiatric diagnoses such as autism spectrum disorder and ADHD, while the controls did not exhibit these issues.

Previous studies have also shown that children with cCMV infection may have intellectual disabilities (Lanzieri et al., 2017), neuropsychiatric diagnoses like autism spectrum disorder, ADHD, and oral motor dysfunctions (Karltorp et al., 2014). A large population-based study by Pesch and colleagues (2024) found that American children with cCMV have a 2.5 times higher risk of developing autism spectrum disorder than those who were uninfected (Pesch et al., 2024). Comorbid conditions are common in children with cCMV, contributing to the high heterogeneity within this population (Karltorp et al., 2014). Some children with cCMV may exhibit no or few symptoms, while others experience multiple severe conditions. It is more likely that children with symptomatic cCMV infection will have more severe conditions (Lanzieri et al., 2017).

However, a unique long-term follow-up study by Falkenius Schmidt and colleagues (2024) found that adults with cCMV infection who had typical hearing and were asymptomatic at birth experienced deficits in word retrieval processes. This cognitive ability is assessed using word fluency tasks such as the FAS task, which requires individuals to retrieve as many words as possible beginning with the letters F, A, and S within one minute. These findings suggest that the long-term effects of cCMV infection may persist even in individuals who initially showed no symptoms, indicating potential subtle cognitive difficulties that may not be easily detected at first glance.

One limitation of the study is its small sample size (n=34), and the findings will need to be replicated in further research. However, a strength of the study was the inclusion of a control group of individuals who were uninfected at birth. When compared to Swedish normative data, 43% of participants with cCMV infection, all of whom were asymptomatic at birth, performed adequately on both FAS and verb fluency tasks, compared to 86% of the controls (Falkenius Schmidt et al., 2024).

Korndewal and colleagues (2017) investigated how daily life is affected for school-aged children with cCMV and their parents. They found that the quality of life for both the children with long-term impairments and their parents was lower compared to controls without cCMV infection. Additionally, parents of children with cCMV reported more physical issues and problems with concentration than those of controls with children without cCMV. These results highlight that cCMV impacts the daily life experiences of both the affected children and their family members, indicating a need for a family-centered intervention approach (Korndewal et al., 2017). While individuals with symptomatic cCMV infection often have more complex needs, it is important to recognize that those with asymptomatic cCMV may also have similar needs.

PREVENTION

Currently, there are no effective CMV vaccines available. Developing such a vaccine is particularly challenging due to the complex nature of CMV immunity, which includes the potential for reactivation of a previous infection and the risk of infection with new viral strains. Although ongoing research into CMV vaccine candidates shows promise (Hu et al., 2024; Schleiss, 2016), it is unlikely that an effective vaccine will be available in the near future. Consequently, the most effective strategy for preventing maternal and congenital CMV infection remains educating pregnant women about the sources of CMV exposure and promoting behavioral interventions to reduce these risks (Calvert et al., 2021). Additionally, increasing public awareness about cCMV infection and promoting hygiene practices are essential to minimize virus transmission.

Despite the need for greater public awareness and the prevalence of cCMV infection, many pregnant women remain unaware of cCMV and its potential serious consequences, as well as preventive actions (Calvert et al., 2021). Studies indicate significant knowledge gaps, with awareness rates ranging from 13% to 60% in various countries (Binda et al., 2016; Cordier et al., 2012; Pereboom et al., 2013). Midwives, who regularly interact with pregnant women, play a crucial role in disseminating information about CMV and can help prevent its spread by educating women on protective actions. Other healthcare professionals, such as physicians, pediatricians, educational audiologists, speech-language pathologists, and early interventionists, also need to be well-informed about the impact of cCMV infection.

An informal International Congenital Cytomegalovirus Recommendations Group, established by Rawlinson et al., (2017) aimed to provide guidelines for the prevention, diagnosis, and treatment of cCMV. The group, based on consensus discussions and review of the literature, concluded universal neonatal screening for cCMV may be considered to facilitate early detection and intervention for hearing loss and/or developmental delay. They also agreed that education and prevention strategies for pregnant women are beneficial and that recommendations should be updated as new data and knowledge become available (Rawlinson et al., 2017).

SCREENING OF cCMV INFECTION

Currently, most children born with cCMV infection worldwide remain unrecognized, resulting in a lack of necessary treatment or follow-up care. Diagnosis of cCMV is ideally made through laboratory tests performed on the infant within the first three weeks of life. If cCMV is suspected after three weeks, retrospective analysis can be conducted using dried blood spots (DBS) PCR, although this is not a highly sensitive method to diagnose cCMV. In Sweden, DBS samples are stored indefinitely, but in most countries, they are retained for only a limited time, making retrospective diagnosis more challenging.

Universal or hearing-targeted screening programs are implemented in only a few regions worldwide, leading to the detection, identification, and treatment of children with symptomatic cCMV infection. Ontario, Canada, launched the world's first universal CMV screening program in 2019, and in 2023 Minnesota became the first state in the United States to implement a universal CMV screening program. Several other U.S. states either have adopted hearing-targeted screening programs, expanded-targeted screening programs, or are campaigning for such programs.

A significant limitation of hearing-targeted screening is that the majority of all children born with cCMV infection pass the universal newborn hearing screening, but may develop progressive hearing loss later. This presents a challenge because caregivers are often unaware of cCMV or the possibility of progressive hearing loss, potentially missing critical early listening and phonological milestones crucial for language development (Blamey et al., 2001; Ching et al., 2017). Additionally, atypical brain patterns associated with cCMV may impact long-term outcomes (Lanzieri et al., 2017), and not only in those with symptomatic cCMV (Falkenius Schmidt et al., 2024).

In Utah, the targeted screening program introduced in 2013 has evolved into an *expanded targeted screening program.* This updated approach includes additional high-risk groups, such as those with abnormal head size (microcephaly), maternal CMV positivity, or infants with petechial rash or blueberry muffin rash (Suarez et al., 2023). The expanded targeted program has shown improved detection rates of symptomatic cCMV cases compared to traditional hearing targeted screening. Suarez and colleagues (2023) argue that expanded screening is a more feasible alternative to universal screening, requiring fewer tests while achieving higher detection rates.

Regardless of type of cCMV screening program, providing a follow-up program for those identified with cCMV infection and their families is essential, as outlined by screening principles (WHO, 2020). Studies indicate that parents generally support cCMV screening programs, even if they must cover costs out of pocket (Diener et al., 2020), and despite initial concerns that the results may be distressing (Beaulieu et al., 2024).

In the patient-led study by Beaulieu et al. (2024), which used a Patient and Community Engagement Research (PaCER) approach encompassing focus-group discussions, interviews, and reflections, parents with lived experience expressed a strong preference to be informed about their child's health condition from the outset. They valued "knowledge as power" and believed that universal screening could reduce stigma by increasing awareness about cCMV. The study participants also highlighted the importance of having a follow-up program after screening to support families (Beaulieu et al., 2024).

FOLLOW-UP PROCEDURES, TREATMENT AND INTERVENTION OPTIONS – CURRENT BEST PRACTICE

If a child is diagnosed with cCMV infection at birth or within the first three weeks of life and exhibits moderate to severe symptoms, international guidelines recommend a six-month course of antiviral treatment with valganciclovir (Leruez-Ville et al., 2024). This treatment aims to reduce the risk of severe neurodevelopmental disorders (Rawlinson et al., 2017). However, the long-term outcomes and potential side effects of this treatment are not fully understood. Consequently, antiviral treatment is not recommended for cases with mild symptoms (Rawlinson et al., 2017). In addition to medical treatment, it is crucial to monitor the child's hearing, vision, and overall development over time. According to Rawlinson et al. (2017), audiological testing should be conducted at six-month intervals during the first three years of life for children with cCMV, and then annually until adolescence. Early ophthalmological examinations and follow-up assessments are also recommended. Developmental assessments should be carried out for those with symptomatic cCMV and should be tailored to each individual case. There are no specific recommendations for asymptomatic cases, though ongoing developmental monitoring is advisable (Rawlinson et al., 2017).

To date, only a few studies have investigated the long-term impact of cCMV in adulthood (Falkenius Schmidt et al., 2024). Existing evidence suggests that individuals with symptomatic cCMV infection are at high risk for enduring and comorbid complications, emphasizing the necessity for early, preventive interdisciplinary interventions. However, early intervention alone may not be sufficient; many individuals with cCMV infection may require ongoing monitoring and support throughout their lives. Those who develop neurodevelopmental disorders in early childhood may also face an increased risk of neuropsychiatric conditions later in adolescence or adulthood, as seen in other populations with co-existing disorders such as autism spectrum disorder or ADHD (Gillberg, 2010). Currently, there are no established best practice guidelines specifically addressing the cognitive and comorbid aspects of cCMV intervention. Nonetheless, research and clinical experience highlight the importance of not only monitoring hearing but also identifying other potential issues, such as non-verbal cognitive difficulties, neuropsychiatric symptoms, atypical spoken language development, poor pragmatics, affected executive functions, and evaluate overall quality of life (Karltorp et al., 2014; Korndewal et al., 2017; Löfkvist et al., 2020).

Given the complexity of cCMV infection and the variability in outcomes, children should be monitored regularly by an interdisciplinary team with specialized knowledge in cCMV. It is essential that individuals with cCMV and their families are actively involved in the intervention process from the beginning. A notable example of this approach is the Hearing House in Auckland, New Zealand, where Dr. Holly Teagle and colleagues initiated a comprehensive CMV program (Raising awareness of congenital Cytomegalovirus on the local and global stage, hearinghouse.co.nz). This program includes workshops where families can engage in education and discussion, and meet with an interdisciplinary CMV team for personalized assessment and intervention. The team comprises audiologists, speech-language pathologists, educational psychologists, occupational and physiotherapists, developmental pediatricians, ENT surgeons, early interventionists, and family members.

The CMV Aoeteara project group is actively investigating various aspects of congenital CMV (cCMV) care in New Zealand. Their research includes examining national CMV database findings, assessing healthcare professionals' knowledge and attitudes, evaluating holistic and child-centered interdisciplinary teamwork, and gathering parental perspectives on CMV screening and diagnosis. Although the program's outcomes have not yet been scientifically evaluated or published, data collection and analysis are currently in progress.

CONCLUSIONS

The global impact of cCMV infection is significant, affecting a large number of infants and often leading to long-term neurodevelopmental disorders and other health complications. With no effective vaccine currently available, preventive measures such as increasing awareness about cCMV transmission and promoting protective behaviors are critical. As the benefits of newborn screening programs—whether universal or targeted—become more evident, their implementation is likely to expand worldwide. While existing research has predominantly focused on the medical aspects of cCMV infection, there remain substantial gaps in knowledge, particularly regarding the long-term cognitive effects in broader populations and the efficacy of various intervention strategies besides antiviral medication and hearing technology. Furthermore, additional research is essential to explore the lived experiences of individuals with cCMV infection and their caregivers, to better identify healthcare gaps, and to address the comprehensive needs of those affected by the infection in clinical settings.

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THE VOLTA REVIEW

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HABILIDADES PSICOSOCIALES Y CALIDAD DE VIDA DE NIÑOS CON SORDERA/HIPOACUSIA: LECCIONES DE LOS PRIMEROS DIEZ AÑOS DEL ESTUDIO LOCHI

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En este artículo se analizan los resultados de los 10 primeros años del estudio Longitudinal Outcomes of Children with Hearing Impairment (LOCHI), que incluye a más de 450 niños australianos con sordera/hipoacusia cuya pérdida auditiva se identificó a través del cribado auditivo neonatal o en procesos posteriores de confirmación e intervención.

El estudio LOCHI (Longitudinal Outcomes of Children with Hearing Impairment, Resultados longitudinales de niños con discapacidad auditiva) es un estudio prospectivo basado en la población de Australia en el que, desde 2005, se evalúa periódicamente una amplia gama de resultados de una cohorte de niños con sordera/hipoacusia para investigar la eficacia de la identificación y la intervención tempranas, así como los factores que explican la variabilidad de los resultados.

Antes del estudio LOCHI, la investigación sobre el efecto de la identificación temprana se componía principalmente de (a) comparaciones retrospectivas de muestras de conveniencia de niños que a menudo procedían de un único servicio de intervención temprana, educativo o audiológico que tendían a mostrar los efectos positivos de los proveedores de identificación temprana (p. ej., Calderon y Naidu, 2000; Moeller, 2000; Yoshinaga-Itano et al., 1998); o (b) estudios basados en la población que tendían a mostrar los beneficios nulos o limitados de la identificación temprana de la pérdida auditiva (p. ej., Kennedy et al., 2006; Korver et al., 2010).

Dada la amplia aceptación del cribado auditivo neonatal universal y el grado en que los gobiernos de todo el mundo se comprometieron a

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aplicarlo, fue una anomalía que las pruebas de su eficacia en las fases iniciales de su desarrollo no fueran sólidas. De hecho, ya en 2008, en una revisión sistemática (Nelson et al., 2008) se señalaba que solo existía «una certeza moderada de que el beneficio neto del cribado auditivo en todos los recién nacidos fuera moderado», y se destacaba la necesidad de disponer de pruebas procedentes de exámenes prospectivos sobre si la identificación temprana de la pérdida auditiva se traducía en mejores resultados educativos y de desarrollo en los niños que nacían con una pérdida auditiva. En este contexto se concibió el estudio LOCHI.

DISEÑO DEL ESTUDIO LOCHI

La necesidad de un estudio prospectivo sobre la eficacia de la identificación temprana de la pérdida auditiva planteaba un importante dilema de diseño para los investigadores en contextos en los que el cribado auditivo neonatal ya se realizaba a nivel poblacional. En tales jurisdicciones, la asignación aleatoria de los niños al cribado o no cribado no sería ética y la idea de asignarlos aleatoriamente a grupos de tratamiento (intervención temprana frente a intervención posterior) sería aún más grave desde un punto de vista ético. Para superar la necesidad de la asignación aleatoria, en el estudio LOCHI se aprovechó un periodo temporal en Australia entre 2002 y 2007, cuando varios estados australianos estaban implementando programas de cribado auditivo neonatal a diferentes ritmos. Los tres estados más poblados (Nueva Gales del Sur, Victoria y Queensland) iniciaron sus programas en un periodo de tres años, entre 2002 y 2005. Nueva Gales del Sur implantó el cribado auditivo neonatal en todas las regiones del estado simultáneamente, mientras que en Queensland y Victoria el despliegue del programa se escalonó por regiones a lo largo de 2 y 6 años, respectivamente (Leigh, 2006).

Al tratarse de un estudio prospectivo, se invitó a participar a todos los niños identificados con pérdida auditiva que habían nacido en esos tres estados durante un periodo de 5 años entre 2002 y 2007, lo que dio lugar a una cohorte de aproximadamente 470 niños que se dividió de forma natural entre los que fueron identificados a través del cribado auditivo neonatal y los que fueron identificados más tarde, lo que permitió que la edad de identificación fuera una variable predictiva principal en un diseño de investigación prospectivo. Además, también se recopilaron datos sobre otras muchas variables predictivas, como (a) variables relacionadas con el niño(a) (es decir, peso al nacer, sexo, edad de identificación/diagnóstico, grado de pérdida auditiva, edad de adquisición del audífono, edad de implantación coclear, capacidad cognitiva no verbal, presencia de discapacidades adicionales y presencia de neuropatía auditiva); (b) variables relacionadas con la familia (es decir, nivel educativo de los padres, estatus socioeconómico y modo de lenguaje/comunicación utilizado en el hogar); y (c) variables educativas/de intervención temprana (es decir, modo de comunicación y cambios en el modo de comunicación a lo largo del tiempo). Cabe destacar que todos los niños participantes, independientemente de si habían sido identificados a través del cribado auditivo neonatal o de la atención sanitaria estándar, recibieron el mismo servicio de intervención auditiva de un único proveedor nacional (Hearing Australia), sin coste alguno para las familias.

En el estudio LOCHI se mide una serie de resultados en varios momentos de la vida de los participantes (es decir, a los 6 y 12 meses después de la primera adquisición de audífonos o implantes cocleares (IC), y posteriormente a los 3, 5 y 9 años de edad), incluyendo: lenguaje receptivo y expresivo, capacidad de comunicación funcional, percepción y producción del habla, lectoescritura, logros educativos y, lo que es más importante en el contexto de esta revisión, desarrollo psicosocial y calidad de vida (CdV).

DIFICULTADES SOCIALES Y EMOCIONALES, Y CALIDAD DE VIDA

En las últimas tres décadas se aprecia que los investigadores prestan cada vez más atención al desarrollo social y emocional de los niños con sordera/hipoacusia. Los estudios se han centrado en conceptos generales de funcionamiento psicosocial, a menudo asociados con el concepto general de CdV y, en ocasiones, más específicamente en la salud mental y la psicopatología. Si bien sus hallazgos distan de ser unánimes, los investigadores suelen destacar la presencia de un mayor riesgo en el caso de algunas dificultades sociales y emocionales asociadas con una disminución de la CdV y, en menor medida, con problemas de salud mental (Castellanos et al., 2018; Fellinger et al., 2009; Hindley et al., 1994; Overgaard et al., 2021; Stika et al., 2021; Theunissen et al., 2014a, b).

A pesar de la consistencia general de estos hallazgos, cabe destacar la existencia de una considerable heterogeneidad en las muestras extraídas de la diversa población de niños y adolescentes con sordera/hipoacusia. Las múltiples dimensiones de la diferencia dentro de esta población significan que las conclusiones generalizadas sean problemáticas. Cada vez más, los estudios se centran en los resultados psicosociales en subsecciones de esta población, concretamente en niños con IC. Otra limitación de la investigación previa en esta área es que la mayoría de los estudios se basan en diseños transversales, a menudo con muestras de conveniencia y una gama limitada de variables predictoras, lo que dificulta la extracción de conclusiones con respecto tanto a la capacidad de generalización como a la causalidad de cualquier dificultad notificada.

Como estudio longitudinal basado en la población, el estudio LOCHI ofrece la oportunidad de considerar el desarrollo social y emocional de una población de niños a la edad de 3, 5 y 9 años, y su CdV relacionada con la salud a los 9 años. El examen longitudinal de estos resultados y de una serie de variables independientes ofrece la oportunidad de considerar tanto la prevalencia de las dificultades sociales y emocionales como los posibles factores predictivos de estas dificultades.

DESARROLLO PSICOSOCIAL A LA EDAD DE 3 AÑOS

Una descripción detallada de los hallazgos relativos al desarrollo social y emocional en el intervalo de edad de 3 años del estudio LOCHI fue aportada por Leigh et al. (2015). En ese momento, además de la encuesta demográfica habitual y la gama de evaluaciones del lenguaje y la comunicación, los progenitores cumplimentaban dos cuestionarios: Child Development Inventory (CDI, Inventario de desarrollo infantil; Ireton, 2005) y Parents' Evaluation of Aural/Oral Performance of Children (PEACH, Evaluación de los progenitores del desempeño auditivo/oral en niños; Ching y Hill, 2007).

El CDI (Ireton, 2005) es una medida basada en un cuestionario que contiene 300 enunciados que describen aspectos observables del comportamiento infantil desde los 15 meses hasta los 6 años. Aunque existan ocho subescalas (Social, Autoayuda, Motricidad gruesa, Motricidad fina, Lenguaje expresivo, Comprensión del lenguaje, Letras y Números), las dos que nos interesaron fueron las que abordaban aspectos del desarrollo psicosocial: (a) la subescala Social, que trata de los comportamientos sociales, la iniciativa, la independencia y la interacción social; y (b) la subescala Autoayuda, que aborda la independencia, las habilidades de autocuidado y la responsabilidad personal. Cada subescala tiene 40 ítems y se requiere que los progenitores respondan Sí o No para indicar si han observado un determinado comportamiento en su hijo(a). El PEACH (Ching y Hill, 2007) incluye 11 preguntas que requieren la valoración por parte de los progenitores de la capacidad de los niños de comunicarse en silencio y en presencia de ruido, y de responder a sonidos en situaciones cotidianas, y dos preguntas que abordan la valoración del uso de sus dispositivos sensoriales. Para cada pregunta se pide a los progenitores que valoren la capacidad del niño(a) en una escala de cinco puntos y la frecuencia de aparición de determinados comportamientos observados a lo largo de una semana. Se obtiene una puntuación global del desempeño funcional calculada a partir de la respuesta a las 11 preguntas.

El hallazgo más notable en relación con el desempeño de los niños a la edad de 3 años fue que las puntuaciones medias en las dos medidas de desarrollo psicosocial estaban dentro del rango típico de los niños de esa edad. Esto contrastaba notablemente con muchas investigaciones anteriores que concluían que los niños con sordera/hipoacusia corrían un mayor riesgo de obtener peores resultados psicosociales que sus compañeros con una audición típica de la misma edad. Sin embargo, en una tendencia que se observaría en intervalos de edad posteriores en el estudio LOCHI, las desviaciones típicas (DT) fueron altas y algunos niños obtuvieron puntuaciones muy por debajo de la media. En contraste con las puntuaciones psicosociales, los cocientes medios de la capacidad lingüística y comunicativa de los niños, así como sus puntuaciones estándar en el PEACH, estaban muy por debajo del rango esperado en sus compañeros de la misma edad con un desarrollo típico (Leigh et al., 2015). Además, las puntuaciones de los niños correspondientes a la capacidad de lenguaje receptivo y expresivo medida por el CDI, así como la capacidad de comunicación funcional medida por el PEACH, se correlacionaban significativamente con ambas medidas de desarrollo psicosocial. En cuanto al PEACH, los niños que fueron puntuados por los progenitores como mejores comunicadores funcionales tendían a puntuar más alto tanto en desarrollo social como en habilidades de autoayuda, lo que plantea la cuestión de si las habilidades de comunicación funcional podrían seguir estando más asociadas con fortalezas psicosociales.

DESARROLLO PSICOSOCIAL A LA EDAD DE 5 AÑOS

A la edad de 5 años, había datos disponibles de 356 niños cuyos padres habían cumplimentado al menos una de las evaluaciones basadas en cuestionarios sobre su desarrollo social y emocional (Wong et al., 2017, Wong et al., 2018). De nuevo, en las evaluaciones se incluyeron el CDI (subescala Social), el SDQ (Strengths and Difficulties Questionnaire, Cuestionario de fortalezas y dificultades; Goodman, 1997) y el PEACH. El SDQ es una medida de cribado de 25 ítems diseñada para identificar problemas conductuales y emocionales en niños. Se compone de cinco subescalas: problemas de conducta, hiperactividad, síntomas emocionales, problemas con los compañeros y comportamiento prosocial. Cada subescala consta de 5 ítems valorados por los padres en una escala de respuesta de tres puntos: 0 = «no es cierto», 1 = «algo cierto», 2 = «totalmente cierto». Las puntuaciones de las cuatro primeras subescalas se utilizan para obtener una «puntuación de dificultades totales», indicando las puntuaciones más altas mayores dificultades sociales o emocionales y una posible psicopatología. Las puntuaciones más altas en la escala prosocial indican fortalezas psicosociales. Las capacidades relativas al lenguaje receptivo y expresivo a los cinco años se evaluaron con la Escala del lenguaje preescolar, cuarta edición (PLS-4; Zimmerman et al., 2002) y la capacidad cognitiva no verbal se midió utilizando la Escala de capacidad no verbal de Wechsler (WNV; Wechsler y Naglieri, 2006).

Las puntuaciones de cada una de las medidas se convirtieron en puntuaciones Z utilizando los datos normativos publicados más recientemente. Para reducir el error de medición y la variación aleatoria entre las distintas subescalas, se obtuvo una «puntuación psicosocial global» promediando las puntuaciones Z de la escala de dificultades totales del SDQ, la escala prosocial del SDQ y la escala de habilidades sociales del CDI.

En un patrón de resultados similares a los de los 3 años, el análisis de las puntuaciones psicosociales globales de los niños a los 5 años demostró que, en promedio, se encontraban dentro del rango de los niños con un desarrollo típico (puntuación Z = -0,67). Sin embargo, una vez más se observó una gran variabilidad entre las puntuaciones (DT=1,05) y una proporción mucho mayor de la esperada de niños que se situaban más de 2 DT por debajo de la media normativa (es decir, el 11,7 % frente al 2,5 % esperado en una distribución normal). Además, en contraste con los resultados a los 3 años, las puntuaciones en la escala de Habilidades sociales del CDI se situaban en promedio 1,4 DT por debajo de la expectativa de edad de los niños con una audición típica (Wong et al., 2017).

Para determinar los posibles predictores de estos resultados tan variables se realizaron análisis de regresión múltiple jerárquica con la puntuación psicosocial global como variable dependiente. Los factores que predijeron un mejor funcionamiento psicosocial fueron una mayor capacidad cognitiva no verbal, la ausencia de discapacidades adicionales, la capacidad lingüística y las habilidades funcionales de comunicación (es decir, según lo medido por el PEACH). En cuanto a los dos últimos predictores, en consonancia con los hallazgos a la edad de 3 años, las puntuaciones tanto en el PLS-4 como en el PEACH fueron predictores altamente significativos de los resultados psicosociales globales a la edad de 5 años. En concreto, las puntuaciones medias del PEACH mostraron la mayor magnitud del efecto sobre los resultados psicosociales globales, lo que sugiere que, incluso si los niños desarrollan buenas capacidades lingüísticas estructurales (es decir, morfosintácticas), pueden experimentar dificultades sociales o emocionales si no se comunican eficazmente a un nivel funcional. La influencia de las capacidades comunicativas funcionales en el funcionamiento psicosocial fue una cuestión que se consideró que era especialmente importante abordar en el nivel de 9 años del estudio LOCHI.

DESARROLLO PSICOSOCIAL A LA EDAD DE 9 AÑOS

Considerando el interés de investigar más a fondo el valor predictivo de las habilidades de comunicación social en el funcionamiento psicosocial de los niños, en la fase de 9 años del estudio LOCHI se amplió el rango de capacidades de comunicación funcional consideradas y el rango de resultados medidos. En cuanto a la primera cuestión, era evidente que la capacidad de comunicación funcional incluye la capacidad de ser comprendido por los demás (es decir, la inteligibilidad del habla) y la capacidad de utilizar el lenguaje socialmente para gestionar las relaciones con los demás (es decir, las capacidades lingüísticas pragmáticas). Estos componentes no solo son importantes en cualquier definición de habilidades funcionales del lenguaje y la comunicación, sino que también otros investigadores han demostrado que están asociados con el funcionamiento psicosocial (Freeman et al., 2017; Zadman-Zait y Most, 2020). Sin embargo, son pocos los estudios en los que se aborda la posible relación entre la capacidad de comunicación funcional y la CdV. Por lo tanto, en el intervalo de edad de 9 años del estudio LOCHI se consideró la relación del uso funcional del lenguaje (incluyendo la inteligibilidad del habla y las capacidades pragmáticas) tanto con el funcionamiento psicosocial como con la CdV mientras se controlaba la influencia de las capacidades lingüísticas estructurales (Ching et al., 2021).

En el intervalo de edad de 9 años había 144 niños de los que se disponía de puntos de datos para cada una de las siguientes variables predictoras y dependientes:

Predictores

- Capacidad de lenguaje estructural: CELF-4, 4ª edición (Clinical Evaluation of Language Fundamentals, Evaluación clínica de los fundamentos del lenguaje; Semmel et al., 2003)
- Capacidad cognitiva no verbal: WNV (Wechsler Nonverbal Scale of Ability, Escala de capacidad no verbal de Wechsler; Wechsler y Naglieri, 2006)
- Uso pragmático del lenguaje hablado: CCC-2, 2ª edición (Children's Communication Checklist, Lista de verificación de la comunicación en niños; Bishop, 2003).
- Desempeño funcional auditivo: PEACH (Parents' Evaluation of Aural/Oral Performance of Children, Evaluación de los progenitores del desempeño auditivo/oral en niños; Ching y Hill, 2007).
- Inteligibilidad del habla: SIR (Speech Intelligibility Rating scale, escala de Puntuación de inteligibilidad del habla; Allen et al., 2001).
- Grado de la pérdida auditiva.
- Edad en el momento de la intervención (edad en la primera adquisición de audífonos).

Variables dependientes

- Comportamiento y emoción (funcionamiento psicosocial): SDQ (Strengths and Difficulties Questionnaire, Cuestionario de fortalezas y dificultades; Goodman, 1997)
- Calidad de vida relacionada con la salud (CVRS): PedsQL, versión 4.0 (Pediatric Quality of Life Inventory, Inventario pediátrico de

> calidad de vida; Varni et al., 2001). Este inventario se compone de 23 ítems en 4 áreas: funcionamiento físico, funcionamiento emocional, funcionamiento social y funcionamiento escolar. Cada ítem se valora en una escala Likert de 5 puntos, de 0 (nunca es un problema) a 4 (casi siempre es un problema). Se calcularon puntuaciones Z para las puntuaciones medias de cada escala y la puntuación total utilizando normas publicadas (Varni et al., 2003).

Tal como se observó a los 3 y 5 años, las puntuaciones medias de los niños en el SDQ (tanto en la escala total como en todas las subescalas) se encontraban dentro del rango normal. Sin embargo, al igual que entonces, una proporción de niños mucho mayor de la esperada presentaba puntuaciones clínicamente significativas de dificultades psicosociales. De hecho, el 4,9 % obtuvo puntuaciones de dificultades totales más de dos DT por debajo de la media, en contraste con la población normativa en la que se esperaría que solo el 2,3 % se situara en ese rango. Del mismo modo, en el PedsQL, el 6,9 % de los niños presentaba unas puntuaciones totales que situaban más de dos desviaciones típicas por debajo de la expectativa normativa.

Relaciones entre las medidas

Tal como se esperaba a partir de los resultados de investigaciones anteriores, unas mejores capacidades lingüísticas estructurales se correlacionaban con mejores habilidades psicosociales, menores dificultades conductuales y mejor CdV. También se observaron correlaciones positivas significativas entre las capacidades lingüísticas pragmáticas y todas las subescalas y puntuaciones totales en el PedsQL y el SDQ (Ching et al., 2021). Los análisis de regresión múltiple jerárquica, que tienen en cuenta los efectos de otras variables, proporcionaron una comprensión mucho más clara de la importancia de la contribución de los distintos predictores. En la Tabla 1 se muestran los resultados de los análisis de regresión múltiple con las puntuaciones de las subescalas de dificultades totales, prosocial e individual del SDQ y las puntuaciones de las escalas total, de salud psicosocial, de salud física e individual del PedsQL. Las capacidades auditivas funcionales (PEACH) y las capacidades lingüísticas pragmáticas (GCC), junto con el grado de pérdida auditiva y el dispositivo auditivo (es decir, IC frente

Tabla 1. Modelo de regresión múltiple utilizando como variables dependientes las puntuaciones totales de la escala y las puntuaciones de las subescalas individuales del SDQ y el PedsQL a la edad de 9 años.

PREDICTOR	**SDQ**												**PedsQL**											
	SDQ Total		**Emoción**		**Conducta**		**Hiperactividad**		**Prob. compañ.**		**Prosocial**		**CdV total**		**Psicosocial**		**Escolar**		**Social**		**Emoción**		**Física**	
	Est	**Valor *p***	**Est**	**Valor *p***	**Est**	**Valor *p***	**Est**	**Valor *p***	**Est**	**Valor *p***	**Est**	**Valor *p***	**Est**	**Valor *p***	**Est**	**Valor *p***	**Est**	**Valor *p***	**Est**	**Valor *p***	**Est**	**Valor *p***	**Est**	**Valor *p***
Edad en la intervención	0,05	0,45	0,02	0,84	0,05	0,51	0,08	0,27	0,001	0,99	- 0,13	0,09	0,08	0,36	0,05	0,52	0,05	0,51	0,03	0,7	0,04	0,67	0,12	0,16
Pérdida auditiva (NAP4F)	**0,47**	**0,003**	**0,45**	**0,01**	**0,42**	**0,018**	**0,33**	**0,04**	0,20	0,26	0,22	0,20	0,29	0,08	**0,34**	**0,04**	0,28	0,08	0,11	0,52	**0,46**	**0,01**	0,22	0,22
Dispositivo (audífono o IC)	**0,40**	**0,10**	**0,52**	**0,005**	0,33	0,07	0,24	0,16	0,11	0,55	0,09	0,61	0,25	0,14	0,29	0,09	0,1	0,53	0,11	0,55	**0,52**	**0,006**	0,2	0,28
CI (WNV)	0,13	0,11	- 0,01	0,95	0,14	0,14	**0,23**	**0,01**	0,001	0,99	0,15	0,09	0,06	0,53	0,05	0,56	0,02	0,82	0,03	0,74	0,08	0,42	0,05	0,63
Leng. receptivo (CELF)	0,13	0,36	0,17	0,31	0,11	0,48	0,05	0,75	0,07	0,65	- 0,32	0,05	0,16	0,31	0,21	0,18	0,2	0,18	0,12	0,47	0,20	0,23	0,03	0,84
Leng. expresivo (CELF)	- 0,13	0,38	- 0,25	0,13	- 0,01	0,94	- 0,06	0,68	- 0,03	0,84	0,22	0,16	- 0,14	0,36	- 0,19	0,21	- 0,07	0,63	- 0,17	0,28	- 0,22	0,18	- 0,01	0,93
Leng. pragmático (GCC)	**0,29**	**0,004**	0,19	0,09	0,06	0,6	**0,35**	**<0,001**	0,20	0,08	0,14	0,19	0,27	0,01	**0,25**	**0,02**	**0,30**	**0,003**	**0,28**	**0,01**	0,03	0,78	0,21	0,07
Desempeño auditivo (PEACH)	**0,33**	**<0,001**	**0,31**	**0,002**	**0,32**	**<0,001**	0,17	0,06	**0,21**	**0,08**	**0,23**	**0,01**	0,33	<0,001	**0,38**	**<0,001**	**0,29**	**<0,001**	**0,28**	**0,004**	**0,37**	**0,004**	0,18	0,08
Inteligibilidad del habla (SIR)	-0,10	0,87	-0,04	0,69	0,04	0,65	0,05	0,55	0,11	0,55	- 0,08	0,36	- 0,01	0,87	0,02	0,78	0,07	0,42	- 0,02	0,82	0,01	0,88	- 0,07	0,45
***R*² ajustado**	**0,332**		**0,129**		**0,166**		**0,262**		**0,135**		**0,194**		**0,249**		**0,268**		**0,308**		**0,160**		**0,114**		**0,094**	

Los valores en **negrita** indican significación a p <0,05. Est. = Estimación del parámetro (beta).

Para las columnas del SDQ: SDQ Total = puntuación de dificultades totales del Cuestionario de fortalezas y dificultades; Emoción = puntuación de dificultades emocionales; Conducta = puntuación de conducta; Hiperactividad = puntuación de hiperactividad; Prob. compañ. = puntuación de problemas con compañeros; Prosocial == puntuación de comportamiento prosocial.

Para las columnas del PedsQL: CdV total = puntuación total del Inventario de Calidad de Vida Pediátrica o PedsQL; Psicosocial = puntuación de salud psicosocial; Escolar = puntuación de desempeño escolar; Social = puntuación de funcionamiento social; Emoción = funcionamiento emocional; Física = puntuación de salud física.

Para los predictores: NAP4F = nivel auditivo con promedio de cuatro frecuencias en el mejor oído (promedio del nivel de umbral auditivo a 0,5, 1, 2 y 4 kHz); IC = implantes cocleares; WNV = puntuación de la escala de capacidad no verbal de Wechsler; Leng. receptivo (CELF) = puntuación de lenguaje receptivo del CELF-4; Leng. expresivo (CELF) = puntuación de lenguaje expresivo del CELF-4; GCC = Compuesto de comunicación general, puntuación de lenguaje pragmático; PEACH = puntuación de audición funcional; SIR = puntuación de inteligibilidad del habla.

a audífonos), fueron predictores significativos de las puntuaciones totales tanto del SDQ como del PedsQL, y representaron el 33,2 % de la varianza total. Cabe destacar que, tras tener en cuenta los efectos de todas las demás variables, las puntuaciones de la capacidad de lenguaje receptivo y expresivo no fueron predictores significativos de ninguna puntuación del SDQ (es decir, ni de la puntuación total ni de ninguna de las subescalas). En el caso del PedsQL, las capacidades auditivas funcionales y las capacidades lingüísticas pragmáticas fueron los únicos predictores significativos de las puntuaciones totales, representando el 24,9 % de la varianza total.

CONCLUSIONES E IMPLICACIONES PARA LA INTERVENCIÓN

Las investigaciones sobre el funcionamiento psicosocial de los niños en cada uno de los tres intervalos de edad del estudio LOCHI aquí revisadas sugieren de una manera sistemática una relación entre las capacidades lingüísticas y comunicativas y las habilidades de los niños en el área psicosocial (Ching et al., 2021; Leigh, et al., 2015; Wong et al., 2017; Wong et al., 2018). Recordando que la génesis del estudio LOCHI se basó en la cuestión de si los resultados en el desarrollo de los niños con sordera/hipoacusia mejoraban con una identificación y una intervención más tempranas, merece la pena destacar que no se evidenció tal relación en el caso del desarrollo psicosocial en los análisis revisados en este documento. Lo que está claro, sin embargo, es que el desarrollo del lenguaje y de la comunicación se ve afectado por la edad a la que se inicie la intervención. Ching et al. (2017) describen cómo, en el intervalo de edad de 5 años, los niños cuya intervención había comenzado a los 3 meses tenían unas habilidades lingüísticas significativamente mejores que aquellos cuya intervención había comenzado a los 12 o 24 meses, con una magnitud del efecto que aumentaba con la severidad de la pérdida auditiva, en concreto, en el caso de los niños que recibían IC. De hecho, en promedio, los niños que habían recibido su primer IC a los 24 meses mostraban unas puntuaciones medias de capacidad lingüística que se encontraban 21,4 puntos de puntuación estándar (es decir, 1,4 desviaciones típicas) por debajo de la puntuación media de los niños que habían recibido su primer IC a los 6 meses. Considerando las relaciones entre las capacidades lingüísticas y las capacidades psicosociales, es evidente que, indirectamente a través de la mejora de las capacidades lingüísticas, la intervención temprana afecta al desarrollo psicosocial y a la calidad de vida.

Lo que queda más claro con los resultados obtenidos en los intervalos de edad del estudio LOCHI es la importancia de las habilidades funcionales del lenguaje y de la comunicación para predecir las capacidades y las dificultades psicosociales. En cada uno de los intervalos de edad de 3, 5 y 9 años, las habilidades comunicativas funcionales, tal y como se evidencian en las puntuaciones de los niños en el PEACH, mostraron una asociación positiva significativa con las habilidades psicosociales y, en última instancia, con la calidad de vida. Además, se demostró que las capacidades lingüísticas pragmáticas (un indicador importante de la capacidad lingüística funcional) también explican una varianza positiva significativa en las habilidades psicosociales de los niños. No obstante, si bien las capacidades lingüísticas estructurales (es decir, las habilidades morfosintácticas y el vocabulario) son fundamentales para el desarrollo de buenas capacidades lingüísticas funcionales, la relación entre estas últimas y el desarrollo psicosocial de los niños ha demostrado ser especialmente sólida. De hecho, en el intervalo de edad de 9 años, las capacidades lingüísticas estructurales no fueron predictores significativos de las puntuaciones en el SDQ o en la CdV después de tener en cuenta los efectos del desempeño auditivo funcional y las capacidades lingüísticas pragmáticas.

Los hallazgos resumidos en esta revisión merecen una atención especial a la hora de determinar las estrategias de intervención en el desarrollo del lenguaje y la comunicación de los niños. Sugieren varias consideraciones críticas para la intervención temprana y los esfuerzos educativos continuos con niños con sordera/hipoacusia, entre ellas: (a) realizar una evaluación de las habilidades lingüísticas y comunicativas de los niños (especialmente sus capacidades comunicativas funcionales) a una edad lo más temprana posible; (b) utilizar estrategias basadas en la evidencia para desarrollar y/o paliar los retrasos o dificultades tanto en las capacidades lingüísticas estructurales como en las funcionales; y (c) garantizar que existan estrategias de evaluación apropiadas para identificar a los niños en riesgo de experimentar dificultades psicosociales o psicológicas significativas, de modo que se pueda realizar una derivación oportuna para la intervención de especialistas y mitigar este riesgo.

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JUGUEMOS Y HABLEMOS: VINCULACIÓN DE LAS CAPACIDADES LINGÜÍSTICAS Y DEL JUEGO SIMBÓLICO DE LOS NIÑOS

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El juego, aunque sea complejo, es esencial para el aprendizaje y el desarrollo de los niños. Está bien establecido en la literatura la existencia de un gran vínculo entre las capacidades del juego simbólico o de simulación de los niños y sus habilidades lingüísticas, especialmente en relación con el uso del lenguaje en contextos sociales. En el caso de muchos niños con sordera/hipoacusia, las capacidades lingüísticas y del juego de simulación suelen sufrir un retraso, lo que pone de relieve el importante papel de los adultos a la hora de apoyar estas habilidades. Para muchos educadores que trabajan con niños pequeños con sordera/hipoacusia, la atención se suele centrar más en apoyar la comunicación y el lenguaje, y menos en el juego. Sostenemos que el juego proporciona un contexto propicio y auténtico que refuerza tanto las capacidades lingüísticas como las del juego de simulación, y que muchos educadores necesitan una formación profesional que les permita adquirir conocimientos e implementar un enfoque basado en el juego para apoyar el lenguaje y el juego de simulación de los niños. En este artículo se esbozan las características clave del juego de simulación y se destaca la relación entre el juego de simulación y el lenguaje. Asimismo, se presentan los hallazgos de varios estudios de investigación recientes, incluidos los resultados de un estudio cualitativo en el que se puso en práctica un programa de aprendizaje profesional de apoyo a los educadores para implementar un enfoque basado en el juego y destinado a fomentar las capacidades de juego y lingüísticas de los niños pequeños con sordera/hipoacusia.

CAPACIDADES DE LENGUAJE HABLADO DE LOS NIÑOS CON SORDERA/HIPOACUSIA

El lenguaje es un sistema complejo de símbolos que se comparten social y culturalmente para ayudarnos a transmitir nuestras ideas, pen-

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samientos, necesidades y deseos con otras personas en nuestro mundo (Paatsch y Nolan, 2020). El lenguaje se desarrolla a través de un proceso multifacético y dinámico que implica el dominio de las reglas de combinación de estos símbolos, tanto en la forma expresiva como en la receptiva (Creaghe, 2020; Paatsch et al., 2023, Pruden et al., 2006). Los niños aprenden el lenguaje cuando están expuestos a entornos frecuentes, propicios, significativos y socialmente interactivos, con el apoyo de usuarios del lenguaje más sofisticados, como son los cuidadores y los maestros (Beiting et al., 2022). Unas capacidades lingüísticas sólidas, en especial el uso del lenguaje en contextos sociales (lo que se conoce como pragmática), son fundamentales para el éxito en las interacciones sociales. La pragmática desempeña un papel fundamental en la capacidad del niño para participar en contextos sociales como el hogar, el preescolar, la escuela y la comunidad en general (Paul et al., 2020), e incluye habilidades como la toma de turnos, el uso de la mirada, la adopción de la perspectiva de los demás y la capacidad de tener en cuenta las contribuciones realizadas por otras personas en la interacción. En conjunto, estas habilidades pragmáticas implican la interacción compleja de capacidades cognitivas, habilidades lingüísticas y conciencia social (Toe et al., 2019). Por lo tanto, no es sorprendente que los resultados del lenguaje tengan una gran vinculación con el éxito académico y las capacidades sociales y emocionales, y se reconoce que son el mejor predictor de la preparación escolar y el éxito académico posterior (Hirsh-Pasek et al., 2015; Levine et al., 2016).

En los estudios de investigación se demuestra que muchos niños con sordera/hipoacusia corren el riesgo de sufrir retrasos en el lenguaje a pesar de la identificación temprana de la pérdida auditiva, la intervención temprana y el mayor acceso al lenguaje hablado a través de tecnologías auditivas más avanzadas, como los implantes cocleares y los audífonos (Paatsch y Toe, 2020; Paul et al., 2020). En concreto, muchos niños con sordera/hipoacusia que utilizan la Escucha y el Lenguaje Hablado como su principal modo de comunicación muestran un retraso en las habilidades pragmáticas, en comparación con sus compañeros con una audición normal, y tienen menos oportunidades de interacción con una amplia variedad de interlocutores conversacionales. Por ejemplo, las puntuaciones de los padres con respecto a las habilidades conversacionales de niños con audición normal y niños con sordera/hipoacusia de entre 12 y 36 meses señalan que, en promedio, las puntuaciones de los niños con sordera/

hipoacusia tienen más de dos desviaciones típicas por debajo de las puntuaciones de los niños con audición normal, con menos de un tercio dentro del rango normal (Nicholas, 2000). En los estudios de investigación en los que se estudia a niños con sordera/hipoacusia en la etapa escolar primaria también se destaca que sus habilidades pragmáticas a menudo difieren de las de sus compañeros con audición normal emparejados por edades (Church et al., 2017; Paatsch et al., 2017; Paul et al., 2020). En los hallazgos de estos estudios se muestra que algunos niños con sordera/hipoacusia son capaces de tomar turnos, solicitar aclaraciones, iniciar temas y responder a preguntas, pero no amplían los temas, retoman las conversaciones cuando existe una interrupción ni proporcionan respuestas que tengan en cuenta las contribuciones del interlocutor. Estos hallazgos sugieren que los retrasos en el lenguaje pragmático de muchos niños con sordera/hipoacusia pueden dar lugar a dificultades sociales a la hora de entablar amistad con sus compañeros y participar en las actividades del aula.

Los investigadores defienden la necesidad de proporcionar a los niños pequeños con sordera/hipoacusia oportunidades abundantes, propicias y auténticas de participar en interacciones significativas con una variedad de interlocutores durante las actividades cotidianas (Paatsch et al., 2017; Paatsch y Toe, 2020). Estas oportunidades proporcionan entornos propicios para que los niños con sordera/hipoacusia colaboren con los demás, tomen turnos, comprendan y utilicen las señales verbales y no verbales apropiadas y necesarias para el desarrollo del lenguaje, adopten la perspectiva de otras personas, reconozcan las contribuciones de los demás durante las interacciones y faciliten respuestas a sus interlocutores. Sostenemos que un contexto crítico y natural para apoyar las capacidades lingüísticas de los niños con sordera/hipoacusia es el juego, especialmente el juego de simulación.

JUEGO DE SIMULACIÓN

La definición del juego, tan difícil como compleja, ha suscitado un gran debate e interés entre investigadores, filósofos, maestros, legisladores y progenitores durante siglos. No obstante, a pesar de la dificultad de aceptar una definición universal, existe un consenso creciente de que el juego es una actividad intrínsecamente motivada, divertida y social, que implica un descubrimiento gozoso y es fundamental para el aprendizaje y el desarrollo de los niños (Yogman et al., 2018; Zosh et al., 2028). Vygotsky (1967) teoriza que es a través del juego que los niños aprenden a usar su imaginación, a crear, a experimentar y a descubrir, especialmente cuando juegan con otros más competentes en estas habilidades. De esta manera, los niños participan en interacciones sociales donde se crea una zona de desarrollo próximo (ZDP), que es la diferencia entre el nivel actual de su desempeño independiente y el nivel en el que pueden aprender cuando reciben el apoyo de compañeros y adultos más competentes.

Yogman y colaboradores (2018) opinan que «el juego no es frívolo, sino que mejora la estructura y la función cerebrales, y fomenta la función ejecutiva (es decir, el proceso de aprendizaje), lo que nos permite perseguir metas e ignorar distracciones» (pág. 1). Existen numerosos tipos diferentes de juegos en los que los niños pueden participar, como los juegos de construcción (p. ej., construir una nave espacial con bloques), los juegos motores físicos o gruesos (p. ej., trepar y correr), los juegos visuales-perceptivos, los juegos con reglas (p. ej., el pillapilla) y los juegos de simulación (p. ej., el uso de símbolos en el juego). El juego de simulación, también conocido como juego simbólico, imaginativo y social, se refiere al «uso no literal de objetos, acciones o atributos... que parece desarrollarse según un programa relativamente fijo y está presente en todas las culturas» (Creaghe et al., 2021, pág. 981). El juego de simulación implica muchos comportamientos distintivos de los niños que incluyen: (1) la sustitución de objetos, en la que utilizan un objeto por otro [p. ej., un palo por una persona; un bloque por una computadora]; (2) el juego de roles, en el que pueden representar el papel de un personaje en el juego [p. ej., una persona o un animal]; (3) las secuencias de juego, en las planifican una serie de acciones en su juego que se vuelven más complejas con el tiempo; (4) los guiones de juego, en los que desarrollan historias con uno o varios problemas y resoluciones que pueden comenzar reflejando lo que hacen (p. ej., simular que se toman una bebida) para luego pasar a historias más complejas; y (5) personajes en el juego, donde utilizan muñecas y figuras para expresar emociones y mostrar una comprensión de los estados mentales de otras personas (Elkonin, 2005; Robertson et al., 2020; Stagnitti y Paatsch, 2018). En conjunto, estas habilidades de juego de simulación implican pensamiento de orden superior, creatividad, imaginación, teoría de la mente, autorregulación, autoconciencia y resolución de problemas, todas ellas habilidades fundamentales para el aprendizaje de las habilidades del siglo XXI (Yogman et al., 2018). Además, el juego de simulación crea una sensación de seguridad, lo que reduce el estrés y la ansiedad, y prepara a los niños para el aprendizaje (Hirsch-Pasek, 2024; Paatsch et al., 2023).

VÍNCULO ENTRE EL JUEGO DE SIMULACIÓN Y EL LENGUAJE

Existe un importante corpus de investigación en el que se muestra la existencia de un vínculo estrecho entre el juego de simulación y la adquisición del lenguaje, que conducirá al desarrollo posterior de la lectoescritura, que incluye la comprensión y el uso del lenguaje narrativo (Creaghe et al., 2021; Lillard et al., 2013). Tanto el lenguaje como el juego, en concreto el juego de simulación, comparten numerosas similitudes conceptuales, entre ellas la comprensión y el uso de símbolos complejos, y la dependencia de la comunicación dentro de contextos sociales (Creaghe y Kidd, 2022; Quinn et al., 2018). Además, ambas actividades dependen del apoyo y el «andamiaje» de usuarios y actores competentes del lenguaje (Creaghe y Kidd, 2022; Creaghe et al., 2021; Paatsch et al., 2023; Quinn et al., 2018).

En un estudio reciente de Stagnitti y colaboradores (2020) se investigó la relación entre las habilidades en el juego de simulación y las habilidades del lenguaje hablado de 30 niños con audición normal al comienzo de su primer año de escolarización formal. Los resultados muestran que el hecho de ser capaces de sustituir objetos por otra cosa, en concreto utilizar muchos objetos para crear un objeto nuevo o utilizar cualquier objeto para cualquier cosa en el juego, predecía las capacidades generales del lenguaje receptivo y expresivo de los niños. En un estudio realizado por Kizildere et al. (2020) se observaron vínculos similares entre el juego de simulación y las habilidades lingüísticas generales, y se descubrió que las habilidades en el juego de simulación de 119 niños a los 4 años predecían sus habilidades de lenguaje receptivo a los 5 años (en la etapa de preescolar). En otros estudios de investigación realizados con niños con audición normal también se observa el vínculo estrecho entre el juego de simulación y habilidades lingüísticas específicas como el desarrollo semántico, incluyendo el vocabulario, la resolución de problemas semánticos y el uso de palabras de estado mental (Creaghe et al., 2021; Melzer y Palermo, 2016); la sintaxis, incluido el conocimiento gramatical (Creaghe et al., 2021; Stagnitti et al., 2016); y el desarrollo pragmático, incluyendo el desarrollo narrativo, la toma de turnos y la interacción social (Creaghe et al., 2021; Fekonja et al., 2005; Holmes et al., 2022).

También se han notificado relaciones similares entre el juego de simulación y las habilidades lingüísticas en niños con sordera/hipoacusia. Spencer (1996) investigó la asociación entre el juego de simulación (simbólico) y el lenguaje expresivo en tres grupos de niños de 2 años: (1) niños con sordera y padres con audición normal; (2) niños con sordera y padres con sordera; y (3) niños con audición normal y padres con audición normal. Los resultados mostraron un patrón sistemático de asociación entre los niveles del lenguaje y el juego simbólico, pero no entre el juego y el estado de audición, lo que confirma el vínculo estrecho entre el lenguaje expresivo y el juego. Brown y colaboradores (2001) investigaron la relación entre el juego de simulación y la producción de palabras en 10 niños con audición normal y 10 niños con sordera/hipoacusia a la edad de 28, 29 y 30 meses. Los resultados mostraron una asociación entre la producción de palabras y los niveles de juego de simulación en ambos grupos, con niveles más bajos de lenguaje y habilidades de juego evidentes en el grupo de niños con sordera/hipoacusia. Conclusiones similares de retraso en el juego de simulación y el aprendizaje de palabras se notificaron en un estudio posterior de Quittner et al. (2016). Específicamente, los niños con implantes cocleares que recibieron los implantes después de los 2 años mostraban un retraso en el logro del juego de simulación y el aprendizaje de sustantivos nuevos en comparación con otros niños con audición normal de la misma edad. En un estudio reciente de Da Silva et al. (2022) en el que se investigaron las interacciones sociales de 12 preescolares con sordera/hipoacusia durante el recreo, los resultados mostraron que estos niños pasaban menos tiempo en interacciones sociales en comparación con sus compañeros con audición normal. En concreto, estos niños evitaban los juegos de simulación y mostraban una elevada prevalencia de quedarse mirando durante el recreo, lo que sugiere que pueden experimentar aislamiento social y dificultades para entablar amistades. Este vínculo entre el desarrollo sociocognitivo, el uso social del lenguaje y las habilidades del juego de simulación también se comparó en un grupo de 82 niños con sordera/hipoacusia y 91 compañeros con audición normal, con edades comprendidas entre los 3 y los 8 años (Sidera et al., 2019). Los resultados mostraron que el grupo de niños con sordera/hipoacusia tenía dificultades para comprender las emociones fingidas, lo que estaba estrechamente relacionado con sus habilidades pragmáticas y de vocabulario expresivo.

En conjunto, estos hallazgos destacan la importancia del juego de simulación como un contexto fértil para apoyar el desarrollo lingüístico y comunicativo de los niños pequeños, ya que participan en amplias interacciones significativas con los demás. No obstante, las implicaciones de estos hallazgos enfatizan la necesidad de que los maestros, los profesores de sordos y los profesionales de la salud relacionados comprendan la importancia del juego de simulación y las formas en que pueden ayudar a los niños a desarrollar habilidades de juego que fomenten el aprendizaje del lenguaje. En las siguientes secciones se presenta un ejemplo de cómo un programa de aprendizaje profesional adaptado ayudó al personal de una escuela para niños pequeños con sordera/hipoacusia a adquirir conocimientos, aplicar un enfoque basado en el juego y cambiar la práctica.

INCORPORACIÓN DE UN ENFOQUE BASADO EN EL JUEGO PARA APOYAR EL DESARROLLO DEL LENGUAJE DE LOS NIÑOS PEQUEÑOS CON SORDERA/HIPOACUSIA

Antecedentes

Este proyecto de investigación de tres años, *Teachers Deliberating Practice (Maestros deliberando sobre la práctica)*, lo realizaron investigadores de la Deakin University, Australia, (Nolan, Paatsch y Robertson) en colaboración con el personal de una escuela para niños con sordera/hipoacusia con edades comprendidas entre el nacimiento y los 7 años. En concreto, la dirección del centro se puso en contacto con los investigadores para ayudarles a integrar en su centro un enfoque de la enseñanza y el aprendizaje basado en el juego como contexto para apoyar el desarrollo de la comunicación y el lenguaje de los niños con sordera/hipoacusia.

Objetivos del proyecto de investigación

El objetivo de este proyecto era establecer una cultura de investigación en la escuela que apoyara la adquisición de conocimientos y condujera a un cambio de prácticas en relación con un enfoque lúdico de la enseñanza y el aprendizaje. En concreto, el proyecto se centró en apoyar a los maestros, los profesores de sordos, los educadores, el personal de apoyo y los profesionales de la salud asociados para que adquirieran conocimientos sobre el juego de simulación y el lenguaje, ya que habían incorporado un enfoque basado en el juego en toda la escuela. Además, en el proyecto se aplicó el enfoque de Investigación-acción participativa (IAP; Kemmis y McTaggart, 2005) para ayudar a los maestros a investigar su propia práctica y documentar cualquier cambio en la misma como resultado de la aplicación de un enfoque basado en el juego.

Contexto del estudio

La escuela ofrece programas para niños con sordera/hipoacusia y sordociegos desde el diagnóstico hasta los 7 años en todo el estado de Victoria, Australia. Los programas incluyen intervención temprana, educación temprana que incluye jardín de infancia y primer año de escolarización, y un jardín de infancia comunitario inclusivo de 3-4 años para niños con sordera/hipoacusia y audición normal. El personal de la escuela incluye maestros, educadores, profesores de sordos, personal de apoyo y profesionales sanitarios asociados, como fonoaudiólogos, trabajadores sociales y fisioterapeutas.

El programa

El proyecto de investigación constaba de tres fases: (1) comprensión y adquisición de conocimientos; (2) práctica de investigación; y (3) adquisición de capacidades para un programa sostenible.

La primera fase se centró en la adquisición de conocimientos por parte del personal en las áreas de juego y lenguaje. En concreto, el primer taller, en el que participaron 60 miembros del personal, incluyó la adquisición de conocimientos sobre los tipos y principios del juego, centrándose en el juego de simulación, la relación entre el juego y el aprendizaje, los componentes del lenguaje y la interacción entre el lenguaje y el juego. El segundo taller consistió en la adquisición de conocimientos por parte del personal sobre su papel en el apoyo al juego y el lenguaje de los niños, la reflexión sobre sus propios patrones de habla que facilitan el aprendizaje del lenguaje y los principios de la aplicación de un enfoque basado en el juego.

En la segunda fase del proyecto de investigación participaron 30 miembros del personal en tres talleres online y dos talleres presenciales de un día de duración. Los objetivos de esta fase eran dar a conocer al personal la evaluación del juego y el proceso de cuatro pasos del enfoque de IAP (observar, reflexionar, planificar y actuar), así como trabajar con ellos para diseñar un proyecto de investigación en torno a un estudio de caso de un niño de su clase. Se incluyó el desarrollo de una pregunta de investigación para explorar su propia práctica en el apoyo al lenguaje de los niños con un enfoque basado en el juego. La exploración incluyó la realización de evaluaciones, la grabación en video de sus prácticas, la recopilación de pruebas, la reflexión y la planificación de la implementación. El personal completó tres ciclos del proceso de cuatro pasos del enfoque de IAP como parte de su propio proyecto de investigación. Al final del tercer ciclo, el personal participó en una «Muestra de prácticas» en la que cada miembro del personal presentó al resto del personal los resultados de su IAP.

La tercera fase del proyecto incluyó cuatro talleres presenciales de un día de duración en la escuela para desarrollar la capacidad de sostenibilidad del programa en el futuro. El personal y los investigadores elaboraron un documento de trabajo que incorporaba el contenido y los recursos introducidos y desarrollados durante las dos primeras fases. En concreto, se elaboró conjuntamente un documento titulado «Juego y práctica: conectando el juego y el lenguaje» para la escuela, que incluía pruebas de investigación y documentos de planificación y evaluación del profesorado específicos para el juego y el lenguaje, así como formas de apoyar un enfoque basado en el juego en la escuela. Otras secciones del documento incluían consideraciones clave para la implementación del IAP y formas de trabajar con colegas en el marco de un enfoque de práctica transdisciplinar.

Los resultados

A lo largo de las tres fases del proyecto de investigación, el personal debatía constantemente su creciente comprensión del juego y el lenguaje. Al final de la segunda fase, se invitó al personal a cumplimentar una encuesta online con el fin de compartir sus aprendizajes como resultado del proyecto. Los resultados generales mostraron que, si bien «había mucho que aprender de todos estos talleres», el personal había adquirido una mayor comprensión de las formas en que se pueden apoyar las capacidades de juego y lenguaje de los niños con sordera/hipoacusia a través de un enfoque basado en el juego. En concreto, el personal señaló que «el juego es una parte integral del desarrollo de las habilidades comunicativas» y que «las oportunidades de juego están en todas partes». El personal también destacó los numerosos elementos de la fase dos que apoyaron su aprendizaje, incluyendo el «nuevo conocimiento y su puesta en práctica», «el proceso de análisis de videos y la redefinición de la pregunta de investigación», «disponer de tiempo para ver videos, debatir y planificar con colegas», y «que el ciclo de investigación-acción me permite reflexionar críticamente sobre mi práctica y, a continuación, implementar un plan que sea específico para mis alumnos».

El personal también comentó el valor de la «Muestra de prácticas». Un miembro del personal comentó que «me encantó conocer las perspectivas de mis compañeros sobre cómo es el juego en sus espacios individuales y cómo la reflexión continua es clave para mejorar nuestros programas y apoyar a nuestros alumnos». Otros miembros del personal comentaron que las presentaciones les ayudaron a «articular lo que entendíamos», a «consolidar el aprendizaje» y a «apreciar el desarrollo de los alumnos en el juego y el lenguaje», mientras que otros afirmaron que escuchar a los colegas fue «interesantísimo», ya que «aprendimos mucho» a partir de «perspectivas diferentes».

También se invitó al personal a reflexionar sobre las diferencias que habían introducido en su práctica durante su participación en la fase 2. El personal apreció la importancia de crear un entorno de juego auténtico para animar a los niños a utilizar el lenguaje con sus compañeros y maestros, y la necesidad de «añadir el juego como parte de sus objetivos, no solo el lenguaje». Algunos también cambiaron sus prácticas para implicarse más con los niños. Como informó un miembro del personal: «reflexioné más sobre lo que estaba pasando, qué papel estaba desempeñando y cómo podía modificarlo para ayudar a los niños». Otro maestro comentó:

> «Era preciso que mi nivel de implicación en el juego de una alumna aumentase, ya que descubrí, al ver los videos de su juego, que necesitaba más modelado para ampliar su juego más allá de las escasas acciones repetitivas que conocía para un escenario determinado. De esta manera, en lugar de limitarme a preparar experiencias para que jugara con sus compañeros, me unía a ella en el juego y el modelado».

CONCLUSIÓN

En este documento se ha presentado la investigación relativa a las características clave del juego de simulación y la relación entre el juego de simulación y el lenguaje. Los hallazgos de la investigación también demuestran que muchos niños con sordera/hipoacusia a menudo presentan retrasos en sus habilidades de juego de simulación y lenguaje, y un gran número de investigadores destaca la necesidad de facilitar desarrollo profesional a las personas que trabajan con estos niños para apoyar estas habilidades. El presente proyecto de investigación que se describe en este documento pone de relieve un ejemplo de cómo los investigadores y el personal de una escuela de Victoria, Australia, trabajaron conjuntamente para adquirir conocimientos sobre el juego y el lenguaje, e implementarlos en la práctica. Los educadores, los profesores de sordos, el personal de apoyo educativo y los profesionales de la salud asociados valoraron el trabajo en equipo transdisciplinar para adquirir sus conocimientos, modificar la práctica actual y desarrollar un programa sostenible para todo el personal que trabajaba en la escuela. Este personal puede marcar la diferencia en el aprendizaje de los niños dedicando tiempo a adquirir conocimientos sobre las capacidades de los niños en el juego de simulación y las formas en que un enfoque basado en el juego proporciona un contexto propicio para el aprendizaje del lenguaje, y a reflexionar sobre sus propias prácticas desde una perspectiva basada en la evidencia. No obstante, es necesario seguir investigando los resultados específicos del juego de simulación y del lenguaje para los niños con sordera/hipoacusia y la relación entre estos resultados y la práctica de los maestros a la hora de implementar un enfoque basado en el juego.

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IMPULSANDO EL DESARROLLO DEL LENGUAJE HABLADO A TRAVÉS DEL ACCESO AUDITIVO

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Los niños solo pueden desarrollar el lenguaje hablado si están expuestos a las señales acústicas que componen el habla y el lenguaje de una manera constante. Hasta hace poco, los niveles de audición obtenidos a través de un audiograma clínico eran la medida principal que se utilizaba para definir la audición normal, así como la presencia o el grado de pérdida auditiva de un niño. Si bien el audiograma clínico sigue siendo una medida diagnóstica importante para los audiólogos, el grado de pérdida auditiva del audiograma es un indicador poco fiable de la capacidad auditiva funcional de un niño, especialmente una vez que utiliza audífonos o implantes cocleares. La audibilidad del habla es una estimación del acceso de un niño a las señales acústicas que componen el lenguaje hablado y se utiliza actualmente para cuantificar con precisión los niveles de audición funcional de un niño con y sin dispositivos. En este artículo se analizan las limitaciones de la cuantificación del acceso auditivo basada en el audiograma clínico, así como los enfoques que existen para estimar la audibilidad del habla en el caso de los niños con pérdida auditiva.

El audiograma es una medida clínica de la sensibilidad auditiva basada en las respuestas conductuales de un niño a estímulos de banda estrecha o de tonos puros en un rango de frecuencias que es el más importante para la comprensión del habla. El audiograma también se puede estimar en base a las respuestas neuronales a estímulos de ráfaga de tonos o chirridos a partir de los potenciales evocados auditivos del tronco cerebral o de los potenciales evocados auditivos de estado estable en bebés o niños que no puedan realizar una prueba de audición conductual. El audiograma se puede dividir en rangos que cuantifican la cantidad o el grado de la pérdida auditiva. En la Figura 1 se muestra el audiograma de un niño con una pérdida auditiva leve en ambos oídos. El audiograma se suele utilizar para realizar un diagnóstico médico de la pérdida auditiva, aunque tiene algunas limitaciones funcionales, sobre todo para medir el impacto de la pérdida auditiva en la comunicación.

https://doi.org/10.17955/tvr.124.1.symp3

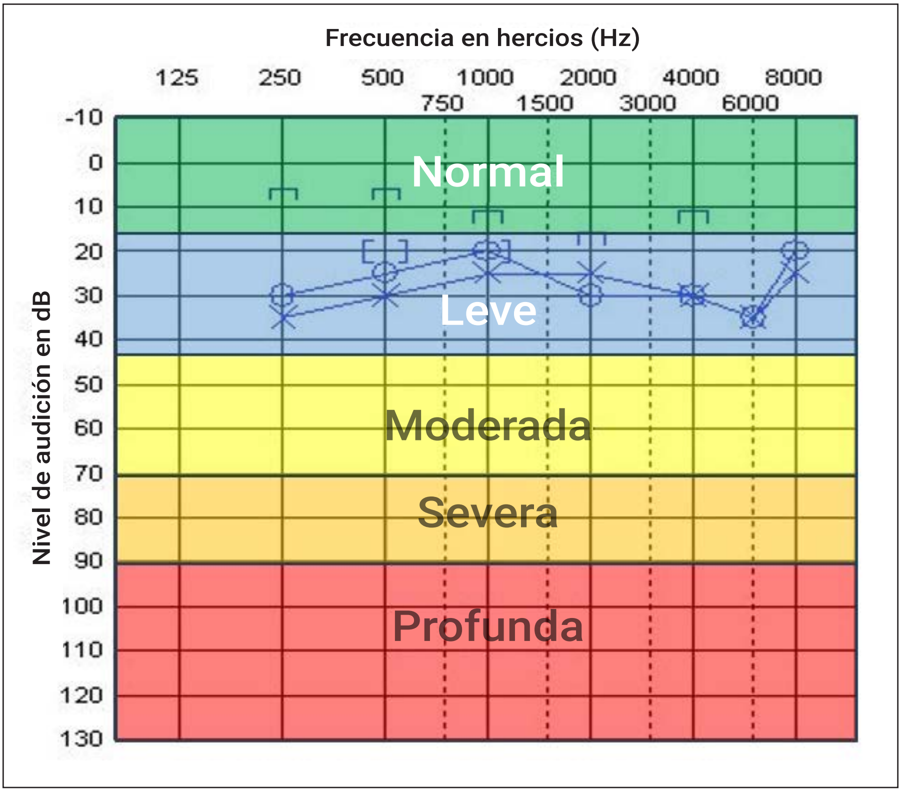

Figura 1. Audiograma de un niño con una pérdida auditiva leve en ambos oídos. Los círculos corresponden a los umbrales de audición del oído derecho y los triángulos a los del oído izquierdo. Los corchetes son los umbrales de conducción ósea. Las áreas sombreadas representan diferentes grados de pérdida auditiva: normal = verde; pérdida auditiva leve = azul; pérdida auditiva moderada = amarillo; pérdida auditiva severa = naranja; y pérdida auditiva profunda = rojo.

Si bien el audiograma mide los niveles de audición relacionados con las frecuencias de sonido discretas que son más importantes para comprender el habla, los estudios de investigación señalan que el grado de pérdida auditiva en el audiograma no es un predictor fiable del grado en que un niño con pérdida auditiva desarrollará el lenguaje hablado o rendirá académicamente en la escuela. En varios estudios se sugiere que los niños con grados leves de pérdida auditiva pueden tener un mayor riesgo de presentar un retraso en el lenguaje hablado (Walker et al., 2020) y dificultades académicas (Tomblin et al., 2020) que los niños con grados de pérdida auditiva moderados o mayores. Es mucho más probable que se pueda predecir el progreso del desarrollo de un niño con pérdida auditiva a través del servicio de intervención que reciba que a partir del grado de pérdida auditiva obtenido de un audiograma. Por ejemplo, Walker y colaboradores (2015) descubrieron que los niños con pérdida auditiva bilateral leve que utilizaban audífonos de una manera sistemática presentaban unos resultados lingüísticos mucho mejores que los niños con pérdida auditiva bilateral leve que no los utilizaban. Del mismo modo, en el caso de los niños que utilizaban audífonos adaptados para optimizar su audibilidad del habla se apreció un mayor desarrollo del lenguaje (Tomblin et al., 2015) y de las habilidades de la función ejecutiva (McCreery y Walker, 2022), así como mejores resultados en matemáticas y lectura (Tomblin et al., 2020), en comparación con los niños con audífonos no adaptados. El grado de pérdida auditiva del audiograma no refleja estas diferencias en la intervención en caso de niños con pérdida auditiva.

Basándose en estos estudios de investigación, entre otros, nuestro equipo de investigación ha desarrollado nuevos métodos para cuantificar el acceso auditivo, es decir, métricas más útiles para los padres y los médicos que prestan servicio a los niños con pérdida auditiva y que muestran una mayor eficacia para predecir el riesgo y la resiliencia de los niños que utilizan audífonos.

AUDIBILIDAD DEL HABLA NO ASISTIDA

La audibilidad del habla no asistida es una medida que representa la proporción de energía del habla que un niño puede oír sin un audífono en una situación de escucha específica. La situación de escucha más habitual que utilizamos para estimar la audibilidad es el habla a un nivel conversacional medio cuando el interlocutor se encuentra a un metro de distancia frente al oyente, que es de aproximadamente 60 dB SPL. Se pueden utilizar los niveles de umbrales auditivos del audiograma para estimar la proporción de habla a un nivel conversacional medio que un niño puede oír con sus umbrales auditivos. En la Figura 2 se muestra el espectro del habla no asistida trazado con el audiograma del oído derecho del niño con pérdida auditiva leve-moderada de la Figura 1.

Se puede medir la cantidad de espectro del habla que se encuentre por encima de los umbrales del niño convirtiendo los umbrales del audiograma en dB SPL para poder compararlos directamente con el habla mediante un gráfico denominado SPL-o-grama (Figura 2). El SPL-o-grama nos permite visualizar qué parte de la señal del habla es

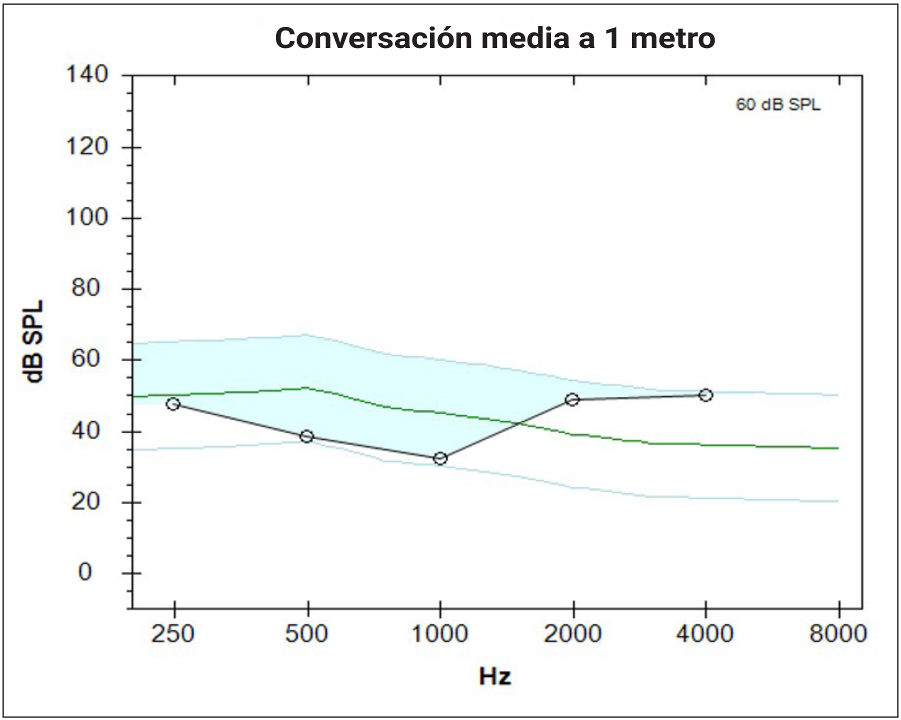

Figura 2. SPL-o-grama del oído derecho del niño con pérdida auditiva leve de la Figura 1. Téngase en cuenta que el eje y muestra la audición normal en la parte inferior y niveles sonoros crecientes a medida que aumenta el eje. Las frecuencias del audiograma permanecen en el eje x. Los círculos que conectan con la línea negra continua son los umbrales de audición del oído derecho convertidos a dB SPL. La línea verde representa el nivel medio del habla a 60 dB SPL a través de la frecuencia, y la línea azul representa el rango de niveles del habla. El área sombreada en azul representa la parte del espectro del habla que es audible en el oído derecho sin audífono.

audible en el oído derecho para una señal del habla media sin audífono (no asistida). También se puede aplicar un cálculo conocido como Índice de inteligibilidad del habla (IIH; ANSI S3.5-2007) que permite cuantificar numéricamente la proporción ponderada del espectro del habla que es audible o que se encuentra por encima de los umbrales del niño. En el ejemplo de la Figura 2, el IIH no asistido del oído derecho es de 0,47, lo que significa que, en el caso de un nivel conversacional medio en una situación de escucha tranquila, el niño puede oír aproximadamente el 47 % de las señales del lenguaje hablado. Visualmente, la mayoría de las señales del habla que son audibles en este ejemplo se encuentran en frecuencias inferiores a 2000 Hz. Los padres de niños con pérdida auditiva a menudo se sorprenden al saber que una pérdida auditiva leve significa que menos de la mitad del espectro del habla es audible en silencio. En un estudio realizado por Sapp y colaboradores (2023) se comparó el nivel de preocupación de los padres basado en las categorías descriptivas tradicionales de la pérdida auditiva (p. ej. leve, moderada, severa y profunda) frente a las descripciones de la pérdida auditiva basadas en la audibilidad y en simulaciones de pérdida auditiva. Los padres consideraron que las simulaciones de pérdida auditiva y las descripciones de la pérdida auditiva basadas en la audibilidad transmitían con mayor precisión el impacto funcional de la pérdida auditiva que el uso de términos como «leve» para describir el problema. Las medidas de audibilidad no asistida pueden ser herramientas de asesoramiento útiles para los padres de niños con pérdida auditiva u otros profesionales que puedan participar en el apoyo médico o educativo del niño.

AUDIBILIDAD DEL HABLA ASISTIDA

La audibilidad también se puede calcular con audífonos basándose en las mediciones de la salida de los audífonos en el conducto auditivo del niño o en un acoplador con los factores de corrección apropiados, lo que se conoce como audibilidad asistida. La audibilidad asistida se basa en el mismo cálculo del IIH que la audibilidad no asistida, pero las medidas de la salida de los audífonos se comparan con los umbrales auditivos del niño, en lugar de con un espectro de habla no asistida estándar. En la Figura 3 se muestra el ejemplo de un SPL-o-grama con audibilidad asistida. En este ejemplo, el espectro del habla amplificado se compara con los umbrales del oído izquierdo del niño en el audiograma. Sin amplificación, el niño puede oír el 2 % del espectro del habla (IIH asistido) y con el audífono puede oír el 73 % del espectro medio del habla (asistida) basándose en el IIH. Como se ha indicado anteriormente, el IIH asistido es una herramienta útil para estimar cuál es el acceso al habla que tiene un niño a través de sus audífonos. Actualmente no existe ningún método estandarizado para calcular la audibilidad del habla de los niños que utilizan implantes cocleares, dado que no hay forma de medir directamente cuánto del espectro del habla es accesible a través del implante coclear.

Una de las limitaciones del IIH asistido es que la cantidad de

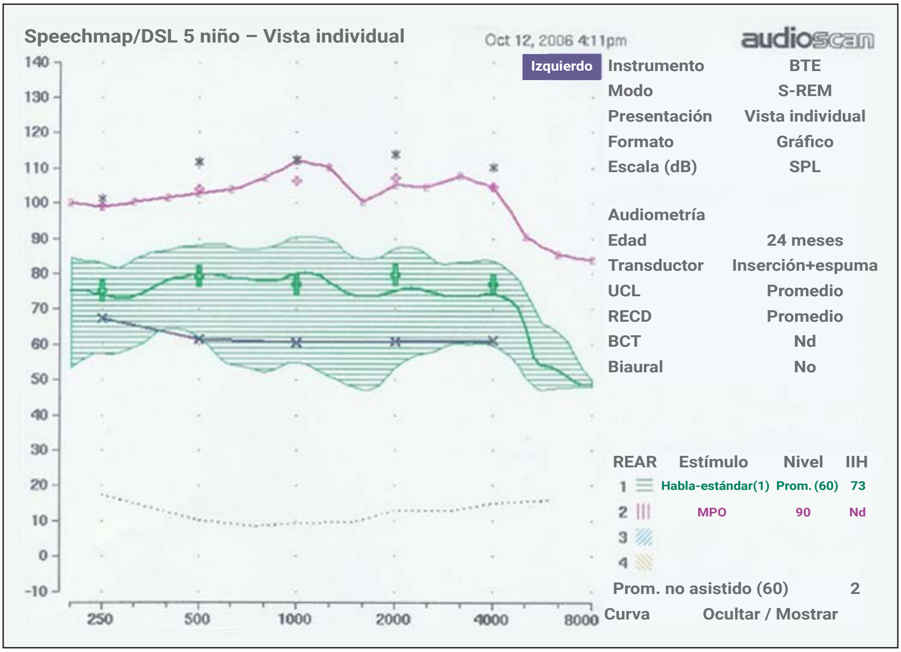

Figura 3. SPL-o-grama que muestra la audibilidad asistida. Los símbolos X conectados con la línea azul representan los umbrales de audición del niño a partir del audiograma del oído izquierdo. La zona verde sombreada es el espectro del habla medido a través del audífono del niño. Los valores de IIH asistido (73) y no asistido (2) correspondientes al habla media (60 dB SPL) se muestran en la parte inferior derecha del gráfico. Significado de las siglas: BCT = umbral de conducción ósea; MPO = salida pico máxima; Nd = no disponible; REAR = respuesta asistida de oído real; RECD = diferencia entre las medidas en oído real y acoplador; SPL = nivel de presión sonora; y UCL = nivel incómodo.

audibilidad del habla que se puede recuperar a través de un audífono disminuye a medida que aumenta el grado de pérdida auditiva del niño (McCreery et al., 2013). Esto significa que no existe un valor óptimo de IIH que se puede alcanzar con niños que tienen diferentes grados de pérdida auditiva. El Protocolo de amplificación pediátrica de Western Ontario ha publicado valores normativos del IIH asistido en función del grado de pérdida auditiva, que están disponibles para los clínicos y se encuentran integrados en numerosos sistemas de verificación de audífonos (Bagatto et al., 2016). En un estudio reciente de Wiseman y colaboradores (2023) se demostró que los niños con pérdida auditiva que utilizaban audífonos y que tenían un IIH asistido inferior al 50 % corrían el riesgo de sufrir retrasos en el lenguaje en comparación con los niños con una audición normal, incluso si utilizaban audífonos de forma sistemática. Este trabajo sugiere que un IIH asistido en silencio, siendo audible menos de la mitad del espectro del habla asistida, puede no proporcionar la audibilidad adecuada para apoyar el desarrollo del lenguaje hablado y puede requerir modificaciones en los servicios de intervención del niño, incluyendo un audífono más potente o la derivación para la evaluación del implante coclear si no se puede conseguir una mayor audibilidad mediante la audición acústica.

El audiograma es una medida importante para el diagnóstico y la cuantificación de la pérdida auditiva. Sin embargo, se sabe que el audiograma no refleja los retos auditivos funcionales que los niños con pérdida auditiva experimentan en su vida diaria. La audibilidad del habla, medida por el IIH, es una forma de utilizar el audiograma junto con espectros del habla amplificados o no amplificados para estimar la proporción del habla que será audible para los niños en diferentes situaciones auditivas. Los médicos pueden utilizar las mediciones de la audibilidad para asesorar a los padres y a los cuidadores de niños con pérdida auditiva que utilizan audífonos, con el fin de ayudarles a que comprendan mejor cómo puede afectar la pérdida auditiva a la comunicación. Los padres de un niño con pérdida auditiva pueden preguntar al audiólogo que le trate o a otros profesionales sanitarios sobre la audibilidad del habla para poder comprender en qué medida los audífonos pueden facilitar su acceso a la comunicación.

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INFECCIÓN CONGÉNITA POR CITOMEGALOVIRUS (CMV): UNA CAUSA FRECUENTE PERO IGNORADA DE PÉRDIDA AUDITIVA Y TRASTORNOS DEL NEURODESARROLLO

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La infección por citomegalovirus congénita (CMVc) es una causa prevalente pero poco reconocida de pérdida auditiva neurosensorial y trastornos del neurodesarrollo en niños. Si bien la infección por CMV suele ser asintomática en personas sanas, su transmisión durante el embarazo puede tener consecuencias importantes y duraderas para la salud del feto. A pesar de su impacto mundial, la concienciación acerca del CMVc sigue siendo escasa, especialmente entre las mujeres embarazadas y los profesionales sanitarios, lo que contribuye a un infradiagnóstico y al tratamiento inadecuado de los lactantes afectados. En este manuscrito se revisan la epidemiología, las manifestaciones clínicas y los resultados a largo plazo de la infección por CMVc, haciendo hincapié en la necesidad crítica de que existan medidas preventivas, entre ellas un mayor grado de concienciación y formación. En el documento también se analiza el papel de los programas de cribado, destacando los beneficios potenciales de los enfoques universales o específicos en la detección y la intervención tempranas. A pesar de los avances en la comprensión del CMVc, sigue habiendo importantes lagunas en el conocimiento de sus efectos cognitivos a largo plazo y la eficacia de las distintas intervenciones. Se necesitan más

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estudios de investigación en los que se exploren las experiencias de las personas con CMVc y de sus cuidadores, se identifiquen las lagunas en la atención sanitaria y se desarrollen estrategias de intervención integrales y centradas en la familia.

¿QUÉ ES LA INFECCIÓN POR CMVC?

El CMV es un herpesvirus que se transmite a través de los fluidos corporales, estableciéndose una latencia de por vida tras la infección inicial, lo que significa que el virus se puede reactivar en cualquier etapa posterior de la vida y que las personas también se pueden reinfectar con diferentes cepas de CMV. En la mayoría de las personas, la infección es asintomática o solo provoca síntomas leves similares a los de la gripe y fiebre, sin problemas de salud graves.

Sin embargo, en el caso de personas con un sistema inmunitario debilitado, como las infectadas por el VIH o que hayan recibido un trasplante recientemente, el CMV puede provocar una enfermedad grave (Taylor, 2003). La infección por CMV congénita (CMVc) se produce cuando el virus se transmite de una mujer embarazada al feto a través de la placenta, un proceso conocido como transmisión vertical. La mayoría de las mujeres embarazadas adquieren la infección a través del contacto con la saliva de niños pequeños que eliminan el CMV más fácilmente que los adultos (Dollard et al., 2014). Por ejemplo, compartir utensilios o besar a un niño pequeño en la boca puede dar lugar a una transmisión del CMV. Para prevenir la infección por CMV, se aconseja a las mujeres embarazadas que practiquen unas buenas rutinas de higiene, como lavarse siempre las manos después de cambiar un pañal o limpiar la nariz a un niño pequeño, y no compartir utensilios, alimentos ni bebidas con otras personas (Fowler & Boppana, 2018).

El riesgo de transmisión es significativamente mayor si la madre contrae una infección primaria por CMV durante el embarazo (32 %) en comparación con una infección recurrente (1,4 %) (Kenneson & Cannon, 2007). Con una prevalencia media del 0,5 %, el CMVc es la infección por virus congénita más frecuente en los países desarrollados (Orb et al., 2024; Ssentongo et al., 2021). En regiones en desarrollo de África, Asia y Latinoamérica, las tasas de prevalencia oscilan entre el 0,6 % y el 6,1 % (Kenneson & Cannon, 2007).

En una investigación de Fowler y colegas (2018) se destaca el impacto de la raza y el origen étnico en la prevalencia del CMVc. Descubrieron que los bebés de raza negra o con un origen multirracial podrían tener un riesgo significativamente mayor de contraer el CMVc. Estos hallazgos subrayan la necesidad de implementar estrategias preventivas destinadas a incrementar la concienciación sobre el CMVc en la población de mujeres en edad fértil y el reconocimiento por parte de los profesionales sanitarios de las diferencias grupales relacionadas con la raza y el origen (Fowler et al., 2018). Cabe destacar que no se encontraron diferencias significativas entre razas o etnias en cuanto a la probabilidad de que los niños presenten una infección sintomática por CMVc o una pérdida auditiva asociada.

La seroprevalencia se refiere al porcentaje de adultos o niños mayores en una población que presentan anticuerpos detectables contra un patógeno específico, como el CMV, en la sangre. Esta medida aumenta con la edad, e indica lo extendido que está el CMV en una comunidad o una población en un momento dado (Fowler et al., 2022). En las regiones con una mayor seroprevalencia suele haber tasas más elevadas de infección por CMVc, si bien el riesgo de infectar a los fetos es mayor en los casos de infección primaria (De Vries et al., 2013).

Aproximadamente 1 de cada 5 bebés que nace con una infección por CMVc puede presentar defectos congénitos o problemas de salud a largo plazo. La infección congénita por CMV también se ha asociado a muerte fetal y mortinatos en algunos estudios (Griffiths et al., 1980; Iwasenko et al., 2011). Los niños que nacen con una infección por CMVc se suelen clasificar en dos grupos: los que presentan un CMVc sintomático (10 %) o un CMVc asintomático (90 %) (Kenneson & Cannon, 2007).

La infección sintomática por CMVc se refiere a los casos en los que el individuo presenta manifestaciones clínicas de la infección al nacer. En los casos congénitos, entre los síntomas se incluyen un agrandamiento del hígado, ictericia, erupción cutánea, pérdida auditiva neurosensorial, problemas de visión y problemas neurológicos como microcefalia, convulsiones o retrasos en el desarrollo (Naing et al., 2016). Estos síntomas se suelen detectar al nacer o poco después y pueden provocar graves problemas de salud a largo plazo (Lanzieri et al., 2017).

La infección por CMVc asintomática se refiere a los casos en los que el individuo infectado no muestra síntomas ni signos observables de la enfermedad en los primeros años de vida (Stoyell et al., 2024) o en la infancia tardía (Lopez et al., 2017; Novelli et al., 2022). Alrededor del 90 % de los niños con CMVc son asintomáticos durante el periodo neonatal (Gantt et al., 2017). Aunque no presenten manifestaciones clínicas en el momento del diagnóstico, pueden ser portadores del virus y desarrollar una pérdida auditiva de aparición tardía. Alrededor del 10-15 % de los lactantes con CMVc asintomáticos desarrollarán una pérdida auditiva neurosensorial permanente (Fowler & Boppana, 2018). Además, si bien se pueden presentar alteraciones visuales y cognitivas, su porcentaje es menor en comparación con las observadas en casos de CMVc sintomáticos (Dollard et al., 2007).

IMPACTO DE LA INFECCIÓN POR CMVC

La infección congénita por CMV es una de las principales causas de pérdida auditiva neurosensorial unilateral o bilateral adquirida en niños, que puede ser de leve a profunda (Goderis et al., 2014). Esta pérdida auditiva puede estar presente al nacer, progresar con el paso del tiempo o desarrollarse como una afección de aparición tardía durante la infancia. Debido a la posibilidad de que la pérdida auditiva sea progresiva o tardía, es fundamental realizar un seguimiento auditivo continuo. Las intervenciones a su debido tiempo son esenciales para favorecer el desarrollo del lenguaje y la comunicación, garantizando que los niños afectados reciban los recursos y cuidados necesarios.

Los implantes cocleares se utilizan en el caso de los niños que presentan CMVc y sordera bilateral, pero también son beneficiosos en los casos de sordera unilateral (Aldè et al., 2023). El implante coclear unilateral en niños con CMVc y sordera unilateral puede mejorar significativamente su capacidad de localizar sonidos y comprender el habla en entornos ruidosos. Además, las personas con CMVc corren un riesgo considerable de perder audición en el oído no afectado con el paso del tiempo. Cuando los niños con sordera unilateral reciben un implante coclear temprano, se preserva la mayor capacidad auditiva posible, y se refuerza el desarrollo auditivo y del habla en general (Aldè et al., 2023).

La pérdida auditiva es un trastorno prominente del neurodesarrollo resultante de la infección por CMVc (Goderis et al., 2014), pero no explica en su totalidad el rango de dificultades cognitivas que se observa en niños con infección por CMVc (Karltorp et al., 2014). Los niños con pérdida auditiva e infección por CMVc corren el riesgo de sufrir diversos trastornos cognitivos, incluido un peor funcionamiento ejecutivo (Löfkvist et al., 2020). En un estudio de Löfkvist y colegas (2020) se evaluaron las habilidades pragmáticas mediante cuestionarios realizados a los progenitores, llegando a la conclusión de que los niños con CMVc tenían unas habilidades pragmáticas y una memoria de trabajo fonológica significativamente peores en comparación con sujetos de control bien emparejados con una causa de sordera genética. Estas dificultades cognitivas pueden, a su vez, repercutir negativamente en el rendimiento académico y en las interacciones sociales (Löfkvist et al., 2020).

Además, el CMVc también se asocia con problemas vestibulares (Dhondt et al., 2021; Karltorp et al., 2014; Kokkola et al., 2023; Shears et al., 2022). En una revisión sistemática, Shears et al. (2022) demostraron que incluso los niños con infección por CMVc asintomática, incluidos aquellos con una audición normal, pueden experimentar una disfunción vestibular, que puede ser progresiva. Karltorp et al. (2014) investigaron las comorbilidades de niños con pérdida auditiva, comparándolas con las de sujetos de control con una causa de sordera genética (conexina 26), que normalmente no incluye problemas cognitivos. Descubrieron que los niños con CMVc presentaban peor equilibrio, pérdida de visión, trastornos alimentarios imprevistos y diagnósticos neuropsiquiátricos, entre ellos el trastorno del espectro autista y el TDAH, mientras que los sujetos de control no presentaban este tipo de problemas.

En estudios anteriores también se ha demostrado que los niños con infección por CMVc pueden tener discapacidades intelectuales (Lanzieri et al., 2017) y presentar diagnósticos neuropsiquiátricos como trastorno del espectro autista, TDAH y disfunciones motoras orales (Karltorp et al., 2014). En un amplio estudio poblacional realizado por Pesch y colegas (2024) se concluye que los niños estadounidenses con CMVc tienen un riesgo 2,5 veces mayor de desarrollar un trastorno del espectro autista que los niños sin infección (Pesch et al., 2024). Las afecciones comórbidas son frecuentes en niños con CMVc, lo que contribuye a la elevada heterogeneidad dentro de esta población (Karltorp et al., 2014). Algunos niños con CMVc pueden presentar pocos síntomas o ninguno, mientras que otros experimentan múltiples afecciones graves. Es más probable que los niños con infección sintomática por CMVc presenten afecciones más graves (Lanzieri et al., 2017).

Sin embargo, en un estudio único de seguimiento a largo plazo realizado por Falkenius Schmidt y colegas (2024) se concluye que los adultos con infección por CMVc que tenían una audición normal y fueron asintomáticos al nacer experimentaban déficits en los procesos de recuperación de palabras. Esta capacidad cognitiva se evalúa mediante tareas de fluidez de palabras como la tarea FAS, que requiere que los individuos recuperen el mayor número posible de palabras que empiecen por las letras F, A y S en un minuto. Estos resultados sugieren que los efectos a largo plazo de la infección por CMVc pueden persistir incluso en individuos que inicialmente no mostraban síntomas, lo que indica posibles dificultades cognitivas sutiles que pueden no detectarse fácilmente a primera vista.

Una de las limitaciones del estudio es el reducido tamaño de la muestra (n=34), por lo que los resultados deberán repetirse en estudios de investigación posteriores. Sin embargo, un punto fuerte del estudio fue la inclusión de un grupo de control de individuos que no estaban infectados al nacer. Cuando se compararon con los datos normativos suecos, el 43 % de los participantes con infección por CMVc, todos ellos asintomáticos al nacer, tuvieron un desempeño adecuado tanto en las tareas de FAS como en las de fluidez verbal, en comparación con el 86 % de los sujetos de control (Falkenius Schmidt et al., 2024).

Korndewal y colegas (2017) investigaron cómo la vida cotidiana de los niños en edad escolar con CMVc y la de sus progenitores se ve afectada. Descubrieron que la calidad de vida tanto de los niños con deficiencias a largo plazo como la de sus progenitores era inferior a la de los sujetos de control sin infección por CMVc. Además, los padres de niños con CMVc informaron de un mayor número de problemas físicos y de concentración que los de los sujetos de control con niños sin CMVc. Estos resultados ponen de relieve que el CMVc repercute en las experiencias de la vida cotidiana tanto de los niños afectados como de sus familiares, lo que indica la necesidad de un enfoque de intervención centrado en la familia (Korndewal et al., 2017). Si bien los individuos con infección sintomática por CMVc suelen tener necesidades más complejas, es importante reconocer que aquellos con CMVc asintomático también pueden tener necesidades similares.

PREVENCIÓN

En la actualidad no se dispone de vacunas eficaces contra el CMV. El desarrollo de una vacuna de este tipo es especialmente difícil por la compleja naturaleza de la inmunidad frente al CMV, que incluye la posibilidad de reactivación de una infección previa y el riesgo de infección con nuevas cepas víricas. Si bien los estudios de investigación en curso sobre vacunas candidatas contra el CMV son prometedores (Hu et al., 2024; Schleiss, 2016), es poco probable que se disponga de una vacuna eficaz en un futuro próximo. En consecuencia, la estrategia más eficaz para prevenir la infección materna y congénita por CMV sigue siendo informar a las mujeres embarazadas de las fuentes de exposición al CMV y promover intervenciones conductuales para reducir estos riesgos (Calvert et al., 2021). Además, el aumento de la concienciación pública sobre la infección por CMVc y la promoción de prácticas higiénicas son esenciales para minimizar la transmisión del virus.

A pesar de la necesidad de una mayor concienciación pública y de la prevalencia de la infección por CMVc, muchas mujeres embarazadas siguen sin conocer el CMVc y sus posibles consecuencias graves, así como las acciones preventivas (Calvert et al., 2021). Los estudios indican importantes lagunas de conocimiento, con tasas de concienciación que oscilan entre el 13 % y el 60 % en varios países (Binda et al., 2016; Cordier et al., 2012; Pereboom et al., 2013). Las comadronas, que interactúan regularmente con las mujeres embarazadas, desempeñan un papel crucial en la difusión de información sobre el CMV y pueden ayudar a prevenir su propagación dando a conocer las medidas de protección a estas mujeres. Otros profesionales sanitarios, como médicos, pediatras, audiólogos educativos, fonoaudiólogos y especialistas en intervención temprana, también deben estar bien informados sobre el impacto de la infección por CMVc.

Rawlinson et al., (2017) crearon un Grupo internacional informal de recomendaciones sobre citomegalovirus congénito con el objetivo de facilitar directrices para la prevención, el diagnóstico y el tratamiento del CMVc. El grupo, basándose en discusiones consensuadas y en la revisión de la literatura, llegó a la conclusión de que un cribado neonatal universal de CMVc se podría considerar para facilitar la detección y la intervención tempranas en caso de pérdida auditiva y/o retraso del desarrollo. También coincidieron en que las estrategias de educación y prevención para las mujeres embarazadas son beneficiosas y que las recomendaciones se deben actualizar a medida que se disponga de nuevos datos y conocimientos (Rawlinson et al., 2017).

CRIBADO DE LA INFECCIÓN POR CMVC

En la actualidad, la mayoría de los niños que nacen con infección por CMVc en todo el mundo pasan desapercibidos, lo que se traduce en una falta de tratamiento necesario o de seguimiento. Lo idóneo es diagnosticar la infección por CMVc mediante pruebas analíticas realizadas al lactante en las tres primeras semanas de vida. Si se sospecha la presencia de CMVc después de tres semanas, se puede realizar un análisis retrospectivo mediante PCR de manchas de sangre seca, si bien no es un método muy sensible para diagnosticar el CMVc. En Suecia, las muestras de sangre seca se conservan indefinidamente, pero en la mayoría de los países se conservan solo por un tiempo limitado, lo que dificulta el diagnóstico retrospectivo.

Los programas de cribado universal o selectivo auditivo se implementan en pocas regiones del mundo, si bien facilitarían la detección, la identificación y el tratamiento de niños con infección sintomática por CMVc. En Ontario, Canadá, se presentó en 2019 el primer programa de cribado universal del CMV del mundo, y Minnesota se convirtió en 2023 en el primer estado estadounidense en implementar un programa de este tipo. Otros estados estadounidenses han adoptado programas de cribado auditivo, programas de cribado selectivo auditivo ampliado o hacen campaña a favor de dichos programas.

Una limitación importante del cribado selectivo auditivo es que la mayoría de los niños que nacen con infección por CMVc superan el cribado auditivo neonatal universal, pero pueden desarrollar una pérdida auditiva progresiva con posterioridad. Esto supone un reto porque los cuidadores a menudo desconocen la existencia del CMVc o la posibilidad de una pérdida auditiva progresiva, por lo que pueden ignorar hitos auditivos y fonológicos tempranos cruciales para el desarrollo del lenguaje (Blamey et al., 2001; Ching et al., 2017). Además, los patrones cerebrales atípicos asociados con el CMVc pueden influir en los resultados a largo plazo (Lanzieri et al., 2017) y no solo en aquellos con CMVc sintomático (Falkenius Schmidt et al., 2024).

En Utah, el programa de cribado selectivo introducido en 2013 ha evolucionado hacia un *programa de cribado selectivo ampliado*. Este enfoque actualizado incluye grupos adicionales de alto riesgo, como aquellos con un tamaño de cabeza anómalo (microcefalia), positividad materna para CMV o bebés con erupción petequial o erupción de hematopoyesis extramedular (Suarez et al., 2023). El programa selectivo ampliado ha mostrado mejores tasas de detección de casos sintomáticos de CMVc en comparación con el cribado selectivo auditivo tradicional. Suarez y colegas (2023) sostienen que el cribado ampliado es una alternativa más factible que el cribado universal, ya que se requieren menos pruebas a la vez que se consiguen mayores tasas de detección.

Independientemente del tipo de programa de cribado de CMVc, es esencial facilitar un programa de seguimiento para las personas identificadas con infección por CMVc y sus familias, tal como se indica en los principios del cribado (WHO, 2020). Los estudios indican que, en general, los progenitores apoyan los programas de cribado del CMVc, incluso si deben sufragar los costos de su bolsillo (Diener et al., 2020), y a pesar de las preocupaciones iniciales de que los resultados puedan ser alarmantes (Beaulieu et al., 2024).

En el estudio dirigido por pacientes de Beaulieu et al. (2024), en el que se utilizó un Enfoque de investigación con participación de pacientes y comunidades (PaCER, por sus siglas en inglés) que abarcaba debates en grupos focales, entrevistas y reflexiones, los progenitores con experiencias vividas expresaron una gran preferencia por estar informados sobre el estado de salud de sus hijos desde un principio. Valoraban «el conocimiento como poder» y consideraban que el cribado universal podría reducir el estigma al aumentar la concienciación sobre el CMVc. Los participantes del estudio también destacaron la importancia de disponer de un programa de seguimiento tras el cribado para apoyar a las familias (Beaulieu et al., 2024).

PROCEDIMIENTOS DE SEGUIMIENTO, OPCIONES DE TRATAMIENTO E INTERVENCIÓN – MEJORES PRÁCTICAS ACTUALES

Cuando a un niño se le diagnostica una infección por CMVc al nacer o en las tres primeras semanas de vida y presenta síntomas de moderados a graves, las directrices internacionales recomiendan un tratamiento antivírico de seis meses con valganciclovir (Leruez-Ville et al., 2024). El objetivo de este tratamiento es reducir el riesgo de trastornos graves del neurodesarrollo (Rawlinson et al., 2017). No obstante, los resultados a largo plazo y los posibles efectos secundarios de este tratamiento no se conocen en su totalidad. En consecuencia, no se recomienda el tratamiento antivírico en los casos con síntomas leves (Rawlinson et al., 2017). Además del tratamiento médico, es crucial controlar la audición, la visión y el desarrollo general del niño a lo largo del tiempo. Según Rawlinson et al. (2017) se deben realizar pruebas audiológicas a los niños con CMVc cada seis meses durante los tres primeros años de vida y, posteriormente, de forma anual hasta la adolescencia. También se recomiendan exámenes oftalmológicos tempranos y evaluaciones de seguimiento. En los casos de CMVc sintomático, se deberán realizar evaluaciones del desarrollo adaptadas a cada caso. No existen recomendaciones específicas para los casos asintomáticos, si bien se aconseja un seguimiento del desarrollo continuado (Rawlinson et al., 2017).

Hasta la fecha, solo en unos pocos estudios se ha investigado el impacto a largo plazo del CMVc en la edad adulta (Falkenius Schmidt et al., 2024). La evidencia existente sugiere que los individuos con infección sintomática por CMVc tienen un alto riesgo de presentar complicaciones duraderas y comórbidas, lo que subraya la necesidad de intervenciones interdisciplinares tempranas y preventivas. No obstante, la intervención temprana por sí sola puede no ser suficiente, ya que muchas personas con infección por CMVc pueden requerir un seguimiento y un apoyo continuos durante toda su vida. Las personas que desarrollan trastornos del neurodesarrollo en la primera infancia también pueden enfrentarse a un mayor riesgo de afecciones neuropsiquiátricas posteriormente en la adolescencia o en la edad adulta, tal como se observa en otras poblaciones con trastornos coexistentes como el trastorno del espectro autista o el TDAH (Gillberg, 2010). En la actualidad no existen directrices de buenas prácticas establecidas que aborden específicamente los aspectos cognitivos y comórbidos de la intervención en el CMVc. No obstante, en la investigación y la experiencia clínica se pone de relieve la importancia no solo de vigilar la audición, sino también de identificar otros posibles problemas, como dificultades cognitivas no verbales, síntomas neuropsiquiátricos, desarrollo atípico del lenguaje hablado, pragmática deficiente, funciones ejecutivas afectadas y evaluación de la calidad de vida en general (Karltorp et al., 2014; Korndewal et al., 2017; Löfkvist et al., 2020).

Dada la complejidad de la infección por CMVc y la variabilidad de los resultados, los niños deben ser controlados periódicamente por un equipo interdisciplinar con conocimientos especializados en CMVc. Es esencial que las personas con CMVc y sus familias participen activamente en el proceso de intervención desde un principio. Un ejemplo destacable de este enfoque es la Hearing House de Auckland (Nueva Zelanda), donde la Dra. Holly Teagle y sus colegas han iniciado un programa integral sobre el CMV (Raising awareness of congenital Cytomegalovirus on the local and global stage, hearinghouse.co.nz). (Concienciación sobre el citomegalovirus congénito en el ámbito local y mundial). Este programa incluye talleres en los que las familias pueden participar en la educación y el debate, además de reunirse con un equipo interdisciplinar de CMV para obtener una evaluación y una intervención personalizadas. El equipo está formado por audiólogos, fonoaudiólogos, psicopedagogos, terapeutas ocupacionales y fisioterapeutas, pediatras del desarrollo, cirujanos especializados en otorrinolaringología, especialistas en intervención temprana y familiares.

El grupo del proyecto CMV Aoeteara investiga activamente diversos aspectos de la atención al CMV congénito (CMVc) en Nueva Zelanda. Su investigación incluye el examen de los resultados de la base de datos nacional sobre el CMV, la evaluación de los conocimientos y las actitudes de los profesionales sanitarios, la evaluación del trabajo en equipo interdisciplinar integral y centrado en el niño, y la recopilación de las perspectivas de los progenitores sobre la detección y el diagnóstico del CMV. Si bien los resultados del programa aún no se han evaluado científicamente ni se han publicado, la recopilación y el análisis de los datos siguen en curso.

CONCLUSIONES

El impacto mundial de la infección por CMVc es significativo, ya que afecta a un gran número de lactantes y a menudo provoca trastornos del neurodesarrollo a largo plazo y otras complicaciones sanitarias. Dado que actualmente no se dispone de una vacuna eficaz, es fundamental adoptar medidas preventivas como aumentar la concienciación sobre la transmisión del CMVc y promover conductas protectoras. A medida que los beneficios de los programas de cribado neonatal (ya sea universal o selectivo) se hagan más evidentes, es probable que su implantación se extienda por todo el mundo. Aunque los estudios de investigación existentes se han centrado predominantemente en los aspectos médicos de la infección por CMVc, sigue habiendo importantes lagunas de conocimiento, concretamente en lo que respecta a los efectos cognitivos a largo plazo en poblaciones más amplias y a la eficacia de diversas estrategias de intervención, además de la medicación antivírica y la tecnología auditiva. Además, es esencial realizar investigaciones adicionales para explorar las experiencias vividas por las personas con infección por CMVc y sus cuidadores, para identificar mejor las lagunas en la atención sanitaria y abordar las necesidades integrales de los afectados por la infección en entornos clínicos.

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Research Proceedings of the 2023 Global Listening and Spoken Language Virtual Symposium

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ALEXANDER GRAHAM BELL ASSOCIATION FOR THE DEAF AND HARD OF HEARING

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HEARING, SPEECH, AND LANGUAGE IN INFANTS AND TODDLERS BORN PREMATURELY

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Worldwide, 15 million babies are "born too soon" every year (Blencowe et al., 2012; Howson et al., 2013). Remarkably, prematurity survival rates now exceed 93% across all countries. More than 360,000 preterm babies survived in the United States in 2020; 10% of all births (Blencowe et al., 2012; Howson et al., 2013). While this is an incredible achievement, preterm birth places the developing infant in a particularly vulnerable state for life-long adverse developmental outcomes. Due to improved survival rates, developmental disabilities, including hearing, visual, motor, cognitive, speech-language, and literacy deficits, have also increased (McCormick & Behrman, 2007). In addition to the medical and developmental consequences, the increased cost of raising a preterm child compared to a term-born child exceeds $134,000 or >60 billion annually in U.S. dollars, adjusted for the year 2023 (Mangham et al., 2009). In contrast, the cost of early intervention is a relative bargain, at $1,240 per preterm infant, adjusted for the year 2023 (Clements et al., 2007). Early intervention is highly likely to be effective, based on the remarkable plasticity of the newborn brain (White et al., 2013).

SPEECH-LANGUAGE DEVELOPMENT AND PREMATURITY

Speech and language disorders are among the most prevalent neurodevelopmental disorders in children, with prevalence rates in the range of 5-8% of all preschool children (Law et al., 1998; Tomblin et al., 1997). In contrast, about 40% of *very/extremely preterm infants* (≤32

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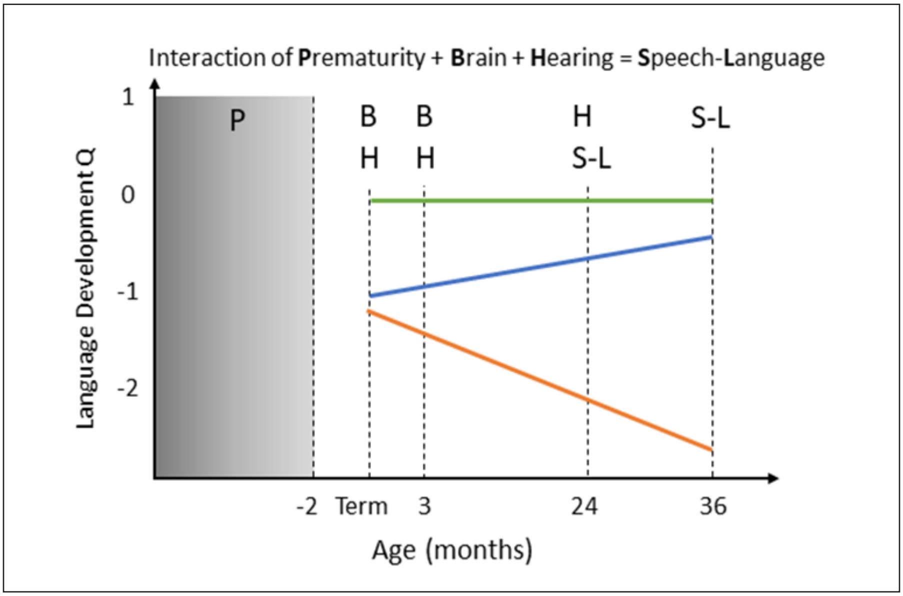
weeks gestational age) develop persistent speech-language disorders (SLD) (Duncan et al., 2012; Nguyen et al., 2018). Based on prevalence estimates, about 40,000 very-extremely preterm babies (≤32 wks. gestational age, hereafter referred to as preterm) develop SLD (Duncan et al., 2012; Nguyen et al., 2018) annually in the United States. Accurate identification of SLD in children born premature often does not occur until age 3-5 years, and persists in about 2 out of every 3 children (Roulstone et al., 2003), placing them at high risk for life-long poor educational, vocational, and social outcomes (Bashir & Scavuzzo, 1992; Young et al., 2002). Currently, it is not possible to accurately predict in the first year of life which children will develop SLD. As a result, speech-language therapy is the most delayed and the least common of the therapy services provided to infants after Neonatal Intensive Care Unit (NICU) discharge (Nwabara et al., 2017). Precious time is lost during the first three years, in which intervention is most effective, resulting in increasing speech, language, and reading deficits at school age (Vohr et al., 2018; Vohr, 2016). Our research program is informed by longitudinal studies (Figure 1) in preterm children that show typical language

Figure 1. Developmental trajectories in prematurity (P), in relation to speech-language and literacy outcomes (S-L): Measures include Brain connectivity/processing (B); Hearing loss (H). Three trajectories are shown: Normal (Green line); Resilient (Blue line); and High Risk for SLD (Orange line).



development in 32%, delay that resolves over time in 28%, and persistent/increasing delay in 40% (Duncan et al., 2012; Nguyen et al., 2018), who need early identification and intervention.

HEARING LOSS AND PREMATURITY

Overall, ten times as many preterm compared to term babies have some degree of permanent hearing loss (Hirvonen et al., 2018; Robertson et al., 2009), compounding the impact on communication difficulties (Hirvonen et al., 2018; Robertson et al., 2009). Prevalence estimates for hearing loss varies widely due to variable diagnostic criteria and risk factors, e.g. brain injury, prenatal infections, hyperbilirubinemia, and exposure to drugs that are toxic to the ear (Cristobal & Oghalai, 2008). Preterm birth can also result in delayed or progressive hearing loss, additionally contributing to SLD (American Academy of Pediatrics, 2007). These delayed and progressive hearing losses may occur due to ongoing problems related to premature birth and these various risk factors (Joint Committee on Infant Hearing, 2019). Current methods of newborn hearing screening reliably only detect hearing loss greater than 30-40 dB HL, thus missing most slight-mild and higher frequency loss (Garinis et al., 2018). Even mild and high-frequency hearing loss increases the risk for SLD (Walker, Holte, et al., 2015; Walker, McCreery, et al., 2015). These so-called “minimal” losses are related to poorer language outcomes (Porter et al., 2013; Walker, McCreery, et al., 2015; Winiger et al., 2016) and are at least three times more prevalent than moderate/greater loss (Su & Chan, 2017; Vohr, 2016). In a large primary school cohort (n=1638), Moore and colleagues reported that prevalence of undiagnosed slight-mild hearing loss (15-25 dB HL) was 16.8% and was significantly associated with poorer language, word and nonword repetition, and speech-in-noise perception compared to better hearing levels (Moore et al., 2020). Statistical models showed that 15 dB HL is an objectively appropriate criterion for diagnosis of significant hearing loss.

Mild and high frequency hearing loss that is missed with standard screening can be detected using newer physiologic methods such as distortion product otoacoustic emissions (DPOAE) (Blankenship et al., 2018) and auditory brainstem responses (ABR) with more sensitive criteria (Sininger et al., 2018). Our goal is to detect these milder forms of hearing loss and examine the relationship to language development in preterm infants.

EARLY DETECTION OF SLD

In stark contrast to Universal Newborn Hearing Screening, there is no universal screening for speech-language risk in infants. The critical lesson from newborn screening is that intervention is more effective *the sooner it can begin after birth* (Joint Committee on Infant Hearing, 2013). Because the infrastructure of newborn screening already exists for newborns in NICUs in all U.S. states and territories, and in many other countries, employing improved screening tools before NICU discharge would be highly feasible (American Academy of Pediatrics, 2007). A greater degree of brain injury or delayed neuromaturation in language pathways, particularly in the presence of hearing loss or poorer speech processing, limits the stimulation needed to support language development. Acoustically enriched language stimulation in the first year is critical for development of normal language and later literacy skills (Nwabara et al., 2017; Vohr, 2016). Increased language exposure has been shown to be associated with increased structural connectivity, particularly in the dorsal language pathway (Romeo, Leonard, et al., 2018; Romeo, Segaran, et al., 2018). Identifying children at greater risk close to NICU discharge would allow for the provision of appropriate early intervention (i.e., increased, targeted language exposure) prior to preschool age.

SPEECH-LANGUAGE AND PRE-LITERACY DEFICITS IN PREMATURE CHILDREN

Developmental testing in NICU survivors with the Bayley Scales of Infant & Toddler Development, Third Ed. is the current clinical standard for guiding early intervention in preterm infants (Anderson & Burnett, 2017). However, there is concern that this tool overestimates cognition and language and is not predictive of later impairment in preterm children (Spencer-Smith et al., 2015). Thus, in our study, we are using two specific language assessments at age 24 months: the MacArthur Child Development Inventory (MCDI) and the Communication and Symbolic Behavior Scales (CSBS). The CSBS is a direct standardized assessment particularly well-suited to the preterm population as it assesses gestures and other non-verbal communication, allowing for a range of performance in children who are delayed in using words. We anticipate the CSBS will be sensitive to variations in communication skills not captured by the Bayley-III language subtests. At 36 months corrected age (CA), speech, language, and literacy skills are assessed using a standardized battery of speech, language, and pre-literacy tests. We are including assessment of pre-literacy skills in this population to identify precursors to later reading deficits known to occur frequently in school-age children with a history of prematurity (Allotey et al., 2018; Borchers et al., 2019; Kovachy et al., 2015; Lee et al., 2011).

SENSITIVE HEARING MEASURES

We recently reported improved diagnosis of slight and greater hearing loss at birth by developing age-appropriate norms in well babies and preterm infants (Blankenship et al., 2018; Hunter et al., 2018). In that prospective, longitudinal study of 279 infants, hearing status was verified at 9 months CA with visual reinforcement audiometry (VRA). At approximately 1 month CA, DPOAE, Wideband Absorbance (WBA), and air- and bone-conduction diagnostic tone-burst ABR (0.5-4 kHz) were measured. The efficacy of DPOAE levels to classify ears as normal or hearing loss was analyzed. We found 100% sensitivity and 76% specificity for detection of slight-mild and greater hearing loss using age-specific normative algorithms, validated against the gold standard of both threshold ABR and behavioral audiometry (Blankenship et al., 2018). In that study, 10% of the preterm babies did not pass the standard newborn screening, while 30% were found to have slight or greater hearing loss. In another recent multisite study of ABR we reported that, using improved ABR techniques, we were able to detect slight or greater hearing loss (Sininger et al., 2018). We are now employing these improved techniques to precisely determine degree of all levels of hearing, assessing prevalence of hearing loss in preterm children from 3 to 36 months CA in a new longitudinal study of very-extremely premature children.

Methods

This preliminary report describes a longitudinal, population-based cohort study of very and extremely premature infants recruited soon after birth from five Cincinnati NICUs. These hospitals care for >90% of Southwest Ohio's very preterm population. We have enrolled a total of 375 infants: 150 infants at birth, in addition to 125 infants at 24-36 months old, who were all born at ≤32 weeks gestational age. Infants with known chromosomal or congenital conditions affecting the central nervous system were excluded, because outcomes are uniformly poor in such cases. We also excluded infants that were too medically unstable to have MRI scanning at term-equivalent age. Due to the language outcome measures, families who did not primarily speak English were excluded. We collected perinatal clinical variables known or suspected to be associated with brain injury, abnormal development, or cognitive deficits (Achenbach et al., 1993; Bapat et al., 2014; Dyet et al., 2006). Extensive data about the mother and infant (Parikh et al., 2013) were collected, along with conditions known to be associated with hearing loss, such as cleft palate or Down syndrome.

Auditory Measures

Comprehensive audiologic assements were completed between 1-5 months CA and at 2-3 years CA using age appropriate measures. At both ages, middle ear function was assessed with wideband absorbance and DPOAEs were used to evaluated inner ear function. At age 3 months CA, hearing levels were estimated using ABR, completed during natural sleep using insert earphones. ABR thresholds were used to categorize hearing status as typical hearing (NH; 0-15 dB eHL) or hearing loss (HL; ≥ 16 dB eHL). At ages 2-3 years CA, the audiology protocol included VRA and/or conditioned play audiometry (CPA) using earphones to obtain ear-specific speech detection and pure tone minimum response levels (MRLs). MRLs were used to categorize hearing status into typical hearing (NH; 0-20 dB HL) or hearing loss (HL; ≥ 25 dB HL).

Speech-Language Outcomes

At 24-30 months CA, families complete the MCDI – Words and Gestures and CSBS. The MCDI, a validated, standardized parent report of early language development, is widely used in studies of young children and may identify preschoolers with language impairment (Skarakis-Doyle et al., 2009) and has been used with children ages 24 months to predict later language outcomes (Goodwin et al., 2002). The CSBS has good psychometric properties (McCathren et al., 2000) and has been shown in 2-year-olds to predict expressive vocabulary one year later (Skarakis-Doyle et al., 2009). The CSBS is video recorded and scored by study staff trained with double scoring for 20% of assessments for inter- and intra-rater reliability using Cohen's kappa. The parent/caregiver also completes the StimQ (home environment questionnaire), Dialog

Parent Report (reading habits questionnaire), and Screen Exposure Parent Report (Horowitz-Kraus et al., 2017; Hutton et al., 2019; Hutton et al., 2018).

At 36 months CA, children's articulation skills are assessed using the Goldman-Fristoe Test of Articulation-3. The Goldman-Fristoe-3 is sensitive to fricative/affricate production that is challenging for some infants with hearing loss (Moeller et al., 2007). Oral language skills will be assessed using the Test of Early Language Development-III (TELD), which is sensitive to language delays in preterm children (Monteiro-Luperi et al., 2016). A parent/caregiver also completes the MCDI – Words and Sentences. Pre-literacy skills are assessed using the Sound Blending, Letter-Word and Writing subtests of the Woodcock-Johnson-IV Tests of Early Cognitive and Academic Development, which assess phonological awareness and early alphabet/word knowledge and writing.

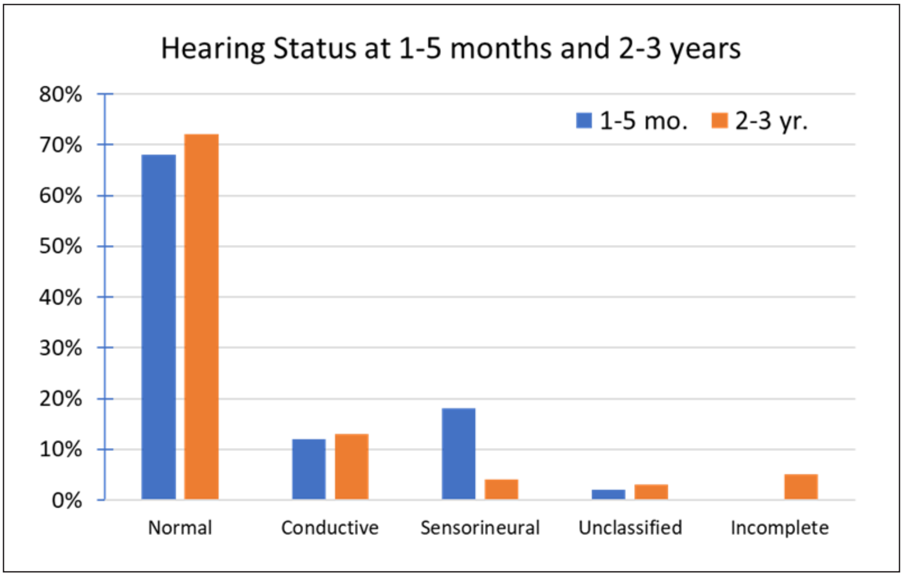
Results

In this preliminary analysis, 150 very/extremely preterm infants were assessed with MRI at fullterm CA. The Global Brain Abnormality Score, or Kidokoro Index, was assessed from the MRI scans (Brouwer et al., 2017). This measure is a composite of four regional MRI measurements including cortical gray matter, total white matter, deep nuclear gray matter and cerebellar scores. In addition, we assessed the older preterm cohort—128 children tested at 2-3 years of age with the full audiologic test battery (average 30 months CA, range 24-42 months).

Auditory Measures

ABRs were completed in 133 preterm infants at 3 months CA (*range*: 1-5 months). Results showed a high percentage of infant ears with hearing loss (32%), with the degree of loss ranging from slight to moderately severe in degree. Most ears had a sensorineural loss (18%), with a smaller percentage having a conductive loss (12%) and 2% with an undetermined type of loss. At age 2-3 years, the behavioral test protocol was highly successful when repeated results were included, resulting in ear-specific hearing levels in 88% of children (*n*=113 of 128), while 11 (9%) had only sound field MRLs and 3% had no reliable responses. At ages 2-3 years, hearing was abnormal in one or both ears in 20% of cases. Bone conduction hearing and/or tympanometry showed a middle ear component in 13%, while 4% were sensorineural (normal

Figure 2. Hearing loss prevalence by type of loss at 1-5 months and 2-3 years, in a cohort of very and extremely premature children.



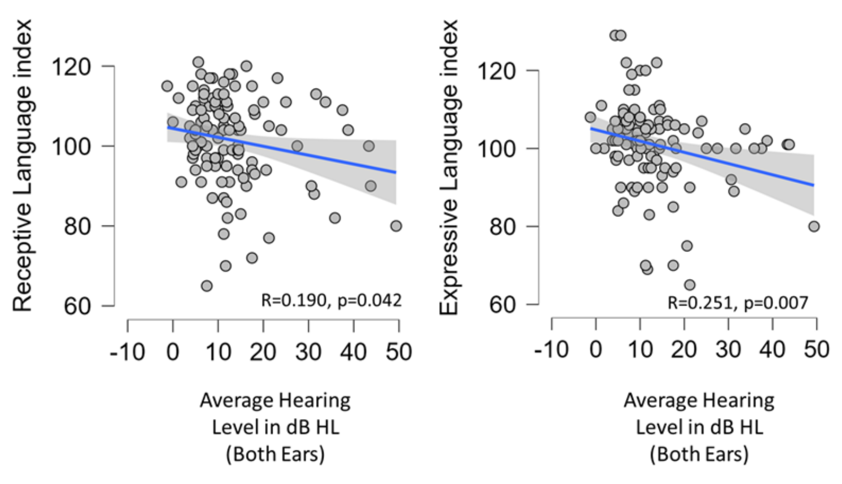
tympanometry and/or elevated bone conduction) and 3% could not be classified. Of the apparent SNHL cases, all but one had passed newborn hearing screening.

A high prevalence of hearing loss, including conductive and sensorineural type, was found in 2–3-year-old preterm children. Only four cases of apparent sensorineural loss had failed newborn hearing screening, indicating either missed or progressive hearing loss in 83% of cases. Age-appropriate otoacoustic emissions levels were highly predictive of hearing levels > 20 dB HL. The lower prevalence of sensorineural hearing loss at age 2-3 years may be due to different birth cohorts for the two age groups, or different hearing measurement techniques (ABR compared to behavioral).

Speech Language Outcomes

Standard scores for the TELD-III for receptive and expressive language subscales at 3 years CA were correlated with average hearing levels (1-8 kHz) for both ears combined in the 2-3 years CA children. As shown in Figure 3, a wide range of scores from severely low to above average was seen for both receptive and expressive language. The mean language scores tended to slope downward, with poorer average hearing levels. Notably, above average expressive language scores were only found in children with excellent hearing levels (better than 20 dB HL). There was also a cluster of severely below average language scores despite excellent hearing levels, thus other factors are clearly at play in this cohort of premature children. Analysis using MRI, EEG, and MMR measures is in progress, and hopefully will shed light on altered neurologic factors that are predictive of the language outcomes, using deep learning approaches for improved prediction of outcomes (He et al., 2020; Li et al., 2018).

Figure 3. Correlations between average hearing level and language standard scores (Mean=100, *SD*=15) on the Test of Early Language Development in 3-year-old children. Hearing was measured at 2-3 years of age.

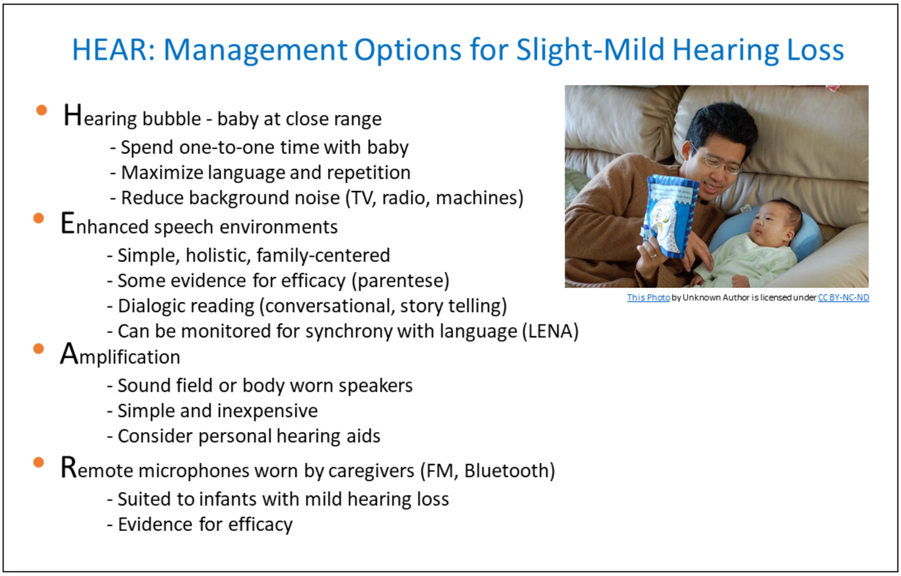


INTERVENTION METHODS

Early language environments and caregiver input play a critical role in shaping children's linguistic, academic, and cognitive development (Cartmill et al., 2013; Kuhl, 2010). For slight to mild hearing loss, there are several viable intervention options, as outlined in Figure 4. These are organized using the mnemonic "HEAR" for easy reference. The first important concept for parents to be aware of is the need for communication within the infant's "Hearing Bubble," which is ideally within 3 to 4 feet. The caregiver's speech will be most audible and clear within this range, and less affected by background noise that can affect speech perception (Leibold et al., 2016). At further distances, the loudness and clarity of speech drops, especially for high frequencies, thus affecting consonant reception. This range is best achieved during one-on-one activities with the baby that also provide language stimulation—book reading in particular. Infants benefit from lots of language stimulation, so caregivers can be coached to provide "play by play" talk whenever they are near the baby, talking about everything they are doing, naming while pointing to objects, and singing. Whenever possible, it is best to turn off extraneous sources of noise, especially electronic devices such as TV, radio, appliances, and machines. Although noise machines are highly popular among parents to encourage sleeping, they are potentially damaging to auditory development, because they produce meaningless extraneous noise, so should be discouraged. Stimulation therapies in the NICU such as Kangaroo Care and Mother-Infant Transaction Programs have been shown to improve cognitive outcomes in preterm infants later in childhood (Benzies et al., 2013; Puthussery et al., 2018).

Enhanced speech environments that are natural and play-based with human caregivers provide important "incidental language cues" that serve to enhance children's language development (Hart & Risley, 1980),

Figure 4. HEAR Recommendations: Management options for slight-mild hearing loss that can be implemented from infancy onwards.



HEAR: Management Options for Slight-Mild Hearing Loss

- Hearing bubble - baby at close range
    - Spend one-to-one time with baby
    - Maximize language and repetition
    - Reduce background noise (TV, radio, machines)
- Enhanced speech environments
    - Simple, holistic, family-centered
    - Some evidence for efficacy (parentese)
    - Dialogic reading (conversational, story telling)
    - Can be monitored for synchrony with language (LENA)
- Amplification
    - Sound field or body worn speakers
    - Simple and inexpensive
    - Consider personal hearing aids
- Remote microphones worn by caregivers (FM, Bluetooth)
    - Suited to infants with mild hearing loss
    - Evidence for efficacy

This Photo by Unknown Author is licensed under CC BY-NC-ND and can even accelerate language growth in premature children with brain injury (Rowe et al., 2009). Quality and quantity of real language, rather than from electronic speech, such as cartoons or other recordings, is preferable particularly for children with hearing loss (Ambrose et al., 2014). To develop language and socialization, interaction with people is always best. The characteristic "baby talk or parentese" that most parents naturally use is universal across all languages, and supports language development through repetition, emphasis of prosodic cues, emotional connections, and support for theory of mind development (Ferjan Ramirez et al., 2020). An intervention approach known as "dialogic reading" involves training caregivers to read to their infants and children using shared storybooks to create conversations (reciprocal communication) with the baby or child. Dialogic reading is associated with attention-focused neural connectivity (Farah et al., 2019), and has positive impacts on both literacy and general language skills.

Amplification is effective for improved language outcomes in mild hearing loss (Tomblin et al., 2014), although it is rare for hearing aids to be recommended and worn regularly in such cases due to a lack of evidence in the past (McKay et al., 2008). Because it is not as clear in such cases that the infant is not hearing well, slight to mild hearing loss is more invisible than moderate to profound hearing loss. Parents may be less willing to invest the financial and time resources needed to obtain and implement hearing aids in such cases. However, the evidence is that infants with mild hearing loss can be as far behind as infants with more severe hearing loss, likely because they are not receiving intervention. Thus, it is always worthwhile to discuss hearing aids and the parents' willingness to try them. It is important to explain that results will be subtle, and it may take time to see the benefit. An alternative approach is the use of remote microphone systems (RMS) to reduce extraneous noise and bring the parent's voice into the hearing bubble even when at a distance. A study using Language Environment Analysis Software (LENA) showed that with the use of RMS, children could have access to approximately 42% more words per day (Benitez-Barrera et al., 2018), and the amount of child-directed speech increases with use of RMS (Benitez-Barrera et al., 2019). In addition, caregivers reported positive perceived communication benefits of RMS, and they tended to talk more from a distance when using the RMS (Benitez-Barrera et al., 2018). Remote microphone technologies are especially beneficial once children are in daycare, preschool or elementary school, and can provide substantial benefits for children with slight-mild hearing loss.

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LISTENING-RELATED FATIGUE IN DEAF AND HARD OF HEARING STUDENTS: UNDERSTANDING AND MANAGING THE "FATIGUE FACTOR"

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Fatigue is often defined as a mood state, a subjective feeling of tiredness or exhaustion that reduces our desire to continue working on a task (Bess et al., 2020; Hockey, 2013). Fatigue is ubiquitous, something most adults and children experience when struggling to complete a long or challenging task. Most of the time, all that is needed to recover from the fatigue is a quick break or cessation of the fatiguing task. In these cases, the negative effects of fatigue are minimal. For some individuals, however, their fatigue experiences may be more severe and sustained. When recurrent, this kind of severe fatigue can have significant, negative consequences, especially for children. Severe fatigue is more common in children suffering from chronic health issues, such as cancer, diabetes, and obesity (Hornsby et al., 2017; Hockenberry-Eaton et al., 1999). Children with such health conditions who also suffer from significant fatigue tend to face additional academic and social struggles compared to their non-fatigued peers (Stoff et al., 1989; Berrin, et al., 2007; Ravid et al., 2009). But what does this have to do with children who are deaf and hard of hearing (DHH)? Evidence continues to mount suggesting that children who are DHH are also more likely to experience severe fatigue as a result of their listening and understanding challenges—a term coined listening-related fatigue.

FATIGUE IN DHH STUDENTS

Parents and teachers have long observed fatigue-related signs and symptoms in children who are DHH, attributing these issues to the increased effort required of the children when listening and trying to communicate with others. These concerns have brought the topic of listening-related fatigue to the forefront of hearing sciences research. To learn more about the fatigue experience of children who are DHH, our

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research group conducted a series of interviews and focus groups with DHH students, their parents, and teachers of DHH students (Davis et al., 2021).

In these discussions, we learned more about the fatigue experiences of children who are DHH, what situations they considered fatiguing, and how they recovered from their fatigue. It may come as no surprise to those in hearing healthcare that listening in background noise was noted as one of the most challenging and fatiguing listening situations. Listening in groups rather than one-on-one situations; in acoustically difficult environments, such as the cafeteria; outside when playing sports; and via media (e.g., telephone) were also fatiguing. Participants' fatigue experiences were vast and varied, and included physical, emotional, social, and cognitive manifestations. For example, physical issues such as headaches, feeling drained, and the need for naps or rest after sustained listening experiences were reported. Children also noted feeling a wide range of emotions that appeared tied to their fatigue, such as becoming frustrated, stressed, or angry. Adults (parents and teachers) also observed social and emotional behaviors that they associated with the children's fatigue, such as acting out or withdrawing in certain situations. Additionally, when listening demands were high, they reported children who are DHH were more likely to zone/tune out or have more difficulty staying on task than children without hearing loss (cognitive fatigue). Some children who are DHH self-reported their "brain feeling tired" after needing to put forth effort to listen and understand at school or in social situations. These reports helped us better understand listening-related fatigue in this population and were the basis for the development of a theoretical framework to highlight the challenges faced by DHH students (Davis et al., 2021). Table 1 shows a modified version of that framework and highlights some of the important situational determinants of listening-related fatigue (i.e., situations that brought about their fatigue) in children who are DHH and some of the more common physical, social-emotional, and cognitive characteristics of their fatigue. For a full description of these, and other factors, see Davis et al. (2021).

Table 1: Situational determinants and experiences of fatigue noted by DHH children, their parents, and teachers. Modified from the listening-related fatigue theoretical framework found in Davis et al., 2021.

What precedes/causes you to feel listening-related fatigue?	How does it feel when you are fatigued?
Participant responses: • Noisy listening situations • Group settings with many people talking • Listening in the cafeteria, gymnasium, during sporting events • When I feel pressure to listen or do well in a situation • When I have to do many things at once (listen and complete another task) • Teachers noted students with additional disabilities, increased motivation to listen, or broken hearing equipment seemed additionally fatigued	**Participant responses:** • Physical: ◦ Drained ◦ Tired ◦ Exhausted ◦ I get headaches ◦ I need to rest • Cognitive: ◦ I will zone out or not pay attention ◦ It is hard to concentrate or focus ◦ My brain "feels tired" ◦ I will day dream • Social-Emotional Experiences ◦ I feel frustrated or angry ◦ I will give up or shut down ◦ I will avoid situations or withdraw from others ◦ Teachers and parents noted increased behavior problems or acting out when child was fatigued

PEDIATRIC VERSIONS OF THE VANDERBILT FATIGUE SCALES: THE VFS-PEDS

The results of these focus groups and interviews confirmed prior anecdotal reports and earlier research suggesting listening-related fatigue was a significant problem for at least some children who are DHH. The study findings highlighted the impact of listening-related fatigue on these children and the need for a sensitive and reliable tool measure the construct. The rich qualitative data from the focus groups and interviews provided us with the foundational information needed to create the pediatric versions of the Vanderbilt Fatigue Scales: the VFS-Peds. The VFS-Peds is a suite of three questionnaires designed to assess long-term, listening-related fatigue (i.e., the fatigue experienced during a typical week) in populations dealing with communication challenges (e.g., hearing loss, speech-language disorders, etc.). The VFS-Peds consists of a child self-report scale (VFS-C), as well as parent and teacher proxy-report scales (VFS-P and VFS-T, respectively). The VFS-C and

VFS-T provide a single, unidimensional, measure of the child's listening-related fatigue. In contrast, the VFS-P is multidimensional, providing separate measures of the child's mental and physical fatigue. A detailed description of the VFS-Peds development and validation can be found in Hornsby et al. (2022). Briefly, quotes from the focus groups and interviews were utilized to create a large pool of potential questionnaire items for assessing listening-related fatigue that were relevant to the target respondent group (i.e., child, parent, or teacher). For example, a parent reported their child had gone to a museum for a field trip and "the gentleman was great, but he spoke so fast—she [the DHH child] was still missing stuff. In a very hectic environment, I can tell it's a lot for her. She has to make an effort, and it wears her out." This, and similar quotes, were transformed into the VFS-P item: *Listening takes a lot of effort for my child*. Once a large pool of potential test items was developed, data were collected from several hundred children who are DHH, their parents, and teachers of children who are DHH. These data were analyzed to identify high-quality items for use in the VFS-Peds and resulted in a high-quality (i.e., valid, reliable, and sensitive), relevant measure of listening-related fatigue (Hornsby et al., 2022).

In addition to the original English version of the VFS-Peds, several research groups have created, or are currently working on creating, culturally and linguistically appropriate translations. To date, translated versions of the VFS-Peds are available in Mandarin, Italian, and Spanish (US/Castilian). Copies of the English VFS-Peds, its translations, and a list of ongoing collaborations to translate the scales into other languages, can be found at our website (https://www.vumc.org/vfs).

USING THE VFS-PEDS SCALES

Please refer to the VFS-Peds User Guide (https://www.vumc.org/vfs) for a detailed description of administration and scoring guidelines. All scales use a simple, five-point (0-4), Likert frequency response format to quantify how frequently a fatigue-related situation or experience occurs in a typical week. Response options include Never (0), Rarely (1), Sometimes (2), Often (3), and Almost Always (4). Using the VFS-Peds, a child's fatigue can be quantified two ways:

1. By using Item Response Theory (IRT) to calculate an IRT scale score. This approach provides the most precise estimate

of an individual's fatigue experience but requires dedicated computer software and algorithms to calculate and is most appropriate for analysis of group data for research purposes.

2. Participant responses can also simply be summed to create a total fatigue score (for the VFS-C and VFS-T) or two subscale scores (mental and physical fatigue scores) for the VFS-P. Individual total or subscale scores can be compared to scores from a standardization sample of typically developing children as described in our user's manual (https://www.vumc.org/vfs). This simple summed score approach is recommended for clinical purposes.

Whenever possible we recommend administering the entire suite of Vanderbilt Fatigue Scales to better understand how, if at all, listening-related fatigue is affecting students who are DHH. The VFS-C is a self-report measure completed by the student, thus giving you information about the child's fatigue from their own perspective. However, not all children will be able to provide reliable responses using the VFS-C. Likewise, results from our focus groups and interviews suggest that some children with long-standing hearing loss may be unaware that the listening difficulties and resultant fatigue they experience are atypical and may underestimate the impact of their fatigue. In these cases, parents and/or teachers of students who are DHH may be better able to identify areas of concerns. The VFS-P is a proxy-report measure, completed by the child's parent/caregiver, that asks questions about the child's after-school responses to listening-related fatigue. The VFS-T is also a proxy-report measure and should be completed only by a teacher/service provider who has direct knowledge of the student's behaviors in a variety of settings. Work examining concordance between the VFS-Peds self- and proxy-report measures is lacking; however, research using generic fatigue scales suggests that some variations between the target respondents is likely (e.g., Hornsby et al., 2017). Given that parents and teachers see the child in very different settings, their impression of the frequency and magnitude of fatigue-related issues may vary, and all could be valid. For these reasons we recommend that you collect responses from all three sources (student, parent, and teacher) whenever possible. Identification of significant fatigue-related issues from any respondent could warrant additional follow-up. The VFS-Peds can be downloaded and printed out to allow for hard-copy completion. In addition, English versions of the scales can be downloaded as electronic files, which can be completed and scored using a computer or tablet device.

In our experience, most parents and teachers can read the VFS instructions and complete the scales with minimal guidance in under 5 minutes. Children completing the self-report scale may require additional guidance depending on their age and cognitive abilities. We discuss additional techniques to administer the VFS-C to children and to enhance the clinical information provided by VFS-Peds below.

Tips for Administering the VFS-C

- In our experience, children younger than 10 years of age may struggle with conceptualizing fatigue. It is recommended that you verify the child understands the concepts and task prior to administering the VFS-C. Asking questions like “what does fatigue mean?” “How does it feel when you experience fatigue?” can get you started.
- Read the instructions and questionnaire aloud to the child. Verify their understanding of the response options (never, rarely, sometimes, often, almost always) before beginning.
    - **IMPORTANT:** If you note that the child is not understanding the concept of fatigue, the task of completing the questionnaire, and/or the available answer choices, discontinue administration.
- Have the child complete the scale in a quiet environment, using their hearing assistive technology (if applicable).
- Throughout administration of the VFS-C, ask probing questions. You can obtain significant information from the student as you talk through the questions together. For example, if a child answers, “almost always” to a question, ask them for an example (“Where did this occur?” “Does this happen every day at school?”) or simply say “Tell me more”. The responses you obtain can help guide important intervention and management strategies. The following section of Case Studies exemplifies the concept of using interviews in conjunction with scoring the VFS-Peds.

VFS-PEDS CASE STUDIES

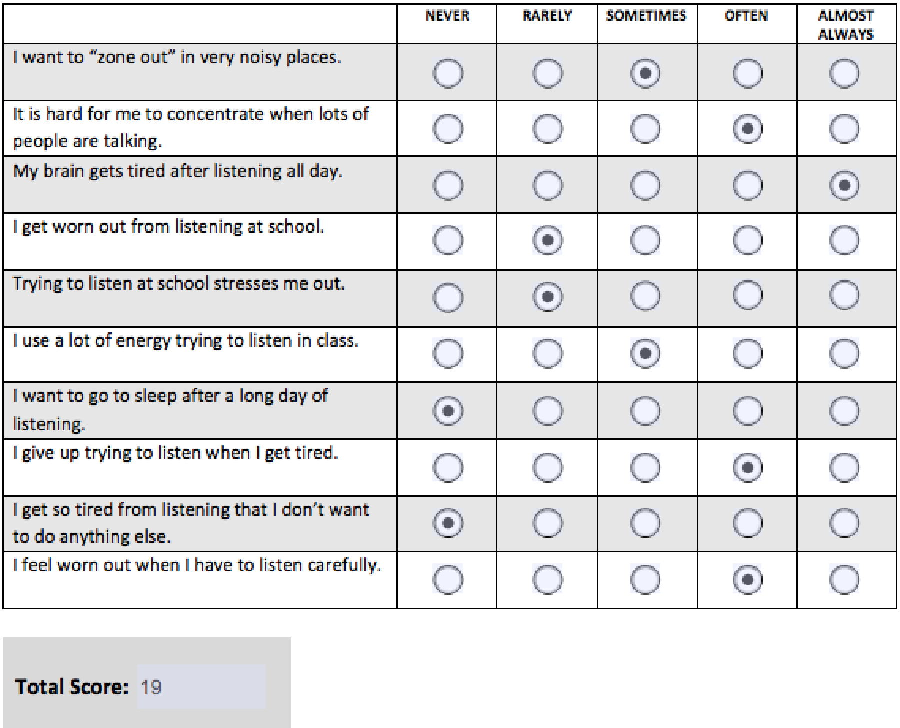
The VFS-Peds have been administered routinely to parents, students, and teachers as part of the assessment battery for eligibility and program planning purposes. When a child's VFS-Peds summed score exceeds standardization norms, it provides a quantitative rationale for initiating additional follow up. However, the score alone does not provide clear guidance as to what follow-up should be taken. In addition, typically developing children can also experience significant fatigue-related issues, as such, there may be cases where a child's fatigue score may not exceed standardization norms even though the child, parent, and/or teacher have voiced concerns. In these situations, we've found the scale scores can be coupled with student interview responses to illuminate "invisible" challenges for the students. This type of information has enabled us to work with educators and caregivers to better understand the child's struggles with listening-related fatigue and how to work more effectively to remediate those issues. The case studies below come from the experiences of the first author (H. Davis) and highlight this approach.

Case Study 1: Jacob

Earlier this school year, a parent contacted me about concerns for fatigue in her child who is DHH. Her son has bilateral, mild-to-moderate sensorineural hearing loss and uses hearing aids. He is currently in the sixth grade and had started exhibiting difficulty with academic coursework, appearing to the educational team as off-task and "not getting it". I completed the VFS-P with the parent and the VFS-C with the student via a virtual meeting after school. The parent's scores were 18 (out of 28) for the mental subscale, suggesting mental fatigue-related issues were relatively common; and 10 (out of 20) for the physical subscale, suggesting physical fatigue-related issues were less frequent. While the overall scores were low (i.e., they did not exceed standardization norms), for two specific items the parent reported the child would "almost always" 1) get tired of listening by the end of the day and "almost always" 2) "give up" in difficult listening situations.

The child completed the VFS-C, scoring 19 out of 40, which is in the low fatigue range per our summed scoring system suggesting that some fatigue-related issues occurred, but not frequently in all conditions. However, a review of his individual responses showed that for 40% of the VFS-C items (4 out of 10), he reported that fatigue-related issues were occurring “often” or “almost always” (See Figure 1 below). He noted his brain was “almost always” tired after listening all day, that “often” it was hard for him to concentrate when lots of people were talking, “often” he would give up trying to listen when tired, and “often” he felt worn out when he had to listen carefully.

Figure 1. VFS-C responses and summed score for Case Study 1



	NEVER	RARELY	SOMETIMES	OFTEN	ALMOST ALWAYS
I want to “zone out” in very noisy places.	○	○	●	○	○
It is hard for me to concentrate when lots of people are talking.	○	○	○	●	○
My brain gets tired after listening all day.	○	○	○	○	●
I get worn out from listening at school.	○	●	○	○	○
Trying to listen at school stresses me out.	○	●	○	○	○
I use a lot of energy trying to listen in class.	○	○	●	○	○
I want to go to sleep after a long day of listening.	●	○	○	○	○
I give up trying to listen when I get tired.	○	○	○	●	○
I get so tired from listening that I don’t want to do anything else.	●	○	○	○	○
I feel worn out when I have to listen carefully.	○	○	○	●	○

Total Score: 19

While completing the VFS-C with the student, I also asked probing questions. See below for examples from our conversation.

Audiologist: What is zoning out?

Student: I stop focusing, not staring, but not looking at anything. If I have a big class with a lot of people in it, sometimes that will happen.

Audiologist: Why did you say that you “almost always” have difficulty concentrating in this situation?

> **Student:** When lots of people are talking, I will zone out or I don't focus all my hearing onto them. I focus on the teacher but not really on the class discussion.
>
> **Audiologist:** Tell me more about what it feels like when your brain gets tired.
>
> **Student:** If I'm doing school classes all day, my brain, my ears get tired after school. It depends on the classes, like a really long lesson can be more tiresome.
>
> **Audiologist:** What do you mean—you use a lot of energy trying to listen?
>
> **Student:** I use a lot of energy—sometimes if a lot of people are talking and I'm really tired, I'll try to push through it and it just takes a lot of my energy. If I keep pushing myself if I'm really listening, then I say "I'm done, I'm done" and then I'm really tired and want to give up. I feel this way at school but not at home or with my friends. I'm tired of hearing so much and I just want to stop listening, I just want to take a listening break.

This information was sent to the student's educational team and the feedback I received was positive—although they knew about fatigue, the team felt the information was very insightful and helped give them a better understanding the struggles the student was facing on a daily basis. Based in part on these results, the school implemented listening breaks and check-ins with the student to better support their listening needs.

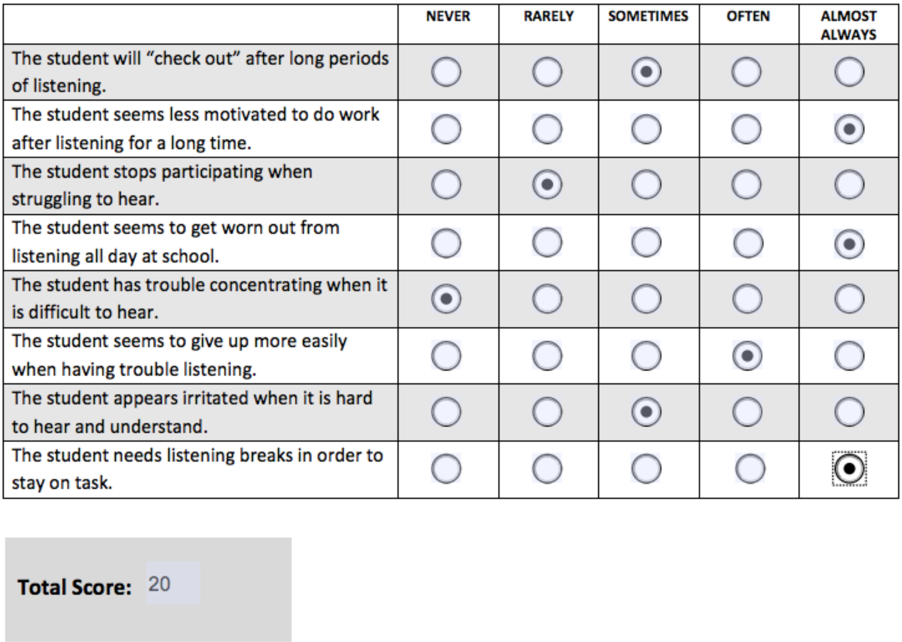
Case Study 2: Miranda

Miranda is a 3rd grade, 8-year-old student with bilateral, profound hearing loss. She utilizes cochlear implants and a remote microphone system at school. As part of an evaluation for her upcoming re-evaluation meeting, the VFS-C and VFS-T were administered to Miranda and her teacher, respectively.

The VFS-C was administered in a quiet space in Miranda's elementary school. I attempted to discuss fatigue with Miranda, providing examples ("Sometimes I feel tired from having to do my work at school. Does this ever happen to you?") but she was unable to understand the concept. At that point, I decided to forgo collecting formal data via the VFS-C.

The VFS-T was emailed to her teacher, who completed and sent it back (see Figure 2), along with a few notes about Miranda's progress in the classroom. She noted that Miranda would often cry during the end of the school day and want to lay her head on her desk. The VFS-T score of 20 was in the low fatigue range; however, the teachers' responses indicated consistent issues with reduced motivation after challenging listening situations, appearance of being worn out, and the need for listening breaks. Although not formally written into her plan, the teacher informed me that she had recently allowed Miranda to take a break around 1:30pm (defined as getting up from her desk and going to get a drink of water from the fountain in the back of the room) in order to help her stay on task. The teacher noted improvement in Miranda's behavior in the afternoon with this recent change.

Figure 2. VFS-T responses and summed score for Case Study 2.



	NEVER	RARELY	SOMETIMES	OFTEN	ALMOST ALWAYS
The student will "check out" after long periods of listening.	○	○	●	○	○
The student seems less motivated to do work after listening for a long time.	○	○	○	○	●
The student stops participating when struggling to hear.	○	●	○	○	○
The student seems to get worn out from listening all day at school.	○	○	○	○	●
The student has trouble concentrating when it is difficult to hear.	●	○	○	○	○
The student seems to give up more easily when having trouble listening.	○	○	○	●	○
The student appears irritated when it is hard to hear and understand.	○	○	●	○	○
The student needs listening breaks in order to stay on task.	○	○	○	○	●

Total Score: 20

At the re-evaluation meeting, the VFS-T results were shared with the educational team, including Miranda's family. The team came to the agreement that an accommodation of a listening break should be added to Miranda's IEP. Two breaks were initialized: one at the end of the day before the last time of group instruction and one after her speech-language therapy services as those required significant effort on Miranda's part.

MANAGING LISTENING-RELATED FATIGUE IN CHILDREN

The information and case studies above highlight the increased risk students who are DHH have for experiencing listening-related fatigue and the impact it can have in a school setting. Unfortunately, to date, systematic research supporting specific interventions or strategies to reduce difficulties associated with listening-related fatigue has not been conducted. We recommend that you observe the student in various settings throughout the school day and use the VFS-Peds, and follow-up interviews, to identify those experiencing significant issues. Probing interview questions can assist you and the child's educational team in tailoring a plan to help minimize their fatigue and any potential negative effects. In the absence of empirically validated approaches, we have used information from our focus group participants (children, parents, and teachers) and our professional experiences to create a management recommendation summary sheet that can be downloaded from our website. Key points from this summary sheet include:

- Consider the acoustics of the classroom/other listening spaces and reduce background noise as much as possible. Background noise was repeatedly noted as fatiguing for students who are DHH.
- The student should receive flexible, preferential seating. This seat provides the best visual and auditory access and may need to change based on the activity.
- Verify the student's amplification (e.g., hearing aids, cochlear implants, bone anchored devices, remote microphone systems) are fit according to prescriptive targets and are functioning appropriately.
- Provide scheduled listening breaks—time where the student is permitted to take a break from attentive listening in the classroom, particularly after a period of difficult listening. Examples include taking a short movement break like standing and stretching or going to the restroom or for a water break.

Alternatively, the child may take a break from work while remaining at their desk and/or remove their hearing assistive technology (e.g., hearing aid, cochlear implant) for a short period of time. Although full time use of amplification is the gold standard, some have reported that taking a brief break from their devices was an effective tool for reducing fatigue development or speeding recovery from fatigue. This recommendation is individualized based on the student's needs and responsibility level. For parents, this may include allowing the child to take a break or nap after school, or occasionally avoid social events in the evening if they are already tired from the day.

- Consider scheduling auditory-heavy therapies or lectures for the morning. Many DHH students report feeling more tired by the end of the school day.
- Educate the child, their parents, and their teachers about listening-related fatigue and its relation to hearing loss and listening difficulties. This education may enhance their awareness of fatigue, its contributors, its consequences, and potential approaches for minimizing its negative effects.

Of note, compared to adults, children who are DHH have more concerns about barriers to utilizing these strategies, citing worries about getting in trouble or missing important content if they didn't continually stay connected in listening situations. Including the student in the decision-making process may help to ensure they understand how to advocate for and use these strategies both at home and school. With your support, these students can learn more about themselves and their needs as it relates to listening-related fatigue, providing with opportunities for successfully navigating this experience.

Resources

The VFS-Peds and related resources are available, free of charge, at our website: https://www.vumc.org/vfs/vanderbilt-fatigue-scales.

Contact

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HEARING LOSS IN THE 21ST CENTURY AND BEYOND: A NEW ERA OF PRECISION DIAGNOSIS AND TREATMENT USING GENOMICS

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The past fifteen years has seen a dramatic improvement in our understanding of hearing and hearing loss. Scientists have worked to identify dozens of genes involved in hearing loss and elucidate the complex molecular machinery responsible for hearing. At the same time, there have been rapid advances in clinical diagnostic tools including imaging and genetic tests. What this means is that for the first time, for the majority of children with hearing loss, we are able to obtain a diagnosis.

HEARING LOSS IS NOT A DIAGNOSIS

In reality, hearing loss is not a diagnosis. Instead, hearing loss is a symptom of an underlying difference in the auditory system. The auditory system is remarkably complex and there are hundreds of possible diagnoses—from ototoxicity due to aminoglycoside drugs to molecular alterations of the mechanotransduction channel in the inner hair cell.

A diagnosis is crucial for those with hearing loss and their families as it provides information on prognosis (will the hearing loss progress over time or stay the same), recurrence risk (chances of having another child with hearing loss), and other clinical features that may be associated with hearing loss. But perhaps most importantly, a diagnosis provides an individual and their family with a sense of empowerment. They are not just a child with a symptom—hearing loss—but instead a person with a diagnosis. There are hundreds or even thousands of different diagnosis that may lead to hearing loss. A diagnosis provides a framework for the care team—physicians, audiologists, speech pathologists—to use when caring for the patient. This allows for truly patient-centered care

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and not care based on a symptom alone. Perhaps one of the best ways to demonstrate the importance of a diagnosis is that children and their families are more likely to utilize hearing aids or cochlear implants because they feel more empowered and involved in their care (Brodie et al., 2022).

Understanding the diagnosis is critical for those who care for children with hearing loss given the myriad of new treatments and treatment decisions available. Hearing aids and cochlear implants have been available for decades, but the decision to move forward with a cochlear implant may be difficult. If a diagnosis of a progressive hearing loss is made (i.e., due to *ACTG1* mutation), then the decision may be different than a diagnosis of a more stable form of hearing loss (i.e., due to *MYH14* mutation). If the diagnosis of congenital cytomegalovirus (cCMV) infection is made, then treatment with valganciclovir and antiviral medication may be indicated. And now, with clinical trials starting for gene therapy for hearing loss (described further below), an accurate genetic diagnosis is critical. A diagnosis allows us to tailor care for patients and families with hearing loss because of instead of just treating the symptom, we are treating the patient.

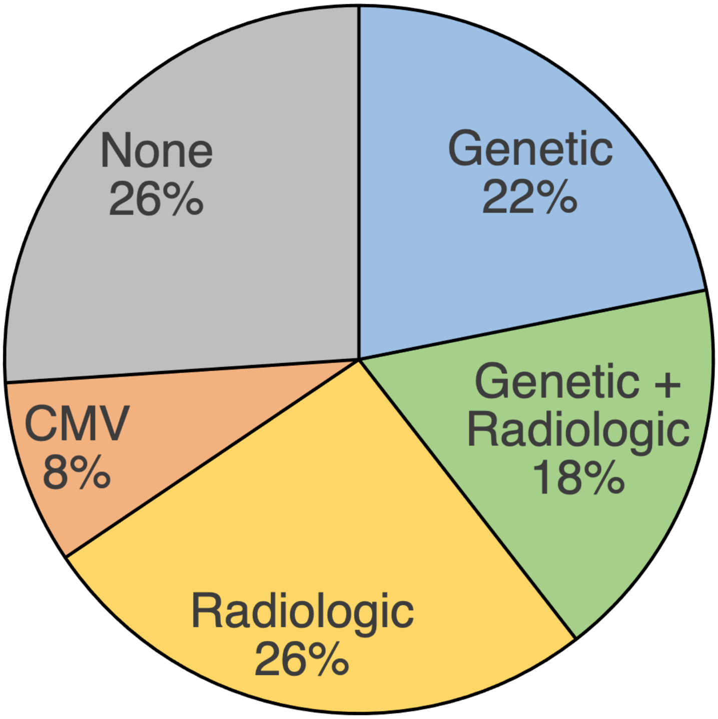
TOOLS FOR EVALUATION OF HEARING LOSS

Advances in imaging, testing congenital cytomegalovirus, and genetic testing mean that the majority of children with severe to profound hearing loss can be provided with a diagnosis. One recent study of 119 children undergoing evaluation for cochlear implants for bilateral hearing loss showed that genetic causes were identified in 22%, genetic and radiologic causes in 18%, radiologic causes in 26%, and CMV in 8% (Kim & Choi, 2022). In total, 74% of children received a diagnosis when a full diagnostic evaluation including high-resolution temporal bone MRI, CMV testing, and genetic testing (Figure 1). This is dramatically different than 20 years ago when the vast majority of children were found to have 'idiopathic' hearing loss after what was a considered a full clinical evaluation (Preciado et al., 2004).

High Resolution Imaging

Imaging of the temporal bone should be thin cut and high resolution. Imaging may be performed with either computed tomography (CT) and/or magnetic resonance imaging (MRI). The benefits of CT are the

Figure 1. Diagnostic yield in 119 children with bilateral severe to profound hearing loss. 74% of children were provided a diagnosis when genetic testing, cCMV testing, and MRI were performed. Adapted from Kim et al., 2022.



detailed view of bony anatomy which may help with surgical planning and the imaging study typically takes 1-5 minutes. However, CT exposes a child to radiation and so is often avoided, if possible, in younger children. The benefit of MRI is that the soft tissue, including the auditory nerve, is visualized. The disadvantage of MRI is that a typical imagining protocol for hearing loss may take 30-45 minutes. This means that a child will need to hold still, sleep, or undergo sedation for an MRI. Many institutions now offer a 'feed and wrap' protocol whereby a young infant is brought to the MRI appointment hungry, fed, and then swaddled tightly with ear-muffs in place. The goal is that the child will sleep naturally during the procedure and in the majority of cases this imaging is successful, with rates > 80%. The overall diagnostic yield of CT and MRI is similar, with different modalities providing higher resolution of different structures (Chen et al., 2014; Kachniarz et al., 2014). Overall, the decision on which type of imaging to pursue may vary depending on the age of the patient, the ability to hold still for a scan, and the type and degree of hearing loss.

Congenital CMV Testing

cCMV infection is the most common infectious cause of pediatric hearing loss, affecting about 20% of children with congenital hearing loss. cCMV infection leads to a spectrum of symptoms from severe brain abnormalities, liver failure, and developmental delay to more mild infections that may just include hearing loss. Testing for cCMV is difficult because the virus is present ubiquitously in our environment and should ideally be completed by 3 weeks of age. Some states have included mandates as part of newborn hearing screening programs to test for cCMV on failure of hearing screening. Many are pushing for cCMV testing to be mandated across the country. Identification of cCMV congenital infections is critical given that the infection may be treated with antivirals medications, ganciclovir and valganciclovir. These drugs have been shown to improve outcomes in severely affected children and also stop, or sometimes reverse, hearing loss in a majority of children (Lanzieri et al., 2023). cCMV testing and imaging form two pillars of diagnostic evaluation for pediatric hearing loss along with genetic testing.

GENETIC TESTING FOR HEARING LOSS

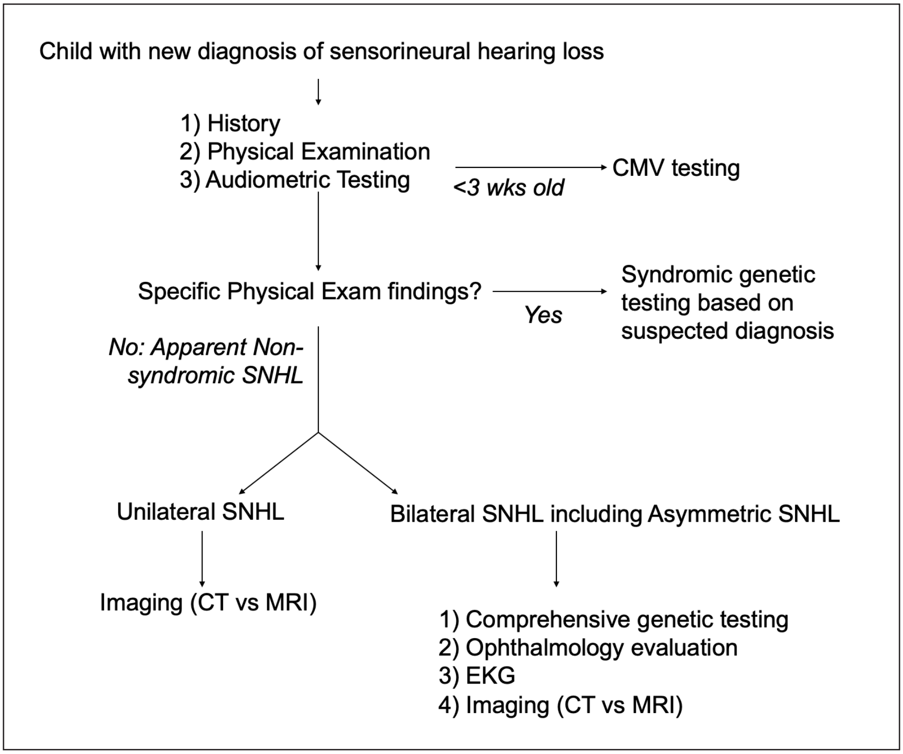
Genetic hearing loss is truly complex, demonstrating remarkable genetic heterogeneity and underscoring the exquisite complexity of human hearing. To date, more than 120 genes have been discovered that cause non-syndromic hearing loss (http://hereditaryhearingloss.org). In addition, there are hundreds more syndromic forms of hearing loss. Each of these genes may contain hundreds of genetic changes (variations) that cause hearing loss. This makes genetic testing for hearing loss difficult because testing a single hearing loss gene is simply not effective, as diagnostic yields (the chance of identifying a cause) is low. However, testing for multiple, or even hundreds, of genes was not possible until about 2010, with the advent of new genomic sequencing technology.

These new DNA sequencing methods, termed next-generation or massively parallel sequencing, provided the ability to sequence all hearing loss genes simultaneously and a new paradigm of genetic testing for hearing loss was developed based on these methods. Instead of testing a single gene, all hearing loss genes could be tested at once, increasing the speed and effectiveness of genetic testing for hearing loss (Shearer et al., 2010). Because of its effectiveness, comprehensive gene panel testing quickly became the standard of care for evaluation of children with hearing loss (Shearer et al., 2015). These genetic testing panels are now used routinely in the evaluation of children with hearing loss as they provide a diagnosis between 40-60% of the time with a single test (Downie et al., 2019; Sloan-Heggen et al., 2016). These panels are available from several different companies and differ based on the number of genes included, from about 100 to more than 200 genes. Typically, all non-syndromic hearing loss genes as well as the most common genes involved in syndromic forms of hearing loss are included. Syndromic hearing loss genes are important, as these diagnoses comprise about 20% of diagnoses in children (Perry et al., 2022).

The use of large gene panels for genetic testing for hearing loss has been established by effectiveness in dozens of studies and thousands of patients. In addition, these panels are key components of two sets of guidelines created for recommended evaluation of children with hearing loss by the International Pediatric Otolaryngology Group (Liming et al., 2016) and the American College of Genetics and Genomics (Alford et al., 2014). A proposed algorithm for diagnostic evaluation integrating these guidelines and used in my clinical practice is shown in Figure 2. This algorithm incorporates CMV testing, imaging, and comprehensive genetic testing using a gene panel.

Genetic testing should be performed on all children with sensorineural hearing loss that is bilateral, including asymmetric hearing loss. If there are syndromic features, then targeted genetic testing based on the suspected diagnosis should be performed; However this is rare, and most children will have suspected non-syndromic hearing loss. For children with unilateral (single-sided) hearing loss, imaging is typically the highest yield study. However, our recent study using exome sequencing showed that about 18% of children with unilateral hearing loss will have a genetic diagnosis and, importantly, 55% of these children will have a diagnosis of a syndromic form of hearing loss (Perry et al., 2022). These diagnoses are very impactful for these patients and suggest that we should consider incorporating genetic testing for unilateral hearing loss into our clinical practice.

Figure 2. Clinical diagnostic algorithm for evaluation of sensorineural hearing loss in children.



Comprehensive genetic testing for hearing loss can be ordered by a physician in the United States. Testing is typically performed from buccal (cheek) swabs, although blood draw may be required in some cases. There are several commercial companies that offer genetic testing in the United States and an updated list is kept by the Genetic Testing Registry available at https://www.ncbi.nlm.nih.gov/gtr/. Importantly, insurance companies have now recognized the effectiveness and importance of genetic testing for hearing loss on clinical care. As such, in the majority of cases genetic hearing loss is now covered by private insurance. Data from our clinic show that private insurance covers comprehensive genetic testing using a gene panel in approximately 80% of cases. Coverage by public insurance varies by state and in some states (e.g., California) is always covered by public insurance. Insurance coverage continues to improve as these companies recognize the utility of this testing in providing a diagnosis for children with hearing loss.

ON THE HORIZON

Genetic testing for hearing loss is very effective, but we are not able to establish a diagnosis in all cases. New technologies like whole genome sequencing and long-read genome sequencing will improve the diagnostic yield. Prices continue to drop for these technologies, making them more accessible for clinical care. Genetic testing for hearing loss also demonstrates unacceptable racial inequity in diagnostic yield—black and Hispanic children are five times less likely to obtain a diagnosis from genetic testing (Florentine et al., 2022). This is primarily due to a lack of focus of study and testing for children from minoritized races and ethnicities. Clearly further work in this area is needed to best serve these children.

The majority of children with hearing loss are identified through newborn hearing screening. Universal hearing screening guidelines were developed in 2000, and by the year 2010 more than 98% of newborns undergo newborn hearing screening in the United States. This has dramatically increased the number of babies identified early who are deaf or hard of hearing. Early identification of children with hearing loss is critical for best language outcomes (Yoshinaga-Itano, et al. 1998; 2018). Clearly, newborn hearing screening has been effective at the goal of identifying more children, earlier, with hearing loss. However, given that hearing loss is far and away the most common disorder identified during newborn screening, it is critical that we continue to work to improve its effectiveness. The current newborn hearing screening relies on a physiologic measure (otoacoustic emissions or auditory brainstem response) and is cost-effective and relatively easy to administer. This physiologic screen will not detect some mild forms of hearing loss, is not designed to detect hearing loss that may occur outside the newborn period, and may miss auditory neuropathy spectrum disorder if otoacoustic emission is used for screening. Given the effectiveness of genetic testing for hearing loss, we, and others, have advocated for incorporation of genetic screening into the newborn hearing screening program (Shearer et al., 2019). In China and some other countries, genetic screening has improved detection rates, decreased time to diagnosis, and decreased loss-to-follow up rates (Guo et al., 2020); Wu et al., 2017). Clearly there would be logistical hurdles to incorporating genetics into newborn screening, but the key is to provide an early diagnosis for hearing loss for children.

Gene therapy for hearing loss has long been a goal of those who are deaf or hard of hearing, scientists who study hearing loss, and clinicians who treat hearing loss. This goal is closer than ever with clinical trials now beginning in the U.S. and Europe for *OTOF* hearing loss. This gene was chosen for initial clinical trials because, although it is a rare form of genetic hearing loss, the mechanism is clearly understood, the key components of the inner ear are present and functioning, and treatment of adult mice with *OTOF* is effective. Clinical trials for other forms of genetic hearing loss will soon following the coming years. Importantly, any gene therapy requires an accurate genetic diagnosis.

CONCLUSION

The last decade has seen a paradigm shift in our evaluation and treatment of children with hearing loss. Now the majority of children with bilateral hearing loss may obtain a diagnosis for their hearing loss if a full diagnostic work up is performed. A diagnosis for children with hearing loss provides the patient and the clinician with valuable information and guides the management of hearing loss. This allows the clinician to treat the child and not just their symptom. New technologies will continue to improve our diagnostic yield leading to a better understanding of hearing loss.

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COCHLEAR IMPLANTS FOR CHILDREN WITH UNILATERAL HEARING LOSS: DEBUNKING THE MYTHS

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Cochlear implants (CIs) have revolutionized the field of audiology, providing a life-changing solution for children with bilateral profound hearing loss. Expanding criteria has allowed children with significant unilateral hearing loss (UHL) to benefit from this technology as well. The practice is not without controversy, however. While we have known since 1984 that children with UHL face educational challenges (Bess & Tharpe, 1984, 1986), watchful waiting until a child is old enough to be fitted with a Contralateral Routing of Signal (CROS) device has been the status quo (Bagatto, 2020; Bagatto et al., 2019). While research has continued to establish the benefits of cochlear implantation in this population, it has yet to become the standard of care. As is inherent in controversial practices, several myths and misconceptions surrounding the use of CIs for children with UHL pervade clinical hearing care systems. Here, we will debunk these myths by examining the evidence-based benefits and considerations associated with cochlear implantation in children with UHL. Through an analysis of current research and clinical evidence, we will highlight the potential advantages of CIs as a viable treatment option, promoting informed decision-making and improving the quality of life for children with UHL.

MYTH 1: CHILDREN WITH SSD WILL BE "JUST FINE" BECAUSE THEY HAVE ONE GOOD EAR

While it is true that access to sound with one ear will allow a child to develop spoken language, children with UHL face numerous challenges that can impact their educational performance, communication skills, and overall quality of life. Research has shown that children with UHL experience difficulties that may initiate in infancy as they start to show delays in auditory development before they even begin to speak (Yang et al., 2020). There have been consistent findings that UHL impacts a

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child's speech perception (Davies et al., 2021; Park, Dillon, et al., 2023), fatigue levels (Bess et al., 2020; Carpenter et al., 2022; Griffin et al., 2019; Sindhar et al., 2021), language development (Fitzpatrick et al., 2019; Lieu et al., 2010; Lieu, 2013; Sangen et al., 2017), perception of speech in noise (Corbin et al., 2021; Ehrmann-Mueller et al., 2020; Griffin et al., 2019, 2020; Park, Dillon, et al., 2021, 2023; Reeder et al., 2015), and sound localization (Corbin et al., 2021; Ehrmann-Mueller et al., 2020; Park, Dillon, et al., 2023; Reeder et al., 2015) when compared to their peers with typical hearing. These challenges can lead to academic struggles (Lieu, 2013; McSweeny et al., 2021), cognitive deficits (Lieu, 2018), and social-emotional issues. Emerging research even suggests neuronal changes in children with UHL (Calmels et al., 2022; Shang et al., 2020; Sharma et al., 2016; Vanderauwera et al., 2020). It is no longer acceptable to assume that a child with UHL will be "just fine." The research clearly shows that there are developmental differences and challenges for this population.

MYTH 2: A CROS/BCHA/REMOTE MIC/PREFERENTIAL SEATING IS GOOD ENOUGH

While devices like CROS systems, bone conduction hearing aids (BCHA), remote microphones, or preferential seating can provide some benefits for children with UHL, they are simply assistive technologies. None of these systems attempt to treat the hearing loss as they do not offer bilateral hearing. In fact, rerouting devices such as a BCHA and CROS can be detrimental in situations where noise is at the poorer ear (Bagatto et al., 2019; Choi et al., 2019; Griffin et al., 2022; Picou, Lewis, et al., 2020). These devices will reroute the noise to the better hearing ear, making listening even more difficult. Rerouting devices also do not provide benefit for localization (Agterberg et al., 2019; Bagatto et al., 2019; Snapp et al., 2017). This is unsurprising considering that only one ear is being stimulated and the head is not being used to provide potential benefits of level and timing cues. Research directly comparing rerouting devices to CIs in individuals with UHL is sparse, but suggests that rerouting devices are not equivalent to the benefits provided from a CI (Arndt et al., 2011). These devices cannot fully replicate the advantages of hearing with two ears, such as improved sound localization and better speech perception in noise.

There is no doubt that a remote microphone is beneficial for children with any degree of hearing loss, including UHL (Bagatto et al., 2019; Griffin et al., 2022; Picou, Davis, et al., 2020; Picou, Lewis, et al., 2020). They should be considered a part of a child's care to provide better access to speech. However, they are tools that do not deliver bilateral hearing. Preferential seating could offer some benefit in a classroom situation by providing an advantageous signal-to-noise ratio (Griffin et al., 2022). However, this is dependent on the better hearing ear remaining oriented toward the teacher and protected from noise, which is not likely a realistic situation in today's busy classrooms. Both remote mic systems and preferential seating can offer benefit in a classroom, but neither afford bilateral input or treatment of the hearing loss during all waking hours. Remote microphones and preferential seating can be used in conjunction with CIs to provide a more comprehensive treatment by providing bilateral hearing and maximizing a child's auditory capabilities.

MYTH 3: A CI DOESN'T HELP AND INTERFERES WITH THE "GOOD" EAR

It is reasonable to be concerned that children with UHL who use a CI would have difficulty fusing the two signals. There are inevitable differences in level, pitch, timing, and sound quality between the ears. However, bimodal listening (use of a hearing aid on one ear and a CI on the other) is a common recommendation in children with asymmetric hearing loss. Studies of this population have not shown evidence of binaural interference (Ching et al., 2001; Hartling et al., 2020). In the limited research investigating cochlear implantation in children with UHL, findings have suggested a lack of clinical interference (Arndt et al., 2011; Deep et al., 2021). Instead, the CI complements the hearing in the non-implanted ear, resulting in better binaural hearing abilities. CIs can provide the opportunity for binaural hearing, enhancing speech understanding, sound localization, and overall auditory abilities (Arras et al., 2021; Benchetrit et al., 2021; Brown et al., 2022; Ehrmann-Mueller et al., 2020; Lee et al., 2020; Park, Dillon, et al., 2023; Thomas et al., 2017).

MYTH 4: KIDS WITH UHL DO NOT LIKE THEIR CIS AND MOST BECOME NON-USERS

Non-use is a concern for those caring for children with CIs. The surgery and presence of the internal device are not without risk (O'Donoghue et al., 2002). Parents and clinicians rightfully want to provide implants to children who will use them. Thankfully, research has demonstrated that children with UHL who receive CIs generally have high rates of device use and positive experiences (Arras et al., 2022; Brown et al., 2022; Deep et al., 2021; Ehrmann-Mueller et al., 2020; Ganek et al., 2020; Park, Gagnon, et al., 2023; Polonenko et al., 2017; Zeitler et al., 2019). However, there is variability in device use for this population (Benchetrit et al., 2021; Gordon et al., 2023; Park, Gagnon, et al., 2023; Thomas et al., 2017). Teens tend to use their devices less often than younger children, which may be attributable to social concerns that come with adolescence (Park, Gagnon, et al., 2023; Thomas et al., 2017). Adequate counseling and ongoing support play crucial roles in promoting successful device use, particularly since device use has been correlated without outcomes in children with UHL (Lee et al., 2020; Park, Gagnon, et al., 2023).

MYTH 5: CHILDREN WITH SSD WHO ARE OVER A CERTAIN AGE OR HAVE A SPECIFIC NUMBER OF YEARS OF HEARING LOSS ARE NOT CANDIDATES

While some smaller scale studies have hinted at limiting CIs for very young children with UHL (Cushing et al., 2022; Rauch et al., 2021), the current research does not support rejecting children from candidacy based solely on age or duration of hearing loss (Benchetrit et al., 2021; Cohen & Svirsky, 2019; Nassiri et al., 2022; Park, Gagnon, et al., 2023; Távora-Vieira et al., 2013; Zeitler et al., 2019, 2023). Duration of hearing loss may impact outcomes (Park, Gagnon, et al., 2023), but benefit can be largely subjective. Research suggests that children with UHL, regardless of age or duration of hearing loss, can benefit from CIs and achieve significant improvements in speech perception, sound localization, and overall quality of life (Arndt et al., 2015; Park, Gagnon, et al., 2023; Thomas et al., 2017; Zeitler et al., 2019, 2023). Each child should be evaluated on an individual basis, considering their unique needs, functional limitations, and potential for auditory benefit. Age and duration of hearing loss are factors to consider but should not be seen as strict exclusion criteria. Counseling families and imparting realistic expectations for children with longer durations of hearing loss will help them make informed decisions about their child's hearing healthcare.

MYTH 6: CHILDREN HAVE TO MEET ALL OF THE FDA CRITERIA BEFORE THEY CAN BE REFERRED

The U.S. Food and Drug Administration (FDA) investigates hearing devices for safety and effectiveness within a predefined set of candidacy criteria. Once a device is available, physicians may opt to use them outside of the approved guidelines if they are well informed about the product and if they feel it is in the patient's best interest (*Understanding Unapproved Use of Approved Drugs "Off Label" | FDA*, n.d.). When it comes to CIs, clinicians and researchers have long been calling for expansion of indications and pointing out the limitations the current guidelines put on children who are deaf and hard of hearing (Anne et al., 2022; Brown & Gifford, 2021; Leigh et al., 2016; Park, Gagnon, et al., 2021; Varadarajan et al., 2021; Warner-Czyz et al., 2022). Large academic institutions frequently provide CIs to children off-label (Carlson et al., 2018). When it comes to UHL, research is pointing to good outcomes in children receiving CIs under the currently approved age of 5 years old (Arras et al., 2021, 2022; Brown et al., 2022; Park, Dillon, et al., 2023; Rauch et al., 2021). Given the early plasticity of the auditory system (Kral et al., 2013; Lee et al., 2020; Sharma et al., 2016; Vanderauwera et al., 2020) and the fact that younger children use their CIs more than older children (Park, Gagnon, et al., 2023,) it is reasonable to assume that outcomes may be better in children who receive a CI under the age of 5. However, research is ongoing and in cases of UHL, children who do not meet the strict FDA criteria for a CI can be referred to an experienced CI team for consideration of cochlear implantation.

CONCLUSION

Debunking the myths surrounding CIs for children with UHL is crucial to promote informed decision-making. General research findings have pointed to a need for intervention in children with UHL that provides bilateral access to sound. The only currently available technology that can do this is a CI. Studies suggest that binaural interference is not a concern, and that most children with UHL do accept and use their CIs. The currently approved FDA criteria are limiting; however, a knowledgeable CI team can evaluate a child with UHL and recommend the best course of intervention while considering individual needs. Cochlear implantation in children with UHL has been shown to provide substantial benefits. While CIs are not a standalone solution, it should be considered as part of a comprehensive treatment plan to maximize a child's listening and communication abilities and improve their quality of life.

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AUDICIÓN, HABLA Y LENGUAJE EN BEBÉS Y NIÑOS PEQUEÑOS NACIDOS PREMATURAMENTE

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En todo el mundo, 15 millones de bebés «nacen demasiado pronto» cada año (Blencowe et al., 2012; Howson et al., 2013). Sorprendentemente, las tasas de supervivencia de los bebés nacidos prematuramente superan ya el 93 % en todos los países. Más de 360 000 bebés prematuros sobrevivieron en EE. UU. en el 2020: el 10 % de todos los nacimientos (Blencowe et al., 2012; Howson et al., 2013). Si bien se trata de un logro increíble, el nacimiento prematuro sitúa al bebé en desarrollo en una situación especialmente vulnerable de sufrir consecuencias adversas en el desarrollo a lo largo de toda su vida. Debido a la alza de las tasas de supervivencia, las dificultades o discapacidades del desarrollo como son: déficits auditivos, visuales, motores, cognitivos, del habla y de la lectoescritura (McCormick y Behrman, 2007) también han aumentado. Además de las consecuencias médicas y de desarrollo, el mayor coste de crianza de un niño prematuro, en comparación con otro nacido a término, supera los 134 000 USD, situándose en más de 60 000 millones anuales en dólares estadounidenses, ajustados al año 2023 (Mangham et al., 2009). En cambio, el coste de la intervención temprana es una «ganga» relativa: 1240 dólares por bebé prematuro ajustados al año 2023 (Clements et al., 2007). Es muy probable que la intervención temprana sea eficaz, basándose en la notable plasticidad del cerebro de un recién nacido (White et al., 2013).

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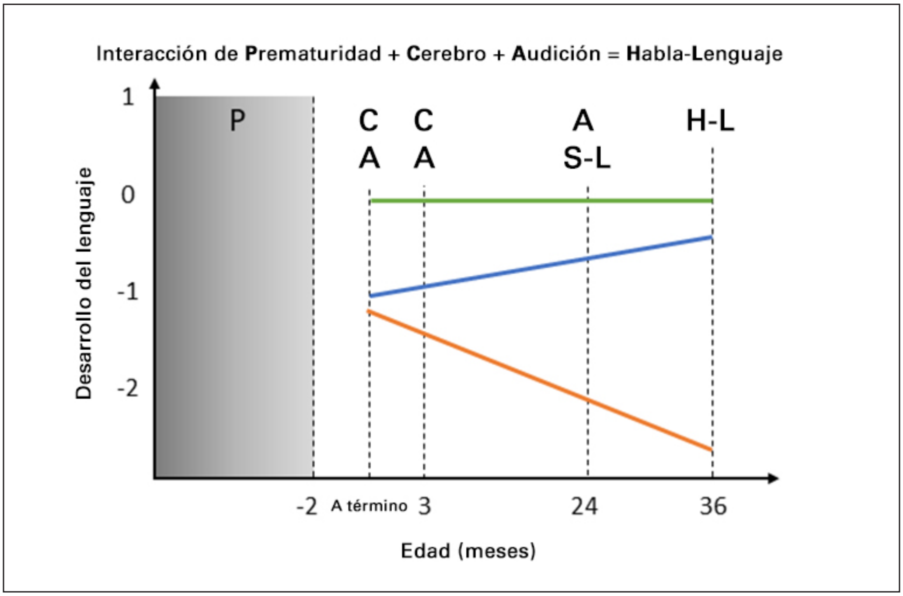
DESARROLLO DEL HABLA-LENGUAJE Y PREMATURIDAD

Los trastornos del habla y del lenguaje se encuentran entre los trastornos del neurodesarrollo más frecuentes en los niños, con tasas de prevalencia que oscilan entre el 5 % y el 8 % de los niños en edad preescolar (Law et al., 1998; Tomblin et al., 1997). En cambio, alrededor del 40 % de los bebés muy/extremadamente prematuros (≤32 semanas de edad gestacional) desarrollan trastornos del habla y del lenguaje (THL) persistentes (Duncan et al., 2012; Nguyen et al., 2018). Según las estimaciones de prevalencia, alrededor de 40 000 *bebés muy-extremadamente prematuros* (≤32 semanas de edad gestacional, en lo sucesivo denominados prematuros) desarrollan THL (Duncan et al., 2012; Nguyen et al., 2018) cada año en los Estados Unidos. La identificación precisa de THL en niños nacidos prematuramente no se suele producir hasta los 3-5 años de edad y prevalece en aproximadamente 2 de cada 3 niños (Roulstone et al., 2003), lo que les expone a un alto riesgo de obtener bajos resultados educativos, profesionales y sociales a

Figura 1. Trayectorias de desarrollo en la prematuridad (P), en relación con los resultados del habla-lenguaje y lectoescritura (S-L): las medidas incluyen Conectividad/procesamiento cerebral (B); Pérdida auditiva (H). Se muestran tres trayectorias: normal (línea verde); resiliente (línea azul); y alto riesgo de trastornos del habla y el lenguaje (línea naranja).



lo largo de su vida (Bashir y Scavuzzo, 1992; Young et al., 2002). Actualmente, no es posible predecir con exactitud en el primer año de vida qué niños desarrollarán THL. En consecuencia, la terapia logopédica es el servicio terapéutico más tardío y el menos frecuente de los que se prestan a los bebés tras el alta de la Unidad de Cuidados Intensivos Neonatales (UCIN) (Nwabara et al., 2017). Se pierde un tiempo precioso durante los tres primeros años, en los que la intervención es más eficaz, lo que se traduce en un aumento de los déficits en el habla, el lenguaje y la lectura en la edad escolar (Vohr et al., 2018; Vohr, 2016). Nuestro programa de investigación se basa en estudios longitudinales (Figura 1) con niños prematuros que muestran un desarrollo típico del lenguaje en un 32 %, un retraso que se resuelve con el tiempo en un 28 % y un retraso persistente o creciente en un 40 % (Duncan et al., 2012; Nguyen et al., 2018), que necesitan servicios de identificación e intervención tempranas.

PÉRDIDA AUDITIVA Y PREMATURIDAD

En general, el número de niños prematuros con algún grado de pérdida auditiva permanente es diez veces mayor que el de los niños nacidos a término (Hirvonen et al., 2018; Robertson et al., 2009), lo que agrava el impacto de las dificultades de comunicación (Hirvonen et al., 2018; Robertson et al., 2009). Las estimaciones de prevalencia de la pérdida auditiva varían ampliamente debido a criterios de diagnóstico y factores de riesgo variables, como lesiones cerebrales, infecciones prenatales, hiperbilirrubinemia y exposición a fármacos ototóxicos (Cristobal y Oghalai, 2008). El nacimiento prematuro también puede provocar una pérdida auditiva tardía o progresiva, lo que contribuye al desarrollo de THL (American Academy of Pediatrics, 2007). Estas pérdidas auditivas tardías y progresivas se pueden producir debido a problemas continuos relacionados con el nacimiento prematuro y estos diversos factores de riesgo (Joint Committee on Infant Hearing, 2019). Los métodos actuales de cribado auditivo neonatal solo detectan de forma fiable la pérdida auditiva superior a 30-40 dB HL, por lo que pasan por alto la mayoría de las pérdidas ligera a leve y de frecuencias agudas (Garinis et al., 2018). Incluso una pérdida auditiva leve y de frecuencias agudas aumenta el riesgo de desarrollar THL (Walker, Holte, et al., 2015; Walker, McCreery, et al., 2015). Estas pérdidas denominadas «mínimas» están asociadas a peores resultados lingüísticos

(Porter et al., 2013; Walker, McCreery, et al., 2015; Winiger et al., 2016) y son al menos tres veces más frecuentes que las pérdidas moderadas/mayores (Su y Chan, 2017; Vohr, 2016). En una gran cohorte de niños de primaria (n=1638), Moore y sus colegas informaron de que la prevalencia de la pérdida auditiva ligera a leve no diagnosticada (15-25 dB HL) era del 16,8 % y se asociaba significativamente con un lenguaje, una repetición de palabras y no palabras, y una percepción del habla en presencia de ruido más deficientes, en comparación con mejores niveles de audición (Moore et al., 2020). En los modelos estadísticos se demuestra que 15 dB HL es un criterio objetivamente adecuado para el diagnóstico de una pérdida auditiva significativa.

La pérdida auditiva leve y de frecuencias agudas que se pasa por alto con el cribado estándar se puede detectar mediante métodos fisiológicos más recientes, como las otoemisiones acústicas de productos de distorsión (OEAPD) (Blankenship et al., 2018) y los potenciales evocados auditivos del tronco cerebral (PEATC) con criterios más sensibles (Sininger et al., 2018). Nuestro objetivo es detectar estas formas más leves de pérdida auditiva y examinar su relación con el desarrollo del lenguaje en los niños prematuros.

DETECCIÓN TEMPRANA DE THL

En marcado contraste con el Cribado Auditivo Neonatal Universal, no existe un cribado universal del riesgo logopédico en recién nacidos. La lección fundamental del cribado neonatal es que la intervención es más eficaz *cuanto antes se inicie tras el nacimiento* (Joint Committee on Infant Hearing, 2013). Dado que en todos los estados y territorios de los EE. UU., así como en muchos otros países, ya existe una infraestructura de cribado neonatal para recién nacidos en las UCIN, sería totalmente viable emplear herramientas de cribado mejoradas antes del alta de la UCIN (American Academy of Pediatrics, 2007). Un mayor grado de lesión cerebral o de retardo de neuromaduración en las vías del lenguaje, sobre todo en presencia de pérdida auditiva o de un peor procesamiento del habla, limita la estimulación necesaria para favorecer el desarrollo del lenguaje. La estimulación del lenguaje enriquecida acústicamente en el primer año de vida es fundamental para el desarrollo del lenguaje típico y de las habilidades posteriores de lectoescritura (Nwabara et al., 2017; Vohr, 2016). Se ha demostrado que una mayor exposición al lenguaje está asociada a una mayor conectividad estructural, en concreto en la vía dorsal del lenguaje (Romeo, Leonard, et al., 2018; Romeo, Segaran, et al., 2018). La identificación de los niños con mayor riesgo cerca del momento del alta de la UCIN permitiría ofrecer una intervención temprana adecuada (es decir, una mayor exposición específica al lenguaje) antes de la edad preescolar.

DÉFICITS DEL HABLA-LENGUAJE Y PRELECTOESCRITURA EN NIÑOS PREMATUROS

Las pruebas de desarrollo en supervivientes de la UCIN con las Escalas de Evaluación del Desarrollo Infantil de Bayley, tercera edición, son el estándar clínico actual para guiar la intervención temprana en bebés prematuros (Anderson y Burnett, 2017). No obstante, existe la inquietud de que esta herramienta sobrestime la cognición y el lenguaje, y no sea predictiva del deterioro posterior en niños prematuros (Spencer-Smith et al., 2015). Por lo tanto, en nuestro estudio utilizamos dos evaluaciones específicas del lenguaje a la edad de 24 meses: MacArthur Child Development Inventory (MCDI, Inventario MacArthur de desarrollo infantil) y Communication and Symbolic Behavior Scales (CSBS, Escala de comunicación y comportamiento simbólico). La CSBS es una evaluación estandarizada directa especialmente adecuada para la población de prematuros, ya que evalúa los gestos y otros tipos de comunicación no verbal, lo que permite obtener una serie de resultados en niños con retraso en el uso de palabras. Anticipamos que la CSBS será sensible a las variaciones en las habilidades comunicativas no captadas por los subtests de lenguaje de Bayley-III. A los 36 meses de edad corregida (EC) se evalúan las habilidades del habla, el lenguaje y la lectoescritura mediante una batería estandarizada de pruebas del habla, el lenguaje y la prelectoescritura. Incluimos la evaluación de las habilidades de prelectoescritura en esta población para identificar los factores precursores de los déficits de lectura posteriores que se sabe que ocurren con frecuencia en niños en edad escolar con antecedentes de prematuridad (Allotey et al., 2018; Borchers et al., 2019; Kovachy et al., 2015; Lee et al., 2011).

MEDIDAS AUDITIVAS SENSIBLES

Recientemente hemos informado de la mejora del diagnóstico de la pérdida auditiva ligera a mayor al nacer mediante el desarrollo de normas adecuadas a la edad en bebés sanos y prematuros (Blankenship et al., 2018; Hunter et al., 2018). En este estudio prospectivo y longitudinal de 279 bebés, se verificó el estado auditivo a los 9 meses de EC con Audiometría por refuerzo visual (ARV). Aproximadamente al mes de EC, se midieron los OEAPD, la absorbancia de banda ancha (WBA) y los PEATC utilizando estímulos de ráfaga de tonos de diagnóstico de conducción aérea y ósea (0,5-4 kHz). Se analizó la eficacia de los niveles de OEAPD para clasificar los oídos como normales o hipoacúsicos. Encontramos una sensibilidad del 100 % y una especificidad del 76 % en la detección de la pérdida auditiva ligera-leve y mayor utilizando algoritmos normativos específicos para cada edad, validados frente al patrón de referencia tanto del umbral de PEATC como de la audiometría conductual (Blankenship et al., 2018). En este estudio, el 10 % de los bebés prematuros no superaron el cribado neonatal estándar y se detectó que el 30 % presentaba una pérdida auditiva leve o mayor. En otro estudio multicéntrico reciente sobre PEATC, informamos de que, utilizando técnicas de PEATC mejoradas, pudimos detectar una pérdida auditiva leve o mayor (Sininger et al., 2018). Actualmente estamos empleando estas técnicas mejoradas para determinar con precisión el grado de todos los niveles de audición, evaluando la prevalencia de la pérdida auditiva en niños prematuros de 3 a 36 meses de EC en un nuevo estudio longitudinal de niños muy prematuros.

Métodos

En este informe preliminar se describe un estudio de cohorte longitudinal basado en la población de bebés muy prematuros y extremadamente prematuros reclutados poco después del nacimiento en cinco UCIN de Cincinnati. Estos hospitales atienden a más del 90 % de la población muy prematura del suroeste de Ohio. Inscribimos a un total de 375 niños: 150 bebés al nacer, además de 125 niños de 24 a 36 meses, todos ellos nacidos con una edad gestacional ≤32 semanas. Se excluyeron los niños con afecciones cromosómicas o congénitas conocidas que afectan al sistema nervioso central, porque los resultados son uniformemente bajos en estos casos. También se excluyeron los niños que eran demasiado inestables desde el punto de vista médico para someterse a una RM a la edad equivalente a término. Debido a las medidas de resultados lingüísticos, se excluyeron las familias que no hablaban principalmente el idioma inglés. Recopilamos variables clínicas perinatales que se sabe o se sospecha que están asociadas con lesiones cerebrales, desarrollo anómalo o déficits cognitivos (Achenbach et al., 1993; Bapat et al., 2014; Dyet et al., 2006). Se recopiló una gran cantidad de datos sobre la madre y el bebé (Parikh et al., 2013), además de las afecciones que se sabe que están asociadas a la pérdida auditiva, como el paladar hendido o el síndrome de Down.

Medidas auditivas

Se realizaron evaluaciones audiológicas integrales a los 1-5 meses de EC y a los 2-3 años de EC utilizando medidas apropiadas para la edad. En ambas edades se evaluó la función del oído medio con absorbancia de banda ancha y se utilizaron OEAPD para evaluar la función del oído interno. A la edad de 3 meses de EC, se estimaron los niveles de audición mediante PEATC, obtenidos durante el sueño natural utilizando auriculares de inserción. Los umbrales de PEATC se utilizaron para clasificar el estado auditivo como audición típica (NH; 0-15 dB eHL) o pérdida auditiva (HL; ≥16 dB eHL). A la edad de 2-3 años de EC, el protocolo de audiología incluía ARV y/o audiometría condicionada por juego (ACJ) utilizando auriculares para obtener la detección del habla específica del oído y los niveles de respuesta mínimos (NRM) de tonos puros. Los NRM se utilizaron para clasificar el estado auditivo en audición típica (NH; 0-20 dB HL) o pérdida auditiva (HL; ≥25 dB HL).

Resultados del habla-lenguaje

A los 24-30 meses de EC, las familias cumplimentan el MCDI (Palabras y gestos) y la CSBS. El MCDI, un informe para progenitores validado y estandarizado sobre el desarrollo temprano del lenguaje y que se utiliza ampliamente en estudios de niños pequeños, permite identificar a preescolares con trastornos del lenguaje (Skarakis-Doyle et al., 2009) y se ha utilizado con niños de 24 meses para predecir resultados de lenguaje posteriores (Goodwin et al., 2002). La CSBS tiene buenas propiedades psicométricas (McCathren et al., 2000) y se ha demostrado que en niños de 2 años predice el vocabulario expresivo un año después (Skarakis-Doyle et al., 2009). La CSBS se graba en video y se puntúa por parte del personal del estudio entrenado con doble puntuación para el 20 % de las evaluaciones en cuanto a fiabilidad entre evaluadores e intraevaluadores utilizando la kappa de Cohen. El progenitor/cuidador también cumplimenta el StimQ (cuestionario sobre el entorno familiar), el Dialog Parent Report (cuestionario sobre hábitos de lectura) y el Screen Exposure Parent Report (Informe de progenitores sobre la exposición a pantallas) (Horowitz-Kraus et al., 2017; Hutton et al., 2019; Hutton et al., 2018).

A los 36 meses de EC se evalúan las habilidades de articulación de los niños mediante el Goldman-Fristoe Test of Articulation-3 (Prueba de articulación-3 de Goldman-Fristoe). El Goldman-Fristoe-3 es sensible a la producción fricativa/africada que representa un reto para algunos bebés con pérdida auditiva (Moeller et al., 2007). Las habilidades lingüísticas orales se evalúan mediante el Test of Early Language Development-III (TELD, Prueba de desarrollo temprano del lenguaje-III), que es sensible a los retrasos lingüísticos en niños prematuros (Monteiro-Luperi et al., 2016). Un progenitor/cuidador también cumplimenta el MCDI (Palabras y frases). Las aptitudes de prelectoescritura se evalúan mediante las subpruebas de Combinación de sonidos, letras-palabras y escritura de Woodcock-Johnson-IV Tests of Early Cognitive and Academic Development (Pruebas Woodcock-Johnson-IV de desarrollo cognitivo y académico temprano), con las que se evalúan la conciencia fonológica y el conocimiento temprano del alfabeto/palabras y la escritura.

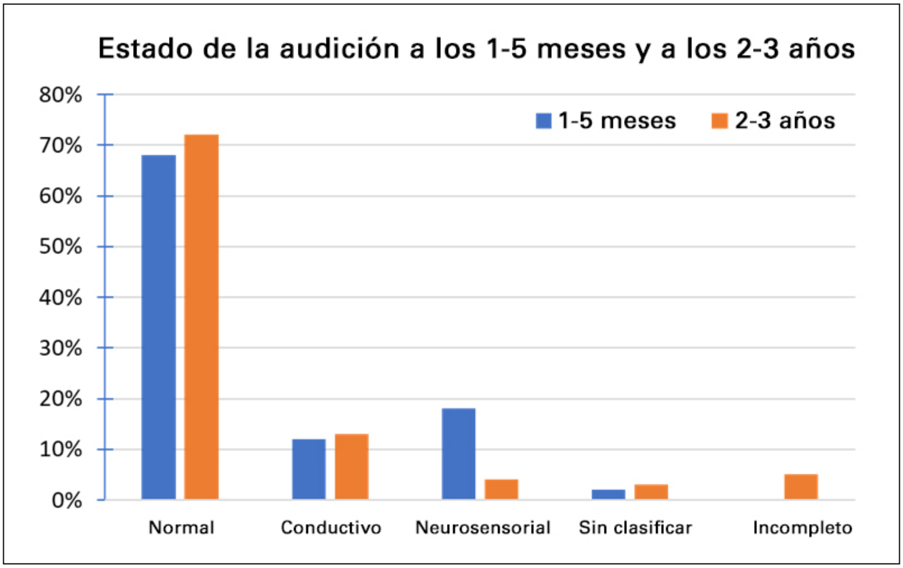
Resultados

En este análisis preliminar, 150 recién nacidos muy/extremadamente prematuros fueron evaluados con RM en la EC a término. La puntuación global de anomalías cerebrales, o Índice Kidokoro, se evaluó a partir de las RM (Brouwer et al., 2017). Esta medida es un combinación de cuatro mediciones regionales de RM que incluyen las puntuaciones de la materia gris cortical, la materia blanca total, la materia gris nuclear profunda y el cerebelo. Además, evaluamos a una cohorte de prematuros de más edad: 128 niños evaluados a los 2-3 años de edad con la batería completa de pruebas audiológicas (media de 30 meses de EC, rango de 24-42 meses).

Medidas auditivas

Se realizaron PEATC a 133 bebés prematuros a los 3 meses de edad EC (*rango*: 1-5 meses). Los resultados mostraron un elevado porcentaje de oídos infantiles con pérdida auditiva (38 %), con un grado de pérdida que oscilaba entre ligera y moderadamente grave. La mayoría de los

Figura 2. Prevalencia de la pérdida auditiva por tipo de pérdida a los 1-5 meses y a los 2-3 años, en una cohorte de niños muy prematuros y extremadamente prematuros.

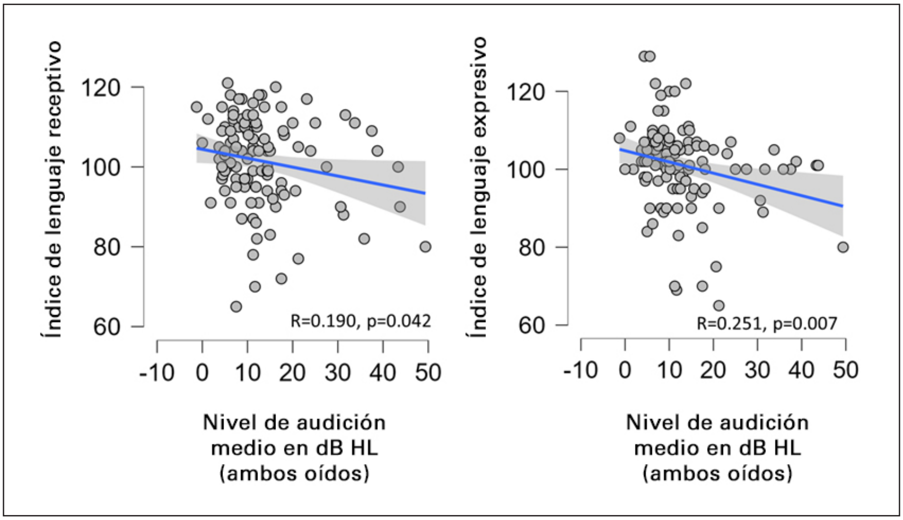


oídos presentaban una pérdida neurosensorial (18 %), con un porcentaje menor de pérdida conductiva (12 %) y un 2 % con un tipo indeterminado de pérdida. Los resultados del protocolo de pruebas audiológicas se muestran en la Figura 2. A los 2-3 meses de edad, el 32 % tenía pérdida auditiva en uno o en ambos oídos; el 12 % era conductiva, el 18 % neurosensorial y el 2% no estaba clasificada. A la edad de 2-3 años, el protocolo de pruebas conductuales tuvo un gran éxito cuando se incluyeron los resultados repetidos, lo que dio lugar a niveles de audición específicos del oído en el 88 % de los niños (*n*=113 sobre 128), mientras que 11 (9 %) solo presentaban NRM de campo sonoro y el 3 % no mostraban respuestas fiables. A los 2-3 años de edad, la audición era anómala en uno o en ambos oídos en el 20 % de los casos. La audición por conducción ósea y/o la timpanometría mostraron un componente del oído medio en el 13 %, mientras que el 4 % era neurosensorial (timpanometría normal y/o conducción ósea elevada) y el 3 % no se pudo clasificar. De los casos aparentes de hipoacusia neurosensorial, todos, menos uno, habían superado el cribado auditivo neonatal.

Resultados del habla y el lenguaje

Las puntuaciones estándar del TELD-III para las subescalas de lenguaje receptivo y expresivo a los 3 años de EC se correlacionaron con

Figura 3. Correlaciones entre el nivel de audición medio y las puntuaciones estándar de lenguaje (Media=100, *DT=15*) en la Prueba de desarrollo temprano del lenguaje en niños de 3 años. La audición se midió a los 2-3 años de edad.

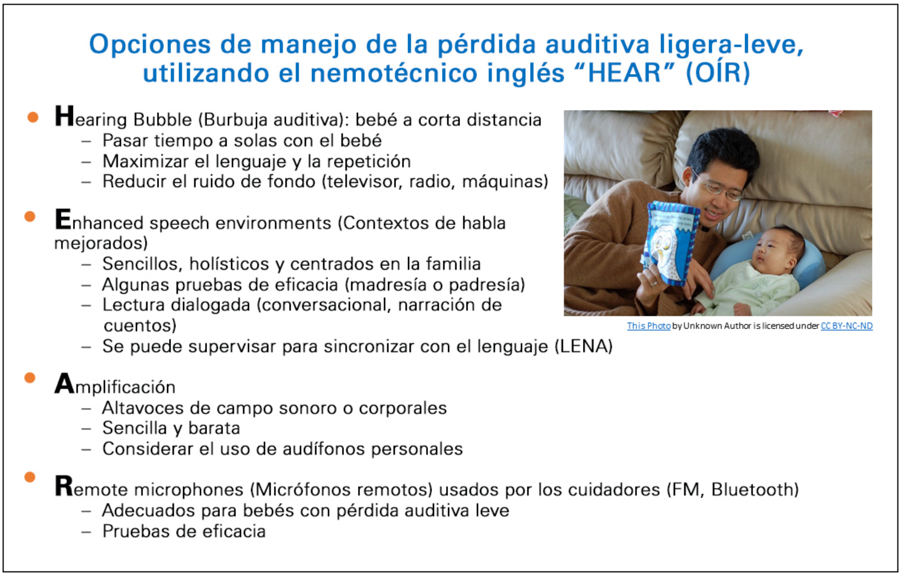


los niveles medios de audición (1-8 kHz) para ambos oídos combinados de los niños de 2-3 años de EC. Tal como se muestra en la Figura 3, se observó una amplia gama de puntuaciones, desde muy bajas hasta por encima de la media, tanto para el lenguaje receptivo como para el expresivo. Las puntuaciones lingüísticas medias tendían a descender con niveles de audición promedio más bajos. En concreto, las puntuaciones lingüísticas superiores a la media solo se encontraron en niños con niveles de audición excelentes (mejores de 20 dB HL). También había un grupo de puntuaciones lingüísticas muy por debajo de la media a pesar de los excelentes niveles de audición, por lo que es evidente que existen otros factores en juego en esta cohorte de niños prematuros. El análisis con medidas de RM, EEG y RMm se encuentra en curso y es de esperar que arroje luz sobre los factores neurológicos alterados que son predictivos de los resultados del lenguaje, utilizando enfoques de aprendizaje profundo para mejorar la predicción de los resultados (He et al., 2020; Li et al., 2018).

MÉTODOS DE INTERVENCIÓN

Los entornos lingüísticos tempranos y las aportaciones de los cuidadores desempeñan un papel fundamental en el desarrollo lingüístico, académico y cognitivo de los niños (Cartmill et al., 2013; Kuhl, 2010). En el caso de la pérdida auditiva leve-moderada, existen varias opciones de intervención viables, tal como se indica en la Figura 4. Para facilitar la consulta, se han organizado utilizando el nemotécnico «HEAR» (OÍR). El primer concepto importante que deben tener en cuenta los padres es la necesidad de comunicación dentro de la «burbuja auditiva» del bebé, que idealmente se encuentra entre 1 metro y 1,5 metros (3 y 4 pies) de distancia. El habla del cuidador será más audible y clara dentro de este rango y se verá menos afectada por el ruido de fondo que puede afectar a la percepción del habla (Leibold et al., 2016). A mayor distancia, el volumen y la claridad del habla disminuyen, sobre todo en las frecuencias altas, lo que afecta a la recepción de las consonantes. Este rango se logra perfectamente realizando actividades individuales con el bebé que también estimulen el lenguaje, en concreto la lectura de cuentos. Los bebés se benefician de una gran estimulación del lenguaje, por lo que se puede enseñar a los cuidadores a que faciliten una conversación descriptiva siempre que se encuentren cerca del bebé, hablando de todo lo que hagan, nombrando objetos mientras los señalan y cantando. Siempre que sea posible, es preferible eliminar las fuentes innecesarias de ruido, especialmente los aparatos electrónicos como el televisor, la

Figura 4. Recomendaciones HEAR: opciones de manejo de la pérdida auditiva leve que se pueden aplicar desde la infancia.



Opciones de manejo de la pérdida auditiva ligera-leve, utilizando el nemotécnico inglés "HEAR" (OÍR)

- **H**earing Bubble (Burbuja auditiva): bebé a corta distancia
  - Pasar tiempo a solas con el bebé
  - Maximizar el lenguaje y la repetición
  - Reducir el ruido de fondo (televisor, radio, máquinas)
- **E**nhanced speech environments (Contextos de habla mejorados)
  - Sencillos, holísticos y centrados en la familia
  - Algunas pruebas de eficacia (madresía o padresía)
  - Lectura dialogada (conversacional, narración de cuentos)
  - Se puede supervisar para sincronizar con el lenguaje (LENA)
- **A**mplificación
  - Altavoces de campo sonoro o corporales
  - Sencilla y barata
  - Considerar el uso de audífonos personales
- **R**emote microphones (Micrófonos remotos) usados por los cuidadores (FM, Bluetooth)
  - Adecuados para bebés con pérdida auditiva leve
  - Pruebas de eficacia

This Photo by Unknown Author is licensed under CC BY-NC-ND radio, los electrodomésticos y las máquinas. Si bien las máquinas que producen ruido son muy populares entre los padres para favorecer el sueño, son potencialmente perjudiciales para el desarrollo auditivo, ya que producen ruidos necesarios y sin sentido, por lo que se desaconsejan. Se ha demostrado que las terapias de estimulación en la UCIN, como el método canguro y los programas de interacción madre-hijo, mejoran los resultados cognitivos de los niños prematuros en la infancia (Benzies et al., 2013; Puthussery et al., 2018).

Los entornos del habla mejorados que sean naturales y se basen en el juego con cuidadores humanos proporcionan importantes «señales lingüísticas incidentales» que sirven para mejorar el desarrollo del lenguaje de los niños (Hart y Risley, 1980) e incluso pueden acelerar el aumento del lenguaje en niños prematuros con lesiones cerebrales (Rowe et al., 2009). La calidad y la cantidad del lenguaje real, en lugar del habla electrónica, como dibujos animados u otras grabaciones, es preferible sobre todo en el caso de los niños con pérdida auditiva (Ambrose et al., 2014). Para desarrollar el lenguaje y la socialización, lo mejor es siempre la interacción con las personas. El «lenguaje infantil o maternés» característico que la mayoría de los padres utilizan de forma natural es universal en todas las lenguas y favorece el desarrollo del lenguaje mediante la repetición, el énfasis en las señales prosódicas, las conexiones emocionales y el apoyo al desarrollo de la teoría de la mente (Ferjan Ramirez et al., 2020). Un enfoque de intervención conocido como «lectura dialogada» consiste en formar a los cuidadores para que lean a los bebés y niños utilizando libros de cuentos compartidos para crear conversaciones (comunicación recíproca) con ellos. La lectura dialogada se asocia a una conectividad neuronal centrada en la atención (Farah et al., 2019) y tiene efectos positivos tanto en la lectoescritura como en las habilidades lingüísticas generales.

La amplificación es eficaz para mejorar los resultados lingüísticos en la pérdida auditiva leve (Tomblin et al., 2014), si bien es poco frecuente que se recomiende el uso regular de audífonos en estos casos debido a la falta de pruebas existentes (McKay et al., 2008). Dado que en estos casos no está tan claro que el bebé no oiga bien, la pérdida auditiva ligera-leve es más invisible que la pérdida auditiva moderada-profunda. En estos casos, los padres pueden estar menos dispuestos a invertir los recursos económicos y de tiempo necesarios para obtener y utilizar audífonos. Sin embargo, está demostrado que los niños con pérdida auditiva leve pueden estar tan atrasados como los niños con pérdida auditiva más severa, probablemente porque no reciben ninguna intervención. Por lo tanto, siempre merece la pena hablar de los audífonos y de la disposición de los padres a probarlos. Es importante explicar que los resultados serán sutiles y que se puede tardar en apreciar los beneficios. Un enfoque alternativo es el uso de sistemas de micrófono remoto (SMR) para reducir los ruidos extraños y llevar la voz de los padres a la burbuja auditiva, incluso a distancia. Un estudio realizado con el programa informático de análisis del entorno lingüístico (LENA) demostró que, con el uso de SMR, los niños podían tener acceso a aproximadamente un 42 % más de palabras al día (Benitez-Barrera et al., 2018); la cantidad de habla dirigida al niño también aumenta con el uso de SMR (Benitez-Barrera et al., 2019). Además, los cuidadores informaron de que percibían positivamente los beneficios de comunicación de los SMR y tendían a hablar más a distancia cuando los utilizaban (Benitez-Barrera et al., 2018). Las tecnologías de micrófono remoto son especialmente beneficiosas cuando los niños van a la guardería, a preescolar o a la escuela primaria, y pueden aportar ventajas sustanciales a los niños con pérdida auditiva ligera-leve.

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FATIGA RELACIONADA CON LA ESCUCHA EN ALUMNOS CON SORDERA O HIPOACUSIA: COMPRENDIENDO Y GESTIONANDO EL «FACTOR FATIGA»

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La fatiga se suele definir como un estado de ánimo, una sensación subjetiva de cansancio o agotamiento que reduce nuestro deseo de seguir trabajando en una tarea (Bess et al., 2020; Hockey, 2013). La fatiga es omnipresente, algo que la mayoría de adultos y niños experimentan cuando se esfuerzan por realizar una tarea prolongada o difícil. La mayoría de las veces, todo lo que se necesita para recuperarse de la fatiga es un breve descanso o el cese de la tarea agotadora. En estos casos, los efectos negativos de la fatiga son mínimos. No obstante, en el ocaso de algunos individuos, las experiencias que generan fatiga pueden ser más severas y sostenidas en el tiempo. Cuando es recurrente, este tipo de fatiga severa puede tener importantes consecuencias negativas, especialmente para los niños. La fatiga severa es más frecuente en niños que padecen problemas crónicos de salud, como lo son el cáncer, la diabetes y la obesidad (Hornsby et al., 2017; Hockenberry-Eaton et al., 1999). Los niños con estas afecciones, que también sufren de fatiga significativa, tienden a enfrentarse a dificultades adicionales académicas y sociales en comparación con sus pares no fatigados (Stoff et al., 1989; Berrin, et al., 2007; Ravid et al., 2009). Pero, ¿qué tiene esto que ver con los niños con sordera o hipoacusia? Cada vez hay más pruebas que sugieren que los niños con sordera o hipoacusia son más proclives a experimentar fatiga severa como resultado de las dificultades para escuchar y comprender, esto se conoce con el término acuñado como fatiga relacionada con la escucha.

FATIGA EN LOS ALUMNOS CON SORDERAO HIPOACUSIA

Tanto los padres como los maestros hace tiempo que observan signos y síntomas relacionados con la fatiga en niños con sordera o hipoacusia,

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atribuyéndolos al mayor esfuerzo que ellos deben realizar cuando escuchan e intentan comunicarse con otras personas. Estas preocupaciones han dado lugar a que el tema de la fatiga relacionada con la escucha actualmente lidere los estudios de investigación en ciencias de la audición. Para conocer mejor la experiencia de fatiga en los niños con sordera o hipoacusia, nuestro grupo de investigación realizó una serie de entrevistas y grupos de discusión con alumnos con sordera o hipoacusia, sus padres y maestros (Davis et al., 2021).

En estos intercambios, aprendimos más sobre las experiencias de fatiga de los niños con sordera e hipoacusia, qué situaciones consideran agotadoras y cómo se recuperan del agotamiento. Es posible que no resulte ninguna sorpresa para aquellos profesionales que se dedican a la salud auditiva que el escuchar en presencia de ruido de fondo sea una de las situaciones auditivas más retadoras y agotadoras. La escucha en situaciones grupales (a diferencia de la escucha en situaciones uno a uno), en entornos acústicamente difíciles como el comedor escolar, al aire libre cuando se practican deportes y a través de dispositivos multimedia (p. ej., el teléfono) también resulta agotadora. Las experiencias de fatiga de los participantes fueron amplias y variadas, e incluyeron manifestaciones físicas, emocionales, sociales y cognitivas. Por ejemplo, se notificaron problemas físicos como dolores de cabeza, sensación de agotamiento y la necesidad de dormir una siesta o descansar después de experiencias de escucha sostenida. Los niños también manifestaron sentir una amplia gama de emociones que aparentemente se encuentran relacionadas con la fatiga, tales como la frustración, el estrés o el enfado. Los adultos (padres y maestros), también observan comportamientos sociales y emocionales que ellos asocian con la fatiga de los niños, como comportarse mal o retraerse en determinadas situaciones. Sumado a esto, cuando las demandas auditivas son altas, los niños con sordera o hipoacusia son más proclives a "desconectarse" o a experimentar más dificultades para mantener la concentración en una determinada tarea en comparación con los niños sin pérdida auditiva (fatiga cognitiva). Algunos niños con sordera o hipoacusia notificaron que «su cerebro se sentía cansado» después de tener que hacer un esfuerzo para escuchar y comprender en la escuela o en situaciones sociales. Toda esta información nos ayudó a comprender mejor la fatiga relacionada con la escucha en esta población y fue la base del desarrollo de un marco teórico que resalte los retos a los que se enfrentan los alumnos con sordera o hipoacusia (Davis et al., 2021). En la Tabla 1 se

Tabla 1: Determinantes situacionales y experiencias de fatiga observadas por niños con sordera o hipoacusia, sus padres y maestros. Modificado a partir del marco teórico de la fatiga relacionada con la escucha que se puede encontrar en Davis et al., 2021.

¿Qué precede/causa la fatiga relacionada con la escucha?	¿Qué se siente cuando se está fatigado?
Respuestas de los participantes: • Situaciones de escucha en un contexto ruidoso • Entornos grupales con muchas personas hablando • Escucha en el comedor, el gimnasio, durante los acontecimientos deportivos • Cuando me siento presionado para escuchar o desempeñarme bien en una situación • Cuando tengo que hacer muchas cosas a la vez (escuchar y realizar otra tarea) • Los maestros observaron que los alumnos con discapacidades adicionales, con una mayor motivación para escuchar o con tecnología auditiva averiada parecen más fatigados	**Respuestas de los participantes:** • Experiencias físicas: ○ Agotado ○ Cansado ○ Exhausto ○ Me da dolor de cabeza ○ Necesito descansar • Experiencias cognitivas: ○ Me desconecto o no presto atención ○ Es difícil concentrarse o centrarse ○ Mi cerebro “se siente cansado” ○ Me pongo a soñar despierto • Experiencias socioemocionales ○ Me siento frustrado o enfadado ○ Me rindo o dejo de esforzarme ○ Evito situaciones o me aparto de otras personas ○ Los maestros y los padres observaban un aumento de los problemas de conducta o mal comportamiento cuando el niño estaba fatigado

muestra una versión modificada de este marco teórico y se destacan algunos de los determinantes situacionales importantes de la fatiga relacionada con la escucha (es decir, las situaciones que provocan fatiga) en niños con sordera o hipoacusia, y algunas de las características físicas, socioemocionales y cognitivas más comunes de la fatiga experimentada. Para obtener una descripción completa de estos y otros factores, véase Davis et al. (2021).

VERSIONES PEDIÁTRICAS DE LAS ESCALAS DE FATIGA DE VANDERBILT: LAS VFS-PEDS

Los resultados de estos grupos de discusión y entrevistas confirmaron informes anecdóticos previos e investigaciones anteriores que sugerían que la fatiga relacionada con la escucha era un problema importante para al menos algunos niños con sordera o hipoacusia. Los resultados del estudio resaltaron el impacto de la fatiga relacionada con la escucha en estos niños y la necesidad de contar con una herramienta sensible y fiable para medir el constructo. Los abundantes datos cualitativos obtenidos de los grupos de discusión y las entrevistas nos facilitaron la información fundamental necesaria para crear las versiones pediátricas de las Escalas de Fatiga de Vanderbilt: las VFS-Peds. Las VFS-Peds consisten en un conjunto de tres cuestionarios diseñados para evaluar la fatiga relacionada con la escucha a largo plazo (es decir, la fatiga experimentada durante una semana típica) en población que enfrenta problemas de comunicación (p. ej., hipoacusia o sordera, trastornos del habla y el lenguaje, etc.). Las VFS-Peds constan de una escala de autoinforme para el niño (VFS-C) y de escalas de informes para padres y maestros (VFS-P y VFS-T, respectivamente). La VFS-C y la VFS-T proporcionan una medida única y unidimensional de la fatiga auditiva de un niño. En cambio, la VFS-P es multidimensional y proporciona medidas separadas de la fatiga mental y física. Se puede encontrar una descripción detallada del desarrollo y la validación de las VFS-Peds en Hornsby et al. (2022). En resumen, citas y comentarios mencionados en los grupos de discusión y en las entrevistas se utilizaron para crear un amplio conjunto de posibles ítems para evaluar la fatiga relacionada con la escucha que fueran relevantes para el grupo de encuestados objetivos (es decir, niños, padres o maestros). Por ejemplo, un padre refirió que su hija había ido de visita a un museo y que «el guía era estupendo pero hablaba tan rápido que ella [una niña con pérdida auditiva] no podía seguir las explicaciones. Cuando en el entorno hay un gran bullicio, lo cierto es que tiene muchos problemas. Debe hacer un gran esfuerzo y termina por agotarse». Esta y otras observaciones similares se transformaron en el ítem de la VFS-P: *Escuchar supone un gran esfuerzo para mi hijo*. Una vez desarrollado un amplio conjunto de posibles ítems para el test, se recopilaron datos de varios centenares de niños con sordera o hipoacusia, sus padres y maestros. Estos datos se analizaron para identificar ítems relevantes y de alta calidad para su uso en las VFS-Peds. El resultado fue una escala de medición relevante de alta calidad (es decir, válida, fiable y sensible) de la fatiga relacionada con la escucha (Hornsby et al., 2022).

Además de la versión original en inglés de las VFS-Peds, varios grupos de investigación han elaborado, o se encuentran elaborando, traducciones cultural y lingüísticamente apropiadas. Hasta la fecha, existen versiones traducidas de las VFS-Peds en mandarín, italiano y español (estadouni-dense/castellano). En nuestro sitio web (https://www.vumc.org/vfs) se pueden encontrar copias de las VFS-Peds en inglés, sus traducciones, y una lista de las colaboraciones en curso para traducir las escalas a otros idiomas.

USO DE LAS ESCALAS VFS-PEDS

Consúltese la Guía del usuario de VFS-Peds (https://www.vumc.org/vfs) para obtener una descripción detallada de las pautas de administración y puntuación. En todas las escalas se utiliza un sencillo formato de respuesta de frecuencia de tipo Likert de cinco puntos (0-4) para cuantificar la frecuencia con la que se produce una situación o una experiencia relacionada con la fatiga en una semana normal. Entre las opciones de respuesta se incluyen Nunca (0), Casi nunca (1), A veces (2), A menudo (3) y Casi siempre (4). Utilizando las VFS-Peds, la fatiga de un niño se puede cuantificar de dos maneras:

1. Utilizando la Teoría de la Respuesta al Ítem (TRI) para calcular una puntuación de escala TRI. Este enfoque proporciona la estimación más precisa de la experiencia de fatiga de un individuo, pero se requieren programas informáticos y algoritmos específicos para su cálculo, y es el más adecuado para el análisis de datos grupales con fines de investigación.
2. Las respuestas de los participantes también se pueden sumar simplemente para obtener una puntuación de fatiga total (en el caso de la VFS-C y la VFS-T) o dos puntuaciones de subescala (puntuaciones de fatiga mental y física) en el caso de la VFS-P. Las puntuaciones individuales totales o de subescala se pueden comparar con las puntuaciones de una muestra de estandarización de niños con un desarrollo normal, tal como se describe en nuestro manual del usuario (https://www.vumc.org/vfs). Este enfoque simple de suma de puntuaciones se recomienda para fines clínicos.

Siempre que sea posible, recomendamos que se administre el conjunto completo de las Escalas de Fatiga de Vanderbilt para comprender mejor cómo afecta (si es que afecta) la fatiga relacionada con la escucha a los alumnos con sordera o hipoacusia. La VFS-C es una medida de autoinforme que completa el alumno, por lo que proporciona información sobre la fatiga del niño desde su propia perspectiva. No obstante, no todos los niños podrán facilitar respuestas fiables utilizando la VFS-C. Del mismo modo, los resultados de nuestros grupos de discusión y entrevistas sugieren que algunos niños con una pérdida auditiva prolongada podrían no ser conscientes de que las dificultades auditivas, ni de que la fatiga resultante experimentada es atípica y pueden subestimar el impacto de esta fatiga. En estos casos, los padres y/o maestros de los alumnos con sordera o hipoacusia pueden identificar mejor las áreas de interés. La VFS-P es una medida de informe de proximidad, cumplimentada por el progenitor/cuidador del niño, que plantea preguntas sobre las respuestas del niño después de la jornada escolar sobre la fatiga relacionada con la escucha. La VFS-T también es una medida de informe de proximidad y solo debe cumplimentarla un maestro/proveedor de servicios que tenga un conocimiento directo de los comportamientos del alumno en una variedad de entornos. Son escasos los trabajos en los que se examina la concordancia entre las medidas del autoinforme y de los informes de proximidad de las VFS-Peds; no obstante, en la investigación en la que se utilizan escalas de fatiga genéricas se sugiere que es probable que existan algunas variaciones entre los encuestados objetivo (p. ej., Hornsby et al., 2017). Dado que los progenitores y los maestros observan al niño en entornos muy diferentes, sus impresiones de la frecuencia y la magnitud de los problemas relacionados con la fatiga pueden variar y todas podrían ser válidas. Por estas razones, recomendamos que se recopilen respuestas de las tres fuentes (alumno, progenitores y maestros) siempre que sea posible. La identificación de problemas significativos relacionados con la fatiga por parte de cualquier encuestado podría justificar un seguimiento adicional. Las VFS-Peds se pueden descargar e imprimir para su cumplimentación en papel. Además, las versiones en inglés de las escalas se pueden descargar como archivos electrónicos para su cumplimentación y puntuación mediante una computadora o una tableta.

Según nuestra experiencia, la mayoría de los padres y maestros pueden leer las instrucciones de las VFS y cumplimentar las escalas con una orientación mínima en menos de 5 minutos. Es posible que los niños que cumplimenten la escala de autoinforme necesiten una orientación adicional en función de su edad y sus capacidades cognitivas. A continuación comentamos técnicas adicionales para administrar la VFS-C a los niños y para mejorar la información clínica proporcionada por las VFS-Peds.

Consejos para administrar la VFS-C

- En nuestra experiencia, los niños menores de diez años pueden tener dificultades para conceptualizar la fatiga. Le recomendamos que verifique que el niño comprende los conceptos y la tarea antes de administrarle la VFS-C. Preguntas del tipo «¿qué significa la fatiga?» o «¿qué sientes cuando experimentas fatiga?» pueden servir para empezar.
- Lea las instrucciones y el cuestionario en voz alta al niño. Compruebe que entiende las opciones de respuesta (nunca, casi nunca, a veces, a menudo, casi siempre) antes de empezar.
    - **IMPORTANTE:** Si observa que el niño no comprende el concepto de fatiga, la tarea de rellenar el cuestionario o las opciones de respuesta disponibles, interrumpa la administración.
- El niño debe rellenar la escala en un entorno tranquilo, utilizando su tecnología de asistencia auditiva (si procede).
- Durante la administración de la VFS-C, haga preguntas de sondeo. Puede obtener información significativa del alumno mientras hablan sobre las preguntas. Por ejemplo, si un niño responde «casi siempre» a una pregunta, pídale que facilite un ejemplo («¿dónde te sucede?», «¿te ocurre todos los días en la escuela?») o simplemente puede decir «cuéntame más». Las respuestas que obtenga pueden ayudar en la orientación de estrategias de intervención y tratamiento importantes. En la sección siguiente de Estudios de casos se ejemplifica el concepto de utilizar entrevistas junto con la puntuación de las VFS-Peds.

ESTUDIOS DE CASOS DE VFS-PEDS

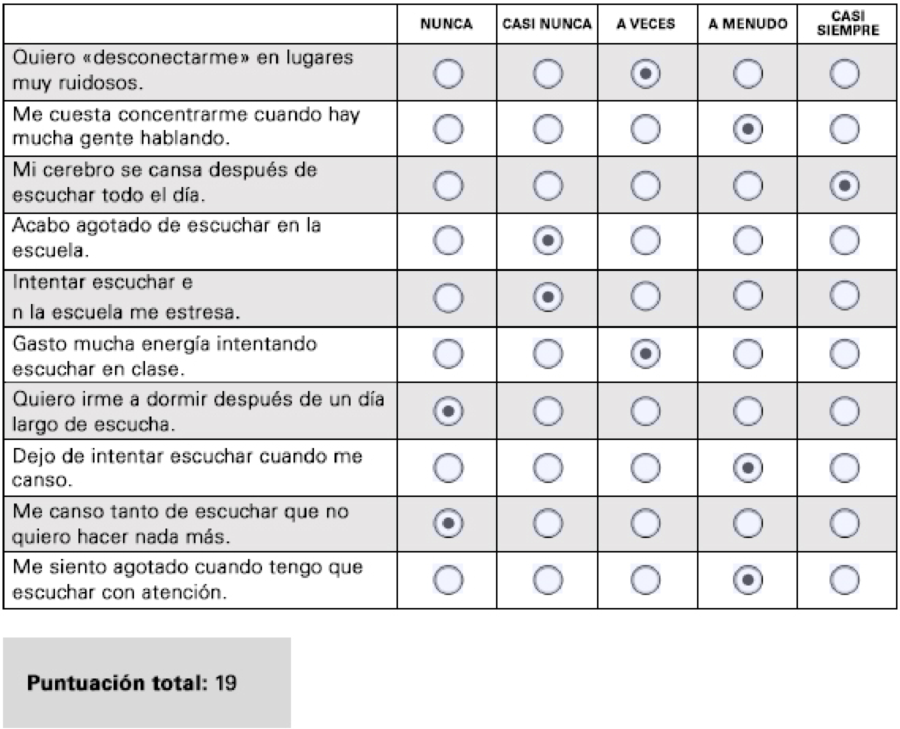
Las VFS-Peds se administran de forma rutinaria a padres, alumnos y maestros como parte de la batería de evaluación con fines de elegibilidad y planificación de programas. Cuando la puntuación total de las VFS-Peds de un niño supera las normas de estandarización, proporciona una justificación cuantitativa para iniciar un seguimiento adicional. No obstante, la puntuación por sí sola no proporciona una orientación clara sobre el seguimiento que se debe realizar. Sumado a esto, los niños con un desarrollo típico también pueden experimentar problemas significativos relacionados con la fatiga, por lo que puede haber casos en los que la puntuación de fatiga de un niño no supere las normas de estandarización aunque el niño, el progenitor y/o el maestro hayan expresado su preocupación. En estas situaciones, hemos comprobado que las puntuaciones de las escalas se pueden combinar con las respuestas de los alumnos en las entrevistas para sacar a la luz problemas «invisibles». Este tipo de información nos ha permitido trabajar con educadores y cuidadores para comprender mejor las dificultades con la fatiga relacionada con la escucha que experimenta un niño y cómo trabajar de una manera más eficaz para remediarlas. Los estudios de casos que se exponen a continuación se basan en la experiencia del primer autor (H. Davis) y resaltan este enfoque.

Estudio de caso 1: Jacob

A principios de este curso escolar, una madre se puso en contacto conmigo porque le preocupaba la fatiga que experimentaba su hijo con hipoacusia. Su hijo tiene una pérdida auditiva neurosensorial bilateral de leve a moderada y utiliza audífonos. Se encuentra actualmente en sexto grado y había comenzado a mostrar dificultades en tareas académicas, lo que el equipo educativo definía como «falta de concentración» y «que no las comprendía». Realicé la VFS-P con la madre y la VFS-C con el alumno en una reunión virtual después de la jornada escolar. Las puntuaciones de la madre fueron 18 (sobre 28) en la subescala mental, lo que sugiere que los problemas relacionados con la fatiga mental eran relativamente frecuentes; y 10 (sobre 20) en la subescala física, lo que sugiere que los problemas relacionados con la fatiga física eran menos frecuentes. Si bien las puntuaciones globales eran bajas (es decir, no superaban las normas de estandarización), en dos ítems concretos la madre informó que el niño «casi siempre» 1) mostraba fatiga de escucha al final del día y «casi siempre» 2) «se rendía» en situaciones de escucha difíciles.

El niño cumplimentó la VFS-C, con una puntuación de 19 sobre 40, que se encuentra en el rango de fatiga baja según nuestro sistema de suma de puntuaciones, lo que sugiere que se produjeron algunos problemas relacionados con la fatiga, pero no de una manera frecuente en todas las situaciones. Sin embargo, en una revisión de sus respuestas individuales se observó que en el 40 % de los ítems de la VFS-C (4 sobre 10), el niño notificaba que las dificultades relacionadas con la fatiga

Figura 1. Respuestas en la VFS-C y puntuación total del Estudio de caso 1



	NUNCA	CASI NUNCA	A VECES	A MENUDO	CASI SIEMPRE
Quiero «desconectarme» en lugares muy ruidosos.	○	○	●	○	○
Me cuesta concentrarme cuando hay mucha gente hablando.	○	○	○	●	○
Mi cerebro se cansa después de escuchar todo el día.	○	○	○	○	●
Acabo agotado de escuchar en la escuela.	○	●	○	○	○
Intentar escuchar e n la escuela me estresa.	○	●	○	○	○
Gasto mucha energía intentando escuchar en clase.	○	○	●	○	○
Quiero irme a dormir después de un día largo de escucha.	●	○	○	○	○
Dejo de intentar escuchar cuando me canso.	○	○	○	●	○
Me canso tanto de escuchar que no quiero hacer nada más.	●	○	○	○	○
Me siento agotado cuando tengo que escuchar con atención.	○	○	○	●	○

Puntuación total: 19

ocurrían «a menudo» o «casi siempre» (véase la Figura 1). Indicaba que su cerebro estaba «casi siempre» cansado después de escuchar todo el día, que «a menudo» le costaba concentrarse cuando había mucha gente hablando, que «a menudo» dejaba de intentar escuchar cuando estaba cansado y que «a menudo» se sentía agotado cuando tenía que escuchar con atención.

Además de completar la VFS-C con el alumno, también le formulé preguntas de sondeo. Véanse a continuación algunos ejemplos de nuestra conversación.

> **Audiólogo:** ¿Qué significa que te desconectas?
>
> **Alumno:** Dejo de concentrarme, de mirar con atención, pero no miro nada en particular. Si estoy en una clase grande con mucha gente, a veces me ocurre.
>
> **Audiólogo:** ¿Por qué has dicho que «casi siempre» tienes dificultades para concentrarte en esta situación?

> **Alumno:** Cuando hay mucha gente hablando, me desconecto o no trato de escuchar lo que dicen. Me centro en el maestro, pero no en la discusión de la clase.
>
> **Audiólogo:** Cuéntame más cosas sobre lo que sientes cuando tu cerebro se cansa.
>
> **Alumno:** Si tengo clases todo el día, mi cerebro y mis oídos están cansados después de la jornada escolar. Depende de las clases; una lección muy larga puede ser más pesada.
>
> **Audiólogo:** ¿Qué quieres decir, qué gastas mucha energía intentando escuchar?
>
> **Alumno:** Gasto mucha energía; a veces, si hay mucha gente hablando y estoy muy cansado, trato de superarlo y eso consume mucha energía. Si sigo esforzándome tratando de escuchar, llega un momento en que me digo «no puedo más, no puedo más». Me siento muy cansado y quiero abandonar. Tengo esa sensación en la escuela, pero no en casa ni con mis amigos. Me canso de escuchar tanto y solo quiero dejar de escuchar y tomarme una pausa de escucha.

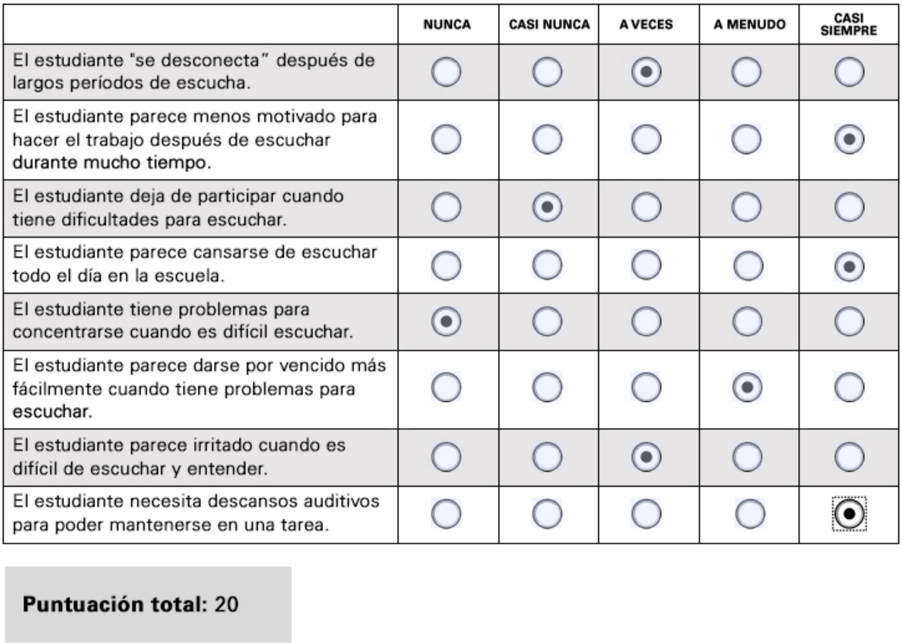
Esta información se envió al equipo educativo del alumno y la respuesta que recibí fue positiva. Si bien tenían conocimientos sobre la fatiga, consideraron que la información era muy reveladora y les ayudaba a comprender mejor las dificultades a las que se enfrentaba el alumno a diario. Basándose en parte en estos resultados, la escuela puso en marcha pausas de escucha y verificaciones con el alumno para apoyar mejor sus necesidades de escucha.

Estudio de caso 2: Miranda

Miranda es una alumna de 8 años que estudia tercer grado y tiene una pérdida auditiva bilateral profunda. Utiliza implantes cocleares y un sistema de micrófono remoto en la escuela. Como parte de una evaluación para su próxima reunión de reevaluación, se administraron la VFS-C y la VFS-T a Miranda y a su maestra, respectivamente.

La VFS-C se administró en un espacio tranquilo de la escuela primaria de Miranda. Intenté hablar del cansancio con Miranda, facilitándole ejemplos («a veces me siento cansada cuando tengo que hacer todas las tareas en la escuela; ¿te pasa esto alguna vez?»), pero la

Figura 2. Respuestas en la VFS-T y puntuación total del Estudio de caso 2



	NUNCA	CASI NUNCA	A VECES	A MENUDO	CASI SIEMPRE
El estudiante "se desconecta" después de largos períodos de escucha.	○	○	●	○	○
El estudiante parece menos motivado para hacer el trabajo después de escuchar durante mucho tiempo.	○	○	○	○	●
El estudiante deja de participar cuando tiene dificultades para escuchar.	○	●	○	○	○
El estudiante parece cansarse de escuchar todo el día en la escuela.	○	○	○	○	●
El estudiante tiene problemas para concentrarse cuando es difícil escuchar.	●	○	○	○	○
El estudiante parece darse por vencido más fácilmente cuando tiene problemas para escuchar.	○	○	○	●	○
El estudiante parece irritado cuando es difícil de escuchar y entender.	○	○	●	○	○
El estudiante necesita descansos auditivos para poder mantenerse en una tarea.	○	○	○	○	●

Puntuación total: 20

niña no podía entender el concepto. En ese momento, decidí dejar de recopilar datos formales a través de la VFS-C.

La VFS-T se envió por correo electrónico a su maestra, que la cumplimentó y la remitió (véase la Figura 2), junto con algunas notas sobre los progresos de Miranda en el aula. Indicó que Miranda lloraba a menudo al final de la jornada escolar y quería recostar la cabeza en el pupitre. La puntuación de 20 en la VFS-T se situaba en el rango bajo de fatiga; sin embargo, las respuestas de la maestra indicaban problemas sistemáticos con una reducción de la motivación después de situaciones auditivas difíciles, un agotamiento visible y la necesidad de pausas de escucha. Si bien no se refleja formalmente por escrito en su plan, la maestra me informó de que recientemente se le permitía a Miranda tomar un descanso sobre las 13:30 (es decir, levantarse del pupitre e ir a beber agua en el lugar situado al fondo del aula) para ayudarla a no desviarse de la tarea. La maestra notó una mejora en el comportamiento de Miranda por la tarde con este cambio reciente.

En la reunión de reevaluación, los resultados de la VFS-T se compartieron con el equipo educativo, incluida la familia de Miranda. El equipo llegó al acuerdo de que debía añadirse al PEI de Miranda una pausa de escucha. Se indicaron dos pausas: una al final del día antes de la última hora de clase en grupo, y otra después de los servicios de logopedia, ya que requerían un esfuerzo considerable por parte de Miranda.

CONTROL DE LA FATIGA RELACIONADA CON LA ESCUCHA EN LOS NIÑOS

La información y los estudios de casos anteriores resaltan el riesgo mayor que tienen los alumnos con sordera o hipoacusia de experimentar la fatiga relacionada con la escucha y el impacto que puede tener en un entorno escolar. Por desgracia, hasta la fecha no se han realizado investigaciones sistemáticas que respalden intervenciones o estrategias específicas para reducir las dificultades asociadas a la fatiga relacionada con la escucha. Le recomendamos que observe al alumno en diversos entornos a lo largo de la jornada escolar y que utilice las VFS-Peds, así como entrevistas de seguimiento, para identificar si experimenta problemas significativos. Las preguntas de sondeo durante la entrevista les podrán ayudar a usted y al equipo educativo del niño a diseñar un plan para minimizar su fatiga y todos los posibles efectos negativos. A falta de enfoques validados empíricamente, hemos utilizado la información facilitada por los participantes de nuestros grupos de discusión (niños, progenitores y maestros) y nuestras experiencias profesionales para crear una hoja resumen de recomendaciones de gestión que se puede descargar en nuestro sitio web. Entre los puntos clave de esta hoja resumen se incluyen:

- Tener en cuenta la acústica del aula/otros espacios de escucha y reducir el ruido de fondo en la medida de lo posible. En repetidas ocasiones se indicó que el ruido de fondo provocaba fatiga a los alumnos con sordera o hipoacusia.
- Al alumno se le debe asignar un asiento o un lugar flexible y preferente. Este sitio le proporcionará el mejor acceso visual y auditivo, y podrá ser necesario modificarlo en función de la actividad.
- Comprobar que la amplificación del alumno (p. ej., audífonos, implantes cocleares, dispositivos de anclaje óseo, sistemas de micrófono remoto) se ajusta a los objetivos prescritos y funciona correctamente.

- Proporcionar pausas de escucha programadas: tiempo en el que se permitirá al alumno tomarse un descanso de la escucha atenta en el aula, sobre todo después de un periodo de escucha difícil. Por ejemplo, hacer una breve pausa para moverse, como levantarse y estirarse, o ir al aseo o a beber agua. Alternativamente, el niño puede hacer una pausa en una determinada tarea permaneciendo en el pupitre y/o quitarse la tecnología de asistencia auditiva (p. ej., audífono, implante coclear) durante un breve periodo de tiempo. Si bien el uso de tiempo completo de la amplificación es la regla de oro, algunas personas han informado que retirarse los dispositivos durante una pausa breve es una medida eficaz para reducir la fatiga o acelerar la recuperación. Esta recomendación se individualiza en función de las necesidades del alumno y de su nivel de responsabilidad. En el caso de los padres, podrán permitir que el niño se tome un descanso o una siesta después de la jornada escolar, o evitar ocasionalmente eventos sociales por la tarde si ya se muestran cansados.
- Considerar la posibilidad de programar por la mañana las clases o las terapias que requieran una atención auditiva intensa. Muchos alumnos con sordera o hipoacusia afirman sentirse más cansados al final de la jornada escolar.
- Educar al niño, a sus padres y a sus maestros sobre la fatiga relacionada con la escucha y su relación con la pérdida auditiva y las dificultades de escucha. Esta educación puede mejorar su concienciación sobre la fatiga, los factores que la provocan, sus consecuencias y los posibles enfoques para minimizar sus efectos negativos.

Cabe destacar que, en comparación con los adultos, a los niños con sordera o hipoacusia se preocupan más por las barreras que les impidan utilizar estas estrategias, y mencionan la preocupación de meterse en problemas o de perderse contenidos importantes si no se mantienen conectados continuamente en situaciones de escucha. Incluir al alumno en el proceso de toma de decisiones puede ayudar a que entienda la manera de defender sus propios intereses y utilizar estas estrategias tanto en casa como en la escuela. Con su apoyo, estos alumnos pueden aprender más sobre sí mismos y sus necesidades en lo que se refiere a la fatiga relacionada con la escucha, facilitándoles oportunidades para superar con éxito esta experiencia.

Recursos

Las VFS-Peds y los recursos relacionados están disponibles de forma gratuita en nuestro sitio web: https://www.vumc.org/vfs/vanderbilt-fatigue-scales.

Contacto

Si desea plantear preguntas sobre este trabajo, póngase en contacto con Hilary Davis, AuD (hilary.davis@vumc.org) o Benjamin Hornsby, PhD (ben.hornsby@vumc.org).

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LA PÉRDIDA AUDITIVA EN EL SIGLO XXI Y MÁS ALLÁ: UNA NUEVA ERA DE PRECISIÓN DIAGNÓSTICA Y DE TRATAMIENTO MEDIANTE LA GENÓMICA

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En los últimos quince años nuestra comprensión de la audición y de la pérdida auditiva ha tenido progresos drásticos. Los científicos han trabajado para identificar docenas de genes implicados en la pérdida auditiva y así dilucidar la compleja maquinaria molecular responsable de la audición. Al mismo tiempo, se han producido rápidos avances en las herramientas de diagnóstico clínico, incluidas las pruebas de imagen y genéticas. Esto significa que, por primera vez, en la mayoría de los casos de los niños con pérdida auditiva, podemos obtener un diagnóstico.

LA PÉRDIDA AUDITIVA NO ES UN DIAGNÓSTICO

En realidad, la pérdida auditiva no es un diagnóstico. La pérdida auditiva es, de hecho, un síntoma de una diferencia subyacente en el sistema auditivo. El sistema auditivo es extraordinariamente complejo y existen centenares de diagnósticos posibles: desde ototoxicidad debida a fármacos aminoglucósidos hasta alteraciones moleculares del canal de mecanotransducción de las células ciliadas internas.

El diagnóstico es crucial para aquellos individuos con pérdida auditiva y sus familias, ya que proporciona información sobre el pronóstico (si la pérdida auditiva progresará con el paso del tiempo o se mantendrá estable), el riesgo de recurrencia (las posibilidades de tener otro hijo con pérdida auditiva) y otras características clínicas que pueden estar asociadas a la pérdida auditiva. Sin embargo, quizás lo más importante sea que el diagnóstico proporciona al individuo y a su familia un sentimiento de empoderamiento. No se trata solo de un niño con un síntoma (pérdida auditiva), sino de una persona con un diagnóstico.

https://doi.org/10.17955/tvr.123.1.symp3

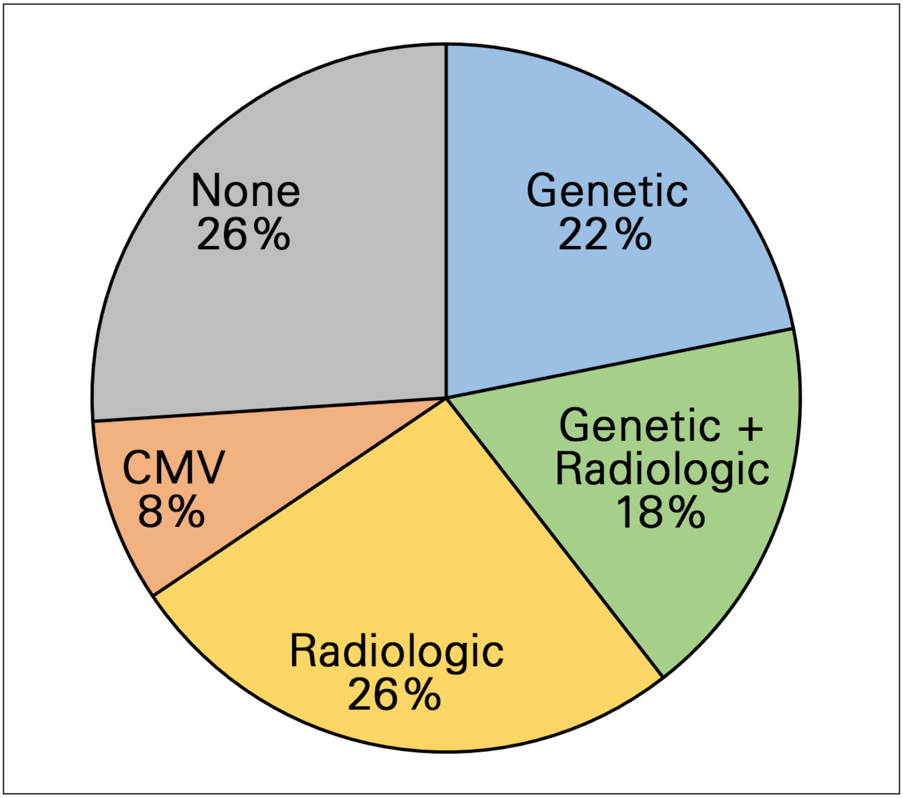
Existen centenares o incluso millares de diagnósticos que pueden dar lugar a la pérdida auditiva. Un diagnóstico proporciona un marco de referencia para el equipo terapéutico (médicos, audiólogos y logopedas) que puede utilizar en la prestación de servicios al paciente. Esto permite una verdadera práctica centrada en el paciente y no una intervención basada en únicamente un síntoma. Quizás una de las mejores formas de demostrar la importancia de un diagnóstico es que los niños y sus familias son más proclives a la utilización de audífonos o implantes cocleares porque se sienten más empoderados e implicados en su cuidado y asistencia (Brodie et al., 2022).

La comprensión del diagnóstico es fundamental para los profesionales que atienden a niños con pérdida auditiva, considerando la gran cantidad de nuevos tratamientos y decisiones terapéuticas disponibles. Hace décadas que los audífonos y los implantes cocleares están disponibles, pero la decisión de seguir adelante con un implante coclear puede resultar difícil de tomar. Si se diagnostica una pérdida auditiva progresiva (por ejemplo, debida a la mutación del gen *ACTG1*), la decisión puede ser diferente a que si se diagnostica una forma más estable de pérdida auditiva (por ejemplo, debida a la mutación del gen *MYH14*). Si se diagnostica una infección congénita por CMV (CMVc), puede estar indicado un tratamiento con valganciclovir y medicación antivírica. Actualmente, con el inicio de los ensayos clínicos de terapia génica para la pérdida auditiva (los cuales se describe más adelante), es fundamental disponer de un diagnóstico genético preciso. Un diagnóstico nos permite personalizar el cuidado y la asistencia del paciente con pérdida auditiva y de su familia porque, en lugar de tratar solo el síntoma, estamos tratando al paciente.

HERRAMIENTAS PARA EVALUAR LA PÉRDIDA AUDITIVA

Los avances en los estudios de imagen, las pruebas del citomegalovirus congénito y las pruebas genéticas resultan en que la mayoría de los niños con pérdidas auditivas de severas a profundas cuenten con un diagnóstico. En un estudio reciente de 119 niños que se encontraban en evaluación para recibir implantes cocleares por pérdida auditiva bilateral se mostró que el 22 % se identificó por causas genéticas, el 18% por causas genéticas y radiológicas, el 26% por causas radiológicas, y 8% por CMV (Kim y Choi, 2022). En total, el 74 % de los niños recibieron un

Figura 1. Rendimiento diagnóstico en 119 niños con pérdida auditiva bilateral severa-profunda. Se facilitó un diagnóstico al 74 % de los niños cuando se realizaron pruebas genéticas, pruebas de CMVc y RM. Adaptado de Kim et al., 2022.



diagnóstico cuando se les realizó una evaluación diagnóstica completa que incluía RM de alta resolución del hueso temporal, una prueba de CMV y las pruebas genéticas (Figura 1). Esta situación es radicalmente distinta a la de hace 20 años, cuando se concluía que la gran mayoría de los niños tenían pérdida auditiva «idiopática» tras lo que se consideraba una evaluación clínica completa (Preciado et al., 2004).

Imágenes de alta resolución

Las imágenes del hueso temporal deben ser de corte fino y de alta resolución. Los estudios de imágenes se pueden realizar mediante tomografía computarizada (TAC) y/o resonancia magnética (RM). Las ventajas del TAC son la vista detallada de la anatomía ósea, que puede ayudar en la planificación quirúrgica, y que la obtención de las imágenes suele tardar de 1 a 5 minutos. Sin embargo, una TAC expone a un niño a radiación, por lo que, si es posible, suele evitarse en los niños más pequeños. La ventaja de la RM es que se visualizan los tejidos blandos, incluido el nervio auditivo. La desventaja de la RM es que, en un protocolo típico de obtención de imágenes en el caso de la pérdida auditiva, los estudios de imagen pueden llegar a tardar de 30 a 45 minutos. Esto significa que el niño deberá permanecer quieto, dormido o sedado para someterse a una RM. En la actualidad, numerosas instituciones ofrecen un protocolo de «alimentación y envoltura» por el que se lleva a un bebé a la cita de RM con hambre, se le alimenta y, a continuación, se le «envuelve» firmemente con orejeras colocadas en su sitio. El objetivo es que el niño duerma de forma natural durante el procedimiento y, en la mayoría de los casos, estos estudios de imagen tienen éxito, con tasas superiores al 80 %. El rendimiento diagnóstico global del TAC y la RM es similar, con diferentes modalidades que proporcionan una mayor resolución de diferentes estructuras (Chen et al., 2014; Kachniarz et al., 2014). En general, la decisión sobre qué tipo de estudios de imagen solicitar puede variar en función de la edad del paciente, su capacidad de permanecer quieto durante la exploración, y del tipo y el grado de pérdida auditiva.

Pruebas de CMV congénito

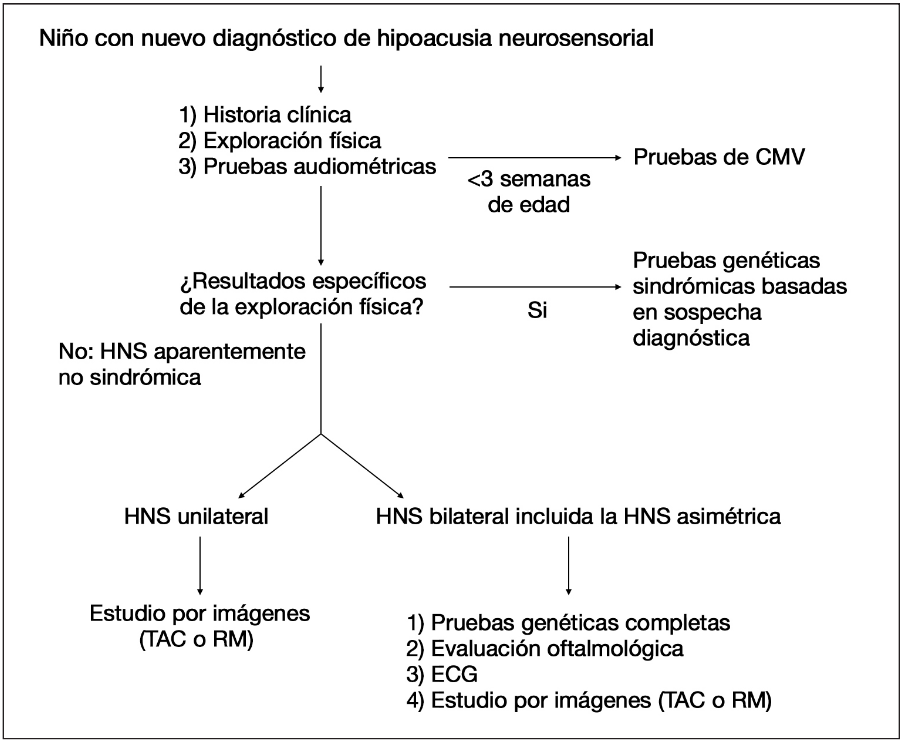
La infección por CMVc es la causa infecciosa de pérdida auditiva pediátrica más común pediátrica y afecta a cerca del 20 % de los niños con pérdida auditiva congénita. La infección por CMVc provoca un amplio espectro de síntomas, desde anomalías cerebrales severas, insuficiencia hepática y retraso en el desarrollo, a infecciones más leves que pueden incluir solo pérdida auditiva. Evaluar para CMVc es difícil porque el virus está presente de manera ubicua en nuestro entorno e idealmente, se debe evaluar a las 3 semanas de edad. Algunos estados de EE. UU. han reglamentado el realizar pruebas de detección del CMVc como parte de los protocolos de tamiz auditivo neonatal en el caso de que un niño no paso el cribado auditivo. Existe bastante presión por parte de la sociedad para que las pruebas del CMVc sean obligatorias en todo el país. La identificación de infecciones congénitas por CMVc es fundamental dado que se pueden tratar con medicación antivírica, ganciclovir y valganciclovir. Se ha demostrado que estos fármacos mejoran los resultados en niños gravemente afectados y también detienen, o a veces revierten, la pérdida auditiva en la mayoría de los niños (Lanzieri et al., 2023). Las pruebas de detección de CMVc y los estudios de imagen constituyen dos de los pilares de la evaluación diagnóstica de la pérdida auditiva pediátrica, junto con las pruebas genéticas.

PRUEBAS GENÉTICAS EN LA PÉRDIDA AUDITIVA

La pérdida auditiva genética es realmente compleja, demuestra una notable heterogeneidad genética y subraya la exquisita complejidad de la audición humana. Hasta la fecha, se han descubierto más de 120 genes causantes de pérdida auditiva no sindrómica (http://hereditaryhearingloss.org). Además, existen centenares de formas sindrómicas de pérdida auditiva. Cada uno de estos genes puede contener centenares de cambios genéticos (variaciones) causantes de la pérdida auditiva, Esto dificulta la evaluación genética para pérdida auditiva, ya que la realización de pruebas para un único gen no tiene eficacia debido al escaso rendimiento diagnóstico (la probabilidad de identificar una causa). No obstante, desde 2010, con la llegada de la nueva tecnología de secuenciación genómica, es posible realizar pruebas para múltiples, o incluso centenares, de genes.

Estos nuevos métodos de secuenciación del ADN, denominados secuenciación de nueva generación o secuenciación masiva en paralelo, proporcionan la capacidad de secuenciar todos los genes de la pérdida auditiva simultáneamente y se ha desarrollado un nuevo paradigma de pruebas genéticas para la pérdida auditiva basado en estos métodos. En lugar de analizar un único gen, se podrían analizar todos los genes de la pérdida auditiva a la vez, lo que aumentaría la velocidad y la eficacia de las pruebas genéticas para la pérdida auditiva (Shearer et al., 2010). Debido a su eficacia, las pruebas de panel genético comprehensivas se convirtieron rápidamente en el estándar de referencia en la evaluación de niños con pérdida auditiva (Shearer et al., 2015). Estos paneles de pruebas genéticas se utilizan actualmente de forma rutinaria en la evaluación de niños con pérdida auditiva, ya que proporcionan un diagnóstico el 40-60 % de las veces con una sola prueba (Downie et al., 2019; Sloan-Heggen et al., 2016). Estos paneles se pueden adquirir en varias empresas y difieren en función del número de genes incluidos, desde unos 100 a más de 200 genes. Normalmente, se incluyen todos los genes de pérdida auditiva no sindrómica, así como los genes más habituales implicados en las formas sindrómicas de pérdida auditiva.

Figura 2. Algoritmo de diagnóstico clínico para la evaluación de la hipoacusia neurosensorial (HNS) en niños.



Los genes de la pérdida auditiva sindrómica son importantes, ya que estos diagnósticos comprenden alrededor del 20 % de los diagnósticos en niños (Perry et al., 2022).

El uso de grandes paneles de genes para las pruebas genéticas de la pérdida auditiva se ha consolidado por su eficacia en docenas de estudios y miles de pacientes. Además, estos paneles son componentes clave de dos conjuntos de directrices creadas para la evaluación recomendada de niños con pérdida auditiva por el International Pediatric Otolaryngology Group (Liming et al., 2016) y el American College of Genetics and Genomics (Alford et al., 2014). En la Figura 2 se muestra un algoritmo propuesto para la evaluación diagnóstica que integra estas directrices y que se utiliza en mi práctica clínica. Este algoritmo incorpora pruebas de CMV, estudios de imagen y pruebas genéticas completas utilizando un panel de genes.

Se deben realizar pruebas genéticas a todos los niños con hipoacusia neurosensorial bilateral, incluida la hipoacusia asimétrica. Si existen rasgos sindrómicos, se deberán realizar pruebas genéticas específicas basadas en el diagnóstico sospechado; sin embargo, esto es poco frecuente y la mayoría de los niños presentarán una pérdida auditiva sospechada no sindrómica. En el caso de los niños con pérdida auditiva unilateral (de un solo lado), el estudio de imagen suele ser el de mayor rendimiento. No obstante, en un estudio reciente en el que utilizamos la secuenciación del exoma se demuestra que alrededor del 18 % de los niños con pérdida auditiva unilateral tendrán un diagnóstico genético y, lo que es más importante, el 55 % de estos niños tendrán un diagnóstico de una forma sindrómica de pérdida auditiva (Perry et al., 2022). Estos diagnósticos tienen una gran repercusión para estos pacientes y sugieren que deberíamos considerar la incorporación de pruebas genéticas de la pérdida auditiva unilateral en nuestra práctica clínica.

En los Estados Unidos, un médico puede solicitar pruebas genéticas completas para la pérdida auditiva. Las pruebas suelen realizarse a partir de hisopados bucales (mejilla), aunque en algunos casos puede ser necesaria una extracción de sangre. Existen varias empresas comerciales que ofrecen pruebas genéticas en los Estados Unidos y en el Registro de Pruebas Genéticas (Genetic Testing Registry) existe una lista actualizada disponible: https://www.ncbi.nlm.nih.gov/gtr/. Es importante destacar que las compañías de seguros reconocen actualmente la eficacia y la importancia de las pruebas genéticas para la pérdida auditiva en la asistencia clínica. Por ello, en la mayoría de los casos, la pérdida auditiva genética está cubierta por los seguros privados. Los datos de nuestra clínica muestran que los seguros privados cubren las pruebas genéticas completas mediante un panel de genes en aproximadamente el 80 % de los casos. La cobertura de los seguros públicos varía en función del estado y en algunos estados (p. ej., California) es sistemática. La cobertura de los seguros sigue mejorando a medida que las compañías reconocen la utilidad de estas pruebas para proporcionar un diagnóstico a los niños con pérdida auditiva.

EN EL HORIZONTE

Las pruebas genéticas para la pérdida auditiva son muy efectivas, pero no se puede establecer un diagnóstico en todos los casos. Las nuevas tecnologías, como la secuenciación del genoma completo y la secuenciación del genoma de lectura larga, mejorarán el rendimiento del diagnóstico. Los precios de estas tecnologías siguen disminuyendo, por lo que cada vez son más accesibles para la asistencia clínica. Las pruebas genéticas en la pérdida auditiva también demuestran una desigualdad racial inaceptable en el rendimiento diagnóstico: los niños negros e hispanos tienen cinco veces menos probabilidades de obtener un diagnóstico a partir de pruebas genéticas (Florentine et al., 2022). Esto se debe principalmente a la falta de enfoque del estudio y de pruebas para los niños de razas y etnias minoritarias. Está claro que hay que seguir trabajando en este ámbito para atender mejor a estos niños.

La mayoría de los niños con pérdida auditiva se identifican a través del cribado auditivo neonatal. En el año 2000 se elaboraron las directrices para el cribado auditivo universal y, en el año 2010, más del 98 % de los recién nacidos se sometieron al cribado auditivo en los Estados Unidos. Con estas directrices, el número de bebés con sordera o hipoacusia identificados en una fase temprana ha aumentado drásticamente. La identificación temprana de los niños con pérdida auditiva es fundamental para obtener los mejores resultados lingüísticos (Yoshinaga-Itano, et al. 1998; 2018). Está claro que el cribado auditivo neonatal ha sido eficaz en el objetivo de identificar en una fase temprana a un mayor número de niños con pérdida auditiva. No obstante, dado que la pérdida auditiva es, con diferencia, el trastorno más común identificado durante el cribado neonatal, es fundamental que sigamos trabajando para mejorar su eficacia. El cribado auditivo neonatal actual se basa en una medida fisiológica (otoemisiones acústicas o potenciales evocados auditivos del tronco cerebral), siendo económico y relativamente fácil de administrar. Este cribado fisiológico no detecta algunas formas leves de pérdida auditiva, no está diseñado para detectar la pérdida auditiva que se puede producir fuera del periodo neonatal y puede pasar por alto el trastorno del espectro de la neuropatía auditiva si se utilizan las otoemisiones acústicas en el cribado. Dada la eficacia de las pruebas genéticas en la pérdida auditiva, nosotros y otros profesionales hemos abogado por la incorporación del cribado genético en el programa de cribado auditivo neonatal (Shearer et al., 2019). En China y en otros países, el cribado genético ha mejorado las tasas de detección, ha reducido el tiempo hasta el diagnóstico y ha disminuido las tasas de pérdida durante el seguimiento (Guo et al., 2020); Wu et al., 2017). Está claro que la incorporación de la genética al cribado neonatal conllevaría obstáculos logísticos, pero la clave reside en proporcionar un diagnóstico temprano de la pérdida auditiva infantil.

Hace tiempo que la terapia génica en la pérdida auditiva es un objetivo de las personas con sordera o hipoacusia, de los científicos que estudian la pérdida auditiva y de los médicos que la tratan. Este objetivo está más cerca que nunca con los ensayos clínicos que se están iniciando en los EE. UU. y en Europa para la pérdida auditiva por mutaciones en el gen de la otoferlina (*OTOF*). Se eligió este gen para los ensayos clínicos iniciales porque, aunque se trata de una forma rara de pérdida auditiva genética, el mecanismo se conoce claramente, los componentes clave del oído interno están presentes y funcionan, y el tratamiento de ratones adultos con *OTOF* es eficaz. Los ensayos clínicos para otras formas de pérdida auditiva genética no tardarán en llegar en los próximos años. Es importante señalar que cualquier terapia génica requiere un diagnóstico genético preciso.

CONCLUSIÓN

En la última década se ha producido un cambio de paradigma en la evaluación y el tratamiento de los niños con pérdida auditiva. En la actualidad, la mayoría de los niños con pérdida auditiva bilateral pueden obtener un diagnóstico si se realiza un estudio diagnóstico completo. El diagnóstico de los niños con pérdida auditiva proporciona al paciente y al médico una información valiosa, y orienta el tratamiento de la pérdida auditiva, lo que permite al clínico tratar al niño y no solo el síntoma. Las nuevas tecnologías seguirán mejorando nuestro rendimiento diagnóstico, lo que se traducirá en una mejor comprensión de la pérdida auditiva.

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IMPLANTES COCLEARES PARA NIÑOS CON PÉRDIDA AUDITIVA UNILATERAL: DESMONTANDO LOS MITOS

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Los implantes cocleares (IC) han revolucionado el campo de la audiología, proporcionando una solución que cambia la vida de los niños con pérdida auditiva bilateral profunda. La ampliación de los criterios ha permitido que los niños con una pérdida auditiva unilateral (UHL, por sus siglas en inglés) significativa se puedan beneficiar también de esta tecnología. Sin embargo, esta práctica no está exenta de polémica. Aunque se tiene conocimiento desde 1984 que los niños con UHL se enfrentan a retos educativos (Bess y Tharpe, 1984, 1986), la espera vigilante hasta que el niño tenga la edad suficiente para que se le pueda colocar un dispositivo de enrutamiento contralateral de la señal (CROS, por sus siglas en inglés) ha sido el criterio imperante (Bagatto, 2020; Bagatto et al., 2019). Si bien la investigación ha demostrado en repetidas ocasiones los beneficios de la implantación coclear en esta población, esta práctica aún no se ha convertido en el estándar de atención. Como es inherente a las prácticas controvertidas, existen varios mitos y conceptos erróneos que rodean el uso de IC en niños con UHL, y éstos impregnan los sistemas clínicos de atención auditiva. Aquí, desacreditaremos estos mitos examinando los beneficios y las consideraciones basados en la evidencia asociados con el implante coclear en niños con UHL. A través de un análisis de la investigación actual y la evidencia clínica, destacaremos las ventajas potenciales de los IC como una opción de tratamiento viable, promoviendo la toma de decisiones informadas y mejorando la calidad de vida de los niños con UHL.

MITO 1: LOS NIÑOS CON SORDERA UNILATERAL «VAN A ESTAR BIEN» PORQUE TIENEN UN OÍDO CON EL QUE ESCUCHAN BIEN

Si bien es cierto que el acceso al sonido con un oído permite desarrollar el lenguaje hablado, los niños con UHL se enfrentan a numerosos retos que pueden impactar en su desempeño académico, sus habilidades de

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comunicación y su calidad de vida en general. La investigación ha demostrado que los niños con UHL experimentan dificultades que pueden iniciar en la infancia, dado que comienzan a mostrar retraso en el desarrollo auditivo antes, incluso, de que empiecen a hablar (Yang et al., 2020). Existen hallazgos consistentes que demuestran que la UHL repercute en la percepción del habla del niño (Davies et al., 2021; Park, Dillon, et al., 2023), en sus niveles de fatiga (Bess et al., 2020; Carpenter et al., 2022; Griffin et al., 2019; Sindhar et al., 2021), en el desarrollo del lenguaje (Fitzpatrick et al., 2019; Lieu et al., 2010; Lieu, 2013; Sangen et al., 2017), en la percepción del habla en presencia de ruido de fondo (Corbin et al., 2021; Ehrmann-Mueller et al., 2020; Griffin et al., 2019, 2020; Park, Dillon, et al., 2021, 2023; Reeder et al., 2015) y en la localización de los sonidos (Corbin et al., 2021; Ehrmann-Mueller et al., 2020; Park, Dillon, et al., 2023; Reeder et al., 2015), en comparación con sus pares con audición típica. Estos retos se pueden traducir en dificultades académicas (Lieu, 2013; McSweeny et al., 2021), déficits cognitivos (Lieu, 2018) y problemas socioemocionales. En estudios de investigación recientes se sugieren incluso cambios neuronales en los niños con UHL (Calmels et al., 2022; Shang et al., 2020; Sharma et al., 2016; Vanderauwera et al., 2020). Actualmente, no es aceptable asumir que un niño con UHL «va a estar bien». La investigación demuestra claramente que esta población presenta diferencias en su desarrollo y que se enfrenta a diversos retos.

MITO 2: UN CROS/BCHA/MICRÓFONO REMOTO/ ASIENTO PREFERENTE SERÁ SUFICIENTE

Si bien los dispositivos como los sistemas CROS, los audífonos de conducción ósea (BCHA), los micrófonos remotos o un asiento preferente pueden aportar algunas ventajas a los niños con UHL, no dejan de ser tecnologías de apoyo. Ninguno de estos dispositivos tiene por objetivo el tratar la pérdida auditiva, dado que no ofrecen la posibilidad de una audición bilateral. De hecho, los dispositivos de redireccionamiento como BCHA y CROS pueden ser perjudiciales en situaciones en las que la fuente de ruido se encuentra cercana al oído con peor audición (Bagatto et al., 2019; Choi et al., 2019; Griffin et al., 2022; Picou, Lewis, et al., 2020). Estos dispositivos redirigirán el ruido al oído con mejor audición, dificultando aún más la escucha. Los dispositivos de redireccionamiento tampoco aportan beneficios en la localización (Agterberg et al., 2019; Bagatto et al., 2019; Snapp et al., 2017). No puede resultar sorprendente si se considera que solo se estimula un oído y que la cabeza no se utiliza para obtener las ventajas potenciales de las diferencias temporales y de nivel sonoro. Los estudios de investigación en los que se comparan directamente los dispositivos de redireccionamiento con los IC en personas con UHL son escasos, pero sugieren que las ventajas que aportan los dispositivos de redireccionamiento no son equivalentes a las ventajas o beneficios que aportan los IC (Arndt et al., 2011). Estos dispositivos no pueden reproducir del todo replicar las ventajas de escuchar con dos oídos, como lo son una mejor localización de los sonidos y una mejor percepción del habla en presencia de ruido.

No hay duda de que los micrófonos remotos son beneficiosos para los niños con cualquier grado de pérdida auditiva, incluida la UNL (Bagatto et al., 2019; Griffin et al., 2022; Picou, Davis, et al., 2020; Picou, Lewis, et al., 2020). Se deben considerar parte de los servicios ofrecidos a un niño para que tenga un mejor acceso al habla. No obstante, son herramientas que no proporcionan una audición bilateral. Un asiento o lugar preferente pueden ofrecer alguna ventaja en el entorno de un aula al facilitar una relación señal-ruido ventajosa (Griffin et al., 2022). No obstante, dependerá de que el oído con mejor audición permanezca orientado hacia el maestro y protegido del ruido, lo que probablemente no sea una situación realista en las ajetreadas aulas de hoy en día. Tanto los sistemas de micrófono remoto como un asiento preferente pueden resultar beneficiosos en un aula, pero ninguno de ellos permite una acceso bilateral ni el tratamiento de la pérdida auditiva durante todas las horas de vigilia. Los micrófonos remotos y los asientos preferentes se pueden utilizar junto con los IC para ofrecer un tratamiento más completo al proporcionar audición bilateral y maximizar las capacidades auditivas del niño.

MITO 3: UN IC NO AYUDA E INTERFIERE CON EL OÍDO «BUENO»

La preocupación de que los niños con UHL que utilicen IC tengan dificultades para fusionar para las dos señales, es razonable. Existen diferencias inevitables de nivel, tono, tiempo y calidad del sonido, entre los oídos. Sin embargo, la audición bimodal (uso de un audífono en un oído y un IC en el otro) es una recomendación habitual en el caso de niños con una pérdida auditiva asimétrica. En los estudios realizados en esta población no se han detectado evidencias de que se produzca interferencia biaural (Ching et al., 2001; Hartling et al., 2020). En los estudios limitados que existen sobre implantes cocleares en niños con UHL, las conclusiones sugieren una ausencia de interferencia clínica (Arndt et al., 2011; Deep et al., 2021). Por el contrario, el IC complementa la audición del oído sin implante, lo que mejoraría la capacidad auditiva binaural. Los IC pueden ofrecer la oportunidad de una audición biaural, mejorando la comprensión del habla, la localización del sonido y las habilidades auditivas en general (Arras et al., 2021; Benchetrit et al., 2021; Brown et al., 2022; Ehrmann-Mueller et al., 2020; Lee et al., 2020; Park, Dillon, et al., 2023; Thomas et al., 2017).

MITO 4: A LOS NIÑOS CON PAU NO LES GUSTAN LOS IC Y LA MAYORÍA DEJA DE USARLOS

El no usar la tecnología es una preocupación constante de los profesionales que atienden a niños con IC. La cirugía y la presencia de un dispositivo interno no están exentas de riesgos (O'Donoghue et al., 2002). Los padres y los médicos desean, como es natural, proporcionar implantes a los niños que los vayan a utilizar. Afortunadamente, la investigación ha demostrado que los niños con UHL que reciben implantes cocleares suelen tener altos índices de uso de los dispositivos y experiencias positivas (Arras et al., 2022; Brown et al., 2022; Deep et al., 2021; Ehrmann-Mueller et al., 2020; Ganek et al., 2020; Park, Gagnon, et al., 2023; Polonenko et al., 2017; Zeitler et al., 2019). No obstante, existe una variabilidad en el uso de dispositivos en esta población (Benchetrit et al., 2021; Gordon et al., 2023; Park, Gagnon, et al., 2023; Thomas et al., 2017). Los adolescentes tienden a utilizar los dispositivos con menos frecuencia que los niños más pequeños, lo que se puede atribuir a las preocupaciones sociales que conlleva la adolescencia (Park, Gagnon, et al., 2023; Thomas et al., 2017). El asesoramiento adecuado y el apoyo continuo desempeñan un papel crucial a la hora de fomentar el uso de estos dispositivos, especialmente porque su uso se ha correlacionado con una ausencia de resultados en niños con PAU (Lee et al., 2020; Park, Gagnon, et al., 2023).

MITO 5: LOS NIÑOS CON SORDERA UNILATERAL QUE SUPERAN UNA DETERMINADA EDAD O PRESENTAN LA PÉRDIDA AUDITIVA DESDE UN NÚMERO CONCRETO DE AÑOS NO SON CANDIDATOS

Aunque en algunos estudios de menor escala se ha sugerido restringir los IC en el caso de niños muy pequeños con UHL (Cushing et al., 2022; Rauch et al., 2021), la investigación actual no respalda el rechazo de la candidatura a IC únicamente basada en la edad o en la duración de la pérdida auditiva (Benchetrit et al., 2021; Cohen & Svirsky, 2019; Nassiri et al., 2022; Park, Gagnon, et al., 2023; Távora-Vieira et al., 2013; Zeitler et al., 2019, 2023). La duración de la pérdida auditiva puede influir en los resultados (Park, Gagnon, et al., 2023), pero el beneficio puede ser en gran medida subjetivo. La investigación sugiere que los niños con UHL, independientemente de la edad o la duración de la pérdida auditiva, se pueden beneficiar de los IC y lograr mejoras significativas en la percepción del habla, la localización de los sonidos y la calidad de vida en general (Arndt et al., 2015; Park, Gagnon, et al., 2023; Thomas et al., 2017; Zeitler et al., 2019, 2023). A cada niño se le debe evaluar de forma individual, teniendo en cuenta sus necesidades únicas, sus limitaciones funcionales y su potencial de beneficio auditivo. La edad y la duración de la pérdida auditiva son factores a tener en cuenta, pero no se deben considerar criterios estrictos de exclusión. Aconsejar a las familias y transmitirles expectativas realistas en los casos de niños con una pérdida auditiva de larga duración les ayudará a tomar decisiones informadas sobre la asistencia sanitaria relacionada con la audición de sus hijos.

MITO 6: LOS NIÑOS DEBEN CUMPLIR TODOS LOS CRITERIOS DE LA FDA ANTES DE QUE SE LES PUEDA DERIVAR

La FDA (La “Food and Drug Administration” en Estados Unidos) investiga la seguridad y la eficacia de los dispositivos auditivos en el marco de unos criterios de candidatura predefinidos. Una vez que un dispositivo está disponible, los médicos pueden optar por utilizarlo fuera de las directrices aprobadas si están bien informados acerca del producto y si consideran que es lo mejor para el paciente (*Understanding Unapproved Use of Approved Drugs “Off Label” | FDA, s.f.*). En lo que respecta a los IC, los médicos e investigadores llevan mucho tiempo pidiendo que se amplíen las indicaciones y señalando las limitaciones que las directrices actuales imponen a los niños con sordera e hipoacusia (Anne et al., 2022; Brown & Gifford, 2021; Leigh et al., 2016; Park, Gagnon, et al., 2021; Varadarajan et al., 2021; Warner-Czyz et al., 2022). Las grandes instituciones académicas suelen proporcionar IC a los niños «fuera del uso aprobado» (Carlson et al., 2018). En lo que se refiere a la UHL, la investigación indica buenos resultados en el caso de los niños que reciben IC por debajo de la edad actualmente aprobada de 5 años (Arras et al., 2021, 2022; Brown et al., 2022; Park, Dillon, et al., 2023; Rauch et al., 2021). Dada la plasticidad temprana del sistema auditivo (Kral et al., 2013; Lee et al., 2020; Sharma et al., 2016; Vanderauwera et al., 2020) y el hecho de que los niños más pequeños utilizan los IC con más frecuencia que los niños mayores (Park, Gagnon, et al., 2023), es razonable suponer que los resultados pueden ser mejores en los niños que reciban un IC antes de cumplir 5 años. No obstante, la investigación sigue su curso y, en los casos de UHL, los niños que no cumplen los estrictos criterios de la FDA para recibir un IC pueden derivarse a un equipo experimentado en IC para que se considere la implantación coclear.

CONCLUSIÓN

Desmitificar las creencias que rodean a los IC en el caso de los niños con UHL es crucial para promover una toma de decisiones informada. Los resultados generales de la investigación apuntan a la necesidad de una intervención para niños con UHL que facilite el acceso bilateral al sonido. Actualmente, la única tecnología disponible con la que se puede lograr son los IC. La investigación sugiere que la interferencia biaural no es un problema y que la mayoría de los niños con UHL aceptan y utilizan los IC. Los criterios vigentes aprobados por la FDA son limitantes; no obstante, un equipo de IC bien informado puede evaluar a un niño con UHL y recomendar el mejor proceso de intervención teniendo en cuenta sus necesidades individuales. Se ha demostrado que la implantación coclear en niños con UHL proporciona beneficios sustanciales. Si bien los IC no son una solución independiente, se deben considerar parte de un plan de tratamiento integral para potenciar al máximo la capacidad de escucha y comunicación de los niños, y mejorar su calidad de vida.

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2022
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LISTENING AND SPOKEN LANGUAGE
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EXCELLING IN A TRANSFORMED WORLD

RESEARCH PROCEEDINGS

CONTENTS

Caregiver Coaching in Early Childhood Intervention

3

Dathan D. Rush, Ed.D., CCC-SLP | *Family, Infant and Preschool Program, J. Iverson Riddle Developmental Center*

Everyday life activities provide development-enhancing learning opportunities for young children. Since parents, teachers, child care providers, and other caregivers are with infants and toddlers more frequently and for longer periods of time than early childhood intervention practitioners, caregivers need to know how they can support their child's learning within the context of their interest-based activities in the home, community, and early childhood classroom. Coaching is an adult learning strategy and interaction style that may be used by early childhood intervention providers to build the competence and confidence of caregivers to use responsive strategies within these development-enhancing activities that will achieve their desired outcomes.

Preliminary Findings: Early Turn Talking is Associated with Later Social-Emotional Development

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Esteban Gômez Muzzio | *Fundación América por la Infancia*

Preliminary findings from a follow-up study of 33 children, assessed for their socioemotional development from 18 months of age, are presented. At 77 months, they were evaluated again in a laboratory context, recording on video a situation of discussion of a conflict with the caregiver and then coding these videos using the CIB instrument. Conversation turns assessed with the LENA software at 18 and 30 months of age are associated with socioemotional cognition, emotion regulation, and emotional communication competencies assessed at 77 months of age. The association is stronger at 30 months than at 18 months, suggesting a possible mediation effect to be explored in future analyses. Implications for professional intervention and public policies for children are discussed.

Foundations for Literacy: A Research-Based Early Reading Program that Improves Outcomes for Children who are Deaf and Hard of Hearing

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Amy R. Lederberg[1], Susan R. Easterbrooks[1], and Stacey L. Tucci[2] | *[1]Department of Learning Sciences, Georgia State University; [2]Training director for* Foundations for Literacy

One avenue for improving reading outcomes is to ensure children who are deaf or hard of hearing (DHH) enter school with the foundational skills needed to learn to read. Our research team developed an early literacy curriculum specifically for DHH children. Teachers use *Foundations for Literacy* (FFL) in a one-hour literacy block for the school year. Student learning objectives include improving spoken phonological awareness, alphabetic knowledge, word reading, vocabulary, and narrative. FFL is more systematic, and its instruction is more explicit, multi-modal, and intensive than might be used with children who have typical hearing. Much of the instruction is embedded in language-rich activities. Differentiation of instruction to the wide variation of language and phonological processing skills observed for children who are DHH is integral to the design. Results from multiple studies, including a randomized-control trial, indicates that FFL is an effective intervention for young DHH students.

The Road to Hearing Health Equity

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Susan D. Emmett, MD, MPH | *Center for Hearing Health Equity, Department of Otolaryngology-Head and Neck Surgery, College of Medicine, and Department of Epidemiology, Fay W. Boozman College of Public Health, University of Arkansas for Medical Sciences (UAMS)*

Of the 1.6 billion people with hearing loss worldwide, over 1.2 billion live in underserved communities with the least access to hearing care. This includes preventable hearing loss in an estimated 42 million children who, without early identification and treatment, are at increased risk of significant detrimental effects that can last a lifetime. These immense hearing health disparities provide a clear and compelling call to action. The new Center for Hearing Health Equity at the University of Arkansas for Medical Sciences is the first of its kind in the world, established to deliver the critical mission of fostering hearing health equity both globally and locally. This article describes the Center's approach, which is built on inclusive, multidisciplinary collaboration guiding a research-driven, public-health oriented agenda to develop evidence that is directly translatable to policy change. The end goal is to generate momentum towards sustained solutions that will create a new future with hearing care accessible to all.

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Caregiver Coaching in Early Childhood Intervention

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Everyday life activities provide development-enhancing learning opportunities for young children. Since parents, teachers, child care providers, and other caregivers are with infants and toddlers more frequently and for longer periods of time than early childhood intervention practitioners, caregivers need to know how they can support their child's learning within the context of their interest- based activities in the home, community, and early childhood classroom. Coaching is an adult learning strategy and interaction style that may be used by early childhood intervention providers to build the competence and confidence of caregivers to use responsive strategies within these development-enhancing activities that will achieve their desired outcomes.

All young children learn new skills and improve existing ones by participating in everyday activities with the important people in their lives. When the development of infants and young children appears delayed or they are diagnosed with a syndrome or condition that may result in a high probability of delay, families may choose to access some support with promoting their children's learning. Part C of the Individuals with Disabilities Education Act (IDEA) helps ensure that children who are eligible and their families receive timely support from a team of individuals representing various disciplines (IDEA, 2004). IDEA also emphasizes that parents and other important people in the child's life are members of the team, are involved in the development of the Individualized Family Service Plan (IFSP), and participate in the intervention process. How services are provided can significantly impact children and their families, thus early childhood intervention practitioners must be certain they are using the most current research-based practices to ensure success. This includes involving caregivers and providing supports within the context of children's and families' typical activities to provide greater opportunities to develop and practice new skills naturally. Caregivers will then know how to maximize the development-enhancing learning opportunities that can occur as part of routine events such as mealtime, dressing, car rides, etc. Coaching is an evidence-based adult learning strategy used by early childhood intervention providers to build the capacity of caregivers to support children's development as part of their daily activities rather than focus on learning only when the provider is present (Rush & Shelden, 2020).

Coaching has become a widely used means of interacting with the important adults in the lives of children enrolled in IDEA Part C early intervention services, and a variety of different coaching approaches are referenced in the early childhood intervention literature (Friedman et al., 2012; Rogers et al., 2021; Rush & Shelden, 2011, 2020; Snyder et al., 2015). The field of early childhood intervention does not have a universally agreed upon operational definition of coaching, which leads to the variability in how the practice may be applied. A frequently cited (e.g., Kemp & Turnbull, 2014; Movahedazarhouligh, 2019; Ward et al., 2019), operational definition of coaching in early childhood intervention is:

> "An adult learning strategy in which the coach promotes the learner's (coachee's) ability to reflect on his or her actions as a means to determine the effectiveness of an action or practice and develop a plan for refinement and use of the action in immediate and future situations." (Rush & Shelden, 2005, p. 3)

This definition was developed and the characteristics of the practice were identified based on a review of over 30 research studies (Rush & Shelden, 2011). When compared to other coaching paradigms across fields of study, the caregiver coaching process described in this article uses an expert-based approach (Ackland, 1991) and contextual model (Akamoglu & Dinnebeil, 2017; Desimone & Pak, 2016, Friedman et al., 2012) with elements of goal-oriented (Ives, 2017; Megginson, 2014) and adult learning coaching approaches (Cox, 2015).

BACKGROUND & RATIONALE FOR CAREGIVER COACHING IN EARLY CHILDHOOD INTERVENTION

Caregiver Coaching is Expert-Based

The early coaching literature delineates the two basic forms of coaching as either expert-based or peer-to-peer (Ackland, 1991). Expert-based coaching uses an individual with specialized knowledge and skills as the coach whereas peer-to-peer coaching involves two individuals who are both learning a new skill at the same time. Caregiver coaching in early childhood intervention is expert-based rather than peer-to-peer. This does not mean that parents and caregivers do not have expertise. Parents are the experts on their child and family, and early intervention practitioners must obtain and can build upon the unique knowledge of families and caregivers to effectively promote child learning. Coaching in early childhood intervention is expert-based because IDEA Part C requires those working in early intervention to have expertise in their role by meeting discipline-specific, program, and state standards (IDEA, 2004). Many professional organizations provide guidelines that document the role of and expertise needed by individuals working in early intervention and some, including the American Academy of Pediatrics, refer specifically to the use of coaching when supporting caregivers (Adams et al., 2013; ASHA, 2008; AOTA, 2013; APTA, 2019; DEC, 2014). The "Mission and Key Principles for Providing Early Intervention Services in Natural Environments" indicate that the role of early intervention practitioners is to help family members and other caregivers promote their child's learning and development as part of their typical daily activities (Workgroup on Principles and Practices in Natural Environment, 2008).

How early intervention practitioners (i.e., therapists, teachers, audiologists, nurses, etc.) provide services is critically important and can determine whether or not early intervention will be successful (Dunst & Trivette, 2009). Coaching is a specific, capacity-building, help-giving strategy found to be effective when used in early childhood intervention. Several syntheses of research studies of caregiver coaching in early intervention have documented positive child and family outcomes (Elek & Page, 2019; Kemp & Turnbull, 2014; Tomeny et al., 2020; Ward et al., 2019). A recent review and meta-analysis found that coaching had positive effects when used to teach caregivers in early intervention as compared to other instructional approaches (Sone et al., 2021), and other authors have referred to the positive impact of coaching caregivers for children with specific diagnoses (Akamoglu & Dinnebeil, 2017; An et al., 2019; Dunn et al., 2012; Graham et al., 2013, 2014; Turner-Brown, 2019).

Caregiver Coaching is Goal-Oriented

Effective help-giving also requires that practitioners are responsive to the priorities of the family and other caregivers. Rather than the practitioner determining the goals and outcomes the child and family will be expected to achieve, IDEA Part C requires the use of an IFSP in which the caregivers identify the outcomes they want for their child and family. Caregiver coaching is goal-oriented. The focus of the coach is on assisting the caregivers to build on their existing strengths and determine how to support the caregivers with achieving the child and family outcomes on the IFSP.

Caregiver Coaching is Contextual

Young children with developmental delays and disabilities as well as those who are typically-developing learn new skills and practice existing ones by participating in everyday activities with the important people in their lives (Campbell & Sawyer, 2007; Dunn et al., 2012; Dunst et al., 2006; Hwang et al., 2013). IDEA Part C requires that services and supports for eligible children and their families be provided in their natural environments (IDEA, 2004). Natural environments are the locations where children would be if they did not have disabilities (IDEA, 2004). Principe 1 of the "Mission and Key Principles for Providing Early Intervention Services in Natural Environments" states, "Infants and toddlers learn best through everyday experiences and interactions with familiar people in familiar contexts" (Workgroup on Principles and Practices in Natural Environment, 2008, p.2). Early childhood intervention providers use natural learning environment practices to support parents of children with disabilities and others in understanding the critical role of everyday activity settings, child interests, and caregiver responsive interaction strategies as the foundation for children's learning (Dunst, 2006).

Caregiver Coaching is Consistent with Adult Learning Principles

Principle 3 of the "Mission and Key Principles for Providing Early Intervention Services in Natural Environments" is, "The primary role of a service provider in early intervention is to work with and support family members and caregivers in children's lives" (Workgroup on Principles and Practices in Natural Environment, 2008, p.2). Most early childhood teachers and pediatric therapists are taught to work directly with young children. In early childhood intervention, however, the principal learners are the adults who are with the child during more of their waking hours than an early intervention provider because development-enhancing learning opportunities happen all day, every day.

The National Research Council (NRC) conducted a research synthesis on human learning to identify teaching practices and environments that promote successful learning (Bransford et al., 2000). Findings of the NRC report indicated that in order for a learner to acquire new knowledge, the individual must develop an understanding of how that knowledge can be used in a specific context and also generalized to other situations (Bransford et al., 2000). The NRC identified three key findings from the research synthesis. First, an individual enters a learning situation with preconceived ideas about a subject matter. As a result, the learner may not develop an understanding of new information and skills being taught if their current understanding is not recognized. Second, to develop a deeper level of understanding in a particular area, the learner must understand the facts as they relate to the particular situation and organize the information for easy recall, use, and generalization to other situations. Third, the learner must develop the ability to self-assess one's current level of understanding, determine what additional knowledge and skills might be needed, and measure progress (Bransford et al., 2000). Coaching is defined as an adult learning strategy and the research-based characteristics of coaching are consistent with the NRC findings (Rush & Shelden, 2011, 2020).

Other research syntheses have also focused on adult learning and found coaching to be a proven strategy. A research synthesis from the implementation science literature examined practice implementation research and found that coaching was an effective approach for supporting the implementation of newly learned skills and practices as part of an intervention (Fixsen et al., 2005). Trivette et al. (2009) examined the research related to the characteristics of four adult learning methods. Findings indicated that the more active participation on the part of the learner, the stronger the outcomes. Coaching provides for active participation through the characteristics of action/practice, reflection, and joint planning (Rush & Shelden, 2011, 2020).

THE CAREGIVER COACHING PROCESS

Because how services are provided can determine the success of the intervention, coaching is the *adult learning* strategy an *expert* in early childhood intervention uses when operationalizing effective help-giving and a capacity-building approach to support caregivers in achieving their desired *goals* for the child by using interest-based everyday activities as the *context* for child learning and development. An early intervention practitioner who uses coaching keeps the caregiver in the lead with the child as much as possible to not only recognize and build their competence and confidence in supporting the child's learning during the visit, but also, most importantly, between visits when they are participating in their usual routines and activities as part of family, community, and early childhood classroom settings. The coach helps caregivers recognize what they are already doing that works to support child development and build on existing or identify new ideas rather than just telling the caregiver what to do or working exclusively with the child while the caregiver watches or carries on with other tasks. Coaching involves asking questions; jointly thinking with the caregiver about what works, does not work, and why; the caregiver trying new strategies with the child; modeling by the coach if necessary; sharing information; and jointly planning next steps (Rush & Shelden, 2020). A coaching interaction occurs during an activity the child and family would typically be doing at the time of the visit and is as hands-on (caregiver and coach) as it needs to be to support the caregiver in knowing how to foster the child's participation in the activity.

Understanding the characteristics or specific activities required to implement a practice is important so a practitioner will know what to do in order to achieve the desired effect (Fixen et al., 2015). The coaching research synthesis by Rush and Shelden (2011) was guided by a process that focused on the extent to which the specific characteristics of the practices are related to differences in their outcomes or consequences (Dunst et al., 2002). More specifically, the research synthesis examined the characteristics of coaching that were related to variations in the use of newly learned practices or improvement of existing skills. The coaching research studies included as part of the synthesis suggest that coaching has five practice characteristics that lead to the intended outcomes: (1) joint planning, (2) observation, (3) action/practice, (4) reflection, and (5) feedback. The characteristics are used in combination to recognize current knowledge, improve existing abilities, develop new skills, and deepen the understanding of the person being coached. These five characteristics were substantiated by a research synthesis conducted by Artman-Meeker and colleagues (2015), and individual characteristics have also been further examined as related to the coaching process (Elek & Page, 2019; Lorio et al., 2020, 2021; Tomeny et al., 2019; Ward et al. 2019).

Joint Planning

Use of a capacity-building approach requires active participation on the part of the caregiver (Dunst et al., 2014; Dunst & Trivette, 2009; Trivette & Dunst, 2007). Active participation occurs as part of several coaching characteristics, but the characteristic of joint planning is used to ensure the caregiver practices new skills that continue to foster child learning as part of everyday activities until the next scheduled visit. Joint planning is the process of determining what the coach and caregiver will each be responsible for doing between visits and in preparation for the next visit. The joint planning process has four parts: (1) revisitation of the previous between visit plan, (2) plan for the current visit, (3) new between-visit plan, and (4) new next-visit plan.

Revisitation of the Previous Plan

Each coaching conversation typically begins by revisiting the previous between-visit plan to determine what worked and if the caregiver might need some additional support to promote the child's learning in that activity. If that activity is the same as what the caregiver and coach had planned to do during the current visit (e.g., snack time), then the coach, caregiver, and child may move into the activity to build on what worked and/or further analyze what did not work as well as identify new ways to improve the child's participation. If the between-visit plan activity is something that is not time-bound (e.g., throwing a ball for the dog to retrieve in the backyard), then the caregiver might engage the child in the activity while the coach is present to receive further support. If the activity that was part of the previous between-visit plan would not typically occur at the time of the visit (e.g., getting ready for bedtime), then the caregiver and coach would engage in a discussion to reflect on the child's participation and caregiver responsive strategies used, and then consider a plan for continued implementation. If the caregiver and child are ready to begin the planned activity immediately upon arrival of the coach or are in the midst of the activity when the coach arrives, then the previous joint plan may be reviewed later in the visit or the caregiver might refer to what they had been doing between visits as it relates to the current activity.

Plan for the Current Visit

Every early childhood intervention visit should be based on a joint plan the caregiver and coach outlined at the conclusion of their previous visit. The plan should not be determined at the beginning of the visit and consist of toys or materials the coach brought; focus on a "lesson" or "exercise" the coach and caregiver create at the moment to work on a particular skill (e.g., putting raisins in a cup to work on counting; simply engaging the child in a play routine, putting together a puzzle, or reading a book because those types of activities can occur "anytime;" or based on the caregiver's response to the question, "What do you want to work on today?"). The plan for the visit is a real-life, interest-based activity that the caregiver and child would be doing at this time if the coach was not present, which is the reason why the coach scheduled the visit with the caregiver on this day at this time. The plan also includes what the caregiver wants the child to learn or be able to do in order to fully participate in the activity as well as what the caregiver wants to learn to help the child be able to successfully participate.

New Between-Visit Plan

All coaching conversations end with a new two-part joint plan. The first part is the new between-visit plan. This plan is developed by the caregiver with assistance from the coach as needed and may be a continuation of what occurred during the visit and/or generalization of newly learned skills to other typical activities and routines. The between-visit plan delineates the specific activities in which the caregiver will practice new strategies for promoting the child's participation, thereby providing the child with opportunities to use and further develop targeted skills. The activities selected for the between-visit plan are generally already occurring and happen frequently enough to provide practice for the caregiver and child. They should not be perceived as an extra task, exercise, or homework the caregiver must perform, thereby decreasing the likelihood of happening.

New Next-Visit Plan

The second part of the new two-part joint plan developed at the end of the coaching conversation is the next-visit plan. The purpose of this plan is to identify the family, community, or early childhood program activities that will serve as the context for the following visit based on the IFSP outcomes and caregiver's priorities for the child. Selection of the activities for the next visit helps to determine the day and time the coach should return as it needs to coincide with when the activity would normally happen for the caregiver and child. The caregiver and coach also discuss how the caregiver wants the child to participate in the activity and what the caregiver wants to learn to support the child's participation. The coach asks the caregiver to determine how much time is needed for practice before the coach should return.

Observation

The purpose of observation is for the coach to see the caregiver and child involved in a real-life activity in which the caregiver wants the child to successfully participate (e.g., walk to the kitchen table, tell the caregiver what she wants to eat, or reach for the spoon). The activity may currently be going well, so the caregiver and coach could determine what is happening to cause the success and perhaps how the strategies or actions of the caregiver could be transferred to other settings. The caregiver may want the coach to observe an activity because it is problematic so they can analyze what is causing a particular behavior or lack thereof, then brainstorm strategies for the caregiver to try. Observation provides the coach with content on which the coach and caregiver may reflect to expand the caregiver's and child's learning opportunities in the moment. Conversely, the caregiver might observe the coach model a technique as an option for the caregiver to use to help the child learn a new skill or behavior, if necessary. Modeling by the coach may be helpful with permission when the caregiver (1) is a visual learner, (2) asks to see the coach demonstrate the strategy, or (3) expresses frustration while trying to engage the child or the child becomes agitated and the coach does not want the caregiver and child to miss an important learning opportunity. Observation of the coach by the caregiver should be intentional. That is, the coach should ensure the caregiver knows what the coach is going to do and why, and then the coach provides the caregiver with the opportunity to practice what the coach just demonstrated while the coach observes.

Action/Practice

Another opportunity for active participation as part of the caregiver capacity-building process is the coaching characteristic of action/practice. As part of the visit, the caregiver immediately practices new ideas for promoting the child's learning during planned and spontaneous real-life activities occurring at that time. Action/practice typically follows observation, reflection, and, if necessary, feedback. The coach may provide modeling, prompts, direct teaching, reflection-in-action, and reflection-on-action as appropriate to support the caregiver's success with promoting the child's learning within the observed daily activity. The caregiver takes action between coaching visits by using what was learned during the visit and included in the joint plan.

Reflection

Reflection is the characteristic that differentiates coaching from other adult learning strategies. Rather than observing, assessing, and then suggesting or telling the caregiver what to do, reflection is consistent with a capacity-building and strengths-based approach by demonstrating respect for what the caregiver already knows and jointly considering how their ideas might work best for the child and family. A key finding of the research synthesis on how people learn was that everyone can be taught to reflect (Bransford et al., 2000). The coach gives the caregiver the opportunity to reflect during the visit to recognize and build upon the caregivers existing knowledge and skills. Reflection increases caregiver competence and confidence so they know what to do when the coach is not present, instead of creating dependence on the coach to solve problems, provide solutions, and work exclusively with the child. The coach prompts reflection by asking open-ended questions that create awareness by the coach and caregiver of what the caregiver already knows to do, is doing, or has tried to do to foster the child's learning and development, and provides opportunities for the caregiver to analyze how previously used strategies have worked in the past or how new approaches are currently working. The use of reflection in a coaching conversation also provides the caregiver with the opportunity to propose new ideas for how to help the child and accomplish the IFSP outcomes.

Reflection occurs on, in, and for action (Schon, 1987). Revisiting the previous between-visit plan at the beginning of the next visit allows the caregiver to reflect on how the targeted interest-based, everyday activity and responsive strategies worked to promote the child's participation in the activity, and determine if more support is needed from the coach related to that activity setting at this time or not. Reflection-on-action can also occur during the visit when the coach asks the caregiver to analyze the child's participation and caregiver's responsiveness *following* an activity observed by the coach. Reflection-in-action involves the coach asking the caregiver to analyze their use of specific responsive strategies while the caregiver and child are participating in a specific activity (e.g., caregiver using the "take a turn and wait" strategy for prompting the child's use of a word to request a bite of food at breakfast). If the strategy is not working as planned, the coach may prompt the caregiver to identify ways to modify the activity or strategy. Reflection-in-action may provide the caregiver and child with multiple opportunities to determine what works and practice within the context of an activity while the coach is present to assist (if needed). Reflection-for-action is used by the coach to help the caregiver think through or develop a plan for how to improve the child's learning and development during a targeted family activity (e.g., helping to find specific items while riding in the grocery cart at the store). The coach may be present if this is an activity planned to be the focus of the visit, so the coach and caregiver would spend a few minutes reflecting on the plan for what the caregiver will be doing with the child during the activity. Reflection-for-action also occurs when developing the between-visit and next-visit plans. The coach asks the caregiver to think about the interest-based activity setting(s) in which the caregiver wants to practice specific responsive strategies that will be best to encourage child learning between the current visit and next visit as well as how much time needs to pass between visits to give ample time for the caregiver and child to engage in that activity. The caregiver also reflects on what interest-based activity setting would be most helpful as the context for the next visit as well as what the caregiver wants the child to learn as a result, and when would be the best time to schedule the visit according to when the activity usually occurs.

Feedback

Feedback is provided by the coach based on an observation or at the request of the caregiver, actions shared by the caregiver, or reflections provided by the caregiver related to the past, current, or planned activity. Feedback may follow an observation (coach's or caregiver's), reflection, action, or practice opportunity. The coach should prompt caregiver reflection or sharing of information prior to giving any feedback. Feedback is provided as necessary to build the caregiver's knowledge, skills, and/or confidence, or in response to a direct question from the caregiver. Feedback takes many forms. It can be information provided by the coach to deepen the caregiver's understanding and should be research-informed rather than the coach's personal opinion. Feedback may also be a verbal or nonverbal acknowledgement or affirmation of what the caregiver is saying or doing. Feedback also includes praise, positive reinforcement, and encouragement, although this type of evaluative feedback should be used carefully and intentionally to avoid creating the caregiver's dependence on the coach's approval or coming across as patronizing. Feedback should not be directive in the form of suggestions or recommendations given prior to the caregiver having the opportunity to share and reflect. Directive feedback creates dependence on the coach and does not ensure the caregiver can or will apply the feedback.

Use of the coaching characteristics is not a linear or step-by-step process, but rather a natural and logical conversation as it transpires within the context of the interaction. Typically, the coach begins and ends with joint planning. Observation and action/practice occur when the caregiver and child are engaged in the real-life activities that are planned as the focus of the visit. Reflection and feedback may be used prior to, during, and/or after the activity as well as part of the re-visitation of the previous between-visit plan and development of the new two-part joint plan at the end of the visit.

CONCLUSION

Caregiver coaching is an expert-based, goal-oriented, and contextualized approach to adult learning that may be implemented by early childhood intervention practitioners to build the competence and confidence of parents, child care providers, and other important people in the child's life and promote child learning and development. Because young children learn by participating in interest-based, everyday activities, the focus of coaching is on the caregiver knowing what to do to promote development as part of their routine interactions, thus providing frequent opportunities for the child to learn and practice new skills. Coaches operationalize the five research-based characteristics of observation, action/practice, reflection, feedback, and joint planning to support caregivers' use of natural, interest-based learning opportunities and responsive strategies. Through the application of these natural learning environment practices and a coaching interaction style, early intervention practitioners empower caregivers to achieve their desired priorities for themselves and the children.

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Preliminary Findings: Early Turn Talking is Associated with Later Social-Emotional Development

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Preliminary findings from a follow-up study of 33 children, assessed for their socioemotional development from 18 months of age, are presented. At 77 months, they were evaluated again in a laboratory context, recording on video a situation of discussion of a conflict with the caregiver and then coding these videos using the CIB instrument. Conversation turns assessed with the LENA software at 18 and 30 months of age are associated with socioemotional cognition, emotion regulation, and emotional communication competencies assessed at 77 months of age. The association is stronger at 30 months than at 18 months, suggesting a possible mediation effect to be explored in future analyses. Implications for professional intervention and public policies for children are discussed.

BACKGROUND

In 1995, Hart and Risley's groundbreaking study showed the world that the number of words adults direct at children predicts the development of their language skills. Subsequent research confirmed the importance of talking to infants and toddlers from an early age to promote their cognitive and linguistic development (Chapman, 2000; Hart & Risley, 1995; Gilkerson et al., 2018; Hirsh-Pasek et al., 2015; Hoff, 2006; Romeo et al., 2017; Rowe, 2008).

Based on these and other pieces of evidence, we conducted a study between 2016 and 2019 that explored the longitudinal relationships between the number of conversational turns (assessed using the LENA software; Weisleder & Fernald, 2013) at 18 months of age and socioemotional development at 30 months. The results of this study were published in *Developmental Science* (Gómez & Strasser, 2021) and received significant attention from the international scientific community as they provided evidence regarding how conversational turns also influence socioemotional development.

In that study, we measured 43 infants' language environment and socioemotional competencies at 18 months and again at 30 months of age. Using hierarchical multiple regressions, we found that turn-taking at 18 months contributed significantly to the development of all socioemotional competencies assessed at 30 months (attachment, emotion regulation, and emotional communication). This relationship held even after controlling for baseline levels of each assessed socioemotional competency, as well as for infant vocalizations, maternal warmth, infant temperament, and cumulative social risk (an index that combined poverty, overcrowding, maternal depression, maternal adolescent, low educational level of the mother, and alcohol abuse or domestic violence in the family) at 18 months of age. Additionally, we performed cross-lagged analyzes that revealed that the direction of the longitudinal relationship between conversational turns and emotional competencies went more strongly from conversational turns to socioemotional development (but not the other way around), providing evidence of a possible causal relationship and initiating a promising line of research in the field.

The initial findings of our work (Gómez & Strasser, 2021) left a set of relevant questions to be considered in further studies: (a) although infants' early vocalizations were used as a control variable, it is important to explore the degree to which children's language skills are associated with turn-taking and to what extent the development of their socio-emotional skills is associated with, for example, language and vocabulary development at later ages; (b) socioemotional competencies were evaluated using a play situation, free of induced stress, making it more difficult to observe their performance under situations of greater emotional and interpersonal demand, such as, for example, in a conflict situation associated with unpleasant or painful emotions or thoughts; and perhaps the most relevant question of all, (c) whether conversational turns in early childhood continue to be significant predictors of social-emotional development in school and beyond.

METHOD

In 2021, the Relational Neuroscience Laboratory of Fundación América por la Infancia in Santiago, Chile, conducted a follow-up study of the children from the original study by Gómez and Strasser (2021), who were now in school and ages 6 to 7 years old.

To do this, a trained research assistant made telephone contact with each family of the original sample to explain the nature of the study, following a written protocol. With those families that were available to participate, they were offered transportation to go from their homes to the laboratory and back, respecting a series of care protocols for reasons of the COVID-19 pandemic. The boy or girl identified as a participant in the original study, plus the mother or primary caregiver, attended the laboratory. The Informed Consent form for adults and the Informed Assent form for children were read and explained in detail. All procedures and instruments were approved by the Scientific Ethics Committee of Fundación América por la Infancia.

Instruments

The following instruments were applied: (a) a sociodemographic characterization sheet, (b) the Peabody test to evaluate vocabulary, (c) the protocol recorded on video for coding with the CIB instrument, (d) the Dp-3 questionnaire, (e) the BarON, (f) the Hearts and Flowers test, and (g) the Gaze Test. In this article, correlational findings are reported and centered on the CIB–Coding Interactive Behavior, an instrument that has been widely used in research due to its precision and versatility in addition to having adequate psychometric properties of validity and reliability (Feldman, 1998; Halevi et al., 2017; Ulmer-Yaniv et al., 2017).

The CIB protocol was designed in the Laboratory of Dr. Ruth Feldman in Israel and consists of videotaping a discussion of a *mother-child conflict situation*. The advantage of this protocol is that, by confronting the child with a discussion about a conflict with their attachment figure, emotional stress is induced, and a cognitive, regulatory, and communicational challenge is generated that summons socio-emotional skills for its resolution. This situation was evaluated by combining specific codes from the CIB instrument according to theoretical relevance so that we could assess the three socio-emotional competencies proposed by Gómez and Strasser (2022): (a) socio-emotional cognition, (b) emotion regulation, and (c) emotional communication.

Socioemotional cognition is defined as the ability to pay attention to social and emotional cues and understand them in a referential framework of meaning. It includes socio-emotional attention, which focuses the organism on social cues and emotional indicators, as well as socio-emotional interpretation, which allows understanding the meaning of social cues and the emotions, intentions, and mental states of oneself and others, combining mentalization and empathy.

To assess socioemotional cognition, the following CIB codes were used: (a) *child's gaze*: the child's gaze is constantly focused on the parent or on the joint activity and maintains eye contact throughout the interaction; (b) *motivation*: the child tries to explain his/her point of view to the parent and makes an effort to understand the parent's perspective during the discussion of the conflict; (c) *initiative/creativity*: the child contributes ideas and solutions to the conflict and develops these ideas, such as being proactive, suggesting what to talk about, contributing ideas and perspectives, to try to resolve the relational conflict with the parent figure. The internal consistency of these three items was very good, obtaining a Cronbach's alpha of α = .83.

Emotion regulation is defined as "the process of initiating, avoiding, inhibiting, maintaining, or modulating the occurrence, form, intensity, or duration of emotional states, emotion-related physiological or attentional processes, motivational states, and/or the behavioral concomitants of emotion, in the service of achieving biological or social adaptation linked to affects" (Eisenberg & Spinrad, 2004). In our model, emotion regulation is made up of inhibitory control and emotional modulation capacities.

To assess emotion regulation, the following codes were used: (a) *affective lability*: the child's affect is labile and shows sudden changes in mood and emotional expression; the child switches from engagement to disengagement or from positive to negative emotionality for no obvious reason or in response to situational events; the reaction is disproportionate to the situation and the affect is unpredictable; (b) *acceptance of limits*: the child respects and accepts the limits that the parent sets regarding ways of talking (e.g., yelling, humiliating the parent, cursing) or other limits set by the parent. The child can accept parental requests during the interaction (e.g., "I want you to sit down and stop walking around the room until we are finished"); (c) *cooperation*: the extent to which the child cooperates with the parent in the task, showing willingness to hold a conversation about a relational conflict in search of a solution or agreement. The internal consistency of these three items was acceptable, obtaining a Cronbach's alpha of α = .63.

Finally, *emotional communication* refers to the use of various communication mechanisms to share the emotional world by connecting, linking, coordinating, and interacting successfully with others, including verbal communication and gestural, corporal, and non-verbal communication skills.

To assess emotional communication, the following CIB codes were used: (a) *conversational involvement*: the child engages in conversation, collaborates, exhibits positive affect, shows interest, and shows that the interaction is important to him or her; (b) *child vocalization*: refers to the child's vocalizations that are part of social interaction with the caregiver and how the child talks to the caregiver (including laughter); it is not about how much you talk, but how you express yourself verbally in the interaction when discussing a conflict; (c) *self-confidence*: the degree to which the child dares to

TABLE 1: Correlations between language variables at 18, 30, and 77 months (*N*=33)

		Child vocalizations 18m	Vocabulary 77m	Conversational Turns 18m	Conversational Turns 30m
Child vocalizations 18m	*R*	1	.326	.597**	.290
	Sig.		.064	.000	.101
Vocabulary 77m	*r*	.326	1	.209	.428*
	Sig.	.064		.243	.013
Conversational Turns 18m	*R*	.597**	.209	1	.660**
	Sig.	.000	.243		.000
Conversational Turns 30m	*R*	.290	.428*	.660**	1
	Sig.	.101	.013	.000	

**Correlation is significant at the 0.01 level (2-tailed)
*Correlation is significant at the 0.05 level (2-tailed)

express his/her point of view clearly and assertively in the presence of the caregiver; confidence is reflected not only by presenting a safe position, but also when the caregiver and the child are expressing different points of view or opinions and when the child disagrees with something the caregiver said. The internal consistency of these three items was good, obtaining a Cronbach's alpha of $\alpha = .79$

RESULTS

First, the data was explored descriptively, to then perform correlational analyzes between the variables. The children were distributed in 54% men and 46% women, with an average age of 6.4 years (*SD* = 0.5), equivalent to 77 months of age on average. The caregivers were 94% women (mainly mothers) and 6% men (grandparents), and their average age was 36.7 years old (*SD* = 8.7).

Language Variables

The vocalizations of the boys and girls at 18 months averaged 161.5 (*SD* = 66.2), with a minimum of 20 and a maximum of 345 vocalizations per hour. Regarding the conversation turns, at 18 months of age they averaged 43.6 (*SD* = 20.7) with a minimum of 7 and a maximum of 105; and an average of 60.9 (*SD* = 26.0) with a minimum of 19 and a maximum of 132 at 30 months, showing a clear upward trend in the number of conversational shifts between 18 and 30 months of age. Regarding the vocabulary evaluated at 77 months, an average of 97.3 (*SD* = 22.6) was observed, with a minimum of 4 and a maximum of 123. The associations between these variables can be seen in Table 1.

Vocalizations at 18 months are only concurrently associated with conversational turns, but do not predict turns at 30 months or vocabulary at 77 months, although the strength of the correlation in the latter case was $r = .326$, with $p = .064$ being marginally significant. In contrast, conversational turns at 30 months do show a significant association with vocabulary at 77 months with $r = .428$ and $p = .013$.

Variables of the Family System and Socio-Emotional Competencies

The cumulative risk averaged 1.6 (*SD* = 1.4), with 30.3% registering zero risk factors, 18.2% one factor, 24.2% two factors, 12.1% three factors, and 15. 2% four risk factors accumulated in the family system. The correlations between the variables of the family system (accumulated risk, the child's temperament, and the mother's emotional warmth) at 18 months, and the children's socio-emotional competencies at 77 months are shown in Table 2.

As can be seen, neither the cumulative risk nor the baby's temperament at 18 months are longitudinally associated with socioemotional competencies five years later. In contrast, the emotional warmth of the mother at 18 months does show a significant association with socio-emotional cognition ($r = .463$), emotion regulation ($r = .392$), and emotional communication ($r = .398$).

Socioemotional Competencies

Regarding the longitudinal measures of socio-emotional development, an association was observed between the variables evaluated within each age considered (at 18 months, 30 months, and 77 months) but not between the ages evaluated (see Table 3). In other words, trajectories of wide variability are observed in the socio-emotional development of boys and girls as time goes by whereas, for example, emotion regulation at 18 months is not associated with emotion regulation at 30 or at the 77 months.

Conversational Turns and Social-Emotional Development

Table 4 shows that children's vocalizations at 18 months are not longitudinally associated nor are their vocabulary at 77 months concurrently associated with socioemotional competencies at 77 months. Additionally, conversational turns at 18 months do not show a relationship with socioemotional cognition or emotional communication at 77 months, but they do with emotion regulation ($r = .393$). On the other hand, the conversational turns at 30 months do show a relevant

TABLE 2: Correlations between family system variables at 18 months and socioemotional competencies at 77 months (*N*=33)

		Cumulative Risk 18m	Temperament 18m	Emotional Warmth 18m	Socioemotional Cognition 77m	Emotion Regulation 77m	Emotional Communication 77m
Cumulative Risk 18m	*r*	1	.153	-.023	-.287	-.138	-.244
	Sig.		.395	.899	.105	.445	.170
Temperament 18m	*r*	.153	1	.039	.129	.042	.142
	Sig.	.395		.831	.475	.817	.432
Emotional Warmth 18m	*r*	-.023	.039	1	.463**	.392*	.398*
	Sig.	.899	.831		.007	.024	.022
Socioemotional Cognition 77m	*r*	-.287	.129	.463**	1	.742**	.856**
	Sig.	.105	.475	.007		.000	.000
Emotion Regulation 77m	*r*	-.138	.042	.392*	.742**	1	.695**
	Sig.	.445	.817	.024	.000		.000
Emotional Communication 77m	*r*	-.244	.142	.398*	.856**	.695**	1
	Sig.	.170	,432	.022	.000	.000	

**Correlation is significant at the 0.01 level (2-tailed).
*Correlation is significant at the 0.05 level (2-tailed).

TABLE 3: Correlations between socio-emotional development variables at 18, 30, and 77 months (*N*=33)

		CS 18m	RE18m	CE 18m	CS 30m	RE 30m	CE 30m	CS 77m	RE 77m	CE 77m
CS 18m	*r*	1	.768**	.746**	.235	-.051	.318	.292	.151	.216
	Sig.		.000	.000	.219	.791	.093	.099	.402	.228
RE 18m	*r*	.768**	1	.703**	.305	.121	.307	.246	.141	.070
	Sig.	.000		.000	.107	.533	.105	.167	.435	.698
CS 18m	*r*	.746**	.703**	1	.276	.069	.469*	.218	.184	.199
	Sig.	.000	.000		.148	.722	.010	.223	.305	.266
CS 30m	*r*	.235	.305	.276	1	.696**	.537**	.228	.351	.001
	Sig.	.219	.107	.148		.000	.003	.234	.062	.994
RE 30m	*r*	-.051	.121	.069	.696**	1	.517**	-.023	.125	-.168
	Sig.	.791	.533	.722	.000		.004	.905	.517	.384
CE 30m	*r*	.318	.307	.469*	.537**	.517**	1	.318	.347	.186
	Sig.	.093	.105	.010	.003	.004		.093	.065	.334
CS 77m	*r*	.292	.246	.218	.228	-.023	.318	1	.742**	.856**
	Sig.	.099	.167	.223	.234	.905	.093		.000	.000
RE 77m	*r*	.151	.141	.184	.351	.125	.347	.742**	1	.695**
	Sig.	.402	.435	.305	.062	.517	.065	.000		.000
CE 77m	*r*	.216	.070	.199	.001	-.168	.186	.856**	.695**	1
	Sig.	.228	.698	.266	.994	.384	.334	.000	.000	

**Correlation is significant at the 0.01 level (2-tailed).
*Correlation is significant at the 0.05 level (2-tailed).

CS = socioemotional cognition; ER = emotion regulation; CE = emotional communication

predictive association of socioemotional competencies at 77 months, both for socioemotional cognition (*r* = .558), emotion regulation (*r* = .558), and for emotional communication (*r* = .561).

DISCUSSION

The relationship found between conversational turns at 30 months and vocabulary at 77 months could account for how everyday conversations that occur between the ages of 2 and 3 years old in boys and girls play a relevant role in the development of their later language and vocabulary, which is in line with findings previously reported in the literature (Chapman, 2000; Gilkerson et al., 2018; Hart & Risley, 1995; Hirsh-Pasek et al., 2015; Hoff, 2006; Romeo et al., 2017; Rowe, 2008).

The study did not find an association between the initial temperament of infants and their socioemotional development at school, which shows that temperament

TABLE 4: Correlations between language variables and socioemotional development at 77 months (*N*=33)

		VN 18m	VB 77m	TC 18m	TC 30m	CS 77m	RE 77m	CE 77m
VN 18m	*R*	1	.326	.597**	.290	.054	-.123	-.030
	Sig.		.064	.000	.101	.767	.496	.867
VB 77m	*R*	.326	1	.209	.428*	.158	-.122	.164
	Sig.	.064		.243	.013	.379	.498	.362
TC 18m	*R*	.597**	.209	1	.660**	.274	.393*	.198
	Sig.	.000	.243		.000	.123	.024	.269
TC 30m	*R*	.290	.428*	.660**	1	.558**	.558**	.561**
	Sig.	.101	.013	.000		.001	.001	.001
CS 77m	*R*	.054	.158	.274	.558**	1	.742**	.856**
	Sig.	.767	.379	.123	.001		.000	.000
RE 77m	*R*	-.123	-.122	.393*	.558**	.742**	1	.695**
	Sig.	.496	.498	.024	.001	.000		.000
CE 77m	*R*	-.030	.164	.198	.561**	.856**	.695**	1
	Sig.	.867	.362	.269	.001	.000	.000	

**Correlation is significant at the 0.01 level (2-tailed).
*Correlation is significant at the 0.05 level (2-tailed).

VN = child vocalizations; VB = child's vocabulary; TC = conversation turns; CS = socioemotional cognition; ER = emotion regulation; CE = emotional communication

and socioemotional development are different theoretical constructs that refer to different aspects of the person (Mangelsdorf & Frosch, 1999; Gross, 2014). Nor was a relationship found between the risk accumulated in family systems at 18 months and subsequent socio-emotional skills, which on one hand could be explained by the dynamism of family systems over the years, and on the other could show that there are proximal processes of greater direct influence than social and family risk in the development of children's socio-emotional skills, such as the emotional warmth of the attachment figure, parenting practices, daily interactions of good or maltreatment, and conversational turns, among other possibilities (Halevi et al., 2017; Gómez & Strasser, 2021, 2022; Gómez, 2022).

The relationship between the emotional warmth of the mother in early childhood and the socio-emotional competencies of children five years later is another interesting finding, which is consistent with research on the importance of positive parenting, sensitivity, and emotional availability of the caregiver in parenting for positive development over time (Gómez, 2022). The association was slightly stronger for socioemotional cognition (attention, mentalization, and empathy), while it was similar for both emotion regulation and emotional communication.

The finding of a stronger association of conversational turns at 30 months with socioemotional competencies at 77 months could be suggesting a mediation effect, in which part of the effect of conversational turns at 18 months passes through conversational turns at 30 months, a possibility that should be explored through mediation analysis in future publications.

The fact that neither vocalizations at 18 months nor vocabulary at 77 months showed a relationship with socioemotional competencies at 77 months reinforces the finding by Gómez and Strasser (2021) that the effect of conversational turns it was sustained even after controlling for the child's individual language skills. Conversational turns at 30 months predict vocabulary and socioemotional competencies of boys and girls at 77 months, but vocabulary is not concurrently associated with socioemotional competencies at that age. This underlines the specific importance of turn-taking as a possible central mechanism in explaining developmental trajectories of socio-emotional competencies. The present study adds to the preliminary longitudinal evidence of Gómez and Strasser (2021) regarding what happens with this relationship beyond early childhood to the school stage.

Combining the study by Gómez and Strasser (2021) (regarding the effect of turn-taking at 18 months on socioemotional development at 30 months) with these preliminary findings from a follow-up study at school (showing an effect of conversational turns at 30 months on socio-emotional development at 77 months), a possible causal mechanism of socio-emotional development throughout the life course is outlined, based on turns and on the exchange of back and forth that would act as a training ground, a laboratory for experimentation and daily discoveries that shape the skills of socio-emotional cognition, emotion regulation, and emotional communication at each age (Gómez & Strasser, 2022).

Future studies should determine the real scope of these preliminary conclusions, expanding the sample and improving the statistical power of the studies, diversifying the characteristics of the participating subjects, making the data analysis models more complex, longitudinally extending the ages studied, diversifying the type of indicators of socio-emotional development considered (to include, for example, psychophysiological indicators, among other possibilities), and including measures not only of the number of turns of conversation but also of the content and quality of the narratives exchanged in those turns.

The implications of these findings, both for professional intervention and for public policies for children, are promising, although much more scientific evidence is still needed to support them. In its current embryonic state, the data suggest that informing and sensitizing the general population about the critical importance of talking to babies—especially of learning to hold dialogues (back and forth conversations with pause and a delicate wait for the intention of the child); training the ability to listen to the baby's vocalizations, paying attention to their signals to exchange sounds in the language, words, and expressions; waiting for the words and expressions of the child, to weave a shared narrative, that is conversational turns, shifts that give a space to the child's internal world as it expresses itself but that also offer the child the possibility of knowing the adult's own world—is key to teaching and professional intervention. If these findings are correct, working on this focus is a key piece to protect and promote positive trajectories of socio-emotional development not only in early childhood, but also in the school stage of boys and girls.

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Foundations for Literacy: A Research-Based Early Reading Program that Improves Outcomes for Children who are Deaf and Hard of Hearing

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One avenue for improving reading outcomes is to ensure children who are deaf or hard of hearing (DHH) enter school with the foundational skills needed to learn to read. Our research team developed an early literacy curriculum specifically for DHH children. Teachers use Foundations for Literacy (FFL) in a one-hour literacy block for the school year. Student learning objectives include improving spoken phonological awareness, alphabetic knowledge, word reading, vocabulary, and narrative. FFL is more systematic, and its instruction is more explicit, multi-modal, and intensive than might be used with children who have typical hearing. Much of the instruction is embedded in language-rich activities. Differentiation of instruction to the wide variation of language and phonological processing skills observed for children who are DHH is integral to the design. Results from multiple studies, including a randomized-control trial, indicates that FFL is an effective intervention for young DHH students.

Children who deaf or hard of hearing (DHH) and are acquiring spoken language need the same foundational skills to learn to read as children with typical hearing (TH). Researchers have found that phonological awareness, alphabetic knowledge, and vocabulary predict reading abilities in young deaf children with cochlear implants and hard-of-hearing children with hearing aids (Ambrose et al., 2012; Cupples et al., 2014; Easterbrooks et al., 2008; Lederberg et al., 2013; Nittrouer et al., 2012; Webb & Lederberg, 2014; Webb et al., 2015). These studies showed that the majority of DHH children are delayed in these skills compared to TH children, with wide individual differences. There is a strong need for intervention in the early childhood years (3-5 years old) and beginning school years (5-6 years old) that focuses on these skills. Our interdisciplinary team developed and assessed the efficacy of an early literacy curriculum for DHH prekindergarten children, called *Foundations for Literacy* (FFL). This paper describes our research and the curriculum used.

BACKGROUND

Our team began the process of developing FFL with a systematic review of research that exists about effective early childhood education programs for TH preschoolers/prekindergartners (National Early Literacy Panel, 2008). Primarily based on the Simple View of Reading (Gough & Tummer, 1986) and Scarborough's Reading Rope (Scarborough, 2001), effective early childhood education programs focus on improving the fundamentals for learning to read by emphasizing both the skills necessary to learn to decode or read individual words (referred to as code-based) and the skills necessary to understand what is read (referred to as meaning-based).

Code-based skills acquired during early childhood include both phonological awareness and alphabetic knowledge (i.e., knowledge of the relations between letter names, sounds, and shapes). Phonological awareness refers to the ability to identify and manipulate the sound units of spoken language. Phonological awareness skills typically taught during preschool include syllable segmentation (e.g., how many word parts are there in *popcorn*?), rhyming (do *cat* and *bat* rhyme?), initial sound identification (what is the first sound in *man*?), and blending sounds (Listen, *c-a-t*, what word do you hear?). Alphabetic knowledge includes letter-name (the name of this letter is m) and letter-sound knowledge (this letter says mmmm). In the United States, early childhood education programs have historically targeted teaching letter-name knowledge. However, evidence suggests that, for TH and DHH children who are acquiring spoken language, letter-sound knowledge has a stronger relation to learning to read because it is foundational to sounding out or decoding written words (National Early Literacy Panel, 2008; Webb, et al., 2015). In a meta-analysis of 78 studies with TH children, Shanahan and Lonigan (2010) found that code-based interventions improved phonological awareness and alphabetic knowledge in TH preschoolers. Interventions with the greatest impact on learning were those that taught phonological awareness and alphabetic knowledge together.

Early childhood programs also build meaning-based skills such as vocabulary and more complex language, which are foundational for reading comprehension (i.e., for understanding the words decoded.) Research suggests structured book-reading (such as dialogic reading) and language enrichment activities are effective in improving language. A meta-analysis of 37 early childhood programs found large to moderate effect sizes for increasing TH children's oral language skills, particularly vocabulary (Shanahan & Lonigan, 2010).

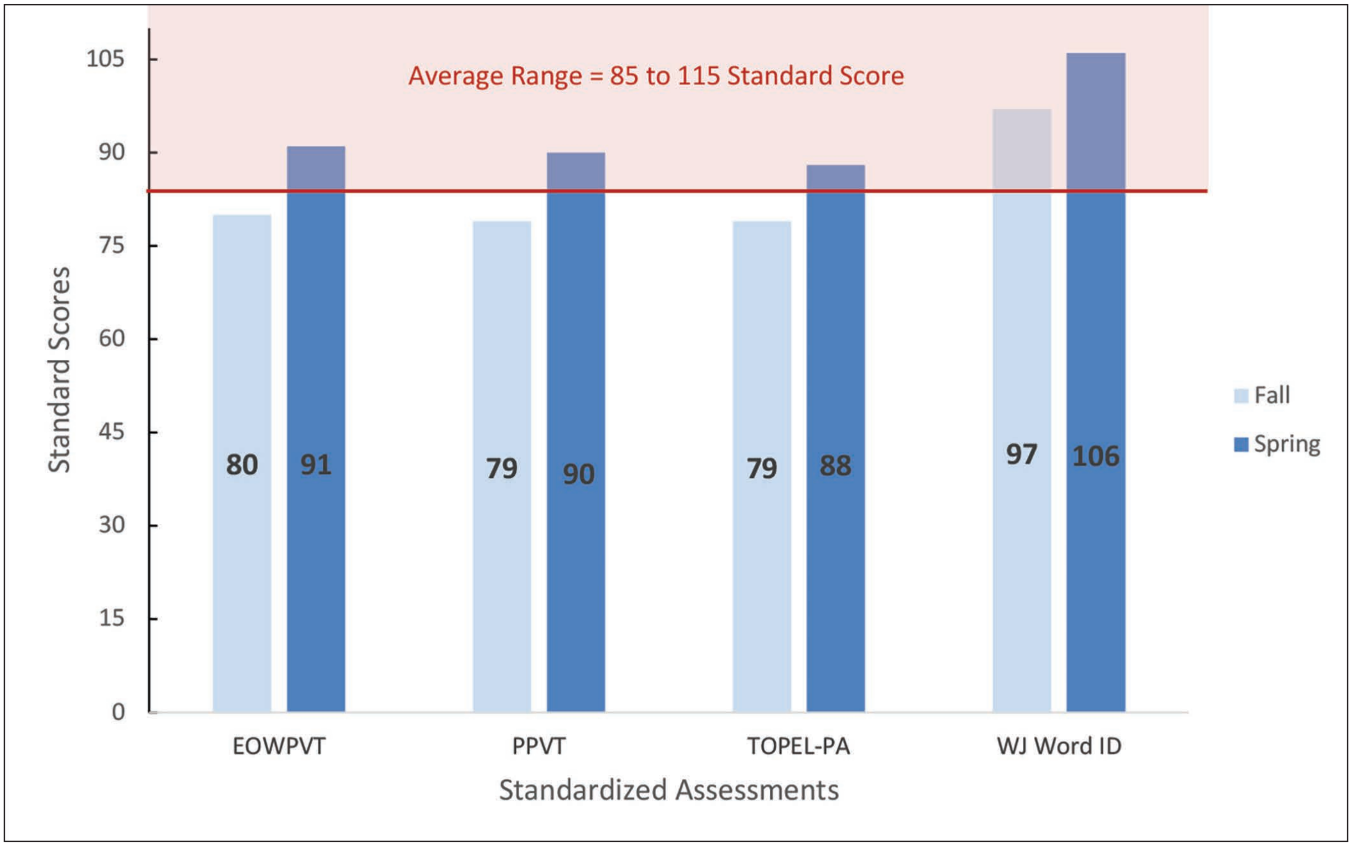
After examining available curricula developed for TH preschool children, our team decided they were not ideal for the majority of DHH children. Commonly used preschool curricula relied too heavily on incidental learning, delivered instruction too quickly, and used instructional language that was too difficult for many DHH children. Our team adopted the literacy objectives of effective, integrated, code- and meaning-based early childhood programs for TH children, but adapted instruction to the unique learning needs of DHH children. FFL is a year-long curriculum designed to be used daily with 3- to 6-year-old DHH children. Hearing loss results in decreased access to spoken language that may cause incomplete phonological representation of words and phonemes, as well as delayed language. Therefore, FFL targets these foundational skills more explicitly and with greater intensity than interventions developed for TH children. Cognitive theories (e.g., Dual-Code theory; Sadoski & Paivio, 2001) and theories of early word reading (Ehri, 2014) suggest that these targeted foundational skills would be acquired best in the context of instruction designed to build multi-modal (visual, auditory, kinesthetic) and semantic (i.e., meaning-based) representations. One novel instructional strategy used in FFL is to build strong semantic associations for the sounds of language. For example, children listen to a story that includes a snake saying *ssss*. The children then make playdoh snakes and pretend their snakes are saying *ssss*. These experiences give children a meaningful connection for the /s/ sound that is reinforced with a picture. During these experiences, the teacher also makes an explicit connection between the letter s and the sound /s/ by saying, "the letter s makes the *ssss* sound, too". Ehri (2014) has shown that TH children remember letters better when they are made more meaningful by pairing a picture with the letter shape. Because DHH children experience greater difficulties remembering sounds than letter shapes, we applied Ehri's semantic association strategy to learning sounds rather than learning to recognize letters. FFL instruction also includes visual and kinesthetic support when teaching phonological awareness, alphabetic knowledge, vocabulary, and narrative elements (i.e., character and setting identification, sequencing of events, and story retell). In order to address most DHH children's language delays, literacy instruction is embedded in language-rich activities that provide a context to teach vocabulary and more complex language while developing code-based skills. Finally, DHH children show large individual differences in early language and literacy skill development. Therefore, each lesson in FFL includes strategies for differentiating instruction based on children's speech perception, developmental language abilities, and language usage. For those children who primarily use sign language and do not have access to sound, we include alternatives to sound-based phonological skills, including fingerspelling and bilingual instructional strategies.

DEVELOPMENT OF FFL

FFL was developed in three phases. During the first phase, research teachers implemented lessons that followed a basic framework developed by an interdisciplinary team of teachers and researchers. Research teachers were certified teachers of the deaf who were part of the development team. They taught children in small groups—four days per week, one hour per day—for the full school year. They gave immediate feedback to the team on child engagement and instructional effectiveness, and lessons were adapted on an ongoing basis. A series of studies indicated that children taught with FFL by research teachers made educationally meaningful gains in phonological awareness, alphabetic knowledge, and vocabulary (Beal-Alvarez et al., 2012; Bergeron et al., 2009; Lederberg et al., 2014; Miller et al., 2013; Tucci & Easterbrooks, 2014). Children who received FFL made significantly more gains in phonological awareness and alphabetic knowledge than children in the comparison group who received business-as-usual instruction.

During the second phase, the research team moved FFL into classrooms where instruction was delivered by the teacher of record (Lederberg, 2016). The team developed a detailed teacher's manual and a two-day professional learning workshop. Eight classroom teachers attended the workshop and received ongoing coaching by members of the research team. These classrooms teachers implemented FFL as their literacy curriculum. The research team used observations of classroom teachers and weekly teacher feedback to improve lesson content and professional development. As is displayed in Figure 1, the 32 DHH children who received FLL from these eight classroom teachers began the year with delays on standardized measures of phonological awareness, alphabetic knowledge, and vocabulary. By the end of the school year, they gained, on average, 10 standard points and ended the year within a standard deviation of the normative average for TH children. In other words, DHH children who received FFL from their classroom teachers showed accelerated learning and achieved end-of-year scores that placed them within the typical range for TH children for phonological awareness, alphabetic knowledge, and vocabulary.

FIGURE 1. Fall and Spring Vocabulary Standard Scores for Children Taught FFL by their Classroom Teachers

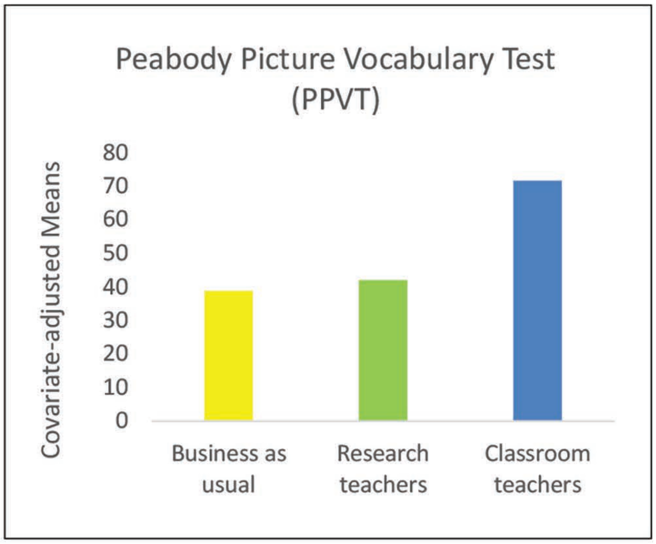


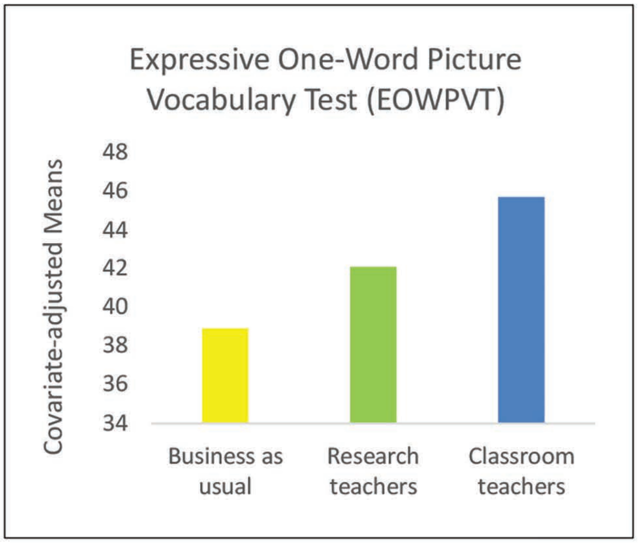
Note. Expressive One Word Picture Vocabulary Test (EOWPVT; Martin & Brownwell, 2011); Peabody Picture Vocabulary Test (PPVT; Dunn & Dunn, 2007); The Test of Preschool Early Literacy-Phonological Awareness (TOPEL-PA; Lonigan, Wagner, & Torgesen, 2007); Woodcock-Johnson Tests of Achievement-III-NU Letter-Word Identification. (WJ-WordID; Woodcock, McGrew, & Mather, 2007). All tests have a mean standard score of 100 with a standard deviation of 15, with 85 representing the lower limit of typical development.

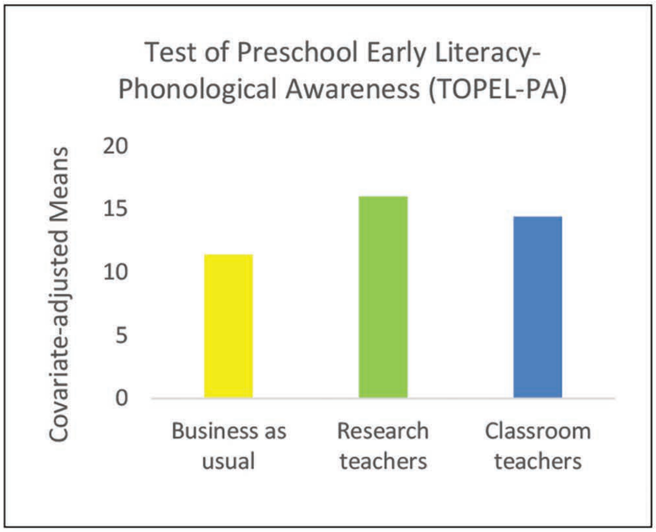
Next, we compared these children's fall-to-spring gains with the gains made by DHH children who were in the business-as-usual group enrolled in phase 1 (see Figure 2). DHH children receiving FLL from their classroom teachers improved their early literacy skills more than the DHH children in the business-as-usual group. Specifically, DHH children who received FLL from classroom teachers made greater statistically significant gains on measures of phonological awareness, alphabetic knowledge, and vocabulary than did DHH children taught by classroom teachers who used their typical instruction. In a second comparison, DHH children who received FLL from their classroom teachers made equivalent or greater gains on these measures than DHH children who received FFL from research teachers (Figure 2).

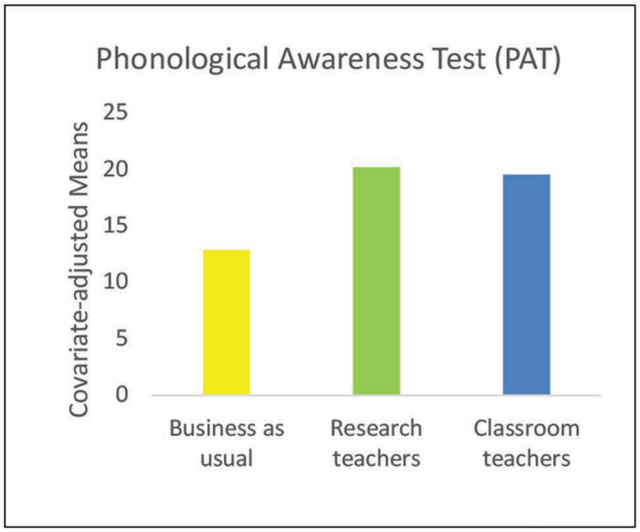
During the third and final phase of our research, the research team conducted a national randomized-control trial (RCT) of FFL (Lederberg et al., 2018). Forty-eight classroom teachers were randomly assigned to either intervention (teachers used FFL as their literacy curriculum) or control (teachers continued their business-as-usual instruction) conditions. Children were enrolled in the condition assigned to their teacher. Classrooms were in rural, urban, or suburban schools located in 14 states. Seventy percent of the teachers used only spoken language with their students, while 30% used both signed and spoken language. The FFL intervention group included 118 DHH children and the control group included 110 DHH children. Children' ages ranged from 3 to 6 years old (Mean age = 4 years, 3 months). Teachers in the intervention group used FFL for one hour a day throughout the school year. Teachers in the control group implemented their typical instruction. DHH children in the intervention group showed greater statistically significant gains on tests of phonological awareness, alphabetic knowledge, and word reading than children in the control group. Effect sizes were moderate to large. Children in both groups showed accelerated gains in vocabulary learning. At the end of the school year, the research team gave each intervention teacher a summative feedback survey. Eighty-four percent of the teachers returned the survey. Of those who replied, 95% of teachers said they enjoyed teaching FFL, felt their children

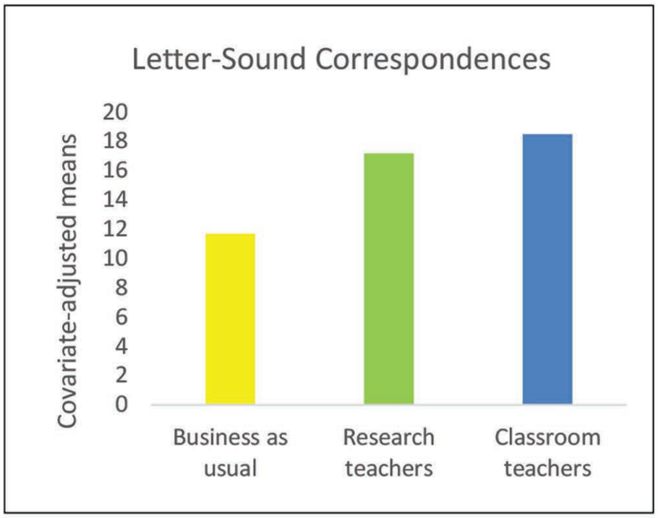
FIGURE 2. Fall to Spring Gain Scores across Three Instructional Contexts











Note. Covariate-adjusted means for each test were the resulting scores when spring scores were adjusted for fall scores. The three groups were children whose teachers delivered their business-as-usual instruction, children who received FFL from research teachers, and children who received FFL from classroom teachers.

PPVT (Dunn & Dunn, 2007); EOWPVT (Martin & Brownwell, 2011); TOPEL-PA (Lonigan, Wagner, & Torgesen, 2007); PAT (Robertson & Salter, 2007).

benefitted from FFL instruction, would recommend it to other teachers, and planned to continue using FFL the following year. Additionally, 62% taught classes with both DHH and TH children (some typically developing, others with disabilities) where all children received FFL instruction and 100% of those teachers agreed that their TH children in their classes benefitted from the FFL curriculum as well.

These studies provide strong evidence that FFL promotes the language and early reading skills of DHH children. Teachers reported enjoying implementing FFL. Teachers also indicated it is appropriate for TH children, so whole class implementation in inclusion classrooms is appropriate. These results also show that early literacy skills, including phonological awareness, letter-sound knowledge, and early decoding, are malleable skills in DHH children, despite their decreased access to sound and spoken language.

DESCRIPTION OF PUBLISHED VERSION OF FFL

Our team released FFL for commercial sale in the summer of 2017 (selling materials at cost). To ensure that FFL is implemented with fidelity, teachers must complete a 16-hour professional learning workshop to purchase it (Lederberg & Tucci, 2022). Hundreds of teachers from across the country have attended the FFL professional learning workshops offered in a variety of delivery models including virtual national trainings, in-person training at local schools, and state-wide trainings. Our trainers have worked with teachers in almost every state and many provinces in Canada. Virtual workshops allow our training team to reach teachers in historically underserved areas. Although only classroom-based teachers were included in our research, FFL is being adapted for use by itinerate teachers and speech pathologists.

We also have created alternative instruction for DHH children enrolled in bilingual (ASL-English) programs. This approach uses fingerspelling phonology and ASL-to-English bilingual strategies to support visual language and early reading skills. These strategies are embedded within the Teacher's Manual. Supplemental materials may be purchased separately.

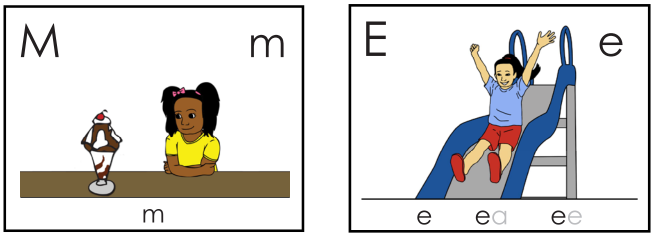
Teachers purchase a 398-page manual, along with all the instructional materials necessary to implement FFL (Lederberg et al., 2020). Figure 3 displays sample material used in FFL. The manual contains detailed lesson plans for 28 instructional units. Each unit consists of four one-hour lessons to be implemented within a week. During the first four weeks, teachers explicitly teach the instructional language needed to understand activities for the rest of the year. For example, students are taught the meaning of *same* versus *different* by sorting pairs of familiar objects into same and different categories. In another activity, students are taught the meanings for *sound* versus *name* through a farm animal activity where students learn the names of different farm animals (e.g., cow, pig, horse) and the sound the animals make (e.g., moo, oink, neigh). Children then identify what sounds are the same. These activities are foundational for later instruction in applying the concepts of same and different to letters and sounds.

The other 24 instructional units have a common structure where each unit is organized around a sound or phoneme (e.g., /m/, /ō/, /ē/, /b/). The target sound is embedded in a unique language experience story that anchors the unit. In the stories, Miss Giggle, an after-school teacher, and her three students, Pete, Kate, and Sue, experience a fun activity in which the target sound occurs. For example, in Unit 5, Pete, Kate, Sue, and Miss Giggle make and eat ice cream sundaes. They are so delicious that the children rub their tummies and say mmmm when they take a bite. Then Miss Giggle says, "Guess what? The letter m makes the same sound mmmm." Miss Giggle draws the letter m and Pete, Kate, and Sue takes turns pointing to the letter and making the /m/ sound.

Teachers begin the week by telling the language experience story using illustrative sequence cards and vocabulary picture cards. During the week, teachers and students retell the Miss Giggle story and participate in a three-day language activity sequence where the teacher and the students plan, do, and recall the same experience in the Miss Giggle story. The Miss Giggle story and related language activity sequence create a personally meaningful semantic association for the target sound (e.g., making sundaes and saying, "*mmmm* that's good!"; going down a slide and saying, "*eeee* that's fun!"). The activities also provide a fun context for children to engage in repeated practice in perceiving and producing individual sounds. Each story is accompanied by a large sound card that displays the associated letters and a picture representing the sound concept in the story (e.g., sundae for /m/, slide for /ē/; see Figure 3.) The picture serves as a visual mnemonic to support student recall of the target sound. Children learn to associate the multiple spellings for long vowels (e.g., e, ee, ea for /ē/). The picture is also used on small sound cards to represent the sound in subsequent reading activities (e.g., blending, decoding, encoding). The small sound card helps to make code-based instruction more transparent as there is a one-to-one correspondence between the sound and the picture on small sound cards (unlike using letters if there are multiple spellings for a sound). Using the small sound cards allows teachers to simplifying the task of identifying and blending sounds in a word. Over time, children transition from small sound cards to letter cards in reading activities.

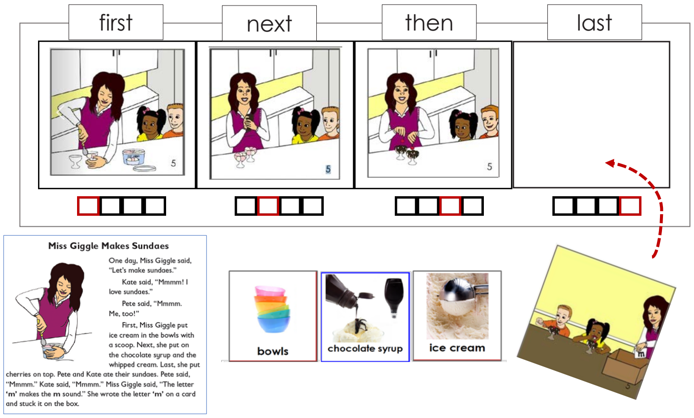
To support story understanding and story retell, teachers target 6-10 vocabulary words, which they embed in the Miss Giggle story and the three-day language experience.

FIGURE 3. Instructional Materials from FFL



Large Letter-Sound Cards

Small Sound Cards



Miss Giggle Makes Sundaes

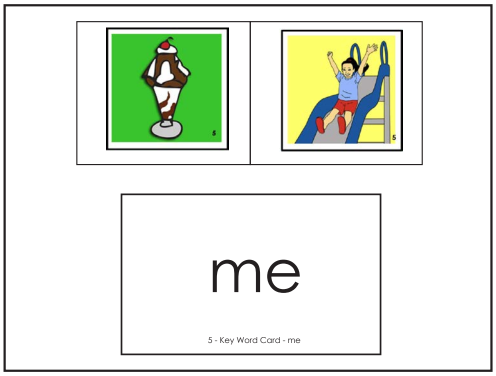
One day, Miss Giggle said, “Let’s make sundaes.”

Kate said, “Mmmm! I love sundaes.”

Pete said, “Mmmm. Me, too!”

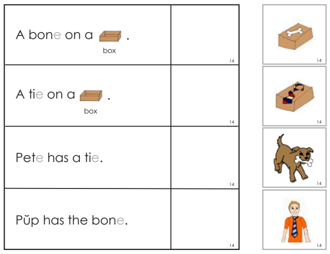
First, Miss Giggle put ice cream in the bowls with a scoop. Next, she put on the chocolate syrup and the whipped cream. Last, she put cherries on top. Pete and Kate ate their sundaes. Pete said, “Mmmm.” Kate said, “Mmmm.” Miss Giggle said, “The letter **‘m’** makes the **m** sound.” She wrote the letter **‘m’** on a card and stuck it on the box.

Miss Giggle Story and Vocabulary Picture Cards

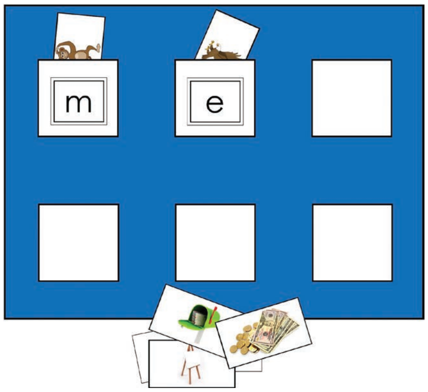


Key Word Blending (Make-a-Word Card)

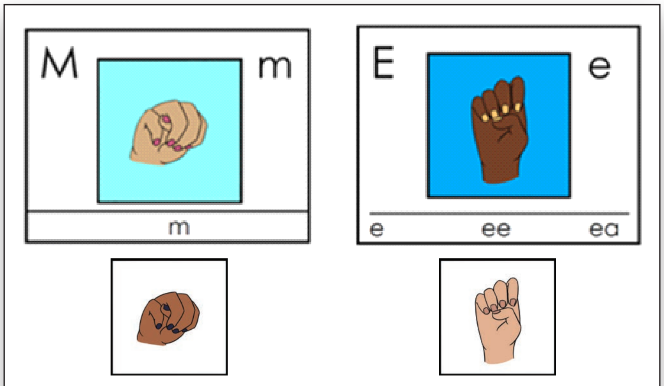
Reading Booklet



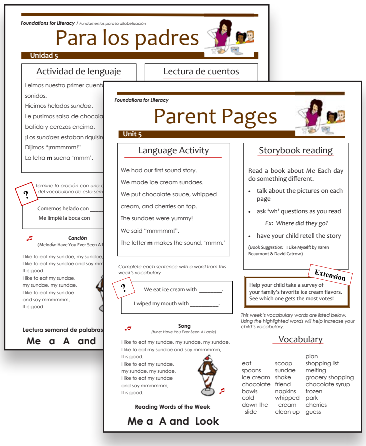
Reading Practice



Initial Sound Identification



Fingerspelling Supplemental Materials



Foundations for Literacy / *Fundamentos para la alfabetización*

Para los padres

Unidad 5

Actividad de lenguaje

Lectura de cuentos

Hicimos helados *sundae*.

batida y cerezas encima.

Dijimos “¡mmmmm!”

La letra **m** suena ‘mmm’.

Foundations for Literacy

Parent Pages

Unit 5

Language Activity

We had our first sound story.

We made ice cream sundaes.

We put chocolate sauce, whipped cream, and cherries on top.

The sundaes were yummy!

We said “mmmmm!”.

The letter **m** makes the sound, ‘mmm.’

Storybook reading

Read a book about *Me* Each day do something different.

- talk about the pictures on each page
- ask ‘wh’ questions as you read

  Ex: Where did they go?
- have your child retell the story

(Book Suggestion: I Like Myself! by Karen Beaumont & David Catrow)

Complete each sentence with a word from this week’s vocabulary

?

We eat ice cream with ________.

I wiped my mouth with __________.

Extension

Help your child take a survey of your family’s favorite ice cream flavors. See which one gets the most votes!

This week’s vocabulary words are listed below. Using the highlighted words will help increase your child’s vocabulary.

Song

(tune: Have You Ever Seen A Lassie)

I like to eat my sundae, my sundae, my sundae,
I like to eat my sundae and say mmmmmm,
It is good.
I like to eat my sundae,
my sundae, my sundae,
I like to eat my sundae
and say mmmmmm,
It is good.

Vocabulary

eat
spoons
ice cream
chocolate
bowls
cold
down the slide

scoop
sundae
shake
friend
napkins
whipped cream
clean up

plan
shopping list
melting
grocery shopping
chocolate syrup
frozen
park
cherries
guess

Reading Words of the Week

Me a A and Look

Parent Pages

Teachers select vocabulary appropriate for their students' language levels from unit-specific vocabulary lists, which include four levels of difficulty: core, target, challenge, and extension. Vocabulary picture cards are provided for the first three levels (see Figure 3). Teachers use evidence-based practices for receptive and expressive vocabulary learning such as using child-friendly definitions, picture support, and repeated opportunities to learn and use new words in meaningful contexts (e.g., Miss Giggle stories and three-day language experience activities) (Duncan & Lederberg, 2018; Schwanenflugel et al., 2010).

Each week teachers engage in activities centered around a decodable word or words that use taught sounds and their associated spellings. New words are introduced with a language activity that provides repeated opportunities to hear, see, and produce the decodable word. For example, after learning /m/ and /ē/, children play a question-and-answer game where the right answer is *me* (e.g., "Who has these eyes?" when shown a picture of the children's eyes). The activities ensure children have strong semantic (meaning-based) and phonological (sound-based) representation of the decodable word (Ehri, 2014) and prime them for the subsequent blending activities. Following the new word activity, teachers model identifying and blending the sounds of the new word using small sound cards (e.g., saying "me, /m/ /e/, me").

After decodable words are introduced and practiced several times using picture support, children spend time reading printed words in isolation and in connected text. These activities target phonological awareness, word reading, and reading comprehension. Reading decodable words in isolation and in connected text provides repeated opportunities to segment and blend the sounds of a word. Research suggests that children learn sound-level phonological awareness skills better when instruction includes letters (Shanahan & Lonigan, 2010), likely because letters serve as visual support for hard-to-discriminate sounds. In addition to reading words in isolation, children read connected text composed of explicitly-taught decodable words and rebuses (i.e., pictures that take the place of words that children cannot recognize through sight or through decoding; see Figure 3). Teachers use reading materials to support their students' reading comprehension by probing children's understanding through pictures and questions.

Teachers also explicitly teach syllable segmentation, initial sound identification, and rhyming. These activities frequently use the vocabulary from previous units to ease the language burden so that students can focus their attention on phonological awareness. Daily practice activities of previously taught skills include reviewing letter-sound correspondences, letter(s)-sound correspondences fluency charts, reading connected text, and phonological awareness activities. These are incorporated into every lesson to ensure students have enough opportunities to build skill mastery.

Teachers further reinforce complex language and vocabulary through daily storybook reading using dialogic reading techniques (Fung et al., 2005; Shanahan & Lonigan, 2010). Teachers select a book that connects to the unit theme and identify 6 to 10 novel vocabulary words contained in the storybook based on their students' language knowledge. They read the story and then revisit the story three or four times throughout the week. Each day the teacher emphasizes targeted vocabulary and increases students' active engagement by asking questions, expanding children's answers, and prompting students to provide more detailed answers and discussions.

FINAL TIPS FOR IMPROVING PHONOLOGICAL AWARENESS, ALPHABETICAL KNOWLEDGE, AND VOCABULARY

Phonological Awareness

- Provide intensive, explicit, and extensive instruction to support phonological awareness skills which takes months to develop.
- Use familiar vocabulary in phonological awareness instruction so the children can focus their attention on the structure of words, rather than their meaning.
- Use letters to facilitate learning to isolate initial sounds of words. For example, once children know the sounds of two letters (e.g., /m/ and /p/) have children sort words (mop, papa) into two categories using the letters m and p as visual support for the two sounds (see Figure 3).
- Teach rhyming through the identification of pairs of familiar words that rhyme and pairs that do not. It is critical to include examples of words that do not rhyme for contrast. Make this a listening activity because pictures tend to distract DHH children in this activity. Exposure to rhyming books is insufficient to teach children to rhyme.

Alphabetic Knowledge and Early Reading

- Teach children to associate sounds with letters by focusing on those sounds in isolation. Sounds are typically taught through reviewing what words start with that letter (e.g., It is M week. What words start with the letter M?--milk, mama). This requires children to isolate the letter from these words. Instead, we suggest focusing on the sound (e.g., This is /m/ week). Embed individual sounds in stories that allow children to hear and say a sound in isolation and then associate it with a letter. Those letter-sound correspondences can then be used to identify initial sounds in familiar words.

- Include opportunities to read simple words during reading instruction. Follow learning letter-sound correspondences with sounding out and blending those sounds in decodable words. This shows children why they are learning the sounds of letters and gives them practice in the important phonological awareness skills of segmenting and blending sounds. Because this is an advanced skill, teachers should model sounding out the letters and blending them into a word. Children should practice reading the same words throughout the year. Preschool children may need continuous teacher modeling to begin to read decodable words.

Vocabulary

- Teach five to 10 words a week explicitly. Instruction should include child-friendly definitions and visual or kinesthetic support to clarify meaning (e.g., pictures, objects, and acting out).

- Expand on children's utterances by repeating them with the addition of at least one or two words or correcting syntactic mistakes.

- Read language-appropriate stories using dialogic reading techniques. Books create a meaningful context for children to acquire and practice using new words. Prior to reading the story, identify targeted vocabulary and create child-friendly definitions and picture cards. Embed these definitions and picture supports while reading the story. Revisit the same book three to four times, encouraging the students to engage in more dialogue with each reading.

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The Road to Hearing Health Equity

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Of the 1.6 billion people with hearing loss worldwide, over 1.2 billion live in underserved communities with the least access to hearing care. This includes preventable hearing loss in an estimated 42 million children who, without early identification and treatment, are at increased risk of significant detrimental effects that can last a lifetime. These immense hearing health disparities provide a clear and compelling call to action. The new Center for Hearing Health Equity at the University of Arkansas for Medical Sciences is the first of its kind in the world, established to deliver the critical mission of fostering hearing health equity both globally and locally. This article describes the Center's approach, which is built on inclusive, multidisciplinary collaboration guiding a research-driven, public-health oriented agenda to develop evidence that is directly translatable to policy change. The end goal is to generate momentum towards sustained solutions that will create a new future with hearing care accessible to all.

There are an estimated 1.6 billion people living with hearing loss worldwide. This means that out of the entire global population, a staggering one in five people experience hearing loss. The numbers of people affected are expected to continue to grow, and by 2050 one in four people are projected to have hearing loss (World Health Organization, 2021). Not only is the scale of hearing loss one of the globe's major health challenges, the burden is also inequitably distributed. Eighty percent of affected individuals live in underserved communities, specifically in low- and middle-income countries, or within underserved communities in high-income countries (Figure 1) (World Health Organization, 2021). As a result, the populations most likely to be affected by hearing loss are also the least likely to have easy access to hearing care.

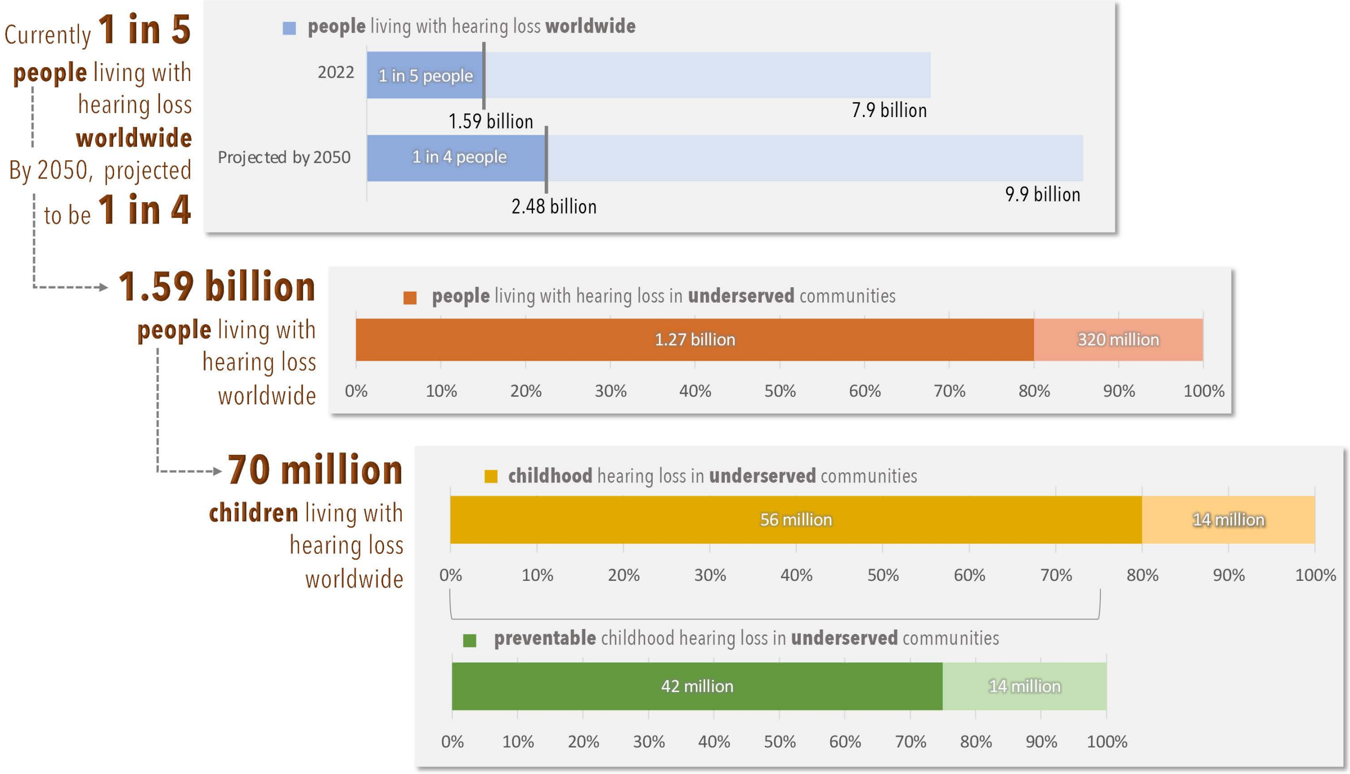
Hearing loss in children is of particular concern given the lifelong consequences. Children with hearing loss frequently experience speech and language delays, poor academic performance, and limited vocational opportunities (Emmett & Francis, 2015; Järvelin et al., 1997; Khairi Md Daud et al., 2010; Tomblin et al., 2015; Woodcock & Pole, 2008). Hearing loss is also associated with lower quality of life and worse behavioral outcomes (Rolland et al., 2016; Wang et al., 2019). Importantly, the World Health Organization estimates that up to 60% of childhood hearing loss is preventable. This estimate rises to 75% in underserved communities, such as in low- and middle-income countries, where the majority of childhood hearing loss is related to ear infections (World Health Organization, 2021).

The scope of hearing health disparities is immense, with more than 1.2 billion people living with hearing loss from underserved communities with the least access to hearing care. This includes preventable hearing loss in an estimated 42 million children, who, without early identification and treatment, are at increased risk of significant detrimental effects that can last a lifetime (see Figure 1).

CENTER FOR HEARING HEALTH EQUITY AT THE UNIVERSITY OF ARKANSAS FOR MEDICAL SCIENCES

As members of the hearing health community, we have a clear and compelling call to action to address these disparities. The new Center for Hearing Health Equity at the University of Arkansas for Medical Sciences is the first of its kind in the world, established to deliver the critical mission of fostering hearing health equity both globally and locally. The philosophy driving our new Center's approach is grounded in several key attributes that work collectively to overcome inequities. First, our approach is multidisciplinary and all-embracing, encompassing collaboration across disciplines, backgrounds, countries, and cultures. This is a global problem that requires global solutions. We believe it is critical to adopt an inclusive mindset that values the input not only of healthcare professionals but also educators, parents, community members, and individuals living with hearing loss themselves. Our approach is research- driven with a specific focus on developing evidence that is directly translatable to policy change. A public health perspective is necessary given the scale of the problem, which

FIGURE 1. Scope of hearing health disparities globally. Estimates derived from the World Health Organization World Report on Hearing.



will not be solved with a traditional clinical mindset of caring for one patient at a time. Finally, our approach is multifaceted and addresses hearing loss from multiple angles.

COMPONENTS OF THE APPROACH

Collaborative

Collaboration is first and foremost for achieving hearing health equity. Our approach at the Center for Hearing Health Equity is rooted in team science, recognizing that accelerated scientific advancement is possible when we invest the time and energy to work in large collaborative teams (Committee on the Science of Team et al., 2015). In 2018, we launched the Global HEAR (Hearing Loss Evaluation, Advocacy, and Research) Collaborative, the only international research network dedicated to reducing disparities in hearing loss worldwide. The Global HEAR Collaborative has grown to now include collaborators from 28 countries (Figure 2). Collaborators' backgrounds include otolaryngology-head and neck surgery, audiology, and speech pathology. In 2021, HEAR-USA began as the United States branch of the HEAR Collaborative focused on addressing hearing loss disparities in underserved rural and minority populations across the United States. We now have three grant-funded initiatives running through these collaboratives, including two randomized trials of school-based telehealth to improve access to hearing care for rural children and a device development project to improve screening for infection-related hearing loss. Our scientific teams also involve experts from multiple other disciplines beyond the hearing fields, such as implementation science and education research. Collaborative initiatives such as HEAR represent the future of hearing health equity, providing an opportunity for individuals across disciplines and countries to partner on large international and domestic multi-site projects that would otherwise not be possible.

Equally vital to the collaborative effort to address disparities is partnership with community stakeholders. At the UAMS Center for Hearing Health Equity, we ground our research in community partnership and feel these relationships are indispensable for generating truly implementable and sustainable policy change. Examples of community engagement include developing stakeholder teams and community advisory boards to inform and guide community-based research. Using mixed methods to incorporate qualitative investigation alongside quantitative randomized controlled trials is another technique we frequently employ to facilitate integration of community input into the research process (Patterson et al., 2022; Robler et al., 2020).

FIGURE 2. Global HEAR Collaborative sites.

Research-Driven

Maintaining a research-driven approach is also critical for achieving hearing health equity. There are many questions in hearing loss to which we do not yet know the answers, and filling these gaps in the evidence can provide actionable solutions to enact policy change. Our Center for Hearing Health Equity focuses on the big picture questions that could ultimately move the needle to address disparities. By working together in large collaborative teams, we will be able to answer these pressing questions and develop the evidence needed to influence policy change. This approach provides an avenue for community partners and healthcare professionals who provide patient care full-time to participate in large-scale initiatives that that will ultimately lead to elimination of inequities in hearing care.

Public Health-Oriented

In addition to being research-driven, we believe a public health lens is essential to successfully address hearing health disparities. The sheer numbers of people affected—20% of the entire global population—dictate that we develop innovative ways to provide hearing care that focus on population-level innovations. Technology can also undergird these efforts, such as supporting task sharing with community health workers by applying artificial intelligence and machine learning techniques for diagnostic support.

Multi-Faceted

New Solutions for Prevention

A multifaceted perspective spanning prevention, identification, and treatment is an essential component of this research-driven, public health approach to hearing health equity. Particularly in children, prevention is critical to addressing disparities because of the vast numbers affected with preventable hearing loss: currently estimated at 42 million children. To be able to prevent hearing loss from developing in the first place, we need to understand why it is so common in underserved communities. Specifically, what are the etiologies that are unique to low resource settings, and what can be done about them?

Two examples, one from Nepal and another from Alaska, illustrate this concept. It is well known that the majority of childhood hearing loss in underserved populations is related to ear infections. What is less understood is why these infections happen, and what the modifiable risk factors are that can be altered to reduce the risk of infection and therefore reduce the risk of developing hearing loss. In the Gangetic flood plain of South Asia where micronutrient deficiencies and undernutrition are common, children with a history of ear infections who received vitamin A supplementation as preschool children were 54% less likely to have hearing loss as young adults than those with ear infections who did not receive supplementation. This evidence from a follow-up study of a large randomized trial cohort has very practical implications: supplementation with a 2-cent vitamin A tablet every six months in early childhood resulted in a 7% absolute reduction in hearing loss among individuals who had a history of ear infections (Schmitz et al., 2012). In Alaska, our analysis from a randomized trial cohort revealed that middle ear disease is more common in children who live in homes without running water (Hicks, 2022). Addressing water availability exemplifies another public health measure that could be used to reduce the burden of ear infections and may ultimately reduce childhood hearing loss in underserved communities.

Improving Identification

In addition to prevention, improving identification is also an essential element of a multifaceted, research-driven strategy to address disparities in hearing loss. For example, school screening is an important public health measure to identify children with previously undiagnosed hearing loss. However, evidence-based guidelines for the optimal protocols to conduct screening are scarce, and loss to follow-up after screening programs is a universal problem. During a recent randomized trial in Alaska, we addressed two key unanswered questions about identification: 1) how do we best identify children with infection-related hearing loss, and 2) how do we address the longstanding problem of loss to follow-up (Emmett, Robler, Gallo, et al., 2019; Emmett, Robler, Wang, et al., 2019)? This trial, which tested mobile health screening and telemedicine referral for rural schools, found that the most accurate screening protocol in an environment with frequent ear infections included not only pure-tone screening, which is commonly used for screening programs, but also tympanometry, which assesses the health of the middle ear. For the perennial challenge of loss to follow-up, the trial found that children randomized to telehealth specialty referral were more than twice as likely to receive follow-up after school screening, and follow-up occurred more than 17 times faster with telemedicine specialty referral than with standard primary care referral (Emmett et al., 2022). These results have important policy implications for improving access to care for rural children, providing clear evidence for the most accurate screening protocol in environments where ear infections are common and a potential solution using telemedicine to improve entry to the healthcare system after screening.

Expanding Access to Treatment

Lastly, a multifaceted perspective would not be complete without addressing treatment. Despite the many unanswered questions around hearing loss, there are nevertheless known treatments that are highly effective. For example, cochlear implantation (CI) has been clearly shown to improve speech and language development as well as improve academic and vocational outcomes in children with severe to profound hearing loss, yet this proven technology is not accessible to the majority of the world's population. In fact, it is estimated that less than 1% of CI candidates globally currently have implants (Bodington et al., 2021). Through the Global HEAR Collaborative, we questioned this gap and sought to address one of the key questions holding up access: the high cost. For many years, a common assumption around cochlear implantation was that the technology would only be available in high resource settings because of cost constraints. Cost-effectiveness in underserved settings had not been evaluated, and filling this gap in the evidence was a critical first step to support policy change for expanding access to CIs. Through a series of analyses spanning 21 countries in Sub-Saharan Africa, Latin America, and Asia, we determined that pediatric cochlear implantation can be cost-effective in underserved settings (Emmett, Sudoko, et al., 2019; Emmett et al., 2016; Emmett et al., 2015). This type of evidence provides ministries of health with essential information to support incorporating cochlear implantation into national health plans.

Mentorship-Focused

Returning to the importance of inclusivity, learners and trainees at all levels of education are vital contributors to achieving hearing health equity. Mentorship of the next generation and fostering long-lasting commitment to addressing disparities are key elements of our approach at the UAMS Center for Hearing Health Equity. From undergraduates to medical students and surgical residents, trainees are involved in all of our projects and bring appreciated energy and perspective to the team.

Advocacy-Centered

Finally, advocacy ties all aspects of our approach together, serving as the connector between new evidence generated and the achievement of hearing health equity. Policy change is needed for the work that we do collectively to have impact, and engaging in advocacy is how the hearing health community can best facilitate policy change. This can happen at multiple levels, from local to global. For instance, many of our local stakeholders serve in state leadership and hold significant influence on the direction of public policy. Involving government leadership from inception of a project provides valuable perspective on the questions that most urgently need to be answered, as well support for scaling and sustainability after the scientific work is complete. On the international level, an example of advocacy is the World Health Organization World Report on Hearing, which involved significant investment by many members of the hearing health community and can serve as an invaluable tool for advocacy at the individual country level.

GOAL OF GENERATING SUSTAINABLE CHANGE

Despite the immense disparities facing us in hearing health globally, achieving hearing health equity is possible. Each of the components of our approach contribute to the ultimate goal of generating sustainable change in the landscape of hearing health disparities. At the UAMS Center for Hearing Health Equity, we are excited to partner with multidisciplinary and community collaborators, leading innovative research initiatives designed to generate momentum towards sustained solutions that will create a new future with hearing care accessible to all.

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