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HEARING LOSS IN CHILDREN (D HORN AND H OU, SECTION EDITORS)

Unilateral Hearing Loss and Single-Sided Deafness in Children: an Update on Diagnosis and Management

Patricia L. Purcell[1] · Sharon L. Cushing[1,2,3,4] · Blake C. Papsin[1,2,3,4] · Karen A. Gordon[1,2,3,4]

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Abstract

Purpose of Review This review highlights our current understanding of the impact of unilateral hearing loss and single-sided deafness in children, offering insight into diagnosis and management.

Recent Findings Children with unilateral hearing loss develop an aural preference toward their better hearing ear, leading to difficulties with spatial navigation, balance, speech/language skills, and quality of life when compared with typical hearing peers. Cochlear nerve aplasia and cytomegalovirus are among the most common etiologies for unilateral hearing loss, which has important implications for treatment. Hearing rehabilitation relies upon early correction of hearing loss with an appropriate auditory prosthesis. In children with single-sided deafness, cochlear implants are the only intervention that potentially offers restoration of bilateral hearing, and studies continue to refine candidacy protocols.

Summary Unilateral hearing loss has important consequences for children. Recent studies emphasize the importance of early diagnosis and investigate ways to appropriately restore bilateral hearing in these children.

Keywords Single-sided deafness · Unilateral hearing loss · Development · Bilateral/binaural/spatial hearing · Etiology of hearing loss · Cochlear implant

Introduction

Unilateral hearing loss (UHL) is relatively common, affecting approximately 1 of every 2000 newborns and at least 3% of school-aged children [1–3]. Previous studies have found that children with UHL have higher rates of grade failure [4] and lower scores on speech/language and intelligence quotient testing [5–7] when compared with children with typical hearing.

There has been growing interest in improving our management of these children [8]. Studying the impact of UHL can be challenging, as clinicians and researchers have often found themselves relying on testing measures that were developed with bilateral hearing loss in mind. In recent years, hearing health providers have gained better understanding of the unique challenges faced by children who have asymmetric access to sound [9, 10]. For example, children spend much of their day in the complex listening environment of the classroom, which often involves group conversations and background noise; therefore, educators are being encouraged to improve auditory access for children with UHL through strategic seating, visual aids, and remote microphone systems [11]. Hearing health providers have also gained better appreciation of the various etiologies of UHL due to improvements in the utilization of brain and temporal bone imaging [12] and

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✉ Karen A. Gordon
karen-a.gordon@sickkids.ca

Patricia L. Purcell
patricia.purcell@sickkids.ca

Sharon L. Cushing
sharon.cushing@sickkids.ca

Blake C. Papsin
blake.papsin@sickkids.ca

[1] Department of Otolaryngology, Head & Neck Surgery, Hospital for Sick Children, University of Toronto, 555 University Avenue, Toronto, Ontario M5G 1X8, Canada

[2] Archie's Cochlear Implant Laboratory, Hospital for Sick Children, University of Toronto, Toronto, Canada

[3] Department of Communication Disorders, Hospital for Sick Children, University of Toronto, Toronto, Canada

[4] Institute of Medical Sciences, University of Toronto, Toronto, Canada

testing for congenital cytomelagovirus (CMV) [13]. More rarely, genetic testing may sometimes be helpful, particularly in children suspected of having syndromic hearing loss such as Waardenburg syndrome [14]. There are also reports of children with GJB2 mutations presenting with UHL [15]. It is probably reasonable to consider genetic evaluation in children who have normal imaging and do not have congenital CMV.

This increased awareness has promoted efforts to provide bilateral hearing to children with UHL through hearing devices. Children with single-sided deafness (SSD) have severe-to-profound UHL with unaidable hearing in the affected ear. Cochlear implants have been provided to children with SSD with reported benefits [16–19], but studies are ongoing to refine candidacy criteria, such as age in children with congenital loss [20]. This review summarizes recent papers that highlight our current understanding regarding the impact of UHL loss and offer insight into management, with particular emphasis on early diagnosis and prompt restoration of bilateral hearing as soon as possible using the most appropriate intervention and/or device.

The Consequences of Unilateral Hearing Loss

Studies have suggested that UHL negatively impacts spatial navigation, balance function, speech/language skills, and quality of life [21]. To better understand why this occurs, it is helpful to discuss the importance of binaural hearing in children [22, 23]. Binaural hearing provides us with a sense of the world in all 360 degrees around us. Time and level differences between the two ears are detected in the auditory brainstem and midbrain and used to code the spatial location of the sound source [24]. Children with monaural hearing do not have access to these binaural cues, and thus cannot make use of spatial hearing to distinguish between multiple sounds in the environment by their distinct spatial locations. Their ability to localize sounds is particularly poor when the sound comes from the side of the poorer hearing ear [25]. In addition, children with monaural or asymmetric hearing loss rely primarily on their better hearing ear, which leads to an aural preference for that side mediated by abnormal strengthening of auditory pathways [26, 27]. In effect, this creates a large "head shadow" which might improve signal-to-noise ratios when the target sound is facing the better ear but has the opposite effect (poorer signal-to-noise) when the target sound is facing the more impaired ear.

The importance of spatial hearing in development is shown by the deficits experienced by children with UHL who do not have access to accurate binaural cues. Without normal spatial hearing in early life, children with UHL show difficulties discriminating speech in noise, understanding speech when it is not directed toward their better-hearing ear, and navigating group conversations [28]. The over-representation of the hearing ear within the auditory system during development, a condition known as "aural preference syndrome" [29], may occur to support speech and language development but simultaneously deteriorates spatial hearing and the potential to restore it later [30].

Difficulties with spatial navigation occur due to UHL alone; however, these problems are often compounded by the fact that children with UHL have higher rates of vestibular and balance impairment [31]. Children with hearing loss and vestibular impairment demonstrate poorer balance skills than typically hearing peers [32]. A comparison of 14 children with UHL and 14 children with typical hearing found that the children with hearing loss demonstrated significantly lower scores on the Bruininks-Oseretsky Test of Motor Proficiency-2 (BOT-2) [33]. The BOT-2 is a standardized test of both static and dynamic balance function, comparing a child's performance with age-standardized norms [34]. In addition, assessment of vestibular end-organ function (otoliths and horizontal canal) in children with UHL found that more than half of the children demonstrated functional abnormality [35]. The dysfunction was more commonly associated with the worse hearing ear.

Without the benefit of binaural cues, listening tasks may require greater mental effort. A meta-analysis found that children with UHL demonstrated lower scores on both the Social and School domains of the PedsQL, a well-validated generic quality of life instrument for children, when compared with typical hearing peers [36]. Researchers also evaluated quality of life using an instrument developed for youth with hearing loss, the HEAR-QL [37]. They found that both children with unilateral sensorineural hearing loss (USNHL) and those with bilateral loss reported a worse quality of life than children with typical hearing on HEAR-QL [38]. While there was not a statistically significant difference between children with unilateral versus bilateral hearing loss, the authors note that the mean HEAR-QL score was > 10 points higher (better) for children aged 7 to 12 years with USNHL when compared with children in the same age group with bilateral loss; this difference was not statistically significant, possibly due to limited sample size.

Children with UHL also seem to experience difficulties with auditory, social, and behavioral tasks. These differences can be detected in early childhood. Using parent questionnaires, Kishon-Rabin found that 21% of children with UHL (median age 9.4 months) demonstrated delays in early auditory skills and 41% demonstrated delays in preverbal vocalizations when compared with normal hearing peers [39]. Fitzpatrick et al. found that children with congenital UHL performed more poorly than children with typical hearing in functional auditory listening and in receptive and expressive language skills at age 48 months [40]. In a separate study, children with UHL were also found to have lower functional auditory skills on both the Parents' Evaluation of Aural/Oral performance of Children (PEACH) and the Teachers' Evaluation of Aural/Oral performance of Children (TEACH) when compared with normative means for typically hearing peers [41].

Advances in Diagnostic Testing

Universal newborn hearing screening has decreased the average age at diagnosis of UHL significantly [42]. In 2013, a prospective cohort study spanning 20 years at Children's Hospital of Eastern Ontario found median age at diagnosis of UHL dropped from 5.3 to 0.3 years after implementation of hearing screening, but it also raised some concern that children with UHL were less likely to undergo early trial of amplification than children with mild bilateral hearing loss [43]. In 2017, a follow-up study suggested a possible shift in practice, perhaps due to increasing awareness of the benefit of binaural hearing and evidence of reduced ability to restore hearing from the poorer ear after a long period of unilateral hearing [44]. In the more recent study, more than 70% of children with UHL eventually received a recommendation for amplification [45]. Importantly, the study also highlighted the importance of monitoring hearing loss over time as 42% of children with UHL demonstrated some degree of progression, with 17% eventually developing bilateral hearing loss [45]. Progression of hearing loss likely depends on the etiology. A study of 128 children with UHL found that 33% demonstrated progression and that children with temporal bone anomalies, including cochlear nerve canal stenosis, may be at greater risk of hearing loss progression in the impaired ear than children with normal temporal bone imaging [46]. Children with CMV-associated UHL are at risk of progressive hearing loss in both ears. One recent study found that 46% of children with UHL associated with symptomatic CMV developed hearing loss in their contralateral ear [47].

As management options expand, increasing importance has been placed on determining the underlying etiology of hearing loss [48], as etiology may have important implications for prognosis and treatment. Studies have consistently found temporal bone imaging to be of high yield in children with UHL, with approximately 30 to 50% of children having radiographic abnormalities of clinical significance [49–51]. One of the most common abnormalities is cochlear nerve hypoplasia/aplasia, often associated with cochlear nerve canal stenosis and other cochleovestibular anomalies; cochlear nerve canal width has been found to strongly correlate with cochlear nerve status, such that a width < 1.7 mm identifies cochlear nerve deficiency with 84% sensitivity and 98% specificity [52]. A recent study compared CT temporal bone imaging of children with unilateral cochlear nerve canal stenosis (< 1.0 mm) and found both the ipsilateral and contralateral cochleovestibular apparatus to be smaller in size than normal controls [53]. The presence of cochlear nerve hypoplasia/aplasia has important implications for auditory rehabilitation. Children with cochlear nerve canal stenosis have been found to have impaired speech discrimination, independent of degree of hearing loss [54], and children with cochlear nerve hypoplasia have poor outcomes with cochlear implantation [55]. Otolaryngologists must obtain appropriate imaging to confirm the status of the nerve prior to counseling families about the option of cochlear implantation.

Some controversy remains regarding the role of magnetic resonance imaging (MRI) versus non-enhanced high-resolution computed tomography (CT) in diagnostic imaging. A recent meta-analysis by Ropers et al. based at Leiden University Medical Center in the Netherlands reported the pooled prevalence of inner ear abnormalities on imaging in children with USNHL [56]. The authors found the pooled yield of radiographic abnormalities to be 37% (95% CI, 25–48%) for CT and 35% (95% CI, 22–49%) for MRI; cochleovestibular abnormalities were found with pooled frequency of 19% (95% CI, 14–25%) for CT and 16% (95% CI, 7–25%) for MRI, while cochlear nerve deficiency or cochlear nerve canal stenosis was found with pooled frequency of 16% (95% CI, 3–29%) on MRI and 44% (95% CI, 36–53%) on CT, respectively.

A multi-institutional study compared the diagnostic yield of CT and MRI in 219 children with USNHL. The authors found imaging abnormalities in 42.7% of children who underwent MRI compared with 36.7% of children who underwent CT; importantly, they found no significant difference in diagnostic yield among the 65 children who underwent both imaging modalities [12]. These studies suggest little difference in yield of imaging modality for some anomalies, lending support to the idea that surgeon preference and resource availability often play a role in determining which type of imaging is performed. However, MRI is the only modality that can definitively assess the status of the cochlear nerve. MRI may also detect imaging findings suspicious for CMV, such as temporal lobe cysts or white matter changes [57]. We have outlined an approach that includes MRI for all children being assessed for cochlear implant candidacy with CT being added for cases at risk of being more surgically challenging [58].

Congenital CMV is becoming well recognized as an etiology of UHL because USNHL predominates in children with hearing loss associated with the "asymptomatic" form of the disease [59]. Children with CMV-associated UHL are at risk of progressive loss and should be monitored over time [60]. The gold standard for diagnosis of congenital CMV is through PCR testing of urine to detect CMV DNA within the first 3 weeks of life [61], so providers must have a high index of suspicion very early in a child's life. Saliva PCR testing can also be performed as this may be easier to obtain from a newborn; however, a positive result must be confirmed with urine PCR due to risk of false positive result from CMV present in breastmilk.

Some institutions have initiated "hearing-targeted" CMV testing, in which a newborn who refers on hearing screen will then undergo CMV PCR testing if they are < 3 weeks of age [62]. The Utah Department of Health was the first to publish results from its first 24 months of targeted screening, diagnosing 14 newborns with asymptomatic CMV out of 509 infants

who never passed a hearing screen [63]. Importantly, this study found that a statewide targeted screening program is feasible. There are currently hospitals in 18 states participating in targeted CMV screening [64].

Concerns have been raised that targeted screening will miss children with asymptomatic congenital CMV who have normal hearing at birth and develop hearing loss in a delayed fashion [65]. Yale New Haven Health System recently published their experience with targeted screening and found it to be low yield, only identifying 3 newborns with positive saliva PCR testing out of 10,964 live births and 171 referred hearing screens [66]. One of the newborns was found to be negative on confirmatory testing. Based on these concerns, the Province of Ontario became the first region in North America to initiate universal newborn screening for congenital CMV based upon dried blood spot PCR. While the sensitivity of dried blood spot PCR is lower than that of urine or saliva PCR, it is more feasible to carry out at the population level because it is already being collected for other forms of newborn screening. The results from this program will be monitored closely over the coming years.

Treatment Recommendations

Hearing rehabilitation in children with UHL centers upon early identification of hearing loss and correction of the asymmetry of hearing by utilizing the appropriate auditory prosthesis. Table 1 lists the various types of auditory prostheses available, and the indication for usage.

Early recognition is particularly important for children with congenital hearing loss who are at risk of developing aural preference that will be resistant to treatment over time [67]. For example, children who receive conventional hearing aids later in childhood do not perceive the same bilateral benefit as those who were fit at a younger age [68]. In similar fashion, children with bilateral hearing loss who are implanted sequentially with long delay continue to demonstrate preference toward the first-implanted ear in the auditory cortices years after they receive their second implant [69].

In 2017, Appachi et al. published a systematic review of studies evaluating the use of auditory prostheses among children with UHL [70]. The review focused on the evidence for frequency modulating (FM) systems, conventional hearing aids, CROS devices, and osseointegrated bone conduction implants. While still limited, there is evidence to support the use of osseointegrated bone conduction implants in children with moderate to profound USNHL; osseointegrated implants were shown to improve objective audiometric outcomes in all seven studies that were reviewed, including pure-tone averages, speech reception thresholds, and Hearing in Noise Test (HINT) scores. For the non-surgical devices, FM systems seem to offer an educational benefit in the classroom. Results were more mixed for the other devices, with conventional aids perhaps showing some benefit in functional measures among children who are candidates. Though they may confer benefit, percutaneous osseointegrated bone conduction implants are also associated with frequent complications in children related to skin overgrowth, infection, and osseointegration failure, so families should be counseled accordingly [71, 72].

Table 1 Indications for auditory prosthesis for pediatric unilateral hearing loss

Type of hearing loss	Auditory prostheses
- Conductive	- Behind-the-ear hearing aid
	- Bone conduction device
	o Non-implanted (band or adhesive-retained)
	o Osseointegrated implant
	▪ Percutaneous
	▪ Passive transcutaneous
	▪ Active transcutaneous
- Sensorineural (mild-to-moderately severe)	- Behind-the-ear hearing aid
- Sensorineural (single-sided deafness)	- Contralateral routing of signal (CROS) device
	- Bone conduction device
	o Non-implanted (band or adhesive-retained)
	o Osseointegrated implant
	▪ Percutaneous
	▪ Passive transcutaneous
	▪ Active transcutaneous
	- Cochlear implant (the only prosthesis that allows for bilateral access to sound)

For children with SSD, cochlear implantation offers the only means for restoration of binaural hearing. Several studies on this subject have been carried out at The Hospital for Sick Children in Toronto, Ontario. Polonenko et al. demonstrated a correction of aural preference and restoration of bilateral auditory input to the appropriate cortices among five fairly homogenous single-sided implant candidates who were 3 years of age and younger at implantation [73]. Ganek et al. evaluated datalogs from follow-up clinical audiology appointments for 23 children who had received an implant with limited durations of SSD and found an average usage rate of 6.22 h per day. There were no significant differences in implant use among this group of children and adolescents based upon age or duration of implant experience [74].

At a separate institution, the University of Wuerzburg in Germany, Ehrmann-Mueller et al. evaluated pre- and post-implantation audiometric measures including speech-in-noise testing and sound localization tasks among 7 children who were implanted between the ages of 3 and 16 years. All of the children demonstrated significant hearing improvements and were consistent device users [75]. A limitation of this study was that the etiology and duration of unilateral deafness were not known for many of the children.

Deep et al. at the New York University School of Medicine recently published a case series of 14 patients who received an implant for SSD [76]. The mean duration of deafness prior to implantation was 3 years. In 8 patients with at least 1-year post-CI follow-up, the mean word recognition score in the CI-only condition was 56%, which was a significant improvement from baseline. In addition, the patients scored as well or better with the CI-on versus CI-off in all speech-in-noise conditions, indicating limited to no interference from the CI.

Though there are studies supporting the benefit of cochlear implantation in children with SSD, it remains important to carefully screen potential candidates and counsel families regarding appropriate risks and expectations of surgery. A retrospective analysis of all potential cochlear implant candidates with SSD at The Hospital for Sick Children found that 61% ultimately did not receive an implant. Thirty percent were excluded due to cochlear nerve aplasia, 29% were not implanted based upon familial preference to avoid surgery, and the remaining 2% were not candidates based upon hearing thresholds or duration of deafness [77]. Children who underwent implantation were more likely to have been diagnosed with congenital CMV, with its recognized risk for progressive bilateral hearing loss, or have experienced sudden loss of hearing.

Conclusion

Recent studies provide further support for the consequences of UHL on many aspects of childhood development, including spatial awareness, speech/language skills, and quality of life. Various factors, such as characteristics of hearing loss, underlying etiology and familial preference, all play a role in determining appropriate methods for evaluation and management of this condition.

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