Current Otorhinolaryngology Reports (2020) 8:267–275
https://doi.org/10.1007/s40136-020-00298-3

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

Implications of Concurrent Vestibular Dysfunction in Pediatric Hearing Loss

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Published online: 9 June 2020

Abstract

Purpose of the Review It is known that the prevalence of vestibular impairment in children with sensorineural hearing loss (SNHL) is high and can lead to balance deficits. In this review, we look beyond balance and consider the impact of this multisensory deficit on neurocognitive function and navigation with the aim of explaining some of the variability in outcomes seen in cochlear implant populations, considering how to ameliorate these outcomes with targeted rehabilitative strategies.

Recent Findings Congenital or early acquired vestibular impairment associated with SNHL impacts multiple cognitive domains including spatial memory. The attentional demands of maintaining postural stability are also significant and receive priority over other competing tasks, leading to a broader impact in everyday life.

Summary Vestibular impairment is common in children with SNHL and impacts their daily function. Early recognition of vestibular deficits is key and several promising therapeutic approaches, including the restoration of bilateral and potentially binaural hearing, are currently under investigation.

Keywords Vestibular impairment · Hearing loss · Cochlear implant · Balance · Postural stability · Development

Introduction

The longstanding history of excellence in research examining the prevalence of vestibular impairment and the implications thereof in children with sensorineural hearing loss (SNHL) dates back well prior to the advent of cochlear implantation (CI). For many clinicians and researchers, the considerations of the impact of vestibular impairment in children with SNHL did not surface until the early 2000s when we clinically began to consider bilateral CI. One concern that arose at the time was the potential risk of inducing a bilateral vestibular loss (BVL) through injury in patients receiving bilateral CI. This concern provided the impetus for many of us to study what was already known about the vestibular system of children with SNHL and to add to that knowledge by more thoroughly examining children in our CI programs.

Indeed, there is a risk of vestibular impairment resulting from CI on both a pathophysiologic (1) and functional level (2–4) and this should not be disregarded. The focus, however, of the current review will be to highlight our current understanding of the interplay between SNHL and vestibular impairment, specifically as it pertains to motor and cognitive development and outcomes in our children with SNHL who receive CI.

Role of the Vestibular System

We often consider the primary role of the vestibular system to be the maintenance of balance, however, its role in the

This article is part of the Topical collection on *Hearing Loss in Children*

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development and daily functioning extends well beyond simply maintaining equilibrium. Specifically, the vestibular system plays several important roles in how we navigate through and interact with our environment. These include but are not limited to maintaining a stable visual view of the world through controlled eye movements; estimating and perceiving self-motion including parameters of speed, distance, and heading direction; maintaining standing balance; supporting mobility (walking, cycling, and driving); and developing and guiding spatial navigational behaviors, strategies, and memories. Notably, all of these aspects of perception and performance typically require multiple sensory inputs (e.g., auditory, visual, and proprioceptive) and occur across a variety of complex conditions including, when overall sensory information is impoverished (e.g., darkness) or challenged (movement/balance perturbations), and during multi-tasking (e.g., walking while talking and physically navigating while route planning).

Development of Postural Stability

Typical Development

To fully understand the impact of sensory deficits in humans, one has to consider the stage of cognitive incline, as in the case of children, or cognitive decline, as in the case of adults, at which the sensory deficit occurred. For many of the children with SNHL who receive CI, the vestibular deficit is either congenital or early acquired. The vestibular system is one of the first systems to be established and responsive with vestibular afferents that are mature and functional in the early stages of human development (5, 6). Morphologically, the vestibular sensory system is fully developed at birth while balance continues to mature with age. In order to understand the gap in development that such a congenital or early vestibular deficit imposes, we must first consider what we know about motor development in the presence of a full complement of senses and intact neurocognition. In typically developing children, acquisition of postural stability occurs in a cephalocaudal progression (7). This process requires an active sensorimotor system that is well organized and functions in a context-specific manner (8). For postural control, an active sensorimotor system requires (1) sensory organization that integrates one or more of visual, somatosensory, or vestibular input and (2) a motor adjustment process that allows for sensory feedback (8, 9). The latter develops earlier and is available during childhood, whereas the higher level integrative processes develop over a longer period of time (10). Infants and young children depend primarily on the developing visual system to maintain balance (9). By age 3–6 years, children begin to use somatosensory information preferentially (9, 10). It has been suggested that the skills needed to ignore misleading sensory inputs and integrate multiple sensory stimuli begin emerging at 4 to 6 years of age; however, the ability to resolve sensory conflict does not reach the adult level until approximately 7–10 years (11). There is some evidence to suggest that one of the roles of the vestibular system is to select and suppress misleading information gathered from the visual and somatosensory systems (9). That is, to select the most appropriate balance strategy for a particular context, a typical adult requires a functional vestibular system. However, the vestibular system's ability to resolve conflict develops slowly and only reaches adult-like levels between 11 and 15 years of age (9, 12).

Developmental Impact of Hearing and Vestibular Loss

In comparison to normal childhood development, the development of motor function and postural stability in the setting of SNHL is complex. It has been demonstrated that, on average, children with congenital hearing impairment and normal vestibular function develop head control and independent walking at ages above the 80th percentile of normal (11). This suggests that the impact of a hearing deficit alone has a positive influence on increased motor development in children with congenital or early acquired SNHL and normal vestibular function.

However, vestibular dysfunction is more common in children with SNHL and is specifically found in up to 70% of those who have severe to profound SNHL (13–15). If we consider patients with severe and bilateral vestibular dysfunction (BVL) across both the canal and otolith systems, then the prevalence of dysfunction among children with SNHL ranges from 35 to 40% (16–18). This makes vestibular function the single most common associated feature of SNHL. With this in mind, it is advisable to include a screen for vestibular impairment and motor delay in the evaluation of children with SNHL (14, 19–21).

In the population of children with SNHL who have abnormal and absent vestibular responses, the age at which motor skills develop is increased (22) (23–25). Despite early reliance on vision in all children for the development of balance skills, those with SNHL-VL have significantly delayed motor milestones—they stand and walk much later than their hearing peers (23, 26, 27). These children must solely rely on visual and somatosensory inputs to select an appropriate balance strategy in any given context throughout childhood development. As development continues, simple tasks of balance reveal that the relative importance of other sensory inputs, such as vision or proprioception, are of equal importance to children with normal vestibular function compared to those with SNHL-VL (28). On difficult tasks of balance, children with SNHL-VL demonstrate an increased reliance on proprioception, whereas their reliance on vision remains the same as that of children with normal vestibular function (28). While

these inputs permit the maintenance of balance for a given level of task difficulty, compensation for the missing vestibular inputs is not complete due to the reweighting of these inputs and consequently, balance performance falls apart.

The resilience of children with SNHL-VL is not to be underestimated. The majority of these children do eventually achieve their milestones even in the presence of significant vestibular dysfunction (22) and their performance on simple balance tasks can be indiscernible from that of children with normal vestibular function (28, 29•). Some studies even suggest that postural disturbances that result from isolated SNHL-VL are usually corrected by the time these children reach their teenage years (23, 24, 28). This is born out in the capacity of children with SNHL-VL to participate in most activities of daily living as long as the threshold of difficulty does not cross a critical threshold for which they can no longer compensate. As a result, clinicians and families may underappreciate or overlook the importance of vestibular deficits in this population. However, when balance tasks become challenging or sensory information is limited (i.e., both vision and proprioception are disrupted), significant differences arise in this population both in the capacity to maintain posture and the effort associated with doing so (28, 29•). Once again, this underlines the importance of using age-appropriate challenging balance tasks in the clinic, such as one leg standing or tandem stance (eyes open, eyes closed), when screening for impairment in children with SNHL (29•). Implementing a challenging balance task in the assessment of this population may reveal the degree of impact of an underlying vestibular deficit which would be of additional value in measuring subclinical impairment and how it relates to the overall quality of life.

Impact of Etiology on the Prevalence of Vestibular Impairment in Children with SNHL

Variability in the relationship between auditory and vestibular function can certainly be linked to differences in the etiology and the severity of the inner ear disorder (30) (31–33). Children with severe to profound SNHL—particularly if the underlying etiology is (1) an acquired infectious cause (i.e., meningitis and cCMV), (2) syndromic (Usher Type 1 Syndrome) or related to (3) cochleovestibular anomalies—are at very high risk of vestibular and balance dysfunction.

A number of acquired infectious causes of SNHL such as meningitis (34–38) and congenital CMV (cCMV) infection also present with associated vestibular and balance impairment (39). Children who become deaf from meningitis nearly universally exhibit profound dysfunction of their vestibular end-organs and depending on the developmental stage at which this occurs this can have variable impact on development of their postural stability (3, 17). Congenital CMV is increasingly recognized as an important cause of SNHL and motor delay and is estimated to affect 0.4% to 2.3% of live births in the USA with the seroprevalence in Ontario, Canada, being 0.6% (40). While the majority of children with cCMV (90%) do not have symptoms at the time of birth, 8 to 15% of them will present later in life with SNHL. In those that do present with symptoms at birth, 30 to 65% will develop to SNHL of patients (41). The distribution of vestibular impairment is likely similar to that of SNHL in both the symptomatic and asymptomatic setting. While much remains to be understood about the pathophysiology of the injury to the labyrinth in the setting of cCMV, active infection has been shown to produce extensive injury throughout the inner ear in addition to the central nervous system (CNS). More specifically, cytomegalic cells containing inclusion bodies, inflammation, and active infection have been identified histopathologically, within the labyrinth (42, 43). A child's ability to compensate for the peripheral deficit might be impacted by the dual insult that also affects the CNS. The sensory deficits resulting from cCMV are acquired in late pregnancy and often progressive, which may also be a clue to the underlyng pathophysiology (44). Progressive, partial, or complete vestibular dysfunction is more common than SNHL in infants with symptomatic cCMV (45–47). This peripheral dysfunction along with the associated central nervous system dysfunction leads to the significant balance disturbances seen in this population with the majority of the children with cCMV infection experiencing a later age at walking (48). Congenital CMV infection should be considered in the differential diagnosis for children with SNHL and/or balance disturbance as well as neurodevelopmental disabilities (49).

While several genetic causes of SNHL have a high prevalence of vestibular dysfunction, the most important of which is type 1 Usher syndrome (USH1). USH1 is a recessive disorder characterized by congenital SNHL-VL and progressive visual impairment due to retinitis pigmentosa (RP). As multigene panels for evaluation of childhood SNHL often include Usher syndrome genes, Usher syndrome may be diagnosed prior to onset of the RP. Alternatively, Usher syndrome may be diagnosed by its clinical manifestations, prompting genetic evaluation and testing. Any child with profound SNHL-VL of unknown etiology should be referred to a specialized ophthalmologist and geneticist with the specific question of USH1 in mind (50).

One might expect the relationship between vestibular and auditory dysfunction to be relatively straightforward in the setting of inner ear dysplasia. However, not all children with SNHL due to inner ear dysplasia demonstrate BVL (31, 51). As an example, children with Pendred Syndrome, an autosomal-recessive cause of SNHL, associated with incomplete partition type 2 (IP-2) of the cochlea and enlarged vestibular aqueduct (EVA) may present with vestibular dysfunction in up to one-third of cases (52).

There are also reports of BVL occurring in the setting of SNHL due to thalidomide fetopathy, kernicterus, nonsyndromic autosomal recessive-type SNHL, as well as SNHL of unknown etiology and this is by no means an exhaustive list (30). Beyond the identification of an important sensory deficit, identifying the etiology of the SNHL may aid in estimating the likelihood of concurrent BVL and reversely, knowing that the child has SNHL-VL may also help narrow the search or aid in the determination of etiology.

Many of the children with the above-mentioned etiology will find their way into our implant programs and receive CIs to rehabilitate their healing loss. In an effort to appropriately counsel and optimize their outcomes, it is important that we have a full understanding of their sensory deficits. This importance will be explored fully in subsequent sections.

Impact of Combined Cochleovestibular Loss on Outcomes Following Cochlear Implantation

In many of the implant candidates that we see today, they, along with their family, not only want but expect more than the ability to communicate orally and be educated in a mainstream environment. These expectations are no longer the definitions of success. These patients wish to function at the same level as their peers, in both the classroom and on the playground. These environments reveal precisely where our current technology falls short when carefully examining these patients in clinic. Clinical tests may be less sensitive to the subtle functional impairments that emerge in the multifaceted challenges of everyday life for these patients. Understanding the ongoing, everyday deficits that children with SNHL who use CI experience will fuel the development of rehabilitation strategies that focuses on generalized function for the future. It is possible, and even likely, that in a significant cohort of children with SNHL who use CI, the ongoing functional impairments—such as those in the domains of learning and cognition—may be over or inappropriately attributed to their rehabilitated SNHL. We know that the single most associated factor of SNHL is vestibular impairment. The projections of the vestibular system and vestibular dysfunction may lead to functional deficits that extend beyond balance which are completely overlooked clinically. Therefore, in order to address the residual subclinical deficits in a portion of children with SNHL who use CI, we may need to consider that an underlying unrehabilitated vestibular impairment may also be playing a role in their overall quality of life.

The role of the vestibular system for overall development continues to be studied and better understood. The vestibular system does play an underacknowledged role in developmental neurocognition by providing perceptual and visuospatial input important for memory and executive function. It follows logically that visuospatial tasks in the presence of vestibular function may be impaired. A number of studies have demonstrated that children with a variety of vestibular impairments reveal deficits in memory and executive function (53, 54), thus, an association between vestibular dysfunction and poor school performance has been documented (55). Overall, individuals with vestibular impairment show poorer performance on all visuospatial tasks specifically including spatial memory, spatial navigation, and mental rotation (53). A thoughtful review of the cognitive impact of vestibular impairment in children suggests that there is likely a critical period to develop accurate spatial representations (56). Individuals with bilateral vestibular dysfunction demonstrate deficits on visuospatial tasks which in some studies have correlated neuroanatomically with decreased hippocampal volume (57). If recognized, the impact of such deficits may be reduced through therapy, particularly in those with acquired vestibular impairment (58).

As is the case with any sensory deficit, vestibular dysfunction demands additional cognitive resources for activities such as staying upright or stabilizing vision. The requirement for additional cognitive resources to maintain balance leaves less available for other tasks and can contribute to overall fatigue. The maintenance of balance even in the presence of an intact vestibular system is not reflexive but draws on cognitive reserve, and additionally, may receive priority over other tasks such as conversing or reading for example (53). When considering the attentional demands of maintaining postural stability, both spatial and non-spatial tasks are equally affected (59).

Children with unilateral hearing loss (UHL) as a population are a great reflection of the deficits from sensory deprivation that extend beyond speech and language. These children do not require treatment to develop speech and language as they have a normal hearing ear, however, they do continue to present with deficits in other domains (60). For example, difficulties with spatial navigation and localization are known to 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 (49, 62•). A comparison of 14 children with UHL and 14 children with typical hearing found that the children with UHL demonstrated significantly poorer standardized balance scores than normal hearing peers (49). In addition, assessments of vestibular end-organ function (otoliths and horizontal canal) in children with UHL found that more than half of the children tested demonstrated functional abnormality. The dysfunction was more commonly associated with the worse hearing ear (62•). In summary, a portion of the deficits that we observe in children with UHL may be due to combined SNHL-VL deficit as opposed to hearing alone (60, 63). In fact, we may even find that associated concurrent vestibular impairment may play into candidacy for CI for UHL in the future.

As a final note, early interest in SNHL-VL was related to the potential negative impact that CI may have on the vestibular system. Beyond what we have learned in this domain, we have also demonstrated that vestibular impairment can consequently put the CI at risk. Specifically, children with SNHL-VL who use CI do not balance as well as their peers and have been shown to be at an increased risk of CI failures which is theorized to be due to increased microtrauma (61).

Therapeutic Strategies

The impetus to identify any sensory deficit is driven by the capacity to improve outcome. Unlike the rehabilitation of SNHL, fewer treatments exist to mitigate the deficits in balance that occur as a result of BVL. With this in mind, we can ask ourselves what value is there in identifying a concurrent vestibular impairment in a child with SNHL? Identifying peripheral vestibular dysfunction and its associated functional implications can add value in developing intervention and rehabilitative techniques that are aligned with everyday conditions and behaviors. More importantly, identification can also prevent the false labeling of children as having consequent learning and cognitive difficulties—such as global delay, central lesions, or multiple handicaps. SNHL-VL also carries with it a number of clinical safety concerns that should certainly be relayed to patients. For example, reports of drowning have occurred in patients where BVL was suspected (64). This is due to the potential for loss of spatial orientation when swimming underwater. Loss of spatial orientation in children with BVL can also happen in the dark when visual inputs are removed.

Different therapeutic approaches can be used for the rehabilitation of children with either loss of vestibular sensitivity or deficits of sensory organization and integration. For example, children with SNHL-VL may benefit from balance strategies in various environmental contexts in an effort to prime their visual and somatic senses, therefore facilitating compensation (65–68). Systematically reweighting these sensory inputs as a form of adaptation for the lack of vestibular input is helpful, but limited. Although rehabilitation is an important component of improving balance in these children, it does not restore the head referenced and gravitational spatial information that the vestibular system provides (69–71). The ability to provide children with SNHL-VL inputs that code the postural alterations that may precede and lead to a fall could translate into significant functional and safety benefits in this population. While several devices whose aim is to stabilize balance through auditory of vibrotactile biofeedback have been studied, they demonstrated variable benefit and have been exclusively trialed in only adult populations (70, 72–75).

Sound inherently carries environmental information, such as timing and level differences, that is important for determining our egocentric position in space. Even stationary auditory cues influence our postural alignment subconsciously (76, 77). A child's ability to maintain balance with diminished vestibular input reflects compensatory adjustments that may have occurred during development (23, 24). Fortunately, children with SNHL-VL can gain access to sound through CIs. The restored sense of audition and the increased spatial awareness that may be gained through bilateral CIs, in particular, may also help provide them with cues to support their balance (13–15). In challenging balance situations and during perturbations, children may rely on and integrate senses, such as hearing, in order to stay upright in a way that does not resemble the strategies of typically developing children (78). There has been some indication that rehabilitation of SNHL with CI positively influences balance function; small improvements in performance have been documented on standardized tests of balance in some individuals when their implants are on and active (13, 14, 79). There are a number of underlying mechanism which could account for this beneficial effect on balance that is seen when hearing is restored with CI. The first is the access to additional spatial cues that occurs in the setting of bilateral implants even when binaural hearing is not restored to normal. A second mechanism that may account for the positive effect on balance from CIs may relate to extracochlear current spread from the intracochlear electrode array. From previous studies, we know that such spread occurs where we demonstrated EMG activity in the facial musculature of nearly 50% of children with CI, with translation into clinically evident facial movement in a much smaller proportion (80). The vestibular end organs, namely the saccule, are in even closer proximity to the implant electrode than the facial nerve and are also housed within the same fluid-filled environment as the cochlea. Indeed, we can infer that current from the intracochlear electrode array can spread to surrounding structures and lead to activation of the vestibular end organs as well as the facial nerve (81–83). In addition, we have also demonstrated that vestibular end-organ responses, specifically cervical and ocular vestibular evoked myogenic potentials (cVEMP, oVEMP respectively) can occur in response to direct CI stimulation, even in cases where acoustic VEMP responses are no longer present (84). Furthermore, stimulation at these levels leads to improvements in the behavioral perception of verticality as measured by the subjective visual vertical test (85). As such, it is certainly possible that electrophysiological changes may occur at the level of the vestibular end organs and afferents in response to an activation of a CI.

In an effort to capitalize on both the possible improvements in balance seen from biofeedback as well as the potential for additional benefits from extracochlear current spread, work at our center has focused on studying the effect of a head-referenced cochlear implant (CI) stimulation system, BalanCI, on balance and postural control in children with

SNHL-VL who use bilateral CI. Children with SNHL-VL demonstrated more stable balance when using BalanCI as measured by an improvement in standardized balance scores, postural control measures, and reductions in falls (86, 87). Ongoing investigations are underway and this may prove to be an intermediary solution for individuals who already have CIs in place.

Beyond substitutive strategies and much in the same way that a CI restores a sense of audition, work is being done on the creation and application of vestibular implants for individuals with BVL. This approach is currently being trialed in adult human subjects by several research groups (88–92). Given the high rate of concurrent bilateral profound SNHL with BVL, treatment often necessitates a device that rehabilitates the SNHL as well—with most experimental devices having the option of both an intracochlear and vestibular stimulating electrode array. There has been success in using the electrical stimulation from these devices to restore vestibular end-organ function (for example, the VOR), reestablishing vestibular percepts, and stabilizing vision and balance (93–97). While results remain promising, much work is required before such devices come into mainstream clinical practice, particularly for pediatric patients. If there is ongoing success of such an approach, one of the challenges to highlight is that the application of these devices in the pediatric population has a prerequisite of early and accurate assessment of the vestibular system which can be challenging due to the behavioral requirements in a majority of vestibular tests.

Conclusions

As humans, our ability to safely and effectively navigate our environment allows us to be interconnected and productive within society. Learning to do so is as important as learning to communicate. Congenital or early acquired vestibular deficits in children with SNHL may impact multisensory integration and neurocognitive function, which inhibits their ability to participate fully in their respective environments. As a first step, we need to recognize which children with SNHL in our hearing loss and cochlear implant programs are suffering from vestibular deficits. Screening and diagnostic methods are becoming increasingly available and easy to use. Secondly, these deficits need to be accounted and controlled for in research methods examining neurocognitive deficits in children with SNHL-VL particularly those who receive CI. This is not to underestimate the complexity and redundancy of the sensory systems at play, but the mechanisms by which multisensory integration happens can create challenges in measuring the full impact of these sensory deficits. Despite these multiple challenges, identifying and assessing these children with SNHL-VL will aid our understanding of the functional implications that vestibular deficits impose on a child's development and outcomes. Incorporating these empirical intricacies into our clinical evaluations will lead to the development of better rehabilitative strategies by differentiating those with isolated cochlear loss from combined cochleovestibular deficits, ultimately improving the quality of life in both clinical populations.

Compliance with Ethical Standards

Conflict of Interest The authors declare that they have no conflict of interest.

Human and Animal Rights and Informed Consent This article does not contain any studies with human or animal subjects performed by any of the authors.

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