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Contemporary Concepts in Pediatric Vestibular Assessment and Management

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Cochlear Implants and Children with Vestibular Impairments

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Abstract

Sensorineural hearing loss (SNHL) in children occurs in 1 to 3% of live births and acquired hearing loss can additionally occur. This sensory deficit has far reaching consequences that have been shown to extend beyond speech and language development. Thankfully there are many therapeutic options that exist for these children with the aim of decreasing the morbidity of their hearing impairment. Of late, focus has shifted beyond speech and language outcomes to the overall performance of children with SNHL in real-world environments. To account for their residual deficits in such environments, clinicians must understand the extent of their sensory impairments. SNHL commonly coexists with other sensory deficits such as vestibular loss. Vestibular impairment is exceedingly common in children with SNHL with nearly half of children exhibiting vestibular end-organ dysfunction. These deficits naturally lead to impairments in balance and delay in motor milestones. However, this additional sensory deficit likely leads to further impairment in the performance of these children. This article focuses on the following:

1. Defining the coexistence of vestibular impairment in children with SNHL and cochlear implants.
2. Describing screening methods aimed at identifying vestibular dysfunction in children with SNHL.
3. Understanding the functional implications of this dual-sensory impairment.
4. Exploring possible rehabilitative strategies to minimize the impact of vestibular impairment in children with SNHL

Keywords: SNHL, cochlear implant, vestibular impairment, balance, equilibrium

Learning Outcomes: As a result of this activity, the participant will be able to (1) list the common etiologies of deafness associated with vestibular impairment in the pediatric population and (2) identify the signs of vestibular impairment in children presenting with SNHL.

Background and Rationale

Sensory deficits in children lead to developmental anomalies that begin at the level of the affected end organ and carry through to impact all pathways en route to and including the primary sensory [1](#) [2](#) [3](#) [4](#) [5](#) [6](#) [7](#) and secondary association cortices. [8](#) [9](#) [10](#) [11](#) The result of such deficits are demonstrated by how children process and perceive decremented or absent incoming sensory information. Their impaired response to this is reflected back in their behaviors and how they interact (or possibly do not interact) with the world around them. While in some instances (i.e., partial vision or hearing loss) sensory deficits can be habilitated (i.e., with glasses, hearing aids, or cochlear implants [CIs]), in other instances rehabilitative options remain more limited. While initially much focus was placed on the capacity to restore as accurately as possible the incoming sensory information, we have become aware that this leads to only a partial reduction in the morbidity and behavioral consequences. The reason for this is that there may be loss of fidelity of the original signal even in the best-habilitated conditions and some features of the sensory signal may be lost to higher order processing functions even with adequate habilitation. Restoration is occurring in the context of a varying duration of deprivation and therefore incoming signals are met by a system that has either experienced lack of development or inappropriate development driven by the lived sensory deprivation. Finally, much overlap exists in the processing of information across our senses; however, sensory deprivation and its rehabilitation are often studied in isolation. It is with this framework that we will examine the far-reaching consequences of dual-sensory deprivation in the setting of the functional deficits we see in children who present with combined cochleovestibular dysfunction.

Vestibular and Balance Impairment in Children with SNHL

The importance of studying the relationship between peripheral vestibular function in the setting of deafness is underlined by the fact that sensorineural hearing loss (SNHL) is the most common congenital sensory impairment occurring in 3 out of every 1,000 live births. [12](#) The prevalence of vestibular impairment in children and individuals with SNHL is high, ranging between 20 and 70%. [13](#) [14](#) [15](#) [16](#) [17](#) Our own studies demonstrated that while 50% of children with profound bilateral SNHL had some abnormality of vestibular end-organ function, 35% displayed severe or complete impairment. [13](#) [18](#) Dysfunction of the vestibular end organs in these individuals translates into impairments of balance. [19](#) [20](#) A great majority of children with vestibular dysfunction will never be vertiginous. This is particularly likely if their vestibular dysfunction is nonprogressive, severe, congenital, and/or bilateral, as is frequently the case in association with significant SNHL. Given this high prevalence and the absence of clinical symptoms, children presenting with SNHL should be screened for vestibular and balance dysfunction. [14](#) [21](#) [22](#) [23](#)

Impact of Etiology

We have previously shown that the risk of having concurrent cochleovestibular loss is dependent on the etiology of the SNHL. [13](#) While our capacity to identify the etiology of deafness in children continues to expand, there remain some children who despite all evaluative tools remain undiagnosed. Our current ability to characterize the etiology in a child with SNHL relies on diagnostic tools such as imaging (i.e., primarily magnetic resonance imaging ± computed to-

mography), molecular genetic techniques (i.e., next-generation sequencing panels for syndromic and nonsyndromic hearing loss), and virology testing for the detection of congenital cytomegalovirus (cCMV) infection. The capacity of these diagnostic tools is enhanced by the most accurate and detailed phenotypic description of the child. As a result while etiology can help us predict the risk of coexisting vestibular impairment in a child, the recognition of vestibular impairment can likewise contribute to the attribution of an etiology of their deafness. The next section will focus on specific etiologies of SNHL where vestibular impairment is highly prevalent and also where vestibular impairment can contribute to reaching a diagnosis. It is beyond the scope of this article to review all known etiologies of SNHL.

Syndromic Hearing Loss

Usher Syndrome

Several syndromic 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 genetic cause of congenital onset SNHL and vestibular impairment, in addition to progressive visual impairment due to retinitis pigmentosa (RP). The multisensory impairments carry with them important considerations for long-term communication. While USH1 is a syndromic cause of SNHL, it falls into the category of nonsyndromic mimicker given that there are no other visibly detectable features. Its recognition relies on the identification of the phenotype in conjunction with genetic evaluation. In the absence of screening for phenotypic features (i.e., vestibular and balance impairment), early diagnosis requires a high clinical index of suspicion. Relying on the development of visual symptoms alone often leads to delayed diagnosis, given its slow progression and necessity for specialized ophthalmologic assessment (i.e., electroretinogram). While most children with SNHL will receive the recommendation to have their vision assessed, such routine assessments will not detect the retinal changes associated with USH1. Recognition of vestibular impairment in a profoundly deaf child should therefore prompt referral to a specialized ophthalmologist and geneticist with the specific question of USH1 in mind. [24](#)

Cochleovestibular Anomalies

Children whose deafness results from cochleovestibular anomalies such as incomplete partitioning defects (types I–III), isolated enlarged vestibular aqueduct, and common cavities have been well described to have coexisting vestibular impairments. In these anomalies where both the vestibular and cochlear impairments may be progressive, changes can occur either simultaneously or independently. While the majority of children with SNHL and vestibular dysfunction will not present with vertigo, the progressive dysfunction seen in cochleovestibular anomalies may be evidenced by acute-onset severe vertigo, lasting days to weeks and is at times mistaken for vestibular neuritis or Meniere's disease. [25](#) Occasionally, transient vestibular complaints are described that can occur with or without a concomitant fluctuation in hearing loss. Most often the deficits improve in time but leave a small permanent decrement (measurable but not usually clinically apparent) which over time contributes to a cumulative substantive loss both in hearing and vestibular function. Cochleovestibular anomalies may occur in isolation or may be associated with an underlying constellation of symptoms such as CHARGE, Waardenburg, branchio-oto-renal, and Pendred's syndromes.

In children with CHARGE syndrome, there is likely near-total absence of vestibular function.²⁶ Given that the posterior labyrinth, which comprises the vestibule, the saccule, and the semicircular canals, is absent and is described as millet seed in configuration. This anatomic feature is pathognomonic for this syndrome on imaging studies and highly correlates with the known genetic defect describing this syndrome, CDH7.

Acquired Hearing Loss

Infectious

Congenital Cytomegalovirus Congenital CMV is increasingly recognized as a common cause of SNHL. The disease incidence is highly correlated with the seroprevalence rate for the virus which is highly geographically variable. Congenital CMV infection is estimated to affect 0.4 to 2.3% of live births in the United States, and up to 90% of those are asymptomatic at birth. Eight percent to 15% of asymptomatic patients will present later in life with SNHL, while symptomatic cCMV infection will lead to SNHL in 30 to 65% of patients.²⁷ Vestibular insult in the setting of cCMV can be expected in patients who are severely affected but can also occur in those children who are classified as "asymptomatic." The impact of the virus on the labyrinth has recently been confirmed histopathologically where cytomegalic cells containing inclusion bodies, inflammation, and active infection produced extensive injury throughout the inner ear in addition to the central nervous system.²⁸ This dual peripheral and central insult may worsen the functional deficit as well as impair or limit compensatory strategies which would typically drive rehabilitation. The sensory deficits resulting from cCMV are also acquired, most commonly in the third trimester, and progressive which may reflect that the underlying injury is degenerative and vascular in nature.^{29 30} One clinical study of infants with symptomatic cCMV infection at birth demonstrated that progressive, partial, or complete vestibular dysfunction was more common than SNHL.³¹ The majority of the children with cCMV infection display balance disturbances, including walking at a later age.^{32 33 34} Congenital CMV affects the general development of the brain and gives rise to a complex pattern of difficulties. Congenital CMV infection needs to be considered in children with hearing impairment and/or balance disturbance and/or neurodevelopmental disabilities.³⁵

Meningitis It is estimated that 5 to 35% of patients who survive meningitis experience partial to profound SNHL.^{36 37 38 39 40} Although not as well studied, the incidence of vestibular loss in the same population appears to be lower and in the range of 3 to 12%.^{41 42} We examined a large cohort ($N = 153$) of children who had deafness following meningitis and were habilitated with CI. In this group, we examined vestibular end-organ function according to etiology and demonstrated that all children with meningitis and 46% with cochleovestibular anomalies had horizontal canal dysfunction, while 45 and 46%, respectively, displayed saccular dysfunction.¹³

It is well known that bacterial meningitis can lead to precipitous ossification of the inner ear. Although the details of inner ear damage are not fully understood, spread of infection appears to occur primarily via the cochlear aqueduct and occasionally via the modiolus.⁴³ Histopathologic temporal bone studies following meningitis suggest that SNHL results from suppurative labyrinthitis in the acute disease phase,⁴³ while biochemical alteration of the inner ear milieu may also contribute. In some cases, new bone and fibrotic tissue is deposited in the inner ear with the horizontal semicircular canal being the first point of ossification and

commonly involved. In studies of temporal bones, the scala tympani, and more specifically the basal turn, was also frequently affected by inflammation, while the scala vestibuli was affected in 50%. Inflammation did not involve the saccule or utricle in any cases.⁴³ These findings parallel what we see radiologically and clinically in this population. In our cohort, horizontal canal function was almost universally affected, whereas saccular function was well preserved in the majority. Static and dynamic balance function was also poor in children following meningitis.⁴⁴⁴⁵ Despite this poor performance on standardized balance testing, these children display functional compensation as witnessed by their participation in a wide variety of balance intensive activities (i.e., horseback riding, dancing, and cycling). This functional compensation suggests that they are able to reweight other sensory information (e.g., vision) for the maintenance of balance. This capacity was demonstrated in our data which demonstrated an overreliance on vision in this population compared with both normal hearing controls and children with cochleovestibular loss from other causes.⁴⁴

While the effect of meningitis remains significant, it must be acknowledged that routine vaccination against *Haemophilus influenzae* and *Streptococcus pneumoniae* has thankfully reduced the burden of disease, and in catchment area with high rates of vaccination there has been a notable reduction in new cases presenting.

Ototoxicity

Drug-induced ototoxicity can occur secondary to the administration of several groups of antibiotic, chemotherapeutic, and diuretic agents. Monitoring for ototoxicity in children who are of necessity on such therapeutic regimens currently typically consists of audiometric assessment alone. Vestibular evaluation is rarely, if ever, considered within the monitoring paradigms despite the fact that many of these agents are more likely to cause vestibular than cochlear toxicity. For children with ototoxic injuries, disequilibrium is the most common impairment with vertiginous symptoms being relatively uncommon. Aminoglycosides are the most common category of antibiotic agents that lead to ototoxicity; however, severe vestibular end-organ damage can also occur with vancomycin and viomycin. While uncommon, some individuals present with increased genetic susceptibility to aminoglycoside toxicity as a result of a mitochondrial DNA defect.⁴⁶ Likewise, most children with cystic fibrosis routinely receive aminoglycosides via a variety of routes. Some series report that vestibulotoxicity in this group is roughly 12%; however, this is likely an underestimate, as this prevalence was determined by survey data and only 15% of cystic fibrosis units participating in the study routinely performed audiometric and/or vestibular ototoxicity monitoring.⁴⁷

Children receiving chemotherapy are also at risk of ototoxicity. Cisplatin in particular is well documented to have ototoxic effects with a prevalence ranging from 26 to 90% where the variability can be accounted for by differences in treatment and monitoring protocols.⁴⁸ Most centers monitor ototoxicity through audiometric outcomes including ultra-high frequency testing (>8 kHz) with few, if any, concurrently monitoring vestibular toxicity.

As a result, in our institution we are embarking on better defining the prevalence of vestibular deficits in these children being screened audiologically. Unpublished preliminary data demonstrate that more than 50% of these children have abnormal horizontal canal function and just under half have abnormal otolithic testing. Depending on the level of task difficulty, between 35 and 65% demonstrated deficits in static balance.⁴⁹

Configuration of Hearing Loss

Just as hearing loss can present in any degree of severity and at any rate of progression, so can vestibular impairment. While on average the risk of vestibular impairment is highest in those with the most significant cochlear deficits (i.e., severe to profound hearing loss), [50](#) [51](#) [52](#) [53](#) [54](#) the coexistence of these two sensory deficits with respect to both degree and time course can follow any number of patterns. The fact that vestibular function is relatively well preserved until the sensorineural hearing loss is advanced likely reflects the phylogenetic primacy of balance and the evolutionary cost of having balance anomalies in terms of survival. The sense of balance and the ability to perceive gravity and orient oneself to it is most primitive. Feeding, reproducing, and survival are all dependent on an organism's ability to perceive and relate to their three-dimensional surround.

Unilateral Sensorineural Hearing Loss

There has been increasing interest in the functional deficits children with unilateral SNHL may experience particularly as we consider our ability to reduce their morbidity using single sided implants. We have recently confirmed that balance deficits occur in children with unilateral profound SNHL. [55](#) While their balance impairments were found not surprisingly to be less severe than those displayed in the setting of bilateral deafness, they remained significant in comparison to their normal hearing peers. [55](#) Multiple factors likely contribute to the poorer balance skills exhibited in children with unilateral deafness. For example, un-rehabilitated unilateral hearing loss in early development can promote an “aural preference syndrome” where hearing is biased to one ear. This likely represents developmental changes at the brainstem and cortical levels and effects processing and binaural perception. [56](#) Without equal binaural input, spatial hearing can be compromised [57](#) and the lack of symmetrical hearing could also affect balance. They may have combined impairment of both hearing and vestibular function. In a recent study, we demonstrated that more than half of the cohort had some abnormality of the vestibular end organs (otoliths and horizontal canal), with the prevalence of end-organ-specific dysfunction ranging from 17 to 48% depending on organ tested and method used. In most children, impairment occurred only on the deaf side. [58](#) The prevalence of vestibular dysfunction in children with unilateral deafness is therefore high and similar to that of children with bilateral deafness. Similarly, up to 45% of adults with unilateral deafness have been shown to have signs of vestibular impairment. [59](#) This high prevalence of vestibular impairment again is likely related to the etiologies seen in children presenting with unilateral hearing loss. Several recent reviews demonstrate that the predominant etiologies of unilateral hearing loss in the pediatric population include cochlear nerve aplasia, cCMV, cochleovestibular anomalies, and sudden SNHL of both a traumatic or idiopathic mechanism. [60](#) [61](#) We have addressed the coexistence of hearing and vestibular deficits earlier in children with cCMV and cochleovestibular anomalies. For children with cochlear nerve aplasia, some may also experience concurrently aplasia or hypoplasia of the vestibular nerve(s). Additionally, individuals who present with sudden, single-sided SNHL also frequently present with symptoms suggestive of an acute change in vestibular function at or around the time of presentation with vertigo, occurring in 20 to 60%. [59](#)

What is becoming clear is that the role of the dual-sensory impairment is underestimated especially in children with unilateral SNHL and concomitant balance anomalies. These children are so easy to consider as “normal,” but when we consider the additive demands on cognition

required in cases where there is diminished unilateral auditory and balance information, we can start to reconsider the now increasingly documented deficits observed in this group of children. These children are likely suffering from a unilateral but multisensory loss which either independently or in combination affects attention, learning, reading comprehension, and school performance. This concept of “cognitive vestibulopathy” will be further addressed in a subsequent section.

Impact of Cochlear Implantation

Just as anomalies of the auditory system due to genetic defects, meningitis, or other causes may be associated with parallel injuries of the vestibular end organs, so too might the act of cochlear implantation iatrogenically cause injury to the vestibular system. Much of our current interest in vestibular function in children with SNHL followed from the advent of bilateral cochlear implantation. In keeping with this, many programs sought to better understand the balance system, given concerns it may lead to bilateral vestibular impairment. There are several potential mechanisms by which surgically induced injuries to the labyrinth may occur, including (1) induction of a serous labyrinthitis due to opening of the membranous labyrinth, (2) introduction of blood into the inner ear, (3) mechanical injury due to the insertion of the electrode array, and (4) high-speed drilling within the temporal bone. [62](#) [63](#)

More specifically, electrode insertion can lead to mechanical disruption of inner ear structures including rupture of the basilar membrane, fracture of the osseous spiral lamina, transection of the scala media, and fracture of the modiolus. [64](#) [65](#) [66](#) [67](#) [68](#) [69](#) [70](#) This postimplantation injury of the cochlear and specifically injury to the osseous spiral lamina correlate with a decrement in perceptual performance after implantation. [71](#) Histopathologic analysis of temporal bones and the vestibular apparatus following CI demonstrates injury in more than half of patients and includes fibrosis of the vestibular apparatus, saccular membrane distortion, osteoneogenesis, and reactive neuromas. [72](#) Involvement of the scala vestibuli as a result of damage to the osseous spiral lamina or basilar membrane in the cochlear basal turn correlated strongly with vestibular end-organ damage. [72](#) The correlation of such histologic injuries with physiologic dysfunction is not well understood. [72](#) Measurable functional damage to vestibular function following CI has, however, been independently described in several studies. [73](#) [74](#) The quality of the evidence varies with initial reports based on subjective complaints of dizziness following CI which occurred in 2 to 49% of patients and were more likely in adults and with increasing age. [75](#) [76](#) [77](#) [78](#) [79](#) [80](#) [81](#) [82](#) [83](#)

The reported and estimated risk of losing or significantly diminishing horizontal canal function based on caloric testing postimplantation ranges between 0 and 77%. [14](#) [79](#) [82](#) [84](#) [85](#) [86](#) [87](#) [88](#) [89](#) [90](#) [91](#) [92](#) [93](#) [94](#) Some of the variability in these data may be explained by the duration of the interval from implantation to post-op testing, variation in surgical technique, coexistent pathology, and variation in vestibular testing technique. Individuals may exhibit a transient loss or decrease in function followed by recovery, [94](#) while others may experience progressive vestibular loss as a result of their underlying etiology of deafness. Longitudinal measures of horizontal canal function by caloric stimulus post-CI suggest that by 4 months postimplantation caloric function is stable and few additional improvements occur. [14](#) In adults and children, postimplant vestibular function was reduced (≥ 21 degree/s) in 29% of those “at risk” of vestibular dysfunction (i.e., those with normal or reduced but not absent horizontal canal function preimplant) and were often paralleled by changes in vestibulo-ocular reflex (VOR) phase and gain in

response to rotation. An additional study examined all semicircular canals using head impulse testing and found preoperatively decreased gain in horizontal canal in 36% as well as decreased gain in one or more of the vertical canals in 50% of cases. Postoperatively, only a single patient (9%) experienced a significant decrease in function of all three semicircular canals on the implanted side accompanied by transient vertigo and oscillopsia. [95](#)

Most studies initially focused on the effect of CI on horizontal canal function, with more recent interest in the impact of CI on the otolithic organs. Indeed, the otoliths, and the saccule in particular, may be more susceptible to damage than the semicircular canals given their proximity to the insertion path of the implant's electrode array. [72](#) In a separate study, 50% of 12 children demonstrated bilaterally normal saccular function as measured by vestibular-evoked myogenic potential (VEMP) prior to surgery, [96](#) which was subsequently obliterated given the disappearance of VEMP responses after CI. In comparison, utricular function measured using off-vertical axis rotation was not found to be impaired following CI. [94](#)

While there is certainly risk of damage to the balance system after cochlear implantation, one of the challenges in the pediatric population is obtaining reliable pre- and postoperative vestibular testing, as many children are undergoing implantation in infancy or early childhood. Some groups have been very successful at obtaining careful pre- and postoperative study of vestibular function in children undergoing delayed sequential implantation. Based on these data, the risk of total bilateral vestibular loss following bilateral simultaneous implantation is estimated to be 2% and is accentuated by a reported case of total bilateral vestibular loss subsequent to, and suspected to be due to, implantation. [17](#) [45](#) [97](#) [98](#) While this 2% should not be discounted when considering bilateral implantation in children, it is exceeded by the risk of total bilateral vestibular loss resulting from the underlying etiology of the deafness itself (i.e., meningitis, cochlear vestibular anomalies, cytomegalovirus, Usher syndrome, etc.).

In summary, the take-home points regarding the risk of vestibular injury in children with SNHL undergoing CI are the following:

1. Adults and children are different. This applies to their risk of vestibular impairment at baseline and following cochlear implantation which reflects the differences in their etiology of deafness. Likewise their ability to compensate following an acute vestibular injury is different. Therefore, decisions regarding timing of implantation, which ear to implant and bilateral implantation should be thought of differently in these two populations.
2. A great majority of children walk into the operating room with vestibular impairment at baseline prior to receiving a CI.
3. Vestibular injury can be induced by CI, when vestibular function is present prior to surgery.

Screening for Vestibular Impairment in Children with SNHL

It is clear that identifying vestibular and balance impairment is important in children presenting with SNHL. However, the practicality of doing so in this challenging population is what keeps it from being routinely performed in many centers. While the appreciation of this importance has increased the number of centers seeking expertise in this area exponentially over the last several years, complete and thorough examination of the vestibular system requires

time and expertise. Given how common SNHL is, there are inadequate resources to apply to all children presenting with hearing loss. As such there is a role for a screening assessment that can be applied by any number of clinicians to better identify those at risk of vestibular impairment who should go on to more thorough testing. While quantitative testing will be thoroughly reviewed in “Quantitative Vestibular Function Testing in the Pediatric Population”, we will focus here briefly on a screening algorithm that can be applied in the busy clinical setting. Our screening assessment includes at least one of the following:

1. Review of motor milestones.
2. Assessment of balance.
3. Assessment of horizontal canal function.

First, asking caregivers specifically about neck control in infancy as well as age at sitting and walking can provide important insight into the coexistence of a vestibular impairment. Normal and abnormal timelines for each of these milestones are outlined in [Table 1](#). It is our experience that while a family may not remember the exact timing of these milestones if they were normal, that is not the case when children's milestones fall well outside the expected norms. It is our experience that just asking about these motor skills will begin an outpouring of valuable information from caregivers who have often seen many specialists to gain some understanding of their child and what may be very subtle behavior variance.

Table 1

Red Flags for Motor Milestones ¹²⁸

Motor milestone	Timeframe
Absence of head control	4 mo
Unable to sit unsupported	7–9 mo
Unable to crawl/bottom shuffle	12 mo
Not attempting to walk	8 mo

Second, evaluating age appropriate balance skills is an important aspect of the pediatric vestibular assessment. The standard or modified Romberg's test used in adults is too difficult in very young children and then quickly becomes too easy in older children, even those presenting with vestibular deficits. When children with SNHL are challenged by sufficiently difficult balance tasks that appropriately emphasize the contribution of the peripheral vestibular system, deficiencies in balance function surface. ^{99 100} Numerous age-normalized balance tests are reported in the literature and will be reviewed in depth in “Vestibular Rehabilitation for Children”. Our preferred clinical test of balance function has been the balance subset of the Bruininks Oseretsky Test of Motor Proficiency-2 (BOT2). This has become our standard for measuring balance in children older than 4 years. ¹⁰¹ The BOT2 is a nine-item test where some tasks are performed eyes open or eyes closed and requires a standardized balance beam and reference normative data. While the use of such a standardized test takes only 10 minutes, this is feasible in a busy clinical setting where a balance screen is indicated. Likewise it does re-

quire specific equipment (i.e., balance beam) and there is easy access to the standardized normative data. To this end, we have reviewed the sensitivity and specificity of each item of the BOT2 and have demonstrated that one-foot standing eyes open and eyes closed is an effective screening tool for balance dysfunction in children. ²³ [Table 2](#) outlines the duration of expected one-foot standing by age.

Table 2

Expected and Red Flag One Foot Standing Times by Age ²³ [128](#)

Age	Duration (s) 1 foot standing
30 mo	1 (briefly)
36 mo	2
4 y	5

The final portion of our screening assessment includes a measure of horizontal canal function. This can be done without the use of any specialized equipment in the form of a clinical head impulse test. In infants less than 6 months of age, we can take advantage of their developmental inability to suppress their VOR response to assess horizontal canal function. This can be done by spinning the child (and the caregiver on whose lap they sit) on a stool and examining for post-rotary nystagmus (fast phase directed away from the direction of the acceleration) which indicates an intact horizontal VOR. ¹⁰² Given the infant's inability to visually suppress their VOR, no equipment is necessary, making it a useful tool to screen vestibular function in infants presenting with SNHL after failing their newborn screen. More complex quantitative and objective assessment can also be performed relatively quickly (i.e., video head impulse test) if the necessary equipment and expertise is available.

In ideal circumstances, all three parts of the screening assessment would be performed; however, completion of any single one step is useful in the identification of children with SNHL at risk of vestibular impairment.

Functional Impact of Vestibular and Balance Impairment

There are several reasons why we should strive to identify children with SNHL at risk of vestibular impairment. First, identifying peripheral vestibular dysfunction prevents the false labeling of children as having global delay, central lesions, or multiple handicaps. Second, different therapeutic approaches can be used for the rehabilitation of children with either loss of vestibular sensitivity or deficits of sensory organization. For example, children with SNHL and reduced or absent vestibular function may benefit from balance strategies in various environmental contexts in an effort to prime their visual and somatic senses facilitating compensation. More specifically, interventions as simple as a 10-day exercise program focused on activities of static balance activities led to significant improvement in standing balance duration in children with SNHL compared with untreated hearing-impaired controls. ¹⁰³

At a minimum, bilateral vestibular loss carries with it several clinical safety concerns that should certainly be relayed to patients. These include the potential for loss of spatial orientation when swimming under water as well as in the dark, and reports of drowning have occurred in patients where loss of bilateral loss of vestibular function was suspected. [104](#) Although such limitations are obvious to clinicians, they often are not to patients and their parents. A recent review of our database demonstrated that an absence of bilateral horizontal canal function (areflexia) increased the odds of CI device failure 7.6 times, [105](#) where failure is defined as mechanical or electrical malfunction of the surgically implanted internal component. Likewise poor balance measured on objective tests of function and saccular dysfunction measured by absence of VEMPs were also significantly more common in children with CI failure. Multiple previous failure studies, including our own, have noted that children with meningitis are more likely to experience higher implant device failure. [106](#) [107](#) The most likely reason for children with postmeningitic deafness to be more at risk for device failure is the fact that this population universally loses their vestibular function, thus increasing their risk of falls and subsequent device trauma. Again, many studies report a higher incidence of CI failure in children citing increased risk of fall as the underlying reason; [108](#) however, they make the connection to poor vestibular end-organ dysfunction. In summary, balance is poor in children with CI failure due to vestibular impairment, which increases the odds of failure nearly eightfold. The likely mechanism is an increase in falls leading to repetitive device damage. Vestibular dysfunction is therefore the largest patient-related factor contributing to CI failure identified to date. [105](#)

The child's brain has access to a remarkable degree of plasticity which proves useful in the presence of sensory impairment. As a result, excellent, although incomplete, compensation appears to occur in many children with bilateral vestibular deficits; therefore, eliciting abnormalities on functional testing may be quite difficult. It is expected that children who are deaf and have concurrent vestibular impairments will have delays in the acquisition of their motor milestones. [20](#) [44](#) [45](#) [99](#) However, given the far-reaching projections of the vestibular system throughout the brain, it is feasible that vestibular impairment may lead to deficits beyond locomotion. In fact, it is possible that the learning and cognitive deficits that are seen in children with SNHL and CI are over attributed to their rehabilitated hearing loss. It may therefore be reasonable to consider that their un-rehabilitated vestibular impairment may also play a role. In parallel to the literature on the cognitive impact of hearing deprivation, there is an entire body of work looking at the impact of isolated vestibular impairment on cognition. Classically, the vestibular system is seen to be important in the maintenance of postural stability and the stabilization of gaze through a variety of broad inputs as well as specific and fundamental reflex arcs (i.e., VOR). There is mounting evidence suggesting that, through its broad reaching inputs, the vestibular system plays a, until now underappreciated, role in neurocognition including not only perceptual and visuospatial ability but also memory and executive function.

It follows logically that visuospatial tasks may be impaired in the setting of vestibular dysfunction. To this end, several studies using the virtual Morris Water Maze Task have demonstrated profound deficits in individuals with bilateral vestibular dysfunction and more mild deficits in those with unilateral deficits. [109](#) Overall individuals with vestibular impairment show poorer performance on all visuospatial tasks specifically including spatial memory, spatial navigation, and mental rotation. [109](#) Neuroanatomically, this is correlated with decreased hippocampal volume in individuals with bilateral vestibular dysfunction. [110](#) Importantly, in those with acquired vestibular impairment, the impact of deficits in visuospatial tasks can be lessened through therapy, if indeed these deficits are recognized. [111](#)

While the impact of vestibular dysfunction on spatial memory is not surprising, the impact of vestibular loss is not limited to this neurocognitive domain. Similar to hearing loss, vestibular dysfunction seems to demand additional cognitive resources for activities such as maintaining ones balance or visual stabilization. The dedication of additional cognitive resources for this purpose leaves less available for other tasks. Even in healthy individuals with intact vestibular function, the maintenance of balance is not simply reflexive but demands cognitive resource and additionally is prioritized in advance of other tasks. ¹⁰⁹ When considering the attentional demands of maintaining postural stability, both spatial and nonspatial tasks are equally affected. ¹¹²

Finally, several studies with variable and mixed methodologies have demonstrated that individuals with a variety of vestibular impairments demonstrate deficits in memory and executive function. ¹⁰⁹ These studies and their results are reminiscent of the literature looking at these neurocognitive domains in children with SNHL and CI. ^{109 113} While there are methodological limitations and a heterogeneous definition of vestibular impairment in these studies, further research in this area is certainly warranted.

The bulk of the literature examining the vestibular-cognitive relationship focuses on deficits resulting from acquired vestibular loss. In contradistinction to these acquired losses, congenital absence of vestibular function may lead to a distortion of typical brain development, as has been demonstrated in hearing or other sensory losses. As a result, one must specifically consider the consequences of such sensory deprivation in the context of the developing child. To this end, there is scant literature focused on this topic which includes a documented association between vestibular dysfunction and poor school performance. ¹¹⁴ In addition, numerous studies in the learning disability literature suggest a correlation with “vestibulocerebellar function”; however, a huge amount of variability in the measurement and definition of vestibular impairment exists across this literature. ¹⁰⁹ A thoughtful review of the cognitive impact of vestibular impairment in children suggests that such as there is with linguistic development, there is indeed likely a critical period to develop accurate spatial representations. ¹¹⁵ Much of the resulting cognitive deficits may result from the brain changes at the level of the hippocampus that result from an absence of vestibular input during that critical time period. The direct impact of this sensory deprivation could be a failure to develop a construct for the relative representation of both the body and other objects in space. However, indirectly, poor hippocampal development may contribute to broader issues with learning, memory, and executive function. ¹¹⁵

Exploring Rehabilitative Strategies

Unlike hearing, which relies nearly exclusively on the cochlea, balance function is the product of multisensory information and is primarily made up, although not limited to sensory input from the visual, vestibular, and proprioceptive symptoms. Given the importance of balance function in our everyday lives, there is sensory redundancy that allows for the resolution of sensory conflicts and in the setting of the topic being discussed, reweighting of inputs in the setting of sensory loss. It is this redundancy that is often the target of therapeutic strategies which can go a long way in the rehabilitation of individuals with vestibular impairment. A thorough review of such strategies will be described in “Vestibular Rehabilitation for Children” and therefore the current section will focus on the restoration of sensory information at the level of the vestibular end organs.

While initial concerns were that CI may negatively impact vestibular function and balance, early on there were several indications that CI may actually positively influence balance function. For example, small improvements in performance have been documented on computerized dynamic posturography in some individuals following implantation when their implants are activated. [14](#) [88](#) We also have demonstrated that children with SNHL and implants perform slightly, although significantly, better on standardized test of balance function (BOT2 balance subset) with their CI on versus off. [99](#) Further and more rigorous study demonstrated that such benefit was achieved both in the setting of directional sound as well as nondirectional white noise. [116](#)

Although a beneficial effect of implantation on vestibular and balance function may exist, at this point we can only speculate as to the underlying mechanism that could account for this. With this in mind, such benefits may relate to the fact that the poorer performance of children with profound SNHL without an identifiable injury of the vestibular end organs may be attributed to a lack or poor quality of spatial cues that occur in the setting of deafness and exist even after rehabilitation with a unilateral implant. Although we are quick to recognize the importance of visual, somatosensory, and vestibular cues in the maintenance of balance, the contribution of hearing is rarely considered and may explain the overall poor balance performance seen preimplantation in this study. A second theoretical possibility is that the improvement somehow is related to extracochlear spread of current. Spread of current to the facial nerve leads to demonstrable electromyographic activity in the facial musculature of nearly 50% of children with CI with translation into clinically evident facial movement in a much smaller proportion. [18](#) The vestibular end organs are within even closer proximity to the implant electrode than the facial nerve and are also housed within the same fluid-filled environment. As such, it is certainly possible that electrophysiologic changes may occur at the level of the vestibular end organs and afferents in response to activation of a CI.

Electrical activity derived from the CI reaches the vestibular end organs. Our own work has demonstrated that VEMPs, a measure of otolithic function, can be elicited with electrical stimulation in a proportion of children with CI, demonstrating current spread from the cochlea to the vestibular system. The presence of electric VEMPs in acoustically nonresponsive ears, along with the shorter latencies of electrically driven VEMPs, suggests that electrical current can bypass the otoliths and directly stimulate vestibular neural elements. [117](#) Beyond stimulation of vestibular end-organ function with CIs, we have also demonstrated that the perception of visual vertical which is a reflection of perceptual tilt in the roll axis due to an imbalance in otolith function can also be improved in the presence of CI stimulation. In a group of children with SNHL and CI, asymmetric spatial orientation deficit was found in nearly half. In those with deficits, their perception of visual vertical as measured by the static subjective visual vertical test performed improved with the delivery of trains of electrical pulses from the implant. These results suggest a role for CI stimulation beyond the auditory system, in particular, for improving vestibular/balance function. [118](#)

While our approach has been to aim to extend the functionality of the intra-cochlear electrode array, several groups have been aiming to more directly and specifically activate the vestibular end organs using separate electrode arrays targeting the labyrinth. Early experiments with such devices began in animals [119](#) [120](#) [121](#) and demonstrated the capacity to restore the VOR and function of the semicircular canals. Work in this domain has moved forward in application to human subjects where the animal data have been confirmed [122](#) [123](#) [124](#) [125](#) [126](#) [127](#) and where appropriate eye movements could be elicited through direct electrical activation of the

labyrinth leading to a restoration of the VOR with a close to normal gain. The hope is that ongoing advancements in all strategies will provide several potential therapeutic options to address the balance impairment due to vestibular dysfunction in both adults and children.

Summary

Vestibular impairment and its resultant balance deficits is the most frequent coexistent clinical feature of SNHL in children. This coexistence of these two sensory impairments most frequently results from a shared etiology but also can occur as a side effect of treatment in the form of CI. Recognition of vestibular deficits in children who are deaf is important, as they have immediate impact on motor development, balance skills, and safety. Their impact also extends beyond the obvious and vestibular and balance deficits have been linked to an increased risk of implant failure as well as having an impact on cognitive development and effort. As a result, early identification and ideal treatment of these vestibular and balance deficits is required to not only understand the outcomes in children who are deaf but most importantly to optimize their function over the course of their lifetime.

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