Vestibular Evaluation and Management of Children with Sensorineural Hearing Loss

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KEYWORDS

• Vestibular dysfunction • Sensorineural hearing loss • Bilateral vestibular impairment
• Cochlear implants

KEY POINTS

- Vestibular impairment is common in children with sensorineural hearing loss
- Screening for vestibular and balance impairment in children is feasible
- Developmental consequences of vestibular impairment
  - Exist
  - Extend beyond balance
  - Impact outcome
- Cochlear implants impact vestibular function
- Abnormal vestibular function increases risk of cochlear implant device failure related to trauma
- Management options exist and are expanding

BACKGROUND

Vestibular Dysfunction is Common in Children with Sensorineural Hearing Loss

Sensorineural hearing loss (SNHL) is the most common congenital sensory impairment, occurring in 3 of every 1000 live births.[1] The prevalence of vestibular dysfunction (VD) in children with SNHL is significant, with estimates ranging between 20% and 70%.[2–6] Our own studies demonstrate that 35% of children with profound bilateral SNHL have severe or absent vestibular function. However, when more subtle dysfunction is included, 50% have evidence of end-organ vestibular abnormalities.[2,7] It is

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important to understand that most children with SNHL and VD (SNHL-VD) do not present with vertigo. Dysfunction of the vestibular end-organs in these children presents with motor milestone delays and balance impairments[8,9]; this is particularly true when VD is nonprogressive, severe, congenital and/or bilateral, which is often the case in children with significant bilateral SNHL.

There are motor, neurocognitive, behavioral, and functional implications of VD on a developing child.[8] Although an early diagnosis is beneficial with regard to counseling, intervention is necessary to improve balance in these children and promote their physical, spatial, and neurocognitive development.[3,6,8]

The relationship between auditory and vestibular function is variable, although patterns linked to the cause and degree of SNHL do exist.[10–13] Specifically, the risk of vestibular dysfunction co-occurring with balance dysfunction is highest in children with severe to profound SNHL. This condition is particularly true when the underlying cause is (1) an acquired infectious disease (ie, meningitis), congenital cytomegalovirus; (2) a syndromic cause (ie, Usher type 1 syndrome); or (3) cochleovestibular anomalies (ie, enlarged vestibular aqueduct).[14–27] In addition, VD is reported to occur in thalidomide fetopathy, kernicterus, nonsyndromic autosomal recessive-type SNHL, as well as SNHL of unknown cause.[10] The vestibular "phenotype" of many causes of SNHL remains to be defined.

Identifying the cause of SNHL may aid in estimating the likelihood of concurrent VD, and conversely, knowing that the child has SNHL-VD may guide the etiologic workup. Given the high prevalence of VD in association with SNHL and the absence of what many clinicians would consider typical clinical vestibular symptoms, children presenting with SNHL should be routinely screened for vestibular and balance dysfunction.[3,28–30]

DISCUSSION

Feasibility of Screening for Vestibular and Balance Impairment in Children

Despite the high probability of concomitant SNHL and VD, and the advantage of early identification of this VD, until recently only a small portion of children with SNHL underwent evaluation or screening of vestibular function. There is now greater appreciation of the benefits of identifying VD, and that screening, as well as more comprehensive evaluation, may be accomplished in children. For these reasons, screening for VD is increasing. This article focuses on a practical screening algorithm that can be applied in the busy clinical setting.

Screening balance and vestibular assessment includes at least one of the following, and ideally all 3 components:

1. Historical review of motor milestones
2. Direct assessment of balance
3. Direct assessment of horizontal canal function (applicable to infants younger than 6 months)

Review of motor milestones

Asking caregivers about neck control as an infant and age at sitting and walking independently can provide important insight into the coexistence of a VD. Timelines for each of these milestones are outlined in **Table 1**.

Assessment of balance

Often lack of age-appropriate balance skills is not obvious during routine office evaluation of children with SNHL. However, when children with SNHL-VD are challenged by difficult balance tasks relying upon the peripheral vestibular system, deficiencies in

Table 1
Red flags for motor milestones[94]

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	18 mo

balance function will surface.[31,32] Our preferred clinical test is the balance subset of the Oseretsky Test of Motor Proficiency-2 (BOT-2). BOT-2 is our standard measure of balance in children older than 4 years.[33] The BOT-2 is a battery of 9 tasks for which reference normative data are available. The tasks are performed under different conditions (ie, eyes open or eyes closed), and some require use of a standardized balance beam. Therefore, use of the entire balance subtest of the BOT-2 in a busy clinical setting is not feasible.

To identify a useful screening assessment of balance in children, our research group evaluated the BOT-2 battery. Sensitivity, specificity, and practicality of each test were reviewed. The one-foot standing with eyes open and eyes closed was the most effective screening tool.[30] This tool does require specific equipment (ie, balance beam), takes several minutes to complete, and standardized normative data are readily available. Therefore, it is ideally suited for screening purposes. Table 2 outlines the duration of expected one-foot standing by age.

Assessment of horizontal canal function

A head impulse test, also known as a head thrust maneuver or the Halmagyi maneuver, can be done to screen for horizontal canal function and will identify side-specific abnormalities of the vestibulo-ocular reflex (VOR). Specifically, this test is performed by having the child fixate on a stationary target and the head is rapidly (high frequency, low amplitude) rotated left or right. Children with abnormal horizontal canal function leading to a deficient VOR will not be able to maintain stationary gaze on the target, and the tester will appreciate corrective saccades back to the target. Use of a novel and interesting target allows this test to be completed in young children.

In addition to the head impulse test described earlier, infants younger than 6 months can also be assessed for postrotary nystagmus without the use of any specialized equipment to eliminate visual fixation (ie, Frenzel goggles) that would be required in older children and adults, because this test capitalizes on the inability of young infants to suppress the VOR response. This evaluation can be done in the office by spinning the child (and the caregiver on whose lap they sit) on a stool and immediately afterward examining for postrotary nystagmus. The key normal examination finding is postrotary nystagmus with the fast phase directed away from the direction of acceleration, which indicates an intact horizontal VOR.[34] Given the qualitative nature of this assessment (present or absent) it is useful in detecting bilateral and complete horizontal canal dysfunction; however, it may be more limited in identifying incomplete or unilateral dysfunction. Owing to the 6-month upper age limit, we often use this test in infants who have come in for hearing loss evaluation following a failed newborn hearing screening.

Ideally, all 3 components of the screening assessment are performed in young children with SNHL. However, completion of any single step can contribute to the identification of children likely to have VD.

Table 2
Expected and red flag one-foot standing times by age[30,94]

Age	Duration (s) 1-Foot Standing
30 mo	1 (briefly)
36 mo	2
4 y	5

It is valuable to refer children with SNHL whose screening identifies them as being at risk of VD for a comprehensive diagnostic assessment. Evaluation of horizontal canal function with caloric evaluation by videonystagmography, rotary chair and video head impulse testing, and otolithic function assessment by ocular and cervical vestibular evoked myogenic potentials are examples of tests that may be useful in evaluating children. Comprehensive testing to confirm that the vestibular end-organ is indeed the reason for children failing screening of balance and vestibular function is becoming more available due to increasing interest in vestibular testing in the pediatric population.

Developmental Consequences of Vestibular Dysfunction

Sensory deficits impact how children's brains perceive and process sensory information. Deficits of sensory information begin at the affected end-organ, impacting all pathways leading to and including the primary sensory and secondary association cortices of the brain, resulting in anomalies of development.[35–45] Beyond balance, the vestibular system plays an underrecognized role in development of cognition by providing perceptual and visuospatial input important for memory and executive function. Several studies have demonstrated that children with VD reveal deficits in memory and executive function.[46,47] Therefore it is not surprising that an association between VD and poor school performance has been documented.[48] Overall, individuals with VD show poorer performance on all visuospatial tasks specifically including spatial memory, spatial navigation, and mental rotation.[46] A review of the cognitive impact of VD in children suggests that there is likely a critical period in which accurate spatial representations may be developed.[49] Individuals with bilateral VD demonstrate deficits on visuospatial tasks that have correlated neuroanatomically with decreased hippocampal volume.[50]

Children with SNHL also have reduced capacity for serial learning even when information is presented visually, as well as deficits in organizational process and retrieval strategies used in verbal learning and memory tasks of recall of spoken words.[51–53] Early deprivation and the reorganization that happens following rehabilitation may lead to the development of deficits in processes needed for rapid encoding and ordering of recalled information.[52] Our laboratory, which studies hearing and balance in children with SNHL, believes that ongoing functional impairments, such as those described earlier, may be over- or incorrectly attributed to the auditory deficits caused by SNHL, rather than VD. If VD is recognized early, its impact may be reduced through interventive therapy.[54]

Children with unilateral sensorineural hearing loss (UHL) are expected to develop speech perception and spoken language even when profound SNHL is present. It is recognized that this group of children are at academic risk, a situation often attributed to their auditory deficit alone. However, this group of children demonstrate significantly poorer standardized balance scores than normal-hearing peers.[25] More than half of the children with UHL demonstrated VD on vestibular end-organ testing (otoliths and horizontal canal) with dysfunction most commonly in the ear with SNHL.[55] These children also present with deficits in other domains.[56] For example, difficulties with spatial navigation and localization, which are typically attributed to the auditory deficit, may instead be caused or exacerbated by vestibular and balance impairment, which is common in children with UHL.[25,55] In summary, a portion of the deficits that we observe in children with UHL may be due to combined SNHL-VD deficit as opposed to the SNHL alone.[56,57]

Another important consequence of VD in children with either unilateral or bilateral SNHL is fatigue. Compensation for VD demands use of cognitive resources to accomplish basic tasks such as staying upright or stabilizing vision. Because maintaining postural stability is a priority, spatial and nonspatial tasks are equally impacted by the reduction in cognitive resources created by VD.[58] The use of cognitive resources to maintain balance contributes to fatigue and takes away from the availability of these resources to perform other tasks such as conversing or reading.[46]

Today, many children with SNHL develop listening skills and spoken language by use of amplification and/or cochlear implants (CIs). However, achieving these goals alone does not equal comprehensive rehabilitative success. VD can impact academic and social skills and limit participation in everyday sports and activities. It is often unrecognized or incorrectly attributed to other factors such as personality or cognition. Even when VD is diagnosed, its impact may be missed because long-term outcome measures typically do include the role of balance in daily life. Therefore, the first step to further improve these children's lives is diagnosis of the underlying cause, followed by effective habilitation. Screening of children with SNHL is therefore an important first step.

Cochlear Implantation and Vestibular Dysfunction

Approximately 35% of CI candidates have absent vestibular function before surgery.[2] Of those with normal vestibular function before surgery, the risk of causing total bilateral loss is 2% in children undergoing bilateral implantation.[6,20,59,60] An inability to develop independent ambulation, a concern in the early days of implantation, is not a concern based on several decades of pediatric implant experience. Whether congenital or acquired, VD in CI recipients may be mitigated by vestibular rehabilitation.[61] In addition, children who have received a CI and have VD are more at risk of experiencing failure of the surgically implanted device.[62] This finding, first reported by our research group, is theorized to be related to increased frequency of falls causing microtrauma to the implant.

Management Options Exist and are Expanding

Diagnosis of peripheral VD and understanding its functional impact is valuable in the current management of children with SNHL and is essential to the development of new and improved treatments. Another benefit of diagnosis is parental counseling regarding safety. For example, individuals with bilateral VD may be at increased risk of losing orientation and drown when swimming underwater.[63] There is also increased risk of loss of spatial orientation in the dark due to reduced visual input.

There are different therapeutic approaches for the rehabilitation of children with VD, and for children with deficits of sensory organization and integration, known sequela of VD. At present, habilitation strategies for VD in children primarily focus on adaptation through compensation. This approach may improve balance, but still requires significant central compensation that may contribute to fatigue and limit cognitive resources available for learning.[64–67] Although habilitation may improve balance, head-referenced and gravitational spatial information by the vestibular end-organs is not restored.[68–70] Providing children with this type of sensory input would be highly beneficial. In adults, research on several devices to stabilize balance with vibrotactile stimulation has demonstrated variable benefit.[69,71–74] Technology that provides useful sensory input to improve balance and prevents falls could result in significant functional and safety benefit for children with SNHL.

Sound stimulation of the auditory system provides the brain with information regarding our position in space. Auditory cues have been shown to influence postural alignment.[75,76] A child's ability to maintain balance with diminished vestibular input requires compensatory adjustments.[77,78] Fortunately, children with SNHL-VD can access sound through amplification and/or CI and thereby improve spatial awareness. Several studies have demonstrated that hearing through bilateral CIs may provide children with cues to support balance.[3,5,79] In challenging balance situations, children with SNHL may rely on and integrate senses, including hearing, to stay upright in a way that is different from the strategies of typically developing children.[80] There is also evidence of limited improvement in balance when electric hearing from CI is being used.[3,79,81] Several underlying mechanisms could account for this benefit to balance. First is improved hearing through CI providing additional spatial cues. Second is spread of current from the electrodes within the cochlear turns. Evidence for the latter is growing. Asymptomatic current spread is known to occur in a significant number of CI recipients.[7] It has also been demonstrated that vestibular end-organs including the saccule are stimulated when the CI is activated by sound.[82–84] Our laboratory has also demonstrated vestibular evoked myogenic potentials in response to CI activation and improved patient perception of verticality.[85,86]

Our laboratory is studying a "BalanCI," a CI system that provides information about head position for children with bilateral CI and VD. More stable balance, improved postural control, and reduction in falls have been demonstrated.[87,88] This system may become an important treatment option to improve balance in implanted children.

Another approach to VD is development of implants with electrode arrays designed to directly activate the vestibular system, either with or without stimulation to improve hearing. This approach would be of benefit to individuals with and without SNHL who have functional deficits from VD. This approach is currently being trialed in adults by several research groups.[89–93] Much work is required before such devices come into mainstream clinical practice, particularly for pediatric patients. A prerequisite for use in children will be the early and accurate assessment of the vestibular system.

SUMMARY

VD is common in children with SNHL. The identification of VD is important to understanding potential developmental problems, which may be ameliorated with rehabilitation. Therefore, screening for VD is an important part of the evaluation of children with SNHL. As therapeutic options expand, early and accurate diagnosis of VD will be needed as a foundation for early intervention.

CLINICS CARE POINTS

- Children with SNHL-VD present infrequently with vertigo.
- Bilateral VD in children does not lead to nonambulation.
- Screening for VD in children who present with SNHL is important and necessary.

- One-foot standing (eyes open eyes closed) is an effective screening tool for VD.
- Objective tests of vestibular end-organ function are possible in infants and children.
- Early intervention for VD may reduce functional, neurocognitive, and motor deficits.
- Clinical safety concerns should be relayed to the families of children who present with bilateral VD.

DISCLOSURE

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