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Vestibular and Balance Dysfunction in the Pediatric Population: a Primer for the Audiologist

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The differential diagnosis of the “dizzy child” is broad and entire textbooks have been written on this topic. As otolaryngologists and audiologists, we are most interested in dizziness that originates from sources within the inner ear and may also have associated hearing loss. However, beyond peripheral inner ear causes, dizziness in children can occur due to any number of cardiac, neurologic and psychiatric conditions. While we will briefly address this broad differential, the focus of the current article will be on those diagnoses most relevant to the audiologist – notably limited to those children that end up in your audiology booth.

Children Who Feel Dizzy

In children whose presenting complaint is “dizziness” the first order of business is to distinguish true vertigo, defined as the sensation of movement (spinning, to and fro either vertically or horizontally) from other sensations such as lightheadness or feeling faint. Once it is established that these children are indeed vertiginous then careful history and physical examination can help determine if there is an underlying peripheral vestibular cause. Specifically, the duration of the vertiginous episodes can be instrumental in determining an underlying cause. Vertigo that lasts seconds to minutes can be attributed to benign paroxysmal vertigo of childhood (BPVC) or benign paroxysmal positional vertigo (BPPV). While similar in name (and acronym), these two entities are very different but often confused. The former (BPVC) is specific to the pediatric population and is felt to be a migraine variant while the latter (BPPV) is very uncommon in children. The exception is for children whose dizziness is acquired after suffering a head injury or concussion or subsequent to surgical intervention such as cochlear implantation. Vertigo that lasts 20 minutes to many hours may represent vestibular migraine, recurrent vestibulopathy or Meniere’s disease which again is uncommon in the pediatric population. Vertigo lasting hours to days may represent vestibular neuronitis or labyrinthitis or progressive vestibular impairment due to an underlying cochleovestibular anomaly which will be described in detail below. There are numerous other causes of vertigo in children that may relate to central nervous system issues (e.g., seizure, multiple sclerosis), mental health symptoms (anxiety, depression) or ophthalmologic abnormalities.

Work-up of the vertiginous child typically includes a full audiologic evaluation including middle ear reflexes and may also include ancillary vestibular end organ testing including caloric, rotary chair, video-head impulse testing and vestibular evoked myogenic potential. Meniere’s disease, labyrinthitis or cochleovestibular anomalies will also typically be associated with a hearing loss while in the remaining diagnoses should present with normal hearing.

Children Who Are “Off Balance”

Not all children who have impairment of their peripheral vestibular system will complain of

vertigo. In fact, a great majority of children with dysfunction of the vestibular portion of the inner ear will never be vertiginous. This is particularly likely if this dysfunction is severe, congenital and/or bilateral. It is this population that we will discuss in detail as these children find themselves frequently in the company of an audiologist. Before we go through the differential of children who present with balance disorders however we will first present a summary of how we can all identify those children at risk of vestibular and balance impairment in the clinic and the booth.

How Do You Identify a Child with Vestibular Impairment?

The gold standard for the detection of vestibular impairments includes horizontal canal dysfunction detected by caloric testing, rotary chair and video head impulse testing. Likewise saccular dysfunction can be measured by vestibular evoked myogenic potentials. Impairment of the vestibular end organs translates into impairment of balance and these children will often have delayed motor milestones (e.g., poor head control [beyond 6 weeks], delayed independent sitting [beyond 9 months] and delayed walking [beyond 18 months]) as commonly reported by caregivers. These delays translate into poorer balance skills that carry throughout childhood. Caregivers may report that their child is “clumsy” prone to fall or injury and also delayed in some common childhood activities such as riding a bicycle without training wheels. An audiologist or otolaryngologist who asks the question about motor milestones in a child presenting with SNHL will open the floodgates when indeed there is concern. In addition, simple maneuvers such as asking the child to stand on foot can also screen for balance impairment. As a reference, a two and a half year old should be able to transiently stand on one foot, a three year old should be able to maintain this posture for 2 seconds, a four year old for 5 seconds, and a five year old for 10 seconds. *Any child with unexplained delay in motor milestones or an inability to stand on one foot should indeed be investigated more thoroughly for vestibular impairment particularly if there is associated hearing loss.*

Balance Disorders and Otitis Media

Otitis media is exceedingly common in the paediatric population. The impact of persistent otitis media with effusion (OME) on the acquisition of language is well established and well accepted as a risk factor for poor development. However, the impact that persistent OME has on motor skills acquisition, particularly if present during critical periods of development has been studied but is less appreciated by the clinicians looking after these children. Middle ear effusions (MEE) are arguably the most common causes and risk factors for the development of balance disturbances in the paediatric population.[1–5] Multiple sources have documented that vestibular end-organ, balance and general motor function are acutely worse in the presence of a middle ear effusion6–11 and that this deterioration reverses upon either spontaneous resolution or treatment of the MEE.[6,7,9–11] A number of different theories have been proposed to explain this relationship.

While the acute effects of MEE on balance have been well established, the consequences of recurrent MEE on balance function following resolution are less clear. There are several studies that suggest the impact of MEE on balance may persist[4,12,13] while others demonstrate return to baseline following resolution of the MEE.[3] It is certainly possible that recurrent and/or persistent MEE that occur early and/or during the time of critical transitions (learning to sit, walk etc.) could translate into overall delays that persist beyond the resolution of the MEE. Children with underlying motor or vestibular dysfunction at baseline may be at particular risk of such effects. Vestibular end-organ function, balance and motor skills should be evaluated and considered in children presenting with MEE and may in some cases, guide treatment.

Balance Disorders and Sensorineural Hearing Loss

Up to 70% of children presenting with sensorineural hearing loss (SNHL) have impairment of their vestibular system with 20–40% having severe bilateral vestibular loss.[14–17] Caregivers may not appreciate the relationship between hearing and balance and may not spontaneously offer details about their concerns regarding their child's balance unless prompted by an astute clinician and it is therefore important to inquire about motor milestones in children presenting with hearing loss as outlined above.

Which Etiologies of Hearing Loss are Most Likely Associated with Balance Impairment?

Despite modern diagnostics, we still remain unable to identify which of the numerous possible etiologies underlies the hearing loss in many affected children. That being said, many identifiable etiologies of hearing loss have well described associated vestibular impairment. In keeping with this, identification of vestibular impairment can aid in narrowing the search for an underlying etiology of the hearing loss. A number of high-risk etiologies of combined cochleovestibular loss are outlined below.

Cochleovestibular Anomalies

Children with underlying abnormalities of the anatomy of the inner ear frequently present with SNHL in addition to vestibular impairment. The majority of these anatomic abnormalities fit into the description of incomplete partition type 1 to type 3 (formerly known as Mondini malformation) and can be recognized through either computed tomography (CT) or magnetic resonance imaging (MRI). In these children, the vestibular impairment, like the hearing loss, may be progressive in nature. When progressive vestibular impairment occurs these children may present with true vertigo which can be quite severe and last multiple days to weeks. It may occur with associated progression of hearing loss or independent of any changes in hearing.

Cochleovestibular anomalies may occur in isolation or may be associated with an underlying syndrome such as Waardenburg, CHARGE and Pendred syndrome.

Syndromic Genetic

The most important diagnosis that fits under this heading is Usher syndrome (US). US is a genetic recessive cause of hearing impairment, meaning that two unaffected parents each pass on a gene change to their offspring which then leads to a manifestation of the syndrome. There are 3 types of US, but for the purpose of this paper we will only discuss type 1. Children with type 1 US present with congenital onset SNHL and vestibular impairment in addition to progressive blindness due to retinitis pigmentosa (RP). The multi-sensory impairments that affect these children obviously carry with them important considerations for communication strategies in the long-term. A diagnosis of type 1 US can be elusive given that it is uncommon and these children show no other outwardly signs. The visual loss is slow and progressive and may be mistaken for more common visual impairments such as near or far sightedness. In many ways, identification of the associated vestibular impairment in these children can lead more expeditiously to a diagnosis. Therefore any child with profound SNHL and vestibular impairments, which can be screened for as outlined above, should undergo more formal vestibular testing and referral to a specialized ophthalmologist in addition to a genetic counselor with the specific question of US in mind.

Congenital and Acquired Infectious Causes

Meningitis

Children who present with SNHL following meningitis are very likely to display an associated impairment of vestibular function.[18] Radiologically this manifests as enhancement of the labyrinth initially on MRI followed by progressive ossification best visualized on CT imaging. Families of children with SNHL following meningitis will report that initially these children suffered huge decrements in their balance function followed by slow gains. Despite these gains, these children will be left most frequently with complete bilateral areflexia of the vestibular end organs and poor balance skills in difficult tasks (e.g., standing on foot with eyes closed, where visual input is diminished). Areflexia implies complete dysfunction or absence of responses of the vestibular end organs.

Cytomegalovirus (CMV)

Congenital cytomegalovirus (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 to 15% of asymptomatic patients will present later in life with SNHL while symptomatic CMV infection will lead to SNHL in 30 to 65% of patients. Vestibular impairment can be expected in patients who are severely affected. One study of infants with symptomatic CMV infection at birth, demonstrated that progressive and partial or complete vestibular dysfunction was more common than hearing loss.[18,19] Given that both the central nervous system and the inner ear can be affected by congenital CMV, the resulting vestibular and balance impairments may be more resistant to rehabilitation.

Unilateral Sensorineural Hearing Loss

The consequences of unilateral hearing loss have been recently underlined[20] one less well understood consequence includes associated vestibular and balance impairment. It is known that approximately 50% of children presenting with congenital “dead ear” have absence of the cochleovestibular nerve which is significantly higher than children with bilateral profound SNHL.[21] These children may also present with unilateral vestibular impairment and some delay in motor development although this will not feature as prominently as children with bilateral impairment.

Auditory Neuropathy Spectrum Disorder (ANSD)

Our understanding of the presentation, treatment and hearing outcomes of ANSD is evolving. What is clear is that many children presenting with ANSD also display impairment of balance function. These children often have multiple risk factors for the development of both hearing loss and vestibular impairment (e.g., prematurity, hypoxia, hyperbilirubinemia etc.) and therefore it becomes difficult to tease out their individual contribution.

Audiologic Screening for Ototoxicity, What about Vestibulotoxicity?

An additional population of children at high risk of vestibular impairment includes those children exposed to ototoxic agents. Those children at risk include those exposed to aminoglycosides (e.g., gentamicin, inhaled tobramycin in children with cystic fibrosis) and chemotherapeutics (e.g., cisplatin) amongst others. Many of these children will be in your audiology screening programs given the hearing risk attributed to these agents. What is less appreciated is the fact that many of these agents are even more injurious to the vestibular end-organs than to the cochlea and that vestibular impairment may therefore occur at a different threshold than hearing loss. Additionally,

children with posterior fossae tumours are at particular risk as they may have received chemotherapeutic agents and the cochleovestibular nerve may be injured or compromised at the time of surgical resection or biopsy.

Conclusions

The presentation of vestibular and balance disorders in children can be subtle; however, the identification of these children has significant implications for their safety and outcome. While the evaluating vestibular and balance function in a child can be intimidating, arming yourself with the few simple tools outlined in this article along with a high index of suspicion will allow you to easily and accurately identify children at risk of vestibular impairment. Audiologists are particularly well suited for this task as they are frequently on the front line evaluating children with high-risk for vestibular and balance impairment and have often established good rapport with families over a number of visits.

It is an exciting time to be interested in vestibular and balance impairment in children. The evaluation and treatment of children with vestibular impairment is currently where the identification, management and treatment of children with hearing loss were decades ago. There is no doubt that audiologists will figure at the forefront of this evolution.

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