Equilibrium-Vestibular Assessment *— for —* Infants

BY RICHARD GANS

An understanding of the vestibular system's role in postural and motor coordination performance can serve as an invaluable contribution for early identification and intervention.

Audiologists have a unique opportunity in the early identification and evaluation of equilibrium function in infants and young children. Especially for those with known congenital sensorineural (SNHL) hearing loss. The global acceptance and success of early neonatal hearing testing has also significantly improved our ability to identify infants at risk for equilibrium dysfunction, due to the comorbidities of hearing loss and vestibular dysfunction. It is estimated that over 500 syndromes and nonsyndromes are known to have an audiovestibular expressivity. Recent investigators have reported as high as 90 percent abnormal vestibular-evoked myogenic potential (VEMP) responses in children with congenital SNHL hearing loss. The emerging use of neonate and infant VEMP data suggests a much higher incidence of vestibular dysfunction than the 30–50 percent previously estimated (Kelsch et al, 2006; Picciotti et al, 2007; Sheykholesami et al, 2005; Zhou et al 2009). Several recent studies are presented in TABLE 1. We recognize the importance of normal hearing for acquisition of speech and language. Intact vestibular function is just as critical to the infant's physical and motor development as is normal hearing for speech and language acquisition.

A child has an audiovestibular system, not just an auditory system. Congenital or acquired deficits can affect all or part of this system. The peripheral end organ of the vestibular system is actually the first sensory system to develop; it precedes cochlear development (the phylogenic development of the cochlea follows that of the saccule) and is developed by 49 days' gestation. The neural connections with the central pathways continue to develop through the eighth month of gestation (Wiener-Vacher, 2008).

The majority of equilibrium problems in infants and children manifest as balance problems not as vertigo or dizziness. Delayed maturational motor milestones typically evidence the equilibrium dysfunction. It is important to ask the parents about the child's motor development timeline as well as to make your own observations. Possible indicators of peripheral-central vestibular dysfunction may include the infant's ability to hold his or her head upright, crawl, stand, and then walk. Benign paroxysmal vertigo of infancy (not to be confused with BPPV), a classification of migraine, is the condition

TABLE 1. Review of Pediatric VEMP Studies

Investigators	Study
Zhou et al, 2009	21/23 (91 percent) SNHL had abnormal amplitudes
Picciotti et al, 2007	3–15 years old
Kelsch et al, 2006	3–11 years old
Sheykholesami et al, 2005	Neonates

Dr. Patricia Castellanos Munoz conducts a balance screening.

Ronald McDonald helps Dr. Munoz make the screening fun.

Guatemalan children with hearing loss are screened for vestibular problems.

most likely to produce symptoms of vertigo in children (Gans, 2002).

Although numerous investigators and authors have reported vestibular evaluation techniques and norms for children, these studies have primarily focused on the application and adaptation of adult tests, that is, VNG, computerized dynamic posturography, and rotary chair (O'Reilly et al, 2011; Valente, 2007; Weiss and Phillips, 2006). Typically children in these studies have been five years of age or older, with the exception of the new normative VEMP data with age ranges from three months through the teen years. The purpose of this article is to provide the reader with an overview of the most common causes of pediatric vestibular dysfunction, a review of common causes, and a discussion of clinical and behavioral assessment tools that have both good sensitivity to underlying vestibular deficits and can be easily performed without the obstacles of technology or cost.

Causes of Pediatric Equilibrium—Vestibular Dysfunction

Congenital and Acquired

FIGURE 1 outlines common causes of vestibular dysfunction in the pediatric population, both congenital and acquired. It is important to remember that the majority of causes result in overall equilibrium dysfunction secondary to bilateral loss or dysfunction rather than acquired unilateral dysfunction resulting in vertigo or dizziness as is the case with adult-onset vestibular disorders.

Congenital disorders by far are the leading cause of pediatric vestibular dysfunction (Pikus, 2002). Syndromes with known and unspecified expressivity can be found in

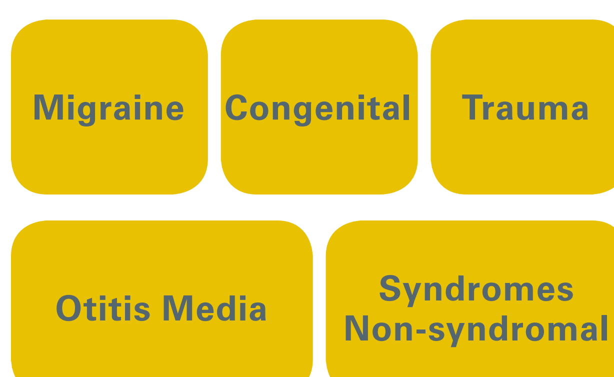

FIGURE 1. Common causes of pediatric vestibular dysfunction.

TABLE 2. A brief description of each syndrome is presented in TABLE 3 to better familiarize the reader with common expressivities associated with the conditions. It has been well established that audiovestibular anomalies are the most frequently found defect across all known mitochondrial diseases. In addition, there are nearly 70 identified different nonsyndromic loci for hereditary audiovestibular impairment. Of these, at least 30 are dominantly inherited, which means that hearing loss may not be consistently seen. So, if there were no failure of a high-risk hearing screening at birth but only the vestibular symptoms, the vestibular loss would probably be missed. Autosomal recessive disorders in the nonsyndromic category account for over two dozen loci.

Acquired conditions may include benign paroxysmal vertigo of childhood, which is the leading cause of pediatric dizziness. It could be argued that since this is a migraine variant that it is actually genetic. As its onset usually occurs between ages one and four, it affects the infant by the age he or she is independently ambulating, and there are no prior developmental delays to forewarn of future occurrences. Head trauma can cause the same form of BPPV as seen in adults. This is usually seen, however, in older children who are involved in sports or activities where they are susceptible to even minor head bumps playing soccer or contact sports. Of great concern, particularly in emerging economies, is overdosing with aminoglycosides for treatment of bacterial infections in infants and young children (Koyuncu et al, 1999). This has become a growing problem and is presently being addressed by a joint effort between the World Health Organization (WHO) and the AAO-HNS Foundation.

Dr. Paty Castellanos Munoz, a well-known audiologist in Guatemala, has added a vestibular screening

TABLE 2. Syndromes with Vestibular Expressivity

Known	Unspecified
Usher	Waardenburg
Branchiootorenal	Von Hippel-Lindau
Pendred	
Neurofibromatosis Type 2 (NF2)	
CHARGE	
Marshall	
Spinocerebellar ataxia	

TABLE 3. Description of Syndromes Affecting Audiovestibular System

Usher	Type I—Congenital-bilateral profound SNHL, retinitis pigmentosa. Type II—Mild-severe progressive high-frequency SNHL.
Branchiootorenal	Preauricular pits or tags, branchial cysts, hearing loss, and / or abnormal development of the kidneys.
Pendred	Congenital, severe-profound SNHL, abnormality of bony labyrinth. Abnormal thyroid development with goiter in early puberty or adulthood.
Neurofibromatosis Type 2 (NF2)	Bilateral vestibular schwanomas, tinnitus, hearing loss and balance dysfunction. Schwanomas of other peripheral nerves, meningiomas, and juvenile cataract.
Waardenburg	Congenital SNHL, pigmentary disturbances of iris, hair, skin. Vestibular disturbances without hearing loss.
Von Hippel-Lindau	Hemangioblastomas of brain, spinal cord, and retina. Renal cysts and renal cell carcinoma (40 percent). Dizziness/imbalance and hearing loss may be initial symptoms, may mimic Ménière's.
CHARGE	Coloboma-heart-atresia-retarded-genital-ear. Vestibular symptoms prevalent.
Marshall	Saddle nose, myopia, early-onset cataracts and short stature. Vestibular symptoms prevalent.
Spinocerebellar ataxia	Complex and progressive. Twenty-three distinct genetic disorders. May also include hearing loss.

component, in collaboration with myself, to her ongoing humanitarian audiological care in the testing and fitting of hearing aids to children. The project is conducted at McDonald's restaurants throughout Guatemala (pictured in the photos on page 24). Balance function testing has now been collected on over 1,000 children with sensorineural hearing loss. This represents the largest study of this type to be conducted and the data will soon be available for publication.

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Behavioral Assessment Techniques

Just as with auditory testing, there is an array of clinical–behavioral tests available. These are age specific with normative data in the form of maturational motor milestones as shown in TABLE 4 (Viholainen et al, 2006). A good case history is essential in speaking with the parents and asking about when these milestones were achieved. Likewise spending some informal time observing the infant, on a mat, playing can provide a great deal of valuable information. Just as we do with hearing testing, much of the evaluation is child directed. Evaluations can be accurately conducted as early as three months of age with neonates who are suspected of congenital hearing loss. Three months of age is preferable because time is needed for the neck musculature to mature enough that the child can begin to hold his or her own head upright. Ideally, as there are three distinct vestibular reflexes, one or more or preferably all three may be evaluated even with behavioral techniques: vestibulocular (VOR), vestibulospinal (VSR), and vestibulocolic (VCR).

TABLE 4. Summary of Maturational Motor Milestones

3 months	7 months	9 months	12 months	24 months
▪ Raises head and chest when lying on stomach ▪ Starts to use eyes and hands in coordination ▪ Begins to support head ▪ Pushes down with legs when feet placed on floor ▪ Moves eyes in all directions	▪ Sits with and then without support of hands ▪ Supports weight on legs ▪ Ability to track moving objects improves ▪ Rolls over ▪ Supports head when sitting	▪ Crawling on hands and knees ▪ Walking with assistance ▪ Upper body—turns from sitting to crawling position	▪ Sits without assistance ▪ Crawls forward on belly by pulling with arms and pushing with legs ▪ Creeps on hands and knees and supports trunk ▪ Pulls self up to standing position ▪ Walks holding on to furniture ▪ Stands momentarily without support	▪ Walks alone by 18 months ▪ Begins to run ▪ Can push a wheeled toy

The best test of the VCR is arguably the VEMP, which is an electrophysiological assessment tool.

Physiology of Equilibrium—Vestibular Reflexes

> Vestibular signals interact in a complex manner with other systems to produce a number of postural reflexes. The cerebellum appears to play a key role in these interactions which can involve limb and neck proprioception, touch, vision and descending cortical influences relayed to the vestibular complex primarily via the reticular formation. The various sensory modalities interact to provide information to the posture control system from three frames of reference. These are: (1) proprioception, the sense of position and movement of one part of the body relative to another, via muscle, joint, tactile and visual receptors; (2) exteroception, the relationship of objects in the environment to each other, via primarily visual and tactile inputs; (3) exproprioception, or information about the body parts relative to the external environment, from all types of sensory receptors. Because the vestibular system subserves a purely exproprioceptive sense that reports velocity and acceleration of the head relative to gravity and inertia, it is especially useful in correcting erroneous information from the other sensory inputs. [Clark, 1985]

Descending pathways responsible for postural reflexes include the vestibulospinal and reticulospinal tracts. Both receive signals from the vestibular end organ, and both are strongly influenced by cerebellar efferents. Descending motor control of the neck musculature is more closely linked to the vestibular end organ and to the semicircular canals. Limb muscle reflexes are more closely linked to input from several sensory systems.

Lifting the child in space or changing the child's position while on a variety of movable surfaces can test righting reflexes and equilibrium responses. An example of this can be seen in FIGURE 2. The three-month-old is placed on a physioball, a dynamic surface. The infant's head and torso remain stable and centered even when he is pertubated in any direction. Lateral tilt, for example, activates utricular receptors, which in turn excite vestibulospinal neurons and influence the activity of limb muscles. Age guidelines for head righting, equilibrium

FIGURE 2. Placing infant on physioball creates a vestibular response.

FIGURE 3. Righting response places the infant toward the ground.

FIGURE 4. Infant produces an upward head turn as part of the intact vestibular righting reflex.

responses, and other postural reactions, which are dependent, at least in part, on vestibular processing, have been documented by many developmental researchers, and are well known to therapists who work in pediatrics.

Another clinical observation relates to the presence of tonic neck postures. Problems in integration of tonic neck reflexes may implicate related vestibular dysfunction because labyrinthine receptors indicate body position only in conjunction with neck receptors. FIGURE 3 and FIGURE 4 show a righting response. This four-month-old is comfortably and safely placed on his mother's lap. The infant is then gently pointed downward. As can be seen in FIGURE 4, there is a clear upturn of the head away from the floor. Muscle tone is another important aspect within the evaluation, as it is closely associated with the integrity of the vestibular system. Loss of vestibular input may result in prolonged muscular debility that may even extend to the visceral muscles.

The visual observation of optokinetic nystagmus (OKN) utilizing a rotating drum, which fills the infant's visual field (at least 80 percent) is also an excellent method of assessing the VOR. It has been demonstrated that its appearance is as early as one month, and it is nicely developed at three months of age. Conditions where there is a bilateral vestibular dysfunction (BVD) will not produce a binocular bidirectional response. In those cases where there may be a noncompensated unilateral vestibular dysfunction, it will be asymmetrical with no or reduced response with the moving stimuli in the direction of the involved labyrinth. It is my experience that it is rare to see infants or young children with noncompensated BVD secondary to an acquired otologic lesion.

Triage

Infants with vestibular, equilibrium, and delayed maturational motor control disorders can now be identified at an earlier age, thanks to the success of newborn hearing screening. Unlike older children or adults with acquired unilateral vestibular deficits, these infants with BVD will not benefit from traditional vestibular rehabilitation strategies. They will benefit, however, from ongoing sensory integration, substitution, and conditioning therapy with trained pediatric physical and occupational therapists. The knowledge of the status of vestibular modality will provide the therapists with valuable information about therapy protocols and ultimately the child's prognosis over time. Although it does require at least two intact sensory modalities to produce normal equilibrium function, this early therapy jump-start will be critical in providing them with a more normal and active lifestyle during their formative years.

Conclusion

It is well documented that the audiovestibular system in infants is just as susceptible to vestibular as to hearing deficits. Audiologists can play an important role in the early identification of infants, especially those with hearing loss, who may be at risk for balance problems as well. Young infants, with the exception of VEMP testing, are not candidates for VNG, posturography, or rotary chair examinations, even if the technology were available. Therefore, an understanding of the vestibular system's role in postural and motor coordination performance can serve as an invaluable contribution for early identification and intervention.

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ALSO OF INTEREST
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