Vestibular Assessment in Infant Cochlear Implant Candidates

Age-appropriate testing can ensure the best possible outcomes and quality of life for even the youngest CI candidates who may have abnormal balance and equilibrium function.

By Richard E. Gans, PhD

Investigations regarding the viability of cochlear implants (CI) began in the 1960s and 1970s. But it was in the early 1980s that CIs became widely embraced, and have since grown in application, acceptance, and utilization, with FDA approval in 1984 for adults and 1990 for children. According to the NIDCD[1], most recent statistics, as of 2019, there have been 736,900 individuals implanted worldwide. In the United States there have been 118,100 and 65,000 implantations for adults and children, respectively. Recently the age for pediatric CI has been reduced from 1 year to 9 months of age.[2]

According to the American Academy of Pediatrics, an estimated three in 1,000 infants are born in the United States each year with moderate, severe, or profound hearing loss.[3] Hearing loss is the most common congenital condition in the U.S.[4] There are over 500 syndromes, non-syndromes, and mitochondrial heritable conditions with audio-vestibular expressivity, with many having greater vestibular expressivity.[5] The most common conditions causing bilateral audio-vestibular loss or hypofunction are listed in **Table 1**.

Video head impulse testing (vHIT) has demonstrated excellent sensitivity for vestibular function in infants and young children and can be easily administered using a fun, game-like approach for infants. This shows how vHIT was adapted for children 24-36 months of age.

Vestibular Function Assessment of Infants

The concern over vestibular and balance function is not due to the CI itself, but those congenital conditions that have vestibular as well as auditory expressivity. Numerous investigators have published data indicating that the CI is not a causation of vestibular loss. In fact, it has been the anecdotal observation that some CI patients' balance improved post-CI that has led to the investigations and encouraging preliminary reports of the success of the combined cochlear-vestibular implants in adults.

With implantations now being performed at younger ages, as early as 9 months, the infant may not yet be at an age where their delayed motor milestones and any equilibrium dysfunction has yet evidenced itself to parents or caregivers, as they face the challenges of an infant with hearing loss and the hopes of a forthcoming cochlear implant. Just as for decades prior to neonate hearing screening the parents of even profoundly deaf children were told by well-intentioned practitioners "not to worry, he/she will talk when they're ready," the same is true for infants and young children who may be quite delayed in their motor development due to lack of vestibular function. **Table 2** provides an overview of normative maturational motor development. This will provide practitioners, family, and caregivers a guideline by which to monitor the infant's development.

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Syndrome	Description
Brachiootorenal	Preauricular pits or tags, branchial cysts, hearing loss, and/or abnormal development of the kidneys
CHARGE	Coloboma–heart–atresia–retardation–genital–ear; vestibular symptoms prevalent
Marshall	Saddle nose, myopia, early-onset cataracts, and short stature; vestibular symptoms prevalent
Pendred	Congenital, severe–profound SNHL, abnormality of bony labyrinth; abnormal thyroid development with goiter in early puberty of adulthood
Spinocerebellar ataxia	Complex and progressive; 23 distinct genetic disorders; may also include hearing loss
Usher	Type I: congenital-bilateral profound SNHL, retinitis pigmentosa Type II: mild-severe progressive high-frequency SNHL
Von Hippel-Lindau	Hemangioblastomas of brain, spinal cord, and retina; renal cysts and renal cell carcinoma (40%); dizziness/imbalance and hearing loss may be initial symptoms, may mimic Ménière's disease
Wardenburg	Congenital SNHL, pigmentary disturbances of iris, hair, skin; vestibular disturbances without hearing loss

Table 1. Congenital genetic conditions with auditory-vestibular expressivity

How to Test Infants

In addition to observation and recording of the infant's motor milestones, infants can be tested with standardized electrophysiological tests as young as 3 months of age with cervical VEMP (cVEMP) testing. There have been numerous publications with normative data for infants through adolescents. This author has found through testing hundreds of infants over 20 years, that it is easy, quick, and comfortable for the infant. As the cVEMP (cervical vestibular evoked myogenic potential) requires the infant to be awake and active, it is much easier than the protocols and requirements for brainstem auditory evoked response (BAER) testing or hearing threshold determination. Video head impulse testing (vHIT) has also demonstrated excellent sensitivity for vestibular function in infants and young children and can be easily administered using a fun, game-like approach for infants as young as 9-10 months of age.

The key to successful vestibular assessment of infants and young children is to be sure that age-appropriate assessment is being undertaken. We have often heard clinicians state that they could not test a child because they were too young for a VNG. Exactly. We should not delay evalu-

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

Table 2. Overview of maturational milestones 3-24 months of age

ations because a clinic or practitioner does not have either the necessary competency or instrumentation to conduct age-appropriate evaluations. **Table 3** provides a summary of age-appropriate protocols for ages 3 months to 60 months old.

Protocols	3	6	9	12	18	24	36	48	60
Behavioral	✓	✓	✓	✓	✓	✓	✓	✓	✓
VEMP	✓	✓	✓	✓	✓	✓	✓	✓	✓
vHIT			✓	✓	✓	✓	✓	✓	✓
Rotary Chair					✓	✓	✓	✓	✓
VNG								✓	✓

Table 3. Age-appropriate protocols for 3-60 months of age

Habilitation of Infants and Young Children

Now the question is, what if the infant or young child does have a bilateral vestibular loss in addition to the bilateral sensorineural hearing loss requiring the CI? What do we do? Unlike an acquired unilateral vestibular loss or hypofunction, with bilateral systemic vestibular loss, the individual cannot have completely normal equilibrium function. They will always be strongly visually and surface dependent, so their lifestyle and activities will need to accommodate these functional limitations. While the combined CI vestibular implant is on the horizon, it is still in the very early stages of development.

Pediatric physical and occupational therapy and family involvement and participation is the historical clinical pathway for these children. It is also important to address any other conditions that may coexist and affect the progression of therapy and maturational norms. This is often seen with premature infants, especially with low birth weights and other modality challenges, e.g., low vision and low muscle tone, who will be playing catch-up in addition to the auditory vestibular deficits. The rigors of scheduling and cost of physical therapy in addition to the speech and language habilitation therapy for the CI may place challenges and hardship on the child's family.

Summary

There are over 500 heritable conditions with auditory vestibular expressivity. It is estimated that as high as 20% to 80% of infants identified with bilateral sensorineural hearing loss and candidates for CI may also have a concomitant bilateral vestibular loss or hypofunction.[6] When a child is identified as a candidate for CI, it is incumbent on practitioners to also evaluate the child's vestibular function. Abnormal balance and equilibrium function can be addressed and treated in parallel with the CI and subsequent therapy, and should not be delayed. Quality of life and the child's safety and well-being are as important as normal speech and language development. ◗

Physical and occupational therapy can greatly benefit pediatric CI candidates with vestibular issues. Abnormal balance and equilibrium function can be addressed and treated in parallel with a cochlear implant and subsequent therapy.

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