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Pediatric Vestibular Disorders

Recognition, Evaluation and Treatment

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The vestibular system is important for the development of normal movement reactions, motion tolerance, and motor control for postural alignment, balance, and vision. A vestibular system that is damaged by disease or injury in childhood can have a major impact on a child's development. Despite advances in testing and documentation of vestibular deficits in children, vestibular problems continue to be an overlooked entity in children.^{1,2} Many children are not receiving treatment that could significantly improve function and address developmental delays caused by vestibular disorders.

Stages of development and the vestibular system

The vestibular organs provide sensory information about motion, and spatial orientation. The organs in each ear include the utricle, saccule, and three semicircular canals. The utricle and saccule detect gravity (vertical orientation) and linear movement. The semicircular canals detect rotational head movements and are located at right angles to each other. When these organs on both sides of the head are functioning properly, they send symmetrical signals to the brain that are integrated with other sensory and motors systems by age six.

If vestibular dysfunction occurs early in development, it slows the development of equilibrium and protective reflexes and motor-control tasks such as sitting unsupported, standing, and walking. In addition, an impaired vestibulo-ocular reflex (VOR) from vestibular dysfunction can have far-reaching impacts on a child's ability to keep pace with schoolwork. The VOR is responsible for maintaining clear vision during rapid head movements. Stable vision is important for learning to read and write and for developing fine and gross motor control. If left untreated, a vestibular disorder can have adverse consequences for a range of functions as the child grows to adulthood.

Signs and symptoms

Vestibular disorders are not as easily recognized in children as they are in adults, in part because children often cannot describe their symptoms well and may be unable to understand the concepts of vertigo and imbalance. Identification of pediatric vestibular dysfunction requires coordinating descriptions offered by the child, symptom reports from parents, and clinical observations by professionals.

Symptoms and signs that may indicate vestibular dysfunction include:

- Dizziness and visual acuity problems, especially with head movements such as when turning to look at something
- Poor spatial relationships, sometimes revealed by skipping words or letters while reading or by having a disorganized writing style.³
- Nystagmus (involuntary, alternating, rapid, and slow eye movements)³⁴
- Difficulty navigating in the dark
- Hearing loss or tinnitus (ringing in the ears)

- Motion sickness or sensitivity (avoids or craves movement)^{5,6}
- Nausea
- Abnormal movement patterns, unsteady gait, clumsiness (including decreased eye-hand and eye-foot coordination), or poor posture— including a tendency to fall, lean, or tilt over
- Ear pressure
- Headaches with associated nausea and/or dizziness
- Developmental and reflex delays^{7,8} that are sometimes revealed by slower achievement of milestones such as riding a bicycle, swimming, hopping, and stair climbing involving alternating left-right leg movements

For each child, the specific set of signs and symptoms will differ based on whether the damage is peripheral (involving the organs in the inner ear), central (involving the brain and brain stem), or both. The signs and symptoms will also depend on whether the damage is unilateral (on one side) or bilateral (affecting both ears), and whether the disease or injury has caused a complete loss of function, reduced function (hypofunction), or increased sensitivity (hyperfunction).⁹

Causes of vestibular dysfunction

The interconnection of the vestibular system with many other body systems can result in vestibular dysfunction secondary to a range of medical conditions and histories. Histories sometimes associated with peripheral or central system dysfunction include:

- Chronic ear infections or otitis media^{10,11}
- Congenital sensorineural hearing loss^{1,12,13,14}
- Cytomegalovirus and other viral infections such as in Ramsay Hunt syndrome (an infection of the facial and cochleovestibular nerves caused by the herpes zoster virus, the same virus that is associated with chicken pox)
- Malformations from acquired or genetic conditions such as branchiootorenal syndrome, Mondini dysplasia, and Waardenburg syndrome
- Other genetic disorders such as Usher syndrome-type I (with severe profound sensorineural hearing loss and balance problems and deteriorating vision by age 10) or type III (with balance and vision problems appearing later in life)
- Anoxia (reduced oxygen at birth) or stroke

- Meningitis (inflammation of the membranes covering the brain and spinal cord, sometimes also affecting membranes in the inner ear)
- Neurological disorders or conditions such as cerebral palsy, hydrocephalus, a posterior brain tumor, or Wallenberg syndrome (caused by a stroke from blockage in the vertebral or posterior inferior cerebellar artery of the brain stem)
- Maternal drug or alcohol abuse during pregnancy^{15,16}
- Immune-deficiency disorders^{17,18}
- Metabolic disorders such as diabetes
- Vascular insufficiencies
- Head-neck trauma from car accidents or sports injuries¹⁹

Specific vestibular disorders that can occur in childhood include:

 Childhood paroxysmal vertigo (CPV), also known as benign paroxysmal vertigo (BPV), is the most common pediatric vestibular disorder associated with dizziness, and is sometimes referred to as migraine equivalent. It is a central vestibular disorder typically seen in children aged 2–12. It is characterized by true spinning vertigo, nystagmus, nausea, and vomiting. A child with CPV is often sensitive to motion, light, and sound, but is typically asymptomatic after sleeping. Children tend to grow out of CPV; however, sometimes CPV progresses into migraine-associated vertigo in adulthood. This disorder is also referred to with a variety of other names and acronyms, but it <u>is not the same as</u> benign paroxysmal positional vertigo (BPPV).

- Vestibular neuritis causes dizziness, and labyrinthitis causes both dizziness and hearing symptoms. Both result from severe ear infections and can cause acute symptoms of vertigo and nausea that usually, but not always, subside within 4– 6 weeks. These disorders are attributed to a viral infection of the trigeminal ganglion, a collection of nerve cells located just behind the ear, (vestibular neuritis) or bacterial or viral infection of the inner ear (labyrinthitis).
- Ototoxicity (ear poisoning) is caused by medications that destroy the hair cells of the inner ear, which transmit balance signals from the inner ear to the brain. Drugs that can cause significant bilateral damage are intravenously administered aminoglycoside antibiotics (such as gentamicin) and certain types of chemotherapies,²⁰ which in children can result in severe imbalance, falls, and visual-motor

problems, including oscillopsia (bouncing vision). Many children who have experienced ototoxicity have difficulties at school, working at a computer, or learning to drive an automobile.

- Ménière's disease is less common in children. It is a peripheral disorder involving an imbalance of an inner ear fluid called endolymph. Symptoms include fluctuating hearing loss, tinnitus, ear pressure or fullness sensations, and episodes of vertigo and nausea.
- Some less common vestibular disorders in children include these peripheral disorders: perilymph fistula (a tear in the oval or round window membranes of the inner ear), and enlarged vestibular aqueduct syndrome (an abnormally large tube connecting the inner ear's endolymph to the endolymphatic sac). Benign paroxysmal positional vertigo (BPPV), a condition caused by dislodged otoconia in a semicircular canal that abnormally stimulate movement-sensing nerve cells, is sometimes seen in children.^{21,22,23,24,25} However, it has a much lower incidence in children than in adults. It is also sometimes observed as a surgical complication of cochlear implantation.1,26,27

Evaluation

Diagnosing and evaluating a vestibular disorder appropriately involves collaboration among specialists. Medical evaluation is essential and will begin with a comprehensive history taking and physical exam by a physician. To help rule out non-vestibular causes of symptoms, this physician may order imaging tests such as a MRI or CT scan. A referral to a specialist (an otolaryngologist, ophthalmologist, otologist, or neurotologist) may lead to diagnostic tests that measure hearing, eye movement, and peripheral vestibular function (performed by an audiologist) and an assessment of balance and functional impairments (evaluated by physical and occupational therapists with advanced training in balance disorders).

Tests of vestibular and balance function

Vestibular function testing is designed to determine if peripheral unilateral or bilateral damage has occurred and whether there is a resulting absence of function, hypofunction, or hyperfunction.^{28,29} In most cases, vestibular function tests can be performed on children aged one year or older by specialists whose training in pediatric care equips them to modify the equipment and techniques as needed, provide assurances that minimize potential apprehension, and ensure cooperation.

Electronystagmography (ENG) or videonystagmography (VNG) is a group of eyemovement tests used to look for signs of vestibular dysfunction or neurological problems. ENG/VNG is used to measure the ability of the semicircular canals to stabilize vision with different stimuli such as with head rotation or movement in different planes, and with warm or cold water circulating in one ear canal (a caloric test). ENG/VNG can evaluate whether signals originating from one side are consistent (symmetrical) with signals from the other side or if responses are absent, hyperactive, or hypoactive. In addition, ENG/VNG can be used with offvertical axis rotation to test the function of the utricle.

Vestibular evoked myogenic potential (VEMP) testing evaluates whether the saccule and the inferior vestibular nerve are intact and functioning normally.³⁰ When sound is transmitted through earphones or via bone conduction with a specialized vibrator placed behind the ear, electrodes placed on the neck muscles record the response of the muscle to the vestibular stimuli.

Balance and posture evaluations address whether sensory information from the vestibular system coordinates and integrates with vision and somatosensory information from the muscles and joints. Such tests require the child to balance under various vision conditions (e.g., with eyes open and closed or with a moving visual field) and varied standing surfaces (e.g., on a firm, soft, or moving surface). For diagnostic purposes, balance testing may be performed with sophisticated equipment such as computerized dynamic posturography (CDP). For treatment evaluation purposes, CDP or simpler equipment such as a pillow of dense foam³¹ may be used. In addition, Standardized Developmental Motor Scales and other assessments employ various eye-hand and eye-foot coordination tasks, balance activities, fine motor tasks, and gaze stability measurements³² to evaluate the child's development, reflexes, and ability to use the vestibular system for balance, coordination, and visual-motor control.

Treatment

Evaluation and assessment results help determine the most effective treatment

plan. For example, dietary changes are critical in children with childhood paroxysmal vertigo.

Depending on the diagnosis, recovery can be complete or nearly complete with unilateral peripheral vestibular dysfunction. With bilateral and central problems, some adaptation is needed to achieve near normal function.

Children with vestibular disorders often respond well to a specialized form of therapy called vestibular rehabilitation therapy (VRT). If surgery is needed to stabilize or correct a condition, VRT can also help with post-surgery recovery. With unilateral peripheral or central dysfunction, VRT may focus on habituation and training in using remaining vestibular function; however, with bilateral dysfunction, VRT must focus on training substitution (i.e., heavier reliance on sensory information from the visual and somatosensory systems).

VRT is an exercise program tailored to address eye-movement control, dynamic visual acuity, balance, developmental reflexes, and body-movement functions. VRT exercises should be based on a child's age, interests, level of comprehension, and test results. For example, appropriate balance or eye-movement exercises for a child aged 4 may involve block designs or balancing on a therapy ball. For a child aged 7, the same goal may be accomplished with a visual maze or balance obstacle course.

VRT can be effective for reducing or eliminating vertigo, improving visualmotor control, improving balance and coordination,³³ improving visual acuity,¹³ and promoting normal development.

Children typically respond more quickly to VRT than adults, because of their greater plasticity—the ability of their neurological systems to more quickly compensate for and adapt to vestibular deficits. In addition, children tend to be less fearful of movement than adults, so they participate well in the balance and movement aspects of therapy, if the therapist keeps it fun and interesting.

Coping

The effectiveness of a child's VRT treatment program depends in part upon the cooperation, patience, and understanding of parents and caretakers in supporting compliance and progress. Parents should be provided with a specific home-exercise program to reinforce the VRT movements and activities performed at the clinic.

Teachers and occupational and physical therapists can also help integrate vestibular training into activities at school. They should work closely with the parents to identify the child's tolerances for vestibular therapy and to develop and modify the individualized therapy program accordingly. Calming activities should be included as part of the program in order to help the child avoid becoming overly stimulated, sick, frightened, or stressed. A well-structured VRT program can make a significant difference in a child's function, learning abilities, development, balance, and selfconfidence.

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