Fact Sheet

Enlarged Vestibular Aqueducts and Childhood Hearing Loss

What are vestibular aqueducts?

Vestibular aqueducts are narrow, bony canals that travel from the inner ear to deep inside the skull. The aqueducts begin inside the temporal bone, the part of the skull just above the ear. The temporal bone also contains two sensory organs that are part of the inner ear. These organs are the cochlea, which detects sound waves and turns them into nerve signals, and the vestibular labyrinth, which detects movement and gravity. These organs, together with the nerves that send their signals to the brain, work to create normal hearing and balance. Running through each vestibular aqueduct is a fluid-filled tube called the endolymphatic duct, which connects the inner ear to a balloon-shaped structure called the endolymphatic sac.

A vestibular aqueduct is considered enlarged if it is greater than 1.5 millimeters in size, roughly the diameter of the head of a pin. If a vestibular aqueduct is enlarged, the endolymphatic duct and sac usually grow large too. The functions of the endolymphatic sac and duct are not completely understood. Scientists currently believe that the endolymphatic sac and duct help to ensure that the fluid in the inner ear contains the correct amounts of certain chemicals called ions. Ions are needed to help start the nerve signals that send sound and balance information to the brain.

How are enlarged vestibular aqueducts related to childhood hearing loss?

Research suggests that most children with enlarged vestibular aqueducts (EVA) will develop some degree of hearing loss. Scientists also are finding that five to 15 percent of children with sensorineural hearing loss, or hearing loss caused by damage to sensory cells inside the cochlea, have EVA. However, scientists do not think that EVA causes hearing loss. Instead, scientists regard EVA as an important clue about hearing loss and its possible causes. This information helps physicians talk with families about how their child's hearing loss may change over time.

The presence of EVA can be a symptom of a genetic disorder called Pendred syndrome, a cause of childhood hearing loss. According to a study by the National Institute on Deafness and Other Communication Disorders (NIDCD) in the United States, approximately one-third of individuals with EVA and hearing loss have Pendred syndrome. With Pendred syndrome, the hearing loss is progressive, which means that a child will have less hearing over time. Some children may become totally deaf.

In addition to its association with hearing loss, EVA may also be linked with balance symptoms in a small percentage of people. However, the brain is very good at making up for a weak vestibular system, and most children and adults with EVA do not have a problem with their balance or have difficulty doing routine tasks.

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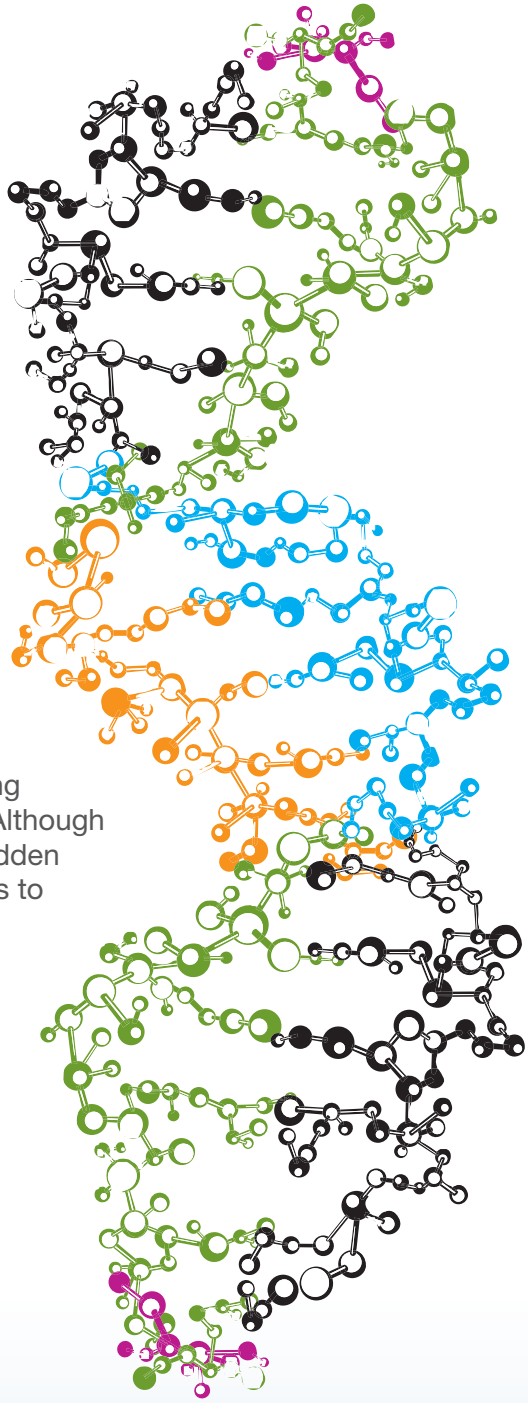
What causes enlarged vestibular aqueducts?

EVA has many causes, not all of which are fully understood. The most well-known cause of EVA and hearing loss is mutations to a gene known as SLC26A4 (also referred to as the PDS gene) on chromosome 7. Two mutations in the PDS gene can result in Pendred syndrome. Scientists believe that other, currently unknown genetic or environmental factors also may result in EVA.



How are enlarged vestibular aqueducts diagnosed?

Medical professionals use different clues to help them determine the cause of an individual's hearing loss. Two tests that are often used to identify the cause of hearing loss are magnetic resonance imaging (MRI) and computed tomography (CT) imaging of a person's inner ear. One or both tests are often recommended to evaluate a child with sensorineural hearing loss. This is particularly true when a child's hearing loss occurs suddenly, is greater in one ear than the other, or varies or gets worse over time. Although most CT scans of children with hearing loss are normal, EVA is the most commonly observed abnormality.

Can enlarged vestibular aqueducts be treated to reduce hearing loss?

No treatment has proven effective in reducing the hearing loss associated with EVA or in slowing its progression. Although some otolaryngologists recommend steroids to treat sudden sensorineural hearing loss, there are no scientific studies to show that this treatment is effective or ineffective when an individual also has EVA. In addition, surgery to either drain liquid out of the endolymphatic sac and duct or to remove the endolymphatic sac and duct is not only ineffective in treating EVA, it can be harmful. Research has shown conclusively that these surgeries can destroy hearing.

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To reduce the likelihood of progression of hearing loss, individuals with enlarged vestibular aqueducts should avoid contact sports that might lead to head injury; wear head protection when engaged in activities that might lead to head injury (such as bicycle riding or skiing); and avoid situations that can lead to barotrauma (extreme, rapid changes in pressure), such as scuba diving or hyperbaric oxygen treatment.

Identifying hearing loss as early as possible is the best way to reduce its effect. The earlier hearing loss is identified in children, the sooner they can develop the skills that will help them learn and communicate with others. Children with permanent and progressive hearing loss, which often is linked with EVA, will benefit from learning other forms of communication, such as sign language or cued speech, or using assistive devices, such as a hearing aid or cochlear implant.

What research is being conducted on enlarged vestibular aqueducts and hearing loss?

While mutations of the SLC26A4 gene are known to cause EVA, not all EVA cases are the result of an SLC26A4 mutation. Some cases may be caused by other genetic or environmental factors. For these reasons, researchers are currently conducting a clinical trial to identify and understand the various factors that can lead to EVA and hearing loss.

Adapted with permission from the National Institute on Deafness and Other Communication Disorders.