

## Enlarged Vestibular Aqueduct (EVA)

### What is an enlarged vestibular aqueduct (EVA)?

The **vestibular aqueduct** is a narrow tube filled with fluid in the inner ear. When that tube is wider than usual, it is called an **enlarged vestibular aqueduct (EVA)** or a **large vestibular aqueduct (LVA)**.

People with an EVA have permanent hearing loss in one or both ears and balance problems. This hearing loss can be present at birth, or it can be **progressive** (get worse over time) throughout life. Sometimes a head injury can cause a permanent change in hearing.

### How is an EVA diagnosed?

An EVA is diagnosed by a medical professional, typically an **otolaryngologist** (an ear, nose, and throat doctor). They will use imaging tests, such as an MRI or CT scan, of the inner ear to figure out the cause of your hearing loss.

- An **audiologist** (a doctor specializing in diagnosing and treating hearing loss) will do a hearing test with you to understand how well you can hear and how much hearing loss you may have.
- It is important to monitor (regularly check) your hearing because hearing can progress (change over time). It can be hard for a child with EVA to explain any changes they notice in their hearing in one or both ears, so routine monitoring is important for management and treatment.

# What are other conditions that someone might have if they have an EVA?

An EVA can commonly happen in people who have Pendred syndrome, so genetic testing can be helpful. Some syndromes can cause other significant health issues, and it is important to follow up with the appropriate specialist. Some other conditions that can happen with an EVA include:

- Waardenburg syndrome
- CHARGE syndrome
- Branchio-oto-renal (BOR) syndrome

An EVA can also happen with another inner ear **malformation** (when a body part develops in a way that is not normal) called **Mondini dysplasia**. This is when the **cochlea** (a spiral-shaped part of your inner ear that sends hearing information to your hearing nerve) has less than the usual 2.5 turns.

Children with an EVA may also have **vestibular** (balance) issues, so a vestibular audiologist or physical therapist can be helpful if needed.

### How is an EVA treated?

- For hearing loss caused by an EVA, there is no treatment to reduce hearing loss or stop hearing loss from getting worse over time.
- Depending on how much hearing loss you have, devices like hearing aids or cochlear implants can help you hear soft sounds. Hearing these soft sounds is important to help you develop speech (talking) and understand others.
- We recommend that you get your hearing tested regularly to see if there have been hearing changes, or sooner if you notice a significant change in hearing. Head trauma (injury to your head) can cause your hearing to get worse with EVA, so also repeat the hearing test if you hit your head.

• If you have balance issues, you may get a recommendation for vestibular therapy. **Vestibular therapy** is a type of physical therapy, involving exercises that improve dizziness and problems with balance caused by inner ear disorders.

#### Where can I find more information on EVAs?

Vestibular Disorder Association (VeDA): <u>vestibular.org</u>	
National Institute on Deafness and Other Communication	
Disorders	
• Enlarged Vestibular Aqueducts and Childhood Hearing	
Loss: www.nidcd.nih.gov/health/enlarged-vestibular-	
aqueducts-and-childhood-hearing-loss	

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