



ENLARGED VESTIBULAR AQUEDUCT (EVA)

This handout is intended as a general introduction to the topic. As each person is affected differently, speak with your health care professional for individual advice.



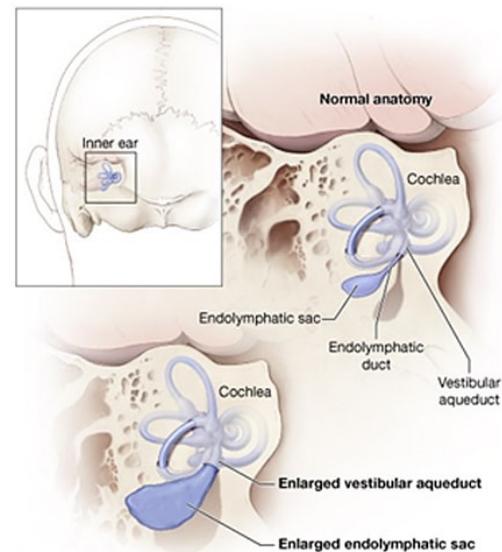
Key points

- Larger-than-normal narrow tube running from the inner ear to deep in the skull.
- Seems to happen before birth during fetal development.
- Likely caused by an underlying genetic problem.
- Sometimes happens on its own and sometimes along with other syndromes.
- Symptoms include sensorineural hearing loss, vertigo (spinning sensation), and balance problems.
- Most often affects young people, from toddlers to those in their 20s.
- Hearing protection may be suggested to prevent sudden hearing loss after a minor head injury or a sudden, extreme change in air pressure.
- So far, no treatment can reduce or slow hearing loss.
- Cochlear implantation may be an option but carries some risk.

What is enlarged vestibular aqueduct?

The vestibular aqueduct (ves-TIB-you-lar AC-wa-duct) is a narrow tube that runs from the inner ear to deep in the skull. It contains the endolymphatic (EN-doe-lim-FAT-ick) duct, which connects the inner ear to a structure called the endolymphatic sac. Endolymph is the fluid in the inner ear.

Normally, the vestibular aqueduct is very narrow: halfway along its length, it is less than 1 millimeter (mm) across—about the width of a pin. But some people have a wider vestibular aqueduct, measuring 1.5 mm to 8 mm across. This is known as enlarged vestibular aqueduct (EVA).



Enlarged vestibular aqueduct

People with EVA often have sensorineural hearing loss, with or without vertigo and balance problems. These problems often start in childhood; between 5% and 15% of children with sensorineural hearing loss have EVA. These problems are more common in girls than in boys, and they are more often found on both sides (bilaterally) than on one side.

Typically, the person's hearing gradually gets worse over time.

EVA was first identified in 1791, but the association between EVA and hearing loss was first described in 1978.

What are the causes?

EVA seems to happen before birth, while the fetus is developing. The enlarged aqueduct does not seem to cause hearing loss. Instead, researchers believe that both EVA and hearing loss are caused by an underlying genetic problem.

Many people with EVA and hearing loss also have other structural changes in the inner ear. For example, EVA is most commonly associated with incomplete partition type 2 (also called the Mondini malformation). This abnormality consists of:

- cochlea (part of the inner ear that looks like a snail) with only 1.5 turns instead of the usual 2.5
- enlarged vestibule with normal semicircular canals
- enlarged vestibular aqueduct containing a dilated (wider) endolymphatic sac

EVA sometimes happens alone and sometimes with other disorders. For example, about 1 in 4 people with EVA and hearing loss have Pendred syndrome, which also causes thyroid problems. Mutations in a gene called SLC26A4 can cause EVA and Pendred syndrome. But not everyone with EVA has these mutations. We are still learning about other things that can cause EVA.

Other conditions that sometimes happen with EVA include:

- CHARGE syndrome
- distal renal tubular acidosis
- Waardenburg syndrome
- X-linked congenital mixed deafness
- branchio-oto-renal (BOR) syndrome
- oto-facial-cervical syndrome

What are the symptoms?

Hearing symptoms of EVA may include:

- sensorineural hearing loss, such as trouble distinguishing words or a child not responding when their name is called
- for children, speech, and language delays

Balance symptoms of EVA may include:

- episodes of vertigo that vary in length
- balance problems
- for children, delays in crawling or walking

Sometimes, the symptoms are first noticed or get worse after a head injury.

How is it diagnosed?

EVA is most often diagnosed in young people, from toddlers to people in their 20s. This section assumes that you are a parent who is concerned about your child's symptoms.

EVA may be diagnosed by a pediatrician or another primary care doctor, but it is more often diagnosed by a specialist, such as an otolaryngologist or an otologist.

The doctor will ask about your child's symptoms. Try to be as specific as possible about the symptoms and when they get better or worse.

The doctor will also ask about your child's medical history, including any medications they are taking or recently stopped taking and any conditions they have been diagnosed with in the past. The doctor will also do a thorough physical and neurological exam.

Your child will probably have some of the following diagnostic tests:

- hearing and vestibular function tests
- an imaging test such as magnetic resonance imaging (MRI) or computed tomography (CT); these tests can show if the vestibular aqueduct is larger than usual and if there are any other abnormalities in the temporal bone

How is it treated and managed?

So far, there is no treatment to reduce or slow hearing loss in people with EVA. It is important to identify hearing loss as early as possible, so that children and their caregivers can develop skills to help with communication. These skills may include sign language and cued speech, which uses hand signals and other visual signs to make the “building blocks” of speech (phonemes) visible.

You or your child will need regular follow-up care to monitor hearing and see if there have been any changes. This is especially important for very young children, who may not be able to tell you if something is wrong. Hearing aids will be recommended when there is significant hearing loss.

Surgery

Surgery to drain excess fluid or remove the enlarged endolymphatic duct and sac is not helpful. It often causes severe hearing loss.

Some people with hearing loss and EVA may lose hearing gradually to a degree where hearing aids are no longer helpful. They may become candidates for cochlear implants. Surgery for cochlear implants does carry some risk. People with EVA are more likely to have serious leakage of cerebrospinal fluid (CSF) during surgery. CSF is the fluid that surrounds the brain and spinal cord. It is important to consider the risks and benefits of surgery carefully and discuss them with the surgeon.

Preventing injury

About 1 in 3 people with EVA have sudden hearing loss after a minor head injury or barotrauma (sudden, extreme change in air pressure). For this

reason, people with EVA are often told to try to protect their hearing by:

- avoiding contact sports
- wearing head protection for activities like bike riding or skiing
- avoiding loud noises
- avoiding situations that cause extreme, fast changes in air pressure, such as scuba diving

avoiding playing wind instruments

taking decongestants when flying if they have sinus or nasal congestion

The risk seems to be higher in people whose hearing changes a lot (fluctuates). Talk to your health care team about whether certain activities are risky.

What to expect in the future

In many people with EVA, hearing changes a lot or suddenly gets worse. It is not possible to predict who will have these sudden changes, so regular monitoring is important.

Researchers are studying the factors that cause EVA and hearing loss. In the future, we may know more about what causes them and how to prevent them.

Visit our website

View this and other articles about vestibular disorders – www.balance&dizziness.org.

In addition, find information about how the balance system works, the journey from diagnosis to treatment, building a wellness toolkit, and more.

Handout updated September 2021

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