WHAT IS AN ACOUSTIC NEUROMA?

Acoustic neuromas, also referred to as vestibular schwannomas, are non-cancerous (benign) tumors that develop from the eighth cranial nerve, between the ear and the brainstem. The eighth cranial nerve, often referred to as the “hearing and balance nerve,” contains three smaller nerves - two balance (or vestibular) nerves and one hearing (or auditory) nerve - and is collectively responsible for sending balance and sound signals from the ear to the brain. Much like a typical electrical wire, the eighth cranial nerve conducts electricity and has an outer layer of insulation. A special layer of cells, called Schwann cells, create the insulation around the nerve. Acoustic neuromas are tumors that develop from the insulating Schwann cells on one of the two vestibular nerves. Therefore, the term “vestibular schwannoma” is technically more accurate than the term “acoustic neuroma,” despite both being used interchangeably.

As acoustic neuromas are non-cancerous, they do not spread to the lungs, bones, or other parts of the body and do not invade the brain. Instead, virtually all the problems that can develop from an acoustic neuroma result from the tumor applying pressure on adjacent important structures, such as the brainstem, cerebellum, and other neighboring nerves.

Acoustic neuromas occur equally in men and women and most commonly develop between the ages of 45 and 70 but may occur at any age. Overall, they represent 8% of all brain tumors and are the most common tumor...
to develop between the ear and the brainstem. While historically thought to be quite rare, today an increasing number of people are diagnosed with acoustic neuroma. Recent research has suggested that approximately one in 2000 adults, and up to one in 500 adults over the age of 70, will be diagnosed with an acoustic neuroma during their lifetime. Therefore, these tumors are more common than previously thought.

**WHAT CAUSES ACOUSTIC NEUROMAS?**

Virtually all acoustic neuromas have at least one mutation involving an important gene located on chromosome 22. When this gene is healthy (not mutated), it prevents the Schwann cells from growing into a tumor. However, when this gene is mutated, a tumor may form. Unfortunately, in most cases, we do not know why this mutation develops in the first place. Approximately 95% of all acoustic neuromas occur sporadically because of random mutations in the affected gene, meaning they are not part of an inherited genetic syndrome. There is no convincing evidence that certain foods, smoking, or other environmental factors increase the risk of developing an acoustic neuroma. Some publications suggest that a history of loud noise or cell-phone use may cause acoustic neuromas to develop; however, this is very controversial, and based on current information, these factors likely do not cause acoustic neuromas. Aside from some rare factors, such as radiation exposure at a young age, there is no identifiable cause to explain why most acoustic neuromas develop.

While 95% of people with sporadic acoustic neuromas only have one tumor, 5% of patients affected will develop acoustic neuromas on both the left and right sides and may also develop other non-cancerous brain and spinal tumors such as meningiomas, ependymomas, and other schwannomas – a condition called Neurofibromatosis Type-2 (NF2). NF2 is a genetic disorder that may be passed on from a father or mother to their child; however, approximately half of people who are diagnosed with NF2 do not have a family history of the disease. The diagnosis of NF2 can be firmly made in people who have two acoustic neuromas, or one acoustic neuroma with a family history of NF2, or one acoustic neuroma and a combination of other brain or spinal tumors. In some cases, a diagnosis of NF2 may be suspected but not yet confirmed in people who develop acoustic neuroma at a young age. In these cases, genetic testing may be ordered. If you are only diagnosed with one acoustic neuroma and have no other brain or spinal tumors and do not have a family history of NF2, it is unlikely that you will ever develop this condition.

**HOW ARE ACOUSTIC NEUROMAS DIAGNOSED?**

In most cases, the first symptoms that develop with an acoustic neuroma are hearing loss and tinnitus (i.e., ringing in the ear) on the side with the tumor. Hearing loss occurs in approximately 90% of cases and tinnitus occurs in approximately 70% of cases. It is important to understand that hearing loss and tinnitus are the most common symptoms of having an acoustic neuroma; however, most people with hearing loss and tinnitus DO NOT have a tumor.

The hearing loss from an acoustic neuroma may occur slowly over time, which may be hard to notice at first. Less commonly (approximately 10% of cases) it may occur suddenly. The hearing loss is most commonly apparent when using a telephone with the affected ear, or when going to bed at night when the “good” ear is against a pillow. These symptoms will often prompt a doctor to order a hearing test, called an audiogram.

If the hearing test confirms that one ear has more hearing loss than the other, then a magnetic
A resonance imaging (MRI) scan of the head is typically ordered. Historically, other tests have been used to help diagnose acoustic neuromas including auditory brainstem responses, but these tests have been shown to be less accurate in diagnosing acoustic neuromas than MRI. Currently in the United States, the gold standard test used to diagnose an acoustic neuroma is an MRI scan of the head with contrast. If you are unable to get an MRI, a computed tomography (CT) scan may be ordered instead. Acoustic neuromas have a classic appearance on MRI such that a tumor biopsy is almost never needed to establish a diagnosis. Rather, in most cases, a diagnosis can be made solely based on the results of the MRI.

HOW ARE ACOUSTIC NEUROMAS TREATED?

It is important to understand that each patient’s situation is unique: the type and severity of symptoms, size and shape of the tumor, age and health of the person, and goals of treatment may all be different from person to person. The internet is overflowing with information ranging from individual experiences to large-scale data, so it can be quite hard for patients to decipher what is relevant to them. Thus, the most important thing for a person diagnosed with acoustic neuroma to do is seek opinions from experienced doctors for a patient-centered, individualized approach to care.

Acoustic neuroma treatment requires an experienced and specialized multidisciplinary team. Broadly, acoustic neuromas can be managed with observation, surgery, radiation, or a combination of these treatment options.

Except for very large tumors, it is not an emergency to treat most acoustic neuromas. After diagnosis, you should take the time needed to understand your condition and feel comfortable with your treatment decision before moving forward. The ANA recommends getting more than one opinion from experienced centers about treatment when possible.

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