

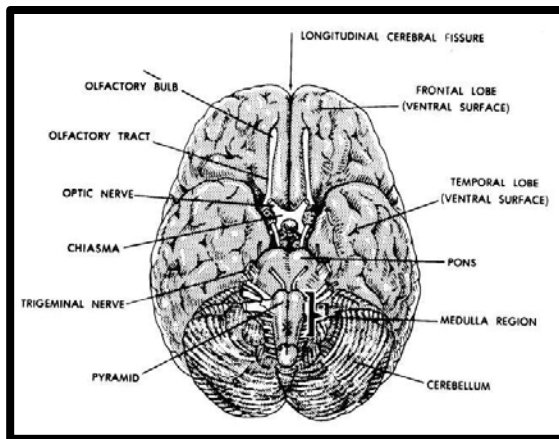
A GLIMPSE OF THE BRAIN

THE ACOUSTIC NEUROMA CONNECTION

GENERAL REVIEW

The nervous system is composed of the brain, spinal cord and peripheral nerves. Each part of the nervous system is linked, either directly or indirectly, to each other part. The brainstem, located at the base of the brain, connects the higher brain centers to the spinal cord. Cranial nerves enter and exit the brainstem to receive sensory information from the head, neck and internal organs, and to send motor signals to the same structures. Spinal nerves enter and exit from the spinal cord to serve similar functions in the rest of the body. An understanding of the anatomy and function of the brain, brainstem and cranial nerves can provide insight about the symptoms an acoustic neuroma may cause. This discussion may also explain the physical changes some patients have after tumor removal.

Figure 1. Bottom view of the brain with cranial nerves indicated

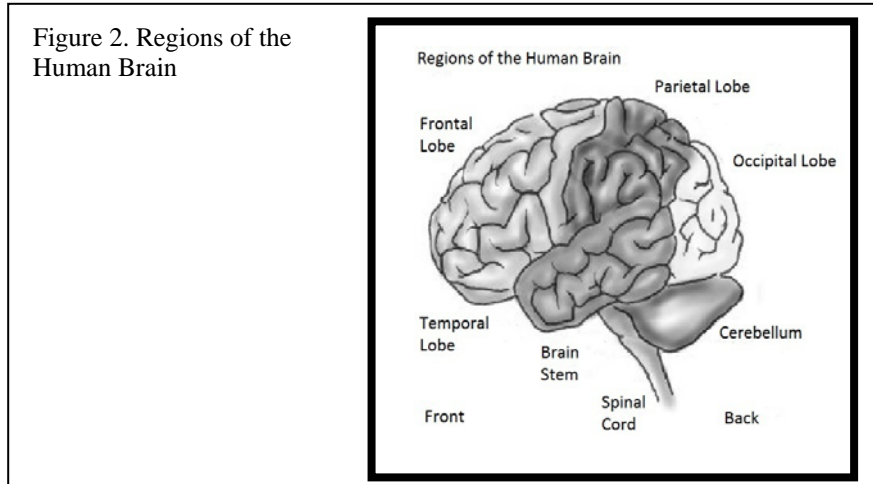


The largest parts of the brain are the cerebral hemispheres. This is the “thinking” part of the brain, most often associated with conscious thought and complex handling of information and reasoning. In general, the right cerebral hemisphere receives information from and controls the left side of the body, and visa-versa. The two hemispheres are connected via a central structure called the corpus callosum. There are, however, many more interactions between the cerebral hemispheres and deeper structures, including the brainstem.

Each cerebral hemisphere is divided into four lobes: frontal, temporal, parietal and occipital. It is not possible in a brief review to discuss all of the anatomic-function correlations. Another important distinction is between eloquent areas and non-eloquent areas. That is, some areas of the cerebral cortex have specific, understood functions, while others serve a more undefined purpose. Damage to an eloquent area will result in a typical neurologic deficit, while damage to non-eloquent areas may not cause any perceptible problem.

In general terms, the frontal lobes are concerned with action and active thought, with imagination and emotions at the very front, and physical movement further behind. The parietal lobes are concerned with perception, including sensation and spatial perception. The occipital lobes are concerned with visual perception.

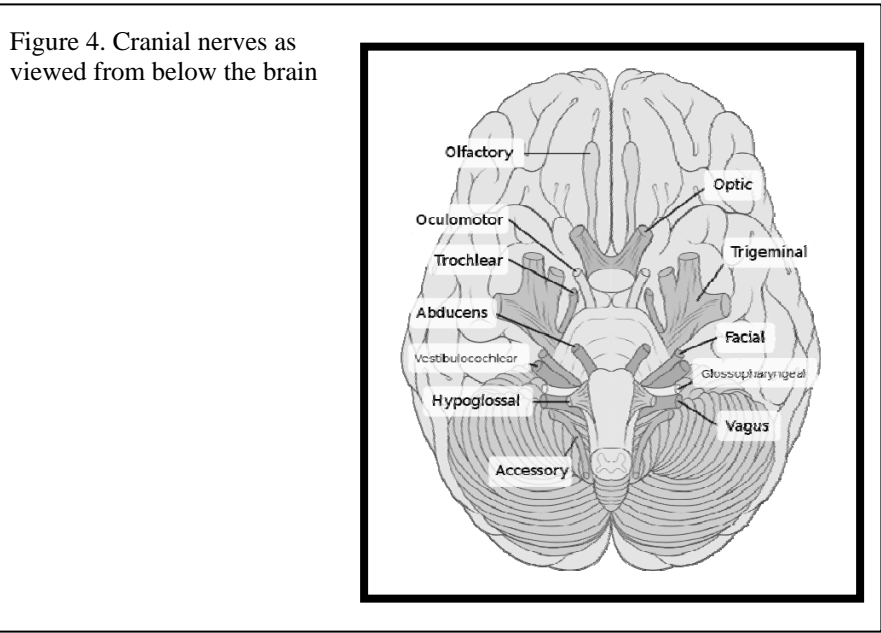
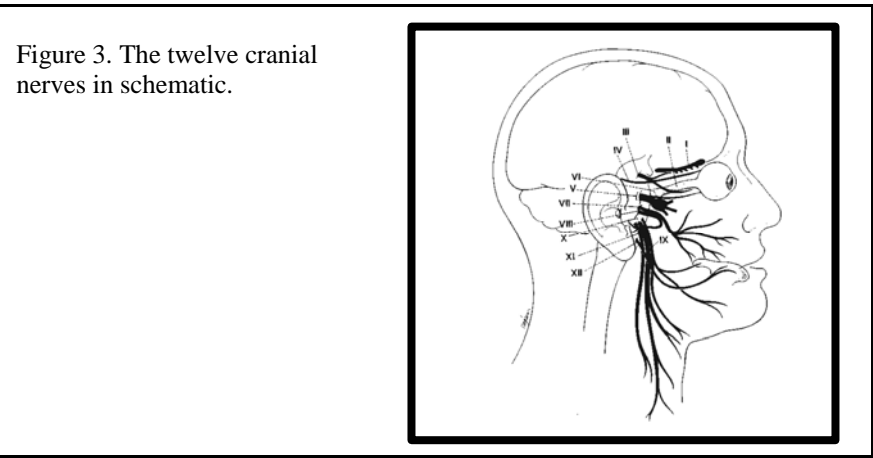
For most people, the ability to speak and understand speech is located primarily in the left cerebral hemisphere (in parts of the frontal, parietal and temporal regions). The temporal lobes are also concerned with other important functions, namely memory and audition.



Within the deeper areas of the brain are central controlling and connecting structures called the basal ganglia and thalamus. These areas coordinate more basic functions of action and sensation that are not controlled consciously. At the base of the front of the brain, in the middle, is an area called the hypothalamus, connected by a short stalk to the pituitary gland. These two structures are concerned with hormonal control of the body.

The brainstem lies at the base of the back of the brain. It is the structure that connects the upper parts of the brain to the spinal cord. Cranial nerves also exit the brain via the brainstem. The brainstem is one of the locations in the nervous system in which pathways cross from right to left, so that “the right side of the brain controls the left side of the body.” In addition, the brainstem contains the areas of the brain controlling the most basic brain functions, including reflexes, or automatic responses, and wakefulness. From top to bottom, the brainstem consists of three parts: midbrain, pons and medulla. The cerebellum is connected to the back of the brainstem at the pons. This structure controls coordination (the right cerebellar hemisphere controls the right side of the body) and the ability to walk. While the brainstem is densely packed with various critical structures, much of the cerebellum is non-eloquent.

The brain, brainstem and the spinal cord are bathed in a watery fluid called cerebrospinal fluid which is continually being formed and absorbed. In addition to flowing over the surface, the fluid circulates in a system of spaces in the brain called ventricles. The brain is supplied by an extensive network of arterial blood vessels and blood is drained by a complex system of veins.



Historically, these cranial nerves are designated with Roman numerals.

- I. Olfactory**—enters the undersurface of the frontal part of the cerebral hemispheres. It carries sensation of smell from the nose to the brain.
- II. Optic**—enters the undersurface of the cerebral hemispheres, very close to the hypothalamus. It carries visual impulses from the retina to the brain.
- III. Oculomotor**—arises from the side of the mid-brain and supplies four of the six muscles that move the eye and the muscle that elevates the eyelid.
- IV. Trochlear**—arises from the mid-brain and supplies one muscle that moves the eye.
- V. Trigeminal**—enters the side of the pons and carries sensation from the inside of the mouth, teeth, anterior aspect of the tongue and skin of the face; supplies neural control for the movement of the muscles of the jaw.
- VI. Abducens**—arises from the front of the pons and supplies one muscle that moves the eye.
- VII. Facial**—arises from the side of the pons and controls the muscles that move the face and the lacrimal (tear) and salivary glands and carries taste sensation from the anterior tongue.
- VIII. Vestibulocochlear** – enters the side of the pons and consists of two parts: the cochlear nerve which carries auditory impulses to the brainstem and the vestibular nerve which transmits impulses from the inner ear compartments which affect balance.
- IX. Glossopharyngeal**—arises from the side of the medulla and supplies the parotid gland and carries taste sensation from the posterior third of the tongue.
- X. Vagus**—arises from the side of the medulla and supplies muscles of the vocal cord and those involved with swallowing; gives motor function to smooth muscles and secretory glands in the respiratory and gastrointestinal tracts; is involved with control of cardiac function.
- XI. Accessory**—arises from the lower medulla and upper cervical spinal cord and supplies muscles that elevate the shoulder.
- XII. Hypoglossal**—arises from the lower medulla and supplies muscles which move the tongue.

ANATOMIC BASIS FOR ACOUSTIC NEUROMA SYMPTOMS

The middle and inner ear structures are located within a bone called the petrous temporal bone, part of the base of the skull that extends from the ear toward the center of the head. An opening in the posterior surface of this bone, called the internal auditory meatus, leads to the internal auditory canal, which contains the vestibulocochlear nerve. The facial nerve also enters the internal auditory canal along its course from brainstem, through the temporal bone, to the face.

Typically, acoustic neuromas grow from Schwann cells that make up the cells that create insulation for the nerves. The tumor usually starts in the bony confines of the internal auditory canal. With time, the enlarging tumor may even cause the bone to remodel and the internal auditory canal to become enlarged. Additionally, the tumor may grow out of the internal auditory canal towards the brainstem.

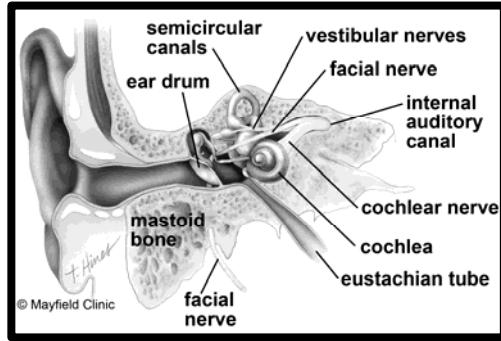
As the tumor grows, it usually first interferes with the function of the vestibular and cochlear nerves. The facial nerve is usually very resilient to a growing tumor, despite the fact that it may be slowly stretched and flattened. When the tumor becomes larger, it starts to compress the brainstem and fifth cranial nerve. A very large tumor may markedly compress the brainstem and the cerebellum and even become life threatening. This is very rare in the developed world, since patients usually present with the earlier less severe symptoms. Usually these tumors are very slow growing such that the brain can accommodate to the enlarging mass by slowly “getting out of the way.”

Functions of the cerebral hemispheres are affected only with large tumors, which may compress the brainstem so much that the normal flow of cerebrospinal fluid is obstructed along its normal course from ventricles, through the brainstem, to the surface of the brain. This causes enlargement of the ventricles, or hydrocephalus, which can be life-threatening.

AUDITORY SYMPTOMS

The first symptom from the tumor is usually some disturbance in hearing, such as the inability to hear on the telephone, tinnitus (ringing, buzzing, roaring, clicking, etc.) and/or fullness in the ear. This is probably due to pressure on the cochlear portion of the eighth nerve. Sensory nerves are typically more sensitive to pressure than motor nerves. The cochlear is much more vulnerable in most patients since all of the blood supply to the cochlear nerve comes from the internal auditory artery.

Figure 5. The normal anatomy of the ear.
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This artery originates inside the head and passes through the internal auditory canal with the nerves, and, therefore, will also be involved with the tumor. Thus impingement of the tumor primarily affects hearing though the origin of the tumor is on the vestibular nerve. This may manifest as sudden severe hearing loss or just fluctuations in hearing or as slow steady progressive hearing loss on one side. Some patients, even with large tumors, have no hearing loss. It is not really possible to predict how any single tumor is going to affect hearing, although typically, hearing will deteriorate over time.

After treatment either with surgery or radiosurgery, the patient may be left with worse hearing due to the intimate relationship of the cochlea, cochlear nerve and the blood supply to these structures to the tumor. In select patients usually with small tumors, it is possible to spare these structures and preserve hearing.

UNSTEADINESS AND VERTIGO

Even though the tumor starts on the vestibular portion of the eighth nerve, there are often no vestibular symptoms due to involvement of this nerve because the nervous system adapts quite well to gradual loss of function in this nerve.

In some patients, unsteadiness or vertigo may be early symptoms due to involvement of the vestibular nerve. It is rare for these symptoms to be severe they are more typically mild or episodic. In patients with larger tumors, pressure on the cerebellum and/or brainstem may cause difficulty with balance. Furthermore the development of hydrocephalus can also compromise balance and walking.

Some patients go through a period of dizziness and difficulty with balance after surgical removal of the tumor. This is typically due to abrupt loss of function in the vestibular nerve fibers that were divided to remove the tumor. These symptoms usually subside over days to weeks. In patients with large tumors, the involvement of the cerebellum and brainstem or its blood supply may be the cause of persisting symptoms. Patients who undergo radiosurgical treatment may experience fewer balance problems immediately after treatment; but may have recurrent episodes of

imbalance or vertigo in the future depending on how their vestibular system and tumor responds to the treatment.

ANA's publication *Improving Balance Associated with AN* offers more details about balance issues.

FACIAL WEAKNESS/EYE TEARING

Because the vestibulocochlear nerve and the facial nerve are intimately anatomically associated, every acoustic neuroma will involve the facial nerve. Pre-treatment facial weakness is extremely uncommon, however, because the facial nerve is quite resistant to pressure or stretching from these slow growing tumors. Exceptions include some patients noting intermittent hemifacial spasm, or slowing or difficulty blinking the eye on the side of the tumor.

In the overwhelming majority of surgeries excising the tumor, the anatomical integrity of the facial nerve is preserved. In cases where there is postoperative facial weakness, it is usually due to the reaction in the nerve after separating it from the tumor capsule. The majority of these cases of facial weakness following surgery are temporary, improving in the weeks to months after surgery. In the rare cases where the facial nerve is cut or otherwise significantly injured, a complete facial paralysis occurs with the affected side of the face drooping. If the nerve is cut, this weakness will be permanent. If it is significantly injured, there is often a significant improvement in function which may take more than a year. A small percentage of radiosurgical patients also experience facial weakness post-treatment. If there is inability to close the eyelids, a minor procedure may be required to improve this function. For nerves that are not expected to recover, surgical procedures may be performed to improve facial function. The risk to the facial nerve with acoustic neuroma treatment is proportional to the size of the tumor.

A dry eye is due to the loss of innervation (nerve supply) through the facial nerve to the lacrimal gland that provides natural lubricating tears to the eye. Occasionally this portion of the facial nerve "over recovers" and carries excessive impulses so that too many tears are produced, especially when chewing. This is known as "crocodile tears."

Facial nerve fibers also serve the secretory glands in the nose, so this function may be affected by either excessive or inadequate secretion.

Taste buds to the tip of the tongue are conveyed by the facial nerve. Temporary taste disturbance frequently occurs following acoustic tumor treatment.

FACIAL NUMBNESS

As the tumor becomes larger, there may be pressure on the trigeminal (fifth) cranial nerve and the adjacent pons, causing numbness of the face and/or numbness of the mouth or tongue. Usually this symptom subsides with removal of the tumor, but occasionally the numbness is worse.

A serious problem occurs when there is a facial paralysis with inability to close the eye and sometimes loss of sensation on the cornea. Special ophthalmologic care is required to protect the cornea.

FACIAL PAIN

This is an uncommon symptom and is due to pressure on the trigeminal nerve. The description of the pain may be similar to that seen with trigeminal neuralgia with sudden stabs of severe pain. Fortunately this problem is rare after surgery.

ANA's publication *Facial Nerve and AN: Possible Damage & Rehabilitation* offers more details about post-treatment facial issues and *Eye Care after AN Surgery* provides information on treatment of eye issues.

INCOORDINATION IN EXTREMITIES

The control of coordination in the extremities is located in the cerebellar hemispheres and adjacent brainstem. For example, a right-side tumor is associated with difficulty in the right hand. In some large tumors, a problem in coordination may occur causing difficulty in using the hand for daily activities or further affecting walking, balance and coordination. Often a program of physical therapy is required to aid recovery in the post-treatment period.

DOUBLE VISION (DIPLOPIA)

Double vision may be due to a loss of function in either the nerves supplying the eye muscles or the brainstem centers that coordinate eye movement. The third cranial nerve comes off the mid-brain above the tumor and is not affected. In large tumors, the fourth cranial nerve may be involved with the superior (upper) portion of the tumor. Patients often complain of "double vision" for the first couple of days after tumor removal even though their eye movements and the nerves that control them are working normally. This is because the balance (vestibular) system and the eye movement (oculomotor) systems are tightly connected in the brainstem. This subjective feeling of double vision usually goes away with the immediate postoperative "dizziness" in a couple of days.

DIFFICULTY SWALLOWING (DYSPHAGIA)

Fortunately this is a very rare symptom both pre-treatment and post-treatment. When it does occur, it is due to involvement of the tenth cranial nerve fibers or occlusion of some of the blood supply to the brainstem.

HEADACHES

Pre-treatment headache is typically uncommon. When it does occur, it is usually associated with a large tumor. Some patients complain of aching in or around the ear, probably from a reflex through the seventh or fifth nerve. When these are present, a CT scan or MRI is done to look for a specific cause, such as hydrocephalus. In most cases none is found. Headaches that occur after acoustic neuroma treatment can be quite varied in their nature. Over the course of decades,

the prevalence of headaches, especially after retrosigmoid/suboccipital surgery, has been noted. Centers with high-volume acoustic neuroma practices now take a number of intraoperative measures to avoid causing headaches. When they do occur, symptomatic treatment is given, and usually there is gradual recovery.

ANA's publication *Headache Associated with Acoustic Neuroma Treatment* offers more details about post-treatment headaches.

CONCLUSION

The symptoms and post-treatment outcomes discussed here are not typical of all acoustic neuroma patients, and each case should be individually evaluated by a medical professional.

WHAT IS THE ACOUSTIC NEUROMA ASSOCIATION (ANA)?

Acoustic Neuroma Association was founded in Carlisle, Pennsylvania, in 1981 by a recovered patient, Virginia Fickel Ehr. She found no patient information or patient support available when she had surgery for the removal of an acoustic neuroma in 1977. She resolved that future acoustic neuroma patients should have easy-to-read medical material about their condition, and support and comfort from each other. With the help of her physician, she contacted eight other patients and formed the organization.

The association is incorporated and is a 501(c)(3) non-profit organization. The patient-focused, member organization now serves close to 5,000 members, is governed by an all-patient Board of Directors and is operated by a small staff in metropolitan Atlanta, GA.

ANA membership benefits include receipt of a quarterly newsletter, patient information booklets, access to a network of local support groups, access to a list of acoustic neuroma patients willing to talk about their experience throughout the country, our website Member Section and an invitation to a biennial symposium on acoustic neuroma. Our exclusive website Member Section includes published medical journal articles on acoustic neuroma and all of our patient information booklets and newsletters and many symposium presentations. ANA also maintains an interactive website at www.ANAUSA.org with an ANA Discussion Forum.

ANA is patient-founded, patient-focused and patient-funded. ANA recommends treatment from a medical team with substantial acoustic neuroma experience. Although the association cannot recommend specific doctors, medical centers or medical procedures, guidelines for selecting a qualified medical professional can be found at the ANA website, www.ANAUSA.org. Now available on our website is a listing of medical resources. The physicians and organizations listed have self-reported data to meet criteria established by ANA for having substantial experience in treating acoustic neuromas. The listings should NOT in any way be construed as an endorsement or recommendation by ANA. It is every individual's responsibility to verify the qualifications, education and experience of any healthcare professional.

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