



VESTIBULAR SCHWANNOMA

This fact sheet is intended to give a possible explanation for symptoms you might experience as a result of having vestibular Schwannomas (formerly called acoustic neuroma). This information is not intended to replace medical advice from your specialist. The symptoms discussed here will vary in each person and are not typical of all vestibular Schwannoma patients. Each case should be individually evaluated by a doctor who is familiar with your symptoms. In Nf2 other benign tumours in the brain or on the spine may cause different symptoms.

Understanding the brain and nervous system

The nervous system is a remarkable structure with specific functions localised to different regions. An understanding of the anatomy and function of the brain and its cranial nerves can provide an insight into the symptoms that a vestibular Schwannoma might cause. The information here can also explain the physical changes some patients experience after the removal of a tumour.

A large portion of the brain is composed of the right and left **cerebral hemispheres** that are interconnected through a central structure called the corpus callosum. The right cerebral hemisphere controls the left side of the body and vice versa.

Each cerebral hemisphere is divided into four lobes: **frontal, temporal, parietal, and occipital**. (It is not possible here to discuss all their functions.)

In general terms, much of the frontal lobes are concerned with personality and emotions; the posterior (back) portion of the frontal and parietal areas are related to movement and sensation. The occipital lobes are concerned with visual perception. The temporal lobes are also concerned with many other functions, including three-dimensional and spatial perception, memory, and auditory function. For most people, the ability to speak and understand speech is located in the left cerebral hemisphere (in parts of the frontal, parietal, and temporal regions).

At the base of the cerebral hemispheres, is an area called the **hypothalamus**, connected by a short stalk to the **pituitary gland**. These structures are concerned with hormonal control of the body.

Deep inside the cerebral hemispheres there are central controlling and connecting structures called the **basal ganglia, thalamus, and mid-brain**. The mid-brain merges into the **brainstem** at the back of the base of the skull until it leaves the head, through an opening called the **foramen magnum**, to become part of the spinal cord.

All of the connections from the brain to the rest of the body go through the brain stem. On each side of the brain stem are the right and left **cerebellar hemispheres**. These help control co-ordination (the right hemisphere controls the right side of the body) and the ability to walk.

The brain is bathed in a watery fluid called **cerebrospinal fluid** which is continually being formed and reabsorbed. 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.

The Cranial Nerves

In the middle of the base of the brain, beginning behind the bridge of the nose and extending to the junction of the head and neck, are a paired series of 12 cranial nerves. These cranial nerves carry impulses from the senses to the appropriate brain centre, and instructions from the brain centre to other structures (such as muscles and glands). Historically, these cranial nerves are designated with Roman numerals.

- I. **Olfactory** — carries sensation from the nose to the brain.
- II. **Optic** — carries visual impulses from the retina to the brain.
- III. **Oculomotor** — supplies four of the six muscles that move the eye, and the muscle that elevates the eyelid.
- IV. **Trochlear** — supplies one muscle that moves the eye.
- V. **Trigeminal** — carries sensation from the inside of the mouth, teeth, and front of the tongue, and skin of the face. It also supplies the muscles of the jaw.
- VI. **Abducens** — supplies one muscle that moves the eye.
- VII. **Facial** — supplies the muscles that move the face and the lacrimal and salivary glands and carries taste sensation from the front of the tongue.
- VIII. **Auditory** — consists of two parts: the cochlear nerve which carries hearing and the vestibular nerve which transmits impulses from the semicircular canals which affect balance.
- IX. **Glossopharyngeal** — supplies the parotid gland and carries taste sensation from the back third of the tongue.
- X. **Vagus** — supplies muscles of the vocal cord and those involved with swallowing. It gives movement to smooth muscles and affects the secretory glands in the respiratory and gastrointestinal tracts; is involved with control of the function of the heart.
- XI. **Accessory** — supplies muscles that elevate the shoulder.
- XII. **Hypoglossal** — supplies muscles which move the tongue.

Symptoms

The middle and inner ear structures are located within a bone that forms part of the base of the skull that extends from the ear toward the centre of the head. An opening in this bone, called the internal auditory meatus, leads to the internal auditory canal, which contains the auditory, vestibular and facial nerves. The vestibular Schwannoma starts near this opening and grows into the canal. Pressure can cause it to enlarge and it can also extend into the cranial cavity.

The tumour originates in the Schwann cells that form the sheath around the vestibular part of the eighth cranial nerve. As it grows, it interferes with the function of the vestibular and cochlear nerves. When the tumour becomes larger, it starts to compress the brain stem and fifth cranial nerve, the trigeminal nerve.

A very large tumour may involve cranial nerves IV to X and markedly compress the brain stem and cerebellum. Cranial nerves I, II, III, and XII are usually not involved with the tumour. The eleventh cranial nerve may rarely be involved with large tumours. Functions of the cerebral hemispheres are affected only in the rare circumstances when hydrocephalus (enlargement of the ventricles containing the cerebrospinal fluid) occurs due to obstruction of the normal circulation of the fluid.

Auditory Symptoms

The first symptom from the tumour is usually some disturbance in hearing, such as the inability to hear on the telephone, fullness in the ear, and/or tinnitus. This is probably due to pressure on the cochlear portion of the eighth nerve. The cochlear nerve is more sensitive to pressure than most other cranial nerves.

The ability to hear requires an intact nerve and an adequate blood supply to the inner ear hearing structures. In most patients, the blood supply to the cochlear nerve and the cells receiving the sound in the ear comes from the internal auditory artery. This artery originates inside the head and passes through the internal auditory canal with the nerves, and, therefore, will also be involved with the tumour. Sudden deterioration or fluctuation in hearing may relate to pressure on this artery.

After an operation to remove the tumour the person is usually deaf in that ear because the cochlear nerve and/or internal auditory artery are intertwined with the tumour. It is difficult to save the hearing when the tumour is removed. Occasionally with a small tumour, it is possible to save these structures and preserve hearing. Radiation therapy may have an effect on the cochlear nerve or the internal auditory artery. This effect may be seen over several years following therapy, thus the initial success with hearing preservation may be lost over time.

Unsteadiness and Vertigo

The tumour starts on the vestibular part of the eighth nerve and in some people, unsteadiness or vertigo may be early symptoms. These symptoms are usually mild or intermittent because the nervous system adapts quite well to gradual loss of function in this nerve. In those with larger tumours, pressure on the cerebellum and/or brain stem might cause difficulty with balance.

Some people will experience a period of dizziness and difficulty with balance after removal of the tumour. This can be due to abrupt loss of function in the vestibular nerve fibres that were still intact and had to be divided to remove the tumour. These symptoms usually subside over days to weeks. Where there is a large tumour, the involvement of the cerebellum and brain stem or its blood supply may be the cause of persisting symptoms. Those who undergo radiosurgical treatment may experience fewer balance problems immediately after treatment; however, because this treatment takes some time to have its effect, balance problems may be delayed in emerging.

Facial weakness / dry eye

Because the auditory nerve and the facial nerve are normally next to each other, every vestibular Schwannoma will involve the facial nerve. Facial weakness before treatment is uncommon because the facial nerve is quite resistant to pressure, although, a few people notice intermittent spasm or difficulty blinking the eye.

Postoperative facial weakness is due to the reaction in the nerve after it has been separated from the tumour capsule or occasionally divided totally from the nerve. If there is inability to close the eyelids, a minor procedure might be required to improve this function. A small percentage of those undergoing radiosurgical treatment also experience facial weakness post-treatment.

A dry eye is due to the loss of innervation (nerve supply) through this nerve to the lacrimal gland. Occasionally this portion of the facial nerve "over recovers" and carries excessive impulses so that too many tears are provided, especially when chewing. Facial nerve fibres also serve the secretory glands in the nose, so this function may be affected by either excessive or inadequate secretion.

Facial Numbness

As the tumour becomes larger, there can be pressure on the trigeminal (fifth) cranial nerve causing numbness of the face and/or numbness of the mouth or tongue. Usually this symptom subsides with removal of the tumour, 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.

Poor co-ordination in legs or arms

The control of co-ordination in the limbs is located in the cerebellar hemispheres and adjacent brain stem. In some large tumours, a problem in hand co-ordination might cause difficulty with some daily activities. The legs may also be affected. Often a program of physiotherapy is required to aid recovery in the post-treatment period.

Weakness or loss of sensation in extremities

In some people with large tumours, this problem is due to brain stem compression. When it occurs postoperatively, it is usually due to blocking of the blood supply to the brain stem. In some large tumours the important arteries in the area supply both the tumour and the brain stem, and it might be difficult to separate them.

Double vision

Double vision can be due to a loss of function in either the nerves supplying the eye muscles or the brain stem centres that co-ordinate eye movement. The third cranial nerve lies above the tumour and is not affected. In large tumours, the fourth cranial nerve may be involved.

Difficulty Swallowing

Fortunately this is a very rare symptom both before and after treatment. When it does occur, it is due to involvement of the tenth cranial nerve fibres or blocking of some of the blood supply to the brain stem.

Headaches

Headaches before having treatment are uncommon. When they do occur, they are usually associated with a large tumour. Some people complain of aching in or around the ear, probably from a reflex through the seventh or fifth nerve. When these are present after treatment, a CT scan or MRI is done to look for a specific cause, such as hydrocephalus. In most cases no special cause is found. Symptomatic treatment is given, and usually there is gradual recovery.

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