

Vestibular Schwannoma (VS)

Introduction

A vestibular schwannoma is a rare inner ear tumour. This condition is also known as acoustic neuroma.

This information is designed to help you answer the common questions that are asked by patients after the initial consultation. It is hoped that it will help you understand your diagnosis.

What is VS?

It is a benign tumour, not a cancer, which grows on the balance nerve. This nerve runs from the brain through a narrow channel in the bone of the skull to the inner ear (the internal auditory canal). These tumours originate inside the bony channel and grow into the space where the brain is situated. Once the tumour reaches a certain size, it can start to press on the brain. It does not grow into the brain tissue itself. The hearing nerve (or cochlear nerve) and the facial nerve are also present within this channel, and it is possible for them to be compressed by the tumour, which may cause you some symptoms.

What causes VS?

These tumours are caused by damage to the genetic material inside the lining of the balance nerve. The cause of this genetic damage is unknown. It is not caused by anything you have done and it cannot be caught from someone else. It is not passed on to your children.

This is different in a rare condition called Neurofibromatosis Type 2 (NF2), where people may have multiple benign lesions. Only 5% of vestibular schwannomas are in people who have NF2, and the condition normally arises in teenagers or young adults. If you need any further investigation for this condition, your clinician will discuss this with you at your outpatient appointment.

How common are VS?

Vestibular schwannomas are very rare. There is one new vestibular schwannoma diagnosed each year for every 100,000 people in the population. They can occur at any age but are most common in people in their 50's and 60's. They affect men and women equally.

Patient Information

How fast do VS grow?

Approximately 40% of vestibular schwannomas grow after diagnosis but the growth rate is usually very slow with an average growth rate of 1-2mm a year. They can occasionally grow faster and the pattern of growth is very variable. However, the rate of growth means that it is generally safe to plan treatment ahead of time.

What symptoms do VS cause?

Single-sided hearing loss is usually the most common symptom that people with vestibular schwannomas experience. Some people may lose their hearing altogether but the majority notice a gradual deterioration in their hearing on the affected side.

Tinnitus is also another common symptom, which is a ringing or buzzing sound in the ear. This can be quite troublesome but there are a number of effective therapies that can help. Some patients also experience imbalance or dizziness. Other symptoms include numbness of the face, aching of the bone behind the ear, twitching of the facial muscles and a feeling of fullness in the ear. Weakness of the muscles of the face is unusual but has been reported by some people with vestibular schwannomas. Some people have no symptoms at all, and the vestibular schwannoma is discovered while they are being investigated for something entirely unrelated.

Are vestibular schwannomas dangerous?

Vestibular schwannomas are not cancerous and do not spread to other areas of the body. If they grow into the space where the brain is situated, they can compress the brain. This may cause symptoms such as headaches.

If a vestibular schwannoma is allowed to grow very large, it can potentially be a life-threatening condition as the tumour can press onto parts of the brain that control breathing and heart rate. However, your tumour will be monitored and treated to prevent this from happening.

How can vestibular schwannomas be treated?

There are three main ways of treating vestibular schwannomas. If the tumour is small, then many patients prefer to undergo a period of observation (known as active surveillance, 'watch, wait and rescan', or WWR). This has no potential side effects unlike the other treatment options but you do require periodic scans to make sure that the tumour is not growing. This is a management option because 60% of vestibular schwannomas do not grow and cause only mild or manageable symptoms.

Patient Information

The other treatments are surgery and radiotherapy. If you have the tumour removed, it is usually possible to remove it completely, but as with any surgery there are risks to having an operation, such as facial weakness. Radiotherapy is very successful at stopping the tumour from growing, but it does not remove the tumour and you will require regular post treatment imaging to check tumour size. In the few cases where it does not stop the growth, it makes any surgery needed more difficult. The chances of complications after radiotherapy are generally less than those from surgery although in the long term there is a very small chance that the tumour could become cancerous. The risk of this is around 1% for each decade after treatment. Some people find it very difficult to decide whether surgery or radiotherapy would be best for them so the doctors and nurse practitioners can help you to make the right decision for you.

Watch and wait

Why is my vestibular schwannoma being observed?

You have a small tumour, which is not compressing the brain. This means that the tumour is unlikely to cause any problems in the short term. In this situation it is reasonable to observe the tumour rather than treat it by surgery or radiotherapy. The options for treatment will have been discussed with you and you have decided to undertake a period of observation to monitor your tumour. It will be discussed with you at your appointment how often you need to be scanned.

An MRI scan is the best type of scan to use for monitoring these tumours. However, occasionally a CT scan will be used instead if an MRI scan is not possible, for instance if you have any magnetic metal work inside your body. The MRI scan takes around thirty minutes and you will have an injection in your hand. The scan is painless but it can be quite loud inside the scanner. Once the scan is done, the specialist will look at the scan and write to you with the result.

What happens if my symptoms change?

If there are any changes in your circumstances, your doctor will be very happy to see you again and discuss any issues you may have. Symptoms can change but that does not necessarily mean that the tumour has grown

What happens if my vestibular schwannoma grows?

If the scan shows there is growth of the tumour, an appointment will be arranged with your doctor to discuss the treatment options available to you.

If there is a small amount of growth and the tumour is not touching the brain, then it may be possible to continue observing the tumour. However, if the tumour is starting to compress the brain, then it will probably be recommended to have either radiotherapy or surgery in order to treat the tumour. These forms of treatment will

Patient Information

be fully discussed with you so that you can make an informed decision about which type of treatment you would like.

Surgical removal of vestibular schwannoma

Complete removal of the tumour at surgery is achievable in the majority of cases and less than 5% of tumours not fully excised will start growing again. In patients who have small tumours and socially useful hearing (i.e. can use the telephone), an attempt can be made to preserve the remaining hearing on the affected side.

However, only around one in three of these patients will have any hearing at all in the affected ear after the operation. In patients who have large tumours, or those who have no socially useful hearing, it can be assumed that hearing will be permanently lost on that side after surgery.

Your surgeon will discuss with you whether it is possible to attempt to save some hearing on the affected side. This affects the technical details of the operation and how we reach the area.

A hearing-preservation operation is described as a retro-sigmoid operation.

Most non-hearing preservation operations are carried out via a translabyrinthine approach. This is through the bone behind the ear.

Most centres will have 2 surgeons performing the operation - a neurosurgeon and an ENT skull base surgeon.

Special measures you need to take after the operation: you will be given more detailed information about any special measures you need to take after the operation. You will also be given information about things to watch out for that might be early signs of problems (for example, infection).

Driving

There is no need to inform the DVLA (Swansea) that you have had surgery, unless specifically instructed to do so by a medical team. However, the advice may differ if you have an HGV licence so please let the team know if this is the case for you. The DVLA's advice is that you do not return to driving until 'fully recovered from the surgery.' This varies enormously between patients and depends on your ability to perform an emergency stop, being able to glance in your mirrors and look right and left without feeling nauseous or dizzy. Once you feel able to do these, you are able to return to driving. To begin with, only drive short distances, and gradually increase the distance.

Flying

Patient Information

Travelling by aeroplane should be avoided for at least three months after vestibular schwannoma surgery.

Check-ups and results: Follow-up appointments will be arranged for you on discharge from hospital.

Serious or frequently occurring risks

The risks of any surgical operation:

- **Haemorrhage** - (less than 1%). Although the risk of bleeding is very small, when it occurs in a confined space, for example next to the brainstem, it can result in serious permanent neurological disability. This can include weakness or paralysis, difficulty in breathing or impaired swallowing.
- **Respiratory complications** – chest infections, which can usually be treated with antibiotics and chest exercises with physiotherapists.
- **Blood clots** – there is a risk of deep-vein thrombosis in the legs, which occasionally pass to the lungs (pulmonary embolism).
- **Wound problems** - including wound infection or leakage.
- **Heart** - for example, abnormal rhythm or heart attack.
- **Death** - (less than 1%)

The risks specifically related to the surgical removal of vestibular schwannomas:

- **Facial weakness:** the facial nerve (which is a nerve that supplies the muscles of facial expression) and the acoustic nerve (the nerve of hearing and balance) run very close together. Due to their anatomical position, the facial nerve is attached to the surface of the tumour and is at risk during tumour removal. There is a 5 - 30% risk of facial weakness after this operation depending on the size of the tumour and how tightly it is stuck to the facial nerve.

With small tumours, it is nearly always possible to preserve the facial nerve anatomically (i.e. not seen to damage it 'by eye'), but the facial muscles may be weak for a number of months afterwards due to bruising.

With some large tumours, and even very occasionally with small tumours, it is not possible to spare the facial nerve. If the facial nerve is completely lost, or fails to recover after the operation, there are a number of plastic surgical operations that can be undertaken to restore some function (your doctor can discuss this with you).

You may experience problems with a dry mouth and dry eye after surgery. Your doctor can prescribe you artificial saliva for your mouth. You should use regular eye drops and ointment at night, and may need to tape your eye closed at night to prevent corneal damage.

Patient Information

- **Loss of hearing on the affected side:** most vestibular schwannomas are diagnosed after the patient experiences a loss of hearing, which can be partial or total. Following surgery, the majority of patients will lose their hearing completely in the affected ear. The doctor will review your hearing tests and look at the appearance of the tumour on the scan.

With this information, the doctor can advise you as to whether an attempt can be made to preserve your remaining hearing in that ear. This will be discussed in detail with you before the operation.

- **Tinnitus:** some patients experience tinnitus (for example a ringing noise) in the affected ear. Even when hearing is lost completely after surgery, it is possible that you will still have tinnitus. Even if you had no tinnitus before the operation, it may develop afterwards. However, it is unusual for tinnitus to be dramatically worse after an operation.
- **Cerebral-spinal fluid (CSF) leak:** CSF bathes the brain in fluid. When the tumour is removed, the CSF pathways around the brain are opened. CSF can leak out either through the entry wound or into the ear and then down the nose. The risk of leakage is around 4-10%. The majority of leaks will settle down over a few days if a drainage tube is placed temporarily in the spinal fluid pathways in your back. However, around one in three leaks will require a second small operation to repair them.
- **Infection:** the operation to remove a vestibular schwannoma is long, and the ear can contain micro-organisms that can get inside the head. These can infect the cerebro-spinal fluid and cause either a local wound infection or meningitis.

If there is a leakage of cerebro-spinal fluid after surgery (see above) this can cause infection. The majority of infections can be treated with antibiotics. Very occasionally, there can be serious and longstanding problems from infection inside the head. Your doctor can discuss this with you on request.

- **Problems with balance:** in many cases with vestibular schwannoma, the balance nerve would have been slowly destroyed by the growing tumour. As the tumour grows slowly the brain has been able to compensate for the reduction in information it receives about balance by relying on the other ear. The tumour can only be removed by cutting through the nerve of balance in the affected ear. Therefore, if before the operation there was some function in the nerve, you will feel dizzy and unsteady after the operation until your brain gets used to it. Your balance may be tested before the operation to see how likely this is to happen and your surgeon will discuss with you the likelihood of you being unsteady or dizzy after surgery. In addition, we will need to temporarily displace the cerebellum (the balance part of your brain). There is a very small risk that this part of your brain could be injured during removal of the tumour which might result in permanent unsteadiness.
- **Difficulty swallowing:** in large tumours (generally those more than 3cm), the nerves that control swallowing and supply the vocal cords might be stuck to the tumour. If this is the case, these nerves might not function after the tumour has been removed. This can result in difficulty swallowing and

Patient Information

hoarseness of the voice for a number of months after surgery. Very occasionally, problems of this kind are permanent.

- **Stroke/major neurological impairment:** there is a very small (around 1%) risk of major neurological impairment following surgery. The greatest risk is if there is any bleeding into the cerebellum or around the brainstem after surgery.

A further small risk is of bleeding from the important blood vessels supplying the brainstem and cerebellum, which can become quite stuck to the tumour, particularly if it is of a large size.

- **Headache and neck pain:** as we need to gain access to the bone behind the ear during surgery, we need to disturb some of the neck muscles in this area. This will cause some neck pain and stiffness. It is common to experience a headache after operations on the head, particularly for the first few days. This will be controlled with painkillers.
- **Numbness of the face:** with large vestibular schwannomas, the trigeminal nerve (the nerve which is responsible for feeling/sensation on the face) can also become stuck to the tumour. If this nerve is damaged during the operation, you can experience numbness on that side of the face. Our greatest concern here is if the surface of the eye becomes numb. If you are not able to feel the surface of the eye, you would not be able to tell if grit or dirt gets into the eye, and this can lead to damage and later, infection.

If you have facial numbness, particularly in combination with facial weakness, you will need to take particular care to ensure that your eye is protected. If this is necessary, you will be taught how to do this. This will involve regular use of eye drops and ointments, and taping your eye closed at night.

Radiotherapy treatment to vestibular schwannoma

What is radiotherapy?

Radiotherapy is the use of high energy x-rays to treat tumours. It can be delivered as a single or small number of treatments and this is normally called stereotactic radiosurgery (Gamma knife or Cyber knife). Alternatively it can be divided into 30 treatments (fractions) which are given daily (Monday to Friday) for several weeks; this is called fractionated stereotactic radiotherapy. To decide which form of radiotherapy is best for you involves an outpatient appointment to see you, look at your MRI scans and to assess the size of your tumour, its location and also to consider your own personal preference.

What is the aim of radiotherapy?

Radiotherapy aims to stop your tumour growing any bigger and for some patients will actually make it shrink, although this may take years following treatment. Radiotherapy will not remove your tumour.

Patient Information

What are the side effects of having radiotherapy?

Radiotherapy does not make you radioactive and there is no need to take any special precautions for the safety of others. The radiotherapy will not make you ill and you will be well enough to travel.

Many patients carry on their normal daily activities before and after the daily treatment session. However sometimes it can cause you to have some side effects.

Fractionated stereotactic radiotherapy – side effects (acute)

Side effects that occur during the course of treatment and in the first couple of months following treatment are called **acute side effects**. Not every patient will experience all of the side effects and if they do happen, the team looking after you will be able to provide help and advice.

Possible acute side effects include:

- **Skin erythema.** This means the skin just in the area being treated may become pink and a bit dry. This is normally very mild, not permanent and will settle about three weeks after treatment finishes.
- **Hair loss.** This is just in the treated area (normally a very small area) and for the majority of patients the hair will grow back normally three to four months after treatment has finished.
- **Increase in tinnitus.** Sometimes the noises you hear in your ear may appear to grow louder or change in tone.
- **Irritation of the treated outer ear canal.** You may experience some redness and irritation of the ear canal on the side being treated. This may mean your hearing seems worse and you feel like your ear is blocked. This should settle a few weeks after treatment finishes.
- **Worsening balance.** You may find that your balance is worse during radiotherapy. This is normally due to some swelling caused by the treatment and should settle a few weeks after treatment.
- **Fatigue and lethargy.** Many patients feel tired and have a lack of energy during and for a few weeks after radiotherapy.
- **Nausea.** Very occasionally patients find the radiotherapy causes them to feel sick and lose their appetite. Tablets can be prescribed if this happens to you. It is very rare to actually be sick.

Stereotactic radiosurgery – side effects (acute)

The acute side effects following SRS are slightly different to FSRT. They may occur in the days and weeks following the single treatment and include:

- fatigue and lethargy
- nausea
- headaches
- soreness at the pin sites if a fixed frame was attached for treatment

Patient Information

Intermediate side effects – following both types of radiotherapy

There are also a few side effects that may happen three to six months after finishing radiotherapy as a result of some swelling of the tumour. These normally settle completely although they may mean having a short course of medication. These are called **intermediate side effects** and include:

- facial weakness, facial numbness, facial pain and decreased hearing
- headaches and vertigo

Late side effects - following all forms of radiotherapy

The last group of side effects are **late side effects** and these occur years after the radiotherapy is given.

Most of these are very rare but include:

- Sudden or gradual decrease in hearing on the treated side.
While for many patients having radiotherapy may initially preserve hearing over time this may deteriorate and a few patients have a sudden loss of hearing at some point following radiotherapy.
- Low risk of long term facial nerve and trigeminal nerve problems. This may include facial pain, facial numbness and facial weakness.
- Persistent feeling of vertigo, dizziness and nausea requiring medication.
- Unpredictable alteration in tinnitus. This may mean a change in volume and pitch.
- Very low risk of causing hydrocephalus. This is when the tumour swells following radiotherapy and can block the flow of fluid around the brain and central nervous system. This normally causes severe headaches and vomiting and usually requires a surgical procedure to correct it.
- Very low risk of causing a second tumour to develop in the area that is treated – this may be another benign lesion but could also be cancerous.
- 2-5% risk of continued tumour growth, which may lead to needing surgery, with increased risks.

If I have radiotherapy do I need follow-up after treatment?

Because the radiotherapy will not remove the tumour, it is recommended that a follow-up schedule of MRI scans are arranged in order to check that the vestibular schwannoma does not show evidence of further growth. It is common in the 1st couple of years after radiotherapy to actually see a slight increase in the size of the lesion as a response to treatment. Normally this will have settled and we would expect the tumour size to be stable from year 3 onwards.

Patient Information

Support groups

BANA

The British Acoustic Neuroma Association (BANA) was formed in 1992. BANA is organised and administered by people affected by acoustic neuroma, is a registered charity and exists for mutual support, information exchange and listening.

British Acoustic Neuroma Association
Tapton Park Innovation Centre
Brimington Road
Chesterfield
Derbyshire
S41 0TZ
United Kingdom

Tel: [01246 550011](tel:01246550011)

E-mail: admin@bana-uk.com

Website: <http://www.bana-uk.com/>

Action on Hearing Loss

Action on Hearing Loss (formerly the RNID) aim to make day-to-day life better for people who are deaf or who are hard of hearing. Their website includes lots of useful information on communication equipment, your rights and help with benefits and services.

Website: <http://www.actiononhearingloss.org.uk/>

Information line telephone 0808 808 0123

Information line textphone 0808 808 9000

Email information@hearingloss.org.uk

Brain and Spine Foundation

The Brain and Spine Foundation provide support and information on all aspects of neurological conditions. They aim to reduce uncertainty and anxiety by providing clear and accurate information on topics such as dizziness and balance, migraine and brain scans. This can be found at their website.

Website: www.brainandspine.org.uk

Helpline: 0808 808 1000