Information for patients, family and carers: Large Acoustic Neuroma (Vestibular Schwannoma)

Acoustic Neuroma/Vestibular Schwannoma, what is it?

Your symptoms and imaging findings are consistent with an acoustic neuroma, also known more correctly as a vestibular schwannoma. This is a benign tumour, usually slow growing, that has not come from somewhere else in the body, and that will also not spread anywhere else. It will most likely have been present for a number of years. Such tumours arise from ‘Schwann cells’ in the vestibular nerve, the nerve to balance, as it runs from your brain into the bone of your skull. The hearing is affected because the tumour stretches the adjacent cochlear nerve, the nerve to hearing.

For most patients with an acoustic neuroma (vestibular schwannoma), the cause is unknown. Very rarely, these tumours occur as part of a hereditary condition known as Neurofibromatosis Type 2. This is usually obvious at the time of initial diagnosis with either the patient’s brain scan showing more tumours than just a single acoustic neuroma (vestibular schwannoma) or other family members having similar problems.

I am aware of the association of hearing loss with an acoustic neuroma. What about the other symptoms that I am experiencing?

As an acoustic neuroma grows to a large size, it tends to put pressure on surrounding structures over and above the nerve to hearing.

It can stretch other nerves nearby. These adjacent nerves are collectively known as the cranial nerves. For example, you may notice numbness of the face or have developed facial pain on the same side of your tumour due to stretching of the trigeminal nerve over the top of the tumour. Less frequently, there may be facial muscle weakness due to stretch of the facial nerve, double vision due to stretch of the abducent nerve (which contributes to control of eye movements), or hoarseness and swallow difficulty due to stretch of the lower cranial nerves (also known as the glossopharyngeal and vagus nerves which are related to the bottom part of the tumour.

You may have noticed problems with coordination of your arm and leg on the side of your tumour reflecting compression of an adjacent part of the brain to do with control of movement known as the cerebellum.

You may have noticed increasing headaches, blurred vision, vomiting, and a general unsteadiness due to increased brain fluid pressure or hydrocephalus (‘water on the brain’). This usually occurs with severe compression of the
brainstem, that part of the brain adjacent to your tumour that connects your spinal cord below with your main brain above.

How is my tumour diagnosed?

Your tumour will have been picked up on a brain scan, either CT (computerised tomography) scan or MRI (magnetic resonance imaging). The scan usually includes injection of a contrast material (‘x-ray dye’) into a vein to allow us to see the tumour better.

An MRI scan is usually needed to allow us to look at the tumour in great detail. Generally, we have a good idea from an MRI scan that a particular tumour is an acoustic neuroma (vestibular schwannoma). An acoustic neuroma (vestibular schwannoma) does have characteristic features on a scan.

We measure the size of your tumour from the scan. A tumour is considered large if it is 3cm or more in diameter. The scan shows the extent to which your tumour is putting pressure on surrounding structures as well as if there is any hydrocephalus.

As and when operation is planned, a CT scan specifically looking at your skull bony anatomy is done to help us decide how we will access your tumour surgically.

However, as and when such a tumour might be operated upon, we do send tumour to the pathologist at the time of surgery to confirm the exact type of tumour. Occasionally, we find out that a patient who we thought had an acoustic neuroma (vestibular schwannoma) has a different tumour type, usually benign, such as a meningioma.

How can my tumour be treated?

In general, for a large acoustic neuroma (vestibular schwannoma), we recommend surgery in the first instance because of the potential life threatening nature of your condition otherwise.

If you have significant general health problems that might make an anaesthetic particularly risky, we may advise that your treatment be stereotactictic radiosurgery (also known as gamma knife), a focussed form of radiation therapy.

You may have a smaller initial operation before your tumour is removed (or if stereotactictic radiosurgery is planned) to manage any brain fluid pressure or hydrocephalus (‘water on the brain’) problems. Such an initial smaller operation would be insertion of a ‘VP-shunt’ or optic nerve fenestration. Your surgeon will explain these operations in further detail if this is an issue for you.
What does an operation involve?

The aim of an operation is to remove the greater part (more than 95%) of your tumour and to decompress the surrounding important nerve and brain structures.

The operation takes around ten to twelve hours. We either drill through the back part of your ear bone (a ‘trans-labyrinthine approach’) or make a trapdoor at towards the back of your skull (a 'retromastoid approach'), with the choice of approach dependent on the appearances of your tumour and your skull bony anatomy. The operation is done jointly by Mr Carroll, Consultant Neurosurgeon, and Mr Yardley, Consultant ENT surgeon.

You will spend a day or two in the neuroscience intensive care or high dependency unit after your surgery (see patient information leaflet on Neuro-Intensive Care Unit). You would be in hospital about one week after the surgery. We would expect you to be independent and self-caring on discharge and be off work subsequently for approximately six weeks.

Any remaining hearing on the side of the tumour is always lost.

The risks of surgery include: clot in the leg/clot in the lung ('DVT/PE') 1:100; infection 1:100; stroke/blood clot (with the possibility of permanent paralysis and also having a very small risk to life) 1:100; brain fluid leakage down nose/hydrocephalus ('water on the brain')/meningitis 1:100; chest infection 1:100, facial nerve palsy 1:100, and other cranial nerve problems such as permanent facial numbness, double vision, swallow difficulty, or voice change 1:100. Because the operation takes a longtime and you are kept in the one position for the duration of the surgery, you can sometimes have pressure areas manifested by skin redness, discomfort, tingling, or occasionally blistering, even with the precautions that are taken such as use of padding and a special operating table mattress. In general, there are a whole range of measures that the surgical team take to stop problems happening and a whole range of measures that are taken to reduce the impact when problems do happen.

After surgery we will carry out a further scan as an outpatient once you have recovered, usually an MRI around the three month mark as a baseline for any future interval scans.

Is there a risk to my facial nerve with surgery?

We take the preservation of facial nerve function very seriously. The facial nerve controls the facial muscles on one side of the face. It is not just about cosmetic appearance, which of course is important. The facial nerve controls your eyelid and tears to your eye, aswell as contributing to eating, speaking, and taste.
Historically, when surgeons completely removed a large acoustic neuroma (vestibular schwannoma), there was facial muscle paralysis in about one third of patients.

The modern practice is not to completely remove your tumour but to leave a very small remnant of tumour against the facial nerve to maximise preservation of its function. With this approach, the risk of significant permanent injury to the facial nerve is 1:100.

This tumour remnant is then followed subsequently by periodic brain scans. In many circumstances this remnant does not grow or can even shrink. Depending on the size of the tumour remnant on the baseline post-operative scan or if the tumour remnant is shown to be growing on subsequent scans, your surgeon will recommend for this small tumour remnant to be treated with stereotactic radiosurgery.

A device for identifying the facial nerve, known as a facial nerve stimulator (with facial muscle electrodes), is always used during an operation to identify the facial nerve and prevent it from being injured.

**What is meant by ‘stereotactic radiosurgery’?**

Stereotactic radiosurgery involves the fixation of a stereotactic metal frame to your skull usually under local anaesthetic, repeating an MRI scan, and then treating the tumour using highly focused beams of gamma radiation.

Radiosurgery aims to stop the growth of any small post-operative tumour remnant, about a 90% chance, and occasionally does result in some tumour shrinkage. Radiosurgery does not get rid of the tumour and does not generally result in improvement in any symptoms that have been caused by your tumour.

Radiosurgery may also be given if you have a large acoustic neuroma (vestibular schwannoma) but are not fit for an operation to remove your tumour but has a significantly reduced effectiveness in treating large tumours as compared to smaller tumours.

The first scan after a radiosurgery treatment is performed at one year following the treatment.

There is less than a 5% chance of problems, usually temporary, with 1-2% being permanent, including facial muscle weakness (a facial palsy).

There is a possible very small risk of long term issues with radiation exposure but in reality this is likely only to be a consideration in a young person.

Stereotactic radiosurgery is generally a single treatment planned and delivered all in one day with a one to two night hospital stay. This is carried
out at the National Centre for Stereotactic Radiosurgery in Sheffield (see www.gammaknife.org.uk or patient information leaflet on Radiosurgery).

**Will I have further scans after my operation?**

Following your surgery and your initial post-operative scan at three months post-surgery, you will then undergo a scan each year initially to see if your small tumour remnant has increased in size. If there is an increase in size demonstrated, you will then undergo stereotactic radiosurgery.

The usual type of scan is an MRI (magnetic resonance imaging), which provides the best detail about your tumour and also avoids exposure to X-rays. A CT (computerised tomography) scan is only performed if you have a pacemaker preventing you from having an MRI or in the work-up for an operation. A CT scan uses X-rays and provides the best detail concerning bone structures.

You should always keep your scan appointments even when you have not had any problems for many years. Note please contact your consultant neurosurgeon’s secretary if you have difficulties with making a scan appointment or your scan appointment is earlier than you anticipated or you do not receive an appointment for a scan when your scan date is due.

It may also be such that you do not need to come to a clinic appointment if your scan is unchanged as compared to previous and that the result of your scan can be communicated to in writing and arrangements can then be made for the subsequent scan. You should enquire with your consultant neurosurgeon or his secretary about the specific arrangements for communication of results and scan follow-up if you feel an appointment would not otherwise be required, in particular if you live at a distance.

**Can my hearing be preserved or improved with treatment?**

You will lose any hearing on the side of your tumour with surgery to remove your tumour. The majority of patients diagnosed with a large acoustic neuroma (vestibular schwannoma) already have severe hearing loss and therefore loss of any functional hearing is not an issue. In addition, treating a large acoustic neuroma (vestibular schwannoma) cannot bring back any hearing previously lost.

**What impact can complete hearing loss on the side of my tumour have on my life?**

Most people with hearing loss in one ear and normal hearing on the non-tumour side are able to live a normal life. Some people report difficulty in blocking out loud background noise when engaging in conversation. Some people have difficulties with ‘directional hearing’, i.e., knowing exactly where a
sound is coming from. If directional hearing is important in a person’s life, e.g., a teacher in a class room or roadside worker, a bone anchored hearing aid (‘BAHA’) may be of benefit.

Can my tinnitus be helped with either stereotactic surgery or open surgery?

Tinnitus (noise or buzzing in the ear) as a result of your tumour will not be helped by either treatment. Tinnitus tends to become less troublesome over time. There are also simple measures that may help the tinnitus have a less impact on your life, e.g., having a radio on in the background if in a quiet environment.

What should my expectations be over the long term?

We would expect that the majority (greater than 95%) of people with a large acoustic neuroma (vestibular schwannoma) to be able to have a normal life in terms of quality of life including employment and relationships and also to not have their lives foreshortened. You will require interval imaging over an extended period beyond ten years after your surgery with longer periods of time between each scan. You may require an additional treatment, usually stereotactic radiosurgery, rarely a second operation.

Is there anybody I can talk with to get further advice and information?

You will be assigned a skull base ‘key worker’ who can provide further advice and information. Your assigned key worker is usually either your consultant neurosurgeon and/or skull base specialist nurse. They should provide you with a means of contacting them. They will also provide you with copies of correspondence such as your clinic letters for your own records and additional information material such as further patient leaflets referred to within the text above.

You may wish to discuss your diagnosis with your GP.

The hospital also provides a chaplaincy service for different faiths or indeed if you do not belong to any faith group. Note that if you are an inpatient the chaplains will not automatically visit you, even if you belong to a particular faith group. If you would like to see a chaplain, please ask a nurse, relative or friend to leave a message on 0114 271 4999.

In addition, there are some local, national and international patient information groups/charities who can offer further advice and information including:

- British Acoustic Neuroma Association
  Website: www.bana-uk.com
  Tel: 0800 652 3143
This information sheet is to be used only in the context of attendance at or admission to the Department of Neurosurgery, Royal Hallamshire Hospital, Sheffield Teaching Hospitals NHS Foundation Trust. No responsibility is held for the advice provided by external support groups listed or the information content provided on their websites. If in doubt, ask your doctor.

Compiled by Mr Thomas Carroll, consultant neurosurgeon, and Mr Mark Yardley, consultant ENT surgeon, 21st September 2012