

Acoustic neuroma

This information sheet has been written to help answer some of the questions you may have about acoustic neuroma and its treatment. If there is anything you do not understand, please discuss it with a nurse or doctor involved in your care.

What is an acoustic neuroma?

An acoustic neuroma is a benign brain tumour that develops from the lining of the auditory nerve, (sometimes called acoustic nerve, vestibular nerve or VIII nerve) which is responsible for hearing. It originates from Schwann cells which line the nerve and gives it the name Schwannoma. It is usually a slow growing tumour and does not spread from the original site to other parts of the body.

Who gets an acoustic neuroma?

Acoustic neuromas are most frequently found in middle-aged adults, although they can occur at any age from 10 onwards. Bilateral acoustic neuromas (affecting both acoustic nerves) are a defining feature of a type 2 neurofibromatosis which may be inherited.

What causes acoustic neuroma?

Apart from the link with neurofibromatosis the cause is unknown. Research is being carried out into possible causes.

What are the signs and symptoms of acoustic neuroma?

Acoustic neuroma is a slow growing tumour and symptoms often develop gradually over several years. The commonest symptom is hearing loss in the affected ear. A buzzing or ringing noise in the ear (tinnitus) may accompany this. Dizziness is also fairly common. Occasionally the tumour may be an incidental finding on a scan performed for another reason.

There are other possible symptoms due to pressure on the facial nerve (VII nerve) and the trigeminal nerve (V nerve) that sit next to the auditory nerve (VIII nerve).

The trigeminal nerve (V nerve) controls sensation on the side of the face and the chewing muscles. This means that damage may lead to altered sensation on the affected side of the face. This may be in the form of numbness, increased sensation or occasionally pain and may also affect chewing.

The facial nerve (VII nerve) controls the facial muscles on the same side of the face. Damage to the nerve causes facial weakness with drooping of the mouth and eyelid and difficulty blinking.

If a nerve has been permanently damaged before treatment starts, it is unlikely that treatment can repair the damage. For example this means that deafness cannot be reversed. However, some damage may be due to pressure on a nerve and when this is relieved, some recovery may occur.

Larger tumours can press on the brainstem but may only rarely lead to problems with walking and balance, and very occasionally to headaches.

People who develop acoustic neuroma related to neurofibromatosis, with tumours on both auditory nerves, may have problems on both sides although only one side may be obvious.

What tests and investigations will I need?

Acoustic neuromas are best seen on an MRI scan (magnetic resonance imaging) as small tumours on the auditory nerve.

A hearing test and sometimes a test of balance measure how much the nerve is affected.

What are the treatment options for acoustic neuroma?

The options are observation without specific treatment, surgical removal, or radiotherapy. Radiation is commonly given in the form of stereotactic radiotherapy either as fractionated stereotactic radiotherapy or as single fraction radiosurgery.

Your doctor will discuss with you and advise you on the best treatment option for you, as an individual, taking into consideration your own preferences. It is important that you feel happy with any decisions that are made.

Observation

If the tumour is small and not causing problems, immediate treatment may not be necessary and a policy of observation may be adopted. This will mean that you will be seen in the clinic and have regular six to twelve monthly scans and hearing tests. This will enable the doctor to assess the rate of growth of the tumour and decide if treatment with surgery or radiotherapy is advisable. Some tumours grow very slowly and may never need treatment.

What are the benefits of observation?

Observation has the advantage of saving treatment until it is necessary and therefore reduces exposure to potential side effects of treatment. In some people it may avoid treatment altogether.

What are the disadvantages of observation?

Some people may feel anxious about not being treated immediately and find the process of observation difficult to come to terms with. Observation requires regular hospital attendance for scans and hearing tests. In most cases, the risks involved with the observation policy are minimal as these tumours are usually very slow growing. Any increase in tumour size is likely to be detected early. However, there is a possibility of gradual hearing loss and larger tumours may cause symptoms as described in the section on signs and symptoms.

Surgery

Surgery has been the standard treatment of acoustic neuroma for many years. The type of surgery done depends on the size and exact position of the tumour. All operations require a general anaesthetic.

There are two main types of operation to remove acoustic neuroma and the opinion varies about which is the best. Both involve a cut behind the ear. The tumour can be reached through the earbone by a translabyrinthine approach. This results in loss of hearing. The tumour can also be reached through the back part of the skull by a retromastoid/retrosigmoid/suboccipital approach. This allows access to the tumour by holding back the brain and it offers the possibility of preserving hearing (depending on the size of the tumour).

Rarely, the neuroma can be removed through a middle cranial fossa approach above the ear bone. While hearing can be preserved, this technique is associated with a small risk of developing epilepsy.

What are the benefits of surgery?

It is possible that the acoustic neuroma can be completely removed and this is particularly true for small tumours, so that no further treatment is required. With modern microsurgical techniques, there is a chance of hearing preservation and less chance of other nerves being damaged compared with older techniques.

What are the risks involved with surgery?

The risks associated with surgery are dependent on the surgeon who performs this specialised surgery and how experienced he/she is in this type of surgery. However, even in the most experienced hands, it is not possible to eliminate all risk. Before undergoing surgery you should discuss the risks with the surgeon who is due to perform the operation.

Surgery for acoustic neuroma may cause temporary or permanent damage to the facial and rarely the trigeminal nerve and this may lead to symptoms previously described (under signs and symptoms of acoustic neuroma).

Following any type of open surgery, there is a risk in the immediate post operative period of infection (wound infection or meningitis), bleeding and external leakage of the cerebro-spinal fluid (CSF leak). All surgery under anaesthetic carries a very small risk of death.

Radiotherapy

Radiotherapy is increasingly used as a treatment for acoustic neuroma. It involves the use of X-rays, targeted onto tumours. It works by stopping cells from dividing and therefore stopping tumours growing. There are two ways of carrying out radiotherapy: conventional radiotherapy and stereotactic radiotherapy. The currently preferred treatment for acoustic neuroma is stereotactic radiotherapy.

More detailed information on stereotactic radiotherapy is on page 4.

Stereotactic radiotherapy for acoustic neuroma

What is stereotactic radiotherapy?

Stereotactic radiotherapy is a high precision treatment, which focuses radiation on the tumour and gives as little radiation as possible to normal brain. You will need to wear a specially fitted head frame, which keeps your head in the same position for each treatment. This means that the area to be treated can be targeted very accurately and radiation of nearby normal brain is minimised.

Radiation can be given as a single high dose in one treatment session and this is described as *stereotactic radiosurgery*. It can also be given in a number of smaller doses and this is called *fractionated stereotactic radiotherapy*.

Stereotactic radiosurgery

Stereotactic radiosurgery is either delivered by a machine called a linear accelerator, sometimes called X knife, or by a machine with cobalt as the source of radiation called gamma knife. Radiosurgery is only appropriate for small acoustic neuromas.

Fractionated stereotactic radiotherapy

Fractionated stereotactic radiotherapy is given with a linear accelerator and usually involves receiving 25 treatments, although sometimes only six treatments are given. A number of radiation beams (usually four to six) are aimed at the tumour at each treatment session.

Each radiation beam is shaped to conform precisely to the shape of the tumour (stereotactic conformal radiotherapy). This is the appropriate technique for treating tumours that are irregular in shape and close to critical structures to be avoided. Fractionated stereotactic conformal radiotherapy has been in routine use at the Royal Marsden Hospital since 1992 for many types of tumour, and since 1995 for acoustic neuromas.

How effective is stereotactic radiotherapy?

The aim of radiotherapy is to stop the tumour from growing. The success of radiotherapy treatment is described in terms of tumour control. A tumour is controlled if it remains the same or smaller size, and does not need further treatment. Following stereotactic radiotherapy for acoustic neuroma, tumour control is in the region of 90% (90 people in 100) at 10 years. Similar tumour control rates have been reported for the two techniques of stereotactic radiotherapy.

There is little hard evidence to show that the rate of decline in hearing is altered by radiotherapy treatment. Decline in hearing may continue regardless of stereotactic radiotherapy.

What are the side effects and risks of stereotactic radiotherapy?

Any radiation treatment has the potential to cause side effects. These may occur during or shortly after treatment (acute side effects) or years later (late side effects). Unlike surgery, where damage is immediate, it may take months or years for the damage to cause symptoms.

Acute side effects are usually temporary and resolve completely. They include tiredness, small patch of hair loss and skin reactions. Large single doses of radiation as used in stereotactic radiosurgery may cause temporary nausea and vomiting. You can read more about this in our leaflet on radiotherapy to the brain.

Following stereotactic radiotherapy, the acoustic neuroma may swell a little before it starts to shrink in size. The swelling may very occasionally press on local nerves and the nearby brain (brain stem). Pressure on the brain stem may block the flow of fluid from the brain. This is described as hydrocephalus and causes headache, nausea and vomiting. This complication is only seen in people with a larger acoustic neuroma and is uncommon. If it occurs, it may need a simple operation to temporarily divert the flow of cerebro-spinal fluid, usually by a shunt.

Late side effects are due to damage to the nerves and part of brain surrounding the acoustic neuroma and may be permanent. Damage to the facial and trigeminal nerves causes symptoms described in the section on signs and symptoms (see page 1).

The auditory nerve (VIII) nerve is also at risk of damage leading to deafness, although sudden deafness is uncommon following fractionated stereotactic radiotherapy.

The techniques of stereotactic radiosurgery and fractionated stereotactic radiotherapy differ in their risk of side effects and the details as reported in the literature are shown in the tables on pages 8 and 9.

There is a concern that radiation may cause another tumour in the brain, but so far there are no reports of radiation induced brain tumours after treatment for acoustic neuroma. The risk of second brain tumour in pituitary adenoma, another benign tumour situated in the centre of the brain, also treated with stereotactic radiotherapy is 2% at 20 years. This is likely to be an over estimate for acoustic neuroma, which is generally a smaller tumour than pituitary adenoma, so the irradiated area is less and we estimate the risk in the region of 1% at 20 years.

Side effects of stereotactic radiosurgery

Radiosurgery has the advantage of being a single treatment, but is more likely than fractionated stereotactic radiotherapy to cause acute side effects of sickness and hydrocephalus. Steroids are usually given to try to reduce this risk.

Large single doses of radiation, as used in stereotactic radiosurgery, are theoretically more likely to damage normal brain and nerves than fractionated radiotherapy. The reported incidence of late side effects following radiosurgery is higher, particularly when a high dose of radiation is given to large tumours. The risk is low with small tumours.

Radiosurgery is an evolving field and current doses of radiation are lower than those previously used with lower reported risk of side effects. However time is needed to see if

lower doses are as effective as higher doses in controlling the growth of an acoustic neuroma.

Side effects of fractionated stereotactic radiotherapy

This technique causes occasional acute effects such as tiredness and temporary hair loss where the small radiation beams enter the head. The details are as mentioned in our factsheet on radiotherapy to the brain. Nausea and vomiting are uncommon. Hydrocephalus is a rare complication of treatment of acoustic neuroma.

In our experience of 29 patients treated up to the end of 2002 with fractionated stereotactic radiotherapy at the Royal Marsden, the incidence of permanent damage to the facial and trigeminal nerves was less than 5% with a tumour control of 100% in the limited follow-up.

The reported incidence of late side effects in the form of permanent damage to the trigeminal nerve and facial nerve is lower compared with radiosurgery (see tables on pages 8 and 9).

Fractionated stereotactic radiotherapy does have the inconvenience of more hospital visits.

How do I decide and what do I do now?

The advice to have treatment is based on the balance of risks and benefits and your doctor would be happy to discuss this with you in further detail.

There may not be a right answer as to which treatment is best for you and the final decision may be left to you. It is important, therefore, that you ask everyone involved in your treatment all the questions important to you, so that you feel equipped to make the best decision for you. You may need to see an expert in each area to come to a conclusion.

More information on the late side effects are summarised in tables on pages 8 and 9.

Useful addresses:

British Acoustic Neuroma Association

Oak House, Ransom Wood Business Park
Southwell Road West
Mansfield,
Nottinghamshire NG21 0HJ
Tel: 01623 632143 or Freephone 0800 652 3143
Fax: 01623 635313
Email: bana@ukan.freeseve.co.uk
Web site: www.ukan.co.uk/bana

Neurofibromatosis Association

Quayside House, City 8, High Street
Kingston-upon-Thames
Surrey KT1 1HL
Tel: (020) 8439 1234
Web site: www.nfauk.org

Contact details

Consultant:

Specialist Registrar:

Senior House Officer:

Clinical Nurse Specialist: Douglas Guerrero

Contact Numbers

Dr Saran's clinical secretary:
Royal Marsden Hospital, Sutton (Surrey) 020 8661 3826

Professor Brada's clinical secretaries:
Royal Marsden Hospital, Fulham Road (London) 020 7808 2271
Royal Marsden Hospital, Sutton (Surrey) 020 8661 3270

Hospital Switchboard:
Royal Marsden Hospital, Fulham Road (London) 020 7352 8171
Royal Marsden Hospital, Sutton (Surrey) 020 8642 6011

If you need to contact the medical team after 5pm or at weekends, please telephone the hospital switchboard and ask for the on-call doctor for the consultant who is looking after you.

Stereotactic radiotherapy for acoustic neuroma – late side effects

To try and help you compare stereotactic radiosurgery and fractionated stereotactic radiotherapy, we have prepared tables summarising the main late side effects that have been reported in the medical literature. It is important that you realise that these results were taken from individual studies of one type of treatment and not studies comparing different treatments.

Complications of radiosurgery for acoustic neuroma as reported in the medical literature

Author and Institution	Preservation of useful hearing (%)	Permanent Facial nerve damage (%)	Permanent Trigeminal nerve damage (%)
Yamamoto and Noren, 1990	32	9	9
Mendenhall et al., 1994	-	16	18
Miller et al., 1999	39	38* 8**	29* 15**
Ito et al., 2000	-	29# 9.5##	44# 15.5##
Prasad et al., 2000	58	1.5	1.7
Rowe et al., 2003	75	1 (4.5)	1.5 (5)
Flickinger et al., 2004 ****	70	0	4.5
Iwai et al., 2003*****	59	0	0

*16-20Gy **12-16Gy

*** Average dose 13Gy

**** 12-13Gy

***** 12 Gy

>16Gy

< 16Gy

() figures shown in brackets are percentages of transient symptoms

Complications of fractionated stereotactic radiotherapy for acoustic neuroma as reported in the medical literature

Author and Institution	Preservation of useful hearing (%)	Permanent Facial nerve damage (%)	Permanent Trigeminal nerve damage (%)
Maire et al., 1995	60	0	0
Varlotto et al., 1996	100	0	8
Kagei et al., 1999	78	0 (8)	0 (16)
Fuss et al., 2000	85	0	5
Andrews et al., 2001	81	2	5

NB. Maire used conventional technique all others were stereotactic
() figures shown in brackets are percentages of transient symptoms

Please note these tables contain only a selection of the available published data and can serve as a guideline towards complication rates but cannot be directly applied to other treatment centres as the doses given and techniques used will vary. In addition, certain individuals may have a higher or lower risk of complications due to the size and position of the tumour.

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