



NF2 Patient Fact Sheet about Radiosurgery and other Radiation

Treatments

Introduction

This fact sheet attempts to explain terms that may be encountered in researching radiation treatments for NF2. It covers both vestibular schwannomas (VS, also called acoustic neuromas) and meningiomas as the commonest tumours in NF2 that are referred for treatment. It addresses commonly raised concerns about radiation treatments in the context of NF2.

Of note most of the experience with radiation treatment in NF2 is the gamma knife experience in Sheffield and reflecting this, the Sheffield service is integrated with the nationally organised NF2 service based in Manchester, London, Oxford and Cambridge. We have therefore included what a gamma knife treatment in Sheffield involves, and more details of this can be found at www.gammaknife.org although unlike this fact sheet the material on our website is not specific for NF2.

Terms and definitions

Stereotactic radiosurgery and **stereotactic radiotherapy** are types of radiotherapy treatment used for treating VS and meningiomas with the aim of avoiding surgery. The term "**Stereotaxy**" allows spacial localization of the lesion being treated either by means of a frame fixed to the patients head or a frameless localising system. The patient is then scanned and this is used to generate a map with a grid system that precisely locates the tumour. This map is used to target the radiation treatment which is focused on the tumour, so sparing the surrounding tissues.

The radiation can be delivered as a single high dose treatment, which is called **stereotactic radiosurgery**; alternatively the treatment can be fractionated that is given as a course of multiple lower dose treatments – called **stereotactic radiotherapy**. The techniques differ in a number of ways.

- A stereotactic radiosurgery treatment is completed in a single day and relies on the frame and precise targeting of the tumour to avoid damage to surrounding structures.

- A stereotactic radiotherapy treatment is delivered over several days or even weeks. Either a re-locatable frame or mask is used or a frameless system employed. Whilst this will increase positional errors, the fractionation, that is the use of multiple low dose treatments, may allow for recovery of the tissues between the individual doses being given.

Importantly, single dose radiosurgery and fractionated radiotherapy may have different biological effects on the tumour being treated. This can be likened to comparing a single hard hit with multiple light blows – the total energy imparted may be the same, but the response of the tissue may be very different. Hence 12Gy given as a single radiosurgical treatment will have a much more marked effect than six fractions of 2Gy given as a course of radiotherapy (Gray, abbreviated to Gy, is the unit used to prescribe and measure the absorbed dose of radiation).

A separate but inter-related issue is the choice of machine used to deliver the radiation. This can be gamma knife, a linear accelerator (also called a LINAC) or Cyberknife.

- The **gamma knife** is a specialised machine that can only be used to treat conditions within the head or upper cervical spine. Gamma knife treatments are almost invariably performed as single fraction treatments, and so the terms “gamma knife” and “stereotactic radiosurgery” have become to some extent synonymous. This technique relies on a rigid frame for stereotactic localisation.
- In the UK most **LINACs** are used for general radiotherapy, treating different parts of the body, but they can be adapted and set up to deliver either single fraction or multi fractionated stereotactic radiotherapy treatments, using either frame or less accurate mask based localisation systems.
- A new technology for delivering stereotactic treatment is now available in the UK, known as **Cyberknife**. This is a light weight LINAC capable of being positioned flexibly by a high precision robotic arm.

In practice, reflecting the clinical experience and issues related to total radiation exposure, the majority of radiation treatments both for sporadic VS tumour in general and for NF2 tumours have been framed based gamma knife radiosurgery delivered as a single fraction.

Gamma Knife Radiosurgery for VS in Sheffield

Since the previous version of this fact sheet was prepared (2003), gamma knife radiosurgery has become the mainstay of active management for VS, this being an increasing alternative to surgical resection. Reflecting this, the Sheffield Unit has been carrying out up to 300 VS treatments per year and to date in total over 3000 of these tumours have been treated. As a result, a radiosurgery treatment for VS is a very routine procedure. Most of these treatments are for unilateral sporadic non-inherited (non NF2) tumours. Whilst the treatment is the same, the results and clinical issues in treating NF2 may be very different.

NF2 issues

In NF2, VS presents typically at a younger age and tend to grow more rapidly than sporadic tumours. As the disease is often bilateral, deafness or hearing preservation is much more of

an issue. Clinical experience is that treatment of NF2 VS either with surgery or radiosurgery is less effective than for sporadic (non-NF2) tumours. For these reasons we feel that it is essential to describe material that is specific for NF2 and not to generalise from the experience with sporadic tumours as this may be falsely optimistic. The results presented here are based on our cumulative experience from 1993 to 2010 treating 185 NF2 VS in 141 patients.

Referral and treatment

Patients are generally referred from specialist services throughout the UK with details of how the individual's condition has progressed, serial MRI scans and audiogram information. In deciding whether an individual is potentially suitable for radiosurgery, tumour size is the main consideration as the larger a tumour is the less likely it is to respond to and be controlled by radiosurgery.

If a patient is thought to be potentially suitable for radiosurgery, they are offered a clinic appointment to come and discuss this. All new patients are seen by Consultant Neurosurgeon, Mr Jeremy Rowe who has a specialist interest in NF2 radiosurgery. It is strongly recommended that all NF2 patients attend clinic before treatment. There is also an opportunity in clinic to meet with and talk to the therapy radiographers.

The treatment is carried out over a 2 night stay in hospital. Patients are admitted the day before treatment. On the day of treatment the stereotactic frame is fitted. This is the reference guide for the whole of the treatment and has to be rigidly fixed to the patient. This procedure is carried out with local anaesthetic, takes around 10 minutes and is very well tolerated. With the frame in place, an MRI scan is carried out to pinpoint the tumour and to map and plan the treatment. The treatment itself which is likened by many patients to "having another scan" is then performed. The frame is used to position the patient very precisely on the bed, which then moves into a tunnel. It is less enclosed than an MRI scanner, and there is no inherent noise (although patients can listen to music, talk over an intercom etc). At the end of the procedure, the frame is removed. Patients, who have travelled a considerable distance, are generally kept overnight, before being discharged the following day. Local patients can be discharged on the day of treatment.

Follow-up is generally performed locally by the patient's referring specialist. It is recommended that all patients are followed up with MRI scanning but this, of course, is part of the normal NF2 follow-up.

Radiosurgery results in NF2

After reviewing our NF2 practice, the most recent results suggest that eight years after the radiosurgery:

- Less than 10% of patients will have undergone surgery or further salvage therapy for the treated tumour.
- In 50% the tumour will be seen radiologically to have stopped growing or have shrunk.

- In 40% there will have been some variable concern about tumour growth, but that they will have been managed conservatively and will not have undergone surgery or other salvage treatment.
- 50% of patients will retain their functional level of hearing.
- In 35% there will be some deterioration in hearing.
- 15% become deaf in that ear.
- No facial weakness was recorded although a one case of facial spasm occurred which later resolved.

Whilst these results are not as good as those achieved treating unilateral sporadic tumours, the evidence is that radiosurgery does slow or stop the growth of the majority of NF2 tumours. The hope is to avoid the need for surgery in most patients. Given the morbidity and mortality associated with surgery, we believe that radiosurgery is an attractive treatment option for selected NF2 patients with growing tumours.

Radiosurgery for Meningiomas

The second most common tumour requiring treatment in the setting of NF2 is meningioma disease. In Sheffield the gamma knife to date has treated around 2000 sporadic meningiomas, again making this a routine treatment. In NF2 the meningioma disease is more likely to be multifocal. Reviewing our practice we have treated 38 NF2 meningiomas in 19 patients between 1987 and 2010. Of these 30 have shrunk on follow up imaging and only one tumour has required a salvage surgical resection. We expect that the demand for NF2 meningioma treatments will increase.

Common questions and concerns about radiosurgery

Questions which we are commonly asked include:

1. *What is the risk of a radiation treatment causing malignancy?*

We believe that the risk of this in the general population is very low indeed. To date there are only a few single patient case reports describing altered tumour behaviour after radiosurgery. World-wide over 500,000 patients have been treated with the Gamma Knife, over 64,000 for vestibular schwannomas.

In a systematic UK population based study we have not found any increased risk despite accumulating most recently 60,000 patient-years of data.

Of course the majority of these patients will not have NF2. Potentially the risk in NF2 may be different. The reason for raising this is the notion that a NF2 patient may inherit one faulty gene but require a mutation in the other healthy gene to develop a tumour. Radiation causes gene damage and is therefore potentially a concern. To address this safety issue, we are actively monitoring these events. To date we have treated over 160 NF2 patients and two have had malignant tumours. One tumour was growing rapidly (increasing in volume 15 fold in the 2 years before radiosurgery) and was found subsequently to be malignant. The rapid growth, however, preceded the radiosurgery and was not caused by it. The second patient developed a glioma 3 years after Radiosurgery but it is impossible to

say if the Radiosurgery was the cause of the 2nd tumour or whether this would have developed anyway. Overall the risks would appear to be low, and in practice have to be weighed against the morbidity and mortality associated with other treatment options.

2. What is the risk of radiosurgery causing scarring, and making subsequent surgery more difficult?

This is even harder to assess and quantify. The published literature is extremely anecdotal and relates to vestibular schwannomas in general, rather than to NF2. Individual surgeons have both reported surgery being harder and easier after radiosurgery (radiosurgery may decrease the blood flow through a tumour making an operation easier, but may cause scarring making it harder).

We are aware that some surgeons claim that there is always damage to the facial nerve after surgery if there has been previous radiosurgery. In evaluating these claims and making decisions about treatment, it is important to remember that it is the minority of patients who undergo radiosurgery who subsequently undergo surgery. Furthermore, in a review of British NF2 patients, most of whom had not undergone radiosurgery, 70% of patients had some degree of facial nerve dysfunction after surgery.

3. Can radiosurgery be repeated?

This is often asked, particularly by patients who have had unfavourable surgical experiences. The answer is yes: repeat treatments in other conditions are safe and there is considerable experience of this. In practice it is rare to re-treat VS. We would stress that this practice of repeating a treatment is very much the exception and that each case has to be judged on its individual merits.

Making a decision

Management decisions in NF2 are difficult because there is no ideal treatment option for all patients. Factors in making a decision include tumour size and how fast they are growing; the hearing in both ears, and other neurological deficits; and the individual's past experience either of their own care or other family members of the different management options.

The final decision rests with the patient. In making this decision we would however encourage the individual to have as many informed opinions as he/she finds helpful. We would hope that this article would clarify some of the issues surrounding radiation treatments.

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Other information

The information summarised in this fact sheet has drawn on a number of sources including;

www.gammaknife.org

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