October 2018

**South Wales Skull Base MDT**

**VESTIBULAR SCHWANNOMA (Acoustic Neuroma)**

**Protocol for management**

**Background**

Vestibular Schwannomas (VS) (sometimes called Acoustic Neuroma) are benign tumours arising from the vestibular nerve in its course between the brain stem and the inner ear. They are slow growing tumours which usually present with ENT symptoms – unilateral hearing loss with tinnitus and imbalance. Once they become large they are more serious, causing difficulty walking, facial pain, headaches, and, if untreated, death as a result of raised intracranial pressure.

These are rare tumours, with an incidence of about 10 – 15 new cases per million per year.

Traditionally, surgery was always the treatment of choice until it became recognised firstly that about a third of patients with small tumours can safely be monitored without treatment, and secondly that for small to medium sized tumours, single session stereotactic radiotherapy is very effective at controlling further growth without the need for surgery. Larger tumours still require surgery in order to prevent life-threatening complications.

**The South Wales Skull Base MDT**

This MDT is based in Cardiff and consists of specialists from Oncology, Neurosurgery, ENT, Ophthalmic surgery, Maxillo-facial surgery, and Radiology. There is a dedicated skull base coordinator. Currently there is a need also to appoint a specialist nurse.

The MDT manages a wide variety of patients with benign and malignant conditions of the skull base that may be arising in the bony skull base, meninges, brain, nasal sinuses, ear, mastoid, or orbit.

The team hold a joint clinic fortnightly on Wednesday afternoons and there is a fortnightly radiology meeting. New and follow-up patients are seen and / or discussed in these forums, and the team are seeing or discussing about 20 – 25 new skull base patients per month, or about 300 cases per year.

New VS patients represent the largest subgroup of these patients, and also require follow-up for at least 10 years (though some of the follow-up can be done closer to the patient’s home).

**Measurement of VS**

VS are classified as intracanalicular if there is no tumour extending beyond the internal auditory canal.

Those with an extracanalicular component are measured as the greatest diameter parallel to the petrous ridge (maximum extrameatal diameter).

**Overview of Management of VS**

There are 3 options for management of unilateral sporadic VS patients – observation, radiotherapy, or surgery.

Patients with small tumours (intracanalicular or <15mm) can safely be monitored with interval MRI scans, as about a third of these patients exhibit no significant growth long term. Regular MRI scanning is the only safe way to monitor growth, because there is no reliable correlation between symptoms and growth. Once the MDT has assessed the patient, surveillance monitoring should be carried out by the referring clinician closer to the patient’s home. Auditory and vestibular rehabilitation (mostly hearing aids and balance exercise programmes) can also be carried out locally. There is no general agreement regarding length of follow-up; older patients with stable tumours can be discharged after 10 years or sometimes earlier, but younger patients will need longer follow-up as tumours which are stable for many years can occasionally become active later.

There is currently no consensus on the best management of tumours in terms of hearing outcome. All patients should have an audiogram and speech discrimination score which should be sent to the Cardiff MDT on referral. Many patients with good hearing at presentation will retain hearing with observation alone.

If on subsequent MR imaging growth is demonstrated then active intervention with radiosurgery is warranted (see below).

Occasionally patients with small tumours may elect to undergo surgical excision, as the morbidity of surgery is much less if the tumour is small, and in some patients there may be the chance of hearing preservation with this approach.

Medium-sized tumours (15 – 30mm), including those which started out as small tumours which have grown to become medium sized, require treatment to prevent them becoming large tumours with much higher disease-related and treatment-related morbidity.

There are a variety of radiotherapy techniques available to treat these tumours. The commonest is single high dose stereotactic radiosurgery (SRS), delivered by a variety of machines (known as Gamma Knife, Cyber Knife, or LINAC). SRS is delivered locally at Velindre Hospital using a Truebeam™. This service is run by the integrated skull base team. The Truebeam™ delivers highly accurate radiosurgery using a frameless technique as a day case. SRS is a highly effective treatment for stabilising growing small or medium sized tumours and preventing further growth, with control rates of at least 95% being reported in most series. However, the patients require ongoing radiological follow-up for 5 – 10 years to check for possible relapse.

Some tumours between 25-30mm can cause compression of the trigeminal nerve with facial pain and numbness or cause brainstem compression. In these cases we do not recommend single session high dose SRS, but consider open microsurgery or fractionated radiotherapy to reduce the risk of permanent trigeminal neuropathy.

Large tumours (>3cm) require surgical excision, as SRS results in adverse radiation effects and neurological decline at higher tumour volumes. Occasionally in a patient unfit or too elderly for surgery, fractionated radiotherapy may be offered. Surgical excision is a major procedure with a joint neurosurgical / ENT team, operations lasting 6 – 8 hours, with a significant incidence of complications, including facial nerve weakness. We most commonly use a retrosigmoid or translabyrinthine approach with full cranial nerve and brainstem electrophysiological monitoring.

We have adopted an approach of facial nerve preserving surgery with complete excision where possible, but near total or subtotal resection is undertaken if needed to preserve facial nerve function, with rates of permanent facial nerve dysfunction (House- Brackman Grade 3 or more) of 5-8% in our centre. Open microsurgery requires an average 5-day hospital stay and patients often experience increased ataxia and headaches following surgery. Recovery from VS surgery is highly variable and return to work can take up to 3 months. Patients are allowed to drive once clinically recovered from surgery – usually around 4- 6 weeks. Where there is residual tumour radiotherapy may be offered after surgery to prevent future regrowth.

Patients with bilateral tumours (Neurofibromatosis type 2) form a small subgroup, which need to be managed differently. These patients usually have more aggressive disease as well as neurofibromata and other tumours elsewhere, making the management much more difficult. They require genetic counselling and other input. They may need to be referred to a quaternary centre such as Manchester, and considered for treatment with Bevacizumab. NF2 associated VS are not included in these guidelines but are managed with joint input from the Manchester centralised NF2 service on a ‘hub and spoke’ basis.

**VS Treatment Algorithm**

All new patients should be discussed and their scans reviewed at the Skull Base MDT. All VS patients will be entered into the National Vestibular Schwannoma audit database.

**Small Tumours** i.e. Intracanalicular and < 15 mm in CPA

Managed by observation.

Audiogram and SDS on presentation and yearly

 Repeat MRI scan every year for 3 years, then every 2 – 3 years, for at least 10 years / age 80.

If any signs of growth, refer back to MDT for decision on whether to offer SRS or continued observation.

Following initial discussion at MDT, follow-up should be done by referring consultant closer to patient’s home.

Surveillance imaging results should be copied to the central skull base MDT for inclusion in national audit.

**Medium Tumours** i.e. 15 – 30 mm in CPA.

 Managed with radiosurgery (SRS).

Some tumours sized 25 – 30 mm may be volumetrically too large for SRS especially if there is brain stem compression or trigeminal symptoms. Treatment options are surgery or fractionated radiotherapy.

Cystic tumours may have better outcome with surgery rather than SRS and the treatment decision is based on MDT discussion

 Follow-up: repeat MRI at intervals as above with small tumours. All scans reviewed by MDT.

Outcome data: MDT keeps database of long term control rates and complications of treatment.

**Large Tumours** i.e. > 30 mm in CPA

Managed with open microsurgery, unless elderly or unfit –where fractionated radiotherapy may be an alternative option.

 Follow-up: After total resection: MRI at 3 months, 2 years, 5 years.

 After subtotal resection: MRI at 3 months, 1, 2, 3, 5, and 10 years.

If nodular residuum, either observe or treat with SRS dependent on volume of residual and patient age.

Outcome data: MDT keeps database of long term control rates and complications of treatment.

**Follow up after SRS/SRT**

Following radiosurgery or radiotherapy patients are seen at the UHW skull base clinic:

6 month clinical review – MRI at earlier interval if symptomatic.

Otherwise MRI at 1 year, then annual MRI for 5 years then every 2 years for 10 years.

Follow-up beyond 10 years dependent on age.