NF2 TREATMENT OPTIONS

Children's Tumour Foundation
CONQUERING NF

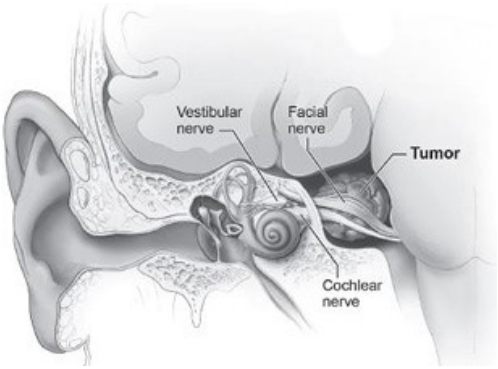
The information provided below is a summary of the presentation given by A/Prof Nigel Biggs on 7 March 2020 at the NF Information Day in Sydney.

Associate Professor Nigel Biggs is Chairman, Department of Otolaryngology, Head and Neck and Skull Base Surgery and the Director of the Microsurgical Skills Laboratory at St Vincent's Hospital. He is also an Associate Professor at the University of Notre Dame, Senior Lecturer at the UNSW, and an examiner in Otolaryngology, Head & Neck Surgery with the Australian College of Surgeons.

WHAT ARE YOU NF2?

NF2 is a genetic condition that is characterised by the development of two separate benign tumours, called **vestibular schwannomas** on the hearing and balance nerves in the brain. This is called the Vestibular Nerve and is also known as Cranial Nerve V111. Vestibular schwannomas were previously known as **acoustic neuromas.**

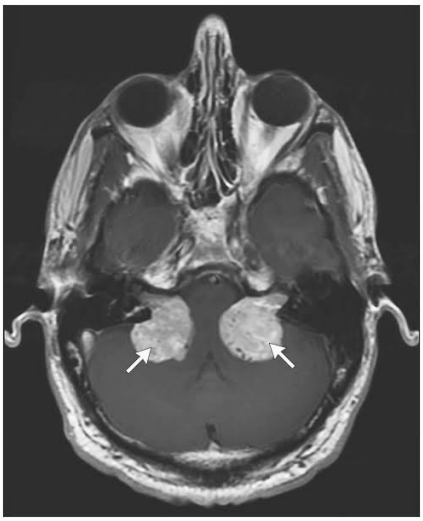
NF2 is rarer than NF1. NF2 is said to occur in approximately 1:25,000 people, it is found on a different chromosome and therefore is an entirely different genetic condition to NF1.



Other than hearing loss, the other main symptoms of NF2 inlcude:

- Tinnitus (ringing in the ears)
- Headache
- Dizziness
- Speech and/or swallowing problems
- Changes in walking ability

NF2 is usually diagnosed in the late teen years or early adulthood. Some children are diagnosed before they are teenagers and these cases tend to be more complex. The age of the individual at diagnosis is generally an indicator of the severity of NF2 and the level of impact the condition will have on quality of life.



NF2 causes tumours to grow within the brain and spinal cord (the Central Nervous System) rather than on the nerves found elsewhere in the body as usually seen in NF1. As there is a lot less space in the Central Nervous System, a growing tumour causes problems by compressing many other nerves and tissues.

NF2 Brain tumours are benign and there is an extremely low risk of them becoming cancerous.

There is another condition which is similar to NF2 where people develop a Sporadic Vestibular Schwannoma. These occur in 1:80,000 and are found on only one of the auditory nerves. These people therefore only have hearing loss on one side and can still hear with the unaffected ear. These schwannomas are not as aggressive as those seen in NF2 and they have better treatment outcomes.

MANAGEMENT OF NF2 IS COMPLEX

Due to the location of the NF2 tumours, management can be very challenging.

NF2 specialists will consider three main questions:

- When to treat?
- How to treat?
- How do we preserve hearing?

NF2 doctors will also take into account the person's age, size and growth rate of the tumour and the hearing and balance implications.

Generally, if tumours are small, the doctors will recommend a period of surveillance and monitoring with annual or biannual MRIs. They can then determine the growth rate of the tumours and intervene if necessary.

Some tumours have periods of rapid growth and then periods of relative stability.

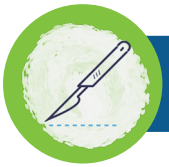
NF2 specialists are reluctant to treat early as intervention will invariably have adverse impacts on hearing function and other complications can arise.

MANAGEMENT OF NF2 IS COMPLEX

Surgery is the main treatment option.

Radiotherapy can be considered in certain areas.

Bevacizsumab (**Avastin**) is a tumour shrinking drug which is becoming more frequently used as a treatment option of NF2.



WHAT ARE THE INDICATORS FOR SURGERY?

NF2 Surgeons will consider operating if there is evidence of compression of brain structures by the tumours. This can be obvious if there is hearing loss, ringing in the ear (tinnitus), loss of balance or facial weakness.

The consequences of surgery can be severe. Surgeons will attempt to preserve the facial nerve and hearing if possible. However, sometimes as they are removing the tumour these nerves are damaged, resulting in facial paralysis and/or hearing loss. This is why surgeons will often elect to monitor and review a person for as long as possible rather than offering surgery in the first instance.

NF2 tumours can be removed in its entirety in approximately 50% of cases.

When the tumour has not been able to be completely excised, surgeons may need to operate a second time. This can be problematic as they will be encountering scar tissue from the previous surgery and there is a greater degree of risk.

Regardless, the goals of surgery are to:

- Remove or reduce the size of the tumour
- Avoid causing any facial weakness
- Retain hearing function if possible
- Maximise outcomes and quality of life

WHEN WOULD RADIATION THERAPY BE OFFERED AS A TREATMENT OPTION?

Radiotherapy may be considered for people who do not want surgery.

There are side effects of radiotherapy (such as skin irritation) and it has a negative impact on hearing.

There is a well-documented risk of developing cancer in radiated areas of the brain later in life. NF2 specialists are therefore concerned about offering this treatment option to young people with NF2 as their lifelong risk is higher than in older people.

Stereotactic Radiosurgery is a single dose of Radiotherapy that is usually delivered by a machine called a Gamma Knife. This delivers a maximum dose of focal radiation to the tumour and attempts to restrict the amount of radiation to surrounding structures and tissues of the brain.

Fractionated Radiotherapy can be offered over the course of a few days.

The goals of radiotherapy are to stop the growth of the tumours, but it will not remove them. If radiation therapy has been attempted prior to surgery, the surgical procedure can be more difficult as radiation changes the character of the tumour. There is a higher risk of facial nerve paralysis following surgery on patients who have also undergone radiotherapy.

WHAT ABOUT AVASTIN FOR NF2

Whilst the use of **Bevacizumab** (commonly called Avastin) is in the early stages, it is likely that it will be the preferred future treatment of NF2.

Early studies of Avastin show that the drug can shrink some tumours by effecting the blood supply. This can arrest progressive hearing loss and therefore improve or maintain quality of life.

Avastin can cost up to $50,000 per annum in Australia.

Avastin can cause side effects - it has been shown to reduce fertility and can cause problems with blood pressure and kidney function.

WHAT ARE THE OPTIONS FOR HEARING REHABILITATION FOR NF2?

Hearing rehabilitation for people with NF2 is challenging as the tumours effect the nerves on both sides. Other types of hearing losses can be more easily remediated as they generally have one functioning auditory nerve.

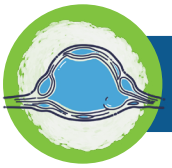
One options is a **Brainstem Implant,** which is an electrode that is implanted into the surface of the brain during surgery. It is available, but not commonly performed in Australia as the results can be variable and requires a highly skilled surgeon.

Cochlear Implants rarely work for NF2 patients as the implant requires a functioning auditory nerve to transmit the signal to the brain. In NF2 the nerve is either impacted by tumours (or has been removed by the surgeon). In instances where the auditory nerve has been preserved, a Cochlear Implant can be trialed. Cochlear Implants require magnets to attach them to the skull. Magnets cast a shadow on MRIs which therefore inhibits future monitoring of tumour size and growth.

A **Cross Aid** is a Hearing Aid that transmits the sound signal to the opposite side of the brain.

Bone Conduction Hearing Aids can be implanted in the bone at the base of the skull. The sound signal travels through the bone to the other ear. The sounds can appear muffled and this type of aid has variable tolerance.

Some of these aids are accessible from Hearing Australia.



WHAT OTHER TUMOURS ARE COMMON IN NF2?

NF2 can cause tumours on other cranial nerves, especially the **facial nerves (V11) and trigeminal nerve (V).**

Many of these other tumours are treated conservatively as facial paralysis following surgery is unfortunately quite common.

Meningiomas are tumours found on the lining of the brain and quite common in NF2. If necessary, these are treated by Neurosurgeons when and if they are threatening the function of other body parts. These tumours are not usually suitable for radiotherapy.

People with NF2 can also develop **spinal tumours.** These are usually quite small and slow growing. They are usually treated conservatively, and surgery is only offered if function is severley impacted.

If you have any questions regarding this information, our Support Team is available to help. You can reach them via phone on (02) 9713 6111 **or by email at** support@ctf.org.au