



NF2-related Schwannomatosis: Sudden Hearing loss

Advice to GPs for management

NF2-related Schwannomatosis (NF2-SWN)

NF2-SWN is a variable and unpredictable condition affecting people in different ways. The main signs of NF2-SWN are benign (non-cancerous) tumours called vestibular schwannomas (formerly called acoustic neuromas) which grow on both hearing nerves. You have one hearing nerve on each side of your brain.

In addition to these, other benign tumours associated with the nervous system may occur, in particular:

- The lining of the brain (meningiomas)
- The spine (meningiomas, schwannomas, ependymomas)
- The skin (schwannomas)

Each hearing nerve has two parts. The *cochlear* nerve carries information about sound and the *vestibular* nerve carries information about balance to the brain.

Over time these tumours cause deafness. A tumour on one side may grow at a different rate to the one on the other side. The size of the tumours may bear little relationship to the degree of deafness. For example, a small tumour may produce deteriorating hearing whilst a large tumour may only cause minimal hearing loss.

Hearing Loss

A sudden loss of hearing may be conductive (e.g. wax blockage, trauma or fluid in the middle ear) or sensorineural (cause unknown, or related to tumours such as a vestibular schwannoma). Often the hearing loss with vestibular schwannoma is gradual, but it can also be sudden. Patients with NF2-SWN are therefore at high risk of sudden sensorineural hearing loss (SSNHL).

Sudden hearing loss for any patient is distressing and management should take place urgently.

An accepted definition for SSNHL would be:

- Rapid onset (within the last 72 hours)
- Decrease in hearing of ≥ 30 dB affecting at least three consecutive frequencies (compare to opposite ear if appropriate)
- Affecting adults (16+)
- Losses of less than 30dB may still be treated but should be discussed with the patient + advice gained from ENT
- Paediatric patients should be discussed with ENT

Around 1/3 to 2/3 of general patients with SNHL recover their hearing in part or in full. This figure is not known for NF2-SWN patients. Given the risk of bilateral profound hearing loss in patients with NF2-SWN, a short course of high-dose steroids is an acceptable intervention. Although many alternative treatments have been tried to manage SNHL there is little evidence for benefit.

With NF2-SWN patients, we recommend that patients are urgently referred to their local ENT service or NF2-SWN team for emergency management; the available guidance below will be useful if this is not possible.

1 If someone with NF2-SWN presents with sudden hearing loss, firstly check for any clinical signs of blockage, infection or effusion. Patients require an urgent hearing test to establish whether the loss is conductive or sensori-neural. It is also possible to use a tuning fork to establish the type of hearing loss, although this can be more challenging. It is useful to compare the hearing test with older hearing tests if they are available.

Consider the patient's clinical history including any associated vertigo or tinnitus, ischaemic (refer urgently to medical team if concerned), previous episodes, risk factors for coagulopathy and previous ear disease.

If it is not possible to get a hearing test e.g. out of hours or over the weekend, the ear canal is free of wax and infection and there is no effusion, **commence oral steroids as below and refer patient for an urgent hearing test as soon as possible**. This is based on the rationale that early intervention is more likely to lead to benefit; this should be balanced against the risk of oral steroids.

2 Prescribe oral prednisolone at 1mg/kg to a maximum of 60mg for 10 days, with appropriate stomach protection if risk factors for gastric complications. It is best for patients to take the dose in the morning, after breakfast.

Prednisolone is used as a short-term preventative treatment of sudden hearing loss. For most people, including pregnant or breastfeeding women, steroids are safe. However, they should only be used if the potential benefits outweigh the risks and there are some circumstances when they should not be used or should only be used with caution.

These circumstances include:

- having an ongoing widespread infection and/or healing wound
- having mental health or behavioural problems – such as depression, psychosis or alcohol dependence
- having certain underlying physical conditions – such as liver problems, heart failure, high blood pressure, a recent heart attack, epilepsy, osteoporosis, obesity, stomach ulcer, glaucoma, underactive thyroid gland or diabetes
- Taking another medication that may interact with corticosteroids – such as NSAIDs, live vaccines, HIV, anti-epileptic, anti-coagulant and diabetes medications.

Current guidelines suggest that patients on doses over 40mg for over one week should be supplied with a Steroid Emergency Card as they are considered at risk of adrenal insufficiency.

What are the possible side-effects?

Side-effects: As with all medications, there are potential side effects in a small number of patients. The commonly reported side effects are: Anxiety; behaviour abnormal; cataract subcapsular; cognitive impairment; Cushing's syndrome; electrolyte imbalance; fatigue; fluid retention; gastrointestinal discomfort; headache; healing impaired; hirsutism; hypertension; increased risk of infection; menstrual cycle irregularities; mood altered; nausea; osteoporosis; peptic ulcer; psychotic disorder; skin reactions; sleep disorders; weight increased.

3 ENT team should review the patient as soon as possible for consideration of intratympanic steroids.

4 Please inform the patient's treating NF2-SWN team as soon as possible. The contact details for the national NF2-SWN hub centres are as follows:

Cambridge (Addenbrooke's Hospital)

01223 348672 – specialist nurses

01223 805128 – NF2 secretary

Manchester (St Mary's Hospital)

0161 701 0422 – NF2 coordinator

NF2 Nurses Advice Helpline – 0161 276 4619

London (Guys and St Thomas' Hospital)

020 7188 1029

NF2 Nurses - 020 7188 9976

Oxford (John Radcliffe Hospital)

NF2 Oxford Office: 01865 231 889

NF2 Oxford Nurses: 01865 231 741 / 01865 231 745

The NF2 service is an outpatient Mon-Fri service. If contacting out of hours or your query is urgent and you cannot reach the relevant team, please contact your local ENT on call service for advice.

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