

Guideline for managing patients with NF2 contacting the service complaining of sudden onset hearing loss.

Background Notes:

A sudden reduction in hearing may be **conductive** (wax blocking ear canal or fluid in middle ear due to an upper respiratory tract infection) or **sensorineural** (idiopathic or associated with underlying pathology such as a vestibular schwannoma).



We know that vestibular schwannomas cause sensorineural hearing loss although we do not fully understand the mechanism. This hearing loss may be gradual but can also occur as a sudden loss. Patients with NF2 are therefore at high risk of this.

For any patient (including non-NF2) sudden onset sensorineural hearing loss (SSNHL) is distressing and should be managed promptly. There is little high quality evidence to guide best practice in this condition and what we do know comes mainly from studies in non-NF2 patient groups.

Diagnosis:

Patients with NF2 can also suffer from sudden onset of hearing loss from wax or fluid in the middle ear (effusion/glue ear). Therefore patients require an urgent audiogram to clarify whether the hearing loss is conductive or sensorineural. If the patient has existing hearing loss it is important to be able to compare the current audiogram with a previous one. Tuning fork tests can also differentiate between a conductive and sensorineural hearing loss although interpretation can be more difficult if there is already significant hearing loss present.

There is not an accepted **definition** for SSNHL but most commonly quoted is that the loss must occur over less than 3 days and be a reduction of more than 30dB across 3 consecutive frequencies. In reality many clinicians would expand the criteria to include less severe losses. Current definitions do not use SDS (speech discrimination score) although it is likely that this would be a useful measure of rapid deterioration in NF2 patients. Access to SDS testing may be limited in some hospitals, especially on an urgent basis, reducing its practicality in this setting.

Management:

It is believed that somewhere between 1/3 to 2/3 of patients with SSNHL recover their hearing partly or completely without any intervention. Given the disability and distress associated with the loss, multiple treatments have been trialled for this condition to try and increase the proportion of patients who recover. Unfortunately there is little evidence to show benefit for any of these. Most ENT doctors in the UK would agree that giving high dose oral steroids (up to 1mg/kg or 60mg for 1 week) as soon as possible after the onset of hearing loss is probably the most accepted intervention. This must of course be balanced against any relative contraindications/risks for the individual patient. Some units admit patients for Carbogen, antivirals, thrombolytics, vasodilators, vasoactive substances, or antioxidants, although the evidence is lacking and these must be balanced with the risk of harm. Hyperbaric oxygen is also used although availability is clearly a problem.

If the hearing loss occurs out of hours or on a weekend, an audiogram may not be possible. Clinical assessment (history, otoscopy and tuning fork tests) to differentiate between a conductive loss and a sensorineural should be performed by the GP/on-call local ENT staff. Associated symptoms of vertigo and tinnitus may point towards an inner ear type of hearing loss. If this assessment suggests SSNHL, most would suggest starting empirical steroids rather than waiting for confirmation with an audiogram. This is based on the rationale that any potential benefit is more likely with early intervention, although this has to be balanced with the risk of oral steroids and the possibility that the hearing loss may not in fact constitute SSNHL.

Proposed Management:

Any patient who contacts the service with sudden onset hearing loss should be managed as an emergency, as should any non-NF2 patient. They should be assessed clinically by their local GP and referred to their local ENT department in order to obtain audiometry as soon as practically possible. If SSNHL is confirmed or clinically suspected (awaiting formal audiometry) then high dose oral steroids should be considered for 1 week.

It would be helpful if patients kept a copy of their most recent audiogram so that this can be used for comparison.

Given the difficulties of obtaining emergency appointments in primary care and the lack of awareness of this condition and the need for prompt intervention, it may be helpful for patients if we provide a letter stating that they are at risk of SSNHL and that they should be assessed urgently and referred if any concerns.

