CHAPTER 12.

INNER EAR CONDITIONS

CONDITIONS

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1. HEREDITARY SENSORINEURAL DEAFNESS

Inherited SND includes a wide variety of conditions, relatively simple or complex. Extensive coverage is found online (see Smith RJH, Shearer AE, et al, “Deafness and Hereditary Hearing Loss Overview). The problems range from GJB2 mutation (Connexion 26) profound loss at birth, to pre-senile (early onset) presbyacusis.

A relatively common variant is termed “cookie bite” losses. These are midfrequency losses, bilateral but often asymmetrical.

Due to residual good upper and lower frequency hearing, the problem may be missed for extended periods. The condition, autosomal recessive in nature, is often progressive and may lead to eventual cochlear implantation (CI).

Figure 1: Cookie Bite SND, showing the typical mid-frequence “sag” in hearing thresholds.

Otosclerosis (covered in Middle Ear Surgery) is a hereditary conductive deafness that commonly incurs sensorineural deafness, but the latter is acquired with time and is thought to be due to extension of the disease into the cochlea, rather than hereditary inner ear deafness per se.
2. CONGENITAL INNER EAR MALFORMATIONS

Deformities of the inner ear are a major cause of profound congenital sensorineural deafness. They may occur in conjunction with outer and middle ear deformity, or in isolation. Their assessment requires meticulous investigation to clarify the extent of loss and their suitability for CI, in the worse cases.

a) Cochleo-vestibular

Figure 2: Advanced Mondini-type deformity. The overall shape of the cochlea resembles the normal, but there is loss of the partitioning of the cochlear spirals, and the inner ear is in continuity with the CSF.

Cochlear deformities display a spectrum of disorders form mild aberrations to total aplasia. Lesser (Mondini) types display loss of the partitions of the cochlear, usually with substantial or total hearing loss. These cases do well after cochlear implantation.

Figure 3: “Comma” cochlear dysplasia, showing the basal turn, but with poor development of the upper spiral.

More major forms permit only partial CI electrode insertion and are less successful. These vary from under-developed spiral length, to globular cochlear patterns that may form a common cavity with the vestibular system.

Another pattern that may be missed at birth is the problem of large ventricular aqueducts. These present with bilateral profound losses in some cases, but alternatively may cause a progressive SND, perhaps over many years, prone to unpredictable hearing fluctuation, then rapid decline, perhaps triggered by minor trauma. The losses respond well to CI.

Figure 4: Large ventricular aqueduct. The duct is in continuity with the CSF. Profound loss at birth, or unstable sensorineural losses later in life, commonly leading to CI.
b) VIIITH Dysplasia

Other cases may have severe dysplasias, often associated with VIIITH nerve abnormalities. The problem is not uncommon in CHARGE association cases. CI is unsuitable in these cases; some will benefit from an auditory brainstem implant.

Figure 5: Gross inner ear dysplasia, stenotic IAC, and VIIITH nerve agenesis in a CHARGE child.

3. AGING DEAFNESS
(PRESBYACUSIS)

With age the great majority of the population will exhibit loss of hearing. Presbyacusis derives from Greek roots: “presbys” (elders) and “akousis” (hearing).

Pathogenesis

The loss is due to degenerative wear and tear damage to the hair cells of the inner ear and has no other health implications. The losses often have genetic origins, but may be exacerbated by general medical conditions, including high blood pressure, diabetes, obesity, smoking, and high cholesterol levels. In a few cases the losses may emerge at relatively young ages, but generally in middle or later age. The male population is more prone, and the problem evidently affects the urban populations more than those in quiet rural situations. A small minority are excepted, retaining excellent hearing into advanced age.

Figure 6: Presbyacusis. Bilateral symmetrical high frequency SND in an aging patient.

Presentation

The deafness emerges as subtle high frequency reduction, slowly progressive over many years. The hearing loss is very symmetrical, into the high frequencies.

Tinnitus may accompany the losses, also slowly progressive; this aspect may be worsened by the reduced external ambient noise as the hearing drops.

Speech discrimination is generally initially good, deteriorating with the advance of the condition. There are generally few other symptoms, although irritability to loud noise (recruitment) may become evident, hence the “don’t shout, I’m not deaf!” scenario.
Management

Treatment for aging losses is by conventional aiding, using either in-the-ear or behind-the-ear air conduction aids. However, in some cases the losses become so advanced that hearing aids are impractical, as the ability to discriminate speech is lost. This group benefits greatly from cochlear implantation, which restores communication and reduces isolation.

4. SUDDEN SENSORINEURAL DEAFNESS (SSND)

Sudden loss of hearing due to nerve damage is a not uncommon occurrence. The cause is uncertain, possibly blockage of the labyrinthine artery, the only blood vessel to the ear. The deafness is usually permanent, with some recovery being gained in a minority of cases. The damage is rapid in onset such that treatment is not available in time to prevent the loss.

The loss may or may not be associated with loss of balance and/or tinnitus.

Management options

a) CROS aid
b) Bonebridge implant system
c) Bone anchored hearing aid (BAHA) implant system
d) Cochlear Implant (CI)
e) Communication strategies and hearing tactics/assistive listening devices

A CROS aid (Contralateral Routing of Sound) works via a receiver device on the deafened ear that transmits to an aid in the better ear (by Bluetooth). This eliminates the dead spot of hearing that is troublesome at a table, in meetings or groups, etc. The device does not restore direction-finding, stereo effect or the better appreciation of sound afforded by two ears. The CROS aid can give many benefits but does not require surgery.

A similar BiCROS aid also amplifies sound for the better ear if this side is partly deaf, e.g. due to age.

Bone conduction implants (Bonebridge, BAHA) work by stimulating the inner ear by vibrating the skull and are applicable only if there is good hearing in the better ear (thresholds < 20db, as above). The Bonebridge device is similar to a cochlear implant and is inserted via a “thumbnail” incision on the rear of the ear, and is powered by a magnetic button type processor on the scalp. BAHA devices require a larger C-incision (and the corresponding hair shave) on the scalp and the magnetic-attached processor is slightly larger than the Bonebridge.

The benefits of the bone conduction devices are higher quality hearing, but with the constraints of the CROS aid as above, and are applicable only with excellent contralateral hearing.

Figure 7. Profound left sensorineural deafness. This will cause a head shadow zone, and loss of stereo effect, direction-finding ability, and binaural summation.
If one of the bone conduction type implant devices (BAHA or Bonebridge), is of interest, the patient is offered the opportunity to take an external bone conduction hearing aid on trial. This simulates what it is like to hear with a bone conduction device. If this is satisfactory, the audiology staff perform aided testing with and without the device to check that the device provides sufficient benefit.

Patients who are considering surgery for a bone conduction implant will be asked to trial a CROS aid for a week, to ensure that they are fully informed about your options. This is mandatory.

Alternatively, a cochlear implant (CI) may be considered. These implants provide better all-round sound awareness and stereo effect but require greater adaptation to the quality of sound received. They are effective even when the other ear is deaf. CI surgery normally eliminates residual hearing.

If the hearing in the ear considered has been impaired for a very long time, or since birth, CI is not an option.

5. NOISE-INDUCED DEAFNESS

Exposure to excessively loud noise levels will produce loss of hearing, due to damage to the sensory hair cells of the inner ear. Losses may be due to chronic exposure, single loud incidents, or as a result of a blast effect. Each tends to present in a different pattern.

Pathogenesis

Chronic noise damage is due to concussive effects of either repeated loud sounds (e.g. factory work, or chronic continuous loud noise. This damages the ear at levels above 85 db, continued for more than 3-4 minutes. The damage worsens with continued exposure, but stabilises if the noise is discontinued. Apart from reducing the noise itself, ear muffs are the optimal mode of damage...
prevention, as even specialised noise suppression earplugs provide less protection.

Single loud incidents may cause marked loss, evidently if exposure occurs with a head-down posture. “Acoustic accidents” of this nature are frequently difficult to prevent, being often one-off events.

Blast injuries may be complex. The force of the incident may compress and rupture the drum, perhaps impacting fragments of drum skin into the middle ear. Conductive (mechanical) losses may result from persistent drum perforation, middle ear scarring, chain dislocation, or from skin fragments forming cholesteatomas that may expand and compress or erode the middle ear structures. Blasts may also cause a perilymph fistula, when rupture of the inner ear orifices produces leakage, with nerve deafness and perhaps severe dizziness.

Investigation

Audiological findings tend to reflect the cause of the noise trauma. Chronic damaging environmental sound produces a symmetrical high frequency nerve deafness, maximal at 4000 cps, forming a characteristic hearing dip at this frequency. This as because the cells that activate in response to this frequency are closest to the entry of sound into the inner ear.

![Figure 10: Chronic noise trauma. Ear muffs are mandatory in these situations.](image)

![Figure 11: Relatively mild chronic noise exposure effects. Characteristic symmetrical V-shaped losses at 4000 cps suggest chronic exposure, both ear suffering equivalent losses.](image)

![Figure 12: Severe unilateral exposure to noise, resulting in losses in one ear that are greater than the other.](image)

In some cases (temporary threshold shift) the initial losses may recover in part or full.

Conversely, single severe episodes tend to cause unilateral losses, reflecting the side of exposure. “Rifleman’s Ear”, for instance, shows a worse pattern in the left side (for a right-handed shooter), as that ear is closer to the muzzle blast of the weapon.
Blast audiological patterns may show a severe unilateral nerve loss, perhaps total, bilateral losses, worse on the blast side, or a mixed nerve and conductive loss in the event of significant middle ear damage. A perilymph fistula may demonstrate a fluctuating low frequency loss, leading to further or total nerve deafness.

6. BENIGN POSITIONAL VERTIGO
(BPV, BENIGN PAROXYSMAL POSTIONAL VERTIGO, BPPV)

Within the balance mechanisms of the inner ear, two structures (utricule and saccule) are used to detect acceleration and head tilt. This is done via jelly-like glycoprotein masses resting on a sheet of cells equipped with fine sensory hair-like filaments that detect movement of the overlying mass. The sensory cells connect to the balance nerves to the brain and signal these movements to the brain as being due to acceleration or tilt movements.

Pathogenesis
The gelatinous material contains crystalline particles that may become dislodged if the structures are subjected to severe vibration, usually as a result of a head injury or noise exposure.

When dislodged, the crystals float in the fluid that bathes the inner ear structures, and frequently accumulate in the posterior semicircular canal. If the head is moved in particular patterns, the crystals accumulate on the sensory hair cells that detect rotatory movement, causing aberrant signals that are felt as severe spinning sensations: rotatory vertigo.

Presentation
The aberrant signals are generated by specific movements, characteristically when arising, looking up sharply, rolling over in bed, (usually to a specific side), or by sudden horizontal turning actions. The sensation of spinning generally lasts for a period of approximately up to 10 seconds, then abating. There is no directly related hearing loss tinnitus or other ear symptoms, but a history of trauma or noise exposure may be given.

The symptoms may be provoked by body and head movement assessments (Hallpike manoeuvres), which may also produce visible nystagmus. More sophisticated electronystagmographic balance testing may confirm this phenomenon, but show no other findings. Audiological testing may confirm previous hearing damage.

Management
Fortunately, the condition generally responds well to specially targeted physiotherapy (Epley manoeuvres), which moves the offending particles away to less troubles sites in the inner ear. These exercises can be repeated at home as required to control the problems.

Less commonly, troublesome dizziness requires surgery on the inner ear to control the problem (posterior canal occlusion). This is not without risk, and care should be taken not to confuse BPV with other forms of positional vertigo that are due to brain damage rather than to the ear.

The problem is diagnosed essentially from its repetitive pattern, brief duration, in specific trigger situations, and the lack of other inner ear manifestations.
7. LABYRINTHITIS

Pathogenesis

The inner ear may in some circumstances, become damaged by either viral or bacterial infection, both of which may cause severe damage and symptoms. Viral infections may occur after common influenza viruses or, mainly in children, from childhood infections such as measles or mumps, fortunately rare after routine childhood vaccinations. Adults may suffer herpes zoster oticus (Ramsay Hunt syndrome) with an associated facial palsy. Bacterial infections originate either from middle ear infections (acute or chronic), or from meningitis; the latter not uncommonly affects both ears.

Viral labyrinthitis in children may pass unnoticed amongst other concurrent illness, being identified after a considerable time lapse. In adults, the problem is obvious from the one-off triple symptoms of deafness, dizziness, and tinnitus. The damage from viral infections is confined to the hearing receptor cells.

Conversely, bacterial patterns tend to be associated with pain or discharge, bringing attention to the site more rapidly. Acute middle ear infection may affect the inner ear directly or via secondary meningitis. Chronic disease usually involves cholesteatoma, an infected cyst of skin in the middle ear that erodes the otic capsule, creating a fistula into the inner ear.

Figure 13: A large lateral semicircular fistula (central), resulting from a cholesteatoma. Dead ear from bacterial labyrinthitis.

Unlike viral disease, bacterial infection commonly causes scarring then bone formation within the inner ear (labyrinthitis ossificans) that may obliterate the hearing structures. This may prevent cochlear implantation. For this reason, meningitis is regarded as a hearing emergency for careful monitoring of the hearing, as the obliterative changes may develop rapidly. Profoundly deafened cases are implanted early (within a few months) after diagnosis, when indicated.

Presentation

Labyrinthitis rapidly produces an unhappy triuplicate of symptoms: deafness, tinnitus and vertigo. Deafness maybe profound and is usually irreversible; tinnitus loud, often persistent and troublesome. The initial acute vertigo, often nauseating and prostrating, gradually regresses through a phase of unsteadiness on movement (disequilibrium) over 6-8 weeks. Skilled physiotherapy may help in this respect.

Any initiating bacterial origin will require urgent medical and/or surgical management according to the clinical situation.
Management of the hearing loss will depend on several factors. Being nerve damage in nature, surgery will not restore the original hearing ability. Lesser losses may respond to hearing aids. If this is ineffective, cochlear implantation is considered, especially in children, but also in selected adult cases, particularly the bilaterally afflicted. Pre-CI assessments may need to ensure that the VIIIth nerve remains undamaged.

For those with normal hearing in the unaffected ear, an active bone conduction implant in the deafened ear offers improved hearing. These devices (Med El Bonebridge, Cochlear (BAHA)) eliminate the “head shadow” deaf zone around the afflicted ear by stimulating the better cochlea, but direction finding, stereo effect and better all-round hearing are not normally achieved.

Hearing loss, tinnitus and an evident cause of deafness distinguish this condition from vestibular neuronitis, where imbalance alone is present.

8. VESTIBULAR NEURONITIS

The inner ears are the major mechanisms of maintaining balance and are connected to the brain by the auditory nerves. Each nerve has two divisions, the hearing (cochlear) nerve and two balance (vestibular) nerves. The balance function of the ears is prone to sudden malfunction that causes profound loss of balance.

Pathogenesis

The precise nature of the problem is ill understood, as the condition is not fatal; the tissues of the ear and the balance nerves cannot be studied at the time of these problems. It is thought that the problem is due to viral infection in the balance nerves, although why the hearing division is not affected simultaneously is uncertain. Another possibility is blockage of the branch of the labyrinthine artery that supplies the balance mechanisms themselves.

Presentation

The onset of the dizziness is sudden, the patient commonly becoming disabled by a severe spinning sensation (rotatory vertigo), accompanied by nausea and vomiting, which may last 1-2 days, worse when moving. There is no deafness, tinnitus or other ear symptoms, and no sensation of loss of consciousness or faintness, and the patient may have no other symptoms of illness that might be related.

The spinning sensation fades, to be replaced by gross unsteadiness if the head is moved (disequilibrium). This is due to loss of balance function on the affected side. With time, as the brain compensates for the altered balance capability, the unsteadiness gradually settles. Intermittent spikes of unsteadiness may trouble the recovery, which may take up to 6-8 weeks, more rapidly in the younger patient. Chronic unsteadiness may trouble the elderly; a “labyrinthine cripple” effect that may also result from other disease (e.g. ELH) or from destructive surgery.

Audiology and radiology show no specific changes but balance tests may show reduced function in the affected ear.
Figure 14: Electronystagmography in Vestibular neuronitis. Severe loss of response from the right ear. Normal audiology. Labyrinthitis would produce a sensorineural loss, perhaps profound.

Management

Medical treatment aims to minimise the nausea and unsteadiness, but do not cure the origins, which fortunately dissipate with time, helped by vestibular rehabilitation physiotherapy.

The clinical picture is thus one of a single episode of sudden vertigo, then lingering imbalance, settling spontaneously but without other attendant ear or neurological symptoms. The condition is distinct from labyrinthitis, as this also destroys hearing.

9. ENDOLYMPHATIC HYDROPS
(ELH, MENIERE’S DISEASE)

The inner ear hearing and balance mechanisms resemble convoluted tubes of membranes immersed in fluid within the otic capsule that encloses the inner ear. The tubes are filled with endolymph, fluid of a consistency necessary for the functions of the inner ear. Outside the tubes, the structures are bathed in perilymph, which closely resembles the cerebrospinal fluid around the brain and which differs substantially from the endolymph.

To maintain its consistency, the endolymph is continually formed within the inner ear, then drained away via a small out-pocketing: the saccus endolymphaticus, which absorbs the fluid back into the bloodstream.

Pathogenesis

For reasons uncertain, the saccus drainage may fail. When this occurs, the membranous inner ear structure swells, and may rupture. The swelling produces episodes of typical symptoms.

As the cause of the problem is uncertain; it is unknown whether the problem is a unique condition or, alternatively, common to several causes (e.g. like a headache). This impacts on the ability to formulate and target treatment adequately.

Presentation

The condition is typified by episodic severe distress. Initially, Low frequency nerve deafness may be mild and fluctuant, becoming more severe with time and affecting all frequencies, possibly leading to total subtotal loss. Speech
discrimination becomes severely compromised. A feeling of fullness, pressure or blockage is felt in the affected ear. Humming or buzzing tinnitus may be severe, worse with the episodes. Severe rotatory vertigo may cause nausea, vomiting and prolonged prostration. Diarrhoea is not uncommon. Later, the condition may develop drop attacks – sudden collapse without warning and without loss of consciousness.

Investigation

Figure 15: Unilateral SND, suspicious for ELH. Few other conditions produce this pattern.

Pure tone audiology will demonstrate a unilateral SND, initially low frequency and possibly fluctuating, later pan-frequency and depressed to 60-70 db. ENG shows a vestibular paresis on the affected side. Nystagmus towards the affected ear may be present in the early irritative phase, later to the unaffected side with the onset of vestibular palsy.

Radiology shows no changes specific to the condition.

Management

Medical management may aim to treat the cause or relieve the symptoms. The former employs diuretics (Moduretic, urea) or vascular agents (betahistidine). Others settle dizziness or nausea (prochlorperazine, chlorpromazine, ondansetron).

Dietary reduction of salt intake appears to be beneficial, but as many foodstuffs have considerable covert salt content, a limited-salt diet is best directed by a specialist dietitian.

Several office procedures may be helpful. Grommet insertions have been used in the past, although with uncertain efficacy.

More commonly, middle ear perfusions are used to control symptoms. Filling the middle ear with steroid solutions is reported as being beneficial in earlier cases, although further verification is needed.

More often, gentamicin perfusions are used. Gentamicin is a common antibiotic with a vestibulotoxic action. If the middle ear is flooded with a gentamicin solution, some diffuses into the inner ear, reducing the vertigo produced by the ELH. Several perfusions may be needed, performed with little discomfort under local anaesthetic as an office procedure, usually employing a vent tube. The aim of the procedure is to produce mild unsteadiness (disequilibrium) that comes on in the days after the perfusion, and after which rotatory vertigo generally diminishes. Further perfusions are given if the vertigo persists. The technique is effective in over 80% of cases, although some require a “top-up” many months later.

The downside of perfusions is the degree of disequilibrium encountered. This balance upset is due to reduction of balance function in the ear
treated, and may be severe in some cases, especially in females, where the lighter bodily tissues presumably admit more of the drug into the inner ear. This more severe reaction may take 6-8 weeks to clear, and the patient needs to know this risk prior to treatment. Fortunately, severe reactions generally herald a complete cure of the rotatory vertigo.

A further complicating factor arises if the patient is unable to distinguish between this disequilibrium and another attack of true ELH: the surgeon may be uncertain whether to administer a further dose that may exacerbate matters.

Gentamicin may also damage the hearing. The effect of perfusions in this respect is uncertain, as active ELH also causes progressive deafness.

Surgery
Operative intervention may be employed in several roles.

i. Saccus Decompression/Drainage.
Removal of the mastoid bone and its air cells from over the saccus endolymphaticus relieves or cures ELH in up to 70% of cases. The surgery is controversial, as the exact beneficial action is uncertain. The efficacy of sac drainage or the insertion of micro-shunts undertaken during this surgery is also questionable. When successful, symptoms are alleviated and hearing stabilised, although cases may relapse later. This is often the treatment of choice in early cases, to retain hearing.

ii. Section of the Eighth Nerve.
Neurosurgical-type division of the nerve attempts to halt the dizziness without loss of hearing. The surgery is more invasive, requires advanced skills, and may be ineffective in salvaging hearing, as the ELH continues to damage this aspect in the ear; the nerve section does not halt this aspect.

iii. Labyrinthectomy.
Removal of the balance elements of the inner ear is effective in eliminating rotatory vertigo, and is done when the hearing is severely damaged, or when the symptoms dictate this course, usually prostration or drop attacks. This surgery will totally remove hearing, therefore concurrent CI or bone-conduction implantation may be considered at the same procedure.

Rehabilitation
a) Hearing
ELH or its management commonly result in severe deafness, with loss of speech discrimination ability, such that conventional hearing aids may be ineffective. Several hearing options are available.

i. CROS / BiCROS aids help to eliminate the “head shadow” dead zone of single sided deafness. These use a receiver on the deaf ear that transmits by Bluetooth to an aid in the better ear (which, in the BiCROS variant also aids the better ear).

ii. Active bone conduction devices (Med EL Bonebridge, Cochlear BAHA) act similarly to the CROS aid using skull vibration, and can be implanted at the initial surgery. They can be used only when the better ear has excellent hearing, and do not restore full audiological ability (direction finding, stereo, or the improved hearing with two ears — binaural summation).
They help greatly at a table, groups, committees etc.

The concern with the bone conduction implants is the possibility of later onset ELH in the contralateral ear, which would negate their effectiveness.

iii. Cochlear implants may also be effective. When the hearing is severely damaged, particularly in rapid onset cases or when contralateral disease is present, cochlear implantation of the worse ear is considered to provide a stable and effective means of communication. The sound quality from a cochlear device is not as clear as the bone conduction implants, but is independent of the other ear and immune to further ELH effects. They help greatly to avoid the socio-economic disruption of bilateral Meniere’s and, particularly in younger patients, are an increasingly employed management.

b) Balance

Gentamicin perfusion, VIII section and labyrinthectomy carry a significant risk of severe disequilibrium, as outlined above. This occurs when the brain has not compensated for the drop in function of the affected ear. In younger patients, recovery may be relatively rapid, but in the older case this is more protracted and long-term difficulties may persist. Expert balance rehabilitation physiotherapy is strongly advocated, and persistence with this management is desirable for best outcomes.

When disequilibrium is severe, however, one’s intellectual centres are used to help compensate. Distraction of these centres can significantly impede compensation; the patient should avoid a number of such situations:

Financial concern: avoid debt, excessive spending.

Friction: minimise family confrontations.

Fatigue: avoid physical over-exertion (e.g. over-zealous rehabilitation).

Movement: Reduce excessive multidirectional movements.

Decision-making: Constant demands for decision impact heavily on executives/managers.

Other worries, stresses: to be avoided

c) Benign Positional Vertigo (BPV):

Lastly, ELH cases may be prone to BPV, due to debris resulting from the disease. BPV occurs when particulate debris irritates balance receptors in the ear, causing rotatory vertigo in specific bodily positions or movements, including arising, looking up, rolling over in bed, or sudden horizontal movements. The problem is often remedied by specialized exercises (Epley manoeuvres) that reposition the offending particles away from the sensitive site.

ELH is a complex, damaging and often distressing condition. Fortunately treatment will terminate the vertigo, or this will burn out spontaneously in the long term, if untreated.
10. VESTIBULAR MIGRAINE

Whilst spinning type dizziness (rotatory vertigo) characteristically arises from the ear, the problem can also arise from CNS origins. Vestibular migraine arises from vascular spasm affecting the balance centres in the lower brain or the artery to the ear. The problem occurs in episodes and there may be a previous history of other migraine events with visual disturbance and ill health. The condition is thought to be triggered by a variety of influences: alcohol, hormonal, stress and fatigue. It seems more common in young women.

The episodes present as spontaneous rotatory vertigo, brief or more prolonged, with attendant nausea, vomiting and prostration. However there is no hearing loss and little tinnitus or other ear symptoms.

Investigations generally show little evidence of ear disease; the tests can only rarely be arranged at the time of relatively brief episodes.

Specific anti-migraine medication is appropriate. Preventative medication is taken daily, and relieving treatment (for pain and nausea) at the time. Efforts are made to avoid any known trigger factors, although these are notoriously difficult to identify.

The diagnosis is essentially based on the pattern of repetitive rotatory vertigo without lasting other ear symptoms and without significant findings on investigation.

11. CNS ISCHAEMIA

General unsteadiness, usually with ataxia, loss of proprioception or incoordination is a common problem of the elderly. It is often confused with ear disease, and these conditions may coexist, hearing loss and tinnitus in the aged being commonplace.

![Figure 16: Advanced CNS ischaemic changes. Imbalance and deafness likely.](image)

Chronic ischaemia manifests as diffuse CNS changes that are very commonly noted on MRI scans when these are undertaken to diagnose unsteadiness. The problems occur with a wide variety of neurologic manifestations, according to the site, severity and extent, being best assessed under a specialist neurologist.

Acute ischaemia may result from cervical spondylosis pinching the vertebral arterial supply. In the past, this was commonly confused with BPV, but the latter has no syncopal effects.
12. ACOUSTIC
NEUROMA/SCHWANNOMA

Pathology

Acoustic nerve tumours arise from the balance section of the nerve and are derived from the Schwann cells that comprise the insulation elements of the nerve. They are neither common nor rare; specialised clinics encounter a steady caseload. The tumours are found in the IAC, within the cerebello-pontine angel, or spread between these two areas. They are usually in middle-aged or older individuals. Most are very slow growing; in the elderly there may be little if any growth. The tumours are benign, but the actively growing examples may expand, attaching to the lower brain, then compressing this tissue, producing major balance upsets. These tumours may be present in certain diseases (Neurofibromatosis 1, von Recklinghausen's disease).

Figure 17: Neurofibromatosis. Multiple skin lesions. Café-au-lait spots may also occur.

Asymmetrical nerve deafness of this nature is a warning sign familiar to all audiologists: such cases are strongly advised to access an ontological opinion.

Figure 18: Unilateral sensorineural deafness in an adult strongly indicates further investigation. An MRI scan is optimal; CT scans are not reliable enough to detect small lesions.

The deafness is accompanied by tinnitus (humming or ringing) and slight unsteadiness on movement. As the disease progresses the unsteadiness becomes more severe, with possible facial paralysis, headache dysdiadokokinesis and ataxia.

Investigation

MRI scans are primary investigations when the symptoms suggest the presence of these tumours. They are also used to track any evidence of growth in the smaller examples, to indicate surgery before the lesions expand excessively.

Presentation

Growth of these tumours is subtle, often unnoticed for some time. The slow compression of the acoustic nerve leads to three main symptoms. Gradual high frequency nerve deafness is typical.
Audiology shows unilateral deafness, as above, that is often complicated by poor speech discrimination. Reflex decay may also be present. ENG may show a vestibular paresis.

Management

With small tumours, especially in the aged, a wait and see approach is optimal, with serial MRI scans undertaken to assess any growth rate. When this occurs the choice is specialised radiotherapy or surgery. The latter is required for larger lesions, the approach being dictated by the pattern of disease present. Although care is taken, the facial nerve is commonly fixed to the tumour and may be damaged, causing facial weakness. Hearing is lost during surgery in the majority of cases. Large tumours that are attached to the brainstem itself are particularly difficult to remove, parasitising the vascular supply, hence the need for early detection.

Because of the reduced speech discrimination, hearing aids before or after surgery may not be effective. A CROS aid may be helpful. If the hearing in the non-affected ear is excellent, an active bone conduction implant (Bonebridge, BAHA) may effectively abolish the “head shadow” on the affected side by stimulating the better ear via skull vibration.

The important point of these conditions is the need to investigate unilateral nerve deafness, to avoid missing small tumours that may be removed with minimal complications, before these enlarge into more difficult situations.

13. GLOMUS TUMOURS

Pathology

Uncommonly tumours derived from the neural crest are found in the middle ear, perhaps extending into the intracranial space. These vascular glomus lesions are paragangliomas or chemodectomas, with a low malignancy rate of 3-5%.

Glomus tympanicum lesions arise from the promontory or hypotympanum. Glomus jugulare tumours arise off the jugular bulb and are often extensive, highly vascular and difficult to remove.

The tumours present as a red globule or mass, seen via the drum. Conductive deafness may be present. VII-XII cranial nerve involvement may be present.
Figure 20: Glomus tumour. A red blush overlying the promontory indicates the presence of a small chemodectoma.

Radiology may demonstrate a vascular blush with considerable vascular contribution from surrounding vessels, possibly with erosion of the skull base.

Management

Glomus tympanicum lesions are removed via a tympanotomy approach, lifting the drum and removing the tumour intact or piecemeal. The jugulare tumours require more aggressive skull base approaches, with pre-operative tumour embolism and/or vascular ligation to reduce haemorrhage, and possible neurosurgical involvement. The outlook is naturally more guarded.