CHAPTER 11.

INNER EAR SYMPTOMS

INNER EAR DISEASE

Cardinal Symptoms

a) Balance disorders
b) Tinnitus
c) Sensorineural Deafness

The ear is an organ of both hearing and inner ear disease damages these functions, causing the cardinal symptoms of tinnitus, sensorineural deafness, and imbalance due to vertigo or disequilibrium. Because of the proximity of the cochlear and the vestibular apparatus, disease processes commonly cause combinations of the above symptoms, although purely auditory or vestibular aberrations are not uncommon. Thorough and structured interrogation will give a clear indication of the diagnosis in many cases. Note, however, that other diseases can closely mimic true ear disease, particularly those of the CNS or VIIIth nerve.

1. IMBALANCE

Disturbances of balance are a common event, often troubling and commonly associated with other health problems. Analysing the problem requires an exact understanding of the symptoms of imbalance plus any attendant events.

Balance is derived from functions of the ear, the nerve connections to the brain, and the brain itself. Other supporting mechanisms include eyesight and proprioception (the ability to judge the position of the body's skeletal structures-joints, ligaments).

Imbalance or dizziness can arise from derangement of any of these components; therefore treatment depends on careful identification of the malfunction.

Manifestations

a) **Syncope**: Common fainting or sensations of about to “pass out”.
b) **Vertigo**: A sensation of spinning oneself, or the world spinning around.
c) **Dysequilibrium**: Unsteadiness on moving the head, or when moving around.
d) **Ataxia**: Unsteadiness on the feet due to muscular incoordination.
e) **Drop Attacks**: sudden collapse but without losing consciousness. Due to Meniere’s disease.
f) **Confusional States**: Impaired clarity of perception due to trauma, Intoxication, etc.
g) **Psychological**: Impaired thought patterns, neurosis etc.
1.1. HISTORY

Interrogation plays a critical part in deciphering unsteadiness. An accurate history, following the usual clinical pathways gives the clinician the vital pattern of events. Obtaining this clinical picture may require some doggedness to clarify matters. Patient digression is common and often persistent.

a) Nature
This is perhaps the most important question: exactly what is the patient experiencing when the balance is disturbed? With true organic disease, a tangible pattern of events usually emerges, such as brief rotatory vertigo induced by specific position changes (benign positional vertigo). Careful questioning plus some patience usually elucidates one of the above. If symptoms are atypical and the patient elusive, begin to suspect psychological elements.

b) Onset and Initiation
Clarify when the problem began, how it happened and whether there was an apparent cause. If the patient had a chronically weeping ear, then became dizzy upon pressing the site, a fistula might be suspected.

c) Duration
The duration of the unsteadiness over time, and the length of any episodes is relevant. Long term unsteadiness when walking after a head injury may indicate ataxia. Prolonged episodes of severe rotatory vertigo suggest active endolymphatic hydrops (ELH) rather than the brief drop attacks that may complicate the same disease in its later phases.

d) Severity
Subtle disequilibrium induced by the gradual growth of an VIIIth nerve tumour differs from the severe example produced by rupture of the otic capsule from a transverse temporal bone fracture.

e) Frequency
Repetitive syncopal episodes in a fair maiden differ sharply from a single confusional state induced by a “spiked” drink.

f) Aggravating/Relieving factors
Dyssequilibrium is characteristically exacerbated by sudden movement of the head. Syncope is relieved by laying the patient down.

g) Associated Factors
The other cardinal features of ear disease are highly important. Vestibular migraine may mimic ELH with repetitive rotatory vertigo, but does not produce the characteristic low frequency SND of the latter. The severe disequilibrium of vestibular
neuronitis does not incur the concurrent deafness and tinnitus of the similarly dizzy labyrinthitis sufferer. Likewise, the major CNS symptoms (headache, ataxia, fits or syncope) may discern intracranial problems from the otological.

1.2. EXAMINATION

Particular points are noted.

a) Otological

The drums are inspected and hearing assessed. Tuning forks test for SND. Hallpike manoeuvres and Unterberger tests performed.

b) CNS

A full CNS examination is frequently desirable (check for treponema), with emphasis on cerebellar testing and also V, VII, IX, X, and XI (because of their relationship to the IAC).

1.3. INVESTIGATIONS

a) Audiology

Pure Tone Audiology (PTA) is essential in most cases of possible otological origin. Classical patterns include the low frequency SND deafness of ELH, or the characteristic 4000cps SND dip of noise trauma (which may be relevant in Benign Positional Vertigo).

Figure 2: Noise trauma. Possible BPV?

i. Asymmetrical HFSND is suspicious for VIII\textsuperscript{th} lesions. SSC dehiscence produces characteristic suprathreshold sensorineural responses.

ii. Reflex assessment: The stapedius and tensor tympani reflexes tend to decay in VIII nerve lesions, readily detected on tympanometry.

iii. Auditory Brainstem Response (ABR) testing notes aberrant results with retrocochlear disease.

iv. Electrocochleography (ECoG) assesses the activity of the cochlear hair cells and produces characteristic changes in ELH.

v. Speech Discrimination ability falls away in retrocochlear disease, advanced ELH and other severe cochlear disease.

vi. Electronystagmography (ENG,VNG) assesses a range of balance aberrations, related to vestibular weakness, cerebellar or other CNS abnormalities.
vii. VEMP (vestibular evoked myogenic potential) testing may indicate a saccular disorder or conditions such as dehiscent superior semicircular canal.

viii. Radiology (CT and/or MRI scans) is frequently employed to detect or exclude CNS pathology, VIII lesions and other retrocochlear problems. CT may also identify chronic ear pathology (labyrinthine fistula) or other problems such as SSCC dehiscence.

ix. Pathology: Serological assessments for Treponema, autoimmune responses and hormonal aberrations are undertaken if indicated.

x. Electroencephalography (EEG) is indicated in some neurological conditions.

Differential Diagnosis of Vertigo and Dysequilibrium

a) Labyrinthine
   a) Labyrinthitis
      • Viral
      • Bacterial
      • Treponema
   b) Trauma
      • BPV
      • Head Injury
      • Post-Stapedectomy
   c) Ototoxins
      • Aminoglycosides
   d) Ischaemia
      • Embolism
      • Thrombosis

b) VIIIth Nerve
   • Vestibular Neuritis
   • Head Injury
   • Acoustic Schwannoma
   • Cerebello-pontine Angle Tumours

c) CNS Disease
   • Vertebro-Basilar Insufficiency
   • Vestibular Migraine
   • CNS Tumours
   • Other CNS disease
   • Treponema, MS, Epilepsy

d) Others
   • Psychogenic
   • Confusional
   • Non-organic
2. TINNITUS

Tinnitus is defined as sound heard in the ears without external sound stimulus. The majority of cases presenting for this symptom are electronic, harmonious, or pulsatile sounds, but others (arising from the EAC or middle ear) may be irregular, staccato or coarse, according to the origins.

There are no known methods of accurate measurement of inner ear tinnitus.

2.1. CLASSIFICATION

Tinnitus may be innate (physiological), acquired, or of psychological origins. Innate tinnitus is evidently heard by many otherwise normal individuals, taking the sensation of a steely monotone, mosquitoes or cicada sound, usually faint and non-irritant, noticed mainly in silent conditions such as the evening, or when listened for in noisier surrounds. This pattern is not accompanied by other audible or otological symptoms, is usually steady in nature and represents ambient neural activity. Another evident pattern presents as bursts of sound in one or the other ear, variable in frequency or perceived volume, and lasting 10-20 seconds before fading. This may be accompanied by a sensation of fullness or pressure that rapidly abates.

Many people also note occasional muffled and irregular rustling/flutter/soft vibration in one ear, also without other symptomatology. This is thought to be minor muscular twitching/spasm of the stapedius or tensor tympani muscles that are attached to the ossicular chain, a phenomenon similar to the legendary “Messerchmitt twitch” of Hollywood fame. This symptom may be triggered by specific volumes, or frequency, perhaps associated with general fatigue.

Other normal bodily sounds from nearby structures, the airway or alimentary tract are not normally classified as tinnitus.

Acquired tinnitus is derived from pathological events and is accompanied by hearing loss, being derived from origins that cause loss or malfunction of normal hearing. Few authorities accept the presence of tinnitus without objective evidence of hearing loss. Acquired patterns may be unilateral or bilateral according to the cause. For example, chronic noise trauma generally causes bilateral tinnitus due to relatively even exposure by both ears over time. Conversely, blast injury may result in severe unilateral tinnitus in the ear closest the origin of the trauma.

Two unusual variants of acquired tinnitus may present. Objective tinnitus may be heard by a nearby observer, being often clicking in nature, and due to palatal muscular twitching causing either palatal or Eustachian related clicks. Pulsatile tinnitus arises from a variety of local vascular pathological phenomena. Some of these may also be audible, e.g. an arterio-venous fistula.
Psychological tinnitus is more problematic. General stress, depression, inadequacy or similar chronic psychological states are commonly associated with perceived greater tinnitus problems, but the latter is difficult to discern from the accompanying problems, and also the commonly associated intake of nicotine, caffeine, alcohol or other tinnitus-enhancing chemicals.

2.3. AETIOLOGY

The common monotonal physiological/innate pattern is of uncertain cause. Neuronal cells maintain low levels of ambient activity, evidently to maintain function, and this is a likely cause. Other speculations include vibration of the cochlear hair cells and the resultant otoacoustic emissions. Innate patterns are commonly enhanced by incidents that cause conductive deafness and hence exclusion of everyday sound from the cochlea that otherwise mask this innate sound.

Acquired patterns result from a wealth of factors that may damage the inner ear or its neural connections. These include noise, age, viruses, and tumours of the acoustic nerve. Listing all of these is not feasible in this context, but the results will be unilateral or bilateral according to the nature of the agent.

2.4. AGGRAVANTS

Whilst many pharmacological products warn of possible tinnitus side effects, in the absence of demonstrable hearing loss, this aspect remains uncertain. However, some agents seem likely stimulants, including nicotine, caffeine, alcohol, higher dose salicylates, plus hypertension, straining/exertion, bending and other posture, and possibly physical fatigue. Psychological stress appears to reduce tolerance.

2.5. ASSOCIATED SYMPTOMS

Acquired tinnitus is frequently attended by the primary ear symptoms: pain, discharge, deafness/blockage or dizziness. These, or their absence, generally indicate the nature of the problem. Other symptoms include those of the central nervous system (CNS) (e.g. headache, unsteadiness) or general health.
2.6 ASSESSMENT/INVESTIGATION

In the absence of other compelling symptoms and signs, investigations centre on the ear and the CNS. Audiology generally indicates the origin, but CT or MRI scans are often required for evaluation of the ear and related structures. The absence of concrete findings strongly indicates an innate or psychological possibility. Whilst objective or pulsatile tinnitus frequently occur without related audiological depression, pathological tinnitus, being due to inner ear damage, is invariably accompanied by hearing loss, unless the process is in its earliest phase, e.g. developing hydrops.

2.7 TREATMENT

At this time there are no known medical or surgical treatments that abolish inner ear origin tinnitus. Some objective or pulsatile varieties may respond to elimination of their original pathology. Research into inner ear remedies is hindered by the difficulty assessing the true effect of a medication, as the symptom cannot be measured. Alternative treatments are therefore needed.

Distraction methods using radio, music or other pleasant sound is often used. A radio alarm clock with a “sleep” feature, activated after a given time, is helpful in the quiet of the night.

Medications for sleep or to reduce irritation may be beneficial.

Professional audiological services offer a range of managements including tinnitus retraining, masking, Neuromonics (complex stimulation at above-audible frequencies) and other measures. Enquire re costs before engaging.

For practical measures, Tinnitus Society websites (preferably UK, Australian) offer a range of helpful hints.

Lastly, tinnitus can cause severe ongoing stress, particularly unilateral trauma or infection related types. Seeking professional psychological management is advisable when chronically distressed.

2.8 PROGNOSIS

Fortunately, in a great number of cases the problem appears to regress with time, albeit with concurrent adjustment by the sufferer.

Figure 5: Tinnitus from severe or traumatic inner ear origins may be distressing.
3. SENSORINEURAL DEAFNESS

Sensorineural, nerve, or perceptive deafness results from damage to the cochlea or the cochlear division of the auditory (VIIIth) nerve. The losses may be congenital or acquired, unilateral or bilateral, stable, fluctuant or progressive.

Most have a high frequency component; a few are low frequency. Stable losses generally result from “one-off” causes, e.g. trauma or vascular occlusion. Fluctuant or deteriorating losses are concerning, as these originate from active pathology that may cause further and permanent deafness if not managed promptly.

3.1 History

Interrogation should follow the structured lines as recommended for vertigo, above. The onset and perceived cause may naturally lead to the diagnosis in many cases, such as deafness in one ear after a blow to the site.

The duration of the loss may also be indicative, e.g. a long standing bilateral loss in the elderly is typical of presbascusis.

Severe losses are frequently easier to diagnose, as the disability produced is obvious and may be readily attributed to a cause. A profound unilateral loss in a well individual suggests a thrombotic or embolic origin.

Concurrently bilateral losses suggest a generalised nature and perhaps cause, whereas a unilateral loss may indicate other aetiological options.

Episodic or frequent losses infer a repetitive origin, probably intermittently active disease, e.g. ELH

Aggravating/relieving factors may be noted; deteriorating losses after aminoglycoside therapy suggests ototoxicity effects.

Associated cardinal ear symptoms or CNS features or other aspects (age, medical history, recent events) are also frequent indicators of a cause.

3.2. EXAMINATION

The external canal is cleared and the drum inspected for disease. In most cases any abnormalities will be coincidental, excepting less common causes such as cholesteatoma or evident trauma.

In unilateral SND the weber will refer to the better ear, but the Rinne will be positive, Beware false negative Rinne responses heard in the better ear (mask this side with rustling paper if necessary). Bilateral cases will demonstrate a central Weber if the losses are similar, and bilateral positive Rinne responses.

Assess the CNS according to the case, with emphasis on balance and coordination if these are suspect.
These follow the assessments for balance, without the specific balance assessments, unless indicated.

Audiology

Pure tone testing is the cardinal inner ear test. This will reveal not only the severity and pattern of the loss, whether high, low or pan-frequency, but may also suggest a diagnosis or concurrent conditions.

The severity of losses may suggest the rate of progression of the condition, e.g. advanced presbyacusis in a middle-aged patient.

The pattern of the PTA is suggestive. Noise trauma delivers a classic high frequency loss, maximal at 4000 cps. ELH is characterised by a low frequency, perhaps fluctuating loss.

Comparison with the other ear is also valuable. Age (presbyacusis) produces bilateral symmetrical high frequency losses, but if these are asymmetrical, the possibility of other pathology such as VIIth nerve lesions must be considered.

Noise trauma patterns that are worse in the left ear suggest rifle fire, if there is a history of shooting, as the muzzle blast is closer to the left ear (in right-handed cases).

Other audiological tests may discern between cochlear and retrocochlear disease. In deaf children, otoacoustic emissions (OAEs) and auditory brainstem testing (ABR) are used to differentiate between dyssynchrony and neuropathy. ABR assessments are also useful to detect retrocochlear lesions.

Radiology

Radiology plays a major part in audiological diagnosis. MRI scans are undertaken routinely for asymmetrical SND, to check for retrocochlear disease, and also some less common labyrinthine conditions. CT scans assess the advance of cholesteatoma, check for superior semicircular canal dehiscence, large centricular aqueducts (in children) amongst other threats.

Figure 6: Classic bilateral symmetrical SND in an older patient: presbyacusis.

Figure 7: Large ventricular aqueduct, causing progressive SND in a child.
Differential Diagnosis of SND

a) Labyrinthine
- Presbyacusis
- Labyrinthitis
- Bacterial (AOM, cholesteatoma).
- Ototoxicity
- Trauma (includes perilymph fistula)
- Vascular occlusion/embolism
- Endolymphatic hydrops
- Auto-immune disease

b) VIIIth Nerve
- Trauma
- Meningitis
- Schwannoma, meningiomas
- Disseminated sclerosis

c) CNS Disease

d) Non-organic