Handbook of ENT

Diseases and Disorders of the Ear, Nose, and Throat

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Acknowledgements

The following comment was passed on to me while I was preparing this textbook: Isaac Newton, when complimented on his achievements, noted that they had only been possible because he was 'standing on the shoulders of giants'. This was particularly apt because, on reading my manuscript, I realized that I had incorporated ideas, words, and phrases from many 'giants' in this specialty which had, consciously and subconsciously, become part of my philosophy and approach. I apologize for the lack of specific references to these sources and trut that anyone reading this book who feels that what has been incorporated originated from them, will accept that this book is an attempt to build a framework of basic knowledge and skills that we, as teachers, need to pass on to future generations of clinicians in the same way that it was passed on to us, in order that their practice of our speciality is of an acceptable standard. There are, however, two people whose contributions require acknowledgement. The first is John Duff, my predecessor in charge of organizing our undergraduate ENT course at the University of Cape Town Medical School, whose notes form the basis for the chapters on anatomy and physiology. The second is Marie-Louise du Plessis, who prepared the original line drawings used to complement the text.

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Introduction

Disorders of the ears, nose, and throat are commonly encountered in primary health care. It is estimated that between 20 and 30 per cent of patients seen in a typical practice present with these disorders. Some examples will serve to support this estimate:

- Almost all patients in a practice will have the occasional cold, which will frequently be followed by a period of purulent nasal discharge. Sometimes this will precipitate an attack of sinusitis.

- More than half of the children in any population will have an episode of acute otitis media before they start school - the peak incidence is in the one- to three-year age group - and many of these children will have had more than one episode by this age.

- We all produce wax in our ears. Some patients become preoccupied with removing this wax and persistently fiddle in their ears. At some stage, such fiddling either impacts the

wax in the ear canal or precipitates an episode of otitis externa, particularly in warm, moist climates or in patients who often go swimming.

- Deafness is a particularly disabling problem, and hearing loss is one of the inevitable accompaniments of ageing that adds to the burdens of these 'golden' years. Recognition of the problem and direction towards assistance, usually in the form of hearing aids, can do much to ease the problem. Age is not the only cause of deafness, and, given some knowledge of the causes such as rubella in pregnancy and noise in industry, many are potentially preventable.

Counselling in this regard is an essential part of the work of a primary care physician.

- Smokers invariably ignore health warnings and some smokers in any practice are likely to develop cancer. Although not as common as cancer in the lung, cancer in the upper aerodigestive tract is sometimes encountered, particularly involving the tongue and the larynx. Early recognition may be life-saving. Treated Stage 1 laryngeal cancer patients have a 90 per cent five-year survival rate, whereas when the cancer has progressed to Stage 4, the survival rate falls below 50 per cent.

Despite all these factors, in most undergraduate medical courses, ENT is regarded as a minor speciality and is squeezed into one of the busy clinical years as a short course. For ENT tutors this poses the problem (and responsibility!) of how, in the short time available, to equip medical students with sufficient essential information and skills to enable them to recognize ENT disorders and to manage patients presenting with these disorders once they have graduated and have their own practices.

Aims

Tertiary education theory maintains that the first step to achieve this goal is to establish aims for the course.

Most students will have forgotten much of the anatomy and physiology they so painfully acquired in the early years of the medical course, so the first aim will be:

- To refresh knowledge of essential ENT anatomy and physiology so that the 'normal' can be recognized.

Eliciting essential signs in this area of multiple body orifices requires special examination skills. In the short time available, these skills can be acquired only if they are taught in a rigid, structured manner. Therefore the second aim will be:

- To teach a structured approach to examination of the ear, the nose, the throat, and the head and neck region.

The range of disorders encountered in ENT specialist practice is vast. Although many of these disorders require specialized knowledge and sophisticated diagnostic and management skills, most of them are common in primary care practice and can be adequately managed in this setting.

Therefore the third aim will be:

- To present students with an essential core knowledge of ENT disorders.

Patients, however, do not present with disorders but with symptoms and signs which must be elicited to make the diagnosis of a disorder. Therefore the fourth aim will be:

- To present a practical approach to ENT disorders.

Some patients will present with disorders that will not be within the management competence of a primary care physician. Early recognition of those disorders requiring referral to a specialist service may be vital to the ultimate prognosis.

Therefore the fifth aim will be:

- To establish guidelines for specialist referral in ENT disorders. In this text, situations calling for referral are flagged with this symbol: *

Patients have the right to expect that their primary care physician is competent to care for them. Therefore the sixth aim will be:

- To assess the competence of students regarding their core knowledge, examination skill, and ability to diagnose and manage common ENT disorders.

The community requires the services of physicians with above average knowledge and skills for the management of specialized ENT disorders. Therefore the final aim will be:

- To stimulate in some students the desire to further pursue ENT through electives, research projects, further training, or specialist training.

Objectives

Education theory further states that aims are achieved by setting objectives. Ideally, these objectives should be formulated in such a way that they act as stepping stones along the way to achieving each aim. However, in practice, most ENT departments have a busy clinical workload and are understaffed for teaching. For this reason it is almost impossible to set out objectives in a logical sequence when the theoretical teaching sessions have to be interspersed with clinical exposure. Whereas theory sessions can conveniently divide the speciality into its components - ear, nose, throat, head and neck - in the clinical sessions patients are going to present with disorders not yet covered by the theory. In practice, therefore, objectives have to be stated as set out below:

By the end of the ENT course students will:

- Have refereshed their knowledge of the essential anatomy and physiology of the ear, the nose, the throat, and the head and neck.

- Be aware of symptoms resulting from disorders of the ear, the nose, the throat, and the head and neck.

- Be able to examine the ear, the nose, the throat, and the head and neck; differentiate normal from abnormal; and recognize the nature of any abnormality.

- Be able to relate these symptoms and examination findings to an essential core knowledge of disorders of the ear, the nose, the throat, and the head and neck.

- Be able to initiate management for these disorders, and to set in motion the following:

- procedures required to examine the area further
- investigations required to confirm the diagnosis
- primary care treatment of the disorder
- follow-up care for the disorder
- referral of patients when their disorders require specialist attention
- undertaking of interim care while awaiting referral.

Assessmnet

According to education theory, these objectives should estabish the criteria for assessment. To be effective, assessment should be 'formative', that is, undertaken throughout the course to afford opportunities for the correction of deficiencies. However, for the reasons stated above, this is often impractical in busy ENT departments so that, for the most part, all that will be offered is 'summative' assessment, that is, assessment undertaken at the end of the course when it is usually too late to correct deficiencies except by repeating the course - the ever-present threat of failing the ENT examination!

An attempt has been made in this textbook to condense the knowledge, which an ENT surgeon will take several years of training and practice to acquire, into an essential core knowledge of theory and skills that can be imparted in a brief ENT undergraduate course to equip students as future medical graduateswith sufficient basic ENT information to enable them to manage these disorders when encountered in primary care. It is strongly recommended that students add to the information contained in this book by referring to the many ENT textbooks available either in medical libraries or in medical bookshops in order to acquire a broader understanding of the different approaches to and ways of treating ENT disorders.

Students who are contemplating the purchase of another textbook to supplement this core knowledge manual should be aware of two considerations, apart from cost, that should influence their choice. The first consideration is the way they prefer to learn from a textbook. ENT books vary in both their style of presentation and in the amount of illustration. Some adopt the traditional style of presenting theoretical discussion of disorders by regions. Others adopt a more practical, symptom-oriented approach. Most fall somewhere between these and vary in the amount and quality of illustration. The second consideration is that some of these textbooks will really only be useful to undergraduates, whereas others will act as a reference

source that will sustain them in primary care practice. Students would be best advised to consider these two aspects and then browse through what is available in the library before making their choice.

The 'ENT Ten'

If you learn nothing else from your ENT course, you should try to memorize these ten points and keep them in your memory bank for the rest of your career.

1. If a mother says her baby is deaf she is usually right.

2. A foul or unilateral nasal discharge in a child is usually due to a foreign body. In an adult suspect tumour.

3. An inflamed, swollen cheek is usually due to dental infection.

4. A painful, swollen eye may be caused by frontal or ethmoidal sinusitis. When this occurs, vision is at risk and there is potential for intracranial spread of infection.

5. Check for ear disease in all cases of meningitis and facial nerve palsy. When ear disease causes facial palsy, exclude TB.

6. If hoarseness persists for more than a month, entertain a suspicion of a tumour (cancer in smokers!) and refer urgently for a diagnosis to be made.

7. Pain referred to one ear is a sinister symptom in anyone over forty with a sore throat. Suspect malignancy!

8. Any neck swelling needs careful ENT evaluation prior to any invasive procedure.

9. Deafness in the elderly can be helped. Refer these patients for assessment.

10. People wanting to poke anything into their ears should use their elbow! Use syringing to remove wax, foreign bodies, and purulent discharges.

Part I

The ear

1. Structure and function of the ear

ENT surgeons describe the ear in three parts: the external, the middle, and the inner ear.

The external ear

The external ear consists of the pinna (auricle), the external auditory canal (external ear canal or simply ear canal), and the tympanic membrane (eardrum).

The pinna

Referring to Figure 1.1, see if you can recognize all the different parts of this structure on a colleague. Names to note are the helix, the antihelix with its two superior crura, the tragus, the lobule, and the conchal bowl which leads into the ear canal.

In the embryological stage, the pinna forms from fusion of a collection of little humps, or hillocks, on the side of the head of the developing foetus. A fairly common congenital abnormality is the pre-auricular pit which arises because of incomplete fusion between two of these humps, usually between the helix and the tragus. It can lead into a multiloculated sac which is often a site for infection and abscess formation. Uncommon, grosser abnormalities arise from failure of development or fusion of the humps to form abnormal, small ear remnants, known as microtia.

The ear canal

The ear canal, for clinical purposes, is considered to have two portions; see Figure 1.1. The first is the outer third, or cartilaginous, portion which is lined with hair-bearing skin containing modified sweat glands - the ceruminous glands. These glands produce cerumen, the major component of ear wax. This is a friendly, touchable part of the ear canal and you may insert a speculum without having the patient jump off the chair. The second is the inner, medial two-thirds, or bony, portion which angles downwards and forwards towards the eardrum. This portion is totally different and the skin is hairless, thin, shiny, and exquisitely sensitive to even light touch - treat it with greatest respect. Because of the angulation, in order to see the eardrum you must pull the pinna a bit upwards and a bit backwards to straighten up the canal.

The ear canal is lined by skin, and as you probably know, skin continually sheds its surface layer of dead skin cells, a process known as desquamation. There has to be a mechanism for clearing all this desquamated debris out of the ear canal. This is achieved by having a pattern of migration from the centre of the eardrum, outwards to its margins, and then up and out of the ear canal. Any desquamated epithelium, debris, particles, hairs, or wax thus naturally migrate out of the ear canal to be shed into the conchal bowl. Interference with this mechanism, such as cotton bud usage, leads to accumulation and the possibility of having to have the ear canal syringed to get rid of the wax.

The tympanic membrane

Almost all of the tympanic membrane, or eardrum (see Figure 1.2), is made up of a slightly concave, translucent membrane. This larger portion of the membrane is the pars tensa and its margins are well demarcated from the canal walls. It has a smooth, squamous epithelial surface that tends to reflect the light of an otoscope - the light reflex (of no clinical significance). Embedded in the pars tensa is the handle of the malleus (one of the ossicles - see below) and superiorly this has a small, outward-projecting bump - the lateral process. Superiorly to the lateral process there is a small area of the drum which blends into the texture of the bony canal skin without any clear demarcation. This area is the pars flaccida (important to ENT surgeons because sinister ear disease may arise here, for example, cholesteatoma; see Chapter 4).

Sound waves (energy) are collected and channelled by the pinna into the ear canal. When it is quiet, try cupping your hands around your ears to increase the size of the pinna and notice the increase in amplification of sounds. The canal has a resonating frequency which selectively amplifies sounds of higher frequencies. These collected sound waves set the drum vibrating and, since the handle of the malleus is embedded in the drum, vibrations are transmitted to the ossicular chain of the middle ear.

The middle ear

The complete ear cleft (see Figure 1.3) comprises the **eustachian** tube, the middle ear cavity, and the mastoid air cell complex. The middle ear cavity is sometimes called the tympanic cavity - anything beginning tymp- refers to the middle ear, anything beginning myring- refers to the eardrum. The eustachian tube opens into the anterior wall of the middle ear cavity and is only open intermittently to the nasopharynx. On the posterior wall of the middle ear is an opening which leads into the **mastoid air cell complex** within the mastoid portion of the temporal bone. The entire middle ear cleft is lined by respiratory mucosa. For the most part, cilia are absent so that mucus transport towards and down the eustachian tube is relatively inefficient. Fortunately, in the normal middle ear there are only scanty mucus-secreting glands so that mucus accumulation is not a problem. The middle ear is a complex cavity that is larger than the drum, which forms only the lower two-thirds of its lateral wall. The area of the middle ear cavity above the pars flaccida is known as the attic.

The middle ear contains two elements vital to the conduction of sound to the inner ear - an ossicular chain and clean, moist, warm air (see below).

The **ossicular chain** consists of the malleus, incus, and stapes. These names are derived from Latin, and mean hammer, anvil, and stirrup. The handle of the malleus is embedded in the drum and its head lies up in the attic where it articulates with the incus. The incus has a long process that angles downwards to articulate with the head of the stapes. The stapes enters a recess in the medial wall of the middle ear - the oval window. Here its footplate has a mobile, circumferential joint that closes off the oval window. The inner surface of the footplate is in contact with the fluid of the inner ear. Thus vibrations along the ossicular chain are transmitted into the inner ear for processing.

What goes in must come out, and to accommodate the pressure changes created by vibration of the foot plate there is another mobile window - the round window. This is closed only by fibrous tissue and mucosa and is located on the medial wall of the middle ear just below the oval window. The two windows are separated by a bony bump, the promontory, which houses the basal turn of the coiled cochlea of the inner ear (see below).

Just above the oval window there is another bony hump and this covers the canal in which lies the facial nerve. The facial nerve originates in the brain stem, enters the internal auditory canal, passes over the cochlea, then angles backwards in the middle ear towards the mastoid where it angles inferiorly towards the stylomastoid foramen. Through the foramen it exists from the temporal bone to enter the parotid gland.

Relations of the middle ear

This complex middle ear cleft has some highly significant anatomical relationships (see Figure 1.4). Their practical importance is that spread of infection is potentially fatal.

Medial to the eustachian tube lies the internal carotid artery. Superior to the middle ear and mastoid is the middle cranial fossa containing the temporal lobe of the cortex (cerebrum). Posterior to the mastoid is the posterior cranial fossa containing the cerebellum, separated from the mastoid by the sigmoid sinus. Medial to the middle ear is the inner ear (labyrinth) separated from it by the facial nerve.

Sound conduction

Why do we need a middle ear when we have an external ear that directs sound wave energy down the ear canal? If you were to stand at the edge of a pool and shout at a friend swimming below the surface, the message would not be heard since the sound waves bounce off the surface interface, and most of the sound energy is then reflected. In a similar manner, if the sound wave were to impinge directly onto the oval window, most of its energy would be reflected and very little would be transmitted into the inner ear - a situation that is seen clinically when middle ear disease causes ossicular chain discontinuity with a resulting conduction hearing loss of about 50 decibels. To overcome this problem there has to be some mechanism for amplifying the sound wave energy sufficiently to overcome reflection and ensure transmission - this is what the middle ear accomplishes. Sound waves impinge on the tympanic membrane, in which the handle of the malleus is embedded, setting in motion vibrations which are faithfully transmitted along the ossicular chain to the stapes footplate. The tympanic membrane is some seventeen times larger than the stapes footplate, giving an energy amplification of about 17:1. The ossicular chain has an additional lever effect of about 1.3:1. The two together provide a transference enhancement of something like 22:1, sufficient to ensure transmission of the sound wave in its original form into the inner ear where it then travels freeely in the cochlear fluids for a distance entirely dependent upon its frequency (this is described later). (See Figure 1.5.)

Two terms that you will need to know in connection with sound wave energy are frequency (pitch), measured in hertz (Hz, cycles per second), and intensity, measured in decibels (dB).

The efficiency of this sound conduction mechanism is dependent on the free vibration of the eardrum and the mobility of the ossicles. It is essential, therefore, that the external and middle ears both contain air. Easy for the outer ear; it is in direct communication with the atmosphere, but the middle ear is a closed system open only to the outside at the whim of the eustachian tube. Air in any closed body cavity is continually absorbed and air in the middle ear is no exception. The eustachian tube is regularly opened by contraction of the palatal muscles, particularly during the act of swallowing, and this allows air to be replaced, consequently equalizing pressure across the tympanic membrane to permit free vibration. Aeration failure from eustachian tube dysfunction not only results in a negative middle ear pressure, but promotes increased production and poor clearance of mucus. The presence of mucus (middle ear effusion) damps down the vibration capability of both the drum and the ossicles.

The inner ear

The inner ear has two functions - hearing and balance. The parts of the ear responsible for these functions - the cochlea for hearing and the semicircular canals, utricle, and saccule for balance - are housed in a labyrinth of fluid-filled tunnels in the dense petrous portion of the temporal bone.

The hearing function

The **cochlea** derives its name from its shape - it looks like a garden snail shell and the Latin word for this is cochlea (see Figure 1.6). Structurally, it consists of a tube doubled over on itself so that the two ends - the oval window and the round window - lie one above the other and the ends are then rolled up for purposes of miniaturization - rather like those irritating things at parties that shoot out when you blow into them. It is easier to understand how the cochlea functions if you think of it straightened out as in Fig. 1.6. The end of the doubled-over tube where the windows are is known as the basal end, the other end as the apex. The fluid in the tube is known as perilymph. Perilymph communicates with cerebrospinal fluid (CSF) through a channel known as the cochlear aqueduct, which is a potential channel for spread of infection into the inner ear from meningitis, which in turn probably explains why deafness is a potential complication of meningitis. The partition between the two tubes is not a membrane but another tube, which contains a different fluid known as endolymph and which has a completely different ionic composition to perilymph. Within this inner endolymphatic compartment is a membrane - the basilar membrane - which supports the hair cells. The hair cells are connected to neurones and the neurones all combine to form the auditory portion of the 8th cranial nerve. (See Figure 1.7.)

The vibrating stapes sends a sound wave through the cochlear fluids, stimulating hair cells which in turn produce a nerve impulse in their neurones. What is special about the cochlea is that each frequency in the sound wave stimulates a different part of the basilar membrane. The basal end responds to high frequencies and the apical end to low frequencies - so-called tonotopic representation. This separation of frequencies is probably what makes it possible for the auditory centre - to which all the nerve impulses eventually travel - to decode the nerve impulses into the sounds that we hear.

Hearing depends on two conditions being met. The first is that the sound wave is conducted from the outside environment (air) to the inside environment of the cochlea (fluid-filled perilymph and endolymph compartments). The second is that the sound wave is accurately processed by the cochlea (sense organ) into impulses in the auditory nerve which are conveyed to the auditory centre in the central nervous system (neural pathways). These two conditions give rise to two categories of hearing loss, namely conduction and sensorineural deafness. Conduction deafness is due to problems with conduction of vibrations to the stapes footplate. Sensorineural deafness arises from problems in the cochlea, the neurones, and the central connections. ENT surgeons can often do something about conduction deafness but for the most part cannot do anything to correct sensorineural deafness apart from fitting a hearing aid to amplify sounds. It may be possible, however, to insert a cochlear implant. For more information on cochlear implants, see Chapter 2, *Audiology).*

The balance function

The vestibular system (organs of balance) consists of the three semicircular canals, and the utricle and saccule. These organs also have highly specialized hair cells but here the hairs are embedded in a covering 'cap' (cupola); see Figure 1.8. Stimulation of the hair cells to produce a nerve impulse occurs when this cap is displaced by acceleration (utricle), gravity (saccule), or rotation (semicircular canals). These nerve impulses will be interpreted as movement in the vestibular centre in the central nervous system.

Balance is a constant battle against gravity. It is made more difficult because we stand upright and because we are very mobile - we rotate and we accelerate. There are more central connections for balance than for any of the other senses. Even a mild disturbance in any of these connections gives rise to dizziness - the most incapacitating of all sensory upsets. **'Dizziness'** and **'dizzy spells'** are words patients use in a very non-specific manner. When doctors speak about dizziness they are referring to symptoms such as faintness, blackouts, feelings of imbalance, or falling. They use the word **'vertigo'** for an abnormal sensation of rotation because this is the predominant symptom of vestibular system upsets.

The most important sensory input into the balance system (see Figure 1.9) besides the vestibular system is vision, but muscle tone and joint proprioception from both the neck and the limbs are also significant. This means that symptoms of what we call disequilibrium may not only arise from the organs of balance in the vestibular system of the inner ear, but also from these other sensory inputs.

All this sensory information is co-ordinated by the vestibular nuclei in the brain stem. From here there are connections to both the cerebellum and the cerebrum. The cerebellum coordinates the body's responses to changes in sensory input. The cerebrum probably exerts some overall control but is predominantly concerned with our awareness of changes.

Rotation

How the vestibular system functions is very complex. For those of you who want to try and understand something about it, the following is a brief description of just one aspect, namely rotation. The first fact to grasp is that within the vestibular system of the inner ear the different receptor organs in each ear are the mirror images of each other, each pair having an equal and opposite action. Next consider the resting state. Since gravity is in constant operation (weightlessness induces profound symptoms of seasickness inthe uninitiated) there is a constant - and equal on the opposite side - discharge from the receptors even in the resting state - because of gravity they are never 'at rest'. So what happens when movement occurs? Displacement of a receptor in one direction increases this discharge rate and in the other decreases it. When the head rotates, the natural inertia of the fluid in a canal causes the receptor organ to displace in the opposite direction to the head movement and to initiate a change in the discharge rate. Because the semicircular canals in the two ears are mirror images of each other, there will be an increased rate of discharge from one ear and a decreased rate of discharge from the other which is fed into the vestibular nuclei in the brain stem. This is what engineers would call a finely tuned servo-mechanism that enables even small rotation movements to be very accurately monitored. From here this complex input of increased or decreased neurological activity is passed to either other brain-stem motor nuclei

or on to higher centres where it is unscrambled, decoded, and translated into some form of instant action response.

One such instant action response regulates eye movements in response to rotation. If the eyes remained in a fixed position in the orbits when the head rotated, the image would be constantly moving across the retina. To overcome this problem, the eyes need to rotate at the same speed but in the opposite direction to the rotation of the head. This is accomplished by what is known as the 'vestibulo-ocular reflex' (VOR), a complex and extensive link in the brain stem between the vestibular and oculomotor nuclei.

When the head rotates there is, however, a comfortable limit as to how far the eye can be swivelled in the opposite direction, and at short, regular intervals a very quick corrective movement returns the eye to the midline for the exercise to commence again. This slow deviation from, and swift return to, the midline is called **nystagmus.** Thus nystagmus is a physiological response to rotation of the head mediated by alterations in the resting discharge rate in the nerves from the receptors in the semicircular canals as rotation movements occur.

A number of pathological processes can alter these resting potentials - infection, trauma, a tumour on the vestibular nerve, ototoxic drugs - and if this happens the brain is fooled into believing that the head is moving, wich consequent mobilization of the VOR and nystagmus. Nystagmus is thus one of the clinical signs that indicates the presence of a vestibular disorder. In investigation of vestibular disorders it is often useful to have a recording of the nystagmus. This is obtained using an instrument that detects minute changes in electrical activity as the eyeball moves - an electronystagmograph. The print-out is an electronystagmogram or ENG.

If you are really interested in the vestibular function, you could build up similar pictures of responses to movements of linear or acceleratory nature which are monitored by the otolith organs in the utricle and saccule.

Nerves of hearing and balance

The nerves of hearing (auditory nerve) and balance (vestibular nerve) constitute the 8th cranial nerve, which passes to the brain stem via the internal auditory canal (meatus). This canal is shared with the 7th cranial nerve, the **facial nerve.**

As mentioned earlier, the facial nerve (see Figure 1.10) pursues a winding course intimately related to inner, middle, and outer ear structures. Because ear disease processes may involve the facial nerve to cause facial nerve palsy, investigation and management of facial palsy is usually undertaken by ENT surgeons. There is more about this in Chapter 4, *Core knowledge of ear disorders.* The anatomy of the facial nerve has some clinical relevance because one aspect to the investigation of facial palsy is to try to localize the site of nerve injury, and testing the function of the various branches of the facial nerve can be useful in doing this:

- The greater superficial petrosal nerve transmits secretomotor fibres to the lacrimal gland.

- The chorda tympani nerve transmits secretomotor fibres to the submandibular gland and returns sensory taste fibres from the anterior two-thirds of the tongue.

- The motor nerve to stapedius muscle is involved in the stapedius reflex (see Chapter 2, *Audiology).*

- The facial nerve terminates in five main branches in the parotid gland that provide motor fibres to all the muscles of the face responsible for facial expression.

2. Audiology

Audiology is an essential component to otology and encompasses:

- Assessment of hearing disorders.

- Assessment of vestibular function.

- Assessment of facial nerve palsy.

- Prescription and fitting of hearing aids and provision of auditory rehabilitation.

Assessment of hearing disorders

With patients who present complaining of hearing loss, or in the case of children suspected of suffering from hearing loss, much can be achieved clinically - history and examination for possible diagnosis, voice testing to gain an idea of the degree of hearing impairment, and use of a tuning fork to differentiate conductive from sensorineural deafness. Once hearing loss is confirmed, patients need to be referred for audiological evaluation to quantify and characterize the hearing loss.

Air-conduction audiogram

On an audiometer one can select pure tones of different frequencies and present these to the patient at different intensities through either headphones or speakers. The patient is asked to identify the air-conduction threshold for each frequency in first one ear and then the other, and these are charted on the audiogram. on the audiogram (see Figure 2.1) the frequencies 250-8000 cycles per second - Hz - are charted across and the intensities - decibels (dB) - are charted down the chart. Zero hearing loss (0 dB0 is biologically defined as the hearing threshold of a normal young adult but decibels also have a physics definition. The intensity of output of the audiometer can be increased up to 120 dB, above which the sound becomes physically uncomfortable - the hearing discomfort level. (The lay public have difficulty with the concept of a decible - who doesn't! - and tend to equate it with a percentage, for example 50 dB hearing loss = 50% deaf.)

Bone-conduction audiogram

If a hearing loss is cofirmed, the next investigation is a bone-conduction audiogram (cf tuning fork test). A bone-conduction transducer plugs into the audiometer and this is

placed over the mastoid bone. The threshold for each different frequency is determined in a similar manner to the air-conduction threshold and charted on the audiogram. Charting bone-conduction thresholds can be a little confusing, as we all know that normally air conduction is better than bone conduction. The audiometer is calibrated in such a way that 0 dB hearing loss with bone conduction is charted on the same line on the audiogram as 0 dB hearing loss with air conduction. Therefore a patient with a sensorineural hearing loss will have similar air- and bone-conduction thresholds - see Figure 2.1. When the bone-conduction threshold is better than the air-conduction threshold - known as 'air-bone gap' (see Figure 2.1) - this indicates a conductive hearing loss.

One factor that needs to be taken into consideration with bone conduction is that the sound will be heard equally well in both ears because the sound is conducted through the skull. If one ear has a hearing loss and the other is normal, the apparent bone-conduction threshold in the deafer ear will be better than the true threshold because the sound is being heard in the good ear. To prevent this from happening, the good ear has to be **masked.** Without going into technicalities of masking, this involves making a louder, but different, sound in the good ear than the intensity of the pure tone being presented to the deaf ear.

Conductive deafness is predominantly an amplification problem. With sensorineural deafness (sensori = hair cells, neural = nerve pathway), there is the added factor that there is nearly always an element of distortion of the sounds heard and this tends to be worse when the neural component is at fault, for example acoutic neuroma.

A **speech audiogram** is the next investigation for sensorineural deafness to try and differentiate between these two components to the deafness. Words can be heard but not understood at the threshold of hearing. It is only when the words are presented through the audiometer at 30 dB above the pure tone threshold that they can be understood. If a list of words is presented at this level to someone with normal hearing, he or she should score 100 per cent correct. Patients with sensorineural deafness due to a hair cell problem will not achieve 100 per cent but should score somewhere close to this, whereas a patient with a nerve pathway problem will achieve nowhere near this score.

Tympanometry

Tympanometry is another routine audiological investigation and as the name implies (tympanum = the middle ear) is an investigation of aspects of middle ear function. A tympanometer can access three components:

- The mobility of the tympanic membrane which can be hypermobile (atrophy), normal, hypomobile (scarring), or immobile (middle ear effusion or middle ear fibrosis) - see Figure 2.2.

- The middle ear pressure, which can be positive (valsalva), normal, or negative (eustachian tube dysfunction).

-What is called the **'stapedius reflex'.** The stapedius muscle, innervated by a branch of the facial nerve, inserts into the head of the stapes. In response to sound presented through the tympanometer probe at 80 dB or greater above the hearing threshold, there will be a reflex

contraction of the muscle (which probably has a protective function for the inner ear) that will show up on a tympanogram as a minute movement of the drum. The presence of a reflex and the threshold at which it is elicited can be determined. If a reflex is absent, the cause is either involvement of the facial nerve (facial nerve palsy) or an elevation of the hearing threshold (deafness). This is the simplest objective investigation for the hearing threshold.

Brain-stem audiometry

A more complex objective investigation is brain-stem audiometry. This is sometimes needed for the very young or the unco-operative. Using special electrophysiological equipment and electrodes, nerve impulses in the auditory neural pathways that are generated in response to sounds can be detected, amplified, recorded, and analysed to provide information regarding both the hearing threshold and the integrity of the neural pathways - the latter being useful in suspected acoustic neuroma.

Assessment of vestibular function

Nystagmus usually indicates the presence of a disorder of the vestibular component of the balance system (vestibuloocular reflex). Nystagmus can be visually observed but more usefully it can be recorded using a special electrophysiological instrument which makes use of the fact that the eyeball has electrical polarity and movements can be detected and amplified. When these movements are displayed and recorded this is known as an electronystagmogram.

If spontaneous nystagmus is present this indicates an active vestibular disorder.

If no spontaneous nystagmus is present, nystagmus can be induced:

- In response to position changes (benign positional vertigo, vertebro-basilar insufficiency).

- By visual 'tracking' of a pendulum or vertical bands on a rotating drum (central causes for vertigo).

- Following cold or warm water **(caloric)** irrigation of the ear canal. The vestibular organ is in close proximity to the deep part of the ear canal and is very sensitive to changes in temperature - see precautions in *Syringing of ears,* Chapter 14. The caloric test localizes the disorder to one or other ear (vestibular neuronitis, Ménière's disease, trauma).

Although vestibular disorders can usually be diagnosed from the history (see Chapter 5, *A practical approach to ear disorders),* investigation of nystagmus may sometimes provide useful additional information.

Assessment of facial nerve palsy

It is sometimes helpful to try and localize the site of injury in facial nerve palsy. This is done by testing responses of each of the branches of the facial nerve - see *Facial nerve* in

Chapter 1. The stapedius reflex, mentioned above, is the test used for the branch to the stapedius muscle.

It is also useful to know what state the palsied nerve is in - neuropraxia or nerve degeneration - because this relates to the prognosis. Neuropraxia always recovers fully, whereas a significant number of patients who have undergone nerve degeneration never regain full function and are left with problems such as incomplete recovery with facial weakness and re-routing of nerve fibres to give mass movement (synkinesia) or 'crocodile tears'. Sometimes surgical intervention with nerve decompression may be indicated in nerve degeneration to try and improve the prognosis. The state of the nerve is most simply determined by the response to nerve stimulation. The nerve trunk is stimulated and one looks to see whether or not this elicits a muscle twitch. This is done a week after the onset of the palsy because degeneration takes this long to show up on testing. A responsive nerve is in a state of neuropraxia, whereas an unresponsive nerve has undergone degeneration. A more quantitative investigation is electroneuronography which involves recording the impulses generated in the nerve using electrodes and an electrophysiological instrument. This can detect the presence of nerve degeneration much earlier than simple nerve stimulation. It is needed when surgical intervention is contemplated because the earlier this is done the greater the chance of benefit.

Prescription of hearing aids

Bone-conduction hearing aids

Bone-conduction hearing aids are used for conditions in which there is either no external ear canal (congenital microtia with canal atresia, traumatic stenosis) or troublesome recurrent ear infection (active chronic otitis media, mastoid cavities), makes the use of an ordinrary hearing aid difficult. Usually a bone-conduction aid uses something similar to the bone-conduction transducer of an audiometer which is held in place with a head band. At their most sophisticated, however, bone-conduction aids are anchored into the skull with titanium screws - bone-anchored hearing aids.

Air-conduction hearing aids

Air-conduction hearing aids originally consisted of a pick-up microphone and amplifier worn on the body which was connected to the output speaker in the ear by a cord. This type of aid is sometimes still used when high amplification is needed of wehn a patient has problems manipulating very small controls. Modern aids have been miniaturized and are worn either behind the ear or in the ear canal. Both of these have to fit very precisely in the ear canal because any sound leak around the canal 'mould' causes electronic feedback to the pick-up microphone in the aid and a loud whistle or whine. Air-conduction aids vary greatly in sophistication (and hence price!) but can often be tailored to the type of hearing loss. High-tone deafness requires an aid with 'low-tone cut-off' and amplification of high tones - this can also be helped by manipulating the mould to make use of the secective high-tone amplification that occurs in the ear canal. Patients whose hearing is very sensitive to loud sounds can be fitted with aids with automatic 'gain' control.

Hearing aids are simply amplifiers and patients with conductive deafness will benefit most from using them because their problem is simply one requiring a louder sound. When

sensorineural deafness is the problem, there is almost always some element of distortion to the sounds heard. Nonetheless, most patients with sensorineural deafness will benefit from using a hearing aid although they may find some hearing situations better than others when using their aid. Audiotherapy involves working with patients so that they derive the maximum benefit from their aid. There will be the occasional patient who finds amplified distortion intolerable. These patients may require to be taught additional skills, such as lip reading, to help them overcome their deafness handicap and to use their aid at lesser amplification.

Cochlear implants

Cochlear implants are used when there has been near total loss of ahir cells but the neural pathways are still intact. These very sophisticated (and hence very expensive!) devices consist of a pick-up component that convers sound into electrical impulses and directs these to a multichanneled electrode (high tones to one end, low tones to the other) which is implanted into the cochlea. The electrodes directly stimulate the nerve fibres and the generated impulses are interpreted as sound in the auditory centre. If one considers thaty 30.000 hair cells are being replaced by 22 electrodes, the quality of the sound can be deduced but with intensive training the quality of an otherwise totally deaf patient's life can be dramatically improved. Many patients are able to understand speech by using a combination of the sound heard and visual clues (lip reading).

3. Structured approach to examination of the ear

The summary in Table 3.1 is the basis for learning how to examine an ear, something that is not usually well taught and which is a difficult skill to master. The rest of this chapter expands on the summary providing both practical details and explaining the significance of findings.

Table 3.1 Aids to examination in otology

The external ear

Normally is determined b exclusion of abnormality. There are really only three categories of abnormality:

- Deformities
    - congenital: remember possibility of deafness
    - old acquired: eg, cauliflower ear
    - recent acquired, traumatic: eg, haematoma
        inflammatory: eg, perichondritis
        neoplastic: eg, squamous cell or basal cell carcinoma

- Inflammatory swelling
    - mastoiditis: abnormal middle ear
    - lymphadenitis: normal middle ear

- Surgical scars: tympanoplasty, mastoidectomy

The ear canal

Normality is determined by exclusion of abnormality. There are really only four categories of abnormality:

- Wax - diagnosis: **wax**

- Foreign body - diagnosis: **foreign body**

- Infection - diagnosi: **otitis externa** - localized (furuncle), generalized

- Others
    - uncommon conditions, eg, exostoses, hyperkeratosis, stenosis, atresia
    - a defect in the posterior wall opening into a mastoid cavity

The eardrum

Normality is determined by exclusion of abnormality. There are really only five categories of abnormality:

- Inflammation - diagnosis: **acute otitis media**

- Perforation:
    - infected - diagnosis: **active chronic otitis media**
    - dry - diagnosis: **inactive chronic otitis media**

- Scarred - diagnosis: **healed otitis media**

- Retracted - diagnosis: **otitis media with effusion**

- Others:
    - uncommon conditions, eg, uninfected cholesteatoma

The hearing

Normal hearing is determined by an accurate response to your **whispered voice test.**

If the resopnse is inaccurate, proceed to test with your **normal voice** - moderate hearing impairment - or your **loud voice** - severe hearing impairment.

Use a **tuning fork** to determine whether the hearing impairment is **conductive** or **sensorineural.**

Step 1

Begin with a general examination of the head from the front, focus in on the ears. Are both ears in a normal position?

Low-set and anteriorly displaced ears are representative of some of the congenital abnormalities that fall under the general term 'funny-looking kid', or FLK. These children are best referred to a paediatrician or geneticist for definition of the abnormality, as there may be other more significant features to the abnormality.

Step 2

View from the side, focus in on the ear. Are both ears normal? If not, is it possible to define the abnormalities when each ear is examined in turn?

Congenital deformity. The significance of finding a congenitally abnormal ear is that these are usually associated with deafness - either conductive or sensorineural. If only one ear is affected, it is essential to have the hearing in the other ear assessed as soon as practical. If both ears are affected, referral to an ENT specialist is mandatory, as hearing aids will need to be fitted before one year of age if there is any hearing impairment. Good hearing is essential for normal speech development. Children develop speech as they mimic the sounds that they hear and if they cannot hear well, their speech development will be retarded or even absent. They develop vocabulary and understanding of words through use of speech and hearing. Since humans think in words, a restricted or absent vocabulary will impair intellectual development.

A more common abnormality is the pre-auricular sinus, usually situated just above the tragal cartilage. These sinuses have the potential to become infected and form an abscess.

Old acquired deformity. Not much can be done for these patients but the significance lies in possible stenosis of the ear canal. If obstructed, the accumulation of desquamated epithelium in the canal may eventually lead to an expanding, destructive mass trapped in the ear canal.

Recent acquired deformity - traumatic. Lacerations are obvious and may require suture. Of more significance is a haematoma of the pinna. If this is not adequately drained, it may potentially heal with fibrosis and contracture to produce the deformity known as a 'cauliflower ear'.

Recent acquired deformity - inflammatory. A painful, swollen, inflamed pinna is caused by underlying perichondritis. If this condition is not promptly and adequately treated, it has the same potential for subsequent deformity.

Recent acquired deformity - neoplastic. The pinna is one of the common sites in the head and neck for development of basal cell and squamous cell carcinoma. Any non-healing ulcers should be regarded suspiciously.

Inflammatory swelling. Inflammatory swelling behind the ear can be caused by either mastoiditis or lymphadenitis. Mastoiditis is always accompanied by some abnormality of the middle ear - middle ear effusion, frank otitis media, or active chronic otitis media. In the absence of any abnormality, it is probably a lymphadenitis and a primary source of infection, usually in the scalp, should be sought.

Surgical scars. These scars may be either behind the ear or in front above the tragal cartilage, and indicate that either a tympanoplasty (middle ear and eardrum reconstruction) or a mastoidectomy have been performed. They provide a clue as to what to expect when examining the ear canal or the eardrum. A successful tympanoplasty will usually leave the drum scarred, whereas if unsuccessful, there will still be a perforation. There may be no signs to see after a simple mastoidectomy but a radical mastoidectomy, performed for cholesteatoma, will have opened the mastoid cavity into the ear canal by removing the posterior wall of the canal. It is surprisingly easy to overlook a mastoid cavity, probably because the instinct when using an otoscope is to look deep and anteriorly for the eardrum. Knowing that there has been mastoid surgery before looking with the otoscope makes it less likely that a mastoid cavity will be missed.

Step 3

Now take the otoscope! Before you start using it, you need to check that it works by switching it on and checking that the bulb delivers a good light - you will have great difficulty distinguishing any of the subtle features that you need to see unless the light is bright. Select the largest size speculum that will fit into the ear canal. The field of view through an otoscope is very narrow and to build up a composite picture you have to move the speculum around and piece together all of the separate images. The larger the speculum, the less it has to be moved around.

To see into the ear canal it has to be straightened. This is done by taking hold of the pinna and gently pulling it outwards and backwards with a little up or down displacement as needed. If this causes discomfort, it is an indication of possible otitis externa. Before inserting the speculum into the ear canal, inspect the opening to the canal. A furuncle may arise in the hair-bearing area here - classified as **localized otitis externa.** Furuncles are easy to miss unless looked for, and squashing them with a speculum is very painful. Inspecting the entrance to the canal will also reveal any discharge that will need to be mopped or syringed before anything meaningful can be seen.

While still holding the pinna, gently insert the speculum into the canal, all the time looking through the otoscope to see where you are going. Do hairs obstruct your view? Gently manoeuvre the speculum past them. Does wax obscure your view? Can you see adequately past it or are you going to have to remove it?

Step 4

Once you can see the ear canal clearly the question you have to answer is 'Is the ear canal normal?'. This is determined by excluding any abnormality.

Wax and foreign bodies should be obvious and are best removed by syringing.

If the canal walls are inflamed, desquamating, and swollen or if there is an accumulation of wet debris in the canal, the diagnosis is **generalized otitis externa.**

There are a few **uncommon conditions** that occur in the ear canal. One of these occurs in people who swim a lot in cold water, who develop 'sweimmer's exostoses' - nodular,

bony protrusions from the deep bony portion of the canal. Another is hyperkeratosis - an accumulation of desquamated squamous epithelium in the deep ear canal resulting from failure of the normal migratory process that 'self-cleanses' the canal. Stenosis of the canal can be either congenital or acquired by atresia is always congenital. If a radical mastoidectomy has been performed, the posterior wall of the ear canal will have been removed, and you should be able to see backwards and upwards into an open mastoid cavity. These open cavities can be the source of problems by either filling up with wax or becoming chronically infected.

Step 5

Having established the status of the ear canal, the next step is to examine the eardrum. To do this you need to angle the otoscope speculum a little forwards until you can see the drum. The eardrum is the window to the middle ear and all of the middle ear disorders can be diagnosed by the characteristic appearances that they impar to the drum. A normal eardrum indicates a normal middle ear, although it will not provide information about the essential hearing function of the ear, which has to be assessed after otoscopy has been carried out.

There are two questions that you have to answer at this point. The first question is 'Is the drum intact?'. Examine the drum closely and move your speculum tip around until you have been able to see all of it. The second question is 'Is the drum normal?'. To answer this, you need to think about what you are seeing.

A **normal eardrum** (see Figure 3.1) is usually described as having a smooth reflective surface with the contour a little indrawn and incorporating the handle of the malleus in the upper part of the pars tensa. The handle of the malleus is recognized by looking for the small lateral projection at its upper end that marks the division between pars tensa and pars flaccida (which is often very insignificant).

However, there are so many variations to the appearance of a normal eardrum that this latter question is best answered by excluding abnormality. There are really only five categories of abnormality, each representing one of the middle ear disease processes. It helps to be dogmatic about these appearances because once again there are so many variations to each of the abnormalities.

Inflamation. As a general rule inflammation of the drum (see Figurwe 3.2) is indicative of **otitis media.** The inflammation of otitis media starts with hypervascularity, goes through a stage of generalized inflammation to the late stage of the inflamed, 'bulging' drum. This is not, unfortunately, infallible and sometimes you will be caught out by inflammation of the drum from variations of otitis externa. Pneumatic otoscopy, which will be mentioned later, helps to distinguish the two, as the drum will be what we call 'mobile' if the cause is otitis externa.

Perforation. (See Figure 3.3.) By definition, a perforation in the drum indicates chronic otitis media - unless it is a recent traumatic perforation or a recently perforated acute otitis media! If the ear is dry and the mucosa of the middle ear, seen through the perforation, is not inflamed, this is an **inactive chronic otitis media.** If the ear is discharging and the middle ear mucosa is inflamed - there may even be visible granulation tissue on the

perforation margins - then this is an **active chronic otitis media.** It is helpful for descriptive purposes to try to define the size of the perforation - 20 per cent, 50 per cent, and so on.

Scarred. (See Figure 3.4.) An intact but scarred eardrum is indicative of **healed otitis media.** These scars may be visible fibrosis or may have become calcified, when they have an appearance described as 'chalk patches'. An atrophic scar in which there is loss of the middle layer of the drum gives rise to the appaearance of a transparent patch in the drum described as a healed perforation.

Retracted. (See Figure 3.5.) Retraction of the drum results from a negative middle ear pressure, after obstruction of the eustachian tube, which sucks the drum inwards. It is recognized by looking both at the angulation of the handle of the malleus - which becomes more horizontal than vertical - and anteriorly at the margin of the drum where the 'sucked-in' appearance is likely to be most noticeable. It is indicative of **otitis media with effusion.** A middle ear effusion is confirmed by assessing the mobility of the drum by pneumatic otoscopy. When there is an effusion, the drum is immobile.

Pneumatic otoscopy. This is a useful technique to master as it provides the essential additional information sometimes needed to confirm a diagnosis and basically assesses the mobility of the drum in response to changing the pressure in the ear canal. Attach a pneumatic bulb to the otoscope and depress it a little. Insert the speculum so that it fits tightly into the ear canal and further depress the bulb. Do the blood vessels blanch and does the drum move away from you? If so, you have a good seal. Release the pressure on the bulb. Is the drum sucked towards you? If it does both of these, the mobility is normal, the middle ear contains air and is normal. The problem comes when the drum appears to be immobile. The chances are that you do not have a good seal, in which case try to reposition the speculum or change up to the next size speculum and try again. If the drum is still immobile and you are sure that you have a good seal, either there is a **perforation** which you have missed (easy to do - which is why it is important to perform this manoeuvre) or there is a middle ear effusion - **otitis media with effusion** (these sometimes have a relatively normal-looking drum and are also easily missed).

Others. (See Figure 3.6.) Is there something else about the appearance of the deep ear canal or drum that makes you wonder whether there is another disease process present? A granulation tissue polyp in the deep ear canal over the eardrum, or even filling the ear canal, often hides an underlying infected cholesteatoma. Cholesteatoma usually presents only once it is infected as an active chronic otitis media, when it is difficult to diagnose unless squamous epithelium can be seen in the middle ear through the perforation. Occasionally it will present as deafness without infection, when it may be possible to identify from the presence of a collection of whitish, hyperkeratotic debris in what looks like a pit or a perforation. Recent trauma or acute pressure changes may cause bleeding in the middle ear, imparting a bluish/black appearance to the drum, an appearance known as a haemotympanum.

Step 6

Hearing. Examination of the ear is not complete until the hearing has been assessed. In the clinical setting the hearing threshold should be considered as being in the range of:

- normal hearing

- moderate hearing loss (hearing impairment)

- severe deafness (deaf).

Perhaps the most useful and easy to administer test to determine the hearing threshold is one based on the fact that if you have normal hearing you should be able to make out what is said to you in a whisper from halfway across the room. If you have a hearing impairment, the person speaking has to come a bit closer and speak in a more normal voice. If you are deaf, the person has to come close and speak very loudly for you to make out what has been said.

Step 7

Stand behind the patient and test the hearing in each ear by a 'Repeat after me' voice test. Since you need to test the hearing in each ear separately, you have to prevent the ear that you are not testing from hearing what you are saying. This is called 'masking' and is done by placing a finger on the tragus, pressing it to occlude the ear canal and at the same time gently rubbing it all the time you are testing to create a noise in that ear.

Standing behind, a little to the side to be tested and at arm's length away, test the hearing by using a complex combination such as a letter, a number, and another letter (for example, B7K, T4S) rather than something that the patient could make a good guess at what you are saying such as the traditional '99' or '21'.

If the patient is:

- able to accurately repeat your whispered voice at arm's length - he/she has **normal hearing.**

- not able to hear your whispered voice but able to repeat your normal voice at arm's length - he/she has a **hearing impairment** (moderate hearing loss).

- not able to hear your normal voice but able to repeat your loud voice at arm's length - he/she is **deaf** (severe hearing loss).

It will be found that children are reluctant to co-operate with a 'Repeat after me' test. This can be overcome by asking them to do something such as touching their nose, or pointing to their mouth in the same sequence of whispered voice, normal voice, loud voice. You may find that you cannot mask younger children while you are testing them, but at least you will be able to get an impression as to where their hearing threshold lies.

Step 8

If there is any hearing impairment, testing with a tuning fork may indicate whether the deafness is conductive or sensorineural. The tuning fork (Rinne) test is useful, but unfortunately not always reliable and is not usually possible when dealing with children. The

importance of carrying out a tuning fork test is that although conductive deafness is usually associated with a visible disease process, this is not always so (for example otosclerosis) and for conductive deafnesses there is the possibility of surgical correction, whereas a sensorineural deafness probably indicates the need for a hearing aid. Points to note are:

- Air conduction better than bone conduction - the tone heard when the tuning fork is held next to the pinna sounds louder than when the base of the tuning fork is pressed against the mastoid bone. This indicates that the conductive mechanism is intact.

- Bone conduction better than air conduction - the tone heard when the tuning fork is held next to the pinna sounds softer than when the base of the tuning fork is pressed against the mastoid bone. This indicates an impairment in the conductive mechanism.

Step 9

Assessment of hearing in babies is a highly skilled procedure that is usually performed by specially trained audiologists. The medical practitioner can obtain a fairly good idea as to whether or not there is a hearing problem by asking the parent or caregiver questions relating to both hearing ability and speech development appropriate for age. The questions to ask for the different ages are given in Table 3.3.

Table 3.3 Questions for use in assessing hearing in babies

Baby only a few weeks old:

Does your baby appear to be listening to you when you talk or sing?

Does your baby open his or her eyes or blink when there is a noise?

Baby about six months old:

Does your baby enjoy your talking or playing word games with him or her?

Does your baby try to see where a noise is coming from by turning his or her eyes or head towards the sound?

Baby about nine months old:

Does your baby appear to enjoy babbling and making other noises?

Does your baby appear to respond to even very soft sounds?

Baby about a year old:

Is your baby beginning to say baby words?

Does your baby respond when you say his or her name and the names of things he or she plays with.

Baby about a year and a half old:

Is your baby beginning to use simple words?

Does you baby pick up or point to things around the house when you ask him or her to do this?

Babies of two years or more:

Is your baby putting words together and trying to talk to you?

Do you think your baby can hear normally even when you speak to him or her in a very soft voice?

Step 10

Piecing together all the information obtained from the history regarding the ear complaint, the findings on examination, and the presence of any hearing loss should enable a diagnosis to be made and a treatment or management plan to be formulated.

4. Core knowledge of ear disorders

The external ear

Impacted wax

Ear wax consists of sebaceous and ceruminous gland secretions combined with hairs and desquamated skin - ear canal epithelium migrates outwards from the drum and deep parts of the canal. The wax varies in colour and consistency and it is normal to have some wah in the outer part of the ear canal. Wax probably has some bactericidal action and may provide some protection for the ear canal. Attempts to 'clean' the ear often simply push any accumulated wax deeper into the canal. Impaction of wax also tends to occur in very dry climates.

Remember that wax dissolves in water. Therefore if wax needs to be removed it is best done by syringing - see Chapter 14. Indications for removal of wax are a need to see the drum, discomfort in the ear canal, or deafness from impaction.

Otitis externa

Otitis externa is described as being localized (a furuncle) or generalized and may occasionally develop complications (cellulitis, adenitis, perichondritis, necrotizing otitis externa). It may be caused by infection (bacterial, fungal, viral) or by allergy (dermatitis). The pathological process is either inflammation and abscess formation in a hair follicle (furuncle - common) or inflammation of the skin and subcutaneous tissues of the canal causing oedema/induration and increased epithelial desquamation (common). Infection may spread into surrounding soft tissue (cellulitis) and there may be adjacent lymphadenitis - pre-auricular, postauricular, subauricular - or it may spread to involve the perichondrium of the auricular

cartilages (perichondritis - uncommon). In a diabetic or immunocompromised patient, pseudomonal infection may spread rapidly into the surrounding temporal bone and beyond (necrotizing otitis externa - uncommon). The symptoms and signs relate to the pathological process, its site, and the extent.

Symptoms

- Itching/irritation

- Pain

- 'Blocked ear'

- Discharge

Signs

- Tenderness on manipulating the pinna or tragus

- Inflammation and swelling - localized or generalized

- Desquamation and accumulation of debris

- Purulent discharge

- Conduction deafness

- Cellulitis

- Tender lymph nodes

- Swollen, inflamed, tender pinna (perichondritis)

- Ulceration, granulation tissue (necrotizing otitis externa)

Treatment

- Cleaning - syringing (see Chapter 14)

- Instillation of:

    - something soothing: glycerine/ichthammol either as drops or soaked into a cotton wool wick gently inserted into the ear canal
    - something antiseptic: tinct Merthiolate ear drops
    - something anti-inflammatory: steroid-containing ear drops (these are usually combined with an antibiotic)
    - something specific: antifungal drops or creams

- Cellulitis requires antibiotics. Perichondritis requires IV antibiotics. Necrotizing otitis externa requires high-dose specific IV antibiotics and debridement surgery.

Foreign bodies

Small children tend to put things into their own and other children's ears. Sometimes the tips fall off cotton buds, and sometimes insect crawl into ears. It is best to attempt to remove all of these by syringing rather than by trying to hook them out as the latter method usually hurts the child and makes subsequent attempts almost impossible. If unsuccessful, refer to an ENT service or, if unavailable, attempt removal under anaesthetic.

Deformities of the pinna and/or canal

Congenital

- Minor:
    - Cosmetic deformities such as prominent (bat) ears.
    - Pre-auricular sinuses (pits). Debris may accumulate and become infected to form an abscess requiring drainage. These sinus tracts need to be excised once the infection has resolved.

- Major:
    - Degrees of what is called microtia with severe deformity of the pinna and stenosis or atresia of the canal. The importance of these major deformities lies in the resulting conductive hearing loss. If this is bilateral, there is an urgency to refer the infant to a specialist service so that management of the deafness can be begun early enough to permit speech development.

Acquired

- Lacerations, burns, etc, produce defects and scarring (cosmetic).

- Haematomas beneath the perichondrium cause cartilage necrosis (cosmetic, 'cauliflower ear'). Haematomas should be aspirated or a suction drainage tube inserted. If unable to aspirate the haemangioma, refer urgently to a specialist service (Plastics, ENT).

- Trauma involving the canal causes fibrosis with subsequent stenosis which interferes with the normal epithelial migration out of the canal. Debris accumulates and can act as a focus for infection. Stenosis requires referral for remedial surgery.

Neoplastic

- The prominent, exposed pinna is a common site for development of basal cell and squamous cell carcinoma. Remember this in consideration of any persistent ulceration.

The middle ear

An overall concept of middle ear disorders may be gained from consideration of the diagram in Table 4.1 below.

Table 4.1 Middle ear disease

Acute otitis media --> Otitis media with effusion
V Mastoiditis V
Chronic suppurative otitis media Cholesteatoma

Acute otitis media

Acute otitis media most commonly occurs in young children - peak incidence one to two years - but it can occur at any age. Although often precipitated by a cold or other viral upper respiratory tract infection, it is usually a secondary bacterial infection **(Haemophilus influenzae, Streptococcus pneumoniae, Streptococcus pyogenes)** that involves the mucous membrane of the middle ear cleft. The infected mucosa produces a mucopurulent exudate which initially drains away down the eustachian tube. When inflammatory swelling obstructs the eustachian tube, a middle ear empyema develops, the inflamed tympanic membrane 'bulges' and may perforate. (See Figure 4.1.) Complications occur when infection becomes trapped within the mastoid air cells - see *Mastoiditis.*

Most acute otitis media will undergo natural resolution but treatment with antibiotic is recommended to prevent both the complication of mastoiditis and to avoid the sequelae of unresolved infection after perforation - see *Chronic suppurative otitits media.*

Otitis media with effusion

The eustachian tube has two functions - aeration of the middle ear cleft and drainage of any middle ear secretion. Middle ear effusions develop when tube function is impaired and this most commonly occurs in children following an episode of otitis media. In many children it takes some time before the middle ear regains normal aeration. Ventilation failure arises either because the middle ear mucosa continues to produce an excess of mucus which 'floods' the eustachian tube and prevents ascent of air, or because the mucosal lining of the tube remains oedematous to cause a mechanical blockage. If the middle ear cannot be ventilated, any residual air is absorbed to create what is called a 'negative middle ear pressure' and mucus accumulates. This impairs vibration of both the drum and the ossicular chain, causing a conductive deafness (which needs to be assessed). The presence of mucus imparts a 'dull', somewhat pinkish appearance to the drum which is generally retracted and immobile. (See Figure 4.2.)

In children, natural resolution is usual given time but there are two problems with expectant management:

- The first is the effect of a persistent hearing impairment on the child's speech and intellectual development. This is managed by inserting a minute ventilation tube through the

drum (often known as a grommet tube because of its shape) to aerate the middle ear and hence restore hearing. These tubes usually extrude spontaneously after a few months.

The second is the effect of a persistent negative middle ear pressure on the retracted drum. The overstretched drum may become atrophic or a retraction pocket may form - see *Cholesteatoma.*

In adults, middle ear effusions are uncommon, sometimes encountered after episodes of otitis media and occasionally seen after barotrauma (flying, diving). In adults the effusion is usually serous rather than mucoid (because there are fewer mucus-secreting glands in the adult middle ear mucosa) and this imparts a yellowish colour to the drum.

In adults with middle ear effusion suspicion has to be high of mechanical obstruction of the eustachian tube by a tumour in the nasopharynx.

Chronic suppurative otitis media

Chronic discharging ears are all better classified as **active chronic otitis media** (see Figure 4.3). There are several causes for a chronic discharge. The most common is the mucosal type of chronic suppurative otitis media (CSOM), which is discussed in this section. Cholesteatoma is another cause, discussed next, and, uncommonly, other causes will be encountered such as tuberculous otitis media, which is discussed under *Unusual middle ear disease.*

Not all cases of acute otitis media that have progressed as far as perforation resolve. There are two reasons for this:

First, while the body defence mechanisms are clearing the middle ear of the bacteria that caused the acute infection, it becomes colonized by coliform bacteria (Proteus, Klebsiella, *Escherichia coli*, Pseudomonas), which find their way in through the perforation. The body appears to be fairly tolerant of these gut commensals and once they establish in the middle ear, infection persists for a long time with continuous production of mucopurulent discharge (otorrhoea).

Second, changes may occur at the perforation margin which will prevent closure of the perforation during the healing phase. Ideally, the raw margins of the perforation - remember that there is a mucosal surface on the inside of the drum and a squamous surface on the outside - should regenerate and grow across the defect to close it. This doesn't always happen and sometimes the squamous epithelium on the outside of the drum forms a junction with the mucosa on the inside to create a 'sealed' margin to the perforation. The perforation has now healed but it has healed 'open' rather than closed. Once this has occurred, the perforation will remain open even after infection is controlled to create a state then known as **inactive chronic otitis media.** This is the origin of the definition that any ear with a perforation is classified as **chronic otitis media.**

A further aspect to chronic middle ear infection is that a persistent perforation contributes to conductive deafness. The degree of conductive deafness will be even greater

if there is either osteitis and erosion of individual ossicles during the stage of infection, or fixation of the ossicular chain by fibrosis during the healing phase.

Treatment involves a combination of aural toilet and instillation of ear drops, supplemented by use of antibiotics.

Aural toilet and instillation of ear drops are used in an attempt to render the middle ear environment hostile to infecting organisms. This is achieved by drying (see *Dry mopping of ears*, Chapter 14) and instilling either acidification agents (acetic acid drops) or antiseptic drops (boracic ear drops) - see *Instilling ear drops,* Chapter 14. Sometimes these inexpensive preparations are not effective and a specific antibiotic has to be used in an ear drop. The more frequently the ear is cleaned and ear dros are instilled, the more effective treatment is likely to be.

It is sometimes difficult to get the ear clean by dry mopping alone and sometimes patients are not co-operative enough to do it adequately. In these circumstances syringing is a very effective way of cleaning out all the infected debris and mucopurulent secretions - see *Syringing of ears*, Chapter 14.

If it appears likely that dry mopping and ear drops will be ineffective, then systemic antibiotics have to be given. This situation applies to most children and also when there is only a very small perforation. The antibiotic selected will have to be one that is effective against a mixed coliform infection. Some of these organisms are sensitive to easily given broad-spectrum antibiotics (Augmentin - if available - amoxycillin, or cotrimoxazole), but many are resistant. When resistant organisms predominate, systemic administration of the specific antibiotic required (gentamicin or cephalosporins) is usually impractical since those that are effective have to be given by injection. In this situation one has to resort to topical therapy - see above - and fortunately there are a number of suitable antibiotics incorporated into ear drops which can be used.

Once the middle ear infection is controlled the degree of hearing loss can be assessed, conductive deafness being one indication for tympanoplasty - surgery for repair of the middle ear. Tympanoplasty surgery comprises both myringoplasty, which is repair of the perforation, and ossiculoplasty, which is reconstruction of a sound-conducting ossicular chain.

Unfortunately, re-infection of ears with a persistent perforation is common, either because contaminated water gets into the middle ear through the perforation or because a further episode of otitis media occurs related to upper respiratory tract infection. Another indication for tympanoplasty would be to close the perforation to prevent re-infection.

Once the acute infection of otitis media has settled, chronic mucosal middle ear infection is painless. The onset of pain in a chronic discharging ear is an indication that there has been spread of infection from mucosa into underlying bone, particularly in the mastoid, and mastoiditis - see below - has to be considered. Such cases need urgen referral to have this excluded.

Cholesteatoma

This weird-sounding word is used for a very destructive middle ear disorder which is always cause for concern in any ear with active chronic otitis media - not only because of the middle ear destruction but also because it has potential for complications such as mastoiditis and intracranial spread of infection.

Negative middle ear pressure - mentioned above associated with a dysfunctional eustachian tube - causes the eardrum to retract into the middle ear space. Sometimes this retraction process is confined just to one area of the drum and results in the formation of a retraction pocket. The more common sites for these retraction pockets to form are either in the postero-superior portion of the drum between the malleus handle and the posterior margin, or in the pars flaccida portion of the drum. Since the outside layer of the drum is covered by desquamating squamous epithelium, this now forms the lining of the pocket. Problems arise because the normal outward migration of desquamated epithelium no longer operates within these pockets and squamous debris accumulates. See Figure 4.4. The pearly white keratin debris was originally thought to be cholesterol - hence the derivation of the name cholesteatoma. These pockets gradually start behaving like epidermoid cysts and invade the middle ear cleft - particularly upwards into the attic area and backwards into the mastoid antrum (hence the derivation of another name for this condition - attico-antral disease).

If the squamous-lined cholesteatoma 'sac' comes into contact with bone, the bone is eroded away. This bone may be an ossicle (conductive deafness), the facial nerve canal (facial nerve palsy), or the lateral semicircular canal (vertigo).

These cholesteatoma sacs tend to become infected and are one cause of actice chronic otitis media. Because the sacs obstruct drainage from the mastoid air cell cystem, they are a potent cause of mastoiditis and, for some reason, predispose to the intracranial complications of mastoiditis.

Cholesteatoma is a difficult condition to diagnose and even ENT specialists often need to use the operating microscope and microsuction before they can identify cholesteatoma. Clues for the non-specialist are a scanty, smelly discharge, failure to respond to medical treatment, and the presence of pain which may be the first indication of spread of infection to cause mastoiditis.

Any patient with a chronically discharging ear that has not responded to one or two courses of treatment should be referred to exclude cholesteatoma.

The treatment of cholesteatoma is surgical with eradication of the cholesteatoma and an attempt to reconstruct a middle ear sound-conducting mechanism. Sometimes this is possible with what is known as a 'closed' technique but more often disease is eradicated by creating a large mastoid cavity that opens into the ear canal - the radical mastoidectomy.

Mastoiditis

Mastoiditis is the dreaded complication of middle ear cleft infection because it carries a high risk of intracranial spread of infection - meningitis, extradural abscess, sigmoid sinus thrombosis, temporal lobe abscess, cerebellar abscess - see under *Relations of the middle ear*, Chapter 1. Mastoiditis occurs when infection is trapped within the mastoid air cell system and then spreads into the surrounding bone to cause a mastoid osteitis (hence mastoiditis). See Figure 4.5. In children, entrapment is usually caused by the swollen mucosa of an acute otitis media (be suspicious of persisting pain or toxicity in a child with acute otitis media). In adults, this can be the cause but it more often arises from swollen mucosa or a cholesteatoma in active chronic otitis media. As the osteitis develops (pain/tenderness), the soft tissue over the mastoid becomes inflamed and swollen (postauricular swelling). The presence of postauricular swelling usually indicates that a sub-periosteal abscess is forming over the mastoid bone. Remember that intracranial spread of infection may be occurring at the same time (headache, meningism, etc). See Figure 4.6.

Once mastoiditis is diagnosed, management involves use of high-dose intravenous antibiotics and urgen interventional surgery to decompress the mastoid abscess and eradicate any osteitic bone (cortical mastoidectomy). If cholesteatoma is present a radical mastoidectomy will be needed. Close monitoring is required to detect any intacranial spread of infection at an early stage, in which case neurosurgical intervention is required.

Otosclerosis

In otosclerosis there is an overgrowth of 'spongy' bone in the bony capsule of the inner ear. When this involves the oval window, the stapes becomes fixed and the result is a conductive deafness. It probably has a genetic origin - most common in people of European origin, less common in those of Asian origin, and almost unknown in those of African origin. Presentation is usually in the 20 to 40 age group and in women the process is accelerated by pregnancy. It can be unilateral but tends to be bilateral. The conductive deafness can be managed with a hearing aid but the preferred option is surgical - stapedectomy. In this delicate operation the stapes is removed and replaced by a minute, piston-shaped prosthesis. See Figure 4.7.

Unusual middle ear disease

Always consider the possibility of TB when granulation tissue is a prominent feature of a chronic suppurative otitis media. Tuberculous otitis media frequently precipitates a facial nerve palsy. (Remember cholesteatoma may also precipitate a facial palsy.)

The combination of a discharging ear and a facial palsy is to be regarded as TB until proven otherwise.

Middle ear neoplasia is rare. Benign neoplasia (glomus tumour - a chemodectoma) presents with pulsating tinnitus and an abnormal, reddish tympanic membrane. Malignant neoplasia presents with persistent, severe pain in an ear full of granulation tissue.

The inner ear

Disorders affecting both cochlea and vestibular components

Trauma

Base of skull fractures (bleeding and CSF leakage from the ear) may transect the temporal bone with disruption of the inner ear to cause deafness (permanent), vertigo (compensatin occurs), and occasionally facial nerve palsy (surgical exploration required). Lesser degrees of head injury may cause labyrinthine concussion.

Labyrinthitis

Labyrinthitis is uncommon. The complex of spaces and channels within the temporal bone that comprises the inner ear is known as the labyrinth. Infection may spread into the labyrinth either from meningitis - the perilymph of the inner ear is in communication with CSF, **hence the need to be aware of deafness as a complication of meningitis** - or from infection in the middle ear cleft, particularly when cholesteatoma has eroded the bony capsule of the labyrinth. Deafness and vertigo accompany such infection, which usually destroys all the membranes and hair cells of both cochlea and vestibular system to cause a 'dead ear'. Management is by intravenous antibiotics, surgical eradication of middle ear disease, and wide exposure of the labyrinth to allow drainage.

Acoustic neuroma (schwannoma)

Acoustic neuroma is rare. The 8th cranial nerve passes from the inner ear via the internal auditory canal to the brain stem (cerebellopontine angle), and comprises both auditory and vestibular nerves. A schwannoma may develop on either of these but as it expands will involve both to cause balance disorder (often very non-specific) and a sensorineural deafness. Management is either bystereotactic radiotherapy or by surgical excision.

Suspect this as the cause in all cases of unilateral sensorineural deafness in adults. Although rare, it does occur in children.

Disorders affecting the cochlea (sensorineural deafness)

Disorders of the cochlea causing sensorineural deafness generally require specialist evaluation. The list of potential causes is included because, among other things, it illustrates the importance of preventive education.

Congenital (genetic). The incidence is about 1:1000 live births. Consanguinity results in a four-fold increase in the incidence. It may be dominant (family history), recessive (tend to be sporadic), or part of a syndrome (two examples are Waardenburg syndrome - deafness with patchy depigmentation, especially of eyes and the forelock of the hair - and Usher's syndrome - deafness with retinitis pigmentosa).

Congenital (acquired). Rubella is the most important of these causes, hence the importance of immunization of all girls before child-bearing age.

Birth asphyxia. There is a relationship to cerebral palsy, hence the need for good obstetric services.

Neonatal jaundice. The brain-stem auditory pathway is one of those that are damaged should kernicterus develop, hence the need for phototherapy and for blood replacement transfusion when billirubin levels rise.

The importance of these causes for deafness lies also in the need for early recognition - see *Deafness in children*, Chapter 5 - in order that rehabilitation may be started before one year of age for optimal speech and intellectual development.

Infantile meningitis. See under *Labyrinthitis.* The severity of deafness as a complication of meningitis can be reduced by the use of steroids. The two organisms most implicated are Haemophilus and Pneumococcus. It is hoped that the introduction of vaccination may help reduce the prevalence of this complication.

Infectious diseases of childhood. Mumps classically causes unilateral deafness, hence the importance of vaccination.

Ototoxic drugs. These drugs should be available only on prescription from a medical practitioner but in some parts of the world their use is still uncontrolled. Ototoxicity is an important reason for monitoring blood levels during intensive therapy. Salicylate-induced deafness is the only kind that is reversible. Ototoxic drugs include aminoglycoside antibiotics (for example gentamicin, streptomycin), quinine and derivatives, some of the diuretics, and salicylates.

Noise-induced hearing loss. Excessive noise damages hair cells - particularly those towards the basel end of the cochlea - hence the predominant deafness around 4000 Hz. Noise restriction and noise protection in the work place are important preventive measures.

Syphilis. This is included because it is about the only cause of sensorineural deafness that can be treated (amoxycillin and steroid) if recognized early. Deafness occurs during the early tertiary phase - **be aware of it in adults, particularly in the age range 40 to 60 years, presenting with sudden or rapid onset of either unilateral or bilateral sensorineural deafness.**

Age. Hair cell loss with ageing causes the predominantly high-tone sensorineural hearing loss known as presbyacusis. The disability of deafness adds to the burden of ageing but can often be helped by prescription of suitable hearing aids.

Disorders affecting the vestibular system (vertigo)

Almost all of the disorders of balance that present with vertigo require specialist evaluation. There are several causes for vertigo - see Figure 4.8.

Vestibular neuronitis. Neither the precise aetiology nor the pathology are known but the condition is best regarded as a viral neuritis (see also *Bell's palsy)* that causes either a neuropraxia or nerve degeneration in the vestibular nerve. These patients present initially with

acute vertigo requiring bed rest and sedation. This lasts for several days. If there is only neuropraxia, complete resolution should take place over the next few weeks but if there is nerve degeneration, the vestibular system will have to undergo central compensation and this takes several months.

Ménière's disease. Simplistically, this disorder is caused by an increase in the pressure within the endolymphatic compartment of the membranous inner ear. (This may have something to do with osmotic pressure - remember the difference in ionic composition between endolymph and perilymph.) As the pressure increases (endolymphatic hydrops) hair cell stimulation causes tinnitus (cochlea) and vertigo (vestibular) with sustained pressure causing hair cell damage (deafness). Predominantly affecting young to middle-aged adults, it is characterized by episodes that occur in clusters with periods of remission between clusters but steadily deteriorating hearing. The vertigo is managed with 'sea-sickness' preparations and sedatives, and the underlying disorder is treated with salt and caffeine restriction, vasodilators, and diuretics. In severe cases, surgical relief may be contemplated. Options include endolymphatic sac decompression, vestibular nerve section, or inner ear destruction (labyrinthectomy).

Benign positional vertigo. The hairs of vestibular hair cells are embedded in a cupola. These structures can fragment from either degeneration or head trauma and the particles released then float loose in the vestibular system. In certain head positions the particles may impinge on and stimulate hair cells. The resulting vertigo lasts only a short time (seconds to minutes) but tends to recur each time the head returns to the precipitating position. Treatment involves positional manoeuvres to try and get the particles to lodge somewhere out of harm's way.

Perilabyrinthitis/serous labyrinthitis. Middle ear infection, particularly when there has been bone erosion by cholesteatoma, may cause a 'neighbourhood' reaction in the labyrinth, manifesting with vertigo. Surgical exploration is required to eradicate infection.

Vertebro-basilar insufficiency (VBI). The vertebral artery travels through channels in the cervical vertebrae and in the osteo-arthritic spine it may be 'pinched', particularly with head extension. This is a common problem in the middle-aged and elderly, and usually manifests with 'drop attacks' or faintness, but this diminution of blood flow to the vestibular nuclei may cause vertigo. Treatment is with physiotherapy and use of a cervical collar.

Facial nerve palsy

Facial nerve palsy occurs at any age with a prevalence of about 1:5000 of the population per year. Of these about 80 per cent will be idiopathic (see below).

Upper motor neurone facial palsy occurs as part of a cerebrovascular accident affecting the motor cortex. The diagnosis should be obvious. (Elucidation of 'forehead sparing' can be misleading as often this is the first area to recover in a lower motor neurone facial palsy.)

The most common cause of a lower motor neurone facial palsy is idiopathic and this diagnosis is given the name **Bell's palsy.** The aetiology and precise pathology are unknown but it is most usefully considered as a neuritis which may manifest as either neuropraxia or

nerve degeneration. Typically the palsy is of sudden onset and there may be associated otalgia. The palsy may be partial or complete. It is unfortunately a diagnosis that has to be made by excluding all the other potential causes for a facial palsy. This is important because all of these require treatment, whereas an idiopathic palsy is usually left to recover on its own, although many specialists advocate the use of steroids. Full recovery within two to three months occurs with neuropraxia, but when there has been nerve degeneration, recovery is uncertain and may take up to a year.

Herpes zoster oticus is a variation on this disorder in which the causative agent is obvious, since herpes zoster vesicles (shingles) erupt in and around the ear canal indicating involvement of the geniculate ganglion of the facial nerve. This is sometimes known as Ramsay Hunt syndrome. Acyclovir is the current drug of choice.

The other causes for a facial palsy are differentiated by systematic consideration of the facial nerve anatomy; see Figure 4.9.

From the brain stem into the internal auditory canal:

- Trauma with avulsion (rare).
- Meningitis (rare, think of TB).
- Aberrant blood vessels (facial tic or spasm).
- Acoustic neuroma (rare).

Within the temporal bone adjacent to the inner ear:

- Trauma with fracture of the petrous portion of the temporal bone. This requires surgical exploration to either decompress or repair the nerve.

Within the middle ear and mastoid:

- Acute otitis media. This is usually seen in children when the treatment is IV antibiotics and drainage of the middle ear.

- Chronic otitis media (particularly cholesteatoma but think of TB).

- Mastoiditis.

- Trauma with fracture of the mastoid portion of the temporal bone.

These require surgical exploration to eradicate disease and to explore the nerve in case repair is needed.

Exit at the stylomastoid foramen into the parotid gland and then distributing to the facial muscle:

- Congenital trauma - within the birth canal the mandibular branch may be compressed (a not uncommon injury), forceps delivery.

- Other trauma - penetrating, blunt.

- Malignant neoplasm.

- Parotitis.

As a general rule, facial nerve palsy requires specialist evaluation as a matter of urgency (if such a service is available) for all causes other than idiopathic palsy.

5. A practical approach to ear disorders

Unless they have had it before or have been told what is wrong with their ear, patients don't usually come complaining of a particular disease. They usually come in wich a collection of symptoms and expect you to tell them what is wrong. A practical approach to ear disorders is largely based on elucidation of patient symptoms, followed up by examining their ears, since this combination should enable most diagnoses to be made. With external and middle ear disorders you cannot rely on the history/symptoms alone because, although to a certain extent history will help in differentiating between external and middle ear disorders, in the end you are going to have to look in the ear and test the hearing to make the diagnosis. On the other hand, history/symptoms are all-important with inner ear disorders since the only methods of examination are either by audiological evaluation or by radiology. Audiometry is essential when the inner ear disorder affects the cochlea (deafness) but vestibular disorders are predominantly diagnosed from just the history.

In this chapter the different disorders that could potentially be causing the symptoms are discussed. Once a diagnosis has been made, refer to Chapter 4, *Core knowledge of ear disorders*, in which each of the disorders is briefly discussed and guidelines for management of the disorder are given.

Symptoms

Symptoms arising from ear disorders are:

- Otalgia (pain)

- Otorrhoea (discharge)

- Impaired hearing and deafness

- Tinnitus

- Disequilibrium and vertigo

- Facial nerve palsy.

Otalgia

Otalgia (pain in the ear) usually arises from inflammation in either the ear canal or the middle ear. The history may provide clues but diagnosis relies largely on examination of the ear. When the appearance of these areas is normal, referred pain has to be considered either from an adjacent structure or one sharing the same innervation as the ear.

Potential causes

The potential causes for otalgia are described below.

External ear

Localized otitis externa - a furuncle. This should be suspected as the cause when there is tenderness on manipulating the pinna or tragal cartilage. Surprisingly, it is easily missed when inserting an otoscope speculum unless the entrance to the ear canal is first inspected.

Later stages of generalized otitis externa. In the early stages the complaint is often merely of an itchy ear, pain starting with the onset of a definite inflammation and swelling of the soft tissues of the ear canal.

Herpes zoster oticus (shingles). This can be a difficult diagnosis to make in the early stages before the vesicular eruption occurs. Since the pain can be intense, the patient may find it strange that you cannot see anything abnormal. The disorder becomes obvious when the vesicles erupt.

Impact wax or a foreign body in the ear canal. Patients usually experience discomfort rather than pain, unless they have been fiddling in the ear canal and have precipitated an inflammation.

Perichondritis. This is an uncommon, inflammatory swelling of the soft tissues of the pinna that may or may not be associated with an otitis externa. It may also be precipitated by trauma. It requires urgent, vigorous treatment, preferably with intravenous antibiotics, as the infection may spread to involve the cartilage with subsequent deforming contracture (cauliflower ear). Be particularly wary when it occurs in a diabetic or immunocompromised patient as the infection in these patients can rapidly spread into the temporal bone with potentially fatal consequences - necrotizing otitis externa.

Tympanic membrane

Bullous myringitis. This is one of the most painful ear disorders and is probably a viral infection that manifests with small blisters on the eardrum that may contain either serum or blood. It is probably the only indication for using ear drops containing a local anaesthetic.

Middle ear

Acute otitis media. In a typical case this should not be too difficult to diagnose - an irritable child with pyrexia and a sore ear who has an obviously inflamed eardrum. But the symptoms caused by an acute otitis media are very variable, which is why examination of the ears is an essential part of the general examination of an unwell child. Although most common in children, acute otitis media occurs at any age.

Developing mastoiditis. This is included as a separate entity from frank mastoiditis - which is a potential, although uncommon, complication of all middle ear disorders - because it should always be in the back of your mind when there is middle ear infection and otalgia. **In acute otitis media, persistent pain that has not settled once antibiotics have had a chance to work is a 'red flag' symptom.** Chronic otitis media is a painless disorder and the onset of pain is again a 'red flag' symptom. Early mastoiditis often responds to antibiotics alone, whereas established mastoiditis requires mastoid surgery, so that, if suspected, you should refer these patients urgently for a specialist opinion.

Mastoiditis. Once mastoiditis has become established, the diagnosis is much more obvious. The patient is pyrexial and obviously ill, and there is an inflammatory swelling behind the pinna. As mentioned, **mastoiditis is uncommon, but unfortunately it often arises in neglected ears and in such cases intracranial spread of infection may have already occurred by the time the patient comes to see you. You need to be aware of this possibility.**

Middle ear efusion. In children, this disorder is often associated with intermittent discomfort which is probably caused by pressure changes in the middle ear. These children may also present with a painful ear as they seem to be more prone to episodes of acute otitis media than children with normal middle ears.

Barotrauma. Anyone who has flown in an aircraft will be aware that a number of people experience intense pain in their ears during descent. During ascent, as the air pressure in the cabin falls, air from the middle ear passively passes down the eustachian tube to equalize the pressure. However, during descent, as the air pressure rises again, the eustachian tube has to be actively opened either by swallowing or by self-inflation - Valsalva manoeuvre - and not everyone is able to do this, particularly small children. If the pressure in the middle ear cannot be equalized, acute retraction of the eardrum causes intense pain. Similarly, during diving, the middle ear pressure has to be equalized every so often during descent. Persistent negative middle ear pressure may precipitate an acute middle ear effusion and sometimes this is accompanied by bleeding from the middle ear mucosa - haemotympanum.

Referred pain

The ear has a complex sensory innervation which includes, among others, branches from the 5th (trigeminal), 7th (facial), 9th (glossopharyngeal) and 10th (vagus) cranial nerves. These latter two also supply sensory innervation to structrues in the aerodigestive tract - it is not uncommon for patients to cough when you fiddle in their ears. Therefore anything irritating or causing inflammation of structures in the oral cavity, pharynx, or larynx may also cause pain to be felt in the ear.

Tonsillitis/pharyngitis and tonsillectomy. It is very easy to assume that otalgia is arising from these disorders but you have to bear in mind that there may be a co-existing otitis media. Always examine the ears to exclude this when otalgia is associated with these disorders.

Dental causes. Erupting teeth, impacted molars, caries, and dental-associated infection are quite common causes for referred pain when the ear is normal on examination.

Temporomandibular joint dysfunction. This is one of the most common causes for referred pain in adults, usually those with dentures. With a full set of natural teeth, no pressure is normally exerted on the articulating surfaces in the temporomandibular joint during mastication, as the upper and lower teeth grind together. When teeth are extracted the alveolar ridge gradually becomes re-absorbed. Dentures may fit well soon after extraction but, after some time, begin to 'overclose' during mastication, exerting pressure on the joint surfaces. This may cause pain which, since the joint is right next to the ear, is experienced as an earache. The management is referral back to the dentist to have the dentures adjusted.

Pharyngeal and laryngeal neoplasia. Look up *The ENT Ten,* and you will see that this is number 7, a definite 'red flag' symptom. Never forget it, as it may make the difference between referral early enough to offer the patient a reasonable chance of cure and late referral with a poor prognosis.

Cervical osteo-arthritis. A number of people with this disorder - and remember that the process starts in the 40s - have referred otalgia among the other symptoms caused by it.

Treatment

Once a diagnosis of the cause for the otalgia has been made, the appropriate treatment can be initiated (see Chapter 4, *Core knowledge of ear disorders,* and Table 5.1).

Table 5.1 Otalgia

Ear canal:

Furuncle - localized otitis externa
Inflamed/swollen - generalized otitis externa
Normal - examine drum

Eardrum:

Inflamed/bulging - acute otitis media
Blister - bullous myringitis
Dull/retracted - otitis media with effusion
Active perforation - potential complications
Normal - referred pain.

Otorrhoea

Otorrhoea (discharge) originates from infection in either the external ear canal or within the middle ear cavity. A history is helpful but diagnosis relies largely on examination of the ear. Discharge from otitis externa is usually scanty and initially associated with itching or discomfort. The middle ear is mucosal lined and the discharge of chronic infection is usually both mucoid and profuse although cholesteatomatous disease may produce only a scanty, smelly discharge. Acute otitis media usually presents with otalgia, which is often relieved when the drum perforates to produce a discharge. Discharges also include bleeding and this is usually related to trauma, either local within the ear canal or from fractures of the base of skull.

Potential causes

The potential causes of otorrhoea are described below.

External ear

Otitis externa. The localized form is a furuncle in the hair-bearing area which, when it points and bursts, will discharge blood-stained pus. The generalized form is an inflammation of the ear canal skin. When this weeps it will produce a pus discharge into an ear canal which is often full of debris-containing desquamated epithelium and wax. With some fungal infections the appearance of the debris has been likened to wet newspaper.

Middle ear

Acute otitis media after perforation. Typically there will have been a pyrexial illness with a sore ear which then 'bursts' to discharge blood-stained mucopus. Surprisingly, in infants not all of the precipitating episodes of otitis media seem to be particularly painful.

Active chronic otitis media. In this case, ears known to have a perforation will begin to discharge either after an episode of upper respiratory tract infection or after water has got into the ear. The discharge is usually profuse and mucopurulent. Many patients with this disorder seem to be particularly neglectful of their ears and the discharge may have been going on for quite some time before they seek medical attention. If the discharge is particularly malodorous, suspect underlying cholesteatoma. Cholesteatoma should also be suspected when the infection fails to respond to treatment.

Previous mastoid surgery and an infected cavity. Scars behind the ear will indicate that either mastoid surgery or reconstructive middle ear surgery (tympanoplasty) has been performed - it is surprising how many patients do not know what type of surgery they have had! It is easy to overlook a mastoid cavity when examining the ear unless the posterior wall of the ear canal is specifically examined. Wax commonly accumulates in these cavities and they are often the site of chronic infection.

Ear trauma and head injury

Bleeding is indicative of either local trauma in the canal, perforation of the tympanic membrane, or a fracture of the base of the skull. This latter is often associated with a CSF leak which creates a 'halo' sign on the ear dressing.

Blood-staining on the cotton wool used to dry mop a discharging ear (see Chapter 14) usually arises from granulation tissue associated with prolonged infection.

Treatment

Once the cause for otorrhea has been established, the appropriate treatment can be initiated (see Chapter 4, *Core knowledge of ear disorders*, and Table 5.2).

Table 5.2 Otorrhoea

Ear canal:

Furuncle - localized otitis externa
Inflamed/swollen - generalized otitis externa
Mastoid cavity - infected mastoid cavity
Normal - examine eardrum

Eardrum:

Short history/fresh perforation - acute otitis media
Longer history/active perforation - active chronic otitis media
Fleshy polyp - active chronic otitis media (cholesteatoma).

Impaired hearing and deafness

Deafness in children is a special topic and will be dealt with next.

Note that the terms 'hearing impairment' and 'deafness' are not strictly synonymous although they are used very loosely. A hearing impairment should refer to mild/moderate hearing loss and deafness to severe/profound hearing loss.

When a patient presents with deafness you want to know not only the cause of the deafness but also the degree of handicap experienced because this may be a determinant factor in management. Not all patients with deafness want or warrant surgery or hearing aids. Potential causes are listed below, some with characteristic features in the history, others with characteristic examination findings or audiometry. It is useful to remember to ask in what situations they experience difficulty as often patients with a conductive deafness are less handicapped in noisy environment (they don't hear the background noise and people tend to speak louder in such situations), whereas it is the opposite for patients with a sensorineural deafness.

Examination of the ear should reveal most of the causes for conductive deafness. Response to voice testing should give you some idea as to the degree of hearing loss, and you should determine whether the deafness is a conductive or a sensorineural loss (response to tuning fork testing). Once deafness is confirmed, referral for audiological evaluation and specialist management is required.

Potential causes

The potential causes described below cover all three parts of the ear.

External ear

Impacted wax or foreign body. These problems will only cause deafness when the entire ear canal is obstructed. The deafness should be relieved when the wax or foreign body is removed.

Occlusion by swelling - infection. Again, deafness is noticed only when the canal is obstructed by either the swelling or the discharge/debris accumulation. Once the disorder has been treated and has resolved, the deafness should be relieved.

Congenital or acquired stenosis. Not all deformed pinnas are associated with complete stenosis of the canal and conversely there may be a complete canal stenosis with a normal pinna.

Tympanic membrane

Perforation. The degree of hearing impairment usually relates to the size of the perforation. This is not always the case, however, as there may be middle ear adhesions or disruptions of the ossicular chain as part of whatever caused the perforation.

Tympanosclerosis with middle ear involvement. The process causing tympanosclerosis in the eardrum (chalk patches) may also extend to involve the middle ear mucosa and impinge on the ossicular chain.

Scarring with middle ear adhesions. Scarring is indicative of healed otitis media and there may be adhesions or disruption of the ossicular chain as well.

Middle ear

Middle ear effusion. This is the most common cause of hearing impairment in children - otitis media with effusion - but it does occur at any age after an episode of otitis media. In adults, it may be associated with barotrauma, usually after flying in an aircraft.

When the effusion is unilateral, especially in adults, pathology in the nasopharynx always has to be excluded.

Active chronic otitis media. There are several causes for deafness in this disorder - the perforation, mucopus in the middle ear, inflammatory swelling of mucosa around the

ossicles, or osteitis causing ossicular chain disruption. When the underlying cause is cholesteatoma, this disease destroys the ossicles.

Inactive chronic otitis media. Here again the deafness may arise from the perforation or the end result of middle ear disease - adhesions or disruption of the ossicular chain.

Ossicular chain discontinuity. Infection and cholesteatoma have been mentioned earlier, but trauma may also be a cause.

Ossicular chain fixation. Tympanosclerosis and fibrosis have been mentioned earlier, but consider otosclerosis as well.

Inner ear

Congenital. The usual classification of congenital disorders apply also to deafness. Genetic causes may be familial or sporadic, dominant or recessive, syndromic or non-syndromic. Congenital deafness may be the result of a disorder in utero (acquired), of which the most common disease is rubella.

Natal causes usually relate to asphyxia or intracerebral haemorrhage. Children who have experienced these disorders usually have some degree of cerebral palsy or retardation.

Neonatal. The neonatal disorder that causes deafness is kernicterus. However, when neonates require admission to neonatal intensive care units they are exposed to a variety of causes for deafness, apart from their underlying illness, of which the most likely cause is ototoxic drugs. All survivors of neonatal intensive care should be regarded as being at risk for deafness and their hearing should be assessed as soon as possible after discharge from the intensive care unit.

Infection. Meningitis is the most likely infection to cause deafness. Mumps classically causes a unilateral deafness. Never forget the possibility of syphilis as a cause for a recent-onset sensorineural deafness in an adult as it is the only one in which the deafness may recover after treatment if it is initiated promptly. The deafness usually comes on fairly suddenly, some 20 to 30 years after the infection was acquired.

Noise exposure. This is diagnosed from the typical 'dip' in the audiogram at 4000 Hz.

Ototoxic drugs. Salicylate-induced deafness is reversible when the drug is stopped, but the others - aminoglycosides, quinine, cytotoxics - are not.

Ménière's disease. Deafness is one of the triad of symptoms - vertigo, tinnitus, deafness - that characterize this disorder, but the deafness does not always recover after each episode so that there will be a progressive sensorineural deafness.

Trauma. Disruption of middle ear structures has been mentioned earlier. The inner ear may be disrupted when base of skull fractures involve the temporal bone. The deafness does not usually recover. There is also a state known as 'inner ear concussion' when there is no demonstrable fracture, from which recovery may occur.

Tumour - acoustic neuroma. Although rare, this is one disorder otologists always consider when there is a unilateral sensorineural deafness because, if untreated, the tumour will eventually expand and compress the brain stem with fatal consequences.

Age - presbyacusis. This is by far the most common cause for sensorineural deafness and affects a large proportion of any ageing population. Typically, the deafness is bilateral and on audiogram will be seen to predominantly affect the higher tones.

Treatment

Once the diagnosis of the cause of deafness has been determined and the degree of handicap evaluated appropriate management can be initiated.

Surgery. Conductive losses are often corrected with appropriate surgery, for example, placement of ventilation tubes for middle ear effusions, or tympanoplasty with or without ossiculoplasty for inactive chronic otitis media. This is not always possible. For example, surgery for cholesteatoma is often destructive rather than reconstructive (radical mastoidectomy), and congenital deformities may not be amenable to surgery and hearing aids may need to be fitted as well as, or instead of, surgery.

Hearing aids. There are also many patients with conductive losses who do not want surgery and would prefer to use a hearing aid. In practice, patients with conductive losses generally derive greater benefit from a hearing aid than patients with sensorineural losses, since their problem is simply one requiring amplification and they do not have the element of distortion that often accompanies sensorineural deafness.

When the handicap of deafness is due to a sensorineural loss, hearing aids are needed. A wide range of hearing aids are available - body-worn aids, postaural aids, in the ear canal aids, spectacle aids. Many sophisticated devices may be built into a hearing aid to make it better able to overcome a speicific handicap, such as high-tone or low-tone cut-off, high-tone or low-tone gain, or to make them more acceptable and easier to use, such as directional microphones, automatic cut-off when the sound level exceeds a certain threshold. Of course the cost factor has to be considered - the more sophisticated the aid, the more expensive it will be.

Provision of a hearing aid should be backed up by audiotherapy which teaches the deaf patient all the added 'tricks' to overcome the handicap - making use of lip reading and body language are examples. This is particularly important when the deafness is in the severe to profound range, when even a powerful hearing aid does not restore the hearing threshold to anything like normal.

Cochlear implant. When the deafness is total or near total, the patient may be a candidate for a cochlear implant. This is a very sophisticated device that converts sound into an electrical stimulus which is applied to the auditory nerve via an electrode implanted into the cochlea.

Often diagnosis of a specific cause for deafness requires specialist expertise. Decisions regarding surgery or prescription and provision of appropriate hearing aids fall largely into

the realm of ENT and audiological specialists. However, primary care practitioners need to be aware of what is available and when and where to direct their patients with a deafness problem or to counsel them when they are uncertain as to aspects of their problem.

Deafness in children

Normal hearing is a fundamental requirement for the development of speech in infancy since speech develops as a mimic of sounds heard - if you want to know what type of sounds deaf people are hearing, listen to their speech - it tells you how they are hearing your speech. Hearing impairment causes both delay in speech development and distortion of the quality of the speech that does develop. A more severe degree of deafness results in no speech development at all (deaf and dumb). There is a limited time span in children's development for optimum acquisition of speech and if they are not speaking by the age of five to six years it is unlikely that they will develop intelligible speech thereafter. Hearing is essential for the acquisition of vocabulary since we learn the words first and then give them meaning. Vocabulary is the basis for all learning and, because we think in words, for intellectual development as well. Since speech and hearing are the most rapid forms of communication, the ability to communicate will be impaired by deafness.

For all of these reasons it is essential that deafness in an infant is detected as early as possible, preferably before one year of age, in order that a rehabilitation programme can be commenced to aid speech and intellectual development.

As a general rule, parents will become aware of hearing problems in their baby early on and it is essential that their opinions are heeded (not ignored or brushed aside) and that these infants are referred for evaluation.

Potential causes

Apart from congenital causes, other infants likely to have hearing problems are those with ear or craniofacial deformities, those who have had meningitis, and those who have required intensive care - the 'high-risk register' for infant deafness.

Testing of hearing in babies and infants is a highly specialized skill and in primary case it is best to question parents to gain an idea as to whether or not there is a hearing impairment. See Table 5.3.

Table 5.3 Questions for use in assessing hearing in babies

Baby only a few weeks old:

Does your baby appear to be listening to you when you talk or sing?

Does your baby open his or her eyes or blink when there is a noise?

Baby about six months old:

Does your baby enjoy your talking or playing word games with him or her?

Does your baby try to see where a noise is coming from by turning his or her eyes or head towards the sound?

Baby about nine months old:

Does your baby appear to enjoy babbling and making other noises?

Does your baby appear to respond to even very soft sounds?

Baby about a year old:

Is your baby beginning to say baby words?

Does your baby respond when you say his or her name and the names of things he or she plays with.

Baby about a year and a half old:

Is your baby beginning to use simple words?

Does you baby pick up or point to things around the house when you ask him or her to do this?

Babies of two years or more:

Is your baby putting words together and trying to talk to you?

Do you think your baby can hear normally even when you speak to him or her in a very soft voice?

Treatment

In childhood, conductive causes for hearing impairment are common, particularly middle ear effusions. At this level of hearing loss there is not the same urgency but a persistent loss over several months will begin to impair speech quality, vocabulary progression and intellectual development. Behaviour problems frequently accompany middle ear effusions and these children are often clumsy. These children need referral for assessment as not only are conductive losses remediable with surgery, for example ventilation tubes, but sometimes a sensorineural hearing loss is mistaken for a conductive loss and in these cases provision of hearing aids may be more appropriate.

Tinnitus

Tinnitus is a subjective ringing or buzzing sound in the ear. To understand tinnitus, think of the auditory system as simply a hair cell with a neurone connecting it to the auditory centre - see Figure 5.1. Any impulse generated in the neurone in such a system will be interpreted by the auditory centre as a sound. Normally such an impulse is only generated by

the hair cell in response to external sounds transmitted via the ear canal, middle ear, and cochlea. Tinnitus arises when impulses are generated by other mechanisms.

Potential causes

The approach to tinnitus involves detection of any of the causes described below from the history and examination. This should be supplemented by audiological investigation to detect any deafness associated with either hair cell loss or neurone impairment. Possible causes are:

Stimulation of the hair cell. This may be internal sounds not normally heard, such as vascular pulastions in the carotid artery or in a vascular middle ear tumour, or it could be from movement of muscles in the middle ear, such as the tensor tympani muscle which attaches to the malleus.

Other types of stimulation. Sometimes an inner ear disorder may stimulate the hair cell by some mechanism other than sound vibration. The pressure changes in the inner ear fluids that occur in Ménière's disease can to this, as can irritative disorders such as labyrinthitis, ototoxics (particularly salicylates), or sensitivity to stimulants such as caffeine.

Damaged hair cell. Neurones connected to sensory organs are spontaneously excitable - by this we mean that they are capable of producing spontaneous, but random, action potentials. Normally the rate at which these are produced is controlled by the sensory organ. In the cochlea this is the hair cell. If the hair cell is damaged (noise trauma, ototoxics) or lost (ageing) these now uncontrolled spontaneous action potentials - impulses - will continue to be produced and eact will be interpreted by the auditory centre as sound.

Irritation. If something irritates the nerve (acoustic neuroma or an abnormal blood vessel) spontaneous discharges may be generated that will also be interpreted as sounds.

Treatment

Removal of causative factors or treatment of the underlying disorder may resolve the problem. When it doesn't, an explanation to the patient will often relieve anxiety and render the tinnitus more tolerable. There are also simple things a patient can do that may help. Tinnitus tends to be masked by external noise (which is why it is usually worse in quiet surroundings) so that having music playing or a radio on is often helpful (sleep buttons on clock radios). Provision of a hearing aid is a way of increasing the external sound input when tinnitus is a complication of deafness. Sophisticated masking devices based on hearing aids are available for really troublesome tinnitus. Where a tinnitus clinic exists, this is a useful resource for referral.

Disequilibrium and vertigo

The brain-stem vestibular centres co-ordinate input into the balance system from the vestibular organs in the two ears, from the eyes, and from joints and muscles in the cervical spine and the limbs. The cerebellum then co-ordinates body movements in response to these sensory inputs. The cerebrum probably exerts some overall control but is primarily where

awareness of balance is centred, the sensation of disequilibrium resulting from a faulty or contradictory input from one or more of the sensory input systems.

Classically, symptoms are divided into either true vertigo (a sensation of rotation or movement often accompanied by nausea) or other symptoms such as lightheadedness (a feeling of fainting), unsteadiness or imbalance (a tendency to fall or feer to one side), and blackouts. This division is made because these other symptoms result from disorders such as hypotension, cardiac arrhythmias, epilepsy, other neurological disorders, and the ageing process with its gradual deterioration in co-ordination (see Figure 5.2), whereas vertigo is usually caused by vestibular disorders (see Figure 5.3).

Potential causes

In ENT we are concerned with disorders causing vertigo. The approach to these disorders primarily involves the history, since examination and investigation add very little further information. There are a limited number of disorders that cause true vertigo, each of which has characteristic features - see Chapter 4. The most useful way of distinguishing between them is from the history regarding the duration of the vertigo.

- Lasts seconds - benign positional vertigo. Floating particles in the vestibular system impinge on and stimulate the vestibular hair cells in certain repeatable head positions.

- Lasts hours - Ménière's disease. Symptom triad of tinnitus, vertigo, and fluctuating deafness. The vertigo is episodic and the episodes typically occur in clusters with periods of remission in between when deafness may recover to some extent.

- Lasts days - vestibular neuronitis. This disorder is probably a viral neuritis affecting one of the vestibular nerves which, by interfering with input into the balance system from one vestibular organ, precipitates acute onset of vertigo which then only gradually settles.

Vertebro-basilar insufficiency. This is caused by cervical osteo-arthritis and interruption of vertebral artery blood flow during cervical spine movements. It is diagnosed from the history regarding the effects of head extension or rotation in precipitating the vertigo.

Concomitant middle ear disease. Examination is most beneficial in detection or exclusion of concomitant middle ear disease that may be the cause for the vertigo:

- Effusions often cause clumsiness and unsteadiness.

- Active chronic otitis media can precipitate a 'neighbourhood' perilabyrinthitis. When this is the cause, it is often because cholesteatoma has eroded the bony capsule of the lateral semicircular canal to expose the membranous labyrinth to the effects of inlammation and infection in the middle ear.

Always think to exclude acoustic neuroma when disequilibrium/vertigo is associated with unilateral deafness and tinnitus.

As far as investigations are concerned, 'positional testing' may aid the diagnosis of cervical osteo-arthritis or benign positional vertigo; audiometry may provide clues in Ménière's disease and acoustic neuroma and caloric testing may identify the affected nerve in vestibular neuronitis.

Treatment

Having arrived at a provisional diagnosis, the vertigo will need treatment and the most beneficial is a combination of sea-sickness preparations, sedation, and bed rest. Thereafter these patients will usually require specialist referral for evaluation and further management.

Facial nerve palsy

Upper motor neurone facial palsy occurs as part of a cerebrovascular accident affecting the motor cortex and the diagnosis should be obvious. Elucidation of 'forehead sparing' on its own can be misleading as often this is the first area to recover in a lower motor neurone facial palsy.

Potential causes

The most common cause of a lower motor neurone facial palsy is idiopathic and this diagnosis is given the name 'Bell's palsy'. It is unfortunately a diagnosis that can only be made by excluding all the other potential causes for a facial nerve palsy. Apart from diagnosing idiopathic facial palsy, it is important to do this because all of these other causes of facial nerve palsy require treatment, whereas an idiopathic palsy is usually left to recover on its own. Many specialists, however, advocate the use of steroids.

The other causes are differentiated by systematic consideration of the facial nerve anatomy - see Chapter 4. Clinically, take a history concerning possible recent head trauma, past ear disorder, current otalgia, deafness, tinnitus, or otorrhea and then examine the ear and the parotid gland. All of these other causes for facial palsy require ENT specialist referral.

Treatment

Having excluded these disorders and arrived at a diagnosis of Bell's palsy (most usefully considered to be a viral neuritis causing either neuropraxia or nerve degeneration) further consideration is necessary. Clinically, the palsy may be incomplete or complete. Incomplete palsies are indicative of neuropraxia and all recover within a month or two. Complete palsies may result from either neuropraxia or nerve degeneration. In this situation specialist referral is needed in order to distinguish between these states. This is done by observing the response to simple nerve stimulation and comparing the threshold stimulation required to elicit a muscle twitch in both the affected nerve and the nerve on the unaffected side. If both thresholds are similar, then the nerve is in a state of neuropraxia and will recover within two to three months. If there is no muscle twitch response from the affected nerve to the highest tolerated stimuluts, the nerve has undergone nerve degeneration. When this is the case, only 50 per cent of such patients will regain an acceptable recovery of facial movement, and this will take 6 to 12 months.

For those patients who do not regain acceptable facial movement, cosmetic surgery (face lift) may help. Eye closure is a problem with exposure keratitis as a complication. In this situation the eyelids have to be partially sutured together to protect the cornea. Sometimes nerve anastomosis is considered. Cross-face nerve grafts from the normally functioning facial nerve on the opposite side or hypoglossal-facial nerve anastomosis are the current options.