The Ear and Associated Structures: Part I

36

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36.1 Applied Anatomy and Physiology

The ear consists of the external, middle and internal ear. The external and middle ear are mainly concerned with the transference of sound to the internal ear, where energy from the sound waves is processed into electrical signals for the brain. In addition to this function, the inner ear also contains the "organ for equilibrium" i.e. the balance organ. Although the tympanic membrane separates the middle ear from the external ear, the middle ear communicates with the external environment (through the nasopharynx) via the pharyngotympanic (Eustachian) tube (Fig. 36.1).

36.1.1 External Ear

This comprises the pinna, external auditory meatus (EAM), external auditory canal (EAC) and the tympanic membrane (ear drum). During embryonic development, six small prominences ("auricular hillocks") appear in the mesodermal tissue around the first pharyngeal cleft, three from the first pharyngeal arch and three from the second. The first arch tubercles form the tragus, helix and crus of the helix. The tubercles from the second arch form the antihelix, antitragus and lobule of the ear. This partly explains the complex innervation to the auricle, as it receives branches from both CN V and CN VII (as well as CN IX, CN X and the cervical nerves C2 and C3). Abnormal development of these prominences can result in microtia and various other anomalies, which may be an indication of malformation in the middle ear (Fig. 36.2).

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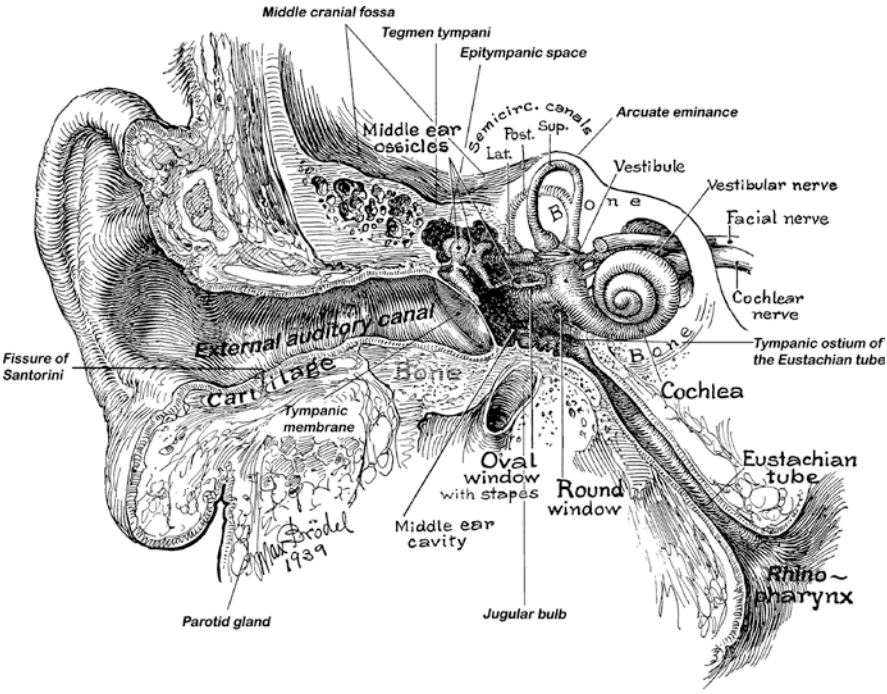

Fig. 36.1 Schematic diagram showing the coronal view of the temporal bone and ear structures

In the adult, the cup-like pinna (auricle) directs sound waves into the external auditory meatus (canal), where they are then conducted to the tympanic membrane. The pinna is comprised mostly of cartilage, covered by skin. Bleeding following an injury may produce a localised collection of blood between the perichondrium and the deeper cartilage—an auricular haematoma. If large, this can compromise the blood supply to the cartilage, resulting in fibrosis and the development of the well known "cauliflower ear". Since the pinna does not contain a subcutaneous layer of fat, it is susceptible to frostbite if exposed to the cold for too long.

The cartilage of the pinna is folded into crescences and valleys. The concha is the deep central depression in the pinna. The helix is the elevated peripheral margin. The non-cartilaginous lobule (ear lobe) consists of fibrous tissue and fat and is a common site for piercings (and subsequent infection or keloid formation). The tragus is the small projection which partially overlaps the external acoustic meatus opening. The main sensory nerves to the skin are the great auricular and auriculotemporal nerves. Lymphatics drain into the superficial parotid lymph nodes, mastoid lymph nodes and the deep and superficial cervical lymph nodes. These sites need to be carefully assessed with suspected cancers of the pinna. Tumours can arise on any of these sun-exposed areas, especially the superior helix.

The external acoustic meatus (EAM) passes approximately 2–3 cm (in adults) through the tympanic part of the temporal bone, towards the tympanic membrane.

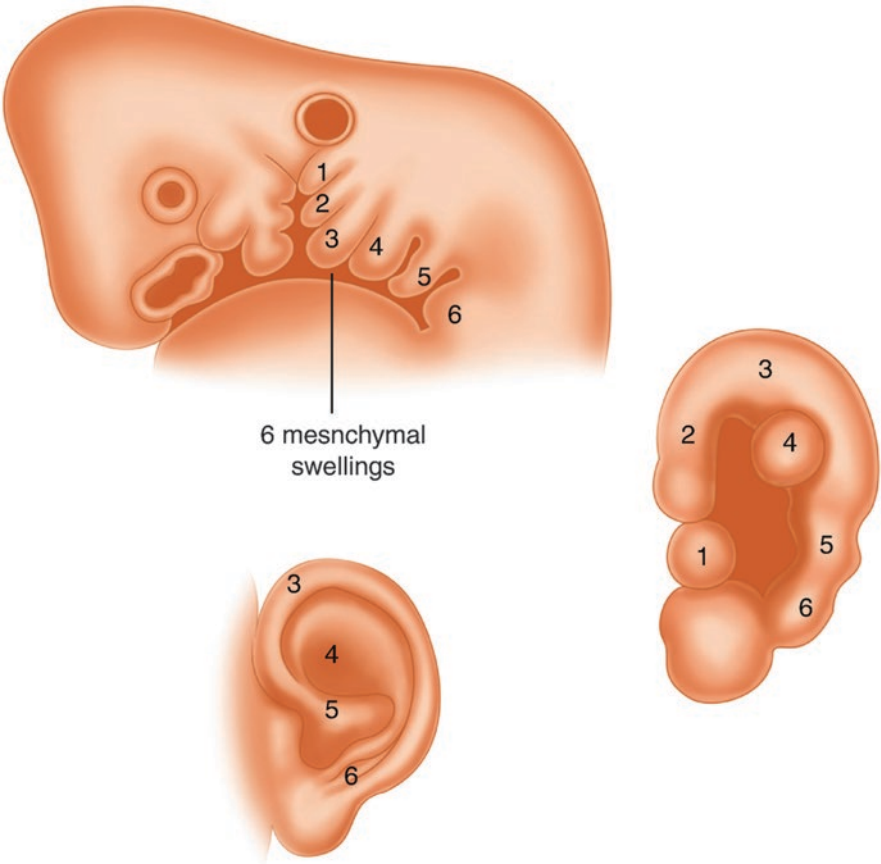

Fig. 36.2 Development of the outer ear

This develops from the ectoderm of the first pharyngeal cleft. Thus the outer lining of the meatus and tympanic membrane are derived from the ectoderm of the first pharyngeal cleft. The inner lining of the tympanic membrane comes from the endoderm of the first pharyngeal pouch (tubotympanic recess). The mesoderm sandwiched between these two layers, forms the fibrous layer of the tympanic membrane. The outer third of the EAM is S-shaped and cartilaginous and is lined with thin skin. Its inner two thirds is bony and lined with thinner skin. Ceruminous and sebaceous glands in the skin of the outer third produce cerumen (earwax). This skin is self-cleansing and contains hair follicles. However, these are absent along the inner part of the canal. Ears are normally self-cleansing and the use of cotton buds should be discouraged. If wax is dislodged into the deeper part of the canal, during attempts at cleaning, it can not be removed by this natural process. Overtime this can become impacted. The external auditory canal mucosa contains exocrine glands, modified apocrine glands and sebaceous glands. Therefore this can be a site for development of an adenoma, such as pleomorphic adenomasl. Apocrine adenomas (ceruminoma)

may also arise in the external auditory canal. These lesions, when large enough, can cause remodelling and expansion of the bony external auditory canal. The lymphatic drainage of the EAC is directed to the periauricular and parotid lymph nodes.

The tympanic membrane is approximately 1 cm in diameter. This is a thin, oval semitransparent membrane which forms a partition between the external acoustic meatus and the tympanic cavity of the middle ear. Embryologically it is derived from the first pharyngeal membrane which persists as an elastic fibrocartilagenous structure, covered by a thin layer of tightly bound skin. When viewed through an otoscope, a healthy membrane appears concave toward the external acoustic meatus with a shallow, cone-like central depression. The edge of the membrane is thickened and attached to a groove in an incomplete ring of bone, the 'tympanic annulus'. This almost completely encircles the membrane and holds it in place. An uppermost small area of the membrane where the ring is open is called the flaccid part (pars flaccida). The remaining larger part is called the tense part (pars tensa) (Fig. 36.3).

Histologically, the membrane consists of three layers (1) an outer layer of skin which is continuous with that of the external canal, (2) an inner layer of mucous membrane continuous with the lining of the tympanic cavity of the middle ear and (3) an intermediate layer between these, which is made of fibrous tissue. These fibres are arranged in a circular and radial pattern which gives the membrane its stiffness and tension. The tympanic membrane moves in response to air vibrations from the external acoustic meatus. These movements are transmitted via the chain of auditory ossicles through the middle ear to the internal ear. The membrane is innervated by several cranial nerves (1) the external surface by CN V and CN X, (2) the internal surface by CN IX. It is extremely sensitive to touch, pain and temperature (via CN V3 and CN IX). The external surface of the tympanic membrane is supplied mainly by the auriculotemporal nerve, a branch of CN V_3. This is important in the diagnosis of Ramsay-Hunt syndrome.

36.1.2 Congenital Anomalies of External Ear

1. Anotia (absence of the auricle): This occurs due to failure of mesenchymal proliferation to form the auricular hillocks. It is rare but commonly associated with first arch syndrome.
2. Otocephaly. Due to a failure in the development of the mandible, the ears fuse in the midline of the neck.
3. Preauricular appendages and pits: These are skin tags and shallow depressions that are usually seen in front of the ear. Preauricular appendages occur as a result of the development of accessory hillocks. Preauricular pits occur secondary to abnormal development of the hillocks. Most anomalies of the external ear are associated with various chromosomal syndromes such as Down's syndrome (trisomy 21), Patau's syndrome (trisomy 13), and Edward's syndrome (trisomy 18) (Figs. 36.4 and 36.5).
4. Atresia of the external auditory meatus: This occurs due to failure of the first pharyngeal cleft to canalise. Clinically this presents as conduction deafness and is often associated with first arch syndrome

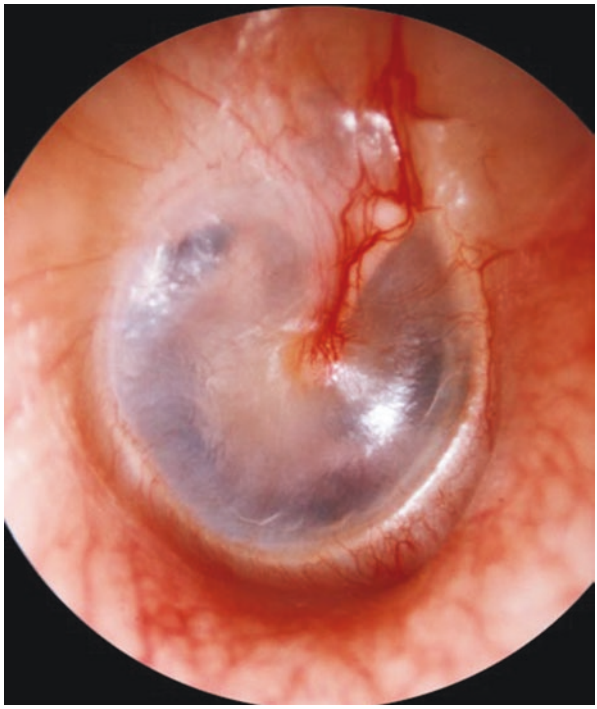

Fig. 36.3 Oto endoscopic view of the Right tympanic membrane with leash of vessels over handle of malleus. Note the light reflex at antero-inferior quadrant

5. Protruding Ears (Bat Ear). This is seen when there is an increase in the distance from the helical rim to the mastoid. It is thought to be due to a lack of the antihelical fold and prominence of the conchal bowl.

36.1.3 Preauricular Sinuses (Congenital Auricular Fistula, or a Geswein Hole)

These are relatively common congenital malformations characterised by a nodule, dent or dimple located anywhere adjacent to the external ear. They most often appear unilaterally, but are seen bilaterally in 25–50% of cases. They result from developmental defects of the first and second pharyngeal arches and can sometimes be associated with renal anomalies (branchio-oto-renal syndrome). Occasionally a sinus or cyst can become infected, but are often asymptomatic. Surgical excision is indicated if there have been recurrent infections.

36.1.4 Middle Ear (Tympanic Cavity)

Embryologically the middle ear, mastoid antrum, mastoid air cells and the inner lining of the tympanic membrane develop from a recess within the first pharyngeal pouch—the tubotympanic recess (with a small contribution from the second

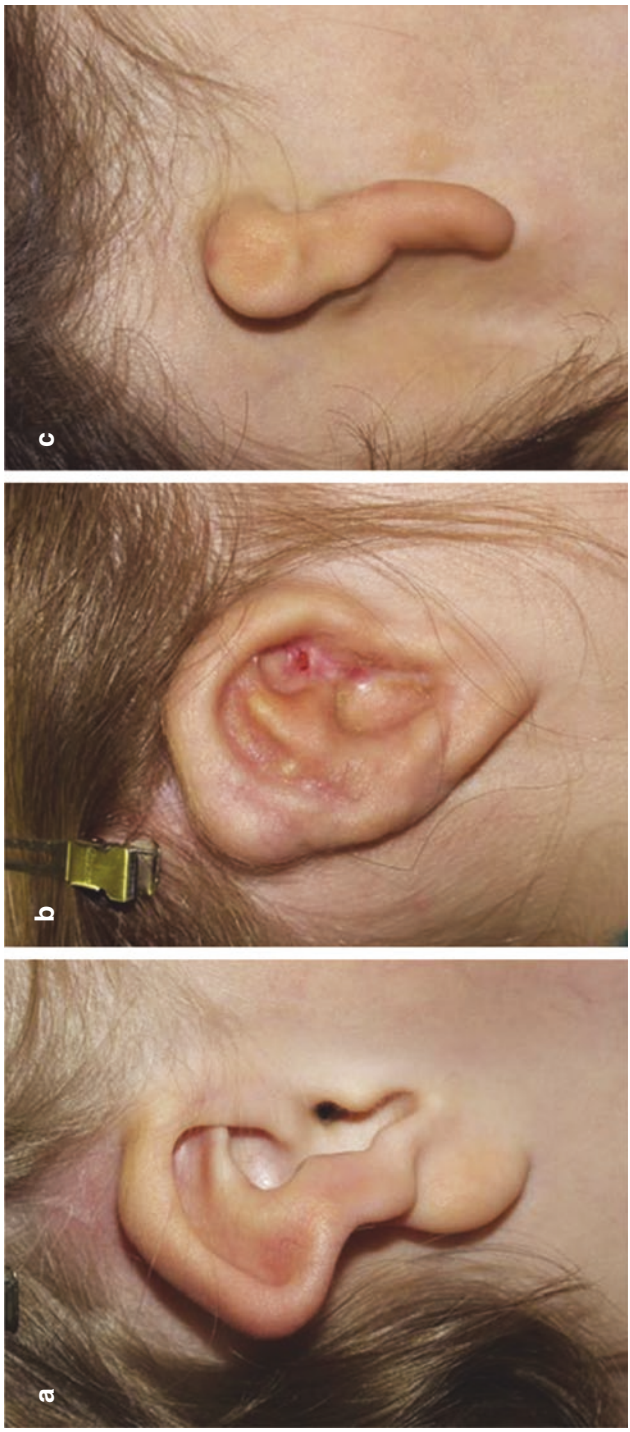

Fig. 36.4 (**a**) Minor auricular malformation (in a case of Goldenhar syndrome). (**b**) Middle grade auricular malformation. (**c**) Severe microtia

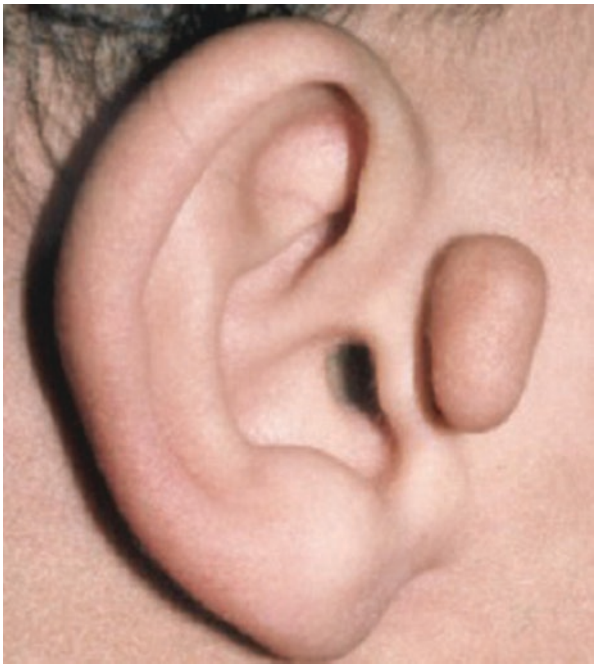

Fig. 36.5
Accessory auricle

pharyngeal pouch). Medially, the tubotympanic recess narrows and forms the auditory (Eustachian) tube (Fig. 36.6). The malleus and incus develop from the cartilaginous bar of the first arch (Meckel's cartilage) while the stapes develops from the second arch cartilage (Reichert's cartilage). The ossicles are initially embedded in mesenchyme, but as this breaks down, the epithelial lining of the developing cavity connects the ossicles to the wall of the cavity in a mesentery-like fashion. Supporting ligaments develop later within these mesenteries. The tensor tympani develops from the first arch and is innervated by the mandibular nerve—the nerve of the first arch. The stapedius muscle develops from the second arch and is thus innervated by the facial nerve—the nerve of the second arch. A posterior extension of the middle ear cavity subsequently forms the mastoid antrum. This is almost adult size at birth, however mastoid air cells are not present in the newborn infants. The mastoid air cells begin to develop at the age of 2 years.

In the adult the middle ear is an air-containing chamber in the petrous part of the temporal bone, which facilitates sound transfer to the cochlea. It is shaped like a box with concave sides and six walls. The cavity has two parts (1) the tympanic cavity proper—the space directly deep to the tympanic membrane and (2) the epitympanic recess—a space superior to the membrane. The tegmen tympani is the bony roof of the cavity. This separates it from the dura of middle cranial fossa. The bone here is thin and thus fractures involving the middle cranial fossa can result in CSF leakage through the ear (CSF otorrhoea). The tympanic cavity is closely related to the brain (middle cranial fossa), jugular bulb (posteriorly) and labyrinth (medially). Infections in middle ear are therefore potentially very serious as they can extend into surrounding structures. The cavity is lined with a mucous secreting membrane. The middle

ear thus contains (1) three ossicles (malleus, incus and stapes), (2) two muscles (tensor tympani and stapedius), (3) part of the facial nerve (chorda tympani branch) and (4) the tympanic plexus of nerves.

The mastoid antrum is a cavity in the mastoid process of the temporal bone. It is also separated from the middle cranial fossa by the thin tegmen tympani. The mastoid air cells are numerous small intercommunicating cavities within the mastoid process of the temporal bone that empty into the mastoid antrum. These are also lined by mucous membrane. Lymphatic drainage of the petrous bone, middle ear, inner portion of the external canal, and mastoid air cells can be either superficial to periauricular and parotid nodes or deep to the retropharyngeal lymph nodes. Mastoid

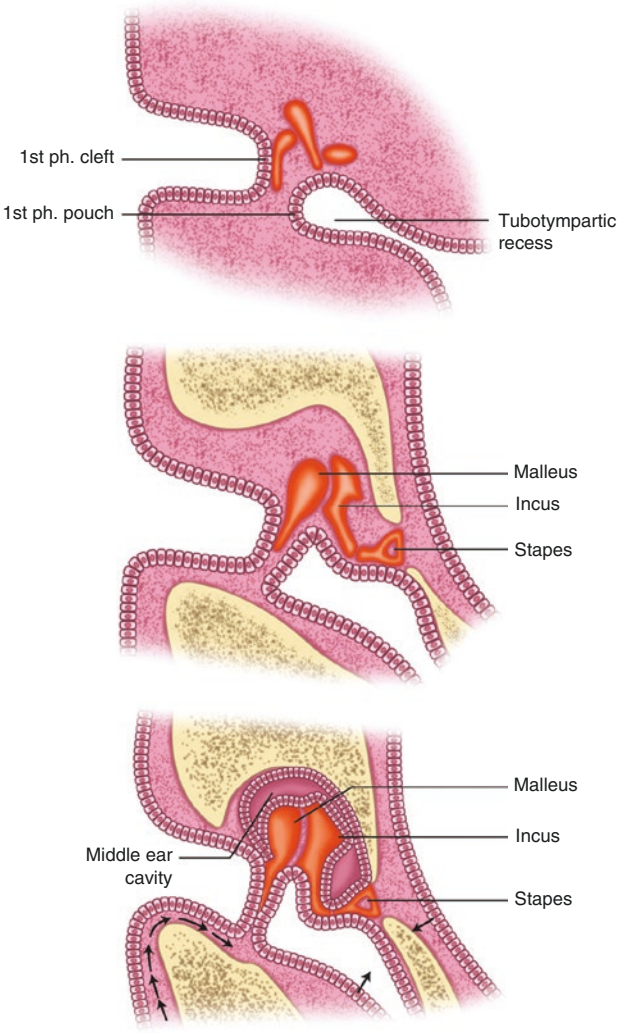

Fig. 36.6 Development of the middle ear

disease may drain to occipital and mastoid nodes. All of these groups tend to drain to both the internal jugular and spinal accessory groups.

Equalisation of pressures with the nasopharynx occurs via the pharyngotympanic (Eustachian) tube. This is derived from the narrow proximal part of the tubotympanic recess and connects the tympanic cavity with the nasopharynx. It is lined by mucous membrane that is continuous with that of the tympanic cavity. The function of the Eustachian tube is to equalise pressure changes between the middle ear and the atmospheric pressure, thereby allowing free movement of the tympanic membrane. The tube is opened by the action of the levator veli palatini, hence equalisation ("popping the ears") is commonly associated with yawning and swallowing (which is why some airlines give out sweets during take off and landing).

The auditory ossicles form a mobile chain of three small bones, which pass from the tympanic membrane to the oval window—a small opening in the bony inner ear (the cochlea). This opening leads to the vestibule of the labyrinth. The three bones are the malleus (hammer), the incus (anvil) and the stapes (stirrup). The stapes, the smallest of the ossicles, has a base (footplate) which snugly fits into the oval window. This opening is considerably smaller than the tympanic membrane. As a result, the vibratory forces that occur in response to sound are increased approximately 10 times. The vibration of the ossicles drives the stapes in and out of the oval window, pushing and pulling upon the liquid in the inner ear. The vibrating ossicles thus transfer the energy of sound from the air-filled outer ear to the fluid-filled inner ear. Two muscles dampen movements of the ossicles and the tympanic membrane—the tensor tympani and the stapedius. These have a protective action in that they dampen large vibrations resulting from loud noises. The nerve to the stapedius is a branch of the facial nerve—a useful fact to know when determine the location of a facial nerve palsy. Paralysis of the stapedius is associated with excessive acuteness of hearing (hyperacusis), as a result of uninhibited movements of the stapes.

36.1.5 Congenital Anomalies of Middle Ear

1. Congenital cholesteatoma is a benign lesion found in the middle ear cavity that results in conduction deafness. This occurs as a result of proliferation of endodermal cells lining the middle ear cavity.
2. Congenital fixation of stapes may occur due to failure of formation of the 'annular ligament'—a ring of fibrous tissue that connects the footplate of the stapes to the oval window.
3. In deaf mutism there is a defect of perception. This defect could be the part of the first arch syndrome in which anomalies of the malleus and the incus are present.
4. Michel Dysplasia is a very rare anomaly in which there is complete absence of the entire middle ear, including the cochlea, vestibule and all semicircular canals. The medial wall of the inner ear and the promontory are flat due to inner ear aplasia.

36.1.6 Inner Ear

This is found in the petrous part of the temporal bone and contains the vestibulocochlear organ. It is concerned with the senses of sound and balance. The structure can be considered as composed of (1) the bony labyrinth—a series of cavities (cochlea, vestibule, and semicircular canals) contained within the otic capsule of the petrous bone and (2) the 'membranous' labyrinth (filled by endolymph), which is suspended (not floating) in the bony labyrinth. This also consists of a series of communicating sacs and ducts. The thin cavity between these two labyrinths is filled with perilymph. Both endolymph and perilymph are important in stimulating the organs for balance and hearing. The cochlea is a shell-shaped part of the bony labyrinth that contains the structure concerned with hearing (organ of corti). The vestibule and semicircular canals are involved in balance. The semicircular canals (anterior, posterior, and lateral) communicate with the vestibule of the bony labyrinth. They are arranged at right angles to each other, thereby occupying the three planes in space.

Embryologically the inner ear is first of the three parts of the ear to develop. It develops from ectodermal thickenings on either side of the rhombencephalon (myelencephalon)—the otic placodes. On each placode, an otic pit appears, which invaginates into the underlying mesenchyme and then separates away from the surface ectoderm to become the otic vesicle. The otic vesicle subsequently divides into two components (1) ventral saccular portion and (2) dorsal utricular portion. The wall of this cyst-like structure gives rise to neuroepithelial cells and incorporates neural crest cells. These form the cells of the vestibulochoclear ganglia. Peripheral process pass to the saccule, utricle and semicircular canals via the vestibular nerve. Perilymph, which is similar in composition to cerebrospinal fluid, communicates with the subarachnoid space via the perilymphatic duct (Fig. 36.7).

At birth, the position of the tympanic membrane is horizontal, whilst the internal ear, tympanic cavity, the middle ear and the ossicles have reached their adult sizes. The inner ear does not receive pain fibres. Therefore, most of the disorders affecting this site do not result in pain. However, some diseases such as Meniere's can produce other sensations, such as pressure or fullness.

36.1.7 Congenital Anomalies of Inner Ear

Congenital deafness. The organ of Corti may be damaged by exposure to rubella virus, especially during weeks 7 and 8 of development. The membranous labyrinth may also fail to develop properly.

36.1.8 The Internal Acoustic Canal

This is a narrow canal that begins at the opening of the internal acoustic meatus (in the posterior cranial fossa), and passes laterally for about 1 cm within the petrous part of the temporal bone. It ends at the fundus, where a thin perforated plate of

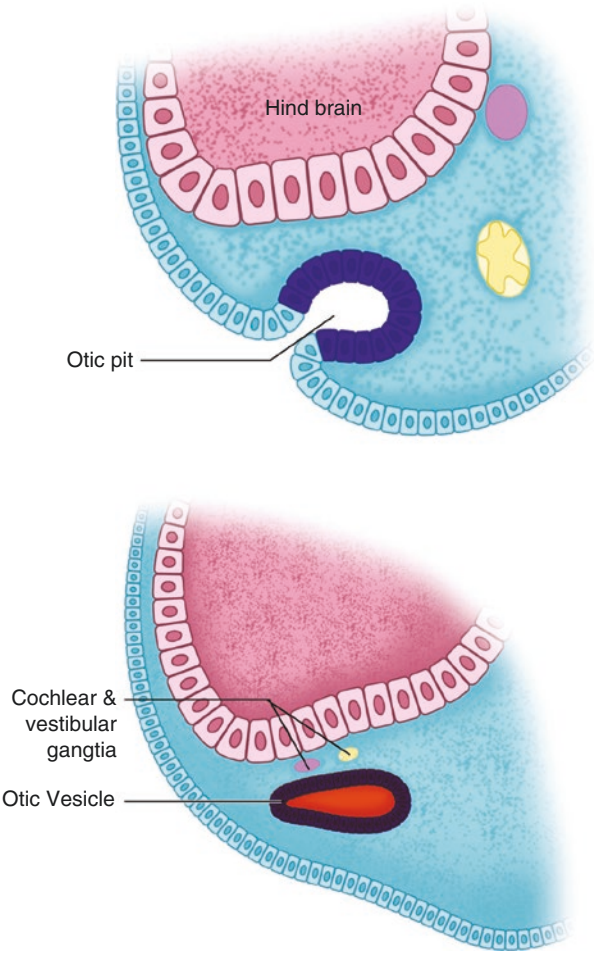

Fig. 36.7 Development of the inner ear

bone separates it from the vestibule. Through this plate pass the facial nerve (CN VII), the vestibulocochlear nerve (CN VIII) and its branches and the labyrinthine artery and veins.

36.1.9 Nerve Supply to the Ear

The ear has a complex and rich innervation including CNs V, VII, VIII, IX, and X and the posterior roots of C2 and C3. As such it is very sensitive to diseases an injuries which can result in severe pain. Otalgia ("earache") is thus a common symptom. It usually arises from local disease ("primary otalgia"). However pain may also be referred ('secondary' otalgia). Pathology occurring in various anatomical sites with the same nerve innervation (notably the TMJ, teeth,

oropharyngeal, laryngeal and hypopharyngeal regions) can present with otalgia. Of not oropharyngeal and nasopharyngeal tumours can initially present with 'ear ache'. If the ear looks normal, it is therefore essential to assess distant sites, especially the temporomandibular joint, oropharynx, nasopharynx, larynx, neck and teeth. With more local pathology facial palsy is a worrying feature and warrant careful consideration.

36.1.10 The Facial Nerve

This nerve is associated with the second pharyngeal arch and therefore innervates all the muscles derived from it. This includes the muscles of facial expression, the stapedius, stylohyoid, platysma and the posterior belly of digastric. The nervus intermedius (of Wrisberg), sometimes erroneously referred to as the "sensory root," is part of the facial nerve. The is so called because it is found between two large nerves (CNs VII and VIII). It carries secretomotor parasympathetic nerve fibres to the lacrimal, submandibular and sublingual glands. The motor component of the facial nerve and nervus intermedius have separate origins in the brainstem but emerge at the cerebellopontine angle. They accompany each another into the internal acoustic meatus of the petrous temporal bone where they join together at the geniculate ganglion. This is the only sensory ganglion of the facial nerve, where sensory neurones transmit taste sensation from the anterior two-thirds of the tongue and general sensation from the back of the ear, nasal cavity and soft palate.

After joining at the geniculate ganglion, the facial nerve passes along the facial canal (within the medial wall of the middle ear), where it gives rise to three branches (1) the greater petrosal nerve, (2) the nerve to the stapedius muscle and (3) the chorda tympani nerve. The fibres in the greater petrosal nerve subsequently enter the pterygopalatine fossa where they synapse in the pterygopalatine (sphenopalatine) ganglion. This then sends fibres to the lacrimal, nasal and palatine glands. The chorda tympani nerve passes across the tympanic membrane into infratemporal fossa where it joins the lingual nerve, a branch of the mandibular division of the trigeminal nerve. The chorda tympani carries preganglionic parasympathetic fibres to the submandibular ganglion.

As the facial nerve emerges from the stylomastoid foramen it gives off small motor branches to supply the occipital belly of occipitofrontalis, stylohyoid and posterior belly of digastric. It also gives off sensory branches to the skin of the external auditory meatus. The nerve then enters the parotid gland where it forms the "pes anserinus", from which five branches emerge from the anterior border of the gland (temporal, zygomatic, buccal, mandibular and cervical). These supply the muscles of facial expression (also erroneously named—their functions are primarily sphincteric to the mouth, nose and eyes). The facial nerve thus has two very important relationships—its intracranial course has a very close relationship to the middle ear. Its extracranial course has a very close relationship to the parotid gland.

36.1.11 The Vestibulocochlear Nerve

This is the sensory nerve for hearing (cochlear) and position sense and balance (vestibular). It arises in cerebellopontine angle and passes into internal acoustic meatus along with the facial nerve. The vestibulocochlear nerve consists of two distinct and separate nerves enclosed within one connective tissue sheath—the vestibular nerve and the cochlear nerve. Both nerves transmit information from specialised ciliated mechanoreceptors ("hair cells"). The cell bodies of the vestibular nerve send peripheral processes which terminate in receptors, in the ampullae of the semicircular ducts, utricle and saccule. Central processes enter the brainstem to synapse in the vestibular complex and the cerebellum. The cell bodies of the cochlear nerve are sited within the cochlear ganglion. Their peripheral processes terminate in the organ of Corti, within the cochlea. Central processes synapse in the brainstem.

36.2 Pathophysiological Mechanisms

36.2.1 Cerumen

Cerumen, or "earwax," is a waxlike substance that lubricates the outer skin of the ear canal and prevents it from drying. It is a composed of lipid-like secretions from the sebaceous and apocrine (ceruminous) glands, mixed with desquamated epithelial cells and debris. This forms an acidic coat that aids in the prevention of infection of the external auditory canal. The secretion also appears to have antibacterial properties and may also prevent the entry of insects. Cerumen is produced only in the outer third of the canal. It is naturally expelled by the ear.

36.2.2 Normal Hearing

Hearing involves the conversion of sound energy to electrical energy. More specifically it involves the conversion of a rapidly varying range of sound frequencies and intensities into 'acoustic signals', which are transmitted to the temporal lobes by neuro-electric impulses. To do this requires several complex mechanical and electrical processes which analyse sound waves, removes noise and then compares these to previous signals stored in the memory (i.e. recognisable sounds, voices etc.). The auditory system can normally perceive a reasonably wide range of sounds from 20 Hz to 20 kHz. Sound intensity is measured in decibels (dB). Tone is the number of vibrations, measured in cycles/sec, or Hz. Timbre allows the differentiation between two sounds with the same tone and intensity.

Sounds are normally transmitted by both Air conduction (AC), and by Bone conduction (BC). Conduction of sound starts at the pinna and passes through the external auditory canal to the tympanic membrane (eardrum). Here, the sound wave vibrations are amplified by the middle ear. Acoustic vibration is thus converted into mechanical vibration by the tympanic membrane and ossicles, which is then

transmitted to the organ of Corti. Within this organ, each frequency stimulates a particular group of receptor cells and their nerve fibres. The cochlear nerve innervates the organ of Corti and converts the received mechanical signals into electrical signals.

Air conduction requires normal function of the external and middle ear (pinna, eardrum and ossicles) to detect sound. Bone conduction can bypass this and allows sound to be transmitted directly to the inner ear via the bones of the skull. This includes the opposite ear. However the volume is reduced. Air conduction should therefore be greater than bone conduction and so normally an individual should be able to hear a tuning fork next to the pinna after they can no longer hear it when held against the mastoid. This forms the basis of the Rinne test.

The acoustic reflex (stapedius or auditory reflex) is an involuntary muscle contraction that occurs in the middle ear in response to high-intensity/low frequency sounds. It also occurs when a person starts to vocalise. The stapedius and tensor tympani muscles contract. The stapedius stiffens the ossicular chain by pulling the stapes away from the oval window. The tensor tympani muscle stiffens the ossicular chain by pulling on the malleus. The effect of both these actions is to dampen sound, thereby protecting the delicate inner ear structures. The acoustic reflex threshold (ART) is the sound pressure level (SPL) at which a sound of a given frequency will trigger the acoustic reflex. This is usually 10–20 dB below the level of discomfort. Individuals with normal hearing have an acoustic reflex threshold (ART) of around 70–100 dB SPL. People with conductive hearing loss have a higher acoustic reflex threshold. The acoustic reflex can be tested with tympanometry (extratympanic manometry—ETM). Since the stapedius muscle is innervated by the facial nerve, measurement of the reflex can be used to locate pathology in the nerve. If this is distal to the stapedius muscle, the reflex will remain normal.

The normal middle ear and mastoid are usually filled with air at atmospheric pressure. The tympanic membrane requires equal pressures on both sides to function. This is maintained by the Eustachian tube. Changes in air pressure during flying can occasionally cause pain in the middle ear. During ascent as the external air pressure drops, it is relatively easy for air to escape from the middle ear into the nasopharynx. However during descent reversing the flow can be more difficult as the increasing external pressure can impair opening of the Eustachian tube. Temporary vasodilation of the vessels in the malleus and tympanic membrane is a common consequence—barotitis. About 10% of adults and 20% of children are affected by this after flying. Symptoms usually resolve spontaneously. However, in some patients a ‘critical closing pressure’ may be reached, when the Eustachian tube cannot open and the pressure differential in the middle ear increases. This can result in fluid accumulating in the middle ear. The individual’s hearing becomes significantly reduced and this may persist for a month or more. In more severe cases tympanic membrane perforation may occur, also resulting in hearing loss. Both these problems can also result in balance problems, with protracted unsteadiness. Contrary to popular belief topical nasal decongestants are no more effective than placebo at preventing symptoms.

36.2.3 Hearing and Voice Production

The auditory system is essential to produce and regulate a normal voice. It provides two types of control over speech production—feedback control and feedforward control. Feedback monitors the voice during speech, which is adjusted as necessary. In the feedforward control, speech is produced from previously learned experiences. The auditory system plays three important roles—(1) providing information regarding the voice. This allows corrections in pitch, volume and intelligibility of speech, (2) providing information about environmental conditions, which is important in noisy situations and (3) and contributing to the generation of "internal models" in voice production, which allows rapid speech without constant auditory feedback. These roles are thus responsible for voice quality, pitch, loudness, resonance, articulation and rate.

36.2.4 Motion Sickness

The maculae are part of the membranous labyrinth which are primarily static organs. These contain small dense particles (called otoliths), which are embedded among the hair cells. The otoliths, under the influence of gravity, bend the hair cells, which in turn stimulate the vestibular nerve and provide awareness of the position of the head (even if the eyes are closed). The hairs are also stimulated by linear acceleration and deceleration and quick tilting movements of the head. Motion sickness results from a perceived difference between vestibular and visual stimuli.

36.2.5 Tympanic Membrane Perforation

Tympanic membrane perforation can present with sudden loss hearing loss and tinnitus. Pain may precede this if there has been a middle ear infection. There may also be a discharge. Diagnosis is confirmed by otoscopic examination and audiography. Several types of perforation are described

1. Central: the perforation does not involve the annulus, it is typically infectious in origin
2. Marginal: this involves the annulus. There is a higher association with cholesteatomas
3. Subtotal: large defect with an intact annulus

Causes include

1. Acute/chronic suppurative otitis media (most common cause)
2. Persistent perforation after extrusion of a grommet
3. Trauma (blow to the ear, barotrauma, diving, water skiing, explosion, forceful irrigation) (Fig. 36.8)

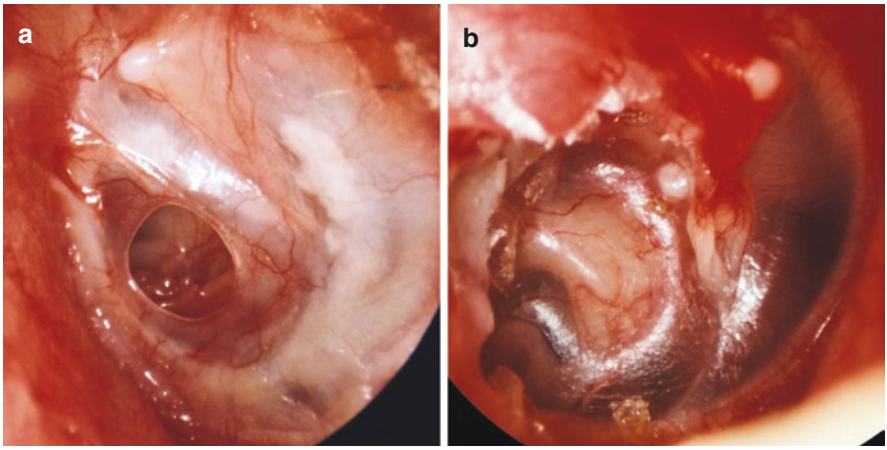

Fig. 36.8 (**a**) Penetrating injury of a left tympanic membrane caused by improper use of a cotton-tipped applicator. (**b**) Right ear. Severe damage to the tympanic membrane and ossicular chain (stapes) caused by an explosion (complete hearing loss)

4. Iatrogenic
5. Cholesteatoma (associated with marginal perforations)

Management involves keeping the ear dry and ear drops if it is infected. Tympanoplasty may be required for a persistent perforation.

36.2.6 Aural Fullness

This is the sensation of a "fullness", or "pressure" in the ear. This may be associated with tinnitus, hearing disturbance, autophony, nasal obstruction or a sore throat. It is often described in the early stages of Ménière's disease and idiopathic sensorineural hearing loss. Pathophysiological mechanisms may involve any part of the ear, from the external auditory canal to the inner ear but most commonly aural fullness results from dysfunction of the eustachian tube, an effusion in the middle ear or a chronic infection in the middle ear. However in around 10% of patients no cause can be found. Rarely it can be a symptom of nasopharyngeal carcinoma.

36.2.7 Ear Itching

Itching can be a symptom of ear infection, commonly otitis externa. Other possible causes include insect bites, contact allergies, and trauma. Chickenpox is accompanied by red, oozing blisters that cause intense itching. Eczema, contact dermatitis, scabies and ringworm are other sources of itching.

36.2.8 Perilymphatic Fistula

This is an abnormal defect between the air-filled middle ear and the fluid-filled inner ear, usually occurring in the round window or oval window membranes. This can be congenital. It can also arise as a complication of middle ear diseases (such as cholesteatoma), or secondary to trauma or rapid pressure changes (that can occur in SCUBA diving). The result of the defect is leakage of perilymph from the semi circular canals into the middle ear. Variations in middle ear pressure can therefore be transmitted directly into the inner ear. This forms the basis of the fistula test (described later). Symptoms of perilymphatic fistula include hearing loss, vertigo and tinnitus. Definitive diagnosis is made by direct visualisation of the fistula. Symptoms can resolve with bed rest, although surgery may be necessary in those with persistent symptoms.

36.2.9 Gout

Gouty tophi (uric acid deposits under the skin), can occur in the pinna, in association with chronic tophaceous gout. These have a white or yellowish appearance and are not usually tender or painful. Other sites of deposition include the olecranon bursa and PIP and DIP joints of the fingers. Rarely, is the appearance of tophi on the pinna the presenting symptom. Small deposits can resolve on lowering the patient's uric acid levels, larger deposits may require surgical removal.

36.3 Important Considerations When Taking a History

Patients may present acutely with a number of ear related symptoms, but by far the commonest are pain, sudden changes in hearing and aural discharge. Dizziness is also a common symptom, but has many varied causes and is therefore not always attributed to ear disorders. In all patients the following issues should be enquired into:

1. Local symptoms (deafness, tinnitus, discharge, pain and vertigo).
2. Previous head/ear injury
3. Previous ear surgery
4. Family history of deafness.
5. Systemic disease (e.g., stroke, multiple sclerosis, cardiovascular disease).
6. Ototoxic drugs (antibiotics (e.g., gentamicin), diuretics, cytotoxics).
7. Exposure to noise

The painful ear (otalgia) needs careful evaluation and its causes are discussed later. When the ear itself is the source of the pain (primary otalgia), examination of the ear is usually abnormal. When the ear is not the source of the pain (secondary otalgia), ear examination is often normal. Often the history will give a good

indication as to the underlying problem, for example a history of trauma, throbbing pain (infection) or deep seated pain (infiltrative pathology). Key points to establish are the patient's age, location of the pain and any radiation, aggravating factors (e.g. chewing) and associated symptoms (both otic and generalised). Discharge, tinnitus, hearing loss and vertigo suggest otic pathology. However the severity of the pain is not always an indication of the seriousness of the problem. Tumour related pain can be mild, whilst dental caries and otitis media can result in severe discomfort.

With primary otalgia the cause is usually apparent on examination (commonly otitis externa or media). However the cause of secondary otalgia can often be difficult to determine due to the ear's complex innervation and diverse sources of referred pain. In such cases questioning needs to be more generalised, noting particularly symptoms related to the throat, temporomandibular joint, teeth and cervical spine. Patients who smoke, drink alcohol, are older than 50 years, have diabetes and whose symptoms persist despite treatment, are at greater risk of having a serious underlying cause to their otalgia.

Abnormal 'ringing in the ear' (tinnitus) and loss of hearing are also common presenting symptoms. With the former it is important to establish the characteristics of the sound (unilateral or bilateral, high or low pitched, buzzing, pulsatile or clicking), its progression and frequency and its effects on sleep and daily life. A sudden onset of tinnitus is concerning, as this may indicate a vascular or traumatic aetiology. Questions regarding the pattern of tinnitus should attempt to differentiate pulsatile from continuous or episodic tinnitus. Pulsatile tinnitus is frequently due to a vascular source whereas Ménière's disease tends to be episodic. Specific associated symptoms to inquire about include hearing loss, vertigo, and aural fullness. The impact of patient positioning on the tinnitus should be asked; specifically, eustachian tube dysfunction is often alleviated by lying down. A past medical history of hyperlipidemia or diabetes may indicate carotid artery atherosclerosis, whereas a thyroid disorder or anaemia may suggest a high output cause. A number of medications are also known to cause tinnitus, although this is rarer than commonly believed. With loss of hearing enquire about its onset and duration (constant or intermittent, progressive), whether it is unilateral or bilateral, any obvious cause (injury or exposure to loud noises) and any family history. Associated symptoms of aural fullness, vertigo, tinnitus, otalgia, otorrhoea and other neurological complaints should be elicited. Autoimmune diseases, hypertension, diabetes, vascular disorders and neurological diseases can also affect the ability to hear.

When the patient presents with an aural discharge the nature of this should be determined. This can vary from clear fluid (CSF), through a more viscid yellow/green discharge (infection), blood-stained discharge (tympanic membrane perforation), to frank blood (trauma). Enquire about the presence of tinnitus, or change in hearing as well as pain. Dizziness has many causes both within and beyond the ear but some characteristics can help with diagnosis. Dizziness lasting only seconds to minutes is more commonly associated with benign paroxysmal positional vertigo (BPPV), or a cardiac arrhythmia. Episodes lasting hours occur in Ménière's and migraine, whilst symptoms lasting days suggest vestibular neuritis, labyrinthitis. Constant dizziness may arise form a central disorder.

36.4 Examining the Ear and Associated Structures

This starts with an inspection of the external ear. Gently remove any discharge, blood or wax. Look for obvious signs of abnormality. Make a note of

1. The size and shape of the pinna (comparison with the other side may help, although pinnae are not completely symmetric).
2. Pre-auricular sinuses or pits.
3. Signs of trauma.
4. Skin lesions of the pinna and external canal e.g. neoplasia.
5. Signs of infection/inflammation of the external ear canal
6. Any discharge.

Examination of the external auditory canal and tympanic membrane (TM) requires an otoscope. The external auditory meatus must be free of blood and wax. In the absence of suspected injury, a Q-tip will usually suffice to clean the canal, although in some patients, a small curette or gentle suction must be necessary. In adults the pinna should be grasped with the thumb and forefinger and pulled up and back. In infants, pull the pinna posteriorly. This straightens the S-shaped outer cartilaginous part of the canal. The speculum of the otoscope is then inserted into the canal slowly and gently advanced, observing as you go. Note the condition of the canal skin and presence of any wax, foreign tissue or discharge (Fig. 36.9). Look for telangiectasia, bulging, erythema, surface white plaques and perforations. The tympanic membrane should appear whitish and taut, like the skin of a drum. The upper region is opaque and referred to as the pars flaccida, whereas the larger lower pars tensa, is translucent. Fine vascular markings may be seen on the surface. At the junction of the two regions is an inferiorly directed linear structure. This represents the attachment of the malleus to the inner surface.

It is important to remember that nasopharyngeal pathology can present with unilateral ear symptoms. Always look at the throat and nasopharynx, especially in adults presenting with unilateral glue ear—this can be a presenting symptom of a nasopharyngeal tumour. A thorough head and neck exam should be performed on all patients presenting with tinnitus. A search for an identifiable source should be performed by auscultation of the auricular region, the mastoid, and the carotid arteries. Objective tinnitus secondary to a venous cause is identified by disappearance of the sound when the ipsilateral jugular vein is compressed. Careful otoscopy should be performed to evaluate for middle-ear infection, cerumen impaction, a dehiscent jugular bulb, or glomus tumor. The oral cavity should also be examined for contractions of the palatal muscles. The cranial nerves should be evaluated for evidence of hearing loss or brainstem dysfunction. Finally, a fundoscopic exam should be performed to look for papilloedema in suspected cases of pseudotumor cerebri.

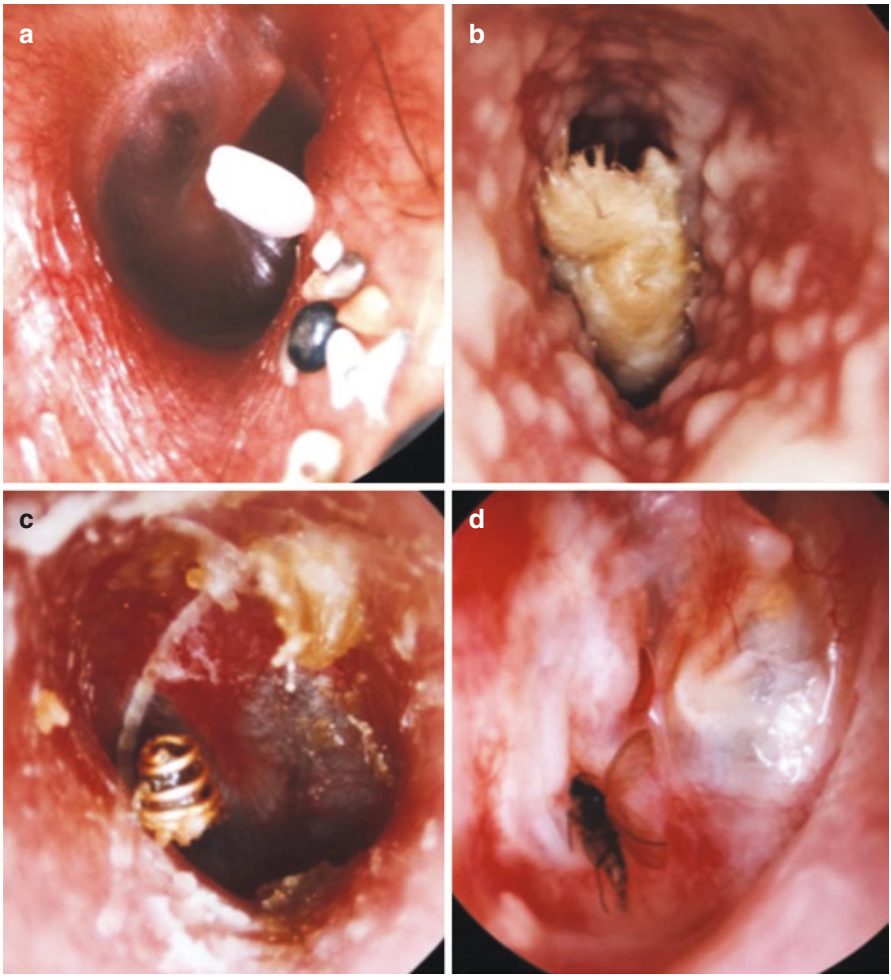

Fig. 36.9 Foreign bodies in external auditory canal. (**a**) Sand particles can be seen along the anterior wall of the external auditory canal. (**b**) A piece of paper has been 'forgotten' inside this external auditory canal; secondary infection (external otitis) of the skin. (**c**) A metallic hearing aid component, with secondary infection of the skin of the external auditory canal. (**d**) A deceased insect on the surface of this tympanic membrane

36.4.1 Battle's Sign (Mastoid Ecchymosis)

Bruising over the mastoid process, is suggestive of a fracture involving the middle or posterior cranial fossae of the skull. Like periorbital haematoma, this sign may take 1 or more days to appear following injury. It can be confused with a haematoma from a fracture of the mandibular condyle, but can be distinguished by palpating for mastoid tenderness, tragal tenderness, the condyle and assessing movements of the temporomandibular joint (TMJ).

36.4.2 Facial Nerve Function

To assess this, ask the patient to give a broad toothy grin. Look for fullness in the cheeks and symmetry. Then ask the patient to screw up his/her eyes. Gently try to prise them open. Finally ask the patient to raise his/her eyebrows. Look for asymmetry and force of movement. Paralysis of the facial nerve causes the face to droop. This is more marked with a lower motor neurone (LMN) lesion than an upper motor neurone (UMN) lesion. The best way to differentiate between the two is to test the muscles of the forehead. These muscles receive a bilateral innervation from the upper motor neurone level. Therefore with any UMN lesion (such as a stroke) they will continue to function. However, a LMN lesion such as Bell's palsy or involvement of the facial trunk by parotid tumour, skull base fracture, or penetrating injury, will involve the forehead.

36.4.3 Vestibulocochlear Nerve Function

Whispering or use of a high-frequency tuning fork can give a very crude assessment of hearing. However more precise assessment requires special tests and manoeuvres.

36.4.4 Pneumatic Otoscopy and the Fistula Test

Pneumatic otoscopy is a painless examination that determines the mobility of the tympanic membrane in response to pressure changes. Immobility can occur as a result of several causes (fluid in the middle ear, perforation, tympanosclerosis). Pneumatic otoscopy is therefore helpful in establishing the diagnosis of otitis media with effusion. This is a sensitive test and can detect the presence of an effusion even when the tympanic membrane appears normal otherwise. A pneumatic speculum is attached to the otoscope. By gently squeezing the bulb on the device, pressure is placed across the tympanic membrane and its mobility observed. A healthy membrane should be seen to move briskly. If diseased, thickening of the tympanic membrane results in loss of compliance and stiffening. If the membrane does not move at all a middle ear effusion should be suspected. In order for this examination to work an airtight seal in the canal is essential. Otherwise, false-positive results may occur. If the patient has a perforation and a perilymph fistula, they may feel unwell as nystagmus, dizziness, vertigo, imbalance, nausea, and vomiting may occur.

The fistula test is undertaken to detect a fistula in the wall of the bony labyrinth in the inner ear. Pressure changes in the external canal are applied intermittently by pressing on the tragus or using a Siegel's speculum. In a normal person, this test is negative because pressure changes cannot be transmitted into the labyrinth. However if a fistula is present the changes in the external auditory canal are transmitted to the labyrinth. This can produces nystagmus and vertigo. It is reported that the nature of the nystagmus can help localise the site of the fistula.

1. If the fistula is over the dome of the lateral semicircular canal this will result in horizontal deviation of the eyes towards the normal side. If pressure is maintained, nystagmus develops with the fast component towards the affected ear. As the pressure is released, the eyes return to normal.
2. If the fistula is in the lateral semicircular canal anterior to the ampulla, this will result in deviation of eyes, to the affected side
3. If the fistula is in the posterior semicircular canal this will result in vertical movement of eyes.
4. Vestibular involvement can result in rotatory horizontal nystagmus towards the diseased ear

36.4.5 Tuning Fork Tests: Weber's Test

The Weber test is a quick screening test for hearing loss. If there is significant loss of hearing in one ear, this should be performed. Hearing loss will 'lateralise', (move to one side), even with a relatively small amount of loss (5 dB). The Weber test can detect both unilateral conductive (external/middle ear) hearing loss and unilateral sensorineural hearing loss (inner ear hearing loss). Strike a 512 Hz tuning fork and place it on the centre of the forehead. Ask the patient in which ear it seems louder. The vibration is conducted through bone. This it will be quieter in an ear affected with sensorineural deafness, but louder with conductive deafness (the affected ear becomes more sensitive). Thus

1. A normal Weber test has sound heard equally in both sides.
2. Conductive deafness—sound heard more in deafer ear
3. Sensorineural deafness—sound heard more in better hearing ear

36.4.6 Tuning Fork Tests: Rinne Test

This test is primarily used for evaluating loss of hearing in one ear. It compares the perception of sound transmitted by air conduction (AC) to that transmitted by bone conduction (BC) through the mastoid. This can quickly screen for the presence of conductive hearing loss. It should always be accompanied by a Weber test to detect sensorineural hearing loss. Using a 512 Hz tuning fork, strike the tuning fork then place it against the mastoid bone. Instruct the patient to tell you when the sound is no longer heard. When this occurs, reposition the still vibrating tuning fork 1–2 cm from the auditory canal and ask again if they can hear it. With normal hearing, air conduction should be greater than bone conduction. Therefore the patient should be able to hear the tuning fork next to the pinna after they can no longer hear it whilst placed on the mastoid. If they are unable to hear the tuning fork next to the pinna, bone conduction is greater than air conduction. This indicates there is something preventing the passage of sound from the ear canal, through the middle ear apparatus to the cochlea. This is called conductive hearing loss. In sensorineural hearing

loss, the ability to sense the tuning fork by both bone and air conduction is equally diminished. Therefore the patient will hear air conduction after loss of bone conduction (similar to normal hearing). However with sensorineural hearing loss the sound will stop much earlier. This can be compared to the examiner's own hearing (if normal).

- AC > BC Rinne +ve (middle/external ear functioning normally)
- BC > AC Rinne –ve (defective middle/external ear)
- Weber and Rinne tests are screening tests and are not replacements for formal audiometry.

36.4.7 Dix-Hallpike Manoeuvre

This is a positional test for benign paroxysmal positional vertigo (BPPV). The patient sits upright with their legs extended. Their head is rotated by approximately 45°. The clinician then helps the patient to quickly lie down backwards with the neck extended approximately 20°. The patient's eyes are observed for nystagmus. If rotational nystagmus occurs then the test is considered positive for BPPV. A modified version exists for patients who are too anxious, or who do not have the mobility or flexibility necessary for the test. This modification involves the patient moving from a seated position to their side, lying without their head extended. In both tests, a positive result is indicated by the development of vertigo or nystagmus.

36.4.8 Head-Shake Nystagmus

This is a sensitive indicator of vestibular dysfunction. Move the patient's head in the horizontal plane for 20–30 s, then suddenly stop and look for nystagmus.

36.4.9 Caloric Reflex Test

The caloric reflex test ('vestibular caloric stimulation') assesses the vestibulo-ocular reflex. This involves irrigating the external auditory canal with ice cold or warm water. The change in temperature creates a convection current in the endolymph of the semicircular canal. This mimics a head turn to the ipsilateral side, with resulting nystagmus. Hot and cold water produce currents in opposite directions and therefore nystagmus occurs in opposite directions. A useful mnemonic to help remember the fast component's direction in nystagmus is "COWS" (Cold Opposite, Warm Same)—If cold water is used the fast phase of nystagmus is to the opposite side and vice versa. Absent eye movements suggest vestibular weakness in the horizontal semicircular canal. In comatose patients the fast phase of nystagmus will be absent. If both phases are absent, this carries a very poor prognosis.

36.5 Investigating Symptoms and Signs

36.5.1 Laboratory Tests

Full blood count may show a raised white cell count (neutrophils). Electrolyte disturbances may result in ototoxicity. Screening for diabetes may be important (this can be associated with hearing loss). Coagulopathies can also be associated with hearing disorders. Other useful tests include

1. ESR
2. Treponemal Studies: Lyme titres/TPHA/VDRL/FTA-ABS.
3. Lipid Profile: Atherosclerotic disease can be associated with sudden senorineural hearing loss
4. Fluid analysis (Beta 2 transferrin in suspected CSF leakage)
5. Microbiology for any discharge

Various metabolic abnormalities can cause sensorineural hearing loss. Thus, the evaluation of an unexplained sensorineural hearing loss should include the following—(1) complete blood count with differential, (2) blood sugar, (3) thyroid function tests and (4) serologic test for syphilis.

36.5.2 Plain Films

Mastoid X-rays are perhaps rarely requested today. These can show opacity of the air cells. These and other plain views of the temporal bone have now largely been replaced by CT and MRI.

36.5.3 CT/MRI of Temporal Bones

Patients with sensorineural and conductive hearing loss make up a significant number of temporal bone referrals. Other common indications for CT and MRI of the temporal bone (and posterior fossa) include facial nerve dysfunction, vertigo, tinnitus, otitis media, mass lesions in and about the temporal bone and unexplained or severe otalgia. Either CT or MRI can be an appropriate choice for these symptoms. CT is best when assessment requires boney detail. As an adjunct, MRI may help characterise the nature or extent of any mass. It also provides information about the membranous labyrinth. MRI is more useful in the assessment of the brain stem, suspected cerebellopontine angle tumours, acoustic neuromas, meningiomas, petrous apex lesions and the intracanalicular course of the facial nerve. Thus the most common use of MRI is in the assessment of sensorineural hearing loss and vestibular symptoms. Pulse synchronous tinnitus or possible vascular masses need careful evaluation to avoid biopsy of a highly vascular mass (such as a paraganglioma) or injury to large vessel such as the internal carotid. CT, with or without

contrast, CT angiography and CT venography may be required. MRI and MR angiography may also be required. CTA may also be used to identify carotid complications in temporal bone fractures.

Focused scans can usually identify congenital disorders and complications arising from suppurative ear disease. They can also define the extent of cholesteatoma, mastoiditis, temporal bone fracture and tumours (notably acoustic neuroma). CT is superior to MRI in revealing skull fractures and radiopaque foreign bodies. A rare complication of skull base trauma is a carotid-cavernous sinus fistula (see the chapter on the head). In this case, MRI and MR angiography are helpful in making the diagnosis. CT and MRI may also be required in patients with parapharyngeal space and nasopharyngeal masses potentially involving the temporal bone.

36.5.4 Nuclear Medicine

Radionuclide studies are used primarily and sparingly for diagnosis and following treatment for skull base osteomyelitis and necrotising otitis externa.

36.5.5 Audiometric Tests

Audiometry tests are carried out in soundproofed rooms using precision equipment. Several tests are available, depending on the individual's age, hearing level and suspected condition. These may be subjective (pure tone audiograms, speech audiometry), or objective (impedance audiometry, evoked response audiometry). Subjective hearing tests provide a rough estimate of the amount of hearing loss. These tests do not provide a quantitative result, but rather assess the cochlea in order to determine the location of the problem. Tuning forks and a speech perception test are commonly undertaken. Speech audiometry measures how well a person hears and understands speech signals. Objective testing is required to delineate auditory function and quantify AC/BC hearing in both ears. It can distinguish conductive hearing loss from sensorineural loss, cochlear versus neural dysfunction and malingering (pseudohypoacusis) Tests include

1. Pure Tone Audiometry: This is a measure of hearing ability using pure-tone signals at frequencies from 250 Hz up to 8000 Hz. The patient wears headphones. This is used to diagnose the degree and type of hearing loss by determining the faintest tones that can be heard at selected pitches.
2. Tymapnometry (Impedance Audiometry): This assesses middle ear pressure and impedance (an indirect measure of Eustachian tube function) and the status and mobility of the tympanic membrane. When sound waves reach the eardrum, some is absorbed and conducted to the inner ear, the rest is reflected back. If there is fluid behind the tympanic membrane, it will not vibrate as much. A probe containing a tiny loudspeaker is placed in the ear canal. This produces sound and a tiny microphone on the probe pick ups the reflected sound. Tympanometry can

be used to measure the acoustic reflex pathway (which involves CNs VII and VIII and the brain stem). It can also distinguish between sensorineural and conductive hearing loss. It is therefore helpful in the diagnosis of otitis media with middle-ear effusion. This test should not be performed in infants below 7 months of age—suppleness of the external canal's cartilage can produce misleading results.

3. Auditory Brainstem Response (ABR): This records the activity of the vestibulocochlear nerve and the central auditory pathway. It is elicited using short bursts of acoustic stimuli, which are detected by electrodes placed on the forehead and near the ears. This test is very useful to screen infants and children under five for hearing loss. It provides information on the cochlea and neuronal pathways.
4. Otoacoustic Emissions (OAE): Otoacoustic emissions (OAEs) are low-intensity sounds produced by the cochlea. They are generated from within the inner ear as a result of the amplification function of the cochlea. They can occur spontaneously or may be elicited by acoustic stimuli. However, if the outer hair cells are damaged or nonfunctional, OAEs cannot be stimulated. Therefore if an emission is present, it is likely that the patient's hearing is normal. Patients with a hearing loss greater than 25–30 dB usually do not produce these sounds. OEAs are measured with a small probe inserted into the ear canal. Because the patient is not required to respond, this is a useful test for neonates and infants and in patients who cannot be tested using simpler techniques. OAE is useful in testing for ototoxicity and damage to the cochlea outer hair cells. Since both ABR and OAE do not need patient co-operation they can be used to identify malingerers and for medico-legal purposes.
5. Videonystagmography. This technique is used to evaluate the function of the vestibular system. The instrument records nystagmus using infrared goggles after stimulating the vestibular system with warm and cool water.
6. Electrocochleography is an electrophysiologic measure of cochlear function. It is usually performed during intraoperative monitoring of cochlear and CNVIII status and in the diagnosis of Meniere's disease.
7. Other tests of the vestibular system include electronystagmography (ENG), rotational chair, and computerised dynamic posturography (CDP).