The Ear and Associated Structures: Part II

37

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37.1 Otalgia ("Earache")

This is a common symptom and usually occurs from local disease (when it is called "primary otalgia"). However pain may also be referred ('secondary' otalgia). If the ear looks normal, it is essential to assess distant sites, especially the temporomandibular joint, oropharynx, nasopharynx, larynx, neck and teeth. Since the ear receives sensory innervation from a wide range of nerves, the potential causes of referred pain are similarly diverse. Most importantly, oropharyngeal and nasopharyngeal tumours can present as 'ear ache'. As with any symptom, it is important to exclude sinister or serious pathology. This includes malignant otitis externa, cholesteatoma, myocardial infarction, temporal arteritis as well as malignancy. Often these diseases can be ruled out following a detailed history and examination rather than extensive testing. 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. Facial palsy, disproportionately severe pain and progressive deafness are worrying features and warrant careful consideration. Common/serious causes of otalgia include.

Local Causes (Primary Otalgia)

1. Chondrodermatitis Nodularis Helicis (affects the pinna only)
2. Perichondritis
3. Acute otitis externa
4. Furuncles (infected hair follicles)
5. Acute and "Malignant" otitis externa

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6. Barotrauma
7. Herpes Zoster
8. Tumours

Distant Causes (Referred Otalgia)

1. Tonsillitis and upper respiratory tract disease
2. Eustachian tube dysfunction
3. Dental pathology
4. Disorders of the TMJ
5. Parotid disease
6. Diseases of the oropharynx and nasopharynx
7. Diseases of the larynx
8. Cervical spondylosis
9. Malignancy of any of the above, but particularly the tongue base.

Important Features to Help Make the Diagnosis Include

1. One or both ears?
2. Is there a discharge?
3. Any associated hearing loss?
4. Systemic symptoms (upper respiratory, fever)
5. Previous surgery, or ear syringing
6. Use of cotton buds
7. Recent swimming, diving, or flying

When examining the ear look for (1) scars, (2) deformity of pinna, (3) cellulitis (4) abnormal appearance of external auditory canals and tympanic membranes. Feel also for lymphadenopathy and mastoid tenderness (mastoiditis).

Common and Serious Causes of Otalgia with an Abnormal Ear Examination

1. Acute otitis media is probably the most common cause of primary otalgia. The tympanic membrane is usually bulging and red, but not always.
2. Otitis externa (swimmer's ear) presents with swelling and redness of the auditory canal. There is often debris in the canal and covering the tympanic membrane. Tugging on the pinna is painful.
3. Foreign bodies in the ear canal are most common in children. Hearing-aid batteries should be removed urgently to prevent alkali burns.
4. Barotrauma can occur following scuba diving or flying. The tympanic membrane is hemorrhagic and there may be blood or serous fluid in the middle ear.
5. Malignant otitis externa is an invasive osteitis of the skull base, usually caused by Pseudomonas infection. There is severe, deep, unrelenting pain and granula-

tion tissue in the external auditory canal. Squamous cell carcinoma of the external auditory canal can present similarly.
6. Ramsay Hunt syndrome (herpes zoster of the geniculate ganglion) results in otalgia, facial paralysis and vesicles in the external auditory canal. There can also be hearing loss, tinnitus, vertigo, taste disturbance and reduced tearing.
7. Relapsing polychondritis is a systemic disease that involves cartilage. It can affect many organs, including the eyes, nose, heart, kidneys and nervous system. The most commonly affected site is the ear. The disorder results in an inflamed looking pinna. Sparing of the earlobe distinguishes this from cellulitis.
8. Cholesteatoma can erode the ossicular chain, inner ear and facial nerve canal, resulting in a sense of fullness rather than severe pain. Consider this in patients presenting with otorrhea and conductive hearing loss

Common and Serious Causes of Otalgia with a Normal Ear Examination

1. Pharyngitis and tonsillitis often cause referred pain to the ear. In many patients otalgia is the main symptom
2. TMJ dysfunction syndrome is described in more detail in the appropriate chapter on the lower jaw. This can often cause ear symptoms, especially on chewing.
3. Dental causes of otalgia generally involve the upper molar teeth. This includes caries, periodontal abscesses and impacted wisdom teeth.
4. Idiopathic otalgia is common. Some cases may be neuropathic in origin and may require a trial of gabapentin or amitriptyline.
5. Tumours in the upper aerodigestive tract, infratemporal fossa, neck or chest can all cause otalgia. The most common sites are the base of the tongue, tonsils and hypopharynx. A history of associated tobacco or alcohol use, dysphagia, weight loss, radiation exposure, hoarseness are highly suggestive.
6. Glossopharyngeal neuralgia can causes pain in the tonsillar area, pharynx and middle ear, often bought on by swallowing on palpation of the tonsillar region. Sphenopalatine neuralgia can results in pain around the eye, nose, ear and mastoid.
7. Bell's palsy (facial paralysis) can be accompanied by postauricular pain, hyperacusis, taste disturbances and decreased tearing.
8. Temporal arteritis often causes pain and tenderness that can include the ear. However less than half patients have tenderness in the temporal arteries. Other symptoms include malaise, weight loss, fever, anorexia and blindness.

37.1.1 Chondrodermatitis Nodularis Helicis

This presents as an exquisitely tender nodule localised on the pinna, which is occasionally crusted. The upper part of the helix is the most frequent site affected, but lesions can arise anywhere around the rim, usually over the convexities in the cartilage, where the skin is thin and at risk of pressure (Fig. 37.1). Most lesion are only

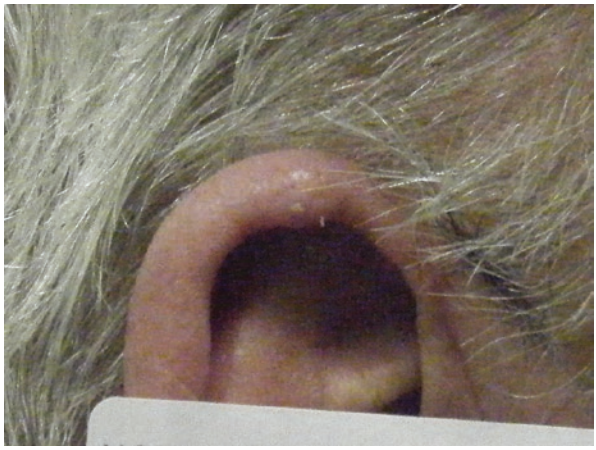

Fig. 37.1
Chondrodermatitis nodularis helicis

a few millimetres in size. Nodules attain their maximum size of 4–6 mm in a few months and remain unchanged if left untreated. Over a period of a few weeks, the nodule develops a central crater, which contains crust-like material. The condition most commonly affects fair-skinned men from the age of 40 onwards and is more common on the right. Bilateral lesions have been reported with in almost 10% of cases. The aetiology of CNCH is unclear. However, it is believed to be related to microtrauma, prolonged excessive pressure, or ischaemia to the dermis. Chronic trauma and pressure (from headphones, pillows etc.) and cold exposure are thought to be precipitating factors. With increasing age there occurs thinning of skin and cartilage, the loss of elastic tissue, and the degeneration of vascular and connective tissue. Pain and tenderness are the key to diagnosis. If the lesion is not painful, consider the possibility of a BCC, or SCC. Treatment includes intralesional steroids, cryotherapy, Nitroglycerin gel, Photodynamic therapy or surgical removal of the inflamed cartilage by excision or curettage. Advise the patient to avoid pressure to the affected area and use pressure relieving padding.

37.2 The Discharging Ear (Otorrhoea)

Discharge from the ear may arise from various causes, including infection, trauma, inflammation, tumours and others. Types of discharge include wax, pus, mucus, blood, cerebrospinal fluid (CSF) and occasionally saliva. However the commonest cause is middle ear infections. The history and nature of the discharge usually establish the diagnosis, however it is important to ask about otalgia, hearing loss, vertigo, or trauma, check the patients temperature and examine the cranial nerves—these may reveal ‘red-flag’ features requiring urgent investigation.

The colour of the fluid often suggests the cause of otorrhoea. A purulent discharge indicates the presence of infection, while a bloody discharge may occur following trauma or from granulation tissue associated with chronic infection. The

presence of a mucoid discharge suggests a perforation of the tympanic membrane (there are no mucous glands in the external ear canal, therefore the fluid must be coming from the middle ear). Clear, watery fluid is highly suggestive of CSF.

Recurrent episodes of purulent otorrhoea suggest CSOM, while acute onset of purulent otorrhoea suggests acute otitis media with perforation of the tympanic membrane, or acute otitis externa. In acute otitis media, the pain characteristically improves when the tympanic membrane ruptures and otorrhoea develops. In otitis externa the pain is persistent. A foul-smelling discharge is usually associated with cholesteatoma or a tumour. Some skin diseases (psoriasis and eczema can also result in discharge).

1. Watery—CSF or eczema
2. Mucoid—Chronic suppurative otitis media
3. Purulent—See below
4. Bloody/Sero-hemorrhagic—Trauma/acute otitis media/malignancy
5. Foul smelling—Cholesteatoma

Common pathological causes of otorrhoea are

1. Chronic suppurative otitis media (CSOM) with/without cholesteatoma
2. Otitis externa
3. Acute otitis media with perforation of the tympanic membrane
4. CSF otorrhoea
5. External or middle ear neoplasms
6. Granulomatous diseases—tuberculosis, atypical mycobacteria, Wegener's granulomatosis.

37.2.1 Purulent Otorrhoea

Diseases Involving the External Auditory Canal (EAC)

1. Diffuse otitis externa
2. Furuncle of the ear canal
3. Malignant (necrotising) otitis externa
4. Trauma to the EAC
5. Tumours of the EAC

Diseases Involving the Middle Ear

1. Acute suppurative otitis media (ASOM)
2. Chronic otitis media (COM)
3. Specific otitis media: e.g. TB, syphilis
4. Tumours

Vertigo and facial palsy associated with otorrhoea are indications for urgent investigation. These may suggest the presence of

1. Labyrinthine fistula or acute labyrinthitis
2. Inner ear erosion by tumour
3. Complication of acute otitis media
4. Malignant otitis externa
5. Cholesteatoma
6. Malignant neoplasm
7. Ramsay Hunt syndrome

If the external canal is filled with discharge, a pus swab should be taken and the ear canal cleaned. Ear toilet is important in the diagnosis and treatment of otorrhoea. This should always be done under direct vision. Syringing of the ear is contraindicated as the introduction of water under pressure may exacerbate an acute infection or water may enter the middle ear through a TM perforation.

37.3 Infections and Inflammation: Otitis

This is a general term referring to inflammation or infection of the ear. It is subdivided into (1) otitis externa, involving the pinna and external auditory meatus, (2) otitis media involving the middle ear (often with fluid in the middle-ear space). This a very common childhood infection, and (3) otitis interna (or labyrinthitis), involving the inner ear. Because this includes the sensory organs for balance, vertigo is a common symptom.

37.3.1 Infections of the EAM and Pinna

37.3.1.1 Acute Localised Otitis Externa (Furuncle)

Acute localised otitis externa is an infection of a hair follicle. This usually begins as a folliculitis but can progress to form a small abscess, a furuncle. The infecting microorganism is usually *Staphylococcus aureus*. Resolution may occur with topical or systemic antibiotics. Any localised abscess should be drained.

37.3.1.2 Acute Diffuse Otitis Externa ("Swimmer's Ear").

Acute otitis externa (AOE) is thought to affect 10% of people at some stage, and can present in acute, chronic, or necrotising forms. While milder forms of acute otitis externa are often short-lived and isolated episodes, a significant number of cases can persist for weeks or even months, despite intensive treatment. Fungal overgrowth can occur, especially after prolonged antibiotic use. Once resolved, there is a significant risk of recurrence. Acute otitis externa (AOE) is a superficial bacterial or fungal (or both) infection with diffuse inflammation involving the skin of the external auditory canal (Figs. 37.2 and 37.3). It is more common in humid countries, hence its

other name "Singapore Ear". Predisposing factors include ear cleaning, lack of protective cerumen, certain dermatological conditions, chronic use of hearing aids or ear plugs, localised trauma (cotton buds, fingernails) and repetitive retention of water (swimming). Local defence mechanisms can become impaired by prolonged ear canal wetness. Skin desquamation then leads to microscopic fissures that provide a portal of entry for infecting organisms. Whilst AOE is primarily a local disease, more serious and invasive disease can occur in certain situations. Diabetics are at risk of a severe form of disease known as malignant otitis externa. *Pseudomonas aeruginosa*, Staphylococcus and Proteus are the most common pathogens, but others include gram-negative bacilli, Staphylococci and fungi. Rare fungal infections have been described with Aspergillus species and Candida species as well as other fungi. These can occur secondary to repeated courses of oral antibiotics and can vary from minimal to extremely severe. Fungi are often found as a harmless commensal, however they can become more active, filling the canal with thick fungal debris. This results in increasing pressure and severe unrelenting pain. Swabs from the external canal should be interpreted with caution because they may grow normal colonising organisms.

Infections may be either acute (less than 6 weeks) or chronic (more than 3 months). Patients present with rapid onset (generally within 48 h) of severe ear pain, aural fullness, tenderness and pruritus. These are classically described as out of proportion to the degree of inflammation seen. Touching or moving the outer ear significantly increases the pain—a useful diagnostic clue in distinguishing AOE from acute otitis media with otorrhea. The most common finding on examination is the presence of greyish white thick debris with its characteristic "wet blotting paper"

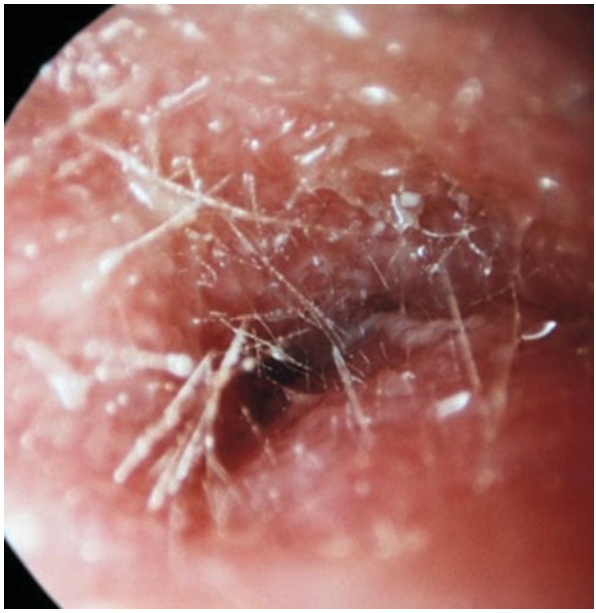

Fig. 37.2 Acute external otitis. Note the cartilaginous external auditory canal is oedematous and the size of its lumen is reduced

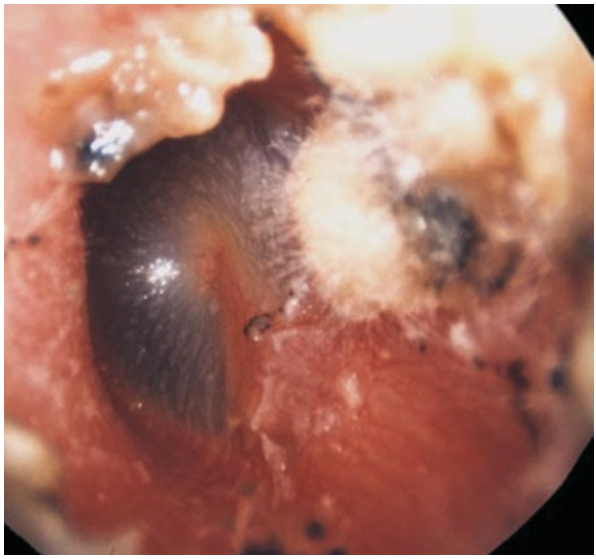

Fig. 37.3 Otomycosis: a creamy whitish exudate covers the skin of the external auditory canal. Fungal hyphae and conidiophores are visible. In this case the tympanic membrane is not affected

appearance. The EAM is also swollen and oedematous with a purulent foul smelling discharge. Patients may also have conductive hearing loss. On direct otoscopy, the canal is oedematous and erythematous and may be associated with surrounding cellulitis. A high fever is not usually present except in severe cases. In all cases of infection it is important to rule out malignant external otitis (look for granulation tissue in the canal, cranial nerve involvement and consider this in diabetic patients).

Management of otitis externa requires thorough and regular cleaning of the canal with fine suction. Topical ear drops (acetic acid, antibiotic, or antibiotic/corticosteroid combination) are effective in mild-to- moderate AOE and may be prescribed. For mild-to-moderate acute otitis externa, the following steps may be taken:

1. First line therapy should be a topical antibiotic with or without topical steroids for 7–10 days. More severe cases should be managed with systemic antibiotics that cover *S. aureus* and *P. aeruginosa*.
2. Adequate pain control can be achieved with systemic acetaminophen, nonsteroidal anti-inflammatory medications or oral opioid preparations. Topical steroid preparations have had mixed effects on hastening pain relief in clinical trials and cannot be recommended as monotherapy.
3. If the canal is very oedematous an absorbent wick may be placed. This helps to remove some of the excess moisture and open the canal. Although aural toileting and wick therapy are common and logical practices, these are not always necessary.

A good response should be seen within 48–72 h, but a full response can take up to 6 days in patients treated with antibiotic and steroid drops. Patients that do not improved should be assessed for obstruction, the presence of a foreign body, or an alternative diagnosis (e.g., dermatitis from contact with nickel, a viral or fungal infection or antimicrobial resistance).

Oral antibiotics are not usually required unless there are signs of cellulitis within or around the canal, or systemic upset. It is important not to prescribe antibiotic and steroid drops if a fungal infection is possible. Steroid drops can make a fungal infection worse and prolonged use promotes growth of fungus. Therefore antibacterial ear drops with steroids should only be used for a maximum of 1 week. Advise the patient also to keep the ear dry and avoid the temptation of self cleaning and probing.

37.3.2 Malignant Otitis Externa (MOE)

This is an uncommon but aggressive form of otitis externa, that occurs mainly in the elderly, diabetics and the immunosuppressed. Pseudomonas is the most common pathogen but aspergillus or mucor can also cause infection. Malignant otitis externa follows a much more chronic but indolent course. Infection can extend into the adjacent bony part of the ear canal and the surrounding soft tissues. Unrecognised and untreated, it can eventually result in osteomyelitis of the skull base. This may then progress to the onset of multiple cranial nerve palsies, including the facial nerve and the vestibulocochlear nerve. Malignant otitis externa is thus potentially life-threatening. The hallmark of infection is its unrelenting severe pain that interferes with sleep and persists even after swelling of the canal has resolved. However MOE does not usually result in a fever or raised white cell count. Management involves oral or intravenous antibiotics and sometimes surgical debridement. Diabetes and other causes of immunosuppression should be screened for and managed accordingly.

37.3.3 Chronic and Eczematous Otitis Externa

Chronic inflammation of the skin of the external ear canal can be caused by a number of problems including (1) bacterial infection, (2) chronic skin disorders (such as eczema), (3) fungal infection (Aspergillosis), (4) chronic irritation (hearing aids, Q-tips), (5) allergy, (6) chronic drainage from middle ear disease, and (7) rarely a tumour. Eczematous otitis externa is a dermatological condition that affect the EAM. This may take the form of atopic dermatitis, contact dermatitis or psoriasis. Often, more than one factor is involved. For example, a patient with eczema may subsequently develop a fungal infection. The ear feels itchy, so the patient then scratches the canal, prolonging irritation and symptoms. In diabetics or immunosuppressed individuals, chronic external otitis can become serious and progress to malignant external otitis.

Management may be difficult and may require trials of more than one strategy. These include

1. Advise the patient to avoid any potential causes of otitis externa.
2. If fungal infection is suspected prescribe a topical antifungal preparation. For uncomplicated infections, consider Clotrimazole 1% solution, Acetic acid 2% spray or Clioquinol. Use steroids with caution.

3. If the cause is an irritant or allergic dermatitis, advise the patient to avoid contact with the irritant or allergen, and prescribe a topical corticosteroid. Seborrhoeic dermatitis can be treat topically with an antifungal/corticosteroid combination.
4. If no cause is apparent prescribe a 7-day course of a topical preparation containing only a corticosteroid without antibiotic. Consider co-prescribing an acetic acid spray. If the response is inadequate, consider adding a topical antifungal preparation.
5. If treatment needs to be continued beyond 2 or 3 months, seek specialist advice.

37.3.4 Perichondritis

This is described in more detail later in the section on injuries to the pinna. It is an infection involving the perichondrium of the ear cartilage, most commonly the auricle (pinna). Untreated, this can become a serious infection, or it can become chronically inflamed, resulting in severe deformity of the cartilage. Infection can arise following trauma, but also following worsening of an otitis externa overlying cellulitis. Clinically the pinna is tender, red, swollen and warm. *Pseudomonas aeruginosa*, *Staph. aureus*, and Streptococcus are the common organisms involved. Treatment requires admission and high dose systemic antibiotics (Fig. 37.4).

37.3.5 Bullous Myringitis

Bullous myringitis is an infection of the tympanic membrane in which small, fluid-filled blisters develop. However, this does not result in an effusion. It is exquisitely painful (Fig. 37.5). The infection is caused by the same viruses or bacteria that cause other ear infections and infections elsewhere, such as the flu, common cold and *Streptococcus pneumoniae*. It is seen more commonly in people who already have an upper respiratory tract infection. This can irritate the eustachian tubes and stop them from draining. Infected fluid from the respiratory tract can then ascend into the ear and cause infection of the membrane. Bullous myringitis is thus also more likely to occur in people with a middle ear infection.

Symptoms are similar to other types of ear infections and may be associated with symptoms of an upper respiratory tract infection. Patients complain of sudden onset of severe pain. This lasts 24–48 h. There may be hearing loss in the affected ear. This will usually resolve once the infection clears. The patient may also develop a fever and discharge from the ear. This will only happen if one of the blisters breaks. If a young child has bullous myringitis, they might be irritable and tug at their ear in an attempt to relieve pain. Treatment usually includes antibiotics and simple analgesia, either orally or topically. Although viruses can cause bullous myringitis, antibiotics are usually prescribed. Symptoms usually improve within 2 days. Rarely hearing loss, meningitis, or sepsis can occur.

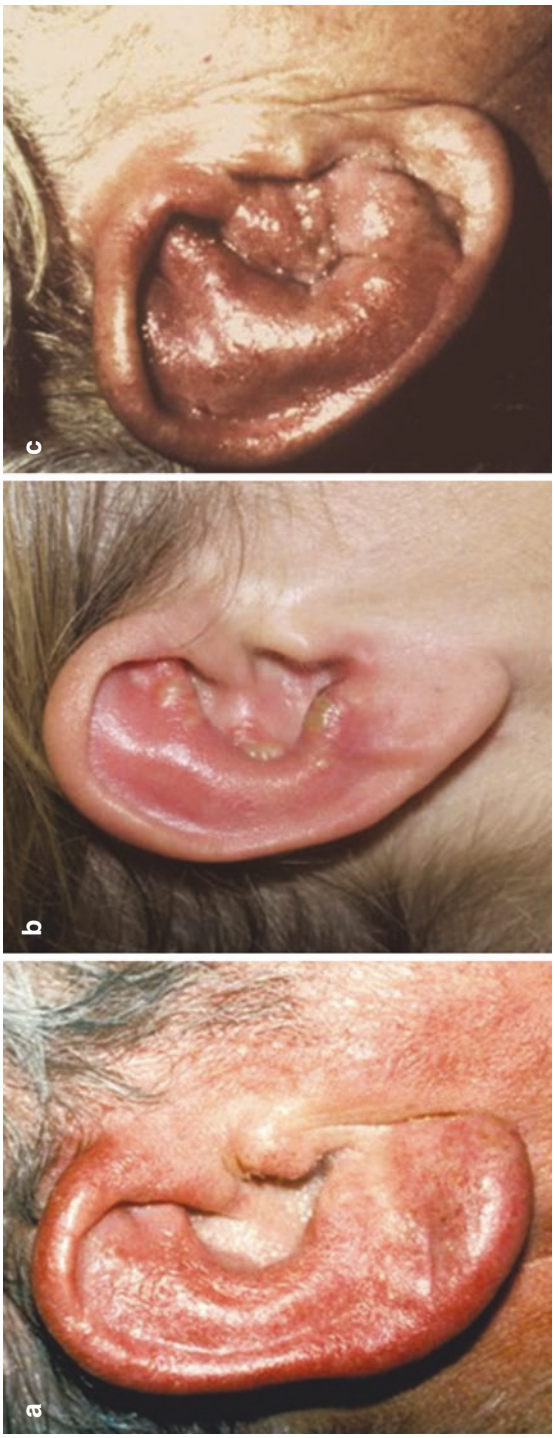

Fig. 37.4 (**a**) Erysipelas; (**b**) perichondritis; (**c**) relapsing polychondritis. Note that in erysipelas (**a**) the whole auricle, ear lobe included, is involved, while in perichondritis (**b**) the ear lobe is normal

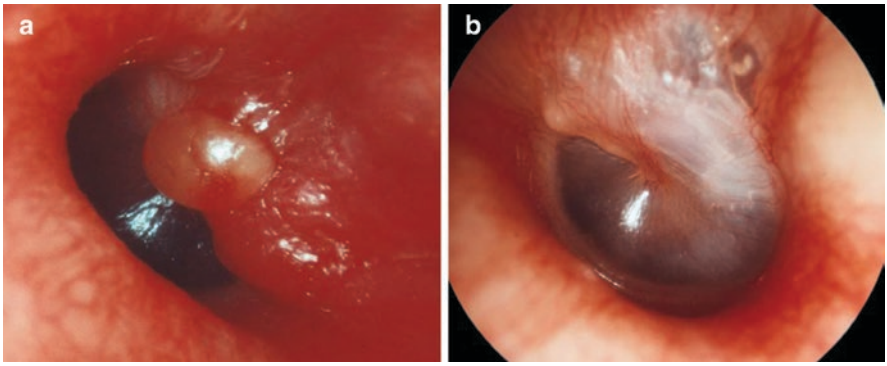

Fig. 37.5 (**a**) Acute bullous myringitis (left ear). Notice multiple bullae filled with serohaemorrhagic exudate. (**b**) Bullous myringitis (same patient) 30 days later recovered without any hearing problem

37.3.6 Granular Myringitis

Granular myringitis is inflammation of the tympanic membrane characterised by persistent granulation tissue on the outer surface of an intact membrane (Fig. 37.6). The cause is not known, but is believed to arise following local trauma or infection which results in loss of the epithelial layer of the tympanic membrane. It can also develop in patients with neglected or inadequately treated acute external otitis. The onset of the disease is insidious and presents with a persistent foul-smelling purulent discharge, raw-looking canal skin and granulation tissue, with an intact tympanic membrane. There may be mild discomfort. These features may also suggest malignancy and malignant otitis externa and therefore biopsy may be indicated. High-resolution computed tomography (CT) may also be performed to exclude cartilaginous and bony involvement (seen in MOE). Cultures usually yield Pseudomonas or Proteus spp. Treatment includes debridement, cauterisation with silver nitrate sticks, topical antibiotic and corticosteroid drops. Treatment with dilute vinegar solution has also been reported as a harmless alternative to conventional antibiotic drops. If successful the raw surfaces re-epithelialise spontaneously.

37.4 Infections of the Middle Ear

37.4.1 Otitis Media

Otitis media refers to a group of inflammatory mucosal diseases of the middle ear with particular involvement of the tympanic cavity. These are common and usually occur bilaterally. They can be acute or chronic. The two commonest types are (1) acute otitis media (AOM) and (2) otitis media with effusion (OME).

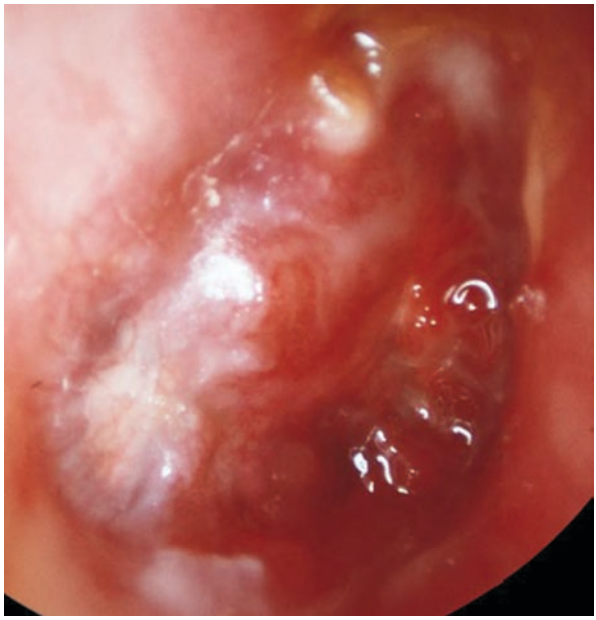

Fig. 37.6 Granular myringitis: a portion of the tympanic membrane (right ear) and of the external canal is covered with granulation tissue

37.4.2 Acute Otitis Media (AOM)

Acute otitis media is a common problem in children. Around two-thirds of all children have an episode of infection before their third birthday. In almost all cases, AOM is preceded by a viral infection of the upper respiratory tract. The most frequent pathogen is respiratory syncytial virus. Other commonly occurring viruses are influenza and parainfluenza viruses, rhinoviruses, adenoviruses, and enteroviruses.The most frequently occurring bacterial pathogens are *Streptococcus pneumoniae* and *Haemophilus influenzae*, followed by *Moraxella catarrhalis*. *Streptococcus pyogenes* and *Staphylococcus aureus* are found in smaller numbers of cases. Prior to introduction of the pneumococcal vaccine the majority of cases of bacterial AOM were caused by pneumococci. Some infections develop tympanic effusion, which may result in middle ear hearing impairment persisting into later childhood.

AOM may occur in isolation, or it can be associated with bullous myringitis. Infection begins with mucosal inflammation and oedema, which results in exudates within the middle ear. Oedema of the mucosa prevents effective drainage and equalisation of pressures through the Eustachian tube, such that, as pus accumulates, the pressure builds up. This results in increasing discomfort and the tympanic membrane to bulge—an important diagnostic sign. Untreated, this continues until eventually the tympanic membrane perforates, with sudden relief of pain and purulent otorrhoea. As the middle ear can now drain the infection will slowly resolve. Patients usually present complaining of an abrupt onset of throbbing ear ache (otalgia). This progresses in intensity until perforation of the tympanic membrane relieves some of

the pressure. Examination of the tympanic membrane is the cornerstone to correct diagnosis. Bulging and reddening, with a scaly, yellowish bulging of the tympanic membrane is commonly seen. In some cases pulsation of the membrane and flattening of the appearance of the manubrium mallei may be visible. Assessment of the tympanic membrane may be difficult in young irritable children, making the diagnosis uncertain. In such cases pneumatic otoscopy may help confirm the restricted mobility of the tympanic membrane. Other useful diagnostic tests are tympanometry and acoustic reflectometry. Palpation of the mastoid and assessment of facial nerve function are important. On tuning fork testing a conductive deafness is usually present, which is often associated with tinnitus. Patients are usually systemically unwell.

Deafness in children is a treatable cause of developmental delay—therefore early detection and treatment is important. Management involves antibiotics (Amoxicillin, or amoxicillin + clavulanic acid), usually orally, but occasionally intravenously. Penicillins are often the first line of treatment, but culture and sensitivities should be taken if possible. Cefuroxime, Erythromycin and Clarithromycin are alternative if allergic to penicillin. Analgesics and antipyretics (Acetaminophen (paracetamol) and ibuprofen) are also required as this is typically a painful condition. Administration of topical local anaesthetics is not recommended. If a bulging tympanic membrane persists, despite adequate antibiotic therapy, myringotomy under general anaesthesia may facilitate the escape of pus and early resolution of symptoms. Patients who present late, with a discharging ear following perforation should be started on broad-spectrum antibiotics after microbiology specimens have been taken. If signs and symptoms do not resolve, it is important to consider the possibility of an underlying infection elsewhere (notably in the mastoid, nasopharynx, sinuses). Complications include

1. Acute mastoiditis—Patients develop unusual prominence of ear and a boggy red swelling over the mastoid. The posterosuperior wall of external auditory meatus may be distorted. There is an elevated CRP and shadowing of the mastoid on plain films. Bezold mastoiditis is swelling of the sternomastoid muscle with torticollis
2. Labyrinthitis—Patients develop vertigo with nystagmus beating initially towards the affected ear, later towards the other side. Progressive inner ear deafness develops.
3. Facial palsy—this may be partial or complete
4. Sinus vein thrombosis—Patients become very unwell, with a general deterioration in their condition. Griesinger sign is retroauricular swelling of the skin over the emissary vein of the mastoid
5. Epidural abscess, subdural abscess, meningitis—Patients develop general deterioration, headache, fever, stiff neck. Seizures may occur
6. Cerebral abscess—Patients develop severe headache, stiff neck, drowsiness and seizures. Raised intracranial pressure may occur.

7. Gradenigo syndrome. This is very rare. Infection spreads to the apex of the petrous temporal bone. Patients develop irritation of the trigeminal nerve with severe pain behind the eye and paralysis of the oculomotor nerve and abducent nerve

37.4.3 Otitis Media with Effusion (OME): "Glue Ear"

This is also known as "serous" or "secretory" otitis media. It is said to affect around one-third of children at some stage during childhood, presenting with loss of hearing and mild otalgia, sometimes associated with tinnitus. OME occurs as a result of accumulation of non-purulent fluid in the middle ear, behind the intact eardrum, often secondary to dysfunction of the Eustachian tube. The eardrum itself usually shows no signs of inflammation. Dysfunction of the eustachian tube plays a key role in the pathogenesis of OME. This may become inflamed or blocked by the adenoids, preventing full aeration of the middle ear cavity and equalisation of pressures. Inflammation and hypertrophy of the middle ear mucosa, combined with hyperplasia of secretory cells can then occur. Allergies, primary ciliary dyskinesia, chronic rhinosinusitis, cleft palate and an immature immune system have also been reported to contribute to the development of this condition. Over several weeks or months, the middle-ear cavity slowly becomes filled with a sorts fluid. This is initially relatively thin but soon becomes very thick and glue-like, hence the name. A retracted but intact eardrum with a shortened malleus handle can be seen. Sometimes an air-fluid level or air bubble may be seen. The effusion can appear clear or yellowish to bluish, depending on its viscosity. The majority of children show a hearing loss about 20–30 dB (Fig. 37.7).

Although many cases will resolve spontaneously, OME can persist or become recurrent. In some cases a short course of antibiotics may prove helpful if there is suspicion of an associated infection. Management may be non-surgical or surgical interventions. Non-surgical management consists of active observation, medical therapy, autoinflation and hearing aids. Short-term (less than 6 weeks) intranasal steroid may be used for OME associated with concurrent allergic rhinitis and adenoid hypertrophy. Oral steroids and prolonged intranasal steroids are not generally recommended. Hearing aids may be considered in persistent bilateral OME and hearing loss where surgery is contraindicated. Surgical intervention may be considered after 3 months of persistent OME in children with hearing loss >25 dB (at three frequency average), continued discomfort and/or structural changes to the tympanic membrane or middle ear. Myringotomy with ventilation tube (grommet) insertion is the procedure of choice—a small incision is made in the tympanic membrane and a plastic grommet inserted. These often self-extrude after around 6 months and therefore repeated insertions may be necessary if symptoms persist. At the same time adenoidectomy should be considered in children with persistent OME and hypertrophied adenoids.

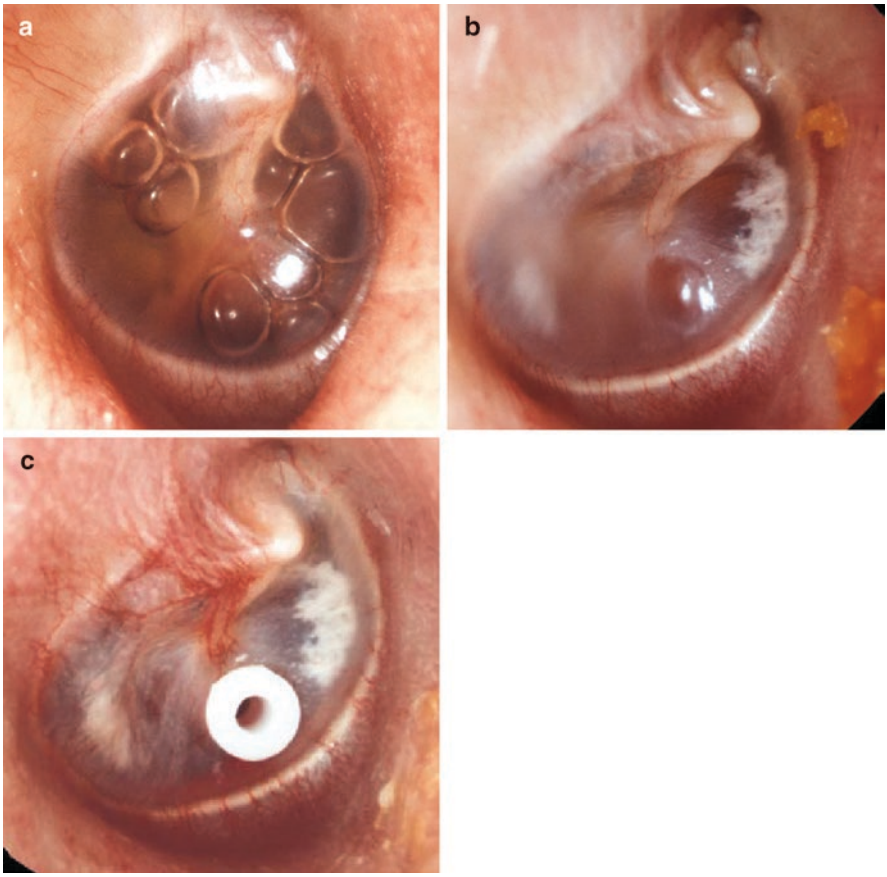

Fig. 37.7 (**a**) Serous otitis media with bubbles following air inflation, right ear. (**b**) Mucous otitis media, (**c**) Same ear after insertion of a Teflon grommet

37.4.4 Chronic Suppurative Otitis Media (CSOM)

Chronic suppurative otitis media (CSOM) is defined as a chronic inflammation of the middle ear mucosa and submucosa. This affects both the middle ear and mastoid cavity. It is characterised by recurrent or persistent ear discharge (otorrhoea) over 2–6 weeks through a perforation of the tympanic membrane. Today the disease is considered to be a multifactorial disease resulting from a complex series of interactions between environmental, bacterial, host and genetic risk factors. It can be a complication of acute otitis media, developing a few weeks after the initial onset of symptoms and resulting in recurrent episodes of aural discharge. Recently, bacterial biofilms have gained attention in the pathogenesis of CSOM. These attach firmly to damaged tissue, such as exposed inflamed bone and ulcerated mucosa, or to

otological implants such as tympanostomy tubes and are resistant to antibiotics and other antimicrobial agents. They therefore difficult to eradicate and may contribute to recurrent infections. Cytokines have also been implicated in the pathogenesis of OM. Typical findings may also include thickened granular middle-ear mucosa and mucosal polyps. If an effusion develops, this prevents the middle ear ossicles from properly relaying sound vibrations from the ear drum to the oval window of the inner ear, causing conductive hearing loss. In addition, inflammatory mediators generated during CSOM can penetrate into the inner ear through the round window. This can cause the loss of hair cells in the cochlea, leading to sensorineural hearing loss. *Pseudomonas aeruginosa* and *Staphylococcus aureus* are the most predominant pathogens that cause CSOM, followed by Proteus vulgaris and *Klebsiella pneumoniae*. CSOM can also arise from co-infections with more than one type of bacterial and viral pathogen. Fungi have also been identified in cultures, possibly as a result of repeated use of antibiotic drops, but also from climatic conditions—a moist and humid environment favours the prevalence of fungal infections of the ear.

Patients can present with painless, persistent or recurrent otorrhoea, associated with perforation of the tympanic membrane. Otorrhea and hearing loss are the most important clinical symptoms of CSOM. Otorrhea occurs intermittently and presents as an odourless mucous or serous discharge. Otalgia is not a usual symptom. Therefore if earache develops, it is important to consider coexisting otitis externa. Typical auroscopic appearances of CSOM show a central eardrum perforation, with an intact annulus fibrosus. Mucopurulent exudate, and inflammation of the middle ear mucosa may be seen. The malleus handle can be displaced. CSOM must be distinguished from cholesteatoma in childhood. It is therefore important to note the site of the tympanic membrane perforation. "Atticoantral" perforations (marginal perforations involving the posterosuperior part of the pars tensa or pars flaccida) are more likely to be associated with cholesteatoma. This is sometimes referred to as 'unsafe' CSOM. 'Safe' CSOM is seen in those cases without evidence or risk of cholesteatoma. Tone audiometry usually confirms a conductive hearing loss under 20 dB. Occasionally CSOM leads to sensorineural hearing loss, presumably as a result of inflammatory mediators from the middle ear damaging the hair cells of the inner ear. If the patient develops otalgia, vertigo, systemic upset or signs of postauricular inflammation, they should be investigated urgently to exclude mastoid or intracranial involvement (Figs. 37.8, 37.9, and 37.10).

Management is initially non surgical. Conservative treatment of CSOM is primarily intended to optimise the conditions for a tympanoplasty. This includes the elimination of any pathogens and drying of the ear. Local treatment with antiseptic or antibiotic (ciprofloxacin) eardrops can reduce infections and chronic inflammation. Routine oral antibiotic treatment is not recommended. Surgical management includes tympanoplasty, sometimes combined with ossiculoplasty. General consensus is that the ideal time for a tympanoplasty is at an age over 6 years. In the long term tympanosclerosis can develop. This is where calcified deposits build up in the tympanic membrane and the ossicular chain, resulting in reduced mobility.

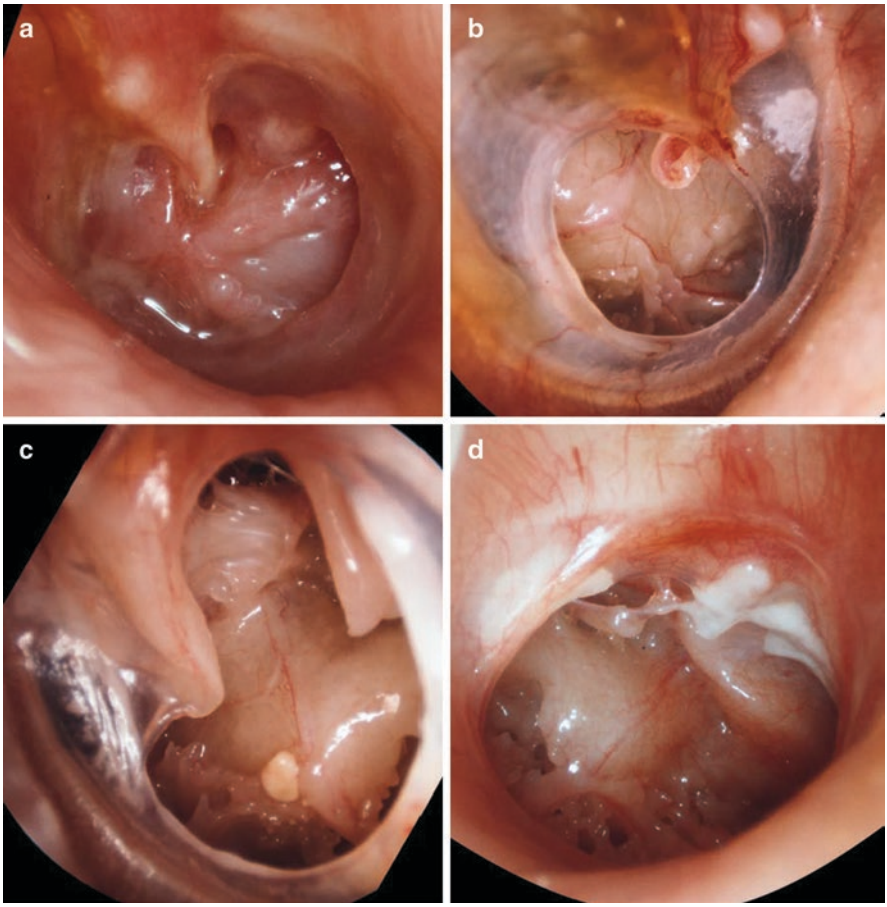

Fig. 37.8 Chronic otitis media (**a**) showing active mucosal disease with discharge through perforation (**b**) shows dry perforation with hypertrophic middle ear mucosa through the perforation (**c**) dry posterior perforation with tympanosclertic posterior margins (**d**) shows stapes head through a subtotal perforation

37.4.5 Herpes Zoster Oticus (Ramsay Hunt Syndrome)

Herpes zoster oticus (HZO), also known as Ramsay Hunt syndrome is a rare complication of herpes zoster infection. Reactivation of latent varicella zoster virus particles (VZV—a member of the herpes zoster viral group) in the sensory geniculate ganglion results in vesicular eruptions in the external meatus, otalgia and facial paralysis. When reactivation occurs neural inflammation, pressure and injury to the confined facial nerve in the temporal bone causes facial palsy. At the same time the virus migrates from the geniculate ganglia into the skin around the ear or into the oropharynx via sensory fibres, where it replicates and produces more virus particles. When quiescent, the virus has the potential to remain latent for years following chickenpox.

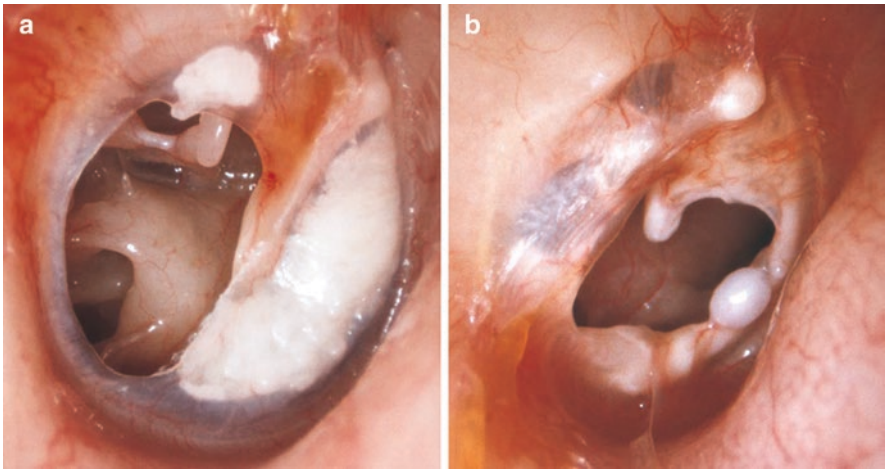

Fig. 37.9 (**a**) Chronic otitis media, right ear, dry perforation (inactive stage). Notice a tympanosclerotic plaque involving the anterior half of the tympanic membrane. A smaller plaque is located at the level of the long process of the incus. Note the in-cudostapedial articulation, the stapedial tendon emerging from the tip of the pyramidal eminence and the round window riche. (**b**) Chronic otitis media, right ear, inactive stage; tympanosdcrotic tissue along the margin of the perforation

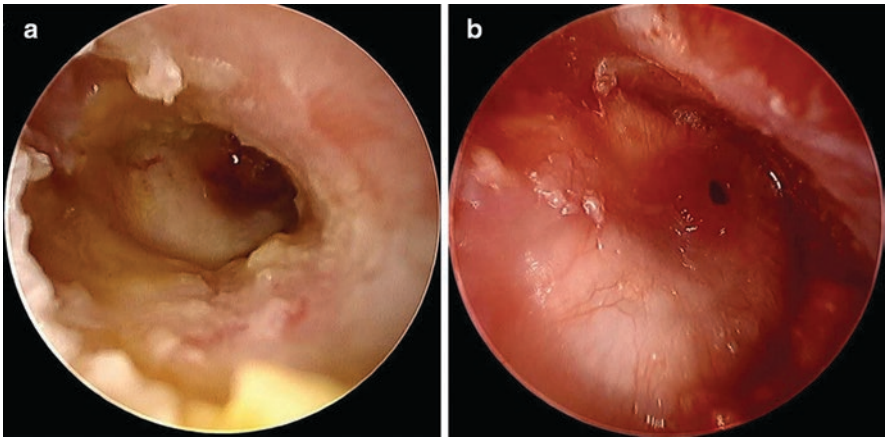

Fig. 37.10 Chronic suppurative otitis media with tympanic membrane perforation, (**a**) Tympanic membrane and external car canal coated in white/yellow purulence and debris. (**b**) Upon debridement of the canal and tympanic membrane, an anterior perforation is revealed with a rim of granulation tissue visible within the perforation

HZO affects both sexes equally. Most patients are aged over 60, although it can affect all ages. It is rare in children. Incidence and clinical severity increases when host immunity is compromised, usually from a decline in cellular rather than humoral immunity. Because not all symptoms present at the onset, this syndrome

can be misdiagnosed, commonly as Bell's palsy. HZO accounts for about 10% of cases of facial palsy, which is usually unilateral. Similar cases have also been reported associated with herpes simplex type 2 virus. Some specialists believe that this condition represents a polycranial neuronitis.

The classic presentation involves a triad of (1) lower motor neurone facial paralysis (2) otalgia and (3) vesicles in the auditory canal and auricle. Facial palsy can occur several days before the onset of eruptions. VZV can also cause facial palsy in the absence of skin lesions—this is referred to as "zoster sine herpete" and is usually diagnosed using serological assays or polymerase chain reaction (PCR). Infection can also spread to involve the vestibulocochlear nerve, resulting in tinnitus, hyperacusis, sensorineural hearing loss and vertigo to varying degress. Involvement of cranial nerves V, IX, X, XI, and XII has also been reported. Diagnosis is usually based on the history and clinical findings. Oral lesions are also present in most cases. Laboratory confirmation is based on increasing antibody titres. PCR can detect VZV in saliva, tears, middle ear fluid and blood (Figs. 37.11, 37.12, and 37.13).

Treatment is controversial in some patients but current recommendations suggest a combination of acyclovir and prednisone. Newer drugs, include valacyclovir, famciclovir, penciclovir and brivudine. However concern center partly on the use of steroids in the presence of a viral infection. It is important to be careful prescribing steroids in VZV (and other viral) infections. If there is viraemia the patient is at risk of developing viral encephalitis. It is therefore important to be up to date with the latest evidence base guidelines and discuss the need for steroids with ENT (or

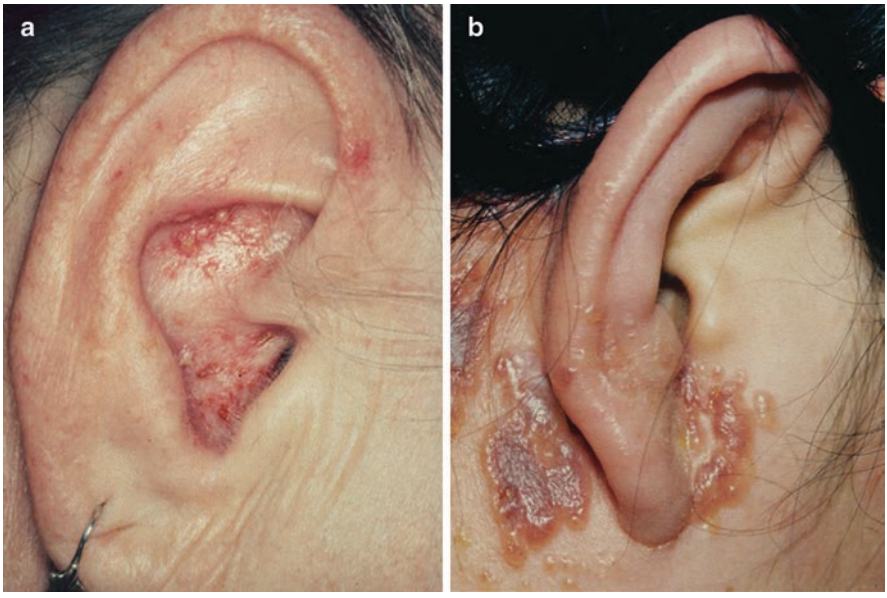

Fig. 37.11 (**a**, **b**) Herpes zoster oticus. (**a**) Numerous vesicles (some of them crusted) are seen scattered throughout the concha. (**b**) Numerous vesicles in the concha. In front and behind the auricle

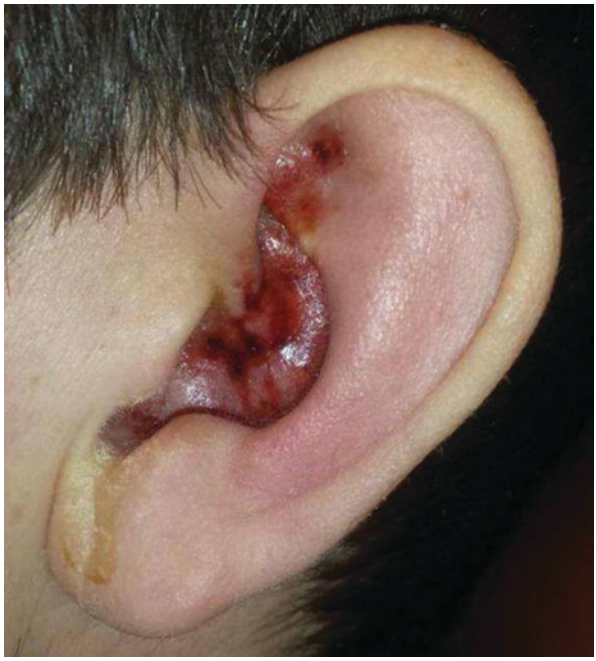

Fig. 37.12 Ramscy-Hunt Syndrome (Herpes Zoster Oticus). Vesicular lesion in the conchal bowl of the pinna

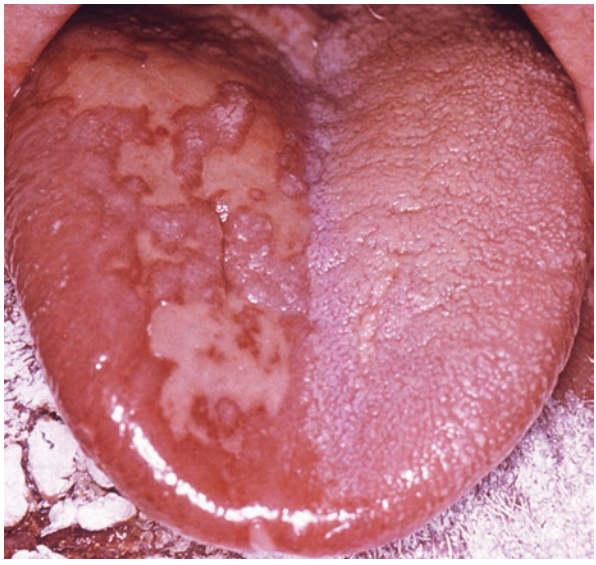

Fig. 37.13 Herpes zoster affecting the right half of patient's tongue and chin. Herpetic lesions involving the throat usually produce acute rather than chronic throat pain. Courtesy of Centers for Disease Control and Prevention (CDC). Public Health Image Library

follow local protocols). Complete recovery of facial paralysis is less likely compared to Bells palsy. Full recovery occurs in only about 20% of untreated patients and is related partly to the severity of palsy at the onset of symptoms. Rehabilitation for facial palsy includes electrical stimulation, infrared radiation and facial

neuromuscular exercises. Immunization of older persons may boost cell-mediated immunity to VZV and provide protection against herpes zoster and postherpetic neuralgia.

37.4.6 Tuberculous Otitis Media

Tuberculous otitis media (TOM) is an important cause of suppuration in many countries. It is an uncommon, insidious and frequently misdiagnosed form of TB. This should always be considered in any ear that fails to respond to standard therapy. TOM usually occurs as a result of spread from adjacent organs (lungs, larynx, pharynx and nose) via aspiration of mucus through the auditory tube, or by haematogenous spread. Primary TOM, in which there is direct implantation through a tympanic membrane perforation, is rare. Patients present with multiple perforations of the tympanic membrane, pale granulations, painless otorrhea, mixed hearing loss and facial palsy. A swab for appropriate culture studies, coupled with chest radiography, will usually confirm the diagnosis. Treatment involves an antitubercular regime and early surgical intervention to decompress the facial nerve if involved. Other complications include labyrinthitis, meningitis, and subperiosteal abscesses.

37.5 Extension of Infection into the Surrounding Bones

37.5.1 Mastoiditis

Mastoiditis is a potentially serious and one of the commonest complications of acute otitis media. Before antibiotics became available it was fatal in around 75% of cases. Most infections are caused by pneumococcus bacteria. Other organisms include staphylococci, resistant enterococci or fungi. Fungal mastoiditis caused by *Aspergillus fumigatus* predominantly occurs in immunocompromised patients. Invasive temporal bone mycoses are rare. Today, pneumococcal conjugate vaccines have significantly decreased infection with pneumococcus bacteria, and mastoiditis is less common. Nevertheless, inadequately treated mastoiditis can result in deafness, sepsis, meningitis, brain abscess and is still occasionally fatal.

Infection from the middle ear passes into the mastoid antrum, infecting the mucosal lining of the mastoid air cells. This can result in rapidly spreading suppuration and bone necrosis. If this is untreated, infection can spread further beyond the confines of the mastoid, both intracranially (meningitis, brain abscess, extra and subdural abscess, sigmoid and lateral sinus thrombosis, dural venous thrombophlebitis) and extracranially (labyrinthitis, osteomyelitis and even subacute bacterial endocarditis) (Fig. 37.14). Two forms of the disease have been described—non-coalescent and coalescent form. Inflammatory thickening of mucoperiosteal layer in the mastoid antrum and cell complexes, characterises non-coalescent mastoiditis, while the coalescent form progresses to destruction of bony walls of the mastoid process. Prolonged infection results in compression of the bone by the swollen

mucosa and retained secretions. This creates a hyperaemic and acidotic environment, which leads to osteoclastic activity and bone resorption within the mastoid (hence the name coalescent mastoiditis). As the inflammation progresses, bone resorption proceeds in all directions. Thus coalescent mastoiditis is the potentially severe form of infection, which can quickly develop into life threatening intracranial complications. This may be suggested by the presence of tympanic membrane perforation, with granulation tissue growth in the middle ear cavity. Other infections and disease processes, such as cholesteatoma and Langerhans cell histiocytosis, can sometimes mimic this process.

Hearing loss is a common complication of extracranial spread and extension of inflammation into the labyrinth can produce vertigo. The infection may also spread to the facial canal and its nerve, resulting in weakness or complete paralysis of the

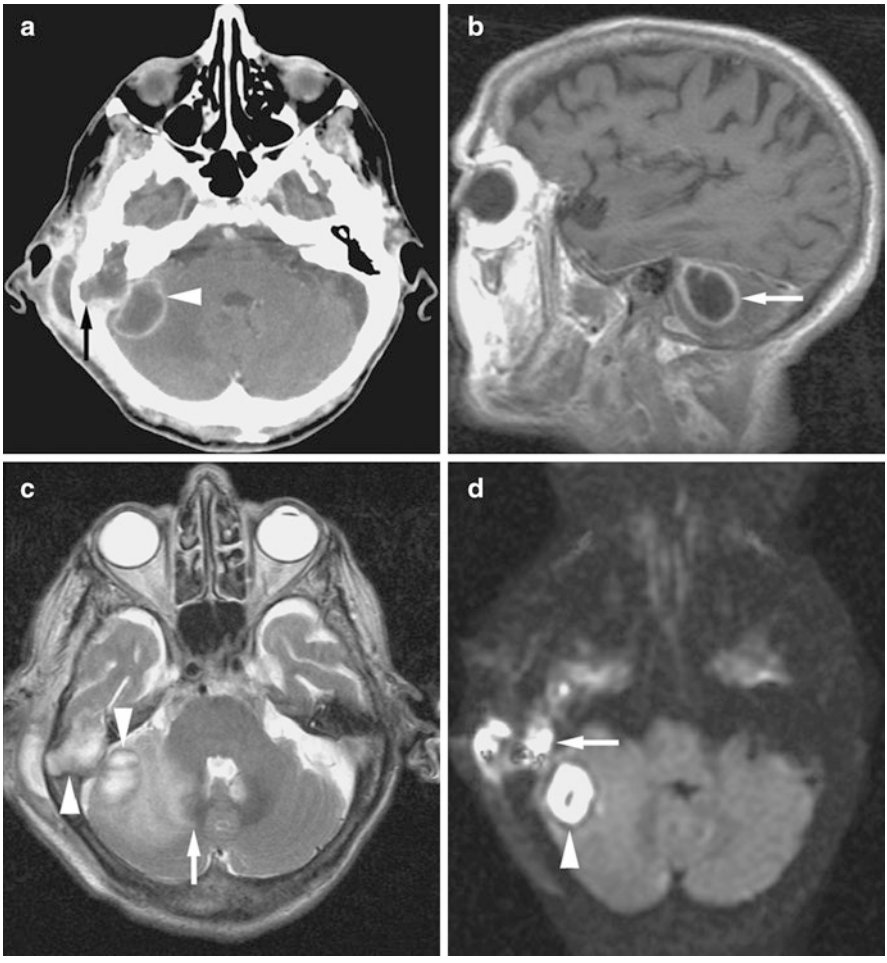

Fig. 37.14 CT and MRI showing temporal lobe abscess from complicated Mastoiditis. Note the reactive brain edema surounding the abscess

face. Other complications include Bezold's abscess (an abscess behind the sternocleidomastoid muscle), Citelli abscess (pus from the mastoid passes along the posterior belly of the digastric muscle to the occipital and cervical region), Luc's abscess (subperiosteal collection beneath the temporal muscle), or a subperiosteal abscess. Mastoiditis following irradiation has also been reported to occur in up to 50% of cases within 12 months of treatment. Life-threatening complications can result if the infection spreads to the dura, dural sinuses or brain. In recent years, IgG4-related disease (IgG4-RD) has become a recognised entity that causes progressive fibrosis and formation of mass lesions. This is diagnosed by raised serum IgG4 and the presence of storiform fibrosis, prominent lymphoplasmacytic infiltrate, obliterative phlebitis and infiltration of excessive numbers of IgG4-positive plasma cells following biopsy. Recent publications have reported cases of IgG4-RD in the mastoid sinus—a relatively new anatomic location for this disease. Presenting symptoms were varied and included tinnitus, hearing loss and cranial nerve palsies. This may be confused with mastoiditis. However patients require immunosuppressive therapy with steroids and Rituximab.

Patients complain of a persistent and deep-seated throbbing pain or pressure in the ear, with increasing deafness, aural fullness and tinnitus. A creamy discharge is usually present which can be profuse. They may also notice a brown discharge on the pillowcase upon waking. Patients are usually systematically unwell, with pyrexia and tachycardia. Post auricular swelling pushes the pinna forwards and is accompanied by tenderness over the mastoid. Otoscopy may show bulging of the roof or posterior wall of the external auditory canal. The tympanic membrane is usually red, perforated and discharging. Mastoid X-Rays may show opacity of the air cells. However CT Scans will give far more information especially if the patient is drowsy, irritable, photophobic, has a persistent high fever despite treatment or is complaining of severe headache. In such cases consider intracranial complications. Any middle ear drainage should be sent for culture and sensitivity.

Tympanocentesis for culture purposes can also be done if no spontaneous drainage occurs. FBC and ESR may be abnormal but are neither sensitive nor specific and add little to the diagnosis. Patients usually require admission for intravenous antibiotics and pain relief. If the causative organism is unknown, broad-spectrum antibiotics are given. Initial therapy with intravenous vancomycin and ceftriaxone may be given until results of the culture and sensitivity studies are available. However the choice of antibiotic may vary. If there is no immediate or complete response, or if a subperiosteal abscess is suspected, surgery may be indicated. A cortical mastoidectomy is performed, in which the abscess is drained, the infected mastoid cells are removed and drainage is established from the antrum of the mastoid to the middle ear cavity. Chronic mastoiditis is treated with oral antibiotics, eardrops, and regular ear debridement.

37.5.2 Petrous Apex Mucosal Disease (Petrous Apicitis)

This is essentially the same disease that occurs in the middle ear and mastoid, which involves the air cells within the petrous apex. As such it has many of the same causes and complications as does mastoiditis and middle ear infections. Infections

and inflammatory diseases can involve the petrous apex as part of more generalised skull base disorder, including necrotising otitis externa (see below), skull base osteomyelitis and rare conditions such as Wegener granulomatosis and Langerhans histiocytosis. In petrous apex mucosal disease, obstruction of the air cells results in resorption of air and chronic inflammation. The mucosa becomes friable and may bleed. Along with inflammation, cholesterol and other blood products accumulate, resulting in cholesterolosis or mucocele formation at the petrous apex. The consequences of these changes vary widely and are similar to those that occur in chronic mastoiditis. The disease can be limited to simple mucosal thickening with or without active inflammation, or localised bone erosion (similar to coalescent mastoiditis) can occur - this is referred to as acute or subacute petrous apicitis. Untreated, osteomyelitis can develop. Petrous apicitis can also induce formation of a nasopharyngeal abscess, dural phlegmon, extradural or subdural empyema. Cranial involvement is thought to result from direct spread of inflammation at the petrous apex.

37.5.3 Osteomyelitis of the Skull Base (Necrotising or Malignant Otitis Externa)

This is an invasive infection of the cartilage and bone surrounding the external ear as a result of extension of an existing infection, usually otitis externa. This rapidly progresses into an osteomyelitis of the temporal bone. Most cases occur in diabetic patients who may have a more favourably increased pH in their cerumen, as well as being immunosuppressed. Since both ageing and diabetes are associated with abnormalities of small blood vessels, it has been suggested that microangiopathy in the ear canal predisposes elderly diabetic patients, making them particularly susceptible. Spread of the disease beyond the external auditory canal is believed to occur through the fissures of Santorini (two vertical fissures in the anterior portion of the external auditory cartilage, filled by fibrous tissue) and the tympanomastoid suture, leading to involvement of the stylomastoid and jugular foramina and eventually affecting nearby cranial nerves. This is therefore a serious complication of otitis externa which if unrecognised can result in progressive osteomyelitis. The most commonly causative organism is *Pseudomonas aeruginosa*, although other organisms such as *Proteus mirabilis*, *Aspergillus fumigatus*, Proteus sp., Klebsiella sp., and staphylococci have been isolated. More recently widespread use of oral and topical fluoroquinolones, used in the treatment of otitis may have contributed to the emergence of *P. aeruginosa* strains resistant to ciprofloxacin. Furthermore, increasing reports of malignant external otitis in patients infected with the human immunodeficiency virus (HIV) suggest that immunocompromised patients (diabetics, immunosuppressive medication, etc.) are at especially high risk. This includes patients who have previously undergone irradiation to the area. These two facts are a recipe for high mortality in at risk groups (Figs. 37.15 and 37.16).

Typically, the disease begins as an infection of the soft tissues surrounding the external auditory canal. Patients then present with severe and unremitting throbbing otalgia, which is seemingly out of proportion to the clinical signs, purulent otorrhea, the sensation of a blocked ear, and hearing loss. Disproportionate pain (anywhere in the body) can often signify major pathology. It is commonly a sign of ongoing tissue

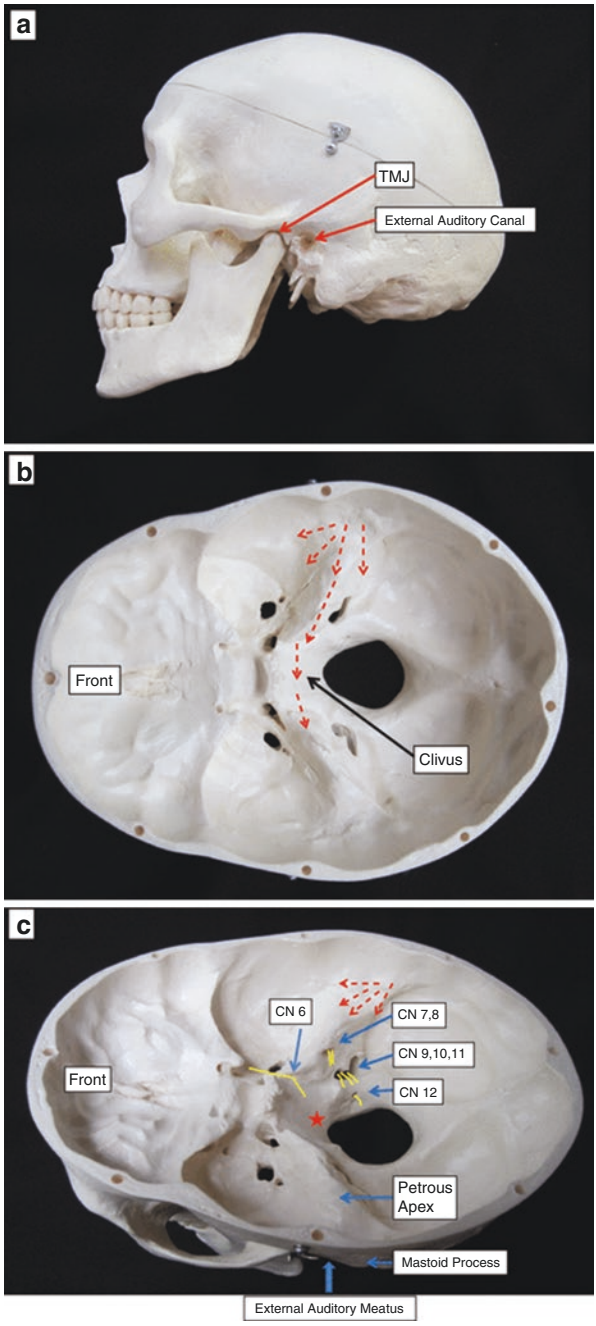

Fig. 37.15 Skull picture showing pattern of spread of necrotising otitis externa (NOE). (**a**) Note the proximity of the external auditory meatus to the temporomandibular joint (TMJ), a structure often involved in NOE. (**b**) Infection may spread (dotted red lines) across the skull base to the clivus and to the opposite side. (**c**) Approximate location of various cranial nerves (CN) that can be involved in MOE. The clivus is indicated by a star, and dotted red arrows indicate potential direction of spread of infection in MOE from the external auditory canal (photographs by Dr. Marlene L. Durand)

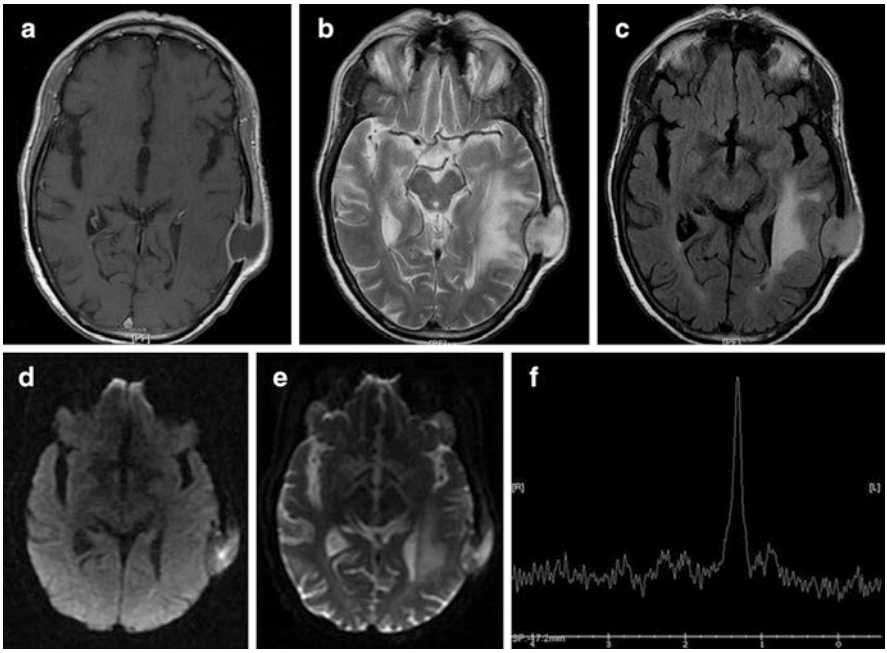

Fig. 37.16 MRI picture of Chronic temporoparietal osteomyelitis

ischaemia and infarction and therefore needs urgent assessment and treatment. On examination there may be granulation tissue in EAM and if extensive, multiple cranial nerve palsies. Thus the combination of deep seated pain, aural discharge and cranial nerve palsies should be taken very seriously. Diagnosis requires urgent CT of the temporal bones. Soft tissue changes may be more obvious on magnetic resonance imaging, but the sensitivity of MRI for detection of early bone destruction is much less than CT. Nevertheless dural involvement and medullary changes of the bone will be shown better on MRI. Radionuclide studies may also be required to assess the response to treatment. Since the clinical features of osteomyelitis overlap with malignancy, urgent biopsy of the bone/biopsy and culture of the granulation tissue will be necessary. Malignant otitis externa requires urgent diagnosis and treatment. Untreated or late diagnosis can lead to multiple cranial palsies, sinus thrombosis, septicaemia, and intracranial infections, including meningitis. This condition therefore carries a significant mortality. Treatment is usually a prolonged (6 week) course of IV antibiotics (e.g., ciprofloxacin), confirmed by culture and sensitivity. Alternatively piperacillin, tazobactam and ahminoglycosides in varying combinations may be prescribed. Treatment also includes topical ciprofloxacin/dexamethasone. Hyperbaric oxygen may be a useful adjunctive, but its definitive

role remains to be seen. Meticulous control of diabetes is essential. Frequent debridement is necessary to remove granulation tissue and any purulent discharge. Surgical debridement to clear necrotic tissue may be required for more extensive infections.

37.6 Disturbances in Hear: Loss of Hearing (Deafness)

The most quiet sounds that people can hear are between 25 and 40 decibel (dB).

Hearing loss is extremely common. It has a wide spectrum of severity, ranging from barely perceptible loss, to profound deafness. It can present at any age. Genetic causes of hearing loss are varied and has received a lot of attention in recent years. Approximately 1% of all human genes are involved in the hearing process in some way and more than 130 genetic loci have been linked to non-syndromic hereditary hearing loss. It has been estimated that at least two-thirds of cases of childhood onset hearing loss have a genetic cause, with the remaining one-third caused by environmental factors, such as cytomegalovirus infection, meningitis and acquired conductive loss. Many cases of late onset progressive hearing loss are also genetic in origin, including hearing loss associated with ageing.

Around 1 in 10 adults have some degree of hearing loss and one-third of the population over the age of 65 has loss of hearing that is sufficient enough to require a hearing aid. Hearing loss can be caused by a number os factors, the commoner ones including genetic-related conditions, age, severe or chronic exposure to noise, specific drugs and chemicals and trauma. It can result from disorders involving the pinna, external auditory canal, middle ear, inner ear, or the central auditory pathways i.e. anywhere along the auditory pathway. In general, lesions involving the pinna, external auditory canal, or middle ear result in conductive hearing loss, whilst sensorineural hearing loss arises from lesions in the inner ear, or conditions involving the vestibulocochlear nerve. Sensorineural hearing loss is a complex process that can result from damage to the hair cells caused by intense noise, viral infections, fractures of the temporal bone, meningitis, cochlear otosclerosis, Ménière's disease and ageing. A variety of drugs can also produce sensorineural hearing loss (notably salicylates, quinine, aminoglycosides, loop diuretics and some chemotherapeutic agents). Tune deafness is a central auditory processing disorder characterised by the inability to discriminate pitch, reproduce melodies or to recognise deviations in melodic structure, in spite of normal hearing. The cause of this disorder is unknown.

Common and Important Causes of Loss of Hearing

1. External ear (pinna, external auditory meatus and canal)—congenital, infection, trauma, tumour, dermatological conditions and cerumen
2. Middle ear (tympanic membrane, ossicles, middle ear cleft)—congenital, eustachian tube dysfunction, infection, tumours, otosclerosis, tympanic membrane perforation, barotrauma and vascular

3. Inner ear (cochlea, vestibule, and semicircular canals)—congenital or hereditary, presbycusis, infection, ménière disease, noise exposure, barotrauma, trauma, acoustic neuroma, meningioma, endocrine/systemic/metabolic/autoimmune conditions, multiple sclerosis, paget's disease, drug induced and neurogenic.

Hearing impairment following mild head injury can be due to central or peripheral causes, middle ear or cochlea being the most common site of peripheral injury. Causes include hair cell damage and degeneration of the organ of corti, ischaemia of the eighth nerve and damage of central auditory pathways (from damage to the blood supply to the inner ear), either partly or totally. In most cases hearing impairment resolves during recovery, but some times it may persist or even progress.

37.6.1 Hearing Loss in Children

Deafness in children is an important and often treatable cause of developmental delay. Early detection and treatment are essential. Congenital malformations of the inner ear can also cause late hearing loss in some adults. More than 200 syndromes are known to be associated with hearing loss. Otitis media with effusion is the commonest cause in children with around 60% of cases going undetected in the first year. This commonly presents with hearing loss and otalgia. There is also a higher incidence of otitis media in children with Down's Syndrome and Cleft palate.

37.6.2 Hearing Loss in Adults

The main causes of deafness in adults are wax impaction and ageing (presbycusis, seen mostly in the over 60s). Otitis media with effusion is rare. Therefore its presence requires suspicion and further investigation to exclude neoplasia. Otosclerosis, noise induced and ototoxic drugs are also common causes. Progressive unilateral sensorineural hearing loss in adults should also raise suspicion for an acoustic neuroma. Hitselberger's sign may be present—loss of sensation in the ear canal, supplied by Arnold's nerve—the auricular branch of vagus nerve, occurs due to involvement at the cerebellopontine angle.

The mechanics and physiology of normal hearing have previously been discussed. Sound vibrations pass from the external ear, through the middle ear to the inner ear. Within the cochlea, thousands of nerve fibres detect a range of frequencies and transmit impulses to the brain. In conductive hearing loss (CHL), vibrations cannot pass from the outer ear to the inner ear. In sensorineural hearing loss (SNHL), there is a dysfunction in the inner ear. In mixed hearing loss, there is a combination of conductive and sensorineural hearing loss. Depending on the cause, hearing loss can be mild or severe, temporary or permanent. It can also be bilateral or unilateral.

37.6.3 Diagnosing Hearing Loss

Hearing loss can be classified according to severity. Hearing loss between 26 and 40 dB is considered mild, 41 and 55 dB moderate, 56 and 70 dB moderately severe, 71 and 90 dB severe and greater than 91 dB profound. In the first instance, clinical assessment can often suggest the underlying cause and severity, but audiometry and imaging are often required. Clinical assessment includes tuning fork tests and repeating certain words at differing intensities and different distances to each ear in turn. By convention this is recorded as, for example "WV @ 300cm"—whispered voice at 300 cm. More precise quantitative evaluation requires audiometry. The patient listens to a number of different pure tone signals at various frequencies through headphones. This assesses air conduction. The threshold of hearing for each ear is noted. Bone conduction is assessed by placing a bone conductor on the mastoid process. This sends tiny vibrations to the inner ear. Comparing air conduction with bone conduction thus provides an indication of whether the hearing loss is due to conduction deafness or nerve deafness. Hearing loss is classified into three main types—conductive, sensorineural, and mixed hearing loss.

37.6.4 Conductive Hearing Loss (CHL)

This is characterised by hearing that is better with bone-conduction, compared to air-conduction (Rinne test). CHL is usually associated with a problem within the external and/or middle ear, whilst inner ear function remains normal. Most cases are treatable with medication, surgery, amplification devices, or a combination of these. A common cause of congenital conductive hearing loss is the absence or malformation of the external ear, acoustic meatus, auditory canal, or middle ear structures (atresia and microtia). In adults, CHL can arise following otosclerosis, cholesteotoma formation and tympanosclerosis. Other common causes include excessive earwax, perforated or scarred tympanic membrane, otitis externa, otitis media, trauma (to the tympanic membrane or ossicles), middle ear effusion, Eustachian tube dysfunction and some tumours.

37.6.5 Sensorineural Hearing Loss (SNHL)

This is hearing loss that occurs as a result of damage to the cochlea, vestibulocochlear nerve or the brain. It can result in complete loss of hearing, despite the external ear and middle ear being normal. Individuals with SNHL demonstrate both air and bone conduction defects, since the 'final common pathway' is disrupted. SNHL can occur following perinatal infections (rubella, herpes, toxoplasmosis, syphilis and cytomegalovirus). In childhood and adults, causes include meningitis, labyrinthitis, mumps, scarlet fever and measles. Other

causes include malformation of the inner ear, ageing, Meniere's disease, drug-induced ototoxicity, and tumours (notably acoustic neuroma). Chronic exposure or sudden exposure to a very loud noise can result in profound SNHL, often accompanied by tinnitus. Due to the delicate pathophysiological mechanisms involved in SNHL, it often cannot be treated.

Most cases of SNHL present with a gradual deterioration in hearing over many years or decades. Diagnosis is usually made using pure tone audiometry. Sudden sensorineural hearing loss is more worrying as this can be associated with acoustic neuroma, perilymphatic fistula, Ménière disease, vascular insufficiency, multiple sclerosis, and other centrally sited pathologies. Although the primary cause of sudden sensorineural hearing loss is almost always viral or ischaemic, patients need to undergo CT or MRI to exclude tumours.

37.6.6 Mixed Hearing Loss

This is a combination of conductive and sensorineural damage in the same ear. While the conductive component may be treated, the sensorineural component often cannot.

37.6.7 External Ear Causes of Hearing Loss

Cerumen (Wax) This is probably the most common cause of conductive hearing loss. Some patients regularly use cotton buds (Q-tips) to clean their ears. However, contrary to popular belief this is not necessary—the other third of the external auditory canal is self cleansing. Instrumentation of the canal can push cerumen further down the canal. Over time this can build up and become impacted, resulting in conductive hearing loss. Patients may then need periodic cleaning.

Infection Infections may result in swelling and blockage of the external auditory canal. If obstruction is complete, conductive hearing loss results.

Congenital Conductive hearing losses that result from congenital malformations range from mild to severe. Embryologically the pinna and external auditory canal are derived from completely different tissues. One may develop fully while the other develops abnormally. It is therefore possible to have a normal pinna but an atretic canal. Malformation of the pinna is termed anotia (complete absence) or microtia (under development). This may result in mild to moderate conductive hearing loss only, because the external auditory canal is still functional. However congenital atresia of the external auditory canal results in moderate to severe conductive hearing loss—sound is unable to reach the tympanic membrane (which may also be defective).

Tumours Malignancy of the external auditory canal is rare. The most common type is squamous cell carcinoma. Others types include basal cell carcinoma, adenoid cystic carcinoma, adenocarcinoma and melanoma. Benign growths of bone may also occlude the external auditory canal, resulting in conductive hearing loss. The two most common benign bone growths are exostosis and osteoma. Benign polyps can occasionally grow large enough to occlude the lumen of the external auditory canal.

Trauma Hearing loss due to trauma is a common phenomenon worldwide. Trauma to the head can result in conductive hearing loss, commonly involving the external ear, middle ear and temporal bone. Penetrating trauma to the external auditory canal or meatus may cause mild or profound conductive hearing loss from a variety of mechanisms (bleeding, swelling, perforation etc.). Trauma to the tympanic membrane and the middle ear can be caused by (1) overpressure, (2) thermal or caustic burns, (3) blunt or penetrating injuries, and (4) barotrauma. Slap injuries usually result in a tear in the tympanic membrane.

37.6.8 Middle Ear Causes of Hearing Loss

Congenital Malformations of the ossicles can cause conductive hearing loss. An abnormal incus or malleoincudal joint is a common cause.

Eustachian Tube Dysfunction Temporary disturbances in function often occur during upper respiratory infections and allergies. Chronic dysfunction or dysfunction resistant to treatment should raise suspicion of a tumour. Negative pressure develops in middle ear as gas is absorbed, resulting in a reduction of tympanic membrane mobility and conductive hearing loss.

Infection Acute otitis media (AOM) results in conductive hearing loss. Middle ear effusion restricts tympanic membrane mobility. In severe cases the middle ear may become filled with thick fluid.

Tympanic Membrane Perforation Conductive hearing loss is very common following perforation. The size, location, and nature of perforation will affect the degree of hearing loss and treatment required. Small perforations in the anterior–inferior quadrant cause the least amount of loss. Most perforations heal spontaneously.

Otosclerosis Otosclerosis is a disease of the temporal bone, which results in ankylosis of the stapes. Hearing loss is the main symptom. Tinnitus and vertigo may develop later.

Cholesteatoma This is persistent growth of desquamated epithelium within the middle ear space. Untreated this will continue to expand with eventual erosion into

bone. This can progress to involve the tegmen, sigmoid sinus, or inner ear, resulting in a labyrinthine fistula (with severe sensorineural hearing loss and vertigo). Conductive hearing loss can also occur if one or all ossicles become involved.

Tumours Malignant tumours (langerhans cell, histiocytosis and squamous cell carcinoma) can occur in the middle ear and can cause conductive hearing loss. These are rare.

Barotrauma Sudden, or large change in the ambient pressure which cannot be equalised with those in the middle ear can lead to an effusion or bleeding. Rupture of the tympanic membrane can also occur.

Vascular Glomus tumours are the most common benign neoplasm in the middle ear. They arise from paraganglionic tissue and may rarely develop malignant potential. As the tumour grows, it tends to fill the middle ear, resulting in pulsatile tinnitus with or without conductive hearing loss. They can also erode bone, resulting in damage to nearby structures (notably large vessels and cranial nerves). Tumours can also disrupt the ossicles and tympanic membrane.

37.6.9 Inner Ear Causes of Hearing Loss

Congenital Non-hereditary conditions involving the developing cochlea include viral infections (cytomegalovirus, hepatitis, rubella, HIV), toxoplasmosis and syphilis. Some teratogenic medications may also affect development in the foetus. These include recreational drugs, alcohol, quinine, and retinoic acid. Sensorineural hearing loss can also be inherited. Congenital malformations of the inner ear vary from complete atresia to a cavity within the cochlea. The most common malformation is a Mondini deformity, where the normal two-and-one-half turns of the cochlea are reduced to a single, or one-and-one-half turns. Malformations of both the inner or the middle ear are at risk of developing perilymphatic fistulas. These can also cause sensorineural hearing loss.

Presbycusis (see later). This is age-related hearing loss. It is a common cause of sensorineural hearing loss. It is a complex and multifactorial disorder, in which there is progressive loss of hearing in both ears over many years. It usually affects high frequencies. Tinnitus is often present.

Infection The most common infection of the inner ear is viral cochleitis in adults, and meningitis in young children. Meningitis can cause profound sensorineural hearing loss by permanently destroying the inner ear hair cells. Viral cochleitis usually manifests as a sudden sensorineural hearing loss and vertigo.

Ménière's Disease (see later). This is diagnosed by (1) spontaneous episodes of vertigo lasting several minutes to hours, (2) low-pitched tinnitus occurring during an attack, (3) fluctuating low-frequency sensorineural hearing loss and (4) aural fullness.

Noise Exposure Constant exposure to loud noises can cause high-frequency sensorineural hearing loss. Loud noises can result in severe to profound sensorineural hearing loss, otalgia, or hyperacusis. Blast energy results in initial mechanical damage to the structures in the cochlear, which is then followed by secondary metabolic disturbances. Excess nitric oxide and oxygen free radicals can damage hair cells and is toxic to membranes. Low magnesium concentrations weaken the hair cells.

Barotrauma This is an uncommon injury to the inner ear, but should be excluded in all cases of middle ear barotrauma. A sudden pressure differential between the inner and middle ear, can result in rupture of the round or oval window. Symptoms include tinnitus, vertigo and hearing loss. Labyrinthine fistula and leakage of perilymph can also occur, resulting in permanent damage to the inner ear.

Trauma This can result in sensorineural loss secondary to concussive forces transmitted through the inner ear fluids and temporal bone fracture.

Tumours Most tumours of the inner ear are benign, although squamous cell carcinoma, sarcomas, adenoid carcinoma and metastasis can occur. Fibrous dysplasia and Paget disease are also rare. The most common tumour that causes sensorineural hearing loss is an acoustic neuroma.

Endocrine Various metabolic abnormalities can cause sensorineural hearing loss.

Autoimmune Wegener granulomatosis, Cogan syndrome, rheumatoid arthritis, systemic lupus erythematosus, and polyarteritis nodosa can all involve the inner ear. Hearing loss is usually sensorineural, bilateral and asymmetric, which fluctuates and progresses.

Ototoxicity and Chemically Induced Many medications are known to cause damage to the ear. Anti-inflammatory, antibiotics, loop diuretics, antimalarials, chemotherapeutic agents, and ototopical medications may all result in ototoxicity. Exposure to chemicals at work can lead to occupational chemical-induced hearing loss. Solvents, pesticides and certain metals (copper, mercury, arsenic, lead etc.) can cause both hair cell dysfunction and central auditory dysfunction.

Neurogenic These include strokes, transient ischaemic attack, Arnold–Chiari malformations (which may stretch the vestibulocochlear nerve) and multiple sclerosis.

37.6.10 Presbycusis

The term presbycusis refers to gradual sensorineural hearing loss that is associated with complex degenerative changes in the hair cells of the cochlea and central auditory connections, that occur with ageing. It is considered the most prevalent sensory

impairment in the elderly, affecting individuals aged 75 years and older. A 1995 UK national study of hearing disorders found that 20% of adults had some degree of hearing impairment (audiometric threshold greater than 25 dB) in the better hearing ear of which 75% were over 60 years of age. By definition, presbycusis is (usually) bilateral, symmetrical, and slowly progressive. It is most noticeable at higher frequencies, with audiometric threshold shift, impaired localisation of sound, deterioration in speech-understanding and speech-perception difficulties in noisy environments. Diagnosis is based on the medical history and physical examination. It is usually made when patients meets the following criteria

1. Symmetric increased hearing threshold
2. Absence of injury, use of ototoxic medications, history of ear disease and previous ear surgery
3. The presence of minimum conductive hearing loss (10 dB or lower) and
4. Aged 65 years or older.

The aetiology of this condition is complex and multifactorial with both environmental and intrinsic factors contributing. It has been classified into six categories, based on results of audiometric tests and temporal bone pathology. These are (1) sensory (degeneration of outer hair cells), (2) neural (degeneration of neurones and spiral ganglia), (3) metabolic or strial (atrophy of stria vascularis), (4) cochlear conductive (thickening and secondary stiffening of the basilar membrane of the cochlea), (5) mixed and (6) indeterminate types. Risk factors include chronic or severe noise exposure, alcohol, hypertension, diabetes, ototoxic medications and a family history. Histological changes associated with ageing have been identified throughout the auditory system from the hair cells of the cochlea to the auditory cortex in the temporal lobe of the brain. These may correlate with clinical findings and auditory test results. Mitochondrial DNA deletions and complex metabolic changes have also been found to be associated with age-related hearing loss. Reduced perfusion of the cochlea as a result of increasing age may result in the formation of reactive oxygen metabolites, which may adversely affect the inner ear neural structures and damage mitochondrial DNA. This may then cause reduced oxidative phosphorylation, which may lead to neural dysfunction in the inner ear. More recently it has been suggested that there is a genetic basis for this disease. Reactive oxygen metabolites have also been implicated, suggesting that antioxidant treatments might help reduce hearing loss. To date presbycusis remains a diagnosis of exclusion. In mild cases reassurance and advice are all that are needed. In more severe cases hearing aids can be of benefit.

37.6.11 Sudden vs. Gradual Onset of Sensorineural Hearing Loss

Sudden onset of unilateral hearing loss, may occur in isolation or be associated with vertigo and tinnitus. This may occur following an inner ear viral infection or a vascular accident, but the differential diagnosis is wide and includes many disorders

both within and extrinsic to the ear, including systemic and metabolic diseases. One important differential diagnosis of sudden hearing loss is intralabyrinthine schwannoma. Patients usually complain of reduced hearing, poor sound localisation and difficulty hearing clearly in the presence of background noise. Diagnosis requires detailed assessment of the patient's history, otoscopic findings, tuning fork tests, and pure tone audiometry followed by targeted audiological and imaging. Work up can be quite complex and may include

1. Tympanoscopy
2. Otoacoustic emissions (OAE), Auditory evoked brainstem potentials (ABR), Speech audiometry, Stapedius reflex measurement
3. Laboratory tests: blood glucose, CRP, procalcitonin, full blood count and differential, creatinine, fibrinogen level
4. Serologic testing: borreliosis, syphilis, herpes simplex virus type 1, varicella zoster virus, CMV, HIV
5. Duplex sonography
6. MRI—to exclude a tumour of the cerebellopontine angle.
7. CT of the skull, temporal bone, cervical spine
8. Glycerol test
9. Electrocochleography to assess for cochlear damage
10. Electronystagmography or video-oculography

Gradual loss of hearing is common with otosclerosis, noise-induced hearing loss, acoustic neuromas and Ménière's disease. Hearing loss associated with otorrhoea is most likely to be due to chronic otitis media or a cholesteatoma.

37.6.12 Management of Hearing Loss

This depends to some extent on the underlying cause. Early detection and intervention of hearing loss is the most important factor in minimising the impact of hearing loss on a child's development and educational achievements.

Reversible conditions should always be treated. To improve sound perception several options are available

1. Hearing Aids: These vary in size, site configuration and strength.
2. Cochlear Implants: Over the last few decades cochlear implants have become recognised as highly successful auditory rehabilitation devices for individuals with severe to profound hearing impairment. Criteria for these are undergoing constant revisions. Cochlear implants convert sound energy to electrical signals and thus can be used to stimulate the auditory component of the vestibulocochlear nerve directly. A microphone picks up sound that is sent to an external speech processor (placed on the body or at ear level). This processor converts the vibrational sound energy into an electrical signal that is transmitted via surgically implanted electrodes into the cochlea and from there to the nerve. With the

latest generation of multichannel cochlear implants, almost three-quaters of patients with these implants can use a telephone.
3. Gene therapy and stem cell research are novel treatments but are still at the early experimental stage.
4. Sensorineural hearing loss treatment has consisted mostly of systemic steroids. This has been limited by their side effects and low concentrations attained within the fluids and tissues of the inner ear. Local application of steroids (dexamethasone and methylprednisolone) through the eardrum, into the middle ear has been used to treat a variety of otologic disorders, including Meniere's disease and sudden sensorineural hearing loss. These are believed to act on the inflammatory processes that contribute to the aetiology of various inner ear pathologies. Direct application bypasses the blood-labyrinthine barrier and results in higher concentrations in the inner ear fluids, thereby avoiding unwanted effects of systemic drugs.

37.6.13 Auditory Processing Disorder

This is also known as Central Auditory Processing Disorder (CAPD). It is a general term used for a variety of disorders that affect the way in which the brain processes auditory stimuli. Individuals with CAPD usually have normal ears and peripheral hearing. However, they cannot process the information in the normal way, resulting in difficulties in recognising and interpreting sounds. This includes speech. It is thought that this condition arises from a dysfunction located in the central nervous system. CAPD can affect both children and adults. Males are twice as likely to be affected than females. The condition can be congenital or acquired. It can arise following ear infections, head injuries and some developmental delays. Due to its somewhat obscure nature is a difficult disorder to diagnose. Symptoms include an intermittent inability to process verbal information, leading the patient to guess what is being said. Consequently this can be confused with other causes of odd behaviour, or early dementia.

37.6.14 Hyperacusis

This is an increased sensitivity to certain frequencies of sound. When severe, everyday sounds can become poorly tolerated and sometimes painful. It can occur following injury to the inner ear and has also been linked to a variety of other disorders (Bell's palsy, Lyme disease, Ramsay Hunt syndrome, perilymphatic fistula, migraine, medications, Addison's disease and hyperthyroidism). The most common cause is overexposure to excessively high levels of sound or a blast wave (explosion, gunfire, airbag). In other cases it can occur as a result of damage to the CNS. In rare cases hyperacusis may be caused by a vestibular disorder (vestibular hyperacusis). This results in the unpleasant perception of both sound and motion.

The pathophysiology of hyperacusis is unknown but may be related to genetic factors, stress, ill-health and abnormal actions of the tensor tympani and stapedius muscles. In cochlear hyperacusis (the commonest form), symptoms include otalgia and a general intolerance to sounds. Everyday sounds (shutting doors, ringing phones, ticking clocks etc.) may cause varying levels of annoyance. In vestibular hyperacusis, the patient may experience vertigo, nausea, or loss of balance when certain sounds are present. Management of hyperacusis includes advising patients to use ear protection and desensitisation (retraining therapy using broadband noise). By regularly listening to this at low levels, patients can develop a degree of tolerance.

37.7 Disturbances in Hearing: Abnormal Sounds (Tinnitus)

37.7.1 Hyperacusis

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37.7.2 Tinnitus

Also called an auditory phantom sensation) is not a disease but is a symptom. It is defined as the perception of an abnormal noise by the patient, in the absence of a corresponding external source. Whilst often described as a 'ringing', it can take many forms such as a clicking or a roaring sound, or a hiss. Tinnitus is reported to be present in 5–15% of the population and in 70% of patients with hearing disturbances. It can occur as a result of a number of conditions affecting the external, middle and inner ear, as well as pathology beyond the ear. Beside the hearing system, possible causes of tinnitus - cardiovascular, musculoskeletal, metabolic, and

endocrine dysfunctions (dyslipidaemia, thyroid dysfunction, diabetes mellitus). Stress also may contribute to the perception of tinnitus. The character of this sound can sometimes provide a clue to the diagnosis—pulsatile tinnitus, which coincides with the patient's heart rate and pulse, suggests a vascular related cause. Severity of symptoms may vary considerably, from a minor annoyance, to symptoms so distressing that some patients have been reported to have considered suicide.

In many cases the pathophysiology of tinnitus is unknown. Several hypotheses have been suggested, many involving abnormalities of the neural pathway from the cochlear to the auditory cortex. These include (1) damage to the hair cells, resulting in excessive stimulation of auditory nerves, (2) increased activity in the auditory complex and (3) overactive auditory nerves. A common cause of tinnitus is noise-induced hearing loss. Other known causes include (1) ear infections, (2) cardiovascular disease, (3) Ménière's disease, (4) tumours, (5) previous head injury, (6) certain medications and (7) earwax. Tinnitus is also more common in patients with depression.

Tinnitus can be classified as subjective or objective. Subjective tinnitus is the perception of sound in the absence of any identified acoustic or external stimuli. It is more common than objective tinnitus and is usually associated with high frequency hearing loss in the elderly. Objective tinnitus is perception of sound caused by an internal source, such as a body sound or vibration (bruit, hum, palatal myoclonus). This can often be identified on examination or testing. If tinnitus is unilateral and accompanied by unilateral sensorineural hearing loss, this may indicate an acoustic neuroma.

37.7.3 Subjective Tinnitus

Subjective tinnitus refers to the perception of sound that is not audible to anyone else. The precise pathophysiology of subjective tinnitus is largely unknown, although in some cases it may involve the subcortical auditory pathways. Known underlying causes are many and can involve the auditory pathway anywhere from the external auditory canal to the auditory nerve. These include

1. Wax impaction
2. Insects
3. Otosclerosis
4. Glue ear
5. Noise induced
6. Presbyacusis
7. Ménière's disease
8. Trauma/TM perforation
9. Ototoxic drugs
10. Labyrinthitis
11. Acoustic neuroma
12. TMJ disorders

Time	No associated hearing loss	Hearing loss present
Seconds	Benign positional paroxysmal vertigo	Perilymphatic fistula Cholesteatoma
Minutes	Vertebral basilar insufficiency Migraines	
Hours	Vestibulopathy	Ménière's disease
Days	Vestibular neuronitis	Labyrinthitis
Weeks	Central nervous system disorders Lyme disease Multiple sclerosis	Acoustic neuroma Autoimmune Psychogenic

Damage to cochlear hair cells is responsible for most of the common causes of subjective tinnitus. A number of systemic illnesses have also been implicated, sometimes referred to as the "metabolic syndrome". These include severe anaemia, obesity, cardiovascular disease, hyperlipidemia, low thyroid function, multiple sclerosis and syphilis. Medications, chemotherapeutic agents and heavy metals can result in ototoxicity and tinnitus. Tinnitus has been suggested to be a "soft" sign for cerebrovascular disease and secondary endolymphatic hydrops. Some nutritional deficiencies have also been implicated in sensorineural hearing loss, vertigo and tinnitus. The biological mechanisms leading to these perceptions of tinnitus are however, not understood. Salicylates, such as aspirin, can also damage cochlea neurones.

37.7.4 Objective Tinnitus

This is tinnitus which can be verified on examination or following investigations. In some patients it can be audible using a stethoscope or Doppler and is often described as a clicking or pulsing sound. Causes include arterial or venous abnormalities, neurological lesions or eustachian tube dysfunction.

Vascular Causes Arteriovenous malformations and fistulas are an important cause that needs to be identified early.

1. Congenital arteriovenous malformations are generally asymptomatic and are uncommon, whereas acquired arteriovenous shunts are more likely to be symptomatic.
2. Dural arteriovenous fistulas can arise secondary to dural venous sinus thrombosis following trauma, infections, neoplasms or surgery. Mortality from intracranial haemorrhage ranges from 10 to 20%.
3. Acquired arteriovenous shunts can also arise from a paraganglioma in the temporal bone (glomus jugulare or glomus typanicum tumour). These are heard as a constant 'blowing' or 'whooshing' sound. As the tumour enlarges patients may develop hearing loss and multiple cranial nerve deficits (VII–XII especially).

4. Rarely, tinnitus may be caused by a dissecting aneurysm in the internal auditory canal or the vertebral artery. Symptoms include tinnitus, pain, Horner's syndrome, cranial nerve deficits, subarachnoid haemorrhage, and transient ischaemic attacks.
5. Aberrant Internal Carotid Artery. This is a rare condition characterised by the presence of the ICA inside the middle ear cavity. It may arise following perforation of the normally thin separating bone by a small branch of the ICA called the "caroticotympanic artery," (a remnant of the embryologic hyoid artery). This anastomoses over the cochlear promontory with the inferior tympanic artery, a branch of the ascending pharyngeal artery and both enlarge. Alternatively, dehiscence of the normal separating bone allows the ICA to herniate inside the middle ear cavity.
6. Tinnitus may also occasionally be the presenting symptom of atherosclerosis in the carotid artery.
7. Idiopathic intracranial hypertension. This is often described as a 'whooshing' noise, rather than a 'ringing' and can be positional or aggravated by head movement. This usually occurs in young obese women. The combination of a headache with pulsatile tinnitus should raise suspicion for this. In such cases it is important to check the fundi for papilloedema. Untreated intracranial hypertension may lead to permanent visual loss.
8. Venous 'hums' may be heard in both systemic or intracranial hypertension. A "dehiscent jugular bulb" refers to its abnormally high location, which extends into the middle ear space. This results in a low-pitched, soft hum that decreases with activity or pressure on the jugular vein.
9. Persistence of the stapedial artery is a rare vascular anomaly that can present in adulthood with pulsatile tinnitus. This artery is part of the initial circulation in the embryonic head and neck. Embryologically, the primitive hyoid artery gives rise to the stapedial artery. Normally the hyoid artery regresses, the middle meningeal joins the internal maxillary artery and the stapedial artery becomes a small branch of the posterior tympanic artery. Sometimes the stapedial and hyoid arteries fail to regress and persist to continue to supply the middle meningeal artery territory. Persistence of the stapedial artery can be an isolated anomaly or accompanied by an aberrant ICA. It can also be seen in cases of trisomy 13, 15, and 21 as well as Paget's disease of the bone.

Venous causes of tinnitus can be differentiated from an arterial causes by placing gentle pressure over the jugular vein. This results in cessation of blood flow and resolution of symptoms. Persistent pulsatile tinnitus is an important group of causes to identify. These result from altered blood flow. Rarely, it can be a symptom of potentially life-threatening conditions such as carotid artery aneurysm or carotid artery dissection. It may also occur in some vasculitides, such as giant cell arteritis. Pulsatile tinnitus can also be an indication of idiopathic intracranial hypertension. Hence this symptom often requires imaging to rule out paragangliomas or vascular anomalies in the middle ear.

Non-vascular Causes

1. Palatal myoclonus is caused by aberrant rapid contractions of the superior constrictor and palatal muscles. These can result in sudden closure of the eustachian tube. This condition may occur secondary to other neurological disorders, such as cerebrovascular disease, central nervous system tumours, and multiple sclerosis.
2. Stapedial muscle spasm is an idiopathic condition often exacerbated by other noises. It is benign and self-limiting. Diagnosis is made by visualising contractions of the tympanic membrane.
3. Dysfunction of the eustachian tube can also result in objective tinnitus. Patients complain of tinnitus, autophony and reverberation. Symptoms usually improve with lying down and recur on rising.

37.7.5 Assessment and Management of Tinnitus

Sudden onset of tinnitus is concerning and may indicate serious pathology. If it is pulsatile, it is often due to a vascular source, whereas Ménière's disease tends to be episodic. Ménière's also has associated symptoms include hearing loss, vertigo, and aural fullness. Patient positioning should also be asked—eustachian tube dysfunction is often alleviated by lying down. A history of hyperlipidemia or diabetes may suggest carotid artery atherosclerosis, whereas a thyroid disorder or anaemia may suggest a high cardiac output aetiology. Many medications are also known to cause tinnitus. During examination, the cranial nerves should be carefully assessed, noting especially any facial weakness, hearing loss or evidence of brainstem dysfunction. It is important to look also for papilloedema, to exclude vision-threatening intracranial hypertension. Auscultation over the auricular region, mastoid and carotid arteries should be undertaken, listening for bruits. Objective tinnitus secondary to a venous cause disappears when the ipsilateral jugular vein is compressed. Otoscopy may identify a middle-ear infection, wax, a dehiscent jugular bulb, or glomus tumour. Diagnostic tests are guided by the history and examination. This may include a full blood count, thyroid function tests, audiology, carotid ultrasonography/transcranial doppler (TCD), CT/magnetic resonance angiography (MRA) and occasionally psychological assessment. CT or MR angiography may be required urgently if there is suggestion of a dissecting aneurysm or arteriovenous fistula.

Management of tinnitus involves treating any identified causes. Tinnitus secondary to ototoxic medications may resolve after discontinuing the drug, if permitted. Other causes may require referral and possibly surgery (arteriovenous fistula/dehiscent jugular bulb/glomus tumors/carotid artery atherosclerosis). Patients with intracranial hypertension require urgent referral to neurology and ophthalmology. Other measures include

1. Psychoeducative counselling. This is recommended as a basic component of every treatment for tinnitus. Often, a sympathetic and understandable explanation of the fundamentally benign nature of idiopathic tinnitus is often all that is needed. Cognitive behavioural therapy (CBT) aims to improve awareness and

facilitate modification of maladaptive patterns on the cognitive, emotional, and behavioural level.

2. Hearing aids. These are used to compensate for any hearing loss by improving the peripheral auditory input in the affected range of frequencies. However, the influence of hearing aids on the perception of tinnitus has not been demonstrated. Observational studies suggest that patients whose tinnitus frequency lies under 6 kHz benefit most from using a hearing aid. If tinnitus is associated with severe loss of hearing, cochlear implants may be highly effective. This masks tinnitus by amplifying normal sounds
3. Individualised auditory stimulation. Most tinnitus patients are particularly bothered by their ear noise in quiet surroundings. Many sufferers find that targeted auditory stimulation reduces their tinnitus. One suggestion is to modify the frequency spectrum of music according to the individual patient's audiometric profile, thus compensating for the individual impairment.
4. Tinnitus maskers. These generate either sounds from the natural environment or individually tailored noises. However, despite their widespread use, only limited data are available from controlled trials and tinnitus masking has not been neither clearly proven to work. Devices are commercially available, but more economical alternatives such as indoor fountains or recordings of waves breaking can also be used. These are generally reserved for intractable tinnitus.
5. Tinnitus retraining therapy. This comprises a combination of counselling and auditory stimulation by maskers or hearing aids. Tinnitus counselling by hearing therapists/relaxation techniques and biofeedback techniques may also help.
6. Neurobiofeedback. Pilot studies have shown significant reductions in the intensity and loudness of perceived ear noise.
7. Repetitive transcranial magnetic stimulation (rTMS) is a procedure in which magnetic impulses are used to modulate the activity of specific brain regions. Several randomised controlled trials have found evidence for clinical efficacy of this. However, the effects are slight, varied and transient.
8. Drug therapy: Despite the investigation of a large number of drugs, no pharmacological approach is yet an established treatment option. Indications for pharmacotherapy is currently restricted to the treatment of co-morbidities such as anxiety disorders, sleep disorders, and depression. Benzodiazepines, Tricyclic Antidepressants, and Carbamezapine may therefore be of benefit
9. Referral to a tinnitus support group
10. Injection of botulinum toxin into the palate has been successfully used in patients with palatal muscle myoclonus.
11. Reported novel therapies include Ginkgo biloba extract and laser therapy.

37.7.6 Pulsatile Tinnitus (Pulse Synchronous Tinnitus)

This is an important subgroup of tinnitus that requires careful evaluation. Many conditions can result in the subjective sensation of a pulsatile noise arising from one or both ears, with or without objective confirmation. Objective pulsatile tinnitus is more likely to have a structural aetiology which can be identified by imaging. Causes can vary from minor developmental anomalies to complex vascular

malformations and acquired diseases. In all cases it is important to consider the possibility of vascular malformations in the temporal bone, paragangliomas, vascular neoplasms (including meningiomas) and metastases. The list of possible causes can be quite complex.

Temporal Bone/Middle Ear

High and/or dehiscent jugular bulb (normal variant).
- Ectopic carotid artery in the middle ear
- Persistent stapedial artery
- Carotid aneurysm
- Arteriovenous malformation
- Paraganglioma (glomus tympanicum or jugulare).
- Other vascular neoplasm
- Osteolytic phase of otosclerosis
- Dehiscent cochlea/carotid canal interface
- Dehiscent semicircular canal

Posterior Fossa and General Intracranial Conditions

Arteriovenous malformation
- Dural venous malformation
- Large draining vessels from a vascular malformation/neoplasm
- Venous variants
- Dural sinus thrombosis
- Pseudotumor cerebri
- Intracranial hypotensionb
- Neoplasm (e.g., meningioma)

Extra Cranial Causes

Paraganglioma
- Other vascular neoplasm
- Arteriovenous malformation—acquired or congenital
- Carotid aneurysm
- Atherosclerotic disease, fibromuscular dysplasia, carotid, or vertebral basilar dissection

By way of contrast, non-pulsatile (pulse asynchronous) tinnitus may be caused by less serious conditions such as palatal myoclonus and a patulous eustachian tube. These patients can often be diagnosed without the need for complex investigations.

Patients presenting with pulsatile tinnitus must be differentiated from those with tinnitus secondary to inner ear and/or cochleovestibular nerve dysfunction. Patients should be initially assessed for simple causes such as wax impaction (which can result in a ‘venous hum’). If the tinnitus can be obliterated simply by

jugular vein compression, serious pathology is less likely. The presence or absence of a mass visible behind an intact tympanic membrane is also important. The absence of one increases the differential diagnosis and can affect choice of imaging. Loss of hearing loss and dizziness should also be enquired about. Most patients present with unilateral tinnitus, but bilateral tinnitus can occur with bilateral paragangliomas or pseudotumor cerebri. Cranial neuropathy, pain and headache can also be associated with paragangliomas, vascular neoplasms, dissections and aneurysms. In most cases high-resolution temporal bone imaging and computed tomographic dual-phase arterial and venous angiography are required. Catheter angiography is rarely required but may be indicted if endovascular interventions are being considered. Management depends on the underlaying cause, some of which are noted elsewhere in this book. Patients with dissections are often anticoagulated for 3–6 months. Vascular malformations and aneurysms generally require endovascular and/or surgical techniques. Underlaying causes also need to be addressed. If an aneurysm is associated with an infection (e.g. mastoiditis with extradural empyema), this must be identified and treated with appropriate antimicrobial therapy and drainage. Dural sinus thrombosis may be an indication of an underlying disease.

37.7.7 Cerumen (Wax) Impaction

Cerumen auris, (ear wax), is a mixture of ceruminous and sebaceous gland secretions mixed with desquamated epithelium. Ears are normally self cleansing. To prevent the deeper part of the external auditory canal becoming filled with dead skin cells, the skin in this region is migratory and gradually moves from the deep canal outwards a process that is aided by jaw movement while eating and talking. Cerumen is sticky, waterproof and has a protective function. Normally there should be a thin coating of wax near the external opening of the canal. If this builds up sound perception can be affected. Accumulation can also reduce the efficiency of hearing aids. If the wax gets wet, the keratin can swell and result in sudden conductive hearing loss, tinnitus, discomfort, dizziness and chronic cough. Keratin can also become infected and develop into otitis externa.

Risk factors for wax accumulation include (1) ear canal hairs, (2) repeated use of cotton buds to clean ear canal, (3) hearing aids and ear plugs, (4) bony growths such as osteoma and (5) abnormally shaped ear canals. Children and the elderly are particularly prone. Impacted wax can become adherent to the ear canal skin and tympanic membrane and make removal more difficult. Since the deep ear canal may be wider than the opening, a large plug of dry, hard wax deep in the canal can be particularly difficult to remove. If wax needs to be removed, options include irrigation (syringing), wax softeners/solvents, irrigation following wax softeners, mechanical removal, or microsuction. Cerumen is best removed by an aural hook or micro suction, which can often carried out in a clinic.

37.7.8 Foreign Bodies/Insects

Foreign bodies in the middle ear are rare but have been reported. These usually occur as a result of higher energy perforation of the tympanic membrane and passage of the foreign body through the external auditory canal to the middle ear space. Incidents with welding bead injuries have been described. Foreign bodies in the external ear however are far more common and may present in both children and adults. In children this is a very common problem. Being curious and experimental, children tend to insert foreign bodies into their own or each other's ears often, without the knowledge of parents or guardians. These include beads, buttons, plastic toys, pebbles, popcorn kernels, paper and vegetable material. Even bullets and Bluetooth devices inserted by exam cheats have been reported. Often these are denied or there is no confirmatory history available. It is therefore important to remember that foreign bodies can present with otalgia or otorrhea. Patients may present with otalgia, discharge, bleeding, hearing loss or tinnitus, although sometimes a foreign body is an incidental finding. If the foreign body has been present for a while, visualisation can be obscured by a build up of wax. If the patient is cooperative it may be possible to attempt removal. If there are no suitable instruments, or the patient is uncooperative, this will need ENT referral for examination and removal. Persistent foreign bodies can lead to infection and the formation of granulation tissue. Batteries in contact with moisture, can result in necrosis of the skin or tympanic membrane. Insects in the ear commonly small cockroaches, can be very distressing due to discomfort created by loud noise and movement. These can present with significant excoriation or swelling of the EAM. These are more common in patients older than 10 years (Fig. 37.17).

Removal may be done with or without general anaesthesia. A good light source and a variety of instrumentation should be available, in view of the variety of objects inserted. These include forceps, wax loop, right-angled ball hook and Frazier tip suctions. Some foreign bodies can be removed by irrigation, although there must be no TM perforation. In children the first attempt at removal is critical because cooperation will decline if this fails. Firm, smooth, non-graspable foreign bodies can be difficult to retrieve and are then more likely to slip deep and lay in close contact with the tympanic membrane. Reported complications of failed removals include canal abrasion, laceration, bleeding, perforation of tympanic membrane, ossicular chain destruction, with removal of ossicles and hearing loss.

Removing a foreign body can often be done with the following steps

1. Try to reassure the patient if anxious, poorly cooperative or a child. It is crucial that they remain still during instrumentation of the canal, otherwise serious damage can occur.
2. Position the patient comfortably.
3. If the object is sticking out and easy to remove, then do so using forceps or a clip.
4. Objects within the canal can sometimes be retrieved with an aural hook, forceps or suction. Advance the instrument slowly through the external auditory canal

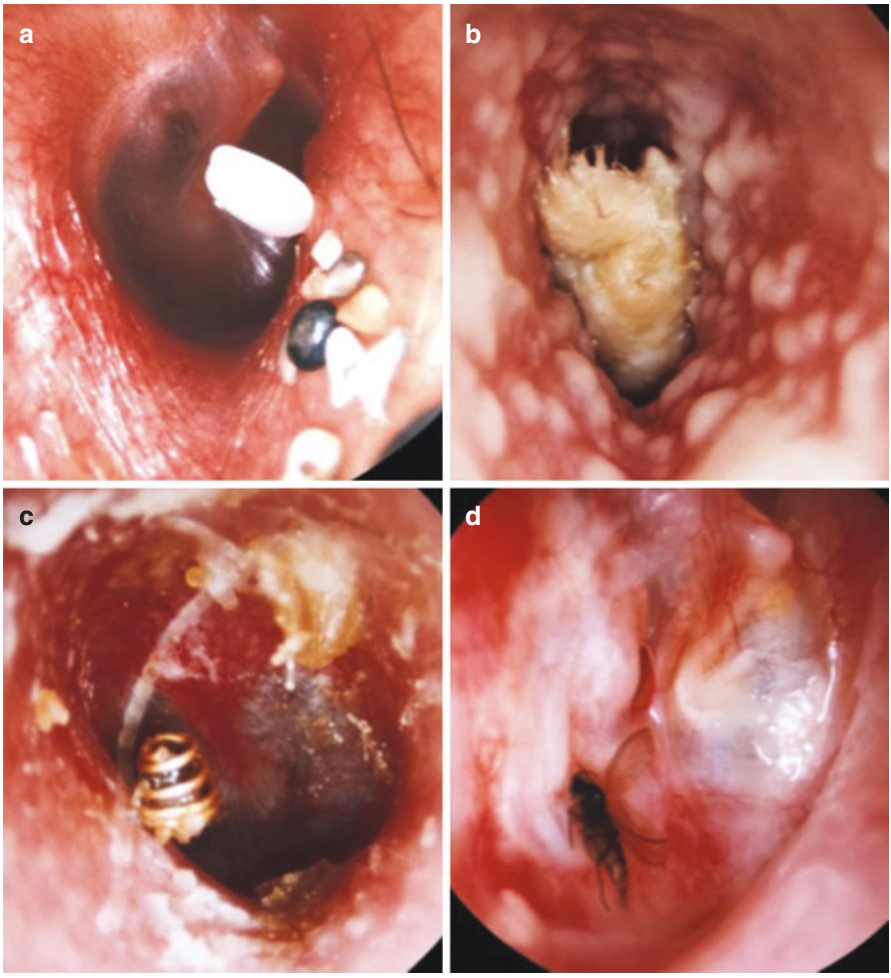

Fig. 37.17 (**a**) Sand panicles can be seen along the anterior wall of the external auditory canal. (**b**) A piece of paper has been "forgotten" inside this external auditory canal; secondary infection (external otitis) of the skin. (**c**) A metallic hearing aid component, with secondary infection of the skin of the external auditory canal. (**d**) A deceased insect on the surface of this tympanic membrane

until the foreign body is grasped. Gently withdraw from the auditory canal with the attached foreign body. If the patient is cooperative and the object smooth and not impacted, a small drop of superglue on the tip of an applicator may help. This should only be attempted if application can be done under direct vision.

5. Dead insects, organic matter, and friable objects can sometimes be broken into smaller evasive pieces are extracted with suction than with forceps.
6. Always check for complete removal of the foreign body, and for any perforation of the tympanic membrane or abrasions of the auditory canal.

7. If the object is impacted deep within the ear, or cannot be seen, do not blindly instrument the canal with forceps. This may do more harm than good. Turn the patients head to the affected side and gently shake the head. This may improve visualisation. If the object does not come out, refer.
8. Non-graspable foreign body can safely be removed by irrigation, so long as the tympanic membrane is intact, there is no history of grommet insertion and the foreign body is free in the canal. The object must also be non-organic and non-electrolytic. Irrigation is contraindicated for organic matter that can then swell through osmosis within the auditory canal. Coating the foreign body with a few drops of hydrophobic lubricant such as olive oil has been reported to facilitate removal by irrigation. To irrigate, attach a small flexible angiocatheter to a 60-mL syringe. Warming the irrigation fluid (water or normal saline) is important to avoid triggering a caloric reflex. Position the patient comfortably, drape the area and place a receiver under the affected ear. Place the catheter tip gently in the external auditory canal and slowly inject the fluid until the foreign body washes out. Do not forcibly apply pressure.
9. Cyanoacrylate adhesives (Superglue) may be removed manually within 24–48 h once desquamation occurs. If adhesive touches the tympanic membrane, remove it carefully, and refer the patient to an ENT specialist.
10. Remove batteries immediately to prevent corrosion or burns. Do not crush the battery during removal.

Removing a live insect can be complicated

1. Do not let patients try to remove the insect themselves, this may aggravate it resulting in a bite or sting.
2. Turn the patient's head so that the affected ear is up and gently shake. Wait for a few minutes and see if the insect flies or crawls out—often they will move towards light, if not trapped.
3. If unsuccessful, try to float the insect out by gently pulling the ear lobe backward and upward (adult), or backward (child) and pouring mineral oil, olive oil or baby oil into the ear. This should be warm, but not hot. However do not use this if grommets are in place or perforation is suspected (suggested by pain, bleeding or discharge). Live insects should be immobilised before removal is attempted. Oil will hopefully drown or dislodge most insects which will then float out. Water will not work. Insects can trap air in their fine hairs and remain alive for many minutes, if not longer.
4. If the insect is dead, it can be sucked or flushed out using warm water and a syringe.
5. Discuss with ENT even if this is successful, as any remains of the insect can still irritate the canal.

37.8 Disturbances in Balance: Dizziness and Vertigo

Acute dizziness/vertigo is a common cause for visiting the emergency department. Vestibular disorders can present with vertigo, chronic dizziness and imbalance. Vertigo is not, as many people think, a fear of heights. It is defined as the sensation

of movement, when an individual is motionless. It is not a single disease entity but the main symptom of a variety of different conditions. These may occur within the inner ear, brainstem, or cerebellum or may be of psychic origin. Postural vertigo may result from dysregulation of systemic blood pressure, or from adverse effects of medications such as antihypertensive or anticonvulsive drugs. Around 30% of people will suffer from rotatory or postural vertigo at some point in their lives.

Patients with any disturbance to the vestibular system can complain of 'dizziness'. Disturbances in both the peripheral or central vestibular system can result in this. Asymmetry in the electrical signals sent to the vestibular centres creates the sensation of spinning, vertigo, or falling towards the side of the injury. Nystagmus and vomiting frequently occur. A wide range of disorders affecting the middle and inner ear can result in these symptoms. They include.

1. Impacted wax
2. Acute otitis media
3. Otitis media with effusion
4. Chronic suppurative otitis media
5. Trauma (temporal bone fracture)
6. Labyrinthitis and Vestibular Neuritis
7. Ménière's disease (endolymphatic hydrops)
8. Otosclerosis/Otosclerotic drugs
9. Benign Paroxysmal Positional Vertigo (BPPV)
10. Iatrogenic (middle ear/mastoid surgery)
11. Cerebellopontine angle tumors/syndrome
12. Perilymphatic Fistulas
13. Oto Syphilis

Unfortunately there is no diagnostic confirmatory tool available for most disorders causing dizziness. Diagnosis is largely based on interpretation of clinical features derived from the history and examination. Vertigo is often associated to migraine, female gender and psychological disorders such as stress, depression and anxiety. Benign paroxysmal positional vertigo (BPPV) is the most common cause, accounting for almost 20% of all cases, followed by phobic vestibular vertigo and central vestibular syndromes (vascular, inflammatory (MS), and degenerative diseases of the brainstem or cerebellum). Vestibular migraine is the most common cause of spontaneously occurring, episodic vertigo. Together with Menière's disease and vestibular neuritis these diseases account for around 70% of all cases of vertigo. Psychogenic vertigo is a form of vertigo, which may occur with hyperventilation, anxiety, panic attacks, claustrophobia, agoraphobia and other psychiatric disturbances including schizophrenia. Metabolic diseases such as uncontrolled diabetes, renal failure, hepatic failure and electrolyte disturbances also can induce a constant feeling of dizziness. Any abnormality in blood or serum levels can drastically alter the central nervous system's ability to function properly.

Vertigo may be classified into peripheral and central. Peripheral vertigo is the most common type. Most cases are caused by a problem within the inner ear, the most common being (1) Benign paroxysmal positional vertigo (BPPV, or "crystal mediated disease"), (2) Vestibular neuronitis (a presumed viral infection) and (3)

Meniere's disease (cause unknown but has been linked to stress and consumption of salt, caffeine and alcohol). Less common causes include (4) Labyrinthitis (a presumed viral infection), (5) Perilymphatic fistula (following trauma and Superior semicircular canal dehiscence syndrome (SSCDS—thought to be secondary to a defect in part of the semicircular canal). Central vertigo is caused by diseases or injuries to the brain (brain stem or cerebellum). These include Head injuries, infections, Multiple sclerosis, Migraines, tumours, Strokes and Transient ischaemic attacks ("mini" strokes).

The traditional approach to 'dizziness' starts with categorising it into four types, (1) vertigo, (2) presyncope, (3) disequilibrium and (4) nonspecific dizziness. However, another a more practical approach in organising a differential diagnosis, is to classify dizziness/vertigo as (1) acute prolonged spontaneous dizziness, (2) recurrent spontaneous dizziness, (3) recurrent positional vertigo, or (4) chronic persistent dizziness and imbalance. The differential diagnosis of childhood vertigo differs from that of adults. Migraine-associated vertigo, psychogenic causes and vestibular neuritis/labyrinthitis are most common. Benign paroxysmal vertigo of childhood is another, but less common cause. In adults -

Acute prolonged spontaneous dizziness/vertigo—Vestibular neuritis/labyrinthitis Stroke
Recurrent spontaneous dizziness/vertigo—Meniere's disease, Vestibular migraine
Psychogenic dizziness, Vertebrobasilar transient ischaemic attack
Recurrent positional vertigo—Benign paroxysmal positional vertigo
Chronic dizziness and imbalance—Degenerative brain disorders, Bilateral vestibulopathy, Psychogenic dizziness

Vestibular neuritis and stroke are important disorders causing acute prolonged spontaneous vertigo. In most patients with recurrent attacks of vertigo, the differential diagnosis is largely limited to benign paroxysmal positional vertigo (BPPV), Meniere's disease (MD) or vestibular migraine (VM). However, an important group to recognise are those secondary to vertebrobasilar ischaemia (transient ischaemic attacks). These patients are at risk of further stroke. It is also important to remember that disorders affecting other systems (notably cardiac, cervical and neurological) can also present with 'vertigo'. In elderly patients vertigo is often multi-factorial with many disorders affecting balance.

History taking should include preceding infections, vascular risk factors (hypertension, diabetes, dyslipidemia, smoking, and cardiac disease) and associated symptoms such as headache or neck pain. Acute vertigo associated with sudden severe headache or neck pain strongly indicates a cerebrovascular disorder. It is important to enquire regarding family history, including hereditary conditions such as migraine and risk factors for cerebrovascular diseases. Associated symptoms such as pain, nausea, vomiting, hearing loss, or neurologic symptoms can help differentiate peripheral or central origin vertigo. If the cause of vertigo is central, nausea and vomiting tend to be less severe. Most causes of vertigo associated with hearing loss are peripheral, whereas neurologic symptoms such as ataxia, weakness, changes in

vision or hearing, paraesthesia, dysarthria, altered level of consciousness, or other changes in sensory and motor function suggest the presence of a central cause. The pattern of severity is also important. In Ménière's disease, vertigo initially increase in severity and decreases later, in acute vestibular neuronitis, initial symptoms are typically are severe and decrease over several days. Key aspects in the history to elicit include

1. Onset and cessation of symptoms—This provide information concerning the likely pathological processes. Vertigo that starts without warning and stops quickly, is more likely to be peripheral vertigo. Central vertigo often develops without warning and may last for a long period of time.
2. Duration and frequency—Dizziness lasting only seconds to minutes is more commonly associated with benign paroxysmal positional vertigo, 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.
3. Any relationship to head movement—if related to turning the head (looking over one's shoulders), consider a cervical spine problem.
4. Circumstance of the vertigo—If related to standing, consider postural hypotension. If related to exercise, consider cardiac causes.
5. Presence of other neurologic signs or symptoms. This may help localise any anatomical defect
6. Hearing loss and otalgia suggest a problem confined to the ear itself
7. Medications—The list of drugs that can cause vertigo or dizziness is impressive. It includes anti-convulsants, anaesthetics, anti-depressants, analgesics, anti-diabetics, contraceptives, anti-inflammatory drugs, cardiovascular drugs, sedatives, tranquillizers, cytotoxic agents, and anti-hypertensive agents.

Based on (1) the duration of each attack and (2) the presence of hearing loss, a practical differential diagnosis is possible.

Patients should undergo a full medical examination, focusing on the head and neck (including the ears, neck, cranial nerves and eyes), central and peripheral nervous system and cardiovascular system. A number of specific tests can be used to help with the diagnosis

1. Spontaneous and gaze-evoked nystagmus. Gaze-evoked nystagmus is when the eye drifts, but only in certain directions of gaze. This may indicate a brainstem related problem.
2. Positional testing—Dix-Hallpike test
3. The head impulse test (HIT), or head thrust test, may be used to differente acute vascular vertigo from a more benign disorder involving the inner ear. The examiner briskly rotates the patient's head while having the patient fixate on a target, usually the examiner's nose. During slow head movements the patient can normally maintain fixation. Rapid head movements results in a "catch-up" saccade to re align vision and the eyes to lag behind during the rapid head movement.

This catch up saccade indicates peripheral vestibular hypofunction on the side towards which the head was rotated.
4. Cerebellar tests—rapid alternating movements (e.g. finger to nose)
5. Romberg's test—A standing patient who becomes unsteady on closing their eyes may have an inner ear or spinal cord problem. The usually fall toward the affected side.
6. Unterberger's test—The patient marches on the spot for 30 s with their eyes closed. If peripheral vertigo is present, there may be sideways rotation, toward the affected side.
7. Dynamic visual activity—Look at Snellen chart with head shake (worsening by >2 lines on chart)

Imaging is not required in every case but MRI may be indicated to exclude large bacterial infections, neoplasms, or developmental abnormalities.

Management of vertigo is based on symptom relief and treatment of any underlying condition. Patients with an acute vestibular disorder associated with a middle ear infection may be prescribed steroids, antiviral drugs, or antibiotics. Symptomatic relief and other measures includes.

1. Take precautions during an attack. Avoid heights, driving, operating heavy machinery when symptomatic
2. Acute Vestibular Suppressants: These medications are prescribed for intolerable symptoms. However they can only be used in the short term. Prolonged use can delay central compensatory mechanisms. Medications include prochlorperazine, phenothiazine, meclizine, cinnarazine, diazepam and antiemetics
3. Vestibular Rehabilitation. Exercise and physiotherapy are indicated for chronic symptoms. These involve positional tasks, head movements, and oculomotor exercises to try and stimulate central compensation
4. Surgical treatment may be indicated for specific diagnoses, such as (1) intra tympanic gentamycin for Ménière's disease, (2) insertion of a bone plug in BPPV, (3) surgery to decompress the endolymphatic sac or (4) division the vestibular nerve.

37.8.1 Benign Paroxysmal Positional Vertigo (BPPV)

Benign paroxysmal positional vertigo (BPPV) is a disorder of the inner ear characterised by episodes of vertigo triggered by changes in head position. It can occur following a head injury or simply arise spontaneously, often in the elderly. It is one of the most common types of peripheral vertigo. The condition is thought to be caused by the presence of endolymphatic debris in one or more semicircular canals. Direct evidence of such debris or canaliths has been demonstrated for posterior canal BPPV. However in some cases a specific cause is never found. The condition can affect any age group but is more commonly seen in those over 50. Patients complain of vertigo lasting seconds when their head is in a certain position, but with no

associated hearing loss. 20% of patients have a preceding history of vestibular neuronitis and another 20% have a history of head injury.

BPPV occurs when small calcium crystals (otoliths) in one or more of the semicircular canals move abnormally within the endolymph. The semicircular canal then becomes stimulated. Movement of debris within the endolymph stimulates the hairs lining the canal which in turn inappropriately stimulates the vestibular nerve. The resultant asymmetrical signals from the affected side combined with the unaffected side results in the sensation of vertigo. Two theories exist—canalolithiasis and cupulolithiasis.

Canalolithiaisis refers to the presence of otoliths in the canals. These are displaced by head movements.

Cupulolithiasis involves otoliths adhered to the cupula, changing its specific gravity. Thus, the cupula is made sensible to linear accelerations, such as the acceleration of gravity.

Canalolithiasis is more common than cupulolithiasis, as the otoliths needs less mass to be symptomatic. It has been calculated that a mass of 0.087 μg is required for canalolithiasis compared to 0.69 μg in cupulolithiasis. These two variants of BPPV result in different characteristics of the nystagmus elicited by the provoking manoeuvres.

BPPV is not a serious condition and typically it resolves in 1–2 weeks. However it may recur in some people. Posterior semicircular canal BPPV accounts for up to 90% of cases. Here the otoliths fall away from the posterior canal cupula of the lowermost ear. Horizontal semicircular canal (HC) BPBV accounts for about 10% of cases. Rarely, the anterior semicircular canal (AC) is involved.

Patients usually complain of a sudden and unexpected onset of vertigo or a spinning sensation that lasts 10–20 s. This is often precipitated by a sudden change in head position, for instance rolling over in bed, getting out of bed, looking up and bending over. Nausea and vomiting are common. Other neurological deficits are absent and audiography and tympanography should be normal. If any neurological deficits are detected more serious pathology (notably TIA or stroke) should be considered. Asymmetric loss of hearing should also raise suspicion and requires further evaluation.

BPPV can usually be diagnosed by the history and by performing specific tests which expose each canal to gravity, causing a nystagmus that matches the stimulation of the canal. The Dix-Hallpike test is generally considered the gold standard for the diagnosis of posterior canal BPPV. It is designed to determine whether the posterior semicircular canal is the site of the problem. The patient is quickly lowered to a supine position, with the neck extended. This position vertically aligns the posterior semicircular canal with the direction of gravity. Thus symptoms are reproduced and there is nystagmus if the posterior canal is the site of the otoliths. Alternatively, the roll test can determine whether the horizontal semicircular canal is the site of the problem. This requires the patient to be in a supine position with their head in 30° of cervical flexion. The head is then quickly rotated 90° to the left side, checking for vertigo and nystagmus. This is followed by bringing the head back to the starting position and rotating to the opposite side. Although the patient may experience

vertigo and nystagmus on both sides, rotating towards the affected side triggers more intense symptoms. The Dix-Hallpike test should be routinely performed if possible in the evaluation of all patients with vertigo/dizziness. Whereas a positive test should be considered sufficient for the diagnosis of BPPV in the clinical setting, a negative test should not rule out BPPV completely. Repeated testing in separate occasions may be necessary to avoid missing the diagnosis.

BPPV affecting the lateral semicircular canal may be diagnosed following a history of episodic vertigo related to changes in head position and the presence of horizontal nystagmus provoked by the supine roll test. This test is performed by rotating the patient's head from the neutral position to one side, while the patient is lying supine. After allowing any nystagmus or vertigo to subside the test is repeated to the opposite side. In a positive test horizontal nystagmus is observed, either beating toward the dependent ear (geotropic) or beating away from the dependent ear (apogeotropic) on both sides. For geotropic nystagmus the side associated with the stronger nystagmus is likely the affected ear.

BPPV is referred to as "benign" because because in many cases it can resolve spontaneously. Routine medications such as vestibular suppressants (e.g., antihistamine and benzodiazepine) are generally not recommended. Vitamin D supplementation has been suggested of possible benefit in selected patients, in a few reports. Symptomatic BPPV is often treated with a number of simple manoeuvres such as the Epley manoeuvre, Semont manoeuvre and Brandt-Daroff exercises. These are designed to reposition the otoliths from the posterior semicircular canal into the vestibule, where it does not cause vertigo. A bone vibrator may also be placed on the mastoid bone during the manoeuvres to loosen the debris. Eighty percent of patients are cured by a single repositioning manoeuvre. If the symptoms persist medications may be useful. Lateral canal BPPV can be treated with Lempert roll, forced prolonged positioning, or Gufoni maneuvers. Surgical treatment is rarely indicated. In one procedure, the singular nerve (posterior ampullary nerve, which selectively supplies the posterior semicircular canal) is identified and divided to prevent aberrant signal generated in the canal from reaching the CNS. In another, the posterior semicircular canal is exposed in the mastoid bone, fenestrated and occluded to prevent the canal from generating aberrant signal. However, surgery can result in severe hearing loss, and thus should be reserved as a last resort option for severe and persistent cases.

37.8.2 Epley's Manoeuvre (Modifications Exist)

The Epley manoeuvre uses gravity to help reposition otoliths thereby making them less stimulating.This can be done in the clinic. However this does not address the actual presence of the crystals, only their location. This should only be performed by experienced practitioners and when there are no contraindications. The steps in this procedure are

1. Sit the patient upright, with the legs fully extended and the head rotated 45° towards the affected side.

2. The patient is then quickly tilted backwards into a supine position with the head held approximately in a 30° neck extension (Dix-Hallpike position). The affected ear faces the ground.
3. Observe the patient's eyes for nystagmus for approximately 1–2 min.
4. The patient's head is then turned 90° to the opposite side so that the affected ear faces up, whilst maintaining the 30° neck extension. Remain in this position for approximately 1–2 min.
5. Keeping the head and neck fixed relative to the body, the patient rolls onto their shoulder, rotating the head with the body. The patient should now be looking downwards at a 45° angle.
6. Observe the eyes for nystagmus for approximately 1–2 min.
7. Finally, the patient is slowly brought back up to an upright sitting posture, while maintaining the 45° rotation of the head. The patient holds this position for up to 30 s (Fig. 37.18).

This manoeuvre may be repeated two more times. Following treatment the patient may wear a soft collar during the day to avoid any head positions that may precipitate symptoms. They are advised to be careful bending, lying backwards, moving the head up and down, or tilting it to either side. Contraindications to Epley's manoeuvre include carotid stenosis, cardiovascular disease, diseases involving the neck and rheumatoid arthritis.

37.8.3 Brandt-Daroff Exercises

These may be undertaken at home. It is designed to break up the crystals and help the patient become accustomed to the position which causes vertigo. The exercise is performed 3 times a day for 5–10 days, until symptoms of vertigo have resolved. The following steps are

1. The patient sits upright on the edge of the bed.
2. The patient then rotates their head to the left as far as possible without causing discomfort.
3. They then lay down on the right side 30 s or until the end of any dizzy sensations.
4. The patient then sits back up and returns their head to a central position.
5. They then rotate the head to the right and lay down on the left side, again for 30 s or until the dizziness has stopped.
6. Finally they sit up and return the head to the centre.

37.8.4 Labyrinthitis and Vestibular Neuritis

These terms are sometimes used interchangeably, due to their clinical similarity. However strictly speaking, vestibular neuritis is inflammation of the vestibular nerve and labyrinthitis is inflammation of the labyrinth within the inner ear. In labyrinthitis the vestibular nerve may or may not also be involved. Because the causes

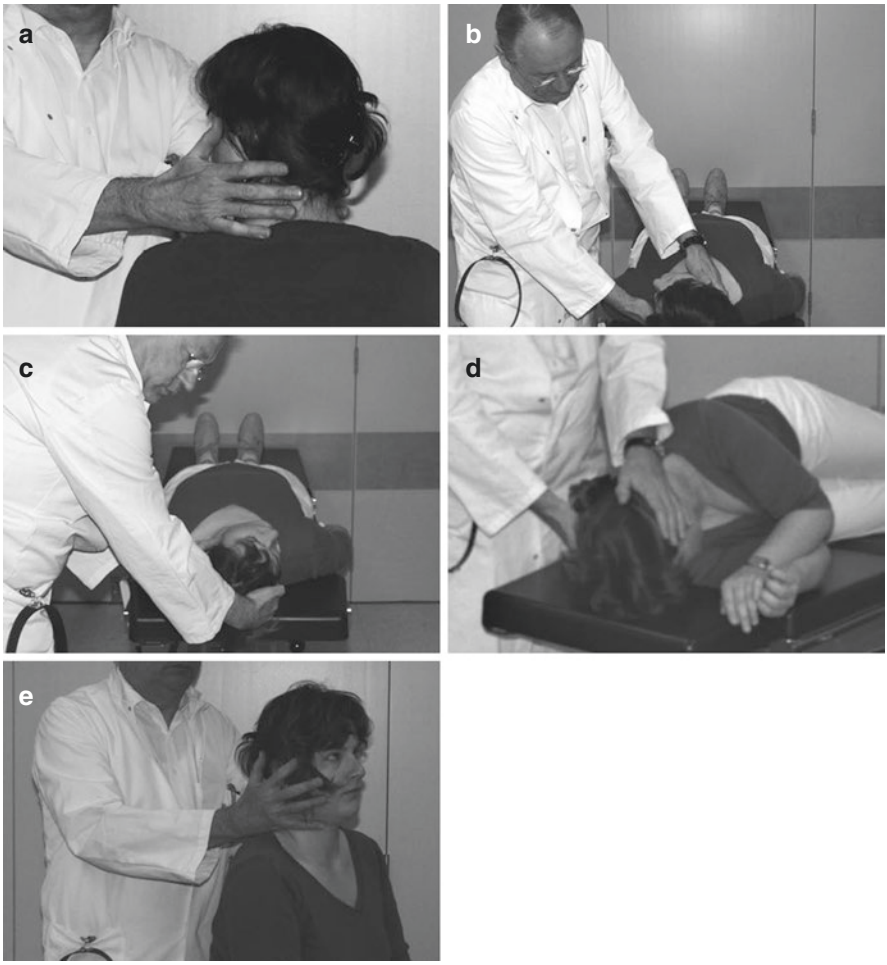

Fig. 37.18 (**a**–**c**) Epleys manoeuvre. (**a**) First step: the patient is sitting in the upright position, the head is 45º turned to the concerned side. (**b**) Second step: the patient is in the lying position, the head still turned to the concerned side. (**c**) Third step: the head is turned 180º to the contralateral side. (**d**) Fourth step: the head and trunk of the patient arc turned 90º to the contralateral side. (**e**) Fifth step: the patient is returned in the upright position. Intervals between each step of 3–5 min

and clinical features of both these conditions are similar, it is often impossible to tell which one is present. Hearing loss is a useful symptom to elicit. If this is present, labyrinthitis is more likely (because the cochlea may also be inflamed). On the other hand, vestibular neuritis is more likely to be associated with nystagmus. In any event, both conditions can result in unilateral vestibular dysfunction, with loss of balance and vertigo as a result of an imbalance in neuronal input between the two sides. In both conditions the underlying cause in the majority of cases is believed to be a viral infection. One third of patients may have had a recent upper respiratory

tract infection prior to developing the disease. Some cases of vestibular neuritis are thought to be caused by an infection of the vestibular ganglion by Herpes Simplex Virus type 1. Both labyrinthitis and vestibular neuritis can also occur following bacterial infection, head injury or with certain drugs.

Both viral and bacterial labyrinthitis may arise primarily in the labyrinth, or occur secondarily from infections in the cerebrospinal fluid, or result in secondary meningitis or meningoencephalitis. Infections may enter the labyrinth via the vestibule nerve, the round or oval window, the CSF, or internal auditory canal. Bacterial endotoxins and the inflammatory response can also irritate the labyrinth without direct infection (referred to as serous/eosinophilic labyrinthitis). Infections can also be blood borne and can complicate stapedectomy. Non-infectious inflammation can occur as a result of vasculitis or immune-related pathology. In both infectious and non-infectious disease, the membranous labyrinths can become severely inflamed which is followed by scarring, atrophy and fibrousosseous obliterative labyrinthitis.

Pyogenic infections of the middle ear and mastoid can cause inner ear dysfunction as a result of involvement of the membranous labyrinth and/or vestibular nerve. This often occurs with meningitis. In the long term, chronic, fibro-osseous obliterative labyrinthitis can result in existent symptoms. Chronic infections of the skull base, Lyme disease and syphilis can also cause inner ear dysfunction from labyrinthitis. All may progress to chronic obliterative labyrinthitis and permanent hearing loss. A number of other but less common inflammatory conditions can also result in labyrinthitis. These include sarcoidosis, Wegener granulomatosis, Langerhans histiocytosis and autoimmune diseases. Trauma and other causes of haemorrhage into the labyrinth comprise non-infectious causes.

Patients generally present with sensorineural hearing loss, tinnitus, vertigo or unsteadiness. When acute, nonspecific symptoms such as malaise, nausea and vomiting may also occur. Nystagmus may be present on examination. Occasionally symptoms, if severe, may initially resemble cerebral infarction. For most patients, the most distressing symptom is severe vertigo. Patients may present with a single attack, repeated attacks, or persistent symptoms that slowly resolve over several days (sometimes weeks). Symptoms can be brought on by pressure changes following flying and scuba diving. Nausea, vomiting, anxiety and a general feeling of being unwell are also common symptoms. Nystagmus suggests vestibular neuronitis. Both vestibular neuronitis and labyrinthitis are generally self-limiting conditions. Recovery time varies, but it is not uncommon for residual symptoms to last for a few months. Medication may not always be necessary, but Prochlorperazine and Cinnarizine may be prescribed to help treat vertigo and nausea. Some authorities suggests that viral labyrinthitis should be treated early with steroids and antiviral medication. Vestibular rehabilitation therapy may also help reduce residual dizziness from labyrinthitis. This works by ‘challenging’ the vestibular system and stimulating adaption. Treatment regimes include various combinations of head and eye movements, postural changes and other exercises (for example, exercises that keep the eyes fixated on a specific target while moving the head). These are believed work by stimulating the brain to use already existing neuronal pathways.

37.8.5 Ménière's Disease (Endolymphatic Hydrops)

Ménière's disease is an idiopathic inner ear disorder which presents with attacks of vertigo, fluctuating hearing loss, tinnitus and aural fullness. It is a chronic condition with a prevalence of 200–500 per 100,000. Patients usually present in the fifth decade of life. Although the precise cause of Ménière's disease is unknown, the pathology is well documented. Excessive accumulation of endolymphatic fluid (hydrops) within the inner ear, results in distension of the endolymphatic compartment. This appears to both mechanically and chemically interfere with the sensory cells involved in balance and hearing. There may also be mixing of the endolymph with the perilymph, which can also lead to temporary dysfunction. It has also been suggested that the endolymphatic sac itself may be defective. The pathophysiology of this disease is poorly understood. Anatomic, infectious, immunologic and allergic factors have all been suggested as the primary cause for these changes. Proposed theories, include intrinsic (genetic, anatomic, metabolic, endocrine, autoimmune, or vascular), and extrinsic (allergic, viral, or traumatic). None of these hypotheses have been fully accepted. Other theories are that the disease is a migraine variant, or a consequence of recrudescent herpes virus infection. Autoimmune mechanisms also appear to be associated in the pathophysiology.

Ménière's disease is characterised by episodic attacks lasting from 20 min to a few hours. During this time there is a triad of (1) vertigo, (2) fluctuating sensorineural hearing loss and (3) increasing tinnitus, typically before or during the vertigo. Patients may also complain of fullness in the ear, gait problems, postural instability, drop attacks and nausea. Usually only one ear is affected, although both may become involved in later episodes. Each episode is usually associated with nausea and vomiting and afterwards, patients feel exhausted for a few days. These symptoms overlap with other conditions such as Migraine-associated vertigo (MAV), transient ischaemic attack (TIA) and stroke.

Ménière's disease is usually a clinical diagnosis. Investigations include pure tone audiometry, electrocochleography and an MRI scan to exclude an acoustic neuroma, tumour of the endolymphatic sac, vascular and other intracranial pathologies. However, there is no diagnostic test specifically for Ménière's disease. Audiology shows a low-frequency sensorineural hearing loss. Electrocochleography and Electronystagmography (ENG) may be needed if the diagnosis isn't obvious, or if ablative therapy is being considered. These are highly specialised tests.

Management is usually medical. Betahistadine and diuretics are usually effective during acute episodes of vertigo. Acute attacks can be treated with either vestibular suppressants (e.g., meclizine and diazepam) or antiemetics (e.g., prochlorperazine). In some patients a short course of oral steroids may help. These may reduce any inflammation or immunologic factors involved in the pathogenesis and consequently the severity of spells. Intratympanic steroids have also been used to treat acute episodes in an attempt to avoid the systemic complications of oral steroids. If these measures do not work intratympanic gentamicin may be given. Dietary modifications (sodium-restricted diet, dietary restrictions on caffeine, nicotine, alcohol and foods containing theophylline e.g., chocolate) may also help. Sodium

restriction is widely used to control symptoms. Reduction of dietary sodium combined with diuretics has been reported to slow progression of hearing loss. This is thought to lower pressure in the hydropic ear, reducing the risk of membrane rupture. A low-sodium diet is also useful in controlling hypertension, a vascular risk factor. Patients who have failed medical and gentamicin treatment may require surgical intervention. Endolymphatic sac surgery and vestibular nerve sections preserve hearing while labyrinthectomy ablates hearing.

37.8.6 Recurrent Acute Vertigo

Recurrent episodes of acute vertigo may be due to benign paroxysmal positional vertigo (BPPV), Meniere's disease (MD) or vestibular migraine. Patients with vertebrobasilar transient ischaemic attacks predominantly manifesting with vertigo are said to be at risk of stroke, so should be assessed urgently.

37.8.7 Vestibular Migraine

Patients with migraine headaches can also develop balance problems, including vertigo. Children commonly have vestibular migraine but rarely develop BPPV. Between attacks patients fully recover and neurological and vestibular examination is normal. However during an attack, there might be nystagmus and headache. Treatments include betablockers, pizotifen, tricyclics, anticonvulsants, cinnarazine, flunarazine, and triptans.

37.8.8 Cerebellopontine Angle Syndrome

This is a distinct syndrome caused by a space occupying lesion at the junction of the cerebellum and pons. Due to its location this can affect multiple cranial nerves and the cerebellum. It can present with unilateral sensorineural hearing loss and unilateral tinnitus, vertigo and ataxia, speech impediments, ipsilateral tremors and loss of motor control. By far the most common cause is an acoustic neuroma. If untreated, extension into the brainstem can result in hydrocephalus, ipsilateral facial and corneal numbness (CN V) and other cranial nerve deficits (CN VII). Bruns nystagmus ('dancing eyes') is an important localising sign, indicative of compression of the flocculi. This is a coarse, horizontal nystagmus with low oscillatory frequency when the patient looks towards the side of the lesion, but a fine, high-frequency nystagmus that increases as the patient looks to the side opposite the lesion.

In most cases, the cause of cerebellopontine angle syndrome is an acoustic neuroma. This may be associated with neurofibromatosis type 2 (NF2). Other reported causes include (1) meningioma, (2) cerebellar astrocytoma (a malignant tumour), (3) intracranial epidermoid cyst (4) glomus jugulare tumour associated with the glossopharyngeal nerve, (5) cholesteatoma, (6) cerebral abscess and (7)

IgG4-related hypertrophic pachymeningitis. Rare causes include chordoma, teratoma and lipoma. Diagnosis is usually made following CT or MRI. Management depends on the cause and the severity of symptoms but is usually surgical or radiotherapy. With benign disease and minimal symptoms, observation and serial imaging may be more appropriate.

37.9 Superior Semicircular Canal Dehiscence

Superior semicircular canal dehiscence (SSCD) is a so called "third window" lesion of the inner ear, in which a small region of the bone overlying the superior semicircular canal is dehiscent (missing), or extremely thin. Normally the bone in the semicircular canals is thin at birth, but it progressively thickens until around 3 years of age. However in approximately 2% of adults the bone may dehisce or remain extremely thin (<0.1 mm), notably in the superior semicircular canal. Any subsequent head trauma or barotrauma which results in sudden changes in intracranial pressure can result in a "two-hit" phenomenon, in which the "second hit" disrupts the weakened bone. In the presence of this bony defect the fluid in the membranous part of the canal can then become disrupted by sound and pressure changes. Normally there are only two discrete regions of compliance within the inner ear—(1) the oval window (through which sound energy is transmitted into the cochlea via the stapes bone) and (2) the round window (which enables the fluid in the cochlea to move in response to this sound energy). SSCD thus creates a "third window"—hence the expression. Symptoms arise as a result of abnormal movement within the endolymph or perilymph following a sudden change in intracranial pressure (for example during coughing, sneezing or straining). These include vertigo, autophony (increased perceived resonance of one's own voice), tinnitus and hearing loss, Some patients may perceive objects to be moving in time with their pulse (pulsatile oscillopsia). Loud sounds can also stimulate motion of perilymph and endolymph between the vestibule and dehiscent superior semicircular canal, resulting in vertigo (Tullio phenomenon).

SSCD may be confused with Meniere's disease, otosclerosis, and perilymphatic fistula. SSCD has also been reported in children and in postpartum females (secondary to the presumed increased intracranial pressure during childbirth). Diagnosis is confirmed with high-resolution CT of the temporal bones. This can often visualise the bony defect in the superior canal, although it can miss a very thin layer of intact bone. Vestibular evoked myogenic potential (VEMP) play an important role in assessment. In SSCD, loud tones evoke a short-latency relaxation potential in the ipsilateral sternocleidomastoid muscle. The amplitude of the VEMP waveform in an SSCD ear is therefore greater than normal. Most patients with SSCD are able to tolerate their symptoms and reduce these by avoiding situations that make them worse. For those that cannot, surgical repair of the dehiscence may help. The canal is plugged with fascia and small bone chips. In patients with bilateral SSCD, surgery on the more severely affected ear may be sufficient to control symptoms.

37.10 Otosclerosis

Otosclerosis is a disease that is unique to the otic capsule of humans.

This term is derived from the Greek for "hardening of the ear." This is caused by abnormal bone deposition within the ear, in which the normally dense layer of endochondral bone is slowly replaced by irregular patches of spongy, vascular bone. Its name is therefore something of a misnomer. Otosclerosis primarily affects the stapes bone. The otic capsule is normally inert with minimal bone turnover, but in otosclerosis there is an increase in bone turnover. In the oval window, this may spread across the annular ligament to immobilise the stapes. Further extension over the inner ear is then accompanied by damage to the membranous labyrinth, which can result in sensorineural hearing loss, tinnitus, and vertigo. This process shares histological similarities to Pagets disease, described elsewhere in this book. The cause of this is unknown although a genetic component has been associated. Several infectious agents have also been implicated, notably the measles virus, but their role is still unknown. Certain drugs may also cause otosclerosis as a side effect. Paget's disease has also been associated.

Otosclerosis usually begins in one ear but bilateral disease eventually develops in most cases. Patients are usually young (under the age of 30) and typically present with mixed hearing loss. However the conductive component is most often predominant. If progressive, the cochlear nerves can become affected. Patients may also experience pulsatile tinnitus as a result of the hypervascularity that occurs develop during an intense osteolytic phase of the disease. The external auditory meatus and tympanic membrane often appear normal on examination but discolouration of the promontory (seen as a reddish retrotympanic mass) is said to be pathognomonic for otosclerosis (Schwartze sign). CT can often visualise deeper calcification. Audiometry is usually required to assess the degree of hearing loss. Depending on the stage of the disease, this may show a mild to moderate conductive hearing loss. Once complete stapes fixation has occurred, there will then often be pseudosensorineural pattern of hearing loss. With progression of conductive loss, the Rinne test demonstrates greater bone conduction than air conduction, and Weber's test lateralises to the affected side. The stapedial reflex may be normal in the early stages but later cannot be elicited as fixation of the bone occurs.

Management of otosclerosis is controversial. Medical treatment is mostly directed at maturing the involved bone and decreasing osteoclastic activity. Sodium fluoride and bisphosphonates (which inhibit osteoclastic activity) and cytokine antagonists may offer hope for the future. Hearing aids are usually very effective in the early stages of the disease. Surgical management includes stapedectomy. The aim is restoration of the sound transmission mechanism from the tympanic membrane to the oval window, by bypassing the resistance of the fixed stapes footplate. Today, a variety of techniques are used to correct stapes fixation.

37.11 Cholesteatoma

A cholesteatoma is a potentially serious condition. This is an erosive, cyst-like structure, composed of desquamated keratinised squamous epithelium, which forms a mass within the middle ear or the mastoid. As it expands it can become locally destructive and invasive due to a combination of pressure necrosis and enzymatic activity. Although cholesteatoma is not pathologically malignant, it can still cause significant problems because of these properties. This can result in the displacement or destruction of the ossicles and erosion through the walls of the middle-ear cavity. Further extension into and through the base of the skull, then intracranially can also occur. Cholesteatoma are often secondarily infected and this can result in chronically discharging ears (Figs. 37.19 and 37.20).

Cholesteatoma are classified as either congenital (2%) or acquired (98%). The latter is most often secondary to chronic otitis media. However it may also occur after tympanic membrane trauma, in which implantation of squamous cells into the adjacent tissues occurs. Cholesteatoma can affect all age groups, from infants through to the elderly, but it is commonly seen in young adults. The hallmark of the disease is

1. Chronic offensive discharge, unresponsive to antibiotics and
2. Conductive hearing loss (from ossicular erosion)

Other symptoms depend on the extent of local invasion. These include vertigo, headache and facial nerve palsy. Otoscopy may show debris leaking through a perforation of the tympanic membrane, or crusting in the attic. It is important to always consider cholesteatoma in a patient with aural discharge and hearing loss. Left untreated, cholesteatoma may erode the tegmen, the sigmoid sinus, or the inner ear, resulting in labyrinthine fistula. Erosion into the lateral semi-circular canal is one of the most common complications of cholesteatoma. It can result in severe sensorineural hearing loss and vertigo. When the overlying bone is lost, pressure can be transmitted directly from the external auditory canal to the endolymph, resulting in vestibular symptoms. A fistula test will be positive in about 70% of patients with lateral semicircular canal erosion. Lateral sinus thrombosis, sepsis, brain abscess, facial paralysis and even death have also been reported. CT and occasionally MRI are required to assess the extent of invasion. Treatment is surgical excision. Following removal of the cholesteatoma, fistulas can be managed with soft tissue grafting.

37.12 Eustachian Tube Problems

37.12.1 Eustachian Tube Dysfunction (ETD)

The eustachian tube has important several functions associated with the middle ear. It enables ventilation to control pressure in the middle ear, drains secretions into the nasopharynx by muscular peristaltic actions and mucociliary clearance, and protects the middle ear from nasopharyngeal sound pressure and secretions. Opening of the tube, is facilitated swallowing or yawning. Pressure in the middle ear is

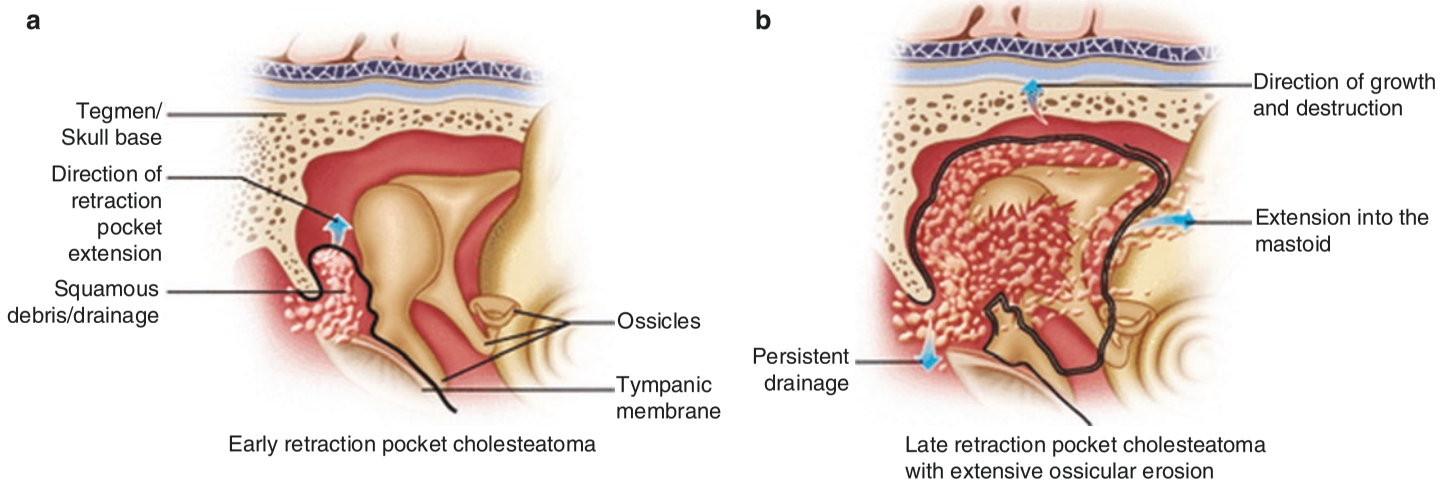

Fig. 37.19 Illustration of the formation of a retraction pocket cholesteatoma, caused by negative pressure in the middle ear space due to Eustachian tube dysfunction. (**a**) In the early phases, the retraction pocket begins to form as the pars flaccida invaginates and wraps around the ossicles. (**b**) As more squamous cells are shed from the surface of the tympanic membrane, they are trapped in the retraction pocket and the cholesteatoma grows, putting pressure on the ossicles and surrounding temporal bone leading to erosion

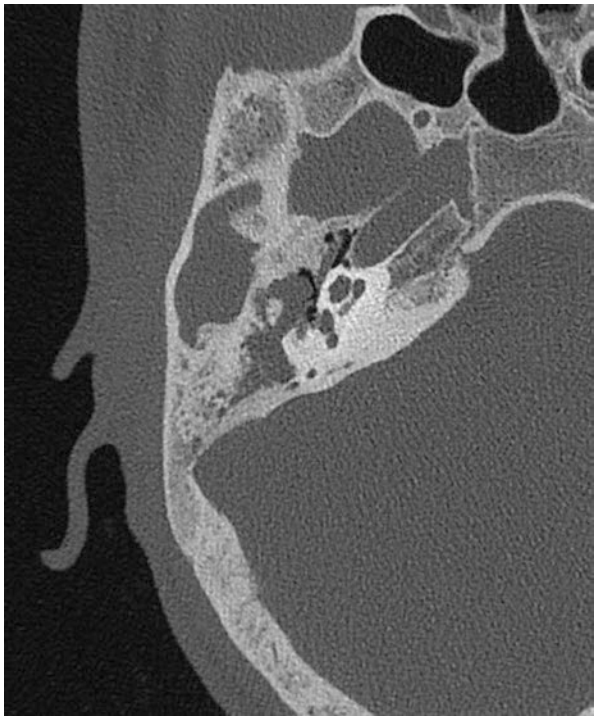

Fig. 37.20 Axial computed tomography scan of the right temporal bone demonstrates a soft-tissue mass of the right middle ear and mastoid cavities that has caused bone and ossicular erosion suggestive of a cholesteatoma

maintained through two mechanisms: middle ear mucosal gas exchange and opening of the Eustachian tube to equilibrate pressure with that in the nasopharynx. In the healthy middle ear, pressure slowly decreases, but periodic opening of the Eustachian tube restores atmospheric pressure. Any disruption in this function can therefore affect homeostasis of the middle ear. The eustachian tube also protects the middle ear against inflammation and infection by viruses, bacteria and gastro-oesophageal reflux. Dysfunction has therefore been associated with most cases of middle-ear disease, including acute or chronic suppurative otitis media, otitis media with effusion, and middle-ear atelectasis. Assessing eustachian tube function is therefore an important steps in evaluating the pathogenesis and prognosis of patients with chronic suppurative otitis media.

Eustachian tube dysfunction is now regarded not as a single entity but rather a syndrome with a constellation of signs and symptoms suggestive of dysfunction of the Eustachian tube. Acute dysfunction may have transient symptoms and signs for less than 3 months, whilst chronic dysfunction, symptoms and signs persist for more than 3 months. Three subtypes of Eustachian tube dysfunction have been described

1. Dilatory Eustachian tube dysfunction
2. Baro-induced Eustachian tube dysfunction and
3. Patulous Eustachian tube dysfunction

Any cause of tube blockage, tube inflammation or failure of the tube to open sufficiently can cause dysfunction. Common causes include upper respiratory tract

infection (URTI), glue ear, allergies or enlarged adenoids. More rarely, in older patients, tumours in the nasopharyngeal region can result in otic symptoms. With severe dysfunction the resulting pressure differential between the middle ear and surrounding environment distorts the tympanic membrane and places it under tension. The membrane becomes less complaint and fails to vibrate fully in response to sound waves. This results in muffled or dull hearing.

Patients present with symptoms of pressure disequilibrium, 'aural fullness', 'popping' and discomfort. They may also report pressure, an 'under water' sensation, crackling, ringing, autophony and muffled hearing. In many patients symptoms are short-lived and no cause is identified. Acute dilatory tube dysfunction is often preceded by an upper respiratory tract infection, or exacerbation of allergic rhinitis, which causes inflammation within the tube. Late onset of sudden deafness should always be carefully assessed with this in mind. Temporary dysfunction is also commonly experienced by many people during take off and landing during flights (baro-induced Eustachian tube dysfunction), but in severe cases barotitis can occur. The tympanic membrane may then be seen to have dilated vessels and bruising. Symptoms can last from a few hours to several weeks depending on the cause.

The ability to auto-inflate the middle ear on Valsalva or Toynbee manoeuvre confirms some degree of patency of the Eustachian tube. Several simple tests can evaluate eustachian tube function. In individuals with an intact tympanic membrane, the eustachian tube swallow test assesses changes in applied pressure in the middle ear while swallowing. Tympanometry is performed repeatedly while the examiner adds positive and/or negative pressure to the external auditory canal and asks the patient to swallow. Another test is to perform the Valsalva maneuver, which increases nasopharyngeal pressure and evaluates whether the eustachian tube opens or not. Mild to moderate conductive hearing loss may also be found in some patients with Eustachian tube dysfunction. Dynamic slow motion video endoscopy is another useful technique for evaluating the pathophysiology of eustachian tube dysfunction. Management depends on the cause. In many cases nasal decongestants can help if allergy or congestion is suspected. In selected cases wires, balloon dilation and stents may be indicated. Antibiotics and sometimes adenoidectomy may be indicated for recurrent middle ear infections secondary to dysfunction.

37.12.2 Patulous Eustachian Tube (PET)

This is a benign condition in which the eustachian tube fails to close normally, instead remaining patent most of the time. It can result in a multitude of symptoms including voice autophony, breathing autophony and aural fullness. Possible aetiologies include atrophy of the peritubal fat, loss of venous tone of the pterygoid venous plexus, and peritubal musculature dysfunction. PET has also been associated with weight loss and pregnancy. In contrast to dysfunction, middle ear problems generally do not occur, since pressures between the middle ear and environment are readily balanced. Symptoms may be better in the supine position or during upper respiratory tract infection and worsen during exercise. However it has been suggested that otitis media may predispose to the condition. Chronic upper respiratory inflammatory diseases are commonly associated with PET.

PET can also be associated with pregnancy, weight loss, mucosal scarring secondary to surgery, radiation and neuromuscular disorders causing muscle atrophy (such as MS or following a stroke). It can easily be misdiagnosed and treated as congestion, although decongestants and steroids spray will be ineffective. Patients commonly complain of autophony (hearing one's own voice and breathing—a "bucket on the head" effect), or alternatively muffled sounds. This is due to variations in pressure that occur during speech or breathing, being transmitted to the middle ear. In severe cases patients usually hear not only their own voice but also their heartbeat, footsteps and chewing. Diagnosis is based on clinical assessment, nasoendoscopy, audiology and tympanogram studies. Mild cases may only require reassurance. Both surgical and non-surgical interventions have been described in the literature, most of which aim to narrow or close the ET orifice. Non-surgical methods include weight gain, topical oestrogen, and insufflation with boric or salicylic acid. Potassium iodide treatment may have a role in thickening secretions in more moderate cases. Surgical management is reserved for severe cases in which there is failure of medical management. Several different techniques are described including injection of bulking agents, fat/cartilage plugging, ligation of the orifice, endoluminal cauterization and hamulotomy. Success rates are generally variable, with current no consensus on management.

37.12.3 Palatal Myoclonus

Palatal myoclonus is a rare cause of pulsatile tinnitus secondary to rhythmic involuntary contraction of the palatal muscles. Two variants are described in the literature, (1) a "symptomatic form" of palatal myoclonus, associated with brainstem or cerebellar lesions in the triangle of Guillain-Mollaret and (2) an "essential form" that occurs in isolation and has no known intracranial cause. This has been described as either idiopathic, psychogenic, voluntary, central, or peripheral in origin but all without any focal neurological lesions on imaging. When associated with eye movements, it is known as oculopalatal myoclonus. Patients present with a sensation of 'clicking' or 'popping' in the ears, or tinnitus. Investigations involve audiometric studies and magnetic resonance imaging of the brain. Treatment with anxiolytics, anticonvulsants, and surgery have been largely unsuccessful. Noise masking has resulted in modest symptomatic relief. More recently botulinum toxin injection into the palatal muscles has been recommended. Side effects include hypernasality, velopharyngeal insufficiency (VPI) with nasopharyngeal regurgitation and dysphagia.

37.13 Tumours and Tumour-Like Conditions of the Ear

Apart from cutaneous malignancies involving the pinna, these are generally uncommon. Cancers arising within the external auditory canal itself are unusual and those within the middle ear are rare. These latter tumours include adenocarcinomas,

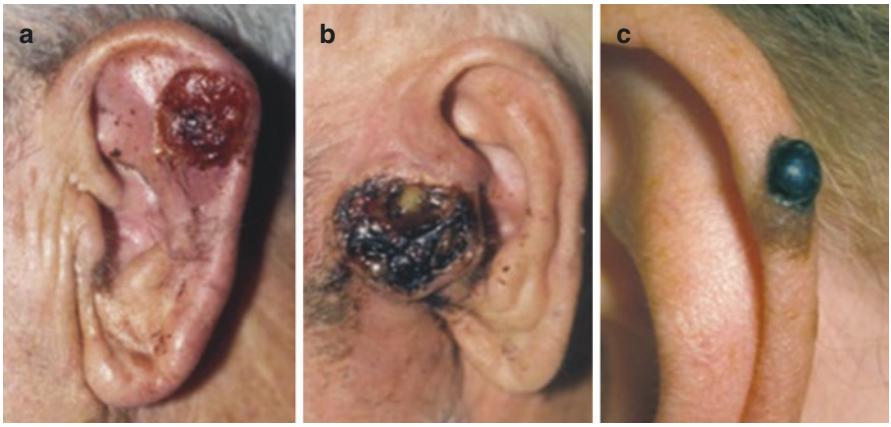

Fig. 37.21 (**a**–**c**) Malignant lesions of the auricle: (**a**) basal cell carcinoma, (**b**) sequences carcinoma, (**c**) malignant melanoma

transitional cell and carcinoid tumours. Rhabdomyosarcoma may also arise in the middle ear in children. The most common malignancies involving the temporal bone are those that arise from the skin of the pinna and surrounding area and which invade into the bone. Ewing sarcoma, osteosarcoma and other mesenchymal-origin cancers may also arise in temporal bone. These are all usually well advanced tumours at the time of presentation and therefore carry a poor prognosis. Some malignancies may be associated with chronic infection and on imaging temporal bone tumours can be confused with chronic inflammation. Haematopoietic, lymphoid and metastases (breast, lung, renal and prostate) to the temporal bone are all rare. Paragangliomas are the most common tumour seen in the temporal bone. These tumours are only occasionally malignant (Fig. 37.21).

37.13.1 Glomus Jugulare Tumours

Paragangliomas are unusual lesions accounting for 0.012% of all tumours, and 0.5% of all head and neck tumours. They are slow growing, vascular tumours with a long natural history. Carotid body tumours account for 60% of head and neck paragangliomas and the rest include glomus jugulare, vagale and tympanicum. Glomus jugulare tumours (also called jugulotympanic paragangliomas) are benign but locally aggressive vascular paragangliomas. They can arise either from the promontory of the middle ear, or the jugular bulb. As these tumours grow, they occupy the middle ear, resulting in pulsatile tinnitus, with or without conductive hearing loss. As they enlarge further they start to erode the surrounding bone, especially inferiorly, to cause lower cranial nerve palsies and involve the major vascular structure in the skull base. With further expansion cerebellopontine angle syndrome may occur. Tumours may also impinge on the ossicles and the tympanic membrane, impairing the motility of both. Patients can therefore present with increasing

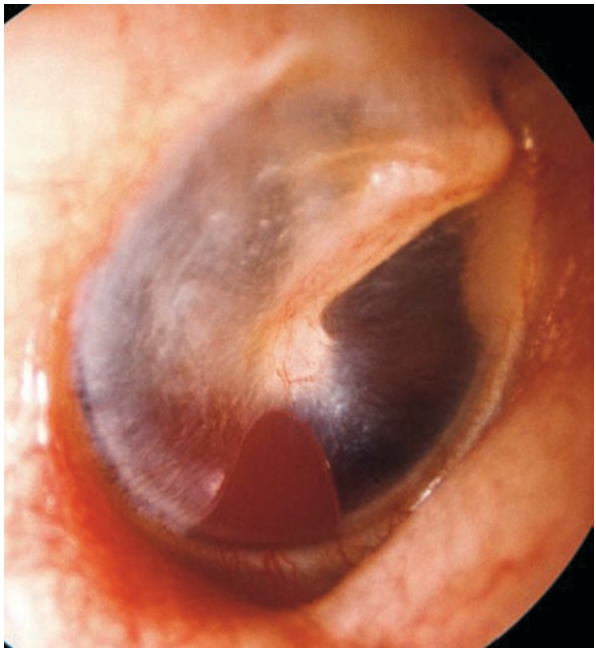

Fig. 37.22 The pulsating red mass behind the inferior quadrants of this right tympanic membrane is a glomus tumour (chemodectoma) limited to the middle ear (class A tumour)

deafness or lower cranial nerve palsies. Involvement of the lower cranial nerves (IX, X, XI, XII) and the internal carotid artery (ICA) and vertebral artery (VA) determine the prognosis and outcome in long term. Brainstem compression and poor contralateral venous circulation are also major risk factors.

Patients are usually middle aged and present with tinnitus, hearing loss, lower cranial nerve palsies, otalgia, auricular discharge and bleeding. On examination there is often pulsatile lesion in the upper neck, hearing loss, cranial nerve deficits and a bruit over the mastoid. A bluish pulsatile or a fleshy granular mass may also be visible on otoscopic examination. Raised blood pressure and postural hypotension can occur with functional tumours. If suspected, urinary catecholamines should be tested for. Diagnosis is confirmed by CT and MRI. High resolution CT is useful for small early lesions. Contrast enhanced CT shows moderate to markedly enhancing tumours in irregular lytic lesions. Treatment involves surgical removal, or stereotactic guided radiation. Malignant transformation has been reported in up to 20% of cases (Fig. 37.22).

37.13.2 Acoustic Schwannoma

A schwannoma (also known as an neurilemoma, neuroma, neurolemoma, and Schwann cell tumour) is a benign nerve sheath tumour which is derived from Schwann cells. These cells normally produce the myelin sheath which surrounds and insulates peripheral nerves. Schwannomas can therefore arise almost anywhere

in the body. In some cases they are associated with neurofibromatosis type II. Schwannomas in the head and neck are relatively common and have been reported to be found incidentally in 3–4% of autopsies. Acoustic schwannoma (involving the vestibulocochlear nerve) is the most common lesion found inside the internal auditory canal. It is usually located in the cerebellopontine angle, but in most cases, will expand into the internal acoustic foramen. It can therefore result in cerebellopontine angle syndrome, or isolated SNHL, vertigo, and tinnitus. These tumours are relatively slow-growing and mostly benign. Less than 1% become malignant (neurofibrosarcoma). The tumour is always encapsulated and on the outer surface of the nerve. Surgical removal is therefore often possible. The differential diagnosis includes vestibular neuritis.

If symptoms are only mild and surgery is not indicated, follow-up imaging should be arranged every 6 months and then annually if the schwannoma appears to be stable. If removal is indicated, hearing preserving microsurgery or stereotactic radiosurgery may be possible. This has been the standard treatment for vestibular schwannoma for many years. Surgery can be difficult. Many important structures (such as the trigeminal, facial and acoustic nerves, arteries and the brainstem) are located around the tumour. Stereotactic radiosurgery is becoming increasingly popular. It is safe and is reported to have a high cure rate.

37.13.3 Tumours of the External Ear

Chronic sun exposure can result in the usual skin cancers seen in the head and neck (discussed in the chapter on the skin). Immunocompromised patients have an increased incidence of these skin cancers, which may be more aggressive than in the general population. Basal cell carcinomas are the most common neoplasms of the auricle (about 45% of pinna tumours), but Squamous cell carcinoma and Melanoma can also occur. All suspicious skin lesions should be biopsied. Treatment options include excision, curettage, cryotherapy, topical 5-Fluorouracil and occasionally radiotherapy. Surgical excision is currently the preferred option in most patients.

Rare tumours of the EAM include glandular tumours (adenoid cystic carcinomas, ceruminous adenomas, ceruminous adenocarcinomas and pleomorphic adenomas). These usually present as a painless skin lesion in the canal. If invasive, they may present with otorrhea, aural fullness, otalgia—symptoms and signs that may mimic inflammatory diseases. Growth is initially contained by the bony/cartilaginous walls and the tympanic membrane and at first the tumour grows within the canal, attached to the underlying perichondrium or periosteum. It then later invades the cartilage and bone, passing through the tympanic membrane and into the middle ear and mastoid. This may involve the facial nerve. Subsequent the tumour may eventually spread intracranially through the roof of the middle ear and mastoid. Hearing loss is an important finding. Conductive hearing loss may occur when the EAC is occluded or the tympanic membrane or middle ear becomes involved. Sensorineural hearing loss signifies deep extension into the inner ear. Tumours may also spread beneath the skin surface, presenting as a preauricular lump or swelling.

In advanced disease trismus may result from invasion of the temporomandibular joint.

37.13.4 Keloids

Keloids are benign overgrowth of fibrous tissue, which occur during healing. Their cause is unknown, although trauma (including surgery) is a well known precipitant. Keloids can occurs in all races, but are much more likely to occur in patients with darker skin, notably Africans and Indians. For some unknown reason the earlobe is particularly predisposed to keloid formation. Ear piercing is the commonest cause of the earlobe keloid. This develops as a hard rubbery lump, which may be itchy, tender or red. Some keloids can reach an impressive size. Simple excision is one treatment, but recurrence rates are high. Intralesional corticosteroid injections may be used alone or combined with surgery. Injection is most effective in the early stages, on younger hypertrophic scars. Triamcinolone acetonide can also be applied topically for a long time. Radiation was previously reserved for keloids resistant to other treatments, but is now rarely undertaken because of the morbidity and risk of carcinogenicity (Fig. 37.23).

37.13.5 Other Tumours of the Middle/Inner Ear

In adults most tumours of the middle ear are benign. These include tympanic paragangliomas, adenomas, meningiomas and schwannomas. Middle ear cancer may also be seen in patients with long-standing middle ear inflammation. Malignant invasion can also occur as nearby tumours can extend into the middle ear. These include carcinomas (arising in the external auditory meatus, parotid gland, minor salivary glands and pharynx), lymphomas and metastasis. Pain, discharge, and facial nerve palsy are common presenting features.

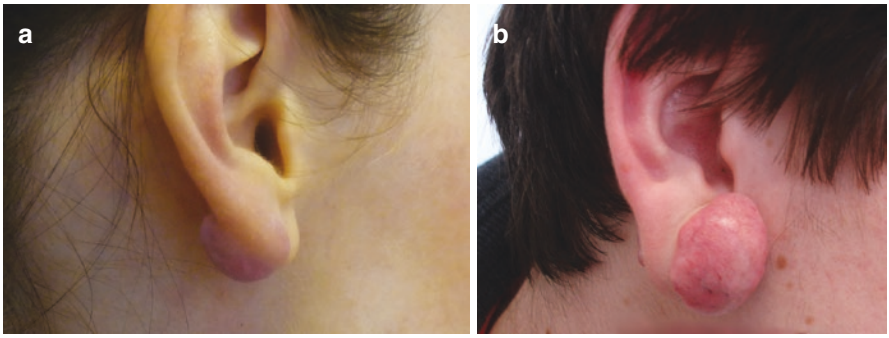

Fig. 37.23 (**a**, **b**) Keloid—the ear lobe is a common site

Carcinoma of the eustachian tube is rare. Patients usually present with pain and otitis with conductive hearing loss. Advanced lesions may develop cranial nerve deficits involving CNVII to XII and Horner syndrome. On examination the nasopharynx will appear normal and imaging is essential to make the diagnosis. Lymphatic spread in tumours arising in the tympanic cavity and mastoid is unusual. Perineural spread along the facial nerve and auriculotemporal branch of the mandibular nerve can occur with all these cancers in this region.

Rhabdomyosarcoma is a malignant tumour of striated muscle origin. It is believed to originate in the middle ear and it will usually have already destroyed bone at the time of presentation. Tumours are more common in children, usually under 12. They usually present with unilateral serous otitis media with or without a visible mass and conductive hearing loss. Pain, discharge, and facial nerve palsy may also occur. Tumours soon extend to the carotid canal, jugular fossa, into the inner ear, and along cranial nerves toward the brain stem. Meningeal involvement and dissemination within the subarachnoid space are also possible. Needless to say, these are poor prognostic findings. Chemotherapy and radiotherapy play a major role in the treatment. Debulking is occasionally performed.

Histiocytosis is an aggressive disease in which histiocytes result in destruction of the mastoid, with or without inner ear extension. Papillary tumours of the endolymphatic sac can occur, particularly in patients with Von Hippel Lindau disease. Around 10% of these patients will develop such a tumour. In some cases this will be bilateral.

37.14 Benign Lesions of Bone

37.14.1 Exostoses and Osteomas

These are bony growths which typically arise in the bony portion of the EAC. They are usually asymptomatic but occasionally may cause wax impaction, otitis externa, or rarely, conductive hearing loss. Osteomas are often pedunculated. Exostoses are often multiple and sessile. They commonly occur in people who frequently spend time in cold water ("surfers ear"). These appear to be formed by reactive bone formation. Most osteomas and exostoses require no intervention. If surgery is necessary, a transcanal or postauricular approach can be used, depending on the size of the lesions.

37.14.2 Fibrous Dysplasia

The temporal bone is sometimes the site of monostotic or, less commonly, polyostotic fibrous dysplasia. The most common presentation is progressive conductive hearing loss secondary to occlusion of the EAC, or impingement on the ossicles. Progressive narrowing of the ear canal can also lead to the development of a cholesteatoma. Diagnosis is based on radiographic and histologic findings. On plain

radiographs, a fibrous dysplastic lesion is based on a "ground-glass" appearance. Surgery is indicated for biopsy, presence of cholesteatoma, hearing loss and correction of any cosmetic deformity.

37.15 Facial Palsy and the Ear

Although the causes of facial paralysis are many and extend beyond the ear, a short synopsis is given hear as several important causes (temporal bone fracture, otitis media, schwannoma and Bells palsy) are related to this anatomical site. Bell's palsy is the commonest cause of spontaneous paralysis. Proper function of the muscles is vital for the protection of the eye, maintenance of the nasal airway, oral continence, and for clear speech.

37.15.1 Nerve Injury

Nerves are made of three main components, from innermost to outermost (1) endoneurium, (2) perineurium and (3) epineurium. The endoneurium is the innermost component that is adherent to the Schwann cell and crucial for regeneration of the nerve. Transection or injury here has a poor prognosis for recovery and often results in aberrant regeneration or no regeneration at all. The perineurium is responsible for providing the tensile strength of the nerve and provides a barrier of resistance to infection. The epineurium or nerve sheath is the outermost layer and is responsible for nourishing the nerve via the vasa nervorum. Nerve injury can be classified into five main categories based on the histological features of injuries.

First degree (Neurapraxia). This follows nerve compression or ischaemia. Nerve continuity is preserved and there is no Wallerian degeneration. Following a short period of dysfunction rapid and complete recovery usually occurs. Clinically there may be partial loss of function (paresis), without muscle wasting or muscular degeneration.

Second degree (Axonotmesis). Here, the axon is severed but the endoneurium of the Schwann cell remains intact. Degeneration of the nerve distal to the injury occurs. There is loss of motor, sensory and sympathetic function at injured site, with muscle denervation distal to the injury, fibrillation and atrophy. Recovery time is dependent on the severity of the injury and usually takes months. However there is usually complete functional recovery.

Third degree (Neurotmesis). Here there is intrafascicular injury with disruption of endoneurium. Wallerian degeneration occurs and axonal regeneration is slow and delayed. Recovery is often incomplete or accompanied by aberrant regeneration. Motor and sensory loss is severe and despite eventual re-innervation, muscle recovery may be incomplete at best.

Fourth degree (Neurotmesis). With this injury the nerve trunk is undisturbed, but the site of injury contains ruptured fasciculi, damaged Schwann cells and regenerating axons. Wallerian degeneration occurs and neuroma formation is common. There

is complete loss of motor, sensory and sympathetic function, with only minimal spontaneous recovery.

Fifth degree (Neurotmesis). This is the worse injury. There is loss of nerve continuity and severe disruption of all structures in the area. Only a few viable neurones survive, the rest are severely damaged including retrograde degeneration. Axons may regenerate but usually fail to re-innervate the correct muscle fasciculi.

37.15.2 Congenital Causes

Congenital facial paralysis is present at birth. This is the most common form of facial paralysis seen in children. It has been estimated that paralysis occurs in 2.0% of live births. It may be isolated with involvement of the facial nerve and its musculature only, or it may be part of a syndrome. In the majority of patients, it is believed to be the result of pressure on the developing foetus from the sacral prominence. During development the facial nerve is superficial and thus easily compressed.

37.15.2.1 Möbius' Syndrome (Congenital Facial Diplegia)

This is a rare congenital disorder, which usually includes bilateral seventh nerve paralysis with unilateral or bilateral sixth nerve paralysis. The cause of Möbius' syndrome is unclear but there appears to be an autosomal dominant inheritance pattern.

37.15.2.2 Hemifacial Microsomia

This syndrome includes unilateral microtia, macrostomia, and mandibular hypoplasia. Goldenhar's syndrome (oculoauriculovertebral dysplasia) is a variant of this. Approximately 25% of patients with hemifacial microsomia have facial nerve weakness.

37.15.2.3 Osteopetrosis

Osteopetrosis is generalised dysplasia of bone, which may have an autosomal dominant or recessive inheritance pattern. In the head and neck, optic atrophy, facial paralysis, sensorineural hearing loss and mental retardation are common. There is also progressive enlargement of the cranium and mandible.

37.15.3 Acquired Causes

37.15.3.1 Trauma

Approximately 90% of all congenital peripheral facial nerve paralysis is attributed to difficult deliveries, high forceps delivery or intrauterine trauma. These are often unilateral and partial. Longitudinal fractures of the temporal bone are the most common type of fracture associated with facial nerve injury. Transverse fractures of the temporal bone are more likely to cause facial nerve injury. Injury to the intratemporal portion of the facial nerve can result in total facial paralysis, as well as dysgeusia. Damage

to the geniculate and proximal to this region can result in decreased lacrimation following injury to the preganglionic parasympathetic fibres that supply the lacrimal gland (carried in the greater superficial petrosal nerve). Damage to the nerve proximal to the take-off of the nerve to the stapedius can result in hyperacusis in the affected ear. Penetrating injuries side of the head and otologic surgery can also result in nerve injury. The House–Brackmann 6-point scale of nerve function is the most commonly used standardised tool for assessing the degree of facial weakness. Electroneurography (ENog) and electromyography (EMG) are usually required to measure facial nerve activity. ENoG is an objective quantitative measurement of nerve function. This measures the summation nerve potential on the normal side of the face on stimulation and compares this to the paralysed side. It is thought to indicate the number of remaining functional nerve fibres. This data is predictive for spontaneous recovery. EMG can be used to determine if a nerve in question is in fact in continuity, shows evidence of Wallerian degeneration, or has signs of re-innervation. Treatment of the injuries is complex and in the intratemporal facial nerve, controversial. Reanimation of the paralysed face is an interesting and evolving field.

37.15.3.2 Cerebrovascular Accidents

Lacunar infarcts may affect fibres in the internal capsule. The facial nucleus itself can be affected by infarction of the pontine arteries.

37.15.3.3 Infection

Facial paralysis can follow otitis media. Infection with the spirochete Borrelia burgdorferi (Lyme disease) can also result in facial paralysis. Herpes zoster oticus (Ramsay Hunt syndrome) is the cause of up to 10% of facial paralysis. Other viral infections (such as chickenpox, mononucleosis, mumps, and poliomyelitis) can also result in facial paralysis that may or may not resolve spontaneously.

37.15.3.4 Tumours

Tumour involvement of the facial nerve should be considered if one or more of the following features are present (1) facial paralysis that progresses slowly over 3 weeks, (2) recurrent ipsilateral facial paralysis, (3) facial weakness associated with muscle twitching, (4) long-standing facial paralysis, (5) facial paralysis associated with other cranial nerve deficits. Several benign and malignant tumours can involve the facial nerve along its intracranial, intratemporal or extracranial course. Schwannoma is one the most common tumours.

37.15.3.5 Hemifacial Spasm

Hemifacial spasm is a disorder in which the facial nerve is hyperactivity, possibly caused by a vascular compression of the facial nerve.

37.15.3.6 Miscellaneous Disorders

Simultaneous bilateral facial paralysis suggests Guillain-Barré syndrome, sarcoidosis, sickle cell disease, or some other systemic disorder. Melkersson-Rosenthal Syndrome is a neuromucocutaneous disease with the triad of recurrent facial oedema, recurrent facial paralysis associated and a fissured tongue.

37.15.4 Bell's Palsy

Also known as idiopathic facial paralysis, Bell's palsy is the sudden onset of facial weakness or paralysis. It accounts for around three quarters of all acute facial palsies, commonly occurring in the 15–45 year old age group. Whilst around 70% of untreated patients will completely recover and 85% will have complete or near normal recovery, the remainder may continue to have persistent moderate to severe weakness, facial contractures, or synkinesis. Women are more affected than men. Interestingly the condition is said to be more common during pregnancy, especially in the latter stages. In most cases, patients completely recover. However about one fifth of patients will be left with persistent weakness. The initial severity of weakness is often a good indication of prognosis.

Although the precise cause of this condition is unknown, latent herpes infection reactivated in the geniculate nerve ganglion has been suggested. Polymerase chain reaction have isolated herpes virus DNA from the facial nerve in patients. Inflammation of the nerve initially results in a reversible neurapraxia, but if this progresses degeneration of the nerve ensues. Varicella zoster virus (VZV) reactivation has also been associated with Bell palsy, but this shows a more aggressive behaviour.

Patients usually present with acute onset of rapidly progressive facial palsy. This progressively gets worse over a few days. Hyperacusis, reduced tear production and altered taste may also occur. In some patients otalgia or a sense of aural fullness may precede weakness. Severe pain suggests herpes zoster virus (Ramsay Hunt syndrome). Vesicles in the conchal bowl, soft palate, or tongue may be seen. Slowly developing weakness, especially if associated with other cranial nerve deficits or atypical headache, should raise the suspicion of a tumour.

On examination there is unilateral lower motor neurone paralysis, with drooping of the forehead, impaired closure of the eye and drooping of the corner of the mouth. Bell's phenomenon is the upward rotation of the eye on attempted closure of the eyelid. This is a protective response. In all cases of facial palsy it is important to carefully assess the ear, other cranial nerves and cerebellar function. Investigations may include polymerase chain reaction of saliva (herpes simplex virus I, or herpes zoster virus). Serological tests for Lyme disease may be required in endemic areas. However urgent imaging is most important. CT or MRI should be requested to exclude tumours.

Treatment aims to speed recovery and prevent corneal complications. Both corticosteroids (oral prednisone) and antiviral agents (acyclovir, valaciclovir, famciclovir and sorivudine) are often prescribed, but some specialists regard this as controversial. Treatment is more effective if commenced early (within 72 h), becoming less effective after 7 days. Eye care focuses on protecting the cornea from drying and abrasion. Lubricating drops should be applied hourly during the day and a simple eye ointment should be used at night. Other reported treatments include Methylcobalamin (an active form of vitamin B-12), Hyperbaric oxygen and Botulinum toxin for hemifacial spasm. Surgical decompression of the facial nerve is occasionally offered but carries risks. Initial severity of paralysis is often related to the final prognosis (Figs. 37.24 and 37.25).

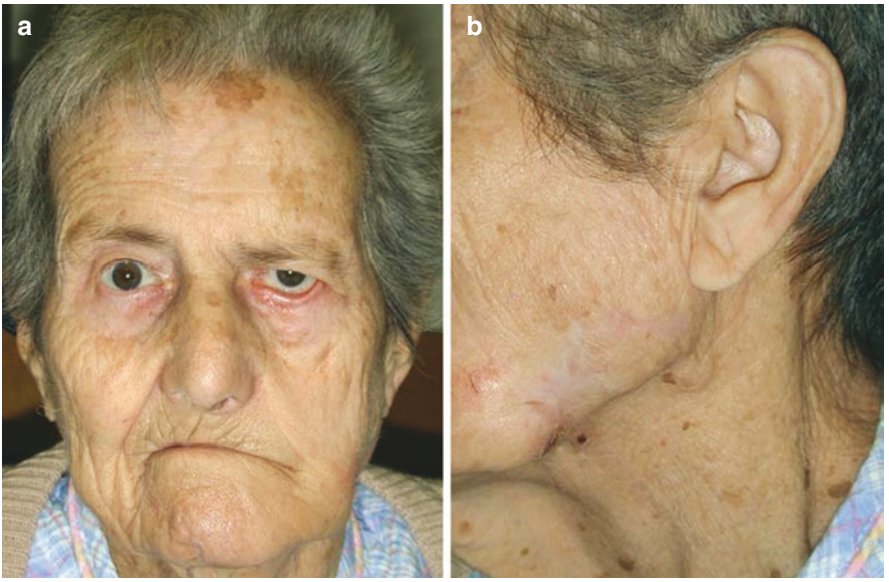

Fig. 37.24 A seventy-year-old patient who presented with a 6-month history of unresolving left-sided facial paralysis associated with facial numbness

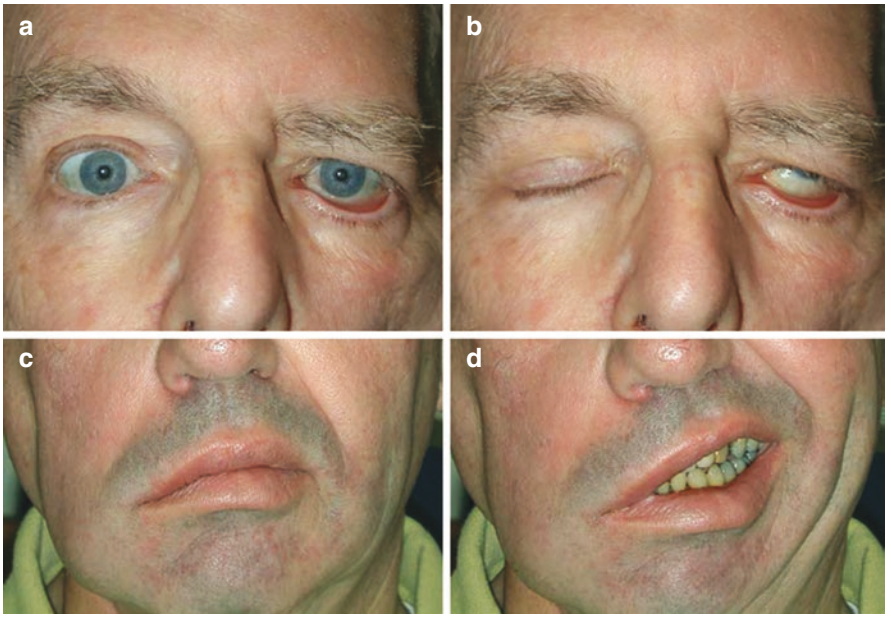

Fig. 37.25 (**a**, **b**) Physical findings of brow and periorbital functional units include brow ptosis, paralytic lagophthalmos, and ectropion. (**c**, **d**) Physical findings of perioral and lower face functional units include loss of nasolabial fold, lengthening of lip, and rotation of nose and mouth. These findings are exaggerated with facial animation

37.15.4.1 Bell's Palsy in Children

Bell's palsy in children under 10 is uncommon. Children therefore require careful assessment to exclude tumours, acute suppurative ear disease and Lyme disease. Complete facial paralysis tends to have a poorer outcome than adults.