Chapter 6: Diseases of the external ear

Valentine Hammond

The auricle

Development

(See Volume 1, Chapter 1.)

The auricle develops from the first and the second branchial arches commencing on the 38th day of fetal life. Three nodules of mesenchymal proliferation develop on the margins of each arch and by day 41 these nodules have reached maximum size, moved in a dorsolateral direction and begun to fuse. The auricle is anatomically complete by the 20th week (Melnick and Myrianthopoulos, 1979a). In the fully developed ear, the first arch contributes only the tragus and possibly a little of the anterior crus of the helix. The rest of the auricle is derived from the second arch. It is important to note that the auricle begins to develop at the level of the future upper neck and migrates in a dorsolateral direction as the mandible develops.

Many minor variations in the shape of the pinna occurs. Some of these are constant enough to warrant description. Darwin's tubercle is seen as a small elevation on the posterosuperior part of the helix. This tubercle is homologous with the tip of the mammalian ear. It is usually an inherited condition. Wildermuth's ear is a distinct entity with prominence of the antihelix and an underdeveloped helix and may be associated with other syndromes and with both sensorineural and conductive deafness. Mozart's ear consists of fusion of the helix and antihelix producing a thickened area in the upper part of the pinna. It affected both Mozart and his father and has a dominant inheritance. Not surprisingly at one time it was thought to indicate musical ability. The lobule may be absent, a condition normal in some races (Bushman of Africa and the Indians of Tierra del Fuego) and rarely it may be bifid. Failure of the lobule to separate from the side of the head is more common in females and may occur in all the females of a family (Potter and Craig, 1975). The upper part of the auricle is sometimes adherent to the head. In females and the young, the auricle is covered with very fine velous hairs but in older males coarse hair may appear on the upper margin of the helix.

Hypertrichosis

Excessively hairy ears occur only in males being a Y-linked inherited trait. Hypertrichosis is present in male offspring and siblings of affected families. The hairs are long and black and usually arise from the margin of the helix, first appearing in early adult life. The condition is most commonly seen in India but also occurs in Iran and Italy (Gates and Badhuri, 1961). An excessive growth of hair also occasionally occurs on the tragus, the so-called harbula hirci (Montagna and Giocometti, 1969).
Congenital anomalies of the auricle

(See Volume 6, Chapter 5.)

Apart from the minor variations already described, major anomalies may occur. Arrested development of the mesenchymal nodules may lead to anotia, the total absence of the auricle, or microtia where the pinna is rudimentary and malformed and usually placed lower and more anteriorly than normal. There is an association between these anomalies, meatal atresia and abnormalities of the middle ear; the degree of abnormality of the meatus and middle ear is usually proportional to the external deformity (Jafek et al, 1975). Minor dysplasias of the second arch development produce folding or defects in the helix and, where the antihelix is poorly formed and there is an excess of conchal cartilage, the typical bat ear appearance is produced.

Auricular appendages

Auricular appendages or accessory auricles occur as small elevations of skin containing a bar of elastic cartilage. They may be single or multiple and most commonly occur just anterior to the tragus or ascending crus of the helix, but may extend along a line from the tragus to the angle of the mouth and can be associated with macrostomia. They may also be associated with other congenital anomalies of the first arch. Auricular appendages may be excised but it must be remembered that they may contain bars of elastic cartilage which can extend deep into the underlying soft tissue.

Congenital aural sinuses and fistulae

Congenital sinuses usually occur in the preauricular region along the ascending crus of the helix. Others may open along a line extending from the lower border of the helix to the angle of the mouth. They are blind tracks lined by squamous epithelium. Collaural fistulae have an upper opening in the floor of the external auditory meatus and a lower one at the anterior border of the sternomastoid behind the angle of the jaw. There is an association between pre-auricular sinuses, branchial fistulae and deafness (Melnick et al, 1975; Fitch, Lindsay and Srolovitz, 1976). Both conductive and sensorineural deafness may occur; the former is a consequence of malformation of the ossicular chain and the latter a result of impaired development of the cochlea. Other associated abnormalities include deformities of the auricle, blockage of the lacrimal ducts and facial palsy. Anomalies of the genitourinary tract have also been described. If congenital sinuses and fistulae are causing no symptoms they do not require treatment. However, they may become infected leading to a persistent discharge and sometimes to abscess formation. When this happens they must be completely excised. The operation requires great care as the tract may extend deep into the soft tissues and at times be related to branches of the facial nerve.
Congenital syndromes associated with microtia and deformities of the pinna

(See Volume 6, Chapter 5.)

Numerous syndromes have been described associating microtia with other congenital defects. Some of the more important of these have been well documented by Melnick and Myrianthopoulos (1979b).

1. The Treacher Collins syndrome: mandibulofacial dysostosis associated with meatal atresia and deafness (autosomal dominant)

2. The otomandibular syndrome of Konigsmark and Gorlin: folding of the pinna, thin external nares, micrognathia and bilateral stapedial fixation (autosomal dominant)

3. The brachio-otic dysplasias: a combination of auricular malformation, cervical fistulae, conductive and sensorineural deafness (autosomal dominant)

4. The LADD syndrome: the lacrimo-auriculo-dental-digital syndrome of cup-shaped ears with deafness, and anomalies of the digits and the enamel of the teeth (autosomal dominant)

5. HRA: the hereditary renal adysplasias with ear malformation; important syndromes in this group include:
   (a) BOR: the branchio-oto-renal syndrome, combining brachial fistula with meatal atresia and renal dysplasias (autosomal dominant)
   (b) dysplastic pinna-hypospadias-renal adysplasia syndrome
   (c) dysplastic pinna-polycystic kidney syndrome (autosomal dominant)
   (d) the oto-renal-genital syndrome (autosomal recessive).

The most important aspect of these syndromes to the otologist is to emphasize the importance of renal investigations in children demonstrating familial branchial arch syndromes and the early audiological assessment of these children and those with known renal anomalies. Most of these syndromes have an autosomal dominant inheritance but, even so, there is a considerable variation in the manifestations of the syndromes within affected families and among individuals. Melnick and Myrianthopoulos (1979b) in an extensive study of these conditions concluded that the underlying developmental abnormality was probably a breakdown of neural crest integrity resulting in aberrant crest cell migration.

Potter's syndrome

This is a well-recognized congenital abnormality of the pinna which is not the direct result of genetic abnormalities affecting the development of the ear. The auricles are large and flat and this may be associated with abnormal limb positioning, pulmonary hyperplasia and compression facies. The syndrome is the result of oligohydramnios where the very small
amount of amniotic fluid present leads to flattening and compression of the pinna. By definition the oligohydramnios is the result of renal agenesis or dysplasia but it may result from loss of amniotic fluid. If the infant survives, the pinna will, in time, return to normal in the vast majority of cases.

**Congenital malformation of extrinsic origin**

The external factors which may affect the development of the fetus and result in abnormalities of the pinna are:

1. drugs
2. X-rays and radioactivity
3. viruses

A variety of drugs has been implicated in the production of congenital anomalies.

1. **Thalidomide**

   Microtia may occur in the thalidomide embryopathy and this is often associated with meatal atresia. Anomalies of both the middle and internal ears may be present. Ear anomalies occur when the drug is administered during the first 30-40 days of pregnancy (Takemori, Ishii and Suzuki, 1976).

2. **Hydantoin**

   The fetal hydantoin syndrome occurs as a result of the ingestion of this group of anticonvulsant drugs by epileptic mothers during pregnancy and may include dysplasia of the auricles.

3. **Folic acid antagonists**

   Auricular abnormalities are also seen following the ingestion of folic acid antagonists such as methotrexate and aminopterin.

4. **Warfarin**

   Warfarin may lead to microtia, although the most striking and consistent finding produced by this drug is severe hypoplasia of the nose.

5. **The fetal alcohol syndrome**

   The result of high alcohol intake during pregnancy can result in microcephaly, maxillary hypoplasia and joint anomalies, but many of these infants show an abnormality of the pinna with a marked ridge running across the concha due to hypertrophy of the crus of the helix.
**Radiation**

Radiation of the maternal pelvis during pregnancy may lead to a higher incidence of congenital malformation of the ear (Jafek et al, 1975).

**Viruses**

Maternal viral infections in the first trimester of pregnancy may be responsible for some isolated cases of external ear deformity, but there is no conclusive evidence of this.

**Steeter bands**

Steeter or amniotic bands are bands of connective tissue that stretch across the amniotic space and can result in clefts or deformities of the auricle as well as of the face, skull and limbs.

**Congenital tumours**

Haemangioma and lymphangioma may be encountered involving the auricle. Dermoid cysts occasionally occur in relation to the pinna, usually just anterior to the helix.

**Trauma to the auricle**

*(See Chapter 8.)*

Accidental trauma to the auricle may result in lacerations, or partial or complete loss. All open wounds of the auricle carry a risk of infection being introduced leading to perichondritis. Wounds should be carefully sutured and antibiotic cover given. Even when the greater part of the auricle has been detached, it has on occasion been successfully resutured into place.

The judicial removal of the auricles for theft was practised in Roman times and persisted in some parts of the world up until the 19th century. Self mutilation of the pinna is widely practised by the insertion of earrings and when this involves the cartilaginous portion of the auricle it carries a definitive risk of perichondritis. Ear piercing unless carried out with scrupulous attention to sterility also carries the risk of spreading viral diseases such as hepatitis and acquired immune deficiency syndrome (AIDS). Septicaemia may also occur (Lovejoy and Smith, 1970) and subacute bacterial endocarditis has been reported following acupuncture to the ear (Lee and McIlwain, 1985).

**Haematoma of the auricle**

Haematoma of the auricle is the result of closed trauma and occurs frequently in contact sports such as boxing and rugby football. In the 19th century it was referred to as the sanguineous tumour of the insane as a result of its prevalence among inmates of the asylums and was, therefore, thought to be a stigma of insanity. The condition results from an extravasation of blood between the cartilage and the perichondrium producing a soft doughy
swelling of the pinna. If untreated the blood clot becomes organized and the ear remains permanently thickened, producing the cauliflower ear deformity.

Treatment

Cases seen shortly after the injury has occurred may be treated by aspiration through a wide bore needle using aseptic precautions. Cases of longer-standing will require incision and the evacuation of the clot. The incision is placed along the margin of the helix and any clot present is sucked out. Again strict asepsis is essential, for if infection is introduced, perichondritis may occur. Following either aspiration or incision, a firm dressing must be applied to the pinna to prevent recurrence of the haematoma. Should it recur, further aspiration may be necessary.

Infections of the auricle

Impetigo

This is an infection of the superficial layers of the skin by staphylococci. Vesicles filled with serum arise on a reddish-purple base. Later the vesicles burst to exude serum which dries to form semi-adherent amber crusts. The condition is most commonly seen in young children and may be secondary to the otorrhoea of a middle-ear infection.

Although the impetigo may involve the whole auricle it does not extend into the external auditory meatus. Commonly the neck and face are also involved.

Treatment

The crusts are removed by bathing with warm sterile saline. The area is then dried and neomycin cream applied. The treatment may have to be repeated daily for several days. If there is otitis media or externa present, this must be treated to prevent re-infection of the skin.

Erysipelas

This is a streptococcal infection of the skin producing a raised red oedematous eruption with a characteristically well-defined edge. The auricle becomes intensely red and swollen and the infection spreads into the adjoining skin of the face. There is usually a marked systemic upset with a high temperature and rapid pulse.

Treatment

The infection usually responds rapidly to penicillin by injection.

Many generalized skin disorders may involve the pinna but require no separate description.
Perichondritis

Infection of the perichondrium of the auricle most commonly occurs when the cartilage is exposed either by laceration or by surgery. The cartilage may also be exposed as a result of frostbite or burns. Infection may be introduced during aspiration or incision of a haematoma auris. Sometimes superficial infections of the meatus or pinna spread deeply to involve the perichondrium.

In the early stages of the infection, the pinna becomes red and tender. This is followed by a generalized swelling of the pinna and eventually by the formation of subperichondrial abscesses. The pus collects between the perichondrium and the underlying cartilage. The cartilage, deprived of its blood supply, may die. Extensive cartilage necrosis results in a marked deformity of the pinna.

Treatment

Cases of perichondritis should be treated promptly with a broad spectrum antibiotic as the infecting organism is rarely sensitive to penicillin. *Pseudomonas aeruginosa* is not uncommonly found in these cases. If there is any discharge from the ear a swab should be taken for culture and the determination of sensitivities. Pending information regarding the sensitivity of the organism, treatment should be commenced.

If subperichondrial abscesses form, they should be incised and drained. Incision should be delayed until definite fluctuation can be elicited, as premature incision may result in a further spread of the infection. In relatively rare instances, pain and suppuration may continue despite these measures and gross deformity is inevitable. In such cases, the whole of the auricular cartilage (except that of the helix) must be excised, through a wide incision on the anterolateral aspect of the auricle.

Chondrodermatitis nodularis chronica helicis

This is the name given to small painful nodular lesions which occur on the upper free margin of the pinna and occasionally on the antihelix.

It is more common in men than in women and usually occurs in middle-aged or elderly subjects with outdoor occupations. The condition is thought to be due to exposure to low temperatures causing local vasoconstriction resulting in small foci of avascular chondritis.

The lesion consists of a tender nodule often covered by an adherent crust overlying a small area of exposed necrotic cartilage.

Treatment

Local excision including a small wedge of underlying cartilage results in cure.
Tophi

Small subperichondrial deposits of sodium biurate crystals may occur on the pinna in cases of gout. Although rarely troublesome they may occasionally become superficially ulcerated.

Treatment is that of the underlying condition.

Tumours of the auricle

Benign

Benign neoplasms of the auricle are uncommon. Papilloma, fibroma and chondroma do occur but need no special description.

Malignant

(See Chapter 8.)

Squamous cell carcinoma

The clinical diagnosis of an epithelioma does not usually present any difficulty. Typically the lesion presents as an indurated ulcer with everted margins. The diagnosis is confirmed by biopsy. The regional lymph nodes may be involved but this is not usually an early occurrence in tumours confined to the auricle.

Treatment

Small lesions on the upper half of the auricle can be removed by a wedge incision with a wide margin of healthy tissue and the edges of the defect sutured together. Large lesions and those involving the lower half of the auricle require total excision of the pinna. Extension beyond the pinna will require more extensive surgery and radiotherapy. Involvement of the lymph nodes is an indication for radical neck dissection. Lesions occurring on the upper half of the auricle carry a far better prognosis than those in the lower half.

Basal cell carcinoma (rodent ulcer)

Common sites for the discovery of these basal cell growths are the tragus, the border of the helix and the meatal entrance. There is at first the typical raised plaque with a rolled-over edge and tendency to central crusting. Bleeding takes place when the central crust is removed. In late cases, the whole auricle may be destroyed while the underlying bone and parotid may be infiltrated.

Cystic forms are sometimes encountered. They are smooth, often pigmented tumours without any crusting or ulceration, and when small may be confused with naevi.

The regional lymph nodes are not involved.
Treatment

Rodent ulcers of the pinna are best treated surgically, the lesion being excised together with a margin of healthy tissue.

Very small superficial lesions may be successfully treated with radiotherapy. Advanced stages with infiltration of the underlying bone and soft tissue require wide excision and postoperative radiotherapy. With adequate excision, small rodent ulcers carry a very good prognosis. When extensive infiltration of the deep tissues has occurred it may be impossible to eradicate the tumour. The patient will eventually succumb, although the progress of the disease is usually very slow.

Malignant melanoma

The auricle is rarely affected by this form of malignancy. When it occurs it is seen as a nodular pigmented lesion which tends to enlarge rapidly and eventually to ulcerate. Involvement of the regional lymph nodes and distant metastasis may occur when the primary lesion is still quite small.

Treatment

Radical excision of the lesion offers the only prospect of cure. This may involve complete excision of the pinna and an *en bloc* dissection of the regional nodes. Even with early lesions the prognosis is poor.

The ear lobe crease

This is a diagonal crease running across the lobule and is frequently present in old age. However, in Europe and North America, there does appear to be some association between the presence of the ear lobe crease in young adults and an increased incidence of coronary thrombosis (Kaukola, 1982; Overfield, 1983). A relationship between high serum cholesterol and the ear lobe crease has been demonstrated but biopsies of the crease show no abnormalities such as cholesterol deposition.

Chen et al (1982) found that, while the ear lobe crease was often seen in younger Chinese coronary thrombosis patients, their serum cholesterol levels were inversely related to the presence of the crease.

The ear lobe crease would not appear to be a reliable indicator of an increased susceptibility to coronary thrombosis.

The external auditory meatus

Development

The meatus begins to develop on the 41st day of fetal life at the dorsal end of the first branchial cleft. Ectodermal proliferation extends inwards approaching the expanding middle
ear cavity at about the 70th day and the central cells degenerate to form the meatus (Melnick and Myrianthopoulos, 1979).

**Congenital anomalies**

*(See Volume 6, Chapter 5.)*

The association between some deformities of the pinna and congenital meatal atresia or stenosis has already been noted (see above). Meatal atresia and stenosis can occur in the presence of a normal pinna.

**Acquired meatal atresia and stenosis**

**The cartilaginous meatus**

**Atresia**

A true acquired atresia of the cartilaginous meatus is rare and nearly always traumatic in origin, particularly as a result of gun shot wounds (Conley, 1946). Burns and radiation may also be responsible. Chronic otitis externa often causes stenosis but is very rarely responsible for atresia. Bilateral atresia of the outer meatus following acute otitis externa has been recorded (Marlowe, 1972).

**Stenosis**

Acquired stenosis of the cartilaginous meatus is not uncommon. It can occur as a result of:

1. **Trauma**
   - accidents: lacerations, gun shot wounds
   - surgery: usually the result of mastoid surgery
   - burns: thermal, chemical
   - radiation

2. **Infection**
   - chronic otitis externa

3. **Neoplasia**
   - both squamous cell carcinoma and adenocarcinoma may present as a progressive narrowing of the meatus.

The commonest cause of stenosis is chronic otitis externa leading to progressive fibrosis with narrowing of the canal. Stenosis may also occur at the junction of the cartilaginous and bony meatus in cases of keratosis obturans. The expansion of the bony meatus leads to exposure of the meatal cartilage at its deep attachment. Infection of the
cartilage with granulation tissue formation occurs with subsequent fibrosis and stricture formation.

**Treatment**

The only effective treatment is a meatoplasty. Repeated dilatation of meatal stenosis rarely produces lasting improvement.

Using a postaural incision, the fibrous tissue and thickened meatal skin are excised but where possible preserving a strip of skin along the roof and floor. The outer part of the bony meatus may be enlarged with burrs but preserving the overlying skin. The outer orifice of the meatus usually needs to be enlarged by excising an ellipse of conchal cartilage and turning back a flap of conchal skin and suturing it in place. The meatus is then packed and allowed to re-epithelialize. Split skin grafts are usually unnecessary.

**The deep meatus**

In the deep meatus, bony swellings, either osteomata or exostoses, represent the commonest form of stenosis. These are described separately.

**Obliterative otitis externa**

In this condition there is a progressive stenosis or atresia of the deep meatus. Bonding and Tos (1974) used the term postinflammatory acquired atresia to describe the condition.

There is always a preceding otitis externa and in some cases chronic middle ear infection is present as well. In the majority of cases there is a history of irritation and discharge, intermittent or continuous over a number of years with an increasing loss of hearing. Occasionally, the condition develops rapidly with deafness persisting after a single episode of otitis externa which may have been present for only a few weeks.

In the active phase of the disease the inflammatory changes are usually confined to the deep meatus. The skin is red and thickened and may bleed easily when cleaned. The tympanic membrane is usually obscured by granulation tissue. Some cases of granulating myringitis may represent a more localized form of the same condition.

As the inflammatory stage settles the deep meatus becomes re-epithelialized, but a mass of connective tissue persists between the outer surface of the tympanic membrane and the new fundus of the meatus with a resulting conductive deafness. No consistent bacteria are found on culture from these cases and histology of tissue obliterating the deep meatus reveals a non-specific vascular connective tissue with inflammatory cells.

At times, instead of atresia, a stenosis or web develops in the deep meatus a few millimetres external to the tympanic membrane. This can progress to complete atresia, in which event, epithelium may persist in the shut-off portion of the meatus and a meatal cholesteatoma may develop.
The precise cause of the condition is not fully understood. The bacterial infection of the deep meatus leads to a loss of epithelium from the tympanic membrane and adjacent meatal wall with granulation tissue formation. During healing the granulations are replaced by fibrous tissue and the surface of the mass is covered by epithelium, but why this type of response to infection should occur in some individuals is unknown.

Treatment

In the active granulating phase treatment is best confined to the removal of granulations and packing the meatus with ribbon-gauze soaked in a topical antibiotic/steroid preparation. There is usually a slow response to treatment and a marked tendency to recurrence. Although a few cases respond to treatment, the majority progress to stenosis or atresia. When the atresia is established surgical treatment can be considered but should be postponed until all evidence of increased vascularity has disappeared. This may take many months.

In quiescent cases with conductive deafness, surgical removal of the obstructing tissue may restore hearing. This is best undertaken via a postaural approach. The oblitative fibrous tissue is dissected away from the meatal walls and off the fibrous layer of the tympanic membrane. The deep meatus is enlarged by burring away some bone. If there is a perforation, this should be closed with a fascial graft. If the area of exposed bone is no more than a few millimetres leaving a pack in place for up to 6 weeks will often result in re-epithelialization, but more extensive bare areas are better covered with thin split-skin grafts.

Necrotic lesions

Radionecrosis

The tympanic plate appears to be unduly susceptible to radionecrosis if it is included in the radiation field during treatment of an adjoining area. Small areas of bare bone appear on the meatal floor, sometimes associated with discomfort or irritation and occasionally with a scanty discharge. These usually develop some time after the original course of radiation and may persist for many years usually healing after the separation of the tiny sequestrum. However, occasionally very extensive necrosis occurs with almost the whole tympanic plate eventually separating.

Benign necrotizing osteitis of the meatus

Patients may present with exposed dead bone in the meatal floor for no apparent reason. The history is usually one suggesting otitis externa and, again, healing may be delayed until a tiny sequestrum separates. These always seem to occur on the floor of the meatus and it has been suggested that the initial lesion is irritation or otitis externa and that it is the constant scratching of the ear with a matchstick, hairpin or some similar object that eventually causes erosion of the skin and periosteum and death of a small area of bone. While this is true in some patients, there are undoubtedly those who produce this change spontaneously over a relatively short period of time for no apparent reason. Quite why the tympanic plate has this susceptibility to necrosis remains obscure. It does not appear to be related to its pneumatization.
**Foreign bodies**

A great variety of foreign bodies may be encountered in the external auditory meatus. Insects may enter the meatus accidentally but most foreign bodies are introduced by the patient. Children and the mentally retarded account for the majority of cases. Otitis externa is often encountered in association with meatal foreign bodies. This may be secondary to the presence of the foreign body such as cotton-wool or a piece of matchstick introduced into the ear by the patient in an attempt to relieve the irritation of a pre-existing otitis externa.

**Treatment**

Insects should first be killed by instilling spirit into the external auditory meatus. Small objects are most easily removed by syringing but this method must not be used if the foreign body closely fits the meatus as it may become more deeply impacted. Vegetable foreign bodies may be hygroscopic and swell if syringed with saline. They should either be removed with small forceps or syringing should be performed with alcohol.

Large foreign bodies should be removed under direct vision with small forceps or a blunt hook, but forceps should never be used for smooth, rounded objects. It is essential that the patient remains completely still during the procedure. General anaesthesia is usually desirable in children and may be indicated in very nervous adults.

When a foreign body is impacted in the deep meatus it may be necessary to open the meatus via a postauricular incision and remove some bone from the posterior wall of the bony meatus in order to facilitate removal.

**Cerumen**

The skin lining the cartilaginous portions of the external auditory meatus contains two types of glands, sebaceous glands and modified apocrine sweat glands or ceruminous glands. Both types of gland contribute to the formation of cerumen. The quantity of wax produced varies greatly from one individual to another. In the majority, the wax dries and separates as small flakes which fall out of the meatus. However, wax may accumulate and cause deafness by blocking the meatus.

**Treatment**

Wax may be removed by syringing but this method should not be used if there is a past history of ear trouble. In these cases, the wax should be removed under direct vision using a ring-ended probe or a blunt hook and aural dressing forceps.

If syringing is undertaken, normal saline at 38° is used as the irrigating solution. Any marked variation from body temperature may cause vertigo due to labyrinthine stimulation. Either a metal syringe or a Higginson's syringe attached to a curved meatal cannula may be used.

Metal syringes should be kept well greased so that they have a smooth action. When in use the nozzle of the syringe should be supported by the hand holding the patient's pinna.
to prevent a sudden head movement forcing the nozzle deep into the meatus. If these precautions are taken, the metal syringe may be used with safety.

Before syringing, the pinna should be pulled upwards and backwards in adults and directly backwards in children in order to straighten the meatus. The stream of solution should be directed along the roof of the meatus. Syringing directly onto a mass of wax will only tend to impact it more deeply. When the wax is very hard, it cannot be removed by syringing until it has first been softened. The patient should be advised to instil a few drops of olive oil into the ear twice daily for a week.

**Keratosis obturans**

In this condition a cholesteatoma-like mass is found filling the deep meatus. The mass consists of desquamated squamous epithelium. Typically the mass has a pearly white surface but this may be obscured by overlying wax.

Keratosis obturans can produce bony erosion so that when the epithelial mass is removed a marked expansion of the deep meatus may be found. The tympanic membrane is usually intact but perforation may occur as a result of pressure necrosis. The cartilaginous meatus is not involved, but granulations may occur at the junction of the eroded bony and cartilaginous parts.

Keratosis obturans appears to arise as the result of abnormal desquamation of epithelium in the deep meatus. Unlike other skin, that in the deep meatus does not normally shed the superficial layer of cells from its surface. There is a constant migration of cells from the surface of the tympanic membrane along the deep meatus (Alberti, 1964). It would seem probable that migration fails to occur in keratosis obturans and so a mass of desquamated epithelium accumulates.

There is a not uncommon association between chronic sinusitis, bronchiectasis and keratosis obturans. Morrison (1956) attributed the keratosis obturans to an excessive secretion of wax which blocks the meatus and leads to an accumulation of desquamated epithelium in the deep meatus. It is postulated that in bronchiectasis there is a stimulation of the efferent vagal nerve ending in the bronchi, producing a reflex secretion of wax in the meatus.

Munro-Black (1964), while supporting the theory of excessive wax secretion, regards sinusitis as the primary lesion.

**Treatment**

Patients with keratosis obturans usually present with either pain or deafness in the affected ear. The treatment consists of removing the mass. This may be very difficult to achieve, especially if there is also some otitis externa present. Syringing is best avoided as it rarely succeeds in shifting the mass and may increase the patient's discomfort. If difficulty is experienced in separating it from the meatal wall it is advisable to complete the removal under general anaesthesia.
After the meatus has been cleared, the patient should be kept under observation, as the keratosis may re-form. Local applications do not appear to be of any value in preventing recurrence.

**Otitis externa**

Otitis externa is the generic term applied to all inflammatory conditions of the external meatal skin. It may arise primarily in the meatus or be a manifestation of a generalized skin condition. Predisposing factors (Peterkin, 1974) may be:

1. genetic: narrow canal, excessive wax, inherited tendency to eczema
2. environmental: heat, humidity and swimming
3. traumatic: matchsticks and hairgrips
4. infective.

The aetiology can be divided into two broad groups:

1. infective: bacterial; fungal; viral
2. reactive: eczema; seborrhoeic dermatitis; neurodermatitis.

More than one factor may be present.

Morrison and Mackay (1976) found a high incidence of excessive negative middle ear pressure in patients suffering from recurrent otitis externa. They have postulated that impaired eustachian tube function may be a factor in causing otitis externa. It is suggested that the negative pressure in the middle ear may cause discomfort inducing the sufferer to probe the ears to relieve it and thus traumatizing the meatal skin. Another possible explanation is that the negative pressure interferes with normal migration of epithelium along the external auditory meatus leading to a build up of epithelial debris in the canal. In any individual case many factors may contribute to the clinical picture. Several predisposing factors may be present and the situation may be further complicated by the development of secondary infection or eczematous reactions to the applications being used in treatment.

**Furunculosis**

A furuncle arises from a staphylococcal infection of a hair follicle. The condition occurs only in the cartilaginous meatus as hair follicles are not found in the skin of the bony meatus. The lesions may be multiple and recur over long periods.

The early symptoms of a furuncle are tenderness in the meatus and pain which is aggravated by movements of the jaw. As the condition progresses the pain becomes more severe and the meatus may become occluded by the swelling causing deafness. In severe cases the oedema may spread to the postauricular sulcus producing forward displacement of the auricle. Eventually the furuncle discharges and unless there are multiple lesions present, the condition rapidly resolves.
**Diagnosis**

Examination reveals a tender red swelling in the cartilaginous portion of the meatus with a normal deep meatus and tympanic membrane beyond it. Pain is produced on pressing the tragus and pulling the pinna upwards and backwards. There may be enlarged, tender lymph nodes palpable anterior to the tragus, over the mastoid process and below the lobule of the ear.

Difficulties in diagnosis arise when there is gross meatal swelling preventing an examination of the tympanic membrane. As swelling and tenderness may also occur in the postauricular region, the condition must be distinguished from an acute mastoiditis. The main points of distinction are shown in Table 6.1.

**Table 6.1. Distinguishing features between furunculosis and acute mastoiditis**

<table>
<thead>
<tr>
<th>Sign</th>
<th>Furunculosis</th>
<th>Acute mastoiditis</th>
</tr>
</thead>
<tbody>
<tr>
<td>Postauricular tenderness</td>
<td>Diffuse</td>
<td>Maximal over mastoid antrum</td>
</tr>
<tr>
<td>Displacement of pinna</td>
<td>Forwards</td>
<td>Typically forwards and downwards</td>
</tr>
<tr>
<td>Enlarged lymph nodes</td>
<td>Present</td>
<td>Absent</td>
</tr>
<tr>
<td>Pressure on tragus and moving the pinna</td>
<td>Pain</td>
<td>No pain</td>
</tr>
<tr>
<td>Mastoid X-rays</td>
<td>Mastoid air cells clear</td>
<td>Mastoid air cells cloudy</td>
</tr>
</tbody>
</table>

**Treatment**

When a furuncle is developing, local heat is helpful in reducing the pain and accelerating the inflammatory process. Heat may be applied by means of a covered hot-water bottle, electric pad or as short-wave diathermy.

In the early stages, local dressings are painful to apply and of no therapeutic value. Incision should be avoided as there is a danger of spreading the infection, especially to the cartilage.

When a furuncle has begun to discharge, the pus should be carefully mopped away. A wick soaked in glycerine may then be inserted. This dressing should be changed daily until the lesion is dry.

Systemic antibiotics are indicated when there is marked oedema or adenitis present, or in cases with multiple furuncles. Penicillin is usually the antibiotic of choice, although an increasing number of penicillin-resistant staphylococcal infections are encountered. A swab should always be taken for culture and sensitivity tests, and penicillin started pending the results. Flucloxacillin is indicated for penicillin-resistant organisms.
Recurrent furuncles

The above methods of treatment are used in recurrent furunculosis, but steps must also be taken to eliminate the staphylococci from the external auditory meatus. The organisms are often carried in the nasal vestibules in these cases and this site also needs attention. A cream containing neomycin or gentamicin should be applied to both the meatus and the nasal vestibules twice daily. Tests should always be carried out to exclude diabetes mellitus.

Diffuse otitis externa

This condition has received a variety of names in the past, emphasizing its frequent occurrence in hot and humid climates, for example 'tropical' ear, Singapore ear. However, diffuse otitis externa is widely encountered in all climatic conditions. Although heat, humidity and bathing are aggravating factors in some cases, the most important factor is local trauma. Scratching the ears, vigorous drying of the meatus with a dirty towel and unskilled syringing are some of the ways in which minor abrasions of the meatal skin may be produced. These abrasions provide access for the causative organisms.

Some cases are secondary to an underlying chronic suppurative otitis media and this possibility should always be considered and excluded by careful examination of the tympanic membrane.

The organisms most commonly found in diffuse otitis externa are Pseudomonas aeruginosa, Bacillus proteus and Staphylococcus aureus.

The condition is seen in two stages - acute and chronic.

The acute stage

The symptom of the acute stage is discomfort developing into pain in and around the ear. The pain is aggravated by movements of the jaw. In severe cases, there may be swelling of the surrounding soft tissues and outward displacement of the pinna. On examination, the meatal skin is red, swollen and very tender. Pus is found in the meatus and, as the disease progresses, the meatal epithelium desquamates forming a mass of cheesy debris in the deep meatus. The tympanic membrane is often dull and injected in appearance.

Treatment

The most important part of treatment is the meticulous cleaning of the meatus. Particular attention must be paid to the deep anteroinferior meatal recess where pus and debris accumulate. A swab should be taken and cultured. After cleaning, the meatus is packed with 12 mm ribbon gauze impregnated with a broad spectrum antibiotic such as neomycin or gentamicin and changed daily or a Pope's wick can be inserted and moistened with drops containing similar antibiotic preparations.

Topical antibiotics must be used with caution as sensitization of the skin may occur and they may encourage the development of fungal infections. The patient must keep the ear dry and avoid rubbing or scratching.
The chronic stage

The chief symptoms of the chronic stage are irritation and discharge. Deafness may occur as a result of the accumulation of debris in the meatus. There is no tenderness, but there may be thickening of the meatal skin with a reduced lumen. Pus and debris are found in the meatus. There may be small granulations on the surface of the tympanic membrane denoting a loss of epithelium.

Treatment

As in the acute phase, careful cleaning of the meatus with clearance of the deep meatal recess is the essential part of treatment. This is best achieved under the microscope with the use of suction. If there is marked meatal swelling, this can be reduced by packing the meatus daily with 12 mm gauze wicks impregnated with an antibiotic such as neomycin or gentamicin or an antiseptic (clioquinol) combined with a steroid or as drops applied to a Pope's ear wick. The addition of the steroid helps both to reduce the inflammatory swelling and to control the irritation. When there is no appreciable meatal swelling the antiseptic and hydrocortisone cream may be applied to the meatus. Ear drops of neomycin or gentamicin, and hydrocortisone, are often effective in clearing up the infection at this stage but may produce a sensitivity reaction in some individuals. This may be difficult to recognize as it may be masked by the presence of the hydrocortisone in the preparation. In cases which fail to respond to treatment the reasons may be:

1. underlying chronic suppurative otitis media
2. fungal infection
3. sensitization of the skin to the topical application being used.

Otomycosis

Although much commoner in tropical climates fungal infections of the meatus are not infrequently encountered in temperate regions.

Otomycosis may develop as a primary infection or as a mixed infection with bacteria. It may result from the prolonged treatment of an initial bacterial infection with antibiotic and steroid preparation, although a recent study did not confirm this (Mugliston and O'Donoghue, 1985). The fungi most frequently isolated in otomycosis are *Aspergillus niger* and *Candida albicans*. Symptomatically, the condition may be indistinguishable from bacterial otitis externa although the irritation is usually more marked in fungal infections.

Occasionally severe pain in the ear is the presenting symptom. On examination a mass of greyish-white debris, resembling wet blotting paper, may be seen filling the meatus. In infection with *Aspergillus niger* the conidiophores may be seen as black specks in the debris. At times, a mass of fine filaments are seen projecting from the meatal wall. The typical appearances are not always present and, in any case of otitis externa that fails to respond to treatment, the possibility of a fungal infection should be considered. The diagnosis can be confirmed by microscopical examination of the debris or by culture.
**Treatment**

Fungi thrive in moist conditions and in the presence of epithelial debris. It is therefore essential to remove all the debris and discharge from the meatus. A specific antifungal agent can then be applied. The most widely used of these is nystatin. Nystatin is particularly effective against *Candida* species, but less active against the aspergillus group of fungi. Econazole nitrate is a broad spectrum antifungal agent which is proving more effective for aspergillus infections. It is also active against some Gram-positive bacteria (staphylococci and streptococci). Amphotericin is also used and has a similar range of activity to nystatin.

These preparations are best applied as powders but may be used in a liquid form. Treatment should be continued for at least a week after the infection has apparently resolved.

**Otitis externa haemorrhagica (bullous myringitis)**

This condition is characterized by the formation of purple blebs on the tympanic membrane and the skin of the deep meatus. The purple colour is due to the haemorrhagic effusion filling the vesicles.

Pain, often severe, is the first symptom and serosanguineous discharge may occur as a result of bursting of the blebs. The pain is not relieved by the onset of the discharge. In uncomplicated cases the middle ear is not involved and the hearing remains normal.

The aetiology of the condition is uncertain but it is thought to be the result of a viral infection. In some influenza epidemics many cases of otitis externa haemorrhagica are seen and there does appear to be an association between the two conditions. Treatment consists of prescribing analgesics for the pain and keeping the ear clean and dry.

Antibiotics have no influence on the course of the disease. The blebs should not be incised as this is of no value in relieving the symptoms and may only introduce secondary infection.

**Herpes zoster oticus**

*(See Chapter 24.)*

Herpes zoster of the geniculate ganglion may give rise to skin lesions with or without involvement of either the seventh or eighth cranial nerves. The herpes eruption occurs on the meatal skin, tympanic membrane and the auricle, particularly in the conchal region. Initially, the rash consists of small tense blisters with surrounding erythema. Lesions may also be found on the buccal mucosa and the hard palate. The blisters gradually dry up, leaving adherent crusts which usually persist for 7-10 days.

The appearance of the rash is often preceded by pain in the ear for several days. Apart from keeping the ear dry, no local treatment is indicated.
Herpes simplex

Herpes simplex occurs most commonly on the lips as the so-called 'cold sore'. Occasionally the skin of the auricle and meatus are affected. The eruption at first consists of a crop of small vesicles which dry up after a few days leaving the skin red and scaly. There is no specific treatment and, apart from keeping the ear dry, no local treatment is necessary.

Seborrhoeic dermatitis

The main feature of this disease is a scaly condition of the scalp usually referred to as dandruff or scurf. This is often associated with scaling in the external auditory meatus, postauricular sulcus and below the lobe of the auricle. The aetiology of the condition is unknown. When the ear is involved, secondary infection may be introduced by scratching, leading to a diffuse otitis externa.

Treatment

The scalp condition always requires attention. Regular washing with a cetrimide shampoo is an effective method of keeping the dandruff under control. In uncomplicated cases, there may be a tendency for debris to accumulate in the meatus. This may require regular removal. The patient should be advised to avoid getting water in the ears and to refrain from attempting to remove the waxy debris.

Eczema

The eczematous reaction occurs as the result of sensitization of the skin cells. This sensitization may be produced by an infecting organism or by contact with an allergenic material. Of the latter group, the substances which most commonly evoke this response are the antibiotics; neomycin is by far the most troublesome in this respect. The topical application of any antibiotic may result in a sensitivity reaction. Clinical the eczematous reaction is characterized by the formation of vesicles. When the vesicles burst, serous discharge exudes from the raw surface. The eruption is usually accompanied by intense irritation.

Treatment

When the eczematous dermatitis is secondary to an infective process, the condition is best treated by cleaning the meatus and applying a cream containing clioquinol and hydrocortisone.

In cases resulting from the topical application of an antibiotic, the preparation responsible must be withdrawn. The ear should be kept dry and either a cream or lotion containing a topical steroid preparation applied regularly until the condition resolves.

Neurodermatitis

In some cases of otitis externa, there is an underlying psychosomatic disturbance which not only initiates the condition but also makes it difficult to clear it up. In these
patients, the initial symptom is irritation in the ear. At this stage, the skin is normal in appearance. Constant scratching may lead to lichenification of the skin or secondary infection may be introduced, causing a diffuse otitis externa.

**Treatment**

Local treatment consists of clearing up any secondary infection and attempting to alleviate the irritation with steroid preparations. In severe cases it may be necessary to bandage the ears to prevent scratching. In management, due attention must be paid to the psychological aspect of the problem.

**Malignant otitis externa**

This term was first applied by Chandler (1968) to describe a severe progressive infection starting in the external meatus and rapidly involving the temporal bone and adjacent soft tissues. Although it usually occurs in elderly, poorly controlled diabetic patients, cases do present in the middle-aged and when the diabetes is well controlled. Zaky et al (1976) in a review of the literature recorded that over 91% of subjects were over the age of 55 years and that 93% were diabetic. The condition is also seen in patients receiving immunosuppressive drugs and cases have been reported in children suffering from malnutrition and anaemia (Joachims, 1976).

The infecting organism is *Pseudomonas aeruginosa*. The infection starts as a cellulitis of the external auditory meatus. It may develop in a pre-existing chronic otitis externa but is often insidious in onset with minimal evidence of meatal infection. Granulomata may appear in the deep meatus usually arising from the floor and overlying areas of osteitis.

There is a rapid spread of infection to the adjacent soft tissues either via the tympanomastoid suture or via the clefts of Santorini, the naturally occurring fissures in the cartilage of the meatal floor, followed by spreading cellulitis of the skull base travelling in the soft tissue planes (Kohut and Lindsay, 1979). The infection also spreads by the vascular channels in the tympanic plate and petrous bone (Nadol, 1980). The pseudomonas organism tends to spread along vascular channels (Teplitz, 1965; Riff, 1971; Zeigler and Douglas, 1979). In addition it produces a number of exotoxins and enzymes, including an elastase, which digest vessel walls, and some enzymes from pseudomonal organisms have a collagenase effect (Lucente, Parisier and Som, 1983).

After the tympanic plate, the next area of bone involvement is usually in the region of the mastoid tip and stylomastoid foramen leading to early facial palsy. This bone involvement would appear to be secondary to the soft tissue infection and there is often no continuity of affected bone between the deep meatus and the stylomastoid region. In this stage of infection, the middle ear and mastoid cells may be spared even when a subperiosteal abscess develops over the mastoid region. Tympanotomy and mastoid exploration may show no evidence of active disease.

Further spread of the infection can lead to involvement of the lateral sinus and the superior and inferior petrosal sinuses. Secondary osteomyelitis at the petrous apex can spread to the floor of the middle cranial fossa or to the basisphenoid with the development of
sphenoidal sinusitis. During the course of the spreading infection cranial nerve palsies may develop. The seventh nerve is most frequently affected followed by IX, X, XI and VI. Even when the infection is apparently well controlled, further evidence of deep extension may appear after many weeks or even months. Spread across the midline to the opposite petrous bone via the basisphenoid can occur presenting with contralateral sixth and tenth nerve palsies and a middle ear effusion due to eustachian tube involvement. Nadol (1980) in post-mortem studies has demonstrated spread of infection anterior to the foramen magnum via the basisphenoid and cavernous sinuses with involvement of the contralateral peritubal area.

Extensive spread of infection may occur with few clinical signs. The main clinical features are the initial otitis externa or meatal granulomata associated with severe pain and the subsequent development of cranial nerve palsies. Radiological assessment is often unhelpful as there may be little involvement of bone and the middle ear and mastoid air cells may remain clear. Computerized tomography (CT) scanning can be useful but may also fail to demonstrate disease until it is very extensive. Isotope bone scanning may demonstrate increased uptake in the region of the skull base in active cases (Garty, Rosen and Holdstein, 1985; Salit, McNeely and Chait, 1985). Magnetic resonance scanning may well prove to be helpful in the future.

**Treatment**

As the infection is predominantly one of soft tissue, antibiotic therapy is the first choice of treatment. Intravenous therapy with the appropriate antipseudomonal antibiotics based on sensitivity tests, should be given for up to 3 months. It has been claimed that 6 weeks' treatment is adequate (Uri et al, 1984), but in well-established cases further activity may develop if the antibiotics are not continued for the longer period. The intravenous antibiotic therapy usually recommended is an aminoglycoside combined with azlocillin or ticarcillin (which have replaced carbenicillin, as they are more effective). Aminoglycosides are both ototoxic and nephrotoxic and the dosage should be carefully monitored by measuring plasma concentrations.

If these drugs cannot be used either because of sensitivity or toxic effects a third generation cephalosporin may be indicated.

Generally, surgery should be confined to the drainage of subperiosteal abscesses and the removal of necrotic or sequestrated bone. A radical mastoidectomy alone has little value in most cases as the disease does not usually spread via the air cells; however a subperiosteal abscess may develop deep to the mastoid process and may require drainage (Raines and Schindler, 1980). A striking feature of the disease is the rapid relief of pain following the commencement of intravenous antibiotic therapy. The severe pain usually associated with the disease rapidly diminishes and may cease completely within a few days.

When treatment has been completed, long-term follow-up is essential as late recurrences may occur between 3-6 months after the end of intravenous therapy. The most reliable indicators of renewed activity are a recurrence of pain, the development of new cranial nerve palsies and elevation of the erythrocyte sedimentation rate (ESR).
Benign tumours of the meatus

Fibroma, chondroma and angioma may occur in the external auditory meatus but require no special description.

Papilloma

Viral papillomata occur in the outer meatus, often associated with similar lesions on the fingers. These viral warts can be removed by curetting under local or general anaesthesia.

Diffuse papillomata of the meatus are occasionally encountered. These have the typical papilliferous appearance but may extend into the deep meatus and obscure the tympanic membrane. They can be removed percutaneously in most cases but may recur locally.

Adenoma

There are two types of gland in the skin of the external auditory meatus and both may give rise to adenomata.

1. Sebaceous adenoma. This tumour arises in the sebaceous glands of the meatus. It is seen as a smooth, painless, skin-covered swelling in the outer part of the meatus. It may be treated by local excision.

2. Ceruminoma. This is a rare tumour arising from the ceruminous glands of the meatal skin. The glands are modified apocrine sweat glands and histologically a ceruminoma closely resembles the sweat gland tumours seen elsewhere in the skin and for this reason the term hidradenoma of the meatus is now generally preferred. Clinically the tumour presents as a firm skin-covered mass in the cartilaginous meatus. Both sessile and polypoid forms have been described (O'Neill and Parker, 1957; Juby, 1957; Arora, 1964). There are no symptoms until the mass enlarges sufficiently to cause a feeling of obstruction in the ear.

Treatment

Ceruminomata show a marked tendency to local recurrence after removal and may become frankly malignant adenocarcinomata (see Chapter 22).

Osteoma

The solitary cancellous osteoma occurs as a smooth, rounded pedunculated tumour attached to the outer part of the bony meatus. It arises from the region of either the tympanosquamous or tympanomastoid suture.

These tumours can be readily removed by fracturing through their narrow attachment to the meatal wall. Very rarely an osteoma of the temporal bone may impinge at the meatus producing a diffuse swelling in the deep meatus arising from one wall. X-rays will demonstrate the true extent of the lesion. This type of osteoma may be associated with similar lesions arising from other bones. Complete removal of these tumours may prove difficult or unwise, but simple burring away of the meatal projection to create an adequate canal may be
followed by recurrence. If the condition is causing symptoms, as much of the osteoma as possible should be removed.

**Exostoses**

Exostoses produce smooth sessile hemispherical elevations in the deep part of the meatus adjacent to the tympanic membrane. The lesions are multiple usually occurring in a group of three. They consist of dense ivory bone covered by a thin layer of normal deep meatal skin. The relationship between cold-water bathing and the formation of meatal exostoses is now widely accepted. Van Gilse (1938) was the first to demonstrate a higher incidence in cold water bathers.

Fowler and Osman (1942), working with guinea-pigs, were able to demonstrate the formation of new bone on the inner surface of the tympanic bulla following irrigation of the external canal with cold water. Harrison (1962) carried out similar experiments with guinea-pigs and found histological evidence of new bone formation in the deep meatus.

Meatal exostoses do not cause any symptoms when they are small and so are usually discovered incidentally during examination of the ears. When large them may completely block the meatus or so greatly reduce the lumen that it is readily blocked by small amounts of wax or epithelial debris. When this occurs the patient will complain of deafness.

**Treatment**

When exostoses are small they require no treatment. If they are large enough to cause deafness or impede the treatment of a chronic middle-ear infection, they should be removed.

As exostoses consist of dense ivory bone, removal must be carried out under magnification using a high speed drill and both cutting and diamond burrs. The meatus is exposed through a postaural incision and the skin overlying the exostoses is elevated and preserved as far as possible. The bone is then drilled away until an adequate meatus is fashioned taking great care not to damage the underlying tympanic membrane. At the end of the operation the skin flaps are replaced and a pack inserted and left in place for 10 days.

**Malignant tumours of the meatus**

*(See Chapter 22.)*