Paparella II: Otology and Neuro-Otology

Section 3: Diseases of the Ear

Part 3: Middle Ear and Mastoid

Chapter 29: Chronic Otitis Media and Mastoiditis

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Long-standing inflammations of the middle ear cleft (eustachian tube, tympanum, attic, antrum, mastoid air cell system) occur in a multitude of forms due to anatomic, physiologic, and bacterial factors. Proper analysis and interpretation of chronically diseased ears requires intensive knowledge of these factors. The fact that no two ears are alike makes this subject both difficult and fascinating.

Chronic ear inflammations behave very differently from acute inflammations. In the latter the process is rapid in onset, quick in resolution and usually free of any significant sequelae. The chronic inflammation, on the other hand, is slow and insidious in its course, tends to be persistent, and is very often destructive. Healing may occur spontaneously, however, and the disease be arrested, but there are often irreversible sequelae. A few of the acute middle ear infections cause destructive processes which persist and are subsequently responsible for a chronic ear inflammation.

As a rule, acute middle ear suppurations resolve within a period of 6 weeks. Suppurations continuing for a longer period of time usually cause irreversible damage to the middle ear cleft mucosa and the underlying bone. These patients usually present themselves with otorrhea and hearing impairment, but not always. The otologist is very familiar with chronic middle ear disease which is detected on routine otologic examination without the patient having been aware of impaired hearing and without history of otorrhea (early incipient keratoma).

Since the onset of chronic middle ear disease is often insidious, a patient may not consult an otologist until symptoms of a complication occur (facial paralysis, vertigo, pain, headache). In these chronic middle ear diseases of an inflammatory progressive nature, the otologist must arrest the disease and also attempt to restore the middle ear to the best possible function.

Some of the middle ear diseased states are potentially dangerous and require urgent surgical intervention. It is therefore very important to distinguish between two main types of chronic middle ear disease, the dangerous and the benign. Mawson (1963) stated that the difference between these two types is dependent on the embryonic pattern of development. Chronic infection in ciliated columnar epithelium seldom, if ever, involves the underlying bone and is confined to the mucosa, whereas infection in the flat pavement epithelium of the attic and antrum is associated with erosion of underlying bone.

We may distinguish between active, inactive, quiescent, and healed states of chronic middle ear cleft inflammations. In the *active* state there is a steady discharge of pus. In the

quiescent state it is an intermission between discharges of pus. The history would indicate a probability of recurrent otorrhea rather than a terminal cessation. An *inactive* state implies that a previously discharging ear is now dry and that a previously discharging ear is now dry and that is probably will not resume discharging. The possibility of its discharging again cannot be reasonably excluded, however. We see this in dry perforations of the drumhead. A *healed* state indicates a permanently controlled middle ear inflammation. The perforation of the tympanic membrane is now healed or repaired.

Anatomic Factors in Chronic Middle Ear Cleft Disease

Middle Ear Embryology

In order to better understand the middle ear cleft with its viscera (ossicles, mucosal folds, and so forth) we will briefly review the embryology.

Between the third and seventh fetal months the gelatinous tissue of the middle ear cleft is gradually absorbed. At the same time the primitive tympanic cavity develops by a growth of an endothelium-lined fluid pouch extending from the eustachian tube into the cleft. Four primary sacs or pouches then bud out. They are the saccus anticus, saccus medius, saccus superior, and saccus posticus (Hammar, 1902). Where these pouches contact each other, mucosal folds are formed. Between the mucosal layers of the folds are remnants of the mesoderm, including blood vessels supplying the "viscera" of the tympanic cavity.

The saccus anticus is the smallest of the pouches. It extends upward anterior to the tensor tendon to form the anterior pouch of von Tröltsch. Its upward extent may be limited at the level of the semicanal for the tensor tympani by contact with the anterior-most saccule derived from the faster-developing saccus medius. The fold formed is the tensor fold and above it is the anterior compartment of the attic. The saccus anticus may, however, extent upward to the tegmen and as far posterior as the superior mallear fold. In this instance there is developed a supratubal space instead of an anterior attic compartment.

The saccus medius forms the attic. It extends upward through the isthmus tympani anticus and usually breaks up into three saccules. The anterior saccule may form the anterior compartment of the attic. The medical saccule forms the superior incudal space by growth over the malleoincudal body to the lateral incudal fold and posterior incudal ligament. The medial saccule usually sends an offshoot forward between the lateral mallear and lateral incudal folds to form Prussak's space. Occasionally the medial saccule extends only to the level of a superior incudal fold, in which case the lateral incudal fold is absent. The superior incudal space would, in such an instance, be developed from the saccus superior. The posterior saccule of the saccus medius extends posteriorly to the anterior crus of the stapes, passes medial to the long crus of the incus, and eventually pneumatizes that portion of the mastoid air cell system which is derived from the pars petrosa of the temporal bone.

The saccus superior extends posteriorly and laterally in the interval between the malleus handle and the tip of the long crus of the incus. It forms the posterior pouch of von Tröltsch and the inferior incudal space. Its upper limit is the lateral incudal fold, although it may extend to the level of a complete superior incudal fold and also extend into Prussak's space. Posteriorly the saccus superior extends medially to pass over the pyramidal eminence

into the antrum. Eventually it pneumatizes that portion of the mastoid which is derived from the pars squamosa. Persistence and further development of the mucosal fold between the saccus superior and the saccus medius in the antrum and the mastoid of the adult will result in a bony partition known as Körner's septum.

The saccus posticus extends along the hypotympanum to form the round window niche, sinus tympani, and greater portion of the oval window niche. When the plica stapedis and membrana obturatoria are present they indicate the furthest advance of the saccus posticus. The saccus posticus, however, often extends under the stapedial tendon to pneumatize the posterior tympanic sinus.

In the region of the oval window niche three pouches play an intimate part in the development: the saccus posticus extends upward from the hypotympanum, hugging the bony medial wall of the tympanum and forming the lower half of the niche itself; the saccus superior passes lateral to the long crus of the incus over the saccus posticus and the stapedial tendon on its way to the antrum; the saccus medius extends medial to the long crus of the incus between the stapes crura and facial canal and superior to the saccus superior to enter the antrum.

The Eustachian Tube

Interferences with normal function of the eustachian tube play an important role in chronic inflammations of the middle ear cleft. The auditory tube is a very complex structure anatomically (Proctor, 1967) and physiologically. Inadequate ventilation of the middle ear via the tube is the basic cause for most middle ear inflammations. Enlarged and inflamed adenoid tissue is a common offender, not because of direct obstruction of the pharyngeal lumen, but because of stasis of the tubal lymphatics. The larger diameter and shorter tube in the infant facilitates the passage of infected secretions up the tubal lumen into the middle ear. For a time it was thought that lymphoid tissue within the lumen (Gerlach's tonsil) was a factor, but Farrior (1943) and others could find no evidence to support this concept. Other causes are malfunction of the tubal muscles (levator and tensor veli palatini), such as is seen in some cleft palate patients; reduction in the size of Ostman's fat pad as a result of starvation; atrophy of the tensor veli palatini muscle following cutting of the motor branch of the trigeminal nerve when doing a retrogasserian neurectomy; collagen diseases affecting the tubal ligaments and elastic tissues; loss of ciliated tubal epithelium; and abnormally thick, tenacious tubal secretions.

The Tympanum

Some temporal bones are relatively small, with a narrow mesotympanum, a shallow hypotympanum, and small recesses in the posterior tympanum. In such ears inflammations have a more difficult time resolving because of the ease of occlusion of the air space by swelling of the mucosa.

Attention is called to the relatively small communications between the mesotympanum and the epitympanum. These spaces are separated by the ossicular chain and their associated mucosal folds. These folds may be nearly complete in a living person, and in childhood they are usually well preserved. There are always two small openings up into the attic, one

between the tensor tympani tendon and the stapes (anterior isthmus), and the other between the short crus of the incus and the stapedial tendon and pyramidal eminence (posterior isthmus). If the middle ear cleft is poorly pneumatized, the isthmuses are particularly prone to occlusion by edematous mucosa, thus closing aeration of the mastoid cells. If air cannot reach the mastoid cells, those cells quickly replace the trapped air with fluid (transudate or exudate if infected). If ventilation of the mastoid is not re-established, these fluids become more and more concentrated (ie, cholesterol granulations) and eventually result in irreversible cicatrixes (adhesive otitis media or tympanosclerosis). It is entirely possible to occlude the isthmuses by keratoma, polyps, or edema and to develop chronic inflammatory disease in the antrum and mastoid air cells and yet have a normally ventilated eustachian tube and tympanum.

Compartmentalization of the middle ear air-cell system tends to limit the extent of disease, at least in its earlier stages. These spaces or compartments are variable but can be explained embryologically. Recognition of disease limited to one compartment may prevent the opening of an uninvolved compartment. For instance, keratoma may be confined to the attic and may not have "dropped" through the isthmuses into the mesotympanum. The matrix can sometimes be peeled away from an intact but perhaps stretched mucosal fold without opening into the mesotympanum. Such patients will often have good residual hearing, and this may be better than the hearing that might result from reconstructive surgery if tissues are removed unwittingly.

Preservation of the tensor fold is particularly important. This fold constitutes the floor of the anterior compartment of the attic. Its removal makes closure of the opening into the anterior mesotympanum much more difficult. This would require removal of the greater portion of the anterior tympanic spine and provision for a graft to close the dehiscence.

Keratomas usually develop by invasion of Prussak's space and thence into the attic, passing through the posterior embryonic dehiscence in this space. Other keratomas originate from a posterior marginal defect in the tympanic membrane to invade the inferior incudal space. They may then extend through the posterior tympanic isthmus to enter the antrum. In some, the attic is spared for some time. These keratomas may extend to involve the various sinuses of the posterior tympanum. In those rare instances where Prussak's space is formed by the saccus anticus a keratoma may extend from Prussak's space directly into the anterior compartment of the attic or into the supratubal space.

Prussak's Space

It is helpful to review the anatomy of Prussak's space. The annulus fibrosus, the dense fibrocartilaginous ring to which the radial fibers of the drum attach, leaves the sulcus tympanicus posteriorly. Its outer fibers insert on the posterior tympanic spine or extend in the stria membrana tympani posticus to the short processus of the malleus. Its inner fibers insert on the medially placed pretympanic spine (Dworacek, 1960; Perkopf, 1960) or radiate out, forming the supporting structure for the posterior mallear fold and attaching on the posteromedial aspect of the upper third of the malleus handle. Between the posterior mallear fold and the drum lies the posterior pouch of von Tröltsch.

Anteriorly the annulus fibrosus leaves the sulcus tympanicus to attach in part to the anterior tympanic spine and then continues on (1) as the stria membrane tympani anticus to the short process, (2) to radiate out to help to form the floor of Prussak's space, (3) to interdigitate with fibers of the lateral mallear fold, and (4) to attach to the bony rim of the notch of Rivinus.

The lateral mallear fold arises from the junction of the malleus head and neck and radiates out to insert on the entire bony rim of the notch of Rivinus, thus forming a firm roof for Prussak's space. The scutum slopes medially at an acute angle above the lateral mallear ligament so that an expanding keratoma in Prussak's space can obliterate this angle by causing adherence of the lateral mallear ligament to a higher level. The lower narrower edge of the scutum can be eroded away without opening into the attic above until it is exteriorized sufficiently to release the trapped keratin plug.

From Prussak's space, keratoma pouches expand in one of three directions, depending on the embryonic route that developed Prussak's space.

Posterior Route. This is the commonest route. The extension would be into the superior incudal space, which lies in the posterolateral portion of the attic. This would be above the lateral incudal fold and incus body.

Inferior Route. This is a frequent route. Prussak's space can pneumatize via the inferior incudal space (saccus superior). A keratoma following this route would easily be seen behind the tympanic membrane in the inferior incudal space. The keratoma would then be in the mesotympanum and not in the attic.

Anterior Route. This is rare. Prussak's space is developed in these patients from the saccus anticus. A keratoma would again extend into the mesotympanum, first into the anterior pouch of von Tröltsch and then into the protympanum. It is important to recognize a keratoma early and to arrest its progression before it extends any further.

Compartments of the Middle Ear

Since we consider progression of keratoma in this chapter, it is well to review the various compartments of the mesotympanum and attic that are invaded by keratomas in their progression.

The attic is separated from the mesotympanum by the ossicular chain, tendons of the tensor tympani and stapedius muscles, anterior and lateral mallear ligaments, and posterior incudal ligament. In addition, there are mucosal folds (tensor, superior and lateral mallear, lateral and medial incudal, plica stapedis, and membrane obturatoris stapedis). The folds are important because they carry blood vessels to the ossicles (Hamberger et al, 1963). They are usually all present in the living. They are best demonstrated on fresh autopsy material which is dissected immediately and not permitted to dry out; otherwise they shrivel up to minute strands or disappear completely.

The attic usually extends forward through the incisura tensoris and anterior to the tensor tendon as the anterior malleolar space or anterior compartment of the attic. This space

lies above the tensor fold, which extends laterally from the semicanal for the tensor tympani muscle to the anterior mallear ligament.

The attic, however, may extend anteriorly only to the level of the tensor tendon, where it is limited by a medial extension of the superior mallear fold instead of by the tensor fold, which in such an instance does not form. The space anteriorly would be in communication with the mesotympanum and eustachian tube. This space, when present, is called the supratubal space.

Posterior to the transversely placed superior mallear fold lies the larger posterior compartment of the attic. That portion of this compartment medial to the superior incudal fold may be considered as the medial incudal space. Laterally the floor of the superior incudal space is formed by the lateral mallear fold and by the lateral incudal fold, which extends posteriorly to the posterior incudal ligament. The entrance into Prussak's space is usually located between the lateral mallear fold and the lateral incudal fold.

Medially the posterior compartment of the attic is separated from the mesotympanum by the dihedral-shaped medial incudal fold, which extends from both crura of the incus to the pyramidal eminence and stapes.

Beneath the floor of the attic and in the upper mesotympanum there are three compartments; they are the inferior incudal space and the anterior and posterior pouches of von Tröltsch.

The inferior incudal space extends from the inferior surface of the incus laterally to the posterior mallear fold. It is limited medially by the medial incudal fold and anteriorly by the interossicular fold which lies between the long crus of the incus and the upper two thirds of the malleus handle.

Between the posterior mallear fold and the tympanic membrane lies the posterior pouch of von Tröltsch. The chorda tympani nerve lies in the free margin of the posterior mallear fold, although it may cross the posterior tympanum independent of this fold. The shallow anterior pouch of von Tröltsch lies between that portion of the drumhead anterior to the malleus handle and the anterior mallear fold which is draped on the anterior mallear ligament. Between the malleus handle and the drumhead and superior to the umbo lies the shallow manubrial fold.

Five folds may be recognized as stapedial folds. They are the (1) obturatoria stapedis between the crura, (2) anterior stapedial folds between the promontory and the anterior crus, (3) posterior stapedial between the promontory and the posterior crus, (4) plica stapedis between the pyramidal eminence and the posterior crus, (5) and superior stapedial folds, which extend from the long crus of the incus to either crus of the stapes or from the facial canal to the crura.

The Posterior Tympanum

The posterior or mastoid wall of the tympanum is complete. It is derived from the second branchial arch (Reichert's cartilage). It completely fills the interval between the bony

annulus tympanicus and the bony labyrinth. This wall is variably shaped and is the extension of the styloid process upward to the level of the fossa incudis and the pyramidal eminence (Proctor, 1969). The wall presents three eminences anteriorly: styloid, pyramidal, and chordal. Hollowed-out areas or sinuses are also present in the posterior wall (sinus tympani, posterior tympanic sinus, lateral tympanic sinus, and facial sinus). These sinuses do not communicate with the mastoid air cell system. Diseased states in these sinuses must be recognized in the management of chronic middle ear disease. The otologist must also know how to clear these sinuses of disease when operating on the ears of the patient.

The Mastoid Air Cells

Surgery for acute mastoiditis has been directed to removal of as much of the mastoid air cells as possible without removal of the posterior bony canal wall or disturbing the "viscera" of the middle ear cleft (ossicles, folds, chorda tympani nerve, muscles, and tendons). Irreversibly damaged bone is removed, thus freeing the aditus so that it does not have to permit slow drainage of pus dammed up in the mastoid. The swollen mucosa in the aditus and attic returns to normal, and aeration into the mastoid is resumed.

In chronic diseased states the mucosa is usually irreversibly altered and the bony septa are thickened, reducing the size of the air cell lumens. Nonetheless, if disease obstructing the attic floor is removed and controlled, it may be possible to prevent recurrences of the "attic block" if a permanent passage can be re-established from mesotympanum into the mastoid. The use of Silastic sheeting across bone denuded of mucosa is expected to prevent such a block. If this objective can be established, the otologist may feel free to preserve a posterior bony canal wall. If the "block" recurs in the presence of an intact posterior bony canal wall, the previous disease state will recur. Keratomas, if they recur, may cause another block, or a "block" from adhesions may cause a retraction pocket from indrawing of the drumhead and, eventually, even a recurrent keratoma.

On the other hand, if the posterior bony canal wall is removed as in extensive keratoma, it becomes necessary to remove all the residual mastoid cells. If a cell with its mucosa is overlooked or not removed, it will continue to form a cyst slowly beneath the epithelium lining the mastoid bowl. Many fenestration cavities are afflicted with recurrent "cysts" until the offending cell or cells are removed along with their mucosa. If these cells are numerous, the transudate or exudate from their mucosal linings prevents the growth of skin over the cells, and these mastoid bowls continue to discharge constantly and to form granulations.

Revision in such ears often reveals residual pathology in the mastoid tip cells. It is easy to overlook cells in the mastoid tip, or the surgeon may wish to avoid creating a large inferior pocket in the mastoid bowl. These cells must be removed along with most of the cortical bone of the tip, so that the soft tissues will fall into the space thus created and help to obliterate it. Another source of discharge is in the perilabyrinthine areas. Here one follows diseased cells to their limits. This will cause some degree of skeletonization of the bony labyrinthine capsule, but this is necessary to control continued suppuration in old mastoidectomy cavities. Other troublesome areas are the retrofacial, infralabyrinthine, paracarotid, and subarcuate.

Pneumatization

The mastoid block proper is pneumatized from two sources: the squamous portion of the temporal bone and the petrosal portion of the temporal bone. These areas are separated to some degree by the petrosquamal lamina. The squamous portion is pneumatized by the saccus superior of the primitive endothelium-lined middle ear pouch. It passes through the posterior tympanic isthmus to pneumatize the outer anterior portion of the mastoid block.

The petrosal portion is pneumatized by the medial sac (saccus medius), which passes through the anterior tympanic isthmus to enlarge into the attic and to pass posterior to pneumatize the inner and posterior portions of the mastoid block.

The two separate areas may be completely separated by the petrosquamal lamina (wall of Schwartze and Eysell, or Körner's septum). This is of considerable clinical importance because one area may be diseased and the other normal. If the more superficially placed squamosal portion contains air, one can easily overlook pathology in the deeper petrosal portion. If the petrosquamosal lamina is not recognized and the surgeon seeks the aditus opening, there is great risk of injury to the facial nerve in its pyramidal segment.

Chronic infections occur most often in poorly pneumatized middle ears. Bateman (1952) stated that chronic otitis media is the result of infection in an acellular mastoid. Others hold that failure of pneumatization is the result of infection. Diamant (1954) found on x-ray examination of average individuals that 20 per cent had acellular mastoids. He believed that the degree of pneumatization is determined by hereditary factors.

In the normal infant the mastoid becomes pneumatized. If pneumatization fails as a result of hereditary factors, the bone remains diploic or it may develop into compact (eburnated) bone by the physiologic process of formation of cancellous bone in the marrow spaces.

On the other hand, infection occurring in the pneumatized, diploic, or compact bone will produce sclerosis as a normal response to infection. Sclerotic bone can be grossly distinguished from compact bone by the presence of thick bony septa and small residual spaces filled with edematous mucosa or retained secretions. Marrow spaces are not present.

Pneumatization of the mastoid implies expansion of air-containing and branching endothelial pouches into the mastoid block. This process apparently can begin at about the fifth fetal month of life. Since air is not present in the fetus, the primitive eustachian tube probably permits passage of pulsion waves through the fluid present in its lumen by means of fetal swallowing. This might be considered its active phase. There may also be a passive activity caused by growth of the tympanic ring away from the cartilaginous labyrinthine capsule. This is promoted by a proportionately greater movement outward by the lower portion of the tympanic ring. Growth of the neck, with hyperextension, causes the sternomastoid to pull the mastoid tip downward and slightly outward.

Rüedi (1963) stated that necrotizing otitis media in infants can arrest pneumatization by destroying mucosa and replacing it with connective tissue and, eventually, sclerotic bone. He felt that the sclerosis of the mastoid bone seen in chronic otitis media is the result of

infection and does not in itself cause an infection to develop.

Another factor to consider that may influence pneumatization is eustachian tube obstruction with a resultant intratympanic vacuum (Tumarkin, 1961). This occurs, especially in young children, as a result of infection and enlargement of adenoids. Thorburn (1965) stated that persistent eustachian tube obstruction is not necessarily a common feature of chronic middle ear disease, although intramastoid negative pressure is probably the limiting factor in pneumatization. The intramastoid negative pressure could occur by persistent obstruction of ventilation between the mesotympanum and the epitympanoantral segment without the persistence of the eustachian tube obstruction that may have initiated the lesion. A similar opinion was expressed by Richardson (1963). Proctor (1964) pointed out that the obstruction is at the anterior and posterior tympanic isthmuses.

Pneumatization of the mastoid varies considerably from cells scattered throughout every portion of the temporal bone to a few small cells radiating out from the mastoid antrum. It is useful to systematize the mastoid block for description and study (Portmann, 1951).

Thick Anterior Portion of the Mastoid Block

Superficial Layer. The superficial layer is located behind the external auditory canal. It is subdivided into three segments by two horizontal lines, one passing above the upper third of the auditory meatus and the other immediately below the lower border of the meatus: (1) superficial antral zone; (2) superficial subantral zone; (3) tip.

Deep Layer. The deep layer lies behind the descending facial canal and the posterior tympanic wall and may be divided into two zones: (1) the deep antral zone, which is subjacent to the cranial cavity and follows the region of the semicircular canals; and (2) the deep subantral zone, which is on the lower surface and faces the neck.

Lamellar Posterior Portion of the Mastoid Block

This is the posterior extension approaching the surface.

Sinus Zone. This may be divided into four zones: (1) upper zone or terminal end of the horizontal portions of the sinus; (2) zone at the angle of the sinus; (3) zone of the descending portion of the sinus; and (4) lower or digastric zone corresponding to the jugular bulb.

Cerebellar or Posteroinferior Zone. This is that portion of the mastoid that lies behind the lateral sinus groove; it is crossed obliquely by the emissary vein in the mastoid canal.

Cells may also extend to the following areas:

Squamozygomatic. Pneumatization may begin in the middle ear or antrum and extend up to the squama above the auditory canal and into the zygomatic arch.

Petrosal. Pneumatization begins at the antrum, middle ear, or eustachian tube. A useful classification of these pneumatic tracts has been described by Lindsay (1941) and by Allam and Schuknecht (1968). The latter classify them as follows:

A. Perilabyrinthine
    1. Supralabyrinthine
        a. Posterosuperior
        b. Posteromedial
        c. Subarcuate
    2. Infralabyrinthine
        a. Hypotympanic
        b. Retrofacial
B. Apical
    1. Peritubal
        a. Anterosuperior
        b. Anterolateral
    2. Apical
        a. Hypotympanic
        b. Peritubal
        c. Perilabyrinthine.

Pathology

The pathology of chronic middle ear cleft infections has been well described and illustrated by Schuknecht (1974). The mucous membrane may be thickened by edema, submucosal fibrosis, and infiltration with chronic inflammatory cells. Mucosal edema may progress to the formation of polyps or the mucosa may become ulcerated, and the infection may extend to involve bone and cause the formation of granulation tissue. Progression of the inflammatory changes may produce a rarefying osteitis of the ossicles, otic capsule, and bony trabeculae of the middle ear cleft air cell system. This activity, if unchecked, will eventually lead to fistulization, invasion, and destruction of the inner ear, dural plates, and facial canal, and extend beyond the confines of the temporal bone. Schuknecht (1974) prefers that the more descriptive term of keratoma replace the older term of cholesteatoma, since it refers to the accumulation of exfoliated keratin in the middle ear cleft which arises from keratinizing squamous epithelium that has invaded the cleft from the external auditory canal. The invading squamous epithelium is usually initially confined to pockets by the bony walls and mucosal folds of the middle ear cleft. If the pocket remains dry the rate of exfoliation is slow, and the keratin may accumulate very slowly and insidiously before eroding adjacent structures. If the outer attic wall or the posterosuperior portion of the bony annulus (scutum) are first eroded, the pocket may exteriorize and evacuate the keratin spontaneously for long periods without any further destruction of adjacent middle ear structures; this process is referred to as epidermization. It may be associated with intermittent suppuration but does not in itself necessarily constitute an indication for surgical treatment. If the keratin becomes infected, the collagenase of its squamous epithelium stimulates a rapid bone erosion (Abramson, 1969; Abramson and Gross, 1971). The hypothesis that keratoma may develop by metaplasia of mucous membrane is untenable to Schuknecht (1974). Histologic studies show that mucous membrane can change to pavement epithelium, but there is no support for the concept that it can convert to keratinizing squamous epithelium.

Adults who develop keratoma usually have minimal pneumatization as a result of childhood middle ear infections that had arrested pneumatization. The middle ear cleft is reduced in volume and the structures are in closer approximation, enabling inflammatory adhesive disease to cause the development of a retraction pocket and to slow retention of keratin. Whenever squamous epithelium invades the middle ear cleft in children, there is a rapid extension throughout the attic and antrum and on into the middle ear cleft. This is probably due to the more active growth of skin in childhood, when keratin formation is most rapid (Proctor, 1975).

Sclerosis in the middle ear cleft has been described by Schuknecht (1974) as either (1) fibrous sclerosis or adhesive otitis, (2) fibrocystic sclerosis, or (3) fibro-osseous sclerosis. Fibrous sclerosis is the result of a healing process and is characterized by the proliferation of fibrous tissue in the middle ear cleft. Submucosal fibrosis thickens the mucous membrane, and dense strands of connective tissue may impair ossicular movement and interfere with ventilation of the middle ear cleft. This change may be associated with perforation or retraction of the tympanic membrane, resorption of ossicles due to obliteration of the blood supply, and deposits of hyalinized collagen (tympanosclerosis).

Fibrocystic sclerosis is characterized by partial or total obliteration of the middle ear and mastoid by fibrous tissue and cystic spaces. The cystic spaces develop as areas that have been isolated by proliferation of fibrous tissue. Hyperplasia of the mucous membrane may form many small cystic spaces within a fibrous matrix. The cystic spaces are lined by nonsecreting flat or cuboidal epithelium, and contain a thick protein-containing acidophilic fluid, which may also contain exfoliated cells and cholesterol.

Fibro-osseous sclerosis is a healing process with fibrous tissue proliferation and new bone growth. This rarely occurs in the middle ear but is a common sequel to infection in other portions of the middle ear cleft. Osteoid tissue is deposited on existing bone by osteoblastic activity, leading to the formation of new lamellar bone. The thickened bone trabeculae coalesce and progressively obliterate the intervening spaces. Central fibrous areas remain in the mastoid antrum and central mastoid tract, even with the most advanced osseous sclerosis. During surgical intervention in such cases great care must be exercised in locating the mastoid antrum, since normal surgical landmarks become obscured by the bone laid down in this region. Fibrosis, fibrocystic sclerosis, fibro-osseous sclerosis, and tympanosclerosis may lead to partial or complete obstruction of the eustachian tubal orifice.

Hyalinization (tympanosclerosis) begins as fibroblastic invasion of the submucosa, followed by thickening and fusion of the collagenous fibers into a homogeneous mass to be followed by a deposition of scattered intracellular and extracellular calcium and phosphate crystals. The pathogenesis is not clear, but a prerequisite seems to be chronic otitis media followed by healing. The ear with hyalinized collagen is free of suppuration. White plaques are frequently seen in the tympanic membrane, as are nodular deposits in the submucosa of the middle ear. When severe, much of the middle ear is filled with layers of this material. The underlying bone usually appears normal. Ossicles embedded in it lose their blood supply and become devitalized. The lacunae are empty, although the bony framework remains.

Rarefying osteitis of the ossicles is a common complication of chronic ear infections. The long process of the incus, crural arch of the stapes, body of the incus, and manubrium

are involved, in that order of frequency. Resorption of the long process may result in a fibrous linkage between the long process of the incus and stapes. It is characterized functionally by fluctuating conductive hearing loss. The ossicles may be fixed by fibrous tissue, hyalinized collagen, and new bone growth.

The following classification of otitis media was proposed by Paparella and Juhn (1978):

1. SOM (serous otitis media) - acute and chronic. Nonsuppurative middle ear inflammation with transudation.

2. MOM (mucoid otitis media). Usually chronic. Nonsuppurative. The result of active secretion by secretory cells, goblet cells, and subepithelial glands.

3. POM (purulent otitis media). Suppuration of the middle ear cleft with erythema and bulging of the tympanic membrane.

4. COM (chronic otitis media). An inflammatory process within the middle ear cleft associated with irreversible tissue pathology. It may be active with continuous suppuration or inactive with sequelae of a "burnt out" infection.

A study of 800 sectioned temporal bones by Meyerhoff et al (1978) revealed 333 (41.6 per cent) to have some type of otitis media: 175, POM (52.5 per cent); 123, COM (36.9 per cent); 20, SOM (6.0 per cent); and 15, MOM (4.5 per cent). The 123 temporal bones with chronic otitis media showed chronic activity in 96 ears (78.1 per cent) and chronic inactivity in 27 ears (21.9 per cent). Osteitis was found in every chronic active type, and granulation tissue was present in 93.5 per cent. Granulation tissue is found most commonly in the epitympanum, mastoid air cells, and round window niche. Keratoma was found in only 18 (14.6 per cent) of the 123 COM bones, and tympanosclerosis in 34 (27.6 per cent). This study indicated that chronic otitis media occurs quite frequently, from a histologic standpoint, in the absence of tympanic membrane perforation. Granulation tissue was found much more frequently in chronic otitis media than was keratoma. Granulation tissue is an indication of underlying bone necrosis, usually progressive, which may ultimately produce intracranial complications. This study dramatically pointed out that complications and sequelae of otitis media tend to occur more commonly secondary to the formation of granulation tissue than to keratoma.

Etiology

The causes of chronic otitis media may be complex and may vary from patient to patient. Impaired aeration of the middle ear cleft will, if unrelieved, lead to a chronic middle ear effusion, organization, and various chronic pathologic states. Some of the causes listed by Proctor (1978) are developmental defects such as seen in cleft palate cases; impaired mobility of the pharyngeal end of the eustachian tube (large adenoid, tumors); infections in the adenoid or paranasal sinuses with tubal lymph stasis; allergic swellings of tubal mucosa and tympanic diaphragm; metabolic disorders such as hypothyroidism; collagen diseases with stiffening of tubal ligaments and cartilage; and ciliary paresis of tubal mucosa, as seen with excessive smoking.

Bacteriology

The bacterial flora found in chronic mastoiditis varies considerably. The predominating organisms are usually gram-negative bacilli.

Friedman (1957a) found the bacteriology of chronic otitis media in 1700 patients as follows:

Staphylococcus aureus - 31.7 per cent.
Staphylococcus aureus, penicillin-resistant - 12.9 per cent.
Bacillus proteus - 25.4 per cent.
Pseudomonas pyocyanea - 12.8 per cent.
Mixed - 8.4 per cent.
Escherichia coli - 8.1 per cent.
Streptococcus pyogenes, hemolytic - 7.0 per cent.
Streptococcus pyogenes and *Streptococcus pneumoniae* - 4.6 per cent.
No growth - 10.6 per cent.

Bacillus proteus and *Pseudomonas pyocyanea* are rarely encountered in acute otitis media. In chronic otitis media they are secondary invaders via the external auditory meatus and abide in the moist disintegrating keratin. They disappear when the keratoma is removed. They are not properly controlled by antibiotic treatment even though they may be sensitive in vitro.

It is advisable to culture a chronic aural discharge in order to identify the offending bacterial infection and to determine its sensitivity to the various groups of antibiotics, which may then be selected for specific treatment before and after surgery, if that becomes necessary.

Previous to immunization for control of measles, diphtheria, influenza, and the like, and previous to the use of antibiotics to control the effects of acute infections in the middle ear cleft, there was a great incidence of necrotizing lesions of the mucosa and underlying bone, which ultimately did not heal and which persisted as chronic inflammations of the middle ear cleft. Today we encounter acute respiratory inflammations of a viral nature that render the middle ear cleft susceptible to secondary invasion by bacteria. These infections are usually confined to the mucous membrane of the nasopharynx, eustachian tube, and tympanum and can be considered as tubotympanic inflammations.

An increased portion of our cases of chronic otitis media and mastoiditis are of an entirely different nature. These patients do not experience such an acute and perhaps stormy onset; rather, the disease is insidious, infiltrative, and slowly destructive in its course. The predominant process is one of excessive keratin activity of epithelium that invades the attic and posterior tympanum. McGuckin (1963) stated that this invasive pathologic lesion originates from the meatus and is not in the strict sense a chronic otitis media. However, this migration of meatal epithelium into the attic and antrum can usually be attributed to pathologic states in the middle ear cleft that induce this invasion.

An otitis-prone population with early onset of otitis media, followed by repeated recurrences, often shows a pneumococcal etiology of the initial episode (Howie, 1975). The incidence of these cases may be effectively prevented by the use of a polyvalent pneumococcal vaccine.

Anaerobic bacteria in polymicrobial infections may be an important factor in failure of resolution during the acute phase of otitis media. Brook (1971) found anaerobes in 26 per cent of ear aspirates in children and Jokipii and colleagues (19770 recovered anaerobes in 33 per cent of 70 consecutive cases of active chronic otitis media. Culturing anaerobes is a difficult process and requires highly trained bacteriologic personnel to obtain reliable results.

Studies by Dennis and coworkers (1976) indicated that changes in permeability and vasodilation of the middle ear cleft mucosa can be produced by inflammatory mediators such as histamine, bradykinin, and prostaglandin E_1 and E_2. Such mediators may be the proximal cause of middle ear effusions, and their continued presence may be responsible for the development of chronic otitis media.

Immunochemical and bacteriologic investigations were carried out by Liu and associates (1975). They found bacteria in 77 per cent of effusions by means of a smear. Bacterial recovery rate was inversely related to the dramatic increase with age of IgA and IgM and lysozyme levels in effusions.

Clinical immunology has become an important segment of medical practice. It can give us a precise description of the defense mechanisms that protect us from microbial invasion. Various specific entities may result from deficiencies in one or more components of the immune system and lead to recurrent infections. Problem cases of otitis media can often be referred to the local clinical immunologist for diagnostic support and ongoing consultation.

Tubotympanic Disease

Two groups of chronic middle ear inflammations can be distinguished: one is benign and safe; the other, progressive and dangerous. They may coexist.

The benign form may be termed "tubotympanitis". Two types have been distinguished; they correspond to types I and II of Lillie. Thorburn (1965) describes them as (1) the permanent perforation syndrome; and (2) persistent tubotympanic mucosal infection.

Permanent Perforation

This type consists of a persistent perforation of the tympanic membrane involving the pars tensa. The margins of the perforation are completely covered with a healed epithelium. The ear may be completely dry for long periods or it may discharge intermittently. Such discharge may be caused by infected water passing through the external meatus or may spread up the eustachian tube from nose blowing or sneezing (facilitated by air readily passing outward through the perforation).

In the dry state the tympanic mucosa is pink, and granulations and debris are not present in the tympanum. Plaques of hyalin may be present on the drum remnants and even beneath the tympanic mucosa. When such an ear discharges, the secretions are mucoid or mucopurulent and may be very profuse. Pulsations in the fluid are commonly seen. There is no odor to the discharge. The mucosa in the tympanum becomes red and diffusely edematous.

The degree of hearing loss varies considerably, depending not only on the size and the position of the perforation but also on the degree of fixation of the drum remnant and ossicles, the presence of chain disruption, and the status of the inner ear. If the pathology is confined to a small anterior perforation, the hearing may be normal. Large posterior perforations cause a greater degree of hearing loss. Some patients may hear better when the ear is discharging. This is because of improved phase relationships between the oval and round windows. In dry ears one can reproduce this effect by placing a small prosthesis of oil-soaked cotton into the round window niche.

If the conductive hearing loss is pronounced, the otologist can determine what hearing might be with an intact tympanic membrane. A paper prosthesis is placed to cover the perforation completely, and the hearing is tested. If the perforation is large so that it cannot be covered completely, one can take a fine bamboo rod with a small bit of fused wax at one end and a sound board of cardboard or x-ray film at the other end. The waxed end is placed on the malleus handle, short process, or even incus or stapes, while the patient listens to a steady noise such as running water. If this type of acoustic probe results in marked hearing improvement, it would indicate the likelihood of a favorable result from reconstructive surgery. If hearing is not improved, it may indicate disruption or fixation of the ossicular chain.

Treatment

All debris is removed from the meatus or exposed tympanum with Hartmann forceps, hooks, or fine suction tips. The patient is instructed to avoid getting water into the ear. When the patient is showering, the meatus should be closed with a rubber insert or with a tightly fitted petrolatum-cotton plug.

If the ear is discharging, the secretions can be removed with a fine sterile suction tip. The patient should be warned that the procedure is apt to be very noisy for a few moments so that he will not suddenly jump during this cleaning. It is also well not to leave the suction in place until a caloric stimulus is elicited. Treatment of the nose, paranasal sinuses, or nasopharynx may also be indicated. Allergic factors may also be sought, and eliminated if possible. Smears of the discharge obtained by aspiration may be examined for eosinophils. The material should also be cultured for the type and sensitivity of the infecting organism. After the ear has been cleaned and dried, the specific antibiotic may be dusted into the cavity, or insufflation of Sulzberger powder (boric acid powder with 1% per cent iodine) may be tried. When antibiotic powders are used, one must be wary of the development of eczema due to allergic sensitivity to the antibiotic. Suctioning may not be possible in children and one may then attempt swabbing the ear with a cotton-tipped applicator.

Placement of the appropriate antibiotic through the middle ear cleft may be tried. The patient lies down with the diseased ear directed upward. The meatus is nearly filled with the

antibiotic solution. With the use of a tight-fitting Siegle's pneumatic ear speculum, alternate pressure and vacuum can be made to move the solution through the middle ear cleft into the nasopharynx.

In a patient with a quiescent, and especially an inactive ear, an attempt should usually be made to stimulate closure of the perforation. Debris must be removed from the surface of the drum and from the margins of the perforation with tiny hooks, delicate forceps, and small suction tips. The epithelium at the margin of the perforation must be removed with hooks, curettes, small punch forceps, or chemical cautery. The latter method consists of dipping a fine wire into a concentrated or supersaturated trichloroacetic acid solution and gently applying it to the margins of the perforation; care must be taken to avoid touching the mucosa on the promontory. This procedure can often be done without local anesthesia. Following destruction or removal of the epithelium at the perforation margin, the drumhead is covered with a thin paper prosthesis such as one made from cigarette or rice paper. These can easily be made with a circular paper punch. The paper prosthesis will adhere better if it is soaked in a bit of blood. This is readily obtained by slightly traumatizing the floor of the bony meatus with a needle. The paper disc is applied to the perforation with a delicate forceps of the Hartmann type, or it can be carried down to the perforation and released in proper position with a pipette such as is used to obtain a blood sample for differential and cell studies. This treatment must be repeated at about monthly intervals, since the paper prosthesis is moved off the perforation by migration of the drumhead and canal wall epithelium. This treatment is very effective in a small- or medium-sized perforation (ie, one quarter or less of the drum surface). In some patients the treatment produces recurrent otorrhea, and in such instances a second attempt should not be made for several months after the ear becomes quiescent. If otorrhea recurs a second time, no further treatments of this type are advisable.

If the perforation remains dry permanently or for long periods and hearing is not a problem, the patient may elect to leave the ear alone and not submit to simple treatments for closure of the perforation as just outlined. He should be told, however, about the benefits and drawbacks of reconstructive surgery to close the perforation and to control disease in the middle ear cleft, if the latter is a factor. Thorburn (1965) stressed these points in this regard:

1. Recurrent otitis media should be controlled if reinfection has been occurring mainly via the external meatus. If clinical observations and history indicate that infection has been recurring by the way of the eustachian tube, then otologic surgery should not be done until the cause in the nose or nasopharynx has been corrected. In some of these latter cases, allergies may be a continuous or recurrent problem in spite of the best allergy programs, so that it may never be possible to undertake surgery in the ear.

2. It is difficult to insist that a child never go swimming, and it is nearly impossible to always keep water out of the ears. Rather than to have a constant hazard it is usually far better to close a perforated drum surgically.

3. Recurrent infections will cause further hearing impairment over the years as a result of mucosal changes in the windows and ossicles. These changes may "stiffen" the windows and ossicles or may lead to disruption of the ossicular chain. Likewise, a middle ear exposed to the relatively drier air in the meatus will gradually change to a drier atrophic mucosa. Closure of a perforation will tend to prevent progressive deleterious alterations to the mucosa

in the middle ear cleft.

Loss of hearing is usually restored to some degree by closure of a simple perforation. There are times, however, when healing is associated with more than the expected amount of fibrosis, so that there may be a drop in hearing. This should be explained to the patient before surgery. It should also be understood that more extensive disease than suspected may be encountered at surgery. Occasionally an unsuspected keratoma is found, so that a considerable amount of additional surgery is required. The otologist and patient both should be prepared for this.

Conservative office treatment for closure of a perforation gives excellent results and closes many perforations. If these conservative attempts fail, there may be underlying pathology in the ear that has not been detected. This may occur when hearing is normal. Therefore, at the time of surgical closure of a perforation the middle ear cleft should be carefully explored to rule out concealed pathology in the posterior tympanum, attic, and aditus.

Tubal obstruction should be ruled out preoperatively. If air cannot be passed up the eustachian tube by politzerization or via a catheter, one can try the reverse by politzerizing through the meatus. Very often the latter is successful when the former fails.

4. It is desirable but not essential that the ear have been free of infection for a few months. It should be in a quiescent stage so that the mucosa is least swollen. Distinction between reversible and nonreversible changes in the middle ear cleft is then more easily made. A quiescent mucosa is less readily traumatized. Some of our failures in tympanoplasty are due to excessive trauma to the mucosa, which causes ulceration and subsequent cicatrization and adhesive formation.

5. Age is not an absolute consideration, and each case must be judged on its own. Until puberty the ear is less stable and there is greater risk of tubal infection. In patients over the age of 60, healing is less satisfactory, with more tendency to fibrosis and, as Thorburn (1965) stated, the cochlear function can be very intolerant of even minor operative trauma.

6. With modern techniques prospects of successful long-term healing after closure of a perforation are excellent.

Chronic Tubotympanic Mucositis (Lillie Type II)

As this term implies, the ear presents a longstanding infection characterized by an odorless mucoid or mucopurulent discharge that becomes very profuse and is associated with upper respiratory infections. A large and often near total defect of the tympanic membrane is usually present. Only the limbus may persist. A shortened malleus may be present. In children the perforation may be smaller and more anteriorly placed. The exposed mucosa on the promontory is markedly thickened and red. The exposed ossicles are buried in this thick, exuberant, and edematous mucosa. Polyps may be present as a result of marked swelling of the tympanic mucosa, and these polyps may be associated with necrosis of the ossicles or underlying portions of the temporal bone. They must not be avulsed if they are attached in the posterior tympanum near the oval window. Removal will result in labyrinthitis if the

stapes is involved. X-ray studies of the mastoid usually show a fairly well-pneumatized mastoid air-cell system, with perhaps some degree of clouding of the cells. In some of these cases, after a long period of suppuration, there is an ingrowth of epithelium around the perforation margin and into the posterior tympanum or attic, producing a secondary keratoma. Such a case should then be classified as Lillie type III because of the associated bone necrosis as a result of keratoma activity.

Treatment

There is considerable variation in the bacterial flora and antibiotic sensitivity but, nonetheless, culture and sensitivity studies are recommended. The appropriate antibiotic should be used for a few weeks if possible. If polyps are present they should be carefully removed under general anesthesia with microscopic control. At that time infected adenoids may be removed and antrostomies done if indicated. Intensive allergy investigation and treatment may be required.

In children, surgery to the mastoid should be delayed until puberty, when respiratory infections become less severe and less frequent. It is hoped that with puberty the otorrhea will subside and the ear become quiescent. Only then can tympanoplasty offer any hope of hearing improvement with a dry ear. If surgery to the mastoid becomes necessary, a simple complete postaural mastoidectomy, or perhaps on occasion a modified radical mastoidectomy, should be done. Reconstructive surgery to close the perforation and to improve hearing should be delayed until after puberty.

In adults, if conservative treatment fails to dry persistent disease in the mucosa, then a radical mastoidectomy may be indicated for the following reasons:

1. In unilateral disease, if the patient wishes a dry ear. To control the otorrhea, all the tympanic and protympanic mucosa must be removed. Peritubal cells may require removal before otorrhea can be expected to cease. Infralabyrinthine cells may also need extensive removal. In some cases all offending cells may not be completely removed, especially by less experienced otologic surgeons, so that the patient should not be promised a dry ear.

2. In the presence of a secondary acquired keratoma.

3. When intracranial complications are imminent (labyrinthitis, facial nerve paralysis, and so forth).

All cells throughout the middle ear cleft must be removed in order to remove all epithelium. If the mastoid is large, obliteration with muscle flaps should be done. In these cases there is severe mucosal disease, and reconstructive surgery for preservation or possibly hearing restoration will seldom be indicated; if tried, it will probably fail to conserve hearing and may result in continued otorrhea.

Radium therapy to the nasopharynx and eustachian tube has in the past been used to treat chronic tubotympanic mucositis with some immediate benefit. However, the hazard of radiation therapy is so great that it is no longer recommended for treatment of this problem.

Attic-Antrum Disease

Disease confined to the attic-antrum is due to primary acquired keratoma, to congenital keratoma, or to occlusion of the attic floor by a chronic pathologic process limited to the anterior and posterior tympanic isthmuses.

Primary Acquired Keratoma

This arises spontaneously and insidiously, without a preceding episode of acute otitis media. The perforation is usually confined to the region of the pars flaccida of the tympanic membrane. The characteristic feature is the presence in the middle ear cleft of keratinizing stratified squamous epithelium. The epithelial layer forms a matrix that constantly desquamates sheets of keratin. The accumulation of keratin in a confined space is called keratoma.

The keratin accumulates in concentric, onion-like layers. The layers contain crystals of cholesterol; hence, the name cholesteatoma. Since these are not tumors in the usual sense and since cholesterol crystals are not always present, the term cholesteatosis has been substituted and also epidermosis (Tumarkin, 1961) and keratosis (McGuckin, 1963). The term keratoma as suggested by Schuknecht (1974) is the most appropriate one and should be used instead of the term cholesteatoma. Normally, skin has a thin surface epidermis consisting of a keratinizing stratified squamous epithelium and a thick deep layer, the corium. Skin is constantly flaking off the thin surface layers of keratin. When stratified squamous epithelium is confined to a pocket with limited access, the squames accumulate to form keratoma.

The skin covering the bony external meatus consists of only the thin surface epidermis. It is in direct contact with periosteum. The corium is absent so that there are no glands, hair follicles, or ceruminous glands. The skin of the meatus appears to originate at the umbo. Litton (1968) and also Alberti (1964) have demonstrated how this epithelium radiates out from the umbo and grows outward along the bony canal walls to the skin of the cartilaginous canal, which contains corium and ceruminous glands. Here the keratin that originates at the umbo is finally discarded. Occasionally an otologist observes an attic retraction pocket of considerable depth and with some erosion of the scutum in which the keratin migrates in a stream out of the pocket and adheres to the thin posterosuperior bony canal wall skin. These may remain quiescent for years until activation of inflammation interferes with migration and the perforation is obstructed by a large mass of keratin.

In primary acquired keratoma the eustachian tube and mesotympanum are often normally aerated and free of pathologic changes. In secondary acquired keratoma there is invasion of stratified squamous epithelium through a marginal and occasionally central perforation into the middle ear (mesotympanum). A disease that produces necrosis of the tympanic mucosa (eg, scarlet fever, measles, tuberculosis, acute exacerbations of chronic tubotympanitis) adjacent to the perforation may cause epithelium of the drumhead or meatus to grow over the ulcerated areas.

Congenital Keratoma

A rare form of keratoma can be recognized. It is not associated with the middle ear cleft and is not related to infections in the ear. It originates from the same ectoderm that forms the primitive notochord. Embryonic cell rests from this ectodermal structure may occur in any of the cranial bones. When they occur in the petrous portion of the temporal bone they may spread in and around the labyrinth and extend into the middle ear cleft or cranial cavity. If they become infected, an intracranial complication may quickly occur. The first sign that usually appears is facial paralysis. Examination usually also reveals cochleovestibular paralysis. These keratomas may be of considerable size and may first be detected on radiologic studies. If the ear is trouble-free, no interference is necessary. These keratomas usually lie dormant for years.

In 1963 Cawthorne reported on 13 cases. He referred to Cushing, who in 1922 had expressed the opinion that keratomas may arise from "epidermal rests laid down in the temporal bone during the early formation of the sense organ which it contains. It is not improbable that in many of the recorded cases the keratoma itself was responsible for the otitis media rather than the reverse". McKenzie (1931), Diamant (1952), and Teed (1936) also believe that congenital epidermal rests cause most cases of attic keratoma that occur when a perforation is confined to Shrapnell's membrane. Cawthorne stated that his cases presented a slowly developing facial palsy and that this, together with the x-ray appearance of the petrous bone, serves to differentiate a congenital keratoma (epidermoid) from an acoustic neurinoma. Cawthorne felt that some attic keratomas may be of congenital origin and that they are not always caused by ingrowth of epithelium through a perforation or by invagination of the pars flaccida of the tympanic membrane. Many otologists have seen a whitish mass behind an intact and normal tympanic membrane and on exploration have found keratoma, presumably in the inferior incudal space. If this occurs in the attic it would not be visible on otoscopic examination until later when the pars tensa of the tympanic membrane had eroded. To my knowledge no one has reported a whitish mass in Prussak's space behind an intact pars flaccida. The argument has also been advanced that epithelium may have migrated through a perforation which subsequently closed and that a keratoma later developed in the middle ear cleft. Cawthorne, however, states that in his case the drum was, to use a philatelist's term, "in brilliant mint condition".

Pathogenesis of Keratoma

Migration Theory

Habermann (1889) was the first to demonstrate migration of stratified squamous epithelium from the skin of the meatus through a perforation into the middle ear and the subsequent development of a secondary acquired keratoma. Bezold (1908) stated that the attic retraction type of keratoma is due to eustachian tube occlusion causing retraction of Shrapnell's membrane into Prussak's space and later into the attic. The outer cornifying layer of epithelium desquamates into the retraction pocket thus performed. Wittmaack (1933) showed that persistence of hyperplastic embryonic mucoperiosteum in the attic might cause adhesions to form pockets in various portions of the attic, with the resultant "attic block". This was confirmed by Richardson in 1963. This would have the same effect as occlusion of the eustachian tube. Wittmaack further stated that this hyperplastic mucosa also caused an

arrest of pneumatization of the temporal bone, thus accounting for the frequency with which attic retraction keratomas are seen in small, poorly pneumatized mastoids. Lange (1925) furnished histologic proof of deeply ingrowing prickle cells in the epidermis of Shrapnell's membrane, which was neither retracted nor perforated. He believed that this epithelial proliferation was caused by an inflammatory stimulus. These proliferating columns of basal cells grow into the submucous connective tissue of Prussak's space, thus forming the basis for another type of keratoma.

Rüedie (1963) reported two predisposing factors to the development of acquired keratoma: (10 the special growth potential of the basal cells in the stratum germinativum in the circumscribed zone of the meatal skin adjoining the upper margin of the tympanic membrane, and (2) the submucous connective tissue layer in the middle ear spaces associated with incomplete pneumatization of the preformed spaces.

The development of keratoma is the same for all types of keratoma. There is an *active phase* of growth. In response to the inflammatory stimulus, the epidermoid basal cells penetrate the submucous connective tissue or newly formed granulation tissue within the middle ear, with simultaneous deposition of sclerosing new bone in the submucous connective tissue. Proliferation of basal cells ceases when available connective tissue or granulation tissue, or both, has been used up. The *passive phase* of growth consists of enlargement of the keratoma sac according to the degree of surface desquamation of horny lamellae from the matrix, and the expense of osteoclastic bone destruction.

A second form of migration theory has been advanced by Tumarkin (1961). He emphasized the preceding collapse of a part or the whole of the tympanic membrane (atelectasis), either in Shrapnell's membrane or in the posterosuperior quadrant. This arises, like middle ear effusion, as a result of tubal occlusion and intratympanic vacuum during the course of an upper respiratory infection during childhood. The "threat" of a keratoma occurs only when the epidermal sac accumulates keratin. When this is activated by secondary infection the patient presents with the typical appearance of atticoantral keratoma. Tumarkin suggested the term "epidermosis" instead of keratoma.

A third form of migration theory was advocated by McGuckin (1963). He considered attic-antrum disease as a nonmalignant destructive disease that starts as a silent hyperkeratotic disease originating in the unique skin of the deep bony canal and outer layer of the tympanic membrane. McGuckin believed that intratympanic negative pressure and infection are not important factors in activation of this disease. He emphasized that pressure by keratin (keratoma) is not the cause of bone destruction, but rather that it is caused by chemical or enzymatic factors derived from the breakdown of the keratin.

Keratomas involving the inferior incudal space are common. This is because the area immediately medial to the posterosuperior quadrant of the tympanic membrane is the narrowest part of the tympanum. The posterior pouch of von Tröltsch is relatively large and deep. Relatively mild otitis can inflame this pouch and ulcerate the mucosal lining. Thus, an adhesive obliteration of the pouch occurs, the medial fibrous layer in the postero-superior quadrant of the drumhead atrophies, and a progressive posterior marginal retraction pocket is born.

Metaplasia Theory

In addition to the migration and congenital theories concerning the genesis of keratoma, there is the metaplasia theory, which was first proposed by Wend in 1873. Squamous metaplasia may be provoked by infections, with ulceration and subsequent replacement of mucous membrane by flat pavement epithelium. Physical factors such as excessive dryness may be important. We are familiar with these changes in bronchiectasis, atrophic rhinitis, and healed ulcerations of the septal mucosa. Metaplasia in the mucosa of the middle ear is difficult to prove, but the possibility of this occurring should be kept in mind. Birrell (1958) found squamous metaplasia of the middle ear cleft mucosa in cases of prickle cells. He found prickle cells only when there was a perforation of the tympanic membrane (ie, keratoma due to migration of squamous epithelium from the meatal skin).

Progression of Keratoma

We have discussed the various ways keratomas are believed to originate; we now consider how they progress.

Perforation of Shrapnell's Membrane

The earliest phase is a pouchlike invagination of Shrapnell's membrane which obliterates Prussak's space. Such a retraction pocket may be quiescent for many years. If the epithelium of this pocket is irritated by moisture or infection, the normal movement of the epithelium may be arrested so that squames accumulate. Many of us have observed these pockets with a stream of keratin adhering to the posterosuperior canal wall (McGuckin, 1963). If this keratin does not move out of the retraction pocket, a "plug" of keratin forms. With this stasis of keratin the pocket can enlarge either by erosion of the scutum or by extension of the epithelial sac into another compartment of the middle ear. It is not uncommon to see a quiescent retraction pocket that has eroded the scutum to a point at which the keratin plug no longer obstructs the orifice. This type of spontaneous marsupialization is nature's simplest form of modified radical mastoidectomy, and occasionally this is all that a surgeon need accomplish if he operates on a very early keratoma that is confined to a "dilated" Prussak's space.

It is of considerable benefit to the otologic surgeon to be able to distinguish between the mucosal folds in normal and in diseased conditions. It is a challenge to recognize them when edematous and distorted. These folds carry an important share of the blood supply to the ossicles (Hamberger et al, 1963). By compartmentalizing the middle ear, these various folds may limit disease for a considerable time to one or more compartments. Keratomas have been followed in various sequences of compartments. Examples are from Prussak's space into the superior incudal space, then into the medial incudal space, and then into the anterior compartment or into the antrum. From Prussak's space they may extend into the inferior incudal space and then, via the isthmus tympani posticus, into the antrum (occasionally eroding the short process of the incus), or into the sinus tympani or inferior aspect of the oval window. If the keratoma is contained in its sac, it may be possible to remove the sac in its entirety, preserve the underlying mucosal folds and the "viscera", and exteriorize the involved compartments to prevent recurrence. This is essentially the older concept of a modified radical mastoidectomy. The surgeon should preserve the functioning structures of the middle ear as

much as possible. Reconstruction is better performed from the point at which the disease process can be controlled.

We now consider briefly the problem of chronic middle ear cleft effusion. Blue drums, cholesterol granulomata, and tympanosclerosis may be included in this group. Whenever the aero- and hydrodynamics of the middle ear cleft are interfered with, the exudates and transudates from the mucosal lining of the mastoid air cell system are trapped. We usually think in terms of eustachian tube obstruction. However, attention is called to the significance of the isthmi tympani anticus and posticus. If these small openings from attic into mesotympanum are occluded by swollen mucosa, keratoma, or tympanosclerotic deposits, these exudates will accumulate above the level of obstruction. If the obstruction is correctable by surgery or if it is reversible, then the surgery can be planned for the mastoid air cell system to communicate normally again with the mesotympanum. However, if necessary, new communications can be created in several ways. The tensor fold may be removed, and often with it the tensor tendon, so as to establish a passage directly from the attic into the anterior portion of the mesotympanum. In several patients with persistent effusion, with a normally functioning eustachian tube, the incus body was removed after cutting the long crus free and leaving it attached to the stapes. The drumhead can easily be shifted the short distance to the preserved long crus. By removal of the incus body the fluids normally formed in the mastoid can once again pass into the mesotympanum. If communication is thus re-established, there will not be the need to remove the mastoid cells in these nonsuppurating ears.

Another method for preserving communications between mastoid and mesotympanum is to remove bone from the facial recess (interval between the descending facial and the chorda tympani nerves) and then removed the facial sinus and the chordal ridge which lies between the chordal and pyramidal eminences.

Progression of keratoma is influenced by negative pressure in the tympanum. The earliest sign of this is invagination of Shrapnell's membrane into Prussak's space. When this occurs without retraction of the pars tensa and without evidence of effusion in the tympanum, one can suspect obstruction of the anterior and posterior tympanic isthmuses instead of eustachian tube obstruction. Some of these ear cases also manifest atrophic changes in the posterior portion of the tympanic membrane. These may be due either to a healed perforation or to destruction of the middle fibrous layer of the drumhead by middle ear effusion. During the latter state there is a relative avascularity and anesthesia of the drumhead. These atrophic posterior drumheads will usually adhere to the incus, and later the stapes, often skeletonizing these structures. They also extend into the posterior tympanum and particularly into the sinus tympani. A pneumatic speculum will usually help to distinguish these from a posterior marginal perforation.

Retraction pockets into Prussak's space or posterior tympanum can remain dormant for years, provided the keratin moves out with the skin migration. If migration is interfered with, the keratin will accumulate to form a plug. Moisture reaching the plug will cause swelling and the keratoma begins its active expanding phase. Mucosal folds will stretch before these expanding keratomas and will temporarily relieve pressure on bone. If the epithelium covering an ossicle or the bony tympanic wall is ulcerated, then osteitis and bone absorption occur. This localized osteitis is characterized by the development of granulation tissue. These vascular polyps of the ear strongly indicate the presence of osteitis and keratoma.

Keratomas extending from Prussak's space into the superior incudal space pass over the incus body to the medial incudal space and then spread anteriorly into the anterior compartment of the attic and posteriorly into the antrum. It must always be remembered that Körner's septum (petrosquamosal lamina) may form a bony wall that would limit attic keratoma to that portion of the mastoid derived from the petrosa (above and medial to the septum).

Posterior marginal keratomas and also some from Prussak's space will involve the inferior incudal space and posterior tympanum. They may then pass through the posterior tympanic isthmus into the antrum. Again, if Körner's septum is intact, the keratoma would be placed inferolaterally below the septum. It is important to be aware of Körner's septum in order to avoid injury to the facial nerve; this can occur if Körner's septum is mistaken for the middle fossa dural plate.

As these keratomas progress they continue to discharge, so that there is no increase in pressure and no pain. The bone necrosis is not due to pressure from a keratoma but rather to the presence of bacteria in the keratoma plus possible enzymatic or chemical changes. Progression is also associated in time with obstruction to the ventilation of the antrum and mastoid air cells. These cells continue to form fluid (exudates and transudates) that cannot escape, so they become more concentrated and irritating and lead to the stimulation of cholesterol granulomas or to condensations of the hyalin material, which we now call tympanosclerosis.

Symptoms of Keratomas

Otorrhea. Usually the discharge from keratoma is not profuse. There is almost always a foul odor. In many cases the patient may not be aware of discharge at all, especially in the earlier stages of attic retraction keratoma.

Hearing Loss. This may vary from a normal hearing threshold to total deafness from old serous labyrinthitis. Early retraction pocket keratoma may not impede ossicular movements and hearing may be normal. Occasionally hearing is found near normal in patients with firm keratoma filling an ossicular chain defect. These are the so-called keratoma hearers. Patients very often show surprisingly little concern when the deafness is slowly progressive. Sudden changes in hearing are more likely to bring the patient to the otologist.

Bleeding. This occurs from trauma to granulations or polyps and may alarm the patient.

Earache. Earache is seen with sudden reactivation of a quiescent keratoma and with inflammation of the meatal skin, or it may indicate an extradural abscess.

Dizziness. With progression of keratoma, and particularly with acute aggravations, a serous labyrinthitis may develop either by extension through the windows (especially when granulations are present on the footplate) or by erosion of the bony lateral semicircular canal with fistula formation. Care must be taken that a cerebellar or even cerebral abscess is not overlooked.

Headache. The onset of headaches in the presence of chronic middle ear suppurations is highly suggestive of intracranial extension.

Examination

Interpretation of the status of a diseased middle ear cleft is often difficult. An otologist must be highly motivated to acquire the skill and the wisdom for exact diagnosis. The history must be taken carefully. The examination must be thorough and accurate; often it has to be repeated before a final diagnosis can be made. Both ears must be considered as a unit in many patients, since very often the second ear gives an indication of what has happened to the affected ear.

When the ear is first examined under proper illumination, it often is full of pus and debris that must be thoroughly removed with fine suction tips. At times this debridement must be completed at a second sitting, particularly when crusts are firmly adherent or when the patients are young children or apprehensive adults. The entire drum surface and the bony canal walls must be carefully scrutinized. All findings must be recorded. A sketch of the drumhead should be in the record and the sites of the pathology indicated. At this point, examination with a pneumatic otoscope (Siegle's) is valuable. This magnifies the drumhead two times and permits movement of the drum and ossicles, moves secretions out of the attic or posterior tympanum, and often indicates patency of the eustachian tube. A Hallpike otoscope is even better, and an operating Zeiss microscope is often the best instrument to use for studying the status of the middle ear.

The following factors should be considered and noted on examination of the ears.

Perforation. Its size, exact condition, and shape, along with a careful evaluation of the remaining portions of the tympanic membrane and the external bony canal walls.

Location of Keratoma. The presence and location of granulations, tympanosclerosis, adhesions, and retractions also should be noted.

Discharge. Its amount, character, pulsation, and apparent source.

Patency of Eustachian Tube. Estimation by autoinflation, politzerization, Toynbee's maneuver, catheterization, and inflation, and the reverse by pneumatic pressure increase via the external meatus. The use of a pneumophone such as that devised by Perlman (1939) or the calibrated inflators of Senturia or Zöllner are useful to indicate the amount of pressure required to open the eustachian tube (normal states +20 to +40 mm Hg).

Granulations or Polyps. These must be distinguished from malignant disease deep in the meatal skin and from glomus jugulare tumors. Malignancies are attached to the meatal wall, whereas granulations arise from the middle ear cleft and are not attached to the bony meatus except at the annulus in some cases.

Pneumatic Movements. Movement of the tympanic membrane with the pneumatic speculum will often reveal the presence of middle ear suppuration in an ear that might otherwise appear inactive. It also may indicate whether the ossicular chain is moving or fixed.

It also may differentiate between middle ear suppuration and chronic myringitis or a severe external otitis. After careful removal of all pus and debris from the meatus and drum surface, the pneumatic speculum is applied and the drum moved back and forth. A minute perforation through which secretions or pus are able to pass can be detected in this manner. Sometimes the meatus is badly swollen as a result of external otitis secondary to a chronic otorrhea or quite often to the use of antibiotic ear drops, which produce severe eczematoid swelling. In this situation it may be difficult to examine the tympanic membrane in its entirety. If a small speculum can be passed and the patient gives a positive response to the Rinne test, one can have tentative evidence that the middle ear is free of involvement.

Hearing Tests

These are carried out after all debris and secretions have been removed from the meatus and middle ear. Tests should be repeated to note the effect of treatment (ie, diminished discharge, reduced edema of mucosa, removal of granulations and accessible keratoma), and also after inflations and after treatments to the nose and nasopharynx, including allergic management. Air and bone conduction tests must be done and the speech reception threshold recorded. The bone conduction test must be carefully evaluated because many of these patients with chronic ear disease have cochlear loss of hearing in addition to the conductive loss. Cochlear hearing loss may be severe enough to prohibit reconstructive surgery for hearing restoration. The surgery in such cases should be directed only to controlling the suppuration and avoiding subsequent intracranial extension. Since most cases of chronic middle ear suppuration are unilateral, it is important that proper masking techniques be used before the hearing status in the afflicted ear is finally evaluated. Confirmation of the audiologist's findings can often be made with the use of the Bárány noise box and tuning forks.

The pure tone air and bone audiograms, the speech reception threshold, and the tuning form tests (Rinne, Weber, and Schwabach) all indicate a conductive type of hearing loss in chronic middle ear disease without loss of cochlear function.

Further information concerning hearing potential can be obtained with the use of an acoustic probe. The delicate Pohlman probe is far superior for evaluation of potential hearing than use of the heavier metal probes to which a bone conductor oscillator has been affixed (Pohlman and Kranz, 1928). The latter are cumbersome to use and distressing to the patient.

Proper hearing evaluation is essential if surgery for control of disease is to be coupled with an attempt at improvement of hearing (tympanoplasty). If hearing is at or near normal, particular pains must be taken to control disease and to prevent an inevitable progression of hearing loss without jeopardizing the existing hearing. It is often possible, for instance, to exteriorize a compartment such as the entire attic, remove the disease, control progression, and avoid subsequent worsening of the hearing. This has been accomplished by otologists for many years with the modified radical operations of Bondy and Heath. The tympanoplasty of today is an extension of these operations made possible by microsurgical techniques. We can now close perforations and reconstruct diseased ossicular chains with a reasonable chance of success provided the middle ear disease has been permanently arrested. When hearing is at or near normal and the ossicular chain and pars tensa are apparently free of disease, surgery is directed to sealing off the mesotympanum from the exteriorized attic and mastoid.

If the hearing tests indicate that conductive hearing loss is complete and that cochlear function is good, the otologic surgeon can proceed with the application of a complete array of techniques to attempt hearing improvement along with control of the middle ear cleft disease.

Bacteriology

Culture and sensitivity studies must be made in all patients with threatening intracranial extension and in patients with an acute exacerbation. These studies are advisable immediately before a surgical intervention so that the proper antibiotic can be used in the immediate postoperative period. The ears are commonly infected with *Bacillus proteus* and *Pseudomonas pyocyanea,* but successful eradication is dependent on elimination of the keratoma that harbors these organisms rather than on the use of specific antibiotics.

Radiology

Routine x-ray studies of the temporal bone are advisable even though they are of limited use after a thorough clinical examination. They will, however, show the degree of pneumatization of the mastoid and petrosa as well as indicate the possible extent of bone erosion.

More information concerning the status of the ossicle and the extent of bone erosion can be obtained from linear tomography of the temporal bone. Even better is the use of the polytome devised by Portmann and Guillen (1959). By this method the dense bone associated with long-standing middle ear cleft disease can be eliminated in part by focusing on the posterior tympanum and ossicular chain. This additional information requires the use of highly sophisticated and expensive equipment, more time for examining the patient, and an adequately trained radiologist.

Treatment

The treatment of attic-drum disease varies considerably and depends on the degree of activity of the disease. Patients with a dry retraction pocket in whom epithelial migration evacuates the pocket can be observed for long periods of time. If keratin debris collects in a retraction pocket it is often useful to irrigate the pocket with 95 per cent alcohol, using a fine attic cannula of the Day type. This method is particularly useful in aged patients, in patients refusing surgery, and so forth. These irrigations are repeated at frequent intervals until the retraction pocket remains free of accumulated keratin. Ear drops are used only with the development of external otitis. Again, the use of 95 per cent alcohol with 2 or 3 per cent salicylic acid helps to dry the ear and to dissolve the keratin. The use of antibiotic ear drops may cause aggravation. Drying powders such as Sulzberger powder (boric acid with 1 per cent iodine) may be tried. If granulations are present, they are carefully removed except when they appear to arise from the window niches. Their base may be cauterized either with 10 per cent silver nitrate or with carefully applied minute amounts of trichloroacetic acid.

The otologist must explain the status of the ear to the patient. If the disease is potentially dangerous, the patient must understand that the diseased ear might suddenly worsen and actually become dangerous and threaten intracranial extension. The patient should

be told the prognosis for hearing loss or improvement.

If keratin continues to accumulate in a retraction pocket in spite of suction treatments, mechanical removal of keratin plugs, and attic irrigations with 95 per cent alcohol, and if there are no serious contraindications for surgery, it is safer to control the disease surgically before the middle ear cleft becomes further diseased and the hearing worsens. Modern otologic surgery is not associated with disturbances of general body physiology, it is precise under magnification, and it gives excellent results in the early stages of middle ear disease. There is little or no excuse for following a policy of "watchful waiting" and leaving the patient in such a potentially dangerous condition.

Some of the benefits of the surgery should be explained in detail:

1. Surgery is done to prevent intracranial complications.

2. Prevention of further hearing loss is very likely possible.

3. The ear will become dry in most instances; at least the fetid odor will be eliminated. The constant use of cotton in the meatus is avoided.

4. Hearing can sometimes be gained by tympanoplasty procedures in one or two stages provided the ear is reasonably free of advanced middle ear cleft disease. The patient must understand that surgery must first be directed to control of the disease.

5. A dry or nearly dry ear may sometimes be desirable so that the patient may be better able to wear a hearing aid. Surgery in such patients is confined to the attic and mastoid so as to avoid risk of further hearing loss by surgery too close to the windows (particularly in children and in the aged).

It is a different matter if a retraction pocket remains free of keratin, but even then the patient should have the ear examined by an otologist at least yearly. Water must be kept out of the ear and the patient must be instructed to return immediately if otorrhea recurs, deafness worsens, or bleeding ensues or if vertigo, pain, or headache develops.

Late Middle Ear Sequelae

Adhesive Otitis Media

Adhesive otitis media has been defined by Cawthorne (1956) as "a long-standing adhesive process as the result of inflammations". In 1953 Ojala stated that chronic adhesive otitis, instead of manifest chronic suppurative otitis media, will become the most common inflammatory disease of the middle ear leading to serious deterioration of hearing. The use of antibiotics in treatment of suppurative otitis media has sterilized the middle ear cleft but has not prevented the inflammatory changes, which, if repetitive, may ultimately result in a chronic adhesive process. We must recognize that deafness resulting from chronic adhesive otitis is a serious consequence of inadequate antibiotic therapy for acute suppurative otitis media. If hearing does not return quickly to normal after adequate treatment, a myringotomy with tube insertion is often indicated; failing that, a complete mastoidectomy with exploration

to correct an attic block should be seriously considered.

The disease may start at any age but its usual onset is during childhood. There may be a hereditary tendency. The history may further reveal previous bouts of middle ear inflammation that were inadequately treated or perhaps not treated at all; in particular, myringotomy was not done. Suppuration in the paranasal sinuses is very common, as is enlarged and inflamed adenoid tissue. Uncontrolled allergic states are frequently seen. The tympanic membrane is intact but pathologic, with atrophic changes and retraction of a variable degree. The audiogram shows a conductive type of hearing loss not unlike that caused by otosclerosis. However, there are often cochlear hearing deficits at an early age, probably as a result of biochemical activity at the permeable round window membrane (serous labyrinthitis). The hearing loss progresses, sometimes rapidly, until in a few cases the patient becomes completely deaf. In some, of course, the disease is arrested in an early phase, and hearing may remain near normal. Frequently there is tinnitus and sometimes vertigo.

Chronic adhesive otitis has been divided into two groups: the one developing after a long period of suppuration, and the other after chronic catarrhal changes. Lumio (1951) reported that in nearly half of his 185 patients there was a history of former ear trouble with earache or discharge. The other half probably had non-suppurative otitis media with effusion that had not been noticed. This fact points out the importance of repeated hearing tests for school-aged children. Care must be exercised in distinguishing chronic adhesive otitis from otosclerosis. Lumio (1951) reported the differential characteristics as shown in Table 1.

Table 1. Differential Diagnosis of Chronic Adhesive Otitis Versus Otosclerosis

Symptoms	**Chronic Adhesive Otitis**	**Otosclerosis**
Familial deafness	Seldom	Often
Age of onset	Any age	Usually 20-30 years of age
Acute otitis	Frequent	Infrequent
Hearing during pregnancy	Unaffected	Worse
Hearing in noise	Not better	Better
Head colds, nasal obstruction	Frequent	Infrequent
Inflatability	Difficult	Often normal
Tympanic membranes	Changes	Nearly normal
Audiogram	Impaired bone conduction at an early age	Impaired bone conduction later
Mastoid pneumatization	Arrested or absent	Frequently extensive.

The reported incidence of chronic adhesive otitis varies considerably. MacNaughton (1956) reported an incidence of about 30 per cent in a hearing aid clinic in Aberdeen, Scotland, whereas Cawthorne (1956) found only 22 cases in 1541 patients seen for deafness during the three years 1953 to 1955. If otosclerosis and perforations are carefully excluded and if the earlier, reversible middle ear inflammations are also excluded so that we diagnose the terminal fibrotic state as this disease, then the incidence is much lower. Pathologic drums (scars, hyalin deposits) with normal hearing should not be included. Perlman (1943) stated

that fibrous tissue bands in the tympanum may be compatible with normal hearing.

In a study of 185 cases, Lumio classified the tympanic membrane changes as shown in Table 2. Cawthorne (19560 stated that the diagnosis is most difficult when the tympanic membrane is uniformly opaque (milky drum). These drums are usually immobile. Absence of movement of the drum can be detected with Siegle's pneumatic otoscope. It is well to call attention at this point to the "fixed malleus syndrome" (Goodhill, 1966), which may be diagnosed only at a surgical intervention for a tentatively diagnosed case of otosclerosis. The fixed malleus may be congenital or acquired.

Pathogenesis of chronic adhesive otitis was discussed by Ojala (1953), who studied 116 histologically sectioned temporal bones of patients with this disorder. He made the following conclusions concerning organization of exudate and pathogenesis of adhesive otitis:

1. The inflammatory changes are irreversible (indicated by the presence of fibrin in the exudate).

2. The subsequent proliferation of connective tissue is rapid, depending on host resistance and virulence of the infectious agent.

3. Drainage is impeded by the drum's rupturing late or not at all.

If the inflammation subsides rapidly and the conditions are favorable for repneumatization, the organized tissue usually atrophies and contracts without affecting the hearing. If the inflammation is protracted or repetitive, the organized tissue in the middle ear becomes denser and forms permanent adhesions. After the inflammation subsides there are regressive changes which finally become stationary.

The treatment of established chronic adhesive otitis poses many difficult problems. When the disorder is bilateral a hearing aid provides satisfactory hearing for many of these patients. The history of this disorder reviewed by Siirala (1964) reveals many attempts at surgical intervention since the seventeenth century. In recent years fenestration has been tried with relatively poor results. Since the advent of microsurgical techniques with tympanoplasty there is some hope for relief of hearing impairment in a few of these patients. The objectives are to free the mesotympanum, including the window niches of adhesions, and to prevent the recurrence of the otitis. An air-filled mesotympanum must be created. It may be possible to repair or to restore a functioning sound conduction system in some of these patients. Prevention of recurrence of adhesions may be accomplished by the insertion of Silastic or similar sheeting in the tympanum. The use of proteolytic enzymes (alpha-chymotrypsin) may also help to prevent recurrences of adhesion during the healing stage (Siirala, 1968). The reversibility of chronic adhesive otitis was accomplished in some cases by Buckingham and Ferrer (1966) by inserting a polyethylene tube through the anterior segment of the collapsed tympanic membrane.

Patients should understand the limits of surgery in solving this problem. They are also subject to further neural losses from surgery at the windows. Prevention of chronic adhesive otitis is, at the present time, the best means of approaching this problem. This involves more intensive and longer use of antibiotics during acute suppurative otitis media, earlier and more

frequent myringotomies, and the performance of a complete mastoidectomy with exploratory tympanotomy earlier in the course of the disease. Particular attention should be paid to overcoming obstructions in the attic floor.

Tympanosclerosis

The term *tympanosclerosis* was originally used to describe sclerotic changes of the middle ear mucosa (von Tröltsch, 1873). The word was revived by Zöllner (9156, 1963) to indicate hyalin changes in the mucosa. These changes come about as a result of acute inflammations in the middle ear cleft that are prolonged or recur frequently. These consist of an invasion by fibroblasts with organization of the subepithelial tunica propria into a thick connective tissue rich with collagen. Hyalin degeneration follows, with a permanent thick, whitish plaque deposited submucosally. The disease is not primarily associated with destructive mucosal or bone lesions. These lesions may calcify or they may be replaced in part by new bone formation. They are commonly seen to a variable extent in patients operated upon for chronic otitis media. Sheehy and House (1962) found 75 such plaques in a series of 227 consecutive patients operated upon for chronic otitis media. They found no differences between this group and the 152 patients without tympanosclerosis with regard to age, type of perforation, presence of keratoma, or the results of surgery. Tympanosclerosis can occur with intact tympanic membranes. In that event it is usually bilateral and is associated with allergic states.

In the management of this disease at surgery, the plaques need be removed only from the vital areas (ossicles, joints, tendons, windows) where its presence may interfere with acoustic function. The plaques can be carefully elevated (sometimes in layers) with a fine needle. Care must be taken to preserve as much mucosa as possible to prevent subsequent adhesions. In those patients with massive plaques around the ossicles, it is idle to hope that refixation would not recur. In such patients incus replacement prosthesis or some type of ossicular reconstruction should be seriously considered. If the footplate is fixed, mobilization may be tried; if unsuccessful, a second stage stapedectomy should be done. It is generally agreed that stapedectomy should never be done at the same time as tympanoplasty or myringoplasty.

Cholesterol Granuloma

A clinical diagnosis of cholesterol granuloma can be made when the tympanic membrane has a dark blue color as a result of the retention of a dark fluid in the tympanum. On operation there is found a granulomatous mass in the region of the ossicular chain and its mesenteries or in the posterior tympanic sinuses. On microscopic examination the granulomatous tissue shows cholesterol crystals surrounded by foreign body giant cells. Cholesterol granuloma is a nonspecific entity occurring in any type of chronic otitis media in which there is stasis with mucosal edema, exudation, and hemorrhage. It seems to occur most frequently in relation to the blood vessels to the ossicles and along the facial canal and particularly in the facial sinus. This may account for its local exuberance.

In a short space an attempt has been made to cover the subject of chronic otitis media and mastoiditis. Much is still unknown about the inflammatory diseases in the middle ear cleft. Very little is known concerning the physiology of the eustachian tube, and our

knowledge of pathology in this area is far from complete. Effusion, cholesterol granulomas, and the nature and behavior of keratin are incompletely understood.

The ramification of the fascinating changes that are observed in the middle ear were discussed at a seminar in Detroit in 1966. The "mucosal" factor indicates disease of the lining of the middle ear cleft, associated with dysfunction of the pressure-equalizing mechanism and with retention of exudates which would normally escape into the nasopharynx. The "squamous" factor means that there is or has been failure of the escalator mechanism (or migration) within the external auditory canal. Retention of the products of desquamation is accepted as a cause of dissolution of the underlying tissues and particularly of bone. The mucosal factors seem to predominate in childhood. Some such system is invaluable as an aid for clarification of the course that any given diseased ear may take, with or without medical or surgical treatment.