Diseases of the Ear

a textbook of otology

Fourth Edition

Stuart R. Mawson
Harold Ludman

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Preface to the Fourth Edition

The philosophy on which former editions of this work was founded has not changed in planning this fourth edition. It embodies the conviction that there is still need for a work that contains within one volume all the fundamental otological knowledge required by a postgraduate student studying for higher qualifying examinations. In this new edition, the two authors have sought to retain the methodicity, uniformity of style, and balance between priorities, which were favourably noted in its predecessors.

During the four years since the preparation of the last edition there has been considerable extension of the applications of the basic sciences to the practices of audiology and of neuro-otology; and now the otologist in training is expected to acquire, and to present to his examiners, more understanding of these basic sciences, and in particular the principles of audiology, than formerly; this is as it should be.

The basic anatomical section has been shortened to exclude much of the formal anatomical description, which is available elsewhere, and it has been modified to emphasize the practical surgical anatomy—applied anatomy. The section on physiology has been reorganized to accommodate a more detailed presentation of acoustics, and of recently acquired knowledge of cochlear and vestibular function. The chapters on diseases of the external, and especially of the middle, ear cover matters where there has been gradual consolidation of ideas and of opinions. These have been presented to provide a balanced and authoritative view of the present state of otological opinion. The section on diseases of the inner ear deals with a fast growing aspect of the speciality: it has been replanned and largely rewritten to present an up-to-date account of these advances, and of the new required knowledge in the fields of neuro-otology, sensorineural deafness, and diseases affecting structures within the internal acoustic meatus. Lastly, after the exuberant developments of surgical technique which characterized the trend of otology in the 1960s, the section on operative surgery has been somewhat pruned. It is not intended, nor would it be desirable, that this section should guide trainees through each step of every operation.

Some of the old line drawings have been redrawn, and others added, by the co-author and previous illustrator, together with new photographs to embellish the text.

This edition initiates a change from the traditional form of bibliography. We have felt that the postgraduate student, to whom the work is principally addressed, would not, at his stage of reading, wish to pursue references that might give him more detail, in a given instance, than would be needed for the purposes of examination. Nor does it now seem required, in this work, to offer, by reference, justification for every statement. Rather, we have elected to provide a general bibliography to broaden and fill in the various sections where necessary. The selection for this bibliography—or recommended further reading—has been from papers published within the past few years, review articles and other textual authorities, and a small number of earlier publications seminal to the subject.

We wish to acknowledge, with gratitude, the valuable assistance of Dr John M. Dawson, F.R.CP., F.R.C.R., Consultant Neuroradiologist at King's College Hospital and the Guy's-King's-Maudsley Neurosurgical Unit, in preparing those sections devoted to radiological examination, and for providing radiographs for reproduction.

We owe special thanks to Miss Gillian Pentelow, A.L.A., Chief Librarian of King's College Hospital Medical School, for her enthusiastic help in tracing and providing relevant publications for the authors. We are also most grateful to our secretaries for their unflagging forbearance and support, and to our wives and families for their tolerance.

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Jucundi acti labores

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Part 1
Introductory

1 Prenatal and postnatal development of the ear

Prenatal development

The components of the ear are derived from all three embryonic primordial layers, ectoderm, mesoderm, and entoderm, and from these three primitive layers the aural derivatives arise as follows (Table 1.1).

Table 1.1

Ectoderm	Mesoderm	Entoderm
External ear Auricle, external acoustic meatus, skin, glands	Middle ear* Upper half, ossicles and mastoid	Middle ear Lower half
Tympanic membrane Outer layer	Tympanic membrane Middle layer	Tympanic membrane Inner layer
Membranous labyrinth Organ of Corti, utricle, saccule, semicircular canals	Bony labyrinth Petrous temporal bone	Auditory (Eustachian) tube

*Some authorities consider the mucosa of all air spaces is derived from entoderm.

Since the ear develops in close relation to the dorsal ends of the first and second branchial arches, these must next be considered (Fig. 1.1). Development begins at 3 weeks with the expansion of the cephalic part of the foregut, an entodermal derivative lying dorsal to the pericardium, to form the primitive pharynx, and is continued by the formation of thirteen depressions in the walls of the pharynx, five bilaterally represented pouches, the rudimentary thyroid, the respiratory tract and the impermanent pouch of Siessel. The entodermal lining of the pouches is separated from surface ectoderm by the secondary mesoderm. On the external aspect of the embryo, opposite the pouches, four grooves or clefts appear in the ectoderm, dividing the intervening side wall of the pharynx into bars or arches. These bars are numbered from the cephalic end, and, as growth proceeds, bars 1 and 2 meet across the midline between the pericardium and the head. Bar 1 persists as the mandible, bar 2 as the hyoid, but bars 3, 4 and 5 remain small and unjoined and soon become overlain and recessed behind a caudal expansion of the second, hyoid, bar. This recess, known as the cervical sinus, is the nursery of branchial cysts and fistulae (Fig. 1.2).

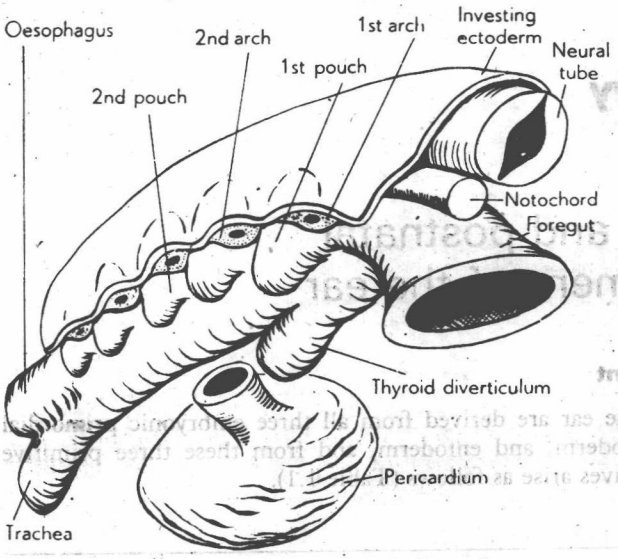

Fig. 1.1. Front end of embryo to show pharyngeal pouches.

Thus the first external groove or cleft is the only one not submerged beneath or overlain by the second bar or arch, and is the primordium of the external acoustic meatus. At the same time, the internal aspect of the second bar forms part of the wall of a lateral extension of the upper part of the cavity of the primitive pharynx known as the tubotympanic recess. This recess also contains the first and third pharyngeal pouches. The tympanic membrane, which

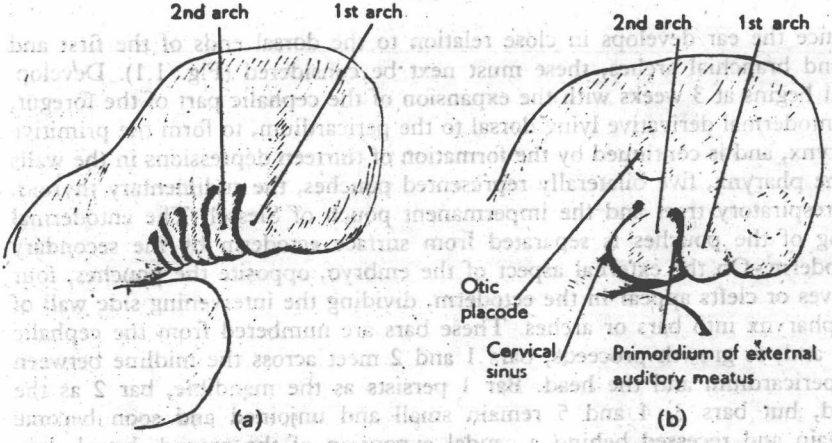

Fig. 1.2. Development of the cervical sinus; (*b*) shows overgrowth of second arch caudally.

separates the tympanic cavity from the external acoustic meatus, is formed in the position of a separating membrane between the first pouch in the tubotympanic recess and the first external groove. But the membrane is a secondary formation since the deeper part of the meatus is first formed by a solid plug of ectodermal cells which later break down in the centre to form the meatus (Fig. 1.3).

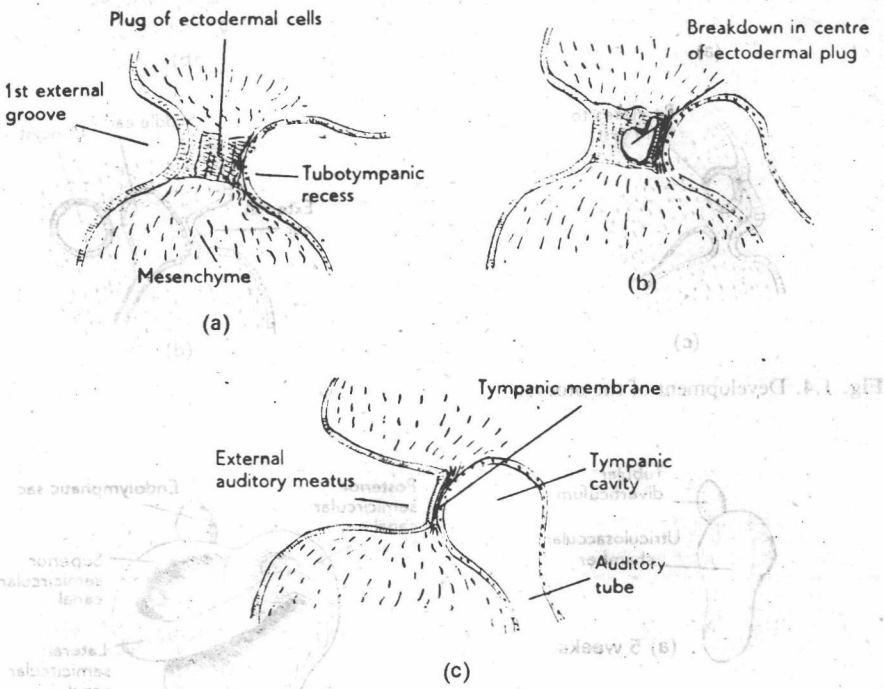

Fig. 1.3. Development of tympanic membrane.

The third bar or arch, meanwhile, although overlain by the second, continues to grow forward under it, and succeeds in narrowing the inner part of the tubotympanic recess to form the auditory tube region, while the outer part of the recess develops into the tympanic cavity. The subsequent development of the ear may now be considered in detail.

Internal ear

At 3 weeks a thickening of ectoderm, the auditory (otic) placode, appears opposite the hind brain immediately above the first external pharyngeal groove. By invagination the placode first becomes a pit, then a closed pit. Sinking beneath the surface ectoderm the auditory vesicle (otocyst), as it is now called, comes to rest in mesoderm. With the intervention of the tubotympanic recess between it and the surface ectoderm, the ultimate gross relations of the three parts of the ear are thus established (Fig. 1.4).

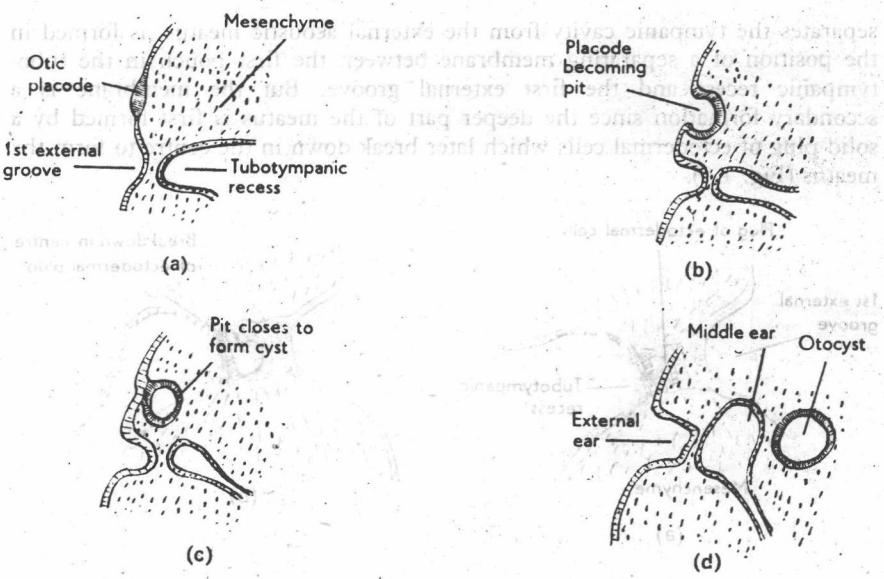

Fig. 1.4. Development of the otocyst.

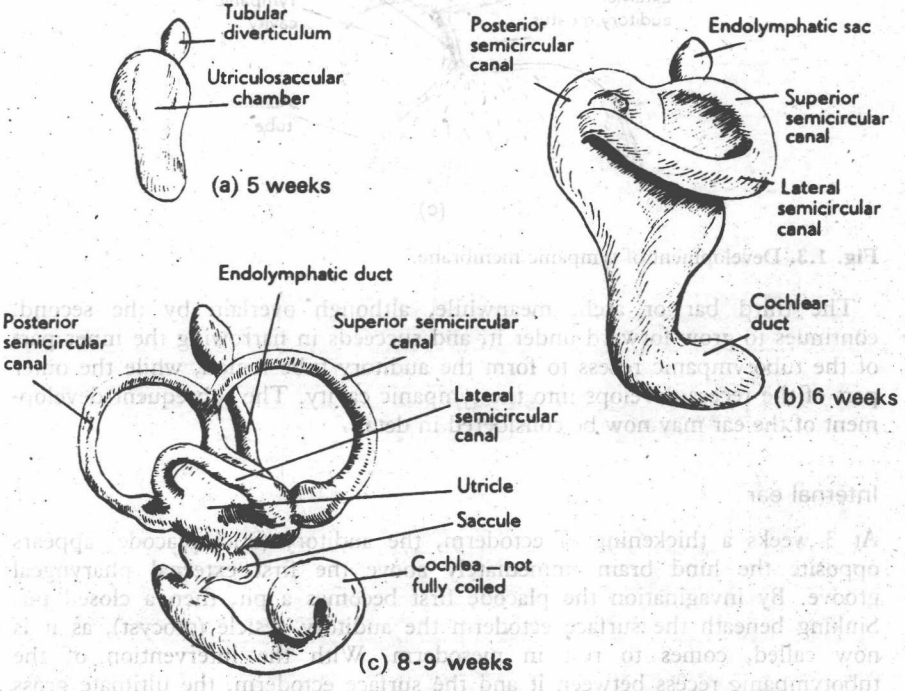

Fig. 1.5. Development of membranous labyrinth (*a*) 5 weeks, (*b*) 6 weeks, (*c*) 8-9 weeks.

The vesicle elongates and develops an indenting groove on its medial side which demarcates a tubular diverticulum, soon further modified into the endolymphatic sac and duct. The major compartment of the vesicle, termed the ultriculosaccular chamber, differentiates into semicircular canals and cochlea (Fig. 1.5). From the dorsal part of the chamber three disc-like evaginations appear in the right-angled disposition of the canals, acquire a corpuscular shape and, with further absorption of their centres, by 9 weeks become free semicircles. From the ventral part of the chamber a single evagination pushes medially as the cochlear duct and by 12 weeks it too, fully coiled, achieves definition. The remaining central portion of the utriculosaccular chamber represents the membranous vestibule, with the utricle developing dorsally and the saccule ventrally. At 8 weeks the mesoderm investing the differentiating auditory vesicle shows signs of conversion into a cartilaginous capsule, later to become ossified. In this cartilage the scala tympani and scala vestibuli gradually extend along each side of the cochlear duct until, when the tip is reached, they approximate and fuse with the formation of an opening, the helicotrema (Fig. 1.6). Ossification of the cartilaginous capsule begins and is generally completed between the 16th and 23rd week.

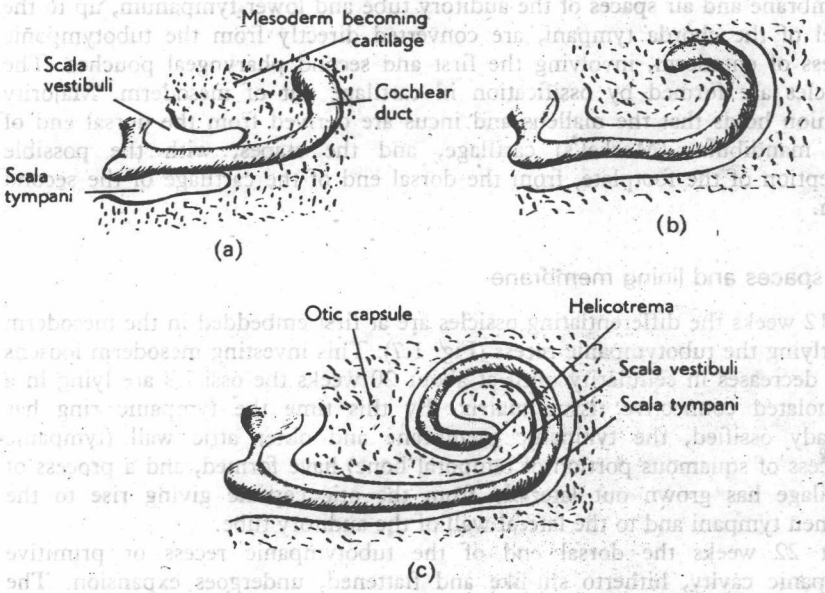

Fig. 1.6. Diagrammatic representation of the development of the scala tympani and scala vestibuli.

Fissula ante fenestram

Between the cartilaginous otic capsule and the membranous labyrinth, the mesoderm differentiates into a mesh-like connective tissue constituting the periotic space or periotic labyrinth. Immediately in front of the oval window,

where the vestibule unites with the scala vestibuli, a streak appears in the cartilage of the otic capsule which bears a close resemblance to the young mesh-like periotic tissue. This streak seems to develop in the direction of the middle ear as an extension of the tissue lining the vestibule, in other words as an evagination of periotic tissue into the capsule. At the same time, when the streak has reached the middle ear it is in turn invaded by a vascular bud of middle ear epithelium which hastens the absorption of cartilage and growth of the streak. From the 16th to 20th week, the period during which cochlea and vestibule are attaining their full dimensions, the fissula is extending until at the end of this period it too has reached maximum size. In the adult the fissula appears as a fibrous slot in the bone. Its importance lies in that, in immediate proximity to the stapedial footplate, it forms a site of predilection for the formation of abnormal, otosclerotic bone.

Middle ear

In the development of the middle ear there are three components to be considered: the air spaces, the lining membrane, and the ossicles. The lining membrane and air spaces of the auditory tube and lower tympanum, up to the level of the chorda tympani, are converted directly from the tubotympanic recess of entoderm, involving the first and second pharyngeal pouches. The ossicles are formed by ossification in cartilage out of mesoderm. Majority opinion holds that the malleus and incus are derived from the dorsal end of the mandibular (Meckel's) cartilage, and the stapes, with the possible exception of the footplate, from the dorsal end of the cartilage of the second arch.

Air spaces and lining membrane

At 12 weeks the differentiating ossicles are at first embedded in the mesoderm overlying the tubotympanic recess (Fig. 1.7). This investing mesoderm loosens and decreases in cellularity until at about 20 weeks the ossicles are lying in a vacuolated connective tissue matrix. By this time the tympanic ring has already ossified, the tympanic membrane and outer attic wall (tympanic process of squamous portion of temporal bone) have formed, and a process of cartilage has grown out laterally from the otic capsule giving rise to the tegmen tympani and to the lateral wall of the auditory tube.

At 22 weeks the dorsal end of the tubotympanic recess or primitive tympanic cavity, hitherto slit-like and flattened, undergoes expansion. The lining epithelial membrane is believed to enter and invade the vacuolated mesoderm and to wrap itself round the ossicles, carrying in potential space of the tubotympanic recess. The subsequent existence of mucosal folds is attributed to a four-pronged invasion of the tubotympanic process by pseudopodial buds or sacs, the folds being formed by the party walls between the buds. This vital process of tympanic expansion by active encroachment of tubotympanic (entodermal) cells into preloosened and vacuolated mesoderm is accepted as the basis for hypotheses attempting to explain lack of mastoid pneumatization on the grounds of either hereditary failure in this active process, or of inter-

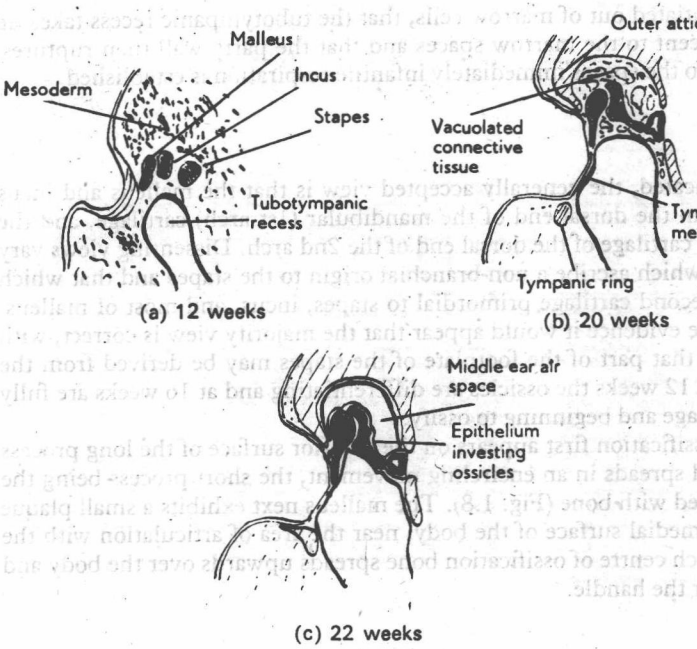

Fig. 1.7. Development of middle ear.

ference with it by infective or other pathological changes. At 29 weeks the otic capsule is well ossified, the middle ear is mostly pneumatized* and the epithelium is extending towards the region of the antrum which is still occupied by the epitympanic connective tissue. The epithelium of the middle ear is then described as reaching the mouth of the antrum and by 34 weeks as having half pneumatized it. Thereafter the pneumatized antrum extends into the capsular bone to form antrum air cells, and laterally into the growing tympanic process of the squamous bone to form air cells in the region of the primitive mastoid process. The concept is firmly one of continuous active expansion and invasion by epithelium cells originating from the primitive tympanum and continuing, in the case of the mastoid air cells, through infancy and childhood. This view of pneumatization indicates the position of present day orthodoxoy. It cannot be denied, however, that this concept of entodermal invasion leaves important questions unanswered, chiefly the histological difference between the epithelium lining the auditory tube and lower part of the tympanic cavity, and the epithelium lining the epitympanum, antrum and mastoid air cells; a difference that is strikingly emphasized by markedly dissimilar response to infection by the two epithelial areas. The process of pneumatization has also been conceived as accomplished primarily in mesoderm, that the vacuolated spaces appearing in the temporal bone simply represent bone marrow spaces similar to spaces in other bones, that these spaces acquire a lining

*Pneumatization here means formation of epithelial lined potential space.

of cells differentiated out of marrow cells, that the tubotympanic recess takes up a position adjacent to the marrow spaces and that the party wall then ruptures, allowing air into the spaces immediately infantile respiration is established.

The ossicles

As already indicated, the generally accepted view is that the malleus and incus are derived from the dorsal end of the mandibular (1st arch) cartilage, and the stapes from the cartilage of the dorsal end of the 2nd arch. Dissenting views vary between those which ascribe a non-branchial origin to the stapes and that which considers the second cartilage primordial to stapes, incus, and most of malleus. On the available evidence it would appear that the majority view is correct, with the reservation that part of the footplate of the stapes may be derived from the otic capsule. At 12 weeks the ossicles are differentiating and at 16 weeks are fully formed in cartilage and beginning to ossify.

A centre of ossification first appears on the anterior surface of the long process of the incus and spreads in an encircling movement, the short process being the last to be invested with bone (Fig. 1.8). The malleus next exhibits a small plaque of bone on the medial surface of the body, near the area of articulation with the incus, from which centre of ossification bone spreads upwards over the body and downwards over the handle.

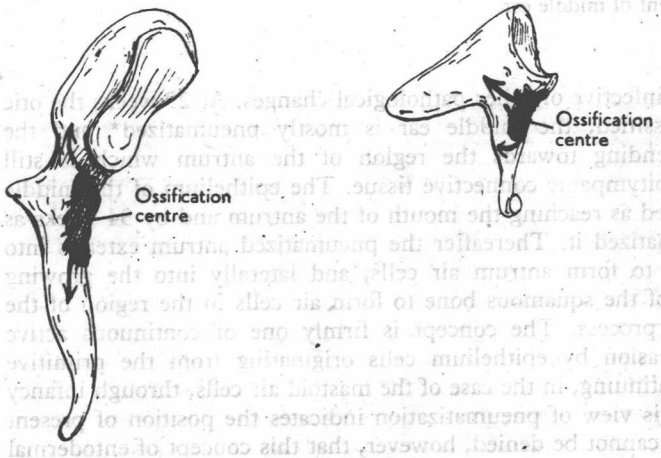

Fig. 1.8. Ossification of malleus and incus.

Last to ossify is the stapes. From a single centre appearing in the footplate, bone spreads along each crus towards the head. Ossification of the stapes is accompanied by complicated remodelling and, having regard to the importance of this bone in the surgery of otosclerosis, more detailed consideration must be given to its development than to that of the malleus and incus.

Stapes (Fig.1.9)

The stapes begins as a ring of cartilage through which passes the stapedial artery. At 16 weeks the ring, by flattening at one end and extrusion of a head at the other, has become a recognizable cartilaginous precursor of the adult form. The changes next to be undergone are to result in the conversion of this solid cartilaginous miniature into the frail and excavated bone so readily fractured under manipulation.

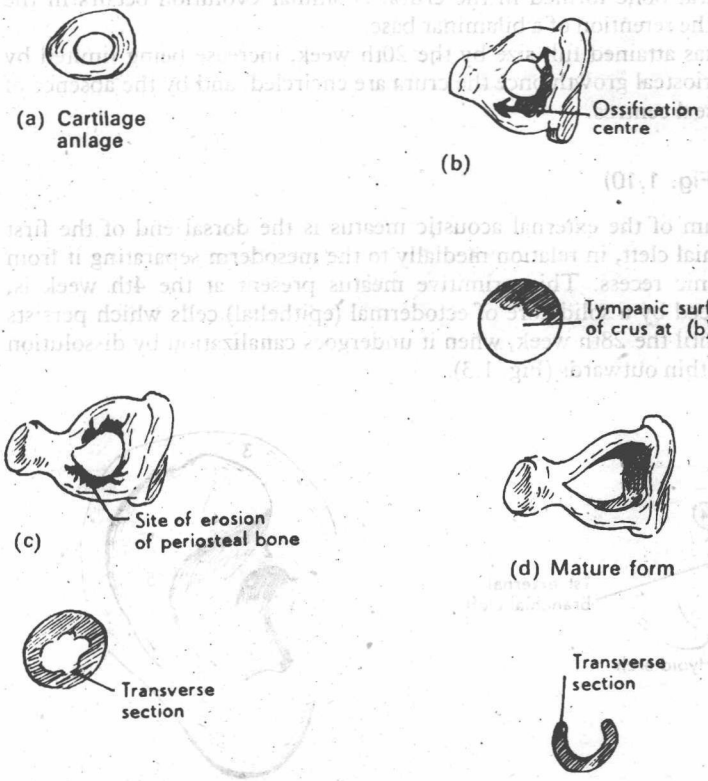

Fig. 1.9. Development of the stapes.

The single ossification centre appears as a plaque on the tympanic surface of the footplate and spreads upwards upon the inner aspect of the crura to extend as an encirclement of periosteal bone over all the surfaces, except for the articular aspect of the head and the labyrinthine aspect of the footplate which remain cartilaginous. At the same time as the bone is spreading superficially there is an invasion deeply, with dissolution of underlying cartilage, by buds of periosteal osteogenic tissue. As the cartilage disappears the crura become converted into tubes of periosteal bone, the core being occupied by marrow spaces. Then, unexpectedly, the early shell of periosteal bone on the inner aspect of each crus

begins to undergo extensive erosion and to become widely cavitated, the marrow space thus exposed being replaced by an invasion of entodermal mucosa (see formation of air spaces and lining membrane, page 6). The crura, therefore, have neither the strength nor stability of a solid cylinder, nor even of a hollow cylinder, but eventually only of a hollow half-cylinder divided in its long axis.

The head of the stapes becomes similarly excavated and invaded from the inner aspect, with the difference that enchondral bone is formed in relation to the articular cartilage thus rendering the articular plate bilaminar; there is little or no enchondral bone formed in the crura. A similar evolution occurs in the footplate with the retention of a bilaminar base.

The stapes has attained full size by the 20th week, increase being limited by cessation of periosteal growth once the crura are encircled, and by the absence of second epiphyseal centres.

External ear (Fig. 1.10)

The primordium of the external acoustic meatus is the dorsal end of the first external branchial cleft, in relation medially to the mesoderm separating it from the tubotympanic recess. This primitive meatus present at the 4th week is, however, replaced by a solid core of ectodermal (epithelial) cells which persists from the 8th until the 28th week, when it undergoes canalization by dissolution of cells from within outwards (Fig. 1.3).

Fig. 1.10. Development of pinna.

At 6 weeks six hillocks or tubercles appear round the dorsal margin of the first cleft, three on the mandibular arch, three on the hyoid. At 12 weeks all the hillocks, save the ventral mandibular hillock, fuse and become incorporated in a general proliferation of the mesodermal element of the 2nd arch which extends crescentically around the meatal core. In the adult ear the tragus represents the persisting ventral mandibular hillock. The rest of the auricle owes its development to the above-mentioned mesodermal output of the hyoid arch carrying an investment of ectoderm (skin) with it.

Postnatal development

Following conversion of the mesoderm surrounding the various parts of the ectodermal labyrinth into a cartilaginous capsule, four centres appear in the cartilaginous capsule at about the 5th or 6th month of fetal life, from which the petrous and mastoid portions of the temporal bone are ossified (Fig. 1.11). One centre appears in the neighbourhood of the arcuate eminence and spreads to the petrous apex, providing cover for part of the cochlea, vestibule, superior semicircular canal and medial wall of the middle ear. A second appears on the medial wall of the middle ear and surrounds the round window, with subsequent extension to form the floor of the middle ear, the investure of the carotid canal, and inferior coverings of the petrous. A third forms the roof of the middle ear and antrum, and the fourth appears near the posterior semicircular canal and extends to form the mastoid process. Meanwhile the squamous temporal has started to ossify in membrane from a single centre appearing near the root of the zygomatic process; while the tympanic part of the temporal bone and the styloid process ossify from one and two centres respectively.

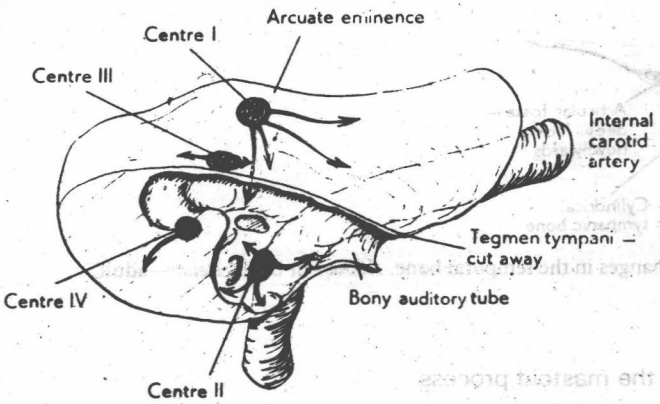

Fig. 1.11. Ossification centres in the petromastoid part of the temporal bone.

The tympanic ring unites with the squamous shortly before birth; the petromastoid fuses with the squamous during the first year and with the proximal part of the styloid process at about the same time. The distal part of the styloid process does not unite with the rest of the bone until after puberty and in some skulls never at all.

Postnatal changes (Fig. 1.12) in the temporal bone consist, apart from increase in size, in alteration in shape of the tympanic ring to form the cylindrical tympanic bone of the adult; deepening and change in direction of the face of the articular fossa; and the growth and pneumatization of the mastoid process. Surgical interest centres in this last, since wide variations in the size and number of air cells present are not only encountered at operation but predetermine the pathological processes that may affect the region. In the infant the absence of mastoid process entails a superficial and unprotected stylomastoid foramen and a vulnerable VIIth (facial) nerve.