DISEASES OF THE EAR

By

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PREFACE

The last two decades have seen a remarkable revival of interest in the ear. Advances in treatment, of deafness in particular, have been as spectacular as those in any branch of surgery or medicine. Originally a separate speciality, otology has so enlarged in scope that it may be said again to occupy a wholly engrossing place within the general field of otorhinolaryngology.

At the same time, for those entering the speciality, and indeed for those already within it, methodical study and methodical teaching have been handicapped by the lack of a single, comprehensive text-book of recent origin. It was felt that a real need existed to assemble and present the subject of otology anew under uniform authorship. This is the task I have undertaken and attempted. Although written with the needs of the specializing postgraduate student primarily in view it is nevertheless also hoped that *Diseases of the Ear* will prove a useful book of reference for general practitioners, house surgeons, registrars, consultants, all, in fact, who may be called upon to treat patients with aural conditions.

Full understanding of the changes resulting from disease requires a very comprehensive knowledge of normality. A relatively large section has, therefore, been set aside for anatomy, physiology and other introductory subjects, corresponding with the requirements of the 1st parts of the examinations for the Fellowship (E.N.T.) of the Royal Colleges of Surgeons, and for the Diploma in Laryngology and Otology of London. The anatomical descriptions have been written with temporal bone dissections at hand, and reciprocal study of such specimens will facilitate assimilation of the text. Operative surgery has been relegated to a separate section at the end, where operating theatre details may be studied without distracting from the primary task of seeing a disease as a whole and of comprehending the principles of treatment upon which detail is based. Elsewhere the classical division of subjects into diseases of the external, middle and internal ear has been followed.

Every single one of the illustrations is original, and has been specially drawn in pen and ink by Harold Ludman, F.R.C.S.. I have been incomparably fortunate in having as illustrator one already specializing in otolaryngology, and I am happy to express my deep gratitude to him for his great help and artistry.

I would also place on record my gratitude for the assistance and encouragement I have received at all times from my colleagues in the Ear, Nose and Throat Department at King's College Hospital, W. I. Daggett, F.R.C.S., Terence Cawthorne, F.R.C.S. and R. S. Lewis, F.R.C.S. For having read my manuscripts with the most critical care, kept alive my interest and answered many questions, despite heavy demands on their time, I am and always shall be greatly in their debt.

To Dr. Stanley A. Mason, F.F.A.R.C.S., Anaesthetist to King's College Hospital, my grateful thanks are due, not only for the section on anaesthesia appearing in Chapter XIX, but for the many harmonious years of co-operation we have enjoyed together in the operating theatre. My preference for general anaesthesia for nearly all operations upon the ear derives from the excellence of the methods advocated. Patients and surgeons who have had experience of a really good, safe general anaesthetic rarely remain in doubt of its advantages.

I would also like to acknowledge my indebtedness to Dr. J. Blewett for the radiographs, and to Dr. D. I. Williams for some of the photographs of skin diseases appearing in Part III. Other photographs have been specially taken by Mr. W. Smith, of the photographic department, whose skill and help have been invaluable.

To Miss Lorna Rutty, my medical secretary, I owe a very heavy debt of gratitude for her spare-time typing of the manuscript. But for her patience and forbearance the book might never have appeared. To her, to Miss Sargeant and the library staff at King's College Hospital, and to all others who have helped me in numerous ways, above all to my wife, I am most sincerely grateful.

STUART MAWSON

Jucundi acti labores

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Introductory

CHAPTER I

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. Following segmentation of zygote into morula, there is differentiation of the blastomeres (morula cells) into an outer, investing layer of cells (trophoblast), and an inner core of cells (inner cell mass) from which the embryo is to develop (fig. 1). At about the same time,

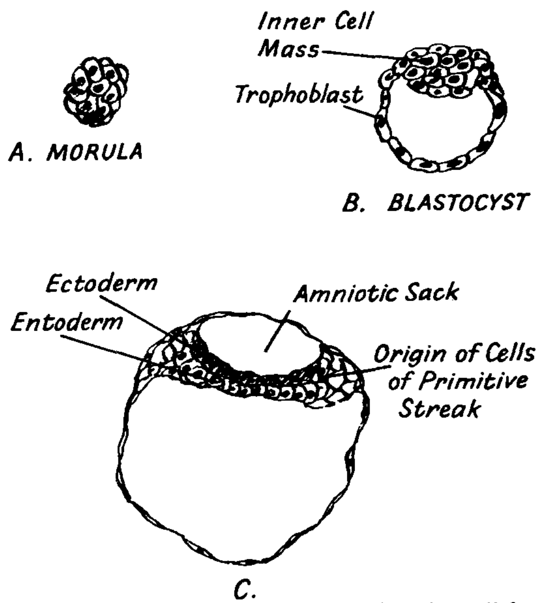

FIG. 1. Development of embryonic primordial layers.

a fluid-filled cavity forms in the morula which, enlarging and dissecting between the inner cell mass and outer cell layer, separates them except at one pole, where they remain in contact. Those cells of the polarized inner cell mass in contact with the outer cell layer (trophoblast) constitute primordial ectoderm, those in contact with the cavity constitute primordial entoderm. This stage is reached at the eighth day of foetal life.

Omitting for simplicity the vesiculations of amnion and primary yolk sack and the brief and partial intervention of primary mesoderm between ectoderm and entoderm, the next event of significance is the appearance of the primitive streak. This condensation of ectoderm at the caudal end of the germ disc, by

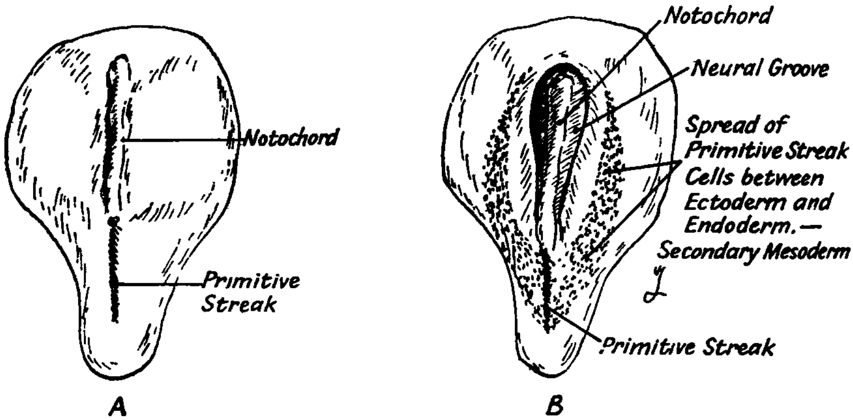

FIG. 2. Dorsal view of germ disc to show primitive streak.

down-growth, becomes intimate with the entoderm and proliferating laterally makes space for itself between ectoderm and entoderm as primordial secondary mesoderm (fig. 2). From these three primitive layers the aural derivatives arise as follows:

TABLE I

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

* 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. 3). 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 13 depressions in the walls of the pharynx, 5 bilaterally represented pouches, the rudimentary thyroid, the respiratory tract and the 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, 4 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 Nos. 1 and 2 meet across the midline between the pericardium and the head.

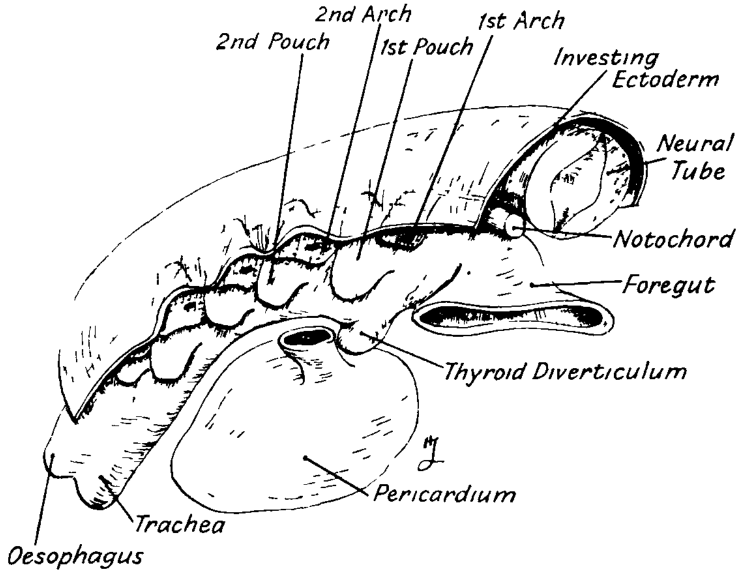

FIG. 3. Front end of embryo to show pharyngeal pouches.

Bar No. 1 persists as the mandible, bar No. 2 as the hyoid, but bars Nos. 3, 4 and 5 remain small and unjoined and soon become overlain and recessed behind a caudal expansion of the second, hydoid, bar. This recess, known as the cervical sinus, is the nursery of branchial cysts and fistulae (fig. 4).

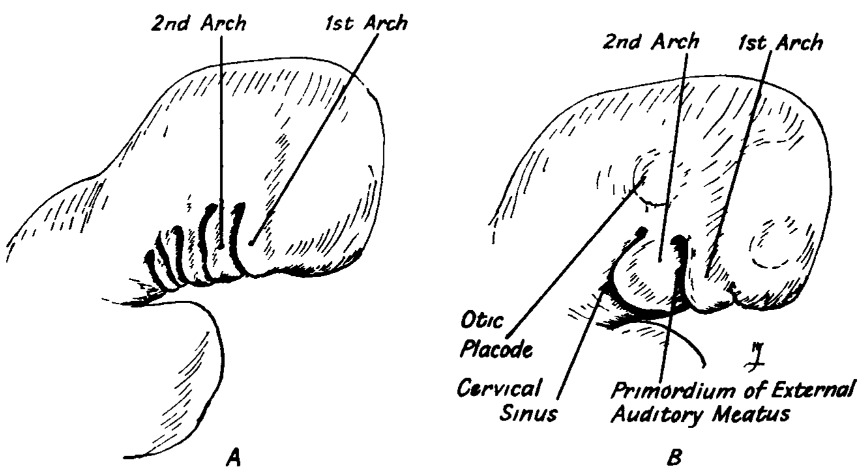

FIG. 4. Development of the cervical sinus. (A) shows overgrowth of second arch caudally.

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 auditory 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 first and third pharyngeal pouches. The tympanic membrane, which separates the tympanic cavity from the external auditory 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. 5).

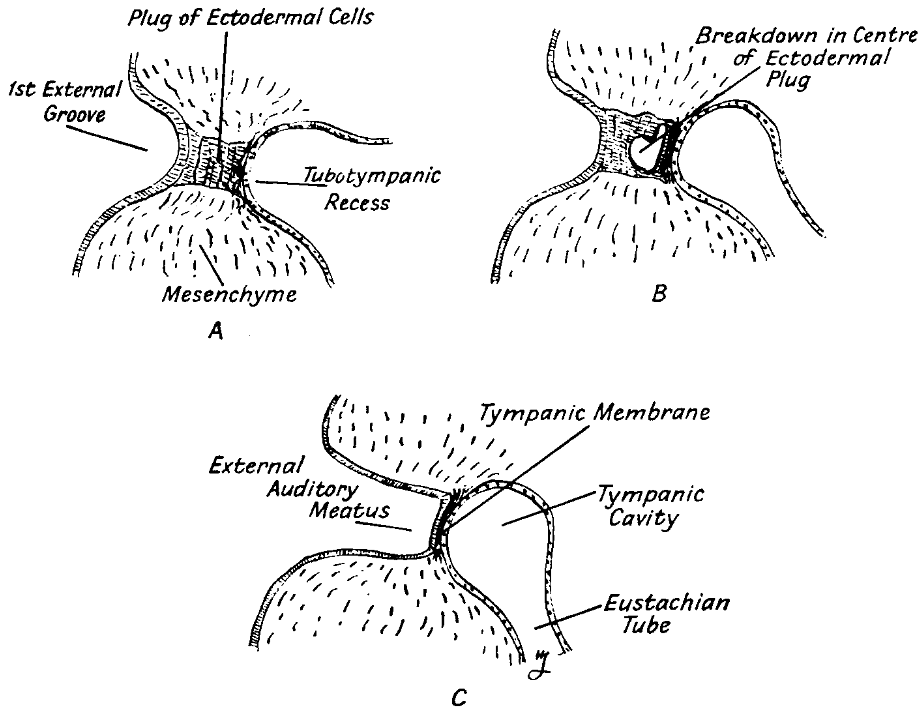

FIG. 5. Development of tympanic membrane.

The third bar or arch, meanwhile, although overshadowed by the second, continues to grow forward under it, and succeeds in narrowing the inner part of the tubotympanic recess to form the Eustachian 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. 6).

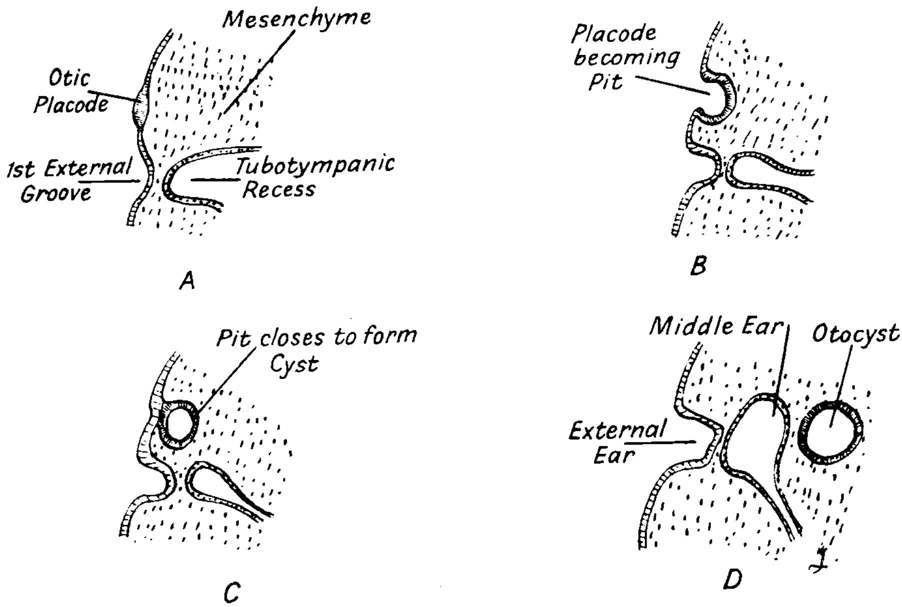

FIG. 6. Development of the otocyst.

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 utriculo-saccular chamber, differentiates into semicircular canals and cochlea (fig. 7). 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 utriculo-saccular 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. 8). Ossification of the cartilaginous capsule begins and is generally completed between the sixteenth and twenty-third week.

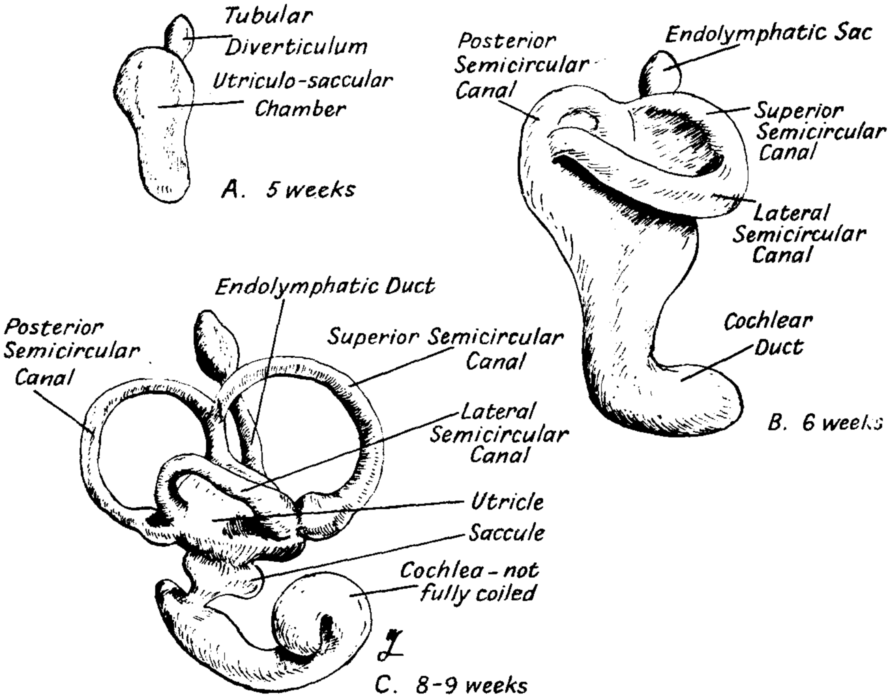

FIG. 7. Development of membranous labyrinth.

Fissula antefenestram

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 (Bast & Anson 1949). 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 sixteenth to twentieth 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 the 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 Eustachian 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 from the dorsal end of the cartilage of the second arch.

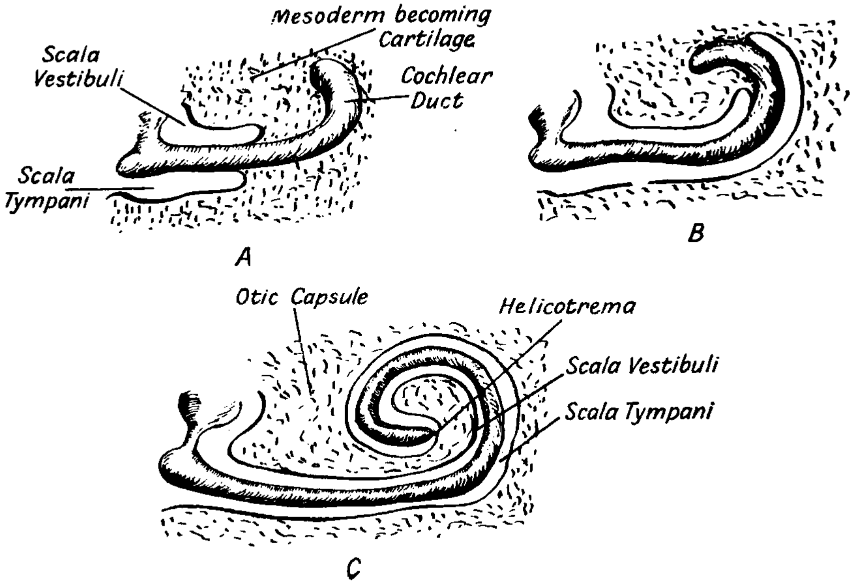

FIG. 8. Diagrammatic representation of the development of the scala tympani and scala vestibuli.

The air spaces and lining membrane

At 12 weeks the differentiating ossicles are at first embedded in the mesoderm overlying the tubotympanic recess (fig. 9). 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, more caudally, to the lateral wall of the Eustachian 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 in the same way as the peritoneal mesothelium covers the intestine (Bast & Anson 1949). This vital process of tympanic expansion by active encroachment of epithelial (entodermal) cells into pre-loosened 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 interference with it by infective or other pathological changes. In the development of the antrum, Bast and Anson, in an impressive study of the anatomy of the developing temporal bone, describe first a tunnelling of loose connective tissue from the epitympanum between the tympanic process of the squamous bone and the ossifying otic capsule. At 29 weeks the otic capsule is well ossified, the middle ear is largely 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

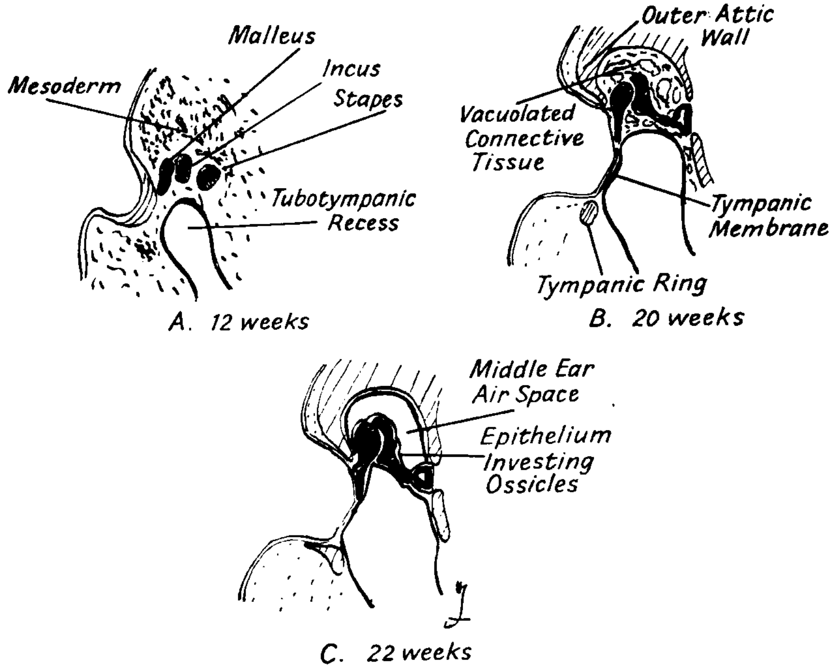

FIG. 9. Development of middle ear.

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 uncompromising support for the classical concept of pneumatization as advanced by Frazer, Wittmaack and others indicates the position of present day orthodoxy. It cannot be denied, however, that the theory of entodermal invasion leaves important questions unanswered, chiefly the histological difference between the epithelium lining the Eustachian tube and lower part of the tympanic cavity, and the epithelium lining the epitympanum, antrum and mastoid air cells;

* Pneumatization here means formation of epithelial lined potential space.

a difference that is strikingly emphasized by markedly dissimilar response to infection by the two epithelial areas. Other observers (Schwarzbart 1959) have suggested that the process of pneumatization is 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 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 (first arch) cartilage, and the stapes from the cartilage of the dorsal end of the second 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. 10). 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.

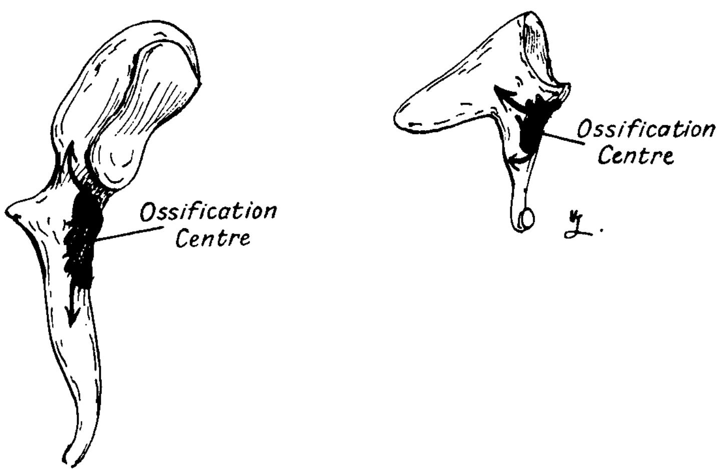

FIG. 10. Ossification of malleus and incus.

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. 11)

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 *hollow* osseous figurine so readily fractured under manipulation.

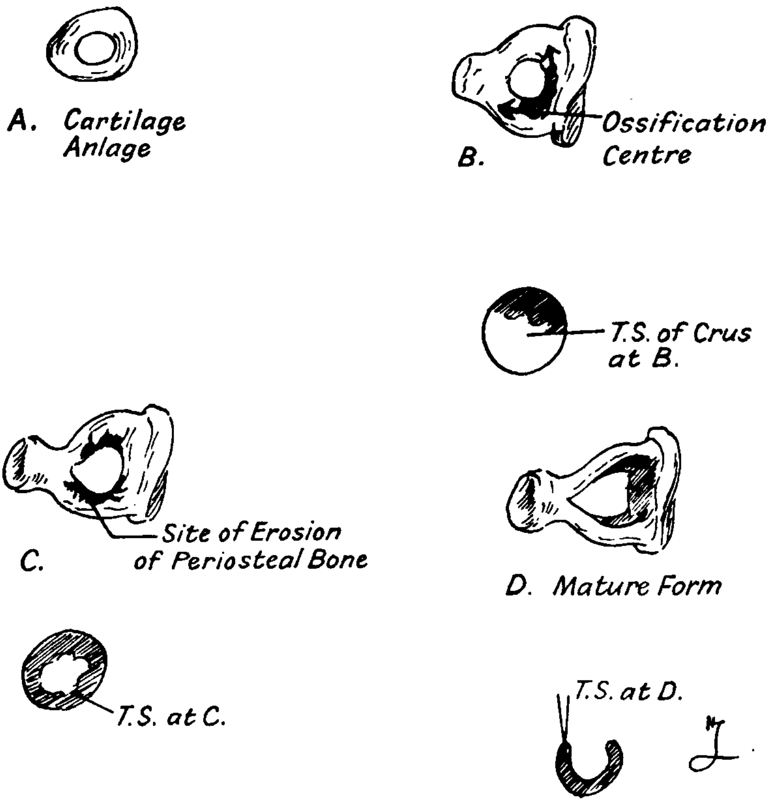

FIG. 11. The development of the stapes.

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 7). 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 (Bast & Anson); 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 secondary epiphyseal centres.

External ear (fig. 12)

The primordium of the external auditory 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 eighth until the twenty-eighth week, when it undergoes canalization by dissolution of cells from within outwards.

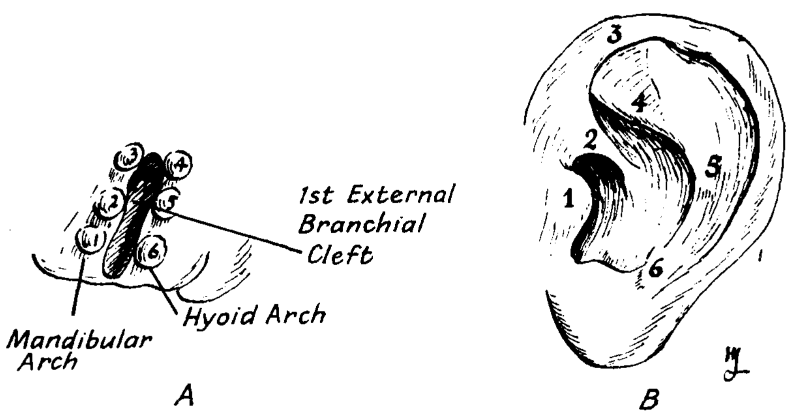

FIG. 12. Development of pinna.

At 6 weeks 6 hillocks or tubercles appear round the dorsal margin of the first cleft, 3 on the mandibular arch, 3 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 second 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.