CHAPTER

1

Embryology of the Ear

Competency achievement: The student should be able to:
EN1.1 Anatomy and physiology of ear, nose, throat, head and neck.

Ear has a very complex source of development. The sound conductive apparatus develops from the branchial apparatus, whereas the sound perceptive apparatus develops from the ectodermal otocyst (pars otica). Because of this dual source of origin, the developmental anomaly produced, commonly affects either the sound conductive system which includes anomaly of the external and/or the middle ear or the sound perceptive apparatus which includes the labyrinth. Both these anomalies rarely coexist because of different source of origin.

Development of the External Ear

This develops around the first branchial cleft.

The Pinna

Around 6th week of intrauterine life, six hillocks or 'tubercles of His' appear around the first branchial cleft (Fig. 1.1). The first tubercle is derived from the first branchial arch and the rest from the 2nd branchial arch. Some authors believe that the first 3 tubercles develop from the first arch and the rest from the 2nd arch.

Structures Derived from Various Hillocks

1. Tragus
2. Crus of the helix
3. Helix
4. Antihelix
5. Scapha and the antitragus
6. Ear lobule

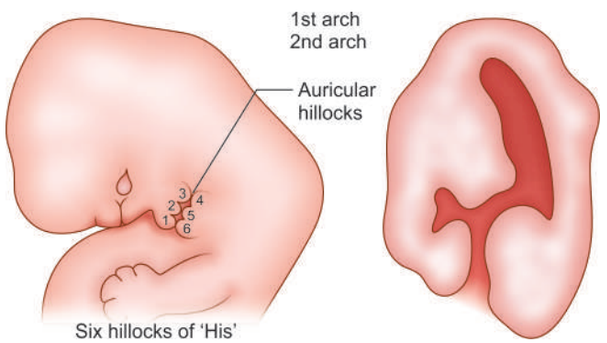


Fig. 1.1: Development of external ear from six hillocks

The ear takes definitive form by the end of third month of intrauterine life. Defective fusion of the tubercles gives rise to preauricular sinus and failure of the development of the hillocks causes anotia. Defective development of 4th tubercle can cause absence of antihelix leading to 'bat ear' deformity.

External Auditory Canal

This develops from the first branchial cleft as an invagination into a funnel-shaped pit to form a primary external auditory canal. Subsequent medial growth with a solid core of ectoderm leads to formation of a meatal plate called the secondary external auditory canal. Between 8th and 10th weeks of IUL, the solid core of epithelium undergoes canalization forming the definitive external auditory canal.

Anomalies of the External Auditory Canal

1. Complete atresia (fibrous and/or bony)
2. Shallow depression
3. Changes in the curvature of the canal
4. Stenosis

Development of Tympanomastoid Cavity and Eustachian Tube (Fig. 1.2)

Around 3rd week of IUL, the first pharyngeal pouch develops which is phylogenetically the aquatic gill apparatus. This outpouching of the first pharyngeal pouch gives rise to two components, namely:

1. The proximal narrow part which forms the eustachian tube.
2. The distal dilated part which gives rise to the developing middle ear cleft and is known as the tubotympanic recess. This forms the definitive tympanic cavity by progressively and systematically invaginating into the adjacent mesenchyme.

Towards the later part of the fetal life a diverticulum appears from the tubotympanic recess which

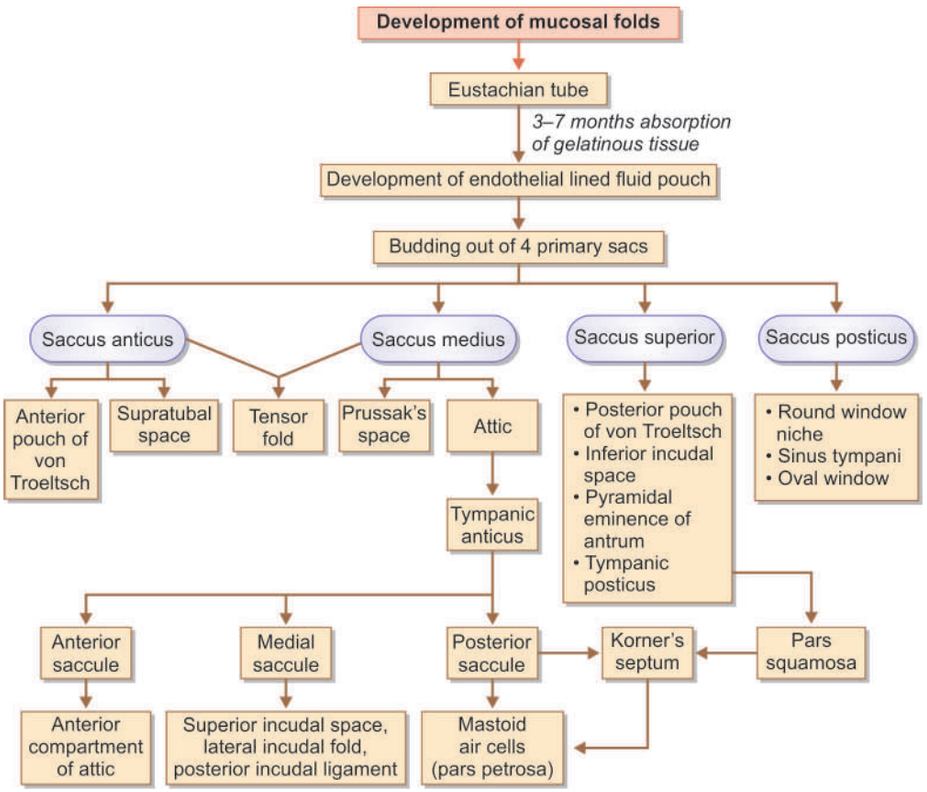


Fig. 1.2: Development of mucosal folds

subsequently forms the mastoid antrum. This antrum is about 3 mm thickness at birth and it increases 1 mm every year till it reaches the adult size of 15 mm thickness.

Development of Ossicles

Anson in 1959 described the details of the development of the ossicles. The first arch cartilage (Meckel's cartilage) forms the head of the malleus and the body of the incus. The second arch forms the manubrium (handle) of the malleus and the long process of the incus and the crurae of the stapes. These sources of development confirm the various developmental anomalies involving the ossicles as encountered during surgery. The footplate of the stapes develops from three sources, namely:

1. The outer periosteal layer of the otic capsule.
2. Middle enchondral layer from the otic capsule.
3. Inner endosteal layer is same as the endosteum of the bony labyrinth and develops from the periotic mesoderm.

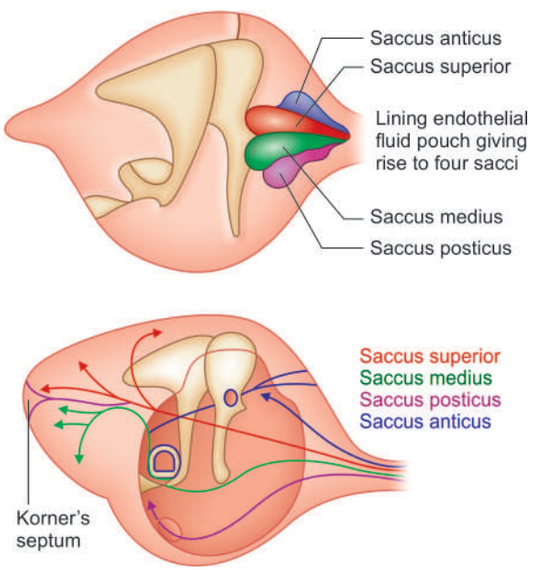
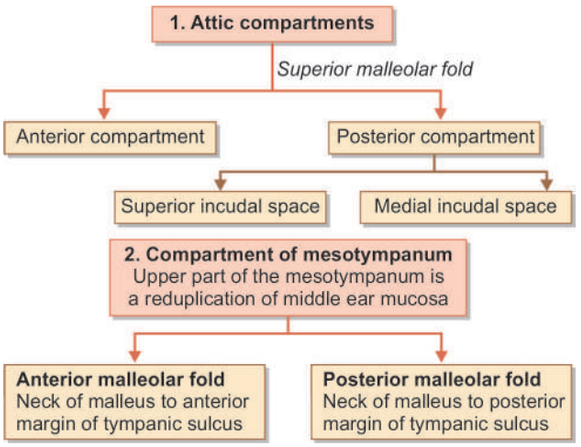


Fig. 1.3: Development of four primary sacs

Development of Middle Ear Spaces and Folds

The envelopment of the ossicles by the mucous membrane lining of the tubotympanic recess occurs between 3 and 7 months of IUL. This mucous lining while encircling the ossicles form numerous folds and spaces as follows (Figs 1.2 to 1.5A to D).



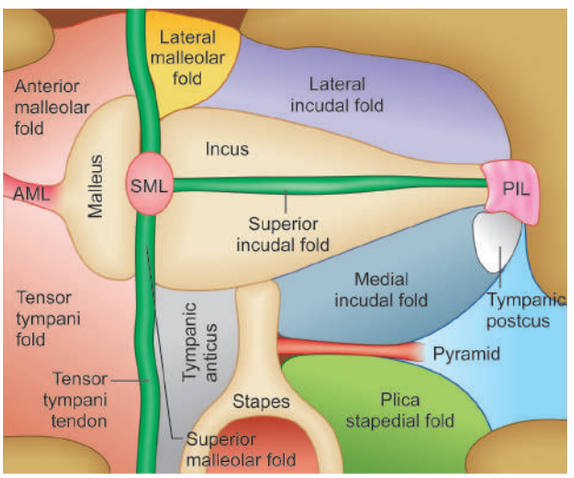


Fig. 1.4: Various middle ear folds and boundaries of tympanic anticus (TA)

COMPARTMENT AND FOLDS OF THE TYMPANIC CAVITY (Figs 1.5A to D)

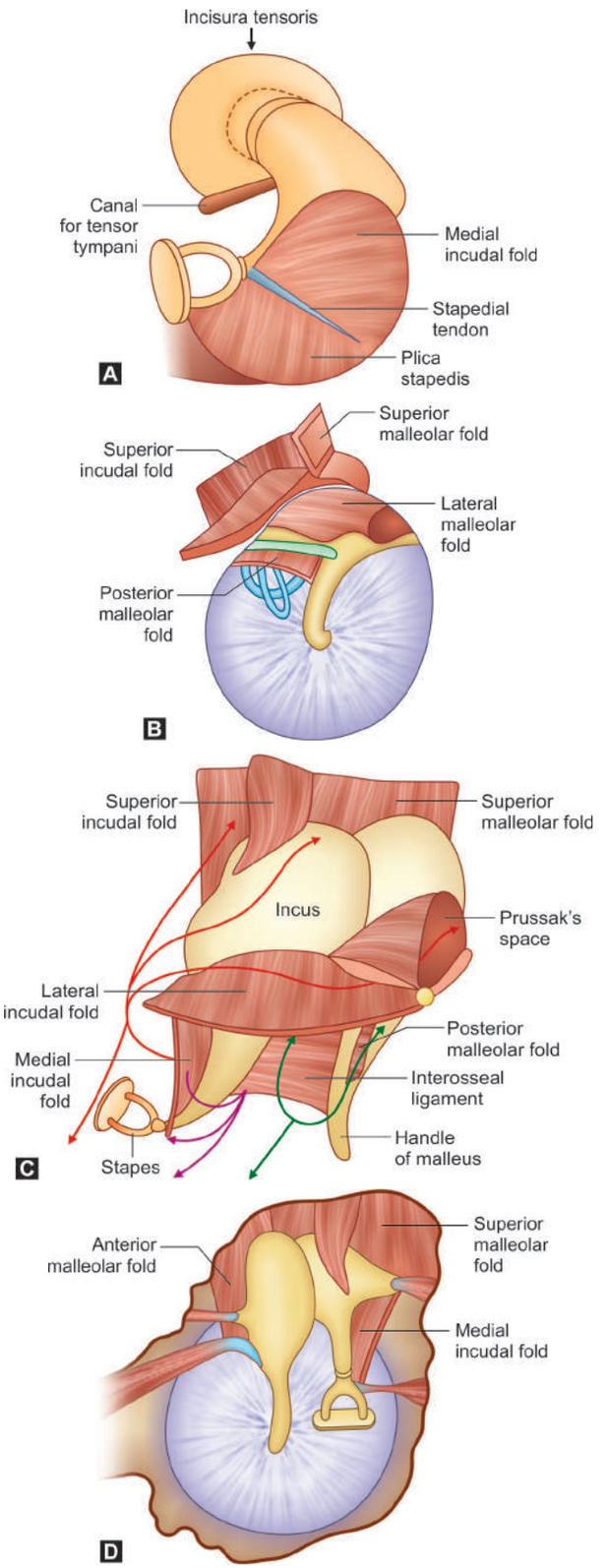
Prussak's Space (Figs 1.5C and 1.6)

It is a potential space which may be the first to involve during the extension of cholesteatoma, it is bounded by (Fig. 1.6):

- Laterally by Shrapnell's membrane (pars flaccida)
- Medially by the neck of malleus
- Superiorly by fibers of lateral malleolar fold
- Inferiorly lateral process of malleus.

Development of the Inner Ear

The inner ear develops from the otic capsule (pars otica). Initially a thickening appears in the ectoderm of the hindbrain known as otic placode. It later invaginates forming otic cyst which is also known as otic capsule. Subsequent differentiation of this otic cyst (vesicles) leads to formation of membranous labyrinth. After the formation of ectodermal otocyst at 4 weeks of fetal life (WFL), the growth of the semicircular canals occurs at 5 WFL, and the elongation and coiling of the



Figs 1.5A to D: Various mucosal folds that are developed during the process of development and the middle ear spaces that are formed from these folds

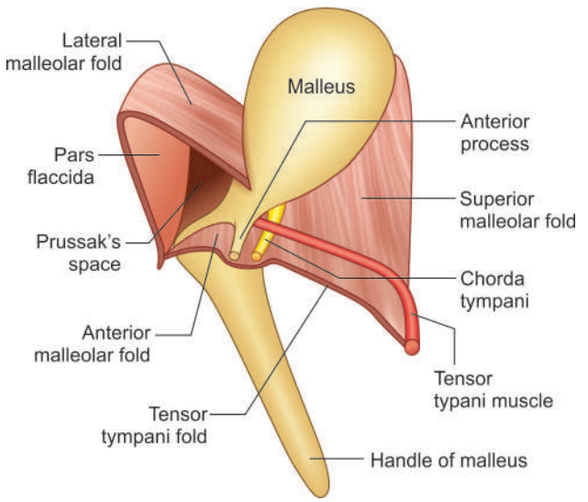


Fig. 1.6: Boundaries of Prussak's space

cochlea at 10 WFL. The mesoderm surrounding the otic capsule forms the bony labyrinth along with endochondral layer of the otic capsule. As the membranous labyrinth takes shape, there is a concomitant development of the sensory neuroepithelia and their associated structures within. The prenatal labyrinth attains an adult equivalent size between 17 and 19 weeks of fetal life (Fig. 1.7). Incomplete development of otic capsule can be associated with various inner ear abnormalities leading to deafness.

Pneumatization of mastoid: It is the process by which the aircell system extends as an outgrowth from the epithelial sac of the middle ear and antrum in conjuction with the enlargement of temporal bone which starts immediately after birth. Both genetic and acquired factors have been implicated to influence pneumatization process. The mastoid process develops at the end of 1st year due to the contraction of the sternocleidomastoid muscles as the child tries to lift the head. In 80% of cases, mastoid is fully pneumatized and in rest of the cases it may be partially pneumatized or sclerotic. Based on this the mastoid can be of three types:

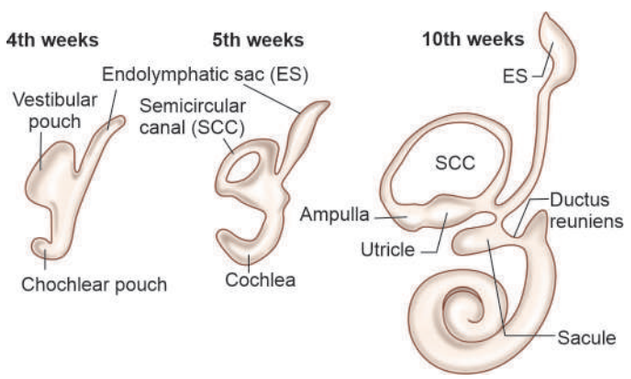


Fig. 1.7: Developing inner ear

1. Cellular (well pneumatized)
2. Diploeic (poorly pneumatized) few small-sized cells
3. Sclerotic (non-pneumatized) absence of cells that are replaced with dense, compact bone. Sclerotic mastoid can be divided further into:
   - Primary sclerotic mastoid (not developed)
   - Secondary sclerotic mastoid (acquired).

Theories of Faulty Pneumatization

1. *Tumarkin theory:* Poor ventilation of the middle ear cleft due to eustachian tube dysfunction leading to arrest of pneumatization.
2. *Wittmaack theory:* He proposed that otitis media in infancy and early childhood leads to poor ventilation of middle ear cleft causing arrest of pneumatization.
3. Diament was of the view that pneumatization of the mastoid bone is determined by hereditary factors.
4. Genetic factors were also implicated by Stern
5. *Graham and Brackmen:* The size of mastoid depends upon the final size of the skull in an individual as in acromegaly (large expansive mastoid) and in microcephaly (underdeveloped mastoid).

Classification of Mastoid Air Cell System

Allam (1969) classified the pneumatized spaces of the temporal bone into five different regions: Middle ear region, mastoid region, perilabyrinthine region, petrous apex region and accessory region (*see* Fig. 2.19).

1. *Middle ear region*: Hypotympanic cells, etc.
2. *Mastoid region:*
   - Central: Antrum and periantral cells.
   - Peripheral: Tegmen cells, sinodural cells, perisinus cells, retrofacial cells and tip cells
3. *Perilabyrinthine region*: Supra- and infralabyrinthine cells
4. *Petrous apex region*: Apical cells.
5. *Accessory region*: Zygomatic, squamous, occipital, styloid and peritubal cells

Points to Remember

1. The sound conducting apparatus develops from the branchial apparatus, whereas the sound perceptive apparatus from the ectodermal otocyst.
2. The pinna develops from the six hillocks around the 1st branchial cleft.
3. Defective fusion of tubercles gives rise to preauricular sinus.
4. The outpouching of the 1st pharyngeal pouch gives rise to a proximal narrow part that forms the eustachian tube and distal dilated part that forms the middle ear cavity.
5. Prussak's space is a potential space lateral to the Shrapnell's membrane and medially by the neck of malleus that can be involved during the extension of cholesteatoma.