Objectives:
- describe normal development of the outer, middle, and inner ear from the 1st pharyngeal cleft, 1st pouch, and the otic vesicle
- name the more common abnormalities of the ear

Internal Ear
The first indication of the developing ear can be found in embryos of approximately 22 days as a thickening of the surface ectoderm on each side of the rhombencephalon (otic placodes).
Each otic placode soon invaginates into the under-lying mesenchyme and forms an otic pit. The edges of the otic pit soon come together and fuse to form an otic vesicle.
The otic vesicle then loses its connection with the surface ectoderm During later development, each vesicle divides into
1. a ventral component (saccular portion) that gives rise to the saccule and cochlear duct and
2. a dorsal component (utricular portion with the endolymphatic duct) that forms the utricle, semicircular canals, and endolymphatic duct.
Together, these epithelial structures form the membranous labyrinth.

Saccule (Cochlear duct)
In the sixth week of development, the saccule forms a tubular outpocketing (the cochlear duct) at its lower pole. This outgrowth, penetrates the surrounding mesenchyme in a spiral fashion until the end of the eighth week, when it has completed 2.5 turns. Its connection with the remaining portion of the saccule is then confined to a narrow pathway, the ductus reuniens.

Scala tympani and Scala vestibuli
Mesenchyme surrounding the cochlear duct soon differentiates into cartilage. In the 10th week, this cartilaginous shell undergoes vacuolization, and two perilymphatic spaces, the scala vestibuli and scala tympani, are formed.

Vestibular membrane and Basilar membrane
The cochlear duct is then separated from the scala vestibuli by the vestibular membrane and from the scala tympani by the basilar membrane.

Spiral ligament and Modiolus
The lateral wall of the cochlear duct remains attached to the surrounding cartilage by the spiral ligament, whereas its median angle is connected to and partly supported by a long cartilaginous process, the modiolus, the future axis of the bony cochlea. Initially, epithelial cells of the cochlear duct are alike. With further development, however, they form two ridges: the inner ridge, the future spiral limbus, and the outer ridge.

Organ of Corti
The outer ridge forms one row of inner and three or four rows of outer hair cells, the sensory cells of the auditory system. They are covered by the tectorial membrane, a fibrillar gelatinous substance attached to the spiral limbus that rests with its tip on the hair cells. The sensory cells and tectorial membrane together constitute the organ of Corti. Impulses received by this organ are transmitted to the spiral ganglion and then to the nervous system by the auditory fibers of cranial nerve VIII.

Utricle and Semicircular Canals
During the sixth week of development, semicircular canals appear as flattened outpocketings of the utricular part of the otic vesicle. Central portions of the walls of these outpocketings eventually appose each other and disappear, giving rise to three semicircular canals. Whereas one end of each canal dilates to form the crus ampullare, the other, the crus nonampullare, does not widen. Since two of the latter type fuse, however, only five crura enter the utricle, three with an ampulla and two without.
Cells in the ampullae form a crest, the crista ampullaris, containing sensory cells for maintenance of equilibrium. Similar sensory areas, the maculae acusticae, develop in the walls of the utricle and saccule.

Impulses generated in sensory cells of the cristae and maculae as a result of a change in position of the body are carried to the brain by vestibular fibers of cranial nerve VIII.

**Statoacoustic ganglion**

During formation of the otic vesicle, a small group of cells breaks away from its wall and forms the statoacoustic ganglion. Other cells of this ganglion are derived from the neural crest.

The ganglion subsequently splits into cochlear and vestibular portions, which supply sensory cells of the organ of Corti and those of the saccule, utricle, and semicircular canals, respectively.

**Middle Ear**

The tympanic cavity originates in the endoderm and is derived from the first pharyngeal pouch. This pouch expands in a lateral direction and comes in contact with the floor of the first pharyngeal cleft.

The distal part of the pouch, the tubotympanic recess, widens and gives rise to the primitive tympanic cavity, and the proximal part remains narrow and forms the auditory tube, through which the tympanic cavity communicates with the nasopharynx.

**Ossicles**

The malleus and incus are derived from cartilage of the first pharyngeal arch, and the stapes is derived from that of the second arch. Although the ossicles appear during the first half of fetal life, they remain embedded in mesenchyme until the eighth month, when the surrounding tissue dissolves. The endodermal epithelial lining of the primitive tympanic cavity then extends along the wall of the newly developing space. The tympanic cavity is now at least twice as large as before.

When the ossicles are entirely free of surrounding mesenchyme, the endodermal epithelium connects them in a mesentery-like fashion to the wall of the cavity. The supporting ligaments of the ossicles develop later within these mesenteries.

Since the malleus is derived from the first pharyngeal arch, its muscle, the tensor tympani, is innervated by the mandibular branch of the trigeminal nerve. The stapedius muscle, which is attached to the stapes, is innervated by the facial nerve, the nerve to the second pharyngeal arch.

**Mastoid antrum**

During the late fetal period, expansion of the tympanic cavity gives rise to the mastoid antrum, located in the petromastoid part of the temporal bone. The mastoid antrum is almost adult size at birth; however, no mastoid cells are present in newborn infants.

By 2 years of age, the mastoid cells are well developed and produce conical projections of the temporal bones, the mastoid processes. The middle ear continues to grow through puberty.

**External Ear**

The external auditory meatus develops from the dorsal portion of the first pharyngeal cleft.

At the beginning of the third month, epithelial cells at the bottom of the meatus proliferate, forming a solid epithelial plate, the meatal plug.

In the seventh month, this plug dissolves and the epithelial lining of the floor of the meatus participates in formation of the definitive eardrum.

Occasionally, the meatal plug persists until birth, resulting in congenital deafness.

The external acoustic meatus, relatively short at birth, attains its adult length in approximately the ninth year.

**Eardrum or Tympanic Membrane**
The eardrum is made up of
1. ectodermal epithelial lining at the bottom of the auditory meatus,
2. endodermal epithelial lining of the tympanic cavity, and
3. an intermediate layer of connective tissue that forms the fibrous stratum.

The major part of the eardrum is firmly attached to the handle of the malleus and the remaining portion forms the separation between the external auditory meatus and the tympanic cavity.

**Auricle**

develops from six mesenchymal proliferations at the dorsal ends of the first and second pharyngeal arches, surrounding the first pharyngeal cleft. 

hillocks 1, 2, and 3 are part of the mandibular portion of the first pharyngeal arch and that the ear lies horizontally at the side of the neck. At this stage, the mandible is small.

These swellings (auricular hillocks), three on each side of the external meatus, later fuse and form the definitive auricle. As fusion of the auricular hillocks is complicated, developmental abnormalities of the auricle are common.

Initially, the external ears are in the lower neck region but with development of the mandible, they ascend to the side of the head at the level of the eyes.

The parts of the auricle derived from the first pharyngeal arch are supplied by its nerve, the mandibular branch of the trigeminal nerve. The parts derived from the second arch are supplied by cutaneous branches of the cervical plexus, especially the lesser occipital and greater auricular nerves.

The facial nerve of the second pharyngeal arch has few cutaneous branches; some of its fibers contribute to the sensory innervation of the skin in the mastoid region and probably in small areas on both aspects of the auricle.

**Clinical Correlates**

**Congenital deafness**

usually associated with deaf-mutism, may be caused by
1. abnormal development of the membranous and bony labyrinths or
2. by malformations of the auditory ossicles and eardrum.
3. In the most extreme cases, the tympanic cavity and external meatus are absent.

Most forms of congenital deafness are caused by
1. genetic factors, but
2. environmental factors may also interfere with normal development of the internal and middle ear.

Rubella virus, affecting the embryo in the seventh or eighth week, may cause severe damage to the organ of Corti. It has also been suggested that poliomyelitis, erythroblastosis fetalis, diabetes, hypothyroidism, and toxoplasmosis can cause congenital deafness.

**External ear defects**

are common; they include minor and severe abnormalities. They are significant
1. from the standpoint of the psychological and emotional trauma they may cause and
2. for the fact they are often associated with other malformations.

Thus, they serve as clues to examine infants carefully for other abnormalities. All of the frequently occurring chromosomal syndromes and most of the less common ones have ear anomalies as one of their characteristics.

**Preauricular appendages and pits**

They are skin tags and shallow depressions, respectively, anterior to the ear. Pits may indicate abnormal development of the auricular hillocks, whereas appendages may be due to accessory hillocks. Like other external ear defects, both are associated with other malformations.