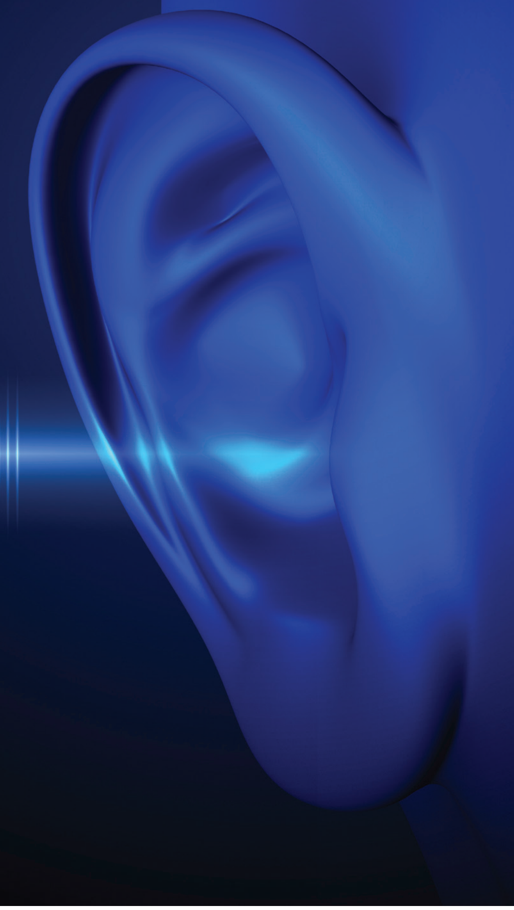
CHAPTER 1



DISEASES OF THE EAR AND HEARING DISORDERS

The physical event initiating the perception of sound is the change in the state of rest of the medium the sound wave passes through. The periodic compression and rarefaction of the particles of the medium create a change in pressure (**Fig. 1.1B)** that gives rise to a phenomenon perceived as vibration by the sense of touch particularly for solid media) and as an acoustic sensation by the sense of hearing (particularly if air is the medium). Sound is produced by a source that disturbs the state of rest of particles (**Fig. 1.1A**) that begin to oscillate back and forth, thus transferring their energy to adjacent particles and propagate the sound wave through space (**Figs. 1.1C, 1.1D**). The changes in the pressure of the medium over time are represented graphically by a curved line above and below a straight line that represents the theoretical state of rest of the medium the sound wave travels through (**Fig. 1.1A**). In simple terms, oscillation of particles with a sinusoidal pattern (pure tone) is characterized by:

- oscillation frequency;
- oscillation amplitude.

The oscillation frequency of a pure tone is measured in *Hertz* (Hz), *i.e.*, the number of cycles per second (**Fig. 1.2**). This frequency of vibration determines its *pitch*).

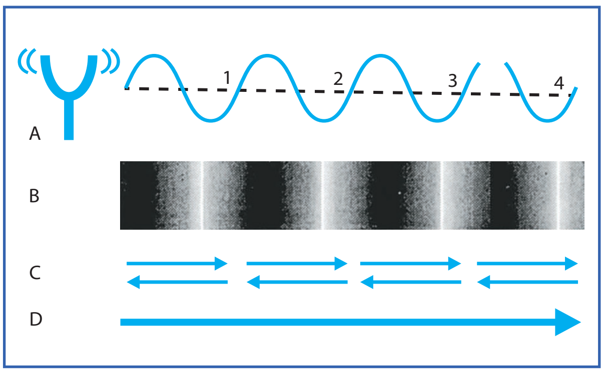


Fig. 1.1. Movement in air of a sound wave produced by a tuning fork: A) sinusoidal wave form of a pure tone; B) compression (dark area) and rarefaction (light area) of particles along the propagation axis of wave propagation; C) movement of air particles; D) direction of wave propagation.

Based on the capability of the human ear to perceive sound, pitch is distinguished as:

- low, up to 500 Hz;
- medium, between 1000 and 3000 Hz;
- high, over 3000 Hz;
- ultrasounds, over 15.000-20.000 Hz;
- infrasounds, below 20-50 Hz.

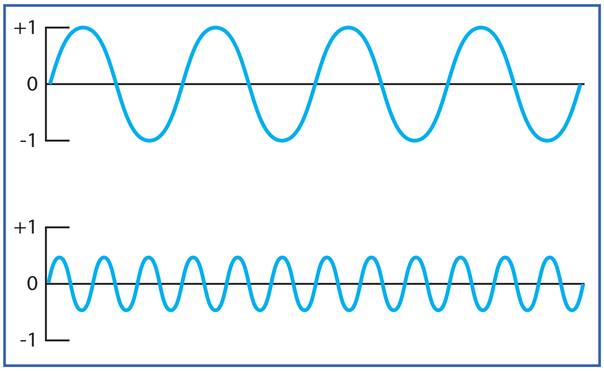


Fig. 1.2. Two pure tones at different frequencies: low frequency (upper half) and high frequency (lower half).

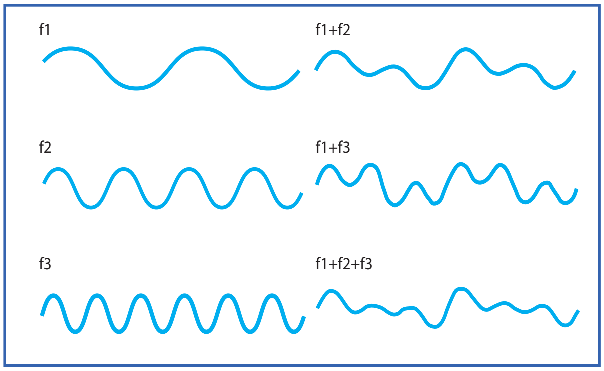


Fig. 1.3. Complex sound waves (right side) can be broken down into pure tones (left side) and summated.

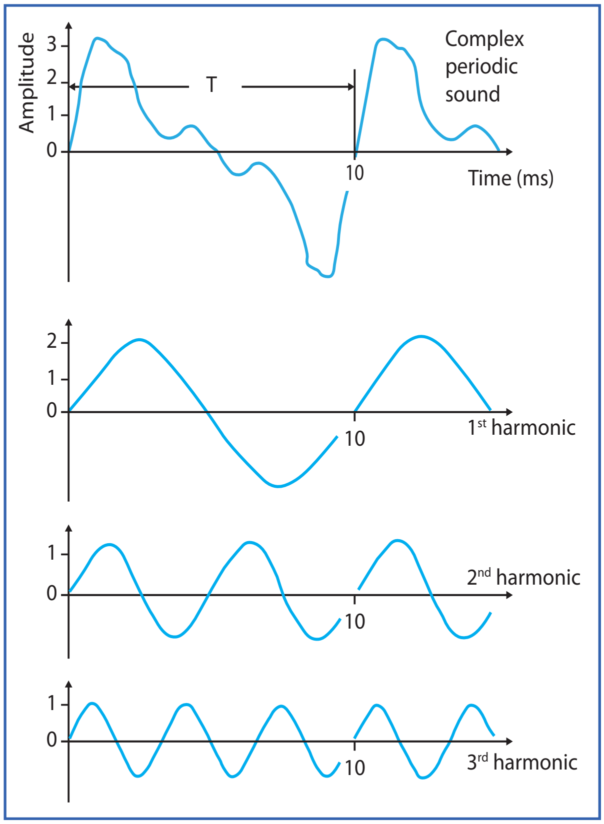


Fig. 1.4. Periodic sound wave. Wave form (from top to bottom), fundamental frequency (lowest frequency of a waveform); harmonics, which are integer multiples of the first harmonic.

Ultrasounds and infrasounds are not perceivable by the human ear as an acoustic signal. The acoustic signals we perceive are usually complex tones composed of irregular sound wave forms (non-sinusoidal). These signals can be recomposed via Fourier analysis in a series of associated pure tones (**Fig. 1.3**).

The signals can be distinguished in:

- periodic sounds, composed of a fundamental frequency (the lowest) and harmonic frequencies, which are integral multiples of the fundamental (**Fig. 1.4**) and are produced by the larynx (voice) and musical instruments;
- noise sound waves lacking periodicity (**Fig. 1.5**).

Acoustic events can also be represented by the frequency domain (spectrum); in which the frequency is reported on the x axis and the intensity on the y axis (**Fig. 1.6**).

The intensity of sound perception (loudness) is determined by the amplitude of a sound wave: the greater the amplitude, the more intense the perception of the sound. Intensity can be measured as power or pressure. Power refers to the capacity of a sound source (Hi-Fi amplifier, machines, vehicles, etc.) to produce noise. Intensity is measured in *Watt* (work/time, W). A more useful unit of measure to define the perception of sound is to express it in *Pascal* (Newton/m^2), a unit of measure that measures the actual intensity of sound at the point where it is measured and is correlated with the distance between the sound source and the point where it is measured. Pascal is a physical unit of measurement that reflects the functional characteristics of the human ear which, when analyzing sound intensity, follows a logarithmic and not a linear curve (Weber-Fechner law), in a very wide range of least perceptible sound and an extremely painful sound pain (at least 10^6).

The perception of sound loudness (intensity) is measured in *decibel* (dB), *i.e.*, 20 times the base 10 logarithm of the pressure measured compared to the *reference pressure* (P0), conventionally set at $2 \bullet 10^{-5}$ Pascal (value defined as the minimum loudness perceivable by a young person not affected by a hearing disorder at 1000 Hz).

$$\text{dB} = 20 \log_{10} \frac{P}{P_0}$$

Decibel (dB) is not an absolute measurement but rather a relative unit of measurement correlated to the cha-

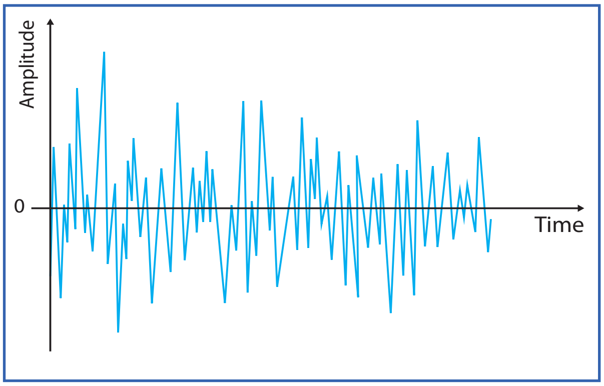


Fig. 1.5. Noise with nonperiodic wave form.

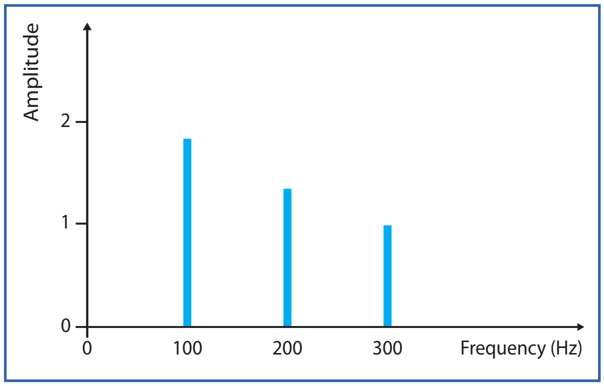


Fig. 1.6. Spectral analysis of a complex periodic sound. Frequencies are reported on the X axis and loudness on the Y axis.

racteristics of the sensitivity of the human ear, in which 0 does not denote the absence of sound (which is possible only in a vacuum) but rather the minimum sound perceivable by the human ear. Furthermore, the use of logarithmic measurement reduces via a simple mode of analysis, like that done by the ear itself, the range of sound loudness. As measured in dB the range of perceivable sound by the human ear is from about circa 0 (minimum sound perceivable) to 100-120 dB (threshold of uncomfortable or painful loudness). A useful way to correlate a value expressed in dB and acoustic perception is to remember the following scale: 40 dB in a quiet room; 60 dB the level of normal conversation; 85-110 dB a particularly noisy environment (e.g., factory, discotheque); >120 dB blasts and explosions (firearms, etc.) noise generated by large engines (airplanes, rockets, etc.).

Table 1-I. Anatomy and physiology of the ear.

System for the transmission of mechanical vibratory energy *Components:* » *external ear* (auricle and extra auditory canal); » *middle ear* (ear drum and tympanic membrane and transmission components, round window, oval window); » *labyrinthine* fluids (perilymph, endolymph) membrane of the inner ear (Reissner's membrane, tectorial membrane, basilar membrane).
System for the transduction of mechanical vibratory energy in nervous energy » sensorial cells of the organ of Corti.
System for the transfer of nervous energy and its transformation in auditory perception components: » *fibers of the cochlear branch of the acoustic nerve*, i.e. peripheral and central prolongations of the cells forming the ganglion organ of Corti; » *afferent acoustic pathway and its nuclei* (dorsal cochlear nucleus, ventral cochlear nucleus, superior olivary complex, nucleus of the lateral lemniscus, inferior quadrigeminal body, medial geniculate body, acoustic cortical areas and related areas, nerve pathways of the connection between the nuclei and the nuclei of the central acoustic pathway).

ANATOMY AND PHYSIOLOGY OF THE EAR

ANATOMY OF THE EAR

The organ of hearing is composed of three systems: a system that transmits mechanical vibratory energy, a system that transforms mechanical energy in nervous energy; a system that transmits nervous energy to the cortex of the temporal lobe where it is transformed in acoustic signals (**Fig. 1.7** and **Table 1-I**).
The anatomical structures that enable hearing are:

- external ear;
- middle ear;
- internal ear;
- acoustic nerve;
- central auditory pathways.

External ear

The external ear is composed of the auricle and the external auditory canal. The auricle consists of a conch-shaped cartilage flap covered with skin that protrudes from each side of the head (**Fig. 1.8**). At the center of the auricle the orifice of the external auditory opens. The canal extends about 24 mm in depth. It is composed of fibrocartilaginous tissue in the lateral third and bone in

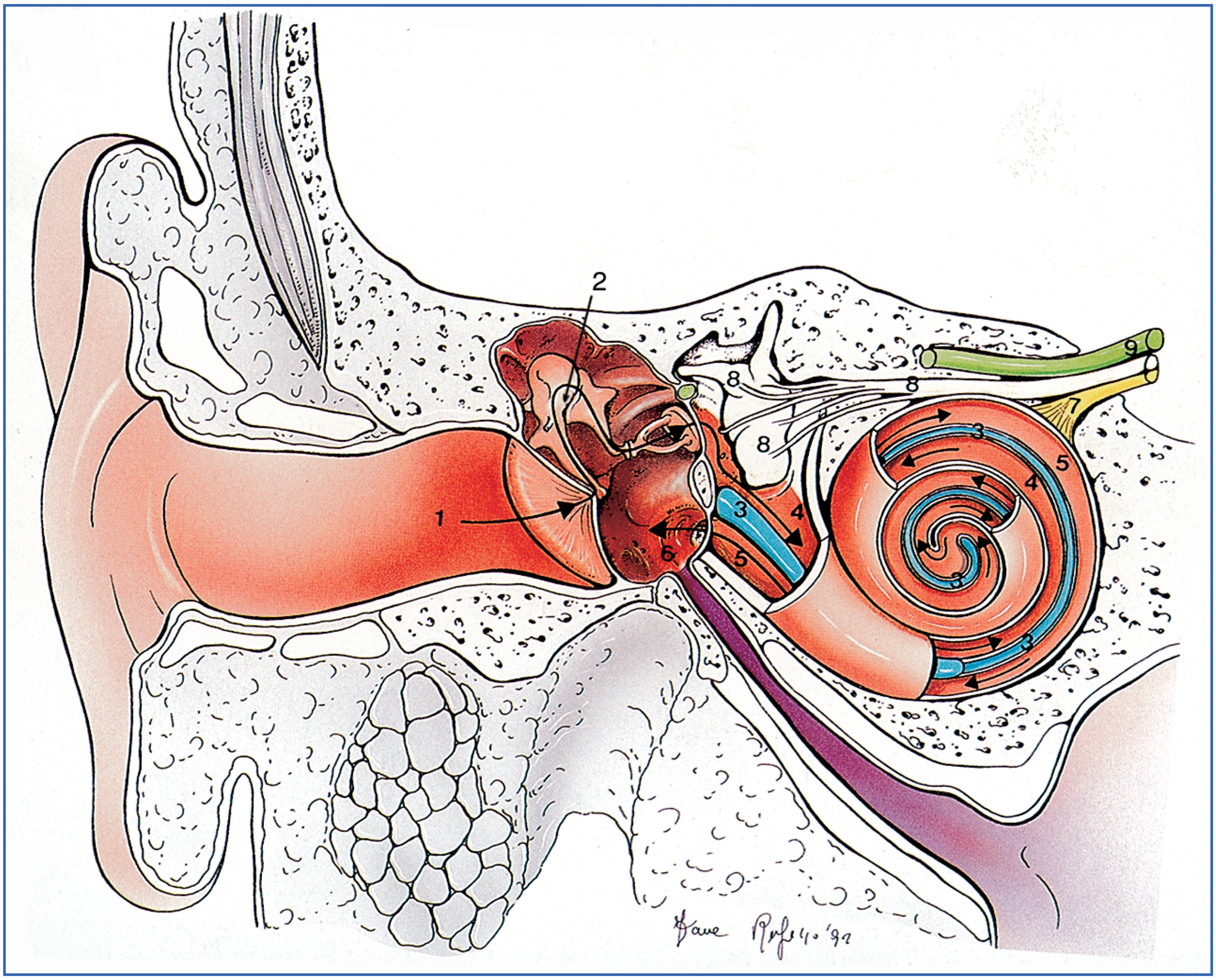


Fig. 1.7. Anatomical diagram of the ear: 1) external auditory canal; 2) ear drum and tympano-ossicular system; 3) cochlear canal; 4) scala vestibuli; 5) scala tympani; 6) round window with secondary tympanic membrane or Scarpa's membrane; 7) cochlear branch of the acoustic nerve; 8) vestibule and vestibular nerve; 9) facial nerve.

the medial two thirds. The narrow passage between the cartilaginous and the bony parts is called the high.

It is helical in shape and follows an oblique course curved forward and downward (**Fig. 1.9**). The canal is closed by the tympanic membrane and is lined with skin along its entire course, including the external surface of the tympanic membrane.

Middle ear

The middle ear is composed of (**Fig. 1.10**):

- auditory or Eustachian tube;
- ear drum;
- mastoid apparatus.

The Eustachian tube connects the rhinopharynx and the ear drum. It is composed of a bony canal in the third adjacent to the cavity of the middle ear and of a fibrocartilaginous canal in the two thirds near the rhinopharynx. Inserted on the external surface of the fibrocartilaginous part of the tube are the tensor and levator veli palatini muscles. At their other end, they insert on the soft palate and are innervated by the motor branches of the third branch of the trigeminus nerve. The ear drum (**Fig. 1.11**) is a flat fissure that has a biconcave lens shape; two walls, the lateral and the medial are distinguished; its circumference is subdivided in quadrants: anterior, posterior, superior, and inferior (**Fig. 1.12**). The lateral wall is composed nearly entirely of the tympanic which (**Fig. 1.13**) is elliptical in shape and is directed downward and forward, somewhat longer (9-10 mm) than wider (8-9 mm). It is formed by three layers:

- cutaneous, outer;
- fibrous, middle;
- mucosal, inner.

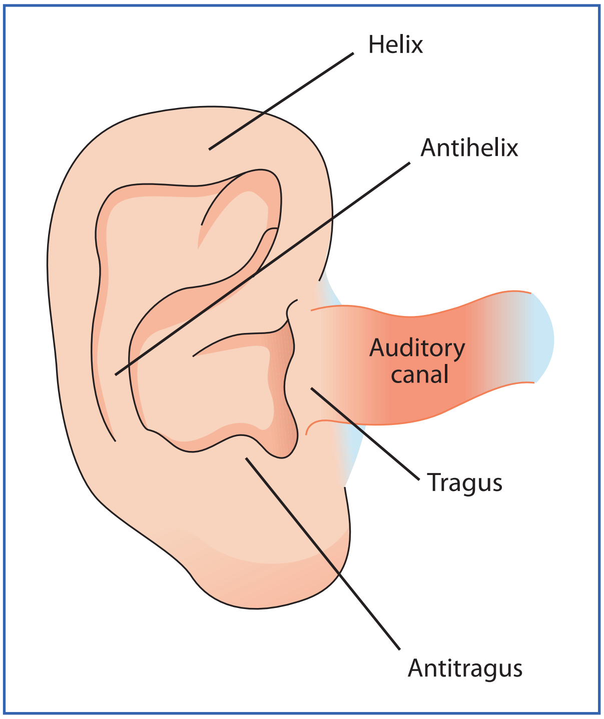


Fig. 1.8. Diagram of the lateral aspect of the auricle and its components.

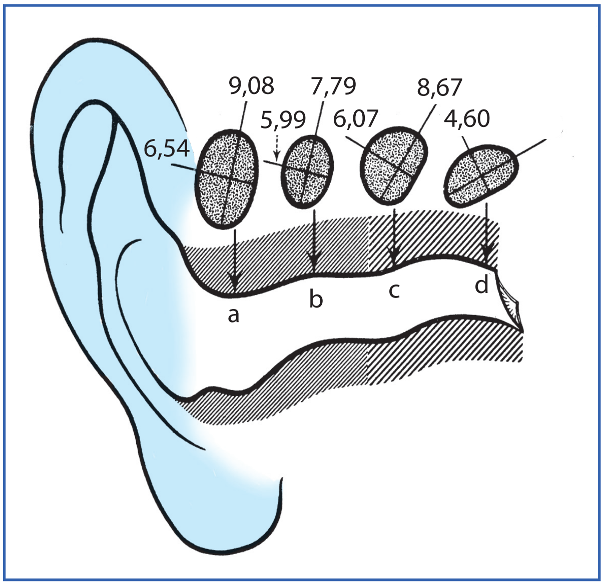


Fig. 1.9. Morphology of the external auditory canal and its diameter (in mm).

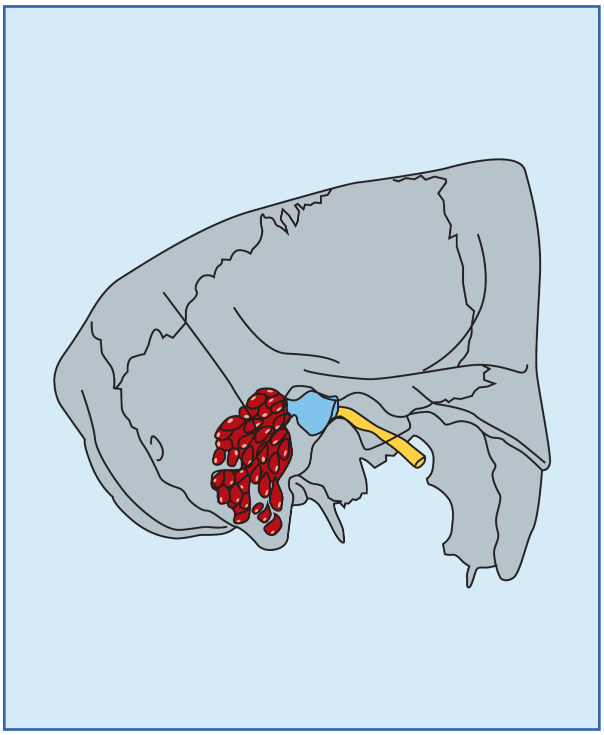


Fig. 1.10. Diagram of the middle ear with Eustachian tube (yellow), cavity of the middle ear (blue), and mastoid apparatus (red).

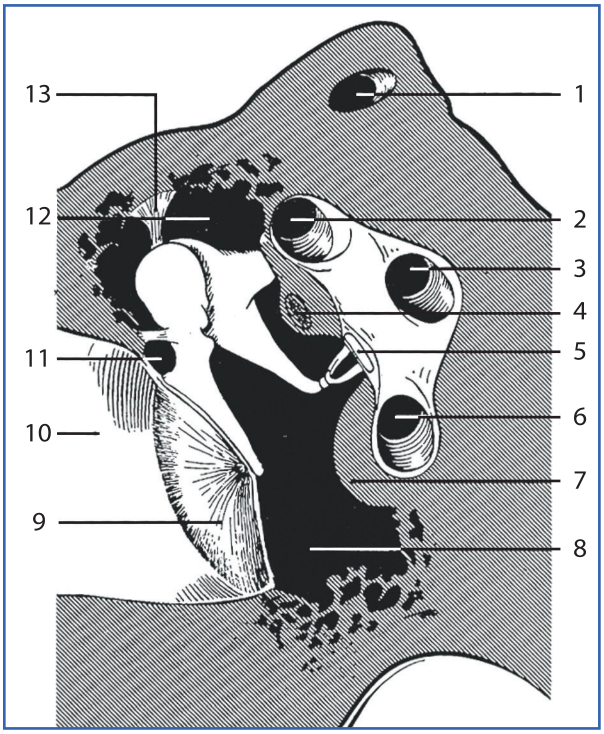


Fig. 1.11. Cutaway diagram of the ear drum, cavities (black) and ossicular chain and ligaments (white): 1) superior semicircular canal; 2) lateral semicircular canal; 3) posterior semicircular canal; 4) facial nerve; 5) stapes; 6) cochlear canaliculus; 7) promontory; 8) hypotympanum; 9) tympanic membrane; 10) external auditory canal; 11) recess of the pars flaccida; 12) epitympanum; 13) superior ligament of the hammer (source: Bairati).

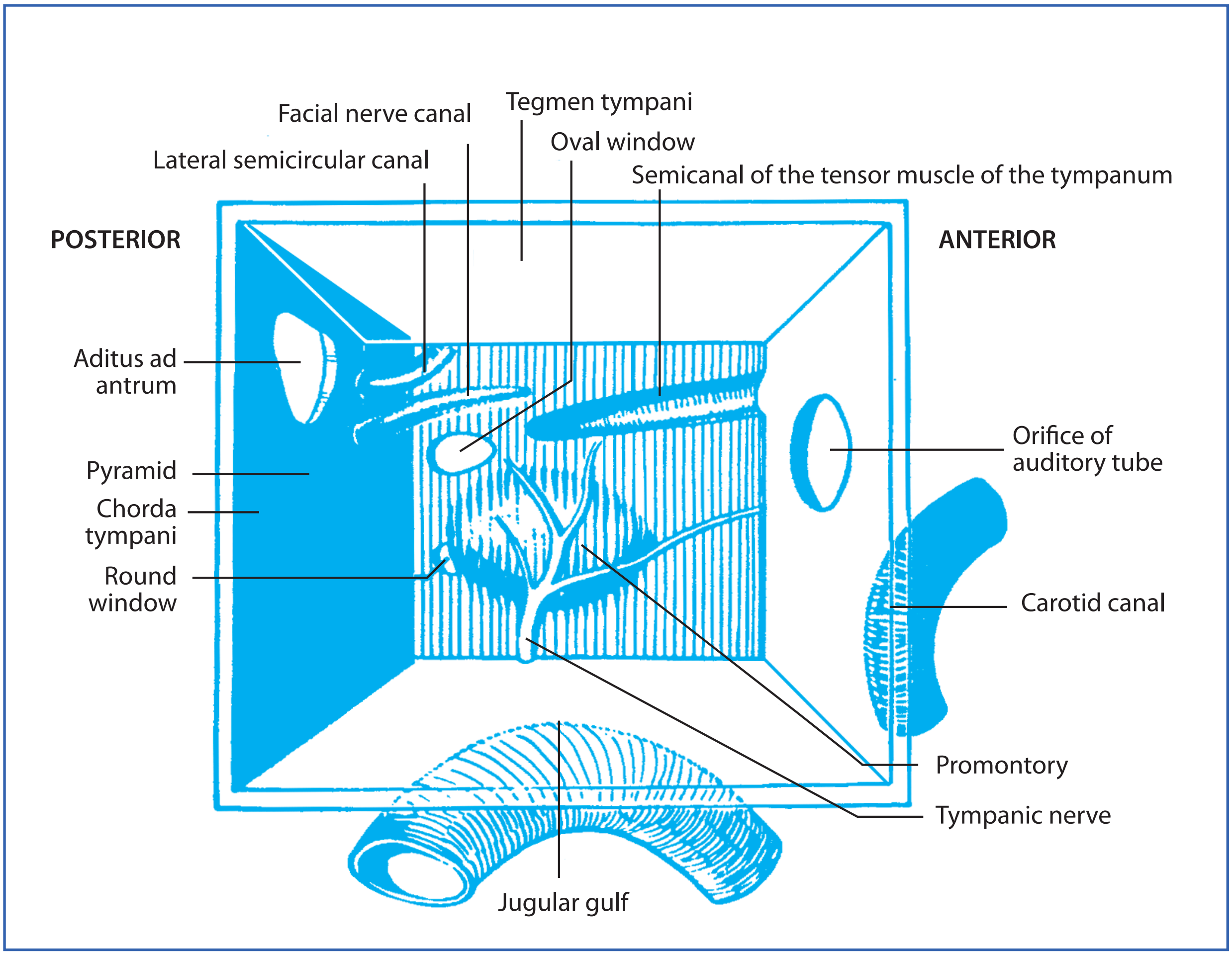


Fig. 1.12. Diagram of the ear drum and the components contained within its walls (source: Gardner et al.).

Two parts are distinguished:

- the *pars tensa* is larger and has a fibrous layer;
- the *pars flaccida*, the upper portion of the tympanic membrane, does not have a fibrous layer.

The handle of the hammer inserts in the upper central part of the *pars tensa.* The handle is directed upward and downward and slightly backward and forward.

The medial wall of the ear drum (**Fig. 1.12**) is rounded in the center, the so-called promontory, and corresponds to the basal turn of the cochlea. Its surface is grooved by the tympanic nerve, a branch of the glossopharyngeal nerve. Posterior and inferior to the promontory is the niche of the round window which communicates with the lower end of the tympanic scale of the cochlea and is closed by the secondary tympanic membrane. Superior to the round window is the oval window, an elliptical orifice where the footplate of the stapes is held in place by the annular ligament. Superior and posterior to the oval window is the arched prominence of the tympanic segment of the bony canal the covers the facial nerve (Fallopian aqueduct). The tympanic ostium of the auditory tube is present on the upper third of the anterior wall. The *aditus ad antrum*, an orifice leading to the tympanic antrum is located on the upper part of the posterior wall. The upper wall is composed of the *tegmen tympani*, a bony layer that separates the ear drum from the medial cranial fossa. The lower wall abuts the jugular gulf. The cavity of the middle ear can be subdivided in five areas (**Fig. 1.14**):

- *the atrium* is the part of the ear drum that can be examined through the external auditory canal; within the atrium are the handle of the hammer, part of the long process of the anvil, and the head of the stapes;
- *the epitympanum* (or *attic*) is located in the highest part of the ear drum and bordered laterally by the wall of the notch in which the *pars flaccida* of the membrane inserts; the epitympanum contains the

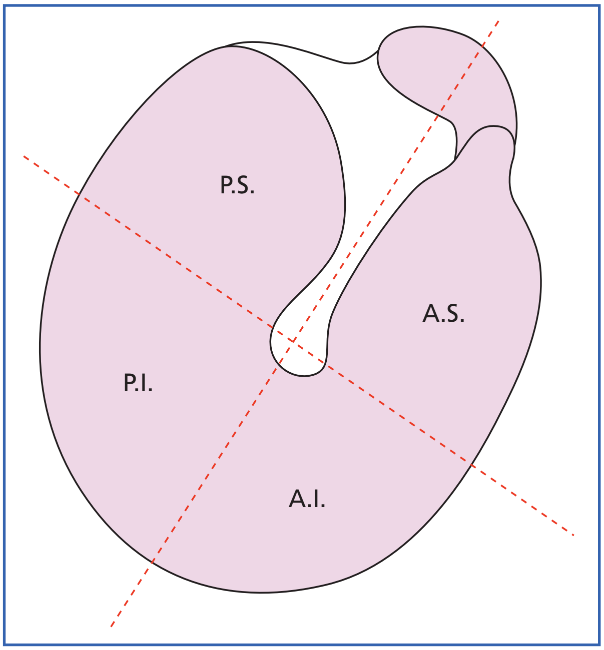


Fig. 1.13. Division of the tympanic membrane (right ear) in four quadrants.

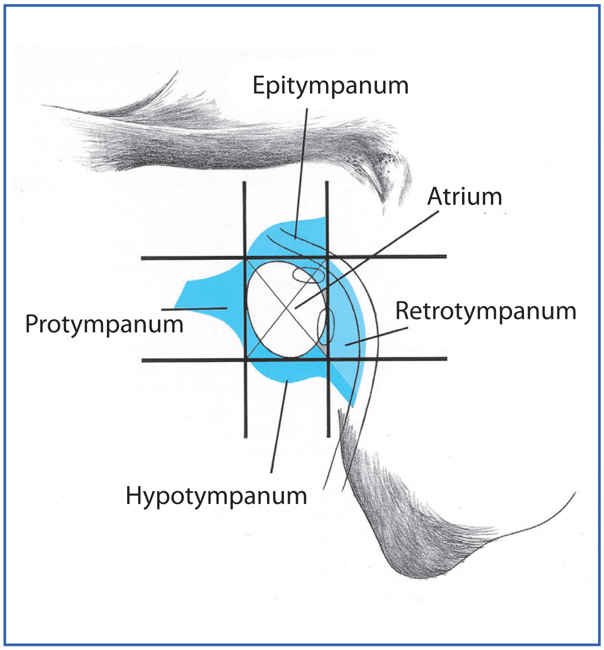


Fig. 1.14. Areas of the middle ear (left ear) (source: Nadol et al.).

head of the hammer and the body and short apophysis of the anvil;

- *the protympanum* is an empty cavity directed anteriorly in which Eustachian tube opens;
- *the hypotympanum*, is an empty cavity directed inferiorly;
- *the retrotympanum* is a morphologically complex cavity behind the ear drum; it contains the oval and the round window and the stapes.

Except for the atrium and sometimes part of the retrotympanum, none of the other areas of the middle ear can be examined though the external auditory canal but can be visualized only by imaging techniques or surgical exploration.

The ossicular chain (**Figs. 1.11, 1.15**) is composed of the hammer, the anvil, and the stapes. The posteromedial surface of the head of the hammer is elliptical in shape and provides the articular surface with the anvil. Beneath the head are the neck and then the handle that rests on the tympanic membrane. The anterior side of the anvil articulates with the hammer; the short apophysis inserted on the posterior side connects via a ligament to the posterior wall of the ear drum. The long apophysis is directed downward. On its terminal end is a lenticular process that articulates with the head of the staff. The head of the staff is composed of two small arches, the *crus anterior* and the *crus posterior*, which unite to form the footplate. The lateral side of the oval footplate is directed toward the ear drum; the medial side is directed toward the inner ear. The stapedial footplate rests on the oval window. Between the stapes and the oval window is a fibrous ring called the annular stapedial ligament. The ossicles are moved by two small muscles:

- the *tensor of the tympanum* that inserts medially on the neck of the hammer;
- the *stapedial* muscle that inserts on the *crus posterior* of the stapes.

The mastoid apparatus is composed of cells occupying the mastoid process. The largest cell, the antrum, communicates with tympanic through the *aditus ad antrum*. The remainder of the mastoid apparatus consists of collections of small bone cells around the antrum that occupy the mastoid process, extending to the root of the zygomatic bone and surrounding the bony labyrinth (**Fig. 1.16**).

The epithelium lining the middle ear is composed of mucosa. The tubular part has the same characteristics as the respiratory of the nose and the rhinopharynx: cylindrical, ciliated cells. The epithelium of the middle ear is composed of mucosa consisting of a layer of flat cells.

Inner ear

The inner ear is composed of a bony labyrinth and a membranous labyrinth and between the two is the perilymphatic space which contains perilymph.

The bony labyrinth is formed by a central cavity, the vestibulum from which three bony semicircular

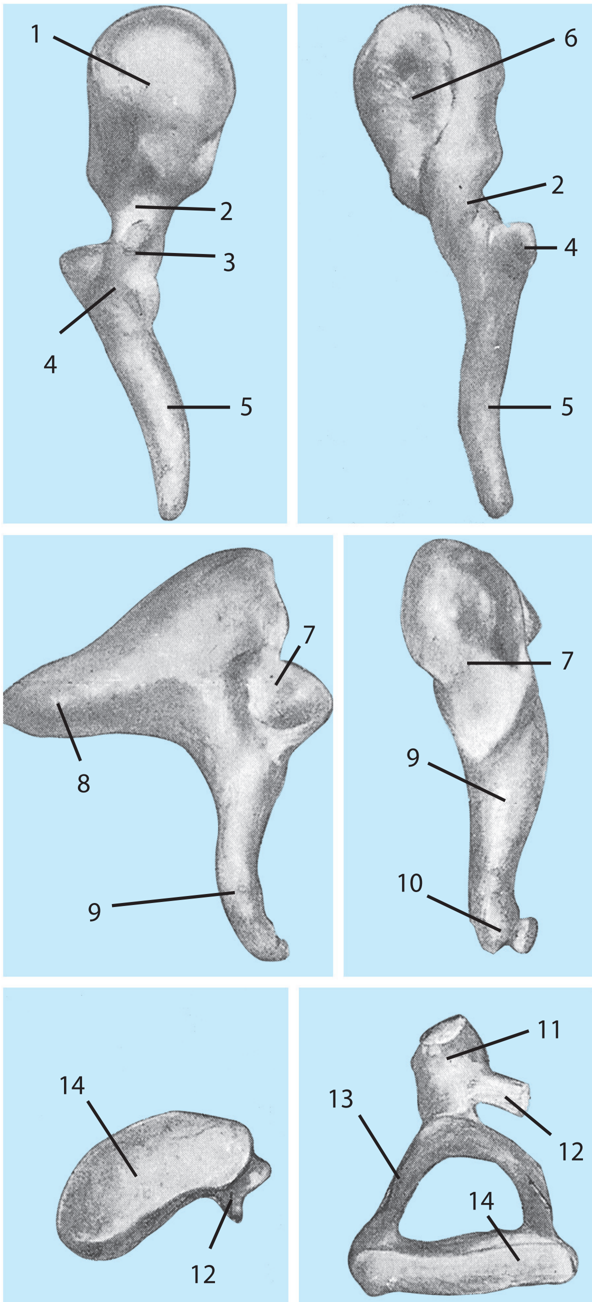


Fig. 1.15. Morphology of the ossicles of the middle ear. The panels show the same bone from two different views. From top to bottom: hammer, anvil, stapes: 1) head of the hammer; 2) neck of the hammer; 3) insertion of the anterior ligament on the hammer; 4) short process of the hammer; 5) handle of the hammer; 6) articular surface of the head of the hammer with the body of the anvil; 7) articular fossa of the anvil; 8) short process of the anvil; 9) long process of the anvil; 10) lenticular process of the anvil with the articular surface of the anvil and the articular surface of the head of the stapes; 11) head; 12) tendon of the stapes; 13) crura (anterior and posterior); 14) footplate.

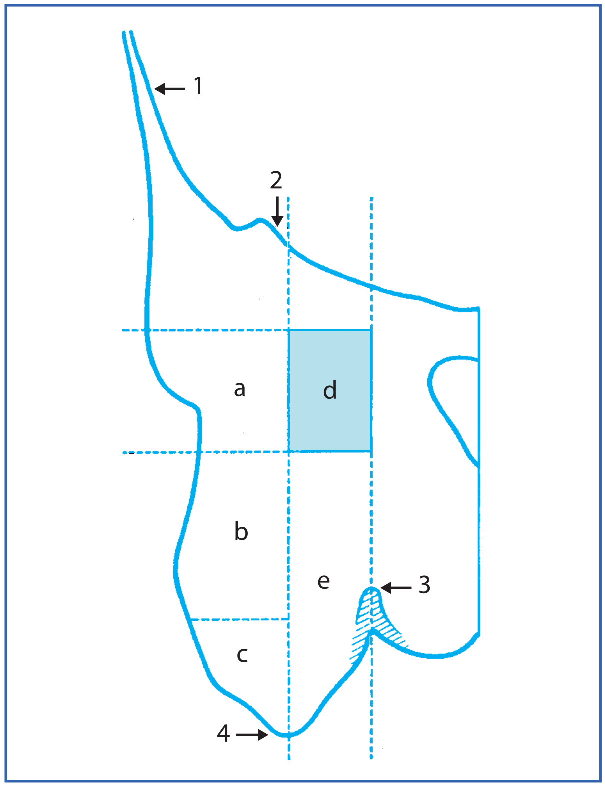


Fig. 1.16. Diagram of the mastoid apparatus: 1) squama of the temporal bone; 2) division between superficial and deep mastoid cells; 3) digastric groove; 4) apex of the mastoid: a) superficial mastoid antrum cells; b) superficial cells of the subantrum; c) cells of the point; d) mastoid antrum; e) deep cells of the subantrum (intersinuso-facial).

canals originate posteriorly and the spiral canal of the cochlea anteriorly. The bony labyrinth communicates laterally with the ear drum via the oval and the round windows.

The bony cochlea is formed by a conical bone block about 1 cm long, inside which is the spiral canal of the cochlea. The modiolus is a central axis. The cochlea turns about 3 times (basal, intermediate and apical around the axis. The inside the bony canal is a bony prominence, the boney spiral lamina, upon which the membranous structures of the cochlea insert (**Fig. 1.17**).

In the basal part of the spiral canal the cochlear aqueduct opens, the narrow, long canal at the medial end of which is the lower margin of the rocca petrosa at the level of the posterior cranial fossa. The aqueduct connects the perilymphatic space with the subarachnoid spaces that contain cerebrospinal fluid.

The membranous labyrinth (cochlear canal), situated inside the cochlea (about 36 mm long) terminates near the apex of the cochlea in a cul de sac. It contains *endolymph*. The cochlear canal divides the