Photoreceptor & Audioreceptor Systems

The Eye

The eye is a complex and highly developed photosensitive organ that permits an accurate analysis of the form, light intensity, and color reflected from objects. The eyes are located in protective bony structures of the skull, the **orbits.** Each eye includes a tough, fibrous globe to maintain its shape, a lens system to focus the image, a layer of photosensitive cells, and a system of cells and nerves whose function is to collect, process, and transmit visual information to the brain. Each eye is composed of three concentric layers: an external layer that consists of the **sclera** and the **cornea;** a middle layer also called the **vascular layer** consisting of the **choroid, ciliary body,** and **iris;** and an inner layer of nerve tissue, the **retina,** which consists of an outer pigment epithelium and an inner retina proper. The photosensitive retina proper is part of the central nervous system and communicates with the cerebrum through the **optic nerve** and extends forward to the **ora serrata.** The optic nerve (and the retina) arises in the embryo as an evagination of the prosencephalon. Consequently, it is not a true peripheral nerve like the other cranial nerves. Because it is a tract of the central nervous system, the myelin of its nerve fibers is produced by oligodendrocytes, not by Schwann cells. This may explain the visual dysfunction often associated with multiple sclerosis, a demyelinating disorder of the central nervous system.

The **lens** of the eye is a biconvex transparent structure held in place by a circular system of fibers, the **zonule,** that extends from the lens into a thickening of the middle layer, the **ciliary body,** and, by close apposition, to the vitreous body on its posterior side. Partly covering the anterior surface of the lens is an opaque pigmented expansion of the middle layer called the **iris.** The round hole in the middle of the iris is the **pupil**.

The eye contains three compartments: the **anterior chamber,** which occupies the space between the cornea and the iris and lens; the **posterior chamber,** between the iris, ciliary process, zonular attachments, and lens; and the **vitreous space,** which lies behind the lens and zonular attachments and is surrounded by the retina. Both the anterior and posterior chambers contain a protein-poor fluid called **aqueous humor.** The vitreous space is filled with a gelatinous substance called the **vitreous body.**

*Note that the terms **outer** (**external**) and **inner** (**internal**) refer to the gross structure of the eye. Inner denotes a structure closer to the center of the globe, whereas outer means closer to the surface of the eyeball.*

External Layer, or Tunica Fibrosa

The opaque white posterior five-sixths of the external layer of the eye is the **sclera**; in humans, this forms a segment of a sphere approximately 22 mm in diameter. The sclera consists of tough, dense connective tissue made up mainly of flat collagen bundles intersecting in various directions while remaining parallel to the surface of the organ, a moderate amount of ground substance, and a few fibroblasts. The external surface of the sclera, the **episclera** is connected by a loose system of thin collagen fibers to a dense layer of connective tissue called **Tenon's capsule.** Tenon's capsule comes into contact with the loose conjunctival stroma at the junction of the cornea with the sclera. Between Tenon's capsule and the sclera is **Tenon's space.** Because of this loose space, the eyeball can make rotating movements. Between the sclera and the choroid is the **suprachoroidal lamina,** a thin layer of loose connective tissue rich in melanocytes, fibroblasts, and elastic fibers. The sclera is relatively avascular.

In contrast to the posterior five-sixths of the eye, the anterior one-sixth,

the **cornea** : Is colorless and transparent. A transverse section of the cornea shows that it consists of five layers: epithelium, Bowman's membrane, stroma, Descemet's membrane, and endothelium. The corneal epithelium is stratified, squamous, and nonkeratinized and consists of five or six layers of cells. In the basal part of the epithelium are numerous mitotic figures that are responsible for the cornea's remarkable regenerative capacity: The turnover time for these cells is approximately 7 days. The surface corneal cells show microvilli protruding into the space filled by the precorneal tear film.

1-This epithelial tissue is covered by a protective layer of lipid and glycoprotein, about 7 µm thick. The cornea has one of the richest sensory nerve supplies of any eye tissue.

2- Beneath the corneal epithelium lies a thick homogeneous layer 7-12 µm thick. This layer, **Bowman's membrane,** consists of collagen fibers crossing at random, a condensation of the intercellular substance, and no cells. Bowman's membrane contributes greatly to the stability and strength of the cornea.

3-The **stroma** is formed of many layers of parallel collagen bundles that cross at approximately right angles to each other. The collagen fibrils within each lamella are parallel to each other and run the full width of the cornea. Between the several layers, the cytoplasmic extensions of fibroblasts are flattened like the wings of a butterfly. Both cells and fibers of the stroma are immersed in a substance rich in glycoproteins and chondroitin sulfate. Although the stroma is avascular, migrating lymphoid cells are normally present in the cornea.

4- Descemet's membrane is a thick (5-10 µm) homogeneous structure composed of fine collagenous filaments organized in a three-dimensional network.

5- The **endothelium** of the cornea is a simple squamous epithelium. These cells possess organelles for secretion that are characteristic of cells engaged in active transport and protein synthesis and that may be related to the synthesis and maintenance of Descemet's membrane. The corneal endothelium and epithelium are responsible for maintaining the transparency of the cornea. Both layers are capable of transporting sodium ions toward their apical surfaces. Chloride ions and water follow passively, maintaining the corneal stroma in a relatively dehydrated state. This state, along with the regular orientation of the very thin collagen fibrils of the stroma, accounts for the transparency of the cornea.

The **corneoscleral junction,** or **limbus,** is an area of transition from the transparent collagen bundles of the cornea to the white opaque fibers of the sclera. It is highly vascularized, and its blood vessels assume an important role in corneal inflammatory processes. The cornea, an avascular structure, receives its metabolites by diffusion from adjacent vessels and from the fluid of the anterior chamber of the eye. In the region of the limbus in the stromal layer, irregular endothelium-lined channels, the trabecular meshwork, merge to form **Schlemm's canal**, which drains fluid from the anterior chamber of the eye. Schlemm's canal communicates externally with the venous system.

Middle, or Vascular, Layer

The middle (vascular) layer of the eye consists of three parts: choroid, ciliary body, and iris, known collectively as the uveal tract.

Choroid

The choroid is a highly vascularized coat, with loose connective tissue between its blood vessels that is rich in fibroblasts, macrophages, lymphocytes, mast cells, plasma cells, collagen fibers, and elastic fibers. Melanocytes are abundant in this layer and give it its characteristic black color. The inner layer of the choroid is richer than the outer layer in small vessels and is called the **choriocapillary layer.** It has an important function in nutrition of the retina, and damage to this tissue causes serious damage to the retina. A thin (3-4 µm) hyaline membrane separates the choriocapillary layer from the retina. This is known as **Bruch's membrane** and extends from the optic papilla to the ora serrata. The **optic papilla** is the region at which the optic nerve enters the eyeball.

Bruch's membrane is formed of five layers. The central layer is composed of a network of elastic fibers. This network is lined on its two surfaces with layers of collagen fibers that are covered by the basal lamina of the capillaries of the choriocapillary layer on one side and the basal lamina of the pigment epithelium on the other side. (See Retina, below, for a description of the pigment epithelium.) The choroid is bound to the sclera by the **suprachoroidal lamina,** a loose layer of connective tissue rich in melanocytes.

Ciliary Body

The ciliary body, an anterior expansion of the choroid at the level of the lens, is a continuous thickened ring that lies at the inner surface of the anterior portion of the sclera; in transverse section, it forms a triangle. One of its faces is in contact with the vitreous body, one with the sclera, and the third with the lens and the posterior chamber of the eye. The histological structure of the ciliary body is basically loose connective tissue (rich in elastic fibers, vessels, and melanocytes) surrounding the **ciliary muscle** . This structure consists of two bundles of smooth muscle fibers that insert on the sclera anteriorly and on different regions of the ciliary body posteriorly. One of these bundles has the function of stretching the choroid; another bundle, when contracted, relaxes the tension on the lens. These muscular movements are important in visual accommodation. The surfaces of the ciliary body that face the vitreous body, posterior chamber, and lens are covered by the anterior extension of the retina. In this region, the retina consists of only two cell layers. The layer directly adjacent to the ciliary body consists of simple columnar cells rich in melanin and corresponds to the forward projection of the pigment layer of the retina. The second layer, which covers the first, is derived from the sensory layer of the retina and consists of simple nonpigmented columnar epithelium.

Ciliary Processes

The ciliary processes are ridgelike extensions of the ciliary body. They have a loose connective tissue core and numerous fenestrated capillaries and are covered by the two simple epithelial layers described above. From the ciliary processes emerge oxytalan fibers (**zonule fibers**) that insert into the capsule of the lens and anchor it in place. The apical ends of the epithelial cells are found at the junction between pigmented and nonpigmented cells, and the cells thus meet each other head-to-head. The zonular fibers have their origin in the basement membrane of the inner cells. The apical ends of the epithelial cells are joined by desmosomes, and elaborate tight junctions are found around the apical surfaces of epithelial cells of both layers. The nonpigmented inner layer of cells has extensive basal infoldings and interdigitations characteristic of ion-transporting cells. These cells actively transport certain constituents of plasma into the posterior chamber, thus forming the **aqueous humor.** This fluid has an inorganic ion composition similar to that of plasma but contains less than 0.1% protein (plasma has about 7% protein). Aqueous humor flows toward the lens and passes between it and the iris, reaching the anterior chamber of the eye. Once in the anterior chamber, the humor proceeds to the angle formed by the cornea with the basal part of the iris. It penetrates the tissue of the limbus in a series of labyrinthine spaces (the trabecular meshwork) and finally reaches the irregular Schlemm's canal, lined with endothelial cells. This structure communicates with small veins of the sclera, through which the aqueous humor escapes.

Iris

The iris is an extension of the choroid that partially covers the lens, leaving a round opening in the center called the **pupil**. The anterior surface of the iris is irregular and rough, with grooves and ridges. It is formed of a discontinuous layer of pigment cells and fibroblasts. Beneath this layer is a poorly vascularized connective tissue with few fibers and many fibroblasts and melanocytes. The next layer is rich in blood vessels embedded in loose connective tissue. The smooth posterior surface of the iris is covered by two layers of epithelium, which also cover the ciliary body and its processes. The inner epithelium, in contact with the posterior chamber, is heavily pigmented with melanin granules. The outer epithelial cells have radially directed tonguelike extensions of their basal region; they are filled with overlapping myofilaments, creating the **dilator pupillae muscle** of the iris. The heavy pigmentation prevents the passage of light into the interior of the eye except through the pupil.

The function of the abundant melanocytes or pigment cells containing melanin in several regions of the eye is to keep stray light rays from interfering with image formation. The melanocytes of the stroma of the iris are responsible for the color of the eyes. If the layer of pigment in the interior region of the iris consists of only a few cells, the light reflected from the black pigment epithelium in the posterior surface of the iris will be blue. As the amount of pigment increases, the iris assumes various shades of greenish-blue, gray, and finally brown. Albinos have almost no pigment, and the pink color of their irises is due to the reflection of incident light from the blood vessels of the iris.

The iris contains smooth muscle bundles disposed in circles concentric with the pupillary margin, forming the **sphincter pupillae muscle** of the iris. The dilator and sphincter muscles have sympathetic and parasympathetic innervation, respectively.

Lens

The lens is a biconvex structure characterized by great elasticity, a feature that is lost with age as the lens hardens. The lens has three principal components.

Lens Capsule The lens is enveloped by a thick (10-20 µm), homogeneous, refractile, carbohydrate-rich capsule coating the outer surface of the epithelial cells. It is a very thick basement membrane and consists mainly of collagen type IV and glycoprotein.

Subcapsular Epithelium

Subcapsular epithelium consists of a single layer of cuboidal epithelial cells that is present only on the anterior surface of the lens. The lens increases in size and grows throughout life as new lens fibers develop from cells located at the equator of the lens. The cells of this epithelium exhibit many interdigitations with the lens fibers.

Lens Fibers

Lens fibers are elongated and appear as thin, flattened structures. They are highly differentiated cells derived from cells of the subcapsular epithelium. Lens fibers eventually lose their nuclei and other organelles and become greatly elongated, attaining dimensions of 7-10 mm in length, 8-10 µm in width, and 2 µm in thickness. These cells are filled with a group of proteins called **crystallins.** Lens fibers are produced throughout life, at an ever-decreasing rate.

The lens is held in place by a radially oriented group of fibers, the **zonule,** that inserts on one side on the lens capsule and on the other on the ciliary body. Zonular fibers are similar to the microfibrils of elastic fibers. This system is important in the process known as **accommodation,** which permits focusing on near and far objects by changing the curvature of the lens. When the eye is at rest or gazing at distant objects, the lens is kept stretched by the zonule in a plane perpendicular to the optical axis. To focus on a near object, the ciliary muscles contract, causing forward displacement of the choroid and ciliary body. The tension exerted by the zonule is relieved, and the lens becomes thicker, keeping the object in focus.

Vitreous Body

The vitreous body occupies the region of the eye behind the lens. It is a transparent gel that consists of water (about 99%), a small amount of collagen, and heavily hydrated hyaluronic acid molecules. The vitreous body contains very few cells, which synthesize collagen and hyaluronic acid.

Retina

The retina, the inner layer of the globe, consists of two portions. The posterior portion is photosensitive; the anterior part, which is not photosensitive, constitutes the inner lining of the ciliary body and the posterior part of the iris. The retina derives from an evagination of the anterior cephalic vesicle, or prosencephalon. As this **optic vesicle** comes into contact with the surface ectoderm, it gradually invaginates in its central region, forming a double-walled **optic cup.** In adults, the outer wall gives rise to a thin membrane called the **pigment epithelium;** the optical or functioning part of the retina the **neural retina** is derived from the inner layer.

1-The pigment epithelium consists of columnar cells with a basal nucleus. The basal regions of the cells adhere firmly to Bruch's membrane, and the cell membranes have numerous basal invaginations. Mitochondria are more abundant in the region of the cytoplasm near these invaginations. These characteristics suggest an ion-transporting activity for this region.

The lateral cell membranes show cell junctions with conspicuous zonulae occludentes and zonulae adherentes at their apexes; there are also desmosomes and gap junctions. These morphological characteristics indicate that the apical and basal regions of this epithelial sheet are sealed off and that there is intercellular communication. These junctional specializations account for the electrical potential difference that results from ion transport between the two surfaces of this epithelium.

The cell apex has abundant extensions of two types: slender microvilli and cylindrical sheaths that envelop the tips of the photoreceptors.

The cytoplasm of pigment epithelial cells has abundant smooth endoplasmic reticulum, believed to be a site of vitamin A esterification and transport to the photoreceptors. Melanin granules are numerous in the apical cytoplasm and microvilli. Melanin is synthesized in these cells by a mechanism similar to that described for the melanocytes in the skin. This dark pigment has the function of absorbing light after the photoreceptors have been stimulated.

The cell apex has numerous dense vesicles of variable shape that represent various stages in the phagocytosis and digestion of the tips of photoreceptor outer segments.

2- The optical part of the retina the posterior, or photosensitive, part is a complex structure containing at least 15 types of neurons, and these cells form at least 38 distinct kinds of synapses with one another. The optical retina consists of an outer layer of photosensitive cells, the **rods** and **cones**; an intermediate layer of **bipolar neurons,** which connects the rods and cones to the **ganglion cells;** and an internal layer of ganglion cells, which establishes contact with the bipolar cells through their dendrites and sends axons to the brain. These axons converge at the optic papilla, forming the **optic nerve.**

Between the layer of rods and cones and the bipolar cells is a region called the **external plexiform,** or **synaptic, layer,** at which synapses between these two types of cells occur. The region at which the synapses between the bipolar and ganglion cells are established is called the **internal plexiform layer**. The retina has an inverted structure, for the light will first cross the ganglion layer and then the bipolar layer to reach the rods and cones. The structure of the retina will now be examined in greater detail.

The rods and cones, named for the forms they assume, are polarized neurons; at one pole is a single photosensitive dendrite, and at the other are synapses with cells of the bipolar layer. The rod and cone cells can be divided into outer and inner segments, a nuclear region, and a synaptic region. The outer segments are modified cilia and contain stacks of membrane-limited saccules with a flattened, disklike shape. The photosensitive pigment of the retina is in the membranes of these saccules. Both rod and cone cells pass through a thin layer, the **external limiting membrane,** that is a series of junctional complexes between the photoreceptors and glial cells of the retina (MÃ¼ller cells). The nuclei of the cones are generally disposed near the limiting membrane, whereas the nuclei of the rods lie near the center of the inner segment.

Rod Cells

Rod cells are thin, elongated cells (50 x 3 μm) composed of two portions. The external photosensitive rod-shaped portion is composed mainly of numerous (600-1000) flattened membranous disks stacked up like coins. The disks in rods are not continuous with the plasma membrane; the **outer segment** is separated from the **inner segment** by a constriction. Just below this constriction is a basal segment from which a cilium arises and passes to the outer segment. The inner segment is rich in glycogen and has a remarkable accumulation of mitochondria, most of which lie near the constriction. This local accumulation of mitochondria is related to the production of energy necessary for the visual process and protein synthesis. Polyribosomes, present in large numbers below the mitochondrial region of the inner segment, are involved in protein synthesis. Some of these proteins migrate to the outer segment of the rod cells, where they are incorporated into membranous disks. The flattened disks of the rod cells contain the pigment **visual purple,** or **rhodopsin,** which is bleached by light and initiates the visual stimulus. This substance is globular and is located in the outer surface of the lipid bilayer of the flattened membranous disks.

The human retina has approximately 120 million rods. They are extremely sensitive to light and are considered to be the receptors that are used when low levels of light are encountered, such as at dusk or nighttime. The outer segment is the site of photosensitivity; the inner segment contains the metabolic machinery necessary for the biosynthetic and energy-producing processes of these cells.

Autoradiographic studies show that proteins of the rod vesicles are synthesized in the polyribosome-rich inner segments of these cells. From there, they migrate to the outer segment and aggregate at its basal region, where they are incorporated into membranes formed by a double layer of phospholipids, producing flattened disks. These structures gradually migrate to the cell apex, where they are shed, phagocytosed, and digested by the cells of the pigment epithelium. It has been calculated that in the monkey, approximately 90 vesicles per cell are produced daily.

Cone Cells

Cone cells are also elongated (60 x 1.5 µm) neurons. Each human retina has about 6 million cone cells. The structure is similar to that of rods, with outer and inner segments, a basal body with cilium, and an accumulation of mitochondria and polyribosomes. Cones differ from rods in their form (conical) and the structure of their outer segments. As in rods, this region is composed of stacked membranous disks; however, they are not independent of the outer plasma membrane but arise as invaginations of this structure. In cones, newly synthesized protein is not concentrated in recently assembled disks, as it is in rods, but is distributed uniformly throughout the outer segment.

There are at least three functional types of cones that cannot be distinguished by their morphological characteristics. Each type contains a variety of the cone photopigment called **iodopsin,** and its maximum sensitivity is in the red, green, or blue region of the visible spectrum. Cones, sensitive only to light of an intensity higher than required to stimulate rods, are believed to permit better visual acuity than do rods.

Other Cells

The layer of bipolar cells consists of two types of cells: **diffuse bipolar cells,** which have synapses with two or more photoreceptors; and **monosynaptic bipolar cells,** which establish contact with the axon of only one cone photoreceptor and only one ganglion cell. Certain numbers of cones therefore transmit their impulses directly to the brain.

In addition to establishing contact with the bipolar cells, the cells of the ganglion layer project their axons to a specific region of the retina, where they come together to form the **optic nerve**. This region, which is devoid of receptors, is known as the **blind spot** of the retina, the **papilla of the optic nerve,** or the **optic nerve head**. The **ganglion cells** are typical nerve cells, containing a large euchromatic nucleus and basophilic Nissl bodies. These cells, like the bipolar cells, are classified as diffuse or monosynaptic in their connections with other cells.

In addition to these three main types of cells (photoreceptor, bipolar, and ganglion), there are other types of cells that are distributed more diffusely in the layers of the retina.

Horizontal cells establish contact between different photoreceptors. Their exact function is not known, but they may act to integrate stimuli.

Amacrine cells are various types of neurons that establish contact between the ganglion cells. Their function is also obscure.

Supporting cells are neuroglia that possess, in addition to the astrocyte and microglial cell types, some large, extensively ramified cells (**Mller cells**). The processes of these cells bind the neural cells of the retina and extend from the internal to the external limiting membranes of the retina. The external limiting membrane is a zone of adhesion (tight junctions) between photoreceptors and Mller cells. Mller cells are functionally analogous to neuroglia in that they support, nourish, and insulate the retinal neurons and fibers.

Accessory Structures of the Eye

Conjunctiva

The conjunctiva is a thin, transparent mucous membrane that covers the anterior portion of the eye up to the cornea and the internal surface of the eyelids. It has a stratified columnar epithelium with numerous goblet cells, and its lamina propria is composed of loose connective tissue.

Eyelids

Eyelids are movable folds of tissue that protect the eye. The skin of the lids is loose and elastic, permitting extreme swelling and subsequent return to normal shape and size.

The three types of glands in the lid are the **Meibomian glands** and **the glands of Moll** and **Zeis**. The Meibomian glands are long sebaceous glands in the tarsal plate. They do not communicate with the hair follicles. ***The Meibomian glands*** produce a sebaceous substance that creates an oily layer on the surface of the tear film, helping to prevent rapid evaporation of the normal tear layer. ***The glands of Zeis*** are smaller, modified sebaceous glands connected to the follicles of the eyelashes. ***The sweat glands of Moll*** are unbranched sinuous tubules that begin in a simple spiral and not in a glomerulus like ordinary sweat glands. They empty their secretion into the follicles of the eyelashes.

Lacrimal Apparatus

The lacrimal apparatus consists of the lacrimal gland, canaliculi, lacrimal sac, and nasolacrimal duct. The **lacrimal gland** is a tear-secreting gland located in the anterior superior temporal portion of the orbit. It consists of several separate glandular lobes with 6-12 excretory ducts that connect the gland to the superior conjunctival fornix. The lacrimal gland is a tubuloalveolar gland that usually has distended lumens and is composed of column-shaped cells of the serous type, resembling the parotid acinar cells. These cells show lightly stained secretory granules, and a basal lamina separates them from the surrounding connective tissue.

Well-developed myoepithelial cells surround the secretory portions of the lacrimal gland. The secretion of the gland passes down over the cornea and the bulbar and palpebral conjunctiva, moistening the surfaces of these structures. It drains into the **lacrimal canaliculi** through the **lacrimal puncta,** which are round apertures about 0.5 mm in diameter on the medial aspect of both the upper and lower lid margins. The canaliculi, which are about 1 mm in diameter and 8 mm long and join to form a common canaliculus just before opening into the lacrimal sac, are lined with a thick stratified squamous epithelium. Diverticuli of the common canaliculus, which may be part of the normal structure, are frequently susceptible to infections.

The lacrimal glands secrete a fluid rich in lysozyme, an enzyme that hydrolyzes the cell walls of certain species of bacteria, facilitating their destruction.

The Ear

The functions of the vestibulocochlear apparatus are related to equilibrium and hearing. The organ consists of three parts: the **external ear,** which receives sound waves; the **middle ear,** in which sound waves are transmitted from air to bone and by bone to the internal ear; and the **internal ear,** in which these vibrations are transduced to specific nerve impulses that pass via the acoustic nerve to the central nervous system. The internal ear also contains the vestibular organ, which maintains equilibrium.

External Ear

The **auricle** (**pinna**) consists of an irregularly shaped plate of elastic cartilage covered by tightly adherent skin on all sides.

The **external auditory meatus** is a somewhat flattened canal extending from the surface into the temporal bone. Its internal limit is the tympanic membrane. A stratified squamous epithelium continuous with the skin lines the canal. Hair follicles, sebaceous glands, and the **ceruminous glands** (a type of modified sweat gland) are found in the submucosa. Ceruminous glands are coiled tubular glands that produce the cerumenâ€”or earwaxâ€”a brownish, semisolid mixture of fats and waxes. Hairs and cerumen probably have a protective function. The wall of the external auditory meatus is supported by elastic cartilage in its outer third, whereas the temporal bone provides support for the inner part of the canal.

Across the deep end of the external auditory meatus lies an oval membrane, the **tympanic membrane** (eardrum). Its external surface is covered with a thin layer of epidermis, and its inner surface is covered with simple cuboidal epithelium continuous with the lining of the tympanic cavity. Between the two epithelial coverings is a tough connective tissue layer composed of collagen and elastic fibers and fibroblasts. The tympanic membrane is the structure that transmits sound waves to the ossicles of the middle ear.

Middle Ear

The middle ear, or tympanic cavity, is an irregular space that lies in the interior of the temporal bone between the tympanic membrane and the bony surface of the internal ear. It communicates anteriorly with the pharynx via the **auditory tube** (**eustachian tube**) and posteriorly with the air-filled cavities of the mastoid process of the temporal bone. The middle ear is lined with simple squamous epithelium resting on a thin lamina propria that is strongly adherent to the subjacent periosteum. Near the auditory tube and in its interior, the simple epithelium that lines the middle ear is gradually transformed into ciliated pseudostratified columnar epithelium. Although the walls of the tube are usually collapsed, the tube opens during the process of swallowing, balancing the pressure of the air in the middle ear with atmospheric pressure. In the medial bony wall of the middle ear are two membrane-covered oblong regions devoid of bone; these are the **oval** and **round windows**.

The tympanic membrane is connected to the oval window by a series of three small bones, the **auditory ossicles** the **malleus, incus,** and **stapes** that transmit the mechanical vibrations generated in the tympanic membrane to the internal ear. The malleus inserts itself into the tympanic membrane and the stapes into the membrane of the oval window. These bones are articulated by synovial joints and, like all structures of this cavity, are covered with simple squamous epithelium. In the middle ear are two small muscles that insert themselves into the malleus and stapes. They have a function in regulating sound conduction.

Internal Ear

The internal ear is composed of two **labyrinths.** The **bony labyrinth** consists of a series of spaces within the petrous portion of the temporal bone that houses the **membranous labyrinth**. The membranous labyrinth is a continuous epithelium-lined series of cavities of ectodermal origin. It derives from the auditory vesicle that is developed from the ectoderm of the lateral part of the embryo's head. During embryonic development, this vesicle invaginates into the subjacent connective tissue, loses contact with the cephalic ectoderm, and moves deeply into the rudiments of the future temporal bone. During this process, it undergoes a complex series of changes in form, giving rise to two specialized regions of the membranous labyrinth: the **utricle** and the **saccule.** The **semicircular ducts** originate from the utricle, whereas the elaborate **cochlear duct** is formed from the saccule. In each of these areas, the epithelial lining becomes specialized to form sensory structures such as the **maculae** of the utricle and saccule, the **cristae** of the semicircular ducts, and the **organ of Corti** of the cochlear duct.

The **bony labyrinth** consists of spaces in the temporal bone. There is an irregular central cavity, the **vestibule,** housing the saccule and the utricle. Behind this, three **semicircular canals** enclose the semicircular ducts; the anterolateral **cochlea** contains the cochlear duct.

The cochlea, about 35 mm in total length, makes two-and-one-half turns around a bony core known as the **modiolus.** The modiolus has spaces containing blood vessels and the cell bodies and processes of the acoustic branch of the eighth cranial nerve (spiral ganglion). Extending laterally from the modiolus is a thin bony ridge, the **osseous spiral lamina.** This structure extends across the cochlea farther in the basal region than it does at the apex.

The bony labyrinth is filled with **perilymph,** which is similar in ionic composition to extracellular fluids elsewhere but has a very low protein content. The membranous labyrinth contains **endolymph,** which is characterized by its low sodium and high potassium content. The protein concentration in endolymph is low.

The Membranous Labyrinth

Saccule and Utricle

The saccule and the utricle are composed of a thin sheath of connective tissue lined with simple squamous epithelium. The membranous labyrinth is bound to the periosteum of the osseous labyrinth by thin strands of connective tissue that also contain blood vessels supplying the epithelia of the membranous labyrinth. In the wall of the saccule and utricle can be observed small regions, called **maculae,** of differentiated neuroepithelial cells that are innervated by branches of the vestibular nerve. The macula of the saccule lies in its floor, whereas the macula of the utricle occupies the lateral wall so the maculae are perpendicular to one another. Maculae in both locations have the same basic histological structure. They consist of a thickening of the wall and possess two types of receptor cells, some supporting cells, and the afferent and efferent nerve endings.

Receptor cells (**hair cells**) are characterized by the presence of 40â€“80 long, rigid stereocilia, which are actually highly specialized microvilli, and one cilium. Stereocilia are arranged in rows of increasing length, with the longest about 100 μm located adjacent to a cilium. The cilium has a basal body and the usual 9 + 2 arrangement of microtubules in its proximal portion, but the two central microtubules soon disappear. This cilium is usually called a kinocilium, but it probably is immotile. There are two types of hair cells, distinguished by the form of their afferent innervation. Type I cells have a large, cup-shaped ending surrounding most of the base of the cell, whereas type II cells have many afferent endings. Both cell types have efferent nerve endings that are probably inhibitory.

The supporting cells disposed between the hair cells are columnar in shape, with microvilli on the apical surface. Covering this neuroepithelium is a thick, gelatinous glycoprotein layer, probably secreted by the supporting cells, with surface deposits of crystals composed mainly of calcium carbonate and called **otoliths,** or **otoconia**.

Semicircular Ducts

Semicircular ducts have the same general form as the corresponding parts of the bony labyrinth. The receptor areas in their ampullae have an elongated ridgelike form and are called **cristae ampullares.** The ridge is perpendicular to the long axis of the duct. Cristae are structurally similar to maculae, but their glycoprotein layer is thicker; this layer has a conical form called a **cupula** and is not covered with otoliths. The cupula extends across the ampullae, establishing contact with its opposite wall.

Endolymphatic Duct and Sac

The endolymphatic duct initially has a simple squamous epithelial lining. As it nears the endolymphatic sac, it gradually changes to tall columnar epithelium composed of two cell types; one of these cell types has microvilli on its apical surface and abundant pinocytotic vesicles and vacuoles. These cells may be responsible for the absorption of endolymph and for the endocytosis of foreign material and cellular remnants that may be present in endolymph.

Cochlear Duct

The cochlear duct, a diverticulum of the saccule, is highly specialized as a sound receptor. It is about 35 mm long and is surrounded by specialized perilymphatic spaces. When observed in histological sections, the cochlea (in the bony labyrinth) appears to be divided into three spaces: the **scala vestibuli**, the **scala media** (cochlear duct) in the middle, and the **scala tympani**. The cochlear duct, which contains endolymph, ends at the apex of the cochlea. The other two scalae contain perilymph and are, in reality, one long tube, beginning at the **oval window** and terminating at the **round window**. They communicate at the apex of the cochlea via an opening known as the **helicotrema.**

The cochlear duct has the following histological structure. The **vestibular** (**Reissner's**) **membrane** consists of two layers of squamous epithelium, one derived from the scala media and the other from the lining of the scala vestibuli. Cells of both layers are joined by means of extensive tight junctions that help preserve the very high ionic gradients across this membrane. The **stria vascularis** is an unusual vascularized epithelium located in the lateral wall of the cochlear duct. It consists of cells that have many deep infoldings of their basal plasma membranes, where numerous mitochondria are located. These characteristics indicate that they are ion- and water-transporting cells, and it is generally believed that they are responsible for the characteristic ionic composition of endolymph.

The structure of the internal ear that contains special auditory receptors is called the **organ of Corti;** it contains hair cells that respond to different sound frequencies. It rests on a thick layer of ground substance the **basilar membrane.** Supporting cells and two types of hair cells can be distinguished. Three to five rows of **outer hair cells** can be seen, depending on the distance from the base of the organ, and there is a single row of **inner hair cells.** The most characteristic feature of these cells is the W-shaped (outer hair cells) or linear (inner hair cells) array of stereocilia . A basal body is found in the cytoplasm adjacent to the tallest stereocilia. In contrast to vestibular receptors, no kinocilium is present. This absence of a kinocilium imparts symmetry to the hair cell that is important in sensory transduction.

The tips of the tallest stereocilia of the outer hair cells are embedded in the **tectorial membrane,** a glycoprotein-rich secretion of certain cells of the spiral limbus.

Of the supporting cells, the **pillar cells** should be singled out for special mention. Pillar cells contain a large number of microtubules that seems to impart stiffness to these cells. They outline a triangular space between the outer and inner hair cellsâ€”the **inner tunnel**. This structure is important in sound transduction.

Both outer and inner hair cells have afferent and efferent nerve endings. Although the inner hair cells have by far the greater afferent innervation, the functional significance of this difference is not understood. The cell bodies of the bipolar afferent neurons of the organ of Corti are located in a bony core in the modiolus and constitute the spiral ganglion.