CHAPTER

03 Anatomy and Physiology of Paediatric Hearing and Vestibular Structures

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'The human body is the most perfect work of art.'

~Leonardo da Vinci

Summary

This chapter provides a comprehensive overview of the audiovestibular system in the paediatric population, delving into its intricate anatomy and functionality. It begins with a detailed discussion of the embryological development of the outer, middle and inner ear, highlighting critical stages of maturation. The chapter then explores the auditory and vestibular nerve pathways, underlining their essential contributions to hearing and balance. Furthermore, it addresses the central processing mechanisms that enable the integration of auditory and vestibular information within the brain. This foundational knowledge is vital for understanding subsequent topics related to assessment, diagnosis and intervention strategies for paediatric populations. By laying this groundwork, the chapter equips hearing healthcare professionals (HHP) to identify and manage hearing and vestibular disorders effectively, ultimately enhancing care and rehabilitation efforts for children.

Learning objectives

From this chapter, the reader will be able to:

1. Understand the embryological development of the ear and the pivotal stages that influence maturation.
2. Obtain a fundamental knowledge of the anatomical structures and nerve pathways that serve as the foundation for auditory and vestibular perception in the paediatric patient.
3. Understand how the audiovestibular system develops during childhood in terms of structure and function, knowledge which is essential for effectively addressing developmental milestones, diagnosing disorders and tailoring interventions.

Introduction

The audiovestibular system is a marvel of biological design, intricately tuned to facilitate the perception of sound and equilibrium from the earliest stages of development. This chapter aims to provide a comprehensive overview of the anatomical and physiological aspects of paediatric hearing and vestibular function, emphasising the complexities of the outer, middle and inner ear. Special attention will be given to the development pathways of the auditory and vestibular nerves. Additionally, the integration of sound processing within the central auditory and vestibular systems will be explored to support the subsequent discussions on assessment methodologies, diagnostic criteria and intervention strategies to support paediatric auditory and vestibular health.

Embryology of the Audiovestibular System

Embryology is a biological discipline dedicated to studying the development of an embryo from the time the ovum is fertilised until it progresses to the foetal stage. A human pregnancy lasts approximately 40 weeks, with the gestation period divided into three trimesters. Each trimester is characterised by distinct developmental milestones and physiological changes.

The first trimester spans from week one to week twelve, the second trimester from weeks thirteen to 26 and the third trimester from week 27 until childbirth. By the conclusion of the first trimester, the foundational structures of the encephalon, or human brain, have begun to take form.

Pharyngeal arches, pouches and clefts are structures that develop in the embryonic stage of human development and are involved in forming various head and neck structures, including the face and ears (Figure 3.1). Inner ear malformations result from developmental arrest at various time points in embryology. Understanding the relationships between anatomical structures is essential for diagnosing developmental disorders.

3.1. (a) **3.1. (b)**

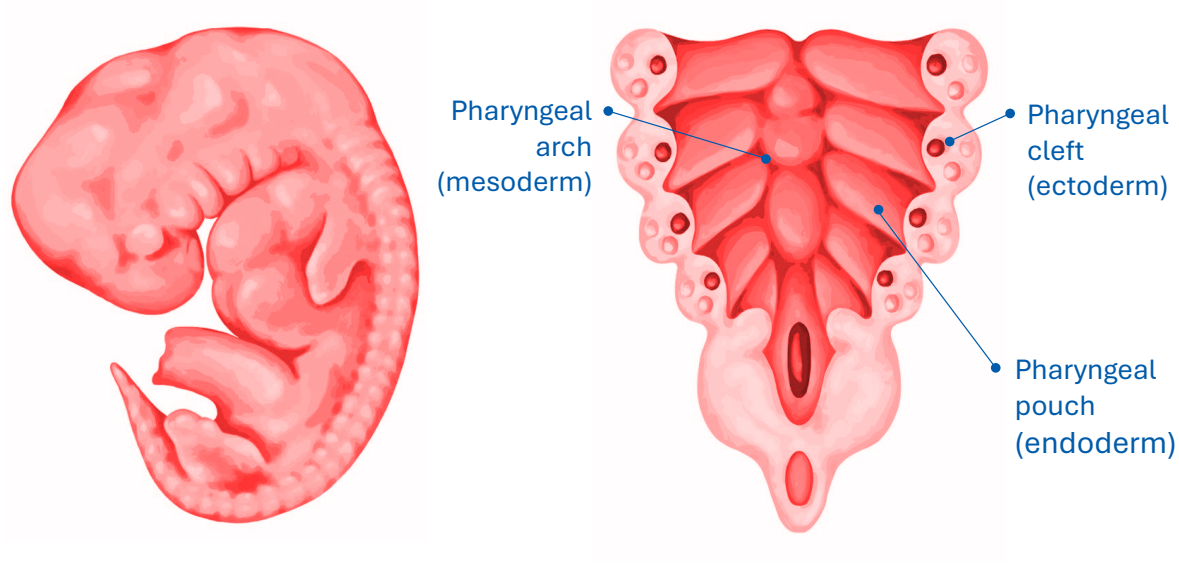


Figure 3.1(a): Five week old embryo, (b): Pharyngeal arches, clefts and pouches

Temporal bone

The temporal bone is a complex structure of the skull that plays a critical role in housing and protecting the auditory and vestibular systems. It comprises several distinct components: the squamous, petrous, tympanic (cartilaginous) and mastoid parts, each of which has unique embryological origins and follows a specific timeline of development during gestation.

Week 4-8: The temporal bone's development occurs. It comprises several distinct parts, each developing from unique embryonic origins.

Week 5: The petrous part, which houses the inner ear, develops. It encases the auditory and vestibular apparatus, and also houses the internal auditory meatus. Ossification begins around week 20.

Week 6: The squamous part, which originates from the prechordal mesoderm, forms the lateral aspect of the temporal bone. It provides support and protection for the underlying structures of the skull and houses compounds of the cranium.

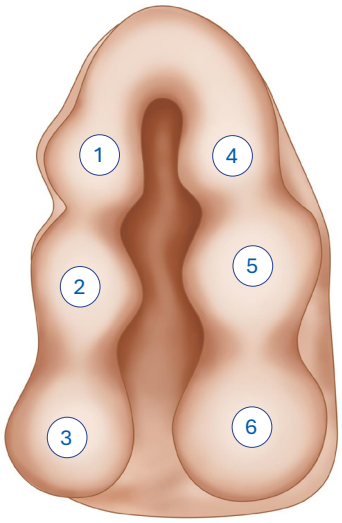
Week 7: The tympanic part develops from a cartilaginous component of the first pharyngeal arch. It helps to form the external auditory meatus (EAM).

Week 24: The mastoid part arises from the temporal region during the latter half of the embryonic period and develops an air cell system that becomes significant postnatally.

Outer ear

The outer ear comprises the pinna (auricle) and the EAM. By the fourth week, the formation of the auricular hillocks begins. The 'Hillocks of His' are six protrusions from the first and second pharyngeal arches (Figure 3.2a). They include the tragus, helix crux, helix, anti-helix, antitragus and lobule and inferior helix of the pinna which are seen by the sixth week. The hillocks gradually fuse to create the rudimentary structure of the pinna, which detaches from the head by the eighteenth week, achieving adult morphology, not size, by 22 weeks (Figure 3.2b).

3.2. (a)



3.2. (b)

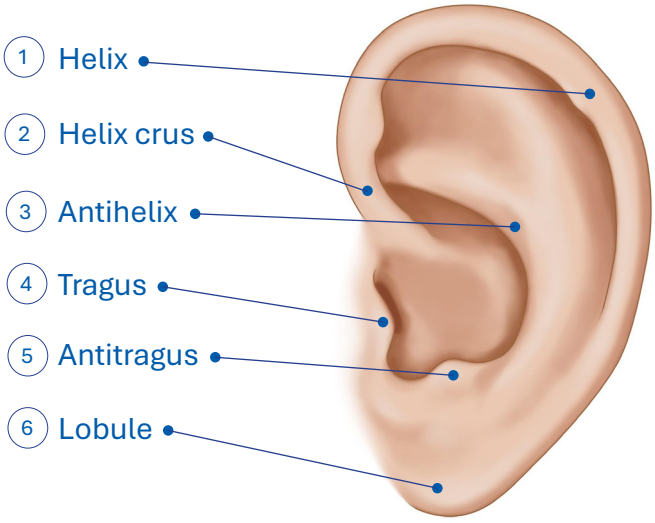


Figure 3.2: (a) The six hillocks of His, (b) The six hillocks of His forms: 1) Helix, 2) helix crus, 3) Antihelix, 4) Tragus 5) antitragus and 6) lobule of the pinna

Microtia, a congenital ear deformity characterised by an underdeveloped external ear, ranges from a small or absent pinna to a partially formed ear structure. Table 3.1 shows how microtia can result from disruptions in development during the sixth to eighth week (Ishinoto et al. 2005). By the sixth week, the EAM develops from the first pharyngeal cleft, which elongates and narrows over time. This canal remains unossified and embedded in mesenchymal tissue during early development. During development, the EAM closes around the eighth week, only to start re-opening in the second trimester. At 21 weeks, it is fully open.

The sebaceous and modified apocrine glands, responsible for the production of cerumen, begin their differentiation at approximately five months gestation, coinciding with the emergence of hair follicles in the outer segment of the external canal.

Table 3.1: Microtia due to disruptions in development, week six to eight

Appearance					
Grade	Normal	Grade 1	Grade 2	Grade 3	Grade 4
Description	Normal size of all parts	External ear is smaller than average. All parts are present. Minor defects.	Outer ear is partially formed and small	Some of the auricular structures are present but no recognised ear parts	Complete absence of the external ear (anotia)
Time of occurrence	Week 21	Week 6-7	Week 6-7	Week 6-8	Before week 6

Middle ear

The ossicles originate from structures derived from the first and second pharyngeal arches. The malleus and incus arise from Meckel's cartilage, which is associated with the first arch. This cartilage is crucial in forming mandibular structures and is a precursor to the malleus and incus. The stapes is derived from Reichert's cartilage, which belongs to the second pharyngeal arch. These structures develop throughout pregnancy, as follows:

Week 4: Formation of the tympanic cavity from the ectoderm of the first pouch.

Weeks 6-8: Formation of the tympanic membrane (TM). Chondrification of Meckel's cartilage, followed by the ossification that gives rise to the malleus and incus.

Week 7: Development of the Eustachian Tube (ET) – completed by Week 16.

Week 16-17: Formation of the Malleus and Incus.

Week 18: Stapes formed.

Week 26: The ossification of the ossicles is complete (Scheuer and Black, 2000).

Inner ear

The inner ear begins forming at the end of the third week of embryonic development, culminating in the development of the cochlear duct's two and a half turns and the three semicircular canals by the end of the eighth week (Jackler, Luxford and House 1987). It is marked by a thickening of the ectoderm at the cephalic end, leading to the formation of the otic placode. This structure is genetically programmed to develop into the membranous labyrinth. As the process continues, the placode invaginates to create the auditory pit, which later becomes encircled by mesoderm, forming the otocyst by the end of the fourth week (Figure 3.3a shows the Otocyst formation).

The otocyst differentiates into different inner ear structures: The ventral part transforms into the saccule and cochlear duct, while the dorsal part develops into the utricle, semicircular canals and endolymphatic duct. In the fifth week, the saccule begins to lengthen and coil, forming the cochlear duct. By the sixth week, the semicircular canals begin as folded

evaginations from the dorsal side of the otocyst, with the superior semicircular canal forming first. By the end of the sixth week, the vestibular system develops into the rest of the semicircular canals, and the cochlear portion continues to elongate. By the end of the eighth week, the cochlear duct has completed two-and-a-half turns.

Figure 3.3: (a)

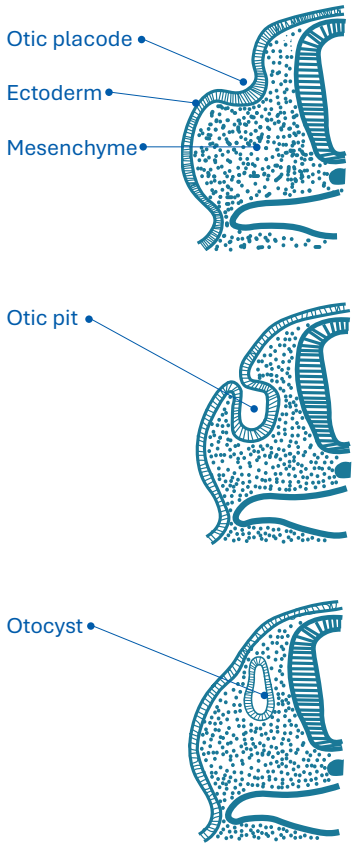


Figure 3.3: (b)

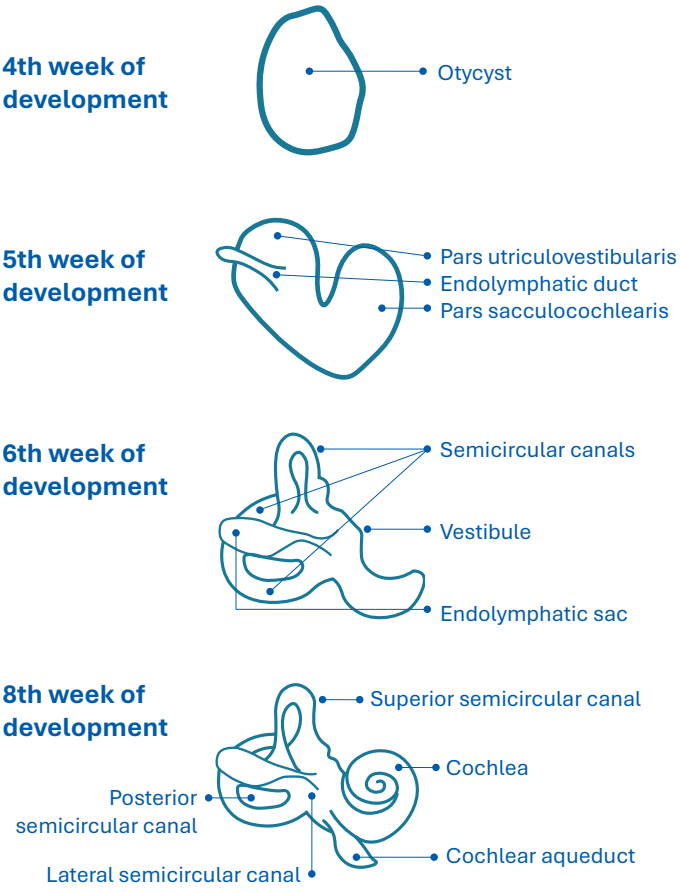


Figure 3.3: (a) Inner ear formation from the 3rd week, developing the otic placode, (b) The Otocyst further differentiates to form two and a half turns of the cochlear duct and three distinct semicircular canals of the vestibular portion by eight weeks of gestation. Source: Dhanasingh (2022)

In the seventh week, the epithelial cells of the cochlear duct differentiate into the Organ of Corti. Initially similar, these cells eventually form inner and outer ridges, with the inner ridge becoming the osseous spiral lamina and the outer ridge developing into inner and outer hair cells. These hair cells interact with the tectorial membrane, while spiral ganglion cells migrate and form a ganglion along the coiled cochlea. By the tenth week, surrounding mesenchyme transforms into cartilage, which vacuoles to create the cochlea's scala vestibuli and scala tympani. The inner ear reaches near-adult size by the end of the fifth month, achieving total cochlear growth within six months.

The vestibular and cochlear aqueducts arise from the otic placode during the third week of development. The cochlear aqueduct, a narrow channel connecting the cochlea to the subarachnoid space of cranial cavity, forms from the posterior aspect of the developing cochlea, typically emerging around the sixth week of development. The vestibular aqueduct arises from a distinct region of the otic vesicle, connecting the vestibule to the posterior cranial fossa. By the eighth week, this structure is well-formed. Although most of the membranous labyrinth is adult-sized by twenty weeks of gestation, the vestibular aqueduct continues to grow throughout embryonic life (Pyle 2000).

Malformations in the inner ear can vary significantly, depending on the timing of developmental disruptions (Table 3.2). By studying the embryological development of the inner ear, HHPs can identify at which stage differentiation has been interrupted, thus aiding in treatment decisions regarding cochlear implants and other interventions (Dhanasingh et al. 2021) (Figure 3.4).

Vestibulocochlear nerve

The spiral ganglion, which develops from the neuroblasts originating from the otic vesicle, forms the auditory aspect connected to the cochlea. Ganglion cells from the auditory nerve emerge from the otocyst in the fourth week, and their fibres enter the cochlea by the fifth week, spreading parallel to its curvature. Simultaneously, the cristae ampullares in the semicircular canals develop from the neurogenic epithelium of the ampullae (Anniko and Nordemar 1982). By the end of the eighth week, the vestibulocochlear nerve is structurally established and ready to relay sensory information to the brain. Connections to inner hair cells form between the 11th and 12th weeks, while outer hair cells make contact at the apex by the 14th week. Synapses are fully formed by the seventh month, with maturation occurring in the last trimester, allowing the foetus to perceive sound outside the womb by the 28th week (Querleu et al. 1988).

The internal auditory meatus develops through inhibited cartilage formation at the otocyst's medial aspect, a process reliant on the presence of the vestibulocochlear nerve (Amarnath et al. 2016). Cochlear nerve deficiency is the most common cause of single-sided deafness in neonates (Yoshimura et al. 2024).

Brainstem nuclei and tracts of audiovestibular function

The brainstem nuclei and tracts of audiovestibular function refer to the specialised clusters of nerve cells and associated pathways within the brainstem that are responsible for the processing of auditory and vestibular information. Stages of development include:

Week 4: The oculomotor nuclei originate from the mesencephalon and are crucial for controlling eye movements. These nuclei mature as sensory input from the developing inner ear becomes functional. By the end of the week, the hindbrain differentiates into the metencephalon and myelencephalon. It is essential for developing pathways for the vestibulo-ocular, vestibulo-colic and vestibulo-spinal reflexes necessary for maintaining balance and co-ordination.

Week 5: The brainstem's vestibular and cochlear nuclei develop from the rhombencephalon, specifically the caudal hindbrain. The oculomotor nuclei are positioned and increasingly organised, reflecting their specialisation for motor functions. Concurrently, the medial longitudinal fasciculus (MLF), a crucial neural pathway for co-ordinating eye and head movements, begins to develop (Ahsan, Riley and Schubert 2007).

Week 6: The integrity of cranial nerve pathways begins to establish, allowing the vestibulo-ocular reflex (VOR) to take form.

Week 8: The cochlear nuclei, trapezoid body, superior olive, lateral lemniscus and inferior colliculus can be identified, establishing foundational connections for sound processing and vestibular function. The connections between the vestibular system and the spinal cord give rise to the vestibulo-colic and vestibulo-spinal reflexes. These structures mature further, allowing for the refinement of auditory and vestibular pathway function.

Vestibulocerebellum

The vestibulocerebellum is an integral part of the cerebellum. It plays a vital role in maintaining balance and co-ordinating eye movements. Its intricate connections with associated

brainstem nuclei and pathways facilitate vestibular processing and motor co-ordination. The development occurs as follows:

Week 4: The neural tube is fully formed, and will eventually differentiate into the central nervous system. As development progresses, the metencephalon begins to give rise to the cerebellum.

Week 6: Precursors of the cerebellar structures are established, including the vermis and the flocculonodular lobe, which will contribute to the vestibulocerebellum (Amore et al. 2021).

Thalamus, auditory and vestibular cortex

The thalamus, auditory cortex and vestibular cortex are integral components of the brain's sensory processing and motor co-ordination systems. The thalamus serves as a crucial relay station, processing and transmitting sensory information. The auditory cortex is primarily responsible for the perception and interpretation of sound. The vestibular cortex, associated with the vestibular system, is involved in maintaining balance and spatial orientation. Stages in development include:

Week 4: The auditory cortex begins developing when the formation of the inner ear structures primarily drives the auditory system.

Week 5: The thalamus begins its development (Antón-Bolaños, Espinosa and López-Bendito 2018).

Week 6: The otic placode develops into the otic cyst.

Week 10: The thalamus is organised into its future nuclei (Sheridan and Tadi 2024).

Week 10-12: The auditory cortex in the superior temporal gyrus appears as connections are made to the thalamus's medial geniculate nucleus (MGN). The MGN is responsible for processing auditory information, particularly in distinguishing pitch and frequency. Neurons in the MGN project target specific regions of the auditory cortex, allowing for precise localisation of sound sources and decoding of complex auditory stimuli, such as spoken language or music.

Week 16: The central vestibular cortex is formed. The central vestibular cortex, located in the parieto-occipital area, receives input from the thalamus's ventral posterior nucleus (VPN). This development lays the groundwork for spatial orientation and balance by establishing links with visual and proprioceptive systems, stabilising vision during motion and enabling posture and movement co-ordination.

Table 3.2: Inner ear malformations

Condition	Description	Time of development
Michel's deformity	Complete labyrinthine aplasia, absent internal auditory canal arrest of the otic placode.	*3rd week*
Cochlear aplasia (CA)	Absence of the cochlear portion while the vestibular portion may be partially formed.	*Late 3rd week*
Common cavity (CC) malformation	Cystic structure lacking distinct cochlear and vestibular differentiation.	*4th week*
Incomplete Partition Type I (IP type I)	Separable cochlear and vestibular structure but lacking the modiolus.	*5th week*
Incomplete Partition Type II (IP type II)	Cochlea displays cystic characteristics, often accompanied by a dilated vestibular aqueduct, potentially risking fluid leakage during electrode insertion.	*7th week*
Incomplete Partition Type III (IP type III)	*Cochlear structure absent from the central modiolus.*	*Genetically inherited disorder linked to the X chromosome*
Enlarged vestibular aqueduct (EVA)	*Abnormal dilation of the vestibular aqueduct.*	*6th to 12th week*
Cochlear hypoplasia	*Arrest in differentiation of semicircular canals of ves-tibular portion and the elongation of cochlear duct.*	*6th week*

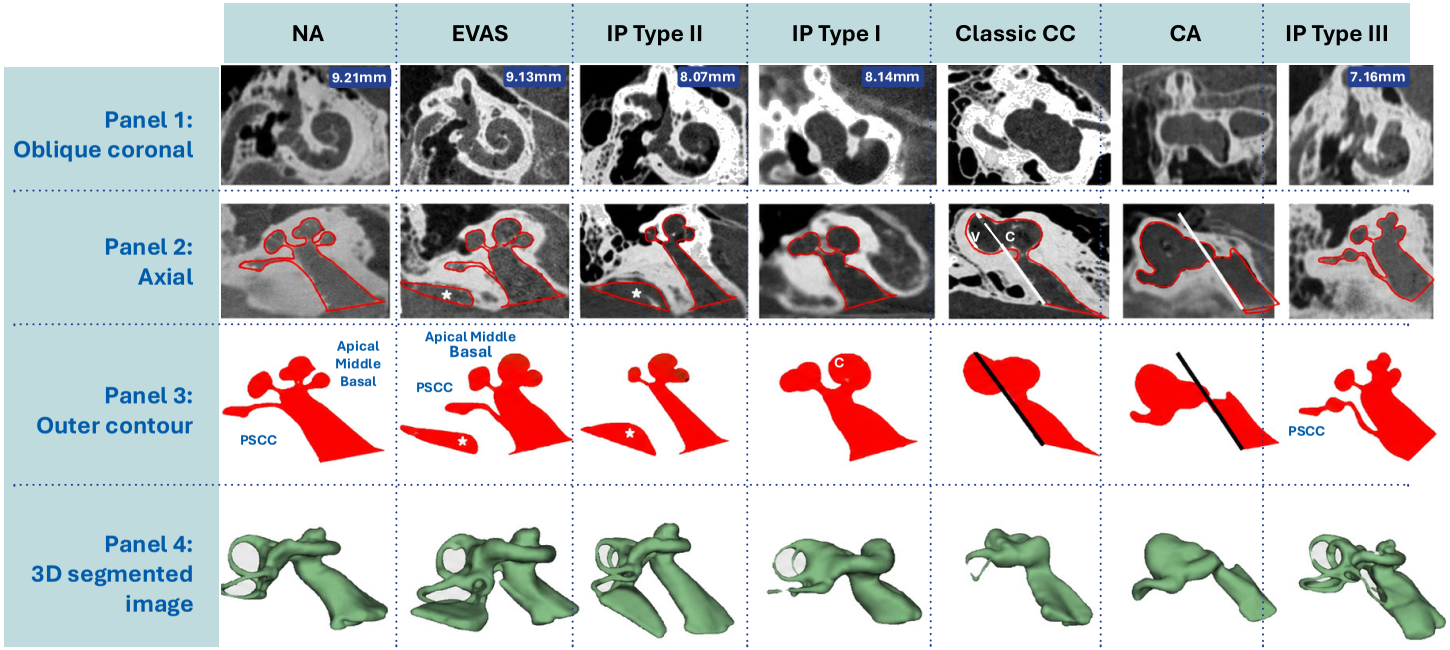


Figure 3.4: Different inner ear malformations. Computed tomography (CT) oblique coronal view (panel 1) and axial view (panel 2). The captured outer contour filled with red (panel 3). The straight line drawn parallel to the posterior edge of the internal auditory canal differentiates the cochlear (C) from the vestibular portion (V). The enlarged vestibular aqueduct (*) seen in enlarged vestibular aqueduct syndrome (EVAS) and the incomplete partition (IP) type II. The 3D segmented images (panel 4). Adapted from: Dhanasingh et al. (2021).

Treasures of Wisdom

Gestational development

- It is important to recognise that the processes involved in gestational development can result in malformations that potentially lead to significant hearing and balance impairments.
- The type of inner ear malformation significantly influences intervention, especially the selection of cochlear implants.

Anatomy and Physiology of the Paediatric Audiovestibular System

Temporal bone

The temporal bone is a paired structure located on the lateral sides of the cranium. It plays a vital role in protecting the delicate structures of the inner ear and houses the auditory and vestibular systems, which are essential for hearing and balance. At birth, the temporal bone is anatomically distinct from its adult counterpart. The newborn's temporal bone comprises smaller, softer, malleable bone structures, accounting for the rapid adaptability needed for the delivery process.

Figure 3.5: (a)

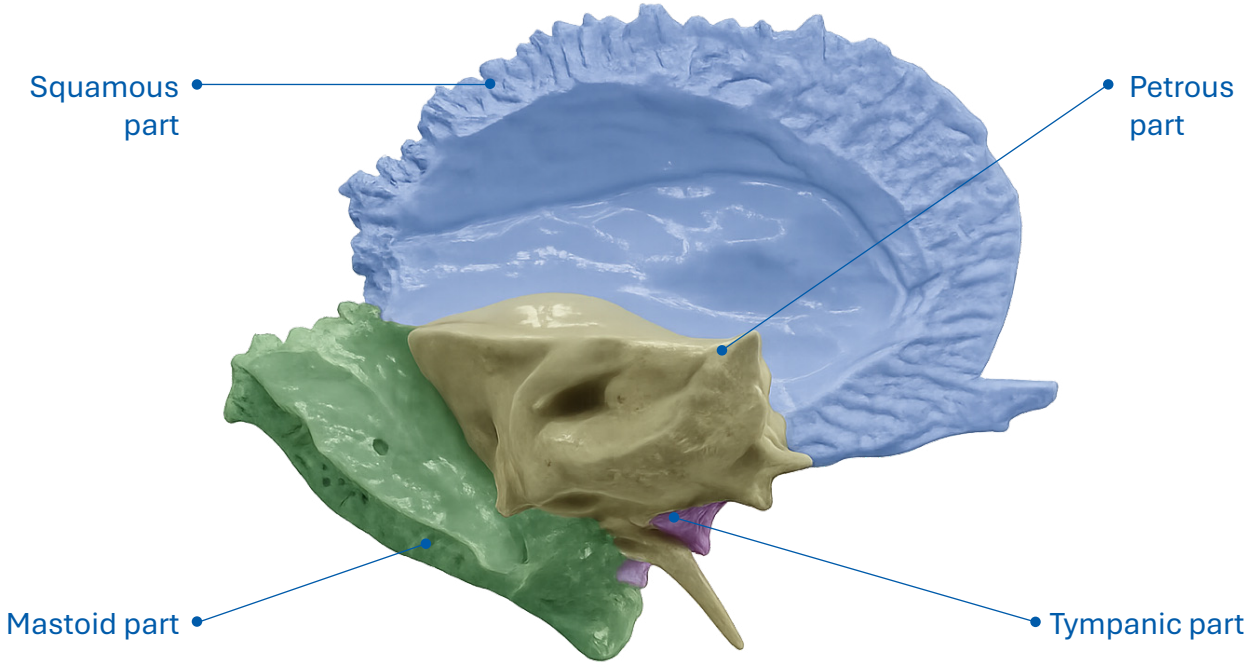


Figure 3.5: (b)

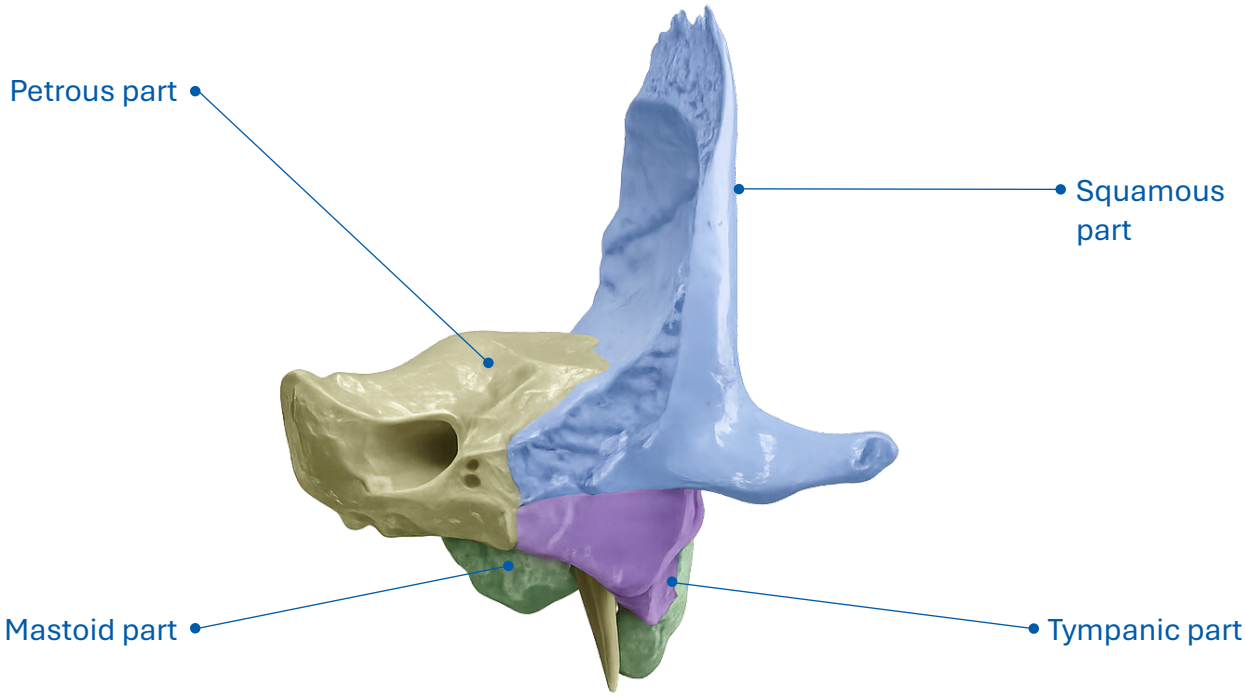


Figure 3.5: (a) The parts of the temporal bone, a coronal view; (b) The parts of the temporal bone, a sagittal view/plan

Each temporal bone is anatomically divided into four parts: the squamous, mastoid, petrous and tympanic parts (Figure 3.5). The squamous portion is flat and contributes to the side of the skull, while the mastoid portion houses the mastoid process, which serves as an attachment point for neck muscles. The mastoid process is not fully developed at birth. The petrous part contains the intricate structures of the inner ear, including the cochlea and semicircular canals. The tympanic part forms the external acoustic meatus, allowing sound waves to travel to the eardrum.

The temporal bone also plays a significant role in various functions, ranging from jaw movement to cranial stability. Its association with the temporomandibular joint facilitates the complex mechanics of chewing and speaking. Additionally, the temporal bone's proximity to

vital cerebral structures underlines its importance, making it essential for anatomical integrity and physiological function.

Outer ear

The pinna is the visible part of the outer ear that protrudes from the side of the head. It consists of skin-covered cartilage. In newborns, the pinna is relatively large in proportion to the head, compared to adults. While it is still developing, the overall structure is present at birth. The pinna has a distinctive shape resembling a funnel that helps capture sound waves from the outside world and directs them into the ear canal. This aids in sound localisation, allowing newborns to determine the direction from which sounds are coming. The pinna also assists in amplifying specific frequencies of sound, which helps with auditory localisation. The pinna acts as a natural barrier that helps protect the inner structures of the ear from dirt and foreign objects. Its shape and orientation help minimise the risk of injury or infection.

The external auditory meatus connects the outside world from the pinna to the middle ear space. Anteriorly, it forms the posterior border of the temporomandibular joint, which means movements during feeding or sucking can impact the canal's function. Medially, the canal leads to the TM, and posteriorly, it is bordered by the structures of the mastoid process. The proximity of these structures can influence the spread of infections and the mechanics of sound transmission.

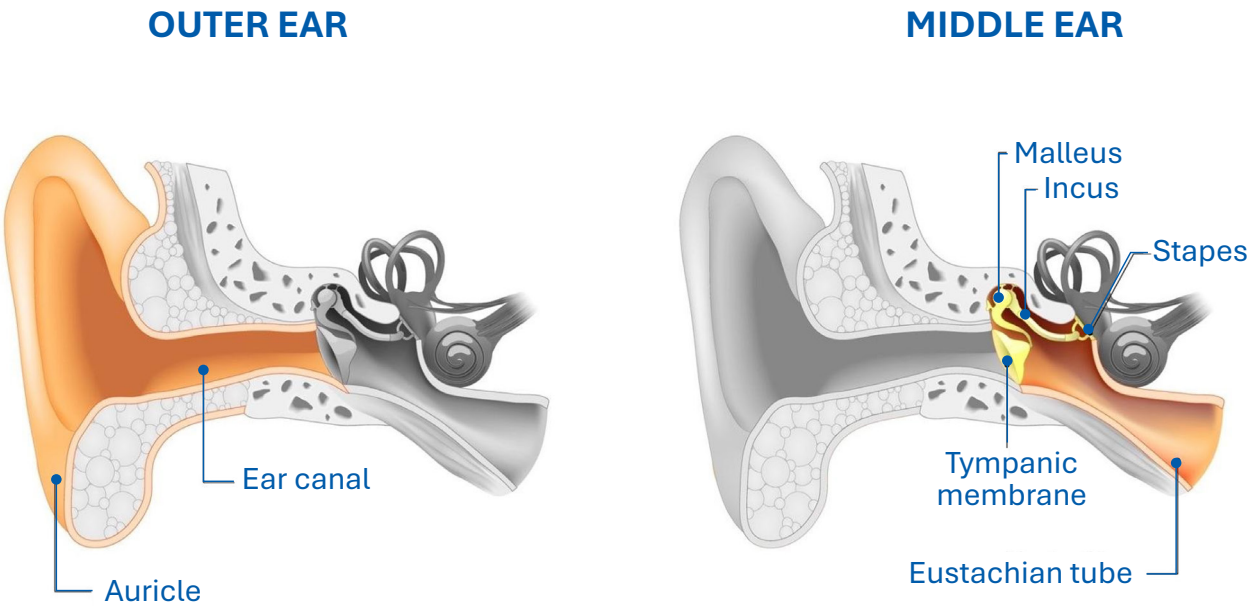
In adults, the ear canal measures approximately two and a half to three centimetres and features a slightly S-shaped curvature. In contrast, the external auditory meatus of newborns is notably shorter, measuring one to one and a half centimetres. Its shape is more linear and horizontally oriented, demonstrating less curvature compared to adults. This anatomical immaturity reflects the ongoing development of the auditory system during infancy. The cartilaginous portion of the ear canal is softer and more flexible in infants, potentially increasing vulnerability to ear infections. This developmental disparity necessitates unique considerations in clinical assessments and interventions for paediatric patients.

The external auditory meatus is covered with skin-lined epithelium characterised by keratinised and non-keratinised squamous cells. This layered structure aids in the consistent turnover and protection of the ear canal. Ceruminous glands in the outer portion of the meatus are modified apocrine glands that, together with sebaceous glands, are responsible

for the production of cerumen, commonly known as earwax. Cerumen is a natural defence mechanism, trapping dust, foreign particles and microorganisms, preventing them from reaching the TM. The pH of the meatus is slightly acidic, ranging from 4.5 to 5.5. This acidity plays a crucial role in maintaining a healthy environment by helping to inhibit the growth of harmful bacteria and fungi. The natural secretions of the ear, including cerumen, have anti-microbial properties and contribute to the acidic environment, acting as a protective barrier against infections.

The external auditory meatus has a natural self-cleansing mechanism that helps to keep it free from excess wax, debris, and bacteria. This self-cleaning process involves the production of cerumen, which gradually moves from the inner part to the outer part where it can eventually be washed away or fall out. The movement of the jaw while talking or chewing also aids this process, assisting in the natural expulsion of cerumen.

The resonance frequency of the external ear canal is within the 2 kHz to 3 kHz range, coincidentally aligning with human speech's most essential components. This resonance enhances auditory perception, allowing neonates to perceive and differentiate sounds critical for early language development and social interaction. The small size of the canal, coupled with its specific geometry, accentuates certain frequencies, fostering an optimal auditory environment for the infant.



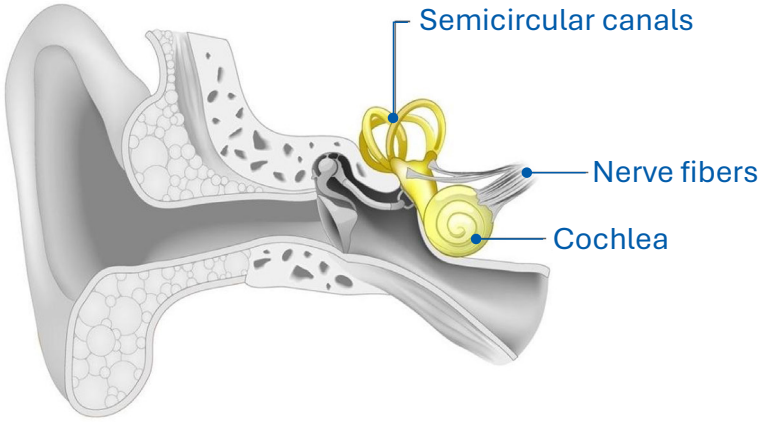


Figure 3.6: The outer, middle and inner ear from the front

Middle ear

The middle ear consists of the middle ear cavity, TM, ossicles, ET opening and the mastoid connection (Figure 3.6). The middle ear cavity is an air-filled space in the centre of the temporal bone. It is situated between the external auditory meatus and the inner ear, functioning as a channel through which sound is transmitted, and air pressure is regulated. The middle ear is well-vascularised and lined by mucosa with secretory properties. Although the ET performs this function primarily, the mucosa also helps with gas exchange in the middle ear. Its blood supply primarily originates from branches of the maxillary and stylomastoid arteries. Innervation is provided by branches of the facial and glossopharyngeal nerves, with sympathetic fibres also present. The TM is the barrier between the external auditory meatus and the middle ear (Figure 3.7). It converts sound waves into mechanical vibrations. In newborns, the size of the TM is smaller than that of older children and adults.

Treasures of Wisdom

Outer ear

- The external auditory meatus exhibits significant variations in size and shape between adults and newborns, reflecting their distinct anatomical and functional characteristics.
- Introducing soap and shampoo into the meatus can disrupt the natural pH balance. Many commercial products are alkaline, raising the pH and compromising the ear's natural defences.
- Using cotton swabs or Q-tips to clean the meatus can disrupt its natural mechanism and push earwax deeper into the canal, causing impaction, discomfort and infections. It can also damage the delicate skin, leading to irritation or lacerations, increasing the risk of infections.

A smaller and thinner TM in newborns influences their auditory capabilities, particularly regarding sound audibility and frequency range. As infants grow, the TM gradually increases in size, paralleling the growth of the surrounding anatomical structures. This maturation process is essential for the enhancement of auditory perception and the ability to process complex sounds.

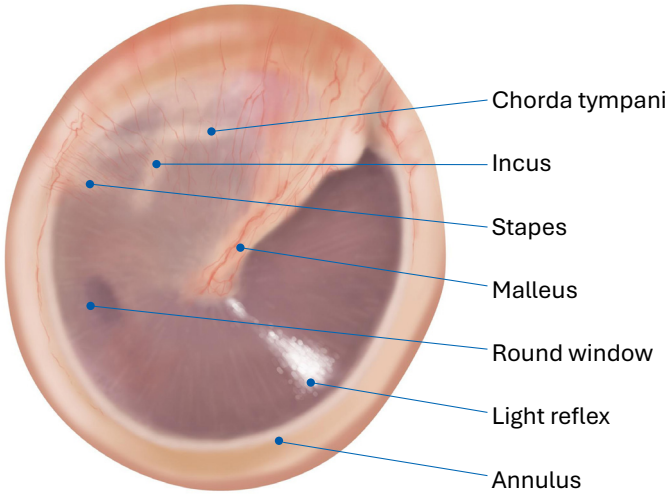


Figure 3.7: Tympanic Membrane

The TM comprises three layers: an outer epidermal layer, a middle connective tissue layer and an inner mucosal layer. In the outer layer, epithelial cells produce keratin, which is shed over time. The keratin migrates outwards to help form the cerumen. Its outer layer provides a protective barrier. The connective tissue in the fibrous middle layer is primarily made of collagen and elastin. It is arranged in an outer radial and inner circular fashion, contributing to the TM‘s tensile strength and vibration capability. The inner mucosal layer protects the interior of the middle ear and connects to the ossicles.

The TM comprises three distinct regions: the annulus, pars tensa and pars flaccida (Figure 3.7). The annulus is a fibrocartilaginous ring that anchors the TM within the external acoustic meatus. It maintains the membrane’s position and ensures it can vibrate effectively in response to sound waves. The pars tensa is the most significant section of the TM, characterised by its tight, fibrous composition. It transforms sound energy into mechanical vibrations. When sound waves reach the membrane, they cause it to vibrate, transferring these vibrations to the middle ear’s ossicles. This mechanical energy is then relayed to the inner ear for

further processing. The robustness of the pars tensa also allows it to withstand atmospheric pressure changes and the mechanical forces exerted by sound waves.

The pars flaccida, or Schrapnell's membrane, is located superiorly and lacks the same tension and structural support found in the pars tensa. It comprises fewer fibres and plays a lesser role in sound transmission, primarily serving as a compliant area that accommodates pressure variations within the ear. Branches from the auriculotemporal and vagus nerves innervate the TM and facilitate sensations like pain. In newborns, the nerve supply is often less developed, increasing vulnerability to infections and affecting hearing. The main blood supply to the TM comes from the deep auricular artery and tympanic branches of the maxillary artery.

The ossicles are three tiny bones in the middle ear: the malleus, incus and stapes (Figure 3.8). They facilitate the transmission of sound vibrations from the external environment to the inner ear. The malleus, commonly known as the hammer, is attached to the TM and receives sound waves, transforming them into mechanical vibrations. It then passes these vibrations to the incus, or anvil, which bridges the malleus and the stapes, or stirrup. The stapes, the smallest bone in the human body, connects with the cochlea's oval window and amplifies the sound vibrations before they enter the fluid-filled inner ear.

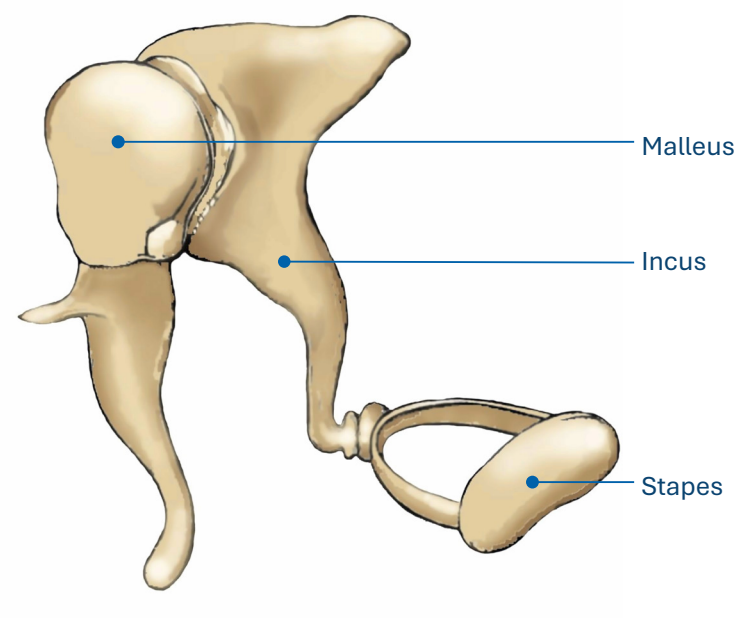


Figure 3.8: The ossicular chain

Amplification occurs in the middle ear through two fundamental mechanisms: the lever action of the ossicles and the differences in surface area between the TM and the oval window. The malleus and incus act as a lever system, magnifying the force exerted on the stapes. Additionally, since the TM is significantly larger than the oval window, the pressure exerted on the oval window is greater than that on the TM, resulting in an effective amplification of sound.

Synovial joints connect the ossicles. They are capable of limited motion, facilitating nuanced changes in sound vibrations. This mobility also enables them to perform a protective function, helping safeguard the TM from pressure fluctuations and maintaining proper ear function during activities such as swallowing or yawning.

The ossicles are further supported by ligaments and connected to two key muscles: the stapedius and the tensor tympani. The stapedius muscle is pivotal in the acoustic reflex, contracting in response to loud sounds, which helps protect the inner ear from potential damage by dampening the vibrations of the stapes. Similarly, the tensor tympani muscle responds to sound and pressure changes, stabilising the TM and reducing the transmission of disruptive vibrations from the throat during swallowing. The tensor tympani muscle is innervated by the trigeminal nerve (CN V), and the stapedius muscle is supplied by the facial nerve (CN VII).

The ET opening lies at the anterior aspect of the middle ear. This opening connects the middle ear to the nasopharynx and facilitates pressure equalisation. It comprises three main sections: the bony, cartilaginous and membranous segments. The bony segment measures about ten millimetres and is found near the middle ear, while the cartilaginous section is the longest segment, connecting to the nasopharynx. The membranous portion is shorter and contains a layer of epithelium, allowing it to maintain patency. The ET is lined with respiratory epithelium, composed primarily of ciliated columnar and goblet cells that produce mucus. This mucus traps particulates and pathogens, while the cilia facilitate the movement of mucus toward the nasopharynx, aiding in the clearance of potential infections. Beneath the epithelium lies a connective tissue layer rich in blood vessels and lymphatics, which plays a role in the immune response.

Figure 3.9: (a)

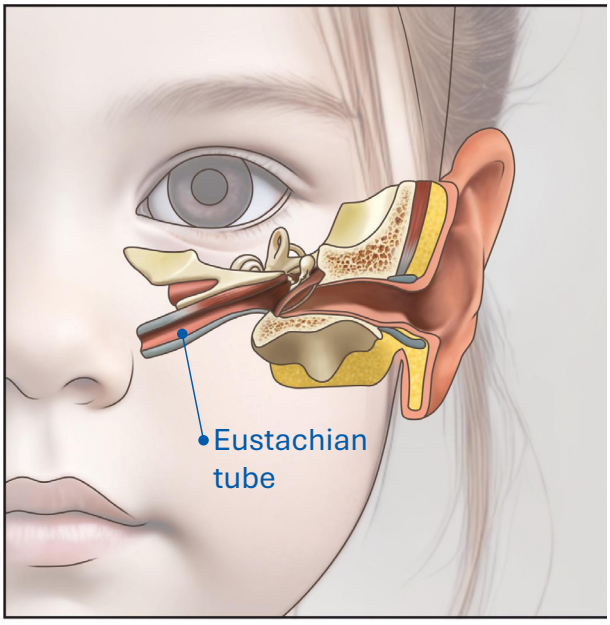


Figure 3.9: (b)

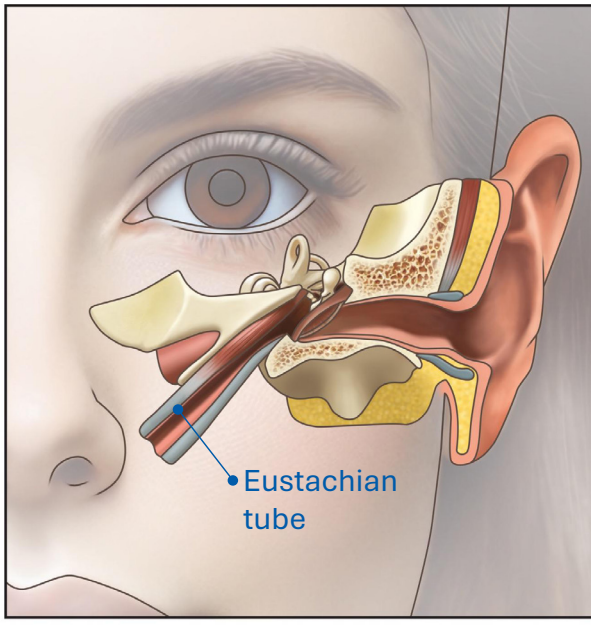


Figure 3.9: (a) The ET in and infant, (b) The ET in an adult

Table 3.3: Differences between the infant and adult ET

	Infants	Adults
Size	Smaller	*Larger*
Length	Shorter	*Longer*
Stiffness	Softer	*Stiffer*
Direction into nasopharynx	Horizontal	*More vertical*
Orifice in nasopharynx	Round	*More oval*

Effusion in the middle ear is an exudation formed under negative pressure exceeding 100 millimetres H2.O. This air vacuum is induced when nitrogen, the gas with the most significant diffusion gradient, is absorbed by microvascular circulation quicker than it can be replaced due to ET obstructive dysfunction.

The posterior aspect of the middle ear leads via the aditus ad antrum into the mastoid air cells, the largest of which is the mastoid antrum. The roof of the middle ear forms the boundary with the middle cranial fossa and temporal lobe. The floor of the middle ear is adjacent to the infratemporal fossa, housing the carotid artery and the jugular bulb of the jugular vein.

Mastoid process

The mastoid process is a prominent bony structure behind the ear, forming part of the skull's temporal bone. It is underdeveloped in newborns compared to its formation in adults, as the ossification process remains incomplete at this stage. The mastoid air cells are integral to the mastoid process and vital to maintaining ear health and functionality.

Mastoid air cells comprise a complex network of small, interconnected cavities encapsulated by a mucous membrane. The lining consists of predominantly ciliated columnar cells interspersed with goblet cells responsible for mucus secretion. This specialised arrangement ensures a healthy respiratory epithelium, as it filters and humidifies incoming air. The cilia facilitate mucus movement and capture particles outward, thereby supporting the hygiene of the middle ear and surrounding epitympanic space. Additionally, these air cells harbour dense vascular and neural tissue, contributing to both immune responses and sensory functions.

These air cells are a pneumatic system that helps balance pressure between the middle ear and the external



Treasures of Wisdom

Middle ear

- The TM is situated more horizontally in newborns than in adults, which may contribute to differences in sound localisation and perception.
- The ET is a slender, flexible tube measuring approximately 31 to 44 mm in adults and eighteen millimetres in infants. In adults, the tube forms an angle of 30-40 degrees with a horizontal plane, whereas in children it only forms an angle of ten degrees.
- The shorter and more horizontal ET in infants significantly impacts ventilation and drainage properties. This anatomical characteristic increases the risk of otitis media.
- The length and angle of the ET change significantly as the mandible and skull base grow, typically reaching adult dimensions by the time a child is around seven years old.

environment. Pneumatisation is when epithelial cells infiltrate the developing bone and create epithelium-lined air cell cavities. This is particularly significant during transitioning from a fluid-filled foetal environment to an air-filled atmosphere after birth, which can introduce pressure imbalances affecting hearing and balance.

The mastoid air cells connect to the middle ear via the aditus ad antrum, thus aiding in the necessary pressure equalisation to ensure proper TM function.

Inner ear

The inner ear or labyrinth is a complex maze consisting of irregular pathways and passages in the inner ear. It contains the hearing and balance organs (Figure 3.10).

The bony labyrinth, also known as the osseous labyrinth or otic capsule, is the rigid, bony outer wall of the inner ear embedded within the temporal bone. The otic capsule reaches its adult size at birth, measuring approximately fifteen millimetres in width. The bony labyrinth is divided into three primary parts: the cochlea, the vestibule and the semicircular canals. The bony labyrinth houses and protects the membranous labyrinth.

Treasures of Wisdom

Mastoid process

- Due to frequent ET dysfunction in newborns, the interconnected nature of mastoid air cells provides an alternative drainage pathway for fluid accumulation in the middle ear, potentially mitigating the risk of infections, such as otitis media.

Figure 3.10: (a)The bony and (b) membranous labyrinth

Figure 3.10 (a)

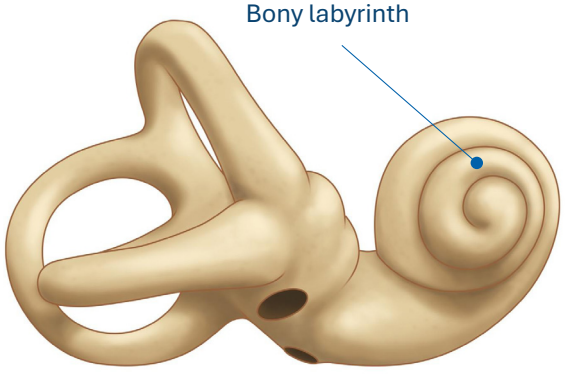
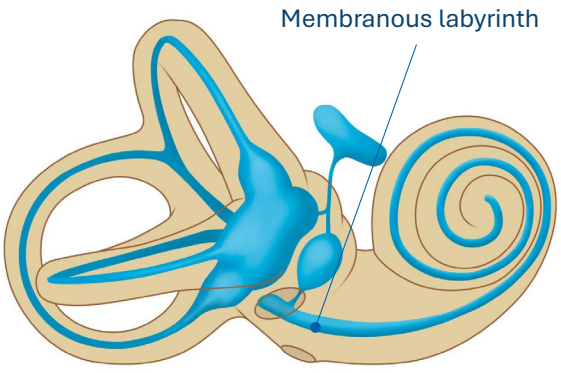


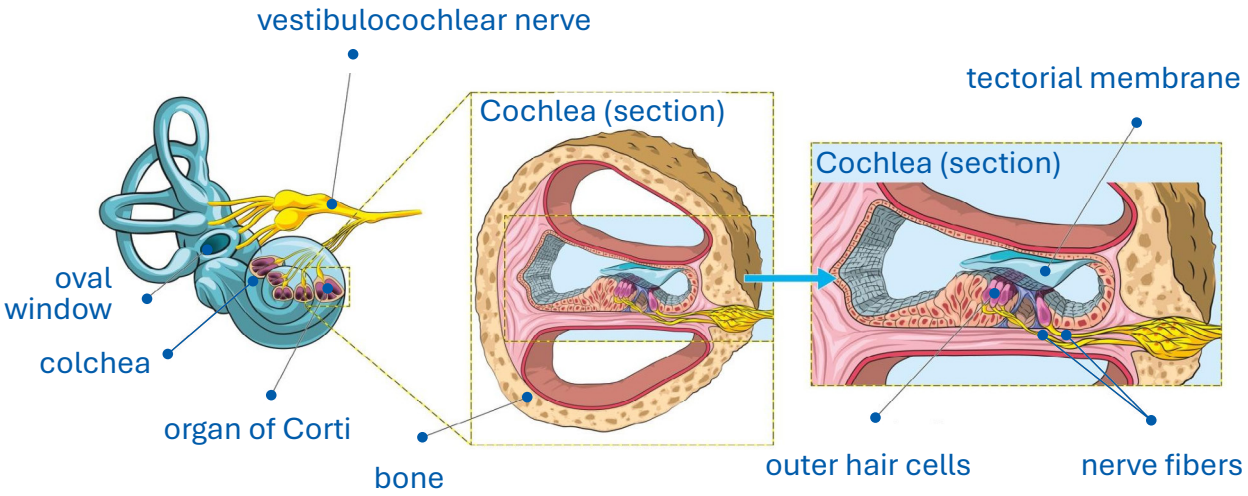
Figure 3.10 (b)



The membranous labyrinth is a complex system of fluid-filled channels and sacs within the bony labyrinth. It is divided into the cochlear duct, saccule, utricle, three semicircular ducts and the endolymphatic duct and sac.

In adults, the cochlea is a 3.2 to 4.2 centimetre-long spiralled, snail-shaped structure composed of two and a half turns (Figure 3.11). It converts sound vibrations into neural signals. The cochlea comprises three fluid-filled chambers: the scala vestibuli, scala tympani and scala media (cochlear duct). The scala vestibuli and tympani are filled with perilymph, similar to extracellular fluid with a high sodium content. In contrast, the scala media contains endolymph, a unique ionic composition with a high potassium content that supports sensory transduction.

Figure 3.11: The cochlea, scala and organ of Corti



The cochlear duct is separated from the scala vestibuli by Reissner's membrane and from the scala tympani by the basilar membrane (Figure 3.11). The latter supports the organ of Corti, the sensory structure that contains hair cells responsible for transducing sound. Hair cells enable the transduction of mechanical stimuli into electrical signals.

These hair cells are arranged in two rows: The inner hair cells, which mainly convey auditory information to the brain and the outer hair cells, which amplify sound vibrations. The human cochlea has 3500 inner hair cells and about 12,000 outer hair cells. The movement of the basilar membrane in response to sound waves causes the stereocilia on these hair cells to bend, opening ion channels that depolarise and, subsequently, release neurotransmitters.

The cochlea also contains supporting cells and the cells of the stria vascularis at the lateral aspect near the wall, which maintains the ionic composition of the endolymph. Connexins are a family of proteins that form gap junctions between adjacent cells, allowing for direct communication and transport of ions, small molecules and electrical signals.

The cochlear aqueduct is a small channel that extends from the scala tympani of the basal turn of the cochlea to the subarachnoid space. Measuring approximately 0.7 to 1.5 mm in diameter, it plays a role in the maintenance of inner ear fluid and balances the pressure between the perilymph space and the cerebrospinal fluid. It is a potential pathway for the spread of infections, such as meningitis, into the cochlea.

The vestibulum forms the central part of the bony inner ear and is essential for balance and spatial orientation. It encompasses the two otolith organs, the utricle and saccule, which detect linear acceleration, of which gravitation is a constant component. The vestibulum is anatomically located between the semicircular canals and the cochlea.

The utricle and saccule contain maculae, specialised sensory patches of hair and supporting cells. The hair cells in these maculae are embedded in a gelatinous layer laden with calcium carbonate crystals called otoliths. When the head moves, gravitational or inertial forces during acceleration cause the otoliths to shift, leading to a relative displacement of the gelatinous layer. This mechanical distortion causes the bending of stereocilia on the hair cells, releasing neurotransmitters and sending signals to the vestibular nuclei in the brainstem.

The maculae of the utricle and saccule exhibit a highly organised structure. The hair cells' stereocilia are arranged in a specific pattern with taller kinocilia positioned in a defined orientation, enabling the detection of directional movements. The utricle contains approximately 33 000 hair cells and the saccule 19 000 hair cells.

The supporting cells provide structural and functional support. At the same time, the overlying otolith membrane is rich in glycoproteins that contribute to the mass difference necessary for the displacement caused by head movements.

There are three bony semicircular canals: the superior (anterior), posterior (inferior) and lateral (horizontal). Each canal contains a membranous duct and is oriented perpendicularly to the others, thus facilitating the detection of angular accelerations during various head movements. At the base of each canal lies the ampulla, which contains the crista ampullaris, where sensory hair cells reside. The three semicircular canalseach contain 7600 hair cells.. The semicircular ducts are filled with endolymph and surrounded by perilymph.

When the head rotates, the inertia of the endolymph causes it to lag behind the motion of the semicircular canals. This relative movement of the endolymph results in bending of the cupula, a gelatinous structure that overlies the crista ampullaris. Such bending stimulates the hair cells within the crista, leading to their depolarisation and the subsequent generation of action potentials. These electrical signals are conveyed to the central nervous system via the vestibular nerve, thus enabling an organism's perception of its position and movement in three-dimensional space.

The crista ampullaris, which consists of hair cells and supporting cells, is similar to the maculae. However, its unique orientation is specifically adapted for detecting rotational movements. The directional arrangement of the kinocilia in the hair cells within the semicircular canals allows for precise motion discrimination.

The vestibulo-ocular reflex (VOR) is an essential mechanism that stabilises vision during head movements, allowing the eyes to maintain a steady gaze on a target (Figure 3.12). It can be categorised into two primary types: angular (rotational) VOR and translational VOR.

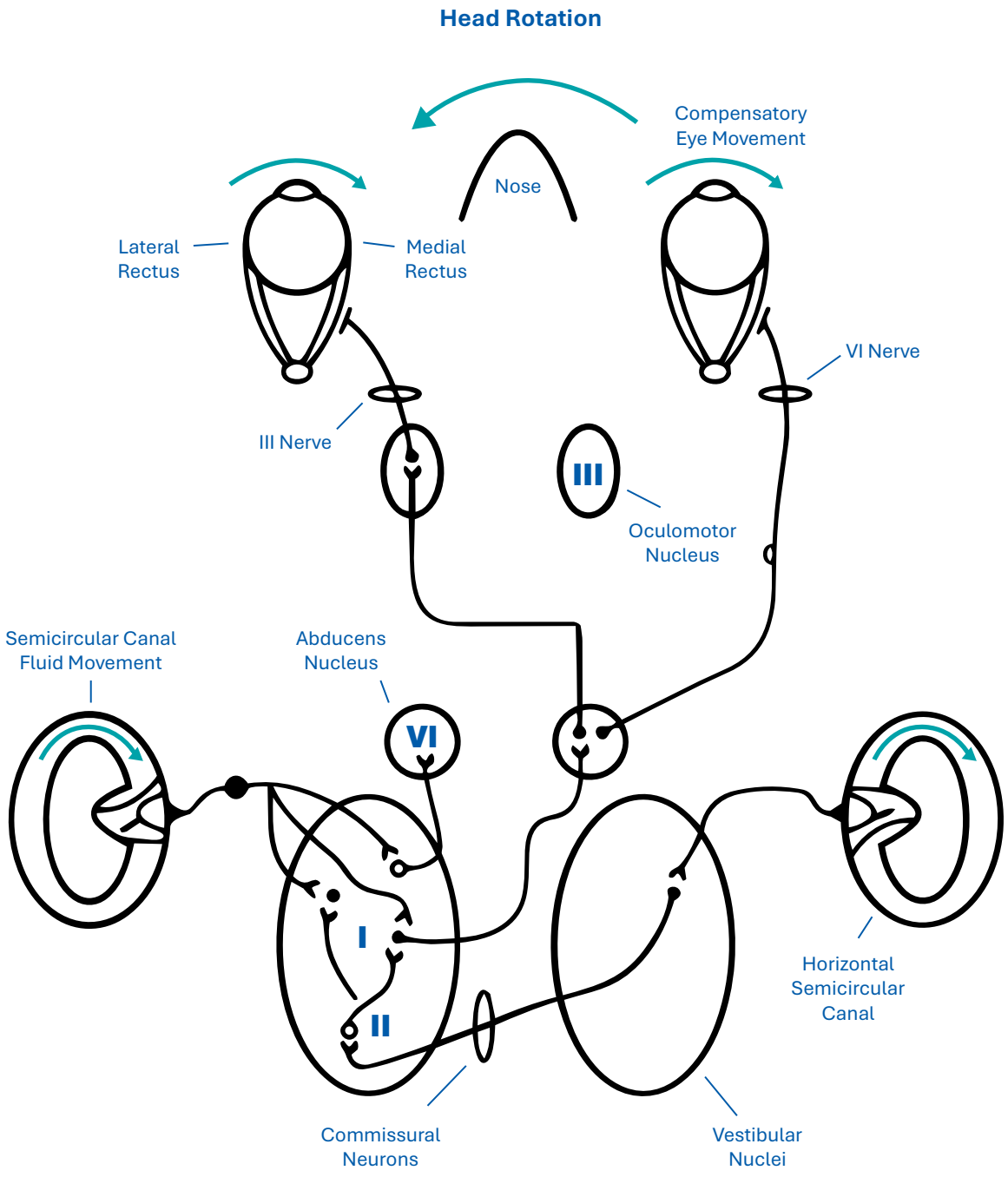


Figure 3.12: The vestibulo-ocular reflex (VOR) of the lateral (horizontal) canal

Rotational head movements activate angular VOR. When the head turns, the semicircular canals in the inner ear detect the angular acceleration and send signals to the oculomotor muscles, resulting in compensatory eye movements. For instance, during a rapid head turn to the right, the right eye will rotate leftward to maintain focus on an object, thus counteracting the head's movement. This reflex is crucial for maintaining visual acuity during high-speed activities, such as sports or driving.

The translational VOR responds to linear accelerations like those experienced during forward or backward head movements. The otolith organs (utricle and saccule) sense gravitational forces and linear motion, eliciting eye movements that stabilise vision in response to shifts in head position. This reflex ensures visual clarity when the head moves up or down or the body is in motion.

The vestibular aqueduct is a bony canal that connects the inner ear with the posterior cranial fossa. The vestibular aqueduct has a variable length averaging one and a half to eight millimetre in the adult. It houses the endolymphatic duct and sac, facilitating the transport of endolymph fluid, which is essential for the vestibular system to function adequately (Figure 3.13). The endolymphatic sac serves as a storage site for this fluid, assisting in pressure regulation within the inner ear and thereby contributing to the homeostasis of the auditory and vestibular systems. It also plays a role in the immunological function of the inner ear.

Figure 3.13: (a)

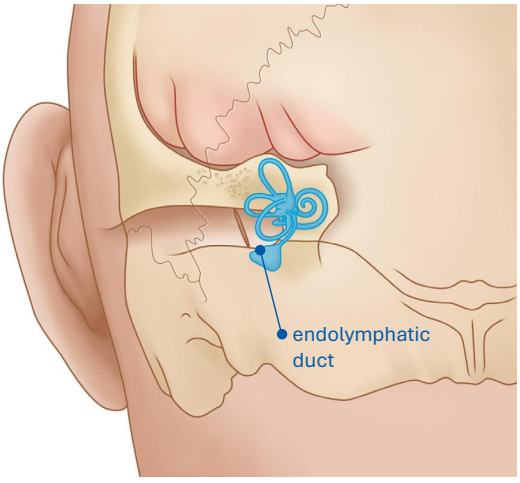


Figure 3.13: (b)

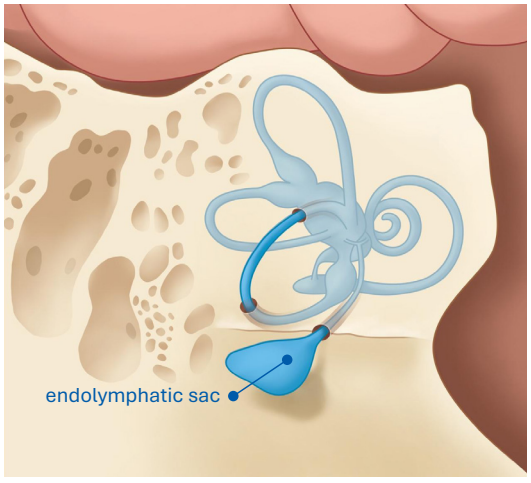


Figure 3.13: (a) The endolymphatic duct, (b) The endolymphatic sac

Internal auditory meatus

The internal auditory meatus is a bony canal located within the petrous portion of the temporal bone. It connects the inner ear with the cerebrospinal fluid-filled cerebellopontine angle of the posterior cranial fossa and the brainstem. The average diameter of the internal auditory meatus in neonates ranges from approximately five to seven millimetres, compared to seven to nine millimetres in adults. The internal auditory meatus is nine millimetres long in a baby and eleven millimetres in an adult.

The contents of the internal auditory meatus include the vestibulocochlear nerve (CN VIII) and the facial nerve (CN VII), both of which are essential for facial motor control (Figure 3.14a). The CN VIII consists of the cochlear nerve, which contains roughly 30,000 to 35,000 nerve fibres, and the vestibular nerve, which contains 20,000 nerve fibres.

Additionally, the canal houses the labyrinthine artery, a branch of the anterior inferior cerebellar artery (AICA), which supplies blood to the inner ear (Figure 3.14b).

Figure 3.14: (a)

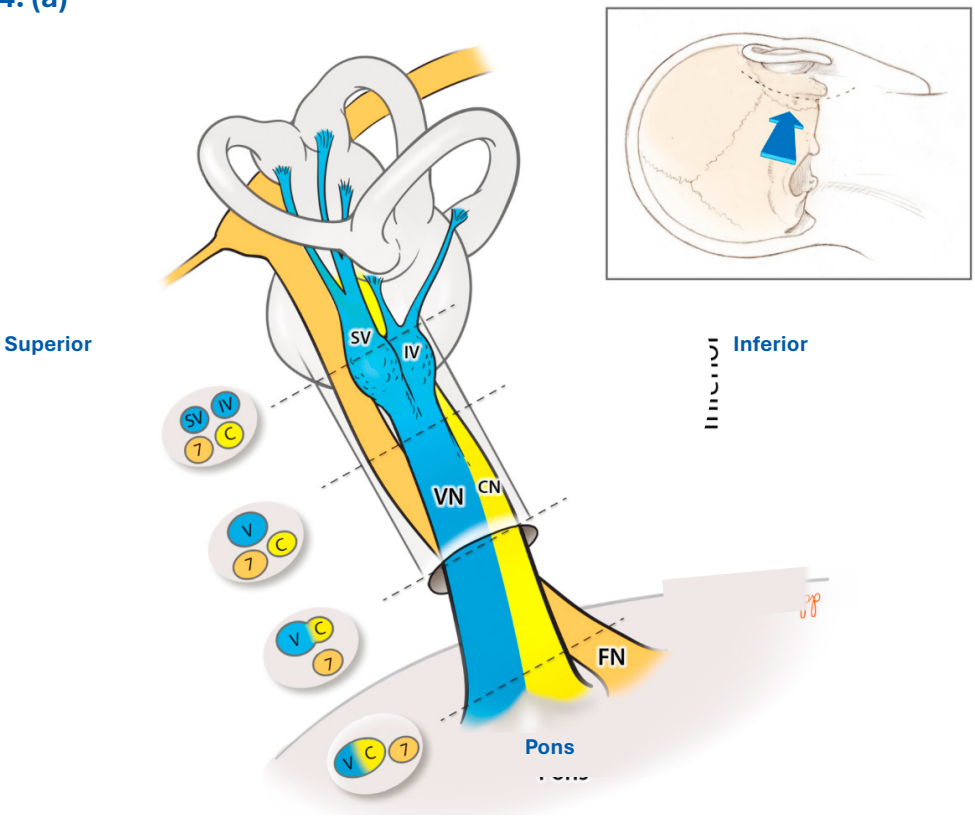


Figure 3.14: (b)

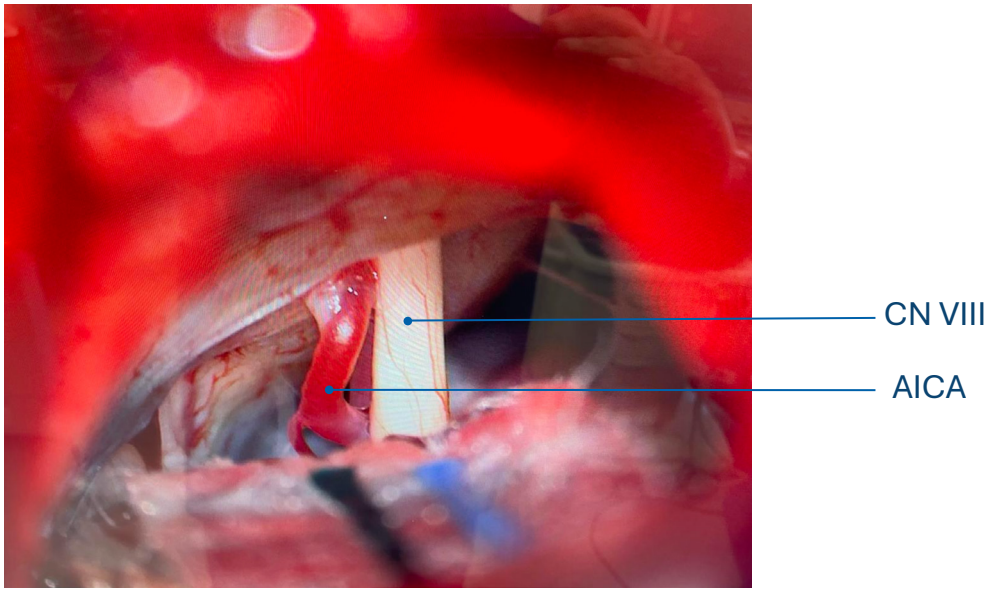


Figure 3.14: (a) Internal auditory meatus with FN and VCN consisting of the VN and CN with VN further dividing into the SV and the IV, (b) Surgical image of CN VIII and AIC

Brainstem nuclei and tracts of audiovestibular function

The brainstem auditory pathway begins in the cochlea, where auditory sensory cells transduce sound vibrations into neural signals, activating the auditory nerve. These signals first converge in the cochlear nuclei, the primary auditory nuclei in the brainstem (Figure 3.15).

The cochlear nuclei have two main components: the dorsal cochlear nucleus (DCN) and the ventral cochlear nucleus (VCN). The DCN processes complex sounds and contributes to sound localisation and discrimination, while the VCN primarily involves sound timing and intensity coding. After initial processing, auditory signals from the cochlear nuclei project to several important brainstem nuclei, including the superior olivary complex (SOC), the lateral lemniscus (LL) and the inferior colliculus (IC).

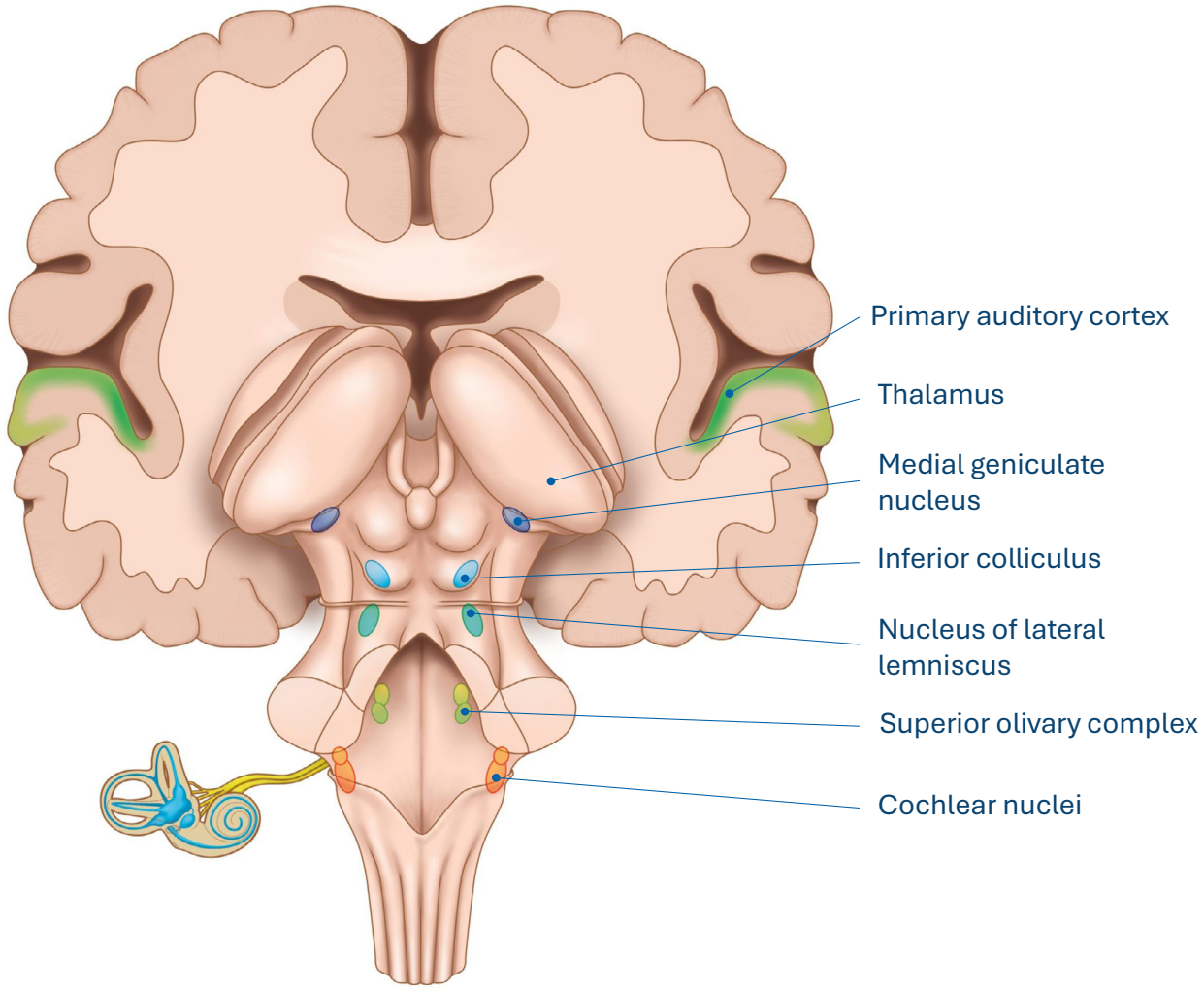


Figure 3.15: Cochlear nuclei, the primary auditory nuclei

The SOC is crucial for binaural hearing, as it receives inputs from both ears and integrates these signals to determine the direction of sound sources. The LL carries information from the SOC and other nuclei to the IC. The IC acts as a relay station that further processes auditory signals, integrating various aspects of sound, including frequency, intensity and timing, before sending information to the thalamus and, ultimately, to the auditory cortex.

The brainstem vestibular nuclei are located in the medulla and pons (Figure 3.16). These nuclei include the superior, lateral, medial and inferior vestibular nuclei. They receive input from the vestibular apparatus in the inner ear. The connections of the vestibular nuclei extend to several pivotal pathways, including the olivary nuclei. From the olivary nuclei, efferent projections travel to motoneurons in the spinal cord via the vestibulospinal tract, facilitating postural adjustments and reflexive head movements.

The medial longitudinal fasciculus (MLF) is the neural pathway that co-ordinates eye movements and integrates sensory information for ocular control. This white matter tract extends longitudinally through the brainstem, interconnecting several cranial nerve nuclei responsible for eye and head movements, notably those of the oculomotor (III), trochlear (IV) and abducens (VI) nerves.

The MLF facilitates communication between the vestibular nuclei and the nuclei of the extraocular muscles, thereby enabling the co-ordination of conjugate gaze and the maintenance of visual stability during head movements. It plays a vital role in the vestibulo-ocular reflex (VOR), stabilising the visual field by adjusting eye positioning in response to head movement.

The vestibular nuclei also connect via the olivary nuclei with the cerebellum, primarily through the inferior cerebellar peduncle, contributing to fine-tuning balance and co-ordination.

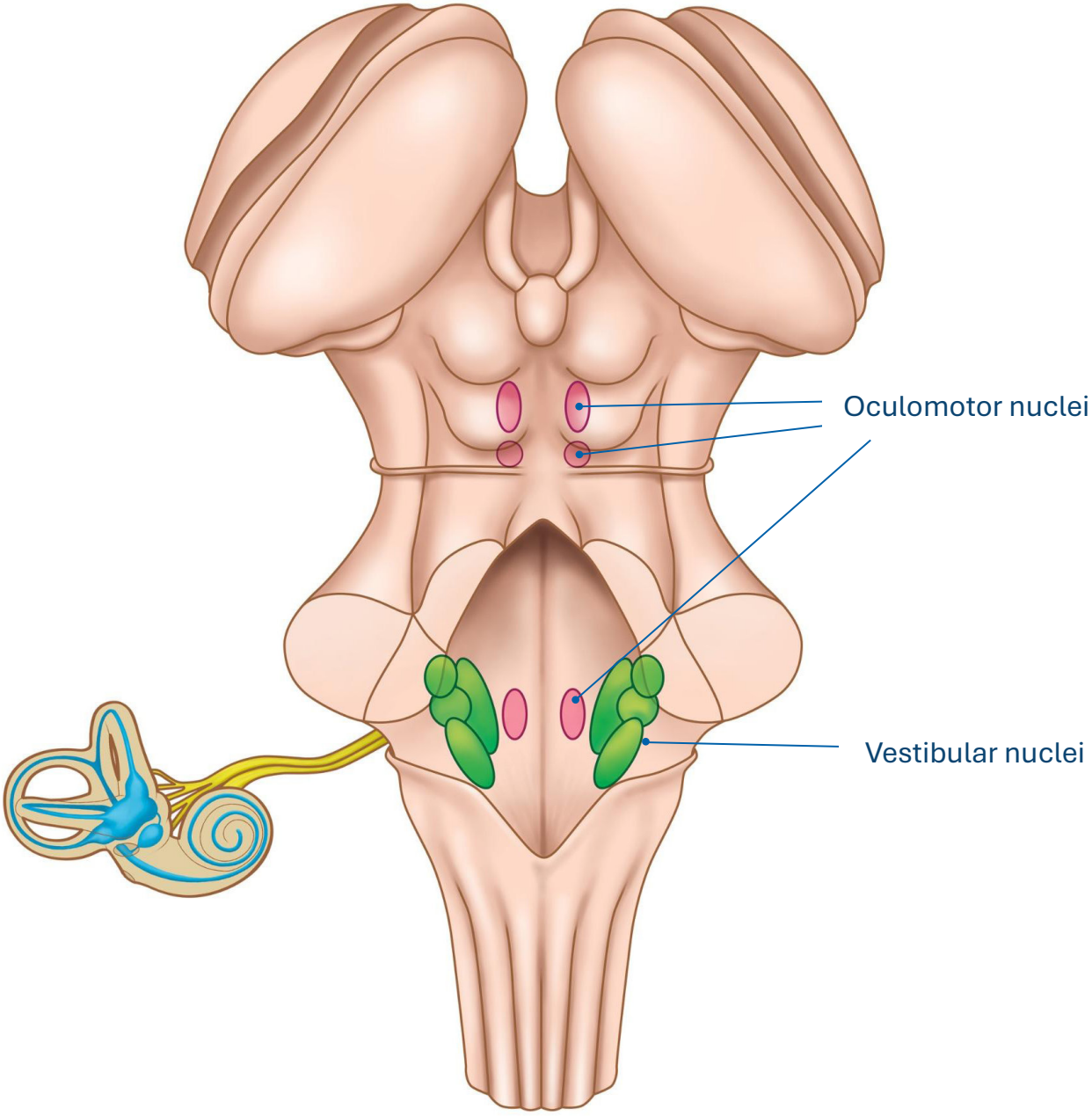


Figure 3.16: Brainstem vestibular and oculomotor nuclei

The vestibulospinal reflex (VSR) and vestibulocollic reflexes (VCR) are two other neural pathways that maintain postural stability and spatial orientation.

The vestibulospinal reflex facilitates balance and posture by transmitting signals from the vestibular apparatus in the inner ear to the spinal cord. This reflex influences extensor muscle activity in response to head and body position changes. This reflex plays a vital role during locomotion, particularly when the body encounters perturbations, ensuring the centre of mass remains stable.

The vestibulocollic reflex is primarily responsible for stabilising the head in relation to the body. It involves bilateral pathways that project from the vestibular nuclei to the neck motoneurons, allowing for co-ordinated head movements that counteract sudden changes in posture or position. Although less important in humans, this reflex is essential during activities such as walking or running, where head stability must be maintained despite the dynamic movement of the body.

Vestibulocerebellum

The vestibulocerebellum is primarily responsible for regulating balance and spatial orientation. (Figure 3.17). It comprises the flocculonodular lobe and is intricately connected to vestibular nuclei in the brainstem, facilitating the integration of vestibular information. Through its modulation of muscle tone and co-ordination of eye movements, the vestibulocerebellum plays a pivotal role in maintaining postural stability and enabling smooth, co-ordinated movements.

The vestibulocerebellum assesses and adapts to head position and motion changes, allowing for real-time adjustments to balance and equilibrium. Dysfunction in this area can lead to disorders such as ataxia, where affected individuals may experience difficulty in maintaining posture or executing coordinated movements. Furthermore, its influence extends to ocular motor control, exemplified by the VOR, which stabilises vision during head movements.

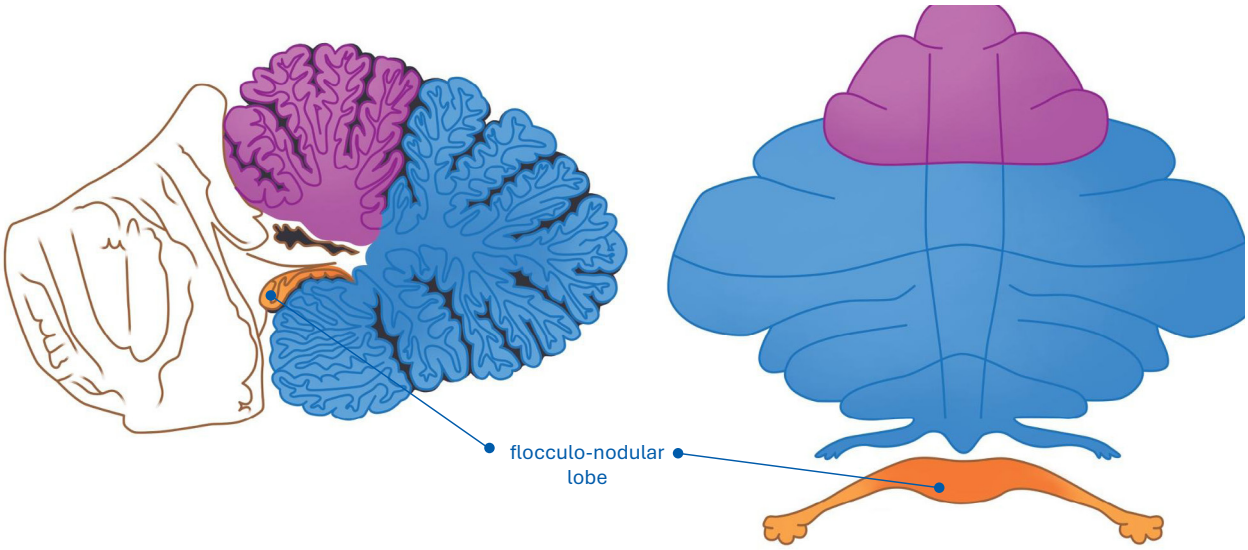


Figure 3.17: The vestibuocerebellum (flocculo-nodular lobe)

Auditory and vestibular cortex

The auditory processing pathway begins with signals originating from the inferior colliculus, ascending to the thalamus's medial geniculate nucleus (MGN). This structure acts as a vital relay station, where auditory signals undergo refinement before transmitting to the auditory cortex. The pathway that connects the MGN to the primary auditory cortex (A1), located in the superior temporal gyrus of the temporal lobe, is called auditory radiation. This neural pathway is essential for auditory perception and localisation, as it integrates information that enables the differentiation of sounds amidst complex auditory environments.

The auditory cortex can be divided into several distinct functional areas, with A1 playing a crucial role in the initial processing of auditory stimuli. The organisation of A1 is tonotopic, which signifies that it spatially arranges different sound frequencies (Figure 3.18). Adjacent regions, such as the secondary auditory cortex, are involved in higher-order processing tasks, such as sound recognition, association and integration of auditory information with other sensory modalities. This hierarchical structure facilitates increasingly sophisticated interpretations of auditory inputs, ranging from basic pitch detection to the complex recognition of patterns inherent in spoken language and music.

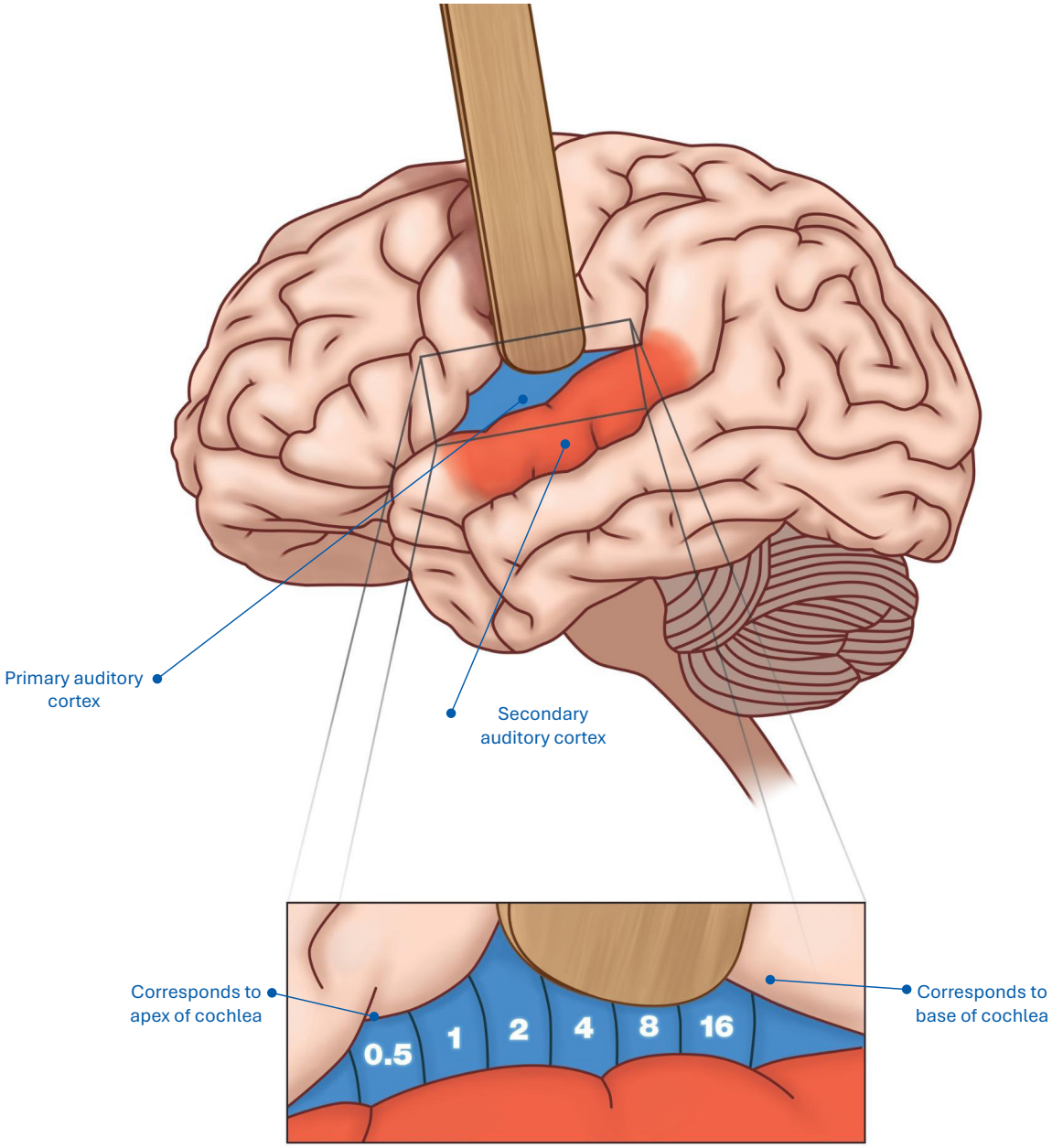
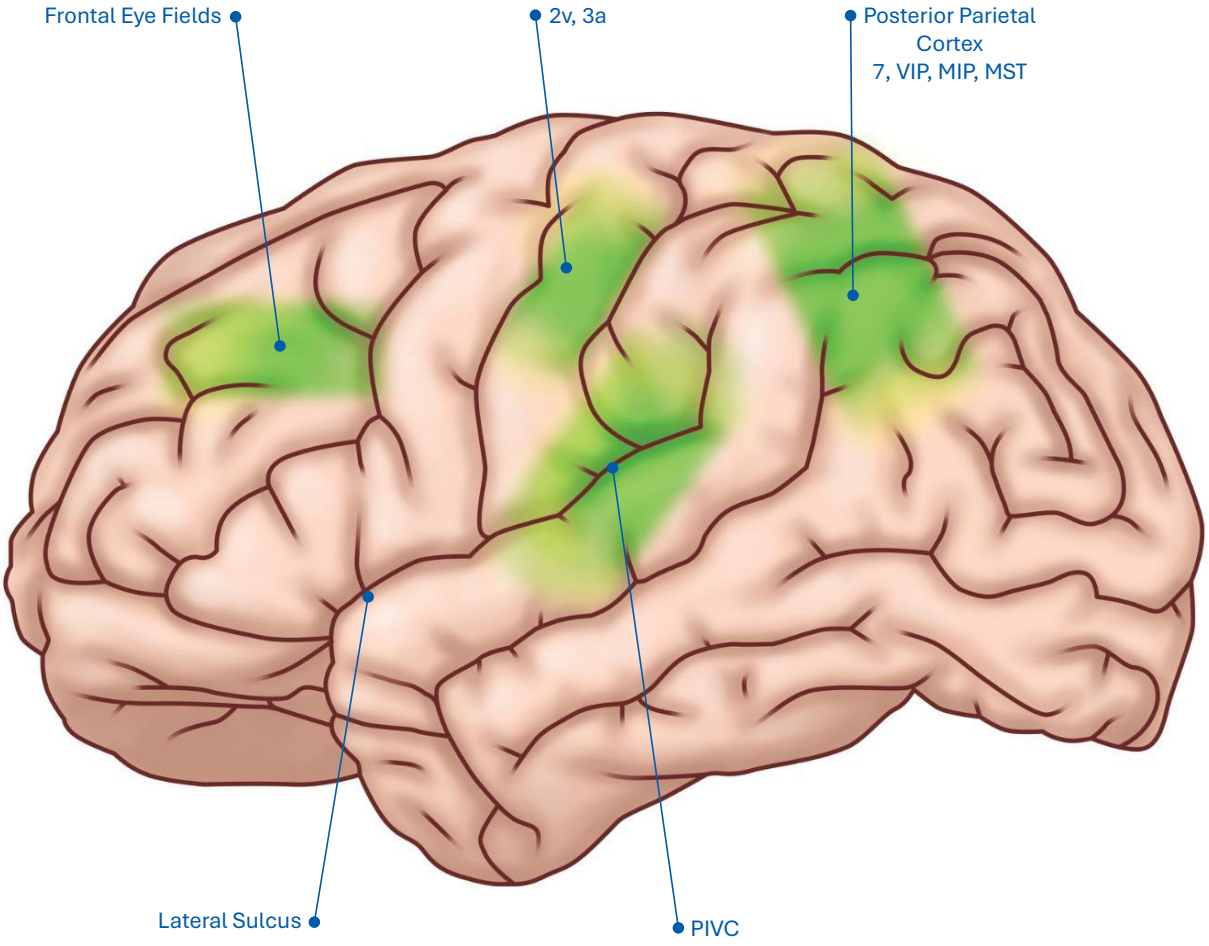


Figure 3.18: The auditory cortex A1 with tonotopic arrangement for different frequencies (kHz)

The vestibular cortex is crucial in processing sensory information related to balance, spatial orientation, head movements and position relative to gravity and motion. The vestibular cortex receives input from the vestibular system, located primarily within the insular and parietal lobes, particularly in areas such as the posterior insula and the parietal operculum.

The vestibular cortex is not isolated in function but works with other sensory modalities, including visual and proprioceptive inputs. Different cortical regions (Figure 3.18) of the brain known to be involved with vestibular processing are summarised in Table 3.4.



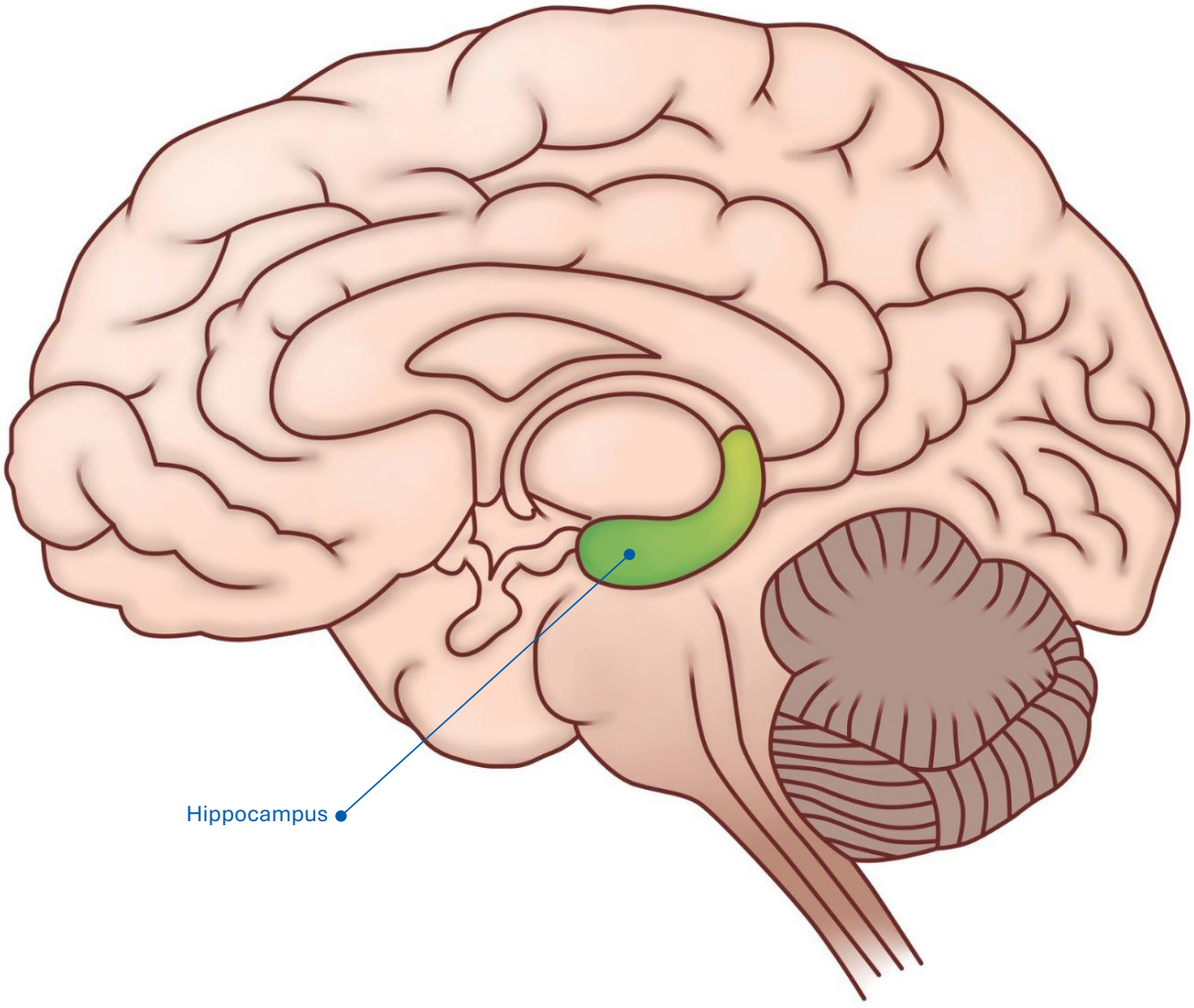


Figure 3.19: The vestibular cortex and associated cortical regions of vestibular processing

This multimodal integration enhances the brain's ability to construct a coherent spatial orientation and movement perception through complex interactions among the vestibular, visual and somatosensory systems.

Table 3.4: Cortical areas associated with vestibular processing. Source: Angelaki and Dickman (2024)

Cortical area	Function
Frontal eye fields (FEF)	Eye movement regulation
Somatosensory areas 2v and 3a	Body location and movement signal mapping
Posterior insular vestibular cortex (PIVC)	Body and head motion processing
Posterior parietal cortex Area 7 Ventral intraparietal area (VIP) Medial intraparietal area (MIP) Medial superior temporal area (MST).	Motion perception
Hippocampus and parahippocampal regions	Spatial orientation and navigation



Treasures of Wisdom

Auditory and vestibular cortex

- The brainstem nuclei are essential in generating different kinds of eye movements. The upper midbrain region is vital for vertical eye movements and the lower pons for horizontal eye movements.
- The cerebellum is mainly involved with the accuracy of eye movements and the ability to suppress the vestibulo-ocular reflex during head movements.
- In addition, damage to the vestibulocerebellum could lead to ataxia.
- Damage to the numerous cortical vestibular areas may also lead to spatial disorientation.

Development of the Paediatric Audiovestibular System

Maturation of the temporal bone

The temporal bone in neonates is predominantly composed of cartilage and is not fully developed at birth (Moore, Persaud and Torchia 2018). The squamous portion, exhibiting minimal ossification, initiates its ossification process shortly after birth through intramembranous ossification, vital for flat bone formation. During the early years of life, this area undergoes continuous ossification, progressively obscuring the sutures that connect it to adjacent bones, thereby

indicating structural integration. Notable ossification occurs in the formation of the roof of the external auditory canal, with significant development evident between ages eight and twelve, approaching the adult morphology. Full maturation occurs in adolescence.

The mastoid process is also underdeveloped at birth. It experiences significant postnatal growth influenced by surrounding anatomical structures, notably the sternocleidomastoid muscle, which exerts mechanical forces that facilitate its development. During the first two years of life, the mastoid undergoes intramembranous and endochondral ossification and is fully pneumatised by age twelve, albeit with individual size variability.

The petrous part, present at birth as the primary ossification centre, remains largely unossified and is one of the last regions to ossify, primarily through endochondral ossification. Its development, crucial for hearing and balance, progresses significantly through childhood, with substantial changes noted by age twelve. Similarly, the tympanic part is largely cartilaginous at birth. It commences ossification through mesenchymal condensation shortly thereafter, culminating by the end of the second year, reaching its final shape in five years.

Maturation of the outer ear

The pinna undergoes significant growth during early childhood, with the most rapid enlargement occurring within the first two years. By age twelve, the pinna reaches six centimetres, approximately 90% of its adult size. The width dimensions mature earlier, at five and a half to eleven years, whereas the maturity age of length dimensions varies from twelve to sixteen years (Purkait 2013). It eventually acquires a more defined shape that enhances its ability to funnel sound waves into the ear canal, facilitating enhanced sound localisation and resonance, which is vital for effective auditory.

The external auditory meatus gradually elongates and becomes more S-shaped as the child grows, reaching approximately three centimetres by age twelve. Despite their anatomical maturity at birth, the ceruminous and sebaceous glands do not achieve their full functional capacity until puberty.

Maturation of the middle ear

The middle ear space, initially small and less ventilated in neonates, expands as the child grows, reaching its adult size by approximately twelve years (Mishra, Dinger and Renken

2017). This process facilitates better aeration and drainage through the ET. This expansion also enhances the ability to transmit sound waves effectively. Different frequencies follow different maturational timelines.

At birth, the TM is thick and relatively flat and horizontal. It gradually adopts a more oblique position and becomes thinner as the child matures, which enhances sound transmission. The ossicles begin developing in utero and fully ossify by the end of infancy. Their arrangement in the middle ear includes size increases and maturation of their joints and muscles, improving their mechanical advantage in sound conduction. The majority of the remodelling of the bone in the ossicles takes place in the first months and years of life, while afterwards, only minor reorganisation occurs (Rolvien et al. 2018).

As the child matures, the eustachian tube elongates and angles downward, improving ventilation and drainage of the middle ear. It reaches its adult configuration by age seven (Takasaki et al. 2007).

Maturation of the inner ear

The inner ear structures are responsible for auditory and balance functions, along with the auditory and vestibular nerves that facilitate sensory perception.

The cochlea undergoes significant development and maturation from birth to age twelve. Intrinsic genetic factors and extrinsic environmental influences can influence cochlear development. At birth, the cochlea is anatomically present but functionally immature. The sensory hair cells reach their highest density shortly after birth, with no regeneration occurring after the first week (Heuermann et al. 2022). Auditory thresholds are up to 25 dB higher in neonates, typically around 40–60 dB HL (hearing level), indicating that while some auditory perception exists, it is far from optimal (Sininger, Abdala and Cone-Wesson 1997).

Critical growth and differentiation processes occur during the first few months of life. Cochlear hair cells begin to establish synaptic connections with auditory neurons (Johnson et al. 2019). The maturation of the organ of Corti leads to enhanced sensitivity to sound frequencies, allowing infants to respond to environmental auditory stimuli and facilitating language acquisition. The audiovestibular nerve undergoes substantial development during the first year of life.

The cochlea is adult-sized at birth, although cochlear size variability is present among individuals.

The frequency selectivity of hair cells improves as they mature, which is crucial for understanding speech among background noise (speech-in-noise perception)—a skill increasingly demanded in social interactions and educational environments. Over the subsequent years, typically within four to eight years, this maturation of the cochlea drives the auditory brain toward attaining its optimal performance levels, contingent upon the specific perceptual tasks involved (Pujol, Lavigne-Rebillard and Uziel 1991). Normal maturation of the cochlea, reaching its maximum performance at birth, will slowly (within four to eight years, depending on the perceptual task) drive a normal auditory brain to the acquisition of its maximum performance.

It is essential to consider the impact of auditory exposure on cochlear development. Children frequently exposed to rich auditory environments typically exhibit better auditory processing and language skills (Atılgan, Cesur and Çiprut 2023). Conversely, those with hearing loss may experience delays in cochlear maturation and subsequent language development, emphasising the interactive nature of auditory experiences during this critical developmental window.

The peripheral vestibular system, essential for balance and spatial orientation, comprises several critical structures, including the utricle, saccule and semicircular canals. These components, along with the endolymphatic duct and sac, develop intricately from birth to age twelve, adapting to the sensory demands of a maturing child.

The morphology of the semicircular canals remains largely unchanged following birth; however, a notable tendency for these structures to tilt anteriorly as a cohesive unit emerges with age. The anterior tilt of the semicircular canals may arise from age-related adjustments in the surrounding cranial and cervical structures, effectively altering the orientation of these canals without altering their fundamental morphological characteristics (Lyu et al. 2016).

Maturation of the central auditory system

The central auditory system undergoes a complex process of maturation and development, beginning in the cochlear nucleus in the brainstem and extending to various auditory nuclei, thalamic relay centres, and, ultimately, the auditory cortex and associated speech/language

areas. This intricate developmental trajectory is crucial for effective sound processing and the emergence of speech and language capabilities.

In neonates, the cochlear nucleus already exhibits a rudimentary tonotopic organisation—a spatial arrangement where different frequencies are processed in discrete regions. As the child matures into early childhood, the connectivity of the cochlear nucleus with subsequent brainstem auditory nuclei, such as the superior olivary complex and the inferior colliculus, becomes more sophisticated. The neural circuits in these areas demonstrate increased synaptic density by twelve months, and myelination—essential for efficient synaptic transmission—is usually completed in two years (Prado and Dewey 2014).

Electrophysiological measures, particularly Auditory Brainstem Responses (ABRs), serve as critical tools in assessing the maturation of the auditory brainstem in infants and young children (Chapter 6). ABRs provide insights into the integrity and development of auditory pathways by measuring neural activity in response to auditory stimuli. In the first year of life, ABRs undergo significant changes, reflecting the ongoing maturation of both peripheral and central auditory structures. Children often exhibit ABR waveforms that closely resemble adult patterns by age two (Johnson et al. 2008), indicating the establishment of fully developed brainstem processing capabilities.

While children exhibit uniform neural responses to click stimuli, distinct differences arise when processing speech sounds. Specifically, three—to four-year-old children show delayed and less synchronous neural onset and sustained responses compared to their five - to twelve-year-old counterparts. These findings highlight the critical period of auditory maturation, with significant strides in the neural encoding of speech around the age of five (Johnson et al. 2008).

Auditory signals ascend to the thalamus's medial geniculate body (MGB). The MGB acts as a critical relay centre, further refining auditory information before it reaches the A1. During the late neonatal period and into early childhood, the thalamic neurons that project to the auditory cortex undergo significant maturation. The connectivity between the MGB and auditory cortex strengthens over time, with enhancing synaptic inputs that assist in complex sound processing. The first two years of life are essential for forming these auditory thalamocortical connections. As neural pathways consolidate, the auditory cortex becomes increasingly responsive to various auditory stimuli. Children start to process sounds akin to adults by age three to four, reflecting the substantial myelination and synaptic development that occurred in previous years.

The primary auditory cortex (A1) is the next stage in auditory processing, where complex sound patterns are interpreted. The maturation of A1 is marked by the development of advanced auditory functions, including sound localisation, frequency discrimination and temporal processing—all vital for language development.

The superficial layers of the auditory cortex do not reach full maturity until adolescence. While myelination is observable across various cortical layers by age six, the process of myelination further progresses into adolescence (Hallet and Proctor 1996). This enhanced myelination facilitates more efficient and rapid neural transmission both within hemispheres and across different cortical areas. Additionally, the complexity of dendritic branching structures continues to evolve until approximately twelve years. These maturation processes are crucial, as they contribute to the establishment of effective auditory processing abilities.

In parallel, significant developments occur in central speech and language areas, like Broca's and Wernicke's areas. While these areas do not process auditory information directly, the maturation of auditory cortex functions is foundational for linguistic development. The sensitivity to phonetic contrasts develops rapidly during the first year, influenced by exposure to the ambient language environment (Kuhl and Rivera-Gaxiola 2008). Children begin to exhibit a categorical perception of phonemes, which is critical for developing language.

Maturation of the central vestibular system

The central vestibular system, encompassing the vestibular nuclei, vestibulocerebellum and connections to various brain regions, including the thalamus and vestibular cortex, undergoes substantial maturation from the neonatal stage until approximately eleven to fifteen years of age. This developmental trajectory facilitates increasingly sophisticated reactions to vestibular stimuli.

Following birth, the vestibular nuclei initiate the processing of signals from the fully developed semicircular canals and otolith organs; however, the central integration of these signals remains immature. As infants grow, synaptic connections within the vestibular nuclei expand, notably with ocular motor nuclei, which are crucial for refining the VOR and enhancing visual stability while moving.

2 years of age: The brain's fundamental structural and functional architecture seems to be in place, and the brain maturation that occurs in later childhood slows.

2-3 years of age: Significant balance and postural control improvements are observed, attributed to both vestibular maturation and thalamic integration (Shumway-Cook and Woollacott 2017).

3 years of age: The vestibular system reaches a crucial developmental milestone. The vestibulocerebellum significantly increases connectivity and functional specialisation. This maturation is evident as children engage in more complex activities like walking and climbing. The system continues evolving into school age, improving spatial orientation and postural control, essential for engaging in sports and dynamic activities (Rine 2018).

7 years of age: Vestibular thalamic functions are largely mature.

10-11 years of age: The hippocampus peaks in volume. Connectivity progresses differently in hippocampal subregions, reaching adult levels

15 years of age: The vestibular function reaches adult-like values (O'Reilly et al. 2011). The vestibulo-ocular, vestibulo-spinal and vestibulo-collic reflexes are now fully developed. Children can now perform intricate tasks requiring balance, co-ordination and spatial awareness with increased competence, thanks to the maturation of the vestibular pathways connecting the brainstem and cortex (Figure 3.20).

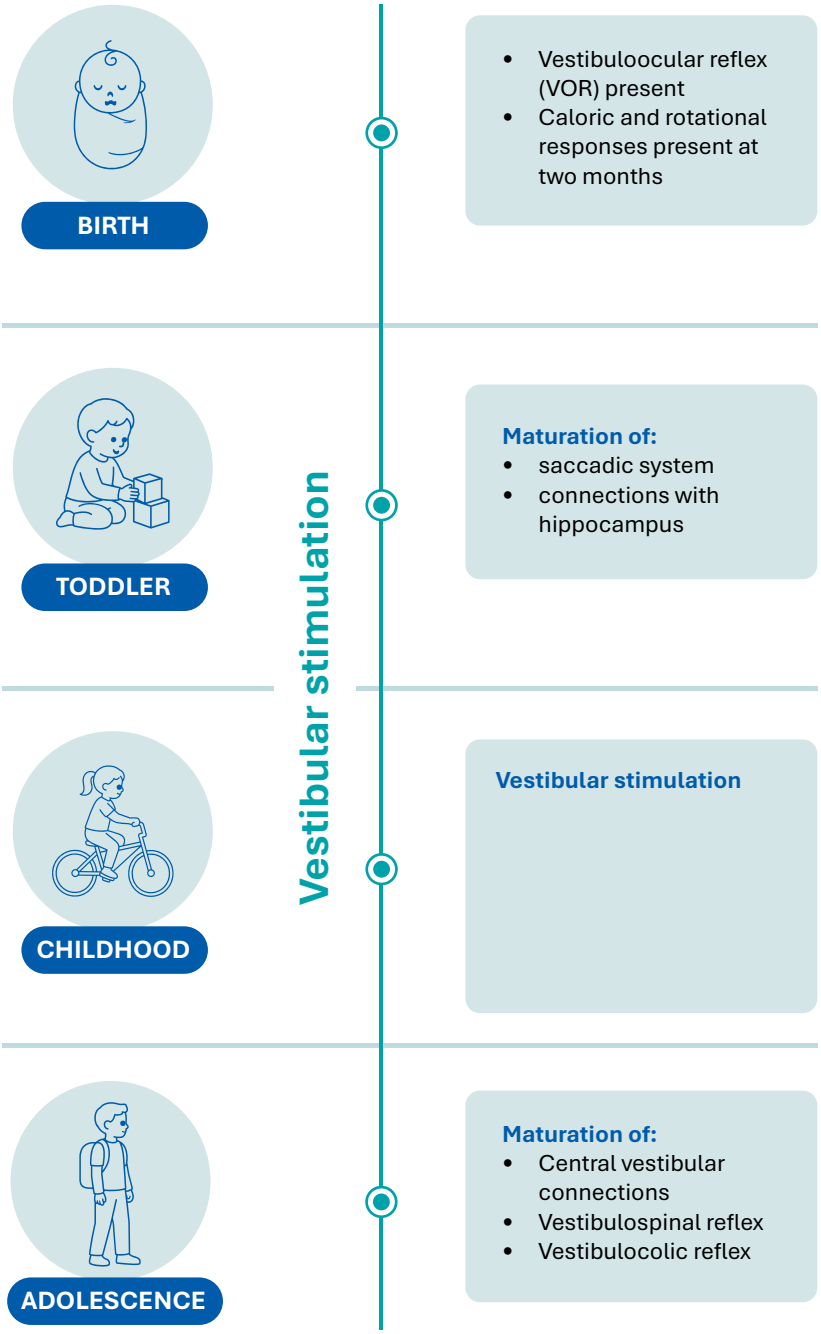


Figure 3.20: Postnatal development of the vestibular system. Source: Urbančič, Battelino and Vozel (2024)

The Human Connectome and the Auditory Cortex

A connectome represents a comprehensive map of effective synaptic connections and neural projections that constitute a nervous system and significantly influence its communicative functions. This intricate network is crucial for understanding the architecture of the brain and the interplay between sensory systems and cognitive processes.

The auditory system plays a pivotal role in the structure and function of the human connectome. Information is processed through auditory pathways, leading to the development of speech and language capabilities. Research highlights the profound impact of auditory experiences on the brain's connectivity, affirming that early auditory input shapes neural circuits essential for communication (Kral et al. 2016).

The implications of childhood hearing loss (CHL) extend beyond mere auditory deficits; they permeate the very fabric of the connectome. Characterising HL as a connectome disease underscores the importance of addressing auditory health holistically. Hearing impairments disrupt the intricate web of synaptic connections, potentially leading to developmental and cognitive challenges.

Neuroplasticity

Neuroplasticity, often referred to as brain plasticity or neural plasticity, is the ability of the central nervous system to adapt and reorganise itself in response to experience, learning and environmental changes. This phenomenon enables the brain to form new neural connections throughout an individual's life, allowing for functional recovery following injury and adaptation to



Treasures of Wisdom

Maturation of the ear

- It is important to frequently check the fit of earmoulds due to rapid growth in the first two years
- Neural maturation involves myelination and synaptic formation, enhancing auditory processing and vestibular reflexes.
- By age two, children can typically localise sounds and differentiate between varying sound properties, which is critical for developing communication skills.
- By age twelve, the otolith organs achieve functional maturity. The co-ordination between the utricle, saccule and semicircular canals is finely tuned, enabling the child to perform complex physical activities and respond efficiently to environmental stimuli.
- By ages five to twelve, children refine their linguistic competencies. The connections between the auditory cortex and these language areas become robust, facilitating language production and comprehension. The maturation of the auditory system and associated language areas occurs concurrently, highlighting the interdependence of auditory processing and language acquisition.

new challenges. Neuroplasticity operates on various levels, from cellular changes (involving individual neurons) to large-scale cortical remapping.

By the end of the first year, the cortex comprises all six layers, signifying the maturation of its structural complexity. In the first three and a half years of life, neurons rapidly multiply in the cortex, especially in the visual, auditory and prefrontal regions. This phase is characterised by creating an overabundance of synaptic connections, laying the foundational architecture for future cognitive functioning.

During this crucial developmental window, myelination of axons also occurs at an accelerated rate, facilitating faster neural communication. Following this period of growth, a significant pruning phase begins around the age of four, during which unnecessary neurons and synapses are eliminated from the visual, auditory and prefrontal cortices. This synaptic pruning is essential for optimising neural efficiency and functionality, ensuring the remaining connections are robust and relevant to the individual's experiences (Sharma and Cardon 2015).

The Human Balance System

Balance is the ability to maintain the body's centre of mass in alignment with its support base. The balance system reaches its peak maturity around the age of twelve to fifteen. By age two, children exhibit significantly improved balance skills, demonstrating foundational co-ordination in activities such as walking and running. The balance system continues to refine through childhood, with critical developments occurring during pre-adolescence.

This balance function is sustained through a sophisticated interplay of sensorimotor control systems, encompassing sensory inputs from vision, proprioception and the vestibular system. Vision provides essential information about the surrounding environment, while proprioception offers feedback on body position and movement through touch. The vestibular system is critical in maintaining motion, equilibrium and spatial orientation.

Effectively integrating these sensory inputs is crucial for achieving stability, which is then translated into motor outputs that engage the eye and body muscles. When these systems suffer from injury or deficiency, the individual's balance can be significantly compromised. This can manifest in difficulties with co-ordination and stability, ultimately affecting one's ability to perform daily activities safely and efficiently.

Conclusion

A thorough understanding of the anatomy and physiology of paediatric hearing and vestibular structures is paramount for the practical evaluation and treatment of patients. The intricate nature of embryological development in these systems lays the foundation for their subsequent functionality and pathology. As children grow, their auditory and vestibular systems undergo significant maturation, influenced by genetic and environmental factors. This maturation process is critical in identifying deviations that may lead to audiological or balance disorders.

Evaluating the developmental milestones of these structures enables clinicians to detect abnormalities early, facilitating timely interventions that can significantly enhance outcomes. Knowledge of the anatomical variations and physiological norms at various developmental stages equips healthcare professionals with the insights necessary to tailor assessment and therapeutic strategies to the unique needs of paediatric patients. Moreover, understanding the interplay between anatomy, physiology and maturation can illuminate the reasons behind specific clinical presentations, guiding more effective treatment approaches.

Paediatric practitioners must comprehensively grasp these fundamental concepts to ensure they can provide optimal care and support for children's auditory and vestibular health.



Test your knowledge:

Based on the content covered answer the following questions:

1. What is the critical period of development of the inner ear and why is knowledge of this important?
2. Describe the anatomy of the middle ear and explain how it differs between a child and an adult.
3. Explain what is the critical period of vestibular development in a child and why it is important.

References

Ahsan, M., Riley, K.L. and Schubert, F.R. 2007. Molecular mechanisms in the formation of the medial longitudinal fascicle. *Journal of Anatomy*, 211(2): 177–187. https://doi.org/10.1111/j.1469-7580.2007.00774.x

Amarnath, C., Sathyan, G., Soniya, R., Periakaruppan, A.L. and Shankar, K.S. 2016. Evaluation of embryological sequences of ear anomalies and its radiological relevance. *Indian Journal of Otology*, 22(4): 248–257. [Online]. Available at: https://pdfs.journals.lww.com/ijoo/2016/22040/evaluation_of_embryological_sequences_of_ear.5.pdf (Accessed 30 December 2025).

Amore, G., Spoto, G., Ieni, A., Vetri, L., Quatrosi, G., Di Rosa, G. and Nicotera, A.G. 2021. A focus on the cerebellum: From embryogenesis to an age-related clinical perspective. *Frontiers in Systems Neuroscience*, 15. https://doi.org/10.3389/fnsys.2021.646052.

Angelaki, D.E. and Dickman, J.D. n.d. *The Vestibular System*. Noba Project. [Online]. Available at: https://nobaproject.com/modules/the-vestibular-system (Accessed 3 February 2026).

Anniko, M. and Nordemar, H. 1982. Formation of the cupula cristae ampullaris: Development in vivo and in vitro. *American Journal of Otolaryngology*, 3(1): 31–40. https://doi.org/10.1016/s0196-0709(82)80030-6.

Antón-Bolaños, N., Espinosa, A. and López-Bendito, G. 2018. Developmental interactions between thalamus and cortex: A true love reciprocal story. *Current Opinion in Neurobiology*, 52: 33–41. https://doi.org/10.1016/j.conb.2018.04.018.

Atılgan, A., Cesur, S. and Çiprut, A. 2023. A longitudinal study of cortical auditory maturation and implications of the short inter-implant delay in children with bilateral sequential cochlear implants. *International Journal of Pediatric Otorhinolaryngology*, 166. https://doi.org/10.1016/j.ijporl.2023.111472.

Božanić Urbančič, N., Battelino, S. and Vozel, D. 2024. Appropriate vestibular stimulation in children and adolescents—A prerequisite for normal cognitive, motor development and bodily homeostasis—A review. *Children*, 11(1). https://doi.org/10.3390/children11010002.

Dhanasingh, A., Erpenbeck, D., Assadi, M.Z., Doyle, Ú., Roland, P., Hagr, A., Van Rompaey, V. and Van de Heyning, P. 2021. A novel method of identifying inner ear malformation types by pattern recognition in the mid modiolar section. *Scientific Reports*, 11. https://doi.org/10.1038/s41598-021-00330-6.

Dhanasingh, A. 2022. Embryology of Inner Ear Malformation Types and its Radiological Relevance. https://doi.org/10.1007/978-981-16-8217-9_1.

Heuermann, M.L., Matos, S., Hamilton, D. and Cox, B.C. 2022. Regenerated hair cells in the neonatal cochlea are innervated and the majority co-express markers of both inner and outer hair cells. *Frontiers in Cellular Neuroscience*, 16. https://doi.org/10.3389/fncel.2022.841864.

Jackler, R.K., Luxford, W.M., and House, W.F. 1987. Congenital malformations of the inner ear: A classification based on embryogenesis. *The Laryngoscope*, 97(S 40): 2–14. https://doi.org/10.1002/lary.5540971301.

Johnson, K.L., Nicol, T., Zecker, S.G. and Kraus, N. 2008. Developmental plasticity in the human auditory brainstem. *The Journal of Neuroscience* , 28(15): 4000–4007. https://doi.org/10.1523/JNEUROSCI.0012-08.2008.

Johnson, S.L., Safieddine, S., Mustapha, M. and Marcotti, W. 2019. Hair cell afferent synapses: Function and dysfunction. *Cold Spring Harbor Perspectives in Medicine*, 9. https://doi.org/10.1101/cshperspect.a033175.

Kral, A., Kronenberger, W.G., Pisoni, D.B. and O'Donoghue, G.M. 2016. Neurocognitive factors in sensory restoration of early deafness: A connectome model. *The Lancet. Neurology*, 15(6): 610–621. https://doi.org/10.1016/S1474-4422(16)00034-X.

Kuhl, P. and Rivera-Gaxiola, M. 2008. Neural substrates of language acquisition. *Annual Review of Neuroscience*, 31: 511–534. https://doi.org/10.1146/annurev.neuro.30.051606.094321

Lyu, H.Y., Chen, K.G., Yin, D.M., Hong, J., Yang, L., Zhang, T.Y. and Dai, P.D. 2016. The age-related orientational changes of human semicircular canals. *Clinical and Experimental Otorhinolaryngology*, 9(2):109–115. https://doi.org/10.21053/ceo.2014.02012.

Mishra, S.K., Dinger, Z. and Renken, L. 2017. Maturation of middle ear transmission in children. *Hearing Research*, 344: 62–67. https://doi.org/10.1016/j.heares.2016.10.029.

Moore, K.L., Persaud, T.V.N. and Torchia, M.G. 2018. The Developing Human E- Book: Clinically Oriented Embryology. Elsevier Health Sciences, Amsterdam.

O'Reilly, R., Grindle, C., Zwicky, E.F. and Morlet, T. 2011. Development of the vestibular system and balance function: Differential diagnosis in the pediatric population. *Otolaryngologic Clinics of North America*, 44(2): 251–271. https://doi.org/10.1016/j.otc.2011.01.001.

Prado, E.L. and Dewey, K.G. 2014. Nutrition and brain development in early life. *Nutrition Reviews*, 72(4): 267–284. https://doi.org/10.1111/nure.12102.

Pujol, R., Lavigne-Rebillard, M. and Uziel, A. 1991. Development of the human cochlea. *Acta Otolaryngologica. Supplementum*, 482: 7–12. PMID: 1897363.

Purkait, R. 2013. Progression of growth in the external ear from birth to maturity: A 2-year follow-up study in India. *Aesthetic Plastic Surgery*, 37: 605–616. https://doi.org/10.1007/s00266-013-0097-1.

Pyle G.M. 2000. Embryological development and large vestibular aqueduct syndrome. *The Laryngoscope*, 110(11): 1837–1842. https://doi.org/10.1097/00005537-200011000-00014.

Querleu, D., Renard, X., Versyp, F., Paris-Delrue, L. and Crèpin, G. 1988. Fetal hearing. *European Journal of Obstetrics, Gynecology, and Reproductive Biology*, 29: 191–212. https://doi.org/10.1016/0028-2243(88)90030-5.

Rine, R.M. 2018. Vestibular rehabilitation for children. *Seminars in Hearing*, 39(3): 334–344. https://doi.org/10.1055/s-0038-1666822.

Rolvien, T., Schmidt, F.N., Milovanovic, P., Jähn, K., Riedel, C., Butscheidt, S., Püschel, K., Jeschke, A., Amling, M. and Busse, B. 2018. Early bone tissue aging in human auditory ossicles is accompanied by excessive hypermineralization, osteocyte death and micropetrosis. *Scientific Reports*, 8. https://doi.org/10.1038/s41598-018-19803-2.

Scheuer, L. and Black, S. 2000. *Developmental Juvenile Osteology*. London: Elsevier.

Sharma, A. and Cardon, G. 2015. Cortical development and neuroplasticity in Auditory Neuropathy Spectrum Disorder. *Hearing research*, 330(Pt B): 221–232. https://doi.org/10.1016/j.heares.2015.06.001.

Sheridan, N. and Tadi, P. 2024. Neuroanatomy, Thalamic Nuclei. [Updated 2023 Jul 24]. In *StatPearls*. Treasure Island, FL: StatPearls Publishing[Online]. Available at: https://www.ncbi.nlm.nih.gov/books/NBK549908/. (Accessed 6 January 2025).

Shumway-Cook, A. and Woollacott, M.H. [1995] 2017. *Motor Control: Translating Research into Clinical Practice*. 5th ed. Lippincott Williams and Wilkins.

Sininger, Y.S., Abdala, C. and Cone-Wesson, B. 1997. Auditory threshold sensitivity of the human neonate as measured by the auditory brainstem response. *Hearing Research*, 104(1-2): 27–38. https://doi.org/10.1016/s0378-5955(96)00178-5

Takasaki, K., Takahashi, H., Miyamoto, I., Yoshida, H., Yamamoto-Fukuda, T., Enatsu, K. and Kumagami, H. 2007. Measurement of angle and length of the eustachian tube on computed tomography using the multiplanar reconstruction technique. *The Laryngoscope*, 117(7): 1251–1254. https://doi.org/10.1097/MLG.0b013e318058a09f

Yoshimura, H., Okubo, T., Shinagawa, J., Nishio, S.Y., Takumi, Y. and Usami, S.I. 2024. Epidemiology, aetiology and diagnosis of congenital hearing loss via hearing screening of 153913 newborns. *International Journal of Epidemiology*, 53(3). https://doi.org/10.1093/ije/dyae052