

Lecture - Sensory Development

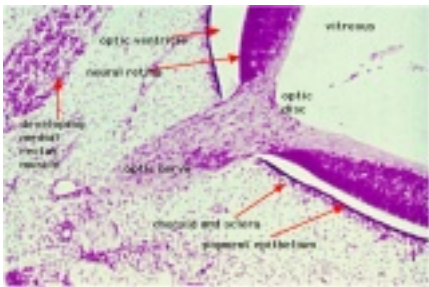
From Embryology

Introduction

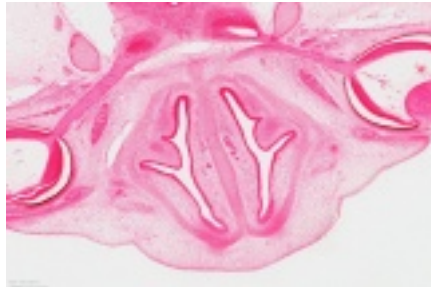
This lecture will introduce development of the special sensory structures associated with hearing, vision, smell and taste. Due to time limitations the lecture will focus on hearing development and if time is available vision and other senses will be introduced in general.



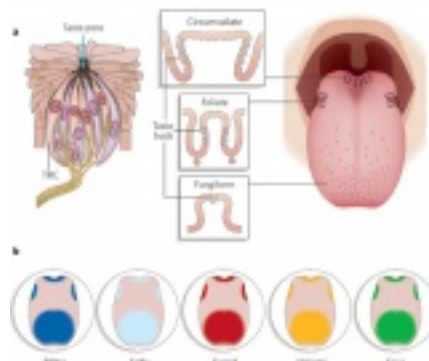
Hearing Development



Vision Development



Smell Development



Taste Development

We use the sense of balance and hearing to position ourselves in space, sense our surrounding environment, and to communicate. Portions of the ear appear very early in development as specialized region (otic placode) on the embryo surface that sinks into the mesenchyme to form a vesicle (otic vesicle = otocyst) that form the inner ear.

This region connects centrally to the nervous system and peripherally through specialized bones to the external ear (auricle). This organisation develops different sources forming the 3 ear parts: inner ear (otic placode, otocyst), middle ear (1st pharyngeal pouch and 1st and 2nd arch mesenchyme), and outer ear (1st pharyngeal cleft and 6 surface hillocks).

This complex origin, organisation, and timecourse means that abnormal development of any one system can impact upon the development of hearing.

Objectives

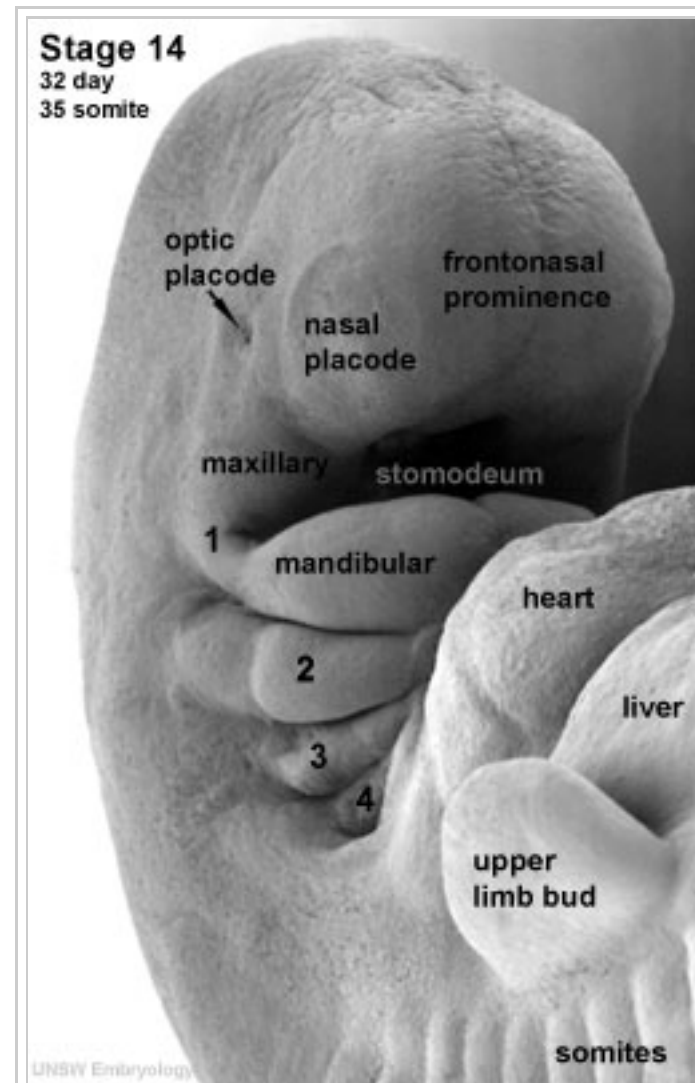
- Understanding of sensory placode development
- Understanding of inner, middle and external ear origins
- Understanding of timecourse of auditory development
- Understanding of abnormalities of auditory development
- Brief understanding of other sensory development

2015 Lecture 20 PDF

Also review your Head development lecture.

Lecture Resources




Movies and Virtual Slides[Expand]



Human embryo sensory placodes (Week 5, stage 14)



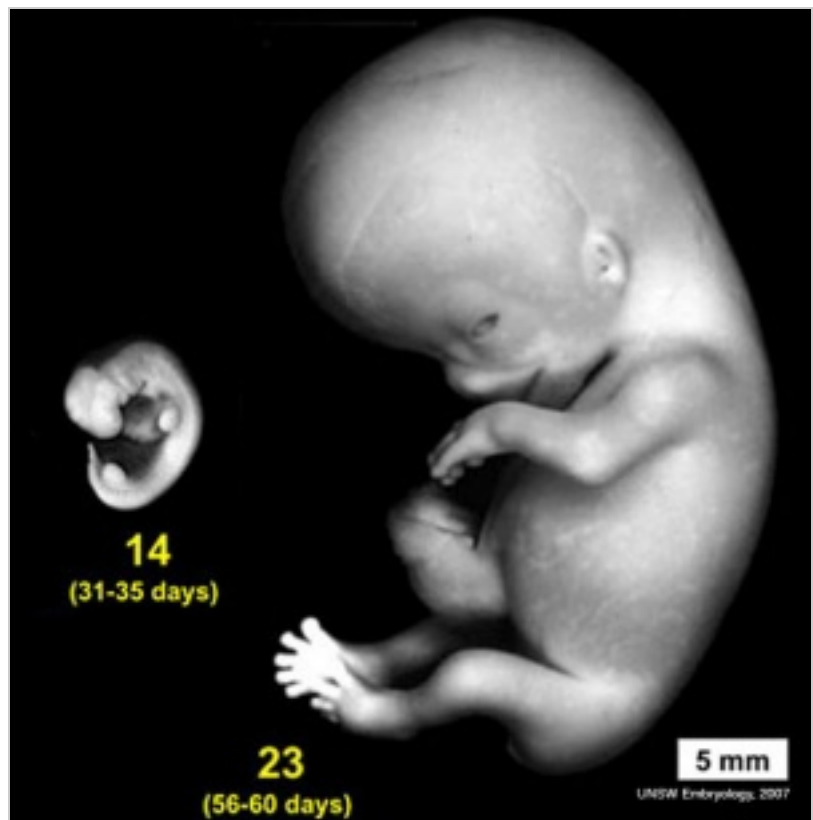
Adult hearing embryonic origins

References	[Collapse]
 <p>Hill, M.A. (2015). <i>UNSW Embryology</i> (15th ed.) Retrieved October 14, 2015, from https://embryology.med.unsw.edu.au</p>	<p>Senses Links: Introduction Placodes Hearing and Balance Vision Smell Taste Touch Stage 22 Category:Senses</p> <p>Hearing Links: Introduction Science Lecture Medicine Lecture Inner Ear Middle Ear Outer Ear Balance Hearing - Neural Pathway Stage 22 Abnormalities Neonatal Diagnosis - Hearing Hearing test Sensory Introduction Placodes Student project Category:Hearing</p> <p>Historic Embryology [Expand]</p> <p>Vision Links: Introduction Lens Retina Placodes Extraocular Muscle Cornea Eyelid Abnormalities Student project 1 Student project 2 Category:Vision</p> <p>Historic Embryology[Expand]</p> <p>Taste Links: Introduction Student project Tongue Development Category:Taste</p> <p>Archive: 2014 (https://embryology.med.unsw.edu.au/embryology/index.php?title=Lecture_-_Sensory_Development&oldid=149414) 2014 PDF</p>
 <p>Moore, K.L., Persaud, T.V.N. & Torchia, M.G. (2011). <i>The developing human: clinically oriented embryology</i> (9th ed.). Philadelphia: Saunders.</p>	<p>The following chapter links only work with a UNSW connection.</p> <ul style="list-style-type: none"> Chapter 18 – Development of Eyes and Ears (http://er.library.unsw.edu.au/er/cgi-bin/eraccess.cgi?url=http://www.mdconsult.com/books/page.do?eid=4-u1.0-B978-1-4377-2002-0..00018-7&isbn=978-1-4377-2002-0&uniqId=330028653-2#4-u1.0-B978-1-4377-2002-0..00018-7)
 <p>Schoenwolf, G.C., Bleyl, S.B., Brauer, P.R. & Francis-West, P.H. (2009). <i>Larsen's human embryology</i> (4th ed.). New York; Edinburgh: Churchill Livingstone.</p>	<p>The following chapter links only work with a UNSW connection.</p> <ul style="list-style-type: none"> Chapter 17 - Development of the Ears and Eyes (http://er.library.unsw.edu.au/er/cgi-bin/eraccess.cgi?url=http://er.library.unsw.edu.au/er/cgi-bin/eraccess.cgi?url=http://www.mdconsult.com/books/linkTo?type=bookPage&isbn=978-0-443-06811-9&eid=4-u1.0-B978-0-443-06811-9..10017-X)

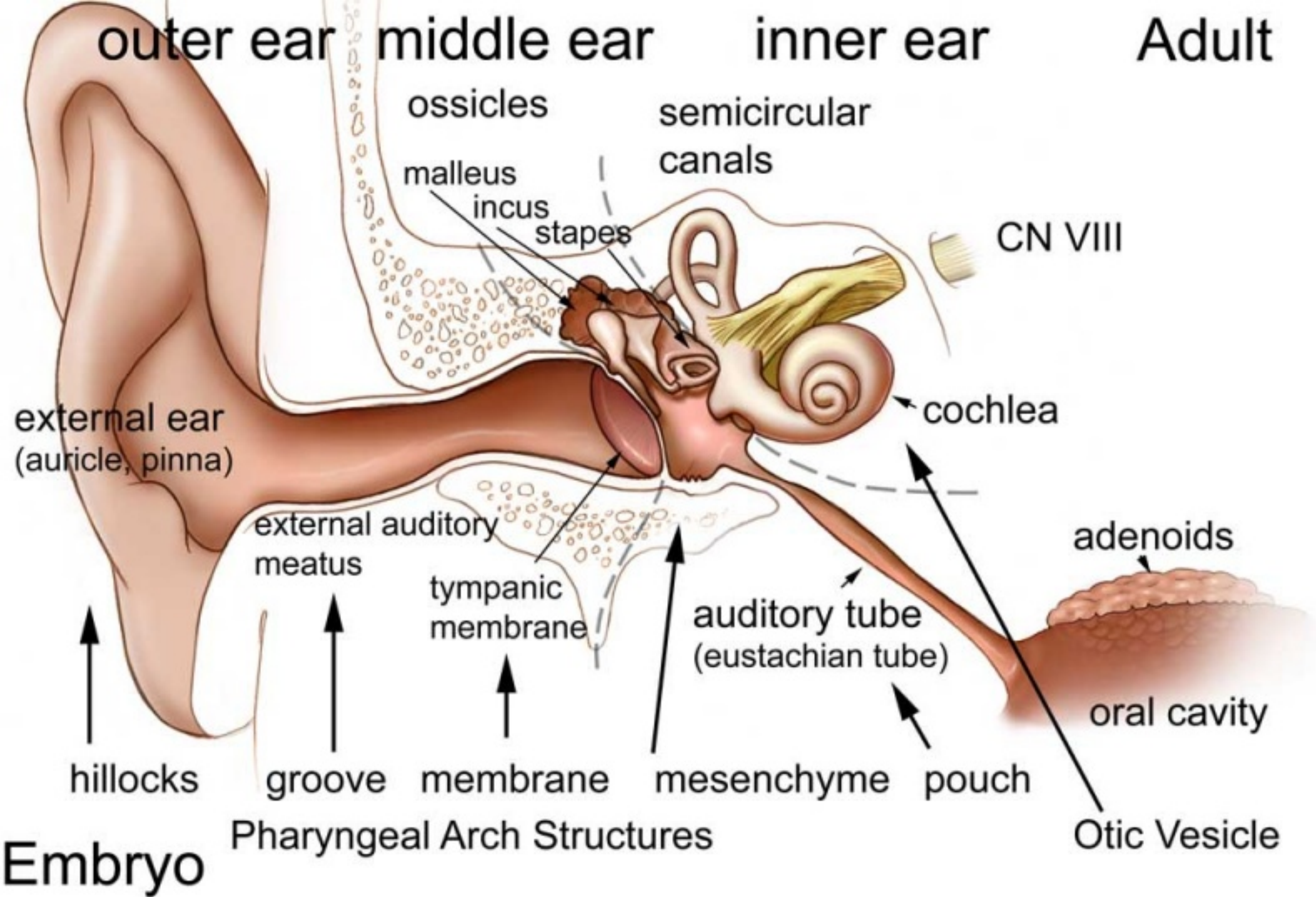
Development Timing

- Week 3** - otic placode, otic vesicle
- Week 5** - cochlear part of otic vesicle elongates (humans 2.5 turns)
- Week 9** - Mesenchyme surrounding membranous labryinth (otic capsule) chondrifies
- Week 12-16** - Capsule adjacent to membranous labryinth undegoes vacuolization to form a cavity (perilymphatic space) around membranous labrynth and fills with perilymph
- Week 16-24** - Centres of ossification appear in remaining cartilage of otic capsule form petrous portion of temporal bone. Continues to ossify to form mastoid process of temporal bone.
- 3rd Trimester** - Vibration acoustically of maternal abdominal wall induces startle response in fetus.

Embryonic Origin Overview



Comparison of size at stage 14 to 23



Adult hearing embryonic origins

Outer Ear

- Auricle - Pharyngeal Arches 1 and 2 (ectoderm, mesoderm)
- External Auditory Meatus - Pharyngeal Arch 1 groove or cleft (ectoderm)
- Tympanic Membrane - Pharyngeal Arch 1 membrane (ectoderm, mesoderm, endoderm)

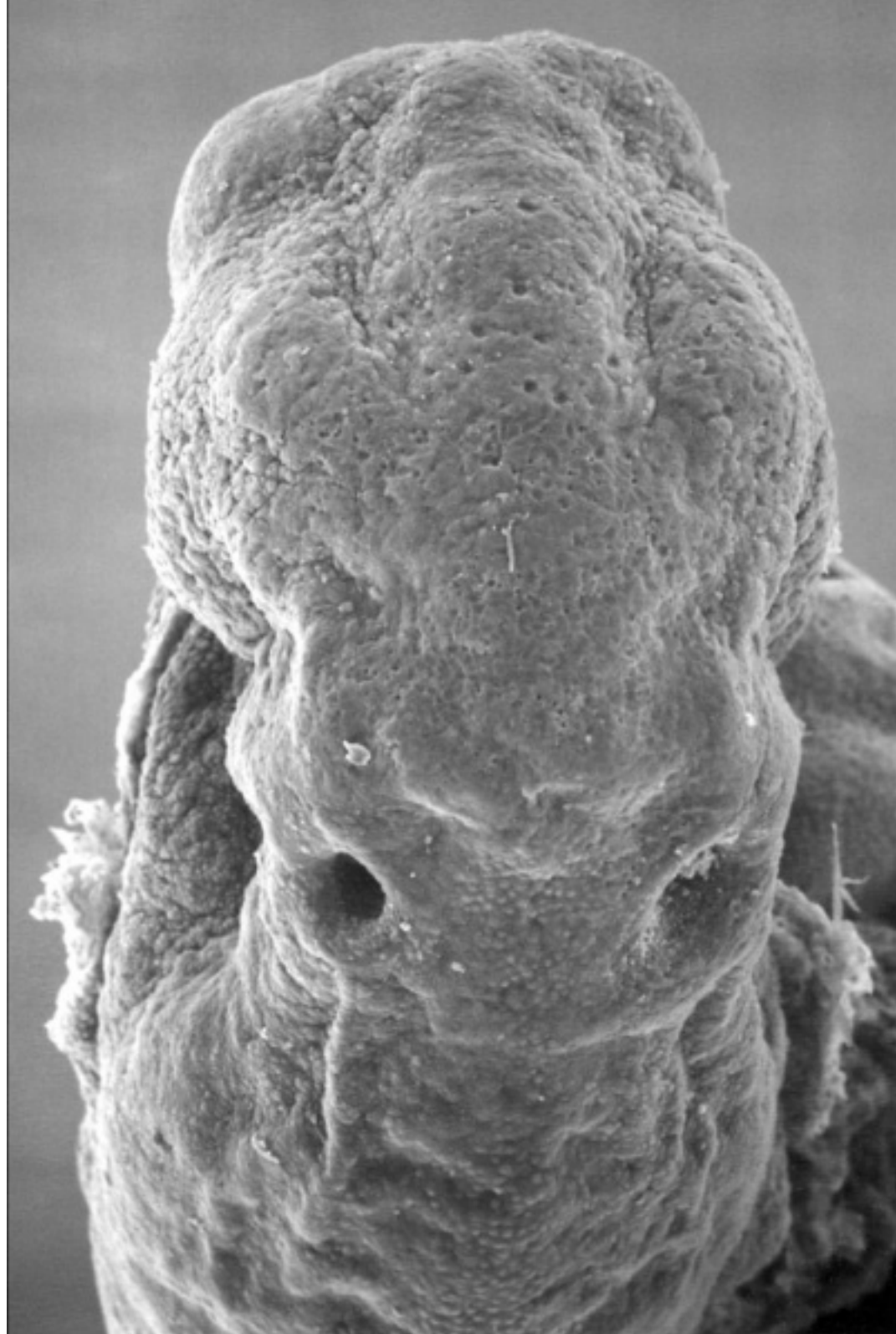
Middle Ear

- Middle Ear Ossicles
 - Malleus and incus - Pharyngeal Arch 1 cartilage Neural crest (ectoderm)
 - Stapes - Pharyngeal Arch 2 cartilage Neural crest (ectoderm)
- Middle Ear Muscles
 - Tensor tympani - Pharyngeal Arch 1 (mesoderm)
 - Stapedius - Pharyngeal Arch 2 (mesoderm)
- Middle ear cavity - Pharyngeal Arch 1 pouch (endoderm)

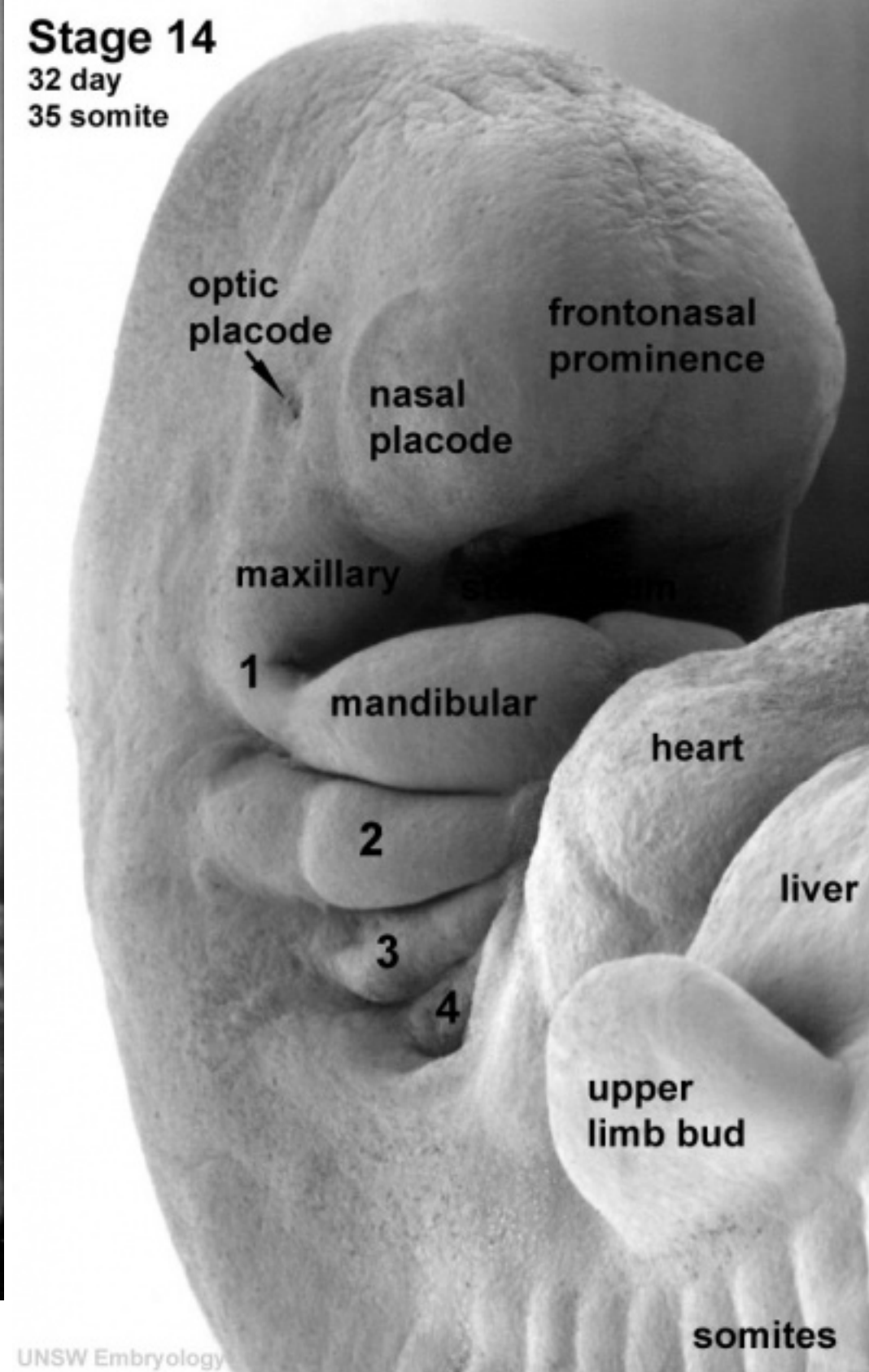
Inner Ear

- Inner Ear Labyrinth
 - Cochlea - Otic vesicle - Otic placode (ectoderm)
 - Semicircular canals - Otic vesicle - Otic placode (ectoderm)
 - Saccule and utricle - Otic vesicle - Otic placode (ectoderm)
- Cranial Nerve VIII
 - Auditory component - Otic vesicle and neural crest (ectoderm)
 - Vestibular component - Otic vesicle and neural crest (ectoderm)

Sensory Placodes



Otic placodes (Stage 11 dorsal view)



Sensory placodes ((Stage 14 ventral view)

- week 4 a series of thickened surface ectodermal patches form in pairs in the head region.
 - Recent research suggests that all sensory placodes may arise from common panplacodal primordium origin around the neural plate, and then differentiate to eventually have different developmental fates. PMID 20801420
- sensory placodes will later contribute key components of each of our special senses (vision, hearing and smell).
- Other species have a number of additional placodes which form other sensory structures (fish, lateral line receptor).
- Note that their initial postion on the developing head is significantly different to their final position in the future sensory system.

Otic Placode

- stage 13/14 embryo (shown below) the otic placode has sunk from the surface ectoderm to form a hollow epithelial ball, the otocyst, which now lies beneath the surface surrounded by mesenchyme (mesoderm).
- The epithelia of this ball varies in thickness and has begun to distort, it will eventually form the inner ear membranous labyrinth.

Lens Placode

- lies on the surface, adjacent to the outpocketing of the nervous system (which will for the retina) and will form the lens.

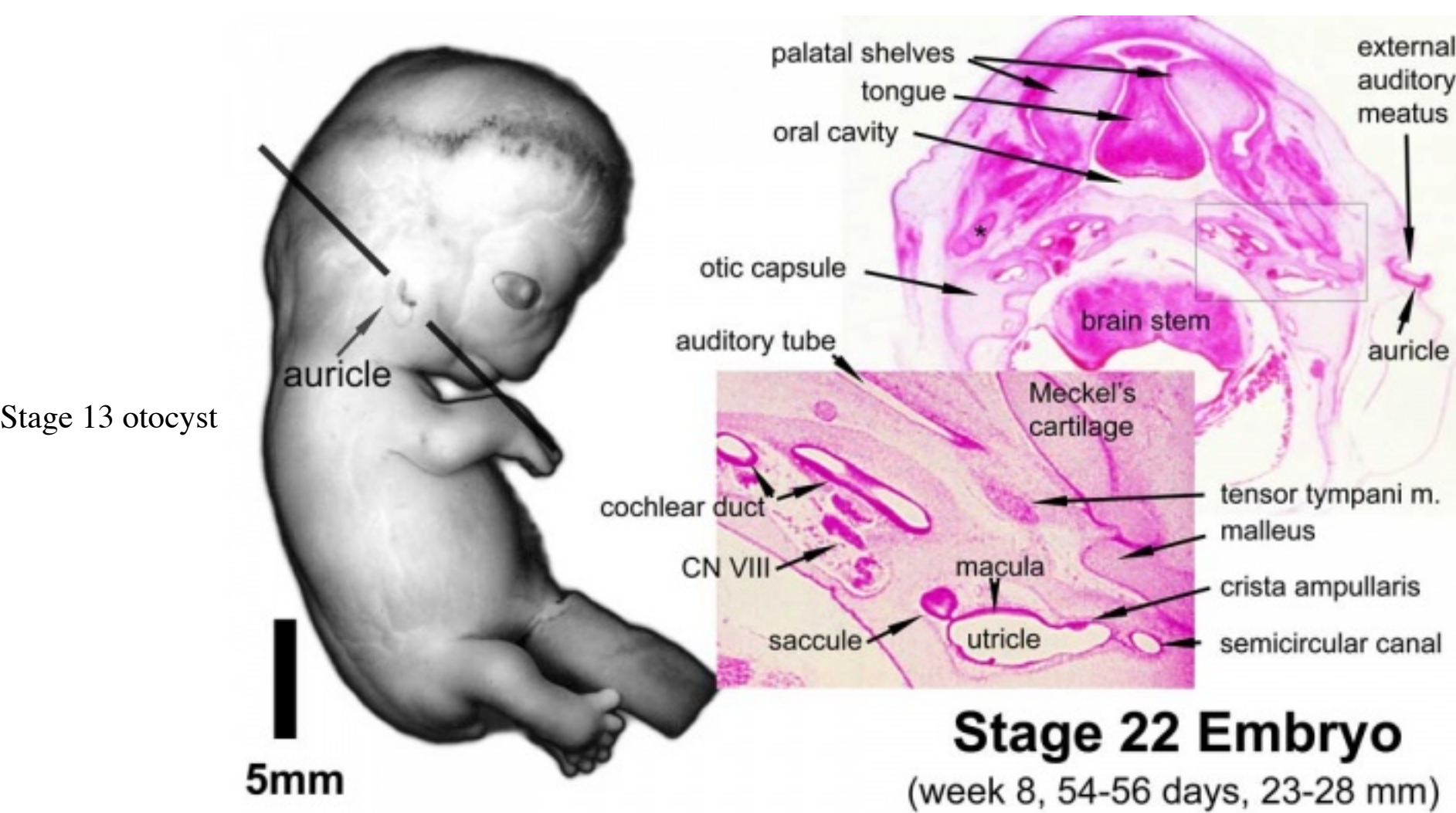
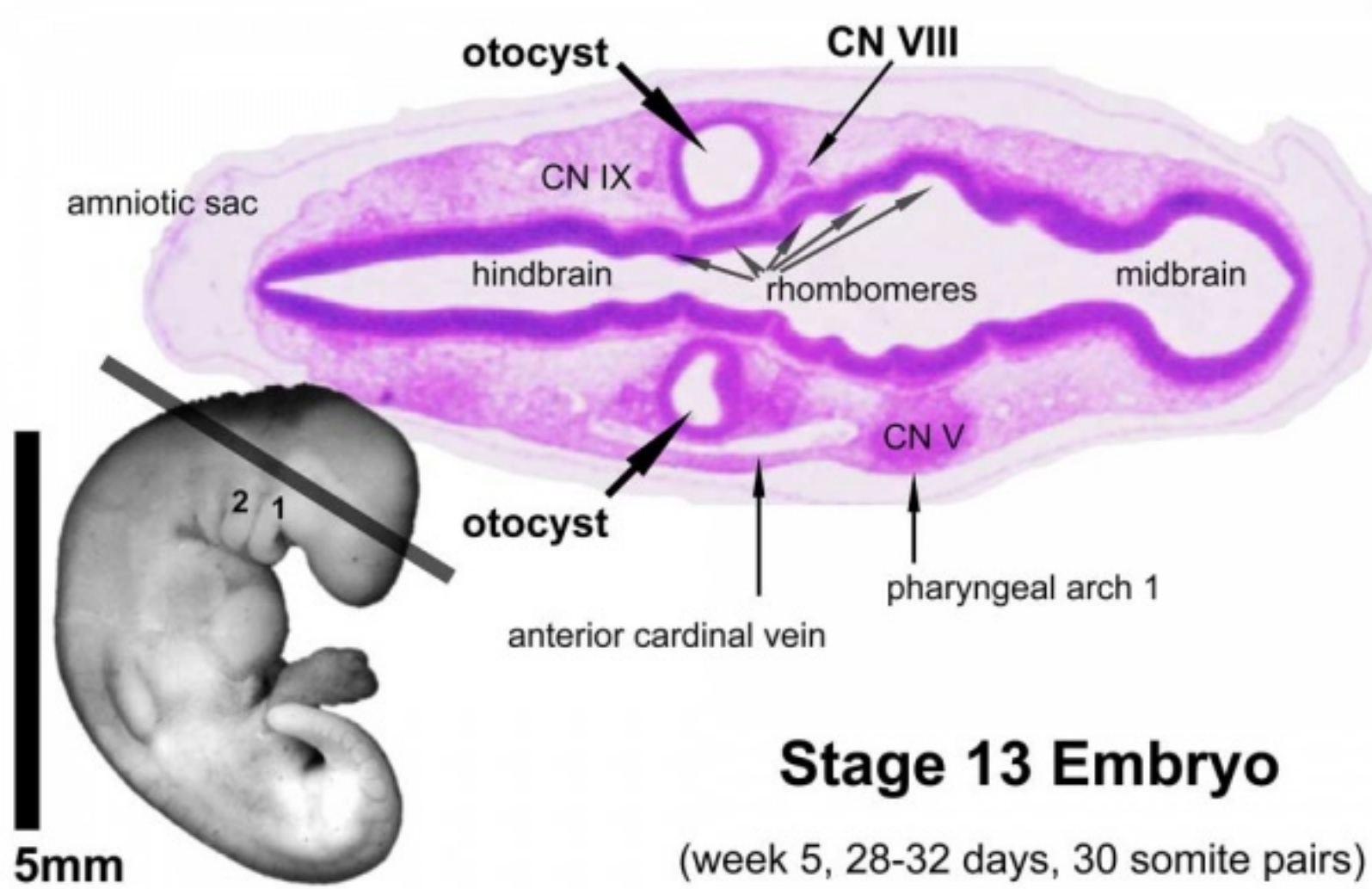
Nasal Placode

- 2 components (medial and lateral) and will form the nose olefactory epithelium.

Links: Placodes

Inner Ear

Described in detail later in lecture.



Stage 13 otocyst

Stage 22 ear

- The inner ear is derived from a pair of surface sensory placodes (otic placodes) in the head region.
- These placodes fold inwards forming a depression, then pinch off entirely from the surface forming a fluid-filled sac or vesicle (otic vesicle, otocyst).
- The vesicle sinks into the head mesenchyme some of which closely surrounds the otocyst forming the otic capsule.
- The otocyst finally lies close to the early developing hindbrain (rhombencephalon) and the developing vestibulo-cochlear-facial ganglion complex.

Links: Inner Ear | Neuroscience - The Inner Ear (<http://www.ncbi.nlm.nih.gov/books/bv.fcgi?rid=neurosci.section.894>)

Middle Ear

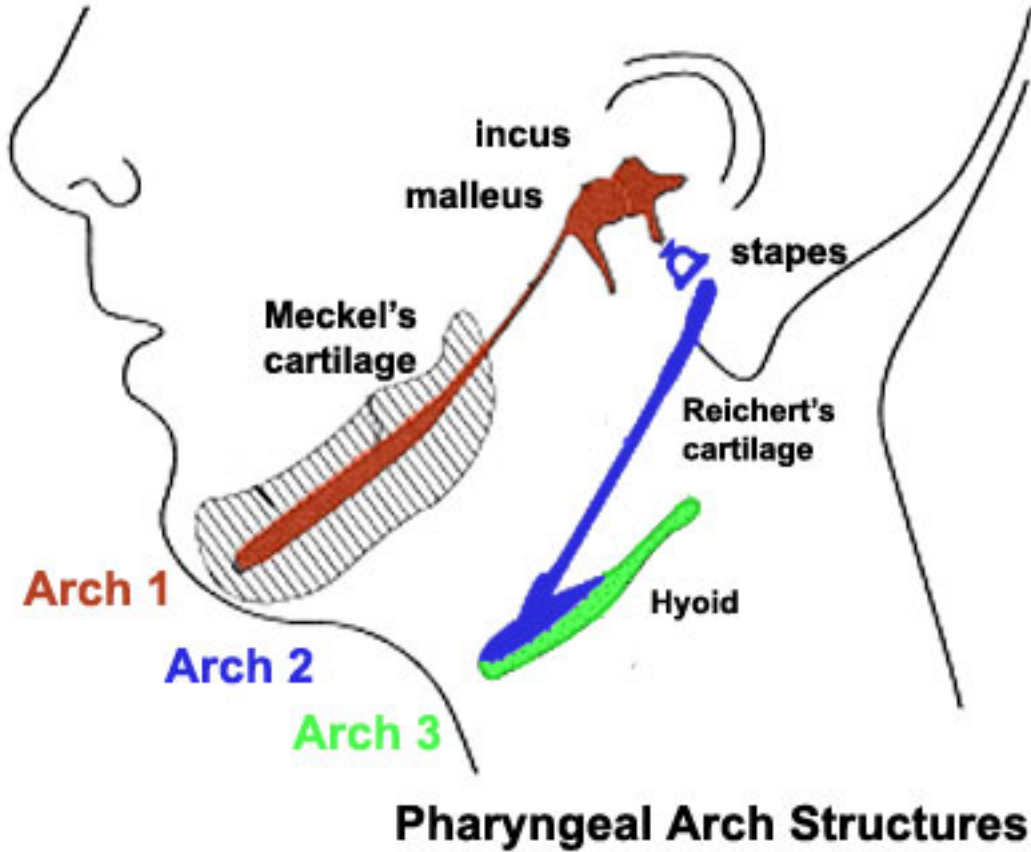
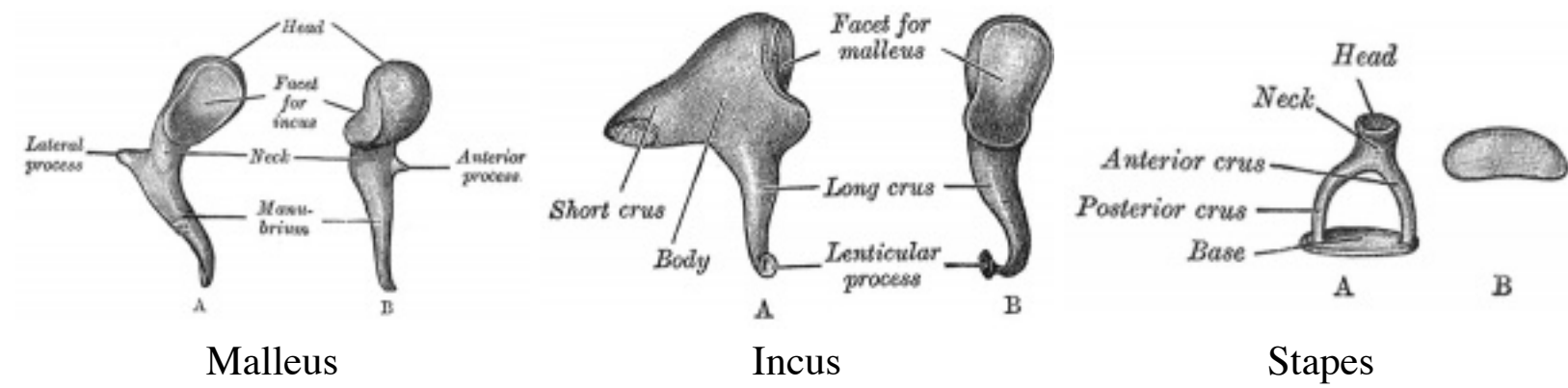
- derived from first pharyngeal pouch and 1st and 2nd arch mesenchyme
- extends as tubotympanic recess - during week 5 recess contacts outer ear canal
- mesoderm between 2 canals forms tympanic membrane
- expands to form tympanic recess
- stalk of recess forms auditory tube(eustachian tube, pharyngotympanic tube)

Ossicles

- The middle ear ossicles (bones) are derived from 1st and 2nd arch mesenchyme.
- The space in which these bones sit is derived from the 1st pharyngeal pouch.
 - remains connected to the oral cavity by the auditory tube.
- ossicles connected with tympanic cavity walls by ligaments (3 for the malleus, and 1

each for the incus and stapes).

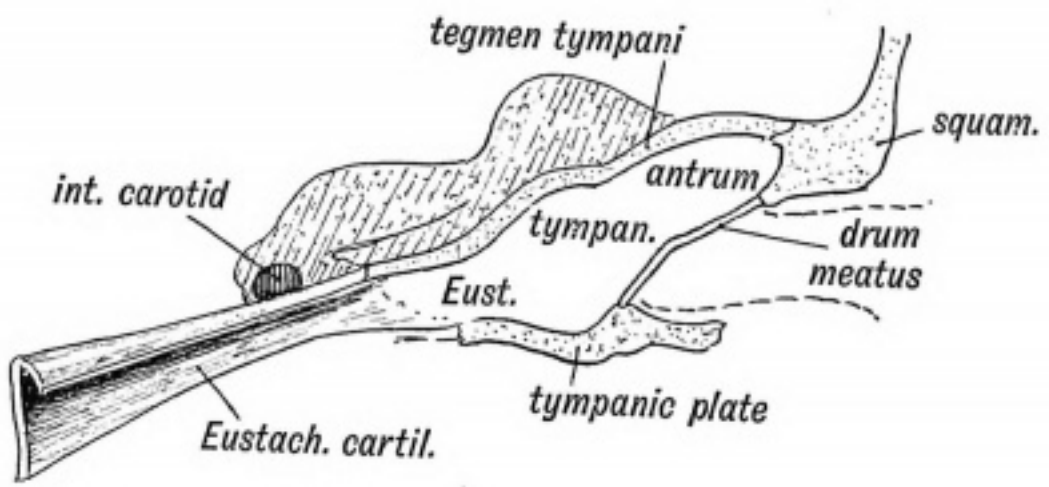
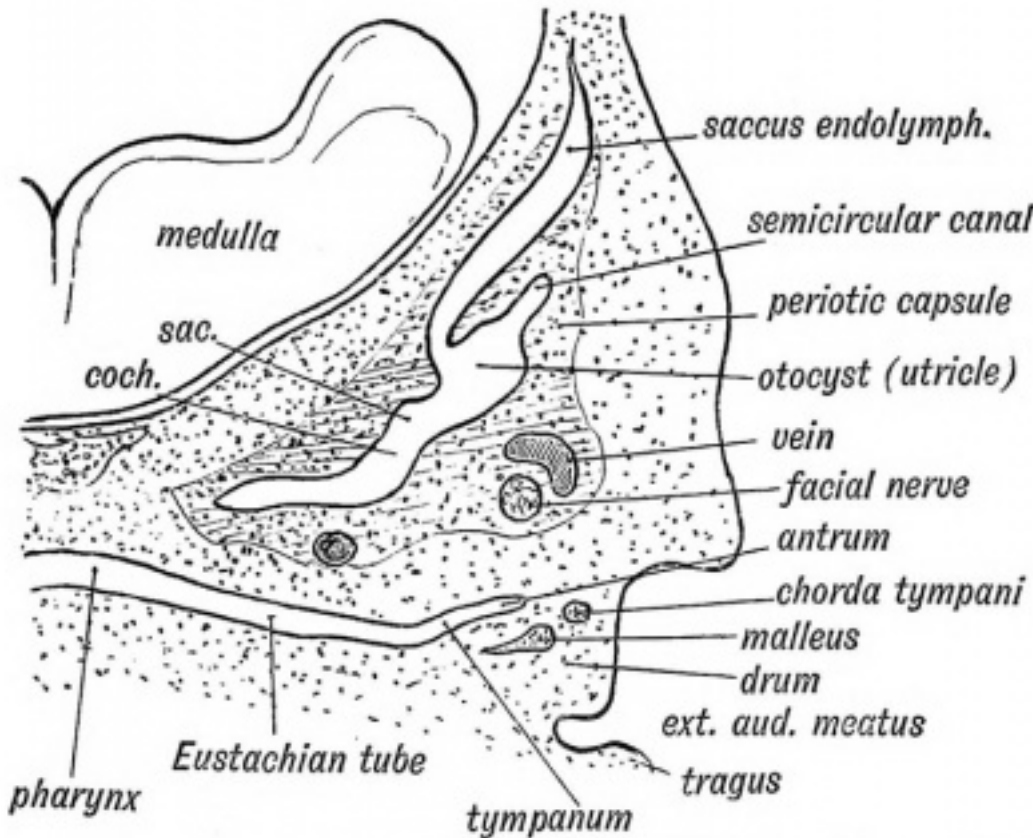
Adult Ossicles



Pharyngeal arch cartilages

Tympanic Cavity

- tympanic cavity enlarges to incorporate
- coats with epithelia
- first arch mesoderm - tensor tympani muscle
- second arch mesoderm - stapedius muscle



Cavities formed from the First Cleft

Middle Ear Genes - goosoid, RARs, Prx1, Otx2, Hoxa1, Hoxb1, endothelial related molecules

Links: Middle Ear | Neuroscience - The Middle Ear (<http://www.ncbi.nlm.nih.gov/books/bv.fcgi?rid=neurosci.section.893>)

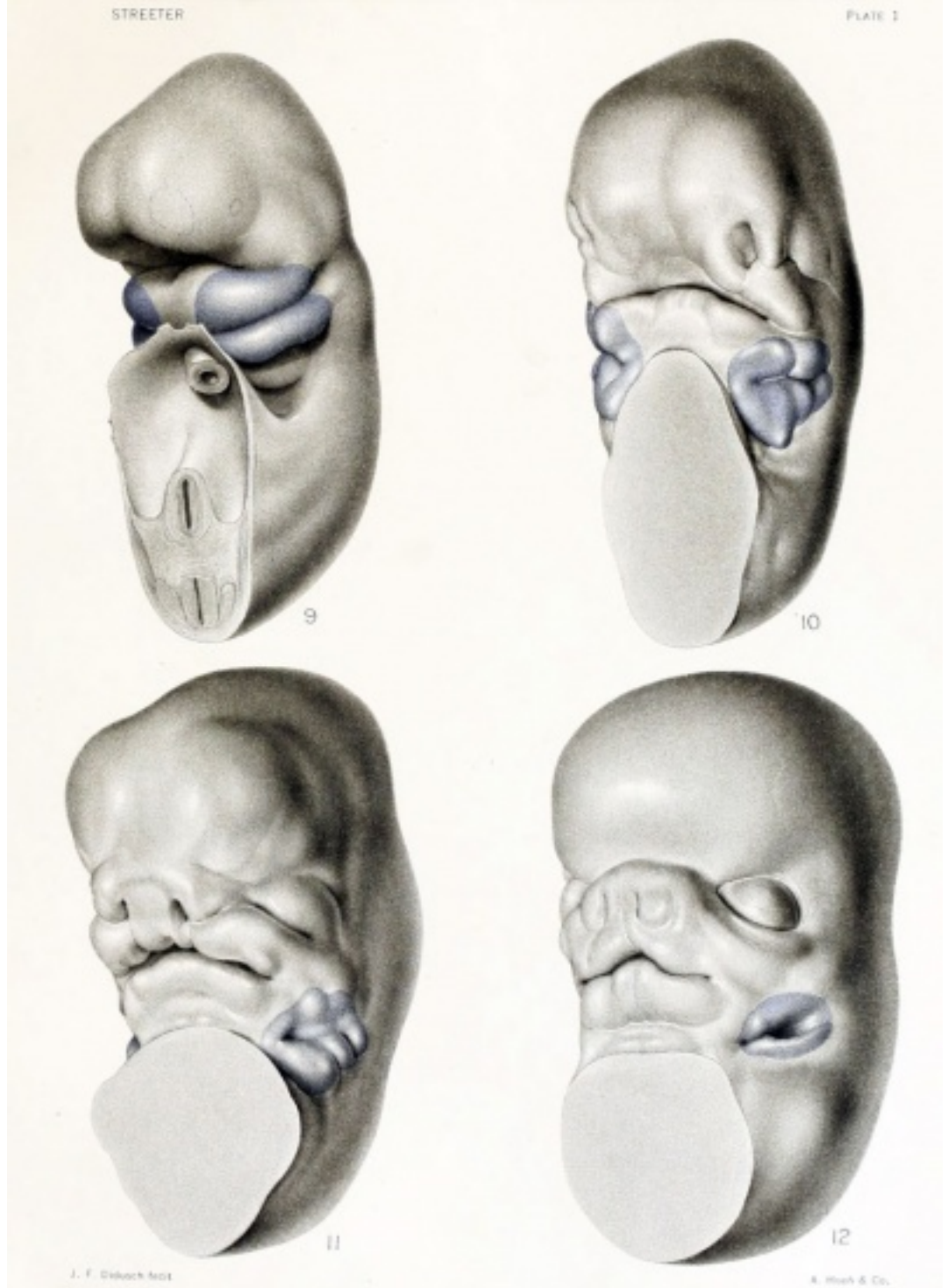
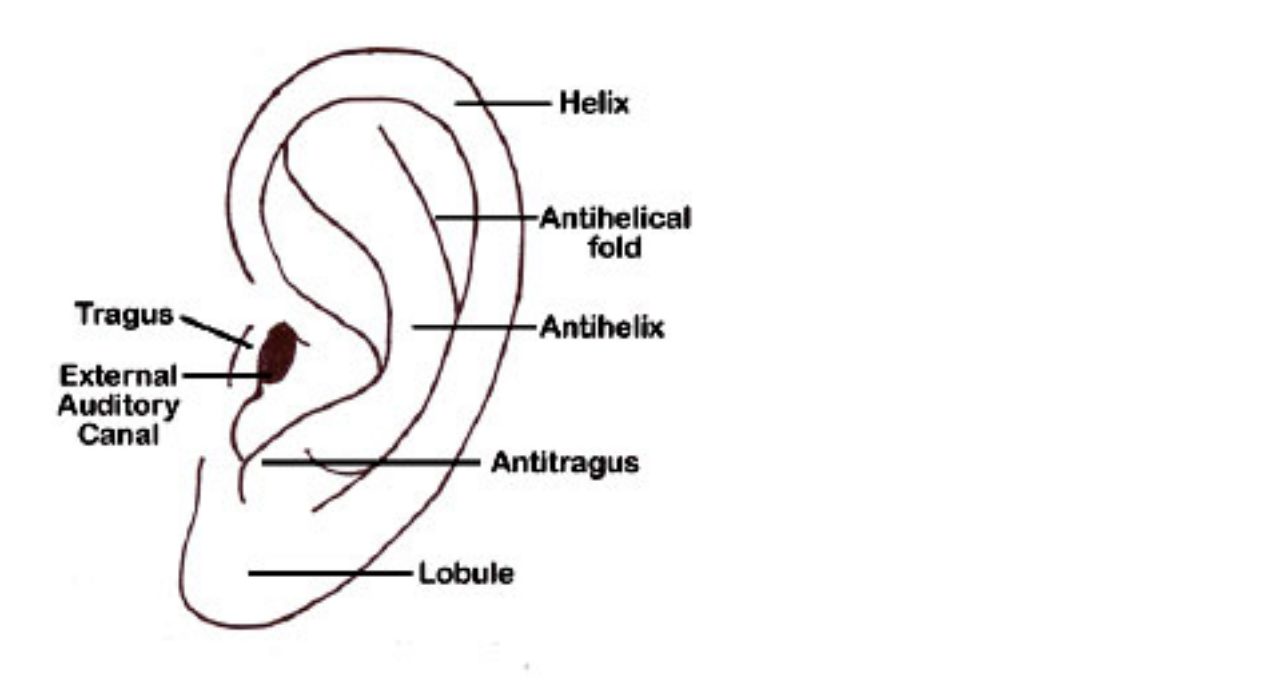
Outer Ear

- The **external ear** is derived from 6 surface hillocks, 3 on each of pharyngeal arch 1 and 2.
- The **external auditory meatus** is derived from the 1st pharyngeal cleft.
- The newborn external ear structure and position is an easily accessible diagnostic tool for potential abnormalities or further clinical screening.

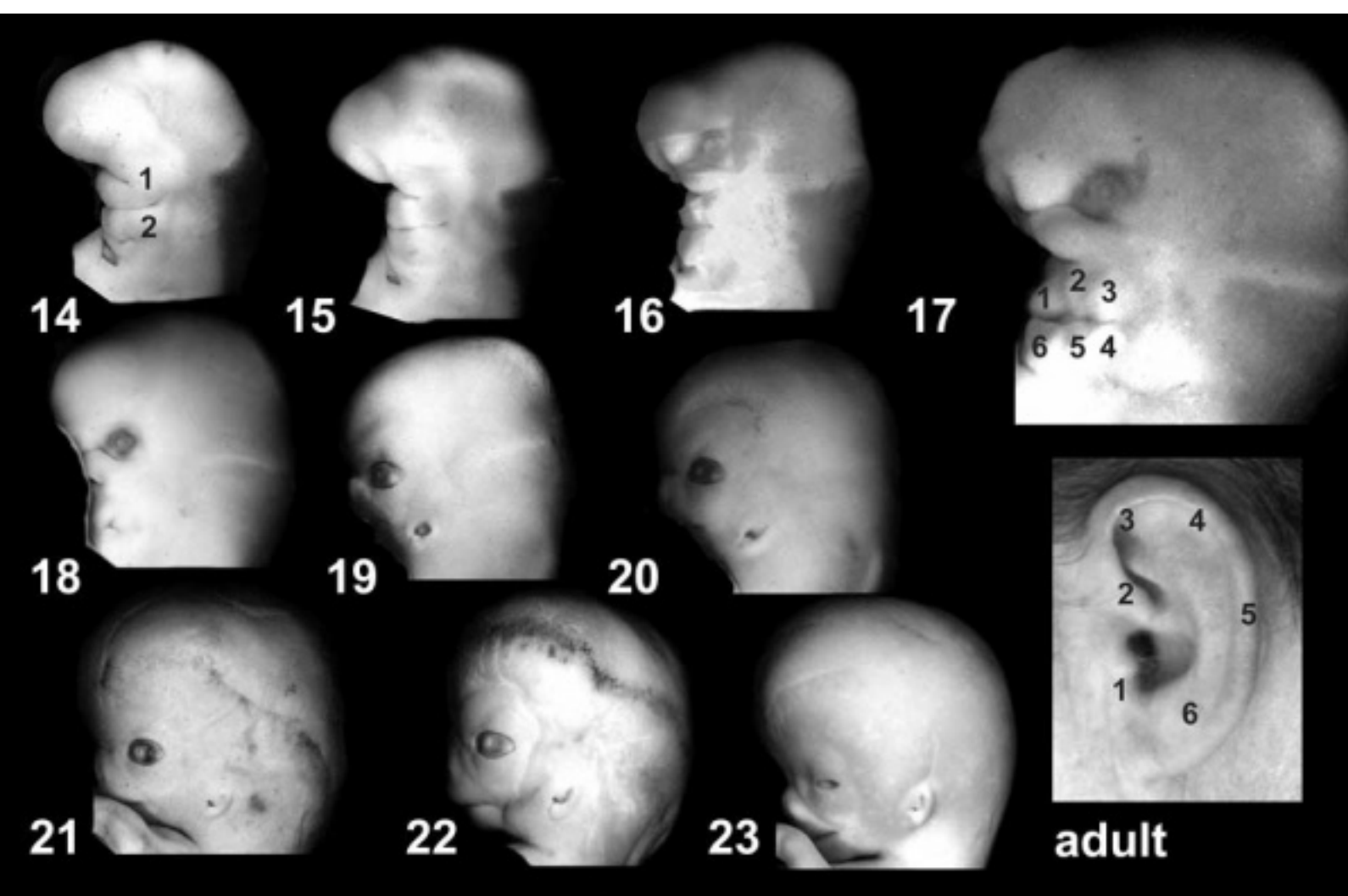
Pinna- Auricle

Develops from six aural hillocks

- 3 on first arch
- 3 on second arch
- originally on neck, moves cranially during mandible development



arch 1 and 2 hillocks



External ear stages 14-23 and adult (not to scale)

Pharyngeal Arch Hillock Auricle Component

Arch 1	1	tragus
	2	helix
	3	cymba concha
Arch 2	4	concha
	5	antihelix
	6	antitragus

- Outer- external auditory meatus
- derived from first pharyngeal cleft
- ectodermal diverticulum
- week 5 - extends inwards to pharynx
- until week 18 has ectodermal plug - plug forms stratified squamous epithelia of canal and outer eardrum

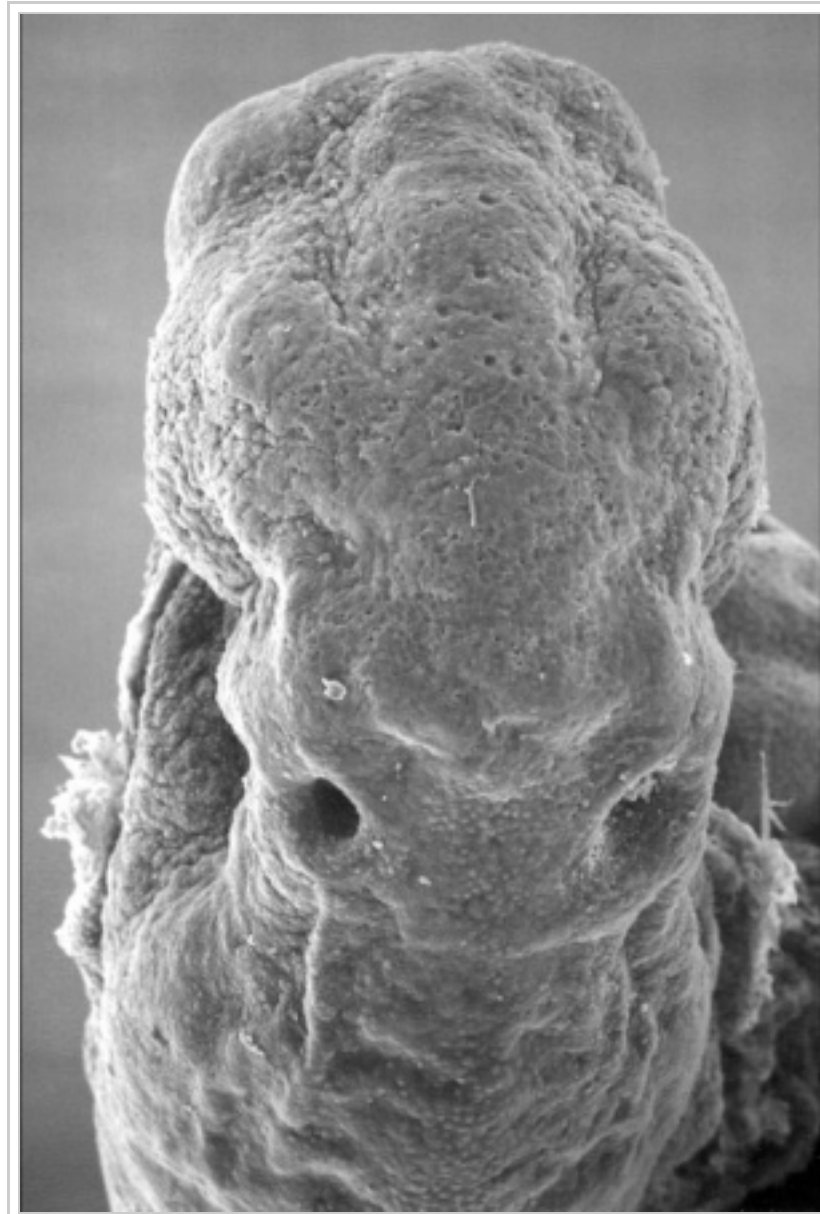
Timeline

-
- This anatomical diagram illustrates the internal and external structures of the human ear. The diagram is a sagittal section showing the following labeled parts:
- Auditory tube**: The tube connecting the middle ear to the nasopharynx.
 - Cowl of mandible**: The bony part of the jawbone.
 - Internal carotid artery**: The major blood vessel carrying blood from the heart to the head.
 - Internal acoustic meatus**: The opening of the internal ear canal.
 - Tympanic cavity**: The middle ear space containing the ossicles.
 - Tympanic membrane**: The eardrum that separates the middle ear from the external ear.
 - Mastoid air-cells**: Small air spaces in the mastoid process of the temporal bone.
 - Transverse sinus**: A large vein in the dural venous sinuses.
 - Part of parotid gland**: The salivary gland located in front of the ear.
 - Tragus**: The small cartilaginous point in front of the ear.
 - External acoustic meatus**: The ear canal leading to the eardrum.
 - Helix**: The outer rim of the ear.

outer ear and external auditory meatus

Inner

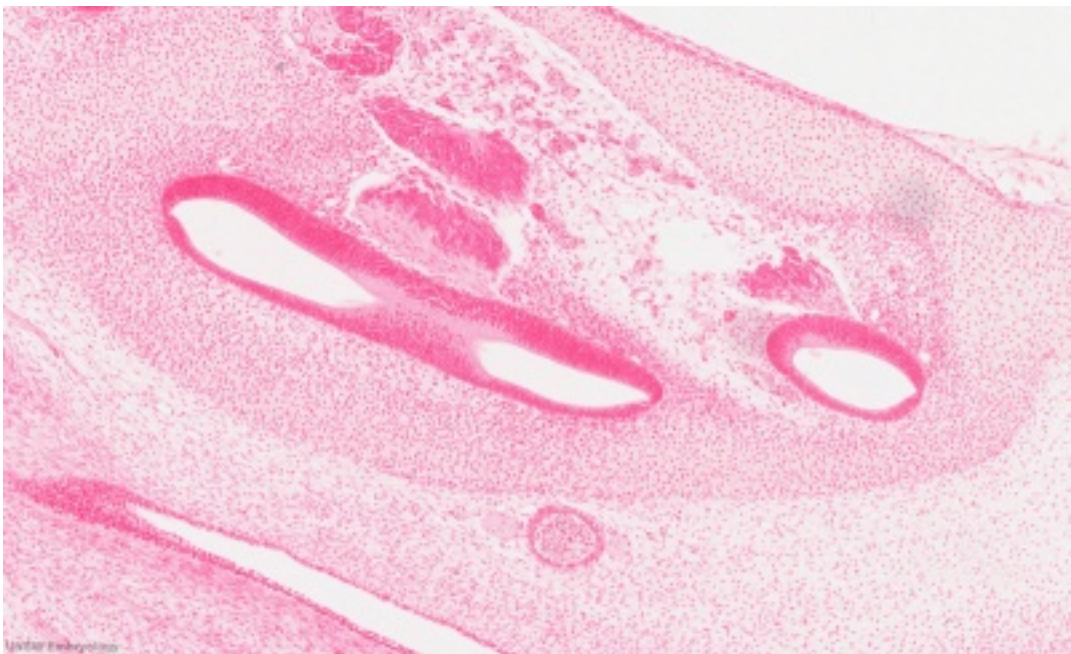
Otocyst



Otic placodes (Stage 11 dorsal view)

- ## Vestibular Sac

- ## Cochlear sac



- ## Bony Labyrinth

- formed from chondrified mesoderm

- Periotic Capsule
- mesenchyme within capsule degenerates to form space filled with perilymph

Vestibulocochlear Nerve

- forms beside otocyst
- from wall of otocyst and neural crest cells
- bipolar neurons
- vestibular neurons
 - outer end of internal acoustic meatus
 - innervate hair cells in membranous labyrinth
 - axons project to brain stem and synapse in vestibular nucleus
- cochlear neurons
 - cell bodies lie in modiolus
 - central pillar of cochlear
 - innervate hair cells of spiral organ
 - axons project to cochlear nucleus

Inner Ear Genes

- hindbrain segmentation occurs at same time placode arises
- otocyst adjacent to rhombomere 5
- may influence development
- Hoxa1, kreisler, Fgf3
- genes regulating neural crest cells (neural genes)
- Pax2 Ko affects cochlear and spiral ganglion, but not vestibular apparatus
- nerogenin 1 affects both ganglia

Semicircular canal

- Otx1- cochlear and vestibular normal
- Hmx3, Prx1, Prx2

Sensory Organs

- thyroid hormone receptor beta
- Zebrafish-mindbomb mutant has excess hair cells but not supporting cells, Notch-Delta signaling
- Gene Expression-inner ear
- Brn-3c and Hair cell development
- Supporting Cells- p27kip
- Thyroid Hormone
- Ganglion neurons require growth factors
- vestibular neurons- BDNF, NT3
 - survival not development

Postnatal Changes

Newborn to adult Eustachian (auditory, otopharyngeal or pharyngotympanic) tube.

- Connects middle ear cavity to nasopharynx portion of pharynx

Functions

- Ventilation - pressure equalization in the middle ear
- Clearance - allow fluid drainage from the middle ear Tube is normally closed and opened by muscles

At birth

- shorter (17-18 mm), narrower and runs almost horizontal Tube is opened by a single muscle, tensor palati muscle

Adult

- longer (twice as long), wider and runs at approximately 45 degrees to the horizontal. Tube is opened by two separate muscles, tensor palati and levator palati

Vision

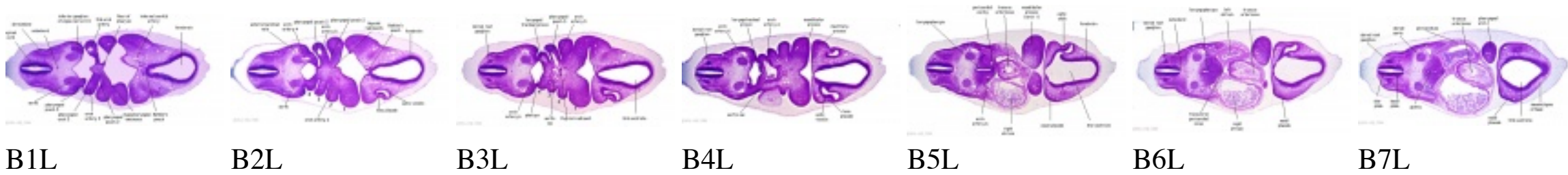
Timeline

- **Weeks 3-4** - Eye Fields-Optic Vesicle
- **Weeks 5-6** - Optic Cup, Lens Vesicle, Choroid Fissure, Hyaloid Artery
- **Weeks 7-8** - Cornea, Anterior Chamber, Pupillary Membrane, Lens, Retina
- **Weeks 9-15** - Iris, Ciliary Body
- **Weeks 8-10** - Eyelids



Carnegie Stage 12 otic placode

Stage 13 (week 5)



Lens

Surface ectoderm -> lens placode (optic placode) -> lens pit -> lens vesicle -> lens fibres -> lens capsule and embryonic/fetal nucleus.

Retina

Neural plate ectoderm -> prosencephalon (forebrain) eye fields -> neural plate growth carries eye field region forward -> eye field invaginates forming optic grooves (sulci) -> diencephalon optic groove interacts with surface ectoderm (induces optic placode) -> optic stalk -> optic vesicle -> folds inward (optic cup) forming double layer -> inner neural retina, outer pigmented retina

Links: Embryo Images - Eye Development (http://www.med.unc.edu/embryo_images/unit-eye/eye_htms/eyetoc.htm)

Neural Crest

Eye connective tissue

Abnormalities

- Inner - common cavity, severe cochlear hypoplasia
 - Large vestibular aqueduct syndrome (LVAS) can be one of the common causes of hearing loss
- Middle - rare and can be part of first arch syndrome, Malleus, Incus and Stapes Fixation
 - Cholesteatoma- Epithelium trapped within skull base in development, erosion of bones: temporal bone, middle ear, mastoid
- Outer - Several genetic effects and syndromes, Environmental Effects

Outer Ear Abnormalities

- Microtia - abnormally small external ear
- Preauricular sinus - occurs in 0.25% births, bilateral (hereditary) 25-50%, unilateral (mainly the left), duct runs inward can extend into the parotid gland, Postnatally sites for infection

Fetal Alcohol Syndrome

- Postion- Lower or uneven height, "railroad track" appearance, curve at top part of outer ear is under-developed, folded over parallel to curve beneath

Congenital Deafness

Sensorineural - cochlear or central auditory pathway

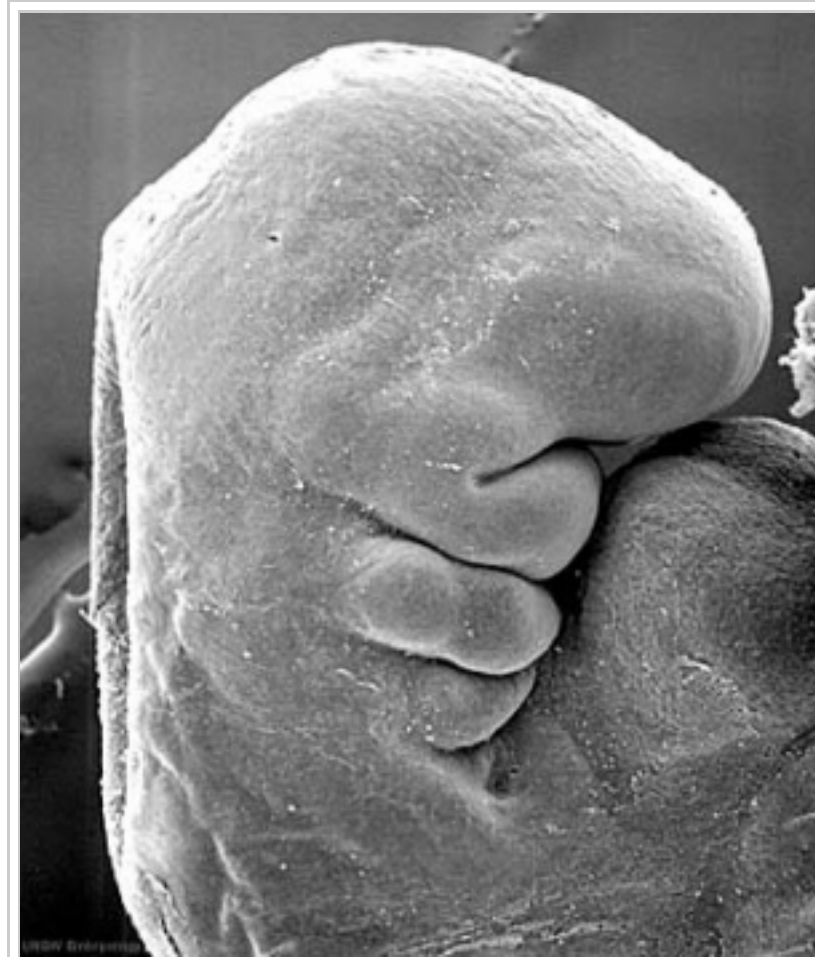
- Hereditary
 - recessive- severe
 - dominant- mild
 - can be associated with abnormal pigmentation (hair and irises)
- Acquired
 - rubella (German measles), maternal infection during 2nd month of pregnancy, vaccination of young girls
 - streptomycin
 - antibiotic
 - thalidomide

Conductive - disease of outer and middle ear

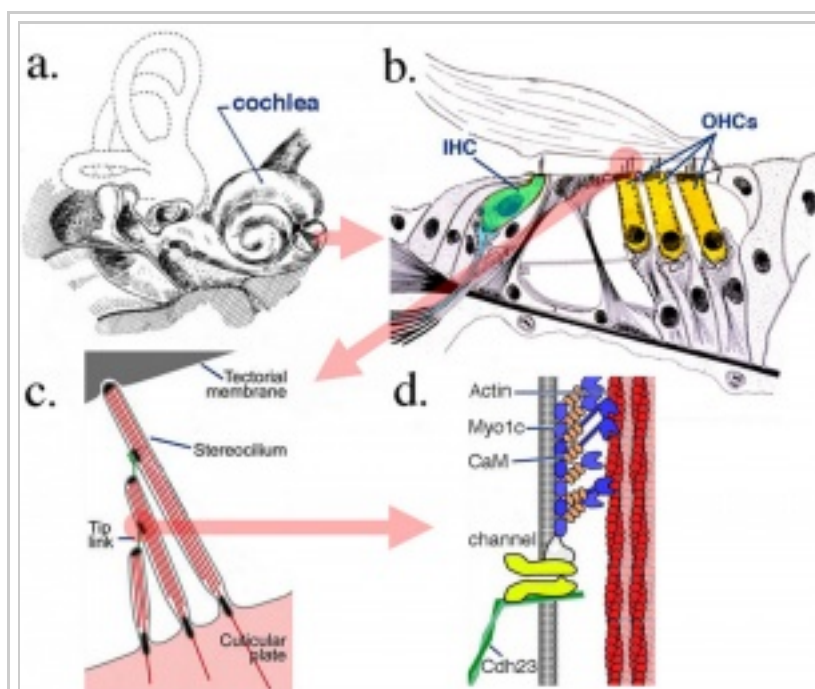
- produced by otitis media with effusion, is widespread in young children.
- temporary blockage of outer or middle ear

Bionic Ear

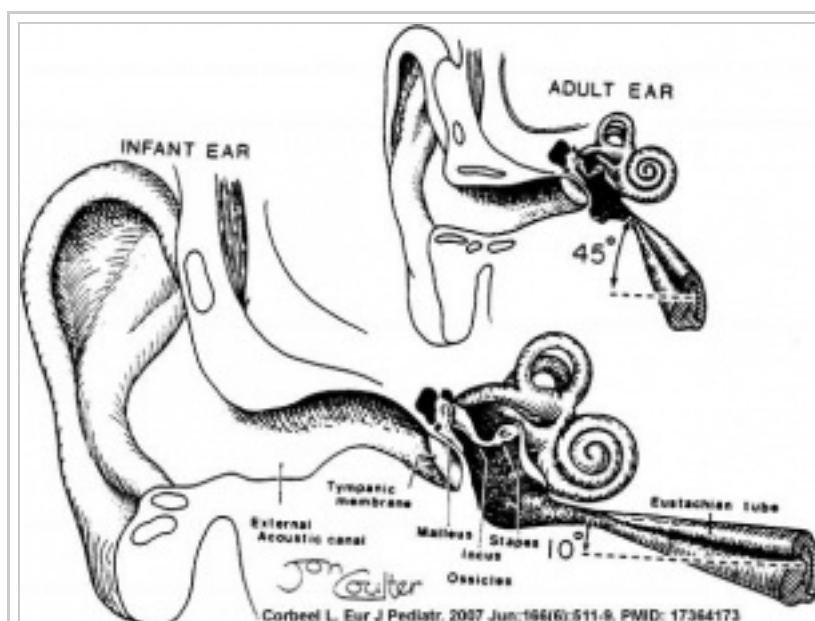
Cochlear Implant - Professor Graeme Clark (1960s, Australia) Array of electrodes implanted within cochlea, direct electrical stimulation to auditory nerve fibres



Carnegie Stage 13 otic vesicle



Inner ear hair cells



Eustacian tube angle changes

Conductive Hearing Loss

- **Conductive Hearing Loss Produces a Reversible Binaural Hearing Impairment** David R. Moore, Jemma E. Hine, Ze Dong Jiang, Hiroaki Matsuda, Carl H. Parsons, and Andrew J. King J. Neurosci. 1999;19 8704-8711 <http://www.jneurosci.org/cgi/content/abstract/19/19/8704>
 - tested ferrets by lon-term plugging of ear canal
 - Repeated testing during the 22 months after unplugging revealed a gradual return to normal levels of unmasking.
 - Results show that a unilateral conductive hearing loss, in either infancy or adulthood, impairs binaural hearing both during and after the hearing loss.
 - Show scant evidence for adaptation to the plug and demonstrate a recovery from the impairment that occurs over a period of several months after restoration of normal peripheral function.

References

Textbooks

- **Before We Are Born** (5th ed.) Moore and Persaud Chapter 20: p460-479
- **Essentials of Human Embryology**, Larson Chapter 12: p252-272

Online Textbooks

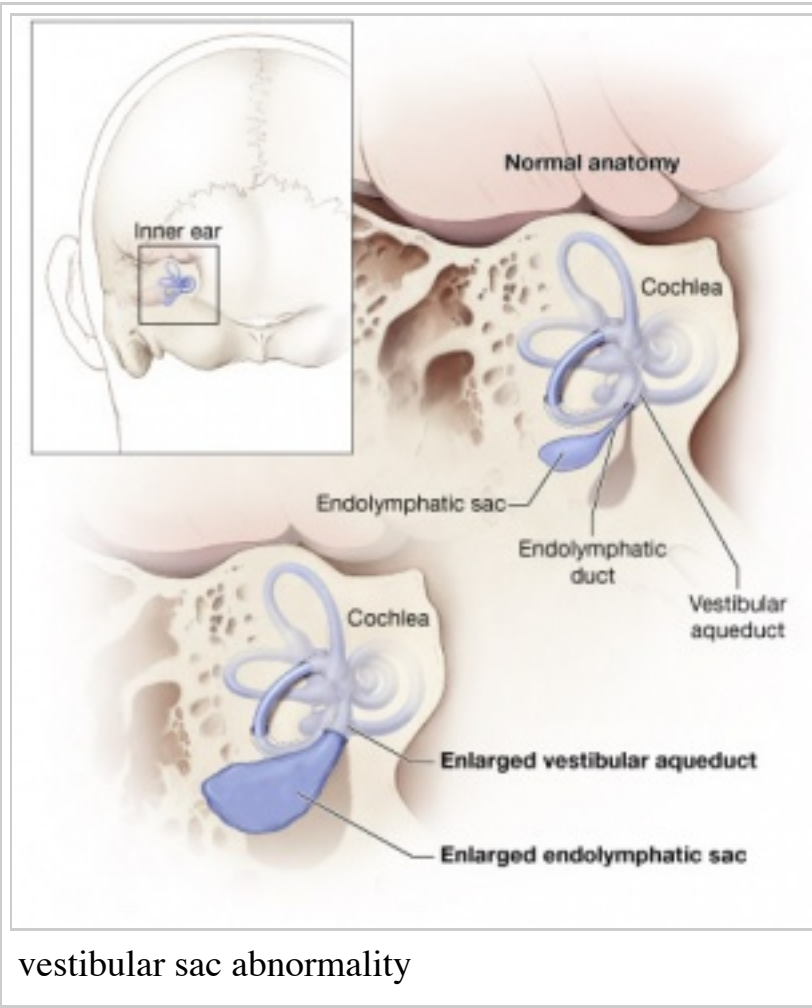
- **Developmental Biology** (6th ed.) Gilbert, Scott F. Sunderland (MA): Sinauer Associates, Inc.; c2000. Evolution of the mammalian middle ear bones from the reptilian jaw (<http://www.ncbi.nlm.nih.gov/books/bv.fcgi?rid=dbio.figgrp.5455%20>) | Chick embryo rhombomere neural crest cells (<http://www.ncbi.nlm.nih.gov/books/bv.fcgi?rid=dbio.figgrp.5460>) | Some derivatives of the pharyngeal arches (<http://www.ncbi.nlm.nih.gov/books/bv.fcgi?rid=dbio.table.3135>) | Formation of the Neural Tube (<http://www.ncbi.nlm.nih.gov/books/bv.fcgi?call=bv.View..ShowSection&rid=dbio.section.2871>) | Differentiation of the Neural Tube (<http://www.ncbi.nlm.nih.gov/books/bv.fcgi?call=bv.View..ShowSection&rid=dbio.section.2884>) | Tissue Architecture of the Central Nervous System (<http://www.ncbi.nlm.nih.gov/books/bv.fcgi?call=bv.View..ShowSection&rid=dbio.section.2894>) | Neuronal Types (<http://www.ncbi.nlm.nih.gov/books/bv.fcgi?call=bv.View..ShowSection&rid=dbio.section.2908>) | Snapshot Summary: Central Nervous System and Epidermis (<http://www.ncbi.nlm.nih.gov/books/bv.fcgi?call=bv.View..ShowSection&rid=dbio.section.2937>)
- **Neuroscience** Purves, Dale; Augustine, George J.; Fitzpatrick, David; Katz, Lawrence C.; LaMantia, Anthony-Samuel; McNamara, James O.; Williams, S. Mark. Sunderland (MA): Sinauer Associates, Inc. ; c2001 The Auditory System (<http://www.ncbi.nlm.nih.gov/books/bv.fcgi?rid=neurosci.chapter.879>) | The Inner Ear (<http://www.ncbi.nlm.nih.gov/books/bv.fcgi?rid=neurosci.section.894>) | The Middle Ear (<http://www.ncbi.nlm.nih.gov/books/bv.fcgi?rid=neurosci.section.893>) | The External Ear (<http://www.ncbi.nlm.nih.gov/books/bv.fcgi?rid=neurosci.section.891>) | Early Brain Development (<http://www.ncbi.nlm.nih.gov/books/bv.fcgi?rid=neurosci.chapter.1447>) | Construction of Neural Circuits (<http://www.ncbi.nlm.nih.gov/books/bv.fcgi?rid=neurosci.chapter.1546>) | Modification of Brain Circuits as a Result of Experience (<http://www.ncbi.nlm.nih.gov/books/bv.fcgi?rid=neurosci.chapter.1640>)
- **Molecular Biology of the Cell** (4th Edn) Alberts, Bruce; Johnson, Alexander; Lewis, Julian; Raff, Martin; Roberts, Keith; Walter, Peter. New York: Garland Publishing; 2002. Neural Development (<http://www.ncbi.nlm.nih.gov/80/books/bv.fcgi?db=Books&rid=mboc4.section.3963>) | The three phases of neural development (<http://www.ncbi.nlm.nih.gov/80/books/bv.fcgi?db=Books&rid=mboc4.figgrp.3966>)
- **Clinical Methods** 63. Cranial Nerves IX and X: The Glossopharyngeal and Vagus Nerves (<http://www.ncbi.nlm.nih.gov/books/bv.fcgi?rid=cm.chapter.1949>) | The Tongue (<http://www.ncbi.nlm.nih.gov/books/bv.fcgi?rid=cm.chapter.3847>) | 126. The Ear and Auditory System (<http://www.ncbi.nlm.nih.gov/books/bv.fcgi?rid=cm.chapter.3777>) | An Overview of the Head and Neck - Ears and Hearing (<http://www.ncbi.nlm.nih.gov/books/bv.fcgi?rid=cm.chapter.3627#3654>) | Audiometry (<http://www.ncbi.nlm.nih.gov/books/bv.fcgi?rid=cm.chapter.3897>)
- **Health Services/Technology Assessment Text (HSTAT)** Bethesda (MD): National Library of Medicine (US), 2003 Oct. Developmental Disorders Associated with Failure to Thrive (<http://www.ncbi.nlm.nih.gov/80/books/bv.fcgi?db=Books&rid=hstat1a.section.25014#25029>)
- **Eurekah Bioscience Collection** Cranial Neural Crest and Development of the Head Skeleton (<http://www.ncbi.nlm.nih.gov/books/bv.fcgi?rid=eurekah.chapter.53006>)

Search

- **Bookshelf** hearing development (<http://www.ncbi.nlm.nih.gov/sites/entrez?db=Books&cmd=search&term=hearing+development>)
- **Pubmed** hearing development (<http://www.ncbi.nlm.nih.gov/sites/gquery?itool=toolbar&cmd=search&term=hearing+development>)

External Links

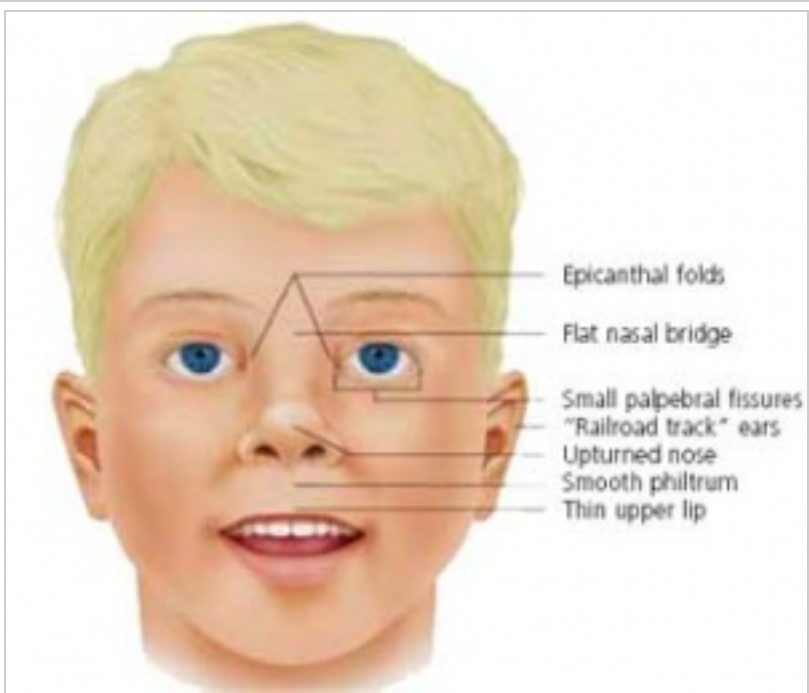
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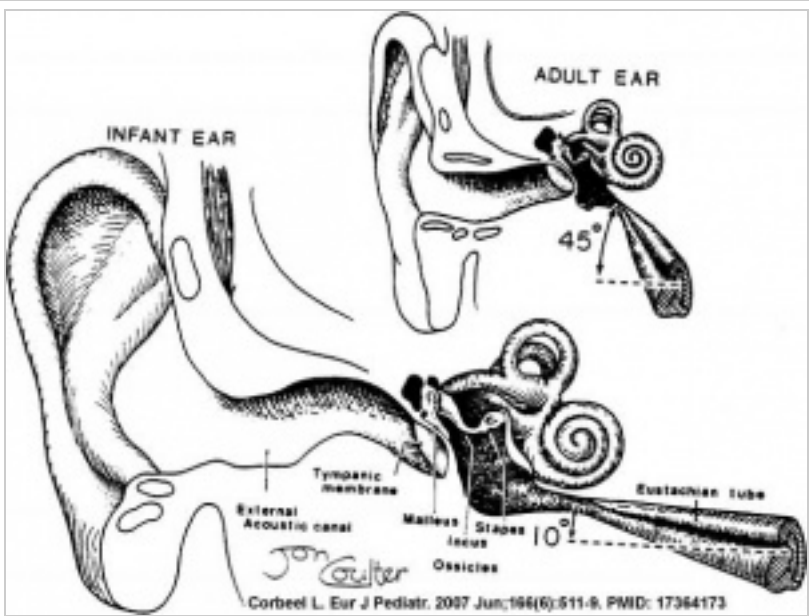
- **NIDCD** - Balance Disorders (http://www.nidcd.nih.gov/health/balance/balance_disorders.asp)
- **Embryo Images Online** (http://www.med.unc.edu/embryo_images/)
 - **Eye Development** - Eye Development Unit (http://www.med.unc.edu/embryo_images/unit-eye/eye_htms/eyetoc.htm) | Eye Fields-Optic Vesicle (Weeks 3-4) (http://www.med.unc.edu/embryo_images/unit-eye/eye_htms/eye001.htm) | Optic Cup, Lens Vesicle, Choroid Fissure, Hyaloid Artery (Weeks 5-6) (http://www.med.unc.edu/embryo_images/unit-eye/eye_htms/eye009.htm) | Cornea, Anterior Chamber, Pupillary Membrane, Lens, Retina (Weeks 7-8) (http://www.med.unc.edu/embryo_images/unit-eye/eye_htms/eye016.htm) | Iris, Cilliary Body (Weeks 9-15) (http://www.med.unc.edu/embryo_images/unit-eye/eye_htms/eye022.htm) | Eyelids (Weeks 8-10) (http://www.med.unc.edu/embryo_images/unit-eye/eye_htms/eye025.htm)
 - **Ear Development** - Ear Development Unit (http://www.med.unc.edu/embryo_images/unit-ear/ear_htms/eartoc.htm) | Inner Ear | (http://www.med.unc.edu/embryo_images/unit-ear/ear_htms/ear012.htm) Middle Ear (http://www.med.unc.edu/embryo_images/unit-ear/ear_htms/ear001.htm) | External Ear (http://www.med.unc.edu/embryo_images/unit-ear/ear_htms/ear014.htm)

Terms

- **altricial animal** - Term used to describe an animal born in a helpless state, with incomplete development of sensory systems at birth. For example rats and mice are born with incomplete development of visual and auditory systems.
- **ampulla** - Term used to describe an anatomical dilation of a tube or canal lumen. Anatomical description of the opening end of the uterine tube lying above the ovary and the enlarged initial segmeny of the semicircular canals of the inner ear vestibular system. (More? [ear6.htm Inner Ear] | [genitalXXuterus.htm Genital System - Female Uterus])
- **aneurism** - (Greek, *aneurysma* = a widening, aneurysm) A term used to describe an abnormal widening of a vessel or anatomical tubal structure.
- **aquaeductus vestibuli** - see vestibular aqueduct
- **auditory neuropathy** - (AN) abnormality of transmission of sound information to the brain.
- **auditory tube** - (eustachian tube) between the middle ear and oral cavity, has a bony (tympanic 1/3) and cartilaginous (pharyngeal 2/3) portion. The main role is equalization of pressure and fluid drainage in the middle ear.
- **auricular hillock** - see hillock
- **atresia** - narrowing, usually of an anatomical tube or cavity.
- **autophagocytosis** - (Greek, auto = self, phagy = eating, also called autophagy) a cell death mechanism that uses the cell's own lysosomes to self digest.
- **border cells** - columnar cells within the organ of Corti on the medial portion of the basilar membrane.
- **canalis reuniens** - (ductus reuniens, canaliculus reuniens, canalis reuniens, Hensen's canal, Hensen's duct, uniting canal, canalis reuniens of Hensen) short narrow canal connecting the cochlea duct to the saccule. (Victor Hensen, 1835-1924)
- **cerumen** - (ear wax) produced by glands in the skin of the outer portion of the ear canal.
- **chondrified** - the developmental differentiation of cartilage from mesenchye, an embryonic connective tissue.
- **cristae ampullaris** - located in the ampulla of the membranous semicircular canals a region with both supporting and hair cells. The hair cell cilia are embedded in the gelatinous cupula.
- **claudius cells** - (cells of Claudius) columnar cells with microvilli overlying the basilar membrane and extend from Hensen's cells to the spiral prominence. Barrier cells that lie external to the organ of corti in endolymph.
- **cochlear sac** - embryonic structure, which will form the coiled cochlear duct and contribute to the saccule.
- **cochlear aqueduct** - a bony channel containing the fibrous periotic duct. It connects the basal turn of the cochlea perilymphatic space with the subarachnoid space of the posterior cranial cavity.
- **cochlin** - major constituent of the inner ear extracellular matrix.
- **collagen type II** - major constituent of the inner ear extracellular matrix.
- **conductive loss** - term used to describe one of the two major classes of hearing loss involving external and middle ear abnormalities (other form is Sensorineural loss).
- **connexins** - channel proteins of the gap junctions that allow rapid communication between adjacent cells. The two connexins Cx26 and Cx30 are the major proteins of cochlear gap junctions.
- **connexin 26** - A strikingly high proportion (50%) of congenital bilateral nonsyndromic sensorineural deafness cases have been linked to mutations in the GJB2 coding for the connexin26
- **cupular deposits** - basophilic material on the cupulae of the semicircular ducts, an postnatal ageing phenomenon seen in some vestibular labyrinth.
- **clinical weeks** - taken from last menstrual period (LMP) and therefore approximately two weeks before fertilization occurs.
- Deiters' cells
- **discoidin domain receptor 1** - (DDR1) a tyrosine kinase receptor activated by native collagen, expressed in the basement membrane and with fibrillar collagens. Found in basal cells of the stria vascularis, type III fibrocytes, and cells lining the basilar membrane of the organ of Corti. {Meyer zum Gottesberge, 2008 #1877}
- ductus utriculosaccularis -
- **endochondral ossification** - the process of bone formation from a pre-existing cartilage template.
- endolymphatic fluid -
- **endolymphatic sac** - inner ear structure that has anatomically both an intraosseous and extraosseous component. Th e sac has functions regulating endolymph that are both secretory and absorptive. Also the site of endolymphatic sac tumors either sporadical occurring or associated with the autosomal-dominant von Hippel-Lindau (VHL) disease, due to a germ line mutation.
- **embryological weeks** - taken from the time of fertilization which typically occurs around the middle (day 14), or just after, of the typical 28 day menstrual cycle.
- **Emx2** - homeobox gene affecting middle ear and inner ear development.
- **eustachian tube** - (auditory tube) A cavity linking the pharynx to the middle ear, which develops from the first pharyngeal pouch. Named after Bartolomeo Eustachi (1500 - 1574) an Italian anatomist. Several functions including the equalization of pressure in the middle ear.
- **external auditory meatus** - (ear canal) develops from the first pharyngeal cleft.
- **ear wax** - see cerumen.
- **espins** - calcium-resistant actin-bundling proteins enriched in hair cell stereocilia and sensory cell microvilli and spiral ganglion neurons (SGNs)
- external auditory canal -
- **fenestra ovalis** - (oval window) separates the tympanic cavity from the vestibule of the osseous labyrinth.



Fetal Alcohol Syndrome Face



Eustacian tube angle

- **fenestra rotunda** - (round window) separates the tympanic cavity from the scala tympani of the cochlea.
- **fetus** - (foetus) term used to describe human development after the 8th week (10th clinical week, LPM) and covers the developmental periods of second and third trimester.
- **fibroblast growth factor 1** - (Fgf-1) a growth factor released from cochlea sensory epithelium which stimulates spiral ganglion neurite branching.
- **fibroblast growth factor 8** - (Fgf-8) a growth factor released by inner hair cells which regulates pillar cell number, position and rate of development.
- **fibroblast growth factor receptor 3** - (Fgfr-3) a tyrosine kinase receptor with a role in the commitment, differentiation and position of pillar cells in the organ of corti
- **fundamental frequency** - (natural frequency) the lowest frequency in a harmonic series, for the female voice this is about 225 Hz.
- **helicotrema** - term used to describe the cochlear apex.
- **Hes** - (hairy and enhancer of split) family of factors, which has been shown to be a general negative regulator of neurogenesis (Zheng, 2000).
- **hillock** - a small hill, used to describe the six surface elevations on pharyngeal arch one and two.
- Hindbrain - Invaginate -
- **Incus** - (anvil) auditory ossicle
- inner phalangeal cells
- **inner pillar cells** - organ of Corti cells arranged in rows and form a boundary between the single row of inner hair cells and three rows of outer hair cells. These cells have surface-associated microtubule bundles.
- inner sulcus - area of the cochlear duct
- interdental region -
- **internal auditory meatus** - (internal acoustic meatus, IAM) Anatomical canal in which CN VII and CN VIII ganglia reside and pass through to the brainstem. This bony canal lies between the posterior surface of the petrous pyramid and the bony labyrinth within the dense petrous bone. Also associated clinically with the site where acoustic neuromas may occur.
- **Kolliker's organ** - (Kollicker's organ, greater epithelial ridge) Developing cochlear structure consisting of columnar-shaped supporting cells filling the inner sulcus and lying directly under the tectorial membrane. This transient organ regresses and generates the space of the inner sulcus. Rudolph Albert von Kolliker (1817-1905)??
- lateral semicircular duct -
- Limbus -
- **LMP** - acronym for last menstrual period, used to clinically measure gestation.
- **malleus** - (hammer) auditory ossicle
- **mastoid process** - of temporal bone
- **Math1** - homolog of the Drosophila proneural gene atonal, necessary and sufficient for the production of hair cells in the mouse inner ear. {Chen, 2002 #1932}Negatively regulated by Hes1 and Hes5
- **meatal plug** - temporary blockage of the external auditory meatus which forms at the end of the embryonic period and remains present until the seventh month.
- **meatus** - anatomical opening, cavity or space (external acoustic meatus,internal auditory meatus)
- **Meckel's cartilage** - first pharyngeal ach cartilage, located within the mandibular prominence. This cartilage first appears at stage 16, stage 20 the beginning of membranous ossification. Named after Johann Friedrich Meckel, (1781 - 1833) a German anatomist. (<http://www.whonamedit.com/doctor.cfm/1840.html>)
- membranous labyrinth - Mesenchyme - Mesoderm - Microtia - Modiolus -
- **mucopolysaccharidosis** - (MPS IIIB, Sanfilippo Syndrome type B) abnormality caused by a deficiency in the lysosomal enzyme N-acetylglucosaminidase (Naglu). Children with MPS IIIB develop abnormal hearing, and mental functioning culminating in early death.
- **netrin-1** - secreted growth factor, expressed in the organ of Corti and spiral ganglion cells, role in process outgrowth.
- neural tube -
- **olivocochlear** - brainstem cholinergic and GABAergic efferent system that innervates sensory cells and sensory neurons of the inner ear.
- organ of Corti - organ of Corti protein II - (OCP-II) cytosolic protein or transcription factor?
- **otolithic membrane** - extracellular matrix that cover the sensory epithelia of the inner ear.
- **ossicle** - (small bone) the individual bone of the three middle ear bones (auditory ossicles), which reduce vibrational amplitude but increase force to drive fluid-filled inner ear.
- ossify - the process of bone formation.
- otic capsule -
- otic cup
- otic placode -
- otic vesicle -
- **otoconin** - inner ear biominerals required for vestibular apparatus function.
- **otogelin** - (Otog) an inner ear specific glycoprotein expressed in cochlea cells at different developmental times.
- **otolithic membrane** - a membrane within the utricle and saccule containing embedded hair cell cilia and small crystalline bodies of calcium carbonate (otoliths). Functions to detect head motion.
- **otoliths** - small crystalline bodies of calcium carbonate found within the otolithic membrane of the utricle and saccule.
- **ototoxic** - compound or drug causing temporary or permanent hearing loss.
- **outer hair cells** - (OHCs) three rows of hair cells that function to increase basilar membrane motion through a local mechanical feedback process within the cochlea, the "cochlear amplifier".
- **outer pillar cells** - arranged in rows and form a boundary between the single row of inner hair cells and three rows of outer hair cells.
- **paratubal musculature** - muscles lying beside the auditory (Eustachian) tube. The tensor veli, palatini (TVP) and tensor tympani muscles.
- perilymph - perilymphatic space - Periotic Capsule - petrous portion - of temporal bone
- **pejvakin gene** - in humans, two missense mutations in this gene cause nonsyndromic recessive deafness (DFNB59) by affecting the function of auditory neurons.
- pharyngeal archpharyngeal pouchpharyngeal membranePharynx
- **pillar cells** - (PC) form an inner and outer row of support cells that form a boundary between inner and outer hair cells.
- Placode
- **preyer reflex** - ear flick in mouse in response to sound.
- presbycusis
- **prestin** - a motor protein structurally similar to the anion transporter family expressed in cochlear outer hair cells.
- **preauricular tag** - skin tags located in front of the external ear opening, are common in neonates and in most cases are normal, though in some cases are indicative of other associated abnormalities.
- primordium-
- **protocadherin 15** - (Pcdh15) required for initial formation of stereocilia bundles and changes in the actin meshwork within hair cells. The Ames waltzer (av) mouse mutant has both auditory and vestibular abnormalities from a mutation in this gene.
- **Reichert's cartilage** - pharyngeal ach 2 cartilage, named after Karl Bogislaus Reichert (1811 - 1883) a German anatomist.
- **Reissner's membrane** - (vestibular membrane, vestibular wall) is a membrane located inside the cochlea separating the scala media from scala vestibuli. Named after Ernst Reissner (1824-1878) a German anatomist. ,ÄÚIt primarily functions as a diffusion barrier, allowing nutrients to travel from the

- perilymph to the endolymph of the membranous labyrinth.
- rhombomere -
- Sacculus macula -
- Saccule - (Latin, sacculus = a small pouch)
- sacculocollic reflex -
- scala tympani - one of the three Cochlea cavities, it is filled with perilymph.
- **Scarpa's ganglion** - (vestibular ganglion) primary afferent vestibular neuron ganglion of the vestibular nerve. Located within the internal auditory meatus.
- **semicircular canals** - series of fluid-filled loops of the inner ear required for balance and sensing acceleration.
- sensorineural - term used to describe one of the two major classes of hearing loss involving the central pathway from the cochlear (other form is conductive loss).
- **space of Nuel** - within the cochlea, an organ of Corti space between the outer pillar cells and the phalangeal and hair cells. Named after Jean-Pierre Nuel (1847-1920) a Belgian ophthalmologist.
- **spiral ganglion neurons** - (SGN) innervate the inner (Type I) and outer (Type II) hair cells of the cochlea.
- **stapedius muscle** - (innervated by CN VII tympanic branch) one of the two muscles in the middle ear, contraction of this muscle pulls the stapes and dampens auditory ossicle movement.
- **stapes** - (stirrup) a middle ear auditory ossicle (bone).stapes footplate - startle response -
- **stereocilia** -finger-like projections from the apical surface of sensory hair cells forming the hair bundle in the cochlea. Formed by tightly cross-linked parallel actin filaments in a paracrystalline array with cell surface specializations (tip links, horizontal top connectors, and tectorial membrane attachment crowns).
- **stratified squamous epithelia** - classification of epithelium which transiently forms a plug in external ear canal to the outer eardrum.
- **stria vascularis** - forms the outer wall of the cochlear duct of the mammalian cochlea is composed primarily of three types of cells. Marginal cells line the lumen of the cochlear duct and are of epithelial origin. Basal cells also form a continuous layer and they may be mesodermal or derived from the neural crest. Intermediate cells are melanocyte-like cells, presumably derived from the neural crest, and are scattered between the marginal and basal cell layers. The stria forms endolymph and also contains a rich supply of blood vessels.
- sulcus -
- **synostotically** - anatomically normally separate skeletal bones fused together.
- **tectorial membrane** - extracellular matrix that cover the sensory epithelial hair cells of the organ of corti within the cochlea.
- **alpha-tectorin and beta-** (TECTA, TECTB) major non-collagenous protein component of the tectorial membrane forming a striated-sheet matrix. Synthesized as glycosylphosphatidylinositol-linked, membrane bound precursors.
- temporal bone -
- **tensor tympani** - (innervated by CN V mandibular nerve) one of the two muscles in the middle ear, contraction of this muscle pulls the malleus and tenses the tympanic membrane, dampening auditory ossicle movement. The muscle arises from auditory tube (cartilaginous portion) and is inserted into the malleus (manubrium near the root).
- teratogens - trilaminar embryo -
- **tonotopy** - term describing the mapping along the tectorial membrane within the cochlea of the different sound frequencies.
- tympanic cavity - tympanic membrane -Utricle -Vacuolization - Vesicle - vestibular apparatus - vestibular evoked myogenic potential (VEMP) test
- **vestibular ganglion** - (Scarpa's ganglion) primary afferent vestibular neuron ganglion of the vestibular nerve. Located within the internal auditory meatus.
- **vestibular membrane** - (Reissner's) extends from the spiral lamina to the outer wall and divides the cochlea into an upper scala vestibuli, a lower scala tympani.
- **Vestibulocochlear Nerve** - Cranial Nerve VIII
- **Whirlin** - A PDZ scaffold protein expressed in hair cells at the stereocilia tips, essential for the stereocilia elongation process. The DFNB31 gene mutations cause hearing loss in human and mouse. This protein can interact with membrane-associated guanylate kinase (MAGUK) protein, erythrocyte protein p55 (p55).
- **Wnt7a** - signaling through the Wnt pathway regulates the development of hair cell unidirectional stereociliary bundle orientation.

2015 Course: **Week 2** Lecture 1 Lecture 2 Lab 1 | **Week 3** Lecture 3 Lecture 4 Lab 2 | **Week 4** Lecture 5 Lecture 6 Lab 3 | **Week 5** Lecture 7 Lecture 8 Lab 4 | **Week 6** Lecture 9 Lecture 10 Lab 5 | **Week 7** Lecture 11 Lecture 12 Lab 6 | **Week 8** Lecture 13 Lecture 14 Lab 7 | **Week 9** Lecture 15 Lecture 16 Lab 8 | **Week 10** Lecture 17 Lecture 18 Lab 9 | **Week 11** Lecture 19 **Lecture 20** Lab 10 | **Week 12** Lecture 21 Lecture 22 Lab 11 | **Week 13** Lecture 23 Lecture 24 Lab 12 | **Projects:** Group 1 | Group 2 | Group 3 | Group 4 | Group 5 | Group 6 | Students | Student Designed Quiz Questions | Moodle page (<http://moodle.telt.unsw.edu.au/course/view.php?id=15814>)

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