Auditory Neuropathy

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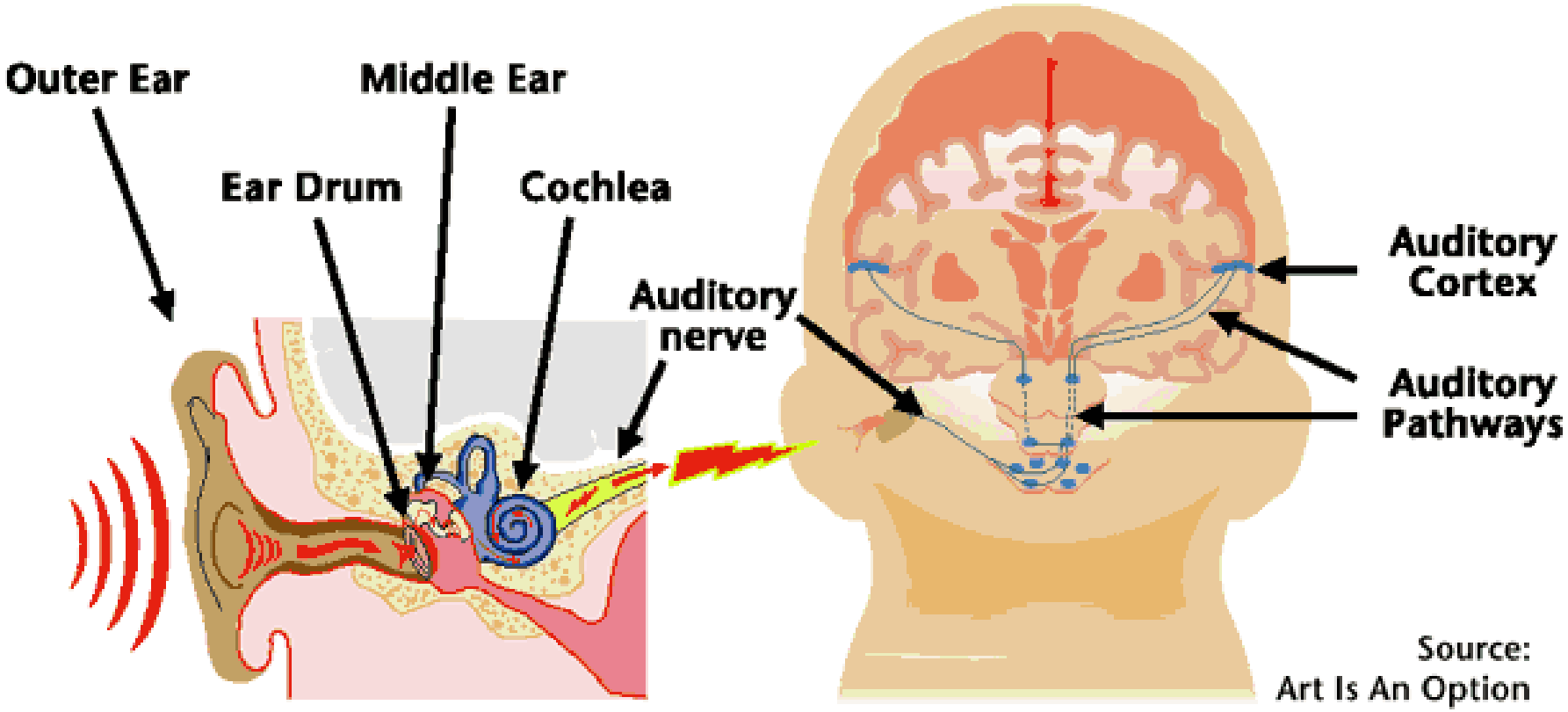
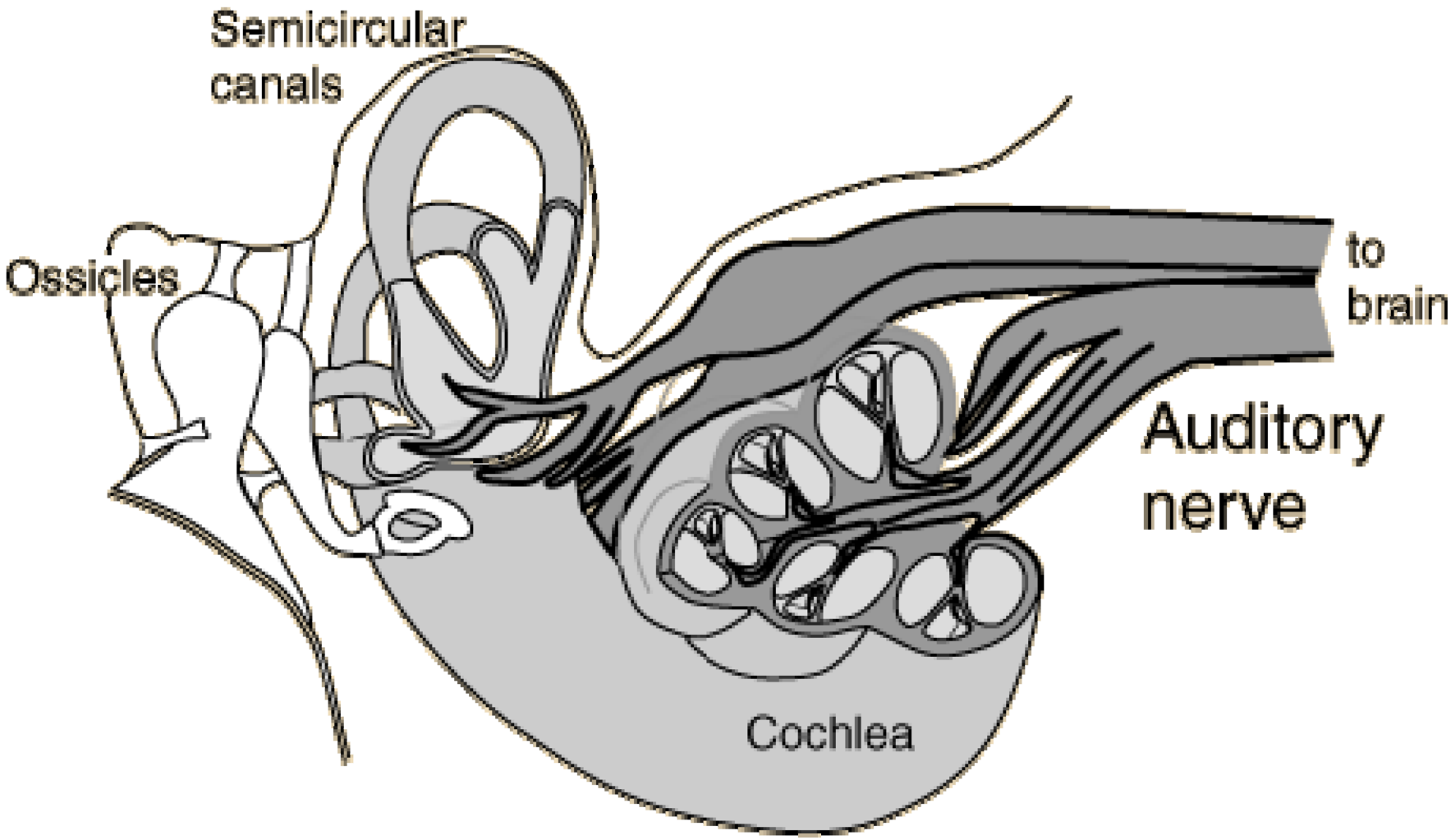
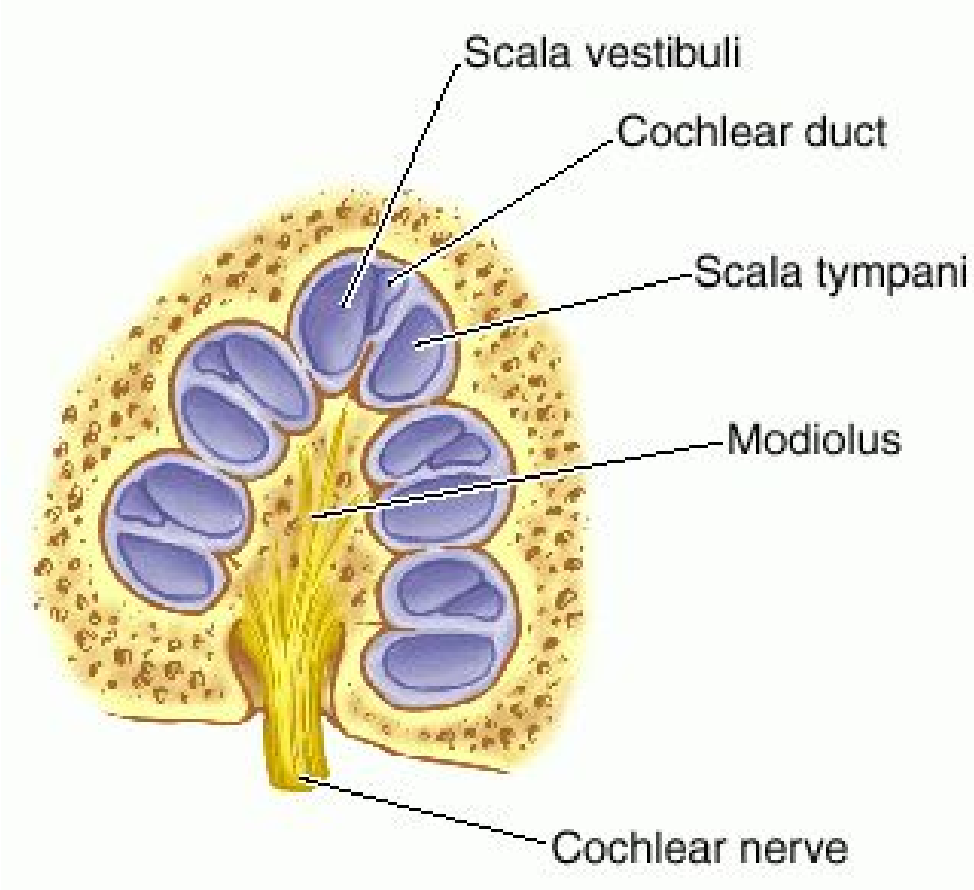
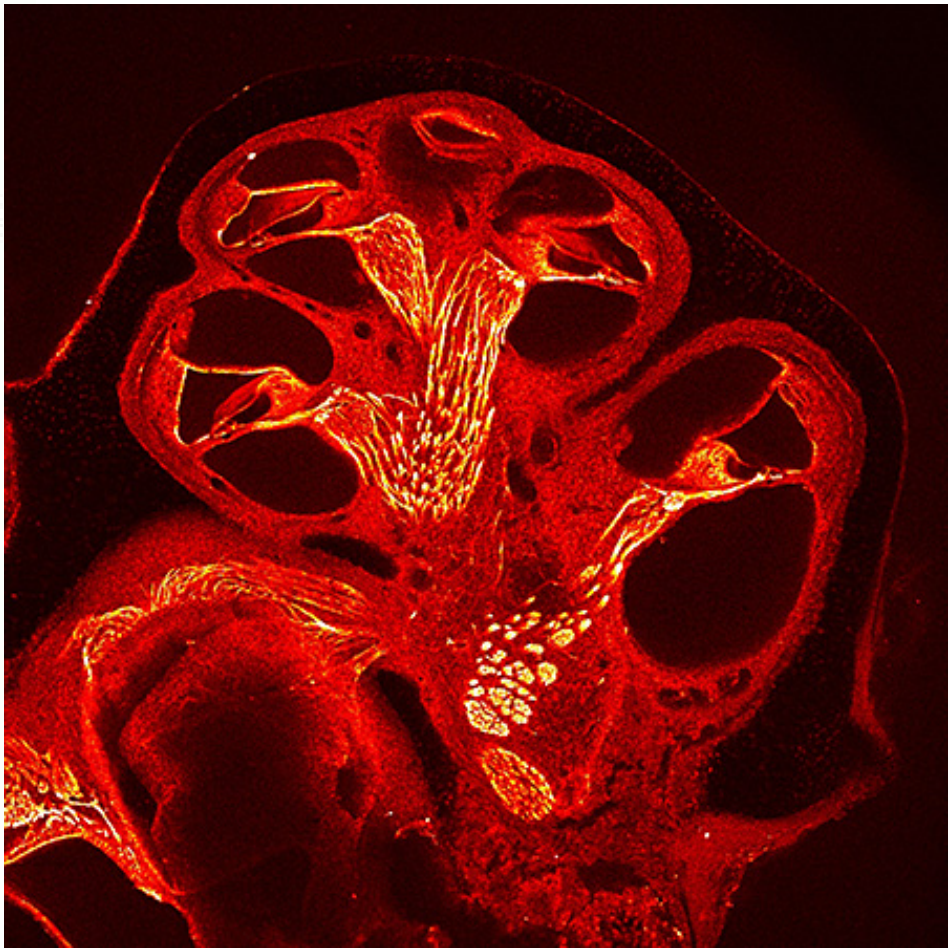
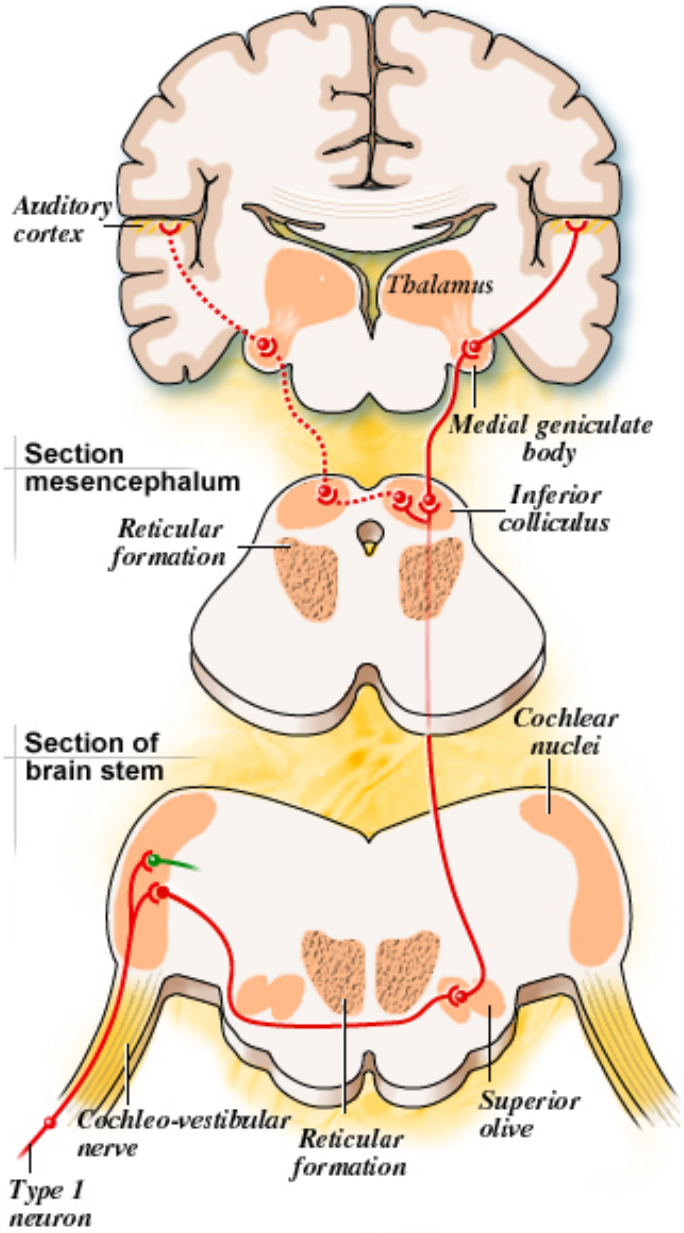
Anatomy


Outer Ear
Middle Ear
Ear Drum
Cochlea
Auditory nerve
Auditory Cortex
Auditory Pathways
Source:
Art Is An Option


Semicircular canals
Ossicles
to brain
Auditory nerve
Cochlea


Scala vestibuli
Cochlear duct
Scala tympani
Modiolus
Cochlear nerve





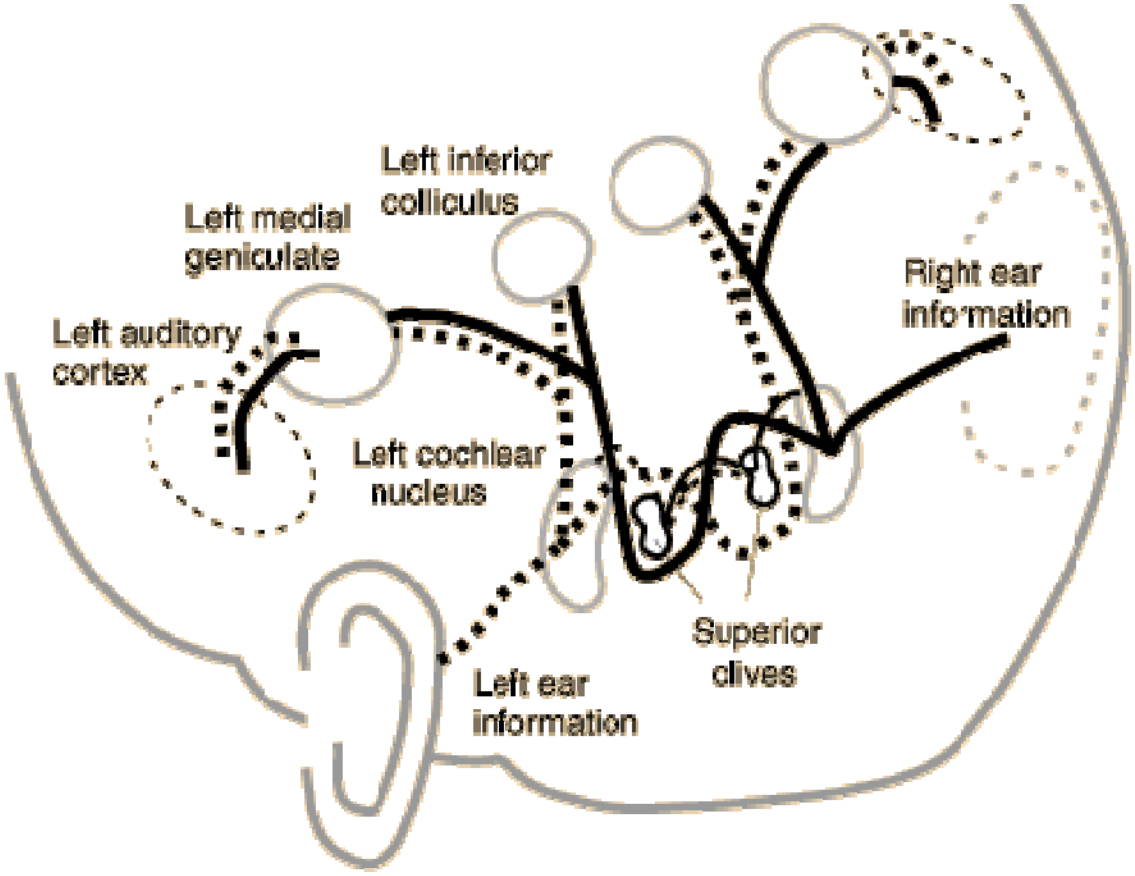
The final neuron of the primary auditory pathway links the thalamus to the auditory cortex, where the message, already largely decoded during its passage through the previous neurons in the pathway, is recognized, memorized and perhaps integrated into a voluntary response.

A final relay, before the cortex, occurs in the thalamus (median geniculate body); it's here that an important integration occurs: preparation of a motor response (eg vocal response).

Leaving this relay, a third neuron carries the mesage up to the level of the mesencephalus (superior colliculus). These two relays play an essential rôle in the localisation of sound.

The second major relay in the brain stem is in the superior olivary complex: the majority of the auditory fibres synapse there having already crossed the midline.

The first relay of the primary auditory pathway occurs in the cochlear nuclei in the brain stem, which receive Type I spiral ganglion axons (auditory nerve); at this level an important decoding of the basic signal occurs: duration, intensity and frequency.


Left inferior
colliculus
Left medial
geniculate
Left auditory
cortex
Left cochlear
nucleus
Right ear
information
Superior
olives
Left ear
information

Definitions

- Neuropathy:
  - Problem with the nerve fibers as they leave the cell body
    - Demyelinating – problem with the cells that make the sheath around the nerve fibers
    - Axonal – problem with the nerve fibers
    - Mixed – combination of both

Definitions

- Dys-synchrony:
  - Inability of the nerve to transmit signals in a coherent fashion

Criteria

1. Recordable otoacoustic emissions or cochlear microphonic (indication of intact cochlear function)
2. Absent or atypical auditory brainstem response (indication of abnormal cochlear nerve or central auditory pathway function)
3. Speech understanding worse than would be expected for the pure tone hearing loss
4. Hearing loss of variable severity and configuration

	Anatomy	Dysfunction	Location	Auditory Test
SENSORY CELLS	***IHC*** *- (auditory receptors)* - each synapses with myelinated dendrites of multiple Type I spiral ganglion neurons (receive a few OCB axons) ***OHC*** *- (modulating)* - groups synapse with unmyelinated dendrites of one Type II spiral ganglion neuron (innervated by OCB axons)	**HAIR CELL DEATH/ DAMAGE**	*IHC* *OHC*	*OAE? CM?* *OAE* *CM*
SYNAPSE	*Hair cells with* **dendrites** of the **spiral ganglion neurons**	**SYNAPTIC BLOCK**	*Synapse*	*No specific test*
SPIRAL GANGLION NEURONS	***Type I*** *(first order auditory neurons) myelinated dendrites synapse with IHC; axons project to brainstem CN* ***Type II*** *(sensory neurons) small unmyelinated dendrites synapse with OHC; physiologic properties unknown; axons project to brainstem CN*	**NEURONOPATHY** (ganglionapathy)	*Spiral ganglion*	*No specific test*
VIIIth NERVE	***Afferent auditory axons*** *(95% = Auditory Type I)*: myelinated axons; synapse with neurons of the CN *(5% = Modulating Type II)*: scarce unmyelinated axons; synapse with neurons of the CN ***Efferent OCB axons (auditory)*** Axons from SOC via Medial OC (myelinated); target: OHC Sparse lateral OC (unmyelinated); target: dendrites of Type I SGN ***Afferent vestibular axons*** Myelinated vestibular axons (first order neurons) synapse with 2nd-order vestibular neurons in BS	**NEUROPATHY** Demyelinating Axonal Mixed	*Auditory Type I* *OCB* *Vestibular*	*ABR Wave I* *ABR Wave II* *MEMR (acoustic)* *OAE suppression* *(OHC activity modified though efferent MOC connections)* *Vestibular tests*
BRAINSTEM PATHWAY	***Cochlear nucleus*** (second order auditory neurons) ***Superior olivary complex*** ***Inferior colliculus***	**BRAINSTEM DISORDER**	*Brainstem*	*ABR Wave III*, *ABR Wave V* } SOC to IC *MEMR (acoustic & tactile)*
THALAMUS	***Medial geniculate*** *body*	**THALAMO-CORTICAL DISORDER**	*Thalamo-cortex*	*MLR* *MMN*
CORTEX	***Primary auditory cortex*** (Heschl's gyrus) ***Secondary auditory cortex*** (superior temporal gyrus)		*Cerebral cortex*	*CAEP* *late obligatory event related potentials*

Diagnosis

- Physical examination can be normal
- Must meet all the criteria
- Can be found on newborn hearing screening if both OAEs and ABR are performed
- Often the diagnosis is delayed

Prevalence

- May be up to 8% of newly diagnosed pediatric hearing loss per year
- Up to 11% of children with a confirmed permanent hearing loss
  - Genetic
- May affect up to 2% of infants in the neonatal intensive care unit
  - Hypoxia (lack of oxygen)
  - Hyperbilirubinemia (too much bilirubin)

Treatment

- Close monitoring of hearing
  - Hearing may change with time
- Amplification with hearing aids
  - Debate regarding efficacy
  - Should be trialed
- Speech and language therapy
- Cochlear implantation
  - Approach cautiously
  - Possible extra testing
  - Results variable