Voice
COCHLEAR IMPLANT REHAB

Handbook for speech and language therapists and parents for teaching children with cochlear implants how to speak

Vocational education and training for speech and language therapists and parents for rehabilitationof children with cochlear implant on how to speak

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"Shoot for the moon. Even if you miss, you'll land among the stars." Les Brown

Introduction

Hearing is a very important sense for the normal development of the modern individual. The first steps in natural hearing are healthy and functional anatomic structures. Pathology involving any key level in sound transmission and formation will result in partial or total deafness, unilaterally or bilaterally. Recognizing normal anatomic architecture will improve diagnosis and treatment possibilities. Depending on the site of the lesion, different treatment options can be used with optimal results.

What are the elements involved in normal sound perception?
How is sound transformed from sound wave to neural input?

1. Anatomy of the Ear and Hearing

The ear is the organ of hearing and balance. It is the peripheral segment of two of the most important sensors in the context of normal social adaptation and integration. Hearing health allows the development of language and communication and an adequate insertion into the environment.

The peripheral part of the analyzer is represented by the ear, which describes three segments: external, medium, and internal. From the inner ear, signals are transmitted through the auditory and vestibular nerves to the brainstem and then to the cerebral cortex.

External Ear

The outer ear consists of the pavilion (pinna) and external auditory canal (EAC). The pinna is located on the side of the head; it is a blade of elastic cartilage covered by skin, forming an angle of about 25–30 degrees with the skull (1). An anterior and a posterior face are described, with a series of reliefs with a role in directing the sound to the middle ear (Figure 1).

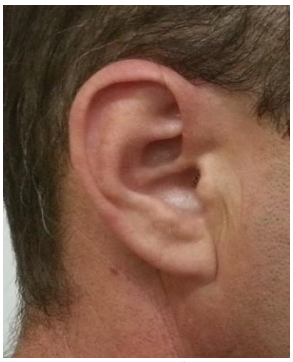


Figure 1. External Ear – pinna

The external auditory canal is about 3 cm long and has a variable diameter. The path to the middle ear is curved, and in order to be able to see the tympanic membrane during the otoscopic examination, it is necessary to pull the pavilion upwards and backwards.

In the external two-thirds, the auditory canal is cartilaginous, and in the internal one-third it is bone. It is covered with skin, which has numerous hair follicles and wax producing glands in the cartilaginous part [2].

These structures have a protective role, with the brushes retaining any particles, and the earwax being antiseptic, so that the external auditory canal in the internal one-third is practically sterile.

The innervation is provided by branches of the trigeminal nerve, the vagus nerve, the superficial cervical plexus, and the facial nerve.

Vascularization of the outer ear is provided by the superficial temporal artery, anterior and posterior auricular arteries (Figure 2). The lymph drains into lymph nodes located in the parotid, retroauricular, and upper cervical ganglia.

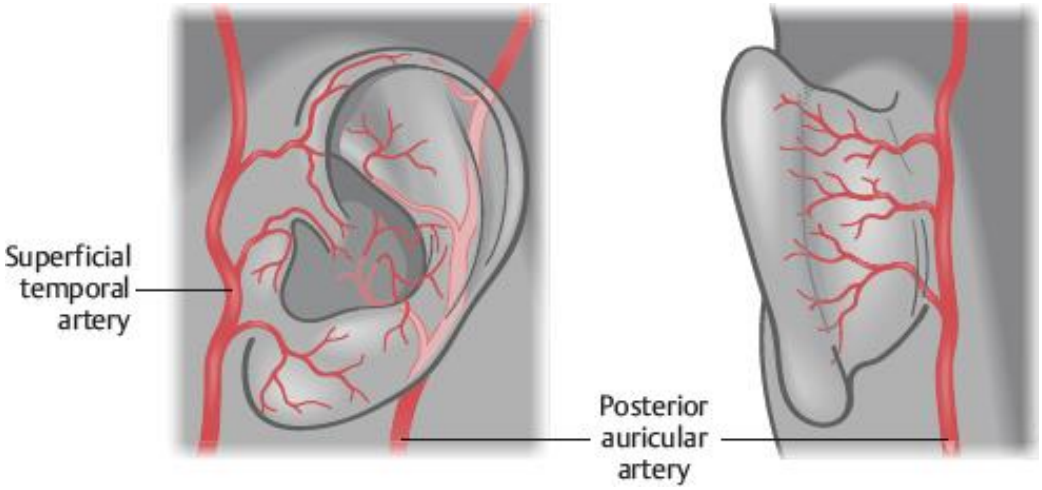


Figure 2. External ear vasculature

Middle Ear

It consists of the tympanic cavity, the mastoid process with the pneumatic system, and the Eustachian tube (Figure 3).

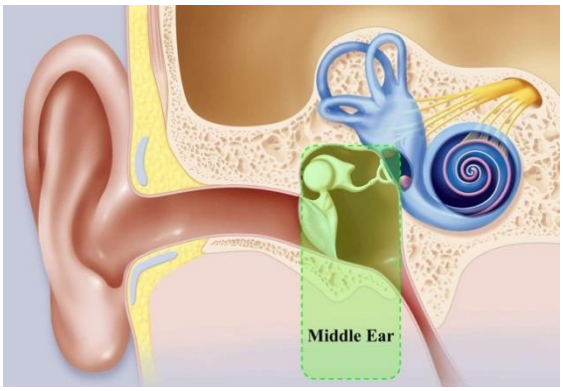


Figure 3: Middle ear

a. The tympanic cavity (TC) has 6 walls:

- the external wall, mostly occupied by the tympanic membrane. It is covered externally by the epithelium, under which is a layer of conjunctival fibers arranged radially, circularly, and parabolically, which gives it strength. The internal surface (towards the middle ear) is lined with mucosa.
- the superior wall, “tegmen tympani”, about 1 mm thick, which separates the tympanic cavity from the endocranium.
- the inferior wall, which is related to the jugular vein bulb.
- the anterior wall contains the tympanic opening of the Eustachian tube (Et) and is in proximity to the internal carotid artery.
- the internal wall houses the oval window with the stapes and, in the inferior part, the round window. Between the two windows there is a bulge called the promontory. The middle ear communicates with the inner ear via the windows. Above the oval window is the bony canal of the facial nerve.
- the posterior wall, through which it comes in relation to the mastoid. On this wall, the "aditus ad antrum" opens, which represents the communication between the mastoid antrum, the largest cell in the groups of cells found in the mastoid, and the TC. Also, at this level, the facial nerve makes an elbow and leaves the TC to enter the mastoid, becoming vertical.

The mucosa covering the walls of the TC is a cuboidal stratified epithelium, becoming pseudostratified, and ciliated only in the lower part of the anterior wall, at the level of the opening of the Et orifice.

The middle ear is vascularized by branches of the maxillary artery (tympanic artery and middle meningeal) and the stylomastoid artery.

The innervation is provided by the tympanic branch of the glossopharyngeal nerve and partially by the trigeminal nerve, through the auriculotemporal branch.

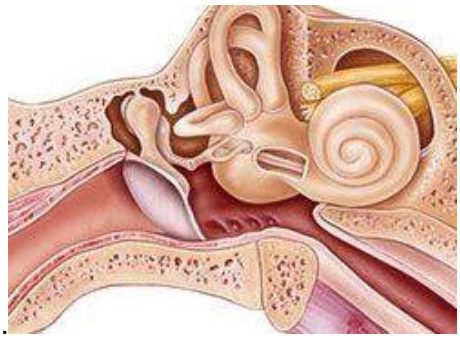
Inside the middle ear, the ossicular chain system consists of hammer, incus, and stapes.

The hammer, through the handle, is fixed on the tympanic membrane and articulates with the incus, which in turn articulates with the stapes. The stapes is fixed to the oval window with a footplate that faces the inner ear.

The ossicular system is fixed by ligaments and muscles to the walls of the TC and is mobilized by the movements of the tympanic membrane, to which the hammer handle is fixed.

The movement of the ossicular chain is transmitted at the level of the oval window to the fluids of the inner ear, making hearing possible (Figure 4).

The ossicular system is located mainly in the upper part of the tympanic cavity (3). Infections in this area (called the attic) are more dangerous because this space is separated by ligaments that suspend the ossicular chain. Drainage will be disturbed, and the persistence of the infection favors osteitis of the ossicular chain and middle ear walls [4].

a. 

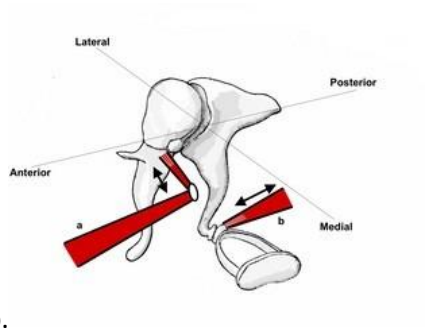
b. 

Figure 4. Ossicular system of the middle ear (a) and hammer and stapes muscles (a, b)

The mastoid process is located posteriorly and inferiorly to the TC. It has a pyramidal shape, with the upper base towards the temporal bone, of which it is part, and the lower tip attached to the sternocleidomastoid muscle. The mastoid contains air-filled cells that are ventilated through the "aditus ad antrum" from the TC, which communicates with the nasopharynx through the Et.

In the posterior part, the mastoid is crossed by the lateral venous sinus (sigmoid sinus), which in the lower part makes a triple elbow and becomes the internal jugular vein at the exit of the skull through the ruptured posterior hole. The facial nerve becomes vertical under the "aditus ad antrum" and runs through the anterior part of the mastoid, heading towards the stylomastoid foramen, to pierce the parotid gland.

b. Eustachian tube is a canal that runs from the anterior wall of the TC to the lateral wall of the nasopharynx, where it opens through the nasopharyngeal orifice of the Eustachian tube. It has a length of 35-45 mm and has a fibro-cartilaginous portion in two-thirds of the internal length, and a bony portion, in one-third of the external length, in relation to the middle ear. The Eustachian tube is lined with epithelium that continues with the rhino-pharyngeal epithelium.

At the level of the fallopian tube, we find the external and internal peristafilin muscles, which constitute the motor system that helps intermittent opening of the orifice [3].

The role of the Eustachian tube is:

- to equalize the pressure in the middle ear with the atmospheric pressure;
- drainage of the middle ear;
- middle ear protection.

Inner Ear or Labyrinth

The inner ear contains receptors for hearing and balance (Figure 5).

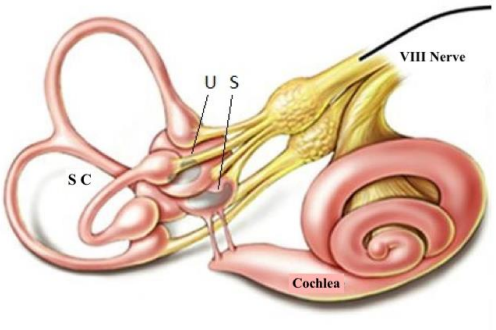


Figure 5. Inner ear (SC – semicircular canals; U – utricle; S – saccule; VIIIth Nerve – acoustic-vestibular nerve)

Hearing aids are located in the cochlea, and those for balance are located in the vestibule and semi-circular canals.

a. The anterior bony labyrinth, or bony cochlea, is a spiral tube that describes two and a half turns around a central conical axis called a columella. Attached to the columella, a bony blade divides the cochlea into two compartments (scala)—one vestibular and one tympanic.

The basal turn (first spire) protrudes on the inner wall of the TC, forming the promontory. In the upper part, the two scala communicate at the level of the helicotrema.

The columella is crossed by fine canals through which the fibers of the cochlear nerve pass to the specialized cells with the role of auditory receptor.

The bony cochlea houses the membranous cochlea, which forms a third compartment, the cochlear duct (CD) or scala media.

Inside the CD the Corti's organ can be found, specialized as an auditory sensor. Corti's organ is composed of the basilar membrane, above which the inner and the outer hair cells are located. They are specialized as auditory receptors, supported by external and internal pillars (support cells), and covered by the tectorial membrane.

The space between the bony and the membranous labyrinth is filled with perilymph, and inside the membranous labyrinth is the endolymph.

Perilymph is a filtrate of cerebrospinal fluid with a chemical composition identical to that of extracellular fluid, in which Na+ predominates, while *endolymph* has a predominantly K+ composition, similar to intracellular fluid, and originating from stria vascularis (vessels located at the external wall of the cochlear duct).

The first neuron of the auditory pathway is located in Corti's ganglion, from which the dendrites go to the hair cells at the level of the Corti organ, and the axons go to the cochlear nuclei in the bulb-protuberant area.

Most axons (90%) go to the inner hair cells, with each cell receiving more nerve fibers.

The rest of the fibers go to the outer hair cells, with each fiber innervating several cells.

The vascularization of the labyrinth is ensured by the internal auditory artery, a branch of the anteroinferior cerebellar artery.

b. The posterior bony labyrinth

The posterior bony labyrinth consists of a central cavity, or vestibule, and three semi-circular canals, which open inside it.

The vestibule is an irregular, rectangular cavity located between the middle ear and the internal ear canal.

On the outer face, we find the oval window, with the inner wall being crossed by nerve fibers from branches of the vestibular nerve.

The three semi-circular canals open in the vestibule through a dilated end called the ampulla.

The plane formed by the three semicircular channels corresponds to the three directions of space: one channel in the horizontal plane and two channels in the vertical plane (Figure 6).

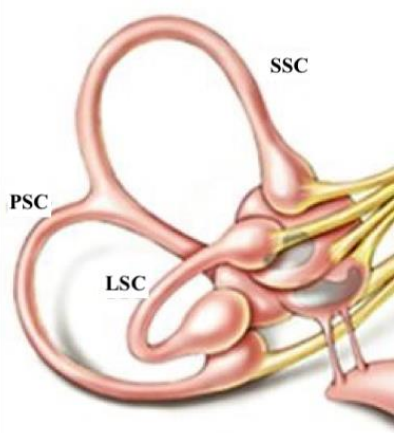


Figure 6. Semicircular canals (SSC – superior semicircular canal; PSC - posterior semicircular canal; LSC - lateral semicircular canal)

The two vertical channels, SSC and PSC, join together in the vestibule through their non-ampullary extremities. CSL has ampullary openings at both ends.

Inside the bony posterior labyrinth, the membranous posterior labyrinth is located. It consists of two vesicles found in the vestibule, respectively the utricle and the saccule, which contain receptors responsible for static balance.

Inside the bony semicircular canals, the membranous semicircular canals are found, with receptors for balance, located at the level of the ampules. Inside the bony labyrinth, we find the perilymph, and in the membranous one the endolymph.

The vestibular ganglion (Scarpa's ganglion) sends its fibers to the sensory cells, located in the macula of the utricle and saccule and in the ampullary ridges at the level of the semicircular canals.

The axons from the vestibular branch of the VIII nerve reach the vestibular nuclei located in the bulbopontine area.

The vascularization of the labyrinth is ensured by the internal auditory artery.

2. Physiology of the Ear and Hearing

The energy produced by molecular vibrations is collected and transformed by the ear into auditory stimulus, which is transported to the auditory neuronal centers and transformed into or interpreted as sound sensation.

This process is accomplished by the three components of the ear: the external ear collects and transports the sound energy, the middle ear transports and amplifies the sound energy, and the inner ear transforms the energy into acoustic stimulus. The acoustic stimulus is transported through auditory nervous

pathways towards the auditory nervous centers, where in conjunction with the cortical layer, it is transformed into sound sensation.

Sound waves are collected by the pinna and directed through the external ear canal towards the tympanic membrane and the middle ear. The **Pinna** architecture on the external side directs the sound waves towards the external ear canal and amplifies frequencies between 1500 and 7000 Hz (with a maximum of 15-20dB at 2500 Hz). The concha of the pinna (middle depression of the pavilion) is a resonating chamber for frequencies between 2000 Hz and 5000 Hz [1].

Another function of the pinna is sound localization. Biauricular localization depends on two factors: differences of sound intensity and differences of sound phase (received by both ears and integrated through cortical processing).

The **EAC** protects the tympanic membrane from direct aggression due to its double curvature. The sebaceous and wax glands offer bacteriological protection to the skin and epidermal layer of the tympanic membrane. There is also an acoustic enhancement function for frequencies between 2000-3000 Hz due to the resonance properties of the canal. These are the most important frequencies for speaking.

At the contact with the **TM,** the sound waves are transformed into vibrations that are transmitted through the ossicular chain towards the oval window and the inner ear liquids.

The TM is both a receptor and a transducer of the sound wave. The vibrations at this level are selective, depending on the frequency of the sound wave. The higher the frequency, the more segmented the tympanic membrane vibration gets [5].

The maximal vibration area of the tympanic membrane is the postero-superior quadrant, regardless of the frequency.

The **ossicular chain** is an intermediate structure that connects two different media: air and liquid. The passing of the sound wave through two different density and elasticity media results in some sound energy reflection.

The difference of surface between the tympanic membrane and the **oval window** [21/1] has also an important role in amplifying the sound energy up to 25dB. This compensates the energy lost during the air-to-liquid media transfer. At this point, sound energy regains up to 60% of its original intensity.

The ossicular chain's architecture and structure have evolved to either facilitate or hinder transmission. This is achieved by the delicate lever mechanism of the malleus and incus body and modulated by contraction of the malleus and stapedius **muscles**. These muscles have a reflex, simultaneous, and antagonist mechanism of action. Contraction of the malleus muscle results in medialization of the TM and a lowering of the vibration amplitude. The stapedius muscle contraction pulls the stapes posteriorly, further blocking vibration [6].

These muscles contribute to frequency selection and the elimination of distorting harmonics (selective muscular contraction results in enhanced sound acuity on certain frequencies), mask low frequencies, and improve the audition of higher frequencies.

Vibrations passing through this system encounter a specific resistance that results from three factors: friction, system mass, and rigidity. This resistance is called impedance, and measuring it is important in

audiology. Measuring the level of the TM offers information regarding the normal functioning of the sound transmission system.

Aside from all the above-mentioned, there is also a direct transfer of sound energy to the inner ear through the bones of the skull (bone conduction).

Footplate movement induces a liquid wave that travels along the cochlea and determines a bulging of the round window. Because liquids are not compressible, the volume moved by the footplate is transmitted through the cochlea to the round window.

The wave produces a movement between the basilar membrane and the tectorial membrane with a specific amplitude according to the frequency of the sound. The maximum height of the amplitude will be isolated by the muscular activity of the external cilia cells, and a specific and selective area of **inner cilia cells** will be stimulated corresponding to the specific frequency of the sound.

The maximum amplitude of the movement will be different and specific for each frequency and will be closest to the apex for lower frequencies and closest to the basal turn of the cochlea for higher frequencies. Each frequency has a specific, corresponding spot on the basilar membrane.

The point of maximum amplitude of the basilar membrane determines a point of excitation in the Corti organ, and this creates an electric stimulus that will be transmitted by the acoustic nerve to the auditory centers.

The mechanic energy produced by the cilia movement produces bioelectric phenomena in the cell. The electric stimulus from the sensory cells of the Corti organ is transformed in the peripheral auditory neuron into the action potential of the auditory nerve. There are several parameters (characteristics) specific to the sound: intensity, frequency, and temporal specificity. These are encoded and transported to the central nervous system.

The nervous influx arriving to the cortex becomes sound sensation [6]. This is achieved by recognizing the sound and integrating it in a meaningful sensation. Through conditioning and memory recording, the sounds are interpreted. Words give the ability of abstract thinking and expressing and offers the opportunity to be integrated in the environment and society.

The vestibular analyzer (VA) is responsible for spatial orientation, keeping vertical position, and the ability to walk. It has a peripheral segment (receptor) located in the posterior portion of the labyrinth, a transmission segment formed by nerves and vestibular nuclei, and a cortical segment where perception and processing occur.

The VA detects linear and angular forces applied to the individual. Along with the information received from the visual system, it contributes to muscle control for a balanced position.

The movement of the head determines endolymphatic movements inside the vestibule. Acceleration in the horizontal and vertical planes determines a movement of the otoliths in the utricle and, respectively, saccule and variable excitation of the cilia cells. The rotation of the head induces a movement through the semicircular canals, which results in the ampullary cilia cells stimulation. The excitation intensity in left and right vestibules varies depending on movement and head position.

Electric signals from the vestibule are collected by the vestibular branch of the eighth nerve and sent to the vestibular nuclei in the bulb. At these levels, they are connected with the bulbo-reticulo-mesencephalic nuclei that account for reflexes: postural, antigravitational, rebounce, and strengthening of pyramidal activity when walking.

Summary

The ear consists of three main elements: external, middle, and internal ear. The anatomy of the external ear is sculptured to collect, amplify, and direct sound towards the tympanic membrane. The architecture of the middle ear (tympanic membrane, ossicular chain, air reservoir, and intermittent pressure equalizer) has evolved to modulate and transmit important sound frequencies towards the inner ear. The cochlea is specialized in transforming and selecting a specific sound vibration corresponding to a specific frequency in an electric signal. This will travel through the acoustic component of the eighth nerve towards auditory centers in the brainstem. The signal will be sent to the cortex for actual signal interpretation and processing, and thus an actual sound sensation appears.

Chapter 2 - Etiopathology of Hearing Loss and Deafness

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"Our children should be properly introduced to the world in which they live." Thomas Berry

Introduction

Congenital hearing loss, without early diagnosis and treatment, can cause negative effects on speech, language, educational, and cognitive development in children. If the implantation is done in a timely manner, and the rehabilitation is done according to the rules, we will finally have an almost normal child from an auditory and verbal point of view, ready to attend stream school. That is why it is important to know the causes that can determine hearing loss. New genetic mutations responsible for hearing loss are discovered every year. More than 100 loci have been identified that include genes encoding proteins involved in the structure and function of hair cells in the inner ear, supporting cells, spiral ligament, vascular striae, basilar membrane, spiral ganglion cells, and auditory nerve. Recent studies have identified dysfunctional proteins that affect calcium and potassium homeostasis, with impaired apoptotic signaling, mechanoelectrical transduction, and electromotility.

Due to the risk of hearing loss after chemotherapy therapy for cancer, do you think that this should be taken into account when performing chemotherapy?

Sensorineural Hearing Loss

Sensorineural hearing loss can be classified as genetic or acquired. Genetic causes can be syndromic or nonsyndromic. They may have autosomal (recessive or dominant), gonosomal or mitochondrial transmission.

1. Genetic Causes

1.1 Non-Syndromic autosomal recessive hearing loss

More than 700 different mutations have been identified in one of 42 genes in individuals with non-syndromic autosomal recessive hearing loss. Mutations reported in GJB2 (Gap Junction Protein Beta-2), which encodes connexin 26, make this gene the most common cause of hearing loss in many populations. Other relatively common deafness genes include SLC26A4, MYO15A, OTOF, TMC1, CDH23 and TMPRSS3 [1].

1.2 Syndromic hearing loss with autosomal recessive transmission

In this case, lesions are present at birth and can range from submicroscopic to total cochlear agenesis.

Usher syndrome is the leading genetic cause in which both hearing and visual loss are present: sensorineural hearing loss, retinitis pigmentosa, and occasionally balance disorders. The syndrome is characterized by genetic heterogeneity, with 12 independent loci with 9 known genes having been identified and having chromosomal localizations of the determinant genes at: 14q32, 11q (myosin), 11p, 1q and other loci [2]. Histopathological examination shows atrophy of the Corti organ, spiral ganglion in the basal tour of the cochlea or at the level of the entire cochlea, vascular striae, limbus and tectorial and Reissner membranes. The electron microscopy shows the absence of supporting cell elements and dendrites in the Corti organ [3].

Pendred syndrome is caused by biallelic mutations in the SLC26A4/PDS gene and is associated with bilateral sensorineural hearing loss, mental retardation, goiter, and abnormal iodine organization with or without hypothyroidism [4, 5]. Malformations of the inner ear counted in an enlarged vestibular aqueduct or cochlear hypoplasia, the cochlea having only one and a half rounds. If a patient has vestibular enlargement of the aqueduct associated with cochlear hypoplasia, this is called a Mondini malformation [6]. Pendred syndrome accounts for up to 10% of cases of congenital deafness and is the most common cause of syndromic deafness [7].

Jervell and Lange-Nielsen syndrome is characterized by: profound bilateral sensorineural hearing loss, prolonged QTc interval (more than 500 msec), which can lead to peak torsades, syncope seizures, and sudden cardiac death in childhood [8]. Most cases are due to deletion mutations in the KCNQ1 and KCNE1 genes, which encode essential proteins for the functions of the heart and cochlea potassium channels [9].

Cockayne syndrome has a multisystemic impairment, and from a genetic point of view, it has been classified mainly into 2 different groups: Cockayne syndrome A, which occurs due to a mutation on ERCC8 on chromosome 5q12 - q31, and Cockayne syndrome B due to a mutation on ERCC6 on chromosome 10q11 [10].

Refsum disease, also known as “phytanic acid storage disease” or “atactic polyneuritiform heredopathy”, is a peroxisomal disorder caused by altered alpha oxidation of branched chain fatty acids, resulting in the accumulation of phytanic acid and its derivatives in cells and tissues. This may be due to deficiencies in phytanoyl-CoA hydroxylase or peroxin-7 activity [11]. The disease is associated with atypical retinitis pigmentosa, cataracts, cerebellar ataxia, peripheral neuropathy, congenital ichthyosis, cardiomyopathy, and progressive sensorineural hearing loss.

Bjornstad syndrome is due to mutations in the BCS1L gene and is characterized by twisted hair (flattening and abnormal twisting of hair around their axis and the absence of any pigment in the hair) and neurosensory hearing loss without intellectual impairment [12].

1.3 Syndromic hearing loss with autosomal dominant transmission—deafness that can evolve progressively throughout the patient's life without a constant evolution.

Waardenburg syndrome has mutations in the following genes: EDN3, EDNRB, MITF, PAX3, SNAI2, and SOX10. The PAX3 gene is located on chromosome 2 and encodes transcription factors. It is responsible for the development of the facies and inner ear and is involved in the onset of Waaldenburg type I and III

syndrome. MITF is located on chromosome 3, is responsible for the development of the ear and hearing and regulates the differentiation and development of the melanocyte of the retinal pigment epithelium. Together with SNAI2, which is located on chromosome 8, it is found in Waardenburg syndrome type II. EDRNB/EDN3 is involved in Waardenburg type IV syndrome. The gene is located on chromosome 20 and encodes the endothelin protein that is part of the vessels [13,14].

Crouzon's disease or Crouzon's craniofacial dysostosis results from a mutation in the gene on chromosome 10, the long arm, locus 26 (10p26) encoding fibroblast growth factor receptor 2 (FGFR2) and the 4p16.3 locus gene on chromosome 4 [15]. The disease has the following features: divergent strabismus, exophthalmos, hypertelorism, maxillary hypoplasia, short upper lip, deformities of the sagittal suture and bregma, nose in the "parrot's beak" and deafness.

Tietz syndrome is the result of a mutation in the MITF gene and is characterized by: albinism, deafness, lack of eyebrows and blue iris [3, 16].

Branchio-oto-renal syndrome is due to mutations in the EYA1, SIX1 and SIX5 genes and is associated with: preauricular fistulas, vestibular and auricular abnormalities, transmissible deafness, sensorineural or mixed hearing loss, cervical brachial cysts, renal canal malformations and stenosis [3, 17].

Marfan syndrome is a mutation in the FBN1 gene. Within this syndrome we find: cardiovascular abnormalities, muscular hypotonia, joint laxity, scoliosis, arachnodactyly, flatulence, ectopia lentis and conductive or sensorineural hearing loss [3].

DiGeorge Syndrome or Deletion Syndrome 22q11.2 is one of the most common human microdeletion syndromes. In infants, DiGeorge's syndrome usually has a triad: immunodeficiency, congenital heart abnormalities, and hypocalcemia due to hypoparathyroidism. Hearing function may be normal or sensorineural, conductive, or mixed hearing loss may occur, mild, moderate, severe, or profound, depending on the severity and location of the abnormalities (middle or inner ear) [3, 18].**1.4 Syndromic hearing loss with the possibility of both autosomal dominant and recessive transmission**

Treacher-Collins Syndrome or **Mandibulo-Facial Dysostosis** - is the result of mutations in the TCOF1, POLR1C, POLR1D genes. When the TCOF1 and POLR1D genes are involved, the mode of transmission is autosomal dominant, and when the POLR1C gene is affected, it is autosomal recessive [20]. Within this syndrome we find the following anatomical changes: retracted mandible, prognosis of the upper jaw, deletion of the glabella, antimongoloid eyelid slit, thin lower eyelid, atrophic, with absence of orbicularis muscle and tear point, abnormalities in the development of atrial or hearing loss or hearing loss [19].

1.5 Syndromic hearing loss with the possibility of autosomal dominant, recessive transmission, or X-linked transmission.

Alport syndrome, also known as hereditary nephritis, is characterized by: sensorineural hearing loss on high frequencies (absence of cochlear neurons, hair cells, and changes in the vascular stria), kidney damage (hemorrhagic nephritis), cataract, anterior lenticonus, and corneal ulcers. It may be associated with skeletal abnormalities, central nervous system abnormalities, and skin abnormalities. This syndrome has three genetic forms: XLAS (mutations in the gene encoding the alpha 5 chain in type IV collagen – X linked transmission); ARAS (mutations in the alpha 3 or alpha 4 chain – autosomal recessive transmission); and ADAS (mutations in the gene encoding the alpha 3 or alpha 4 chain – autosomal dominant transmission) [16,20].

1.6 Norrie syndrome—oculo-acoustic-cerebral degeneration—is a rare X-linked recessive genetic disorder that affects the male sex. It is characterized by congenital blindness accompanied by mental retardation and deafness. At least 30-50% of reported cases have varying degrees of intellectual disability with psychotic features, and about 25% develop progressive neurosensory hearing loss, beginning in the second or third decade of life [21, 22].**Deafness-associated genetic syndromes in which transmission is uncertain or spontaneous or the pattern of transmission is irregular**.

Pierre Robin syndrome represents a mutation on chromosomes 2, 4, 11, or 17 or can be caused by intrauterine compression of fetal mandible, and is characterized by: staphylosis, glossoptosis, and micrognathia. These can be associated with: ear, ocular, cardiac, vascular, and skeletal abnormalities. Ear disorders include malformations of the common arm of the posterior and vertical semicircular canals, modiolus, undeveloped common scale, hypoplastic internal auditory canal, and stapedial abnormalities [17].

Wildervanck syndrome or cervico-oculo-acoustic dysplasia is characterized by: congenital fusion of two or more cervical vertebrae, Duane retraction syndrome (retraction of the eyeballs and impaired abduction), deafness (deformity and ankylosis of the armpits, rudimentary inner ear, presence of a single incompletely developed semicircular canal). They can also be associated with: spina bifida, facial asymmetry, torticollis, cervical ribs, meningocele, mental retardation, cardiovascular and pulmonary abnormalities [18].

Nager syndrome—acrofacial dysostosis—is a rare disease that involves severe micrognathia and shortening of the upper limbs. The transmission can be autosomal dominant or autosomal recessive. It is usually confused with Treacher Collins Syndrome, with which it shares the same craniofacial phenotype. However, patients with Treacher Collins syndrome do not have hand abnormalities, which are seen in patients with Nager syndrome [23]. Recent studies have identified mutations in the SF3B4 gene (binding factor 3b, subunit 4) as the cause of Nager syndrome in approximately 60% of affected individuals, which encode a component of the pre-mRNA spliceosomal complex [23,24].

Potter's syndrome is characterized by pulmonary and renal dysplasia, hypoplastic mandible, bilateral cataract and lower inserted pavilions, and malformations of the middle ear. It is incompatible with life because newborns with Potter syndrome have pulmonary hypoplasia that leads to respiratory distress within one hour of birth [25].

CHARGE syndrome is due to mutations in the CHD7 gene, with a broad phenotype that can involve almost all organs and sensory systems, which causes a large and highly variable comorbidity [25].

The name CHARGE stands for:
C = coloboma and cranial nerve defects
H = heart defects, especially tetralogy of Fallot
A = atresia of the choanae (blocked nasal breathing passages)
R = retardation of growth and development
G = genital underdevelopment due to hypogonadotropic hypogonadism
E = ear abnormalities and sensorineural hearing loss

Atrial malformations are represented by: atrial hypoplasia, implanted lower ears, malformations of the hammer, anvil and ladder, hypoplasia of the eardrum, poor mastoid pneumatics, vascular abnormalities,

stenosis of the oval window and/or round cochlear malformation Mondini, hypoplasia or aplasia of the semicircular canals [25].

Goldenhar syndrome or oculo-atrial-vertebral dysplasia is a rare congenital disease that occurs as a result of the abnormal development of the first two brachial arches. Etiopathogenesis is considered multifactorial but is not yet fully established and involves genetic and environmental factors. In sporadic cases, 5p deletions, 14q23.1 duplications or chromosome 18 and 22 abnormalities were observed [27]. The patient clinically has the following malformations: atrial and facial malformations, vertebral abnormalities, laryngomalacia, vertebral abnormalities, cardiac abnormalities, of the ocular central nervous system. In Goldenhar syndrome we can encounter the following otic malformations: microtia, atresia of the external auditory canal, hammer, anvil, and ladder may be malformed or missing, mastoid is poorly pneumatized, facial nerve has a vicious trajectory, developmental abnormalities of the bone labyrinth and small size of the internal auditory canal [3].

VATER (VACTERL) syndrome

The name VATER stands for:

- **V**ertebrae, in which the bones of your spine are missing or irregular
- **A**nus, where you have a blocked anus called imperforate anus, or an anus that doesn't open to the outside of your body, called anal atresia
- **T**rachea, where you have an irregular connection between your trachea and esophagus called a fistula
- **E**sophagus, where your esophagus doesn't have an opening, called esophageal atresia
- **R**enal, defects that affect your kidneys

Studies have found that the syndrome is associated with decreased motor and sensory sensitivity of the facial nerve, enlargement of the duct and endolymph sac, and minor malformations of the ladder and vestibular system [3].

Congenital hypothyroidism is classified according to etiology and symptomatology into four categories [3]:

- athyreotic cretinism—is associated in over 40% with neurosensory, conductive or mixed hearing loss;
- family goiter - associates: family goiter, neurosensory hearing loss and abnormal perchlorate test;
- endemic cretinism—has mental retardation, spasticity, motor dysfunction and frequently mixed hearing loss;
- childhood goiter hypothyroidism—in this case, no hearing loss occurs.

1.7. Syndromic hearing loss with mitochondrial transmission

- **Kearns-Sayre syndrome,** or oculo-cranio-somatic syndrome, is a rare mitochondrial DNA deletion with multisystem damage. The first symptom is unilateral ptosis, that progresses to a bilateral ptosis. Deafness is due to cochleo-saccular degeneration [18].

Inner Ear Malformations

According to the literature, malformations of the inner ear are classified in [28]:

1. Complete labyrinthine aplasia which includes the absence of the cochlea, vestibule, semicircular canals, and vestibular and cochlear aqueducts:
   - complete labyrinthine aplasia with hypoplasia or petrosal bone aplasia;
   - complete labyrinthine aplasia without otic capsule;
   - complete labyrinthine aplasia with otic capsule;

2. Rudimentary otocyst of a few millimeters without the formation of the internal auditory canal;

3. Cochlear aplasia:
   - cochlear aplasia with normal labyrinth, vestibule, and semicircular canals are normally developed;
   - cochlear aplasia with dilated vestibule;

4. Common cavity - is defined as a single chamber, with round or ovoid shape, representing the cochlea and the vestibule. Theoretically, it has cochlear and vestibular neural structures. The semicircular canals may be present or may have a rudimentary appearance.

5. Cochlear hypoplasia:
   - the cochlea has the appearance of a small bud with round or oval shape, resulting from the internal auditory canal (bud-like cochlea);
   - cystic hypoplastic cochlea—in this case the cochlea is smaller, with defective modiolus and interscalar septa, but with a normal external appearance of the contour. There may be a complete absence of the modiolus, creating a wide connection with the internal auditory canal, making it possible to release and move the electrode into the internal auditory canal. The vestibular aqueduct can be enlarged, and the vestibule may be dilated.
   - the cochlea has less than 2 turns of spiral, but with external and internal contours similar to those of a normal cochlea. The vestibule and semicircular canals are usually hypoplastic;
   - the cochlea has the basal tour of normal appearance, but the middle tour and the apex are hypoplastic. We often find the labyrinthine segment of the anterior cochlear facial nerve.

6. Incomplete division of the cochlea
   - type I or cystic cochleovestibular malformation: in this anomaly, there is a clear differentiation between the cochlea and the vestibule. The cochlea is situated in its typical location and does not have the entire modiolus and the interscalar septum, giving the appearance of an empty cystic structure. The external dimensions (height and length) are similar to those of a normal cochlea. The cochlea is accompanied by an enlarged and dilated vestibule.
   - type II - the apical part of the modiolus presents anomalies giving the cochlea a cystic appearance.
   - type III - the cochlea has interscalar septum, but the modiolus is absent.

7. Enlarged vestibular aqueduct with cochlea, normal vestibule, and semicircular canals.

8. Cochlear opening abnormalities.

2. Acquired Causes

2.1 Viral infections: cytomegalovirus, rubella, lymphocytic choriomeningitis virus, human immunodeficiency virus, measles, varicella zoster virus, mumps, West Nile virus.

Viruses can cause hearing loss by infecting the fetus during pregnancy (congenital hearing loss), either as a result of infection during childhood or adulthood (acquired hearing loss). Hearing loss is due to damage to the ear (significant damage to the organ Corti, stria vascularis, cochlear nuclei) or autoimmune mechanism (cytomegalovirus) [29].

2.2 Ototoxic – which can be determined by [29]:

Medications: aminoglycoside antibiotics (streptomycin, gentamicin, kanamycin, tobramycin, amikacin), vancomycin, loop diuretics (furosemide, ethacrynic acid), cytostatic agents (cisplatin, cyclophosphamide, methotrexate), salicylate, quinine, thalidomide, isotretinoin, diphenylhydantoin.

Industrial substances: heavy metals (arsenic, mercury, lead), solvents (amino and nitrobenzenes), carbon monoxide, organosulfur compounds, carbon tetrachloride.

Psychotropic substances: cocaine, heroin, tobacco, alcohol. Once administered, ototoxics pass the blood-labyrinth barrier and through the Reissner's membrane enter the endolymph where they reach the neurosensory cells where they often produce cell apoptosis [30].

2.3 Causes which may appear during pregnancy: prematurity, prolonged pregnancy, low birth weight, hyperbilirubinemia at birth, prolonged labor, cesarean delivery, placenta previa, pregnancy toxemia, neonatal asphyxia, use of sedatives or anesthetics during labor.

2.4 Bacterial infectious pathology of the middle and inner ear

Infections in the middle ear are responsible for pure conductive hearing loss. In the absence of treatment, they can cause cochlear degeneration (the onset of sensorineural hearing loss) by the degeneration of ciliated sensory cells and the Corti organ and the destruction of the ion pump and Reissner's membrane. The mechanisms responsible for cochlearization are: perilymphatic fistula, Wallerian neural degeneration, thrombosis, or sclerosis of vascular striae, intracochlear hemorrhage [29].

2.5 Perilymphatic fistula - can be congenital (Mondini dysplasia), spontaneous, or acquired (temporal bone fractures, stapedectomy, barotrauma). They can communicate with the subarachnoid space [29].

2.6 Sound trauma - causes damage to the cochlea in case of exposure to a very high intensity of sound, either unique and impulsive (acute sound trauma), or prolonged, due to the profession (chronic sound trauma). In the case of chronic sound trauma, at the beginning, the effects of noise on the ears determine the auditory adaptation, then the auditory fatigue appears, and at the end the hearing loss is installed due to the permanent cellular alteration of degenerative or reparative nature [3].

2.7 Hematological diseases cause neurosensory hearing loss through ischemia that settles in the inner ear [29].

2.8 Autoimmune diseases can be systemic (Wegener's granulomatosis, systemic lupus erythematosus, Sjogren's syndrome, multiple sclerosis, sarcoidosis), or localized (Meniere's disease, Cogan's syndrome, cochlear vasculitis). But only 15-30% of the patients clinically experience sensorineural hearing loss

resulting from immune responses generated by the inner ear, which damage the cochlea and spinal ganglion [31].

2.9 Paget's disease has the mechanism of decreasing bone intensity in the ossicular system or cochlea [28].

2.10 Diabetes may cause hearing loss and balance disorders in patients due to poor use of glucose. In diabetes, the vascular walls of the vascular membrane and stria vascularis are thickened, which will lead to endothelial dysfunction [32]. Cochleo-vestibular disorders in diabetic patients are favored by angiopathy and diabetic neuropathy [29].

2.11 Tumors that cause hearing loss are: atrial-meatal tumors, benign cavotympanic and antro-mastoid tumors (facial neurofibroma, solitary neurofibroma belonging to the Jacobson tympanic nerve, osteoclastoma, osteoma, adenocortistoma), malignant tumors of the ear (epithelium, sarcoma) or secondary (enlargement of a tumor in the adjacent areas of the nasopharynx, skull base, parotid gland), tumors located in the internal auditory canal or pontocerebellar angle (vestibular schwannoma, dermoid cysts, arachnoid cysts, meningiomas), extensive basal skull tumor (chondrosarcoma, rhabdomyosarcoma, hemangioma) [3].

2.12 Radiation therapy for head and neck cancers performed at doses greater than 45 Gy may be responsible for damage to the auditory nerve and brainstem (29).

2.13 Trauma. Trauma may result in labyrinthitis, perilymphatic fistulas, damage to the cochlear nerve, damage to the myringo-ossicular system, stapediovestibular joint dislocation or subluxation, fractures of the cavum tympani, rupture of labyrinthine windows (3). Traumas of the skull base represent 45% of the craniocerebral fractures and over half of them affect the temporal bone, resulting in longitudinal, transversal, oblique, atypical fractures. The fractures are often indirect, splitting into continuously irradiated fractures, from the vault or base, and independent, ipsilateral, contralateral, or bilateral fractures [3].

Conductive Hearing Loss

The causes of conductive hearing loss can be classified into (3):

1. **By meatal blockage**: agenesis or stenosis of the external auditory canal complete the posttraumatic or postotitic sequelae scar

2. **By myringeal damage**: myringeal blockage, perforation of the tympanic membrane

3. **By ossiculo-cavotympanic injury**: ossicular blockage (fixed malleus syndrome due to malformative, tympanosclerotic or posttraumatic cause, posttraumatic, congenital, tympanosclerotic incudo-mallear fixation, genetic stapedo-vestibular ankylosis syndrome, or tympanosclerotic)

4. **Through tubal dysfunction**

5. **By window blockage**: osiculosclerotic, malformative, tympanosclerotic

Summary

There are two important classes of causes that can cause sensorineural hearing loss: genetic causes and acquired causes.

The genetic causes of hearing loss are highly heterogeneous. Hereditary causes are hereditary congenital diseases with autosomal recessive and dominant transmission and deafness-associated genetic syndromes in which transmission is uncertain or spontaneous or the pattern of transmission is irregular. Mutations reported in GJB2 (Gap Junction Protein Beta-2), which encodes connexin 26, make this gene the most common cause of hearing loss in many populations.

Genetic studies have greatly increased understanding of inner ear functions at the molecular level and provide important information for patients. Personalized and accurate genetic counseling is becoming more common. Prevention of genetic hearing loss is feasible through pre-pregnancy and prenatal diagnosis and genetic counseling.

Acquired causes are viral infections, ototoxic (medications, industrial and psychotropic substances), causes of intrapartum, bacterial infectious pathology of the middle and inner ear, perilymphatic fistula, sound trauma, hematological diseases, autoimmune diseases, Paget's disease, diabetes, tumors that cause hearing loss, radiation therapy and trauma.

It is important that hearing loss be diagnosed as soon as possible, and that hearing rehabilitation be done as early as possible.

Chapter 3 - Treatment of Hearing Loss

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"Blindness separates us from things, but deafness separates us from people" Hellen Keller

Introduction

Normal hearing at birth is one of the prerequisite conditions for speech acquisition.

Hearing does not have a fixed or absolute value, but rather is situated at different values in normal hearing parameters. The auditory threshold is represented by the weakest sound stimulus that can be perceived by the human ear. Hearing loss is defined by an auditory threshold shift with a decrease in hearing sensitivity.

Disabling hearing loss, based on 42 population studies, has a prevalence of 5.3% worldwide; statistically, 91% of those with hearing loss are adults and 9% are children [1,2]. According to the WHO, disabling hearing loss is defined as an increase of the auditory threshold in the better hearing ear of more than 40 dB in adults (patients over 15 years of age) and of more than 30 dB in children (patients under 15 years of age). Due to the nature of prelingual hearing loss, children have delayed language development, cognitive impairment, and low school performance [3]. In adults, hearing loss can lead to social isolation and difficulties in maintaining a job [4]. Diagnosis of hearing loss requires collaboration between the ENT physician, audiologist, geneticist, ophthalmologist, and pediatrician. The patient requires a thorough clinical examination, audiometry, CT scan, MRI, cytomegalovirus screening, molecular genetics tests, and so on [5,6]. In this context, early identification, and treatment of deafness (the most frequent sensorial pathology encountered at birth – 1-3‰ of newborns), represents one of the most important preoccupations for the public health policy in the developed countries.

Neuroscience research indicates optimal periods for auditory development and, consequently, the importance of maximizing early hearing experience by early hearing loss treatment. Nowadays, speech development reports show better results for infants with congenital deafness treated very early, bringing evidence that deaf children under 12 months benefit most from cochlear implantation [7].

Early intervention provides better language development due to maximum cortical plasticity in the early years of life with immediate and late consequences, which is reflected in the cognitive-behavioral, social, and daily activities.

How can we prevent hearing loss?
How can we convince parents of the necessity of the early treatment?

What treatments are available for hearing loss?

Hearing loss can be sensorineural (SNHL), conductive (CHL) or mixed (MHL) - unilateral or bilateral; regarding the degree of hearing loss, it can be mild (26-40 dB), moderate (up to 60 dB), severe (up to 80 dB), or profound (81 dB or greater) [8]. The treatment of hearing loss is dependent on multiple factors. First of all, the site of the lesion (external, middle, inner ear, and/or auditory nerve) is important and also the cause, type, and the severity of hearing loss. A team comprised of an otorhinolaryngologist surgeon, an audiologist, and a speech therapist decides on the appropriate treatment on a case-by-case basis.

Treatment of conductive hearing loss is focused on the cause, such as removal of the foreign body, or of the cerumen from external auditory meatus. In case of congenital atresia of the external auditory canal surgery can be attempted in accordance with the Jahrsdoerfer grading scale score. In order to recommend surgery, the score may be higher than five [9]. A score of 5 or less disqualifies the patient for aural atresia surgery. In cases of mucous otitis media, myringotomy followed by tube placement to ensure the drainage and ventilation of the middle ear until the maturation of the pharyngotympanic tube function. In otosclerosis/otospongiosis – stapedotomy and stapes replacement with a special piston can be the solution. When the cause of the hearing loss is a cholesteatoma, then surgical removal is necessarily followed by hearing restoration (ossiculoplasty) depending upon the degree of destruction of the middle ear structures. Inoperable, conductive hearing loss can be treated with conventional hearing aids or bone-conduction hearing aids, depending on the case's particularities.

Treatment of sensorineural hearing loss - Ninety percent of hearing losses are sensorineural, and surgical and medical corrections are much more challenging than in conductive hearing loss.

What are conventional hearing aids?

Conventional hearing aids are electronic devices that are worn in or behind the ear (see figure); they amplify, with good results, sounds up to 40 to 60 dB.

Choosing a conventional hearing aid depends on the lifestyle, budget, and hearing loss degree. It is also important to consider what features would be most beneficial - from directional microphones to waterproof options, there are numerous varieties to meet everyone's personal needs. Hearing aids are even equipped with Bluetooth connectivity to work with wireless technology like a cell phone or television.

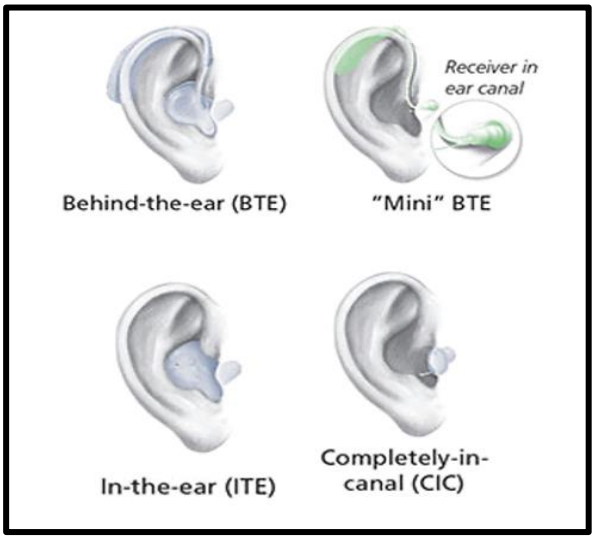

Figure 1 Styles of hearing aids (Source: NIH/NIDCD)

They are considered sleek, compact, and innovative – offering solutions to a wide range of hearing aid users. Typically, a hearing aid contains a microphone, an amplifier, and a speaker. The microphone picks up the sound waves and converts them into electrical signals, which are then processed, amplified, and then sent to the speaker, which converts them back into sound waves, providing a louder and clearer sound.

There are four basic types of hearing aids: behind-the-ear (BTE), receiver-in-canal (RIC), in-the-ear (ITE) hearing aids, and completely-in-canal (CIC) (Figure 1). Regarding behind-the-ear (BTE) hearing aids—all tech components are stored behind the ear in a casing that connects to the ear mold via a thin tube. Receiver-in-canal (RIC) hearing aids are also worn behind the ear but connect via a form-fitted wire to a small speaker that sits in the ear canal. In-the-ear (ITE) hearing aids are nearly hidden inside the ear canal with no components behind the ear. Completely-in-canal (CIC) hearing aids, which are custom-made, are housed entirely inside the ear canal. To find the proper hearing aid it is important to try more than one and to benefit from a trial period.

What are cochlear implants?

Since December 2019, about 736,900 registered cochlear implants have been implanted worldwide. In the United States, roughly 118,100 devices have been implanted in adults and 65,000 in children. (Estimates provided by the U.S. Food and Drug Administration [FDA], as reported by cochlear implant manufacturers approved for the U.S. market).

The success story of the congenitally, profoundly deaf children who have overcome deafness began in the early 1960s when doctor William House learned that his colleague from France, doctor Charles Eyres, together with the electrophysiologist Andre Djourno had succeeded in 1957 in producing a sensation of hearing by placing an electrode in contact with the eighth nerve in a patient operated for a huge cholesteatoma that destroyed the inner ear [10]. Based on the French experiments, doctor House began his work in the field of cochlear implantation. It was in 1961 when, together with Jim Doyle, he implanted the first ever cochlear implant (CI) with one channel in Los Angeles [11]. Later, some other teams, especially from Germany and the US, joined the CI research. It was the experimental period of CI. The second period began in 1970 and was the trial period when a cohort of patients were cochlear implanted [12]. In 1973, during the first conference on electrical stimulation of the acoustic nerve as a treatment for the profound sensorineural deafness in human, organized in San Francisco, the term “cochlear implant” was introduced in medical literature. In the same period the multichannel single wire electrode was initiated by Clark in 1978 [13] and Simmons and White in 1979 (14). Their studies launched the third period - that of commercialization. In this third stage, other teams from Germany, Belgium, and Austria started their experiments [15,16].

A cochlear implant is a small, complex electronic partial implantable device that can help to provide a sense of sound to a person who is profoundly deaf.

An implant consists of an external part and an internal part. The external part is composed of a microphone, a speech processor, a battery housing, and a transmitter. The implantable part consisted of a receiver/stimulator, an internal coil, and an electrode array. The microphone picks up the environmental sounds. The sounds are prepared by the sound processor and the resulting information is transferred via the antenna to the receiver/stimulator which converts it into an electric impulse. From the receiver/stimulator, the stimulus travels through the electrode array to the neural elements by passing

through the cochlea. An implant does not restore normal hearing but helps the recipient to understand speech and to be aware of environmental sounds.

Are there any differences between a cochlear implant and a hearing aid?

A cochlear implant is very different from a hearing aid. Hearing aids amplify sounds so they may be perceived by people that have moderate hearing loss. Cochlear implants bypass the inner ear and directly stimulate the auditory nerve. So, cochlear implants are indicated for those who have no benefit from hearing aids because the cochlea is so damaged and no longer useful. The auditory nerve sends the signals further to the brain, which interprets them as sound. Hearing through a cochlear implant is different from normal hearing and takes time to interpret the means of it. Recipients of cochlear implants recognize different sounds in the environment and understand speech even over the telephone.

Which are the indications for cochlear implantation?

Today, cochlear implantation is continuously changing its indications and is attempting to improve the way a patient hears as compared to a conventional hearing aid. More authors recommend a personalized evaluation of every case, establishing: the type and localization of the lesion that led to hearing loss, the presence of some other comorbidities, the level of cognition, motivation, family support, individual expectations, etiology of deafness, the morphology and size of the cochlea, residual hearing and so on. Audiological parameters are of the utmost importance for cochlear implantation indications. According to the FDA, adult candidates have to have an average hearing loss of 70 dB or more, bilaterally (both ears), on frequencies of 500-1,000-2,000 and 4,000 Hz, the tonal auditory threshold in free field with hearing aids greater than 55 dB on the same frequencies, and disyllabic discrimination value of less than 40% on the speech audiogram in free field with hearing aids at an intensity of 65 dB (the hearing aids have to be worn a minimum of 6 months before cochlear implantation approval).

The audiological criteria in children are different from those for adults because behavioral tests are difficult to be done and interpret and due to a lack of tests that can examine the entire range of frequencies. In this case, the FDA recommendations for children that are candidates for cochlear implantation are the following: SNHL, when the average auditory threshold is calculated on frequencies 500, 1000, 2000, and 4000 Hz and is more than or equal to 90dB [17].

Furthermore, from the possible candidates for cochlear implantation, those with acute or chronic inflammation of the middle or external ear are temporarily excluded. Once the inflammatory process heals, the patient can be considered for cochlear implantation. Other contraindications for cochlear implantation are: cochlear aplasia, cochlear nerve agenesis, cochlear ossification [18]. Candidates for cochlear implantation undergo rigorous examination to determine the cause of hearing loss and to establish the extent to which it may hinder cochlear implantation, for example: a) In temporal bone fractures, the facial nerve could be stimulated during implant activation and the stimulation parameters would need to be modified; b) Fibrosis or ossification of the cochlear duct after meningitis changes the procedure of cochlear implantation; c) Patients with neurofibromatosis type II and those with cochlear nerve aplasia may benefit by a brainstem implant. Age is not an impediment for cochlear implantation. Currently, patients younger than 12 months old and as old as 84 to 86 years old have been implanted; the usual upper age limit is 95 years [19].

Indications for cochlear implants in children have changed over the years as well [20][21]. Until the 1990's, only adults were accepted as candidates for cochlear implantation, but now, children have a higher priority and are the favored candidates. The auditory system and the child's brain are genetically preconfigured, but their development is modulated by the external environment being either restrictive or permissive. Speech development starts at birth and is almost completely developed by the time the child reaches the age of 6 years. Due to this fact, children with congenital deafness need to be implanted when cerebral plasticity is at its greatest. After the ages of 5 to 8 years, the chances of speech development decrease significantly [22,23]. Until 2000, the smallest recommended age for implantation was 2 years, and after 2000, the FDA approved cochlear implantation starting with the age of 12 months. Implantation in younger infants is now accepted, although the audiological and anesthesiologic implications are still in debate. In hearing loss due to meningitis, cochlear implantation is considered an emergency [24]. The success of rehabilitation after cochlear implantation is strong corelated with a good familial and community support as well as a high level of maternal education. The remarkable progress made in implant technology led to the implementation of certain selection criteria for the candidates and thus, some of the patients that would not have been able to benefit from a cochlear implant years ago can now be accepted as valid candidates for cochlear implantation, according to the literature.

The current indications refer to bilateral cochlear implantation, especially in cases of SNHL post-meningitis or in cases of concomitant blindness. Bilateral cochlear implantation improves sound localization and speech discrimination in noise [25]. It is a delicate decision whether to bilaterally implant a child simultaneously or sequentially at a certain interval between the two. The preferred approach has still not been established [26, 27].

Cochlear implantation in unilateral deafness has been gaining ground recently. Studies in which cochlear implants are compared to the traditional hearing aids used for unilateral hearing loss (CROSS or IAO) show that cochlear implants are more efficient [28]. Moreover, in most studies on unilateral hearing loss with tinnitus, electrical stimulation plays an important role as a tinnitus reliever. In most cases, tinnitus is suppressed (in 46-95% of cases) with only 5.6% of cases reporting an aggravation of tinnitus after electrical stimulation [29,30].

Auditory neuropathy (AN) is a group of disorders that have an incidence of 2.4-15% in children diagnosed with SNHL [21,31]. A study comprising 260 children diagnosed with AN showed that 5% of these children developed normal speech without the use of hearing aids or other auditory prostheses [32]. The results of children with AN who were implanted were variable [33].

Implantable Prosthesis for the Middle Ear

Bone-active implants (BAI) require an external energy source which transforms input signals into mechanical energy that is transmitted directly to the ossicular chain or to the cochlea [34]. BAI's can be totally or partially implantable, and these different types use different forms of transductors to convert electrical stimulus into mechanical energy. This is represented by piezoelectric or electromagnetic vibrations with their specific electromechanical variation [35]. The main indications for BAI are as follows: moderate to severe SNHL, - vocal audiogram with disyllabic words with 50% or better.

Regarding VSB, a transducer (FMT) has to be placed in connection with the ossicular chain (traditionally coupled to the long process of the incus, but new couplers have been developed that enable the FMT to be coupled to the short process of the incus) in both cases an anatomically normal middle ear is mandatory

[36]. The main indications are for patients who cannot wear external earmolds that come with conventional hearing aids (due to atresia or stenosis of the external ear canal, chronic otitis externa or an allergy to the material the earmolds are made from), and for patients who have acoustic feedback. Every type of implantable device has its own specifications and specific auditory criteria. Benefits of BAI's are: correct sound amplification, no acoustic feedback, speech discrimination, and no foreign body in the external ear canal [36][37].

Bone-Conduction Auditory Implants (Baha/Bonebridge/Osia) Bone-conduction implants transmit sound through the bone to the cochlea through direct stimulation of the skull [35]. Indications for this type of hearing device are: mixed hearing loss or conductive hearing loss in which ossicular reconstruction was not possible; radical open-cavity mastoidectomy; atresia or stenosis of the external ear canal or patients who cannot wear hearing aids for other medical reasons or who have not achieved sufficient benefits from other hearing systems and the bone conduction is better than 50-65 dB [37,38]. Another distinct indication is single-sided deafness, in which the auditory information is transmitted from the side of the deaf ear to the contralateral normal ear via skull vibration [35–37,39].

Assistive Listening Devices include telephone and cell phone amplifying devices, smart phone, or tablet "apps," and closed-circuit systems (hearing loop systems) in classrooms and auditoriums. These FM systems are discreet and work well together with the child's hearing aid or cochlear implant, overcoming the poor acoustics with lots of background noise. The speaker has a discreet microphone that transmits the voice directly to the child's hearing aids or cochlear implant.

Summary

The treatment of hearing loss is dependent on multiple factors. First, the site of the lesion is important (external, middle, and/or inner ear), and also the cause, the severity of hearing loss, and the age of the patient. The proper treatment is decided on case-by-case basis, by a team composed by the otorhinolaryngologist surgeon, audiologist, and speech therapist.

Although progress in medical technology has created new opportunities in the treatment of deafness, allowing us to prompt intervention, it is important to emphasize that technology does not "cure" hearing loss but may help a child with hearing loss to make the most of their residual hearing. In the last years the concepts of personalized treatment and of holistic treatment of deaf child have emerged. The new possibilities for deafness treatment, whether conservative, involving the use of assistive listening devices and amplification with powerful conventional digital hearing aids or surgical treatment with implantable prosthesis in cases where medical or surgical treatments could not be done or have failed, or in cases where people do not benefit from conventional hearing aids have become the norm worldwide.

Implantable ear devices for the middle ear (like BAHA - bone anchored hearing aid), for the internal ear (cochlear implant), or for the eighth nerve (brainstem implants), followed by personalized fitting and speech rehabilitation with direct input from all members of the family along with speech therapists and audiologists, are attempting to improve the way a patient hears as compared to a conventional hearing aid.

Chapter 4 - Hearing and Brain – Neurosciences input and evidence

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"The handicap of deafness is not in the ear; it is in the mind".
Marlee Matllin

Introduction

Concas et al. (2021) described hearing as ''One of the most complex mechanisms in humans is the sense of hearing, which has become a cornerstone of our communication, integration, and sociality. At the base of this fundamental ability is the auditory system, an intricate apparatus aimed at converting mechanical soundwaves into electrical impulses that the brain can process.'' In other words, the auditory system gives us the ability to detect and recognize sounds and identify their location.

How does the neural pathway of hearing work?
What parts are involved in the neural pathway of hearing?
What role has attention in general and specifically when listening?

1. Neural pathway of hearing

The two main parts of the auditory system are: the ears and the brain. Specifically, the ear's task is to convert sound waves into neural signals. The brain's task is to receive and process these neural signals and the information they contain. Thus, physiologically, the brain is the true hearing organ because it processes the sounds. By following a sound on its journey into the ear, we can understand how that works. The source of a sound creates vibrations that travel as waves of pressure through particles in air, liquids, or solids [2,3]. These vibrations travel through the ear, contact the tympanic membrane, and travel through the three ossicles that cause depression of the oval window, which then creates waves that travel to the cochlea.

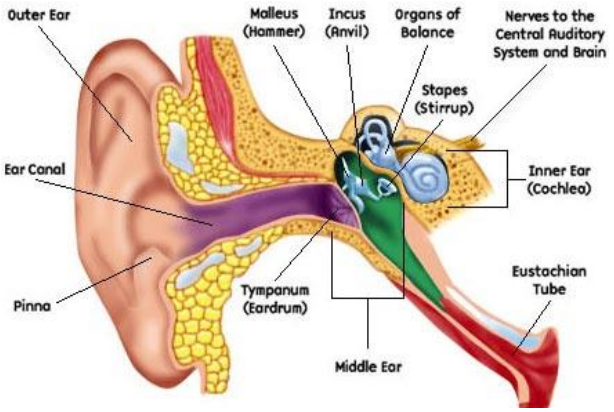

Figure 1: The inner ear. [Connolly, B. 2015]

The cochlea consists of three fluid-filled canals that run parallel to one another; the scala vestibuli, scala media, and scala tympani. The scala media contains a fluid called endolymph. The scala vestibuli and scala tympani contain a fluid called perilymph [5]. So, once the sound vibrations get to the cochlea, they are

converted into vibrations of the fluids in the cochlea, and they travel like a wave from one end of the cochlea to the other. The basilar membrane starts to move as well because of these waves. This membrane is lined with hair cells that have stereocilia which move along with the vibrations. The basilar membrane responds to different frequencies of the sound wave created by the original stimulus, when the sound wave progresses to the particular section of the membrane that responds to that frequency [2,5]. This way, the frequency of sounds can be accurately translated into representative neural activity that can be sent to the brain. The translation into electrical impulses is the currency of signaling that is common throughout the nervous system. The organ of Corti is the receptor organ of the ear. Figure 2 below [6] shows the cochlea and its inner parts. Figure 3 shows the organ of Corti [7]. Thus, the incoming vibration, provoked by the incoming sound, is reinterpreted as an electrical signal.

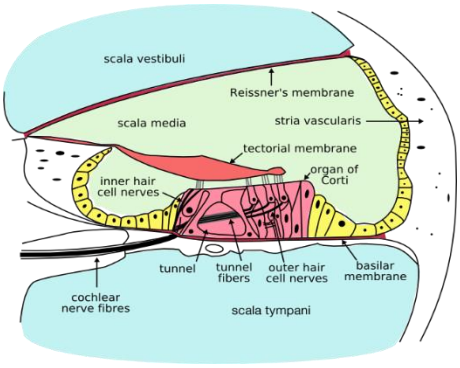

Figure 2: The cochlea and its inner parts [6]

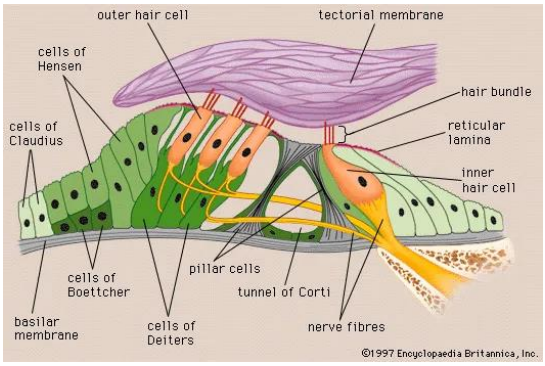

Figure 3: The organ of Corti [7]

The organ of Corti is located on top of the basilar membrane. Naturally, this receptor organ contains receptor cells. These receptor cells are called hair cells, which are named like that because on top of each hair cell, a small collection of small hairs protrudes. These mechanically sensitive organelles are called stereocilia [3,8,9]. When the stereocilia start to move, electrical signals are produced across the membrane trough the action of ion channels which open and cause the release of neurotransmitters. The hair cell excites when the hair bundle displaces towards the top of its taller edge. An inhibitory effect occurs when the motion takes place in the opposite direction [8].

Once the electrical signal has occurred from the organ of Corti, it is transferred to the Vestibulocochlear nerve or N. VIII. This cranial nerve consists of a vestibular and cochlear component. Naturally, their function is to carry information to the brain from the vestibular system and the cochlea. The information coming from the cochlea deals with hearing, while the information coming from the vestibular system deals with vestibular sensations which include information about head position and movement. The cochlear part of the nerve travels from the cochlea to the dorsal and ventral cochlear nuclei, which are found between the pons and medulla. From there, the auditory information is sent to areas in the brainstem trough the cerebellum to the cortex that are involved in auditory processing [10,11,12].

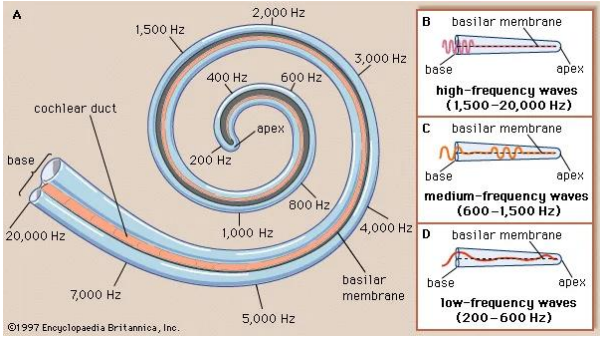

Figure 4. The tonotopy of the Cochlea [7]

As discussed in Chapter 1, the cells responsible for the high-frequency components of speech are located at the base of the basilar membrane. For example, a

voiceless fricative (/s/ or /f/). The cells that are responsible for the low-frequency components are located at the head of the basilar membrane. A vowel with lower-frequency components would activate the cells at the head (image 4). So, a voiced fricative (/z/ & /v/) would stimulate hair cells in both the base and the head of the cochlea [14].

The cerebellum has a left and a right part, which are called hemispheres. Four lobes are distinguished on each hemisphere: the frontal lobe, temporal lobe, parietal lobe, and occipital lobe (Figure 5).

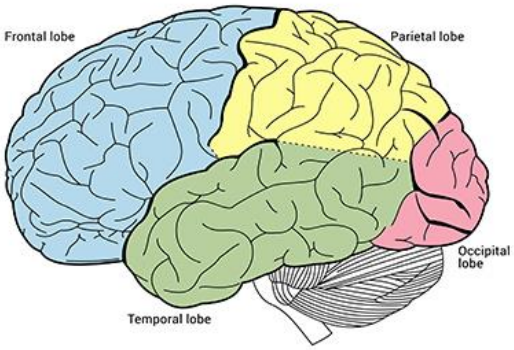

Figure 5: The lobes of the brain [15]

The outer surface is formed by the cerebral cortex, which is highly folded. These so-called folds form the sulci, which are grooves in the hemispheric surface, and gyri, which are folds in the hemispheric surface. The sulci form boundaries between the lobes; the sulcus centralis forms the boundary between the frontal and parietal lobe; the sulcus lateralis is the boundary between the parietal and frontal lobes on the one hand and the temporal lobe on the other hand. The lobes that receive and process auditory input are the temporal lobes. The temporal lobes are also critical for the comprehension of music and language. Each temporal lobe receives the input coming from both ears. Located on the superior temporal gyrus and buried in the Sylvian fissure, is the primary auditory cortex. The Sylvian fissure is the most distinct and consistent barrier of the hemispheric surface on the lateral side of the brain, which contains important vascular structures surrounded by cerebrospinal fluid [15]. The medial geniculate nucleus sends the auditory input to the Sylvian fissure. Corresponding with other cortical areas, the superior temporal gyrus and sulcus as well as the prefrontal cortex are involved during challenging listening tasks. Lateral to the primary auditory cortex is located the association cortex. The association cortex is involved in the higher-order processing of auditory input [3,12,17,18]. Wernicke's area is also a higher-order auditory association cortex and is located on the left superior temporal gyrus. Wernicke's area is essential for language comprehension. Regarding music perception, both association cortexes within the left and right hemispheres are involved [10, 17, 19].

2. Cognitive functioning

Attention

Evidence supports the idea that the essential characteristics of the sound sources in the surroundings, as well as the overall goal of the listener, determine what auditory object has the focus of attention [20].

There are two types of attention, selective attention, and sustained attention. With selective attention, an individual purposely focuses on a specific stimulus stream to process its information more thoroughly. Doing so, they ignore other stimulus streams that could be a potential distraction. Sustained attention means that an individual is able to maintain their attentional focus as time strikes [21]. An example could be whether a listener is able to perceptually segregate his/her conversation partner's voice from the sound mixture in crowded surroundings. This can help to determine how well and effective the listener can tune out the surrounding noise coming from other people's chatter, for example [20].

There are two main skills that a listener should have to be efficient at selectively attending a conversation in crowded surroundings. First, a listener has to be able to amplify the characterization of the sound coming from the source of interest. Furthermore, a listener must be able to suppress noise from sources that are not the focus of attention on the one hand, while maintaining some awareness to be able to refocus rapidly when necessary. This means that the impact on the effectiveness of selectively attending depends on the way in which the auditory objects are formed into the desired element, in both "complex" and "easy" scenes. Thus, auditory object formation must be strong in order to selectively attend to a source of interest [20]. Speech perception needs the rapid integration of bottom-up and top-down attention components. As hearing is based on real-time perceptual judgments, the dependence on the information gained from the context of this process increases [10].

Working memory

Working memory is a tool used by everyone to help us perform efficiently and effectively in all aspects of our lives. This essential tool is defined as the ability to maintain and manipulate information in the mind for a brief period of time, often termed "short-term memory" [50]. Working memory is a vital ability for storing short-term information, words, and meanings [51].

Working memory is critical for basic language skills that are required for recognizing words and understanding sentences and paragraphs. Also, because language learning and development require the individual to follow, retain, and integrate a stream of auditory information, working memory is likely to be a core component of language development following cochlear implants [52].

Reading comprehension is closely linked to working memory. Reading comprehension and word decoding correlate with phonological skills in hearing children. Children with CI also use phonological (coding) skills in reading [53,54].

In the multicomponent working memory model (Figure 6) from Baddeley, (2000), short-term memory (STM) is described as a temporary storage system with limited storage time and capacity (100). Working memory (WM) is described as a temporary storage system with a limited capacity in which information is stored and processed currently (100). In WM, there is one main component, the central executive and three subsystems: the phonological loop, the episodic buffer, and the visuospatial sketchpad. The central executive is the attentional controller with a domain-general limited capacity. This system is responsible for mental set-shifting, information monitoring and, when necessary, for updating and the inhibition of irrelevant information. The three subsystems are subordinate to the central executive and their main task is to store less attentional-related aspects of experience. Simultaneous processing and storage demand a lot of attention, while storage on its own does not demand that much attention, unless the number of items to be stored is too numerous.

The verbal STM system primarily stores auditory-verbal information for a short period as a phonological code, which is crucial for vocabulary and language acquisition in children.

The domain-specific verbal STM contains phonological storage (briefly, storage of auditory-verbal material) and a sub-vocal rehearsal process (stored material is to be sustained and refreshed).

The second subsystem is the episodic buffer, which is an interface between the long-term episodic and semantic memory with three WM subsystems. The third subsystem is the visuospatial sketchpad. This sketchpad is assumed to be responsible for storing both visual and spatial information. The visuospatial sketchpad may also be involved in language acquisition. Semantic knowledge is likely to play a role in understanding objects, figuring out machinery mechanisms, organizing items and acquiring geographic knowledge [100, 101].

Both the processing and storage components of WM depend on attention. When attention is reduced for whatever reason, memory traces decay [100].

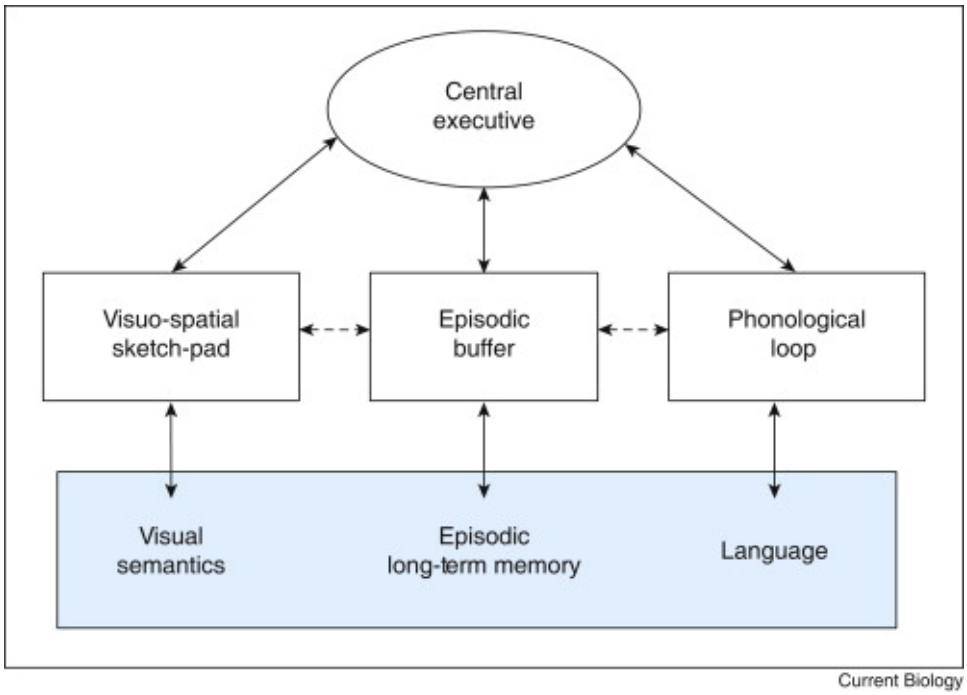

Figure 1: Working memory (Baddely, 2000) [101]

The working memory deficit is related to the type of language (signed vs. spoken) used for communication [55]. Consequently, the demands of sign language on memory may be higher than those of spoken language.

Finally, phonological awareness for spoken language is affected in signing deaf persons [56,57]. Although it is difficult to exclude comorbid disorders completely, these results indicate that higher-level functions (even non-auditory ones) possibly require an auditory reference (via top-down interactions) for proper function [26].

Working Memory with attention

Evidence implies that in order to successfully control auditory attention, the availability of the working memory (WM) is important. WM is important, regardless of the task or the sensory modality. The WM's specific role in successfully directing attention is to hold in mind what the current priority of the

assignment is. Therefore, it refers to the ability of an individual to hold and manipulate a set of stimuli currently in their mind [22,23]. Specifically, it serves as a primary intermission between the initial sensory input and the stored knowledge systems of individuals. In hearing, this can refer to knowledge systems about a listener's language. As the WM processes all kinds of sensory and perceptual information, it is momentary and very fragile. So, naturally, the WM can display solid limitations within its capacity. Information from sensory and perceptual stimuli is rapidly lost from the WM when long-term memory (LTM) processes did not take place. For instance, the rehearsal process, elaboration process, and the overall "contact" with the LTM. The WM also forms an important link between subsystems used in different skills such as production, perception, and action [24].

What happens when the WM is "loaded" or "full"? Evidence suggests, on the one hand, that when the working memory capacity (WMC) is "loaded", the degree of interference caused by irrelevant stimuli in auditory speech increases. On the other hand, it is being suggested that cognitive load can reduce distraction from irrelevant stimuli when people need to attend to a single stimulus. This would mean that the load on the WM can facilitate attentional focus due to a higher demand on the attentional resources which are involved (Bayramova et al., 2020). Recent studies related to children, showed that WMC and vocabulary could determine important predictors when listening to speech in challenging circumstances [23].

Plasticity of the auditory system

By now, it should be clear that the ability to focus attention on relevant sounds in acoustic surroundings is important for everyday life. Especially when communicating. Short term plasticity might be the explanation for this foundational function [12]. Plasticity means that the brain is able to reorganize functions and structures following a given event or set of events [19]. Thus, when learning, a sensory stimulus gains new behavioral meaning. This results in the reorganization of the representation of all that is involved and associated with that specific sensory stimulus. Cortical mechanisms as well as subcortical mechanisms contribute to this process [26].

Within the neurosciences, research has been done to examine whether the short-term plasticity caused by selective attention was limited to nonprimary auditory areas in the cortex or whether selective attention was already modulated when processing sound in the primary auditory cortex. The outcome was that it seems that during selective listening, the nonprimary auditory cortex displays more vigorous modulation in which the effects also involve the primary auditory cortex. This simply means that selective attention can modulate the processing of present sounds across multiple levels of the auditory system. Thus, it can be assumed that short-term plasticity of the auditory cortex contains the ability to filter the relevant stimuli out of the countless other simultaneous stimuli when listening [12].

Evidence for plasticity within hearing has also been gathered. Some cells are not able to be "restocked". Hair cells are those kinds of cells. When one loses hair cells, they also lose the advantages afforded by the cochlea and its active process. This results in reduced sensitivity and, thus, difficulty in hearing [8]. A cochlear implant (CI) can be used to avoid the potentially harmful effects of this sensory loss The CI can directly stimulate the auditory nerve and therefore, the brain. It is critical that the CI be placed in young children as soon as possible. The juvenile brain has a greater capacity and ability to reorganize. There are, in fact, specific time periods where sensitivity is greater, and so developmental changes are possible because of this higher neuronal plasticity. These periods are called "sensitive periods". The first sensitive

period where the auditory system is maximally plastic takes place in the first 3-4 years. This allows the cortex to progress in "maturing" [26].

Cross modal plasticity

Land et al. (2016) suggest that cross-modal plasticity caused by congenital sensory loss triggers the deprived sensory cortex. Apparently, as best documented in deaf individuals, cross-modal reorganization is related to superior abilities in the remaining senses. This reorganization may either complement or compete with the "original" inputs to the deprived area after sensory reconstruction. This means that it can either be beneficial or unfortunate for the sensory reconstruction. The results demonstrated that, despite cross-modal reorganization, the auditory fields preserve their auditory responsiveness after congenital deafness. A disconnection was shown in the effects of deafness on auditory and visual responsiveness in the Dorsal Zone (DZ) field, but with an increase in visual responsiveness in this zone.

3. Brain plasticity and cochlear implantation

A sensitive period for cochlear implantation

Studies of children fitted with cochlear implants have established the existence of and the time limits for a sensitive period for cochlear implantation. The optimal time for cochlear implantation is within the first 3.5–4.0 years of life (and best before the second year of life), when central auditory pathways show the maximum plasticity to sound stimulation. The eventual end of the sensitive period (at approximately 6.5–7.0 years of age) has consequences for the reorganization of cortical areas and pathways [26].

Early implantation enables hearing to be put within a behavioral framework by allowing the hearing modality to interact with the environment, which provides constant feedback. This is a prerequisite for appropriate learning. The brain is hard-wired for hearing and learning to listen.

Several large population studies have confirmed that children benefit most when cochlear implantation takes place as early as possible, to use the maximum of the plasticity of the brain up to 3.0-3.6 years, when the central auditory pathways show maximal plasticity. The latency of the P1 component of the cortical auditory evoked potential (considered a biomarker of cortical maturation) decreases rapidly and reaches the normal age range in children who receive an implant before 3.5 years of age [99]. By contrast, children who receive implants after the age of 7 show abnormal cortical responses, even after many years of cochlear implant use [28,29,30,31]. Children who receive implants at this young age show significantly higher speech perception scores and better language skills compared with children implanted after 6–7 years of age [32,33,34,35].

Early implantation within a brief sensitive period allows for more adequate cortical maturation, resulting in the development of speech perception and the acquisition of spoken language.

Reorganization of the brain

Prior to cochlear implantation, the absence of auditory input to the auditory system leaves the brain vulnerable to reorganization [36,37,38,39,40,41]. Secondary and association auditory areas, which respond to multi-sensory input including hearing, vision, and touch become recruited by the visual [39,40,42] and somatosensory [43,44] systems to perform non-auditory functions.

A reorganization toward a new modality might interfere with the restoration of the original sensory input and thus could close sensitive periods for the therapy with cochlear implants [45,46]. This is related to a fundamental question of developmental neuroscience: can experience cause a sensory area to change the major driving modality and thus extensively restructure the brain's connectome? A differential role of auditory areas in cross-modal reorganization has been demonstrated, with visual reorganization specific to higher-order auditory fields in congenital deafness [47,48,49].

4. Binaural hearing

Bilateral cochlear implants attempt to restore binaural hearing by providing information to both ears. Normally, the auditory system compares, processes, and integrates subtle differences between the level and timing of sounds reaching each ear. In this way, binaural hearing allows: the identification/localization of sound sources in space [58,59]; increased perception of loudness through binaural summation [60, 46]; and improved hearing in quiet and noisy environments through the head shadow and squelch effects [61,62].

Binaural hearing also makes communication less tiring, which enables listening and communication to be a more pleasant experience. Although restoring binaural hearing is the goal of bilateral implantation, this has not yet been completely realized in children [63,64].

Outcomes improve when both implants are provided with limited delays and at young ages [63,65,58, 66,67]. As the duration of the inter-implant delay decreases, the two ears develop more symmetrical speech perception abilities, and children show increasing advantages of bilateral over unilateral implantation [65]. Significant improvements on standardized speech perception tests are seen as early as 6 months following bilateral cochlear implant stimulation in children who receive their second implants simultaneously or within short delays [65]. Furthermore, children implanted with both cochlear implants simultaneously derive significantly more benefit from the spatial separation of noise compared to children who have longer delays between implants [67].

Sequentially implanted children also seem to depend more on their first implanted ear than their second for speech perception and show less bilateral improvement (relative to unilateral implant use) on speech outcomes than children implanted simultaneously or with limited delay [66].

Limited or distorted binaural hearing challenges development. Children spend much of their time interacting and learning in dynamic environments, such as the playground and classroom [68, 69, 70] in which they listen to sounds coming from multiple directions. Binaural hearing supports detection of these sounds and the ability to distinguish one sound from another based-on differences in their spatial locations. To do this, the auditory system largely depends on comparing the level and timing of sounds arriving at both ears. Even if this binaural coding is impaired, children can benefit from hearing with both ears by taking advantage of the ear with the better signal-to-noise ratio [59]. Because the intensity at the further ear is attenuated by the head (head shadow effect), the ear closest to the sound source will pick up the sounds best.

Intact binaural hearing further improves listening compared to only listening with the ear that has the better signal-to-noise ratio (binaural squelch) and provides better audibility by combining the signals from each ear (binaural summation) [71]. Restoring children's access to spatial/binaural cues is thus important to aid hearing in most common situations. Spatial/binaural hearing is clinically evaluated by measuring

speech recognition in difficult noise conditions (speech-in-noise) or while listening with both ears over each ear alone (binaural benefit).

The improved speech perception in difficult listening situations enables the incidental learning processes to take place. Children with poor listening abilities might have problems piecing together poorly heard information and, as a result, might not profit sufficiently from incidental learning. The inability to "overhear" spoken conversations limits the access of these children to many avenues of incidental learning, and this restricts their acquisition of knowledge in the fields of advanced language development, verbal cognition, social interaction, and Theory of Mind skills [72,73].

Young children with asymmetric hearing loss have impaired access to bilateral sound and are at risk of developing poor sound localization and speech detection in noise [74,75], as well as social, educational and language deficits [76,77]. These hearing difficulties and associated challenges are likely to reflect cortical reorganization with prolonged unilateral hearing. In children with congenital bilateral deafness, early hearing through one CI for > 2 years increases activity in the contralateral auditory cortex [79, 80] and both left and right auditory cortices develop an abnormal preference for stimulation from the hearing ear [79].

Given that impairments in these networks correlate with educational outcomes [81], and that bilateral hearing is important for social and educational development [78], it makes sense to avoid cortical reorganization resulting from unilateral hearing in children. Treating asymmetric hearing loss with bimodal devices may restore bilateral access to sound, but it remains unclear how the two very different signals are processed and integrated in the cortex.

For children who have sufficient residual hearing in their non-implanted ear, there may be several advantages to bimodal hearing. First, bimodal hearing protects against unilaterally driven changes by promoting expected cortical organization, which could support specialization of the right versus left auditory cortices and cortical integration of bilateral input [94]. Second, bimodal hearing improves detection and understanding of speech in noise by providing access to sound from both sides of the head [82, 83]. Third, bimodal hearing preserves low-frequency acoustic hearing, which works better than CI stimulation for pitch perception in music and speech [83,84,85].

Evidence shows that access to sound within limited durations of bilateral deafness in early life promotes normal-like development of activity along the auditory pathways in children who have many years of hearing experience with a unilateral cochlear implant. At the same time, however, the unilaterally driven stimulation leaves the opposite pathways deprived of input and susceptible to reorganization. Providing bilateral cochlear implants to children after a period of unilateral deafness of longer than 1.5 years drives abnormal mismatches inactivity at the level of the brainstem and cortex. These abnormalities in auditory development are associated with more asymmetrical speech perception, poorer hearing in noise, abnormal sound localization, and an inability to identify inter-aural timing cues.

Cochlear implantation has become standard treatment for childhood deafness. One cochlear implant promotes significant gains in speech understanding and language development when provided early in development [86,87,88].

On the other hand, access to sound in only one ear results in impaired binaural hearing [90, 91] which is the foundation for sound localization. Without access to spatial hearing, children with asymmetric hearing

are at risk for social and educational deficits [77,90,92]. We thus sought to promote bilateral hearing development by providing the most appropriate device in each ear to our large cohort of children with deafness [90]. A cochlear implant was provided in ears with severe/profound deafness; children with bilateral deafness received two cochlear implants whereas children with better hearing in one ear received one cochlear implant and a hearing aid in the other, better ear (bimodal hearing) [83,90].

Summary

Hearing is the cornerstone of our communication, integration, and sociality (Concas et al. 2021). The ears and the brain are the two main parts with the ears converting the sound into neural signals and the brain receiving and processing these neural signals and the information they contain (Oliver 2018; Oluwule et al. 2021). Attention is crucial when listening. We use selective attention when we want to purposely focus on stimuli. Sustained attention is used when we want to maintain the focus on the stimuli (Gomes et al. 2000). Another vital ability is that of the WM. Evidence implies that in order to successfully control auditory attention, the availability of WM is important. It is clear that the ability to focus attention on relevant sounds in acoustic surroundings is important for everyday life, especially when communicating. Short term plasticity might be the explanation for this foundational function (Jääskeläinen & Ahveninen, 2014). Specifically in hearing, hair cells are cells that cannot be “restocked”; thus, one is left with reduced sensitivity and accordingly, difficulty in hearing (Hudspeth, 2015). To avoid the harmful effects of sensory loss, a CI can be placed. The younger the patient, the better. After all, infants’ and young children’s brains have greater capacity and ability to reorganize (Krall & Sharma). Limiting the period of bilateral deafness in early life is essential to drive maturation in the auditory pathways and promote optimal hearing, speech, and language development.

Bilateral and unilateral deprivation should be limited to promote optimal binaural hearing in children who use cochlear implants and enable them to function better and more naturally in challenging listening situations such as the playground or classroom environments.

A more comprehensive understanding of the neural correlates of individual variability will be critical to developing better habilitation options that are aimed at, and customized for, individual patients. An individualized approach to the patient, which takes into account the individually different central compensation processes, would optimize rehabilitation success. Future directions in the patient’s rehabilitation may include the use of electrophysiological and brain imaging measurements as cortical biomarkers of the functional developmental state of the individual patient, and hence, allow customization of the habilitation options after cochlear implantation.

Chapter 5 - Fundamentals in Speech Acoustics

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In the speech sound wave, one word runs into the next seamlessly; there are no little silences between spoken words the way there are white spaces between written words." Steve Pinker

What are speech acoustics and how do they relate to children with a CI?
What characteristics can you use to describe vowels?
What is the role of auditory feedback in children with a CI?

What is speech acoustics?

Speech is produced by forcing air from the lungs through the traches and then the rest of the vocal tract. For some speech sounds, the vocal cords vibrate, and for others, they don't. The vibration of the air provides the sound waves that is defined as speech [1].

Definition: Acoustic phonetics is the study of the acoustic characteristics of speech, including an analysis and description of speech in terms of its physical properties, such as frequency, intensity, and duration [2].

The shape of the vocal tract between the glottis (vocal cords) and the lips determines which speech sound is produced. The vocal tract constricts and expands in crucial places to change the resonant frequencies associated with a speech sound.

The two basic types of speech sounds are vowels and consonants. Most often, a vowel is a must have component in a syllable. In typically developing children, the acquisition of vowels is relatively easy, and they are acquired earlier than consonants, regardless of the language spoken. Generally, vowel acquisition is complete before the age of 3 years. Corner vowels are mastered easier and are produced with higher accuracy than other vowels; tense vowels are acquired earlier than lax vowel [3].

Speech features can be categorized into classifications:

- High and low vowels
- Voiced and voiceless consonants
- Plosives and fricatives

With these features most vowels and consonants can be described across different languages [4].

CONSONANTS (PULMONIC) © 2018 IPA

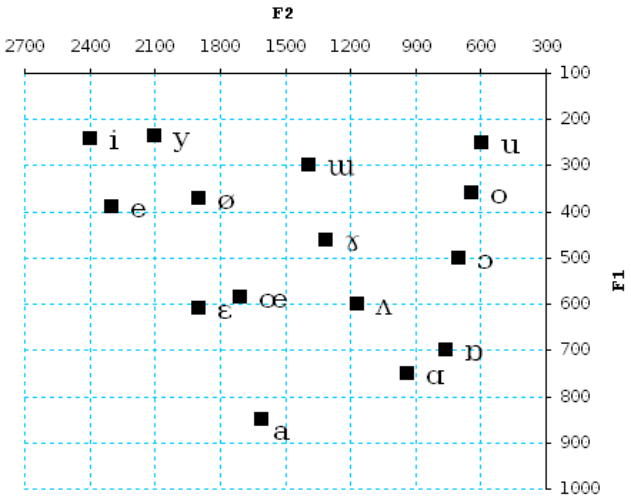
	Bilabial	Labiodental	Dental	Alveolar	Postalveolar	Retroflex	Palatal	Velar	Uvular	Pharyngeal	Glottal
Plosive	p b			t d		ʈ ɖ	c ɟ	k g	q ɢ		ʔ
Nasal	m	ɱ		n		ɳ	ɲ	ŋ	ɴ		
Trill	ʙ			r					ʀ		
Tap or Flap		ⱱ		ɾ		ɽ					
Fricative	ɸ β	f v	θ ð	s z	ʃ ʒ	ʂ ʐ	ç ʝ	x ɣ	χ ʁ	ħ ʕ	h ɦ
Lateral fricative				ɬ ɮ							
Approximant		ʋ		ɹ		ɻ	j	ɰ			
Lateral approximant				l		ɭ	ʎ	ʟ			

Symbols to the right in a cell are voiced, to the left are voiceless. Shaded areas denote articulations judged impossible.

Figure 1: Consonants (Source: https://linguisticsstudyguide.com/manners-of-articulation)

All phonemes of speech have identifying resonant frequencies, known as formant frequencies. They are distinctive for every different language. The movement of the articulators can change these formant frequencies and therefore result in different sounds [1]. Formants are distinctive frequency components of the acoustic signal produced by speech. Speech is a complex acoustic signal, with features we rely on to recognize them individually. If a formant is compared to a single tone on the piano, then speech is a chord on the piano, with each formant representing a single note [12].

A formant is the broad spectral maximum that results from an acoustic resonance of the human vocal tract. The fundamental frequency of the voice is not called a formant. The next peaks in frequencies are called formants. The formant with the lowest frequency is called F_1, the second F_2, and the third F_3. Mostly the first two formants are sufficient to identify the vowel [4,5].



Vowels			Formant 1 (Hz)		Formant 2 (Hz)	
			Female	Male	Female	Male
oo	/u/	boot	459	378	1105	997
ou	/ʊ/	book	519	469	1225	1122
oh	/o/	boat	555	497	1035	910
aw	/ɔ/	ball	781	652	1136	997
uh	/ʌ/	bug	753	623	1426	1200
ah	/ɑ/	bot	936	768	1551	1333
er	/ɝ/	bird	523	474	1588	1379
ae	/æ/	bat	669	588	2349	1952
eh	/ɛ/	bed	731	580	2058	1799
ay	/e/	bay	536	476	2530	2089
ih	/ɪ/	bit	483	427	2365	2034
ee	/i/	beat	437	342	2761	2322

Figure 2: Formants in vowels (Source: https://en.wikipedia.org/wiki/Formant)

Figure 3: Average formant values of English vowels for male and female voices (Source: https://blog.medel.pro/introducing-the-building-blocks-of-speech)

To detect a speech sound, at least one formant must be heard, but to identify a speech sound, multiple formants need to be distinguished [12].

Plosives modify the placement of formants by the surrounding vowels. This sometimes applies for fricatives as well. In bilabial sounds, the formants are lower. For velar sounds F_2 and F_3 are almost always combined in a velar pinch before the velar. Alveolar sounds cause fewer systematic changes in neighboring vowel formants. The rapid changes in vowel formant frequencies are referred to as "formant transitions." [5].

When talking, people listen to themselves—what they say and how they say it. Real time feedback plays a crucial role in this process.

Auditory feedback has a key role in feedback control and in updating feedforward/predictive control. Perturbations of the auditory feedback can cause auditory errors or discrepancies between expected and perceived feedback. Speakers can learn from errors that are produced by consistently applied perturbations, and they adapt their motor plans for future utterances to incorporate information from those errors [6].

Speech acoustics and hearing loss

Normal hearing people exploit the formant transition detection to identify the place of articulation for stop consonants. The formants of naturally produced stop consonants undergo rapid formant transitions (FT) after the consonant release. These formant transitions reflect the change of vocal tract shape from consonant to vowel (or from vowel to consonant). They are highly effective cues for the perceived place of articulation. For hearing-impaired listeners, it is generally difficult to identify the place of articulation of stop consonants. This could be caused by the rapid formant transitions and the difficulties in the perception of these rapid changes. There is also psychological evidence suggesting that poorer speech intelligibility in hearing-impaired people may be due to a degraded representation of the speech signal in the auditory nerve [7,8].

Hearing aids can help to compensate for disorders through the amplification of a sound, but the effectiveness of the hearing device also depends on the ability of the auditory nerve system to integrate the given spectral and temporal information. There is a strong correlation between the speech performance of cochlear implant clients and temporal processing capabilities. This suggests that phoneme recognition, especially consonant recognition, relies on the client's ability to detect temporal modulation [7,8].

As research shows, many factors play a role in the speech perception and language development of children with CI. The age of implantation appears to be one of the most important and crucial ones. Inadequate stimulation of the hearing system before implantation causes both pathological and atrophic changes to occur both in the cochlea and central auditory pathways. Inadequate stimulation leads to impairment of temporal processing skills. There is a significant relationship between temporal processing and speech recognition performance in users of cochlear implants [9].

Temporal auditory processing is the ability to resolve and track rapid temporal changes that occur over time. This process is crucial for the process of the speech perception ability and understanding speech in noise [10].

Temporal processing contains two aspects: temporal resolution and temporal sequencing. Temporal resolution is an acoustic spectral time analysis that shows the difference between sounds in milliseconds. Temporal sequencing is the ability to process sounds in order of occurrence. During the speech, voices follow each other with short gaps, that is why the ability of accurate temporal ordering and temporal resolution is important for speech perception. And speech perception—both of your own speech and the speech from others— is crucial for speech production [3,6,9].

Cochlear implants have helped hearing-impaired children to restore auditory sensations, which contributes to the improvement of their communication skills. Communication skills include perception and production, but also language abilities and academic performances [3].

Research shows that the accuracy performance for vowels and diphthongs improves post-implantation. Front and back vowels show the biggest improvement, diphthongs the smallest. Among different types of single vowels, central vowels are the most produced types of vowels pre-implantation. Post-implantation front vowels are the most frequently produced vowels.

As expected, monophthongs are acquired more rapidly and earlier than diphthongs. This is for normal hearing people, as well as for cochlear implanted people.

Within the first 12 months after implantation, the monophthongs emerged and were acquired within 36 months post-implant. After 48 months post-implantation, 75% of the diphthongs met the targetless criterion, and 63% met the target criterion. This shows that children with CI follow the same developmental steps as normal hearing children, but with a delay in their acquisition [3].

Studies show a significant improvement in vocal and phonological development after implantation [10]. To determine whether an individual has sufficient auditory access to recognize vowel sounds, the audiogram can be compared with the formant information described for a particular vowel [12].

Looking back at the formants, CI children produce F1 in a wider range and F2 in a reduced range, compared to age-matched normal hearing children. After 3 years of CI use, there was a progressive shift in both F1 and F2 in the direction of age-matched norms. But still, the F1 and F2 values were still higher than the normative data for low, back vowels [3].

The CI children produce clearer vowels when there is auditory feedback, and their speech is more intelligible [11].

Children with a CI produce the three corner vowels differently from children with normal hearing controls. They have a significant slower speaking rate, smaller vowel space area, and lower speech intelligibility. The vowel space area in sustained phonation was significantly correlated with ratings of vowels and words. This shows that children with a CI show deviation from their normal-hearing peers, even years after implantation, on the fine-grained acoustic-phonetic characteristics of vowels. This could be associated with the slightly distorted speech signals they receive from the device [3].

For example, vowels show a longer duration, and the overall vowel intelligibility was lower than in normal hearing peers. Differences in consonants will also appear in children with unilateral CI as well as children with bilateral CI, both implanted before the age of 5 years [10].

For children with a cochlear implant, normal speech development is less likely to occur without intensive and good training and habilitation [3].

Good speech perception is a prerequisite for children to learn to produce intelligible speech. CI children with better tone perception tend to have higher accuracy in tone production [11].

Auditory feedback is also important for good production, and it helps to produce clearer vowels. Intelligibility increases when the child has access to good auditory feedback [10]. During the rehabilitation, the use of visual aids and tools to guide the child to appropriate articulation targets may be useful [3]. The importance of both tone perception and production supports this vision [11].

Another important aspect of the rehabilitation is the mode of communication during the rehabilitation. Purely looking at the speech production, oral communication shows better results in speech development versus total communication. Age of implantation and early habilitation are crucial both to better speech production and high intelligibility of discourse [10].

Summary

Speech is produced by forcing air through the vocal tract. A different shape of the vocal tract leads to different sounds. The two basic types of speech sounds are vowels and consonants. In learning speech, vowels are easier to learn than consonants. Every speech sound has its own formants, which means distinctive frequencies. In learning speech, auditory feedback plays an important role. Perturbations in receiving speech can lead to perturbations in your own speech. Children with a hearing loss miss this auditory feedback, at least until the CI is implanted. The earlier the implantation, the better the results on speech production and intelligibility.

Chapter 6 - Clinical Instrumentation to Explore Auditory Function

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„The works must be conceived with fire in the soul but executed with clinical coolness." Joan Miró

Introduction

This chapter describes the tests available to identify hearing loss caused by a peripheral pathology and/or a central lesion, but also presents clinical information on the anatomophysiologic mechanisms of audition and the physical basis of a test to understand what and when each audiologic investigation is needed. Clinic examination as well as audiometric tests are required to evaluate hearing, because the structure and function of the auditory system are interconnected. The medical examination will be guided by the anamnesis, the assessment of affect, the intellectual performance, the inspection of the face, the memory capabilities, and the assessment of central language functions.

The detailed examination of the ear using an otoscope, or more recommended an otomicroscope, begins with auricle inspection and continues with the external conduct, the tympanic membrane, and the middle ear evaluation. Wax and multiple diseases such as otitis media with effusion, acute and chronic otitis media, or cholesteatoma should be identified, treated, or noted when we do audiometric tests.

What are the three most important multidimensional planes in the PRIMIR framework, and why are they important?

When is the best time to do the hearing tests?

The Joint Committee on Infant Hearing (JCIH) endorses early detection for infants with hearing loss. The goal is to ensure that all infants are identified as early as possible, and appropriate intervention is initiated, no later than 3–6 months of age.

Why is early detection so important?

The importance of Early Hearing Detection and Intervention (EHDI) is to maximize linguistic competence and literacy development for children who are deaf or hard of hearing.

Which are the worst consequences of a late hearing loss diagnosis?

Without appropriate opportunities to learn language, these children will fall behind their hearing peers in communication, cognition, reading, and social-emotional development [1]. Such delays may result in lower educational and employment levels in adulthood [2]. Regardless of previous hearing-screening outcomes, all infants, with or without risk factors, should receive ongoing surveillance of communicative development beginning at 2 months of age during well-child visits in the medical home. EHDI systems should guarantee seamless transitions for infants and their families through this process [3].

It has long been recognized that unidentified hearing loss at birth can adversely affect speech and language development as well as academic achievement and social-emotional development. Historically, moderate-to-severe hearing loss in young children was not detected until well beyond the newborn period, and it was not unusual for the diagnosis of milder hearing loss and unilateral hearing loss to be delayed until children reached school age.

All children with hearing loss should have access to the resources necessary to reach their maximum potential. The following principles provide the foundation for effective EHDI systems and have been updated and expanded since the 2019 JCIH position statement [1]:

All infants should have access to hearing screening using an objective physiologic measure at no later than 1 month of age.

All infants who do not pass the initial hearing screening and the subsequent rescreening should have appropriate audiological and medical evaluations to confirm the presence of hearing loss at no later than 3 months of age.

All infants with confirmed permanent hearing loss should receive intervention services as soon as possible after diagnosis but no later than 6 months of age. A simplified, single point of entry into an intervention system that is appropriate for children with hearing loss is optimal.

The EHDI system should be family-centered on infant and family rights and privacy guaranteed through informed choice, shared decision-making, and parental consent in accordance with state and federal guidelines. Families should have access to information about all intervention and treatment options and counseling regarding hearing loss.

The child and family should have immediate access to high-quality technology including hearing aids, cochlear implants, and other assistive devices when appropriate.

All infants and children should be monitored for hearing loss in the medical home. Continued assessment of communication development should be provided by appropriate professionals to all children with or without risk indicators for hearing loss.

Appropriate interdisciplinary intervention programs for infants with hearing loss and their families should be provided by professionals who are knowledgeable about childhood hearing loss. Intervention programs should recognize and build on the strengths, informed choices, traditions, and cultural beliefs of the families.

Information systems should be designed and implemented to interface with electronic health charts and should be used to measure outcomes and report the effectiveness of EHDI services at the patient, practice, community, state, and federal levels.

Hearing Screening

The Universal Newborn Hearing Screening (UNHS), component of EHDI programs, requires multidisciplinary teams of experts, including audiologists, physicians, and nursing personnel. To guarantee that screening programs are of high quality and successful, all team members collaborate. Each component of the hearing-screening program should include an audiologist, especially at the statewide level and, whenever possible, at the individual hospital level. A physician should be designated by hospitals and organizations to oversee the medical parts of the EHDI program [1]. The hospital infrastructure should be reviewed in relation to the screening program by each team of specialists responsible for the hospital-based UNHS program. Screening technology (such as OAE or automated ABR testing) should be considered in hospital-based programs. The validity of the specific screening device, screening protocols, including screening timing in relation to nursery discharge, the availability of qualified screening personnel, the suitability of the acoustic and electrical environments, the follow-up referral criteria, the referral pathways for follow-up, the information management and quality control, the improvement of the content of reports to physicians and parents, the documentation of outcomes in medical charts, and the procedures for reporting to state registries and national data sets all require well-defined reporting and communication standards [4].

Physiologic measures must be used to screen newborns and infants for hearing loss. Such measures include OtoAcoustic Emissions (OAE) and automated Auditory Brain Responses (ABR) testing. Both OAE and automated ABR technologies provide noninvasive recordings of physiologic activity underlying normal auditory function, both are easily performed in neonates and infants, and both have been successfully used for UNHS [5,6,7].

The two measures, however, have significant disparities. OAE measurements are collected from the ear canal using a sensitive microphone within a probe assembly that records cochlear responses to sonic stimuli. As a result, OAEs represent the state of the peripheral auditory system, which includes the outer hair cells of the cochlea. ABR measurements, on the other hand, are taken using surface electrodes that record neural activity in the cochlea, auditory nerve, and brainstem in response to acoustic stimuli provided through an earphone. The state of the peripheral auditory system, the eighth nerve, and the brainstem auditory pathway are all reflected in automated ABR measures [8,9].

Audiologic Tests Used for Children

Audiologic tests describe abnormalities within the auditory system, establishing the location of the lesion and quantifying the loss. Specifically, the purpose of the testing is to: a) quantify the audiometric threshold at each frequency; b) differentiate conductive from sensorineural hearing loss; c) differentiate cochlear from retrocochlear abnormality; d) identify central auditory dysfunction in the brainstem, midbrain, or auditory cortex; e) identify any nonorganic hearing impairment [9,10].

Very young infants, of course, do not have the cognitive or motor abilities to provide an audiogram as older children and adults do. Thus, the information about the amount, type, and configuration of hearing loss must be inferred from objective, physiologic measures [11]. The test is “objective” in that it does not require the active participation of the child.

Pure Tone Audiometry

The pure tone audiogram (PTA) is the fundamental component of audiologic testing. It is used to screen for hearing loss and confirm or deny the presence of hearing loss, but it cannot be used on infants and small kids.

During hearing testing, one of the goals is to determine the degree of hearing impairment, in general, as a function of frequency or pitch. If there is a hearing loss, the test will also verify or deny the presence of this condition.

The type of loss (conductive or sensorineural hearing loss) and the location of the problem can also be determined. Medical diagnosis and pathology involve the use of this information, as well as management strategies such as the selection and fitting of hearing aids. An audiogram represents the results of a basic pure tone test of hearing, namely a graph of threshold levels as functions of frequency.

A soundproofed room is used for pure-tone audiometry as noise in the room affects low-frequency thresholds particularly.

There are two ways to deliver pure tones: either using headphones (for air conduction thresholds) or applying a bone conductor to the mastoid process (for bone conduction thresholds).

Pure tones are delivered through headphones (for air conduction thresholds) or a bone conductor applied to the mastoid process (for bone conduction thresholds).

Pure tone oscillators can be configured to display a wide range of frequencies, and the signal strength can be adjusted on a calibrated scale from –10 to +90 dB HL [9,10,11].

The pure-tone oscillator can be set to one of several different frequencies, and the level of the signal can be adjusted according to a calibrated scale from –10 to +90 dB HL. A threshold is defined as the lowest level at which a pure tone can still be heard on at least 50% of presentations at a particular frequency. To determine a subject's AC or BC threshold for a frequency, the subject responds to the quietest tone presented in the manual audiometry procedure. It is considered normal to have air conduction threshold values better than 20 dB HL. Bone conduction threshold values that are significantly greater than air conduction thresholds suggest a malfunction of the middle ear-inner ear communication, conductive hearing loss. The similarity between bone and air conduction thresholds implies normal hearing (<20 dB) or sensorineural hearing loss(>20dB) (Figure 1). Transmission or mixed hearing loss are indicated when there is difference between bone and air conduction thresholds (Figure 2).

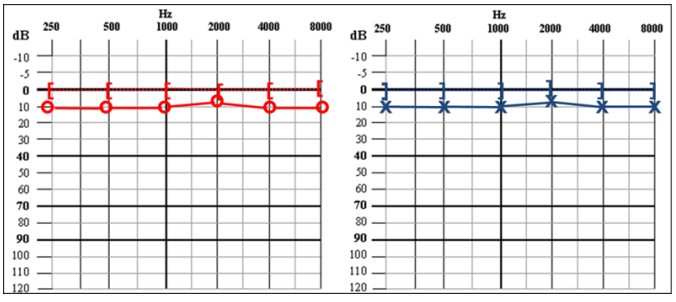
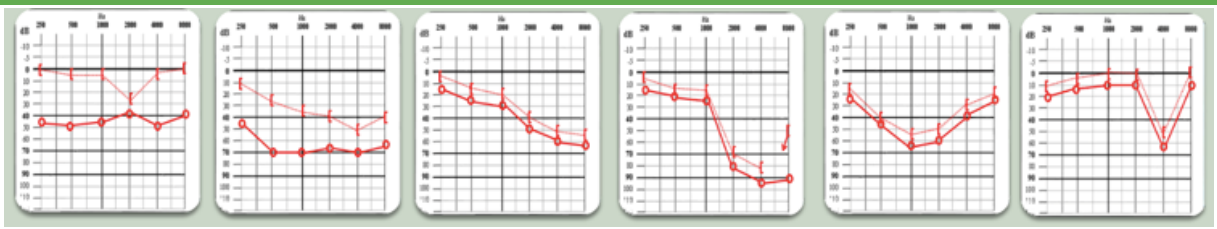


Figure 1 – Normal hearing (Archive of Audiology compartment, Recuperare Hospital, Iasi)



1 2 3 4 5 6

Figure 2 - Different types of tonal audiometry:

1-Conductive hearing loss 2-Mixed hearing loss 3-Presbiacusis 4-Sensorineural hearing loss sky slope type 5-Congenital Sensorineural hearing loss 6-Acoustic trauma hearing loss.

The following standard format is used to plot the results of pure-tone audiometric measurements:

- X = left-ear air conduction threshold
- O = right-ear air conduction threshold
- △ = unmasked bone conduction threshold
- [= right masked bone conduction threshold
-] = left masked bone conduction threshold

Typically, hearing loss is classified into one of four categories (mild, moderate, severe, or profound), based on its relationship to the normal hearing range (0 -20 dB HL) [12].

Severity

- Normal: better than 20 dB HL
- Mild: between 20 and 40 dB
- Moderate: between 41 and 70 dB HL
- Severe: between 71 and 90 dB HL
- Profound: in excess of 90 dB HL.

Frequency

- Low-frequency (250 and 500 Hz)
- Mid-frequency (1000 and 2000 Hz)
- High-frequency (4000 and 8000 Hz).

Acoustic Immittance Measurement (tympanometry)

Tympanometry, or measurement of the acoustic immittance of the ear, obtains information about the state of the middle ear as a function of ear canal pressure. The difficulty that a sound wave (acoustic energy) faces as it travels through the ear is referred to as “acoustic immittance”. The ear's "stiffness" is the property that keeps it in shape and allows it to be restored after a force has been applied. The middle-ear system's mobility is represented by compliance, which is the reciprocal of stiffness. Acoustic immittance detects both high-impedance and low-impedance middle ear disorders, such as otitis media and otosclerosis, as well as low-impedance abnormalities like ossicular interruption [12]. The advantages of tympanometry are that it is objective and thus requires no behavioral response, it is noninvasive and well tolerated, and it is quick and inexpensive. A flexible tube attached to one of three orifices in a probe tip assembly delivers a variable-intensity, low-frequency pure tone (220–226 Hz) created by a small sound source to the ear’s acoustic meatus (EAM).

The other two components include a tube attached to a pump that changes the ear pressure in the EAM and measures it manometrically, as well as a tube that transports sound waves from the EAM to a microphone for transduction into electric activity.

The electronic signal represents the level of sound pressure. It is compared to a reference voltage provided by an impedance meter, and the result is recorded on a balance meter. The probe assembly is inserted into the EAM to make an airtight seal. In a normal ear, the middle ear pressure (MEP) will range from +50 to –50 mmH2O (adults) and from +50 to –100 mmH2O (children). The tympanogram is obtained by varying the external meatus pressure from –600 to –200 daPa and is now recorded automatically, giving a hard copy, and also evidence of rhythmic fluctuations in compliance due to conditions such as tensor tympani myoclonus or the pulsatile tympanogram of a glomus tumor.

High-impedance abnormalities**:**

- Perforated TM: flat tympanogram with middle-ear compliance >2.5 up to 4.5 mL
- Middle-ear effusion: high impedance with a flat pressure peak inversely correlated with the amount of middle-ear effusion
- Ossicular fixation: shallow tympanic peak, MEP peak around 0 mL (type As).

Low-impedance abnormalities:

Thin, atrophic TM, i.e., flaccid: high tympanic pressure peak with sharp notching.

Ossicular disruption: high tympanic pressure peak, low static impedance measurements.

The most commonly used classification system was introduced by Liden and modified by Jerger with Feldman [13] describing an analytic approach:

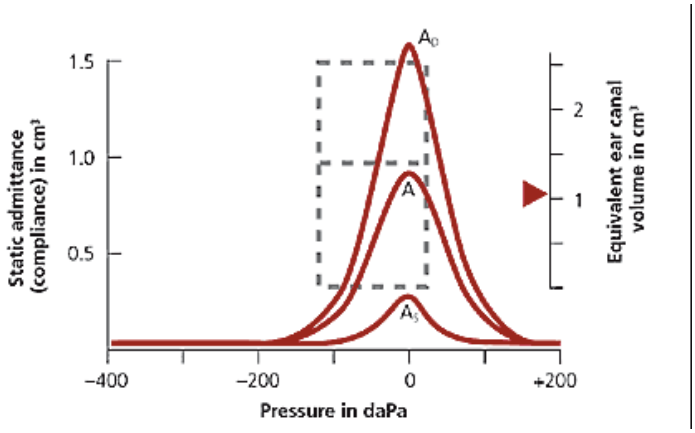


Figure 3 (Am Fam Physician 2004;70(9):1713-1720)

Type A is normal – the peak immittance is at or near 0 daPa. (Figure 3)

Type AD shows an unusually high peak pressure, e.g., a flaccid TM or ossicular discontinuity. (Figure 3)

Type AS shows a reduced pressure peak, e.g., ossicular fixation and some forms of otitis media. (Figure 3)

Type B shows a flat pressure peak such as with a middle-ear effusion or other space-occupying lesions of the middle ear. Alternatively, this may be seen with obstructing wax. (Figure 4)

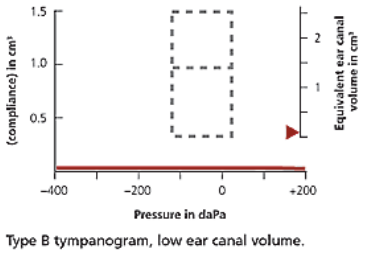


Figure 4 Type B

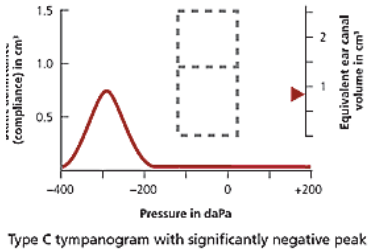


Figure 5 Type C (Am Fam Physician 2004;70(9):1713-1720)

Type C shows a negative peak pressure and indicates negative MEP, such as with a retracted eardrum (Figure 5).

Type D shows sharp notching, characteristic of scarred eardrums or hypermobile TM.

Type E is characterized by broad, smooth notching and is most commonly found in cases of partial or complete ossicular discontinuity.

Stapedius reflex thresholds

Measurements of the stapedius reflex reveal information on the middle and inner ear, as well as the eighth and seventh nerves (which innervate the stapedius) and brainstem function. The dynamic changes that occur when the stapedius muscle contracts in response to stimuli of 500, 1000, 2000, and 4000 Hz at intensities of 70–115 dB sound pressure level are assessed, and activation thresholds are described. Stapedius reflex thresholds arise from stimulation across the four frequencies of 500, 1000, 2000, and 4000 Hz.

Behavioral observation audiometry

It is a test used to observe hearing behavior to sound, often utilized for infants under the age of 6 months or who are not able to turn their heads towards a sound. In many ways, this test is restricted. Diverse types of stimuli elicit different responses in infants of various ages. Infants are more likely to respond to broadband complex stimuli (particularly speech) than to narrow bands of noise or pure tones. As a result, the most frequent specific signals, such as those employed in audiometry, are the least likely to elicit responses from newborns, especially at low stimulus levels (near threshold). Because there is so much variation across participants, detecting mild and moderate degrees of hearing loss is effectively impossible. Individual newborn reactions are likewise highly variable, and responses are prone to habituation. A final limitation of BOA is that examiner bias, knowledge of the stimulus level, or knowledge about previous test results may result in underestimating or overestimating hearing loss [15,17].

Electroacoustic and Electrophysiologic Test

Otoacoustic emissions

An otoacoustic emission (OAE) is a low-intensity, mild- frequency sound, generated from within the inner ear by the active movement of the outer hair cells. OAEs were first demonstrated experimentally in 1978 by Kemp. TEOAEs are elicited by transients of 80–86 dB summating potential, which are collected in the first 20 ms after the stimulus is presented. If the response amplitude is >4 dB higher than the background noise, they are present.

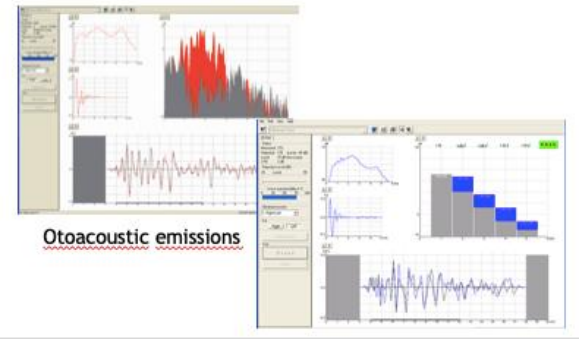


Figure 6 -Otoacoustic emissions
(Archive of Audiology compartment, Recuperare Hospital)

These tests are employed in newborn hearing screening because the presence of a response can be interpreted as showing adequate cochlear function, sufficient to imply hearing levels normally better than 30 to 40 dB (Figure 8). These levels are in accord with newborn screening results and correlate to no more than mild hearing loss. They are considered a more sensitive measure of cochlear hearing since they can sometimes be diminished despite a normal audiogram. If there is a conductive loss, false negatives result [17,18]. In the clinic, transient evoked OAEs (TEOAEs) are frequently utilized to assess proper cochlear function. OAEs diminish after damage to the inner ear, but they survive after a section of the eighth nerve, hence they have been used to show auditory neuropathy.

Auditory Evoked Potentials/BERA

This test can cover the entire auditory pathway, from the cochlear hair cells to the cerebral cortex (Figure 7). Separate auditory potentials that are time-locked to the stimulus and background noise are used in the approaches. Varying levels of the auditory system have different timing and waveforms of activity, as well as different orientations and positions of the generators. As a result, electrode sites, filter settings, and analysis timings can be configured to selectively record activity from a single generator along the auditory pathway, assisting with site pathology. ABRs are detected by surface electrodes and represent electric activity transmitted by the eighth nerve and brainstem auditory relay centers in the 10 seconds immediately after an acoustic stimulus [20]. Most of the ABR waveforms generators have been the subject of long-running controversy, with only Wave I having a clear attribution—to the cochlea. Wave II is supposed to originate in the cochlear nucleus, while waves III, IV, and V are thought to originate in the brainstem auditory circuits' generator locations (Figure 10). ABRs generated in infants for threshold prediction may require specific recording parameters that differ from those that are standard for use with adults. Infant ABRs are slower than those of adults and generally require a long analysis window of 20-30 ms and a lowered band-pass filter (30 to 1000 Hz). Infants' recordings, especially those made during natural sleep, may be noisy, requiring on average a longer time to achieve an adequate signal-to-noise ratio for reliable response detection. The ABR can sometimes be absent or severely abnormal, even when the inner ear is functioning well, due to auditory neuropathy. Children with this disorder will have an abnormal or absent ABR but usually will also show a present OAE. When this condition exists, neither the

ABR nor the OAE can be used to predict hearing levels. The hearing loss in patients with auditory neuropathy can be of any degree, and their speech perception ability is severely disordered. Therefore, it is very important to include the measurement of OAEs in any assessment of hearing using ABR. If the ABR and OAE are disparate in the prediction of threshold, then auditory neuropathy must be suspected, and neither will be a good indicator of hearing level. Depending on the purpose of the ABR, two protocols can be followed: in the neuro-otology clinic, ABRs with stimulus intensity of 90 or 100 dB are recorded both ipsi- and contralaterally. Measurements are made of the absolute latencies of the waveforms and the interwave latencies, identifying any abnormalities. Interaural wave latencies and interwave intervals are compared. Prolongation of the I–III interval can be seen in auditory nerve and cochlear nucleus pathology. Prolongation of the III–V interval is usually indicated when pathology is located above the level of the cochlear nucleus, while absent IV and/or V waves are found in cases with involvement of the mid-upper pons. Interaural latency comparisons of wave V are of value in the diagnosis of vestibular schwannoma but may not be useful in detecting brainstem involvement [12]. In audiology clinics, for neonates or others who cannot perform subjective audiometry, threshold ABRs are recorded to determine the level of hearing. The wave V latency shortens with the maturation of the eighth nerve and is considered to reach permanent values by the age of 12 months. In young children, auditory evoked potentials are utilized to estimate the audiogram. Electrical potentials can be captured non-invasively, starting at the level of the cochlea and auditory nerve and progressing to higher brain levels. The Auditory Brainstem Response (ABR) is the most used method to appraise hearing sensitivity. Because it is unaffected by consciousness, sedation, or sleep state, the ABR is particularly well suited for newborn audiometry. As in behavioral audiometry, a comparison of reactions to air-conducted stimuli and responses to bone-conducted stimuli provides knowledge about the kind of hearing loss the child has, conductive or sensorineural [12].

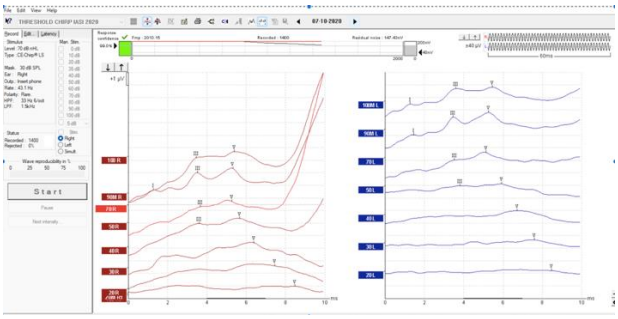


Figure 7 - ABR example test (Archive of Audiology compartment, Recuperare Hospital, Iasi

The broadband click stimulus, which is commonly employed for screening, may be utilized for two reasons in diagnostic testing: the assessment of the auditory nerve and brainstem structures' integrity, and the measurement of the ABR threshold. The second is accomplished by eliciting the ABR with high-level click stimuli, quantifying the latency of the waveform's three main peaks, and comparing it to age-dependent norms. Many audiologists utilize the ABR threshold to click stimuli to obtain a rough estimate of the level of hearing loss. But because the click consists of energy at several frequencies, its threshold can over- or underestimate the degree of hearing loss, particularly when there are dips and risings in the audiometric configuration.

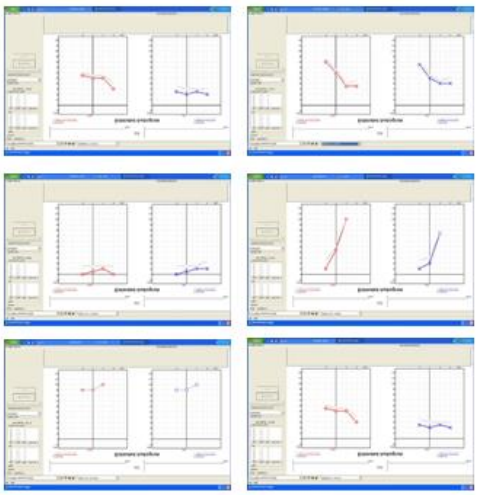
As a result, diagnostic ABR should use short-duration tonal stimuli (tone bursts) at specified frequencies to provide threshold estimations that are more equivalent to an audiogram. If it is not possible for the

four most important audiometric frequencies to be completed, then at least responses to 500 Hz and 2000 Hz need to be acquired.

Another drawback of ABR testing is that it cannot be used to diagnose severe to profound hearing loss. For defining thresholds higher than 90 dB HL, ABR testing is insufficient. Hearing losses that surpass the maximum output might be difficult or impossible to assess due to this constraint.

Because of its unusual waveform shape, especially in very young children, tone-burst ABR testing has yet to be established in common clinical practice. Furthermore, tone-burst ABR testing is a time-consuming procedure that yields no results in severe and profound hearing loss.

Auditory Steady State Response



The ASSR, formerly known as the steady-state evoked potential (SSEP), is another method for objectively evaluating frequency-specific responses. The optimum modulation frequency range for newborns and youngsters is around 80-100 Hz. The continuous electroencephalographic activity is sampled and examined in the frequency domain while long chunks of these stimuli are given. Auditory steady-state response (ASSR) testing is a technique for determining frequency-specific hearing thresholds by measuring auditory evoked potentials. Pure-tone sounds are utilized as the stimulus in ASSR testing (Figure 8). The amplitude and frequency domains of a pure-tone sound are modified. The spectral splatter narrows when a pure-tone sound stimulus is modulated, resulting in the activation of a highly limited and narrow region of the basilar membrane. As a result, ASSR testing might evoke a more frequency-specific hearing threshold than ABR testing. The neuronal activity from the brainstem is recorded if the rate of modulation is greater than 60 Hz [12].

Because brainstem responses are unaffected by the patient's mental state, they may be captured even in sedated babies and children. Hearing thresholds for the four important frequencies are determined using an objective, statistically based mathematical detection technique.

Figure 8 – Different aspects of auditory steady state response test (Archive of Audiology Compartment, Recuperare Hospital, Iasi)

Furthermore, ASSR testing permits stimulation at high-intensity levels of 120 dB HL, but ABR testing is unable to distinguish between severe and profound hearing loss. It is crucial to be able to distinguish between these major forms of hearing loss. The ability to differentiate between a 75-dB hearing loss and a 95-dB hearing loss may influence decisions like whether to fit a traditional hearing aid in an infant who has a 75-dB SNHL or consider cochlear implantation for someone with a 95-dB SNHL.

Ultimately, the objective detection of ASSRs in the frequency domain is assured by statistical tests. The waveforms or reaction patterns are not examined subjectively.

At any frequency, ASSR was unable to determine thresholds in the youngsters below 40 to 50 dB nHL (stimuli calibrated relative to thresholds in properly hearing adults). At present, using ASSR to detect hearing loss in newborns and young children, particularly those with mild to moderate hearing loss, is not recommended.

Electrocochleography

Transtympanic electrocochleography (ECochG) is a surgical method that is used to evaluate cochlear potentials. A needle electrode is introduced through the TM (if intact) and placed on the promontory of the middle ear using a microscope.

Cochlear Microphonics

Cochlear microphonics are the early components of auditory brainstem responses that occur in the 0.7–1 ms interval after the stimulus and have waveform characteristics that are comparable to the stimulus. Cochlear microphonics in the presence of an abnormal ABR are typical of an auditory neuropathy [19].

Summary

The hearing evaluation of newborns and children needs the use of specialized equipment and trained professionals. If the practitioner lacks the necessary expertise and equipment, the newborn and family should be directed to an audiologist who is trained in and experienced with infant audiologic evaluation.

Loss of hearing can be more harmful to newborns and children during important learning stages for speech, language, and general communication abilities than it can be to adults. All forms of learning for school-aged children rely on communication abilities.

The most crucial duty for an infant is the development of communication skills, as it is the foundation for practically all later learning. Physicians should inquire for answers to their queries rather than referring for a specific test, leaving the decision about the most appropriate test battery to the pediatric audiologist.

The strongest evidence that even mild bilateral hearing loss is debilitating for young children comes from the American Academy of Pediatrics' endorsement of programs for early detection of mild hearing loss of 30 to 40 dB due to evidence that this degree of loss causes significant communication and educational delays.

Cochlear implantation is the only treatment possible for bilateral profound or severe hearing loss in children and could provide very good language development when it is done at the right time.

The need for assistance for children with hearing loss is especially apparent during the baby and preschool years, when language and speech gains are at their apex. The elementary-school-age child should be able to tolerate slightly more hearing loss, especially if language skills are developing well, and the criteria level for disability is changed to moderate (50 dB) rather than mild.

Several investigators have compared the performance of children with cochlear implants to that of their peers who use conventional amplification. The speech perception abilities of pediatric cochlear implant recipients met or exceeded those of their peers with unaided pure-tone average thresholds ≥ 90 dB HL who use hearing aids [21].

It should be noted that obtaining a thorough description of the infant's hearing may necessitate more than one visit. Referrals for a medical diagnosis (etiology) and the start of early intervention services should be made as soon as it is obvious that a hearing loss exists (without waiting for all components of the examination to be completed).

Chapter 7 - Actual Rehabilitation Possibilities for Children with Hearing Aids/Cochlear Implants and Their families

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"It won't be easy, but it will be worth it."
Samantha Mckinney

What different communication approaches do you know?
What are the basic principles of auditory (re)habilitation?
How does one learn to listen and develop spoken language skills?

Different Communication Approaches

Babies and parents share a common language so that they can understand each other. Hearing babies begin to pay attention to the spoken language of their environment as soon as they are born. Deaf babies from deaf parents also begin to pay attention and learn as soon as they are born through sign language exposure. But 95% of the parents of deaf children are both hearing. These hearing parents need to find effective ways to communicate and model language for their babies [1].

Decisions about how to communicate with your young child with a hearing loss are very important for your child's early development. As parents choose a cochlear implant for their deaf child, they also choose to learn to listen and to develop the spoken language of their environment.

Approaches can also vary in the amount of emphasis placed on listening, spoken language, and/or on visual communication modalities. Any decision will require commitment and expertise from the parents and from the professionals.

We often think of these approaches on a continuum from auditory (A) emphasis in learning, to visual (V) emphasis in learning (Figure 1). By considering the child's unique learning strengths and needs, parents and communicators can use the best approach for the child. It is even possible that children might start at one place on the continuum and move along as they get older and their needs change.

Figure 1: approaches on a continuum from auditory (A) emphasis to visual(V) (adapted from Nussbaum, et al, 2004).
-A and Av: A lot of emphasis on learning to listen through the auditory (hearing) sense. e.g., auditory-verbal, or auditory-aural approach
-Av: A lot of listening practice with some visual support, like speech reading. e.g., Natural approach
-AV: Equal emphasis on auditory and visual learning. E.G. Total Communication (spoken language supported with signs) or Cued Speech
-VA and V: The emphasis on visual communication. Signed Bilingual approach which combines sign and auditory stimulation (VA), or primarily learning through Sign Language (V).

Auditory (re)habilitation: basic principles

Auditory (re)habilitation is a term used to describe the creation of a listening and learning environment in which parents, therapists, and educators help children with impaired hearing to acquire listening and speech skills. Many of these processes consist of obtaining audiometric information, providing appropriate hearing aids or cochlear implants, maintaining the child's listening device and communicating through speech and spoken language [2]. When parents and professionals are able to establish these listening and learning conditions, most children are able to develop good listening and spoken language skills.

Habilitation refers to health care services that help a person acquire, keep, or improve skills related to communication and activities of daily living partially or fully. These services address the competencies and abilities needed for optimal functioning in interaction with their environments.

Rehabilitation refers to regaining skills, abilities, or knowledge that may have been lost or compromised as a result of illness, injury, or acquiring a disability [3].

In this chapter we will use the term 'habilitation' instead of 'rehabilitation', because we focus on young deaf children receiving cochlear implants and they don't regain skills or abilities, but they will acquire these skills for the first time.

Habilitation will vary depending on the service delivery model employed by the team and on the individual needs of the child. It can be provided on an individual basis and through group work, in a live session or by using tele-practice.

Auditory habilitation can be implemented either by a top-down and/or a bottom-up approach. The top-down approach, known as a synthetic training approach, aims to improve the efficiency of central processing [4]. Participants are trained to develop active listening strategies (e.g., attention to lexical or contextual cues). This approach targets higher levels of auditory processing and selectively uses lower-level auditory processing as needed. For example, the connected discourse tracking used by De Filippo and Scott [5] and Rosen et al. [6]reflects a top-down approach.

Analytic training is termed a bottom-up approach because the intent is to improve the overall speech comprehension by focusing on the acoustic parts of speech messages. The reason is that if someone can reliably distinguish the acoustic elements of speech, then he or she should be better able to comprehend the larger units, such as sentences and paragraphs.

The listening hierarchy

Hearing is viewed in terms of four different levels of perception in a hierarchy, which was already introduced by Erber (Figure 2).

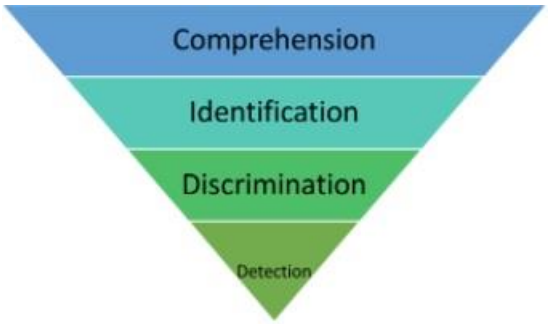

Figure 2: Erber's hierarchy

Detection is the most basic level of perception. It involves being able to notice and perceive the presence or absence of auditory stimuli, and it stimulates the exploration and discovery of the world of sound. Localization is the ability to detect the location of a sound source. This is an important topic in the case of bilateral stimulation. **Discrimination** is the perception of differences and similarities between sounds. Environmental sounds, phonemes, and words are all examples of different types of sounds that can be discriminated. In a discrimination task, the CI user must indicate if two or more items (such as /t/ and /k/ or /bi/ and /bo/) are the same or different. **Identification** of sounds is the ability to repeat or point to the stimuli heard. It involves the ability to match the perception of a sound to its actual source (or a picture of its source) and also to associate a speech sound (i.e., phonemes, words, or phrases) with a larger context of language. **Comprehension/Interpretation**, also called processing or comprehension, is the ability not just to hear the auditory stimuli, but also to understand their meaning. Understanding the content and meaning of a sound, phoneme, word, phrase, expression, sentence, or a larger utterance is central to interpretation [2]. A good interpretation level stimulus, then, is one that requires the CI user to respond in a way to show that it was correctly processed or comprehended (e.g., following directions), giving a specific response, or summarizing something in their own words. These responses would indicate understanding that goes beyond just hearing.

Listening to different sounds

When developing listening skills, you not only learn to listen to different environmental sounds, but also to different speech sounds. Sounds can consist of non-verbal (environmental sounds, music instruments, music etc.) and verbal speech sounds (onomatopoeia, phonemes, words, phrases, sentences, text etc.).

Prosody also plays an important role in the learning to listen. It refers to the rhythm, stress, and intonation in connected speech and includes supra-segmental (rhythm, duration, pause, and pitch) and segmental (vowel, diphthong, and consonant) characteristics [7].

Listening to different sound sources

Auditory habilitation should not only be conducted in ideal listening conditions (a quiet environment, 1 M distance, or good speech intensity) but also under conditions that more closely resemble everyday-life situations; that includes background noise, larger distances, and people who whisper. So, habilitation can be conducted while the person is wearing his/her own listening device, such as cochlear implant(s), or a CI and a conventional hearing aid, as well as by using assistive listening devices such as Bluetooth devices,

FM-systems, infrared systems, or T-loops. Purely auditory signals, purely visual (speech reading), or a combination of both can be used. Speech reading adds the visual component (including eye contact and gestures) into the auditory habilitation and makes listening easier. You can do the listening activities with the child in a closed, open, or semi-closed set. In a closed-set activity, the child practices with a limited number of items (known by the person) via a set of objects, photos, drawings, or a list of written possibilities. In a semi-closed set activity, the person does not know exactly which items are being presented. Information is given about the stimuli to assist the person prior to the start of the exercise, for instance, by agreeing on a topic, theme, or a rule for the first few letters or phonemes of the possible responses. In an open-set activity, which is of the highest difficulty level, the person does not know the names of the items or anything about them beforehand. This would mean that there may be an unlimited number of possible responses.

Auditory habilitation always starts with large contrasts. These are large differences in sound characteristics that are more easily perceivable, and the degree of difficulty becomes bigger by reducing contrasts [7]. Listening activities can be done in person (live) or by using the radio, TV, computer, tablet, mobile phone etc.

When creating listening activities with a child using a CI, the difficulty level should always be kept in mind. Here they are listed, from the easiest to the most difficult:

- From closed to open-set
- With and without speech reading
- Without and with background noise
- From a short distance to a longer distance
- From easy to difficult practice material (from large to small auditory differences, from short to long utterances, from an easy to a difficult language level).

The Listening Cube

The Listening Cube (Figure 2) is a three-dimensional auditory program, developed at the KIDS Centre for the Deaf in Hasselt Belgium [7]. It helps to combine the listening hierarchy (dimension 1), with different sounds (dimension 2) and different sound sources (dimension 3). It also alerts the therapists to increase their expectations and to allow improved perception of soft sounds and speech, improved speech understanding in background noise, as well as localization of sound and speech [8].

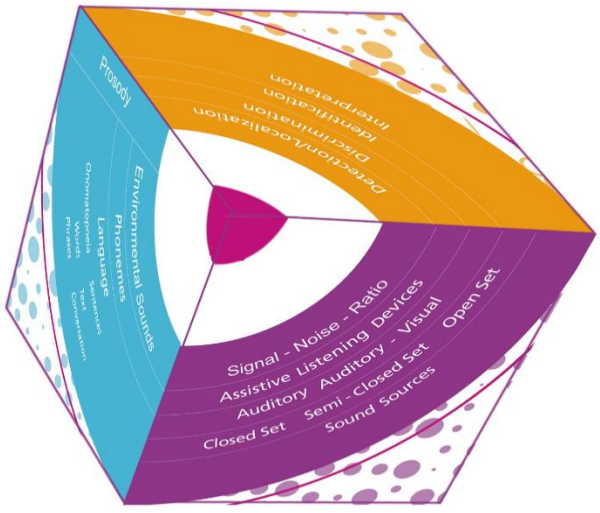

Figure 2: The 3 dimensions of the KIDS Listening cube (De Raeve et al, 2012)

The Listening Cube is a tool for planning therapeutic sessions that are both challenging and appropriate for CI users' individual needs. The three dimensions of the cube—levels of perception, practice material, and practice conditions—can serve as a visual reminder of the task analysis and other considerations that play a role in structuring therapy sessions.

The Ling 6 sounds

The Ling Six Sounds Test provides a well-established means for determining the audibility of the speech signal at any time and in any situation. When a child is able to detect and/or identify all six sounds (ah, oo, ee, sh, ss, mm), presented with a natural, unforced vocal effort, the parent/professional has a good indication that sound energy across the speech spectrum is available within the given distance of instruction [9]. The sounds used were selected because they encompass the entire speech spectrum (Figure 3).

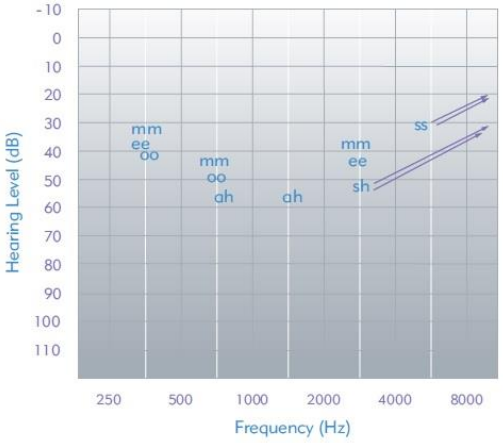

Figure 3: Ling Six Sound test (AB, 2016)

The test can be assessed at the detection level ("I can detect the sound") or at the identification level ("I can identify the speech sound"):

- Detection Task: elicit a conditioned response to the sound, e.g., putting a peg in a pegboard.
- Identification Task: have the child point to pictures or repeat what is said.

This simple test should be performed routinely throughout each school day, e.g., before each small group or one-to-one instructional situation.

Recently, Madell & Hewitt (10) upgraded the Ling Six Sound test into the Ling-Madell-Hewitt (LMH) Test Battery or Ling 10 sound test. They added four additional consonants (/z/, /h/, /n /, and / dʒ) to the 6 Ling sounds to provide additional information about mid-frequency perception.

Learning to listen and to develop spoken language

To stimulate listening and spoken language development in young deaf children receiving cochlear implants, we have to focus on 6 goals, which parents usually learn quickly if the therapist coaches them in using various strategies.

Create a listening environment

Once a child has access to sound, it is critical to help him/her develop focused auditory attention, so the child learns to listen as quickly and efficiently as possible. Listening improves when the child is quiet and attends to the adult who is talking [11].

The closer you talk to the microphone of the child's hearing device, the more easily the child will understand the speech signal. The best understanding of speech occurs in a quiet room and when the person is talking between a 1- and 3-meter distance from the child's hearing device [12].

Facilitate auditory attention

In order to develop a listening attitude, the child's attention must be directed to focus on sounds (e.g., "Listen! Hey!") and adults need to expect that the child will pay attention to what people say. These auditory hooks direct attention to the environmental sound or to the spoken language, which creates a longer time of shared and joined attention.

Auditory attention can also be drawn by listening first and last to the presentation of information. The child first listens to the spoken language (even two or three times), and if the child does not understand the spoken message after three presentations, then add a visual cue. Once the child understands the message, always return to the auditory-only presentation [13].

Enhance auditory perception of speech

We are able to enhance the auditory speech perception of the child by using **infant-directed speech (IDS)** or motherese. This is a higher-pitched, repetitive, rhythmic, and slower speech mode (e.g., "helll-llloo"), which adds several additional acoustic cues to speech and increases speech understanding.

Vocal play can enhance the auditory perception of speech. Adults need to reinforce and imitate their child's vocalizations and should vary their own vocal play in order to facilitate speech perception and production [14].

Sound-object associations help infants to strengthen their auditory focus, develop an auditory imprint for specific sounds, recognize different sounds and phonemes, and maintain their interest while enhancing speech perception. It also promotes language comprehension because the child learns to associate meaning with each sound [12].

Babies and toddlers enjoy **whispering and singing**, even deaf children wearing cochlear implants. Both are important strategies to enhance speech perception.

Additionally, stressed morphemes, syllables, words, or phrases (**acoustic highlighting**) enhance speech perception. They are longer, louder, and have a higher pitch than those that are unstressed and help the child to hear particular features of language more clearly.

Pre-verbal communication skills

Before babies learn to talk, they communicate in a variety of non-verbal ways. The most important are: eye contact, shared attention, imitation, and turn taking.

Around the age of four months, we see a change in **eye contact**. The baby slowly starts to track you or even an object with his/her eyes and starts pointing at it. In fact, this is the first way the child takes turns. From the age of six months, we see that the child is able to look at an object that the parent is looking at. This milestone is often referred to as "shared" or "joint" attention. It forms the basis for spoken language development since he/she can then make the connection between the object and the words he/she hears from you.

When parents communicate with their child, they look where the child is looking or at the object of the child's interest. Afterwards, they usually start talking about whatever it is they are looking at together. Talking about things the child is looking at teaches the child to understand the words being said. We also call this "**shared" or "joint" attention** because, in these moments, the parent and the child are both

focused on the same thing. Later, the child may even point at the object, either to confirm that the adult is also looking at it or to request that the adult look at it. What is important here is that the child hears your voice clearly. He/She can look at the object you are talking about while listening to you. If he/she does not respond to you talking, you can bring the object in the visual field between you two or use gestures to point out the object. The child not only reacts to what the parent says but can also take initiative himself/herself to communicate. The child's ability to point to an object in a communicative way is a crucial aspect in the development of communication. The parent responds by labelling the object ("ball") or action ("bouncing") that the child is pointing out.

Now that your child can hear with the cochlear implant, encouraging and playing imitation games are important first steps towards developing "first words" or language**. Imitate** the sounds your child is making.

Good communication assumes that it is not always the same person who is talking, but that both parties (parent and child) take turns saying something. Even very young children that do not speak yet can take their turn by pointing at something, smiling, or crying. You should react to this as if your child spoke. When it is his/her "turn", you may need to wait a bit, so he/she has enough time to react.

Taking turns like this teaches your child how a conversation works: one person communicates while the other awaits and then the next time, the roles reverse. Children are able to start taking turns by the age of three months; turn-taking at this age may not involve vocalizations. Verbal turn-taking will initially happen by making vowel-like sounds (ie, oe, eeh, ooh, aah, ae, uuh) and later, with consonants (such as d, b, m) [15].

Stimulate incidental learning

Normal-hearing children continue to learn speech and language all day through structured and unstructured activities. Incidental learning refers to the acquisition of language skills occurring through auditory information derived from the surroundings by overhearing conversations and speech while at home, in the classroom, or in/at the playground for example.

Incidental learning of spoken language requires excellent auditory skills. Because of the reduction in signal intensity and integrity with distance, a hearing-impaired child may have a limited range or distance of hearing, which subsequently reduces the amount of auditory information they are exposed to on a daily basis relative to the normal-hearing child and, as such, misses out on the development of incidental learning skills [16]. Children need good auditory conditions to stimulate and develop the cognitive skills needed for incidental learning: they should be able to detect and discriminate the acoustic signal. To enable incidental learning, the ideal acoustic environment for hearing impaired children would have minimal background noise or reverberation with a Signal-to-Noise Ratio (SNR) of +15 dB, i.e., the speech is 15 dB louder than the surrounding noise level [17].

Binaural hearing

While the specifics of the (re)habilitation programme including auditory training do not vary greatly from that used with individuals using unilateral stimulation alone, there are some unique considerations for training binaural hearing skills. In comparison to unilateral stimulation, the main benefits of binaural stimulation are related to enabling binaural hearing to develop further to allow improved perception of soft sounds and speech, improved speech understanding in background noise, as well as localization of

sound and speech. It is therefore important that the auditory training programme tailored specifically for the bilaterally stimulated child should involve listening exercises which include auditory tasks that train binaural hearing skills, ultimately using conditions that more closely represent listening conditions in daily life. The following list includes a range of listening activities that can be incorporated into a long-term rehabilitation programme for the bilaterally stimulated child.

Softer sounds and speech

It is recommended that sound stimuli be presented from one meter away and in front of the child at a conversational level (i.e., approximately 60-70 dBHL) using either live voice or recorded stimuli. Due to the loudness summation effect of receiving sound stimulation to/by two devices simultaneously, the child is better equipped to learn to perceive and recognize softer sounds and speech compared to using only one device. As an example, the sound stimulus level may be gradually reduced by decreasing the volume or by increasing the distance of the stimuli presented to the child. Training may be performed ideally in larger spaces, indoors or outdoors, enabling the therapist to adjust and modify the training exercise according to the child's performance level.

Speech perception with background noise

In the case of a child receiving bimodal or bilateral stimulation, it is very important to provide listening practice in more difficult listening situations than would be used with a unilaterally implanted bilaterally deafened child for example. Training environments could include rooms with standard acoustics in the presence of various background noises, or outside spaces that more closely represent the daily listening situations. Furthermore, increasingly difficult listening environment options to train binaural listening skills can be created by choosing a room with a high level of reverberation, such as a hard-walled gymnasium, or by moving to a noisy playground, or creating background noise indoors by using a radio or a CD player at various loudness levels.

Localization

Identifying the direction of a sound stimulus is another aim of training binaural hearing skills for the bilaterally stimulated child. While this may be automatically acquired for the short-term deafened child receiving bilateral stimulation, this is not the case for the older child with a considerable time interval between the implantation of the first and second devices. Hence, localization of sound stimuli from multiple directions is a binaural hearing skill that needs to be practised during auditory training over time. This can be done by the therapist but also at home with family and friends. To train localization skills, different stimuli may be used, such as: environmental sounds, musical instruments, speech sounds, or conversational speech.

General rules:

- the shorter the interval between implant surgeries, the quicker the inter-aural performance equivalence is observed;
- the younger the child at second implant, the lower the need for individual second ear therapy;
- the older the child at second implant, especially in the case of good performers, the higher the need for individual second ear therapy.

Listening in the classroom

The typical classroom presents a difficult listening situation for a child with a hearing impairment. Classrooms tend to be very noisy places, especially in preschool. The level of background noise within the classroom setting is often louder than the teacher's voice by up to 15 dB creating a SNR of –15 dB (18).Such an environment is considered sub-optimal for many children with even a mild hearing loss, and highly undesirable for children with significant hearing impairment. Furthermore, children are often seated at a distance of 3 meters or more from the teacher. The greater the distance between the child and teacher, the greater the decrease in the quality and intensity level of the teacher's voice at the child's ear(s). For characteristically practical reasons, classrooms are full of hard-surfaced walls and furnishings and are prone to a high level of reverberance (i.e., reflection and echoes) further reducing the quality of the acoustic conditions and leading to distortion of the teacher's voice.

For normal-hearing children, the use of two ears in such a sub-optimal acoustic environment enables them to distinguish the teacher's voice from the surrounding noise. Therefore, to increase the potential to cope in such an acoustic environment, ideally the hearing-impaired child should also be provided with auditory stimulation to both ears.

Further improvements in the listening conditions can be created by keeping the distance between the teacher and the hearing-impaired child to a maximum of one meter, together with the use of carpets and soft furnishings to reduce the level of reverberation and background noise.

Additionally, to further improve the listening conditions in the classroom for the hearing-impaired child, the use of assistive listening devices, such as an FM-system, (comprising a Frequency-Modulated (FM) radio transmitter worn by the teacher and a receiver worn by the child) in combination with the child's cochlear implant(s) or hearing aid(s) can, be considered. Through the use of an FM system, the level of the teacher's voice is enhanced while the level of the surrounding sounds is reduced, leading to the potential for improved speech comprehension ability for the child. Essentially, the SNR is improved relative to the situation without the FM-system.

The decision to use a FM-system on one ear versus the other or with both ears combined via connection to acoustic or electrical hearing devices should be determined individually on a case-by-case basis and the FM-system adapted accordingly.

For the simultaneously implanted child, the FM-system may be connected to both CI implants to balance the input to/in both ears. Similarly, in the case of the bimodally stimulated child, the FM-system may be connected to each device. In the case of the sequentially implanted bilateral CI user, in our experience, the continued use of the FM-system with the first CI ear is usually recommended. Following fitting of the second CI, when the sound processor programme is stable and the child begins to demonstrate speech identification skills through the second ear, FM-system use is encouraged with both implants in combination. In contrast, some clinicians may prefer to initially recommend FM-system use with the second CI ear alone, as a way of strengthening the signal delivered to the second ear relative to that perceived by the experienced first CI ear. To date, there is no evidence to suggest one or the other method of FM-system use following sequential implant provides superior or quicker outcomes.

Music

Speech and music share features such as rhythm, intensity, timbre, and pitch. However, the pitch spectrum in music is much greater than in speech. Access to these pitches of music helps us to hear birds, sirens, and other important sounds in the world. Musical activities can even drive neural networks to function with higher precision than needed for spoken communication.

Music activities can stimulate listening skills, speech production, and language development. Separate from these skills, other domains such as socialization, cognition, and emotional-behavioral development can be stimulated by music. The type of activity and responses required should reflect the current developmental level of each child and help the child to move gradually toward the next level.

Goals of listening, speech, and language can be addressed through playing instruments, movement, and singing. Out of all musical experiences, the voice is the instrument that is most accessible and most related to talking. Because singing resembles speech in production, but with greater inflection, the link between singing and communicative goals is obvious. Singing requires coordination of the same vocal mechanisms used in spoken communication and singing along with others involves careful listening.

The vocabulary and concepts found in children's songs are rich sources for linguistic development [19]. One benefit of using songs to promote speech and language development is the inherent repetition of vocabulary.

Summary

Hearing babies begin to pay attention to the spoken language of their environment as soon as they are born. As parents choose a cochlear implant for their deaf child, they also choose to learn to listen and to develop the spoken language of their environment [20].

There are different communication approaches for children with a hearing loss, from only auditory to only visual communication. When parents choose a cochlear implant for their child, they choose to stimulate listening and spoken language. Auditory habilitation is the creation of a listening and learning environment in which parents, therapists, and educators help children with impaired hearing to acquire listening and speech. The Listening Cube is an auditory training approach for children with a hearing loss, consisting of 3 dimensions: the levels of perception, the practice material, and the practice conditions.

Speech and music share features such as rhythm, intensity, timbre, and pitch. Access to these pitches of music helps us to hear birds, sirens, and other important sounds in the world. Music activities can stimulate listening skills, speech production, and language development.

Chapter 8 - Family Centered Intervention

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"An attitude of compassion does not mean looking down on someone, pitying them in their misery. Compassion is based on respect. We discuss life as equals, learn from each other and strive together to improve our lives" Daisaku Ikeda

Introduction

The existence of profound deafness in early childhood has an impact on children's development and important consequences for the child, his family, and society in general. It is well known that there are critical periods for speech and language development during which the developing nervous system is particularly responsive to auditory stimulation. Inadequate sensory input during these periods leads to lifelong linguistic and communicative deficits. Most hearing losses can be satisfactorily managed with hearing aids and rehabilitation. Until the advent of the cochlear implant, however, the prospects for children too deaf to hear speech through a hearing aid were less promising and often failed to develop intelligible spoken language. However, we assisted to a revolution with the emergence of very sophisticated and powerful hearing aids, namely cochlear implants [1,2]. These advances meant the need to change the paradigm in the care of these children and their families. Auditory Verbal Therapy (AVT) represents this change par excellence and bases its proposals on increasingly robust scientific evidence and particularly on evidence of very well achieved and thoughtful clinical practices. The family becomes our main target of action, and the work developed is to equip each family with the knowledge and strategies that allow their child to develop to his/her greatest potential. The ultimate goal is the full inclusion of all those with hearing loss, with levels of social participation equal to those of their hearing peers [3,4].

Throughout this chapter, you will find the fundamentals of coaching in working with families as well as some work processes that allow the greatest possible involvement of these children and their families. It also addresses the challenge presented to the professionals who work with them and some conceptual references that underlie this "lifestyle" model.

"Parental coaching plays a central role in intervening with families".
What role should the professional play in order to ensure family involvement in therapy?

Coaching strategies for parents of children with cochlear implants

Coaching

Coaching is defined by Rush & Shelden (2019) [5] as "an adult learning strategy that is used to support the parent/teacher (coachee) in identifying, obtaining and mobilizing the knowledge and skills necessary to achieve an intended outcome. It promotes the ability to reflect on his or her actions as a mean to determine the effectiveness of an action or practice and to develop a plan for refinement and use of the action in immediate and future situations".

Rush & Shelden (2019) define five key characteristics of coaching:

1. Joint planning: agreement on the actions they will take or opportunities they will have to practice between coaching visits.
2. Observation: examination of another person's actions or practices that can be used to develop new skills, strategies, or ideas.
3. Action/practice: spontaneous or planned events that occur within the context of a real-life situation and that provide opportunities to practice, refine or analyse new or existing skills.
4. Reflection: analysing of existing strategies to determine whether they are consistent with evidence-based practices and whether these strategies may need to be implemented without change or modified to obtain the intended outcomes.
5. Feedback: information provided by the coach that is based on his/her observations of the coachee, actions reported, or information shared. The aim is to expand the coachee's current level of understanding about a specific evidence-based practice or to affirm the coachee's thoughts or actions related to the intended outcomes. Video interaction guidance is an evidence-based means of coaching for optimal interactions between parents and children with a hearing loss.

A recent scoping review, elaborated by Noll and collaborators in 2021, examined how parental training and coaching is conceptualized and implemented in the listening and spoken language practice. In describing the impact of caregiver coaching, it suggested that families of children who are DHH can effectively learn to implement specialized communicative strategies through coaching [6].

In addition, Giallini et al. [7] highlighted in their systematic review on parent coaching the great potential of parent coaching, to promote learning and change in the parents of children with a hearing loss, driving them toward responses and behaviours that are more facilitative of their children's communicative development. Parents should be considered the primary agents of change engaged in the promotion of their children's development, and this is particularly true for early communication and language development, where the quality and quantity of adult language input strongly influences language growth

in both young, typically hearing children and in children with a hearing loss. Within the general framework of family-centred practice, to train the parents of children with a hearing loss to use communicative and language facilitative strategies should be considered a key component in early intervention programs.

Coaching families and teachers

Parent coaching can promote parents' confidence and competences in supporting child learning and development. The role of the coaching is to identify what the parents already know and what they are doing in relation to the child's development, to share new information and ideas, and to work together to support and promote opportunities for learning. Parent coaching is also useful to support parents in identifying, assessing, and evaluating needed resources for their child and family.

Coaching can also be used with teachers in childcare settings, preschools, or early childhood intervention programs. Within these environments, coaching may be used as a follow-up to professional development activities to help teachers incorporate new skills into their current teaching practice.

Parents have a central role in intervention of children with a hearing loss

When utilizing a coach in deaf education, the positive outcomes for parents or professionals supporting children who have a hearing loss are abundant. The coach in a deaf education setting could be a teacher of the deaf, an audiologist, a speech-language pathologist, or other professionals with knowledge of this topic. When all of these professionals come together, they become part of a family-centred team. The family-centred approach is commonly used in early intervention, a service provided to families of children from birth to three-years-old who have special needs [8].

It is likely that parents and other professionals (who are coached about relevant and useful information) are able to learn, retain, and apply the given information when it is close to where they are. Developing a partnership sets the stage for facilitating learning through the implementation of best practices in coaching. This coaching model provides interactive learning sessions in which the parents or other professionals have the opportunity to practice the needed skills with the coach there to guide them. This approach not only helps the adult learn the techniques and skills, but it also builds confidence in the adults regarding their ability to help the child succeed.

Coaching provides opportunities for supported practice in real-life environments when it is tailored to the specific needs and settings relevant for each family. When parents and other professionals are given positive but honest feedback, those adults will be able to reflect on their strengths and think about how to improve their skills. As reflective practice becomes an interactive procedure between the coach and the adults, collaboration is strengthened, and coaching will become more effective. Fostering reflective practice demonstrates the real meaning of life-long learning and embodies the concept of the growth mindset. The more one is able to reflect on their performance, the more self-directed the learning will be [8].

Parent coaching in Auditory Verbal Therapy

Doreen Pollack, one of the pioneers of Auditory Verbal Therapy (AVT), urged practitioners to coach parents in creating the right learning environment, one in which their child, who has a hearing loss, could grow up in a world with sounds, in which the environment believes the child could hear, expect the child

to listen and respond in a relevant and meaningful context of daily experience. Parents have to learn strategies that promote the child's development of listening, talking and thinking skills so that they match the child's needs and real-life situations in an individualised manner, and they also have to learn how to integrate listening into their child's being so that listening becomes a way of life [4].

Parents develop confidence in the planning, delivery, and evaluation of strategies that promote learning in children with a hearing loss. This confidence has been identified as a significant variable that influences outcomes in listening, spoken language, and cognitive skills of children with a hearing loss. The concept of coaching alongside the parent creates more opportunities for promoting the development of listening, spoken language, and learning than practitioner-led intervention. Even the strategic placement of the adults in an AVT session stimulates the development of spoken language through listening. Successful parent coaching in AVT is the work of the practitioner who knows how to use strategies to stimulate listening and spoken language, and how to coach the parents in ways to help them acquire confidence and competence in stimulating their child's listening and spoken language.

Throughout the process of coaching, the practitioner remains ever mindful of the many stressors on the family and always approaches coaching with kindness by paying attention, showing patience, communicating respectfully, and expressing compassion and concern for others. Parent coaching is also a dynamic process. Each individual session, situation, or family presents a number of human variables that can determine how the coaching components occur [2].

The family's involvement in the process an Auditory Verbal approach

Rotfleisch and Martindale [9], in their last book (2022), dedicate the entire second chapter to the work to be developed in order to effectively involve the family as a partner in a verbal-auditory therapy perspective. They point out some important aspects and steps to working with families of children with hearing loss: how to plan each session, how to give emotional support to families, apply for an ecological transition in an andragogic model of teaching adults, intervention-based in activities of daily living (ADL), and taking into consideration the cultural background of each family.

Planning is a mandatory starting point for each session. Professionals should review typical and atypical child development and prepare subtle take-home messages that are precious day-to-day "states of mind" for parents (e.g., informative post-its on the refrigerator door). Instead of doing "lectures for parents" or academic/scientific long explanations, professionals must model target behaviours or strategies, and allow parents to experiment by themselves, turn over the control of the sessions' activities to them and praising all their efforts and achievements. It is important to rebuild parental self-confidence as parents and first promotors of child development. In order to guarantee effective support for parents, we need to master all the fundamental aspects of teaching adults, rescuing what Knowles called *andragogy.* Adults learn differently from children and are in great need of direct-action models, directed to their inner needs and motivations and rely only on solving their problems. They do not need classes or scientific findings *per se*. All information provided must meet their specific needs. Academic knowledge should only be the pillar of security and function as a resource for the professional's performance with families [10,11].

Each family represents a particular cultural identity, and the impact that the diagnosis of deafness has on your child is not the same for all. We need to be able to provide each of them with the necessary emotional support so that they believe in and invest in their fundamental role with their child. These needs vary throughout the child's life and sometimes become more acute during transition periods in their academic

life. In a model of auditory verbal therapy (AVT) we witness a change in the roles usually assumed by parents. They are no longer mere spectators of the professionals' expertise and must take a prominent place in your child's education. The information left at the end of the sessions, or the work requested to be done at home makes no sense. Parents are an integral part of planning and running the sessions. We are facing what Neuss calls an *ecological transition*. They assume new roles in addition to the ones they have or are expected to have, and our partnership is very important in your training and guidance. They must be guided on what they are supposed to know and know how to do when they take the front line in child's development, advocate for their child's rights and become primarily responsible for their inclusion in mainstream classes in schools. In Neuss words "It requires significant long-term parental participation and has been described as a "way of life", (Goldberg & Flexer, 1993) whereby parents become so familiar with the techniques learned in therapy that they employ them with their children during the course of their interactions throughout the day" [12,13].

Following the precepts of the 10 AVT (Auditory Verbal Therapy) principles, we were able to plan our intervention centered on a deep understanding of the impact of hearing loss on each child and their families, their routines, activities, and preferences. Guiding and coaching each member of the family in our intervention requires a very specialized training as well as accurate planning from the professional who is sensitive to the particularities of each one. [14,15]

Our starting question should be: why is it important for parents to be part of the session and intervention plan?

First of all, because they ensure, or can guarantee the continuity of stimulation of the child's development throughout the day. In other words, they ensure 24/7 learning for their child. This is not possible if parents and professionals do not work together. In the work of Moeller and Holzinger [8,16,17], we can find evidence that parental involvement is a significant predictor of good language development. Following the ADL formula suggested by Rotfleisch & Martindale we can easily illustrate the number of good repetitions of meaningful language examples that can be found in the routines of daily life:

R = Repetitions per day
N =Number of days activity performed
P = Potential language repetition within ADL
X = Exposure to language Structure
R x N x P=X
For example, mealtimes:
Activity – meals R = 3
Number of Days per Year N = 365
Language Repetitions P=10
3x365x10 = 10,950
(example extracted from *Listening and Spoken Language Therapy for Children with Hearing Loss- A practical Auditory-based Guide,* p. 62)[9]

On the other hand, the involvement of parents as partners is the only way to respect their culture. "Culture as abroad construct, involves ethnicity, religion, language, skin color, socioeconomic status, age, gender, orientation, and wholeness/disability status as well as family structure and dynamics."(in Estabrooks et al., 2016, chapter 1, p13)[4]

Revisiting the conceptual field created by Moses (1994) for professionals who work directly with families, we find the anagram ENUF (E= Empathizing N=Nonjudgmental U= Unconditional F= Feeling Focused), a very useful tool to guide our practice [18]. What does this mean? It means standing by the family's side; supporting them in their grief for the perfect child; understanding exactly what the other is feeling at any given moment; allowing their feelings because they are our equal; not judging the feelings and/or actions of others and remaining neutral regarding what is right or wrong for the family; being focused on the principle of establishing mutual trust; and being aware that parents trust us to share very strong feelings, whatever they may be. We need to step out of our comfort zone, being very knowledgeable and experienced (and this implies a lot of supervision throughout our practice) and assume that no matter how great the cultural difference with certain families is, they will always recognize our compassionate effort. Practical examples are, for instance, using foods that the family eats and not ours; learning and respecting the rules of communication within the hierarchy of each family culture; continuously reading each parent's emotional level; and accepting times of depression or exaggerated anxiety in trivial aspects of everyday life (such as starting swimming lessons or organizing a birthday party). Allow them to meet other families in same journey. Allow them enough space and time to decide if they want or not to follow our proposals, because they are facing so many challenges and decisions to make about schooling, hearing technology, socialization, other child’s needs, and issues throughout life.

Chapter 9 – Intervention Models and Framework in Speech and Language Therapy

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"Coming together is a beginning, staying together is progress, and working together is success." Henry Ford

Introduction

The speech therapist's performance over time has undergone immense variations and has been influenced by different intervention paradigms. The work developed by Judy Duchan is an invaluable resource for us to understand our roots and fields of activity since ancient times [1].

Through the study of her historical research, we realize that we are in the Era of the Pragmatic Revolution where the main objective is to achieve the most developed and appropriate social performance possible and not so much non-functional and merely technical performances. In this historical evolution, the influence of different conceptions of intervention on children with hearing loss is notorious. As Judy Duchan herself states:

"*The most recent period, from 1975 to the present, saw the emergence of a new type of clinical practice, that having to do with pragmatic concerns such as how messages were used, and how they fit into the situational and cultural contexts of everyday life communication. New therapies were developed, in keeping with the notion of communication in context. Rather than conducting practice sessions in contexts separate from the client's ordinary communication situations, the clinical services were delivered in classrooms, homes and in community settings*." [1,2]

We have moved from an intervention that aimed to develop speech skills as the most important thing, the understanding of hearing loss as separate from cognitive functions and the almost absurd lack of knowledge of its impact on the global development of children. Our work was completely separate from schools, parents, families, and the contexts in which each one grew up. The basic model of action was the medical model, and the objective was to "treat" the pathology and not accompany the person who had it.

The transdisciplinary practice, integrated in an in-depth knowledge about hearing loss, its medical precepts, technologies to support listening, the typical and atypical neurodevelopment of children, the different teaching and learning methodologies, practiced only a few decades in speech therapy and in Auditory Verbal Therapy (AVT) is considered the maximum exponent of a biopsychosocial model of action, currently advocated.

Point out the major importance of promoting education in regular schools with peers who have typical hearing and with appropriate services from early childhood onwards.

Auditory Verbal Therapy

The Auditory Verbal approach started in 1978 as result of the efforts and knowledge of Daniel Ling, Doreen Pollack, and Helen Beebe. Since then, several professionals in the fields of medicine, education, psychology, and speech therapy have contributed to the evolution of this approach, and their clinical research has allowed to cement all the pillars of knowledge that underlie Auditory Verbal Therapy as we know it today. The work developed by the AG Bell Association in promoting the path taken and the international certification in AVT, ensuring quality practices and a uniform level throughout the world should be highlighted. Ellen Rhoades and Warren Estabrooks are two major examples of the diffusion of these practices, certifying speech therapists and teachers of the deaf around the world, and spreading new trends in Listening and Spoken Language [3–5].

Obviously, the entire technological revolution in bioengineering related to technical aids to support listening, as well as advances in medical genetics and medical imaging, have allowed unprecedented progress in intervention for children with hearing loss and their families. What was previously assumed by practice is now known objectively.

The AVT program advocates a certain philosophy of life for both families and professionals who accompany them. It is therefore guided by 10 principles. They are [3,6,7]:

1. Promote early diagnosis of hearing loss in newborns, infants, toddlers, and young children, followed by immediate audiologic management and auditory-verbal therapy.
2. Recommend immediate assessment and use of appropriate, state-of-the-art hearing technology to obtain maximum benefits of auditory stimulation.
3. Guide and coach parents to help their child use hearing as the primary sensory modality in developing listening and spoken language.
4. Guide and coach parents to become the primary facilitators of their child's listening and spoken language development through active, consistent participation in individualized auditory-verbal therapy.
5. Guide and coach parents to create environments that support listening for the acquisition of spoken language throughout the child's daily activities.
6. Guide and coach parents to help their child integrate listening and spoken language into all aspects of the child's life.
7. Guide and coach parents to use the natural developmental patterns of audition, speech, language, cognition, and communication.
8. Guide and coach parents to help their child self-monitor spoken language through listening.
9. Administer ongoing formal and informal diagnostic assessments to develop individualized auditory-verbal treatment plans, to monitor progress and to evaluate the effectiveness of the plans for the child and family.
10. Promote education in regular schools with peers who have typical hearing and with appropriate services from early childhood onwards.

As we can easily see, everything is based on the existence of newborn hearing screening implementation. This tracking is increasingly a reality, although there are countries that have not yet adopted it. This must be done at birth and/or after the first two weeks of life. It is vitally important in signaling and monitoring

all children who do not respond to sound. Negative results in this screening imply a subsequent and immediate audiological follow-up and the use of hearing aids or other listening technology. There is a lot of evidence of the advantage of this audiological detection and control as early as possible [8–13]. There is a report of guidelines for good practices in early hearing detection and intervention that support what Carol Flexer called *neurological emergency* [14]. This report, elaborated by the Joint Committee on Infant Hearing, highlights the need to follow the 1-2-3 formula: detection in the first month of life, audiological adaptation in the second and guarantee of auditory-verbal intervention from the third month [15].

From principles 3 to 8, we are alert about how, when, and how much it is important to guide and coach families. We find the expression *guide and coach parents to*. Throughout these principles, professionals are called to pay attention to the need for their work to address parents directly and lead them to create a good listening environment, lead the child to be responsible for their hearing and assistive technology, and to consider hearing as a first way of acquiring and developing knowledge and skills and advocating for the rights of their children along their journey.

Principle 9 enlightens the need to continually obtain diagnostic information (in sessions and from parents and teachers) and to carry out specific formal assessment times as well. Close monitoring of the areas of hearing, speech, language, cognition, and literacy is essential to ensure a harmonious development at the level required by the school and society in general.

The last principle represents the culmination of all the others, that of the possibility that each child has to attend classes in a mainstream school with academic success. Working from an auditory-verbal perspective implies educating the child to listen in all environments, but it also means fighting to ensure that optimal acoustic conditions and human and technological resources are guaranteed to these children. The collaboration between all specialized professionals, such as teachers of the deaf, educational audiologists, speech and language therapists, mainstream classroom teachers, among others, creates a net who allows full opportunities and access. This is what inclusion means: providing the means, and equipping the physical, educational, and social contexts so that everyone has their degree of participation in society guaranteed [16,17].

From Assessment to Intervention: Some Foundational Pillars of AVT

Assessment is an essential component of good AVT practice. Traditionally, the assessment of children's communication skills boils down to establishing the articulatory profile and the levels of language comprehension and expression. In AVT therapy, evaluating is much more than that! It includes all the parameters mentioned and also informal and formal tools that evaluate and monitor the children's development progress in an integrated way in the areas of hearing, communication, speech and language, cognition, play and literacy.

The professional must also be able to obtain information about the family's needs and culture.

The assessment of the child and his/her family starts at the first meeting between all of them. In this session, the professional must ensure information about the (1) child's developmental level, regarding the references of neurotypical children in the areas of hearing, language, communication, must be able to (2) establish long-term goals in their individual intervention plan and must obtain the (3) line of basis to monitor progress. This progress is evaluated on an ongoing basis and is based on the diagnostic

information obtained in each session. That is why it is called *ongoing assessment* [18]. The AVT sessions are diagnostic sessions in which the professional collects information from informal assessment and observation in order to be able to adjust his intervention objectives/strategies/activity whenever necessary (and may need to change some objectives set for the session if the child and the family demonstrate that this level is already acquired). This is providing a better and more accurate, authentic profile of child´s abilities and competences.

AVT session

AVT sessions are individual, personalized, and purpose-built for that child in his/her family, within his/her culture. AVT is not group therapy and it is not a simple exercise prescription completely out of the child's life and needs.

In this way, each session is unique to the children and families for which it is designed and therefore there is no one session formula that fits all. On the other hand, each professional is unique in their knowledge and experience and in their AVT practice, of course, they bring a lot of themselves to each session, as well as their beliefs and attitudes [19]. It is why Warren Estabrooks called AVT practice an art! Often the therapist finds that the session plan is falling apart and has to activate the alternative plan all in order for it to be meaningful for the child and his or her family. In AVT, the therapist is experienced in expecting the unexpected and must develop art in order to save the session, find solutions in order to "command" again. It's why Warren Estabrooks called AVT practice an art! [20,21]

Each AVT session is planned according to the overall intervention plan. This should be reviewed every 3, 6, or annually, depending on the child's level of progress. Family-only sessions are also provided for the assessment and monitoring of progress, as well as for guidance and counseling on specific monitoring matters or aspects.

In fact, in each AVT session, the professional aims to create a listening environment; facilitate auditory processing; enhance the auditory perception of speech; promote linguistic knowledge; facilitate spoken language and cognition; encourage incidental, autonomous learning [20,22,23].

We should plan the session for three different and complementary moments: the pre-session, the AVT session in progress, and the post-session. In the schematics in Figures 1, 2, and 3 provided by Estabrooks and colleagues, we can find and understand the entire algorithm of the sessions and the content of each of these phases [20].

As we can see, in pre-session time, the professional must review the current competences of the child according to the last sessions, as well as the tasks and objectives indicated since the last session, and the information sent by colleagues from other areas, namely the audiologist, teacher, and psychologist. He/She must also review his/her own notes on the goals to be achieved, the goals set for himself/herself and for the family, on the tasks and materials to be used, and also on the selection of strategies that allow him/her to achieve all these objectives. Based on all these aspects, the next session is then planned in order to make sure that any intervention takes place in the child's *zone of proximal development.*

Pre-Session

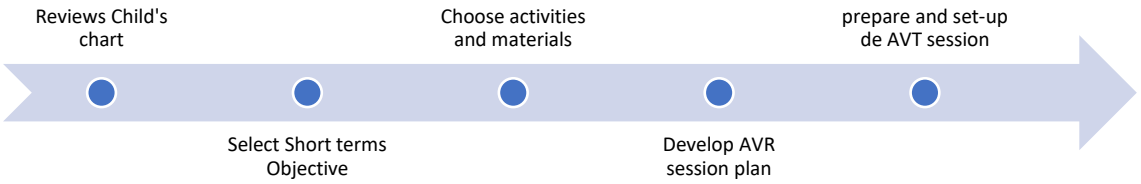

Figure 1

The session begins when the professional greets the parent and child. This welcome should be done in a calm, friendly way, with a well-toned voice and a smiling face, excited for another reunion. In this way, we modeled the form of greeting that parents should ask their child as well. Then there should be a little conversation about what has happened since the last time, especially what has been achieved, how, when, and what needs to be changed.

In all sessions, we must make sure that the child has guaranteed hearing access, that the devices work, and that the child understands what he hears.

It is very important to observe the interaction between the parents and the child and obtain information about the relevant aspects of it. The plan should be presented and quickly discussed with the family. The professional must initiate the proposed activities and apply the strategies thought out in the plan and model for the country. Then you should pass the activity on to the parents so that they train the behaviors and know how to proceed (at this stage, it is very important to practice online coaching in order to support the parents and guide them in solving the problems that they see not forgetting that our main objective is to make them suitable and autonomous in promoting your child's development).

Getting Started in AVT Session

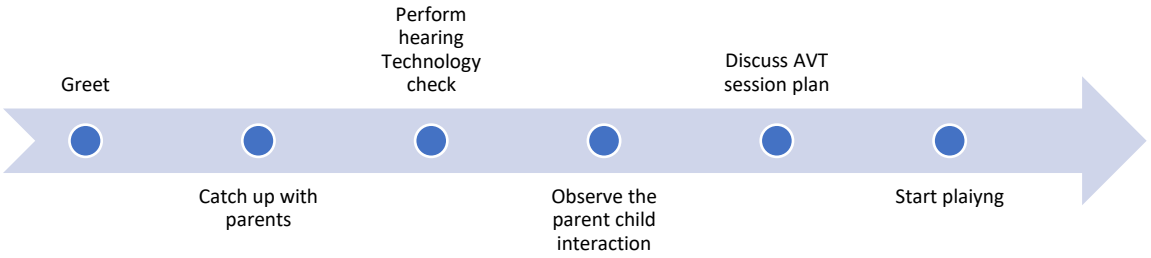

Figure 2

Figure 4 shows a schematic of the cycle of activities throughout the session. As the diagram shows, these activities are cyclic for each activity, group of strategies and techniques over the duration of the session (between 60 and 90 minutes). All sessions must be video recorded with permission of the family since the first day.

Post AVT Session

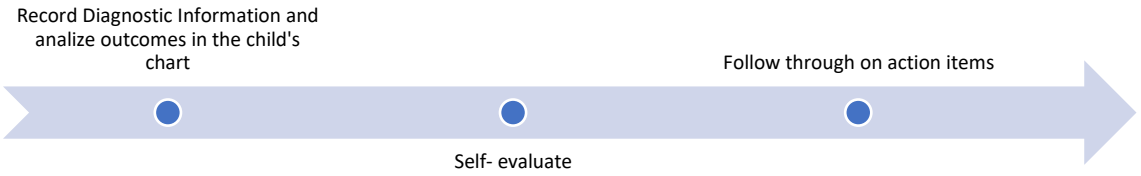

Figure 3

In post-session time, the professional must analyze and obtain diagnostic information in all domains of the child treatment plan (audition, language, speech, cognition, and communication/behavior) and the techniques and strategies that worked or not. It is fundamental to improve (*"raise the bar"*) their own personal performance (what I did that it was a good service delivery and what must be improved and how). The professional must answer about what he did that allowed child to learn to listen, think and talk. He/She must be sure that his/her coaching and guidance to parents are good, and finally, he/she must take notes on the action for the next steps and sessions and prepare information to share with others professionals, if necessary [20,21,24].

AVT Session Cycle (AVTSC)

Figure 4[1]

[1] all information contained in figures 1,2,3, and 4 are by Estabrook and colleagues and are taken from the book Estabrooks W, McIver-Lux K, Morrison HM, editors. Auditory Verbal Therapy- Science, Research and Practice. 1st ed. San Diego: Plural Publishing Inc.; 2020

Chapter 10 - Cognitive skills and social communication

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"Our minds influence the key activity of the brain, which then influences everything; perception, cognition, thoughts and feelings, personal relationships; they're all a projection of you." Deepak Chopra

Introduction

During daily life, people use mental skills to function. People use these skills to learn, to work, to manage their daily lives etc. The set of the mental skills that include working memory, flexible thinking and planning are called executive functions. These functions are important when learning the language. Besides these functions, it is also important to understand other people's state of mind, the so-called "theory of mind". Possessing a functional theory of mind is considered crucial for success in social interaction.

How do children with a CI come to the complex process language development and how do they develop social interaction skills?
What is the impact of binaural hearing on incidental learning?

Executive functions

Executive functions (EF) are a diverse collection of cognitive control and self-regulatory processes that enable humans to manage their behavior and monitor their thoughts during active, goal-directed problem solving. [1]

Thinking Skills	Behavioral Control Skills
Planning	Response inhibition
Organization	Self-regulation of affect
Time management	Task initiation
Working memory	Flexibility
Metacognition	Goal-directed persistence

Figure 1: Executive function skills (1)

Executive skills begin to develop in early infancy and develop throughout adolescence. These processes are mediated in regions in the brain that connect to other parts and functions in the brain, such as speech and language development. These processes interact with each other. For children with hearing loss, it is likely that they have neural, cognitive, and affective delays and disturbances as a result of early auditory deprivation and/ or delayed language development. Therefore, it is useful to look at the basic,

fundamental, and underlying neurocognitive processes that play a role in this process. To be more specific, consider the executive organizational-integrative (EOI) processes involved in speech perception and spoken language processes (figure 2) [1, 3].

EOI Processes	Description
Memory	Verbal and non-verbal working memory; visual memory
Fluency-efficiency-speed	Speed of visual-motor reproduction; word naming; visual recognition; visual matching
Concentration-vigilance-inhibition	Inhibition of automatic/over-learned responses
Organization-integration	Perceptual-organizational skills; visual-motor integration; visual organization planning skills

Figure 2: Executive organizational-integrative processes involved in speech perception and spoken language processing [1]

It is likely that EOI abilities are important for speech-language development after a CI surgery, because of the dynamic relationship between early auditory experiences, the development of spoken language and spoken language processing skills, and the development of EF (figure 3) [1].

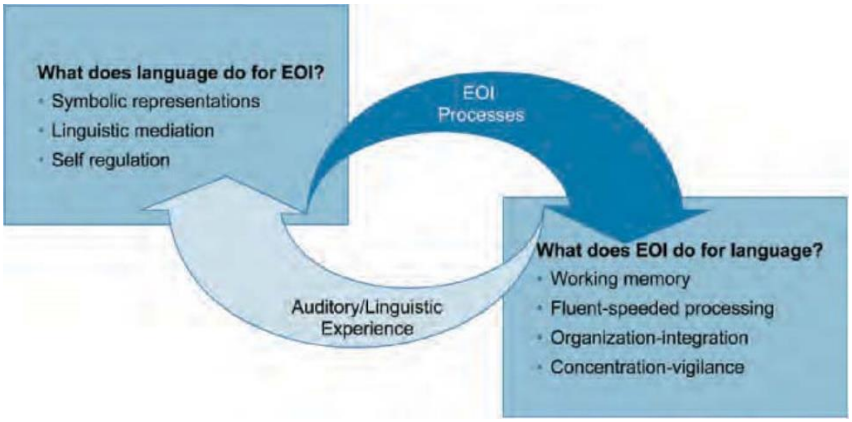

Figure 3: Dynamic relationship between language and EOI processes [1]

Research [1] indicates that four EOI areas are involved in speech-language outcomes after implantation: memory, fluency-efficiency-speed, concentration-vigilance-inhibition, and organization-integration. These abilities allow us to process spoken language quickly (fluency-efficiency-speed) into meaningful parts (organization-integration), stored (memory), and actively assigned meaning (organization-integration and working memory), while the individual maintains a focus on the relevant information (concentration-vigilance) and resists distracting impulses (inhibition).

The development of speech-language skills after implantation relies on these EOI areas to process new auditory input. The individual's ability to coordinate and utilize the EOI abilities will also impact the speech and language outcomes [1,3,4].

Children with CI show significantly lower scores on EF inhibition and working memory. Both of these processes are core neurocognitive processes that are strongly mediated by language. Due to auditory

deprivation, children with CI might have more difficulties with speech perception and spoken language tasks. Especially when tasks have a high cognitive load, such as hearing in noise, perceiving spoken sentences, or formulating sentences based on a picture, deficits in working memory may become problematic [2, 3]. Deprivation in early auditory experiences may influence the development of more basic elementary neurobiological and cognitive functions, extending in learning spoken language. Verbal working memory capacity and fluency-speed are significantly related to language and speech perception skills in children with a CI. The relation between verbal working memory and language seems to be stronger than in normal hearing peers [4,9]. This can be explained by slower subvocal rehearsal, longer memory scanning and irrelevant sound effects. Memory scanning is a gap or pause between words that may reflect the time spent attempting to recall them. Longer memory scanning is likely to affect the efficiency of the memory process [9].

Verbal working memory is a crucial and foundational component of language development following a CI. The development of verbal working memory is strongly dependent on language throughout childhood. CI users who communicate in spoken language depend strongly on phonological and lexical representations during verbal processing and language learning. The capacity demands on the active verbal working memory during routine spoken language processing are likely to be greater than the demands on the memory of normal hearing peers [4,9].

As mentioned before, many children with CI lag behind on their hearing peers in speech, spoken language, and cognitive tasks (verbal and visuospatial short-term memory (STM) and working memory (WM)). The WM is associated with high-level cognitive processing (learning, decision-making, mathematical problem-solving). STM and WM are assumed to be important components that are crucial for speech perception and language skills. WM skills also correlate with language and reading comprehension, vocabulary, language acquisition, following instructions, spelling, writing, and taking notes. All these skills are important for (academic) learning. The WM interacts with the STM, that connects to the long-term memory (LTM) to store and sustain a spoken message. Research shows that long-term CI users with better verbal and visuospatial WM skills have fewer attention problems, which is crucial for learning. The central executive system, WM, STM, and LTM are independent of each other; they are also related [9].

Limitations in the STM may affect the verbal WM of long-term CI users. Long-term CI users probably spend more time and energy during the storage stage, which could mean that the processing might be intact or less affected. Perceiving incoming speech can be more challenging, attentional difficulties and limited speech perception in CI users may therefore lead to fewer items being stored in the STM storage. Severe limitations in STM can be caused by four factors: slower sub-vocal rehearsal, longer memory scanning, poor perception of prosody, and irrelevant sound effects. When CI users speak slower, they engage in vocal rehearsal, and therefore store less items in STM. Let children (when able to read) read the content with melody and visual support; this could lead to remembering more items [9].

Bilateral implantation

It seems that bilateral implantation may ameliorate the effects of auditory deprivation faster and in a more comprehensive manner than unilateral implantation. Stimulating both ears simultaneously appears to create a convergence of the input at the level of the auditory cortex that promotes the normal development of central pathways and creates more chances for incidental learning [6,8].

Limited speech perception and experiencing irrelevant sound effects may affect incidental learning in a negative way and reduce exposure to spoken language, especially when the listening conditions are challenging. These factors may lead to limitations in the semantic LTM, which prolongs the processing time as more time is needed to match the incoming information to the information already stored in the semantic LTM. And this may lead to problems in encoding, storage, sub-vocal rehearsal, and recall. All these factors together (slower sub-vocal rehearsal, longer memory scanning, poor prosodic perception, limited speech perception, spoken language skills) can lead to the decay of verbal materials within verbal STM. Storing, maintaining, and retrieving verbal information could be more demanding, cost more time, and require more effort [9].

Taking all this information into account, the following suggestions should be made for educational programs for CI users:

1. Reduce cognitive load by using both visual and verbal modalities during lessons. Plan activities to improve visuospatial STM and verbal STM.
2. Use short songs, nursery rhymes/poems to improve verbal STM and use more than one modality.
3. Play memory card games to improve visuospatial STM/WM.
4. Train discrimination between important and unimportant information.

Theory of Mind

Children will find out that different people have different viewpoints, beliefs, thoughts, desires, feelings, and so on [10,11,12,13]. The cognitive ability that is responsible for this cognitive function is called the "Theory of Mind" or ToM. ToM is defined as the mental capacity within a cognitive function that affords children the ability to assign and understand the inner mental states of themselves and other people. The ToM abilities are required in infancy. Developmental psychologists consider this cognitive ability to be the most important development in infancy and is the first step in social cognition [11,12,14,15,16]. According to the gathered evidence, social competence is one of the main developmental factors that helps children to succeed in school. The performances on ToM tasks correlated with leadership and (negatively) with loneliness, social competencies, and popularity among peers have even been demonstrated. Therefore, it seems that there is a bidirectional relation between the children's understanding of learning judgements and the ToM development [16,17]. Liu et al. (2018) even stated that the ToM is the first condition for EF development and that, in return, EF is necessary to provide correct answers on ToM tasks.

Theory of Mind Development

Research by Asher et al. (2020) shows that the first sensitive period for ToM exists from birth up to the age of 3.6 years. The brain is the most plastic at most levels, during this period [13]. When infants get near their first birthday, they start to engage in joint attention. Joint attention is the ability of an infant in a triadic interaction to share the focus on an object or event with an adult. Afterwards, infants will develop the ability to pretend play. This happens around the age of 18-24 months [15]. Subsequently, around the age of 2-3 years, an understanding of diverse desires arises. This is when children begin to understand that their own desires are not always consistent with the desires of someone else [14]. The next sensitive period for the development of ToM typically emerges between the ages of 4 and 6 [11,13,15]. The

understanding that people's actions are a result of their beliefs, even by mistake, emerges around the age of 4-5 years. This is called "false belief". The hidden emotion is a later skill that emerges around the age of 5-6 years. This involves the understanding that an individual can mask their own feelings for social reasons even though they display a different emotion towards the outside [12,14]. In the next stage, at the age of 6-7 years, children find out that others can think and give reasons for their mental states. Finally, when children are at the ages of 9-11 years, the meaning of metaphors and jokes will be learned and eventually understood [12].

Language and Theory of Mind

It has been suggested that language skills are fundamental abilities connected to ToM development [13,15]. '*In literature, researchers emphasized the contribution of executive function skills or working memory capacity to false-belief reasoning, and they argued that language facilitates cognitive processing and consequently ToM.'* [13]. It has been suggested that there are multiple reasons why language acquisition is an important ToM performance that goes beyond the general cognitive maturation. Furthermore, it is being described that language ability of deaf children with hearing parents can give a better prognosis for ToM than chronological age [18,19]. Since the quality of everyday conversations has been shown to contribute to an infant or child's later understanding of mental states, the role of language can be speculative in terms of social constructivist consideration [15]. It has even been shown that ToM development benefits from conversations between young children and their older sibling(s) or from frequent use of mental-state language by their mothers [19].

Deaf children can have access to a form of language, through sign language. It is often argued that social competences and other sensory organs develop very well, even when language delays are severe. When they have intact language skills, deaf children can have a well-developed ToM provided by early rehabilitation and cochlear implantation [13]. On the contrary, impairment in ToM has not been described in deaf children whose parents are also deaf because they could freely communicate with each other. This could be an explanation why the limited access to language could impact ToM development in deaf children of hearing parents [10,15].

Theory of Mind development in CI Children

Any form of language from birth should enable infants and children to develop a regular ToM, regardless of their hearing ability [16]. However, the social and pragmatic communication skills of CI children may not rise to the same level as normal hearing peers, especially when there is a language deprivation in the early years of live during the development of language and speech perception. CI children could also experience difficulties engaging in spontaneous interaction and conversation with peers, which could interfere within the development of ToM and other cognitive abilities [10]. On the contrary, throughout the years, a lot of researchers have reported that even though a CI does not provide typical hearing, there are always effects of improved communication, weaker social competence, and spoken language development. When children have had the benefit of their CI for a longer period of time, several young bilaterally implanted children catch up in ToM development with respect to their typically hearing peers. In research, it appeared that with infants who received their first CI before their first birthday and second birthday, ToM skills were equally developed [12,16].

Finally, ample evidence has been found that there is, in fact, an association between EF and ToM in young CI children. In particular, inhibitory control appeared to play a massive part and appeared to be

significantly correlated with ToM. Inhibitory control emerged as the main predictor of ToM. This means that EF is a prerequisite for ToM. In conclusion, interventions that may be effective for supporting ToM development are cognitive interventions. The following statement has been made: *'This suggests that parents and teachers could increase their access to opportunities of speech and language practice, which may help children to improve their cognitive abilities by focusing on development of VA, which in turn may strengthen the ability of conversation with their parents and teachers.'* [14].

Summary

EF are a diverse collection of cognitive control and self-regulatory processes that enable humans to manage their behavior and monitor their thoughts during active, goal-directed problem solving. These begin to develop in early infancy and throughout adolescence. The processes are mediated in different brain areas that interact with each other. Children with hearing loss are more likely to have cognitive and affective delays and disturbances as a result of delayed language development and/or early auditory deprivation. The EF that are most important for speech- and language development after CI surgery are the EOI abilities. After all, there is a dynamic relationship between early auditory experiences, the development of spoken language and spoken language processing skills, and the development of EF. The development of speech-language skills after a CI implant relies on domain-general EOI areas to process new auditory sensory input. The individual's ability to coordinate and utilize the EOI abilities will also impact the speech and language outcomes. EF difficulties are associated with social maturity after long use of CI. This could be more at risk due to limited social-emotional experiences.

Within 3,5 years of bilateral deafness, CI implantation can provide the development of age-appropriate cortical responses over the first 3 to 6 months after implantation. Bilateral implantation can restore binaural hearing, which allows us to: (1) localize/identify sound sources in the space; (2) increase the perception of loudness through binaural summation; and (3) improve the hearing in quiet and noisy environment through the head shadow and squelch effects. This makes listening less tiring, and consequently, communication more pleasant.

At some point, children will find out that people have different viewpoints, beliefs, thoughts, desires, feelings, and so on. The 'Theory of Mind' (ToM) ability is responsible. This cognitive ability is considered the most important development in infancy and is the first step in social cognition. Research reveals that ToM is the first condition for EF development. In return, EF is necessary to provide correct answers on ToM tasks. It has been suggested that there are multiple reasons why language acquisition is an important ToM performance that goes beyond the general cognitive maturation. Deaf children can have access to a form of language, through sign language. It is often argued that social competences and other sensory organs develop very well, even when language delays are severe. If deaf children have intact language skills, they can have a well-developed ToM provided by early rehabilitation and cochlear implantation. Conclusion: when children have had the benefit of their CI for a longer period of time, they catch up in ToM development with respect to their typically hearing peers.

Chapter 11 – Learning Language and communication by Playing

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"Let the child be the scriptwriter, the director and the actor in his own play." Magda Gerber

Which are some of the practical activities that parents can engage in to enhance their children's social skills?
Which is the most suitable (physical) place in the classroom for students with cochlear implants?

What is play?

Play is, for children, about having fun in a spontaneous and engaging way. Even though it appears to be something simple and unintentional, play can be explained as "the expression of intentional states – the representations in consciousness constructed from what children know about and are learning from ongoing events – and consists of spontaneous, naturally occurring activities with objects that engage attention and interest" [1]. Play may or may not involve caregivers or peers, may or may not involve a display of affect, and may or may not involve pretense and the use of symbolic representations for objects, people, and/or activities [1].

Jumping around in a trampoline is play …

Building a tower piling up building blocks is play …

Sharing a table game with a friend is play …

Creating an imaginary scenario with miniature animals is play …

Making pretend play with a plot of doctors and nurses is play …

Play can be as much as the child wishes … especially when adults can support and enrich these activities, from a very young age. Through play, children learn about themselves and the world in the best way they can. They, in fact, actively construct "new knowledge about objects, people, and events by integrating new experiences with what they already know" ([1], p.228).

However, there are many misconceptions and erroneous ideas about play that influence the way that we, as adults, think about this fundamental activity in childhood.

Common myths about play!

There are some common myths about play that influence the way adults think about this very important activity, interfering with the way "grown-ups" enhance play and support the development of play skills (Parenting Counts, N/D). Some of the wrong ideas some of us might believe in are that:

- … play wastes time and prevents learning

In fact, play is a fundamental tool for helping children understand their environment and relationships, so, in a way, it is actually harder for children to learn if the approach is not playful.

- … play is really nothing more than fun and games

Yes, it is fun, and it is about learning … However, it is also the ideal environment for helping children understand how to live happy, healthy lives. There is no chance for children' well-being if there is no time to play in their lives!

- … play is not important until children are older

Play actually helps infants build strong bonds with caregivers, and even simple games like peek-a-boo stimulate a baby's brain and help it develop. Therefore, playing is really important from a very, very early age.

- … children only benefit from structured play directed by adults

There is a role for structured play, but unstructured, independent play is important too; it helps children discover their own interests and develop their imagination.

Given the importance of play to children, it is fundamental that we stop thinking of it as something that is necessarily purposeful; otherwise, it is a waste of time. More than an interest, play is a need for young people, considering its importance in terms of development and well-being.

How is play important for development and well-being?

According to Goldstein [2], play is essential for the child's development since it contributes to the cognitive, physical, social, and emotional well-being of children and youth. It also provides an ideal opportunity for parents to engage fully with their children while doing something everyone enjoys, which is important for the promotion of healthy attachment and relationships.

Play has potential advantages in many domains of the child's development, namely:

Emotional-behavioral benefits:

- reduction of fear, anxiety, stress, and irritability;
- creation of joy, intimacy, self-esteem, and mastery not based on others' loss of esteem;
- improvements in emotional flexibility and openness;
- increase in calmness, resilience, adaptability, and ability to deal with surprise and change;

Social benefits:

- increase in empathy, compassion, and sharing;
- creation of options and choices;
- modeling of relationships based on inclusion rather than exclusion;

- enhancement of verbal and nonverbal skills;
- increase in attention and attachment.

Physical benefits:

- increase the efficiency of the immune, endocrine, and cardiovascular systems, related to experiencing positive emotions;
- decrease in stress, fatigue, injury, and depression;
- increase in the range of motion, agility, coordination, balance, flexibility, and fine and gross motor exploration.

Besides all that has already been mentioned, play has also been closely connected to communication and language development. Research from diverse countries shows a relationship between early social play and communication skills at a later stage. Lyytinen et al. [3] reported that symbolic play at the age of 14 months is a good predictor of the children's language development at the age of two. In another study, children aged one and a half to two and a half, who were provided with plastic building bricks with which to play, had significantly better language scores six months later [4]. Westman [5] linked the children's confidence and motivation when playing to their language development. Children who are motivated by play and try to expand their play actions tend to be more confident and to show higher linguistic development.

There are so many potential benefits of play in terms of development, health, and well-being that there are actually "no excuses" not to take play very seriously. Let's now explore how play develops over time to understand how to support children better.

Ages & Stages: Different types of play developing over time

Play develops over time and begins at a young age when adults and children interact and play together.

The first type of play manifested between children and their caregivers is called People Play, consisting of the dyads playing with other people in simple routines (e.g., peek-a-boo) in social back-and-forth exchanges between two or more people [6]. People Play has evolved over time. Early People Play usually involves some physical sensations that children appreciate, implies shared attention, turn taking, and the exchange of nonverbal communication cues, and may introduce objects (e.g., bubbles). At a later stage, this kind of play may evolve into a more complex game (e.g., chase and tag), imply more use of language and pretending, and involve other children as play partners [7.]

The second kind of play exhibited by the child is Toy Play, meaning that children are now able to explore toys and objects, with physical activities such as banging, throwing, shaking, trying to understand how objects feel and work [6]. Toy Play may be Exploratory, when children perform actions on the toys and objects using all their senses, first, performing the same actions (e.g., banging) with different objects and later beginning to differentiate. In time, Toy Play evolves and becomes Constructive, with the child being able to perform actions on the toys and objects as expected (e.g., build a tower with the blocks); at first the child uses one expected action on one object, and later he/she starts to use multiple expected actions in diverse objects, which contributes to the child's conceptual development. Gradually, more language will be used in relation to the Toy Play taking place [7].

Lastly, the more evolved form of play that emerges as the child develops is Pretend Play, which involves symbolic and "make believe" play and consists of playing using symbolic representations (e.g., a spoon

can be an airplane), therefore being very much related to the development and use of language []8. In Early Pretend Play is very similar to Constructive Toy Play in that the child uses objects and realistic toys in expected ways, creating sequences of pretend ideas; it is similar to "Reality play", using imaginary representations of objects and scenarios on the child and others. Later Pretend Play involves thinking at a higher level, meaning that the child does not need real or realistic objects to pretend (e.g., a cardboard box may be a car). The newly developed imagination enables the child to pretend and create short sequences of ideas and stories with objects. The child is using symbols to play and create ideas, and this ability to hold ideas in mind allows the child to pretend play even without objects (e.g., the child may lick an imaginary ice cream) [8].

From Early People Play, to Later Pretend Play, many skills are developed between children and caregivers, while playing. There is, nevertheless, the possibility that play skills do not develop as expected over time, with damaging effects on the child's development and well-being.

Are there any potential threats to the development of play skills?

Can a Hearing Impairment have an impact on play and play skills?

There are many internal and/or external factors related to the child that may interfere with play and hinder the development of play skills. Drake [9] stated some of the variables that may influence play development, namely:

1. Motor skills problems that limit the child's abilities and opportunities to participate in playful exchanges
2. Cognitive development delays with impact on the development of play skills
3. Sensory processing difficulties that create obstacles for the child's engagement in play activities
4. Social interaction issues that can challenge the child, as well as the caregivers and peers, while trying to share different types of play
5. Miscommunication and difficulties with language expression and understanding

Many researchers have dedicated time and efforts to explore the topic of play specifically by children with hearing problems. As it is with language, play is very much influenced by the hearing status and the modality of communication – sign or spoken language – used by the infant. Less optimal early experiences, such as "deprivation of linguistic stimuli by the caregiver, lack of exposure to sign language, linguistics re-education delay, could impede normal cognitive and linguistic development of hearing-impaired children" ([10], p.99). On the other hand, "better language development in both sign language and spoken language is a predictor of better play behavior", according to the same author. This does not mean that children with a hearing impairment will think, learn, or play differently than their typically hearing peers; they may need more visual cues, gestures, speech reading, or just more time for turn taking, but the play exchanges may be of equal quality, especially when created with suitable communication and language modalities from very early ages.

As a conclusion, it is especially important to remember that for hearing impaired children, as well as for children with other types of disabilities, play development will be dependent on the children's current play skills — "what the child already knows" about playing — and the actual competence of the caregivers to join in and support play — "what the caregiver knows" about playing with that child. If the right balance is created, there will be fertile ground to have fun and evolve through play.

How can we support communication and language development using play?

In order to join in and support the development of play skills in children, the following strategies may be useful. Read through the next table and try to find your own practical examples.

Strategy	Description	How would I use this strategy?
Early People Play		
Face to Face	Position yourself face-to-face and at the child's eye level	
OWL	Wait, while observing the child's behaviors and listening to any communicative attempt	
Interpret	Interpret the child's behavior, giving it an "as if" meaning, even if you are not sure it was intentional	
Imitate	Imitate the child's play	
Insist	Insist carefully, creating longer chains of play between the dyad.	
Later People Play		
Sensory Preferences	Respect and take advantage of the child's sensory preferences while creating playful activities	
Early Toy Play: Exploratory		
Follow the Child's Lead	Include the child's interests in the play activities. Try to joyfully intrude, joining in and playing with the same object Interpret the child's behavior, giving it meaning or an "as if" intention Imitate the child's play	
Comment	Give your child a word or short phrase that relates to what your child is doing or saying at that very moment…then wait!	
Later Toy Play: Constructive		
Level UP	Level 1: Imitate existing single-expected play actions on different toys Level 2: Imitate new single-expected play actions on different toys Level 3: Imitate combinations of 2 or more expected play actions on different toys	
Early Pretend Play		
Early Pretend Play 1	Add pretend actions to create sequences of pretend actions	
Early Pretend Play 2	Pretend by substituting one object by another. Pretend with invisible objects	
Later Pretend Play		
Expanding Later Pretend Play	Create new and original ideas Create role plays and act out situations Include other children in pretend play.	

Table 1. Simple strategies to support play and play development
(Adapted from Sussman & Weitzman, 2014a; Sussman & Weitzman, 2014b; Weitzman & Drake, 2014)

Chapter 12 – Learning Language with Music, Singing and Sharing Books

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"Coming together is a beginning, staying together is progress, and working together is success." Henry Ford

Mention two different challenges (and the related solutions) teachers might face when working with students with cochlear implants.

Using books to promote emergent literacy

What is emergent literacy?

It is now widely recognized that starting early with structured, varied reading and writing experiences, supported by more competent readers is critical to fostering emergent literacy skills and educating young readers [1], those who, from an early age, enjoy the pleasure of literature.

There are several models in the light of which the concept of Emergent Literacy is described and explained, as well as the programs and intervention proposals that, internationally, gather scientific evidence regarding their effectiveness in stimulating "pre-reading" competence. Whitehurst and Lonigan [2] propose the renowned Inside-Out/Outside-In model, which considers the following basic skills as the fundamentals to make learning to read and write easier:

Inside-Out Skills	**Outside-In skills**
Allow the translation of graphic symbols into sounds, facilitating decoding of the written code, namely: - Knowledge of the alphabet; - Phonological awareness.	Allow understanding of the meaning of writing, facilitating understanding and interpretation of written content, namely: - Mastery of vocabulary; - Linguistic competence.

Table 1 - Basic Skills

Picking up on this important concepts, [3][4] organized two intervention programs that focus on developing emergent literacy skills in kindergarten children. According to the authors, the skills to be worked on include:

- Pragmatic skills to participate in conversations;
- Knowledge of vocabulary;
- Understanding of narratives;
- Abilities for linguistic inference;
- Phonological awareness;
- Mastery of graphic notions and print concepts.

Having a clear concept of emergent literacy, one can wonder about different activities and resources that may be used to foster these skills. One can also understand why some traditional ideas may be considered myths or false beliefs and induce caregivers into fallacious thinking around this issue.

Common myths about literacy!

There are many beliefs and dogmas associated with learning to read and write, despite contradictory research evidence. Some of the most popular and potentially pernicious myths that influence reading education are presented below [5].

Myth #1: Kindergarten emergent literacy promotion is teaching reading and pushing down the first-grade curriculum.

Fact: Kindergarten literacy instruction responsively promotes young children's linguistic skills (and other skills) that support their literacy development.

Myth #2: Kindergarten literacy instruction is developmentally inappropriate.

Fact: High-quality literacy experiences in the kindergarten context are developmentally appropriate and supported by decades of solid research.

Myth #3: Kindergarten literacy experiences make for less time for play, fun, or social-emotional development.

Fact: Kindergarten literacy experiences should be fun, playful, and engaging!

Myth #4: One size fits all when it comes to literacy experiences and instruction.

Fact: Kindergarten literacy instruction and experiences are individualized, and each child —and each group—deserves a personalized approach.

Myth #5: Learning to read is a natural process.

Fact: Simply put, it is just about the most unnatural thing humans do. This belief is related to the idea that learning to read is a natural process that comes from rich text experiences. However, learning to read and write requires systematic and more or less formal instruction and a lot of effort from the learner.

Myth #6: Children will eventually learn to read if given enough time.

Fact: It is true that children should be taught to read in developmentally appropriate ways, and that instruction should always address each child's zone of proximal development; however, one should not

simply wait for children to develop reading skills in their own time. A child who does not develop reading skills along with his or her peers is a reason for great concern, and measures should be taken.

What about literacy and hearing impairment?

Is there any real relation between hearing disability and learning to read and write?

Can a Hearing Impairment have an impact on emergent literacy skills?

There is vast research around the topic of literacy, reading and writing proficiency, and hearing impairment. Long-term literacy outcomes for children with hearing impairment, in particular those with severe-to-profound hearing loss who use cochlear implants, are below those of children with normal hearing. The causes for these long-term deficits are not completely understood, though differences in the auditory access may be "only" a partial cause [6].

In recent research, Ingvalson et al. [6] concluded that children who use cochlear implants show performance deficits on nearly every component of the Whitehurst and Lonigan's [2] emergent literacy model, namely phonological skills and vocabulary. Though this can be caused partially by the differences in children's auditory experiences, the fact that many high schoolers who were implanted with cochlear implants at an early age do not read at grade level suggests that improving auditory skills by itself may not be enough. It also reinforces the idea that interventions that target emergent literacy skills may be necessary for children with hearing impairment, in order to give kindergarten children who use cochlear implants good foundations for literacy instruction.

How are books important to promote language and literacy?

When it comes to promoting language and emergent literacy skills, books are, without a doubt, an invaluable resource. May it be by the hands of parents, by its use by professional caregivers, or even "just" by having the child "giving it a go" and exploring them independently, choosing the right books and creating engaging reading activities is one of the best ways to introduce young children to the universe of literacy and literature.

Recently, through an intentional collaboration between teachers and Speech and Language Therapists (SLTs), Murphy, Pentimenti, and Chow [7] proposed to support the language and literacy development of young children with or at risk for learning disabilities using shared reading activities. The authors concluded that this interprofessional collaboration created a high-potential setting for students with and without disabilities to learn, allowing teachers and SLPs to target key early literacy and language skills using evidence-based strategies. By collaborating, the professionals were also able to support each other's practices and enhance the quality and effectiveness of literacy instruction and intervention.

In a similar approach, professionals may cooperate with parents and other caregivers in order to promote the best literacy experiences in the home setting. Ferreira and Silva [8], authors of *Conto Contigo*, a familiar literacy program, highlight the importance of good literacy routines at home and how it is possible to improve these routines, guiding the caregivers on how to implement literacy activities at home and as a family "ritual".

The next sections present a very simple approach to promoting emergent literacy skills, just by choosing the right books and creating high-quality shared reading experiences.

Different books for diverse readers Choosing the right book for different audiences is key to ensuring the success of the early literacy experiences. Different children will have diverse preferences, competences, and needs; therefore, it is important to explore the array of literature available and then to choose wisely.

Younger children, even at a pre-verbal level in their linguistic development, will be able to participate in early literacy experiences as long as the adult chooses books that look like toys (i.e., are made of plastic, fabric, or wood), have colorful showy pictures, have photographs of familiar people and real objects, have rhymes, rhythm, and repetition, and are explored in an interactive way. A bit later on, and after the first book sharing experiences, it is possible that children will be available and prepared to explore books that already have a few words [9].

Figure 1. Examples of book choices for the younger pre-verbal children

As the child grows older, he/she gains "literacy experience" and evolves in the understanding and use of language, the book choice should evolve and incorporate books that have pictures and photographs of children doing familiar actions, include funny rhymes, rhythm, and repetition, explore a nuclear topic (e.g., the family, the toys around the house etc.), and tell a simple story, with a very basic narrative structure (i.e., a few characters, a few scenarios, clear timeline, one problem, a few actions from the characters, and a simple resolution). At this level, it is also interesting to provide the children with books that have central themes (e.g., going to the doctor), exploring it with repetitive and predictable words and sentences [9].

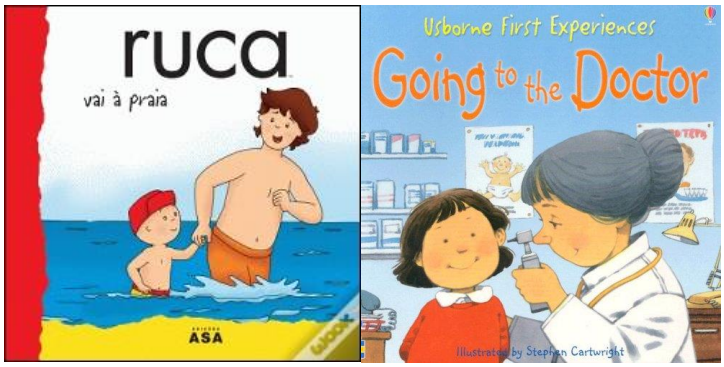

Figure 2. Examples of book choices for the younger verbal children

With time and diverse, high-quality shared reading experiences, it is likely that the child will become more and more familiar with how literacy operates. At some point, it is possible that the child becomes a more active "reading" partner, trying to guess the letters/words, formulating hypotheses about the story, naming, and describing pictures etc. At this stage, as an early reader, it is interesting to offer books with "*onomatopoeias*" (e.g., sound-like words), possible translation with movements, many action verbs, diverse contextualized nouns, different adjectives, short and simple sentences, and written content that can be complemented by deictic and paraverbal elements. Books that challenge the child with consultation hypotheses (e.g., "What is there in the country?"; "What is there in the city?") are also good ideas, promoting further vocabulary exploration.

Figure 3. Examples of book choices for the younger verbal children

Choosing great books is a very good start! Then … one just needs to know how to use it effectively!

How to support emergent literacy skills using books?

One great activity to promote the development of emergent literacy using books is the practice of Shared Reading. Shared reading allows adults to model vocabulary and language structure and to relate both to the children's lives and experiences. It is also a good activity to teach young children how to interact around books and to learn the structure and rules of conversations. Besides contributing to the improvement of the conversational and linguistic skills, shared reading supports the understanding of diverse print concepts, helping young people to understand how books and literacy "work" [10].

Considering Cole, Madox, Lim, and Notari-Syverson´s [11] C.A.R. strategy to follow the child's lead, and incorporating additional elements, Corine Watson (cited by [10]) suggested the "***Put the C.R.O.W.D. in the C.A.R.***" approach to improve children's linguistic skills by taking part in high quality Shared Reading experiences. The next picture [2] presents this approach in a clear and simple way.

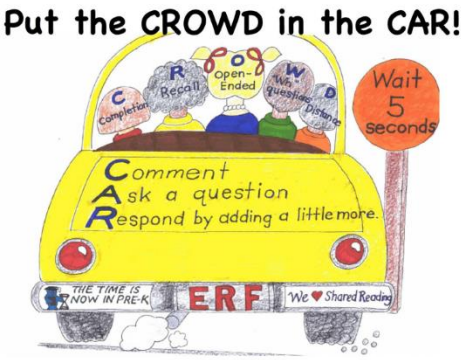

Figure 4. Following the child's lead to enhance participation in literacy experiences

As defined by Cole et al. [11], a simple way to follow the child's lead during a book reading activity is to:

1) **C**omment on something the child did or said and then wait for five seconds (e.g., "*Wow, that's a really big dog*!").

2) **A**sk a question directly to the child and then wait for five seconds (e.g., "*Do you like big dog*?").

3) **R**espond to the child by adding additional information (e.g., "*I like the big dog. He looks friendly.*").

Related to the **C.A.R.** strategy, the **C.R.O.W.D.** acronym stands for:

i) **C**ompletion – the child is asked to complete a repetition line (e.g., "*… and then the wolf blew, blew, and …*").

ii) **R**ecall – talking about what has already happened in the story (e.g., "*I wonder if you remember where the first little piggy ran to?"*).

iii) **O**pen-ended Wh- Questions – the adult reader asks open-ended questions about the story (e.g., "*How do you think the piggies were feeling*?").

iv) **D**istancing – relating the content of the book to the child's life (e.g., "*Do you remember when we went to the farm and saw the piglets*?").

Using the very simple "**Put the C.R.O.W.D. in the C.A.R.**" strategies and supplementing them with the use of gestures, symbols, cued speech etc. will most likely support the development of language and emergent literacy skills with children with a hearing impairment!

Using music to promote communication

What is music?

Music listening is one of the most pleasurable activities in our lives. Music is a field that has its roots in nature. In order to achieve what we now call music, composers laid the foundations using the sounds of nature. One of the most widely accepted definitions of music is that it is the art through which feelings and ideas can be expressed through a chain of musical notes to result in a pleasing aesthetic form. Among its many roles, such as entertainment or educational, music also has a therapeutic role.

Music, an abstract stimulus, can cause feelings of euphoria similar to the tangible rewards involved in the striatal dopaminergic system. It has been empirically demonstrated that music can effectively generate pleasurable emotional responses, and neuroimaging studies have shown the involvement of emotional and reward circuits in the brain while listening to music [12].

Quality parent-child interactions with the help of books, rhymes, songs, stories, daily routines have an essential role in the child's development in all areas: cognitive, emotional, socio-affective, relational, and linguistic. In today's world of distractions and technology, therapy time should be a micro-universe in

which parents and children build relationships based on secure attachment and quality time, focusing on what really matters.

Common myths about music!

Despite all the positive thoughts and ideas shared about music, when it comes to connecting it to the child's development there may be some erroneous ideas in the minds of adults. Here are some of these false beliefs:

- *… Music distracts the children, and they can no longer focus on what they are doing.*

In fact, music is an important tool that helps children to focus. The right rhythm and words can help your child focus on learning. Music holds great therapeutic potential and can improve communication skills.

- *… Children with cochlear implantation cannot play music.*

Children with cochlear implantation have a different experience of music.

Researchers from New Macquarie University have revealed how music helps children with cochlear implantation. The children showed significant improvements in various aspects of their sound perception, such as their ability to identify whether a sentence was a question or statement based on the rhythm, linguistic stress patterns, and tone. Their instrumental identification success increased by 8 percent, indicating improvement in their timbre perception. Even a modest amount of music training has benefits for music and speech outcomes. Music training is a suitable complementary means of habilitation to improve the outcomes for children with hearing loss [13].

How is music important for communicative and linguistic development?

Pleasant music can bring improvements in long term memory [14]. Musical activities can positively affect a child's whole development. Participation in musical activities enhances learning by promoting language skills, cognitive skills, communication skills, social and emotional skills, gross and fine motor skills, and creativity. The child will bond with the teacher and parent while sharing music together.

Recent findings indicate that brain regions involved in the processing of musical and language at the cortical and subcortical levels tend to overlap. We can appreciate that there is a significant degree of affinity between language and music. The researchers' findings indicate that children with implants or hearing aids display poorer music perception skills compared to their peers, but this seems to be related to differences in acoustic hearing prior to cochlear implantation. When offered musical training, their perception of musical acoustic features improves, and this transfer effect of musical training seems to affect not only language but also other cognitive functions [15].

Music training may be particularly helpful for hearing as it requires students to be receptive to quick and detailed changes in sound.

Musical stimulation is based on two musical values: rhythm and melody. Musical stimulation makes possible and improves: hearing, speech perception, speech production, natural speech development, rhythm, intonation, intensity and articulation, and auditory memory. Music and musical structures and games create order in the motor, musical, and verbal expression of the child.

Music has a strong emotional power. The most common goal of musical experience is related to music's ability to modulate emotional state in the listeners. There are many studies that have shown that, beside distraction, mood regulation, and recreation, music-evoked emotions can also drive memory enhancements in both healthy and clinical populations [14].

Many recipients say that listening to and participating in musical activities is something that would make them feel more "normal" and a part of society [16,17]. Adults, who once could hear and now are implanted use their memory of music as a contextual cue. Children who were prelingually deafened cannot use these contextual cues to process the sound, but they have the advantage of a more plastic nervous system.

Participation in music activities helps children to develop communication skills, language skills, cognitive skills, and motor skills, and also helps them in the social, emotional, and creativity areas.

Can a Hearing Impairment have an impact on music skills?

Children enjoy rhythmic sounds and frequently bang various objects together to make noise. Parents can play by clapping hands, stamping feet, rubbing hands together, or opening and closing their mouths with a popping sound.

Researchers found that hearing-impaired children with cochlear implants who sing regularly have better perception of speech in noise compared to children who do not sing. This is an important skill in daycare or school settings where children discuss and receive instructions in noisy conditions. They found that communication skills and the ability to perceive speech in noise have a vital importance in education, because all children should have the possibility to learn music and singing [18].

There are a lot of studies that examined the effect of music therapy on children with cochlear implantation. The findings revealed that music therapy has a positive effect on communication as long as parents and educators learn how to expose children with cochlear implantation to music. The results of a study by Kerem [19] suggested that music therapy promoted communication by increasing the frequency and duration of spontaneous imitation, initiation, turn-taking, and synchronization of children with CI. It is necessary to include music therapy in daily experiences to obtain the best results.

Robins and Robins [20] suggested that all the instruments that can be used successfully with children with normal hearing can also be used with children with CI. They also claim that it is of great importance to use instruments that produce clear, distinctive, interesting, and enjoyable sounds. Teachers could present children with more instruments, and they could choose the one that they like best. We can expose children to instruments that produce a variety of sounds. It is important that the instruments suit the children's developmental stages.

How to support communication using music?

Parents can have activities with their young children, like singing lullabies before sleep. This will create a secure space and will help the child to become familiar with the mother's voice.

Parents can help their child take notice of sounds. They should choose a quiet environment, show children different sounds, and encourage listening and responding. They can ask the children to repeat sounds after they hear them and help them to pronounce them correctly. They can start by imitating animals, like dogs or horses, and while the child hears the sound, the parent can show a picture or a toy with the animal. Some other strategies to support the child's interests and abilities with music are presented below.

Using visual stimuli while singing

Meaningless or basic rhymes (spoken or sung) use syllables in the rhythm of kindergarten rhymes. These rhymes are said or "sung" accompanied by the beat of the rhythm, which it can be made by using musical toys like maracas, chopsticks, drums, and clapping.

BA BA

BA BU

BU BU

Kindergarten rhymes that make the transition from music to speech. In these games, the introduction of meaningful words with syllables begins.

BA-BA BA-BA

BU-BA *BU-BU*

Kindergarten rhymes with meaning and *short songs using meaningful words*. It is used after the children have mastered the correct pronunciation and rhythm of each existing word in the rhyme structure. It is recommended that the text be accompanied by a graphic representation of the correct rhythm and images representing the words in the text.

ROW, ROW, ROW YOUR BOAT, ROW, ROW, ROW YOUR BOAT

Interacting with the child using child directed speech. Implanted child directed speech should be clearer and more repetitive, use a higher pitch, use a slower tempo, more melodious with more varied intonation, a sing-song rhythm. Songs like „Twinkle, twinkle, little star", „Wheels on the bus", „Five little ducks", „Row, row, row your boat" can be used.

Listening, identification, and playing percussion instruments. It is important for children to have access to a few percussion instruments like drums, triangles, wood blocks, electronic keyboards, xylophones, bells, and whistles. Also, parents can improvise instruments using a bucket or a pot turned upside down, and wooden spoons. Children will be encouraged to try to say the sounds that they hear. First, they will start with one instrument, and after the child has learned the specific sound, another instrument will be introduced. After that, the children will have to distinguish the differences between instruments.

Chapter 13 - Transition to School

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"The highest result of education is tolerance."
Hellen Keller

Introduction

Transitioning from the early childhood phase to primary school is one of the significant steps that each child must take during their educational journey. Similarly, one of the most critical steps in children's (re)habilitation following cochlear implantation is related to their transition to school. Therefore, this specific transition can also be seen as a part of the (re)habilitation and an objective. Therefore, preparing both families and children for this experience is essential for a successful school start. Furthermore, since teachers are also a significant factor in this process, we will also focus on their involvement.

Given that the parental needs for the transition of children using cochlear implants from preschool to elementary school have been explored in a growing number of studies, and most of them highlighted parents' need for information on most of the aspects related to school transition [1], this chapter will answer the following questions:

- Which are the most significant factors parents should consider before their CI children's school transition?
- When transitioning to school, which are some of the most efficient practical techniques the parents might use to encourage their CI children's language development and independence when transitioning to school?
- Which are some of the most efficient practical techniques the classroom teachers might use to facilitate CI children's transitioning to school?

Mention two different challenges (and the related solutions) teachers might face when working with students with cochlear implants.

Building self-confidence and self-esteem

Self-confidence and self-esteem are essential for children transitioning to school, with or without a cochlear implant. These personality traits shape children's perceived ability to handle situations successfully (e.g., without leaning on others) and their positive self-evaluation. Previous research

highlighted various strategies and practices that parents can apply to enhance these traits in their siblings, regardless of the children's status (with or without a cochlear implant). For example, when parents praise their children's efforts, they encourage them to adopt incremental motivational frameworks [2]. In other words, children learn that their abilities are malleable and attribute their success to their hard work, learning to enjoy challenges, and generating new improvement strategies. Additionally, as research shows, parents should account for other significant factors regarding their children's self-confidence and self-esteem related to a warm, secure, and positive family environment [3-4].

However, research also suggests that there should be a reasonable, balanced approach considering the ways parents might raise their children's self-esteem and confidence. More specifically, praise might also backfire in children with low self-esteem [5]. It seems that adults are often prone to giving a person praise (e.g., "You are so intelligent!") as well as inflated praise (e.g., "That is amazingly beautiful!"). However, paradoxically, such praise might have a counter-effect, i.e., it might decrease the self-esteem of children with low self-esteem by lowering their motivation and feelings of self-worth when facing setbacks (e.g., when they struggle, when they have to handle a failure). Therefore, parents should approach a more equilibrated way to enhance CI children's self-confidence (e.g., realistic feedback that is relatively close to objective benchmarks) [6]. This way, children will also be able to handle potentially less successful academic or social contexts.

In addition to the global idea of self-confidence, self-confidence is highly significant when transitioning to school for CI children. In this case, self-confidence can also be translated into sufficient spoken language skills to help them be effectively independent and competent during their daily school interactions. Therefore, parents should identify the potential gaps in their CI children's language before school starts. In order to do that, a visit to the school and good (previously acquired) knowledge of the school curriculum would reduce the potential related limitations. Another efficient approach is practicing language and vocabulary together with the child in different school-related contexts, which might make this transition smoother.

Enhancing autonomy

Children with hearing impairments need to be as self-sufficient as possible in controlling their hearing aids at school. For example, children should be able to turn the gadget on and off, change the batteries, and notify the teacher if there is a problem (depending on their age, of course). Furthermore, encouraging children to communicate their needs vocally is critical because it allows children to achieve the independence they need to succeed in school.

At the same time, children should be encouraged to be their own best advocates at school and answer inquiries regarding the device and why they use it with confidence. For instance, parents can teach them the correct terminology when discussing their cochlear implant components (if there is an issue, children should be able to indicate which part is malfunctioning). In addition, parents can teach their children how to self-advocate by modeling independence at home. For example, children may engage in role-playing scenarios, and as a part of their school preparation, they can practice interacting with their peers and teachers.

Enhancing children's conversational skills

Encouraging language development and conversational skills implies that parents should be trained as co-therapists to ensure consistent daily practice. To achieve this goal, adequate parental training is needed, adapted, and personalized to the social and educational characteristics of the family [7]. Without this adaptation, there is a risk of rejection or anxious reactions hindering full family involvement. However, parental involvement can facilitate good social and school integration with the correct information and trained skills, decreasing the risk of social, academic, and professional disengagement [8-9].

Conversational skills (also called conversational competence) combine the abilities to talk and listen actively. When we want to develop this competence in children with cochlear implants, we consider the receptive language, expressive language, and practical side of communication. Several intrinsic factors influence the results of language stimulation activities. These factors are related to the child's age, hearing loss, other associated medical problems, and level of nonverbal cognitive performance. In addition, the dynamic of intrinsic factors is shaped (amplified or diminished) by extrinsic factors. Among the most important related factors is the consistency of wearing the implant, the available educational programs, the services, and support that the child benefits from, and their behavior in the family dynamics [10].

However, the development of conversational skills is a systematic, continuous process. Therefore, parents should take every opportunity to encourage and practice several aspects of language development, i.e., articulation (orthoepic pronunciation) - intelligibility of speech; speech rhythm - speech fluency; the qualities of the child's voice – resonance, intensity, pitch, timbre, melody; the child's posture; nonverbal elements of language - facial expressions, gestures; indicators of language development - mean length of utterance, the total number of words, number of different words.

Pronunciation development in children with cochlear implants follows the same guidelines as in the typical child. However, there are several challenges to be confronted. For example, depending on the child's hearing age, he or she may have difficulty distinguishing between voiced and voiceless consonants. In addition, consonants with articulation points in the back of the mouth may also be more difficult to perceive. Thus, parents should constantly provide the correct pattern of utterance. Following the phono-articulatory exercises performed at speech therapy sessions, any other circumstance can be used at home to practice pronunciation.

The home-practiced activities should follow a well-established hierarchy/continuum, from simple to complex. For example, phonemes should be introduced first in isolated forms, then in vowels, monosyllabic words, polysyllabic words, simple sentences, developed sentences, storytelling, and conversation. The vocabulary would then be composed of terms relevant to the child's immediate experience, then extended. As a basic principle, any word must be introduced in a meaningful structure. Practicing isolated words is less effective and less attractive to children. Instead, the parent shows the correct way of saying the words and tries to involve as many sensory modalities as possible (auditory, visual, tactile-kinesthetic) to help the child learn how to pronounce them.

The intelligibility in Context Scale [11] is a valuable tool for assessing speech intelligibility (in the Romanian context, the instrument has been translated, adapted, and validated and can be easily used by parents, including to monitor their children's progress). Verbal fluency is also a skill that can be trained and

improved. Children with cochlear implants will develop good speech fluency if their language proficiency and performance are close to the typical development. During the daily interaction with their child, parents can also contribute to developing their verbal fluency. Choosing the topic of the discussion, giving enough time to formulate the message, and reformulating are some efficient strategies that parents might use.

Another helpful discussion topic is voice specificity, a topic that has been the focus of various research studies [12-13]. The primary characteristics of the voice include *the pitch of the voice* (which is directly dependent on the movements/ripples of the vocal cords), *intensity* (which we perceive as the volume of the voice, being dependent on glottal pressure), *quality* (dependent on the resonant cavities), *timbre* (it gives color to the voice, a unique voice imprint to the speaker, distinguishing him from other speakers) [14]. However, some of these voice qualities can be affected in children with cochlear implants. The suprasegmental elements corresponding to the prosody are essential from a communication point of view. These elements require systematic learning in as diverse contexts as possible. The parents should use intuitive concrete support to facilitate their child's understanding.

Other practical activities that parents might engage in are related to role-playing games and the child's interpretation of some favorite characters. In addition, some applications allow the visualization of the child's speech (spectrogram), which can be used successfully in practicing the appropriate vocal emission (these applications usually run on Windows, Android, or iOS). Furthermore, the nonverbal and paraverbal language components are essential in empowering children with cochlear implants. The hearing parent will usually adhere to the verbal strategy for the child's language development. However, the use of natural and evocative gestures can facilitate the acquisition of verbal content by a child with a cochlear implant. We must not lose sight of the fact that some languages, including Romanian, are inflectional, and the accent and intonation contribute to clarifying the meaning of some words or sentences. Parents should also teach children to adopt and maintain a correct posture in various situations to communicate in the best possible conditions and get the most out of the cochlear implant.

Investigating school options

Children's transition to school also includes considering several educational options. First, the parents can opt for the nearest (geographically speaking) school, where children are usually placed by default. The choice of the neighborhood school has some advantages, such as the proximity to the house, the possibility for the child to acquire a certain level of autonomy gradually, and the existence of potential interactions with colleagues in extracurricular contexts. However, at least in Romania, not all schools are inclusive schools or places suitable for children with cochlear implants. For example, an integrative educational institution has specialized teachers for activities with various children with special educational needs (e.g., speech therapists, support teachers, school counselors). Parents might choose another school other than the neighborhood school when these specialists are absent. Nevertheless, several legal impediments make this step rather difficult in many cases.

Though the so-called special schools (designated exclusively for the hearing impaired) are an available option, they should not be the only choice or the first choice for several reasons related to children's rights and integration policy. Once the option for a particular school is formulated, parents can work with the school representatives to maximize all the available resources to ensure a smooth transition for their children. Molina and their collaborators [15] suggested that the benefits of including children with cochlear implants in public schools, along with typical children, include the following: (1) children learn to

respect others, accept differences, and recognize different skills, thus creating opportunities for the development of new friendships; (2) children learn about skills related to helping others participate and learn; and (3) children with cochlear implants benefit from the cognitive effort required to make themselves understood in typical contexts [15].

Knowing the school and the teachers

There are various ways in which parents might get involved in the relationship with the educational institution their implanted children will further attend, both formal and informal. For example, parents might participate in various extracurricular activities organized by the school, as volunteers or co-therapists, in different working and decision groups, such as the parent committee. Ideally, the parent is also part of a multidisciplinary team that includes classroom teachers, audiologists, general practitioners, pediatricians, support teachers, and speech therapists. Each of these specialists has a well-defined role in developing language and communication skills and adapting the children with cochlear implants to the student status requirements and school regulations and requirements. Before the actual school start, the child can visit the future classroom, get to know the teachers, and get acquainted with the new environment. These simple actions will further facilitate their children's transition because their final purpose is to facilitate social and educational integration by equalizing opportunities.

Different educational systems imply different educational structures, with various ways of teaching. In Romania, for example, from the preparatory class (i.e., the first grade, when children are 6-years old) to the fourth grade, the child interacts daily with the same primary teacher. For any child, and even more so for the child with a cochlear implant, it is essential to establish a close and supportive relationship. Experience and research showed that teacher empowerment is essential for integrating the child with a cochlear implant [16]. From the very beginning, parents and teachers should find effective ways to communicate, and the related discussions should have a concrete, operational character, which would further help clarify potential integration issues.

The parent-teacher communication will further help to determine how well the child responds to the instructions given to the whole class and/or individually, to determine the level of children's activism, the situations in which they have hearing difficulties, how their interactions with their classmates and other teachers who teach in that class are, how good their understanding about hearing impairment and hearing aids is, and which are the major difficulties that teachers face in their relationship with the child.

Parent-teacher discussions should also include the physical positioning of the child in the classroom, the volume of the voice, and the level of directivity in the child-teacher relationship. In many cases, the parents can be really helpful due to their better knowledge of their children's needs and sharing relevant information can empower teachers to provide the best education to their child with a cochlear implant.

Academic preparation: practical guidance for parents

Academic preparation for the transition to school is a summative process that requires practical efforts. The more the child practices the relevant skills at home, the more prepared they will be to participate in real future school situations. The training of these skills can be done naturally at home by involving the children in various activities and daily routines. School performance is also based on these skills, which can be practiced and developed as early as possible. In this case, we can consider (a) Self-care skills (washing, dressing, wearing shoes, eating, preparing the bag, and so on); (b) Development of motor skills

(speed, strength, endurance, dexterity, suppleness); (c) Development of fine motor skills and optical-manual coordination (pre-writing requirements); (d) Development of language and communication skills.

Furthermore, receptive and expressive oral language can be developed and/or enhanced by practicing the following components: (a) Perceptual focus on verbal data; (b) Receptive and expressive vocabulary; (c) Auditory verbal comprehension; (d) Verbal fluency (with executive and linguistic determinants); (e) Naming; Repetition (auditory attention and phono-articulatory praxis) (f) Self-expression through language. Examples of such related activities include listening games (for example, *Simon says*...), verbal-auditory integration exercises (verbal description of complex sounds, with imagistic support at hand), exercises for repeating phrases and sentences, poetry and puns, providing patterns of correct expression and developing the child's verbal formulations, using daily routines as a pretext for verbalization, reading picture books with the child, and last but not least, playing with the child.

Practical guidelines for teachers' involvement at school

As we have already pointed out, the parent knows their child better than anyone else. Likewise, teachers (dedicated teachers, especially) aspire to this status, as they also aim to know their students as well as possible. Therefore, when there is a solid and balanced relationship with the child's school and teachers, parents can provide information to help their child get the most out of education. Thus, the child's parents and teachers work together to support his or her learning and well-being. The practical results of a solid and efficient parent-teacher collaboration and team effort are usually demonstrated by the child's regular attendance at school, academic performance, a positive attitude towards school, good social and relational skills, and overall well-being. Nevertheless, the family's socioeconomic status and the related cultural characteristics can also impact the nature and degree of parental involvement. Therefore, even if not all parents can get involved as much as possible, everyone can convey the implicit and explicit message that school is essential for both the family and the child.

Formal learning must respond to seemingly contradictory commands. On the one hand, we have frontal group training, and on the other hand, the needs, the learning style, and the particularities of each student. Teachers should respond to this challenge and facilitate an effective, active, and lifelong teaching process [17]. In the case of students with a cochlear implant, teachers must judiciously combine traditional teaching methods and strategies with some that are adapted to these children's learning specifics. The teaching objectives are not limited to training but the formation and development of learning situations favorable to each individual in formal, informal, and non-formal contexts.

Active learning is a concept with a large area of coverage that refers to several instructive-educational models that involve the learner's responsibility and accountability in learning. To learn effectively, children must do more than listen. They must see, read, write, discuss, and be trained in problem-solving, covering three major key areas: knowledge (information), skills, and attitudes. Therefore, active learning involves children in two primary ways: by doing and also by analyzing (learning to synthesize and evaluate their activities). Thus, they will learn effectively, having clear objectives related to their learning and using various learning strategies and good management of resources (time, information sources, materials). Finally, children will build a realistic image of their strengths and weaknesses and will further relate positively to the learning contents, assuming the responsibility of training, planning, structuring, and evaluating the learning process.

The practical ways teachers can support the school integration of children with cochlear implants is also a teamwork effort. Thus, the primary intervention areas of interest are established together with the speech therapist, starting from the results of the integrated evaluations of *psychomotor skills* (e.g., motor coordination; perceptual-motor structures and behaviors; body scheme; temporal and spatial orientation), cognitive skills (e.g., associating contexts with specific actions; matching objects to the designated functions; reproduction of graphic signs and symbols), *language and communication skills* (motility of the phono-articulatory organs; phonatory breathing; speech intelligibility; passive and active vocabulary; enabling total contextual communication), *affective-emotional and behavioral skills* (e.g., personal and social autonomy; participation in structured activities).

Some suggestions can be made for meeting these goals in class. First, to ensure that messages are well passed on to parents (the child with a cochlear implant will not always fully understand the requirement and send it home), the teacher can use a dedicated social media channel to communicate (i.e., an updated version of the communication booklet). Also, children with cochlear implants should be placed in front of the class. The teacher can use a natural voice intensity and a medium speech rhythm (a high voice intensity tends to blur the consonants, the vowels remaining dominant). If it is a class in which the students usually sit, the teachers should choose a less noisy classmate for the child with a cochlear implant. The child should be placed in the middle row, not at the window or the classroom door, and their position should be correlated with the type of activity so that they can choose the position with the best audition in each situation. Teachers should also talk to the class and stop when they turn their back (for example, to write something on the board). Children with cochlear implants distinguish words better when they see the speaker's face. Furthermore, the teacher’s face should not be obstructed by any obstacles (hands, books, other teaching materials). Teachers can use natural gestures to emphasize the meaning of the words spoken without abusing them or moving excessively around the class while speaking. Some children have a single cochlear implant. In this situation, the teacher will consider the fact that the child hears best on the side with the implant. Also, because combining auditory and visual information maximizes the child's ability to understand, classroom lighting is also important. However, too little, or too much light (for example, sunlight enters directly through windows) makes it challenging to receive visual cues.

Despite the most careful preparations, there are situations in which children will not understand a particular message. It would be desirable to be taught how to signal this from the first days of school. An easy way would be, for example, to have two cards at hand, one with a smiling face and the other with a puzzled face, that would help communicate these issues effectively. Furthermore, various approaches can support their ability to understand what is happening around them. Each time the subject of the discussion changes, this should be marked. If it is an interactive lesson sequence, it is good for the teacher to synthesize the ideas expressed by the child’s classmates. In addition, from the very beginning of the preparatory class, the children are trained to have access to written information; children with a cochlear implant may also benefit from such visually presented information.

As there is a gap between the active vocabulary of the hearing child and that of the hearing-impaired child, it may also be helpful to recommend practicing various word lists, specific terms, or general topics at home. In addition, verbal messages can be repeated and reworded to make them easier to understand. Whenever possible, these messages will be accompanied by visual aids. The teachers should also keep in mind that the multisensory approach is the most effective in learning and teaching children with cochlear implants. However, if children do not understand some of the contents, they should have access to compensatory strategies that will help them overcome these issues.

Summary

A cochlear implant is a reliable way to restore hearing in children with severe or profound hearing loss. However, it does not automatically turn the child into a speaker. In order to form their speech and language, a sustained and combined effort is needed from the family, the school, and the specialists in hearing (re)habilitation. Family-centered intervention and specific communication development strategies generally work best for the child's desired development in the social and educational environment and school transition. The empowerment and rehabilitation programs family-centered intervention programs include two primary objectives. The first one is to strengthen the confidence of parents and children in their abilities. The second one is to provide parents with the knowledge needed to implement the language stimulation strategies and techniques needed for effective communication with their children [18]. The involvement of parents brings a multitude of benefits for both family and child, as well as school and community. First, it helps decrease parents' misconceptions about the school staff and vice versa regarding motives, attitudes, intentions, and abilities. Furthermore, it might also increase parents' roles as long-term resources for their children's academic, social, and psychological development. Moreover, parental involvement facilitates the development of parents' own skills and self-confidence, sometimes encouraging them to continue their education and get a better job, thus providing their children with better role models. This also further enhances parents' roles as school supporters in the community.

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