Hearing loss: etiology, identification and interventions

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Abstract

Hearing loss is characterized by increasing difficulty in hearing, interpreting and understanding the sounds. Although many acousticians are familiar with hearing trauma caused by loud sounds, a significant part of the acoustics community has limited knowledge on how hearing loss is identified, its consequences in everyday life, or the latest innovations in terms of management. This article summarizes the types and degrees of hearing loss, and its impact on quality of life. The main methods of evaluation (audiometry, tympanometry, acoustic reflex threshold, auditory evoked potentials) and treatments (hearing aids, cochlear implants, auditory brainstem implants) are also discussed to raise awareness of the day-to-day clinical audiology reality to the Canadian acoustic community.

Keywords: audiology, hearing loss, evaluation, hearing aids, bone conduction implants, cochlear implants, auditory brainstem implants

Résumé

La perte d'audition se caractérise par des difficultés croissantes à entendre, interpréter et comprendre les sons. Bien que de nombreux acousticiens soient conscients des traumatismes auditifs causés par les sons forts, une partie importante de la communauté acoustique a des connaissances limitées sur l'identification de la perte auditive, ses conséquences dans la vie quotidienne ou les dernières innovations en termes de prise en charge. Cet article résume les types et degrés de perte auditive, ainsi que leurs impacts sur la qualité de vie. Les principales méthodes d'évaluation (audiométrie, tympanométrie, réflexe stapédien, potentiels évoqués auditifs) et de traitement (prothèse auditive, implant cochléaire, implant du tronc cérébral) sont également abordées afin de sensibiliser la communauté de l'Acoustique Canadienne aux réalités de l'audiologie clinique quotidiennes.

Mots clefs: audiologie, surdité, évaluation, prothèses auditives, implants auditif en conduction osseuse, implants cochléaires, implants auditifs du tronc cérébral

1 Introduction

Hearing loss is an invisible disability that affects 1.5 billion people worldwide [1]. According to the WHO, this number could rise to 2.5 billion people by 2030. In addition to depriving a person's main sense used for communication, hearing loss is associated with depression [2], and social withdrawal [3]. More drastically, hearing loss is predictive of dementia [4]. For children, hearing loss is a major barrier to normal speech and language development [5]. This can lead to learning problems that ultimately hinders academic achievements. Annually, the cost of the global burden imposed by hearing loss is estimated to be US$ 960 billion [1].

This paper aims to report the most methods employed to identify hearing loss as well as the existing treatments and interventions. Section 2 reports what is hearing loss. The types, degrees and impact of hearing loss are presented in Sections 2.2, 2.3, and 2.4 respectively. Most common methods employed to evaluate the degree and type of hearing problems are described in Section 3. Sections 4 address the possible treatments and interventions. Conclusions are reported in Section 5.

2 What is Hearing Loss ?

Hearing involves the transduction of mechanical energy coming from sound waves to electrical signals that are interpreted by the brain. The peripheral auditory system can roughly be divided into three parts : the external, middle, and inner ear. Hearing loss arises when one or more of these components are disrupted resulting in lowered sensitivity to sounds when compared to normal hearing, or reduced speech intelligibility.

The hearing sensitivity, or degree of hearing, is evaluated by measuring the hearing threshold for pure tones of different frequency, typically from 250 to 8000 Hz. The degree of hearing loss is based on these thresholds measured with earphones, which are also referred to as air conduction (AC)

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thresholds. Classification of hearing loss is based on a continuum from mild (thresholds between 25 and 40 dB(HL)) to profound (95 dB(HL) or more). It is common for the degree of hearing loss to be different from one frequency region (low, mid or high frequencies) to the other.

Another type of transducers used in audiometry is the bone conduction (BC) vibrator. The BC vibrator is placed against the skull, usually the mastoid bone, to obtain BC thresholds, which essentially represent the response of the inner ear. Bone conduction testing bypasses the conductive portions of the auditory pathways (outer and middle ear) [6]. Hearing loss is characterized by the nature (type) of hearing loss, and the degree of hearing loss at different frequencies in each ear. This information allows clinicians to approximate expected functional difficulties, to recommend personalized treatment plans, and to establish baselines for future auditory surveillance. Hearing loss is often visually represented on an audiogram where the tested frequencies are on the x-axis and the hearing threshold is on the y-axis (see Fig. 1).

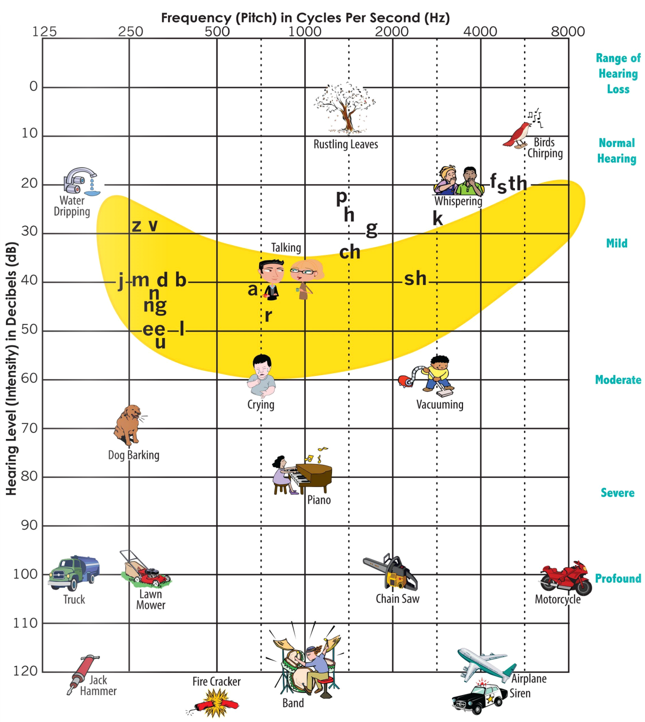

FIGURE 1 – Audiogram of familiar sounds. Image courtesy of John Tracy Center (https ://www.jetv.org).

2.1 How is hearing loss measured ?

Audiometry is performed in a soundproof booth where hearing acuity is assessed for different tones and/or different speech stimuli. Pure tone audiometry is a fast and simple way to assess a patient's hearing. Sensitivity to sounds is generally assessed at different frequencies (usually between 0.25 – 8 kHz) for each ear, but more and more clinics also evaluate the hearing thresholds in the extended high-frequency range (10 to 20 kHz) to monitor the effect of chemotherapy on the hearing system for example, or other ototoxic medication [7]. The threshold is defined as the lowest intensity at which the patient hears the tone at least 50% of the time [8]. The thresholds are used to determine the degree of hearing loss.

Pure tone audiometry can be performed via air conduction (AC) or bone-conduction (BC). Air-conduction thresholds are determined as the sounds travel through all the structures of the ear. They are evaluated using headphones. Bone-conduction thresholds are obtained by using a bone vibrator which transmits the sound through the skull directly to the inner ear, bypassing the external and middle ear [9]. By comparing the results obtained by these methods, clinicians can determine the nature of the hearing loss.

The results for each ear are represented on an audiogram where the x-axis represents the tested frequencies, and the y-axis represents sound intensity. The symbols on the audiogram will usually represent the thresholds. This graphical representation allows clinicians to quickly identify hearing loss configurations. A hearing impairment that is progressively worse in the high frequencies (commonly called "ski slope" due to its particular shape) is often associated with the age-related hearing loss. Hearing loss centered around 4kHz can be observed in cases of noise-induced hearing loss. A hearing loss in the middle frequencies with normal thresholds in the lows and the highs (cookie-bite) can be associated to congenital hearing loss [10].

Validity of the test can be compromised if the patient does not fully understand the instructions (e.g. language barrier or young children), the patient is not attentive (e.g. fatigue), or if the patient is a malingerer. It is always important to crosscheck the results of pure tone audiometry with other tests.

2.2 Types of Hearing Loss

Sensorineural Hearing Loss

A sensorineural hearing loss is identified when the hearing thresholds are at the same level in air and bone conduction (see Fig. 2a). It occurs when there is damage to the inner ear and/or damage to the nerve that relays information from the ear to the brain. Noise exposure, ototoxic medication, and aging are, among others, causes of this type of hearing loss. The hair cells of the inner ear (the cochlea) are not only responsible for the detection of sounds, but also for providing the ability to discriminate between different sounds [11]. Thus, degradation of these hair cells will not only result in a reduction of perceived loudness but also a reduction in the clarity of sounds. In some rare cases, the information transmitted from the cochlea to the brain can become desynchronized. This condition called Auditory Neuropathy Spectrum Disorder (ANSD) results in compromised speech understanding accompanied by variable hearing loss [12]. For most cases, sensorineural hearing loss is permanent.

Conductive Hearing Loss

A conductive hearing loss is identified when the hearing thresholds obtained with bone conduction are significantly bet-

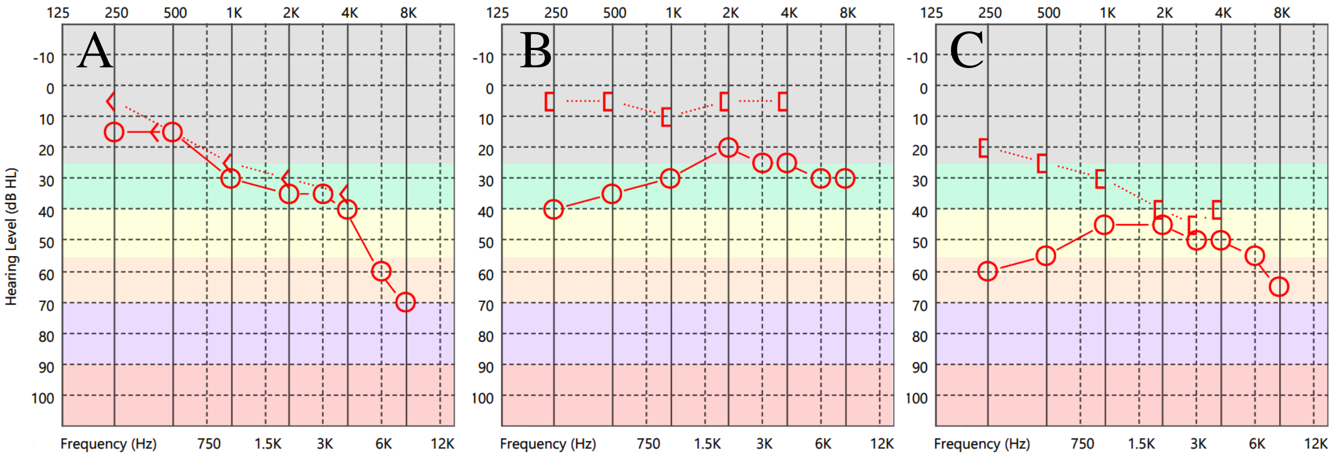

FIGURE 2 – Example of audiograms. A : sensorineural hearing loss in high frequency assessed on the right ear. Both air-conducted (◯ symbols) and bone-conducted (< symbols) thresholds show normal hearing from 250 to 500 Hz followed by a mild sloping to moderately severe sensorineural hearing loss from 1 to 8 kHz. B : conductive hearing loss assessed on the right ear. The masked bone-conducted thresholds are within normal range but the air-conducted thresholds are in the mild range. C : mixed hearing loss assessed on the right ear. Both air-conducted and masked bone-conducted ([symbols) thresholds are in the abnormal range, with a significant air–bone gap, also called AB gap, greater $\geq$ 15 dB in the low frequency range. Normal hearing is indicated by the grey area. Green, yellow, orange, purple and red areas respectively denote mild, moderate, moderately severe, severe, and profound hearing loss.

ter (difference greater than 10 dB) than those obtained with air conduction (see Fig. 2b). It occurs when sound waves are not transmitted efficiently through the outer and/or the middle ear. These first two parts of the peripheral auditory system mainly serve as conductive parts of the ear, as well as amplifiers for the sound waves before reaching the inner ear. Examples of outer ear complications include excessive accumulation of cerumen and auditory canal malformations, such as exostosis which are common in cold water divers. Ear infections (otitis media), tympanic membrane perforation, and damage to the ossicles are potential contributors to conductive hearing loss involving the middle ear [11]. When a sound is loud enough, it can overcome the loss of natural amplification. Most cases of conductive hearing loss s are reversible and can be treated. However, when no intervention is possible, the hearing impairment will persist.

Mixed Hearing Loss

A hearing loss that occurs in both the conductive and sensorineural arts of the ear is called a mixed hearing loss. The hearing thresholds obtained with bone conduction are significantly better (difference greater than 10 dB) than those obtained with air conduction (see Fig. 2c), but in both cases that are showing a hearing loss. Mixed hearing loss (see Fig. 1c) is the combination of conductive and sensorineural hearing loss. While the conductive component can possibly be treated, the sensorineural component is considered a permanent hearing loss. An example of mixed hearing loss is an older individual with age-related hearing loss that presents with a perforated ear drum. The sensorineural component of the hearing loss is associated to presbycusis whereas the conductive component of the hearing loss is associated to the perforated ear drum.

Central Hearing Loss

Auditory information is relayed from the peripheral auditory system to the central auditory system via the auditory nerve. The signal passes through several structures before reaching the auditory cortex. These structures include cochlear nucleus, superior olivary nuclei, lateral lemniscus, inferior colliculus, and medial geniculate nuclei. More complex processing of the auditory information is carried out as it ascends towards the central auditory system. Additionally, auditory information is also influenced by top-down control via the medical olivocochlear (MOC) efferent system. It is believed that this efferent system contributes to protect the cochlea from intense sounds as well as facilitate speech-in-noise recognition [13].

Central hearing loss occurs when there are alterations or damage to these higher order structures. In a typical audiology clinic, it is not possible to directly access the integrity of these structures. However, central hearing loss often manifests itself as behavioral difficulties that include, but that are not limited to worse-than-expected difficulties with hearing in noise, difficulties with sound localization, difficulties following rapid speech, academic difficulties related to reading or spelling, difficulties paying attention, and frequent requests for repetition. These symptoms are observed despite normal results for the peripheral auditory system evaluation [11]. Speech in noise tests can be used to document these difficulties. Additionally, evaluation of the auditory processing abilities can be performed to attempt to identify specific weaknesses relating to higher order sound processing.

2.3 Degree of Hearing Loss

In addition to its nature, hearing loss is also characterized by a degree or severity. The degree of hearing loss ranges from mild to profound. It is determined as how loud a sound needs

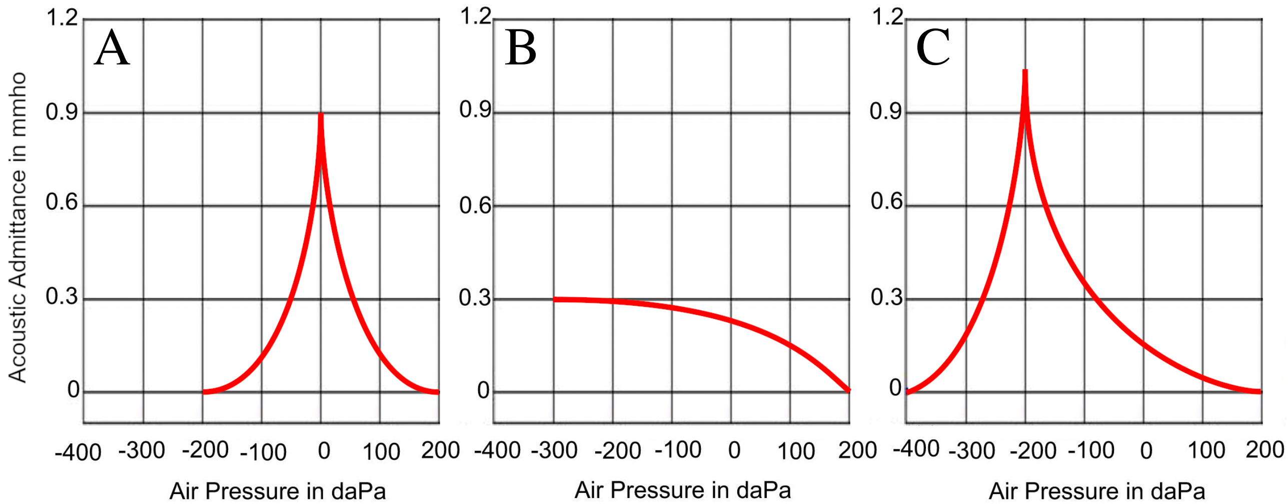

FIGURE 3 – Common types of tympanographs. Example A illustrates a tympanogram showing normal middle ear pressure. Example B is typical in case of otitis media with effusion, space-occupying lesions of the tympanic cavity, and tympanic membrane perforations. Example C present a tympanogram showing negative middle ear pressure as reflected by the negative pressure peak, which is associated with eustachian tube disfunction.

to be for an individual to hear it (in dB HL). The cut-offs for the various degrees of hearing loss can be found in Table 2 from [14]. The severity of hearing loss allows clinicians to describe a hearing loss and obtain a general idea of the expected difficulties.

2.4 Impact of Hearing Loss

The first listening difficulties experienced by individuals suffering from hearing loss may include : muffled speech sounds, difficulty understanding speech in noise, confusing certain words, asking others to repeat frequently, having to increase the TV or radio volume. As hearing loss worsens hearing loss can lead to withdrawal from conversations, social isolation, and is even associated with an increased risk of dementia. For children, hearing loss can interfere with access to spoken language, which can lead to cognitive delays as the areas of their brain used for communication may not develop appropriately [15].

3 Audiological evaluation

Beyond tonal audiometry, different tests are used to gather information about the auditory system, and to validate results. These tests include speech audiometry, immitancemetry, otoacoustic emissions, and auditory evoked potentials. The combined interpretation of these various tests help establish a portrait of one's hearing. Unfortunately, our current evaluation methods are not able to fully assess the intricacies of the auditory system. In the following section, we will discuss the main techniques used in audiology for differential evaluation, and the shortcomings of our current assessment methods.

3.1 Speech Audiometry

Two common speech evaluations performed in clinic is the speech reception threshold (SRT) and word recognition score (WRS). For the SRT, patients are asked to repeat spondaic words (two-syllable words with equal stress on each syllable). The SRT is used to validate the results obtained via tonal audiometry. It has been shown that the SRT is approximately equal to the pure tone average (500, 1000, 2000 Hz). For the WRS, patients are asked to repeat mono-syllable words at a comfortable intensity level. The goal is to determine if there is any degradation of clarity when enough volume is provided for the patient [9].

The speech measures described above are presented in quiet conditions. There are, however, other speech measures being used to evaluate how well a patient can hear when in presence of background noise. The use of speech-in-noise tests such as the HINT [16] or the QuickSIN [17] are being progressively included in routine audiological assessments (see below). Speech in noise tests provide a more accurate representation of an individual's real-world performance [18] as it is rare for many patient's daily listening situation to be as quiet as a sound booth. When using speech material, it is particularly important to considerate the patient's most comfortable language. Poor performances may be due to poor understanding of a language rather than hearing difficulties.

3.2 Immittancemetry

Tympanometry

Tympanometry is used to evaluate the middle ear function by measuring the compliance of the tympanic membrane (see Fig. 3). The tympanometer probe includes a miniaturized speaker (for emitting the stimulating sound), a probe microphone (for recording the reflected sound) and an air pump (for altering the pressure in the sealed ear canal). Tympanometers measure the energy absorbed by the eardrum to the sound stimulus emitted by the probe speaker (typically a 226 Hz or a 1000 Hz pure tone but multi-frequency tympanometry can be used to identify ossicular abnormalities, e.g. malformations/diseases affecting the ossicles [19]). When a sound is emitted, part of it is absorbed while the remaining part is reflected and can be captured by a miniaturized microphone to compute the energy absorbed by the eardrum. In a normal situation the eardrum will have a maximum absorption and the collected sound will be very weak. In the case of serous otitis or poor ventilation of the middle ear, the pressure in the external auditory canal and in the middle ear will be unbalanced, resulting in a lower absorption of the eardrum and therefore a higher reflected sound. In order to evaluate compliance, the pressure in the external auditory canal is adjusted using an air pump to compensate and re-equilibrate the pressure on both sides of the eardrum. This pressure variation is presented in "mm of water" (mmH_2O) and can be negative or positive. The peak of a tympanogram is where the eardrum is balanced and allows to determine if the tympanic membrane moves freely or if a pathology is present.

Additionally, wide band tympanometry (WBT) can provide more complete information about the middle ear. With one recording, middle ear function is assessed using a range of frequencies (typically between 226 and 8000 Hz) rather than the traditional 226 Hz or 1000 Hz. WBT is believed to be more reliable for assessing the middle ear in infants [20]. Furthermore, WBT could be a useful method for tracking the progression of otosclerosis [21]. It has even been suggested that WBT can help with the diagnosis of Menière's disease [22].

Acoustic Reflex Threshold

The acoustic reflex threshold (ART) is defined as the minimal sound level pressure required to trigger the acoustic reflex (usually between 70-105 dB SPL). This reflex aims to protect our ears from loud sounds. In response to intense sounds, the stapedius muscle contracts to stiffen the ossicular chain [9]. This results in a dampening of the vibrations that are transmitted to the inner ear. The acoustic reflex pathway (see Fig. 4a) involves several key structures of the ear, such as the middle ear, the inner ear, the vestibulocochlear nerve (VIII), the cochlear nucleus, the superior olivary complex, and the facial nerve (VII).

By evaluating the presence of the acoustic reflex in different stimulation and recording conditions, clinicians can get additional information regarding the site of lesion. To evaluate the acoustic reflex, a tone is presented at different intensities (usually 70 to 105 dB) in one ear. While the tone is being presented, acoustic immittance is being measured in the same ear (ipsilateral) or the opposite ear (contralateral). A reflex is considered present when the change in immittance is greater than a pre-determined cut-off criterion. The acoustic reflex is evaluated in both ipsilateral and contralateral conditions for both ears resulting in a pattern of absent or present reflexes. The acoustic reflex pattern provides additional information to complete / confirm the results of other audiological evaluations (see Fig. 4b). It is important to note that the site of lesion cannot be determined based solely on the results of this test.

Unfortunately, the acoustic reflex is known to yield many false positives. It has mostly fallen out of favor. In the event that retrocochlear involvement is suspected, the MRI is the gold standard, especially since the availability of MRI has improved. Since a loud sound is used to trigger the ART, this method is avoided with hypersensitive patients and those suffering from tinnitus.

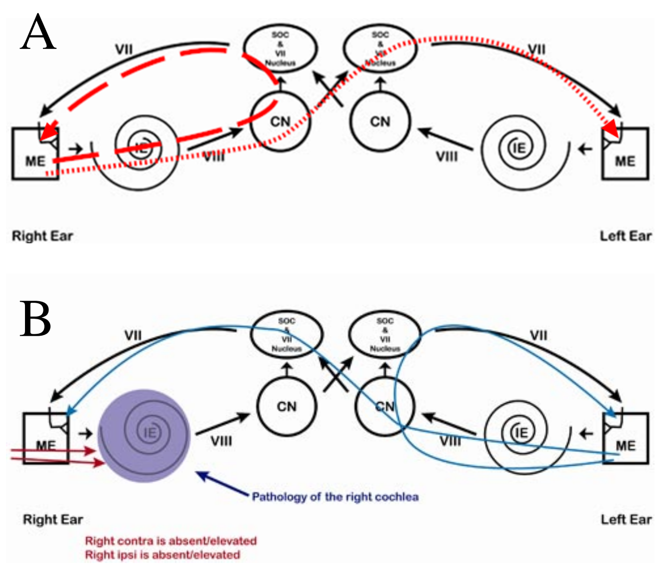

Figure 4 – A : Acoustic stapedial reflex model. The ipsilateral and contralateral pathway are indicated respectively by the long dashes and dotted lines. B : Cochlear pathology, right ear. Note that right ipsilateral and right contralateral ARTs are elevated/absent and left ipsilateral and left contralateral ARTs are present. ME = middle ear, IE = inner ear, VIII = vestibulocochlear nerve, CN = cochlear nucleus, SOC = superior olivary complex, VII = facial nerve. Modified from the original figure, courtesy of D. Emanuel [23].

3.3 Distortion Product Otoacoustic Emissions

The role of the outer hair cells is to amplify weak sounds thanks to their actions on the basilar membrane. The counter reaction to this amplification is a "relaxation" of the basilar membrane which induces a movement of the cochlear fluid, the oval window, the ossicles, and finally the eardrum. Consequently, the eardrum will act like as a loudspeaker that will diffuse a sound instead of vibrating because of a sound. This sound generated by the eardrum is very weak and remains inaudible to the human ear, but a microphone can capture it. Such sounds created by the ear itself are called otoemissions and can be spontaneous (i.e. generated in the absence of any stimulation), or provoked (i.e. produced in response to a stimulus).

When two simultaneous tones of different frequencies are used, the resulting otoacoustic emission is called Distortion product otoacoustic emissions (DPOAEs). DPOAEs are extensively used in pediatric hearing screening routine as a way to obtain non-invasive, quick and reliable information regarding the cochlear status of the neonates. Additionally, DPOAEs are used in several ototoxicity monitoring protocols. Damage to the fragile hair cells caused by ototoxic medication can occasionally be detected via DPOAE even before having any impact on the audiogram (see Fig. 5).

The DPOAEs are a very valuable objective screening methods, especially with the pediatric population. The presence of responses can rule out any hearing loss worse than a mild hearing loss at the tested frequencies. DPOAEs also provide ear specific information. The main drawback of this method is that it requires normal middle ear function for a valid test. A valid test also requires a relatively quiet environment. This is usually not an issue with adults but can become problematic with a child resisting the placement of the probe in the ear.

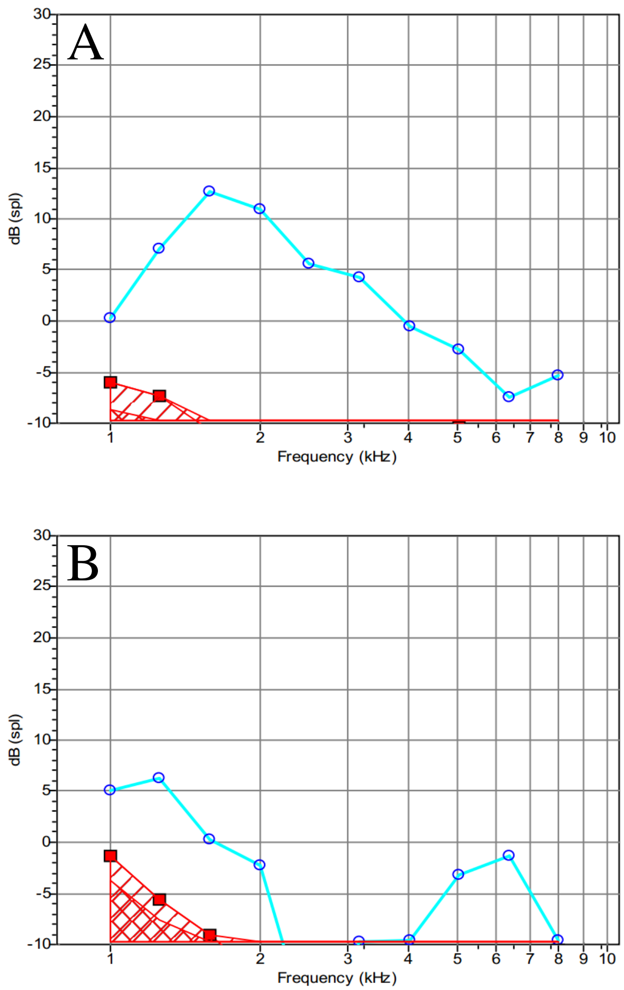

FIGURE 5 – Examples of DPOAEs. A : Presence of DPOAEs for most tested frequency suggest the integrity of inner cochlear hair cells. B : Absent DPOAEs in the 2-5 kHz range suggest damage to inner cochlear hair cells. Noise levels are indicated by the red line and squares.

3.4 Auditory Evoked Potentials

Auditory evoked potentials (AEPs) are an electrical manifestation of the brain response to an auditory stimulus. AEPs can be elicited by using brief acoustic stimuli as clicks or tone pipes to monitor the function of the inner ear or the neural auditory pathways during surgery. When a sound is processed by the auditory system, it elicits a signal arising from different anatomical generators and at latencies that range from a few milliseconds to hundreds of milliseconds.

The earliest components are generated in the cochlea and can be recorded using electro-cochleography from the middle ear ("transtympanic") or from the auditory canal ("extratympanic"). Electrocochleography recordings are often use for the diagnosis of Ménière disease, and for the intraoperative monitoring of the cochlear and eighth nerve. Subsequent neural responses can be divided into three latency classes [24] : early-latency responses, arising in the first ten msec after the stimulation, long-latency responses, with latencies greater than 50 ms, and middle-latency responses, with intermediate latencies (see Fig. 6).

The early-latency responses are the most often used for clinical purposes. They are relatively easy to record, and their wave shapes and component peak latencies are highly consistent across normal subjects. Sedation and surgical anesthesia produce only minor changes in early-latency responses. Early evoked potentials consist of seven small positive deflections, numbered I to VII, reflecting the passage of auditory information in different neural structures : wave I is generated by the fibers of the auditory nerve afferent to the inner hair cells ; wave II is generated by the passage of nerve impulses through the auditory nerve as well as by the entry into the cochlear nucleus ; wave III is mainly generated by the exit of the cochlear nucleus, as well as by the entry into the superior olivary complex ; wave IV is thought to be generated primarily by the lateral lemniscus and wave V by the inferior colliculus ; and although this is poorly defined because of their instability, waves VI and VII are thought to be generated in the medial geniculate body [25]. Early-latency responses are most often called "auditory brainstem responses" (ABR), even though the term is not completely accurate since wave I and part of wave II are generated in the auditory nerve rather than the brainstem itself.

ABRs latency decreases as the stimulation level increases [26] : the presence of the five well individualized waves is obvious at 70 dB but as the stimulation level decreases, the latency increases, and the amplitude of the waves decreases (see Fig. 7). The objective audiological threshold is defined by the minimum stimulation level that allows a clearly identifiable wave V, which is 20 dB in this example. Tone-ABR thresholds in patients with sensorineural hearing loss are typically 5 to 15 dB higher than their pure-tone behavioral threshold [27].

Auditory Evoked Potentials recorded in normal hearing subjects differ significantly from those recorded in hearing-impaired subjects : the five waves are easily discernable using a stimulation level of 60 dB when ABRs are scalp-recorded on a normal hearing child, but the same measurement performed on a deaf child does not allow to detect any wave (see Fig. 8). Interpeak latency between wave I and V can be used in the diagnosis of retrocochlear (i.e. beyond the cochlea) lesions : values higher than 4.70 ms indicate existence of retrocochlear lesions [29]. The comparison of wave V latency values in both ears can also be used in diagnosis retro-

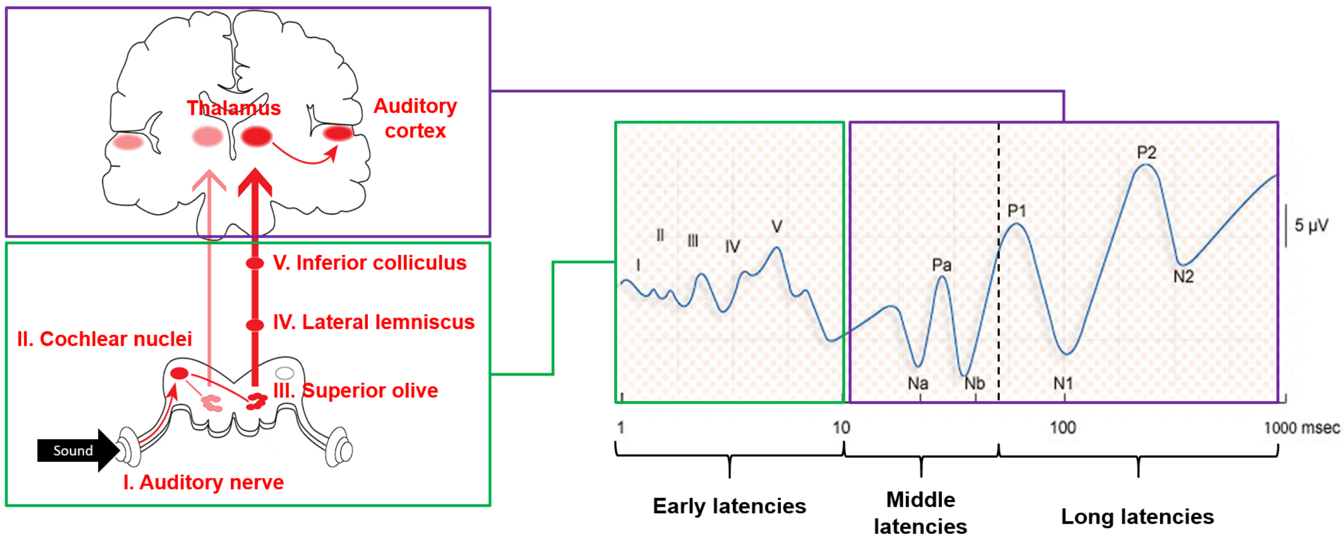

FIGURE 6 – Auditory evoked potentials in normal hearing subjects. Modified from the original figures by S. Blatrix and P. Minary by adding arrows, text, and anatomical brain structures on the left.

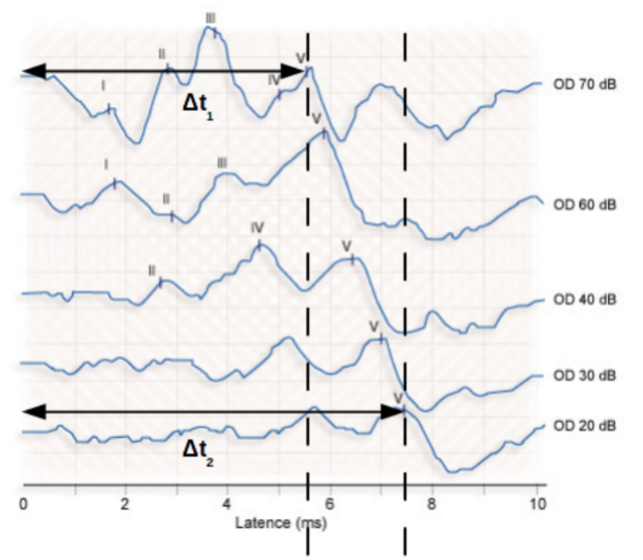

FIGURE 7 – ABRs recorded at different stimulation levels. The latency (Δt) decreases as the stimulation level increase. Modified by adding arrows and text from the original figure by S. Blatrix and P. Minary.

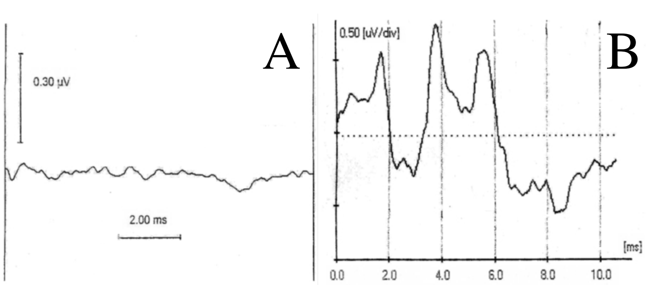

FIGURE 8 – ABRs recorded on the right ear of a deaf child (A) versus those recorded in a normal-hearing child (B) with a stimulation level set to 60 dB. Original figure from O. Valentin [28].

cochlear lesions : an interaural difference great than 0.3 ms indicates retrocochlear lesions [30]. Patients with auditory neuropathy due to a neurodegenerative disease or with a progressive demyelinating pathology present absent or severely abnormal ABR [31]. Acoustic neuroma, a rare slow-growing non-malignant tumor of the 8th cranial nerve also called ”vestibular schwannomas” induces a prolonged I-III and I-V delay [32].

One of the main drawbacks of AEPs is that this method requires time-consuming preparation of the skin surface to reduce skin–electrode electrical impedance, and EEG signal acquisition systems that are bulky. Part of this drawback can be overcome by using a portable EEG amplifier [33] and unobtrusive sensors [34], but such devices are not yet available for daily clinical use. Furthermore, like all electrophysiological measurement, AEPs are very sensitive to motion, making this method highly challenging with uncooperative patient.

3.5 Shortcomings

The current clinical test battery poorly represents one’s real-world hearing abilities. Patients are tested in a controlled clinical environment that lacks the complexity of real-world listening. Although, there are attempts at developing more ecological-valid assessment methods (e.g. speech-in-noise testing), our assessments method still do not take into account important factors contributing to one’s listening abilities. Listening effort, cognitive ability, and one’s ability to utilize visual cues are rarely assess and may help explain the discrepancy between clinical results and real-world performance [35].

Furthermore, the audiogram cannot fully encompass the complexity of hearing impairment. In other words, different individuals with the same audiogram may have very different functional difficulties. In the case of hidden hearing loss or cochlear synaptopathy, a patient may present with signifi-

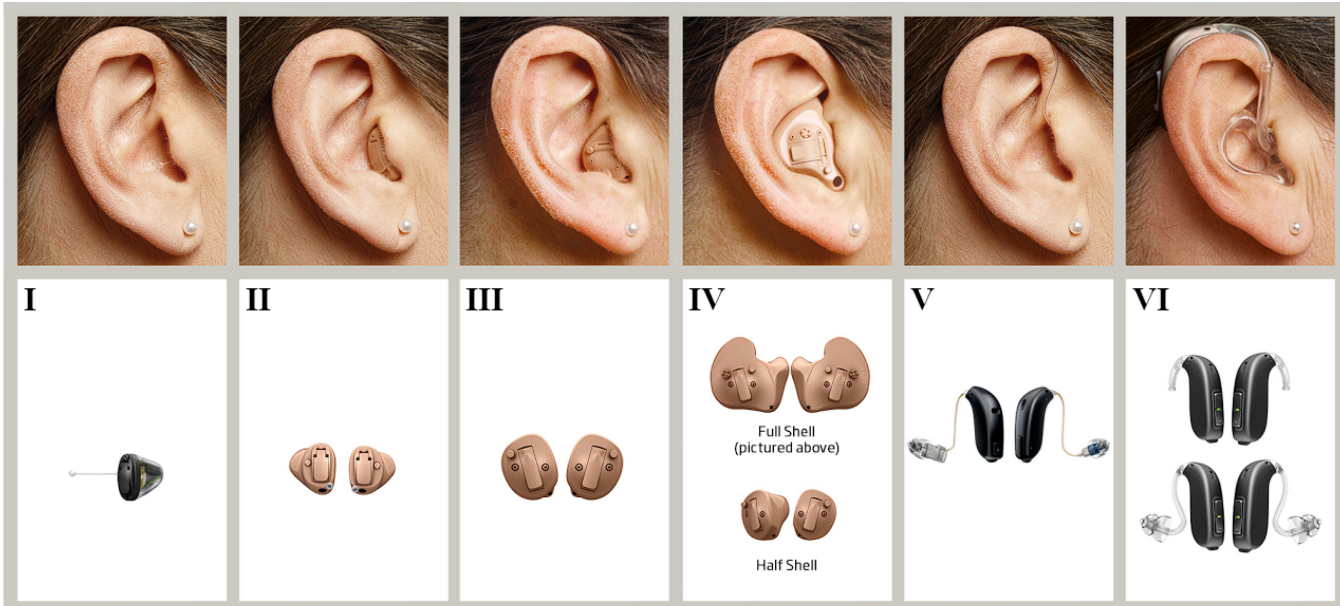

FIGURE 9 – In-the-ear hearing aids (I to IV) versus behind-the-ear hearing aids (V and VI). Modified from the original figure, courtesy of Oticon Medical.

cant listening difficulties despite a normal audiogram. It is believed that the hearing impairment arises from a disconnect between the auditory-nerve fibers and the cochlear hair cells [36]. Also, animal studies have shown that an audiogram is insensitive to inner hair cell damage. Normal hearing thresholds could be obtained following carboplatin-induced selective destruction of inner hair cells, the main afferent pathway originating from the cochlea [37]. Currently, there are no non-invasive methods of evaluating these deficits.

4 Management of hearing loss

4.1 Hearing Aids

Hearing aids are the most common intervention used to help with hearing impairment. The hearing aid is a small device that is equipped with a small microphone and a receiver. Its goal is to pick up the sounds from the environment and deliver an amplified version to the hearing aid user. There are several different types of hearing aids that each have their advantages and disadvantages. Discussion with an audiologist or a hearing aid dispenser is essential for choosing the right hearing aid (See Fig. 9).

The hearing aid is not only a simple sound amplifier. Sounds that are picked up by the microphone are decomposed into several channels. Each channel covers a different frequency range that is processed and amplified differently. Effectively, this allows the hearing aid to be personalized to everyone's hearing loss, as well as perform additional processing such as noise reduction and feedback cancelling. Additionally, hearing aids also employ an amplification strategy called wide dynamic range compression (WDRC) where soft sounds are amplified more than louder sounds. WDRC restores audibility and, at the same time, maintains comfort [39].

Hearing aids can also be programmed with different "presets" for different environments. For example, a standard program will use omnidirectional microphones with mild noise reduction. This allows users to benefit from amplification in most listening situations. However, said program would likely struggle in a noise environment such as a restaurant. For these situations, the hearing aid user would rather benefit from a directional microphone with more aggressive noise reduction. Modern hearing aids are even able to detect different listening situations and automatically switch to the appropriate program.

It is important to note that it requires some adaptation time to get used to the hearing aid. Often, patients are overwhelmed by all the new sounds they can hear again. With time, the brain will get used to these sounds and tune them out [40].

CROS and BiCROS Hearing Aids

In cases of single side deafness or asymmetric hearing loss, a "contralateral routing of signal" hearing aid (CROS) can be considered. The main purpose of these devices is to capture sounds from the individual's hard of hearing side and transfer it to the better side [41]. To do so, the user must wear one hearing device on each ear : a "dummy" hearing aid that serves as the microphone on the hard of hearing side, and a standard hearing aid on the better hearing side. One unfortunate consequence of this hearing system is the need to wear a hearing aid on the good ear. Furthermore, CROS aids only transfer information from one side to the other – it does not restore binaural hearing. Despite these inconveniences, some patients appreciate the CROS as it is non-invasive solution that restores sound awareness on their worse hearing side. The BiCROS functions similarly to the CROS but also provides amplification on the better side.

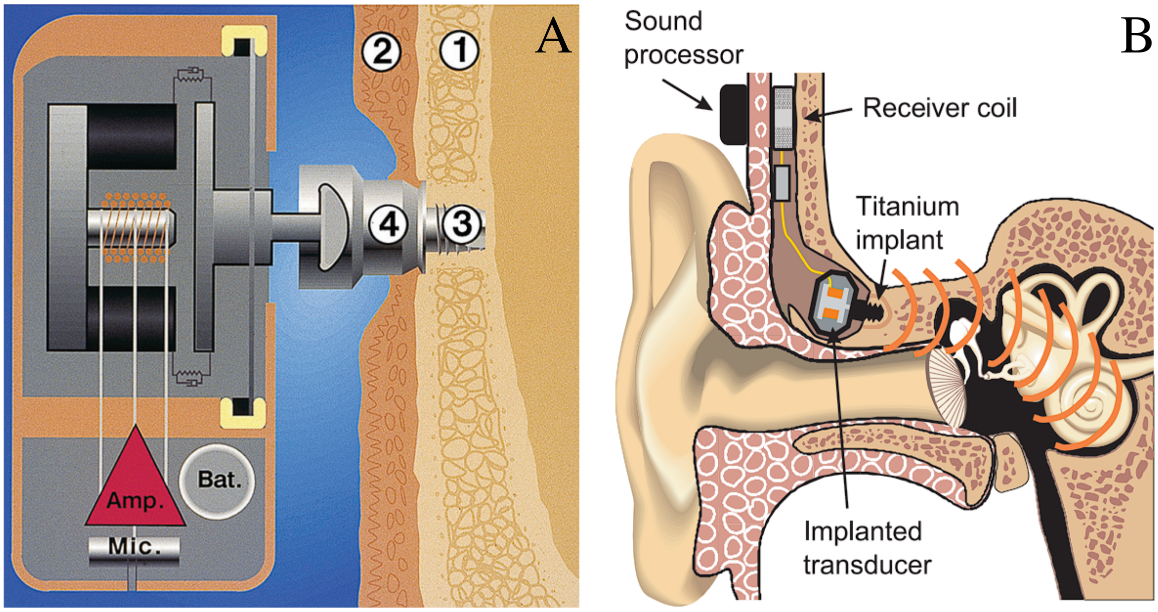

FIGURE 10 – Percutaneous (A) versus transcutaneous (B) bone conduction implants. Image courtesy of B. Håkansson [38].

4.2 Bone Conduction Implants

Bone conduction hearing devices are specialized types of hearing aid. Instead of outputting sound through air conduction these devices vibrate the skull transmits the sounds directly to the cochlea through bone conduction. These devices are considered for patients who have recurrent ear infections or malformation of the external auditory canal as they allow amplification to bypass the outer ear. In most cases, bone conduction hearing devices require a surgical procedure to implant an abutment into the skull. The bone conduction hearing device is then attached to the abutment allowing the vibrations to more effectively be transmitted through the skull to the cochlea. Bone conduction implants can be percutaneous (i.e. the transducer is directly coupled to the bone by means of a permanent skin penetration, like the BAHA®system from Cochlear™ or the Ponto™system from Oticon Medical, see Fig. 10A) or transcutaneous (i.e. one part of the transducer is implanted and the other part is kept outside the intact skin and soft tissue, like the Osia®system from Cochlear™ or the ADHEAR system from MED-EL™, see Fig. 10B).

4.3 Cochlear Implants

The cochlear implant (CI, see Fig. 11) is a biomedical device used to overcome profound deafness by replacing the function of a damaged or destroyed cochlea [42]. The implant consists of two main components. The external part includes the sound processor and microphone and is placed behind the ear. The internal part is placed under the skin and includes an electrode array that is inserted into the cochlea (see Fig. 12).

How does a cochlear implant work ?

First, the acoustic information is picked up by a miniaturized microphone placed in the behind-the-ear component. The sound is then transmitted to a speech microprocessor which analyses and digitises the sound captured by the microphone. After this signal processing step, the stimuli are sent to the external antenna located in the retroauricular region and then to the receiver/stimulator implanted in the temporal bone by radio wave, under the skin. Finally, the receiver/stimulator converts the signals transmitted by the antenna into electrical impulses that are sent to the electrode array to directly stimulate the auditory nerve : the electric impulses transmitted to the brain are then interpreted as sound.

Speech processors and coding strategies

The cochlear implant sound processor, which patients wear over-the-ear like a hearing aid, plays a critical role in the restoration of hearing. Many speech coding strategies have been developed over the past thirty years to the mimic firing patterns inside the cochlea as naturally as possible. Existing coding strategies can be regrouped into three categories : rate coding strategies which emphasis on the temporal representation of the signal [43, 44], place coding strategies which emphasis on the spectral representation of the signal [45], and hybrid coding strategies who combine both place and rate strategies [46]. Among the wide variations of coding strategies, some use a fixed number of channels to reproduce the original sound spectrum, while others used a virtual channel technique to increase the number of electrodes to achieve a better spectral resolution.

The default strategy in all implants uses a fixed set of electrodes to deliver biphasic electrical pulses in a non-overlapping fashion, e.g., by only stimulating one electrode at a time to avoid interactions. The sound signal collected by the external microphone is first pre-emphasized for frequencies above 1.2kHz at 6dB per octave and then separate into several bands using band-pass filters (one per stimula-

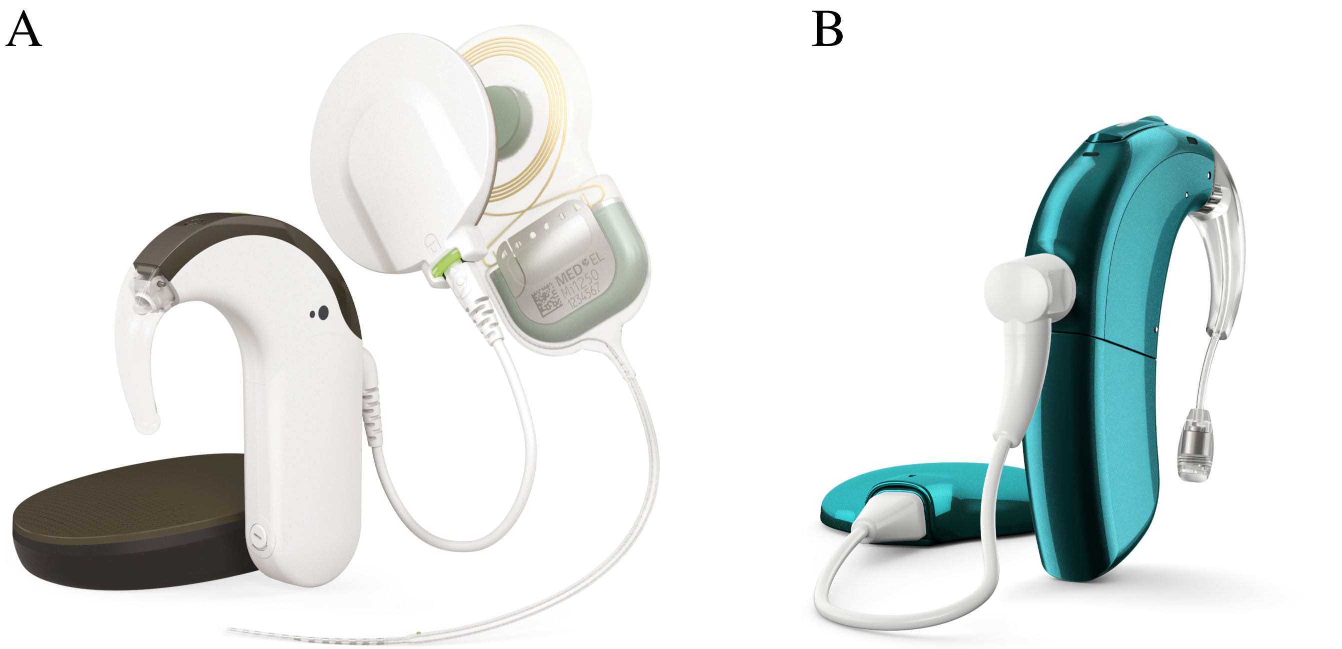

FIGURE 11 – A : external and internal components of the MED-EL SYNCRONY 2 cochlear implant system (image courtesy of MED-EL). B : CI's external components can be tailored specially for children, as the Sky CI™ M sound processor designed for kids by Advanced Bionics (image courtesy of Advanced Bionics).

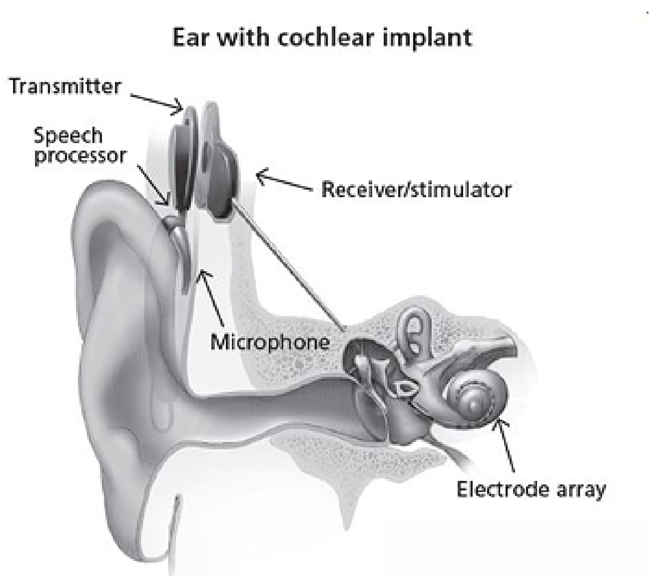

FIGURE 12 – Ear with cochlear implant. Image courtesy of the National Institute on Deafness and Other Communication Disorders (NIDCD, https ://www.nidcd.nih.gov/).

ting electrode). Each band is then rectified and low-pass filtered to obtain the signal envelope which is dynamically compressed using a nonlinear mapping function in order to map the wide dynamic range of sound in the environment (up to about 100 dB) into the narrow dynamic range of electrically evoked hearing (about 10 dB). The envelopes are finally used to modulate the biphasic electrical pulses sent to the stimulating electrodes. One limitation of this strategy is the number of implanted electrodes used to reproduce the sound signal versus the number of auditory nerve fibers (around 30,000). Since the electrodes can only stimulate a limited number of auditory nerve fibers, the resolution and information received by a CI user remains limited.

Drawbacks and limitations

It is undeniable that the cochlear implant is a life-changing device. However, this technology is not without its drawbacks and limitations. Acquiring a cochlear implant involves multiple visits for candidacy evaluation, surgery, device programming. This process can be quite prohibitive for patients who live far away from implant centres. Furthermore, patients must commit to a long and arduous rehabilitation process to learn or "re-learn" how to hear. Unfortunately, even after rehabilitation, there remains a large variability in benefits experienced by cochlear implant recipients. Some patients perform poorly than expected or will show little improvement. Finally, patients will have to adjust to living with implanted electronic device. The cochlear implant can be problematic with MRIs. It is also non compatible with certain medical intervention such as neurostimulation, electrical surgery, and ionic radiation therapy.

4.4 Auditory Brainstem Implants

Auditory brainstem implants (ABIs, see Fig. 13) were first developed nearly 40 years ago to provide hearing to people with hearing loss who can't benefit from a hearing aid or cochlear implant [48]. This is most commonly due to a missing or very small hearing nerve or severely abnormal inner ear (cochlea). The auditory brainstem implant directly stimulates the hearing pathways in the brainstem, bypassing the inner ear and hearing nerve (see Fig. 14). Similar to a CI, an ABI consists of an external ear-level worn device and an internal receiver-

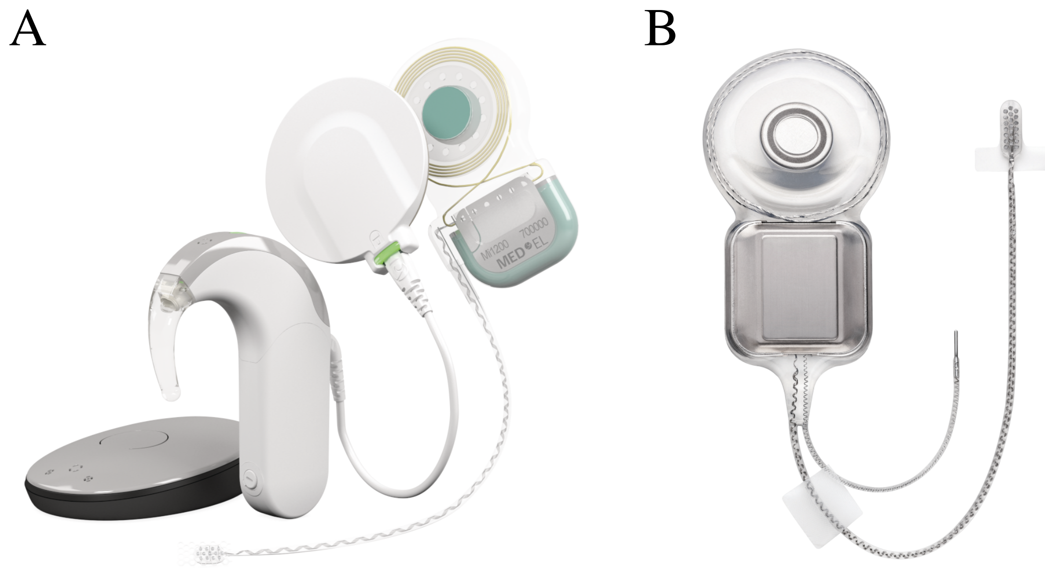

FIGURE 13 – The auditory brain stem implant. A : internal and external components of the ABI made by MED-EL (image courtesy of MED-EL). B : internal components of the ABI made by Cochlear (image courtesy of Cochlear Americas).

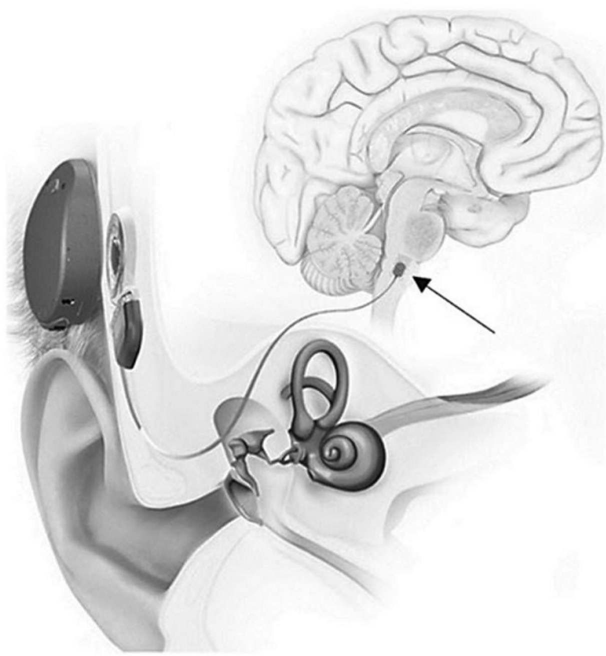

FIGURE 14 – The auditory brainstem implant, with the electrode array placed on the cochlear nucleus (black arrow). Source : Original drawing from Dhanasingh et al. [47] (image courtesy of MED-EL).

stimulator implant. ABI external part is identical to the external CI and includes the same elements : battery, microphone, speech processor, and transmitter. ABI internal part consists of a surgically placed receiver-stimulator placed against the skull and above the craniotomy defect, a multichannel electrode array placed through the lateral recess of the fourth ventricle, and a ground electrode inserted against the calvarium and under the temporalis muscle. Unfortunately, the overall outcomes of the ABI are inferior to the CI. Most users will see benefits in terms of sound awareness but will rarely see great improvements in speech discrimination [49].

4.5 Other interventions

In the event that a patient cannot benefit from a hearing device (medical contraindication, financial limitations), has limited benefits with the hearing device, or does not want to utilize a hearing device (limited perceived benefits, stigmatization), other strategies can be used to improve communication. Certain communication strategies can be employed such as favoring face-to-face conversation, avoiding noisy environments, and repeating and rephrasing. Lip-reading is often done unconsciously but can also be practiced. This provides complementary information to what is heard. Another device-oriented solution is the wireless remote microphone. These devices transmit sounds from a desired source wirelessly to the ears of the user. Wireless remote microphones are particularly effective for improving speech understanding in noise [50]. A more modern solution to communication problems comes under the form of live transcription applications available on the cellphone. These applications essentially provide real-time closed captions of the conversation unfolding around the cellphone. This method provides written information that complements auditory information [51].

5 Conclusions

Hearing loss is one of the most common chronic conditions which can be particularly critical at both ends of the age spectrum. We proposed this overview of the evaluation and interventions/solutions for hearing loss, in an attempt to demystify the audiological world to the Canadian acoustical community. Collaboration between the multiple professionals involved in hearing health and hearing research is vital for the development of better evaluations and interventions.

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