AUDITORY FUNCTIONS OF THE PERIPHERAL HEARING SYSTEM AND THE COMMON CONDITIONS AFFECTING SOUND CONDUCTION

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Résumé

Le système auditif périphérique combine une transmission mécanique et électrique via les différentes structures de l'oreille externe, moyenne et interne. L'Organisation Mondiale de la Santé estime que 6,1% de la population présente une atteinte auditive bilatérale. Cet article résume le rôle des différentes composantes du système auditif périphérique et présente les causes les plus communes de perte auditive touchant la transmission mécanique des sons chez l'humain. Quelques causes fréquentes d'atteinte auditive touchant la transmission neurale sont aussi abordées. Plus précisément, l'occlusion du conduit auditif externe, l'otite externe (dans le cas où l'enflure est si importante qu'elle bloque le conduit auditif), la dysfonction tubaire (trompe d'Eustache), l'otite moyenne séreuse et aiguë, la perforation tympanique, le cholestéatome, la discontinuité ossiculaire, l'otosclérose, la maladie de Ménière, la presbyacousie et la perte auditive causée par l'exposition au bruit sont brièvement abordés dans cet article afin de sensibiliser l'ensemble de la communauté de l'Acoustique Canadienne à ces problèmes et pathologies.

Mots clés : système auditif, anatomie, physiologie, conduction mécanique, pathologies

Abstract

The peripheral hearing system combines mechanical and electrical transmission through the different structures of the outer, middle and inner ear. The World Health Organization estimates a prevalence of 6.1% of the world population living with a bilateral disabling hearing loss. Here, the roles of the different peripheral hearing system structures are reviewed and the most common causes of hearing loss related to mechanical transmission in the human ear are presented. Some common causes of sensorineural hearing loss are also discussed. More precisely, ear canal blockage, external ear infection (when the ear canal is blocked due to severe swelling), Eustachian tube dysfunction, serous and acute otitis, tympanic membrane perforation, cholesteatoma, ossicular chain discontinuity, otosclerosis, Meniere's disease, presbycusis and noise-induced hearing loss are briefly presented in this paper in an attempt to highlight these problems and pathologies to the Canadian acoustical community.

Keywords: hearing system**,** anatomy, physiology, mechanical transmission, pathologies

1 Introduction

The prevalence of hearing loss worldwide is difficult to estimate. Studies on the subject use different measuring tools, ranging from subjective questionnaires (completed by the subject or by someone in the household) to clinical evaluations in a sound-attenuating booth. Moreover, the criteria to conclude the occurrence of hearing loss vary in terms of intensity and frequencies.

For example, Goman and Lin (2016) determined the prevalence of hearing loss in the US by using an average criterion to sort the impairments by severity. This average criterion was computed using pure-tone thresholds estimated at 500, 1000, 2000 and 4000 Hz in a sound-attenuating booth. If the average criterion was superior to 25 dB HL, individuals were considered to present a hearing loss [1]. Using such methodology, Goman and Lin concluded that the prevalence of hearing loss in the US for 12 year olds and older is estimated at 23% [1]. Their study also reported that mild hearing loss was more frequent (estimate of 25.4 million cases), except for individuals aged 80 years or older for whom a moderate hearing loss (mean threshold between 41 and 60 dB HL) was more frequent than a mild hearing loss (mean threshold 26 to 40 dB HL). Using the same severity criterion and average frequency method, Feder et al. (2015) estimated that 19.2% of Canadians aged between 20 and 79 years old presented a hearing loss and that 12% of Canadian adults suffer from a mild hearing loss [2]. Their study also reported that the prevalence was more important in younger subjects (less than 10%) and reached 50 to 65% in 70-79 years old [2].

As for children and teenagers, Feder et al. (2017) studied 2434 individuals aged between 6 and 19 years old with valid audiometric results. In this study, they defined the hearing loss as a pure-tone average (for each ear separately) of more than 20 dB HL in individuals aged between 6 and 18 years old and of more than 25 dB for individuals aged 19 years old, using different pure-tone average frequencies. A global average was computed using thresholds estimated at 500,

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1000, 2000 and 4000 Hz. A high frequency average was computed using thresholds estimated at 3000, 4000, 6000 and 8000 Hz. A low frequency average was computed using thresholds estimated at 500, 1000 and 2000 Hz [3]. Hearing losses were found by using at least one of the pure-tone averages in 7.7% of the individuals aged between 6 and 19 years old, and 4.7% for the 4 frequencies pure-tone average [3]. The majority of these hearing losses were unilateral and the most frequent severity was mild. The authors report a possible underestimation of the true prevalence since many subjects were excluded because no audiometric valid results could be measured or because the ear canal showed excessive earwax or pus [3]. In newborns, the prevalence of permanent hearing loss is estimated to 133 per 100 000 live births [4].

The World Health Organization estimates that 6.1% of the world population has a disabling hearing loss (defined as a 40 dB and 30 dB hearing loss in the better ear in 15 years or older and 14 years and under respectively) [5].This paper aims to report the more frequent hearing loss etiologies affecting the mechanical transmission of the sounds in the peripheral hearing system, in an attempt to sensitize the Canadian acoustical community to the various origins of such hearing impairments that are widely reported in the literature. The first part of the article describes the normal function of the peripherical hearing system and the common hearing dysfunctions are presented in the second portion. Pathologies affecting the sound transmission, rather than the mechanical transmission part, were excluded in this paper, except for the most prevalent ones: NIHL and presbycusis. Rare afflictions and malformations will also be excluded.

2 Hearing function

As can be seen in figure 1, from the time a sound wave enters the outer ear to the time it reaches the inner ear and is processed by central auditory pathways, many mechanical and electrical functions (neural transmission) are required.

2.1 External ear

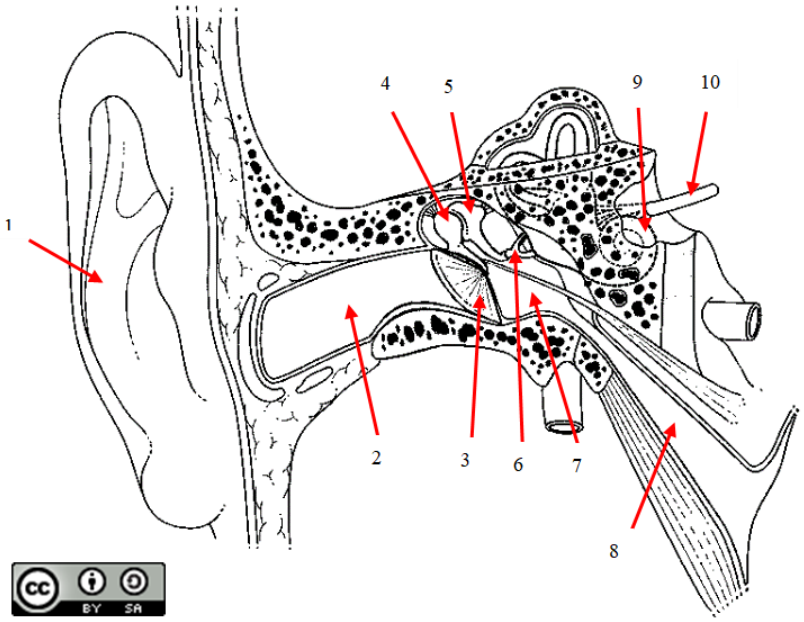
When a sound reaches a normally functioning ear, it first meets the external ear. The external ear is comprised of the pinna and the external ear canal (figure 1) [6]. The pinna has a peculiar shape that allows both sound localization in the vertical plane and sound amplification: the mid frequencies from 2000 and 7000 Hz are slightly amplified [6, 7]. The sound then travels in the ear canal, another source of sound amplification composed of an external cartilaginous portion and an inner bony portion [8].

2.2 Middle ear

At the end of the external auditory canal, the sound reaches a thin translucid membrane called the tympanic membrane, or eardrum (figure 1, figure 4a) [7, 8]. The tympanic membrane is the first structure of the middle ear and is composed of three layers. The condensations and rarefactions of the sound make the tympanic membrane vibrate, and the three middle ear ossicles (called malleus, incus and stapes) are put into motion as well, transforming the acoustical energy into mechanical energy [9]. The three ossicles are linked with one another and the ossicle chain is suspended in the middle ear cavity by ligaments [10]. The last ossicle of the chain, the stapes, transfers the movement to the oval window, a small opening in the inner ear which is covered with a flexible membrane.

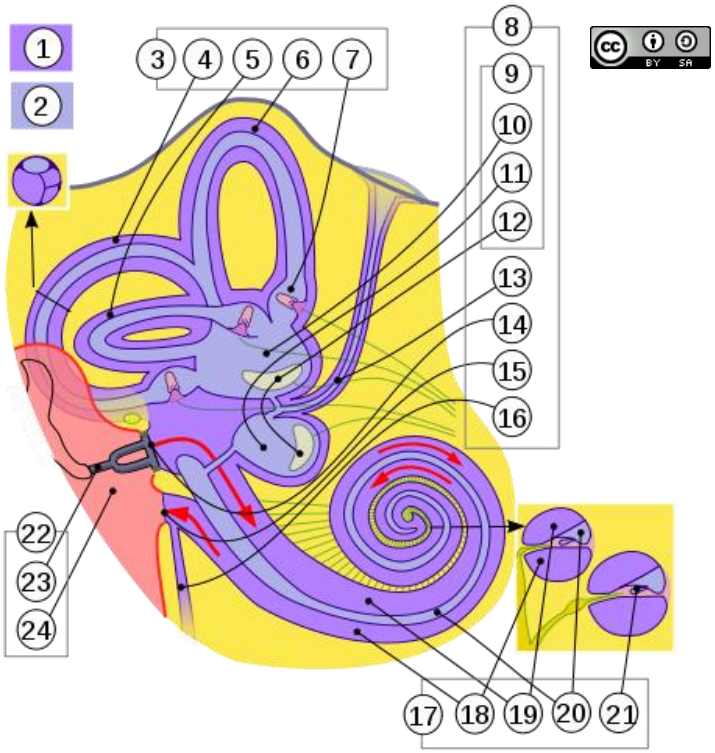
During the transfer of the movement to the oval window, the ossicles also amplify the movement, mainly around 1-2 kHz [9, 11, 12]. When the stapes presses on the oval windows, it causes the liquid of the inner ear (the perilymph) to move.

During this change in medium, the middle ear uses two principles to match the impedance: firstly the tympanic membrane has a much larger surface than the oval window, and secondly the lever action of the incus and malleus [9].



1- Pinna
2- External auditory canal
3- Tympanic membrane (eardrum)
4- Malleus
5- Incus
6- Stapes
7- Tympanic cavity (middle ear)
8- Eustachian tube
9- Cochlea (inner ear)
10- Auditory nerve

Figure 1: Peripheral hearing system. Modified by adding arrows and numbers from the original drawing by Didier Descouens, licensed under CC BY-SA.



1 Perilymph
2 Endolymph
3 Semicircular canals
4 Posterior semicircular canal
5 Horizontal semicircular canal
6 Superior semicircular canal
7 Ampula
8 Vesibule
9 Otolith organs
10 Utricule
11 Saccule
12 Vestibular maculae
13 Endolymphatic duct
14 Oval window
15 Round window
16 Perilymphatic duct
17 Cochlea
18 Scala tympani
19 Scala vestibuli
20 Cochlear duct
21 Oran of Corti
22 Middle ear
23 Stapes
24 Tympanic cavity (middle ear)

Figure 2: Inner ear. Original drawing by Jmarchn, licensed under CC BY-SA.

Middle ear pressure

The equalization of the air pressure in the middle ear is ensured by the Eustachian tube [13]. This structure is a small tube (24 mm long, with a bony and cartilaginous portion) between the nasopharynx and the middle ear which has a protective role: protection against self-generated body sounds such as voice, breathing and heartbeat (also called autophony), protection against infections and protection against inner body sudden pressure change (like when coughing) [14-16]. Most of the time, the Eustachian tube is closed. It normally opens when swallowing or following jaw movements or yawning [15]. These occasional openings allow one to equilibrate pressure outside and inside the middle ear, and to bring in fresh air and oxygenate the walls of the middle ear, recovered with mucosal tissue [17].

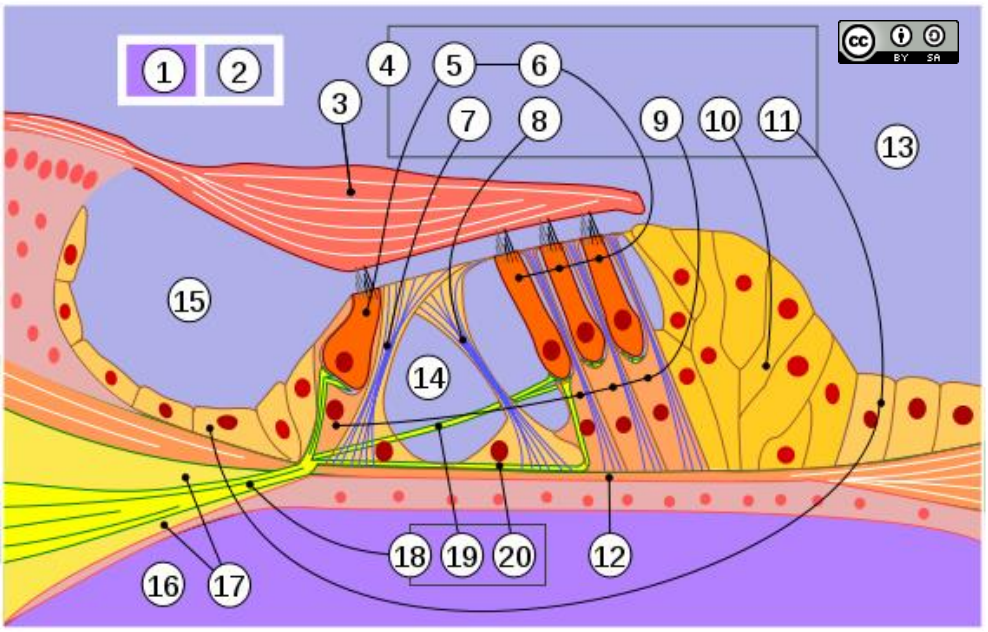
Reduction of loud noises

In the middle ear, the stapedial muscle (not shown on Figure 1) contracts in the presence of a loud noise (generally 85 to 100 dB SPL for pure tone stimuli), thus stiffening the ossicular chain and reducing the intensity of the noise reaching the cochlea [9, 18]. In order for this reflex to occur, other structures need to be functioning well, such as the cochlea, auditory nerve, facial nerve and brainstem structures [18]. However, this reflex has a few limits: it does not protect for long noise exposures, has a small activation delay which does not protect against impact noises and mostly protects against low frequency noises [7, 9].

2.3 Inner ear

The inner ear, also called the cochlea, is filled with two different kinds of fluids: perilymph and endolymph. These liquids travel in three canals all along the two and a half spires of the cochlea: the endolymph in the cochlear duct and perilymph in the scala vestibuli and scala tympani (Figure 2) [7]. The scala vestibuli and the scala tympani are connected at the apex of the cochlea, a point named helicotrema [7]. Along with the oval window, the round window allows perilymph to move when the stapes moves. The movement of the stapes and the presence of the two windows allow a movement of the fluids, resulting in a vibration of the basilar membrane, on which the Organ of Corti rests [7, 9]. Even if the organ of Corti is similar along the two and a half turns of the cochlea, the properties of the basilar membrane differ. It is narrow and stiff at the base. At the apex the basilar membrane is wider, more flexible and has more mass. These properties allow a frequency distribution (low-frequencies being perceived with the stimulation of the apex and high-frequencies being perceived with a base stimulation of the cochlea) [7, 9].

In the organ of Corti, there are hair cells and supporting cells: hearing sensitivity depends on the good function of those cells, and more specifically on inner and outer hair cells [7]. The hair cells are in contact with two different fluids with different ionic composition: perilymph and endolymph. These fluids have a 80 mV potential difference (endocochlear potential) [7].



1 Perilymph
2 Endolymph
3 Tectorial membrane
4 Organ of Corti
5 Inner hair cells
6 Outer hair cells
7 Inner pillar
8 Outer pillar
9 Deiter's cells
10 Hensen's cells
11 Claudius cells
12 Basilar membrane
13 Cochlear duct
14 Inner tunnel (Corti)
15 Internal spiral tunnel
16 Scala tympani
17 Bony (osseous) sprial lamina
18 Auditory (cochlear) nerve
19 Efferent fibers
20 Afferent fibers

Figure 3: Organ of Corti. Original drawing by Jmarchn, licensed under CC BY-SA.

The tip of the hair cells bathes in endolymph and the base bathes in perilymph, allowing a passive entrance of potassium ions (K^+) as well as a passive exit of the same ions, following the concentration gradient [7]. This way, rapid successive stimulations are possible in the hair cells [7]. The stria vascularis needs energy to create the endolymph, which is rich in K^+, but the presence of the different fluids allows hair cells to save ATP [7].

Inner hair cells have stereocilia at their apex and, when there is a vibration, the stereocilia move in the stria vascularis' direction (driven by the movements of the basilar and tectorial membranes, see Figure 3) and pull open potassium channels, allowing for the cell to depolarize [7]. There is a release of neurotransmitters in the synaptic space, thus transforming a mechanical energy into an electrical energy: the sound travels through many auditory relays up to the brainstem or the brain [7]. Those higher functions are beyond the scope of this paper.

In the inner ear, outer hair cells also depolarize by the stereocilia movement. Unlike the inner hair cells, the outer hair cells' electric message feeds energy back to the cochlear partition [7]. The outer hair cells play an amplification role for faint to moderate sounds. These cells contract (by electro-mechano transduction), allowing less powerful soundwaves to stimulate the inner hair cells more easily [7]. Lost external hair cells do not regenerate in humans, nor for all mammals [7].

3 Conductive & mixed hearing dysfunctions

There is a large number of hearing loss etiologies. Some only affect the mechanical transmission of the sound to the cochlear hair cells: this origin of hearing loss is called "conductive hearing loss". In this particular case, patients have good hearing thresholds when measured with bone conduction (stimuli are presented with a bone vibrator placed on the mastoid), but not when measured with air conduction (stimuli are presented with earphones) [7]. Other causes lead to sensorineural hearing losses, a hearing loss affecting the cochlear or auditory neural pathways; in these cases, bone and air-conduction are similarly affected [7]. Finally, a mixed hearing loss is when there is both a bone-conduction hearing loss and an air-bone gap in the hearing thresholds [7].

The focus of the present paper is on the more prevalent causes of hearing loss affecting the mechanical transmission of sounds, from the external ear to the cochlea. The choice of presented conditions was inspired by Isaacson & Vera (2003) [19]. Some pathologies will change the structure's mass or stiffness and may therefore alternate the perception of sounds in different ways by modifying the transfer function of the middle ear.

By definition, the resonant frequency of a vibrating object depends on mass and stiffness: mass is an obstacle to high-frequency sound transmission and stiffness is obstacle to low-frequency [9]. Therefore, if the middle ear is considered as a mass-spring system, the stiffness resides in the tympanic membrane (it's elasticity), the ligaments of the ossicles and the pressure changes in the middle ear during the tympanic membrane vibration [9]. For example, the tympanic membrane may become stiffer when the air pressure in the middle ear is very different than the ambient pressure [9]. Changes in stiffness may also come from a modification in the tendons and ligaments linked to the ossicular chain, or by changes in the ossicular chain itself.

The mass of the mass-spring system of the middle ear resides mainly in the ossicles and the tympanic membrane modifications. Changes in the ossicles' mass may affect transmission of high frequencies [9]. Additionally, changes in the mass of the tympanic membrane may also change the middle ear function. Frictions may also occur in the middle ear due to the viscosity of the mucous membranes or the presence of narrow passages [20].

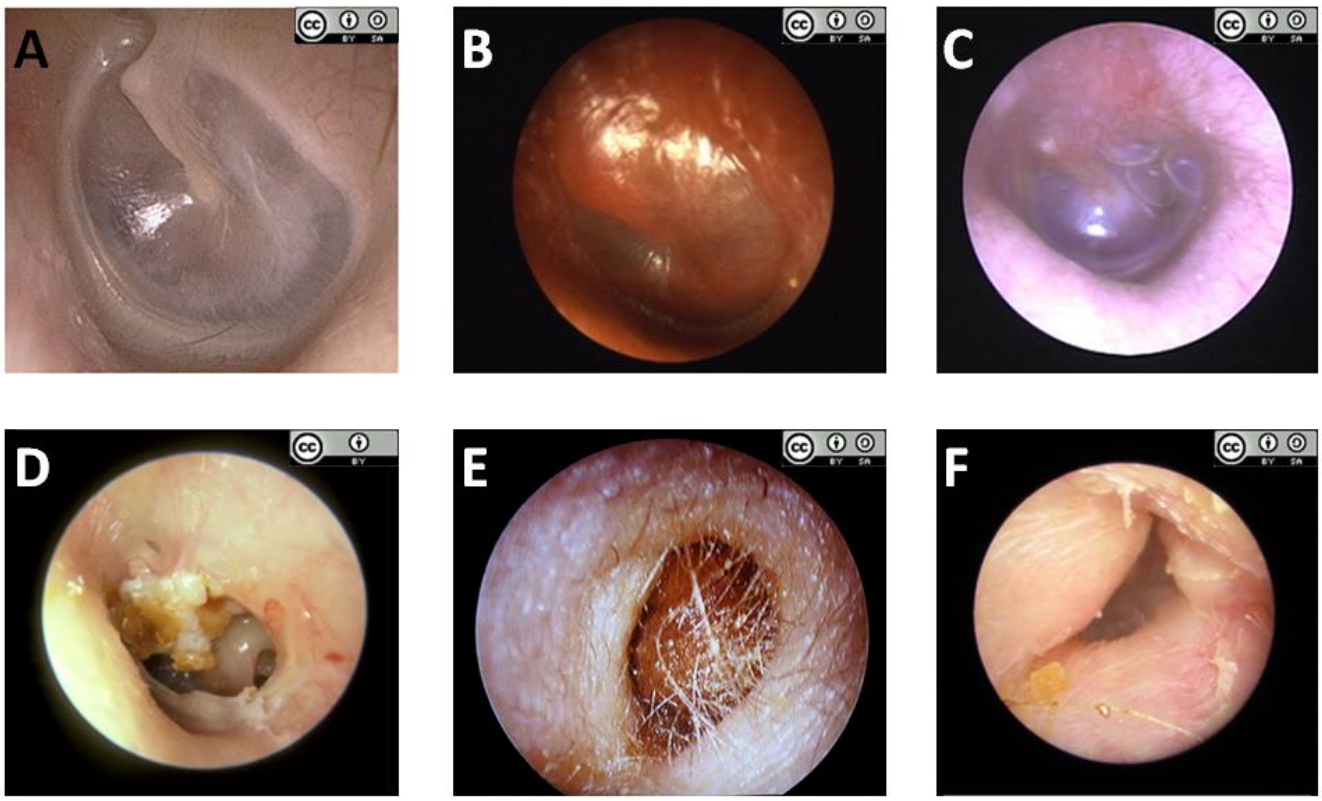


Figure 4: External ear and middle ear

A- Normal tympanic membrane. Original picture by B. Welleschik, licensed under CC BY-SA.
B- Acute otitis media. Original picture by B. Welleschik, licenced under CC BY-SA.
C- Serous otitis media. Original picture by B. Welleschik, licensed under CC BY-SA.
D- Tympanic membrane perforation with cholesteatoma. Original picture by Michael Hawke, licensed under CC BY-SA.
E- Cerumen (earwax) blockage. Original picture by Didier Descouens, licensed under CC BY.
F- Exostosis. Original picture by B. Welleschik, licensed under CC BY-SA.

Additional hearing dysfunctions may modify the natural external ear transfer function, such as when the ear canal is blocked or when the tympanic membrane is perforated [6].

3.1 Ear canal blockage

There are many causes for a temporary or permanent ear canal partial or total blockage: cerumen (earwax) accumulation, an external ear malformation, a foreign body (e.g. necklace pearl, blueberry), etc. [8]. A partially occluded ear will generally have a high frequency hearing loss, while a total occlusion will also affect middle and low frequencies [21]. Such hearing loss is conductive.

Cerumen is produced in the outer third of the ear canal, where hair and ceruminous glands are found. It is constituted of ceruminous glands secretions and oily sebaceous secretions [10, 22]. The cerumen forms a protective coating with antibacterial properties [23]. The ear is generally self-cleaning; in some cases, a partial or total obstruction of the ear canal is noted (Figure 4e) [22]. This situation is temporary if the ear canal is cleaned.

Another slightly less frequent cause of partial blockage is exostosis (figure 4f). The exostosis is a benign bony outgrowth, often noted in patients with a history of cold water swimming [8]. This problem is generally bilateral [24]. While often asymptomatic, in rare cases the ear canal may be completely blocked or cause repeated infections or cerumen impaction [8, 24].

3.2 External ear infection

Also known as swimmer's ear, the External ear infection or otitis externa is an infection of the external ear (often of the ear canal) [25]. The cerumen coating generally creates an acidic environment in the ear canal, but frequent swimming (bath, shower) may rinse it off and make the ear more vulnerable to ear canal infection, especially when contaminated water stays in the ear [23, 25].

Early on, the patient notices rather sudden itching and swelling of the ear canal, as well as pain [24]. Moreover, movements such as mouth opening or touching the pinna may be painful [24]. In most cases, there is also generally an ear discharge that may be thick or serous.

If there is an important accumulation of ear discharge and debris or if the tympanic membrane is thicker, a conductive hearing loss might be noted [24]. This infection is generally treated with eardrops since the more common cause is bacterial [24]. A fungus or a virus may also be the cause. This condition may be prevented by the use of ear drops changing the acidity in the ear canal [25].

3.3 Eustachian tube dysfunction

While an Eustachian tube may have a closing dysfunction, the most prevalent trouble is an obstruction or occlusion of the Eustachian tube. This problem is often temporary, like in the case of nasal congestion or in young children. The obstruction of the Eustachian tube may result in a serous otitis, as will be discussed later.

In an occlusive dysfunction, the forces that maintain the Eustachian closed (pressure from surrounding structures, cartilages elasticity, surface tension offered by mucous in the Eustachian tube, some muscle contraction and relaxation) and those which allow opening (mainly muscle contraction) are unbalanced [16]. The cause may be in the Eustachian tube itself, like in an upper respiratory tract infection with acute inflammation in the Eustachian tube (edema, mucus change), or located nearby (hypertrophy of adenoids blocking the nasopharyngeal opening) [26]. Allergic rhinitis may also cause a nasal inflammation and an edema in the nasopharynx [26]. Another frequent and permanent cause is malformations affecting the muscles in charge of the Eustachian tube opening or the tube itself. For example, cleft palate is a malformation that often affects the Eustachian tube function [14].

In the presence of Eustachian tube dysfunction, a negative pressure in the middle ear is frequent and may cause a rise in the middle ear stiffness and a conductive hearing loss, mainly in low frequencies [9].

Vila et al. (2017) estimated the US number of consultations to more than 2 million per year in patients under 20 years old, and a similar number of consultations in patients of 20 years old and older, for Eustachian tube dysfunction and related complications (otitis media with effusion and tympanic membrane retraction) [27].

3.4 Acute otitis media

When there is an acute inflammation of the middle ear, it is called an acute otitis media (figure 4b) [28]. The symptoms generally appear suddenly: ear pain, impression of ear fullness, fever, hearing loss and, in some cases, ear discharge (if the tympanic membrane is perforated) [15].

Acute otitis generally causes a temporary conductive hearing loss present at all frequencies and is more prevalent in children [15, 29].

In most cases, the infection is viral and resorbs by itself in less than 48 hours [29]. When the condition persists or in the presence of a perforated tympanic membrane, antibiotics are generally prescribed. An untreated bacterial infection may have serious complications such as mastoiditis and meningitis.

By their first birthday, 62% of children have had at least one acute otitis media episode and 83% before their third birthday, with 46% having at least three episodes [30].

3.5 Serous otitis media

If the Eustachian tube is occluded or is not able to open for a prolonged period, the middle ear mucosal walls use all the oxygen available and a negative pressure builds up in the middle ear, causing a tympanic membrane retraction in the middle ear direction [17]. Once this pressure is important enough, the body mucosal walls of the middle ear transudate a serous liquid in the middle ear [13, 27]. The presence of this serous liquid, in the absence of infection, is called serous otitis media (figure 4c) [31]. This condition is very common in children in whom the Eustachian tube is shorter and closer to a horizontal orientation [14].

There are causes for serous otitis other than Eustachian tube dysfunction. For example, serous otitis may follow an acute otitis, if the infection was treated but the middle ear remains filled with fluid [14, 28]. Following an acute otitis media diagnosis, a middle ear effusion is often present for 1 to 3 months (45% and 10% of the cases respectively) [30]. There is also a possibility that the effusion may have been present before the infection causing the acute otitis media [14]. Malformations of the skull base, ear or Eustachian tube (including the muscles that allow it's opening) may also cause frequent serous otitis [14].

Once the middle ear fills with fluid, the mass and the stiffness of the tympano-ossicular system changes, and a conductive hearing loss is typically present at all tested frequencies [9].

This condition in children must not be taken lightly because it can delay language acquisition if present for a prolonged period [32]. In many cases, the serous otitis will resorb by itself [29]. However, the presence of residual uninfected fluid in the middle ear is a risk factor for another episode of acute otitis [14, 28]. Also, if the serous fluid persists many months, the Ear, Nose and Throat doctor (ENT) will often place ventilation tubes to restore the hearing sensitivity [13]. It must be noted that fluid that persists in the middle ear for years may cause an ossicular erosion over time [33].

The serous otitis media has a prevalence in adults of approximately 0.6% and a prevalence in children of 20% (for 2-year-olds), with more than 90% of the children having a first occurrence of serous otitis media before the age of two [31, 34].

3.6 Barotrauma

A barotrauma is a trauma caused by extreme air pressure changes and affects particularly enclosed cavities. The Eustachian tube has elastic properties, but may block in the presence of a sudden pressure change [15]. If the pressure difference between the middle ear and the environment becomes severe, particularly if the extra-tympanic pressure increases rapidly, there is a risk for barotrauma [15]. The trauma may cause middle ear transudation in the lighter cases, with an obstruction persisting up to several weeks. In more severe cases, hemorrhages in the middle ear, ossicular dislocation, tympanic perforation or perilymphatic fistula (opening in the round or oval window resulting in perilymph loss from the inner ear, accompanied by a hearing loss and vertigo) may occur [24, 35].

The impact of barotrauma on the hearing thresholds will vary according to the type of consequences: middle ear fluid, tympanic membrane perforation, etc.

The time of recovery and the number of medical interventions vary as well.

3.7 Tympanic membrane perforation

The main causes of a tympanic membrane perforation include the use of an ear swab or the insertion of a foreign body in the ear, a barotrauma, an acute otitis or a head trauma [8]. The presence of tympanic membrane perforation consecutive to chronic suppurative otitis media is estimated to 1.78% [36].

The impact on the hearing sensitivity varies greatly according to the perforation localization and size [8]. For example, ventilation tubes are known to cause little to no hearing changes. In most cases however, a conductive hearing loss is measured in the presence of a tympanic membrane perforation [37].

The recovery is often spontaneous but depends on the size of the perforation, the cause and the presence of constant ear discharge [24, 36].

3.8 Cholesteatoma

The cholesteatoma is a non-cancerous tumor which develop in the middle ear (figure 4d). It is often a consequence of otitis media or tympanic perforation [36]. The cholesteatoma is formed of accumulated epithelial debris and keratin in the middle ear (often beginning on the tympanic membrane) and can destroy surrounding structures, even bone, by erosion and compression [15, 36]. The presence of this mass in the middle ear leads to an augmentation of the impedance. It causes a hearing loss and it is generally accompanied by a strong scented ear discharge [38].

The treatment for cholesteatoma includes ear surgery to remove all of the tumor and to prevent recidivism [15]. If the mass is large, a mastoidectomy – removal of bone in the skull, behind the ear – can be necessary. Since the surgery often leads to a predominantly conductive hearing loss (for example in the case of ossicle removal), a reconstructive surgery may be offered to regain some hearing sensitivity.

The prevalence of cholesteatoma is estimated to be 0.34% in the population of 4 year olds and older [36]. The retraction pocket in the tympanic membrane, a condition that may follow tympanic membrane perforation and is a risk condition for cholesteatoma, has an estimated prevalence of 1.21% in the population of 4 years old and older [36].

3.9 Ossicular chain discontinuity

The three middle ear ossicles are normally linked one to another to transmit the sound vibration from air (outer ear) to a liquid medium (inner ear). However, sometimes two ossicles may lose their link from one another, or the malleus with the tympanic membrane, or even the stapes with the oval window [15, 39]. One of the possible etiologies is the cholesteatoma, especially after a surgical tumor removal. Other causes include congenital malformations of the ossicles (often with atresia – a permanent occlusion of the ear canal), head trauma, otitis media (with important erosion), barotrauma or penetrating object in the middle ear. Depending on the cause, sometimes a fibrous joint may remain between the ossicles even if they are detached [39].

In the case of a complete discontinuity, a conductive hearing loss is noted on all frequencies [39]. Partial discontinuity is known to cause a hearing loss more important on the high frequencies [39].

3.10 Otosclerosis

Otosclerosis is a disease of the temporal bone that leads to a progressive hearing loss [40, 41]. It consists of bone resorption, followed by new bone formation, often causing a partial or total mechanical blockage of the stapes footplate movement on the oval window (the mobility slowly decreases with time) [42, 43]. Mechanical blockage also may touch the other middle ear ossicles. The presence of otosclerosis stiffens the ossicular chain, leading to a conductive hearing loss more important in the low frequencies [9, 37].

The prevalence of this disease is of 0.3 to 0.4% in Caucasians [42]. It is one of the more frequent etiology for hearing loss apparition in adults, and the more common conductive hearing loss in Caucasian adults [42, 43].

Women are twice more often affected than man by otosclerosis, and the evolution of the pathology is faster in women due to their hormonal changes [44]. Otosclerosis progress faster with endocrine activity, for example during puberty, pregnancy and menopause. In some cases, the cause of otosclerosis is genetic. An autosomal dominant transmission with incomplete penetrance have been identified in some cases [42, 43].

Many patients suffering from otosclerosis undergo a stapes replacement surgery to regain some hearing sensitivity [40]. The extension of the pathology to the inner ear leads to a sensorineural component in the hearing loss, in addition to sound transmission alterations [41, 42].

4 Sensorineural hearing dysfunctions

4.1 Meniere's disease

Meniere's disease is characterized by an excessive presence of endolymph, or hydrops of the inner ear [45, 46]. A hydrops is an excessive accumulation of fluid. There are several hypotheses about the apparition of the hydrops: either too much endolymph produced by the stria vascularis or not enough (absorbed by the endolymphatic sac), or there is a circulatory problem of the endolymph [47]. Meniere's diseases is characterized by three main symptoms: a fluctuating sensorineural hearing loss (generally in low frequencies), intermittent tinnitus (whistling, buzzing, but most frequently a humming in Meniere's disease) and intermittent episodes of vertigo [48]. Symptoms manifest in the form of crisis that may last from a few minutes to many days, and are often accompanied by an aural pressure [47, 48].

Since some patients present an incomplete expected symptoms portrait, the prevalence of Meniere's disease is not well known. Estimations vary between 8 and 218 cases per 100 000 individuals [49, 50]. In the US, the estimation is 73

per 100 000 individuals and the prevalence is lower for men than women [50].

Therapeutic treatments include sodium intake restriction and medication (betahistine, intratympanic gentamicin, steroids) [46, 51].

4.2 Presbycusis

It is well documented that hearing loss is more frequent in older individuals. Ageing will impact the neural transmission of sounds by auditory neurons loss, vascular changes in the cochlea and central hearing pathways' function [7, 52]. However, it also impacts the mechanical transmission of sounds.

In the external ear, the cerumen becomes drier, harder and impactions are more frequent [52]. In the middle ear, the tympanic membrane may become stiffer, thinner and less vascularized [52]. The joints between the ossicles may show calcification and arthritic changes. Middle ear muscles and ligaments show atrophy and degeneration [52]. However, despite all these changes the impact on ageing in the audiometric results are typically of sensorineural predominance [7, 52, 53].

In the inner ear, ageing will lead to hair cells loss, support cells loss, basilar membrane rigidity and calcification of some structures in the cochlea [52]. The organ of Corti is the most susceptible structure to age changes. The hearing loss is generally sloping in the high frequencies [21, 52].

Homans et al. (2017) found a prevalence of 33% of men and 29% of women with a hearing loss ≥ 35 dB HL (pure tone average: 500, 1000, 2000 and 4000 Hz) [53]. Lin et al. (2011) estimated that 63.1% of the US population of age 70 and older present a hearing loss (pure tone average > 25 dB HL in the better ear) [54].

4.3 Noise-induced hearing loss (NIHL)

Noise exposure is known to cause a hearing loss, affecting importantly the outer hair cells function, and eventually the inner hair cell function [55]. At first, a noise-exposed individual may experience a temporary threshold shift (TTS), where symptoms such as hearing loss, tinnitus and impression of ear fullness last from a few hours to a few days [55]. These effects were believed to be temporary until recently. Animal studies however suggest that there may be permanent effects on the neural processes of supra-threshold signals [56]. In the case of exposure to very intense noise (e.g. the noise of a gunshot nearby), the hearing damage may be immediate and even affecting the middle ear - this situation is known as an acoustic trauma [55]. The acoustic trauma is less frequent than the permanent threshold shift (PTS) observed in most noise-exposed workers: 10% of the world's population would be exposed to noise levels associated to a noise-induced hearing loss risk [57]. Typically, the PTS occurs progressively, is permanent and impacts more importantly the hearing thresholds at and near 4000 Hz [55]. The hearing loss is sensorineural and affect both ears in most cases [57]. Sensorineural hearing loss or progression may be prevented in reducing the noise exposition time and the noise intensity [55].

5 Conclusion

This overview of the peripheral hearing function and dysfunctions focuses on the most prevalent etiologies affecting mechanical transmission. Such mechanical changes may touch the ear canal, the tympanic membrane, the middle ear ossicles and/or the cochlea. Presbycusis and noise-induced hearing loss are also discussed because of their high prevalence. Other hearing loss causes that are less prevalent or related to the auditory nerve or higher auditory pathways are not included in this paper.

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