6 Understanding the Aeration Avenues of the Middle Ear with the Aid of the Endoscope

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Introduction

The study of the ventilation pathways of the middle ear, in particular of the epitympanic diaphragm, has long been one of the main interests of otologists, as, in the early 40s, it was first understood how its structure can affect the pathology of the middle ear. The inter-attico-tympanic diaphragm, later renamed the epitympanic diaphragm, was first identified in a histological study in newborns by Chatellier and Lemoine [1]. They described a set of membranous structures that, along with the incus and the malleus, shaped the floor of the epitympanum. Their study was based on the previous histological description of the development of middle ear spaces in fetuses at different stages of development by Hammar [2] and on anatomical treatises, like the one by Prussak [3]. Palva et al. further defined this structure through studies of microdissection of both healthy and diseased specimens, focusing on the ventilation pathways and their implication in the pathological processes of the middle ear. The epitympanic diaphragm divides the middle ear into two compartments: the epitympanum and the medial tympanum. It was Palva et al. who underlined the association between the blockage of the aeration pathways and the lesions of the upper compartment, due to dysventilation [4, 5].

During the last 10 years, the endoscope has gained increasing importance, first as a support to traditional microscopic surgery [6, 7] and then as a tool of its own, to carry out surgical procedures [8, 9].

The endoscopic approach to the middle ear has provided a unique way of evaluating its anatomy and disease, as it has made spaces, which cannot be reached with the use of traditional microscopes, easily accessible; in fact, 30°, 40°, and 75° endoscopes enable the surgeon to reach blind spaces in the middle ear, without disrupting any anatomical structures. While traditional approaches with a microscopic postauricular tympanomastoidectomy provide a limited access to the attic, especially to its anterior portion, the endoscope allows the surgeon to assess the integrity of the whole epitympanic diaphragm [10].

Since 2009, Marchioni et al. have published several studies on the endoscopic anatomy of the middle ear and have developed a nomenclature of the structures they identified. This classification has made it possible to standardize surgical approaches to lesions that require careful exploration of all the compartments of the middle ear, such as acquired or congenital cholesteatoma [11].

Ventilation Pathways

Anatomy of the Epitympanic Diaphragm (Fig. 6.1)

The epitympanic diaphragm, along with the malleus and the incus, forms the floor of the epitympanum, and it consists of the following folds (Fig. 6.2):

- Anterior, posterior, and lateral malleal ligament folds
- Posterior incudal ligament fold
- Tensor fold
- Lateral incudomalleal fold [12]

The anterior malleal ligament fold defines the anterior limit of Prussak space, as it extends from the anterior portion of the malleus neck to the anterior attic bony wall.

The posterior malleal ligament fold inserts into the posterior portion of the malleus neck and extends into the posterior tympanic spine. von Tröltsch's pouch is medially bounded by the posterior malleal ligament fold.

The lateral malleal ligament extends from the lateral wall of the malleus head to the medial wall of the scutum. It pos-

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M. V. Goycoolea et al. (eds.), *Textbook of Otitis Media*, https://doi.org/10.1007/978-3-031-40949-3_6

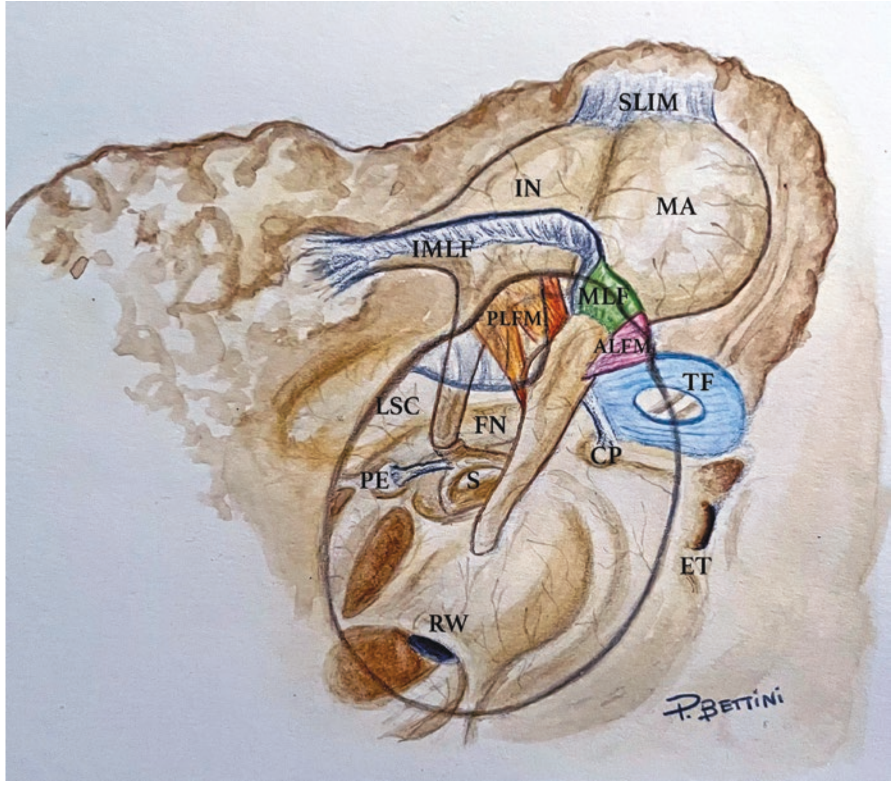

Fig. 6.1 The right ear. A drawing showing the epitympanic diaphragm seen in a caudal-to-cranial orientation and from a lateral to a medial one. The tensor fold is incomplete, showing a possible further ventilation pathway of the anterior epitympanum. *SLIM*, superior malleal ligament; *IN*, incus; *MA*, malleus; *IMLF*, lateral incudomalleal fold; *MLF*, lateral malleal fold; *PLFM*, posterior ligament of the malleus; *ALFM*, anterior malleal ligamental fold; *TF*, tensor fold; *LCS*, lateral semicircular canal; *FN*, facial nerve; *PE*, pyramidal eminence; *S*, stapes; *RW*, round window; *CP*, cochleariform process; *ET*, Eustachian tube

teriorly turns downward and merges with the anterior portion of the posterior incudal fold, whereas it anteriorly converges with the posterior portion of the anterior malleal fold.

The incudomalleal lateral fold is superior to the lateral malleal ligament fold, and it separates into two more compartments: the upper and lower lateral attic spaces. This fold has its insertion in the body of the incus and in the incudomalleal joint; it runs horizontally and extends to the medial wall of the scutum. Anteriorly, this structure bends and merges with the posterior margin of the lateral malleal ligament. During an endoscopic approach to the middle ear, the lateral incudomalleal fold must be disinserted from the body of the incus, to fully expose the upper lateral attic.

The anatomy of the tensor fold was defined owing to the introduction of the endoscope; in fact, its orientation and location make it undetectable through traditional retroarticular approaches. The tensor fold separates the supratubal recess in the protympanum from the anterior epitympanum, which is the space between the head of the malleus and the anterior bony wall of the scutum [12]. The tensor fold can appear as either complete or incomplete and can have three different orientations: vertical, horizontal, or oblique [13].

The relationship between the tensor fold and the cog (transverse crest) is important. The latter is a bony septum that cranially detaches from the tegmen tympani and vertically descends just anterior to the cochleariform process; it usually defines two compartments of the epitympanum: the anterior epitympanic space and the posterior epitympanic space. When the tensor fold has a vertical orientation, it is inserted into the cog, thus leading to a large supratubal recess. In this case, the cog does not separate the anterior from the posterior epitympanic space, rather, it represents the bony boundary between the supratubal recess and the epitympanic space. In the majority of cases, the tensor fold has an oblique orientation and attaches to the anterior tegmen tympani; therefore, the cog represents the division between the anterior and the posterior epitympanic spaces. When the tensor fold has a horizontal orientation and attaches to the tensor tympani semicanal, the supratubal recess is small or nonexistent. Thus, the anterior epitympanic space is large, and it is divided from the posterior epitympanic space by the transverse crest. As it is, the more vertical the tensor fold is, the wider the supratubal recess becomes.

Ventilation of the Upper Unit

It is known that three main factors influence the ventilation of the middle ear: the Eustachian tube function, the buffer mechanism provided by the mastoid, and the mucosal gas exchange.

The Eustachian tube plays a role in providing equalization of the pressure between the middle ear and the outside air. When the Eustachian tube fails, the pressure in the tympanic cavity becomes negative, causing a retraction of the tympanic membrane. The pressure in the middle ear is also related to the transmucosal gas exchange through the mas-

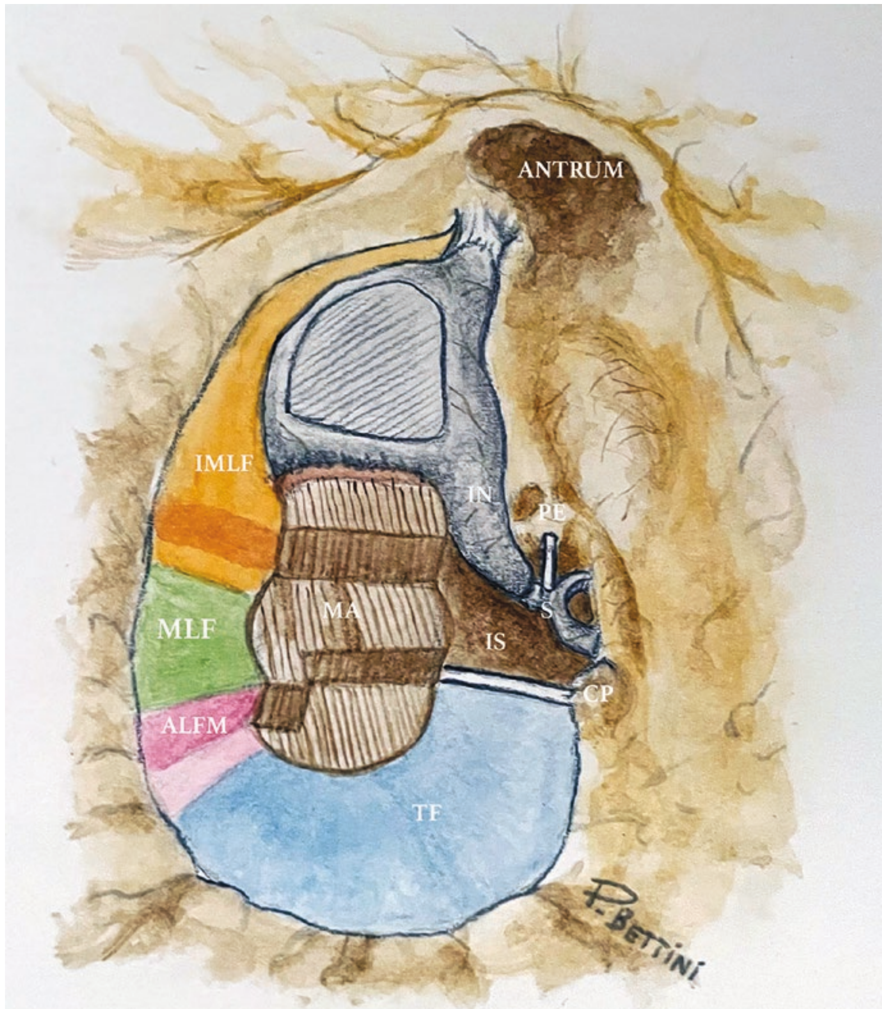

Fig. 6.2 The right ear. An epitympanic diaphragm from a superior view. The malleus is severed at different heights to show the attachment levels of the epitympanic diaphragm. The incus is sectioned at the level of its upper body. *IMLF*, lateral incudomalleal fold; *MLF*, lateral malleal fold; *ALFM*, anterior malleal ligamental fold; *TF*, tensor fold; *MA*, malleus; *IN*, incus; *IS*, tympanic isthmus; *S*, stapes; *PE*, pyramidal eminence; *CP*, cochleariform process

toid mucosa. These two systems achieve a balance if the ventilation pathways in the middle ear are open [14].

Two separate areas can be identified for what concerns the ventilation: an upper unit and a lower unit. The upper unit is made up of the superior lateral attic, along with the medial attic, and is connected to the mastoid. In case of a complete tensor fold, the upper unit is only aerated through two narrow spaces called the anterior and posterior isthmus, as the epitympanic diaphragm separates the epitympanum from the mesotympanum. The inferior lateral attic, which is divided from the superior one by the lateral incudomalleal fold, by contrast, is the only compartment of the epitympanum, which is aerated by the mesotympanic region [12].

The tympanic isthmus (Fig. 6.3) was first described by Chatellier and Lemoine; it was then outlined by Aimi as a narrow passage between the upper tubotympanic cavity and the atticomastoid space [15].

It is laterally bounded by the short process of the incus and the malleus head, anteriorly by the tensor tympani tendon and posteriorly by the posterior incudal ligament. The isthmus is divided into an anterior isthmus, which is the space defined by the incudostapedial joint and the tensor tympani tendon, and a variable posterior isthmus, which is just behind the incudostapedial joint.

The tensor fold plays a key role in the ventilation of the upper unit (Fig. 6.4), as it provides a separation between the anterior epitympanum and the protympanum. When the tensor fold is complete, the only ventilation pathway is through the tympanic isthmus, whereas in case of an incomplete tensor fold, additional ventilation is created between the protympanum and the anterior epitympanum.

Ventilation of the Lower Unit

The lower unit is represented by Prussak space; it is located below the epitympanic diaphragm, since its roof corresponds to the lateral malleal ligament fold. Prussak space is inferiorly defined by the short process of the malleus and medially by the malleus neck. The area is anteriorly bounded by the membranous anterior malleal ligamental fold, and its lateral wall is represented by Shrapnell's membrane, whereas the

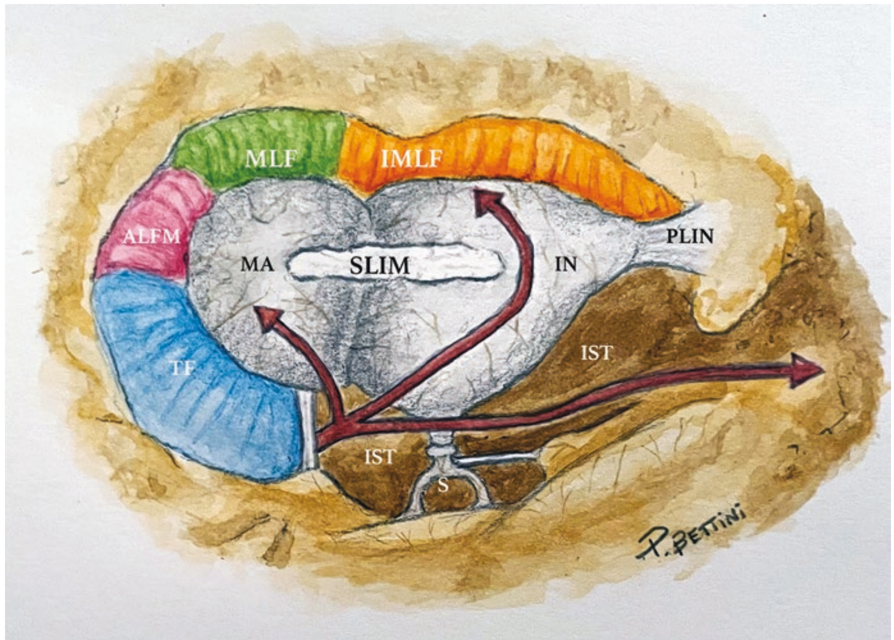

Fig. 6.3 The right ear. An epitympanic diaphragm seen from above. The arrows indicate the main ventilation of the epitympanum through the tympanic isthmus (one arrow is for the anterior epitympanum and the others are for the posterior epitympanum and mastoid). *ALFM*, anterior malleal ligamental fold; *MLF*, lateral malleal fold; *IMLF*, lateral incudomalleal fold; *TF*, tensor fold; *MA*, malleus; *SLIM*, superior malleal ligament; *IN*, incus; *IST*, tympanic isthmus; *S*, stapes; *PLIN*, lateral and medial posterior incudal ligaments

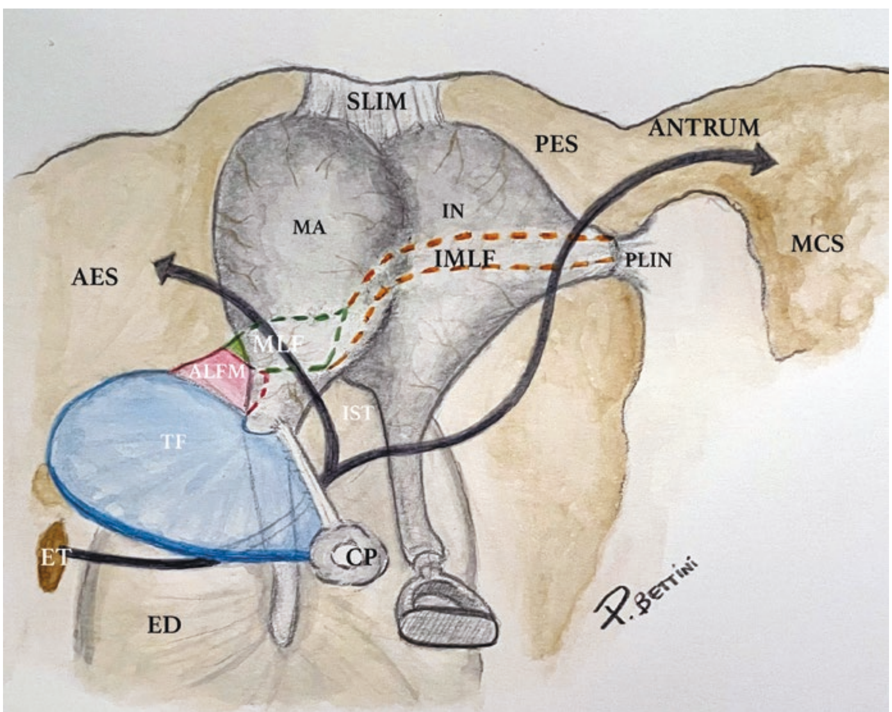

Fig. 6.4 The right ear. View of a complete tensor fold from a medial to a lateral direction and the main ventilation pathway of the epitympanum, through the tympanic isthmus. The epitympanic diaphragm is also seen in transparency as lateral to the incus and the malleus (dashed line). The black arrows show the ventilation of the anterior and posterior epitympanum. *AES*, anterior epitympanic space; *ET*, Eustachian tube; *TF*, tensor fold; *ALFM*, anterior malleal ligamental fold; *MLF*, lateral malleal fold; *MA*, malleus; *SLIM*, superior malleal ligament; *IN*, incus; *IMLF*, lateral incudomalleal fold; *IST*, tympanic isthmus; *ED*, eardrum; *PES*, posterior epitympanic space; *PLIN*, lateral and medial posterior incudal ligaments; *MCS*, mastoid cell; *CP*, cochleariform process

posterior wall corresponds to the posterior pouch of von Tröltsch.

The ventilation of Prussak space occurs through communication with the mesotympanum, represented by von Tröltsch's pouch, which is posteroinferiorly open, at the level of the most cranial portion of the mesotympanum, and it is laterally bordered by the pars tensa and the pars flaccida of the eardrum and medially by the posterior malleal ligament

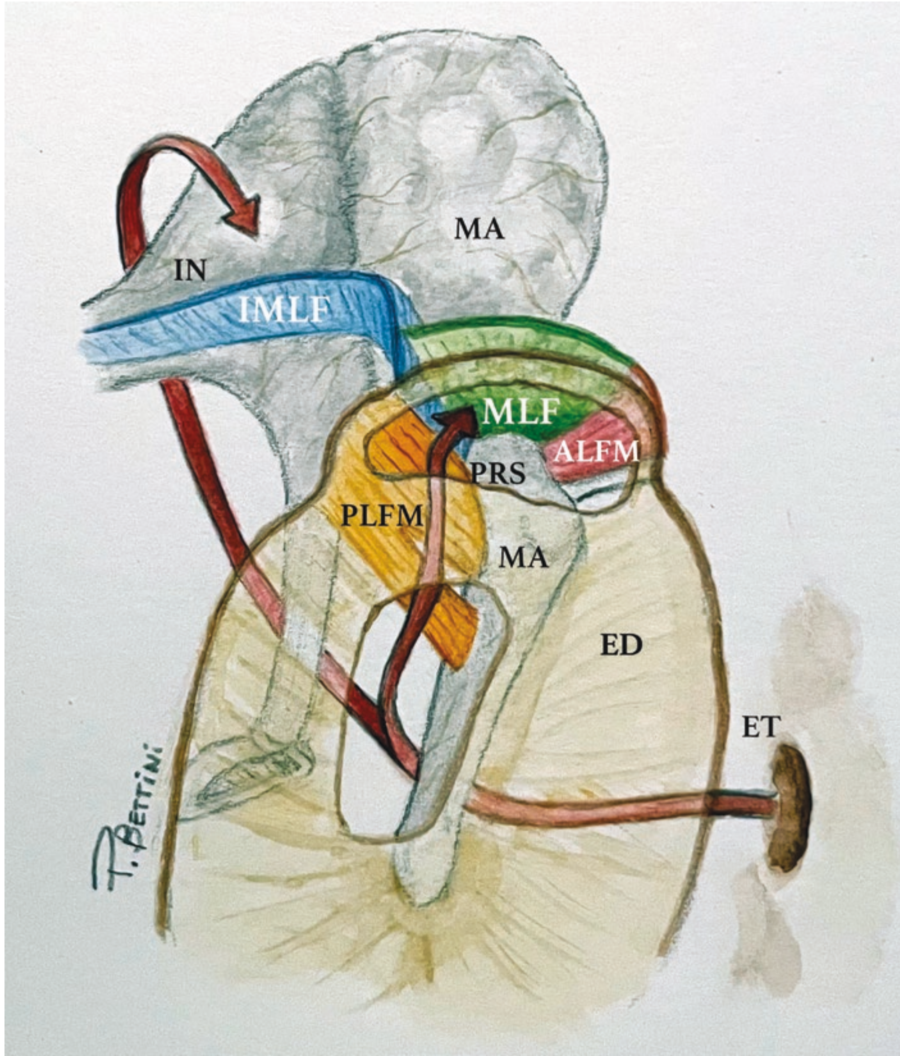

Fig. 6.5 The right ear. Lower and upper unit ventilation and epitympanic diaphragm. The tympanic membrane has been partially removed both in the pars flaccida (showing Prussak space) and in the pars tensa. The red arrows indicate the main ventilation pathway for the upper unit through the isthmus and for the lower unit (Prussak space) through von Tröltsch's pouch. *IN*, incus; *MA*, malleus; *IMLF*, lateral incudomalleal fold; *MLF*, lateral malleal fold; *ALFM*, anterior malleal ligamental fold; *PRS*, Prussak space; *PLFM*, posterior ligament of the malleus; *MA*, malleus; *ED*, eardrum; *ET*, Eustachian tube

(Fig. 6.5). The ventilation path of Prussak space is narrow and can easily tighten until it completely closes [16].

Even if the lower unit is an important component of the epitympanum, its aeration pathway can be blocked without affecting the function of the compartments, which lie superior to the epitympanic diaphragm or the one of other middle ear structures and the mastoid air system.

The Role of the Mastoid and Transmucosal Gas Exchange

The aerated mastoid is one of the three main factors influencing the ventilation of the middle ear.

Sadé and Ar [17] observed that the mastoid plays the role of an air reservoir to buffer rapid pressure changes within the middle ear (Fig. 6.6). Since volume and pressure are inversely proportional, a small, poorly aerated mastoid does not grant an optimal buffer function, allowing for wider pressure variations in the tympanic cavity (Fig. 6.7). Consequently, a largely aerated mastoid requires great changes in volume to achieve the same pressure as a small mastoid [12].

Another buffer mechanism is performed by the tympanic membrane since, and, if it is mobile enough, it has the possibility of bending inward and outward according to the pressure changes of the middle ear. Conversely, if the tympanic membrane is rigid and cannot bend properly, then the dilution of the pressure changes entirely depends on the aerated areas of the mastoid.

Since a small mastoid is most vulnerable to pressure changes, compensatory buffering mechanisms may develop, such as additional tympanic membrane retraction or middle ear volume reduction through fluid accumulation. A correlation exists between a poorly pneumatized mastoid and pars flaccida retraction, with a wider risk of atelectasis and subsequent cholesteatoma formation [18]. In the same way, it has been observed that children with recurrent otitis media, also complicated with cholesteatoma formation, usually have more sclerotic (less pneumatized) mastoids than do healthy children [19]. These examples provide further evidence of

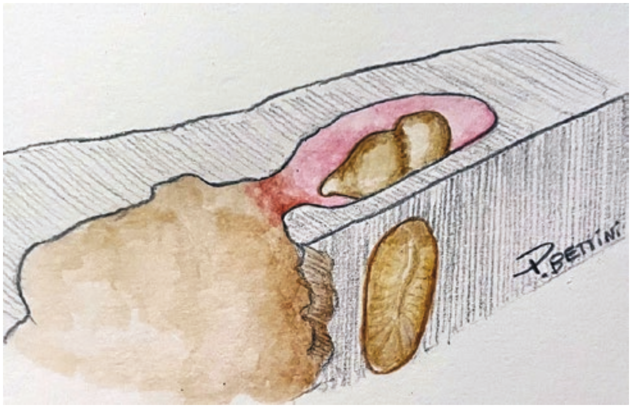

Fig. 6.6 The right ear. Regular buffer function of the mastoid. Sagittal section at the level of the tympanic membrane. Pink: middle ear; red: mastoid antrum; brown: mastoid

Fig. 6.7 The right ear. Sagittal section at the level of the tympanic membrane. Worse buffer function of a small mastoid that leads to wider pressure variations in the tympanic cavity, with a consequent retraction of the tympanic membrane (arrows) and a reduction of volume in the middle ear (arrows)

the fact that the mastoid plays an important buffering role in regulating the pressure of the middle ear [20].

As previously stated, the pressure balance is the result of the interaction between gas exchange in the mastoid air cells' mucosa, directly related to the degree of mastoid pneumatization, and a functioning Eustachian tube. Since these two mechanisms are interdependent, an isthmus blockage, even with a working Eustachian tube, could prevent mastoid cells' ventilation, leading to sclerotization of the mastoid itself. Görür et al. [21] classified the degree of radiological mastoid pneumatization shown in computed tomography (CT) scans as follows:

- Type 1 (normal pneumatization): The aerated mastoid cells reach the mastoid tip and the zygomatic, perisigmoid, and periantral regions and the aditus ad antrum is open.
- Type 2 (hypopneumatization): The aerated mastoid cells are found only in the antral and periantral regions and the aditus ad antrum is open.
- Type 3 (sclerotic): Mastoid pneumatization is absent and the aditus ad antrum is blocked.

Marchioni et al. ran CT scans on a group of subjects with an impairment of the middle ear aeration pathways. The vast majority of this group (18 out of 22) presented a hypopneumatized or scleroid mastoid. The degree of mastoid pneumatization was worse in the pathological ear, when compared to the contralateral one [22].

Blockage of the Ventilation Pathways

A primarily acquired cholesteatoma is morphologically characterized by epithelial cell proliferation. Unfortunately, despite a large number of studies, our understanding of the mechanisms underlying the pathogenesis of cholesteatoma is still limited. In the past, Bezold and Young suggested that the etiology of attic cholesteatoma could be attributed to an Eustachian tube dysfunction, resulting in an increased negative pressure in the tympanic cavity [19, 23]. On the other hand, Palva et al. observed how an attic retraction pocket could occur even in the case of a regular Eustachian tube function. Therefore, there must be other factors that affect the pathogenesis of attic cholesteatoma. The group led by Palva studied children with a blockage of the isthmus, who had undergone a tympanostomy tube placement, and they observed the persistence of inflammatory materials and cholesterol granuloma in the superior attic, despite the tympanostomy tube. They believed that an epitympanic diaphragm obstruction might be the origin of an attical cholesteatoma, stemming from an attical retraction. In contrast, children with an incomplete or absent tensor fold showed good ventilation of the anterior attic [24].

The theory that the obstruction of the aeration route between the mesotympanum and the attic is a contributing factor in the pathogenesis of attic retraction and cholesteatoma was severely challenged years ago by Yamasoba [25] and Kobayashi [26]. Yamasoba pointed out that it is possible for a cholesteatoma to stem from an attic retraction pocket despite the aeration of the mesotympanum in the middle ear. However, the importance of tensor fold removal during the surgical treatment of middle ear attic cholesteatoma remains of paramount importance, to restore the ventilation route from the protympanum to the anterior epitympanum, thus preventing the development of an attic cholesteatoma.

In microscopic ear surgery, since it is difficult to visualize the tensor fold due to its anterior position, some authors proposed different surgical approaches to microscopically visualize that fold. Morimitsu et al. [27] proposed an "anterior tympanotomy;" they removed the bone from the lateral attic to the zygoma, allowing for a possible drilling in front of the malleus head to remove the tensor fold. Palva et al. [28] suggested a tensor fold excision to create a large, new attic aeration pathway during cholesteatoma surgery through an endaural atticotomy extending to the supratubal recess.

With the introduction of an endoscope during middle ear surgery, it has become possible to study the anatomical variations and the ventilation pathways in pathological patients, owing to the magnification of critical areas such as the tensor fold and tympanic isthmus [16]. The exclusive transcanal endoscopic approach allows for a wide exposure of the anterior epitympanic compartment, especially the tensor fold and its inclination and its complete or incomplete structure, also granting the possibility of tensor fold removal with an angled instrument, thus restoring the ventilation of the upper unit.

The endoscopic approach to the tensor fold and to the anterior epitympanic compartment mainly depends on the dimensions of the anterior epitympanic space [29]. In most cases, it is wide, and the inferior endoscopic approach to the tensor fold area provides a good exposure to the inferior edge of the tensor fold, allowing for its removal and restoring direct ventilation from the protympanic space to the anterior epitympanic space.

In subjects with an extremely narrow anterior epitympanic space, associated with a vertical orientation of the tensor fold, it is only possible to explore the fold component close to the cochleariform process. In this case, a removal of the incus and the head of the malleus is necessary to obtain a good visualization of the superior edge of the tensor fold.

Selective Epitympanic Dysventilation Syndrome

As stated above, a physiological middle ear pressure seems to be connected to a functioning Eustachian tube and to the transmucosal gas exchange through the mastoid mucosa, which is directly related to the degree of pneumatization. An inflammatory middle ear chronic disease may thus be due to an Eustachian tube dysfunction and subsequent poor tympanic ventilation, which is often associated with the hypopneumatization or sclerotization of the mastoid cells [15, 16]. Conversely, in subjects with an excellent Eustachian tube function, two different conditions could give rise to an epitympanic selective dysventilation. The first condition is an obstruction of the posterior pouch of von Tröltsch, which closes the gap between the tympanic membrane and the posterior malleal ligamental fold, creating a selective dysventilation of the lower unit (Prussak space) [24]. The second state is an obstruction of the isthmus associated with a complete tensor fold, determining a selective dysventilation of the upper unit [30]. Several factors may obstruct the tympanic isthmus or von Tröltsch's pouch, such as mucosal fold variations, inflammatory webs, a retracted tympanic membrane, a pathological attic mucosa, and a cholesteatoma. In the two abovementioned conditions, a limited disease in the respective areas, possibly associated with an attical retraction, might occur [14]. As a confirmation of this, a tympanic isthmus obstruction is a consistent finding in patients with a limited attic cholesteatoma (Fig. 6.8).

A selective epitympanic dysventilation syndrome thus appears with four simultaneous distinctive features: an attic retraction pocket or cholesteatoma, a type A tympanogram or a regular tubal function test, a complete epitympanic diaphragm, and an isthmus blockage. In patients with this syndrome, while the epitympanum is poorly ventilated, with a subsequent decreased pressure that also extends to the mastoid, the mesotympanum is well-aerated, receiving air directly from a functioning Eustachian tube. In clinical practice, it is not uncommon to find an isolated retraction pocket of the pars flaccida and/or a cholesteatoma in this region, with a normal pars tensa and mesotympanum. Conversely, when the Eustachian tube function is impaired, the middle ear mucosa is usually widely involved in the inflammation process, and the tympanic membrane is completely retracted.

So, in patients with a selective epitympanic dysventilation, it is usually possible to intraoperatively appreciate an open Eustachian tube, a good protympanic and mesotympanic mucosa, and a partial or complete isthmus blockage that could alone explain it, thus strengthening the preoperative evaluation of a working Eustachian tube [14, 31]. From this, we can understand the importance of a surgical approach that can completely visualize the epitympanic diaphragm and surgically restore the ventilation pathways.

Selective Dysventilation of the Mesotympanum

Pars tensa cholesteatoma is a primarily acquired (retraction pocket) cholesteatoma that has a different pathogenesis from that of the attic, and it is associated with a negative middle ear pressure, due to a chronic Eustachian tube dysfunction. However, a ventilation dysfunction in the mesotympanum could also be attributed to fibrous or mucous webs between the medial wall of the tympanic cavity and the tympanic membrane that can exclude the tubal orifice and can create selective dysventilation areas in the anterior or posterior mesotympanum. This often leads to selective mesotympanic retraction pockets rather than the whole mesotympanum [32].

Fig. 6.8 The right ear. Lateral-to-medial view with a slight anterior rotation. Selective epitympanic dysventilation syndrome: blockage of the tympanic isthmus with a complete tensor fold. The arrow indicates the ventilation of the mesotympanum, through the Eustachian tube. The epitympanum is not properly ventilated due to the tympanic isthmus blockage. Prussak space is regularly aerated through von Tröltsch's pouch. ****: blockage of the tympanic isthmus. *PLIN*, lateral and medial posterior incudal ligaments; *IMLF*, lateral incudomalleal fold; *IN*, incus; *MA*, malleus; *MLF*, lateral malleal fold; *PRS*, Prussak space; *TF*, tensor fold; *ET*, Eustachian tube

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