The Laryngoscope

Selective Epitympanic Dysventilation Syndrome

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Objectives/Hypothesis: Although middle ear aeration is certainly related to eustachian tube (ET) function, other anatomic factors may play an important role in ventilation of these spaces, particularly the epitympanum.

Study Design: A prospective case series study group with retrospective case series comparison.

Methods: Eight patients affected by non–self-cleaning attic retraction pockets or attic cholesteatoma of the middle ear and normal tubal function (verified by type A tympanogram or Williams' test) underwent endoscopic surgery. Anatomic findings were noted and collected, with particular attention paid to middle ear folds. A control group was used for comparison. The prevalence of an isthmus blockage with a complete tensor fold in pathologic ears was compared between the study group and control group.

Results: The higher prevalence of an isthmus blockage associated with a complete tensor fold in the study group was found to be statistically significant ($P = .001$).

Conclusions: A selective epitympanic dysventilation syndrome is hypothesized; normal ET function seems to be insufficient for complete ventilation of the middle ear, and an open tympanic isthmus or an incomplete tensor fold may be necessary for epitympanic recess ventilation. The selective epitympanic dysventilation syndrome would consist of the contemporary presence of the following four conditions: attic retraction pocket or cholesteatoma, normal tubal function tests, complete epitympanic diaphragm, and isthmus blockage.

Key Words: Selective dysventilation, isthmus blockage, eustachian tube.

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INTRODUCTION

Aeration of the tympanic cavity and mastoid cells and anatomic pathways for middle ear ventilation have been studied since the end of the 19th century starting with the work of Prussak in 1867.[1] More recently, Palva and colleagues were the first to describe middle ear anatomy focusing on ventilation patterns and their implications for middle ear disease.[2–5] It is well known that the eustachian tube (ET) plays a crucial role in maintaining middle ear aeration and atmospheric pressure. Usually, inflammatory middle ear chronic disease is related to ET dysfunction due to poor tympanic ventilation. This is also related to hearing impairment and poor postoperative outcomes.[6] Although middle ear aeration is certainly related to ET function, other anatomic factors may play an important role in ventilation of these spaces. In fact, epitympanum aeration is strictly dependent on the ventilation pathways clarified by Palva and colleagues: if the tensor fold and the lateral incudomalleal fold are complete, the only ventilation pathway to the epitympanum is through the tympanic isthmus.[3,4] In such cases, when an isthmus blockage occurs, the ventilation of the epitympanum may be impaired, and the only gas exchange would come from the mucosa of mastoid cells.[7]

The aim of this report is to study middle ear anatomy, focusing on middle ear folds in patients with attical retractions or cholesteatoma and with a normal tubal function test, who underwent endoscopic surgery. This scenario might describe a selective epitympanic dysventilation syndrome, possibly not related to ET impairment.

MATERIALS AND METHODS

The setting was the otolaryngology department of the tertiary referral University Hospital of Modena in Italy. A study and a control group were selected and then compared. A three-chip high-resolution monitor and camera (Karl Storz, Tuttlingen, Germany) were used for all of the procedures.

Study Group

From December 2008 to March 2009, 21 patients affected by a non–self-cleaning attic retraction pocket or frank attic

cholesteatoma of the middle ear underwent endoscopic tympanoplasty. All patients who were candidates for ear surgery underwent high-resolution computed tomography, audiometric, and impedenziometric evaluations. Subjects affected by a disease of the epitympanic compartment (non–self-cleaning attic retraction pockets, epitympanic cholesteatoma) and with a normal tubal function tests (type A tympanogram or Williams' test)[8] were included in present study. Subjects affected by a disease involving the protympanic, the mesotympanic, and the retrotympanic region, or patients who had previously undergone middle ear surgery were excluded from the study. The final study group consisted of eight patients. During the endoscopic approach, the appearance of the tympanic isthmus, the tympanic folds, and the ET mucosa and opening were carefully observed in all patients. These features were precisely described in the operative report and reported in a database soon after the operations. All of the operations were recorded to form a video archive.

Control Group

A retrospective control group of eight patients who had been operated on for simple chronic otitis media without epitympanic disease, and whose operations had been video recorded and archived in our department, was selected. Patients who had already had middle ear surgery in the past were excluded from the control group. Evaluation of the tympanic isthmus, the tympanic folds, and the ET mucosa and opening was done based on the videos and a database.

Surgical Approach for the Study Group

A 3-mm 0° endoscope was used to create the tympanomeatal flap with the superior limb at the 1 o'clock position and inferior limb at the 6 o'clock position. The tympanomeatal flap was elevated under endoscopic control until the annulus was visible. We entered over the annulus into the tympanic cavity inferiorly transposing the tympanomeatal flap. A transcanal atticotomy was performed using a microdrill system under endoscopic vision until the epitympanic compartments and the incudomalleal joint were visible. When a cholesteatoma sac was present, angulated instruments were used to remove the sac from the epitympanic compartments. The incus and malleus head were removed if required. At the end of the procedure, after the pathologic tissue had been removed from the epitympanic compartments, we focused on exploring the isthmus and the tensor fold to better understand the anatomy of the epitympanic diaphragm. The lateral endoscopic approach to the tympanic cavity allowed us to check the isthmus, the protympanic space, and the tensor fold region.

The tympanic isthmus was explored using 3-mm 0° and 45° endoscopes. In this way, exploration of the tympanic isthmus was possible between the medial part of the posterior incudal ligament posteriorly and the tensor tendon anteriorly. The 0° endoscope was used for better magnification of the space between the incudostapedial joint and the cochleariform process with the tensor tendon (Proctor's anterior isthmus). A 3-mm 45° endoscope was used for better magnification of the space between the pyramidal process and the short process of the incus, and also to examine the medial attic space.

To explore the protympanic space, a 3-mm 45° endoscope was used and rotated anteriorly. During the surgical procedure, we also observed the opening of the ET and the supratubal recess.

Endoscopic examination of the tensor fold area was possible using a 3-mm 45° endoscope. Two different approaches already described by the authors were used to examine the orientation of the tensor fold[9]: 1) With the inferior approach to the tensor fold, a 45° endoscope is inserted into the protympanic region. After identification of the eustachian tube and the supratubal recess, the endoscope is rotated superiorly. This procedure allows good exposure of the inferior aspect of the tensor fold. 2) With the superior approach to the tensor fold, after a wide anterior atticotomy exposing the incudomalleal joint and the anterior epitympanic space, a 45° endoscope is inserted into the epitympanic compartments, rotating the endoscope inferiorly. This procedure allows good exposure of the superior aspect of tensor fold.

At the end of the endoscopic procedure, a composite tragal cartilage/perichondrial graft was placed underneath to close the epitympanic defect of the lateral bony wall.

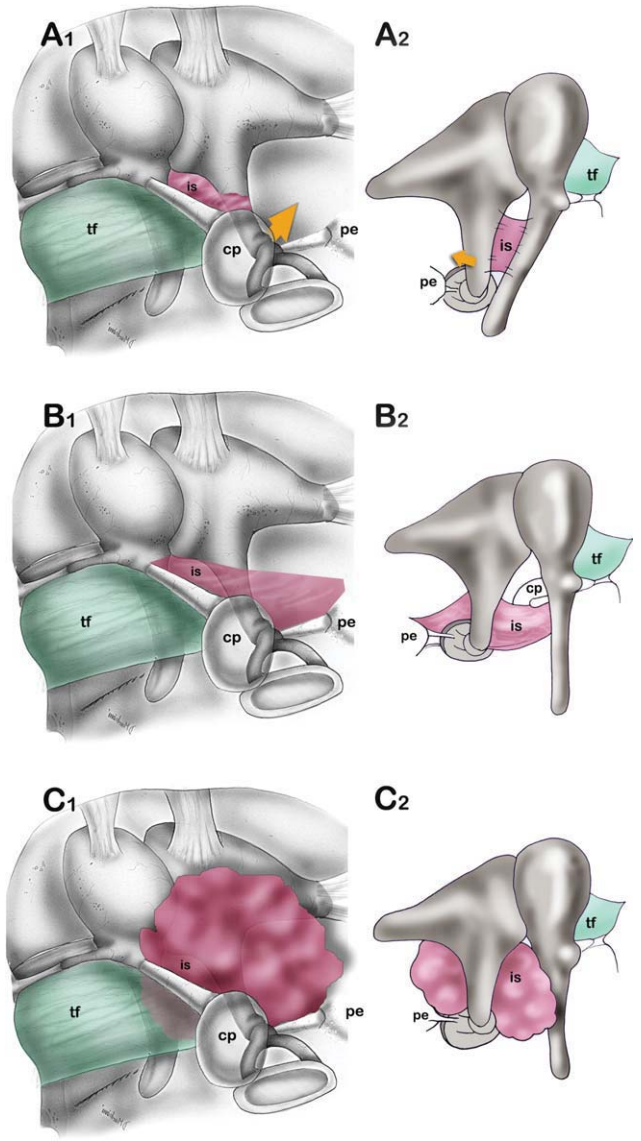

Fig. 1. Anatomical findings in study group. (A) Partial isthmus blockage with a small opening in the space between the pyramidal eminence and the incudostapedial joint. A1: medial view; A2: lateral view. (B) Complete isthmus blockage. Mucosal fold is inserted posteriorly on the pyramidal eminence, anteriorly involving the space between the incudostapedial joint and the cochleariform process. B1: medial view; B2: lateral view. (C) Granulation tissue closing the isthmus. C1: medial view; C2: lateral view. tf = tensor fold; is = isthmus; pe = pyramidal eminence; cp = cochleariform process. [Color figure can be viewed in the online issue, which is available at www.interscience.wiley.com.]

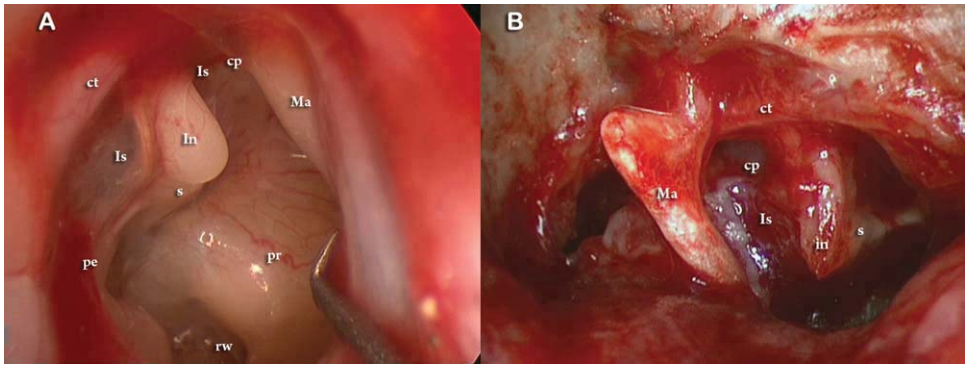

Fig. 2. Endoscopic findings in study group. (A) Isthmus blockage with a complete mucosal fold. (B) Complete isthmus blockage by granulation tissue. ct = chorda tympani; Is = isthmus; cp = cochleariform process; Ma = malleus; in = incus; s = stapes; pe = pyramidal eminence; pr = promontorium; rw = round window. [Color figure can be viewed in the online issue, which is available at www.interscience.wiley.com.]

Surgical Approach for the Control Group

A 3-mm 0° endoscope was used to create the tympanomeatal flap with the superior limb at the 1 o'clock position and the inferior limb at the 6 o'clock position. The tympanomeatal flap was elevated under endoscopic control until the annulus was visible. We entered over the annulus into the tympanic cavity inferiorly transposing the tympanomeatal flap. The same examinations of the middle ear anatomy were made as described above. At the end of the endoscopic procedure, a perichondrial graft was placed underneath to close the tympanic membrane.

Statistical Analysis

Intergroup results were compared using a Fisher exact test (χ^2 analysis was not applicable because of the small number of subjects). A value of $P < .05$ was considered statistically significant.

RESULTS

Study Group

The patients ranged in age from 25 to 55 years (mean age, 44.3 years). Five subjects were men and three were women. In four subjects a type A tympanogram was obtained, whereas in four subjects with type B tympanogram a negative Williams' test was obtained; therefore, they were included in the study group. Ear pathology included five subjects affected by cholesteatoma limited to the epitympanic compartments and three subjects affected by a severe retraction pocket involving the epitympanic compartments. In all eight subjects, it was possible to analyze the epitympanic diaphragm, observing the tensor fold area and the isthmus.

We observed a complete blockage of the isthmus associated with a complete tensor fold in all eight subjects. In these subjects there was a mucosal fold reducing the dimensions of the isthmus space or closing it completely. In two subjects there was also granulation tissue closing the isthmus (Fig. 1C). In six subjects the isthmus was completely blocked; in these patients the mucosal fold was inserted posteriorly on the pyramidal eminence, anteriorly involving the space between the incudostapedial joint and the cochleariform process (Fig. 1B). In two subjects the isthmus was partially blocked; there was only a small opening in the space between the pyramidal eminence and the incudostapedial joint, but there was a complete obstruction of the space between the incudostapedial joint and the cochleariform process (Fig. 1A). In these patients there was a medialization of the malleus to the incudostapedial joint. In all eight subjects we found a complete tensor fold separating the anterior epitympanic space from the protympanic space (Fig. 2). In three subjects the tensor fold had a vertical orientation, arising posteriorly from the tensor tendon of the malleus and attaching anteriorly to the anterior bony wall of the epitympanic space; a wide supratubal recess was present in these patients. In five subjects the tensor fold had a horizontal orientation, arising posteriorly from the tensor tendon of the malleus and attaching anteriorly to the tegmen tubae. In two of these patients there was a very small supratubal recess, whereas in three there was no supratubal recess.

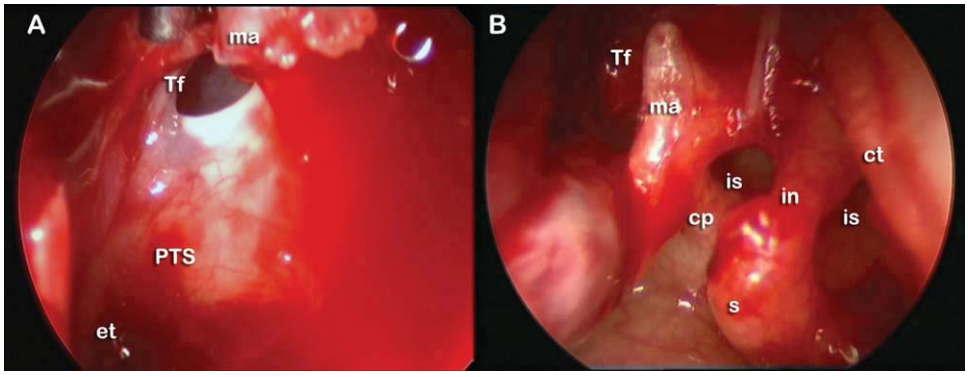

Fig. 3. Endoscopic findings in control group. (A) Partial tensor fold. (B) Complete isthmus opening. ma = malleus; Tf = tensor fold; PTS = partial tensor fold; et = eustachian tube; ct = chorda tympani; is = isthmus; cp = cochleariform process; in = incus; s = stapes. [Color figure can be viewed in the online issue, which is available at www.interscience.wiley.com.]

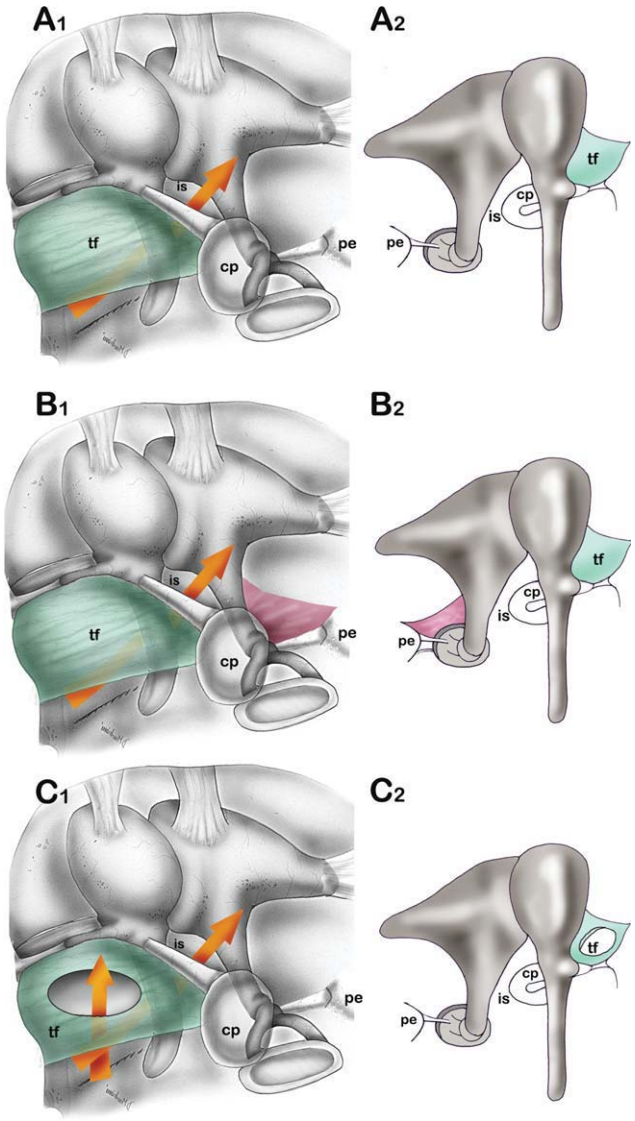

Fig. 4. Anatomical findings in control group. (A) Wide open isthmus and complete tensor fold. A1: medial view; A2: lateral view. (B) Thin mucosal fold closing the space between the incudostapedial joint and the pyramidal eminence blocking the posterior portion of the isthmus. B1: medial view; B2: lateral view. (C) Wide open isthmus and incomplete tensor fold. C1: medial view; C2: lateral view. is = isthmus; tf = tensor fold; cp = cochleariform process; pe = pyramidal eminence. [Color figure can be viewed in the online issue, which is available at www.interscience.wiley.com.]

In all eight subjects, we observed the mucosa of the protympanic and mesotympanic space to be in good condition, whereas in six subjects the mucosa of the epitympanic space presented degeneration areas with epithelialization. Endoscopic exploration of the opening of the eustachian tube showed normal conditions in all eight subjects, and we did not find any inflammatory tissue obstructing the eustachian tube.

Control Group

Patients ranged in age from 20 to 56 years (mean age, 40.2 years). Four subjects were men and four were women. It was possible to analyze the epitympanic diaphragm in all eight subjects in the control group, observing the tensor fold area and the isthmus.

In seven subjects in the control group, we found a wide isthmus with a good communication from the mesotympanic compartment to the epitympanic compartments (Fig. 3). We observed a wide opening in the isthmus associated with a complete tensor fold in six of these subjects (Fig. 4A). In one of these subjects, there was an open isthmus associated with an incomplete tensor fold, and we found a wide opening inside the tensor fold permitting a direct communication from the protympanic space to the anterior epitympanic space (Fig. 4C). The endoscopic exploration of the opening of the eustachian tube and mucosa showed normal conditions in all eight subjects, and we did not find inflammatory tissue obstructing the eustachian tube. In one subject, the isthmus was almost blocked, presenting a thin mucosal fold closing the space between the incudostapedial joint and the pyramidal eminence blocking the posterior portion of the isthmus, whereas the space between the incudostapedial joint and the cochleariform process was wide and open, possibly with adequate communication from the mesotympanum to the epitympanum; also in this patient, there was a complete tensor fold (Fig. 4B).

Intergroup Comparison

The prevalence of an isthmus blockage with a complete tensor fold in pathologic ears was compared between the study group (8/8) and control group (1/8). The higher prevalence of an isthmus blockage associated with a complete tensor fold in the study group reducing the ventilation to the epitympanic compartments was found to be statistically significant ($P = .001$).

DISCUSSION

In 1946, Chatellier and Lemoine formulated the concept of "the epitympanic diaphragm." The authors described different ligament and membranous folds, which together with the malleus and incus form the floor of a large epitympanic compartment. This space represents the upper unit and is aerated from the protympanic space through the tympanic isthmus.[10] Aimi described the tympanic isthmus as a narrow passage between the tubotympanic cavity and the atticomastoid air space.[11] He observed that obstruction of the tympanic isthmus is common in various types of middle ear disease and causes significant air-diffusion disturbance within the temporal bone pneumatic system. Aimi also noted that the factors that caused an obstruction of the tympanic isthmus were mucosal fold variations, inflammatory webs and exudate, retracted tympanic membrane, diseased attic mucosa, and cholesteatoma.[11]

A functioning ET is an integral part of a normal middle ear, and thus is an essential requirement for optimum results in tympanoplasties. ET plays a crucial role in maintaining middle ear aeration and atmospheric pressure, and inflammatory middle ear chronic disease may be related to ET dysfunction due to poor tympanic ventilation. Middle ear pressure seems to be related not only to a functioning eustachian tube but also to

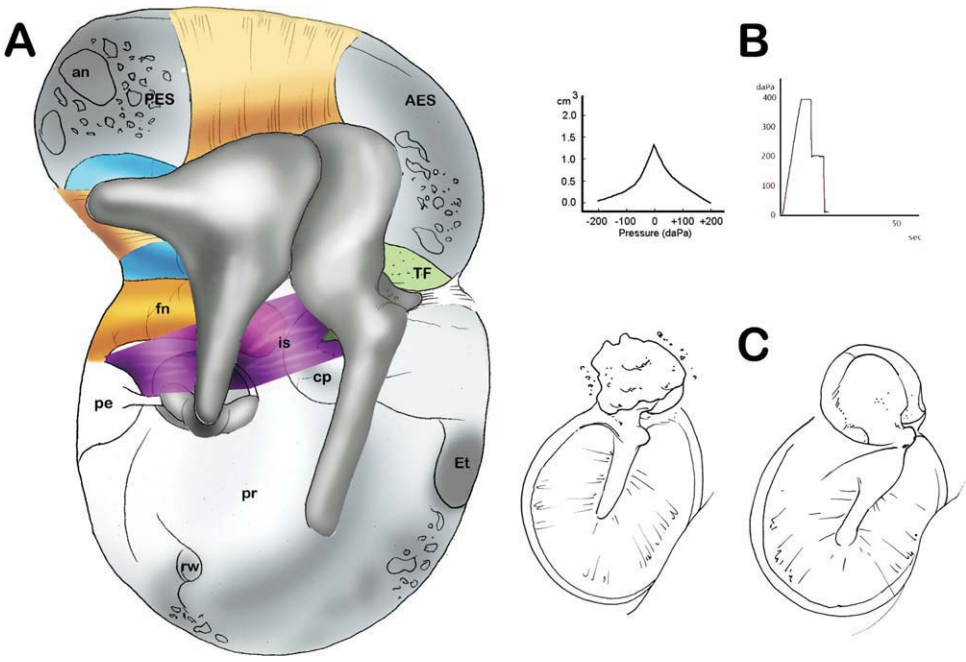

Fig. 5. Selective epitympanic dysventilation syndrome. (A) Middle ear with complete isthmus blockage, epitympanic diaphragm, and complete tensor fold. (B) Type A tympanogram and Williams' test. (C) Attic cholesteatoma and retraction pocket. an = antrum cell; PES = posterior epitympanic space; AES = anterior epitympanic space; lsc = lateral semicircular canal; TF = tensor fold; fn = facial nerve; is = isthmus; pe = pyramidal eminence; cp = cochleariform process; pr = promontorium; Et = eustachian tube; rw = round window; *isthmus blockage. [Color figure can be viewed in the online issue, which is available at www.interscience.wiley.com.]

transmucosal gas exchange through the mastoid mucosa. The mucosal gas exchange is related to the degree of mastoid pneumatization,[12] which begins on the 33rd gestational week and continues up to 8 or 9 years.[13] These two gas pressure systems should reach equilibrium, which is possible if ventilation pathways of the middle ear are open. In a recent work, the authors found that obstruction of the tympanic isthmus is a consistent finding in patients affected by limited attical cholesteatoma.[9] We also recently studied middle ear ventilation route blockage and its relationship to mastoid pneumatization. We found and described three major types of isthmus blockage related to different pathologic conditions. All of them were significantly associated with hypopneumatization or sclerotization of mastoid cells.[14]

In our earlier studies, ET function was not tested as in the present series. Based on the emerging data obtained from our former publications and the present article, we hypothesize a selective epitympanic dysventilation syndrome. If an isthmus blockage occurs in an ear with complete tensor and incudomalleal folds, a selective epitympanic dysventilation may manifest even with a functioning ET. So the syndrome would occur with the contemporaneous presence of four conditions: an attic retraction pocket or cholesteatoma, a type A tympanogram or a normal tubal function test, complete epitympanic diaphragm, and isthmus blockage (Fig. 5).

In the majority of the subjects, the tensor fold and the lateral incudomalleal fold are complete, and the only ventilation pathway to the epitympanum is through the tympanic isthmus. When the isthmus is open and ET function is normal, the epitympanic compartments and the mastoid cells are ventilated and the pressure level is homogeneous; this should probably guarantee normal conditions in the tympanic cavity. Otherwise, when an isthmus blockage occurs, the epitympanum is not well-ventilated, creating different conditions from the mesotympanic compartment, which receives air directly from the ET. In clinical practice it is not uncommon to find an isolated retraction pocket of the pars flaccida and/or a cholesteatoma, limited to the posterior epitympanum, with an otherwise normal pars tensa and mesotympanum. When ET function is impaired, middle ear mucosa is usually widely involved by the inflammation process and the eardrum is completely retracted; however, if the ET is functioning the retraction pocket might possibly be related to a selective dysventilation process.

As confirmed intraoperatively, an open ET and a good protympanic mucosa appearance were found in all eight patients in the study group, strengthening our preoperative evaluation of ET function, whereas a selective dysventilation of the epitympanic compartments could be explained by a complete isthmus blockage in six patients and by a partial blockage in the others. These findings, associated with a complete tensor fold and an epitympanic retraction pocket of the eardrum, effectively describe a selective epitympanic dysventilation syndrome. In these patients, a functioning ET normally equalizes middle ear pressure, but a selective dysventilation of the excluded epitympanic region might cause a retraction pocket, or in some cases, an epitympanic cholesteatoma after an undefined period of time. On the other hand, seven patients in the control group had an isthmus that was wide open, and in one case, there was a partial isthmus blockage. In one case, the tensor fold was incomplete, and an additional ventilation pathway was possible. All of these patients had a normal epitympanic recess without any signs of dysventilation.

Although the results to date are encouraging, they must be considered preliminary as the number of patients in our study and control group was small. A possible bias may be caused by the retrospective selection of the control group, and thus our results need to be confirmed in the future by prospective trials using a greater number of patients. Moreover, the sole

tympanogram is an indirect measure and may have underestimated the number of patients with abnormal tubal function; on the contrary, Williams' tympanometric swallow test[8] may have overestimated the number of patients with abnormal ET function with some false positives.[15]

We strongly believe that the physiopathology of middle ear disease requires a better understanding, and endoscopic middle ear surgery may help us in this purpose, allowing the surgeon to look behind the corner without radically changing middle ear anatomy, and thus completely explore all ventilation pathways. From this point of view, we strongly suggest that during middle ear surgery, special attention is paid to restoring an isthmus ventilation pathway, removing inflammatory tissue, or recreating a new isthmus with an ossiculoplasty; the tensor fold should usually be removed to create an accessory ventilation route to the epitympanum. The abovementioned procedures are necessary for good epitympanic ventilation. Awareness and early diagnosis of selective middle ear dysventilation problems in the future could prevent the development of chronic otitis and cholesteatoma.

CONCLUSION

A selective epitympanic dysventilation syndrome is hypothesized, consisting of the contemporaneous presence of an attic retraction pocket or cholesteatoma, normal tubal function tests, complete epitympanic diaphragm, and isthmus blockage. Normal ET function may not be sufficient for complete ventilation of the middle ear, but an open tympanic isthmus or an incomplete tensor fold may be necessary for epitympanic recess ventilation. To treat this condition and perhaps to prevent cholesteatoma formation, a new ventilation route should be created during surgery, and this can be performed by endoscopic middle ear surgery in a very preservative way.

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