

Original article

MANAGEMENT OF LABYRINTHINE FISTULA IN CHRONIC OTITIS WITH CHOLESTEATOMA: CASE SERIES

Francesco Dispenza¹, Walter Mazzucco², Sebastian Bianchini¹,
Sergio Mazzola², Ettore Bennici¹

Summary

Labyrinthine fistula is a complication of ear cholesteatoma that increase the risk of sensorineural hearing loss. The management of the fistula must be done contextually with mastoidectomy by: leaving cholesteatoma matrix over the fistula, or remove the matrix reconstructing the defect. *Objective:* analysis of the two techniques to treat labyrinthine fistula. *Methods:* retrospective review with case series analysis. *Results:* a labyrinthine fistula was present in 14% of cholesteatoma patients; CT scan was predictive in all cases; the hearing preservation was obtained with both techniques; a recurrence was detected only in one case; postoperative nystagmus incidence was higher in those cases with matrix left in situ and when the size of the fistula was larger than 2 mm. *Conclusions:* the labyrinthine fistula have to be treated contextually with cholesteatoma removal, both techniques had good postoperative hearing preservation rate. The postoperative vertigo with nystagmus is more frequent in larger fistulas.

Introduction

According to different published studies, labyrinthine fistula (LF) prevalence in patients diagnosed with otitis media ranges from 5.8% to 21%. In an epidemiological prospective observational study conducted on a total of 1.816 patients diagnosed with otitis media, it was found that cholesteatoma is the most frequent etiology of intratemporal complications such as labyrinthine fistula¹. Although incidence of intratemporal complications in developing remains significant when compared with developed countries, more recent studies documented discordant results, as similar high incidence of complications associated with chronic otitis media was highlighted in a US metropolitan public hospital as well as lower occurrence was reported in southern Iran^{2,3}.

The natural history of the cholesteatoma includes temporal bone progressive erosion and also the otic capsule may be involved leading to LF. The opening of inner ear exposes the patients to lose the hearing and vestibular function. The symptoms com-

Address of the authors:

¹ U.O.C. Otorinolaringoiatria, Ospedale San Giovanni di Dio, Agrigento.

² Department of science for Health Promotion and Mother to child care G. D'Alessandro; University of Palermo.

Send correspondence to: Francesco Dispenza, francesco.dispenza@gmail.com

Received: 28th December, 2015 — **Revised:** 30th December, 2015 — **Accepted:** 31th December, 2015

plained by patients with cholesteatoma include: firstly progressive hearing loss and otorrhea, followed by vertigo or some major complications (i.e. facial palsy, intracranial infections) after worsening of the disease. A labyrinthine fistula exposes the inner ear fluids to potential infection and to pressure variation; both factors may induce alterations of inner ear functions. In patients with middle ear cholesteatoma complaining vertigo could be present a LF. The vertigo spell with nystagmus may be elicited with a simple pressure on the tragus (fistula test), as well as by increasing air pressure in the external ear canal (Hennebert sign). Although this clinical signs may be indicative, the tests are positive only in 50% of cases⁴. According to literature, LF prevalent localization in patients with acquired cholesteatoma is in the lateral semicircular canal (LSC)⁵. The imaging, not always required for middle ear cholesteatoma, is suggested in case of suspected LF in those patients with vertigo and positive fistula sign. The high resolution CT scan without contrast enhancement is the gold standard to display the otic capsule status to plan the more appropriate surgical treatment in case of hearing preservation attempt, evaluating the likely size⁶.

One of the first surgical treatments of a labyrinthine fistula has been attributed to Nylen⁷, which removes the matrix over the fistula, leaving the site uncovered. In the last century the operation of inner ear fenestration become common to treat otosclerosis, but the trend of the treatment of LF was to leave matrix over the fistula⁸. For this reason the debate is still to whether to remove the matrix or leave it in situ, because several Authors reported a high hearing preservation rate also after matrix removal^{9,10}.

We report our experience in management of LF in cholesteatoma surgery, adopting both techniques.

Material and Methods

A retrospective review of charts in the period 2011-2014 was done in the ENT clinic of our Hospital among patient treated for ear cholesteatoma. Only patients reporting a LF in the description of operation were collected. The analysis

included: patients data (age, sex, side of the disease), presenting symptoms, pre- and post-operative nystagmus evaluation with Frenzel lens, pre- and post-operative hearing tested by pure tone audiometry (PTA) reported by average of 0.25-0.5-1-2-4 kHz frequencies, Hennebert sign recording, type of operation, size of fistula, site of fistula. Postoperative variation of more than 10 dB in average PTA was considered worsening of the hearing. Only patients with at least 12 months of follow-up were included in the review. All patients undergo to CT scan without contrast enhancement and the evidence of otic capsule erosion was reported. All patients were treated on by the same surgeon (first Author). The surgical management was done by a canal wall down mastoidectomy in all cases because of preoperative evidence of cholesteatoma at micro-otoscopy and with CT scan predictive for LF. The generally adopted approach to the fistula was to leave the matrix over the fistula in all cases with normal or near-normal bone conduction at PTA, while matrix removal was chosen in those patients with preoperative profound sensorineural hearing loss.

Results

Among 57 patients treated at our institution for cholesteatoma between 2011 and 2014, 8 (14%) patients were positive for LF. All patients with preoperative evidence of cholesteatoma and CT scan suspect for LF were managed with canal wall down mastoidectomy. The Hennebert sign was positive in 3 patients, of which 2 had less than 2 mm LF. The management of the fistula was the matrix removal in 3 cases: in two cases with preoperative profound sensorineural hearing loss and in one patient with less than 2 mm fistula size and intraoperative evidence of intact endosteal layer after matrix elevation (Figure 1 A-B). In 5 patients a thin layer of the matrix was left over the fistula to prevent open labyrinth (Figure 2 A-B). In all cases the fistula involved the lateral semicircular canal (LSC). Three cases of 8 treated had a less than 2 mm fistula. Two patients had a multiple site fistula: one with posterior semicircular canal in-

volvement and one with anterior arm of superior semicircular canal fistula (Figure 3); both patients had a preoperative pro-

found sensorineural hearing loss. In all case the CT scan was predictive for LF. A postoperative nystagmus was recorded in

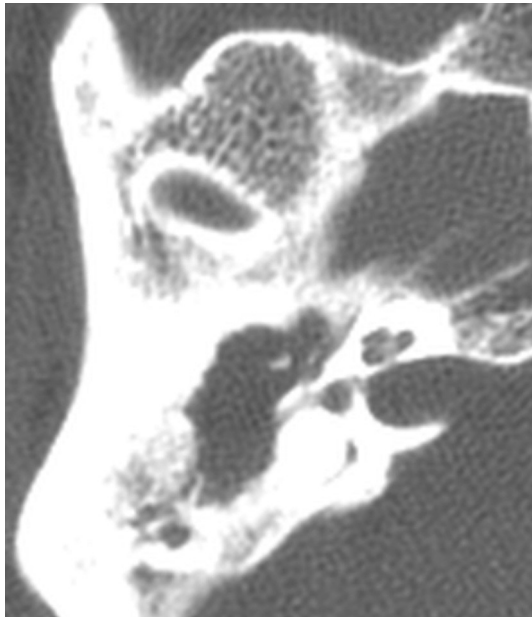


Figure 1 (A-B): Case with < 2 mm labyrinthine fistula managed with modified radical mastoidectomy and matrix removal over the fistula. A (left): CT scan shows the horizontal canal bone interruption. B (right): intraoperative view of the open fistula after matrix removal over the horizontal canal.

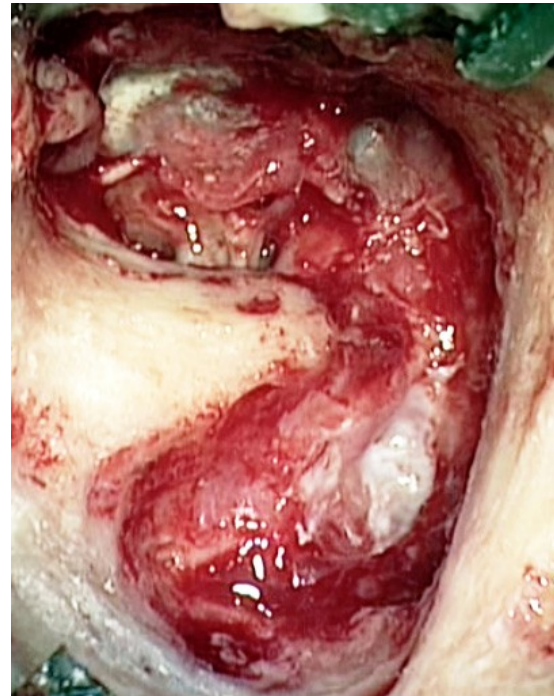


Figure 2 (A-B): Case with > 2 mm labyrinthine fistula managed with modified radical mastoidectomy and matrix left over the fistula. A (left): CT scan showing the bone erosion on horizontal canal and also erosion of the middle cranial fossa plate. B (right): intraoperative finding of the horizontal canal fistula with matrix left in situ.

3 patients treated leaving the matrix over the LF. Only one patient experienced a worsening of the hearing after surgery. In the follow-up one patients managed with matrix over the fistula had residual cholesteatoma after 16 months since surgery, but the site of residual (pearl of cholesteatoma) was far from the LF site. All case series data are summarized in Table 1.

Discussion

Labyrinthine fistula is one of the most frequent complications of cholesteatoma progression, occurring in 14% of our cases. The horizontal canal is involved in about 90% of patients because of its anatomical position expose directly the otic capsule to cholesteatoma action¹¹. The LF could be present in both cholesteatoma and chronic granulomatous otitis media without signs of cholesteatoma, and the fistula is often observed without reactive changes in the inner ear histology. The bone destruction is a multifactor product by: acid pH, proteolysis via cathepsin and matrix metalloproteinase 9 and acid phosphatase degradation of collagen¹². The clinical sign as fistula sign or Henne-

bert test have low specificity and are present in about 25% of cases as in our case series. The reason might be that transmission of pressure changes from the external ear canal to the fistula is interrupted by the cholesteatoma mass. Furthermore, the positivity of the fistula test was not related with the fistula type or size. Symptoms as vertigo spells and positivity of fistula sign are poor indicators of LF, as the literature confirmed¹³. A first staging system of LF was proposed by Palva and Johanson in 1986¹⁴, which categorized fistulae by the depth of otic capsule erosion; but this staging system has not been adopted systematically in literature. The commonest categorization of LF is based on fistula diameter reported in millimeters. The size of the LF gives the entity of bone destruction and the depth of the involvement of labyrinth structures; indicatively a fistula diameter less than 2 mm is unlikely to involve the endosteal membrane because the cholesteatoma matrix is still sustained by the edge of eroded bone¹⁵. Thus we adopted the limit of 2 mm size to categorize our patients and we noted an increased rate of postoperative nystagmus in those with more than 2 mm fistula size, all treated

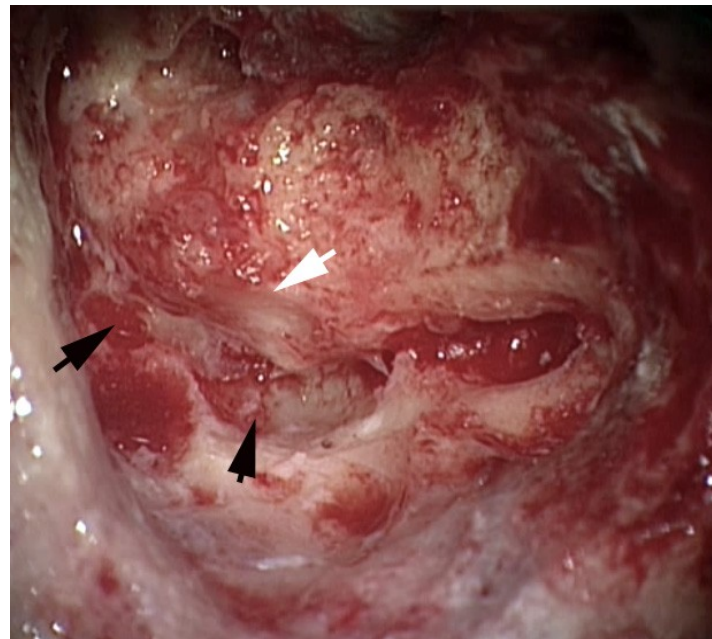


Figure 3 (A-B): Intraoperative findings of large fistula of lateral canal and posterior semicircular canal (black arrows); the facial nerve canal was eroded by cholesteatoma (white arrow).

by leaving the matrix in situ and with hearing preservation.

Hearing loss due to a LF is variable, and in cases of concurrent infection of labyrinth preservation of hearing is not realistic. Some studies reported that 12% to

30% of patients with LF were deaf on a preoperative hearing test ^{8,16}. Imaging with CT scan is important to localize the LF and to have cognition about extension and depth of the otic capsule resorption. In our case series all CT scan done had a

Labyrinthine Fistula characteristics of 8 cases										
Site	LSC	LSC	LSC	LSC	LSC	LSC	LSC +SSC	LSC +PSC	LSC	LSC
<i>Size (mm)</i>	< 2	> 2	> 2	< 2	< 2	> 2	> 2	> 2	> 2	< 2
<i>Fistula Sign</i>	Yes	Yes	No	No	No	No	No	No	No	Yes
<i>PTA Bone conduction Pre-op</i>	10	70	20	25	25	90	90	60	15	25
<i>PTA Bone conduction post-op</i>	10	70	45	25	25	90	90	60	15	25
<i>CT scan prediction</i>	Yes	Yes	Yes	Yes	Yes	Yes	Yes	Yes	Yes	Yes
<i>Matrix over the fistula</i>	Yes	Yes	Yes	No	No	No	No	No	Yes	Yes
<i>Post-operative Nystagmus</i>	Yes	Yes	No	No	No	No	No	No	Yes	No
<i>Cholesteatoma Recurrence</i>	Yes	No	No	No	No	No	No	No	No	No

Table 1. Characteristic of 8 patients managed for labyrinthine fistula. LSC: lateral semicircular canal; SSC: superior semicircular canal; PSC: posterior semicircular canal; PTA: pure tone audiometry; mm: millimeters.

good predictive value in identifying site and size.

The mastoidectomy technique adopted in management of LF in cholesteatoma can be either open or closed. Our surgical strategy agreed with those Authors reporting that only radical or modified radical mastoidectomy is indicated when a LF is present^{14,17,18}. A closed technique is adopted by other Authors depending on the contralateral hearing, the extent of the fistula, the size of the mastoid and the degree of sensorineural hearing loss in the affected ear^{19,20}.

The management of LF includes generally two approaches. The first advocate the complete removal of the cholesteatoma matrix over the LF with contextual repair of bone defect; the repair could be done in the same operation or in a second stage operation. The complete matrix removal eliminates a potential risk of recurrence or source of infection. The second approach is to leave a thin layer of the matrix over the fistula site. The rationale for this approach is supported by the thought that the removal of the cholesteatoma sac reduces the pressure effect to the bone resorption and the risks for labyrinth functions are reduced by not opening the inner ear.

The general address reported by Authors is the adoption of both techniques: advanced disease is treated more often with matrix over the fistula, limited cholesteatoma with LF are managed by matrix removal and fistula reconstruction. In case of preoperative profound hearing loss the matrix is generally removed, because no risks for hearing is presents. The hearing preservation after LF treatment is near to 85% with both techniques^{4,21}, although the interpretation of the data should be done critically considering the disease severity and the preoperative hearing. Postoperative deterioration of sensorineural hearing with complete removal of the cholesteatoma over the fistula varies between 0 and 66% in the literature^{9,22,23}, while it varies from 2.4 to 26% when the matrix is left in situ¹⁵⁻¹⁷. Intuitively, the opening of the labyrinth could lead to sensorineural hearing loss, although several Authors open the labyrinth intentionally with hearing pres-

ervation as in management of positional vertigo, far advanced otosclerosis and in neurotologic approaches with partial labyrinth destruction²⁴⁻²⁶. The removal of the matrix under great magnification and brightness, followed by with rapid repair is often adopted when the hearing preservation is presumable by limited disease and good preoperative hearing, although should be always considered that a number of dead ears have resulted also in selective treatments^{4,27}.

Conclusion

Labyrinthine fistula is a severe complication of extended cholesteatoma and lateral semicircular canal is frequently involved. CT scan is very important for diagnosis because there are no specific preoperative symptoms of labyrinthine fistula, although it has limits on accuracy of depth evaluation. The choice of surgical technique is suggested by preoperative hearing status, extension of cholesteatoma, size and site of the LF. Considering our results, and reports in literature, leaving or removing the cholesteatoma matrix over the fistula could warrant the hearing preservation. Although the recurrence seems not related to type of LF management, a canal wall down procedure is suggested to achieve complete cholesteatoma removal and wide field management of labyrinthine fistula.

References

1. Maranhao A, Andrade J, Godofredo V, Matos R, Penido N. Epidemiology of intratemporal complications of otitis media. *Int Arch Otorhinolaryngol* 2014; 18:178-183.
2. Greenberg JS, Manolidis S. High incidence of complications encountered in chronic otitis media surgery in a U.S. metropolitan public hospital. *Otolaryngol Head Neck Surg* 2001; 125:623-627.
3. Faramarzi AH, Heydari ST, Rusta M. The prevalence of labyrinthine fistula in chronic otitis media surgery in shiraz, southern iran. *Iran Red Crescent Med J* 2011; 13:582-585.
4. Copeland BJ, Buchman CA. Management of labyrinthine fistulae in chronic ear surgery. *Am J Otolaryngol* 2003; 24:51-60.

5. Portier F, Lescanne E, Racy E, Nowak C, Lamblin B, Bobin S. Studies of labyrinthine cholesteatoma related fistulas: report of 22 cases. *J Otolaryngol* 2005; 34:1-6.
6. Sanna M, Dispenza F, Flanagan S, De Stefano A, Falcioni M. Management of chronic otitis by middle ear obliteration with blind sac closure of the external auditory canal. *Otol Neurotol* 2008; 29:19-22.
7. Nylen CO. The labyrinthine fistula symptoms. *Acta Otolaryngol* 1923; 111: (suppl).
8. Sheehy JL, Brackmann DE. Cholesteatoma surgery—management of the labyrinthine fistula—a report of 97 cases. *Laryngoscope* 1979; 89:78-87.
9. Soda-Merhy A, Betancourt-Suarez MA. Surgical treatment of labyrinthine fistula caused by cholesteatoma. *Otolaryngol Head Neck Surg* 2000; 122:739-742.
10. Parisier SC, Edelstein DR, Han JC, Weiss MH. Management of labyrinthine fistulas caused by cholesteatoma. *Otolaryngol Head Neck Surg* 1991; 104:110-115.
11. Meyer A, Bouchetemplé P, Costentin B, Dehesdin D, Lerosey Y, Marie JP. Lateral semicircular canal fistula in cholesteatoma: diagnosis and management. *Eur Arch Otolaryngol*; DOI 10.1007/s00405-015-3775-6.
12. Jung J, Chole R. Bone resorption in chronic otitis media: the role of the osteoclast. *ORL J Otorhinolaryngol Relat Spec* 2002; 64:95-107.
13. Jang CH, Jo SY, Cho YB. Matrix removal of labyrinthine fistulae by non-suction technique with intraoperative dexamethasone injection. *Acta Otolaryngol (Stockh)* 2013; 133:910-915.
14. Palva T, Johanson LG. Preservation of hearing after removal of the membranous canal with cholesteatoma. *Arch Otolaryngol Head Neck Surg* 1986; 112:982-985.
15. Sanna M, Zini C, Gamolletti R, Taibah A, Russo A, Scandellari R. Closed versus open technique in the management of labyrinthine fistulae. *Am J Otol* 1988; 9:470-475.
16. Ritter FN. Chronic supportive otitis media and the pathologic fistula. *Laryngoscope* 1970; 80:1025-1035.
17. Gacek RR. The surgical management of labyrinthine fistulae in chronic otitis media with cholesteatoma. *Ann Otol Rhinol Laryngol* 1974; 83:11-19.
18. Abramson M, Harker LA, McCabe BF. Labyrinthine fistula complicating chronic suppurative otitis media. *Arch Otolaryngol* 1974; 100:141-142.
19. Hakuba N, Hato N, Shinomori Y, Sato H, Gyo K. Labyrinthine fistula as a late complication of middle ear surgery using the canal wall down technique. *Otol Neurotol* 2002; 23:832-835.
20. Law KP, Smyth GD, Kerr AG. Fistulae of the labyrinth treated by staged combined approach tympanoplasty. *J Laryngol Otol* 1975; 89:471-478.
21. Moon IS, Kwon MO, Park CY et al. Surgical management of labyrinthine fistula in chronic otitis media with cholesteatoma. *Auris Nasus Larynx* 2012; 39:261-264.
22. Magliulo G, Celebrini A, Cuiuli G, Parrotto D. Surgical management of the labyrinthine fistula complicating chronic otitis media with or without cholesteatoma. *J Otolaryngol Head Neck Surg* 2008; 37:143-147.
23. Quaranta N, Liuzzi C, Zizzi S, Dicorato A, Quaranta A. Surgical treatment of labyrinthine fistula in cholesteatoma surgery. *Otolaryngol Head Neck Surg* 2009; 140:406-411.
24. Parnes LS, Mc Clure JA. Posterior semicircular canal occlusion in the normal hearing ear. *Otolaryngol Head Neck Surg* 1991; 104:52-57.
25. Ars B, Claes J, Casselman J, Ars-Piret N. Preservation of cochlear function after extensive labyrinthine destruction. *Am J Otol* 1996; 17:40-45.
26. Sanna M, Dispenza F, Mathur N, De Stefano A, De Donato G. Otoneurolological management of petrous apex cholesterol granuloma. *Am J Otolaryngol* 2009; 30:407-414.
27. Anthony PF. Partitioning the labyrinth for benign paroxysmal positional vertigo: Clinical and histologic findings. *Am J Otol* 1993; 14:334-342.