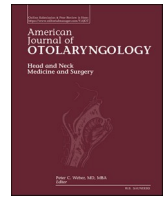




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Labyrinthine fistulas: Surgical outcomes and an additional diagnostic strategy

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ABSTRACT

Purpose: To evaluate perioperative findings and audiological and vestibular outcomes in patients operated for cholesteatoma with labyrinthine fistulas. Also to assess radiological fistula size.

Materials and methods: Patients who underwent surgery for a labyrinthine fistula caused by a cholesteatoma between 2015 and 2020 in a tertiary referral center were retrospectively investigated. Fistula size was determined on preoperative CT scan. Bone and air conduction pure tone average thresholds were obtained pre- and postoperatively. Clinical outcomes, such as vertigo and otorrhea were also evaluated. Main purpose was to determine whether there is a correlation between fistula size and postoperative hearing. Furthermore, perioperative findings and vestibular outcomes are evaluated.

Results: 21 patients (22 cases) with a labyrinthine fistula were included. There was no significant change after surgery in bone conduction pure tone average (preoperatively $27.6 \text{ dB} \pm 26.7$; postoperatively $30.3 \text{ dB} \pm 34.3$; $p = 0.628$) or air conduction pure tone average (preoperatively $58.7 \text{ dB} \pm 24.3$; postoperatively $60.2 \text{ dB} \pm 28.3$; $p = 0.816$). Fistula size was not correlated to postoperative hearing outcome. There were two patients with membranous labyrinth invasion: one patient was deaf preoperatively, the other acquired total sensorineural hearing loss after surgery.

Conclusions: Sensorineural hearing loss after cholesteatoma surgery with labyrinthine fistula is rare. Fistula size and postoperative hearing loss are not correlated, however, membranous labyrinthine invasion seems to be related to poor postoperative hearing outcomes. Therefore, additional preoperative radiological work up, by MRI scan, in selected cases is advocated to guide the surgeon to optimize preoperative counselling.

1. Introduction

One of the complications of chronic otitis media with cholesteatoma is a labyrinthine fistula. These are described by a bony defect or dehiscence of the otic capsule. The labyrinth consists of a cochlear and a vestibular part, which conceal respectively the hearing and equilibrium apparatus. Therefore, lesions to the otic capsule have serious impact on these two important structures for communication and balance function. Patients with labyrinthine fistulas present with a myriad of symptoms related to the aforementioned functions: sensorineural hearing loss, tinnitus, vertigo, dizziness or instability triggered by augmented

intracranial or middle ear pressure (Hennebert or fistula sign) or loud noise (Tullio phenomenon), hyperacusis, and oscillopsia [1–4]. Often, patients already have a history of different treatments to alleviate longstanding otorrhea before these complaints occur.

The incidence of an otic capsule fistula varies between 5 and 9% of all ear surgeries for chronic otitis media, mostly caused by cholesteatoma [1,3,5–7]. In most cases (80–90%), the lateral semicircular canal (LSCC) is involved [1–3,5,8]. However, damage to the posterior (PSCC) and superior (SSCC) canal as well as the cochlea is also described, sometimes with concomitant facial nerve weakness. Labyrinthine fistulas might serve as “porte d’entrée” to the intracranial space

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leading to meningitis, cerebritis or cerebral abscess. Therefore, early and adequate diagnosis with timely treatment is advocated.

The ultimate goal in patients with a fistula is to eradicate the cholesteatoma matrix and cover the fistula, and attaining a dry and safe ear, while hearing and vestibular function are preserved. The best treatment to achieve this is still a subject of debate and depends on the location and size of the fistula. Some surgeons perform canal wall down procedures leaving the matrix overlying the fistula to prevent damage to the labyrinth [9,10]. However, most recent studies advocate removing the cholesteatoma matrix entirely and subsequently closing the bony defect [3,7,11,12].

It seems plausible that a larger fistula can cause more hearing loss. Interestingly the clinical symptoms or burden patients experience does not seem to correlate with the size of the fistula. There might even be a negative correlation between the amount of labyrinthine damage, size of the fistula, and postoperative hearing preservation [13]. Depending on the extent of structural damage to the petrous bone and labyrinth, more or less aggressive surgical management is the appropriate treatment of choice.

We present a case series of patients with chronic otitis media with cholesteatoma and labyrinthine fistulas. We describe the preoperative symptoms and fistula size, perioperative findings, surgical techniques and the audiological outcome. Furthermore, current classifications are evaluated regarding the prognostic applicability and an additional preoperative diagnostic approach is proposed.

2. Materials and methods

A retrospective cohort study was performed in a tertiary referral center. The study did not need approval of the Medical Research Ethics Committee, since the Medical Research Involving Human Subjects Act (WMO) does not apply to it (MvdL/mb/21/500229). All surgeries, performed by two otologists (X and X) in our tertiary referral center between January 1, 2015 and December 31, 2020 were evaluated. Patients with a labyrinthine fistula, caused by cholesteatoma, were included. Patients below the age of 18 at the time of surgery were excluded. Pre- and postoperative hearing, location and size of the fistula on high resolution computed tomography (HRCT), iatrogenic injury of the membranous labyrinth, associated complications of surgery, clinical outcomes, including vertigo and dizziness, and occurrence of recurrent cholesteatoma were evaluated. Investigated parameters were sex, age, side, comorbidities, preoperative clinical signs, type of surgery, material used for fistula closure, preoperative associated complications of the fistula and/or cholesteatoma, duration of hospitalization, previous ear surgeries, type and dose of corticosteroids, and administration of antibiotics peri- and postoperatively. The recently published STAMCO classification was used to report the extent of the cholesteatoma [14].

2.1. Fistulas

All patients had a preoperative HRCT scan of the temporal bone using a Siemens-force CT scanner at 120 kV and 150mAs or a Philips scanner at 120 kV and 300mAs. The images were reconstructed in the axial and coronal plane with a slice thickness of 1.0 mm (mm). The extent and location of the fistulas were described by assessing the involvement of the inner ear structures: the three semicircular canals and cochlea. To assess inter-rater reliability, the fistula size was measured independently by two otologists (X and X) and one specialized neuroradiologist (X) in both the axial and coronal plane. The mean of the three measurements was calculated in each plane. The largest dimension was used to determine the size of the fistula. When a patient had two fistulas, the largest one was taken into account for the analysis.

2.2. Hearing outcome

Hearing was assessed preoperatively, as well as six weeks

postoperatively (also one year postoperatively if available). Bone conduction (BC) and air conduction (AC) thresholds were measured at pure tone average (PTA) 0.5, 1, 2, and 4 kHz.

2.3. Surgical technique

In most cases, the surgical technique was the canal wall up (CWU) tympanomastoidectomy, both with or without epitympanum and mastoid obliteration (BOT). Whether or not the obliteration technique was used, did not depend on any aspect of the fistula. In cases with previous canal wall down (CWD) surgery, the posterior canal wall was reconstructed followed by obliteration of the mastoid. In one case with a pre-existent deaf ear, a subtotal petrosectomy with abdominal fat obliteration was performed. In all cases, the cholesteatoma matrix was completely removed and the fistula was covered with different types of material.

2.4. Statistical analysis

For the comparison between pre- and postoperative hearing, paired samples *t*-tests were performed. Spearman's rho test was used to determine the correlation coefficient. SPSS 26.0 was used. A *p* value of less than 0.05 was regarded as statistically significant.

3. Results

3.1. Patient population

During the study period, 238 patients with chronic otitis media with cholesteatoma were treated in our hospital. 21 patients (9%) presented with a labyrinthine fistula. One patient had surgery for a fistula twice. The mean age at the time of surgery was 48 years [range 19–84]. An obliteration of the epitympanum and mastoid was performed in 14 ears

Table 1
Perioperative parameters and patient factors.

Total	22
Gender	
Female	5
Male	17
Side	
Left	11
Right	11
Surgical technique	
CWU tympanoplasty	8
CWU with obliteration	11
Mastoid obliteration with CW reconstruction	2
Subtotal petrosectomy	1
Surgery	
Primary	8
Revision	14
Associated complications of the cholesteatoma	
Tegmen destruction	9
Canal wall destruction	7
Postauricular abscess	2
Cerebrospinal fluid otorrea	2
Adhesive otitis	1
Facial palsy	1
Postoperative antibiotics	
Amoxicillin/clavulanic acid	14
Clindamycin	3
Ceftriaxon	1
Ciprofloxacin	1
Comorbidities	
Diabetes type 2	2
COPD GOLD I	1
Allergy to antibiotics	3
Atopy	2
Smoking	11

CW: canal wall, CWD: canal wall down, CWU: canal wall up.

(64%, Table 1). In 14 cases (64%) it was a revision surgery. Associated preoperative complications, caused by the cholesteatoma, can be seen in Table 1. All cholesteatomas were graded STAMCO stage III.

Antibiotic regimen was administered perioperative in all cases (cefazolin 2 g, intravenous). From 19 cases that received prophylactic antibiotic treatment during the postoperative course, 1 had a retroauricular wound infection after 8 days and 1 after 9 days. One patient, who did not receive prophylactic postoperative antibiotics, had a retroauricular wound infection 19 days postoperatively. The average duration of hospitalization was 1.18 days [range 0–4].

3.2. Hearing outcome

There was no significant change in BC PTA 6 weeks after surgery (preoperatively $27.6 \text{ dB} \pm 26.7$ versus postoperatively $30.3 \text{ dB} \pm 34.3$; $p = 0.628$). Also, there was no difference in AC PTA (preoperatively $58.7 \text{ dB} \pm 24.3$ versus postoperatively $60.2 \text{ dB} \pm 28.3$; $p = 0.816$).

Looking at individual cases in Fig. 1, three cases (14%) had a postoperative PTA BC deterioration of $>10 \text{ dB}$. Out of these three cases, two [#4 and #8] had a deterioration of $>10 \text{ dB}$ only at 4 kHz (their speech reception remained stable); one [#13] had only a temporarily deterioration $>10 \text{ dB}$, since there was no adequate masking possible during the audiogram due to temporarily conductive hearing loss on the contralateral ear. One patient acquired a deaf ear [#3]. One patient [#18] was preoperatively deaf in the affected ear.

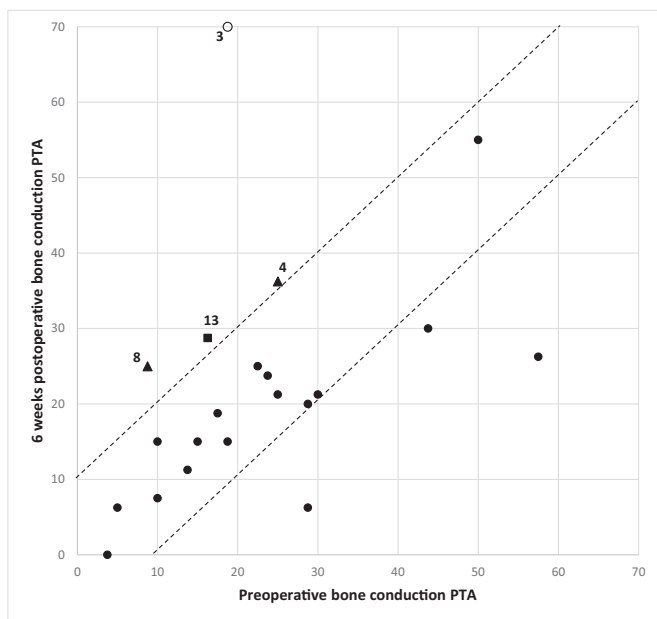


Fig. 1. Preoperative bone conduction pure tone average against 6 weeks postoperative bone conduction pure tone average.

Pure tone average (PTA): pure tone average is determined as the mean dB hearing level on 0.5, 1, 2, and 4 kHz.

The numbers refer to the case numbers from Table 3.

The triangles represent case numbers with postoperative deterioration of the bone conduction threshold $>10 \text{ dB}$ only at 4 kHz [#4 and #8].

The square represents the case [#13] with a temporarily deterioration of the bone conduction threshold $>10 \text{ dB}$ (the bone conduction could not be properly masked at 6 weeks postoperatively, because of temporarily conductive hearing loss on the contralateral side).

The open circle represents the case [#3] in which the patient acquired a deaf ear.

BC: bone conduction, PTA: pure tone average.

3.3. Invasion depth

Membranous invasion could not be detected on preoperative CT, but was encountered during surgery. In case 3 and 18, the cholesteatoma had grown into the membranous labyrinth. When removing these cholesteatomas during surgery, the membranous labyrinth was opened by the surgeon. In one case [#20], invasion and damage during surgery were doubtful; postoperative BC and AC PTA did not deteriorate in this case.

3.4. Fistulas

In 77% of the cases the LSCC was affected (Table 2). In four patients [#4, #10, #18 and #20], more than one inner ear structure was affected; one was already deaf preoperatively, in the other three both BC and AC PTA did not deteriorate $>10 \text{ dB}$ postoperatively. One patient had two fistulas in the cochlea. In the majority of the cases (13 out of 22 fistulas), the fistula was covered with fascia and bonedust. The radiological fistula size varied between 0.9 mm and 6.2 mm. The intraclass correlation coefficient (single measures) revealed a high agreement of the measurements between the three independent observers (0.96 for measurements in the axial plane; 0.85 in the coronal plane). Two fistulas could not be measured on CT (#10 in the SSCC, #18 in the PSSC), because they were too large. There was no correlation between fistula size and postoperative change (post- minus preoperative) in BC PTA ($\rho_s = 0.00$, $p = 1.00$) or AC PTA ($\rho_s = -0.24$, $p = 0.31$).

3.5. Clinical signs: pre- and postoperative

Preoperatively, 20 patients suffered from subjective hearing loss (Table 3). 14 patients encountered postoperatively persisting subjective hearing impairment, of whom five experienced a deterioration compared to preoperative. Two out of these five had unchanged hearing when objectively measured; one had a 9 dB deterioration in AC PTA, but not in BC PTA; in one both AC and BC PTA temporarily deteriorated [#13]; and one became deaf.

Before surgery, also vertigo ($n = 17$), otorrhea ($n = 11$), otalgia ($n = 9$) and tinnitus ($n = 7$) were common subjective complaints; every patient suffered from at least one of these clinical signs ($n = 22$). Three months postoperatively 50% ($n = 11$) did not have any of these complaints anymore. Five patients had ≥ 3 complaints before surgery: one with deteriorated BC PTA and four with stable or improved BC PTA. Two patients had persistent postoperative vertigo; they were not administered postoperative additional steroids. Three patients continued to have recurrent episodes of otorrhea.

3.6. Steroid treatment

During the operation, every patient was administered four milligrams dexamethasone. Six patients were also administered additional steroid treatment: one pre-, peri- and postoperative because of acute endangered hearing; five postoperatively because of postoperative vertigo (4/5 recovered within 4 days, 1 after 6 weeks).

4. Discussion

4.1. Hearing and structure preservation

A labyrinthine fistula can cause severe hearing loss and disabling complaints. The best approach to address a labyrinthine fistula has been part of debate for many years. In line with most literature, Fig. 1 shows that after carefully removing the entire cholesteatoma matrix from a labyrinthine fistula and covering the fistula, severe or profound hearing loss is rare [1,3,5,7,11,13]. Moreover, vestibular function is rarely affected. Only in one case [#3], a patient acquired a deaf ear. The cholesteatoma in this case had invaded the membranous labyrinth,

Table 2

Overview of surgical approach, size, location, hearing, material for covering, and follow-up time per fistula.

Case number	Surgical approach	Size per location (mm)				Hearing (dB)			Material for fistula closure	Follow up (years)
		LSCC	SSCC	PSCC	Cochlea	AC pre-operative	AC post-operative	ABG		
1	CWU	4.7				96	45	19	Tutoplast & bonedust	0.99
2	CWU	2.2				76	28	21	Fascia & bonedust	1.27
3	CWU with BOT	6.2				25	Deaf ear	Deaf ear	Bonewax	1.11
4a	CWU	2.4				50	59	16	Bonedust	1.00
4b			1.1							
5	CWU with BOT	1.2				50	51	45	Fascia & cartilage	0.27
6	CWU	1.7				70	63	48	Cartilage & perichondrium	1.50
7	CWU with BOT	1.5				55	40	25	Fascia & bonedust	1.11
8	CWU with BOT	3.5				53	65	40	Fascia	0.19
9	CWU with BOT	5.0				59	55	35	Fascia & bonedust	0.95
10a	CWU	1.7				39	45	21	Fascia & bonedust	0.67
10b			**							
11*	CWU	4.8				39	50	35	Fascia & bonedust	1.01
12*	CWU with BOT	1.6				51	35	24	Fascia & cartilage	1.61
13	CWU	1.4				44	61	33	Fascia & bonedust	0.76
14	CWU with BOT	2.6				26	34	26	Fascia & bonedust	0.16
15	CWU with BOT	1.2				60	60	35	Fascia & bonedust	0.38
16a	CWU				0.9	60	61	40	Fascia & bonedust	2.22
16b					2.3					
17	CWU with BOT	1.4				73	74	44	Fascia & bonedust	1.14
18a	Subtotal petrosectomy	2.0				Deaf ear	Deaf ear	Deaf ear	Bonewax	0.12
18b				**						
19	CWdown with BOT	2.6				95	109	54	Fascia & bonedust	1.00
20a	CWdown with BOT	2.0				43	36	36	Fascia & cartilage	1.13
20b				2.1						
21	CWU with BOT	4.9				43	50	31	Fascia & bonedust	0.11
22	CWU with BOT	3.1				55	43	21	Fascia & bonedust	0.18

Each case number represents a single fistula surgery. A and b are used to indicate multiple fistulas in one patient. Case 11 and 12, indicated by an asterisks (*), are one and the same patient, who had surgery for a fistula twice, at two different time points. The two cases indicated by two asterisks (**) had a bony defect of the labyrinth that was too large to measure. In case 10b the SSCC was concerned, in case 18b the PSCC.

ABG: air-bone gap; AC: air conduction, BOT: bony obliteration technique, CW: canal wall, CWU: canal wall up, dB: decibels, LSCC: lateral semicircular canal, mm: millimetres, PSCC: posterior semicircular canal, SSCC: superior semicircular canal.

Table 3

Clinical signs, counted pre- and postoperatively.

Clinical signs (n = 22)	Preoperative (%)	6 weeks postoperative (%)	3 months postoperative (%)
Subjective hearing loss	20 (91)	14 (64) ^a	14 (64) ^a
Vertigo	17 (77)	7 (32)	2 (9)
Otorrhea	11 (50)	4 (18)	3 (14)
Otalgia	9 (41)	2 (9)	1 (5)
Tinnitus	7 (32)	3 (14)	3 (14)
Hyperacusis	2 (9)	3 (14)	3 (14)
Loss of taste	1 (5)	0 (0)	0 (0)

^a 23% (n = 5) had subjective hearing loss compared to preoperative.

thereby destroying the inner ear structures, and the membranous labyrinth was opened during surgery. Another patient [#18] was already deaf before surgery, presumably after an episode of labyrinthitis. As in case 3, the cholesteatoma had grown into the membranous labyrinth. In all the other cases, the sensorineural hearing thresholds remained stable or improved. Two cases [#4 and 8] admittedly had a deterioration of their BC PTA, but this was only at 4 kHz, therefore not having clinical consequences. In fact, they had subjective hearing improvement. The other case [#13] with worse postoperative BC PTA could not be measured well six weeks after surgery, because of temporarily conductive hearing loss on the contralateral side, making it difficult to mask the affected ear properly. One year postoperative the BC PTA was measured again, showing no BC PTA deterioration of >10 dB. Profound sensorineural hearing loss caused by cholesteatomas that have grown into the membranous labyrinth has been observed in previous studies [3,7,11].

Also, extensive fistulas causing total deafness have been reported [1,13]. However, often no other option than removing the cholesteatoma is left, as leaving matrix to attempt to preserve hearing increases the risk of complications – such as labyrinthitis and intracranial complications – during follow up [13]. Furthermore, most patients were postoperatively free from any other complaints than hearing loss, being an extra argument to remove the entire cholesteatoma.

4.2. Radiological findings

The incidence of a labyrinthine fistula in patients treated for cholesteatoma in our tertiary referral center (9%), as well as the incidence of involvement of the LSCC (77%), concurs with the literature [1–3,5]. There was no correlation between the radiological fistula size and postoperative sensorineural hearing loss, which is in line with most previous studies [7,12,15,16]. Al Zaabi suggested a negative correlation between fistula size and postoperative sensorineural hearing loss; our study did not demonstrate that [13]. Fistula size was measured by using the images of a standard two-dimensional CT-scan so the used method will be clinically usable in general hospitals with a CT-scan. However, the labyrinth is a three-dimensional structure. In some cases it was impossible to perform an accurate fistula measurement, especially when the fistula was not located in the LSCC. This fact did not change even when reconstructions were made in the plane of the semicircular canal of interest. However, the extent to which the fistula has grown into the membranous labyrinth, rather than the radiological size, seems related to poor functional hearing postoperatively [#3 and #18] and therefore important for the prediction of clinical outcomes. In the patient with acquired deafness, deep invasiveness of the disease could have been

indicated by the massive bony destruction (>6 mm) visualized on the preoperative CT imaging. The patient with preoperative deafness had a PSCC fistula that was too large to measure. All the other measurable fistulas were ≤ 5.0 mm. Although our cohort did not include many invasive cases, it can be suggested that defects of the LSCC >5.0 mm or immeasurable defects on preoperative CT point towards invasion of the membranous labyrinth and thus an endangered labyrinth during the surgery. It is mentioned in previous studies that a membranous invasion could be suspected on the basis of radiological size, however, no clear consensus about the specific size exists. Stephenson reported a sensitivity of 66% and specificity of 71% for a membranous fistula when the

radiological size is 3.55 mm or more [5]. In Sone et al. most membranous fistulas were >3.0 mm on CT [16]. The article of Geerse described three cases with poor postoperative BC PTA in which the membranous labyrinth was inadvertently opened: two of them were >2 mm, one 1–2 mm, but they do not mention the exact size [7]. It also is not clear whether or not these three cholesteatomas had actually grown into the membranous labyrinth. Based on these studies and our own results we suggest that in cases with defects >3.5 mm membranous invasion is likely.

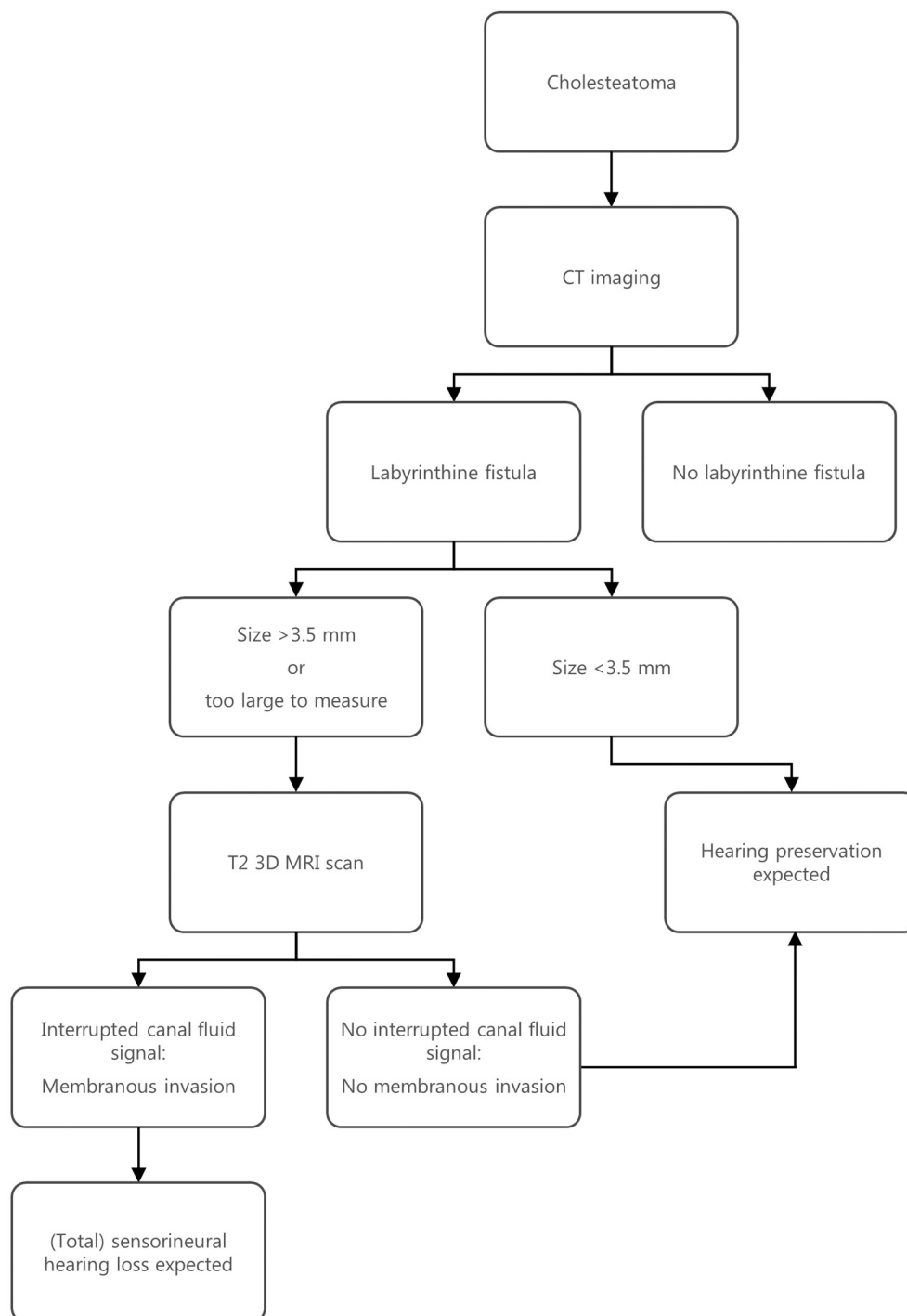


Fig. 2. Diagnostic flowchart: radiologic assessment.

CT: computed tomography, mm: millimetres, T2 3D MRI: T2 weighted three-dimensional magnetic resonance imaging.

4.3. Classifications and an additional strategy

Different classification systems have been published to predict odds for inner ear iatrogenic damage. Sanna uses three size categories: small (<1 mm), medium (1–2 mm) and large (>2 mm) [17]. Since our data, in line with most literature, did not show a correlation between fistula size and postoperative hearing, this classification does not have a clinical implication. Dornhoffer does not take into account the fistula size, but merely invasion depth [3]. Whether or not the cholesteatoma has invaded the membranous labyrinth is clinically important, however, it has to be determined preoperatively instead of during surgery to be useful for preoperative patient counselling. Since this cannot be conclusively demonstrated based on HRCT images, T2 3D magnetic resonance imaging (MRI) sequences (0.5 or 1 mm per coupe) can be used to visualize the possible invasiveness. Sone et al. previously described that patients with a higher signal intensity ratio on three-dimensional fluid-attenuated inversion recovery MRI tend to have higher chance of invasion of the membranous labyrinth (stage three fistula) [16]. Considering the costs, we do not recommend performing an MRI in addition to CT in every patient who will have surgery for cholesteatoma; we propose to make an additional MRI only when there is a risk for membranous labyrinth involvement (Fig. 2).

4.4. Hearing and vestibular symptoms

Slightly more than half of the patients suffered postoperatively from persisting hearing impairment, of which 23% ($n = 5$) experienced a subjective worsening compared to the preoperative situation. The subjective hearing did not always match with the objectively measured change in hearing thresholds. Therefore, in future research it would be valuable to focus on patient related outcome measures and disease specific quality of life questionnaires. Other common complaints in descending order were vertigo, tinnitus, otorrea, and otalgia. All patients had at least one of these complaints before surgery, but, apart from possible hearing impairment, most patients were postoperatively free from complaints. Only two patients had persistent vertigo; they did not use postoperative additional steroids. The vertigo seemed not related to fistula size. The other five with postoperative vertigo were administered with additional steroids and recovered within a week. Therefore, we again advocate complete removal of the cholesteatoma in each patient with a fistula and also strongly recommend treating postoperative vertigo with additional steroids.

4.5. Limitations and future perspectives

The relatively short follow up time of this study can be interpreted as a limitation. Not every patient had a one-year postoperative audiogram. However, the main outcome (iatrogenic damage causing sensorineural hearing loss) can be objectified after six weeks. Moreover, the hearing outcome six weeks postoperatively almost never differs from the outcome one year after surgery [18]. Prospective research is needed to investigate the possibilities of MRI scanning to predict the extent of labyrinthine destruction and assess its clinical relevance.

5. Conclusion

Subjective hearing loss and vertigo are the most common clinical signs indicating a labyrinthine fistula. We advocate complete removal of the cholesteatoma matrix and covering of the fistula, since iatrogenic sensorineural hearing loss is rare. No correlation between fistula size and postoperative hearing outcome was found, but membranous labyrinthine invasion seems to affect hearing. A preoperative additional MRI is advocated for fistulas with a size more than 3.5 mm to detect membranous invasion. This can guide the surgeon and acquire optimal patient informed consent.

Declaration of competing interest

None.

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Author contributions

All names authors meet the International Committee of Medical Journal Editors (ICMJE) criteria for authorship for this article, take responsibility for the integrity of the work as a whole, and have given their approval for this version to be published.

All authors contributed to the study conception and design. SW analyzed and interpreted the patient data and was a major contributor in writing the manuscript. HT and LS performed measurements of the CT images and were major contributors to the manuscript. JWD performed measurements of the CT images and also of the MRI scans. All authors read and approved the final manuscript.

Compliance with ethics guidelines

Approval by the Ethical Committee was not required for this study under Dutch Law (Mvdl/mb/21/500229). Written informed consent that allowed the use of their entire bodies for educational and research purposes was obtained during life from the persons that were used for this study.

Data availability

All data generated or analyzed during this study are available from the corresponding author on reasonable request.

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