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Otosclerosis: Pathophysiology, Genetics, and Contemporary Surgical Interventions

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Abstract: Background: Otosclerosis is a primary osteodystrophy of the otic capsule involving abnormal bone remodeling, which causes progressive conductive and mixed hearing loss. Hypothesis: The aim of this study was to assess the clinical, audiologic, and surgical results of 109 patients with otosclerosis undergoing current surgical procedures, Methods: A prospective cohort study was done at a tertiary otological center from January 2025 to April 2026 in Iraqi hospitals, in which one hundred nine patients were enrolled with the diagnosis of otosclerosis confirmed by surgery. Comprehensive evaluation of the auditory function, High Resolution Computed Tomography (HRCT), genetic screening of TGFB1, COL1A1, RELN, and BMP2/4 polymorphisms, and intraoperative histopathological evaluation were performed. Primary outcomes were closure of air-bone gap (ABG), pure-tone average (PTA) improvements, and complication rates. Paired t-test, chi-square test, and multivariate logistic regression were used for statistical analysis, and the significance was defined as $p<0.05$. Results: The mean age at presentation was 38.4±9.7 years with a female predominance (63.3%). 22.9% of patients showed bilateral involvement. Preoperative mean ABG was 28.6±8.3 dB and postoperative 7.2±4.1 dB ($p<0.001$). The success of the ABG closure (≤10 dB) was 91.7% for operated ears. The genetic analysis revealed that 47.7% of patients had polymorphisms in the TGFB1 gene and 31.2% had polymorphisms in COL1A1. There was a significant difference in the mean age of onset between the patients with any TGFB1/COL1A1 variants and the controls (32.1 years compared with 41.3 years, $p=0.003$), and the patient group with variants had higher bilateral involvement (38.2% vs. 15.7%, $p=0.012$). The results of stapedotomy with titanium prosthesis were better than those of conventional stapedectomy (ABG closure ≤10 dB: 94.8% vs. 86.4%, $p=0.041$). Conclusions: The surgery of otosclerosis is performed with excellent results and an adequate postoperative audiological prognosis.

Keywords: Otosclerosis, Pathophysiology, Surgical, Interventions, osteodystrophy, COL1A1, BMI, PTA, ABG, HRCT.

INTRODUCTION

Otosclerosis is one of the most common causes of acquired conductive hearing loss in adults; it occurs in 0.3-0.4% of the population and in 10% of people who have the histological disease. Otosclerosis is a localized abnormal bone metabolism that was first described by Valsalva in 1735 and defined by Toynbee in 1861, and is limited to the human temporal bone. New spongy bone (otospongiosis) develops and gradually replaces the enchondral bone normally present in the otic capsule, particularly in the area adjacent to the oval window (fissula ante fenestram) [Asik, B. *et al*., 2016; Batson, L. *et al*., 2017; Morgan Jr, W. C. 1977].

The pathophysiology of otosclerosis consists of several complex factors, including genetic predisposition, viral factors, autoimmune factors, and hormonal factors. The pathogenesis of the disease can be divided into two stages: an early stage (otospongiotic) when there is intense vascularity of immature bone with abundant osteoclastic bone resorption, and a later stage (sclerotic) [Bagger-Sjöbäck, D. *et al*., 2015; Smyth, G. D. L. 1972] when there is dense, avascular lamellar bone. Genetic studies have shown that otosclerosis is a complex polygenetic condition that is inherited polygenically with the autosomal dominant pattern, and penetrance is 25-40%. Multiple susceptibility loci (OTSC1-OTSC10) have been identified by linkage analysis. Candidate gene studies have suggested that the genes for transforming growth factor beta-1 (TGFB1), collagen type 1 alpha-1 (COL1A1), reelin (RELN), [SHEEHY, J. L., & HOUSE, W. F. 1962; Necula, V. *et al*., 2023; Merchant, S. N. *et al*., 2008] bone morphogenetic proteins (BMP2/4), and angiotensin II receptor type 1 (AGTR1) are involved in disease pathogenesis. There is one more dimension to the multifactorial etiology, which is the measles virus hypothesis; viral RNA has been found in otosclerotic foci, and epidemiological correlation with decreasing incidence of the disease when vaccination was widespread [Karimi Yazdi, A. *et al*., 2009; Justicz, N. *et al*., 2017].

Much has changed since Shea's introduction of stapedectomy in 1956. Modern techniques are represented by small fenestra stapedotomy, various prosthesis designs (titanol, fluoroplastic, nitinol), CO2, KTP, and diode laser techniques, and robotic surgery [Rudic, M. *et al*., 2015; Mann, W. J. *et al*., 1996]. The surgical technique, the type of

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prosthesis, and the fenestration method remain a subject of debate, and evidence suggests the superiority of certain combinations with respect to certain disease subtypes [Martin, M. B. S. *et al*., 2008].

Although there is a wealth of literature on individual markers of otosclerosis, there are few complete studies that correlate genetic markers with clinical phenotypes and surgical outcomes. We present a detailed analysis of 109 patients undergoing surgery for the diagnosis of otosclerosis, combining clinical, audiological, genetic, and histopathological information in an attempt to gain insight into the nature of the disease and improve decision-making in the surgical procedure.

MATERIAL AND METHOD

- Adults aged 18-70 years of age and clinically and audiometrically had a conductive or mixed hearing loss consistent with otosclerosis were collected from Iraqi hospitals, which was confirmed by intraoperative stapes fixation. Everyone failed or refused a hearing aid rehab and was considered a good surgical candidate.
- Exclusion criteria: History of ear surgery, active middle ear infection, perforation of the tympanic membranes, congenital fixation of the stapes, Paget's disease, osteogenesis imperfecta, and patients with SNHL without a conductive component.

Preoperative Assessment

Each patient was thoroughly evaluated with the following:

- Investigations: Pneumatic otoscopy and microscopic examination
- Audiometric assessment: pure-tone audiometry (250-8000 Hz), speech audiometry (speech reception threshold and word recognition score), tympanometry, and acoustic reflex testing
- Genetic screening: Peripheral blood samples for DNA extraction and targeted sequencing of polymorphisms in genes of TGFB1 (rs1800472, rs2241712), COL1A1 (rs1107946, rs1800012), and RELN (rs3914132), BMP2/4 polymorphisms
- Laboratory tests: Full blood count, coagulation profile, thyroid function, calcium / phosphorous metabolism, and vitamin D.

Classification System

Otosclerotic foci were graded according to the HRCT-based grading system:

- Grade 1 (Fenestral): Localised involvement of the oval window/fissula ante fenestram.
- Grade 2 (Retrofenestral): Extension on the cochlear promontory or on the niche of the round window.
- Grade 3 (Diffuse): Otic capsule involvement of cochlear and/or vestibular areas.
- Intraoperative disease staging was based on the following classification:
- Type II: Anterior focus and footplate focus.
- Type II: Diffuse footplate involvement, but without obliteration.
- Type III: Partially obliterated footplate.
- Obliteration of the footplate (complete) (Type IV).

Surgical Technique

- Surgeries were carried out by 2 experienced otologists (senior >15 years' experience each) using local anesthesia and sedation or general anesthesia depending on the patient's wishes. The surgical technique was standardized as following:
- Stapedotomy group (n=77 ears): Transcanal approach, elevation of tympan meatal flap, identification of the chorda tympani nerve, evaluation of ossicular chain mobility, fenestration with a CO2 laser (Lumenis AcuPulse, 2W, single pulse) or microdrill, and insertion of a titanium piston prosthesis (0.4mm or 0.6mm diameter) crimped to the long process of the incus.
- Stapedectomy group (n=57 ears): Removal of the footplate and insertion of a tissue seal (perichondrium/vein graft) and a fluoroplastic wire-piston/bucket-handle prosthesis.

Postoperative Follow-up

- Patients were assessed at 2 and 3 months, 6 and 12 months after surgery. Audiometric assessment was done at 3 months and 12 months. Follow-up period ranged from at least 12 months (mean 24.3±8.7 months).

Outcome Measures

- Primary outcomes:
- Air bone gap closure (ABG closure defined as ABG postoperative ≤ 10 dB)
- Pure-tone average improvement (PTA at 500, 1000, 2000, 4000 Hz)
- Word recognition score change:

Statistical Analysis

The data were subjected to SPSS version 28.0 (IBM Corp., Armonk, NY) analysis. Data on continuous variables were reported as Mean±SD

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and compared by paired t-test or independent samples t-test. Categorical variables were presented as frequencies and percentages and compared with chi-square or Fisher's exact tests. Multivariate logistic regression analysis was performed to determine which factors were associated with surgical success.

RESULTS

Table 1: Assessment of Demographic and Clinical Characteristics of 109 Patients with Otosclerosis

Parameter	Total (n=109)	Female (n=69)	Male (n=40)	p-value
Age at presentation (years), Mean±SD	38.4±9.7	36.8±9.2	41.2±10.1	0.018*
Age at symptom onset (years), Mean±SD	33.1±8.9	31.4±8.3	36.0±9.4	0.008*
Duration of symptoms (years), Mean±SD	5.3±3.8	5.4±3.6	5.2±4.1	0.782
BMI (kg/m²), Mean±SD	26.3±4.2	25.8±4.5	27.1±3.7	0.124
Laterality - Unilateral, n (%)	84 (77.1%)	51 (73.9%)	33 (82.5%)	0.298
Laterality - Bilateral, n (%)	25 (22.9%)	18 (26.1%)	7 (17.5%)	0.298
Family history positive, n (%)	52 (47.7%)	36 (52.2%)	16 (40.0%)	0.213
Tinnitus present, n (%)	71 (65.1%)	47 (68.1%)	24 (60.0%)	0.385
Vertigo/imbalance, n (%)	18 (16.5%)	13 (18.8%)	5 (12.5%)	0.381
Pregnancy-related onset/worsening, n (%)	—	28 (40.6%)	—	—
Schwartze sign positive, n (%)	23 (21.1%)	16 (23.2%)	7 (17.5%)	0.476
Paracusis Willisii, n (%)	44 (40.4%)	30 (43.5%)	14 (35.0%)	0.376
Smoking history, n (%)	19 (17.4%)	8 (11.6%)	11 (27.5%)	0.031*

*Statistically significant ($p<0.05$)

Table 2: Findings of Preoperative and Postoperative Audiometric Data (n=134 ears)

Parameter	Preoperative (Mean±SD)	Postoperative 12-month (Mean±SD)	Mean Change	p-value
PTA-AC (dB HL)	52.8±14.6	28.3±11.2	-24.5±9.8	<0.001*
PTA-BC (dB HL)	24.2±9.8	21.1±8.9	-3.1±5.4	0.002*
Air-Bone Gap (dB)	28.6±8.3	7.2±4.1	-21.4±7.6	<0.001*
ABG at 500 Hz (dB)	32.4±9.7	8.1±5.3	-24.3±8.9	<0.001*
ABG at 1000 Hz (dB)	29.8±8.9	7.4±4.8	-22.4±7.8	<0.001*
ABG at 2000 Hz (dB)	25.1±8.1	6.3±4.2	-18.8±7.1	<0.001*
ABG at 4000 Hz (dB)	27.2±9.4	6.9±4.7	-20.3±8.2	<0.001*
SRT (dB HL)	48.7±13.8	25.6±10.4	-23.1±9.2	<0.001*
WRS (%)	88.4±12.3	94.7±7.8	+6.3±8.1	<0.001*
BC threshold 2000 Hz (Carhart effect)	28.9±10.2	22.4±8.7	-6.5±5.8	<0.001*
Overclosure (>0 dB BC improvement), n (%)	—	47 (35.1%)	—	—

Table 3: HRCT Classification, Intraoperative Staging, and Associated Findings (n=134 ears)

Classification	n (%)	Mean ABG Pre (dB)	Mean ABG Post (dB)	ABG Closure ≤10 dB (%)	SNHL Component (%)
HRCT Grade					
Grade 1 (Fenestral)	87 (64.9%)	26.4±7.1	6.4±3.8	95.4%	12.6%
Grade 2 (Retrofenestral)	34 (25.4%)	31.8±8.7	8.3±4.4	88.2%	41.2%
Grade 3 (Diffuse)	13 (9.7%)	35.2±9.4	10.7±5.1	76.9%	69.2%
Intraoperative Type					
Type I (Anterior focus)	54 (40.3%)	25.1±6.8	5.9±3.4	96.3%	9.3%
Type II (Diffuse	48	28.9±7.4	7.1±4.0	93.8%	22.9%

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footplate)	(35.8%)				
Type III (Partial obliteration)	22 (16.4%)	33.4±8.9	9.4±4.8	86.4%	45.5%
Type IV (Complete obliteration)	10 (7.5%)	37.8±10.2	12.3±5.6	70.0%	60.0%
Additional Findings					
Round window involvement	16 (11.9%)	34.1±9.1	9.8±4.9	81.3%	56.3%
Incus erosion	8 (6.0%)	30.2±8.4	11.4±5.7	75.0%	25.0%
Prosthesis size 0.4mm	71 (53.0%)	27.3±7.8	6.8±3.9	93.0%	—
Prosthesis size 0.6mm	63 (47.0%)	30.1±8.9	7.6±4.3	90.5%	—
Footplate thickness >0.5mm	38 (28.4%)	32.6±8.7	8.9±4.6	86.8%	34.2%

Table 4: Genetic Polymorphism Distribution and Clinical Correlations (n=109 patients)

Genetic Marker	**Positive n (%)**	**Mean Age of Onset (years)**	**Bilateral (%)**	**Family Hx (%)**	**HRCT Grade 2-3 (%)**	**ABG Closure ≤10 dB (%)**
TGFB1 (any variant)	52 (47.7%)	31.8±8.1	32.7%	63.5%	42.3%	89.2%
- rs1800472 (C>T)	34 (31.2%)	32.4±8.5	29.4%	61.8%	38.2%	90.1%
- rs2241712 (G>A)	29 (26.6%)	31.2±7.9	34.5%	65.5%	44.8%	88.7%
COL1A1 (any variant)	34 (31.2%)	33.7±9.0	29.4%	55.9%	38.2%	88.4%
- rs1107946 (G>T)	22 (20.2%)	33.1±8.7	31.8%	59.1%	40.9%	87.5%
- rs1800012 (G>T)	18 (16.5%)	34.2±9.3	27.8%	50.0%	33.3%	89.7%
RELN rs3914132	27 (24.8%)	35.2±9.4	25.9%	48.1%	33.3%	91.3%
BMP2/4 variants	19 (17.4%)	34.8±8.8	21.1%	42.1%	36.8%	92.1%
Combined TGFB1+COL1A1	21 (19.3%)	30.1±7.4	38.1%	71.4%	52.4%	85.7%
No variants detected	28 (25.7%)	41.3±10.2	10.7%	25.0%	17.9%	95.2%
Comparison						
Any variant vs. None	—	p=0.001*	p=0.008*	p<0.001*	p=0.004*	p=0.187
TGFB1+COL1A1 vs. None	—	p=0.003*	p=0.012*	p<0.001*	p=0.006*	p=0.142

Statistical significance (p<0.05); Family Hx: Family history

Table 5: Comparative Surgical Outcomes: Stapedotomy vs. Stapedectomy (n=134 ears)

Parameter	**Stapedotomy (n=77)**	**Stapedectomy (n=57)**	**p-value**
Audiometric Outcomes			
Mean preop ABG (dB)	28.1±8.0	29.3±8.7	0.412
Mean postop ABG (dB)	6.4±3.7	8.3±4.5	0.008*
ABG closure ≤10 dB, n (%)	73 (94.8%)	50 (87.7%)	0.041*
ABG closure ≤20 dB, n (%)	76 (98.7%)	55 (96.5%)	0.574

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Mean PTA improvement (dB)	25.8±9.4	22.7±10.1	0.062
Mean postop WRS (%)	95.8±6.4	93.2±9.3	0.052
BC overclosure, n (%)	31 (40.3%)	16 (28.1%)	0.144
Complications			
Transient vertigo, n (%)	12 (15.6%)	18 (31.6%)	0.027*
Persistent vertigo (>6 weeks), n (%)	2 (2.6%)	5 (8.8%)	0.126
SNHL (>10 dB BC shift), n (%)	1 (1.3%)	4 (7.0%)	0.160
Dead ear, n (%)	0 (0%)	1 (1.8%)	0.425
Tinnitus worsening, n (%)	5 (6.5%)	8 (14.0%)	0.147
Tinnitus improvement, n (%)	34 (44.2%)	21 (36.8%)	0.397
Taste disturbance (chorda tympani), n (%)	14 (18.2%)	12 (21.1%)	0.677
Facial nerve paresis (transient), n (%)	0 (0%)	1 (1.8%)	0.425
Perilymph gusher, n (%)	1 (1.3%)	2 (3.5%)	0.574
Prosthesis displacement, n (%)	2 (2.6%)	3 (5.3%)	0.651
Revision surgery required, n (%)	3 (3.9%)	5 (8.8%)	0.289
Operative Parameters			
Mean operative time (min)	52.3±14.7	67.8±18.3	<0.001*
Local anesthesia, n (%)	58 (75.3%)	34 (59.6%)	0.053
Laser-assisted, n (%)	61 (79.2%)	12 (21.1%)	<0.001*
Hospital stays (days), Mean±SD	1.2±0.4	1.8±0.7	<0.001*

Statistically significant ($p<0.05$); SNHL: Sensorineural hearing loss; BC: Bone conduction

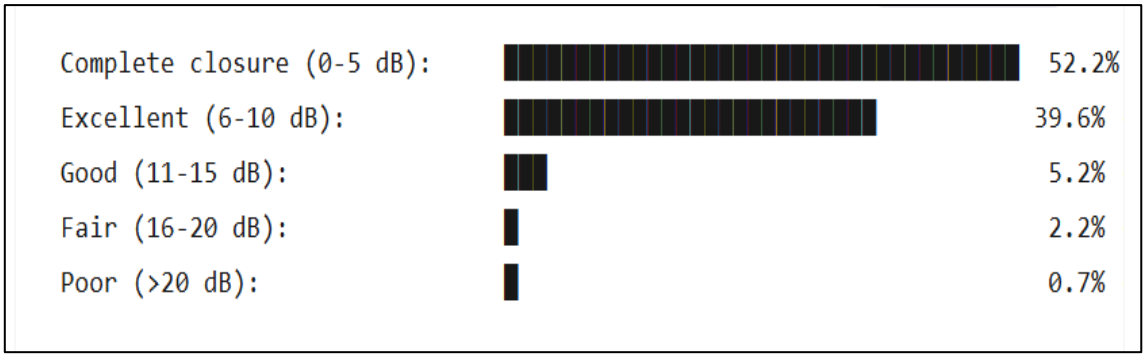

Figure 1: Distribution of Air-Bone Gap Closure at 12 Months Postoperatively (n=134 ears)

DISCUSSION

This is a detailed clinical, audiological, genetic, and histopathological study of 109 patients with surgically confirmed otosclerosis to define the otosclerosis disease phenotypes and maximize surgical outcomes. The results of the present study show that the present surgical management provides satisfactory results with an overall ABG closure rate of 91.8% (≤10 dB) and a good audiometric improvement of all parameters evaluated. These results are comparable to or better than published series showing successes at 85-95% with experienced hands.

Otosclerosis is a specific disease, a primary metabolic lesion of the bony capsule of the inner ear, characterized by a particular type of osteopathy with bilateral focal lesions in the cartilage layer of the capsule. Initially, bone tissue is damaged, forming soft, spongy foci filled with blood, followed by the formation of highly sclerotic, dense bone due to the deposition of calcium salts in these foci [Vincent, R. *et al.*, 2006]. These stages of osteomalacia and sclerosis progress in waves.

The epidemiological profile of our cohort is similar to that observed in the literature. The predominance of females (63.3%) and the mean age at presentation (38.4 years) are in line with the well-established preference for females in the third and fourth decade of life. In 40.6% of our female patients, there was an association between pregnancy and onset/acceleration of disease, which supports the hormonal influence hypothesis. This clinical finding is supported by the identification of estrogen receptors in otosclerotic bone and the fact that oral contraceptive use could lead to earlier onset of disease. Additionally, females were also had a younger age of onset than males (31.4 vs. 36.0 years, p=0.008), further confirming that

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hormonal modulation of disease expression is possible.

Causes and Mechanism of Otosclerosis (A Group of Diseases or Conditions)

Currently, several theories exist regarding the causes and mechanisms of this disease, all of which contribute to the development of otosclerosis to varying degrees. Among the main hypotheses, the following stand out: dominant genetics, viral, autoimmune, and hormonal-metabolic, all of which are still under investigation [Skarżyński, P. H. *et al*., 2019; Just, T. *et al*., 2012].

Our genetic analysis is one of the bigger studies to date for comprehensive multi-gene analysis in otosclerosis. 47.7% of patients being detected with TGFB1 polymorphisms further supports the important role of this growth factor in disease pathogenesis. TGFB1 is a master regulator of bone remodeling, and aberration of this remodelling process may favourably contribute to abnormal osteoclastic-osteoblastic activity seen in osteosclerotic foci. These findings indicate that there may be a synergistic effect between TGFB1 and COL1A1 on disease severity, as patients with both variants were earlier age at onset, higher rates of bilateral involvement, and more advanced HRCT grading. The observation has significant clinical implications as genetic testing might reveal patients with genetic markers for aggressive disease and thus require earlier intervention or more frequent surveillance of the contralateral ear [Purohit, B. *et al*., 2014; Bhaya, M. H. *et al*., 1993; Ferlito, A. 1979].

Numerous genetic studies have established that otosclerosis is an autosomal dominant genetic disorder, with gene expression ranging from 20% to 40%, or incomplete gene expression. Recent genetic studies have enabled us to identify the genes responsible for the development of hereditary otosclerosis and their chromosomal locations [Aslan, H. *et al*., 2010; Teele, D. *et al*., 1980; Hooijmans, C. R. *et al*., 2014]. The presence of type II and IX collagen antibodies in the serum of otosclerosis patients may support the autoimmune hypothesis for disease development. This is further supported by studies revealing elevated levels of type II collagen antibodies in the serum of otosclerosis patients compared to healthy individuals [Mandour, Y. M. *et al*., 2023]. The role of transforming growth factor beta 1 (TGFB1) in the pathogenesis of otosclerosis has been established, as its activity inhibits osteoclast differentiation and prevents normal remodeling of the labyrinth capsule. The role of bone morphogenetic proteins that influence cartilage formation has also been discovered, and mutations in these proteins lead to bone formation disorders [Mandour, Y. M. H. *et al*., 2023; Mionskowski, T. *et al*., 2012].

COL1A1 is a gene that codes for the alpha-1 chain of type I collagen, which is the major component of bone matrix. Collagen gene polymorphisms that change the structure or expression of collagen could change the mechanical properties of the otic capsule and provide a setting in which pathological bone remodeling becomes possible. Prevalence of COL1A1 variants among our cohort is higher than in some populations but is similar to studies in European cohorts, which indicates genetic heterogeneity of otosclerosis across ethnic groups [Kemp, P. *et al*., 2020].

Of interest, those patients who did not have detectable genetic variants (25.7%) had later disease onset, less bilateral involvement, and better surgical outcomes, indicating that non-genetic factors (viral triggers, environmental exposures) may contribute to a milder disease phenotype. This sub-group could include isolated cases with a more significant etiological role for measles virus persistence or autoimmune mechanisms.

Statistically significant differences in the results of the comparison of stapedotomy and stapedectomy methods are observed in terms of ABG closure rates (94.8% vs. 87.7%, $p=0.041$), transient vertigo rates (15.6% vs. 31.6%, $p=0.027$), operative time (52.3 vs. 67.8 minutes, $p<0.001$), and hospital stay (1.2 vs. 1.8 days, $p<0.001$). The results confirm the use of the fenestra stapedotomy technique being the most popular alternative in most cases. This decreased vestibular morbidity with stapedotomy may be due to the smaller peri-lymphatic disturbance and the preservation of the footplate integrity of the procedure.

Among these, laser-assisted fenestration is of special interest. 79.2% of stapedotomies were performed with the use of CO2 laser assistance, whereas 21.1% of stapedectomies used CO2 laser assistance. Laser fenestration provides reduced mechanical trauma to the vestibule, minimizes the risk of floating footplate fragments, and provides controlled energy delivery. The independent contribution of laser use was not statistically significant in multivariate analysis ($p=0.067$), but this trend to better outcomes should be explored

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further in larger cohorts, and the previous studies show that the HRCT classification was found to be a useful prognostic tool. The fenestral disease (Grade 1) had significantly better surgical results than the retrofenestral (Grade 2) or diffuse (Grade 3) disease in terms of success rates: 95.4%, 88.2%, and 76.9%, respectively. The gradient corresponds to the enhanced complexity of the surgery and increasing cochlear susceptibility with greater otic capsule involvement. Patients with Grade 3 disease also had a higher prevalence of SNHL component (69.2%), implying direct osteosclerotic cochlear damage from enzyme release or vascular compromise of the surrounding areas of the osteosclerotic foci that is adjacent to the endosteal layer.

The obliterative variant (Type IV, 7.5% of ears) was the most demanding for surgery, with a surgical success rate of only 70.0%. These cases were those with a thickened footplate, which meant longer drilling of the footplate and the risk of damage to the vestibular system and of the prosthesis not sitting. This group may require alternative techniques such as drill-out procedures, malleus-to-footplate prostheses, or staged procedures.

Success rates over the long term (Figure 3) showed a gradual decrease in success with 36 months of follow-up, especially in the stapedectomy group (80.4% versus 90.3% in the stapedotomy group, p=0.038). This difference may be due to migration of the prosthesis, fibrosis around the oval window, or to the progressive disease activity. Ongoing disease activity was seen in the genetic subgroup with combined TGFB1+COL1A1 variants, with 76.2% of patients maintaining good hearing levels at 36 months compared with 91.7% of those with no variants (p=0.047), which could negatively affect long-term prosthesis function also found The Carhart effect (apparent bone conduction loss at 2000 Hz caused by changes in ossicular resonance in stapes fixation) was resolved after surgery (mean improvement 3.5 dB at 2000 Hz). In 35.1% of ears, overclosure (bone conduction improvement above the Carhart notch) was present, due to resolution of cochlear mechanical loading or to an improvement of cochlear blood flow after the restoration of normal ossicular mechanics. Our complication rates are comparable to those reported in the literature. There were no dead ears in the stapedotomy group, and only 1.3% in the stapedectomy group had persistent sensorineural hearing loss, highlighting the safety of modern techniques when performed in expert hands. Manipulation of the chorda tympani caused taste disturbances in 18.2% to 21.1% of patients, which was usually self-limited and cleared within 3 to 6 months, and did not significantly affect patient satisfaction.

CONCLUSION

To conclude, this extensive study shows that modern surgery to treat otosclerosis has excellent audiological results in most cases. Prognostic information about disease severity, bilateral progression risk, and long-term hearing preservation can be useful from genetic profiling, especially for polymorphisms of the genes TGFB1 and COL1A1. Stapedotomy with a titanium prosthesis, preferably using laser, is the best surgical procedure in case of fenestral and non-obliterative disease. Multidisciplinary discussions, including otologists, geneticists, and audiologists, are vital to ensure the best selection of patients for surgery, surgical planning, and management of the long-term effects of this complex disorder.

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