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ORIGINAL ARTICLE

Surgical treatment in children with otosclerosis and congenital stapes fixation: our experience and outcome

Authors' Contribution:
A – Study Design
B – Data Collection
C – Statistical Analysis
D – Data Interpretation
E – Manuscript Preparation
F – Literature Search
G – Funds Collection

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ABSTRACT:

Aim: To evaluate outcomes of stapes surgery in children with congenital stapes fixation and juvenile otosclerosis.

Methods: A retrospective chart review was performed from 1987 to 2013 to identify patients under 18 years of age who underwent stapes surgery. Patient's age, gender, pre- and postoperative audiograms, intraoperative findings including etiology of stapes fixation, prosthesis type, and complications were analyzed.

Results: Eighteen children (6 - 17 years old), all with bilateral conductive hearing loss were identified and 34 stapes surgeries were performed (2 patients underwent surgery only on one side). Causes of fixation included juvenile otosclerosis in 88% and congenital stapes fixation in 12%. The mean preoperative air-bone gap (ABG) was 36.24 dB (SD: 10.86) compared to a postoperative mean ABG of 7.74 (SD: 3.3) ($p < 0.000$). Profound sensorineural hearing loss was not observed in long-term follow-up.

Conclusions: Pediatric stapes surgery has comparable results to stapedectomy in adults regardless of the cause of stapes fixation; however, the better hearing outcome was observed for cases of juvenile otosclerosis rather than congenital stapes fixation.

KEYWORDS: stapedotomy, stapes surgery, congenital stapes fixation, congenital stapes ankylosis, juvenile otosclerosis, children, pediatric stapes surgery

INTRODUCTION

Otosclerosis, described for the first time by Adam Politzer is defined as osteodystrophic changes localized in the bony labyrinth and footplate of the stapes [1]. Specific only for humans, it may hinder the function of hearing or balance, depending on the site, size, and histologic features of the pathologically involved area.

Abnormal bone remodeling, which includes bone resorption, new bone deposition, and vascular proliferation in the temporal bone are pathologic manifestations of the disease [1]. Otosclerotic plates are mainly localized anterior to the oval window (fissula ante fenestram region), and on the stapes footplate (80%), at the round window (30%), pericochlear region (21%) and the anterior part of the internal auditory canal (19%) [2].

Clinical presentation usually begins in early adulthood and is observed bilaterally. Otosclerosis has a symmetrical tendency and begins at the fissula ante fenestram in 70-90% of cases. Ethology of otosclerosis remains unclear, but family predisposition of autosomal dominant inheritance with incomplete penetrance and variable expressivity.

Prevalence of histologic otosclerosis is significantly higher than the clinical manifestation of the disease, indicated as 0.3% to 0.4% of the population [3]. Guild et al. reported findings of otosclerosis in 0.6% of temporal bones in children under 5, and in 4% of children between 5 and 18 years [4]. In the more recent study, 2.5% of the 236 temporal bones (or 3.4% of patients) have revealed otosclerotic foci [5]. The result is much lower than the previously published figures on histologic otosclerosis but correlates well with clinical studies of otosclerotic families.

Stapes surgery for improvement of conductive hearing loss is a standard procedure in adults. Nevertheless, stapes surgery in children is less accepted, mainly because of the risk of postoperative sensorineural hearing loss (SNHL); however, incidence in children cannot be overlooked.

Differential diagnosis in children with conductive hearing loss with normal tympanic membrane and aerated tympanic cavity include juvenile otosclerosis, congenital stapes fixation, anomalies of the ossicular chain, osteogenesis imperfecta, round window atresia [6].

Congenital stapes fixation can be distinguished from juvenile otosclerosis by the progressive character of the latter. Therefore, information about the non-progressive character of deafness constitutes an essential element of diagnosis. Consequently, it is reasonable to allow a few years of follow up (about three years) and for the following audiograms to be performed.

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The purposes of investigation were to evaluate the results of stapes surgery in children who underwent the procedure for stapes fixation and to analyze postoperative audiologic results.

MATERIAL AND METHODS

Participants

The study constitutes a retrospective review of 18 cases of children (6 - 17 years old) who underwent surgical treatment because of juvenile otosclerosis or congenital stapes fixation. Data include patients' age when surgery was performed, side affected, surgical technique, intra-operative findings pre- and postoperative air--bone gap (ABG). Material was collected between 1987 and 2013 based on available data in a tertiary pediatric otolaryngology department in Barcelona. All of the participants suffered from bilateral conductive hearing loss. For that reason, 34 stapes surgeries were performed by the same senior, most experienced surgeon. In 2 cases patients underwent the procedure on only one side. Children with congenital malformations of the ossicular chain, different than stapes fixation were excluded from the study.

One patient from the study group (F, 8 years old), three months after the primary procedure developed stenosis of the external auditory canal on the right side and had to undergo canaloplasty. The middle ear was not opened during the second intervention. When operating on the left side, the surgeon performed large canaloplasty first and stapedotomy, no adverse symptoms were present at follow-up. Two additional cases of juvenile otosclerosis with perilymphatic gusher (Gusher syndrome) diagnosed from CT imaging were operated. Cerebrospinal fluid leakage was controlled with oval window closure with temporal fascia graft, and titanium prosthesis was fixed over the graft. Air-bone gap (ABG) remained the same after the procedure, no symptoms of sensorineural hearing loss were present in follow-up. However, these cases were excluded from this study.

Pure tone audiometry results were collected from all cases. Primary audiometry performed the day before surgery and the second audiometry performed after six months of follow-up were analysed in the study. ABG was calculated pre- and postoperatively. Follow-up was scheduled at the office after a week, one month, six months and one year. Ideal follow-up was planned for 36 months after the procedure.

Surgical technique

All surgeries were performed by the same, senior, most experienced surgeon in a tertiary referral center. Modified Fisch-incision technique (reversal steps stapedotomy) was applied [7]. Transcanal approach was used in every case. In one case, canaloplasty was made prior to the stapedotomy, as was mentioned before. Stapes movement was evaluated by delicate palpation of the stapes head with a curved needle after incudostapedial joint separation. Otosclerosis was confirmed intraoperatively by the presence of sings of white otosclerotic foci. Stapedotomy was performed in 23 cases (68%) with a microdrill and 11 cases (32%) using a CO_2 laser (Sharplan CO_2 Laser System).

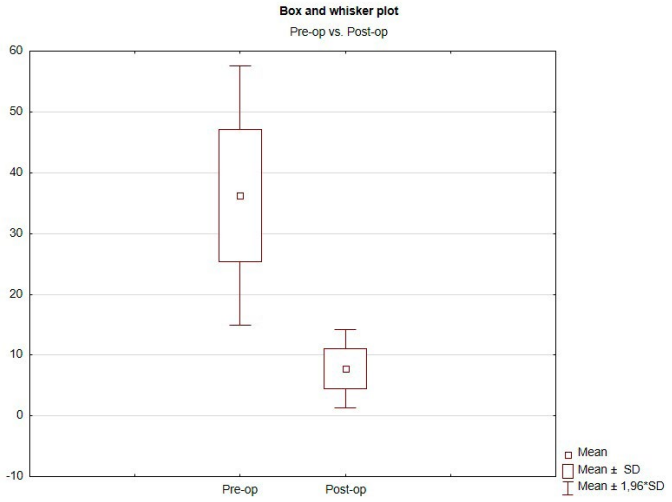

Fig. 1. Results of paired sample t-test.

In all 34 cases, a 0.4-mm-diameter Teflon piston was used. The oval window was sealed with Gel foam and blood clots in every case.

Intraoperative findings were prospectively reported by the surgeon. The senior surgeon divided the cases into three groups: obliterating otosclerosis (OF), anterior otosclerosis foci (AFP) or absence of annulus (AA, characteristic for congenital stapes fixation).

Audiometric evaluation

Four-frequency (0.5, 1, 2, and 4 kHz) pure-tone audiograms were used to estimate hearing using AC and BC thresholds. The successful result of stapes surgery was stated when the closure of the air-bone gap was 10 dB or less, as well as in air-bone gap closure to 20 dB or less.

These ABG closure levels are generally considered as a successful outcome of stapes surgery in the literature [8]. AC and BC thresholds measured six months after time surgery were used for calculation of the air-bone gap closure.

Pure-tone audiometry was performed according to the protocol recommended by the Spanish Society of Audiology, which does not differ significantly from the guidelines of the British Society of Audiology [3]. Testing was performed by the same, certified audiometric technician. The air-bone gap was calculated using AC and BC thresholds at 0.5, 1, 2 and 4 kHz, and the result was divided by 4 to receive a mean value from 4 different frequencies.

Statistical analysis

Data was collected in an Excel sheet and implemented to the Statistica 13.1 (Statsoft) software for statistical analysis. Statistical significance was reported at the alpha level of 0.05 (α=0.05). A p-value below 0.05 was considered significant ($p<0.05$). Continuous variables were shown as means with standard deviations (SDs), and categorical data were presented as percentage values. Pre- and postoperative air-bone gaps (ABG) were analyzed using a paired sample t-test.

RESULTS

Research included 18 children: 5 boys and 13 girls. The patient's mean age at the moment of surgery was 11.17 years (median: 11y, standard deviation ± 2.90). In every case, both ears were affected. However, in two cases only one side was operated. CT imaging showed signs of otosclerosis in 91% of cases.

SURGICAL FINDINGS

Among 34 procedures, 18 were performed on the right ear (53%) and 16 on the left ear (47%). Intraoperative findings show: anterior fixation in one-third of stapes footplates identified in 41% of cases (AFP), involvement of entire footplate in 47% (OF), and absence of annulus (AA) which is characteristic for congenital stapes fixation in 12%. Furthermore, any other ossicular chains anomalies were not identified.

The classical technique which incorporated a microdrill for stapedotomy was performed in 68% of cases. Laser stapedotomy was applied in 32% of interventions. The decision to use a CO_2 Laser was based on the availability of equipment and surgeon preferences which were influenced by the patient's unique anatomy.

Pure tone audiometry evaluation

Tab. I. Pure tone audiometry results: air bone gap calculation: preoperatively (1 day before surgery) and postoperatively (6-month follow-up).

	MEAN (DB)	+CI	-CI	MEDIAN	SD
Pre-Op	36.24	32.45	40.03	17.5	10.86
Post-Op (6 month)	7.74	6.59	8.9	7.3	3.3

dB- Decibel's, CI- confidence intervals, SD- standard deviation

Statistical analysis

Tab. II. The result of paired-sample t-test.

	MEAN	SD	N	σ^	T	p
Pre-op	36.24	10.86				
Post-op	7.74	3.30	34	11.46	14.5	0.000

SD- standard deviation, N- sample size, σ^- sample standard deviation of differences, t- t-statistic (t-test statistic) for paired sample t-test, p- p-value (probability value) for t-statistic

DISCUSSION

Conductive hearing impairment with an intact tympanic membrane and no history of chronic ear disease in children is an uncommon case of hearing loss and can be caused by multiple conditions including congenital stapes fixation, juvenile otosclerosis, tympanosclerosis, lateral ossicular chain fixation, or inner ear abnormalities such as semicircular canal dehiscence or an enlarged vestibular aqueduct. The most common diseases among those listed are congenital stapes fixation (CSF) and juvenile otosclerosis (JO). Progression of conductive hearing loss and a positive family history are the essential clinical grounds to differentiate between CSF and JO. Congenital stapes fixation and juvenile otosclerosis are potentially correctable with surgical intervention. However, childhood surgical treatments remain controversial.

The options for managing the pediatric stapes fixation include observation, implementation of hearing aid, bone anchored hearing aid, or stapedectomy. Before a decision about surgery, a number of factors should be considered: monoaural -binaural character of the disease, child's age, degree of hearing impairment, cochlear involvement, concomitant otitis media, Eustachian tube function, child's social and academic performance and level of speech development. We do not recommend the surgery before the age of 5 due to a high risk of otitis media and Eustachian tube dysfunction. In unilateral cases, surgery should be postponed till adulthood [9].

The presented research aimed to evaluate cases of children who underwent stapes surgery. The patients' mean age was 11 years. Juvenile otosclerosis was discovered in 88% of cases; congenital stapes fixation involved 12% of ears. Obliterative otosclerosis was seen in 47% of cases. CT scanning was useful to confirm signs of otosclerosis. Statistical analysis showed significant differences in hearing results between the pre- and postoperative group.

Congenital stapes fixation in comparison to juvenile otosclerosis has an earlier onset of hearing loss (3 vs. 10 years), a higher incidence of other malformations of ossicular chain (25% vs. 3%) and a greater air-bone gap after stapedotomy [9].

Juvenile otosclerosis is characterized by a higher incidence of a positive family history of deafness (53% vs 10%) and has better postoperative results (85% vs. 53% with a postoperative air-bone gap ≤ 10 dB) [8]. In a most recent meta-analysis of surgery success rate in juvenile otosclerosis, and congenital stapes fixation 72.1% of ears were improved to the desired ABG (10 dB) after surgery [8]. Surgery-related hearing loss was reported for JO in 3.4% of cases and for CSF in 2.1% [8].

To increase safety of surgical treatment, we would like to highlight a method of "reversal steps stapedotomy" proposed by Fisch which includes perforation of the footplate when the ossicular chain is still intact, followed by immediate placement and stabilisation of the prosthesis and, lastly, removal of the stapes superstructure to increase safety of the procedure. The technique reduces risk of "floating footplate" and complication related to dislocation of the incus. In literature, many research showed that the application of less traumatic surgical techniques such as stapedotomy has not decreased the incidence of these complications [10]. Fisch's reversal steps stapedotomy was proven to lower the incus and footplate difficulties [7][11]. That is why this method is our technique of choice in pediatric stapes fixation cases.

Several studies reported higher prevalence of an obliterative type otosclerosis in juvenile otosclerosis, which makes the surgery more challenging [12]. In our study, signs of obliterative otosclerosis were seen in 47% of cases which is confirmed in previous reports.

Computed tomography imaging may be used for diagnosis and to guide treatment. Imaging allows diagnosing alternative pathologies in patients with similar audiological findings to those of otosclero-

sis, such as ossicular discontinuity, ossicular fixation or labyrinthine anomalies. Research conducted by Dudau stated that a specific CT diagnosis of otosclerosis or alternative clinically relevant pathology was obtained in 63% of cases and 24% underwent targeted surgery. CT made a broader contribution by detecting important extensions of otosclerosis which impact the surgical risks and outcomes, whilst also predicting audiometric findings [13]. CT provides additional information by detecting various anatomical variants (e.g., tympanic facial nerve overhanging, dehiscent facial nerve canal, narrow oval window niche, and persistent stapedial artery), which can influence on the surgical approach to a stapedectomy [13][14]. In all pediatric cases with conductive hearing loss, CT imaging is necessary before surgical exploration to reduce the risk of a stapes gusher and sensorineural hearing loss [15]. Nowadays, the perilymph gusher is considered to be the direct result of abnormal communication between the perilymphatic system and the subarachnoid space through the internal auditory canal [16]. In research concerning temporal bone computed tomography (CT) scanning, Talbot and Wilson emphasized the fact that most cases of perilymph gushers are not correlated with an abnormally patent cochlear aqueduct. Common CT findings were an enlarged IAC, cochlear hypoplasia, absent bony modiolus, abnormal vestibular aqueduct, and enlarged labyrinthine facial nerve canal[15]. We recommend to always consider CT imaging because it allows to exclude the cause for severe complications; furthermore, when confirming otosclerosis, it also provides information about the otosclerotic foci and the presence of anatomical variants which can potentially impact on the surgical options, difficulties, and outcomes. Numerous studies have shown excellent results after stapes surgery. House et al. were one of the first to describe hearing results after stapedectomy in children. The results seem to be good as hearing improvement is seen after adult stapedectomy [17]. In a recent literature review, the pediatric stapes surgery was recommended as an effective and relatively low-risk option for carefully selected children with bilateral CSF or JO, who are at least 5 years old, free of otitis media, have a speech reception threshold >35 dB, and an ABG >30 dB [18][19]. In a recent study, significant prognostic factors for postoperative hearing outcome in otosclerosis patients were preoperative ABG, preoperative air conduction threshold, cochlear otosclerosis, male sex, laterality and age [20]. Notably, preoperative ABG was a significant prognostic factor at multiple frequencies. Authors assumed that patients with larger preoperative ABG have some other otosclerotic lesions outside the oval window niche [20]. We would like to highlight the role of the child's conscious participation in treatment. We encourage older children to engage in the decision-making process. Usually, children over the age of 14 are asked to sign informed consent with their parents. From our experience, it also occurs that with thorough patient's qualification, stapes surgery in children can give excellent results with a high level of safety.

CONCLUSIONS

Stapedotomy for juvenile otosclerosis is safe and efficient. A worse hearing outcome and higher risk of SNHL were observed for congenital stapes fixation. Hearing aid should be considered for the youngest children. The decision about surgery should be based on the character of the disease, child's age, degree of hearing impairment, cochlear involvement, Eustachian tube function, child's social and academic performance and level of speech development. In unilateral cases, surgery should be postponed till adulthood.

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