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REVIEW ARTICLE

Otosclerosis in children and adolescents: A clinical and CT-scan survey with review of the literature

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Summary

Objective: To assess the role of the CT-scan in the preoperative evaluation of juvenile otosclerosis and to study additional outcome data.
Design and setting: We performed a retrospective case series study from an academic referral hospital using data from 1992 to 2005.
Patients and methods: We selected patients younger than 18-year-old who had undergone primary stapedectomy for otosclerosis among the 10 stapedectomies performed over the study period and analyzed the patients' systematic pre- and post-operative audiograms and CT-scan findings.
Results: For this survey, complete data was available for 7 children, totaling 10 primary stapedectomies for otosclerosis. Their ages at diagnosis ranged from 10 to 17 years. In 4 children, CT-scan demonstrated bilateral findings typical of otosclerosis: poorly calcified foci near the fissula ante fenestram, associated with a hypodense edging surrounding the labyrinthine capsule in 2 children. The youngest patient had no CT-scan abnormalities. Stapedectomy was performed in one case and laser stapedotomy in 9 cases. Seven children were immediately improved following surgery and no postoperative facial palsy or prolonged vertigo was reported. The mean (S.D.) postoperative ABG was 6.5 dB (±3.7). The mean closure was 19 dB (±11.2). The mean change in high-tone bone conduction level was 1.8 dB (±7.5). Six children had a postoperative ABG less than 10 dB while in one, the ABG was inferior to 20 dB.
Conclusion: Preoperative CT-scan is useful for the preoperative diagnosis of otosclerosis in children. The images seen must be distinguished from other footplate

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pathologies or deformities of the ossicular chain which are often associated with poorer surgical results. This survey provides additional evidence that stapes surgery is an effective procedure for treating juvenile otosclerosis.

Contents

1. Introduction

Otosclerosis in childhood and adolescence or juvenile otosclerosis is rarely reported. Its histologic demonstration by Guild [1] dates from the beginning of the 20th century. The conductive hearing loss (CHL) is caused by stapes ankylosis and is associated with normal tympanic membranes at otoscopy. In this group age, the diagnosis is usually suggested by a positive family history of otosclerosis but must be distinguished from more frequent etiologies of stapes ankylosis with a normal tympanic membrane such as tympanosclerosis of the oval window or minor aplasia. Consequently, a thorough work-up including a search for cranio-facial abnormalities [2], examination of the pinna, the external auditory meatus and the tympanic membrane should be performed in order to eliminate major atresia and middle ear effusion, tympanosclerosis within the tympanic membrane, perforation, retraction or cholesteatoma which are frequently associated with CHL in this age group.

Despite a careful and thorough clinical examination, unexplained CHL requires appropriate imagery studies of the ear. Imagery plays an important role in the etiologic diagnosis of deafness in children and in its subsequent clinical management. CT-scan is the accepted gold standard [2,3] for assessing the status of the oval window, the inner ear, the IAC, the vestibular and cochlear aqueducts, the ossicles and the course of the facial nerve.

To our knowledge, there is no published data on the radiological findings in juvenile otosclerosis and the potential usefulness of imagery studies for establishing the diagnostic before surgery in this group age. This is important because clinical series on stapes surgery in children report best results when otosclerosis is present compared with other etiologies of stapes ankylosis [4]. This may explain the ongoing controversy concerning the optimum management of CHL: hearing aid or surgery? Consequently, we decided to assess the usefulness of the CT-scan as an imaging tool in the diagnosis of juvenile otosclerosis and provide additional outcome data in stapes surgery in children by performing a retrospective review of stapedectomy for otosclerosis during a 13-year period.

2. Patients and methods

We reviewed the medical records of all patients who had undergone stapedectomy for otosclerosis at the *CHRU de Tours* (a tertiary referral center in Tours, France) between 1992 and 2005. During this time period, 690 procedures were performed including 42 stapedectomies and 648 stapedotomies. Only 1 surgeon (EL) was involved in the pediatric procedures that concerned 7 subjects younger than 18 years. The diagnosis of otosclerosis was based on surgical criteria: stapes fixation and visualization of a macroscopic focus of otosclerosis (Fig. 1) in the absence of other external or middle ear abnormalities. None of the enrolled patients had cranio-facial abnormalities. Six of the 7 children had undergone a CT-scan prior surgery. Eight surgical procedures were performed under general anesthesia and 2 under local anesthesia, via an endaural incision. The technique used for footplate opening was different according to the time-period studied: stapedectomy was performed during the early study period while carbon dioxide laser stapedotomy was performed during the later (Fig. 1).

Each patient's chart was reviewed to determine the subject's gender, age at presentation, familial history and main complaints. In addition, we gathered surgical and CT-scan data concerning malformations in the otic capsule or abnormalities of the ossicles and round or oval windows.

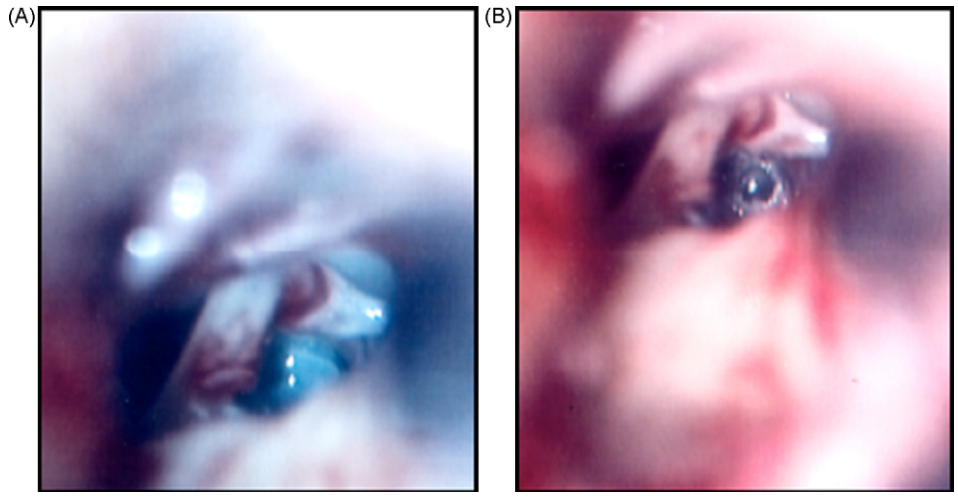

Fig. 1 Photograph during a surgical procedure for a right juvenile otosclerosis of a 9-year-old girl (Patient #6). (A) The otosclerotic foci invade the entire footplate. (B) A 0.6 mm stapedotomy opening has been vaporized into the center of the footplate.

We based our results on a comparison of the preoperative and postoperative audiometric data (using 1995 AAO-HNS Committee on hearing guidelines) [5] and otology symptoms.

3. Results

A total of 6 girls (9 of the ears operated on) and one boy were studied. Bilateral CHL was observed in 4 patients, although only 3 of them had bilateral surgery. Only one child had a family history of otosclerosis. In all the cases, deafness was the chief presenting complaint. Tinnitus was noted in 3 cases and dizziness in one case.

Among the children who had a CT-scan prior to surgery, 4 showed typical bilateral radiographic evidence of otosclerosis, namely poorly calcified foci near the *fissula ante fenestram* (Fig. 2). In 2 children, these lesions were associated with a hypodense edging surrounding the labyrinthine capsule (Fig. 2). There were no CT-scan abnormalities discovered in the 2 other patients. In addition, CT-scan failed to demonstrate any abnormalities in the malleus or incus or any potential risk for a peri-lymphatic gusher during footplate opening. Tympanotomy revealed a thin, blue, fixed footplate with a white anterior focus in 2 cases and diffuse otosclerosis in the remaining cases. No malformation or fixation of the malleus and/or the incus or abnormalities of the windows were encountered. Clinical and CT-scan data are summarized in Table 1.

All the patients showed immediate improvement after surgery and no facial palsy or prolonged vertigo

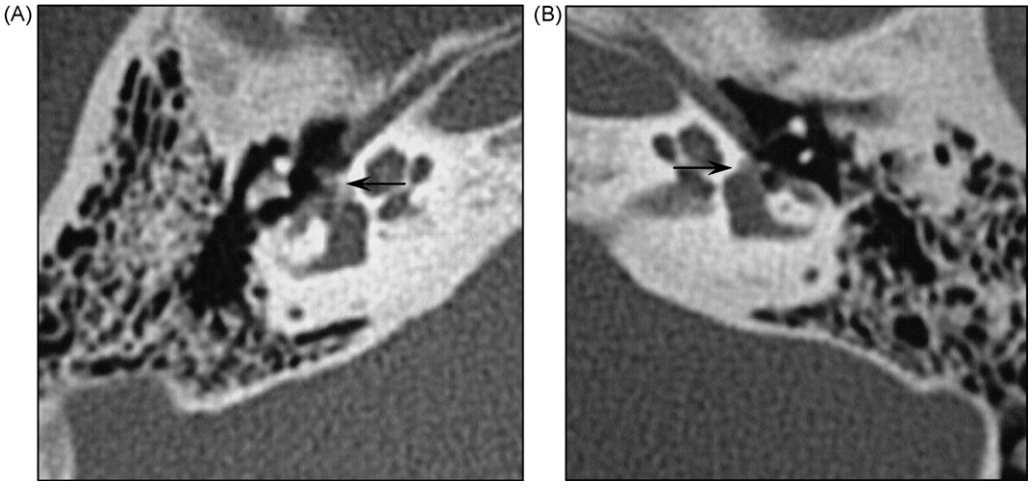

Fig. 2 (A and B) Axial computed tomographic scan of a 13-year-old girl (Patient #2) with bilateral otosclerosis. Black arrows indicate, on both sides, poorly calcified foci in the cochlear capsule just anterior to the oval window.

Table 1 Clinical and CT-scan data[a]

Patient ID	Gender	Deaf ear	CT evidence of otosclerosis	Age at surgery (years)	Surgery (year)	Observations at tympanoscopy	Preop ABG (dB)	Postop ABG (dB)
1	Female	Left	NA	15	Stapedectomy, 1992	Anterior FP otosclerotic focus	35	2.5
2	Female	Left	+	13	Laser Stapedotomy, 1996	Obliterative focus	16.3	15
		Right	+	14	Laser Stapedotomy, 1997	Obliterative focus	10	7.5
3	Female	Left	+	18	Laser Stapedotomy, 1996	Anterior FP otosclerotic focus	32.5	8.7
		Right	+					
4	Male	Left	+	15	Laser Stapedotomy, 2000	Obliterative focus	20	2.5
		Right	+					
5	Female	Right	+	14	Laser Stapedotomy, 2000	Entire FP otosclerotic focus	26.3	2.5
6	Female	Right	−	9	Laser Stapedotomy, 2002	Entire FP otosclerotic focus	32.5	3.8
		Left	−	12	Laser Stapedotomy, 2005		18.8	6.3
7	Female	Right	−	12	Laser Stapedotomy, 2003	Entire FP otosclerotic focus	33.8	3.8
		Left	−	14	Laser Stapedotomy, 2005	Entire FP otosclerotic focus	30	5

[a] NA, not-available.

was reported although one girl complained of tinnitus. All of the patients operated on received at least 12 months of follow-up (range: 2−15 years; mean: 7.4 years). Hearing results are presented in Table 2. The mean (standard deviation) postoperative air-bone gap was 6.5 dB (±3.7). The mean closure was 19 dB (±11.2). The mean change in high-tone bone conduction level was 1.8 dB (±7.5). ABG postoperative was inferior to 10 dB in 9 cases while in one it was inferior to 20 dB. The Speech Reception Threshold improvement was 13.7 dB (±14.5).

4. Discussion

Otosclerosis occurs in less than 0.6% of the population before the age of 5 years and in only 4% between the ages of 5 and 18 years [1]. It is therefore rarely seen in young patients referred to pediatric otolaryngologists and radiologists specializing in the evaluation of CHL. This low incidence explains the uncertainties concerning the optimum management of this cause of hearing impairment with the attendant risks of inadequate diagnosis and treatment.

This survey suggests that the CT-scan is helpful for establishing the preoperative diagnostic of otosclerosis. We performed a CT-scan in 6 children and found evidence of otosclerosis in 4 of them (a total of 8 ears). Then, in children with otosclerosis but normal CT-scan, this rather low sensitivity of CT-scan must be underlined. In adults, the sensitivity of CT-scan is higher than 85%, so that density measurement of fissula ante fenestram does not permit the diagnosis of all temporal bones with otosclerosis [6].

Table 2 Audiometric patterns in 7 juvenile otosclerosis patients

Patient ID	Deaf ear	Preop bone conduction/preop air conduction				Postop bone conduction/postop air conduction			
		0.5 kHz	1 kHz	2 kHz	3 kHz	0.5 kHz	1 kHz	2 kHz	3 kHz
1	Left	15/40	5/50	10/45	10/45	15/15	5/5	5/10	10/15
2	Left	10/40	20/35	20/30	20/30	20/40	30/40	40/50	35/55
	Right	10/25	10/30	20/20	25/30	15/20	20/25	35/40	25/40
3	Left	−5/45	15/45	0/25	5/30	5/20	30/35	5/20	20/20
4	Left	25/60	20/55	40/50	35/35	20/40	25/25	25/30	25/35
5	Right	0/40	5/35	0/20	0/15	10/10	15/20	10/10	5/10
6	Right	5/60	20/55	20/40	15/35	10/20	15/20	10/10	10/10
	Left	5/45	15/40	20/20	10/20	5/15	5/20	15/15	15/15
7	Right	15/55	10/55	10/35	5/30	10/10	10/10	5/5	0/15
	Left	10/50	15/50	15/50	10/30	10/20	10/20	5/5	5/5

These CT scanning limits giving false-negative data correspond to small hypoattenuation areas. Nevertheless, the radiographic appearance was the same as seen in adults [7]: the association of foci of otosclerosis and otospongiosis in the absence of any congenital or acquired abnormalities of the malleus, incus or stapes.

Otosclerosis involved the margin of the OW without narrowing its aperture and was associated with poorly calcified foci located at the margin of the OW, near the *fissula ante fenestram*. As has been described in adults [7,8], we observed in 4 cases, hypodensities extended to the promontory and surrounded the labyrinthine capsule. Nevertheless, we previously showed a high prevalence of such pericochlear hypodensity in this age group without clinical correlation [9]. In the youngest patient, tympanotomy suggested otosclerosis (an otosclerotic focus in an entire footplate associated with ankylosis of the stapes) however preoperative CT-scan failed to reveal any abnormality. Nevertheless, CT-scan is mandatory since it furnishes the best anatomic details for planning the surgical approach and thus avoiding unfortunate intra-operative events secondary to variations in the course of vessels and nerves [10]. It is also useful for detecting any potential communication between the middle ear and the subarachnoid space in order to prevent potential surgical complications. Advanced scanning techniques can allow visualization of abnormalities of the footplate and both crura of the stapes in children and, in addition, can provide magnified views of both the inner and middle ear. The low-dose scanning technique minimizes the irradiation level and provides good definition for distinguishing the different tissues encountered. As illustrated in our cases, it can give useful details on axial sections as well as reformatted coronal images.

Preoperative confirmation of otosclerosis with appropriate imaging has therapeutic implications but this indication has not been previously mentioned in published data concerning otosclerosis in children. Indeed, in cases of congenital stapedial fixation, it can predict good results instead of poor results after surgery [11]. In addition, it eliminates the non-negligible risk of peri-lymphatic gusher associated with these congenital abnormalities and an even greater risk in certain situations [12]: males, defect in the internal auditory canal and pre-existing sensorineural hearing loss.

It remains uncertain whether stapes surgery in children can be an effective procedure for correcting CHL due to stapedo-vestibular ankylosis. In addition, since children and parents commonly choose hearing aids in lieu of surgery [13], data on the surgical outcome is lacking. The absence of any consensus on the optimum management of this disorder in children adds to these uncertainties. Millman, in a series of 40 stapedectomies in children and adolescents younger than 21 years [14], reportedly closed the air-bone gap to within 10 dB in 58% of the ears and to within 20 dB in 90% of the ears. These good results are consistent with other reports in the literature [3,4,11,13–19] and are comparable to our results (Table 3). As reported in stapedectomies in adults, the long-term results are equivalent when the immediate postoperative gaps are compared [14]. On the other hand, while stapes surgery in otosclerosis is highly effective, the rare complications encountered can be devastating. Hence, the opening statement in House's 1980 review of the subject [16]: "Stapedectomy on a child? Never!" However, according to this author, we believe that age was not the main reason for the very poor outcome described in one child [16]. The concept of "no touch" was the idealistic goal for stapes surgery, before laser surgery became available. Our previously published studies confirmed the benefits of laser stapedotomy in adults [20,21]. In all procedures performed, laser stapedotomy in young patients has been technically successful and has minimized mechanical trauma to the footplate.

Table 3 Results of published pediatric otosclerosis studies[a]

References	Ears	Time of survey (years)	Age groups, mean ±S.D. (years)	Postop ABG <10 dB, %	Surgical technique
[16]	24	5	9–18, NA	92	24 S
[15]	62	NA	6–20	77	62 S
[19]	35	20	5–18, 13.7	100	35 S
[17]	9	3	5–19, NA	56	4 S, 5 s
[14]	40	18	7–21, 16.7 ± 3.2	58	40 S
[13]	60	35	7–17, 13.1 ± 3.3	92	60 S
[11]	95	14		82	
[18]	11	6	9–17	63	11 S
Author's case-series	10	13	9–17, 13.6 ± 1.9	90	1 S, 9 ls

[a] S indicates stapedectomy; s, stapedotomy; ls, laser stapedotomy; NA, not-available.

Stapes laser surgery has enhanced the safety of the surgical procedure and is particularly adapted to surgeons (pediatric otolaryngologists) with limited experience. This technique can thus help keep postoperative complications to a minimum. Nevertheless, the stapedotomy carried out by an experienced otosurgeon, with or without laser, remains still the most reasonable strategy to expect a good functional result.

Finally, it should be stressed that postoperative recommendations after stapes surgery in children are exactly the same as those following other procedures in pediatric ear surgery: limit straining and vigorous activity, avoid heavy lifting, sniffing or wiping by obstructing both nostrils and to be very cautious around water. Both the child and parents should be instructed to alert the surgeon if any symptom or sign suggesting peri-lymphatic fistula appears (dizziness or vertigo, sudden hearing loss).

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