Juvenile otosclerosis: radio-clinical features and outcomes of surgical treatment

Otospongiose juvénile: aspects radio-cliniques et résultats du traitement chirurgical

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ABSTRACT

Purpose: To review the radio-clinical features and to evaluate the outcomes of surgery approaches for the treatment of otosclerosis in pediatric population.
Methods: It's a retrospective study including 12 patients under 18 years of age, diagnosed with otosclerosis and using data over a 24 – year –period (1996-2020). A clinical examination, an audiometric assessment and a computed tomography of the temporal bone (CT-Scan) prior to surgery had been performed in all cases. All patients underwent surgical treatment under general anesthesia. Follow –up was essentially clinical and audiometric with a mean period of 4years.
Results: Mean age of our patients was 15.3 with a ratio of 0.5. Only three of them had a family history of otosclerosis. Major functional sign was hearing loss; tinnitus was noted in only 5 cases. CT-Scan had shown typical radiographic evidence of otosclerosis grade Ia (Veillon classification) in 9 cases, grade Ib in 1 case and no abnormalities in 2 cases. Surgery was performed in all cases: 8 patients underwent stapedotomy and 4 had stapedectomy. The audiometric results were good, air-bone gap closure to within 10 dB was achieved in 84% of cases (10 cases) and to within 20dB in 100% of cases at last follow-up.
Conclusion: Audiometric and radiological assessments are essential to guide the diagnosis and the treatment of juvenile otosclerosis. Stapes surgery is a good option for closing the air-bone gap in children with bilateral juvenile otosclerosis.
Keys –words: Otosclerosis, Children, Hearing loss, Stapes surgery

RÉSUMÉ

But: Etudier les aspects radio-cliniques et évaluer les résultats de la chirurgie dans le traitement de l'otospongiose juvénile.
Méthodes: Il s'agit d'une étude rétrospective incluant 12 patients âgés de moins de 18 ans suivis pour otospongiose entre 1996 et 2020. Un examen clinique, une évaluation audiométrique ainsi qu'une tomodensitométrie ont été réalisés en préopératoire chez tous les patients. Ils ont tous eu un traitement chirurgical sous anesthésie générale. Le suivi post-opératoire était essentiellement clinique et audiométrique avec un recul moyen de 4 ans.
Résultats: L'âge moyen des patients était de 15,3 ans avec un ratio à 0,5. Trois patients avaient des antécédents familiaux d'otospongiose. Le principal signe fonctionnel retrouvé était l'hypoacousie alors que les acouphènes n'ont été notés que dans cinq cas. La tomodensitométrie a montré des signes radiologiques d'otospongiose grade Ia selon la classification de Veillon chez neuf patients, grade Ib chez un patient et était normale chez deux patients. Tous les patients ont été traités chirurgicalement: huit patients ont eu une platinotomie et quatre ont eu une platinectomie. Les résultats audiométriques étaient satisfaisants, une fermeture du Rinne à 10 dB a été obtenue pour 84% des patients (10 patients) et à 20dB pour 100% des patients jusqu'au dernier contrôle.
Conclusion: Une évaluation audiométrique et radiologique est essentielle afin de guider le diagnostic et le traitement de l'otospongiose juvénile. La chirurgie stapédienne semble être une bonne option pour la fermeture du Rinne chez les enfants avec une otospongiose bilatérale.
Mots-clés: Otospongiose, Enfants, Surdité, Chirurgie stapédienne

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INTRODUCTION:

Otosclerosis is a primitive osteodystrophia of the labyrinthine bone. It is commonly diagnosed in the third to fourth decade of life. The onset of hearing loss is usually after puberty. Juvenile otosclerosis is rarely reported. Just as adult population, young patients frequently present to otolaryngology services with progressive hearing loss and tinnitus with normal tympanic membranes at otoscopy. Otosclerosis along with otitis media are the main causes of conductive hearing loss in the pediatric population [1]. Therapeutic options for children with otosclerosis consist in stapes surgery or hearing aids. Surgery has been consistently debated.

The purpose of this study was to review radio-clinical features and to evaluate the outcomes of surgery approaches for the treatment of otosclerosis in pediatric population.

METHODS:

We conducted a retrospective study including 12 patients under 18 years of age, with otosclerosis treated during the period from January 1996 to December 2020. Age, sex, family history, previous surgical approach, time of onset of symptomatology and the side of hearing loss were reviewed. The diagnosis was based on a history of hearing loss, physical examination, otoscopy, audiometric and radiographic findings. All patients had undergone surgical treatment. The surgery was done under general anesthesia. A transcanal procedure was chosen in all cases. Per-operative data were examinated. Hearing results were evaluated using pre- and postoperative audiometry. The criteria of success were closure of the airbone gap to within 10 dB of the preoperative bone conduction in the speech frequencies (500 to 2000 Hz). Follow-up for a minimum one year with clinical examination and audiogram for all patients was required.

RESULTS:

The mean age of the patients was 15.3 years (12- 17) with a ratio of 0.5 (8 girls and 4 boys). Only three children had a family history of otosclerosis. One child had a history of acute otitis media. There was a mean period of hearing impairment of 5.9 years. Two patients (22.2%) had unilateral hearing loss. Bilateral hearing loss was found in 10 patients (83.3%). In all cases, hearing loss was the main symptom. Tinnitus was noted in five cases; it was unilateral in one case and bilateral in four cases. General examination was abnormal in a single child and showed facial dysmorphia, prognatism and convergent strabismus. Otoscopic examination was normal in all cases. Audiometric assessment showed that two patients had a mixed hearing loss, with a threshold of 90 dB and 60dB, and 10 patients had a conductive hearing loss. Mean threshold was 44.17 dB. The evaluation of preoperative audiometric bone conduction revealed that 10 children had a normal cochlear reserve and two children had respectively 70 dB and 40 dB as bone conduction thresholds across 500,1000,2000 frequencies. Stapedial reflex was absent in all cases. Speech audiometry was practiced in 9 cases with a mean threshold of 50.2. No disturbance of intelligibility has been found in all cases. All the children had a CT-scan before surgery: in nine cases, it showed typical radiographic evidence of otosclerosis grade Ia (Veillon classification) with isolated involvement of the footplate, which is thickened (> 0.6 mm) and hypodense while one patient had isolated anterior fenestral hypodensity grade Ib (Figure 1).

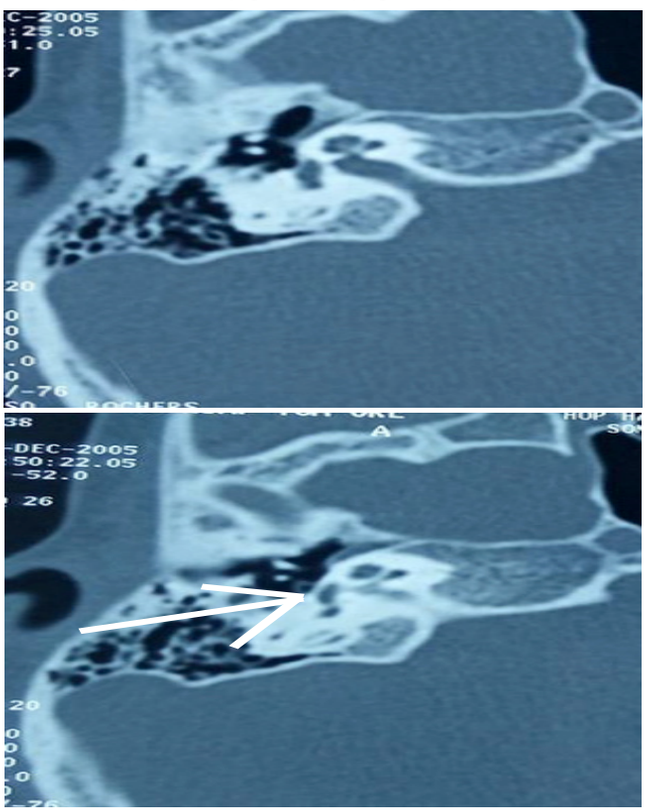

Figure 1 (a and b) Axial computed tomographic scan of a 16-year-old girl with otosclerosis; isolated thickening of footplate hypodensity (↑)

CT-Scan was normal in two cases with no abnormalities detected. In addition, CT-scan found no abnormalities in the malleus or incus or any potential risk of a peri-lymphatic gusher during footplate opening. The surgical treatment was performed on all patients under general anesthesia, there were no concurrent anomalies of the stapes superstructure, malleus or incus malformations. The facial nerve had a normal course in all operated ears. A total stapedectomy was performed in 4 cases and stapedotomy was done in 8 cases.

All the patients showed immediate improvement after surgery and no facial palsy nor prolonged tinnitus. One patient complained of post operative vertigo.

All operated patients have at least 12 months of follow-up (1 – 10 years). The mean follow–up time was 4 years. Hearing results are presented in Table I. Postoperative air-bone gap closure to within 10 dB was achieved in 83.3% of cases and to within 20dB in 100% of cases at last follow-up. The mean postoperative air-bone gap was 5.83 dB [0-20]. The mean air bone gap gain was 35.82 dB [30-55] (Figure 2). Mean air conduction threshold after surgery was 16.66dB [10-60].

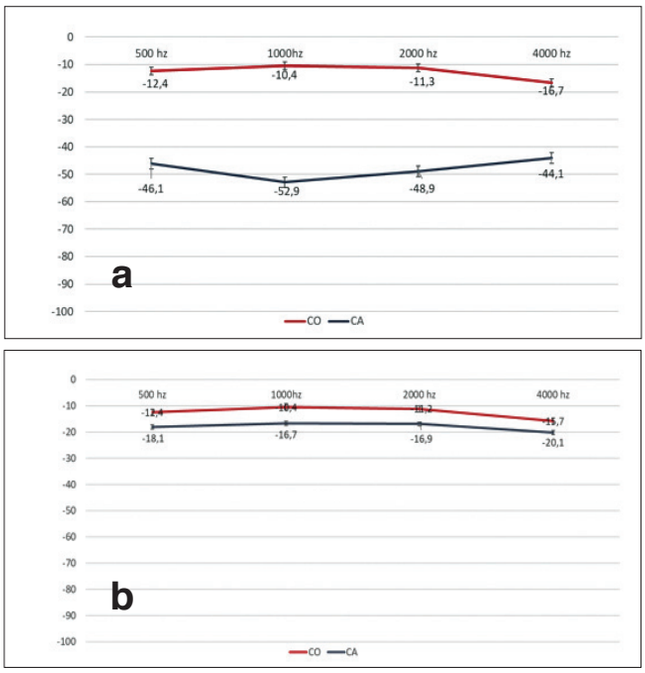

Figure 2: a: Mean air conduction threshold and air bone gap before surgery for each frequency.
b: Mean air conduction threshold and air bone gap after surgery for each frequency.

Furthermore, success rate remained stable over the course of follow-up time. Postoperative sensorineural hearing loss didn't occur in any case.

Table I:
Postoperative results for patients with juvenile otosclerosis.

	Age (years)	Air conduction thresholds before surgery (dB)	Air bone gap before surgery	Air conduction thresholds after surgery (dB)	Air bone gap after surgery	Air bone gap gain
1	12	90	30	60	0	30
2	16	60	60	20	20	40
3	14	40	40	0	0	40
4	16	30	30	0	0	30
5	14	55	55	0	0	55
6	16	55	55	0	0	55
7	14	50	50	20	20	30
8	17	55	40	10	0	40
9	16	40	30	10	0	30
10	13	50	40	20	10	30
11	17	50	40	20	10	30
12	17	60	40	40	20	20

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 [2]. The incidence of juvenile otosclerosis is low, but it is significant when we consider that 15.1% of patients who have undergone stapedectomies developed their hearing losses before the age of 18 [3]. In our study, all patients were aged 12 to 17 with a mean age at the time of surgery of 15. In the study reported by R. Vincent [4] the mean age was 14.4 years (range 8 to 18 years) while G. Namyslowski reported a study of 13 patients suffering from otosclerosis, the children were aged 9 to 17 years. There is a very high incidence of bilateral otosclerosis (92%) when the symptom of hearing loss appears underage of 18 [3].
The etiology of otosclerosis remains undetermined, but hereditary tendency, endocrine factors, immune disorders, viral involvement, and connective tissue disorders are advanced as potential causes of otosclerosis. The sex difference has been reported in many studies with highest incidence in women confirmed by R. Vincent [3,5]. In our study, we found a higher incidence in women with a sex-ratio of 0.5.
The differential diagnosis of juvenile otosclerosis includes all conditions with clinical conductive-type hearing loss; otitis media with effusion, suppurative otitis media, congenital cholesteatoma, ossicular chain anomalies, connective tissue disorders and all conditions with pediatric stapes footplate ankylosis such as congenital stapes fixation (CSF), tympanosclerosis, minor aplasia and less commonly Paget's disease and osteogenesis imperfecta [2,6,7].
The diagnosis is suspected on a family history, otoscopic examination, audiometric results and radiologic findings. Progressive hearing loss is the main symptom, associated frequently to tinnitus. CT-scan is an excellent procedure for preoperative evaluation; assessing the status of the oval window, the inner ear, the internal auditory canal, the vestibular and cochlear aqueducts, the ossicles and the course of the facial nerve [2]. It demonstrates the same appearance seen in adults: the foci of otosclerosis in the absence of any congenital or acquired abnormalities of the malleus, incus or stapes. These foci involve the margin of the oval window without narrowing its aperture and with poorly calcified foci located at the margin of the oval window, near the fissula ante fenestram. A high prevalence of pericochlear hypodensity was noted in this age group without clinical correlation. In adults, the sensitivity of CT-scan is higher than 85%,on the contrary many false negative data were found in pediatric population and they correspond to small hypo attenuation areas [2,8].
Surgical management of juvenile otosclerosis is controversial. Stapedectomy is still rarely performed in children younger than 16 years. Many treatments were suggested including anti-enzyme or bone resorption moderating drugs and personal Sound Amplification Devices (*PSAD*). Surgery is not recommended in children below the age of 5 years; in such cases, PSAD are the treatment of choice. Stapedotomy is controversial after the age of 5 years [9]. Pediatric otologists delay the surgery preferring the participation of young patients in the decision, especially in unilateral cases because of the risk of postoperative sensorineural hearing loss caused during surgery as the inner ear is opened [10]. Many authors doubt the validity of surgery in very young patients because of the particularly evolutive nature in this age group [11]. Arguments against pediatric stapedectomy also include a higher incidence of Eustachian tube dysfunction, otitis media, increased risk of refixation, greater risk of stapes gusher and resultant sensorineural hearing loss [11]. Thus, hearing aids are safer and offer a very good rehabilitation[10]. The otologists who prefer surgery argue that juvenile otosclerosis represents a more aggressive process and if stapedectomy is delayed until

adulthood, surgery will be more challenging with a higher likelihood of obliterative disease requiring ‘‘drill-out’’ [12]. Hence, therapeutic approach depends on the child’s own motivations, on whether they can minimize activities in the immediate postoperative period and on their ability to understand the risk information [10]. Recommendations after surgery include limiting straining and vigorous activity, avoiding heavy lifting, sniffing or wiping by obstructing both nostrils and to be very cautious around water. Both the child and parents should be educated about these precautions to avoid complications [2,13].

The results of primary stapedotomy in children with otosclerosis are comparable to the results of primary stapedotomy in adult otosclerosis patients. Various studies have shown that stapedotomy adequately treats conductive hearing loss in up to 90% of these patients [14]. Robinson reported high success percentages with an air-bone gap closure to within 10 dB in 93% of stapedotomies and air-bone gap closure within 20 dB in 98% of stapedotomies: 80% of his patients have an excellent cochlear reserve and do not show a deterioration of sensorineural function following stapedectomy [4]. Similar results were published in several studies. Lescanne [2] published a serie of 7 patients with juvenile otosclerosis, 10 of the ears operated on, 8 surgical procedures were performed under general anesthesia and 2 under local anesthesia. In his study, the mean closure was 19 dB. Air bone gap postoperative was inferior to 10 dB in 9 cases, while in one it was inferior to 20 dB.

In our study, Surgery was performed in all cases: 8 patients underwent stapedotomy and 4 had stapedectomy. Air-bone gap closure to within 10 dB was achieved in 83.3% of cases (10 cases) and to within 20dB in 100% of cases at last follow-up.

Stapes surgery can be performed endoscopically. Anthony et al [15] compared microscopic and endoscopic surgery in a study of 22 endoscopic and 52 microscopic surgery all performed in juvenile population. This study found that Improvement in pure-tone average (25.9 dB vs 18.5 dB, p =.382) and air bone gap (21.7 dB vs 14.7 dB, p =.181) was similar. Post-operatively, the median air bone gap was 11.3 dB and 15.0 dB for endoscopic and microscopic cases (P =.703) respectively. Air bone gap closure to 20 dB (72.7% vs 65.2%, p =.591) was also similar.

Age not only influences the immediate postoperative result, but also the future course of hearing thresholds[16]. Many causes may explain delayed sensorineural hearing loss after surgery such as perilymph fistula, middle ear granulation, aseptic labyrinthitis, iatrogenic trauma or acute labyrinthitis, especially in children that are prone to acute otitis media [6]. In a long-term study of children with otosclerosis, a gradual mean sensorineural hearing loss of 0.7 dB per year was also observed post-stapes surgery, which is likely due to cochlear otosclerosis [17,18].

Conclusion: Audiometric and radiological assessments are essential to guide the diagnosis and the treatment of juvenile otosclerosis. Stapes surgery is a good option for closing the air-bone gap essencially in children with bilateral juvenile otosclerosis.

Compliance with ethical standards

Conflict of interest: The authors stated that there is no conflict of interest.

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