CASE REPORT

Juvenile otosclerosis: a case presentation and review of the literature

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SUMMARY

Otosclerosis in childhood and adolescence or juvenile otosclerosis is a rare disorder resulting in conductive hearing loss. A 9-year-old boy presented to our clinic, suffering from moderate hearing loss. According to his parents, his hearing acuity had progressively deteriorated over the past 2 years. Otoscopy and tympanometry revealed bilateral secretory otitis media and the patient underwent bilateral grommet insertion. However, he continued to report of hearing loss and a right exploratory tympanotomy was performed. Stapedial fixation was confirmed, being compatible with juvenile otosclerosis, and we proceeded to a right stapedotomy. One year later, follow-up showed satisfactory outcome with an air-bone gap closure to 10 dB. Juvenile otosclerosis with the coexistence of persistent secretory otitis media can be overlooked. Affected children from 9 years of age are strongly motivated to undergo stapes surgery for juvenile otosclerosis, following parental consent.

BACKGROUND

Otosclerosis in childhood and adolescence or juvenile otosclerosis is a rare clinical entity. In adults, otosclerosis is a well-known cause of conductive hearing loss (CHL) mainly affecting patients aged between 15 and 45 years. However, microscopic foci of otosclerosis have been found in 0.6% of autopsies of individuals less than 5 years of age.¹

With regard to the genetic background, juvenile otosclerosis is a multifactorial disease, associated with autosomal dominant inheritance with incomplete penetrancy of approximately 40–45%.² In adults, stapedectomy is the treatment of choice, in which the stapes is surgically removed and replaced with a prosthesis. In children, stapedectomy remains a challenging procedure and a ground of dispute with questions about the indicated age of operation, the operating technique and the management of possible complications. Alternative conservative treatment options are sound amplification with a hearing aid, a bone anchored hearing aid or a 'watch-and-wait' strategy.

The aim of this study is to report on a child suffering from juvenile otosclerosis, obscured by secretory otitis media, and to review the literature on this topic.

CASE PRESENTATION

A 9-year-old boy presented to our clinic, suffering from moderate hearing loss. According to his parents, the hearing loss had progressively

deteriorated over the past 2 years, while he had fully developed normal speech. No family history of hearing loss was reported. The clinical examination revealed bilateral secretory otitis media and tympanometry revealed a type B pattern. Pure tone audiogram showed CHL with air-bone gap of 45 dB in the right ear and 22 dB in the left ear (figure 1A). The patient underwent examination of the ears under general anaesthesia, bilateral myringotomy and grommet insertion, during which a significant amount of glue was aspirated; a small section of the adenoids was also removed at the same time. Two months later, the child still reported of hearing loss and pure tone audiogram demonstrated slight improvement of only 10 dB of the ABG in all frequencies (figure 1B). High-resolution CT scan did not show any otosclerotic foci around the stapes footplate, and no congenital dysplasia of the external and middle ear.

DIFFERENTIAL DIAGNOSIS

The high degree of hearing loss and the progressive character of the hearing loss led us to suspect that the cause of CHL was not limited to secretory otitis media. The differential diagnosis mainly included congenital stapes fixation and juvenile otosclerosis. Juvenile otosclerosis was more likely, as it tends to affect patients older than 6 years and is often associated with progressive hearing loss. On the contrary, congenital stapes fixation appears in younger patients and does not involve progressive CHL; facial nerve dysfunction and mild ossicular anomaly may also be encountered.

CT scan did not demonstrate fusion of the middle ear ossicles to the epitympanum or stenosis/aplasia of the external auditory meatus, so congenital disorders such as Crouzon's disease, Marfan syndrome, Treacher Collins syndrome, achondroplasia, Apert syndrome, Pierre Robin syndrome, Duane syndrome and otopalatodigital syndrome were excluded. In addition, the clinical examination, completed with paediatric evaluation, did not show any facial and/or skeletal deformities. Moreover, CT scan did not show abnormal dilation of the internal acoustic canal, which could result in increased perilymphatic pressure. This sequence can be the trigger to the observed 'gusher' during the opening of the stapes footplate (X-linked deafness-2).

Less common causes of CHL in children include stapes fixation from osteogenesis imperfecta (presence of blue sclera), tympanosclerosis (usually with a history of chronic otitis media), congenital cholesteatoma and juvenile Paget's disease.

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Figure 1 (A) Preoperative audiogram, depicting bilateral conductive hearing loss, more significant on the right side. (B) Pure tone audiogram after insertion of the ventilating tubes.

TREATMENT

In order to verify the cause of the conductive deafness, an exploratory tympanotomy was planned, which confirmed fixation of the incudostapedial joint and stapes footplate. No incus and malleus abnormalities were identified, and both anterior and posterior crus were present. There was no obvious otosclerotic focus around the oval window or the stapes footplate. Moreover, although facial nerve malposition is reported in cases of congenital stapes fixation, perioperatively, the facial nerve was neither overlying the oval window niche nor coursing downwards to the oval window. The family was informed about the treatment options in case of juvenile otosclerosis, including a hearing aid trial, and they opted for surgical treatment. After an endaural incision, a right stapedotomy was carried out. Excision of the stapedius muscle tendon and removal of the stapes suprastructure were performed. A 0.7 mm perforation on the footplate was performed manually and a 4.75 mm Teflon piston was anchored to the long process of the incus. Temporalis fascia was put around the piston so as to prevent any leak from the oval window. The whole procedure was performed using compatible cold instruments. The surgeon opted for leaving the grommets in place in order to prevent secretory otitis media recurrence.

OUTCOME AND FOLLOW-UP

The postoperative period was uneventful. Follow-up audiogram 3 weeks after surgery depicted an air-bone gap closure to 20 dB. The second follow-up, 1 year postoperatively, showed further improvement with an air-bone gap closure to 10 dB (figure 2); the otoscopy confirmed the extrusion of tubes and the tympanic membranes had normally healed. The parents were also informed about the possibility of stapedotomy on the left side, as the pure tone audiogram and an absent stapedial reflex on that side were compatible with juvenile otosclerosis.

DISCUSSION

Secretory otitis media is the most common cause of CHL in children and is characterised by fluid accumulation behind the tympanic membrane, which remains after the resolution of acute otitis media, once the acute inflammation has resolved. In our case, juvenile otosclerosis was overlooked since secretory otitis media is more common in children and could also cause

conductive hearing loss. The persistence of conductive deafness after the insertion of ventilating tubes and the high degree of hearing loss led us to suspect adjacent juvenile otosclerosis, so the child underwent exploratory tympanotomy, which confirmed the diagnosis.

The first series of paediatric stapedectomy was published by House in 1980, where characteristically the author concluded mentioning 'Stapedectomy on a child? Never'.³ However, although reports of the results of this surgery for otosclerosis in children are rare, a review of the literature confirmed that stapedectomy could also be performed in this population. Table 1 displays these reports and their results.³⁻¹⁵ The reason for such delay of stapes surgery in children, although it is a common procedure in adults, is the risk of severe sensorineural hearing loss, the great incidence of otitis media and the risk of refixation. Alternative conservative treatment options are amplification with a hearing aid, a bone anchored hearing aid or a 'wait-and-watch' strategy.

Figure 2 Satisfactory air-bone closure 1 year postoperatively.

Table 1 Air-bone gap closure (dB) in various studies after stapedectomy for juvenile otosclerosis

	Age (years)	N (cases)	ABG preop	ABG postop
Murphy and Wallis ⁴	≥5	9	11–20 N=1 21–30 N=4 31–40 N=3 41–50 N=1	0–10 N=5 11–20 N=4
Welling <i>et al</i> ⁵	3 to 17	4	41	13.1±3
Denoyelle <i>et al</i> ⁶	>10	6		<10 N=4 10–20 N=1 lost N=1
An and Lee ⁷	>5.9	6	36±10.9	8.7±6.0
Neilan <i>et al</i> ⁸	6.7–18	6	34.4	7.2
Lescanne <i>et al</i> ⁹	9–18	10	25.52	6.5±3.7
House <i>et al</i> ³	9–18	24		22<10 1 no improvement 1 dead ear
Cole ¹⁰	6–20	62	–	77% ABG closure <10 dB
Robinson ¹¹	5–18	35	–	100% ABG closure <10 dB
Millman <i>et al</i> ¹²	7–21	40	–	58% ABG closure <10 dB
Lippy <i>et al</i> ¹³	7–17	60	–	92% ABG closure <10 dB
Namyslowski <i>et al</i> ¹⁴	9–17	11	–	63% ABG closure <10 dB
Vincent <i>et al</i> ¹⁵	NA	41	–	93% ABG closure <10 dB

NA, not applicable.

One year postoperatively, our patient showed improvement with an ABG closure to 10 dB. Our review of the literature regarding the postoperative results showed a wide variation in the ABG closure. A very good result (ABG closure <10 dB) was noted in 55–100% of children (mean 77%; [table 1](#)). A good result (ABG 11–20 dB) was reported in 5–45% of children (mean 33%; [table 1](#)). These hearing results are consistent with those in adults, although slightly worse, as approximately 85% and 15% of adults have demonstrated ABG <10 dB and ABG 11–20 dB, respectively.¹⁶ This may occur because children and teenagers have a much higher incidence of footplate pathology and diffuse otosclerosis with oval window obliteration and cochlear involvement compared to the general stapedectomy population.^{2 3 8 17}

Fenestration on a fixed footplate may be performed with the help of a perforator, microdrill, pick or piezoelectric device. Although the laser technique is not yet used for stapes surgery in our clinic, it could be used for children as it is used for adults.^{2 3 6 8 17} Lescanne *et al*⁹ successfully performed laser stapedotomy on six children suffering from juvenile otosclerosis; in the same study, it was contended that laser stapedotomy could minimise mechanical trauma to the footplate and lessen postoperative complications as well. Recently, Fang *et al*¹⁸ presented a meta-analysis on laser versus non-laser stapedotomy. The authors concluded that there was a significant difference between the two methods with regard to the ABG closure, with the former achieving significantly better hearing results. According to the authors, the laser technique reduces the risk of mobilisation of the footplate and maintains a bloodless operating field.¹⁸ On the other hand, some disadvantages of lasers worth mentioning are the thermal effects on deeper structures and the difficulty in reaching certain structures outside the direct visual field, especially when it comes to paediatric stapedotomy.¹⁹

Reviewing the literature, the encountered complications during and after stapes surgery for juvenile otosclerosis are rare and limited to sporadic cases. Perioperatively, House described

persistent stapedial artery in two cases and abnormally placed facial nerve in one case.³ De la Cruz reported perilymphatic gusher during surgery in two children, attributed to possible wide communication between the subarachnoid and perilymphatic spaces through the fundus of the internal auditory canal; however, in that study, not mentioned was which condition the two children suffered from, juvenile otosclerosis or congenital stapes fixation.²⁰

In the short-term postoperative period, Neilan *et al*⁸ described profound hearing loss associated with acute vertigo 3 weeks after stapes surgery in a child aged 10 years; in the revision stapedectomy, they found a long prosthesis, which was removed and replaced with a shorter one, with no sign of infection but also without hearing improvement.

In the long-term follow-up, CHL developed in a child 8 years after the primary stapes surgery,¹² in which the revision surgery revealed a displaced prosthesis, which was subsequently replaced and the ABG closed. Worsening in sensorineural hearing loss has been reported to occur with a mean of 0.7 dB per year for 25 years after stapes surgery;¹³ Millman *et al*¹² reported even better cochlear reserve and nerve function as they showed cochlear deterioration of only 1 dB to 2 dB over an average of 25 years. Although the hearing results of stapes surgery in children may worsen over time, children who underwent this surgical procedure did not show increased ABG over time.⁷

The minimum age for safe stapes surgery is still under discussion. In [table 1](#), the minimum age is demonstrated, indicating that children as young as 3 years old could be operated with satisfactory outcomes. Prior to scheduling stapes surgery, one should take into account the child's motivations and intention to follow the postoperative recommendations (limiting intense activity, not blowing against blocked nostrils, avoiding heavy lifting). Some authors suggest waiting for the child to reach an age where he or she is able to make an informed decision before stapes surgery. During this period of time, an alternative option is a hearing aid trial. As in our case, children aged 9 years are strongly motivated to undergo stapes surgery for

juvenile otosclerosis, although stapedotomy could be equally effective for those under this age.

A matter of paramount importance in the whole treatment process is consultation and the offered management options. It is essential to explain to parents that children undergoing stapes surgery for juvenile otosclerosis have about an 80% or better chance to close the ABG to within 10 dB, which is in accordance with the first author's experience. The surgeon should mention that there is also a small chance of profound hearing loss as well as progressive sensorineural hearing loss over the next several years. With regard to age of operation, there are no specific guidelines. Most surgeons prefer to offer a hearing aid trial, especially to young children, rather than rushing to surgery. However, it is preferred to operate at an age around 10 years, when the child can understand the information given and minimise the activities as required, postoperatively.⁶

Learning points

- ▶ Secretory otitis media is the most common cause of conductive hearing loss in children. Since juvenile otosclerosis is also a cause of conductive hearing loss, it may be overlooked when it coexists with secretory otitis media. It is advisable to suspect juvenile otosclerosis in case of persistent conductive hearing loss even after the treatment of secretory otitis media.
- ▶ Stapedotomy can be performed in children as young as 3 years of age, but is better at an age of around 10 years, when the child can understand the information given and minimise his postoperative activities.
- ▶ It is essential to explain to parents that children undergoing stapes surgery for juvenile otosclerosis have about an 80% or better chance to successfully close the air-bone gap to within 10 dB.

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reviewed and revised the manuscript, and approved the final manuscript as submitted.

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