Stapedotomy in Congenital Stapes Ankylosis with Mobile Footplate: A Case Report

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Patient: Female, 15-year-old
Final Diagnosis: Fixation of the middle ear ossicles
Symptoms: Conductive hearing loss of the right ear of up to 40 dB • tomography confirmed the presence of the middle ear ossicles

Medication: —
Clinical Procedure: —
Specialty: Otolaryngology

Objective: Congenital defects/diseases
Background: This paper presents a case of a 15-year-old child with a rare congenital anomaly of the middle ear in which the stapes was fused to the medial wall of the tympanic cavity.

Case Report: This defect coexisted with partial malleus fixation in the attic and caused conductive hearing loss at an average level of 35-40 dB. Two exploratory tympanotomies were performed, where excess bone between the stapes and promontory was removed and the head of the malleus was released in the attic. The good effect of these procedures was unstable, mainly due to re-attachment of the stapes to the medial wall of the tympanic cavity. At the next surgery it was decided to perform stapedotomy, despite the mobile stapes footplate. The operation was performed with a small-fenestra stapedotomy technique. Perforation of the footplate was done using a microdrill with a balanced speed. A KURZ prosthesis with a diameter of 0.5 mm was used. The postoperative period passed without any complications. Three and 6 months after the surgery, control pure tone audiometry was performed and showed significant improvement in hearing thresholds. During the follow-up period of more than 6 months, hearing improvement remained stable.

Conclusions: We concluded that it was safe to perform stapedotomy in the presence of a mobile stapes footplate when congenital anomaly of the stapes superstructure caused its severe fixation in the middle ear. In our opinion, in a child with congenital ear anomaly, consideration should be given to the multifocal origin of the hearing loss.

Keywords: Stapedotomy • Congenital Ear Anomaly • Middle Ear Malformation • Stapes Surgery • Hearing Loss

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Background

Conductive hearing loss in children is usually caused by episodes of otitis media with effusion [1]. When diagnosing a child suffering from hearing loss but with no symptoms of otitis media or trauma, the possibility of a congenital anomaly should be considered, especially if the eardrum appears normal. Congenital anomalies of the middle and inner ear that cause hearing loss are rare, but recently have been diagnosed more often. That is undoubtedly the result of the greater availability of specialized devices and the growing experience of professionals. As hearing loss caused by a congenital anomaly can be severe and may negatively affect social interactions, it is important to minimize its adverse effects. Surgical treatment is the preferred treatment since it can permanently improve hearing. This is especially important for young people as it eliminates the need to wear hearing aids. However, surgical treatment is not possible in all cases of anomalies, due to the limitations resulting from the depth of the malformation and the risk of complications.

Ear defects in children are extremely diverse. When discussing hearing impairment in this group of patients, one should not forget otosclerosis, which can also occur in younger individuals. The therapeutic option is then stapedotomy [2,3], which can improve quality of life [4]. The most common middle ear anomalies are deformations of the long process of the incus [5]. Other include malleus – incus fusion, ossicular chain adhesions with the bony wall of the tympanic cavity [6], and those associated with fixation of the stapes and its footplate [7,8].

When qualifying a young patient for middle ear reconstructive surgery, the potential benefits to hearing and the operational risks should be carefully considered. There are some inoperable malformations, such as those associated with the inner ear hydrops [9,10]. In such cases, opening the inner ear may cause inner ear fluid leakage (gusher) with the risk of deafness and meningitis. High-resolution computed tomography (HRCT) of the temporal bones is extremely helpful in diagnosis of these anomalies.

Case Report

A 15-year-old girl was admitted to the clinic due to suspicion of hearing loss in the right ear. There was no inflammation or effusion. Family history was irrelevant. Physical examination showed no abnormalities and the appearance of the tympanic membrane was normal in both ears. Audiometric results showed conductive hearing loss of the right ear of up to 40 dB (Figure 1, black lines).

Due to a suspicion of congenital anomaly of the middle ear, HRCT of the temporal bones was performed. The tomography confirmed the presence of middle ear ossicles. A linear structure was present between the malleus and the anterior wall of the epitympanum, possibly representing an adhesion or ossification in this area (Figure 2). In addition, thickening of the posterior crus of the stapes and discreet widening of the incudomalleolar joint was observed (Figure 3).

The child was qualified for an exploratory tympanotomy of the right ear. The procedure was performed at the age of 15 years. During the surgery, it was discovered that the superficiation of the stapes was completely fixed due to the presence of abnormal bone tissue between the stapes and the promontory. The malleus was also partially fixed in the attic. Ossiculoplasty was performed with removal of excess bone tissue blocking the stapes in the oval window niche and with mobilization of the malleus in the epitympanum (transcanal approach, removing the adhesions with a small hook after a limited widening of the lateral attic wall). The mobility of the stapes footplate was good. The postoperative course was uneventful. For about 2 months the patient had improved hearing, but subsequent hearing examination 3 months after the procedure showed almost no change in comparison to the condition before surgery (Figure 1, red line). The child was therefore qualified for re-operation, which was performed the same year.

During the second surgery, mobilization of the stapes was performed since it had again adhered to the medial wall. The excess bone was removed with a drill. Additionally, some adhesions around the head of the malleus were removed to re-establish its mobility in the attic. Mobility of the stapes footplate was again assessed as very good. However, 3 months after the second procedure, audiometric examinations did not show any improvement in hearing (Figure 1, yellow lines), suggesting that the stapes had scarred quickly and stiffened again.

After in-depth analysis of the case, it was decided to undertake another surgery, which was finally carried out when the child was 16 years of age. During the surgery, re-attachment of the stapes to the medial wall, in the form of a bone block covering the lower and middle part of the ossicle, was observed. This abnormal tissue was removed, improving the mobility of the ossicle. The stapes footplate had good mobility in the oval window. Considering the scope and course of previous surgeries, it was decided to remove the superficiation of the stapes and perform stapedotomy. Its crura were cut and the ossicle. The malleus was also partially fixed in the attic. Ossiculoplasty was performed with removal of excess bone tissue blocking the stapes in the oval window niche and with mobilization of the malleus in the epitympanum (transcanal approach, removing the adhesions with a small hook after a limited widening of the lateral attic wall). The mobility of the stapes footplate was good. The postoperative course was uneventful. For about 2 months the patient had improved hearing, but subsequent hearing examination 3 months after the procedure showed almost no change in comparison to the condition before surgery (Figure 1, red line). The child was therefore qualified for re-operation, which was performed the same year.

After removal of the crura, a band of abnormal, previously invisible bone tissue was revealed, appearing low, just next to the footplate of the stapes from the promontory side. A hole in the mobile footplate of the stapes was made using a 0.6-mm
Figure 1. The patient’s pure tone audiometry, before and after surgeries: black lines: before operations; red lines: after first operation, yellow lines: after second operation. Green lines: after stapedotomy. Note almost unchanged bone conduction.

Figure 2. Right ear HRCT, sagittal view. A linear structure present between the malleus and the anterior wall of the epitympanum (white arrow). It was the cause of the malleus head immobilization in the attic.

Figure 3. Right ear HRCT, coronal view. Thickening of the posterior crus of the stapes (white arrow).
diamond burr. A KURZ Skarżyński-type prosthesis with a diameter of 0.5 mm was inserted and fastened to the long crus of the incus. The area around the prosthesis was sealed with venous blood clot. The postoperative period passed without complications. At 3 and 6 months after the surgery, a control PTA was performed and showed significant and stable improvement in hearing (Figure 1, green lines).

Discussion

Cases of fixation of the middle ear ossicles, especially the stapes, are not common. When suspecting this condition and considering the surgery, it is important to determine its pathogenesis. It could be inflammatory, resulting from a congenital anomaly, or may be the result of juvenile otosclerosis. The presence and surgical treatment of juvenile otosclerosis has been well documented in the literature since House’s research in 1980 [11]. The results of this treatment are generally good and do not differ much from those obtained in adults [12,13]. Since juvenile otosclerosis is a separate problem, as is otitis media with effusion, we do not describe it in detail here (although both are common causes of conductive hearing loss).

When discussing congenital anomalies of the stapes, it should be mentioned that fixation of this ossicle may have a different etiology. The most common is congenital fixation of the stapes footplate. Less frequent are isolated defects of the superstructure, and even less frequent is its immobilization due to the inner ear hydrops. Congenital fixation of the stapes footplate is a condition described by many authors as one that should be differentiated from juvenile otosclerosis [7,8,14]. Fixation of the stapes footplate is one of the most common isolated defects of the middle ear [15], and its surgical treatment is similar to that of otosclerosis, although the postoperative results are slightly worse [16]. The stapes footplate defects (apart from fixation) may also be related to stapes underdevelopment. The footplate can be partially membranous, with the risk of vertigo when middle ear pressure is changing [17]. The spectrum of congenital anomalies of the stapes varies widely. Park et al described 66 patients, distinguishing 16 subtypes depending on the complexity of the defect [18]. Abnormalities of the structure took the form of a stapes with only 1 crus present, deformed crura, missing part of the ossicle, lack of continuity with the incus, underdevelopment of the footplate, and even the absence of a stapes with the footplate (agenesis). Approximately 80% of these cases were related to fixation of the stapes footplate and the treatment included stapedotomy or stapedectomy. Ossiculoplasty was performed in cases where the stapes footplate was mobile.

Two situations must be distinguished: the first, in which the stapes is fixed within its footplate, and the second, in which the stapes footplate is mobile and only the upper part of the ossicle is immobile. In the literature, the term ‘stapes fixation’ generally refers to the immobilization of the stapes footplate along with the entire ossicle. That term can cause confusion because a case of isolated fixation of the stapes superstructure can also be referred to as ‘stapes fixation’. However, these 2 are different situations, and cases with isolated fixation of the stapes superstructure are rare. Nandapalan and Tos describe a similar case to the one discussed in this paper, where the stapes superstructure is in the form of a single block adhering to the promontory [19]. They treated the patient by removing the excess bone tissue, with good results. Lee et al described the attachment of the stapes to the facial nerve canal, in the absence of the long crus of the incus [20]. In both cases, the footplate of the stapes was mobile. Other rare stapes anomalies found in the literature included a case of elongated pyramidal eminence causing adhesion of the ossicle [21] and a familial stapes fixation to the medial wall [22,23].
The efficacy and safety of surgical treatment of stapes fixation as a result of juvenile otosclerosis or congenital footplate fixation is generally good. There are many papers in the literature in this field [11,18,24]. However, we found no papers evaluating the results of stapedotomy in a case of a congenital stapes anomaly with isolated fixation of its superstructure and with a mobile stapes footplate. This procedure is technically difficult and can be risky. However, this surgery was performed here because the effects of previous ossiculoplasties were unstable. The presented results clearly show that correctly performed surgery can have good results, with improvement of hearing.

It should also be emphasized that the observed “fixation” of the stapes may be in fact the result of the inner ear hydrops. This situation is encountered in some cases of enlargement of the vestibular aqueduct [25] or incomplete partition type III [26]. These cases require proper diagnosis and should not be treated surgically, because surgical intervention will cause gusher with the risk of hearing loss and meningitis. Our experience indicates that the risk of gusher exists even when radiological results are normal and there are no indications of its occurrence before surgery. The surgeon must be prepared for such a situation.

Conclusions

The result of stapedotomy in a child with congenital anomaly of the stapes superstructure and its fixation in the middle ear were good. The presence of a mobile stapes footplate was not a contraindication for stapedotomy. In this patient, the fixation of the malleus in the attic was also observed, so consideration should be given to the multifocal origin of the hearing loss in a child with congenital anomaly.

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